Behrooz A. Akbarnia
Muharrem Yazici
George H. Thompson
Editors

The Growing Spine

Management of Spinal Disorders in Young Children

Second Edition

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As with our first edition, the driving force behind the second edition of The Growing Spine textbook is our **patients and their families**. This book would mean nothing without their need for our care and expertise. We therefore dedicate it to our patients for their trust and participation and for giving us the opportunity to learn from experience and to share the knowledge we have gained with others. It is our sincere hope that this book will continue to inspire those who care for young children with early-onset scoliosis and thoracic deformities, leading to better outcomes and improved quality of life.

Much has changed over the past 5 years, and we believe this second edition textbook reflects our better understanding of the issues and advances in the field. We owe a great deal of gratitude to many who helped us reach this level of understanding, including our specialty societies: the Scoliosis Research Society (SRS), the Pediatric Orthopaedic Society of North America (POSNA), the European Pediatric Orthopaedic Society (EPOS), and the American Academy of Orthopaedic Surgeons (AAOS). These organizations endorsed the International Congress on Early Onset Scoliosis (ICEOS) meeting, supporting both the educational effort during their meetings and the formation of a task force for pediatric devices where their collaboration with regulatory agencies and the industry has enabled an environment for innovation and change. The ability to bring diverse groups together to achieve the common goal of improving our patients' quality of life has led to an enriching experience, one which we have attempted to share on the pages of this book.

Many thanks to our colleagues in North America and around the world at both the Growing Spine Study Group (GSSG) and the Children Spine Study Group (CSSG) (including investigators and study coordinators), as well as to the faculty and participants at ICEOS. These important groups supported the creation of a forum of exchange addressing issues surrounding very young children with spine and thoracic deformities and thereby advancing the research and knowledge in this field. Thanks also to the related specialties showing increased interest and participation in teaching at

ICEOS, including pulmonology, cardiology, anesthesiology, radiology, and genetics. The list is too long to mention all specialties, but suffice it to say that the help, expertise, and guidance of our specialist colleagues have been invaluable. We particularly wish to thank the Growing Spine Foundation (GSF) board and staff, who have supported our work with assistance, financial support, and guidance over the years.

This book would not be in your hands without our dedicated, world-expert authors. We are grateful to them for their contributions and for their invaluable efforts to prepare excellent chapters and to respond to multiple queries and in a timely manner.

Last, but not least, we are grateful to our associates and staff for putting up with us throughout the preparation of this book; we give them all special thanks and are deeply indebted to them for their support.

And now, most importantly, a special note from each of us to our respective families:

Behrooz Akbarnia, MD*:* My deepest gratitude, as always, to my wife Nasrin, for her continuing support, love, and encouragement throughout the completion of the second edition. Thanks also to my three children and their spouses and, last but by no means least, to my four wonderful grandchildren—Simia, Kian, Leila, and Luca—the last two born after the publication of the first edition. They were always a source of inspiration and energy for me to continue the work on this book in spite of the time it took away from them.

Muharrem Yazici, MD*:* The greatest of thanks should go to my family Ruya, Yildiz Naz, and Mehmed Emir. I have stolen time from them, and although I have, on more than one occasion, chosen academia over personal life, they have never complained and always provided their love and support for me.

George H. Thompson, MD*:* As with the first edition, my thanks and appreciation to my wife, Janice, for her support and encouragement in completing the second edition. She and our children and six grandchildren were deprived of my attention during the editing of this edition. They never complained as they understood the importance of completing this project due to the improved care for others that it will provide.

Foreword 1

The growth of the spine demonstrates symmetry in all dimensions to reach a harmonious balance not only for volume and mass (cosmetics) but also for function (motion) during growth as well as when growth is completed. This is especially important for the development of the thoracic cage and subsequent pulmonary function.

Spinal pathology from any etiology can affect growth and development in different ways. For example, if the disorder affects a very limited area, such as a localized congenital malformation (i.e., unbalanced hemivertebra or unsegmented bar), a limited operative procedure (i.e., hemivertebra excision) may have significantly different results. Localized congenital deformities may result in a significant deformity with growth, while a limited procedure may correct a deformity and allow adequate development of the spine. In other situations, the etiology may affect a larger part, if not the entire spine such as in neuromuscular, syndromic, or idiopathic deformities (infantile idiopathic scoliosis). For such situations, it must be remembered that any surgical procedure on a still growing spine will create a disturbance in the growth. Sometimes the spine may not be well balanced in three dimensions and may not be reversible following an extensive early fusion.

The other point is that scoliosis is mainly a horizontal (coronal) plane deformity where the most important mechanism is spinal torsion or rotation. Until now, surgically speaking, we have developed devices able to correct the collapsing spine with elongation or distraction rods or by growth modulation devices with a compression mechanism used anteriorly on the apical vertebral bodies on the convex side. But as yet we do not have true derotation devices.

This is why, until now, the best three-dimensional correction for a spinal deformity in a growing child, especially in the thoracic area without congenital malformations, is by repeated or serial casts and bracing (more or less mixed over time). These are used until appropriate definitive posterior spinal fusion and segmental spinal instrumentation can be performed close to the end of growth.

Despite that, some situations require surgical treatment during growth, particularly early onset or at a young age, and this book will give some proposals and even some solutions for such cases. But the main goal of this book is to advise the reader to evaluate carefully and have a clear understanding of the pathophysiology, natural history, and treatment options before treating an "early-onset scoliosis" case. We need to think about the consequences of

immediate and delayed treatment, either nonoperative or mostly operative, with its subsequent and sometimes irreversible dangers or complications. Remember that operating on a growing spine can result in repeated surgeries with exponential complication risks. Perform surgery only when truly necessary.

Finally, this book will open your mind regarding the need for further research, which is always necessary to help the children who are depending on our care.

Paris
March 2015

Jean Dubousset

Foreword 2

This is the second edition of *The Growing Spine*, being published 4 years after the first edition in 2011. With the publication two questions are posed – why so soon, and what is different?

The field of EOS has expanded greatly with the Study Groups and the ICEOS meetings, as well as the knowledge disseminated with the first edition of *The Growing Spine*, all of which helped establish the growing spine as a recognized subspecialty in the field of spinal surgery. More physicians and clinics are concentrating on the care of these challenging problems. In addition, the SRS Growing Spine Committee Early Onset Scoliosis Consensus statement published earlier this year covers the problem well (1). The definition of EOS, "spine deformities that is present before the age of 10 years of age," is stated, and in addition the organization of EOS into diagnostic categories, evaluation, treatment goals, and treatment options is laid out. With the great expansion in knowledge and interest in the field, the editors viewed the explosion of knowledge in the field as being so rapid that the first edition is not current.

The second edition has been expanded to 57 chapters with international authors comprising 30 % of the total authorship. The chapters have been revised and updated with the addition of new chapters. The organization of the chapters into different sections is very helpful and follows the outline of the consensus statement above. There are certain chapters that still represent the definitive work of definitive experts in that particular area:

Chapter 4 on "Normal Growth of the Spine and Thoracic Cage" by Dr. Alain Dimeglio

Chapter 5 on "Normal Lung Growth and Thoracic Insufficiency Syndrome" by Dr. Gregory Redding

Chapter 16 on "Neurofibromatosis" by Dr. Alvin Crawford

Chapters 17 and 18 on "Spine Deformities in Syndromes" by Dr. Paul Sponseller

Chapter 29 on "Casting for Early-Onset Scoliosis" by Dr. James Sanders

Chapter 38 on "Traditional Growth Rods" by Drs. George Thompson and Behrooz Akbarnia

Chapter 39 on "VEPTR Expansion Thoracoplasty" by Dr. Robert Campbell

The new chapters add to the knowledge of the growing spine and are an impressive addition to the text. They expand on areas partially addressed in

the first edition, as well as add additional knowledge. Notable among these are:

Chapter 7 on "New classification System" by Dr. Michael Vitale

Chapter 15 on "Intraspinal Pathology" by Drs. Nejat Akalin and Amer Samdani

Chapter 50 on "Complications Following Distraction-Based Growing Technique" by Dr. John Emans

Chapter 52 on "Anesthetic Considerations in Growing Children and Repetitive Anesthesia" by Dr. Lena Sun

The surgical management is well covered, with chapters on "traditional surgery" as these children do not always present at a time when growth-friendly surgery can be performed. The coverage of the techniques for growth-friendly surgery is extensive with many choices presented, both proven and innovative.

The most important consideration is the outcome of the treatment as pointed out by Dr. Vitale in Chapter 55 assessing the radiographic, pulmonary, and HRQOL results. It is important to remember that the critical time is at the end of growth and later when the children become adults.

It is an honor and pleasure to be asked to write the foreword to the second edition of *The Growing Spine*, and you have in your hands the latest comprehensive knowledge in this growing unique field.

1. Skaggs DL, Guillaume T, El-Hawary R, et al (2015) Early onset scoliosis consensus statement, SRS growing spine committee. 2015. Spine Deformity 3:107.

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Preface to the Second Edition

The field of spinal deformities in young children is one of the newer areas in spinal surgery literature. For a long time, early-onset spine deformities have had to endure neglect by researchers and only recently achieved priority in the scopes of spinal deformity surgeons. However, despite this late start, early-onset deformity literature has accelerated and gained popularity at a greater rate, and this early neglect has been largely negated in the appearance of an explosive number of epidemiological, clinical, and experimental studies in the past 5–10 years.

Undoubtedly, the collaborative efforts of physicians joined together under the auspices of the Growing Spine Study Group and Children's Spine Study Group and International Congress on Early Onset Scoliosis (ICEOS) meetings held annually since 2007 have had stimulating effects on this newly kindled research effort. However, we should not be remiss in mentioning the 2010 first edition of this book, which has contributed greatly to the recognition of the growing spine as an independent subspecialty in the practice of spinal surgery.

Our book, the first edition of which was eagerly perused by the spinal surgery reader, was sold out quickly and among thousands of Springer-published textbooks and soon took its place on the top rungs of the most-downloaded list of eBooks. It was even later translated into Chinese and is being translated to other languages. Both due to this quick depletion of printed books and the rapid evolution of information in this field, preparations for the second edition began only 3 years after its initial publication, a time period that can be considered quite short for scientific medical textbooks. The edition you hold in your hands today boasts extensive reviews of all of its chapters and includes all-new developments that have found their place in the EOS literature recently in the form of updates and in all-new chapters. The editorial process was performed even more diligently, with close attention to the congruity of sections written by authors across the globe to achieve an integrated text. As distinct as this book is by presenting the classical and emerging ideas on the growing spine, it is also unique in that it boasts exceptional international contributions.

The dissemination of scientific knowledge is in constant flux. As soon as a research project is turned into written word, it is doomed to become outdated. Given time, doubtless the information that has found its way onto the pages of this book will suffer that very same fate. However, we, the editors, are confident in our belief that our book will provide current and satisfying

answers to basic questions posed by clinicians and academicians interested in the field.

Although all three editors have reviewed all chapters meticulously, mistakes overlooked are inevitable in a text of this scope. All accolades for the book belong with the authors. We are grateful to them for their invaluable and diligent contributions. All errors and omissions, however, are the responsibility of the editors. For these, we beg your forgiveness and understanding. Please do not hesitate to share with us any and all errors and omissions that you note in this book, thereby allowing us to correct them in future editions.

Finally, without the hard work of our dear friend Pat Kostial, RN, BSN, the publication of this book would not have been possible. There are no words to convey our gratitude to Ms. Kostial for her indispensable help. With Michael D. Sova's transparent, collaborative work style and diligence, who worked with us for this edition, obstacles disappeared and impossibilities only took a little time. We consider ourselves very lucky for having had the opportunity to work with such a team. It would also not have been possible to print these sentences without the professional approach of Springer, which is doubtless among the world leaders in the publication of scientific medical literature.

La Jolla, CA, USA — Behrooz A. Akbarnia, MD
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Preface to the First Edition

Early onset scoliosis (EOS) is a major topic in pediatric spine deformity today. These challenging deformities occur in almost all differential diagnostic categories.

Unfortunately, each diagnosis has a different natural history, making it even more demanding. This is the first textbook on this topic. It is a compilation of the current concepts of evaluation and treatment of the various deformities of the growing spine.

We have tried to explore the normal growth of the spine and other associated organs as well as natural history of the various differential diagnostic categories and possible treatment options. It is anticipated that this textbook will need to be updated every 2–3 years in the future as concepts and treatment guidelines change. Treating the spinal deformity is not the major issue, but controlling the deformity to allow for growth of the spine and the associated organ systems, such as pulmonary, cardiac, and gastrointestinal, is the major goal. Controlling deformity allows for improved spinal growth of the involved child and the controlling associated development of these organ systems. A short trunk has an adverse effect on these organ systems. As a consequence, EOS requires a multidisciplinary care. It involves genetics, pediatrics, pulmonology, cardiology, neurology, neurosurgery, as well as orthopaedic surgery.

Treatment options for very young children are controversial. Bracing, serial Risser casts, and surgery (growth modulation and the use of distraction-based or growth-guided techniques such as growing rods) are explored in this textbook.

Preliminary treatment results have demonstrated that growth-friendly surgical techniques are effective in controlling or modulating curve progression and allowing for spinal growth. Spinal growth allows for improved capacity of the thoracic and abdominal cavities. Cosmesis is less than ideal as crankshaft remains a significant problem even in the growing rod systems. Surgical treatment complications are high, particularly infection and implant failure, especially rod breakage. Management of complications is an important aspect of the treatment of EOS. Because of the high complication rate, it is important to make the right decision regarding patient and family selection. They must be cooperative and understanding and be willing to be cooperative during the postoperative period.

Future research is important. The Growing Spine Study Group (GSSG) and other databases will hopefully guide future investigations. Only by

defining the results of treatment in a relatively large volume of children over a long period of time can the true effectiveness of each of these techniques be determined. Predicting who will worsen, improving spinal tethers to control progressive deformities and the development of self-expanding or remotely controlled devices that would obviate the need for repeated surgical procedures.

We thank our contributors who are all specialists and experts in a variety of areas involved with early onset scoliosis. We also acknowledge the contribution of the members of Growing Spine Study Group who have continuously provided the information that is the basis for a significant portion of the data presented in this book.

Special thanks for assistance in preparing and organizing this textbook are to Sarah Canale and Pooia Salari; without their assistance, the completion of this project would have been very difficult.

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Part I

General

Embryology and Anatomy: Spine/ Spinal Cord

1

Shay Bess and Breton Line

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Key Points

1. Development of the spine and spinal cord begins during the third week of gestation.
2. Early development includes formation of the axes of the embryo, formation of primitive neural tissue, and notochord development.
3. The axial skeleton eventually arises from the somites, and normal vertebral and neural formation is dependent upon normal development of the paraxial mesoderm and somites.
4. Errors in the formation of the paraxial mesoderm and somites and errors in the formation of the cartilaginous precursors to the vertebrae and neural arch structures lead to congenital scoliosis and spinal dysraphism conditions, as well as abnormalities in other developing organ systems.
5. Neurocentral joints allow continued growth of the spinal canal, and secondary vertebral ossification centers persist until the third decade of life.

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1.1 Early Development

The initial development of the spine begins during the third week of gestation. The embryo at this stage of development exists as a two-cell layered structure called the bilaminar germ disc. On approximately day 15, a groove forms in the midline of the germ disc called the primitive groove. The primitive groove forms an initial deepening at the cranial end of the embryo and then extends caudally and grows along the length of the germ disc forming the cranial and caudal axes of the embryo. This central deepening is termed the primitive pit, and the collection of cells that surround the primitive pit forms the primitive node (Fig. 1.1). The head of the embryo eventually forms at the primitive pit and primitive node. The entire structure (primitive pit, node, and groove) is called the primitive streak. The primitive streak establishes the embryonic longitudinal axis, giving rise to left and right sides of the embryo. Therefore, the cranial/caudal, left/right, and ventral/dorsal axes are formed during this third week of gestation.

A three-layered embryo is formed by the proliferation and migration of epiblast cells through the primitive streak (Fig. 1.2a–c). Epiblast cells invade and replace the hypoblast cell layer, forming the definitive endoderm. Migration of epiblast cells between the epiblast and endoderm layers continues, forming a third cell layer, mesoderm. Upon establishment of the mesodermal layer, the epiblast is renamed the ectoderm or ectodermal layer.

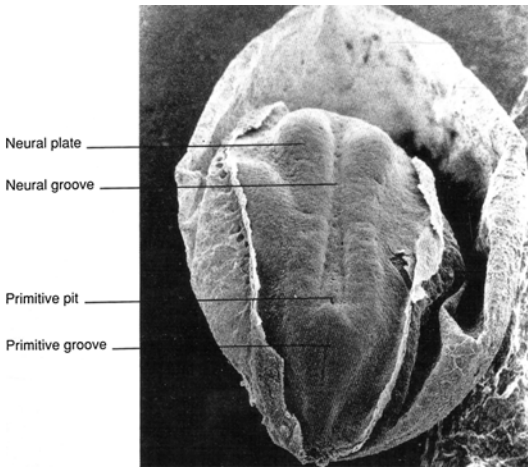

Fig. 1.1 Photomicrograph of primitive streak in the bilaminar germ disc. The primitive pit, primitive groove, and primitive node form the primitive streak. The head of the embryo will eventually form at the primitive pit and primitive node, and the entire structure (the primitive streak) establishes the embryonic longitudinal axis (Adapted from Tamarin (1983) With permission from John Wiley & Sons)

Two midline structures develop in the mesoderm: the prechordal plate and the notochordal process. The notochordal process begins as a hollow mesodermal tube and goes on to become a solid rod structure, called the notochord. The notochord induces the formation of the vertebral bodies, and subsequently, the vertebral bodies coalesce around the notochord inducing the notochord to form the nucleus pulposus (Fig. 1.3a, b).

Following the development of notochord, three distinct structures form in the mesoderm: the paraxial mesoderm, intermediate mesoderm, and lateral plate mesoderm. As pertains to the spine and spinal cord, the paraxial mesoderm, which lies adjacent to the notochord, gives rise to cell lines that form the critical structures called the somites. The somites are responsible for formation of the axial skeleton, voluntary musculature, and the skin dermis (Fig. 1.4). The intermediate mesoderm and lateral mesoderm are involved in the development of the urogenital and cardiopulmonary systems. As a consequence, defects that alter the development of the mesoderm resulting in vertebral abnormalities may also result in concurrent abnormalities in the urogenital and cardiopulmonary systems. VACTERL syndrome is an acronym with each letter representing an associated defect secondary to abnormalities in the mesodermal development including Vertebral anomalies, imperforate Anus, Cardiac abnormalities, TracheoEsophageal fistula, Renal dysplasia, and Limb malformations. Approximately 30–60 % of vertebral abnormalities diagnosed in childhood will have an additional organ system abnormality, with the genitourinary system most commonly involved. This underscores the need to evaluate for additional organ system involvement in children and infants with congenital vertebral abnormalities, including cardiac and renal ultrasound.

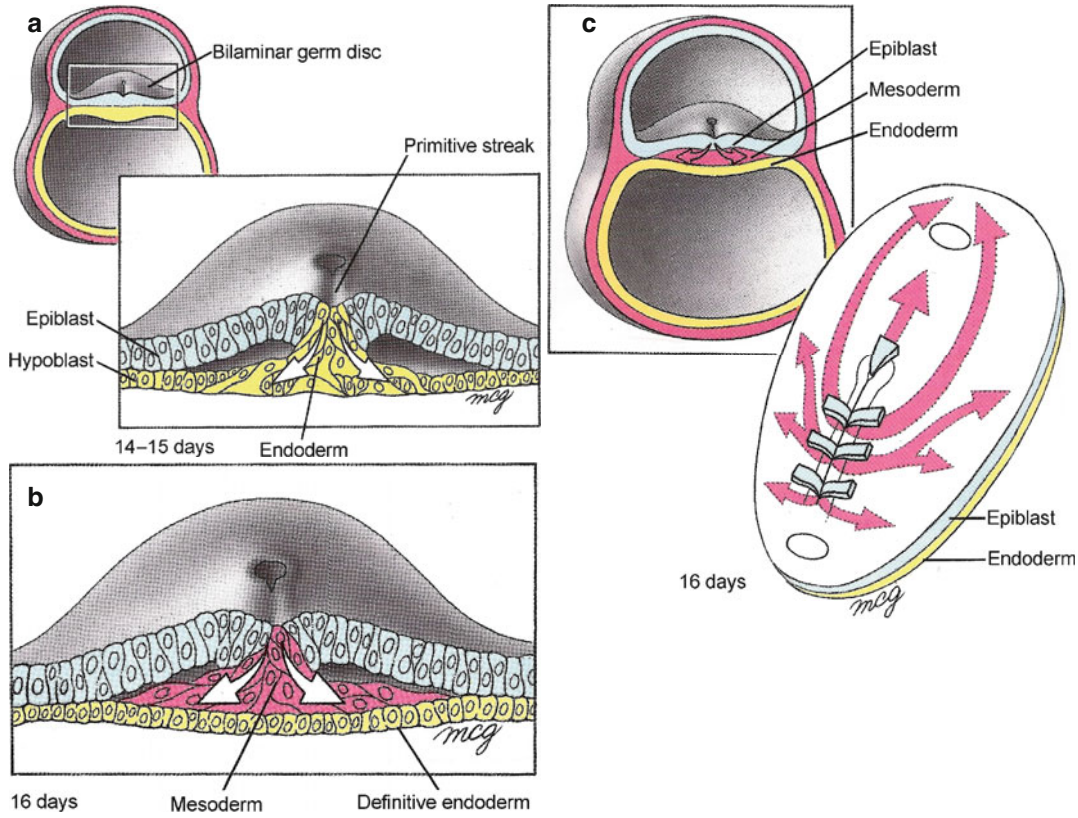

Fig. 1.2 (**a**–**c**) Proliferation and migration of epiblast cells. Epiblast cells proliferate and migrate through the primitive streak eventually forming the endoderm, mesoderm, and ectoderm; the definitive three-cell layered embryo (Adapted from Larsen (1993). With permission from Elsevier)

1.2 Somite Formation and Differentiation

The axial skeleton, voluntary muscle, and the dermis of the neck and trunk are derived from the somites. The somites emerge as paired on approximately gestational day 20, arising from the paraxial mesoderm and developing in a cranial to caudal fashion at a rate of approximately 3–4 somites per day (Fig. 1.5).Initially 42–44 somite pairs exist adjacent to the notochord. The cranial-most somite pairs eventually form the base of the skull and extend caudally to a rudimentary structure, the embryonic tail. However, caudal 5–7 somites regress, leaving a total of 37 somite pairs for development. Somite pairs 1–4 form the occiput as well as the bones of the face and inner ear. Somites 5–12 form the cervical spine (there are eight cervical somites but ultimately only seven cervical vertebrae because the first cervical somite participates in occiput formation). Somites 13–24 form the thoracic vertebrae, somites 25–29 form the lumbar vertebrae, and somites 30–34 form the sacral vertebrae. The remaining three terminal somite pairs form the coccyx and persist after regression of the terminal embryonic tail. The consecutive somite pairing on the embryo creates an anatomic template that organizes the vertebral alignment and the corresponding peripheral nervous system (PNS), which persists to maturity.

As the embryo develops, the somites separate into subdivisions. Accordingly, the ultimate tissue structure that develops from each somite is produced from the respective somite

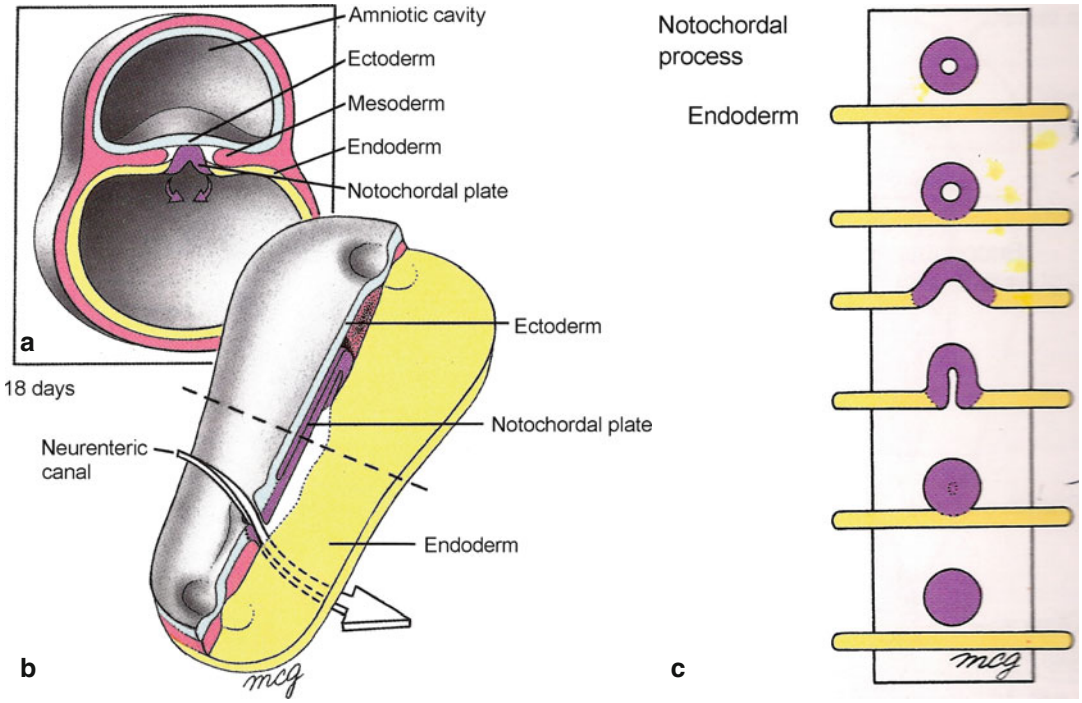

Fig. 1.3 (**a**–**c**) Formation of the notochordal process and notochord. The hollow notochordal process forms within the mesoderm and goes on to form the solid notochord. The notochord induces vertebral body formation and eventually becomes the nucleus pulposus (Adapted from Larsen (1993). With permission from Elsevier)

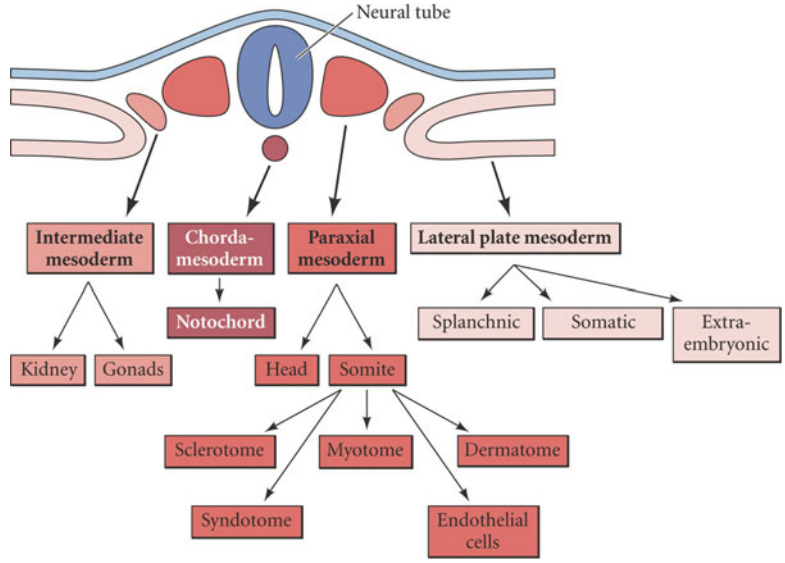

Fig. 1.4 Paraxial mesoderm, intermediate mesoderm, and lateral plate mesoderm formation, location and eventual structures (Adapted from Larsen (1993). With permission from Elsevier)

subdivision. The first somite subdivision that appear are the sclerotomes. The sclerotomes ultimately give rise to the bony spinal column. Sclerotomes are formed when a hollow central cavity forms within the somite. This cavity develops in the medial region of the somite adjacent to the midline notochord and neural tube. The central cavity fills with cells, termed

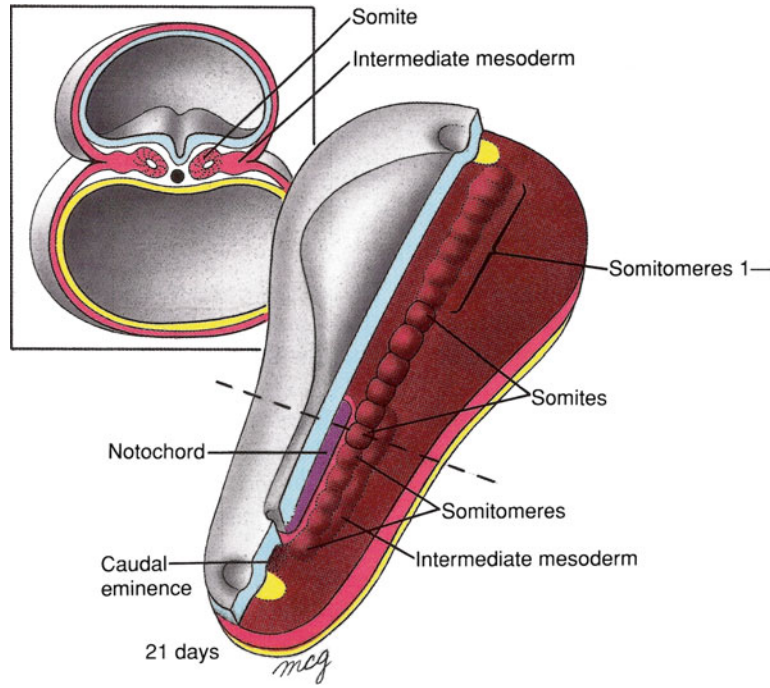

Fig. 1.5 Somite formation. The paired somites arise from the paraxial mesoderm and form the axial skeleton, voluntary muscle, and the dermis of the neck and trunk (Adapted from Larsen (1993). With permission from Elsevier)

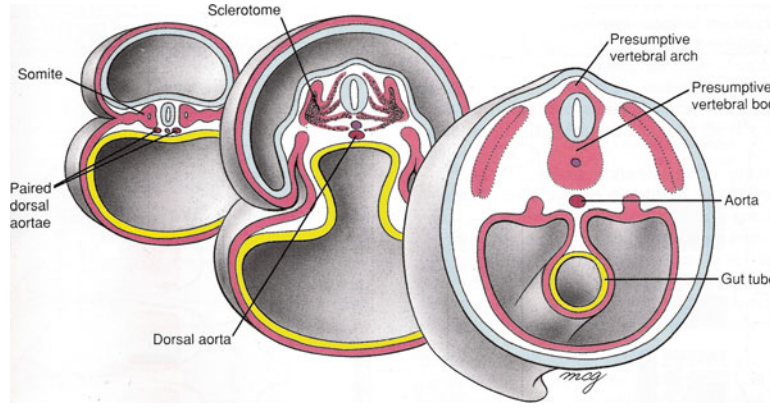

Fig. 1.6 Sclerotome formation. The central cavity within the somite fills with loose core cells and eventually ruptures. Core cells migrate toward the midline and envelop the notochord and neural tube forming a sclerotome. The ventral sclerotome forms the vertebral body, and the dorsal sclerotome becomes the vertebral arch (Adapted from Larsen (1993). With permission from Elsevier)

loose core cells, and eventually ruptures, allowing the core cells to migrate toward the midline and envelop the notochord and neural tube (Fig. 1.6). The cellular structure that eventually surrounds the notochord and neural tube is termed sclerotome. The ventral sclerotome that surrounds the notochord eventually becomes the vertebral body, and the dorsal sclerotome that envelops the neural tube eventually becomes the vertebral arch.

Normal vertebral body and vertebral arch development are dependent upon sclerotome induction by the underlying notochord and neural tube. Abnormalities in this sclerotomal-notochord induction signaling process creates spinal dysraphism, which is a spectrum of birth defects caused by failure of neural tube closure. Spina bifida is defined as incomplete closure of the neural arch leaving the underlying neural elements uncovered. The severity of spina bifida ranges from spina bifida occulta, in which the neural arch fails to completely close, to more severe conditions of spina bifida, in which the contents of the neural canal extend out of the

canal and become continuous with the overlying skin. The type and severity of spina bifida are classified by the neurological tissue that extends out of the canal, which may include the neural meninges (dura and arachnoid), as well as nerve roots. The neurological tissue that extends out of the spina bifida defect is contained within a membranous tissue called a cele. The cele is what is visible on the skin surface overlying the spina bifida defect and, as indicated above, may contain meningeal tissue, in which case, the cele is termed a meningocele. The cele may also contain neural tissue and meninges, called a meningomyelocele.

Once the sclerotomes form and become positioned adjacent to the notochord and neural tube, each sclerotome divides into a cranial and a caudal portion. This cranial and caudal division allows the spinal nerves to emerge from the neural tube and exit at their respective level (Fig. 1.7a–d). Once the sclerotome division is complete, the caudal portion of the suprajacent sclerotome merges with the cranial portion of the subjacent sclerotome. This sclerotomal merging forms the vertebral precursor. This sclerotomal division and then the subsequent re-fusion explain why there are eight cervical nerves but only seven cervical vertebrae (Fig. 1.7d). The cranial division of the first cervical somite forms a portion of the base of the occiput, while the caudal division of the first cervical somite and the cranial division of the second cervical somite form the atlas. The first cervical nerve exits above the C1 vertebra, the second cervical nerve exits between C1 and C2; this pattern persists to the C7-T1 foramen where the C8 nerve root exits. The sclerotomal cells that remain following the sclerotome division surround the notochord and form annulus fibrosis, which is the fibrous portion of the intervertebral disc. The portions of the notochord that becomes enveloped by the sclerotomal tissue form the nucleus pulposus. Then during the process of maturity, the original notochord cells of the nucleus pulposus are replaced by fibrocartilageous cells.

1.3 Central Nervous System Development

Two key structures originate in the mesoderm during early development: the notochordal process and the prechordal plate. The prechordal plate induces the overlying epiblast cell layer to form the neural plate, then the neural plate cells differentiate into neurectoderm. Once formed, the neurectoderm proliferates in a cranial to caudal fashion. The cranial portion of the neural plate is broad shaped and gives rise to the brain, while the tapered caudal region of the neural plate forms the spinal cord. The positioning of the neural plate as it develops is such that the caudal portion of the neural plate overlies the notochord and is bordered by the somite pairs. This positioning allows the caudal portion of the neural plate to become enveloped by the sclerotomes forming the spinal canal, and then the neural plate itself becomes the spinal cord (Fig. 1.8). The neural plate becomes the neural tube by a process called neurulation, in which the neural plate involutes, until the lateral edges of the folded neural plate and overlying ectoderm meet and fuse in the midline forming the tubular shape of the neural tube (Fig. 1.9).

Once the neural tube fuses in the midline, it separates from the overlying ectoderm and differentiates into three distinct layers (Fig. 1.10). The innermost cell layer of the neural tube, called the ventricular layer, lays adjacent to the lumen of the neural tube (the neural canal). The ventricular layer is comprised of neuroepithelial cells, which are the precursors to the cells that eventually comprise the CNS. The first generation of cells produced by the neuroepithelial cells are neuroblasts. Neuroblasts eventually become the neurons in the CNS. Once formed, neuroblasts migrate away from the ventricular layer to form the mantle layer. The mantle layer eventually becomes the gray matter of the CNS. The neuroblasts in the mantle layer organize into four columns during the fourth week of gestation, forming paired dorsal and ventral col-

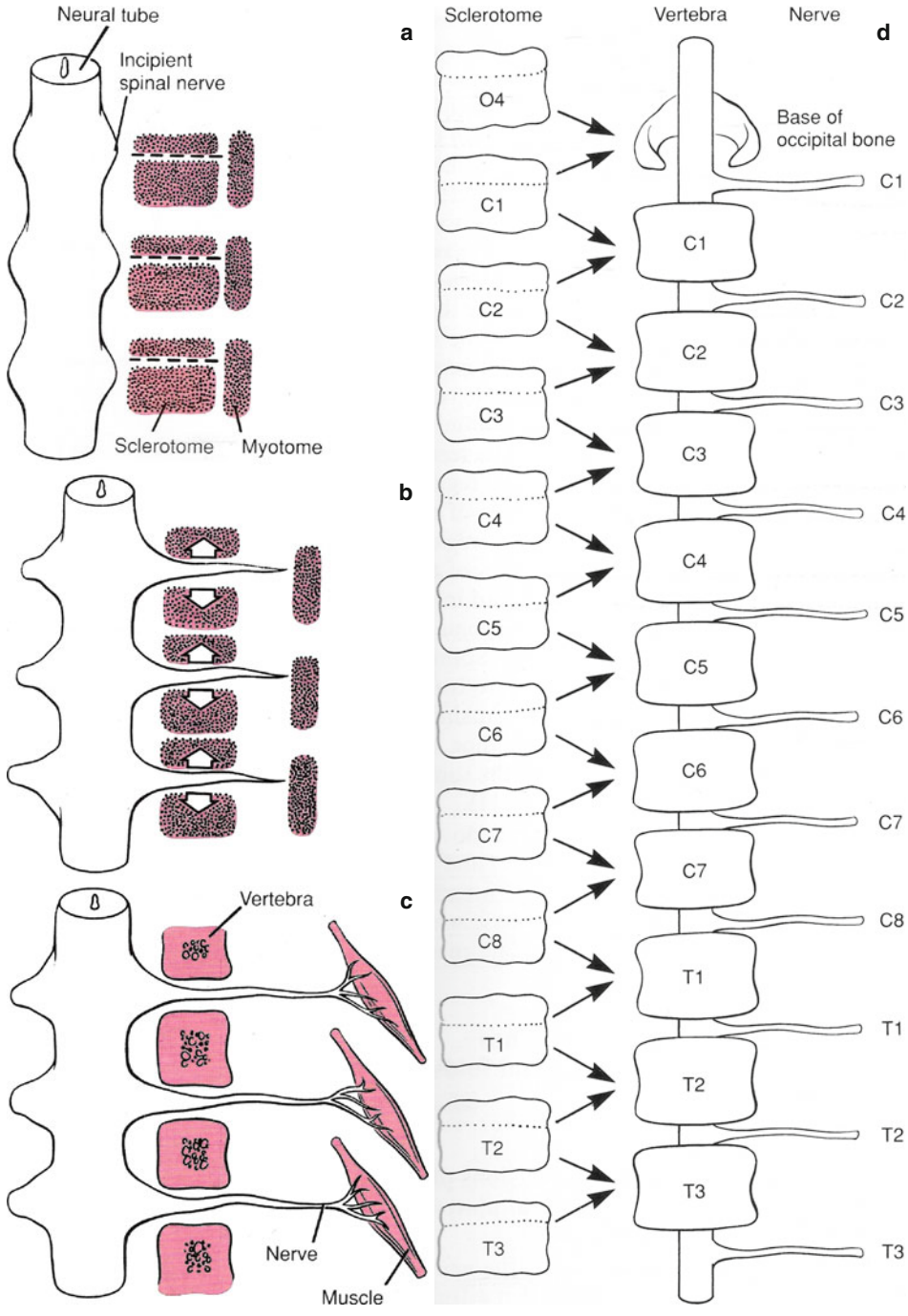

Fig. 1.7 (**a**–**d**) Sclerotome division and re-convergence. Sclerotome division allows the spinal nerves to emerge from the neural tube and extends to the periphery. The sclerotomes then reconverge to form the final vertebrae (Adapted from Larsen (1993). With permission from Elsevier)

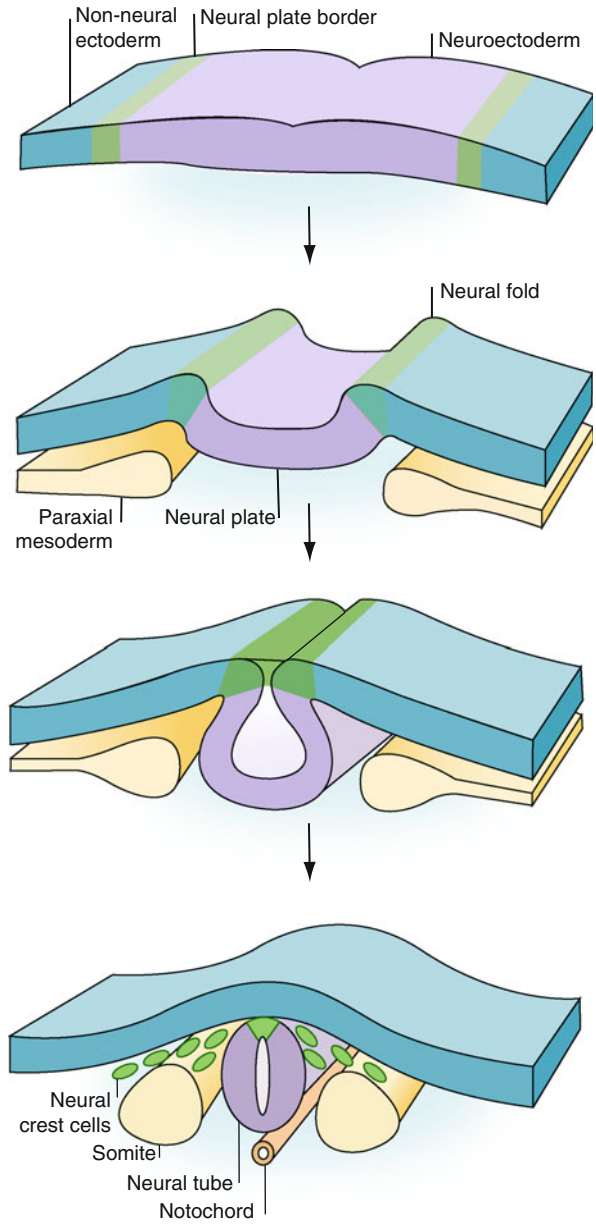

Fig. 1.8 Neural plate and CNS formation. The neural plate differentiates from the epiblast, neurectoderm cells and migrates in a cranial and caudal fashion, giving rise to the cranial (brain) and caudal (spinal cord) neural plate. The caudal neural plate is eventually enveloped by the sclerotomes, forming the spinal cord and bony spinal canal, respectively (Adapted from Gammill and Bronner-Fraser (2003). With permission from Nature Publishing Group)

umns. The cells of the dorsal column form association neurons that serve to interconnect the motor neurons of the ventral columns with the sensory neurons in the dorsal root ganglia (DRG). Neuronal processes that germinate from the neuroblasts extend peripherally to form the third layer of the neural tube, the marginal layer. The marginal layer becomes the axonal white matter of the CNS.

1.4 Peripherial Extension of the CNS; Formation of the Peripheral Nervous System

Formation of the PNS begins on approximately gestational day 30. Somatic motor neurons in the ventral gray columns extend axon sprouts toward the adjacent sclerotome tissue (Fig. 1.11). The

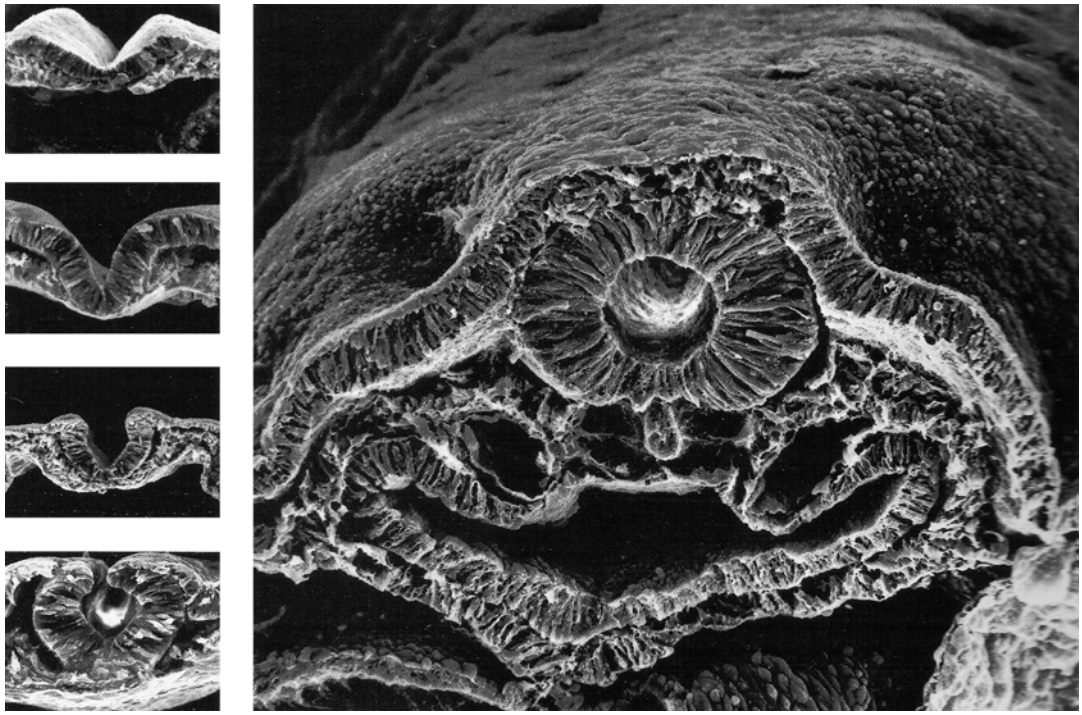

Fig. 1.9 Neurulation. The neural plate becomes the neural tube during neurulation, in which the neural plate involutes and the lateral edges of the folded neural plate fuse in the midline (Adapted from Larsen (1993). With permission from Elsevier)

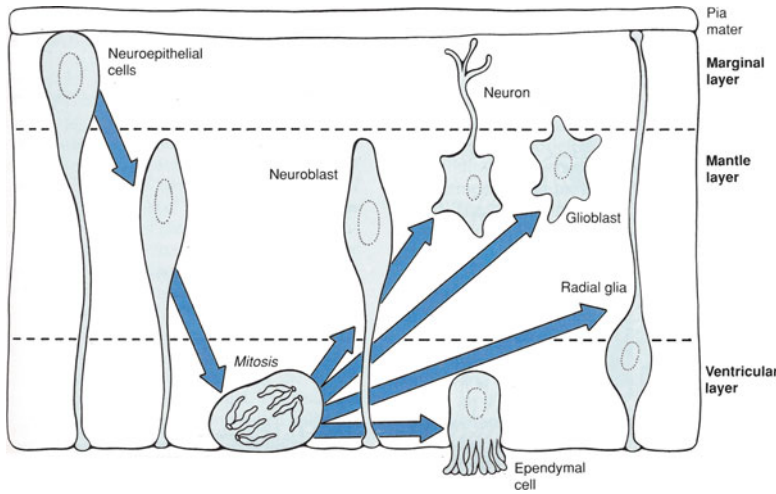

Fig. 1.10 Neural tube differentiation. The neural tube differentiates into three distinct layers. The ventricular layer forms the precursor cells that eventually populate the mantle and marginal layers and comprise the CNS. The mantle layer forms the gray matter of the CNS. The marginal layer becomes the axonal white matter of the CNS (Adapted from Larsen (1993). With permission from Elsevier)

axon sprouts begin the cervical region and extend in a cranial to caudal manner. The ventral axons coalesce as they reach the adjacent sclerotomes, forming distinct segmental nerves and the ventral roots. The somatic system is formed as ventral roots extend past the DRG. Unlike the somatic neurons in the ventral column, the neurons in the DRG are derived from neural crest cells. The neural crest cells arise from the lateral margins of the neural folds during neurulation. These cells detach from the neural plate and migrate to different regions of the developing embryo, forming melanocytes, sympathetic and parasympathetic ganglia, and the sensory neurons that reside in

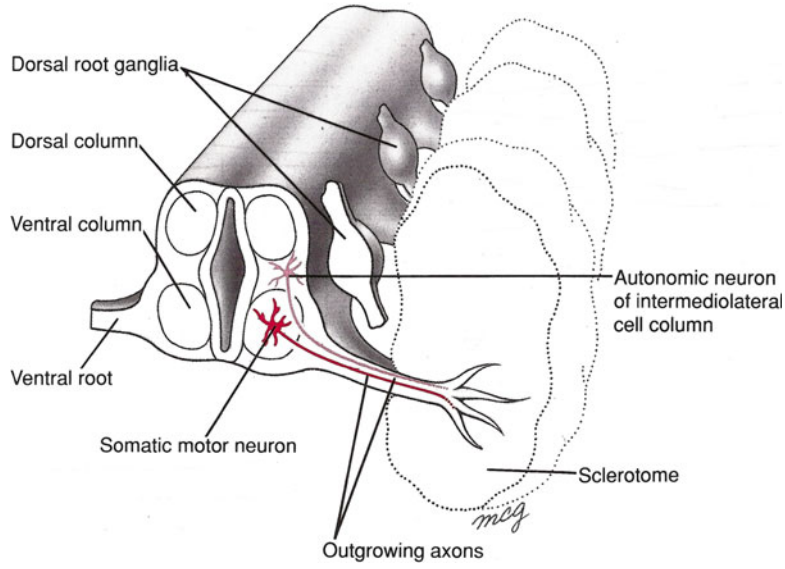

Fig. 1.11 PNS formation. Axon sprouts emerge from the primordial spinal cord and coalesce as they reach the adjacent sclerotomes, forming segmental nerves and providing end organ innervation (Adapted from Larsen (1993). With permission from Elsevier)

the DRG. The axons that extend ventrolaterally from the DRG join the axons in the ventral roots to form mixed spinal nerves. The mixed spinal nerves extend to and penetrate the adjacent sclerotomes and eventually function to innervate the end organs. Other DRG axons grow medially, extending into the dorsal column to synapse with the newly formed association neurons.

1.5 Vertebral Ossification

At approximately the sixth week of gestation, the mesodermal spine precursor transforms into a cartilage model via chondrification centers within each vertebra. Two chondrification centers develop in the vertebral body, called the centrum. The centrum then goes on to fuse in the midline to form a single vertebral body cartilage precursor. This single fused cartilaginous centrum develops into the centrum ossification center. If one of the cartilaginous centrums fails to form, only the contralateral centrum will then develop and ossify, thereby forming a hemivertebra, which then may generate a congenital scoliosis (Figs. 1.12 and 1.13). The vertebral arches derive from chondrification centers adjacent to the vertebral body; one chondrification center exists for each neural arch. Chondrification centers for the transverse processes and spinal process

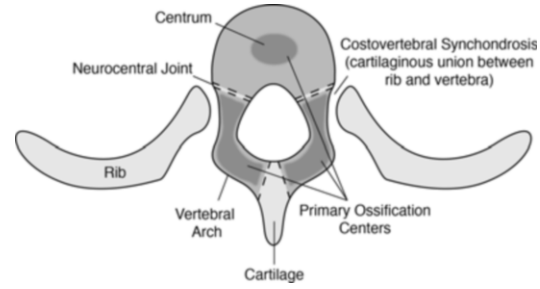

Fig. 1.12 Vertebral chondrification centers. The mesodermal spine precursor transforms into a cartilage model via the chondrification centers. The chondrification centers are eventually ossified forming the mature vertebrae

subsequently form, completing the cartilage anlage for the vertebra.

Ossification centers appear in the cartilaginous templates at approximately the ninth week of gestation. Each vertebra is derived from three primary ossification centers – one for the body (centrum) and two adjacent centers for the vertebral arches (Fig. 1.14). The centra are first ossified in the lower thoracic and upper lumbar regions. Centra ossification progresses more rapidly in the caudal vertebrae, while the vertebral arches are more rapidly ossified in the cervical spine. Dorsal, midline fusion of the lamina initially occurs in the lumbar spine, then progresses cranially. Once ossified, the lamina does not fuse to the centrum. Instead an embryologic joint, the neurocentral joint, persists between the centrum and each

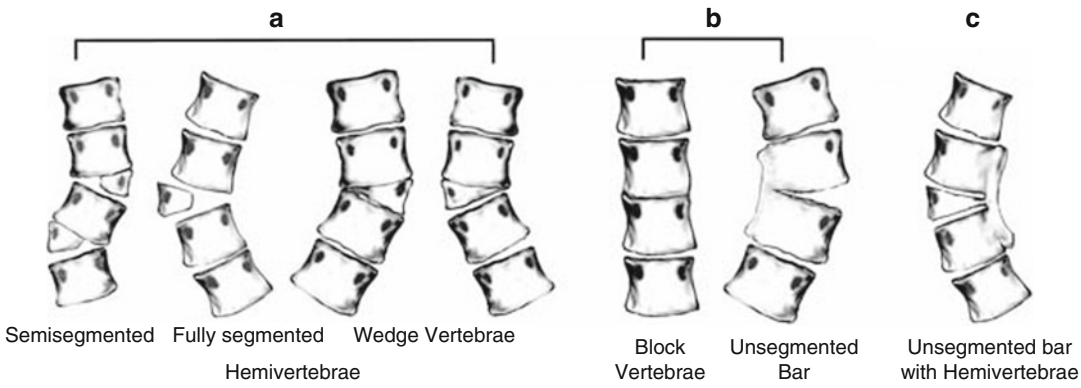

Fig. 1.13 (**a**–**c**) Congenital scoliosis (Adapted from Erol et al. (2002). With permission from The University of Pennsylvania Orthopaedic Journal)

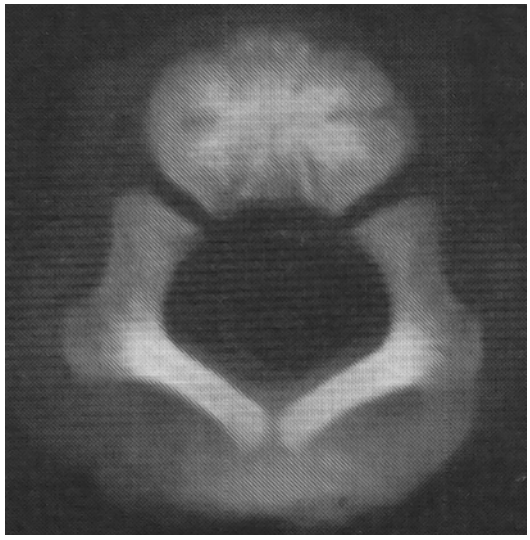

Fig. 1.14 Vertebral ossification centers. The three primary ossification centers of the vertebrae (Adapted from Herkowitz et al. (1999). With permission from Elsevier)

lamina. The neurocentral joints allow the expansion of the spinal canal during growth of the centrum and lamina and eventually disappear by 6 years of age. Secondary ossification centers at the tips of the transverse processes; spinous process and ring apophysis develop after birth and eventually fuse during the third decade of life (Fig. 1.12).

Conclusion

Embryological formation of the spine and spinal cord progresses in an organized manner, beginning with formation of the primitive streak, notochord, somites, and scleotomes. Normal vertebral and neural formation is dependent upon the development of these early structures to induce the adjacent cell lines to form the neural arch and distinct vertebral bodies. Errors in formation of these structures lead to induction failure and subsequent spinal dysraphism and congenital scoliosis.

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Genetics

2

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Key Points

- DNA is the blueprint of our human body. Variations in DNA are the source for the phenotypes of different individuals.
- The two most common types of variations, also called polymorphisms, are microsatellites and single nucleotide polymorphisms (SNPs) in our DNA.
- Mendelian disease refers to a simple form of disease in which alternation or mutation in a single gene is enough for its manifestation. Linkage analysis has been proven to be a useful method for studying this type of disease.
- Complex genetic disorders are caused by multiple genes with small effects combined with environmental factors. Candidate-gene and genome-wide association studies using case-control design are best used to analyze these disorders.
- Whole genome sequencing will discover all types of variations for comprehensive genetic studies
- Idiopathic scoliosis is likely a complex genetic disorder. With the recent developments in human genetics, the cause of idiopathic and congenital scoliosis will likely be elucidated in the not-too-distant future.

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2.1 Basic Genetics

Today's human genetics and genetic epidemiology are based on the Mendelian laws of inheritance, which explain the pattern of segregation of genes through generations. With the help of modern technology, we are able to determine the status of potential or causative genetic variants that might lead to the development of disease. This chapter aims to provide the reader with a basic concept of the terminology and principles involved in disease gene hunting, so that readers will have a much better understanding of the advances in genetics and scoliosis, which will no doubt appear in the literature in future.

2.1.1 The Chromosome and DNA

To understand the principle of genetics and disease gene mapping, concepts concerning the chromosome and its structure, DNA, genetic polymorphisms, and different types of diseases need to be clarified.

The human genome consists of 23 pairs of homologous chromosomes. This complete set of chromosomes is called diploid, while the halved set of a gamete is called haploid. For each pair of these homologous chromosomes, one is derived from the father (paternal) while the other is from the mother (maternal). The chromosomes are made up of DNA (deoxyribonucleic acid), which is a long stretch of nucleotide sequence of four bases – adenine (A), cytosine (C), guanine (G), and thymine (T). Each single strand of DNA has two ends, namely, 5′ and 3′. Hydrogen bonds make the pairing of A with T and C with G. With the existence of base complementarity between bases of two DNA strands running in opposite directions (5′ to 3′ and 3′ to 5′), a double-stranded DNA is formed. This is the primary structure of DNA. Upon further interaction with histone and scaffold protein, which are the major proteins to package DNA into a smaller volume to fit in the cell, the DNA is tightly wound together into chromosomes (Fig. 2.1).

The DNA contained in the chromosomes provides the blueprint for making all the structures inside the human body, as well as all the "software" needed to regulate its processes at the molecular level. All the necessary information is stored in the DNA sequence as a series of codes. A sequence of DNA that contains coding information is called "exons," while non-coding sequences are called "introns." A series of steps occur inside each cell to decode these information and translate them into protein products that are essential for metabolism as well as other normal functions of the human body. Through the process of *transcription*, a single DNA strand is used as a template for constructing a complementary RNA strand. Apart from the intrinsic chemical difference between DNA and RNA, the most important difference is that U (uracil) replaces T in RNA. In a certain region of the genome, which we call a gene, the transcribed RNA sequence encodes for information (codons – every three bases of RNA determine an amino acid) on

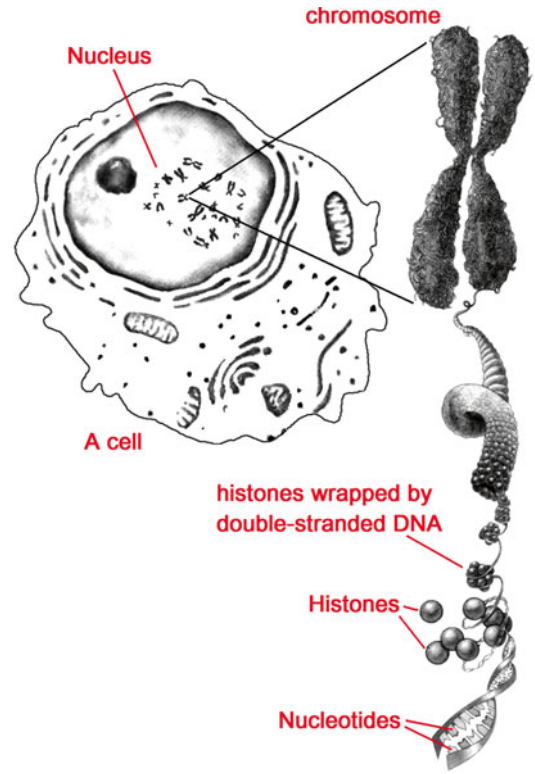

Fig. 2.1 Chromosomal organization

how to make a certain protein (depending on the gene) with a specific amino acid sequence. Any genes will contain regulatory sequences and variable numbers of intervening exons and introns. The transcription of genes forms messenger RNA (mRNA) and would include all the intron and exon regions. Although the introns are non-coding region and are removed eventually, they may have regulatory functions during the processes of transcription. Eventually with post-transcriptional modification called splicing, these intron regions are removed, and the coding exons are linked together to form mature mRNA. The mRNA then undergoes *translation*. This is the process in which the specific amino acid sequence coded for by the mRNA is translated by ribosomes into amino acids. The amino acid sequences assemble into peptides and proteins (Fig. 2.2). Upon further post-translational folding, twisting, and interacting with other proteins, the secondary, tertiary, and quaternary structure of proteins are formed. These are important for the proper functioning of the active protein.

These proteins may form part of the structural elements of the tissue, such as collagen types 1 and 2; they may contribute to the extracellular matrix to form proteoglycans; or they may form regulatory enzymes, such as metalloproteinases, that help to regulate the metabolic processes inside the tissue.

2.1.2 Genetic Polymorphisms and Their Relation to Diseases

The haploid genome is comprised of about three billion base pairs (bp). There are about 30,000 genes located in the genome [40, 81]. Majority of the genome is shared in common among the population, with only a small part of it having variation. Such small proportion of difference may cause great influence on the phenotype of different individuals. The two most common types of variation, also called polymorphisms, are microsatellites and single nucleotide polymorphisms (SNPs). At the locus where the polymorphism is located, variants are called alleles. These alleles of the polymorphism are inherited through generations with each individual having two alleles at each locus and are determined by both the paternal and maternal lineages. Microsatellites are tandem repeats of short sequence of 2–8 bp, and the number of tandem repeats differentiates alleles (Fig. 2.3). It is highly polymorphic. SNPs are polymorphisms that differ at a single nucleotide (Fig. 2.4), and the number of known SNPs exceeded ten million in the human genome [15, 79]. Although an individual SNP is not as polymorphic as a microsatellite due to the limited number of alleles, they are compensated for by the large numbers of SNPs scattered throughout the gene and the genome; thus with high-throughput genotyping technology, SNP markers

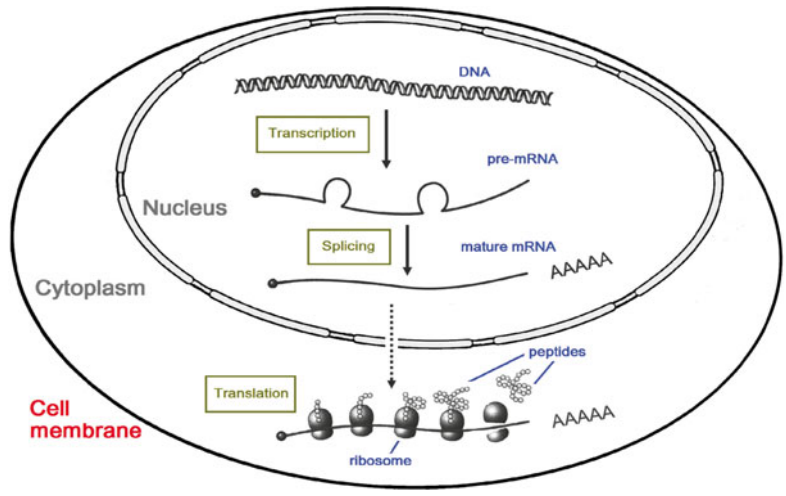

Fig. 2.2 Central dogma from DNA through RNA to protein

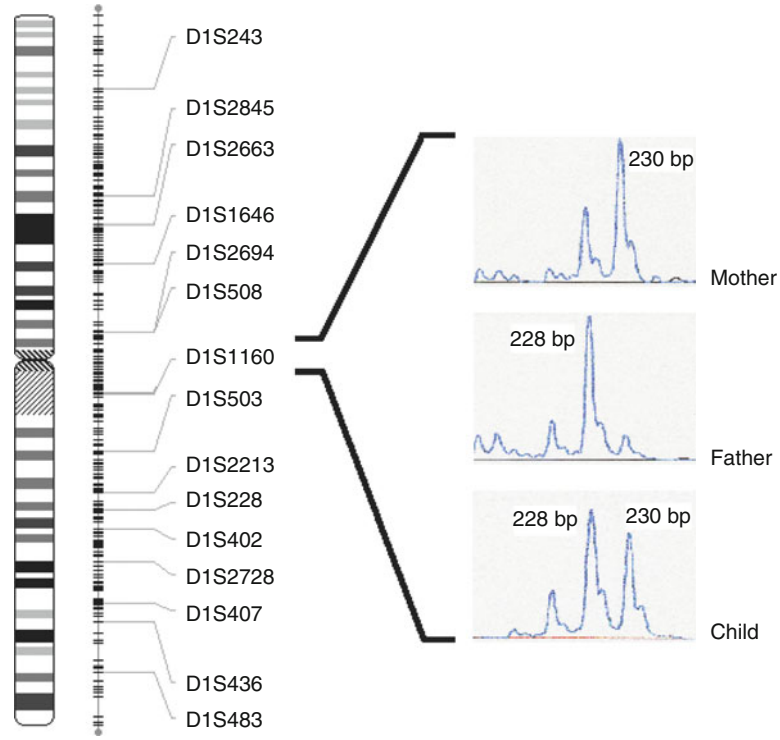

Fig. 2.3 Microsatellite markers and their inheritance. The mother is homozygous for 230 bp allele at marker D1S1160, while the father is homozygous for 228 bp allele. As a result, their child inherited one copy of both alleles simultaneously from the parents and hence heterozygous at the marker

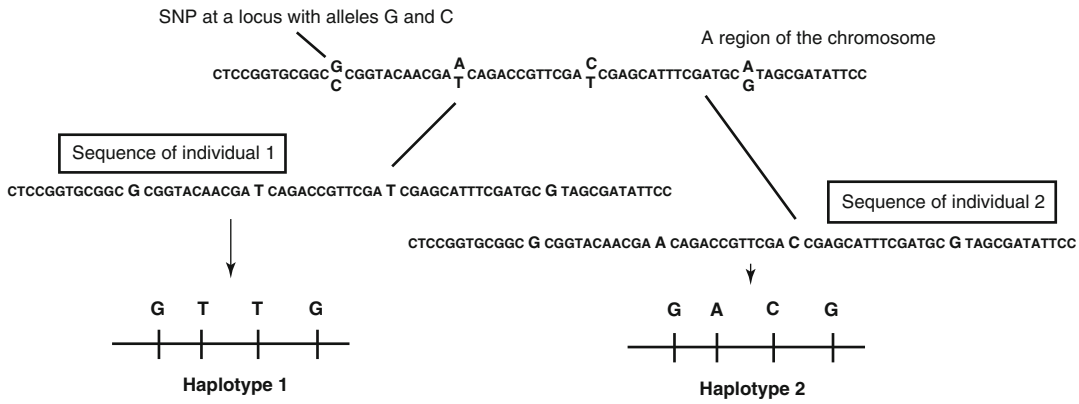

Fig. 2.4 SNP and haplotype. On the sequence of a chromosome region, there are four SNP loci. When considering the haplotype of these four loci, individual 1 is having GTTG, while individual 2 is having GACG

are more commonly used for genetic analysis nowadays. There are various types of SNP, such as non-synonymous coding SNPs that change the amino acid sequence encoded, synonymous coding SNPs that do not modify the encoded amino acid, intronic SNPs located in the introns that might affect proper splicing, SNPs located in the 5′ and 3′ untranslated region (UTR), and intergenic SNPs that are not located within a gene.

Changes in gene sequences that result in disease are generally called mutations, while changes in the gene sequence without significant external effects are termed polymorphisms. Nonsense mutations result in an amino acid change to a stop codon. Deletion mutations delete one or more nucleotides from a sequence, and insertion mutations insert one or more nucleotides into a sequence. The most recorded

pathogenic mutations are detected in the coding sequence such as nonsynonymous mutations and frameshift mutations. Promoter is the regulatory region that is located upstream of a gene, and it provides regulation to transcription so that gene expression is controlled. Mutations in promoter which controls gene transcription would prevent the promoter from working, resulting in a change in the level of gene expression. These products of mutations may have reduced or no function, called loss of function mutation [7], while gain of function mutation takes place when the gene product has a positively abnormal effect [30, 68].

If we are referring to a single locus, the term genotype is used to define the status of the two alleles. On the other hand, when more than one locus on a chromosome is considered, haplotype denotes their allele configuration according to their order in physical position (see Fig. 2.4). Since a single SNP locus has only two alleles, it is not very polymorphic, with a limited number of variations. Thus several SNP loci can be combined into a haplotype and used to increase the power to detect associations (see Fig. 2.4).

With the advance of sequencing technologies [5, 35], sequencing of individual human genomes or whole exons (1–2 % of the human genome) is becoming more and more affordable. The technologies allow discovering every type of DNA variations in genome, not just most common variations (SNPs and microsatellites) we mentioned above but also other variations such as rare alleles (low-frequency alleles), copy number variations (CNVs). These variations may explain some more genetics effects for disease phenotypes and can be used as markers to study the relationship between genotypes and phenotypes.

2.1.3 Types of Disease

Phenotypic variations between individuals are determined by polymorphisms of particular genes resulting in differences in genotypes. These phenotypic outcomes may be expressed subtly at the molecular level (e.g., expressional level difference of a protein) or more obviously resulting in notable body changes (e.g., height) or even symptoms of diseases. However, risk-conferring genotype in one individual may not necessarily result in disease or symptoms of disease in another. This is because of a phenomenon known as penetrance. Incomplete penetrance refers to the case in which the risk-conferring genotype is not fully expressed and therefore does not actually cause disease. This maybe because the risk-conferring genotypes may require additional exposure to environmental factors, as well as interaction with other susceptibility genes in order to develop disease. Such conditions requiring an interaction of multiple genes with the environment are known as complex genetic disorders. Osteoarthritis, degenerative disc disease, hypertension, and diabetes are examples. Mendelian disease refers to a simpler form of disease in which alternation or mutation at a single gene is enough for its manifestation [56, 61]. They represent rare diseases with more severe phenotype, such as osteogenesis imperfecta or Duchenne muscular dystrophy. Furthermore, phenotypic outcomes may be qualitative (i.e., with or without the disease) or quantitative, presenting with a spectrum of severity from mild to severe, which can be measured by number of units (e.g., blood pressure, scores of intervertebral disc degeneration) [72].

2.2 Disease Gene Mapping

Before claiming that a disease has a genetic component and trying to find the gene, commonly called mapping, it is important to estimate the relative importance of the genetic risk factor on the disease. One way to do so is to assess familial aggregation; if the disease occurs in multiple members of the same family, this is an indication. However, one needs to remember that members of the same family are likely to be exposed to similar environmental factors, such that the appearance pattern of a disease may not be ultimately due to genes but merely to non-genetic factors. On the one hand, an even better method is to examine disease occurrence between twins, especially as monozygotic (identical) twins share the same genes; on the other hand, dizygotic

(non-identical) twins only share 50 % of similar genes. Therefore, for a purely genetic disease, the monozygotic twins should both have the disease (high concordance rate). But if both monozygotic and dizygotic twins have similar concordance rates, it would be stronger evidence for shared environmental factors being a major factor. If low concordance rates are found among twins, the disease could be affected by some unshared environmental factors. In summary, for a disease predisposition to be genetic, high and low concordance rates must be obtained from monozygotic and dizygotic twin pairs, respectively [9]. These classical twin studies are a common feature of many diseases that are suspected to have a genetic component.

Once a disease has been confirmed to have a substantial genetic component, one can attempt to map the disease gene to a particular location in the genome by the use of a number of strategies including linkage analysis on familial subjects, case-control association studies using population-based subjects on either biologically relevant candidate genes, or case-control association studies on a genome-wide scale using gene-chip arrays.

2.2.1 Linkage Analysis on Familial Subjects

Linkage analysis is a classical method for mapping disease genes, and it has been successfully used to identify numerous disease genes in the past decades. Families, preferably large and having multiple affected members, are recruited and genotyped for hundreds of microsatellite markers. If a disease gene is located in the proximity of one of these markers, so that recombination is unlikely to occur at a position in-between the marker and the disease gene, that region of the chromosome is likely to be transmitted to affected members within the family together with the marker. Hence, the marker is said to be in linkage with the disease gene and produces a characteristic pattern of transmission (Fig. 2.5). With the use of microsatellite markers covering the whole genome, genome-wide linkage analysis can locate the rough chromosomal localization of an unknown disease gene without any prior knowledge. It is a powerful strategy that can maximize the chance of finding a disease gene.

If the marker is on the same chromosome as the disease gene, recombination will be responsible for breaking them up so that their alleles will not be transmitted together on the same chromosome. The further apart they are, the higher the chance that they will be affected by recombination. Hence, from the rate of recombination, we can estimate the distance between the marker and the unknown disease gene. Generally speaking, a 1 % recombination rate (θ) is referred to as 1 centimorgan (cM) apart, and it is roughly equivalent to one million bp distance on the chromosome [55, 69].

In a parametric linkage analysis, one tests whether the test hypothesis (that the marker is linked to the disease gene) or the null hypothesis (that the marker is not linked to the disease gene) is true. After making the assumption of disease model (e.g., mode of inheritance, penetrance, and disease allele frequency), one performs sequential test at various θ to compare the likelihoods of the test and null hypotheses. The likelihood of the test hypothesis to the likelihood of the null hypothesis is called the likelihood ratio or odds. Taking the logarithm to base 10 of this likelihood ratio will give us the LOD (logarithm of odds) score. The point with the highest LOD score indicates the most likely distance between the marker and the disease gene locus [51]. To achieve a genome-wide significant level equivalent to $p = 0.05$, an LOD score of 3.3 is required [39].

The advantage of the LOD score method is that one can combine the results from different studies to strengthen the significance (considering they are studying the same disease with the same disease model assumption) [51]. For instance, one may have relatively small sample sizes across studies and find suggestive linkage evidence with LOD score <3.3. Although the LOD score does not reach the threshold of 3.3 in an individual study, their LOD scores can be added up so that the combined LOD score may reach statistical significance.

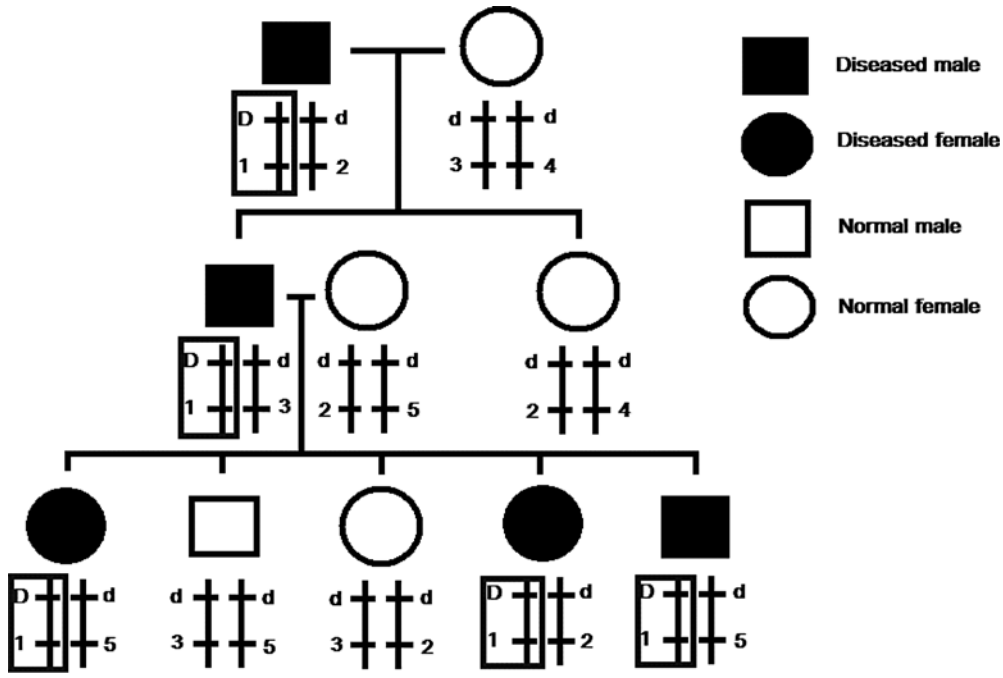

Fig. 2.5 Typical inheritance pattern of linkage. In the figure, two loci are considered. The upper locus is the disease gene locus with disease allele *D* and normal allele *d*, while the lower one is a nearby microsatellite marker locus with many alleles (*1*–*5*). If the two locus is in linkage, they will be transmitted together to the offsprings without being disrupted by recombination during meiosis. In this example, the two loci are in linkage, and allele *D* is linked with allele *1*. The resulting haplotype D-1 is transmitted to every diseased family member. Since the disease gene locus is unknown, linkage analysis relies on correlating the marker locus inheritance (genotype) with disease status (phenotype) to detect linkage. As the marker locus is in linkage with the phenotype, it is an evidence that the unknown disease gene locus is somewhere nearby and thus its rough chromosomal localization can be determined. By using large amount of markers covering the whole genome, genome-wide linkage analysis can be performed. As the markers are not the actual disease-causing mutation and they only denote a certain status of the chromosome, each family may have a different allele in linkage with the disease-causing allele

An alternative to parametric linkage is nonparametric linkage analysis which dose not make an assumption on the disease model. This still allows the usage of the power of linkage analysis. The affected sib pair (ASP) method [38, 58] and the affected pedigree member (APM) method [83, 85] were developed. The latter case uses all affected members instead of only affected siblings for analysis. The idea of nonparametric linkage analysis is that if there is a disease-causing mutation at a locus near a marker, they are in linkage and their alleles on the same chromosome are likely to be transmitted among affected pedigree members unless recombination disrupts them. As a result, the affected members within the same family are expected to share marker alleles in common more often than by chance alone (50 % for siblings) if there is a disease-causing mutation at a gene nearby.

A number of computer programs Allegro [29], Genehunter [37], and Merlin [1] have been developed for performing linkage analysis.

The merit of linkage analysis is that we need not have any prior knowledge on where the disease gene is, and we can determine its location based on evidence of linkage with markers; thus it is a method to discover new and unexpected predisposing genes. However, it works best in diseases where relatively few genes are involved and where these genes exert relatively major effects (i.e., disease causing). It lacks the power to detect the effect of common alleles with modest effects on disease, and these may be important as our understanding of genetic predisposition

increases. It is likely that common diseases such as hypertension, diabetes, osteoarthritis, and intervertebral disc degeneration are the result of multiple genes with modest effects interacting with the environment to produce a phenotype. For these, a population-based genome-wide case-control association studies maybe the better approach.

2.2.2 Case-Control Association Studies on Population-Based Subjects

Instead of testing for allele sharing within families in linkage described above, population-based association looks for an allele to be associated with a trait (symptom or characteristic of a disease) across the population. The principles are similar to linkage, but it searches for an allele for a disease-causing gene mutation in an extended family (i.e., individuals of a population believed to share a common ancestry). In this special kind of linkage study, the "family" considered is the whole population, and the linkage is so tight (distance between the disease-gene locus and the marker locus is extremely close) that it will not be disrupted by recombination even after thousands of generations. To test for association, we test whether a particular allele of a locus (i.e., a marker) is overrepresented in cases and, at the same time, underrepresented in controls. If so, we can claim that such locus is associated with the studied disease.

In general, there are two types of association – direct and indirect [14]. Direct association targets polymorphisms that have functional consequences and predisposes to disease. This kind of association is the most powerful, but the chance of selecting a marker, which is also a disease predisposing allele, is not high. On the other hand, in indirect association, the association is between the marker and the nearby disease predisposing allele. It relies on the principle of linkage disequilibrium (LD) whereby, due to the proximity of the marker to the predisposing allele, the marker will be associated with the predisposing allele and, therefore, the disease is in a higher frequency than would be expected. Thus identification of such a marker would provide clues that a disease causing polymorphism is nearby and would narrow the search for this polymorphism.

There are two methodologies for association studies. The first is by the use of a candidate-gene approach, in which one guesses the likely genes that are involved in the disease and directly screen them for disease association using a set of markers as described above. The identification of such genes is usually based on previous studies that suggest the candidate genes are biologically involved in the disease or that they reside within the functional pathway of the disease process. One such example would be for the testing of the Asporin gene in degenerative disc disease [72], when this gene has already been shown to be involved in osteoarthritis [34].

Once the candidate genes are selected, the next step would be the selection of markers within the gene or region of interest. The most commonly used markers are called single nucleotide polymorphisms (SNPs). These are single nucleotide changes within the human genome which do not have a functional consequence. These have been identified and are provided within the HapMap database [79]. This type of association study is also often the final part of a linkage analysis study. While the linkage analysis described above can identify a region within a particular chromosome, it is unable to identify a particular gene. Thus the best candidate genes can be selected within the confined interval and tested using a case-association approach. Such a two-stage approach would minimize the candidates to be tested as well as maximize the chance of disease gene hunting.

The second type of case-association study is the so-called genome-wide association study. The principle is the same as that described above, except that due to advancements in technology, rather than to test single candidate genes, a high-density SNP map of the whole genome is generated, and all these SNPs are tested for association with disease by comparing their frequencies between the disease and control cohorts. This type of study is only made possible recently by the availability of high-throughput genotyping

platforms such as DNA genechips [59]. It is now feasible to genotype hundreds of thousands of SNPs at a reasonable cost and time. By using large amounts of SNP markers that cover the whole genome, genome-wide association studies need not select candidates and thus does not rely on "best guess" selection of candidate genes. With the availability of initial results highlighting a particular chromosomal region, the indicated genes can be studied in detail by a direct association approach, in which polymorphisms that result in a change in the coding sequence of the gene (often referred to as non-synonymous SNPs) are examined.

2.2.3 Genetic Mutations to Spinal Abnormalities

Non-synonymous coding mutations, SNPs in the introns of splicing sites, SNPs in the 5′ and 3′ untranslated region (UTR) may affect gene function. As genes encode peptides and proteins that may form structural elements of the spine (e.g., collagens), extracellular matrix components (e.g., proteoglycans), or enzymes in regulating metabolic processes (e.g., metalloproteinases), alterations in their gene function may result in altered levels of expression or altered structure of the involved protein, leading to disease.

For example, radiological studies of spondyloepiphyseal dysplasia Omani type (SED Omani type) showed minor metaphyseal changes but major manifestations in the spine and the epiphyses. With age, the vertebral endplates became increasingly irregular, the intervertebral space diminished further, and individual vertebrae started to fuse resulting in a severe short-trunk dwarfism with kyphoscoliosis [65]. A mutation (R304Q) in the *CHST3* gene was identified in these patients [80]. *CHST3* encodes chondroitin 6-*O*-sulfotransferase 1 (C6ST-1), which catalyzes the modifying step of chondroitin sulfate (CS) synthesis by transferring sulfate to the C-6 position of the *N*-acetylgalactosamine of chondroitin. The mutation is essential for the structure of the cosubstrate binding site leading to defective sulfation of chondroitin sulfate (CS) chain and chondrodysplasia with major involvement of the spine [80].

CHD7 gene is widely expressed in undifferentiated neuroepithelium and in mesenchyme of neural crest origin. Towards the end of the first trimester, it is expressed in dorsal root ganglia; cranial nerves and ganglia; and auditory, pituitary, and nasal tissues as well as in the neural retina [67]. Gao et al. [21], in 2007, identified a single-nucleotide polymorphism (SNP), an A-to-G change in intron 2 of the *CHD7* gene that was predicted to disrupt a caudal-type (cdx) transcription factor binding site, which affects *CHD7* gene expression leading to association with late-onset idiopathic scoliosis (IS) [22].

Osteogenesis imperfecta type IIB is an autosomal recessive form of perinatal lethal osteogenesis imperfecta with excess posttranslational modification of type I collagen, indicative of delayed folding of the collagen helix [6]. CRTAP protein interacts with the enzyme responsible for posttranslational prolyl 3-hydroxylation of collagen. Without CRTAP protein, collagen structure was abnormal. A homozygous single-base pair (T) deletion in exon 4 (879delT) caused a frameshift and was expected to cause a null allele due to nonsense-mediated decay [6]. Other homozygous or compound heterozygous mutations in the *CRTAP* gene also have been identified to cause low levels of *CRTAP* mRNA and a lack of CRTAP protein [49].

2.2.4 Genetics of Early-Onset and Congenital Scoliosis

Over 80 % of scoliosis conditions are idiopathic in nature and are conventionally classified according to the age of disease onset – infantile (aged 0–3), juvenile (aged 4–10), and adolescent (aged older than 10). The clinical presentation is quite different depending on the onset of the disease, e.g., infantile idiopathic scoliosis is more common in boys with left-sided thoracic involvement but adolescent idiopathic scoliosis is more common in girls and with right-sided involvement. Up until now, there is no established genetic evidence explaining the difference in the onset of the disease [24].

According to the Scoliosis Research Society (SRS), early-onset scoliosis (EOS) refers to lateral curve of the spine that is diagnosed before the age of 10. In general, it includes both infantile and juvenile idiopathic scoliosis as well as congenital scoliosis. The spine in idiopathic scoliosis appears normal in morphological appearance, whereas congenital scoliosis has malformation in the vertebrae due to failure of segmentation or formation. Under some circumstances, neuromuscular scoliosis, syndromic scoliosis, and thoracic insufficiency syndrome are also included as early onset scoliosis since these deformities can be identified at birth or present quite early in life. Very little is known about the inheritance of early onset scoliosis. Wynne-Davies et al. examined 114 patients with idiopathic scoliosis and noticed that there were more boys being affected in the early-onset group (infancy to 8 years of age), whereas the late-onset group (8 years of age and older) had more girls involved [90]. Same study also concluded that the incidences of scoliosis among the first-, second-, and third-degree relatives were higher in the late-onset scoliosis, in particular, the first-degree relatives. Another study reviewed 87 families with early-onset idiopathic and congenital scoliosis and concluded that the recurrence risk for scoliosis was low but there was an increased risk of neural tube defects in families with congenital scoliosis [13]. Furthermore, kyphoscoliosis resulting from solitary hemivertebrae and localized anterior defects of the vertebral bodies were mainly sporadic [91]. In another review of 1250 patients with congenital spinal deformities, only 13 patients were found to have a first- or second-degree relative with vertebral defects [87].

Congenital scoliosis usually represents sporadic occurrence with an incidence of 0.5–1/1000 live births [25, 70]. The etiology is still unknown, but it is likely due to multifactorial including genetic and environmental factors. Hypoxia, hyperthermia, carbon monoxide, and alcohol are some common environmental factors that can lead to vertebral anomalies during fetal development [31]. Gestational hypoxia is known to cause congenital scoliosis over a century ago [23], and recent evidence suggested gene-environmental interactions such as gestational hypoxia could potentiate the development of congenital scoliosis in genetically susceptible mice through abnormal FGF signaling [73].

In the embryo, vertebral bodies are developed from somites through a complex interaction of various signaling pathways including FGF, Wnt, and notch [60]. A number of notch pathway genes including MESP2 [86], LFNG [74], and HES7 [75] were identified to be important in the normal somite segmentation and vertebral development in mice. Mutations of these genes can lead to spinal deformities. In humans, notch pathway gene mutations have now been identified in spondylocostal dysostosis (SCD) [8] and Alagille syndrome [41, 53], which are known to have congenital vertebral malformation and scoliosis. Based on the assumption that the genetic components of the development of scoliosis are conserved across species, Giampietro et al. used the mouse-human synteny analysis to identify potential human candidate genes from the patterning genes of Wnt, FGF, and Notch signaling pathways in mice somitogenesis [24, 25, 28]. A number of candidate genes including PAX1, DLL3, and TBX6 were studied using association analysis [18, 20, 26, 27, 43]. In the analysis of 254 Chinese Han subjects (127 congenital scoliosis patients and 127 controls), two SNPs of TBX6 gene (rs2289292 and rs3809624) were found to be in strong linkage disequilibrium ($d' = 1.0$; $\gamma^2 = 0.984$; 95 % confidence interval, 0.96–1.0; LOD = 57.48) in the controls. The authors suggested that the genetic variants of TBX6 gene might play an important role in the development of congenital scoliosis in Chinese Han population [20].

2.2.5 Genetics of Adolescent Idiopathic Scoliosis

Adolescent idiopathic scoliosis is the most common pediatric spinal deformities affecting 2–3 % of the school age children [84]. Twin studies gave evidence for a genetic etiology in adolescent idiopathic scoliosis (AIS) [3, 90]. The severity of the disease within families can change and

sometimes miss or skip generations. It is also possible that more than one gene is involved in the disease.

Ogilvie and Braun in 2006 investigated a cohort of 145 AIS probands to ascertain whether they have a family history of AIS and found that nearly all (97 %) AIS patients have familial origins [54]. The authors suggested at least one major gene with different penetrance and expressivity. They also detected a major gene effect by segregation analysis using a model with age and gender effects in 101 pedigrees ascertained through a proband. Their model indicates that only 30 % of the male and 50 % of the female carriers of the predisposing allele develop pronounced forms of the disease [4].

Family linkage analysis and case-control association have been used to detect disease susceptibility genes. Miller [46] and Cheung et al. [12] in 2007 gave good reviews on the genetics of familial idiopathic scoliosis in four published data sets. Significant linkage regions were identified through a genome-wide analysis of a large family on chromosomes 6, 10, and 18, with the highest LOD score on chromosome 18 [88].

Genome scans of seven multiplex families of southern Chinese descent with AIS were carried out. A two-point linkage gave a LOD score of 3.63 with a flanked region (5.2 cM) between D19S894 and D19S1034 on chromosome 19p13.3 [10]. This region was later confirmed to be significantly linked to a subset of families with probands having a curve > or =30° [2]. The X chromosome was reported to link to a subset families with a maximum LOD score of 1.69 (theta = 0.2) at marker GATA172D05 [33], and chromosomes 5 and 13 were found to link to a subset of families with kyphoscoliosis [45]. A positive LOD score of 3.20 at theta = 0.00 was detected with marker D17S799 in a three-generation IS Italian family. Then six additional flanking microsatellites confirmed the linkage between D17S947 and D17S798 [66]. More recently, significant linkage was detected to the telomeric regions of chromosomes 9q at marker D9S2157 with a maximum LOD score of 3.64 and 17q at marker AAT095 with a maximum LOD score of 4.08 in AIS pedigrees of the British population. The 9q region was further narrowed down to approximately 21 Mb at 9q31.2–q34.2 between markers D9S930 and D9S1818, and the 17q candidate region was 3.2 Mb between the distal to marker D17S1806 on chromosome 17q25.3–qtel. [52]

In addition, evidence of linkage and association with (multipoint LOD 2.77; $p = 0.0028$) was detected in a cohort of 52 families of AIS on 8q12 loci of genome-wide scans. Haplotypes of the CHD7 gene were detected to be associated with the CHARGE syndrome after fine mapping in the region. The re-sequencing of CHD7 gene revealed at least one potentially functional polymorphism that is over-transmitted ($p = 0.005$) to the affected offspring and predicts disruption of a caudal-type (cdx) transcription-factor binding site. These results suggest etiological overlap between the rare, early-onset CHARGE syndrome and the common, late-onset IS [22].

Several genes have been studied by using case-control design. MATN1 gene was analyzed in a population of 81 trios, each consisting of a daughter or son affected by idiopathic scoliosis (IS) and both parents. An allele of a microsatellite marker in MATN1 was found to have been significantly over-transmitted from parents to affected probands. The results suggest that familial idiopathic scoliosis is associated with the MATN1 gene [48]. With the support of the chicken pinealectomy model, melatonin deficiency was suggested to play a significant role in AIS. In a case-control analysis, melatonin receptor 1B (MTNR1B) was found to be one of the candidate genes [62]. The authors initially screened 472 cases and 304 controls with five tag SNPs and replicated the study with 342 cases and 347 controls. All subjects were Chinese. A promoter SNP (rs4753426) was found to be more in AIS patients, and the CC genotype significantly increased the risk of AIS by an odds ratio (OR) of 1.29. However, similar results could not be confirmed in different ethnicities, such as the Japanese and Hungarian population [50, 77]. The XbaI polymorphism site of the estrogen receptor gene was studied in 304 girls with idiopathic scoliosis using Cobb's method. The authors detected that this polymorphism was associated with

curve severity [32]. This association was confirmed by a Chinese data set consisting of 202 patients with AIS and 174 healthy controls [89]. However, the association with curve severity could not be replicated in 364 Chinese AIS patients and 260 controls [78]. Although other candidate genes such as IGF-I [92], growth hormone receptor [63], and aggrecan [44] did not appear to be significantly associated with AIS in a particular study data set, further studies are still required.

In summary, several loci have been detected by linkage analysis as listed in Table 2.1. While the linkage analysis to map AIS was proven to be successful, nonetheless, the progress of detecting disease genes remains slow and, so far, only *CHD7* gene has been discovered by linkage approach.

This maybe because in complex diseases, multiple genes with only moderate effects are involved, and as previously mentioned, linkage analysis may not have the power to detect all of them in an individual, leading to inconsistent results. Case-control studies also identified genes associated with sporadic (non-familial) AIS; however, there is a need to carefully interpret these results, as case-association studies can give rise to false-positive information. It is important to remember that for genetic association studies to be successful, one needs large sample sizes, small p values, reported associations that make biological sense, and alleles that affect the gene product in a physiologically meaningful way [16]. Therefore, many such studies and identified genetic risk factors need further confirmation with larger samples and different populations.

With the completion of the human HapMap project and recent new technologies development, one can foresee that case-control studies using a set of high-density SNP marker to cover the whole genome (Genome Wide Association Study or GWAS) will become an increasingly popular approach. Using this approach, a risk locus (rs10510181 in the proximity of the *CHL1* gene) was discovered in 419 AIS families [71]. Another SNP (rs11190870) near *LBX1* gene with OR = 1.56 in Japanese was detected by GWAS [76] and was replicated in Chinese [19, 21]. A meta-analysis of rs11190870 in six Asian and three non-Asian cohorts confirmed that both genders yielded $p = 1.22 \times 10^{-43}$, and $p = 2.94 \times 10^{-48}$ for females, and this would be the first susceptibility locus replicated by many data sets for AIS [42]. The third SNP (rs6570507) detected by GWAS was in *GPR126* (encoding G-protein-coupled receptor 126) and was discovered in Japanese (odds ratio (OR) =1.28), which was replicated in Han Chinese and European-ancestry

Table 2.1 Published linkage loci and genetic risk factors

Linkage regions	**Reference**
6p, 10q, and 18q	Wise et al. [88]
19p13.3	Chan et al. [10]
Xq23-26	Justice et al. [33]
5p13, 13q13, and 13q32	Miller et al. [45]
17p11	Salehi et al. [66]
9q31.2-q34.2, 17q25.3-qtel	Ocaka et al. [52]
8q12 (CHD7)	Gao et al. [22]
12p	Raggio et al. [64]
5q13-q14, 3q11-13	Edery et al. [17]
17q24.3	Miyake et al. [47]
Case-control studies	**Reference**
MATN1	Montanaro et al. [48] Chen et al. [11]
MTNR1B*	Qiu et al. [62], Takahashi et al. [77], Morocz et al. [50]
Estrogen receptor	Inoue [32], Wu et al. [89], Tang et al. [78], Zhang et al. [93]
G-protein-coupled estrogen receptor	Peng et al. [57]
IGF-I[a]	Yeung et al. [92], Takahashi et al. [77]
GH receptor	Qiu et al. [63]
Aggrecan	Marosy et al. [44]
TPH1	Wang et al. [82]
CHL1	Sharma et al. [71]
LBX1	Takahashi et al. [76], Fan et al. [19], Gao et al. [21], Londono et al. [42]
GPR126	Kou et al. [36]

[a]SNPs in the genes for MTNR1B and IGF-I were found to be associated with AIS in Chinese but not in other ethnics

populations [36]. We anticipate that more and more susceptibility loci will be discovered by GWAS or whole genome sequencing in future.

Conclusions

With international collaboration using well-defined phenotypes and large sample sizes, study of gene interaction and gene-environment interaction will be performed to speed the discovery of genetic risk factors for scoliosis. The study of genetic factors that predispose to scoliosis has come a long way in the past 10 years. Our understanding of the human genome and the development of new high-throughput techniques and computational methods have provided us with the tools to really embark on large-scale studies on scoliosis. The cause of idiopathic scoliosis and congenital scoliosis, factors that may influence their progression, will likely be elucidated in the not-too-distant future.

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Relevant Biomechanics to Growth Modulation

3

Brian Snyder, Robert M. Campbell, and Tricia St. Hilaire

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Key Points

- Growth modulation systems use engineering principles to maximize correction, modulate growth, and avoid implant failure. Customize the structural properties of the device, the mode of fixation, and loading conditions to the individual patient's needs to attain treatment goals.
- Avoid sharp transitions in the structural geometry of the system that could lead to stress risers, and minimize the contact of dissimilar materials to decrease potential for corrosion.
- In distraction-based systems, the rod diameter should be large enough to have the structural rigidity capable of withstanding biomechanical loads seen over time. Over time, rods of increasing diameter must be substituted based on patient size and activity level to compensate for increased working length.
- In compression-based systems that inhibit growth, the treating physician should also have an idea of the expected growth remaining in each spine segment

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to be able to predict expected correction over time and avoid overcorrection.
- The interface between both compression- and distraction-based systems and their attachment points to the patient is complex. Stability and strength of screws, hooks, or rib cradles depend on directional load and reactive properties of the bone to stress.
- New guidelines and procedures developed specifically for pediatric patients are needed so that the biomechanical properties of growth modulation systems can be compared in a consistent manner.

3.1 Background

In children with complex early onset scoliosis (EOS), treatment is a challenge. When surgical correction is indicated, the goals are to maximize correction, preserve thoracic growth, and avoid complications such as device dislodgement, fatigue fractures, infection, and other soft tissue problems. Spinal arthrodesis is seldom the best option for young children, as it inhibits thoracic growth contributing to decreased pulmonary function [1, 2]. The concept of growth modulation is based on the goal of maintaining the growth of the spine, lungs, and thorax and preserving mobile spine segments. Growth modulation systems, such as growing rods, VEPTR, MAGEC, SHILLA, staples, and tethers, offer many options for treatment and can be distinguished by their anatomic placement and loading mode. These systems may be used independently or in combination, both anteriorly and/or posteriorly. They may be used to distract the concavity or compress the convexity of the spinal deformity. Systems may also be used in a static state or change dynamically over time.

In determining the best system for a child with early onset scoliosis, the surgeon should consider the applied forces and potential growth patterns of the spine and thorax in that child. Unfortunately, the interaction between mechanical loading and biology is not well understood. Mechanotransduction describes how manipulating the stress (or strain) state affects biology. This chapter will focus on the biomechanics of growth modulation devices and how their application affects the efficacy of treatment and preservation of spine and thoracic growth. There are two aims: (1) assist physicians in understanding the basic biomechanical principles relevant to choosing the correct system for their patient that corrects spinal deformity, maintains growth, and avoids device complications both in the short and long term and (2) establish the groundwork of potential performance criteria for new growth modulation devices.

3.2 Engineering Principles of Material Properties

The modulus of elasticity (E), or Young's modulus, is a measure of stiffness and is defined by the ratio of stress over strain. It is a constant value for a specific material and is not related to the size or shape of the device. A lower modulus of elasticity makes the implant more flexible and closer to that of bone, therefore minimizing stress shielding. Compared to bone, titanium is about four to five times stiffer, while stainless steel is about ten times stiffer. Stress shielding occurs when there is a large difference in modulus between the implant and bone, resulting in a redistribution of load away from the bone, removing the normal stress. This can result in osteopenia and/or failure of the device. Fractures just proximal or distal to a bone plate are due to this "stress shielding" effect. The shear modulus (G) indicates the stiffness of a material in shear and is defined by the ratio of shear stress over shear strain. A shear force is a force applied perpendicular in line with a surface.

Stress values that are used in device design to predict failure are referred to as strength values.

Yield strength – stress where plastic deformation begins

Ultimate tensile strength – maximum stress a material can withstand before failure

Fracture strength – stress at which material failure occurs

These strength values are critical in defining material behavior.

When selecting a device, the type of metal or other material should be considered. The main metallic biomaterials used in growth modulation systems are titanium (Ti) alloys. While there are over 25 titanium alloys, two are predominant in orthopedic implants, TAN (Ti-6Al-7Nb) and TAV (Ti-6Al-4 V). Pure titanium, cobalt alloy, nitinol, and stainless steel are used to a lesser extent. Titanium alloys are used because of their biocompatibility and corrosion resistance. When compared to stainless steel, titanium alloys have a higher tensile and yield strength. This helps Ti implants withstand cyclic loading over an extended period of time. Titanium also has a lower modulus of elasticity (E) as compared to stainless steel, by approximately 50 %. Another important reason to consider titanium is that it is non-ferromagnetic, so MRI scans can still be used diagnostically in patients with growth modulation implants. Cobalt-chrome alloys are also non-feromagnetic (Table 3.1).

In addition to material, the surgeon should consider the structural geometry of the implanted device. The cross-sectional shape (A) and thickness of a rod affect the ability of the rod to sustain axial, bending, and torsional loads. The area moment of inertia (I) describes the spatial distribution of a material relative to a bending axis, called the neutral axis. The farther the mass is located from the neutral axis, the larger the moment of inertia about that axis. The polar moment of inertia (J), or second moment of inertia, is a measure of a material's ability to resist torque, a twisting moment about the neutral axis that produces shear stress. The larger the polar moment of inertia (rod thickness), the better the rod's resistance to torque. For the cross-section of a circular rod, a small increase in thickness of a rod makes a large difference in rigidity since the radius has a fourth-power effect on the moment of inertia and polar moment (Fig. 3.1). For example, a 5-mm uniform diameter rod is 1.5 times more rigid than a 4.5-mm rod made of the same material in resisting applied bending moments.

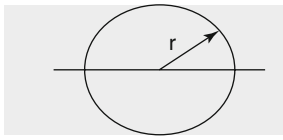
Table 3.1 Modulus of elasticity (E) for common orthopedic biomaterials

Orthopedic biomaterial	Modulus of elasticity
Ceramic	400
Co-Cr alloys	210
Stainless steel	190
TAV (Ti-6Al-4 V)	110
Tendon	50
Cortical bone	12–24
PMMA bone cement	2.2
Ultrahigh molecular weight polyethylene (UHMWPE)	1.2
Cancellous bone	0.005–1.5

When a rod is subjected to a torque or twisting, such as when a child with a growing rod turns his or her trunk in play activities and one end rotates relative to the other, a shear stress is induced on the cross-section of the rod. The shear stress varies from zero in the axis to a maximum at the outside surface of the rod. Rod breakage can occur because of shear stress alone or in combination with bending of the rod (Fig. 3.2).

Each rod used to correct a deformity is able to withstand a certain amount of load as determined by the shape and composition of the rod. The load is the force that the deformed spine places on the rod as well as any additional forces from the child's play activities. The structural rigidity determines the load capacity of the rod. It is a function of the material modulus *and* the rod geometry. Structural rigidity takes into consideration the loading mode: axial, bending, and torsion. Axial rigidity (EA) is the product of E×A. It reflects the ability of the device to resist axial loads in line with the longitudinal axis of the rod. Tensile or compressive axial loads are not usually a failure mode in growth modulation systems. Bending rigidity (EI), the product of E×I, reflects the ability of the device to resist bending moments applied perpendicular to the central axis of the rod. Most device failures occur when the bending rigidity of the device is too small to resist the applied bending moments. Torsional rigidity (GJ) is a product of G×J, and it reflects the ability of the device to resist torques around the axis of the rod. The more rigid a structure, the less it twists under a specific torque. Torsion may lead to device failure when a rod changes from a small radius to a larger radius in a non-gradual way (Fig. 3.3).

a

Shape	Area moment of inertia (I)	Polar moment of inertia (J)
	$I = 1/4\,\pi r^4$	$J = \pi / 2\, r^4$

b

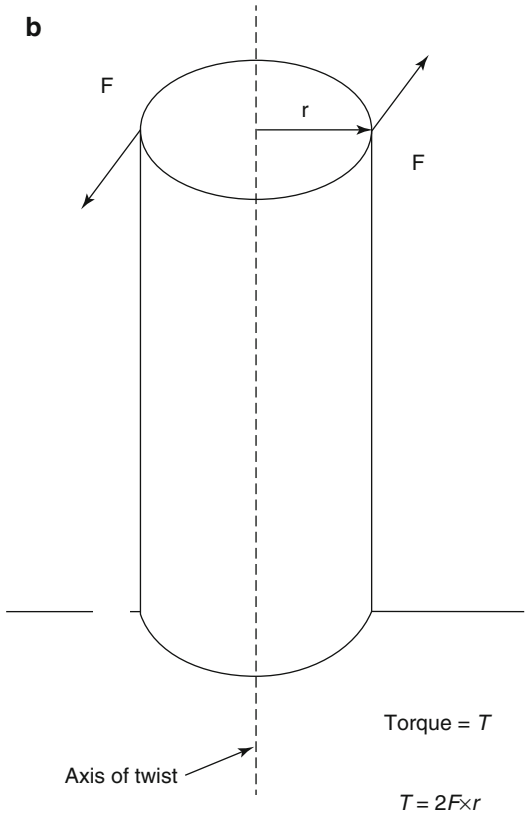


Fig. 3.1 (**a**) Moment of inertia and torque can be calculated using the shape of an object. (**b**) Torque is created by two forces acting in opposition around an axis of twist (Reprinted from Lucas et al. [3]. With permission from Springer)

Failure can also occur at a stress concentration. A stress concentration is created when there is an abrupt change in structural geometry, such as cross-sectional diameter. It can also be induced by stress risers predisposing the device to failure. Localized stress concentrations may occur at holes, bends, and coupling sites (Figs. 3.4 and 3.5).

Fatigue failure, the most common mechanism of rod breakage, can occur in a material subject to a cyclic stress. Although the peak stress in each cycle is less than what is needed to make the material fail in a static test, constant repetition of load weakens the material and causes a sudden failure. It is like repetitively bending a coat hanger wire and eventually having it break. The fatigue life of a system depends on the fluctuation of stress, the mean stress level, and the way it varies over time. In an active child, small degrees of rod bending and torsion with normal activities can fracture a rod in time. Fatigue failure can be accelerated by stress concentrations, stress fluctuations, corrosion, and surface stress.

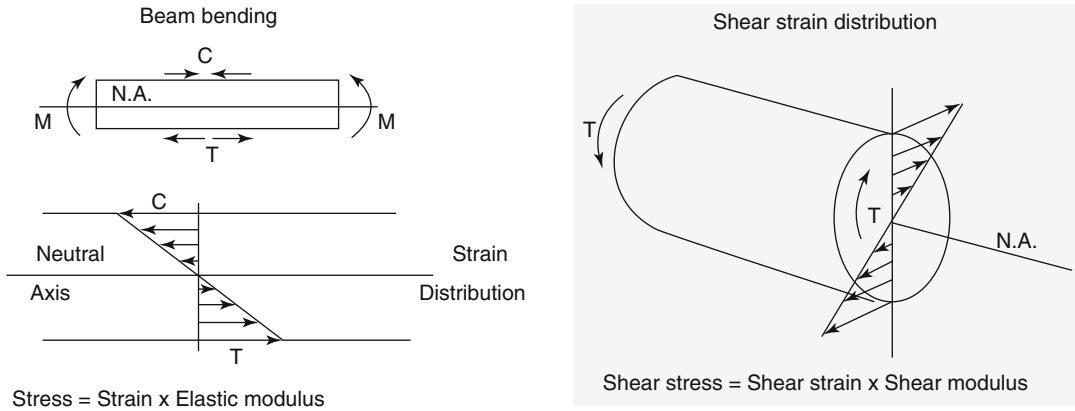


Fig. 3.2 Shear stress is the largest on the outside surface of a rod. Failure can occur because of shear stress or due to a combination of shear stress and rod bending

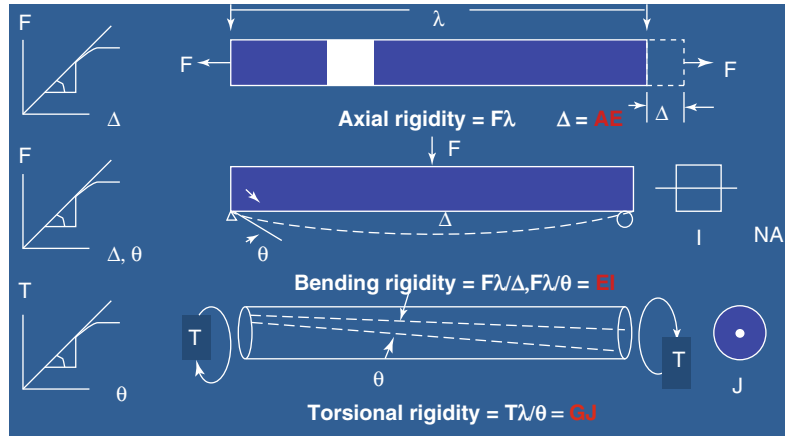


Fig. 3.3 Structural rigidity (modulus × geometric property). The more rigid a rod, the less it will twist. Failure of a rod may occur when there is a rapid transition between a small radius and a large radius

Another factor that can contribute to system failure is corrosion. Corrosion can occur because of a combination of both material and structural properties of the device. Corrosion is a result of the interaction between the metallurgical, mechanical, geometric, and electrochemical properties of the implant. When corrosion occurs, the material weakens. While the majority of materials used in growth modulation systems are Ti-based, the fabrication and finishing processes can vary widely. When two dissimilar metals are in contact, such as steel and titanium, an electrochemical reaction or galvanic corrosion can occur. A second type of corrosion, mechanically assisted crevice corrosion or fretting, happens as a result of relative motion of two metals. Modular junctions increase the potential for fretting. This happens across a mechanical joint, such as a screw and a rod, when intermittent load is applied. Corrosion can lead to the failure of the device because of material weakness, wear due to particles released from moving surfaces, and/or localized inflammation and osteolysis.

3.3 The Engineering Principles of the Spine and Its Remodeling Response to Deformity

The spine has six degrees of freedom (DOF). It can rotate axially, laterally, and sagittally and can translate (move) axially, laterally, and anteroposteriorly. A functional spinal unit (FSU) is made up of superior and inferior vertebrae and an intervertebral disc and has two DOF. Each FSU, with the

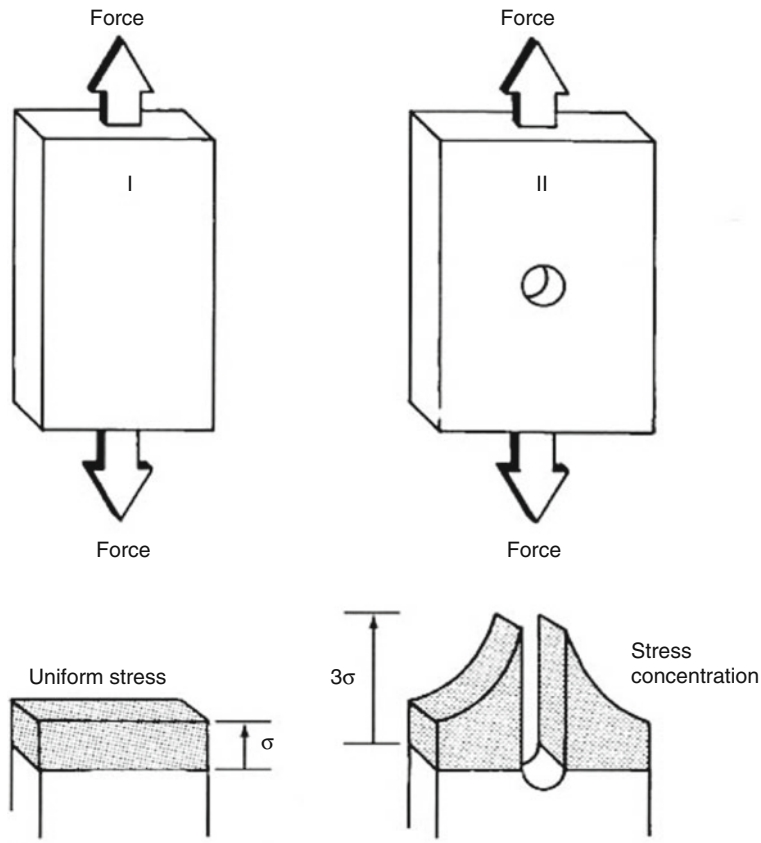


Fig. 3.4 Stress concentration. In the first image, uniform stress is applied across the entire block. In the second image, the center hole creates a stress concentration creating a much larger stress around the area of the hole

exception of the cervical spine, is supported by ten ligaments. These ligaments protect the FSU by restricting motion and absorbing energy when loads are out of the normal range. The rib cage, attached to the spine through joints and ligaments, also increases the stability of the spine by restricting motion in all directions. Specifically, the costovertebral joint is critical in limiting motion during flexion and extension [4]. Using computer simulations, Andriacchi et al. [5] determined that the stiffness properties of the spine were greatly improved by the rib cage during flexion, extension, side-bending, and axial rotation [6].

The three-column concept of Denis assists in assessing the stability of the FSU when subjected to distraction, flexion, and extension forces and moments. Usually the neutral axis is located in the mid-dorsal region of the vertebral body. It bears much of the axial load and does not significantly distract or compress during flexion or extension.

When an applied constant load is placed on the spine over a period of time, there will be deformation. This is defined as creep. In scoliosis, a force is applied to correct the deformity. After the initial correction, a constant force remains on the spine. Over time, the progressive correction of the deformity that occurs is due to creep. When a constant load is applied, there is a continued progressive strain. Clinically, this occurs in the case of halo traction. When the initial applied load reduces over time, this is called stress relaxation. Clinically, this has implications for sequential or a series of incremental applied loads (Figs. 3.6 and 3.7). When constant deformation is applied, stress progressively decreases from initial load. Clinically, this occurs when spinal instrumentation loosens during scoliosis correction.

Creep reflects the viscoelastic nature of a material. While bone is weakly viscoelastic, soft tissue behaves partially like a solid and partially like a

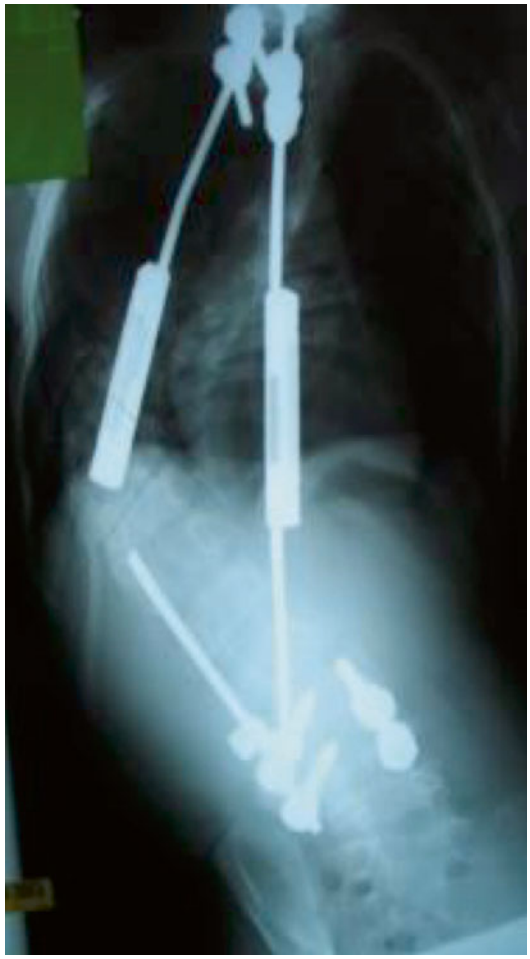


Fig. 3.5 Clinical example of a stress concentration. When a device changes abruptly from a smaller diameter to a larger diameter, a stress concentration is created. Here it is shown that the abrupt transition of the coupling point to the rod caused the device to fail. A rod of larger diameter may have minimized the potential for failure by reducing the stress concentration between the coupling and the rod

liquid and exhibits viscoelasticity. Viscoeleastic materials withstand higher loads to fracture and less elongation when stretched faster.

A compressive load is transferred between vertebral endplates through the intervertebral disc (IVD). Excessive compression will lead to degradation of the disc. The biomechanics of the disc change when subjected to bending or torsion. When bending loads are applied during flexion, extension, and lateral bending, the discs are subjected to both tension on the convex side and compression on the concave side. The torsional stiffness of the spine is mainly dictated by the orientation of the facets, which determines how much rotation can occur. Facets limit the degrees of freedom of motion. Facet design changes when comparing the thoracic spine to the lumbar spine. Thoracic facets are oriented in frontal plane and permit lateral bending and torsion. Lumbar facets are oriented in the sagittal plane and permit flexion and extension. The greatest change in orientation is seen at the thoracolumbar junction. Biomechanically, this creates a sudden change in stiffness creating a stress concentration. This is why the highest frequency of spine injury occurs here. Orientation of facets leads to coupled motion. Coupling occurs when two or more individual motions (i.e., lateral bending and rotation) occur simultaneously and cannot be produced individually. In scoliosis, there is twisting and warping as a consequence of this coupled motion. Surrounding muscles act as dynamic springs and dampen or attenuate deformation in response to applied load. At slow rates of loading, the vertebral body is more likely to fail. At higher rates of loading, muscles will be injured.

The surgeon must also take into consideration how the spine growth will be modulated by the applied mechanical loads. The Hueter Volkmann Law describes how loads affect the growth of bones. This was demonstrated in a study that showed how epiphyseal growth is affected by tensile and compressive forces [7]. Increased pressure or compression inhibits growth, whereas decreased pressure or tension accelerates growth. Using a rat tail model, Stokes et al. [8] showed that sustained compression of physiologic magnitudes inhibited growth by 40 % or more, while distraction increased growth by a much smaller amount.

In contrast, Wolff's law states that bone remodels over time in response to mechanical loads. When the trabeculae are oriented along principal stress trajectory, bone is subjected to loads above or below what is normal and the bone responds. For example, increased intermittent stress stimulates bone formation where reduced intermittent stress causes bone resorption. For example, the ribs of attachment for the VEPTR tend to thicken with time, reflecting the

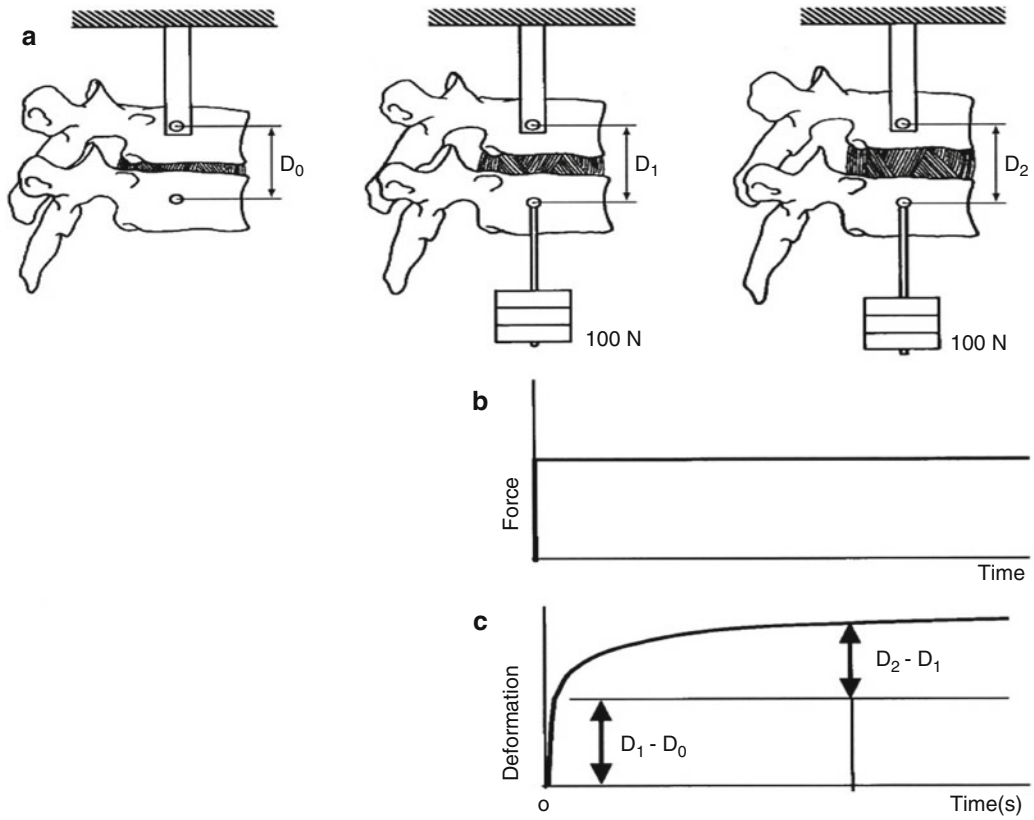


Fig. 3.6 (**a**) Creep is the deformation that occurs over time when there is a constant applied load, as in halo traction. (**b, c**) show that when a constant force is applied there is continued deformation over time

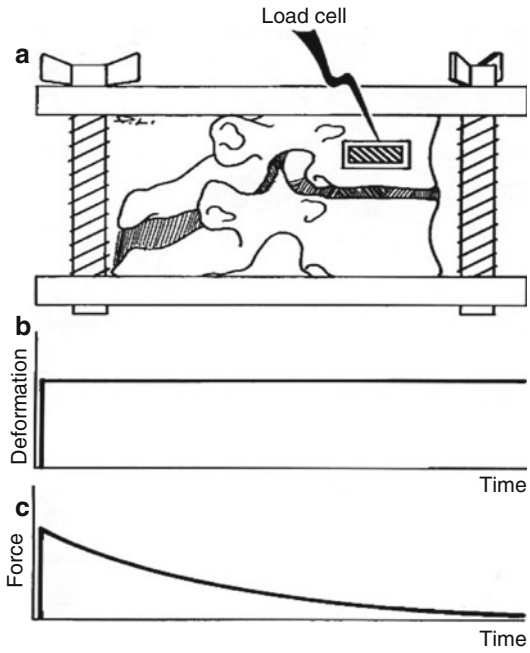


Fig. 3.7 When an applied load reduces over time, stress relaxation occurs

reaction to stress of the rib cradle on the ribs. Adjacent muscles neutralize tensile loads and allow bone to carry increased compressive loads.

Correction obtained by growth modulation is dependent on growth remaining at each level. This dictates the number of FSUs and time of inhibition required to correct a deformity. Balasubramanian and Dimeglio estimated the growth per level per year, and this information can be used to mathematically predict the number of levels and years required to correct asymmetrical growth of the spine.

3.4 Specific Device Constructs and Their Response to Load Application Over Time

How the growth modulation system is fixed and loaded in vivo will have a large impact on the correction of the deformity and the longevity of

the device. However, the patient's anatomy, specifically the bone-anchor interface, limits the amount of distraction force that can be applied. Using a screw versus a hook at this interface will affect the amount of force that can be expended.

3.4.1 Bone Anchors

A pedicle screw is rigidly fixed in both cortical and cancellous bone. The outer diameter of the screw predicts pull-out strength, while the inner diameter determines fatigue strength. Screws function by converting the insertion torque into internal tension in the screw and into elastic reactions in the surrounding material [3]. In comparison, a hook is semi-constrained, allowing some motion or stress relief to occur at the bone-implant interface. Sublaminar wires are often used as a tension band across posterior spinal elements. As wire tension increases, it becomes a stiffer anchor point. For all anchors, the bone quality affects the stability of the bone anchor. Bone stiffness and strength vary with density squared. Using pedicle canal fill by using the widest screw possible maximizes the friction between the anchor and the bone, therefore minimizing the potential for failure.

In posterior-based systems, bending rigidity and the unsupported working length between anchor points dictate the stability of the system. Specifically, the longer the unsupported working length of rod is, the less rigid the system and the larger the deflection. Increasing the rod diameter or fixing the rod in the center of the deformity, as with Shilla, can create a more stable condition (Fig. 3.8).

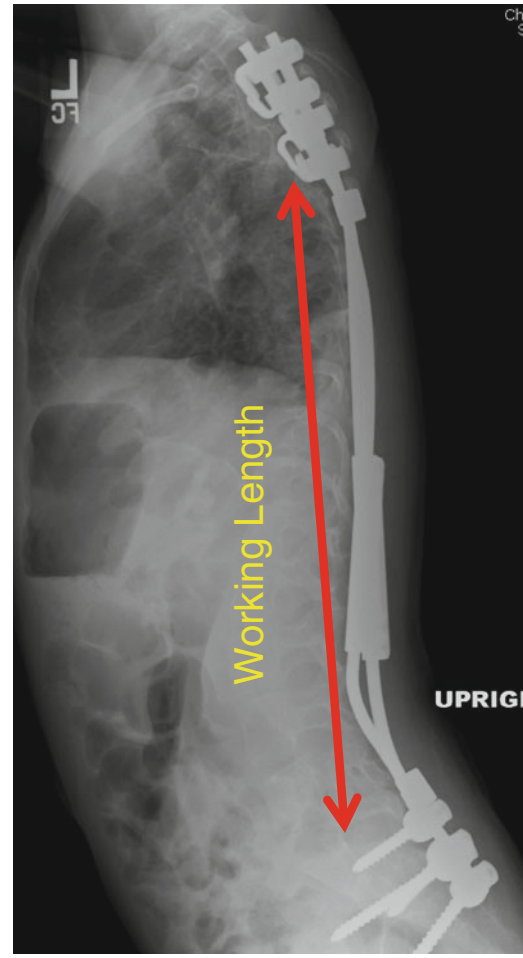


Fig. 3.8 Clinical example of unsupported working length. The longer the unsupported working length of the rod is, the less rigid the system and the larger the deflection. This increases the chances for rod failure

Anterior systems function much differently. Their mode of growth modulation is based on compression across the convexity in an attempt to correct the asymmetric growth of the spine. Tethers and staples are placed on the anterior spine along the convexity of the deformity, based on the Hueter Volkmann Law that compression inhibits growth. As was shown by Stokes, this can be a powerful mode of correction. A study published by Coombs et al. [9] studied the effects of a titanium clip-screw construct placed anteriorly on the biomechanical properties of porcine thoracic motion segments. The study analyzed the stiffness of the segments in flexion and extension, as well as the axial translation. It found that the device decreased range of motion by less than 20 % and increased stiffness by less than 33 %. In one published example, a patient with scoliosis treated with vertebral body stapling had overcorrection of the deformity due to the compressive force inhibiting the growth remaining to a greater extent than the initial deformity being treated [10]. An additional clinical report by Crawford and Lenke [11] using tethers to correct scoliosis acknowledges the potential for overcorrection in association with anterior tethering (Table 3.2).

Table 3.2 Growth modulation systems

Posterior systems	**Placement**	**Loading mode**
Rib-based (i.e., VEPTR)	Placed on concavity of the curve; connected superiorly at rib; inferiorly at rib, spine, or ilium	Static distraction loads at fixed intervals placed on superior and inferior connection points attempts to achieve and maintain immediate correction
Spine-based (i.e., growing rod)	Placed on concavity of the curve; connected superiorly and inferiorly at spine	Static distraction loads at fixed intervals placed on superior and inferior connection points attempts to achieve and maintain immediate correction
Spine- and rib-based magnetic (i.e., MAGEC)	Placed on concavity of the curve; connected superiorly at rib or spine and inferiorly at spine	Static distraction loads at fixed intervals placed on superior and inferior connection points attempts to achieve and maintain immediate correction
Spine-based; semi-rigid (i.e., SHILLA)	Placed on concavity of the curve; connected superiorly, inferiorly at spine. Connected also at apex of curve	Dynamic distraction over time at superior and inferior ends via sliding rods attempts to achieve correction over time
Anterior systems	**Placement**	**Loading mode**
Staples	Spans disc in vertebral bodies on convexity of curve	Dynamic compression at each instrumented vertebral segment attempts to achieve correction over time
Tether	Screws placed in vertebral bodies on convexity of curve	Dynamic compression at each instrumented vertebral segment attempts to achieve correction over time

Correction can be immediate by tensioning the tether which applies an initial corrective moment in compression, followed by correction that is passive, so that compression is generated as longitudinal growth is inhibited by the staple or the tether over time. Growth modulation depends on the number of endplates spanned and the growth remaining at each level. Some of the initial correction is related to compression of the IVD and stress relaxation. This limits the motion of the IVD and leads to degeneration. What is unknown is how persistent or responsive the apophysis is to "reversal" or removal of growth modulating forces. The compressed, dehydrated IVD may not be capable of re-expanding, and the inhibited cells at the apophysis may not be capable of turning back on. This may be dependent on the magnitude and duration of applied load and the age of the patient. Work performed by Newton et al. [12] showed that an anterolateral tether limits motion primarily in lateral flexion. When tether is removed, total lateral bending returned to levels similar to control. In subsequent research, tethered discs in a calf model had similar water content to control discs and did not demonstrate gross degeneration [13]. Wall also used an animal model to demonstrate that spinal hemiepiphysiodesis decreases growth plate and disc height [14].

The applied load that induces growth modulation creates a time-dependent increase in strain (tensile or compressive) resulting in continued deformation or viscoelastic creep. Over time, the applied load changes proportionately to the longitudinal growth that is inhibited (see Figs. 3.6 and 3.7). For distraction-based systems, it dissipates. While in compression-based (anterior) systems, the load increases. This load is dynamic over time and varies from patient to patient. In addition to the load required to modulate growth, the system must also withstand the multitude of superimposed loads and moments generated during daily life activity. As a result, it is unknown what loads and number of cycles the implant must withstand during the course of treatment for the growing child. Research performed by Marco Teli measured the forces applied during distraction of growing rods under general anesthesia.

Twenty measurements were obtained showing a linear increase of the load at each subsequent distraction. The mean peak force was 485 N at 12 mm of distraction [15]. A similar study found that the intraoperative force required at the fifth lengthening (368 N) of a growing rod construct was double that required at the initial lengthening and that the mean length distracted decreased over time [16].

3.4.2 The Theory of "Diminished Returns" in Growth Modulation Surgery: Is This a Valid Concept?

In the past, the classic outcomes in growth modulation surgery measured the ability to correct the Cobb angle of scoliosis, the growth rate of the T1-S1 spinal segment, and the complication rate of treatment. A new interpretation of expected outcomes has recently been published [17]. To the existing measures, the authors of this report have added a new implied outcome measure for successful growing rod treatment. The new measure suggests that the mechanical lengthening of the growing rod at each subsequent surgery must equal the original lengthening at implant surgery or, at a minimum, should not decrease with time. The findings of the study show a gradual decrease in the mechanical lengthening distances of the growing rods over time with lengthening surgeries (Fig. 3.9), and its authors have coined the term "The Law of Diminishing Returns" (LODR) to describe this observation, implying that the effectiveness of the growth modulation system is diminishing, suggesting that autofusion is the etiology.

Paradoxically, the report noted a reduction in the Cobb angle from 74° to 36° and a growth rate of the T1-S1 spinal segment of an average of 1.76 cm/year – considered a normal growth rate by the authors. Growth of the spine is critical for pulmonary outcome in scoliosis [1], so the latter is the most important. Complications were not mentioned, but based on the scoliosis correction and growth rate of the spine, their growth modulation treatment could be termed successful based on past accepted outcome measures. However, the article concludes that the mechanical problem with rod lengthening diminishes the overall effectiveness of growth modulation.

From a biomechanic perspective, the correction achieved at each growth rod lengthening distraction is a function of the moment arm and the force applied, which determines the corrective bending moment. The moment arm is correlated with the lateral offset, or the distance from the rod

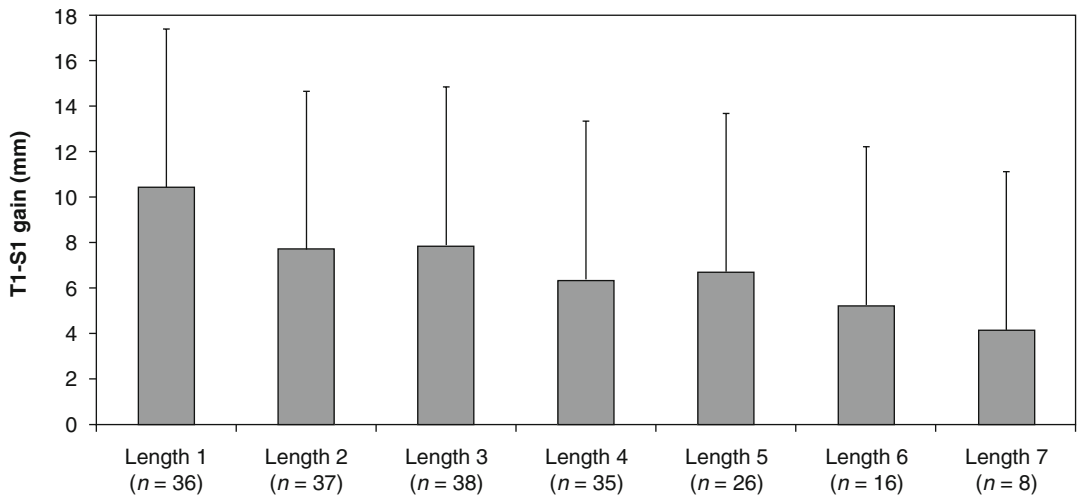


Fig. 3.9 The graph shows a gradual decrease in the mechanical lengthening distances of the growing rods over time with lengthening surgeries (Reprinted from Sankar et al. [18]. With permission from Wolters Kluwer Health)

to the apex of the spinal deformity (Fig. 3.10). As the spine straightens and elongates with each distraction, the moment arm decreases. Therefore, to maintain a constant corrective moment, the distraction force required needs to increase proportionate to the decrease in the lateral offset. In a way, early corrective success of the rods requires less force, "turning the spine" to correct a curve, but with subsequent lengthenings, the surgeon is "stretching" a relatively straight spine, creating more reactive force and actively axially loading the spine. More directly, using the analogy of a rope or cable, once the slack is removed by pulling fully to length, the axial stiffness increases. Distracting a straighter spine loads the IVD and facets. The corrective moment required to straighten the spine increases proportionate to the decrease in stress. Biomechanics helps explain why it gets harder to mechanically lengthen the growing rods once the spine is fairly straight from treatment. Therefore, the moment arm of distraction in growth modulation surgery is "diminished" by successful treatment. However, to extend this to the effectiveness of the total

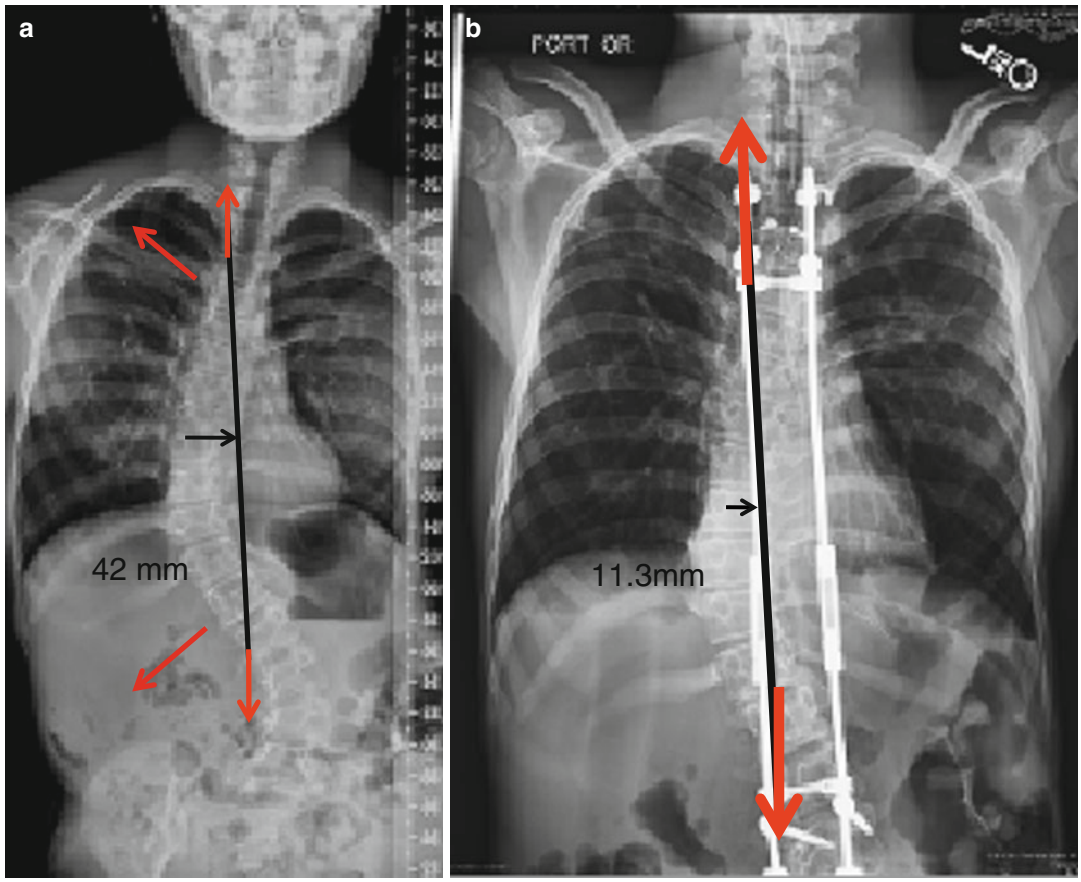


Fig. 3.10 Working length of rod and LODR. As the rods are distracted, the working length between anchor points increases, destabilizing the construct, yet more force is required to continue to achieve correction because of the decreasing moment. Corrective moment = distraction force × moment arm (lateral offset). (**a**) The moment arm from the pedicle of T9 is 42 mm to the planned axis of a growing rod. With this large moment arm, little force is needed for growing rod distraction, since it is "turning the spine." (**b**) Once the rods are inserted, then later at the time of lengthening surgery, the moment arm is diminished to 11.3 mm, and more force is needed for distraction since the relatively straight spine has to be "stretched" by lengthening distraction, elongating the intervertebral discs and the joint capsules

growth modulation system is overreaching. The topic deserves further critical study before accepting the theory of diminished returns. A recent study by Chukwunyerenwan et al. [18] found that although the AP spinal lengthening decreased over time with growth modulation surgery, supporting the theory of LODR, the changes were not as apparent in the sagittal plane X-rays, and true spine length increased considerably, suggesting that the AP deficit finding may be somewhat artifactual in nature. Parents should be reassured that growth modulation surgery is an effective treatment, and the LODR is controversial and, at this time, remains a theory that needs more evidence.

3.5 Reducing Growth Modulation Complications to the Minimum

It is important to understand engineering as applied to growth modulation systems to maximize correction, modulate growth, and avoid implant complications such as fatigue fracture and anchor point pull-out. The system, the application, and its use over time should be evaluated as a whole to make the best choice for the patient. Treatment objectives are often attained by customizing the structural properties of the device, the mode of fixation, and loading conditions to the individual patient's needs (see Fig. 3.5). A successful growth modulation system should avoid sharp transitions in the structural geometry of the system that could lead to stress risers and minimize the contact of dissimilar materials to decrease potential for corrosion. The rod diameter in posterior systems should be large enough to have the structural rigidity capable of withstanding biomechanical loads seen over time. In distraction-based systems, since lengthening the rod increases the working length, which increases the stress on the construct and destabilizes the construct, rods of increasing diameter must be substituted over time based on patient size and activity level. When using anterior systems that inhibit growth, the treating physician should also have an idea of the expected growth remaining in each spine segment to be able to predict expected correction over time and avoid overcorrection.

3.6 The Timing of Growth Modulation Surgery and Its Effect on Implant Complications Testing: What Is the Intended Duration of Use of the Device?

The duration of device use has a large influence on the incidence of growth modulation spine instrumentation complications: fatigue failure, deformation such as bending, and anchor point pull-out or migration. The longer the instrumentation "life-span" needed, the higher the risk of breakage or pull-out. Existing guidelines, established by the American Society for Testing and Materials (ASTM), evaluate static and dynamic (fatigue) strength. Recommendations are based upon spinal fusion constructs used in adults. Growing constructs undergo three static and one fatigue mechanical test, as governed by ASTMF1717. The static tests are compression bending, tension bending, and torsion. The fatigue test is a compression bend test at a constant load ratio for a minimum of five million cycles. The constructs are fixed in a corpectomy-like model. Posterior spinal constructs, both for fusion and growth, are tested using the same parameters, regardless of intended use or patient (i.e., adult vs. pediatric). The assumption in spinal device testing is that clinically, a load-sharing situation develops as the fusion heals, with fusion mass taking over the load from the instrumentation as it matures. Therefore, while the fatigue resistance of spinal rods used for spinal fusion does not need to last a lifetime, the fatigue resistance for non-fusion growth modulation rods may need to be greater.

The load forces in the pediatric spine from activities of daily living are poorly understood but likely include marked sagittal as well as lateral flexion and extension, rotation, and compression. The change in these loads due to spinal deformity is also poorly characterized, but likely, they are

increased and become eccentric in nature. Growth modulation instrumentation, for the most part, is not shielded by fusion mass and must be able to withstand repetitive complex loads encountered in pediatric patients for the lifetime of the instrumentation, often years. Past testing has been limited. The VEPTR device, "over-engineered" to best survive long-term use with the hybrid version having a 6-mm-diameter titanium alloy rod to resist fatigue failure, has a very low rate of breakage. Growing rod pedicle screw pull-out has been studied [19], but no fatigue testing is available for growing rods to our knowledge, and limited test information is available for any other form of growth modulation instrumentation. Until the forces experienced by growth modulation instrumentation are better characterized and standardized testing is developed for them, including the number of cycles needed for valid fatigue testing, it is best to design for the worst case scenario, adding strength through component thickness and avoiding sharp changes in transition points to avoid fatigue fracture.

3.6.1 What Are the Clinical Implications of the Timing of Intervention?

While there is a theoretical biomechanical benefit to limiting growth modulation instrumentation lifespan, the clinical ramifications must also be considered. Two schools of thought have emerged: those who argue that early intervention is best to aid lung growth and pulmonary function by correcting deformity at a very young age and those who argue that late intervention is better since it shortens total duration of instrumentation use so there is statistically decreased risk of device breakage and pull-out and probably less risk of soft tissue problems such as infection and skin slough. Very little specific data exist to support either position. For the "early intervention" camp, one VEPTR series of fused ribs and scoliosis, it was noted that those operated on below the age of 2 years had an FVC% predicted of an average of 58 %, but for those operated on after the age of 2 years, the FVC% was 44 % [20]. Motoyama et al. [21] reported the results of 24 VEPTR patients with an average age of 4.6 years (1.8–10.8 years) at first surgery and, at an average of 3.2-year follow-up, found that those younger than 6 years at the time of first surgery had a 14 %/year increase in percent predicted FVC, while those older than 6 years at time of surgery had only a 6.5 %/year increase. To our knowledge, the late intervention camp does not appear to have pulmonary literature supporting their position. Until more data are available about these issues, the timing of intervention with growth modulation instrumentation will remain controversial, but it is recommended that parents of very young children with severe scoliosis be made aware that lung growth is rapid early in life, and the constriction of the chest by the scoliosis may have a negative effect on lung growth.

3.7 The Future

In the development of new systems for the growing child, there is a need for sound performance criteria based on these principles. Preclinical testing is essential to ensure that devices are able to withstand both the corrective and superimposed forces seen during distraction and over time. Mechanical testing would ideally reproduce the same stress levels and fluctuations that are seen in vivo. New guidelines and procedures developed specifically for pediatric patients are needed so that the biomechanical properties of growth modulation systems can be compared in a consistent manner. New growth modulation systems, such as the magnetically controlled devices [22], show promise but require the same careful biomechanical study as older systems to optimize performance.

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Normal Growth of the Spine and Thorax

4

Alain Dimeglio, Federico Canavese, and François Bonnel

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Key Points

- The growing spine is a mosaic of physes.
- Spine and thoracic growth are interrelated. The thorax is part of the spine; it is the "fourth dimension of the spine."
- T1–S1 segment is a strategic segment; it makes up 49 % of the sitting height at maturity.
- The T1–T12 segment represents 30 % of the sitting height, and the lumbar spine represents 18 % of the sitting height.
- About 50 % of trunk growth occurs during the first 5 years of life. It is a critical period for early-onset spinal deformities.
- As the spinal deformity progresses, by a "domino effect," not only spinal growth is affected but size and shape of the thoracic cage are modified as well.
- The pathologic spine is dominated by the crankshaft phenomenon.

4.1 Growth Holds the Basics

It is growth that distinguishes pediatric from adult orthopedics. It is this ongoing 17-year adventure, punctuated by upheavals, that gives this discipline its originality and makes it so interesting. Growth analysis is the evaluation of the effects of time on the growing child. Growth is a complex and well-synchronized phenomenon with a hierarchical pattern that organizes the different types and rates of growth in various tissues, organs, and individuals through time [6, 20, 26].

Growth can be considered as "microgrowth," which is mainly the growth at the cellular level (e.g., in the physes). Although the histologic structure is the same, each physis has its own characteristics and dynamics [20]. The study of height, weight, and body proportions may be considered as the study of "macrogrowth." This study is the culmination of all the effects of microgrowth on the individual: the combined effect of growth of the lower limbs, the trunk, and the upper limbs, increase in weight, and so on [20, 26].

The scope of this process called *growth* and the changes it brings about can be better perceived by considering these facts: from birth onward, height will increase by 350 %, weight will increase 20 fold, and the spine will double in length [24, 26].

Growth is an essential element in the natural history of any orthopedic disorder in the growing child [24, 26]. It would be a mistake to assume that only growth in terms of increase in height is important. It is equally important to consider the manner in which the skeletal system develops, that is, the timing of growth in various parts of the body and the changing proportions of various body segments.

The spine surgeon needs to know the normal values for many parameters and how to measure them. He or she needs to know the significance of these values, for example, the effect of a ten-level spinal fusion in a boy who has a bone age of 10 years. Bone age, Tanner classification, stages of puberty, and measurement of the upper and lower portions of the body are all parameters that may need to be considered in the analysis of any particular case [20, 24, 26].

Knowledge of the synchronization of the various events in growth will also allow the orthopedist to anticipate certain events, for example, the onset of puberty characterized by an increase in growth velocity in a girl with early breast development. However, these values vary with the individual, and average values may not apply to a particular individual. What is most important is the pattern and rate of growth for a particular individual. It is the rate of growth that will influence orthopedic decisions, more than the final height. A sequence of measurements of the important parameters is far superior to a single measurement [20, 24, 26].

The criticism often directed at growth data is that such data are ethnically specific, and it is difficult to transfer parameters from one population to another. For example, bone age atlases are not

Table 4.1 Clinical examination must answer these basic questions

How tall is the child?
What is the child's sitting height?
How long is the subischial leg length?
How much has the child grown in a single year?
What is the child's chronologic age?
What is the bone age?
How much growth does the child have left in the trunk and in the lower limbs?
Exactly what point has the child reached on his or her developmental peak?
Where is the child in relation to puberty and the pubertal peak?
What about the Tanner signs?
Are the child's proportions within normal limits?
How much does the child weigh?

transferable between populations nor are growth curves, from one country to another. A comparison of data relating to children in England, Switzerland, France, and the United States reveals no significant differences in final heights, bone ages, or other parameters of growth [33, 50, 55, 56, 58]. Looking beyond racial diversity, there are growth constants (i.e., stages through which every child must pass regardless of chronologic age) that are the same in ethnic groups.

A few simple tools are required at the time of consultation: a height gauge, scales, a metric tape, and a bone age atlas. With these tools, the specialist will be able to form a rapid mental arithmetic and reach a reasonable decision. A few simple questions will provide the orthopedic surgeon the information that is required [20, 24, 26] (Table 4.1).

4.2 Biometric Measurements

There is not much useful data that can be obtained from a single measurement. A single measurement can be an error, and two measurements constitute an indication, while three measurements define a tendency.

Measurements of growth should be taken at regular intervals. Checking the child every 6 months, one of the two checkups being preferably around his or her birthday, allows an easy assessment of the growth velocity of the child and the different body segments [20, 24, 26]. These measurements provide a real-time image of growth, and when carefully recorded in a continually updated "growth notebook," they provide charts that make decisions easier. Growth velocity is an excellent example, because it provides the best indicator of the beginning of puberty, on which so many decisions rest. The first sign of puberty is the increase in growth rate of the standing height to more than 0.5 cm/month or more than 6 cm/year.

The spine surgeon should be familiar with the measurements of these parameters. Regarding standards of growth, several good references are available [3, 20, 24, 26, 35, 37, 50].

4.2.1 Standing Height

Standing height is necessary but is not sufficient to assess growth.

Measuring the height is to the orthopedic specialist as listening to the heart is to the cardiologist. In children younger than 5 years, standing height is measured with the child lying down because in this age group, this position is both easier and more reliable [20, 24, 26].

Between birth and maturity, the body will grow by approximately 1.20 m, or even 1.30 m. Growth is brisk up to 5 years of age. After that, it slows considerably until the onset of puberty, which occurs at approximately 11 years in girls and 13 years in boys. At 2 years of age, the standing height is approximately 50 % of the adult height; at 5 years of age, it is approximately 60 %; by the age of 9 years, approximately 80 %; and at puberty, approximately 86 %. In this latter period, standing height increases more rapidly.

Standing height is a global marker and is composed of two specific measurements known as subischial height (i.e., the growth of the lower limbs) and sitting height (i.e., the growth of the trunk). These two different regions often grow at different rates at different times, which is valu-

able information for decisions in orthopedics. Values for the standing heights of girls and boys at various ages were given in previous publications [59–61].

4.2.2 Sitting Height

Sitting height is the most reliable parameter to monitor trunk growth.

In children 2 years of age or younger, the sitting height is measured with the child lying down for the same reasons that the standing height is also measured supine in this age group. After 2 years of age, the child to be measured should be placed on a stool or table at a convenient height. The most important consideration is that the child should always be measured under the same conditions using the same measuring instruments. The sitting height averages 34 cm at birth and averages 88 cm for girls for a standing height of 165 cm at skeletal maturity and 92 cm (sitting height) for boys at the end of growth for a standing height of 175 cm [20, 22, 24, 26, 52] (Figs. 4.1, 4.2 and 4.3).

In patients with scoliosis, it can be instructive to follow the changes in the sitting height rather than in the standing height. If a 6-year-old girl with juvenile scoliosis is being treated, her sitting height will be approximately 64 cm and will increase to about 88 cm. Therefore, the spine surgeon will have to control the spinal curve while her trunk grows 24 cm. The measurement of sitting height can also be useful in anticipating the onset of puberty. In an average population, puberty starts at approximately 75 cm sitting height in girls and 78 cm in boys. When the sitting height is approximately 84 cm, 80 % of girls have menarche (Figs. 4.4, 4.5 and 4.6).

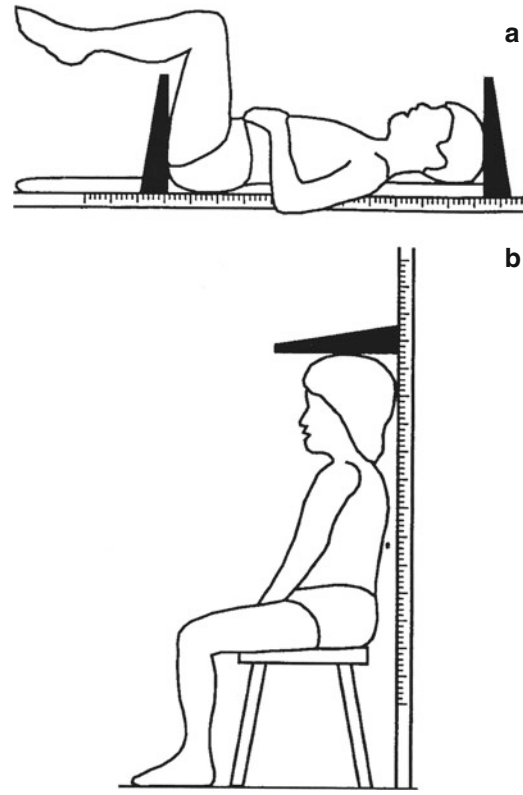


Fig. 4.1 Sitting height measurement: in children younger than 2 years (**a**) and in children older than 2 years (**b**)

4.2.3 Subischial Limb Length

Where does growth come from? Is it from the trunk or from the legs?

The segment of the body consisting of the lower extremities is measured to determine the subischial limb length. As implied by the name, subischial limb length is measured by subtracting the sitting height from the standing height.

At birth, the subischial limb length averages 18 cm. At the completion of growth, it will average 81 cm in boys and 74.5 cm in girls. These 63 cm of growth in boys and 56.5 cm of growth in girls contribute to a far greater percentage of growth in height than does trunk growth. This accounts for the changing proportions of the body during growth (see Figs. 4.4, 4.5, and 4.6).

4.2.4 Arm Span

Measuring arm span is useful in non-ambulatory children.

The measurement of arm span provides an indirect control parameter for the measurement

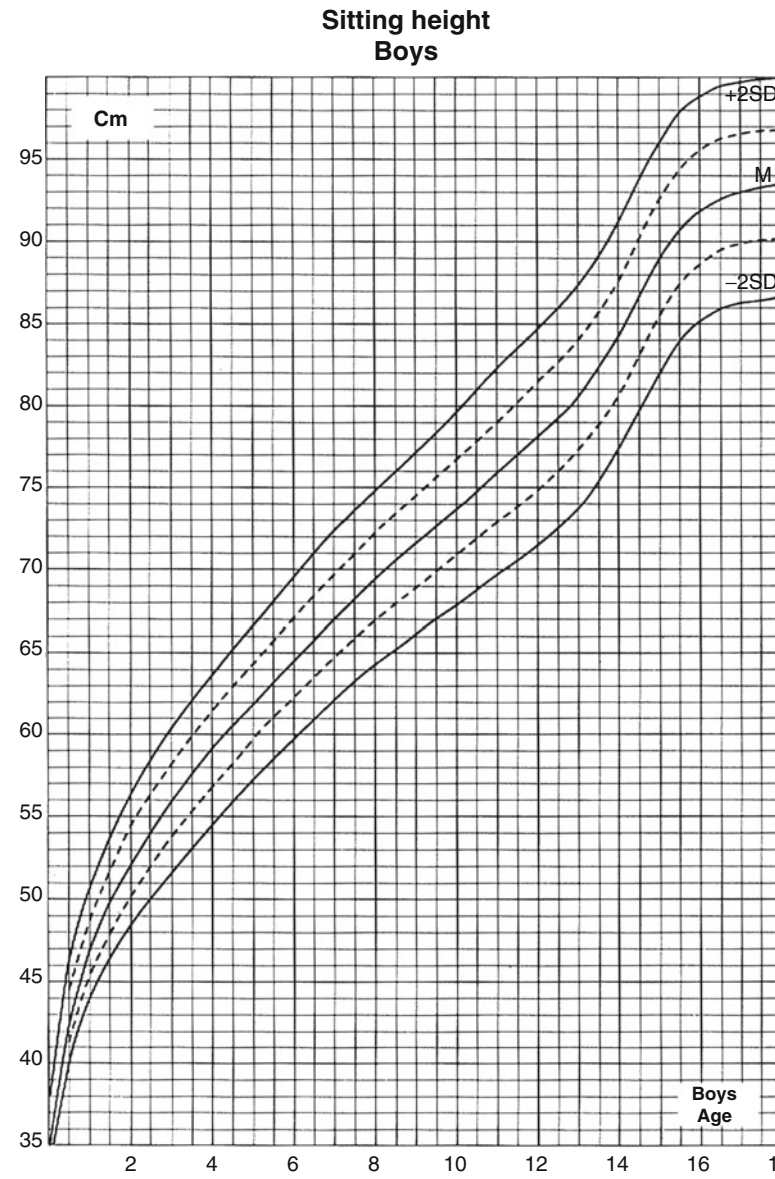


Fig. 4.2 Sitting height-for-age (birth to 18 years: boys)

of standing height. Combining these two measurements avoids virtually all errors. To measure arm span, the patient simply raises the arms to a horizontal position, and the distance between the tips of the middle fingers is measured with a tape measure. There is an excellent correlation between arm span and standing height, as standing height is about 97 % of arm span. If the trunk is normal (i.e., without deformity), its length will be equal to approximately 52 % of arm span, and the lower limbs will be equal to approximately 48 % or will be the same as their proportions in the standing height.

The relation of arm span to normal height is useful in determining the normal height of a child who is wheelchair bound; this allows the calculation of the child's height [39]. It is a routine used for any child who has a spine deformity (e.g., scoliosis) for calculating the normal values for pulmonary function. With spinal deformity, arm is a good estimate of what the standing height would be if there were no scoliosis.

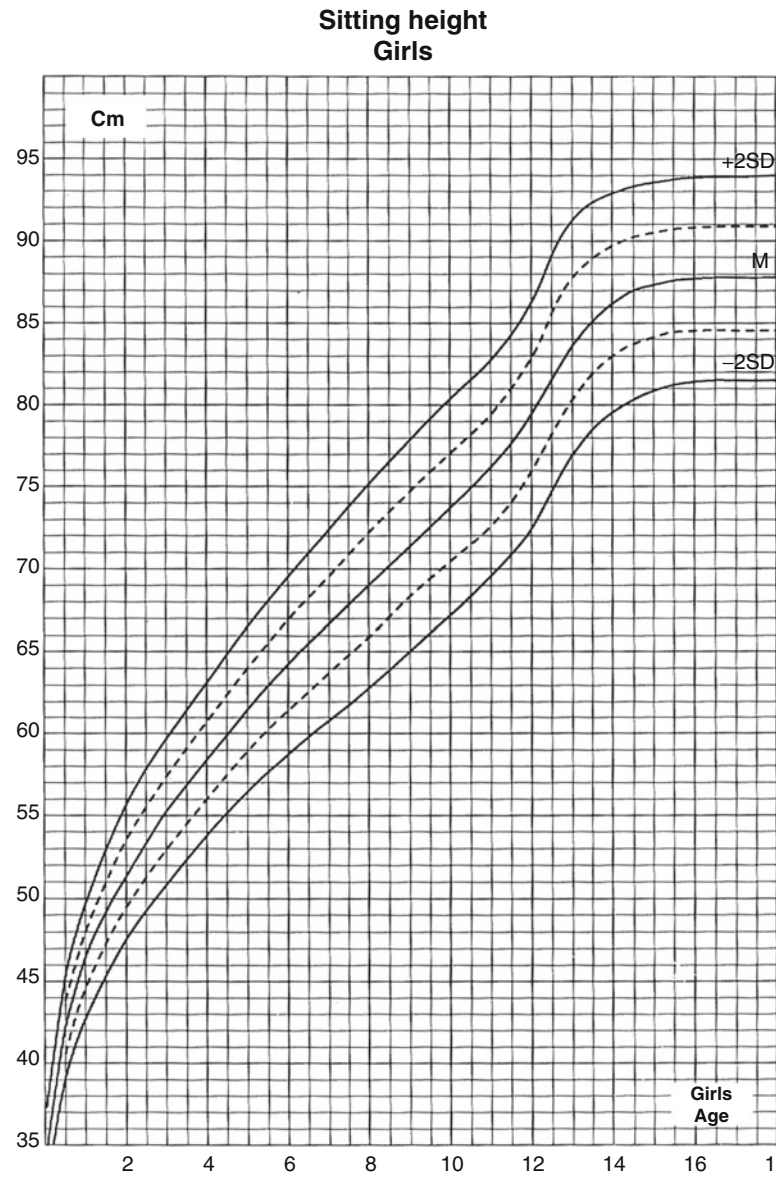


Fig. 4.3 Sitting height-for-age (birth to 18 years: girls)

4.2.5 Weight

Weight is often an underestimated parameter.

Weight should always be brought into the equation when making a surgical decision, whether the orthopedic is dealing with a case of idiopathic scoliosis or paralytic scoliosis. Children should always be weighed at consultations. There may be striking morphologic changes from 1 year to the next. If weight evaluation becomes an integral part of each consultation, changes will become obvious and can be incorporated into the orthopedic specialist's deliberations. A simple trend in the increase in a boy's weight is 18–20 kg at 5 years of age, 30 kg at 10 years of age, and 60 kg at 17 years of age [26]. Note that weight doubles between 10 and 17 years of age. At 5 years of age, the child's weight reaches 32 % of the final normal weight, yet only 48 % of the final normal weight is achieved at 10 years of age. In a patient whose

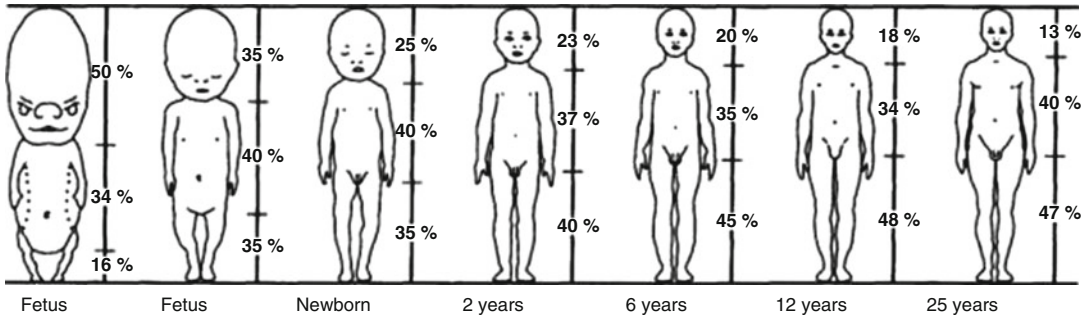


Fig. 4.4 Sitting height and lower limb length proportion. Sitting height is 65 % of standing height at birth and 52 %, at 12 years

Fig. 4.5 Growth velocity of sitting height (*red*) and lower limb (blue) in boys (1–18 years). Green arrow indicates beginning of growth spurt (*P*)

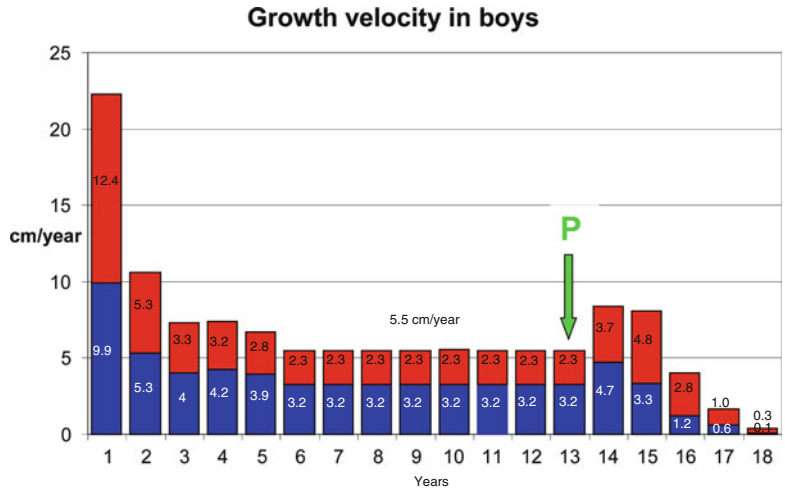
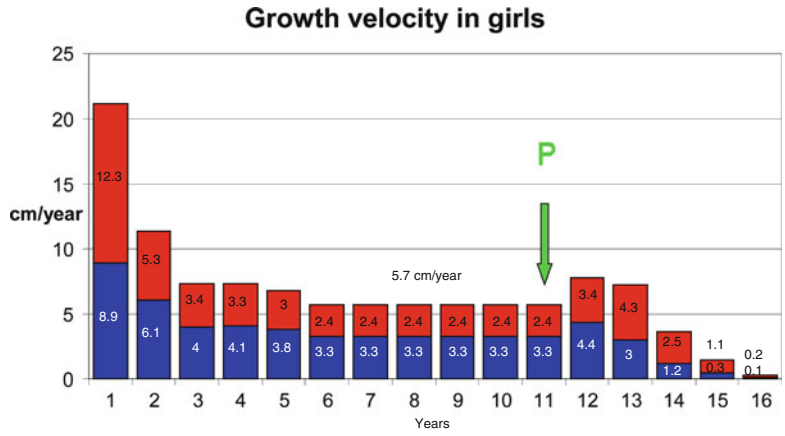


Fig. 4.6 Growth velocity of sitting height (*red*) and lower limb (*blue*) in girls (1–16 years). Green arrow indicates beginning of growth spurt (*P*)



weight is 10 % or more above normal, a scoliosis brace may no longer correct the spinal curve as it did before. A low weight, on the other hand, can explain the delay in the onset of menarche because girls generally need to attain a weight of 40 kg for menarche to occur. Hypotrophy is frequent in severe infantile scoliosis. A generally accepted estimate of body fat is expressed in the Quetelet body mass index: weight (kg)/height (m^2). Using this index, 20–25 kg/m^2 is normal,

25–30 kg/m^2 is moderate obesity, 30–40 kg/m^2 is major obesity, and more than 40 kg/m^2 is morbid obesity. Obesity is a major problem in Willi–Prader syndrome with scoliosis [20, 24, 26].

4.2.6 The Multiplying Coefficient

Lefort [42] outlined the concept of "multiplying coefficient," which can be applied to growth measurements in children at any age. This has also been extensively described by Paley et al. [49]. It is easy to calculate this coefficient, which is obtained by considering the percentage of growth that has been attained. For example, once a child has reached 40 % of his or her expected adult standing height, the multiplying coefficient can be calculated as 100/40 = 2.5. The multiplying coefficient can be applied to all biometric data – standing height, sitting height, subischial limb length, and length of the femur, tibia, humerus, radius, and ulna.

At birth, the sitting height of boys reaches 37 % of its final value. The multiplying factor is 2.85. At 10 years, sitting height of boys reaches 77 % of its final value; the multiplying factor is 1.28.

4.3 Chronology

4.3.1 Intrauterine Development

The most significant growth occurs during intrauterine life.

Growth starts before birth. During the first trimester of gestation, the systems are busy organizing themselves and develop at a brisk pace [34, 46]. During this period, the fetus makes daily progress, so that when the infant is born, it reaches a weight 6 million times that of the original egg. By the second month of life, the sitting height increases at a rate of 1 mm daily, which subsequently increases to 1.5 mm/day. Were this rate of growth to continue until the age of 10 years, the child would ultimately stand 6 m tall [6, 20, 24, 26, 34, 62].

From the third month onward, the embryo becomes a fetus and turns into a miniature adult. At the end of the second trimester of gestation, the fetus reaches 70 % of its expected length at birth (it measures 30 cm at this stage) but achieves no more than 20 % of the expected birth weight (it weighs approximately 800 g). During the third trimester, the fetus gains weight at the highest rate (700 g/month). This means that various stages of growth do not occur simultaneously during intrauterine life. Length increases steadily and rapidly during the first 6 months in utero, whereas weight gain is most rapid during the final 3 months of gestation.

With high-resolution ultrasonography, it is possible to follow the growth of the fetus and to detect even the slightest abnormality. It can be anticipated that many orthopedic spine conditions characterized by abnormal growth will be diagnosed prenatally.

4.3.2 From Birth to 5 Years

About 50 % of trunk growth occurs during the first 5 years of life. It is a critical period for early-onset spinal deformities.

Birth marks a very obvious transition in the growth of the child. After birth, not only does the overall rate of growth vary at different ages, but also the rates at which various segments of the body grow differ. For example, during the first 5 years of life, sitting height and subischial leg length increase at about the same rate; from 5 years of age to puberty, the sitting height accounts for one-third of the gain and the subischial limb length accounts for two-thirds; from puberty to maturity, the ratio is reversed, with the sitting height accounting for two-thirds of the gain in height and the subischial limb length accounting for one-third. The extent of increase in sitting height and subischial leg length for boys and girls of various ages are shown [20, 22–24, 26].

At birth, the standing height of the neonate (50–54 cm) is 30 % of the final height. By 5 years of age, the standing height increases to 108 cm,

which is double the birth height and 62 % of the final height. The first year of life sees particularly vigorous growth rates, with the infant's height increasing by 22 cm. This means that the height gain during a single year is as great as it is during the entire surge of puberty. After the age of 1 year, the growth rate starts to slow down but remains strong, with the infant growing another 11 cm between 1 and 2 years of age and 7 cm between 3 and 4 years of age.

At birth, the sitting height of the neonate is approximately 34 cm, which is roughly two-thirds of the standing height and 37 % of the final sitting height. The sitting height gains about 12 cm from birth to age 1 year; 5.3 cm from 1 to 2 years; 3.3 cm from 2 to 3 years; 3.2 cm from 3 to 4 years; and 2.8 cm from 4 to 5 (average) years. In 5 years, the trunk gains about 28 cm for girls and 29 cm for boys, much more than during the puberty spurt (11.5 cm for girls, 13 cm for boys). During the first 5 years of growth after birth, the proportions change. The cephalic end of the body becomes relatively smaller, whereas the subischial leg length increases. During this period, growth is not only a vertical phenomenon but also a volumetric one.

At birth, weight is between 3000 and 3500 g, which is 5 % of the final figure. At 5 years of age, the weight averages 18–20 kg, which is 32 % of *the* final adult weight. In 5 years, the weight gain is 15–17 kg. Birth weight triples in a single year and quadruples by the age of 3 years. The circumference of the chest is 32 cm at birth but increases by 25 cm to reach 57 cm by the age of 5 years [2, 3, 8, 45]. Chest morphology has undergone dramatic changes (see Figs. 4.5 and 4.6).

4.3.3 From 5 Years to Beginning of Puberty: A Steady Period

Before puberty, trunk growth slows down.

Between 5 years of age and the onset of puberty, 11 years of bone age in girls and 13 years of bone age in boys, there is a marked deceleration in growth, with standing height increasing at approximately 5.5 cm/year. About two-thirds of this growth (3.2 cm) occurs in the lower limb, and about one-third (2.3 cm) occurs in the sitting height. The trunk is now growing at a slower rate, whereas the lower limbs are growing faster than the trunk, thereby altering the proportions of the body. During this period, in boys, standing height will increase by 27 % (approximately 44 cm); sitting height, by 20 % (approximately 18 cm); and subischial limb length, by 32 % (approximately 26 cm); in girls, standing height will increase by 22 % (approximately 34 cm); sitting height, by 17 % (approximately 14 cm); and subischial limb length, by 28 % (approximately 20 cm) [20, 22–24, 26]. By 5 years of age, the sitting height increases to 60 cm, approximately 66 % of the final sitting height, with only another 26–30 cm to grow. This information is useful in anticipating the effects of deformity and the consequences of arthrodesis in spinal deformity in young patients.

From 5 years of age to the beginning of puberty, the average weight gain is about 2.5 kg/year [26]. At 10 years of age, the weight represents about 50 % of the final weight. In contrast, the standing height at this age is 78 % of the final standing height in the case of boys and 83 % in the case of girls [26].

4.3.4 Puberty: A Turning Point

Puberty is a very challenging period for children with early-onset scoliosis.

At the beginning of puberty, approximately 22.5 cm of growth remains to be attained in standing height (12.5 cm in sitting height and 10 cm in lower limb) in the case of boys and 20.5 cm (11.5 cm in sitting height and 9 cm in lower limb) in the case of girls (Figs. 4.7, 4.8, 4.9, and 4.10).

Chronologic age is a poor indicator of puberty. We may start anticipating puberty at 10 years of age in girls and 12 years in boys. The acceleration in growth velocity best characterizes the beginning of puberty. From a clinical viewpoint, puberty will

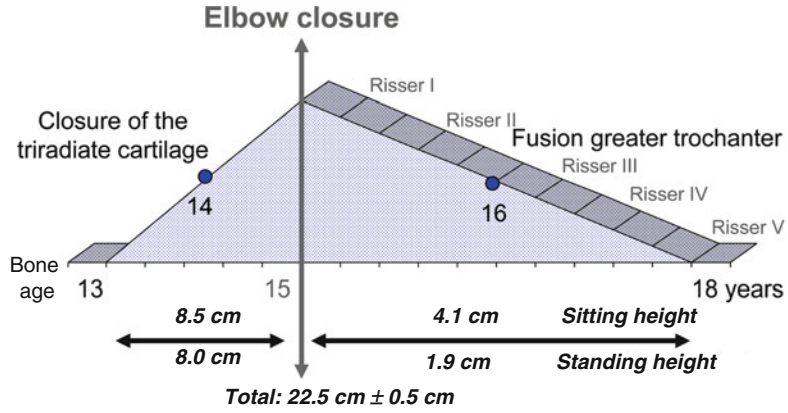


Fig. 4.7 Pubertal diagram in boys

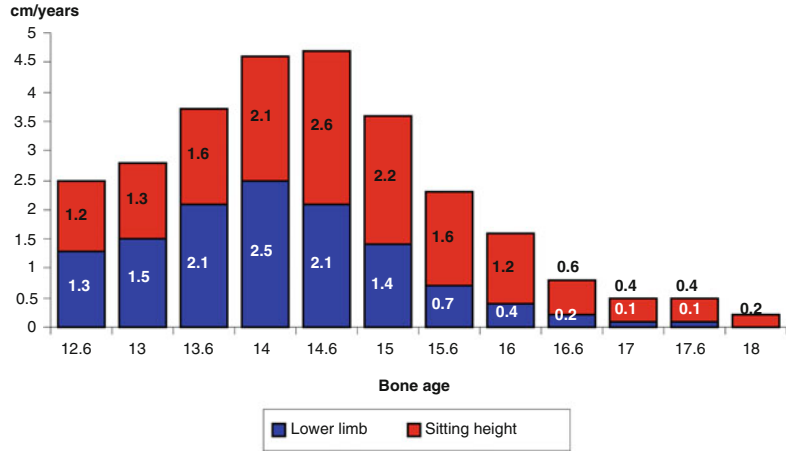


Fig. 4.8 Growth velocity for bone age (boys). The peak velocity of growth during puberty occurs between 13 and 15 years of bone age in boys

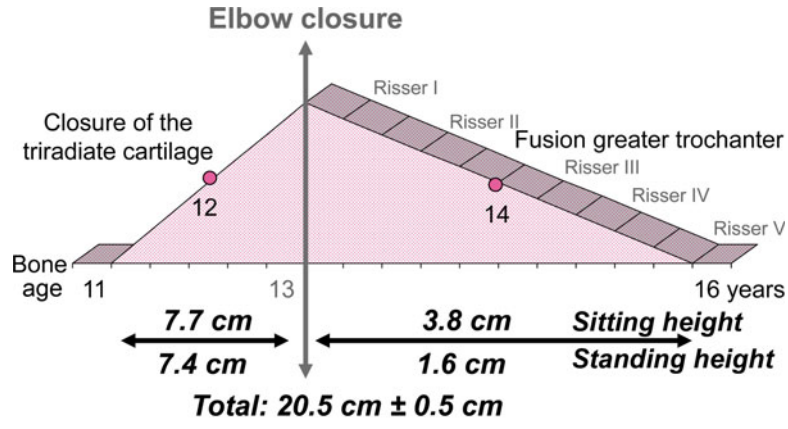


Fig. 4.9 Pubertal diagram for girls

be recognized by a combination of factors other than growth: sexual development, chronologic age, and bone age. After 11 years of age, the growth patterns of boys and girls proceed differently. On an average, girls will experience the onset of puberty at 11 years (bone age) and boys, at 13 years (bone age). Puberty, and its accompanying rapid growth, is a period of great importance to the orthopedic surgeon. It is therefore crucial to recognize the period just before puberty.

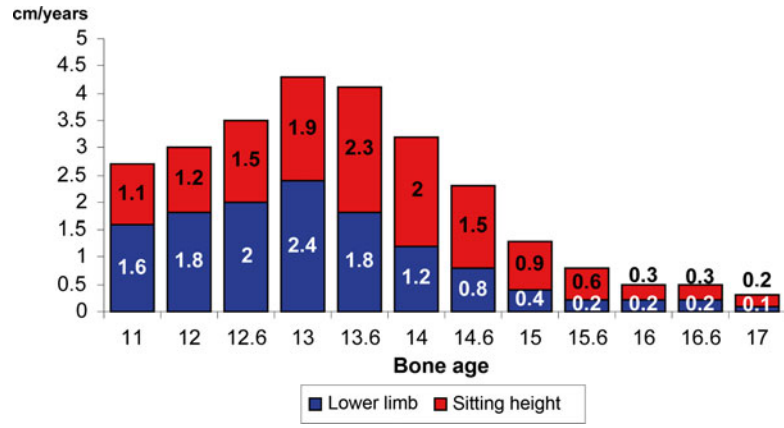


Fig. 4.10 Growth velocity for bone age (girls). Menarche usually occurs on the descending side, after elbow closure at 13 years 6 months bone age, around Risser I

There are four main characteristics that dominate the phase of growth called puberty:

1. Dramatic increase in stature
2. Change in the proportions of the upper and lower body segments
3. Change in overall morphology: biachromial diameter, pelvic diameter, fat distribution, and so on [32]
4. Development of secondary sexual characteristics

During puberty from 11 to 15 years of age in girls and from 13 to 17 years in boys, there is a dramatic increase in the growth rate. However, during this period, the growth is far more noticeable in the trunk than in the lower limbs: two-thirds of the growth goes toward increasing sitting height, and only one-third is toward increasing subischial limb length. It is during this period that boys overtake girls in height. On an average, boys are between 12 and 15 cm taller than girls. This is accounted for by two factors. First, boys have approximately 2 years of growth more than girls. Second, boys have a slightly greater increase in the rate of growth during puberty than do girls, accounting for approximately 2 cm of additional height [20, 24, 26].

During puberty, the standing height increases by approximately 1 cm/month. At the onset of puberty, boys have 14 % (±1 %) of their remaining standing height to grow; this is approximately 22.5 cm (±1 cm), made up of 12.5 cm in sitting height and 10 cm in subischial limb length. Girls have 13 % (±1 %) of their standing height to grow. This is approximately 20.5 cm (±1 cm), made up of 11.5 cm in sitting height and 9 cm in subischial length.

The peak velocity of growth during puberty occurs between 13 and 15 years of bone age in boys and between 11 and 13 years of bone age in girls. After bone age of 13 years in girls and 15 years in boys, there is a considerable decrease in the annual velocity of height gain. The lower limbs stop growing rapidly; the total remaining growth is 5.5 cm, about 4 cm in the sitting height and about 1.5 cm in the lower limb. This variation in growth velocity is an extremely important factor to consider in the treatment of many disorders, especially scoliosis and limb-length discrepancy.

These figures, ratios, and rates provide only a partial view of the growth phenomenon. Precise evaluation of the characteristics of puberty, using the bone age assessment, the Tanner classification, the onset of menstruation, the Risser sign, and the annual height velocity, is something that needs to be undertaken with a great deal of care and consideration. One of the major problems with using only the onset of menarche and the Risser sign is that they occur after the growth associated with puberty has begun to slow.

4.3.5 Secondary Sexual Characteristics

Observe the child and consider biological age.

Secondary sexual characteristics develop throughout the course of puberty; the first appear-

ance of pubic hair, the budding of the nipples, and the swelling of the testes are the first physical signs to signal the onset of puberty. The first physical sign of puberty in boys, testicular growth in 77 %, occurs, on average, 1.7 years before the peak height velocity and 3.5 years before attaining adult height [53]. The bone age will be approximately 13 years at the onset of puberty; the Risser sign is 0, and the triradiate cartilage is open.

The first physical sign of puberty in girls, breast budding in 93 %, occurs about 1 year before peak height velocity [26]. This averages 11 years in bone age. The Risser sign is still 0, and the triradiate cartilage is still open at the onset of puberty. Menarche occurs about 2 years after breast budding, and final height is usually achieved 2.5–3 years after menarche. After menarche, girls will gain the final 5 % of their standing height, about 3–5 cm [26]. The appearance of axillary hair, although variable, occurs after the peak of the pubertal growth diagram.

The secondary sexual characteristics generally develop in harmony with bone age, but there are discrepancies in 10 % of cases. Puberty may be accelerated and growth can end more quickly than usual, catching the unwary physician off guard. In fact, it has been demonstrated that it is not uncommon to see an acceleration of the bone age during puberty.

4.3.6 Pubertal Diagram and Peak Height Velocity

Peak height velocity is not a single point on a chart.

Using all of these landmarks, it is possible to draw a diagram relating the events occurring during puberty. Even if one indicator is missing or does not match the other, it is still possible to have a good idea of where the child is, on his or her own path through puberty. By plotting the gains in standing height and sitting height every 6 months, a picture of the period of puberty is developed (Figs. 4.7, 4.8, 4.9 and 4.10). It is also easy to divide this into two parts. The first phase (i.e., the ascending phase of the growth velocity curve) is characterized by an increase in the velocity of growth and is the major portion of the pubertal growth spurt. The second phase (i.e., the descending phase of the growth velocity curve) is characterized by a slowing of the rate of growth [57].

The first phase of the pubertal growth spurt is the ascending phase, which corresponds to the acceleration in the velocity of growth. This phase lasts 2 years, from approximately 11 to 13 years of bone age in girls and from 13 to 15 years of bone age in boys. The gain in standing height in girls during this phase is about 15.1 cm, made up of 7.7 cm in sitting height and 7.4 cm in subischial limb length. The gain in standing height in boys during this phase is about 16.5 cm, made up of 8.5 cm in sitting height and 8 cm in subischial limb length. During this first phase of pubertal growth spurt, the increase in sitting height contributes 53 % and the increase in subischial length contributes 47 %. Therefore, more growth comes from the trunk than from the legs during this phase growth.

The peak height velocity occurs on the ascending side of the growth velocity curve. It does not occur at just one point on the curve but takes place during a period of 2 years [26, 54]. It can be roughly identified by accurate assessment of standing height and sitting height at 6-month interval.

Triradiate cartilage closure occurs about halfway up the ascending phase of the pubertal growth velocity diagram. This closure corresponds to an approximate bone age of 12 years in girls and 14 years in boys. After closure of the triradiate cartilage, there is still a considerable amount of growth remaining: greater than 12 cm of standing height in girls and more than 14 cm in boys. Sanders et al. [54] have shown that the crankshaft phenomenon decreases substantially after closure of the tri-radiate cartilage.

The second phase of the pubertal growth spurt is the descending side, which corresponds to the deceleration of the velocity of growth. The closure of the elbow (discussed in subsequent text) divides the ascending and descending phases of puberty. The descending phase lasts 3 years from 13 to 16 years of bone age in girls and from 15 to

18 years of bone age in boys. During this phase, both boys and girls will gain about 6 cm in standing height, with 4.5 cm attained from an increase in sitting height and 1.5 cm attained from an increase in subischial limb length. During this phase, the increase in sitting height contributes 80 % of the gain in the standing height [20, 22, 24, 26].

Menarche usually occurs after closure of the olecranon apophysis, on the descending phase of the growth curve when the rate of growth is slowing. This decrease in rate of growth is usually between bone ages of 13 and 13 years 6 months and corresponds to Risser sign I on the iliac apophysis. After this stage, the average girl will gain an additional 4 cm of sitting height and 0.6 cm of subischial limb length. Menarche is not as precise as many other indicators during puberty. Forty-two percent of girls experience menarche before Risser I; 31 %, at Risser II; 8 %, at Risser III; and 5 %, at Risser IV [20, 22, 24, 26]; after 2 years of menarche, there is no more growth.

The descending phase of puberty is characterized by a significant growth of the thorax (Fig. 4.11).

During puberty, the peak growth is a combination of three micropeaks: the first peak involves the lower limb at the very beginning of this period, and the second peak involves the trunk (these two peaks are on the ascending phase of the growth velocity curve), the third peak involves growth of the thorax and occurs during the descending phase of the curve. At skeletal maturity, the final standing height is about 175 cm (±6.6 cm) for boys and about 166 cm (±6 cm) for girls.

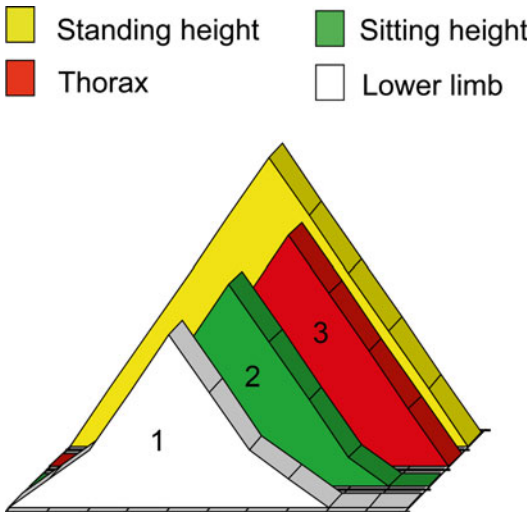


Fig. 4.11 During puberty, the peak growth is a combination of three micropeaks: *1* lower limb growth peak; *2* trunk growth peak; and *3* thorax growth peak

Puberty is characterized by a great increase in weight. At the beginning of puberty, the average weight is 40 kg for boys and 33 kg for girls. At skeletal maturity, the average weight of boys is 65 kg (a gain of 25 kg), and the average weight of girls is 56 kg (a gain of 23 kg). During the growth spurt of puberty, the average gain in weight each year is 5 kg [20, 24, 26].

4.4 Estimation of Skeletal Maturity

Bone age does not always match with chronological age. It is a controversial although necessary parameter.

In pediatric orthopedics, chronologic age is of no significance. Everything depends on bone age. Personal data indicate that about 50 % of children have a bone age that is significantly different from their chronologic age. Delayed bone age is characteristic of severe cerebral palsy (total body) involvement. All, reasoning, analyses, forecasting, and decision making, should be based on bone age [2, 14, 19, 20, 22, 24, 25, 55, 56, 58].

Accurate assessment of bone age is not easy. The younger the child, especially before puberty, the more difficult it is to determine future growth, and the more likely it is for errors to be made. In addition, children are often bone age mosaics. Bone age determinations carried out in the hands, elbows, pelvis, and knees will not always agree with one another.

Often, the bone age determination is made too quickly and with too little information. The standard deviations for determining bone age must be understood, as well as the nuances of what to

look for in the interpretation of the radiograph. When using a particular method (e.g., the Greulich and Pyle Atlas [34]), it is important to read the entire book to understand what to look for and to know the standard error, rather than simply comparing radiographs. If there is a major decision to be made, it is better to have two interpretations of the child's bone age and to enlist the support of pediatric radiologists with experience in bone age determination. Cundy et al. [19] demonstrated that four radiologists' interpretations of skeletal age differed by more than 2 years in 10 % of patients. Carpenter and Lester [14] evaluated bone age in children younger than 10 years. They showed that taking separate readings of the distal radius and ulna, the carpal bones, the metacarpals, and the phalanges could magnify these errors and that the ages of the carpal bones and the distal radius and ulna often lag behind the ages of the metacarpals and phalanges. This means that excessive haste in reading the bone age can result in fatal strategic errors.

There are three basic approaches to the radiographic assessment of skeletal maturity: atlas, sum of scores, and statistical combination of scores. Knowledge of these methods and their limitations is important for the orthopedist, especially in difficult cases. The Greulich and Pyle Atlas [34] is the most familiar and commonly applied approach and involves qualitatively matching the subject's hand and wrist radiographs against a series of gender-specific standards. This atlas is based on a collection of radiographs of children born between 1917 and 1942. In comparing this atlas with its French counterpart, Sempé and Pavia Atlas [56], we learned that there is no major difference between these two atlases. One of the shortcomings in using the Greulich and Pyle Atlas is that there are few changes in the hand during the critical time of puberty (ascending side of pubertal growth velocity diagram) [20, 26, 28].

For this reason, the author has found the method by Sauvegrain et al. [25, 55] to be of enormous value in assessing children during puberty. This method is the scoring system that evaluates anteroposterior and lateral views of the elbow and assigns a value to the epiphyses. This value is then plotted on a chart to give the bone age. Four ossification centers are taken into consideration: condyle and epicondyle, trochlea, olecranon, and radial epiphysis. This method is reliable and is based on the skeletal maturation of the elbow, which occurs during a 2-year period corresponding to the ascending phase of the growth velocity curve. Therefore, it is extremely helpful in boys aged 13–15 years and in girls aged 11–13 years, a period in which many of the clinical decisions involving future growth are made (spinal arthrodesis). In addition, it shows good correlation with the Greulich and Pyle Atlas but is much easier to use.

At the beginning of puberty, growth centers of the elbow are wide open, but 2 years later, when the peak velocity of the pubertal growth spurt is reached and growth begins to slowdown, they are all completely closed. This complete closure occurs 6 months before Risser I. In the method by Sauvegrain et al., the olecranon is the bone that shows the most characteristic and clear-cut sequences during the first 2 years of puberty [17, 25]. For this reason, the author has described the olecranon method, at the beginning of puberty (at bone age of 11 years in girls and 13 years in boys), two ossification centers appear (Fig. 4.12). Six months later (at bone age of 11.5 years in girls and 13.5 years in boys), they merge to form a half-moon shape. At bone age of 12 years in girls and 14 years in boys, the olecranon apophysis has a rectangular appearance. Six months later (at bone age of 12.5 years in girls and 14.5 years in boys), the olecranon apophysis begins to fuse with the ulna, a process that takes another 6 months, being completed by the bone age of 13 years in girls and 15 years in boys. In our clinical practice experience, the olecranon alone can give rapid and valuable information about bone age. The olecranon method is more accurate in itself because it allows differentiation of bone age in semesters, which is not true for the Greulich and Pyle Atlas for the considered time of puberty [33].

Other methods are described as follows: the Tanner et al. [58] system scores 20 indicators on hand and wrist radiograph, yielding total scores ranging from 0 to 100. The Fels method is a sophisticated approach, scoring hand and wrist

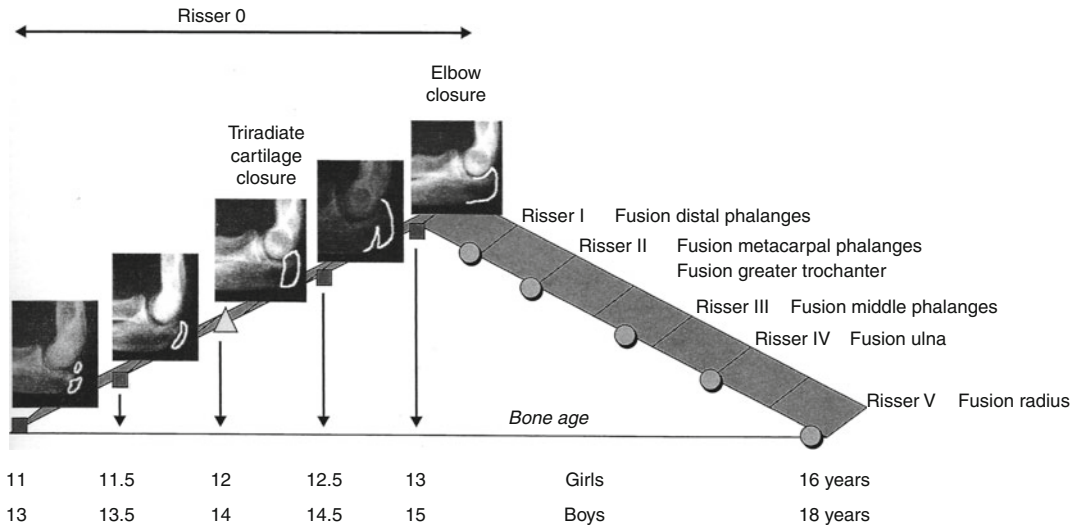


Fig. 4.12 Examination of skeletal maturity (bone age) by Olecranon method (Described by Dimeglio)

radiographs and using a computer program. Both of these methods are time consuming and not useful in daily practice.

The Oxford scoring method for assessing skeletal maturity from pelvic radiographs is based on nine indicators, three of which are useful during puberty: the triradiate cartilage, the greater trochanter, and the Risser sign [2]. The triradiate cartilage closure occurs on the ascending side of the pubertal growth diagram at bone age of 12 years in girls and 14 years in boys. After its closure, a significant amount of growth in standing height still remains: 13 cm in girls and 14 cm in boys. The greater trochanter closure occurs on the descending phase of the pubertal growth diagram, at bone age of 14 years in girls and 16 years in boys (i.e., between Risser II and Risser III).

4.5 The Concept of Risser Sign Is Misleading

Risser 0 covers two-thirds of pubertal growth spurt.

The Risser sign is one of the most commonly used markers of skeletal maturation, especially in the treatment of scoliosis. The sign appears on the radiograph of the pelvis, which is often studied during the assessment of this disorder, thereby obviating the need for an additional radiograph. The duration of excursion of the Risser sign is also variable and may range from 1 to 3 years [4]. However, the value of this sign in accurate decision making has been questioned. Little and Sussman [43] concluded that, all things considered, it is better to rely on chronologic age. Although the author does not agree with their conclusions, when important decisions are made, the Risser sign should be supplemented with the bone age, as determined by the method of Greulich and Pyle [34] (Figs. 4.13 and 4.14).

Risser 0 covers the first two-thirds of the pubertal growth, which corresponds to the ascending limb of the pubertal growth diagram. However, this period of Risser 0 is important in decision making in many conditions; therefore, it is important to have more precise markers of stage of puberty (growth) during this period, such annual growth velocity, elbow maturation (olecranon), and changes in morphology of the triradiate cartilage [16]. Risser 0 gives little information, other than indicating that the peak of the growth velocity curve has not been reached [16]. The author has recommended dividing this period of the ascending phase of the pubertal growth diagram, characterized by

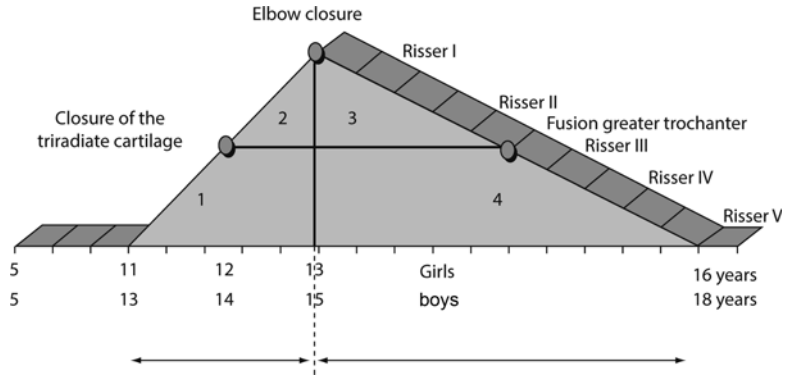


Fig. 4.13 Pubertal diagram and Risser stages. An anteroposterior (*AP*) radiograph of the pelvis provides useful information regarding the pubertal stage of the patient

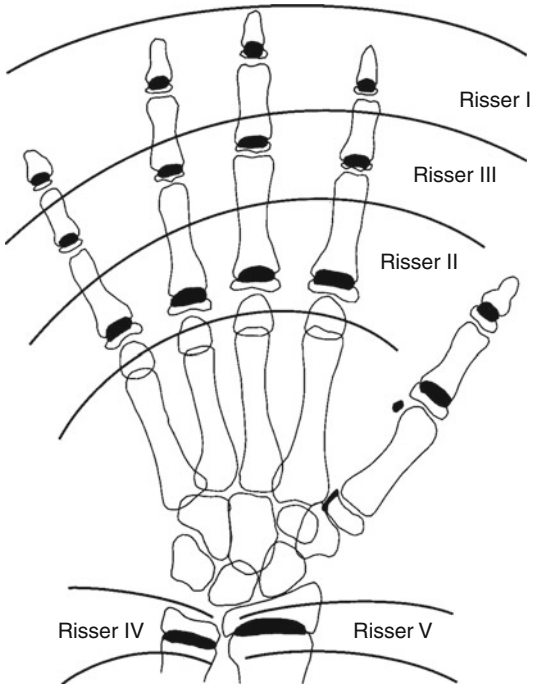


Fig. 4.14 Epiphyseal closure and Risser stages in hand

Risser 0, into three periods, based on the triradiate cartilage and the closure of the olecranon apophysis: triradiate cartilage open, triradiate cartilage closed but olecranon open, and olecranon closed [17, 20, 22, 24, 26].

Risser 1 heralds the beginning of the descending slope of the pubertal growth velocity diagram. It generally appears after the elbow closure, when the epiphyses of the distal phalanges (II, III, IV, and V) of the hand fuse. The rate of growth in sitting height and standing height decreases abruptly. Axillary hair generally appears during this period [17, 20, 22, 24, 26].

Risser 2 corresponds to a bone age of 14 years in girls and 16 years in boys. It generally appears when the greater trochanteric apophysis unites with the femur. When the proximal phalangeal epiphyses fuse in the hand, there is approximately 3 cm left to grow in sitting height and no more growth of the lower limb.

Risser 3 corresponds to bone ages of 14.5 years in girls and 16.5 years in boys. The phalangeal epiphyses of P1 and P2 fuse during this period; the greater trochanter is closed, and 1 year of growth and an increase of 2 cm in sitting height still remain.

Risser 4 corresponds to a bone age of 15 years in girls and 17 years in boys. The distal epiphysis of the ulna is united to the shaft. At this stage, the remaining growth in sitting height is 1 cm.

Risser 5 is very much like Risser 0: it is a long period that does not provide much information to the clinician. The distal radial epiphysis generally fuses around Risser 5. The iliac apophysis may fuse at age 22 or 23 years, but in some cases, it never fuses [7].

Regardless of the method of its determination, bone age is meaningless as an isolated parameter. It should be constantly measured against chronologic age, the rate of annual growth in standing height, and secondary sexual characteristics [53].

Before 10 years of age, evaluation of bone age is difficult; the appearance of the ossification center on the hand or on the elbow can give useful information, but the most important is to look at the growth curves of the standing and sitting

height, and mainly the weight, to evaluate the biological age.

On the ascending side of puberty, olecranon evaluation is more precise than the hand; on the descending side of puberty, ossification centers of the hand must be balanced with the Risser sign [25].

4.6 Growth of the Trunk

4.6.1 Growth in the Spinal Column

The growing spine is a mosaic of growth plates.

Measurement of sitting height provides an indirect indication of spinal growth. The spine makes up 60 % of the sitting height, whereas the head represents 20 %, and the pelvis represents 20 % [22]. If we accept the fact that there are at least four growth zones per vertebra, the resulting morphology of the spinal column is the product of 130 physes. The pattern of growth in the posterior arch, where closure is linked, in particular, to the presence of the neural stem, differs from that seen in the body of the vertebra, which behaves like a long bone [21, 22].

If one were to compare any of the vertebrae of a newborn, one would find very little morphologic variation between them. The process by which cervical, thoracic, and lumbar vertebrae acquire their individual identities is gradual. In the vertebral body, ossification first appears in the dorsal region; from this hub, the process of ossification radiates to the cranial and caudal parts of the spine. The process of ossification is extremely slow and does not finish until the 25th year of life.

At birth, the lumbosacral vertebrae are relative smaller than the thoracic and cervical vertebrae. However, during the first years of growth, they grow more rapidly. Between 3 and 15 years of age, the lumbar vertebrae and their discs increase in size by about 2 mm/year, whereas the thoracic vertebrae and their discs increase by 1 mm.

The discs account for approximately 30 % of the height of the spinal segment at birth. At maturity, this proportion decreases to 25 %, with the discs constituting 22 % of the cervical spine, 18 % of the thoracic spine, and 35 % of the lumbar spine.

The anterior and posterior portions of the vertebrae do not grow at the same rate. In the thoracic region, the posterior components grow at a faster pace than their anterior counterparts. The reverse occurs in the lumbar region. Growth potential therefore varies from one level to the next, differing from anterior to posterior. In addition, as the vertebrae develop, there is a constant remodeling of the anatomic organization of the spine; for example, the articular apophyses change in both morphology and direction.

The neurocentral synchondroses are at the junction between the vertebral body and the posterior arch. There are two physes developing in two directions; the neurocentral physes contribute to 30 % of the ossification of the vertebral body and participate mostly in the posterior arch ossification. Zhang et al. [64] have studied in normal children the evolution of the neurocentral synchondroses by MRI axial images. The neurocentral growth plate is opened in all patients less than 3 years. It closes from the lumbar and proximal thoracic spine to the middle and distal thoracic spine. At 4 years, the neurocentral synchondroses had 50–74 % closure in the lumbar region. At 5 years, the proximal thoracic (T1–T6) had 25 % less closure with the middle T7–T9 and distal (T10–T12) thoracic demonstrated no closure. At 9 years, the neurocentral synchondroses of the spine are closed. This excellent study provided significant information for growing modulation treatment of early-onset deformities.

The length of the spine will nearly triple between birth and adulthood. At birth, the vertebral column is approximately 24 cm long. In the newborn, only 30 % of the spine is ossified. There is little substantial difference in morphology between one vertebra and another. The length of a thoracic vertebra is about 7.6 mm, and that of a lumbar vertebra is about 8 mm [22]. The average adult spine is approximately 70 cm long in men, with the cervical spine measuring 12 cm; the thoracic spine, 28 cm; the lumbar spine, 18 cm; and the sacrum, 12 cm. The average female spine is approximately 63 cm long at maturity [21, 22].

4.6.2 Cervical Spine

At birth, the cervical spine measures 3.7 cm; it will grow by about 9 cm, to reach the adult length of 12–13 cm. The length of the cervical spine will nearly double by 6 years of age. It will gain an additional 3.5 cm during the pubertal growth spurt. The cervical spine represents 22 % of the C1 S1 segment and 15–16 % of sitting height.

The diameter of the cervical spinal canal varies with location, typically decreasing in width from Cl to C7 or from C1 to C3 and then widening slightly. These differences are important in the clinical setting because the room available for the spinal cord can be very consequential. It should be remembered that, regardless of the size of the child (e.g., in dwarfing conditions), the spinal cord will attain the usual adult diameter. The average width of the cervical cord is 13.2 mm, and the average anteroposterior depth is 7.7 mm [22]. Therefore, the transverse and sagittal diameters of the cervical canal are important. In an adult, at C3, the normal transverse diameter is 27 mm and the average sagittal diameter is approximately 19 mm. The cervical canal is wide enough to permit the entry of the thumb of an adult finger.

4.6.3 T1–S1 Segment

It is a strategic segment.

The Tl–S1 segment is very important because the most frequent disorders of the spine during growth originate in this segment (Figs. 4.15 and 4.16). The Tl–S1 segment measures about 19 cm at birth,

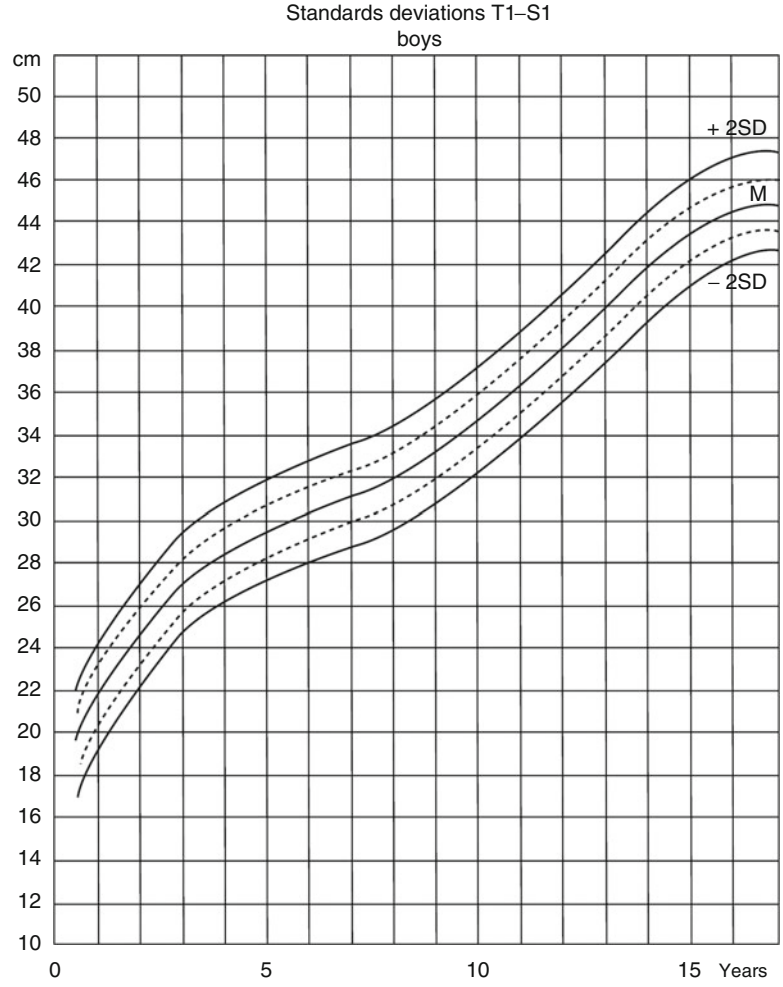


Fig. 4.15 T1–S1 segment length-for-age (birth to skeletal maturity: boys)

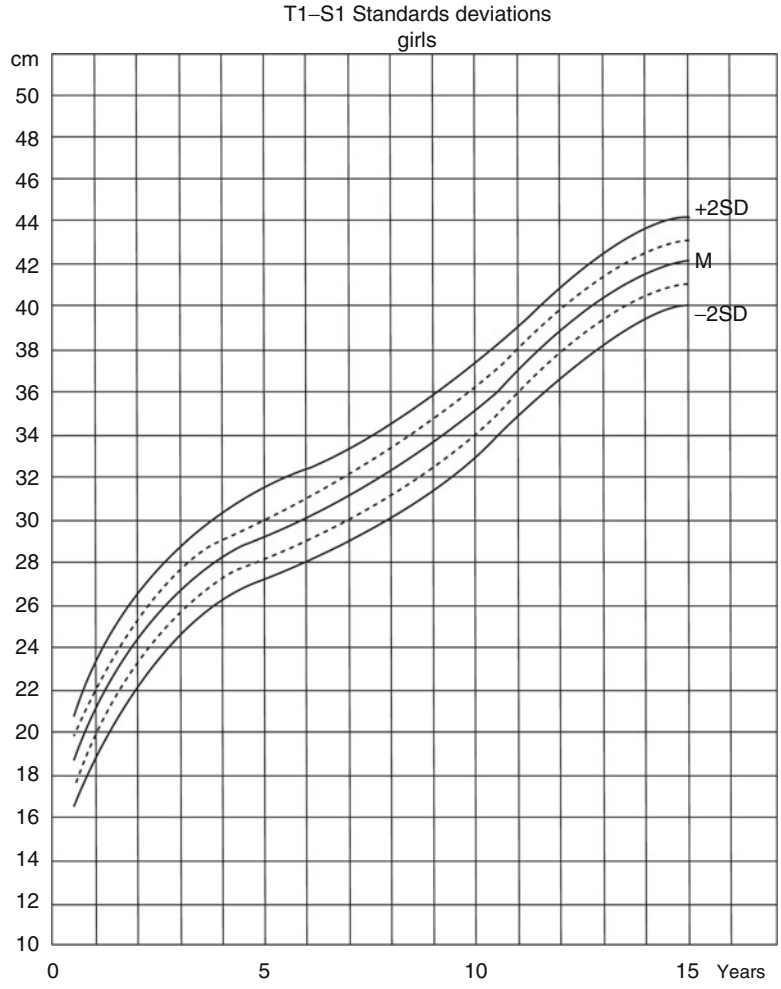


Fig. 4.16 T1–S1 segment length-for-age (birth to skeletal maturity: girls)

45 cm at the end of growth in the average man and 42–43 cm in the average woman. From birth to 5 years of age, the gain is about 10 cm; from 5 to 10 years of age, about 5 cm; and from 10 years of age to skeletal maturity, about 10 cm. This segment makes up 49 % of the sitting height at maturity. Knowledge of the effects of arthrodesis on this segment of the spine requires precise knowledge of the growth remaining at various ages (Figs. 4.17 and 4.18).

4.6.4 Thoracic Spine T1–T12

It is the posterior pillar of the thoracic cage.

The thoracic spine is about 11 cm long at birth and reaches a length of about 28 cm in boys and 26 cm in girls at the end of growth. Its length more than doubles between birth and the end of the growth period. The growth of the thoracic segment has a rapid phase from birth to 5 years of age (7 cm), a slower phase from 5 to 10 years of age (4 cm), and rapid growth through puberty (7 cm) [22–24, 26]. The T1–T12 segment represents 30 % of the sitting height, so a single thoracic vertebra and its disc represents 2.5 % of the sitting height (Figs. 4.19 and 4.20). By knowing the amount of growth that each vertebra contributes to the final height, the effect of a circumferential arthrodesis, which stops all growth in the vertebrae and discs, can be calculated [21, 22, 24, 26].

Posterior arthrodesis results in only one-third of this deficit (2.5 % of sitting height for each thoracic vertebra), which is about 0.8 % of the remaining sitting height.

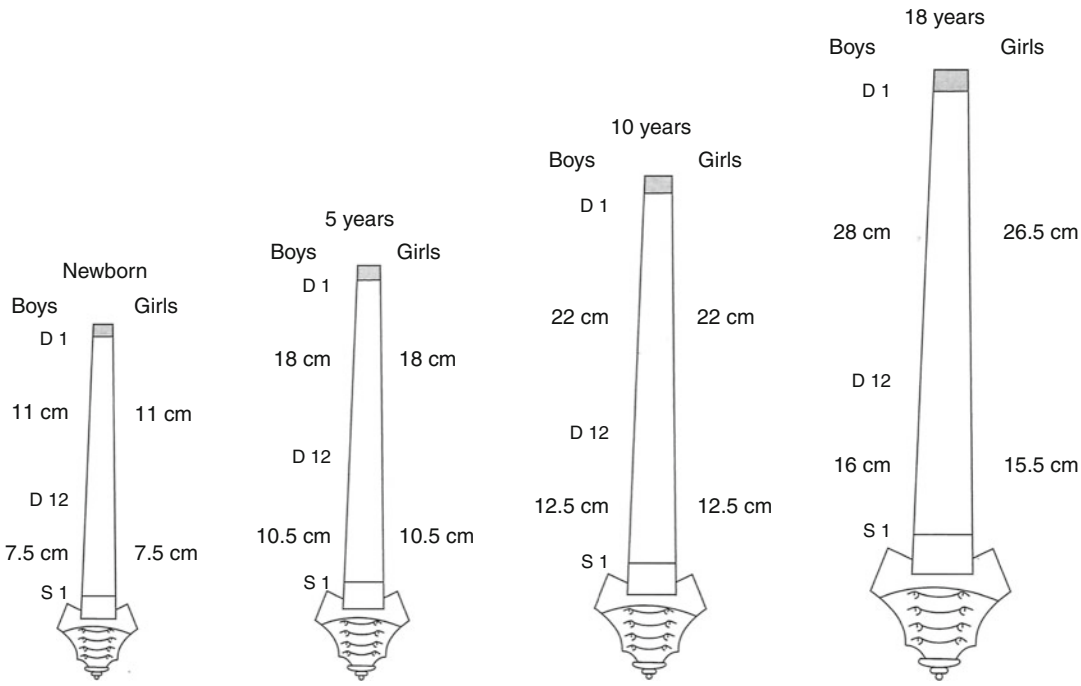


Fig. 4.17 Evaluation of T1–S1; thoracic segment T1–T12, and lumbar segment L1–L5 at birth, 5, 10, and 18 years (the figures are average values)

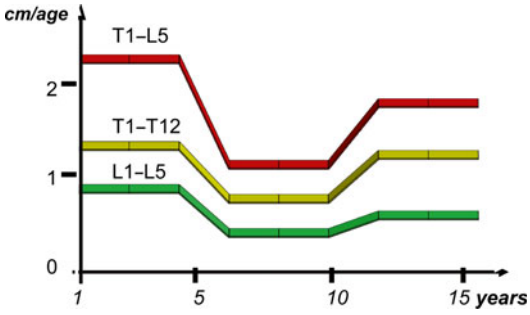


Fig. 4.18 Growth velocity of T1–L5, thoracic segment T1–L5, thoracic segment L1–L5; T1.L5 : from birth to 5 years : 2,2 cm per year; from 5 to 10 years : 1,2 cm per year; from 10 to skeletal maturity : 1,7 cm per year. T1 T12 represents 2/3 of T1 L5; L1 L5 represents 1/3

The thoracic spinal canal is narrower than either the lumbar or the cervical canals. At the age of 5 years, this canal attains its maximum volume and is wide enough to permit the entry of the little finger of an adult hand. The average of the transverse and anteroposterior diameters at T7 is approximately 15 mm.

4.6.5 Lumbar Spine (L1–L5)

The lumbar vertebra grows more than the thoracic vertebra.

The L1–L5 lumbar spine is approximately 7 cm in length at birth, and it grows to approximately 16 cm in men and 15.5 cm in women. As in the thoracic spine, growth is not linear; there is rapid growth from 0 to 5 years of age (gain of about 3 cm), slow growth from 5 to 10 years (gain of about 2 cm), and rapid growth again from 10 to 18 years of age (gain of about 3 cm). The height of the lumbar spine doubles between birth and maturity (Figs. 4.21 and 4.22).

The lumbar spine represents 18 % of the sitting height, and a single lumbar vertebra and its disc account for 3.5 of the sitting height. Values for the remaining growth of the lumbar segment at various ages are given in figures. A posterior vertebral arthrodesis results in a deficit of only one-third of this value, that is, slightly more than 1 % of the remaining sitting height.

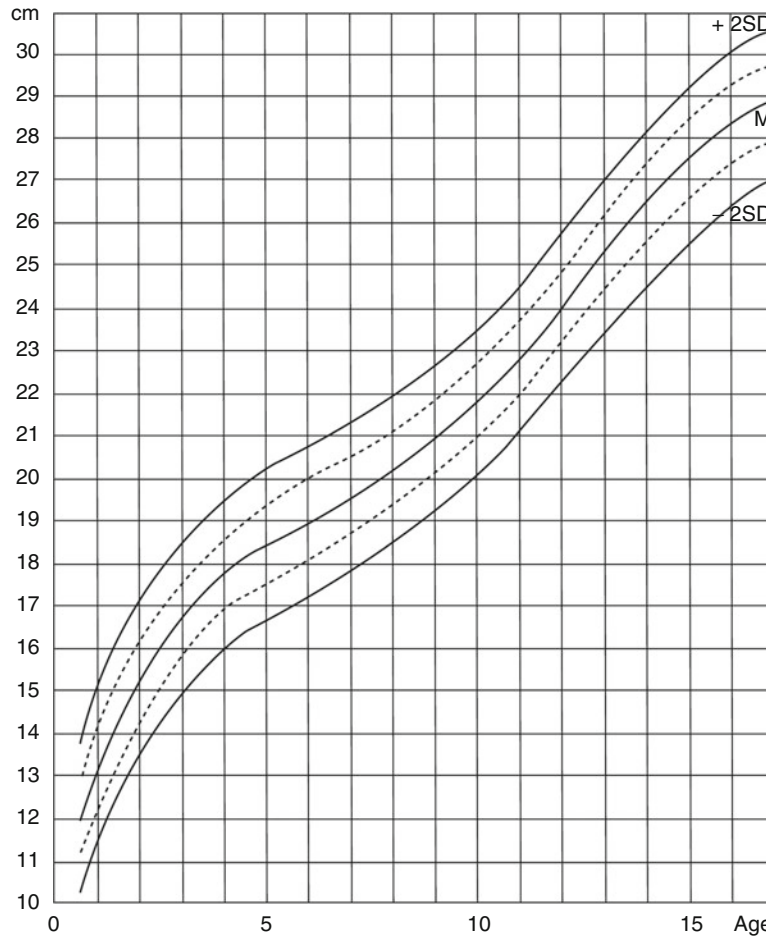


Fig. 4.19 T1–T12 segment length-for-age (birth to skeletal maturity: boys)

At the skeletal age of 10 years, the lumbar spine reaches 90 % of its final height but only 60 % of its final volume. The medullar canal in the lumbar spine is wider than that in the thoracic spine. At skeletal maturity, the dimensions of the canals are such that the adult thumb can be introduced into the cervical canal, the forefinger into the thoracic canal, and the thumb into the lumbar canal. At birth, the spinal cord ends at L3, and at maturity, it ends between L1 and L2.

4.6.6 The Thorax is the Fourth Dimension of the Spine

Spine and thoracic growths are interrelated.

The thoracic circumference is a rough but valuable indicator of this fourth dimension of spinal growth. The thorax has a circumference of 32 cm at birth, and it will grow to 56 cm in boys and 53 cm in girls, that is, to almost three times of its birth size [20].

In boys, the thoracic circumference is 36 % of its final size at birth, 63 % at 5 years of age 73 % at 10 years, 91 % at 15 years, and 100 % at 18 years. From birth to 5 years of age, the thoracic circumference grows exponentially and increases by 24 cm. From the ages of 5 to 10 years, the increase is slower; the thoracic circumference is 66 cm at 10 years of age, which means that its growth is only 10 cm in 5 years. At that stage, it is at 73 % of its final dimension. Another spurt occurs between the ages of 10 and 18 years, particularly during puberty. The thoracic circumference then increases by 23 cm, that is, as much as between birth and 5 years.

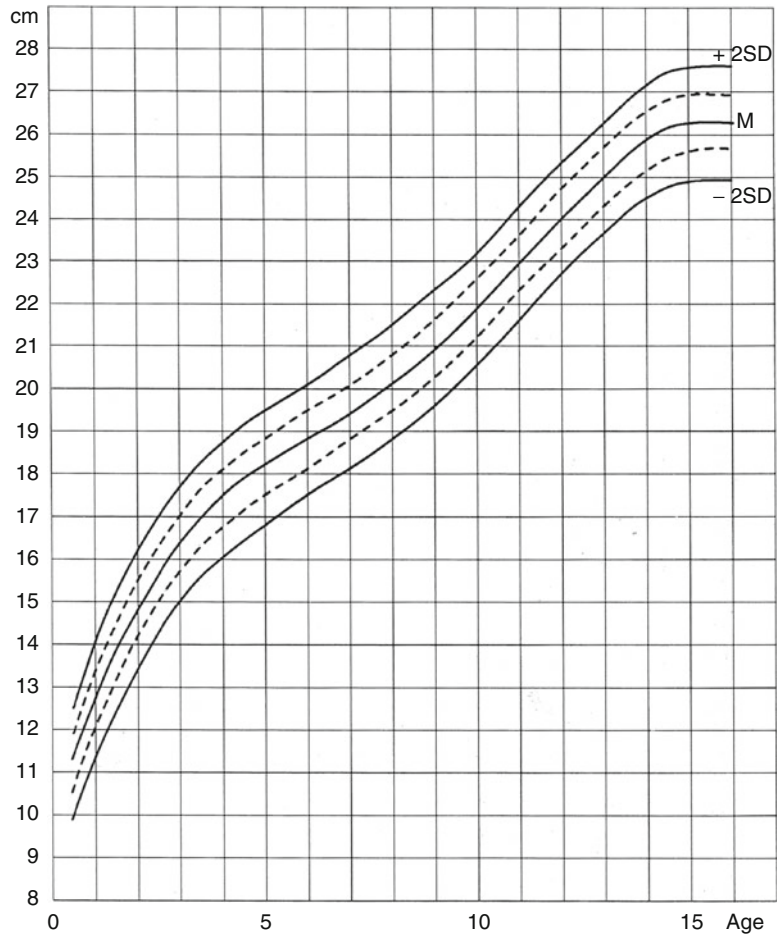


Fig. 4.20 T1–T12 segment length-for-age (birth to skeletal maturity: girls)

The thoracic circumference measures approximately 96 % of the sitting height [22, 26]. These two measures do not grow simultaneously, especially during puberty. At 10 years of age, the thoracic circumference is at 74 % of its final size, whereas the sitting height is almost at 80 % of the expected measurement at the end of growth. The transverse and anteroposterior diameters, which can be measured with obstetrical calipers, are two more parameters to assess the growth of the thorax. At the end of growth, the thorax has an anteroposterior diameter of about 21 cm in boys and 17 cm in girls, that is, it has increased by 9 cm since birth. The transverse diameter is 28 cm in boys and 24 cm in girls at the end of growth, that is, it has increased by 14 cm since birth. The transversal diameter makes up 30 % of the sitting height, and the anteroposterior diameter constitutes 20 %. The sum of the measurements of the transverse and anteroposterior diameters of the thorax should equal 50 % or more of the sitting height.

All parameters do not progress at the same speed, at the same pace. At 5 years of age, the increased weight and thoracic volume remain offset relative to the other parameters: sitting height and standing height. At birth, the thoracic volume is about 6 % [23]. At 5 years, it is 30 %. From birth to the age of 5, thoracic circumference grows exponentially and thoracic volume increases fivefold. During this period, the thorax experiences its most rapid growth. At 10 years, it is 50 %. The thoracic volume doubles between 10 years and skeletal maturity. At 5 years, the remaining growth of the thorax is about 70 % and the remaining sitting height is about 35 %. In treating scoliosis, the

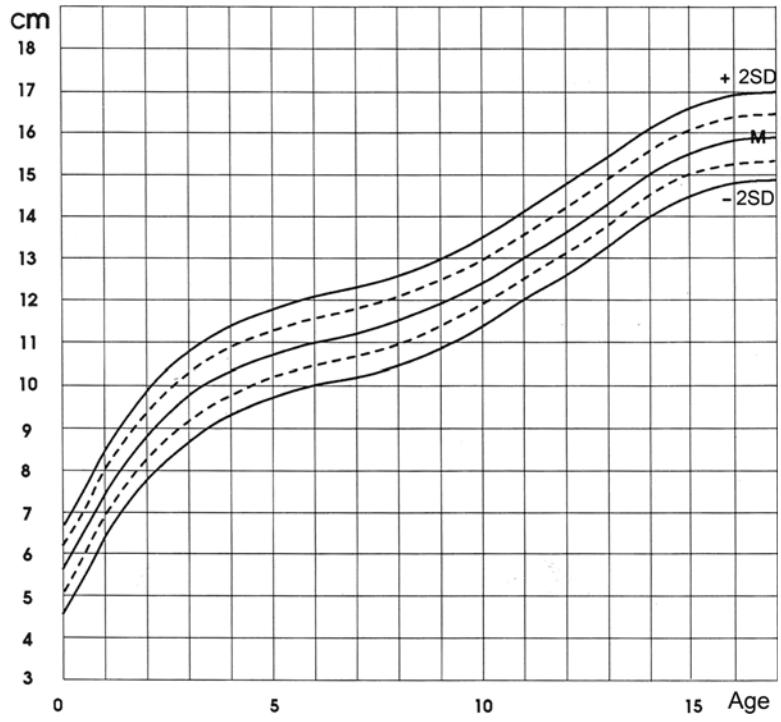


Fig. 4.21 L1–L5 segment length-for-age (birth to skeletal maturity: boys)

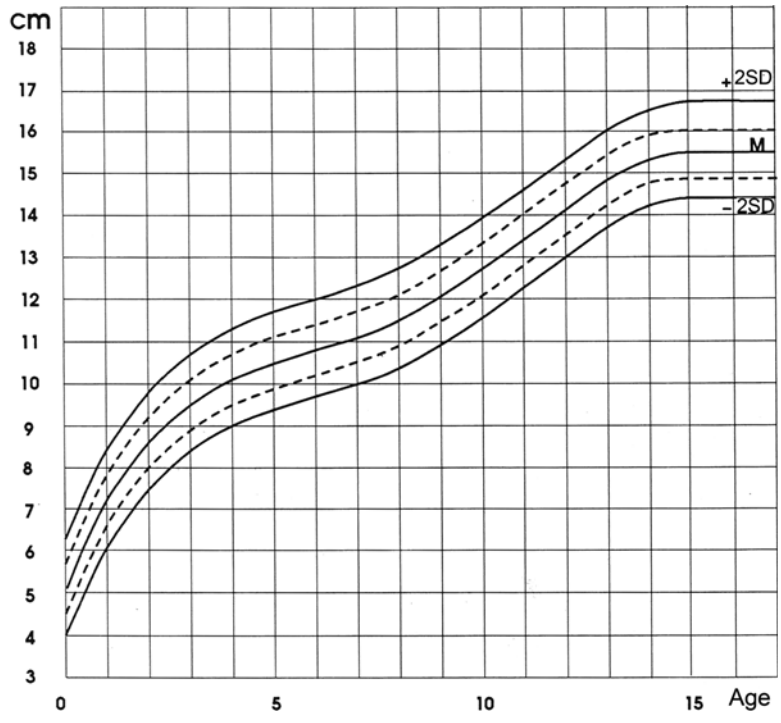


Fig. 4.22 L1–L5 segment length-for-age (birth to skeletal maturity: girls)

morphology of the thorax must be taken into consideration [22]. As the curve progresses, not only the growth of the spine is affected but also the size of the chest cavity is diminished. This will affect the development of the lungs which can create significant respiratory problems [8–11, 28].

Campbell et al. [8–11] have described the thoracic insufficiency syndrome, defined as the inability of the thorax to support usual respiration and lung growth. The opening wedge thoracostomy increases the volumetric thoracic growth (parasol effect); Dubousset et al. [28] have shown that severe scoliosis leads to penetration of the vertebra inside the thorax and have described the spinal penetration index. Severe scoliosis has a negative effect on the sitting height and the morphology of the thorax.

4.6.7 Scoliosis and Puberty

During puberty, trunk growth is significant.

The sitting height plays an essential part in the treatment of scoliosis; unfortunately, it is not recorded often enough. Gain in sitting height always needs to be compared with angular development of the spine. This relation is all that is needed for the proper assessment of treatment efficacy. If there is an increase of the sitting height without worsening of the curve angulation, the treatment is definitely working well. If, on the other hand, it is accompanied by deterioration of angulation, the treatment needs to be reconsidered.

When we treat scoliosis, we must also think of growth. In congenital scoliosis, the intrauterine growth and that occurring in the first few years of life can reveal a great deal about the future behavior of the spinal curvature. In idiopathic infantile and juvenile scoliosis, the growth during the first 10 years of life can be very important and may give dues to the behavior of the spinal curvature during the pubertal growth spurt [22–24, 26, 30].

However, in adolescent idiopathic scoliosis – the most common form of scoliosis – there is no such information available before the spine begins to curve in puberty. The ultimate outcome of the curve will be determined during the pubertal growth spurt. Therefore, monitoring the behavior of the spinal curve during this short and decisive period gives the only dues to its natural history. To detect these dues, it is necessary to know the onset of puberty.

The natural history of the curve of the spine can be judged on the ascending side of the pubertal growth velocity diagram corresponding to the first 2 years of puberty (from 11 to 13 years of bone age in girls and from 13 to 15 years of bone age in boys). Any spinal curve increasing by 1° each month (12°/year) during the ascending phase of the pubertal growth diagram is likely to be a progressive curve that will require treatment. Any curve that increases by 0.5° each month during this phase must be monitored closely, whereas a curve that increases by less than 0.5° each month during this phase can be considered mild [16, 26]. This observation of the natural history of the spinal curve during the early part of puberty gives information about the behavior of the curve during the last phase of puberty, as growth is slowing, and thereby gives guidance about the frequency of follow-up visits and the duration of bracing.

4.6.8 The Scoliotic Risk

It is therefore clear that scoliotic risk evaluation plays an essential role in the treatment of the disease. During the ascending phase of the pubertal growth diagram, a 5° curve is associated with a 10 % risk of progression, a 10° curve represents a 20 % risk, a 20° curve carries a 30 % risk, and a 30° curve raises the risk to virtually 100 % [15, 16, 44].

The risk of scoliosis decreases on the descending phase of the puberty growth diagram. At Risser 1 (13.6 years of bone age in girls and 15.6 years of bone age in boys), there is a 10 % risk of progression for an angulation of 20° and a 60 % risk for a 30° curve [20, 24, 26, 45] (Fig. 4.23).

At Risser 2 (14 years of bone age in girls and 16 years of bone age in boys), there is still a 30 % risk of progression (5° or more) for a 30° curve and a 2 % risk for a 20° curve [57]. At Risser 3 (14.6 years of bone age in girls and 16.6 years of bone age in boys), there is a 12 % risk of a curve of 20° or greater progressing by 5° or more [11]. At Risser 4 (15 years of bone age in girls and 17 years of bone age in boys), the risk of the pro-

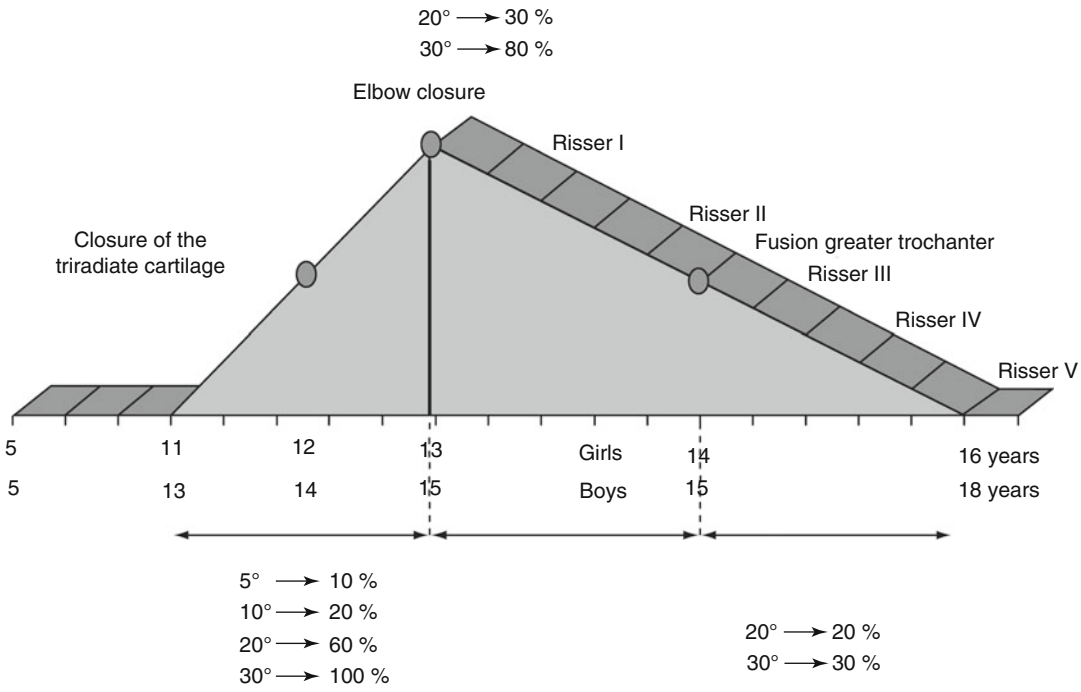


Fig. 4.23 Evaluation of scoliosis progression risk in pubertal growth diagram

gression of scoliosis is markedly decreased, although, for boys, a slight risk remains. At Risser 5 (16 years of bone age in girls and 18 years of bone age in boys), it would be futile, if not naive, to wait until the iliac crest is completely ossified before discontinuing the treatment of scoliosis, but there is still a risk of worsening in boys who have idiopathic scoliosis between Risser 4 and Risser 5 [40].

However imprecise and approximate the Risser sign may be, it is widely used as a deciding factor in many reports of brace treatment or surgery. Nevertheless, its limitations must be understood. The data of the studies by Lonstein and Carlson [45] relating Risser sign and the curve magnitude have been discussed. As was discussed previously, because two-thirds of the pubertal growth spurt occurs before the appearance of Risser I and often ambiguous relation exists between Risser stage and bone age, its value in both clinical decision making and research should be questioned [22–24, 43]. Bone age, abnormal growth velocity, and secondary sexual characteristics are the most reliable parameters. Risser stages must not be regarded as a first-choice indicator; they must always be compared with bone age [53] especially when making decisions that will have major consequences, such as ordering or removing a brace or scheduling vertebral fusion. Does growth stop at Risser 5? What is the best parameter? Growth stops when standing height does not progress (the best parameter). When the distal epiphysis of the ulna and the radius are closed, at the same period, the proximal epiphysis of the humerus closes [38].

4.6.9 Growth in the Paralytic Child

The growth pattern is abnormal in many children with paralytic disorders (e.g., cerebral palsy, spina bifida, and poliomyelitis). In these children, therefore, it is essential to record and follow the parameters of growth closely to establish an indication for surgery with as much accuracy and safety as possible. There are two problems that make it difficult to mea-

sure and evaluate the parameters of growth in such children. First, contractures and deformities make morphometric measurements difficult or even impossible. Second, reference values of children with normal growth and development are not applicable to these children [26, 48].

Nevertheless, it is still possible to gain valuable information about growth, first by measuring arm span when the child is in a wheelchair and, second, by scrutinizing the child carefully from head to foot. The length of even one bone that has been more or less spared by the deficit could be sufficient to determine the standing height of the child. For example, after 8 years of age, the proportions of the body segments remain the same; therefore, the length of the femur represents 28 % of standing height, and the length of the tibia and fibula represents 24 % of standing height. For the upper extremities, the humerus represents 19 % of standing height and 36.5 % of sitting height, the radius represents 14.5 % of standing height and 27.8 % of sitting height, respectively, and the ulna represents 15.5 and 29.5 %, respectively [18, 39]. These figures are particularly useful when sitting and standing height cannot be assessed such as in nonambulatory patients with mental delay and multiples comorbidities.

Weight is an important parameter to take into consideration. Many children, especially those with cerebral palsy and total body involvement, have a deficit of 20–30 kg. Surgical procedures are not the same for children who weigh 20, 40, and 60 kg. Being underweight as a result of malnutrition creates a risk of infection after surgery. There are many parameters that are used to assess nutritional status, such as measurements of the triceps and the subscapular skinfold or determination of the total lymphocyte count. Whichever tests the surgeon relies on should be carried out on underweight children before surgery, especially on those children with chronic conditions.

On the other hand, obesity can also be a problem in surgery. The obesity of children with muscular dystrophy or spina bifida may restrict the choice of surgical approaches and instrumentation. The gain in weight during puberty is the main enemy of children with diplegia or ambulatory quadriplegia.

The assessment of bone age is more difficult in paralyzed children. Bone age retardation is frequent in severe cerebral palsy. These patients sometimes display a wide range of bone ages, with the bone age of the hand not matching that of the elbow or the pelvis (this observation is made from the author's personal experience). The real bone age, therefore, must be approximately assessed, and this information must be correlated with the results of anthropometric measurements.

4.7 Lessons Learned from Growth

Charts and diagrams are only models or templates. They do not by themselves define a true age. They define trends and outline the evolution of growth. They should be taken as just what they are: a convenient means to map the route through puberty. They record ephemeral points in the processes of growth and anticipate the events that lie in the future.

Their use helps the surgeon in avoiding uncertain or unnecessary treatments and aids in developing successful strategies. Nothing can produce worse results than decisions leading into uncharted territory.

Annual growth velocity is an essential parameter, mainly to detect the pubertal peak velocity. Birthdays are a convenient reminder for annual evaluations such as measurements of growth. Percentages provide an extremely valuable and objective tool for evaluating residual growth, particularly with respect to the proportions between the lengths of various segments of the limbs and between the limbs and the trunk. The multiplying coefficient can be applied to all biometric data. However diverse their ethnic origins and although stature has been increasing in succeeding generations over the centuries, boys of all generations and ethnic backgrounds will always have approximately 14 % of outstanding growth in standing height

(13 % for girls). Neither the percentage nor the proportions change, and even the ratios are stable. The humerus is equivalent to, and will always be equivalent to, approximately 19 % of standing height and 36.5 % of sitting height [20]. Whatever be the population profile, *the chronology of growth and stages of puberty remain the same*.

A figure in isolation is meaningless; a ratio is more reliable. For instance, the length of the thoracic segment in relation to sitting height provides more objective values. To gain this information, the examiner should try to obtain a general overview of the child's growth and plot the child's anthropometric chart. The ratios of the various body segments are important in many conditions, especially the various types of dwarfism. The ratio of sitting height to subischial leg length is essential when analyzing chondrodystrophy. Special curves can be used to follow such patients. Dwarfism can be divided into two families: short-trunk dwarfism, the prime example of which is represented by Morquio syndrome, and normal-trunk dwarfism, which is characterized by the limbs being shorter than normal. The prime example here is achondroplasia.

The various processes that make up growth are well synchronized, organized, and interdependent, but they vary widely in the time of occurrence during growth. For instance, the growth of the trunk accounts for most of the increase in standing height during the last part of puberty. Also, weight gain lags behind growth in length until puberty, after which the percentage gain in weight far exceeds the percentage gain in height. All changes are gradual. Growth itself is a succession of phases, periods of deceleration or acceleration, spurts, and alternating processes.

The pubertal growth velocity diagram is very useful in decision making. The peak height growth velocity takes place during the first 2 years of puberty. By measuring the standing height, the sitting height, and the subischial leg length every 6 months, it becomes much easier to understand the puberty growth spurt. Bone age must be analyzed with a critical mind and constantly compared to the rate of annual growth in standing height and secondary sexual characteristics [20, 24, 26].

Treatment of children often requires a consideration of remaining growth. Puberty is the time when most of these decisions will be made. In children in whom growth disturbance is anticipated, it is best to record several parameters over time in order to have an accurate picture of growth.

Treatment is easiest when it is done in anticipation of future growth. The milestones that mark the growth path during puberty must be noted and understood by the orthopedic surgeon [20, 24, 26].

4.8 Growth of the Spine with an Early-Onset Deformity

Patients with early-onset scoliosis are heterogeneous population characterized by multiple etiologies. There is not only a single management strategy. There is no absolute truth. It is necessary to adapt treatment to each patient's need.

4.8.1 Early-Onset Scoliosis Has Negative Effects on a Growing Child: It Is a Physeal Disorder

Early-onset spinal deformities have very negative effects on growing children. In young children with progressive deformity, there is a decrease of longitudinal growth and a loss of the normal proportionality of trunk growth. Abnormal growth leads to a deficit that sustains the deformity.

4.8.2 Early-Onset Scoliosis Has a "Domino" Effect and It Becomes a Pediatric Disease

As the spinal deformity progresses, by a "domino effect," not only spinal growth is affected but size and shape of the thoracic cage are modified as

well. This distortion of the thoracic cage will ultimately interfere with lungs development and cardiac function. Over time, the spine disorder changes its nature; from a mainly orthopedic issue, it becomes a severe pediatric, systematic disease with Thoracic Insufficiency Syndrome, Cor Pulmonale, and – in most severe cases – death. These deformities can be lethal in the most severe cases as a result of reciprocal interactions and influences among the various skeletal and organic components of the thoracic cage and cavity that are not well understood. The development of the thoracic cage and lungs is a complex process that requires perfect synergy among the various components of the rib-vertebral-sternal complex. Alterations in any of these elements affect and change the development and growth of the others.

4.8.3 The Pathologic Spine Is Dominated by the Crankshaft Phenomenon

The crankshaft phenomenon is a constant concern. Theoretically, when dealing with a severe scoliosis, the best arthrodesis is the perivertebral arthrodesis. Posterior arthrodesis in the immature spine induces the cranckshaft phenomenon. For severe cases, and, particularly in congenital scoliosis, early hemiarthrodesis before 5 years of age has been proposed, but experience has shown that early arthrodesis has negative effect [27, 51].

There is currently no instrumentation that is able to control the three-dimensional nature of early-onset spinal deformities. The ideal method of early-onset scoliosis treatment has not been identified yet.

4.8.4 Surgical Management Depends on the Age of the Patient

Age is an important parameter that must be taken into account. Clearly, the 1-year-old child, the 5-year-old child, and the 9-year-old child with spinal deformity are different facets of the problem and represent different opportunities for care.

4.8.4.1 From Birth to 5 Years of Age

During this period, priority should be given to the lungs. The weight of the lung will increase by tenfold, from 60 g at birth to 750 g. Up to 85 % of alveoli develop after birth. Alveoli are added by multiplication after birth until the age of 8 years. The lungs volume increases by sixfold during the first years of life (Fig. 4.24 and Fig. 4.2). The golden period of the thoracic

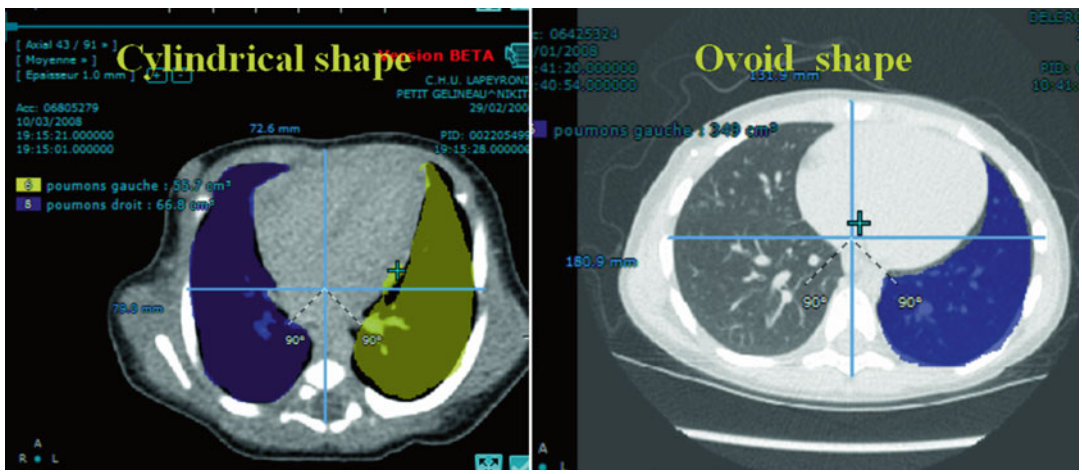


Fig. 4.24 Morphology of the thorax. It is cylindrical at birth and becomes ovoid at 5 years. Between birth and 5 years of age, lungs volume increases by sixfold

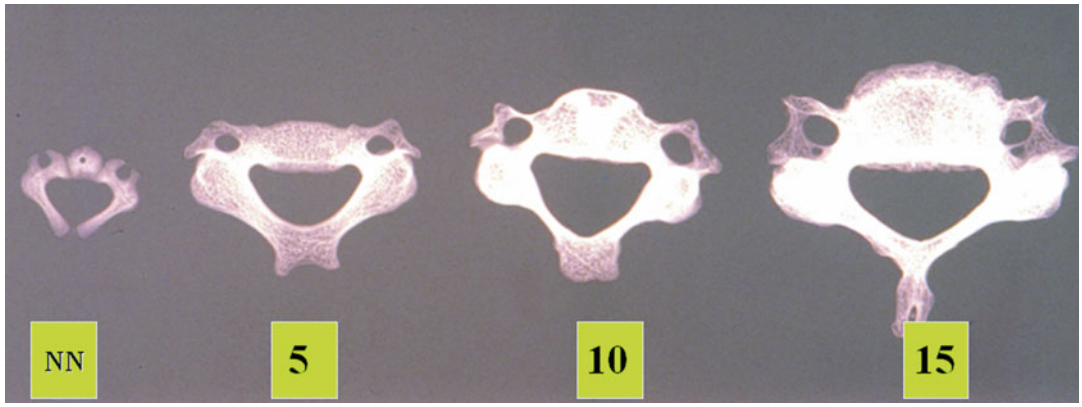


Fig. 4.25 Vertebral body ossification and spinal canal size from birth to skeletal maturity; by the age of 5 years, ossification raises up to 65 % and the spinal canal has grown to 95 % of its definitive size

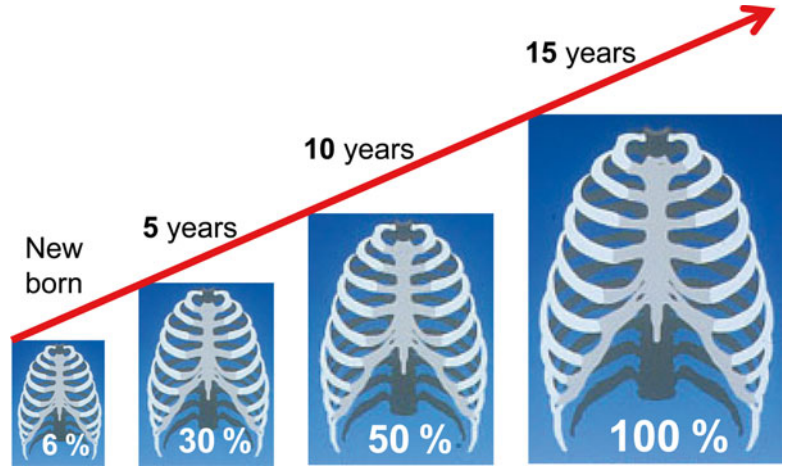


Fig. 4.26 Volumetric growth of the thoracic cage between birth and skeletal maturity

spine and rib cage coincides with lung development. The source of the respiratory failure is twofold: intrinsic alveolar hypoplasia and extrinsic disturbances of the chest wall function. The chest deformity prevents hyperplasia of the lung tissues (Campbell (2000) personnal communication) [8, 10, 11].

Patients with early-onset spinal deformities tend to have reduced body weight. Growth requires an enormous amount of energy. The nutritional requirements in the first 3 years of life are much greater than those of adulthood: calories, 110 vs. 40 cal/kg/day; protein, 2 vs. 1 g/kg/day; and water, 150 vs. 5 ml/kg/day. Skeletal mineralization alone requires storage of 1 kg of calcium between birth and adulthood.

During the first 5 years of life, surgery can be challenging due to poor bone quality and reduced size of vertebral bones. At birth, the spine is mainly cartilaginous. Only 30 % of the spine is ossified; by 5 years of age, ossification raises up to 65 %. At this age, the spinal canal has grown to 95 % of its definitive size [21–23] (Fig. 4.25).

From birth to 5 years, the morphology of the thorax changes; it is cylindrical at birth and becomes ovoid at 5; the frontal diameter grows more than the anteroposterior diameter [22] (Fig. 4.24). During this period, the thoracic cage is mainly cartilaginous and susceptible to morphological modifications (Fig. 4.26).

It must be reminded that during the first 2 years of life, sitting height increases by 20 cm,

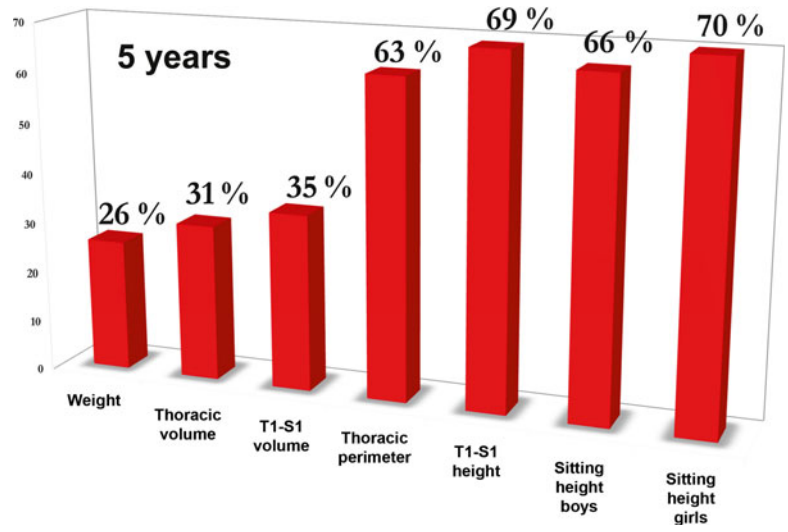


Fig. 4.27 After birth, growth parameters do not proceed at the same speed

12 cm of which is during the first year. The gain of 12 cm during the first year of life corresponds to the gain occurring during puberty. Moreover, during the first 5 years of life, the gain in sitting height is 28 cm, which corresponds almost to the growth occurring between 5 years of age and puberty (30 cm). All growth parameters do not progress at the same peace (Fig. 4.27).

4.8.4.2 From Five to Puberty, a Quiescent Period

During this quiescent phase of vertebral growth, it is important to improve both weight and respiratory function with intensive physiotherapy. The growth velocity of the sitting height and T1–S1 slows down. Vertebral growth is reduced and is between 0.7 and 1 mm/year per vertebra. T1–S1 will increase from 5 to 10.6 cm. The annual growth velocity on the standing height is 5.7 cm: 2.4 cm on the trunk, 3.3 cm on the lower limbs [21–23].

Remaining growth is an important parameter to choose the best management strategy. At 5 years of age, the remaining growth on T1–S1 is about 15 cm, two-thirds on the thoracic spine (10 cm), one-third on the lumbar spine (5 cm). Remaining standing growth is about 65 cm at 5 years. Remaining sitting growth is about 32 cm [21–23].

At 10 years of age, the thoracic volume will increase from 30 to 50 % [17, 21–23]. A distraction of 1 cm/year is recommended [1]. Effect on the impact of the distraction can be appreciated by measuring the sitting height [21–24, 26]. At 10 years of age, the remaining growth of T1–S1 is about 11 cm for boys, 7 cm for girls. Remaining standing height is 38 cm for boys and 26 cm for girls. Remaining sitting height is 20 cm for boys and 15 cm for girls. The remaining growth on the thoracic perimeter is about 33 cm for boys and 31 cm for girls.

4.8.4.3 Puberty, a Period Characterized by Progression of the Curve: The Last Increase in Height, the Last Challenge to Overcome

Puberty is characterized by a significant increase in annual growth velocity. However, many patients with early-onset spinal deformities have multiples comorbidities and may not undergo a real pubertal growth spurt, i.e., Rett syndrome, cerebral palsy [22–24, 29, 30].

During puberty, weight increases by 5 kg/year and thoracic cage volume doubles its size. A curve of 30° at the onset of puberty or a curve progressing more than 1°/month has a surgical risk of nearly 100 % [16, 32, 54]. T1–S1 will increase from 10 to skeletal maturity, about 9.5 cm for boys, 6.5 cm for girls [22–24, 26]. Aggressive scoliosis in the first year of puberty should be detected soon and offensive strategy be chosen. Severe scoliosis should be treated early and, if necessary, on the ascending side of puberty.

4.8.5 Growth Disturbances After Early Spinal Arthrodesis

Mehta et al. [47] have shown that in a growing rabbit model, there is an interaction between growth of the spine and thorax; a unilateral deformity of the spine or the thorax induces both scoliosis and thoracic cage deformity with asymmetric lung volumes. Karol et al. [41] have shown that early arthrodesis reduces the AP diameter and shortens the T1–T12 index. Fusion is a cause of respiratory insufficiency and adds to the spinal deformity, the loss of pulmonary function. The forced vital capacity may decrease to less than 50 % of predicted volume if more than 60 % of the thoracic spine (i.e., eight thoracic volume) is fused before the age of 8 years. Emans et al. [31] have confirmed this negative effect of early arthrodesis. Canavese et al. [12, 13] have shown that dorsal arthrodesis in prepubertal rabbits changes thoracic growth patterns in operated rabbits. The dorsoventral diameter grows more slowly than latero-lateral diameter does. The sternum, as well as the lengths of the thoracic vertebral bodies in the spinal segment, where the dorsal arthrodesis was performed, grew less. The cranckshaft phenomenon is evident at the fused vertebral levels where there is a reduction of thoracic kyphosis [12].

4.8.6 How to Calculate a Deficit Induced by a Vertebral Arthrodesis in a Growing Child

When planning a perivertebral arthrodesis, we should know what the deficit of the trunk will be [63]. The remaining sitting height and the figure 2.5 % of sitting height for thoracic vertebra and 3.5 % for lumbar vertebra are the elements to take into consideration.

For example, a thoracic spine arthrodesis of five vertebrae at age 5 years will generate a deficit in sitting height of 3.2 cm and 3.8 cm in girls and boys, respectively. Remaining sitting height is about 26 cm for girls and 31 cm for boys. The thoracic spine makes up 30 % of the sitting height. The remaining growth of the thoracic spine (12 vertebrae) corresponds to $remaining\ sitting\ height \times 30\ \% = \underline{Y}$. The deficit of the sitting height for n vertebral will be $Y/12 \times n$. The same calculation can be done for the lumbar spine knowing that it makes up 18 % of sitting height. For girls, a perivertebral arthrodesis at the beginning of puberty will cause a deficit of 3.6 cm on the thoracic segment and 2.1 on the lumbar segment. For boys, a perivertebral arthrodesis at the beginning of puberty will cause a deficit of 3.9 cm on the thoracic segment and 2.3 on the lumbar segment. The deficit on the trunk will be outbalanced by the correction of the deformity (Figs. 4.28 and 4.29).

4.8.7 A Thorough Pediatric Evaluation Is a Priority

Because of the great diversity of diseases involved in early spine deformities, a complete pediatric investigation is strongly recommended as patients with early-onset scoliosis have frequently associated comorbidities.

4.8.8 What We Know, Where We Are, and Which Way to Follow?

Growth is a change in proportions; it is a volumetric revolution, and it is not linear. Growth parameters do not proceed at the same peace. All growths are interrelated. Any abnormal growth, by a "domino effect," leads to another abnormal growth. The irregular growth of vertebral bodies is the basis of a distorted development. Severe, progressive early-onset spinal deformities lead to abnormal spine growth that alters thoracic and lungs growth, which finally affects the cardio-pulmonary system. Only perfect knowledge of normal growth parameters allows a better understanding of both normal and abnormal spine and thoracic cage growth and of the pathologic changes induced on a growing spine and chest by an early-onset spinal deformity. There is a normal interaction

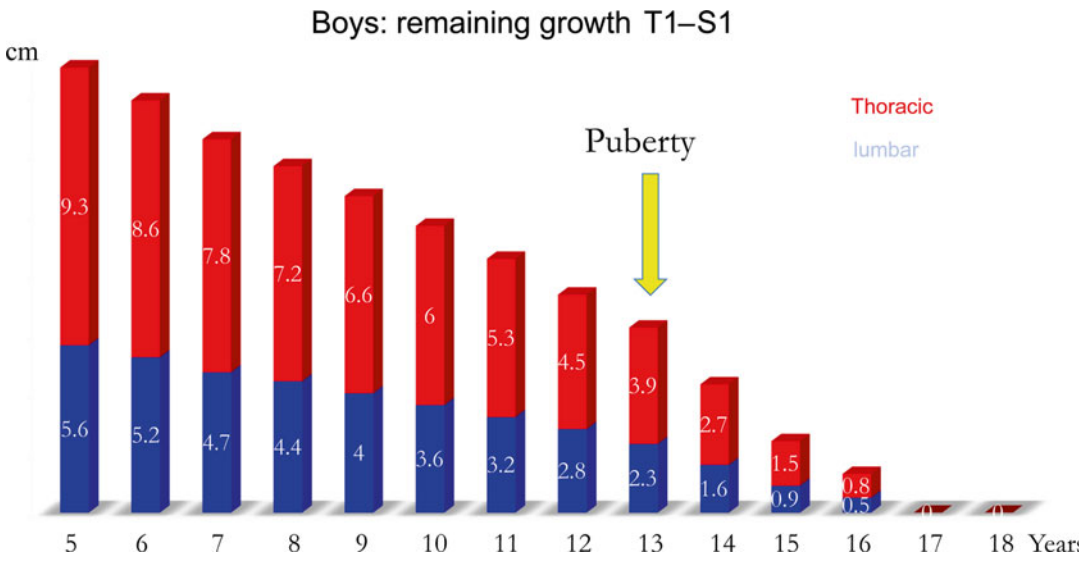


Fig. 4.28 T1–S1 remaining growth in boys (*red*: thoracic spine; *blue*: lumbar spine)

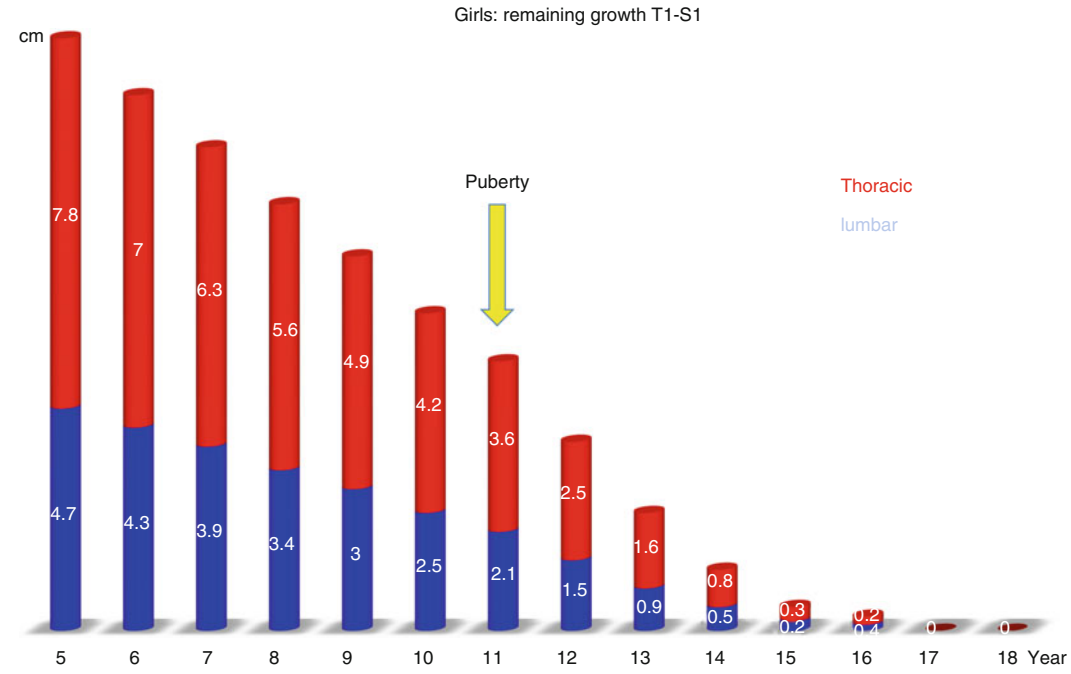


Fig. 4.29 TI–S1 remaining growth in girls (*red*: thoracic spine; *blue*: lumbar spine)

between the organic components of the spine, the thoracic cage and the lungs.

Deformities of the spine adversely affect the development of the thorax by changing its shape and reducing its normal mobility [32]. The rib vertebral complex which fits the thoracic cavity three dimensionally tends to constitute an elastic structural model similar to a cube in shape, but in the presence of scoliosis, it becomes flat and rigid and turns elliptical, thus preventing the lungs

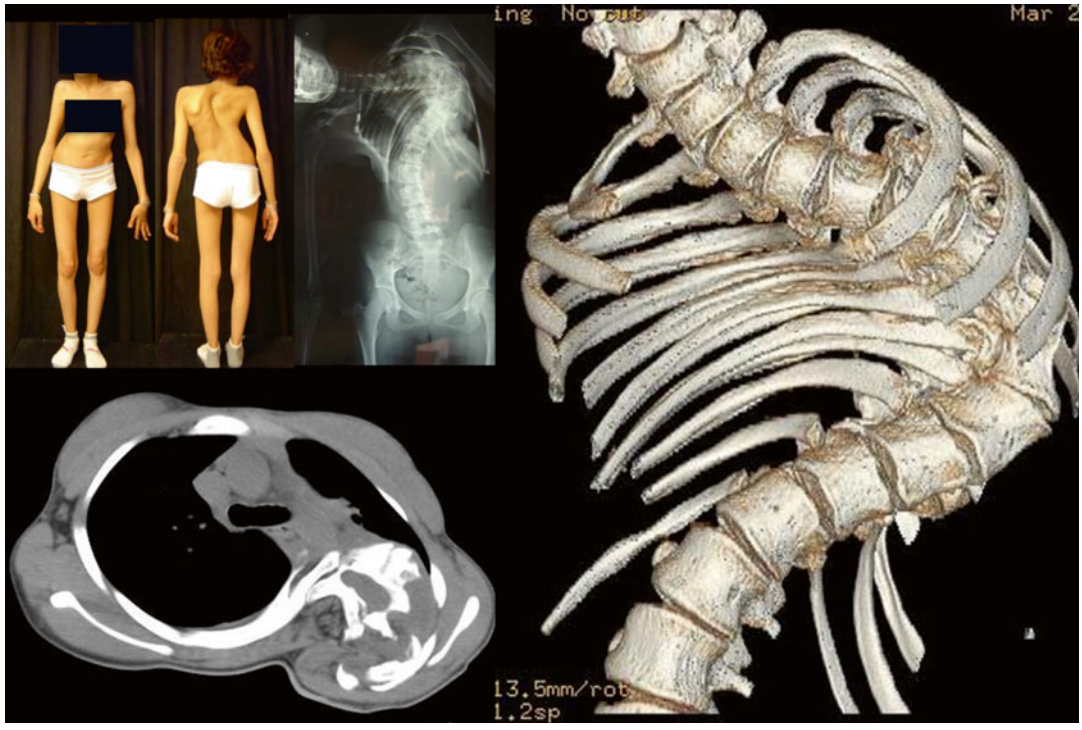


Fig. 4.30 On the C-T scan, we can see the penetration of the vertebra inside the thorax with crushing of the right lung

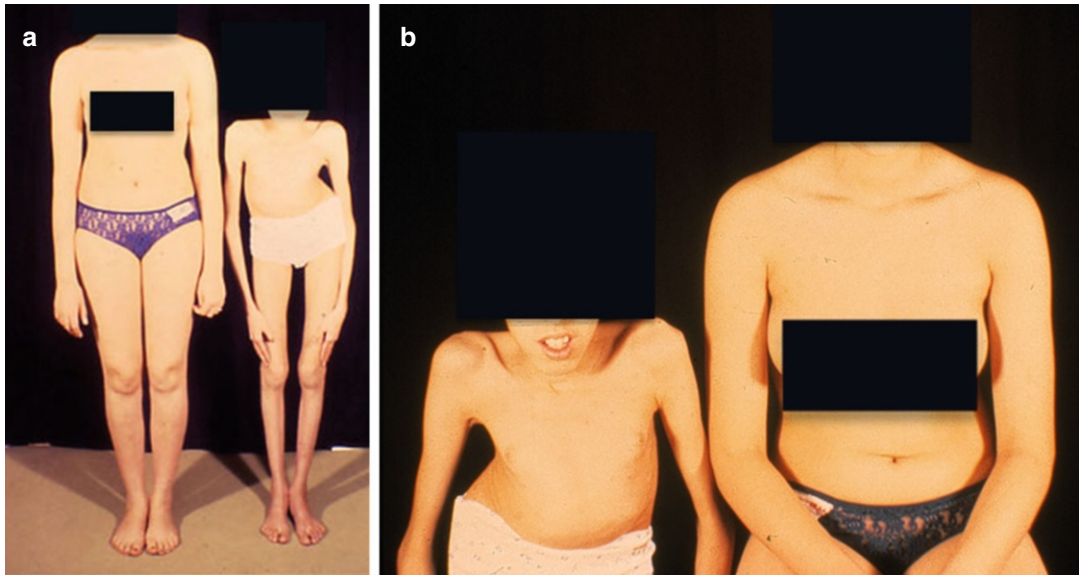


Fig. 4.31 Comparison between normal patient and patient with early-onset spinal deformity. The lower limb has normal growth for the patient with severe early onset scoliosis but there is a significant deficit of the sitting height

from expanding [13] (Figs. 4.30 and 4.31). Early posterior arthrodesis in the central portion of the spine (T1–T6) disturbs significantly the morphology of the thorax and blocks the thoracic volume [5]. Before the age of 5 years, the retractions of the thorax should be treated to preserve the pulmonary growth [8].

Innovative techniques such as expansion thoracoplasty [8] and dual rod distraction [1], stapling, screwing of the neurocentral growth plate, offer the possibility of preventing thoracic insufficiency and spinal deformity. However, the ideal surgical treatment of early-onset scoliosis has not yet been identified. Magnetically controlled, remotely distractible growing rod systems have been developed to reduce the number of repetitive surgeries under general anesthesia, to decrease the number of hospitalizations, to facilitate out-patient rod distractions, to reduce the number of wound complications and psychological problems. It is very close to the principles of distraction osteogenesis introduced by Ilizarov about four decades ago. This technique still presents some technical imperfections that need to be improved.

Surgery must be mini-invasive. Due to repetitive surgical procedures, however, the surgeon gradually seizes almost the whole spine and forgets that is necessary to spare levels as well as spinal motion. It is important not to forget that between T1 and S1, there are only 18 vertebrae.

Challenging the growing spine means how to maintain the spinal growth, the thoracic growth, the lung growth and to keep the spine supple [22]. The principle that a short spine produced by early fusion is better than a long curved spine is no longer generally accepted [36].

The obsession of the centimeter does not have to distract the surgeon away from the fundamental priorities. The surgeons must keep in mind that simple objectives have to be achieved in particular (a) improvement of the clinical picture, (b) a thoracic spine height of 18–22 cm (in order to avoid severe respiratory insufficiency), (c) a vital capacity of at least 50 %, and (d) weight gain of about 2.5 kg per year or a weight of at least 40 kg.

The child with severe early-onset spinal deformities must not become a full-time patient or a juxtaposition of surgical procedures. The ultimate goal of treatment is to improve the natural history of the patient's spinal deformity as well as the quality of life and to have these sick children become independent adults.

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Thoracic Insufficiency Syndrome

5

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Key Points

- Thoracic insufficiency syndrome (TIS) represents a variety of skeletal and muscular disorders that are severe enough to impair respiratory function to the point that treatment is necessary.
- TIS is a progressive disorder that varies in the rate of progression but leads to restrictive lung disease in most cases and additional obstructive disease due to large bronchi distortion in a minority of children.
- Forced vital capacity is the diagnostic index of choice to serially monitor changes in lung function over time. However, overnight sleep studies (polysomnograms) are useful to guide respiratory treatment before, during, and after surgical intervention.

Abbreviations

BIPAP	Bi-level positive airway pressure
EOS	Early-onset scoliosis
MEP	Maximum expiratory pressure
MIP	Maximum inspiratory pressure
REM	Rapid eye movement
TIS	Thoracic insufficiency syndrome

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5.1 Thoracic Insufficiency Syndrome

Thoracic insufficiency syndrome (TIS) is defined as the inability of the thorax to support normal respiratory function and postnatal lung growth in children with skeletal immaturity [1]. It is a term meant to encompass multiple disorders of the spine, ribs, sternum, and surrounding muscles that restrict lung volume, expansion, and postnatal lung growth. TIS also includes abnormalities of spine and thoracic structures that occur secondary to underlying neuromuscular disorders which produce either weakness or spasticity. Children with either primary or secondary TIS may have additional pulmonary diseases, such as aspiration pneumonias or primary pulmonary hypoplasia, but the functional assessment of breathing in children with TIS presumes that these other pulmonary issues have been optimally treated and that they contribute minimally to the restrictive respiratory disease.

The major spine deformities that produce TIS are congenital and infantile scoliosis involving the thoracic vertebrae with or without kyphosis, also known as early-onset scoliosis (EOS). The major thoracic cage abnormalities producing TIS are forms of thoracic hypoplasia and rib anomalies. These deformities vary in severity on initial presentation but often progress over time. EOS, in particular, is complex with different apices of spine curvature, numbers of vertebrae involved, and the types of vertebral anomalies, e.g., block vertebrae, unsegmented vertebrae, and hemivertebrae, all with or without rib fusion [2]. These lead to variable degrees of scoliosis, kyphosis, lordosis, and rotation of thoracic vertebrae and distort the normal configuration of the ribs and sternum. The specific impact of a single structural feature on lung function is difficult to assess. Multiple studies have shown that different lung function measurements have little relation to the Cobb angle alone in children with TIS [3, 4]. Given the variety of combinations of structural abnormalities of the vertebrae and ribs producing TIS and the variations in severity of each structural deformity, it is not surprising that thoracic and spine structural features in general do not correlate well with multiple respiratory measures of function.

An alternative approach is to use both structural and respiratory functional measures in conjunction with one another to determine the severity and the rate of progression of TIS in young children. This section describes the respiratory functional abnormalities that result primarily from EOS, as a form of TIS, and the tests available to quantify these abnormalities. Respiratory function is not limited to function of the lung; examples of respiratory functions are listed in Table 5.1. The most commonly used tests measure respiratory mechanics (and hence the work of breathing) and gas exchange. It is likely that as the spine and thoracic deformities progress, respiratory function deteriorates. However, some abnormalities appear only when severe deformity develops, e.g., pulmonary hypertension and retention of carbon dioxide. Most of the tests mentioned below have not been used in a standardized or uniform way to assess children with TIS. Figure 5.1 summarizes the relative frequency of respiratory abnormalities reported to date in these patient populations.

Table 5.1 Pulmonary function domains

Lung and chest wall mechanics
Lung and chest wall compliance
Chest wall excursion
Lung volumes
Gas exchange
Oxygenation efficiency
Carbon dioxide removal
Regional lung function
Ventilation and perfusion distribution
Respiratory muscles
Inspiratory and expiratory force generation
Thresholds for respiratory muscle fatigue
Pulmonary hemodynamics
Pulmonary hypertension and cor pulmonale
Pulmonary host defenses
Cough effectiveness

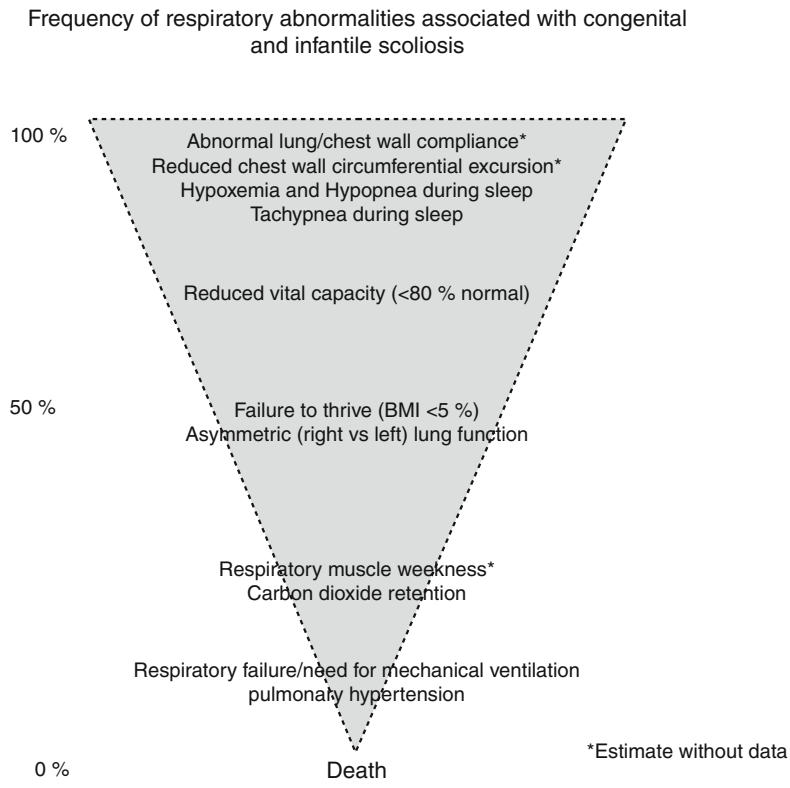

Fig. 5.1 Frequency of respiratory abnormalities associated with congenital and infantile scoliosis and other children with thoracic insufficiency syndrome

5.2 Lung and Chest Wall Mechanics

The shape and compliance of a child's thorax change with increasing age. The depth and width of an infant's chest are equal at birth and achieve the adult's AP/lateral dimension ratio of 0.7 by 2–3 years of age [5]. Ribs project at right angles from vertebrae at birth and achieve degrees of angulation seen at adulthood by the third year of life. Chest wall compliance is greatest in infancy and becomes stiffer with increasing thoracic muscle development and ossification of bony structures. "Normal" stiffness of the chest wall is therefore age dependent. In human newborns, the chest wall is seven times more compliant than the lung and active chest wall and diaphragmatic muscle tone is required to maintain resting lung volumes [6]. In children up to 5 years of age, the chest wall is almost twice as compliant as the lung during quiet breathing [7]. Between 6 and 15 years of age, the chest wall normally falls by approximately 30 % [8]. In adults, the ratio of chest wall to lung compliance is close to 1.0.

In children with scoliosis, chest wall compliance is reduced, as is rib cage excursion. In response to progressively stiff chest walls, children choose to breathe shallowly to reduce the work needed to expand the chest wall with each breath. Consequently, they breathe rapidly to maintain normal minute ventilation. Respiratory rate during wakefulness is higher due to activity, excitement, and speech and is more reflective of respiratory mechanics when measured during sleep. The normal respiratory rates of children, which are age dependent, are portrayed in Fig. 5.2. Tachypnea due to scoliosis is common, but the exact prevalences of tachypnea during wakefulness and sleep have not been reported.

Lung volumes increase as the thorax increases in size. Intra-thoracic volume (at maximum inspiration) at birth increases 33-fold to reach the adult size [9]. Processes that slow thoracic cage growth therefore reduce lung volume.

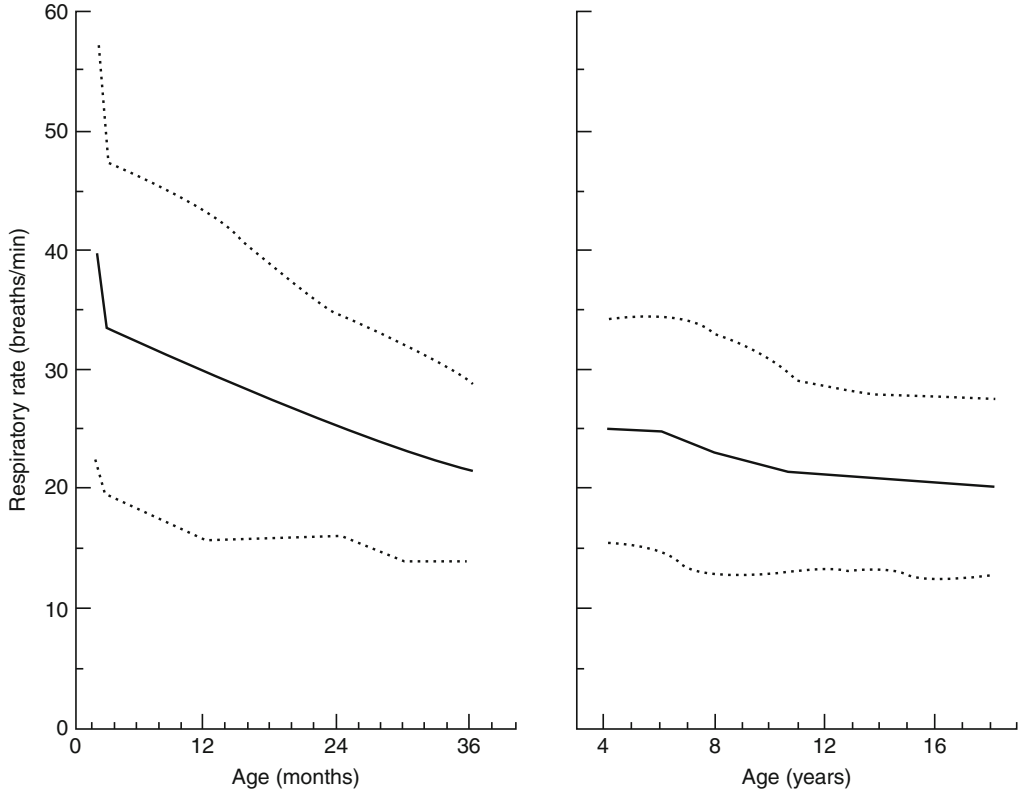

Fig. 5.2 Normal respiratory rates in awake children (panel **a**) [43] (panel **b**) [44]

Respiratory compliance or distensibility is composed of lung and chest wall components. Lung compliance, apart from chest wall compliance, is volume dependent. At very low lung volumes, the lung resists expansion more when intra-thoracic pressure changes. At normal lung volumes during tidal breathing, the lung is maximally distensible. Lung volumes and lung compliance have been measured in one series of young children with early-onset scoliosis under anesthesia and found to be low [10]. Surgical procedures that increase thoracic volume, such as growth friendly constructs, also increase resting lung volume [11]. However, these same procedures also reduce chest wall compliance with insertion and serial expansion of metal into the spine or chest. [12]

With prolonged constraint of the lungs by a small thorax, alveolar development can be hampered postnatally [13]. Lung growth after birth primarily occurs in the distal or acinar regions of the lung and with increased alveolar number, size, structural complexity, and alveolar-capillary surface area. These attributes of alveolar growth change at different times, so that alveolar number and complexity increase early in life while increased alveoli enlarge later in proportion to height. [14] Lung histology from adult rabbits with scoliosis who underwent unilateral rib fusion at 7 weeks of age shows simplified larger but fewer alveoli, illustrating how small thoracic size due to EOS produces postnatal pulmonary hypoplasia [15].

Scoliosis impacts respiratory mechanics in several ways and does so more when it begins in early life than with onset during adolescence [16]. Abnormal lung volumes have been measured using infant lung function techniques as early as 6 months of age in infants with chest wall disorders [17]. Lung volumes are subdivided as illustrated in Fig. 5.3; combinations of lung volumes are described as lung "capacities." *Vital*

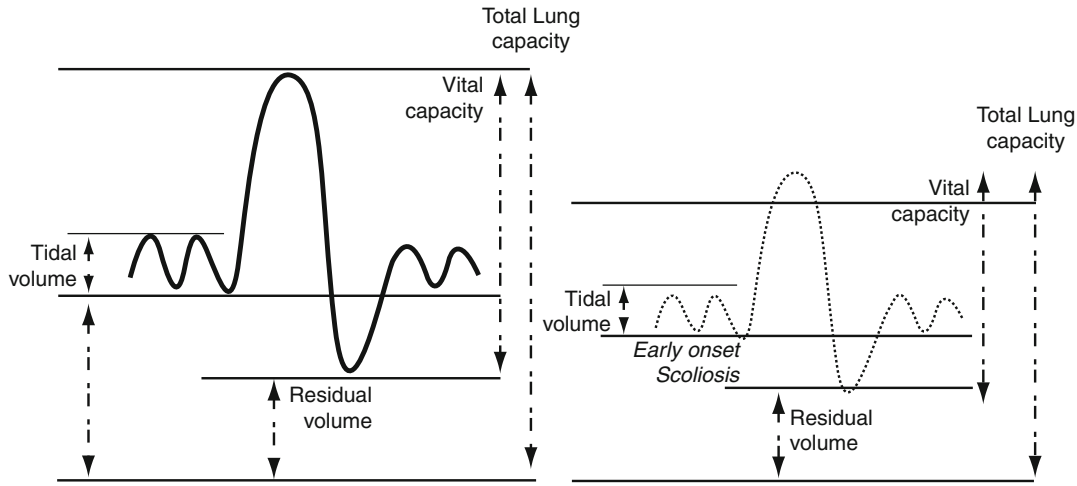

Fig. 5.3 Lung volumes and capacities in normal children and children with early-onset scoliosis

capacity is that portion of total lung capacity that can be voluntarily inhaled or exhaled with maximal efforts and reflects ventilatory reserve that could be used if needed. In contrast, residual volume is that volume of gas in the chest that remains after maximal exhalation. Residual volume provides a gas reservoir to sustain transfer of oxygen and carbon dioxide from air to blood compartments.

In children and adults with scoliosis, vital capacity is preferentially reduced as a result of reduced lung volumes, reduced lung and chest wall compliance, and reduced respiratory muscle function. This is illustrated in Fig. 5.3 compared to normal values. *Residual volume* is reduced less than the reduction in vital capacity, as it depends on thoracic cage volume and stiffness and less on respiratory muscle function. *Total lung capacity*, the combination of vital capacity and residual volume, is reduced less than the vital capacity because residual volume is least compromised [18]. Vital capacity is a more sensitive respiratory test to follow patients with TIS serially, as it reflects multiple changes in respiratory function. Studies in children with TIS suggest that there is greater reduction in vital capacity per degree Cobb angle if scoliosis begins earlier in life than when it begins in adolescence [16].

Vital capacity is measured by spirometry in children old enough to understand and perform maximal inspiratory and expiratory efforts. Reproducible artifact-free efforts that reflect lung function rather than how a child performs the test occur between 4 and 6 years of age. The major source of variability in spirometric results is the experience or lack thereof of the child performing the tests [19]. With practice, variability in test performance declines and coefficient of variation for vital capacity results can be as low as 5–8 % in well-practiced normal children [20]. All spirometric indices, including vital capacity, are compared to published norms that are created using height, ethnicity, and gender to normalize across different ages. In children with EOS, height reflects spine curvature as well as growth and is inappropriate to apply to reference values of normal children. Surrogates for expected height, such as arm span and ulnar length, are used instead to estimate expected height from which percent of predicted values can be derived [21–23]. In contrast to spirometry, measures of total lung capacity and all of its volume components require either gas dilution techniques or measurements with a body plethysmograph. These two techniques are available for children in tertiary care centers with a pediatric focus but are much less available than routine spirometry. Vital capacity is therefore the most clinically available spirometric test for clinicians managing children with TIS.

Spirometry also measures inspiratory and expiratory airflow. Diseases that narrow conducting airways produce obstructive lung disease. The degree of airway obstruction is usually quantified using the forced expiratory volume at 0.5 and 1 s (FEV 0.5 or FEV1) after the onset of a forced expiratory effort and the forced expiratory flow in the mid-half of the exhaled vital capacity (FEF, 25–75 % VC). FEV1 is usually normalized for vital capacity, and the normal ratio of FEV1/FVC is 80–85 %. In 4- and 5-year-olds who can perform spirometry, this ratio is 90 % or more. Importantly, FEV1/FVC is independent of age or height. Early-onset scoliosis that results in TIS produces obstructive lung disease in up to 30 % of children [24]. However, asthma is also a common condition among children, and if spirometric indices of airway obstruction are abnormal, then asthma should be ruled out using a bronchodilator challenge. Obstructive lung disease related to EOS is usually due to compression of one or both mainstem bronchi by vertebrae and mediastinal structures [24].

Pulmonary function tests in children can be subdivided by those that do and do not require cooperation and those that are more invasive in nature. Spirometric measures are non-invasive but require cooperation and maximal effort. Measures of maximal respiratory muscle strength are also effort dependent. Lung and chest wall compliance measurements do not require cooperation but do require placement of a pressure transducer within the esophagus to estimate pleural pressure. The invasive nature of esophageal balloons has led to measurements of lung compliance in the operating room under anesthesia but not during outpatient visits with children who were awake [10]. Passive inflation and deflation performed in sedated or anesthetized children do not account for the respiratory muscle dysfunction that occurs in TIS. Consequently, results obtained in supine children with EOS in the OR cannot be compared to results obtained in the same awake upright children in the clinic. The issue of cooperation in awake patients is particularly important as it limits the use of many tests in very young children with TIS who are being considered for surgical therapies. Table 5.2 lists commonly used tests of respiratory function based on the need for cooperation and for invasive procedures.

5.3 Gas Exchange

Respiratory gas exchange is measured by efficiency of oxygenation and sufficiency of ventilation. Oxygenation is usually measured as percent of hemoglobin saturated with oxygen (SpO_2) using non-invasive pulse oximeters. These devices have replaced direct measures of partial pressure of oxygen in arterial blood except in intensive care units. Oxygenation varies with wakefulness and sleep, and SpO_2 may fall by 2 % in normal children and adults during sleep [25]. In children with TIS, SaO_2 is normal (>96 %) in most children when awake. In the initial study of 218 children with TIS who underwent expansion thoracoplasty,

Table 5.2 Invasive and pulmonary function tests

	Pulmonary function testing in infants and children	
	Invasive	Non-invasive
Effort—Dependent[a]	Trans-diaphragm pressures	Spirometry Respiratory muscle strength and fatigability Exercise tests
Effort—Independent[a]	Chest wall and lung compliance	Respiratory rate Oximetry Lung volumes Blood gas tensions Lung ventilation/perfusion scans Sleep study Echocardiogram

[a]Voluntary effort at 5–6 years of age

only 22 % received supplemental oxygen therapy [26]. However, 10 of 11 children with TIS whose breathing was studied during sleep experienced recurrent but brief episodes of hypoxemia, particularly during rapid eye movement (REM) sleep [27]. Additionally, elevated hemoglobin levels were reported in 23 % of children with EOS, suggesting recurrent hypoxemia during sleep with increased erythropoietin levels and mild polycythemia [28]. Non-invasive positive pressure ventilation such as bi-level positive airway pressure (BIPAP) effectively improves hypoxemia during sleep in children with EOS [29].

Ventilation is assessed using arterial or arterialized measurement of carbon dioxide partial pressures in blood. Capillary sampling of arterialized blood is commonly used to measure PCO_2 in the outpatient setting. Retention of carbon dioxide above normal levels, i.e., PCO_2>50 mmHg, is uncommon and a late finding in children with TIS, reflecting acute and/or chronic respiratory failure. An alternative way to measure CO_2 status is with a capnograph which measures CO_2 concentration in exhaled gas. The end-tidal PCO_2 can be used as a non-invasive surrogate measure of arterial PCO_2 in children who have no underlying lung disease and only spine and chest wall disease. End-tidal PCO_2 has been used during sleep studies to identify frequency, duration, and severity of hypercapnia. Subacute and chronic hypercapnia can be assessed by the total CO_2 content in the blood, a measure of compensatory metabolic alkalosis in response to sustained respiratory acidosis. The utility of this measure is that it is less susceptible to sampling errors than capillary or arterial CO_2 tensions. It may also be elevated after transient hypercapnia during sleep when assessed early in the morning.

5.4 Lung Function Asymmetry

One consequence of progressive chest wall deformity due to EOS is an asymmetric chest wall shape and increasingly different volumes in the right and left hemi-thoraces. Different capabilities to move the right and left chest walls due to differences in rib alignment, hemi-diaphragm configuration, and regional chest wall compliances lead to asymmetric lung volumes and also to asymmetric ventilation and perfusion between the right and left lungs. Regional lung ventilation and perfusion are measured with lung scans. Ventilation is measured using inhalation of a radiotracer gas; regional lung perfusion is measured using intravenous radiolabeled agents. Lung perfusion scans require minimal cooperation and can be used in children of all ages. In infants, the normal distribution of ventilation and perfusion to the right and left lungs is 50:50 %. As the chest wall becomes more oblong transversely, there is a shift to the adult distribution of 55 % right lung:45 % left lung for both ventilation and perfusion [30]. Perfusion is normally well matched to ventilation.

Among children with congenital or infantile scoliosis, lung function asymmetry can be severe with <10 % of function residing in the right or left lung [4]. Twenty of 39 children with EOS studied with lung scans had asymmetric right and left lung distribution of ventilation. Reduced function occurred more often in the lung in the concave chest, but this was not invariable. The degree of lung function asymmetry did not correlate with Cobb angle [4]. The child with TIS due to scoliosis can thus be described as someone with increased work of breathing, with minimal chest wall excursion, and who relies increasingly on one lung as the spine and chest wall deformity progresses.

5.5 Respiratory Muscle Function

The more rigid the chest wall, the less effective are the intercostal and accessory respiratory muscles. Children with TIS become increasingly dependent on function of the diaphragm. Force is generated in the costal region of the diaphragm, which lies parallel to the chest and abdominal walls. The diaphragm attaches at the base of the sternum anteriorly and descends to attach at the level of T8–T12 in the lateral and posterior aspects of the thoraco-abdominal wall [31]. Contractile fibers are normally aligned in the caudad-to-cephalad direction, and shortening moves the diaphragm downward during

inspiration. In normal individuals, diaphragmatic motion also raises the lower ribs, prompting the "bucket handle" motion that increases the thorax in the anterior-posterior and lateral dimensions. This action is lost when the chest wall becomes rigid. The diaphragm is the principal muscle of inspiration accounting for 65 % of inspiratory muscle force generation in upright individuals and more in the supine position [32].

Inspiratory muscle force can be measured non-invasively with inspiratory efforts against a closed system that measures pressure. The greatest inspiratory force is generated when the effort originates at the end of exhalation and is termed the maximum inspiratory pressure (MIP). The invasive way to assess diaphragmatic force more specifically is the trans-diaphragmatic pressure measuring the pressure differential above and below the diaphragm during maximum effort. Data in adults with severe scoliosis have shown that as trans-diaphragmatic pressure falls, vital capacity diminishes and hypercapnic respiratory failure ensues [33]. Normal MIP values are age- and gender dependent with pre-adolescents and girls having lower values [34]. Children with EOS old enough to perform MIP measures have reduced force generation of respiratory muscles that is directly proportional to their respective loss of vital capacity [35]. Causes of reduced respiratory muscle force generation in children without underlying neuromuscular conditions likely relate to the disorientation of the diaphragm fibers, tethering of the muscle at points of attachment, and abnormal insertion of the diaphragm into the abdominal wall [36].

Maximum expiratory pressures (MEPs) are also reduced in children with EOS. Sufficient reduction in MEPs will reduce the effectiveness of the cough reflex and predispose children to retained airway secretions and atelectasis during respiratory infections and post-operatively.

5.6 Pulmonary Hypertension

Respiratory disease that is severe enough to produce respiratory failure can produce pulmonary hypertension and cor pulmonale. These conditions are markers of severe and prolonged hypoxemia, but they can also directly predispose a patient to right heart failure and death. Previous reports of lung pathology in adults with advanced scoliosis have noted the remodeling of pulmonary vessels characteristic of pulmonary hypertension [37]. However, the long-term use of oxygen therapy at home has made pulmonary hypertension a rare complication. Echocardiograms are currently used to assess both right heart morphology (right ventricular hypertrophy) and abnormal septal motion as evidence of chronic pulmonary hypertension. Echocardiograms also provide estimates of pulmonary artery pressures using the velocity of the regurgitant jet from the tricuspid valve. In addition, the echocardiogram identifies unsuspected congenital heart disease that may complicate surgical and medical management of children with TIS. In one report, 26 % of 126 children with congenital spine deformities had associated congenital heart disease [38]. The non-invasive nature of the test and the need for minimal patient cooperation make the echocardiogram easy to use for screening of pulmonary vascular disease and associated cardiac conditions.

5.7 Hypoplastic Thoraces

Chest dimensions are also reduced in children with hypoplastic thoraces. Hypoplasia results in reduced intrathoracic volume due to one of several abnormal structures. In children with Jeune's syndrome (asphyxiating thoracic dystrophy), abnormally short curved ribs lead to circumferential narrowing of the chest wall. In spondylocostal dysostosis and spondylothoracic dysplasia, abnormal ribs number and shape, fused ribs, and vertebral deformities lead to shortened thoracic height and abnormal shape. Reviews of the many syndromes that produce thoracic hypoplasia are published elsewhere [39].

There are few reports of lung function measurements in children with hypoplastic thoraces. Most reviews of Jeune's syndrome report substantial mortality due to hypercapnic respiratory failure in the first 2 years of life. In one series, of

the 118 patients with Jeune's syndrome compiled, 56–80 % of those that died did so at <2years of age [40]. The range of mortality was due to incomplete data on reasons for death in among cases reviewed from the literature. There are no studies depicting the lung mechanics or lung volumes of children with this disorder.

Among children with spondylothoracic dysplasia, there is also a continuum of thoracic cage hypoplasia [41]. In a series of 28 patients, eight (28 %) died during the neonatal period from respiratory failure. Of the survivors, nine were 12–49 years old. Thirteen performed spirometry with vital capacity values ranging from 17 to 51 % of predicted norms. There are no published reports of changes in lung function as a result of surgical interventions in any population of children with hypoplastic thoraces.

5.8 Practical Pulmonary Approach to TIS

The pulmonary evaluation for individual children with TIS depends on how severe respiratory function is impaired on presentation. Table 5.3 lists the clinical and laboratory options for pulmonary evaluation on initial presentation. Children with significant tachypnea, inspiratory work of breathing, and use of abdominal muscles at rest have significant respiratory impairment that ideally is quantified for serial assessments over time and before and after spine interventions. Electrolytes, hematocrit, and oxy-hemoglobin saturation can be assessed regardless of age. Polycythemia and hypercapnia mandate further assessment of gas exchange during sleep and an echocardiogram to rule out pulmonary hypertension.

For children old enough to perform spirometry, assessment of FVC and FEV1/FVC (to measure airway obstruction) is needed to categorize the degree of impairment. If the FEV1/FVC is less than 80 %, then a bronchodilator challenge with pulmonary functions 20 min later will identify likely asthma. If the FEV1/FVC is less than 80%, which is more likely in EOS, then describing the site and nature of central airway narrowing by CT scan or bronchoscopy is an option. Pre-operative risk factors for pulmonary complications in adolescent idiopathic scoliosis triple in frequency when FVC < 40 % predicted using arm span for height. [42] Until data are generated to better depict thresholds of severity for EOS, this seems a reasonable value at which to measure respiratory function in more detail, as described in Table 5.3.

The optimal frequency of monitoring lung function serially is based on the severity of the chest wall and spine deformities, the severity of respiratory impairment on previous evaluations,

Table 5.3 Use of pulmonary functions to initially assess and monitor TIS

Initial evaluation[a]
Clinical features:
Respiratory rate
Retractions (suprasternal, intercostal, subcostal)*
vAsymmetric breath sounds
Abdominal push on exhalation*
SaO_2 in room air
Body mass index (weight/arm span)
Laboratory features
Spirometry (ages ≥5 years):
FVC < 80 % is abnormal
FEV1/FVC < 80 %, then bronchodilator trial and repeat spirometry
Total CO content (electrolytes) when FVC < 40 % and/or *clinical findings
Hemoglobin level (if not on supplemental oxygen or BIPAP at night)
Maximum inspiratory pressure (age ≥7–8 years; <60 cm H_2O is abnormal but values are age dependent)
Echocardiogram when FVC < 40 %
Polysomnogram when FVC < 40 %, polycythemia, or persistently elevated CO content
Lung perfusion scan (left and right contributions as %total) if FVC < 50 % or clinical* findings with asymmetric breath sounds
Thoracic CT scan or bronchoscopy if FEV1/FVC < 60 % with no response to bronchodilator

[a]Assessment every 6–12 months depending on curve progression, new respiratory symptoms, surgical or brace intervention, and degree of abnormality on previous pulmonary evaluation

Repeat polysomnogram, bronchoscopy, bronchodilator response as clinically indicated

*Indicates significant respiratory risk or impairment

and the rate of progression of the spine deformity. In addition, the onset of new respiratory symptoms, such as exercise intolerance, serious intercurrent respiratory infections, or interventions to improve the deformity (e.g., bracing, surgery), should prompt one to assess respiratory function within 1–2 months of the intercurrent event. These recommendations are derived from clinical experience and should not be used as formal guidelines for care. The field of TIS has advanced rapidly, as have the devices used to treat it. The specific utility of each of these tests and thresholds for their use needs prospective study.

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What Can We Learn About Ribs and Vertebra Growth from an Osteological Collection?

6

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Key Points

- This is one of the first studies to use normative data from an osteological collection to make inferences about growth and development of the human pediatric ribs.
- Thirty-two pediatric spine and rib specimens from the Hamann-Todd Osteology Collection of the Cleveland Museum of Natural History were studied.
- There was symmetry and coupled growth between the upper and lower thoracic ribs. The mid-thoracic ribs appeared to grow linearly and as rapidly as that seen in the distal femur.
- The projected rib area appeared to increase in volume by the mathematical representation of a logarithmic spiral, similar to how many rigid biological structures grow in volume.
- The vertebral canal is relatively large in the cervical and lumbar spine compared to the thoracic spine, likely for neural protection and to accommodate for increased motion.
- Unlike other studies that have shown 95 % of the spinal canal area has been completed by age 5 years, our study showed that the canal does not reach 95 % of adult area until age 10 years and continues to increase in area until age 15 years in the thoracic region

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6.1 Introduction

The study of rib, thorax and spine growth is important for a better understanding of the development and function of the child as well as of the many related critical internal structures, including the spinal cord, heart, lungs, abdominal organs and the diaphragm. There are a variety of methods to study the normal growth of these structures. These include animal experimental studies, human longitudinal or cross-sectional anthropomorphic studies [1–3], plain radiography, CT scans [4] and MRI [5]. Each modality provides distinct advantages as well as disadvantages. During the period from 2008 to 2014, we had an opportunity to utilize the Hamann-Todd (HT) Osteology Collection to investigate in great detail the normative growth and development of the pediatric ribs and vertebra in a historical sample of specimens collected from earlier in the twentieth century. This allowed us to make inferences about the unique shape and growth pattern of the pediatric ribs and spine and determine how this growth contributes to the shape and size of the developing pediatric thorax.

The Hamann-Todd (HT) Osteology Collection is housed in the Cleveland Museum of Natural History and contains over 3100 human specimens, including 62 cataloged pediatric specimens aged 1–18 years. Many were depression-era children suffering from malnutrition, and each died prematurely. While environmental factors such as tuberculosis may have had an effect on growth and development for the entire collection, there was no evidence of gross pathologic abnormalities in any of the 62 pediatric specimens. None of the specimens included in the normative data exhibited a spine deformity. The majority of these specimens are over 10 years of age, and unfortunately, there are no 2- or 9-year-old specimens in the collection.

A total of 32 of the most complete specimens representing 1–18 years of age were selected for more detailed study. At least one specimen from each available age group was included to provide a cross-section representation of a growing population. High-quality digital photographs with a background grid provided reliable information. This avoided transcription errors, and any discrepancy and any outlying value could be remeasured directly from the archived photographs (Fig. 6.1).

A total of 6226 high-resolution photographs of ribs and vertebra of the 32 pediatric specimens were obtained. All available ribs and spines were photographed. Ribs were sorted right to left and proximal to distal, labeled as ribs 1 through 12, and were imaged from three orthogonal angles, with the digital camera mounted on separate. A tripod vertebra were arranged from C1 to L5 and photographed in six positions – anterior, posterior, right lateral, left lateral, inferior, and superior. Each photograph was calibrated from a fixed background grid, and quantitative measurements were taken. Scandium Image Analysis Software (Olympus, Soft Imaging Solutions) was used for all measurements, reported to 0.1 mm. The final data set has resulted to date in over 32,000 separate measurements.

There are limitations to using osteology collections in the study of human growth. These include a small sample size, assumption that all specimens are normal, selection bias inherent in using only available specimens (i.e., a bias to poor undernourished children) and cross-section data being used to infer longitudinal normal growth of both individuals and of populations. Other limitations include the following: specimen height and weight were not included in the

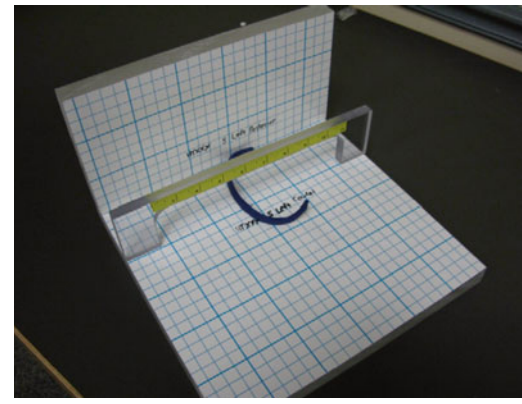


Fig. 6.1 Stationary mount used to photograph the specimens. Shown is a rib model on the grid used to obtain high-resolution photographs in six projections

measurement analysis and the actual chest volume could not be assessed due to inability to measure soft tissue structures. The method used for measurement was subjective, with the points of interest being chosen by the investigators. Test, re-test, inter- and intra-observer reliability was not performed; however, the variability of these lineal measurements in preliminary pilot testing was small.

6.2 Rib Growth

6.2.1 Background

During human evolution of *Homo erectus*, the rib cage transitioned from a triangular funnel shaped to a barrel shaped chest approximately 1.6 million years ago [6]. This is believed to be a result of environmental changes to a more rich and efficient diet that allowed the abdominal organs and waist to narrow. The resulting change in trunk contour is believed for the first time to allow efficient running, which likely provided further evolutionary advantages. There is little detailed information how the triangular-shaped neonatal thorax assumes the barrel shape of adulthood. With increasing age, the chest transitions from a circular to an elliptical cross-section shape with greater transverse width in the mid-thoracic spine at ribs 6 and 7 [7]. Individual ribs have very distinguishing features with the more proximal ribs being flatter and more curved than the straighter lower ribs (Fig. 6.2).

The rib articulates with the thoracic vertebra at the superior lateral aspect of the vertebra and the tip of the transverse process. This provides both stability to the thorax and an axis of rotation for the rib for providing chest motion for respiration (Fig. 6.3). Dimeglio has provided contemporary information on how the chest grows in volume during childhood [1]. The neonate has approximately 6 % of eventual adult chest volume. Although the 10-year-old child is nearing adult height, by maturity, there has been a doubling of chest volume beyond age 10 years. The changing length and shape of the rib create a greater cross-section of the thorax, that when added to the increase in thoracic height, rib separation and costo-sternal growth contribute to overall thoracic volume. Whereas the growth of the long bones is well described, basic rib growth has not [8]. In the human femur, 70 % of the growth occurs distally (approximately 1 cm/year), with 30 % of growth occurring proximally.

Fig. 6.2 Specimen from the Hamann-Todd Collection showing ribs 1–12. Rib 1 (inside in this photograph) is flatter and more curved than the lower ribs (outside ribs). The longest ribs are in the mid-thorax, creating the frontal plane barrel shaped thorax seen beyond infancy

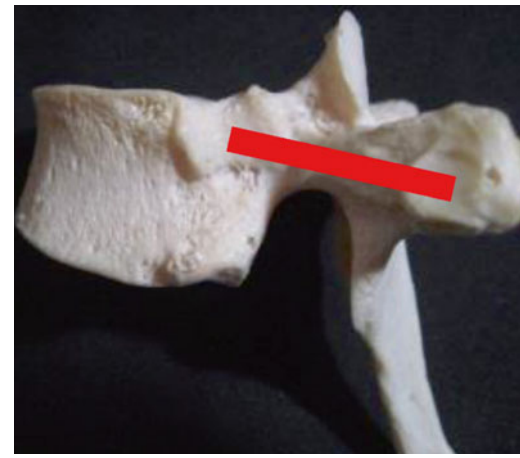


Fig. 6.3 Left lateral view of a mid-thoracic vertebra. The *red line* defines the axis of rib rotation. This axis is relatively more oriented in the coronal plane in the upper thoracic spine and more sagittal oriented in the lower thoracic spine

For the humerus, 80 % of linear growth is proximal and 20 %, distal. Rather than describing normal rib growth, the majority of published rib studies have described abnormalities of rib or thorax growth and asymmetry in children with scoliosis [9–14]. Rib deformities created in experimental animals have demonstrated that abnormal thoracic development is a common feature of scoliosis [15, 16].

Although our study did not look at specific growth in an individual, by looking at a spectrum of growth over the entire period of childhood for a small population, these data can provide some insight into how the individual child's ribs grow. Introducing standardized definitions of the dimensions of the rib has allowed us to describe normative data for how the rib appears to grow during childhood. The area described by the shape of the rib does not describe the total chest volume, which includes the anterior cartilage and sternum, spine, diaphragm, rib spacing and other soft tissues of the thorax. Sandoz et al., in a normative cross-section CT study, showed that the costal cartilage accounted for 45–60 % of rib length, which is a feature of thoracic volume that cannot be studied in an osteology collection [8]. Since we were most interested in the area enclosed by the rib shape, we defined two measurements that represented this space. The outer costal length (OCL) is the total curved length of the rib, and the base diameter (BD) is the linear distance connecting one end of the rib to the other (Fig. 6.4). These two measurements influence the shape and size of the chest and therefore are two of the key parameters in thoracic growth and development. Of the 32 pediatric specimens, 714 total ribs were present and intact for measurement. This allowed us to make inferences about the unique shape and growth pattern of pediatric ribs and determine how the growth of the rib contributes its part to the volume and shape of the developing pediatric thorax.

6.2.2 Linear Rib Growth

Each rib has its own linear growth rate, which more than tripled in length from birth to adulthood. The fastest growing ribs are the mid-thorax ribs, which may influence the triangular shape of the neonatal thorax to become the barrel shape of the older child and adult thorax [9]. There is coupled growth among the upper, middle and lower ribs. Thus, rib pairs number 1 and 12, 2 and 11, 3 and 10 and the

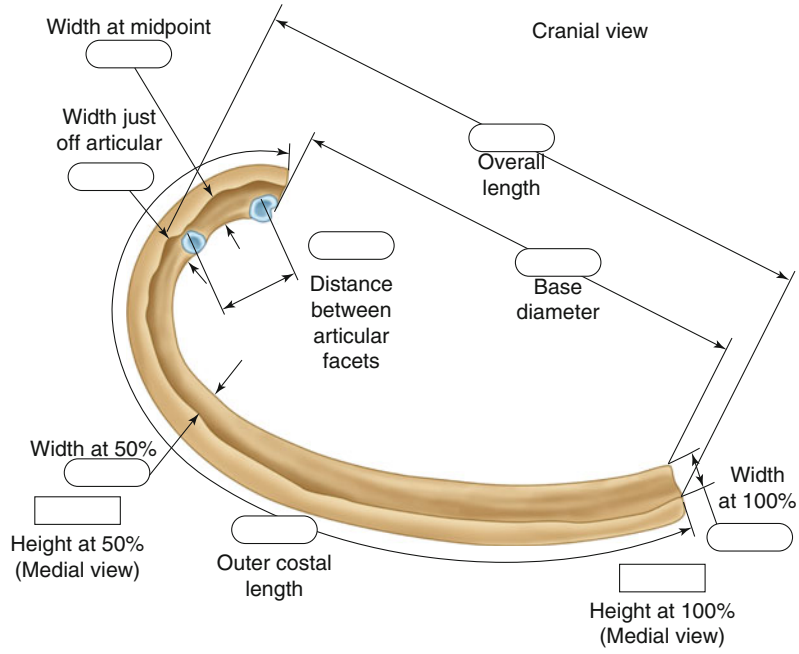


Fig. 6.4 Measurements of the rib outer costal length (*OCL*) and base diameter (*BD*) are illustrated

middle ribs as a group are noted to have the projected area increase with increasing specimen age. The lower ribs where the diaphragm is attached are straighter than the more curved upper ribs. Table 6.1 shows the linear rates of rib growth by vertebral level. The middle ribs increase in length as well as projected area the greatest, similar to the growth rate of the distal femur (Fig. 6.5).

Table 6.1 Growth rate of ribs 1–12, aged 1 year to 18 years

Rib	Starting length (mm)	Growth rate (mm/year)	Correlation coefficient
1	56.7	4.1	0.761
2	86.9	7.5	0.823
3	99.5	9.4	0.869
4	105.0	10.3	0.823
5	112.0	11.0	0.864
6	114.2	10.7	0.854
7	113.9	10.6	0.821
8	109.6	10.5	0.885
9	106.4	9.4	0.842
10	94.5	8.1	0.823
11	69.9	6.1	0.760
12	39.0	4.0	0.575

The middle ribs R4–R8 are growing the fastest. $N = 60$

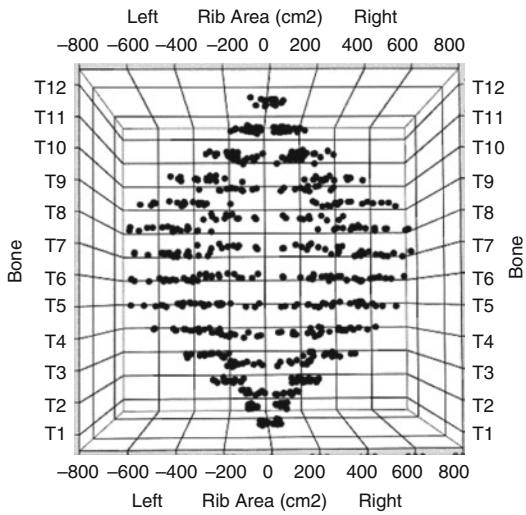


Fig. 6.5 The mid-thoracic ribs increase the greatest in projected area for all ages. This effect is especially notable in the older adolescent, where the barrel-shaped chest is apparent. There is symmetry in projected area between left and right ribs. Younger subjects (aged 1 and 3 years) appear near the center of the graph, while older subjects (aged 17–18 years) are at the far left and right

6.2.3 Coupled Symmetry of Growth

The two physical measurements taken from the ribs for this study (OCL and BD) showed that ribs in these children follow specific patterns of growth. Both the OCL and BD increase with age. An important finding was the linear and volumetric growth and concept of coupled symmetry (Fig. 6.6). The ribs of the upper and lower thorax are shaped very differently, yet they attain the same projected area through different means. The upper thorax contributes to area more by an increase in the OCL, while the lower thorax adds area by an increase in the BD. The lungs are broader at the diaphragm than at the proximal end, and the shape of the ribs helps determine this. The projected rib area greatly increases after age 10 years, reflecting the known doubling of thoracic growth after this age (Fig. 6.7).

6.2.4 Logarithmic Spiral

We observed that the changing curvature of the rib resembles the well-described principle of growth of solid structures in nature, the

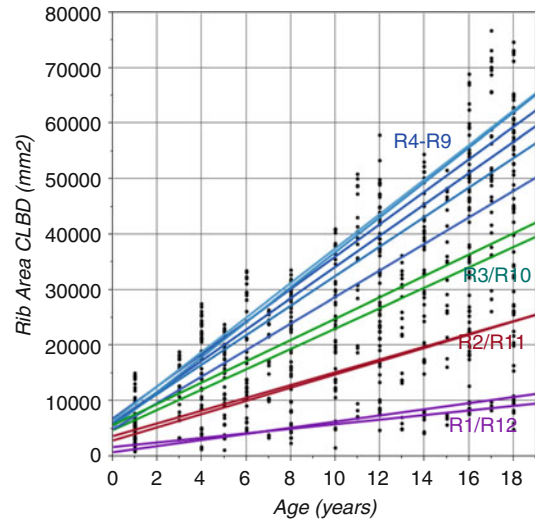


Fig. 6.6 Increasing projected rib area with increasing age. The middle ribs (R4–R9) are increasing the greatest with age. There is coupling of growth between the upper ribs and the lower ribs

logarithmic spiral (Fig. 6.8). This is a geometric principle that is seen throughout nature in animals with shells and is a common mechanism for how rigid organisms increase in volume without changing their basic shape. A logarithmic spiral pattern is also seen in nature in spider webs, low-pressure weather patterns and some interstellar structures such as the Whirlpool Galaxy [17]. Although often described as a golden spiral, these spirals in nature are more accurately logarithmic spirals. Their shape is sometimes approximated by the Fibonacci sequence of numbers, the golden rectangle or the golden ratio. The golden ratio is based on the number phi (1.618) and has a ratio of sides of 1.618 to 1.0. A sampling of over half of the available 714 ribs were evaluated for how closely they followed these spiral forms. The ribs deviated from the expected spiral with an average error of ~8 %. Most of the 1 cm/year of growth of the middle ribs seems to be creating volume by growth of the anterior sternal aspect of the chest, similar to how a nautilus grows. This amount of linear growth of the middle ribs is comparable to that of the distal femur, the fastest linear growing long bone in the human.

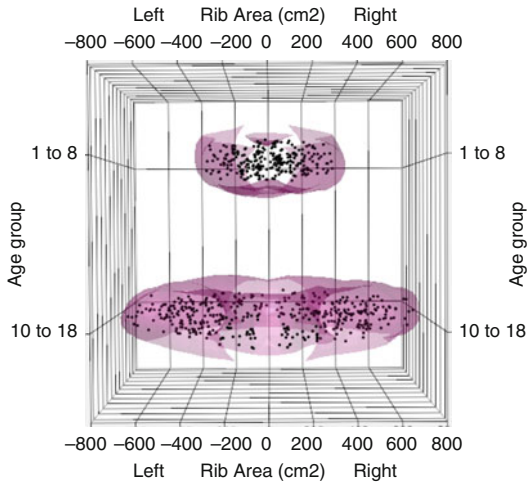


Fig. 6.7 Projected rib area by age group, 1–8 years and 10–18 years. There is a marked increase in projected area after age 10 years, which parallels the known doubling of thoracic volume that occurs after age 10 years

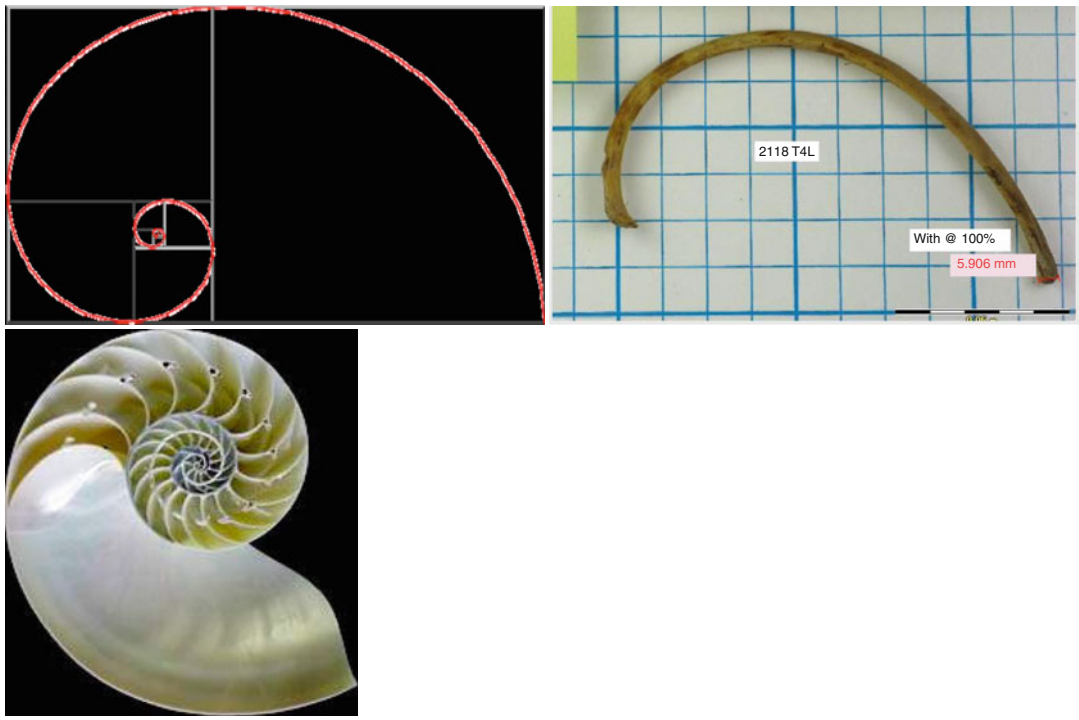


Fig. 6.8 The human rib appears to follow the elliptical spiral structure found in nature with the majority of its growth at the sternal end

Other rib measurements of interest are the rib height and width, which were included in our measurements. We determined that rib height reaches one-half of adult rib height by 1 year of age but still increases in height into adolescence. In contrast, rib thickness reaches close to adult size by about a year of age. This confirmed a CT study that we had previously performed on a normative pediatric population that showed an average rib height of 6.9 mm (one-half adult rib height) by 1 year of age and reaching three-fourths of the adult rib height by 3 years of age. Compared to the growth of the more distal body segments such as the femur, there appears to be early preferential growth of the thorax and ribs. This is particularly important since the upper ribs are frequently used for hook anchor sites. Ferguson et al. [18] showed that the greatest available width of the rib for implant attachment is just lateral to the transverse process: T2 width was 8.4 SD 1.2 mm; T3, 6.3 SD 1.0 mm; T4, 5.6 SD 1.1 mm; T5, 5.0 SD 0.7 mm.

6.3 Vertebra Growth

Initially, we used electronic calipers to obtain morphometric measurements of the vertebral bodies (Fig. 6.9). With time, it became apparent that high-resolution photographs could more quickly provide this information. Six photographic images were obtained for each specimen: anterior, posterior, right lateral left lateral, superior and inferior. Data were plotted for vertebral body width and height with age (Fig. 6.10). The data show linear growth of the vertebral bodies. However, the data are insufficient to determine if this is continual linear growth or is curvilinear growth with different velocities at different ages. By age 1 year, the vertebral width, but not height, is one-half the eventual adult size. Vertebral body width and height increases are greater with more distal levels, and growth continues into late adolescence (Fig. 6.11). Transverse process height and width dimensions were also obtained at levels T1, T4, T7, T10 and L3 for 16 representative specimens. By 1 year of age, these TP dimensions had attained one-half of their eventual adult dimensions, similar to rib height and vertebral body width.

6.4 Spinal Canal Growth

6.4.1 Background

The spinal canal enlarges early in life through endosteal remodeling of the posterior elements and longitudinal growth of the pedicles through the neurocentral synchondrosis (NCS). Osteological studies have traditionally been used to describe normal vertebral canal dimensions and growth [19–22]. Most osteological studies show that the lumbar AP canal diameter reaches adult size very early at approximately 3–5 years of age, whereas transverse diameter continues to grow into adolescence [23]. Porter et al. [23] demonstrated that by 4 years of age, the mean interpedicular diameter was 87 % of adult size and that the vertebra canal area was completely mature. Dimeglio [3] has stated that the spinal canal has reached 95 % of its adult area by 5 years of age. However, there is evidence in osteological collections that inter-pedicular diameter increases until about age 10 years, suggesting that the canal area is also increasing [24]. Environmental factors and health may also contribute to growth inhibition and spinal stenosis for which catch up growth is difficult [22, 24]. Papp et al. [24] also showed that the proximal spine segments mature before the more distal lumbar segments.

Besides the canal becoming wider with growth, the pedicles also increase their transverse width with age. The increase in pedicle size occurs lateral to the spinal canal through remodeling (Fig. 6.12). The implication of this outer pedicle wall remodeling is that this allows the medial pedicle wall to become thicker and more resilient to inadvertent medial pedicle wall implant penetration and also provides for increased canal transverse diameter as the pedicles enlarge. Human and animal studies have shown that unilateral surgical closure of the neurocentral synchondrosis can cause asymmetric growth and scoliosis [25]. However, these procedures do not cause significant spinal canal stenosis, likely due to preserved posterior element appositional growth.

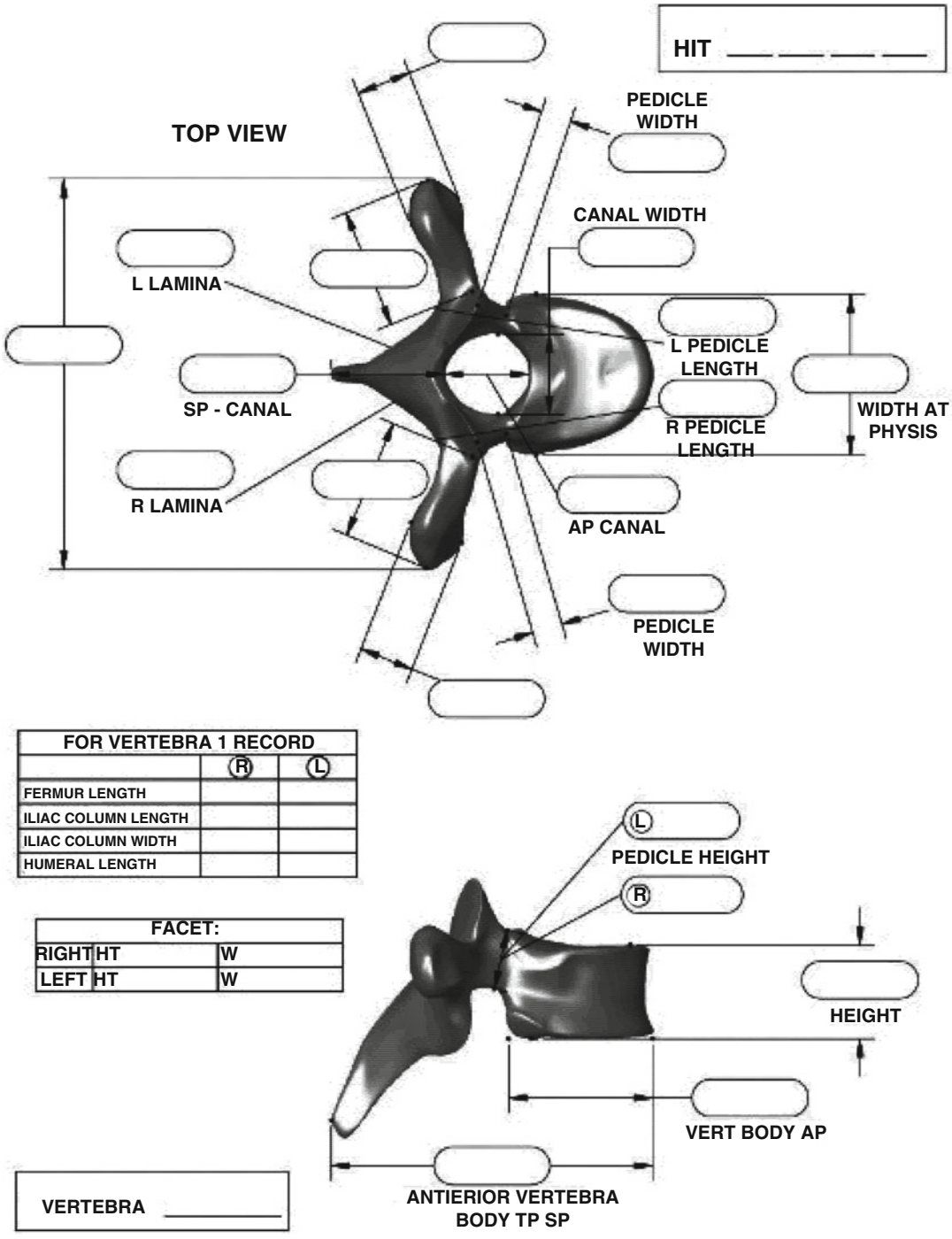


Fig. 6.9 Superior and right lateral views of vertebral body showing the measurements obtained by both calipers and photo imaging

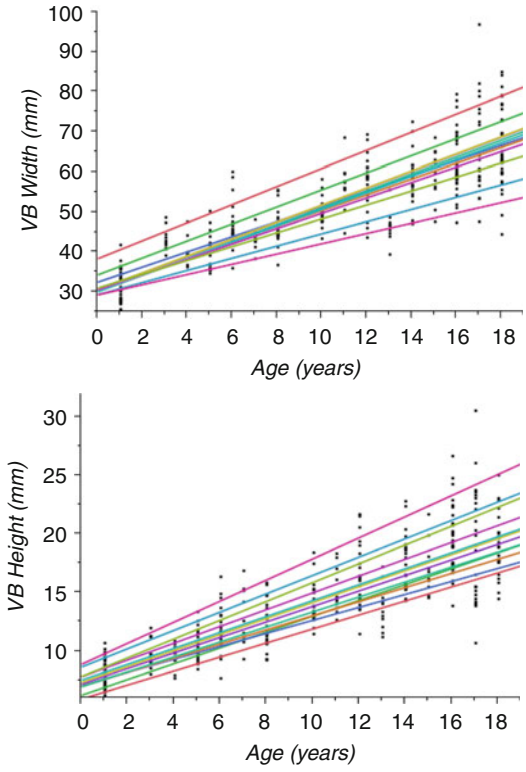


Fig. 6.10 Vertebral body width increases at 1.9–2.1 mm/year $r^2 = 0.78–0.86$. Vertebral body height increases at 0.55–0.90 mm/year, $r^2 = 0.8–0.87$. Each line in each figure represents the linear regression for T1–T12, with T12 the *bottom line* and T1 the *top line*

6.4.2 Canal Width and Area

For the 32 specimens, we noted that the lateral canal width increased with age and also varied by the spine level (Fig. 6.13). We have shown that canal area increases proportional to the lateral canal width but is independent of the AP width (area vs. lateral width $r^2 = 0.68–0.83$ but r^2 is only 0.22 for AP canal width). By age 5 years, the canal was 71 % of its final size, and by age 10 years, 95 %. After age 5 years, the increase in canal area is almost completely through an increase in lateral canal width, not AP width. The growth of the canal area for all vertebral bodies and ages is approximately 6.2 mm²/year. The cervical spine and L5 have the greatest transverse diameter with growth, possibly related to the need for both volume and motion in these areas of the spine. The cervical spine has the most canal width compared to the vertebral body width (Fig. 6.14).

Initially the canal area was calculated based on the equation for an ellipse (Area = Pi*lateral radius*AP radius) (Table 6.2). The Scandium imaging system was able to provide an actual measured canal area. The measured canal area was plotted against the actual canal area with very good correlation $r^2 = 0.852$ (Fig. 6.15). Paired *t* testing showed that the calculated area was very close to the measured area, overestimating the area by only 21 mm². For practical purposes, either the calculated canal area or

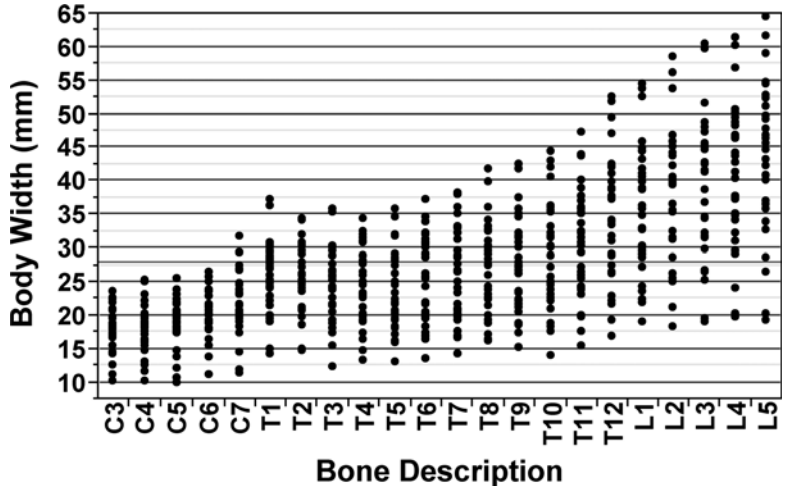


Fig. 6.11 Vertebra body width for each vertebral level. The spread for each vertebral level indicates increasing width with age from 1 to 18 years. C1 and C2 data are not included. Vertebral body increases in width with more distal segments. Younger subjects (aged 1 and 3 years) are at the bottom of the graph and older subjects (aged 17–18 years) are at the top

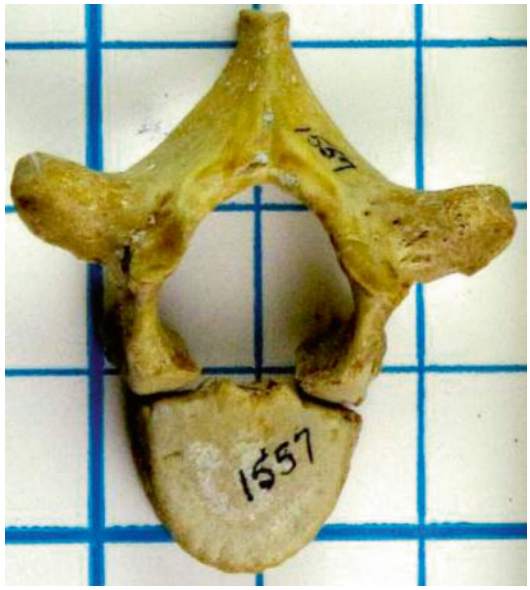


Fig. 6.12 Specimen showing relatively large canal dimensions compared to the width of the vertebral body. The neurocentral synchondrosis (NCS) is still open. Most of the increase in canal area seen into adolescence occurs through increase in the lateral inter-pedicular distance and pedicle remodeling

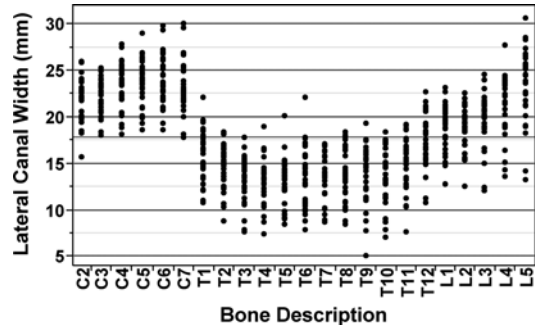
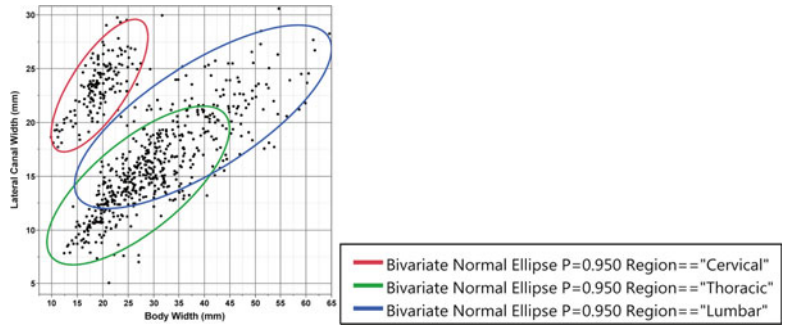


Fig. 6.13 Lateral canal width for each vertebral level. One-way analysis of vertebral canal width. The spread for each vertebral level indicates increasing canal width with age from 1 to 18 years. C1 and C2 data are not included. Canal width is greatest in the cervical and lower lumbar spine. Younger subjects (aged 1 and 3 years) are at the bottom and older subjects (aged 17–18 years) are at the top

Fig. 6.14 Canal width compared to its vertebral body width. Cervical spine canal width is much larger compared to the width of its vertebral body, indicating relatively large canal size



the software measured canal area can be used to determine canal area. Since the AP diameter of the spinal canal does not change with age, the transverse diameter is a surrogate measure for increase in canal area. The canal area is larger in the cervical and thoracic spine (Fig. 6.16).

1–18 years. This indicates that the canal area continues to increase throughout childhood.

C1 canal area is much larger than all other level to accommodate the odontoid and for cervical motion.

6.4.3 Canal Area Increases into Adolescence

Measured canal areas by age groups indicate that the canal continues to increase in area at least up to 10 years of age and possibly beyond. This is apparent when specimens are grouped in 5-year increments (Fig. 6.17) or by two groups of 8-year increments (1–8 years of age vs. 10–18 years of age) (Fig. 6.18). Canal area is larger for the age group older than age 10 years for each region of the spine (cervical, thoracic or lumbar) (Table 6.3).

Table 6.2 Calculated mean canal area for the entire group

Level	*N*	Mean (mm^2)	SD
C1	27	509	122
C2	29	275	58
C3	31	232	39
C4	32	233	43
C5	32	237	44
C6	32	241	44
C7	30	233	42
T1	31	177	44
T2	31	174	42
T3	31	171	42
T4	31	170	45
T5	31	171	47
T6	31	179	49
T7	31	179	49
T8	31	180	48
T9	31	177	48
T10	31	175	41
T11	31	179	47
T12	31	234	56
L1	30	258	46
L2	29	236	52
L3	31	214	49
L4	31	223	66
L5	31	209	57
Cervical[a]	186	241	47
Thoracic	372	180	48
Lumbar	147	228	57

SD for C1 is wide due to skewed data for 1-year-old
Also shown are average canal areas for cervical, thoracic and lumbar regions
[a]Data for 1-year-old are excluded due to difficulty reconstructing with open synchondroses

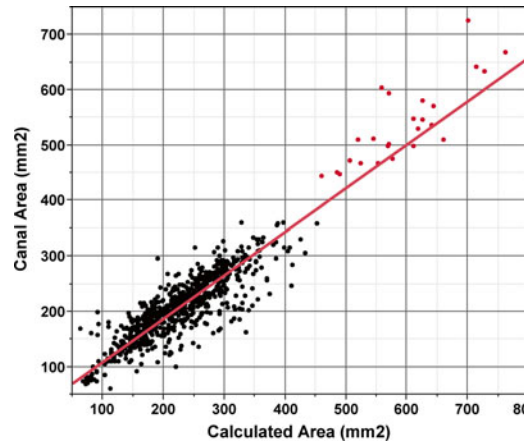


Fig. 6.15 Software measured canal area vs. calculated canal area. $R2 = 0.852$, $N = 732$ observations. Cervical area measurements are plotted in red at the far right and top of the graph

6.5 Neurocentral Synchondrosis in the Growing Vertebra

Development, growth, and closure of the neurocentral synchondrosis (NCS) are essential for proper maturation of the human spine. The NCS is a unique physeal structure, lying at the posterior aspect of the immature vertebral body where it joins the posterior elements, and it is primarily responsible for the growth of the posterior arch [25] (Fig. 6.12). The age of closure in humans has been debated, and the effects of alteration of normal NCS growth are not completely understood. There have been some human cadaveric studies of NCS growth and development including our work with the Hamann-Todd collection as well as animal studies.

Osteological collections can provide unique information about the growth of the spine and ribs in children. Maat et al. [26] conducted a histological and osteological study which included a stillborn infant and dry bone specimens from three skeletally immature individuals. In contrast to the physeal plate of long bones, they noted that the NCS is a bipolar physeal plate, with growth columns present on both sides, with the NCS closing by age 7 years. However, Cañadell et al.'s [27] 1974 study of an osteological collection noted that the NCS closed at 11–14 years of age. Osteologic studies have also documented that pedicle morphology is significantly altered in scoliotic spines, with changes including smaller and dysmorphic concave pedicles, but it does not necessarily follow that this represents a growth disturbance [28]. Huynh et al. [29] performed finite element model analysis of asymmetric pedicle growth rates, both independently and in combination with other deformations (apical rotation or vertebral wedging) in an idiopathic scoliosis model. Based on their results, the authors postulated that asymmetrical growth of the NCS did

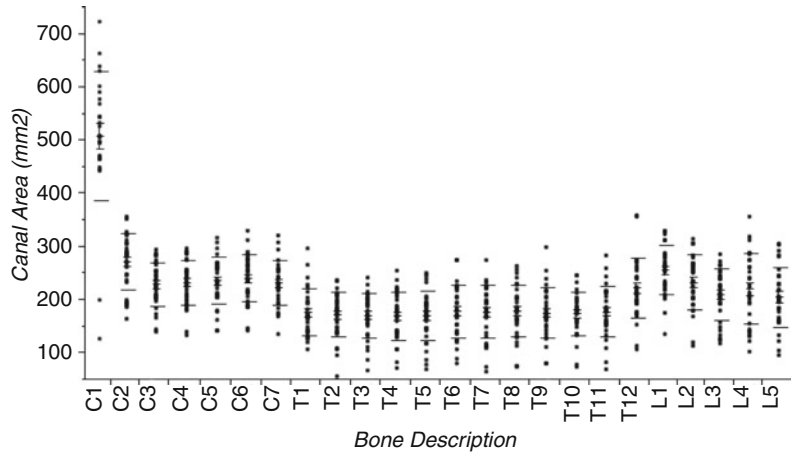


Fig. 6.16 Canal area by vertebral level. Large spread in the areas by level is due to age spread from 1 to 18 years

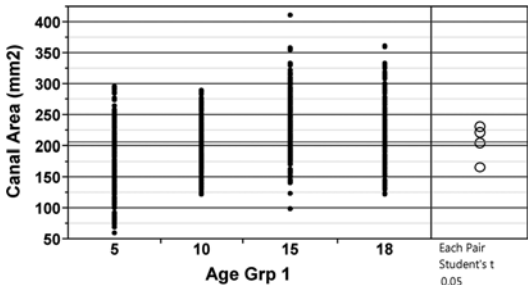


Fig. 6.17 Average measured canal area by 4 age groups: 1–5 years old, 6–10 years old, 11–15 years old, 16–18 years old. C1 data are not included. Average canal continues to enlarge in area beyond age 10 years but reaches adult size by age 15 years. There is no specimen for ages 2 and 9 years. Student *t* test, $p < 0.05$

Table 6.3 Canal area vs. spinal region for younger and older age groups

	Mean ± SD	*N*
Cervical		
1–8 YO	221.9 ± 32.4	62
10–18	282.3 ± 35.0	88
	Thoracic	
1–8 YO	140.8 ± 41.8	152
10–18	208.9 ± 44	203
Lumbar		
1–8 YO	243.7 ± 62.1	58
10–18	301.3 ± 49.5	82

Canal area is uniformly larger in the adolescent compared to the less than 9-year-old age group

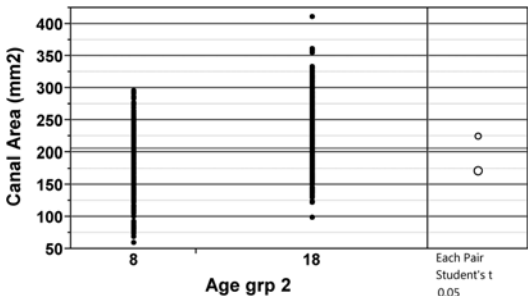


Fig. 6.18 Average measured canal area by 2 age groups. 1–8 years ($N = 250$, mean 172, SD 53) and 10–18 years ($N = 460$, mean 224, SD 50). Student *t* test, $p < 0.05$. There is no specimen for ages 2 and 9 years

not appear to be an etiologic factor in the development of scoliosis. Radiographic imaging has also been used to estimate the age of NCS closure. Yamazaki et al. [30] utilized MRI imaging to calculate the %NCS open and concluded that that the thoracic NCS closed at 11–16 years of age in female and at 12–16 years in male patients. More recently, Zhang et al. [31] assessed the thoracic and lumbar spine neurocentral synchondroses (NCS) with weighted T2 MRI, showing that the lumbar spine NCS is closed by age 10 years, but thoracic spine NCS remains 50 % open at that age.

The manipulation of a growing NCS and associated implications for the growth or distortion of growth of the vertebral body or spinal canal have been explored in both animal and human studies with posterior pedicle screw instrumentation. Cil et al. [32] inserted pedicle unilateral lumbar pedicle screws with and without compression in newborn pigs and demon-

strated significant hemi-canal narrowing (26 % on average) and 5 % pedicle shortening on the instrumented side. Zhang and Sucato [33] were able to produce scoliosis by placing unilateral thoracic pedicle screws across the open NCS in their immature pig model, as well as a direct correlation between greater closure of the neurocentral synchondrosis and greater scoliotic deformity. However, in published reports of children, instrumenting the open NCS will neither cause canal narrowing nor scoliosis, even in the very young [34, 35]. Ruf and Harms [34] described 16 patients aged 1–2 years who received thoracic and lumbar pedicle screws, and in three patients, they were able to assess the vertebral anatomy by MRI and described no canal stenosis or deformity. It is unclear whether those screws were located in the thoracic or lumbar spine. A subsequent small series 1–6 years of age, undergoing 28 hemivertebrectomies and transpedicular instrumentation followed at 3.5 years showed no neurologic complications [35]. The authors believed that spinal stenosis would not develop due to early cessation of spinal growth. Olgun et al. [36] followed 15 children with early-onset scoliosis treated with thoracic or lumbar pedicle screw instrumentation at an average age of 46 months and followed at 2 years with axial imaging. There was growth disturbance indicating that the majority of clinically important growth is concluded by age 5 years.

Manipulation of the growing NCS has also been explored with anterior instrumentation. Elsebaie et al. [37] studied the effect of anterior fixation on the NCS in children ages 21–34 months, with average follow-up of 3 years. CT imaging determined a difference of 10–20 % between the surface areas of the hemicanals at six unspecified levels where the screw heads were passing through or encroaching on the NCS. Zhou et al. [38] used unilateral double pedicle screw fixation across the NCS in the thoracic vertebrae of immature pigs. This resulted in 97 % neurocentral synchondrosis closure with a 20 % decrease in the canal area and a 15 % decrease in the canal depth [38]. Use of unilateral double vertebral body screws across the neurocentral synchondrosis through an anterior approach resulted in 71 % neurocentral synchondrosis closure with a 15 % decrease in the canal area and an 8 % decrease in the canal width.

Assessment of the NCS was included in our study of the Haman-Todd collection. Utilizing the 32 pediatric HT specimens, 1–18 years of age, we evaluated the NCS in all three regions of the spine, including the cervical spine. A total of 733 vertebral body (VB) specimens from C1 to L5 were photographed and were available for analysis. The NCS growth plate attaching the pedicles to the vertebral body was readily detected in the specimens (Fig. 6.12). The length of the NCS was compared to the width of the pedicle at the growth plate: %NCS Open = NCS/pedicle width × 100. Calculations showed that the NCS in the thoracic spine is still partially open at age 16 years, contrary to previous reports of closure by age 10 years [31]. Figure 6.19 shows that during early childhood, the NCS is active and open in all three regions of the spine. By age 5 years, the cervical spine has closed with only 10 % of the NCS visible. The lumbar spine, while closing rapidly, is still nearly 50 % open at age 5 years and is closed by age 10 years. The thoracic spine is only 25 % closed at age 5 years, 80 % closed at age 12 years, 85 % closed at age 16 years and can remain open through age 17 years of age. There was no difference between the left and right NCS data (*t* test = NS). These data are consistent with MRI findings of Zhang et al. [31] and supports the theory that axial plane vertebral growth occurs in the thoracic spine at the age of peak growth velocity, a period of increased risk of deformity progression in scoliosis [39].

6.6 Summary

This is one of the first studies to use normative data from an osteological collection to make inferences about growth and development of the human ribs. Thirty-two pediatric spine and rib specimens from the Hamann-Todd Osteology Collection of the Cleveland Museum

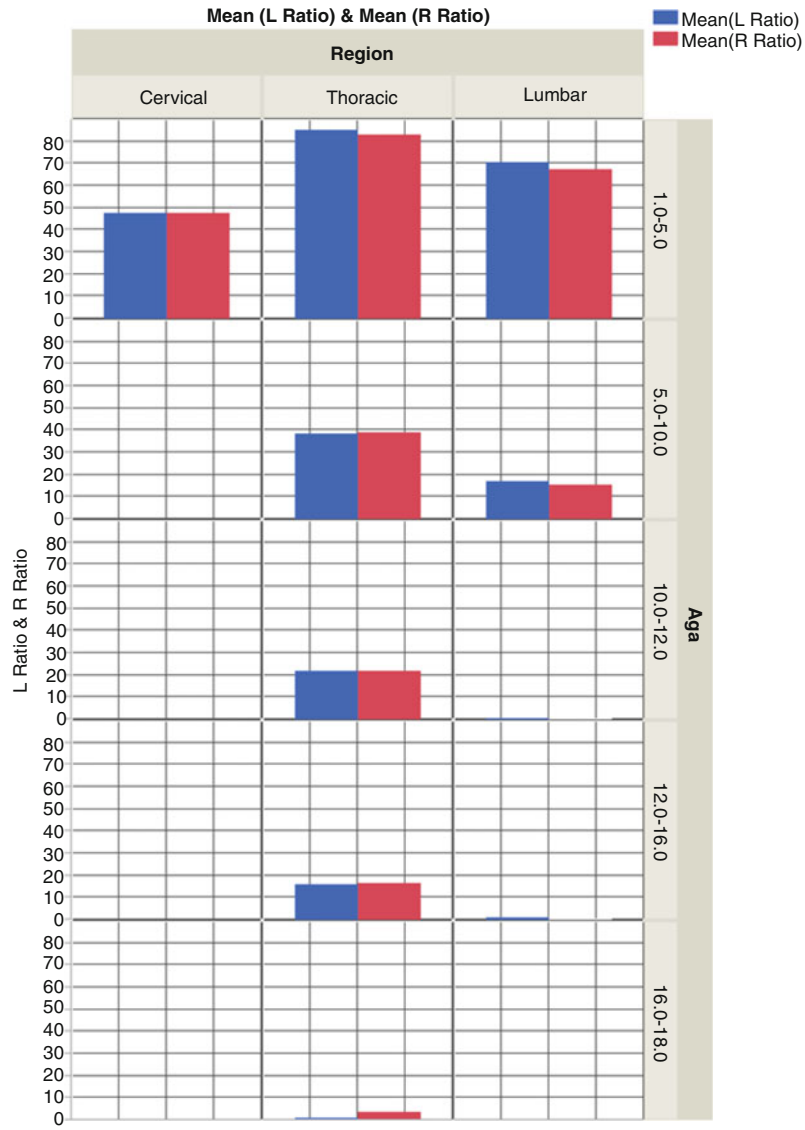


Fig. 6.19 Neurocentral synchondrosis percent open by age group. There is symmetry between the left (*blue*) and right (*red*) NCS. The thoracic NCS remain at least partially open until age 17 years; cervical, until 5 years; and lumbar, until 10 years of age

of Natural History were studied. There was symmetry and coupled growth between the upper and lower thoracic ribs. The mid-thoracic ribs appeared to grow linearly as rapidly as seen in the distal femur and increased in volume resembling the mathematical logarithmic spiral, similar to how other rigid biological structures seen in nature grow in volume. The vertebral canal is relatively large in the cervical and lumbar spine compared to the thoracic spine, likely to accommodate for greater motion. Unlike other studies that have shown 95 % of the spinal canal area has been completed by age 5 years, this study showed that the spinal canal is 95 % of adult cross-section area by age 10 years; however, the spinal canal continues to increase in area up to about age 15 years. Whereas the cervical neurocentral synchondrosis (NCS) closes by age 5 years and the lumbar, by age 10 years, the thoracic spine neurocentral synchondrosis remains at least partially open through age 17 years.

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Part II

Patient Evaluation

Classification of Early-Onset Scoliosis

7

Michael G. Vitale and Evan Trupia

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Key Points

- Early-onset scoliosis is a complex disease that, until this point, has been difficult to classify and therefore difficult to study.
- Classification allows stratification of patients, such that researchers are able to compare outcomes before and after intervention.
- Fifteen experienced surgeons participated in a nominal group technique consisting of iterative rounds of meetings, surveys, and reliability assessments to identify factors most important to managing EOS.
- The C-EOS consists of a continuous age prefix, etiology (congenital or structural, neuromuscular, syndromic, and idiopathic), major curve angle (1, 2, 3, or 4), kyphosis (−, N, or +), and an optional progression modifier (P0, P1, or P2).
- By providing a stratification tool to guide ongoing research, the Classification of Early-Onset Scoliosis aims to improve outcomes for children with EOS.

Substantial portions of this research were previously published in Vitale MG. Development and initial validation of a novel classification system for early-onset scoliosis: Classification of Early-Onset Scoliosis (C-EOS). JBJS 2014;96(16): 1359–1367. With permission from Rockwater, Inc.

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7.1 Introduction

Early-onset scoliosis (EOS) is a complex and heterogeneous condition of considerable diversity with regard to etiology, manifestation, and natural history. Until recently, an integrated and consistent definition of EOS was unavailable. In light of this need, the leadership of the Growing Spine Study Group (GSSG), the Children's Spine Study Group (CSSG, formerly known as the Chest Wall and Spine Deformity Study Group [CWSDSG]), and the Scoliosis Research Society (SRS) collaborated to endorse the definition early-onset scoliosis as "scoliosis with onset less than the age of 10 years, regardless of etiology." Despite this, a more comprehensive system to classify children with EOS remained necessary. In a field with considerable variation in management, the lack of an organizing structure to condense the varied manifestations of EOS further contributed to clinical uncertainty [1–3]. To foster communication and conduct the higher level of evidence studies necessary to establish best practices, reliable stratification of children with EOS is essential [2, 4]. It is with this in mind that the Classification of Early-Onset Scoliosis (C-EOS) was developed.

Left untreated, the natural history of severe EOS is characterized by progressive deformity, cardiopulmonary disease, and early mortality [5]. Recently, in recognition of the relationship between the growing thoracic spine and pulmonary development, the standard of care has shifted away from early fusion and toward more growth-friendly options [6, 7]. Despite a growing body of evidence supporting the safety and efficacy of non-fusion techniques [8–11], there remains controversy over the indications, timing, and techniques that yield the best outcomes for different patients with EOS [1, 2, 4, 12]. Indeed, variability in treatment preferences and collective equipoise among leaders in the field of pediatric spine surgery has been well documented [1–3]. Much of this uncertainty has undoubtedly been rooted in the relative dearth of high level of evidence studies; the available EOS literature consists mainly of Levels III and IV case series and case control studies [4].

It is through prospective, outcomes-based research that the evidence base for the treatment of early-onset scoliosis will be improved. Randomized controlled trials are the gold standard for examining the comparative efficacy of treatment options, and the success of these studies hinges on the control of potentially confounding variables. That is, differences between patients which might impact both treatment decisions and clinical outcomes must be accounted for. The overarching diagnosis of EOS encompasses a great deal of variety with respect to not only etiology and severity of spine deformity but also medical, cognitive, and functional involvement. Without a reliable classification schema, this phenotypic diversity has made it difficult, if not impossible, to draw meaningful comparisons of outcomes before and after treatment.

Classification systems are highly valued instruments employed throughout the orthopedic community. By characterizing the nature of a musculoskeletal condition, accurate classification provides a common language through which surgeons can communicate, guide management, and conduct research [13, 14]. Valid classification also predicts the natural history of a condition or injury such that the results of various treatments can be uniformly reported. This permits the outcomes from different institutions treating the same entity to be reliably compared [14]. Many classifications have already been described for both adult and adolescent idiopathic scoliosis (AIS); however, the inherent complexity of EOS precludes the utility of these systems. In addition, none of these systems lend themselves to the comparison of EOS treatments with or without arthrodesis [15–17]. This is particularly relevant, as surgeons now avoid fusion in the immature spine due to the deleterious effects on cardiopulmonary development. A need thus arose for a comprehensive, practical, and predictive novel classification system designed exclusively for young patients with scoliosis that can adapt to the growing child.

7.2 Development of the Classification of Early-Onset Scoliosis (C-EOS)

Using a model proposed by Audigé et al. [13] for the creation of fracture classifications, a three-phase developmental framework for the creation of a novel classification for patients with EOS was designed (Fig. 7.1). A panel of 15 surgeons from 13 institutions was chosen on the basis of having at least 10 years of experience treating EOS patients, major contributions to the literature, and membership in one of two EOS study groups: the GSSG and CSSG.

7.2.1 Phase 1A: Content Library

A thorough literature review of existing scoliosis classifications identified nine unique systems [15–17, 19–23]. From them, a list of factors important to the management of spinal deformity was compiled and narrowed by the panel through structured discussions and qualitative interviews. A final library of 13 potential EOS variables was assembled (Table 7.1) for evaluation during *Phase 1B*.

7.2.2 Phase 1B: Nominal Group Technique

The nominal group technique is a well-established method of building consensus among professionals in a given area of study. Consisting of iterative rounds of item-rating and group discussion, it has been successfully implemented in multiple medical fields to establish treatment indications and guidelines [24–29]. Using the library generated in *Phase 1A*, participants rated variables on a 3-point Likert scale: not useful, useful, and essential. These results were used to calculate the content validity ratio (CVR), a well-described measure of how essential an item is to the topic under study [30], for each variable to determine group-wide importance.

Using a minimum CVR of 0.51 to satisfy the 5 % level (i.e., exceed chance expectation) [30],

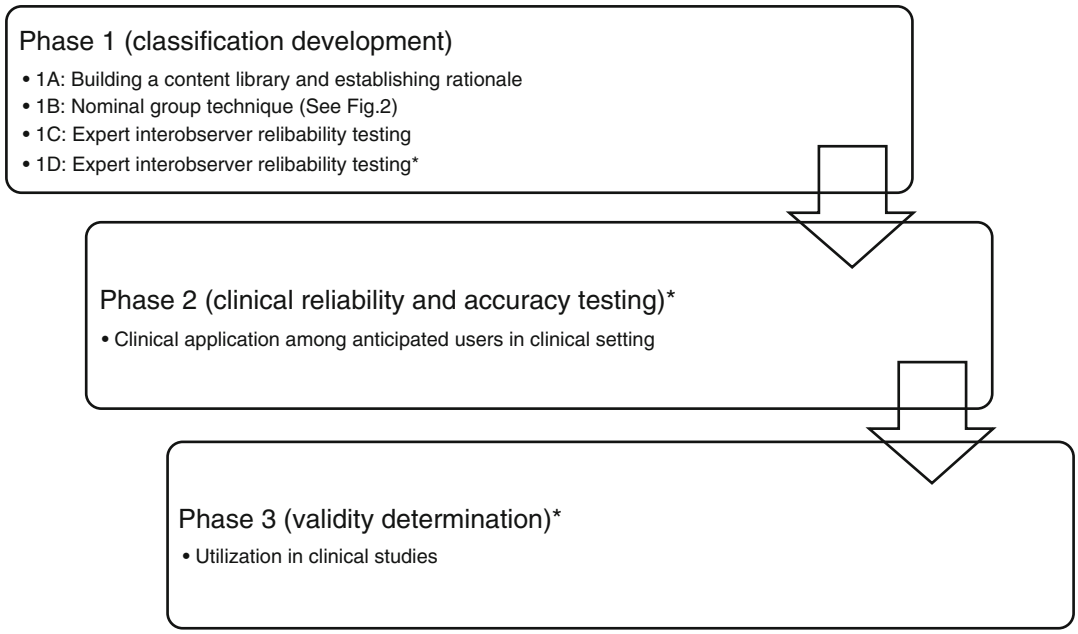


Fig. 7.1 Developmental framework and validation of the C-EOS: validation model. Phases 1A, 1B, and 1C are completed. *The remaining phases will be described in future studies (Reprinted from Vitale [18]. With permission from Rockwater, Inc.)

Table 7.1 Variable content validity rankings: participant ratings of the 13 proposed variables included on the primary survey on a 3-point Likert scale used to assess content validity as proposed by Lawshe [24]

Variable	Not useful	Useful	Essential	CVR
Cobb angle	0	1	13	0.86
Etiology	0	3	11	0.57
Kyphosis	0	3	11	0.57
Age	5	0	9	0.29
Progression	3	5	6	−0.14
Curve flexibility	3	6	5	−0.29
Chest wall abnormalities	2	8	4	−0.43
Other co-morbidities	3	8	3	−0.57
Pulmonary function	3	8	3	−0.57
Nutritional status	5	7	2	−0.71
Ambulatory ability	2	11	1	−0.86
Mental function	9	5	0	−1.00
Bone quality	10	4	0	−1.00

Based on data from Ref. [24]

the variables with significant CVRs on the primary survey were major curve angle (0.86), etiology (0.57), and kyphosis (0.57) (see Table 7.1). Although age had the fourth highest-ranking CVR (0.29), it did not meet the CVR significance threshold. Further discussions determined that because age carries import implications regarding patient management and outcomes, it would still be included as a continuous classification prefix. Curve progression and flexibility also did not meet the content validity cut-off point but were later deemed important to decision-making nonetheless. They were instead considered as optional modifiers to be included at a provider's discretion. Curve flexibility was later discarded.

The preliminary classification underwent multiple rounds of modifications based on e-mail discussions, in-person meetings, and a secondary survey. Participants reconvened and reached consensus regarding final modifications, variable subgroups, and cut-points. The Classification of Early-Onset Scoliosis (C-EOS), consisting of a continuous prefix (age), three core variables (etiology, major curve angle, and kyphosis), and an optional modifier (curve progression), is illustrated in Fig. 7.2. A case example of the C-EOS in practice is shown in Fig. 7.3.

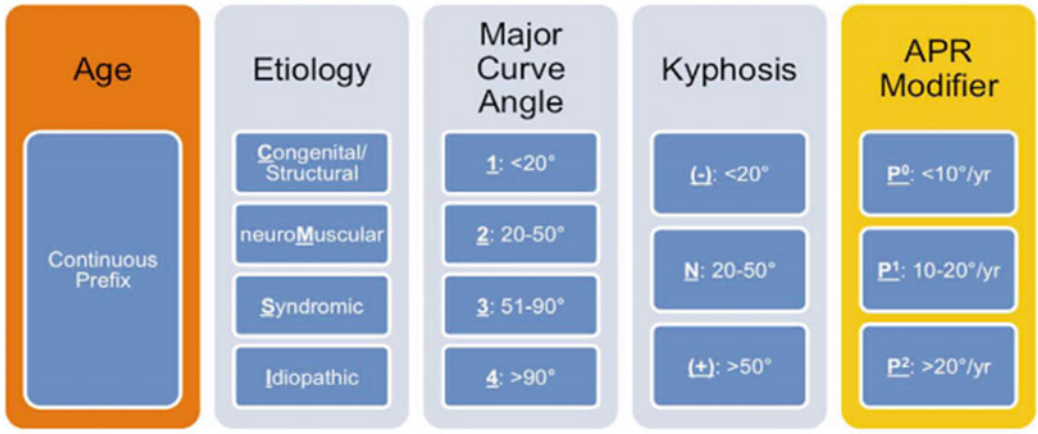
7.2.2.1 Age

There is no question that patient age at the time of evaluation carries important implications regarding treatment decision-making and prognosis. Several different age-based groupings were proposed during the iterative consensus-based process; however, agreement for any single system was lacking. It was clear that the perceived importance of where to draw the "line in the sand" varied greatly among participants. Because of this, the group decided to include age in the C-EOS as a *continuous prefix*. Future efforts to provide meaningful subgroup structuring for the age prefix are still encouraged.

7.2.2.2 Etiology

The impact of etiology on the natural history and management of EOS cannot be understated. The severity of scoliosis and response to treatment is often determined by factors very specific to a particular diagnosis [6, 31–34]. For example, multiple studies have shown derotational casting to be a potentially curative intervention for infantile idiopathic scoliosis, while its utility for non-idiopaths remains in question [35–38].

When finalizing etiologic subgroups, priority was given to pairing those etiologies which are managed most similarly. To improve clarity in cases of unclear or mixed etiology, three final modifications were made. First, neuromuscular patients with abnormally high or low tone were collapsed into a single group to remove any ambiguity. Second, a number of commonly encountered conditions were assigned and catalogued as belonging to a particular etiology (see Fig. 7.2). Third, in cases of mixed etiology, an order of priority was assigned which, from highest to lowest, is congenital/structural (C), neuromuscular (M), syndromic (S), and idiopathic (I). Patients with multiple disease states are then assigned to the subgroup of highest priority.



Age: The age of the patient should be included as a continuous classification prefix (e.g. 3yo).

Etiology: Etiologies are listed in prioritized order from highest to lowest. When etiology is mixed and/or unclear, etiologic assignment should be made starting from the top of the list.

- **Congenital/Structural:** Curves developing due to a structural abnormality or asymmetry of the spine and/or thoracic cavity
  - *Hemivertebrae, fused ribs, post-thoracotomy, thoracogenic, iatrogenic (post-thoracotomy), tumor (pre or post resection), amniotic band syndrome, hemihypertrophy, NF (dysplastic type), congenital diaphragmatic hernia, congenital heart defect (s/p repair), Proteus syndrome, Jeune syndrome, constrictive chest wall syndrome, Jarcho-Levin Syndrome, Spondylothoracic dysplasia, Spondylocostal dysplasia, VATER/VACTERL*
- **Neuromuscular:** Curves without congenital or structural abnormalities in which the deformation is primarily attributable to a neuromuscular abnormality of high or low tone
  - *Flaccid spinal cord injury, spinal muscular atrophy, muscular dystrophy, spina bifida, low tone CP, Freidrich's Ataxia, familial dysautonomia, syringomyelia, Charcot-Marie-Tooth syndrome, CHARGE Syndrome, Spastic CP, spastic spinal cord injury, Rett Syndrome*
- **Syndromic:** Syndromes with known or possible association with scoliosis that are not primarily related to congenital/structural or neuromuscular etiology
  - *Spinal dysraphism, Ehlers danlos (and other connective tissue disorders), Prader-Willi syndrome, Marfan syndrome, Achrondroplasia, Arthrogryposis, Diastrophic dysplasia, Ellis Van Creveld, Neurofibromatosis, Osteogenesis Imperfecta, Spondyloepiphyseal dysplasia, Down's Syndrome, Goldenhar, Klippel-Fiel*
- **Idiopathic:** No clear causal agent (can include children with a significant co-morbidity that has no defined association with scoliosis)

All radiographic assessments should be posteroanterior and performed in the most gravity dependent position possible for patient (i.e. standing preferred to sitting preferred to supine).

Major Curve: Measurement of major spinal curve in position of most gravity

Kyphosis: Maximum measurable kyphosis between any two levels

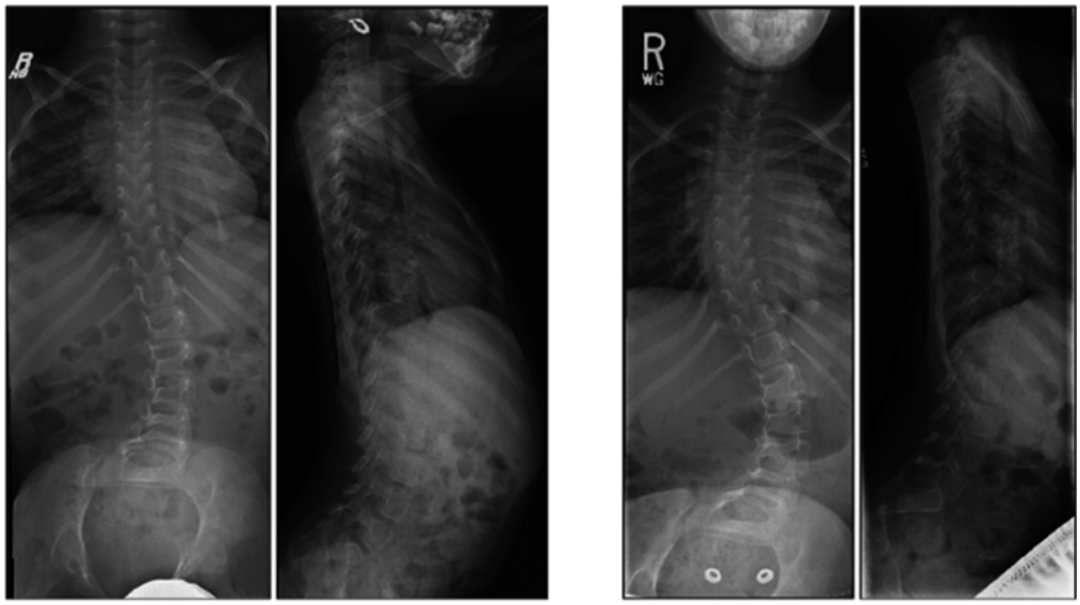
Annual Progression Ratio (APR) Modifier (optional): Progression calculations should be made with two separate clinical evaluations at times t_1 and t_2 that are spaced a minimum of 6 months apart

$$APR = (\text{Major Curve @ } t_2) - (\text{Major Curve @ } t_1) \times \frac{12 \text{ months}}{[t_2 - t_1]}$$

Fig. 7.2 The Classification of Early-Onset Scoliosis (C-EOS) (Reprinted from Vitale [18]. With permission from Rockwater, Inc.)

7.2.2.3 Major Curve Angle

Subgroup cut-points were initially based on previously described ranges of major curve angles for guiding the management of scoliosis [12]. Modifications were made after group discussion during the nominal group technique. Consensus was reached for the following major curve subgroupings: Group 1 as <20°, Group 2 ranging from 20° to 50°, Group 3 from 51° to 90°, and Group 4 as >90°. Future studies utilizing the C-EOS will test the hypothesis that these subgroups will correlate with outcomes of various treatment strategies.



This is a six year old female with a history of merosin-deficient muscular dystrophy. On initial presentation (Left), seated films revealed a major curve of 18 degrees with 7 degrees of kyphosis. On follow-up 14 months later (Right), repeat x-rays showed progression of the major curve to 33 degrees with 2 degrees of kyphosis. Muscular dystrophy is a neuromuscular etiology. The patient is hypokyphotic and would be assigned a P^1 progression modifier based on the following calculations:

APR = [33 degrees – 18 degrees] X (12/14)
≈ 13 degrees/yr

Classification of this patient is **M1-** at first visit and **M2-P^1** at second visit.

Fig. 7.3 Sample case: use of the C-EOS in a sample case presentation (Reprinted from Vitale [18]. With permission from Rockwater, Inc.)

7.2.2.4 Kyphosis

The C-EOS defines kyphosis as the highest measurable sagittal Cobb angle between any two levels. This is in contrast to the classification system for AIS developed by Lenke et al. [16] and reflects the observation that kyphosis often extends into the lumbar spine in patients with EOS. Normative data from the available literature suggest that the pediatric measures of kyphosis are different from adults' and generally increase with age [39]. With this in mind, the normokyphotic (*N*) range was selected to be 20–50° [40], with hypokyphosis (–) and hyperkyphosis (+) falling on either side. Of note, hyperkyphosis has recently been studied as a metric of operative importance in EOS patients undergoing growing-rod surgery [41]. Ongoing validation studies will investigate the impact of kyphosis as it fits in the C-EOS schema and the effect on treatment outcomes.

Curve Flexibility

Despite the importance of curve flexibility in clinical decision-making, there remains considerable variation in the flexibility imaging techniques between institutions. At many centers, flexibility imaging is not routinely performed throughout the course of care in children with EOS. In addition, most imaging is both effort and gravity-dependent, and depending on the child's physical and cognitive involvement, he or she

may be unable to achieve a proper flexibility assessment. For these reasons and other barriers to widespread and reliable calculation, curve flexibility was ultimately excluded from the C-EOS.

7.2.2.5 Curve Progression

The clinical significance of the velocity of curve progression is well documented in the literature [3, 6, 35, 42]. Among patients with congenital EOS, McMaster et al. [31] determined that the rate of curve progression depends on anomaly type and location and that the highest risk occurs during periods of rapid growth (e.g., the first 2–3 years of life, puberty). Rodillo et al. [33] showed that rates of progression in children with spinal muscular atrophy (SMA) ranged from 5° to 15°/year, depending on disease type, relation to puberty, and ambulatory status (e.g., increased rate after loss of ambulation). In young children with idiopathic scoliosis, Scott et al. demonstrated that although a curve can be stable for years, when it starts to progress, it will do so in a fairly constant fashion. They believed that the end result depends primarily on the ages at which progression begins and growth finishes and that any variation from a steady rate of 5°/year was likely due to flexibility or measurement error [43].

Unfortunately, the extensive variability among providers in the reporting of curve progression presents an obstacle to reliable assessment. That is, the meaning of "the curve has progressed 15° since the last visit" cannot be scaled unless the duration of time between the visits is duly reported. To control for any potential inconsistency, a simplified annual progression ratio was developed to standardize the calculation of curve progression (see Fig. 7.2). A minimum of 6 months of follow-up between points is required for inclusion. Future efforts will aim to characterize definable patterns in the rates and timing of change in the annual progression ratio, utilize the C-EOS to identify children with inherently higher risk of accelerated decompensation, and examine the rate of progression as it contributes to the C-EOS in predicting treatment outcomes.

7.3 Reliability and Validity of the C-EOS

For a classification system to be successfully adopted in a given medical field, it must be both reliable and valid. Reliability generally refers to the inter- and intraobserver reliability of a classification schema and is a measure of precision. Validity is a measure of accuracy and ensures that a given system characterizes the true pathologic process [14].

7.3.1 Phase 1C: Expert Interobserver Reliability Testing

Kappa coefficients are measures of statistical agreement within or between observers [44]. Initial exploration of the C-EOS revealed substantial to excellent interobserver kappa values for etiology, major curve angle, and kyphosis. Interobserver reliability for the calculation of the annual progression ratio was moderate, which was surprising given the high reliability of its component variables (major curve angle, time). It is suspected that this is a reflection of rough estimation, rather than strict adherence to the formula among participants – which will be emphasized in the future.

7.3.2 Phases 1D, 2, 3

Studies examining the intraobserver reliability, clinical reliability, and validity are either under construction, currently enrolling, or in various stages of manuscript completion and submission. Among them is a reliability study of the C-EOS in concert with a novel schema for classifying complications in children with EOS developed by Smith et al. [45] Another will utilize the C-EOS to identify groups of patients with inherently higher risk of complications following growing instrumentation surgery.

7.4 A Research Initiative

Beyond simply facilitating communication in the clinical setting, validated classification systems are a critical component of the research process. Reliable stratification of study subjects is the foundation for establishing evidence-based treatment guidelines. Much in the way the classification systems for adult scoliosis were borne out of advances in treatment [46], the development and implementation of the C-EOS were the logical next step to improve outcomes among children with scoliosis. Building off of work to identify areas of clinical equipoise among treating surgeons [3], the structure and common language provided by the C-EOS will provide a scaffold around which future clinical trials of treatment modalities can be built. By fostering a collaborative environment in all areas both clinical and academic, the C-EOS will ultimately lead to improved care for children with early-onset scoliosis.

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8 Clinical Examination and Associated Comorbidities of Early Onset Scoliosis

Hazem B. Elsebaie and Jeff Pawelek

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Key Points

- The general clinical examination of a child with early onset scoliosis is of paramount importance and should be carefully noted in the medical record.
- Birth, family and medical history should be well understood and documented prior to initiating treatment.
- Associated other systems abnormalities need to be searched for individually with reference to their occurrence in different types of scoliosis.
- Proper dealing with detected comorbidities can avoid potential problems that might affect and jeopardize the results of management.
- Diagnostic laboratory tests, imaging, and pulmonary function tests when feasible are crucial in developing a comprehensive treatment plan.

8.1 Introduction

The clinical examination of a child with early onset scoliosis (EOS) begins with a thorough medical history, which should include information specific to the spinal deformity such as age of onset, history of progression, and previous nonoperative and operative treatment. All previous

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spine-related diagnostic imaging should also be reviewed, if available. Examination of the neural axis, pulmonary, cardiovascular, urogenital, musculoskeletal, gastrointestinal, cutaneous, and other systems that warrant evaluation should be performed to identify and assess all spinal and non-spinal comorbidities. The overall health and nutrition of the child must be examined with special consideration of the child's height, weight, and dietary regimen. If there is any indication that these systems are abnormal, further workup should be pursued with the appropriate subspecialists. All information obtained from the initial clinical evaluation should be carefully noted in the medical record for future reference for the treating physician and other practitioners involved with the child's care.

8.2 Medical History

8.2.1 Birth History

The child's medical history should be well-understood and documented prior to initiating treatment. Significant facts regarding the birth of the child, including neurologic and musculoskeletal abnormalities, should be noted. The Apgar score (activity, pulse, grimace, appearance, respiration) may reveal important information related to the child's primary diagnosis and secondary musculoskeletal abnormalities (e.g., clubfoot, loss of limb) by assessing muscle tone, heart rate, reflex irritability, skin coloration, and ability to breath immediately after childbirth [1]. Additional results from physical examinations of an infant may be of special interest in identifying possible neurologic causes of the spinal deformity.

8.2.2 Family History

Spinal deformities that exist within the child's family may increase the risk of progression of the child's deformity and should be considered when determining when to initiate treatment and which treatment would be most appropriate [2]. Syndromes, neuropathies, and myopathies that are present in immediate family members may also provide clues for identifying the etiologic diagnosis and possible associated comorbidities.

8.2.3 Spinal Deformity History

Previous spinal deformity treatments, including Risser casting, bracing, and surgery, should be carefully reviewed before planning new treatment. Historical details of each treatment should be noted, including prior hospitalizations and complications. The age of onset and initial magnitude of the spinal deformity are important to assess the rate of progression [3].

8.3 Review of Systems

The spinal deformity and other abnormalities can share the same origin, have a cause-and-effect relationship, or have no clear link. To add further complexity, the timing of presentation for spinal and non-spinal anomalies may not be synchronous. The child's overall health, with particular attention to nutrition and pulmonary status, is an important factor if surgical treatment is being considered. Current medications and known allergies should be documented, and necessary pharmaceutical modifications should be made prior to initiating treatment. History of pneumonia, dysphagia, or recurrent infection must be accounted for particularly when considering surgical intervention.

8.3.1 Comorbidities

The most common and clinically relevant comorbidities associated with various etiologies of spinal deformity will be covered in the following section. Associated comorbidities will be reviewed individually with reference to their occurrence in different types of scoliosis under the following headings:

1. Neural axis and neurological status
2. Cardiac

3. Urogenital
4. Musculoskeletal
5. Gastrointestinal
6. Cutaneous
7. Mental status, disability, and pain

8.3.1.1 Neural Axis and Neurological Status

The treating physician must be aware that a spinal deformity in a growing child can be the presenting sign of an asymptomatic neural axis abnormality. These abnormalities include Arnold–Chiari malformation, syringomyelia, hydromyelia, low-lying conus, tethered cord, and tumors. If these neural axis abnormalities remain undetected, there is a risk of undue neurological sequelae resulting from the use of instrumentation for correction of the scoliosis [4, 5]. Detailed neurological examination of scoliosis should consist of an evaluation of the motor, sensory, and reflex function of the upper and lower extremities as well as an evaluation of abnormal neurologic signs such as sustained hyperactive reflex, unilateral superficial abdominal reflex, muscle atrophy, motor weakness, sensory loss, and sometimes elicited gag reflex. Abnormal abdominal reflexes may suggest the presence of an intraspinal disorder [6]. History of complaint of severe headache, backache, and the presence of neurologic symptoms should be noted. The child's pre- and posttreatment ambulatory status should also be monitored to identify significant changes.

Idiopathic Scoliosis

For infantile and juvenile idiopathic scoliosis with curves greater than 20°, previous literature has demonstrated an approximately 20 % (range, 17.6–26 %) prevalence of neural axis abnormalities [7–9]; of additional concern is the reported need for neurosurgical intervention between the time of birth and 10 years of age in more than 50 % of idiopathic patients who have neural axis abnormalities on magnetic resonance imaging (MRI). Screening by total spine MRI examination in these children is recommended at the time of presentation in early onset scoliosis with curves greater than 20° even if the findings of neurological examination are normal and despite the fact that many children at that age may need intravenous sedation or general anesthesia [7]. Once neural axis abnormality is detected, neurosurgical consultation is mandatory for evaluation, treatment, follow-up, or possible intervention usually in the form of decompression of the posterior fossa, surgical decompression and/or shunting of a syrinx, and cord untethering.

The association between "idiopathic" scoliosis in all age groups and craniovertebral abnormalities has been well established. It has been reported that the mean position of the cerebellar tonsils in patients with "idiopathic" scoliosis was 4 mm below the foramen magnum and that 50 % of patients with "idiopathic" scoliosis had cerebellar tonsils below the foramen magnum [10]. With the development of MRI, neural axis abnormalities such as syringomyelia or Chiari malformations, tonsillar ectopia, and low conus-medullaris are increasingly being found in patients with asymptomatic "idiopathic" scoliosis [11].

At 10 years of age and older, the routine use of MRI in patients with idiopathic scoliosis before surgery remains controversial; the classical guidelines for MRI screening in scoliosis are valuable; and the proposed indications for ordering MRI in the literature include neurologic deficits, infantile and juvenile onset, male gender, abnormal sagittal profile of the spine, atypical curve pattern (left-sided curve), rapid curve progression, and the presence of pain [11]. Advocates of a safer routine MRI point out that a higher risk of neurologic complications has been reported during surgery of scoliosis associated with syringomyelia; in addition, to prevent potential neurologic complications [12], neural axis malformations need to be addressed before the treatment of scoliosis; therefore, every attempt should be made to identify these patients clinically or by MRI [4, 13]. A selective approach is advised by some investigators suggesting that MRI study is not necessary for a neurologically intact patient with "idiopathic scoliosis"; however, this might entail little risk of neurologic complications as a result of scoliosis surgery if

these patients have a neural axis malformation on MRI [14–16].

Congenital Scoliosis

Congenital scoliosis is often associated with intraspinal abnormalities. The embryonic development of vertebrae is closely related with that of the spinal cord and the organs of the mesoderm [17]. An incidence of intraspinal anomalies between 30 and 38 % is reported in association with congenital spinal deformities when using total spine MRI for the assessment of these patients [18–20]. The intraspinal anomalies can cause progressive neural loss with growth and curve progression. In addition, they greatly increase the risk of neurologic injury during surgical correction of the deformity.

Neurocutaneous stigmas and neurological findings of intraspinal lesions can appear from history or examination; these include hairy patches (Fig. 8.1) and pigmentation overlying the spine, bladder symptoms, paraesthesia in one leg, foot deformity, obvious wasting of one leg, asymmetrical abdominal reflexes, and abnormality of posterior column sensation. However, the presence of neurocutaneous stigmas is not a reliable indicator of intraspinal abnormality [11, 19, 21, 22].

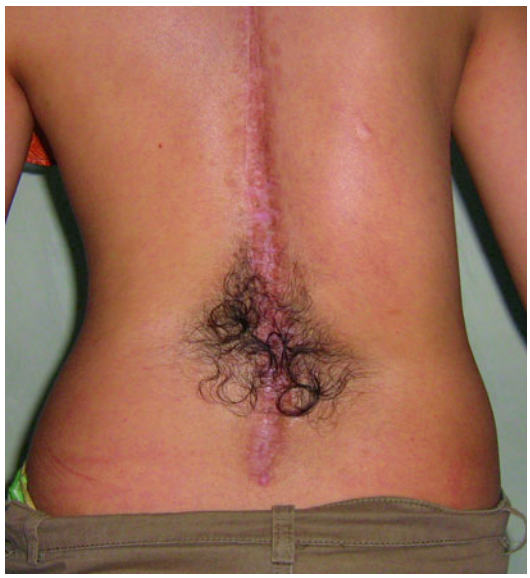

Fig. 8.1 A hairy patch found in the back of a child with congenital spinal deformity; it can be associated with occult spinal dysraphism. The presence of neurocutaneous stigmas is an unreliable indicator of intraspinal abnormality

Tethered spinal cord (Fig. 8.2a) is the most common MRI-identified intraspinal anomaly in congenital spinal deformity in many reports; syrinx (Fig. 8.2b) is the second; then thickened and fatty filum, low conus, diastematomyelia (Fig. 8.2c), intradural mass/lipoma, extradural mass, Chiari malformation arachnoid cyst, and Dandy–Walker malformation [18, 19, 23].

Neurologic deficit caused by tethered spinal cord in congenital spinal deformity may not be manifested in very young children, and in older ages, there is lack of clear association between intraspinal anomalies and detectable clinical manifestations; therefore, MRI is generally recommended in the evaluation of patients with congenital spinal deformity even in the absence of clinical findings [23]. The performance of MRI in young children, especially 5 years of age or less, involves administration of sedation or general anesthesia, with the attendant risks of respiratory complications in these children who already may suffer from pulmonary compromise. As a result, a selective approach probably is wise. An MRI scan must be obtained in older age groups before surgical correction of the spinal deformity in cases with established or developing neurologic signs and probably also in cases with progressive deformity, in which surgery is to be considered sooner, but for younger children, MRI with the patient under general anesthesia is to be considered only if surgery is imminent or neurologic signs develop [18].

Marfan Syndrome

Dural ectasia is ballooning or widening of the dural sac, fibrillin deficiency resulting in connective tissue abnormality, and weakness in the dural sac has been suggested as the cause for duralectasia in Marfan syndrome. It usually occurs in the most caudal portion of the lumbosacral spinal column, at the point of greatest cerebrospinal fluid pressure in the upright patient. The neural symptoms are thought to be related to stretching and traction mechanisms, which may be clinically

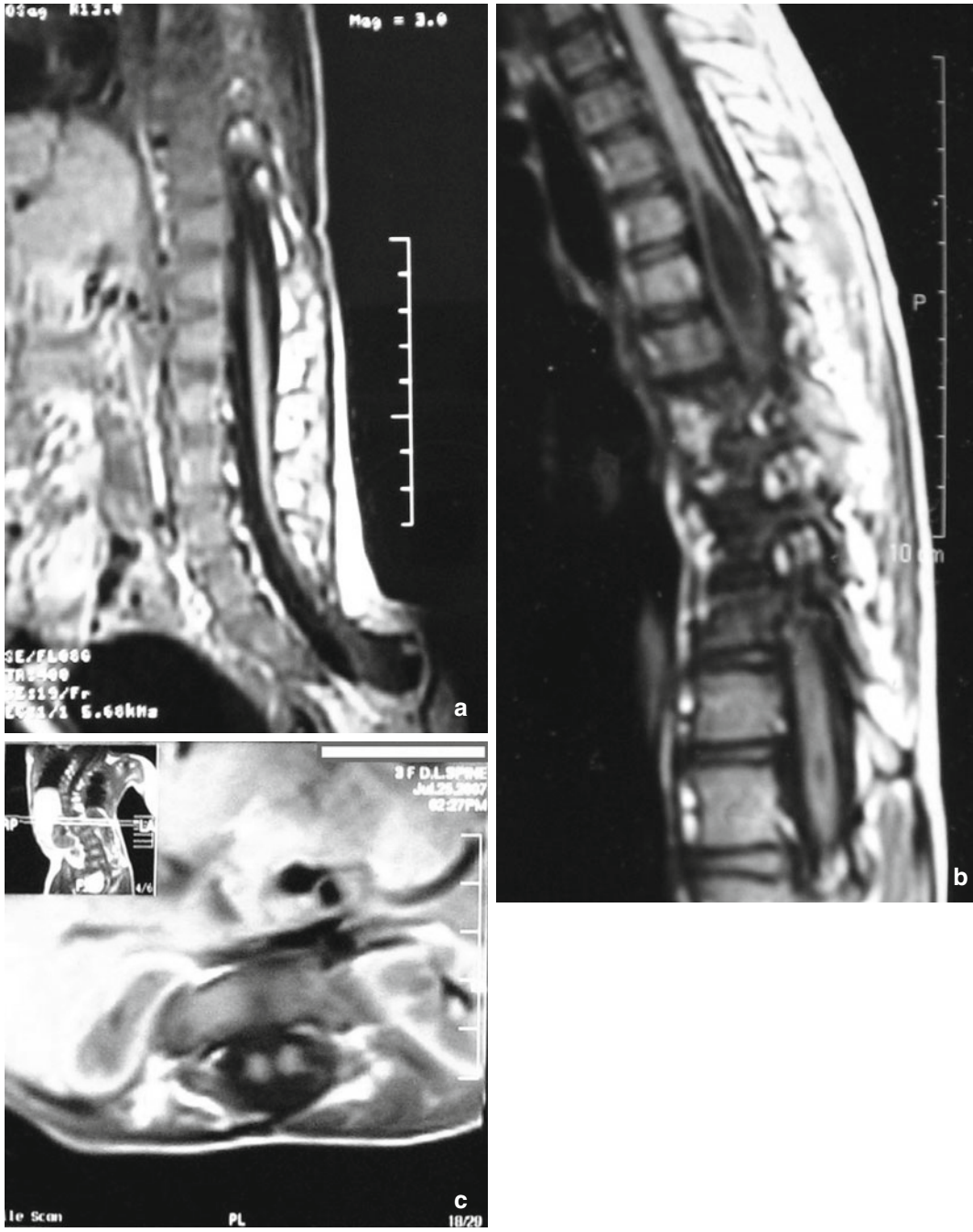

Fig. 8.2 Intraspinal anomalies associated with spinal deformities requiring neurosurgical consultation. (**a**) A sagittal T1-weighted MRI showing a tethered spinal cord, the most common MRI-identified intraspinal anomaly in congenital spinal deformity. Syrinx and tethered cord can also be found in case diagnosed as idiopathic scoliosis. (**b**) A sagittal T1-weighted MRI showing multiple variable sized dorsal syrinx associated with congenital scoliosis. (**c**) An axial T1-weighted magnetic resonance image (MRI) showing diastematomyelia with complete split of the cord at the dorsal region in a child with congenital scoliosis

manifested with back pain and headaches. The consequences of duralectasia include bony erosion or anterior meningoceles. Widened interpediculate distance, increased vertebral scalloping, and increased sagittal diameter can detect duralectasia in patients with Marfan syndrome. Dural ectasia is a major diagnostic criterion used in Marfan syndrome, especially in patients who previously have not had sufficient major and minor diagnostic criteria [24, 25].

An incidence of duralectasia of 63 % was reported in Marfan syndrome [26]. This incidence was noted to be 76 % in patients with Marfan syndrome and back pain and 41 % in patients with Marfan syndrome without back pain [27]. Because the calculation of dural volume requires sophisticated software programs that are not widely available and because duralectasia is important as a major diagnostic criterion for Marfan syndrome, many guidelines were developed to detect the presence of duralectasia on computed tomography or MRI scan with trials to establish normative values for lumbosacral dural sac dimensions [28]. An abnormal dural sac ratio (the dural sac diameter corrected for vertebral size) has also been used to identify duralectasia in patients with Marfan syndrome. In symptomatic duralectasia, posterior laminectomy has been sometimes implemented as a means of relieving back pain.

Neurofibromatosis

Dystrophic neurofibromatosis scoliosis is characterized by early onset, rapidly progressive curves that are difficult to treat and has a tendency to progress to a severe deformity. Dystrophic curves may be associated with kyphosis and have a higher incidence of neurologic injury. Most of these patients present with skin lesions (Fig. 8.3) as well as associated neurofibromas that envelope the bone or come from the canal (dumbbell lesion) [29].

Enlargement of the spinal canal caused by intraspinal tumors or duralectasia is common. It erodes the bony and ligamentous structures causing vertebral scalloping and meningocele formation. Meningoceles, pseudomeningoceles, duralectasia, and dumbbell lesions are related to the presence of neurofibroma or abnormal pressure phenomena in and around the spinal canal neuraxis. Paraplegia is an uncommon finding in patients who have dystrophic curves; it is more prevalent in patients who have severe vertebral angulation (kyphosis), vertebral subluxation, and soft tissue tumors in the spinal canal [30]. Occasionally, these intraspinal elements may compromise the cord directly when instrumentation and stabilization are attempted, or they may cause erosive changes in the bone, preventing primary fusion. A rare, but important, cause of paraparesis in scoliotic patients is spinal cord compression due to rib penetration [23, 31]. A CT scan is the most sensitive tool to diagnose intraspinal rib dislocation. A resection of the rib will prevent or improve paraparesis in most patients who have dislocation.

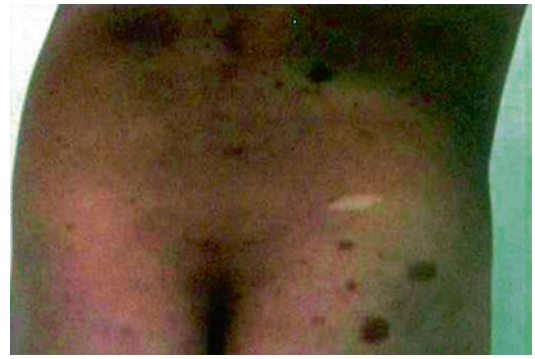

Fig. 8.3 Café au lait spots, cutaneous markers of systemic disorders observed in children with neurofibromatosis scoliosis

It is the surgeon's responsibility to correct and stabilize the spine with the most expedient, safe, and permanent method without causing neurologic injury. Therefore, it is imperative to evaluate such a condition in the preoperative period. MRI should be used in the investigation of all dystrophic curves before surgical treatment [32, 33].

Neuromuscular and Myelomeningocele Scoliosis

By definition, neuromuscular disorders are a group of diseases that affect any part of the nerve and muscle. These nerve tissue disorders include motor neuron diseases such as amyotrophic lateral sclerosis and spinal muscular atrophy, which

may involve motor neurons in the brain, spinal cord, and periphery, and ultimately weaken the muscle. Many of these diseases can cause early onset scoliosis due to a primary affection in the neural axis.

Spinal deformity also may be caused by paralysis secondary to the spinal cord injury. Scoliosis is secondary to spinal cord affection in patients with very young age; traumatic paralysis of the spinal cord may also lead to syringomyelia or traumatic tethering in 20 % of patients and should be looked for when patients with spinal cord injury have worsening symptoms. Another type of scoliosis is defined by the presence of a clear anomaly in the spinal cord – "myelomeningocelescoliosis." Both types of scoliosis, neuromuscular and myelomeningocele, have their own characteristics, complications, and way of management meriting a separate detailed discussion.

8.3.1.2 Cardiac

The relationship between cardiac abnormalities and scoliosis is a complex one. Both can originate from the same tissue defects in Marfan syndrome due to connective tissue disorder or in neuromuscular scoliosis due to different types of myopathy; scoliosis and congenital heart disease (CHD) can occur as a part of multiorgan congenital anomalies; in addition, there is an unexplained higher incidence of minor cardiac anomalies with idiopathic scoliosis. Conversely, scoliosis has a higher tendency to be present in children with congenital heart disease (CHD) with or without previous thoracotomy.

Idiopathic Scoliosis

Mitral valve prolapse (MVP) is known to be associated with thoracic skeletal anomalies, and MVP is four times more common in patients with severe idiopathic scoliosis than in the normal adolescent population. MVP and other valvular anomalies have been detected by echocardiogram and or ultrasound Doppler in 13.6–24.4 % of patients with idiopathic scoliosis as compared to 3.2 % in age- and weight-matched controls [14].

Patients with MVP are mostly asymptomatic, and only a systolic click or murmur can be detected on examination. Electrocardiogram (ECG) is abnormal in 21 % of patients with MVP as compared with only 1.6 % of patients with idiopathic scoliosis but no MVP. The persistent nature of MVP, even after corrective spinal surgery, may be related to factors other than geometric changes of the heart caused by abnormal thoracic curvature [12]. Looking at other comorbidities associated with idiopathic scoliosis, a significant relationship was found between valvular anomalies and other comorbidities. Valvular anomalies were detected in 17.2% with no comorbidity and in 50% with a comorbid condition; in this latter group of patients, routine echocardiography study seems advisable in the preoperative evaluation [34].

Congenital Scoliosis

CHD was found to be associated with congenital spinal deformity in 7–26 % of the patients. These include ventricular septal defects, atrial septal defects, patent ductus arteriosus, Fallot transposition of great arteries, pulmonary stenosis, sick sinus syndrome, and dextrocardia. Almost half of these children need medical therapy; some would require surgery for the cardiac condition in the future, and others will need to be kept under observation. This underscores the importance of a systematic clinical cardiac assessment and use of echocardiography for these patients. All patients in whom surgery is planned for correction of congenital spinal deformity should have echocardiography as part of preoperative workup. In addition, it has been suggested that patients with congenital scoliosis resulting from mixed bony defects and those with congenital kyphosis should have a routine echocardiogram because of the higher risk for CHD. These patients should be referred subsequently to a cardiologist for further management [18, 35].

Neuromuscular Scoliosis

Cardiac involvement may occur in most of the primary myopathies, including Duchenne muscular dystrophy (DMD), Becker muscular dystrophy (BMD), myotonic muscular dystrophy (MMD), and some cases of limb girdle muscular dystrophy. Dystrophin has been localized to the membrane surface of cardiac Purkinje fibers;

this localization probably contributes to the cardiac conduction disturbances seen in DMD and BMD.

A high (60–80 %) occurrence of cardiac involvement is present in patients of all ages with DMD and BMD; this can be detected via ECG and echocardiogram. However, only about 30 % of patients with DMD have clinically significant cardiac complications. Pulmonary hypertension also has been implicated in the cardiorespiratory insufficiency associated with DMD, and some investigators blame congestive heart failure as the cause of death in as many as 40 % of patients with DMD. The cardiac compromise may be disproportionately severe relative to respiratory compromise in some patients with BMD. Thus, ECG and echocardiography screening are indicated at regular intervals for all patients with BMD because severe cardiac involvement in BMD may occasionally precede the clinical presentation of skeletal myopathy. Patients with myocardial involvement need close follow-up and treatment by a cardiologist with expertise in this area. Some patients with BMD may be suitable candidates for cardiac transplantation [36].

A high prevalence of abnormalities found via ECG exists in MMD. Studies have shown that about one-third of patients with MMD have first-degree atrioventricular block, while about one-fifth have left axis deviation. Only 5 % have left bundle branch block. Complete heart blockage, requiring pacemaker placement, is rare but can occur. Patients with MMD should receive routine cardiac evaluations [36, 37].

Marfan Syndrome

Marfan syndrome is characterized by connective tissue disorder with classic triad affection ocular, skeletal, and cardiac. Cardiovascular system anomalies account for a significant proportion of the shortened life span with Marfan syndrome.

The most prominent cardiovascular manifestations of Marfan syndrome are known to be caused by defects in fibrillin 1. MVP occurs in 35–100 %, aortic dilatation in 75 %, mitral regurge in 44–58 %, and aortic regurge in 15–44 %. Many patients present with silent MVP, diagnosed through echocardiography (78–100%), largely exceeding the auscultatory diagnosis (45–70 %). Therefore, it is recommended that all patients suspected for Marfan syndrome be evaluated echocardiographically [38], as progressive aortic root dilatation, aortic regurgitation, dissection, or rupture is the most common life-threatening feature. Aortic regurge is an indicator of high risk for subsequent complications such as dissection. In general, morbidity and mortality are associated with aortic abnormalities rather than with mitral valve dysfunction. Investigators reported that sporadic cases of Marfan syndrome have more severe cardiovascular involvement compared to familial cases [39].

Due to early diagnosis, the awareness for milder forms of the disease, advances in aortic surgery, and medical treatment, the life expectancy of Marfan patients has increased from 37 years in the seventies to more than 60 years in the nineties [38].

Congenital Heart Disease

It has been well established that the incidence of scoliosis is higher in patients with CHD than in normal subjects. The incidence of scoliosis in patients with CHD has been reported in the literature to be from 2 to 19 %. The relatively wide range in the incidence of scoliosis associated with CHD is thought to be due to differences in types of CHD, criteria of patient's selection, effect of cardiac surgery, and the definition of scoliosis. In other words, the etiology of scoliosis associated with CHD is still unknown, and many factors such as CHD itself, cardiac surgery, thoracotomy, cyanosis, and other abnormalities may affect the onset of scoliosis. Some reports found strong correlation between thoracotomy done for cardiac surgery and the development of scoliosis in up to 22 % of their patients [39], whereas others found no correlation [40]. A number of theories have been proposed to explain the etiology of scoliosis associated with CHD, impaired oxygenation, and deficient or asymmetrical blood supply to the vertebral bodies or supporting tissues may be causative factors [41].

8.3.1.3 Urogenital

Congenital Scoliosis

The genitourinary and musculoskeletal systems are both of mesodermal origin and develop at the same time in the embryo. As a result, any genetic defect or other insult acting at a crucial stage of organogenesis, which results in a congenital vertebral abnormality, may also lead to a congenital genitourinary malformation. There is also the possibility that other developing organ systems will be affected. Thus, a cluster of disparate congenital abnormalities may occur. Renal anomalies are mostly nonhereditary, which supports the suggested etiology of an insult to the embryo between the fifth and seventh weeks. This period corresponds to the stage of organogenesis when the stem cell population is being established for the primordial organs. These interactions are sensitive to insult from genetic and environmental influences. In a 4-week-old embryo, the mesonephros is located from the sixth cervical vertebra to the lumbar spine. A stimulus in the lower cervical or upper thoracic area between the fourth and seventh weeks of gestation could simultaneously affect the developing mesodermal structures [42].

The frequent occurrence of congenital genitourinary abnormalities in patients with congenital scoliosis was reported in the 1980s [43]. The incidence of genitourinary abnormalities between 20 and 34 % has been found to occur in patients with congenital vertebral anomalies in different series using intravenous pyelography (IVP) and ultrasound. The most common urogenital anomalies associated with congenital spinal deformities are renal hypoplasia, horseshoe kidney, single kidney, congenital Megaureter, ectopic kidney (pelvic), hypospadius, pelviureteric junctional obstruction, posterior urethral valve, cloacal anomaly, epispadius, exstrophy of the bladder, hydronephrosis, and undescended testis [35]. It has been observed that the association of some malformation of the urinary system seems to be directly related to the occurrence of hemivertebrae; the location of the hemivertebra also seems important in relation to the side of agenesis of the kidney [43].

While these anomalies may remain asymptomatic, some can be associated with significant morbidity. Infection, obstruction, and the formation of calculus are the main reported problems. These patients are also at increased risk of proteinuria, hypertension, and renal insufficiency, and it is essential to have prolonged and careful follow-up. Some patients with urogenital anomalies (up to 25 % in some series) required surgery, others had abnormal renal function requiring medical therapy including dialysis, and the rest had abnormalities that do not affect renal function or do not require treatment [18].

Historically, IVP has been the investigation of choice in the evaluation of the morphology of the urinary tract, but diagnostic ultrasonography has been shown to be an acceptable alternative method of screening. Some centers reserve IVP for confirmation in those patients in whom an abnormality has been identified ultrasonographically or when the study is inconclusive. Ultrasonography is noninvasive, is less expensive, and has a reduced exposure to radiation. This is relevant in patients in whom multiple anomalies have been identified, and repeated imaging is required. Recently, the trend has been for an initial ultrasonographic evaluation. This can be difficult in the overweight patient and in those with severe spinal deformity in whom the chest is abutting against the pelvis. In these circumstances, IVP is recommended [42].

Neuromuscular Scoliosis

Urinary tract infections are frequently seen with paralytic spinal deformities as a consequence of paralysis of the muscles that control the bowel and bladder. Patients with paralytic spinal deformity classically have a higher postoperative infection rate, and the chronic urinary tract infections in these patients are often thought to be a potential cause. Patients with urinary tract infections should be treated preoperatively in an effort to eradicate the infection and eliminate the urinary tract as a potential source of bacteremia. It can be helpful to consult with a urologist who can evaluate the renal function of the child with regard to the child's ability to withstand major spinal surgery and can also be instrumental in optimizing the renal function of the child [44].

8.3.1.4 Musculoskeletal

A thorough musculoskeletal examination begins with height and weight measurements and should be collected at each visit to monitor weight gain and nutritional status. Standing height is measured to quantify linear extremity growth and total height over time. Torso length – measured while the patient is in the supine position – is used to assess linear truncal growth and is particularly useful in patients who cannot sit upright due to poor postural control (Fig. 8.4a, b).

Clinical photographs can be used to document a child's growth over time as well as the correction, or progression, of the spinal deformity including axial rotation, sagittal balance, and shoulder height. Routine photographs include posteroanterior (PA), anteroposterior (AP), lateral views, and "sunset" views of the thoracic and lumbar prominences seen on an Adams forward bend test. Photographs can also capture coronal balance and trunk shift, which are used to measure the translation of the head and thorax relative to the center of the pelvis, respectively (Fig. 8.5). Baseline and annual photographs of these parameters are helpful tools in monitoring the efficacy of the treatment plan.

Axial rotation of the trunk is often the most obvious spinal abnormality. The magnitude of trunk rotation can be visualized with an Adams forward bend test and measured using a Scoliometer. Rib prominence of the thoracic spine and lumbar prominence of the lumbar spine can be recorded photographically to document correction and progression.

Chest and rib deformities are evaluated by the observation of a child's breathing, respiratory rate, circumference of the chest, and pulmonary

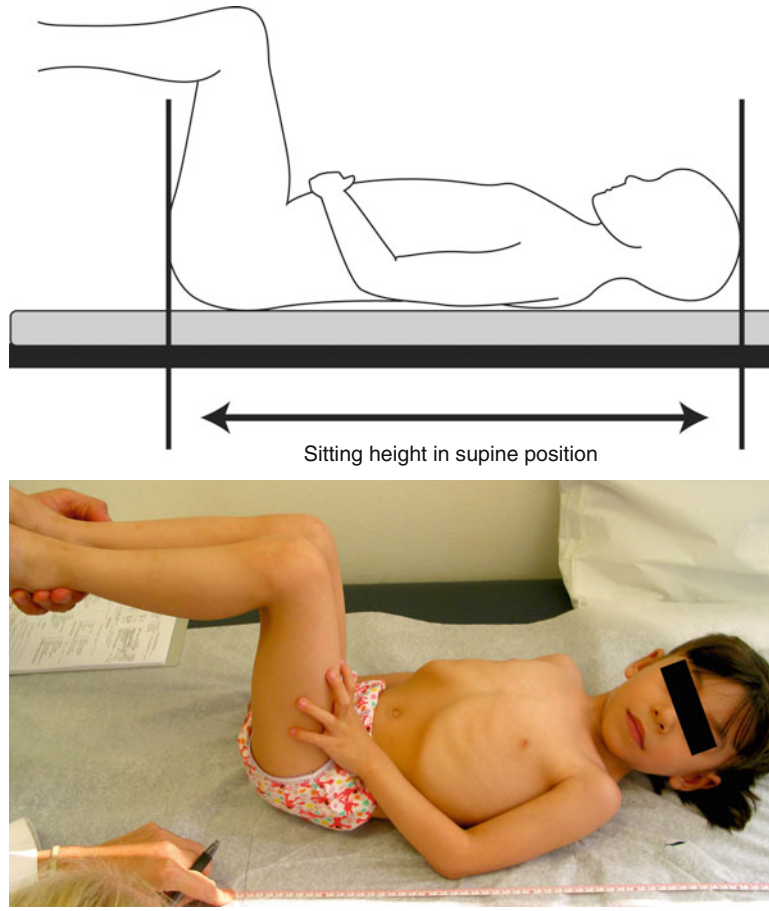

Fig. 8.4 (**a**) Measurement of sitting height (drawing). (**b**) Measurement of sitting height (clinical photo)

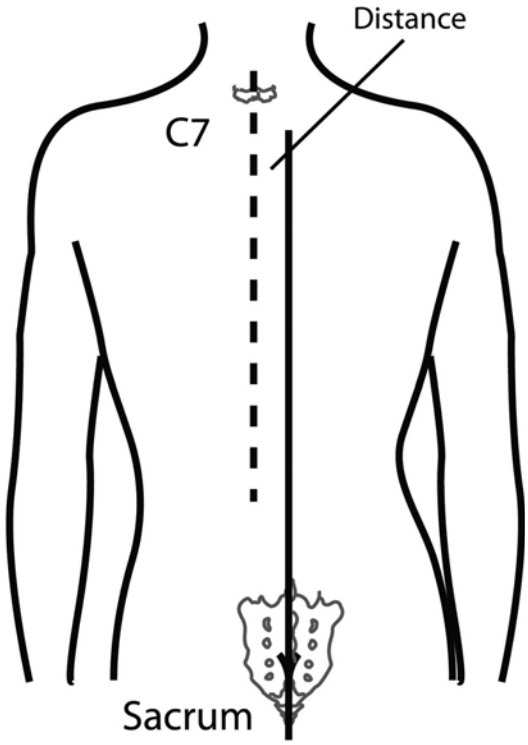

Fig. 8.5 Measurement of coronal balance

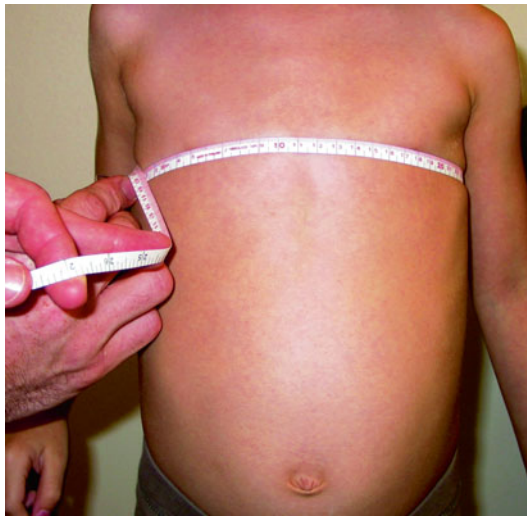

Fig. 8.6 Measurement of chest wall circumference

function testing (Fig. 8.6). The expansion of the thorax over time may be useful in gauging the growth of the child as well as correlating growth with pulmonary function.

A unique method used to determine chest-wall motion in a flail chest with absent ribs is known as the "Thumb Excursion Test" and was described by Campbell et al. [45]. To perform this test, the base of the chest is encircled from the back by the examiner's hands, with the examiner's fingers just anterior to the anterior axillary line of the patient (Fig. 8.7). The tips of the examiner's thumbs are positioned equidistant from the spine. The distance between each thumb tip is graded during inhalation as the thumbs move laterally away from the spine.

Idiopathic Scoliosis

Early onset scoliosis has been noted to be associated with ipsilateral plagiocephaly (Fig. 8.8) (an asymmetric and twisted head in reference to the spine), which is very common in children with pelvic flattening and obliquity and hip adduction. Subsequently, a correlation between infant positioning and early onset scoliotic deformities has been proposed, which was later questioned warranting further research [46]. Developmental dysplasia of the hip is also found at a higher frequency in patients with early onset idiopathic scoliosis than in children without scoliosis. Other comorbidities associated with idiopathic scoliosis include isthmic spondylolisthesis, hereditary exostosis, and slipped capital femoral epiphysis [34].

Congenital Scoliosis

Hypoplasia of upper extremity and lower extremity; wasting of one leg, clubfoot, and other foot deformities; Sprengel's deformity, dislocated hip, and polydactyly can be associated with congenital spinal deformities [18].

Neuromuscular Scoliosis

In neuromuscular scoliosis, there is an affection of different groups of muscles caused by the disease itself causing weakness, deformities, and difficult ambulation, or the affection can occur secondary to the spinal deformity resulting in pelvic obliquity, hip subluxation and dislocation, equines foot, and apparent leg length discrepancy.

Marfan Syndrome

The association of protrusioacetabuli and Marfan syndrome has been confirmed in many studies

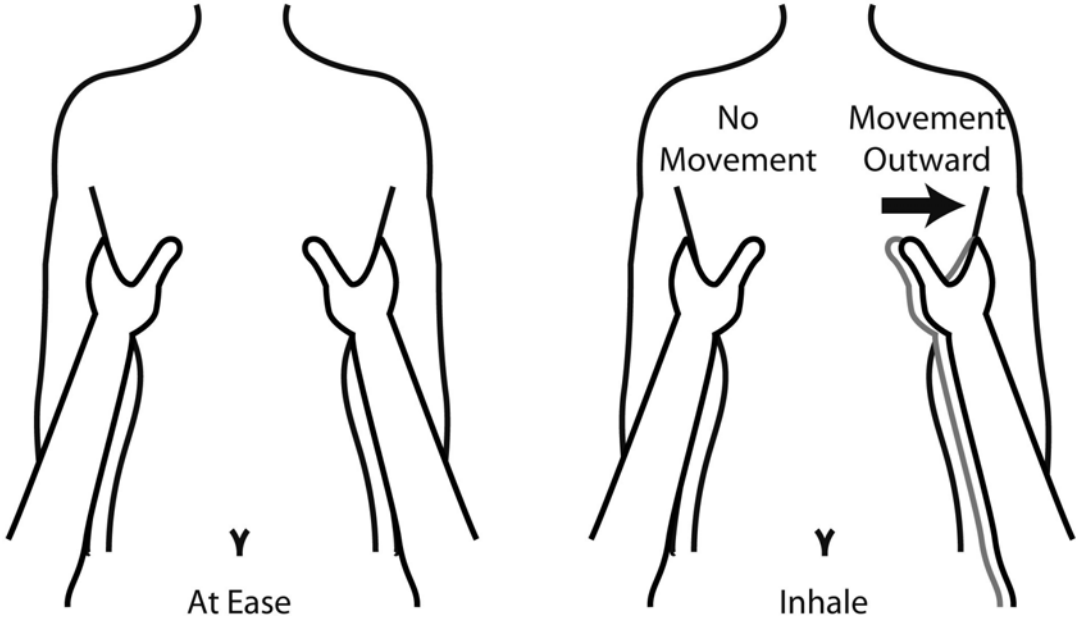

Fig. 8.7 Thumb excursion (Adapted from Campbell et al. [45]. With permission from Journal of Bone and Joint Surgery)

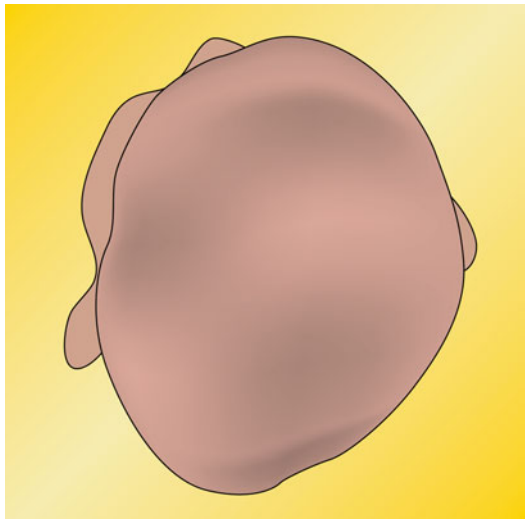

Fig. 8.8 Plagiocephaly, an asymmetric flattening and twisting of the head in reference to the spine, is common in children with early onset scoliosis; it may be related to the long-standing tilted position of the soft head of the infant when lying supine

with an incidence reaching around 30 % of the cases. Planovalgus foot deformity is sometimes observed in patients with Marfan syndrome with a reported incidence reaching 25 %, and it has been postulated to be caused by increased ligamentous laxity resulting from underlying connective pathology. Several investigators also reported decreased bone mineral density (BMD) in patients with Marfan syndrome; the significance of this finding in relation to fracture risk remains uncertain [24].

Neurofibromatosis

Patients with neurofibromatosis scoliosis may present with some type of bony dysplasia. The orthopedic complications with neurofibromatosis usually appear early; they include congenital tibial dysplasia with bowing and pseudarthrosis of the tibia, forearm, other bones, as well as overgrowth phenomenon of an extremity, and soft tissue tumors (see Fig. 8.3) [32].

8.3.1.5 Gastrointestinal

Inguinal hernia is found at a higher frequency in patients with early onset idiopathic scoliosis than in children without scoliosis; celiac disease, cystic fibrosis, and lactose intolerance were also found to be associated more with idiopathic scoliosis [34]. Imperforate anus, hernia, esophageal

atresia, and situs inversus were found to occur with congenital spinal deformities [18].

8.3.1.6 Cutaneous

A thorough physical examination in early onset scoliosis should be performed, beginning with a search for cutaneous markers of systemic disorders, such as the café au lait spots and axillary or inguinal freckling observed in neurofibromatosis (see Fig. 8.3) and the hairy patch (Fig. 8.1) associated with occult spinal dysraphism. Additionally, pigmented nevus, hemangioma, and dimples can be associated with congenital spinal deformities, sometimes denoting an underlying neurological abnormality. Skin scarring and defects can also be associated with myelomeningocele scoliosis, clearly affecting the outcome of these patients and warranting detailed assessment.

8.3.1.7 Mental Status, Disability, and Pain

The child's mental status should be noted, especially if the child has limited communication abilities or evidence of a developmental delay. Changes in verbal communication should also be monitored before and after treatment. The presence of a developmental delay may suggest an underlying syndrome with more global involvement. Mental disabilities can be associated with some types of neuromuscular scoliosis due to brain affection. The presence of cognitive delay has been shown to correlate with curve progression in early onset scoliosis and particular attention should be paid to whether the child has appropriately reached developmental milestones [46].

Idiopathic pediatric scoliosis patients have more pain than asymptomatic pediatric age group without scoliosis, and those with Schmorl's nodes often had greater pain than those without; however, the overall degree of disability is clinically mild. This is different from neuromuscular scoliosis, which usually results in a considerable degree of both pain and disability. The etiology for painful idiopathic scoliosis is thought to include muscular pain due to eccentric loading about the apex of a curvature, asymmetric facet joint loading resulting in facet arthritis or synovitis, discogenic pain, or a combination of these factors. Human studies have shown disc degeneration at the concave aspect of scoliotic discs; however, the progression from disc degeneration to discogenic pain is still not fully understood. The location of the pain is mainly over the apex of the primary curvature; to a lesser extent, at the midline at levels corresponding to patients' Schmorl's nodes; and finally, in the interscapular and low back regions.

Overall, disc degeneration was similar in scoliosis and asymptomatic control groups; however, specific aspects of degeneration, such as Schmorl's nodes and inflammatory end plate changes, were more common, suggesting that symptoms in the scoliosis patients may, in part, have a discogenic etiology and that pain in scoliosis may occur in an abnormal loading environment combined with abnormal endplates [36].

8.4 Diagnostic Laboratory Testing

A child who is to undergo surgical intervention will require a preoperative complete blood count (CBC) and a metabolic panel to identify disorders such as anemia, infection, and other diseases. If the patient is cooperative, a pulmonary function test (PFT) should be performed to assess lung capacity and expiratory volume.

8.5 Diagnostic Imaging

8.5.1 Radiographs

Routine primary evaluation radiographs should include a standing posteroanterior and a standing lateral – both of which should span from the lower cervical spine down to the femoral heads. If planning surgical treatment, preoperative films should also include coronal supine left and right bending films. Traction or bolster films are also useful to determine the degree of curve flexibility, appropriate levels of instrumentation, and expected degree of correction.

8.5.2 Magnetic Resonance Imaging (MRI)

If there is concern for an intraspinal anomaly (e.g., syrinx, tethered cord, tumor, diastemetomyelia), an MRI should be performed to identify neural element abnormalities.

8.5.3 Computed Tomography (CT)

If the child's radiographs demonstrate congenital anomalies, a CT scan (preferably with three-dimensional reconstruction) is obtained to further investigate bony abnormalities, such as failure of vertebral segmentation or formation. Intravenous contrast is not routinely needed. However, intrathecal contrast may be warranted if an MRI is contraindicated for medical reasons and further evaluation for intra-spinal anomalies, such as cord tethering or diastematomyelia, is required.

8.6 Developing a Comprehensive Treatment Plan

Managing early onset spinal deformities requires the commitment and flexibility of the surgeon and the family. A carefully developed treatment plan is likely to span over several years until the child reaches skeletal maturity or even adulthood. Short-term and long-term goals of the treatment plan should be identified and clearly discussed with the patient's family. A multi-disciplinary approach may be warranted in complex cases, which will require additional oversight by the spine surgeon.

8.7 Postoperative Examination

If the child undergoes surgery, postoperative evaluations should include an examination of the skin incision, assessment of neurologic status, and continual evaluations of implant fixation for signs of prominence, junctional problems, loosening, and migration.

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Imaging of the Growing Spine

9

John T. Smith, Peter O. Newton,
Christine L. Farnsworth, and Kevin Parvaresh

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Key Points

- Innovative imaging technology, such as the EOS system, allows for postural assessment of spinal morphology.
- Advances in three-dimensional (3D) imaging are being utilized to improve deformity progression analysis, surgical planning, and translational research potential.
- Functional imaging such as dynamic MRI shows promise for a novel method for assessment of outcomes in the management of pediatric deformity.
- New technology for 3D analysis is significantly reducing radiation exposure in children.

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9.1 Introduction

Management of deformity and disease in a child with a growing spine presents unique challenges to the treating physician. Since the introduction of X-rays by Wilhelm Roentgen in 1895, there has been a steady improvement in our ability to quantify and analyze spinal deformity. More advanced imaging techniques now make it possible to understand these deformities in three dimensions, allowing for a better understanding of strategies for three-dimensional (3D) correction.

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There have been significant advances in our ability to image spine and chest deformity in growing children. Rather than simply measuring the nature and degree of deformity, imaging can be used to assess volume and function in children who are unable to cooperate with normal test such as pulmonary function studies. This is being done with dynamic magnetic resonance imaging (MRI) and a significant reduction of exposure using the EOS system. The purpose of this chapter is to provide an overview of current imaging techniques for managing children with deformity in the growing spine.

9.2 Plain Radiography

The initial evaluation of suspected spinal deformity in a child begins with a thorough history and physical examination. If there is clearly a spinal deformity present, then plain radiographs should be obtained. It is the responsibility of the requesting physician to specifically tell the imaging department the type of film needed. When possible, these are best taken as orthogonal views in the upright or standing position (posterior–anterior (PA) and lateral) (Fig. 9.1). The PA exposure lessens the amount of radiation exposure to the breast tissue and reproductive organs. The entire spine from C1 to the pelvis should be included as a single image and should include the entire chest wall. Many spinal deformities in children impact the shape and volume of the chest. Rib anomalies and fusions are easily seen, and they provide a sense of relative chest hypoplasia. Many congenital deformities in the thoracic spine shorten the spine and result in constricted lung volumes. Hypoplasia of the chest restricts normal alveolar multiplication during growth and may result in thoracic insufficiency syndrome as described by Campbell et al. [1].

Plain radiographs allow for assessment of the etiology of the deformity (idiopathic, congenital, neuromuscular, etc.) and analysis of both coronal and sagittal balance. Curve severity can be measured and documented by measuring the various curves using the Cobb technique. Skeletal maturity is assessed by looking at various growth plates such as the tri-radiate cartilage of the hip or the iliac apophysis (Risser Sign). It is valuable to have a visible marker on the film to account for magnification and allow for accurate measurements of spinal height and growth over time. This allows for measurement of spinal growth using T1-S1 heights. Campbell et al. [1] described the space available for the lung (SAL), a ratio of the distance from the top of the lung to the apex of the diaphragm, comparing left to right (Fig. 9.2a, b). This measurement may have direct implications on the development of the lung.

Differences in the relative angle of the ribs are referred to as rib-vertebral angle differences

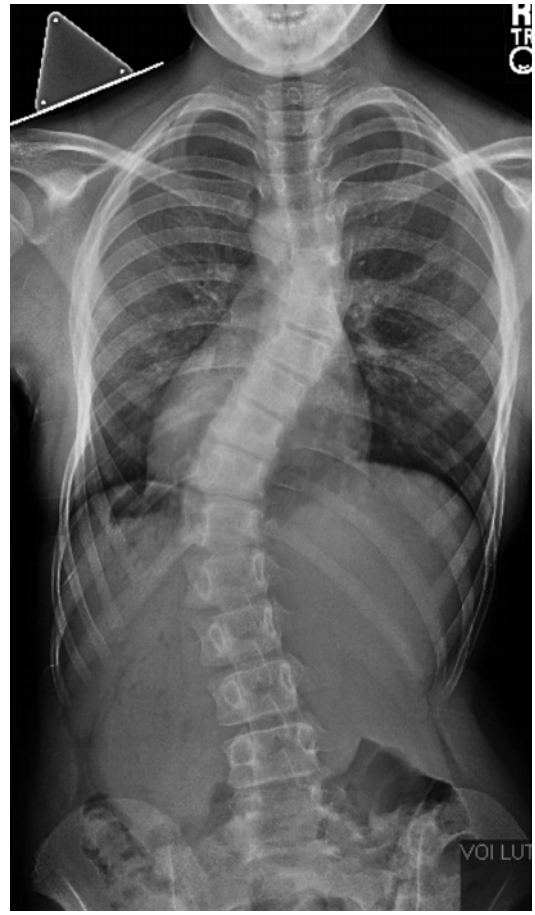

Fig. 9.1 PA radiograph of the spine. The PA radiograph allows for assessment of diagnosis, curve type, magnitude, coronal balance, and skeletal maturity. The PA view has significantly less radiation exposure to sensitive tissues including the thyroid and breasts

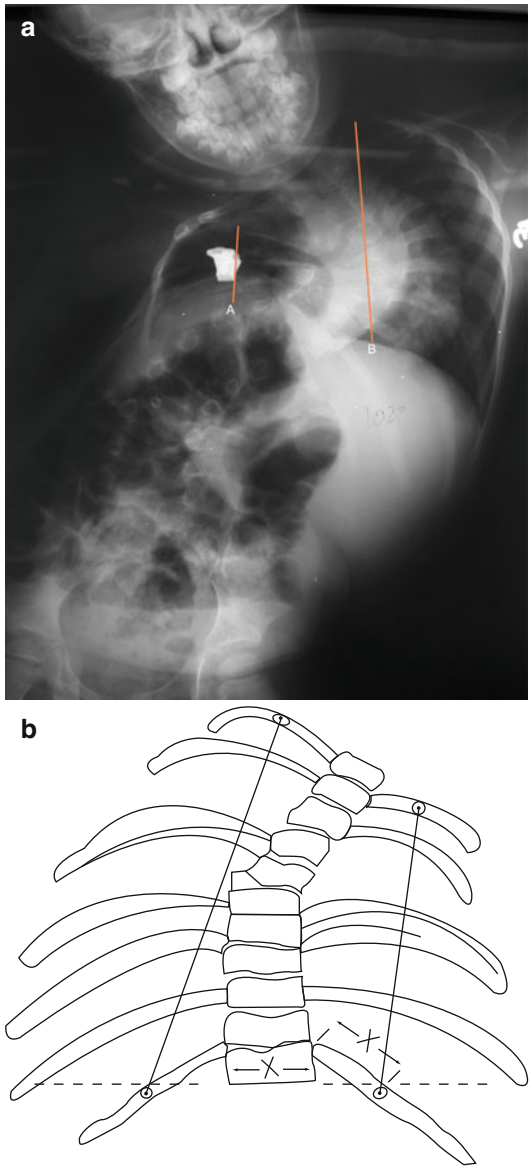

Fig. 9.2 (**a**) Space available for the lung (SAL). This is a ratio that documents the effect of the three-dimensional deformity of the chest on potential lung growth. (**b**) Space available for the lung (SAL). The SAL is a ratio expressed as a percentage of the distance from the diaphragm to the apex of the lung (A/B) measured on an upright radiograph of the chest when comparing one side to the other. In this example, the SAL is 70 % when comparing the right to the left

(RVADs). An RVAD >20° in children with idiopathic early onset scoliosis is felt to correlate with a greater risk of the curve becoming progressive during growth rather than spontaneous correction that may be seen when the RVAD is <20°. An RVAD >20° might warrant an earlier use of correction techniques such as elongation-derotation casting.

9.2.1 EOS System

The EOS system (EOS Imaging, Paris, France) is a recent advancement in imaging technology that allows three-dimensional (3D) assessment of scoliosis deformity through the acquisition of two-dimensional (2D) images. The system utilizes two orthogonal X-ray beams moving in unison with particle detectors along the length of the imaging area, producing two linked orthogonal radiographs. Three-dimensional reconstruction is then performed using SterEOS® software (EOS Imaging, Paris, France), in which a 3D base template is manipulated to produce an anatomic 3D model (Fig. 9.3). The base template for this technology was generated using contour detection from 2D radiographs and subsequently validated with CT images using a synthetic spine phantom model [2]. The template is contoured to match multiple discrete points measured manually on the 2D radiographs to generate the final 3D model (Fig. 9.4). The resultant 3D model may then be manipulated in all three planes based on the operator's preference for assessment.

The 3D EOS model offers significant clinical utility for 3D analysis of spinal deformity. The SterEOS® software measures many clinically significant scoliosis parameters including various curves, axial rotation of the apical vertebrae, sagittal alignment (T1–T12, T4–T12, L1–L5, and L1–S1), pelvic incidence, sacral slope, sagittal pelvic tilt, lateral pelvic tilt, and pelvic axial rotation. Individual vertebral orientation and rotation between adjacent vertebrae from T1 to L5 are calculated in the frontal, lateral, and axial planes (Fig. 9.5). Importantly, the reproducibility of SterEOS® software measurements compared to manual measurements using conventional radiography has been verified [3, 4].

There are several advantages as well as limitations to this new technology. Standing image

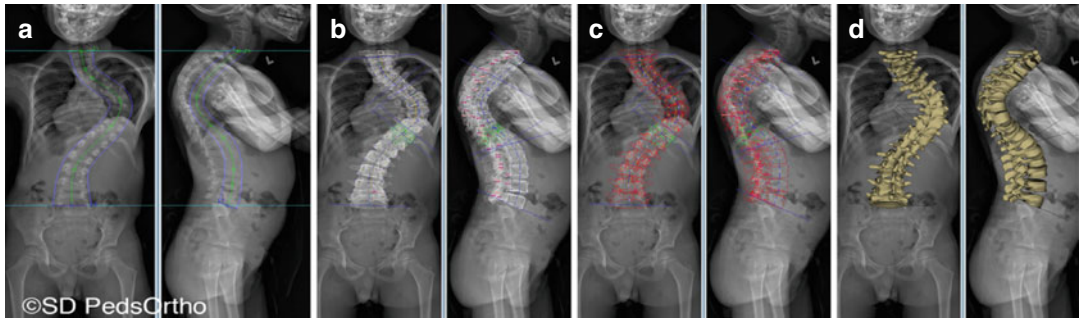

Fig. 9.3 Biplanar imaging (EOS Imaging, Paris, France) of a 4-year 4-month-old child with early onset scoliosis. (**a**) Vertebrae are identified by the operator. The spine is then detected by the software (SterEOS®). Next, a template is superimposed, which the operator adjusts to match the radiograph contour. (**b**) Radio view, (**c**) contours view, and (**d**) surface views of the adjusted template are shown

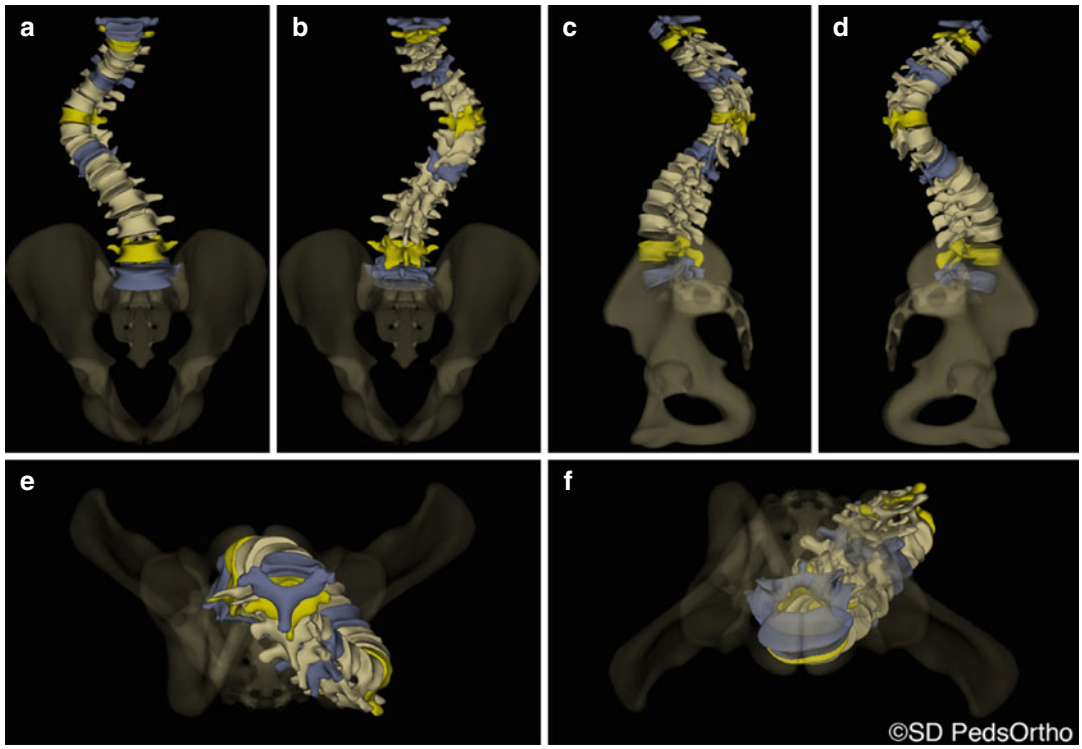

Fig. 9.4 3D model using the template created (Fig. 9.3) morphed to the patient's images. (**a**) Anteroposterior view, (**b**) posteroanterior view, (**c**) lateral left, (**d**) lateral right, and axial views from (**e**) overhead and (**f**) underneath

acquisition is a protocol, allowing proper assessment of gravity and posture relative to deformity but requiring patients to be stable upright and weight-bearing. Image distortion is reduced due to enhanced particle detection and limited scatter. Radiation exposure is reduced by eight to ten times that of conventional radiographs and 800–1000 times that of CT [5, 6]. Even lower "micro-dose" modalities have been developed further reducing radiation fivefold and are currently undergoing safety and efficacy evaluation. Although validated, the SterEOS® 3D model is based on a standard template that may not be truly accurate for deformities without

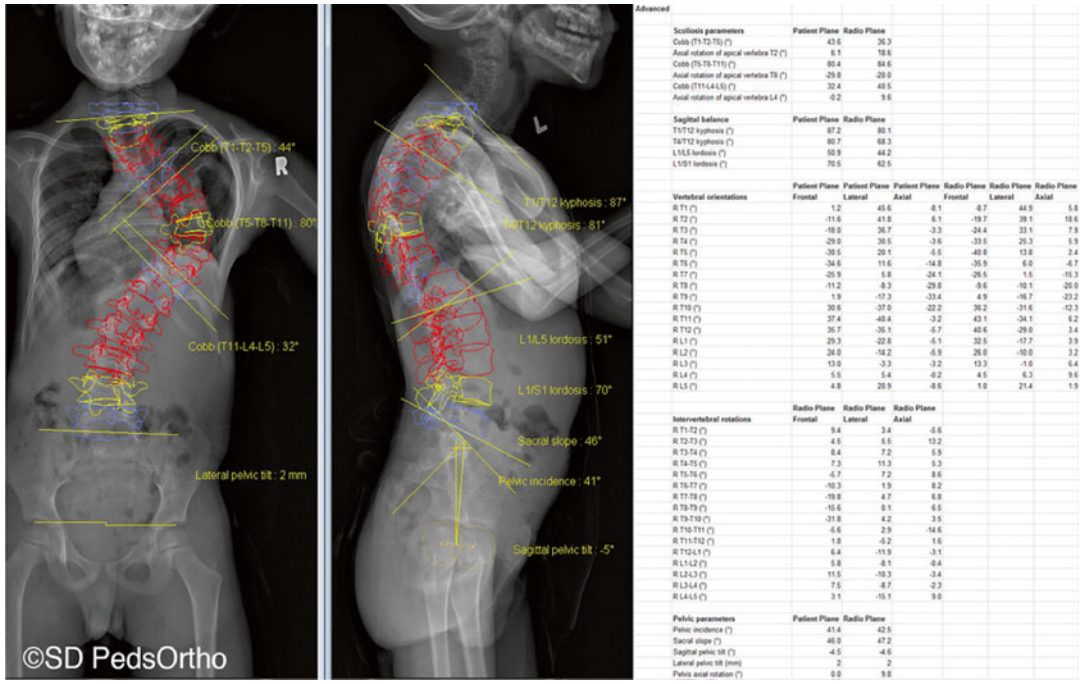

Fig. 9.5 Contours view template with measurements and measurements output

normal osseous development, such as congenital scoliosis. Altogether, this EOS system exists as a powerful tool for 3D evaluation of scoliosis deformity.

9.3 Computerized Axial Tomography

CT scans are an invaluable tool for the diagnosis and treatment planning for complex spinal deformity in the growing child. The CT scan allows for accurate measurement of spinal deformity including spinal length, rotation, and lung volume and to characterize types of vertebral and chest wall malformation. These determinations come with a significant radiation cost. The ability to visualize a 3D reconstruction of the spine and manipulate the image is very useful for pre-operative planning (Fig. 9.6).

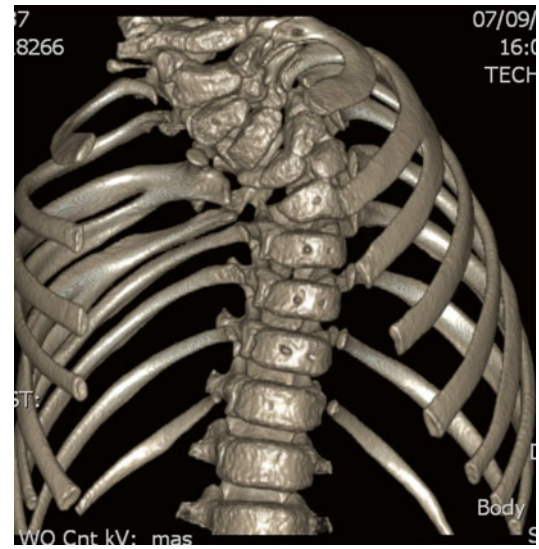

Fig. 9.6 3D reconstruction of a CT scan of a child with a complex congenital scoliosis with multiple vertebral anomalies and fused ribs

There have been several unique measurements described based on CT scans. Ilharreborde et al. [7] described the spine penetration index as a way to measure the space occupied by the spine inside the thorax, which correlates with increased thoracic lordosis. Gollogly et al. [8] described the thoracic distortion index, which measures the cross-sectional area of the thorax and compares

this to age-matched normal children. This index correlates closely with the amount of impairment in pulmonary function.

CT scans can be used to measure lung volumes in children, which then become a useful tool to evaluate change in lung volume after treatment. Gollogly et al. [8] first reported using CT scans to measure lung volumes after treatment for congenital scoliosis and fused ribs using the VEPTR device. Normative data for CT lung volumes are now available for comparison [9]. Smith et al. [10] showed an average increase in lung volume following expansion thoracoplasty of 257 cc at 12-month follow-up.

CT scans can be used to generate a plastic model of the spine for study prior to complex corrective surgery such as vertebral column resection. Rapid prototyping technology had improved greatly in recent years simplifying the process to obtain these models.

9.4 Magnetic Resonance Imaging

The use of magnetic resonance imaging (MRI) is the standard of care for the evaluation of complex spinal deformity in the growing child (Fig. 9.7). The incidence of intraspinal pathology varies with the etiology of the deformity. In congenital scoliosis, the associated incidence of intraspinal malformations ranges from 3 to 52 %. This is dependent on the pattern of malformation, with the highest incidence associated with multiple unilateral hemivertebra and contralateral failure of segmentation (bars). Congenital kyphosis is also associated with a high incidence of intraspinal anomalies. MRI has replaced more invasive and less specific imaging modalities such as myelograms and CT myelograms as screening tools for intraspinal pathology. However, these remain useful tools when MRI is contraindicated.

Dynamic MRI is a new tool being used to noninvasively evaluate the effect of spinal deformity on chest wall and diaphragmatic function. The excursion of the diaphragm can be measured during inspiration and expiration and motion of the chest wall. These measurements allow for quantification of the effects of different treatments on pulmonary and diaphragm function (Fig. 9.8).

9.5 Radiation Risk

Children with significant spinal deformity often require repetitive exposure to radiation during growth. Although difficult to quantify, different forms of radiation such as CT scans have significantly different degrees of risk over time. Radiation doses are cumulative over time. Doody et al. [11] reported that females with multiple scoliosis radiographs had a twofold increase in mortality from breast cancer when compared to age-matched controls. This risk is correlated with increasing numbers of radiographs and cumulative radiation dose. Levy et al. [12] calculated that if the AP view was replaced by the posterior–anterior (PA) view, a threefold to sevenfold reduction in cumulative doses to the thyroid gland and the female breast would be achieved, yielding threefold to fourfold reductions in the lifetime risk of breast cancer and a halving of the lifetime risk of thyroid cancer.

Reducing the frequency of obtaining plain radiographs is one way to decrease radiation exposure. A minimum interval for comparison radiographs is 6 months. As a general rule, a rapidly progressing curve would change about 1–2° per month in the growing child. With the average intra-observer measurement error for the Cobb angle of 3°, it is logical that to see a significant difference in a curve that is not simply measurement error, 6 months would be a minimum interval. Presciutti et al. [13] and Pace et al. [14] compared radiation exposure for adolescent idiopathic scoliosis between operative, braced, and observation cohorts. The operative group averaged 1400 mRads per year; braced group, 700 mRads per year; and observed group, 400 mRads per year [14]. For the operative group, 78 % of their radiation exposure was due to intraoperative fluoroscopy. Overall, the operative group received 8–14 times more radiation than did braced or observed patients.

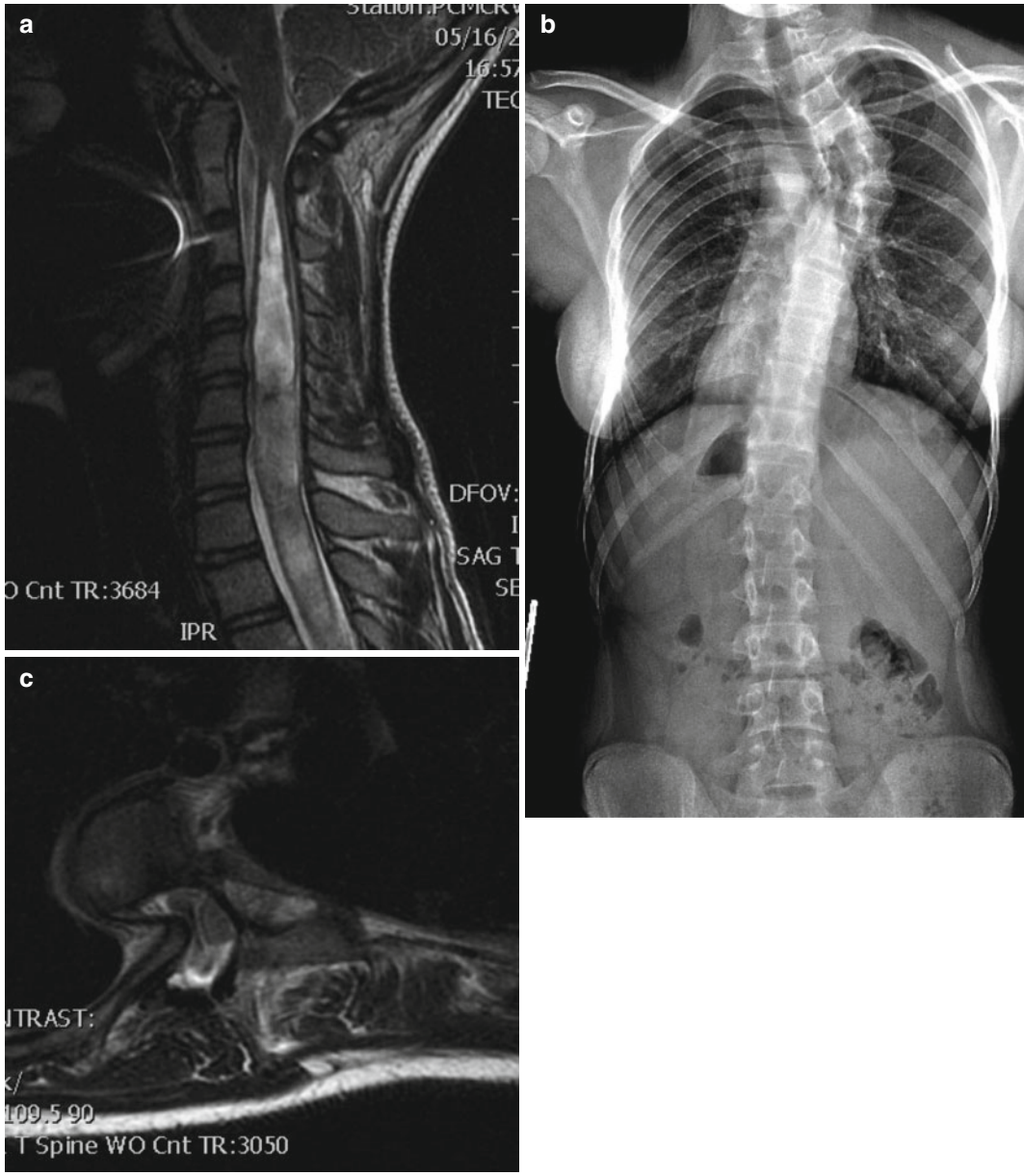

Fig. 9.7 (**a**) MRI is an important screening tool to look for associated malformations of the spinal cord. This figure demonstrates a Chiari malformation and associated Syrinx. (**b**) Plain radiograph of a 14-year-old female with neurofibromatosis, Type 1. It was not evident that she had intrathoracic migration of four ribs into the spinal canal until she had a pre-operative MRI (see Fig. 9.4c). (**c**) An axial MRI in a child with dysplastic scoliosis detected migration of the ribs into the spinal canal, which was not apparent on a plain radiograph (Fig. 9.4b)

CT scans significantly increase radiation exposure for the child. The actual amount of radiation varies with the type of scanner, extent of the spine imaged, number of slices obtained, and settings of the machine. The additional radiation of a CT scan is estimated to be the equivalent of 15–600 chest radiographs. The average radiation from a single chest radiograph is 0.02 mSV; a thoracic spine film is 0.07 mSV; and a CT scan of the chest is 7 mSV, the equivalent of 400 chest

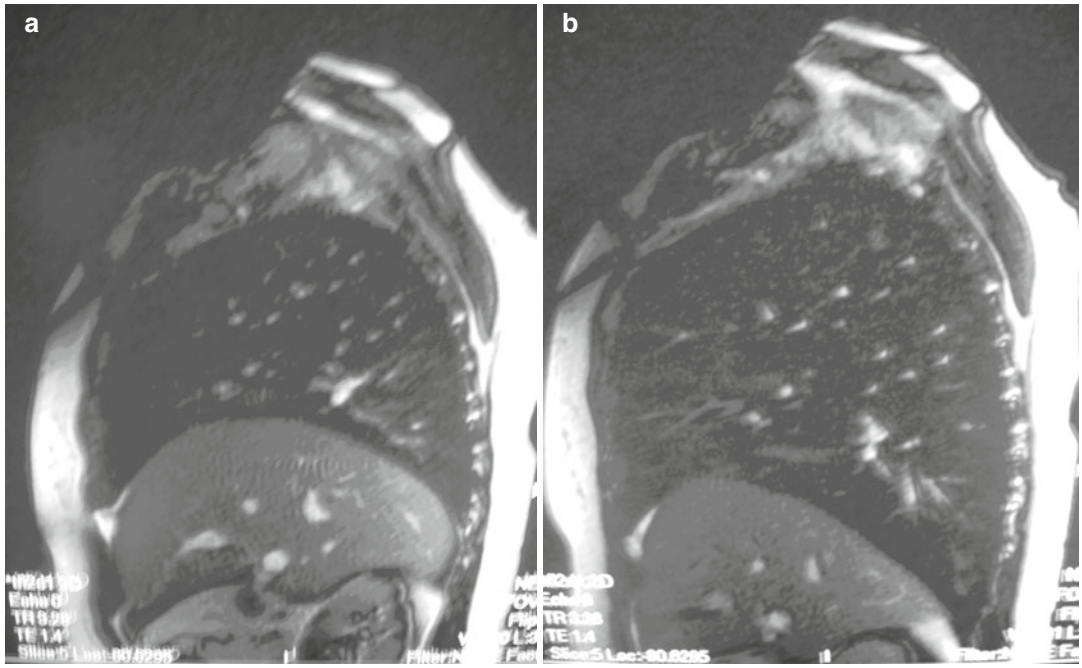

Fig. 9.8 (**a**) Dynamic MRI imaging used to measure the movement of the diaphragm in maximum expiration. (**b**) Dynamic MRI showing diaphragm excursion in maximum inspiration

radiographs. The increasing popularity of using the O-Arm intra-operative CT scan for navigation has the potential to dramatically increase radiation exposure when multiple imaging "spins" are obtained during a single surgery. In our institution, we attempt to reduce radiation exposure by asking for non-enhanced CT scans using 5-mm cuts. This technique still allows for measurement of CT lung volumes, 3D reconstruction of the spine, and accurate characterization of pedicle anatomy.

The EOS system offers the promise of greatly reducing radiation to children when compared to CT scans while allowing for 3D analysis of the spine. The dose of radiation is estimated to be 800–1000 times less than a CT scan with 3D reconstruction.

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Part III

Spinal Deformities in the Growing Child

Idiopathic Early Onset Scoliosis

10

Gregory M. Mundis Jr., Laurel C. Blakemore, and Behrooz A. Akbarnia

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Key Points

- Careful history and physical examination are imperative to rule out other etiologies of scoliosis in growing children.
- Quality PA and lateral spinal radiographs are essential to the evaluation and management decision making.
- In younger children with (infantile) scoliosis, curves with an RVAD of 20° or more, Cobb angle of 25° or more, or a phase 2 rib head should be followed closely for progression. Curves with an RVAD of less than 20° and a phase 1 rib head typically do not progress.
- All curves with a Cobb angle of 20° or more should be evaluated with advanced imaging (typically an MRI of the entire neural axis) to rule out brain and spine anomalies.

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- Our current recommendation for treatment of progressive idiopathic early onset scoliosis is a distraction-based technique of dual growing rods placed subfascially using a two-incision technique with a skin bridge. Our preference when possible is to place magnetically controlled growing rods to minimize the return to surgery rate for routine lengthenings.

10.1 Introduction

> "It develops rapidly and relentlessly, causing the severest form of orthopaedic cripple with dreadful deformity, marked dwarfing and shortening of life."
> J.I. James, MD on infantile scoliosis, 1959

Management of spine deformity in children 5 years of age or less presents one of the most challenging tasks in spine surgery. It requires a thorough knowledge of normal spine development as well as the etiology, natural history, clinical evaluation, and available nonoperative and operative treatments for infantile scoliosis. Early recognition by both parents and pediatricians is essential, and immediate orthopedic referral is mandatory as early treatment will ultimately affect patient outcome.

Harrenstein [26] in 1936 coined the term infantile idiopathic scoliosis (IIS). He treated 46 children with bracing with mixed success and attributed the deformity primarily to rickets [25]. In 1952, James [30] reported on 33 cases of scoliosis in infants aged 3 years and younger. They were predominantly boys with left-sided thoracic curves. Four cases resolved spontaneously, but the remainder progressed aggressively. In 1954, he first described scoliosis according to chronologic age at a presentation, including infantile from birth to 3 years, juvenile with onset up to 8 years, and adolescent with onset from 10 years to maturity [31]. Interestingly, no reference was made for those between 8 and 10 years [31]. Dickson [13] later recommended that scoliosis in children be classified as early (5 years or less) or late (>5 years) onset. The rationale for this is twofold. As Dimeglio and Bonnel [16] have shown, growth velocity in the spine is highest from birth to 5 years, followed by a deceleration between age 6 and 10 years. From 11 to 18 years, there seems to be another peak in growth velocity but not equal to that of early life. Early onset, therefore, more accurately describes this growth. Similarly, this group is at a higher risk for developing significant cardiopulmonary complications if thoracic curves progress, whereas these complications are rare in the late onset group. Complications include pulmonary hypoplasia, restrictive pulmonary disease, pulmonary artery hypertension, corpulmonale, and thoracic insufficiency syndrome.

A recent publication supported by the Children's Spine Study Group and the Growing Spine Study Group as well as SRS and POSNA has called for consensus in terminology of early onset scoliosis. Their summary suggested that EOS should be inclusive of all etiologies. Furthermore, EOS should refer to all patients with the diagnosis of scoliosis before the age of 10. It was felt that 10 years represents an age that also triggers different interventions. Using Dickson's nomenclature, children between 0 and 5 years and 5 and 10 years are likely going to receive similar growth friendly treatments, so differentiating early and late onset may not be as relevant. However, children 10 years and older are likely to have fusion as the primary surgical intervention and under 10 years, growth friendly interventions [19]. This is also supported by a recent publication by Skaggs et al., who defined EOS as "scoliosis of any etiology developing before the age of 10. The SRS Growing Spine Committee has further sub categorized EOS into: idiopathic, congenital, thoracogenic, neuromuscular, and syndromic. Using this universal terminology will be essential for communication, teaching, and research [62].

This chapter aims to equip the spine deformity surgeon with all the relevant knowledge to diagnose, educate, and effectively treat the child with early onset idiopathic scoliosis.

10.2 Natural History

10.2.1 Growth and Development

Dimeglio [15] and Dimeglio and Bonnel [16] illustrated that spine growth velocity is greatest from birth to 5 years, averaging >2 cm growth per year during those years. From the age of 6 to 10 years, velocity decreases to 0.5 cm per year and then increases to 1.3 cm per year from the age of 11 to 18 years. Chest growth is most easily assessed as thoracic volume, which shows a similar trend as spine growth. At birth, it is 5 % of adult volume. By 5 years of age, it has reached 30 %, a staggering 600 % increase in volume [15]. At 10 years of age, lung volume is 50 % and doubles to 100 % of adult capacity by the age of 15 years in both males and females. Lung development is best measured by change in alveolar volume and number. It is estimated that 20 million alveoli exist at birth and increase to 250 million by the age of 4 years and complete development by 8 years of age. A similar increase in alveolar volume also occurs. Respiratory branches also increase from 20 at birth to 23 by 8 years.

10.2.2 Epidemiology

Several authors have reported the incidence and prevalence of infantile idiopathic scoliosis (IIS) and juvenile idiopathic scoliosis (JIS) [13, 33, 48]. In the United States, IIS comprises less than 1 % of idiopathic cases. A slightly higher incidence has been reported in Europe [33, 48]. Unlike late onset, it is more common in males with a ratio of 3:2, and curves tend to be left sided. It occurs in the mid to lower thoracic spine in 75–90 % of cases [13, 33, 61]. Since the initial description by James [31] in 1951, it appears that the incidence has decreased. McMaster [48] most recently reported on a declining prevalence of patients with IIS scoliosis in Edinburgh, at a major referral center for scoliosis in the United Kingdom. Between 1968 and 1972, they averaged 16.5 new patients per year with a 34 % incidence of progressive curves. From 1980 to 1982, there was an average of two referrals per year. During this same time period, referrals for adolescent idiopathic scoliosis increased.

JIS accounts for 12–21 % of reported idiopathic cases [33, 54]. It is more prevalent in females with a 2:1–4:1 female-to-male ratio. Between 3 and 6 years of age, the gender difference is neutral, and after 10 years of age, females are affected at a rate of 8:1 [22, 66]. Males are usually diagnosed by 5 years of age, and females, by 7 years of age. This difference, as well as the age of skeletal maturity, likely explains a higher rate of progression in males. Right-sided thoracic and double major curves are the principal curve patterns associated with JIS [21, 44].

10.2.3 Prognosis

James [30] in 1951 reported his initial series of 33 patients, in which 18 (55 %) were progressive; 11 (33 %), stable; and 4 (12 %), spontaneously resolved. In 1954, he increased his numbers to include 52 children who were treated with physiotherapy, plaster-of-Paris beds, and orthoses [31]. Curves in 43 patients progressed (83 %), with all curves being >70° at the age of 10 and several progressing >100°. In the remaining nine patients (17 %), the curves resolved spontaneously without treatment. In 1959, James et al. [33] reported on 212 infantile cases from two separate institutions. Seventy-seven (31 %) patients had spontaneous resolution, and the remainder progressed aggressively (135/212). Of these 135 patients, 47 were between 0 and 5 years, and 23 of these already had a curve >70°. Thirty-seven patients were between 5 and 10 years, and 27 of 37 had a curve >70°, and in 14, the curves were >100°. Of the 23 children aged 11 years and older, 12 had a curve >100°, and two at skeletal maturity had curves in excess of 151°.

Scott and Morgan [61] reported on 28 patients with IIS, of which 14 were followed to skeletal maturity, and were the first to describe the poor cardiopulmonary outcomes in patients with untreated disease. All had severe scoliosis with a mean of 120°. The remaining 14 were still

growing. At 6 years of age, the average Cobb measured 65° with the largest being 112°. Three patients died in the late second and third decades of life from cardiopulmonary complications. All patients in their series had small thoracic cages with reduction in both pulmonary and cardiac function. Younger age at diagnosis and progression were found to be predictors of poorest outcome.

In 1965, Lloyd-Roberts and Pilcher [40] reviewed 100 patients with idiopathic curves who were diagnosed before 12 months of life. Ninety-two of these curves resolved spontaneously. Several other authors have subsequently reported their rates of resolution ranging from 20 to 80 % [14, 32, 40]. James [32] followed 90 patients with nonprogressive curves and found that all resolved by the age of 6 years. Diedrich et al. [14] reported 34 patients with resolving curves followed through maturity and found that none progressed during the adolescent growth spurt. Of the 34, 20 were treated with an orthosis, and no children had significant disabilities related to their spine.

Fernandes and Weinstein [21] reviewed the literature and summarized the data on nonprogressive and progressive infantile idiopathic curves. They identified 573 patients with nonprogressive curves with a male-to-female ratio close to 3:2. Ninety percent were thoracic curves, 80 %, apex left with greatest Cobb angle ranging from 20 to 48°. A large majority had associated intrauterine molding features. Perhaps the most significant finding was age at diagnosis that averaged 5.5 months compared to 12 months among the progressive group. Furthermore, the progressive group showed greater variability compared to historic reports. Gender ratio was closer to 1.2:1 (male to female), 81 % with thoracic curves and 75 % left sided. It is important to recognize that girl infants with right-sided thoracic curves may have a worse prognosis and may not follow the typical rate of spontaneous correction.

Juvenile idiopathic scoliosis differs from IIS in its natural history [38]. The curves progress at a slow to moderate rate [22, 28, 31, 36, 53]. The earlier onset usually leads to more severe deformity than adolescent idiopathic scoliosis. Tolo and Gillespie [66] reported on their series of 59 patients, of which 71 % (42) progressed to require surgery. Similarly, Figueiredo and James [22] found that 56 % (55) of 98 JIS patients progressed. The size of the curve at presentation appears to affect prognosis; Mannherz et al. [44] reported on a series of JIS patients who did not progress. All patients presented with curves <25°.

Pulmonary complications are the most morbid results of untreated infantile scoliosis. As previously described, the spine, chest wall, and respiratory system rapidly develop during the first 5 years of life [15]. Alteration in normal development of one of these can have deleterious effects on the others. Scoliosis that presents and progresses during this time period has a higher chance of causing cardiopulmonary compromise [52]. Infantile scoliosis alters normal development of alveoli and pulmonary vessels resulting in ventilation defects. The severity of pulmonary involvement is directly related to the age of onset of scoliosis. The earlier the onset and progression, the more the disability. Pulmonary dysfunction usually presents as restrictive lung disease with reduced vital capacity (VC), total lung capacity (TLC), and increased residual volume (RV). The loss of compliance of the chest wall and both lungs contributes to the restrictive pattern of disease. Persistence of restrictive lung disease usually results in pulmonary hypertension and corpulmonale. Hypoxemia is related to reduced tidal volume, as gas exchange is typically normal. Respiratory failure is a late development, as these patients have significant pulmonary reserve. This pattern of disease has been consistently shown in the literature; however, it is a rare finding in curves that present after maturation of the lungs (8 years) [12, 34]. Similarly, it differs from thoracic insufficiency syndrome, which presents with respiratory failure at a very early age [9].

10.2.4 Etiology

Browne [8] in 1956 was the first to suggest that infantile scoliosis was initially attributed to an intrauterine packaging problem. He found in his

series that 83 % of infants had some form of intrauterine crowding deformity such as plagiocephaly, plagiopelvy, decreased hip abduction, and abnormal rib molding with infantile scoliosis. Mehta [49] later agreed that intrauterine crowding was responsible. In 1965, Lloyd-Roberts and Pilcher [40] termed this association "molded baby syndrome." Further study would refute this theory, as scoliosis was not found to be present at birth and did not explain the gender difference or the variance in geographic regions. The difference in incidence in Europe and the United States gave rise to an environmental theory. Mau [46] in 1968 proposed that infantile scoliosis was linked to how an infant was positioned for sleeping. In the United States, it was more common to place the infant prone in bed, which decompresses the spine. This is in contrast to the Europeans who were placing their infants supine. Children in this position tend to turn to a slight oblique position with a tendency to lie oblique to the right. He suggested that the molding deformities noted were caused by constant pressure on the soft bones of infants. He also added four other components to the molding theory: unilateral contracture of neck muscles, associated oblique posture of the head, calcaneus foot deformity, and the subsequent development of fixed dorsolumbar kyphosis. These concepts sought to raise awareness and prompt intervention for earlier diagnosis of infantile scoliosis.

The geographic differences further influenced Wynne-Davies [69] to analyze 180 medical records from the Edinburgh Scoliosis Clinic. She identified 114 eligible patients and studied the prevalence of scoliosis between first, second, and third-degree relatives. She analyzed these patients in two groupings: early (before the age of 8 years) and late onset. In the early group, 88 % had left thoracic curves with a slight male predilection. She identified a 2.6 % prevalence of scoliosis in the infantile group compared to 0.39 % of controls, a 30-fold higher risk. The late/adolescent group had an even stronger association at 6.94 %. Plagiocephaly was found in 100 % of patients compared to 11 % among controls. Mental retardation and epilepsy were found in 13 % of patients. Advanced maternal age was also commonly associated with progressive curves.

Ward et al. [67] have made recent advances in genetic testing among the adolescent idiopathic group. Several gene locuses have been identified to strongly predict those patients with progressive curves. In the future, this technology may be expanded to aid in the early detection and treatment of infantile and juvenile scoliosis.

10.3 Clinical Evaluation

10.3.1 History

A thorough and systematic history prior to physical examination is imperative in the diagnosis of infantile and juvenile scoliosis. Careful attention to detail in the history will lead the spine surgeon to pursue further diagnostic testing. Idiopathic scoliosis is a diagnosis of exclusion, and therefore, all etiologies need to be exhausted for accurate diagnosis. Differential diagnoses include neuromuscular scoliosis, syringomyelia, spinal tumor, congenital spinal deformity, other intraspinal anomalies, neurofibromatosis, syndromic disorders, and spinal infection. Patients need to be carefully screened for any other associated anomalies including cardiac defects, history of hip dysplasia, cognitive deficits, congenital muscular torticollis, and other molding abnormalities. This information is often overlooked during an interview, and we recommend having history forms that are conducive to eliciting this information.

During history taking, careful attention should be directed to prenatal history of the mother, including any health problems, previous pregnancies, and medications. Birth history should include length of gestation, delivery type (vaginal or cesarean), weight, and any complications. Like developmental dysplasia of the hip (DDH), there has been an association between scoliosis and breech presentation. Unlike DDH, however, infantile scoliosis is more common in premature low birth weight males. Careful attention should be given to developmental milestones and cognitive function. Wynne-Davies [68] found mental retardation in 13 % of males with infantile scoliosis.

10.3.2 Physical Examination

Physical examination should be performed systematically with special attention given to the skin, head, spine, pelvis, extremities, and neurological examination. Findings in this group of patients are often subtle, and workup is largely dependent on examination findings in order not to miss an underlying cause for scoliosis. The skin examination should include careful inspection for café-au-lait spots and axillary freckling seen in neurofibromatosis. A hairy patch along the spine may indicate spinal dysraphism, and bruising may indicate trauma. The head examination aims primarily to identify any plagiocephaly, where the recessed side of the head is often on the left side of patients. Wynne-Davies [69] found a 100 % incidence of plagiocephaly among the infantile idiopathic group.

The spine examination should begin with inspection, palpation, and careful evaluation of the child's posture, head, shoulder, trunk, and pelvic symmetry. In very young patients, an Adam's forward bend test (looking for prominence of ribs in the thoracic spine or transverse processes in the lumbar spine) is not possible, but the test can be simulated by lying the child prone over the examiner's knee as well as positioning the child with the convex side downward. Lateral pressure in this position allows assessment of curve flexibility. The more rigid the curve, the higher the likelihood of progression. Chest or flank asymmetry and limitation in chest excursion should make the examiner aware of the association with syndromic scoliosis. Abdominal reflex abnormalities should initiate a more thorough neurological examination. Absence of this reflex has been reported as the only objective finding in patients with Chiari malformations [51]. The abnormal reflex is typically found on the convex side of the curve [71]. Further workup is appropriate in this setting with total spine magnetic resonance imaging.

Other physical findings that should not be overlooked include plagiopelvy and developmental hip dysplasia, both with strong associations to idiopathic infantile scoliosis [7, 8, 10, 29, 70]. Hooper [29] found a 6.4 % prevalence of congenital hip dislocation among 156 patients with infantile scoliosis. This is approximately ten times higher than the general population. Wynne-Davies [70] similarly reported on four patients among her infantile scoliosis cohort who had DDH. In 1980, Ceballos et al. [10] reported on 113 patients with a 25 % prevalence of DDH. Interestingly, the dislocations were found mainly among females and with resolving curves. There was no correlation with side of dislocation and direction of curve. Limb length inequality must be ruled out as an etiology for scoliosis. When it is the cause, the lumbar prominence is found on the side of the longer limb. Other means of testing this include a sitting forward bend test or a test by placing a lift under the short limb to equalize limb lengths.

Finally, the physical examination should also include evaluation for different surgical treatments. Current surgical management of IIS and JIS involves the use of implants such as pedicle screws, hooks, and spine-to-spine and rib-to-spine devices. These implants can be prominent and require attention to detail during the physical examination. Patients with very short stature may not be able to accept a rigid 70- or 90-mm rod connector. Other patients may have significant skin contractures or have had previous surgeries that may complicate the use of spinal implants and may prompt consultation with plastic surgery.

10.4 Diagnostic Testing

10.4.1 Radiologic Evaluation

Plain radiography is a simple and reliable tool in the workup of a child with suspected scoliosis. Patients are typically diagnosed in the first 6 months to 1 year of life, and early recognition and treatment are essential for optimal outcomes. Radiographs will help rule out congenital scoliosis as well as establish baseline measurement for future comparisons. Treatment decisions are traditionally based on progression of Cobb angle and rib vertebral angle difference (RVAD) obtained at subsequent visits. Progression has

been associated with compensatory curves (including lumbar, double thoracic, and thoracic), greater vertebral rotation, and shorter length of curves.

High-quality radiographs are essential for thorough radiographic analysis. Initial evaluation should include postero-anterior (PA) and lateral radiographs of the spine (including cervical spine and pelvis). In children too young to stand, films should be obtained supine. Special attention should be paid to the cervical spine for anomalies, as well as to the lumbosacral junction for spinal dysraphism, and the pelvis and hips to ensure a reduced position of the hips. Measurements should include both Cobb angle and RVAD (Fig. 10.1a). Mehta [49] is credited for developing this powerful tool for predicting progression of infantile curves. Out of frustration with the inability to predict progression with Cobb measurements, she evaluated the relationship of the rib attachment to the vertebral body. She noted variability in the takeoff angle of the ribs from the convex vs. the concave side of the curve. The rib vertebral angle measures the angle of a line drawn perpendicular to the apical thoracic vertebra end plate and a line drawn down the center of the concave and convex ribs. The RVAD is calculated by subtracting the convex from the concave angles. An RVAD of less than 20° indicates a curve that is most likely to resolve (85–90 %), while an RVAD of 20° or more is frequently associated with progression. She also described a second radiographic parameter to assist in prediction known as the phase of the rib head (Fig. 10.1b, c). This radiographic tool uses the relationship of the head and neck of the rib to the vertebral body, at the apex of the convexity of the scoliosis. In phase 1, there is no overlap of the rib head or neck on the apical vertebra. In this group of patients, the RVAD should be measured to detect progression. In phase 2, the head or neck of the rib is overlapped on the apical vertebra. It has been shown that phase 2 rib head is a certain predictor for progression and RVAD does not need to be measured. Mehta [49] reported on 46 infantile patients with phase 1 rib heads whose scoliosis resolved. She found that 83 % had an RVAD of less than 20°. Of the remaining patients

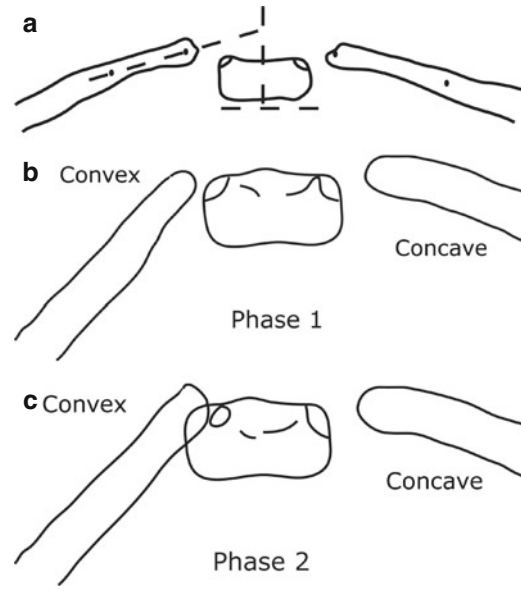

Fig. 10.1 (**a**) Rib vertebral angle difference (RVAD). (**b**) Phase of rib head: phase 1. (**c**) Phase of rib head: phase 2 (Redrawn from Ref. [49])

with an RVAD of 20° or more, the angle was found to consistently decrease with follow-up. The decrease in RVAD also preceded the decrease in Cobb angle. Of the group with progressive curves, 84 % had an initial RVAD of 20° or more (range, 18–30°).

Ceballos et al. [10] corroborated Mehta's findings reporting 92 % of their resolving curves having an RVAD of 20° or less. Of the remaining 8 % with an RVAD greater than 20°, all showed improvement at the 3-month follow-up. Robinson and McMaster [57] in 1996 found that the curves that progressed among their 109 patients had a mean initial RVAD of 31°, while those that resolved had a mean of 9° on initial examination.

Mehta [49] recognized a special radiographic feature among the less common (and more aggressive) double major and lumbar curve patterns. She recognized that the RVAD at the apical thoracic vertebra was frequently less than 20° and found that there is significant asymmetry at the 12th vertebra. Here, she found the rib on the concave side becoming more vertical than the rib on the convex side, making the RVAD negative. The 12th rib is initially part of the upper curve

but becomes the apex of a secondary curve developing caudally to the first. Consequently, the rib that is on the concavity of the upper curve drops secondary to the progression of the vertebral rotation and increases in magnitude of the caudal curve.

10.4.2 The Role of Advanced Imaging and Neural Axis Abnormalities

The role of advanced imaging in infantile and juvenile scoliosis is directly related to the presence of neural axis abnormalities. As IIS and JIS are a diagnosis of exclusion, all attempts must be made to identify possible etiologies. The incidence of neurological abnormalities has been reported as high as 20 % in patients under the age of 10 years [20, 37, 39, 49]. Lewonowski et al. [39] reported a magnetic resonance imaging (MRI) study of 26 consecutive patients with idiopathic scoliosis under the age of 10 years. They found five patients (19 %) with neuropathology and only two patients with atypical curves. Four of their patients were infantile, and two patients had abnormal findings: a 4-month-old boy with a terminal lipoma and a 3-year-old girl with a syrinx.

Gupta et al. [24] conducted a prospective and retrospective MRI study to evaluate the prevalence of neural axis abnormalities in patients 10 years of age or younger with idiopathic scoliosis and a normal clinical examination. In the prospective arm, he followed 34 patients with a mean age of 9 years and found abnormalities in six patients (18 %). Within this group, six patients were infantile and three patients had identifiable neuropathology. Among the 64 retrospective patients, 20 % were found to have neural axis pathology.

Most recently, Dobbs et al. [17] in multicenter study identified 11 of 46 infantile scoliosis patients with neural axis abnormalities. All patients were clinically asymptomatic and had curves of 20° or less. Five patients had an Arnold-Chiari type-I malformation, three with syringomyelia, one with a low-lying conus, and one with a brain tumor. Of these ten patients, eight required surgical intervention. On the basis of the findings of this paper and other reports, it is our recommendation that all patients with IIS or JIS with a curve of 20° or less have both a brain and a complete spine magnetic resonance imaging (MRI).

Other imaging modalities exist to aid in management and provide continued relevant information in the care of these children. Computed tomography (CT) scans can be helpful for preoperative evaluation in selected patients where the spine will be instrumented. Pedicular anatomy and bony anomalies are made very clear. CT scans can also be used to assess the three-dimensional lung volumes and can be a marker of treatment; however, their use must be weighed against the risk of significant associated radiation exposure.

10.5 Management Themes (Fig. 10.2)

10.5.1 Selecting Surgical Candidates

Management of children with early onset idiopathic scoliosis is based on anticipated or actual curve progression. Mehta's [49] prognostic criteria, as discussed earlier, are very helpful in identifying curves at risk. Curves with an RVAD of less than 20° and a Cobb angle of less than 25° are at low risk of progression. These patients are safely treated with observation; however, they should be followed clinically every 4–6 months for progression. Once the curve has resolved, the follow-up interval can be extended to 1–2 years. We recommend following these patients to maturity to ensure that there is no recurrence during the adolescent growth spurt. Diedrich et al. [14] reported on 25-year follow-up of infantile scoliosis, validating the use of RVAD, and demonstrated that there was no advantage to supine plaster bed treatment over physiotherapy, in regard to time to resolution or functional outcome.

Infants with an RVAD of 20° or more or a phase 2 rib–vertebral relationship and a Cobb angle between 20° and 35° have a higher risk of progression. This group of patients should be followed closely at 4- to 6-month intervals for clinical and radiographic evaluation. Active treat-

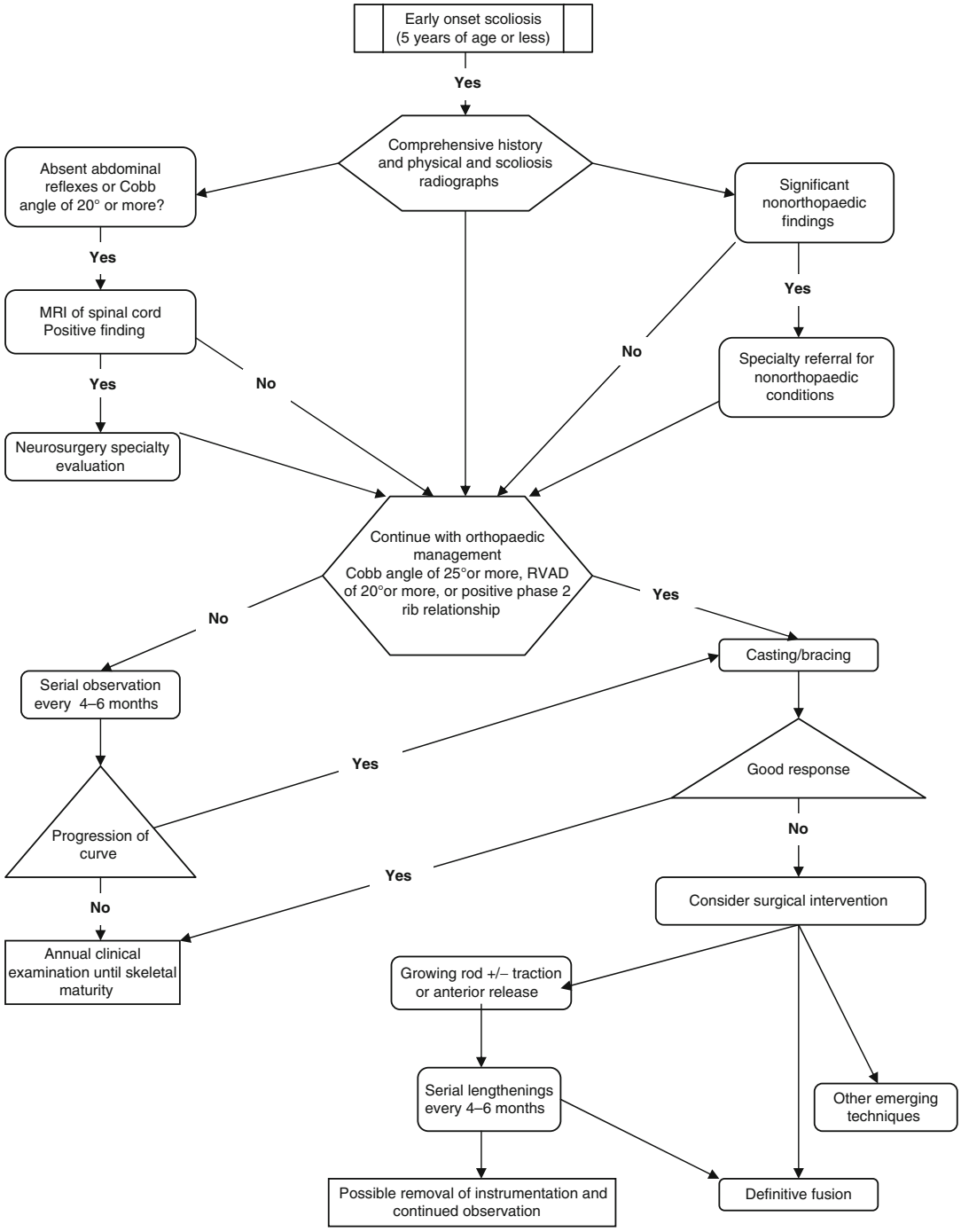

Fig. 10.2 Treatment algorithm for infantile and juvenile idiopathic scoliosis (Adapted from Ref. [23])

ment should be initiated when progression of Cobb angle of 5° or more is documented over 1 year [1]. Active treatment at this point is usually in the form of casting or bracing, which will be discussed thoroughly in separate chapters.

10.5.2 Surgical Treatment: Historic Perspective

The goals of surgical treatment of idiopathic early onset scoliosis are multifold: to stop curve progression and to allow for maximum growth of the spine, lungs, and thoracic cage. Surgery is often recommended in children with progressive curves of 46° or more; however, there are other factors that influence decision making. The risks and benefits of curve correction vs. continued growth should be considered by the treating surgeon to make a final decision. This statement reflects the current trend toward more aggressive operative management since the techniques for fusion-less surgery have become refined and the natural history of this disease, more clearly understood.

Historically, the goals of surgery were a straight shortened spine rather than a deformed spine of near normal length. Isolated posterior spinal fusion in this age group quickly went out of favor, after Dubousset et al. [18] described the crankshaft phenomenon. This phenomenon seen in skeletally immature patients describes progression of deformity following posterior spinal fusion due to continued anterior growth of the spine. Sanders et al. [59] further correlated open triradiate cartilage and Risser 0 to high risk of crankshaft in the presence of an isolated posterior spinal fusion. Anterior arthrodesis was therefore recommended, in addition to posterior fusion to prevent crankshaft. Anterior and posterior fusion, however, results in a significant amount of height loss and thoracic underdevelopment. As discussed earlier, Dimeglio [15] nicely outlined spinal growth throughout childhood with two noticeable peaks of growth (0–5 years and 10–15 years). Using his formula for calculating normal growth, expected loss of height can be determined for patients treated with anterior–posterior fusion. Winter [68] similarly described a formula for calculating amount of projected height loss. To calculate projected shortening in centimeters, multiply 0.07 × number of segments fused × number of growth years remaining. These data are very valuable in educating family and caretaker of the potential ramifications of fusion in this very young patient population. It should also be noted that the effect of fusion on the spine could have morbid effects on lung and thoracic cage development. This has been a motivating factor over many decades to devise other surgical methods that avoid circumferential fusion.

Over 45 years ago, Roaf [56] attempted to modulate spine growth, much like one would modulate an angular deformity in a pediatric lower extremity with hemiepiphysiodesis. He proposed that the spinal deformity was the result of asymmetric growth between the convex (faster growing) and concave (inhibited) sides of the curve. His technique of modulation involved ablation of the convex epiphyseal cartilage and adjacent discs at the vertebrae near the apex of the curve. Only 23 % of his treated patients showed improvement of Cobb angle, while 40 % showed little or no improvement (Cobb angle <10° change). Marks et al. [45] built upon this idea and used hemiepiphysiodesis and simultaneous Harrington internal fixation. No significant improvement was measured in 13 consecutive patients with 12 demonstrating progression of deformity.

Harrington [27], in 1962, described a fusionless technique in 27 idiopathic and postpolio patients, placing a single distraction rod on the concavity of the curve connected to hooks at both ends. The hooks and rods were placed after a subperiosteal approach to the spine. The idea was to instrument the spine without arthrodesis in an attempt to preserve spinal growth, correct deformity, and control the residual deformity. Although no longitudinal results were reported, he believed that children under 10 years could be managed with instrumentation alone and those 10 years and older required arthrodesis.

Moe et al. [50] modified the technique described by Harrington and limited subperiosteal exposure to the site of hook placement and passed the rod subcutaneously. Furthermore, they modified the rod to have a smooth, thicker central portion to prevent scare formation to the threads and allow for sagittal contouring. Patients were lengthened when a loss of Cobb angle >10° occurred. Of the two patients treated with idiopathic infantile scoliosis, both were reported as

having a notable decrease in curve magnitude. They, furthermore, reported a complication rate of 50 %, including rod breakage and hook dislodgement from the rod or the lamina.

In 1997, Klemme et al. [35] reported on 20-year experience of the Moe technique. Sixty-seven patients were followed from initial instrumentation to final fusion, with an average of 6.1 procedures per patient. Curve progression was arrested or improved in 44 of 67 patients with an average curve reduction of 30 %. Of the remaining 23 patients, 12 were neuromuscular, and the curves progressed on average 33 %.

In 1977, Luque and Cardosa [42] described their technique of fusionless treatment of scoliosis with segmental spinal instrumentation. In 1982, Luque [41] modified this technique by adding sublaminar wires and replacing the Harrington rod with L-shaped rods, later to be known as the Luquetrolly. His initial series included 48 paralytic patients who grew by an average of 4.6 cm over the immobilized segment with an average curve correction of 78 %. This system became less favored after reports that subperiosteal exposure and sublaminar wire passage created scar tissue and weakened the lamina, which made revision and later definitive fusion difficult. There were also several reports of spontaneous fusion and substantially less growth preservation than predicted. These findings were attributed to the exposure that was required at each level to pass wires [54].

Patterson et al. [53] combined segmental spinal instrumentation with anterior apical convex growth arrest and fusion in 9 of 13 patients who had previously undergone surgery at an average age of 5 years and 5 months. Curve correction averaged 46 % at 2-year follow-up. Less curve deterioration was identified in those patients who had anterior apical growth arrest compared to those who had segmental instrumentation alone.

In 1999, Pratt et al. [55] performed a retrospective review of patients treated with Luque trolley instrumentation with and without convex epiphysiodesis in 26 patients. Eight were treated with Luque trolley alone, and all showed significant curve deterioration. Of those treated with combined convex epiphysiodesis and Luque instrumentation, the Cobb angle worsened in seven of 13, remained unchanged in four and improved for two. Growth was found to be 49 % among those predicted in the Luque trolley alone group and 32 % among those undergoing combined surgery.

Blakemore et al. [6] further reported periodic lengthening with a submuscular rod with and without apical fusion. Apical fusion was performed on curves 70° or more and in those whose curves were stiff on bending radiographic testing. The rod was placed within the muscle above the spine periosteum, placing the rod closer to the spine for better contour and alignment without inducing spontaneous fusion. He reported on 29 children, ten idiopathic, all treated in a Milwaukee brace postoperatively. Mean Cobb angle improved from 66 to 38° immediately postoperatively with most recent follow-up showing a slight deterioration to 47°. Complication rate was 24 % including hook dislodgement (5), rod breakages (3), and superficial wound infection (1).

10.5.3 Current Approaches to Surgical Management

Once the decision for surgery has been made, several factors have to be considered before choosing the correct surgical approach. The rigidity of the curve plays an important role in decision making, as curves that have little flexibility will not likely be as amenable to a growing construct alone. In this situation, there may be a role for anterior release prior to posterior fusionless surgery. Marks, in unpublished results, discussed the use of annulectomy vs. nucleotomy as anterior release options. No long-term results exist, however, to make any definitive recommendations (D Marks, 2009, personal communication).

The next decision to make is which lengthening procedure is ideal for the patient. Salari et al. [58] recently reported on the results of a survey sent to 40 qualified surgeons on ideal treatment of 11 different case scenarios of infantile scoliosis. Seventeen surgeons responded with a wide

variation in treatment recommendations for each patient scenario. The most common treatment selected was a dual growing-rod construct (56.7 %), followed by nonoperative management (16.6 %), SHILLA (15.5 %), VEPTR (7 %), fusion or resection, and immediate fusion (4 %). This study is important to highlight the lack of standardized treatments offered to our patients by highly qualified surgeons [57].

The next two sections briefly describe the various fusionless surgeries. They are subdivided into two categories: distraction-based growing rods and growth-directed surgery. VEPTR, a form of distraction-based growing rod, will be discussed in a separate chapter.

10.5.4 Distraction-Based Growing Rods

The unpredictability and high implant-related complication rate associated with single rod distraction techniques led Akbarnia and Marks [3] to popularize a dual growing rod technique, building on concepts formulated by Asher (Fig. 10.3a–d). Subperiosteal dissection is limited to the proximal and distal foundations (anchor sites). Hooks or pedicle screws are placed on both ends over two or three spinal levels. Foundation sites are fused using local bone graft supplemented with synthetic graft. Upper and lower contoured 3/16 in.-diameter rods are placed submuscularly on both sides of the spine. The rods are joined on each side with extended tandem connectors placed at the thoracolumbar junction to avoid disturbing sagittal balance. The first lengthening is typically performed at the index procedure. A distractor designed to fit within the longitudinal opening in the tandem connector is used at time of lengthening that typically occurs at 6-month intervals starting with the index surgery. The intent of the original lengthening is to obtain modest correction of the scoliotic curve without unduly stressing the foundations. We have found approximately 50 % correction of coronal Cobb angles at the original surgery. More aggressive lengthening can be performed starting with the first lengthening after fusion. Somatosensory-evoked potential monitoring is performed during each lengthening. Lengthening can be performed as outpatient surgery with appropriate anesthesia and

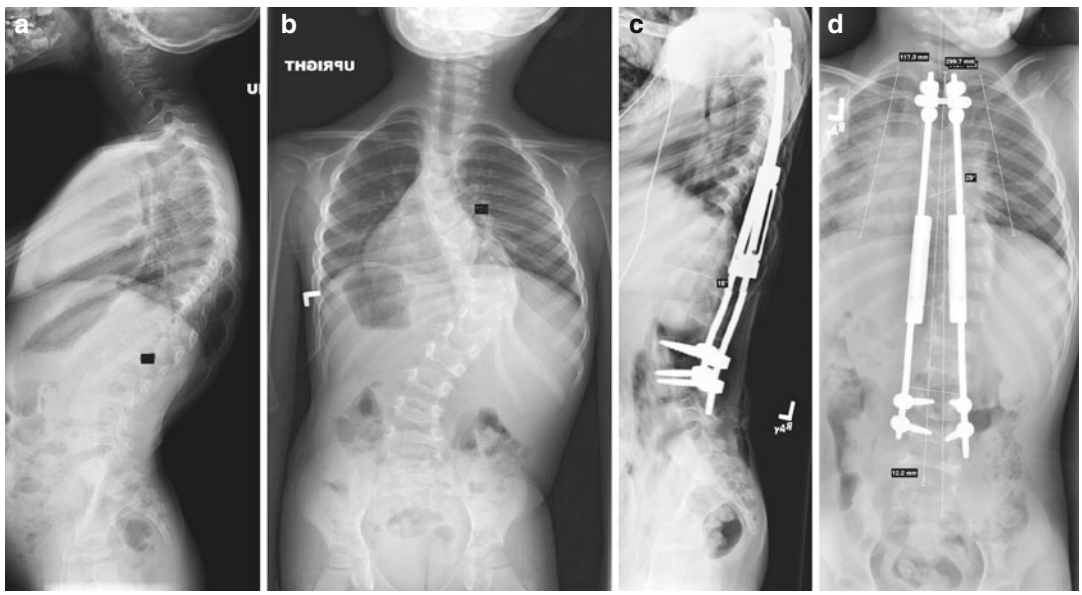

Fig. 10.3 (**a**, **b**) Severe progressive scoliosis in a 4-year-old patient with idiopathic infantile. (**c**, **d**) Post-initial surgery radiographs

nursing support. Bracing is utilized until fusion is achieved at the foundation sites.

Recent advances in technology have led to the development of magnetically controlled growing rods (MCGRs). The index surgery for placement of the MCGR is similar to placement of dual growing rods with the exception that the implant consists of a single preassembled rod [11]. The use of MCGR has become the preferred method of treating IIS and JIS when feasible, to avoid repeat exposure to anesthesia and surgery traditionally associated with growing rod surgery.

Akbarnia et al. [2] reviewed 13 patients with no previous surgery and noncongenital curves who were followed to final fusion. They found a mean spinal growth of 5.7 cm during a 4.4-year treatment period. The curve improved from 81 to 36° after initial surgery and to 28° at final fusion. T1–S1 length improved from 24 to 29 cm after initial surgery to 35 cm at final fusion. Those patients lengthened at 6-month or less intervals experienced significantly more growth and curve correction than those lengthened less frequently [2].

A recent report by Sankar et al. [60] reviewed 782 growing rod surgeries in 252 patients where neuromonitoring was performed. Surgeries included 252 primary rod implantations, 170 implant exchanges, and 362 lengthenings. Neuromonitoring changes occurred in two primary implant surgeries (0.8 %), one implant exchange (0.6 %), and one lengthening (0.3 %). The change noted in the case of implant exchange also resulted in a clinical deficit, which resolved within 3 months. The monitoring change that occurred in the lengthening was in a child with an intracanal tumor that also had a change during the primary surgery. The final recommendation was that the overall rate of neuromonitoring change seen in primary and implant exchange surgeries justifies its use. No definitive recommendations could be made for lengthenings because of sample size.

Akbarnia et al. [4] reported on a multicenter study with 2-year follow-up (24–111 months) of 23 patients, seven of which had idiopathic infantile scoliosis. The average age at initial surgery was 5 years and 5 months, with an average of 6.6 lengthenings. Mean Cobb angle improved from 82 to 38° following initial surgery and 36° at latest follow-up. Growth averaged 1.21 cm per year as calculated by T1–S1. Seven patients completed treatment and averaged 11.8 cm of total growth (T1–S1) from preoperative to postfinal fusion (1.66 cm per year). Among 14 patients with thoracic curves, the space available for lung as described by Campbell et al. improved from 0.87 preoperatively to 1.00 at latest follow-up or final fusion. Complications occurred in 11 of 23 patients between initial surgery and final fusion. They included three anchor (hook or screw) displacements, two rod breakages, two deep wound infections, four superficial wound problems, one crankshaft, and one junctional kyphosis requiring an extension of instrumentation. Although the complication rate is high, the authors contested that it is safe and effective and carried with it a lower complication rate than single rod systems.

Thompson et al. [64] compared the results of single and dual growing rod systems in 28 patients followed to definitive surgery. Five had a single rod construct with anterior and posterior apical fusion, 16 had single rod without apical fusion, and seven had dual rod without fusion. Mean Cobb angle, respectively, improved from 85° to 65°, 61° to 39°, and 92° to 26°. Spinal growth, respectively, was 0.3, 1.0, and 1.7 cm per year. The authors concluded that the improved results seen in dual rod systems are likely attributable to its greater strength and more frequent lengthening.

Mahar et al. [43] published results of a biomechanical study investigating the construct of the foundation in a porcine model. They investigated four constructs: (1) hook–hook with cross-link, (2) hook–screw with cross-link, (3) screw–screw with cross-link, and (4) screw–screw without cross-link. They found that a four-screw construct in adjacent vertebral bodies provides the strongest construct in pullout testing. A cross-link did not provide any additional strength to the all screw construct. They also found that the hook construct had significantly higher pullout strength in the lumbar spine compared to the thoracic spine.

In a multicenter study, Bess et al. [5] (Growing Spine Study Group) reported on complications in

910 growing rod surgeries in 143 patients with minimum 2-year follow-up. They divided the group as single ($n=73$ patients) or dual rod ($n=70$ pts) and subcutaneous ($n=54$) or submuscular ($n=89$). Complication rate per surgery was <20 %. Complication rates were equivalent among single and dual rod constructs. Significance was found in a number of implant-related complications requiring unplanned return to the operating room for single rod constructs compared to dual. The subcutaneous group had more complications per patient (1.6 vs. 0.99) and more wound problems (13 vs. 4 patients). Furthermore, subcutaneous placement of dual rods had higher overall complication rate, higher wound problems, prominent implants, and patients undergoing implant-related unplanned return to the operating room. The conclusion was that the overall complication rate is comparable to historic reports; dual rods reduce unplanned trips to the operating room, and submuscular position of implants is preferred over subcutaneous placement.

10.5.5 Growth-Directed Surgery

Growth-directed surgery is the phrase used to describe procedures where reduction of the spinal deformity relies on the remaining growth available. The classic example of this is the Shilla procedure described by McCarthy et al. This surgery involves limited instrumentation and reduction of the apical segment with specialized polyaxial Shilla screws that house two rods and allow those rods to glide within the construct. The concept is to improve the deformity of the spine by naturally directed growth along a new path (the rods that are placed).

McCarthy et al. [47] recently reported on ten patients with 2-year follow-up. Three of these patients were either infantile or juvenile idiopathic scoliosis. Initial curve correction went from 70.5° (40–86°) to 27° (5–52°) at 6 weeks, and 34° (18–57°) at 2-year follow-up. Two patients had a staged anterior apical release. Complications included rod revision for growth off the end of the rods, rod exchange for a shorter rod due to prominence, one broken rod, and two wound infections for a total of five surgeries among all ten patients beyond the index procedure. It was predicted that this same group of patients would have required 49 additional surgeries in a distraction-based growing rod model. For more details regarding this technique, please see Chap. 41.

10.6 On the Horizon and Conclusion

Significant strides have been made in the last decade regarding understanding and management of idiopathic early onset scoliosis. This unique disease entity, however, still leaves many areas undiscovered including genetic etiology, accurate scientific predictability of progression, ideal treatment for individual curves, and refinement in surgical technique.

The ideal surgery would include a minimally invasive approach with a durable and inert implant that rarely requires reoperation. Takaso et al. [63], in 1998, reported on the development of a rod containing a direct-current motor attached to a radio-controlled receiver. They performed successful correction of experimental scoliosis in beagles. The main issues with this device were its size (16 mm) and the placement of the receiver in the abdominal cavity. Akbarnia has recently explored the idea of remote lengthening, and animal studies are under way investigating this promising technology.

Ward et al. [67], as discussed in another chapter, are currently studying this very unique patient population to identify any markers for progression and the genetic basis of IIS and JIS. It is the hope of all treating physicians that success in this arena will be as productive as it has been in identifying these markers in adolescent idiopathic scoliosis.

In conclusion, idiopathic early onset scoliosis is a disease entity that, if left untreated, can result in devastating and life-threatening complications. Early recognition and timely treatment are essential to management and for good outcome. Exciting new technology and improved surgical

technique will result in lower complication rates, avoidance of natural history, and ultimately improved patient outcome [61, 65].

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Congenital Scoliosis

11

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Key Points

- Congenital scoliosis (CS) is caused by vertebral malformations, which interrupt the longitudinal growth of the spine.
- The progression of CS depends on the type of the anomaly.
- The evaluation of a patient with CS should include detailed spine and neurological examination, radiographic evaluation, and investigations for associated anomalies.
- There are many treatment alternatives for CS. The age of the patient, type of anomaly, experience of the surgeon should be considered while choosing the most appropriate method of treatment.

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11.1 Introduction

Congenital scoliosis (CS) is a lateral curvature of the vertebral column. It is caused by vertebral malformations, which interrupt the longitudinal growth of the spine. Congenital vertebral malformation (CVM) is always present at birth; however, scoliosis develops in some patients as the spinal column grows longitudinally.

CS is caused by early embryologic development failure of vertebral column. Failure of formation, segmentation, or both can cause anomalous development [4]. Either these malformations can be a benign anomaly that results in mild curvatures and does not affect spinal balance, or they can be one with high potential to deteriorate and affect spinal balance [29].

The exact incidence of CS is unknown [36], while the prevalence rate of congenital scoliosis is thought to be approximately 1 in 1000 live births [16]. It occurs more often in girls than in boys, with a ratio of 2.5–1 [32].

11.2 Etiology

The causes of CVM have not yet been investigated in detail. The etiology is thought to be multifactorial. It is believed that the genetic and teratogenic factors play a role in the development of CS. Vertebral malformations may be an isolated finding or, in some cases, may occur with other cardiac, renal, and intraspinal malformations. They may also occur as part of an underlying chromosome abnormality or syndromes such as Alagille, Jarcho-Levin, Klippel–Fiel, Goldenhar, Trisomy 18, diabetic embryopathy, and VACTERL (vertebral, cardiac, renal, limb anomalies, anal atresia, tracheo-esophageal fistula) association [1, 21]. Ingestion of antiepileptic drugs, alcohol during pregnancy, maternal insulin-dependent diabetes mellitus, and gestational diabetes have also been accused as a possible cause of CVM [59]. The risk of having CVM is also increased in monozygotic and dizygotic twins. Vertebral segmentation defects associated with rib anomalies have been reported in spondylocostal dysostosis which is an autosomal recessively inherited disorder. These patients have short stature, shortened trunk, and protuberant abdomen in addition to CVM [23].

Carbon monoxide (CO) and hypoxia are the two most common teratogenic factors believed to be possible causes of congenital vertebral malformations. CO is a well-known teratogen [51]. It is a colorless and odorless gas and has 200–300 times greater affinity for hemoglobin than oxygen. Therefore, CO binds to hemoglobin in the lungs easily, while it does not unbind in peripheral tissues, thus interfering with tissue oxygenation [19]. CO crosses the placenta. Even though how the CO leads to spinal malformations is unknown, the relation between CO exposure and spinal malformations has been shown in different studies. Studies of maternal carbon-monoxide exposure have demonstrated vertebral and rib malformation in offspring of mice and rabbits. Loder et al. found spinal malformations in 70 % of offspring of mice exposed to 600 ppm CO on gestation day 9 [38]. The dose and the timing of CO exposure seem critical. Maximum effect is seen on day 9 of gestation, which corresponds to fourth week in human embryonic fetal life, with an exposure to 600 ppm of CO [19]. Hypoxia was shown to be an etiologic factor in experimental animal models. These reports showed a relation between time and dosage of hypoxia and vertebrae and rib malformations. The malformations were segmentation and formation defects, similar to those found in man [31, 50].

Based on mouse studies, a series of candidate genes, known to cause vertebral malformations, has been identified [5, 22]. *Wnt3a*, *PAX1*, *DLL3*, *Sim2 genes* have been proposed to be responsible for vertebral malformation in mouse models. Mutations of these genes may disrupt early somite development, leading to rib fusion and deficient development of the anterior vertebral elements and failure of formation of the dorsal neural arches [22, 24–26, 40].

11.3 Classification

CS is often rigid, while some forms are inclined to deteriorate, and others do not disturb spinal balance. Therefore, it is crucial to anticipate when a

CS is at risk for rapid progression. A proper classification system of the malformations is mandatory, for an accurate estimation of the progression risk. CS is mostly classified under three main categories: failure of formation, failure of segmentation, and complex malformations. MacEwen described a classification system for CVM, which was modified by Winter et al. [60] in 1968 and was later accepted by the Scoliosis Research Society.

11.3.1 Failure of Formation

Failure of formation arises as a result of an absence of a part of the vertebra. Anterior, anterolateral, posterior, posterolateral, and lateral region of the vertebral ring may be affected [37]. Failure of formation may be incomplete or complete.

Wedge vertebra is an incomplete formation defect. The anomalous vertebra has two pedicles, but one side of the vertebrae is hypoplastic. A butterfly vertebra is another type of incomplete formation defect. These vertebras have two pedicles with hypoplasia or cleavage in the center of the vertebral body. Plain frontal radiographs show that these bones have the shape of a butterfly. There are two types of butterfly vertebra: symmetrical and asymmetrical.

Hemivertebra is a complete formation defect. The anomalous vertebra has one pedicle, and there is only half of the vertebra. There are three types of hemivertebra (HV): fully segmented, partially segmented, and unsegmented. Fully segmented hemivertebra has growth plates, both cranially and caudally. In this way, a fully segmented hemivertebra continues to grow longitudinally both cranially and caudally and has great effect on spinal balance. Alternatively, a nonsegmented hemivertebra is not separated from cranial and caudal vertebra; thus, it has lower growth potential and less effect on spinal balance. A partially segmented hemivertebra has functional disc only on one side and fusion on the other side. In hemimetameric shift, a hemivertebra is counterbalanced by contralateral hemivertebra on the other side of the spinal column. They are separated by at least one normal vertebra and are most commonly seen in the thoracic region [52].

11.3.2 Failure of Segmentation

Segmentation defects present an abnormal connection or bar between vertebras. The bar stops the growth on the affected side and causes a tethering effect. A bilateral bar (block-vertebra) has a significantly lesser effect on spinal balance [29] and the progression of spinal deformity. They may occur in combination with congenital rib union, and fused ribs may deteriorate spinal deformity as well as chest wall deformity [34, 56]. If the bar is bilateral (block vertebra), it has much less effect on spinal balance [29].

11.3.3 Complex Malformations

Complex malformations contain segmentation and formation defects in the same patient. These patients have a great risk for rapid progression. Since only 30 % of the spinal column is ossified at birth, it is difficult to diagnose these kinds of malformations at the beginning of life. This category includes a unilateral unsegmented bar with contralateral hemivertebra, which represents the most severe progression of scoliosis [41].

Oftentimes, the posterior element anatomy is neglected in classification systems of congenital scoliosis. Understanding the posterior anatomy is especially helpful for planning the surgery. Nakajima et al. [45] analyzed the three-dimensional morphology of congenial vertebral anomalies in formation failure and detected two anatomical characteristics. The first is a variety of posterior structure; posterior elements that may be totally normal, hemi-laminas, fused laminas, or bifid areas with exposed neural structures (Fig. 11.1). The second is discordancy between the anterior structure and the posterior structure (Fig. 11.2). They emphasized that discordant vertebral anomalies may lead to selection of the wrong level of surgery and the importance of a three-dimensional analysis of the congenital scoliosis in a group of patients.

Kawakami et al. [35] indicated that failure of formation in Winter's classification includes those with segmentation failure such as nonsegmented hemivertebra, although those can be classified

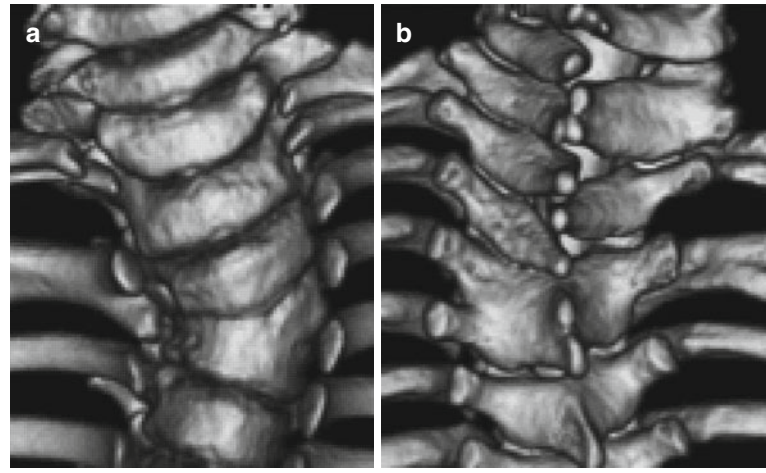


Fig. 11.1 The anterior formation defect (**a**) is accompanied with fused lamina on the posterior spine (**b**)

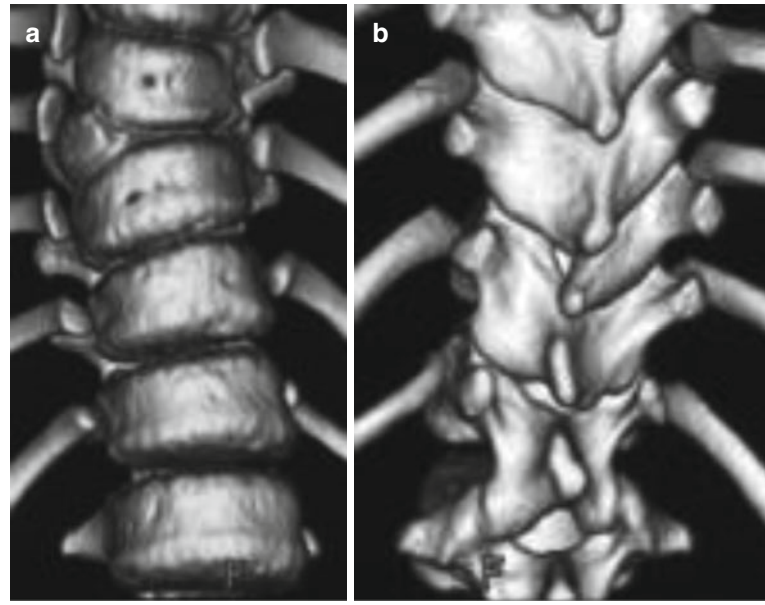


Fig. 11.2 The level of the anterior (**a**) and the posterior (**b**) vertebral malformation is discordant

into failure of segmentation or mixed type. The report classified congenital spinal deformity into the solitary simple, multiple simple, multiple complex, and pure type of segmentation defect based on these three-dimensional morphological findings. This new classification is based on the concept that vertebral anomalies in failure of segmentation do not have any type of abnormal vertebrae with the characteristics of formation failure (Fig. 11.3). Although this classification is slightly more complicated, it demonstrates necessity of a preoperative three-dimensional morphological analysis for determining the optimal surgical strategy for treating congenital vertebral anomalies.

Winter et al. Classification [60]

I. Unclassifiable: There is a collection of many types of segmentation defects. There is no dominating type.
II. Fusion of ribs.
III. Unilateral failure of formation of a vertebra, partial: this produces a wedge or trapezoid-shaped vertebra. A vestigial pedicle may be present.
IV. Unilateral failure of formation of a vertebra, complete: this produces a hemivertebra.

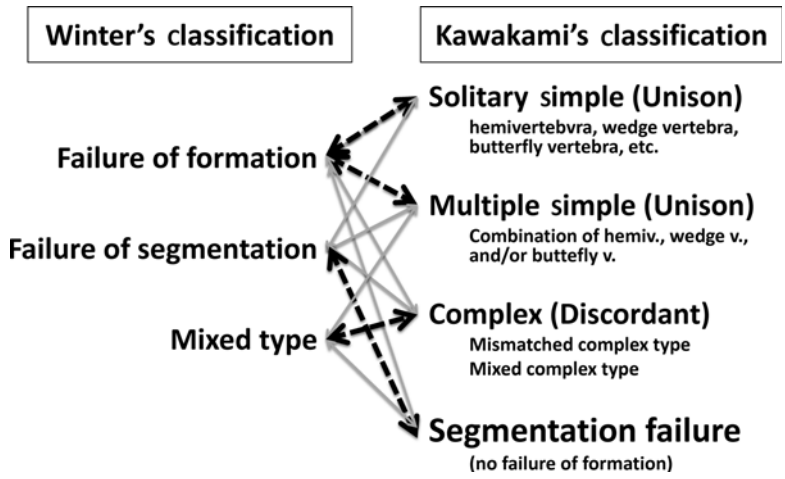


Fig. 11.3 The new classification of congenital spinal malformation by Kawakami et al. and Winter et al.

V. Bilateral failure of segmentation: this refers to the condition in which there is absence of the disc space between adjacent vertebral bodies.
VI. Unilateral failure of segmentation: this produces an unsegmented bar and may involve two or more vertebrae and only the bodies or only the posterior elements.

Kawakami et al. Classification [35]

Type 1. Solitary simple type: there is only one abnormal vertebra in the entire curve.

Type 2. Multiple simple type: there are multiple abnormal vertebrae with a consistent anterior and posterior structure.

Type 3. Multiple complex type: there are multiple abnormal vertebrae with a combination of formation and segmentation defects with or without discordancy.

Type 4. Segmentation failure type: there are multiple abnormal vertebrae without any type of formation failure.

11.4 Natural History

Longitudinal spine growth comes from superior and inferior end plates. Curve progression is caused by unbalanced growth of one side of the spine. Well-formed and normal appearing discs suggest healthy growth plates and potential for asymmetric growth. On the other hand, the presence of bar or fused ribs is a sign of restricted growth on this side and may cause progressive deformities [29]. Therefore, the progression of CS depends on the type of the anomaly. The location of vertebral malformation and the growth potential of the patient are the other two most important factors in predicting the deterioration potential of the curve.

The two most important natural history studies were published by Winter et al. [60] in 1968 and McMaster and Ohtsuka [41] in 1982. Winter et al. [60] followed 234 patients with CS and found that thoracic and thoracolumbar curves progressed more than cervico-thoracic and lumbar curves did. A mild cervico-thoracic curve might cause serious cosmetic deformity because of head tilt, prominence of the neck, and dropping of one shoulder. It was also found that the rate of progression was not related to the severity of the curve, since some of the mild curves progressed more rapidly than the severe ones. Progression was most likely to occur when there were multiple unilateral anomalies in the thoracic spine. It was also reported that most severe deterioration of the curve was seen during preadolescence and infancy period. McMaster and Ohtsuka [41] followed 216 patients for a mean of 5.1 years and reported that the rate of curve deterioration was found to depend on both the level and the type of malformation. For each type of deformity, the deterioration of the curve was less severe in upper thoracic regions, more

severe in mid-thoracic region, and worst in thoracolumbar region. Block vertebra and bilateral failure of segmentation are the most benign forms of anomaly, and the progression rate is less than 2° per year. Wedge vertebra, hemivertebra, and unilateral bar cause more severe deformities, respectively. A unilateral bar and contralateral hemivertebra were the most severe anomalies and have a progression rate of 5–10° per year. On the other hand, Winter et al. [62] later reported spontaneous improvement of scoliosis in seven patients with a hemivertebra in a review of 1250 with congenital spinal deformity. Therefore, predicting curve progression is still difficult. This may be due to the variety of not only vertebral body morphology but also posterior structure. Further analysis of the three-dimensional vertebral morphology using computed tomography (CT) and magnetic resonance imaging (MRI) images may reveal more precise determination of the natural history of each type of vertebral anomaly.

11.5 Patient Evaluation

The evaluation of a patient with CS focuses on physical examination including detailed spine and neurological examination, radiographic evaluation, and investigation of associated anomalies.

11.5.1 Physical Examination

Since the spinal growth is a major concern in CS, physical examination should start with recording the sitting and standing height and weight. The growth of the child should be monitored, as there is a close relationship between growth and curve progression, as discussed in natural history part.

CS may cause spinal imbalance; therefore, sagittal and coronal spinal imbalance should be recorded. Spinal balance in sagittal and coronal plane, and pelvic balance, head tilt, and shoulder balance must be keenly recorded. The rigidity of the curve is assessed.

Rib cage deformities can be seen with vertebral malformation; therefore, any anomaly of rib cage should be recorded. Inspiratory and expiratory capacity of lungs should be evaluated by pulmonary function tests to detect any restrictive lung disease.

A detailed neurological examination including muscle forces, sensation of the skin, abdominal and deep tendon reflexes should be recorded to rule out any spinal dysraphism. Patient's back should be examined carefully for any hair patches, lipomata, dimples, and abnormal pigmentation, which can be the first signs for intraspinal pathology. Physical findings such as asymmetrical calves, cavus feet, clubfeet, and vertical talus can also be the manifestations of spinal dysraphism, so a detailed lower extremity examination is mandatory.

11.5.2 Imaging

Appropriate imaging techniques should be used during patient evaluation in order to define the pathologic anatomy, classify the malformation, and make a logical surgery plan.

Routine radiographs are essential to evaluate the deformity. Radiographs in infants can be taken supine. As the child becomes older and can stand independently, standing posterior–anterior (PA) and lateral views should be obtained [29]. Measurement of Cobb angle is often more difficult in patients with CS, because of the distorted end plates and the malformed pedicles. However, with high-quality radiographs, it is possible to determine the type of the malformation, the magnitude of the curve, and the growth potential of the vertebral anomaly. It is also a reliable method for follow-up of curve progression [18].

In a study investigating intra- and interobserver variability in measurement of the Cobb angle in CS, Loder et al. [37] found a variability of ±9.6° and ±11.8°, respectively. According to the authors, to ensure with 95 % confidence that the increase in the curve is not due to error of measurement, at least 23° of change is necessary. Facanha-Filho et al. [18] stated that the variability in Loder et al. study was very high and

performed another study to assess variability in measurement of the Cobb angle. They found a mean intraobserver variance with an average of 2.8°, and the interobserver variance was 3.35°.

It is possible to estimate the growth potential of malformed vertebrae from disc spaces and their relative sizes on direct radiographs. If they are narrow and poorly defined, they do not have much growth potential. On the other hand, visible, wide, normal appearing discs associate a high potential for growth and curve progression. Even though the conventional method of evaluating CS is direct roentgenograms, these images can be difficult to interpret in patients with small size, in overlying structures obscuring the deformity, and in complex deformities. In patients who are candidates for surgery, more detailed imaging techniques are necessary.

New improvements in computed tomography (CT) and magnetic resonance imaging (MRI) technology made both of these modalities indispensable, especially in patients undergoing spinal stabilization and with complex deformities.

Three-dimensional computed tomography (3D-CT) is the best modality for defining the osseous anomalies and their relationships [46]. It is mostly recommended for complex deformities but not for routine observation or serial follow-up. Hedequist et al. [28] compared the findings in direct radiogram and 3D-CT of patients with CS with the findings in surgery. In all patients, anterior and posterior anatomy correlated with the CT findings. It is clear that a three-dimensional analysis of abnormal vertebrae can demonstrate the relationship between the anterior and posterior structures. These findings are absolutely necessary to evaluate even a single hemivertebra, whether it is a discordant type or not, to ensure that the patient undergoes the appropriate level of surgery [35] (Fig. 11.3). On the other hand, it should be kept in mind that serial X-ray and frequent CT evaluations loads significant amount of radiation to those patients with CS and every effort should be spent to decrease the unnecessary radiographic evaluations.

EOS (EOS Imaging, Paris, France) is a novel low-dose biplanar digital radiographic imaging system that can scan the patient in a standing position in two orthogonal planes. Although it has been shown that the EOS system can be reliably used in adolescent idiopathic scoliosis and limb length measurements, its reliability has been questioned in CS where each individual patient shows different curve and malformation patterns [17, 30]. The main advantage of EOS is that it decreases radiation exposure by up to 85 %.

MRI is the standard diagnostic tool for the assessment of intraspinal pathology [11]. Specific indications for MRI include the presence of neurological signs such as weakness, sensory loss, bowel or bladder dysfunction, a skin abnormality over the spine like dimple, hairy patch or nevus, leg or back pain, lumbosacral kyphosis, interpedicular widening. MRI is also very important in any patient undergoing spinal correction and stabilization. In addition, MRI may become a key modality for the assessment of natural history of congenital vertebral anomalies, particularly hemivertebrae because it also demonstrates various types of congenital disc and tissue anomalies [44] (Fig. 11.4).

Since the genitourinary system abnormalities are found in 18–40 % of patients with CS, screening renal ultrasonography is recommended to all patients [15, 39]. The incidence of congenital heart disease in patients with CVM was found to be 26 % in a recent study of Basu et al. [8] revealing the high importance of detailed cardiac examination and an echocardiography for a thorough examination.

11.5.3 Associated Anomalies

The development of vertebral column is closely associated with the development of the spinal cord; therefore, the neural and vertebral malformations often coexist. These malformations may cause neurological findings. However, absence of any neurological finding does not rule out intraspinal pathology [29]. Mesoderm, which is responsible for the formation of vertebra, is also responsible for the formation of urogenital, pulmonary, and cardiac systems. Malformation of these systems can also accompany congenital vertebral malformations [33]. Therefore,

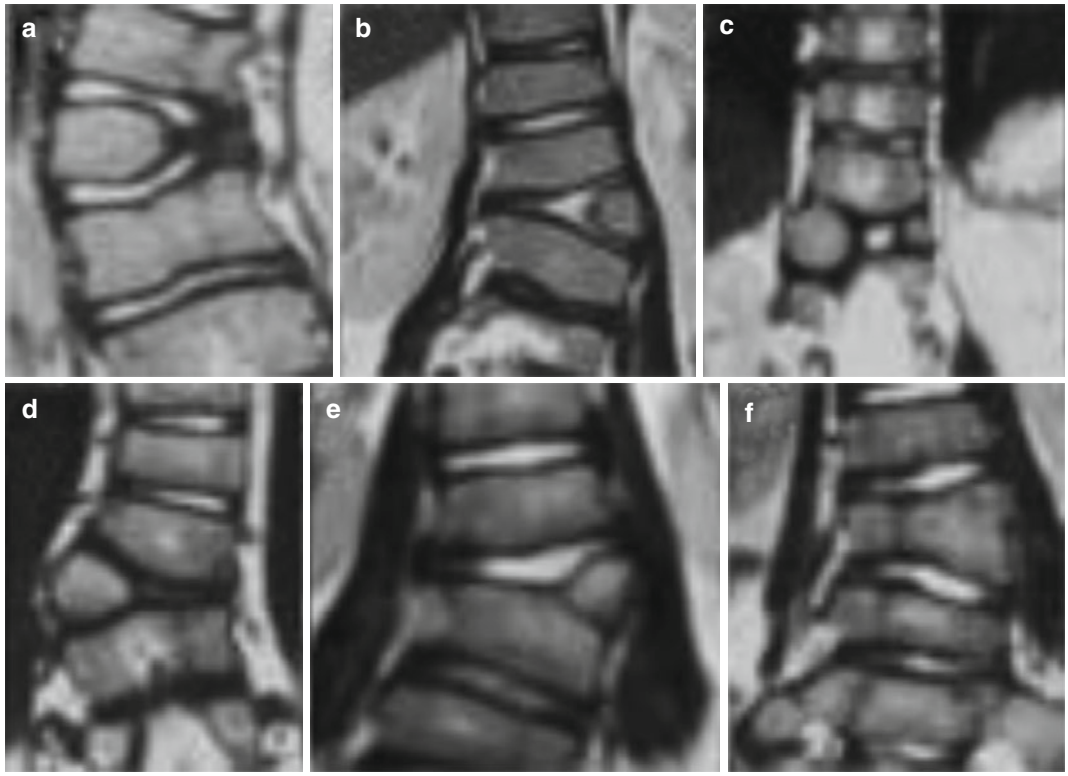


Fig. 11.4 (**a**–**f**) Magnetic resonance imaging (MRI) of different types of hemivertebras and the discs around malformed segments

systemic evaluation of the patients with proper imaging techniques is mandatory.

In a study using direct radiographs and myelograms, McMaster et al. [42] found intraspinal pathology in 18.3 % of 251 CS patients. When MRI was used as diagnostic tool for evaluation of the CS, neural axis abnormalities increased to 30–38 % [11, 48]. Bollini et al. studied the relationship of the level of the hemivertebra (HV) and the incidence of the intraspinal pathology. They showed that the incidence of intraspinal pathology is higher in patients with HVs located at lumbosacral region than any other part of the spine (33 % for lumbosacral, 13 % for lumbar, and 10 % for thoracic HV). They were unable to show any relationship between the intraspinal or other visceral pathologies and the type (segmented-semisegmented), number (single-multiple), side (right-left) of the HV, or gender of the patient [10].

The most common malformation of spinal cord is diastematomyelia (split cord), which is defined as partial or complete split of spinal cord or cauda equina with a bony or fibrous spur [42]. Diastematomyelia is found in approximately 20 % of patients with congenital scoliosis [27]. In patients with diastematomyelia, the normal movement of spinal cord is restricted. The spinal cord stretches while the spinal column grows longitudinally. Any corrective manipulation of spine may cause more stretching of the cord, and this may result in neurological deterioration. Therefore, it is very important to evaluate the whole spine before any corrective surgery. Other congenital intraspinal anomalies associated with CVM are epidermoid cysts, dermoid cysts, neuroenteric cysts, tethered spinal cord, lipomas, and teratomas [42] (see Chap. 15).

Evidence from a number of retrospective studies shows diminished pulmonary function in patients with CS. Because of the complex interconnections between spine, sternum, and ribs, the displacement and rotation of the vertebrae in

scoliosis have profound effects on the shape of the thorax. In a retrospective study, 192 (50.3 %) of 382 patients were reported to have rib anomalies. Missing rib was the most common anomaly among others. The anomalies were located more frequently on the concave side and mostly associated with thoracic and thoracolumbar CVMs [63].

Individuals with CS and chest wall deformities are believed to have a thoracic deformity that limits lung growth and rib deformities leading to thoracic instability and alteration in respiratory mechanics. The expansion of the thoracic cavity is limited as the movement of the ribs is impeded, which in turn decreases chest wall compliance and makes breathing significantly harder despite the absence of any lung disease. Altered development and morphology in patients with scoliosis can lead to measurable changes in lung function most consistent with restrictive lung defect.

Renal system abnormalities may be found in 18–40 % of patients with CVM. Anomalies may affect the kidneys, ureters, bladder, and urethra. Unilateral renal agenesis, duplicated kidneys, and ureteral obstruction are the most common renal abnormalities associated with congenital scoliosis [15, 39].

Congenital heart disease is present in 10–26 % of patients. Atrial and ventricular septal defects are the most common cardiac abnormalities. More complex cardiac malformations like tetralogy of Fallot and transposition of great vessels can also be seen in CS patients [8, 49].

Musculoskeletal anomalies like clubfeet, Sprengel's deformity, Klippel-Feil syndrome, developmental dysplasia of the hip may all be seen in these patients [29].

11.6 Treatment Alternatives

Once the CS is diagnosed, it is very important to note the patient's age and spinal balance and to classify the anomaly. Treatment should be started regardless of age, if the patient has CVM with high potential to deteriorate such as unilateral hemivertebra and contralateral unsegmented bar. Patients with anomalies less inclined to deteriorate should be carefully followed with serial radiographs, and Cobb angle measurement of their curves should be obtained at each visit to detect any progression.

There are many treatment alternatives for CS. The age of the patient, type of anomaly, limitations of the surgeon, and surgery room should be considered to choose the most appropriate way of treatment.

11.6.1 Observation

Patients with balanced spine and vertebral malformations, less prone to deteriorate like hemimetameric shift or block vertebra, can be followed with serial plain radiographs at 4- to 6-month intervals. It is important to assess the spinal balance of the patient and the Cobb angle measurements of the curves. Most recent radiographs should be compared with the earliest radiographs of the patient in order to detect any progression.

11.6.2 Bracing and Casting

Short and rigid curves rarely respond to brace treatment. Bracing can be considered for long, flexible curves and for compensatory curves, which are located proximal or distal to anomalous segment. Serial casting, which is an effective treatment modality in early-onset idiopathic scoliosis, can also be used in very young patients with CS until surgical procedures could be applied. Demirkiran et al. recently showed that the serial casting can effectively control both the main and the compensatory curves while providing longitudinal spine growth in 11 patients with CS [14].

11.6.3 Surgery

Surgical techniques applied in CS will be discussed in detail in consecutive chapters, so only brief summaries regarding the surgical alternatives will be mentioned here. The reader is advised to refer to specific chapters for technical details (Chaps. 30, 31, 32, 33, 39, and 40).

11.6.4 Growth Inhibition

In situ spine fusion via posterior exposure is historically the oldest surgical technique used in the treatment of CS. Because the in situ fusion technique provides no correction and the pseudarthrosis rate is high, the treatment results are unsatisfactory [6].

Even though the addition of anterior fusion to posterior fusion degreased the pseudarthrosis and

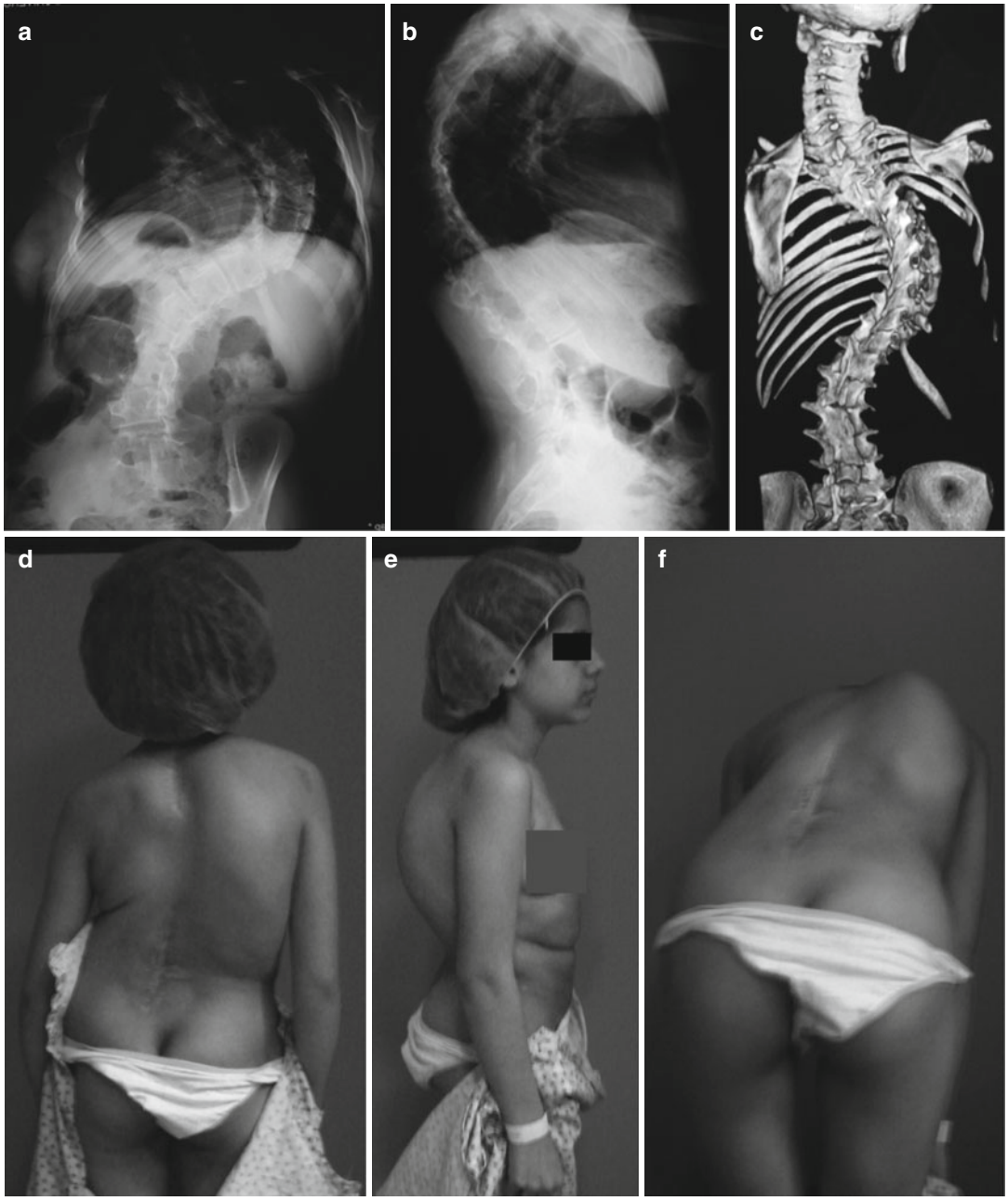


Fig. 11.5 A 12-year old girl with congenital spinal deformity (**a**–**f**). During the posterior instrumentation and fusion, additional multiple Chevron osteotomies and concave rib osteotomies were performed (**g**–**k**)

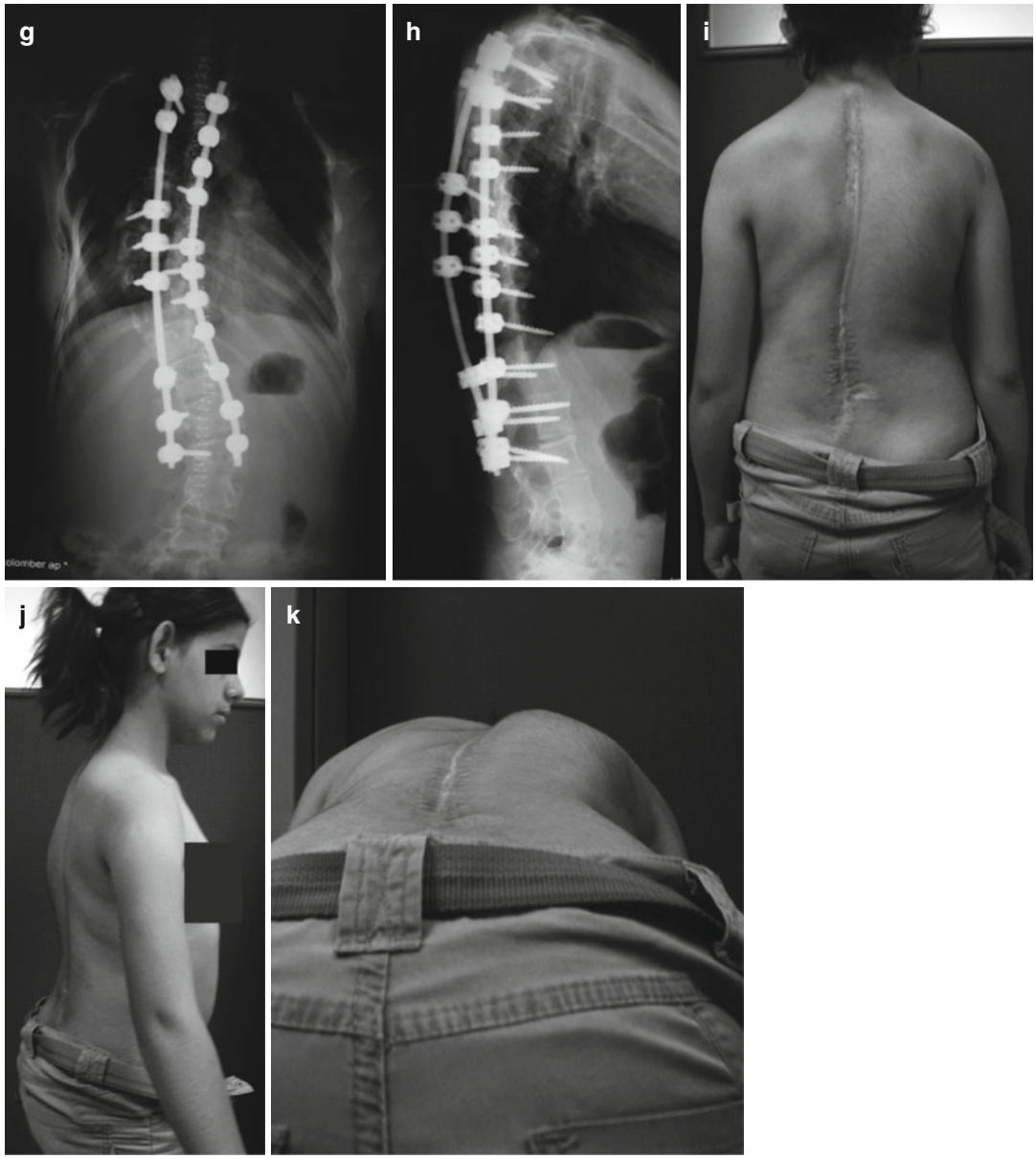


Fig. 11.5 (continued)

crankshaft rate in the following years, the reality that the pre-existing deformity is unchanged makes the in situ fusion technique undesirable for patients with severe deformity and trunkal off-balance. With the help of recently developed reconstructive techniques, in situ fusion is no longer a preferred choice of treatment in CS.

CS has long been accepted as a spine deformity where the instrumentation should be avoided due to complexity in deformity and anatomy, rigidity, and accompanying intraspinal pathologies. Today, with the help of improved visualization techniques, better equipped anesthesia and intensive care units, neuromonitorization, and with improvements in implant technologies, instrumentation of CS is safer than before and obtaining significant correction is possible [6, 7]. Figures (11.5 and 11.6) instrumentation of long

segments in the early period of life is accompanied by spinal fusion, which is undesirable at early ages, and different surgical alternatives have been sought.

11.6.5 Growth Modulation

Growth inhibition on the convex side of the deformity by means of in situ fusion (convex growth arrest) without instrumentation, or by pedicle screws or staples, is another option, especially for deformities with normal growth potential on concave side [13]. Patients 5 years of age or less with scoliosis of 70° or less and without any lordosis or kyphosis are ideal candidates for growth modulating surgical treatment [61]. Convex growth arrest appears to be an effective procedure to halt the progression of the curve with an expected correction over time in scoliosis patients without signs of advanced skeletal maturity. The overall main problem seems to be the unpredictability of the results [57]. Alanay et al. added a concave rod for distraction in addition to instrumented convex fusion and proposed a new modification of convex growth arrest. They showed better correction in scoliosis and balance [3].

11.6.6 Growth Preservation/ Stimulation

11.6.6.1 Growing Rod

The growing rod technique, which was described for idiopathic or idiopathic-like deformities where the vertebral anatomy is normal, is based on reconstructing the deformity via a distraction maneuver (Fig. 11.7). However, detailed examination of the growing spine series, published in the past few years, revealed that the method has also been applied in patients with CS [2, 55]. A recent multicentric study, where 19 congenital scoliosis patients were treated with growing rod technique and were followed for at least 2 years, reports 31 % correction in Cobb angle and 12 mm yearly elongation of #T1-S1 segment [16]. Also, space available for lungs ratio increased from 0.81 preoperative to 0.94 postoperative. None of the patients in the study group had neurologic injury. In another study of 30 patients with CS in which the dual growing rod was applied, 1.49 cm of longitudinal spinal growth was obtained per year. There was significant correction in main curve [58]. The growing rod technique is a safe and reliable method for young children, who present some flexibility in the anomalous segment, or when the congenital anomaly involves a vertebral segment too long for resection or with compensating curve with structural pattern concomitant to the congenital deformity.

11.6.6.2 Expansion Thoracoplasty

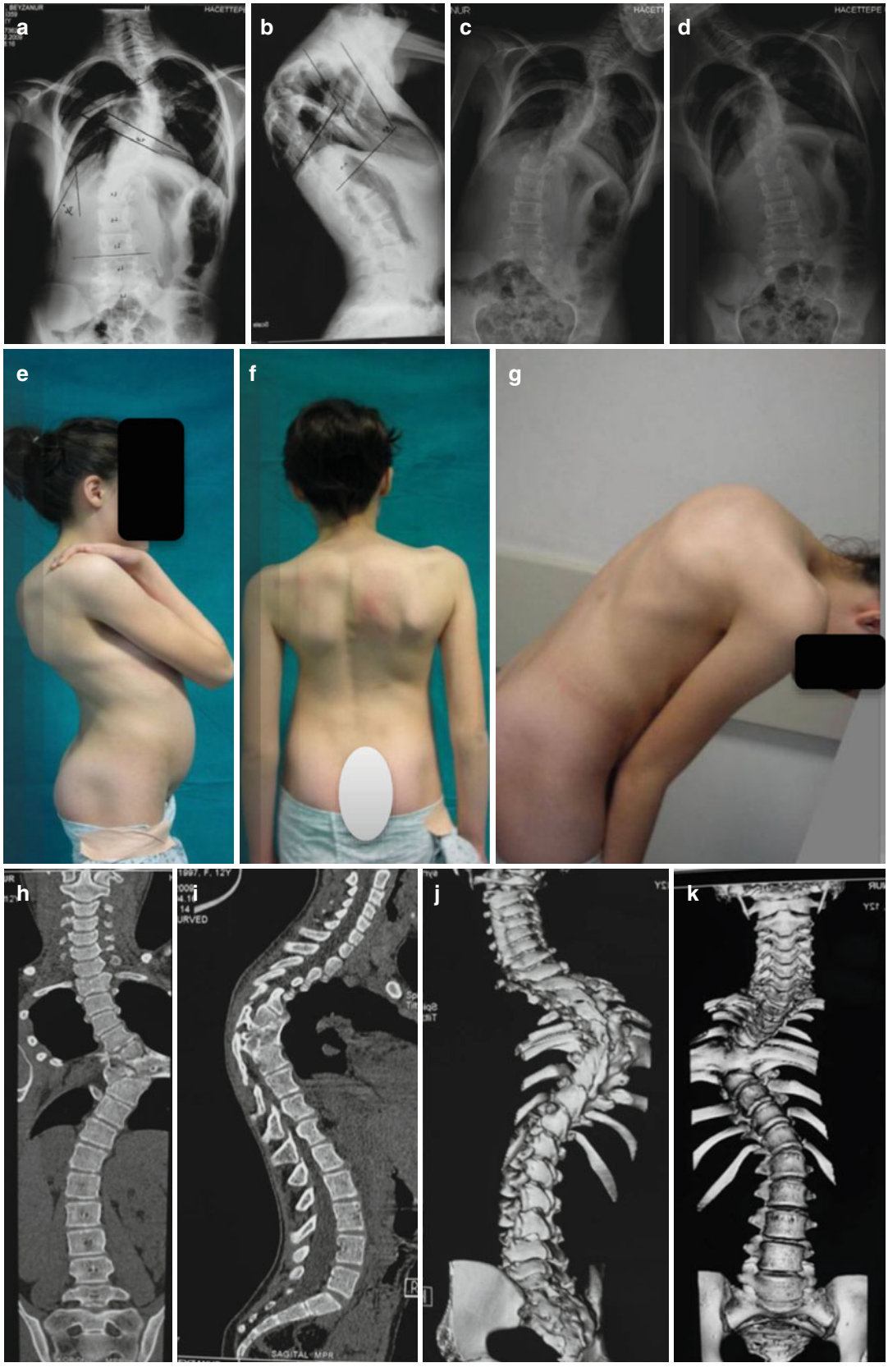
Growing rod technique is a spinal instrumentation method addressing the deformity in the spine and therefore should be used in patients where the primary problem is at the vertebral column. If the patient has rib fusions and/or thoracic insufficiency syndrome, in other words, if the primary problem involves the thoracic cage, it would be a preferable approach to employ the treatment method addressing to the thorax deformity, which is the thoracic expansion [12, 20].

11.6.7 Reconstruction

11.6.7.1 Hemivertebrectomy

Since the scoliosis due to hemivertebrae present bone excess on the convex side (or shortfall in concave side), the most reasonable and ideal treatment method is the resection of this bone (hemivertebrae). Although hemivertebrectomy has been technically defined as a procedure long time ago, it has not been applied frequently at the beginning, since it involved a long and challenging surgery on young children who face other comorbidities.

Fig. 11.6 A 14-year-old female patient with kyphoscoliosis. The patient has mixed-type congenital scoliosis on the upper thoracic region accompanied with rib fusions. Bending X-rays show that the curve is rigid. She underwent multiple chevron osteotomies and concave rib osteotomies through all-posterior approach, and the alignment in coronal and sagittal plane was constituted (**a**–**g**). Preoperative sagittal and coronal computed tomography (CT) reconstructions (**h**–**l**). Immediate postoperative clinical pictures and spine X-rays (**m**–**t**). Four-years follow-up spine X-rays (**u**, **x**)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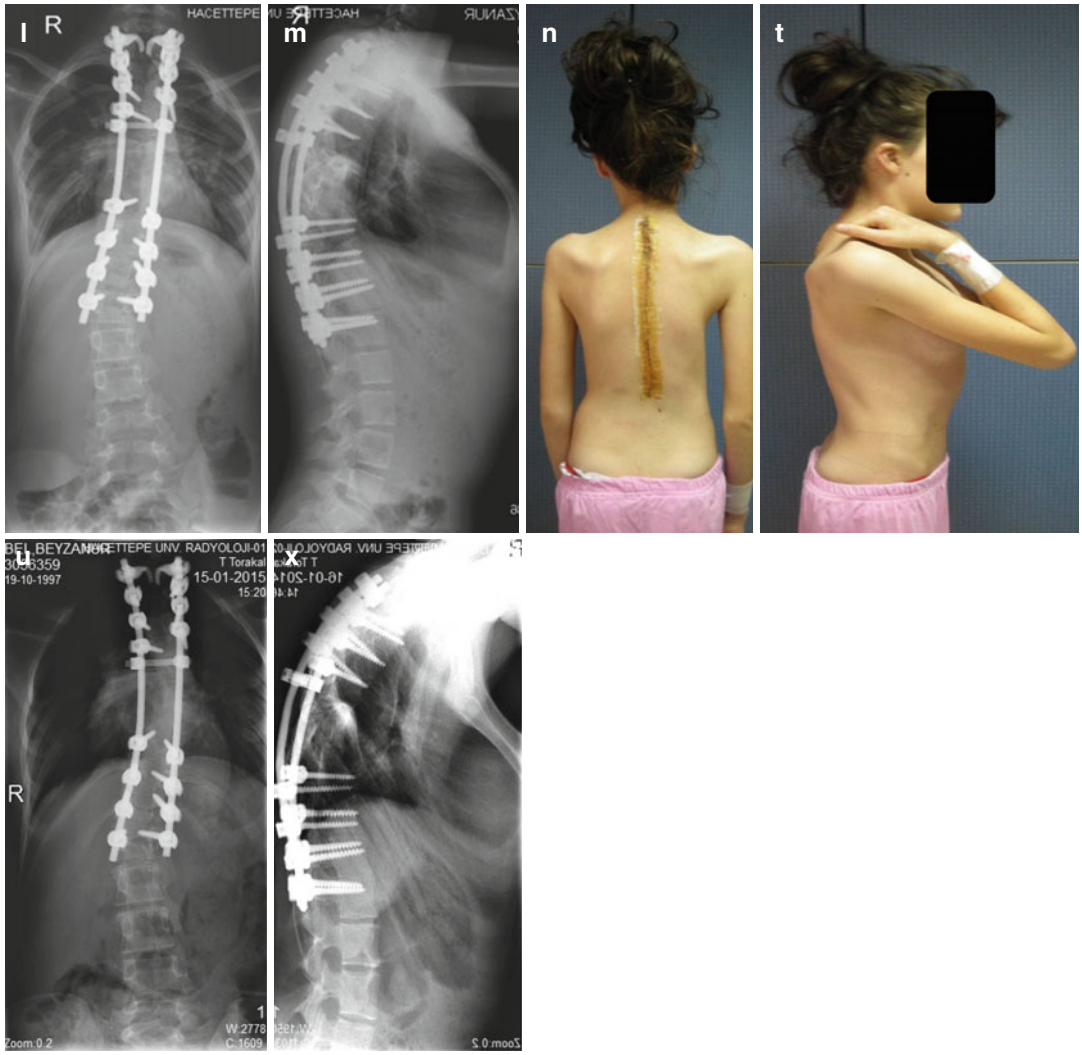


Fig. 11.6 (continued)

Because of the developments in anesthesia and postoperative care, today, surgery has become the standard treatment method for the single hemivertebrae of the thoracolumbar and lumbar regions [9] (Figs. 11.8 and 11.9). Hemivertebrectomy can be applied through a posterior approach or combined anterior and posterior technique can be utilized, with each technique having its own advantages and disadvantages [43].

11.6.7.2 Vertebral Column Resection

CS may emerge due to CVM much more complicated than a simple hemivertebrae. Simple hemivertebrectomy would definitely not suffice for the treatment of multiplanar complex deformities occurring in case of multiple hemivertebrae and unsegmented bars. Vertebral column resection is a technically challenging procedure [53, 54]. It takes a long time and involves excessive bleeding. Moreover, it is open to severe complications, including serious neurological injury [47, 54]. Despite all these risks, it is being widely practiced in experienced centers and turning into a standard procedure for the complex spinal deformities leading to serious trunk imbalance (Figs. 11.10 and 11.11).

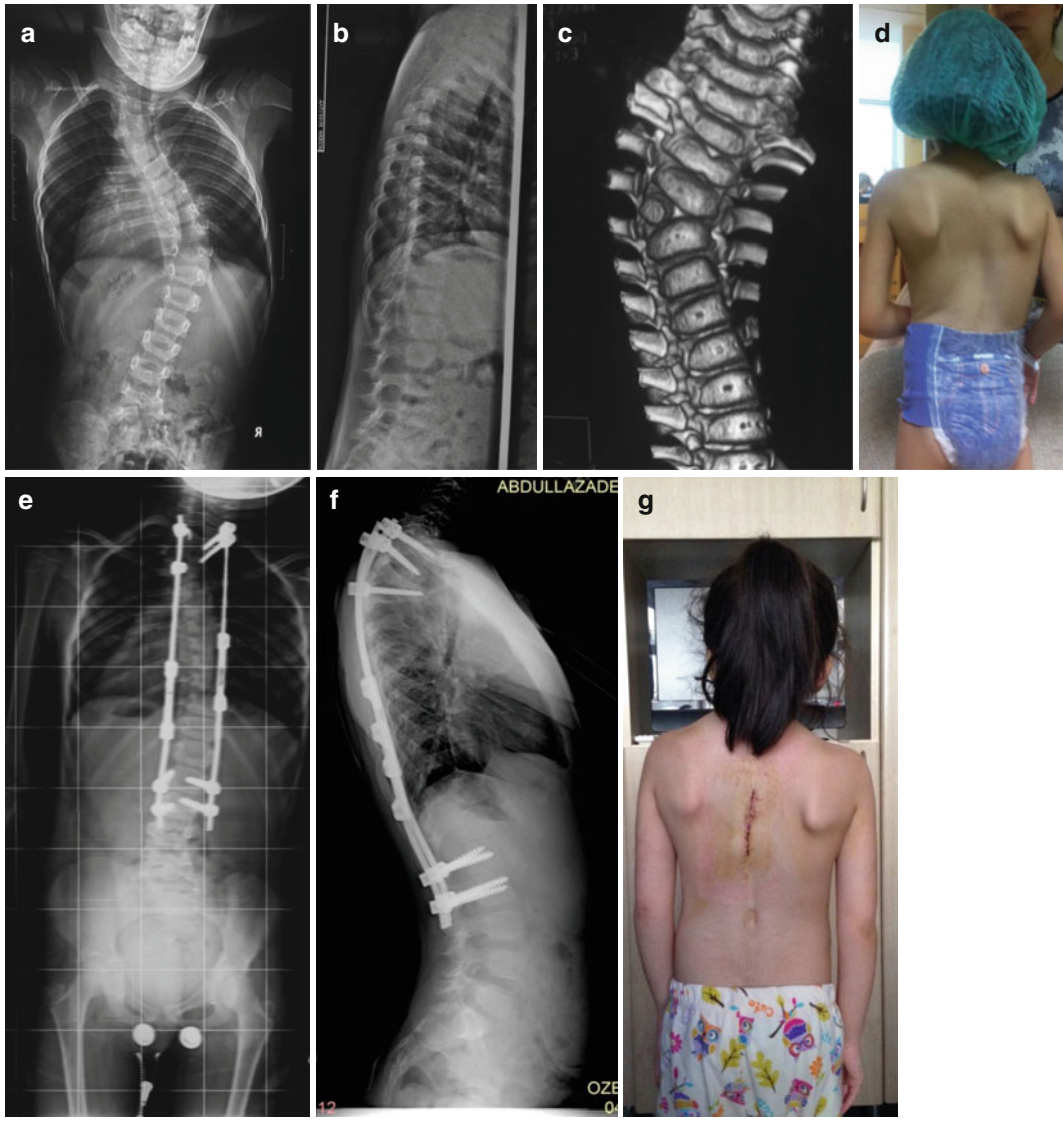


Fig. 11.7 A 4-year-old female patient with upper thoracic congenital formation anomaly accompanied with compensatory thoracolumbar curve (**a**–**d**). She underwent hemivertebra resection in addition to growing rod instrumentation. Clinical picture and radiologic result after fourth lengthening procedure (**e**–**g**)

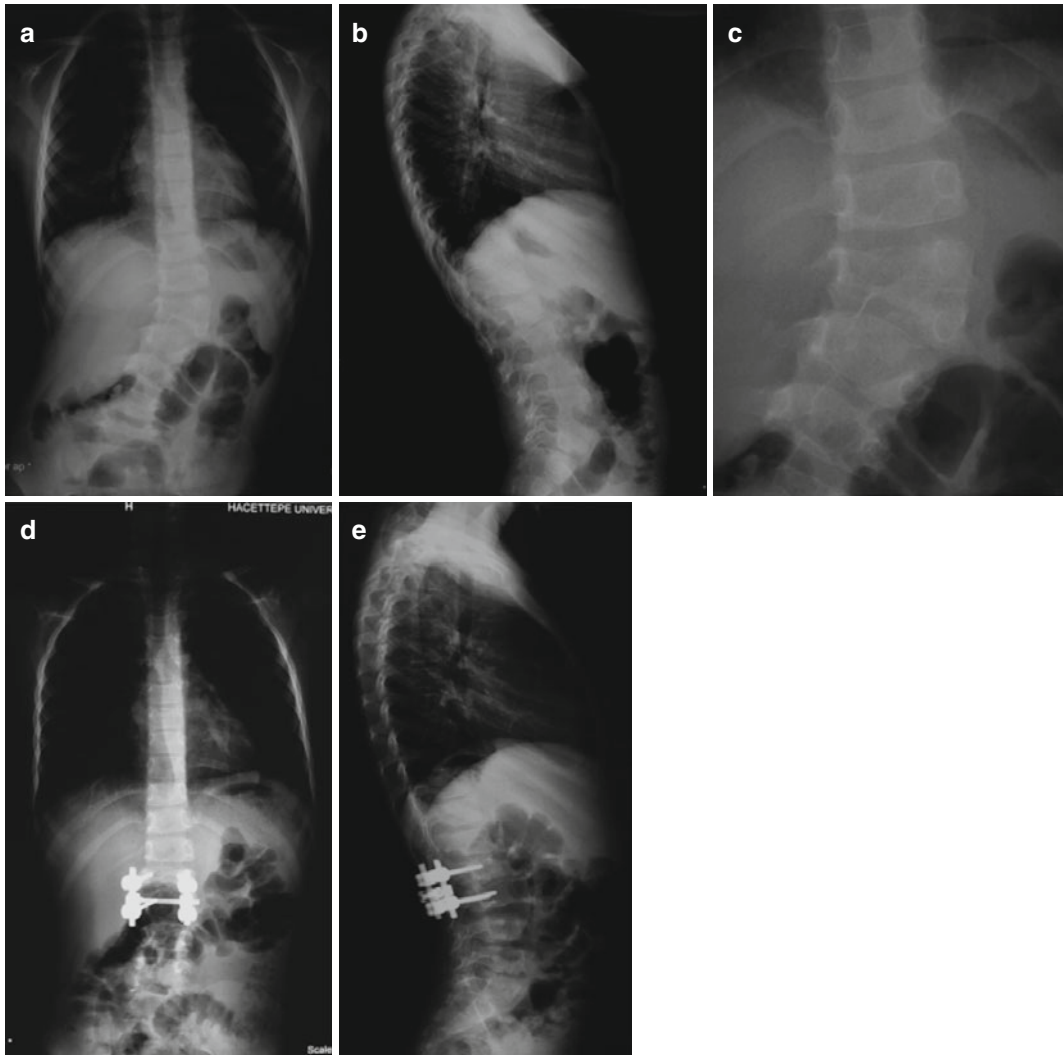


Fig. 11.8 A 4-year-old girl (**a**–**c**). Single hemivertebra at L1 was treated with hemivertebrectomy and posterior instrumented fusion (**d**, **e**). Since we were able to obtain perfect anterior bone contact after hemivertebrectomy, no additional anterior structural support was used

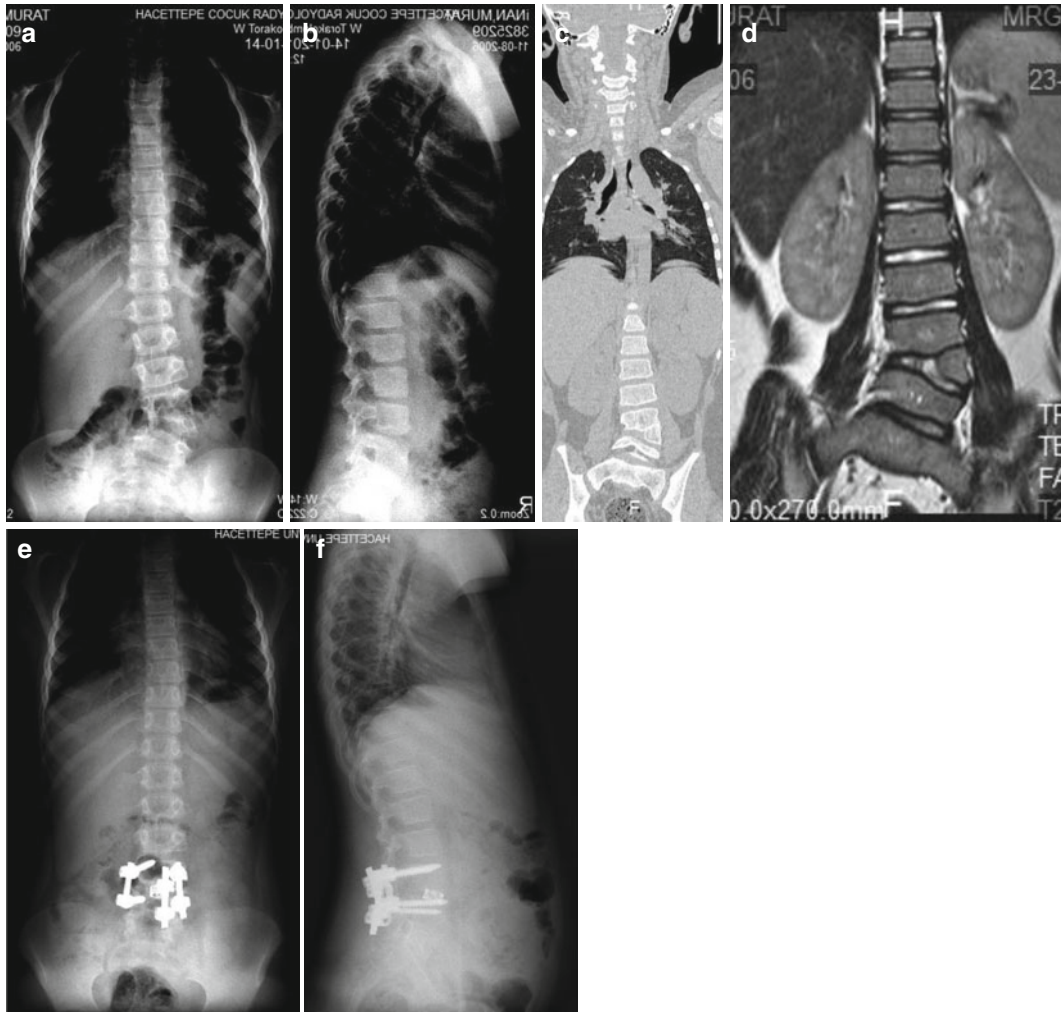


Fig. 11.9 A fully segmented unincancerated hemivertebra causing significant off-balance and severe waist asymmetry in a 14-year-old male patient (**a**–**d**). Reconstruction of the sagittal and coronal plane alignment using posterior three rods and anterior cage through an all-posterior approach (**e**, **f**)

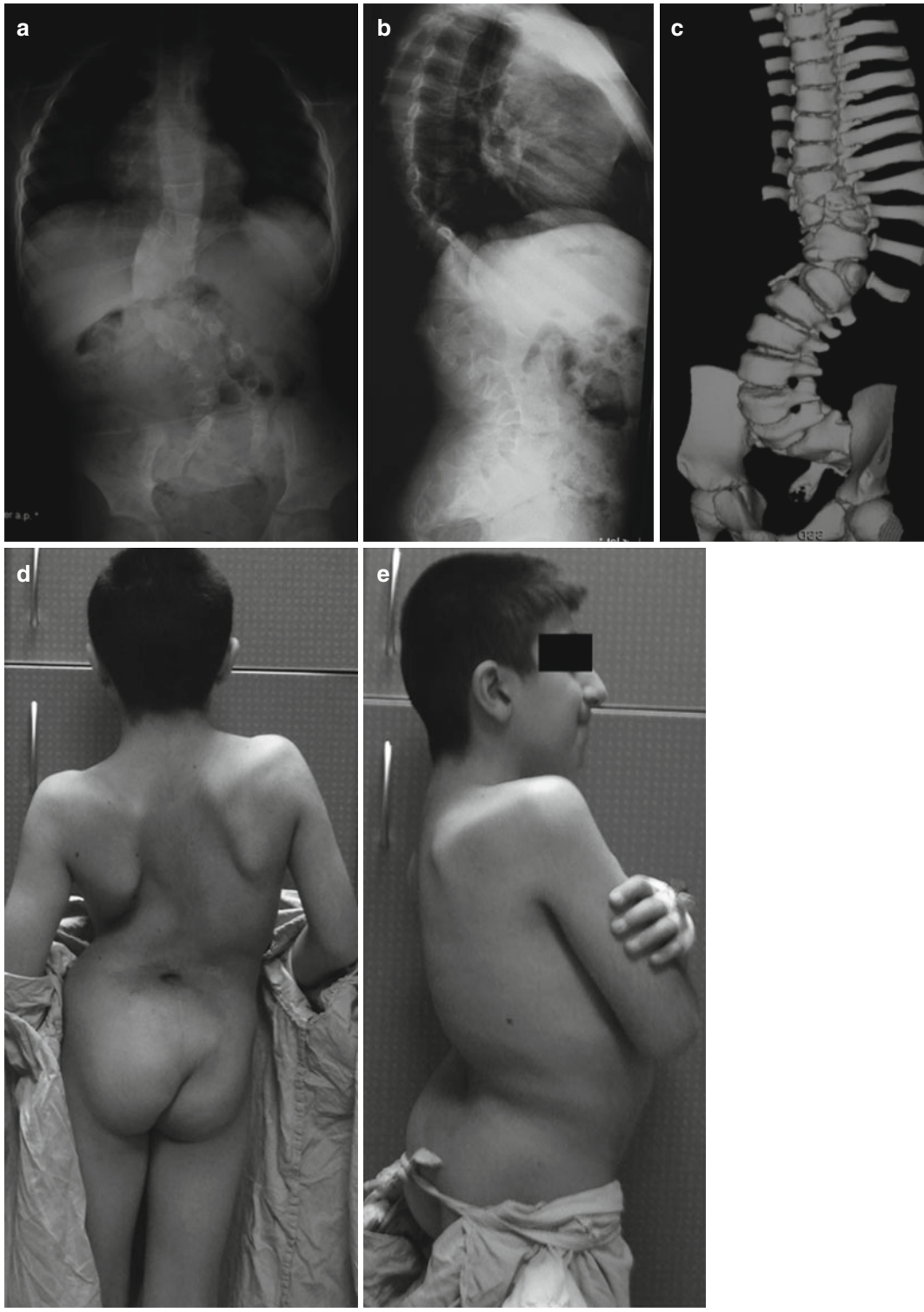
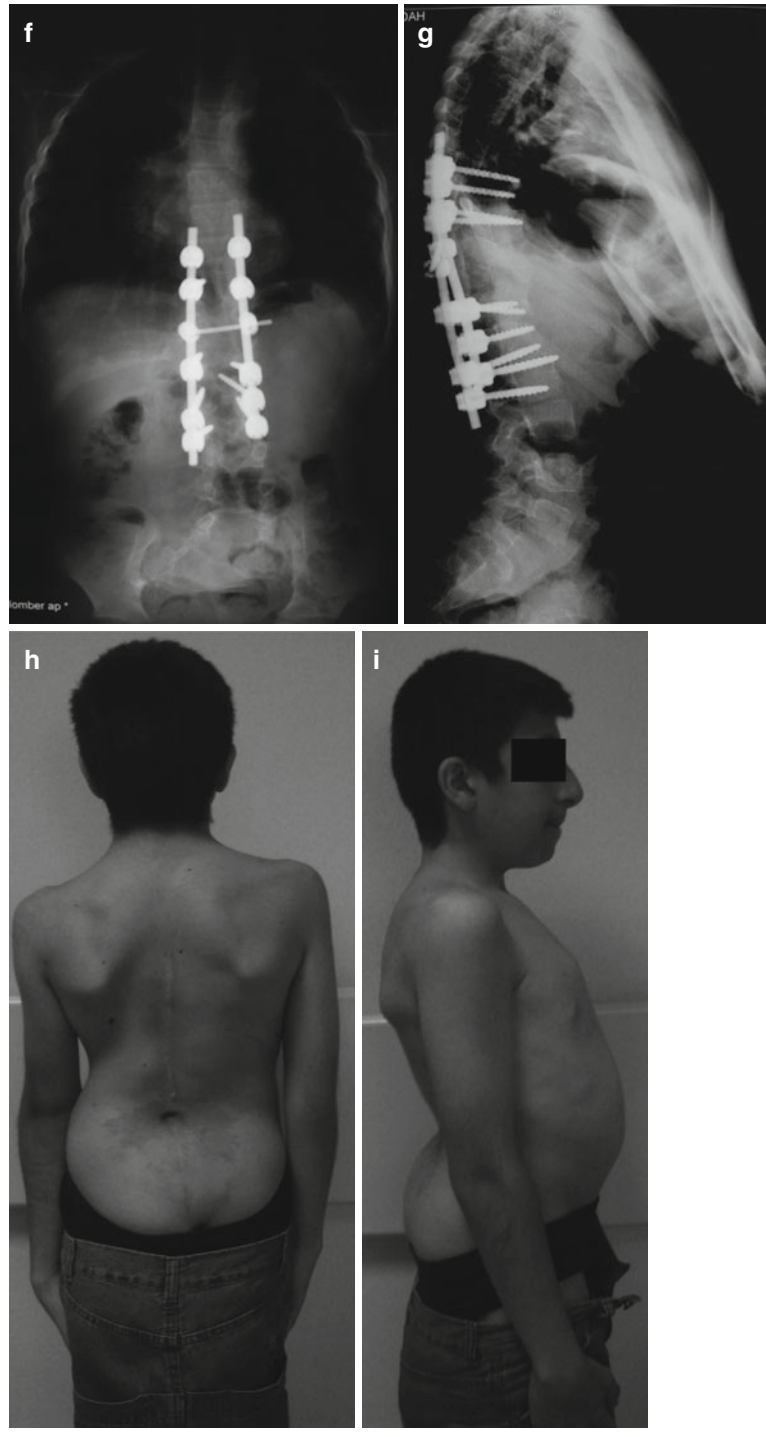


Fig. 11.10 An 11-year-old boy with congenital scoliosis (**a**–**e**). He underwent a neurosurgical intervention previously for diastematomyelia. After vertebral column resection, his deformity was improved both clinically and radiologically (**f**–**i**)



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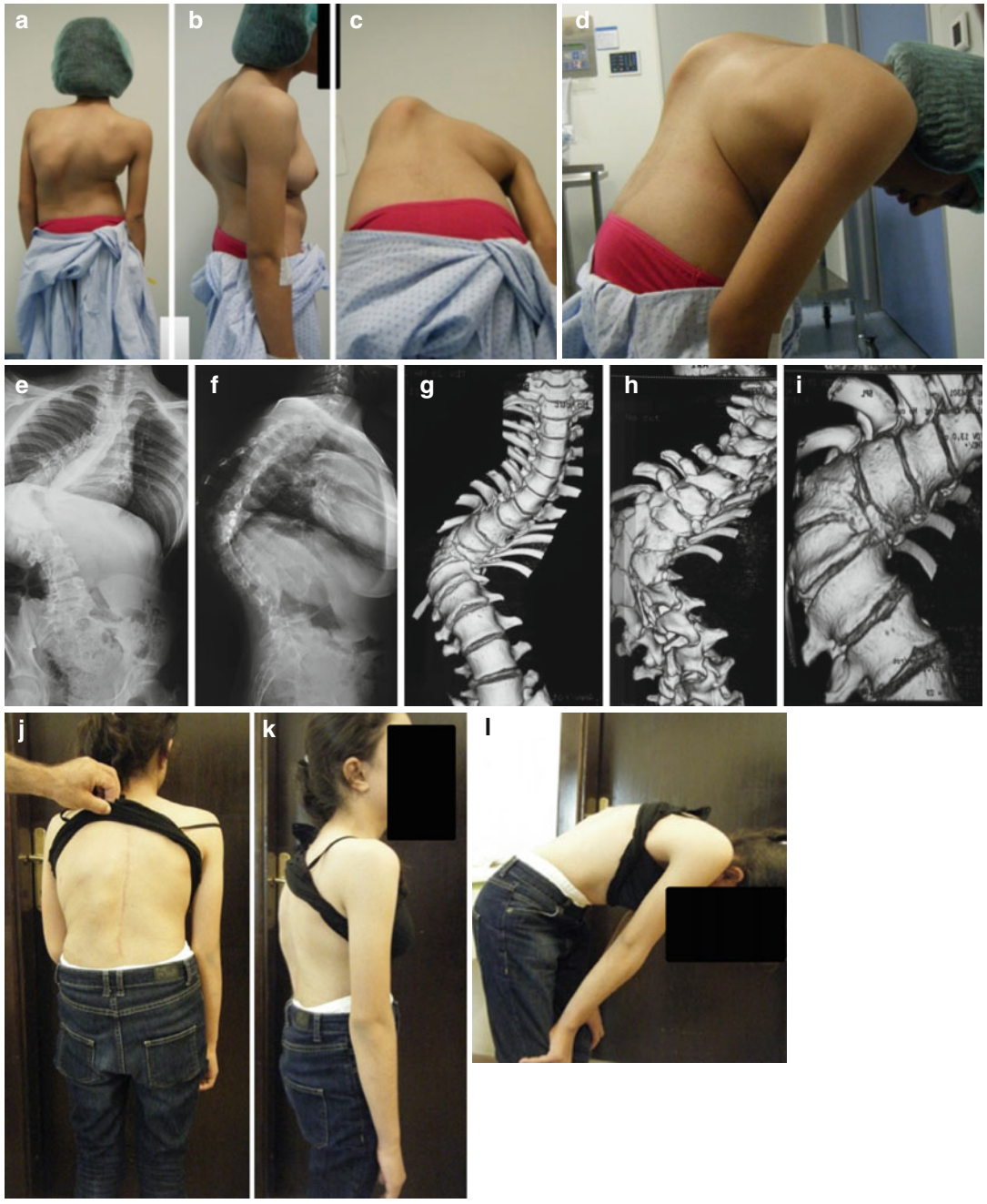

Fig. 11.11 A 15-year-old female patient has congenital spine anomaly resulting in sagittal and coronal plane deformities (**a**–**i**). Normal sagittal and coronal balance were created after vertebral column resection and posterior instrumentation (**k**–**o**)

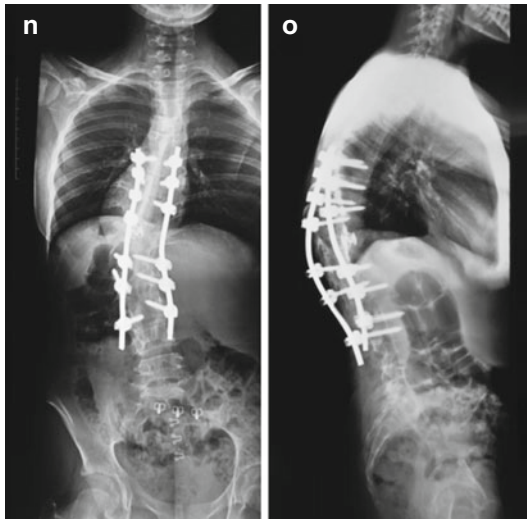

Fig. 11.11 (continued)

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Treatment of Spinal Deformity in Cerebral Palsy

12

Suken A. Shah and Leok-Lim Lau

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Key Points

- Scoliosis in cerebral palsy is related to the severity of neurological involvement; dependent sitters with poor head control have a very high rate of scoliosis. Curve progression leads to subsequent deformity and trunk imbalance with associated loss of function.
- The goals of surgery for the higher functioning patient are to provide more normal spinal balance, alter the progression of disease, and preserve function with respect to ambulatory potential; in a wheelchair-bound patient, the aim is to maintain independence in sitting and facilitate care. Improved sitting may correlate with attentiveness in class, easing of care, improvement of self-image, and decrease in the rate of decubitus ulcers.

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- Although it is generally accepted that bracing likely will not alter the progression of the curve in the cerebral palsy patient with scoliosis, it is reasonable to utilize an orthosis to improve muscle balance and sitting in patients with flexible curves while closely following them.
- In general, surgical intervention is considered for curve magnitudes greater than 40° or 50° and in patients with significant deterioration in function. Delay of intervention is possible when the curve is flexible and still can be performed with all posterior surgical approach with curve magnitudes up to 90°.
- Anterior fusion for the so-called crankshaft phenomenon is not necessary, even for young patients, when rigid, segmental instrumentation, such as a unit rod or pedicle-based system, is used posteriorly.
- A pedicle screw-based hybrid construct is the authors' preferred method of instrumentation and offers a powerful mechanism of correction in both the coronal and sagittal planes with primarily cantilever and translational correction mechanics. Iliac screws or S2 alar-iliac screws provide rigid fixation to the pelvis especially in patients with lumbar hyperlordosis. The construct allows powerful correction of deformities in multiple planes with less implant-related complications.
- The risk of complications, both perioperatively and postoperatively, is substantial but manageable with a careful preoperative workup, multidisciplinary care, and attention to details.
- Caregiver satisfaction is high after this procedure and affords a good long-term outcome with a positive impact on the patient's sitting ability, physical appearance, comfort, and ease of care.

12.1 Introduction

A spinal deformity arising in the clinical setting of muscle imbalance secondary to an underlying neuropathic or myopathic disease can be classified as neuromuscular spinal deformity. The associated muscle imbalance in neuromuscular disease causes abnormal biomechanical loading of the spine. According to the Heuter–Volkmann principle, abnormal biomechanical loading secondary to this muscle imbalance and spinal collapse results in asymmetric vertebral body growth (and hence anatomic deformity) in a skeletally immature individual. Progressive deformity is believed to be the result of both progressive muscle imbalance and anatomic deformity.

Of the neuropathic and myopathic disorders associated with scoliosis (Table 12.1), cerebral

Table 12.1 Neuromuscular disorders associated with scoliosis and their classification

Neuropathic
Upper motor neuron
Cerebral palsy
Spinocerebellar degeneration
Friedreich ataxia
Charcot–Marie–Tooth
Roussy–Levy
Syringomyelia
Spinal cord tumor
Spinal cord trauma
Lower motor neuron
Poliomyelitis and other viral myelitides
Traumatic
Spinal muscle atrophy
Werdnig–Hoffmann
Kugelberg–Welander
Dysautonomia
Myopathic
Arthrogryposis
Muscular dystrophy
Duchenne
Limb-girdle
Facioscapulohumeral
Fiber-type disproportion
Congenital hypotonia
Myotoniadystrophica

palsy is the most prevalent. Scoliosis represents the majority (93 %) of the patients with spinal deformity. Isolated sagittal deformity including thoracic hyperkyphosis or lumbar hyperlordosis constitutes the remaining cases [1]. This chapter will mainly focus on the operative treatment of scoliosis due to cerebral palsy.

Cerebral palsy (CP) has an estimated incidence of two per 1000 of live births [2], with an incidence of scoliosis 15–28 % on clinical or radiological examination in a general CP population [3]. In an institutionalized CP population, Madigan and Wallace [4] found a 64 % incidence of scoliosis. Of the physiological classification of CP, spastic quadriplegic CP has the highest incidence of scoliosis [3, 4]. The risk of scoliosis correlates with the level of ambulatory ability of the patients graded by Gross Motor Functional Classification System (GMFCS). Children with mild gross motor function limitation (GMFCS levels I-II) have no higher risk of developing scoliosis than the general population. In children with limited motor function (GMFCS levels IV and V), the risk of developing clinically moderate or severe scoliosis is 50 % [3].

Lonstein and Akbarnia [5] classified scoliotic curves as a result of CP into two groups: Group I curves, which are double curves with thoracic and lumbar components (S-curves) that behave like idiopathic scoliotic curves with higher likelihood of preservation of ambulation ability, and Group II curves with more lumbar or thoracolumbar curves that extended into the sacrum with associated pelvic obliquity (C-curves). The long, sweeping, and collapsing curves are more typical of neuromuscular curves in patients who are wheelchair-dependent or bed-ridden. The apex of these curves centered at the thoracic (T2–T10) or thoracolumbar (T11–L1) and pointed to the right (Fig. 12.1). Among institutionalized CP population, Group I and II curves have the same incidence [4]; however, Group II curves form the majority (94 %) [6] of the patients with CP who required surgical intervention attributable to pelvic obliquity, poor coronal balance, and large magnitude of the curve.

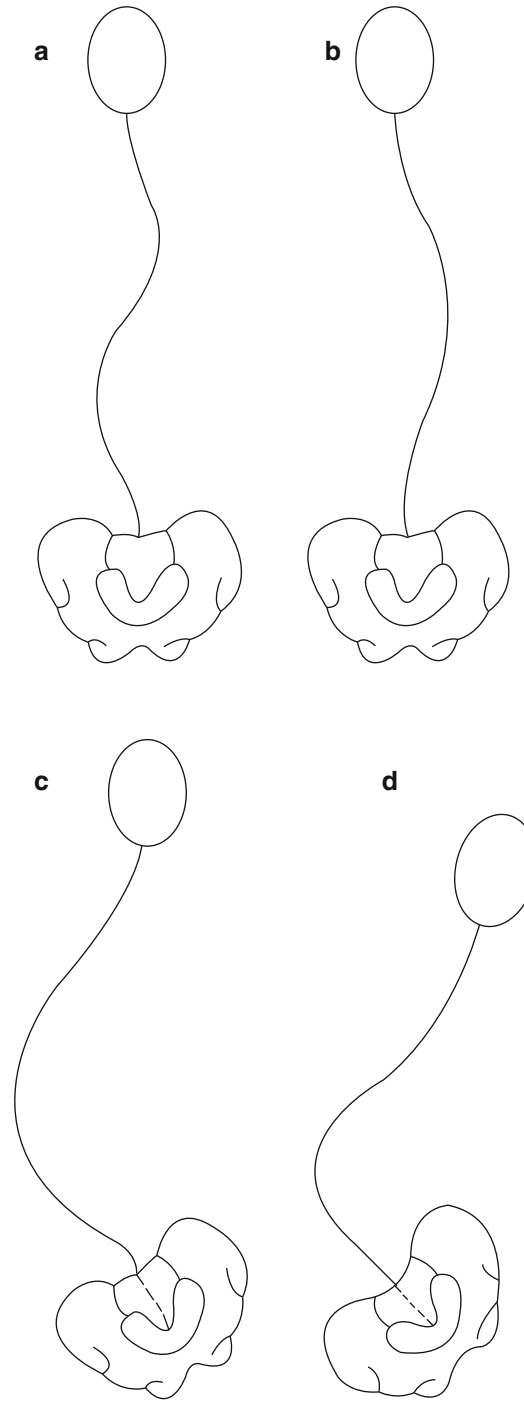

Fig. 12.1 Curve patterns in cerebral palsy scoliosis. Group I curves are double curves with little pelvic obliquity that may be balanced (**a**) or unbalanced (**b**). Group II curves (**c**, **d**) are large lumbar or thoracolumbar curves with marked pelvic obliquity (Adapted from Lonstein and Akbarnia [5], p 800)

12.2 Natural History

Although the age of onset can vary widely from 3 years old to 20 years old, neuromuscular scoliotic curves generally develop at a younger age than adolescent idiopathic scoliosis [7]. Some of the patients present with significant curves prior to the prepubescent growth spurt as a result of earlier curve onset. With the growth spurt, which is typical delayed, the flexible, postural curve tends to develop into a torsional structural deformity. Finally, a stiff curve of considerable magnitude develops before the growth complete.

The single most important factor that affects the magnitude of the curve is the severity of CP. There appears to be proportional relation between the severity of involvement of CP and the curve severity; 67 % of the quadriplegic and 18 % of the non-quadriplegic spastic CP in this study had curves that exceeded 60° [7].

While the rate of curve progression is highly variable, the average progression cited in one report is 0.8° a year in curves less than 50° and 1.4° a year in curves more than 50° [8]. During periods of rapid growth, much more severe progression can occur. Eighty-five percent of patients who had curve of more than 40° by age 15 years progressed to 60°, while only 13 % of those with a curve of less than 40° by age 15 progressed to 60° [7]. Some authors reported curve progression after intrathecal baclofen therapy was instituted to control severe spasticity even in skeletally mature patients [9]. In an adequately powered, case–control study, it was found that the rate of progression was not affected by the use of intrathecal baclofen therapy and that curve incidence and progression were rather related to neurologic involvement [10].

Curve progression increases the magnitude of deforming forces and leads to subsequent deformity, truncal imbalance, and pelvic decompensation. The pelvis is often the end vertebra – the most tilted vertebra with residual axial rotation of the C-curve. This was described as the pelvic vertebra by Dubousset. Less commonly, pelvic obliquity presents as a compensatory fractional curve to the C-curve. Pelvic obliquity alters the sitting position and the pressure at the typically well-distributed sitting tripod at both ischial tuberosities and pubic symphysis. The undue increased pressure at the ipsilateral ischial tuberosity is further exacerbated in patients who have increased pelvic tilt and results in pressure sores.

Depending on the dominant deforming forces and the interplay of spasticity, patients may present with kyphoscoliosis or lordoscoliosis. In kyphoscolosis, progressive deformity with associated pelvic obliquity and retroversion may compromise the often-limited ambulatory function particularly standing to transfer. Significant pelvic obliquity makes sitting adaptation difficult and sometimes impossible. In lordoscoliosis, patients may present with extensor posturing clinically. The progressive deformity renders sitting impossible. Patients may need to be nursed in a semi-reclined position in a wheelchair. This group of patients may present with acute pain that is not alleviated with any sitting adaptation.

Significant spinal deformity is known to compromise cardiopulmonary function, affect gastrointestinal motility, and result in rib-pelvis impingement. The morbidity associated with these are difficult to quantify in this vulnerable and low-demand population especially those with pre-existing difficulties in swallowing, dependence on G or J-tube, and multiple medical comorbidities and are unable to articulate their symptoms.

12.3 Clinical Presentation and Evaluation

Given the universal progressive nature of neuromuscular scoliosis, early diagnosis of the deformity is essential. Initial evaluation should consist of clinical monitoring by physical examination. During physical examination, the patient is examined in a sitting position for a curve and pelvic obliquity. When a curve is identified, the crux of the examination is to assess the flexibility of the curve clinically and the remaining growth potential by serial height and weight measurements and radiographic markers.

Curve flexibility is assessed by holding the patient up at the axillary areas in a sitting position.

In a smaller framed patient, a clinical fulcrum bend test over the examiner's knee is possible. Pelvic obliquity is assessed by lying the patient in a prone position with the hips and knees hanging free. Infrapelvic causes of pelvic obliquity such as hip subluxation/dislocation or adductors contracture are evaluated and managed appropriately. Suprapelvic causes of pelvic obliquity arise from the scoliosis and are assessed clinically for flexibility and reducibility.

When a significant curve is identified, standing position (when possible) 36-in. posterior–anterior (PA) and lateral radiographs of the spine should be obtained. Radiographs in sitting position may be obtained if the patient is unable to stand; it may be necessary to support the head and trunk in severely affected children with poor truncal control. At our center, we use a standardized sitting frame with lateral support straps to obtain films in the sitting position with minimal external support.

Radiographically, the curve characteristics (curve type, magnitude, and progression), spinal balance (sagittal and coronal), pelvic balance (pelvic obliquity and tilt), and the growth remaining indicators (status of the tri-radiate cartilage and Risser sign) are documented (see Fig. 12.2a, b). Vertebral rotation with rib deformity and wedging suggest that the deformity is structural rather than positional. Among the various techniques of pelvic obliquity measurement, horizontal pelvic obliquity has the least intra-observer and inter-observer variability [11]. The patient with established scoliosis due to CP requires at least yearly follow-up examination to assess curve progression, but with severe curves or during periods of rapid growth, biannual follow-up is desirable.

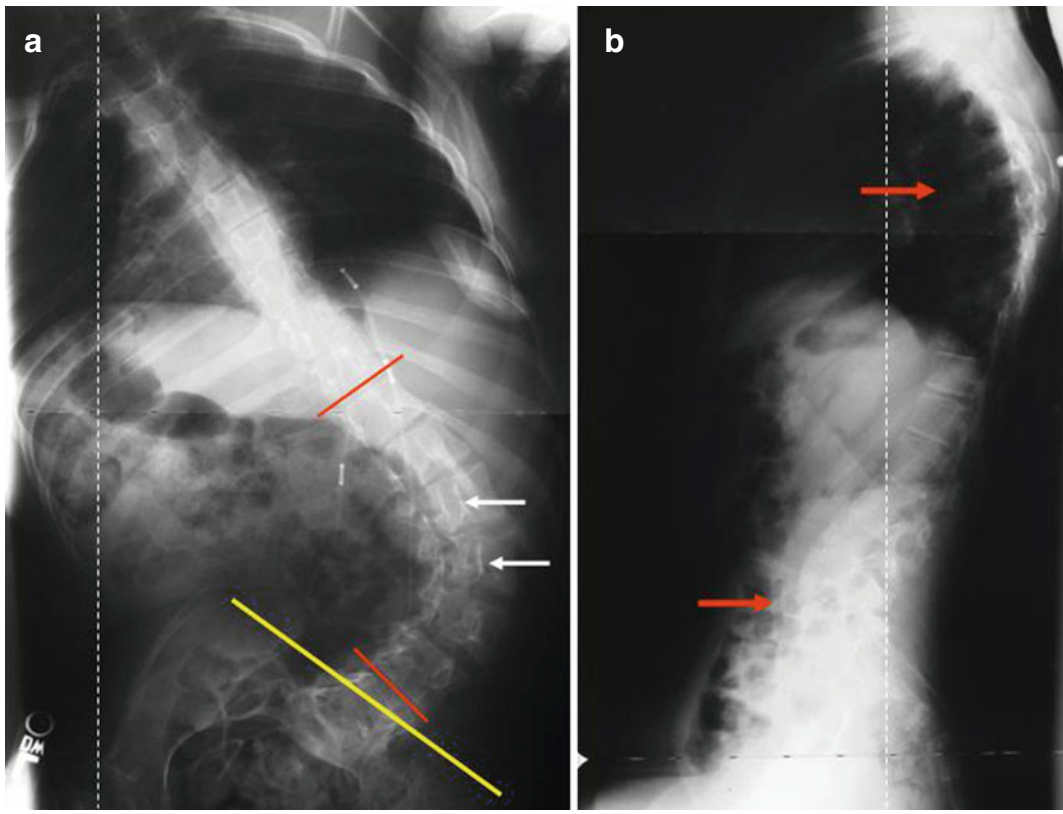

Fig. 12.2 When evaluating a patient with CP scoliosis, there are other radiographic parameters in addition to the Cobb angle (*red lines*). Note the severe apical rotation (*white arrows*), pelvic obliquity (*yellow line*) (**a**), hyperkyphosis, and hyperlordosis (*red arrows*) (**b**) and coronal and sagittal imbalance (*dashed lines*)

A magnetic resonance imaging (MRI) should be obtained if there is any suspicion of intraspinal pathology, such as very rapid progression at a young age, increasing lumbar hyperlordosis, or a change in neurologic status, which could be harbingers of a tethered cord.

12.4 Non-operative Care

In the global planning of disease management, several factors need to be considered. Of paramount importance are the alleviation of pain, preservation of function, and facilitation of daily care. Non-operative management of patients with neuromuscular spinal deformities should be directed at maximizing sitting ability and postural control to facilitate interaction with the surrounding environment. A normalized eyebrow-chin angle allows visual and cognitive stimulations with motor response.

Initial close observation of curves that are 20° or less is reasonable; if progression occurs, initial intervention with a brace may be an option. The role of bracing in CP scoliosis is dependent on the severity of the curve and neurological involvement. In a patient with spastic quadriplegic CP, it is generally accepted that bracing is ineffective but may slow the rate of progression. Miller et al. found no impact of a rigid thoracolumbosacral orthosis (TLSO) on scoliosis curve, shape, or rate of progression in spastic quadriplegic patients that were braced 23 h per day over a mean period of 67 months compared to a similar cohort that were not braced and were followed to spinal fusion [12]. Terjesen et al. [13] retrospectively examined a cohort of 86 patients with spastic quadriplegic CP and found a mean rate of progression per year of 4.2° with a custom-molded polypropylene TLSO. Interestingly, 25 % of the patients had no progression or progression of less than 1° per year. The degree of curve correction in the orthosis appeared to correlate with non-progression of the curve. Of note, Terjesen et al.'s study had a mean initial Cobb angle of 68.4°.

Although it may not alter the final disposition, a soft (polypropylene foam) TLSO can provide seating support and augment function. Improved sitting in a child may correlate with attentiveness in class, ease of care, improved self-image, and decreased rate of decubitus ulcers.

Another option for patients with flexible curves in need of seating support is the adjustment of offset lateral chest supports and modular seating systems on the wheelchair. This three-point control of the coronal deformity will prop up the child and address sitting balance. The wheelchair should be the primary seating device. In an ambulatory patient (GMFCS level I-II), it is believed that a hard brace may slow the progression of the curve similar to the patients with adolescent idiopathic scoliosis. The brace is indicated beyond 25° in immature patients with significant growth remaining. The brace should be worn for a minimum of 12 h. An optimal brace time is 16–18 h per day. Therapeutic stretching, electrical stimulation, or botulinum toxin is lacking scientific validity and should have no role in the management of deformity.

12.5 Rationale of Operative Care

Goldberg argued that the potential gains from interventions should be assessed by the following components: functional health gain, patient satisfaction, and technical success [14]. Given the wide spectrum of disease presentation and progression as well as the concomitant variability in functional status of the patient, the decision to proceed with operative correction and stabilization is based, in large part, on patient-specific factors with the broad aim of maintaining the functional health against the progressive deformity and its associated morbidity, achieving reasonable patient/caregiver satisfaction, and minimizing the complications associated with the surgical intervention.

For higher functioning patients, operative intervention aims to provide a more normal spinal balance and alter the progression of disease with the goal to preserve function with respect to ambulatory potential. The parents or caretakers can make an informed decision weighing the risks and benefits for their children.

For patients with no ambulatory potential (GMFCS 5), the aim is to maintain independence in sitting and facilitate care. As expected, the burden of care in this group of patients with severe learning disability may change significantly [15, 16]. As observed by Madigan and Wallace [4], the severity of scoliosis is directly proportional to the severity of involvement of CP. Concern has been raised regarding the risk of an extensive surgical procedure in a medically compromised patient. Surgical treatment in this group represents a palliative measure that allows the family to provide maximal medical treatment with the intent of caring for the child at home and keeping the child involved in school and other outside, community activities.

A prospective study by Larsson et al. in a cohort of neuromuscular scoliosis with a varied spectrum of learning disabilities found that the overall care burden decreased with improved sitting position and lung function (vital capacity) on follow-up [15]. Comstock et al. [17] assessed both patient and caregiver satisfaction in a cohort of 100 patients with total-body-involvement spastic CP who underwent spinal fusion. The satisfaction of both caregivers and patients was assessed via interview responses to standardized questions, and physical examination was used to assess functional status. Eighty-five percent of the parents interviewed indicated that they were satisfied with the results and would repeat the surgery again. There was an impression by caregivers that the patients had an improved self-image, and patients who were able to respond to questions confirmed this. Both parents and caregivers felt that the surgery had a positive impact on the patient's sitting ability, physical appearance, comfort, and ease of care. Multiple authors including Bulman et al. [18], Sussman et al. [19], and Watanabe et al. [20] found similar satisfaction rates in their studies.

12.6 Indication for Surgery and Specific Considerations

In general, surgical intervention is considered for curve magnitude greater than 50° with significant deterioration in function [17, 22, 27]. There is sufficient evidence that these curves will progress, even if the child has completed his growth. For curves 60–90°, surgery is indicated when the deformity becomes stiff by physical examination, even if substantial growth remains. If the spine displays continued flexibility on physical examination during growth, surgery can be delayed until 90° and can still be performed with a posterior-only procedure. In a flexible curve of greater than 90°, sitting may be a challenge and further exacerbated by the associated pelvic obliquity.

In planning for surgery, specific considerations should be given to the level of instrumentation, early-onset scoliosis, sagittal plane deformity correction, pelvic and infrapelvic coronal deformities, intraoperative neuromonitoring, the necessity of anterior release, intraoperative femoral traction, and intrathecal baclofen therapy.

12.6.1 Level of Instrumentation

Patients with neuromuscular scoliosis are traditionally fused long, typically from T1/T2 to the sacrum including pelvic fixation. An increased incidence of proximal curve progression, especially proximal junctional kyphosis, has been observed if the cephalad level of instrumentation does not extend to at least T2 [21], since most of these children lack sufficient head control. In patients with pathological thoracic kyphosis, it may be necessary to extend the instrumentation to C7 for adequate thoracic kyphosis control.

Historically, there has been debate regarding when to extend the posterior spinal fusion to the pelvis. Pelvic obliquity has been noted to progress in neuromuscular scoliosis if the pelvis is not fused [8, 22, 23]. Many authors have recommended fusion to the pelvis in nonambulatory patients. In the ambulatory patient with pelvic obliquity, fusion to the pelvis has been traditionally avoided due to the belief that it will adversely affect ambulatory function [3, 24]. At our institution, a retrospective study by Tsirikos et al. [25] demonstrated preserved ambulatory function in ambulatory patients with CP that were fused with

unit rod instrumentation, documented by gait analysis.

A subset of patients may be instrumented to L5 particularly if they use the gluteus maximus to propel their gait due to weak gastrocnemius. Significant pelvic rotation is expected clinically during ambulation. McCall and Hayes [26] retrospectively examined a cohort of patients with neuromuscular scoliosis in whom those with a stable lumbosacral articulation were instrumented with a "U-rod" (unit rod without the pelvic limbs) with L5 pedicle screw fixation. The L5–S1 interspace mobility was assessed on the basis of L5 tilt; patients with more than 15° of L5 tilt were instrumented with a standard unit rod construct. McCall and Hayes [26] found in follow-up that the patients that were instrumented to L5 with the U-rod had similar results to those fused with the standard unit rod construct.

12.6.2 Early-Onset Scoliosis in CP

A severe curve in pre-pubertal growth period presents a management dilemma. The options include continued observation, surgical intervention with growth-friendly spine implants to control the curves, or premature spinal correction and fusion.

The experience of growing rods in 27 children with CP at the mean age of 7.6 years showed 47 % correction of the Cobb angle from a mean of 85° [27]. The multicenter study highlighted complications in 19 of the 27 patients. Eight patients experienced deep wound infection (8/27, 30 %). Other complications include rod-related complications (11 occurrences) and anchor-related complications (six occurrences) in this group of vulnerable patients. Eight patients in the cohort had no complications and had four rod lengthenings at an interval of 11 months [27]. A similar complication rate was noted with the "Eiffel Tower" VEPTR construct [28].

Early spinal fusion in a cohort of 33 patients with a mean age of 8.3 years and mean curve of 85° with a minimum follow-up of 5 years was reviewed. Patients with early-onset scoliosis in this group of neurologically severely involved patients (31 patients GMFCS V) had a 28 % mortality rate, and six patients died between 1 and 5 years and 2 died between 10 and 15 years after surgery. Deep infection was reported in three patients (3/33, 9 %) [29].

Clearly, the ideal management plan is yet to be determined. An optimal surgery should be a growth-friendly spinal implant without the need for subject patients to undergo repeated surgeries whether for lengthening purposes or implant-related problem. A magnetic-driven growing rod has recently been FDA approved and can lengthen without surgery. Its role in early-onset spinal deformity in children with CP is yet to be defined.

12.6.3 Sagittal Plane Deformities

Sagittal plane deformities such as hyperkyphosis or lordosis may develop in patients with neuromuscular disorders, either with or without scoliosis. Flexible, postural deformities may be addressed in younger patients with tight hamstrings by lengthening the posterior thigh musculature and addressing the associated posterior pelvic tilt and pelvic retroversion in these patients or by appropriate modifications to the wheelchair or shoulder harness, but in older children, these adaptations do not work as well.

The spinal column lengthens with lumbar hyperlordosis correction and shortens with thoracic hyperkyphosis correction. Exclusion of a tethered cord is important prior to embarking on the surgical correction of lumbar hyperlordosis. Patients who have undergone a previous dorsal rhizotomy for spasticity can be at particular risk for developing a pathological hyperlordosis and associated spondylolisthesis. This has implications during posterior surgical exposure. The authors have experience with postoperative radiculitis after correction of hyperlordosis and relative lengthening of the lumbar spine with presumable nerve root tension.

Lumbar hyperlordosis and its associated pelvic anteversion and obliquity alter the trajectory of the pelvic fixation significantly and can be a risk factor for pelvic fixation-related complications [6]. Medial breach of the ilium resulting in bowel perforation by the limb of a unit rod has

been described. A modular screw-based system is recommended [30] to decrease morbidity with pelvic screw placement, allow customization, and afford deformity correction.

12.6.4 Pelvic and Infrapelvic Coronal Plane Deformities

Compensatory scoliosis arises from coronal plane deformities of the pelvic and infrapelvic origin. Asymmetrical forces of the gluteus medius and hip adductors coupled with infrapelvic pathology such has hip subluxation and dislocation contribute to pelvic obliquity. In young patients, soft tissue procedures such as adductor and iliopsoas release could be attempted to achieve coverage of the femoral head and level the pelvis. With growth, the deformities can become stiff and need to be addressed by osteotomy of the proximal femur and pelvis. In such cases, spine surgery to restore spinal balance and pelvic obliquity is performed prior to the osteotomy of the pelvis for femoral head coverage.

12.6.5 Intraoperative Neuromonitoring

Spinal cord monitoring with intraoperative transcranial motor-evoked potentials and somatosensory-evoked potentials is controversial in this population [31] since meaningful monitoring is difficult. Up to 30 % of the patients with severe CP may have weak or absent signals at baseline, particularly transcranial motor-evoked potentials in the most severely affected children [32, 33].

Intraoperative neuromonitoring changes present a significant management dilemma. The Stagnara wake-up test is usually not possible. In the subgroup that responds to intraoperative optimization of physiological parameters and surgical correction, it could potentially advert neurogenic bladder (requiring urinary catherization) and maintain protective sensation even in the most neurologically involved patients. In the subgroup that has lost signals despite optimization, staging the procedure in this medically challenging group versus in situ correction is debatable. Problematically, the patients may not have reliable signals during the staged procedure. Involvement of the family in the potential decision-making is helpful to determine the course of action.

12.6.6 Anterior Release

Anterior release at the apical levels is indicated for stiff curves or curves greater than 90° not reducible with a pull or fulcrum bend film to gain flexibility and allow correction. Anterior release at the lumbosacral region includes psoas muscle recess at its origin, annulus release, and complete anterior discectomy, which helps with correction of pelvic obliquity and pelvic tilt [34]. With anterior surgery, complications and morbidity increase. Keeler et al. reported significantly higher infection, pulmonary and cardiovascular (coagulopathy or hypotension) complications when anterior release was employed [35]. Thoracoscopic anterior release is possible from the intervertebral disc of T4/5 to T11/12 and could reduce the operative time and morbidity associated with open thoracotomy.

It is unclear whether to stage the anterior and posterior procedures separately (1–2 weeks apart) or to do both the procedures on the same day. Evidence exists to support both strategies, and it is our practice to stage surgeries for patients with severe involvement and multiple medical comorbidites [36]. For relatively healthy patients, we usually perform both stages on the same day, provided that the time under anesthesia or blood loss is not too substantial after the anterior release. Anterior fusion for the so-called crankshaft phenomenon is not necessary, even for young patients, when rigid, segmental instrumentation such as a unit rod or pedicle screws are used posteriorly [37–39].

12.6.7 Intraoperative Halofemoral Traction

Intraoperative halofemoral traction is useful particularly in patients with kyphoscoliosis or significant pelvic obliquity [35, 40, 41]. Its use,

however, is less optimal in patients with lumbar hyperlordosis where the traction on both legs may aggravate the lordosis. Anecdotal experience suggests that unilateral traction prior to corrective maneuvers is useful in leveling the pelvis.

12.6.8 Intrathecal Baclofen Pump

Intrathecal baclofen pump therapy is increasingly being employed to control muscle spasticity while maintaining muscle function. For patients with intrathecal baclofen therapy, great care is taken to ensure adequate padding at the site of the pump during prone positioning. The concurrent insertion of the pump with the spinal deformity surgery does not increase the rate of infection, and simultaneous procedures are not substantially difficult [42]. No significant cerebral spinal fluid (CSF) leakage is expected during insertion of the intrathecal component of the tubing. The pump including the connecting tubing at the intrathecal sac can be safely inserted or exchanged even post-spinal fusion below the conus medullaris.

12.7 Surgical Evolution and Outcome

Spinal instrumentation and fusion are indicated for collapsing deformities and painful sitting when no other alternatives exist [43]. Historically, fusions with Harrington instrumentation had an unacceptably high rate of pseudarthrosis in 18–27 % of cases [5, 17, 44, 45]. The advent of segmental instrumentation with Luque rod and sublaminar wiring yielded improved results over the Harrington system [19, 21, 46–48] and obviated the need for prolonged postoperative casting. Comstock et al. [17] found a mean correction of 51 % in a posterior-only instrumentation cohort 57 % and in an anterior–posterior cohort.

Multiple authors have noted progression of pelvic obliquity if the fusion was not extended to the pelvis [17, 19, 23]. The Galveston technique to extend the fusion across the pelvis by placing each Luque rod between the pelvic tables [49] demonstrated acceptable fusion rates across the L5–S1 segment and provided good control of pelvic obliquity. It was associated with a high incidence of loosening secondary to micromotion at the ilium at the sacroiliac joints, which was described radiographically as the "windshield-wiper" effect. While the impaction of two Luque rods into the pelvis with associated segmental fusion via sublaminar wires provides a strong construct in the sagittal plane, there exists a moment arm of rotation about the two rods allowing for rod translation with respect to one another, loss of torsional control, and subsequent progression of pelvic obliquity, pseudarthrosis, and implant failure [50]. The use of Luque rods smaller than one-fourth-inch diameter may increase the incidence of implant failure [21, 23, 51], but the intraoperative bending of one-fourth-inch diameter steel rods to the optimal geometry for pelvic implantation presents a technical challenge. Lonstein et al. found in a cohort of 93 patients a 50 % correction of the major scoliotic curve with a mean preoperative scoliosis of 72° and 40 % correction of pelvic obliquity at a mean follow-up of 3.8 years using a dual Luque-Galverston instrumentation technique [52]. With a similar construct, Sanders et al. [23] found that postoperative residual curve greater than 35°, preoperative curves greater than 60°, crankshaft deformity, and not fusing to the pelvis were the factors associated with postoperative curve progression. It is clear that rigid fixation is essential for surgical success.

The unit rod developed by Bell et al. [50] addressed some of the potential limitations of dual Luque rod instrumentation. The implant design of a proximally connected, pre-contoured rod provides for better rotational control, as the degree of rotational freedom between two independent Luque rods is eliminated. Tsirikos et al. found in a cohort of 241 patients, a mean correction of 68 % from a mean scoliotic curve of 76° and pelvic obliquity correction of 71 % at a mean follow-up of 3.9 years [6]. This correction was more effective than dual Luque rod instrumentation. Westerlund [39] and Dias [37] found similar results.

In neuromuscular scoliosis, segmental instrumentation system using Cotrel-Dubousset instrumentation of hooks is limited to patients with S-shaped curves without the need to extend to the pelvis. In patients with associated pelvic obliquity, hybrid constructs using iliosacral screws for pelvic fixation with hooks allows pelvic obliquity correction of 40 % [53, 54] with the posterior only approach and 47 % in an anterior–posterior approach. There is a tendency of in situ rod derotation to bring about coronal correction. As expected, the reduction technique does not allow a significant biomechanical advantage in reducing the pelvic obliquity over a unit-rod with sublaminar wires.

The smooth Galveston rods and iliosacral screws were precursors to iliac screws. The dissection required for iliac screw placement is less than that of iliosacral screws and Galveston rods. Biomechanically, a threaded iliac screw has significant better pullout strength than a smooth Galveston rod for pelvic fixation, as it extends anteriorly beyond the pivot point of lumbosacral motion [55]. The shorter pelvic limbs of Galveston rods may pull out and become prominent posteriorly. The use of segmental pedicle screw constructs has shown substantial improvement of fusion rates at the lumbosacral junction while accomplishing the goals of leveling pelvic obliquity and addressing seating problems. The modular systems can navigate some of the substantial challenges in these patients, such as osteoporotic bone, three-dimensional deformity of the pelvis including a rotated pelvis [56], and lumbar hyperlordosis, and avoid some of the risk in early instrumentation failure.

The iliac screw is offset and connected to longitudinal rod members with a connector. Careful rod engagement with adequate length caudad to the tulip head of the connector is required to avoid rod disengagement. Using four iliac screws at the pelvis improved bony purchase at the construct foundation but did not eliminate the windswept wiper effect at the iliac screws [57]. Implant prominence can be a problem in a thin patient. Placement of the screw caudad to the natural prominence of the PSIS with bony recession may avoid this problem.

Sacral alar-iliac (SAI) fixation was popularized by Kebaish et al. [58, 59] and has the advantage of an iliac screw without the prominence of the implant as it is placed 15 mm deeper to PSIS. The tissue dissection is less than that required for iliac screw and pull out strength is similar to an iliac screw, and it may result in lower infection rate. The screw extends across sacro-iliac joint anteriorly beyond the pivot point of the lumbosacral junction and serves as an effective flexion moment against movement at the lumbosacral junction. The tulip screw head is aligned with the instrumentation array without the need of an offset connector.

In combination with iliac screws or SAI screws, lumbar pedicle screws with reduction tabs allow an effective sagittal and coronal control while allowing reduction of pelvic obliquity. This is particularly useful in patients with lumbar hyperlordosis and pelvic anteversion where the trajectory of the iliac anchor could be challenging.

In a cohort of adolescent CP patients who were managed with posterior-only all-pedicle screw constructs, Tsirikos et al. [30] reported 72 % correction of the major curve with the mean of preoperative of 76; the correction of pelvic obliquity was 80 % from a mean of 22° in the posterior-only subgroup. In our practice, pedicle screws hybrid constructs achieve a similar result. Pedicle screws are inserted at the most caudad thoracic vertebra and lumbar vertebrae. Sacropelvic fixation is achieved with S2Al screw using favored angle iliac screw with reinforced shank. A stable pelvic fixation allows a strong cantilever force to level the pelvis using dual custom rods, which are proximally connected. The reduction of the rods starts at the caudad lumbar screws and proceeds in the cephalad direction. The intervening thoracic levels are instrumented with sublaminar wiring. All screws construct are used when the significant correction is needed at the thoracic region as well.

Pedicle screw-based modular construct allows superior Cobb correction and leveling of pelvic obliquity with considerable economic cost than that achieved by unit rod. This is mitigated by less implant-related complications and lower rate

of infection, as shown by Sponseller et al. in a multicenter series [60], which are the two most common complications encountered aside from pulmonary-related issues [61].

12.8 Preoperative and Perioperative Concerns

The individual with CP scoliosis is medically complex and may pose significant preoperative risk. The risk and complications of a procedure of this magnitude are directly related to the severity of neurological impairment. Lipton et al. [62] have reported that a child who is not fed orally is severely mentally retarded, cannot speak, and has seizures and one who cannot sit independently, by far, has the highest rate of complications. Medical management of seizures, respiratory problems, nutrition deficiency, gastroesophageal reflux, and motility issues should be addressed before surgery.

Some of these children may be on a ketogenic diet for seizure control. Anesthesiologists need to be aware of this, as these children are more susceptible to hypoglycemic episode intraoperatively and medical and nutritional management is needed to manage tight metabolic control.

Standard preoperative laboratory work including hematology, metabolic profile, urinalysis, and a coagulation panel should be obtained as well as an assessment of nutrition, but we have found that the laboratory values are not always a reliable assessment of the preoperative status of the child. Blood loss can be substantial, and a type and crossmatch of 1–1.5 times the patient's blood volume should be available prior to the start of surgery [43]. Coagulation factor replacement and core body temperature maintenance are also important if substantial bleeding is encountered. Blood loss tends to be earlier and larger in this population during posterior spinal surgery resulted from qualitative defect despite a normal PT and PTT [63, 64]. The use of cell salvage and antifibrinolytics can be an important adjunct in decreasing allogenic transfusion requirement and blood loss during surgery. Antifibrinolytics such as tranexamic acid (TXA) and epsilon-aminocaproic acid (A) can be used. Aprotinin has been withdrawn due to the concern of higher mortality rate in cardiac surgery [65]. TXA is shown to be more effective then AMICAR with the loading dose of 100 mg/kg given over 30 min followed by a 10-mg/kg infusion until closure of the incision began. The infusion should be limited to maximum of 8 h [66]. The authors have used this regimen for 3–4 years with an excellent safety profile.

Many parents and caretakers have noted that they were not prepared for the complexity of the patient's postoperative course [17]. Preoperative counseling of the family and caretakers should stress the potential of a prolonged intensive care unit stay, as well as the significant possibility of postoperative complications, which can prolong the hospitalization.

Intraoperatively, the surgeon must maintain constant communication with the anesthesia staff. Intraoperative hypothermia is the most commonly encountered problem by the anesthesiologist (55 %) and could contribute to the coagulopathy. Active warning blanket with monitoring is essential to prevent hyperthermia in this group of patients with impaired thermoregulation [67]. The patient is most at risk during induction of anesthesia and preparation of the patient during IV line access before skin preparation and draping.

Intraoperative hypotension is encountered in 15 % of the cases and frequently secondary to inadequate volume replacement from chronic underhydration, increased sensitivity to anesthetic agents, and greater blood loss [67]. Correction of a kyphotic deformity can also impede venous return to the heart with resultant hypotension [43], which could be mitigated by increasing the pre-load volume prior to the start of the correction. In the event of hypotension during curve correction, an attempt to release pressure on the spine should be made and an increase in the rate of intravenous fluid and/or blood replacement should be performed; after the blood pressure has been stable for 5–10 min, it may be safe to proceed with a gradual correction to allow time for the soft tissues to stretch. If an episode of sudden hypotension with our without bradycardia

occurs, anaphylaxis should be considered, perhaps due to an unknown latex allergy or reaction to colloid or blood product replacement.

12.9 Surgical Technique

After intubation, appropriate monitoring leads, and establishment of large bore IV access and arterial and central venous catheterization, the patient should be placed prone on a radiolucent table or four post frame. Care should be taken to ensure that all bony prominences are well padded to avoid skin breakdown, especially in thin patients or patients with baclofen pump and the abdomen should hang free. The hips can be allowed to gently flex with knee and thigh support to passively correct lumbar hyperlordosis. We used unilateral intraoperative skin traction on the side with high pelvic obliquity during the corrective maneuver.

A standard posterior exposure of the spine from T1 to the sacrum is done subperiosteally, out to the transverse process with the use of Cobb elevators and electrocautery for hemostasis. Care is taken to preserve the cephalad inter and intraspinous ligaments. An aggressive posterior release with facetectomies and ligamentum flavum resection is important in creating flexibility in the rigid apical portion of the curve, and in all but the largest, stiff curves makes a posterior-only approach sufficient for correction of the scoliosis. Concave osteotomy at the apical segments may be necessary. Concave release of the taut iliolumbar ligaments at the tip of the L5 transverse process may be needed for severe, stiff pelvic obliquity.

At the inferior margin of the incision, posterior superior iliac spine (PSIS) is exposed when placing iliac screws. A notch is cut out with an osteotome 1 cm caudad to the most prominent part of the PSIS to avoid screw head prominence, and the cancellous bone between the inner and outer table is cannulated with a drill or pedicle gearshift. We have found that successful iliac screw fixation is possible with a mini-access approach to avoid extensive muscle dissection of the paraspinal muscle at the lumbosacral junction and outer table of the pelvis by using intraoperative fluoroscopy. By tilting the image intensifier obliquely in the plane of the iliac wing and cephalad so that it is parallel to the cortical bone of the sciatic notch, the "teardrop" of the ilium can be visualized (Fig. 12.3a–c). Iliac screw placement in this area ensures excellent fixation in strong cancellous bone, adequate length to extend past the pivot point of the lumbosacral junction, and safe avoidance of the sciatic notch and acetabulum (Fig. 12.4a–c). The iliac screw can then be connected to the longitudinal members of the thoracolumbar construct to level the pelvis (Fig. 12.5a–c).

When an S2 alar-iliac (SAI) screw is placed in lieu of a traditional iliac screw, the exposure at the caudad margin of incision is minimal. The starting point is at the midway of S1 and S2 foramen in line with S1 pedicle screws. The screw traverses SI joint and has the same end point as iliac screw. The trajectory is guided by the radiographic "teardrop" with a cannulated gearshift and pointed to anterior inferior iliac spine (AIIS) for maximum bony purchase while avoiding sciatic notch and hip joint. The bony isthmus is usually at 60-mm mark. A curve gearshift that points cephalad in relation to the plane of ilium allows longer screws to be inserted while gliding away from the direction of the hip joint [68]. A guidewire is then inserted prior to the drill and screw insertion. A typical screw of 8 mm in diameter with a length of 65–80 mm is used (Fig. 12.6a–d). In hyperlordosis, a more horizontal trajectory is expected. In a neutral pelvis, 30° lateral angulation is expected, which often needs to be adjusted for rotational deformity.

The freehand technique is used for thoracic and lumbar pedicle screw insertion. Meticulous preparation of the entry landmarks (pars, mammilary bodies, and transverse process) is warranted. The pars interarticularis leads to the lumbar pedicle entry point, which is at the intersection of mid-transverse process and mid-facet joints. The entry point is cannulated with a curve gearshift with the curve part pointed laterally. The gearshift is removed at 20-mm mark to point medially. Polyaxial reduction pedicle screws are placed in the lumbar vertebrae. At the thoracic

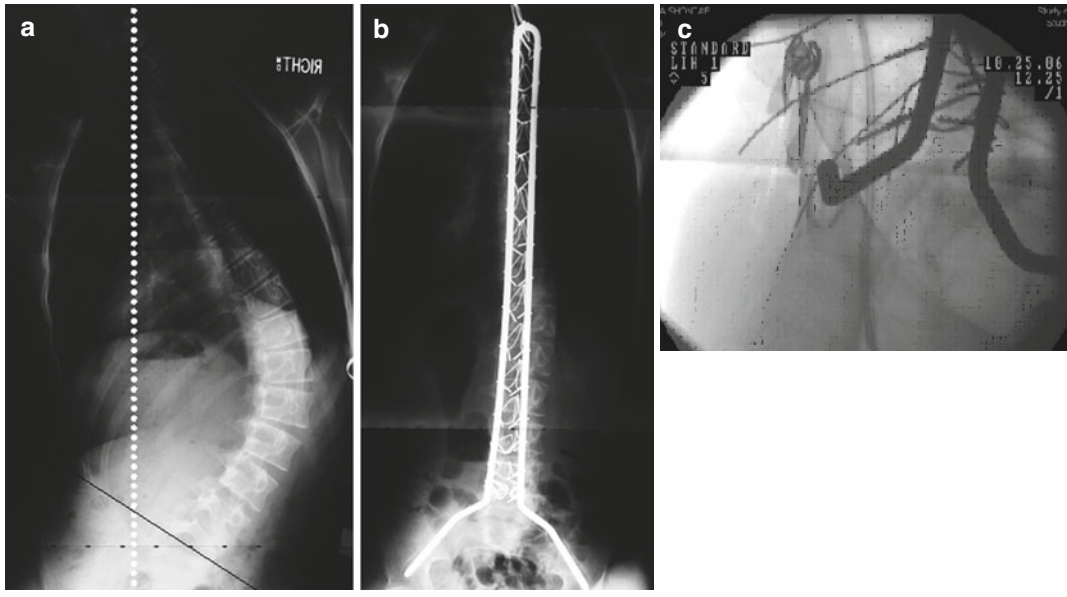

Fig. 12.3 (**a**, **b**) Preoperative and postoperative PA sitting radiographs of an ambulatory child with cerebral palsy scoliosis after posterior spinal fusion with the unit rod. Note the restoration of truncal balance and pelvic obliquity. (**c**) Intraoperative view of a pelvic limb of the unit rod placed between the inner and outer table of the ilium, in the "teardrop"

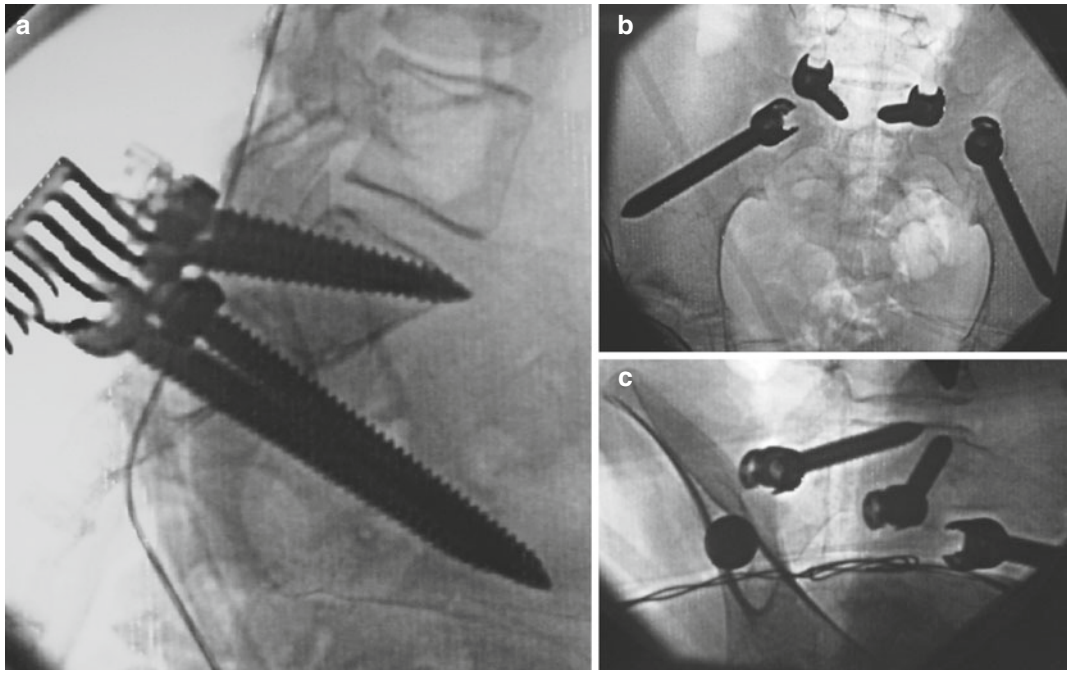

Fig. 12.4 (**a**–**c**) Lateral, AP, and oblique views showing satisfactory placement of pedicle screws in S1 pedicle and iliac screws

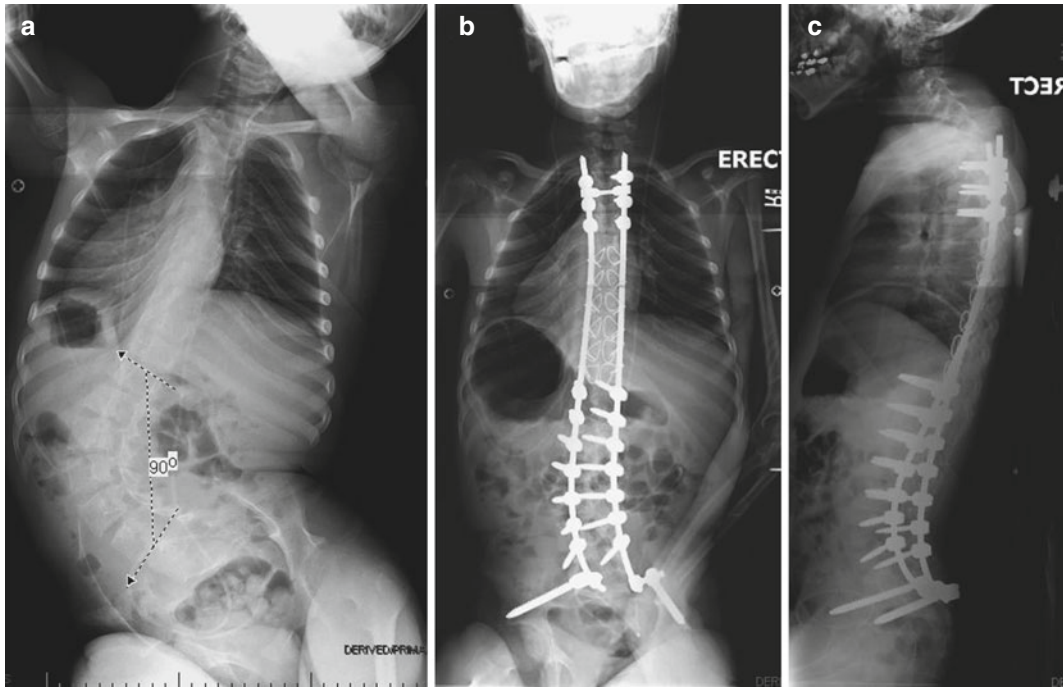

Fig. 12.5 (**a**–**c**) Radiographs of a 12-year-old male with quadriplegic cerebral palsy and scoliosis with a 90° curve, lumbar hyperlordosis, 30° pelvic obliquity, and seating difficulties. Wide posterior releases were used to make the curve more flexible, and the spine was corrected with modular segmental instrumentation using pedicle screws, sublaminar wires, and iliac screws

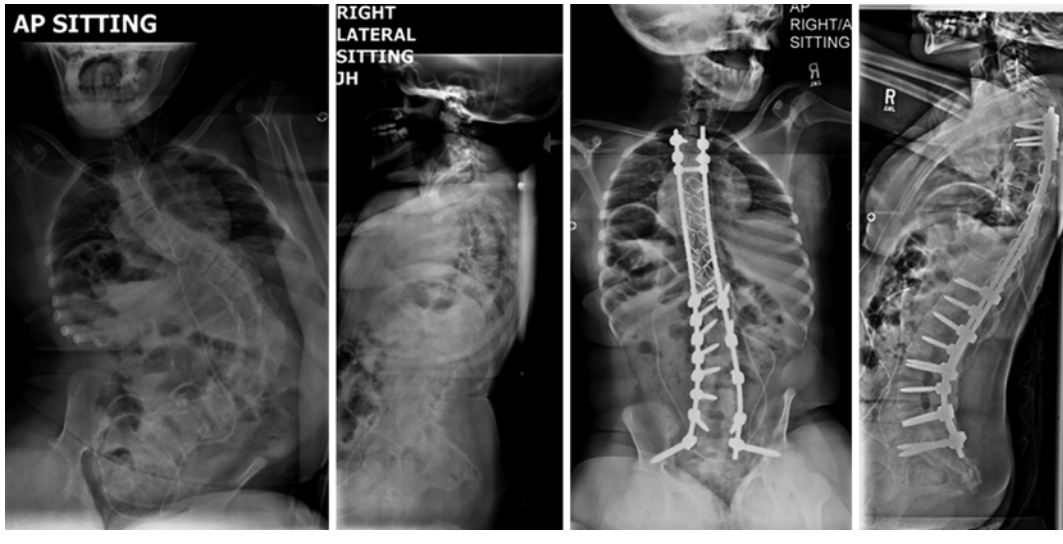

Fig. 12.6 (**a**–**d**) Radiographs of a patient with quadriplegic cerebral palsy and severe scoliosis, lumbar hyperlordosis, 35° pelvic obliquity, and seating difficulties. The spine was corrected with modular segmental instrumentation using pedicle screws and SAI screws

T2, a straight thoracic gearshift is used. The entry point of the thoracic pedicle screw is at the intersection of mid-transverse process and slightly lateral to the mid superior facet point. Fluoroscopic guidance occasionally may be required particular at the concave side of T2.

The spinous process of the thoracic level is removed to expose the ligamentum flavum. Care must be taken to preserve the laminas as they are key to the strength of fixation, especially the supralaminar cortex, which even in osteoporotic bone, can be strong. After the removal of the spinous processes and exposure of the sublaminar space, the sublaminar wires are passed at each level. A 16-gauge double Luque wire is passed at each level from T5 to T12. The wire is passed from inferior to superior. After passing the wire, it is contoured back over the lamina and the ends of the wire are contoured to the edges of the incision; this will maintain the intraspinal portion of the wire against the undersurface of the lamina as the remaining levels are instrumented. While passing the wire, care must be taken to avoid levering off the lamina and impinging against the cord; the diameter of the contoured bend should approximate the length of the lamina.

The implant metallurgy property should be carefully matched to the individual patients. Custom-bent 5.5-mm cobalt chrome or stainless steel rods are typically used. Note that the correction of a thoracic hyperkyphotic deformity will shorten the spine and the correction of a lumbar hyperlordotic deformity will lengthen the spine. The rods are differentially bent and connected to the SAI screws or iliac screws. A cross-link is placed at the cephalad part of the rods. The reduction-tab lumbar pedicles screws allow insetting of the rod. The spine is manually corrected to rod. Pushing the rod to the spine can generate substantial force at the lever arm of the pelvic insertion with subsequent fracture. The set screws over reduction pedicle screws at the lumbar levels and sequential tightening of the thoracic sublaminar wires allow significant load sharing prior to generating substantial cantilever forces. After tightening and retightening the wires, the wires are cut 1-cm long and bent down to the lamina to avoid implant prominence.

Facetectomies and decortication are performed, and a preliminary grafting of the area under the rod is performed. Copious crushed cancellous allograft mixed with antibiotics such as vancomycin [69, 70] is packed, and the wound is meticulously closed. A drain is generally not used.

12.10 Postoperative Care

Extubation in the operating room should be considered. Postoperatively, the patient should be maintained in an intensive care setting for 24–48 h and volume status and urine output closely monitored. The hemoglobin should be maintained over 9 g/dL to ensure adequate perfusion, and the coagulation parameters and platelet count should be corrected as needed, as these patients are frequently coagulopathic. Prophylactic antibiotics are continued for 24 h. In patients with poor nutritional status, enteral hyperalimentation should preferably be started early in the postoperative period either via a G- or J-tube. There is no need for immobilization postoperatively, and the patient should be mobilized out of bed and into a wheelchair as soon as medically appropriate. The child's personal wheelchair should be readjusted to accommodate his new trunk proportions and pelvic alignment. Children can return to school in 3–4 weeks, when sitting tolerance is attained, and no postoperative restrictions or orthoses are employed.

12.11 Complications

As previously discussed, a patient undergoing spinal fusion for neuromuscular scoliosis frequently has significant associated medical comorbidities, and postoperative complications are prevalent and should be anticipated. The incidence of postoperative complication has been noted to range from 18 % to 68 % [17, 21, 46, 71]. Curves 70° or greater, severity of neurologic involvement, and severity of recent history of medical problems have been shown to increase the risk of postoperative complications [62]. Respiratory complications are frequent, namely, atelectasis, or more

severe problems requiring prolonged ventilatory support. Postoperative ileus, pancreatitis, superior mesenteric artery syndrome, pulmonary compromise, and cholelithiasis can occur [72–75], and the physician must be vigilant in evaluating any clinical abnormalities.

Postoperative wound infections are of particular concern and tend to be the highest among patients with neuromuscular scoliosis [76, 77]. The rates of infection have declined steadily from 90 % in the 1970s to a current rate of 6-11 % [37, 47, 78–81]. Patients with G- or J-tube, unit rods, significant residual curve, skin breakdown [80], and implant prominence [82] are at risk. Most deep infections in the early postoperative period respond well to drainage and irrigation with delayed wound closure over drains or a vacuum-assisted device with intravenous antibiotic therapy and retention of the instrumentation. Patients with a previous infective episode are more likely to have pain at follow-up [83]. Pseudarthrosis is more commonly found in the setting of previous infection, and a workup should be undertaken for persistent pain or radiographic evidence of implant failure.

12.12 Outcomes

Correction of neuromuscular scoliosis with a pedicle screw-based hybrid construct is typically 75–80 % with leveling of the pelvis and excellent sagittal alignment. With proper surgical technique and rigid instrumentation, fusion rates are superior, and pseudarthrosis can be avoided. The parent and caregiver satisfaction is very high for this procedure, and over 85 % of the caregivers noted benefits beyond sitting and facilitation of care for the child postoperatively [15, 17, 37]. In a group of children that included even the most severely involved, there was a predicted 70 % survival rate at 11 years following surgery [84].

12.13 Summary

In summary, scoliosis is common in this group of children with neuromuscular disorders. The majority of these children have progressive spinal deformities that interfere with sitting and other functions and will require surgical stabilization to address these problems and facilitate care. A pedicle screw-based hybrid construct is the preferred method of instrumentation and offers a powerful mechanism of correction in both the coronal and sagittal planes. The risk of complications both perioperatively and postoperatively is substantial but manageable. Caregiver satisfaction is high after this procedure and affords a good long-term outcome.

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Other Neuromuscular Diseases

13

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Key Points

- Progressive neurologic and muscle diseases can cause progressive and severe scoliosis.
- These diseases commonly affect the pulmonary and cardiac systems, which need to be considered when managing the scoliosis.
- Early intervention in Duchenne muscular dystrophy is recommended to optimize cardiac and pulmonary function.
- Pelvic obliquity is commonly associated with scoliosis, and in many cases, pelvic fixation is recommended.
- Segmental fixation is recommended in the osteopenic bone commonly found in patients with neuromuscular scoliosis
- New methods of treating early-onset neuromuscular scoliosis are being evaluated including growing rods, VEPTR, and Shilla technique, but further studies are needed to determine its efficacy.

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13.1 Duchenne Muscular Dystrophy

Duchenne muscular dystrophy (DMD) is an X-linked inherited disorder isolated to the dystrophin gene causing an absence in the protein dystrophin [1]. DMD is usually first diagnosed by the age of 5 years. Initial complaints by parents include delayed walking, clumsiness, or flat feet. It has been suggested to screen any boy not walking by 18 months for DMD [2]. A later concern by parents, usually at age 4 or 5 years, is their son's inability to keep up with his peers or increased difficulty climbing up stairs. Other clinical findings seen on examination include pseudohypertrophy of the calves, proximal muscle weakness, Achilles and iliotibial band contractures, and a positive Gowers' sign.

In suspecting the diagnosis of DMD, the initial laboratory test is evaluating serum levels of creatine phosphokinase (CK). The diagnosis is then confirmed by genetic testing. In the remaining one-third of patients, a muscle biopsy is needed to specifically assess the quantity and quality of dystrophin present.

13.1.1 Spinal Deformity

Spinal deformity is the most critical orthopedic issue for the patient with DMD. The incidence of scoliosis is about 95 %. The onset of spinal deformity usually occurs at the same time that patients lose the ability to walk between the ages of 10 and 14 years. The risk of progression of scoliosis is also very high. Smith et al. reviewed the natural history of 51 patients with DMD and scoliosis who had no surgical treatment and were followed until their death [3]. Seventeen of these patients had curves greater than 90°. The mean rate of progression was 2.1° per month. In many cases, the curves continued to progress until the rib cage contacted the ilium.

The spinal deformity associated with DMD differs from the deformity seen in adolescent idiopathic scoliosis (AIS). The rate of progression is greater in muscular dystrophy patients [4, 5]. Unlike the typical hypokyphotic or lordotic patient with AIS, most progressive scoliosis in DMD patients is kyphotic in the sagittal plane. Wilkins and Gibson suggested two types of spinal deformity in DMD [6]. The more stable deformity is associated with an extended position, while the unstable pattern is characterized by a progressive kyphosis [6, 7]. Oda et al. also utilized sagittal alignment to help differentiate the deformity in DMD into three types, recommending surgery for the kyphotic deformities [8].

Considering that scoliosis develops once the patients becomes wheelchair bound, screening is not needed while the patient is ambulatory. However, once the patient is unable to walk, radiographic screening should occur every 6 months.

13.1.2 Medical Considerations

In addition to the orthopedic manifestations, there are considerable medical complications associated with DMD. The problem that is most concerning for the spine surgeon is the progressive worsening of pulmonary function. Muscle weakness, contractures, and spinal deformity result in a restrictive disease pattern. This progressive decline typically occurs in the second decade of life, worsens with increasing age, and ultimately leads to the patient's death [9–11].

Kurz et al. strongly suggested that age and curve severity negatively affect pulmonary function [4]. Forced vital capacity peaked when the patient became unable to stand. Each year of age following this then resulted in a forced vital capacity decline of 4 %. If the patient developed scoliosis, an additional decline of 4 % occurred for every 10° of thoracic scoliosis. A study by Yamashita et al. also supported the relationship of scoliosis and pulmonary function [12].

Since age and thoracic scoliosis were the best predictors of pulmonary decline in their study, Kurz et al. recommended early surgical intervention in the DMD patient [4]. Others have also made similar recommendations. Galasko et al. demonstrated slightly improved survival and maintenance of forced vital capacity for the first 36 months in those patients that underwent

surgery before progression occurred [13]. Rideau et al. found static vital capacity at 2 years in five surgically treated DMD patients [14]. Recently, Velasco et al. supported spinal stabilization, demonstrating a significant decrease in the rate of respiratory decline post-surgery compared with pre-surgery rates [15].

Some authors have contradicted the positive effects of surgery on pulmonary function. Their studies found no significant difference between the surgical and nonsurgical group in terms of declining respiratory function [16–18]. Kennedy et al. demonstrated a similar decline in forced vital capacity of 3–5 % per year in both operative and non-operative patients [19]. The criticism of this study was that the surgical patients had severe scoliosis with pulmonary function too poor to benefit from surgery [20]. A recent Cochrane Review by Cheuk et al. was unable to give an evidence-based recommendation regarding the effect of surgery on pulmonary function since no randomized controlled clinical trials have been performed [21].

Prior to any spinal surgery, preoperative pulmonary function tests should be performed. Typical problems encountered included prolonged intubation and the need for permanent tracheotomy. Recently, studies have suggested that with aggressive postoperative pulmonary management, patients with low forced vital capacity could successfully undergo spinal fusion [22, 23]. Of the 45 patients prospectively collected, Harper et al. found no difference in outcomes between patients with a forced vital capacity greater than 30 % compared with those less than 30 %. We recommend that if spinal fusion is contemplated, early intervention should be performed before further decline in pulmonary function. Short ventilatory assistance followed by early extubation and aggressive pulmonary management minimize the risk of atelectasis and pneumonia.

Patients with DMD should also undergo a cardiac evaluation including echocardiogram. Cardiac involvement includes cardiomyopathy and conduction abnormalities [16, 24, 25]. In those patients with severely reduced cardiac function that cannot be controlled pharmacologically, surgery may not be an option.

Similarly to other myopathies, there is an increased risk of malignant hyperthermia in DMD [26, 27]. In extreme cases, patients have died intraoperatively from sudden cardiac arrest. Typically, anesthesiologists refrain from using anesthetics that trigger malignant hyperthermia. Awareness of the risk will maximize the preparedness of the entire team for these medically complicated patients.

13.1.3 Nonsurgical Management of Scoliosis

Spinal deformity in the DMD patient rarely develops in the ambulatory patient. Therefore, close screening of these patients should begin when the patient begins using the wheelchair fulltime. In those rare cases when scoliosis develops in the ambulatory patients, bracing should not be utilized. It has been suggested that bracing is ineffective and may end the ability to walk [20]. For the non-ambulatory scoliosis patient, bracing has also been discouraged. Multiple published reports have shown that while there is a decrease in the rate of progression, orthotics do not prevent the development of severe scoliosis [2, 28].

Since Drachman demonstrated positive outcome in the use of steroids for the treatment of DMD, there has been increasing work in investigating the effects on scoliosis [29]. Corticosteroids have been found to stabilize muscle strength for a period of time [30]. A recent Cochrane Review found evidence to support the use of steroids to improve muscle strength and function in the short term (6–24 months) [31]. However, it is not completely clear whether this will have any change in the management of scoliosis. Some studies have suggested that similarly to bracing, steroids can delay the progression of scoliosis [32, 33]. A previous prospective study compared 30 DMD patients treated with deflazacort with 24 similar control patients. While they suggested that steroids slowed the progression of scoliosis, they were unable to demonstrate the prevention of spinal deformity [33]. More recently, Lebel et al. compared long-term follow-up of ambulatory

DMD patients receiving deflazacort versus those not receiving the glucocorticoid [34]. At this long-term follow-up, they found that 20 % in the deflazacort group had developed scoliosis compared to 92 % in the non-treatment group. Currently, there are no data to support corticosteroids as a long-term option for the management of scoliosis. However, based on recent literature, it may be beneficial in the prevention of scoliosis. In addition, the use of steroids has to be balanced with the potential complications including weight gain, behavioral problems, fracture, glucose intolerance, gastrointestinal symptoms, skin changes, and cataracts [30, 31].

13.1.4 Surgical Management of Scoliosis

Posterior spinal fusion (PSF) and segmental spinal instrumentation (SSI) are the standard surgical treatments of DMD. For a patient with documented progressive scoliosis that can tolerate surgery, there is little controversy for the need for surgical stabilization. The goal is to maintain sitting balance and patient mobility and minimize the effect of scoliosis on pulmonary function. Due to the high likelihood of developing scoliosis, some authors have suggested performing procedures when patients lose the ability to ambulate [3]. This time period is when patients have maximum lung function and are most fit to withstand surgery from a pulmonary standpoint. Most authors, however, recommend surgery with radiographic evidence of scoliosis at about 20–30° [20, 35–37].

With the development of SSI by Luque, there have been major improvements in the surgical stabilization of DMD patients [38, 39]. SSI has improved the fixation in otherwise osteopenic bone and has minimized the need for prolonged immobilization. Currently, surgeons continue to effectively use the more traditional sublaminar wires with unit rods, while others have equal success with more modern instrumentation such has hooks or pedicle screws [40].

There is little controversy where the fusion should begin. It is recommended that the instrumentation should extend into the upper thoracic spine, typically at T2 [11, 20, 36]. Stopping short of this may allow for cephalic progression of the curve due to progressive trunk and neck muscle weakness, causing the patient to lose head control.

The caudal extent of the fusion, however, continues to have some controversy. Specifically, should the instrumentation end at L5 or the pelvis? Fixation to the pelvis is technically more demanding, increasing both operative time and the potential risk of complications [41, 42]. Sussman suggested that spinal fixation to L5 was sufficient in the early treated patients [37]. Mubarak et al. similarly concluded that instrumentation to L5 was sufficient if treatment was early when there was minimal pelvic obliquity (<15°) [36]. They prospectively followed 12 patients with fusion to the sacrum and 10 patients with fusion to L5 only. The mean follow-up was 7 years. Fusions to the pelvis took an additional 30 min longer. Review of the patients' sitting balance and postoperative pelvic obliquity demonstrated only minor differences between the groups.

Sengupta et al. evaluated fixation to L5 utilizing modern pedicle screws and compared them to standard Galveston fixation or L-rod configuration to the pelvis [41]. The minimum follow-up was 3 years. The pedicle screw group had a mean preoperative curve of 19.8° and pelvic obliquity of 9°. The pelvic group had a mean preoperative curve of 48° and pelvic obliquity of 19.8°. The pelvic group was about 2.5 years older at the time of surgery. The authors documented improved correction of the major curve and pelvic obliquity in both groups. They also acknowledged the difference in deformity between the two groups. Their conclusion was that pedicle screw fixation to L5 provide a solid foundation for those patients that undergo surgery when performed early with minimal pelvic obliquity.

Other studies have recommended fusing to the pelvis at the initial time of surgical intervention [43–47]. Patients are healthiest at the first surgery. Any attempts to later fuse to the pelvis in those that have progressive pelvic obliquity will pose a greater risk with their worsening medical condition. Alman and Kim reported on 48 DMD patients that underwent spinal fusion [43].

Thirty-eight patients with less than 10° of pelvic obliquity and 40° curvature underwent fusion and instrumentation to L5. Of these patients, 32 had progression of their pelvic obliquity. They found that curves with an apex below L1 were at the greatest risk of progression. Therefore, Alman and Kim recommended fusion to the pelvis for all curves with an apex below L1.

Gaine et al. evaluated 85 patients that underwent spinal fusions to either L4, L5, sacrum, or ilium [45]. They demonstrated that the more proximal the implant ended, the worse the correction of major curve and pelvic obliquity. Intrapelvic fixation maintained the best correction in pelvic obliquity. Interestingly, they found no difference in correction of the pelvic obliquity between instrumentation and fusion that terminated at L5 compared with those that ended at S1.

Brook et al. reported on the results of ten patients that underwent fusion above the pelvis with an L-rod and seven patients that had Galveston fixation to the pelvis [44]. Six of the L-rod patients experienced some curve progression and sitting imbalance. The criticism of this study is that eight of the ten patients had curves greater than 40° and that preoperative pelvic obliquity was not recorded [20]. In addition, four of the ten patients had their fixation end at either L3 or L4.

SSI in the thoracic and lumbar spine has traditionally been with the use of sublaminar wires. With advancements made in instrumentation, some have chosen to use hooks or pedicle screws for the stabilization of the deformity. Recent studies involving pedicle screw fixation have demonstrated improved major curve correction in patients with DMD [48–51]. Another study reported on improved patient function, sitting balance, and quality of life with pedicle screw constructs [52].

Selection of implants is related to surgeon preference, cost, deformity, and patient anatomy and is beyond the scope of this chapter. Currently, our preferred technique is to utilize pedicle screws in the lumbar spine as well as at the cephalad portion of the construct. Depending on the deformity as well as bone quality, we will utilize either sublaminar wires or pedicle screws in between in the thoracic spine (Fig. 13.1).

There are similar choices for instrumentation to the pelvis. Options include the Galveston techniques with either Luque or Unit Rods, Dunn-McCarthy technique with an S-rod, sacral screw, and iliac screw fixation [42, 44, 53–55]. Each has advantages and disadvantages. Galveston technique is subject to loosening and migration of the rod [11]. In addition, the Galveston technique sometimes requires complex three-dimensional contouring to fit the altered pelvic anatomy. Iliac screws, on the hand, are placed individually into each iliac wing and then connected to the rod through connectors. A recent study by Peelle et al. demonstrated equal effectiveness in controlling pelvic obliquity between the Galveston technique and iliac screw fixation [55]. Our preferred method is to utilize iliac screws when instrumenting to the pelvis in DMD patients.

Another important consideration in the preoperative planning for scoliosis is the risk of blood loss during surgery. Of all pediatric spine surgeries, Duchenne muscular dystrophy has demonstrated, on average, to have the highest mean level of blood loss [56, 57]. This is important considering their poor cardiac reserve. These patients require a large exposure from the upper thoracic spine to the lower lumbar spine or pelvis. The paraspinal muscles are difficult to elevate subperiosteally. Dysfunction of vascular smooth muscle as well as decreased platelet adhesion is thought to contribute to increased blood loss [57, 58]. Besides diligent hemostasis intraoperatively, the use of antifibrinolytics may help to minimize the blood loss. Shapiro et al. retrospectively evaluated the use of transexamic acid in 20 DMD patients and compared them with 36 control patients [59]. Transexamic acid was found to reduce intraoperative blood loss and the need for homologous transfusions. Other options which have been published for adolescent idiopathic scoliosis but not DMD include the use aminocaproic acid [60–62]. Vitale et al. investigated the efficacy of preoperative erythropoietin on hematocrit and transfusion rates in neuromuscular patients. They found no clinical benefit in their treatment group. We currently work with anesthesia preoperatively to ensure that each patient is administered an antifibrinolytic during surgery. Intraoperative blood loss is also collected in a cell

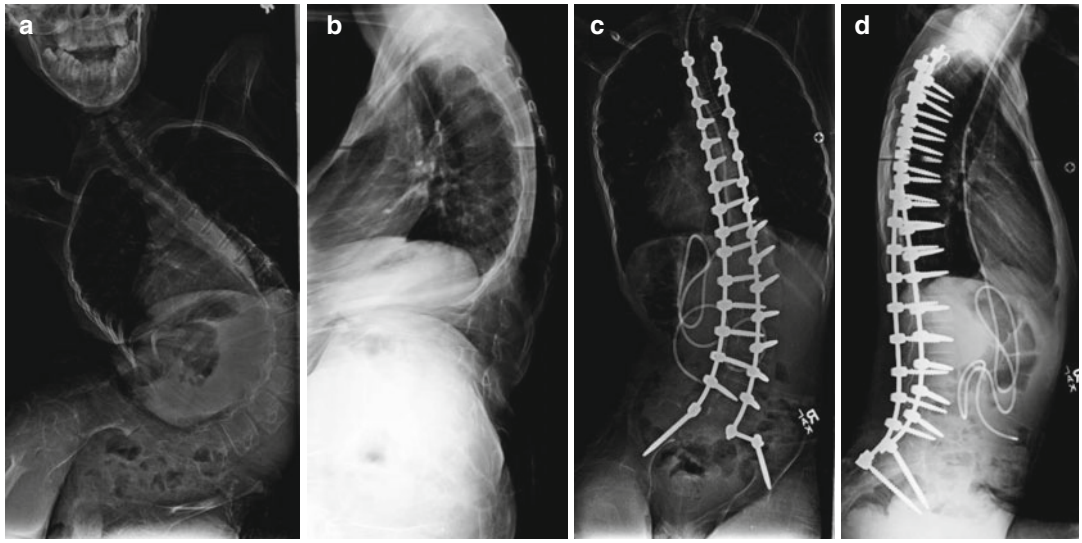

Fig. 13.1 (**a**, **b**) A 15-year-old male with DMD and delayed presentation of his progressive scoliosis. His lumbar curve is 128° and he has significant pelvic obliquity. (**c**, **d**) The patient underwent posterior spinal instrumentation and fusion from T3 to the pelvis with iliac screws. To assist with correction, intraoperative traction as well as multilevel Ponte type osteotomies were performed

saver and given back to the patient. Postoperatively, hematocrits are monitored closely to ensure that cardiac function is not overly stressed.

13.1.5 Long-Term Outcomes

As previously discussed, there is controversy whether scoliosis surgery improves pulmonary function in the DMD patient. A recent Cochrane Review by Cheuk et al. was unable to provide an evidence-based recommendation for scoliosis surgery in DMD. Their reasoning was the lack of randomized clinical trials. Of the 36 relevant studies addressing the outcomes of scoliosis surgery, none met the inclusion criteria for review.

Studies have suggested that scoliosis surgery does benefit patients beyond pulmonary function [63–65]. Bridwell et al. sent questionnaires to 33 patients with DMD evaluating function, self-image, cosmesis, pain, quality of life, and satisfaction [63]. Patients reported benefits in all categories with the highest ratings in cosmesis, quality of life, and satisfaction. Granata et al. and Takaso et al. found that sitting position, aesthetic improvement, and quality of life were all improved following spinal fusion [52, 64]. More than 90 % of their patients/parents would give their consent again for surgery.

13.1.6 Summary

Spinal deformity commonly affects the male patient with DMD. Treatment of this deformity is complicated by the progressive muscle weakness and deteriorating pulmonary function. Current literature suggests that surgical management of the deformity can maintain upright sitting posture, improve quality of life, and positively affect short-term pulmonary function. Unfortunately, a lack of randomized controlled trials has prevented any formal evidence-based recommendation from being made by a Cochrane Review. If surgery is contemplated, however, it should be performed early when the patient is at his or her maximal health. In addition, if there is more than mild pelvic obliquity, one should consider including the pelvis in the instrumentation and fusion.

13.2 Spinal Muscular Atrophy

Initially described by Guido Werdnig, spinal muscular atrophy (SMA) is a rare autosomal recessive disorder characterized by degeneration of the anterior horn cells of the spinal cord and the neurons of the lower bulbar nuclei [66, 67]. Two genes are associated with this disease: the survival motor neuron gene and the neuronal apoptosis inhibitory protein gene [68]. SMN protein appears to interact with RNA-binding proteins and is found in both the nucleus and cytoplasm of cells [69]. It is considered the most common fatal neuromuscular disease of infancy and the most common neuromuscular disease in children [66].

13.2.1 Classification

Common to all SMA patients is a symmetric muscular weakness predominantly affecting the lower limbs and proximal muscles compared with the upper limbs or distal muscles. Patients usually have normal intelligence with no effect on sensibility. The age of onset and clinical course can have a variable presentation. Due to this heterogeneity, spinal muscular atrophy is most commonly divided into three types [70–72].

13.2.1.1 Type I, Acute Werdnig-Hoffman Disease

Type I SMA is the most severe form of the disease, usually presenting at birth or within the first 2–6 months of life. These patients do not meet early motor milestones with the inability to gain head control, roll over, or sit up. It has been suggested that in utero osteoporosis from decreased movement is responsible for postnatal pathologic fractures [73]. Patients with Type I SMA usually do not survive beyond the age of 3 years. Respiratory failure from intercostal weakness and rib collapse is responsible for their mortality. Due to their early mortality, orthopedic intervention is rarely indicated in these children.

13.2.1.2 Type II, Chronic Werdnig-Hoffman Disease

The clinical onset of Type II SMA occurs between the ages of 6 and 24 months. Patients reach early motor milestones but are never able to walk independently. Weakness usually starts in the lower extremities, affecting the gluteal and quadricep muscles initially. Life expectancy is variable from adolescence to adulthood with some patients living into their fourth decade [66, 74]. The cause of mortality is respiratory failure.

13.2.1.3 Type III, Kugelberg-Welander Disease

The clinical onset of Type III SMA occurs following the age of 18 to 24 months. In nearly all cases, the diagnosis is made before the age of 10 years. As expected, children attain greater motor milestones compared with Type II SMA. Patients are able to walk independently until early adolescence. Russman et al. reported that 50 % of those children with an age of onset before 2 years lost their ability to walk without assistance by age 12 [75]. Those children that presented after age 2 typically were ambulating into the fourth decade. Patients that never reached independent ambulation lost their ability to walk by age 7.

13.2.1.4 Functional Classification

Evans et al. described a functional classification based on the maximum physical function achieved [76]. The purpose was to give insight into the patient's prognosis. Group I patients never sit independently, have poor head control, and develop early progressive scoliosis. Group II children have head control and ability to sit but cannot walk or stand. Group III patients can stand by themselves and are able to walk with external support. Group IV children can walk and run independently.

13.2.2 Diagnosis

For those patients that do not present at birth, presenting concerns by families are a delay in

reaching motor milestones. Depending on the age of the patient, these include an inability to gain head control, roll over, sit, stand, or walk independently. Physical examination should then assess motor strength as well as deep tendon reflexes. For those patients that present early (Type I or II), gross fasciculations of the tongue or tremors of the finger are commonly present [66, 77].

Once SMA is suspected, further diagnostic workup includes laboratory studies, nerve conduction studies, electromyography (EMG), and DNA testing. Creatine phosphokinase and adolase are usually normal or slightly elevated in Type III patients [78]. Motor and sensory nerve conduction velocities are normal. EMG findings demonstrate fibrillation potentials associated with denervation as well as large polyphasic motor units associated with renervation [66, 78]. DNA testing is highly sensitive for SMA with PCR the diagnostic procedure of choice [79]. Muscle biopsy is also highly diagnostic. Histologic findings include muscle fiber degeneration and atrophy with no evidence of primary myopathy [78].

13.2.3 Spinal Deformity

Scoliosis is the most common orthopedic problem in patients with SMA [80]. Nearly 100 % of Type II SMA patients and half of Type III SMA patients develop a spinal deformity [76, 81–83]. The deformity is typically a right-sided C-shaped thoracolumbar curve. Ninety percent of the patients have a single curve. The curve is usual progressive and in approximately a third of the cases associated with a progressive kyphosis [81, 82]. Similarly to DMD patients, development of scoliosis in type III SMA occurs with their loss of ambulation [80]. Pulmonary function is similarly compromised in patients with SMA [84]. The worsening of lung function is secondary to muscle weakness as well as the progressive scoliosis.

As there is a difference in the clinical presentation between the three types of SMA, there is similar heterogeneity in the risk and progression of scoliosis. Evans et al. demonstrated that the age of scoliosis onset correlated with the severity of muscle disease [76]. Type I SMA patients typically had scoliosis by the age of 2 years, while Type III SMA patients developed scoliosis between the ages of 4 and 14 years. The rate of progression was also highly associated with the disease severity ranging from 8.3° per year in severe cases to 2.9° in more mild cases.

As for the severity of the scoliosis, a study by Granata et al. reported curves ranging from 10 to 165° [81]. Schwentker and Gibson reported on 50 patients with SMA [80]. Seventy percent had scoliosis measuring greater that 20°, and 40 % had curves greater than 60°. The natural history of these large curves suggests that they can be quite disabling [76]. In addition to trouble sitting, patients can lose upper extremity function to maintain trunk balance as well as develop back pain or pain from rib impingement on the pelvis.

13.2.4 Nonsurgical Management of Scoliosis

Orthotics has generally been thought to be ineffective in preventing the development or progression of neuromuscular scoliosis [81, 85, 86]. It, however, has been shown to be effective in improving sitting balance. Letts et al. demonstrated an improvement in sitting stability in 80 % of patients with a collapsing neuromuscular scoliosis with the use of a soft Boston orthosis [87]. They also thought that a soft brace was more tolerable than a rigid orthosis and resulted in less skin breakdown.

Some studies have suggested that the use of orthotics may slow the rate of progression of scoliosis [82, 86, 88]. Slowing the rate of scoliosis progression has the advantage of allowing patients to get older when they are more suitable for a surgical intervention. This is especially critical in the early-onset patients (Type I and II SMA). Unfortunately, most of these studies report opinion and have not given reliable data to demonstrate that bracing truly slows the progression of spinal deformity. Bracing is also not without its morbidity. Aprin et al. reported on five

patients that had to discontinue their brace secondary to respiratory difficulty [86].

Our preferred nonsurgical treatment is to initiate bracing in patients with spinal deformities on sitting films between 30 and 40°. Typically, the curves in SMA are quite flexible and amenable to the orthosis. We find that the brace in addition to wheelchair supports help to maintain sitting balance. This is especially critical in pre-adolescent patients where attempts are made to delay surgery until the patient is more mature. In some cases, especially in the Type I patient where long-term survival or surgical tolerance is not expected, bracing may be the definitive management of the spinal deformity.

13.2.5 Surgical Management of Scoliosis

Similarly to other neuromuscular scoliosis, the decision to operate on an SMA patient is dependent on multiple factors. In general, the radiographic parameters for spinal fusion are not controversial and simple to follow. We recommend spinal fusion for curve magnitudes greater than 50° that are refractory to conservative measures and demonstrate progression. These indications for surgical fusion have been recommended by other authors as well [66, 76, 89].

Unfortunately, patient factors may not make the above rules simple to follow. In some patients with type I SMA, their early-onset scoliosis and grim long-term survival have made surgical intervention unreasonable. Type II patients may also present with a progressive scoliosis at an early age. Spinal arthrodesis would have a considerable negative effect on trunk growth as well as lung growth. These patients are also at significant risk of developing a crankshaft deformity necessitating an anterior fusion [90]. In these cases, spinal fusion is indicated, but an attempt at delaying surgery with the use of an orthosis is made. The goal is to maintain some control of the curve until a definitive procedure can be done at about the age of 10. Of course, this may mean watching a curve progress to greater than 80°.

13.2.5.1 Growing Spine Techniques

Previously, there has been some thought about using an expandable or "growing rod" construct in young SMA patients that developed significant scoliosis. Fujik et al. reported using an expandable or "telescoping" device in type II SMA patients [91]. The device was abandoned due to its technical demands and inability to prevent progression of the deformity and crankshafting. They concluded that a brace should be used until the age of 10 when a fusion can be performed.

However, more recent publications on early-onset scoliosis in SMA suggest increasing use of growing spine constructs. Growing rods may be the answer for these young patients. Chandran et al. found excellent deformity correction in 11 patients (mean age of 6 years) from 51° to 21.6° with a low complication rate at the initial surgery [92]. Sponseller et al. demonstrated in six patients that growing rods can be fixed to the pelvis and result in improved coronal balance, sagittal balance, and pelvic obliquity [93]. With similar constructs, McElroy et al. found improved major curve by nearly 50 %, improved trunk height, and improved space available for lung ratio at final follow-up. However, they did not find any halt in rib collapse, which is common in SMA. Additionally, they found that patients with SMA had longer hospital stays than did patients with early-onset idiopathic scoliosis undergoing the same procedure [94]. Recently, Tobert and Vitale published a case series of three SMA patients (aged 8, 7, and 3 years) undergoing rib to pelvis growing construct. They found stabilization of pulmonary function and overall improvement in quality of life and caregiver burden [95].

Additional literature is needed to know the long-term benefits and complications related to these surgeries. The tolerance for multiple anesthetics needed to expand the device on these already pulmonary compromised patients is also not known. To minimize this need for multiple anesthetics, we have utilized a modified Shilla technique to manage the early severe scoliosis (Fig. 13.2).

13.2.5.2 Posterior Spinal Fusion

In the older SMA patient that requires definitive spinal stabilization, the standard is PSF and

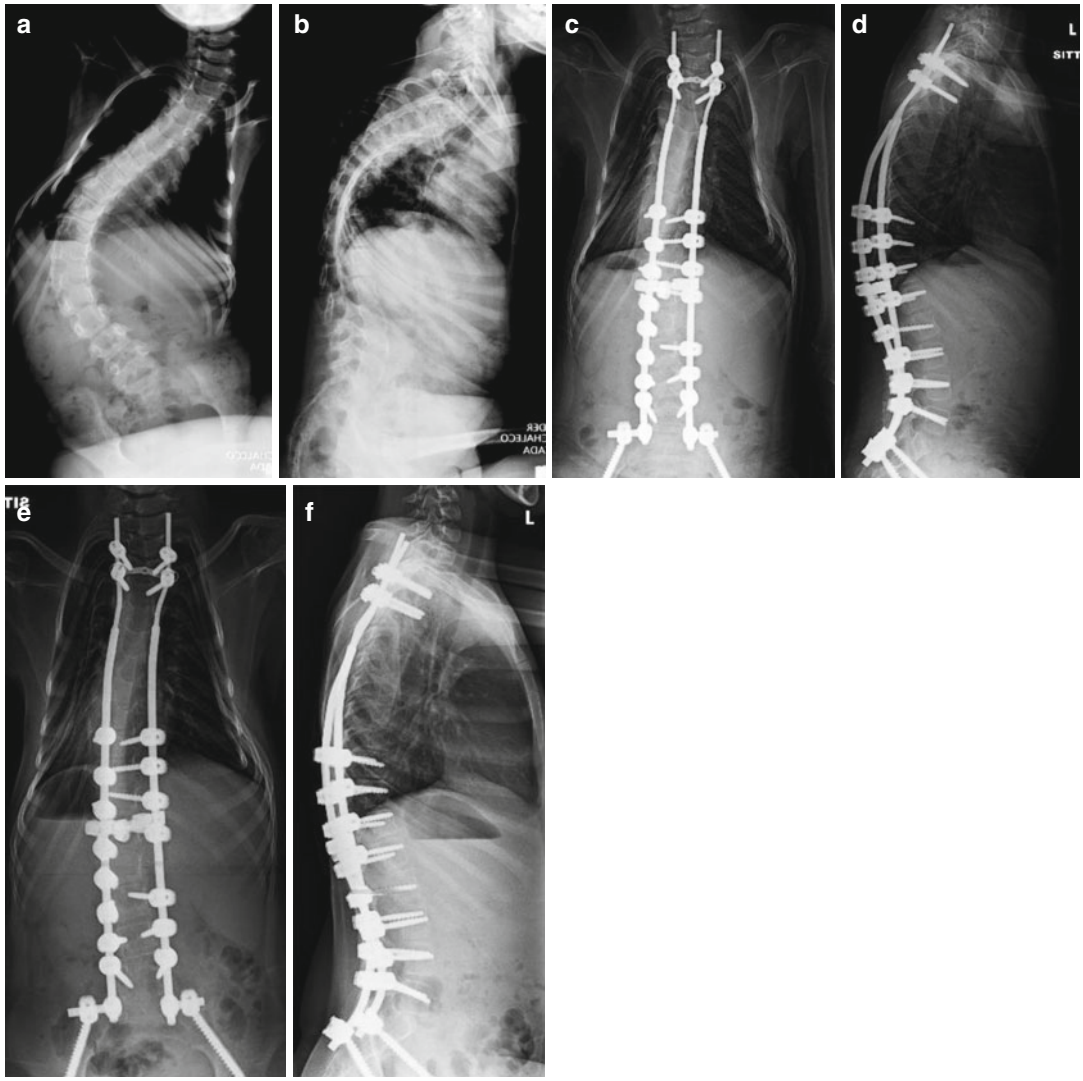

Fig. 13.2 (**a**, **b**) An 8-year-old female with SMA and progressive kyphoscoliosis. Her coronal major curve measures 90°. (**c**, **d**) The patient underwent a modified Shilla technique with instrumentation and fusion from T10 to the pelvis. The instrumentation was extended proximally to pedicle screws at T3 and T4 with fusion across these two levels. The pedicle screws are allowed to slide along the rod as the spine grows. The goal of this procedure is to attain correction with the distal fusion but allow thoracic growth with the Shilla technique. (**e**, **f**) At 1 year postoperative, the patient had grown nearly 1 cm as measured by the movement of the top screws

SSI. The goal is to prevent progression and obtain an alignment that will improve or maintain balance and sitting ability. In the non-ambulatory patient, this typically involves segmental instrumentation from T2 to the pelvis. Many spinal deformity surgeons report good outcomes using sublaminar wires with Luque rods or a unit rod for the treatment of neuromuscular scoliosis [40, 83, 96]. Others are transitioning to the use of pedicle screws to provide more rigid fixation [40]. The improved fixation to bone with pedicle screws has decreased the use of postoperative bracing for some neuromuscular patients [66]. We continue to brace all neuromuscular patients for 3 months postoperatively to prevent excessive stress on the osteopenic bone during transfers, including those with all pedicle screw instrumentation. Pelvic instrumentation is recommended to

prevent progressive pelvic obliquity and difficulty with sitting [80]. Similarly to patients with DMD, options include Galveston technique or iliac screws.

The use of an anterior approach has traditionally been reserved for severe curves or for patients at risk of developing crankshaft deformity. In the case of patients with SMA, other factors need to be considered. These patients typically have poor pulmonary reserve associated with weakness of their respiratory muscles. This places them at increased risk of developing pulmonary complications. The use of segmental fixation may decrease the risk of crankshaft deformity. Smucker and Miller reported on 43 patients with neuromuscular scoliosis and open triradiate cartilage treated with a unit rod [97]. They found no evidence of crankshaft deformity at 2-year follow-up. Some believe that pedicle screws may further decrease the risk by providing three-column fixation. However, this needs to be evaluated. There is also increasing evidence that severe spinal deformity can be completely managed from a posterior approach. Multilevel posterior osteotomies or single-level vertebral column resections stabilized with pedicle screws have been shown to adequately treat the severely deformed, rigid spine [98]. However, this, too, has not been adequately studied in patients with spinal muscular atrophy.

In preparation for spinal fusion, all patients with SMA should be evaluated by a pulmonologist, neurologist, and anesthesiologist. This will ensure that patients are optimized for surgery especially regarding their pulmonary function. In the immediate postoperative period, patients are most at risk of developing pulmonary complications. Aprin et al. reported a 45 % incidence of respiratory problems following surgery [86]. Four of their 22 patients required intubation. Brown et al. reported that tracheostomy was needed in 30 % of their patients [96]. The use of preoperative traction has been suggested to increase spinal flexibility and improve pulmonary function, possibly diminishing their risk of respiratory complications [86, 99]. Postoperatively, these patients should have aggressive pulmonary therapy and early mobilization. Ventilatory assistance with the guidance of a pulmonologist may be needed several days following the surgery. Other long-term complications following spinal arthrodesis include crankshafting, pseudoarthrosis, prominent implants, narrowing of the chest, gastric volvulus, and diaphragmatic rupture [63, 82, 86, 100]. Except for crankshafting, these complications were more commonly seen in older patients with larger deformities.

13.2.6 Long-Term Outcomes

In general, the literature supports spinal fusion in SMA patients with progressive scoliosis. Multiple authors have reported improvements in sitting, balance, comfort, and cosmesis [81, 99]. Bridwell et al. evaluated 21 SMA patients with an average follow-up of 7.8 years after surgery [63]. Patients reported benefits in all categories with the highest ratings in cosmesis, quality of life, and satisfaction. In contrast, some authors have reported a decline in some functional activities, specifically upper extremity activities. Brown et al. demonstrated a decline in self-feeding, drinking, and self-hygiene at 2-year follow-up with some improvement at 5 years [96]. Furumasu et al. reported similar findings suggesting that the lack of spinal flexibility diminished gross upper extremity motor function due to a change in trunk position. What is unclear in these patients is the influence of a progressive muscle disease in the diminished functional activities.

Pulmonary function also appears to benefit from stabilization of the scoliosis. Robinson et al. demonstrated a significant improvement in lung function in the patients that underwent spinal fusion [84]. They also demonstrated a significant inverse linear relationship between curve magnitude and percentage of predicted vital capacity.

13.2.7 Gene Therapy

Spinal muscle atrophy is caused by a mutation in the survival motor neuron (SMN) 1 gene that results in a reduction of the SMN protein. Patients also can have variations in the copies of the

SMN2 gene, which produces reduced levels of SMN protein. This production, however, is insufficient for normal motor neuron function [101]. There are promising gene therapy pathways that are being tested to increase the number of SMN proteins produced [102, 103]. One approach is to antisense oligonucleotides to redirect SMN2 translation and increase production of fully functional SMN protein [104].

13.2.8 Summary

Spinal muscle atrophy is a heterogeneous disease commonly affected by progressive scoliosis. Depending on the severity of the disease, patients can have significant deformity at a very early age. While ineffective at preventing scoliosis, bracing is utilized to delay surgery. When severe scoliosis develops at a young age (<10 years), growing spine constructs can improve spinal deformity and sitting balance and may improve pulmonary function and quality of life, but this needs further study. The gold standard for spinal stabilization remains posterior spinal instrumentation and fusion. Current literature suggests that surgical management of the deformity can maintain upright sitting posture, improve quality of life, and positively affect pulmonary function. Whether this improvement in pulmonary status improves life expectancy is still unclear.

13.3 Arthrogryposis Multiplex Congenita

Arthrogryposis or "arthrogryposis multiplex congenita" (AMC) is a heterogeneous group of diseases with the similar phenotype of multiple congenital joint contractures [105, 106]. Currently, there are more than 150 subtypes that result from a failure of normal movement in utero. The etiology for this lack of movement may be myopathic, neurologic, or secondary to connective tissue abnormalities [107]. Amyoplasia is the term used to describe the more classic disease entity seen in orthopedics. These patients have a dysgenesis of anterior horn cells resulting in replacement of muscle with adipose and fibrous tissue [108].

Patients with arthrogryposis multiplex congenita (AMC) have significant musculoskeletal deformities secondary to the contractures. The majority of patients have all four limbs involved (84 %) [105]. Severe equinovarus feet, hip dislocations (unilateral or bilateral), and scoliosis are commonly seen. Non-orthopedic abnormalities include hypoplasia of the labial folds, inguinal hernias, abdominal wall defects, cryptorchidism, gastroschisis, and bowel atresia [105].

13.3.1 Spinal Deformity

The incidence of scoliosis in AMC is reportedly between 30 and 67 % depending on the definition used [109, 110]. The deformities are similar to other neuromuscular conditions with lumbar and thoracolumbar curves predominating [111, 112]. The curves are frequently stiff. Progression of the deformity can be rapid, up to 6.5° per year [112]. The earlier the presentation of scoliosis, the more severe the curve may become and be associated with pelvic obliquity. Increased lordosis is frequently seen.

Scoliosis is typically refractory to orthotic management [111, 112]. Patients with arthrogryposis will frequently develop scoliosis early in life. Little literature has evaluated the treatment of early-onset scoliosis in these patients. Recently, Astur et al. and the Chest Wall Spinal Deformity Study Group evaluated ten children with arthrogryposis that underwent treatment with the use of the vertical expandable prosthetic titanium rib (VEPTR) device and found it to be an effective treatment method in these patients [113]. Using this rib-based distraction device, they obtained 37 % correction of scoliosis and 29 % correction of kyphosis. They also found improved thoracic volume. Six complications occurred in four patients in a total of 62 procedures performed. Proximal junctional kyphosis appeared to remain a problem, however, in this cohort. Other than this series, few studies have evaluated growing spine techniques in patients with AMC.

PSF and SSI remains the standard and appears to be effective in preventing progression of the scoliosis. However, correction of the curves appears to be modest, about 35 % [111]. Yingsakmongkol and Kumar reported slightly increased correction (44 %) with a combined anterior and posterior fusion [109]. These series are dated, however, and do not assess surgical outcomes with current segmental instrumentation. In some cases, instrumentation was not used. If pelvic obliquity is present, fusion to the pelvis should be attempted. Care should also be taken when positioning patients. Their stiff joints and osteopenia place them at increased risk of developing pathologic fracture.

13.4 Rett Syndrome

First described in 1966, Rett syndrome is a progressive neurologic disorder that affects one in 20,000 females [114, 115]. Patients initially appear normal at birth but then proceed through four stages of deterioration. The first stage typically has an onset between 6 and 18 months with developmental stagnation. The second stage (1–3 years of age) is characterized by lost language skills and autistic behaviors. In the third stage (2–10 years of age), patients may have seizures, exhibit some mental retardation, and have repetitive hand motions. In the fourth stage, patients develop spasticity and muscle wasting. Scoliosis is most likely to present in this final stage.

13.4.1 Spinal Deformity

The musculoskeletal manifestations of Rett syndrome include lower extremity contractures, coxavalga, and scoliosis [115, 116]. The spinal deformity is similar to other neuromuscular diseases with a long C-shaped curve being the most common [117, 118]. However, patients can also present with a single thoracic or double major curve. Large curves are frequently associated with pelvic obliquity. As a patient get older, so does the prevalence of scoliosis. Curve progression has been suggested to be more rapid than in idiopathic scoliosis or other neuromuscular scoliosis. Lidstrom et al. demonstrated greater that 15° per year of progression in the final stage of Rett syndrome [119]. For this reason, it has been recommended that patients are evaluated every 6 months following the age of 5 [120].

Bracing has been found to be largely unsuccessful in preventing the progression of scoliosis [117, 118, 120]. It, however, can be used to delay the need for surgical intervention to allow for more truncal growth. Posterior spinal fusion and segmental spinal instrumentation are the treatment of choice for the progressive scoliosis. In those patients that are non-ambulatory, it is recommended to fuse from the upper thoracic spine to the pelvis to prevent delayed decompensation or pelvic obliquity. Ambulation is possible in patients with Rett syndrome and can be positively affected by surgery. Harrison et al. demonstrated no loss of ambulation in all five patients that walked preoperatively and improvements in some patients [118]. Overall, PSF and SSI are successful in halting curve progression and improving spinal balance in the sitting and walking patient. Improvement in activities of daily living has been seen following spinal fusion in Rett syndrome patients [121]. However, medical complications can be high especially pulmonary (63 %) and gastrointestinal (37 %) in some series [122].

13.5 Congenital Myopathies

Congenital myopathies are a heterogeneous group of disorders characterized by weakness and hypotonia from birth [123]. Typically, the diseases have similar clinical findings but are classified based on histologic and microscopic findings. Central core disease, nemaline myopathy, and myotubular myopathy are just a few of the multiple congenital myopathies wherein scoliosis has been described [124–128]. They are genetically transmitted and can have variable penetrance.

13.5.1 Spinal Deformity

The musculoskeletal abnormalities associated with these disorders include congenital hip instability, foot deformities, other joint contractures, as well as scoliosis [127]. The curves are similar to other neuromuscular curves with a long, thoracolumbar shape. Kyphosis can also be associated with the deformity. As scoliosis progresses, it often becomes rigid. Rigid spine syndrome, as described by Dubowitz, has often been associated with these diseases and other congenital muscular dystrophies [129, 130].

If the patients present early and have flexible curves, scoliosis may be amenable to management with an orthosis. Those patients that fail bracing or present with large, rigid curves should undergo spinal fusion. Similar consideration as with other neuromuscular scoliosis must be given to the health and age of the patient. Poor pulmonary function has been associated with congenital myopathies [128]. At a minimum, patients should undergo preoperative pulmonary function testing. These patients are also at increased risk of developing malignant hyperthermia [124]. The anesthesiologist should be made aware of this before the day of surgery so that adequate preparation can be done.

Depending on the severity of the disease, patients may present with early-onset scoliosis. Those that demonstrate progression with the use of an orthosis may require surgical treatment with an expandable device or "growing rod." However, there have been no studies adequately evaluating the use of a "growing rod" in these patients. Those patients that present later in life do well with a posterior spinal instrumentation and fusion. Anterior release can be done for the large rigid curve if the patient can tolerate the exposure. In the non-ambulatory patient with pelvic obliquity, the fusion should be extended to the pelvis.

Similar principles to other neuromuscular diseases should be followed when treating patients with congenital myopathies. Posterior fusion is the treatment of choice. The need for traction or fusion to the pelvis should be determined on an individual basis. Depending on bone quality, a brace can be used postoperatively to support the instrumentation. Particular attention, however, has to be made towards the increased risk of hyperthermia.

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Myelomeningocele

14

Lawrence I. Karlin

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Key Points

- Myelomeningocele is a multisystem disorder with an extremely variable clinical presentation.
- The unifying pathology is a defect in the posterior vertebral bony elements and a malformation in the exposed, underlying neural structure. A level-dependent neurological deficit results.
- Associated abnormalities which may also affect the neurological status and lead to a potentially preventable spine deformity include the Arnold–Chiari malformation, hydrocephalus, symptomatic spinal cord tethering, and syringomyelia.
- Most involved children have some form of spinal deformity that is either present at birth or develops by age 10 years.
- Surgery is the definitive treatment of problematic spinal deformity, but is complicated by the anatomic abnormalities and the orthopaedic, neurological, and medical comorbidities.
- Segmental spinal instrumentation is a significant technical improvement that allows instrumentation of the dysplastic posterior bony elements and earlier mobilization.
- Growth preserving technology offers another option for the treatment of significant early onset deformity.

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14.1 Introduction

Myelomeningocele is the most severe form of spina bifida, the group of birth defects that have in common the abnormal fusion of the posterior neural tube. In children with myelomeningocele, the neural tissue protrudes through the deficient posterior vertebral elements and has no protective skin coverage. The neural tube is open; the neural folds are attached to the adjacent ectoderm; and the neural placode is exposed (Fig. 14.1). There will be motor and sensory deficits related to the level of the spinal cord lesion and accordingly the consequences of paralysis in growing children; developmental orthopaedic deformities, spinal malalignment, and some degree of neuropathic bladder dysfunction. Congenital orthopaedic abnormalities are also encountered.

In most neuromuscular diseases, the neurological status is predictably either stable or progressive. The clinical course of myelomeningocele is far less certain since a number of different central and peripheral nervous system abnormalities may be present. In addition to the spinal cord lesion with an established neurological level present at birth, there will usually be the Arnold–Chiari malformation and hydrocephalus and quite often syringomyelia and symptomatic spinal cord tethering. These additional problems will vary in their severity and may or may not cause neurological deterioration and orthopaedic deformities. They often require surgical treatment. When not causing neurological complications, they have the potential to do so during spinal surgery. Associated systemic anomalies occur (Table 14.1) and in some cases the defect is part of a syndrome involving chromosomal or single gene abnormalities (Table 14.2).

Myelomeningocele was a uniformly fatal diagnosis until the second half of the twentieth century and the advent antibiotic therapy and advances in urological and neurosurgical care. A seminal event, and an example of the remarkable commitment seen in the families of children with myelomeningocele, is the development of a shunt for the treatment of hydrocephalus. In 1955, John Holter was working as a technician in a hydraulics factory when his first son, Casey, was born with myelomeningocele and hydrocephalus. With encouragement of Dr. Eugene Spitz, a neurosurgeon, Holter dedicated himself to improving

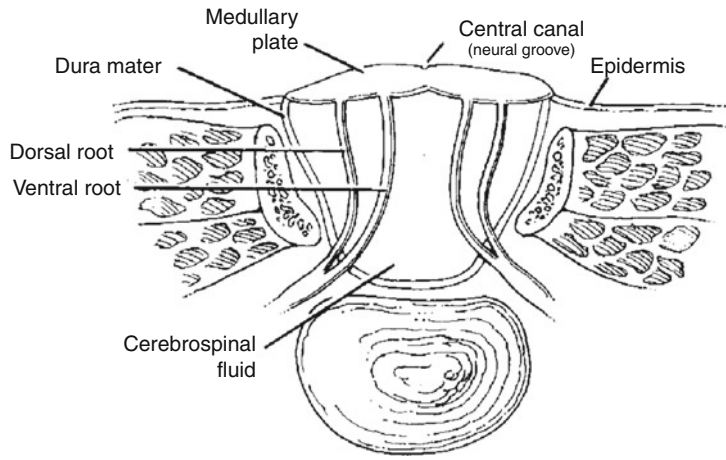

Fig. 14.1 The basic lesion of most children with myelomeningocele is the open neural placode. The dorsal surface is the interior of the neural tube while the ventral surface is what would have been the entire outside of the neural tube had it closed. There is no skin overlying the defect and the placode is covered by an extremely thin arachnoid that will breakdown shortly after birth allowing infection, meningitis, and death if closure is not performed. Passing down the center of the placode is a narrow groove that is continuous with the central canal of the closed spinal cord. Cerebral spinal fluid passing down the central canal is discharged through a small opening at the upper end of the placode and bathes the external surface of the neural tissue (Reprinted from Lindseth [45]. With permission from Lippincott Williams & Wilkins)

Table 14.1 Systemic anomalies associated with spina bifida

Skeletal	Gastrointestinal	Pulmonary	Craniofacial	Cardiovascular	Genitourinary
Clubfeet, vertical talus + other foot deformities Lower extremity contractures Hip dislocation Scoliosis Kyphosis Spondylolisthesis Pectus excavatum Syndactaly Rib anomalies Charcot arthropathy	Inguinal hernia Mickel's diverticulum Malrotation Omphalocele Imperforate anus	Tracheoesophageal Fistula Situs inversus	Synostosis Cleft palate Stabismus Low-set ears Hypertelorism	Ventriculo-septal defect Atrial-septal defect Patent ductus Coarcation	Hydronephrosis Hydroureter Horseshoe kidney Undescended testes Hydrocle Malrotation Exstrophy

Adapted from Reigel [76]. With permission from Elsevier

Table 14.2 Recognized syndromes including a neural tube defect

Genetic syndromes
Meckel
Median cleft face
Robert's
Anterior sacral meningomyelocele and anal stenosis
Trisomy-13
Trisomy-18
Triploidy
Others including unbalanced translocation and ring chromosome
Nongenetic syndromes
Syndrome of the amnios rupture sequence
Oculoauriculovertebral dysplasia

Adapted from Luciano and Velardi [48]. With permission from Springer Verlag

Table 14.3 Delayed neurological complications of spina bifida

Seizure disorder
Hydrocephalus
± Shunt malfunction
Arnold–Chiari malformation
Tethered spinal cord
Tethered spinal cord with tumor
Lipoma
Dermoid
Neurenteric cyst
Fibroma
Diastematomyelia
Arachnoiditis
Hydromyelia
Dermal sinus and stalks

Adapted from Reigel [76]. With permission from Elsevier

the current treatment options. Using Silastic, he produced a safe and functional shunt, and revolutionized the treatment of children with myelomeningocele [8].

14.2 Associated Abnormalities

The myelomeningocele child will have a level of paralysis based on the position of their spinal cord lesion, but the neurological status is notoriously unstable. These individuals must be monitored carefully to avoid the often preventable neurological deterioration or spinal deformity that may occur due to untreated hydrocephalus, syringomyelia, Arnold–Chiari malformation, or spinal cord tethering (Table 14.3).

14.2.1 Arnold–Chiari Malformation

In 1891 and 1896, Chiari described various anatomic patterns of herniation of the brain stem through the foramen magnum [23]. The Chiari II malformation, also known as the Arnold–Chiari malformation, is characterized by the displacement of the medulla oblongata into the cervical

canal and an upward course of the cervical nerve roots. Almost all children born with myelomenigocele will have the Chiari II malformation which can cause periodic stridor, apnea, swallowing difficulties, upper extremity paresis, hypertonia, nystagmus, and opisthotonis. The symptoms are worse in early childhood. In most affected children treatment of the secondary hydrocephalus with a ventriculoperitoneal shunt will alleviate the symptoms, but in some a decompression of the posterior fossa will be necessary.

14.2.2 Hydrocephalus

The Arnold–Chiari malformation obstructs the cerebral spinal fluid circulation. At birth, the open communication between the fourth ventricle and the central canal allows for decompression of the cerebrospinal fluid into the myelomeningocele sac. Once the sac is closed, this avenue of decompression is lost and hydrocephalus occurs. A ventriculoperitoneal shunt has been the standard treatment to avoid cortical damage, though recently endoscopic third ventriculostomy alone or in combination with choroid plexus cauterization has shown promise as a better treatment option [98, 99].

Hydrocephalus frequently recurs secondary to shunt malfunction. In children, the clinical signs of acute hydrocephalus are bulging fontanelles, altered mental status, nausea, vomiting, and severe headaches [65]. Hydrosyringomyelia may occur when the cerebral spinal fluid pressure within the central canal increases. Symptoms include increasing lower extremity weakness, spasticity, back pain, rapid progression of scoliosis, and upper extremity weakness. Early correction of hydrocephalus by shunt revision is usually curative.

14.2.3 Tethered Spinal Cord

The spinal cord is considered tethered when the conus medullaris is located at an abnormally distal level and fixed there by an inelastic structure. In the child with myelomeningocele, the spinal cord is tethered not by an expendable and easily resected thickened filum terminale, but by an adherence between the neural placode, the surrounding tissues, and the repaired dural layer. These children may also have dermoid inclusion cysts that add to the tethering or cause pressure on adjacent neural tissue. Most children who have undergone operative closure of a myelomeningocele will demonstrate a low lying spinal cord on magnetic resonance imaging (MRI) examination. In these individuals, the diagnosis of the tethered cord syndrome requires the presence of both anatomic tethering and neurological deterioration, and the absence of other causes of deterioration such as hydromyelia, shunt malfunction, and symptomatic Chiari II malformation. An MRI assessment will define the nature of the tether and assess the presence of possible associated pathologies including diastematomyelia, lipomas, dermoids, granulomas, inclusion cysts, and teratomas [37, 76].

Neurological deterioration due to a tethered cord will occur in up to 30 % or more of children with repaired myelomeningocele [90]. Release should be performed before major functional loss occurs. Although the conus level does not significantly change after detethering [17], the procedure can be expected to produce improvement or stabilization in most cases as illustrated by a comparison of outcomes in two long-term studies of sacral level myelomeningocle populations. In patients that had detethering procedures, none of 62 lost ambulatory ability and 61 were community ambulators [92]. In another similar study of 36 patients that did not have routine untethering, one-third had gait deterioration, 11 became wheelchair-dependent, and spinal deformities and lower extremity contractures developed in 44 % [16]. Results from untethering seem to vary with respect to the symptoms. The results are excellent for improvement of pain and motor deficits [54], but less successful for urological functional [20, 52] and accordingly timely intervention is key.

In properly selected myelomeningocele patients, tethered spinal cord release can stabilize a progressive scoliosis. Pierz et al. evaluated the effect of tethered cord release on scoliosis in 21

myelomingocele patients. Three had improvement in the curvatures, and six stabilized. Twelve patients progressed greater than 10°. Eighty-six percent of patients with initial curvatures over 40° and 100 % of those with a thoracic level required spinal fusion [72]. McLone et al. reported on 30 patients with myelomeningocele and scoliosis. In patients with curvatures greater than 50°, 1 out of 6 improved, while 14 of 23 with less severe curvatures stabilized [53].

14.3 Other Associated Abnormalities

14.3.1 Latex Allergy

Allergy to latex rubber has been reported in up to 28 % of individuals with myelomeningocele [10, 21, 51, 56, 58, 94]. These are IgE-mediated reactions characterized by urticaria, bronchospasm, rhinoconjuntivitis, laryngeal edema, and systemic anaphylaxis. The risk factors for intraoperative anaphylaxis are number of surgeries and atopic predisposition [32, 33, 39, 43, 63]. In the past, an attempt was made to identify latex-sensitive patients, but this strategy proved inefficient. A negative history for reaction does not eliminate the possibility of intraoperative anaphylaxis; skin testing lacks sensitivity and safety; and preoperative prophylaxis is not dependable. Accordingly, the present recommendation is that all myelomeningocele patients be treated in a latex-free environment.

Holzman, Gerber, and others have reported successful reoperation in patients that had previous anaphylactic reoperation by perioperative latex avoidance alone [14, 31, 40]. Additionally, a latex-free environment may diminish the number of myelomeningocele patients that become sensitized [64].

14.3.2 Precocious Puberty

Precocious puberty in girls with myelomeningocele is common. Proos et al. noted breast development by age 9 and related the risk of precocious puberty to increased intracranial pressure and shunt malfunction [74]. Furman and Mortimer noted that affected girls began menstruation at an average age of 10 years 3 months and earlier than their mothers and siblings [28].

14.3.3 Spinal Deformity

Spinal deformity occurs commonly and early in children with myelomeningocele. Piggott noted a 90 % incidence of some form of spinal deformity by age 10 years, and in half the patients the curvature was significant enough to merit surgical treatment [73]. The common deformities are scoliosis and hyperkyphosis which are often classified as congenital and developmental. Congenital deformities include scoliosis and kyphosis due to failure of formation or segmentation, and the rather unique form of kyphosis found in the myelomeningocele population due to dysplastic posterior elements. Developmental curvatures occur without vertebral malformation presumably due to muscle paralysis, muscle malposition, or hip deformity and pelvic obliquity [73, 75]. More recently, hydrocephalus, syringomyelia, and spinal cord tethering have been appreciated as causative factors. Spondylolisthesis has also been reported [42, 88].

Problems caused by spinal deformity include recalcitrant ulcerations over the gibbus in individuals with kyphosis or of the ischium or sacrum in wheelchair sitters with scoliosis and trunk imbalance. Pulmonary compromise can occur with significant scoliosis or kyphosis. Those with trunk imbalance in either the sagittal or frontal plane may need to use their hands for support and in doing so lose upper extremity function [6, 38, 41, 81]. Scoliosis is the most common deformity and will be discussed. Kyphosis is covered elsewhere in this text.

14.3.3.1 Scoliosis

Scoliosis of 10° or more will occur in 50–80 % of myelomeningocele children by 10 years of age [59, 73, 84]. Congenital scoliosis is present in 7–38 % of these [75, 80]. This will progress when spinal growth is unbalanced. Treatment is similar

to that of congenital scoliosis not associated with spinal dysraphism, but in the myelomeningocele group posterior fusion alone in the dysplastic portion of the spine is likely to fail due to the deficient bone stock. The addition of anterior fusion has been recommended [46].

Developmental scoliosis is the more common variety, and its incidence relates to the level of the neurological and anatomical lesion. Trivedi et al. defined scoliosis in this group as a curvature greater than 20° and noted the three most predictive factors for the development of scoliosis to be motor level, ambulatory status, and last intact laminar arch (LILA). The scoliosis prevalence was 93 %, 72 %, 43 %, and 7 % in patients with thoracic, upper lumbar, lower lumbar, and sacral motor levels respectively, and 89 %, 44 %, 12 %, and 0 % at similar LILA, respectively [95]. A rate of progression during growth of 12.5° per year in curvatures over 40° can be expected with a maximum rate in the 11–15-year-age group [61].

14.3.3.2 Orthotic Management

While many believe orthotic treatment to be at best temporizing [27, 34, 45, 65, 77], Muller and Nordwall [60] reported that the Boston brace arrested progression if initiated before the curvature reached 45°. Whether or not it prevents progression, an orthosis can improve function by providing improved support for seating. The rigid "active" control braces utilized in idiopathic deformities are problematic in the neuromuscular population, but total contact "soft" orthoses seem to be well tolerated. Letts et al. reported on the use of the "soft Boston orthosis" in the management of neuromuscular scoliosis. The brace is constructed of "Aliplast," a material similar to Styrofoam, and reinforced with polyethylene. While the improvement of the scoliosis averaged 15°, postural position or seating stability was enhanced 90 % [44]. Orthoses may be modified to accommodate individual needs (Fig. 14.2).

14.3.3.3 Surgical Indications and Planning

Surgery is frequently suggested for curvatures over 50° [9, 65] presumably because a deformity of this magnitude will be problematic or will progress to the point of being so. There is little evidence-based data to support either premise [103]. There are no long-term studies that document either the progression or clinical consequences of curvatures in adulthood. Improved sitting balance is an immediate benefit of scoliosis surgery but is obtained with considerable risk. Skin ulceration due to seating imbalance is another indication for surgical correction of deformity, yet the problem can be exacerbated following surgery if rigid residual pelvic obliquity persists [24, 66] or lordosis is diminished [52].

There are arguments that can be made for and against surgical treatment and discussions with families need to be balanced. Surgeons that believe that the natural history of untreated scoliosis is sufficiently problematic to require surgery should temper their discussions with reference to the paucity of evidence-based studies and the very real risks of complications and functional loss.

Preoperative Evaluation

This is a multisystem disease that requires an extensive preoperative evaluation. The skin in the area of the surgery must be assessed for scarring and fragility as large corrections and bulky spinal instrumentation can make closure difficult. Tissue expanders used preoperatively may be helpful in allowing better soft tissue coverage, though reported experience is limited [35, 68].

The gait and method of active transfers should be observed to determine the consequences of the loss of lumbosacral motion following extensive fusions. Hip contractures may become more problematic when the lumbosacral spine is fused. Diminished hip extension will be further compromised if the lordosis is surgically reduced and stance and ambulation will become more difficult. Increased lordosis following fusion will make sitting more difficult in those with reduced hip flexion at baseline.

A thorough neurological examination is performed. The Arnold–Chiari malformation, hydrocephalus, or spinal cord tethering may be the cause of the progressive scoliosis and their treatment may preclude the need for the surgery. If symptomatic and untreated, they may complicate

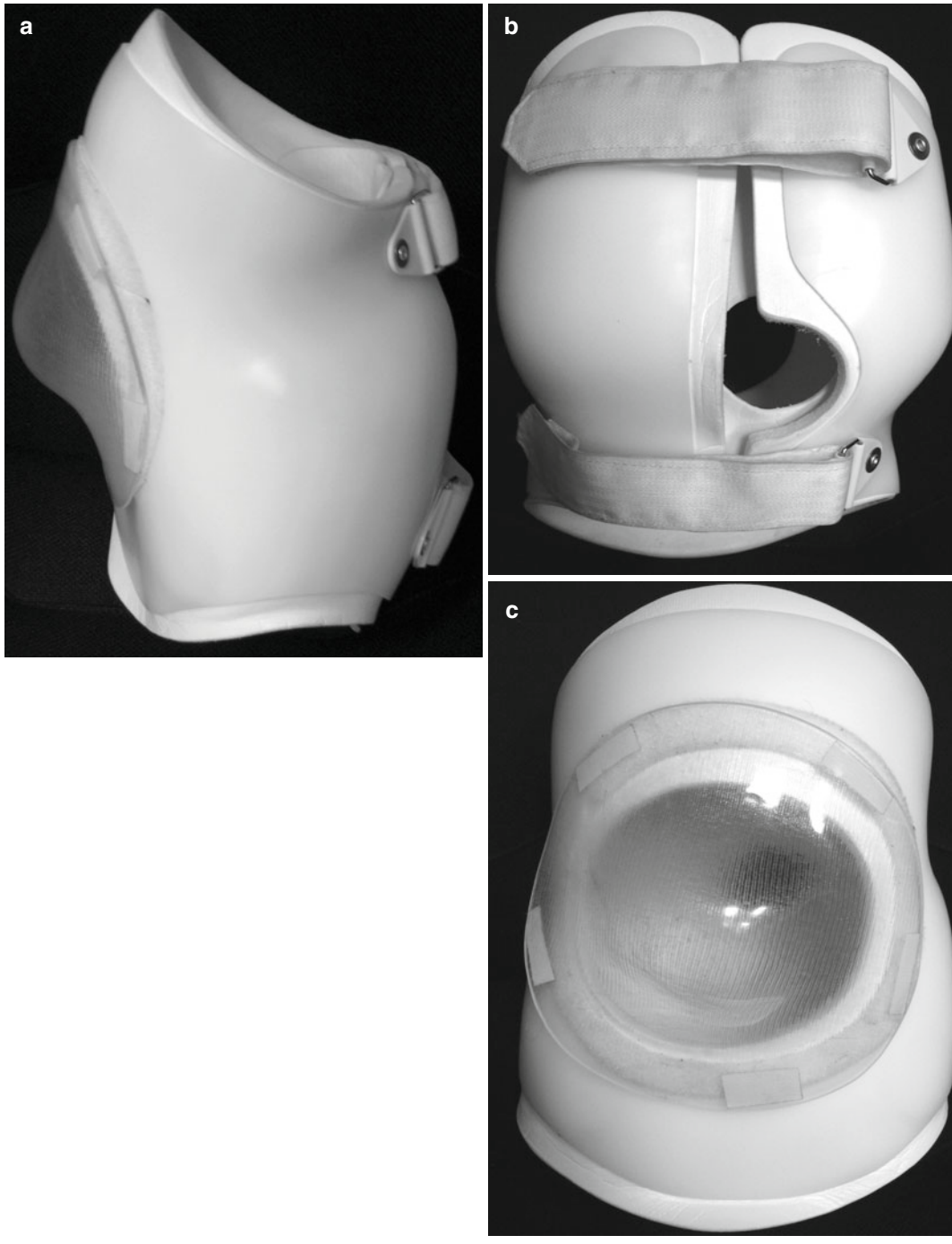

Fig. 14.2 Spinal orthoses may not prevent deformity progression, but may improve function. Here a "soft" brace has been reinforced with polypropylene and adapted to accommodate a rigid kyphotic deformity with skin ulceration (**a**–**c**). The removable clear plastic bubble over the gibbus protects the skin while allowing for its easy inspection. The orthosis is further modified to allow for drainage catheters. The device allowed this child with skin ulceration over the gibbus to maintain full function during treatment

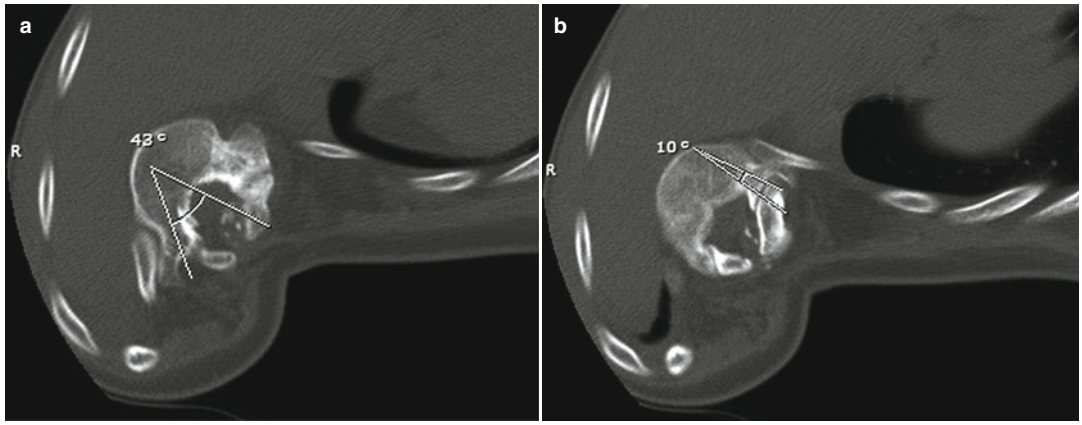

Fig. 14.3 While pedicle screws offer superior fixation in the dysplasic portion of the spine, placement can be challenging. The pedicle orientation and size may vary significantly within (**a**, **b**) and between levels. The vertebral body depicted is at the apex of the deformity where the posterior elements are essentially intact, but the angles for pedicle screw insertion and allowable screw length differ significantly

the procedure. Shunt function must be evaluated prior to cordotomy or intraoperative spinal cord manipulation to avoid catastrophic complications [30, 102]. An MRI should be performed to evaluate for syringomylia and intraspinal tumors as well as spinal cord tethering. The conus will essentially always be further distal than the norm, and this finding must be put in context with other findings to determine if there is symptomatic tethering. The role of prophylactic detethering prior to surgery is not established [79].

Laboratory assessment must include a urine culture and nutritional evaluation. Hatlen et al. have demonstrated that nutritional deficiency and preoperative positive urine cultures were related to an increased risk of infection. The organism responsible for the deep wound infection was the one found in the preoperative urine culture in 66 % of the cases [36].

The radiographic assessment should include full-length views of the spine in the position of function: standing for ambulators and sitting for non-ambulators. A sitting upright will eliminate the affect of hip contractures on the spinal alignment. This view when performed anterior/posterior as opposed to the posterior/anterior fashion affords better assessment of the lumbosacral articulation and pelvic obliquity. Flexibility is assessed with side bends, traction, or fulcrum bends as needed to determine the need for releases or other destabilizing measures and the extent of the fusion. Maturity is best evaluated by bone age as chronological age is less accurate in a population with frequent precocious puberty. A spinal CT scan is helpful to better analyze the three-dimensional anatomy. It is especially useful in determining the size and orientation of the pedicles in the dysplastic portion of the spine when pedicle fixation is planned (Fig. 14.3).

Surgery

The surgical treatment of spinal deformity in children with myelomeningocele is arguably the most challenging type of spinal surgery. The deformities are often long, severe, and rigid. There is poor soft tissue coverage and the skin is often insensate, scarred, and fragile. The posterior elements are deficient and dysplastic, offering a poor mass for fusion and difficult instrumentation purchase and placement. There may be tenuous neurological status and associated urological abnormalities.

The timing of surgery is another matter for consideration. Children that develop scoliosis will usually do so before age 10 years. The decision will be to use either growth preserving techniques or definitive surgery. One consequence of early fusion will be shortened trunk length that translates into less lung volume and perhaps lung growth. The crankshaft phenomenon has been noted in immature patients with scoliosis that have undergone posterior fusion. The continued anterior growth and posterior tether results in a rotational deformity. This deformity has not been docu-

mented in the myelomeningocele population and there is some evidence that posterior instrumentation to the pelvis in the neuromuscular population protects against it [86, 100]. The benefits of additional trunk growth must be weighed against the risk of repeated surgeries in a population notoriously at risk for operative complications.

Definitive Surgery

Advancements in technique have had a significant impact on the results of deformity surgery. Combined anterior and posterior surgery has lowered the rates of nonunion. Segmental instrumentation allows for secure fixation through the dysplastic posterior elements, dramatic corrections, and reduction or elimination of postoperative immobilization.

The choice of surgical methods must be individualized based on the patient's needs and function, the vertebral deficiency, and deformity characteristics. Attempts to spare mobile lumbar segments seem appropriate in ambulatory patients. More rigid and severe curvatures with extensive dysplastic segments will require more secure posterior fixation obtained through increased segmental anchors and, usually, the inclusion of the pelvis. The stresses on those fixation anchors can be lessened through anterior releases or posterior destabilization techniques.

Combined Anterior Posterior Instrumentation and Fusion

In the myelomeningocele population, combined anterior and posterior spinal procedures have been the standard of care. In his evidence-based literature review, Wright demonstrated that the best available series were consistent Level II and III studies. These studies clearly demonstrated the superiority of fusion rates and correction in the combined anterior and posterior techniques (Table 14.4).

While good results in terms of correction and fusion rates can be obtained with non-segmental posterior instrumentation combined with anterior surgery, the limited stability provided requires prolonged postoperative immobilization which is particularly problematic in the myelomeningocele population. Insensate skin leads to pressure sores beneath casts or braces. Contractures develop and children with limited strength may lose function by increased weakness caused through prolonged immobilization. Additionally, there are significant rates of instrumentation failure [22, 52, 82]. These problems were addressed with the development of segmental instrumentation which provided similar results and required less or no immobilization (Fig. 14.4) [4, 7, 13, 26, 49, 69, 70].

The improved fixation of pedicle fixation may preclude the need for pelvic fixation in selected patients. Wild studied a series of patients fused short of the pelvis with anterior instrumentation and posterior pedicular segmental instrumentation (Fig. 14.5). The mean curvature prior to surgery was 81.8° and 34.7° at last follow-up. The pelvic obliquity was corrected from 32° to 4.8°. There were no skin ulcerations postoperatively despite minimal correction in one patient due perhaps to seating modifications permitted by the more mobile lumbosacral area [101].

Selective Anterior- or Posterior-Alone Fusion and Instrumentation

Improved fixation permitted by newer segmental systems has lead to a renewed interest in anterior- or posterior-alone procedures with their associated diminished morbidity. Most studies reveal less correction of the deformity and pelvic obliquity with posterior only procedures though the complication rate may be less [15, 69, 93, 104].

In the report of pedicle screw fixation by Rodgers et al., 14 of 24 patients had posterior only surgery. These authors concluded that pedicle fixation produced results from posterior-alone surgery that were comparable to those obtained with combined anterior–posterior techniques (Fig. 14.6) [78].

Anterior-alone instrumentation would seem well suited to a group of patients with normal anterior vertebrae and compromised posterior midline skin and deficient posterior spinal elements, but historically this technique has been problematic due to its kyphosing tendency and the adding-on of segments above and below the end-instrumented vertebrae. Newer more rigid systems have addressed some of these concerns by providing superior rigidity and have demonstrated a lower infection rate (Fig. 14.7) [51]. Basobas et al. reported their results of anterior-alone instrumentation and fusion in 21 patients with neuromuscular scoliosis of which 12 were children with myelome-

Table 14.4 Myelomeningocle scoliosis surgical correction: a comparison of instrumentation techniques

	Author	Year	Scoliosis correction (%)	Pelvic obliquity (%)	Infection	Pseudo/ instrument failure
Post non-segmental	Osebold [67]	1982	23	29	33 %	46 %
	Mazur [52]	1986	32	39	7 %	33 %
	Parisini [69]	2002	17	50	30 %	70 %
Post non-segmantal and anterior fusion	Ward [97]	1989	57	60		
	Osebold [67]	1982	48	46	50 %	100 %
Post non-segmental and anterior instrumentation	Osebold [67]	1982	56	47	18 %	23 %
	DeWald [22]	1979	55			
	Mayfield [50]	1981	75		0	31 %
	Mazur [52]	1986	42	41	11 %	11 %
	McMaster [55]	1987	63	72		
Anterior only	Mazur [52]	1986	48	14		29 %
	Sponseller [87]	1999	57	44	7 % (superficial)	36 % revision
	Basobas [12]	2003	59	64.3	5 %	24 % revision
Post-segmental and anterior fusion	Ward [97]	1989	51	56		
	Banta [11]	1989	53		11 %	16 %
	Parsch [70]		37			33 %
	Geiger [30]	1999	52			33 %
	Yazici [104]	2000	68	96		
Post-segmental and anterior instrumentation	Ward [97]	1989	63	71		18 %
	Geiger [30]	1999	59			
	Parsch [70]	2001	57			18 %
	Wild [101]	2001	56	81	27 % (superficial)	
Post-segmental	Geiger [30]	1999	52			
	Parsch [70]	2001	41			40 %
	Rodgers [78]	1997	58	64	0	14 %
	Parisini [69]	2002	47	40	10 %	10 %
	Yazici [104]	2000	69	50		

ningocele. The mean primary curvature of 60.4° was reduced to 19.9° and was 24.6° at final follow-up, and the pelvic obliquity was corrected from 15.1° to 5.4° at final follow-up. The mean number of segments preserved below the fusion mass was 3.2. There was no loss of lumbar lordosis. There was one infection and one pseudarthrosis. Four of the twenty-one patients required subsequent extension of the fusion [12]. Stark and Saraste reported adding-on in five or six patients treated by anterior-alone methods and concluded that the technique was inadequate in this population [89]. Sponseller et al. reviewed 14 myelomeningocele patients that had anterior-alone instrumentation and fusion and found neurological deterioration in two, proximal decompensation in two, and screw pull out in one. All poor results were in patients with either syringomyelia or curvatures greater than 75° [87].

Posterior Instrumentation Techniques

The unique characteristics of myelomeningocele spinal deformity is the tenuous skin, deficient soft tissue, and the dysplastic bone as well as the complex deformities which are a combination of neuromuscular and congenital etiology. The surgical technique must address these elements. If the midline skin is scarred or tenuous, consideration should be given to a triradiate incision. This will avoid the midline skin and minimize the chance for myelomeningocele sac penetraton, while

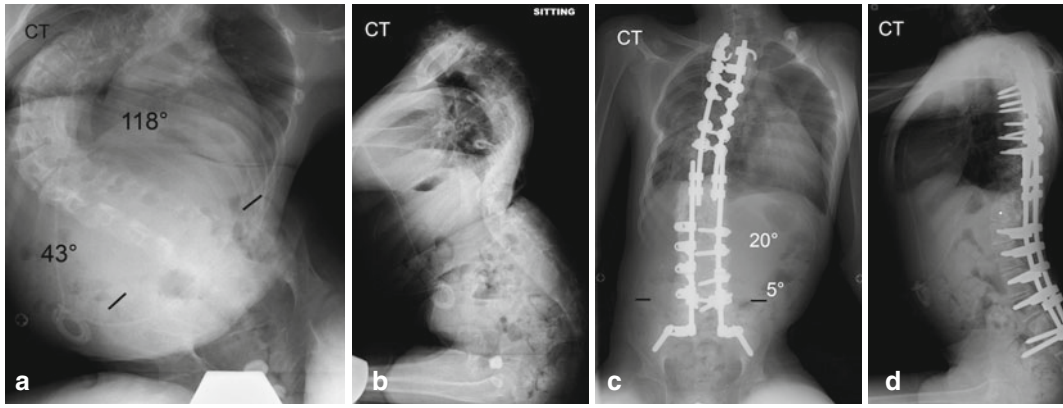

Fig. 14.4 CT has a dramatic scoliosis of 118° and a pelvic obliquity of 43° (**a**, **b**). This is a rigid deformity that corrects at best on supine traction to 90°. The scoliosis and pelvic obliquity were corrected more than 80 % through a combination of spinal destabilization procedures that included anterior releases, vertebral column resection, and rib osteotomies, and rigid segmental instrumentation provided by pedicle fixation (**c**, **d**). Four spinal rods were utilized allowing fixation through multiple anchors and maximum distribution of the forces generated by the manipulation. A cantilever manipulation then brought the two limbs of the deformity into alignment. Permitting a deformity to progress to this severity is not recommended. The example is intended to show the corrections possible with present generation techniques

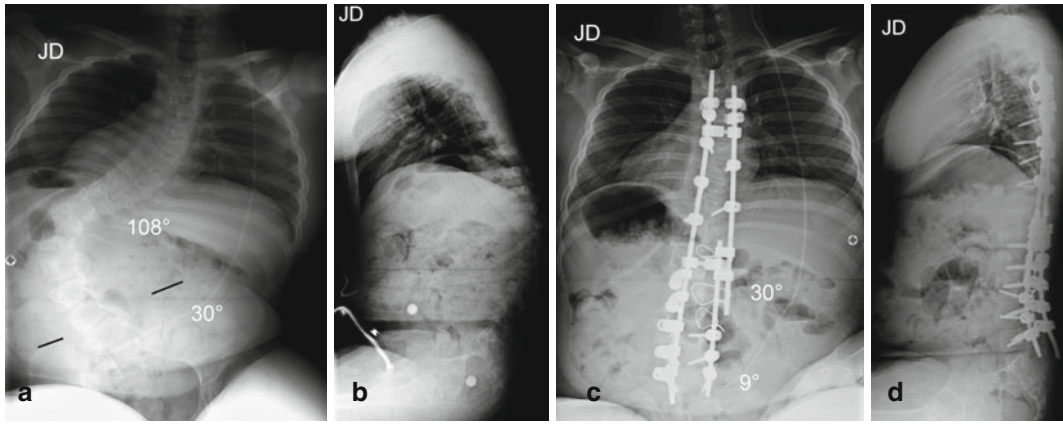

Fig. 14.5 JD has scoliosis, pelvic obliquity, and thoracolumbar kyphosis (**a**, **b**). A correction of approximately 70 % was possible with anterior releases and fusion coupled with posterior spinal segmental instrumentation (**c**, **d**). The pelvis was not felt to be part of the major curvature and the pedicle fixation in the lumbar spine provided excellent stability. Accordingly, the instrumentation and fusion did not cross the lumbo-sacrum and flexibility through that area was maintained. Although some pelvic obliquity persists, there have been no seating or skin irritation problems

affording excellent exposure of the lateral bony masses of the dysraphic spinal elements. Ward et al. noted that 6 of 15 with this incision had wound necrosis, but only 2 had a significant clinical problem. They felt that its advantages merited its use and cautioned against undermining the skin and subcutaneous tissues between the two inferior limbs and advised that the inferior limbs subtend as large an angle as possible [97]. Dissection of the dural sac is most easily accomplished by proceeding from the portion of the spine with intact posterior elements toward the deficient area.

In the portion of the spine with intact neural arches any anchor system may be used, but due to

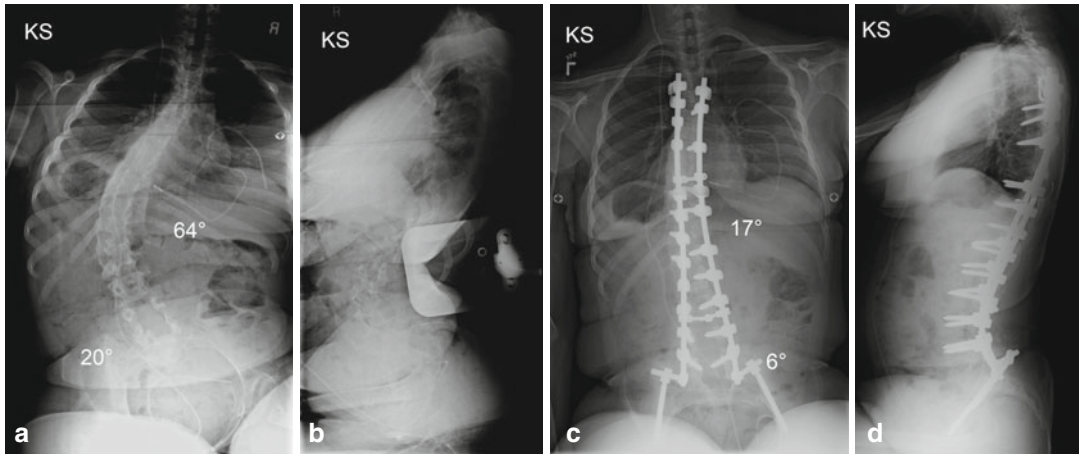

Fig. 14.6 KS has a moderate deformity with a 64° scoliosis and a 20° pelvic obliquity (**a**, **b**). Segmental fixation allowed excellent correction and stabilization without the need for an anterior procedure (**c**, **d**). The youngster was mobilized immediately following the procedure without a spinal orthosis. These children with their dysplastic posterior lumbar spinal elements do present deficient bone mass for fusion. The bulky segmental instrumentation further compromises the available bone surface. Only long-term follow-up will determine if late instrumentation failure and pseudarthroses will occur (Courtesy Dr. John Emans)

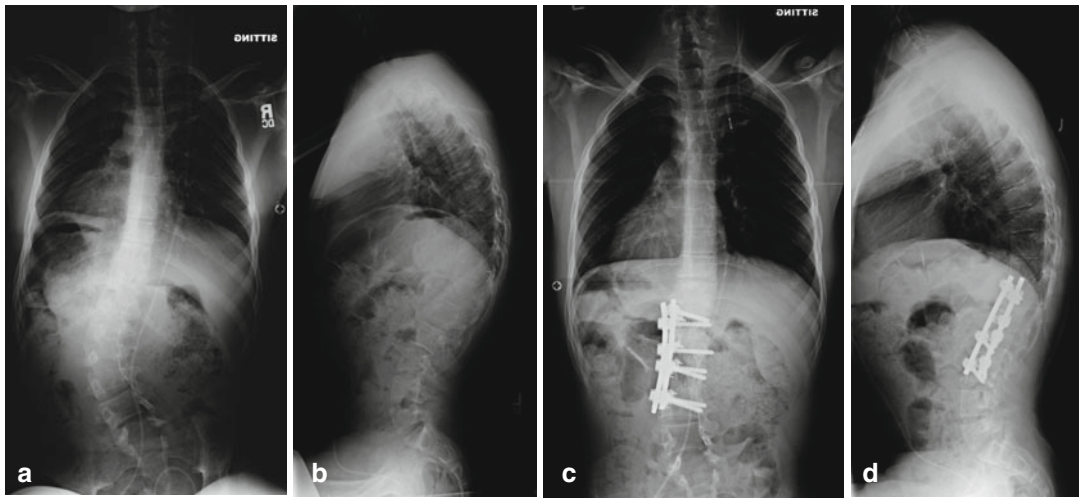

Fig. 14.7 (**a**–**d**) E.R., an avid wheelchair athlete, has not yet had his adolescent growth spurt. His lumbar scoliosis has progressed to 50°, his pelvic obliquity to 20°, and he is having early seating difficulties. Anterior instrumentation and fusion has leveled the pelvis, corrected the deformity, and spared motion segments

the osteopenic bone multiple fixation points are needed to distribute the forces. In the dysraphic portion, a number of options for segmental fixation exist. At present, pedicle screws offer superior fixation though placement is challenging as the dysraphic segments present dramatically altered pedicle size, position, and orientation. Direct visualization by dural retraction and medial wall dissection allows proper insertion. The variable intersegmental orientation of adjacent pedicles may be addressed by using polyaxial screws or adjustable transverse connectors. When pedicle anatomy precludes placement, alternatives for segmental instrumentation are Drummond wires placed with the button medially [25] and Luque wires placed through the neuroforamina beneath

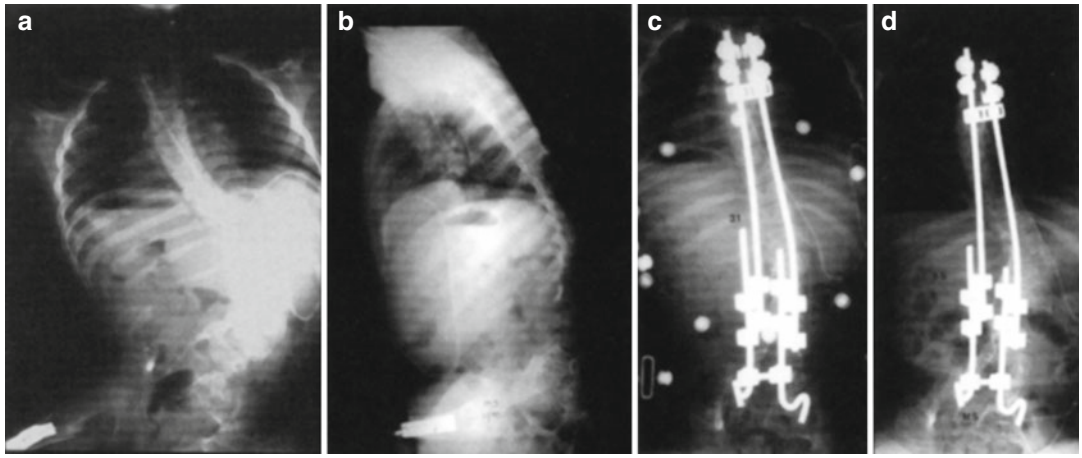

Fig. 14.8 The curvature (**a**, **b**) has been corrected by the use of spine-based dual growing rods (**c**) and growth provided by subsequent lengthening procedures (**d**) (Courtesy of Dr. Richard McCarthy)

the pedicles. Another technique involves placement of bone screws, which can be of smaller diameter, with wires looped around the screw head [91] or the use of specially designed screws with a hole in the head for wire placement (Depuy Spine). Cross-links are utilized for extra stability but must be contoured to avoid pressure on the sac.

Sacro-pelvic fixation is often required for the extra stability afforded by instrumentation anchorage and for pelvic obliquity correction, especially when the lumbo-sacral segment is part of the curvature. The Galveston method will provide good coronal plane stability and has the benefit of being low profile in an area of soft tissue deficiency [5, 7, 29, 97]. Visualization of the lateral cortex is advisable as the iliac orientation is variable and the bone stock deficient. Iliac and ilio-sacral screws provide excellent anchorage points and unlike the Galveston method may be left in place to allow for the complex rod contouring maneuvers often necessary in the dysraphic spinal area [57]. When compared to the Galveston technique, ilio-sacral screws demonstrate improved pelvic obliquity correction and less loosening [71]

Growth Preserving Instrumentation

The dilemma of early onset scoliosis is well covered in this chapter and certainly pertains to the child with myelomeningocele. At present, experience with spinal growth preserving surgical techniques in this population is limited and short term. Anticipated obstacles are obtaining and maintaining secure anchorage sites in the dysplastic posterior bony elements and avoiding wound complications secondary to incision through and instrumentation beneath scarred and tenuous soft tissues (Figs. 14.8 and 14.9). Theoretically, rib to pelvic-based constructs that avoid the midline should minimize these problems and are the most common construct used (Figs. 14.10 and 14.11).

Abol and Stuecker studied 20 non-ambulatory patients with neuromuscular scoliosis treated with a bilateral rib-to-pelvis vertical expandable prosthetic titanium rib (VEPTR). Seven of these children had myelomeningocele. Nine of the twenty had complications in 1.33 years of follow-up including proximal cradle migration in five, implant breakage in five, deep wound infection in three, and dislodged iliac hooks in two [1]. Campbell et al. reported ten patients with myelomeningocele who had thoracic insufficiency syndrome, spine deformity, and pelvic obliquity and were treated with a hybrid VEPTR construct with s-hook iliac crest fixation. The average follow-up was 5.75 years. The pelvic obliquity improved from 34° to 11°, the scoliosis from 73° to 46°, and the lumbar kyphosis from 43° to 26°. There were three s-hook migrations, two rib-cradle migrations, one skin slough, and four wound infections

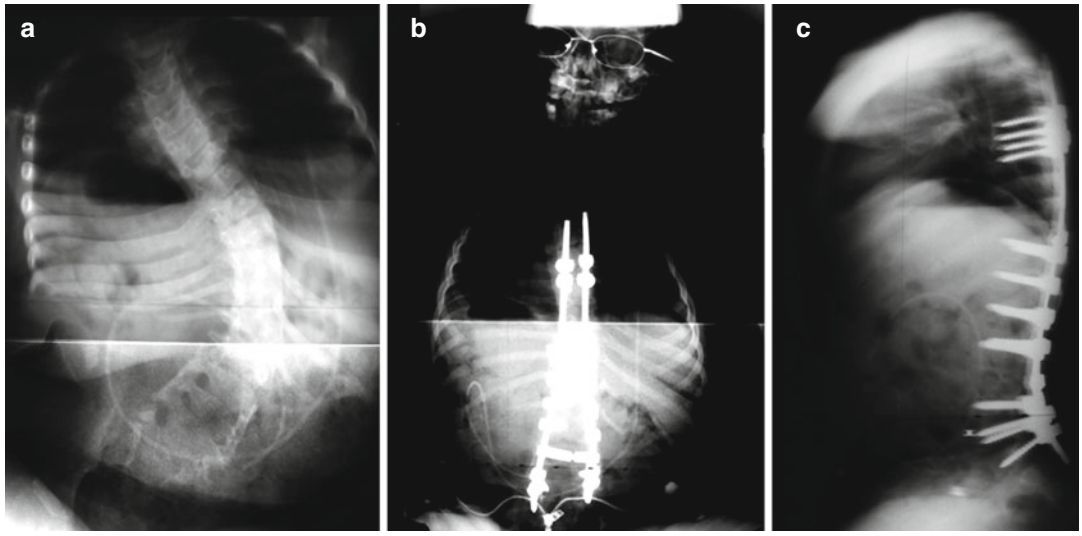

Fig. 14.9 The Shilla procedure allows continued growth without the need for repeated lengthenings. The apex of the deformity (**a**) is corrected and fused and the proximal and distal limbs of the deformity grow under the "guidance" of the specifically modified anchors (**b**, **c**)

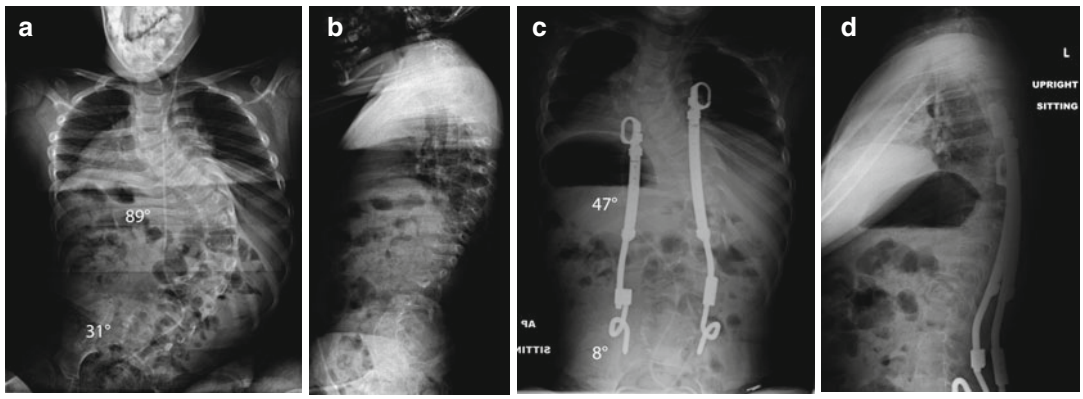

Fig. 14.10 (**a**–**d**) BD has a significant deformity at age 7. A growth preserving technique was utilized to improve seating and avoid spinal fusion at an early age. The VEPTR technique was selected in part because it could be performed away from the midline scarred tissues. To date there have been no major complications. If the correction can be maintained, definitive surgery may be less involved than it would have been had the curvature been allowed to progress

[18]. Less proximal instrumentation complications in a small series of four myelomeningocele patients have been reported with a four-rib hybrid construct [2]. Smith et al. reported on the use of spinal- and rib-based distraction systems for early onset scoliosis in a myelomeningocele population. While deformity stabilization was obtained, there were 66 complications in 34 patients. This included 24 infections and device migration or rib fractures in 15. The authors emphasized that most of these problems were easily managed [85]. Clearly, the decision to embark upon this form of treatment requires careful comparison of the benefits of growth preservation and perhaps the avoidance of more extensive future surgery versus a single definitive procedure.

14.3.3.4 Surgical Complications

Enthusiasm for correction of spinal deformity must be tempered by the considerable problems encoun-

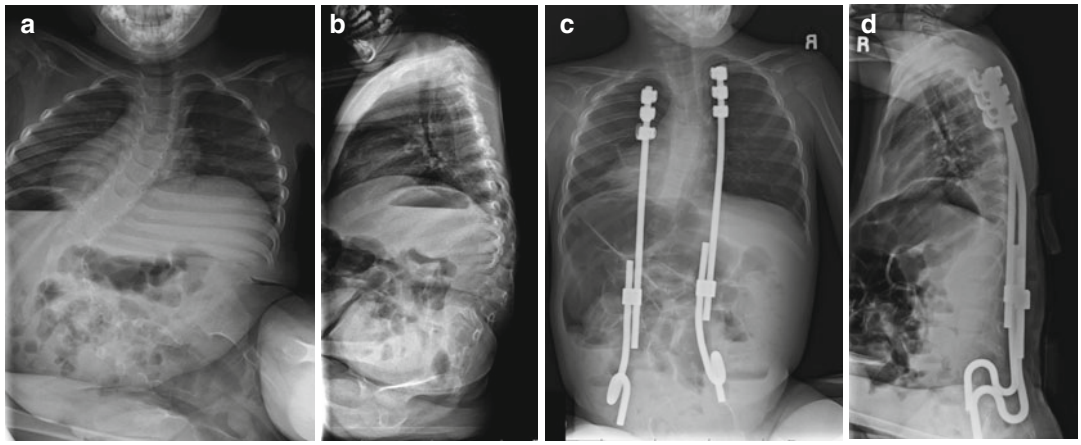

Fig. 14.11 (**a**–**d**) PL presented at 15 months of age with a 90° scoliosis and 40° pelvic obliquity. He underwent spinal cord detethering, but as anticipated with a curvature of that magnitude, there was continued progression and loss of seating balance. The spinal radiographs at 3 years 4 months are shown. A hybrid pelvis to rib construct avoided surgery on the scarred midline tissues and resulted in significantly improved alignment. The child now sits unaided and uses a standing frame

tered. Surgery for myelomeningocele spinal deformity has rates of complications, including infection, pseudarthrosis, instrumentation failure, and neurological deterioration, that are among the highest of all deformity surgery [7, 47, 50, 52, 55, 62, 82, 83, 97]. In defense of surgery, it must be realized that some of these studies relate to methods no longer used. Improved spinal instrumentation, earlier mobilization, attention tourosepsis, nutritional support, neurosurgical evaluation, prophylactic antibiotic treatment, and proper tissue handling have combined to decrease the complications. Nevertheless, this particular population continues to be a difficult one to treat despite the use of modern surgical techniques. Even when compared to the neuromuscular population, a group known to have high complication rates, the myelomeningocele patients stand out as the most problematic. Stevens and Beard reported on 76 patients with neuromuscular spinal deformity undergoing segmental spinal instrumentation. Most of the serious complications occurred in 30 patients with myelomeningocele. Ninety percent of these had a complication and half of the unplanned re-operations were in this group (Table 14.5) [91]. In a similar series, Benson et al. noted that in a study group of 50, 13 patients with myelodysplasia had over half the complications [13].

Table 14.5 Segmental spinal instrumentation for neuromuscular spinal deformity—complications by diagnosis

Complication	Myelodysplasia (30 patients)	Other (46 patients)
Deep infection	10	1
Skin breakdown	6	1
Neurological deficit	6	4
Permanent	4	4
Temporary	2	0
Inadequate correction	3	6
Minor revision rod/wire	2	8
Broken rod	1	4
Mechanical failure	1	4
Curve progression	0	3
Pseudarthrosis	0	3

Adapted from Stevens and Beard [91]. With permission from Wolters Kluwer Health

In this study, a notable preponderance of serious complications occurred in the myelomeningocele subgroup. Ninety percent of these patients had one or more complications. Of 27 patients that had an unplanned procedure, half of them were in the myelomeningocele group

Outcomes

Reasonably good technical results have been documented following spinal surgery, but there are few studies that specifically address func-

tional outcomes. Kahanovitz and Duncan evaluated 39 patients over age 16 years with myelomeningocele and scoliosis. Improved function correlated with curvatures less than 40° and pelvic obliquity less than 25°. Of the 15 patients that had spinal surgery 8 maintained, 7 lost, and none improved function. Twelve of these surgeries were in situ fusions and the others employed first-generation instrumentation techniques. The conclusions were contradictory; surgery was not helpful, but deformities of greater than 40° should be avoided. The intimation is that improved surgical techniques would improve the future outcomes [41]. In a study of 98 children with spina bifida and scoliosis, Wei et al. used statistical analysis to evaluate the relationship of spinal deformity to sitting balance and multiple functional assessment measures. The authors concluded that in the short run the only benefit of surgery was improved seating balance [96].

A number of studies that address surgical techniques have also included functional outcomes. Separate series by Mazur, Schoenmaker, and Muller report that a minimum of 50 % of patients lost ambulatory ability following surgery [52, 62, 82]. Only rarely was this deterioration based on neurological status changes. Of note is that these patients had prolonged postoperative immobilization, developed contractures, and presumably were deconditioned. Many of these children were at best borderline community ambulators and loss of ambulatory function may be the natural history as these children age. Other studies provide a more optimistic outlook with improved self-esteem [3], self-care [52, 80, 81], and modestly improved pulmonary function [11, 19].

Conclusion

The complex pathophysiology of myelomeningocele is now much better understood. It is appreciated that several different neurological abnormalities are present and that proper intervention can often prevent neurological deterioration. Technical advances in the surgical treatment of scoliotic deformity in children with myelomeningocele allow both improved deformity correction and early mobilization and may lead to better outcomes. Spinal growth sparing techniques offer a promising alternative treatment for severe early onset curvatures. Treatment should be tailored to the individual's needs keeping in mind the very real difficulties involved in this population with challenging and unique problems.

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Intraspinal Pathology

15

Nejat Akalan, Luke Macyszyn, Steven Hwang, and Amer F. Samdani

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Key Points

- The term "spinal dysraphism" covers two types of spinal congenital malformations, traditionally grouped as "open" and "closed" forms.
- Open spinal dysraphism, or myelomeningocele, is primarily a neural tube closure defect with treatment aimed at preserving the neurological and clinical status of the newborn.
- Chiari decompression improves syringomyelia in up to 90 % of patients, which increases safety of deformity surgery.
- Type 1 diastatomyelia should be resected prior to deformity surgery, whereas Type 2 diastatomyelia may be left alone.

15.1 Introduction

Congenital spinal disorders that interfere with neurological function result from imperfect development of either the neural tissue itself or tissues that are designed to cover and support the spinal cord. In current terminology, the terms "spina bifida" and "spinal dysraphism" are used interchangeably to cover all spinal malformations derived from neuroectodermal and mesodermal

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origin. Those that are believed to be of neuroectodermal maldevelopment form the "open dysraphism" subgroup: "open = Appert" implying a visible, exposed lesion of the neural tissue. This group is also called "neural tube defects" along with "anencephaly," which results from failure of fusion of the cranial neural tube. Mesodermal tissue-derived embryological anomalies that directly or indirectly hamper normal neurological function form the "closed dysraphism" group with the assumption of the maldevelopment occurring over a normal neuroectodermal differentiation period. The term "closed = occult" describes an obscured lesion covered with intact skin unlike the open counterpart (Fig. 15.1). Although such a simplified classification scheme of spinal dysraphism, according to the embryological origin, may help in understanding and standardizing the diagnostic and therapeutic measures, the diverse clinical manifestations, natural course, and treatment protocols do not necessarily provide prescribed algorithms. Moreover, normal development of the spinal cord and its surroundings is far more complicated and intermingled rather than a consecutive differentiation of neuroectoderm and mesoderm. The insult at a certain point of differentiation may contribute to both neural and adjacent tissue maldevelopment resulting with open and closed types of dysraphism exhibited in the same patient.

The aim of this chapter is to discuss the contemporary treatment protocols of this heterogeneous group with emphasis on changing concepts due to accumulating data on their embryogenesis, natural course, and therapeutic alternatives.

15.1.1 Open Spinal Dysraphism (Myelomeningocele)

Formation of the spinal cord is identified as primary neurulation in classical embryology, wherein ectoderm of the bilaminar embryo undergoes a series of complex differentiation,

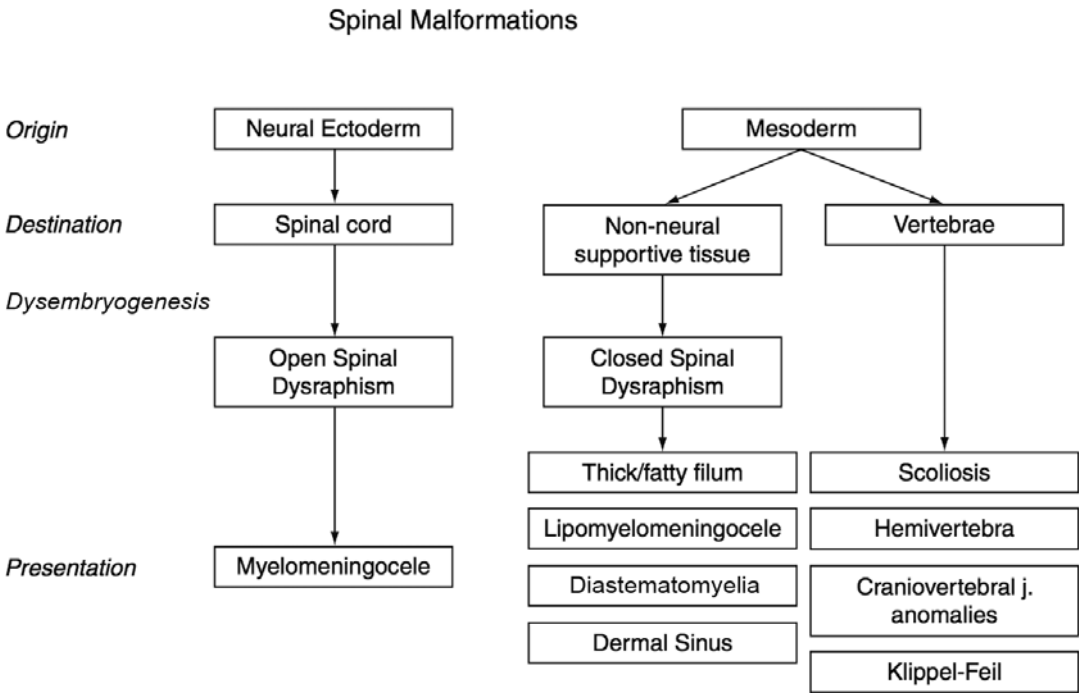

Fig. 15.1 Congenital spinal malformations; disordered embryogenesis during formation of neural tube from neural ectoderm results with myelomeningocele, while different forms of occult dysraphism may occur due to mesodermal disarrangement

yet not through a fully understood mechanism, to form the neural tube. This process requires molecular, biochemical, and mechanical interactions between neuroectodermal cells and adjacent ectodermal derivatives [1–4]. Although primary neurulation itself represents neural tissue formation, it is closely dependent on the ongoing mesodermal activity that is responsible for the differentiation of the nonneuronal surrounding tissue. While notochordal induction is essential for initiating primary neurulation, a completed neurulation is required for appropriate differentiation of the surrounding tissue (also see Chap. 14).

15.1.1.1 Pathogenesis

Neural tube formation starts at the 17th day of gestation during which the notochord induces the overlying ectoderm to differentiate into neuroectoderm to form the neural groove in the dorsal midline of the embryo. Neural folds on either side of the groove elevate and meet, the cells fuse dorsally, and a primitive spinal cord is formed as a hollow cylinder. This process is completed by days 27–28 of gestation. Interruption of the neural tube formation dorsally at a particular segment(s) prevents normal neural tissue differentiation at that level. While this maldevelopment of the spinal cord structure causes a more or less complete neurological impairment below the affected level, additional abnormalities appear as a result of altered induction of the neural tube to the surrounding tissues [2, 5]. The consequence is a visible spinal cord segment, placode, representing an unclosed primitive neural tube remnant without meningeal, bony, or cutaneous enclosures dorsally. The term "spina bifida," although denoting only the missing posterior bony elements over the unclosed neural tissue, is confusingly used to describe the whole anomaly. Likewise, myeloschisis, spina bifida cystica, and myelomeningocele are terms attributed to the different morphological appearance of the same pathology contributing no practical purpose in terms of decision making, surgical technique, or outcome other than confusion. The term "myelomeningocele" is currently preferred to represent almost all variations of open spinal dysraphism that result from a common mechanism of maldevelopment [1, 6, 7].

Disordered embryogenesis of the segmental neural tube formation in myelomeningocele is attributed to either primary failure of neural tube closure or secondary opening after appropriate tube formation. The nonclosure theory is probably for the majority of human myelomeningoceles; however, overdistention may contribute to some experimental neural tube defect models [2, 6]. Regardless of the causative mechanism, unclosed neural tube triggers a cascade of events concerning the nonneural tissue that is designated to cover the spinal cord dorsally. The cutaneous ectoderm remains attached to the open neural tube segment and fails to form the future skin over the lesion. Moreover, the normal mechanism in which the cutaneous ectoderm detaches from the dorsal side of the neural tube to allow paraxial mesoderm to move in between to give rise to bone and soft tissue is also distorted. The final pathological anatomy is an exposed lesion at birth, representing the inner surface of the spinal cord on the dorsal midline covered with membranous tissue as the epidermal remnants or exudate, with a groove continuous with the central canal of the unaffected segments. Immature laminar remnants and paraspinal muscles occupy the lateral border of the widened spinal canal and can be palpated under the border of the skin defect (Fig. 15.2).

15.1.1.2 Etiology and Epidemiology

Current hypotheses suggest that complex interactions between extrinsic and intrinsic variables are responsible for myelomeningoceles. Clinical and epidemiological data in humans imply maternal illnesses, medications, environmental toxins, and dietary factors such as lack of folic acid that play causative or at least contributing roles in the development of myelomeningocele. On the other hand, increased incidence demonstrated in certain families situates neural tube defects into complex genetic disorders in which genes and the environment interact through an unknown relationship [2–4].

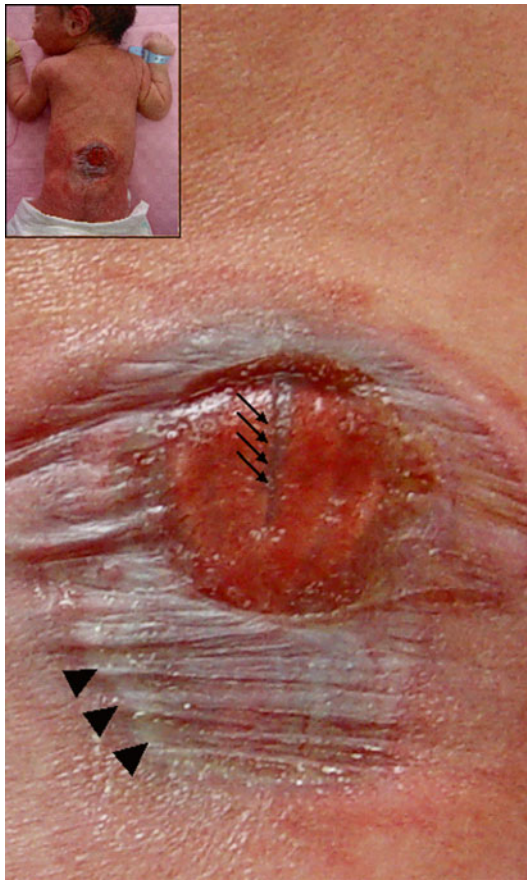

Fig. 15.2 A newborn with myelomeningocele (*upper left*). Unclosed primitive neural tube remnant, placode, is exposed without any dorsal covering expect for membranes of epidermal remnants or exudate. The groove (*arrows*) represents the central canal. The healthy skin border (*arrow heads*) denotes the lateral edges of the widened spinal canal segment

The incidence of myelomeningocele is generally accepted as 1/1000 live births regardless the ethnic and geographic variability. Although several studies demonstrate a variation in different parts of the world depending on the geographical region, seasons at conception, gender of the affected infants, ethnicity, and socioeconomic status of the parents, maternal age and parity and population-based surveillance studies fail to confirm a definitive correlation with the incidence [8]. A decrease in frequency of myelomeningocele has been reported recently in some areas, while the incidence has been stable elsewhere [9]. Although this decrease has been attributed to increased prenatal diagnosis, selective terminations, genetic counseling, and mostly folic acid supplementation during pregnancy, there are no hard data to indicate that the decrease is due to a single factor [10, 11].

15.1.1.3 Presentation

Myelomeningocele represents one of the most devastating congenital malformations that are compatible with life. This is due to the fact that neurological impairment is inevitable for a patient with myelomeningocele and the severity is proportional to the affected level of the spinal cord. With up to 80 % of the disclosure taking place at the thoracolumbar spine, paraplegia is the result. The level of neurological deficit descends as the lesion level moves caudally; at best, sacral localization avoids a major motor disturbance but does result in a neurogenic bladder [7, 12, 13].

The neurological deficit in myelomeningocele is thought to be due not only to incomplete differentiation of the neural tube but also to exposure of the uncovered neural tissue to amniotic fluid. Furthermore, associated anomalies extending to 63 % as reported in fetal autopsy series contribute to the disability of the myelomeningocele cases [3]. Besides morphological abnormalities of the adjacent vertebral elements, almost all patients with myelomeningocele have associated Chiari II hindbrain malformation. The simplest representation of Chiari II malformation is herniation of the cerebellar tonsils and vermis to the cervical spinal canal through a tight foramen magnum. Additionally, medullary kinking, low-lying tentorium, tectal beaking, brain stem nuclei changes, polymicrogyria, and gray matter heterotopias may be associated with the Chiari malformation. The main contribution of the tonsil herniation to the clinical picture of myelomeningocele is hydrocephalus, which is present in almost 90 % of patients either during delivery or becoming apparent after surgical treatment [1, 12]. Hydrocephalus are one of the major coexisting factors that are responsible for morbidity and overall unfavorable outcome in myelomeningocele cases. Craniolacunae, a mesodermal self-limiting skull abnormality, is also a frequent finding in the newborn.

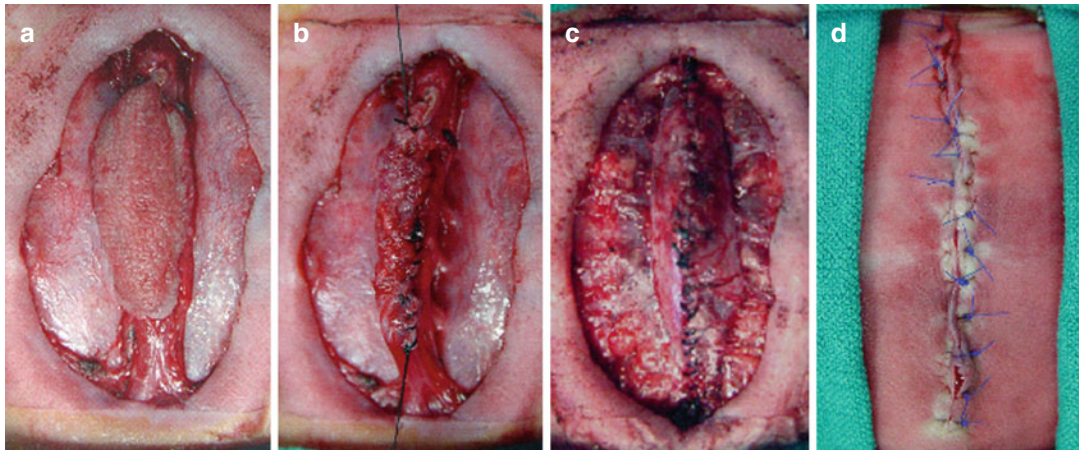

Fig. 15.3 Basic steps of myelomeningocele closure. (**a**) Removing membranes and debris and exposure of the placode, (**b**) reapproximation of the flat placode to a tubular form with fine sutures for an easy closure, (**c**) detaching the dural layer from paravertebral fascia and watertight closure over the exposed neural tube, and (**d**) primary closure of the skin defect

From the practical viewpoint, a patient with myelomeningocele is born with signs of functional cord transection at the lesion level and neurogenic bladder, and has a very good chance of hydrocephalus. The open lesion carries a substantial risk of getting infected; cerebrospinal fluid exposure to the external environment through the incomplete dural barrier can initiate meningitis and ventriculitis. Meningitis not only complicates a probable treatment for hydrocephalus but also adds the potential risk of seizures and further neurological impairment in terms of intellectual outcome.

15.1.1.4 Surgical Treatment

The aim of surgical treatment for myelomeningocele is to stabilize the clinical and neurological status of the newborn and prevent the potential risks of deterioration. This is best achieved by reconstructing the open neural tube and its coverings as soon as possible after birth. The initial management aims to stabilize the infant, avoiding contamination of the lesion and excluding the associated malformations. First, the important concern at this point is the decision to treat. This has medical, ethical, and legal ramifications to be discussed among the parents and the physicians. In cases with a prenatal diagnosis, the parents have already acknowledged the consequences of a congenital anomaly and treatment options. Otherwise, the anticipated problems in myelomeningocele with the limited role of surgical repair in the final outcome may result in the refusal of surgical treatment. From the physicians' side, severe forms with multisegment large lesions complicated with associated vertebral anomalies and hydrocephalus may cause hesitation to treat. Unless there is a life-threatening coexisting malformation, the current ethicolegal opinion is to provide surgical treatment to all cases [6, 7, 12]. Once treatment has been decided, the second concern is the timing of the surgical procedure. Risks of immediate repair in a newborn should be weighed against the risk of contamination at delayed closure. Surgical repair within 48–72 h after birth is universally accepted and does not necessarily carry increased risk of contamination compared to very early treatment (within the first 24 h). Furthermore, the time interval provides sufficient postnatal evaluation and stabilization of the infant. The simplest description of the surgical technique is to mimic the normal embryological pattern of development. This is to isolate the nonfused segment, establish the original tube shape for the placode, and recreate and close the dural envelope followed by approximation and closure of the skin over the lesion (Fig. 15.3a–d). Even very large defects can be closed by relaxation incisions along the axis, avoiding complex muscle and skin

flaps once advocated but proved to have major consequences. Almost 10 % of cases exhibit hydrocephalus at birth, necessitating simultaneously shunting with repair of the spinal defect. Operative mortality is nearly zero, but major morbidity includes progressive hydrocephalus, wound infection, breakdown, and leakage of cerebrospinal fluid.

Following myelomeningocele repair, the treatment of the other conditions may range from simple observation to extensive surgical procedures. The vast majority of patients will require shunts for hydrocephalus before being discharged; future treatment might be required for associated kyphosis, Chiari malformation, foot deformities, and secondary tethering of the spinal cord for CM-II, syringomyelia, and/or tethered cord syndrome. While myelomeningocele may be regarded a static and nonprogressive defect, clinical worsening is caused by associated problems. Owing to the fact that myelomeningocele is located at thoracolumbar segments in almost 80 % of the cases, the child faces lifetime complications of paraplegia and neurogenic bladder. Therefore, the initial closure is just the beginning, and outcome and long-term results greatly depend on the management of the associated conditions [7, 12]. At least 75 % of children born with an open spina bifida can be expected to reach their early adult years. Survivors have a high incidence of problems related to pressure sores, obesity, severe renal disease, hypertension, depression, and visual impairment. Mortality is mostly related to shunt dysfunction and infection, urinary complications of neurogenic bladder, or respiratory tract infection [13].

Fetal Myelomeningocele Repair

Fetal surgery is routinely performed for various conditions, and myelomeningocele is also a good candidate for in-utero repair since it is associated with considerable morbidity after postnatal care, it is compatible with life, and can be detected before the 20th week of gestation. Furthermore, there is enough experimental evidence that some function of the placode is preserved initially but can deteriorate during gestation or at birth. Fetal closure of the open neural tube can prevent secondary damage and preserve neurological function, while existing fetal potential for wound healing and axonal regeneration might reverse preexisting injuries to a certain degree. Since 1994, more than 330 cases of intrauterine repair have been performed by standard multilayer reconstruction through a hysterotomy between 19 and 25 weeks gestation in certain centers worldwide. Preliminary findings suggested that intrauterine myelomeningocele repair may lessen the degree of Chiari malformation and reduce the incidence of shunt-dependent hydrocephalus [14]. Therefore, a multicenter randomized controlled trial was initiated in the United States to compare intrauterine with conventional postnatal care in order to establish the procedure-related benefits and risks. The Management of Myelomeningocele Study (MOMS) was terminated early in 2010 due to the efficacy of prenatal surgery. The researchers found that intrauterine repair was associated with a 52 % risk reduction with regards to shunt placement and at 30 months the patients in the prenatal group had better motor function and were more likely to walk without an orthotic device [15]. These findings are especially important since the anatomical lesion in the prenatal group was significantly higher. However, the benefits of prenatal repair of myelomeningocele must be weighed against a higher rate of prematurity and maternal morbidity.

Type 1 Chiari malformation is the most common neural axis abnormality in infantile and juvenile idiopathic scoliosis [16]. Furthermore, between 50 and 75 % of these patients will have concomitant syringomyelia, and anywhere from 40 to 80 % will have scoliosis. The incidence of syringomyelia-associated scoliosis falls to less than 20 % for patients over the age of 20 [17]. As compared to adolescent idiopathic scoliosis, this patient population tends to have more atypical curve patterns with a higher curve apex, but a right thoracic remains the predominant curve [17, 18]. The severity of the spinal deformity, however, has not been shown to be correlated with the length of the syringomyelia cavity.

It remains prudent to first perform a Chiari decompression, with or without duraplasty, since this has been associated with a decrease in the size of the syrinx and possible improvement or stabilization of the spinal curvature. Suboccipital decompression is preferred over shunting, as the benefits on curve progression have not been observed when shunting of the syrinx is performed alone [19]. Likewise, the possible benefits of neurosurgical intervention do not extend to patients with congenital scoliosis and syringomyelia.

In several studies of children under the age of 10, over 90 % experienced an improvement or stabilization of their spinal curve following Chiari decompression [16, 17]. Although others have reported more modest results following decompression, averaging about 50 %, there appears to be a consensus in the literature that patients who progress tend to be older and have larger, double curves [17, 19, 20]. It is important to note that more extensive decompression of the cervical spine, laminectomies below C1, and extensive muscle stripping have been reported to worsen the spinal deformity, especially in the sagittal plane [18]. Finally, the effects of decompression may be temporary; hence, a follow-up period of at least 5 years is advised [19].

15.1.2 Closed (Occult) Spinal Dysraphism

Occult spinal dysraphisms represent a wide spectrum of malformations within congenital spinal disease, the only common feature being a mesodermal developmental error covered with normal skin [21]. Unlike the neuroectoderm that is designated to initiate the spinal cord, embryonic mesoderm gives rise to a variety of structures. Any disarrangement during the differentiation of this pluripotent layer triggers diverse forms of disease with regard to anatomy, clinical presentation, and treatment options. While the neurological impairment in open dysraphism is straightforward related to the incomplete differentiation of the neural tissue, the mechanism of neurological consequences in closed forms are far more complex and controversial. This complexity, in turn, generates an ongoing controversy in establishing universal algorithms for treatment.

15.1.2.1 Pathogenesis

Neurulation is responsible for the formation of the spinal cord until the future second sacral segment and the most distal segment of the spinal cord develop by a process called secondary neurulation from the neural ectoderm cell mass caudal to the neural tube, the caudal eminence. Caudal eminence is formed from pluripotent cells derived from the regressing primitive streak. The mesenchymal neural cord then becomes an epithelial cord, acquires a lumen by canalization and regression process, attaches to the primary neural tube, and forms the remaining sacral and coccygeal segments of the spinal cord including the filum terminale [1, 3, 22]. Developmental errors during secondary neurulation, besides several anomalies, lead to the formation of a fatty and short/thick filum, a classical representative of occult spinal dysraphism. Other major occult forms of dysraphism include split cord malformations (SCMs, diastematomyelia), lipomyelomeningoceles, and dermal sinuses which represent disordered mesodermal differentiation belonging to different stages during primary neurulation, before secondary neurulation begins [23]. The setoff time for the mesodermal maldevelopment and the stage of neurulation at that instance are critical for the neurological consequences. The more the primary neurulation is disrupted, the chances are higher that the child is born with a neurological compromise. This is one of the main reasons why, in different forms of occult dysraphism, resultant neurological status ranges from normal to severe impairment, sometimes compatible with myelomeningocele. Segmental, asymmetrical involvement of neurulation results with lower extremity changes, including leg or buttock asymmetry, hip and knee problems, and foot deformities that typically worsen due to muscle imbalance, weight bearing, and gravity as the child grows [6].

15.1.2.2 Diastematomyelia (Split Cord Malformation, or SCM)

SCMs represent a mesodermal anomaly belonging to the earliest stages of embryogenesis. The terms diastematomyelia and diplomyelia refer to a segment formed in two separate hemicords either in individual dural sleeves separated by a bony-cartilaginous septum in between (SCM type I) or hemicords separated by fibrous septa in a single dural sac (SCM type II). Pang et al. [24] introduced this new nomenclature and a new theory for the formation of these anomalies. An adhesion between the ectoderm and endoderm leads to an endomesenchymal tract that divides the spinal cord. In the earlier weeks of gestation, the primitive neurenteric canal temporarily connects the yolk sac of endodermal origin with the amnion, which is ectodermal in origin. While the primitive neurenteric canal regresses, a second endodermal–ectodermal communication, the accessory neurenteric canal appears. The persistence of the anterior end of the accessory neurenteric canal causes intestinal duplication, the formation of a fibrous band that interferes with intestinal rotation, or the development of a neurenteric cyst and the persistence of the posterior end results in cutaneous abnormalities such as angiomas, umbilical lesions, and hypertrichosis. The notochord is forced to develop in two separate pieces by the persistence of the intermediate part at that level. The neural ectoderm over the separated notochord is forced to form two separate neural tubes in return. The duplication of the notochord further initiates abnormal vertebral body formation, like hemivertebrae, bifid, hypertrophic or hyperplastic vertebrae, fusion of adjacent vertebral bodies comprising associated congenital spinal deformity. In this context, a hairy patch marking the level of the malformation mostly at the thoracolumbar area and scoliosis occasionally with lower extremity changes are the hallmarks of classic SCM.

15.1.2.3 Spinal Lipoma (Lipomyelomeningocele)

Often used as a general term for all lumbosacral lipomas, lipomyelomeningocele refers to a malformation in which a subcutaneous mass of fat extends through a deficient dorsolumbar fascia and lamina to attach to an open neural placode similar to a myelomeningocele. In this most frequently encountered form of occult dysraphism, the lipoma often tethers the cord asymmetrically, leading to rotation of the cord and the unequal development of nerve roots. Among various speculative theories, a current theory that accounts for the surgical anatomy is that of McLone and La Marca [25] and Naidich et al. [26]. It is proposed that the separation of the neural tube from the surrounding ectoderm, disjunction, occurs prematurely, leaving the neural plate open posteriorly and allowing mesenchymal cells to enter this cleft, where they are induced by the primitive ependyma to form fatty tissue, while the remaining anterior half of the neural tube induces the development of normal meningeal and vascular structures. The resulting anatomy is a skin-covered lumbosacral mass continuous through a defective bone and muscular tissue to adhere a partially open spinal cord segment (Fig. 15.4a–c). The subsequent neurological picture at birth varies from normal to asymmetrical lower extremity involvement with neurogenic bladder, similar to that seen in SCM.

Thick/Fatty Filum

Thick/fatty filum represents a true defective secondary neurulation process where normally the caudal mass, upon completion of the neural tube—primary neurulation—undergoes a canalization and retrogressive differentiation process to form the spinal cord below the lumbar enlargement including the filum terminale. Development of thick/fatty filum is a poorly understood process. Current theories on the maldevelopment of filum terminale with lipomatous lesions center on faulty retrogressive differentiation, with differentiation of pluripotent caudal mass cells into adipocytes. Such a theory is consistent with the observations that these lesions are less frequently associated with cutaneous stigmata, as secondary neurulation occurs after the closure of the overlying ectoderm, and that they often occur in conjunction with other malformations of the caudal cell mass, such as sacral agenesis and VATER

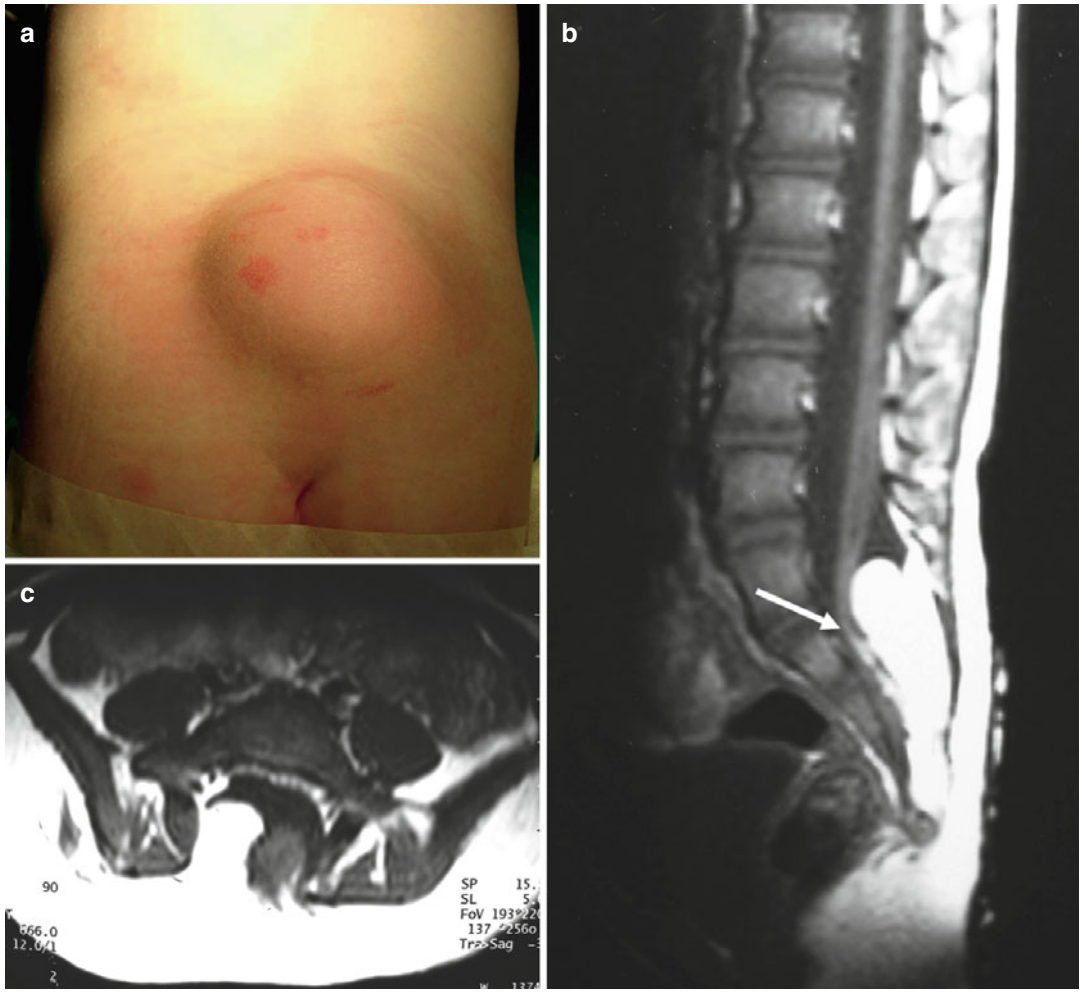

Fig. 15.4 (**a**) Lipomyelomeningocele in a 2-year-old girl. (**b**) Sagittal T1-weighed image reveals a fatty mass attached to conus where conus (*arrow*) lies at almost S1 level instead of L1–L2. (**c**) Axial view demonstrates intimate fat–conus interface with a marked torsion of conus and the roots due to lipoma

syndrome. The consistent finding is a low-lying conus ending well below the level of L1–L2 vertebral segments, attached to a thickened, short, and fatty filum terminale (Fig. 15.5). This is also reflected in the clinical presentation in almost all cases present with exclusively neurological signs and symptoms of conus involvement apparently at late childhood, without any orthopedic or vertebral deformity [21, 27, 28]. Fatty filum is also referred to as "filum terminale lipoma" and is included as a subgroup of spinal lipomas along with lipomyelomeningoceles and are believed to be of common embryological origin.

Spinal Dermal Sinus

Spinal dermal sinuses are believed to occur due to incomplete disjunction of the cutaneous ectoderm from the underlying neural ectoderm following the dorsal closure of the neural tube. The disjunction process involves detachment of the cutaneous ectoderm from the neural tube to enable the paraxial mesodermal tissue to slide in between to give rise to bone and soft tissue to form the dorsal aspect of the future spinal canal. If the ectoderm fails to detach at a given point, most often the future lumbar area, the resulting malformation is a

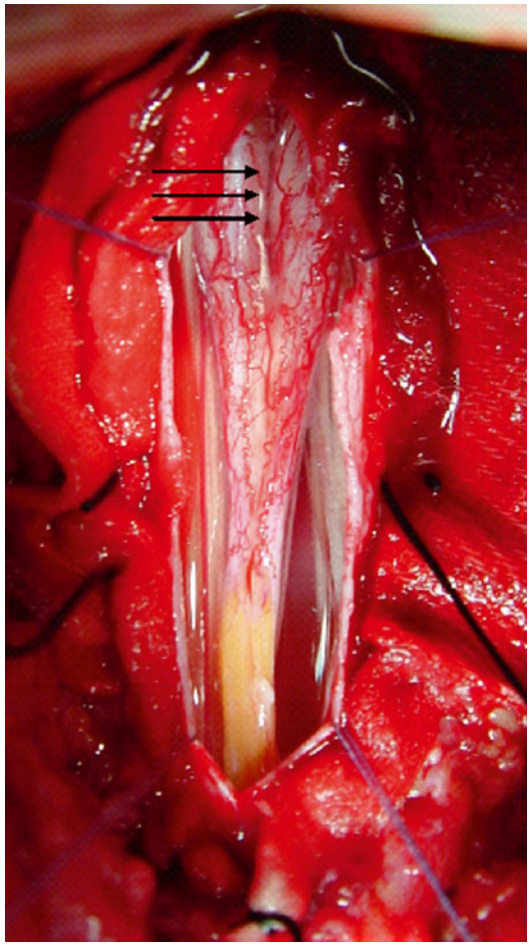

Fig. 15.5 Thick/fatty filum with an additional type II split cord malformation where two hemicords separated by fibrous bands lying within a single dura (*arrows*)

skin-derived tract connecting the skin surface to the dura through a bony opening and fascia defect [22].

Besides the earlier mentioned common occult dysraphic states, meningocele manqué, neurenteric cysts, terminal syrinx, and caudal regression syndrome are other forms that occur with disordered embryogenesis at various stages of mesodermal differentiation. Although these pathologies may occasionally have a similar pattern in terms of neurological insult, they usually require a different algorithm for treatment and will not be discussed here.

15.1.2.4 Epidemiology

The true incidence of occult dysraphism is very difficult to assess. Unlike open dysraphisms, closed defects may be asymptomatic throughout life and diagnosed with the onset of symptoms or incidentally found during workup of unrelated problems. Unlike the reported decline of the incidence of myelomeningocele, the incidence of closed dysraphisms has been increasing due to greater clinical awareness and incidental detection provided by magnetic resonance imaging (MRI) [29].

15.1.2.5 Presentation

Although malformations within the occult dysraphism group exhibit diverse pathological and clinical properties due to the different embryological step involved, there is a general tendency to unite all these malformations due to a similar pathophysiological mechanism by which symptoms arise. Almost all of the described malformations have a high rate of associated anomalies that would initiate a set of events as scoliosis, lower extremity deformities, and genitourinary anomalies apparent at birth, but a substantial number of cases are born with no sign at all except for some cutaneous lesions. The existing neurological symptoms or a potential for developing neurological symptoms in time is attributed to a great extent to cord tethering, a term which is used interchangeably with occult spinal dysraphism.

The theory of the tethered spinal cord is based on the pathological fixation and mechanical stretching of the lumbosacral cord. During embryogenesis, the spinal column elongates and grows much faster than the neural tissue. While neural and corresponding vertebral levels lie on the same plane until the end of the third gestational month, the different rate of growth results with conus medullaris to ascend and move to almost the L2 vertebral level at term. Mesodermal disarrangement during secondary neurulation in which spinal cord tissue lies adjacent to the same vertebral segments prevents the conus from ascending, and it remains at its original low-lying

position (see Fig. 15.1). Within this context, it is postulated that any inelastic structure like a thick and fatty filum, bony septum, or lipoma anchoring the caudal end of the spinal cord that prevents cephalad movement causes chronic and progressive ischemic spinal cord injury. The resulting clinical outcome includes lower limb motor and sensory deficits, incontinence, and musculoskeletal deformities of various degree and combination [2, 30, 31]. Although the tethered spinal cord concept and its pathophysiology are universally accepted, there is major controversy on certain aspects, especially regarding treatment algorithms of this syndrome [32–34].

15.1.2.6 Treatment

There are different clinical presentations in a given type of occult dysraphisms. For Appert spinal dysraphisms, where underlying embryological disarrangement, pathological anatomy, and the clinical consequence are almost identical for any case, the algorithm for surgical treatment is straightforward. Unlike the Appert forms, occult dysraphism presents within a wide spectrum of clinical findings from asymptomatic to severe neurological dysfunction. The true incidence and natural course is not clear, which further perplexes the decision-making process.

The surgical intervention for occult dysraphism is more or less surgery for the tethered spinal cord, with the ultimate goal being to improve or stabilize deficits in the symptomatic patients and to prevent future deficits in the asymptomatic ones by detethering the spinal cord. Within this context, decision making is straightforward in those with significant dysraphic abnormality and clear clinical deterioration. Potential benefits of surgery are expected to outweigh the risks. For those with normal neurological findings or stable deficits and in those with an incidentally discovered abnormality, the decision-making process becomes less clear and more controversial [32, 35]. While it has been commonly accepted that clinical deterioration is inevitable in asymptomatic cases and therefore prophylactic surgery should be undertaken, conflicting data exist in this regard. Certain forms of disease like thick/fatty filum can be prophylactically treated with a high level of confidence, especially the complex malformations such as lipomyelomeningoceles that carry a significant neurological morbidity (Fig. 15.6a–d). Complications in patients with preexisting deficits might be more tolerable, while complications in neurologically normal patients are particularly distressing to surgeons as well as to the patients [32, 36–38]. A common scenario is a child with coexisting malformations of different patterns of maldevelopment. SCMs and coexisting vertebral segmentation with scoliosis and urogenital malformations in caudal regression syndrome require different specialties to work simultaneously. There is no evidence-based treatment algorithm regarding staging or precedence of approach by different surgical teams. While there is no conflict on removing a bony spur and dural reconstruction of a type 1 SCM before correction of scoliosis at the corresponding segments, the benefits gained by removing those remote and neurologically intact SCMs (either type 1 or 2) or other forms of radiologically diagnosed dysraphisms just for the sake of safe scoliosis correction are questionable.

Diastematomyelia has long been known to be associated with spinal deformity. In a series of patients with diastematomyelia, Hood et al. have shown this disorder to be associated with scoliosis in 60 % of cases [39]. Patients present with either scoliosis or a neural deficit as their primary complaint, with the diastematomyelic spicule most frequently located at the first or third lumbar vertebra. McMaster reported the presence of an occult intraspinal anomaly in one-fifth of patients presenting with congenital scoliosis, with diastematomyelia accounting for over 90 % of these anomalies [40]. Most patients had abnormal neurological findings (65 %), while 75 % also had cutaneous abnormalities. Recently, in a large series of Chinese patients with congenital scoliosis, diastematomyelia was found to be the most common intraspinal abnormality, occurring in over 40 % of patients [41]. The higher rate of detected abnormalities is likely due to the use

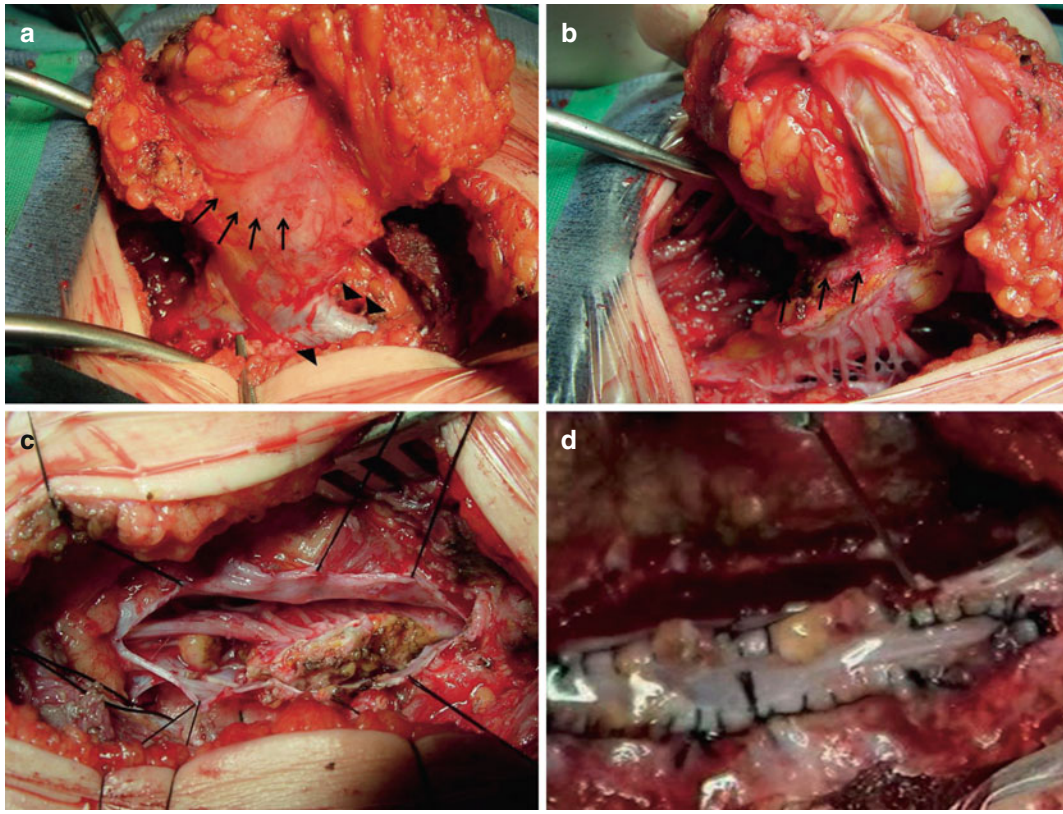

Fig. 15.6 Lipomyelomeningocele, basic steps for excision and untethering. (**a**) Subcutaneous part is freed from surrounding soft tissue and subcutaneous fat and followed to the entrance into intraspinal compartment through the enlarged multilevel spina bifida (*arrowheads*) and dural defect (*arrows*). (**b**) Intradural component is removed from the spinal cord tissue leaving a layer of fat at the cord–lipoma interface (*arrows*). (**c**) Final appearance following resection. Note the transverse orientation of the roots unlike the normal longitudinal arrangement of cauda equina due to ascendance of conus during embryogenesis. (**d**) Dural closure should be done with a generous dural patch to prevent cerebrospinal fluid leakage and retethering

of MRI, which is a much more sensitive modality for detecting and evaluating developmental spinal anomalies. In this series, a bony septum was found 40 % of the time, while a fibrous septum accounted for the remainder of cases. Importantly, extra-spinal defects occur in 40 % of these patients, with cardiac anomalies being most common, and hence a thorough clinical workup is warranted, especially for surgical candidates.

Previously, correction of any spinal deformity in a patient with diastematomyelia was felt to be unsafe until the intraspinal anomaly was first surgically addressed [39, 42]. Resection of the bone spur leads to marginal neurological improvement in about 50 % of patients and has no effect on curve progression. Winter et al. [43] advocated resection of the bony spicule first and a return to the operating room in 3–6 months for correction of any associated spinal deformity. However, this approach necessitates two operative procedures, during which time retethering of the spinal cord can occur. Hamzaoglu et al. [44] have reported the simultaneous treatment of both the diastematomyelic spicule and the concomitant spinal deformity. The group treated 13 patients with diastematomyelia using a posterior approach in which the bony spicule was first resected, followed by instrumentation and correction of deformity. No patient suffered neurological deterioration following surgery. More recently, Ayvaz et al. [45] have questioned whether a diastematomyelic abnormality needs

to be addressed at all prior to deformity correction. In their series of 32 patients with congenital spinal deformity, patients with a type 2 diastematomyelia underwent deformity correction without surgical intervention for the intraspinal abnormality while patients with a type 1 diastematomyelia underwent resection of the bony spicule with subsequent instrumentation and correction. Patients with type 2 diastematomyelia did not suffer any neurological deterioration as a result of corrective spinal surgery alone. Therefore, surgical treatment of diastematomyelia remains a controversial topic. Generally, however, there is some consensus that patients with scoliosis and diastematomyelia as their only abnormality should be treated surgically. Likewise, patients with a type 1 diastematomyelia and concomitant developmental abnormalities should undergo simultaneous neurosurgical intervention and orthopedic correction, while patients with a type 2 diastematomyelia may be safely treated with corrective surgery alone.

Recurrent tethered cord syndrome represents an especially difficult entity to treat. Following surgical intervention, recurrent tethering can occur in up to 50 % of patients who were completely detethered and in up to 80 % of patients where the spinal cord was only partially untethered [46, 47]. Although laminectomy and detethering is the mainstay approach for initial detethering, this method is fraught with complications when employed in patients with recurrent tethering, often leading to progressive neurological decline and retethering. Grande et al. [47] demonstrated in a cadaveric study that vertebral column shortening can lead to a significant reduction of tension in the nerve roots. To achieve this, a vertebral column resection and reduction was performed at the T11/T12 level, leading to about 20–25 mm of vertebral column shortening. Hsieh et al. [46] have reported the use of this technique in the treatment of two patients with recurrent tethered cord syndrome. A posterior vertebral column subtraction osteotomy was performed at the thoracolumbar junction using a staged approach, leading to 20 mm of vertebral column shortening. One patient had a stable neurological examination 1 year after surgery, reporting slightly improved urodynamic function, while the other reported significantly improved motor, sensory, and bladder function after 1 month. Thus, vertebral column resection, a well-known and described technique in spinal deformity surgery, represents a viable, alternative approach to the treatment of recurrent tethered cord syndrome.

No clear diagnostic or treatment strategy based on basic research and prospective clinical trials exists for occult dysraphism and tethered spinal cord. Until the results of such research are available, indications for treatment remain confined to personal experience, expertise, and complexity of the lesion, with the substantial risk of over- or undertreatment.

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Neurofibromatosis

16

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Key Points

- Early onset scoliosis in NF-1 patients would more likely be dystrophic.
- Modulation may be the result of unrecognized intra and/or extra spinal pathology. Total spinal and brain MRIs are recommended in all NF-1 patients with spinal deformities.
- Dural ectasia and paraspinal tumors may erode the anchor purchase sites. Preoperative CT is indicated for all dystrophic spinal deformities to access potential anchor sites.
- The distorted anatomy of the posterior elements in these patients may require the use of hooks, screws and sublaminar wiring to establish anchors.
- The current state-of-the-art treatment for significant deformity or progressive deformities in the patient achieving

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skeletal maturity is combined anterior and posterior spinal arthrodesis.
- If the child is very young (under 5–6 years), a corrective cast or bracing may be attempted, most often with little to marginal success.
- Growing rods have been used to obtain correction without definitive fusion and to lengthen or "grow the spine" every 6 months, but with varying success and a high rate of complications.
- In severe early dystrophic deformities, we have successful experience performing annulotomies for anterior release, traction, and posterior growing rod stabilization. This allows us to achieve further spinal length before the subsequent anterior discectomy and fusion at maturity.

16.1 Introduction

Neurofibromatosis is a multisystemic, autosomal dominant genetic disorder defined as a spectrum of multifaceted diseases involving neuroectoderm, mesoderm, and endoderm. The clinical features of neurofibromatosis type 1 (NF-1), the most common form of the disease, were reported in several family members by German pathologist Virchow in 1847, [1] but it was his student von Recklinghausen [2] who 35 years later described the histological features of the syndrome that often bears his eponym.

NF-1 is characterized by extreme variability of expression. The proposed mechanisms for this variability include germline-modifying genes, environmental agents, second hit somatic mutation events in *NF-1* or other genes, epigenetic modification, and post-zygotic mutations [3]. The NF-1 phenotypes vary to a greater degree with increasing distance from a proband, thus documenting that the specific familial *NF-1* mutation is not the primary cause of variability [4]. Common clinical manifestations include café-au-lait macules, neurofibromas, and schwannomas. Skeletal complications usually present early in life and can be attributed to abnormalities of bone growth, remodeling, and repair in NF-1 or can be secondary to nearby soft-tissue abnormalities complicating NF-1.

Skeletal complications can be categorized as generalized or focal manifestations [5]. Generalized skeletal abnormalities include osteoporosis/osteopenia, osteomalacia, shortness of stature, and macrocephaly. These features are common in individuals with NF-1, with decreased bone mineral density in both sexes reported in up to 50 % of the patients, but usually mild [6–9]. Focal abnormalities of the skeleton are less common than generalized abnormalities, but may cause significant morbidity. Focal manifestations include spinal deformities, dysplasia of the tibia and other long bones, sphenoid wing dysplasia, chest wall deformities (pectus excavatum), dental abnormalities, periapical cemental dysplasia, and cystic osseous lesions. The effect of generalized abnormalities in the occurrence or progression of focal skeletal manifestations remains elusive.

The incidence of spinal deformities in association with NF-1 varies from 2 to 36 % with scoliosis being the most common musculoskeletal manifestation of NF-1 [10, 11]. The intent of this chapter is to present the spinal deformities that are most commonly associated with NF-1 and to identify the current management of spinal disorders based on the most recent literature.

16.2 Classification

Five distinct clinical forms of neurofibromatosis are currently accepted by most investigators: NF-1, NF-2, segmental NF-1, Legius syndrome, and schwannomatosis.

16.2.1 Neurofibromatosis 1 (NF-1)

NF-1 or peripheral neurofibromatosis is a common autosomal dominant single-gene disorder with an estimated prevalence of 1:3,000 [12]. It is the most common form of neurofibromatosis and

the one most likely to be encountered by the orthopedist. It is predicted to affect over two million people worldwide in all racial and ethnic groups. The *NF-1* gene is large in size, in the range of 350,000 base pairs with 59 exons, and its locus was discovered on chromosome 17q11.2 [12–14]. *NF-1* is a tumor-suppressor gene that encodes neurofibromin, a large cytoplasmic protein with 2,818 amino acids. Exons 21 through 27a encode a 360 amino-acid domain with homology with guanosine triphosphatase (GTP)-activating proteins (GAPs). The relevant domain, known as GAP-related domain (GRD), downregulates p21-Ras oncogene which promotes cell growth, proliferation, and differentiation. GAPs, including neurofibromin, inactivate Ras oncogene through their GTPase activity. Decreased synthesis or complete absence of neurofibromin expression, as in NF-1, results in unopposed activation of p21-Ras oncogene through GTP binding. This, in turns, leads to aberrant growth-promoting signals and the development of NF-1-associated neoplasms, including benign neurofibromas, malignant peripheral nerve sheath tumors, pheochromocytomas, and optic nerve gliomas, as well as to other clinical manifestations [15, 16].

The *NF-1* gene displays almost complete penetrance. Individuals with NF-1 are constitutionally heterozygous for an *NF-1* gene loss-of-function mutation. Approximately 50 % of affected individuals inherited the gene from an affected parent and 50 % arise sporadically due to spontaneous mutations [16–19]. De novo mutations in the *NF-1* gene are associated with advanced paternal age [19].

The diagnosis of NF-1 is established when at least two of the most commonly presenting features of the disease as defined by the 1987 Consensus Development Conference of the National Institutes of Health are present (Table 16.1) [20]. In 97 % of patients, a diagnosis is made by age 8 [21]. Molecular diagnosis with direct sequencing of the causative mutation is possible in 95 % of patients with NF-1 and is indicated in uncertain cases and for prenatal diagnosis [22]. Differential diagnosis includes tuberous sclerosis and other conditions of pigmentation, such as McCune–Albright syndrome and mastocytosis. NF-1 is closely related to a number of other genetic syndromes involving mutations of the Ras pathway, such as Noonan syndrome, cardio-facial-cutaneous syndrome, and LEOPARD syndrome. Of interest, these other Ras pathway syndromes can have overlapping orthopedic manifestations with NF-1.

Table 16.1 Diagnostic criteria for NF-1 as defined by the 1987 Consensus Development Conference of the National Institutes of Health [20]

1	Six or more café-au-lait macules more than 5 mm in greatest diameter in prepubertal individuals and more than 15 mm in postpubertal individuals
2	Two or more neurofibromas of any type or more than one plexiform neurofibroma
3	Freckling in the axillary or inguinal regions
4	Two or more Lisch nodules (iris hamartomas)
5	Optic glioma
6	A distinctive osseous lesion, such as sphenoid dysplasia or thinning of long bone cortex, with or without pseudarthrosis
7	A first degree relative (parent, sibling, or offspring) with NF-1 by the above criteria

Based on data from Ref. [20]

16.2.2 Neurofibromatosis 2 (NF-2)

NF-2 or central neurofibromatosis has an estimated incidence of 1 in 33,000 individuals and is associated with bilateral vestibular schwannomas and multiple spinal shwannomas [23, 24]. The NF-2 locus is located on the long arm of chromosome 22. Fifty percent of cases involve a new mutation. NF-2 is not associated with primary skeletal disorders; however, multiple paraspinal and intraspinal tumors (schwannomas and ependymomas) are common in this disorder. NF-1 and NF-2 are genetically distinct disorders with different gene loci, despite similarities in names.

16.2.3 Segmental Neurofibromatosis

Segmental neurofibromatosis is characterized by features of NF-1 involving a single body segment. Typically, only a single segment of the

body (such as left upper extremity) is affected with café-au-lait spots and freckling, and lesions usually do not cross the body midline. Other segmental forms may involve deep neurofibromas in a single body segment. It is considered as a somatic mosaic form of NF-1, and typically is not associated with cognitive effects or learning disabilities seen in NF-1.

16.2.4 Legius Syndrome

Early neurofibromatosis literature recognized that a mild form of NF-1 existed, consisting primarily of familial café-au-lait spots. In recent years, multiple families with such mild involvement have now been found to have mutations in the *SPRED1* gene. Initially discovered by Legius et al. [25] this condition, now called Legius syndrome, can present with multiple café-au-lait spots, freckling, macrocephaly, and mild learning disabilities, but does not present with any of the benign or malignant tumors associated with NF-1. This condition is quite a bit less common than NF-1, with an estimated prevalence of about 1/50,000. Since patients with Legius syndrome can actually meet the clinical diagnostic criteria for NF-1, it can be appropriate to perform molecular testing if there is any question about diagnosis.

16.2.5 Schwannomatosis

Schwannomatosis is a distinct form of neurofibromatosis which typically involves multiple schwannomas throughout the body, but without the vestibular schwannomas typical of NF-2. Initially thought to represent a mosaic form of NF-2, it has now been determined that familial schwannomatosis is due to mutations in the *INI1* gene, linked to *NF-2* on chromosome 22. It is a disease of adulthood that consists of multiple deep painful peripheral nerve sheath tumors that may occur in a generalized form or in a segmental distribution. Differential diagnosis from NF-2 can be difficult, and genetic testing of NF-2 and *INI1* is now available to help in making this distinction.

16.3 Spinal Abnormalities in NF-1

16.3.1 Epidemiology

Spinal abnormalities are the most common orthopedic manifestation of NF-1. It is quoted as from 2 to 36 % in the literature [10, 11]. In the NF clinic at our institution, it is 23 % [12]. In a report in 1988, Winter et al. [26]. found only 102 patients having NF-1 by clinical criteria in a pool of approximately 10,000 patients with scoliosis. Functional scoliosis resulting from limb hypertrophy or long-bone dysplasia leading to limb length inequality must be ruled out in patients with NF-1. Rarely, unrecognized extra-pleural thoracic tumors can present as focal scoliosis. These lesions are usually plexiform neurofibroma and are not visible on plain radiographs [27]. The spinal deformities tend to develop early in the life therefore, all preadolescent children with NF-1 should be evaluated by scoliosis screening or the Adam forward-bend test to rule out the presence of a spinal deformity.

It is important to emphasize that there is no standard pattern of spinal deformity in NF-1. All manner of spinal deformities in multiple planes and in any part of the spine may occur with NF-1 [28, 29]. The characteristic deformity tends to be a short-segmented, sharply angulated curvature that usually involves four to six vertebrae in the upper third of the thoracic spine [30]. We have traditionally classified the deformities into dystrophic or non-dystrophic types based on the coronal plane x-rays. The entire spine may be affected by deformity in the coronal and sagittal planes. There are nine radiographic criteria most often used to classify the deformity as dystrophic. These include rib penciling (the rib being smaller in diameter than the second rib), vertebral rotation, posterior vertebral scalloping, vertebral wedging, spindling of the transverse process, anterior vertebral scalloping, widened interpedicular distance, enlarged interverteral foramina, and lateral vertebral scalloping. Recently, two more magnetic resonance imaging (MRI) findings have been added to the criteria used to classify the deformity as dystrophic: the presence of dural ectasia and the presence of paraspinal tumors (Table 16.2) [31]. More

Table 16.2 Diagnostic criteria of dystrophic spine

1	Rib penciling
2	Posterior vertebral scalloping
3	Vertebral wedging
4	Spindling of transverse processes
5	Anterior vertebral scalloping
6	Widened interpedicular distance
7	Enlarged intervertebral foramina
8	Lateral vertebral scalloping
9	Vertebral rotation
10	Paraspinal tumors
11	Dural ectasia

than three of these dystrophic features are considered diagnostic of dystrophic scoliosis. Non-dystrophic curves are considered similar to idiopathic scoliosis.

Ten to thirty-three percent of children with NF-1 have spinal deformity [16]. All preadolescent children with NF-1 should be evaluated by the scoliosis screening or the Adam's forward-bend test to rule out the presence of a spinal deformity, which usually occurs earlier in children with NF-1 [30].

16.3.2 Etiology

The cause of spinal deformity remains unknown. Several theories including metabolic bone deficiency, osteomalacia, endocrine disturbance, and mesodermal dysplasia have been proposed and are at best inconclusive [32–36]. The dystrophic changes may be attributed to intrinsic factors or may be associated with anomalies of the spinal canal secondary to abnormalities of the spinal cord dura mater.

Pressure erosive effects of dural ectasia and paravertebral tumors have been frequently found to be adjacent to and approximated to the deformities, initiating instability and subsequent deformity. Dural ectasia, a disorder unique to certain conditions, is an expansion or dilatation of the dural sac. The changes in the spinal canal induced by dural ectasia may increase the difficulty in obtaining adequate purchase for fixation of anchors during spinal deformity correction.

Scalloping was initially thought to represent the result of erosive pressure or direct infiltration of the vertebra by adjacent neurofibroma [37–41]. A neurofibroma-derived locally active biochemical substance or hormone that triggers dystrophic features in the adjacent vertebra has also been proposed [37]. The presence of an altered response of the vertebral bone in NF-1 to a paraspinal tumor has been hypothesized. An interactive pathophysiological mechanism between a genetically compromised bone and a neuroectodermal derivative, such as a contiguous neurofibroma or an abnormal meningeal sheath, is suggested by some authors [37, 39].

The etiological theory of vertebral scalloping being a primary developmental defect was supported by the presence of scalloping without adjacent lesions [42]. This was also supported by an MRI study in patients with NF-1, in which posterior vertebral scalloping was highly associated with dural ectasia, lateral scalloping was related to dural ectasia or neurofibromas in 50 % of cases, and anterior scalloping was unrelated to dural ectasia or tumors [43]. The authors could not identify any association with dural ectasia or paraspinal tumors in more than one-third of their patients with MRI evidence of vertebral scalloping. Nevertheless, dural ectasia without associated vertebral scalloping was recorded in 10 % of the cases.

A recent study in ten monozygotic twins with NF-1 demonstrated mixed concordance and discordance for presence of scoliosis [3]. The affected twin pairs were discordant for presence of dystrophic features, degree of curvature, and need for surgery. This finding suggests that both heritable and nonheritable factors contribute to the pathogenesis of spinal deformities in NF-1 patients. Dystrophic curves most likely require a nonhereditary event, such as an adjacent tumor or dural ectasia, or a second hit event in local bone cells leading to the underlying dysplasia. If occurrence and progression of dystrophic spinal deformity is affected by adjacent neurofibromas, then therapies targeting to reduction or stabilization of paraspinal tumors could provide a promising approach to spine deformity prevention in patients with NF-1.

Apart from its tumor suppressor activities through the Ras signaling, the role of neurofibromin

may converge with other bone biochemical pathways, such as bone morphogenetic protein (BMP) signal transduction [44]. This theory suggests that intrinsic bone pathology due to loss of a functional *NF-1* allele with subsequent Ras deregulation may be responsible for osseous manifestations in NF-1 through altered osteoblastic/osteoprogenitor differentiation, overgrowth of cellular tissue due to preferred fibroblast differentiation of mesenchymal cells, and impaired bony callus formation. Double inactivation of NF-1 by somatic mutation of the *NF-1* gene in a population of cells which depends on neurofibromin-regulated Ras signaling to maintain normal bone was suggested to contribute to the occurrence or progression of tibia pseudarthrosis [45]. Although such second hit events have been demonstrated in pathological tissue from NF-1 tibias, it is unknown whether spinal deformities of NF-1 require a second hit event.

16.3.3 Mouse Models

The NF-1 heterozygous mouse has a minimal skeletal phenotype. In order to better understand the mechanism of skeletal abnormalities in NF-1, researchers have developed more complex mouse models with knock-out of the second *NF-1* allele in osteoprogenitor cell lines, using a process called cre-recombination. In one such model, Col2.3Cre(+) mice showed multiple vertebral anomalies, including: short vertebral segments, reduction in cortical and trabecular bone mass of the vertebrae, increased numbers of osteoclasts, and decreased numbers of osteoblasts in vertebrae [45]. These mouse models provide additional insight to the underlying pathophysiology of spinal abnormalities in humans with NF-1.

16.3.4 Imaging

Most often plain standing posterior–anterior and lateral radiographs are sufficient for screening the curvature. An angle of greater than 10° assigns the deformity as structural. When treatment is to be initiated, multiple planar films in supine bending modes and traction are necessary to determine flexibility. If there are adjacent structures requiring further clarification, higher levels of imaging are required, such as computed tomography (CT) for bony deformity or high-resolution contrast CT or MRI for soft tissue delineation.

16.3.5 Dural Ectasia

Dural ectasia is a circumferential dilatation of the dural sac which is filled with proteinaceous fluid. The slow expansion of the dura results in erosion of the surrounding osseous structures resulting in widening of the spinal canal, thinning of the laminae, and ultimately destabilization of the spine. Dural expansion through the neural foramina can cause meningoceles giving the radiographic dumbbell appearance. However, enlargement of a single neural foramen on an oblique radiograph is usually caused by neurofibroma exiting from the spinal canal rather than from the dural ectasia (Fig. 16.1). Similar lesions are seen in other connective tissue disorders, e.g., Marfan's syndrome and Ehler–Danlos syndrome, although cause of these lesions in NF-1 is not known.

During this process, the neural elements are not affected. As a result of slow nature of this process and enormous widening of the spinal canal the neural elements have adequate room for accommodation, and there may be severe angular deformity and distortion without neurological deficit. The patients remain neurologically intact until later in the course of the disease process when destabilization of the vertebral column jeopardizes the neural elements. Dislocation of the vertebral column due to dural ectasia has been reported in the literature [46]. The destabilization at the costovertebral junction can result in penetration of the rib head into the spinal canal with neurological compromise (Fig. 16.2) [47, 48]. The presence of rib head or the neurofibroma in the spinal canal can result in intraoperative neurological deficit if instrumentation is used for correction of the curve without adequate decompression.

Dural ectasia can be readily seen on high-volume CT myelography or contrast-enhanced MRI and is recommended before surgical intervention is undertaken for dystrophic curves. Higher imaging studies help to demonstrate extremely thin laminae; in which case dissection by electocautery rather than by periosteal eleva-

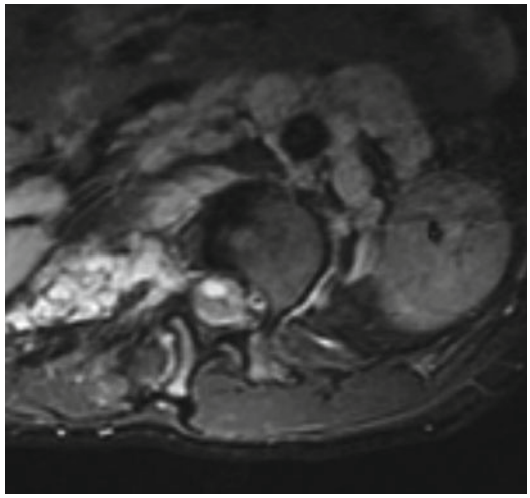


Fig. 16.1 MRI of the spine with the neurofibroma in canal. Bright shadow depicts neurofibroma exiting through the spinal canal. The constriction of the neurofibroma in the foramen gives it the appearance of a dumbbell

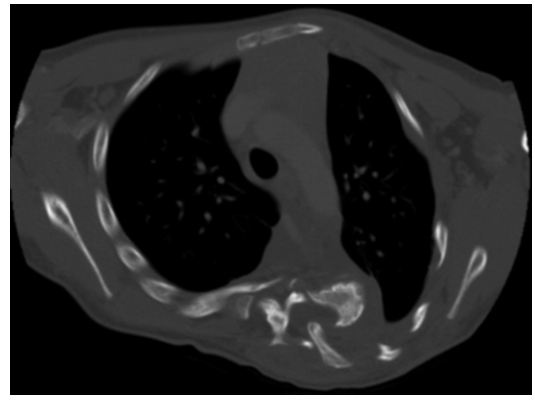
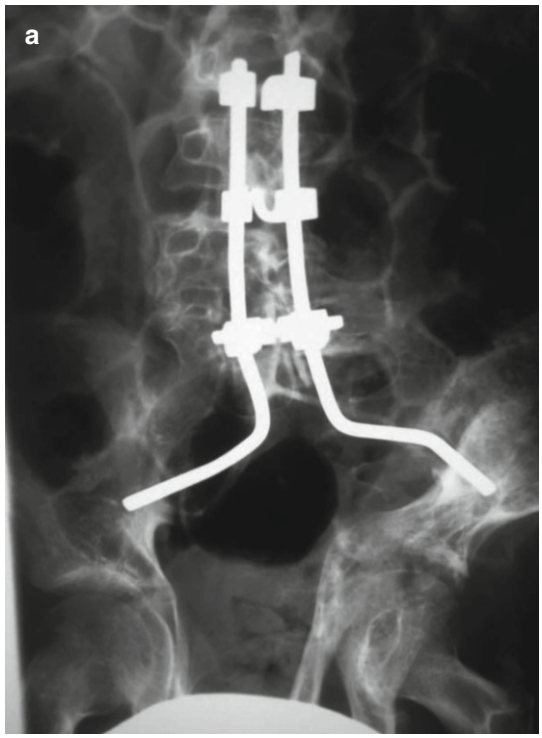


Fig. 16.2 Dislocation of rib head in the spinal canal in a severe dystrophic thoracic curve. Careful evaluation of the preoperative imaging including CT scan is essential to identify this pathology. Decompression prior to correction is essential to prevent intraoperative neurological complications



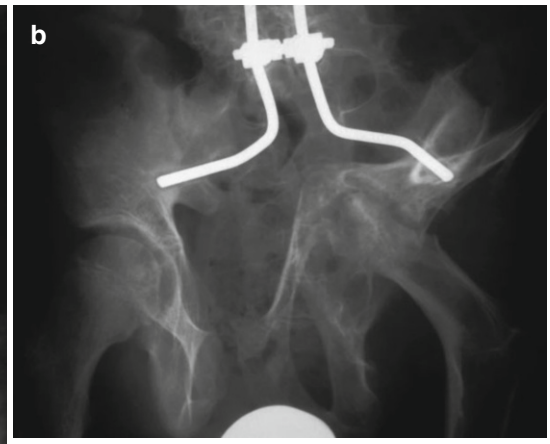


Fig. 16.3 (**a**) Neurofibromatosis in the lumbosacral spine. This patient was treated by fusion and instrumentation extending to the pelvis. (**b**) A few years later, the fusion mass and the vertebrae are eroded completely by expanding dural ectasia leaving behind the instrumentation. Note also the destruction of the hip joint by tumor

tors are recommended during surgical exposure to avoid direct injury to the neural elements/dura by plunging into the spinal canal. Surgical spinal stabilization and fusion does not alter the course of dural ectasia. Dural ectasia can result in failure of the primary fusion or the expanding dura ultimately can destroy a solid fusion leaving behind the instrumentation (Fig. 16.3).

Spinal affections in NF-1 can be described under following regions: cervical, thoracic/thoracolumbar, lumbosacral, and spinal canal.

16.4 Cervical Spine Abnormalities

The cervical spine abnormalities in NF-1 have not received enough attention in the literature [49, 50]. Usually, the cervical lesion is asymptomatic. When the lesion is symptomatic, pain is the most common presenting symptom [51]. Cervical abnormalities are likely to be missed in presence of scoliosis or kyphoscoliosis lower regions of the spine where the examiner's attention is focused on the more obvious deformity. In a study of 56 patients with NF-1, Yong-Hing et al. [52] reported that 17 patients (30 %) had cervical spine abnormalities. Out of these, seven patients were asymptomatic, whereas the rest had limited motion or pain in the neck. Four patients had neurological deficits that were attributed to cervical instability. Four of the 17 patients required fusion of the cervical spine. Curtis et al. [53] described eight patients who had paraplegia and NF-1. Four of these patients had cervical spine instability or intraspinal pathology in the cervical spine. The upper cervical spine should also be examined carefully. Isu et al. [54] described three patients with NF-1 who had C1–C2 dislocation with neurological deficit. All patients improved after decompression and fusion. We recommend that the cervical spine should be evaluated at the initial scoliosis assessment.

A lateral radiograph of the cervical spine is the initial screening tool. The NF-1 can be manifested on a plain radiograph in the form of dystrophic changes or malalignment [55]. If any suspicious area is noted on plain radiographs, right and left oblique views should be obtained to look for widening of the neuroforamina which may represent dumbbell lesions. MRI is the definitive study to evaluate these lesions.

Anteroposterior and lateral radiographs of the cervical spine should be obtained in all NF-1 patients who: (1) are placed in halo traction; (2) undergo surgery; (3) require endotracheal intubation; (4) present with neck tumors; (5) complain of neck pain; and (6) present with symptoms indicating intra- or extraspinal neurofibromas, such as torticollis or dysphagia [56]. If there is any suspicion of instability, CT and/or flexion-extension MRI are indicated. Erosive defects of the skull may be present in some patients with NF-1. Thus, plain radiographs of the skull prior to halo or Gardner–Wells tong traction pins application are strongly recommended.

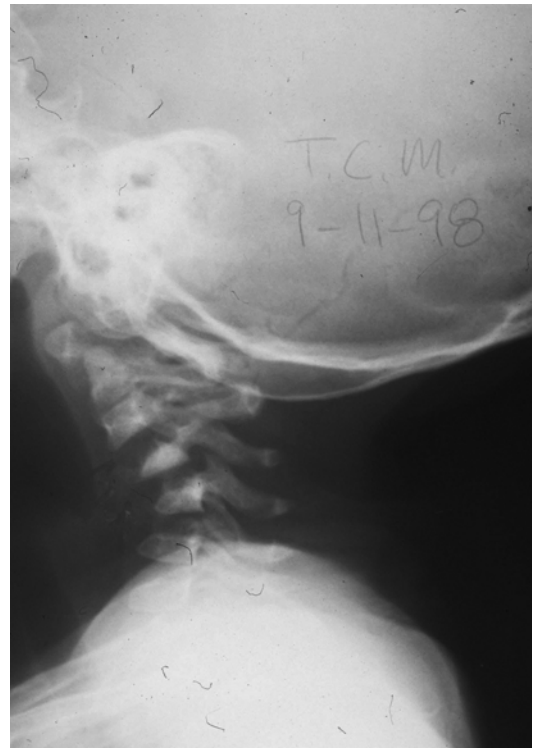


Fig. 16.4 Lateral radiograph of the cervical spine 6 months following laminectomy and excision of the neurofibroma, demonstrating marked kyphosis of the entire cervical spine. Note the dystrophic appearance of the vertebrae

The most common spinal abnormality in the cervical spine is a severe cervical kyphosis (Fig. 16.4), which is often seen following a decompressive laminectomy without stabilization for an intraspinal lesion and is highly suggestive of the disorder [57]. We recommend stabilization of the spinal column at the same time of surgical removal of tumors from the spinal canal.

Ogilvie reported on the surgical treatment of cervical kyphosis by anterior fusion with iliac-crest or fibular bone graft or both [51]. He considered halo traction to be a useful preoperative step if the kyphosis was greater than 45°. In the presence of progressive cervical kyphosis, we recommend preoperative halo traction only if the deformity is flexible as judged by the radiographs. This should be followed by posterior fusion. If the deformity is rigid, then an anterior soft-tissue release followed by traction is safer.

Internal fixation with pedicle and lateral mass screws is preferred for posterior instrumentation. Sublaminar wire fixation may be difficult secondary to dural ectasia and osseous fragility. For anterior fixation, we currently use bioabsorbable plates. Even with rigid instrumentation, postoperative halo immobilization is recommended until a fusion mass with trabecular pattern is seen on cervical CT.

16.5 Thoracic/Thoracolumbar Spinal Abnormalities

The two varieties of spinal deformity are well distinguished in these regions of the spine. Also, the natural history of spinal deformities is well studied for thoracic/thoracolumbar region.

Patients more likely to develop progressive scoliosis of the thoracolumbar spine are children under 7 years of age who have thoracic lordosis (sagittal plane angle of less than 20° measured from T3 to T12) and paravertebral tumors. There is a strong association between modulation and progression of the spinal deformity. More specifically, curves that acquire either three or more penciled ribs or a combination of any three dystrophic features will almost certainly progress [28]. Other factors that have been associated with substantial curve progression include: (1) high Cobb angle at presentation; (2) early age of onset; (3) abnormal kyphosis; (4) vertebral scalloping; (5) severe apical rotation; (6) location of the apex in the middle-lower thoracic spine; (7) penciling of one rib or more on the concave side or both sides of the curve; and (8) penciling of four ribs or more [34].

More recent MRI studies have questioned the theory of modulation [43]. Patients with radiographically labeled non-dystrophic curves have been found to have significant dysplastic changes on MRI. Having in mind the higher sensitivity of MRI in identification of dystrophic features than x-rays, we recommend characterization of the curve as dystrophic or not based on a combination of MRI and x-ray findings [31].

16.5.1 Non-dystrophic Scoliosis

This is the common variety of spinal deformity observed in NF-1. These curves behave similar to idiopathic curves with some differences [7, 9, 58]. This form usually involves 8–10 spinal segments. Most often, the deformity is convex to the right. However, these curves usually present earlier than the idiopathic curves and are more prone to progression. Furthermore, the rate of pseudoarthrosis following a fusion surgery is higher in these patients [49]. These differences can be attributed to the process of modulation and the underlying bone pathology. Compared to dystrophic curves, non-dystrophic curves tend to present in older children with less angulation and rotation of the deformity [59].

16.5.2 Dystrophic Scoliosis

This is an uncommon but malignant form of spinal deformity. It is characterized by early onset, rapid progression and is more difficult to treat [60, 61]. Typically, the dystrophic curve is a short-segmented, sharply angulated type that includes fewer than six spinal segments. Dystrophic curves may be associated with kyphosis and have a higher incidence of neurological injury [61, 62].

Dystrophic vertebral changes develop over time (Table 16.2). Dystrophic curves are found most commonly in the thoracic region (Figs. 16.5, 16.6, 16.7, 16.8, 16.9, and 16.10) [63].

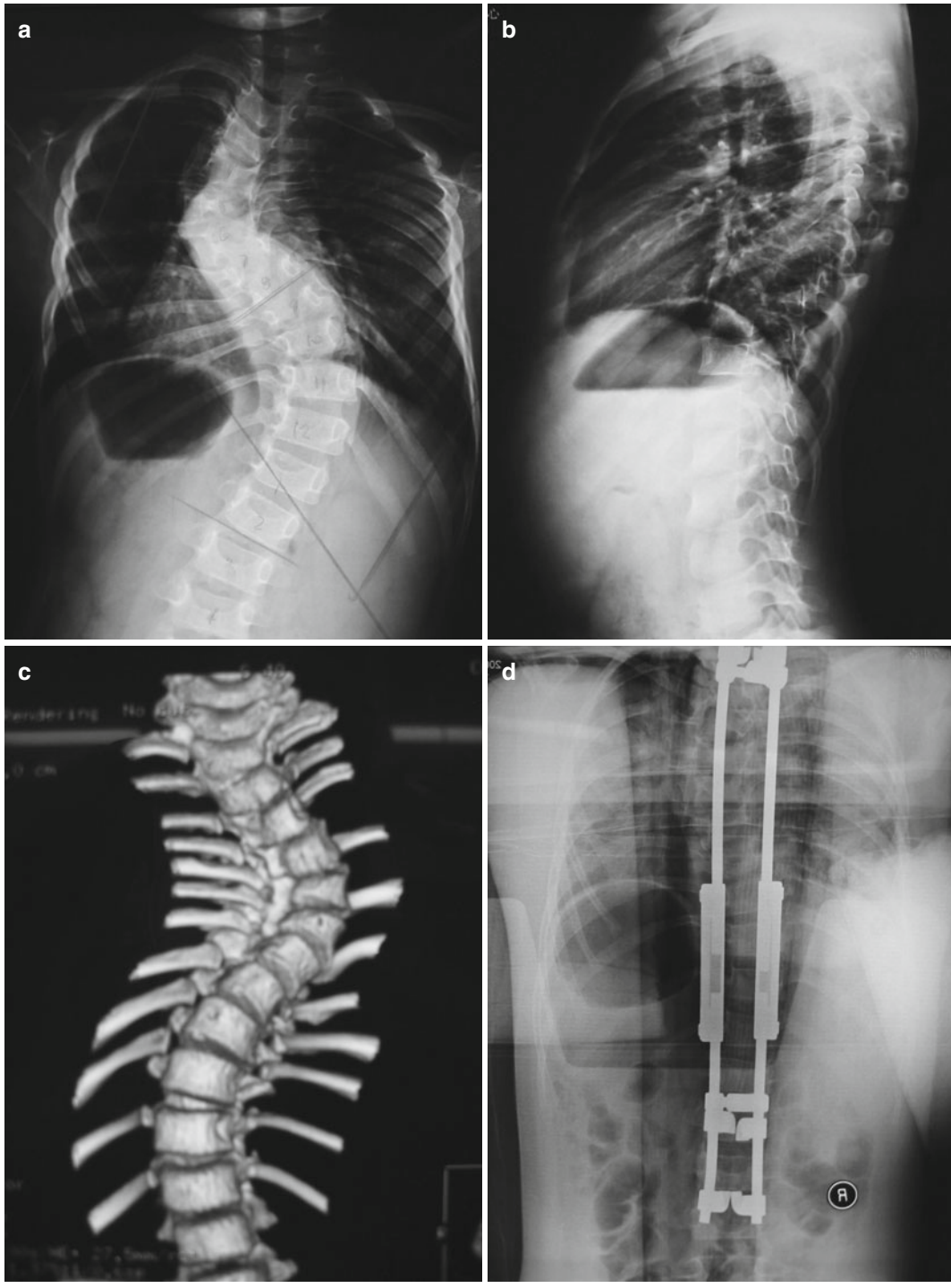


Fig. 16.5 A 6-year-old female with 80° thoracic scoliosis which was untreated. Preoperative 3D CT scan and x-rays (**a**–**c**) show presence of typical radiological features of dystrophic vertebral bodies. She underwent a growing rod instrumentation with hook anchors (**d**). At 2-year follow-up, the correction has remained stable and spinal length has increased following serial lengthenings at every 6-month intervals (**e**, **f**)

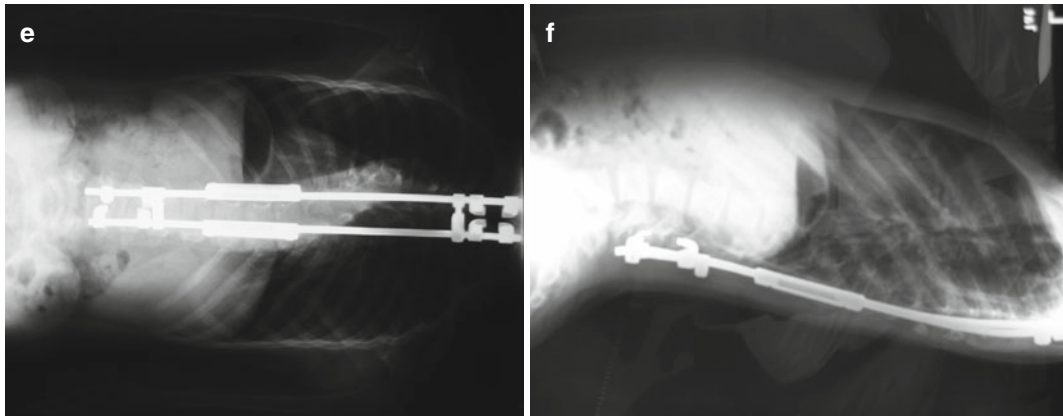


Fig. 16.5 (continued)

16.5.2.1 Natural History

The onset of spinal deformities may occur early in patients with NF-1. Usually early onset scoliosis is associated with kyphosis giving rise to kyphoscoliotic deformities. Calvert et al. [63] presented a series of treated ($n=34$) and untreated ($n=32$) patients who had NF-1 and scoliosis. Seventy-five percent of patients in the nontreated group had kyphoscoliosis. The investigators reported that patients, who had severe anterior vertebral scalloping noted on the lateral view, progressed an average of 23° per year for scoliosis and kyphosis. All other patients had an average rate of scoliosis progression of 7° and kyphosis progression of 8° per year.

Some of the non-dystrophic curves exhibit the phenomenon of modulation. Durrani et al. [28] defined modulation as a process by which a non-dystrophic curve acquires the features of a dystrophic curve and behaves as a dystrophic curve. They reported that modulation occurred in about 65 % of their patients. Modulation occurred in 81 % of patients who presented with scoliosis before 7 years of age and in 25 % of those diagnosed after 7 years of age. Rib penciling acquired through the modulation period was the only factor that was statistically significant in influencing the progression of the deformity. The rate of progression for "modulated" scoliosis and kyphosis was 12° and 8°, respectively, versus 5° and 3° for non-modulated spines. These authors based their report on plain radiographic findings. Some of the recent reports with the use of MRI of spine have shown the presence of dystrophic findings in the spine before they are apparent on the plain radiographs. Based on these reports, it can be speculated that true modulation may be rare, and many of the apparent non-dystrophic curves are actually dystrophic curves which subsequently present themselves with radiographical changes of dystrophic curve giving an impression of modulation.

A retrospective review of 694 patients with NF-1 revealing 131 patients (19 %) with a scoliosis ranging from 10° to 120° was performed at the Cincinnati Children's Neurofibromatosis Clinic [31]. Mean age at diagnosis of scoliosis was 9.0 years, with 18 patients (15 %) having onset before 6 years of age. Forty-six patients (35 %) required surgical repair, usually anterior and posterior spinal fusion with instrumentation. Six patients had growing rods successfully placed. Tumors near the spine were found in 65 % of patients requiring surgery. A subset of 56 patients with complete imaging and curvatures of greater than 15° was reviewed. Of this group, 70 % had three or more dystrophic features on plain radiographs or MRI. The data confirmed the existence of at least two distinct types of scoliosis; the first being a curve similar to idiopathic scoliosis and a second type with dystrophic changes that were more likely to progress. The presence of three or more dystrophic features on radiographs or MRI was highly predictive for the need for surgery.

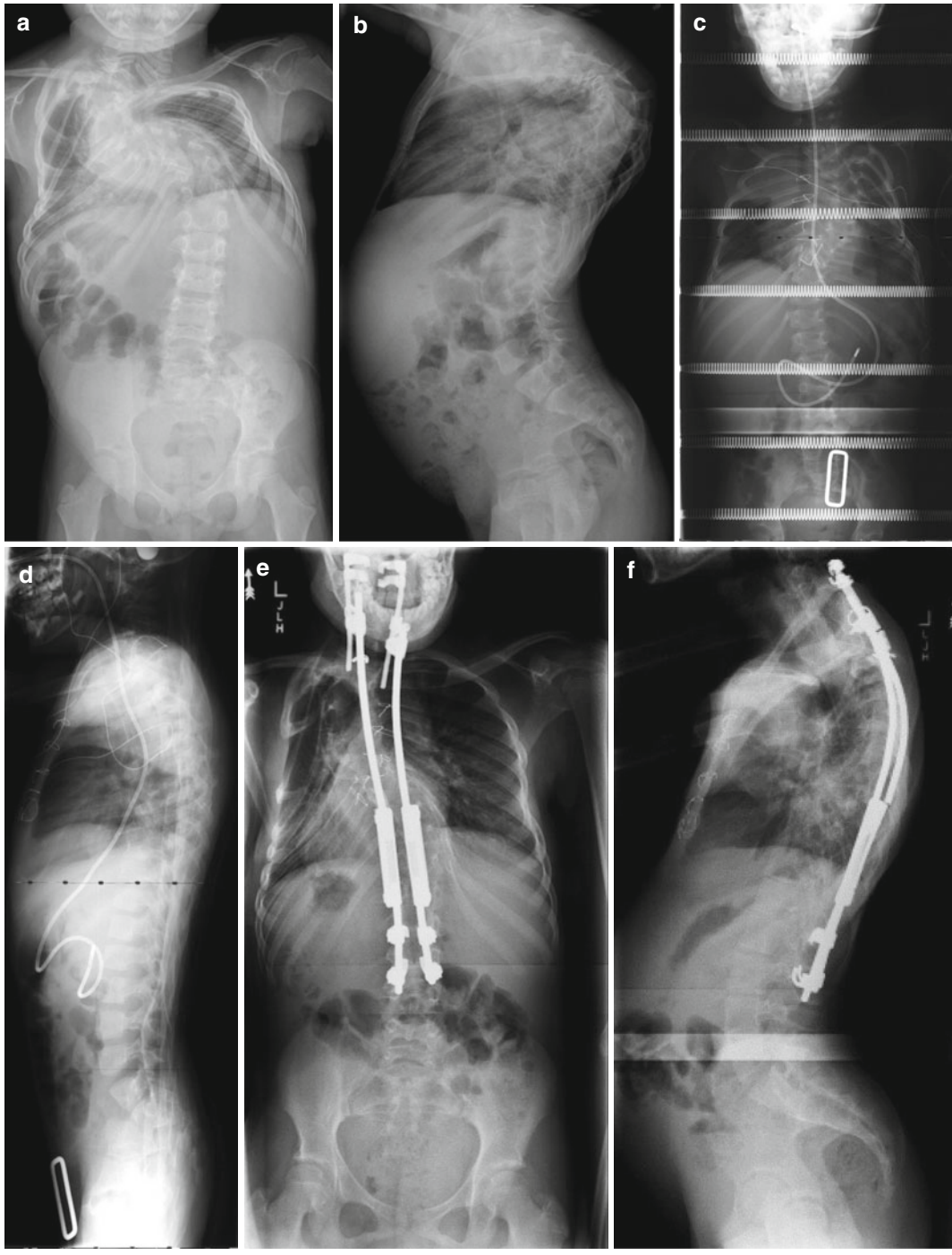


Fig. 16.6 (**a**, **b**) A 6-year-old female with severe thoracic dystrophic kyphoscoliosis. The deformity involves mid and upper thoracic spines. The curvature measures more than 100° in both planes. (**c**, **d**) She underwent an anterior release (annulotomies) through a double "trap door" approach for her upper thoracic and mid thoracic curve followed by a period of 2 weeks of halo-femoral traction. Anterior release with gradual traction alone resulted in significant correction of the deformity. (**e**, **f**) After the traction, the patient underwent growing rod instrumentation with proximal anchors in her lower cervical spine. At 5 years of follow-up, although one rod is broken, her correction is well maintained and her spinal height has increased as measured by digital radiographs

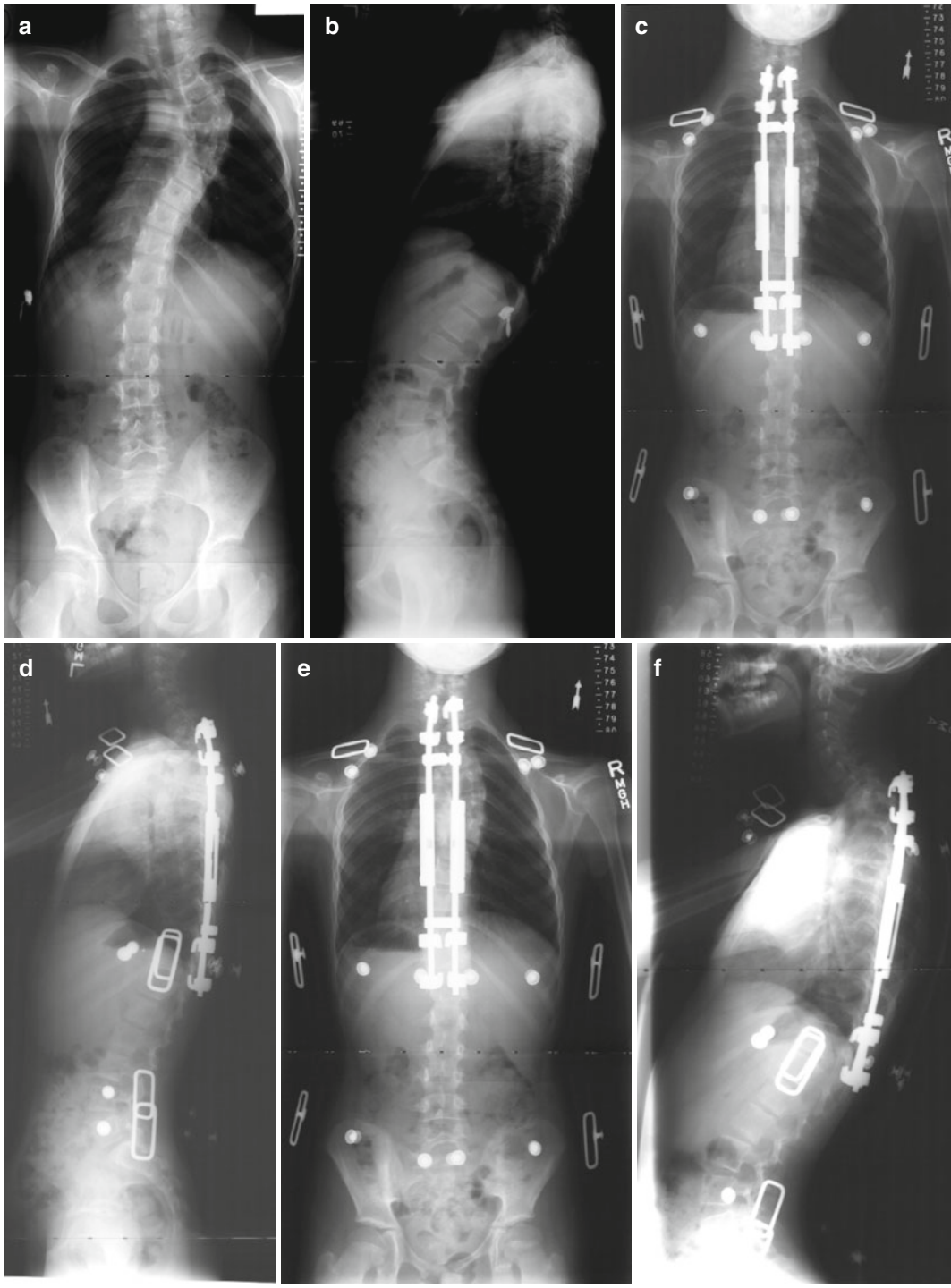


Fig. 16.7 (**a**, **b**) A 7-year-old female with high thoracic dystrophic scoliosis. The brace is usually ineffective in controlling the high thoracic curves. (**c**, **d**) The patient underwent a growing rod instrumentation with brace. Decent correction of the curve was achieved with the index procedure. Note that the proximal hook is at T1. (**e**, **f**) After 1-year postoperation, the correction is well maintained after two lengthenings. On the lateral x-ray, gradual development of junctional kyphosis is evident at both proximal and distal end instrumented segments. Patient is asymptomatic at this point in time. (**g**, **h**) The proximal instrumentation was extended to C7 with supralaminar hooks, which pulled out 2 years after surgery. The instrumentation was then extended to C5 with fusion. (**i**, **j**) Final fusion was performed 5 years after the index procedure

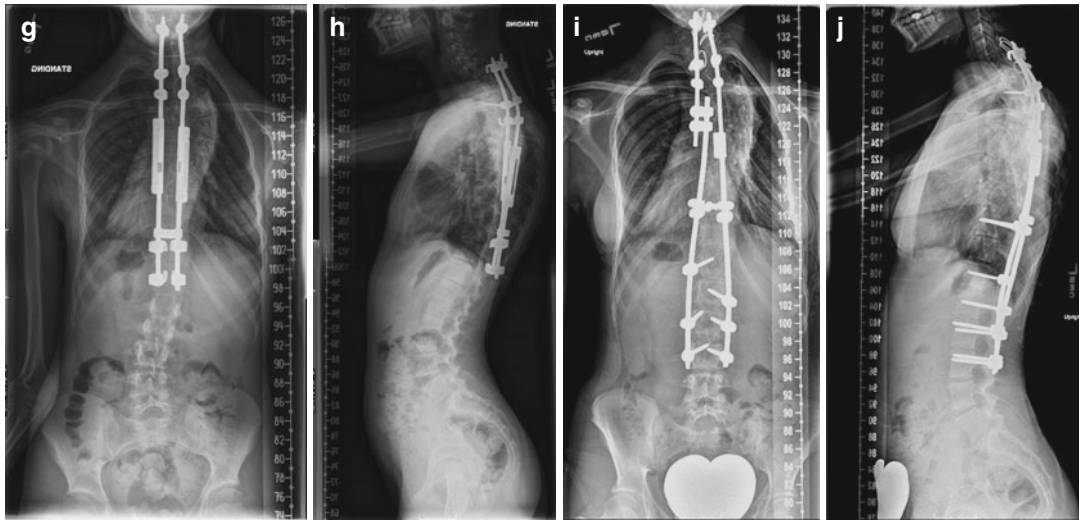


Fig. 16.7 (continued)

It is well known that despite apparent solid fusion, some dystrophic curve shows progression. This tendency is more noted in patients with kyphosis (>50°). The vertebral subluxation, disk wedging, and dystrophy of peripheral skeleton are other factors associated with progression of the deformity after fusion [64].

16.5.2.2 Treatment

The treatment of non-dystrophic curvatures is very similar to idiopathic scoliosis. The curve of less than 25° should be observed. Curves between 25° and 40° can be treated with brace successfully [35]. Once beyond 40°, surgery by posterior spinal fusion is usually indicated [65]. Curves >55°–60° are treated with anterior release with bone-grafting, followed by an instrumented posterior spinal fusion [49]. This is necessary because the curve is usually more rigid than is a similar-sized curve in idiopathic scoliosis. We recommend postoperative orthotic immobilization, although others have managed these patients without postoperative immobilization, with good early results [29].

Dystrophic curvatures of less than 20° should be treated by observation. Serial spinal radiographs at 6-month intervals should be obtained to check for progression of the deformity [49]. Bracing of progressive dystrophic curvatures is ineffective and surgery is usually recommended [10, 35, 66]. For adolescent patients with dystrophic curvature greater than 20°–40° of angulation, a posterior spinal fusion with segmental spinal instrumentation is recommended [10, 62]. In more severe dystrophic scoliosis, anterior fusion should be performed in addition to posterior fusion, to increase the fusion rate, and to reduce the risk for progression despite solid posterior fusion. Preoperative halo traction may be beneficial for the treatment of severe curves, including those with kyphoscoliosis [10, 58, 67, 68]. It allows gradual and controlled soft tissue relaxation and curve correction before surgery or between staged surgeries; however, it is contraindicated in patients who have cervical kyphosis. Daily neurological evaluations are mandatory to avoid spinal or cranial nerve injuries. Nutrition is also paramount during this time. We use supplemental nasojejunal feeding in between stages to decrease the protein depletion that is seen in staged patients [34, 69]. We recommend anterior release, nasojejunal tube alimentation, and craniofemoral traction for rigid curves of >90°. For curves >100° in any plane, anterior as well as posterior release followed by nasojejunal tube alimentation and craniofemoral traction is recommended (see Fig. 16.6).

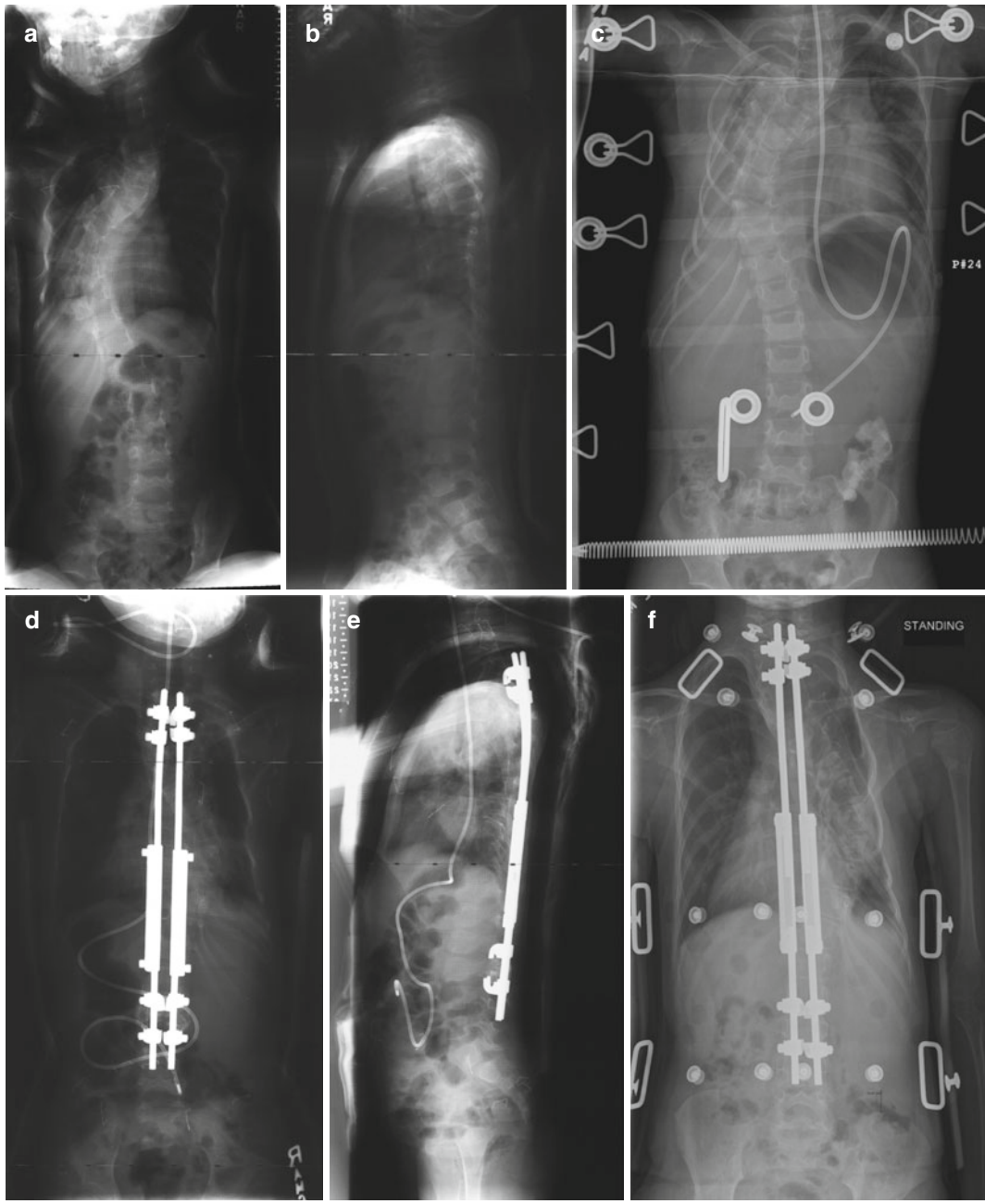


Fig. 16.8 (**a**, **b**) A 4-year-old female with thoracic dystrophic scoliosis who failed cast-brace treatment. (**c**–**e**) She underwent anterior annulotomies at the thoracic apex by thoracoscopic procedure followed by traction for 10 days. This was followed by growing rod instrumentation. (**f**, **g**) At 2-year follow-up, the correction has remained stable and spinal length has increased following two lenghtenings. Development of proximal junctional kyphosis at this point in time is asymptomatic. (**h**) Continued junctional kyphosis lead to prominent hooks. (**i**, **j**) Eight years after the index procedure, the patient had menses and she underwent final fusion with exchange of all instrumentation (4.5–5.5 system with transitional rod) with proximal extension to C6. Proximal junctional kyphosis has been corrected satisfactorily. The thoracolumbar spine was solidly fused due to prolonged immobilization by the growing rods and did not require any further anchors

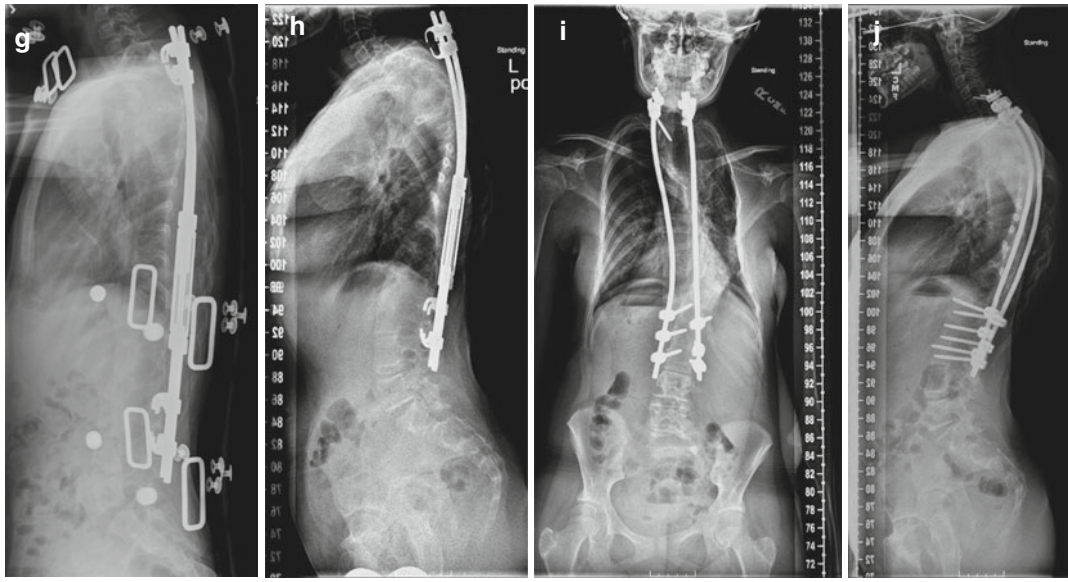


Fig. 16.8 (continued)

The dystrophic curves that are present in late juvenile and early adolescent period pose a challenge to the surgeon. These curves have a high rate of pseudoarthrosis following a posterior spinal fusion [49, 61, 65]. A combined anterior and posterior spinal fusion has been recommended in these patients to decrease the rate of pseudoarthrosis and crank-shaft [70–73]. We also recommend the use of segmental instrumentation to reduce the rate of curve progression after arthrodesis. In our experience, an early fusion of the spine in this age group does not significantly alter the final height and its benefit outweighs the risk of severe progression. Furthermore, the dystrophic segments have very limited growth potential to begin with [32]. The incidence is higher in the presence of kyphosis of more than 50°. It is suggested that the primary reason for fusion failure is an inadequate anterior procedure [74]. However, erosion from enlarging neurofibromas, dural ectasia, and meningoceles may play a role. The best results are obtained when a preplanned combined interverte-bral fusion and posterior arthrodesis is performed. Despite the circumferential arthrodesis, solid fusion is not obtained in every patient, and some patients require repeat operative procedures [74].

Dystrophic curves in infants, toddlers, and early juvenile patients present even more of a challenge. In this age group, a spinal fusion can certainly have a significant effect on overall height as well as the size of the thoracic cage. Smaller size of the vertebrae can pose difficulty in the instrumentation. On the other hand, progression of the curve itself can significantly distort the thoracic cage which can lead to cardio-thoracic decompensation.

The current state-of-the-art treatment for significant deformity or progressive deformities in the patient achieving skeletal maturity is combined anterior and posterior spinal arthrodesis. Most centers recommend observation initially for spinal deformities to determine whether or not it will progress. If the child is very young (under 5–6 years), a corrective cast or bracing may be

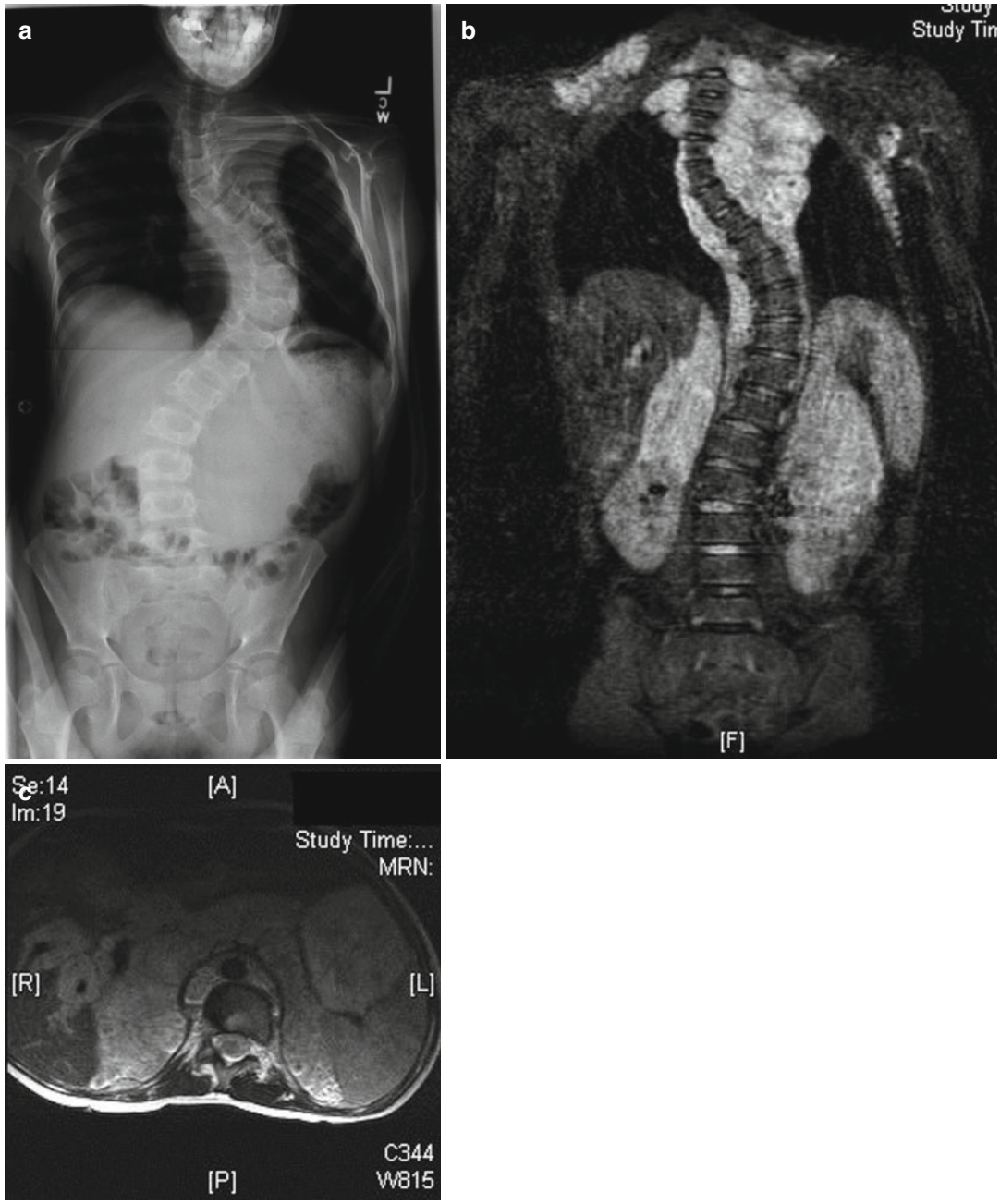


Fig. 16.9 (**a**–**c**) A 9-year-old female with dystrophic thoracic scoliosis. The MRI examination shows extensive involvement of the thoracic cavity with the tumor. (**d**, **e**) This patient underwent a single-stage growing rod instrumentation. Since the posterior elements were involved by the tumor, pedicle screws were used as an anchor point for the growing rods. At 5-month follow-up, the correction is well maintained and there are no complications. Note the presence of sublaminar wires and hooks to augment pedicle screw fixation

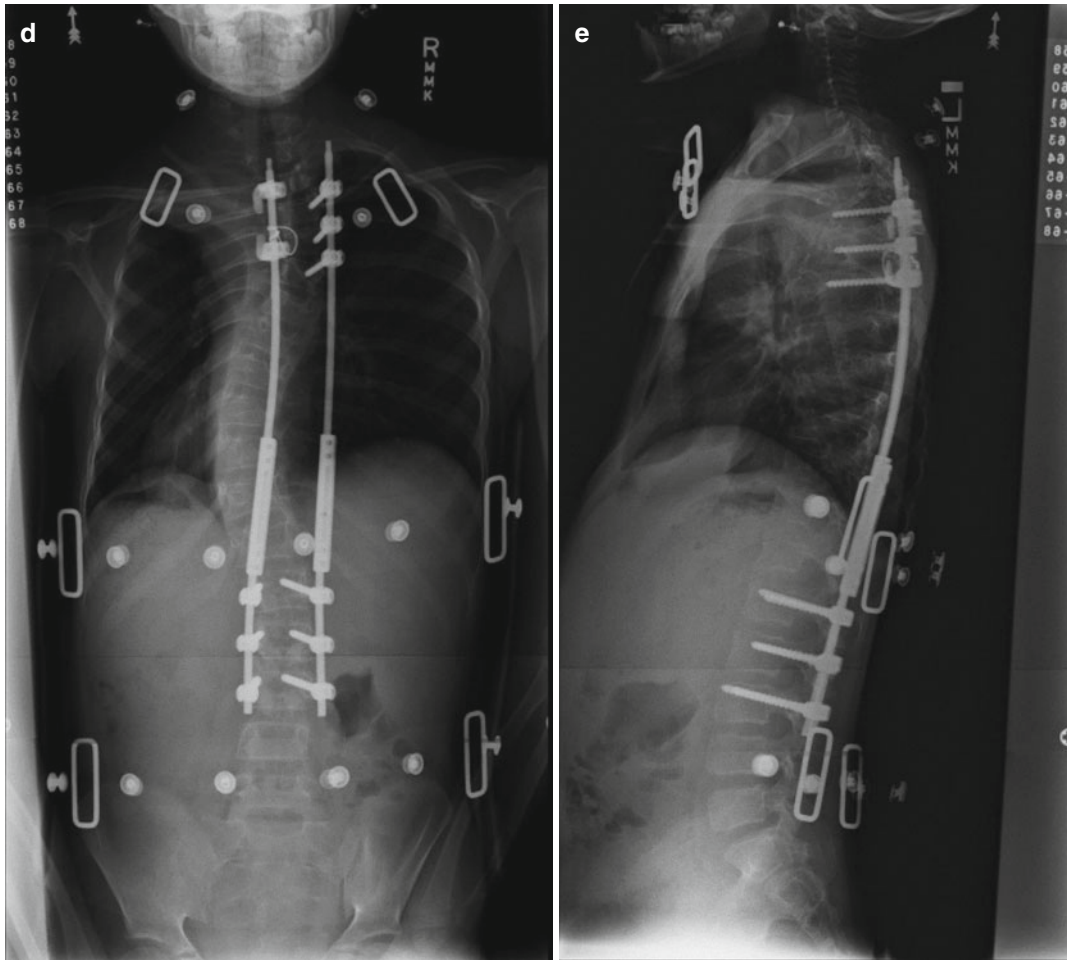
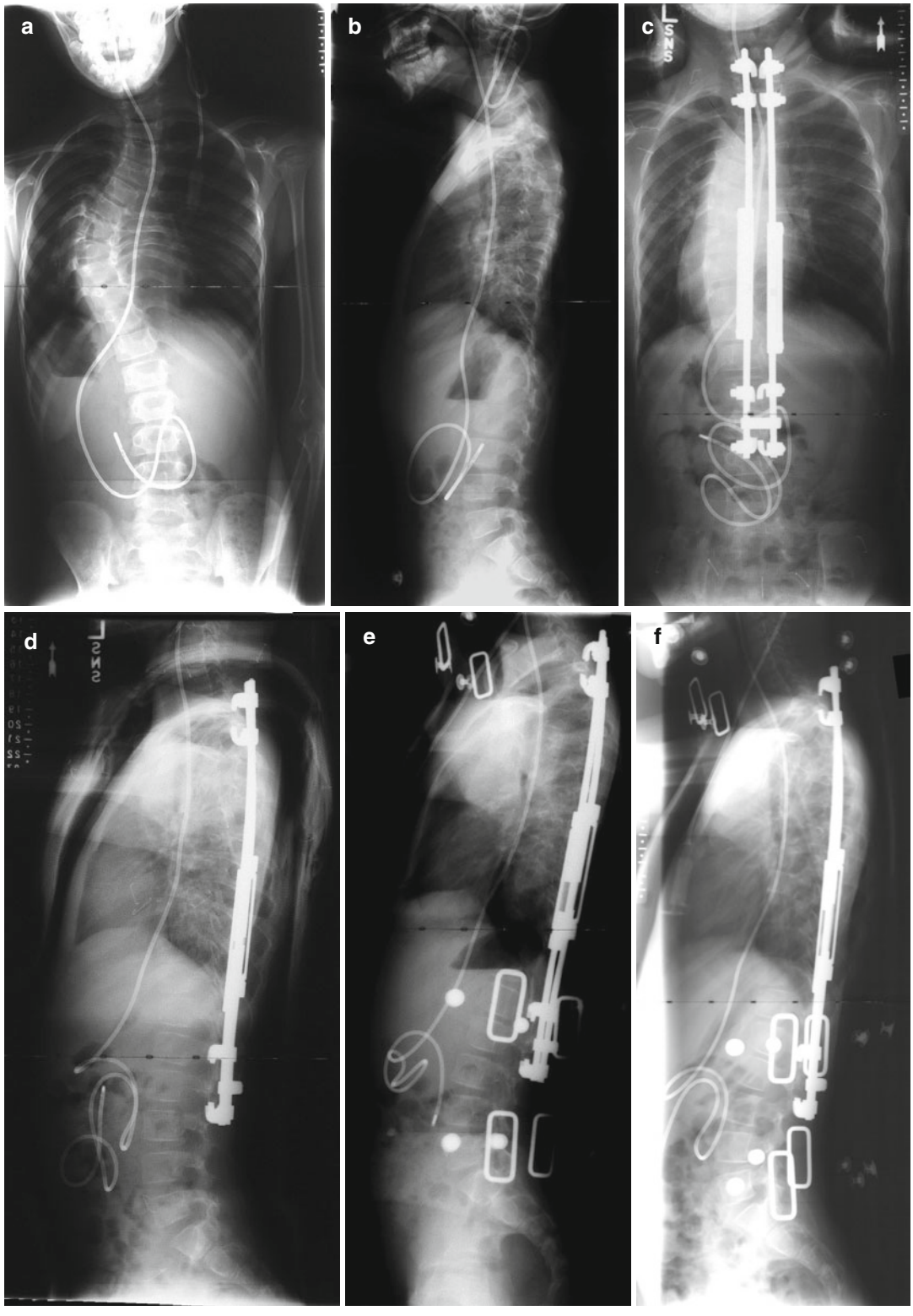


Fig. 16.9 (continued)

Fig. 16.10 (**a**, **b**) This is a 8-year-old male patient with thoracic scoliosis and dystrophic NF1. Patient also had feeding difficulty requiring preoperative alimentation to increase his BMI. (**c**, **d**) He underwent a growing rod instrumentation. (**e**–**g**) During the subsequent follow-ups after a few lengthenings, he was noted to have proximal junctional kyphosis with progressive pulling out of the proximal hooks. He therefore was revised at his proximal end and the anchor point was moved more distal leaving his kyphosis alone. (**h**, **i**) Following his last lengthening, he developed an abscess at his proximal anchor site which could be attributed to his poor nutritional state. This was treated by removal of the anchor, washout, and reinsertion of the anchors. Patient responded well to this treatment and is currently asymptomatic. At 2-year follow-up, his correction is well maintained, although the increase in spinal length is negligible. (**j**, **k**) The hook anchors proximally were eventually changed to pedicle screws due to pullout. Seven years after the index procedure the patient is skeletally mature, there has been no change in implant positioning and curve magnitude. The family has elected to keep the growing rods in as permanent instrumentation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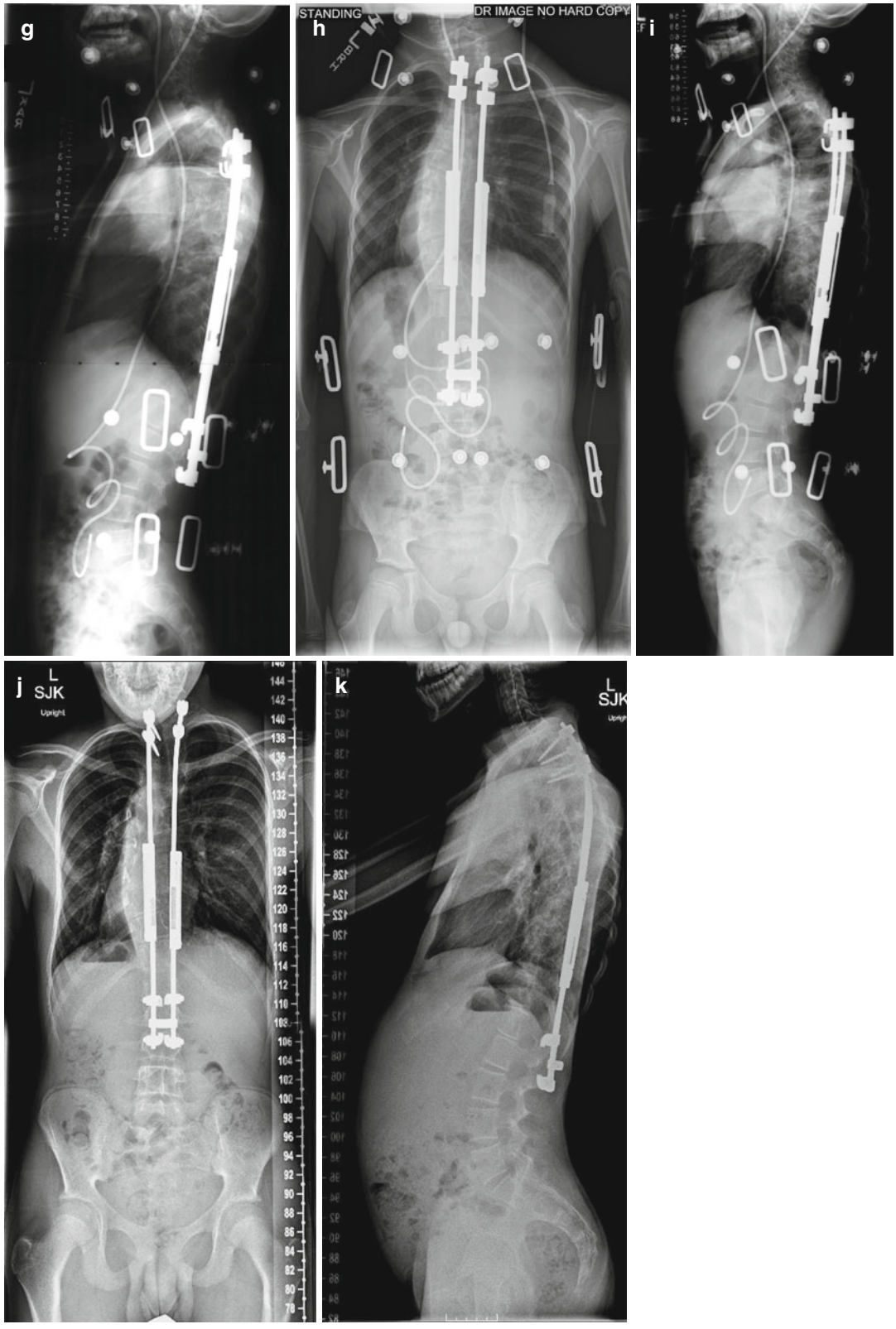

Fig. 16.10 (continued)

attempted, most often with little to marginal success. However, it may allow the surgeon to buy some time. Growing rods have been used to obtain correction without definitive fusion and to lengthen or "grow the spine" every 6 months, but with varying success and a high rate of complications. None of the major spine centers have attempted modulating the growth of the spine with convex side tethers using staples or synthetic constraints to date.

16.5.2.3 Growing Rod Instrumentation

The growing rods have been used successfully in the treatment of early onset idiopathic curves. These devices have been shown to prevent the progression of the curve while preserving the longitudinal growth of the spine [75]. The currently available dual growing rods have been shown to be superior to the previous versions of submuscular single growing rods [76]. We have used dual growing rods on early onset dystrophic curves with a great deal of optimism [62].

We have used the growing rods directly with fusion of the cranial and caudal anchors only in the patients with flexible curves less than 60° (see Fig. 16.7). Traditionally, this is followed by a period of bracing and lengthening every 6 months. In larger and stiffer curves, we recommend anterior annulotomies (with or without thoracoscope) without fusion to preserve growth (see Figs. 16.6 and 16.8). Annulotomies should be performed with Bovie dissection and the use of a thin rongeur through the annulus fibrosis instead of sharp dissections of the endplate apophysis. Sharp dissection may cause significant bleeding from the often friable cancellous matrix of the vertebral bodies. Care should be taken to preserve the segmental vessels as much as possible. This is followed by insertion of the growing rods and routine lengthening at 6-month interval. In certain cases, traditional use of hooks as anchor point may not be feasible (see Fig. 16.9). In these cases, use of pedicle screws as anchor points in the spine is advantageous.

The use of growing rod instrumentation in NF-1 is also associated with a high incidence of complications. The high rate of complications has also been reported for idiopathic patients [75]. The most common complication we have encountered is proximal junctional kyphosis. This is especially common in the patients with high thoracic or cervicothoracic curves (see Figs. 16.7, 16.8, 16.10, and 16.11). We believe that it is the result of excessive stress put on the proximal anchors by routine lengthening. This abnormal stress results from the difficulty of applying adequate proximal kyphosis to the rods above the lengthener because of not enough length. The subsequent lengthening drives the rod directly vertical as opposed to physiologic mechanics. The proximal anchor places a vertical load on the lamina forcing hinging of the vertebra into kyphosis. This is our experience with proximal hook capture systems. An all screw construct may be less problematic. In these patients, we currently do not perform routine lengthening. Other complications encountered are infection (see Fig. 16.10) and rod breakage (see Fig. 16.6).

Although the use of growing rod instrumentation is associated with higher complication rate, its benefits outweighs the risk in patients with early onset dystrophic scoliosis. Our early results with the use of growing rods remain encouraging. This is a promising technique made especially useful because most dystrophic curves are early onset.

16.5.2.4 Trapdoor Procedure

A few of the NF-1 patients develop very high thoracic curve extending in the cervicothoracic junction. These patients need circumferential fusion and instrumentation in the lower cervical and upper thoracic spine. This group of patients may benefit from a "trap door" sternal split approach if anterior fusion is needed (see Fig. 16.6) [71, 77]. This approach allows anterior exposure of the lower cervical and upper thoracic spine. Bracing may need to be extended to the cervical region in cases of severe dysplastic curves that are instrumented into the upper thoracic and cervicothoracic region. Cervical bracing, halo vest, or Minerva casting may help to prevent the possibility of screw/hook pullout. This is especially true for dystrophic curves that have low bone mineral density [7].

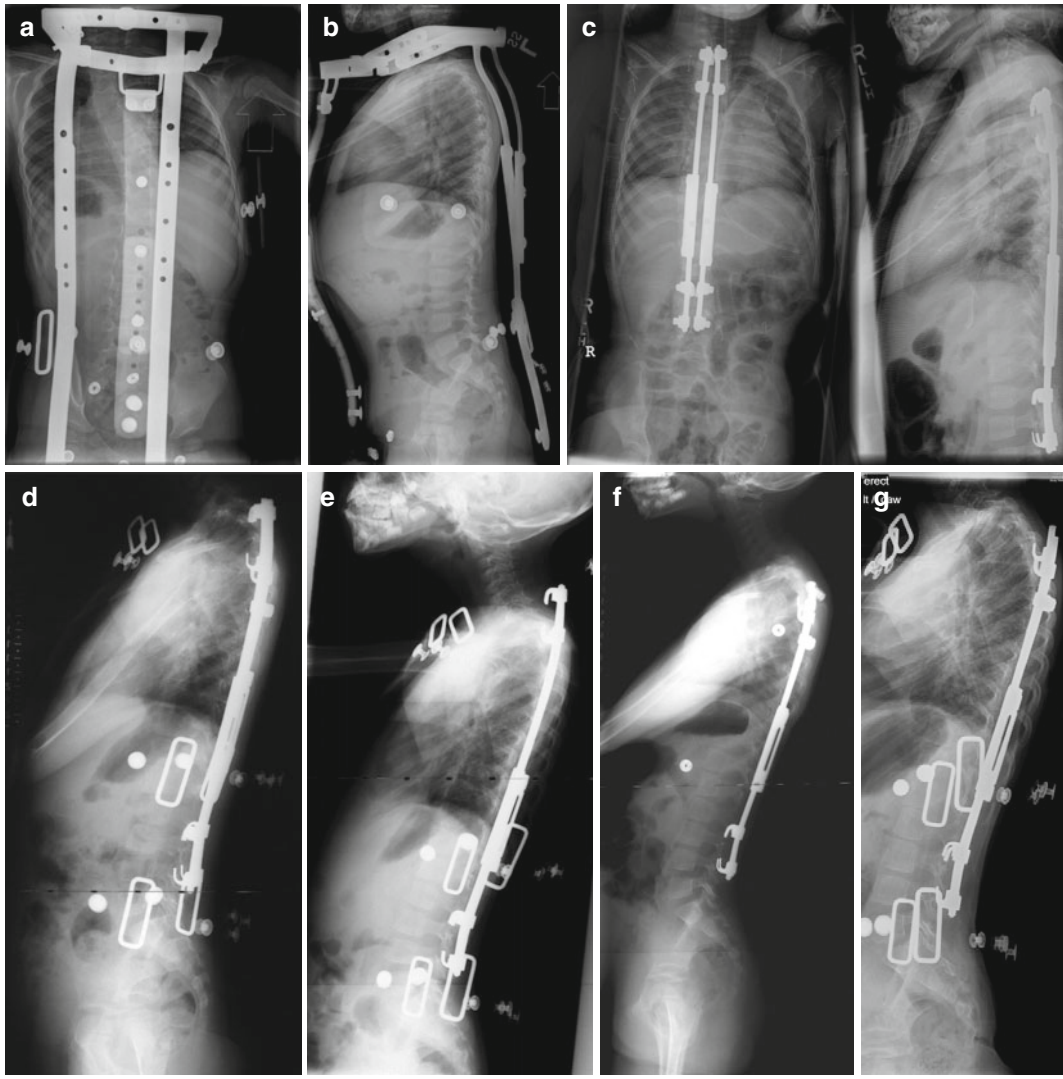

Fig. 16.11 (**a**, **b**) A 6-year-old female with thoracic dystrophic NF1. Patient was earlier treated in a Risser cast followed by a CTLSO for two cycles. Despite in cast and brace, her curve progressed. (**c**) She ultimately underwent a growing rod instrumentation. (**d**, **e**) Following first few lengthenings, she was noted to develop proximal junctional kyphosis with gradual pulling out of her top hooks. There are two options at this time. First is to extend the instrumentation to the cervical spine or (**f**) second to extend the instrumentation down one segment in order to allow the junctional segment to remain free from the stress and prevent further kyphosis. (**g**–**i**) The patient developed further kyphosis resulting in the pulling out of the proximal hook. At that point in time, her entire proximal construct was revised and she was given a CTLSO. After a total of 3 years of follow-up, her curve remains stable and there are no further complications at this time. Furthermore, the patient has shown an increase in her spine length as measured on the digital x-rays. (**j**, **k**) Due to continued proximal anchor failures, these were changed to pedicle screws. (**l**, **m**) Final fusion was performed 7 years after the index procedure with rod exchange. Almost complete fusion was noted at the time of surgery at the segments between the anchors. The sublaminar instrumentation was used at the apex as there were extremely narrow pedicles available secondary to progressive dural ectasia

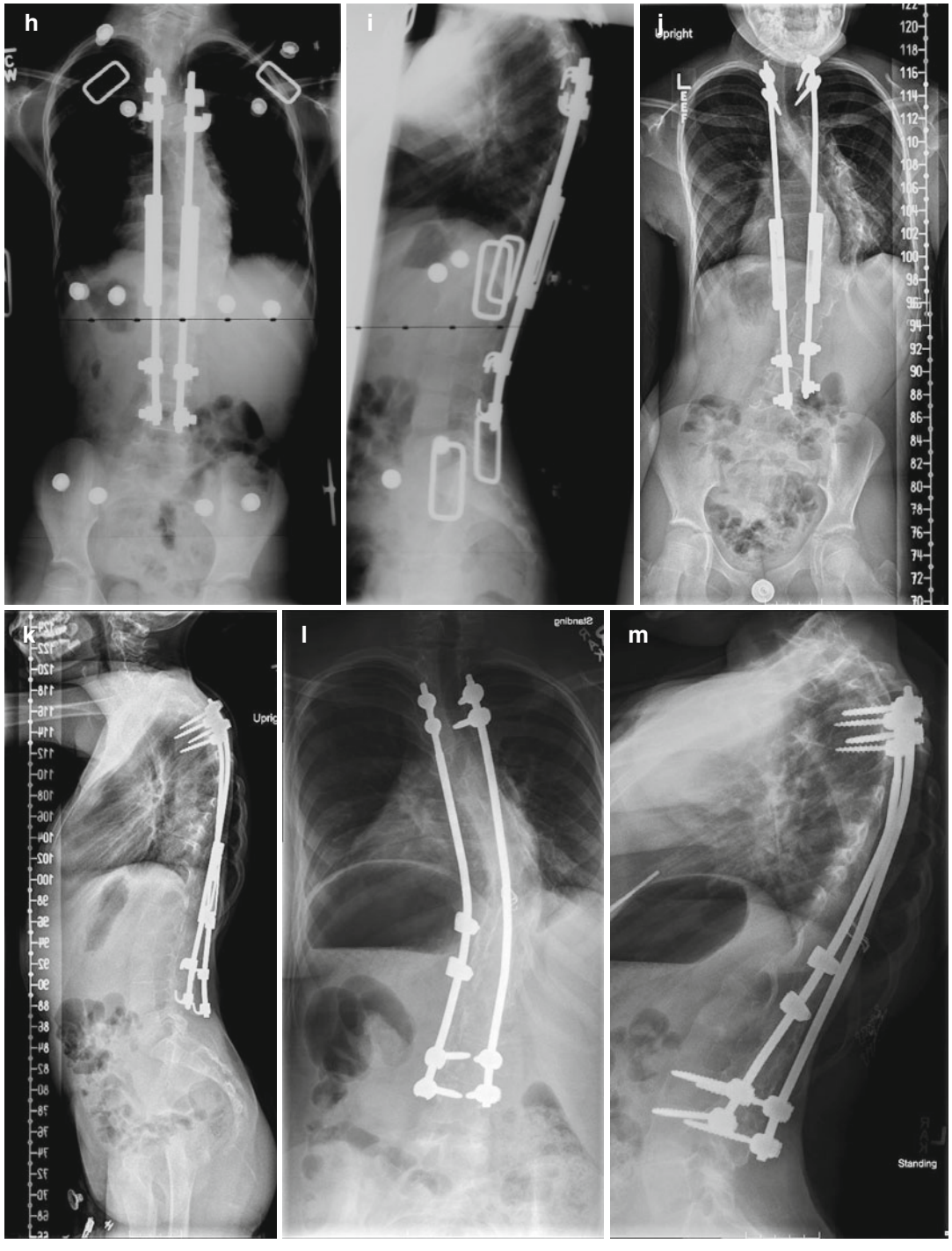

Fig. 16.11 (continued)

16.6 Other Spinal Deformities

16.6.1 Kyphosis

Kyphoscoliosis is defined as scoliosis accompanied by a kyphosis of greater than 50°. It may occur by gradual scoliotic rotation and progression or it can be found early in the disease with an abrupt angular kyphotic curve [78]. Vertebral bodies may be deformed so severely that they are confused with congenital deformities. Severe kyphosis is the most common cause of neurological deficits in NF-1 [62]. Use of traction in patients with rigid and severe kyphosis can increase the tension on the spinal cord leading to neurological deficits. Traction following anterior release is safe when monitored appropriately. For curves greater than 50°, anterior surgery (release and fusion) is recommended, followed by posterior segmental instrumentation one or two levels above and below the end vertebrae [32, 49, 58, 64]. Assessment of the fusion mass by CT at 6 months postoperatively is recommended. If pseudarthrosis is noted, augmentation of the fusion mass is indicated.

We recommend that the anterior procedure should be undertaken from the convex side of the deformity, since the exposure is extremely difficult from the concave side [79]. The anterior fusion should include the entire structural area of the deformity with complete disk excision and local strut grafting. Multiple grafts or cages should be placed into the vertical weight-bearing axis of the torso, with the strong autologous fibula or rib graft placed more anteriorly [62, 66]. Strut grafts should have contact with each other and with the vertebral body to prevent resorption noted when graft material is surrounded by pathological tissue. Anterior release and fusion should be followed by posterior instrumented fusion using a large amount of autologous iliac crest bone graft and BMP in selected cases.

16.6.2 Lordoscoliosis

Lordoscoliosis has not been so frequently reported in patients with NF-1 compared to kyphoscoliosis (see Fig. 16.6). However, lordosis of the thoracic spine predisposes to significant respiratory compromise and mitral valve prolapse [77, 80].

Anterior release and intervertebral fusion followed by posterior instrumented fusion is considered as the most reliable surgical option to achieve correction of dystrophic lordoscoliosis [32]. Sublaminar wires, pedicle screws, or rod-multiple hook constructs can be used.

16.6.3 Spondylolisthesis

Spondylolisthesis in patients with NF-1 is rare. It is characterized by pathological forward progression of the anterior elements of the spinal column. Spondylolisthesis in patients with NF-1 is most often associated with pathological elongation and thinning of the pedicles or pars interarticularis by lumbosacral foraminal neurofibromas or dural ectasia with meningoceles [32]. The vertebral bodies may also be small and dystrophic. MRI and/or CT scan are absolutely necessary for preoperative evaluation.

Fusion may also be delayed because of the forward traction effect of the vertebral bodies and the slow healing and remodeling of bone in NF-1. We recommend a combined anterior and posterior fusion from L4-to-sacrum using intervertebral body grafting and lumbosacral instrumentation. Postoperative immobilization is indicated until the fusion is absolutely solid.

Conclusion

NF-1 is the most common human single-gene disorder. Skeletal complications usually present early in life and can be attributed to abnormalities of bone growth, remodeling, and repair in NF-1 or can be secondary to nearby soft-tissue abnormalities associated with NF-1. Scoliosis is the most common osseous manifestation of NF-1. It is important to recognize the dystrophic curve and to distinguish it from the non-dystrophic curve.

The management of spinal disorders in young children in NF-1 continues to be problematic. The use of growing rods allows more longitudinal growth than fusion and more life freedom than bracing. The problems we have

encountered are mechanical and could be expected when proximal and distal fixation is performed over an otherwise completely mobile spinal column. The multiple surgeries increase the potential for complications including infections. We continue to pursue solutions to our problems.

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The Growing Spine in Marfan and Loeys–Dietz Syndromes

17

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Key Points

- Bracing for scoliosis in Marfan syndrome may be best instituted before curves reach 25°.
- Patients with Marfan syndrome should have cardiac clearance within 6 months of surgery.
- Patients with Loeys–Dietz syndrome should have periodic examinations of the entire spine, including the neck.

17.1 Introduction

The following two chapters will review a set of unique congenital syndromes that have a high association with scoliosis due to their effect on the connective and neurological systems. There are some general principles that govern the management of spinal deformities in syndromes. These syndromes have wide reaching, systemic manifestations with the potential to cause significant morbidity and mortality if not diagnosed. Since musculoskeletal manifestations are often the most conspicuous, many of these patients may first present to an orthopedic physician. Therefore, it is incumbent upon orthopedic surgeons be alert and knowledgeable about the unique diagnosis, referral, and management of these patients. This chapter reviews Marfan syndrome and Loeys–Dietz syndrome

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(LDS). The following chapter will review Schprintzen–Goldberg, Ehlers–Danlos syndrome, Prader–Willi, Rett, and Down syndromes.

17.2 General Principles of Syndromic Deformity Management (Table 17.1)

17.2.1 Role of Nonoperative Management

Patients with syndromic disorders often present with significant curves at a young age. Therefore, they often require more than simply orthotic treatment or spinal fusion near the age of maturity. Syndromic curves can present as early as infancy. There are virtually no studies showing efficacy of orthotic treatment in syndromic curves. In addition, D'Astous and Sanders have shown that Mehta casting is less effective for infants with scoliosis due to syndromes than it is in idiopathic infantile scoliosis [1]. Practically, bracing is commonly recommended in young syndromic patients with curves between 35° and 50°. Orthotic treatment may show in-brace correction but there have been no studies that document an improvement in the expected natural history. The physician should consider refraining from overzealous application of bracing for large curves (over approximately 50°) in young patients at the expense of quality of life.

17.2.2 Comprehensive Evaluation

Another principle of treating patients with syndromic curves is that the whole spine is at risk of developing differently. The cervical, thoracic, and lumbar spine should be examined and imaged as indicated. "Coned" films (focal images centered on area of interest) should be obtained of any area that requires further definition. In addition, there is a greater chance of abnormality of the neuraxis. For this reason, whole-spine magnetic resonance imaging (MRI) should be considered prior to surgical intervention. Findings such as duralectasia, stenosis, spondylolysis, instability, and disk pathology are more likely to be seen than in idiopathic deformity. Computed tomography with multiplanar or three-dimensional reconstruction may be invaluable in defining dystrophic bony features, if suspected. Flexibility is often best assessed with traction films rather than bending films in young patients with syndromes, due to age and curve magnitude.

17.2.3 Medical Considerations

Patients mature at different rates; skeletal maturation in syndromic patients may be earlier or

Table 17.1 General spinal considerations in patients with syndromes

Role of nonoperative management	Curves become large at a young age Brace early or not at all; avoid in low-yield situations
Comprehensive imaging	Image entire spine MRI before surgery in most cases CT to define abnormal bony anatomy Traction films more useful than bending
Medical considerations	Communicate with specialists Consider genetic consultation Consult OMIM for information (Online Mendelian Inheritance in Man) Assess nutrition, respiratory status
Operative considerations	Blood loss likely more Bone density often lower Have appropriate size implants Failure of fixation more likely Do not "fuse short" in syndromes Have ICU available postoperatively Consider rehab needs postoperatively

later than in idiopathic deformity. Medical comorbidities are more often seen in syndromic patients. The surgeon should take advantage of pediatric consultants in genetics, pulmonary, and cardiology specialties. A geneticist can be a great help both preoperatively and postoperatively in managing patients with syndromes, tying all of the disparate features together. A good source of genetic information is Online Mendelian Inheritance In Man (OMIM), rapidly available to all on the Entrez Pub Med series of applications. This site allows one to search for diagnoses by listing a series of physical findings. A set of matches, discussion, and references will appear.

Specialists can also provide helpful input in determining the proper role of surgery for a given patient. It is helpful to ask, "what other specialists are you seeing?" so that the orthopedic management plan can be integrated with that of other specialists. Testing prior to surgery may also include echocardiography or sleep study for patients at risk of cardiac or pulmonary difficulties. Specific cardiovascular manifestations of specific syndromes will be discussed further in the subsequent sections. Finally, nutritional and gastrointestinal issues can affect this group of patients as well. Specifically, severe curvatures can cause abdominal compression, including gastric reflux. At least one case of postoperative superior mesenteric syndrome has been reported in literature following scoliosis surgery in a patient with Marfan syndrome (MFS) [2]. Malabsorption has been reported in Ehlers–Danlos syndrome (EDS) and MFS secondary to bacterial overgrowth in large jejunal diverticula. Feeding problems and indigestion have been reported in most syndromic scoliosis disorders. The authors recommend a gastrointestinal consultation if these issues come to light.

17.2.4 Operative and Postoperative Management

Implant size may be a problem in young patients with poor nutrition, such as infantile Marfan patients. A range of implant diameters should be available. Osteopenia and increased blood loss may affect surgery. Failure of fixation is another complication commonly seen in syndromic patients following spinal instrumentation. The number and types of anchors should be chosen to minimize this risk. Because of balance and connective tissue factors, principles of instrumentation and fusion that apply to idiopathic patients may not apply to syndromic patients. Attempts to "save levels" do not always work as predictably as in idiopathic deformity. Patients with syndromes often require more involved postoperative care and intensive care stay may be appropriate. Return to function may also be significantly slower than in idiopathic patients. Inpatient rehabilitation may occasionally be indicated after discharge from hospital.

17.3 Specific Syndromes

17.3.1 Marfan Syndrome

MFS is one of the most common connective tissue disorders. It is characterized by its classical involvement of the skeletal, ocular, and cardiovascular organ systems due to its effect on microfibril formation.

17.3.1.1 Etiology/Genetics

MFS has been linked to heterozygous mutations of the *FBN1* gene on chromosome 15 that encodes the fibrillin-1 protein, which undergoes polymerization to produce micofibrils [3]. Therefore, fibrillin-1 is an essential component of elastic connective tissue. Additionally, fibrillin-1 plays a role in transforming growth factor beta (TGF-β) binding by stabilizing latent growth factor β-binding proteins, which hold TGF-β in the inactivated state [3]. It was previously hypothesized that the mutations in FBN1 cause structural abnormalities in the microfibrils that lead to the Marfan phenotype; however, it is now accepted that the faulty regulation of TGF-β by fibrillin-1 may be the dominant mechanism [4].

Family history of MFS should be considered significant, as it is a heritable disorder, however it has been approximated that up to 27 % of cases arise from a de novo mutation [4].

17.3.1.2 Presentation/Diagnosis

A diagnosis of MFS may be considered in a young patient who is tall and thin, with long arms and digits, pectus deformities, scoliosis, and other skeletal features [5] (see Table 17.2). The Ghent Nosology is the most widely accepted diagnostic criteria. A Revised Ghent Nosology was published in 2010 [6]. Under the new criteria, cardiovascular manifestations of MFS receive greater attention and, as demonstrated in Table 17.3, aortic root aneurysms and ectopialentis are considered principle features for diagnosis. When no family history is noted, a combination of aortic root aneurysms and ectopialentis are sufficient to establish a positive diagnosis. If these features are not present, demonstration of a FBN1 mutation or a number of systemic features (see Table 17.2) may establish the diagnosis [6]. Under the Revised Ghent Nosology by Loeys et al. [6], a new scoring system has been devised to and is summarized in Table 17.3.

Notable exceptions to Table 17.3 include when a patient presents distinguishing features that suggest LDS (described in this chapter), Shprintzen–Goldberg syndrome, or EDS (described in this chapter that follows). These syndromes often demonstrate significant overlap with MFS. Additionally, molecular testing for TGFBR1/2, collagen biochemistry, and COL3A1 may be required before a positive diagnosis of MFS can be made [6].

Table 17.2 shows the scoring system for systemic features, as defined for the Revised Ghent Nosology [6]. A score of ≥7 is required to fulfill the criteria for systemic involvement.

The early assessment of patients with a suspected diagnosis of MFS should include a detailed personal and family history, due to the age-dependent and heritable patterns of this disorder. Furthermore, an ophthalmological examination and transthoracic echocardiogram are warranted to identify ocular and cardiovascular criteria. Molecular genetic testing may be indicated; however, testing for FBN1 mutations has not proven definitive by itself due to the fact that FBN1 mutations may not be unique to MFS, and 5–10 % of patients may not demonstrate mutations via current testing methods [7–9].

Table 17.2 Systemic features scoring for Marfan syndrome

Scoring for systemic features in Marfan syndrome	
Feature	Points
Combined wrist and thumb sign	3
Wrist or thumb sign	1
Pectus carinatum deformity	2
Pectus excavatum or chest asymmetry	1
Hindfoot deformity	2
Plain pes planus	1
Pneumothorax	2
Dural ectasia	2
Protrusio acetabula	2
Reduced upper segment/lower segment ratio + increased arm/height + no severe scoliosis	1
Scoliosis or thoracolumbar kyphosis	1
Reduced elbow extension	1
Facial features (3/5): olichocephaly, enophthalmos, down-slanting palpebral fissures, malar hypoplasia, retrognathia	1
Skin striae	1
Myopia >3 diopters	1
Mitral valve prolapse	1

Table 17.3 Diagnosis of Marfan syndrome

No family history	Family history
1. Aortic root dilation *Z score* ≥ *2* + *Ectopia lentis*	1. *Ectopia lentis* + *family history* of Marfan syndrome
2. Aortic root dilation *Z score* ≥ *2* + *Causal FBN1 mutation*	2. *Systemic features score* ≥ *7* + *family history* of Marfan syndrome
3. Aortic root dilation *Z score* ≥ *2* + *systemic features score* ≥ *7*	3. *Aortic root dilatation Z score* ≥ *2* (if above 20 years old), ≥ *3* (if below 20 years old) + *Family History* of Marfan syndrome
4. *Ectopia lentis* + *causal FBN1 mutation* + *aortic root dilation*	–

MFS cannot be ruled out in young patients via the Ghent Nosology due to the age-dependent development of its features. If MFS is on the differential diagnosis, children should be kept under clinical review until age 18 or until a positive diagnosis has been made [4].

17.3.1.3 Skeletal/Spine Manifestations

Skeletal manifestations include pectus deformity, spinal deformity (scoliosis, spondylolisthesis, kyphosis, reduced lumbar pedicle, and laminar thickness), joint hypermobility, dolichostenomelia, arachnodactyly, highly arched palate with crowding of teeth, and abnormalities in facial appearance (dolichocephaly, malar hypoplasia, enophthalmos, retrognathia, down-slanting palpebral fissures).

Scoliosis exists in two-thirds of MFS patients. The curve patterns resemble that of idiopathic curves, but with earlier onset, and may be associated with pain in the region of curvature. There appears to be no familial pattern and the scoliosis shows approximately equal prevalence in male and female patients, as opposed to larger idiopathic curves, which tend to be more common in females. Similar to idiopathic causes, scoliosis in MFS tends to demonstrate right-side thoracic curvature and left-side lumbar curvature. The curvature in Marfan infants tends to progress most rapidly, at approximately 20° per year, followed by the curvature of adolescents, which progress at approximately 6° per year (which is not dissimilar to the rate of idiopathic curves during growth spurts). Curves of magnitude greater than 20° are likely to progress during growth and curves greater than 30°–40° tend to progress during adulthood. In general, curves of more than 30° demonstrate mild progression, while curves of more than 50° demonstrate more rapid progression.

The incidence of spondylolisthesis does not appear to be increased. However, the amount of slip tends to be approximately double that of patients without MFS [5]. There is also a tendency for thoracolumbar kyphosis. Dural ectasia, while rare in the general population, is present in at least 60 % of Marfan patients and is often associated with back pain. Patients with duralectasia demonstrate increased rates of bony erosion and anterior or posterior meningoceles [10]. Lumbar pedicle widths and laminar thicknesses tend to be significantly reduced in patients with MFS and may be associated with duralectasia and vertebral scalloping [11]. Additionally, bone mineral density tends to be lower in the spine and pelvis. However, an increase in fracture rate has not been demonstrated [12, 13].

17.3.1.4 MFS and Sports

Patients with MFS are not recommended to participate in high-intensity static exercises, such as weight lifting and hill climbing exercises, due to their effects on blood pressure and vascular resistance. Regular non-strenuous and noncompetitive aerobic activity should be encouraged. Contact sports should be avoided due to risk of damage to the aorta and eyes, and scuba diving should be avoided due to increased risk of pneumothorax [4].

17.3.1.5 Spinal Deformity Treatment and Complications

Bracing for scoliosis in MFS is successful in only 17 % of patients [14]. Approximately one-eighth of patients eventually develop a severe curve requiring surgical intervention [14]. Marfan patients with infantile spinal curvature are a special group (Fig. 17.1). They more often have no family history (spontaneous mutation) and more severe phenotypes. For these patients, rib-based distraction is not widely successful, as it tends to cause progressive kyphosis of the lax spine. Growing rods have documented effectiveness (Fig. 17.1). The authors prefer to delay surgery until at least age 7 if possible, or until curves reach 80°. In the experience of the author, one of two types of constructs can be used. For patients with excessive thoracolumbar kyphosis or severe lower lumbar curves, the distal anchor may be placed in the pelvis (Fig. 17.2). For those with no significant sagittal abnormality, more "typical" growing rod anchors in the thoracic and lumbar spine have been used. It is critical to have an anesthesiologist who is an expert at managing cardiovascular problems, and to have pediatric intensive care and cardiology physicians available. Outpatient lengthening of the growing rods is advised only in the most stable of patients.

When inserting growing rod anchors in this population, the surgeon should be prepared for fixation challenges. The laminae are often thin, creating a risk of hook dislodgement. For this reason, pedicle fixation is preferred if at all possible. However, if hooks are used, a three-level "claw" is recommended [15]. Pedicles are often thin in MFS, especially proximally. Small-diameter

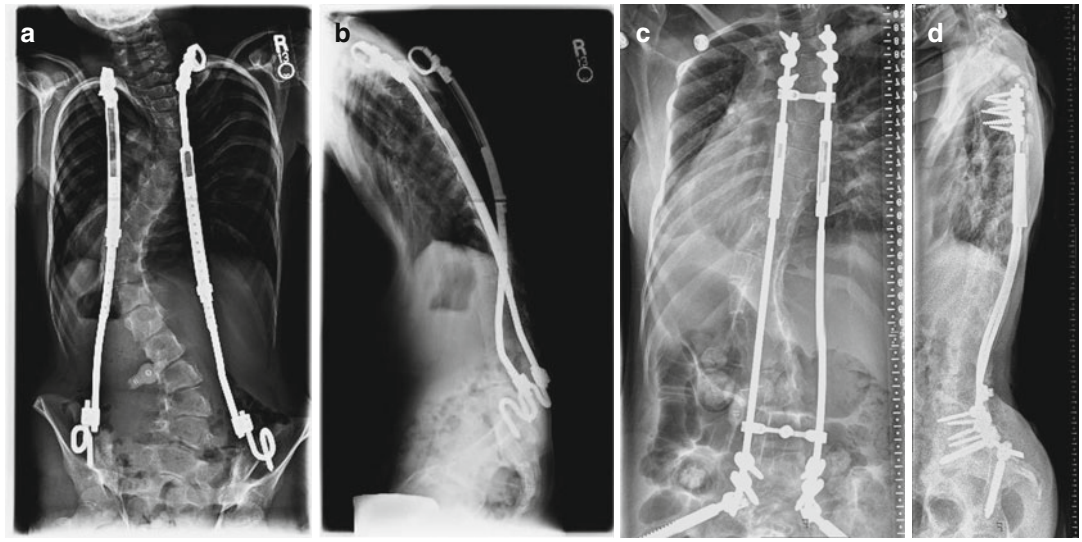

Fig. 17.1 The 11-year-old female Marfan patient previously underwent rib-based distraction for thoracic insufficiency. Progressive kyphosis resulted (demonstrated in images **a** and **b**) and was severe enough to warrant further intervention. The VEPTR were removed and growing rods were inserted with pedicle screws from T2–T4 to pelvis. Recontouring of the lumbar spine was done to maintain lordosis, and sagittal balance. This was aided by the pelvic fixation (images **c** and **d**)

screws and even the use of cervical systems may be necessary. Because of the thin, dysplastic pedicles, the author advises liberal use of image guidance when inserting pedicle screws. Three-level proximal foundations are also advised if pedicle screws are used in dysplastic cases. Submuscular placement of growing rods is recommended due to problems with implant prominence in this population related to the asthenic habitus. It can usually be accomplished in a minimally invasive fashion with two small, midline incisions. Intraoperative leakage of cerebrospinal fluid occurs with a higher frequency in this population. Due to duralectasia, the dura in MFS often expands to fill the entire spinal canal and is extremely thin and fragile. This is often seen when dissecting under the lamina or attempting to cannulate the narrow pedicles. CSF leakage is especially common when dissecting on the sacral lamina, which may be paper-thin or absent. Placing the operating table in 15° Trendelenburg during dissection may help to minimize the risk of dural leak. Fibrin glue is typically used along with sutures to deal with dural leaks. Sometimes the dura is too friable to hold a suture, but patients virtually always respond to a period of postoperative recumbency. Bleeding is also more extensive in the Marfan population.

Postoperative bracing is typically not used in this population. Patients who are on Coumadin are usually lengthened only yearly, in order to minimize the risks of stopping and restarting the anticoagulation. Periodic in-situ recontouring of the lumbar segment of the rods into lordosis is necessary to maintain sagittal balance (Fig. 17.1). This may also require osteotomy of the caudal foundation near maturity. It is not known whether Marfan patients follow the "law of diminishing returns" at the same rate as non-Marfan patients, due to the connective tissue laxity. Final treatment of Marfan spine deformity at the end of growth may be nonsurgical due to spontaneous ankylosis, with the growing rods in place, if the following criteria are met: no recent rod breakage within the past 2 years, acceptable alignment, postmenarchal status for females, and Risser sign of two or greater. If, however, there is a history of recent rod breakage to suggest that the spine is mobile, or the deformity is not well corrected, a traditional

fusion may be appropriate. This may be performed with additional anchors and optimal rod diameter and material to ensure fusion.

Complications of rod breakage and implant dislodgement have been low in the experience of the author. This may be due to the fact that patients

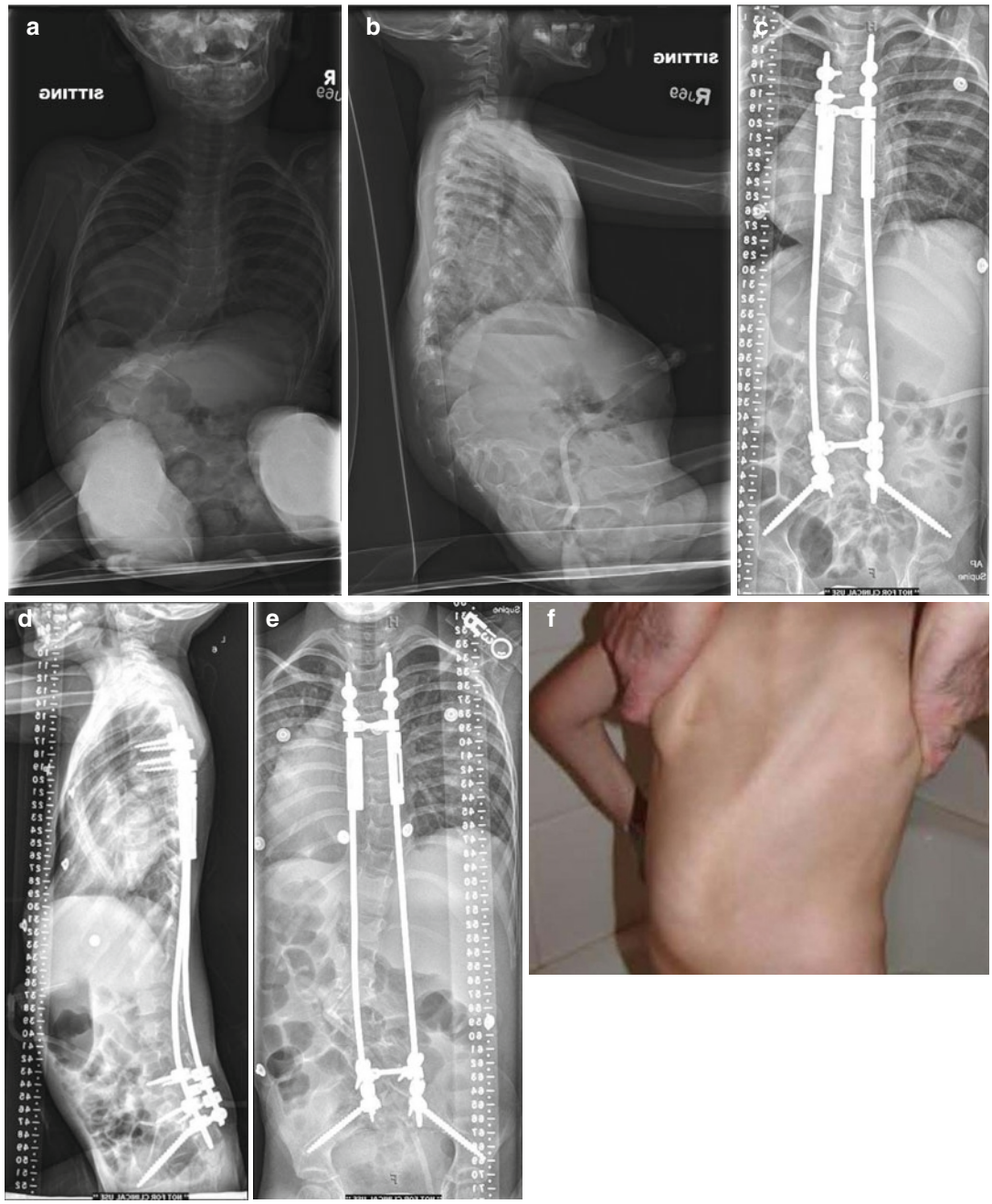

Fig. 17.2 The 3-year-old female Marfan patient with severe kyphoscoliosis (primary cobb angle 95°). Postoperative images are seen in **c** and **d** with a follow-up cobb of 34°, as seen in image **e**. Pedicle screws were used at T3–T4 to the pelvis. The S2 screws were used as distal anchors in the ileum. In addition to radiographs (**a**–**e**) the following clinical photographs (**f**–**i**) further elucidate the effects of growing rod surgery in this patient

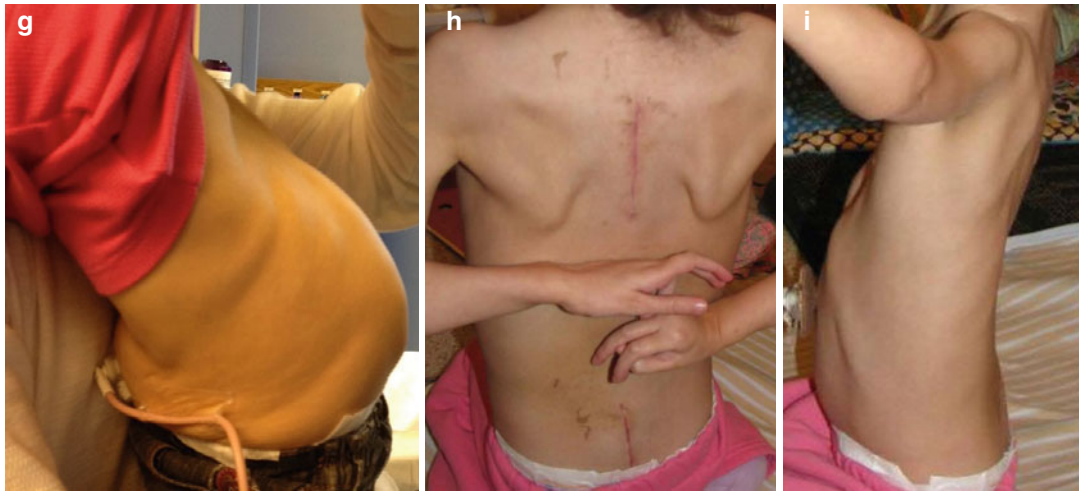

Fig. 17.2 (continued)

are restricted from high impact activities to avoid stress on the aorta or the eye. Aneurysm of the ascending aorta can cause aortic regurgitation, dissection, or rupture [16]. Management of vascular disease in MFS includes regular echocardiography to monitor the aorta, β-adrenergic blockade to decrease arterial pressures (as prophylaxis or in patients with preexisting dilated aorta), and prophylactic valve or aortic surgery. Early treatment with Losartan, a blood pressure medication that also antagonizes TGF-β, may slow the rate of aortic root dilatation [17]. Further randomized trials are underway. Additionally, patients have an increased risk of pneumothorax [4].

17.3.2 Loeys–Dietz Syndrome

LDS is a newly recognized entity that was defined originally in a subset of MFS patients. Though it shares many systemic and skeletal findings with MFS, it is characterized by a triad of findings: (1) hypertelorism, (2) bifid uvula with or without cleft palate, and (3) generalized arterial tortuosity with widespread vascular aneurysms [18].

17.3.2.1 Etiology/Genetics

LDS is an autosomal dominant genetic disorder characterized by mutations in the TGFBR1, TGFBR2, SMAD3, and TGFB2 genes. These mutations make up four subtypes of LDS, called Types 1–4, respectively. TGFBR1 and 2 are genes that encode receptors for TGF-β; SMAD3 is an intracellular signaling intermediate in the TGFB pathway; and TGFB2 is TGF-β Ligand 2. This signaling pathway transduces signals to the nucleus that regulate cellular proliferation, differentiation, and apoptosis. Importantly, it plays a role in the extracellular matrix and is implicated in connective tissue development and function, (including bone and blood vessel formation and function) suggesting a mechanism for disease phenotype. Approximately two-third of LDS cases arise out of de novo mutations and tend to be more severe, while the remaining one-third are familial in origin and are milder [19].

17.3.2.2 Presentation/Diagnosis

As may be expected, LDS shows clinical overlap with MFS including features of aortic root aneurysm, pectus deformities, scoliosis, and arachnodactyly. Despite these similarities, LDS may be distinguishable by unique features such as craniosynostosis, hypertelorism, cleft palate or bifid uvula, cervical spine instability, clubfeet, and arterial aneurysms with tortuosity [19]. As set forth by the LDS Foundation, clinical findings of LDS may be found in Table 17.4 [20].

At present, there exist no specific clinical criteria to establish a diagnosis of LDS, and diagnosis

Table 17.4 Clinical features of LDS

Characteristic triad: *Arterial tortuosity* most commonly in neck *Aneurysms* most commonly in aortic root but may be seen throughout arterial tree *Hypertelorism* *Bifid uvula*	Cardiovascular Dilatation or dissection of the aorta—most commonly at aortic root Other arterial aneurysms and tortuosity—most prominent in head/neck vessels but found throughout arterial tree Congenital heart defects, including patent ductus arteriosus (PDA), atrial or ventricular septal defect (ASD/VSD), and bicuspid aortic valve (BAV)
	Skeletal Pectus excavatum or pectus carinatum Scoliosis Joint laxity or contracture (typically fingers) Arachnodactyly Talipes equinovarus Cervical spine Osteoarthritis Typically normal stature
	Craniofacial Malar hypoplasia Slight downward slant to the eyes Craniosynostosis—most commonly sagittal suture, but all can be involved Bifid uvula/cleft palate Blue sclerae Micrognathia and/or retrognathia
	Skin Translucent skin Soft or velvety skin Easy bruising Dystrophic scarring Hernias
	Other Food or environmental allergies Gastrointestinal inflammatory disease Hollow organs such as intestine, uterus, and spleen prone to rupture

hinges on molecular testing for mutations in one of the four aforementioned genes [21]. If LDS is suspected, a detailed personal and family history must be conducted, as well as a physical examination to identify possible skeletal, craniofacial, or cutaneous manifestations of LDS (see Table 17.4). An echocardiogram with cardiology consult should be performed to assess for aortic enlargement, and a 3D CTA or MRA of the entire arterial tree may be conducted to identify aggressive vascular tortuosity that is characteristic of LDS.

A diagnosis is made in the presence of characteristic LDS findings, and/or a family history of LDS, in conjunction with molecular testing for mutations in TGFBR1, TGFBR2, SMAD3, and TGFB2. Molecular testing methods remain the "gold standard" for confirming diagnosis. All four mutations may cause similar clinical presentations, so it is difficult to distinguish mutations based on clinical findings; however, mutations in SMAD3 may present with early osteoarthritis, and mutations in TGFB2 may present with milder phenotypes [19, 20]. If a genetic test is positive, it is recommended to test the parents and offspring of the affected individual as well [20].

The following guidelines may be employed to determine when genetic testing for LDS is appropriate [22]:

1. Patients who demonstrate the characteristic triad of LDS: hypertelorism, cleft palate/bifid uvula, and aortic/arterial aneurysms/tortuosity.
2. Patients with aortic/arterial aneurysms and variable combinations of other LDS features including arachnodactyly, camptodactyly, clubfeet, craniosynostosis, mental retardation, blue sclerae, thin skin, atrophic scars, easy bruising, joint hypermobility, bicuspid aortic

valve (BAV) and patent ductus arteriosus (PDA), and atrial and ventricular septal defects (ASD/VSD).
3. Patients with a vascular Ehlers–Danlos like phenotype and normal type III collagen biochemistry, but with joint hypermobility and characteristic skin findings (thin skin, atrophic scars, easy bruising).
4. Patients with a Marfan-like phenotype, particularly:
   (a) those without ectopialentis but with aortic/arterial aneurysms, craniofacial features, and skeletal features who do not satisfy the previously described Ghent criteria for MFS.
   (b) as a secondary test for patients with a negative FBN1 molecular test, despite a Marfan-like phenotype.
5. Families with autosomal dominant thoracic aortic aneurysms, especially those who demonstrate aortic/arterial dissection, aortic disease beyond the aortic root, aortic/arterial tortuosity, and ASD/VDS/PDA. Mild Marfan-like skeletal features may be present.

17.3.2.3 Skeletal/Spine Manifestations

Cervical spine malformations and/or instability have been reported in 15–50 % of cases, and are especially severe in patients who demonstrate more pronounced craniofacial abnormalities. Scoliosis has been cited to occur in 25–70 % of patients. One study by Erkula et al. [23] found an average Cobb angle of 29.6° ± 17.9°, with a thoracic curve being the most common pattern. Dural ectasia has been reported in 67–73 % of patients, similar in frequency to MFS, and severity has been demonstrated as a marker of overall connective tissue disease severity [24]. Spondylolisthesis is also noted to occur.

Marfan-like features tend to be seen in patients with LDS; however, the magnitude of limb overgrowth tends to be less severe. The digits tend to be more affected than the limbs, as the rates of arachnodactyly (approximately 50 %) are greater than dolichostenomelia, which is in contrast to MFS where increased arm-span-to-height ratio is a prominent finding. Combined Steinberg (thumb) sign and Walker–Murdoch (wrist) sign is present in one-quarter to one-third of patients, which is more common than in the general population, but less common than in MFS. Approximately half of patients demonstrate joint hypermobility, and congenital hip dislocation and joint subluxations are common features. Joint contractures also occur in some individuals, and camptodactyly and clubfeet are the most common manifestations of reduced joint mobility. This is unusual because joint contractures occurring in conjunction with hypermobility are common in LDS but rarely seen in the general population [19, 23]. A more complete list of findings may be found in Table 17.4.

17.3.2.4 Loeys–Dietz and Sports

While Loeys–Dietz patients should remain cardiovascularly active, they should not exercise to the point of exhaustion, should not participate in competitive/contact sports, and should not do isometric exercises (sit ups, pull ups, push ups, weight lifting, etc.) [25].

17.3.2.5 Spinal Treatment and Complications

Cervical spine instability has been associated with LDS. It is recommended that patients obtain cervical spine films in flexion and extension at the point of diagnosis to assess for cervical abnormalities, subluxations, or instabilities. Children who do not show cervical instability at baseline may be recommended to repeat images every 3–5 years during growth to assess for changes that may warrant further care [25].

Scoliosis and kyphosis often requires treatment. While there is currently no data on the efficacy of bracing, it may be helpful for growing children with mild curves (<25°). However, it is recommended that patients be monitored yearly until skeletal maturity due to the higher propensity for progression [25]. In the authors' experience, early-onset scoliosis in LDS can be treated similarly to MFS with bracing followed by growing rods if it becomes significant (Fig. 17.3).

While surgery tends to be tolerated in Loeys–Dietz patients, there is a propensity for increased bleeding intraoperatively. Also, delayed bone

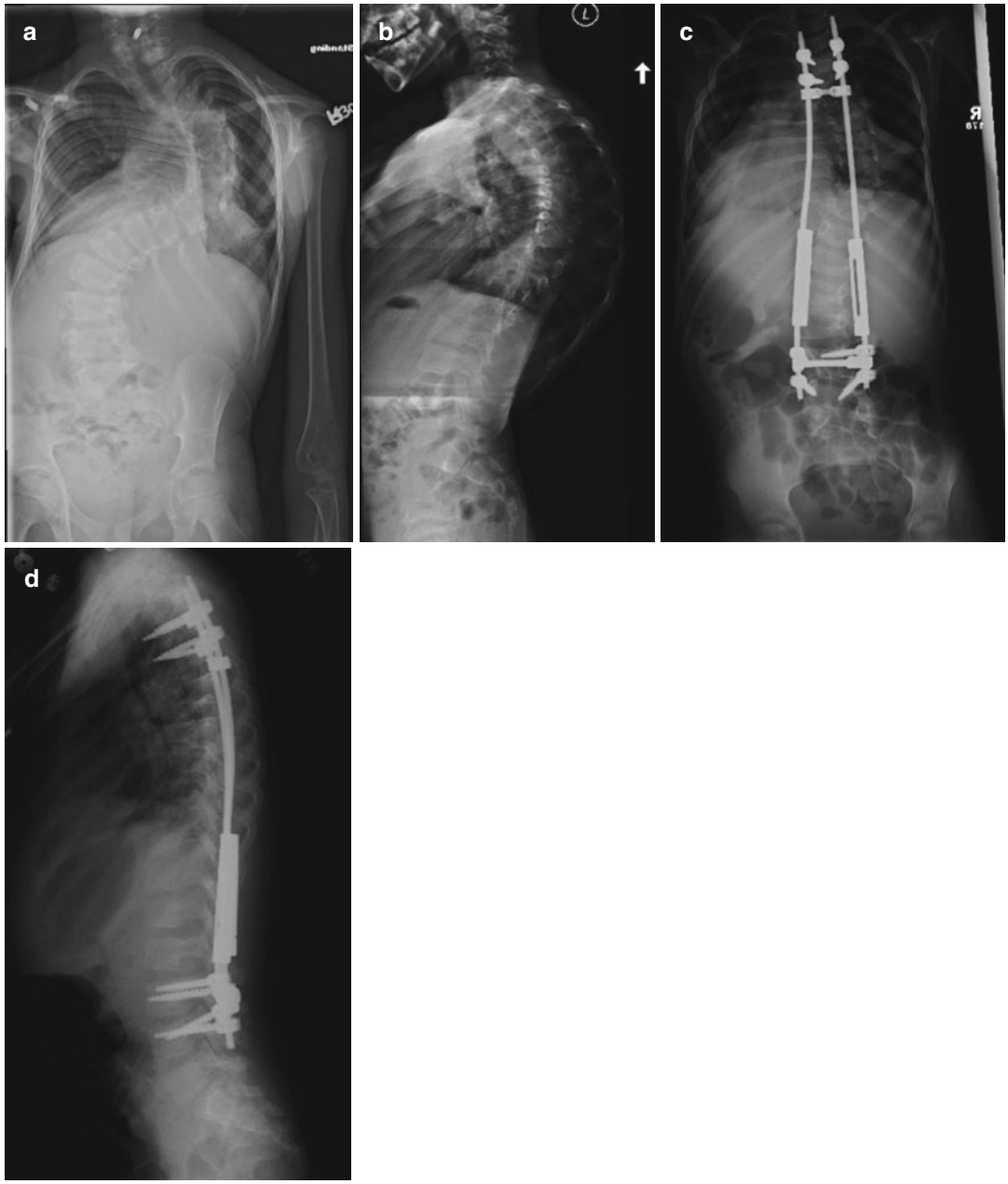

Fig. 17.3 The 6-year-old girl with LDS (Preop: **a**, **b**) was treated with growing rods from T3–T4 to L3–L4. Pedicle screws were used for both proximal and distal fixation. Rods were inserted below the submuscular layer and autologous bone graft was liberally used. Cross-links were used distally and proximally for stable fixation. A small dural leak was encountered due to duralectasia. Major curve correction using the growing rods in the patient was significant from 101° to 40°. Postoperative complications included a small superficial wound infection that was successfully treated with antibiotics (Follow-up: images **c** and **d**)

healing and pseudarthrosis have been reported in association with lack of fixation of pedicle screws [23]. Due to the tissue laxity, kyphosis at the proximal implant junction or several levels above is common (Fig. 17.4). The fusion of lumbar spondylolisthesis can usually be combined with

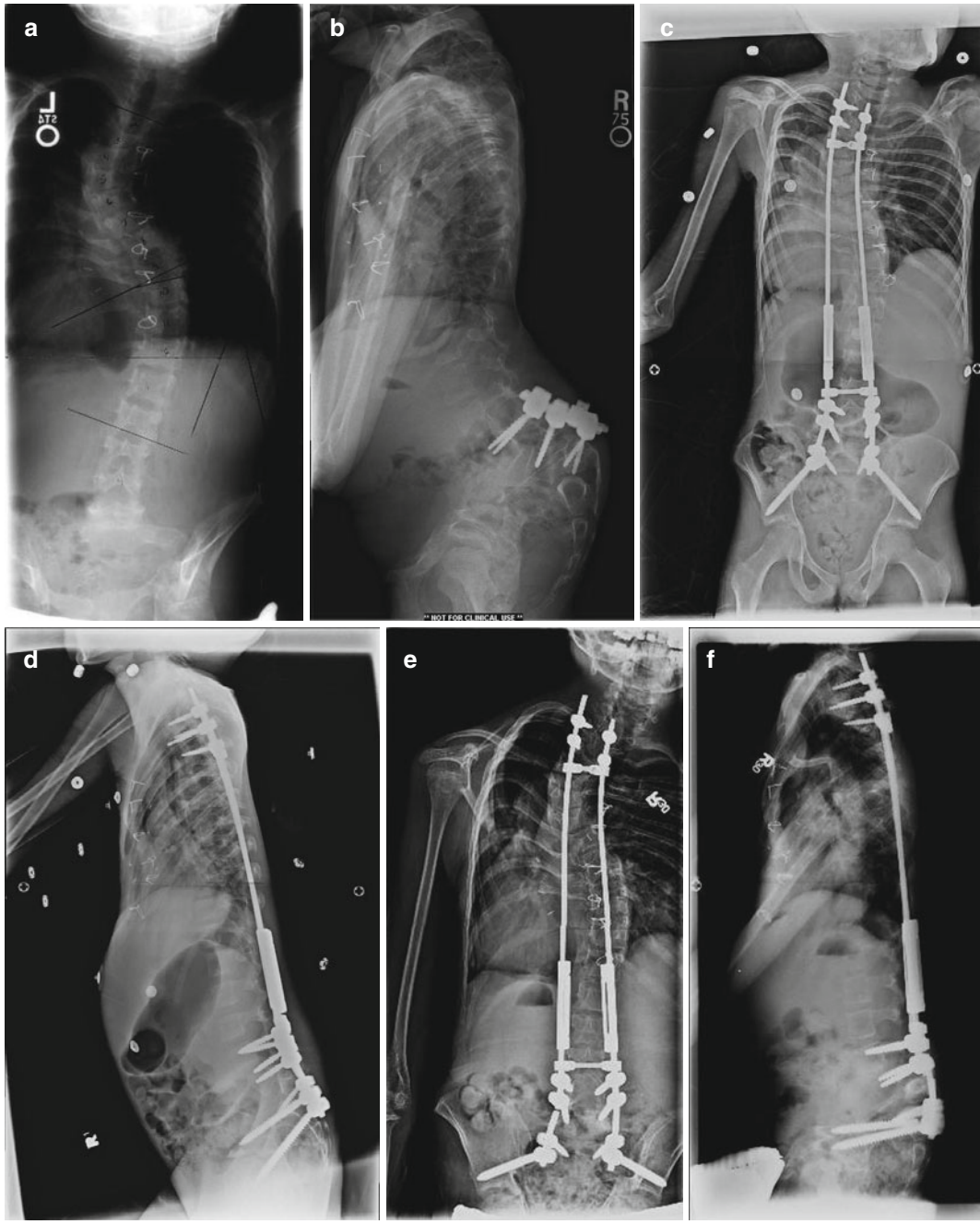

Fig. 17.4 The 13-year-old female with LDS **a**. Image **b** demonstrates a grade 4 spondylolisthesis that was previously repaired, but redeveloped at the L5–S1 junction following an injury. The patient underwent insertion of growing rods with proximal pedicle screws at T2–T3 and distal screws at L4–S2 and in the sacroilium (images **c** and **d**) to better correct her sagittal deformity. Images **e**–**h** show two occasions of instrumentation failure. The patient experienced increased cervical instability (image **i**) superior to the proximal implant junction and underwent cervical fixation, but ultimately necessitated whole-spine fusion from occiput to sacrum, seen in images **j** and **k** (most recent postoperative cobb 57°)

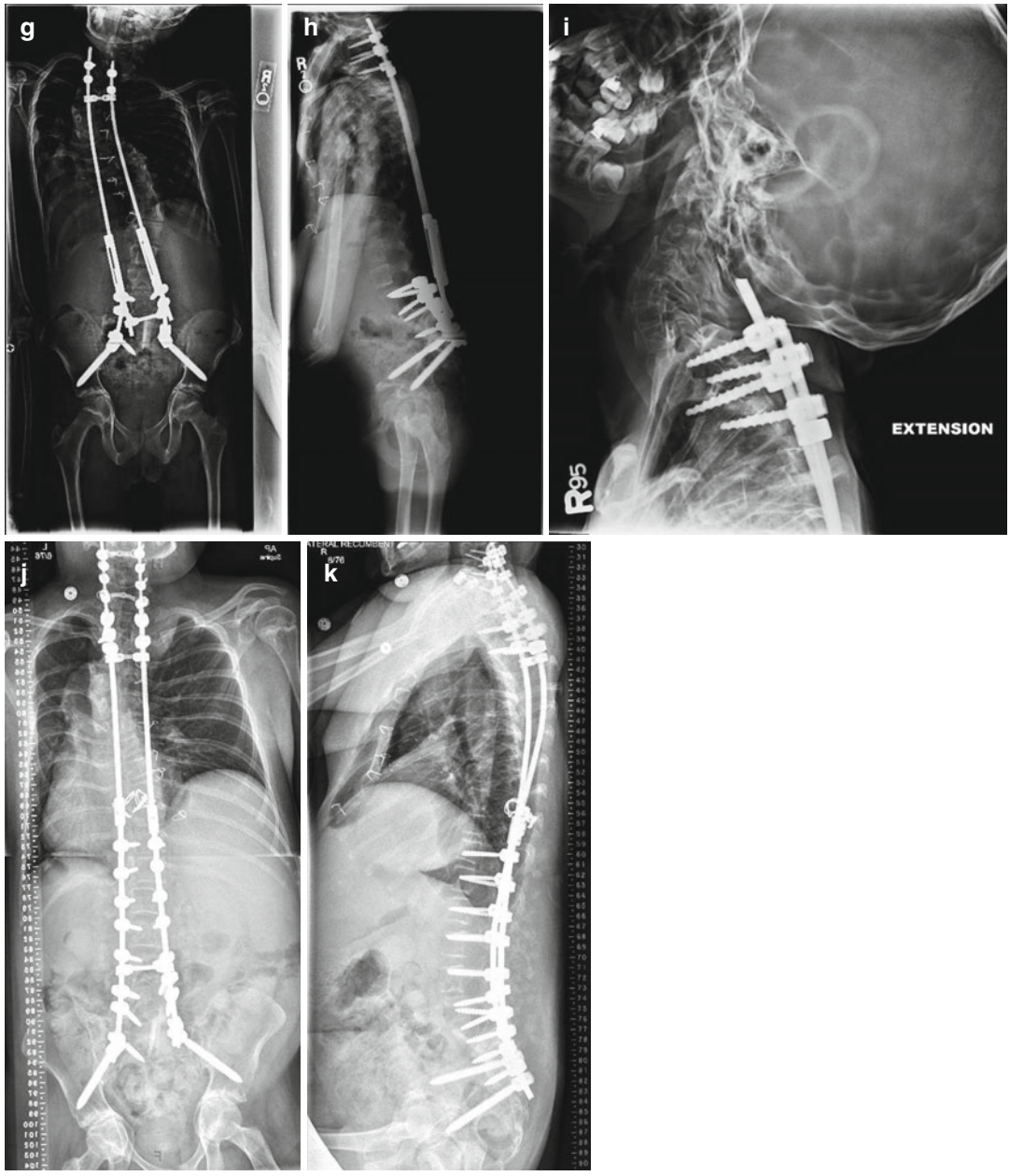

Fig. 17.4 (continued)

growing treatment of the thoracolumbar kyphoscoliosis (Fig. 17.4). Due to the frequent cervical anomalies, these patients may eventually require instrumentation of their entire spine from occiput to pelvis (Fig. 17.4). Dural ectasia is also seen in LDS and patients are at increased risk for tear.

Osteoporosis and osteopenia are reported with greater frequency in LDS compared to MFS. LDS patients have a high risk for fracture, with a reported rate of 50 % by age 14 [26]. Of note, disk degeneration has been noted to occur at early onset in LDS patients [25].

Affected patients have a high risk of aortic dissections or ruptures at an early age and at blood vessel sizes that are not associated with risk in other conditions. Typically, vascular involvement is also more progressive in LDS patients than in MFS patients. Similar to MFS, strict management of blood pressure is advised. Medications that negatively affect the cardiovascular system should be avoided, such as stimulant medications and vasoconstrictors [25].

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Other Syndromic Disorders of the Growing Spine

18

Ehsan Jazini, Jarred Bressner, and Paul D. Sponseller

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Key Points

- Each syndrome has key medical points for which specialists should be involved.
- Bracing has less success in controlling curves in syndromic scoliosis than in idiopathic scoliosis.

18.1 Shprintzen–Goldberg Syndrome

Shprintzen–Goldberg syndrome (SGS) is characterized by craniosynostois, marfanoid habitus, intellectual disability, skeletal, cardiovascular, and connective tissue anomalies [1].

18.1.1 Etiology/Genetics

Genetic mutations in both the FBN1 and TGFRB2 have been associated with SGS [2, 3]. More recently, de novo missense mutations in *SKI* gene (SKI protein is a known repressor of

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TGFβ signaling) were found in most individuals with SGS [4]. There are no pathognomonic signs of SGS. Therefore, diagnosis is dependent on recognition of examination patterns and molecular anomalies. Craniosynostoses and cognitive delay are distinguishing features [5]. The phenotypic features overlapping with LDS, MFS and EDS may make the diagnosis challenging. Mode of inheritance of SGS is usually sporadic, with some rare instances of autosomal dominant described [6].

18.1.2 Presentation

Facial features include hypertelorism, downslanting palpebral fissures, high arched palate, micrognathia, and low-set ears. Other reported features include neonatal hypotonia, abdominal hernias, and minimal subcutaneous fat [5]. While in Marfan syndrome the eyes are characterized by enophthalmos, SGS patients present with proptotic eyes [7]. Unlike in MFS or LDS, cognitive delay is nearly universal in SGS.

18.1.3 Skeletal/Spine Manifestations

In the skeletal system, SGS is associated with arachnodactyly, pectus deformity, camptodactyly, and joint hypermobility [5]. However, none of these features are specific. Spinal abnormalities include: scoliosis, C1–C2 abnormality, 13 pairs of ribs, square-shaped vertebral bodies, and osteopenia.

18.1.4 Surgical Deformity Treatment and Complications

Developmental scoliosis is common in SGS and can be treated similarly to MFS. Significant coronal and sagittal imbalance may occur (Fig. 18.1). Due to the poor quality of bone and thin habitus, risk of implant failure and loss of correction are of major concerns. In one clinical series by Watanabe et al., the patients were treated surgically for scoliosis with both growing rods and posterior spinal fusion. The curve patterns were double major or triple major with average Cobb angles 102.8 ± 16.9 with kyphosis at the thoracolumbar area in all the patients, with a mean kyphosis angle of 49° ± 16° [8]. The authors noted that three out of four patients had dural ectasia. They reported high complication rates, including implant dislodgement (3/4 of patients), postoperative infection (2/4 patients), pseudarthrosis, and loss of kyphosis correction. The use of multiple points of fixation (screws, sublaminar wiring, hooks) is recommended to increase chances of curve correction and to prevent implant failure. Careful soft tissue dissection and appropriate soft tissue coverage of implants are vital to decrease development of pressure ulcers and subsequent infection in SGS patients characterized by thin habitus.

18.2 Ehlers–Danlos Syndrome

Ehlers–Danlos syndrome (EDS) is a class of connective tissue disorder caused by defects in collagen synthesis. It is characterized by distensible and thin skin, easy bruising, hyperextensible joints, facial features, and severe arterial complications. The eye, gastrointestinal, respiratory, and cardiovascular systems can also be affected.

18.2.1 Etiology/Genetics

EDS is not a homogeneous disorder and can be thought of as a group of related entities that share, to varying degrees, the same complex of physical anomalies. Therefore, various subclassifications exist with different clinical presentations and different genetic mutations. According to the Villefranche classification, Type VI is characterized by progressive infantile scoliosis. It is inherited in an autosomal recessive fashion with mutation in *PLOD* gene (encoding lysyl hydroxylase important in collagen cross-linking) [9].

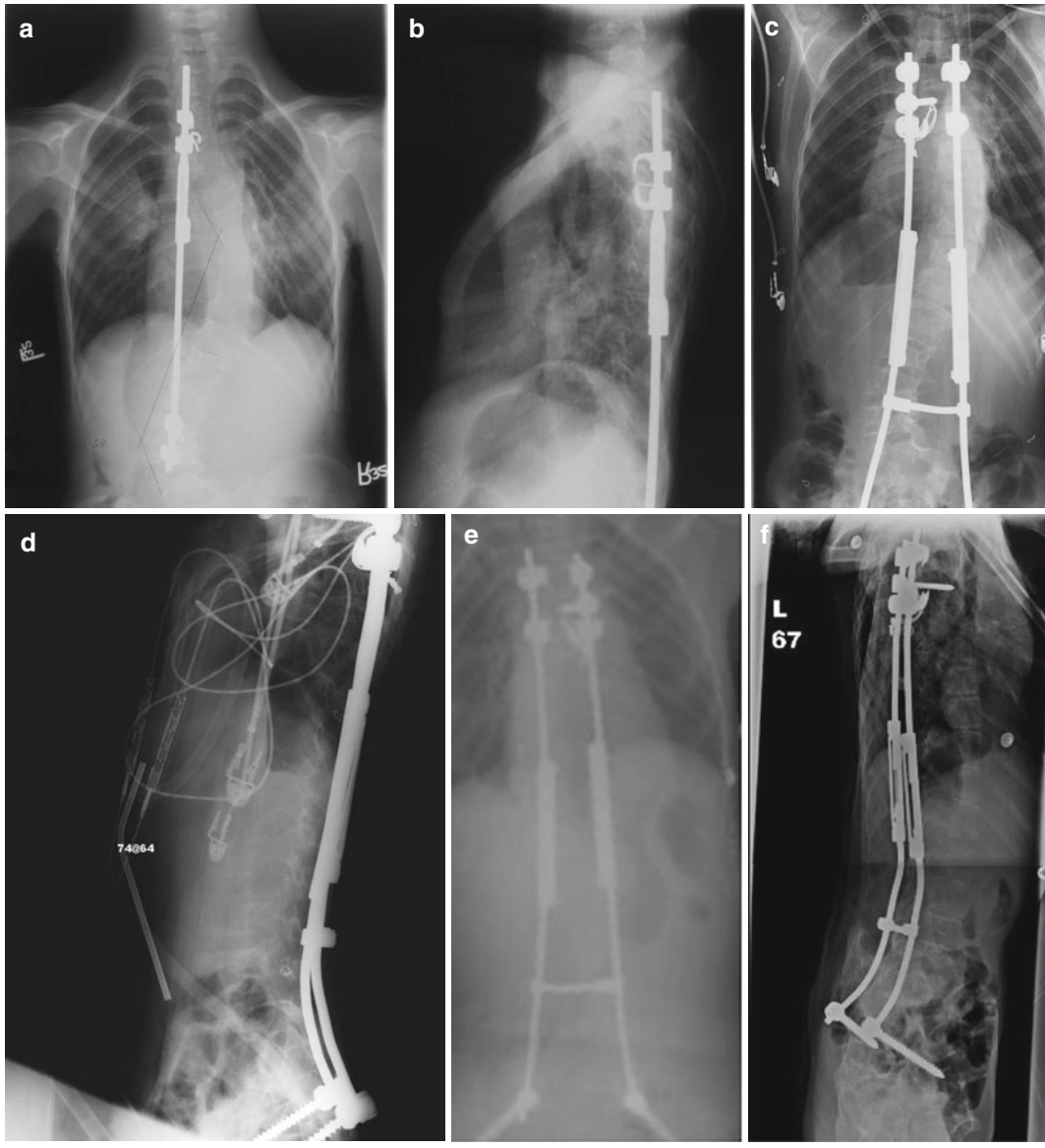

Fig. 18.1 The 9-year-old female patient with SGS. The patient had a previous growing rod instrumentation to treat pelvic obliquity (**a**, **b**), however, developed lordosis and significant decompensation with a right trunk shift. (**c, d**) show images following initial growing rod surgery. In postoperative follow-up, this patient has had three lengthenings in 3 years with a total increase of 4 cm in T1–S1 length (**e**, **f**). Complications since initial surgery have included distal screw breakage and rod erosion that required successful revisions

18.2.2 Skeletal/Spine Manifestations

Skeletal manifestations include developmental dysplasia of the hip, club foot, pes planus, joint hypermobility and dislocation, generalized ligamentous laxity, and scoliosis [9]. Kyphoscoliosis is a hallmark feature of type VI EDS; however, scoliosis also often presents at an early age in patients with other classes of EDS, most notably Types I, II, and III [10] (Fig. 18.2). Osseous fragility is often seen.

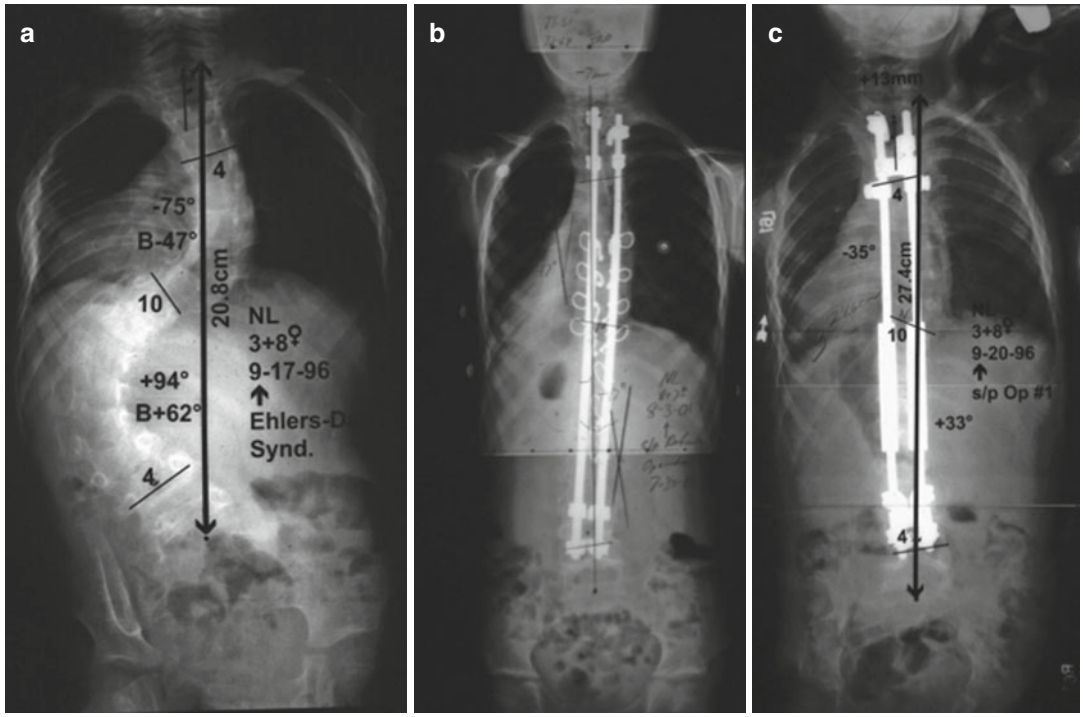

Fig. 18.2 The 3-year-old female with EDS with preoperative curve over 90° (**a**) underwent placement of growing rods (**b**). The patient later went on to successful posterior fusion at the age of 8 (**c**) with curve at last follow-up of 25° (Case courtesy of Marc A. Asher, MD)

18.2.3 Surgical Treatment and Complications

With scoliosis surgery, it is important to keep in mind the vascular fragility that is inherent in this disease. Although spine surgery in MFS can be associated with increased bleeding, anterior approaches to the spine should be avoided when possible in EDS because such surgery can be catastrophic involving large arteries and veins. Akpinar's review of five cases with Type VI EDS who underwent surgical treatment of scoliosis had two cases of vascular complications during the anterior approach, one involved avulsion of the segmental arteries from the lower aorta and common iliac vein requiring gortex graft repair [10]. Vogel et al. also reported major vascular complication in one out of four patients associated with the anterior approach due to the inherent vascular fragility in EDS patients. They also reported major neurological complications including permanent paraplegia in two patients [11].

Growing procedures in EDS patients, if begun early enough, may allow posterior-only approaches so that the patient never requires an anterior procedure or a complex posterior osteotomy. Measures such as hypotensive anesthesia and careful dissection of segmental arteries are advised [10]. Recently, a report of using Factor VIIa to help control massive bleeding following spontaneous large vessel rupture in Type IV EDS has been published [12].

18.3 Prader–Willi Syndrome

Prader–Willi syndrome (PWS) is characterized by early hypotonia, developmental and motor delay, small hands and feet, and later hyperphagia resulting in massive obesity.

18.3.1 Etiology/Genetics

PWS is caused by a lack of paternal expression of a region of chromosome 15. Pituitary dysfunction results in many generalized (listed earlier) and orthopedic manifestations [13].

18.3.2 Natural History/Skeletal Manifestations

Orthopedic manifestations include growth retardation, hip dysplasia, and scoliosis. Scoliosis is seen in 66 % of patients with PWS by time of skeletal maturity, according to longitudinal study by Odent et al. [14]. Its onset is often in the infantile or juvenile period with a mean age of onset 10.2 ± 6.2 years [14]. Increased body mass index (BMI) was a risk factor for developing associated kyphotic deformity which led to a higher likelihood of surgical treatment. Annual systematic clinical examination for scoliosis is recommended.

18.3.3 Spinal Deformity Treatment and Complications

Orthoses have a role if body habitus does not prohibit it. Administration of human growth hormone (HGH) has been shown to aid in the management of many aspects of this condition [13]. Initially, there was concern that HGH may increase the prevalence and severity of scoliosis [13]. However, it appears to help control many aspects of the disease and does not increase the incidence or severity of scoliosis [14]. In fact, a study out of South Korea purports that in their clinical series preoperative treatment with HGH before surgical fixation may reduce postoperative complications [15]. Massive obesity is now rarely seen in PWS since the advent of HGH treatment.

Treatment of scoliosis in this condition should follow usual clinical guidelines. If curve size increases beyond orthotic range in the early juvenile period, there may be a role for growth-guiding surgery (Fig. 18.3). At the time of any surgical procedure, monitoring for sleep apnea is important. Other relevant important perioperative considerations are that these patients have higher likelihood of osteopenia, depression, and diminished pain sensitivity [16]. Proximal junctional kyphosis with acute cord stenosis has been reported after spine surgery in PWS.

18.4 Rett Syndrome

Rett syndrome (RS) is a rare progressive neuromuscular disorders first described a generation ago by Austrian physician Andreas Rett. It is often confused with cerebral palsy.

18.4.1 Etiology/Genetics

Its etiology has been recently defined as a defect in the transcription repressor *MECP2* gene at chromosome Xq28 [17]. Two mutations (R294X and R306c) of the *MCEP2* gene predicts a less severe form of RS and less worsening of scoliosis [18].

18.4.2 Presentation

Virtually all patients are female, and manifest stereotypic hand movements, little to no expressive language, seizures, and a neurological picture combining dystonia and spasticity.

Between 50 and 90% of patients develop scoliosis during the juvenile growth with rapid worsening during adolescence with continued progression into adulthood [18]. With no treatment, this leads to severe impairment including difficulty sitting, pain, and respiratory difficulty. Worse severity of disease, characterized by earlier onset of regression, and inability to achieve walking also portend rapid progression of scoliosis, with long single curves down to the pelvis [19].

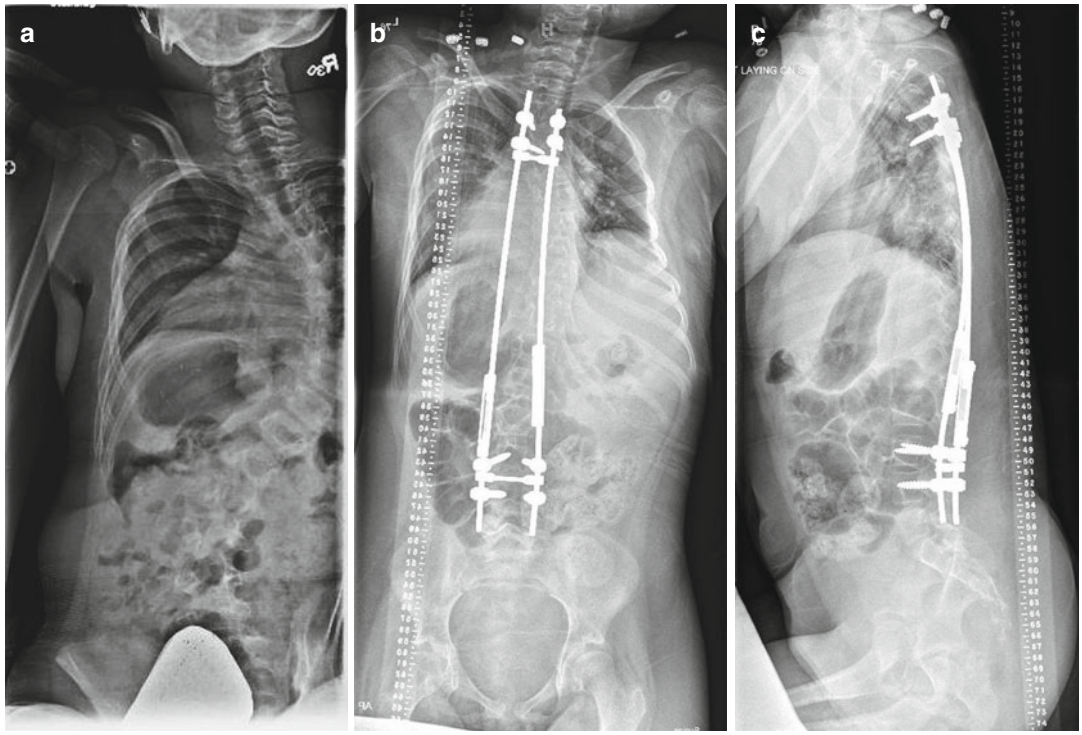

Fig. 18.3 The 9-year-old male with PWS. The patient had a collapsing 90° scoliosis that progressed despite attempted bracing (**a**). Growing rods were inserted and **b** and **c** display images after second growing rod surgery with a Cobb angle reduced to 36°

18.4.3 Spinal Deformity Treatment and Complications

Neither physical therapy nor bracing has been proven to affect curve progression. Growing instrumentation is an option to control curves (Fig. 18.4). Because of the profound neurological disorder, pelvic obliquity should be controlled, and therefore strong consideration was given to pelvic fixation. Osteoporosis is common (Fig. 18.4). Experienced pulmonary backup or support is needed even after simple lengthening procedures. If patients develop surgical curves in the juvenile period, the author's preference is to postpone surgery until the peak height velocity (approximately age 10) does not exceed 90°. At that time, a single-stage fusion can be performed. Growing spinal implants are associated with more complications in this disorder than in other disorders and we prefer to avoid these.

18.5 Down Syndrome

18.5.1 Etiology/Genetics

Down syndrome is the most common chromosomal disorder. Most cases involve complete trisomy of the 21st chromosome, but a smaller number are translocations and carry less pronounced manifestations. *COL6A1* and *COL6A2*, the genes that encode Type VI collagen, are encoded on chromosome 21 and are believed to play a role in the joint laxity responsible for many of the skeletal manifestations in these patients [20].

18.5.2 Skeletal/Spine Manifestations

Musculoskeletal findings are common in Down syndrome. Cervical spine instability is the most significant finding and may occur at both the atlantoaxial joint (C1–C2) and the occiput–C1 junction.

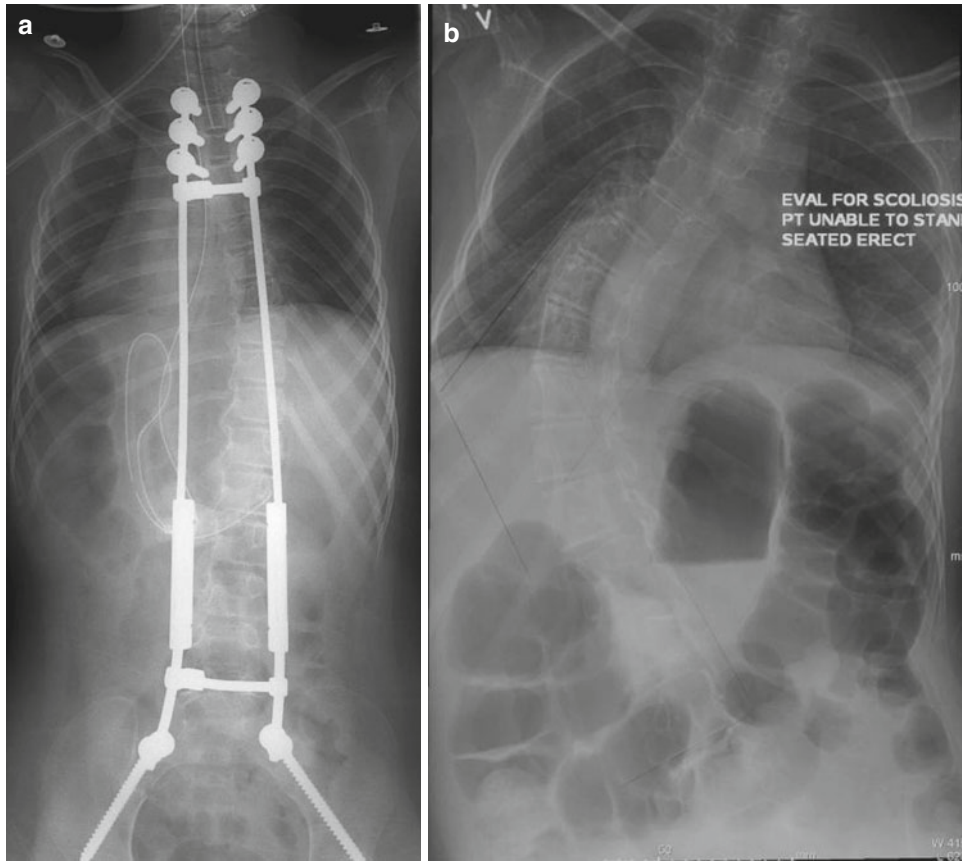

Fig. 18.4 This is a female Rett patient who had a 70° curve, convex to the right, that corrected to 30° with traction. Growing rods were inserted at 9 years old (image **a**). After five distractions, the patient required removal of the growing rods due to a deep back wound infection (*Enterobacter cloacae*) that could not be suppressed and necessitated surgical debridement. Although the posterior elements appeared fused, a deformity redeveloped; however, no further surgery was performed (image **b**)

Incidence has been reported in 10–30 % of patients; however, the majority of patients with cervical instability are asymptomatic [21]. Hypermobility and instability are believed to occur due to ligamentous laxity [20]. Scoliosis is also present with increased frequency in Down syndrome patients and reviews have estimated that 10 % of Down syndrome patients have scoliosis over 20° [22, 23].

18.5.3 Spinal Deformity Treatment and Complications

Identification of cervical instability is critical. As stated, most patients are asymptomatic; however, unidentified cases have risk of progression and irreversible damage due to cord compression. Screening via lateral radiographs in neutral position, flexion, and extension is recommended to diagnose cervical instability. Down syndrome patients with atlanto-axial translation greater than 5 mm should be monitored periodically. Surgical fusion of this interval is indicated if the patient has myelopathy, translation greater than 8 mm, space available for the cord of less than 13 mm, or may incur head impact. When no cervical instability is found, it is recommended that the patient's family be alert for symptoms of neurological compromise. In asymptomatic patients with instability, symptom surveillance, neurological examination, and additional radiographic evaluation are recommended [20]. Surgical

stabilization may be indicated in patients who display neurological symptoms such as neck pain, deterioration of motor activity, failure to meet motor developmental milestones, or demonstration of neurological findings associated with brainstem or spinal cord compression [21]. It must be noted that arthrodesis is challenging in these patients and complication rates are very high. Noted complications include infection, delayed wound healing, nonunions, loss of reduction, bone graft resorption, junctional instability, and neurological deterioration [20]. There is currently no data on the efficacy of nonfusion options for this population. Additionally, during any surgical procedure, cervical instability should be managed and the neck should be maintained in a neutral position.

Scoliosis is also found at increased frequency in Down syndrome patients, though the incidence is not known at present. It has been associated with previous thoracotomy for cardiac surgery [20]. Scoliosis should be screened at regular visits to the pediatrician or orthopedic surgeon. Treatment efficacy is undocumented. For curves of 25°–40°, bracing may be offered for compliant patients. Surgical fixation is indicated in curves greater than approximately 55° [24]. As in cervical stabilization, surgical fixation for scoliosis is accompanied with a high risk of complications including wound infections, delayed healing, pseudarthrosis, implant failure, and junctional instability [21].

In summary, care of children with syndromes is challenging and rewarding. Many of them present with significant spinal deformities at a young age and they usually have associated medical problems. By building the appropriate care team and skill set, the pediatric spine surgeon can successfully manage deformities which would otherwise cause significant morbidity and even mortality.

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Spinal Deformity in Metabolic Disorders

19

James O. Sanders

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Key Points

- Scoliosis is the primary deformity from soft bone diseases in children.
- Bracing has little role in these disorders both because it is ineffective and it can create significant chest wall deformity.
- Medical treatment of these disorders has a very important role in both preventing spinal deformity and providing improved fixation when surgery is necessary.

19.1 Generalities

19.1.1 Soft Bone and Spinal Deformity

Soft bone disease comes in many forms and may cause or coexist with spinal deformity. The poor bone quality may cause macrofractures, microfractures, or plastic bone deformation creating a spinal deformity. Bracing is typically ineffective and the pressure from bracing creates secondary deformities of the chest wall. Surgery is complicated by bone bleeding when fractured and the soft bone making correction difficult from poor bone purchase. There are several strategies to minimize these problems, but some appear

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unavoidable with current knowledge and techniques. This chapter outlines some generalities, which are discussed in more detail with the specific diseases in this chapter.

19.1.1.1 Scoliosis

Unlike adults in whom the primary disorder from soft bone is kyphosis, scoliosis is the primary disorder in children with soft bones. Although older studies recommended bracing, the evidence for bracing, particularly in this population, is poor. Bracing exerts pressure upon the ribs, which can deform and constrict lung volume. If bracing is attempted, it must be done with great caution and careful evaluation for its effects. Halo traction using multiple halo pinsto providing sufficient surface area contact has been used successfully in obtaining preoperative correction in soft bone disease. This is likely because the ligaments are often relatively strong despite the bone abnormality and ligament laxity. Scoliosis correction can be difficult, and most authors recommend operating upon curves before they become large. Early reports had little correction, but this appears improved with modern techniques.

19.1.1.2 Kyphosis

While rarer than scoliosis, kyphosis still occurs in children with soft bone. When occurring over a short segment, typically from fractures, it can cause spinal cord compression requiring decompression. Kyphoplasty reported use in children with soft bone is limited to a case reports [1, 2]. Correction of kyphosis in the young may require an anterior fusion for structural support. This can be difficult because the anterior bone is often substantially weaker than the posterior bone.

19.1.1.3 Spondylolysis and Spondylolisthesis

Pathological bone is a separate classification in the major spondylolisthesis classifications. We have only seen it below posterior fusions, but there is no reason to suppose it does not occasionally occur otherwise. It is usually best treated by observation.

19.1.1.4 Basilar Invagination

Basilar invagination occurs when the heavy skull settles on the upper cervical spine. The foramen magnum is compressed by the dens pushing into the brain stem and creating abnormal CSF flow. Although patients are often asymptomatic [3], they may present with headache, lower cranial nerve problems, hyperreflexia, quadriparesis, ataxia, and nystagmus.

19.1.1.5 Poor Bone Purchase

Fixation in soft bone can be quite difficult. The basic principles are to purchase the strongest bone with the widest surface area possible. This can be achieved with multiple wires, screws, and hooks. The anterior cancellous bone is often quite thin and does not provide good purchase. Screw purchase can be maximized using diameters large enough to engage the pedicular cortex and catching the endplates that are stronger than the vertebral bodies. Polymethylmethacrylate (PMMA) has been used to improve purchase of both hooks and screws. Since the advent of bisphosphonate treatment of soft bone disease in children, we have not used PMMA, but have instead used two or three preoperative courses of intravenous pamidronate preoperatively to improve purchase.

19.2 Rickets and Rickets Like Syndromes

19.2.1 Rickets

The classic metabolic bone disease is rickets. Rickets includes several conditions that lead to abnormal bone mineralization in a growing child. Rickets refers to a failure or delay in mineralization of newly formed osteoid at the growth plates, while osteomalacia is a delay in mineralization of newly formed osteoid as a part of bone remodeling. Children suffer from both problems while skeletally mature individuals have osteomalacia only. Vitamin D plays a vital role in the development of rickets and osteomalacia. Vitamin D can be consumed in the diet and can also be formed in the skin by UV-B irradiation from sunlight. Vitamin D is transported to the liver where it is hydroxylated to 25(OH) vitamin D, and then to the kidney where it is again hydroxylated by 1-alpha-hydroxylase into $1,25(OH)_2$ vitamin D. This active form of vitamin D increases

intestinal absorption of calcium and phosphorus, and renal reabsorption of calcium (and phosphorus) to maintain a supersaturated state of a calcium–phosphorous product in the serum that results in passive mineralization of newly formed osteoid [4]. 1,25$(OH)_2$ vitamin D also directly effects osteoblast activity, increasing expression of several bone proteins. Levels of calcium, phosphorus, and parathyroid hormone (PTH) closely regulate the 1-alpha-hydroxylase enzyme, allowing for homeostatic balance. There are different forms of rickets, including vitamin D deficiency, vitamin D-dependent and -resistant rickets, and hereditary hypophosphatemic rickets.

19.2.1.1 Vitamin D-Deficient Rickets

The most common rickets is vitamin D deficiency rickets, which results from deficient intake, absorption, or production of vitamin D. Diets low in vitamin D (especially the exclusively breastfed infant or the strict vegetarian), dark skin pigmentation, limited sun exposure, or regular strict use of sunscreen are all risk factors as are malabsorptive syndromes, anticonvulsants, and steroids. Children may have failure to thrive, short stature, delayed development, muscular hypotonia, or hypocalcemic seizures. Widening of the wrists and ankles, chest deformities, and bowing of the long bones are common skeletal manifestations. In advanced disease, calcium and phosphorus are both low, AP and PTH are elevated, and 25(OH) vitamin D is low. 1,25$(OH)_2$ levels are not helpful as they are generally normal or even elevated. Treatment includes vitamin D and calcium. Serum 25(OH) vitamin D levels are the best clinical indicators of nutritional vitamin D status and should be maintained at levels >32 ng/ml (75–80 nM) [5]. The author has treated one African American child with nutritional rickets and progressive infantile scoliosis who responded completely to serial casting and therapeutic vitamin D supplementation.

19.2.1.2 Vitamin D-Dependent Rickets

This form of rickets results from deficient or abnormal function of the renal 1-alpha-hydroxylase resulting in low 1,25$(OH)_2$ vitamin D levels. Vitamin D-resistant rickets are caused by a defect in the vitamin D receptor that prevents 1,25$(OH)_2$ vitamin D binding. Both types are caused by genetic mutations and lead to hypocalcemia, secondary hyperparathyroidism, hypophosphatemia, and the typical skeletal manifestations seen in vitamin D-deficient rickets. Treatment includes pharmacological doses of vitamin D or 1,25$(OH)_2$ vitamin D (calcitriol), as well as calcium.

19.2.1.3 Hereditary Hypophophatemic Rickets

In this disorder there abnormal renal phosphate reabsorption occurs, and phosphate "wasting" ensues. This is typically a X-linked recessive disorder, but rarely, autosomal dominant inheritance can occur. Patients have hypophosphatemia, hyperphosphaturia, and elevated serum alkaline phosphatase, but normal calcium, PTH, and 25(OH) vitamin D levels, and decreased 1,25(OH) vitamin D levels. Skeletal manifestations are similar to those seen with the other forms of rickets; symptoms related to hypocalcemia (seizures, tetany), however, do not occur. Interestingly, there is no evidence of increased osteoclast activity, and in treated patients, bone mass can be normal [6]. Treatment is with large quantities of oral phosphate.

In the late-nineteenth through mid-twentieth century, rickets was considered a common cause of scoliosis [7].. This was supported by experiment evidence from bipedal rats on a rachitogenic diet which produced scoliosis in contrast to either quadrupedal rachitic or bipedal nonrachitic rats [8]. Pehrsson et al. [9] found an increased mortality rate in scoliosis with rickets and an increased incidence of severe scoliosis, but the study is hampered by uncertain diagnosis of the rickets with other dwarfing conditions. Rickets does result in an increased incidence of asymmetric posture [10]; however, human scoliosis in practice is rarely if ever caused by rickets [7]. We have treated a number of families with familial hypophosphatemic rickets and have not identified any with scoliosis, perhaps because their growth spurt is markedly diminished compared to other children.

19.2.2 Hypophosphatasia

Hypophosphatasia results from abnormally low activity of tissue nonspecific alkaline phosphatase (TNSALP), which leads to rickets and/or

osteomalacia. Several mutations and inheritance patterns are present and depend on the type of hypophosphatasia. The different forms of this disorder are classified by age at onset of skeletal manifestations: perinatal, infantile, childhood, and adult. Two other forms include odontohypophosphatasia and pseudohyphosphatasia. The perinatal form is lethal; those that are stillborn may have bone spurs of the extremities and characteristic radiographic findings – abnormal ossification of bones, round and flattened vertebral bodies. Those that survive birth suffer from respiratory failure. The infantile form may present as failure to thrive, poor feeding, and hypotonia in the first 6 months of life. Craniosynostosis, dolicocephaly, short stature, and fractures may occur. This form carries a 50 % mortality rate, usually from rachitic rib changes and subsequent respiratory compromise. The childhood form may present as delayed motor development and early loss of deciduous teeth. Many patients suffer from severe bone pain. The adult form presents in the fourth–fifth decade of life with symptoms resulting from stress fractures or joint pain. Premature loss of deciduous teeth also occurs. Odontohypophosphatasia is characterized by premature loss of teeth and no evidence of skeletal disease. Pseudohypophasphatasia is clinically indistinguishable from the perinatal form, except for normal alkaline phosphatase activity. It is thought that although TNSALP functions normally in vitro, it has abnormal activity in vivo. There are only case reports of scoliosis in hypophosphatasia [11, 12]. Whether this was caused or coexisting with the disease is unknown, although Arun et al. [12] suggest a genetic linkage rather than a common etiology. Scoliosis has also been reported rarely in hepatobiliary rickets [13].

19.3 Lowe's Syndrome

Patients with Lowe's oculocerebrorenal syndrome may have scoliosis or kyphosis, but more commonly have upper cervical abnormalities similar to patients with storage diseases which are discussed later [14].

19.4 Osteogenesis Imperfecta

Osteogenesis imperfecta (OI) is a disorder of congenital bone fragility, usually resulting from abnormal type I collagen. It commonly results in spinal deformity. The molecular mechanisms of these disorders are being increasingly understood, and for surgeons dealing with various severities of spinal deformity, it is important to understand something of this new knowledge. The most common types of OI result from problems with type I collagen. Collagen is a triple helix, and more severe forms of OI can result when one abnormal collagen monomer destabilizes the entire triple helix. Mutations in the *COL1A1* and *COL1A2* genes, coding for type I collagen, can lead to both qualitative and quantitative disturbances of the protein. Types of OI are distinguished by the severity of their manifestations and inheritance. Normal but insufficient type I collagen is typical of type I OI, the mildest form of OI, where fractures are most common in infancy. Patients have blue sclera and occasionally abnormal dentition. Long bone deformities are less prevalent and severe than in other types. Type II is often fatal in the perinatal period. In utero fractures are present, and lung hypoplasia and CNS malformations are common causes of death. Type III is considered a severe type of OI with more frequent fractures, including in utero. Long bone deformities are characteristic and muscle weakness and bone pain can be debilitating. Kyphoscoliosis may be severe enough to cause respiratory compromise and basilar invagination may be fatal. Sclera are not typically blue. Type IV has phenotypic similarities to Type I, with more severe bone involvement and lack of blue sclera. There are now 15 different OI's listed on Online Mendelian Inheritance in Man (OMIM) with new types regularly being identified. Types V–XV do not have primary defects in type I collagen and are inherited autosomal recessively. Table 19.1 lists the currently described types of OI, links to the Online Mendelian Inheritance in Man (OMIM, and their known association with spinal deformity.

Pulmonary failure is the leading cause of death in adults with OI, and it is closely associated

Table 19.1 Genetic metabolic and connective tissue disorders associated with spinal deformity in children

Disorder and OMIM link	Pathophysiology and inheritance	Typical features	Treatment	Association with spinal deformity
Rickets like disorders				
Vitamin D-deficient rickets	Vitamin D deficiency from poor dietary intact, often breastfed, dark-skinned	Widened physes, lethargy, limb deformities	Physiological doses of vitamin D and calcium	Historically believed to be a strong etiological factor in scoliosis. Spinal deformity will occur in bipedial rats on a rachetogenic diet [8]. Human scoliosis is rarely if ever caused by dietary deficiency rickets [7]
Vitamin D-dependent rickets *#264700 (Type I)* *#277440 (Type II)* *%600785 (Type II with normal vitamin D receptor)* *#193100 (autosomal dominant type)*	Type I – AR, 12q13; genetic deficiency of enzyme 1-alpha-hydroxylase Type II – abnormality of calcitriol receptor; end-organ resistance	Type I – low levels $1,25(OH)_2$ vitamin D Type II – increased levels $1,25(OH)_2$ vitamin D; severe hypocalcemia	Type I – Calcitriol Type II – Calcitriol, calcium	Spinal deformity rarely associated, possibly because of decreased growth velocities
X-linked hypophosphatemic rickets *#307800* *Autosomal dominant* *#193100*	X-linked dominant, PHEX gene, Xp22 coding for a protease AD, *FGF23* gene, 12p13	Hypophosphatemia, decreased levels $1,25(OH)_2$ vitamin D; normal calcium	Calcitriol; phosphate	Spinal deformity rarely associated, possibly because of decreased growth velocities
Lowe's syndrome *#309000*	Mutation in *OCRL1* gene; Xp26; deficiency of phosphatidylinostitol 4,5-biphosphanate 5-phosphatase	Phosphaturia, hypophosphatemic rickets; aminoaciduria; decreased ammonia production by kidney – acidosis; carnitine wasting; cataracts; MR	Replacement therapy – phosphate, carnitine, alkali; supportive care	May develop increased lordosis and occasionally scoliosis [14]
Hypophosphatasia *#241500 (infantile)* *#241510 (childhood)* *#146300 (adult and odonthypophosphatasia forms)*	Defect in alkaline phosphatase production	Identified in perinatal, infantile, childhood, adult, and odontohypophosphatasia forms. Perinatal hypophosphatasia lethal. Infantile form has a roughly 50 % mortality rate with symptoms appearing within the first 6 months after birth. The other forms are generally nonlethal. Adult form and odontohypophosphatasial form are marked by premature teeth loss. Common symptoms include bone malformations and higher chance of bone fracture		Isolated cases reports of scoliosis in infantile form [11, 15]. Suggested as a genetic linkage rather than a common etiology [12]

(continued)

Table 19.1 (continued)

Disorder and OMIM link	Pathophysiology and inheritance	Typical features	Treatment	Association with spinal deformity
Osteogenesis imperfecta	Over 100 different mutations lead to OI	Incidence 1:10–20,000		
Type I OI *166240 (IA), #166200 (IB)*	AD. COL1A1 (chromosome 17) or COL1A2 (chromosome 7) functional null alleles causing reduced amounts of normal collagen I	Mildest and most common form of OI. Blue sclera, conductive hearing loss, with or without dentigenesis imperfecta	Bisphosphonate treatment currently with either pamindronate or alendronate for more severe cases	Occurrence of scoliosis is related to the severity of bone involvement
Type II OI *166210*	Typically new mutation in either the *COL1A1* gene or the *COL1A2* gene	Fatal perinatally with intrauterine fractures, intracranial hemorrhage		Fatal too early for scoliosis
Type III OI *#259420*	AD, rarely AR. Mutation in most cases lies in one of the genes for type I collagen, *COL1A1* or *COL1A2*	One-eighth as common as type I. Severe involvement with progressive deformities, dentigenesis imperfecta, hearing loss, easy bruising, triangular facies	Bisphosphonate treatment currently with either pamindronate or alendronate	Scoliosis is very common. Kyphosis may also occur but is less common
Type IV OI *#166220*	AD. COL1A1 or COL1A2	Similar to type I without blue sclera and more severe osseous involvement	Bisphosphonate treatment currently with either pamindronate or alendronate	Similar to type III
Type V OI *%610967*	AD. Rare. Uncertain molecular mechanism	[12, 16] Similar to type V but with hyperplastic callus formation at fracture sites, calcification of the interosseous membrane between the radius and ulna, and the presence of a radio-opaque metaphyseal band adjacent to the growth plates	Bisphosphonate treatment currently with either pamindronate or alendronate	Similar to type III
Type VI OI *%610968*	AD with parental mosaicism. Rare. Uncertain molecular mechanism	[17] Similar to type IV	Bisphosphonate treatment currently with either pamindronate or alendronate	Similar to type III

Type VII OI #610682	Eight affected individuals in a small consanguineous First Nations community in northern Quebec. Mutation in the *CRTAP* gene (also cause type IIB)	Bone fragility and low bone mass	Uncertain	Uncertain
Type VIII OI #610915	AR. Mutation in the gene encoding leprecan (*LEPRE1*)	Lethal	Uncertain	Uncertain.
Type IX OI #259440	AR. Abnormality in *PPIB* gene	Lethal	Uncertain	Uncertain
Type X OI #613848	AR. Mutation in the serpin peptidase inhibitor (SERPINH1)	Lethal	Uncertain	Uncertain
Type XI OI #610968	AR. Homozygous mutation in the *FKBP10* gene	Severe OI without dentogenesis imperfecta	Uncertain	Scoliosis in 5/8 reported patients
Type XII OI #613849	AR. Mutation in the *SP7* gene	Milder deformity without dentogenesis imperfecta. White sclera	Uncertain	Scoliosis reported
Type XIII OI #614856	AR. Homozygous mutation in the *BMP1* gene	Variable	Uncertain	Kyphoscoliosis (in some patients). S-curve scoliosis of thoracic and lumbar spine (in some patients)
Type XIV OI #615066	AR. Homozygous mutation in the *TMEM38B* gene	Variable	Uncertain	Uncertain
Type XV OI #615220	AR. Homozygous or compound heterozygous mutation in the *WNT1* gene	More severe, blue sclera	Uncertain	Scoliosis reported
Other osteopenic syndromes				
Bruck syndrome #259450	Rare. Possibly gene encoding bone telopeptidelysyl hydroxylase	Bone similar to type I OI, multiple joint contractures, severe scoliosis [16–18]	Uncertain	Severe scoliosis similar to OI type III
Osteoporosis-pseudoglioma syndrome #259770	Familial gene encoding low-density lipoprotein receptor-related protein-5	Blindness. Brittle bones	Uncertain	Scoliosis is reported
Idiopathic juvenile osteoporosis #259750	Uncertain etiology	Idiopathic osteoporosis which resolves with adolescence	Uncertain. Protect spine	May develop spine compression fractures

(continued)

Table 19.1 (continued)

Selected Mucopolysaccharidoses				
Mucopolysaccharidoses	Pathophysiology	Typical features	Treatment	Association with spinal deformity
Type I (Hurler's syndrome) *#607014*	Gene encoding alpha-l-iduronidase	Course features are evident early. Bulging fontanels, neurological compression, corneal clouding; upper airway obstruction; pulmonary edema postoperative; short stature; carpal tunnel	Bone marrow transplant Enzyme therapy may help ? Gene therapy	Gibbus deformity lower spine. Upper cervical instability, odontoid hypoplasia and compression from instability or dural and ligamentous hypertrophy may occur
Type II (Hunter syndrome) *+309900*	Deficient activity of iduronate 2-sulfatase X-linked; mapped to Xq27–28 Dx: Enzyme assays in cultured fibroblasts/leukocytes Increased urinary heparan and dermatan sulfate	Two forms exist (Type A – severe; Type B – mild) Type A – clinical features as in Type IH; onset age 1–2 years; death in adolescence, third decade Type B – may be diagnosed in adulthood Course facial features; hearing loss; mental retardation (Type A); absence of corneal clouding; upper airway obstruction; pulmonary edema postoperative; HSM; dysostosis multiplex; short stature; HCP; carpal tunnel; ivory skin lesions; Mongolian spots; hypertrichosis	Bone marrow transplant Enzyme therapy may alter disease progression; not curative Idursulfase ? Gene therapy	Similar to other mucopolysaccharidosis (MPS)
Type III (Sanfilippo syndrome) *#252900* *#252920* *#252930* *#252940*	Type A – deficiency of heparin *N*-sulfatase (17q25.3) Type B – deficiency of alpha-N-acetylglucosaminidase (17q21) Type C – deficiency of aceyl-CoA:alpha-glucosaminide acetyltransferase (14) Type D – deficiency of *N*-acetylglucosamine-6-sulfatase (12q14) Dx: Increased urinary heparan sulfate	Type A – most severe, aggressive Behavioral issues at 2 years, neurological manifestations at 6 years Death in second decade Corneal clouding not a common finding; attentional/behavioral problems; hyperactivity; seizures; diarrhea; URI	Supportive care Not improved by bone marrow transplant	Similar to other MPS

Type IV (Morquio syndrome) Numerous types	Type A – deficiency of galactosamine-6-sulfatase (GALNS gene, 16q24.3) Type B – β-galactosidase (GLB1 gene, 3p21.33) Increased urinary excretion of keratin sulfate (cartilage/cornea) mild may not excrete ELISA Enzyme assay in cultured fibroblasts/leukocytes Genetic testing to detect mutations in *GALNS*, *GLB1*	Clinically, both forms may be similar; great variability in severity within both groups Mortality related to atlantoaxial instability, myelopathy and pulmonary compromise For severe, death in second or third decade of life No course facial features; normal intelligence; spondyloepiphyseal dysplasia; ligamentous laxity; odontoid hypoplasia; shortened trunk dwarfism; genu valgus; greater incidence of spinal involvement; bowel/bladder incontinence; OSA; pulmonary infections – chest wall deformity; heart valve thickening/defects; corneal clouding; enamel Less common – hearing loss, hernias	Supportive treatment	Similar to other MPS. These patients survive long enough that orthopedic treatment, particularly for cervical instability may be necessary
Type VI (Maroteaux–Lamysynd) #253200	AR Deficiency of *N*-acetylgalactosamine 4-sulfatase Accumulation of dermatan sulfate	Acceleration in growth during first year; followed by regression and short stature; course features, HSM, corneal clouding; normal intelligence Other: hearing loss, respiratory infections, valvular disease	Bone marrow transplant reported as successful. Enzymatic treatment approved	Similar to other MPS
Type VII (Sly disease) #253220	Deficiency of β-glucoronidase, required for degradation of dermatan sulfate, heparin sulfate, and chondroitin sulfate Mutation in gene mapped to chromosome 7	Phenotype varies; severe form (subtype 1) present at birth – jaundice, anemia, hydrops; milder forms present later, before (subtype 2) or after (subtype 3) age 4 years; course features; HSM; hernias; MR; dysostosis multiplex	Bone marrow transplant reported as successful in improving daily function but not the mental retardation	Similar to other MPS

with thoracic scoliosis. Widmann et al. [19] found a high negative correlation between pulmonary function and thoracic scoliosis and but not with chest wall deformity or kyphosis. They also found diminution in vital capacity below 50% in thoracic curve of 60° or more.

Spinal deformity in OI is directly related to the severity of osseous involvement [20–22], though body weight is also a factor in bone density [23]. Anissipour et al. [20] recently evaluated progression of scoliosis in a large cohort of patients with various types of OI and found the most rapid progression in the more severe type III. While ligamentous laxity, also from type I collagen deficiency, may also contribute, a series of patients with various types of OI found less scoliosis in those with ligamentous laxity [21]. There is a tendency for later development of scoliosis in those with early development of motor milestones, particularly supported sitting [21]. Overall, the worse the osseous involvement in terms of intrinsic vertebral body deformity and decreased *Z*-scores for bone density, the worse the scoliosis and the more difficultly achieving good fixation for correction [22, 23]. The morphology of the vertebral bodies ranges from normal contours to flattened, wedged, and biconcave with the biconcave vertebra more likely to develop severe scoliosis [23–25]. Six or more biconcave vertebra before puberty appears prognostic of developing scoliosis of greater than 50° [23]. The spine may also become kyphotic particularly [26] in those with more severe involvement [27]. Cervical fractures [28] including spontaneous paraplegia from chiropractic manipulation [29], thoracic fractures with spinal cord injury [30], and multiple flexion stress endplate fractures of the thoracic and lumbar spine [31] have been reported. Daivajna et al. [32] reported a modified anterolateral approach for decompression of myelopathy from severe cervical kyphosis in a 9-year-old with OI.

Bracing has been attempted for spinal deformity from OI. However, the corrective force applied through the pathological ribs only leads to further rib deformity and may contribute to worsening pulmonary function [33–35]. Current braces cannot achieve this. Patients who are not operative candidates can be fitted with custom seating for comfort and function [36, 37]. Bracing's role in OI is primarily limited to postoperative temporary support though there is some suggestion that an orthosis can slow the progression of basilar impression [3].

Positioning patients for surgery can be very difficult and must be carefully supervised because of chest wall deformities, fragile extremities, ribs, and frequent contractures. Unpadded blood pressure cuffs can result in fractures. Surgery for these patients should typically occur in institutions accustomed to their care [38]. Fatality has been reported from intraoperative rib fractures [39]. Classically, correcting curves in severely soft bone has been difficult at best with most authors recommending stabilization rather than relying upon correction [34]. The stability of fixation depends upon both the strength of the bone and the quality of the fixation's purchase. Polymethylmethacrylate has been used for additional fixation [33, 40, 41]. Historically, each new type of fixation has been attempted. As might be expected, the greater the purchase and the more segmental the fixation, the less stress required on any single level and the greater the correction achieved [42] (Figs. 19.1a–c and 19.2a–d). Preoperative halo gravity traction using multiple pins may be useful [43, 44] though both sixth and fourth nerve palsies have been reported in OI patients [45]. Postoperatively, there is little change from preoperative ambulatory ability or activity [34, 37, 46], though patients report less pain, fatigue, and dyspnea [47], and the improvements gained at surgery are usually maintained into adulthood [43, 47]. Spondylolysis [48] or an elongated pars with spondylolysthesis [49] can occur below a long spinal fusion. Anterior interbody fusion has been described [49], but the spondylolisthesis may not be symptomatic (Fig. 19.3).

Basilar invagination with the odontoid protruding above Chamberlain's line is often difficult to see in these children with short necks, wide chests, and poor bone quality. If not present in preschool films, it is not likely to develop over time and further routine screening is probably unnecessary [50]. If it is suspected, an MRI can

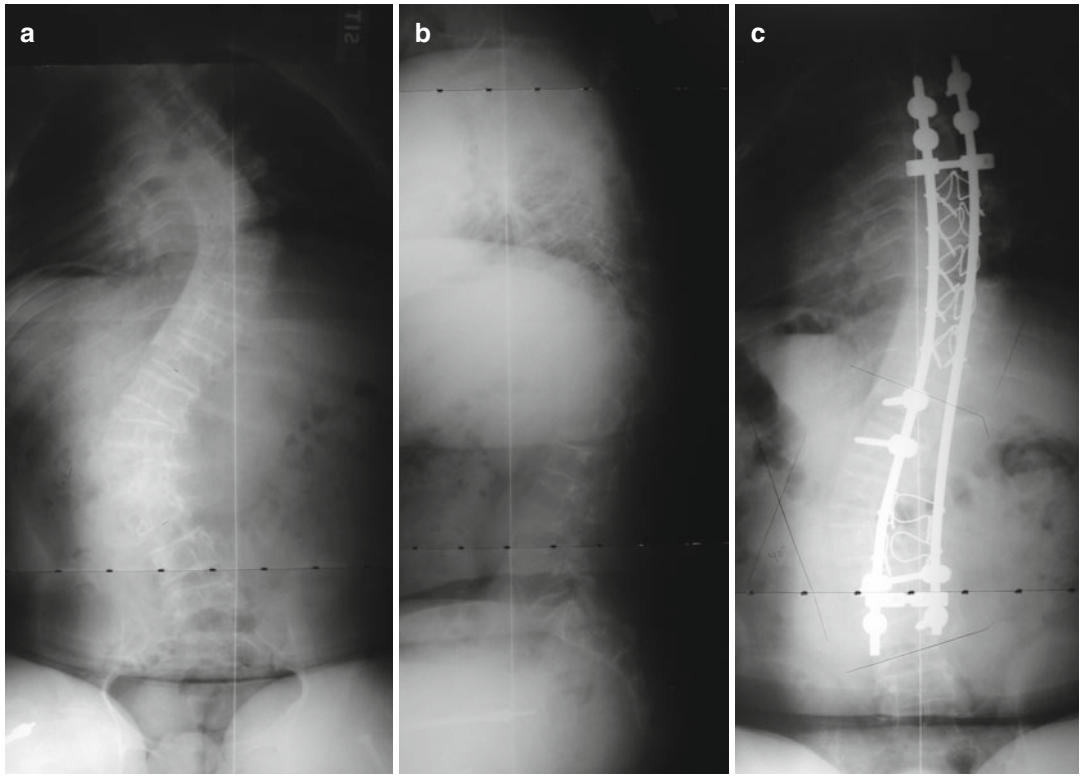

Fig. 19.1 (**a**, **b**) Type III OI scoliosis AP and lateral. (**c**) Same patient as in (**a**, **b**) with multiple fixation types to maximize bone. Patient had been pretreated with bisphosphonates

be helpful [51] (Fig. 19.4a, b). If present, the follow-up is individualized. Basilar impression from platybasia can be very difficult to treat and shunting may be necessary [41]. The symptoms typically develop in early adolescents and the signs include headache, lower cranial nerve problems, hyperreflexia, quadriparesis, ataxia, nystagmus, although many patients remain asymptomatic [3]. It occurs primarily in those with more severe involvement and kyphosis, particularly with type III OI [27]. In general, patients with reducible deformities can be treated with instrumentation transferring the weight of the head to the cervical [52, 53] or thoracic spine [52]. When reduction is not feasible, transoral [52] and an extended maxillotomy approach [54] for anterior decompression both have been used. More recently, successful endoscopic decompression has also been described [55]. Unfortunately, the basilar invagination can progress despite a solid fusion [3]. Prolonged immobilization, particularly during adolescence, with a custom Minerva may help improve symptoms and slow progression [3]. Our current strategy is to manage minimally symptomatic patients with bisphosphonates though there is no evidence published about its effectiveness.

In our experience, bisphosphonate therapy seems to improve the bone quality for fixation and certainly seems to improve the patients' bone pain and overall quality of life [56, 57]. Pamindronate may actually help reverse some of the pathological changes of the deformed bone [58] and may prevent scoliosis in some younger children [59]. Postoperative bisphosphonates have been suggested because progression may occur after surgery [47, 60]. Bone mineral density continues to improve for up to 2 years after pamidronate discontinuation, but not as much as in those with continued treatment [60]. Bisphosphonates should be continued until skeletal maturity is reached.

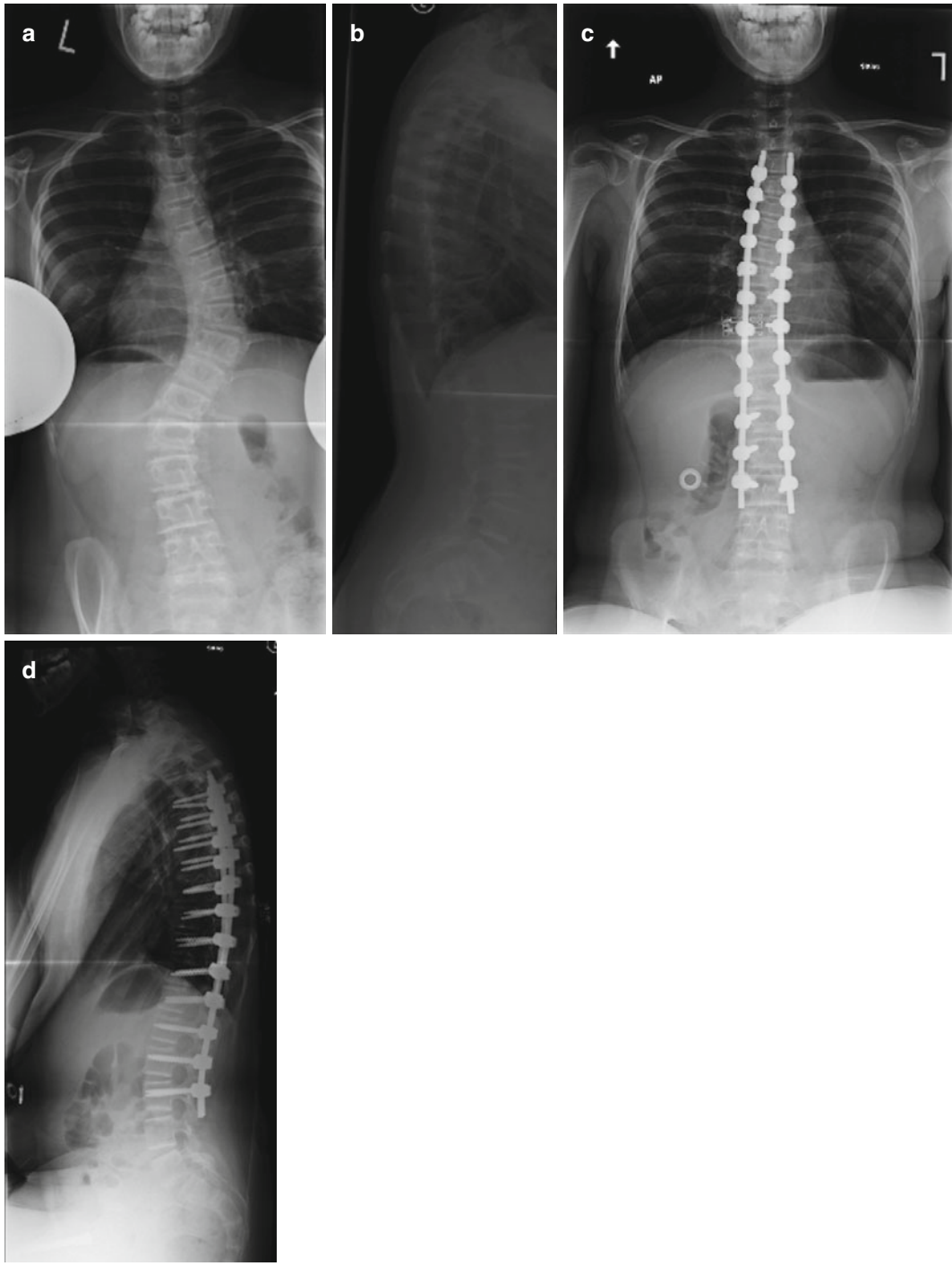

Fig. 19.2 (**a**–**d**) Patient with more severe type I OI scoliosis treated with multiple screws after bisphosphonate treatment

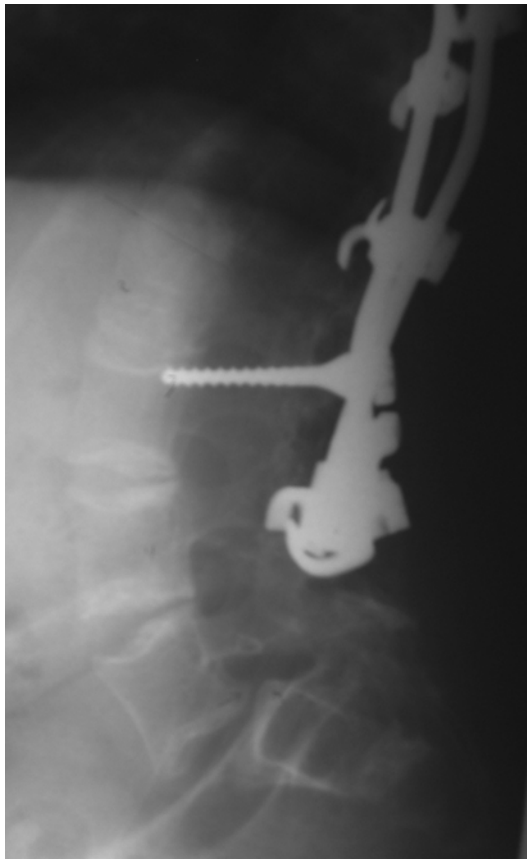

Fig. 19.3 Spondylolisthesis occurring below instrumentation for scoliosis in a patient with type III OI

19.5 Storage Diseases

The mucopolysaccharidoses (MPS) constitute a family of storage diseases with spinal abnormalities similar to those seen in spondyloepiphyseal dysplasia. In these disorders, enzymatic abnormalities prevent the normal breakdown of glycosaminoglycans with subsequent accumulation within lysosomes. Mucopolysaccharidoses include a group of disorders that occur when specific lysosomal enzymes are deficient. Lysosomal enzymes are responsible for the degradation of glycosaminoglycans (GAGs), long-chain carbohydrates comprising a major component of connective tissue. The different types of MPS vary in severity as well as in clinical manifestations. Some features that many of the MPS share include course facies, skeletal involvement – dysostosis multiplex and short stature, organomegaly, corneal opacification, and varying degrees of mental retardation. Table 19.1 lists the different types of MPS, as well as their pathophysiology, clinical manifestations, and treatment. Enzymatic treatment can prevent the mental deterioration many of these patients demonstrate but may not significantly change their musculoskeletal problems.

The various types are listed in the Table 19.1. At birth the children are normal, but the accumulated glycosaminoglycan products produce soft tissue swelling which can cause direct spinal cord compression particularly around the craniovertebral junction and odontoid [61–71]. They may also have cervical compression from odontoid hypoplasia [68, 72, 73] and atlantoaxial instability [69, 72, 74]. A thoracolumbar gibbus is often present on presentation with a classic bullet-shaped vertebra (Fig. 19.5). Occasionally, thoracic or thoracolumbar gibbus can be progressive, cause spinal cord compression, and require decompression and fusion [68, 75, 76]. Despite the improvements in MPS I (Hurlers) from bone marrow transplant, spinal abnormalities may persist because of poor enzymatic penetration of bone and require treatment [77]. These children pose substantial anesthesia risks because of their enlarged pharyngeal soft tissues [61, 78–80].

19.6 Juvenile Osteoporosis

Juvenile osteoporosis is an unusual but self-limiting disease, which may result in pathological kyphosis. The diagnosis is one of exclusion particularly looking for malignancy. Results of medications including bisphosphonates, calcitriol, fluoride, and calcitonin are equivocal with mixed results reported [81–86]. Treatment is aimed at protecting the spine until remission [83] and the patients should be referred to an endocrinologist.

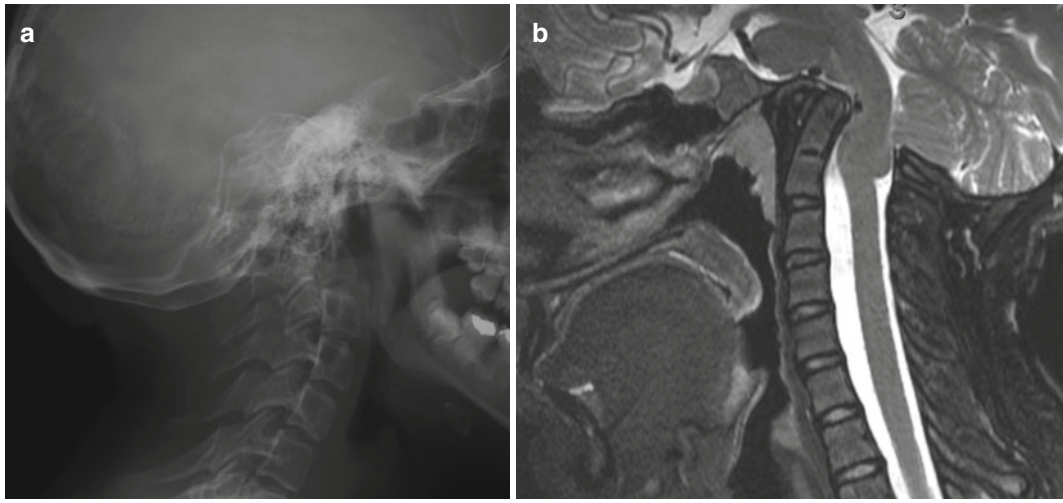

Fig. 19.4 Basilar invagination in a patient with type IV OI on plain radiographs (**a**) and MRI (**b**). The patient is asymptomatic

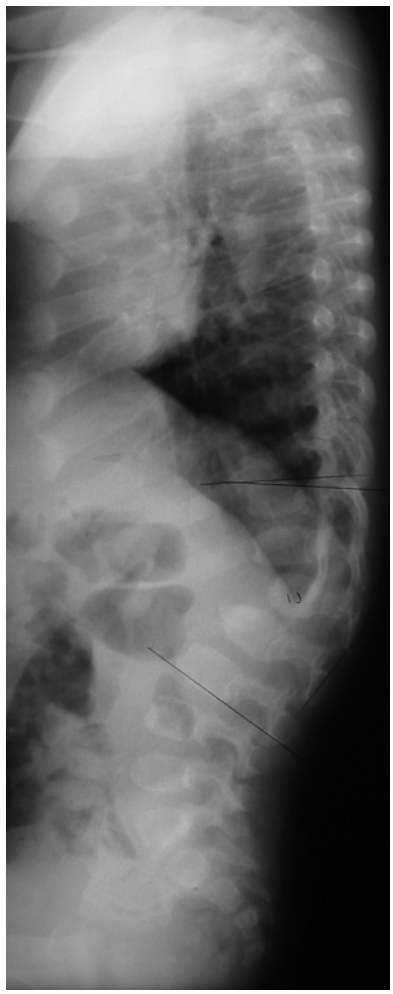

Fig. 19.5 Wedge-shaped vertebrae in patient with Morquio's MPS

19.7 Anorexia Nervosa

Anorexia nervosa can cause decreased bone density and is most frequent in the same population as girls with idiopathic scoliosis and should be considered in patients with a low BMI and osteopenia. It can result in pathological kyphosis from multiple compression fractures [87–90].

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Spinal Manifestations of Skeletal Dysplasias

20

Leok-Lim Lau and William Mackenzie

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Key Points

- If the diagnosis is known, the typical spinal deformities present can be expected and managed early in the course of evolution.
- All skeletal dysplasias other than achondroplasia can have upper cervical instability and stenosis and this needs to be evaluated in every child.
- Restrictive lung disease is present in many forms of skeletal dysplasia and it is essential to preserve thoracic growth in these children.
- Spinal stenosis is very common and must be evaluated when surgical management involves instrumentation that may enter the canal.
- When managing sagittal, coronal, and rotational deformity of the spine, contractures about the pelvis and lower extremities must be considered.

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20.1 Introduction

Skeletal dysplasias are a diverse group of conditions that affect cartilage and bone development. Spinal involvement is common and manifest in a variety of ways. Commonly, the affected patients have a mature height of less than 2 standard deviations on the normative chart. Each of the conditions is relatively rare. Collectively, it is estimated that 2.3–7.6 per 10,000 population are affected. This is comparable to the prevalence of cystic fibrosis, neural tube defects, or Down syndrome. The most common skeletal dysplasia is achondroplasia which is readily recognizable and can be diagnosed by practicing orthopedists. Others may be more exotic. Online resources such as Orphanet or Online Mendelian Inheritance in Man (OMIM) provide updated clinical information. This chapter aims to provide a framework to assess and manage spine problems of the common skeletal dysplasias that affect cartilage predominantly. It is not the aim of this chapter to give an exhaustive description of each type of skeletal dysplasia. Relevant spinal manifestations are distilled to provide insight to the management.

20.2 Nomenclature and Classification

The plethora of the phenotypes and genotypes of skeletal dysplasia result in 33 groupings, under which more than 400 conditions are subclassified [1]. Achondroplasia falls under the FGFR3 group with five other conditions including thanatophoric dysplasia, SADDAN, hypochondroplasia. OMIM codes each of the conditions with a six-digit number. For instance, achondroplasia is coded [100800]. The [100000] to [200000] range denotes autosomal-dominant phenotype. The systematic classification allows better communication among clinicians and researchers.

Skeletally, the patients can be affected proportionately and disproportionately. In the latter group, it could affect predominantly axial skeleton (such as brachyolmia) or appendicular skeleton. In the appendicular skeleton, shortening of the hand, forearm, and arm are named acromelic, mesomelic, and rhizomelic, respectively. See Table 20.1 for summary for the conditions affected by the subcategory.

The following broad groups (Table 20.2) are the conditions seen in a skeletal dysplasia clinic.

Table 20.1 Disproportionately short (Dwarf)

Short trunk
Brachyolmia spondylodysplasia
Short appendicular skeleton
Acromelic (short hand)
Acromelic dysplasia
Mesomelic (short forearm)
Dyschondrosteosis (Leri Weill)
Rhizomelic (short arm)
Achondroplasia and related disorders
Achondroplasia
Hypochondroplasia
Short trunk and limb
Spondyloepiphyseal dysplasia
Kniest dysplasia

Table 20.2 Relative common skeletal dysplasia groups

FGFR3 mutation
Hypochondroplasia, achondroplasia, SADDAN (severe achondroplasia, developmental delay, acanthosis nigricans), thanatophoric dysplasia
Metatropic dysplasia group and SMED
Diastrophic dysplasia
Type 2 collagenopathies
SED congenita, SEMD, Kniest dysplasia
Pseudoachondroplasia
Multiple epiphyseal dysplasia
Metaphyseal dysplasia
Cartilage hair hypoplasia (McKusick), Schmidt, Jansen
Chondrodysplasia punctata
Dysostosis multiplex group
Mucopolysaccharidoses
Mesomelic dysplasia
Leri–Weil dyschondrosteosis
Dysplasia with predominant membrane bone involvement
Cleidocranial dysplasia
Bent-bone dysplasia group
Campomelic dysplasia

Significant overlapping occurs within the group. Precise diagnosis is determined in consultation with an experienced geneticist.

20.3 Spinal Problems in Skeletal Dysplasia

Spinal problems in skeletal dysplasia include instability, sagittal and coronal deformity, and stenosis. Some deformities are transient, for example, thoracolumbar kyphosis in achondroplasia which typically resolves at onset of ambulation and cervical kyphosis in diastrophic dysplasia. Cervical instability and stenosis in mucopolysaccharidoses is progressive that require early recognition and intervention to prevent or reverse neurological deterioration.

20.3.1 Base of Skull Abnormalities

Achondroplasia is a rhizomelic, disproportionate short statue with frontal bossing and mid-facial hypoplasia. In early life, they are at risk for hydrocephalus. Symptoms include irritability, lethargy, and vomiting. Head circumference measurement should be performed in reference to the normative chart of people with achondroplasia. Any upward crossing of the percentile would warrant further investigation with magnetic resonance imaging (MRI) scan of the brain. The defective endochondral ossification in patients with achondroplasia from the FGFR-3 mutation and early closure of synchondroses leads to foramen magnum and jugular foramen stenosis. It is thought that diminished jugular return due to jugular foramen stenosis results in hydrocephalus [2]. With growth, the risk diminished significantly. Management includes a ventriculoperitoneal shunt.

Platybasia literally means flattening of the skull base. It has been reported in patients with Kniest dysplasia [3] and cleidocranial dysostosis [4]. The sagittal relationship between the anterior and posterior fossa at the skull base (as measured by the base of skull angle, normal range <143°) pathologically widen. Indirectly, the relationship of the posterior fossa with upper cervical spine is altered as measured by clivus-canal angle (also known as craniovertebral angle, normal range 150–180°). Basilar invagination can ensue with the odontoid impinging on the ventral aspect of the cervicomedullary junction. In patients with cleidocranial dysostosis, the skull base angle is more likely to be wider than the normal population, possibly related to the abnormal flexure of the clivus, a midline structure that is malformed in conjunction with the clavicle and pubis [5].

20.3.2 Atlantoaxial-Occipital Complex Abnormalities

20.3.2.1 Developmental Anatomy

The upper cervical spine forms differently from the subaxial spine embryologically and is closely linked to the occiput [6]. The C1 atlas has three primary ossification centers (anterior atlas, arch, and two lateral masses). The closure of posterior synchondroses occurs at 3–5 years. An opening at the cartilaginous cleft can be found prior to the closure of the neural arch [7]. The closure of neurosynchondroses of the anterior atlas arch and the two lateral masses achieved by 5–7 years old [8]. The spinal canal area at C1 expands rapidly in the first 3 years of life with no further significant expansion beyond 6 years of age [9].

C2 has five primary ossification centers (two lateral masses, centrum, odontoid dens that is formed from two vertically aligned columns). C2 has secondary ossification centers that are located at the tip of odontoid, the roof and the base of the centrum, and the inferior ring apophysis. The atlantoaxial column is developed from separate embryological entities named X, Y, and Z. They become chondrum terminale, odontoid dens, and centrum of C2, respectively. Failure of fusion of XY and Z by ages of 5–7 results in an osodontoideum [10]. Failure of fusion of X and YZ by age of 12 results in an ossiculum terminale. The spinal canal expands rapidly in diameter prior to closure of the neurosynchondroses from birth to 8-year-old, but attains significant width by 2 years [11].

20.3.2.2 Foramen Magnum Stenosis and Cervical Stenosis

Achondroplasia classically has foramen magnum stenosis and cervical stenosis both in the upper and subaxial cervical regions. The symptoms may be subtle. The symptoms and signs include developmental delay, central apnea, neurological signs (hypotonia, hyperreflexia, clonus, hemiplegia, tetraplegia), or a combination of the above. Risk of sudden death is high when the stenosis is undetected [12]. Similar to the hydrocephalus, the risk is highest in the first 2 years of life and diminishes with the expansion of the spinal canal. In the suspected case, sleep study (central apnea) and MRI (Fig. 20.1) help to clarify the situation. Foramen magnum decompression is indicated in symptomatic patients with documented stenosis.

20.3.2.3 Occipitalization of C1 and Pro-Atlas

C1 abnormalities are closely linked with occipital development. Failure of segmentation at the occipital sclerotome 4 and cervical sclerotome 1 result in occipitalization of C1 and potential basilar invagination. Occipitalization is reported in patients with skeletal dysplasia such as Goldenhar syndrome and Russell-Silver syndrome [13].

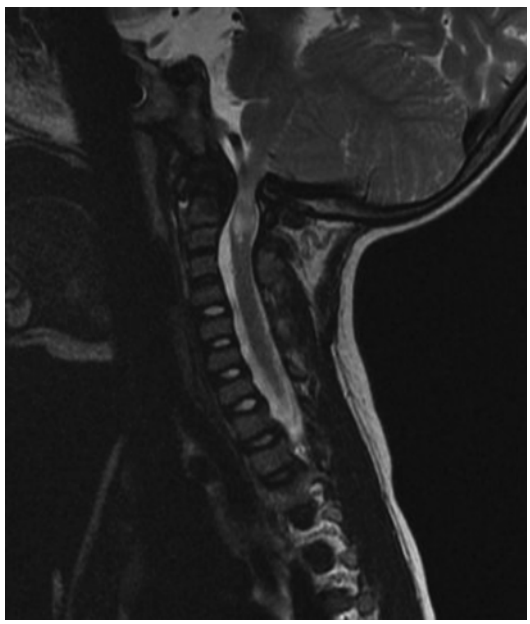

Fig. 20.1 Foramen magnum stenosis in achondroplasia. The MRI showed significant cervical magnum stenosis with cervical myelopathy in a patient with achondroplasia

20.3.2.4 Upper Cervical Spine Instability

Upper cervical spinal instability includes occipito-cervical instability (O–C1) and/or atlantoaxial instability (C1–C2). This should be suspected in all cases of skeletal dysplasia with the exception of achondroplasia. The spinal instability may result in cervical myelopathy. The clinical manifestation ranges from asymptomatic to tetraparesis. In older children, the earliest symptom may be gradual loss of physical endurance. In toddler and younger children, gross and fine motor developmental delay can occur.

O–C1 stability is determined by ligamentous strength. Uniquely, there is lack of an intervertebral disk at this segment. The bony articulation of the atlas and occipital condyles are shallow and broad with a relatively lax capsule to allow motion. The cord-like alar and apical ligaments arise from the odontoid to the anteromedial aspects of the occipital condyles, and the tectorial ligament which is a continuation of the posterior longitudinal ligament to basion, are the most important internal ligaments at this joint. Instability at this joint is thought to be rare and includes Kniest dysplasia which is a type 2 collagenopathy [14].

C1–C2 stability depends on combinations of bony and ligamentous restraints. It is thought that the instability and associated delayed in ossification often seen in skeletal dysplasia result in odontoid abnormalities that ranges from odontoid aplasia, hypoplasia, to os odontoideum [15]. Failure of closure at the dentocentral synchondroses by 7 years results in os odontoideum. The line of separation may be caudad or cephalad to the superior articular facets of the axis (C2). This is in contrast to traumatic cause of dentocentral separation where the separation is caudad to the superior articular facets of the axis. The instability may result in delay or failure of ossification of chondrum terminale and anterior arch of the atlas. Dentocentral separation has been reported in chondrodysplasia punctata [16].

Abnormal appearance of the odontoid radiographically can be an indication of potential

underlying upper cervical instability. However, not all abnormal odontoid morphology leads to instability. Skeletal dysplasias associated with odontoid abnormalities include pseudoachondroplasia, MPS, Cartilage-Hair dysplasia, diastrophic dysplasia, metatropic dysplasia, Larsen syndrome, and type 2 collagenopathy (SED, Kniest Dysplasia). Congenital nasopharyngeal abnormalities such as cleft palate may be associated with upper cervical spine abnormalities such as that is seen type 2 collagenopathy [14].

In atlantoaxial instability, there is either posterior or anterior subluxation of C1 on C2. Posterior subluxation of C1/C2 occurs when the atlas overrides the centrum of axis. This is a more uncommon phenomenon due to the odontoid process acting as a posterior constraint; however if there is odontoid aplasia (uncommon) or osodontoideum, there is mobility. Anterior subluxation can be due to odontoid abnormalities, ligamentous laxity, or absence of ligaments. In a lordotic cervical spine, a fixed posterior subluxation is probably more tolerable than an immobile anterior subluxation.

Cervical spine instability may not cause symptomatic spinal cord compression. In a normal C1 spinal canal, Steel's rule of third dictates that one-third of the space is occupied by dens, one-third by spinal cord, and one-third is a reserve space that may be sufficient to buffer the instability created by a pathology. In Down syndrome for instance, symptomatic atlantoaxial instability is only present in 18 % of the patients with atlantoaxial instability [17]. Significant atlantoaxial instability (ADI>8 mm) without symptoms is said to be an absolute indication for surgery while moderate atlantoaxial instability (ADI 4–8 mm) without symptoms is a relative indication for surgery. The authors do not agree. The most important factor is spinal cord compression and instability without spinal cord compression with a capacious canal does not require surgical management.

The surgeon must be aware of the concomitant presence of extradural impingement (see following section) or an abnormally small spinal canal at C1 which may be seen in spondyloepiphyseal dysplasia congenita [18] or metatrophic dysplasia [19]) (Fig. 20.2). Laminectomy of C1 may be necessary to create extra room to accommodate the spinal cord [18, 19]. In a detailed dynamic CT myelography study of patients with Type IV mucopolysaccharidoses, the source of impingement of the spinal cord was shown to arise from the abnormally thick posterior neural arch globally or at the unossified posterior neurosynchondroses focally [15]. It is our experience that cervical myelopathy presents earlier in patients with spondyloepiphyseal dysplasia congenita compared to patients with Morquio syndrome.

20.3.2.5 Abnormal Extradural Impingement

Extradural impingement of the spinal cord may be observed in mucopolysaccharidoses (Fig. 20.3). This is almost universal in mucopolysaccharidoses type IVa (Morquio syndrome) and to a lesser extent in type IH (Hurler syndrome) and type VI (Maroteux-Lamy syndrome). The glycosaminoglycan accumulation intracellularly results in mechanically incompetent cartilaginous tissue and ligamentous tissue that encourages reactive tissue formation. In cases where biopsy was performed via trans-oral approach, reactive tissue composed of fibrocartilaginous tissue accumulated extradurally without evidence of meningeal involvement [15]. It was also noted that the reactive tissue could extend caudadly to the dorsal aspect of the centrum of axis (C2). Extension to C3 and C4 is possible.

20.3.2.6 Torticollis: Atlantoaxial Rotatory Fixation/ Subluxation/Dislocation

In patients with metatropic dysplasia, torticollis may occur as part of a possible mechanism to protect the airway [19]. Patients present with torticollis in extension. This can develop postoperatively in patients who had C1–C2 decompression without fusion or when fusion fails to occur. Patients may have torticollis secondary to a unilateral lateral mass defect rather than ligamentous rotational instability [20].

It is postulated that recurrent upper viral respiratory infection involving Waldeyer's tonsillar ring seen in the Griesel's syndrome may

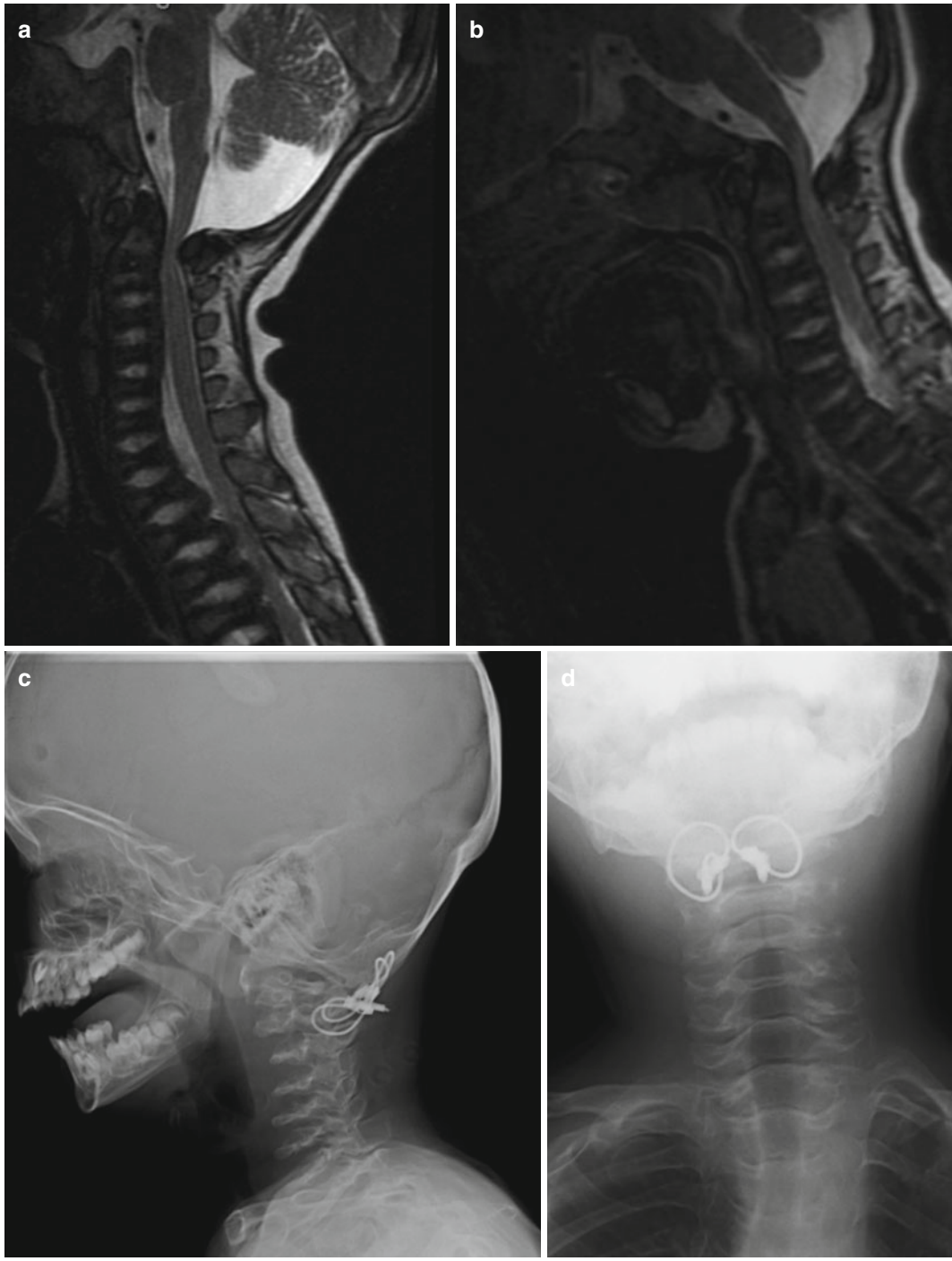

Fig. 20.2 C1 stenosis without instability in metatropic dysplasia. MRI views in flexion (**a**) and extension (**b**). C1 laminectomy was performed. Stabilization was achieved with posterior O–C2 arthrodesis using autologous bone graft and titanium cable. (**c**) Cervical X-ray lateral view and (**d**) cervical X-ray AP view postfixation

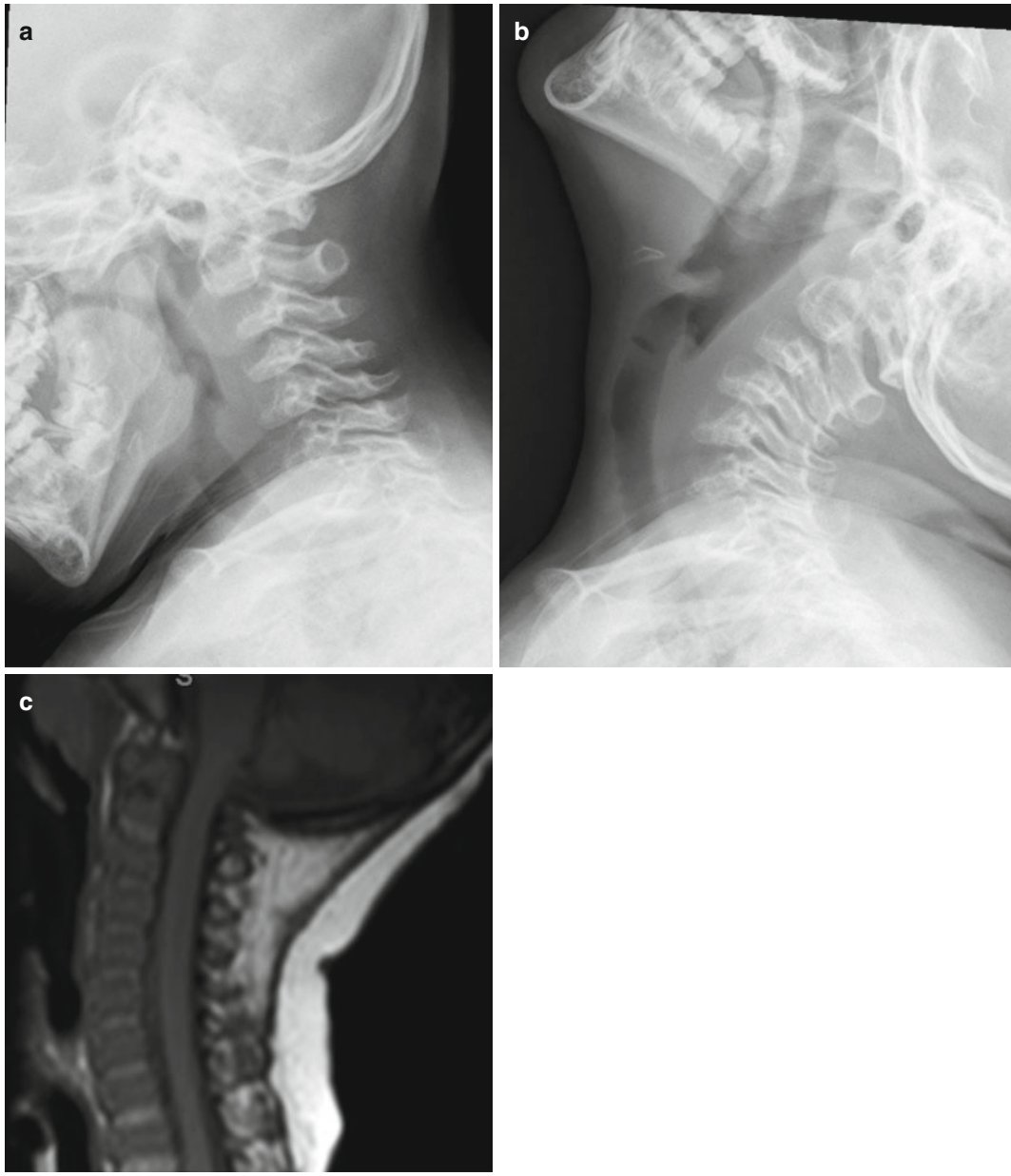

Fig. 20.3 Lateral cervical radiographs showing C1–C2 cervical instability in Morquio syndrome. SAC decreased in flexion (**a**) from extension (**b**). Extradural mass shown cranial to the hypoplastic odontoid process on T1 MRI cervical spine (**c**)

potentially predispose the at-risk patients with cleft palate or recurrent otitis media to atlantoaxial rotational subluxation. The hyperemic tissue at the posterior wall of nasopharyngeal space irritates the atlantoaxial joint asymmetrically resulting in instability. Atlantoaxial rotational subluxation after otoplastic surgery has been reported in patients without skeletal dysplasia (21). The diagnosis is confirmed by dynamic CT scan.

20.3.2.7 Radiological Signs of Upper Cervical Spine Instability

Radiological investigation using plain radiography may underestimate the underlying cervical instability. The occipitocervical junction is difficult to visualize. The immature bone with delayed ossification centers at various stages and abnormalities of the odontoid process compound the issue. When in doubt, dynamic MRI of the cervical spine should be performed [3] (Fig. 20.4). In very young children, this entails general anesthesia. MRI is done in neutral position first. Compression seen at this position may obviate the need for further dynamic positioning.

The standard measurement for O/C1 horizontal instability includes Power ratio, Basion-Dental interval, or Wiesel-Rothman method. The latter is the easiest to measure.

Vertical instability that results in basilar invagination is ascertained by Mc Rae's line, Chamberlain's line, McGregor's line, or Wachenheim's line. C1 involvement from occipital condylar hypoplasia, for example, is determined by Kaufman's technique.

C1/C2 horizontal instability is measured by anterior atlantoaxial distance, an indirect measure,

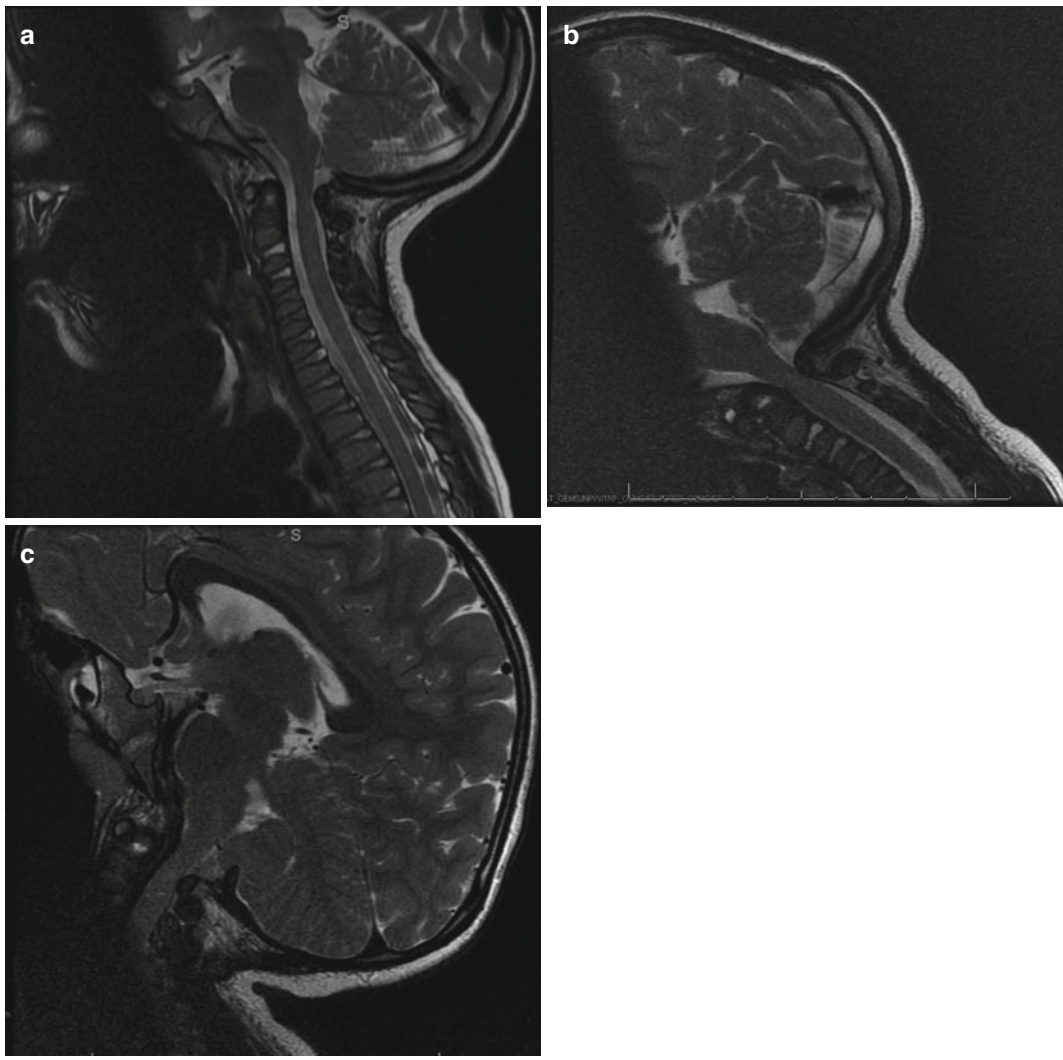

Fig. 20.4 Dynamic MRI. Space available for cord (SAC) in neutral position of the MRI (**a**). The stenosis is most severe in flexion view (**b**) and improved with extension (**c**)

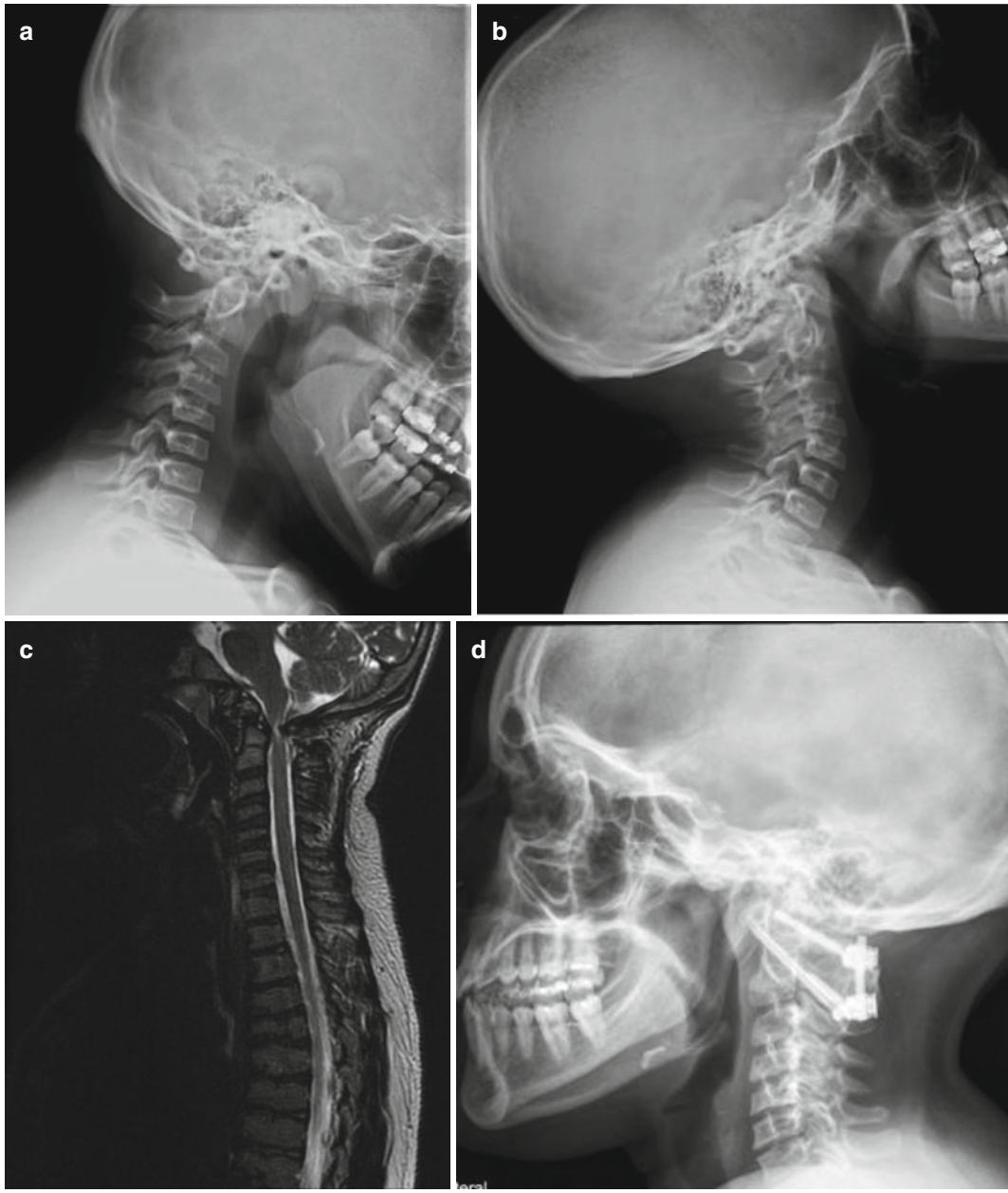

Fig. 20.5 SEDC with C1–C2 instability in flexion (**a**) and extension (**b**) views. MRI (**c**) shows cervical stenosis and myelomalacia. C1–C2 decompression, instrumentation, and fusion were done as shown in (**d**)

or posterior atlantoaxial distance (also known as space available for cord [SAC]).

20.3.2.8 Surgical Options

Cervical arthrodesis at C1–C2 include Gallie's technique, Brooks-Jenkins's technique, Sontag modification of Gallie's technique, Magerl's transarticular screw [21], and Harm's modification of Goel's technique [22]. Gallie's technique has poor rotational stability at the C1–C2 joint with a high rate of nonunion. Screw-based techniques (Magerl's and Harm's) are effective and can be used in young patients but careful preoperative evaluation of the vascular anatomy needs to be done (Fig. 20.5).

The Brooks-Jenkins's technique [23] and the Sontag modification of Gallie's technique [24] are effective wire-based techniques for C1–C2 arthrodesis. These two techniques are indicated for horizontal or vertical instability without the need for C1 ring decompression. C1 ring decompression is required if there is compression in a reduced position or if it is secondary to an irreducible subluxation. Fixation is ideally achieved with the Magerl or Harms technique but in young children this can be difficult especially if extension is needed to the occiput which may be needed in patients with spondyloepiphyseal dysplasia congenita, Morquio syndrome, or metatropic dysplasia.

To achieve occiput-cervical fusion after decompression, we used a modified occipital-based technique with a fashioned autologous iliac bone graft notched between O and C2 using cable (Fig. 20.2). Immobilization was performed using halo-vest postoperatively. No nonunion is reported [22]. Other techniques for O to C2 fusion include the use of rib graft and wire-based fixation of a fashioned iliac crest bone graft. Occipito-cervical fixation is often too bulky in young children.

20.3.3 Subaxial Cervical Spine Abnormalities

20.3.3.1 Developmental Anatomy

The subaxial cervical spine develops differently from the upper cervical spine. The subaxial cervical spine derives from sclerotomes C3 to C8. Primary ossification centers of the centrum first appear in the lower cervical/upper thoracic region and progresses caudad-cranially to C3. The centrum unites with the neural arch by 3 years of age. They are usually wedge-shaped with some loss of lordosis until about the age of 8–10-year-old. Ossification of the apophyseal rings appears at 10–12 years and the rings are fused by skeletal maturity.

20.3.3.2 Subaxial Cervical Spine Kyphosis and Stenosis

Subaxial cervical spine kyphosis should be screened for in patients with diastrophic dysplasia and Larsen syndrome. Other skeletal dysplasias with cervical spine kyphosis include Kniest dysplasia, chondrodysplasia punctata, and campomelic dysplasia [19, 25]. Rarely, Morquio syndrome may present with cervical kyphosis [26].

In mild cervical kyphosis, spinal alignment is altered without neurological consequence. With progression, the cervical kyphosis may result in instability with ventral cord impingement in flexion and relieved with extension. A significant deformity causes cervical stenosis in both flexion and extension. The progression of the deformity depends on the individual skeletal dysplasia and includes the magnitude of the deformity at presentation, the loss of anterior column support, the failure of facet and capsular restraints, associated posterior element deficiency, and the loss of the posterior tension band.

In diastrophic dysplasia, one-quarter of the patients have cervical kyphosis at birth. The kyphosis has its apex at C3 to C4, less commonly at C5 (Fig. 20.6). With the preservation of lordosis at the caudad level, a S-shaped or swan-neck appearance is evident. The vertebral body at the apex is hypoplastic resulting in triangular or round shape, also termed "loss of four corner" [27]. Concurrent spinal bifida occulta is often present from C3 to upper thoracic spine [28]. Most of the cases resolved spontaneously by the age of 6 years. In the largest series to date [27], cervical kyphosis of more than 60° (Fig. 20.7) at presentation had poor prognosis with either progression of the kyphosis or respiratory failure from associated severe tracheal and bronchiomalacia in early life.

Larsen syndrome may not be difficult to diagnose with the presence of multiple major joint dislocations (hips, knees, and elbows), joint laxity, and facial dysmorphism. An accessory calcaneal apophysis is the characteristic radiological sign. Screening cervical X-rays should be performed (Fig. 20.8). The findings include a hypoplastic anterior column at the apical vertebra, segmentation defects, and spinal bifida occulta. In the most severe cases, the associated ligament laxity, hypotonia, and posterior column deficiency result in spondylolisthesis and spondyloptosis [29]. The natural history of the cervical kyphosis is a progressive course [28]. Clinically, progressive hypotonia or loss of ambulation should not be attributed solely to joint dislocation or deformity. Consideration should be given to cervical myelopathy. Surgical intervention includes

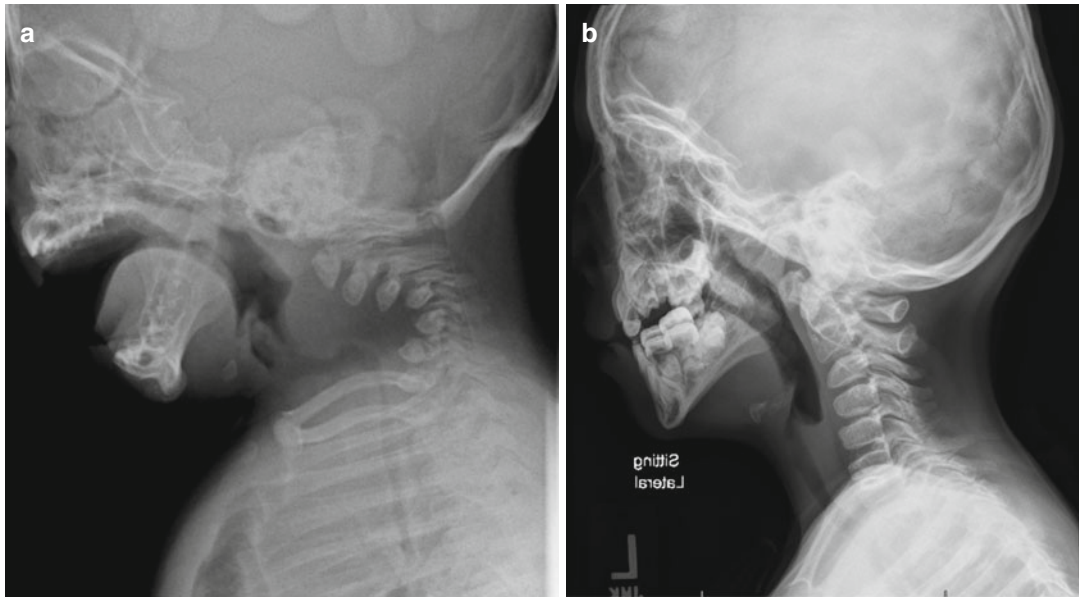

Fig. 20.6 Diastrophic dysplasia with cervical kyphosis measuring 60° at 3 months old (**a**). The kyphosis improved over time with residual kyphosis at the age of 8-year-old (**b**). The shape of vertebrae reconstituted

in situ posterior arthrodesis without instrumentation in milder deformity without cord compression. Halo immobilization is essential [30]. Restoration of lordosis is possible with anterior column growth. In patients with significant cervical kyphosis and cord compression anterior decompression and fusion followed by posterior fusion and halo immobilization has been described [31]. In young children, the posterior elements are not sufficient for rigid instrumentation.

Metatropic dysplasia may present with subaxial cervical kyphosis and stenosis. The kyphosis is associated with the distinctive severe platyspondyly. Other abnormalities including subaxial cervical spinal stenosis without kyphosis have been reported [19].

20.4 Cervicothoracic Abnormalities

Cervicothoracic stenosis has been reported in Morquio syndrome and chondrodysplasia punctata [30]. Stenosis in cervicothoracic area and in thoracic-lumbar junction has been reported in patients with Morquio syndrome in addition to upper cervical spinal abnormalities. The stenosis is usually associated with junctional kyphosis. It is assumed that relative ligament laxity globally affects the area of particular high tensile stress at the transitional vertebral segment where the facet orientation changes. The most common affected level is C7–T1 but it can extend to T4 [32]. A whole-spine MRI is necessary to screen for pathology. The age of onset can start at 2 years of age. Majority of the patients with Morquio syndrome are diagnosed from 2- to 5-year-old [33]. The most common presenting symptoms include an unsteady gait or failure to walk. Upper motor neuron signs are often present.

Cervicothoracic stenosis without associated kyphosis has been reported in chondrodysplasia punctata [34]. The focus of the stenosis arises from the dysplastic vertebrae.

20.5 Thoracic, Thoracolumbar, and Lumbar Abnormalities

20.5.1 Developmental Anatomy

The thoracic vertebrae are formed by a centrum and a neural arch. The neural arch is formed by two synchondroses. Closure of the centrum and

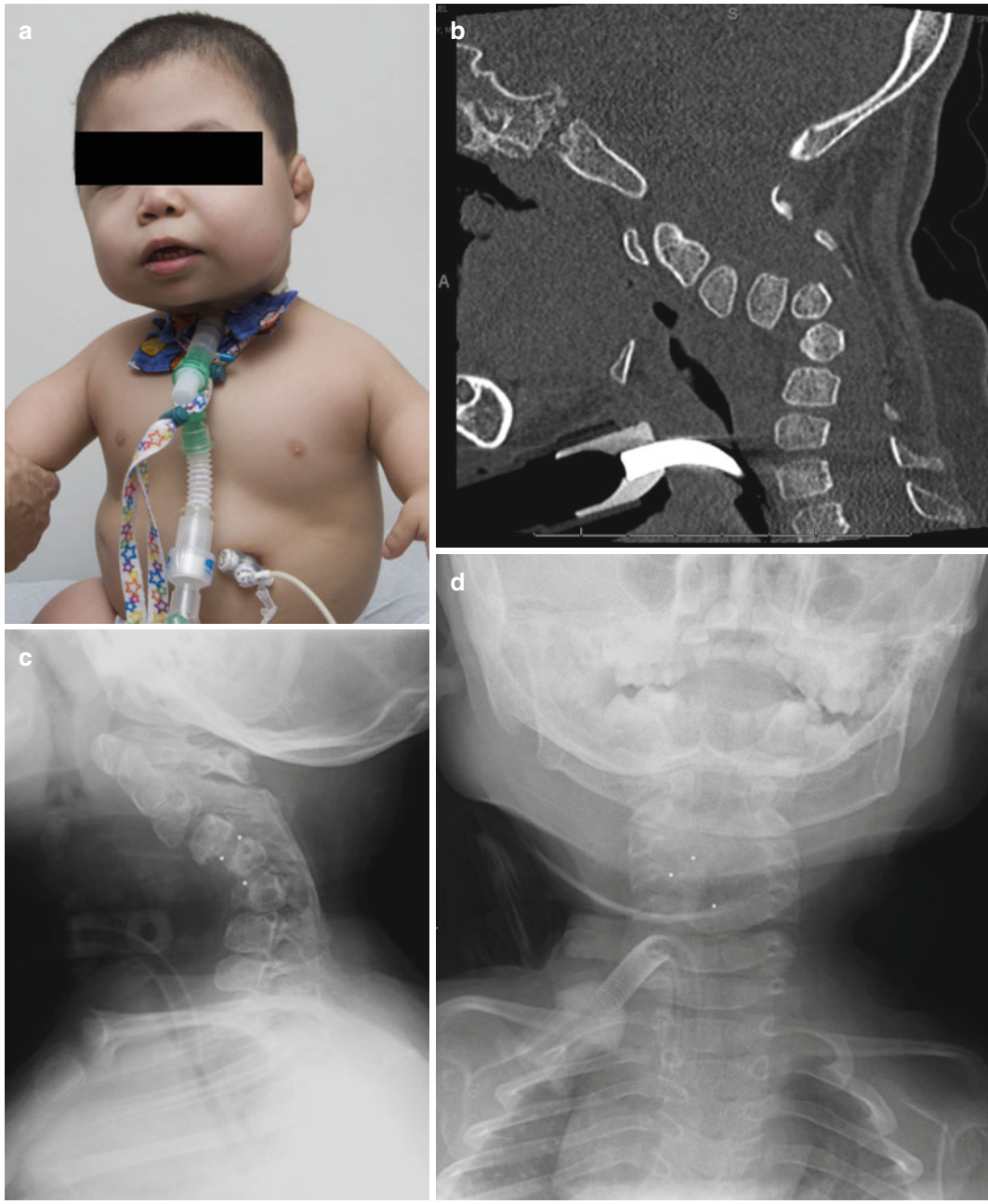

Fig. 20.7 Progressive cervical kyphosis in a patient with diastrophic dysplasia with tracheomalacia. The clinical photo showed a 3-year-old that needed tracheostomy for ventilation (**a**). Other features include cauliflower ear and hitchhiker thumb are evident. He presented with mid-cervical kyphosis of 90° with loss of "four corners" at C4 and C5 vertebral bodies (**b**) that was managed with anterior C4 vertebrectomy and fusion and posterior cervical arthrodesis. Postoperative radiographs in AP view (**c**) and lateral view (**d**)

the neural arch is achieved by 5–6 years. The posterior synchondroses of the neural arch united by 2–3 months of age. The spinal canal of thoracic vertebra attains adult dimension earlier than in cervical spine. A similar process of development is seen in the lumbar vertebrae.

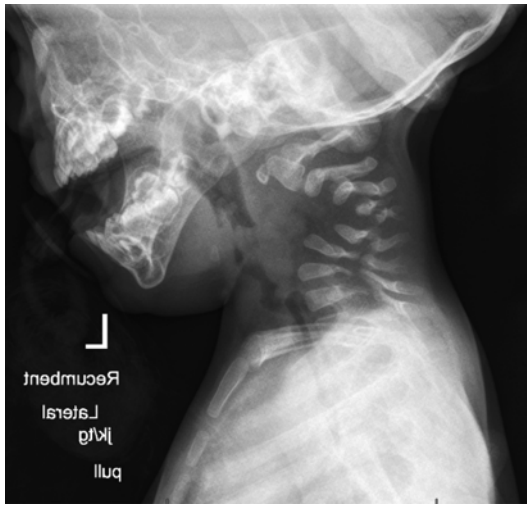

Fig. 20.8 Screening lateral cervical X-ray in Larsen showed mid-cervical kyphosis that has a progressive natural history

The level of the conus medullaris in skeletal dysplasia has been investigated and found to be stable at the L1 vertebra regardless of patient's age or the type of skeletal dysplasia involved in this cohort of 467 patients. Two cases of tethered cord in this series were noted in patients with chondrodysplasia punctata and diastrophic dysplasia [35].

20.5.2 Vertebral Abnormalities

Vertebral abnormalities are present in skeletal dysplasia. Platyspondyly is seen in metatropic dysplasia, pseudoachondroplasia, spondylodysplasias, Kniest dysplasia, and mucopolysaccharidoses. Different types of platyspondyly are evident in Kniest dysplasia (Fig. 20.9), likely related to abnormally soft cartilage, hence the name anisospondyly. In metatropic dysplasia, the decoupling of the perichondral growth and enchondral ossification results in what is termed the "overfaced pedicle" where the vertebral bodies are wider than the pedicles in posterior-anterior view of the radiograph [36]. In campomelic dysplasia, hypoplasia of the thoracic pedicles is present.

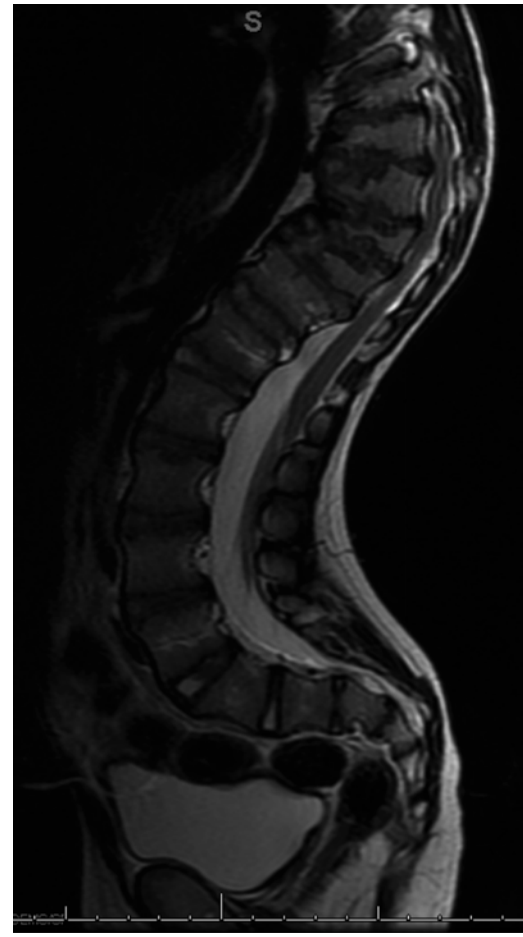

Fig. 20.9 MRI showed Kniest dysplasia with vertebral bodies flatten to various sizes, a condition known as anisospondyly

20.5.3 Thoracolumbar Kyphosis

In patients with achondroplasia, thoracolumbar kyphosis may develop in the first year. The incidence is reported to be as high as 94 %. The apical segment is typically at L2 with contribution from L1. Anterior wedging may occur. It is suggested that generalized hypotonia in the presence of relatively large head size with abdominal protuberance and ligamentous laxity contributes to the development of the kyphosis in a sitting position.

Spontaneous resolution occurs in majority of these patients when the child achieves independent ambulation [37]. The risk of persistent

thoracolumbar kyphosis includes thoracolumbar wedging of more than 60 % and developmental delay in motor function including inability to sit without support by 6 months and inability to walk independently by 18 months [38]. Early bracing may be required although its role in relation to the natural history is unclear [53].

Surgical management is indicated when the kyphosis has a large magnitude, fails to resolve or progresses, and is symptomatic (pain, symptoms of spinal stenosis) [54]. Classically, this is managed with anterior release and fusion with posterior decompression, instrumentation, and fusion. With the advent of modern instrumentation systems, posterior osteotomy or posterior vertebrectomy with anterior instrumentation can be done safely (Fig. 20.10) [39]. Careful attention to the sagittal alignment (adequate correction of the kyphosis and appropriate lumbar lordosis) is important to avoid misalignment. In achondroplasia, there is often compensatory thoracic lordosis above the kyphosis which may need to be corrected.

Other skeletal dysplasias with kyphotic deformities include chondrodysplasia punctata, diastrophic dysplasia, mucopolysaccharidoses, spondyloepiphyseal dysplasia, and metatropic dysplasia. Spontaneous resolution of the thoracolumbar kyphosis with bracing has been reported in a patient with spondylometaphyseal dysplasia, Kozlowski type [40]. Bracing is rarely indicated except in flexible deformities.

20.5.4 Thoracic, Thoracolumbar, and Lumbar Stenosis

In achondroplasia, the area of the spinal canal is reduced. This is more marked at the lumbar region and the narrowest level is reported at L4 [41]. The reduction in the canal volume is a result of abnormal enchondral ossification at the neurosynchondroses aggravated later in life by facet capsular thickening, facet hypertrophy, ligamentum flavum thickening, and intervertebral disk bulge. The interpedicular distance is reduced by 5 mm and the pedicle length is shorter by an average of 10 mm compared to controls. A trefoil-shaped spinal canal is evident with minimal space for the epidural content. Posterior vertebral body scalloping is typical. The characteristic progressive reduction of interpedicular distance from L1 to S1 is pathognomonic of achondroplasia.

The resultant spinal stenosis causes symptoms that can have a significant impact on quality of life. The age of onset of symptoms ranges from the first decade to adulthood. Twenty-five percent are symptomatic in the second decade and in 80 % over 60 years old. The most common symptom is neurogenic claudication [42]. Patients may present with lumbar pain, sensory disturbance, weakness, urgency, and bowel dysfunction. Cauda equina syndrome and myelopathy may occur especially in patients with associated thoracolumbar kyphosis [43].

Surgical management entails adequate posterior decompression, fusion, and instrumentation in the skeletally immature symptomatic patient to avoid post-laminectomy kyphosis (Fig. 20.11). Post-laminectomy kyphosis was seen in all 10 skeletally immature children decompressed without fusion in one study [44]. Skeletally mature patients without sagittal deformity can be managed by decompression without fusion as long as the facets are not damaged. Pedicle screw instrumentation is ideal [45]. Encroachment of the narrow canal with implants such as wires may result in neurological injury [46]. The standard pedicle screws starting points used in the average stature population are not ideal for patients with achondroplasia. The transverse axis of the pedicle screw deviates from the average stature population with less convergence in the lumbar region [47]. Fluoroscopic guidance or 3D navigation is recommended. Intraoperative complications such as dural tear is reported up to 30 % secondary to a thin and stretched out dura [55].

20.5.5 Thoracic, Thoracolumbar, or Lumbar Scoliosis

Scoliotic deformities are common and typically present as kyphoscoliosis [54] (Figs. 20.12 and

20.13). *Patients with diastrophic dysplasia, chondrodysplasia punctata, spondyloepiphyseal dysplasia congenita, acromesomelic dysplasia, pseudoachondroplasia, cartilage hair hypoplasia, Larsen syndrome, and Kniest dysplasia have a variable course*.

In metatropic dysplasia, the progressive spinal deformity changes the rhizomelic children with a

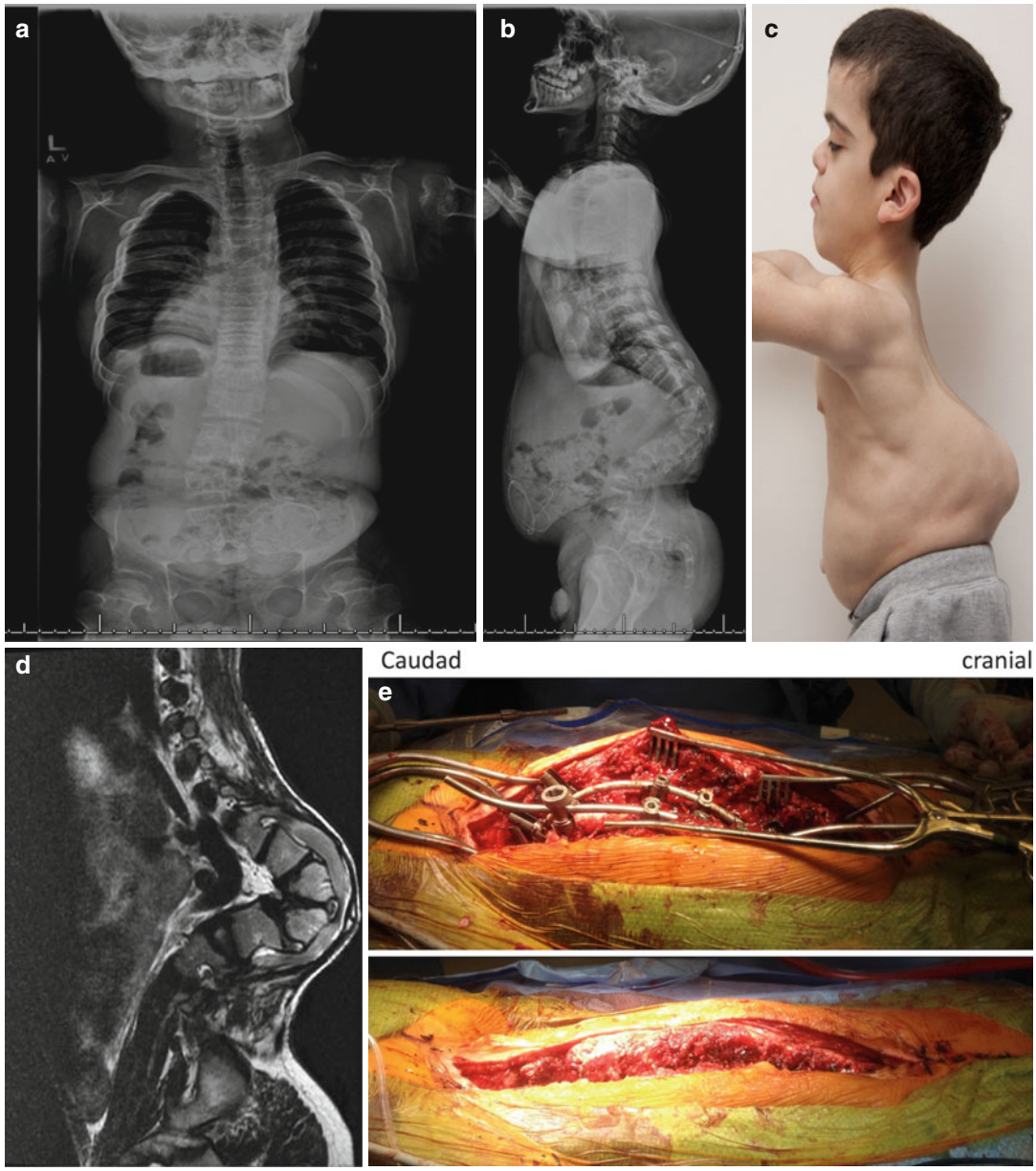

Fig. 20.10 Significant thoracolumbar kyphosis after previous attempts at management in a 15-year-old with achondroplasia and symptoms of neurogenic claudication as shown in AP (**a**) and lateral (**b**) X-rays. Clinical photo of thoracolumbar kyphosis (**c**). Other features include bifrontoparietal bossing and mid-face hypoplasia. He underwent posterior laminectomy and posterior L1 vertebral column resection and reconstruction using an expandable cage. Reduction was achieved by cantilever reduction technique. (**d**) MRI showed thoracolumbar kyphosis and spinal stenosis. (**e**) Left rod temporary rod in situ. Right rod cantilevered to reduction screw. (**f**) Alignment achieved post reduction

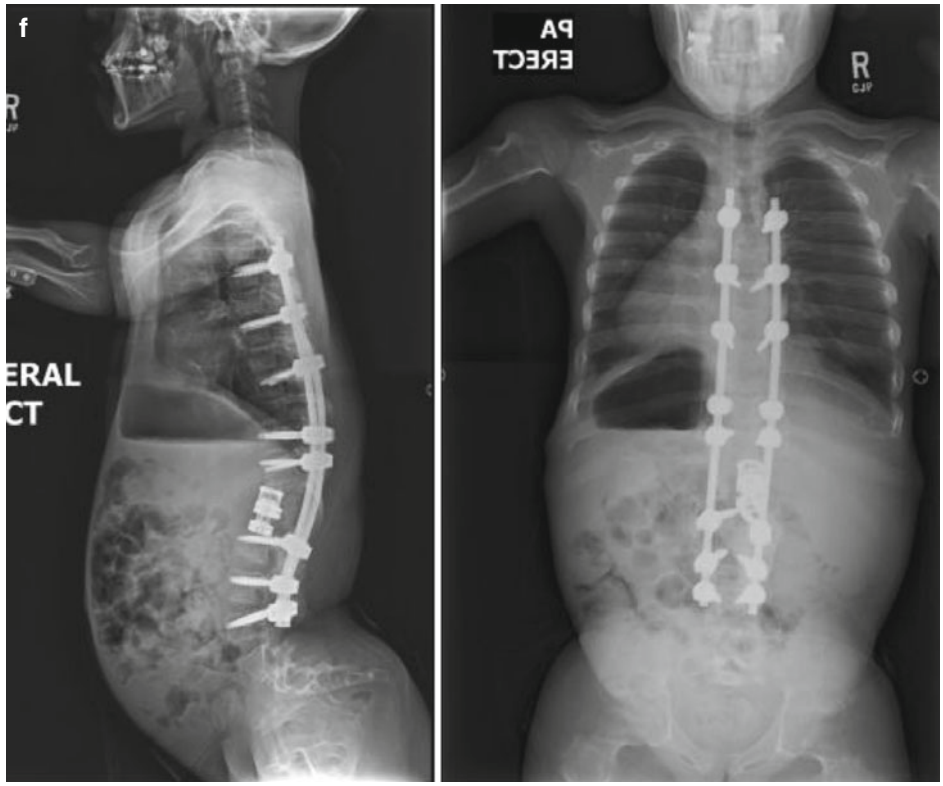

Fig. 20.10 (continued)

normal trunk alignment and length at birth to a severe kyphoscoliosis [49, 52]. The age of onset can be in the first year but there is a wide clinical spectrum. The children have a long and narrow thoracic cage with severe restrictive lung disease. Also the deformity is stiff and bracing is not a good option. Surgical intervention is challenging given the early onset of the spinal deformity. Spinal stenosis is common and with the severe platyspondyly instrumentation can be difficult. Fusionless techniques should be used to preserve trunk growth whenever possible.

In chondrodysplasia punctata, two curve patterns are recognized [16]. The nondysplastic has minimal kyphosis with mild vertebral body abnormalities. The curve responds to standard posterior spinal fusion technique. The dysplastic curve is less predictable. It has an early onset during infancy, is progressive, and is associated with severe vertebral body deformity that results in apparent hemivertebra.

In diastrophic dysplasia, three curve patterns are present [48]. An early progressive curve pattern that resembles the progressive form of infantile idiopathic scoliosis is seen. It is evident by the age of 3 years. Without intervention, significant kyphoscoliosis of more than 100° is expected. The idiopathic form presents from age 3 to 10 and the final curve rarely exceeds 100°. The curve pattern that presents in older children can develop in response to pelvic obliquity, vertebral deformity, and asymmetrical intervertebral disk collapse. It is managed using standard techniques.

Early onset and progressive curves warrant early intervention. Use of bracing is limited to lower magnitude and less progressive curve patterns in children without cardiopulmonary compromise. Early thoracic spinal fusion may result in or exacerbate thoracic insufficiency syndrome. Surgical intervention should involve growth-friendly spinal instrumentation such as dual

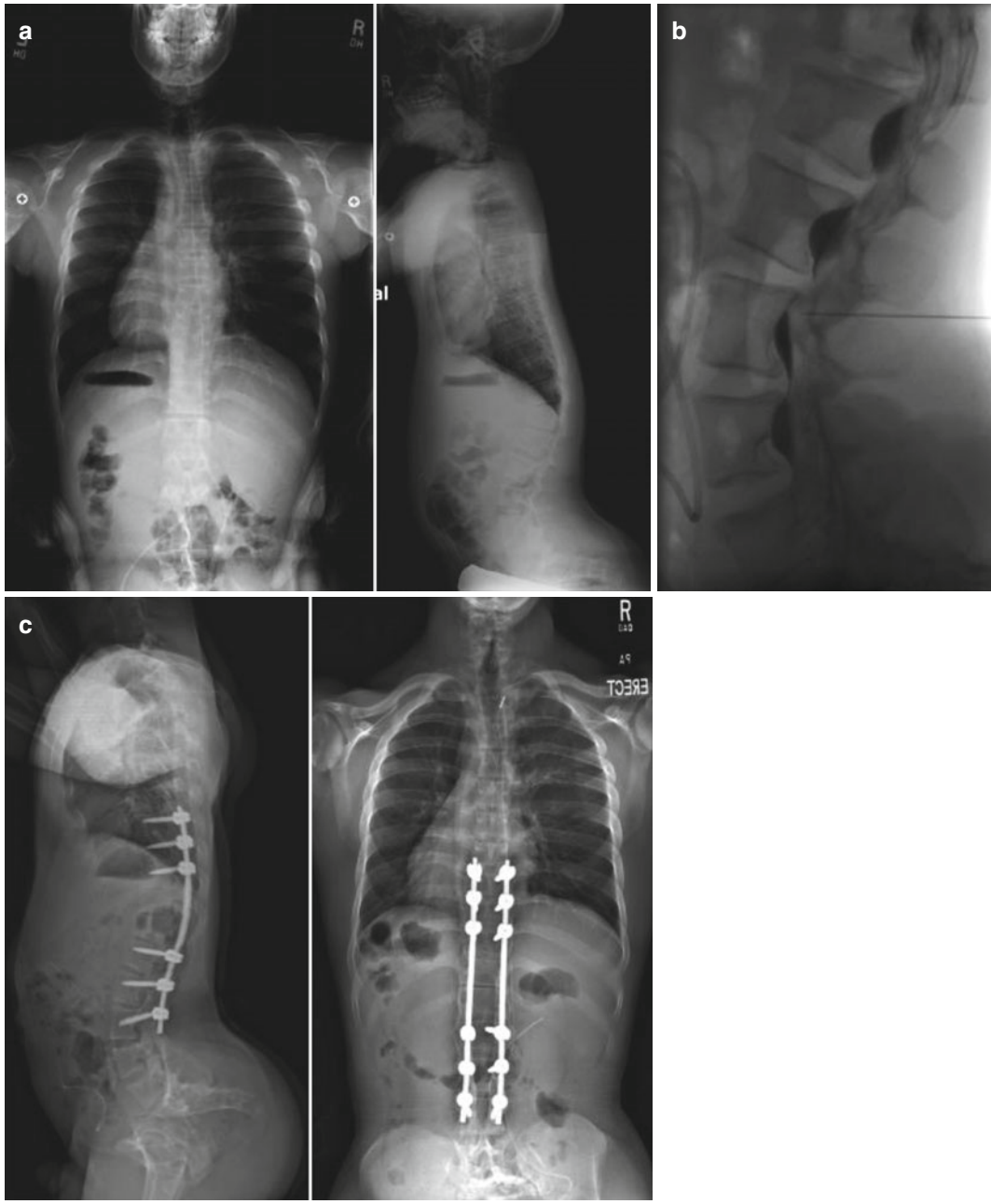

Fig. 20.11 Achondroplasia in a 14-year-old with symptoms of spinal stenosis. Note the thoracolumbar kyphosis and thoracic lordosis in the AP and lateral views (**a**). Intraoperative myelogram (**b**) demonstrates the spinal stenosis with scalloping at the posterior vertebral bodies. The patient had posterior osteotomies, decompression, instrumentation and fusion as shown in (**c**)

growing rods or hybrid dual growing rods in skeletally immature children. The growth-friendly instrumentation is effective in controlling curve progression while allowing truncal growth (Fig. 20.13). Complications are comparable to diagnoses in patients without skeletal dysplasia. Complications such as wound infection from repeated lengthening or implant-related

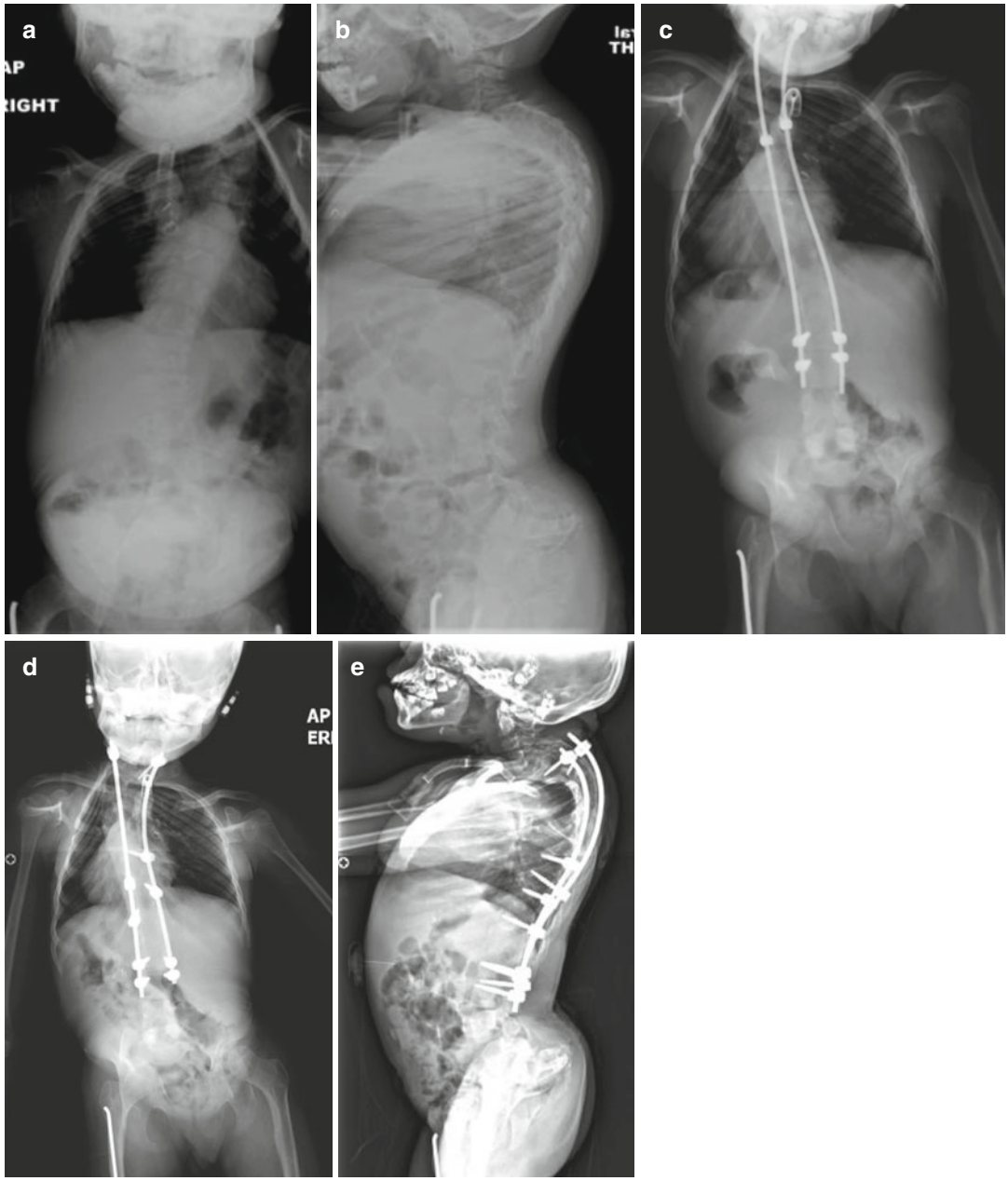

Fig. 20.12 Scoliosis in campomelic dysplasia. (**a**) Preoperative AP and (**b**) preoperative lateral. The hypoplastic pedicles made instrumentation difficult. Hooks were used with a customized growing rods construct (lengthened distally) (**c**). Pedicle screw construct was used at the final fusion stage with the aid of navigation. (**d**) Lateral postoperative definitive fusion

issues (rod breakage, hook or screw dislodgements) and alignment abnormalities (proximal junctional kyphosis) are reported at 40 % [49]. The role of the magnetic growing rod is unclear but an attractive procedure to avoid repeated surgical procedures. Standard fusion techniques are used in skeletally mature (or close to maturity) children.

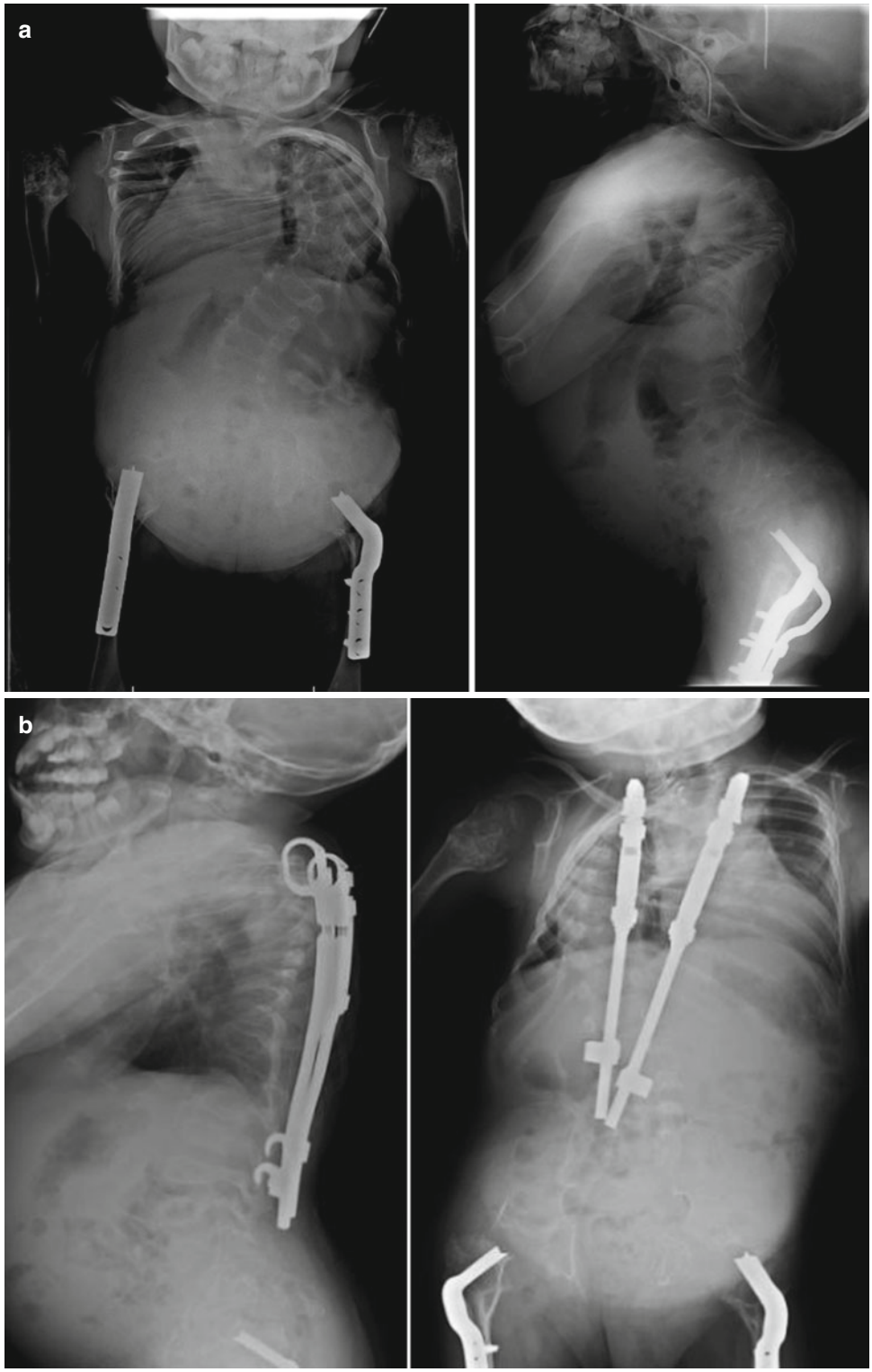

Fig. 20.13 Kyphoscoliosis in a patient with spondyloepimetaphyseal dysplasia. AP standing lateral views before (**a**) and after (**b**) VEPTRs were implanted at the age of 8. Note the proximal junctional kyphosis. Final fusion was done at the age of 13 as shown in radiographs AP standing (**c**) and lateral (**d**)

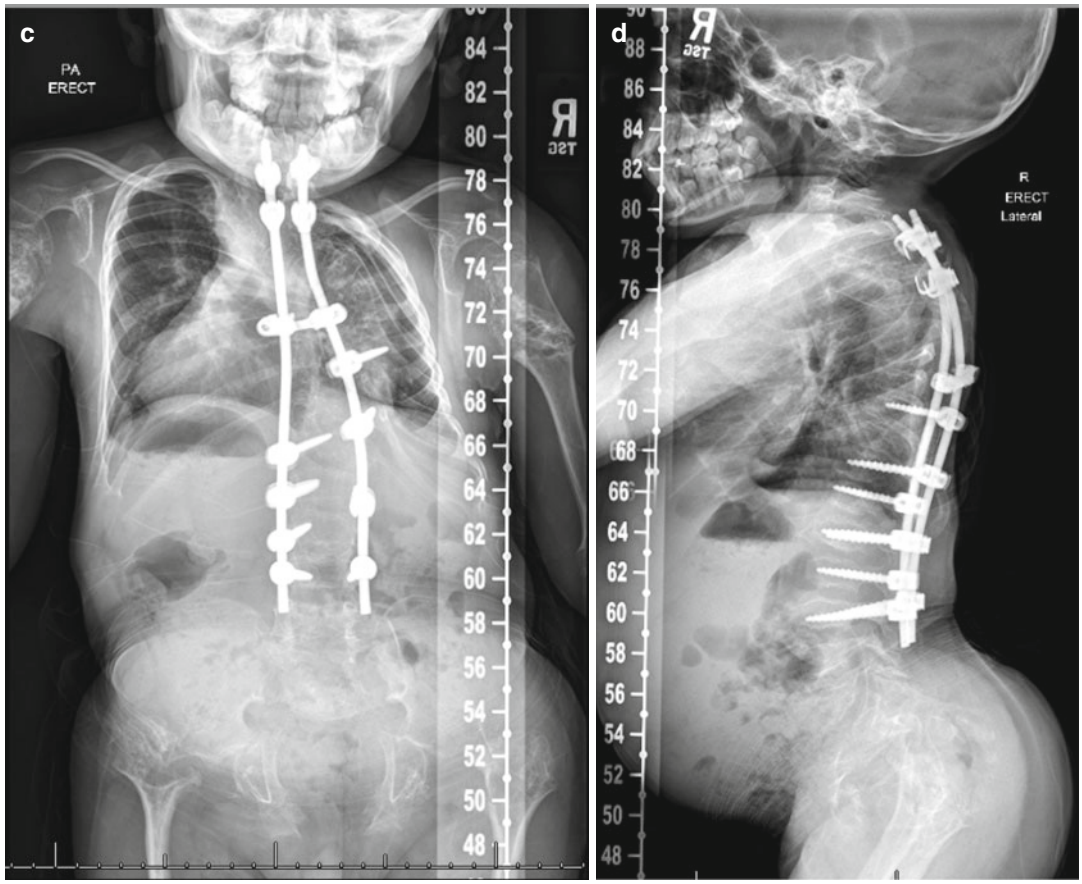

Fig. 20.13 (continued)

20.6 Lumbosacral Abnormalities

Hyperlordosis is frequently seen in many skeletal dysplasias. It may be primary lordosis or secondary to hip flexion contractures, particularly in children with coxa vara (Fig. 20.14).

In achondroplasia, the spinopelvic relationship is not well understood. Pelvic incidence in this cohort averaged 43° compared to average stature controls of 52°. The excessive lordosis compensated the thoracolumbar kyphosis and generated a negative 22 mm sagittal balance which has a poor correlation with the pain score [50]. A component of the lumbar lordosis is probably generated by the fixed flexion contracture of hips. Some authors believed that the abnormal hyperlordosis causes pain and observed that achondroplasts often squat to reduce lordosis and achieve symptomatic relief (also probably using this maneuver to increase the canal volume) [51].

20.7 Clinical Presentation

Patients with skeletal dysplasia present with spinal problems at different stages of life. Antenatal deformity screening with ultrasound at second trimester may identify deformity and structural defects. Anxious parents may seek an orthopedic consult to discuss future medical problems and expected quality of life.

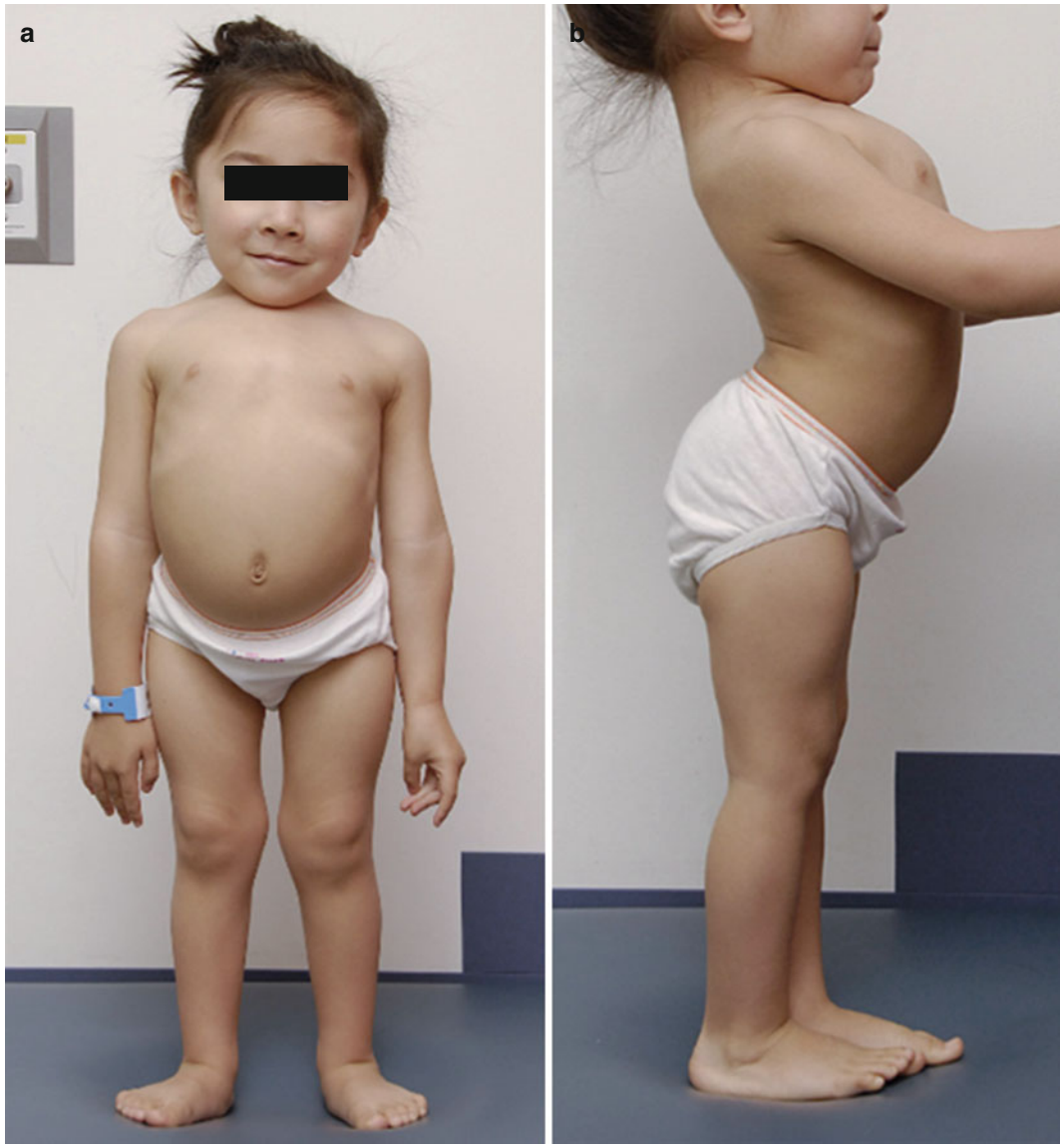

Fig. 20.14 Clinical photos showing hyperlordosis secondary to coxa vara in a girl with spondyloepiphyseal dysplasia congenita. Significant spinal shortening is evident (**a**) with lumbar hyperlordosis with abdomen protuberance seen (**b**)

At birth, characteristic facies and disproportionate body appearance allow many types of skeletal dysplasia to be recognized (see Table 20.2). Forty to fifty percent of patients however do not live beyond the first year of life secondary to severe skeletal involvement of the rib-cage complex and its associated thoracic insufficiency syndrome. Significant abnormalities such as multiple major joint dislocation in Larsen syndrome or cleft palate present in Type 2 collagenopathy and diastrophic dysplasia allow diagnosis to be made early with skeletal survey.

Skeletal dysplasia with mild shortening and deformity and normal facies may not be recognized in the first year of life. This includes pseudoachondroplasia, multiple epiphyseal dysplasia,

and metaphyseal dysplasia. Storage disorders such as mucopolysaccharidoses result in damage to target organs over time and are clinically evident at 2–3 years of age. The child may first present secondary to other associated disorders especially respiratory symptoms. Chest radiography may reveal the underlying radiographic skeletal abnormalities. More commonly, developmental delay, short stature, or abnormal gait may be the first clinical evidence of underlying abnormalities.

Radiographic evaluation is the most powerful single tool for the diagnosis of the skeletal dysplasia. A complete skeletal survey should be done in children at first evaluation. In neonates or infants less than 6 months of age, an anteroposterior (AP) and lateral babygram that shows the whole spine, hands, and lower extremities should be obtained. Lateral cervical spine radiographs that include the skull in flexion and extension are essential.

20.8 Management

Patients with skeletal dysplasias need to be managed by a coordinated multidisciplinary team involving the orthopedic surgeon, genetics, neurosurgeon, ENT surgeon, cardiology, pulmonology dentist, rehabilitative physician, nutritionist, and geneticist. Retinal detachment, for example, needs to be screened for in patients with SEDc and Kniest dysplasia. Cardiopulmonary issues have direct impact on patient care and need to be addressed. Genetic counseling is important for parents in regard to future pregnancy.

The majority of spinal problems can be addressed nonoperatively. Surgical intervention when needed should be planned meticulously. Anesthesia consult is paramount prior to intervention. Patients with skeletal dysplasia can have difficult airways. Tracheomalacia and cervical instability entail use of special intubation techniques. Joint contracture and disproportionate body frame make positioning difficult. Trial positioning prior to the operative day with egg crate foam or gel pad is useful. Intraoperative neuromonitoring is the standard of care. Prepositioning and postpositioning baseline signals are acquired. Different stimulation regimens may be required in myelopathic patients. Blood conserving strategy by using cell-saver avoids allogenic blood transfusion. Postoperative management may be required in an intensive care setting.

Conclusion

Skeletal dysplasia has varied spinal manifestations that involve the entire spinal column. It is important to understand the developmental anatomy in the context of individual skeletal dysplasias for effective management. Plain radiography is the gold standard for diagnosis. Advanced imaging will be required in the upper cervical spine. Perioperative issues should be addressed to ensure a successful outcome. A coordinated multidisciplinary team is essential for clinical effectiveness.

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Pediatric Spine Trauma

21

Emmanouil Grigoriou and John P. Dormans

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Key Points

- A majority of pediatric spine injuries occur at the cervical spine, with most fractures occurring at the upper cervical spine.
- Ligamentous laxity, horizontal orientation of vertebral facets, wedge-shaped vertebral body, and underdeveloped paraspinal muscles in the immature spine result in a relatively high incidence of cervical spine injuries.
- The clinician should be cognizant of normal anatomic variants of the immature spine when interpreting pediatric spine radiographs.
- Unfused physeal lines, pseudosubluxation, and absence of cervical lordosis may be normal findings in the developing spine.
- Odontoid fracture, a common injury pattern in the immature cervical spine, typically occurs at the synchondrosis and can be managed conservatively with immobilization.
- Atlantoaxial rotatory subluxation is managed conservatively if diagnosed early. Chronic cases may require manual reduction under anesthesia, or fusion.
- SCIWORA is a relatively common injury in the pediatric population.

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- Thoracolumbar injuries are usually the result of MVCs or sports-related injuries and are more prevalent in the adolescent population.
- Thoracolumbar spine injuries are the result of significant trauma and the presence of extraspinal injuries are common.
- In a flexion-distraction injury, the anterior column fails in compression, while the middle and posterior columns fail in tension.
- Stable injuries of the thoracolumbar spine can be treated conservatively with brief bed rest, spasmolytics, thoracolumbosacral orthosis; mechanically unstable fractures with neurological deficits should be managed surgically.
- Close follow-up is necessary for early detection and management of posttraumatic scoliosis in the patient with thoracolumbar trauma.

21.1 Introduction

Incidence of pediatric spinal fractures is relatively low and ranges from 1 to 10 % in all traumas. They often result from motor vehicular accidents, falls, athletics, and occasionally child abuse. Unique features of the pediatric spine results in specific injury patterns. The ligaments, disks, and soft tissue of the pediatric spine are laxer in comparison with the adult spine. This laxity, however, also accounts for the increased incidence of spinal cord injuries without radiographic abnormalities (SCIWORA) [1–3].

Of note is the different pattern of injuries in cervical trauma between young children and adolescent patients [4, 5]. Increased elasticity, a large head-to-torso ratio in the child of 8 years of age and less, results in preponderance of upper cervical spine pathology in this cohort. In contrast, the adolescent patient, with more mature structures, presents with injury patterns similar to that of the adult. Nitecki and Moir [5] showed an 87 % incidence of upper cervical spine injuries in patients younger than 8 years of age in a series of 227 consecutive C-spine fractures in children. Cervical fractures commonly seen in the pediatric population include atlanto-occipital (AO) dissociation, upper cervical spine injuries, SCIWORA, and the thoracolumbar compression fractures.

Pediatric thoracolumbar trauma has also unique features differentiating it for the adult spine trauma; these injuries are more likely in children older than 8 years and are usually associated with high-impact sports or MVCs [6]. Thoracolumbar injury patterns vary and can include minor compression fractures, apophyseal fractures, flexion-distraction injuries, burst fractures, and major combined fracture dislocations.

21.2 Cervical Spine Trauma

21.2.1 Epidemiology

The overall incidence of cervical trauma in the pediatric trauma patient has been estimated to range from 1 to 2 % [7]. However, a majority of pediatric spine injuries (60–80 %) occurs at the C-spine [8]. A difference in the pattern of cervical spine injury has been established in patients below and above 8 years of age. In children presenting with spinal injuries, cervical injuries appear to be more common in younger children compared with adolescents. In addition, specifically upper cervical injuries appear to be more prevalent in the younger population. Osenbach and Menezes [9] reported a 79 % incidence of cervical spine trauma in children 8 years of age or less presenting with spinal trauma. In contrast, the incidence of cervical trauma in those 8 years of age or older was 54 %. They also demonstrated higher rate of upper cervical spine injury in children 8 years of age or less. Subluxations and neurological injuries were more common in the younger population. Knox et al. showed increased variability of injury patterns even in very young children; infants and toddlers (age 0–3) were more likely to sustain ligamentous injuries, whereas young children (age 4–9) were more

likely to sustain compression fractures [4]. SCIWORA injuries were also more common in the younger population. Hadley et al. [10] corroborated the above findings in their study of 122 cases of vertebral column injuries in adolescent patients. Brown et al. [11] reported a 68 % prevalence of upper cervical spine injuries in children. Cervical spine injury, especially upper cervical spine, SCIWORA, and neurological injuries were more common in patients <9 years of age. SCIWORA is also a common finding in the abused child presenting with C-spine trauma. Other patterns of injury reported in the abused child include epidural and subdural hematoma of the spinal cord at the cervicomedulary junction and ventral spinal contusion in the upper C-spine [9–13].

Motor vehicle crashes (MVC), athletics, and falls are the most common causes of injuries in the pediatric population [11, 14]. Brown et al. [11] attribute 52 % of cervical trauma to MVC, 27 % to sports-related injuries, and 15 % to falls. A recent multi-institutional study of 540 children with cervical spine injuries showed that, although for children <7 years old MVCs were the most common injury mechanism, for children 8–15 years old sports accounted for as many injuries as MVCs (23 %, 23 %). Mortality after pediatric spine trauma has been estimated at 7–28 % in various studies [11, 13, 14].

21.2.2 Cervical Spine Anatomy

The atlas is formed by three ossification centers, located at the anterior arch and each neural arch. Ossification of the anterior arch is often absent at birth and may not appear before the first year. Union of the anterior ossification center and the lateral centers occurs by 7 years of age. The posterior arch results from posterior fusion of the neural arch and occurs by 3 years of age.

The axis is formed by four ossification centers, located within the central mass, the neural arches, and the dens. These centers are present at birth. Two ossification centers, fused in utero at 7 months, form the odontoid process and are separated from the body by a cartilaginous physis. Often the two odontoid ossification centers may persist after birth. The dentocentral basilar synchondroses, located below the atlantoaxial facet joint, fuse between 3 and 6 years of age. Knowledge of the location of the odontocentral physeal line, which may be present radiographically until 11 years of age, prevents erroneous diagnoses of a transverse dens fracture. The ossiculum terminali, a secondary ossification center at the apex of the dens, appears between 3 and 6 years of age. The neural arches fuse with the body between 3 and 6 years of age, and fuse posteriorly to form the posterior arch by age 2–3 years (Fig. 21.1). C3–C7 have similar patterns of development with three ossification centers: one located within the vertebral bodies and two within the neural arches. Union of the neurocentral synchondrosis occurs between 2 and 6 and the posterior arches fuse between 2 and 4 [1, 16].

Distinctive anatomic features in the immature C-spine account for the pattern of injuries seen in this population. The horizontal orientation of the vertebral facet joints in the pediatric cervical spine and the relative ligamentous laxity results in a relatively high incidence of upper cervical subluxation and cervical injuries in this population. Before 8 years of age, the articular facets have an orientation of ~30° and progressively become more vertical. By adolescence, the orientation is similar to that of an adult at 55–70° [1]. Furthermore, the relatively high head-to-body ratio of the pediatric patient localizes the fulcrum of spinal flexion at C2–C3 as opposed to C5 and C6 in the adult [17]. Other anatomic features contributing to the pattern on injuries seen in this population include delayed ossification of the uncinate process (a stabilizer in the adult spine), anterior wedge of the immature vertebral body, and underdevelopment of cervical para-spinal muscles. These factors contribute to the relatively high incidence of upper cervical instability/injuries in the pediatric patient [1, 18, 19].

21.2.3 Clinical Evaluation

Cervical spine injury in the pediatric patient often results from considerable trauma such as motor

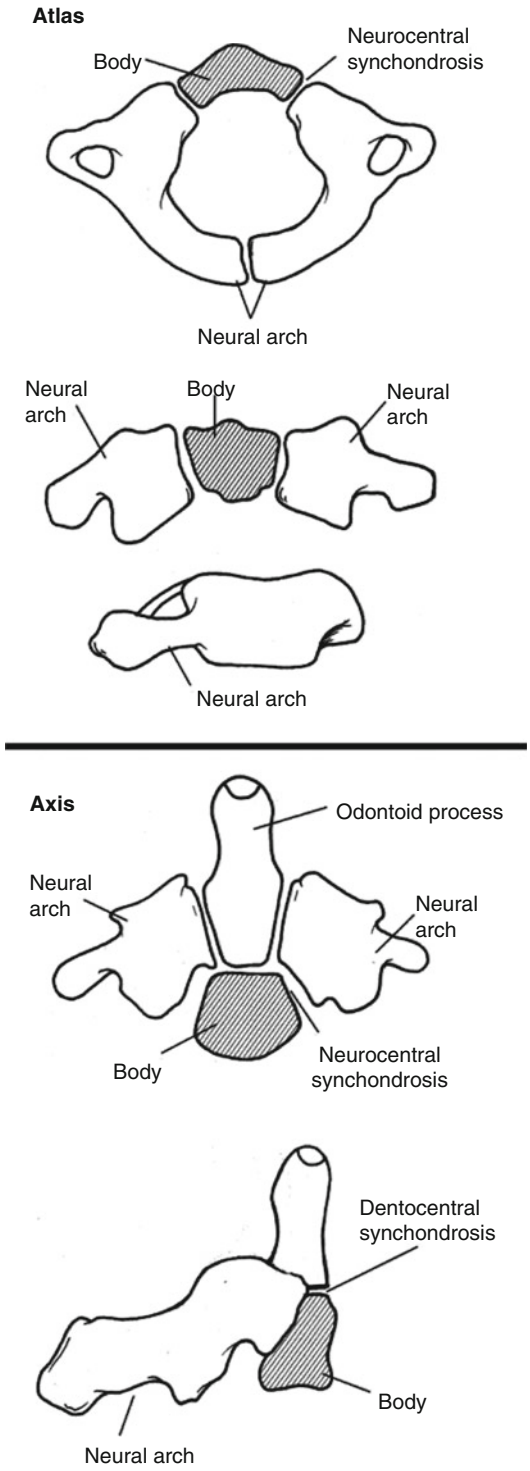

Fig. 21.1 Ossification centers of the atlas and axis (Reprinted from Copley and Dormans [15]. With permission from Wolters Kluwer Health)

vehicular accidents, fall from height, and penetrating injuries. Athletic injuries and child abuse are also potential causes of trauma in the pediatric spine. Patients may be unconscious at presentation. When conscious, they report neck pain or spasm. Signs of trauma to the head and face, bruising caused by seat or shoulder belts may also be present. Often these findings are absent in the young child, and instead, the child may report occipital headaches. Additionally, increased suspicion is crucial in children with syndromes that have a genetic predisposition for cervical spine injuries like Down syndrome, Klippel-Feil syndrome, and Morquio syndrome.

In addition to strict adherence to ATLS protocol, a comprehensive neurological examination should be performed on all patients with suspected spinal injuries, the patient should also evaluated for other associated injuries [21].

The child presenting with a suspected cervical spine injury should be immobilized in a spine board. The indications proposed by Lee at al. [22] for immobilization of the pediatric cervical spine are presented in Table 21.1.

Because of the relatively large head of children <8 years of age, there is a tendency for flexion of the head, when these patients are immobilized in

Table 21.1 Indications for immobilization of the pediatric cervical spine

Abnormal neurological examination findings (complete testing of motor, sensory, and reflex functions of all extremities is required)
Unreliable examination due to inconsolable or unconscious children, or substance abuse
Neck pain or focal neck tenderness
Mechanism of injury potentially associated with CSI (high-speed motor vehicle collisions, falls greater than body height, bicycle or diving accidents, forced hyperextension injuries, acceleration–deceleration injuries involving the head)
History of transient neurological symptoms suggestive of SCIWORA (weakness, paresthesias, or lightening/burning sensation down the spine/extremity or related to neck movement)
Physical signs of neck trauma (ecchymosis, abrasion, deformity, swelling, or tenderness) or significant trauma to the head or face

Based on data from Ref. [22]

a traditional spine board. This can be prevented by elevating the torso relative to the head with a thin mattress or using a board with a recess [23]. A halo ring and vest is also a viable method of immobilizing the pediatric cervical spine. The construct requires the use of 8–12 pins applied with low insertional torques (1–5 in. lbs). A CT scan of the head obtained prior to application of the halo can help identify cranial sutures and thin areas of the skull. These areas should be avoided when inserting halo pins. Advantages of a halo vest immobilization include relative ease of application, earlier mobilization of the patient, access to wounds of the neck and scalp, and freedom of mandibular motion. Pin site infection is the most common complication associated with halo vest immobilization [24, 25].

21.2.4 Radiological Evaluation

The standard trauma series, including an AP, lateral, and odontoid views of the cervical spine, including the T1 level, can be obtained when a C-spine injury is suspected. In the lucid and older patient without neurological deficits, flexion and extension views of the C-spine should also be obtained if instability is suspected. Indices of instability include segmental kyphosis, anterior soft tissue swelling, and equivocal subluxation in the standard radiographic views [1, 26]. In small infants, odontoid views and lateral views may be difficult to interpret because of the overlapping skull. In these instances, a lateral view of the skull may be utilized to evaluate the upper cervical spine [1].

Radiographic parameters used for the evaluation of upper cervical stability include the atlanto-dens interval (ADI), Powers ratio, and the basion-axis distance. The ADI describes the distance between the anterior surface of the dens and the posterior surface of the anterior arc of the atlas and should be <5 mm. The Powers ratio defines the ratio between the distance of the basion and the posterior arc of the atlas, and the distance between the opisthion and the anterior arc of the atlas. This ratio averages 0.77 and a value exceeding 1 or <0.55 is indicative of atlanto-occipital dislocation. The basion-axis distance is the distance between the basion and a vertical line extended cephalad from the posterior border of the dens. This line should be 12 mm or less in children 13 years of age or less (Fig. 21.2) [19, 21].

Normal anatomical variants in the developing spine should be recognized and differentiated from pathological findings. The most common anatomical variation in the pediatric spine is the physiological displacement between C2 and C3, and less often C3 and C4; pseudosubluxation of C2 on C3 was observed in 22 % of pediatric polytrauma patients by Shaw et al. [28] and in 46 % on lateral dynamic views in patients <8 years of age by Cattell and Filtzer [29]. In order to differentiate this pseudosubluxation from an injury, one must evaluate the spinolaminar line (i.e., Swischuck's line) (Fig. 21.3). The spinolaminar line is drawn along the posterior arch of C1 to C3 and normally passes within 1 mm of the anterior cortex of the posterior arch of C2. That said, a displacement of >1.5 mm should raise concerns and a displacement of >4 mm is always an abnormal finding [29].

Absence of cervical lordosis, a pathological finding in the adult patient, may be normal variant in some pediatric patients younger than 16. In these patients, an intraspinous distance of 1.5 times the distance of the adjacent intraspinous distances or less confirms stability [29, 30]; additionally, normal lordosis is restored with the neck in extension.

In the subject of normal CT measurements and parameters specifically of the pediatric cervical spine, recent literature suggests the need to define age- and sex-specific values. Vachhrajani et al [20] found that there are age-independent (LMI = lateral mass interval and ADI = atlanto-dental interval) but also age-dependent (BDI = basion-dental interval; CCI = craniocervical interval, and PADI = posterior ADI) normal CT measurements of the upper cervical spine in children [21]. Further research is needed to validate these preliminary values in a larger and geographically diverse patient cohort so as to better direct patient care.

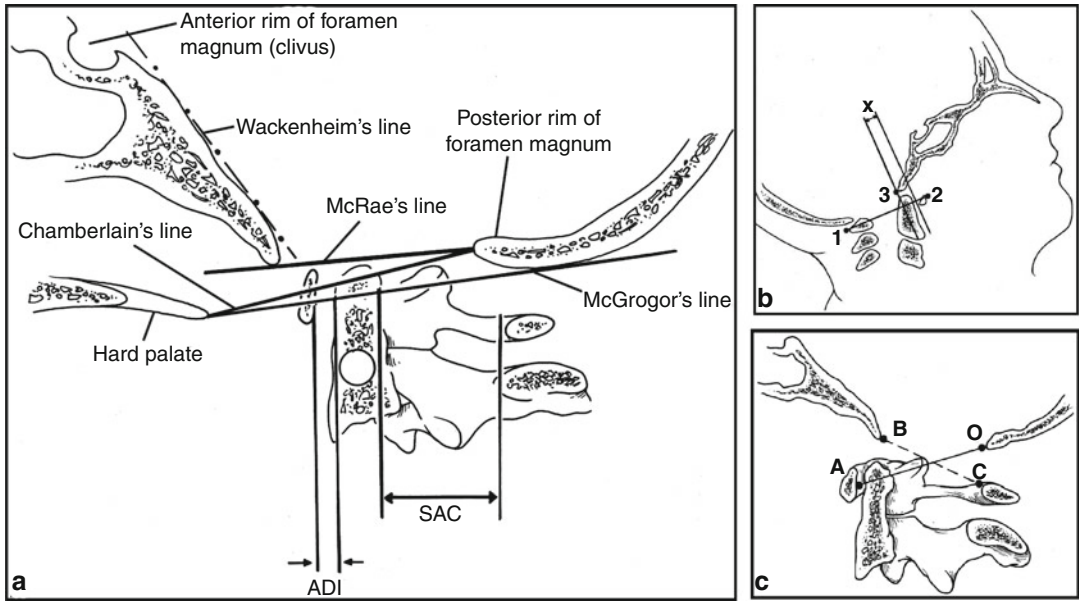

Fig. 21.2 Cervical spine landmarks and measurement parameters. (**a**) The lines commonly used to determine basilar impression and the measurements for determining atlantoaxial instability. *ADI* atlanto-odontoid interval, *SAC* space available for cord. (**b**) Method of measuring atlanto-occipital instability according to Wiesel and Rothman [27]. The atlantal line joins points *1* and *2*. A line perpendicular to the atlantal line is made at the posterior margin of the anterior arch of the atlas. The distance (*x*) from the basion (*3*) to the perpendicular line should not vary by 1 mm or more in flexion and extension. (**c**) The ratio of powers is determined by drawing a line from the basion (*B*) to the posterior arch of the atlas (*C*) and a second line from the opisthion (*O*) to the anterior arch of the atlas (*A*). The length of line *BC* is divided by the length of line *OA*. A ratio of 1.0 or greater is diagnostic of anterior occipitoatlantal dislocation (Reprinted from Copley and Dormans [15]. With permission from Wolters Kluwer Health)

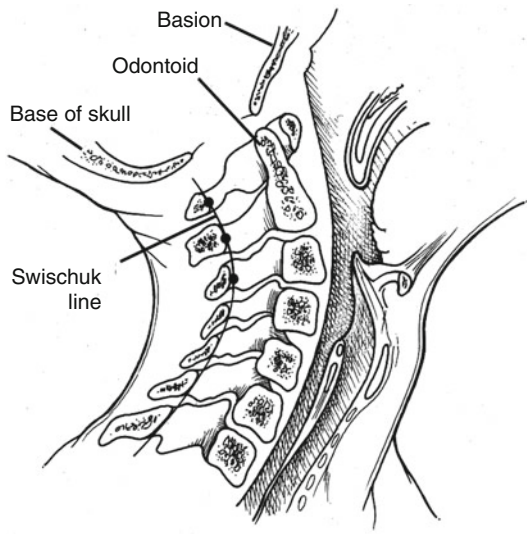

Fig. 21.3 Spinolaminar line (Swischuck line) used to differentiate pseudosubluxation of C2 on C3 from true cervical injury (Reprinted from Copley and Dormans [15]. With permission from Wolters Kluwer Health)

Differentiation of true fractures from unfused physeal line is essential in the evaluation of the pediatric spine. Secondary ossification centers in the spinous processes and unfused ring apophyses of vertebral bodies may mimic fractures on radiographs. Normal physeal plates are smooth, regular areas of radiolucency with underlying subchondral sclerotic lines and occurring at predictable locations. In contrast, acute fractures are irregular, without sclerosis, and occur at any location within the C-spine [21, 30].

Finally, although advanced imaging (CT and MRI) can be used to identify subtle injuries, undetected from the physical examination and the plain radiographs, CT should not be used to clear the cervical spine of a pediatric trauma patient with a potentially existing instability secondary to soft tissue injury [31]. Additionally, since flexion and extension radiographs are inappropriate to assess instability in intubated,

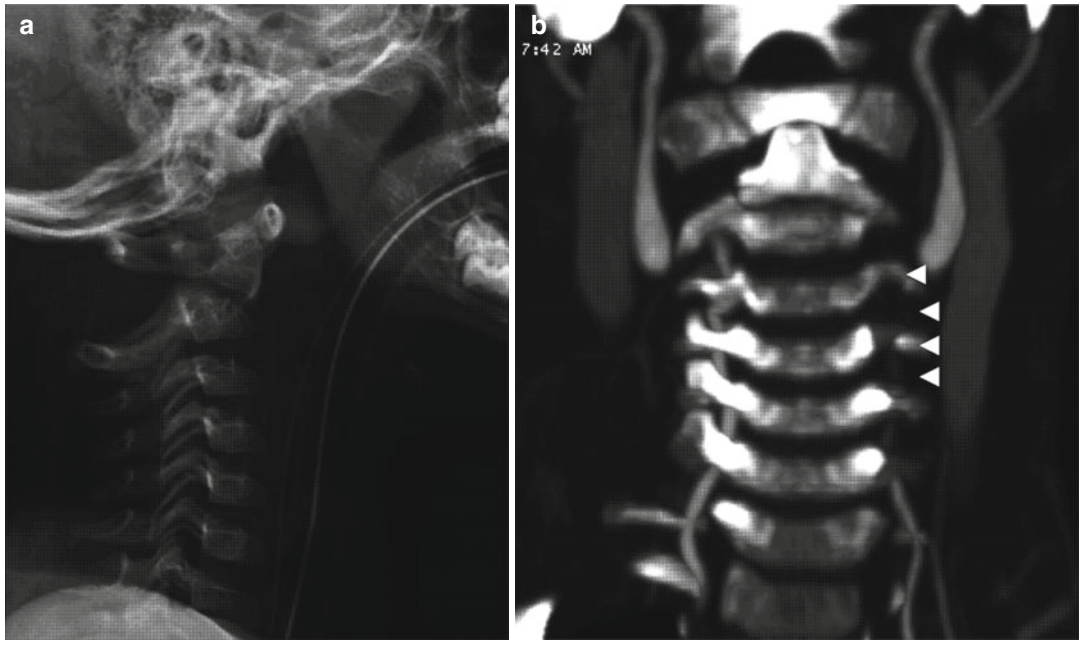

Fig. 21.4 Lateral radiograph of the cervical spine (**a**) of a 4-year-old male after being struck by a car demonstrating an odontoid fracture, with posterior dislocation of the C2 vertebral body with respect to the dens and increased widening of the posterior elements of C1 and C2. CT angiogram of the neck (**b**) demonstrating a dens fracture and accompanied compromise of the left vertebral artery with long-segment luminal irregularity (*arrows*)

obtunded, or uncooperative children, MRI can be very helpful in identifying soft tissue injury. Flynn et al. [32] reported that MRI was able to demonstrate cervical injuries not seen in radiographs in 15 of 64 children (24 %). Finally, CT angiograms can be very helpful in delineating vascular compromise (Fig. 21.4).

21.2.5 Specific Injury Patterns

21.2.5.1 Atlanto-occipital Dislocations

A historically fatal injury, current management protocols have decreased the mortality associated with these injuries. These protocols include improved resuscitation at the scene of injury, early immobilization, high index of suspicion, and improved diagnostic modalities. These injuries often result from a rapid deceleration during an MVC or MVC-pedestrian collision. The head is hyperflexed relative to the torso resulting in dislocation of the atlanto-occipital joint. The lack of inherent bony stability of the atlanto-occipital joint, and ligamentous laxity of the pediatric cervical spine, predisposes the young child to this injury. Three types of atlanto-occipital dislocation (AOD) have been described. Type 1 involves anterior displacement of the occiput relative to the atlas; type 2 describes longitudinal displacement of the occiput from the atlas; and type 3 describes a posterior displacement [19].

The paired semilunar occipital condyles located at the inferior surface of the cranium articulate with the concave lateral masses of C1. At birth, the surfaces of the lateral masses are relatively flat and progressively become more concave with age. This absence of bony articular congruence contributes to the increased risk of atlanto-occipital instability in the young pediatric patient, especially under the age of 6 years [33].

Numerous ligaments provide atlanto-occipital stability. The tectorial membrane, a cranial extension of the posterior longitudinal ligament, attaches C1 to the anterior surface of the foramen magnum. Deep to the tectorial ligament are the paired alar and apical ligaments. These ligaments

originate from the dens, with the former attaching to the medial aspects of the occipital condyles and the latter attaching to the foramen magnum. The tectorial, alar, and apical ligaments constitute the major stabilizers of the atlanto-occipital joint. The tectorial membrane limits extension, while odontoid impaction on the basion limits flexion. The alar ligaments limit lateral bending. Both the alar ligaments and tectorial membranes limit distraction [33].

The patient with AOD is often polytraumatized with numerous associated injuries including head injuries. Hemodynamic instability, often secondary to neurogenic shock, is a common finding. On presentation, the patients may be intubated and on pressors. The earlier clinical state often conceals the presence of an AOD and prevents a comprehensive neurological examination. High clinical suspicion is therefore needed to diagnose AODs and optimize management. Patients often present with quadriplegia or quadraparesis. In addition, cranial nerve palsies, especially the 6th, 9th–12th, are often present. The lower CN palsies may be secondary to stretch injury, while the sixth palsy may result from head injury. The vertebral arteries may also be injured [34–36].

Plain radiographs are the mainstay in the evaluation of patients with AOD. Power ratio, the Wackenheim line, and the occipitocondylar distance are radiographic parameters used in the evaluation of atlanto-occipital instability. On the lateral view, the Wackenheim line is drawn along the clivus and tangentially intersects the tip of the odontoid. Anterior or posterior displacement of this line relative to the odontoid signifies a corresponding displacement of the occiput on the axial spine. The occipital condylar distance defines the distance between the occipital condylar facet and the C1 facet. A distance greater than 5 mm signifies disruption of the atlanto-occipital joint (see Fig. 21.2) [19, 21, 30]. Finally, the power ratio is defined as the ration of the distance from the basion to the midcervical portion of the posterior laminar line of the atlas to the distance from the opisthion to the midvertical portion of the posterior surface of the anterior ring of the atlas. A ratio >1.0 is suggestive of anterior subluxation of the occiput on the atlas. CT scan is a sensitive method of evaluating atlanto-occipital instability. Fine-cut CT with coronal and sagittal reconstruction clearly demonstrates malalignment of the atlanto-occipital joints. MRI has also been utilized to evaluate atlanto-occipital instability. It provides excellent visualizations of the ligaments and soft tissue structures above the atlanto-occipital joint. Visualization of a complete defect of the tectorial membrane on MRI is diagnostic of AOD. Furthermore, injuries to the spinal cord can be visualized on MRI [33].

As with all polytraumatized patients, ventilation should be optimized and hemodynamic stability maintained. The C-spine should be immobilized in a neutral position; halo immobilization with or without traction or Minerva casting are the two prevalent options. Use of cervical traction remains controversial with some advocating its utility in type l and type 3 injuries. That said, cervical traction has no role in the management of type 2 injuries because alignment is adequate [33, 35–37].

Definitive management of AOD is controversial with many authors advocating early posterior fusion since many of these injuries ultimately become unstable due to the associated soft tissue injury. Techniques described in the literature include contoured loop fixation, atlantoaxial transarticular screw placement, and wire fixation with bone grafting. Two techniques using wire fixation and autologous iliac crest graft or rib grafts have been described. Both the procedures require the use of cables or wires affixed to two burr holes drilled on the occiput. In the former, a single-shaped iliac graft is attached to a trough prepared at the base of the occiput and to the spinous process of the axis. In the latter procedure, paired autologous rib grafts with a natural curve resembling the spinal anatomy are harvested, and fixed to the occiput and axis with sublaminar wires (Fig. 21.5a–d) [16, 34, 35]. Factors favoring a decision to proceed with immediate posterior fusion include 3 years of age or later, and a complete AOD with neurological deficits. Some have, however, recommended initial nonoperative management with an orthosis as it is believed that the pediatric spine has a higher

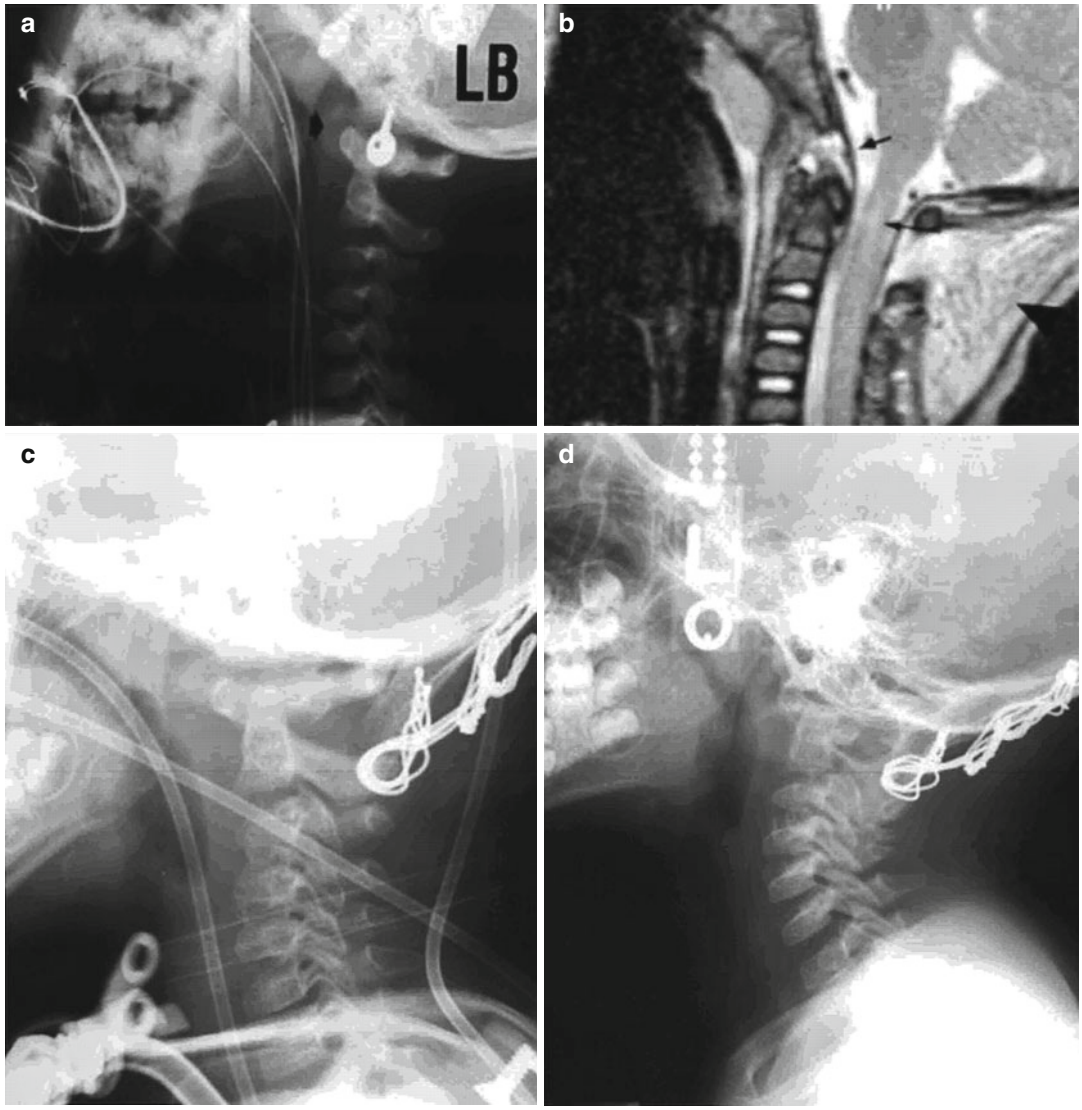

Fig. 21.5 A 1-year-old male pedestrian stuck presenting with the AOD. (**a**) Lateral C-spine radiographs showing an AOD and atlantoaxial disruption. (**b**) T2 MRI demonstrating spinal cord contusion. (**c**, **d**) Postoperative (12 and 55 weeks after posterior fusion) radiographs showing normal alignment and graft consolidation (Reprinted from Hosalkar et al. [35]. With permission from *Journal of Bone and Joint Surgery*)

repair capacity. Healing can thus be obtained without instrumentation. Because of the technical difficulties involved in instrumentation of the very immature spine, and the high healing capacity of very young children, nonoperative management is often a viable option in this population. Nonoperative management may also be indicated in patients with incomplete injuries [16, 33–35, 38].

21.2.5.2 Fractures of the Atlas

Fractures of the atlas, classically the Jefferson fracture, are uncommon injuries in the pediatric population. It is usually secondary to an axial compression load resulting from a fall onto the top of the head or hyperextension after a motor vehicular crash. Force is transmitted from the occiput to the lateral masses, resulting in a fracture at the weakest point of the atlas. This is

usually the anterior or posterior arch. Diastasis of the lateral masses can also result in avulsion of the transverse ligament, leading to atlantoaxial instability. Current studies show a 6.9-mm displacement of the lateral masses and suggest a transverse ligament rupture, which result in instability [15, 39, 40].

Pediatric patients with a fracture at the atlas usually present with neck pain, cervical muscle spasm, torticollis, and reduction in range of motion. Neurological deficits are uncommon with C1 fractures. Though the earlier-mentioned signs and symptoms are nonspecific, they should raise the index of suspicion and prompt assessment for the presence of this fracture pattern especially since they are commonly missed due to inadequate imaging of the occipitocervical junction [41].

Isolated fractures of C1 is often absent on plain radiographs. Findings suggestive of these fractures include a prevertebral hematoma on the lateral cervical spine views, or separation of the lateral masses on an odontoid view. A CT scan of the C-spine and MRI is needed for accurate diagnosis of these fractures (Fig. 21.6a, b) [39, 40, 42].

There are few reports of isolated pediatric C1 ring fractures in the literature and conservative management of these fractures with external immobilization seems to be the consensus recommended treatment. Successful use of soft cervical color, halo vest, and rigid cervical collar for varied duration has been documented. Recovery of full function and return to activities are expected with conservative management [41–45].

21.2.5.3 Odontoid Fractures

Odontoid fractures are common in the pediatric population. Like most cervical spine injuries in this population, the cause of injury often involves an MVC via rapid deceleration with flexion, or a fall from height. Odontoid fractures usually occur at the base of the dens at the synchondrosis. The weak synchondrosis and the relatively large head size of the young pediatric patient predispose this location to injury.

The clinical presentation of patients with odontoid fractures can range from neck pain to significant spinal cord injuries (SCI). Fasset et al. [46] demonstrated 33 % incidence of neurological deficits in patients presenting with odontoid fractures. When present, spinal cord injuries (SCI) tend to occur at the cervicothoracic junction. It has been proposed that this may be secondary to a traction injury of the cord resulting from hyperflexion [47]. In the Fasset et al. [46] series, 53 % of SCIs occurred at the cervicothoracic level.

Radiographic evaluation of odontoid fractures begins with a plain radiograph of the C-spine. On

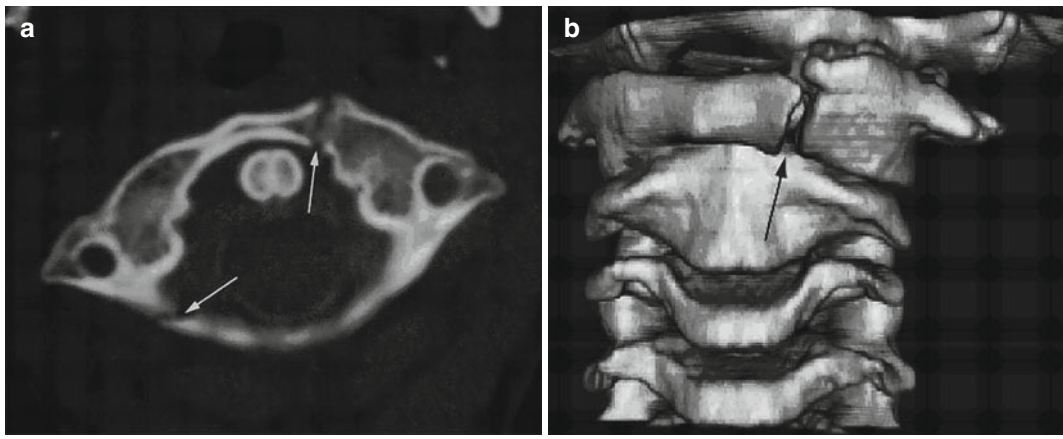

Fig. 21.6 Axial CT scan through C1 (**a**) and three-dimensional reformatted CT scan of the upper cervical spine (**b**) obtained in a 6-year-old boy showing anterior and posterior ring fractures of C1 (*arrows*) (Reprinted from Lustrin et al. [21]. With permission from *Radiological Society of North America*)

the lateral view, anterior angulation or displacement of the dens is often apparent. A CT scan with a sagittal and 3D reconstruction may be needed to further delineate the fracture site and demonstrate diastasis of the synchondroses. In equivocal cases, MRI imaging can be useful. Soft tissue changes at the C1 and C2 levels combined with a high signal at the synchondroses in the appropriate clinical setting suggest a fracture [21, 46, 48].

Odontoid fractures can be managed nonoperatively. Closed reduction can be obtained with extension or hyperextension of the C-spine (Fig. 21.7). Fifty percent apposition of the fracture segment is usually sufficient for healing. The C-spine is subsequently immobilized in a halo vest or pinless halo for 2–3 months. Frequent radiographic follow-up is required to insure stability is maintained. Fasset et al. [46] showed a fusion rate of 93 % with conservative management.

Operative management is indicated in fractures where reduction is not obtained with external immobilization, or with nonunion (no evidence of healing after 3–6 months). Viable surgical options include posterior C1 and C2 fusion with bone grafting and wiring. Motion-sparing procedures have not been proven to be efficacious in the pediatric population [46]

21.2.5.4 Hangman's Fracture

Traumatic anterior spondylolisthesis of the axis due to bilateral fracture of the pars interarticularis is extremely rare in the pediatric population with only a few cases reported. It occurs most commonly in children 2 years of age and or less. Various factors account for this pattern of injury in the developing spine. As indicated earlier, the relatively large head-to-body ratio of the young child localizes fulcrum of flexion over the upper cervical spine. This is further compounded by the relative ligamentous laxity and weak neck muscles of the young child. That said, injury results from cervical hyperextension [19, 50].

Radiological diagnosis of pediatric Hangman's fracture is complicated by unique features of the

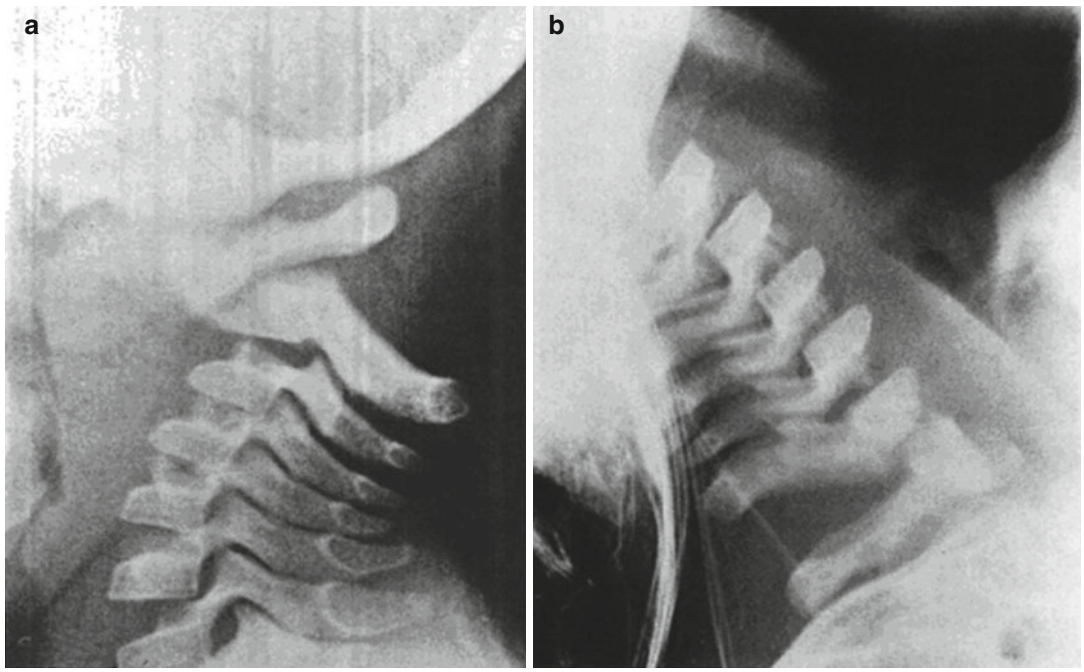

Fig. 21.7 Lateral C-spine radiographs of a young child with a severely displaced odontoid fracture after a fall. The second image shows a reduced fracture with hyperextension (Reprinted from Sherk et al. [49]. With permission from *American Journal of Bone and Joint Surgery*)

pediatric spine, as discussed earlier. Hangman's fractures involve fractures of the pars interarticularis and anterolisthesis of C2 on C3. Evaluation of the pediatric Hangman's fracture begins with standard plain radiographic views, with CT and MRI reserved for more detailed evaluation. On radiographs, a radiolucent line can be observed anterior to the pedicles of the axis. Pathological anterior displacement of C2 is likely if the tip of the spinous process of C2 is greater than 2 mm anterior to a line (Swischuk's line) connecting the spinous processes of C1–C3 (see Fig. 21.2) [16, 19].

The Levine classification system for traumatic spondylolisthesis can be extrapolated to the pediatric population. Type 1 injury describes 3 mm or less translation between C2 and C3 without angulation. In type 2 injuries, there is greater than 3 mm of translation between C2 and C3 and greater than 10° angulation. Type 3 fractures include the characteristics of type 2 fractures as well as bilateral facet dislocation and have greater angular components [51].

Management of nondisplaced Hangman's fracture is usually conservative with immobilization in a halo ring and vest or pinless halo. Levine type 3 fractures are managed surgically with open reduction and posterior fusion [19].

21.2.5.5 Atlanto-axial Rotatory Subluxation

Injuries of the atlantoaxial joint include ligamentous disruptions resulting in instability and odontoid fractures. The atlantoaxial joint is a relatively mobile joint with 50 % of cervical rotation occurring at this joint. Furthermore, its diameter, one-third occupied by the dens, one-third by the spinal cord, and one-third by subarachnoid space, allows for significant cervical motion before cord injury [21, 52].

Atlantoaxial rotatory subluxation (AARS) is a rotational deformity of C1 on C2 most commonly secondary to infection or trauma leading to lateral neck flexion and contralateral rotation. Upper respiratory infections, retro-pharyngeal abscesses are common causes of atlantoaxial instability. However, it also results from trauma in ~20–45 % of the patients [1, 19].

Children with atlantoaxial subluxation present with torticollis, with the chin rotated to one side and head laterally deviated to the contralateral side. In addition, there is spasm of the sternocleidomastoid muscle opposite to the side of chin rotation. They will typically report headaches and neck pain. In a fixed subluxation, rotation of the atlantoaxial joint is prevented or limited by impingement of the C1 facet joint on C2. An attempt at manual reduction of the deformity is usually painful. Neurological deficits are rare [1, 36, 52].

Radiographic assessment of patients presenting with suspected atlantoaxial dislocation includes standard cervical spine series complemented with a dynamic rotation CT scan. MRI is useful to rule out other diagnosis and assess further the surrounding soft tissues. Findings indicative of subluxation on lateral radiographs include absence of a defined craniocervical junction and lack of orientation of the anterior arch in a true lateral plane. On the AP plane, the anteriorly subluxated lateral mass may appear wider and more proximal to the midline, while the opposite mass appears farther and smaller. Significant displacement is however unlikely in the presence of a normal ADI. Plain radiography is often difficult to interpret because of the tilted position of the head (Fig. 21.8a–d) [19, 21].

Fielding and Hawkins [53] classified atlantoaxial rotatory subluxation (AARS) into four categories. Type 1, the most common, shows no displacement of C1 and a normal ADI. It results from unilateral facet subluxation without disruption of the transverse ligament. Type 2 demonstrates a greater than 3–5 mm anterior displacement with compromise of the transverse ligament. Type 3 describes bilateral anterior facet dislocation associated with complete rupture of the transverse ligament and leads to significantly reduced space for the spinal cord. It is manifested by a >5-mm ADI. Type 4 demonstrates posterior displacement of C1 and is rare.

Atlantoaxial rotatory fixation (AARF) can be demonstrated with a dynamic rotation CT. A fixated C1 and C2 unit rotates as one on a CT obtained at rest, and with attempted neck rotation. It results from an untreated AARS [21].

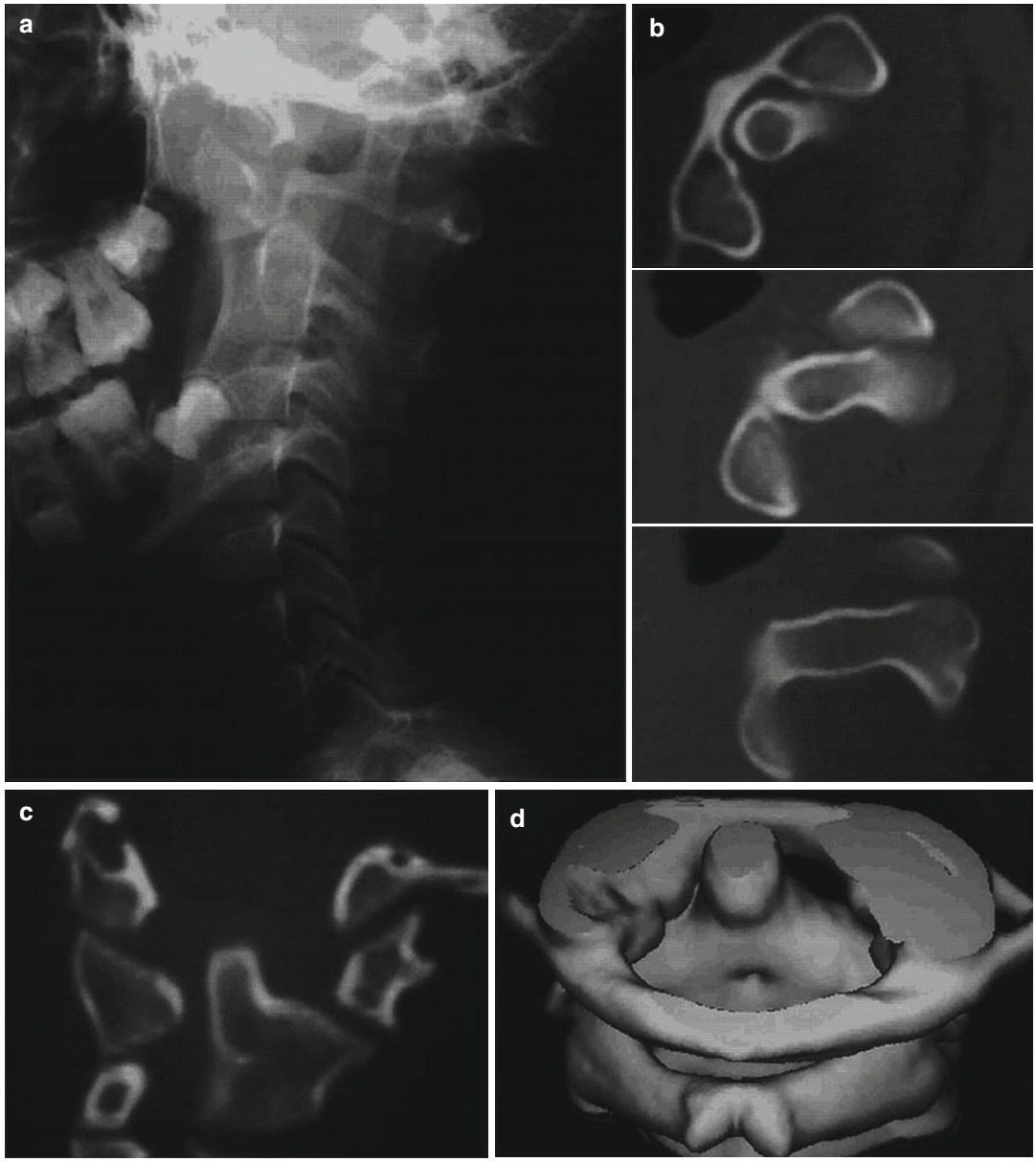

Fig. 21.8 Lateral radiograph of the cervical spine (**a**), axial CT scans (**b**), coronal CT scan (**c**), and 3D reformatted CT scan (**d**) through C1 and C2 in a 12-year-old girl with atlantoaxial rotatory subluxation (Reprinted from Lustrin et al. [21]. With permission from *Radiological Society of North America*)

Management of atlantoaxial rotatory subluxation may involve conservative management or in rare instances surgical stabilization. Often, mild subluxations may reduce spontaneously and patients do not present for formal intervention. Patients presenting with symptoms of <1 week duration can be treated with a soft cervical collar, NSAIDs, and muscle relaxants. If symptoms have persisted for >1 week, patients may be treated with head halter traction, supplemented with muscle relaxants and analgesics. Conservative therapy is unlikely to be successful in patients presenting with symptoms persisting for >1 month. In this setting, reduction can be

attempted with a halo traction or manual manipulation under anesthesia. If successful, the patient can be immobilized in a halo vest for 6 weeks. If this modality is unsuccessful, surgery may be indicated. Surgery is also indicated in the presence of persistent instability, neurological deficits and may include bilateral facet reduction with fusion. The atlantoaxial joint may also be fused in a subluxated position [19, 54].

21.2.5.6 Spinal Cord Injury Without Radiographic Abnormalities (SCIWORA)

Spinal cord injury without radiological abnormalities is a relatively prevalent injury in the pediatric population. It was initially defined by Pang and Wilberger [55] as the presence of myelopathy as a result of trauma without evidence of fracture or ligamentous instability on plain radiographs or tomography. Its incidence has been estimated from 4 % to as high as 67 % of all pediatric spinal traumas [11, 55, 56]. Anatomical and biomechanical features already described earlier predispose the developing spine to SCIWORA. The osseous and ligamentous structures of the spinal column are laxer than the spinal cord. Consequently, these structures can undergo significantly more deformation, than the spinal cord, without failure. Biomechanical studies have shown that the bony and soft tissue structures can stretch for ~2 in. before failure. The cord, however, can be ruptured after 0.25 in. of displacement. SCIWORA results from transient traction or compression of the spinal cord resulting from cervical hyperextension or hyperflexion. Hyperextension of the spine results in compression of the cord by the ligamentum flavum, while flexion results in a traction injury to the spinal cord [19, 21, 55, 56]. Younger children with high-energy injuries typically present with more significant cord injuries as compared to the adolescent patient with injuries secondary to athletic activities [11, 19].

MRI is the study of choice for the patient presenting with spinal cord injury without evidence of trauma on plain radiograph and CT. Grabb and Pang have described five different spinal cord injury patterns on MRI: complete spinal cord disruption, major cord hemorrhage, minor cord hemorrhage, only edema, and no abnormality. Approximately 30 % of patients with SCIWORA have no obvious spinal cord abnormality on MRI [32, 57].

In the absence of osseous abnormalities, or mechanical instability, management of SCIWORA involves prolonged rigid external immobilization for 2–3 months. If instability is noted, surgical stabilization may be necessary. Long-term prognosis following SCIWORA is dependent on the neurological status at presentation [55]. Especially for patients with minor hemorrhage or edema only on MRI, Grabb and Pang [57] found that MRI was a better predictor of outcome than the neurological status at the time of presentation.

21.2.6 Cervical Spine Injuries Outcomes

Overall, the reported mortality rate in pediatric patients with cervical spine trauma ranges from 15 to 20 % [5, 9, 11]. The reported incidence of concomitant neurological injuries in children with cervical spine injuries is 35–66 % [8, 11, 58]. In their series of 103 consecutive C-spine injuries, Brown et al. [11] reported a mortality of 18.5 %. In this series, 18 % of patients required operative intervention with the most common indication being instability. Closed head injuries were often associated with cervical spine injury and were a significant adverse prognostic factor. In the same series, Brown et al. [11] reported a mortality rate of 49 % in the presence of closed head injuries. Other identified risk factors for increased mortality in pediatric patients with cervical spine trauma are upper cervical injuries and atlanto-occipital (AO) dislocation [5, 9, 11]. Platzer et al. [58] reported a 66 % incidence of neurological deficits in a study of 56 pediatric patients with cervical spine injury over a 25-year period. Complete recovery of neurological function occurred in 68 % of these patients. A 75 % mortality rate was reported in patients with complete spinal injuries.

In a large multicenter review of cervical spine injuries, $n = 1.098$, Patel et al. [13] noted a 35 %

incidence of spinal cord injury. Of these, 50 % were SCIWORAs, and 76 % of these patients had incomplete injuries with 24 % suffering complete neurological injuries. Complete neurological injury was a significant predictor of mortality; 53 % of patients with complete neurological injuries died. In contrast, a 15 % mortality rate was noted in patients without neurological deficits, and 16 % for those patients with incomplete deficits. Overall mortality in these series was 17 % [13].

21.3 Thoracolumbar Fractures

21.3.1 Epidemiology

Thoracolumbar injuries in the pediatric population occur primarily in children between the ages of 14 and 16 and account for 1–2 % of all pediatric fractures [7, 8, 59]. The most common location is T4–T12 and then T12–L2. In a retrospective review of 610 cases of pediatric spine injuries in adolescents, the majority (63 %) of injuries occurred in males [60]. Compression fractures are the most commonly reported, followed by process-only fractures and then unstable fractures [61, 62]. Most of the injuries occurred during sporting activities (53 %), and then MVC (26 %). Falls accounted for 13 % of the injuries. Fractures occurred in 67 % of these cases with 26 % incidence of neurological injuries. Overall, MVCs are thought to be the cause of pediatric thoracolumbar fractures in up to 50 % of the patients. Child abuse also accounts for some thoracolumbar fractures in infants and younger children. Types of injuries associated with battering and shaking include fractures of the spinous processes, pars, pedicles, or compression fractures of multiple vertebral bodies [60]. Finally, contrary to the cervical spine trauma, thoracolumbar injuries are more common to children older than 8 years [63].

21.3.2 Anatomy

Anatomic differences between the immature thoracolumbar spine and the adult spine result in different patterns of injury. As with the subaxial cervical spine, the thoracolumbar spine has three centers of ossification: one centrum and two neural arches. Fusion typically occurs between the ages of 2 and 6. The facet joints of the thoracolumbar spine are more horizontally oriented and incompletely ossified resulting in increased intervertebral mobility. The articular facets begin to attain a mature configuration at the age of 8 with complete adult orientation occurring at the age of 15 [60]. Additionally, one should remember that the spinal cord in newborns ends at L3 and migrates during childhood to his final position at L1–L2.

The developing vertebral body consists of two physes located at the superior and inferior end plates. The physes becomes apparent radiographically between the ages of 8 and 12 when apophyseal ossification begins to occur at the cartilaginous end plates. At this time, the end plate is bordered superiorly by hyaline cartilage subjacent to the overlying nucleus and inferiorly by physeal cartilage. Fusion begins at the age of 14–15 years, and physeal lines may be misinterpreted as fractures until the age of 15–21 when complete fusion occurs. Before 12 years of age, the pediatric vertebra has significant remodeling potential after compression fractures. If the wedging deformity is <30°, physeal injury is avoided and complete reconstitution of vertebral morphology can occur. However, a more significant deformity causing physeal injury may result in a deformed vertebral body during the adolescent growth spurt [60]. Furthermore, the immature, wedge-shaped vertebral bodies of the pediatric spine predispose to compression fractures [10].

21.3.3 Mechanisms of Injury

The three main mechanisms of injury are flexion with or without compression, distraction, and shear with hyperflexion being the most common. Hyperflexion results in failure of the anterior column with preservation of the middle column. Although the posterior column may remain intact, a distraction injury may occur with greater degrees of flexion. Fractures resulting from hyperflexion most commonly occur at the

thoracolumbar junction. More significant forces may result in a burst fracture. A burst fracture involves failure of the anterior and middle columns as a result of axial loading. The axial loading drives the nucleus pulposus into the vertebral body with subsequent fractures of the anterior and middle columns. The posterior cortex of the vertebral body is fractured with retropulsion of the fragments into the spinal canal (Fig. 21.9). An increase in the interpedicular distance on plain radiograph typifies this fracture pattern [17, 45].

Distraction injuries, i.e., chance fractures or "seat-belt injuries," usually occur during rapid automobile deceleration in a restricted patient. In this injury pattern, the posterior and middle columns are distracted as the torso is hyperflexed over a lap belt. This results in tension injury to the posterior ligamentous and bony structures. Compression loading may also result in fracture of the vertebral apophysis. This injury is common in the adolescent population and is discussed at length in the sports section. That said, it most commonly occurs at the L4 level, is diagnosed with CT and MRI and increased suspicion is necessary for the well-established association between concurrent abdominal trauma and flexion-distraction injuries [36, 60, 64].

21.3.4 Clinical Evaluation

Comprehensive evaluation of the pediatric patient presenting with thoracolumbar injuries should include institution of the pediatric ATLS protocols. These injuries are often a result of significant trauma such as motor vehicular accidents and associated extraspinal injuries are frequent. A comprehensive history should be obtained when possible. The lucid pediatric patient with thoracolumbar injury will report back pain and should be able to localize the pain. Mechanism of injury, spinal maturity, time of injury, presence of neurological complaints, and associated extraspinal symptoms should be ascertained. A systematic physical examination includes assessment of the airway, breathing, circulation, presence of disability, and the ABCDs of ATLS. This prevents omission of associated injuries. Santiago et al. showed that physical examination was up to 87 % sensitive and 75 % specific for detecting thoracolumbar spine fractures [65]

Subtle spinal injuries are often missed or diagnosed late in the polytraumatized patient. Evaluation of the spine begins with a careful neurological examination. Initial evaluation for sensation at all the four extremities includes assessing for perception of light touch. The patient should also be instructed to move his/her fingers and toes. Inspection and palpation of the entire spine as well as the paraspinal region occurs during the log roll. Bruising, swelling, step-offs, crepitus, or deformity over the spine should be noted. The entire spine should be palpated for tenderness and deformity. A rectal examination should also be performed by paying particular attention to rectal tone. The bulbocavernosus reflex should be elicited as its absence may indicate the presence of spinal shock. Perianal sensation should be assessed. Perception of pain in a dermatomal fashion should be assessed. Reflex testing of the upper and lower extremities should be performed and strength should be graded in a myotomal fashion. Injury to the spine should be suspected in patients with abdominal seat-belt abrasions [66].

Historically, patients with neurological deficits have been administered by intravenous methylprednisolone to minimize cord edema. Patients presenting within 3 h of injury are given 30 mg/kg bolus over 15 min for the first hour, followed by an infusion of 5.4 mg/kg/h for 23 h [67]. Treatment should be extended for an additional 24 h if treatment is started within 3–8 h from injury [68]. Recent studies have, however, questioned the efficacy of the earlier regiment. Sayer et al. [69] and Pettiford et al. [70] in a systematic review of the current literature found insufficient evidence to support the use of methylprednisolone in acute spinal cord injuries.

21.3.5 Radiographic Evaluation

Most vertebral fractures can be visualized on plain radiographs; however, MRI or CT scan may be needed to further define the character of these

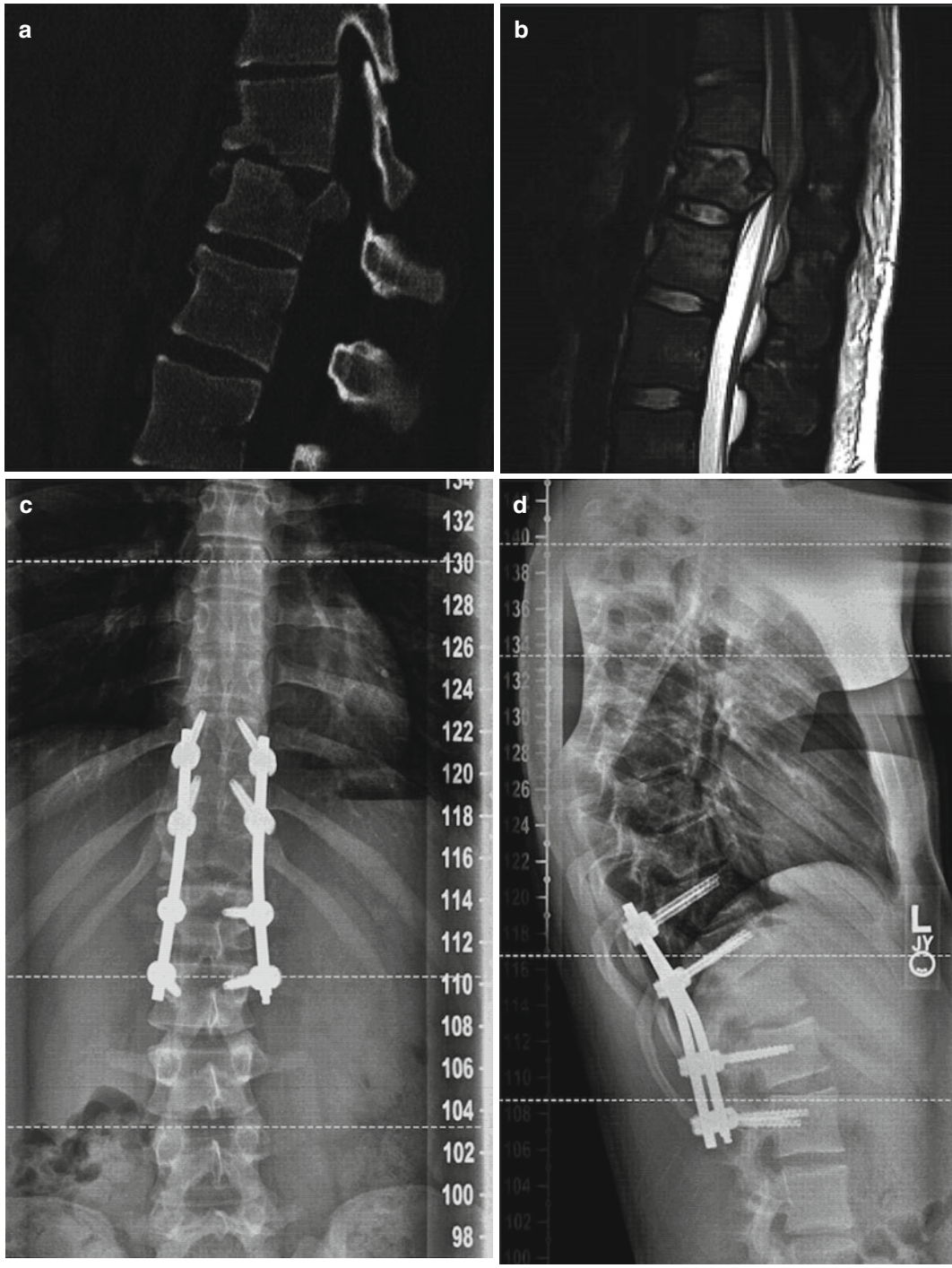

Fig. 21.9 Sagittal CT scan (**a**) demonstrating a burst fracture involving the T12 vertebral body with focal kyphosis and loss of the disk space at the T11–T12 level. There is retropulsion of a 1.3 × 1.0 cm fragment from the T12 vertebral body into the spinal canal. Sagittal T2-weighted MRI (**b**) demonstrating focally increased T2 signal intensity within the spinal cord for a distance of approximately 2.2 cm in the craniocaudad dimension, reflecting cord contusion caused by the retropulsed fracture fragment. Anteroposterior (**c**) and lateral (**d**) radiographs of the spine 5 months after posterior spinal fusion with pedicle screws and instrumentation extending from T10 to L2

fractures. CT is especially useful in the evaluation of the spinal canal after a burst fracture (Fig. 21.9). However, CT benefits should be weighed against the risks associated with ionizing radiation especially in infants and young children. In cases where neurological deficit is present, an MRI should be obtained to assess the extent of cord injury. On T2-weighted images, a high signal suggests cord edema, a mixed signal indicates contusion, and a low signal indicates acute hemorrhage [60].

Radiographic studies can be used to assess for stability in the injured thoracolumbar spine. Findings indicative of instability on radiographs include vertebral body collapse with widening of the pedicles, >33 % canal compromise by fragments of the lamina or middle column, translation of more than 2.5 mm between vertebral bodies in any plane, bilateral facet dislocation, significant distraction of the posterior components with greater than 50 % collapse of the vertebral body. Presence of neurological deficits also suggests spinal instability. On plain radiographs, disruption of two or more columns indicates instability. However, fractures involving more than 2 columns cephalad to T8 can be stable, if the sternum and ribs are intact. Integrity of the sternocostal joint tends to stabilize the thoracic vertebra. In addition, fractures at L4 and L5 can be stable if the posterior elements are intact and if normal lumbar lordosis is maintained [17, 60].

Absence of neurological deficits in a patient who is able to ambulate after injury usually suggests a stable fracture. However, it should be noted that even in the absence of neurological injuries, a fracture may be unstable if there is evidence progression of deformity will occur. These fractures include hyperflexion compression fractures where the posterior ligamentous structures are disrupted. This is reflected on plain radiographs by a greater than 50 % deformity of the anterior column, with an intact middle column. Greater than 20° of flexion of L1 on L2 also indicates disruption of the posterior ligamentous structures. In burst type fractures with compromise of the middle column and retropulsed fragments, neurological insult may occur if axial load is applied prematurely before healing. Consequently, these fractures are considered unstable even in the absence of neurological deficits at presentation [60].

Finally, in the pediatric patient with spine trauma, the entirety of the spine should always be assessed for possible multilevel, contiguous or not, concomitant injuries [10, 65].

21.3.6 Management

Stable injuries of the spine in an adolescent can be managed nonoperatively. Nonoperative management in this setting includes bed rest, adequate analgesia, and spasmolytics for muscle spasms. After adequate analgesia is obtained, mobilization in a thoracolumbosacral orthosis (TLSO) bracing is started and continued for 6 weeks. Fractures amenable to this treatment include minor spinous process fractures, transverse process fractures, wedge compression fractures, and chance fractures. Chance fractures specifically may also be treated with hyperextension casting for 8 weeks; however, despite the immobilization method, one must ensure with standing radiographs proper alignment and well-reduced fracture in the cast or brace before discharge of the patient from the hospital [60, 66]. Burst fractures can also be treated conservatively in the absence of neurological compromise. Integrity of the posterior ligamentous structures especially the posterior longitudinal ligament must be ascertained before nonsurgical management is instituted. Treatment involves early mobilization in a TLSO brace for 8–12 weeks. Rarely, when surgery poses a significant risk because of other comorbidities, unstable fractures may be managed conservatively. This involves 6–10 weeks of bed rest and subsequent immobilization in a TLSO brace for a similar length. The fracture should be assessed frequently with standing radiographs for change in position [71].

Expeditious surgery for thoracolumbar pediatric trauma is indicated in the presence of mechanical instability or neurological deficits. Ultimate outcome is optimized when surgical intervention is expedited. It is recommended that decompression and reduction should be

performed within 12–48 h after injury. The length of instrumentation should be minimized but be adequate for stabilization and correction of the deformity. Pedicle screws, hook, and rod constructs one or two levels above and below the fractured level have been utilized. Indirect reduction of retropulsed fragments is possible with distraction if the posterior longitudinal ligament is intact. In the immature spine, the displaced fragments are often attached to the posterior longitudinal ligament and can be reduced with traction. Surgical management of unstable chance fractures involve compression fixation of the posterior elements to facilitate ligamentous healing. Several different new techniques have been described for the surgical treatment of unstable burst fractures, like minimally invasive percutaneous pedicle screw fixation [72], and anterior or far lateral corpectomy and strut grafting followed by anterior or posterior instrumentation [73]. However, large outcome studies with long-term follow-up have not been reported about these techniques yet. Severe burst fractures with >40 % canal compromise and >15° of kyphosis may require anterior fusion [60, 71].

21.3.7 Outcome Studies for Thoracic and Lumbar Spine Trauma in Pediatric Patients

As in the adult population, favorable outcomes have been reported for stable fractures of the thoracic and lumbar spine managed nonoperatively [74–76]. Parisini et al. [76] recently reported the outcomes of 44 pediatric and adolescent patients treated for spinal trauma. Of these, 29 involved the thoracic and lumbar spine; 58 % of these fractures were unstable. Of the 12 unstable fractures, 41 % had spinal cord injuries. Favorable outcomes, absence of significant deformity, and persistent stability were reported in stable fractures managed conservatively. Likewise, Dogan et al. [74] reviewed the outcomes of 89 pediatric patients treated for thoracic, lumbar, and sacral spine injuries. A majority of these patients (85.4 %) were neurologically intact at presentation. Neurological deficits were more common in patients with injuries at the thoracic level (53.8 %). A majority of the patients were treated conservatively with a 12.6 % average loss of vertebral body height at subsequent follow-up. Moller et al. [77] supported the earlier findings in a long-term outcome of 30 adolescent patients with thoracic and lumbar fractures managed nonoperatively.

Contrary to the earlier findings, conservative management of unstable burst fractures, even in the absence of neurological deficits, often yields unfavorable outcomes. In the Parisini et al. series [76], conservative management of unstable fractures without neurological deficits in the thoracic and lumbar spine resulted in significant deformity at 4 months follow-up. An average of 18–20° of global kyphosis was noted in the four patients with unstable fractures of the thoracic and lumbar spine managed conservatively. That said, equivocal outcomes have been reported with surgical management of unstable thoracolumbar fractures. Erfani et al. [78] reported excellent functional and radiographic results in 20 patients with a mean follow-up of 49 months, while Parisini et al. [76] reported a high incidence of subsequent progressive deformity in unstable fractures managed surgically regardless of neurological status at presentation. Six of the nine patients with unstable thoracic and lumbar spine fractures managed surgically had significant spinal deformity at follow-up. Dogan et al. [74], however, showed a low incidence of progressive deformity (8.6 %) in patients managed surgically in his series. Of note is the high incidence of spinal deformity in patients with neurological injury before puberty [76, 79]. In Parisini et al. series, 72 % of patients with neurological deficits proceeded to develop significant scoliosis and/or kyphosis regardless of management. This is consistent with the >90 % incidence of post-SCI deformity reported in the literature. Deformity in this setting has been attributed to impaired muscular function [76, 79, 80]. Some have advocated the prophylactic bracing of curves <10° in order to prevent surgical correction. Mehta et al. [81] showed that in children with scoliosis subsequent to acquired spinal cord injury initiation of bracing in curves less than 20°

(and more than 10°) resulted in delayed surgery whereas bracing initiated later, after 20° of spinal deformity, was less likely to control the curve and prevent surgical correction. A recent study by Angelliaume et al. [82] showed that the group with Risser sign 3 or above, a single vertebral fracture, and lumbar fracture had more severe post-SCI coronal deformity.

Conclusion

In conclusion, spinal trauma though rare in the pediatric population can have devastating consequences. Neurological impairment due to spinal cord injury is highly related to participation and quality of life, as well as social integration [83]. Hwang et al. [84] reported that in adults with pediatric-onset spinal cord injury not only employment odds decreased with occurrence of autonomic dysreflexia, spasticity, or chronic medical condition, but also odds of depression increased over time in those who remained unemployed.

Clinicians treating trauma in the immature spine should be cognizant of the anatomical and biomechanical features that account for the pattern of injuries seen in this population. Furthermore, the relative high prevalence of SCIWORA in this population underscores the need for comprehensive history and physical examination. Prompt diagnosis of neurological deficits and expeditious management in the setting of trauma enhances outcomes.

An emphasis should be placed on prevention. As indicated earlier, motor vehicular accidents in the young child and sports injuries in the adolescents are the most common mechanisms of injury in the pediatric population. Attempts at prevention should therefore be mechanism directed. Given that MVCs are major contributors of pediatric spine trauma seat belt and child safety policies and guidelines are of utter importance. Characteristically, Brown et al. [11] found that 81 % of patients who sustained cervical spine injuries in MVCs were either unrestrained or inappropriately restrained. Table 21.2 presents the recommendations issued by the American Academy of Pediatrics regarding proper use of seat belts and child safety [85].

Table 21.2 Car safety seats guidelines for growing children

Best practice recommendation	Details
Infant-only or convertible CSS used rear-facing	All infants and toddlers should ride in a rear-facing car safety seat (CSS) until they are 2 years of age or until they reach the highest weight or height allowed by the manufacturer of their CSS
Convertible or combination CSS used forward-facing	All children 2 years or older, or those younger than 2 years who have outgrown the rear-facing weight or height limit for their CSS, should use a forward-facing CSS with a harness for as long as possible, up to the highest weight or height allowed by the manufacturer of their CSS
Belt-positioning booster seat	All children whose weight or height is above the forward-facing limit for their CSS should use a belt-positioning booster seat until the vehicle lap-and-shoulder seat belt fits properly, typically when they have reached 4 ft 9 in. in height and are between 8 and 12 years of age
Lap-and-shoulder vehicle seat belt	When children are old enough and large enough to use the vehicle seat belt alone, they should always use lap-and-shoulder seat belts for optimal protection
All children younger than 13 years should be restrained in the rear seats of vehicles for optimal protection	All children younger than 13 years should be restrained in the rear seats of vehicles for optimal protection

Based on data from Ref. [85]

CSS car safety seat

In the adolescent athlete, use of appropriate sporting equipment such as helmets, as well as rules targeted at safety should be instituted. Furthermore, proper conditioning with attention to strengthening of paraspinal muscle groups should be encouraged. Proper and safe techniques such as avoidance of head-first tackling should also be practiced [33].

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Sports and Sports-Related Injuries in the Growing Spine

22

R. Justin Mistovich and Keith Baldwin

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Key Points

- Up to 53 % of spinal cord injuries are a result of sports-related injuries; the sideline physician must know how to appropriately diagnose and treat.
- Back pain has many different causes, but it affects 80 % of adolescent athletes. Proper physical examination and appropriate diagnostics and direct treatment algorithms.
- Vertebral apophyseal avulsion fractures can present similarly to an acute herniated disc in adolescents.

22.1 Introduction

Participation in recreational and competitive sports continues to rise among children and adolescents, resulting in an increased incidence of musculoskeletal injuries. Sports-related injuries are the most common cause of pediatric and adolescent injury-related emergency room visits, accounting for nearly 10 % of all emergency room encounters in this age group. Most concerning, nearly 40 % of all life-threatening injuries children and adolescents sustain are sports-related [1]. For example, among high school and college athletes playing American football, there is one fatality

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per 100,000 participants each season typically resulting from injury, heat illness, or underlying cardiac condition. While most injuries are not fatal, they still can result in substantial time lost from sport participation and carry long-term sequelae. Furthermore, they are the predictable genesis of many questions from both the patient and his or her family.

We review specific injuries affecting the young athlete that are both characteristic and common in the growing spine and provide recent evidence on both the diagnosis and management. The scope of sports injuries ranges from acute on-field trauma to classic office chief complaints.

22.2 Acute On-Field Trauma

22.2.1 Cervical Spine Injuries

Nearly 10 % of all new cases of paraplegia and quadriplegia in the United States result from athletic injury [2]. Other studies of all spine and spinal cord injuries have found 27–53 % resulted from participation in sports. These injuries were more common in the adolescent population, and most commonly involved the upper cervical spine [3, 4]. In American football, the incidence of catastrophic cervical spine injury peaked in 1976 as a result of improved helmets imparting a false sense of safety upon athletes who spear tackled (leading with the head into a tackle). With the banning of spear tackling, the injury rate dramatically dropped, illustrating the importance of training young athletes in proper techniques [2]. While football has the highest incidence of cervical spine injuries, many other sports also put the athlete at risk, including gymnastics, cheerleading, diving, hockey, pole vaulting, skiing, and snowboarding.

22.2.1.1 Cervical Spinal Cord Neuropraxia

Cervical spinal cord neuropraxia (CSCN), also referred to as transient neuropraxia or transient quadriplegia, is a transient albeit real spinal cord injury. CSCN results from hyperextension, hyperflexion, or axial loading of the cervical spine. The bony elements of the spine act as a pincer mechanism on the cord. While older patients with cervical stenosis are at elevated risk, in contrast to the adult athlete, current evidence suggests that many cases in young patients result from the hypermobility of the developing cervical spine rather than stenosis [5]. That is, there is no anatomic stenosis in the child, but the injury combined with their ligamentous laxity results in cord neuropraxia.

Patients complain of symptoms affecting more than one extremity, which can include sensory changes such as numbness or burning and a variable degree of motor weakness. Symptoms may last from seconds to 36 h, though complete recovery is the rule [6]. It is critical to distinguish CSCN from a burner or stinger. CSCN causes multiple extremity symptoms. Even if mild or transient, bilateral symptoms indicate a spinal cord injury. In contrast, a burner or stinger is a transient brachial plexus neuropraxia, causing *single* upper extremity numbness, tingling, and/or weakness. Patients with burners or stingers may return to play when symptoms have completely resolved [7]. However, patients with bilateral extremity symptoms must be treated as a spinal cord injury and are not cleared for return to play.

Initial on-field management should follow ATLS protocols. The cervical spine must be immobilized. If a football injury, the helmet should not be removed. Rather, the patient should be immobilized on a rigid board with the neck in neutral alignment and the helmet left on. To access the airway, the facemask alone on the helmet should be removed [8]. Once acutely stabilized, the patient should be taken to the emergency department for a complete clinical and radiographic evaluation, including MRI imaging of the cervical spine, to fully assess for bony or ligamentous injuries as well as space available for the cord [6, 9, 10]. While controversial, the use of high-dose steroids can be considered, weighing the potential benefits, comorbidities, and specific institutional protocols [11].

In the absence of radiographic evidence of fracture or ligamentous disruption, management consists of immobilization in a hard collar.

Immobilization may be discontinued when symptoms completely resolve and the patient has a pain-free ROM with normal strength. If pain persists, immobilization should be continued and the patient should be revaluated in 2 weeks. Collar immobilization can be discontinued in the presence of a normal neurological examination, normal dynamic and static cervical spine radiographs, and complete resolution of symptoms. Some authors have suggested completion of a physical therapy program that includes strengthening of the paracervical muscles before returning to sports [12]. Dailey et al. made a strong recommendation that patients with CSCN and without cervical stenosis can safely return to sports after symptoms have resolved. They also recommended (with weaker evidence to support it) that patients with CSCN and radiographic evidence of cervical canal compromise should not continue participation in contact sports [6].

22.2.1.2 Spinal Cord Injury

Complete and incomplete spinal cord injuries may result from athletic injury. One study identified that nearly $700 million is spent annually treating sports-related spinal cord injury in the United States [2]. Patients with spine fractures and spinal cord injuries require standard ATLS management, including rigid spinal immobilization and prompt transfer to a trauma center for assessment, imaging, and definitive treatment.

Spinal Cord Injury Without Radiographic Abnormality

One subset of spinal cord injuries unique to the pediatric population is spinal cord injury without radiographic abnormality, or SCIWORA. Described by Pang and Wilberger in 1982, SCIWORA is described as cord and neural damage in children without radiographic fracture, malalignment, or dislocation [13]. It results from the elasticity of the vertebral columns in children, allowing the mobile vertebral elements to impinge on the spinal cord. Spinal cord injury may continue to evolve and progress after an initial injury.

Affected children, particularly those younger than 9 years of age, may have complete cord injuries, as neurological lesions are more severe in younger children because the cervical spine is more unstable in these children. Outcome has been related to neurological status at presentation. Children with complete lesions seldom improve, those with incomplete lesions improve but not to normal levels, and those with mild or moderate deficits often have a full recovery. The MRI appearance of the spinal cord has also been demonstrated to predict neurological outcomes, with absence of cord signal changes indicative of excellent outcome [14].

Treatment has been controversial, with the duration of cervical immobilization (and even its need altogether if no instability is present) unclear; however, a recent study has made level III recommendations that immobilization is recommended for up to 12 weeks, with the possibility of earlier discontinuation in patients who become asymptomatic and have normal flexion and extension imaging, indicating a stable spine. Furthermore, high-risk activities should be avoided for 6 months [15].

22.3 Common Chief Complaints in the Growing Spine

22.3.1 Back Pain in the Pediatric Athlete

Back pain is common in pediatric athletes, affecting up to 80 % of participants in some studies [16]. Kujala et al. found a higher incidence of back pain in adolescent athletes than nonathletes (18 % of whom are still afflicted) [17]. Back pain can result in lost playing time for up to 40 % of athletes. Sports including gymnastics, wrestling, football, soccer, and tennis have high associated rates of back pain. Gymnasts have the highest risk of back pain, with one study demonstrating over 80 % of enrolled gymnasts reported low back pain in a 7-week period [16, 18]. Goldstein et al. showed a higher incidence of lumbar spine abnormalities on MRIs of gymnasts than swimmers. This difference was attributed to the repetitive stress placed on the lumbar spine by the gymnast [19]. A history of prior low back pain

has been demonstrated to be a risk factor for recurrent injuries in varsity athletes [20]. These numbers, however, must be considered in the global perspective: the lifetime prevalence of low back pain in the adult population is estimated to be 85–90 % [21].

Micheli and Wood demonstrated a higher incidence of identifiable pathology in a population of young athletes presenting with back pain when compared with adult patients with similar complaints [22]. However, more recent studies suggest that the incidence of both pediatric and adolescent back pain is increasing, while the proportion of patients having a diagnosable pathology is decreasing. In fact, one recent large cohort found no diagnosable pathology in 78 % of patients [23]. Therefore, for unclear reasons, but possibly as a result of higher intensity athletic participation, the pattern of back pain in the pediatric population is becoming more similar to that of adults. This only adds to the complexity facing the clinician, who must perform a careful history and physical examination, order appropriate tests for "red flags," and not miss serious pathology.

Common causes of back pain in the pediatric athlete include muscle strain and stress fractures such as spondylolysis and pediculolysis. Less common causes include spondylolisthesis and lumbar Scheuermann's disease. Importantly, one must identify neoplastic or systemic diseases presenting with back pain. Lumbar vertebral fractures are also a common cause of back pain in the adolescent athlete (*please see* Chap. 21).

22.3.1.1 Evaluation

Evaluation of the adolescent athlete with back pain involves obtaining a thorough history and physical examination, complemented by the appropriate, indicated imaging studies.

22.3.1.2 History

The exact location, onset, duration, and severity of pain should be elucidated. Pain localized to the lower back and gluteal regions is likely secondary to a mechanical etiology, while leg pain is likely a result of nerve compression or irritation. Aggravating activities and relieving factors should also be determined. In general, pain exacerbated by flexion suggests discogenic pathology, while pain worsened by extension suggests an injury to the posterior elements.

Additional important historical information includes the type of sport and level of competition the patient is involved in. The literature has demonstrated an association between certain sports and specific types of injuries. For example, spondylolysis and spondylolisthesis have been frequently associated with weightlifting, diving, wrestling, and gymnastics. Congeni et al. reported a 47 % incidence of spondylolysis in athletes participating in gymnastics and diving [24]. These sports involve repetitive hyperextension, a proposed mechanism of injury for spondylolysis.

A comprehensive review of systems should be performed in the adolescent athlete presenting with back pain. The presence of significant weight loss, night pain, fever, or urinary symptoms should spur further investigation of systemic causes of back including neoplasm, infection, or renal pathology. Finally, a history of previous treatments received by the patient should be reviewed.

22.3.1.3 Physical Examination

Examination begins with inspection of the back. Assess spinal curvature, as a scoliotic or kyphotic spine in the adolescent athlete can result in back pain. Next, palpate the thoracolumbar spinous processes, paraspinal muscles, and sacrum for point tenderness or masses. Quantify range of motion in the coronal plane by evaluating lateral bending, and the sagittal plane by evaluating forward flexion and extension. Any pain associated with range of motion should be noted. Test provocative maneuvers, including the straight leg raise test for disc herniations and the single leg hyperextension test for spondylolysis. Perform a comprehensive neurological examination, including motor strength testing, sensory examination, and reflex testing. Examine the hip, assessing range of motion and palpating bony anatomy, as hip pathology may be manifested as back pain. Finally, assess the patient's gait.

22.3.1.4 Imaging

Obtain standard plain radiographs, including AP and lateral views of the lumbar spine upon the

initial examination for patients with "red flags," including history of trauma, pain unrelieved by rest, night pain, constitutional symptoms such as fever, chills, or weight loss, neurological dysfunction, bony tenderness or step-off, pain with provocative tests, or abnormal spinal alignment. Patients with suspected benign, mechanical back pain, or muscle strain who fail to improve after a 4–6-week course of rest and non-narcotic analgesics also warrant radiographic evaluation. However, routine, reflexive radiographs for all patients presenting with back pain prior to physical examination and identification of "red flags" expose many young patients to unnecessary ionizing radiation without changing their clinical management [25].

A recent study evaluated the necessity of the traditional oblique views for diagnosing spondylolysis, concluding that two-view PA and lateral studies had the same sensitivity and specificity as four-view studies that added oblique views. While a classic teaching, the "Scotty dog" view may not have a diagnostic benefit [26]. Flexion and extension views are warranted if instability is suspected. Bone scan, SPECT (Fig. 22.1), CT scan, and MRI should be obtained when indicated.

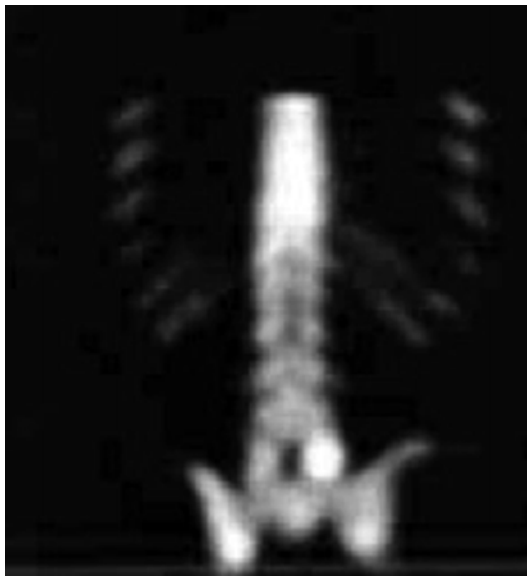

Fig. 22.1 SPECT

22.3.2 Muscle Strain

Muscle strains are a common cause of back pain in the pediatric and adolescent athlete. They have been estimated to occur in 27 % of adolescent athletes presenting with lower back pain [16, 27]. Strains result from disruption in muscle fibers within the muscle belly or musculotendinous junction.

Numerous factors such as acute trauma, repetitive stress, poor technique, obesity, muscle imbalance, and poor footwear can contribute to muscle strain. Patients with a spinal muscle strain present with progressively worsening lower back pain and possible spasms. Symptoms peak 24–48 h after onset. Diagnostic testing such as plain radiography, bone scan, and MRI are usually negative and do not contribute to the diagnosis. However, if patients present with any "red flags," imaging is necessary to rule out other etiologies [16].

Many children presenting with "mechanical back pain" are experiencing failure of their core muscles during exertional activities. After a growth spurt, the adolescent athlete often develops transient lower extremity muscle contractures, further altering the stresses acting upon the lumbosacral junction. Treatment should focus on lower extremity stretching and core strengthening exercises.

22.3.3 Lumbar Scheuermann's Disease

Lumbar Scheuermann's disease is another potential cause of back pain in the adolescent athlete. It is an overuse injury involving the lumbar spine that occurs in sports requiring repetitive flexion and extension. Male weight lifters and football players are most commonly affected. Patients present with localized back pain exacerbated by forward flexion. Pain may be associated with muscle spasm and loss of lumbar lordosis. Neurological deficit is a rare finding.

Radiographic evaluation of Scheuermann's disease involves a plain lateral radiograph of the lumbar spine. Interestingly, lumbar Scheuermann's

disease may not present with the classic anterior wedging deformity (greater than 5° of anterior wedging across three consecutive vertebral bodies) as is associated with the thoracic variant. The original paper by Blumenthal, Roach, and Herring found that only 6/13 patients had classic anterior wedging. The remainder presented with anterior Schmorl's nodes—and all of these atypical cases presented with pain [28–30].

Lumbar Scheuermann's disease can be managed conservatively. Activity modification and lumbar bracing may facilitate pain control. Physical therapy should be directed at pelvic and lumbar stabilization exercises. The patient may return to play when pain subsides and after completion of physical therapy [29].

22.3.4 Back Pain Secondary to Stress Fractures

Spinal stress fractures are a common cause of back pain in the immature athlete (Fig. 22.2). It is particularly common in athletes participating in sports requiring repetitive hyperextension and flexion [31]. Stress fractures result from repetitive low-intensity load and microtrauma to the bone. Two theories have been postulated to explain the occurrence of stress fractures. In the overload theory, repetitive rhythmic contractures of muscles result in stress at their osseous insertions, consequently reducing the mechanical resistance of the bone. The muscle fatigue theory attributes stress fractures to diminished shock absorbing properties of a fatigued muscle subjected to repetitive stress. This leads to aberrant loading of bone and subsequent failure [22, 32].

The immature adolescent spine is particularly susceptible to stress fractures because of the absence of complete ossification. Areas of incomplete ossification within the vertebrae are weak points that become susceptible to failure when subjected to repetitive compressive, torsional, or distraction forces [31]. Adolescent athletes participating in sports such as gymnastics, weightlifting, or diving among others expose their spine to such forces thereby increasing the risk of stress injury. Of note, there is an additional risk seen in competitive female athletes suffering from the female athlete triad syndrome: disordered eating, amenorrhea,

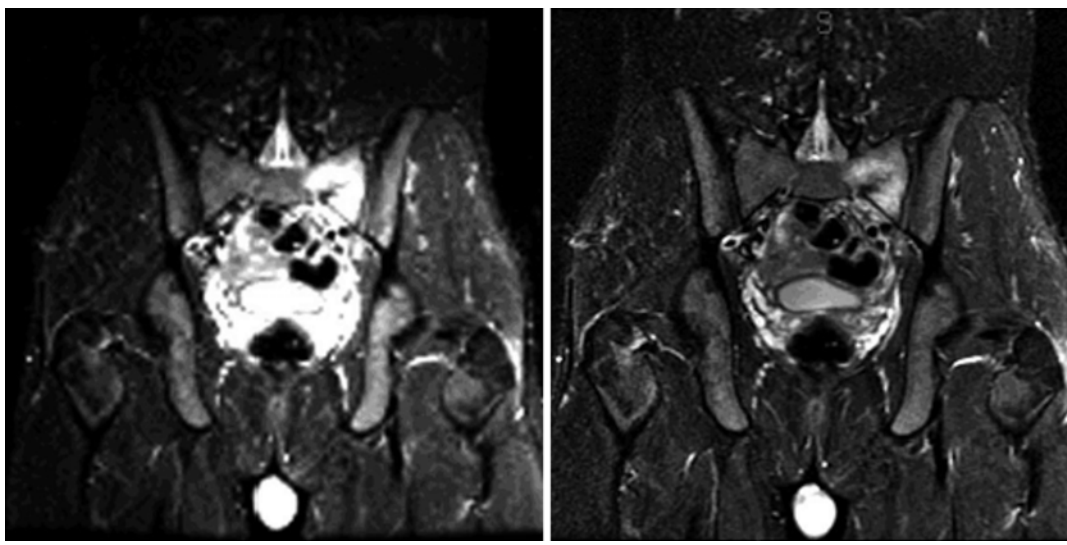

Fig. 22.2 A 17-year-old boy presented with 3 weeks of left lower back and sacral pain. While he had no specific injury, his symptoms began during a 13-mile cross-country run. He notes the pain was 6 out of 10 at its worse. Past medical history reveals no prior illness, injuries, surgeries, or hospitalization. On physical examination, his pain was localized with palpation along the left posterior superior iliac spine. Radiographs show no evidence of stress fractures or other osseous abnormalities. MRI of the pelvis reveals a left sacral stress fracture with soft tissue edema

and osteoporosis. Both osteopenia and their propensity for overexertion increase the risk of stress fractures in this population. It is important to evaluate female adolescent athletes for this condition with both the history and physical examination, and begin a multidisciplinary approach to treatment if symptoms are present [22].

22.3.4.1 Spondylolysis/ Spondylolisthesis

Spondylolysis is a common cause of back pain in the adolescent athlete (Fig. 22.3). Many studies have investigated the incidence of spondylolysis in the adolescent athlete. Notably, Micheli and Wood reported 47 % incidence of spondylolysis in adolescent athlete presenting with back pain [22]. However, Rossi reported a much lower incidence of 15 % in a review of radiographs of adolescent athletes [33]. Additionally, Drummond et al investigated nearly 3,000 adolescent patients presenting with low back pain, diagnosing only 7.8 % of them with spondylolysis [25].

Spondylolysis is a defect in the vertebral pars interarticularis. It most commonly occurs at L5–S1 resulting from an L5 pars defect, though it may also occur at more cephalad levels. Subsequent translation of the cephalad vertebral body on the caudad vertebral body describes

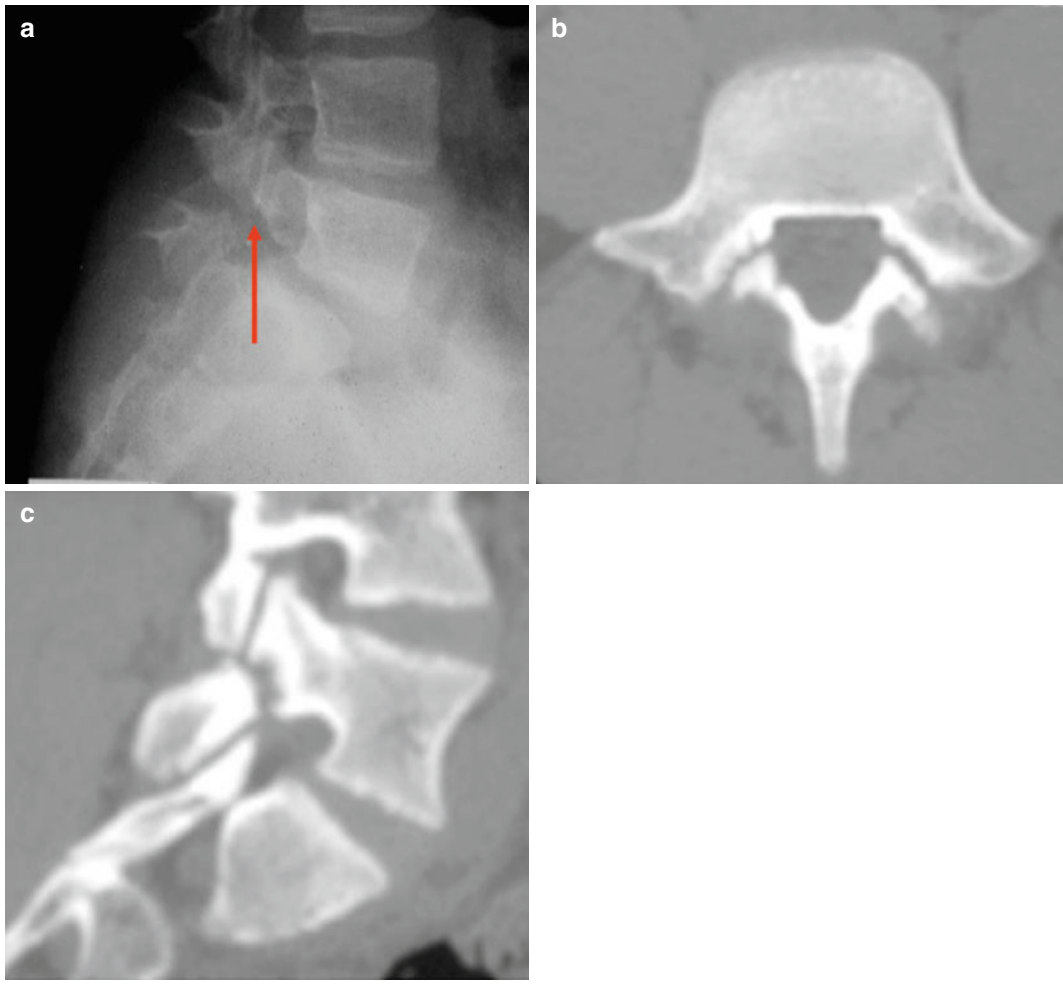

Fig. 22.3 Imaging of a 13-year-old elite level gymnast with 2 years of back pain who was diagnosed with spondylolysis. (**a**) Lesion visible on lateral radiograph with arrow demonstrating spondylolysis (**b**) and (**c**) CT images further define the stress fracture. Note the sclerotic margins on the transverse cut. This is a typical finding of a long-standing fracture with failure to heal

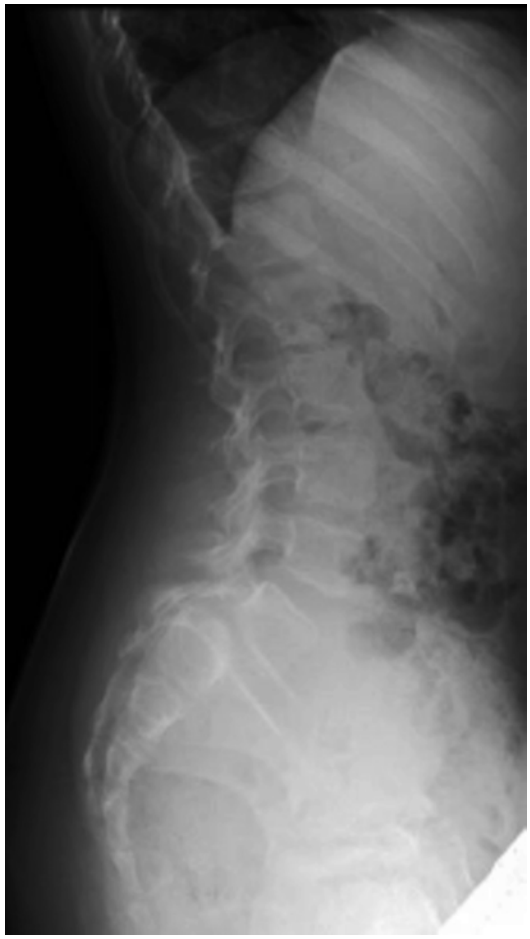

Fig. 22.4 Plain radiographic lateral view of an 8-year-old female with Grade III spondylolisthesis

spondylolisthesis (Fig. 22.4) [34]. For more detail, please refer to Chap. 24 on Spondylolysis/Spondylolisthesis.

Clinical Presentation

Spondylolysis is often asymptomatic and found incidentally. Athletes who do develop symptoms typically report focal back pain with occasional radiation to the buttocks or proximal thighs. The pain may either be insidious in onset or associated with an inciting traumatic event. Patients may also report an acute worsening of chronic mild pain after a new injury. Pain is often exacerbated by activities requiring repetitive flexion and extension of the lumbar spine [35]. Hamstring tightness is also a common complaint of patients with spondylolysis, especially in the presence of advanced spondylolisthesis. This may be manifested by a shortened gait stride with flexion at the hips and knees. Radicular symptoms are rare in spondylolysis and low-grade spondylolisthesis, but may be present with high-grade spondylolisthesis [34].

The one-legged hyperextension maneuver is considered by many to be pathognomonic for spondylolysis. In this maneuver, pain is elicited with a one-legged stance and lumbar extension. If the lesion is unilateral, pain is often localized to the ipsilateral side. Other findings include a hyperlordotic thoracolumbar posture, which compensates for a loss of lumbar lordosis. Neurological deficits are usually absent unless in the presence of significant spondylolisthesis.

22.3.4.2 Pedicle Fractures

A component of the neural arch, the pedicle is also vulnerable to fracture from the cyclic loads stressing the lumbosacral spine during sports. However, pedicle stress fractures are much less common than pars stress fractures [22]. This is partly secondary to the greater intrinsic strength of the pedicles and shorter moment arm from the vertebral body. The pedicles are therefore capable of resisting greater shear forces than the pars. In a biomechanical study evaluating the mechanical strength of 74 vertebral units subject to repetitive mechanical loads, Cyron and Hutton demonstrated five pedicular stress fractures compared to 55 pars fractures [36]. Contralateral pedicular fractures have been associated with isthmic pars fractures. This phenomenon has been attributed to increased and aberrant loading of the pedicles that result from an unstable neural arch following a pars fracture [37–39]. Ulmer et al. showed a 40 % incidence of reactive changes on MRI in pedicles with contralateral spondylolysis [40].

Radiographic evaluation of pedicular fractures involves plain radiographs and CT scan. CT scan has a higher accuracy in the evaluation of these patients. When visible, hypertrophy or sclerosis is seen on the involved pedicle. Pediculolysis is clearly demonstrated as a linear lucency at the base of the involved pedicle on CT. In addition,

presence of a concomitant pars defect can also be identified [39, 41]. When evaluating patients with pediculolysis, one must consider osteoid osteoma and osteoblastoma in the differential diagnosis. These entities can be differentiated from a pediculolysis by the absence of a nidus, the presence of contralateral pars defect, or the presence of a linear defect in a pedicular stress fracture. Differentiating these lesions is important because pedicle excision—a management option for symptomatic tumor—may further destabilize an already unstable neural arch [39, 42].

Early diagnosis of a pedicular stress fracture and prompt treatment is necessary to facilitate rapid return to sports. Management of pedicular stress fractures is similar to that of a pars stress fracture. Initial management is conservative and involves rest and bracing. Surgery is indicated in the setting of failed conservative treatment, though surgical treatment is rarely necessary. Techniques described in the literature include compression fixation of the defect, excision of the hypertrophic defect with lateral fusion, and bone grafting of the pars and pedicular defects with subsequent pedicle screw fixation [37, 39, 43].

22.3.5 Radicular Pain in the Growing Athlete

22.3.5.1 Disc Herniation

Disc herniation is a rare cause of back pain in the adolescent athlete (Fig. 22.5). Only 1 % of disc herniations occur in the second decade [44]. As opposed to an incidence 48 % in the adult population, only 11 % of adolescent athletes present with symptomatic disc herniation. The most common location of herniation in the young athlete is the L4–L5 and L5–S1 level. In the skeletally immature patient, the disk may be herniated into the vertebral end plate with axial loading, demonstrating a Schmorl's node on radiographs [29, 45].

Young athletes with disc herniations typically present with localized back pain and paraspinal muscle spasm. Impingement of the nerve roots may result in radicular symptoms. Radicular symptoms warrant an MRI evaluation. In the absence of neurological compromise, conservative management is recommended, consisting of rest and activity modification. Prolonged bed rest is discouraged. NSAIDs, muscle relaxants, and oral corticosteroids may also be beneficial. The majority of adolescents can be treated nonoperatively. However, those with progressive neurological deficits or debilitating pain that has failed conservative management are candidates for a microdiscectomy. Patients are usually able to return to full participation in sports [45, 46]. While more applicable to adult patients as it examined an adult patient population, the SPORT trial examined the effects of operative versus nonoperative management for patients with a mean age of 42 who presented with symptomatic lumbar herniated discs. This multicenter randomized controlled trial concluded that at 2 years, patients treated surgically and nonoperatively both demonstrated substantial improvement and could not make conclusions about the superiority of one treatment over the other [47].

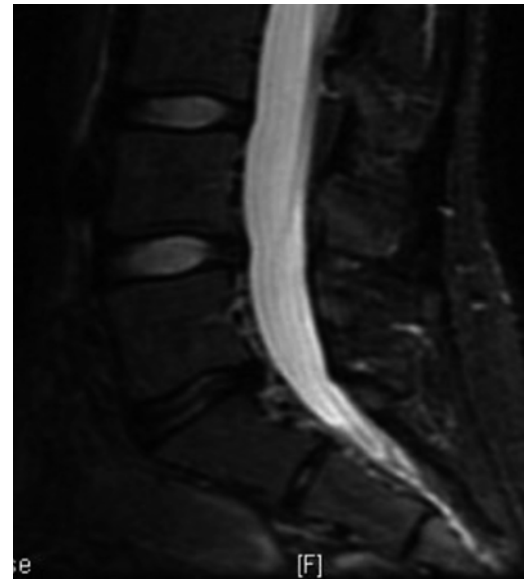

Fig. 22.5 Disc herniation

22.3.5.2 Vertebral Apophyseal Avulsion Fractures

An apophyseal ring fracture is a rare injury in the pediatric population, occurring primarily in the male adolescent athlete (Fig. 22.6) [48]. It most

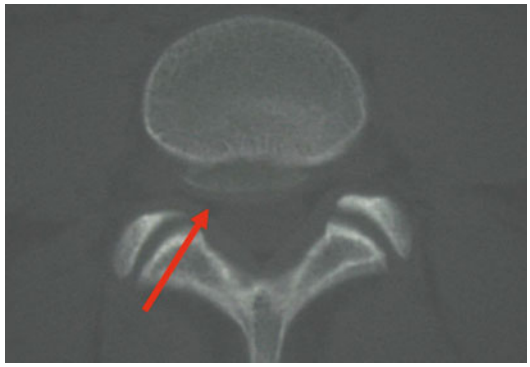

Fig. 22.6 Vertebral apophyseal ring avulsion fracture with arrow demonstrating fracture fragment

frequently affects L4 and L5. The incidence of apophyseal fractures may be underreported as it is frequently misdiagnosed as lumbar disc herniation [49].

In the developing lumbar spine, the superior and inferior endplates are bordered by the vertebral ring apophysis. Ossification of the apophysis commences at approximately 5 years with fusion by age 18. During the rapid adolescent growth phase, the apophysis is separated from the underlying vertebral body by a physeal zone. This area of weakness is susceptible to avulsion injuries. It has been suggested that hyperextension of the lumbar spine or rapid flexion with axial load may be the causative mechanism [48].

Clinical presentation includes low back pain with potential radicular symptoms. Neurogenic claudication, paresis, and cauda equina syndrome has also been reported. The most common finding on examination is restricted lumbar range of motion. The straight leg raise test may be positive in these patients though frank neurological deficits are usually rare [49, 50].

Radiographic evaluation involves the use of both the plain radiograph and CT scan. Bony defects can be visualized on the lateral radiograph; however, the axial CT scan images have been proven to be more sensitive [51].

Takata et al. reported on a large series of apophyseal injures, classifying these injuries into three categories based on CT findings. Type 1 fracture is a separation of the posterior rim of the vertebra body with negligible defect in the vertebra body. This type was noted primarily in patients between the ages of 11 and 13. Type 2 describes an avulsion fracture of the posterior rim of vertebra body including the posterior segment of the overlying annulus fibrosis. It is more prevalent in older patients. Type 3 fracture is a small localized fracture posterior to an irregularity in the cartilaginous endplate [49].

In the absence of neurological deficits, initial management is conservative and includes use of NSAIDs, lumbar bracing, and activity restriction. However, many patients will not improve with conservative care as a result of the bony component of the injury. Surgery is indicated for pain refractory to conservative management and the presence of neurological deficits. Surgery includes laminectomy and excision of the fractured fragments. A discectomy should be avoided in the adolescent patient without evidence of disc degeneration [52, 53].

22.4 Return to Sports After Spine Surgery

Dailey et al. examined the literature to determine optimum criteria for return to play after single-level anterior cervical decompression and fusion performed for athletes who had cervical stenosis. They made a strong recommendation that patients who had undergone a single-level ACDF to correct cervical neurological compression could return to full-contact sports, provided that the athlete had a solid fusion, normal cervical range of motion, and no residual neurological deficits [6].

The Spinal Deformity Study group recently published on return to sports after surgical correction of adolescent idiopathic scoliosis. A survey of 23 expert spinal surgeons was administered. They found that using pedicle screw instrumentation allowed for earlier return to all sports. Most surgeons allowed a return to running by 3 months, contact sports by 6 months, and collision sports by 12 months. However, 20 % of surgeons surveyed never allowed return to collision sports regardless of the construct used. On the contrary, all surgeons allowed eventual return to contact sports. Only one respondent reported hardware failure after surgery

as a result of athletic activity (in a patient who was snowboarding 2 weeks postoperatively) [56].

Rubery and Bradford surveyed a number of SRS members on the appropriate timing of return to play after spine surgery. Factors affecting the decision to initiate sports after scoliosis surgery included time of surgery, instrumentation, and type of sport. They additionally noted that evidence of radiographic union and time from surgery were important determinants in allowing return to play in patients undergoing surgery for spondylolisthesis. Most surgeons initiated non-contact sports after 6 months and allowed return to contact sports after 12 months [54]. However, a small percentage prohibited resumption of contact sports permanently. A majority of physicians discouraged resumption of collision sports such as football.

Li and Hresko examined return to play criteria in athletes undergoing lumbar spine surgery. Their review of the literature for athletes undergoing discectomy noted that 90 % of collegiate athletes who underwent a single-level discectomy were able to return to varsity play; however, athletes who had multilevel procedures were unable to return to play secondary to continued pain. They also noted that most athletes were released to full sports participation 8–12 weeks postoperatively.

Their literature review for spondylolisthesis noted that return to play criteria for contact sports is controversial, with nearly half of surgeons in one survey prohibiting return to collision sports and many waiting between 6 months and 1 year before allowing return to contact sports [55].

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Sagittal Plane Alignment and Deformities in Growing Children

23

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Key Points

- Physiologic sagittal plane alignment in children is age-related.
- Kyphosis in children may be secondary to idiopathic, congenital, neuromuscular, infectious, posttraumatic, syndromic, and postsurgical etiologies.
- An evolving understanding of sagittal plane alignment and of the measurement of spine growth is improving our knowledge of the effects of surgery on sagittal plane alignment in children.

Abbreviations

C-EOS	Classification for early onset scoliosis
CT	Computed tomography
MRI	Magnetic resonance imaging
PA	Posteroanterior
PI	Pelvic incidence
PJK	Proximal junctional kyphosis
PSO	Pedicle subtraction osteotomy
PT	Pelvic tilt
PVCR	Posterior vertebral column resection
SS	Sacral slope
TB	Tuberculosis
TLSO	Thoracolumbosacral orthosis

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23.1 Normal Sagittal Plane Alignment in Growing Children

At birth, the sagittal profile of the spine is C-shaped and globally kyphotic. Once infants obtain the ability to maintain independent head control, cervical lordosis develops and when infants are able to stand, lumbar lordosis develops. Cervical lordosis continues to develop in order to balance the head over the center of the pelvis and to optimally position the head for horizontal gaze. In a well-balanced adult spine, the C7 plumb line passes vertically from the centroid of the C7 vertebral body and intersects the posterior superior corner of the first sacral end plate [1]. Young children have been found to have positive sagittal balance, which diminishes throughout childhood and adolescence (age 3–6 years = +2.5 ± 4.3 cm, age 7–9 years = +0.7 ± 4.6 cm, age 10–12 years = −0.1 ± 4.1 cm, and age 13–15 years = −0.9 ± 4.4 cm) [2].

The pelvis, which is the foundation for the spine, is considered to be of crucial importance for the formation of this normal sagittal profile [3]. A key concept in the understanding of sagittal plane alignment is the relationship between pelvic incidence (PI), pelvic tilt (PT), and sacral slope (SS) [3] (Fig. 23.1). Pelvic incidence is a measure of the orientation of the pelvis in space and, regardless of age, it is the sum of pelvic tilt and sacral slope. Pelvic tilt is a measure of pelvic retroversion and its role is to keep the sacrum posterior to the axis of the hips in order to maintain the center of gravity over the lower extremities to maintain standing balance [4]. Sacral slope is a measure of the sagittal tilt of the first sacral end plate with respect to the horizontal reference plane.

These sagittal spinopelvic values for children have been found to differ from those reported for adults but the correlations were similar [5]. Prior to walking age, the sacrum is less curved and the first two sacral vertebra are more oblong than in adults, which results in smaller pelvic incidence and, ultimately, smaller pelvic tilt and sacral slope than in adults [6]. Spinopelvic parameters on 167 children aged 3–10 years demonstrated

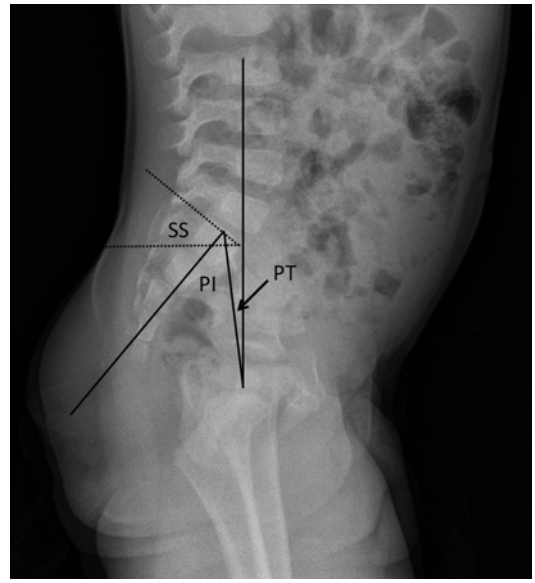


Fig. 23.1 Radiograph of a skeletally immature patient demonstrating sagittal plane pelvic parameters. *PI* pelvic incidence, *PT* pelvic tilt, *SS* sacral slope

pelvic incidence of 44° ± 9°, pelvic tilt of 6° ± 8°, and sacral slope of 38° ± 8° [7], while a similar study on healthy children younger than 10 years old demonstrated pelvic incidence of 45° ± 11°, pelvic tilt of 4° ± 8°, and sacral slope of 40° ± 9° [8]. Throughout childhood and adolescence, pelvic incidence has been observed to increase toward the adult value of 52° [9]. Pelvic tilt also increases throughout childhood toward the adult value of 12° [9]. This change in pelvic tilt with age may help to avoid significant displacement of the center of gravity and may be responsible for the reduction in sagittal vertebral axis observed throughout childhood.

Another contributing factor to this age-related reduction in sagittal vertebral axis may be the effect of lumbar lordosis. With upright posture, lumbar lordosis develops and has been found to increase from 44° for children aged 3–9 years to 53° by age 10 years [7]. As lumbar lordosis has a negative effect on the sagittal vertebral axis, this increase in lumbar lordosis throughout childhood may contribute to the age-related changes that have been identified for sagittal vertebral axis [2].

Thoracic kyphosis has also been found to change with patient growth and development.

The published value for thoracic kyphosis in healthy children less than 10 years of age is 38° ± 10° [8], which has been found to increase from 42° in children 3–9 years of age to 48° by age 10 years [7]. Cervical spine alignment in patients aged 10 years old shows significantly more lordosis of 6° versus 1° in those older than 10 years. This may be strongly influenced by craniocervical orientation and thoracic shape [10]. In 181 asymptomatic children with mean age 11.7 years, organized as a function of age, increased cervical lordosis was associated with increased thoracic kyphosis. Subanalysis of those aged 3–7 years have lower rate of cervical hypolordosis and kyphosis than older patients and that cervical lordosis continued to correlate with thoracic kyphosis [11].

When children less than age 10 years were compared with older children, the main areas of sagittal plane alignment which differed were the cervicothoracic region (T1–T2), the thoracolumbar region (T10–L2), and the lower lumbar region (L4–S1) [2]. The hierarchically increasing importance of knowing all of these regional values, the correlation of these values within the region, the global sagittal balance, and their correlation to health-related quality of life have recently been emphasized [7].

In an effort to refine the way that we measure sagittal plane parameters, recent work has been performed which may shed further light on the interrelationships of sagittal alignment. These include the spinosacral angle, spinal tilt, and spinal pelvic tilt, which use as landmarks the first sacral endplate, a horizontal to the center of the first sacral endplate, and the midpoint of a line joining the center of the femoral heads, respectively, as reference for a line connecting to the centroid of the C7 vertebral body. The normative values for these in patients aged 3–10 years are spinosacral angle of 130° ± 10° and spinal tilt of 92° ± 6° [7]. Although angular parameters have become increasingly popular to limit potential measurement error inherent in pure linear descriptors, the true spine length linear measurement tool may improve our understanding of the relationship between spine alignment and spine growth. By measuring the length of a curve formed by regular points along the spine rather than along a straight line, a true representation of the length of the spine will be obtained [12].

23.2 Sagittal Plane Alignment in Growing Children with Scoliosis

A biomechanical study of three-dimensional modeling of growth suggests that perturbations in the thoracic spine in the coronal plane are more important in the development of scoliosis than in the sagittal [13]; however, recent studies in the adolescent population have shown that there are sagittal plane differences between predominantly lumbar and thoracic curves even early in the scoliotic process, supporting the role of the sagittal plane in curve development [14].

Young children with scoliosis have been found to have a positive sagittal balance of 2.2 ± 4 cm, which was not related to the etiology of scoliosis as defined by the classification for early onset scoliosis (C-EOS) diagnosis [4]. Children with scoliosis age 1–10 years were observed to have increased pelvic tilt (11° ± 14°), decreased sacral slope (36° ± 12°), and similar pelvic incidence (48° ± 16°) as compared to young children without scoliosis [4]. This increased pelvic tilt may be important as increased retroversion has been linked to spondylolisthesis in adolescents and young adults as well as to poor health-related quality of life in adults [15]. In addition, there is some evidence that these parameters may be linked to failed pediatric spinal operations [16]. Despite this, there is some evidence in adolescent idiopathic scoliosis that a direct radiographic link between sacropelvic and thoracic parameters is not yet obvious [17]. An MRI study looking at gender-related variation of supine lumbosacral parameters seemed to confirm the need to consider pubertal development stage rather than simply age when studying normal values for males and females [18]. It was found that sagittal plane alignment was similar between congenital, neuromuscular, and idiopathic scoliosis in children younger than 10 years old. Syndromic scoliosis was found to have a higher pelvic tilt and a higher

pelvic incidence than the other etiologies [4]. In that same study, thoracic kyphosis (38° ± 21°) and lumbar lordosis (49° ± 17°) were not found to be different than for normal children of this age [4].

23.3 Conditions That Affect Sagittal Plane Alignment

23.3.1 Postural Kyphosis

Postural kyphosis is a mild form of kyphosis in which no structural changes can be seen. This includes adolescent round back, which can be distinguished from structural kyphosis be the Adam's forward bending test. A postural kyphosis will not create a sharp, angular deformity and will often disappear with this test (Fig. 23.2). The prognosis for postural kyphosis is generally favorable and no surgical treatment is recommended. Physiotherapy may be efficacious to maximize core muscle strength and to improve upon posture.

23.3.2 Scheuermann's Condition

Scheuermann's condition has been regarded as an idiopathic condition, although a genetic predisposition for the condition has also been identified. This disorder typically develops during adolescence and can affect the thoracic, thoracolumbar, or lumbar spine (Fig. 23.3). Thoracic Scheuermann's condition creates a kyphotic deformity, while the latter two show more symptoms such as pain. Radiographic features include endplate irregularity, Schmorl's nodes, and vertebral body wedging. Magnetic resonance imaging (MRI) allows visualization of disk dehydration as well as better delineation of the radiographic features of Scheuermann's.

Pain and cosmetic appearance are the most frequent symptoms that lead to an orthopedic consultation. Lung function is generally not greatly reduced and it is rare to develop neurological symptoms.

In most cases of Scheuermann's condition, there is no need for treatment. Bracing is sometimes recommended; however, the literature on the subject is controversial. The main indications for surgical treatment of Scheuermann's kyphosis are pain and appearance. Surgical methods have classically included an anterior release with posterior spinal fusion and instrumentation. Correction by anterior approach alone using an anterior double rod system has been advocated by some [19], while posterior surgery alone has been proposed by Ponte [20]. With his technique of multiple posterior osteotomies and posterior instrumentation, the spine is shortened and correction is performed. In rigid cases, we perform a thoracoscopic release combined with Ponte's technique. Health-related quality of life, specifically self-image and mental health domains of the Scoliosis Research Society Questionnaire, has been found to significantly increase 2 years postoperative for Scheuermann's kyphosis; however, the surgical treatment of Scheuermann's kyphosis has 3.1 times the likelihood of major complication compared to surgical treatment of adolescent idiopathic scoliosis [21, 22]. Careful counseling of patients is recommended prior to performing surgery for Scheuermann's kyphosis.

23.3.3 Congenital Kyphosis

The most common type of congenital kyphosis is failure of formation but failure of segmentation and mixed types can also be seen [23] (Fig. 23.4). Malformations may result in an isolated sagittal plane deformity even though these are most often combined with a scoliosis. MRI of the spine is recommended, as other intraspinal pathology is prevalent in at least 25 % of congenital spine cases. Renal ultrasound and cardiac echocardiogram evaluations are recommended as concomitant malformations of the genitourinary tract and the heart must be excluded. Close observation with regular reassessments is recommended and, during periods of rapid growth, investigations should be performed even more vigilantly. Upright PA and lateral radiographs should be obtained during these assessments. There is no evidence that brace treatment is able to halt the progression

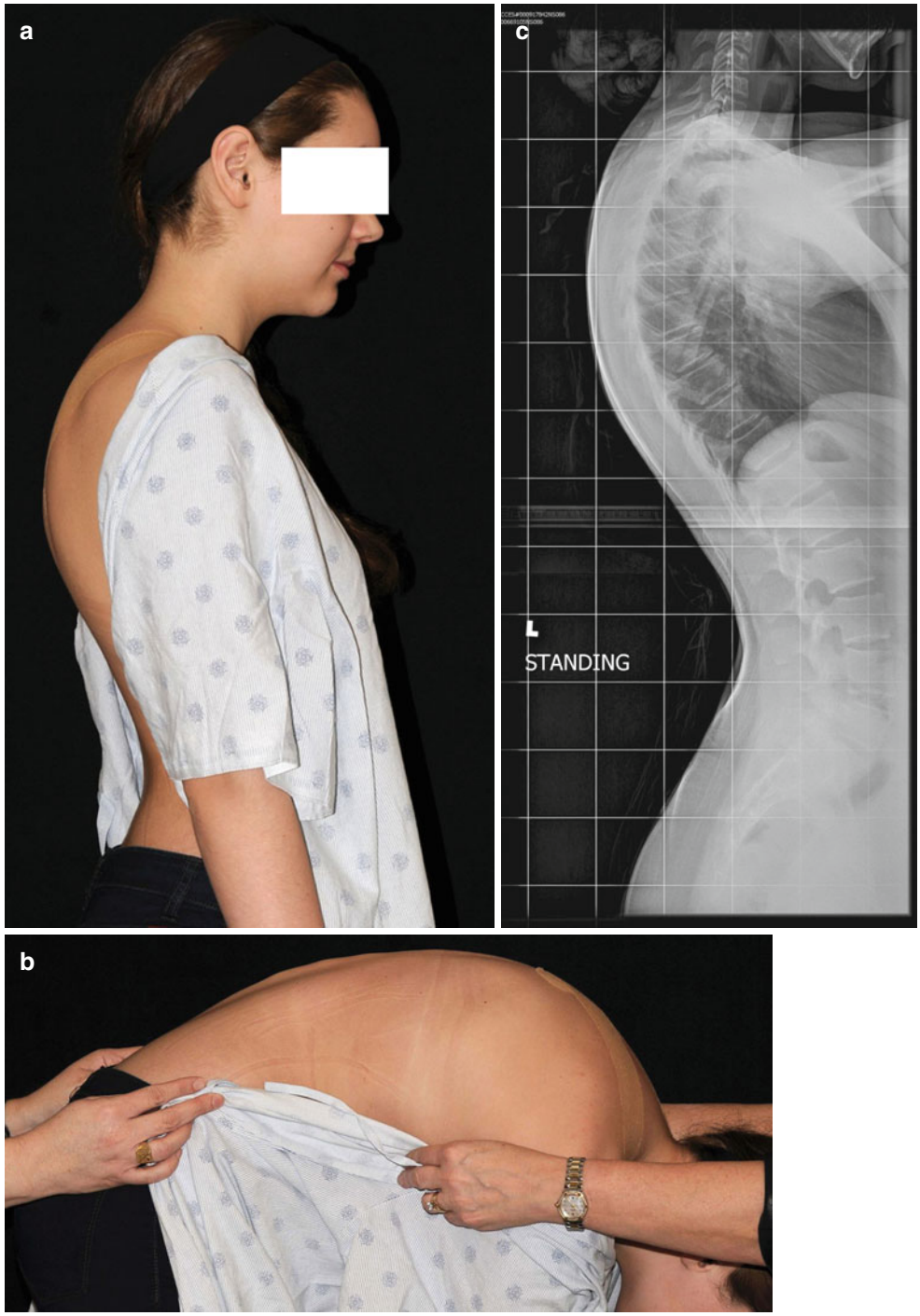
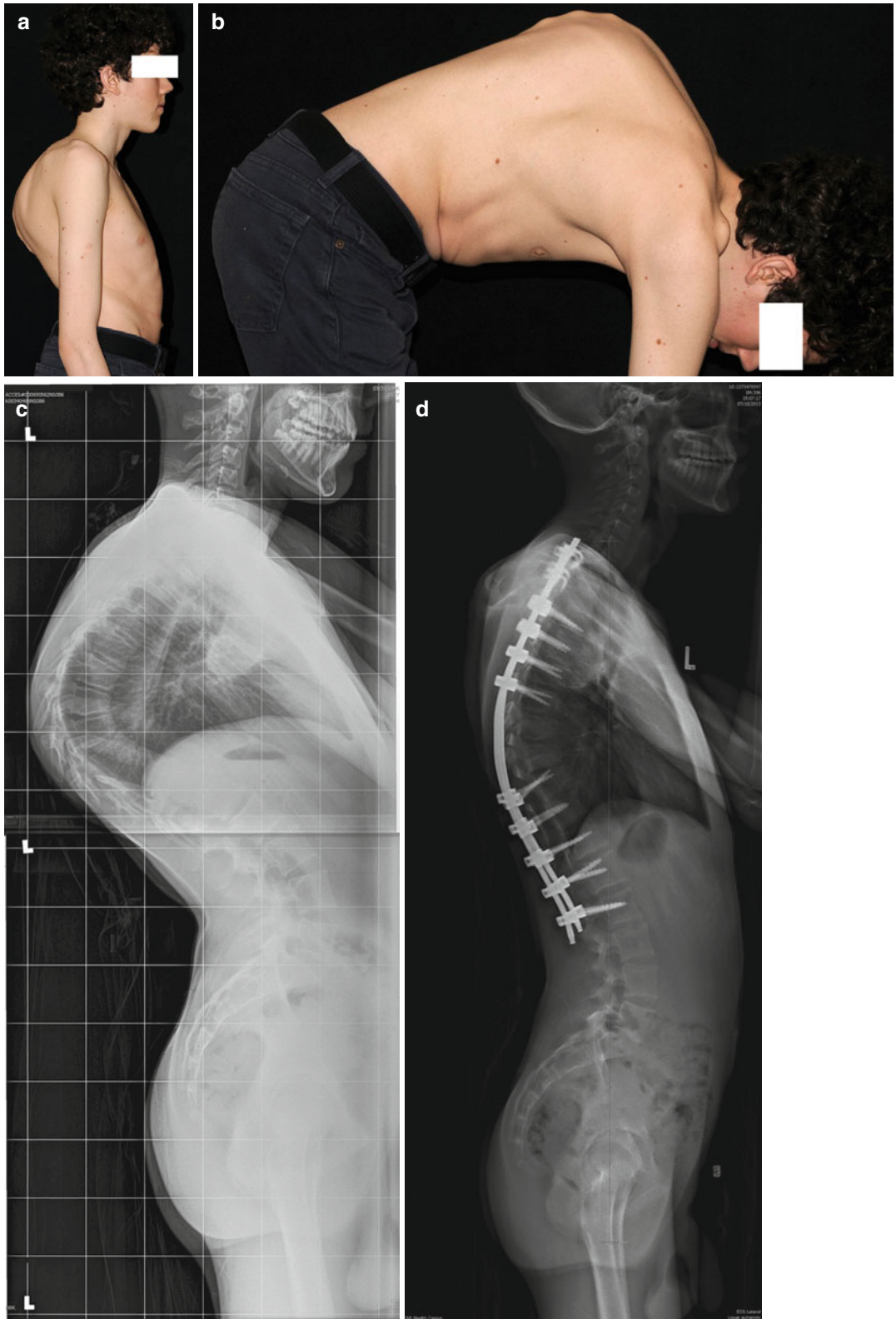


Fig 23.2 A 13-year-old girl with hyperkyphosis. (**a**) Clinical photograph. (**b**) Clinical photograph with forward bending demonstrating loss of hyperkyphosis. (**c**) Standing lateral radiograph demonstrating T5–T12 kyphosis of 45°. Endplate irregularities are present; however, strict criteria for Scheuermann's kyphosis are not present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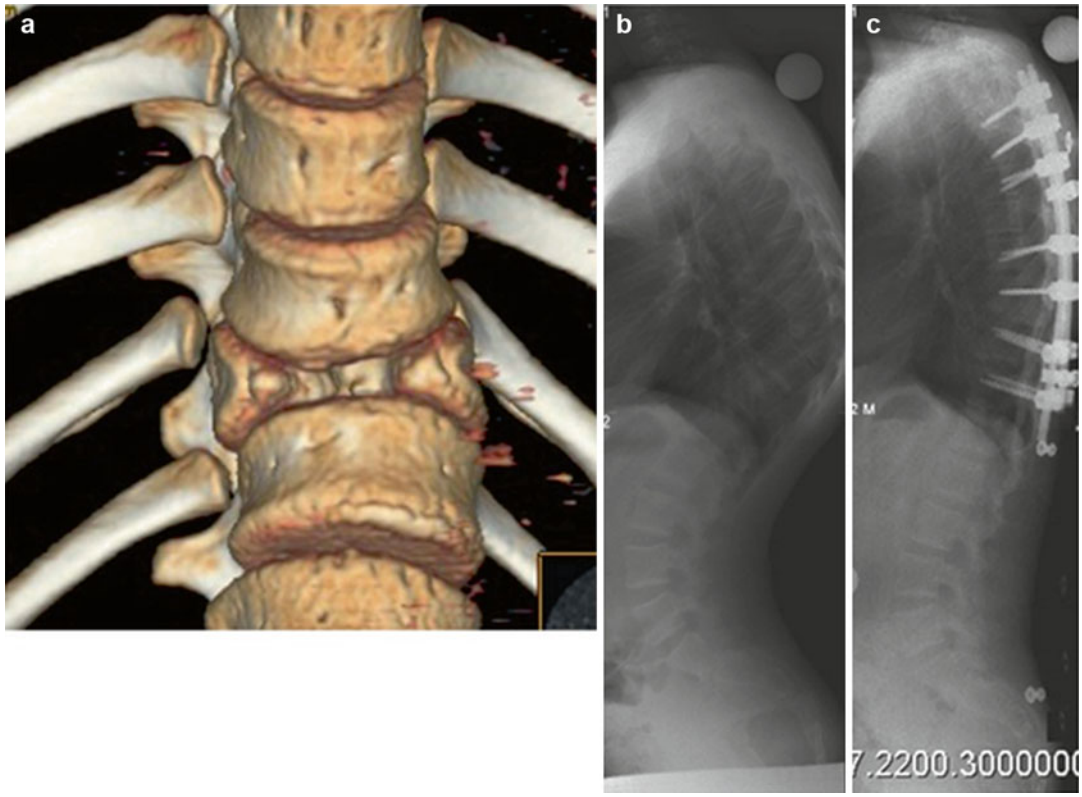


Fig. 23.4 (**a**) A 3D reconstruction of a case with kyphosis due to failure of formation. (**b**) Preoperative lateral view and (**c**) correction by means of pedicle subtraction osteotomy (PSO) and posterior instrumentation

of the kyphotic deformity [24]. Congenital kyphosis due to a failure of segmentation from a progressively ossifying anterior bar is an unusual phenomenon and can be difficult to diagnose in younger children. Thoracic and thoracolumbar bars can lead to mild kyphosis and surgery is rarely indicated. In the event of the lumbar spine with abnormal or ossified disk space, then surgery, including osteotomy, is recommended; whereas, if the disk beyond the bar is normal, bar resection and cement interposition can be utilized [25].

Indications for surgery include a well-documented progression of deformity or for any new neurological symptoms and signs. Historically, noninstrumented posterior fusion and casting were recommended for a kyphosis exceeding 50° in children under the age of 6 years. For older children, a combined anterior and posterior approach was most often recommended. Early surgery is recommended for progressive curves. Anterior approach for the surgical treatment of congenital kyphosis has been a mainstay of treatment and is effective if

Fig 23.3 A 17-year-old boy with Scheuermann's kyphosis. (**a**) Clinical photograph demonstrating a sharp, angular thoracic hyperkyphosis. (**b**) Clinical photograph with forward bending demonstrating persistence of a sharp, angular kyphosis typical of Scheuermann's kyphosis. (**c**) Standing lateral radiograph demonstrating radiographic evidence of Scheuermann's kyphosis (vertebral body wedging and endplate irregularities). His T5–T12 kyphosis measured 95°. (**d**) Standing lateral radiograph postoperative left prone thoracoscopic release T6–T10, Ponte osteotomies T7–T10, and posterior spinal fusion and instrumentation T2–L2

performed early. Posterior-only techniques are technically challenging but are becoming more popular [26]. These contemporary and improved surgical techniques, such as pedicle subtraction osteotomy (PSO), the "eggshell procedure," and posterior vertebral column resection (PVCR), have now limited the requirement for anterior surgery.

Perhaps the most dramatic of congenital deformities of the growing spine is the rare congenital dislocation, a single-level developmental failure of the spine and spinal cord at a single spinal level. Any initial neurological involvement is thought to be associated with cord malformation rather than mechanical factors [27].

23.3.4 Myelodysplasia

In myelodysplasia, there is greater than 10 % rate of kyphosis. This deformity is usually located in the upper or middle part of the lumbar spine (Fig. 23.5). In many cases, the deformity in myelodysplasia is accompanied by skin and soft tissue problem (ulceration) due to a lack of sensitivity, poor blood flow, and thin and stretched subcutaneous tissue. Three types of kyphosis in myelodysplasia have been described: paralytic, sharp-angled, and congenital [28]. The paralytical type has an almost normal curvature at birth. The sharp-angled type has a rigid curvature at birth due to the pathological position of the erector spinae, quadratus lumborum, and the thoracolumbar fascia anterior to the spinal column. This pathological anterior position creates a flexion moment through normally lordotic segment of spine. The congenital type is the most severe spinal deformity in myelodysplasia as there is also an anterior defect of segmentation, which allows this deformity to progress rapidly during the first year of life.

In general, conservative treatment has not been successful; however, bracing can theoretically reduce the rate of progression, with skin and respiratory problems as probable or expected side effects. Many different surgical techniques have been described (see Chap. 34). The absence of posterior elements, poor skin, a large thecal sac, and a concomitant osteoporosis make this surgery very challenging [29]. Preoperatively, tissue expansion by means of soft tissue expanders can be applied; although, with the shortening of the spine during correction, the "tissue gain" is, in most cases, good enough to provide the surgeon with enough skin for coverage. Close cooperation with an experienced plastic surgeon is most helpful. Preoperatively, a computed tomography (CT) of the brain should be obtained so that any hydrocephalic expansion could be diagnosed if neurological changes occur postoperatively.

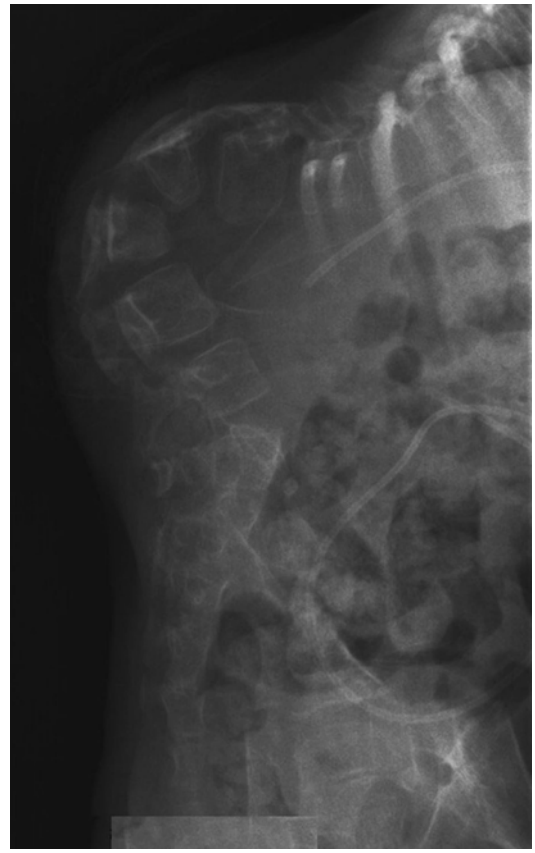


Fig. 23.5 Congenital kyphosis in myelodysplasia, lateral view

As early as 1968, Sharrard [30] described kyphectomy in the neonatal period, by having no other stabilizing techniques than sutures. Crawford et al. [31] reported a series of kyphectomies, utilizing a sophisticated technique with sutures, at the time of first intervention. The

operation is most often performed at the age of about 6 years. Previously, division of the thecal sac was recommended; however, with modern techniques such as PSO or PVCR, the thecal sac can be left intact. The distal fixation can be performed with various techniques, but is challenging because of poor quality of soft tissue, insensitivity, poor vascular supply, and the absence of posterior bone. Proximally, instrumentation to the upper thoracic spine is recommended in order to minimize adjacent segment kyphosis. Kyphectomy in myelomeningocele carries a large range of serious complications up to and including death [32, 33]. Posterior kyphectomy with anterior fixation and cordotomy has been performed in myelomeningocele with similar results to other procedures however with less blood loss [34]. As well, in an effort to preserve growth and pulmonary function, growth-friendly surgery may be considered for the upper part of the instrumentation [35]. Growth-friendly surgery may be instituted without kyphectomy, even in severe gibbus deformity, whereas the sagittal effect when used primarily for scoliosis correction requires further attention [36, 37].

23.3.5 Postinfectious Kyphosis

Historically, spinal tuberculosis (TB) has been a great problem worldwide, being the main cause of kyphosis in the developing world (Fig. 23.6). In 2001, Rajasekaran [38] described the natural history of pediatric spinal TB, showing that 15 % of cases treated conservatively still had a considerable increase in kyphotic deformity. He demonstrated that there is a spontaneous remodeling capacity of partially destroyed vertebrae in children under the age of 15 years. He also reported that a child having more than two of the four severe radiographic signs – dislocation of facets, retropulsion of vertebral fragments into the canal, lateral translation, and "toppling" of the superior vertebra – was at significant risk of severe kyphosis development and thus requires surgery. A review of operative Pott's cases in India showed a majority present in their first decade of life and the most common region in the operative pediatric patient is thoracic (33 %), followed by craniocervical junction (27 %) [39]. Thoracolumbar TB has the worst prognosis with regards to the development of significant kyphosis.

The primary basic treatment is chemotherapy with a combination of streptomycin, isoniazid, and rifampicin. The patients should also be braced for at least 6 months. Surgical therapy, such as irrigation and debridement, is indicated for significant paraspinal abscesses. Other surgical indications include neurological signs, progression of kyphosis, or significant bony destruction [38]. In the Western world, these patients often present after the period of active disease. Surgery is often considered in cases with sharp kyphosis exceeding 30°–40°.

23.3.6 Posttraumatic Kyphosis

Young children are not often exposed to high-energy trauma, although minor trauma is not infrequent. Spine fractures account for 1–3 % of all childhood fractures [40]. In contrast to adolescents and adults, fractures in the growing child occur more often in the mid-thoracic region. Due to the elastic nature of children's spines, the trauma force will be transmitted over several segments and result in multiple but less severe fractures [41]. There is a great remodeling capacity of vertebral fractures during growth and, as a result, there is often no residual deformity remaining at skeletal maturity [42]. In the rare case of significant deformity in the immature child, surgical correction and stabilization are indicated. Vander Have et al. reviewed 37 young patients with thoracolumbar burst fractures, of which two subjects were less than 10 years old. One subject was treated with a thoracolumbosacral orthosis (TLSO) and the other with posterior spinal fusion and instrumentation [43]. A multicenter review of 35 patients, with average age 9 years old (range 1.6–17 years), who had Chance fractures, revealed an initial kyphotic deformity that averaged 11° in the nonoperative group and 22° in the operative group [44]. Surgery can often be posterior only and instrumentation generally extends one to two segments cephalad and one to

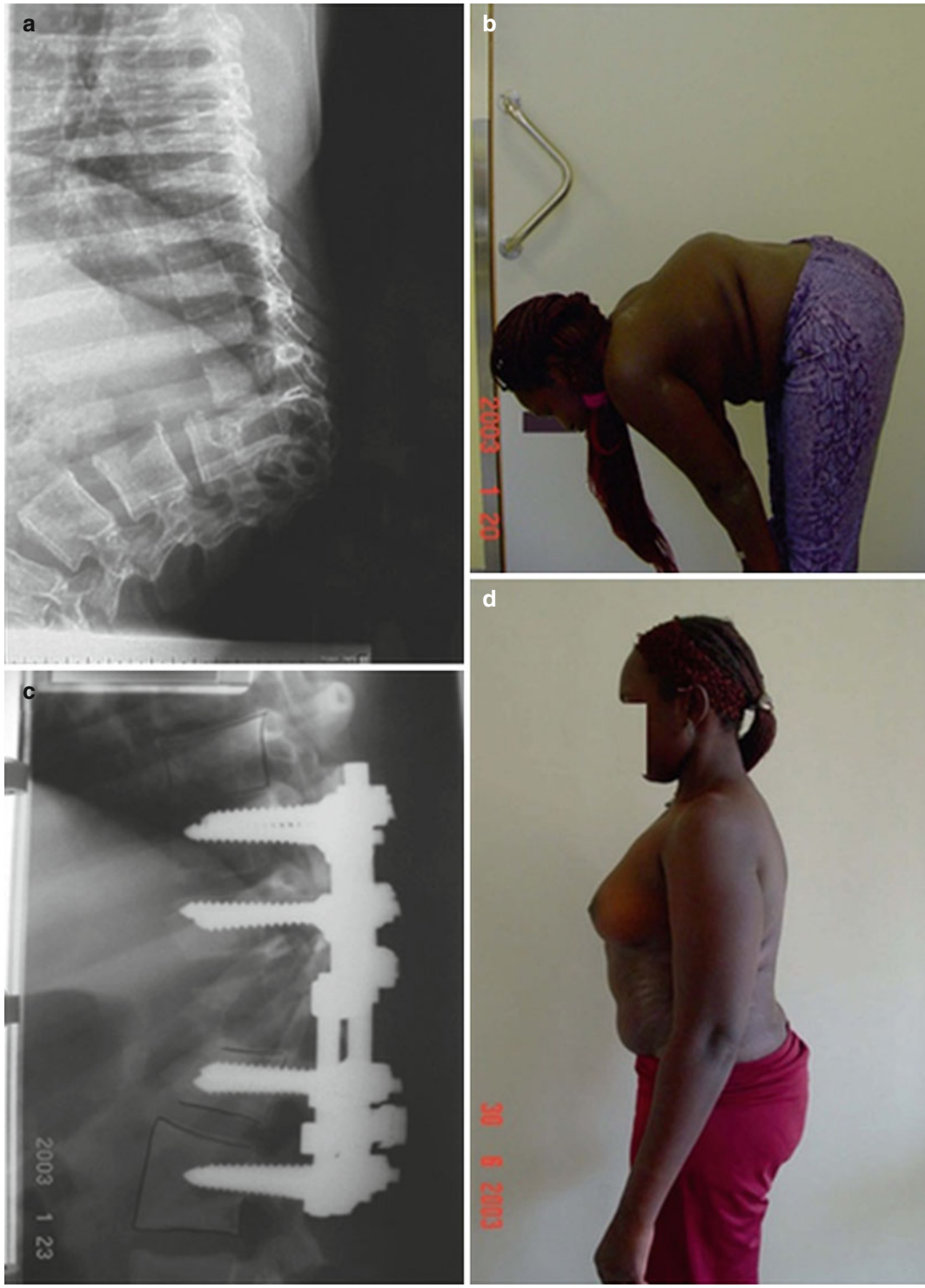


Fig. 23.6 (**a**) Post-tubercular kyphosis. Lateral radiograph. (**b**) The sagittal profile of the patient. (**c**) After operation with PVCR, lateral radiograph. (**d**) The sagittal profile of the patient 1 year after surgery

two segments caudal to the injury (Fig. 23.7). In the face of a complete, permanent neurological deficit at the time of injury, a secondary scoliosis can develop over time.

23.3.7 Syndromic Kyphosis

23.3.7.1 Achondroplasia

Thoracolumbar kyphosis is very common in infants with achondroplasia. Persistent kyphosis is often observed in patients with achondroplasia who are unable to independently ambulate prior to age 2 years. Spinopelvic parameters in children with achondroplasia reveal a dichotomous distribution of pelvic morphology, with one group exhibiting an extremely horizontal sacrum and a negative pelvic tilt. The clinical implications of this finding are not yet understood [45] (Fig. 23.8).

23.3.7.2 Hurler's Syndrome

Thoracolumbar kyphosis in treated Hurler's syndrome averages 38° at 17 months of age. Those patients presenting with greater than 45° of kyphosis are more likely to progress and can be treated with growth-friendly surgery or with anterior or anterior/posterior fusion (Fig. 23.9) [46].

23.3.7.3 Larsen's Syndrome

Thoracolumbar and cervical kyphosis both can occur with Larsen's syndrome. Screening for cervical kyphosis in patients with Larsen's syndrome should be considered due to the possibility of paralysis or death secondary to cord impingement at the apex of the deformity. These deformities are usually progressive and often require surgical stabilization [47].

23.3.8 Postsurgical Kyphosis

23.3.8.1 Postlaminectomy Kyphosis

In all ages, laminectomy alone in a kyphotic region or the unstable zone (T10–L2) of the spine, without any stabilization and fusion, will inevitably bring the patient to be at risk of developing kyphosis. In the pediatric population, the risk of developing deformity is much higher during periods of rapid growth. Despite this knowledge of post-laminectomy deformity, we still observe patients having had laminectomies performed for treatment of spinal cord tumors (Fig. 23.10). As these patients can present years later with large kyphotic deformities, a close relationship with the neurosurgical team at the time

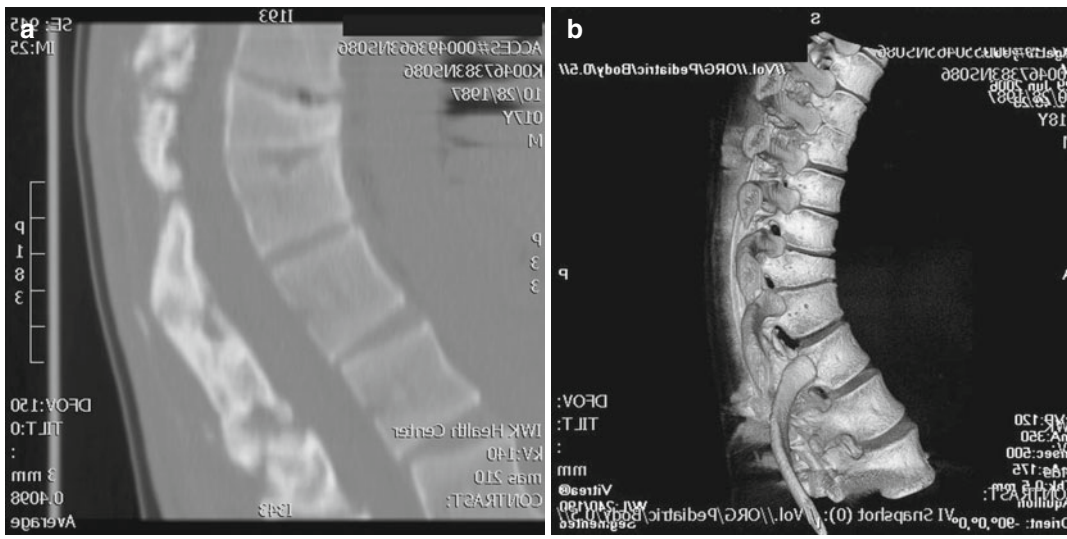


Fig. 23.7 (**a**) Sagittal CT scan image of a 17-year-old boy who was 2 years postinjury with resultant thoracic hyperkyphosis. (**b**) Three-dimensional CT scan image 1-year postoperative osteotomy and posterior spinal fusion and instrumentation

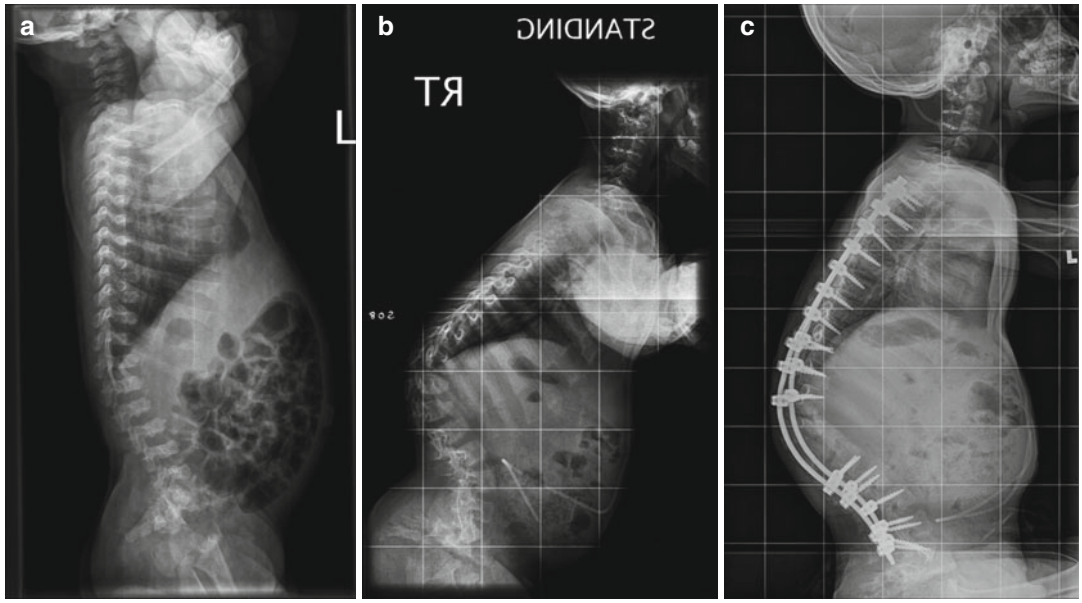


Fig. 23.8 Lateral radiographs of a patient with achondroplasia (**a**) 10 weeks of age with typical anterior vertebral body dysplasia at the thoracolumbar junction; (**b**) 5 years of age with progressive thoracolumbar kyphosis and sagittal imbalance; (**c**) 7 years of age and 2 years postoperative anterior spinal fusion with rib strut graft and posterior spinal fusion and instrumentation with apical Ponte osteotomies

of tumor resection is of paramount importance in order to prevent this deformity. Once a deformity develops, it can be challenging to treat as the tissues have often been irradiated and the patients may have been immunosuppressed as part of their cancer treatment.

23.4 Effects of Growth-Friendly Surgery on Sagittal Plane Alignment

Growth-friendly surgery includes distraction-based surgery such as spine and rib-based growing rods, compression-based surgery such as vertebral body stapling, and guided growth surgery such as Shilla and Trolley techniques. As these techniques are placed in the immature spine in an effort to control coronal plane deformity, surgeons have also observed postoperative effects on the sagittal plane.

Sagittal balance is initially improved after implantation of spinal growing rods, although sagittal balance increases again with subsequent lengthening surgeries [48]. An analysis of 23 growing rod patients with minimum 2-year follow-up demonstrated changes in kyphosis from preoperative, postoperative to final follow-up of 50°, 35–45°. Lordosis (45°, 42°, 48°) and sagittal balance (3.72, 2.33, 3.92 cm) also changed slightly during those intervals. It is now recognized that posterior distraction techniques are kyphogenic by nature. Patients with spinal growing rods do not appear to have changes in cervicothoracic parameters and may have improvements in their spinopelvic parameters [49].

A review of 14 largely non-idiopathic kyphoscoliotic curves that were treated with rib-based distraction surgery demonstrated an increase in thoracic kyphosis by a mean of 22°. As a result of this ubiquitous hyperkyphosis, a frequent problem seen in this cohort was migration of the proximal rib hooks. The authors recommended adding more fixation points, placing rib anchors more cephalad, placing distal foundations more caudal, and utilizing more hybrid rib to spine constructs [50].

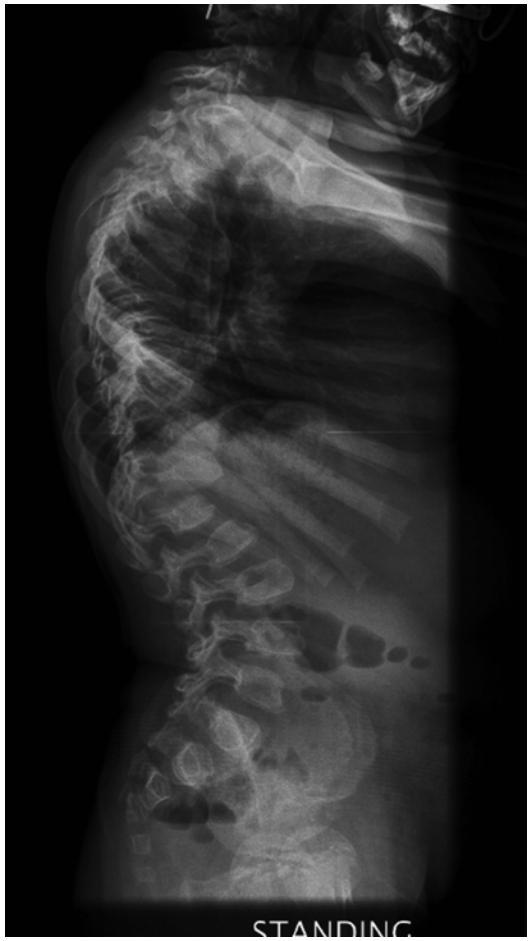


Fig. 23.9 Standing lateral radiograph of a 5-year-old girl with kyphosis secondary to Hurler's syndrome demonstrating dysplastic vertebral bodies with resultant thoracolumbar junction kyphosis

In 28 patients with idiopathic scoliosis, in whom approximately half were juvenile, anterior vertebral body stapling appeared to modulate growth in both the thoracic and lumbar spine toward increasing kyphosis or decreasing lordosis. After stapling, there were more patients with normal thoracic kyphosis than hypokyphosis, and the average lumbar lordosis was 49° with all patients having >10° change in lordosis. Vertebral body stapling is contraindicated in patients with preexisting thoracic hyper kyphosis [51].

There is some early evidence to suggest that guided growth and nonoperative distraction-based techniques (i.e., magnetically controlled growth rods) may affect sagittal alignment in children with early onset scoliosis. The Shilla growth guidance concept is based on a three-plane correction with apical fusion using dual rods. These rods are connected to proximal and distal gliding anchors to allow for continuing spinal growth. An early report of ten patients, at 2-year follow-up, revealed no significant change in thoracic kyphosis [52]. In a study examining 14 patients with the magnetically controlled MAGEC implant (Ellipse Technology, Irvine, CA), the mean thoracic kyphosis was 39° preoperatively, 31° immediately postoperatively, and 48° at latest follow-up [53].

Hyperkyphosis secondary to growth-friendly surgery can increase the rate of postoperative complications. In a study utilizing the Classification for Early Onset Scoliosis system, it was observed that patients who were hyperkyphotic were at a higher risk for device-related complications that required a trip to the operating room [54]. A further study on the effect of thoracic kyphosis on spinal growing rod surgery showed that hyperkyphosis above 40° was associated with implant complications such as rod breakage. The complications were observed to increase linearly with increasing kyphosis and that syndromic patients were at an ever higher risk [55].

The complication of proximal junctional kyphosis (PJK) has been observed during growth-friendly surgery (Fig. 23.11). For spinal growing rods, rates of PJK as high as 56 % have been discussed with 44 % of these having upper anchor failure. Some of these patients had upper rib-based anchors, which were thought to be protective against PJK. Rates of PJK were also twice as common with dual growing rods, which, may be secondary to the increased rigidity of the dual versus single rod, construct [56]. In a large study by Bess et al., PJK developed in only 3 of 140 growing rod patients [57]; however, in a multicenter database review of 88 patients, PJK occurred in 24 % with independent risk factors being greater thoracic kyphosis, greater proximal thoracic scoliosis, more proximal level of lowest instrumented vertebrae, and proximal anchors other than pedicle screws [58]. Proximal junctional

kyphosis secondary to rib-based surgery has been found to be associated with preoperative hyperkyphosis and possibly to an upper-instrumented level caudal to T6 [59, 60]. A study of rib-based distraction in 21 patients showed that clinically significant proximal kyphosis was observed in four patients and positive sagittal off-balance in five patients [61]. In a head to head study of rib-based and spine-based distraction surgeries, a similar rate of PJK was observed (25 % vs. 31 %); however, spinal growing rods may afford a better initial sagittal plane correction [62, 63]. In a small series of ten patients treated with Shilla growth guidance, there were no cases of

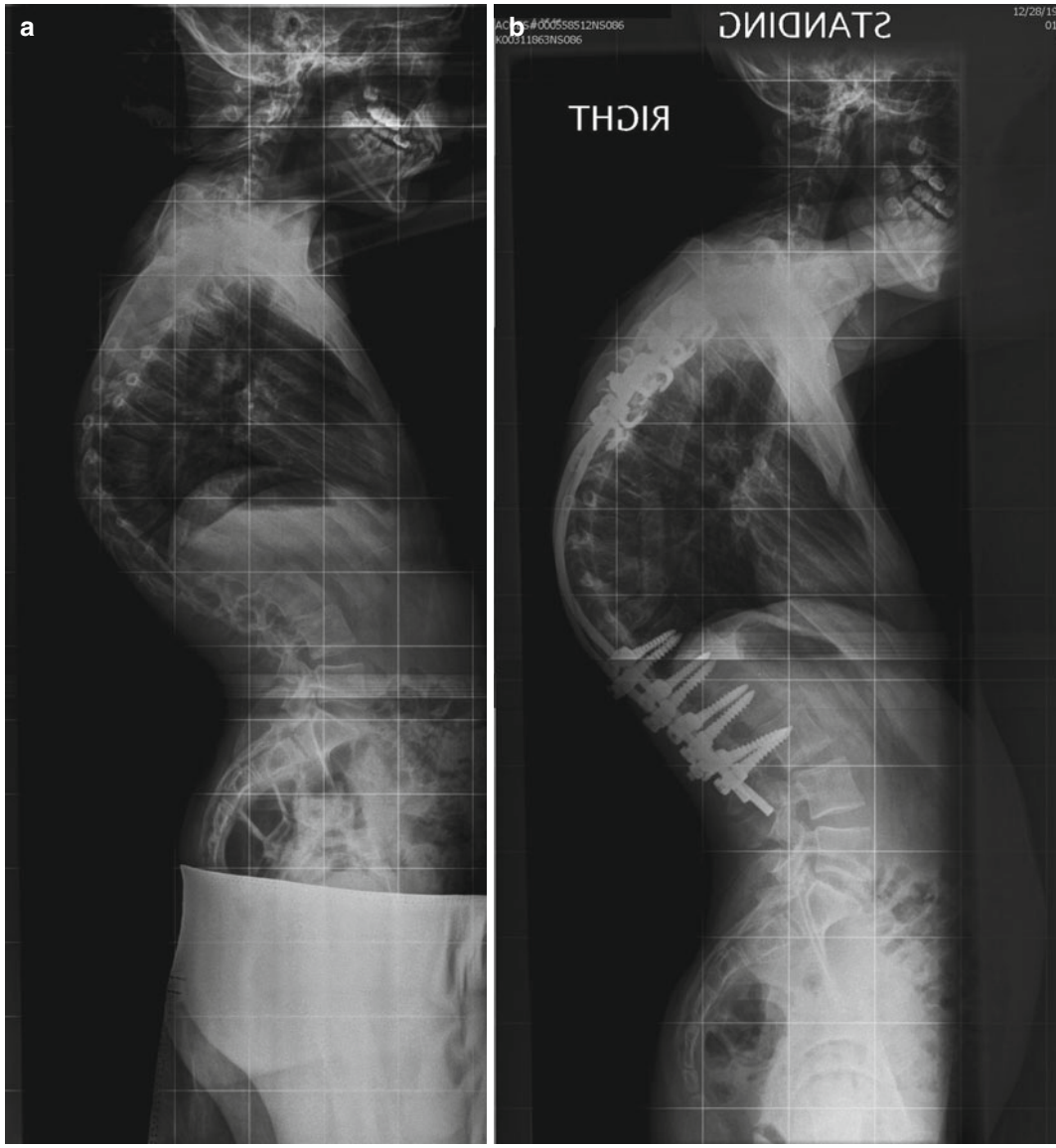
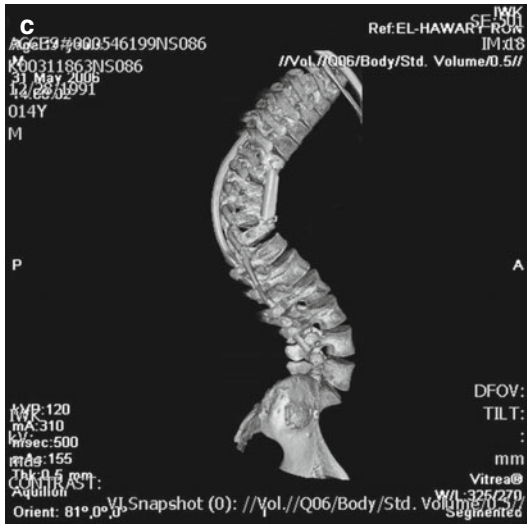


Fig. 23.10 A 13-year-old boy who was treated as a toddler with laminectomy and radiation for a Ewing's sarcoma. (**a**) Standing lateral radiograph demonstrating 90° of thoracic kyphosis. (**b**, **c**) Standing lateral radiograph and three-dimensional CT image 2-year postoperative right thoracotomy, discectomy T8–T10, in-situ fusion with vascularized fibular graft, and posterior spinal fusion and instrumentation T1–L3 with iliac crest bone graft



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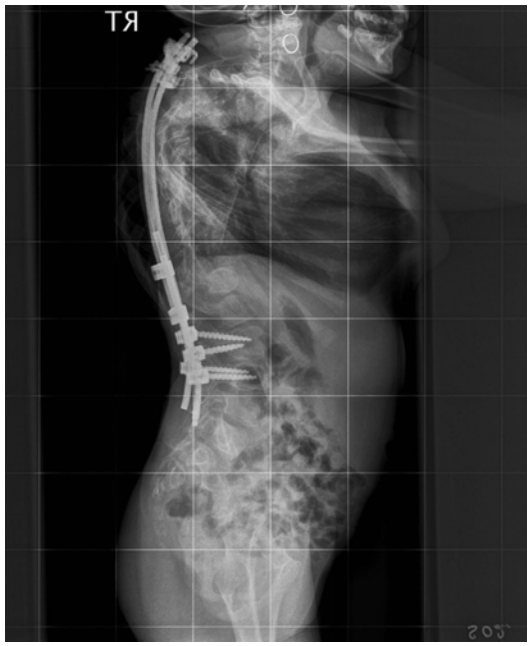

Fig. 23.11 Standing lateral radiograph of a 9-year-old girl with Hurler's syndrome who was treated with growth-friendly surgery for thoracolumbar kyphosis (same patient as illustrated in Fig. 23.9). Image demonstrates proximal junctional kyphosis with subsequent upper instrumented vertebral failure

PJK at short-term follow-up [50]. One study on the MAGEC implant (Ellipse Technology, Irvine, CA) observed no cases of PJK in a group of 14 subjects [53], while in another study one case of proximal junctional kyphosis was identified with the use of this device [64]. Current investigations into the high variability of radiographic measurement variability and the clinical importance of radiographic PJK are ongoing [65, 66].

23.5 Neuromuscular Hyperlordosis

Neuromuscular hyperlordosis without scoliosis has been described, but it is extremely rare (Fig. 23.12). Brace treatment is of limited value and surgical indications are not clearly defined. Surgery generally consists of anterior lumbar discectomy and/or posterior paraspinal muscle release. Halo femoral traction is a useful intraoperative adjunct to improve correction of the hyperlordosis. In case of an existing posterior nonsegmented bar, Langenskiöld has described resection of the bar with deformity correction and posterior instrumentation. In young children, spinal or rib-based growing rods can be utilized to correct the deformity until skeletal maturity.

23.6 Summary

Physiologic sagittal plane alignment in healthy children evolves with age as pelvic incidence, pelvic tilt, thoracic kyphosis, and lumbar lordosis all increasing during childhood and reach typical adult values by the adolescent years. Sagittal plane alignment in children with scoliosis demonstrates increased pelvic tilt and decreased sacral slope as compared to children without scoliosis; however, pelvic incidence, thoracic kyphosis, and lumbar lordosis are similar. A variety of etiologies including postural kyphosis, Scheuermann's condition, congenital kyphosis, myelodysplasia, postinfectious, posttraumatic, syndromic, and postsurgical can result in alterations in the sagittal plane alignment of growing children. Each of these etiologies is unique and treatment of each of these conditions is individualized. An evolving understanding of sagittal

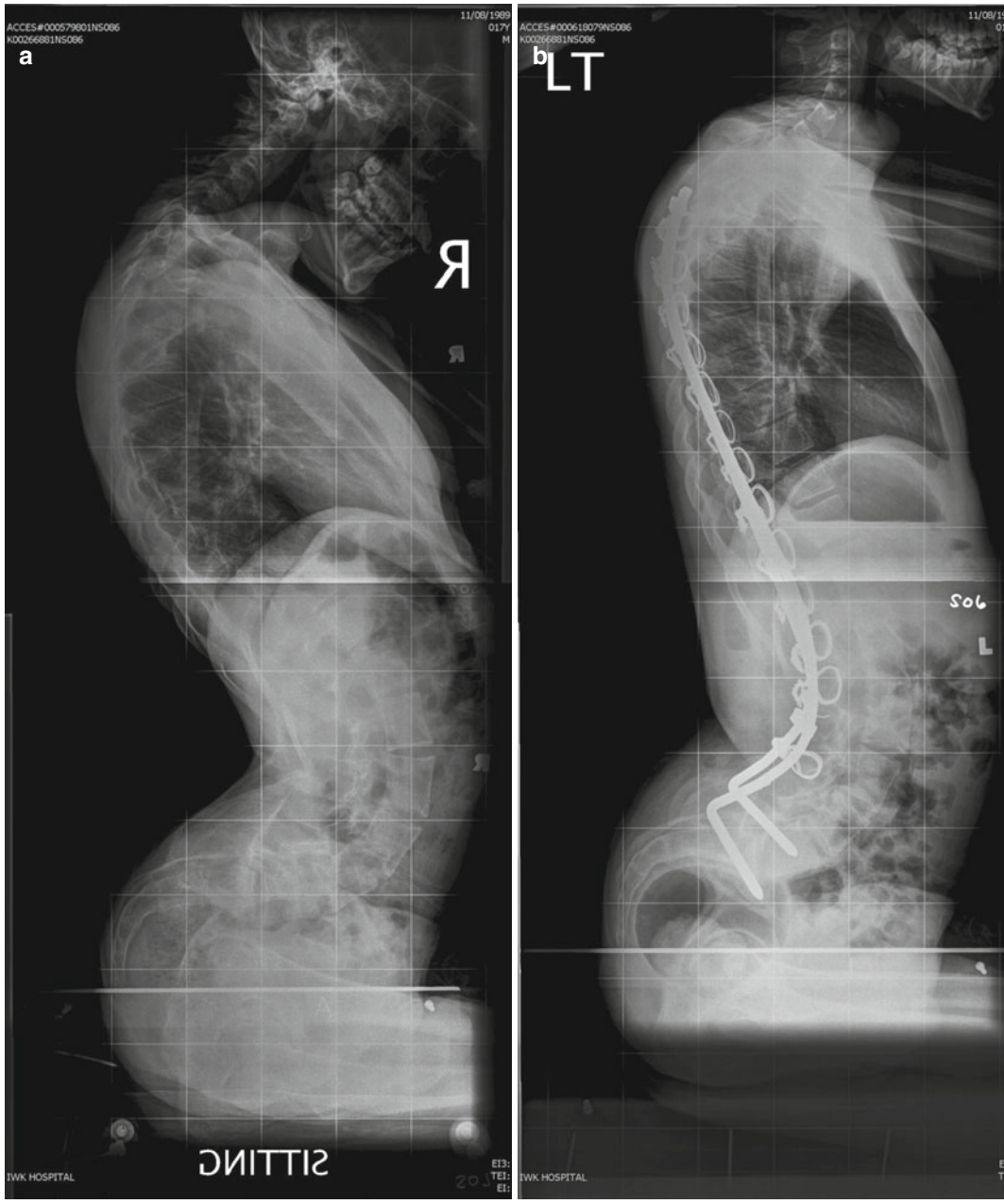

Fig. 23.12 (**a**) A 17-year-old boy with cerebral palsy and lumbar hyperlordosis. Note the significantly increased sacral slope and decreased pelvic retroversion. (**b**) Two years postoperative posterior spinal fusion and instrumentation with Luque–Galveston technique

plane alignment and of the measurement of spine growth is improving our knowledge of the effects of surgery on sagittal plane alignment in children.

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Spondylolisthesis

24

Dietrich Schlenzka

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Key Points

- Isthmic lumbar spondylolisthesis occurs in 4.4 % of children and about 6 % in the adult Caucasian population. In general, it is a benign condition.
- Children before or during the growth spurt need radiographic follow-up for documentation of possible progression.
- Uninstrumented posterolateral in situ fusion is the treatment of choice for pain not responding to conservative measures in slips up to 50 %.
- In severe slips (>50 %), anterior or combined fusion is the most reliable way to prevent further progression of lumbosacral kyphosis.
- Slip reduction in high-grade slips is controversial. It is accompanied by a higher risk of complications. It has not been shown to be superior to in situ fusion.
- Spondylolisthesis in syndromes has various aspects. It should be assessed and treated according to basic principles considering the underlying condition. Long-term follow-up is mandatory in these rare cases.

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24.1 Background

Spondylolisthesis in growing Individuals – at first glance – appears to be a straightforward topic. However, it proves to be difficult when limited to children only, i.e. to the age group up to 12 years. Traditionally, the majority of publications dealing with this diagnosis combine children and adolescents as a single group. In series with a substantial number of young patients, children are represented only as a small minority, the majority being adolescents or young adults [29, 58, 60, 62, 97, 118, 120–123, 134]. There are a few publications reporting on infants or "very young" children [5, 11, 52, 55, 60, 61, 79, 98, 140, 144, 151]. To the best of the author's knowledge, there is only one study presenting a significant number of patients under the age of 12 years [120].

According to the present knowledge, there should not be a significant difference in the incidence of the condition between pre-adolescence and adolescence. The explanation for the fact that children are underrepresented in postoperative follow-up series may be that there is a higher probability of developing a further slippage and becoming symptomatic during adolescence than during childhood. It is impossible to say whether this has to do with the natural history (e.g. influence of the growth spurt), with increasing physical activities during adolescence, or with both of them.

It must be remembered that the information concerning children presented in this chapter is fragmentary due to the limited data in the literature. The information we do have does not give any reason to assume that there should be significant differences in the approach to spondylolisthesis in children as compared to adolescents. Patients' outcomes after fusion surgery do not seem be different in children if compared to adolescents [47].

Despite that, given recommendations should be applied critically and with great reluctance, especially when dealing with very young patients.

24.1.1 Epidemiology

Spondylolisthesis affects only humans. It seems to be extremely rare in non-ambulatory persons [18, 74, 107]. Spondylolisthesis has never been described in animals except in experimental models [94, 111]. Lumbar spondylolysis affects the fifth lumbar vertebra in 90 %, the fourth in 5 % and the third in 3 % of cases. The risk to developing symptoms during adulthood is higher if the changes are located in the segments above L5 [38, 112].

It has never been found in newborn. The youngest patient with spondylolisthesis reported to date was 15 weeks old [11]. Laurent and Einola published a case of unilateral spondylolysis with 4-mm slip in a 10-month-old girl [61]. In Caucasians the prevalence of spondylolysis is 4.4–5 % at early school age [1, 17, 27]. It increases during growth being 6.0–7.2 % in adult Caucasians [29, 85, 141].

In certain ethnic groups the prevalence is much higher (Alaskan Inuit: 32.9 %, Ainnos in Japan: 41 %) [82, 132]. Isthmic spondylolisthesis is more common in males but severe slips occur more frequently in females.

24.1.2 Classification

Traditionally, spondylolisthesis is classified according to Wiltse, Newman and Macnab [149] (Table 24.1).

According to their classification, the majority of slips belongs to the *isthmic* type in which an interruption (spondylolysis) or elongation of the pars interarticularis (isthmus) of the vertebral arch is present. The *dysplastic* spondylolisthesis develops due to congenital changes of the upper part of the sacrum and the vertebral arch of L5. Subluxation of the facet joints is always present in this form. True dysplastic spondylolisthesis is rare. Further types are *traumatic* spondylolisthesis

Table 24.1 Classification of spondylolisthesis

I. Dysplastic
II. Isthmic
A. Spondylolysis
B. Isthmus elongation
C. Acute fracture
III. Traumatic
IV. Degenerative
V. Pathologic
VI. Iatrogenic

According to Wiltse, Newman and Macnab [147, 148]

in acute fractures, *degenerative* spondylolisthesis as a result of disc and facet joint degeneration in elderly people, and *pathologic* spondylolisthesis caused by infection or tumour destruction of parts of the vertebral arch [90, 149]. *Iatrogenic* spondylolisthesis may occur after excessive resection of posterior vertebral elements [149]. This classification has been criticised rightly for being inconsistent and mixing aetiologic (e.g. dysplastic) and anatomic (e.g. isthmic) terms. As its inventors already realised, the distinction between isthmic and dysplastic forms is not always possible. And no specific treatment guidelines are derived. To overcome these shortcomings recently improved classification systems have been proposed [71, 73].

The Marchetti-Bartolozzi classification (Table 24.2) has gained much popularity especially in North America [74]. It breaks spondylolisthesis down into two main aetiologic groups: *developmental* and *acquired*. For all developmental forms, the authors assume a more or less severe congenital dysplasia (i.e. weakness) in the posterior elements ("bony hook") of the vertebra leading with time under physiologic loads to spondylolysis and/or spondylolisthesis. The developmental form is subdivided into *high dysplastic* and *low dysplastic*, each with spondylolysis or elongation of the pars. The acquired forms of spondylolisthesis are *traumatic*, *post-surgery*, *pathologic* and *degenerative* with their respective subgroups.

The classification proposed by Mac-Thiong and Labelle has its roots in the Marchetti-Bartolozzi classification. It was refined by adding criteria concerning the sagittal spino-pelvic balance and recommendations for operative treatment based on current practice [71].

Table 24.2 Classification of spondylolisthesis

Developmental	Acquired
High dysplastic	Traumatic
With lysis	Acute fracture
With elongation	Stress fracture
Low dysplastic	Post-surgery
With lysis	Direct surgery
With elongation	Indirect surgery
	Pathologic
	Local pathology
	Systemic pathology
	Degenerative
	Primary
	Secondary

According to Marchetti and Bartolozzi [72]

A drawback of these newer classifications is that they are rather complex in view of daily clinical use. The distinction between different types is partly arbitrary and not always clear-cut. There are still "grey zones" [73], and the real benefit for clinical decision making is not obvious. The patient's age, a very important factor, is neglected. And the treatment recommendations given for the different types of spondylolisthesis have not been verified yet prospectively in a sufficient number of patients.

Another new classification, designed especially for children and adolescents, was proposed by Herman and Pizzutillo [42]. It includes pre-spondylolytic stress reactions of the isthmus seen in single-photon emission computed tomography (SPECT) and in magnetic resonance imaging (MRI). It focuses on non-operative treatment. According to the inventors, its validation concerning treatment recommendations will take several years.

The degree of anterior translation as described by Meyerding is a commonly used classification system to describe the deformity and to assess for progression [80]. This classification divides the lower vertebral body into four parts to describe the percentage of slippage: grade I is <25 %, grade II is 26–50 %, grade III 51–75 % and grade IV 76–100 %. Grade V >100 % which is often used for spondyloptosis was not mentioned in Meyerding's original publication.

In the author's experience, for practical decision making at present, the essential factors are the degree of slip, the sagittal alignment (lordosis/kyphosis) at the level of the slip, patient's age and symptoms. In this context, it is of secondary interest whether a slip is to classify, e.g. as dysplastic or not.

24.1.3 Natural History and Risk of Progression

The natural history of isthmic spondylolisthesis is benign in the majority of cases due to a tendency towards self-stabilisation of the affected

segment [119, 141]. Despite that, isthmic spondylolisthesis is the most important cause of low-back pain and radiating leg pain in children and adolescents [62]. The average prognosis of the adult individual with isthmic spondylolisthesis concerning low-back problems and working ability does not differ from the rest of the population [7, 28, 38, 141]. There is no explanation yet why some people with spondylolysis or isthmic spondylolisthesis become symptomatic while the majority remains symptom free. As sources of pain the lytic defect itself, the intervertebral disc, the nerve roots and the ligaments are all possibilities [13, 83, 92, 114, 115, 145].

Spondylolysis may be present without vertebral slip. If the slipping occurs it happens mainly during the growth period and is usually mild [7, 123]. Participation in competitive sports does not seem to increase the risk for progression [87]. Risk factors for progression in young individuals are high degree of slip (>20 %) at admission and age before growth spurt [44, 123]. The trapezoid shape of the slipped vertebral body and rounding of the upper endplate of the sacrum in more severe slips are frequently interpreted as "dysplastic" changes and/or predictors of progression. They are, however, in most cases secondary changes. They express the severe slip; they do not predict it [7, 12, 27, 43, 61, 99, 123].

24.2 Clinical Presentation

24.2.1 Symptoms

Pre-school children are usually pain-free. In this young age group, the condition is mostly detected by chance or due to posture changes and/or gait abnormalities (see Sect. 24.4.4). In older children, the unset of the symptoms is often spontaneous. A history of sports activities is very common. Sometimes acute trauma is reported.

The leading symptom is low-back pain during physical activities as well as while standing and/or sitting for a longer period of time. The pain may radiate to the buttocks and to the posterior or lateral aspect of the thigh, seldom more distally to the lower leg, ankle or foot. In the severe slip (>50 %, Meyerding III and IV), gait disturbances, numbness, muscle weakness and symptoms of cauda equina compression may be present. There is, however, no direct relationship between severity of subjective symptoms and the amount of slip.

24.2.2 Physical Examination

In low-grade slips (Meyerding I and II), the patient's gait and posture are usually normal unless radicular symptoms are present. The mobility of the lumbar spine is free or decreased due to muscle spasm and pain. Maximal extension may induce pain at the lumbosacral junction. There is local tenderness during palpation, and in many cases a step can be felt between the spinous processes at the level of the slip. Tightness of the ischiocrural muscles (hamstrings), typical for high-grade spondylolisthesis, is sometimes seen also in the symptomatic patients with a low-grade slip. Muscle strength, reflexes and skin sensation of the lower extremities are normal in the majority of patients.

In a high-grade slip (Meyerding grades III–IV slips and spondyloptosis), the clinical picture is very variable despite the severe local malalignment of the spine seen in the radiograph. In many cases, the patient's posture is disturbed in a typical way (Fig. 24.1): The sacrum is in vertical position due to retroversion of the pelvis. There is a short kyphosis at the lumbosacral junction and a compensatory hyperlordosis of the lumbar spine usually reaching up into the thoracic region [76]. The spine is scoliotic and often out of balance in the frontal as well as in the sagittal plane. The patient is unable to fully extend hips and knees during standing, and she/he walks with a typical pelvic waddle. In those patients the hamstrings are always extremely tight [100]. Signs of neural impairment (muscle weakness, disturbances of skin sensation, incontinence) may be present. Some patients look clinically normal and show, e.g. only some milder hamstring tightness. Astonishingly, even in severe slips objective neurologic findings are rare. Many patients are subjectively almost free of pain symptoms despite

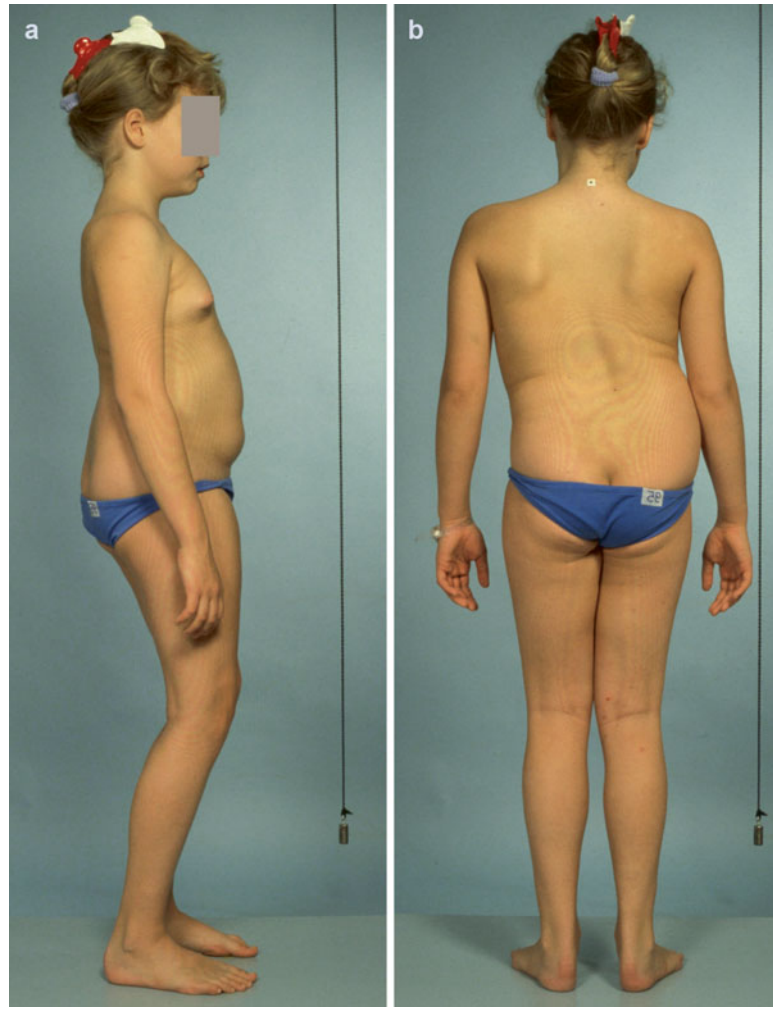


Fig. 24.1 (**a**) Typical clinical appearance of a symptomatic 11-year-old girl with high-grade isthmic spondylolisthesis. Vertical position of the sacrum due to retroversion of the pelvis, the patient is forced to stand with hips and knees flexed. (**b**) The spine is out of coronal balance, there is a secondary "sciatic" lumbar scoliosis

significant posture changes and hamstring tightness.

In some patients lumbar or thoracolumbar scoliosis is seen as a secondary phenomenon to spondylolisthesis. "Sciatic" forms (mainly in high-grade slips) are due to pain and muscle spasm and disappear usually after relieve of symptoms. Structural ("olisthetic") curves caused by rotational displacement of the slipped vertebra have to be followed closely and lumbosacral fusion operation is indicated if progression occurs [121]. Thoracic scoliosis in a patient with lumbar spondylolisthesis is assessed as a separate entity and treated according to the guidelines for scoliosis management.

24.3 Imaging

24.3.1 Plain Radiographs

Plain radiographs (PA and lateral) of the lumbar spine in standing position focused on the lumbosacral junction should be obtained. The images show the alignment of the lumbar spine and the true amount of vertebral slip if any. In most cases the lateral projection will also reveal the spondylolysis (Fig. 24.2). The use of traditional oblique plain radiographs to verify a lysis not visible in the standing lateral view is obsolete. At the author's institution, the slip is measured according to Laurent and Einola as the quotient between the sagittal slip and

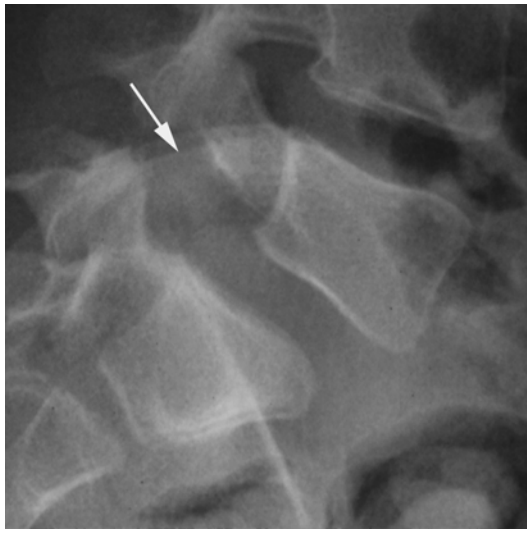


Fig. 24.2 Spondylolysis (*arrow*) and a low-grade L5 slip on a standing lateral radiograph

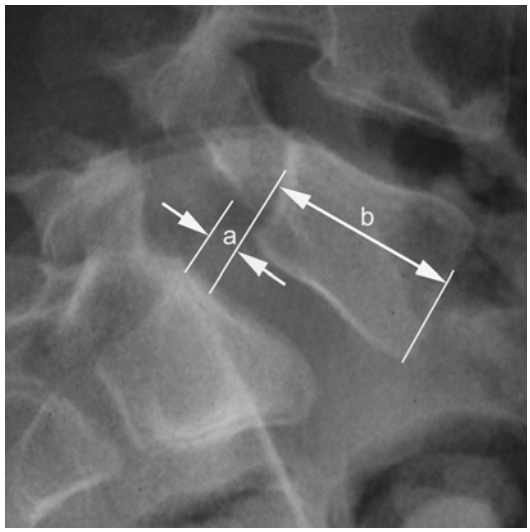


Fig. 24.3 Calculation of the percentage of vertebral slip according to Laurent and Einola [73]. Slip [%] = $a/b \times 100$

the sagittal length of the slipped vertebral body expressed in per cent (Fig. 24.3) [61]. This method allows for an exact measurement to detect and document also smaller changes especially if one thinks about follow-up radiographs to identify slip progression during growth. The sagittal lumbosacral alignment (lordosis/kyphosis) is assessed from the same radiograph and measured as the angle between the posterior border of the first sacral

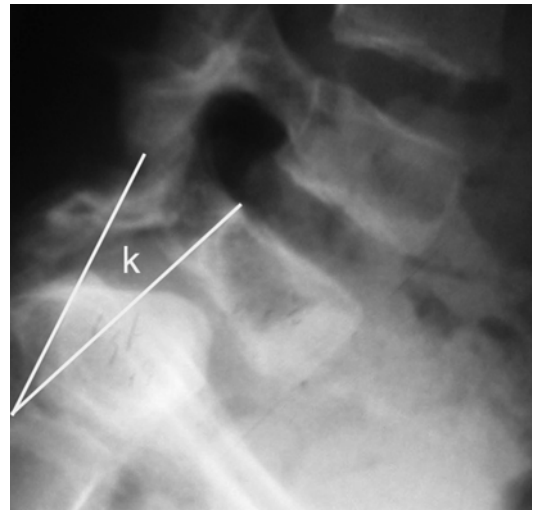


Fig. 24.4 Measurement of lumbosacral kyphosis as the angle (*k*) between the posterior border of S1 and the posterior (or anterior) border of L5

vertebral body and the anterior or posterior border of the fifth vertebral body (Fig. 24.4). Long-standing films of the whole spine in two planes are taken if the spine is clinically significantly out of balance and/or if scoliosis is present.

24.3.2 Functional Radiographs

Flexion-extension radiographs have been traditionally used in order to detect possible "instability" in the olisthetic segment. They are not in use anymore as we could not see any value for decision making concerning the patient's treatment. However, in high-grade slips with lumbosacral kyphosis, a lateral hyperextension radiograph in supine position is taken preoperatively (Fig. 24.5). It demonstrates the reducibility of the slipped vertebra to judge whether the disc space below the vertebra will be accessible during a planned anterior procedure without the need for instrumented reduction.

24.3.3 Computed Tomography (CT)

In most cases the lysis can be easily seen from the standing lateral radiograph. If in doubt, a CT image with the gantry tilted to obtain slices in the

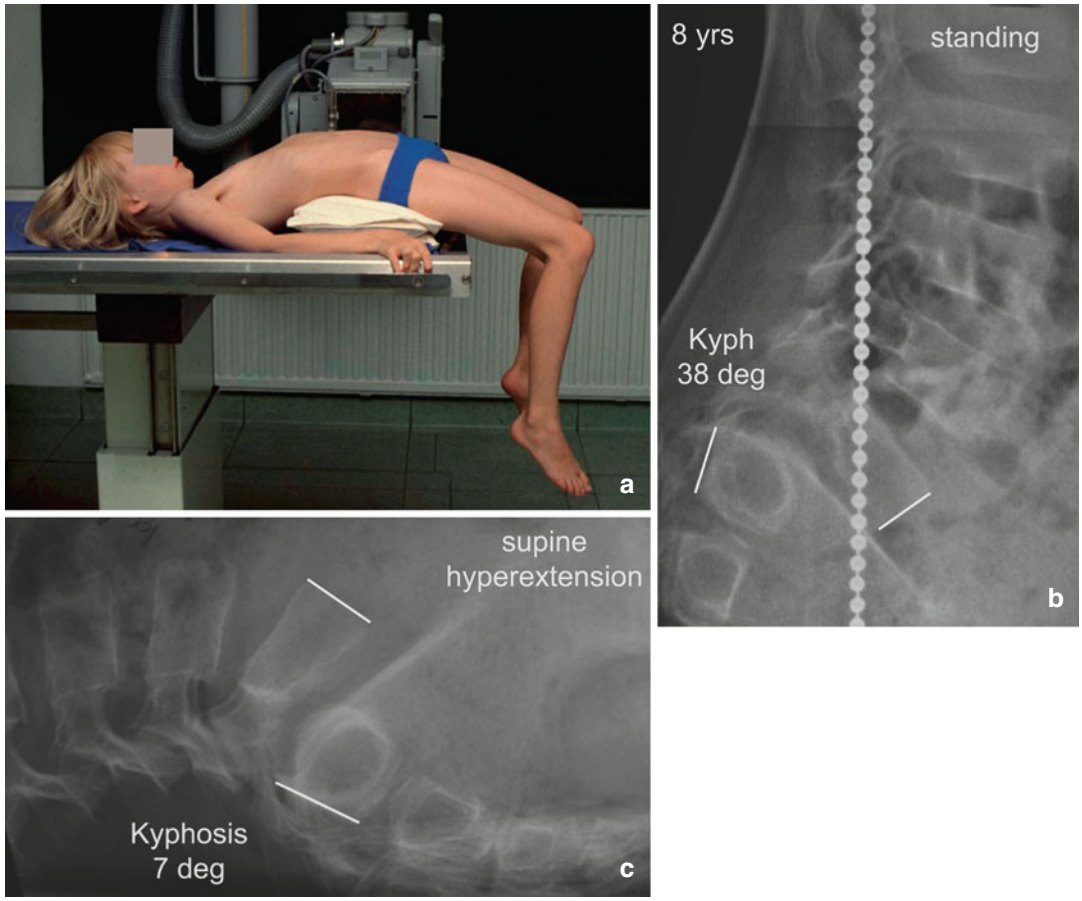


Fig. 24.5 (**a**) Patient positioning for the lateral supine hyperextension radiograph of the lumbosacral junction in high-grade slips. (**b**) Standing lateral radiograph shows significant lumbosacral kyphosis. (**c**) On the supine hyperextension radiograph of the same patient, a marked decrease of the kyphosis is visible

longitudinal direction of the isthmus should be taken. It is the most reliable imaging mode for demonstrating the spondylolysis (Fig. 24.6). CT is also very valuable to assess possible healing of the defect [5, 37].

24.3.4 Magnetic Resonance Imaging (MRI)

Increasingly, MRI is used as a primary imaging mode for children with low-back pain. Especially in young athletes increased signal intensity is seen frequently in the area of the isthmus or the pedicles. This is interpreted as a stress reaction. Its importance and natural history are unclear so far. There are difficulties to distinguish these stress reactions from true spondylolysis in MRI. Prospective studies are needed to clarify this phenomenon.

In low-grade slips without neurologic signs, there is no rational indication for MR imaging. MRI is indicated in cases with neurologic symptoms, cauda equina syndrome, or if disc herniation is suspected. It is helpful to demonstrate the shape of the spinal canal, the intervertebral foramina, and possible compression of neural structures (Fig. 24.7).

MR also allows to assess the condition of the intervertebral discs at and adjacent to the olisthetic segment. The disc below the slipped vertebra is often pathologic already in young

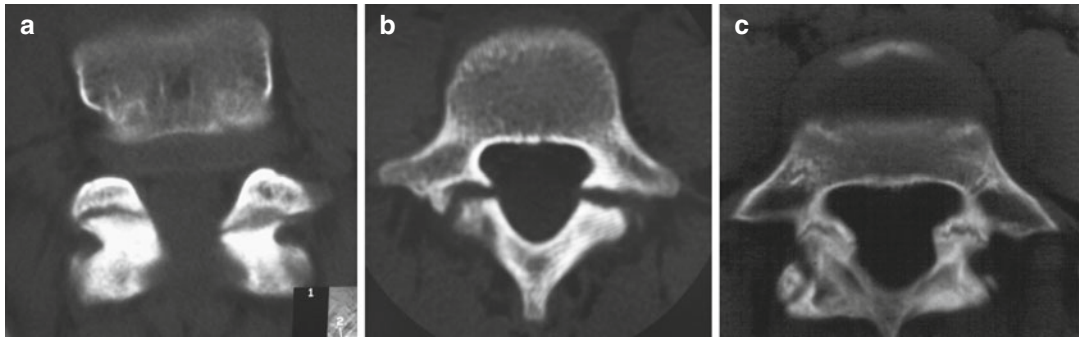


Fig. 24.6 (**a**) Isthmus CT-image of a 10-year-old boy, "early-traumatic" spondylolysis. (**b**) Isthmus CT-image of an 11-year-old girl, "atrophic" spondylolysis. (**c**) Isthmus CT-image of an 14-year-old boy, "hypertrophic" spondylolysis

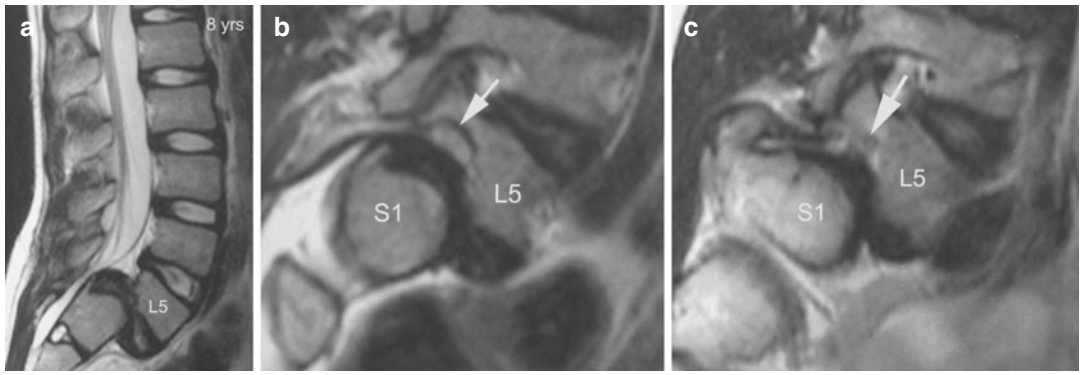


Fig. 24.7 (**a**) Midsagittal lumbar MR image in high-grade spondylolisthesis. The central spinal canal is narrowed; the L5-S1 disc is severely damaged, (**b**, **c**) Right and left parasagittal lumbar MR images in high-grade spondylolisthesis. The L5 roots (*arrow*) are caught between the pedicle and the disc

individuals regardless of whether they do have pain symptoms or not. Dehydration of the adjacent disc above the slipped vertebra is relatively common in symptomatic patients [114, 115]. As the clinical relevance of disc dehydration seen on MR images of young persons is unclear, MR is not of value for clinical decision making in spondylolisthesis in this respect [105].

Symptomatic disc herniation at the level of the slip is very rare in patients with isthmic spondylolisthesis [101].

24.3.5 Single-Photon Emission Computed Tomography (SPECT)

The SPECT technique is nowadays often used for evaluation of low-back pain especially in young athletes. It shows increased uptake in stress reactions, microfractures and fractures. It allows differentiating chronic spondylolysis (pseudarthrosis) from fresh, active lesions which theoretically should have a higher healing potential. However, its predictive value concerning healing of the spondylolysis has not been established yet [8, 19, 81, 133, 134].

24.4 Treatment

The benign natural history of the condition should always be kept in mind when weighing the necessity for treatment and the treatment options. The parents are usually very worried after learning that there is something "broken" in the lower back of their child. In every case, it is very important to explain the basically benign nature of the course to the patient and to the parents. In many cases symptoms resolve after

several months without any special treatment. At the same time it should be made clear that the condition may not be ignored either. Follow-up for a certain period of time is necessary to act appropriately if significant progression occurs. The parents should also be informed that effective treatment is at hand if prolonged severe subjective symptoms are present or marked slip progression is seen.

The only case for immediate decision towards active intervention is a high-grade slip with lumbosacral kyphosis and/or a significant neurologic deficit.

24.4.1 Observation

Rapid growth and a slip of more than 20 % have been identified as risk factors for progression [43, 123]. Therefore, children before or during the growth spurt have to be checked at regular intervals until the rapid growth is over [146, 123]. Plain standing lateral radiographs of the lumbar spine are obtained every 6–12 months depending on the degree of slip at admission and the age of the patient. There is no need for restriction of physical activities during follow-up. At the end of the observation period, the patient and the parents should be assured that there are no restrictions in view of future sports activities or choice of occupation.

24.4.2 Non-operative Treatment

Symptomatic spondylolysis or low-grade spondylolisthesis is primarily typically treated non-operatively by decreasing the level of physical activities, strengthening of back and abdominal muscles, and sometimes a brace [6, 23, 72, 131]. Patients participating regularly in sports are advised to modify their training program to avoid pain-causing exercises. But there is no reason to stop all physical activities. According to the literature, the functional outcome after brace treatment of spondylolysis in young athletes is good or excellent in 80 % or more. Healing of the defect can be demonstrated radiologically in 16–57 % of involved patients. Unilateral defects seem to heal more often than bilateral defects as do defects at L4 in comparison to L5 defects. There is no correlation between healing and good clinical outcome. Neither the efficacy of bracing nor the predictive value of increased activity of the lysis in SPECT scans can be demonstrated definitely as there are no prospective comparative studies available [7, 30, 81, 109, 131, 133]. Klein et al. presented a meta-analysis of 15 observational studies (665 patients) on non-operative treatment for spondylolysis and Grade I spondylolisthesis [54]. They concluded that non-operative treatment is successful in 83.9 %, bracing has no influence on the result, and most of the defects do not heal.

24.4.3 Operative Treatment

The data on operative treatment of spondylolysis and spondylolisthesis in children (up to 12 years of age) are very sparse due to the fact that in the vast majority of reports children and adolescents are treated as one group. The impact of patients' age at operation on the results is usually not analysed. No randomised trials comparing operative treatment to natural history are available thus far. In his retrospective long-term follow-up study, Seitsalo investigated 149 patients with low-grade slips after a mean follow-up of 13.3 years [119]. Seventy-two patients (mean age 13.8 years, mean slip 16.2 %) had conservative treatment or no treatment at all while 77 patients (mean age 14.6 years, mean slip 16.6 %) were treated by uninstrumented posterior or posterolateral fusion. At follow-up, 75 % of the conservatively treated patients and 87 % of the operatively treated patients were free of pain. None of the primarily conservatively treated patients had an operation at a later date. In the conservative group, 6 of 72 patients (8.3 %) patients and 4 of 77 (5.2 %) patients in the operative group reported decreased working ability. In a recent long-term cohort study, Jalanko et al. compared the results after fusion surgery between children operated on before onset of the pubertal growth spurt (females <12.5 years old, males <14.5 years old) and adolescents [47]. They could not find any differences of clinical importance in patients'

functional, radiographic and health-related quality of life outcomes between the two age groups, neither for low-grade nor for high-grade slips.

The indication for operation in children and adolescents depends on the amount of slip (high-grade or low-grade), the age of the patient (before, during or after the growth spurt), and the clinical signs and symptoms. Neurologic symptoms (cauda equina syndrome, peroneus paresis) are a clear indication for operation. However, those occur very rarely even in high-grade slips. The most common reason for operation in low-grade slips is pain not responding to non-operative measures. In children with a slip of 50 % or more, operation is recommended also to prevent further progression even if the patient has only minor symptoms or no symptoms at all. Operation should also be considered in a very young patient with a slip of over 20 % if progression occurs during follow-up.

The choice of the operative technique depends on the percentage of slip and/or lumbosacral kyphosis and on the personal experience and preferences of the surgeon. Table 24.3 represents the recommendations developed at the author's institution. It can be used as a guideline for decision making. The listed numbers for slip percentages and degrees of lumbosacral kyphosis are not based on scientific evidence. They mark a smooth transition which cannot be defined with mathematical accuracy. The final decision is always made according to the individual situation of the patient taking into consideration patient's stage of skeletal maturity, gender, individual anatomic features of the slip, ability to co-operate, patient's and parents' hopes and desires, and, last but not least, the surgeon's personal experience.

Table 24.3 Management of isthmic spondylolisthesis in children and adolescents

Slip (%)[a]	Symptoms	Treatment
0–25	No	Follow-up during growth
0–25	Yes	Non-operative Operative[b] Uninstrumented posterolateral fusion
>25–50	Yes/no	Consider post-lat fusion before growth spurt
>50 L-s kyphosis <20°	Yes/no	Uninstrumented anterior fusion
>50–90 L-s kyphosis >20°	Yes/noL-s kyphosis	Uninstrumented anterior/posterior fusion
>90–100 (ptosis)	Yes/no	Partial reduction or resection +
		Instrumented anterior/posterior fusion

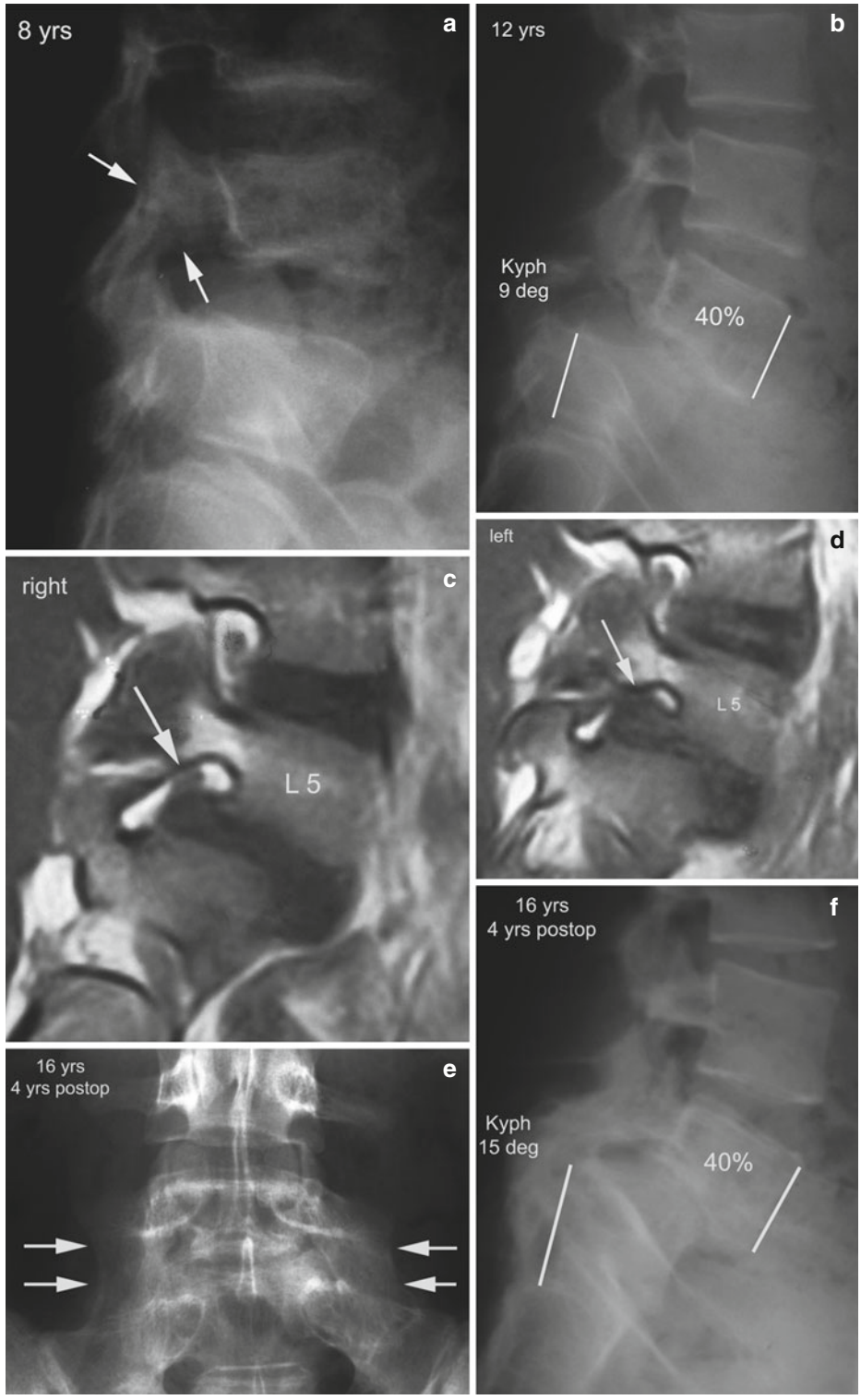
[a]The listed values for slip percentages and degrees of lumbosacral kyphosis are not based on scientific evidence. They mark a smooth transition which cannot be defined with mathematical accuracy. The final decision has to be made after assessing the overall picture of the individual patient

[b]If significant symptoms not resolving under non-operative treatment

24.4.3.1 Spondylolysis and Low-Grade Slip (≤50 %, Meyerding I and II)

Uninstrumented segmental posterolateral fusion in situ using autogenous bone from the posterior iliac crest is the method of choice for cases with a percentage of slip up to 50 % (Fig. 24.8). The operation is performed through the bilateral paraspinal muscle split approach as recommended by Wiltse [146, 147]. The segment above the slipped vertebra is usually not included into the fusion even if the disc shows signs of dehydration in

Fig. 24.8 Uninstrumented posterolateral fusion for symptomatic low-grade slip in a 12-year-old male. (**a**) L5 spondylolysis at 8 years of age. (**b**) tanding lateral radiograph at 12 years of age. Then slip has progressed. The patient suffers from low-back pain and left leg pain. (**c**) On right parasagittal MR image, the right L5 nerve root (*arrow*) is free on MRI. (**d**) On left parasagittal MR image, compression of the left L5 nerve root (*arrow*). (**e**) Plain AP radiograph 4 years after posterolateral fusion without decompression. Note bilateral mature fusion mass (*arrows*). (**f**) Standing lateral radiograph 4 years postoperatively. Minimal progression of the kyphosis. The patient is free of symptoms


8 yrs
a
12 yrs
b
Kyph
9 deg
40%
left
d
L 5
right
c
L 5
16 yrs
4 yrs postop
e
16 yrs
4 yrs postop
f
Kyph
15 deg
40%

MRI. The patient is mobilised 1–2 days after the operation wearing a soft brace for 3 months' time. Sports activities are forbidden for 6–12 months depending on the radiologic development of the fusion. There are no restrictions of physical activities after solid bony healing. The method is very safe and effective. There are no specific complications. In this young age group it leads to bony fusion in 80–90 %. Subjective results and functional outcome are good or satisfactory in 82–96 % of the patients [62, 65, 116, 117, 126]. A recent long-term study in 107 children and adolescents with a mean age at operation of 15.9 years (range, 8.1–19.8) and a mean follow-up of 20 years has proven the lasting effectiveness and reliability of this method [58]. The mean Oswestry Disability Score [25] was 7.6 (range, 0–68) at last follow-up. It was in the normal range (0–20) in 100 out of 107 patients (93 %). Six (6 %) out of 107 patients had an Oswestry score of 20–40 (moderate disability); one patient had a score of 68 (crippled). Pseudarthrosis (17 % after posterolateral fusion) and adjacent disc degeneration on plain radiographs (12 %) did not correlate with poor outcome. The Scoliosis Research Society outcome instrument [35] yielded a mean of 94.0 points (range, 44–114 points) at follow-up [40]. Degenerative changes in MRI at follow-up did not have any significant influence on patients' outcome [105].

In low-grade slips, decompressive laminectomy is indicated in young patients only in rare cases with true impingement of neural structures. However, this has not been seen by the author in a low-grade slip during a period of over 30 years. Pseudoradicular symptoms (radiating pain to the posterior aspect of the thigh) and hamstring tightness resolve without laminectomy due to stabilisation of the segment by fusion. If decompression is performed during growth segmental fusion has to be added always to prevent subsequent progression of the slip [95].

The use of instrumentation has not been shown to give any advantages in low-grad slips in this age group. Nor is there any reason for reduction of low-grade slips. The author agrees fully with this statement published by Wiltse and Jackson in 1976 [146]. Internal fixation, with or without reduction, is connected with longer operation time, more severe muscle trauma, an increased risk of complications, and higher costs. It would probably increase the fusion rate. But the disadvantages mentioned above would not countervail this, as pseudarthrosis does not have a measurable negative effect on the outcome in this group of patients [58, 65, 119, 124].

In cases of spondylolysis without a slip or with a slip of less than 25 %, the direct repair of the isthmic defect is recommended by some authors reporting favourable results using different methods of internal fixation (screws, cerclage wire, butterfly plate, hook plate, pedicle screws and rods) [16, 33, 48, 51, 67, 86, 91]. Several authors reported favourable outcome especially in younger patients [14, 39, 46, 91]. There are no series published dealing exclusively with children. At the author's institution Scott's wiring technique with autologous bone grafting has been used [91]. For postoperative treatment a plastic TLSO was applied for 3–6 months. In a comparative study in children and adolescents, the results after mid-term and long-term follow-up were very good in the majority of cases, but not better than the results of uninstrumented segmental fusion. Thus, the benefit from saving the lytic-olisthetic motion segment could not be demonstrated so far [116, 117]. At present, the direct repair is used by the author only in cases of spondylolysis or minimal slips in the segments above L5.

24.4.3.2 High-Grade Slip (>50 %, Meyerding III and IV)

If the slip approaches 50 % the biomechanical situation changes profoundly with far reaching consequences for the sagittal alignment of the entire spine [66]. The physiologic lumbosacral lordosis decreases and, dependent of the amount of displacement, a kyphosis develops due to absence of anterior support for the slipping vertebra (Fig. 24.9). In the growing patient, this kyphotic deformity has a risk for progression in almost 100 %. Operation should be considered even in patients with minimal subjective symptoms or no symptoms at all [146]. There is no data showing that non-operative measures (exercises,

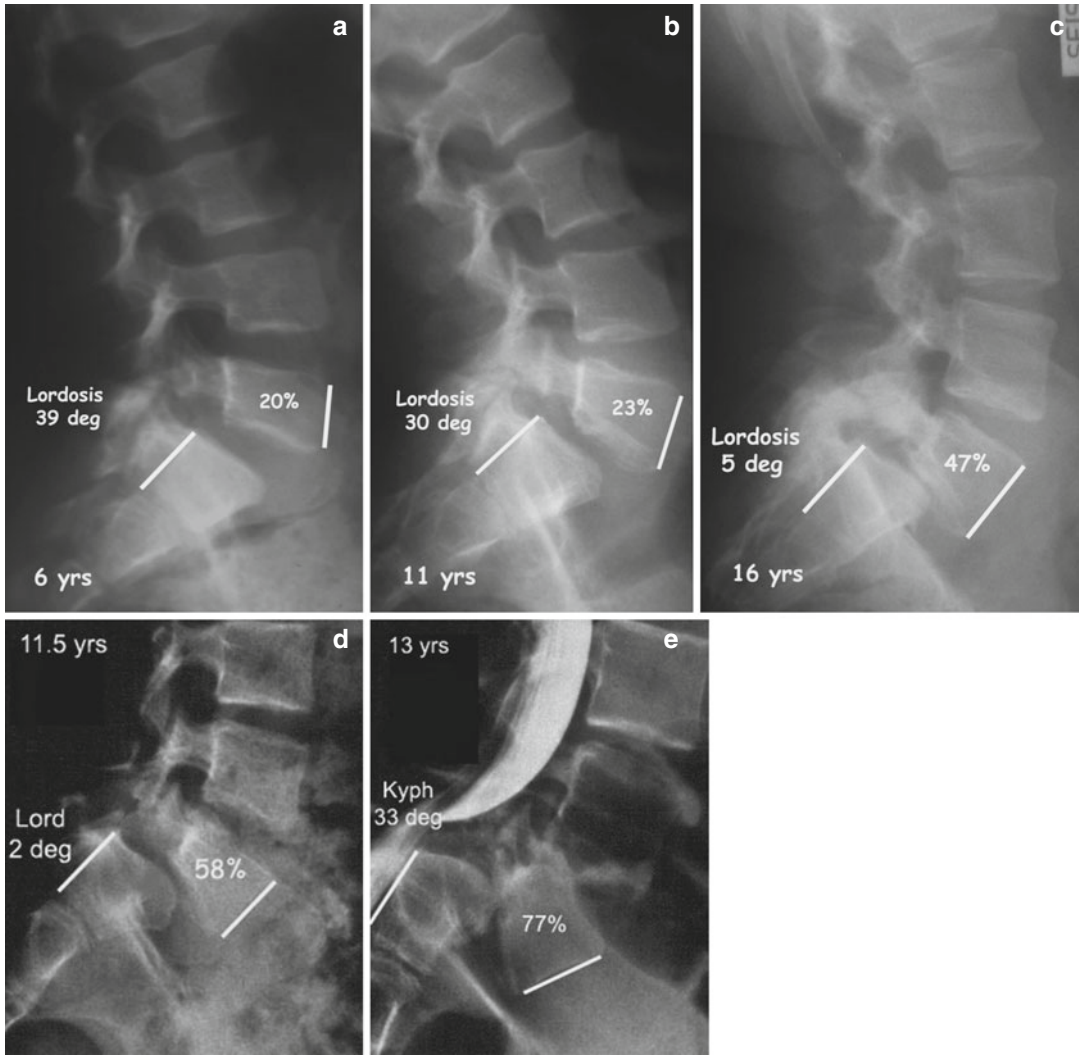


Fig. 24.9 Radiographs demonstrating changes of lumbosacral alignment during slip progression. Standing lateral radiographs of a female patient at 6 years of age (**a**), 11 (**b**) and 16 (**c**) years show marked loss of the physiologic L5-S1 lordosis during slip progression. In another female patient, rapid deterioration of the sagittal alignment from 2° of lordosis (**d**) to 33° (**e**) of lumbosacral kyphosis within 18 months time

bracing) or restriction of sports activities would stop the progression. One should not wait and see too long. Proceeding progression makes the necessary operation technically more difficult, increases the risk of complications, and leads possibly to an inferior result. It has, however, to be noted that in some cases even patients with high-grade slips or even spondyloptosis remain subjectively symptom-free. The author agrees with Bridwell that there is no "right way" to treat all high-grade slips [15]. But overtreatment for the sake of radiologic correction should be avoided. The methods applied should be assessed critically for their benefit for the patient in the long run in terms of clinical outcome and function.

A considerable variety of methods for operative treatment of high-grade slips has been published: uninstrumented posterior or posterolateral fusion from L3 or L4 to S1 [34, 41, 45, 59, 65, 106, 122, 146], uninstrumented anterior interbody fusion [41, 102, 103, 136], uninstrumented

combined fusion [57], uninstrumented posterolateral fusion L4-S1 and anterior fusion L5-S1 with cast immobilisation after preoperative gradual conservative reduction [59, 138], uninstrumented posterolateral fusion L3 or L4 to S1 with postoperative cast reduction [17], anterior reduction with anterior screw fixation and interbody fusion, posterior pedicle screw reduction with or without decompression and posterior or combined fusion using bone graft or cages [24, 77, 84, 88, 102, 103, 108, 110] or the Bohlman technique utilising a transsacral fibular strut graft [10, 36, 128], or a special titanium cage [2, 127], and anterior and posterior reduction with decompression, and double-plating [139].

According to Bradford, the goals of treatment for high-grade spondylolisthesis are to prevent progression, to relieve pain, to improve function and to reverse the neurologic deficit if there is any [13]. These goals can be achieved in the vast majority of cases safely by in situ fusion which wrongly is of bad repute. The reason for the negative attitude of many surgeons towards in situ fusion seems to be that many of them have seen symptomatic adult patients with a high-grade slip up to spondyloptosis who had a posterior or posterolateral *"in situ"* fusion of a more or less severe slip as teenagers. Usually, in the early years after the primary operation, they were symptom free. But with time, posture deteriorated and symptoms reappeared, sometimes they became even worse than before the operation. When analysing the radiographs, one sees that slow progression of the slip and the kyphosis happened over the years despite a solid-looking fusion mass. Retrospectively, one can say that although in situ fusion was attempted it was not achieved. Several of such cases with slow progression after posterior or posterolateral so-called in situ fusion L4 to S1are are shown in the very instructive papers by Taillard and Burkus et al. [17, 134]. The cause for the failure is misunderstanding of the biomechanics of a high-grade slip which in fact is a progressive kyphosis. Anterior bony support is insufficient or missing totally. The disc below the significantly slipped vertebra is always severely damaged. The disk will degenerate further and atrophy due to loss of functional motion after fusion. This all together induces increasing flexion moments on the posterior fusion mass which will bend and elongate. It must be stressed here that in the author's language successful in situ fusion does mean that a solid bony fusion is achieved and even after long-term follow-up the position of the fused vertebra is not significantly worse than before the operation.

Biomechanically, the most reasonable procedure to stop the progression of a kyphotic deformity is to provide anterior support. This is the rationale for anterior fusion. At the author's institution, uninstrumented anterior interbody fusion in situ without decompression is the method of choice for high-grade slips with no ore minimal (up to 10–20°) lumbosacral kyphosis (Fig. 24.10). The operation is performed through a transperitoneal or retroperitoneal approach using two to three autogenous tricortical iliac crest grafts. Uninstrumented combined anterior and posterolateral fusion in situ without decompression is preferred for slips with greater lumbosacral kyphosis (more than 20°). Combined fusion has been shown to be more effective in preventing the postoperative progression of the lumbosacral kyphosis, i.e. to achieve a true and lasting in situ fusion without late deterioration [41, 106]. Using this technique there is no need to include more than the olisthetic segment into the fusion. After anterior or combined procedures the patient is mobilised at the second or third postoperative day wearing a plastic TLSO for 3–6 months. Hamstring tightness disappears and spinal balance is regained within a few weeks although no decompression has been performed (Figs. 24.11, 24.12, and 24.13). The clinical short- and mid-term results of anterior and combined fusion in the severe slip are comparable to the results of posterior or posterolateral fusion [49, 59, 64, 122, 125, 136]. In a recent long-term follow-up study (67 patients, slip 50–100 %, mean age at operation 14.4 years, range 8.9–19.6, follow-up of 10.7–26 years.), the outcome after three different uninstrumented in situ fusion techniques (posterolateral, anterior, combined) without decompression was compared. At final follow-up, 14 % in the posterolateral and in the anterior fusion

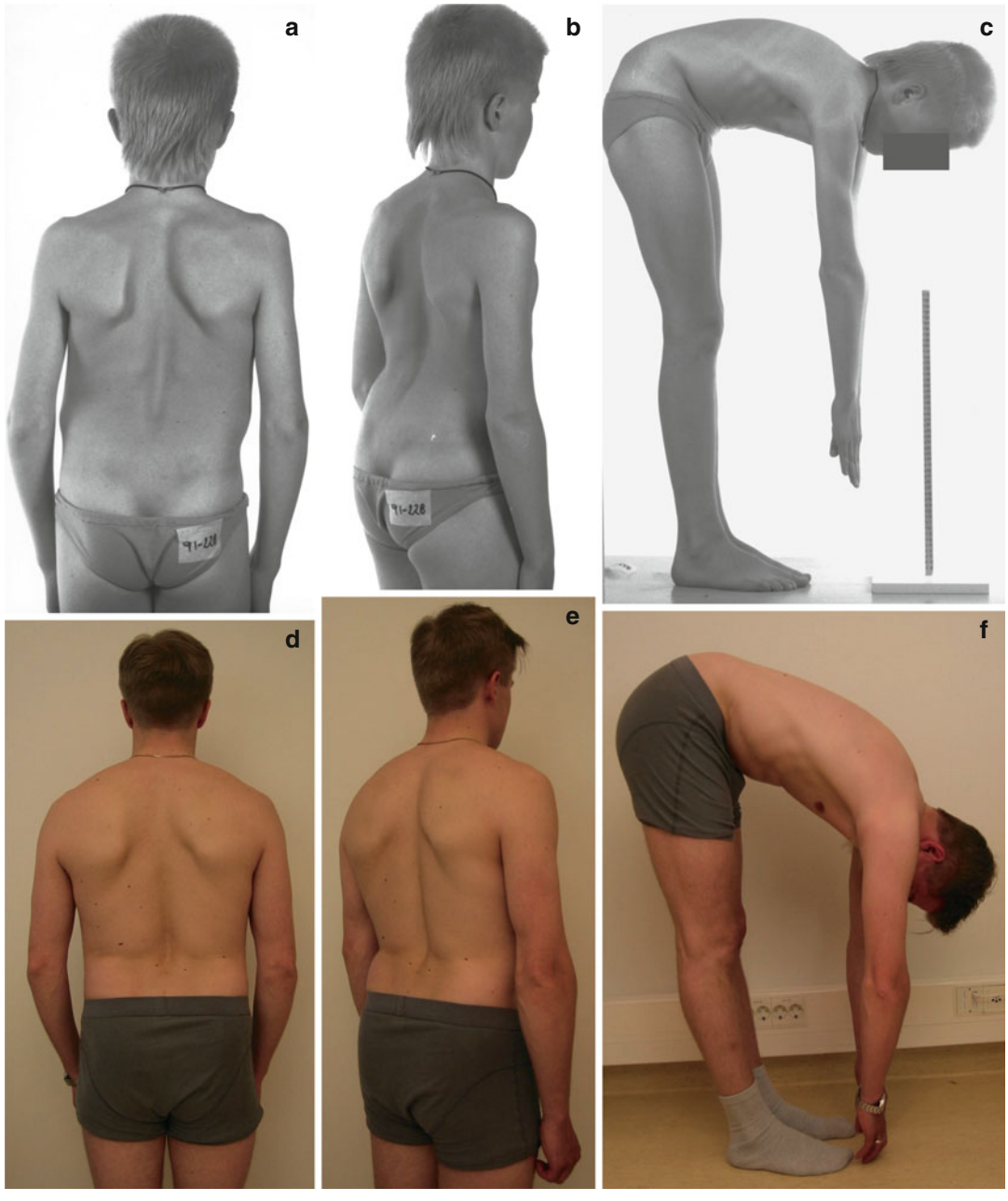


Fig. 24.10 Uninstrumented anterior in situ fusion without decompression for high-grade slip in an 11-year-old boy. Preoperative photographs (**a**–**c**) show typical posture changes. (**a**) The spine is slightly out of balance to the right. The spine appears to be lordotic. (**b**) he lordosis of the thoracolumbar spine is clearly seen. Note pelvic retroversion and positive sagittal balance. (**c**) C) Forward bending is restricted due to hamstring tightness preventing anterior rotation of the pelvis. (**d**) D) Eleven years postoperatively, the patient is free of symptoms. The spine is balanced in the coronal plane. (**e**) The sagittal profile is normal. (**f**) Full forward bending is possible. Hamstring tightness has resolved. (**g**) G) Preoperative standing lateral radiograph showing L5 slip of 66 % and a lumbosacral kyphosis of 18°. (**h**) At follow-up 11 years postoperatively, standing lateral radiograph shows solid anterior fusion. No progression of the deformity

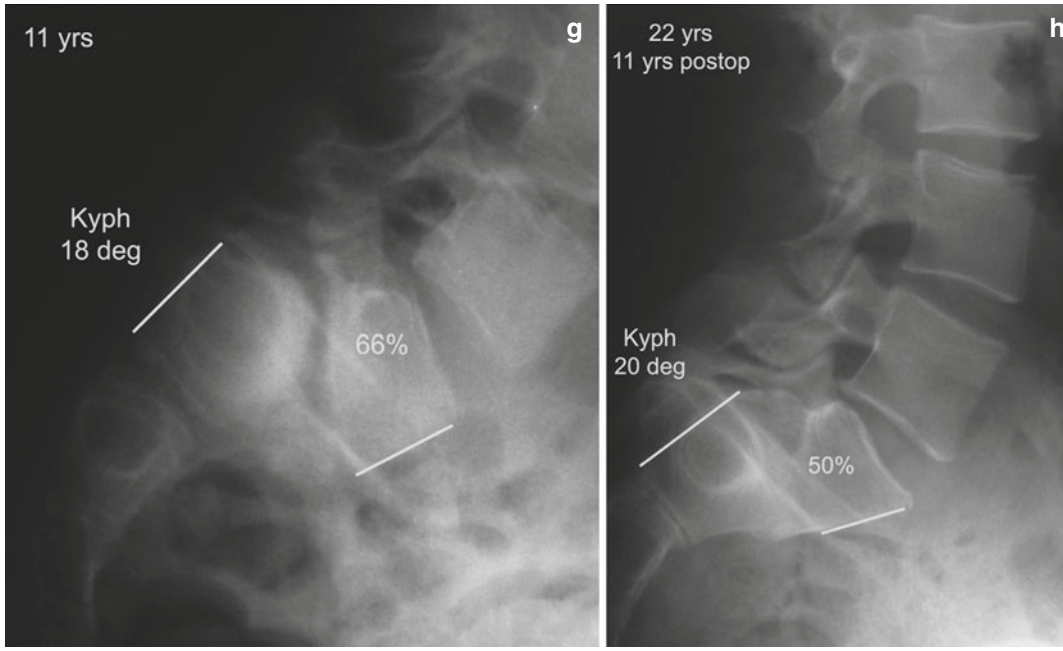
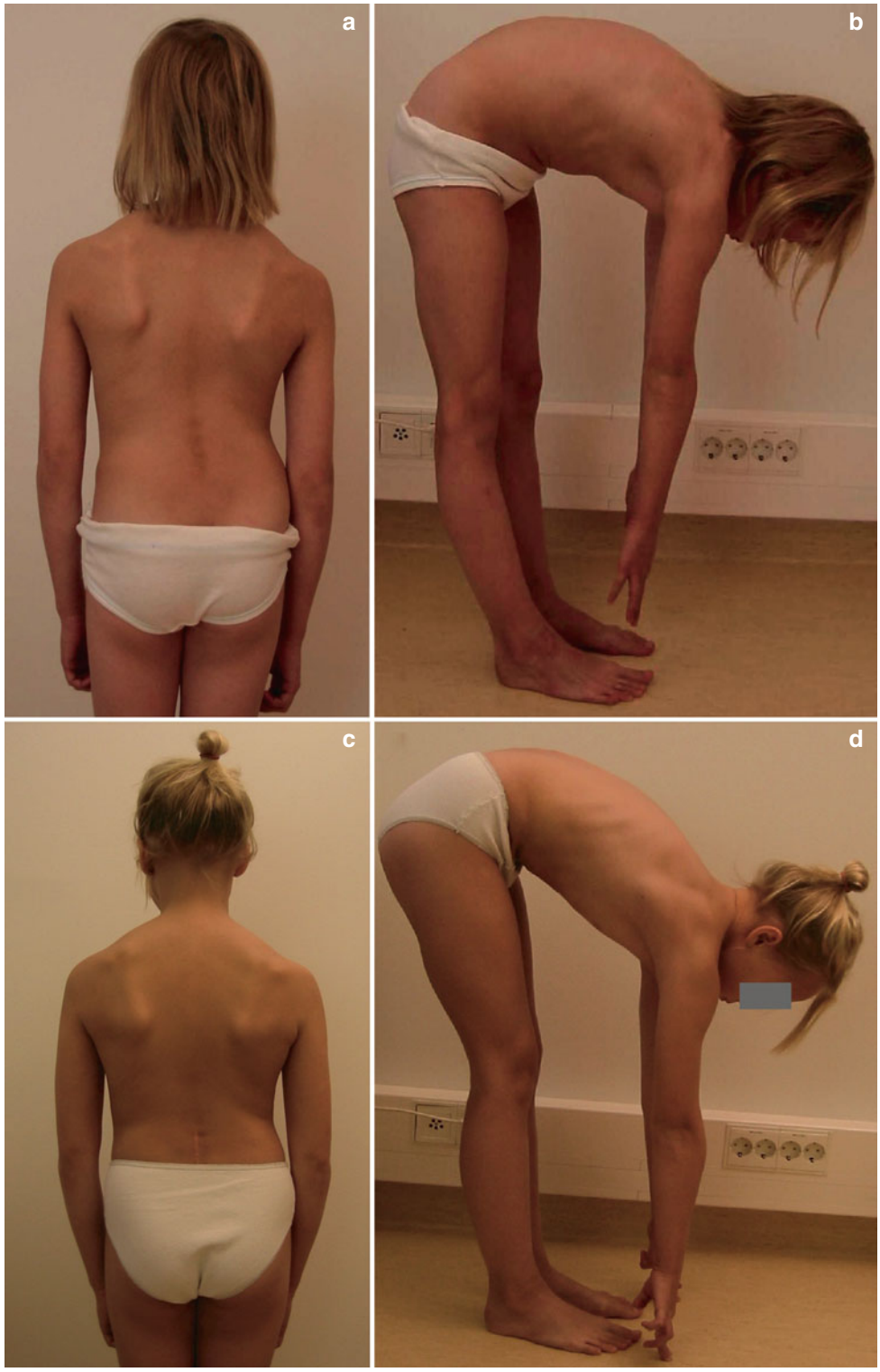


Fig. 24.10 (continued)

Fig. 24.11 Uninstrumented anterior-posterior in situ fusion without decompression for a high-grade L5 slip in a 9-year-old female athlete. Before the operation the patient was free of pain symptoms. A scoliosis was detected by the school nurse. In addition, she had mild bilateral hamstring tightness. (**a**) C-shaped left convex secondary scoliosis. (**b**) Mild hamstring tightness prevents maximal forward bending. (**c**) One year postoperatively, the scoliosis has resolved. (**d**) Free forward bending 1 year postoperatively. (**e**) Four years postoperatively, the spine is clinically balanced. (**f**) Four years postoperatively, the sagittal profile is clinically normal. (**g**) Four years postoperatively, forward bending is free. (**h**) Preoperative standing lateral radiograph. L5 slip of 52 %. Lumbosacral kyphosis of 24°. (**i**) Standing lateral radiograph six months postoperatively. Anterior fusion is healed. No progression of the deformity. (**j**) Two years postoperatively, on standing lateral radiograph, solid fusion, no progression of the deformity. (**k**) Two years postoperatively, solid posterolateral fusion on AP radiograph. (**l**) Whole spine lateral radiograph 4 years postoperatively shows satisfactory sagittal alignment. (**m**) On preoperative standing PA whole spine radiograph, secondary scoliosis is seen. (**n**) Spontaneous improvement of the scoliosis on whole spine PA radiograph 3 months postoperatively. (**o**) Unimportant residual scoliotic curve on whole spine PA radiograph 1 year postoperatively


a
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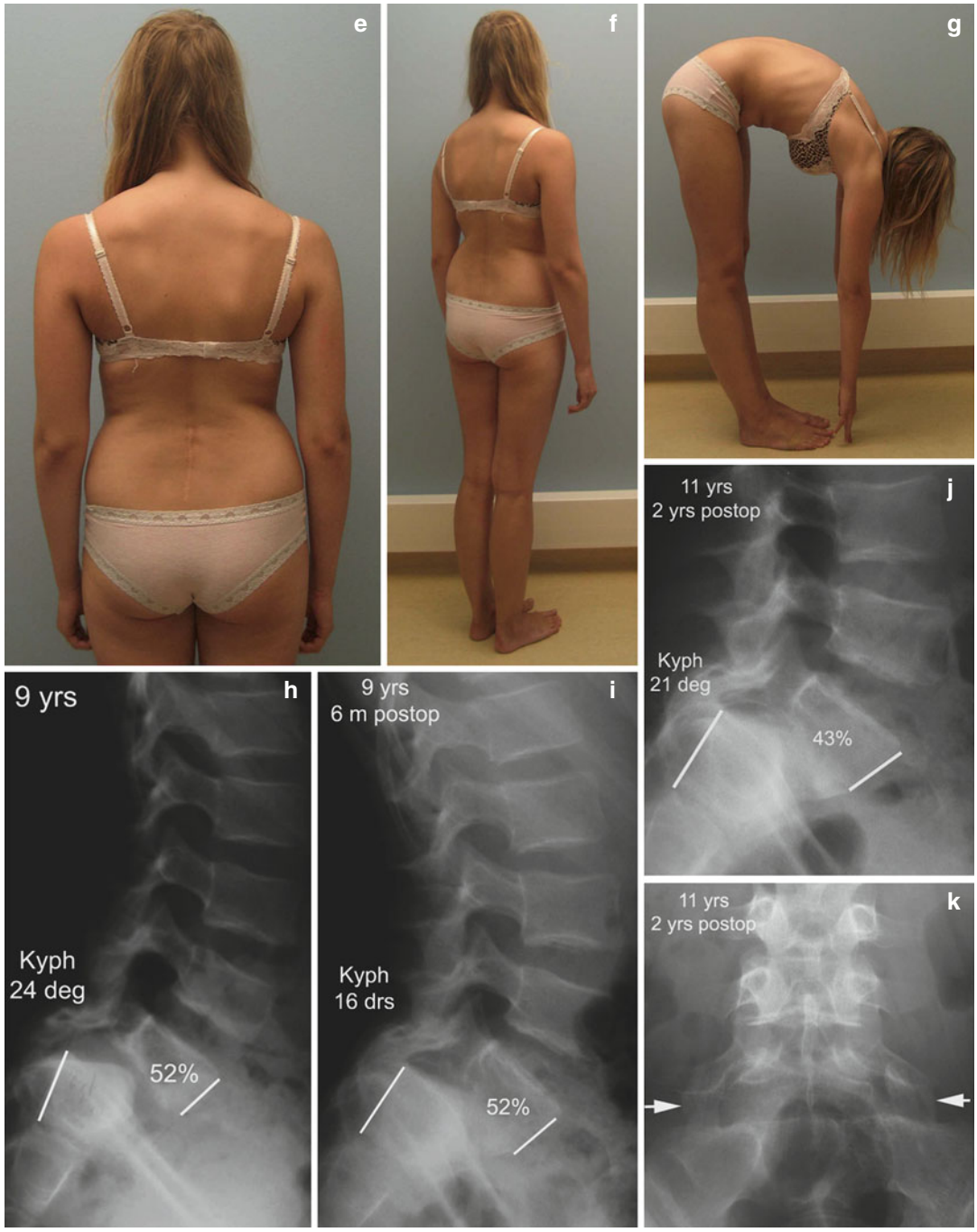

Fig. 24.11 (continued)

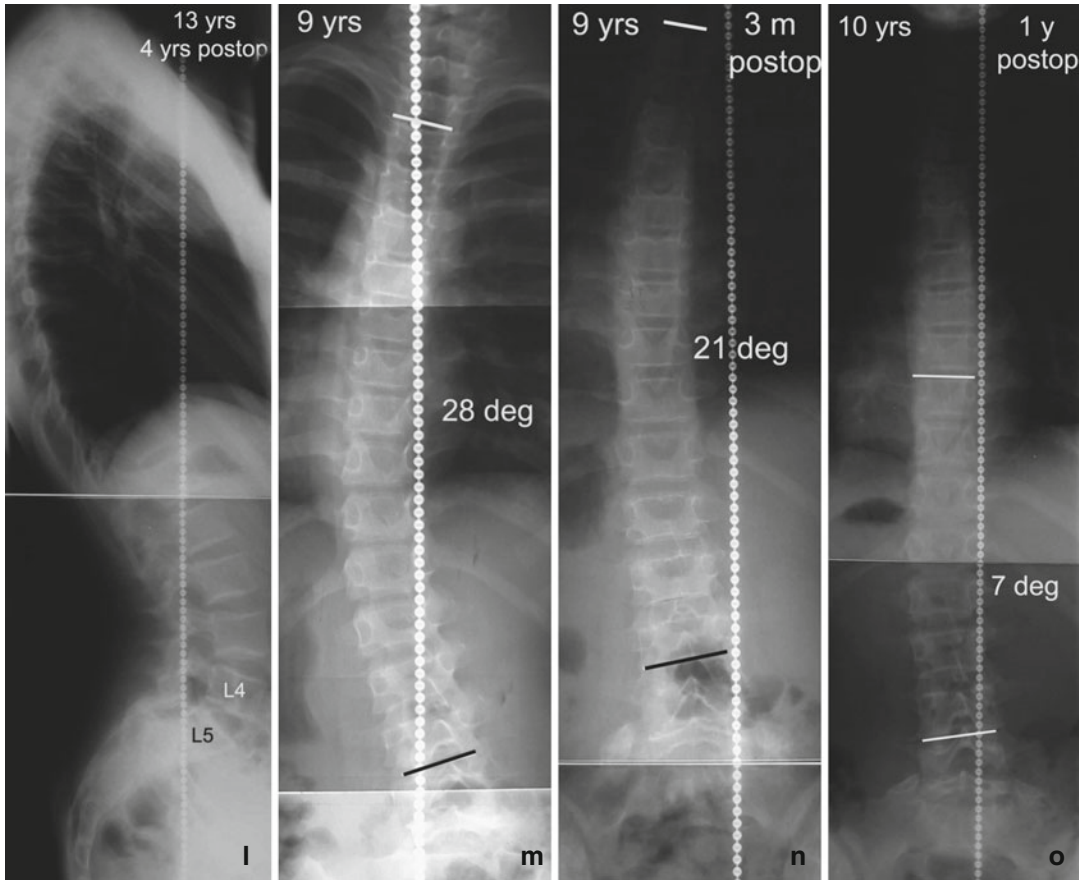

Fig. 24.11 (continued)

group reported low-back pain at rest often or very often, but none in the circumferential fusion group. The mean Oswestry index was 9.7 (0–62), 8.1 (0–32) and 2.3 (0–14) respectively, indicating combined fusion being slightly but not significantly superior. Radiographs showed some progression of the mean lumbosacral kyphosis during follow-up in the posterolateral and in the anterior only fusion group. No progression of the lumbosacral kyphosis was detected after combined fusion [106]. A comparison of the three groups using the Scoliosis Research Society questionnaire yielded the same kind of results with a slightly better outcome in the circumferential fusion group [40, 57]. It has, however to be noted that those patients are still in their thirties. And no data is available showing what happens when they reach midlife and seniority.

The risk of complications is obviously higher if using the anterior approach. Massive intraoperative bleeding, postoperative thrombosis and retrograde ejaculation in male patients may occur. However, in experienced hands these complications are very rare [136]. The anterior approach can be avoided by performing a posterior interbody fusion (PLIF) or by utilising Bohlman's transsacral strut graft technique [10, 35]. These procedures, however, make it necessary to resect the posterior structures in order to open the spinal canal with all its drawbacks like exposing the neural structures, significant muscle trauma and loss of interspinous ligament continuity. In contrast, the direct anterior interbody fusion in combination with a posterolateral fusion through the paraspinal muscle split approach without touching the midline structures causes

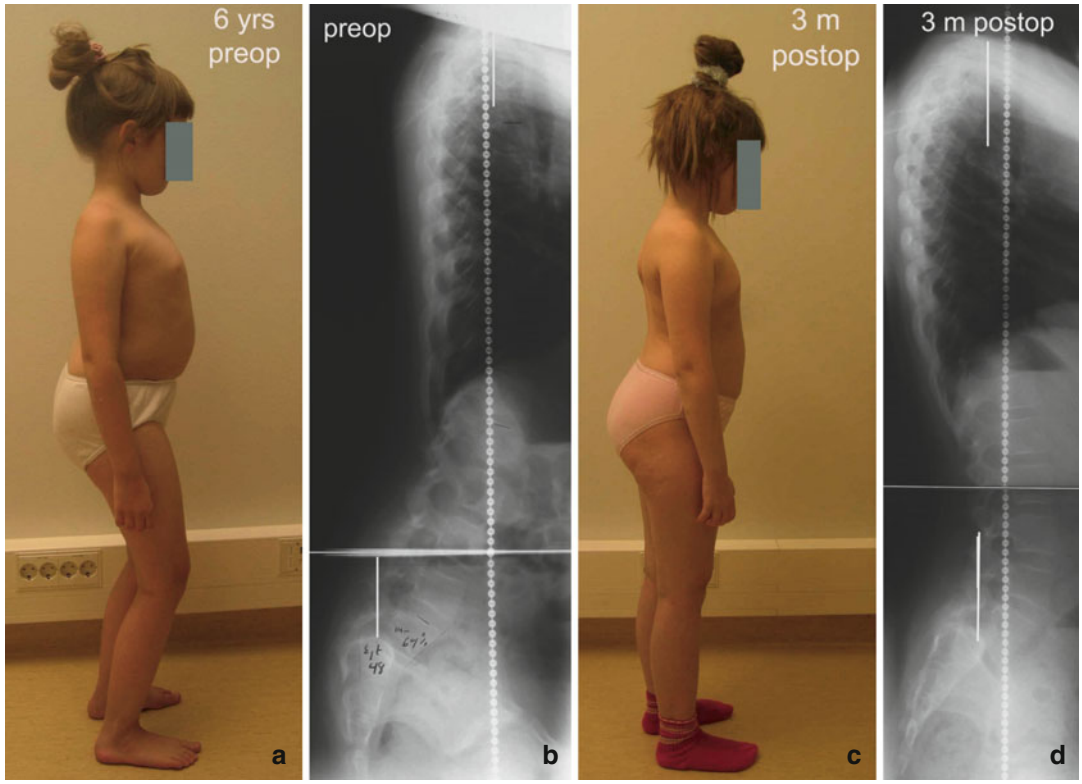

Fig. 24.12 Sagittal rebalancing of the spine in a 6-year-old female with a high-grade slip after combined uninstrumented in situ fusion without decompression. (**a**) Preoperatively the patient can stand upright only by flexing her hips and knees. (**b**) The preoperative lateral radiograph was taken with the patient's hips and knees straight. Note significant positive sagittal balance. (**c**) Three months postoperatively there is marked improvement of the sagittal balance clinically. (**d**) Improvement of sagittal balance radiographically 3 months after operation

only very limited soft tissue damage and does not lead to additional destabilisation.

Reduction of the slipped vertebra is technically possible [9, 12, 17, 24, 77, 84, 88, 93, 102, 108, 110, 113]. It includes a considerable risk of neurologic complications. It is recommended by several authorities [20, 74].

The question is, whether reduction is necessary. There are no prospective randomised controlled trials available comparing in situ fusion with reduction and fusion. Six retrospective comparative studies failed to show any measurable benefit from reduction in clinical outcome [17, 84, 88, 102, 103, 137]. The numerous publications on high-grade slip reduction do show that patient's outcome will be good if solid fusion is achieved and no complications occur. Partial reduction is less dangerous than full reduction. But not a single study so far was able to prove that the reduction itself, i.e. the improvement of the position of the slipped vertebra in terms of slip percentage and/or lumbosacral kyphosis, is in anyway related positively to the outcome concerning pain or function. The very satisfactory results of in situ fusion in young patients are mainly due to stabilisation. Additionally, there is a significant capacity for remodellation [96]. Possibly, we underestimate the adaptive capability of the growing spine. Lubicky, in his comment on this topic, stated: "Are we so stubborn and arrogant that we cannot accept the possibility that we feel has to be better on anatomic improvement is in fact not better when viewed from the patients' eyes? Or are we right and the outcomes instruments just cannot demonstrate it? Who knows?"[69].

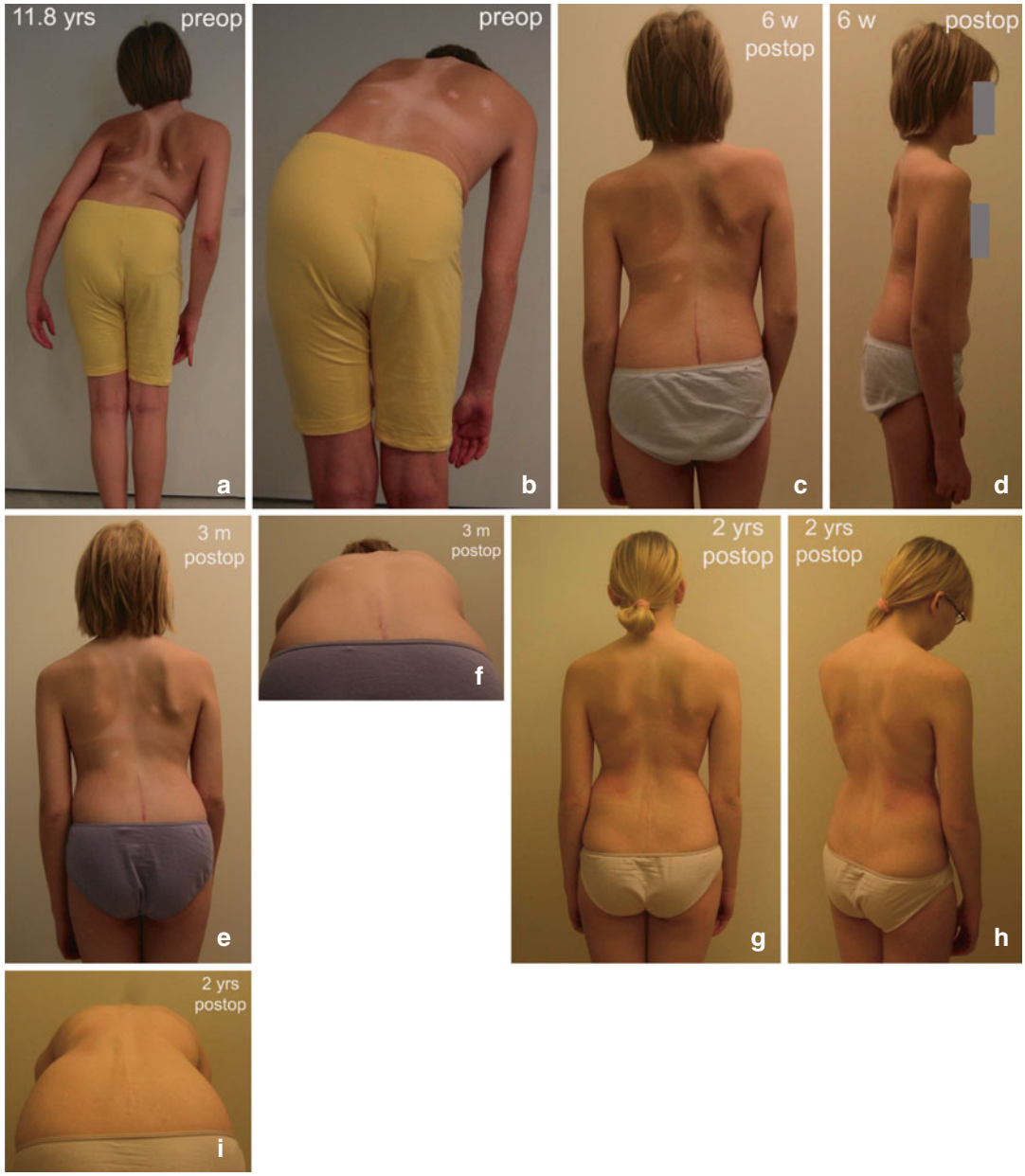

Fig. 24.13 An 11-year-old girl with a painful high-grade L5 slip, severe balance problems and a significant secondary scoliosis. Clinical radiographs, preoperative to 2 years postoperative follow-up (**a–i**). The patient was treated by uninstrumented anterior-posterior in situ fusion without decompression. Plain radiographs, preoperative to 4.4 years postoperative follow-up (**j–r**). Note rapid preoperative deterioration within 4 months. Because of considerable residual scoliosis 6 weeks after surgery, a Boston-brace was applied. The brace treatment was stopped after 13.5 months when overcorrection of the curve was observed (**p**). Very satisfactory clinical (**g–i**) and radiographic (**q**, **r**) outcome

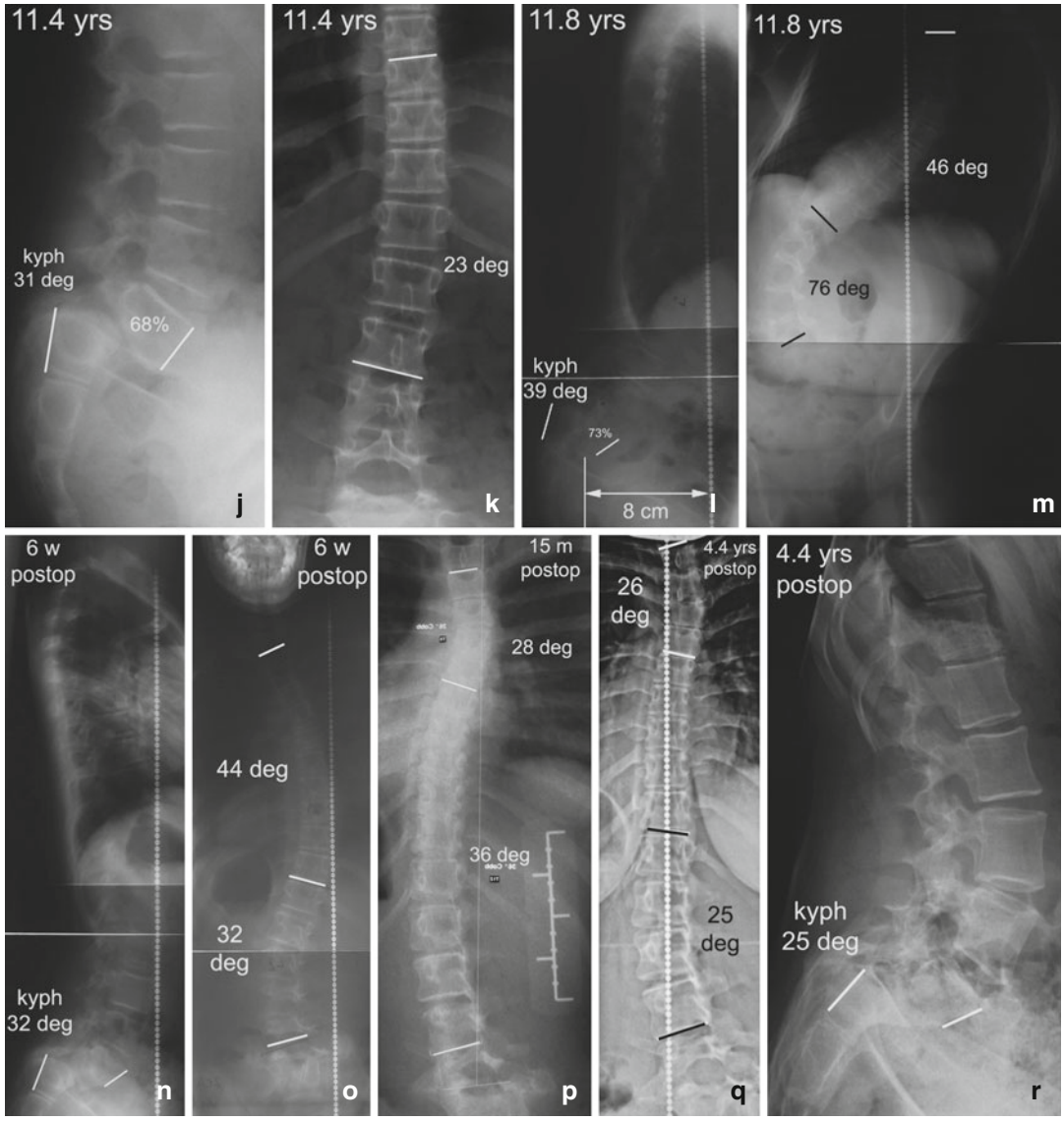

Fig. 24.13 (continued)

At this stage of knowledge, the author considers active instrumented slip reduction in children and adolescents only when the slip is not reducible in the supine-hyperextension radiograph sufficiently to allow for adequate anterior fusion. The aim of the (partial) reduction in such rare cases is to improve the position of the slipped vertebra to facilitate successful anterior interbody fusion. Reduction should then always be combined with decompression to allow visual control of the nerve roots during the manoeuvre.

24.4.3.3 Spondyloptosis (>100 %)

Spondyloptosis is a very special and rare situation. It requires thorough investigation and serious consideration of individual solutions as there is not a single approach. There are different degrees of ptosis: The vertebra may be just "fallen off" the sacrum and is still very mobile. However, it may be also situated very caudally in front of S2 and appear almost unmovable. The clinical presentation is variable, too. It reaches from very mild symptoms to severe neurologic impairment.

A traction radiograph and a hyperextension radiograph in supine position should be taken to demonstrate the mobility of the slipped vertebra. MRI shows the configuration of the spinal canal, the cauda equina, and the exiting nerve roots.

If there is no neurologic impairment, one can perform a posterolateral in situ fusion from L3 to S1 and add a transsacral strut graft according to Bohlman [10, 35]. This is effective if solid fusion is achieved. It does, however, not improve the cosmetic aspect very much. If the vertebra appears to be mobile enough one can reduce it preoperatively by halo-femoral traction or intraoperatively with pedicle screw instrumentation depending on the severity. Decompression should be always performed in these cases. Combined fusion ensures a stable result (Fig. 24.14).

Another option is the resection of the slipped vertebra and instrumented fusion of L4 onto the sacrum as proposed for spondyloptosis by Gaines [31, 32, 63]. This seems to be the *ultima ratio* if it is not possible to get the vertebra by means of reduction safely into a satisfactory position for fusion (Fig. 24.15). These complex procedures, however, should be performed only by experienced spine surgeons familiar with the special pathoanatomical features of the deformity.

24.4.4 Spondylolisthesis in Very Young Children

As mentioned in the introduction, the literature on spondylolisthesis in children up to 12 years of age is sparse. Publications on children of preschool age (under 7 years) including a greater number of individuals are extremely rare. Several case reports are available [4, 11, 26, 52, 55, 61, 70, 79, 98, 119, 143, 144, 151].

King analysed lumbar radiographs of 500 (250 girls, 250 boys) normal first-grade school children who were between 5.5 and 6.5 years old [52]. Twenty-two (4.4 %) of them had spondylolisthesis, 9 out of 250 girls and 13 out of 250 boys. In "several children", a slip up to 75 % was present. There was no history of trauma. None of the children had any back symptoms. At reexamination after 6–10 years of follow-up, four additional cases were detected. All the children were still asymptomatic. Radiographs from some of the parents and siblings of the children with spondylolisthesis were obtained, too. The incidence of spondylolisthesis in those siblings was up to 69 %. The author stressed that this finding supports the opinion that spondylolisthesis is hereditary. He also stated that in many children

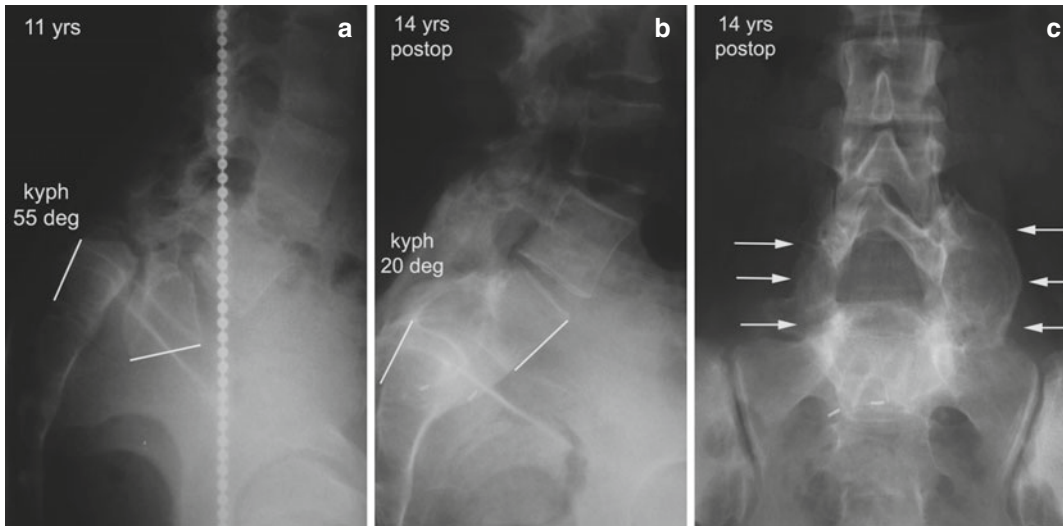

Fig. 24.14 Bilateral L5 laminectomy, instrumented reduction and anterior/posterolateral fusion L4-S1 for spondyloptosis. (**a**) Standing lateral radiograph of an 11-year-old girl with L5 spondyloptosis. (**b**) Lateral radiograph 14 years postoperatively shows satisfactory alignment and healed anterior fusion. (**c**) Solid posterolateral fusion (*arrows*) is seen on the AP radiograph

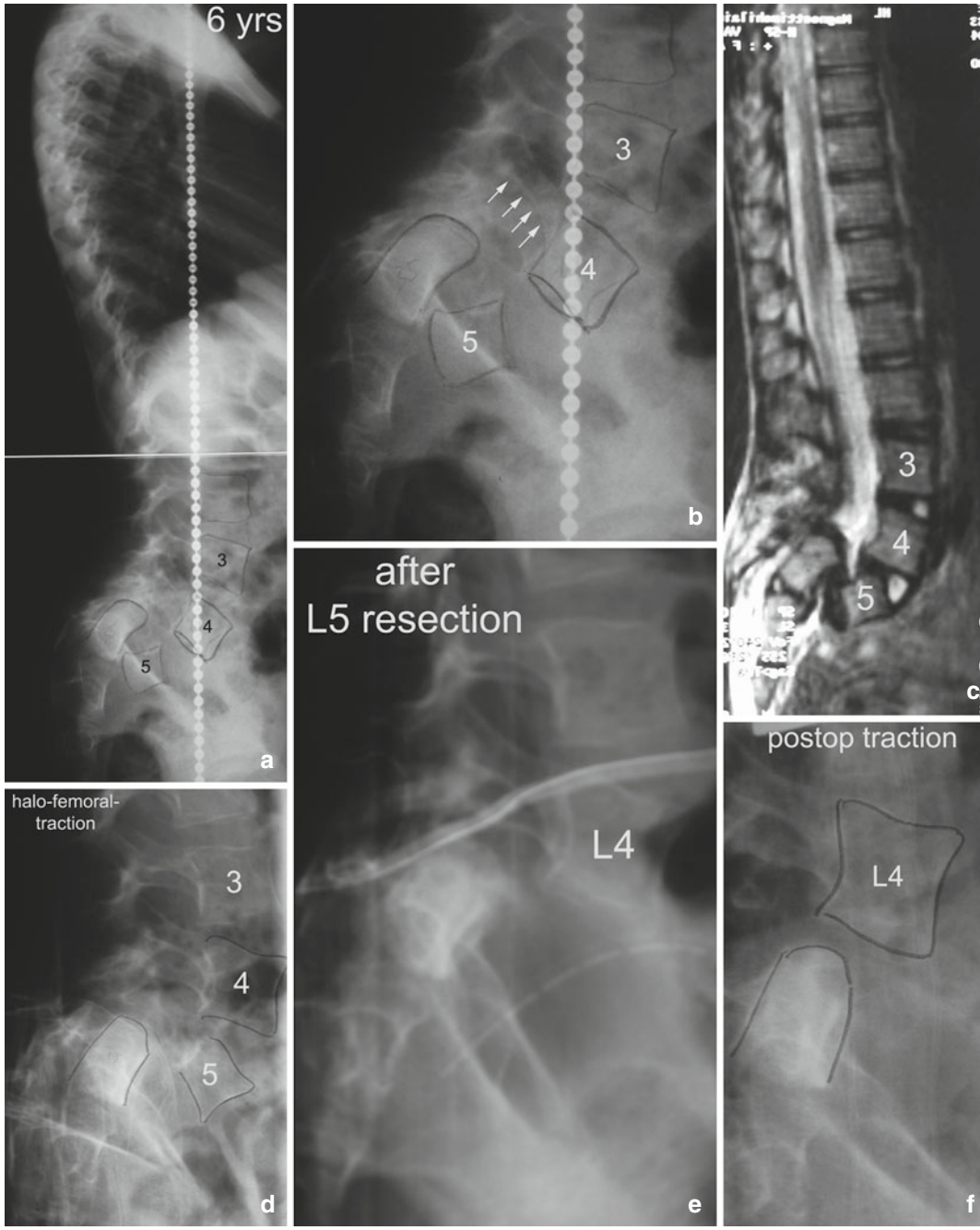

Fig. 24.15 Resection of L5 vertebra (Gaines' procedure) in a 6-year-old male with osteogenesis imperfecta and spondyloptosis. (**a**) Preoperative standing lateral radiograph shows L5 spondyloptosis but acceptable sagittal balance. (**b**) Close-up of the preoperative lateral radiograph, note elongated pedicles (*arrows*). (**c**) Preoperative midsagittal MRI shows L5 on the level S1/S2 and total obstruction of the spinal canal. The patient did not have neurological symptoms. (**d**) Lateral radiograph in halo-femoral traction shows partial reduction of L5. Traction had to be stopped because the patient developed bilateral peroneal muscle weakness. (**e**) Lateral radiograph after L5 resection. The lower endplate of L4 is just above S1. (**f**) Lateral radiograph after postoperative halo-femoral traction shows reduction of L4 on S1. (**g**) Lateral radiograph 2 years after anterior spondylodesis using an autologous fibular inlay-graft shows solid fusion and satisfactory position of L4. (**h**) Lateral radiograph 5 years postoperatively shows severe impairment of the sagittal profile due to bending of the sacrum at the level S1–S2. The patient is neurologically intact and free of pain

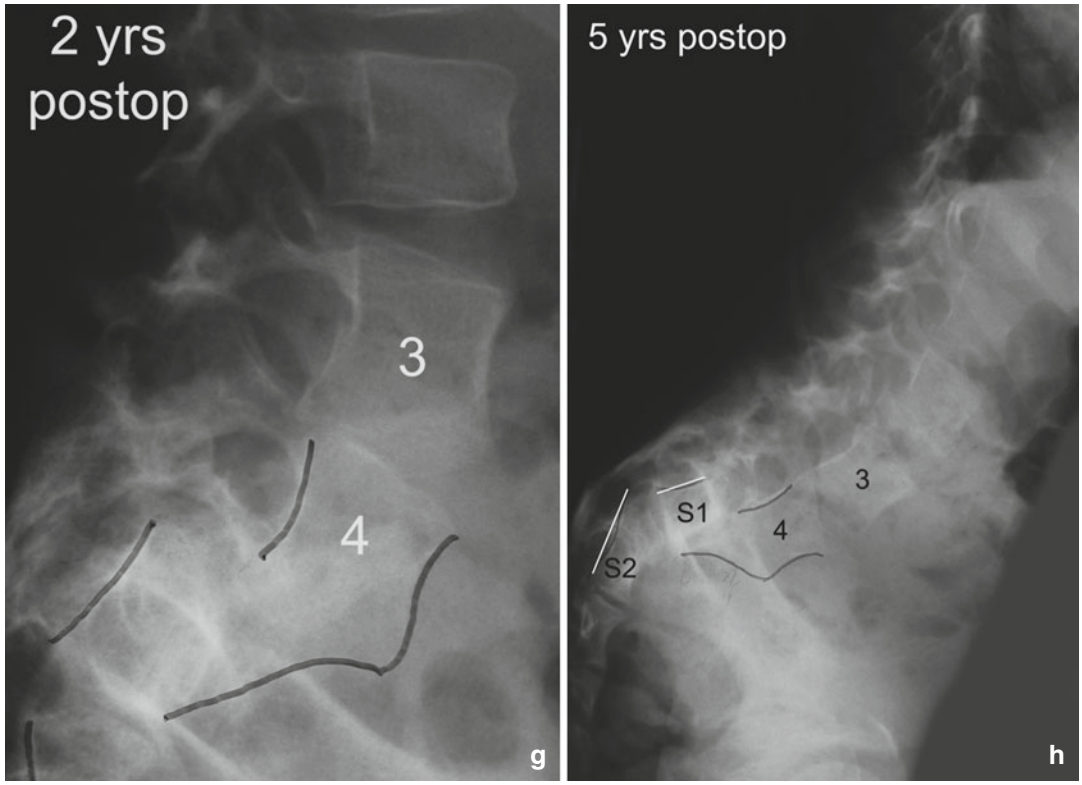

Fig. 24.15 (continued)

the slip occurred mainly before 6 years of age. However, one has to note that the follow-up in this study was not long enough to exclude slip progression during the adolescent growth spurt in all children.

Zippel and Abesser obtained AP- and lateral lumbar radiographs from 530 children of a paediatric orthopaedic outpatient clinic [151]. All these children were free of any low-back symptoms. Their age ranged from 1 to 10 years. If the isthmus was not seen clearly in the primary radiograph, additional oblique films were taken. Low-grade spondylolisthesis was found in 3 of 293 children (1 %) in the age group 1–6 years of age and in 7 of 237 children (3 %) in the age group 7–10 years. In the younger age group, all slips were at L5. One child in the elder age group had undergone operative treatment for myelomeningocele earlier. There were no follow-up data available of this series.

Pfeil took lumbar radiographs from 500 normal children up to 6 years of age [98]. Bilateral spondylolysis and low-grade spondylolisthesis were found in nine children (1.8 %). The youngest child was a male of 1.5 year. There were further four males of 3 years, one of 4 years and two of 5 years of age. The only girl in this group was 6 years old. The mother of one of the 3-year-old males had a spondyloptosis. None of the nine children had any subjective symptoms.

Beguiristáin and Diaz-de-Rada presented a series of eight pre-school children with a mean age of 3.5 years (range, 9 months to 5 years) out of 188 spondylolisthesis patients younger than 20 years of age at diagnosis [4]. All patients had slips at L5. There was some confusion in the publication concerning the classification of the cases. According to the text, four slips were isthmic, two were dysplastic, one was traumatic and one was iatrogenic. In Table 24.1 of the same paper, three cases are categorised as being isthmic, three

dysplastic, one traumatic and one iatrogenic. Two slips were high-grade, one dysplastic 87 % and one traumatic 57 % slip. In the remaining four, the slip ranged from 18 to 48 %. The two patients with the iatrogenic and the traumatic slip resp. had pain on admission. In the other six cases the reason for investigation was scoliosis in four, kyphosis or skin alteration in one each. The mean follow-up was 11.5 years (range, 9–14 years). Three patients were treated operatively. A 4-year-old patient was operated at the time of diagnosis because of high-grade slip (87 %). In situ arthrodesis was performed. The technique (levels? approach?) was not described in detail. At 11 years postoperatively, the patient was asymptomatic, the slip measured 95 %. The youngest patient was a girl with a 15 % slip was diagnosed at 9 months of age due to a skin alteration. She was operated at the age of 11 years because of progression to 53 %. Posterior fixation with hooks and rods from L3 to the sacrum was performed. The patient developed a pseudarthrosis and was reoperated successfully 9 months later with pedicle screw fixation from L4 to S1. After 2-year follow-up, the patient was free of symptoms. No further progression of the slip occurred. The third surgically treated patient was a female with a 25 % slip at 3 years of age. Progression of the slip to 50 % was detected at 11 years. Combined fusion L5 to S1 with pedicle screw instrumentation was performed. Three years after the operation, she was asymptomatic. The slip was 28 %. Three non-surgically treated patients had slips from 18 to 47 %. To one of them, a 3-year-old female with 47 % of slip, restriction of sports activities was recommended. The remaining two patients were allowed to live without any restrictions. No braces were used. They were followed until maturity, and no slip progression occurred. The 5-year-old male with the lumbosacral luxation-fracture (so-called traumatic spondylolisthesis, slip 57 %) was treated by reduction under general anaesthesia and plaster immobilisation for 3 months. He had a complete unilateral motor deficit and impaired sensibility at L5 and a partial S1 motor deficit at admission. Neurologic symptoms resolved. He was asymptomatic at 14-year follow-up with a slip of 15 %. The patient with the iatrogenic slip of 25 % had undergone tumour surgery at the age of 4 years. He was followed for 14 years. He remained symptom free, and there was no progression of the slip.

Out of a clinical series of 63 spondylolisthesis patients aged 0–19 years, McKee et al. found 28 children under the age of 10 years (7 children in the age group 16 months to 4 years, 21 children in the age group 5–9 years) [79]. The gender distribution was equal. Out of these 28 children under age 10 years, 18 children were symptom free. The ten symptomatic children had lumbar pain, none had radiating pain. Three had spondylolysis without any slip, 24 had a low-grade slip, and 1 child had a high-grade slip. Out of the whole patient population (63 patients), only 5 patients required surgery. However, it is not known if any operated child belonged to the group under the age of 10 years as the ages of the operated patients are not presented in the article. In their discussion, the authors stress that usually children under the age of 10 years do not have radiating pain which seems to be present more in elder children and adolescents.

In the study by Seitsalo, of the 5 children less than 7 years of age, four were asymptomatic at diagnosis. After postoperative mean follow-up of 14.5 years, no difference in outcome after fusion was detected if compared with children over 7 years of age [119].

The youngest patient in the literature was described by Borkow and Kleiger [11]. In this male, a kyphotic deformity of the lumbosacral region was detected after birth. At the age of 15 weeks, a lateral radiograph of the lower lumbar spine and the sacrum revealed a kyphotic displacement of L4. The child developed normally, but its gait was described as “wide-based shuffling”. The hip radiographs were normal. At the age of 11 years a lax right patella was stated. One year later, recurrent dislocations of the patella started. Although no back problems were reported nor any slip progression occurred, posterolateral fusion from L3 to the sacrum was performed at the age of 13.5 years. Fusion was successful. And after short-term follow-up the patient was free of symptoms. Looking at the radiographs in the publication, one gets the impression that this was

a congenital lumbosacral kyphosis due to an anterior failure of formation of the L5 vertebral body. In fact, the authors also discussed this possibility in their article, but they preferred the explanation that a real slip had happened in utero.

A patient with a unilateral spondylolysis and 4-mm slip of L5 at the age of 10 months was reported primarily by Laurent and Einola [61]. When she was re-examined at 10 years of age, the lysis and the slip were still visible. At final follow-up, 25 years after the first presentation, the isthmus was healed and there was no slip anymore. The patient was free of any complaints and played volleyball actively [120].

Wild et al., in a case report, described an 18-month-old male with L5 spondyloptosis and spina bifida occulta L4 to S1 [144]. The condition was deemed congenital although there is no proof that the deformity has been present already at birth. The boy had episodes of recurrent falls and inability to stand and walk lasting no longer than 30 min. The history was otherwise uneventful. There were no objective neurologic findings. At the age of 5 years, the child was hyperactive and demonstrated a waddling gait and lower limb muscle hypoplasia. There was L4/L5-weakness, a mild foot deformity and ileopsoas contracture which resolved after stretching. The degree of vertebral slip was unchanged. A three-stage procedure (back-front-back) was performed: bilateral L5 laminectomy and root decompression, partial resection of the L5 vertebral body, reduction of L5, interbody fusion L5 to S1, and posterior fixation from L2 to the sacrum. Nine months later the instrumentation was removed and posterolateral fusion L5 to S1 was added. Nine years after the operation, the patient had a normal alignment of the lumbosacral spine and was practising sports actively.

Wertzberger and Peterson published a case report of a girl developing a spondylolysis and a low-grade slip at the age of 18 months during an observational period after repeated radiotherapy and chemotherapy for histiocytosis X [143]. She was free of back symptoms. During a 2-year follow-up the radiographic picture did not change. Nor did the patient develop symptoms from her spine. No treatment was applied.

Kleinberg described a case of a 17-month-old female who was admitted for treatment of a congenital dislocation of the hip [55]. The back was clinically normal. The child had been walking for a few weeks. No trauma was known. On routine radiographs a forward slip of L5 of more than 50 % of its length was seen. A "gap or cleft" in the pedicle visible on the lateral radiograph was interpreted as being the inborn cause for the slip. Therefore, the case was claimed to represent a true congenital spondylolisthesis.

Finnegan and Chung reported a case of a very special L5 in a 3-year-old female [26]. The child was born small-for-age at gestation week 37 and developed erythroblastosis fetalis. The mother detected a lump in the back of the child when it started to walk at the age of 1 year. No trauma was reported. A doctor was consulted because of out-toeing at the age of three. The child exhibited a "wide-based shuffling gait" with stiff knees. The muscle tonus was slightly increased in the lower extremities, as were the tendon reflexes. But there was no hamstring tightness. The skin sensation in the perineal area was decreased. The plain radiograph showed a spondyloptosis of L5 being situated in front of S1. At the same time L5 was also displaced anteriorly in relation to L4, i.e. L5 was slipped forward out of the vertebral row. Due to that the posterior inferior corner of the L4 vertebral body was riding on top of the dome-shaped sacrum. This is extraordinary as usually the whole upper vertebral column is moving forward with the slipping vertebra. Myelography demonstrated a filling defect at the slip level. Decompression was performed by unilateral laminectomy L3 to L5 and excision of the posterior part of the L5 vertebral body. A posterior and a posterolateral fusion from L3 to L5 were added. The outcome was favourable after 3 months of follow-up. Concerning the aetiology, the authors did not believe that it was possible to determine the cause of the slip exactly. They questioned if this was a usual spondylolytic slip and favoured the explanation that this was possibly a "complete congenital spondylolisthesis".

The vast majority of very young children with a vertebral slip seem to be free of pain. An exceptional case of low-back pain symptoms in a very

young patient with spondylolisthesis was published by Lucey and Gross [70]. They encountered a female of 2 years and 8 months who had an uneventful history until she started to complain about back pain at the age of 2 years. The pain was not related to activities nor was there any trauma. On physical examination, the spine looked normal. There was no tenderness but a pilonidal cyst at the lower back. Neurology and her gait were normal. The popliteal angle was 20° bilaterally. On the primary radiograph, there was a "unilateral (?) Grade 1 spondylolisthesis" of L5. Two months later, the slip had progressed to 27 %. MRI was normal. An antilordotic cast (later a brace) was applied. The girl became pain free. No healing of the pars defect was seen during a period of 8 months.

In conclusion, children of pre-school age with spondylolisthesis may have low-back pain, but the majority seems to be pain free. Often, the lysis or the slip is diagnosed incidentally during investigations for other diseases. Some cases are detected because of posture anomalies, scoliosis or gait problems. Slip progression does not appear to be a common phenomenon during this age period. Despite that, regular radiographic follow-up is advisable. If operation is deemed to be necessary uninstrumented posterolateral fusion is the method of choice.

24.4.5 Exotic Spondylolisthesis

The term exotic spondylolisthesis was coined by Lubicky for unusual cases with vertebral slips due to bone or soft tissue anomalies often related to syndromes [68]. There can be developmental disturbances in the facet joints or elongation of the pars interarticularis or the pedicles due to poor bone quality and/or spina bifida. Pathologic increased soft tissue laxity may also be a contributing factor.

24.4.5.1 Osteogenesis Imperfecta

Some cases of spondylolisthesis in osteogenesis imperfecta have been published in the literature [3, 53, 68, 104]. Usually elongation of the pedicles is seen in those cases. Assessment and diagnosis is often difficult as a significant other spine deformity may be present. No specific recommendations for treatment are existing. In asymptomatic non-ambulators, operative treatment is probably not indicated. In walking children with symptoms it seems reasonable to stabilise the segment with combined fusion.

King and Bobechko found cases of spondylolisthesis due to elongated pedicles among 60 patients with osteogenesis imperfecta [53]. Three of the four also had scoliosis. No treatment for spondylolisthesis was applied. The age of the patients was not mentioned.

Rask published a case report of a 40-year-old male with osteogenesis imperfecta who has had a back injury at 4 years of age [104]. Back pain persisted for many years following the injury. At the time of presentation, a low-grade slip of L5 was found. It is assumed that he had sustained a pars interarticularis fracture during that childhood injury. No treatment was necessary.

Basu et al. reported on two cases [3]. A 10-year-old female had elongated pedicles of L4 and L5 causing a low-grade slip on both levels. A successful uninstrumented anterior L3 to S1 fusion was performed at 11 years of age. Later, a posterior instrumentation and fusion from T1 to L1 was carried out for thoracic scoliosis. At 3-year follow-up she had no back problems. The second patient was an 11-year-old female suffering from low-back and coccygeal pain since 1 year. She had elongated lumbar pedicles and a high-grade slip of L5. In addition, a lumbar hyperlordosis and thoracic scoliosis were present. She was developing also leg pain. In situ fusion was planned.

At the author's institution, one case of spondyloptosis in a 6-year-old boy with osteogenesis imperfecta was treated by resection of L5 and fusion of L4 to the sacrum. Fusion was successful and the patient is still clinically free of symptoms. No neurologic complications occurred. However, obviously due to the poor bone quality, the sacrum started to bend into kyphosis causing significant cosmetic impairment (Fig. 24.15). This example shows that especially in young patients with systemic disease long follow-up is mandatory to ensure the lasting benefit of an operative procedure.

24.4.5.2 Neurofibromatosis

A few cases of spondylolisthesis in neurofibromatosis type I have been described [68]. As in osteogenesis imperfecta, they are often accompanied by other severe spinal deformity. Treatment should follow common rules. In operative treatment the poor bone quality should be taken in consideration. Therefore, combined fusion is advisable if operative treatment is deemed to be necessary.

McCarroll analysed radiographs of 46 patients with neurofibromatosis [78]. Four of them had spondylolisthesis. Two were accidental findings; the other two had low-back pain. The age of the patients is not reported. No details are given concerning follow-up or treatment.

Crawford published a series of 82 patients with neurofibromatosis [21]. Fifty of them had spinal deformities. One out of the 50 had a vertebral slip. The case was not described in detail. In a later publication including another 34 patients no additional cases of spondylolisthesis were found [22]. The authors conclude that spondylolisthesis is not as common in neurofibromatosis patients as compared to the general population.

24.4.5.3 Marfan's Syndrome

Spondylolisthesis has been reported also in connection with Marfan's syndrome [56, 135, 150].

Sponseller et al. investigated two different groups of patients with Marfan's syndrome [129]. Among 82 skeletally mature patients, 5 (6 %) had a low-grade slip, the mean slip being 30 % (a radiologic follow-up of more than 2 years), three had spondylolisthesis, one low-grade and two high-grade (mean 60 %). No information is provided on slip progression, symptoms or treatment. The authors hypothesise that the frequency of spondylolisthesis in Marfan's may not be higher than in the general population. But the degree of slip is greater probably due to the altered ligament properties and shear resistance of the disc.

Winter presented a report on two operated cases of spondyloptosis with Marfan's syndrome: a 13-year-old and a 16-year-old male [150]. Both patients showed the typical posture of a high-grade slip. Both had back pain and leg pain. Their hamstrings were tight. There were no neurologic findings, nor bladder dysfunction. One was treated by decompression and uninstrumented posterolateral fusion from L3 to S1. At 4 years of follow-up, the patient had no complaints and the fusion was sound. The other patient had preoperative halo-femoral traction for 2 weeks, bilateral laminectomy L4 and L5, posterolateral fusion L3 to S1, postoperative traction for 3 weeks more, and subsequent anterior fusion L5-S1. This was followed by 4 months of bed rest in a plaster cast and after that a body cast with one leg included for further 3 months. At follow-up after 2 years the patient was pain free and had normal neurology. The fusion was solid.

Taylor published a case of an 11-year-old female with Marfan's and a high-grade L5 slip and a 70° c-shaped lordoscoliosis [135]. She had pain in her left buttock, leg and foot. Hamstrings were tight, and ankle jerks were absent. L5 laminectomy and posterolateral fusion from L4 to S1 was performed. She was mobilised in a plaster spica after 2 weeks. The fusion healed within 6 months. The patient was free of symptoms. Further follow-up of the spondylolisthesis was not presented. The scoliosis progressed and was operated on 14 months later.

24.4.5.4 Ehlers-Danlos Syndrome

Spondylolisthesis in Ehlers-Danlos syndrome was reported by Nematbakhsh and Crawford [89]. A 2-year-old female was evaluated for transient paraparesis of the upper extremities. An unstable slip of C2 was found and treated by fusion from C1 to C3. Due to subsequent instability of the adjacent segment the fusion had to be extended to C5. At 4 years of age, she started having back pain, and at years of age radiographs revealed a low-grade L5 spondylolisthesis with pars defects. At that time she was diagnosed having Ehlers-Danlos Type VI. The slip progressed during 4 years to 75 %. A successful uninstrumented posterolateral fusion L4 to S1 was performed when she was 13 years old. No complications occurred. The long-term outcome was not reported. Lubicki reported having treated at least one other patient with Ehlers-Danlos and spondylolisthesis [68]. He underlines the risks

related to poor bone quality and wound healing and recommends treating them in a similar way as Marfan patients.

24.4.5.5 Myelomeningocele

In patients with myelomeningocele, spondylolisthesis should be very common due to the developmental disturbance of posterior vertebral elements. The incidence reported ranges from 5.9 to 28.6 % [50]. In the study of Mardjetko et al., ambulatory patients with spina bifida had an incidence of spondylolisthesis twice as high as non-ambulatory patients [75]. All slips were low-grade. During 5 years of follow-up no progression was seen. There was no correlation between the level of the spina bifida and the degree of slip. According to Lubicky, two patients of that series had been operated on, one because of pain, the other for tethered cord release. Operative stabilisation is advised for pain, progressive deformity or neurologic impairment [68].

Stanitski at el performed a radiographic evaluation of 305 patients aged 7–22 years with myelomeningocele [130]. They found L5 spondylolisthesis in 18 (5.9 %) patients. The majority (75 %) of them were between 7 and 14 years of age. All affected patients were walkers. The mean slip was 37 %, ranging from 12 to 56 %. Fourteen were low-grade and four high-grade slips. Patients with spondylolisthesis had a greater lumbar lordosis than patients without a slip. Furthermore, lumbar lordosis was positively related to the slip percentage. During a follow-up from 2 to 7 (mean 2.5) years, no slip progression was detected. None of the patients had symptoms from the spondylolisthesis. No comments concerning treatment are given in that paper.

Overall, spondylolisthesis in various syndromes represents a very mixed conglomerate of problems. As a general recommendation, the author agrees fully with Lubicky who wrote that one "will need to use common sense and well-accepted general principles when encountering such clinical problems" [68]. Long-term follow-up is to be recommended in operated as well as in non-operated patients to ensure a favourable final outcome.

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Management of Spine Tumors in Young Children

25

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Key Points

1. Tumors are relatively rare in the pediatric spinal column but still constitute a substantial portion of pediatric spinal disorders.
2. Presentation may be very unspecific with vague symptoms; a high level of suspicion is required.
3. Treatment guidelines are not particularly different compared to primary tumors in appendicular skeleton, or those of adults.
4. Surgery is the treatment of choice in most of pediatric spine tumors, and should not be withheld on the basis of potential complications.

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25.1 Introduction

Vertebral neoplasms in the pediatric age are uncommon and feature a significant clinical challenge for the surgeon involved in their diagnosis and treatment. Primary malignant tumors like osteogenic sarcoma (OGS) and Ewing's sarcoma (ES) are less frequent than benign tumors like osteoid osteoma (OO), osteoblastoma (OBL), aneurysmal bone cyst (ABC), and eosinophlic granuloma (EG). Other conditions like hemangioma (HE) are rarely found before the maturity. Metastases in the pediatric spine can develop in

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the clinical course of Wilms tumor and neuroblastoma.

As in the adults spinal tumors are difficult to diagnose, it is important to know their clinical and imaging pattern in order to suspect the diagnosis: It is quite remarkable that the spine tumors occurring in the pediatric spine have some strongly suggestive peculiarities.

As frequently in this age group the onset symptoms of a spine tumor are not specific, a persistent back pain, notably if independent by activity and increasing during the night, should be seriously considered and the suspect of a tumor should be ruled out.

Plain radiographs are inadequate to detect small lesions, but the progress of imaging techniques in the last 20 years has improved the possibility of early diagnosis: isotope scan, positron emission tomography (PET)-scan, computerized tomography (CT)-scan, and magnetic resonance imaging (MRI) can combine a series of data in many cases suggestive for a lesion or a for a restrict number of possible diagnosis. Technetium one scans are very helpful in detecting small bony lesions as a cause of otherwise unknown pain. PET scan is able to find out – from the entire body – areas of pathologic metabolism: Its role is increasing in detecting and staging of bone lesions.

Angiography today is abandoned as a diagnostic tool as MRI is less invasive and exceeds it in pointing out the vascular pattern of the lesion. Conversely the role of selective arterial embolization (SAE) is growing as adjuvant to intralesional surgery of highly vascularized lesions and as curative technique in the treatment of ABC.

Treatment includes surgical and non-surgical options: Combination of both is frequently considered in the perspective to enhance positive effects and reduce the morbidity. This strategy obviously requires involving the activity of a multidisciplinary team.

As in the adults, planning surgical treatment includes: the selection of an oncologically appropriate surgical target as suggested by diagnosis and oncological staging and to design consequently a surgical technique aiming to preserve function and to maintain alignment, stability, and motion.

In the pediatric age the peculiarity of a growing spine must also be considered and the surgical techniques must be modified accordingly.

Non-surgical options like radiotherapy and chemotherapy are included in the protocols of malignant tumors. Radiotherapy is limited by the risk of interfering with growth processes, resulting in asymmetrical or symmetrical deformities, possible damage to the spinal cord, and post-irradiation sarcoma. Selective arterial embolization and direct injection of steroids or stem cells have been recently suggested in specific cases.

25.2 Evaluation

25.2.1 Clinical Presentation

The most common symptom is pain, which has been reported to be the presenting problem in 46–83 % of patients [1–3]. Persistent back pain should alert the physician for an underlying pathological process. The characterization of pain in spinal tumors may be progressive pain, predominant night pain, or unrelenting [4, 5]. Severe pain is usually associated with microfractures induced by the rapid growth of neoplasms. In some patients palpation of the affected segments may induce pain. Only in case of OO the pain feature can be pathognomonic.

Another possible finding is neurological involvement. Several studies report incidences of 54 and 67 % for motor weakness and neurological involvement in pediatric patients with spinal neoplasms [1, 5]. However, neurological involvement is most commonly associated with malignant bone tumors, as radicular symptoms and myelopathy may arise from the involvement of neural foramina and spinal canal concordant with the level of involvement. As most of tumors in the pediatric spine are benign, a detailed history and physical examination is essential.

Children with spinal tumors may present with spinal deformity, especially in cases of OO and OBL [1, 2, 6–9]. Scoliosis associated to these tumors is quite peculiar, possibly associated to reactive muscular spasm; the rotational component is minimal. It has been reported to be the

presenting symptom in 27–63 % of these patients [9, 10]. Other tumors such as Langerhans cell histiocytosis or eosinophilic granuloma (EG) may also cause subsequent deformity (scoliosis and kyphosis) due to destruction of the vertebral body and collapse.

25.2.2 Imaging Studies

The evaluation of a patient with suspected spinal tumor should start with high-quality AP and lateral radiographs. In cases with stigmata which may be associated with tumors such as atypical spinal deformity, masses, bony destruction, vertebral collapse, widened interpedicular distances, erosion of pedicles ("winking owl" sign), sclerosis, enlarged neural foramina, and scalloping of vertebral bodies, advanced imaging studies must be performed. The sensitivity of plain radiographs for detection of spinal tumors varies between 55 and 98 % [1, 11]. CT, MRI, and technetium bone scans are the most commonly used advanced imaging studies. CT is well tolerated as it is noninvasive and usually fast and very efficient in the demonstration of bony lesions such as erosive masses, bone destruction, sclerosis with central radiolucency (nidus), periosteal reaction, and widened spinal canal. On the other hand, CT scan has disadvantages of an increased exposure to radiation as well as not being very sensitive in the demonstration of soft tissue lesions. MRI appears to be the most sensitive of all imaging studies and should be the first choice, especially in the presence of neurological involvement. It may show the bony and soft tissue masses, bone destruction, lesions extending to or originating from surrounding soft tissues, as well as the spinal canal and neural elements. Furthermore, in patients with malignant neoplasms, MRI may be useful to monitor the response to chemotherapy and/or radiotherapy. Along with gadolinium enhancement, MRI is the most sensitive and specific measure to detect a metastatic disease. The major disadvantage of MRI is the need for sedation or general anesthesia for younger children. Technetium bone scans are very useful for patients with vague symptoms for whom the presence or location of a problem cannot be ascertained with other studies.

Spinal angiography is obsolete for diagnostic purpose, as MRI is less invasive and equally appropriate to evaluate tumor vascularity, but is frequently performed for preoperative selective embolization to reduce intraoperative bleeding and facilitate surgical resection.

PET scan can be performed if multiple tumor localization or metastases are suspected. Moreover Standardized uptake value (SUV) Maximum (Max) evaluation can be useful to differentiate between tumors and infections.

25.3 Staging

25.3.1 Oncological Staging

Oncological staging defines the biological behavior of neoplasms. The most commonly used for primary musculoskeletal tumors is the Surgical Staging System (SSS) introduced by Enneking and coworkers, which will form the basis of staging in this manuscript so forth [12]. In this staging system, benign tumors are evaluated in three stages (latent, active, and aggressive), whereas primary malignant tumors are divided into two stages of localized disease with two sub stages each and a third stage for metastatic tumors (Table 25.1). This staging was originally described for long bone tumors but proved to be applicable to the primary tumors of the spinal column as well, as demonstrated by several studies [13–15]. These studies also find it very useful for incorporating all the knowledge acquired by clinical and radiological investigations and forming a simple and understandable basis for communication between health professionals. On the other hand, it has to be mentioned that this or any of the other staging systems have not been evaluated for their accuracy in the estimation of the oncological prognosis of tumors located in the spinal column.

25.3.1.1 Benign Tumors

Benign tumors are divided into three stages. First stage tumors (S1) are "latent" tumors which are

Table 25.1 Surgical Staging System (GTM) of primary musculoskeletal tumors

Benign:			
S1: Latent tumors	(G0 T0 M0)		
S2: Active tumors	(G0 T0 M0)		
S3: Aggressive tumors	(G0-1 T0-1 M0-1)		
Malignant:			
Stage I	Low grade	A: Intracompartmental	(G1 T1 M0)
	Low grade	B: Extracompartmental	(G1 T2 M0)
Stage II	High grade	A: Intracompartmental	(G2 T1 M0)
	High grade	B: Extracompartmental	(G2 T2 M0)
Stage III	Any Grade	Any T	(G1-2 T1-2 M1)

inactive and usually asymptomatic. Their feature is to be surrounded by a true capsule, expression of latency. Once identified as latent tumors, these do not need to be submitted to a treatment based on oncological principles as they tend to grow very slowly if at all. Intraosseous enchondromas, osteochondromas, hemangiomas, or lipomas (extremely rare) can be staged into this category. Palliative surgery is required for spinal cord compression or spinal instability due to pathologic fractures, while in most cases these tumors must be followed with clinical and imaging observation.

Second stage (S2) "active" benign tumors tend to grow at a certain rate albeit slowly and often become symptomatic. These are surrounded by a thin layer of fibrous capsule and induce the formation of a reactive inflammatory tissue, which may be seen on MRI ("pseudocapsule"). Oncologic treatment of S2 tumors consists of surgical excision. Embolization, cryotherapy, and radiofrequency ablation are other modalities that may be used in conjunction with surgery or alone.

Third stage (S3) benign tumors ("aggressive") are rapidly growing tumors with a very thin or absent capsule. These tumors invade the neighboring compartments and usually have a wide reactive hypervascularized "pseudocapsule." They usually grow to become large enough to be visible on radiographs, technetium bone scans are usually significantly positive, and CT and MRI demonstrates the aggressive nature of the tumor. Treatment consists of surgical excision of adequate aggressiveness along with the help of one or more of the surgical adjuvants delineated above.

25.3.1.2 Malignant Tumors

Malignant tumors are studied based on the concept of "grade" and classified into two groups as low grade and high grade. These are further subdivided into two categories of A and B based on the relation of the tumor with the compartment it has originated from. A is used for the tumors that are still within the compartment at the time of diagnosis, and B for those that have extended beyond that compartment or has originated at a location with no natural boundaries and therefore does not constitute a compartment (e.g., intrapelvic). Based on this, a low-grade stage IA tumor is one that remains inside the vertebra itself and by contrast, a stage IB tumor invades paravertebral compartments. These tumors usually have thick pseudocapsules of reactive tissue and small, microscopic tumor islands within that reactive zone called "satellite nodes." The acceptable resection margin is therefore wide resection if possible. High-grade tumors are likewise divided into stages IIA and IIB. These are very rapidly growing tumors with no reactive tissue; on the other hand they do have not only satellites lesions but also a significant risk of skip metastases (foci of tumor outside the main mass, completely isolated). High-grade tumors are identifiable on plain radiographs, but MRI is needed in addition to show the entire extension of the tumor and the absence of reactive zone. Treatment is wide en bloc excision as radical excision in the spinal column is impossible [16].

25.3.2 Surgical Staging

After the definitive diagnosis and oncological staging has been established, the next step before biopsy should be surgical staging. The most widely used scheme was developed by Weinstein for primary spine tumors and later modified by Boriani and coworkers to become the WBB (Weinstein, Boriani, Biagini) system [17]. This system is useful for defining the local extent of the tumor and therefore eventually dictates the type of resection needed. Vertebra is divided into 12 radiating zones in a clockwise order (from 1 to 12), five concentric layers from paravertebral extraosseous to the intradural region (A to E) (Fig. 25.1), and the longitudinal extension of tumor as expressed by the number of involved spinal segments is added as a separate parameter. The major advantage of this system is that it delineates the relation of the lesion with the spinal cord and therefore intrinsically marks out the amenability of the tumor to wide resection. These authors recommend that in order not to endanger the spinal cord and to control the epidural space, the surgeon should aim to resect wedge sectors of the vertebra [16].

Another classification system is proposed by Tomita and coworkers. This system is composed of a two-part numeric system and incorporates a detonation of tumor location providing a simplified scheme for describing the extent of vertebral involvement. The first numeric part describes the affected anatomic site, comprising 1 vertebral body, 2 pedicles, 3 lamina, transverse and spinous processes, 4 spinal canal, and 5 paravertebral area. The second numeric part describes tumor extension in numbers ranges from 1 to 7. These authors tended to consider type 1, 2, and 3 lesions as intracompartmental, type 4, 5, and 6 lesions as extracompartmental, and a type 7 lesion as a multisegmental tumor or one with multiple-skip lesions [18]. For the sake of simplicity only the WBB system will be used in this text.

25.4 Biopsy

Biopsy is an essential step before planning the treatment. The purpose of this procedure is to provide the pathologist with an amount of pathological tissue both quantitatively and qualitatively representative for diagnosis. Its volume must be adequate for different staining and for immunohistochemical studies. The tissue must not be removed from necrotic, reactive, or fibrotic areas; it must be taken from the core of the vital tumor. Biopsy is by definition an intralesional procedure and therefore includes the high risk of spreading tumor cells in the surrounding tissue,

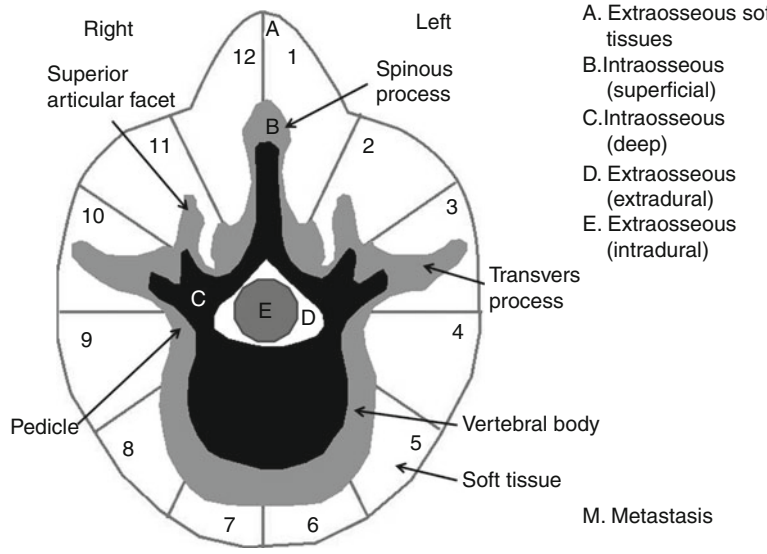

Fig. 25.1 The WBB staging system for primary spine tumors (Adapted and redrawn from the original in Ref. [6])

with the obvious consequence of increasing the risk of local recurrence. The most important surgical principle is to include the biopsy route within the line of incision that will be used at the time of definitive surgery, particularly in case of en bloc resection, whose specimen will necessarily include the entire biopsy tract from the skin to the tumor mass. For this reason, the biopsy approach should never be performed along the anatomical extracompartmental spaces, as performed in elective non-oncologic orthopedic surgeries. The biopsy approach must always be performed inside muscles, in order to make easier the removal of the tract. Following these principles, biopsy can be performed through percutaneous or open techniques [19]. Open biopsy should be performed by the surgeon who will perform the definitive surgery; it may result in substantial blood loss and morbidity, but the surgeon can obtain a relatively large amount of tissue for diagnosis decreasing the likelihood of a sampling error. A particular care should be adopted to control bleeding and avoid hematoma, which is a severe complication of biopsy as the tumor cells can be seeded on a wide area, almost impossible to resect later on.

Percutaneous biopsy (performed by fine core needle or best by a trocar) is a relatively simple procedure and has been proven to be safe and effective when performed under CT-scan image guidance [20]. On the other hand, selection of the optimal biopsy technique depends on the differential diagnosis, the location and extension of the lesion, and the potential definitive treatment plan. Although there is the theoretical possibility of having intraoperative frozen sections, the present authors do not recommend it on a routine basis as in our hands it has been associated with a substantial rate of diagnostic errors, totally unacceptable for tumor surgery in spinal column. Therefore it should be reserved for cases whose imaging is pathognomonic (like some ABC or some OO) or for the confirmation of the adequacy of surgical margins if necessary. Open biopsy should be avoided when a malignant bone tumor is suspected on clinical and imaging studies due to the highest risk local recurrence. Transpedicular image-guided trocar biopsy allow to remove adequate sample from anterior elements without contaminating the thoracic or abdominal cavities; to reduce the risk of seeding from the empty pedicle, it can be filled with acrylic cement [16].

25.5 Surgical Treatment of Spinal Tumors

The goal of the surgery for pediatric spinal primary tumors is to allow the best local and systemic control; conversely, the treatment of metastatic and systemic diseases is mostly palliative, aiming at pain relief, decompression of neural structures, provide correct alignment, stability, and possibly mobility to the spine [21].

Tumor removal in case of metastatic and systemic disease is reasonable if the specific tumor type has low sensitivity to chemotherapy and radiation therapy.

A commonly accepted terminology for surgical procedures and for definition of tumor extent is needed for surgical planning. Lesions in the pediatric spine are often more challenging to treat than lesions of similar behavior elsewhere in the musculoskeletal system. “Curettage” describes the piecemeal removal of the tumor. As such, it is always an intralesional procedure. “En bloc” indicates an attempt to remove the whole tumor in one piece, together with a layer of healthy tissue. The term “intralesional” is appropriate if the surgeon has been within the tumor mass at any time during surgery; “marginal” is appropriate if the surgeon has dissected along the pseudocapsule, the layer of reactive tissue around the tumor; and “wide” is appropriate if ablation could be performed outside the pseudocapsule, removing the tumor with an undisturbed shell of healthy tissue. This wide en bloc procedure can be called “excision” or “resection.” “Radical resection” means the en bloc removal of the tumor and the whole compartment of tumor origin which is virtually impossible for a spine tumor because of the ring shape of the vertebral body around the neural structures and also because of the fact that some compartments such as the epidural or subdural spaces extend from sacrum to cranium [16]. Slow-growing but

locally aggressive primary tumors that may be easily treated elsewhere in the skeleton may be unresectable and potentially lethal in certain locations in the spine. The surgical approach should be planned carefully and must achieve the prescribed appropriate margins. While intralesional removal may be associated with excellent outcomes for many patients with benign latent tumors and benign active tumors, more aggressive surgery is indicated for some locally aggressive benign tumors and many malignant tumors. Non-metastatic malignant tumors are ideally removed with wide surgical margins, when technically feasible.

As described above, the treatment depends on diagnosis, natural history, location, size of the lesion, as standardized in the Surgical Staging System (SSS) which dictates the type of surgical treatment to be used. Benign latent lesions (S1) do not require oncologic treatment as they are latent. Benign active tumors (S2) do have growth potential: Intralesional excision usually can be performed with a low rate of recurrence. Benign aggressive tumors (S3) infiltrate neighboring compartments and have wide reactive hypervascularized pseudocapsules. En bloc resection with wide/marginal margins is often indicated for reasonably acceptable recurrence rates when possible. If not, intralesional excision and additional local adjuvant therapies such as phenol or alcohol administration, cryotherapy with liquid nitrogen or abundant use of polymethylmethacrylate (PMMA) may be necessary. Depending on specific sensitivity, radiation therapy can be helpful as an adjuvant. The side effect of radiation therapy on growing bone must be considered, together with the risk of secondary radio-induced tumor in patients with a long life expectancy [22].

Management of malignant spinal tumors is more complex and requires a multidisciplinary approach. Early detection of the tumor followed by complete excision is advisable [18]. Again the SSS helps to delineate the progressive stages of a given tumor and the specific implications for surgical treatment and provides guidelines for the use of adjuvant therapy. Advances in chemotherapy and techniques for resection and reconstruction have expanded the role of local surgical management. For optimal surgical treatment oncological staging of lesion is essential. The grade of the tumor as a sign of the general behavior, presence of metastatic lesions, location and extension of the tumor in the spinal column are important factors. Local control of the malignant pediatric spinal tumor can only be achieved with a well-planned wide resection. However, wide en bloc excision procedure may be impossible in some cases because of the location and the extent of the tumor. Even if the spinal cord and nerve roots are sectioned above and below, the epidural space represents a compartment extending from the skull to the coccyx. Therefore, a tumor-free margin en bloc resection is not possible when a stage II malignant tumor is encroaching the canal.

Three different en bloc resection surgeries have been defined: vertebrectomy, sagittal resection, and resection of the posterior arch. Vertebrectomy implies removal of all the elements of the vertebra, which may be total, or hemi- in the sagittal plane. These procedures can be performed in staged, sequential, or simultaneous anterior and posterior approaches or in a single stage through posterior approach [18, 23–26]. Lesions involving the posterior elements of the spine are obviously submitted to en bloc excision by posterior approach [16].

Reconstruction of the spine after resection before maturity requires a further concern as long fusions can be followed by secondary impairment of sagittal balance, while short fusions – particularly if associated with muscle and ligament sacrifice as required in tumor resection – can create shortly a segmental instability [27].

25.5.1 Specific Spinal Tumors

25.5.1.1 Benign Tumors

Eosinophilic Granuloma (Langerhans Cell Histiocytosis)

It represents the Langerhans cell histiocytosis form localized only in the skeleton, different from Hand Schuller Christian disease and

Letterer Siwe disease. (Table 25.2) This lesion is a reactive proliferation of Langerhans cells forming granulomas and may produce focal destruction because of this essentially inflammatory character. Clinical manifestations range from a single bony lesion to multiple granulomas in bones and soft tissues to systemic forms of disease. The incidence of spinal involvement ranges from 7 to 25 % [28–33]. It is commonly seen in children less than 10 years of age and is more common in males [34–36]. The most common presenting symptom is pain [34, 35, 37, 38]. Vertebra plana is the typical radiological appearance caused by the partial or complete collapse of the vertebral body [28]. Vertebra plana is not the onset image, it is the image of the collapse following the erosive initial activity of the EG (Fig. 25.2). The initial imaging is hard to be distinguished from a lymphoma or Ewing's sarcoma and biopsy is mandatory. It is important to note that the collapsed vertebral body is located between two normal discs in order to differentiate from infectious disease. Asymmetrical vertebral collapse can lead to the scoliosis or severe kyphosis but the most common deformity is mild to moderate kyphosis [39] (Fig. 25.3). Neurological symptoms due to vertebral collapse may rarely be seen.

Table 25.2 Frequently encountered benign and malignant tumors of pediatric spine

Benign lesions
Eosinophilic granuloma (Langerhans cell histiocytosis)
Aneurysmal bone cyst
Osteoid osteoma
Osteoblastoma
Osteochondroma
Giant cell tumor
Fibrous dysplasia
Non-ossifying fibroma
Malignant lesions
Ewing's sarcoma
Osteosarcoma
Leukemia
Neuroblastoma (metastatic)
Wilms' tumor (metastatic)
Teratoma (metastatic)
Lymphoma (metastatic)

A skeletal survey or bone scan should be done to rule out other lesions of EG which is associated with multifocal disease. MRI is helpful for the differential diagnosis from malignancy. Histology of these lesions has three main components that are lipid-containing histiocytes with "coffee-bean" appearance, eosinophils, and Langerhans giant cells.

Solitary lesions are usually a self-limited disease. Treatment is somewhat controversial, but it is clear that many patients heal their lesions without any treatment (see Fig. 25.2) or for that matter, any treatment other than biopsy. Observation with or without spinal immobilization with cast, body jacket, orthosis, or collar that can be used for few months to several years has been the standard modality of treatment. This conservative treatment allows the load sharing of the anterior column and may produce an enhancement in the growth plate activity leading to a possible restoration of vertebral height [40]. Raab and coworkers have reported that 18.2–97 % vertebral body height restoration was possible in conservatively treated patients. It appears that the age of the patient is an important factor in this context. If the lesion had been identified at least 4 years before skeletal maturity, remaining growth capacity is usually enough for adequate remodeling regardless of location at the cervical, thoracic, or lumbar regions [37]. Radiotherapy, chemotherapy (only for disseminated form), and steroid injections have been advocated with no proven benefits over observation for solitary lesions. Operative treatment is only necessary in the rare instance such as neurological involvement secondary to vertebral collapse, compression of the spinal cord, extraosseous extension and instability of spine, or persistent pain.

Osteoid Osteoma (OO) and Osteoblastoma (OBL)

These lesions are most frequently seen in first two decades of life and show a propensity (greater for OBL) for the posterior elements of the spine. They may be located in the pedicles, transverse processes, laminae, and spinous processes. The

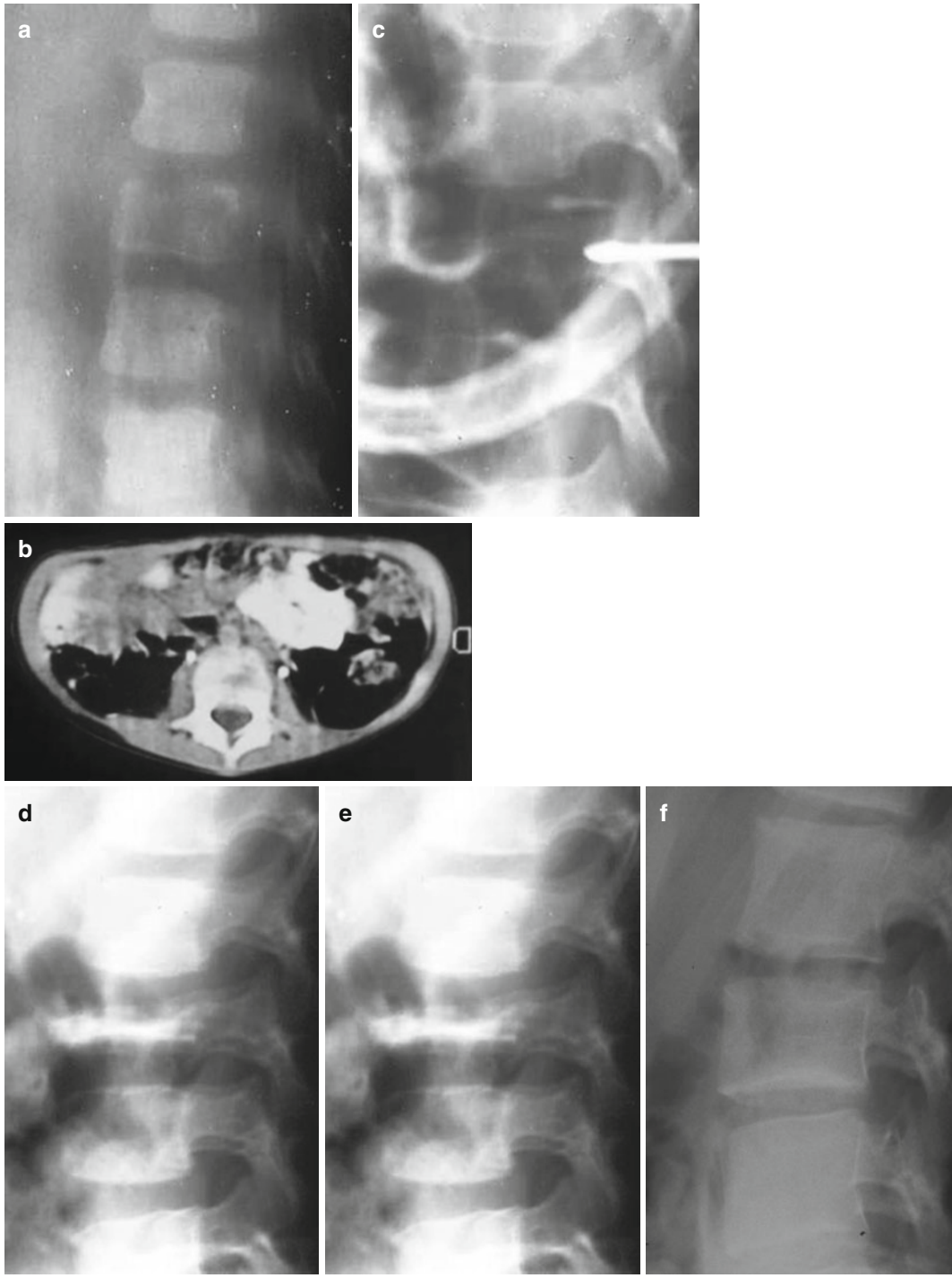

Fig. 25.2 A 4-year-old male, back pain and muscle spasm. Case observed in 1992 (**a**, **b**) Sagittal Tomography and CT scan show initial lytic changes in L2 vertebral body. (**c**) Biopsy was performed by transpedicular open approach and allowed the diagnosis of Eosinophilic granuloma. (**d**) An orthosis was advised. The standard radiogram 2 months later shows the tyical aspect of a vertebra plana. No pain. The patient was allowed to leave the orthosis. (**e**) Three years later initial reconstruction is evident. (**f**) At 10-year follow-up the reconstruction is complete, no functional loss

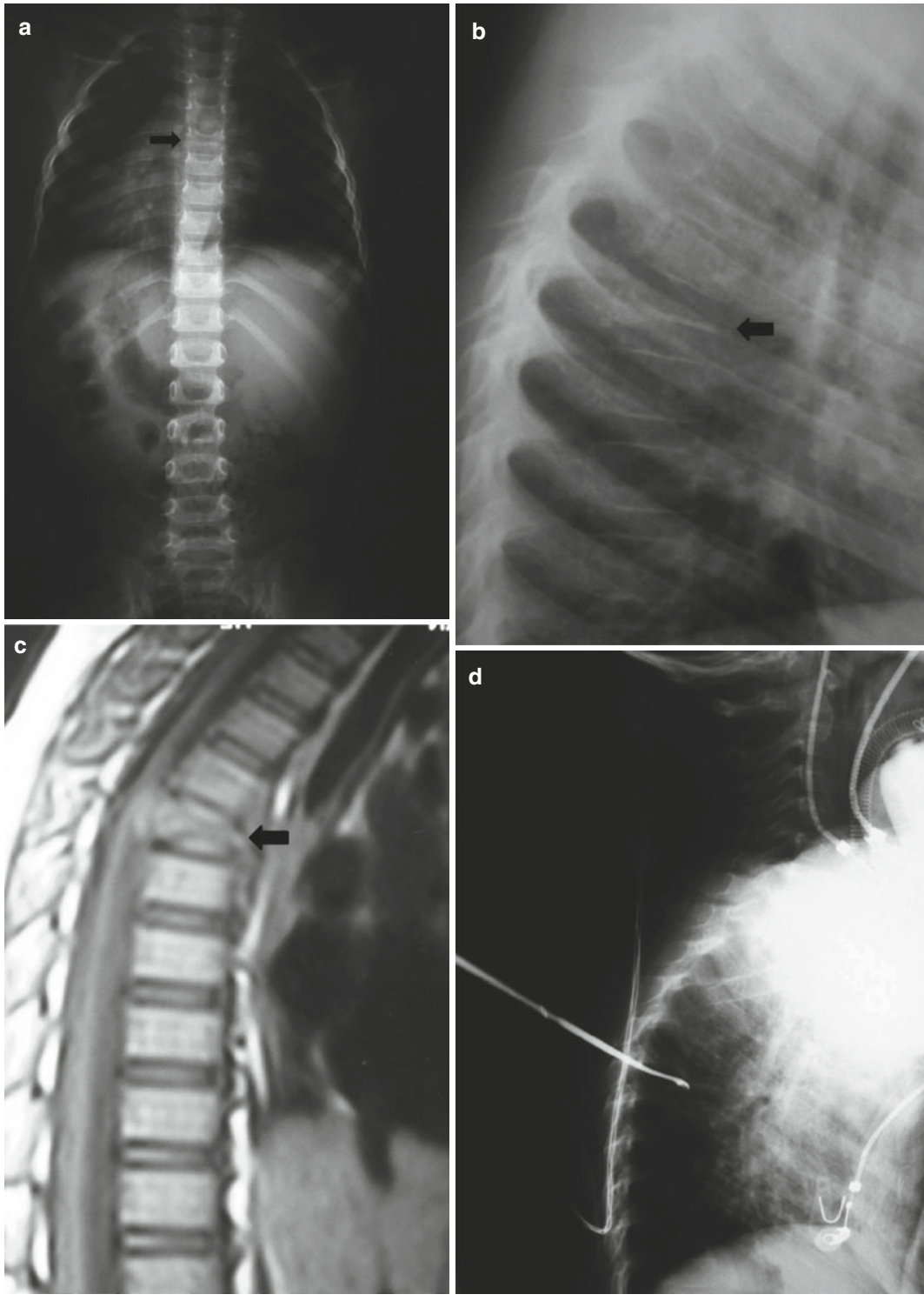

Fig. 25.3 A 5-year-old male presenting with back pain. (**a**, **b**) AP and lateral plain X-rays demonstrating the typical appearance of vertebra plana (*arrow*). (**c**) T1-weighted sagittal MR image demonstrating the totally preserved disc spaces (*arrow*) (**d**) Intraoperative lateral X-ray taken during transpedicular biopsy

overall rate of spine localization ranges between 10–41 % for OO and 30–50 % for OBL [23]. Pain is the predominant symptom and is usually worse at nights and activity. It may resolve with the use of nonsteroidal anti-inflammatory drugs (NSAID). Night pain and dramatic response to NSAID should evoke the clinical suspect of OO. On the other hand, OBL pain has a lower response to NSAIDs. These tumors often produce pain before they become visible on plain radiographs. A CT scan is often required to diagnose the lesions which have the typical appearance of osteosclerosis surrounding a radiolucent nidus of less than 2 cm in diameter for OO and greater than 2 cm for OBL (Figs. 25.4 and 25.5). Technetium bone scan can be useful in establishing the diagnosis while roentgenograms are still negative, and the pain definition by the patient is vague and non-localizing by showing a nonspecific but intense, well-defined focal uptake of activity [41–43]. OO is typically a benign latent self-limiting lesion that has a tendency to spontaneously regress over several years whereas OBL are usually locally aggressive tumors.

Painful scoliosis is another fairly well-recognized presentation of OO and (less frequently) OBL [6, 7, 44], the incidence of being the initial symptom of spinal OO and OBL ranges from 25 to 63 % [8, 45]. Lesions with scoliosis are more common in thoracolumbar spine than cervical spine and have been identified as the most common cause of pain provoked reactive scoliosis [46]. Saffuidin and coworkers have reported on the typical findings associated with scoliosis in OO and OBL, based on a series in which 63 % of patients had scoliosis overall. Lesions were mostly located on the concave side near the apex. Asymmetrical location of lesion within the vertebral body or neural arc appeared to be the most significant factor for the development of scoliosis whereas a lesion at the center of vertebral body (e.g., spinous process) had the least likelihood. It was postulated that asymmetrical inflammatory effect of the lesion caused asymmetrical muscle spasm and secondary scoliosis. Cervical lesions were associated with a minor chance of developing scoliosis, predominantly at the lower cervical spine, but asymmetrical inflammation may lead to torticollis [9]. Considering that congenital and idiopathic scoliosis are always painless, a painful scoliosis should always rise the suspect of OO and induce to submit the young patient to an isotope scan.

If the patient's symptoms can be managed with NSAIDs without any significant side effects, a trial of medical treatment may be prescribed. Long-term medical treatment was found to be as effective as surgical treatment [47]. However, such prolonged use of drugs is often associated with at least gastrointestinal irritation and may lead to severe hemorrhage. Surgical treatment should be considered if medical treatment cannot be used or is not successful, that of OO being intralesional excision. There is no need for the removal of the entire sclerotic reaction; however, the nidus should be completely removed to ensure good pain relief and to prevent recurrences. As pain is usually radically improved after complete resection of the nidus, it can attest to the completeness of the excision as well. Spinal deformity improves in almost all patients within 15 months [6, 8, 23].

Contrary to OO, treatment of OBL consists of complete surgical excision. Curettage has been advocated in the past, but with an unacceptable rate of recurrence. Even marginal excisions carry a recurrence risk of about 10 % [48]. Radiotherapy has been advocated in the past because of these relatively high rates of recurrence after surgery but has been mostly abandoned now in the era of modern spinal surgery, as it may be associated with the danger of malignant transformation of these lesions.

Aneurysmal Bone Cyst (ABC)

ABC is a benign and highly vascular bony lesion that is relatively rare and often mistaken for a malignant tumor due to its both radiological and pathological aggressiveness. Three to twenty percent of lesions are located in vertebral column [39, 49, 50]. These lesions usually occur in the second decade of life and have a tendency to be located at the posterior elements of the vertebra [21, 23, 51–53]. It is important differentiate primary benign ABC with specific immunoistochemical pattern from reactive ABC associated to

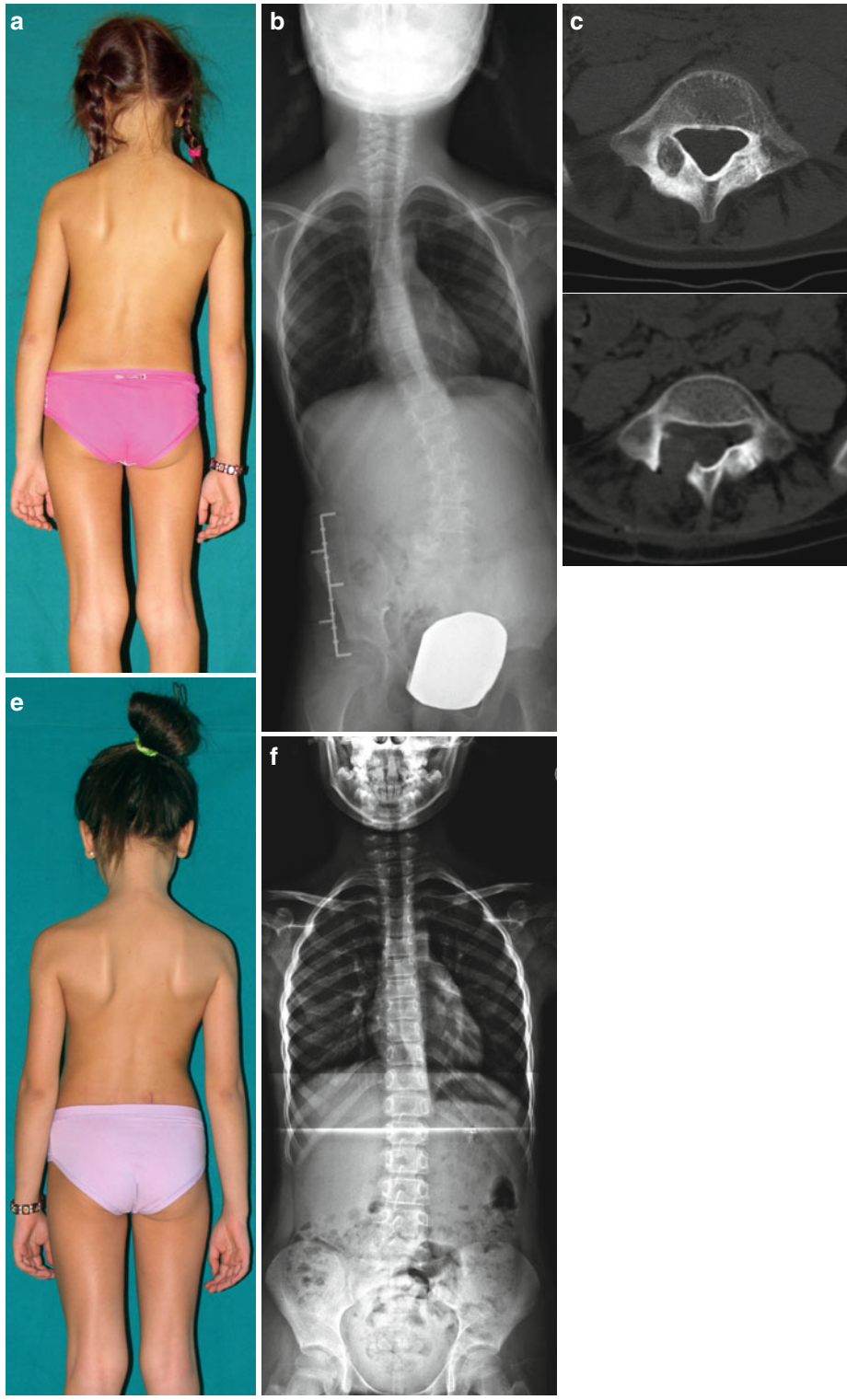

Fig. 25.4 A 6-year-old female. (**a**) Painful scoliosis. (**b**) Standard standing radiogram shows a left lumbar scoliosis with relevant torsional component. (**c**) CT scan shows a small lytic area surrounded by a sclerotic reactive bone, located in the right posterior arch of L5. Imaging is consistent with the diagnosis of osteoid osteoma. (**d**) Postoperative CT scan; complete excision of the nidus. (**e**) Six months later the deformity is reduced. (**f**) Standing radiograms showing recovering of normal alignement in the coronal and in the transverse plane

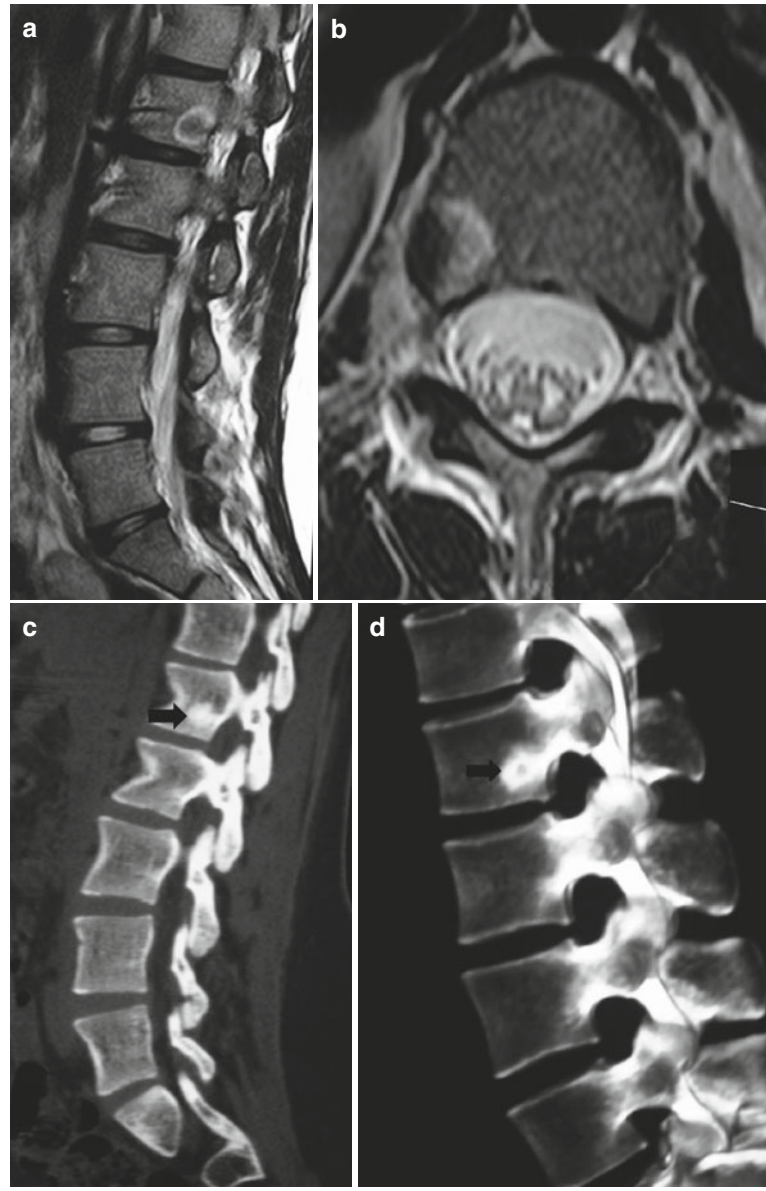

Fig. 25.5 A 14-year-old female presenting with back pain that is worse at night time and reasonably responsive to salicylates. MR imaging was prescribed, revealing a completely hypointense nucleus surrounded by a relatively hyperintense zone on sagittal (**a**) and axial (**b**) T2-weighted images. (**c**, **d**) Sagittal and 3-D reconstructions of CT images demonstrating a sclerotic zone surrounding a relatively lytic nidus typical for osteoid osteoma (*arrows*)

different benign and malignant tumor (such as chondroblastoma, osteosarcoma, and mostly giant cell tumors).

ABC is mostly located in the cervical and thoracic spine, less frequently in the lumbar spine and sacrum [21, 51, 52].

ABC is a benign but locally aggressive lesion [54, 55]. The most common complaints are pain and neurological symptoms as a result of spinal cord or nerve root compression both of which usually almost immediately regress with surgical decompression [52, 56]. Radiography reveals an expansile, lytic lesion with bubbly appearance surrounded by a thin shell of reactive bone. Fine bony septations give the lesion a soap-bubble appearance. Occasionally they may involve three or more adjacent vertebra. CT and MRI provide optimal evaluation of lesion content and expansion of tumor. MRI reveals multiloculated, septated, expansile lesions with fluid-fluid levels whereas CT is important in demonstrating the intact bony shell as a telltale sign of a benign

lesion. Definitive diagnosis may be established with CT-guided needle biopsy, trocar biopsy, or open biopsy and/or frozen section at the time of the resection procedure. A thorough histological study coupled with clinical and radiographic data is essential for a correct diagnosis, as the differential diagnosis includes tumors with similar histological appearances such as giant cell tumors as well as osteosarcoma [52]. Angiography may also be helpful in the surgical planning of these lesions. Selective embolization is now considered the gold standard and can be used as a stand-alone treatment or to reduce the intraoperative bleeding if performed before the surgery. Radiotherapy has been advocated in the past but it is now established that lesions treated with radiotherapy have a risk of malignant transformation [39, 52, 56]. Early surgical intervention with intralesional curettage of all affected bone is recommended after the diagnosis has been established by biopsy. Intraoperative excessive bleeding may be a problem, which can be solved with selective arterial embolization before surgery or early removal of the thin bony wall of the lesion before the resection of the cystic lesion itself thereby shortening the period of surgery with significant blood loss. Recurrence rates of 10–50 % have been reported with intralesional excision [39, 52, 54, 55]. As a real danger of recurrence after simple intralesional curettage exists, cauterization of the osseous cysts wall, extended curettage with high-speed diamond burrs, and administration of dilute (5 %) phenol and absolute alcohol administration have been advocated in order to decrease this rate [57]. More aggressive resection margins should be reserved for cases in which it is feasible without creating any iatrogenic instability or recurrent cases in which intralesional surgery has definitely failed. Spinal fusion should be performed when the lesion (Fig. 25.6) or the surgical procedure has rendered the spine unstable. Complete excision with stabilization can provide cure of aneurysmal bone cyst and good outcomes with low recurrence [58].

In selected cases Denosumab (off label treatment) could represent an alternative to surgical and other non-surgical procedure [59] but this drug cannot be used before skeletal maturity.

Hemangioma

These lesions are usually clinically silent and found incidentally. The spine is the most commonly affected part of the skeleton. HE are predominantly located in the vertebral body and especially in thoracic region. Symptomatic HE of bone is a relatively uncommon entity. Cases of soft tissue extension with nerve root or spinal cord compression have been documented [17, 23] but they are extremely rare before skeletal maturity [60, 61].

Patients' main symptom is pain, which may or may not be associated with pathological fractures mostly reported during pregnancy. CT scan and MRI are diagnostic. Axial CT views demonstrate the typical honeycomb pattern, which is diagnostic, whereas MRI reveals the fluid content and increased vascular blood flow. Most patients do not require treatment as long as they remain symptom free. Fairly large lesions may be considered as candidates of treatment because of the possibility of a fracture, but specific guidelines regarding the size and location of the lesion in regard to the risk of impending fracture have not been established. Embolization may be used before surgery to reduce bleeding. Vertebroplasty or kyphoplasty are becoming increasingly popular and have the potential of rendering any open surgery unnecessary [62, 63], but are limited to cases with intact posterior wall. Open surgery with resection of the lesion should be reserved for patients with pathologic fractures and neurological compromise.

Osteochondroma

Osteochondromas (exostoses) are the most common benign bone tumors. They occasionally occur in the spine, especially in the pediatric population. Multiple hereditary exostosis is more likely to involve spine. Posterior elements of cervicothoracic spine are the most frequent location. A simple painless mass may be the only presenting symptom. They may as well be diagnosed incidentally or, very rarely, may cause neurological symptoms. Plain radiographs are usually adequate in demonstrating the sessile or pedunculated mass. CT may be used to understand the exact location of the lesions, whereas MRI may be

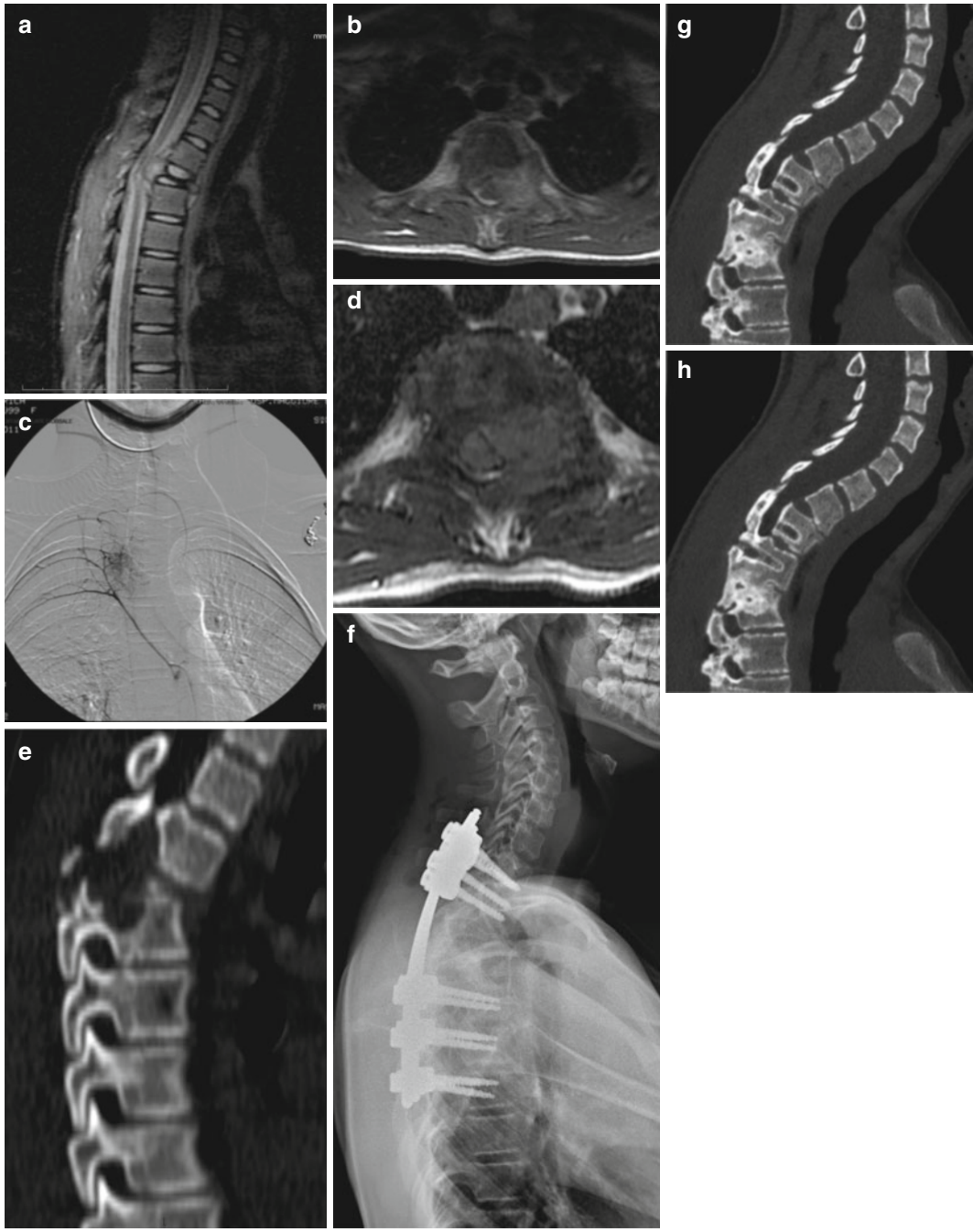

Fig. 25.6 A 12-year-old female. (**a, b**). Pathologic fracture of T3. Vascularized soft tissue eroding bone and invading the canal. Pain, no neurologcal symptoms. Histological diagnosis of Aneurysmal bone cyst (**c**). Angiogram and embolization repeated three times without significant effect on bone reconstruction. (**d**) MRI shows increasing of the volume and of the encroachment of the canal (**e**) CT scan sagittal reconstruction demonstrating sagittal misalignement with subluxation. Lower limb weakness, numbness. (**f**) Intralesional excision, fixation, grafting, and local injection of stem cells. (g, h) One year later the implant was removed. In these images the last CT scan reconstruction 2 years after excision. Fusion with acceptable kyphotic deformity. No recurrence of the aneurysmal bone cyst

necessary to evaluate the thickness of the cartilage cap. There has always been a debate on the importance of the thickness of the cartilage cap, most current information suggesting that a cap thicker than 2 cm may be associated with malignant degeneration [20]. Osteochondromas of spine tend to grow fairly slowly until skeletal maturity and remain dormant thereafter. Painful lesions with neurological compromise and those that had been demonstrated to commence growth after skeletal maturity need to be treated operatively. An essential feature of excisional surgery is the necessity of the removal of the entire cartilaginous cap. Recurrence risk is low in children in whom it could completely be excised [20].

Other Benign Tumors

Giant cell tumors occur very rarely in the pediatric population. This tumor is composed of a highly vascularized network with numerous multinucleated giant cells. Most giant cell tumors are benign and locally aggressive tumors with a predilection for the vertebral body. Pain, local tenderness, swelling, and neurological problems are the usual presenting symptoms [21, 23]. Sacrum is the most commonly affected area. As occasional pulmonary metastases have been reported, chest radiograms or CT should be performed in oncological staging [64, 65]. Pediatric patients with giant-cell tumors should be managed with wide en bloc resection whenever possible. If obtaining tumor-free margins en bloc resection is not possible, an intralesional excision can be performed (always planning embolization as a mandatory preoperatory step to reduce bleeding) but the patient and family should be warned on the possibility of recurrence. Recurrence rates are high in adult population but little is known for the pediatric population because of the rarity of the tumor. Adjuvant surgical treatments like phenol, polymethylmethacrylate or liquid nitrogen may be used. For large giant cell tumors or tumors with critical locations that would render them virtually unresectable (e.g., sacrum), repeated arterial embolizations has been reported as a useful stand-alone treatment modality with favorable results [23, 66]. Radiotherapy is the last resort because of the potential for increased malignant transformation and should be reserved for those uncontrollable lesions with multiple recurrences.

Another uncommon lesion is fibrous dysplasia of the spine. Some cases of fibrous dysplasia have been reported. Pain is usually the main complaint. Monostotic and polyostotic forms may be seen in children with the most common site of location at the lumbar and the thoracic spine. The possibility of neurological involvement has also been reported [67, 68]. Scoliosis has also been reported in extensive fibrous dysplasia and may require management with standard scoliosis surgery [69].

25.5.1.2 Malignant Tumors

Osteosarcoma

Osteosarcoma is the most common primary malignant bone tumor [23]. Primary osteosarcomas are relatively uncommon in the spine consisting of 3 % of all osteosarcomas and often affect the thoracic and lumbar spine [21, 70]. They are most frequently encountered in the second decade of life. Vertebral body is predominantly involved. Most osteogenic sarcomas of spine in children are metastatic as a relapse of the most common locations in the limbs.

Pain is the most common and earliest presenting symptom [23]. Night pain will be present in approximately 25 % of patients and up to 40 % will be expected to have neurological symptoms [20, 70] Laboratory findings usually are not helpful except for the possibility of elevated serum alkaline phosphatase levels. Osteosarcomas are locally and systemically aggressive, high-grade malignancies. They radiographically present with a lytic, blastic, or mixed lesion with an ossifying matrix. All patients in whom a primary or metastatic osteosarcoma is included in the differential diagnosis should be evaluated with an MRI of the involved region so as to define the lesion and extension. Osteosarcomas have a very high tendency to metastasize, almost exclusively to the lung, rarely to the other bones. Pulmonary CT and bone scans (or total body MRI) are essential requirements for staging the disease.

For those highly malignant lesions, biopsy should be performed at the center and by the

surgical team who will assume the definitive responsibility for the treatment. Every effort should be made to remove the tumor by en bloc excision with tumor-free margins, but even the most successful surgery will not be adequate if complete chemotherapy pre- and postoperative courses are not performed [71].

Primary osteosarcomas of spine present complex therapeutic problems in management, mainly because of the difficulties posed by surgical resection. It has to be understood and relayed to the patients and families that spinal osteosarcomas have a very poor prognosis. Median survival of spinal osteosarcomas has been reported to be 6–10 months, however new surgical techniques and technologies and modern treatment regimens may improve this outcome [21, 70–73].

Ewing's Sarcoma

Ewing's sarcoma is the most common primary malignancy of the childhood to be located in the spinal column. It is most frequently seen in the first two decades of life but is uncommon under 5 years of age. Approximately 8–10 % of all Ewing's sarcomas occur in the spinal column with sacrum being the most common site. The tumor histologically consists of uniform, small, round, highly undifferentiated cells [23, 74]. These may resemble neuroblastomas, rhabdomyosarcomas, and lymphomas – malignant tumors with similar small round cells. Pain and neurological deficits are the most common presenting symptoms. Unlike osteosarcoma, Ewing's sarcoma may present with systemic symptoms such as fever and weight loss and may be mistaken as a systemic infection at the early stages. It is very common to have the systemic inflammatory signs such as ESR and CRP elevated.

Plain radiographs reveal lesions with a moth-eaten appearance, a shadow of the soft tissue mass and aggressive periosteal reaction. The occurrence of vertebra plana is also possible and may lead to a misdiagnosis of eosinophilic granuloma. As the lesion usually starts at the bone of the vertebral body, disc spaces are usually preserved until late disease. Like osteosarcoma, MRI is mandatory to evaluate the soft tissue extension and the spinal canal. Ewing's sarcoma's staging strategy is similar to osteosarcoma and necessitates chest CT and bone scans (or total body MRI) so as to detect lung and bone metastases. Although specific staging systems have been advocated and commonly used in the past, the use of the Surgical Staging System is getting to be more popular and accurate because of the advances in the general understanding of tumor behavior and in treatment. The traditional treatment of Ewing's sarcoma consists of neoadjuvant chemotherapy followed by local control with radiation therapy or surgery or both. There has been an ongoing debate on the usefulness of ablative surgery for such tumors that are reasonably responsive to radiotherapy but recent studies have demonstrated clearly better results in favor of surgical resection [75] (Figs. 25.7, 25.8, and 25.9). It is advisable to use neoadjuvant chemotherapy for a better treatment outcome and especially to decrease the volume of the tumor mass for easier and safer surgical resection [21, 76]. In our hands radiotherapy is only recommended if the lesion could not be resected with adequate margins (wide/marginal), which has been relatively frequent but becoming less so. Spinal metastases of Ewing's sarcoma have poorer prognosis compared to primary Ewing's sarcoma of the spine [20].

Leukemia

Leukemia is the most common cancer in young children. Peak incidence is between 2 and 5 years of age. All organ systems may be affected, the skeleton being one of the most frequent diagnostic sites of the acute form of the leukemia. Bone pain may be the presenting symptom in 25 % of patients. The others symptoms are lethargy, anemia, and fever. Changes in laboratory tests such as increased white blood cells (WBC) and decreased platelet count are characteristics of leukemia when present. Elevated ESR and CRP levels may also be seen. Plain radiograms often do not have any definitive diagnostic appearance. Diffuse osteopenia, osteosclerosis, osteolysis, and periosteal reaction may be seen in and around the vertebral bodies of leukemia patients. Pathologic fractures with or without vertebral body collapse also may occur. Definitive treatment is specific to the type of leukemia but in

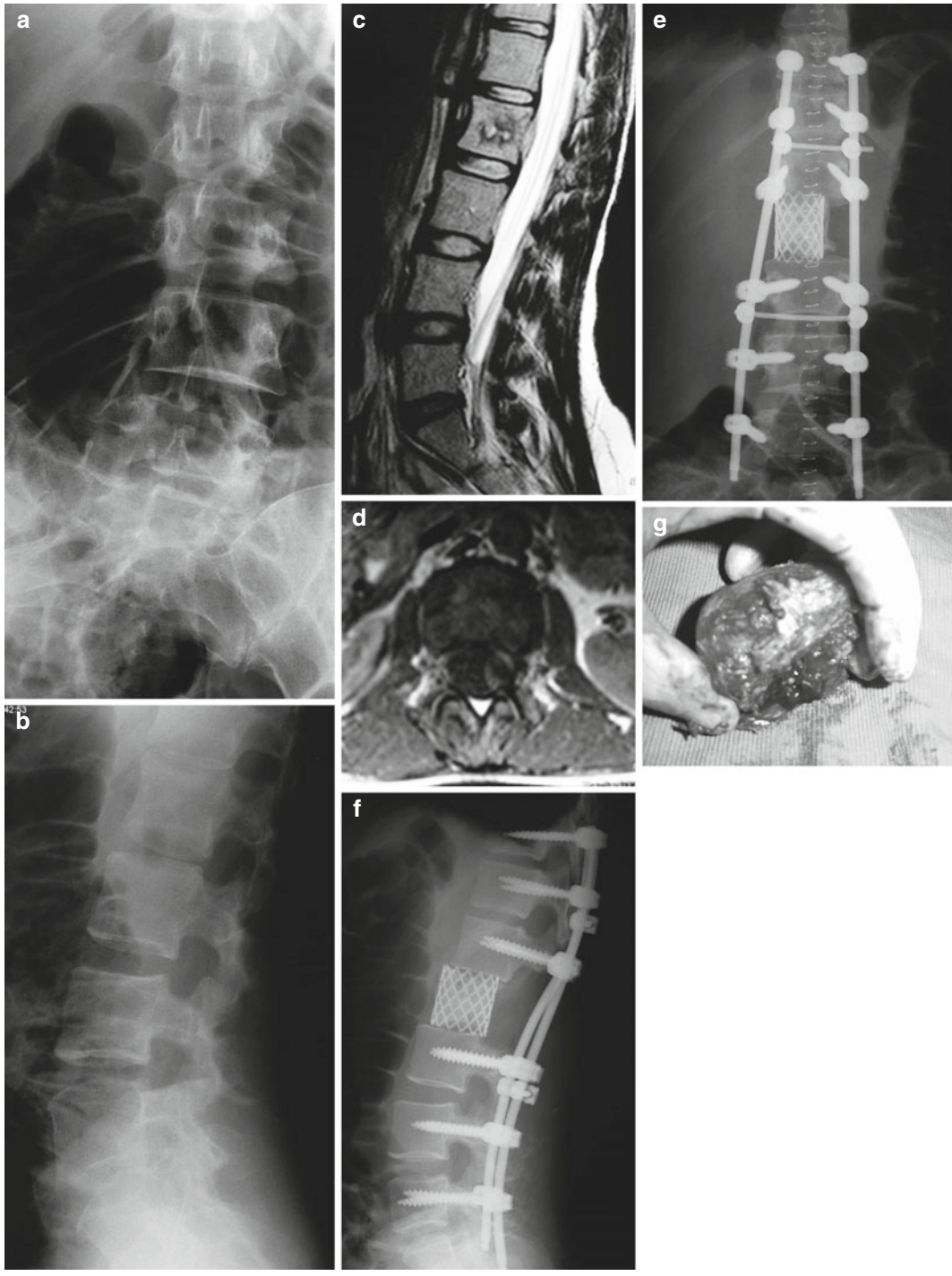

Fig. 25.7 A 15-year-old female presenting with back pain. (**a**, **b**) AP and lateral plain X-rays negative for any significant finding. (**c**, **d**) T2-weighted sagittal and axial MR images demonstating the tumor located within the body of L2. Oncologically staged as IIB, surgically (WBB) staged as zones 4–10 from B to D because of the epidural component. Needle biopsy under CT guidance confirmed the diagnosis as Ewing's sarcoma. (**e**, **f**) Postoperative X-rays following en bloc excision. (**g**) Resected vertebral body

some cases surgery may be needed for the treatment of spinal instability or pain associated with pathologic fractures [75, 77, 78].

Other Malignant and Metastatic Lesions

Metastatic involvement of the spine in children may occur in rhabdomyosarcomas, neuroblastomas, Wilms tumors, lymphomas, and teratomas. Pain is the main symptom because of micro- or macropathologic fractures. Vast majority of metastatic tumors are sarcomas with varying levels of radioresistance. Treatment depends on the definitive diagnosis of the primary tumor as well as the stage of the disease and the general condition of

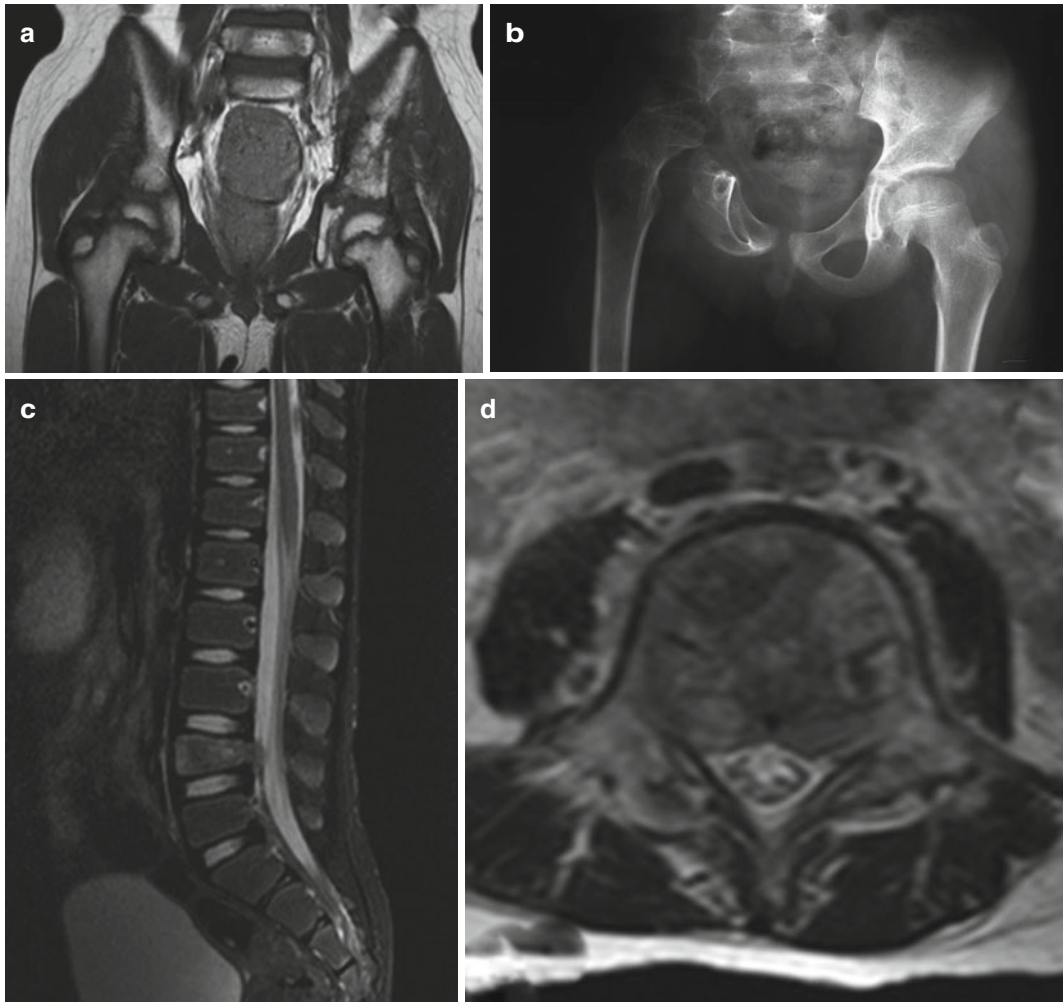

Fig. 25.8 An 8-year-old male presenting with right-sided hip and buttock pain. (**a**) T1-weighted MR image demonstating a mass located at the iliac wing, infiltrating the surrounding muscle. Oncologically staged as IIB, biopsy revealed a Ewing's Sarcoma. (**b**) Plain AP pelvis view following wide excision of the tumor after neoadjuvant chemotherapy. Developed mild back pain 1 year after index surgery. (**c**–**d**) Lateral plain X-ray and STIR axial and sagittal MR images demonstrating metastatic lesion at L4. Oncological staging III, surgical (WBB) staging zones 4–9, B and C. Patient was not referred to surgery at this stage, received radiotherapy followed by another episode of chemotherapy. (**f**, **g**) STIR coronal MR images at 6 months after the radiotherapy. Note that the the lesion has spread not only both of the neighboring levels but also completely surrounds the dural sac at his point. (**h**) T1-weighted sagittal MR image confirming the involvement of additional levels as well as the epidural space (WBB staging three levels, 1–12, B to D). This patient was considered to have become in-operable at this stage, became paraplegic soon after and expired 3 months later

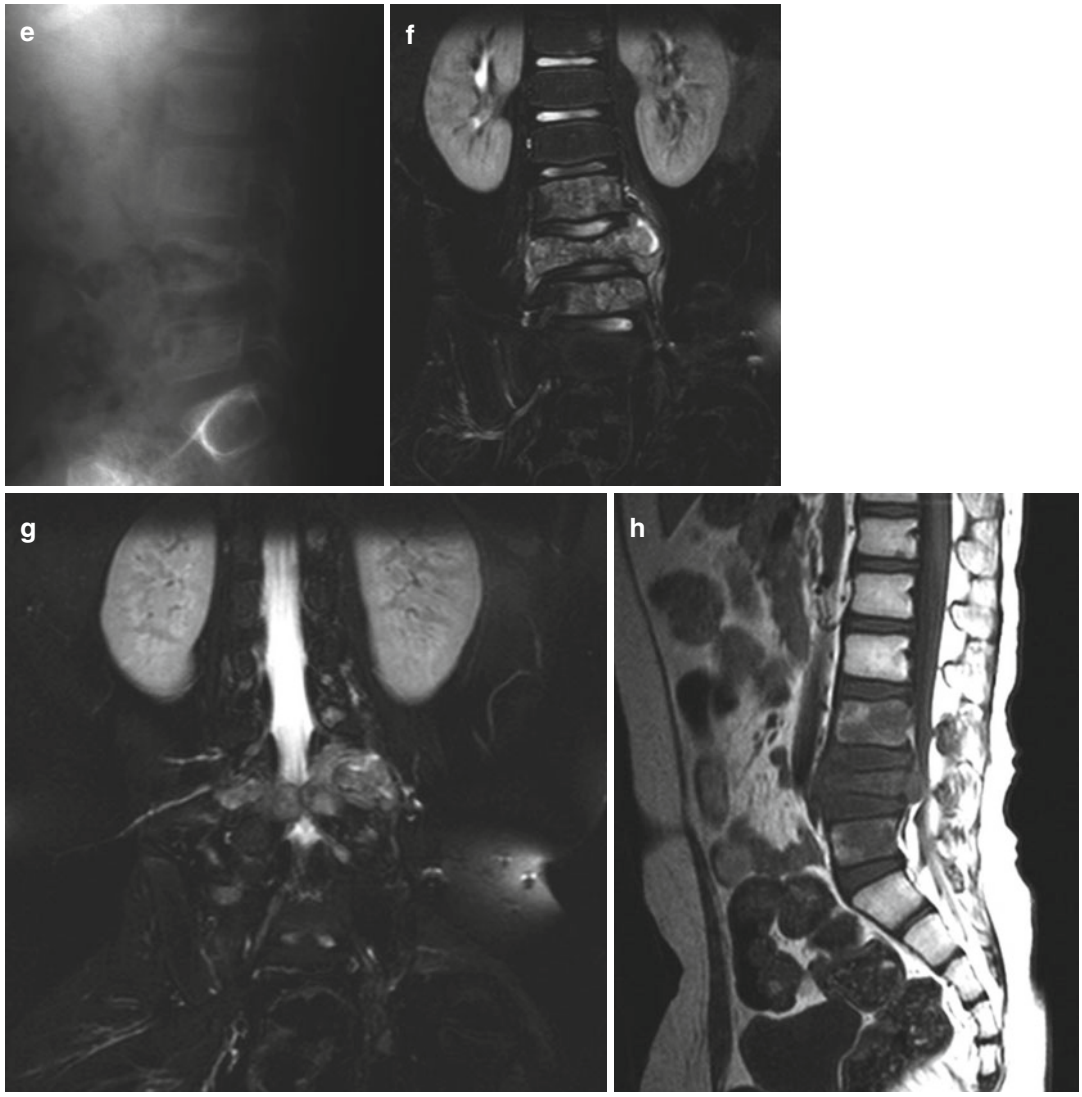

Fig. 25.8 (continued)

the patient. In cases of instability, spinal stability should be reinstituted with segmental spinal instrumentation [21, 79].

25.6 Summary

Tumors of the spinal column in pediatric age have distinct features in regard to diagnosis and management. Primary lesions are far more common compared to adults and benign tumors are more frequent than malignant. Some tumors, originating from other systems but affecting bone such as leukemia may be almost specific to this age group as well. Spine is also a frequent location for the metastases of pediatric tumors like Wilms and neuroblastoma.

On the other hand, the general principles of evaluation, staging, biopsy, and treatment are the same predicted for the adults. Profound knowledge of the principles of musculoskeletal tumor surgery as well as an understanding of the specific difficulties of oncological surgery in the spinal column is essential. Limitations due to

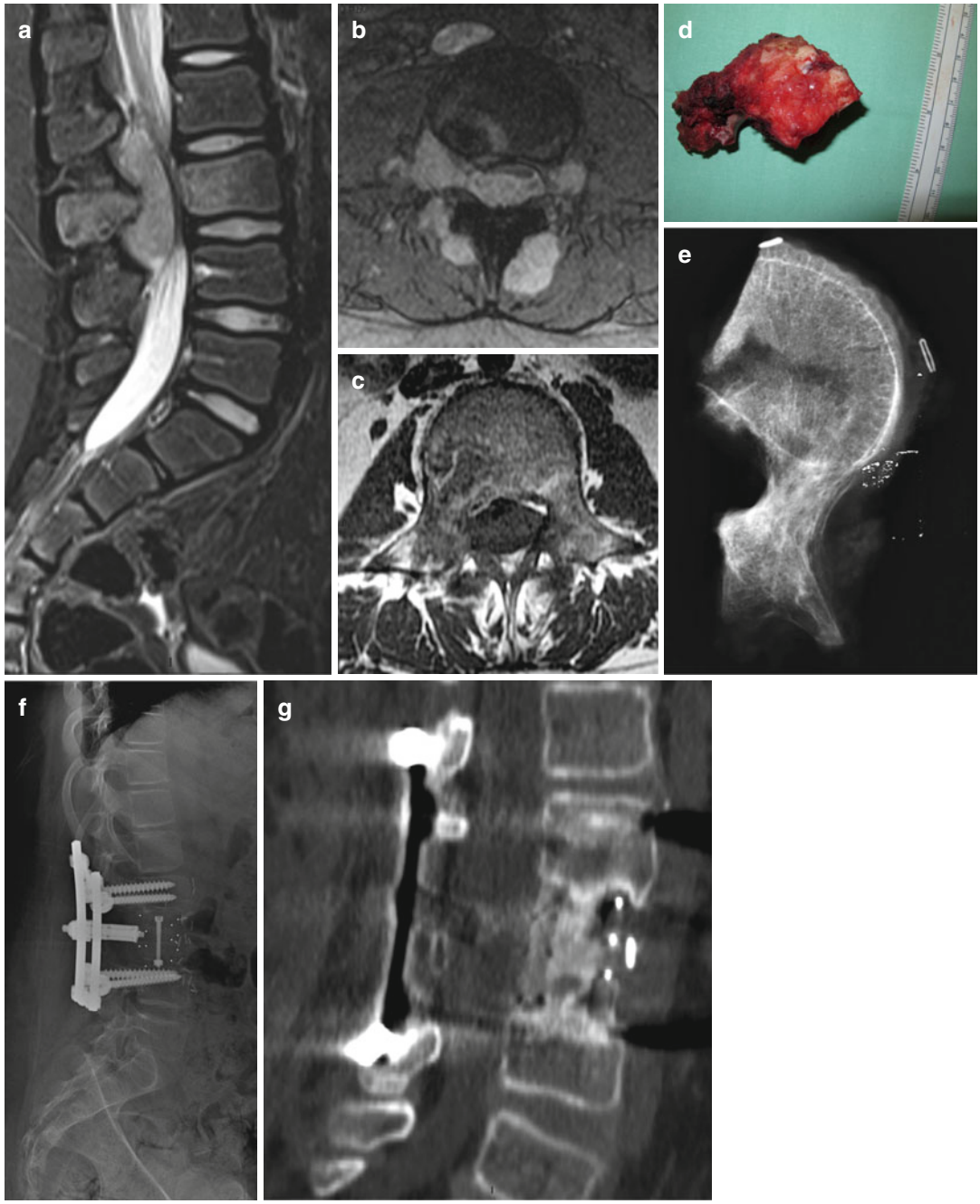

Fig. 25.9 A 9-year-old female complaining pain and progressive lower limb weakining. (**a**, **b**) L3 tumor with huge soft tissue extension invading the canal and compressing the cauda equina. Expansion outside the bone without destruction the cortex suggests the diagnosis of small cell malignant tumor. Trocar biopsy confirms the diagnosis of Ewing's sarcoma. (**c**) After three course of chemotherapy the soft tissue component has disappeared, neurological symptoms are regressing. (**d**, **e**) En bloc resection. (**f**) Three-year follow-up: no evidence of local disease. Lung metastases regressing under chemotherapy, able to walk. (**g**) Fusion of the graft inside and across the carbon fiber cage

skeletal immaturity must be imposed to radiotherapy, while surgery must consider the side effect of fusion in the pediatric spine on sagittal balance.

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Acute Pediatric Spinal Infections

26

Ahmet Alanay and Caglar Yilgor

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Key Points

- The umbrella term spondylodiscitis covers vertebral osteomyelitis, spondylitis, and discitis that are different manifestations of the same pathological process.
- Extraskeletal manifestation of the disease is also possible.
- Discitis is seen in 1–2:30,000 with a mean age of 2.8 years, and mostly the infection is localized in the lumbar or lumbosacral region.
- Commonly, low-virulent, slow-growing atypical microorganisms cause discitis; however, delay in diagnosis and treatment may lead to of restriction of the spinal mobility with persistent disc space narrowing, and/or with partial or complete fusion.
- Vertebral osteomyelitis is seen in 1:250,000 with a mean age of 7.5 years, and can manifest in the lumbar, thoracic, or cervical regions.
- Most patients with vertebral osteomyelitis respond well to antibiotic treatment and functional outcome is generally acceptable; however, the radiological outcome is not as innocent.

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Abbreviations

CRP	C-reactive protein
ESR	Erythrocyte sedimentation rate
MRI	Magnetic resonance imaging
PCR	Polymerase chain reaction
PPD	Purified protein derivative

26.1 Introduction

Spinal infections are uncommon in childhood; therefore, the clinical workup should begin with a high level of suspicion. The umbrella term spondylodiscitis covers vertebral osteomyelitis, spondylitis, and discitis (infections of the vertebral body, the joints, and the disc space, respectively) that are different manifestations of the same pathological process [1]. The differentiation between these entities in the early course of the disease process is often difficult [2]. Yet, vertebral osteomyelitis and discitis have distinct epidemiologic, clinical, and radiographic features. Although the infection affects the disc and adjacent vertebral bodies initially, it may spread into the paravertebral and epidural spaces, the meninges, and the spinal cord [3–5]. Extraskeletal manifestation of the disease is also possible [6]. Mostly, the spreading route is hematogenous; however, direct implantation after trauma and surgery and contiguous spread is also possible [7]. From an etiological point of view, infections of the spine can be divided into pyogenic, granulomatous, and parasitic infections. Since this chapter covers the acute pediatric spinal infections, the pathogenesis, evaluation, treatment, and outcomes of pyogenic discitis and vertebral osteomyelitis will be reviewed in this chapter.

26.2 Discitis

Discitis is a rare condition that has an approximated incidence of 1–2 cases in 30,000 [8]. The mean age of children is 2.8 years [2] and most children are younger than 5 years [7]. Yet, discitis displays a bimodal age distribution, in which there is a peak incidence in neonates and infants, and a smaller peak in juveniles [2, 9–11]. Although any level in the spinal column can be affected, in 75 % of the cases infection is localized in the lumbar or lumbosacral region [2, 12]. Since erroneous first diagnosis is often reported or diagnosis can be established after a significant delay, obtaining an early magnetic resonance imaging (MRI) scan is important when suspected as not to delay the start of treatment [11, 13, 14].

26.2.1 Pathogenesis

For decades, the topic pediatric discitis has sheltered consistent disagreement regarding to its etiology and optimal treatment [12, 15–24]. Due to the facts that the affected children are generally incapable of communication, that the symptoms are variable, and are not necessarily localized to spine, and that the laboratory investigations are not helpful, discitis was believed to be an inflammatory condition or secondary to trauma [9, 15, 23]. The reports of high percentages of afebrile and/or self-limiting discitis cases where the children do not appear to be acutely ill, and negative blood and disc cultures furthermore supported this view [2, 21, 23, 25]. In a recent report, however, the authors used specific real-time polymerase chain reaction (PCR) on blood samples of culture negative cases, and identified *Kingella kingae* as an etiological factor [26]. *Kingella kingae* is a slow-growing Gram-negative coccobacillus with a low virulence. The afebrile, culture negative children reported in the past might have been due to low-virulent and atypical organisms like *K. kingae* that does now grow in classical culture agars. Especially young infants may be at risk of infection by organisms that are less virulent and often part of the normal flora [27, 28]. Thus, today, the etiology of pediatric discitis is generally accepted as a bacterial infection involving the disc space and adjacent vertebral end plates [7, 29].

Discitis is mainly hematogenous in origin, where the preceding infections may be otitis media, urinary tract infections, or pulmonary infections [30]. Histological examinations revealed that nutrient arteries are the route of

infection for the hematogenous spread rather than the paraspinal venous system, which is primarily the course in adult discitis. Furthermore, septic embolus in the arterial system is proposed [31] to be involved as seen in osteomyelitis of the long bone metaphysis in children. The cause of infection may rarely be direct inoculation or a contiguous spread from an adjacent infection locus.

Intervertebral discs are avascular with hyaline cartilage end plates lying on both sides of the disc in children. Those cartilaginous plates, however, contain a vascular supply by means of numerous canals that appear before the 16th gestational week and persist until the third decade at which point the ring apophyses fuse [32, 33]. This rich vascular supply via canal systems to the disc is the unique property of pediatric end plates. Thus, blood-borne bacteria also reach the intervertebral disc via these roots to subsequently spread and contaminate the avascular disc with the hematogenous pathogen [34].

Pediatric vertebral bodies have a higher vascular supply through intraosseous enormous anastomosis compared to the adult vertebral body [35]. High vasculature of the body prevents the risk of pyogenic infection in the vertebrae. The pediatric vertebral body is also less prone to infarction. Additionally, cartilage caps on both sides prevent the spread of the infection to the vertebral body from the disc space [32, 33]. Instead bacteria are prone to be deposited at the superior and inferior parts of the disc that is adjacent to the vertebral end plates [33]. So, after the septic emboli, the vertebral bodies are often spared due to their rich blood supply and hyaline cartilage-capped end plates. In the avascular disc, however, the bacteria may be relatively free from the host defense mechanism and develop infection. As bacterial enzymes alter the disc biology and annulus fibrosis, the typical disc space narrowing is observed in plain radiographic images. If the infection persists, end plate erosion occurs, and a saw-tooth pattern of end plate destruction is seen on the radiograph. Redundant vascular supply at the vertebral body is consequently exposed to the infection at this level. Depending on the proficiency of the host mechanism, the outcome of the infection is determined. The infection may clear up or progress to the classical vertebral osteomyelitis with or without a concomitant soft tissue abscess.

26.2.2 Evaluation

26.2.2.1 History

The presentation of a child with discitis is variable and generally late, with few pathognomonic features. The family may report an antecedent or concurrent illness. Lumbar and lumbosacral region is the affected site in approximately 75 % of the patients [2, 12]. The child's ability to communicate determines the presenting symptoms. Children who are 3 years of age or younger often present with an acute onset of limping or refusal to bear weight [12]. Eventually, symptoms continue to progress and the children become uncomfortable in all positions other than lying supine. Children between 3 and 8 years of age may present with vague abdominal or back pain and have a decline in physical activity or exhibit abnormal posture. Older children may better localize the pain, or complain of buttock and leg pain due to nerve irritations [7].

26.2.2.2 Physical Examination

Physical examination may reveal low-grade fever; however, generally the child is afebrile. Refusal to walk and discomfort with hip movements can be observed but not to the degree that would suggest septic arthritis. Local spinal tenderness and paraspinal muscle spasms with a decreased spinal range of motion and hamstring tightness are common findings. Moreover, examination may also yield a positive result for the straight leg raise test. Although rare, lower motor neuron signs of limb weakness, reduced tone, and absent reflexes were also reported due to inflammatory tissue surrounding spinal nerves [9]. A stiff back may be observed for those children who can walk. Additionally, the child typically shows difficulty in picking up an object from the floor and probably would bend the knees then squat while keeping the back straight [7].

26.2.2.3 Laboratory Workup

Laboratory tests should involve a complete blood count with differential white blood cell count, blood cultures, erythrocyte sedimentation rate (ESR), and C-reactive protein (CRP). Typically, white blood cell count is in the high normal range and a left shift with mild leukocytosis [24]. ESR and CRP might be high providing nonspecific information. These markers can be more useful in monitoring the efficacy of the treatment [6, 10, 29]. It was suggested that a weekly 50 % reduction in CRP represents adequate improvement [1, 36]. Blood cultures should be expanded to involve fungi and mycobacteria, and purified protein derivative (PPD) skin test should be requested whenever subacute infection is suspected [8, 9]. Newer data suggests that PCR for low-virulent and atypical organisms should also be obtained for possible pathogens that does now grow in classical culture agars [26].

Performing disc space aspiration or biopsy is not routinely necessary due to low positivity rates, potential morbidity, and the need for sedation and/or anesthesia [9, 34, 37, 38]. It is furthermore discouraged in toddlers because of its low influence on the choice of antibiotic regime and the unknown long-term effects of the procedure [9]. When performed, *Staphylococcus aureus* is the most commonly isolated pathogen [12, 16, 18, 20, 36, 39]. Computed tomography (CT)-guided needle biopsy or open surgical biopsy may be used for children that fail to respond to initial empirical intravenous antibiotic treatment to rule out neoplasm, fungal infection, tuberculosis, brucellosis, and nonstaphylococcal pyogenic infections [7].

26.2.2.4 Imaging Studies

Plain radiographs are usually abnormal including loss of disc height and possible endplate irregularities especially when the symptoms have persisted for 2–3 weeks [2, 7, 9, 10]. Crawford et al. [21] described four radiographic phases for discitis: (1) latent phase: radiographs are normal, (2) acute phase: 2–4 weeks after the onset of symptoms characterized by narrowing and erosion of the disc space, (3) healing phase: 2–3 months after radiographic changes occurred, characterized by sclerosis of the vertebral contours, and (4) late phase: narrowing of the affected disc space with possible ankylosis.

Technetium 99 m-labeled bone scans are safe and highly sensitive in discitis, although they cannot differentiate discitis from other possible causes of back pain. Although scintigraphy may precede the radiological changes and can aid in an early diagnosis [8, 40] and scan with Indium-marked leukocytes increases the specificity, the use of MRI has led scintigraphy falling into disuse [41]. Bone scans may be helpful in toddlers when physical examination findings fail to localize the symptoms to the spine [42].

CT scans are easy to perform and may show bony endplate erosions, but do not add much value to management decisions [7, 36]. Therefore, CT may be preserved only to aid percutaneous needle biopsies.

MRI scanning is the most useful imaging method especially in the early stages, and is routinely recommended to confirm the diagnosis [10]. Early use of total spinal MRI under general anesthesia is essential to avoid any delay in the diagnosis [25, 43]. The use MRI decreases the need for biopsy. MRI may also provide additional information regarding the anatomy of the surrounding tissues such as the presence of epidural collection, paravertebral abscess, and nerve-root entrapment [2, 9, 10].

26.2.2.5 Differential Diagnosis

The differential diagnosis of a child with back pain differs according to the age of the child and may involve both infectious and noninfectious conditions. Scheuermann's kyphosis is a noninfectious condition manifesting with back pain that is mostly seen among adolescents. Plain radiography reveals end plate irregularity, wedging of the vertebrae, and the presence of Schmorl's nodules in lumbar spine involvement. Vertebral body involvement rather than endplate erosion is more likely in patients with metastatic tumors and leukemia, and multilevel involvement is more frequent.

Other etiologies of infection must also be considered since hip or sacroiliac joint septic arthritis might mimic discitis.

26.2.3 Treatment

The initial treatment for children with suspected discitis is empirical. The goals of treatment are eradication of infection, relieving the pain, and minimization of morbidity. Even though a debate in antibiotic treatment existed, Ring et al. showed that intravenous antibiotics provides a more rapid recovery [24]. Delay in antibiotic therapy may lead to prolonged hospitalization, recurrence and progression of infection [7]. Therefore, more recently antibiotics are more widely recommended [6, 9, 29, 36]. The treatment should start via parenteral route until a favorable clinical response is noted. The treatment is then continued orally. Yet, there are no absolute recommendations to guide the duration of antibiotic treatment. One to eight weeks of parenteral antibiotics and up to 3–6 months of oral antibiotics were recommended [2, 9, 24, 33, 34].

Relative bed rest [15, 23], spinal brace or cast [2, 9, 20, 36], or a combination of both have almost universally been recommended to improve comfort and pain relief. More recent reports, however, do not recommend any form of immobilization since they have comparable results with good range of motion [10].

Paraspinal fluid collection documented in MRI do not necessitate surgical debridement in light of clinical improvement with antibiotics [7]; however, it may suggest longer durations of antibiotic therapy [9]. Surgical debridement should only be considered in the rare patient with neurological deficit due to medullary or root compression, a documented abscess who is systemically ill, or in the setting of failure of conservative treatment [7, 29].

26.2.4 Outcomes and Follow-Up

Once the symptoms are resolved, patients are followed for 1–2 years. Periodic plain radiographs will demonstrate residual disc space narrowing and persistent endplate sclerosis. Partial reconstruction of the disc height may gradually occur; however, full recovery is rare [7]. Early degenerative changes may be revealed as persistent narrowing of the disc space, sclerosis with partial fusion or complete fusion [43]. It is suggested that with the amount of the loss of the disc space 2 years after the treatment, the outcome could be predicted [9]. Disc spaces that lost more than 50 % of their height are more prone developing ankylosis or fusion. The overall rate of fusion varies from 14 to 44 % [9, 12, 16]. Long-term follow-up studies displayed 20 % of restricted spinal mobility [6].

A routine follow-up MRI is not recommended [7, 9]. After complete resolution a normal vertebral body with low disc signal will be seen on T2-weighted images [39, 44].

26.3 Vertebral Osteomyelitis

Vertebral osteomyelitis in childhood is rare when compared to discitis, and the overall incidence is approximated to be 1:250,000 [45, 46]. The patients are generally adolescents who are systemically ill with fever, pain, and muscle aches in the lumbar, thoracic, or cervical regions [2]. The mean age of children is 7.5 years [2]. Yet, a fulminant form of vertebral osteomyelitis is also reported in infancy [47, 48]. Diagnostic workup and management is more in favor of invasive procedures when compared to discitis [38, 47].

26.3.1 Pathogenesis

Vertebral osteomyelitis is thought to occur when microorganisms lodge in the low-flow, end-organ vasculature adjacent to the subchondral plate region. Three routes of infection were identified: (1) hematogenous spread, which is the most common, (2) direct implantation, and (3) contiguity [49]. Transient minor bacteremias from ear, throat, or urinary tract infections, or antecedent traumas such as minor cuts and abrasions may be associated with vertebral osteomyelitis although it is not clearly documented [2, 24, 50].

Vertebral osteomyelitis typically affects the vertebral body. Although rare, the infection can affect the posterior parts of the vertebrae as well [49, 51]. Nonsurgical treatment of vertebral arch

osteomyelitis may be more prone to incomplete resolution since the posterior elements are less vascularized [49, 51].

26.3.2 Evaluation

26.3.2.1 History

In contrast to discitis, children with vertebral osteomyelitis are more likely to be febrile and ill appearing at the time of presentation. The duration of the symptoms may be longer for children with vertebral osteomyelitis and prolonged fever may be the presenting complaint [2]. Back pain is the predominant complaint and children may present with neck, shoulder, rib, or abdominal pain depending on the affected spinal region [2].

Vertebral osteomyelitis is rarely seen among children before 3 years of age, while discitis is uncommon in children 8 or more years of age. Children of intermediate ages should be carefully evaluated [7].

26.3.2.2 Physical Examination

The clinical symptoms are variable and generally unspecific. The most common findings are difficulty walking and irritability. Back pain may extend to the abdomen, hip, leg, scrotum, or perineum and be exacerbated by spinal movements [29]. Palpation of the spine may be painful, anterior trunk decompensation may be noted. Mobility of the spine may be limited, and paravertebral or psoas muscle spasms may be present [29]. Presentation with flaccid paraplegia in a thoracic vertebral involvement with abscess and cord compression was also reported [36]. If cervical spine is affected, dysphagia and neck stiffness may be present [1]. Occurrence of spinal nerve or medullary compensation and meningitis is approximated to be 12 % [52]. Pyogenic pleural effusion and chylothorax caused by erosion of thoracic duct were also reported [48, 53].

26.3.2.3 Laboratory Workup

Similar to children with discitis, white blood cell count, ESR, and CRP provide nonspecific information for vertebral osteomyelitis. Blood cultures and aspiration or biopsy should be strongly considered in most cases; however, as with discitis, definitive results cannot always be obtained [38]. Most commonly isolated organism is *S. aureus* [2]. Cases of *S. epidermidis*, *Salmenella* group, *B. henselae*, *Streptococcus*, *Clostridium*, and *Propionibacterium acnes* are also reported [2, 36]. Cat scratch disease was also reported to cause vertebral osteomyelitis and epidural abscess [54–56]; therefore, in children with cat exposure serologic testing should be performed.

26.3.2.4 Imaging Studies

Although abnormal findings may be present in as low as 46 % of the initial radiographs [2], spine radiographs should be obtained in all children with suspected spinal infection. Radiographs demonstrate localized rarefaction of the vertebral body approximately 3 weeks after the onset of the symptoms, and later, bone destruction and osteophytic bridging. Nuclear bone scans provide nonspecific information. CT imaging is best for defining the extent of destruction of bone.

MRI has been shown to have a sensitivity of 96 % and a specificity of 93 % for the diagnosis of vertebral osteomyelitis, making it more sensitive and specific than nuclear bone scans or routine radiographs [57]. MRI is a fast, accurate, and noninvasive method that can distinguish discitis from pyogenic bone involvement, and it provides information about presence, extent, and location of abscess formation. MRI may provide sufficient detail to guide the need for invasive diagnostic procedures [2, 58, 59]. According to Donovan et al. [60], the optimal MRI technique for evaluation of spinal infection is thin-section surface coil imaging with T1-weighted images in sagittal and axial views, and sagittal T2-weighted images. Edema and pus in the marrow or disc space will appear dark on T1-weighted images and bright on T2-weighted images. A contrast-enhanced MRI may provide more detailed information [42, 57, 60].

26.3.2.5 Differential Diagnosis

Similar to discitis, differential diagnosis should include Scheuermann's kyphosis as well as

metastatic tumors and leukemia. Additionally eosinophilic granuloma is characteristic on radiographs as vertebra plana. Osteoid osteomas and osteoblastomas commonly involve the posterior elements of the vertebrae and should be considered in the differential diagnosis of vertebral arch osteomyelitis.

26.3.3 Treatment

Isolation of the pathogen is vital in planning a management strategy for vertebral osteomyelitis. Hence, percutaneous or open biopsies may be needed when blood cultures are inconclusive. The treatment should include antibiotics specific to pathogen and rest and/or immobilization [2, 6]. Antibiotics should initially be administered intravenously and later by oral route. Abscess formation may look serious, but is not an absolute indication for surgical debridement unless the patient has neurologic signs [36]. Clinical and radiological progression of the disease in spite of antibiotic therapy may necessitate a surgical intervention. Following the debridement, potential instability is assessed and if present, instrumentation and fusion is necessary for stabilization. Post-infection deformity is another indication for surgical stabilization.

26.3.4 Outcomes and Follow-Up

Most patients with vertebral osteomyelitis respond well to antibiotic treatment and functional outcome is generally acceptable [36]. Radiological outcome, however, is not as innocent. Infantile vertebral osteomyelitis is more prone to bone destruction. Vertebral fusion, block vertebra, a posterior wedge remnant resembling a hemivertebra, and anterior fusion resembling a failure of segmentation are among reported outcomes [36, 47, 48].

Since the ability of natural remodeling is limited in the presence of a previous infectious process [48], severe kyphotic deformities and instabilities should be addressed through spinal fusion.

26.4 Sacroiliac Joint Infection

Presentation and laboratory workup of children with sacroiliac joint space infection is similar to those with discitis and vertebral osteomyelitis. Pain production on direct palpation of the joint and/or with lateral compression of pelvis should drive suspicion. Bone scan demonstrating increased uptake that corresponds to sacroiliac joint may be adequate for establishing a diagnosis. Treatment consists of antibiotics and rest. If a prompt response is not obtained, a joint aspirate may be considered. Ankylosing spondylitis must also be considered when formulating a differential diagnosis [38].

26.5 Other Pathogens Involved in Spondylodiscitis

Mycobacterium tuberculosis, fungus such as *Aspergillus*, *Cryptococcus*, *Candida*, parasites causing hydatid cysts, bacteria including *Brucella* and *Actinomyces*, and even viruses are reported to cause infection in the pediatric spine [6, 7, 27, 28, 61–64]; however, they are not covered in the scope of this chapter since they cause subacute and chronic infections.

26.6 Summary

As the use of routine MRI is widespread, the previously used umbrella term spondylodiscitis is today recognized as two different manifestations of the same pathological process that have distinct epidemiologic, clinical, and radiographic features. Both entities are uncommon in childhood, and laboratory is little helpful; therefore, the clinical workup should begin with a high level of suspicion to avoid delay in diagnosis. Percutaneous core needle or open biopsies are not always necessary. Treatment should include intravenous followed by oral antibiotics, and rest and/or immobilization may be used to improve comfort especially in the initial stages. The efficacy and duration of treatment can be followed

via the levels of ESR and CRP. Functional deficits generally do not occur regardless of the radiological changes.

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Pediatric Spinal Infections (Chronic)

27

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Key Points

- Mycobacterium tuberculosis infection of the spine is common in the developing countries but increasingly being recognized in developed countries due to AIDS, cancer chemotherapy, and medical illnesses.
- Next to the lungs, the spine is the most common site affected by the organism.
- Pain, kyphotic deformity, and neurological deficit are the three main manifestations of spinal tuberculosis. In children, due to the cartilaginous nature of the vertebral bodies, the disease can cause rapid vertebral destruction.
- Short course ambulant anti-tubercular chemotherapy is the mainstay of treatment in the early and uncomplicated stages of the disease. Multi-drug management is essential to prevent drug resistance and disease recurrence.
- The natural history of spinal tuberculosis in children is different as they are prone to worsening of kyphosis throughout the remaining period of growth even after cure of the disease. "Spine-at-risk radiological signs" will allow early identification of children prone to progressive deformity, where surgical stabilization is advocated.

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- Surgical debridement, decompression, and stabilization are needed in patients with severe neurological deficits, extensive vertebral destruction, and deformity.

27.1 Introduction

Spinal infections are a rare but serious cause of back pain in children. They can be of pyogenic origin with an acute presentation or chronic of granulomatous nature (tuberculous or fungal). Tuberculosis is the most common chronic granulomatous infection of the pediatric spine and is predominantly observed in developing regions of the world [1]. Fungal infections of spine are rare and observed in children with decreased immunity due to leukemia, lymphoma, myelosuppression due to drugs, radiotherapy, or HIV infection.

27.2 Spinal Tuberculous

27.2.1 Background

It is important to recognize that childhood spinal tuberculosis differs from adult infection in both severity and clinical behavior. The pediatric vertebral bodies are very vulnerable for rapid and complete destruction during the acute phase and children frequently develop major defects of the anterior column. The pediatric spine is also more flexible making it susceptible to greater deformity and instability than adults during the active phase of the disease. While the deformity does not change after healing and consolidation in adults, children continue to exhibit a progression in deformity for the better or worse until the end of growth (Fig. 27.1a–c). As a result, children not only require surgery more frequently than adults but also require a careful follow-up until growth is complete.

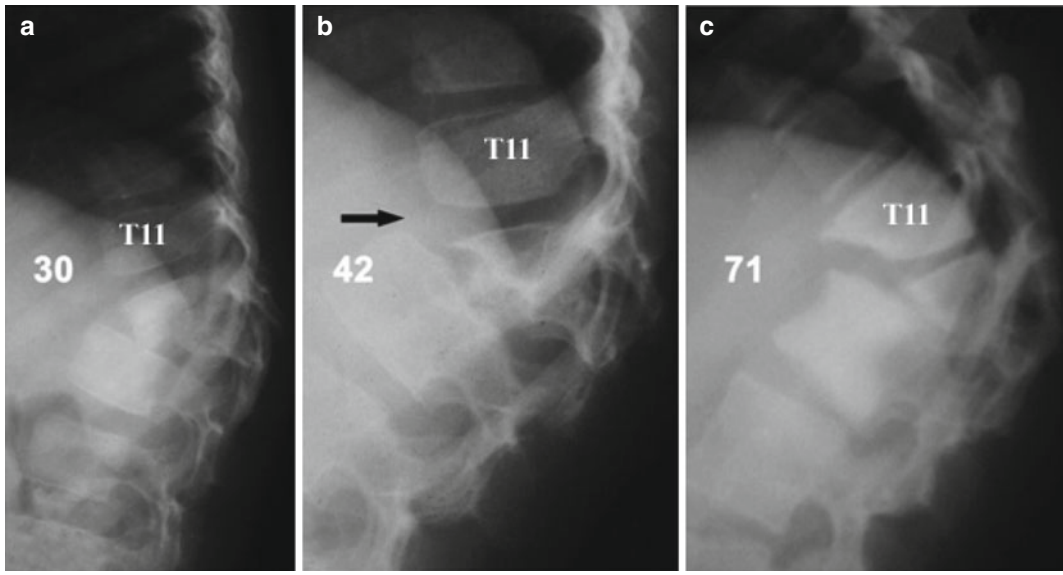

Fig. 27.1 Lateral radiographs of the thoraco-lumbar spine demonstrating kyphosis progression in a 8 year old child. (**a**) The deformity measures only 30 degrees at the completion of chemotherapy. Over a period of 5 year follow-up, the deformity gradually worsens from 42° at 5 years (**b**) to 71° at the end of 10 years (**c**) Note that the T11 vertebra (*arrow*), which was uninvolved in the disease, is showing progressive destruction due to biomechanical influence during growth

27.2.2 Epidemiology

The global burden of tuberculosis still remains huge. The World Health Organization's (WHO) Global Tuberculosis Report (2012) observes that there were an estimated 8.6 million new cases of tuberculosis and 1.3 million people died due to the disease [2]. The prevalence of tuberculosis is inversely related to the socioeconomic status of the society and the standard of public health. In developing countries, many children present late with significant deformity and neurological deficits. In developed countries, the diagnosis can be missed as it does not feature as a common diagnosis in the clinician's mind.

In developing parts of the world with a high burden of pulmonary tuberculosis, the incidence of spinal infections is expected to be proportionately high, with India and China accounting for 26 % and 12 % of the global disease burden respectively in 2012 [2]. Though the exact incidence and prevalence of spinal tuberculosis in children are not known, the incidence of pediatric spinal tuberculosis is reported as 58 % of all spinal tuberculosis in Korea, 30 % of all patients treated for spinal tuberculosis in India, and 26 % in Hong Kong [3–5].

27.2.3 Microbiology and Pathophysiology

Tuberculosis is caused by a bacillus of the Mycobacterium tuberculosis complex. There are approximately 60 known species among the *Mycobacterium* genus but only a minority of these cause human tuberculosis (M. tuberculosis being the most common). Vertebral infection by the bacillus results from hematogenous dissemination from a primary focus elsewhere in the system, commonly the lungs and the kidneys. Spread of the organism can also occur through the lymphatics from viscera to the adjacent vertebral segments (e.g., pulmonary tuberculosis can spread to the thoracic spine).

Following the infection in the vertebral marrow, the inflammatory response is characterized by chronic accumulation of macrophages and monocytes. The tubercle bacilli are phagocytosed and their lipid is dispersed throughout the cytoplasm of macrophages, transforming the macrophages into *epitheloid cells*, which are characteristic of the tuberculous reaction. Another characteristic feature of tuberculous lesion is the presence of *Langerhans giant cells*, which are formed by the coalescence of a number of epitheloid cells. The typical histopathological lesion of tuberculosis is called the *tubercle*, which is formed by the conglomeration of macrophages, epitheloid cells, Langhans giant cells, lymphocytes, and inflammatory exudate. With progressive destruction, caseation necrosis occurs in the center of the tubercle. Adjacent tubercles then coalesce to form a large abscess and since it is a chronic infection, the acute features of inflammation like warmth and redness are absent (*cold abscess*).

27.2.3.1 Clinical Pathology

The most common pattern of tubercular spinal infection in adults is the "paradiscal" type, where the bacilli lodge in the sub-chondral marrow on either side of the disc as the disc is avascular. In children, the disc retains its blood supply till approximately 9 years of age and so the bacilli affect and destroy the vertebral body and disc simultaneously ("centrum" or "complete" type) (Fig. 27.2a–f). Due to the weaker immune response of the child and cartilaginous nature of the vertebral body, extensive vertebral destruction and exuberant abscess formation is more common in children [6]. The other types of spinal tuberculosis are the anterior type (abscess formation beneath the anterior longitudinal ligament), posterior type (isolated involvement of posterior elements), and the non-osseous type (extensive abscess formation with very little bony destruction).

27.2.4 Clinical Presentation

Unlike pyogenic spondylitis, tuberculous lesions have a much more insidious onset and the clinical symptoms often develop over a period of 1–2 months. Back pain localized to the affected

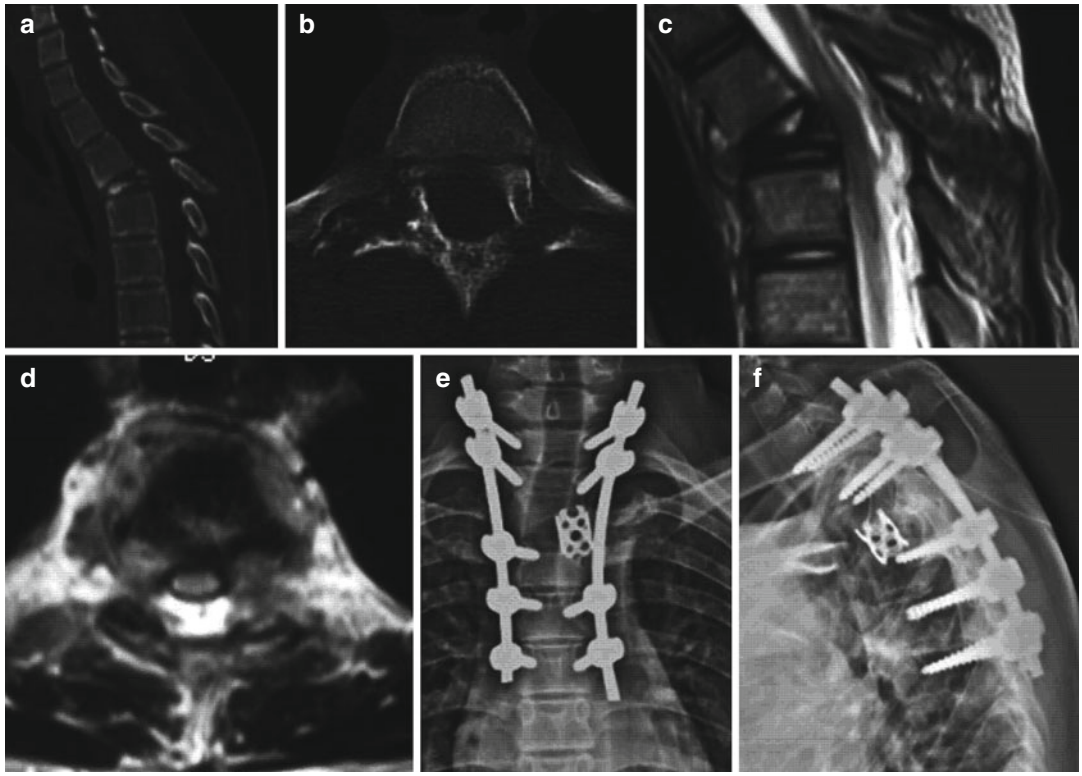

Fig. 27.2 Centrum (complete) type of tuberculosis in a 10-year-old child. Sagittal CT (**a**) and coronal CT (**b**) images show complete collapse of the T3 vertebra with involvement of the pedicles and lamina on the right side. Sagittal and axial MRI images (**c**, **d**) show complete collapse of the T3 vertebral body with prevertebral abscess formation. Unlike a classical adult paradiscal type of spinal tuberculosis, the centrum type is characterized by vertebral body destruction with intact adjacent disc spaces. The lesion has been treated by posterior stabilization, corpectomy, and reconstruction with cage as shown in the AP and lateral radiographs of the thoracic spine (**e**, **f**)

site and aggravated with spinal movements is the usual presenting feature. In the initial stages, the pain is due to inflammation, distension of the capsule by the abscess and pressure on neighboring structures. Later with development of instability, the pain can become quite severe. The affected child may need to support his trunk by placing the hands on the couch while sitting (*Tripod sign*) or hold the neck by the hands when the cervical spine is affected (Fig. 27.3a, b). Constitutional symptoms of malaise, loss of appetite and weight, evening rise of temperature and night sweats is also observed in up to 60 % of patients [7].

A paravertebral cold abscess is a diagnostic feature of spinal tuberculosis. It may be clinically evident, either in the paraspinal area or the abscess may tract distally along the perineural, perivascular, intermuscular, subpleural, subperitoneal, and natural areolar tissue spaces to present remotely away from the vertebral lesion. The common areas of presentation include the retropharyngeal abscess from a cervical lesion, a paravertebral abscesses in the thoracic spine tracking along the intercostal neurovascular bundle along the chest wall, and pre-sacral and pelvic retroperitoneal abscess from a lumbar lesion. A psoas abscess is common in thoracolumbar lesions below the diaphragmatic attachment to the spine. Psoas abscesses are pathognomonic of spinal tuberculosis and can present bilaterally. They can present externally in the inguinal region, at the Petit's triangle (See Fig. 27.3b), in the ischiorectal fossa, or in the buttock under gluteus maximus (Fig. 27.4a–d).

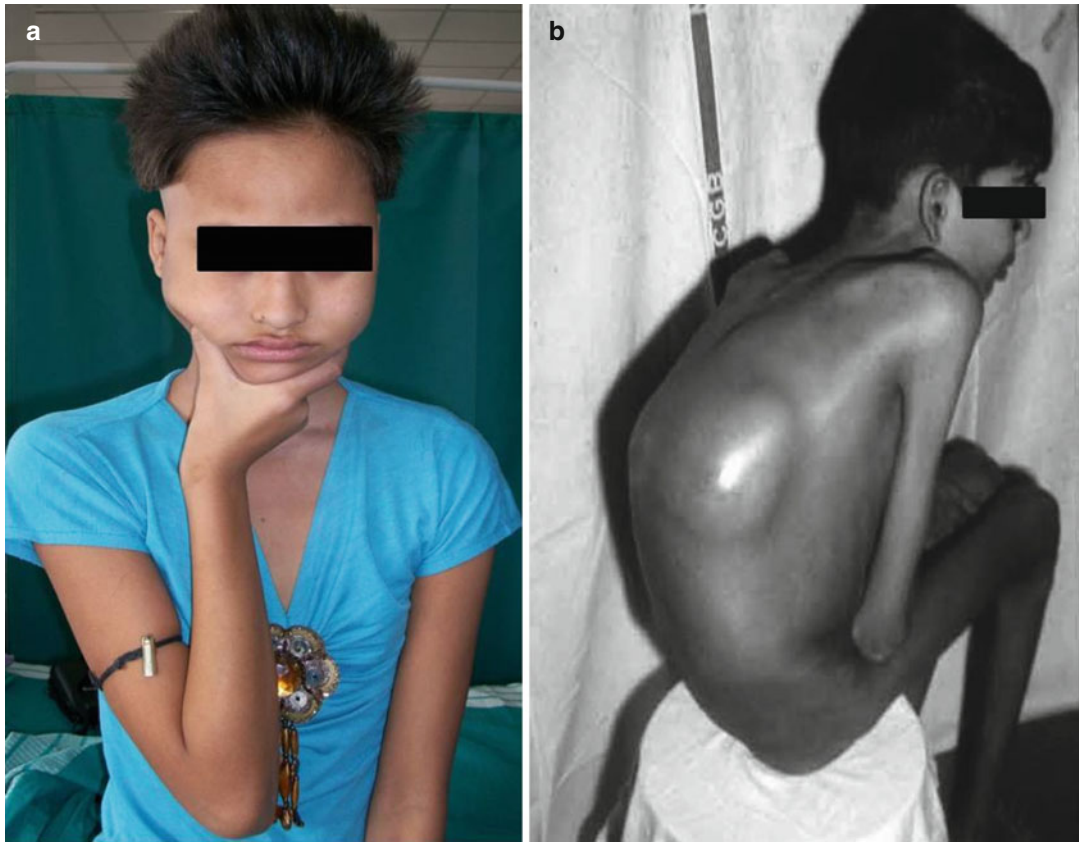

Fig. 27.3 (**a**) A 13-year-old girl with upper cervical tuberculosis and cervical instability holding her neck because of severe instability pain. (**b**) Another 9-year-old boy with prominent thoracolumbar kyphosis with a large lumbar abscess formation. Note that the patient is supporting his trunk with his elbows and the generalized wasting of the muscles. (**b**) (Courtesy: Prof. V.T. Ingalhalikar, India)

Deeper abscesses are not clinically palpable but can cause pressure symptoms. A retropharyngeal abscess arising from cervical tuberculosis can produce dysphagia and dysphonia. In children with high thoracic spinal involvement with paraspinal abscess formation, the abscess in the prevertebral region may cause significant bronchial compression. These symptoms may simulate bronchial asthma as the dyspneic symptoms exacerbate when the patient lies down at night.

While most abscesses resolve gradually with chemotherapy treatment, they may also rupture with neglect leading to the formation of a sinus. The sinus may heal spontaneously with medical treatment after all the necrotic material is discharged or may persist if there is any residual infection or secondary pyogenic infection. The tubercular pus is white or light gray in color, watery, and has no specific smell unlike a pyogenic abscess.

27.2.4.1 Neurological Involvement

Neurological compromise occurs in up to 30–75 % of the patients with spinal tuberculosis [7–9]. Though children have more severe destruction, they also have a lesser incidence of neurological involvement, probably due to the relative larger canal diameter and more flexibility of the spine. Cervical tuberculous lesions manifest with quadriparesis but since the thoracic and thoracolumbar regions are commonly affected, lower limb weakness with bladder and bowel involvement is more common. Initial symptoms are incoordination and clumsiness while walking

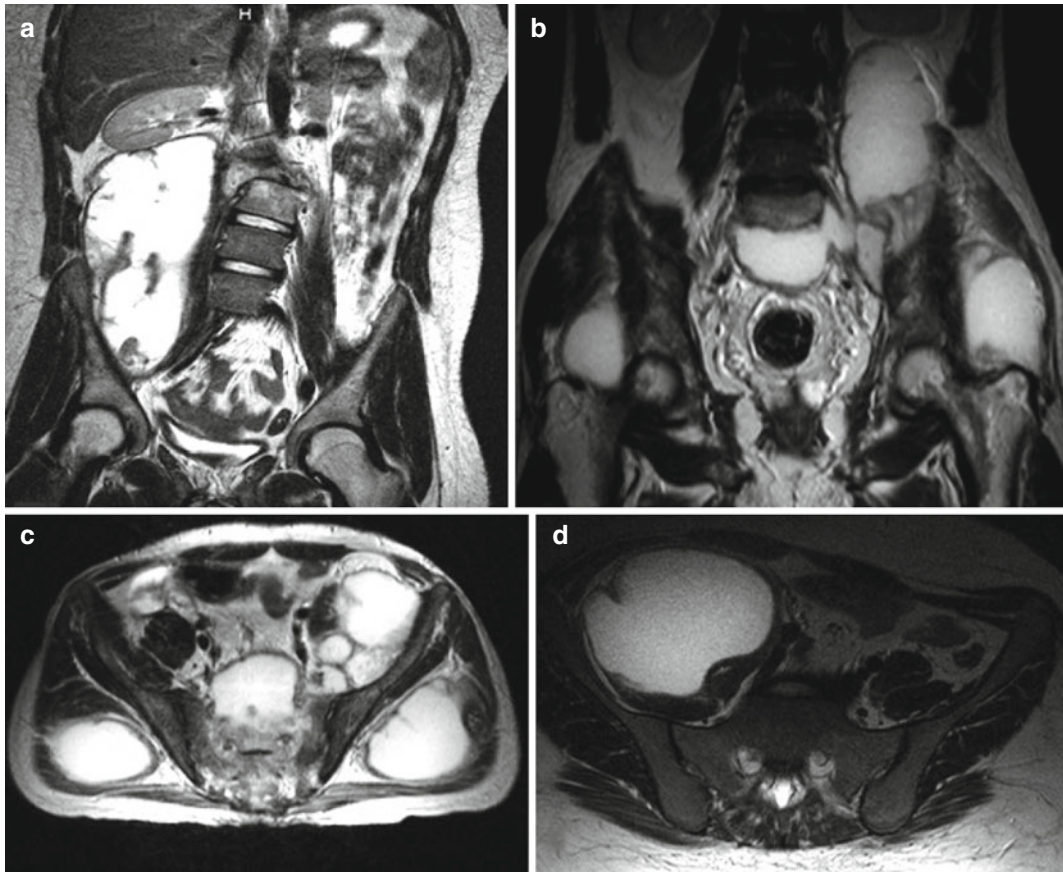

Fig. 27.4 Tubercular cold abscess can be present in multiple locations depending on the pathway of its spread across areolar spaces. In this 40-year-old patient, who presented with back and bilateral gluteal pain, extensive cold abscess formation was observed. Coronal T2 MRI sections of the lumbar spine (**a**, **b**) showing a right retroperitoneal psoas abscess (**a**), bilateral trochanteric abscesses (**b**). Axial T2 sections through the sacrum and pelvis shows multiple abscesses in iliac fossa (**d**), presacral and sub-gluteal regions (**c**), and large pre-sacral abscess (**d**)

which slowly progresses to paraplegia and loss of sphincter control.

Children can present with neurological involvement both in the active and healed phase of the disease. In active lesions, it is due to the result of direct compression of the spinal cord by an abscess, inflammatory granulation tissue, a dislodged sequestrum, or canal compromise due to instability. In the healed disease, it occurs after many years and is usually due to stretching of the cord over a bony ridge at the apex of the deformity.

27.2.4.2 Kyphotic Deformity

The etiology and progression of kyphosis is different in the active and healed phase of the disease. Tuberculosis affects and destroys the anterior structures of the vertebral column in more than 90 % of patients. Collapse of the vertebral body is evident as a localized kyphotic deformity. Involvement of two or three adjacent vertebral bodies manifests as a sharp, angular kyphosis called the *gibbus*. Chemotherapy will cure the disease but vertebral collapse will continue until the healthy vertebral bodies in the region of the kyphosis meet anteriorly and consolidate. The severity of collapse during the active phase is mainly influenced by the *severity of vertebral destruction*, *the level of the lesion*, *and the age of the patient* [10].

During the active phase, the deformity increases in proportion to the severity ranging about 25–35° for each vertebral body loss in thoracic and thoracolumbar lesions. The kyphotic collapse is less in lumbar lesions due to the lordotic nature of the lumbar spine, large size of the intervertebral discs, and the sagittal orientation of the facet joints which allows vertical subsidence. In the thoracic and the thoracolumbar regions, the deformity tends to be more severe due to the inherent kyphotic nature of the thoracic spine and the coronal orientation of the facets which lead to subluxation and kyphosis. Deformities in children less than 10 years of age have been observed to have greater deformity than those over 10 years of age due to soft vertebral bodies, weaker posterior stabilizing structures, and the secondary increase in deformity during the adolescent growth spurt [11, 12].

In a long-term follow-up of 15 years of 63 children, Rajasekaran reported three types of collapse and healing of the anterior column with different implications for deformity progression during the period of growth [10, 13, 14] (Fig. 27.5a–c). Type A healing was seen in minimal lesions and paradiscal type of involvement, where the facet joints were intact and there was large area of contact of vertebral bodies anteriorly. These patients showed minimal deformity in the active phase and frequently an improvement during the growth period (Fig. 27.6a–c). Type B healing was seen when the vertebral body loss is equivalent to the loss of one vertebral body. Here during the process of collapse, the facet joint at the level of destruction subluxed or completely dislocated. The superior vertebra rotates during the process of descent so that its antero-inferior margin comes into point contact with the superior surface of the inferior normal vertebra. This resulted in growth depression at the point of contact and the deformity could progress by up to further 30° during the growth period (Fig. 27.7a–c). Type C restabilization occurred when the vertebral body loss increases to more than "two." The large anterior column defect necessitates the dislocation of two or more facet joints before anterior column restabilization can occur. The superior normal vertebra rotates 90° so that the anterior surface of superior vertebra comes into contact with the superior surface of the inferior vertebra. This was seen frequently in children less than 7 years

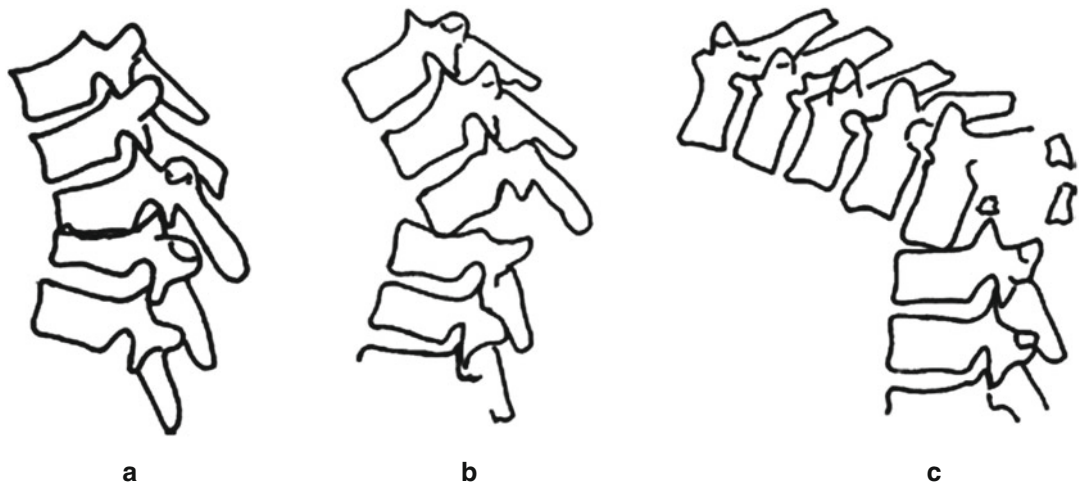

Fig. 27.5 Following destruction of anterior column, restabilization and healing occurs by one of the three methods. (**a**) In patients with minimally destroyed vertebrae with intact facet joints, restabilization occurs with wide contact area. (**b**) In patients with dislocation of single facet joint, restabilization occurred by point contact. (**c**) In patients with loss of two or three vertebrae, the facets dislocate at multiple levels and the superior segment rotates by 90° so that its anterior surface can rest on the superior surface of inferior vertebra

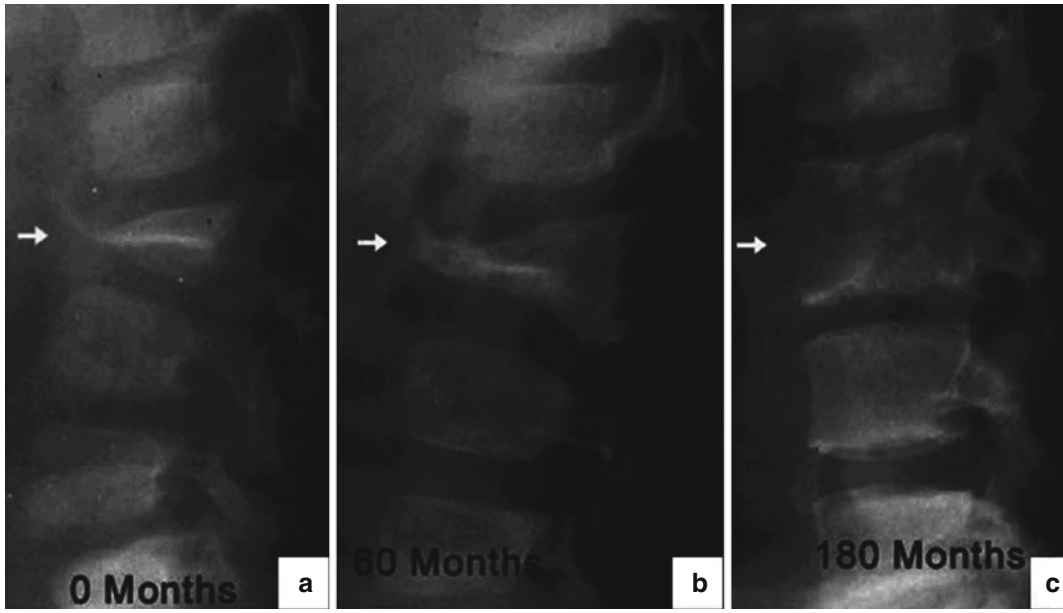

Fig. 27.6 Type A restabilization. In patients with partially destroyed vertebrae, restabilization occurs with wide contact area and the kyphosis gets corrected automatically or remains unchanged. (**a**) Lateral radiograph of thoracolumbar spine showing a healed tuberculous lesion at L2-3 which has healed into a triangle shaped fusion mass (*arrow*). The bony fusion has occurred over a wide contact area with intact facet joint. (**b**) During growth, the fusion mass shows "accelerated growth phenomenon" (*arrow*) and achieves spontaneous improvement in vertebral height at 60 months. (**c**) At 180 months, the fusion mass has become a large "single" vertebra (*arrow*) with two subjacent pedicles

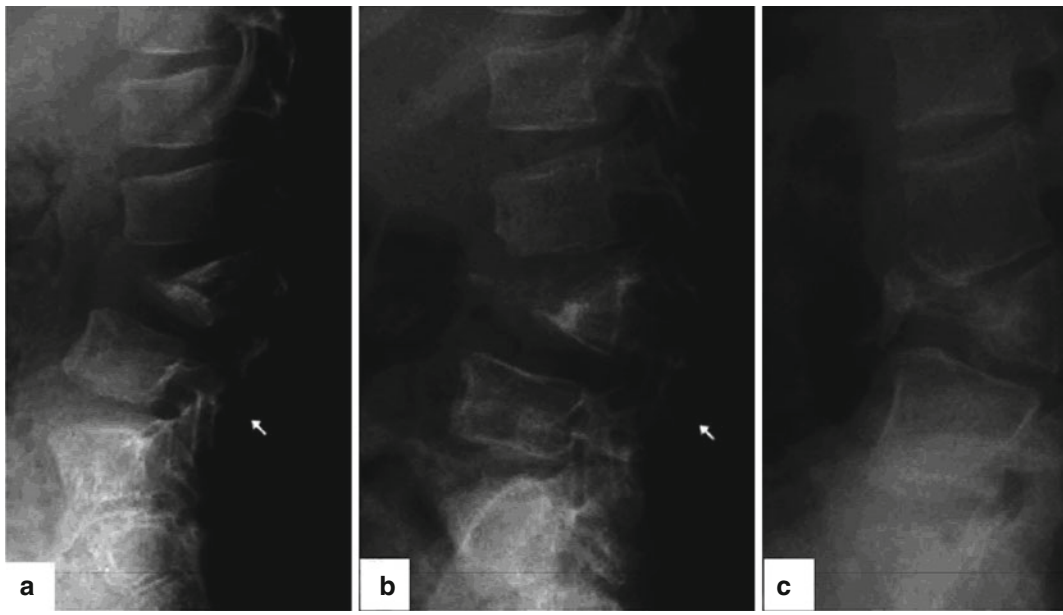

Fig. 27.7 Type B restabilization. Type B healing is seen when the vertebral body loss is between 1 and 1.5. (**a**) Lateral radiograph of the lumbar spine shows significant destruction following L3-4 spondylodiscitis which has resulted in local kyphosis. (**b**, **c**) Lateral radiographs performed at 60 and 180 months show that the facet joint at the level of destruction is dislocated (*white arrow*) during the process of collapse and the superior L2 vertebra gradually rotates by 90 degrees so that its antero-inferior margin comes into point contact with the superior surface of the fusion mass

of age with thoracolumbar disease. In children with multiple vertebral body destruction, a peculiar pattern of collapse is seen which has been termed as "Buckling Collapse" [14, 15]. Dislocation of facet joints occurs sequentially at multiple levels leading to a kyphosis of more than 120° and the entire spine is converted to two large compensatory curves. Many vertebral segments become horizontally oriented with stress shielding of their growth plates. Longitudinal overgrowth of the vertebral segments is noted leading to stretching of the spinal cord at the apex of the kyphosis with possible secondary late-onset paraplegia. Risk factors for "buckling collapse" included an age of less than 7 years at the time of the disease, thoracolumbar involvement, loss of more than two vertebral bodies, and presence of radiographic spine-at-risk signs (Fig. 27.8a–c).

Unlike adults in whom the deformity is static after cure of the disease, post-tuberculous kyphosis in children is a dynamic deformity with variable progression during growth. Three different patterns of progression have been observed depending on the pattern of healing [10]. Type 1 progression, where worsening of deformity occurs during growth, is seen in 39 %. This increase can occur after a lag period of few years after the disease control. As a result, a severe increase in deformity may be missed if the child is not followed-up carefully till the completion of growth. Forty-four percent of children had a Type II progression where after an increase in deformity during the active phase, the deformity showed a progressive and spontaneous correction. This was mostly observed with Type A healing pattern and in children younger than 7 years. Type III progression, where there was no major change during growth, was seen in the remaining 17 % who either had a minimal disease or a lower lumbar lesion (Fig. 27.9a, b).

Four radiological signs which indicate spinal instability have been identified by Rajasekaran to predict the risk of late and progressive development of deformity in childhood spinal tuberculosis [10, 13]. They basically indicate the presence of facet joint dislocation and disruption of the posterior arch. These signs are easy to identify in radiographs, appear early in the course of the disease and are useful to identify children at risk for progression so that surgical stabilization can be suitably advocated. These four "spine-at-risk" signs are: (1) dislocation of one or more facet joints in the lateral view, (2) retropulsion of the diseased vertebra, (3) lateral translation seen in antero-posterior view, and (4) the "Toppling Sign" (Fig. 27.10a–d).

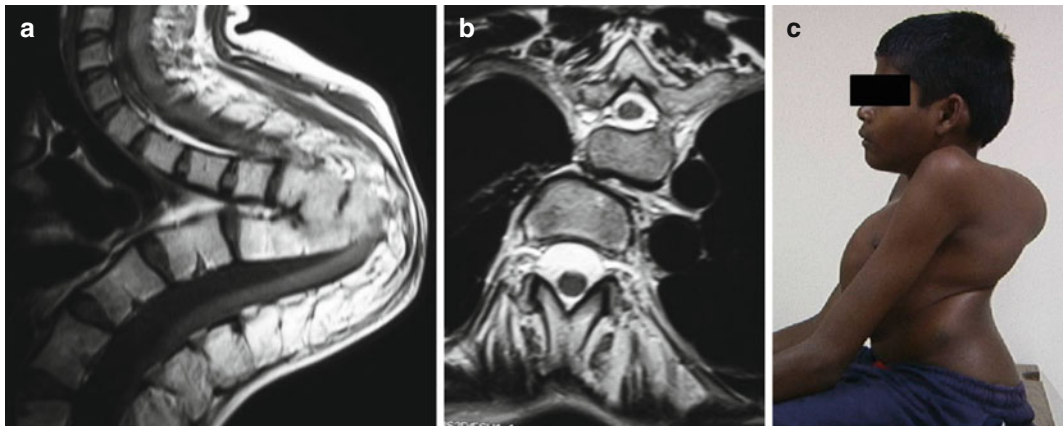

Fig. 27.8 Buckling collapse due to neglected tubercular kyphotic deformity in a child. The sagittal MRI of spine shows buckled spine with two long spinal segments lying on each other (**a**). The spinal cord is stretched and compressed at the apex of the kyphosis. The axial MRI image shows two vertebral segments at the same level straddled over each other because of buckling (**b**). Clinical picture of the same patient shows the shortened trunk because of buckling (**c**)

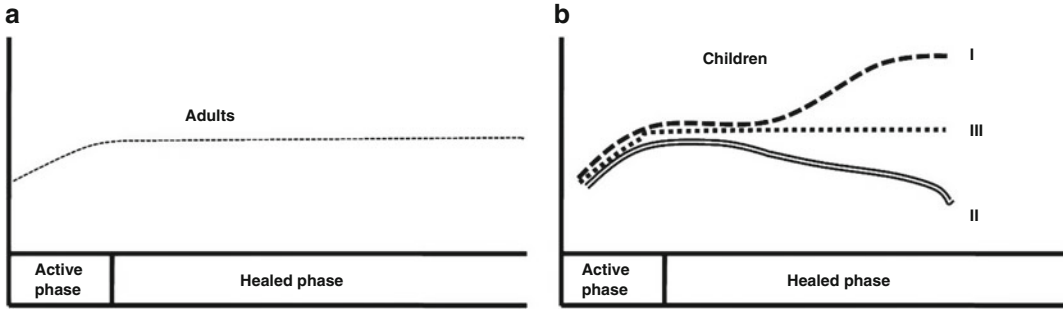

Fig. 27.9 Deformity progression in healed tuberculosis in adults and children. (**a**) In adults, the deformity remains the same during the healed phase. (**b**) In children, the deformity can either worsen (Type I), remain static (Type III), or improve (Type II) during the healed phase

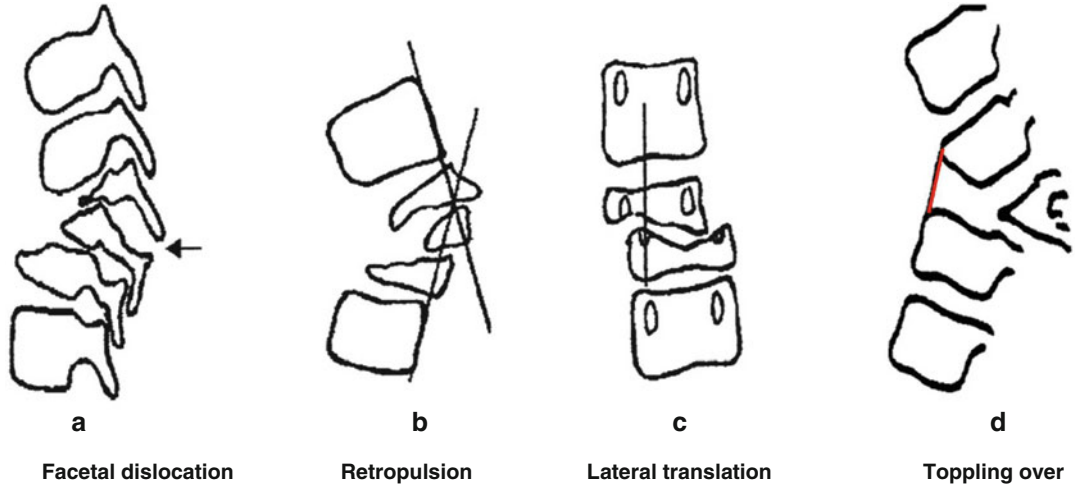

Fig. 27.10 Rajasekaran's 'Spine at risk' radiological signs. (**a**) Separation of the facet joint. The facet joint dislocates at the level of the apex of the curve, causing instability and loss of alignment. In severe cases the separation can occur at two levels. (**b**) Posterior retropulsion. This is identified by drawing two lines along the posterior surface of the first upper and lower normal vertebrae. The diseased segments are found to be posterior to the intersection of the lines. (**c**) Lateral translation. This is confirmed when a vertical line drawn through the middle of the pedicle of the first lower normal vertebra does not touch the pedicle of the first upper normal vertebra. (**d**) Toppling sign. In the initial stages of collapse, a line drawn along the anterior surface of the first lower normal vertebra intersects the inferior surface of the first upper normal vertebra (*red line*). 'Tilt' or 'toppling' occurs when the line intersects higher than the middle of the anterior surface of the first normal upper vertebra

27.2.5 Diagnostic Investigations

Diagnosis may be difficult in children, especially in the early stages of the disease. Systemic symptoms of tuberculosis include malaise, easy fatigability, weight and appetite loss, and sometimes low grade fever. Clinical signs of frank sepsis are uncommon.

27.2.5.1 Laboratory Investigations

Anemia and elevated erythrocyte sedimentation rate (ESR) are the two common abnormalities noted in blood investigations. ESR may be markedly elevated (>70 mm/h) and serial ESR measurements are helpful in assessing the response to treatment. ESR and low hemoglobin levels however lack specificity [16]. A positive Mantoux (tuberculin skin) test merely indicates cell-mediated immune response due to a previous tuberculous infection and in endemic regions, the test can be positive even in patients without active tuberculosis. Its diagnostic value is useful only in regions where tuberculosis is rare. Polymerase

chain reaction (PCR) analysis from infected tissue is considered very sensitive and specific for the diagnosis of spinal tuberculosis [17].

27.2.5.2 Bacterial Cultures

Bacterial culture of the infected tissue is useful to confirm the diagnosis and to acquire antibiotic sensitivities to guide therapy. Since spinal tuberculous infection is paucibacillary (less bacilli in infected tissues), it is essential to culture material from deep structures such as bone and abscess walls. Culture media such as BACTEC™ (Becton-Dickinson and Co., USA) now are the standard culture media [18]. An important advantage is that they allow drug susceptibility assessment. This helps in identifying drug resistant strains and start early alternate second-line medications.

Histopathology and Microbiology

The confirmation of tuberculosis infection is through identification of bacillus in the tissue or by histological confirmation of typical tubercles in the infected tissue. The typical histopathological findings are large caseating necrotizing granulomatous lesions with epitheloid and multinucleated giant cells with lymphocytic infiltration [19].

27.2.5.3 Imaging Studies

Earliest features observed on plain radiographs are vertebral osteoporosis, narrowing of the disc space and indistinct paradiscal margin of vertebral bodies. With progress of the disease, destruction is associated with vertebral collapse, kyphosis, and sagittal or coronal instability. In the cervical spine, the prevertebral soft tissue shadow can be enlarged due to distension of the abscess in the retropharyngeal region. In the thoracic spine, the cold abscess is visible on anteroposterior plain radiographs as a fusiform or globular radiodense shadow (*bird's nest appearance*) (Fig. 27.11a–e). In children, attention should be paid toward the "spine-at-risk signs" as it indicates chances of deformity progression.

Longstanding abscesses may produce concave erosions around the anterior surfaces of the vertebral bodies called *the aneurysmal phenomenon* (See Fig. 27.11). In healed disease, vertebral bodies sometimes have spontaneous fusion, simulating a hemivertebra. Autofusion following healing is seen as a single vertebra with two subjacent pedicles (*vertebra-within-a-vertebra phenomenon*) (See Fig. 27.11). In children with severe deformity, the vertebral bodies just above the kyphosis lie more horizontally and achieve more increase in height than width due to the weight-relieving effect (*horizontalization of vertebral body*) (See Fig. 27.11).

CT and MRI can detect lesions at an earlier stage. CT is useful in assessing accurately the extent of bony destruction, early identification of posterior element involvement, and in tuberculosis of certain regions like the craniovertebral and cervicodorsal junction, the sacro-iliac joints, and the sacrum, which are not easily defined in the radiographs (Fig. 27.12a–c). MRI is the gold standard investigation for demonstrating the extension of disease into soft tissues, the spread of tuberculous abscess, identification of multilevel non-contiguous involvement, and evaluating neural compression (Fig. 27.13a–e). MRI with contrast is also helpful in differentiating vertebral lesions from pyogenic and other non-infectious causes.

27.2.6 Management Principles

Multi-drug anti-tubercular chemotherapy in adequate dose and duration forms the foundation for the treatment of spinal tuberculosis. Attention to good nutrition, brace to support the spine and prevention of neurological involvement and deformity are other essential aspects of treatment.

27.2.6.1 Chemotherapy

The first-line drugs (Isoniazid, Rifampicin, Ethambutol, Pyrazinamide, and Streptomycin) are the most effective group of agents active against tuberculosis (Table 27.1). Short-course chemotherapy has many inherent advantages such as improved patient compliance, lower failure rates, lower cost, and a lower incidence of drug resistance [20–22]. It is important to establish that there is adequate drug sensitivity and the patient is compliant in consuming the drugs in both proper dosage and duration. The

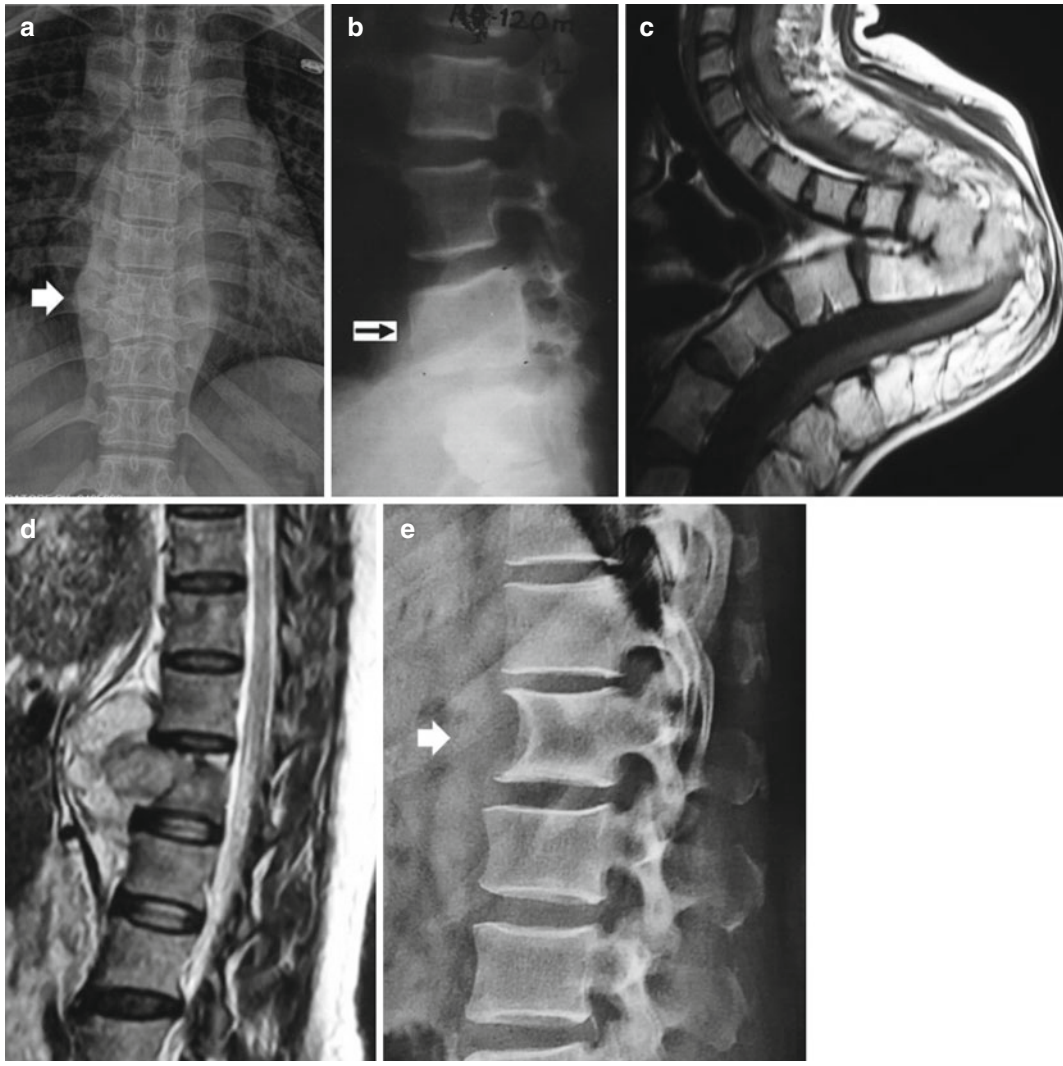

Fig. 27.11 Radiological appearances in spinal tuberculosis. (**a**) Bird's nest appearance: In the anteroposterior radiograph of the spine, the paravertebral abscess formation is seen as a fusiform shaped radiodense shadow (*thick white arrow*). (**b**) Vertebra within vertebra appearance – Healing of a paradiscal type of tuberculosis results in fusion of two adjacent vertebral bodies, which is seen as a single vertebra with two subjacent pedicles (*black arrow*). (**c**) Horizontalisation of vertebral body in buckling collapse – Chronic buckling collapse can lead to increase in the supero-inferior height of the horizontally placed vertebral bodies. (**d**, **e**) Aneursymal phenomenon – Sagittal MR image shows prevertebral abscess formation under the anterior longitudinal ligament. Such longstanding abscesses can cause erosion of the anterior surface of the vertebral body (E) (*thick white arrow*) similar to an aortic aneurysm

WHO has provided guidelines for the type and duration of anti-tuberculous chemotherapy and considers spinal tuberculosis to be severe extrapulmonary (category 1). Treatment is advised for 6 months and in cases of relapse or treatment failure, treatment is prescribed for 9 months (Category 2). The currently recommended first-line drug regime is four-drug therapy. This includes Isoniazid 5 mg/kg/day, Rifampicin 10 mg/kg/day, Pyrazinamide 20–25 mg/kg/day, and Ethambutol 15 mg/kg/day for 2 months (Intensive phase) followed by Isoniazid and Rifampicin for four (Category 1) to 7 months (if Category 2) (Continuation Phase). In children,

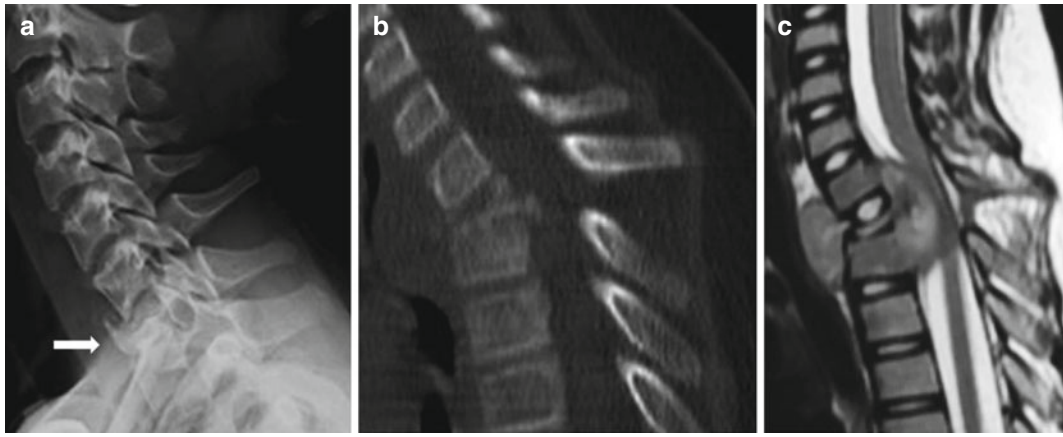

Fig. 27.12 CT and MRI are useful in evaluating junctional lesions not easily visualised in the radiograph. Here there is collapse of the C7 vertebral body (*white arrow*) which is noted in the lateral radiograph (**a**). However the sagittal CT and MR image shows the clear extent of vertebral body damage, kyphosis, abscess formation, and cord compression (B, C)

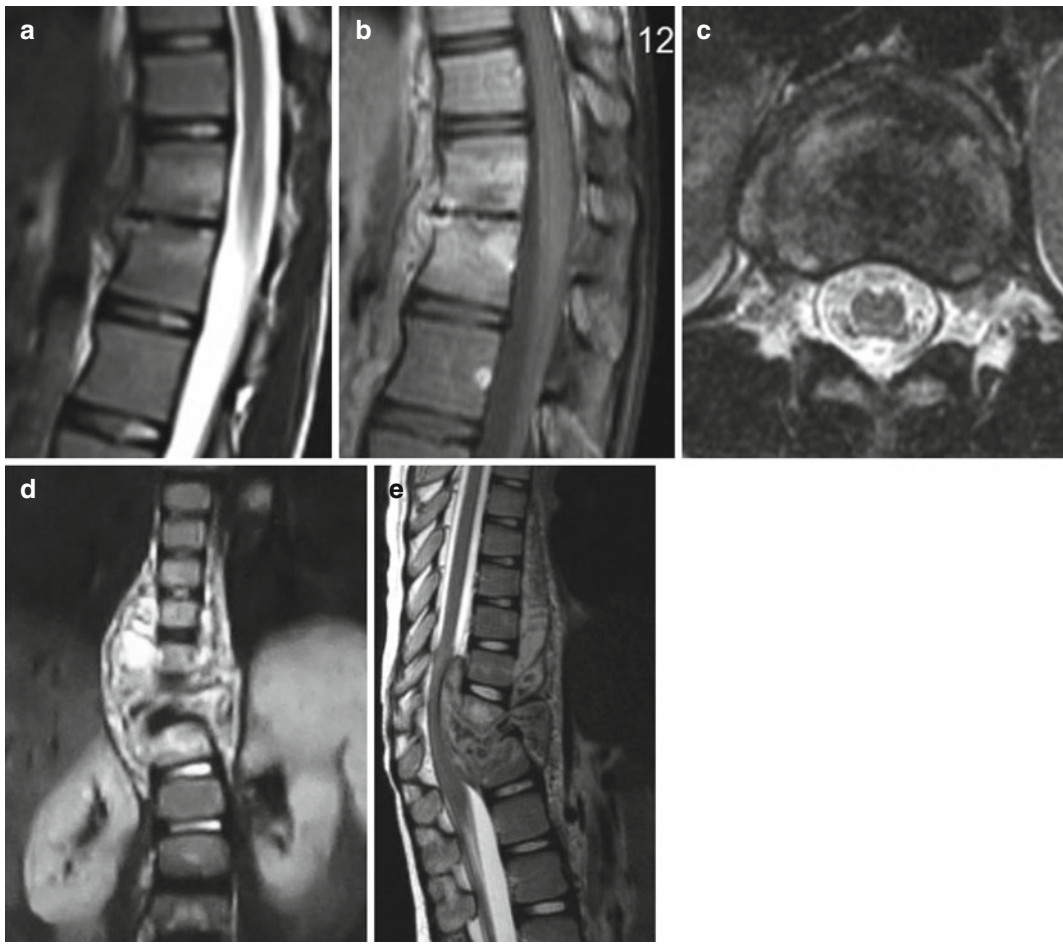

Fig. 27.13 MRI features of spinal tuberculosis include bright signal on T2-weighted images (**a**) and low signal on T1-weighted images (**b**) in the affected vertebral bodies, end plate disruption (**b**, **c**), the presence of septate pre- and paravertebral or intra-osseous abscess with a subligamentous extension and epidural abscess formation causing cord compression (**d**, **e**)

Table 27.1 Chemotherapeutic agents against tuberculosis: first-line drugs

Name of the drug	Mechanism of action	Important adverse effects
Isoniazid 5 mg/kg/day	The drug penetrates the cell wall and inhibits synthesis of mycolic acid. It is bactericidal against rapidly dividing bacilli and bacteriostatic against resting bacilli.	Chronic intake of the drug can result in peripheral neuropathy. This can be prevented by concurrent administration of pyridoxine (vitamin B6).
Rifampicin 10 mg/kg/day	After entering the bacillus, the drug blocks DNA-dependent RNA polymerase thus affecting protein synthesis. It is highly bactericidal against slowly multiplying bacteria.	Orange discoloration of body secretions including sweat and urine, hepatotoxicity, skin rashes and abdominal pain, flu like symptom.
Pyrazinamide 20–30 mg/kg/day	The drug disrupts the membrane potentials, thus inhibiting membrane transport function of the bacilli. Very effective drug; it is bactericidal and can penetrate even macrophages harboring the mycobacteria.	The drug can result in elevated serum uric acid levels manifesting as arthralgia.
Ethambutol 15–20 mg/kg/day	The drug interrupts cell wall formation and is bacteriostatic against rapidly multiplying bacteria.	It can result in optic neuritis, which manifests as reduced visual acuity, central black spots and loss of ability to differentiate color. If identified in early stages, it is potentially reversible. So it is not administered in children because children may not be able to report visual disturbances.
Streptomycin 15–20 mg/kg/day	Streptomycin affects protein synthesis thus disrupting ribosomal function and cell wall formation. It is bactericidal against the rapidly dividing extracellular bacteria.	It can affect alterations in renal function and disturbances in vestibule-cochlear function.

Ethambutol is not prescribed as it may cause optic neuritis. The response to chemotherapy must be carefully assessed both clinically and radiologically. Failure of adequate response demands investigation for drug resistance or poor patient compliance.

Second-line drugs are less effective, more toxic, and more expensive. Examples of second-line drugs include Ciprofloxacin, Levofloxacin, Kanamycin, Capreomycin, Cycloserine, etc. (Table 27.2). Usage of second-line drugs must involve a physician well versed in the treatment of resistant tuberculosis and the management of the side effects and toxicity of the second-line drugs.

27.2.6.2 Operative Management

While chemotherapy is the mainstay of treatment, surgery has a larger role in children to prevent deformity, neurological deficit, and chronic pain. The main indication for surgery in children would be:

1. Lesions with significant vertebral body loss
2. Severe lesions of the cervical spine
3. Junctional lesions of occipitocervical, cervicothoracic, and thoracolumbar regions (Fig. 27.14a–f)
4. Presence of "spine-at-risk" radiological signs
5. Impending or presence of neurological deficit
6. Documentation of progressive deformity

The standard operative procedure initially advocated was "universal anterior radical excision surgery" [20]. Though radical surgery provided good disease clearance, complications due to morbidity of approach, vascular complications, prolonged surgeries, neurolog-

Table 27.2 Chemotherapeutic agents against tuberculosis: second-line drugs

Drug	Dosage	Mechanism of action	Adverse effects
Para-aminosalicylic acid	10–12 g/day	Bacteriostatic drug	Gastrointestinal disturbances, drug hypersensitivity, hepatotoxicity, hypernatremia
Ethionamide	15–20 mg/kg/day	Bacteriostatic agent	Gastrointestinal disturbances, drug hypersensitivity, hepatotoxicity, metallic taste
Cycloserine	0.5–1 g/day	Penetrates blood brain barrier and is distributed widely in CSF	Psychosis, convulsions, depression, headaches, rash and drug interactions
Kanamycin	12–18 mg/kg/day	Aminoglycoside (needs parenteral administration), bactericidal	Toxicity Auditory Vestibular Renal
Amikacin	12–18 mg/kg/day	Aminoglycoside (needs parenteral administration), bactericidal	Toxicity Auditory Vestibular Renal Electrolyte imbalance Dizziness
Capreomycin	12–18 mg/kg/day	Aminoglycoside (needs parenteral administration), bactericidal agent	Toxicity Auditory Vestibular Renal
Ciprofloxacin	1–1.5 g/day	Bactericidal agent. Prevents synthesis of DNA through the inhibition of DNA gyrase	Gastrointestinal disturbances, drug hypersensitivity, dizziness, headaches
Ofloxacin	5–10 mg/kg/day	Bactericidal agent	Same as Ciprofloxacin

ical deficits, and the problems of bone defects and grafting was increasingly recognized. Subsequently the "middle-path regime" was described, where chemotherapy is the mainstay of the treatment, and limited surgery aimed at debridement of infected tissues, abscess evacuation, and spinal stabilization was advised [23]. This approach is now currently adopted.

The various options for the surgical treatment include:

1. Anterior debridement, reconstruction, and anterior instrumentation
2. Anterior debridement, reconstruction-supplemented posterior instrumentation
3. Debridement, reconstruction; and instrumentation through a posterior only approach

Currently, operative treatment in spinal tuberculosis is performed to achieve debridement and drainage of large cold abscess, decompression of the spinal cord, prevention of instability, and to correct or prevent deformity. The traditional anterior approach techniques have now given way to predominantly posterior approaches due to the development of newer approaches such as transpedicular or transfacet decompression and anterior reconstructions. Pedicle screw instrumentation and anterior cages can be safely used for stability and reconstruction despite the presence of infection [24–28].

Anterior Techniques

Anterior techniques allow maximum exposure for adequate debridement of infected tissues and reconstruction of the vertebral defect.

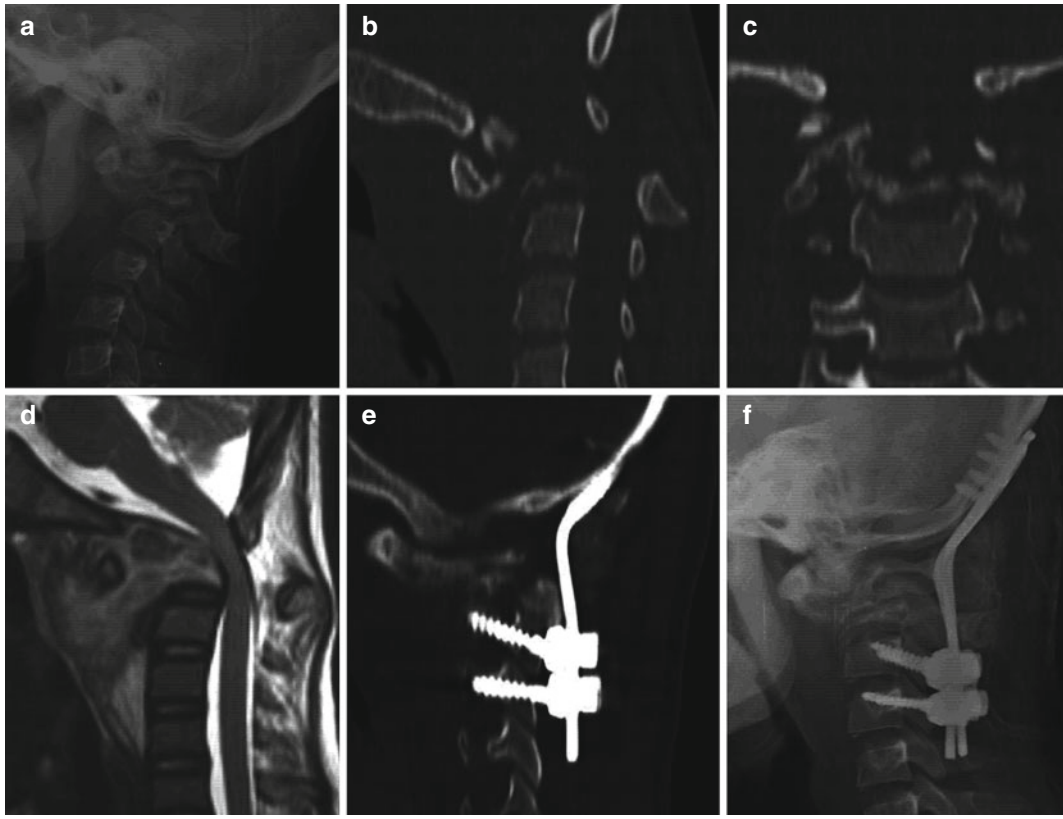

Fig. 27.14 A 13-year-old child presented with upper cervical tuberculosis. The lateral radiograph (**a**), sagittal and coronal CT (**b**, **c**) shows the complete destruction of C1 and C2 vertebra with atlanto-axial instability and dislocation. The sagittal MR image shows extensive prevertebral abscess formation and cord compression at the level of C1 (**d**). The patient has been treated by posterior decompression, abscess drainage, correction of instability and fusion from occiput to C4 (**e**, **f**). This case is a good example to demonstrate that despite extensive destruction, the anterior bone gradually reforms new bone with adequate antitubercular chemotherapy and posterior stabilization

Debridement alone without reconstruction of the anterior column is rarely done as it does not prevent the development or progression of deformity. Reconstruction of the anterior column can be done with either autograft, structural allograft, or a cage, with or without anterior instrumentation (Fig. 27.15a–e). While autografts from iliac crest and ribs are commonly used for achieving interbody fusion, titanium cages with bone grafts are also used more frequently as they provide secure, accurate, and dependable deformity correction [24–28]. Anterior instrumentation with plate or rods provides additional stability and prevents graft collapse and dislodgement. Single rod-screw system suffices in children. If the vertebral body is too small to accept a screw, then a combined posterior stabilization either with pedicle screws or Hartshill rectangle can be used. Concomitant posterior instrumentation after anterior reconstruction is indicated to protect the anterior bone graft, prevent graft-related complications in long segment disease and correct kyphosis (Fig. 27.16a–h). Combined anterior and posterior procedures can be performed at the same time or can be staged appropriately.

Posterior Techniques

In the last decade, the development and refinement of posterior surgical techniques allow the performance of adequate decompression, debridement, reconstruction, and stabilization to be achieved by an all posterior approach. The

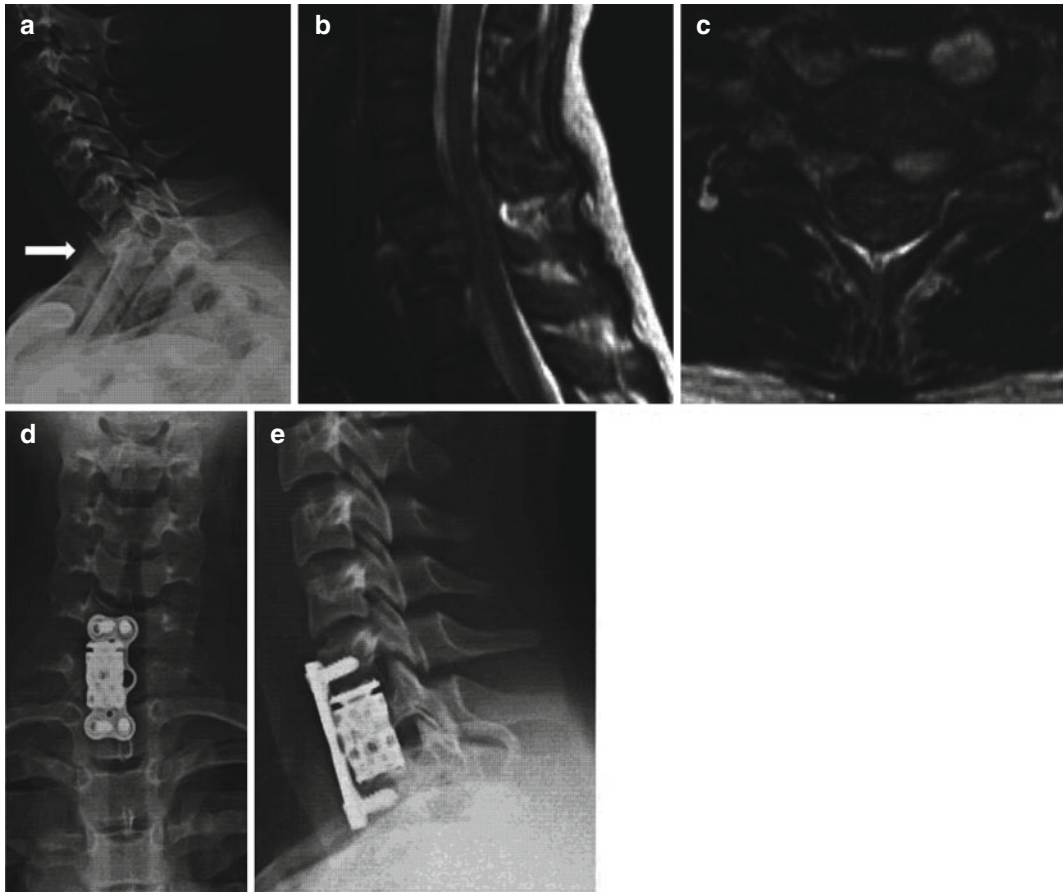

Fig. 27.15 A 10 year old child presented with C7 tuberculosis and quadriparesis. (**a**) Lateral radiograph shows collapse of C7 vertebra with widened prevertebral shadow (*white arrow*). (**b**, **c**) Sagittal and axial MR images show vertebral destruction, perivertebral abscess formation causing cord compression. (**d**, **e**) The patient was treated by anterior corpectomy, decompression of abscess and stabilisation with cage and plate. Along with anti-tubercular chemotherapy, titanium cage and plate can be safely used in active spinal tuberculosis

main advantages include the familiarity of the approach, less morbidity as opening of the body cavities are avoided, excellent exposure for circumferential spinal cord decompression, instrumentation which can be easily extended for multiple levels, better control of deformity correction, and safe performance of simultaneous anterior reconstruction. In patients with early disease with less deformity, a posterior transpedicular decompression with stabilization alone provides immediate pain relief, prevents deformity and any neurological sequelae. In patients with advanced disease, the transpedicular/extrapedicular route can also be used to place bone grafts or an interbody cage to achieve deformity correction and anterior vertebral reconstruction (Fig. 27.17a–f).

27.2.7 Kyphosis in Healed Tuberculosis

Deformities with more than 60° kyphosis are usually the result of childhood spinal tuberculosis and often require correction. Severe kyphosis is a major cause for cosmetic and psychological disturbance in a growing child. It also causes pain due to costo-pelvic impingement, respiratory impairment, and late-onset paraplegia. Surgical correction of established severe kyphosis in

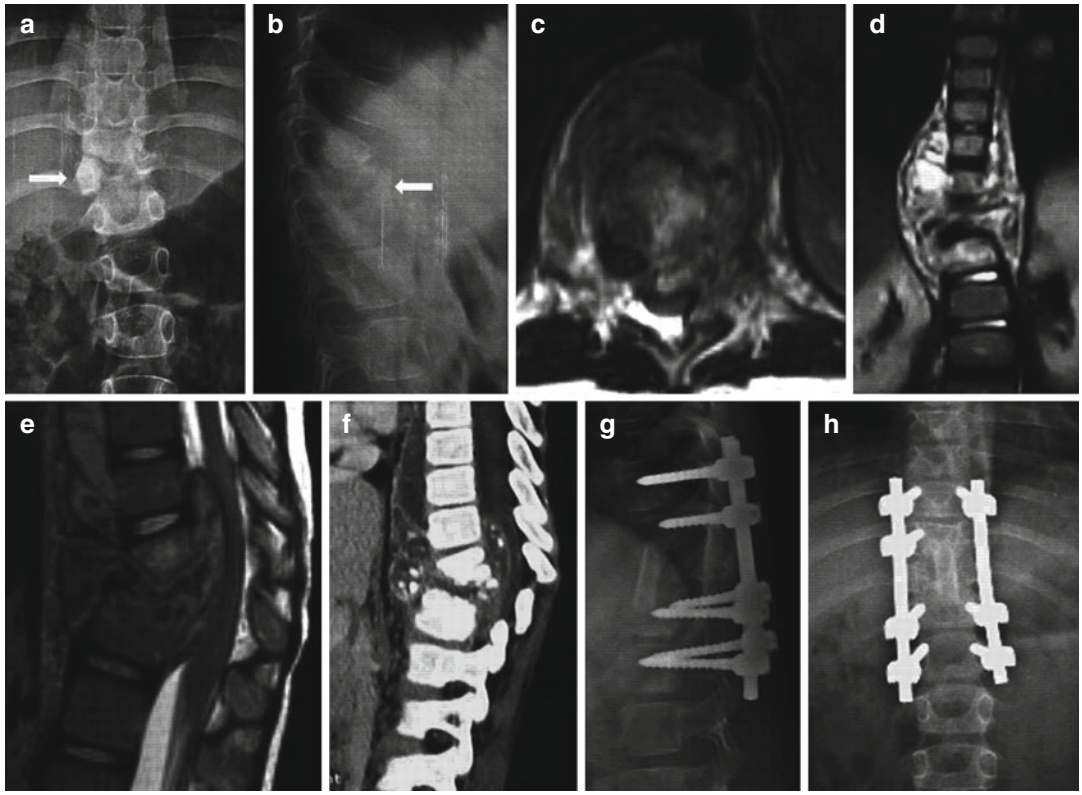

Fig. 27.16 (**a**, **b**) This 9 year old child presented with T12-L1 tuberculosis and paraplegia seen in the radiographs as vertebral collapse and local kyphosis (*white arrows*). The MR images (**c**-**e**) shows extensive abscess formation in the perivertebral space, epidural abscess, cord compression and multiple vertebral destruction. The sagittal CT shows the extent of bony destruction (**f**). (**g**, **h**) The patient has been treated by modified Hongkong surgery through anterior debridement and reconstruction with auto-fibular graft. Supplemental posterior stabilisation has been performed to prevent graft failure

children is challenging in many ways. The insertion of pedicle screws is fraught with danger as the vertebral anatomy is grossly altered. The pedicles are often small in size and osteoporotic providing additional challenges in fixation. During the healing process, there can be tethering and adhesions of the dura over the periapical segments with possibilities for dural tear or cord damage during decompression. The procedure obviously should be performed only by the experienced surgeons.

Different surgical techniques are available for the correction of severe kyphotic deformities. Anterior decompression in the presence of severe kyphosis is not only difficult but also dangerous due to fibrosis and adhesions near the apical fusion mass. In severe cases, two or three vertebral bodies are frequently destroyed causing a severe shortening of the anterior column but the posterior column is preserved. Correction by purely opening up the anterior column can cause severe stretching of the spinal cord with risks of neurological deficits. A combined anterior–posterior procedure to achieve anterior decompression and correction and stabilization by a posterior approach was common. However an all posterior approach is gaining more popularity because of higher ease and safety. Posterior closing osteotomies have been used effectively for kyphosis due to trauma, osteoporosis, and ankylosing spondylitis. However, in tuberculosis, where the anterior column destruction can be extensive, pure posterior closing wedge osteotomies can result in kinking of the cord with potential for neurological compromise. Hence an

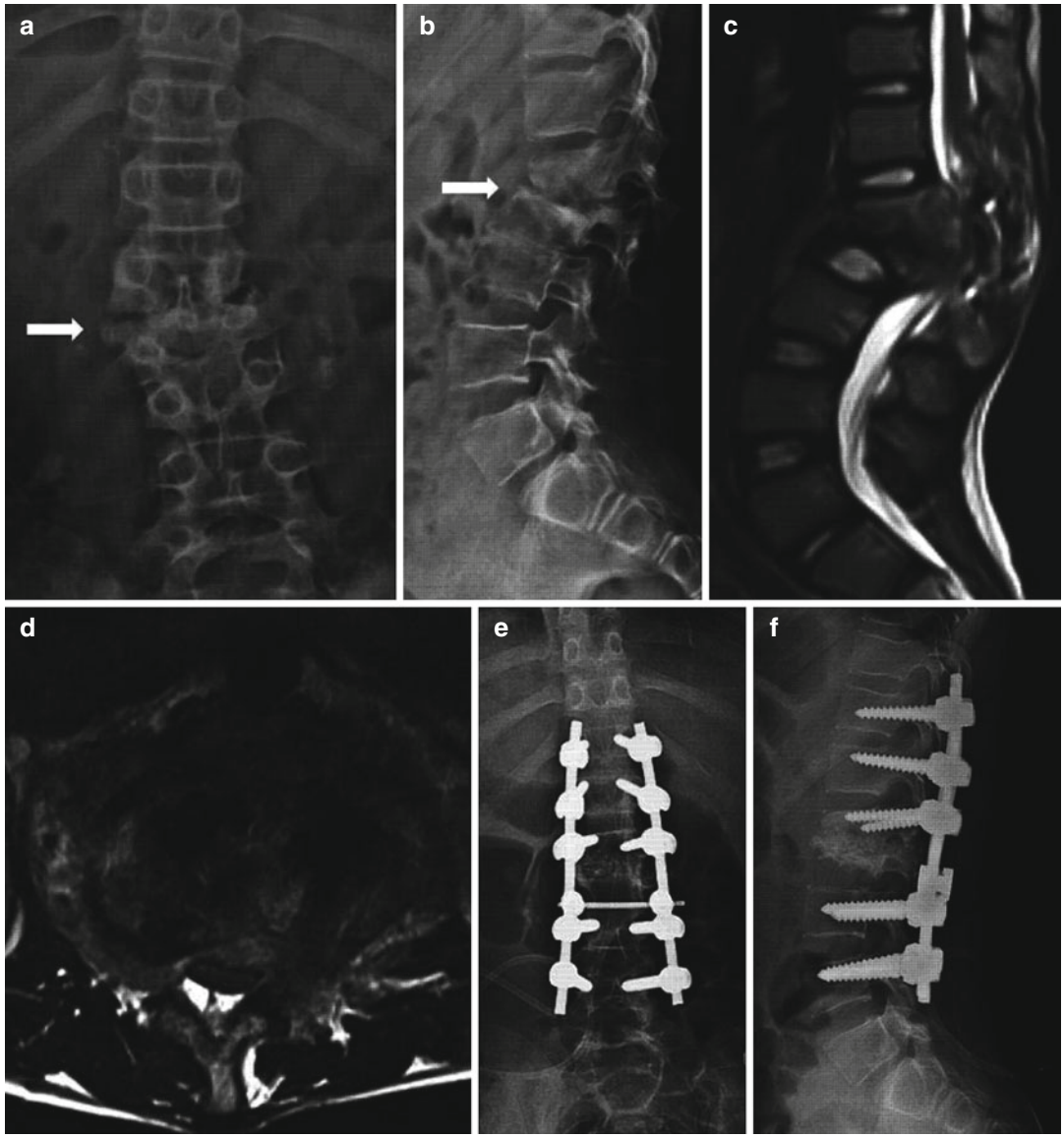

Fig. 27.17 This 13 year old boy had developed L1-2 tuberculosis and kyphosis with conus medullaris syndrome. (**a**, **b**) The anteroposterior and lateral radiographs show the vertebral damage and local kyphosis (white arrows). Sagittal and axial MR images show abscess formation and vertebral collapse causing cord compression (**c**, **d**). He has been treated by posterior decompression, transpedicular abscess drainage, posterior column shortening with Ponte's osteotomy and stabilisation from T11 to L3 (**e**, **f**)

"opening-closing osteotomy" that achieves correction by closing the posterior column but also lengthening the anterior column appropriately is preferred (Fig. 27.18a–d). Kawahara et al. originally described the closing-opening wedge osteotomy [29], and Rajasekaran et al. [30] has reported its usefulness in post-tuberculous kyphosis in which the mean preoperative kyphosis improved from 69.2° ± 25.1° to 32.4° ± 19.5° postoperatively. The percentage correction of kyphosis achieved was 56.8 ± 14.6 % (range, 32–83 %). The authors recommended this procedure as posterior only single-stage procedure, allowing for significant kyphosis correction with minimal complications.

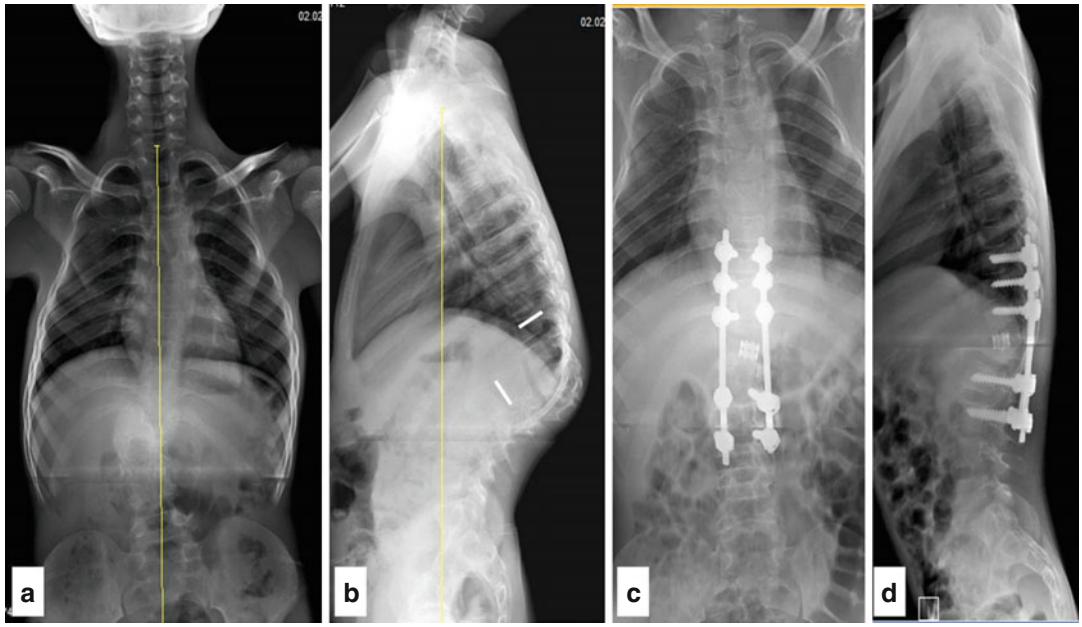

Fig. 27.18 Closing opening wedge osteotomy to correct a post tubercular kyphotic deformity. (**a**, **b**) Pre-operative AP, lateral radiograph of the patient shows a kyphotic deformity of 118 degrees at the thoraco-lumbar junction between T9 and L3 vertebrae (marked between the two white lines). The yellow lines represent the central sacral vertical line. (**c**, **d**) Post operative AP and lateral radiograph shows good correction of the deformity with pedicle screw instrumentation placed at least three levels proximal and distal to the apex. The 'opened' anterior wedge has been reconstructed with a titanium mesh cage

27.2.7.1 Technique (Fig. 27.19a–f)

The patient is placed prone over padded bolsters to avoid pressure over the abdomen and all bony points and superficial nerves are protected. The procedure is performed under spinal cord monitoring. The spine is exposed through a standard posterior midline approach at least three or four vertebrae above and below the region of wedge resection. The operative exposure is wide enough bilaterally up to the tips of the transverse processes to allow a costotransversectomy approach on both sides and pedicle screws inserted carefully. An extended laminectomy is performed to include at least one level above and below the osteotomy level. A contoured rod is temporarily fixed unilaterally to maintain spinal stability. The rib heads along with the transverse processes are then carefully excised, remaining strictly extrapleural. The parietal pleura is bluntly separated from the vertebrae. The exiting nerve roots at the level of kyphosis are dissected from the vertebral body and gently retracted to make room for the paravertebral dissection. If the surgery is at the thoracic level, one or two nerve roots at the apex can be doubly ligated and cut and held with the long ends of the ligature. Blunt dissection is performed anteriorly on both sides through the plane between the pleura and the vertebral body and the plane maintained by retraction with a spatula retractor.

The apical wedge resection is carefully achieved using an osteotome, curette, rongeur, and high-speed drill keeping a thin layer of the posterior vertebral cortex intact till the end to avoid troublesome bleeding from the epidural veins. The thin cortex also acts as a natural retractor protecting the dura. Once the dissection has been performed on one side, the temporary rod is shifted to the other side and similar steps performed. Once the apex of the wedge has been resected from either side, the posterior vertebral cortex is finally drilled out using a diamond burr. Correction is obtained by the use of appropriately contoured rods. Care is taken to avoid kinking or over-shortening of the cord that can cause neurological compromise.

27.2.8 Conclusion

After the availability of anti-tubercular drugs, the outcome of tuberculosis of the spine has dramatically improved. Infections in children can lead to

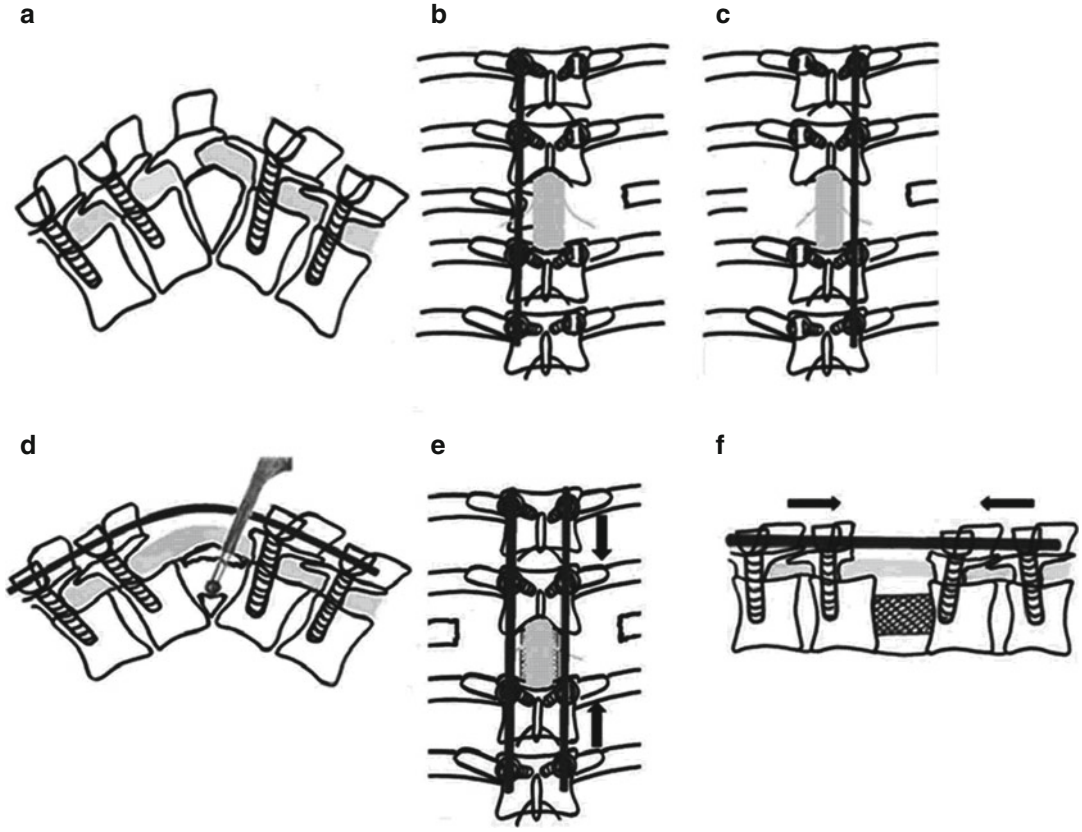

Fig. 27.19 (**a**) In the surgical procedure, a temporary stabilisation of the spinal column with a pedicle screw construct is first performed before decompression. (**b**) A stabilising rod is placed on one side and rib, transverse process and vertebral body resection performed contra laterally. (**c**) The stabilising rod is switched to the opposite side and decompression performed contralaterally. (**d**) The planned wedge resection of the vertebral body is performed using a high-speed burr and sharp curettes. It is important that a thin shell of posterior cortex of the fusion mass is maintained till the very end so that it forms a natural protection to the cord. Do not start burring at the apex of the deformity. If decompression is started from the posterior part of the fusion mass, there will be continuous bleeding from the epidural veins and the cord also is prone for injury as it descends down and comes into contact with the working instruments. (**e**) After thorough decompression, the posterior rim of the fusion mass is removed. (**f**) An appropriate sized cage or bone graft is used to open the anterior column so that kinking of the cord is avoided. Posterior compression of the pedicle screws is performed to achieve further correction

significant vertebral destruction with risks of deformity development and progression. Children need periodic follow-up till completion of growth. Uncomplicated tuberculosis of the spine is a medical disease and surgery is required only to prevent and treat complications of deformity progression or neurological deficit. Pan-vertebral lesions, risk or presence of severe deformity, a severe or progressively worsening neurological deficit, lack of improvement or deterioration despite adequate chemotherapy are indications for surgery.

27.2.9 Brucella Spondylitis

Brucella can affect the spine and its clinical and radiological presentation can mimic tubercular spondylitis. The lumbar region is the most commonly affected region but multiple site infections can also happen. The patients present with constitutional symptoms of chronic fever, weight loss, polyarthralgia, and significant back pain. A strong index of suspicion is required especially in endemic regions and diagnosis is based on radiographs, serology, and culture. Step like erosions

of the margin of the vertebral body, vertebral sclerosis, disc space collapse, and vertebral segment ankylosis by bridging osteophytes are typical radiographic features of spinal brucellosis. The diagnosis usually is confirmed by serum Brucella antibody titers of 1:80 or greater. Most patients can be treated by medical management for 3 months with tetracycline, rifampicin, or streptomycin. Surgical management is reserved for those with neurological compromise or persistent back pain.

27.3 Spinal Fungal Infections

Fungal infections of the spine are not common. *Coccidioides immitis*, *Blastomyces dermatitidis*, *Cryptococcus*, *Candida*, and *Aspergillus* are the common fungal pathogens that afflict the spine. Infection occurs in immunologically weak persons such as on cancer chemotherapy, HIV infection, chronic steroid abuse, and diabetes. The organisms spread through hematogenous means along intravenous lines, implants, and prosthetic devices, or during surgery, or spread from a primary pulmonary infection. Infection usually occurs in the vertebral bodies which results in vertebral compression fractures and kyphotic deformity of the spine. Early recognition of the disease requires a high index of suspicion, a detailed physical examination, and radiological evaluation, and confirmed by typical histological findings of fungal hyphae and spores. Treatment relies on the prompt institution of appropriate anti-fungal chemotherapy while surgery is indicated for resistance to medical management, spinal instability, and neurologic deficits.

27.3.1 Aspergillosis

Aspergillus fumigatus is a rare cause of spinal infection occurring in children with immunological compromise such as chronic granulomatous disease (CGD). The increased frequency of vertebral involvement may be due to a specific immune defect, as patients with CGD are susceptible only to certain organisms [31]. Aspergillosis is also the second most common invasive fungal infection in cancer patients accounting for 30 % of fungal infections [32]. Bone marrow transplant recipients can develop invasive aspergillosis following immunosuppressive therapy. Pulmonary involvement is the most frequent form of aspergillosis and vertebral with rib involvement occurs due to contiguous spread.

In children, isolated spinal involvement without deformity may be difficult to diagnose in the initial stages. Pain and tenderness over the spine and ribs with limitation of spinal movements occur initially followed by alterations in gait pattern. The infection may involve single or multiple vertebral bodies.

CT and MRI with contrast are useful in delineating bone and soft tissue pathology respectively. The chest radiograph may reveal single or multiple round opacities, or a fungal ball, especially in the immunocompromised host. The immunodiffusion (ID) test is effective and specific for the diagnosis of Aspergillosis in patients with an intact immune system. Counter immunoelectrophoresis (CIE) and radioimmunoassay (RIA) should be performed in immunocompromised patients with invasive Aspergillosis when the sera are negative for antibody. Isolation of the organism and histopathological demonstration of the hyphae in biopsies and cultures are consistent with a diagnosis of Aspergillosis.

Early surgical drainage of pus and spinal decompression are essential in controlling infection in children with CGD. Multiple debridements, and a partial or complete lobectomy, may be required for a contiguous pulmonary lesion. Spinal stabilization is advised in patients with extensive bone destruction and instability. Amphotericin B and 5-Flucytosine have been used as combination therapy because of their synergistic effect. The recommended dose of Amphotericin B is 0.1–0.2 mg/kg body weight administered intravenously by slow infusion. Five-Flucytosine is administered orally and the daily dosage varies between 100 and 200 mg/kg body weight. There is no consensus on the total dose or the duration of anti-fungal treatment. The duration is dependent on toxic side effects, the clinical response, and the radiological outcome.

Infections by other fungi are less common and management principles are similar to that of spinal aspergillosis.

27.3.2 Coccidioidomycosis

Coccidioidomycosis is caused by *Coccidioides immitis*. The organism enters the body through the respiratory tract and causes local infection. It can spread hematogenously resulting in systemic infection. The disseminated systemic form is progressive and potentially lethal. About 20 % with disseminated disease have osseous lesions. Diagnosis is made through serum complement fixation test and positive coccidioidin skin test. In the radiographs, most bone lesions are lytic in nature and involve the vertebrae and posterior elements of the spine. Treatment with amphotericin B or fluconazole is recommended. Surgical debridement and stabilization is indicated in patients with neurological deficit due to cord compression and spinal instability.

27.3.3 Blastomycosis

Blastomycosis is caused by *Blastomyces dermatitidis*. This fungus usually causes chronic respiratory infection but is capable of systemic dissemination. Systemic spread results in generalized symptoms of fever, night sweats, anorexia, and weight loss. Infection in the bone is common in disseminated blastomycosis, where the spine is also frequently involved. In spinal blastomycosis, the disc is usually involved early, and large paravertebral masses involving ribs is present. Treatment for blastomycosis is oral ketoconazole or itraconazole and in severe cases amphotericin B may be needed. Indications for surgery are similar to those for coccidioidomycosis.

27.3.4 Cryptococcosis

Cryptococcosis is caused by *Cryptococcus neoformans*. Similar to other fungal infections, it is a chronic fungal disease with a primary focus in the respiratory tract. Though it may affect all ages, it is most prevalent in the fifth and sixth decades of life. It is commonly seen in children afflicted with leukemia, Hodgkin's disease, or sarcoidosis. In 10 % of patients with disseminated infection, bone involvement happens. Apart from systemic symptoms of infection, local features include pain, swelling and restriction of spine motion. Similar to all the fungal diseases, cryptococcosis is treated medically with amphotericin B or fluconazole. Surgical indications are similar to those for other fungal infections.

27.3.5 Candidiasis

Candida is a common commensal organism but can produce infection in immunocompromised or patients on chronic antibiotic usage. Medical treatment is amphotericin B or fluconazole. Surgery is indicated in situations similar to other fungal infections.

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Part IV

Management of Spinal Deformity in the Growing Child: Non-surgical

Orthotic Management for Early Onset Scoliosis

28

Michael P. Glotzbecker, John B. Emans, and M. Timothy Hresko

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Key Points

- Orthotic treatment is a useful adjunct to cast treatment of early onset scoliosis.
- Orthotic treatment is most successful in idiopathic early onset scoliosis, particularly in single curves in the middle of the spine.
- Successful brace treatment of early onset scoliosis requires an effective brace, a committed multidisciplinary team, and an involved family.
- Bracing can cause irrevocable harm to the growing thorax if pressure is inappropriately applied or continued too long in spite of worsening thoracic deformity.

28.1 Introduction

This chapter will mainly discuss the contemporary role of orthotic treatment of idiopathic early onset scoliosis (EOS) [1, 2]. Orthotic treatment has limited efficacy in congenital scoliosis. Orthotic treatment of paralytic deformities, although useful, varies greatly by etiologic diagnosis, type of deformity, and goals of treatment.

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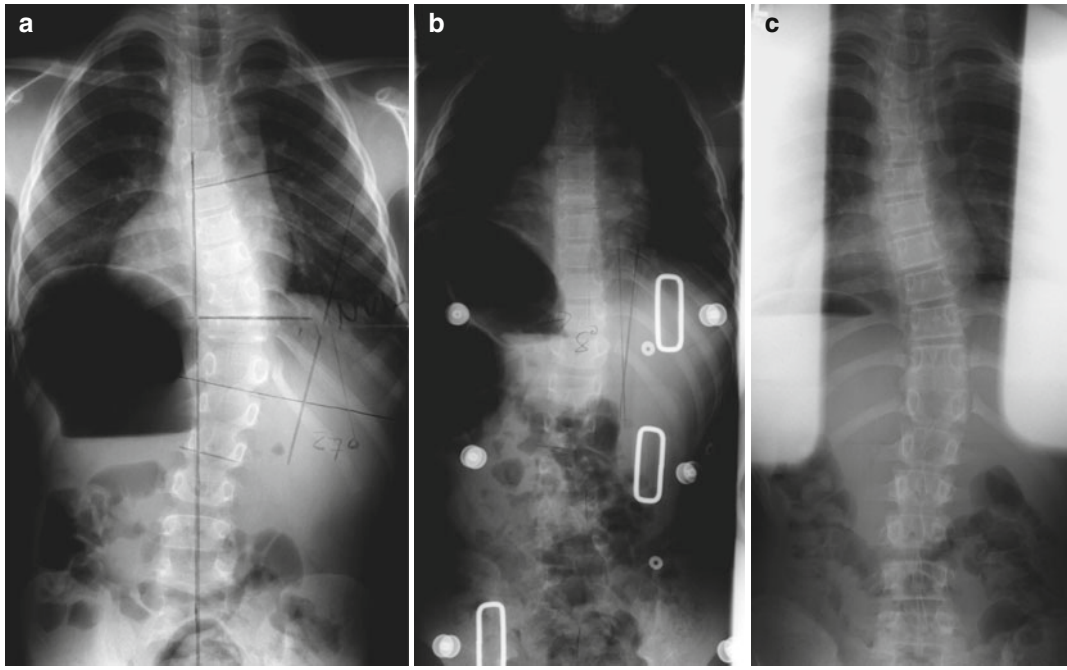


Fig. 28.1 Idiopathic EOS. Scoliosis was noted at the age of 6 years and progressed to 30° by the age of 7 years (**a**). Full-time brace treatment began at the age of 7 years (**b**) and continued through the age of 13 years, then part-time at the request of the patient. At follow-up at the age of 18 years after 1 year out of brace, there is a stable 25° curve (**c**)

These latter uses of bracing are beyond the scope of this chapter.

Bracing has always occupied a prominent position in the treatment of idiopathic EOS. Over the last two decades, orthotic treatment has come to be questioned as an effective treatment for adolescent idiopathic scoliosis for the lack of prospective, randomized evidence supporting the efficacy of orthotic treatment [3–6] in spite of a preponderance of nonrandomized series suggesting efficacy [7]. However, a recent prospective study has demonstrated efficacy for bracing and may therefore serve as better evidence to support its use [8].

Incontrovertible evidence for the efficacy of bracing in idiopathic EOS is lacking. The recent prospective study included patients greater than age 10 with adolescent scoliosis, and therefore its application to the EOS population is not known. Retrospective series, experience, and expert opinion strongly suggest that bracing can be effective in EOS, particularly in older children. Brace treatment of idiopathic EOS is approached with enthusiasm in continental Europe, yet greeted with skepticism in much of the United States.

With increasing evidence that there may be a law of diminishing returns when using "growth friendly implants," [9] as well having personal experience where patients have spontaneous fusion after an initial period of successful lengthening, nonoperative measures which at a minimum delay surgical treatment have appeal. We approach the subject with the impression that nonoperative treatments (casting or bracing) are the preferable treatment and can be effective in moderate idiopathic early onset deformity (Figs. 28.1 and 28.2).

Success or failure in bracing depends partly upon the goals chosen for treatment. Establishing realistic, specific, and transparent goals early in orthotic treatment of early onset deformity facilitates rational expectations by the practitioner and family. Is the goal complete correction, prevention of worsened deformity, or slowing of progressive deformity, acknowledging that surgery will eventually be needed? Complete, lasting correction with repetitive casting is a reasonable goal in early, selected idiopathic scoliosis as demonstrated by Mehta [10], and is anecdotally occasionally also achieved in moderate EOS in the older age range treated by bracing alone (see

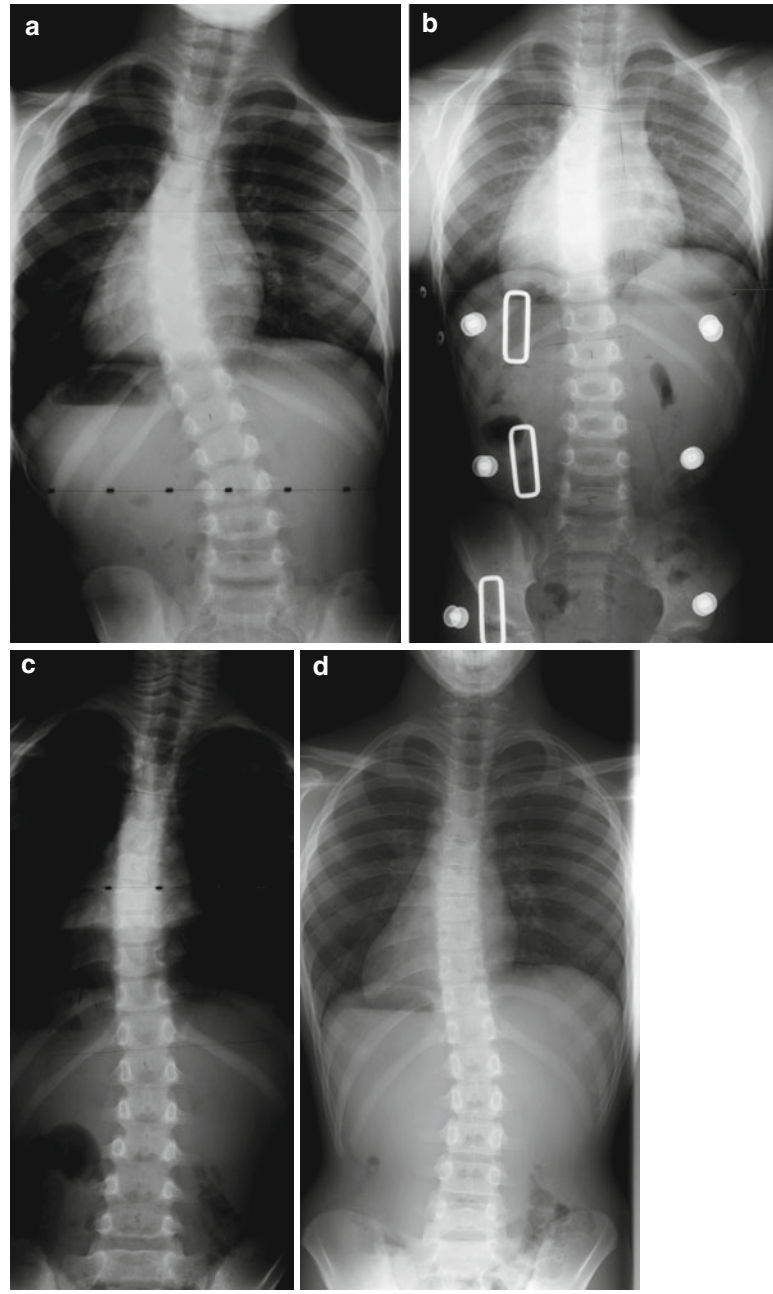


Fig. 28.2 Idiopathic early onset scoliosis. Scoliosis noted at the age of 3 ½ years (**a**) and full-time bracing initiated with good in-brace correction (**b**). At the age of 6 years, correction was maintained (**c**). Part-time bracing and correction were maintained through adolescence (**d**)

Fig. 28.2). Complete correction is rarely achieved in progressive idiopathic scoliosis in the younger age range by orthotic treatment alone and repetitive casting may be a better choice. Complete correction as a goal can help motivate families and patients assuming there is some chance of achieving that goal. Bracing is usually used in conjunction with casting even when complete correction is obtained, and the brace is used to maintain this correction after the cast is discontinued. In more severe EOS where complete correction is unlikely, nonoperative treatment is sometimes viewed as a temporizing measure, stabilizing deformity for years, allowing for more growth before initiating surgical treatment, and allowing the child to remain free of the need for repetitive surgical intervention with growing rods [11] (Fig. 28.3). Nonoperative treatment to allow for more growth with eventual surgery anticipated can be successful in achieving

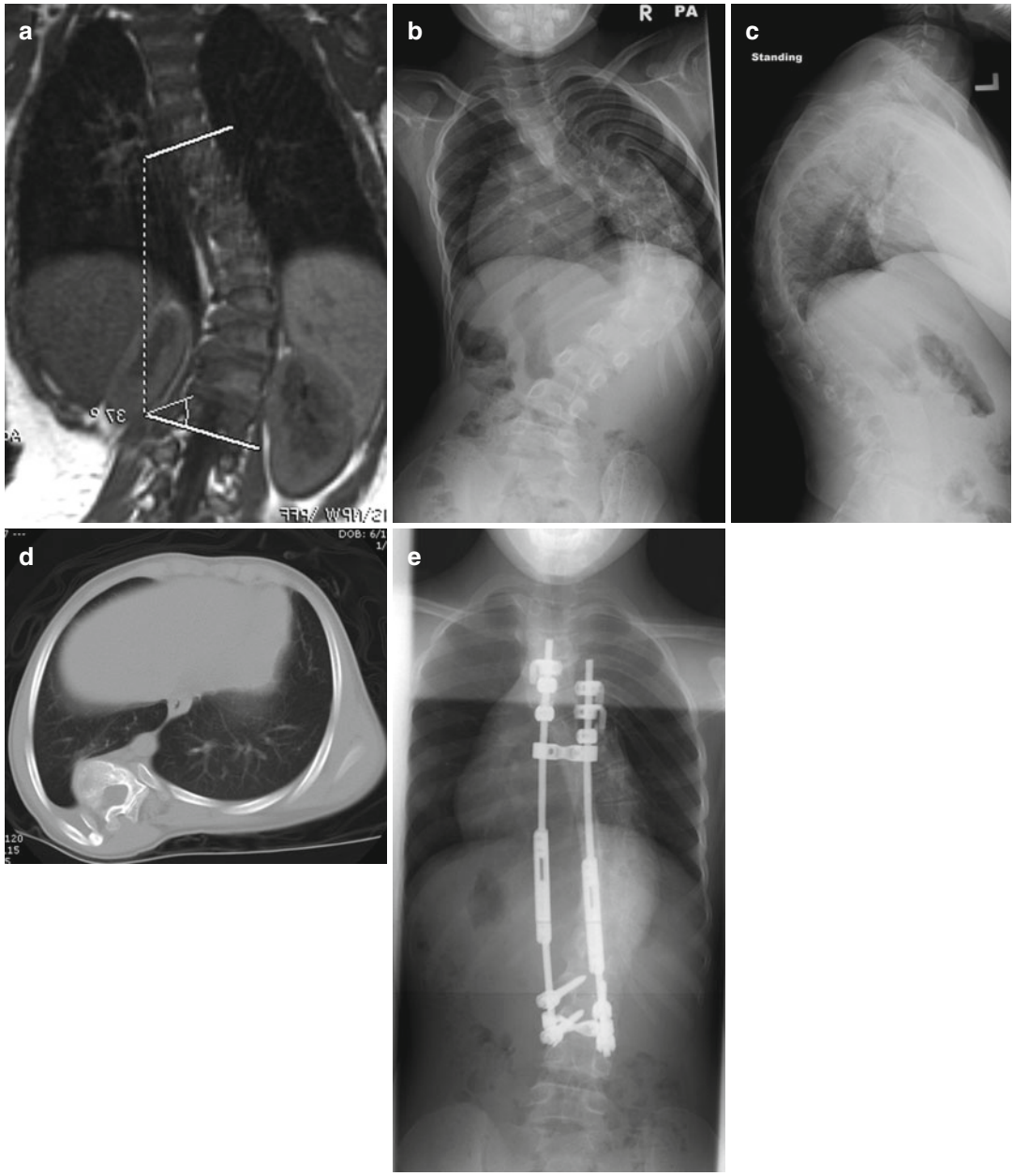


Fig. 28.3 Idiopathic early onset scoliosis. Treatment began at 18 months with full-time brace treatment (**a**). Referred for growing rods at the age of 7 years when rib prominence and thoracic deformity had worsened (**b**–**d**). Spine and thoracic deformity have been fairly well-controlled by dual growing rods from the age of 7 to 11 years (**e**). Growing rods had been suggested at the age of 2 years, but declined by the family. Earlier surgical intervention would have resulted in an easier initial surgical procedure, but the child was spared 5 years of surgical interventions and 10 surgical lengthening procedures by brace treatment from the age of 18 months until 7 years

the dual goals of a longer spine and fewer operations, but also may lead to inappropriate delay and worsened, irrevocable, thoracic deformity (Figs. 28.4 and 28.5). The availability of modern growth friendly surgical treatments for spinal deformity such as expandable spinal rods or

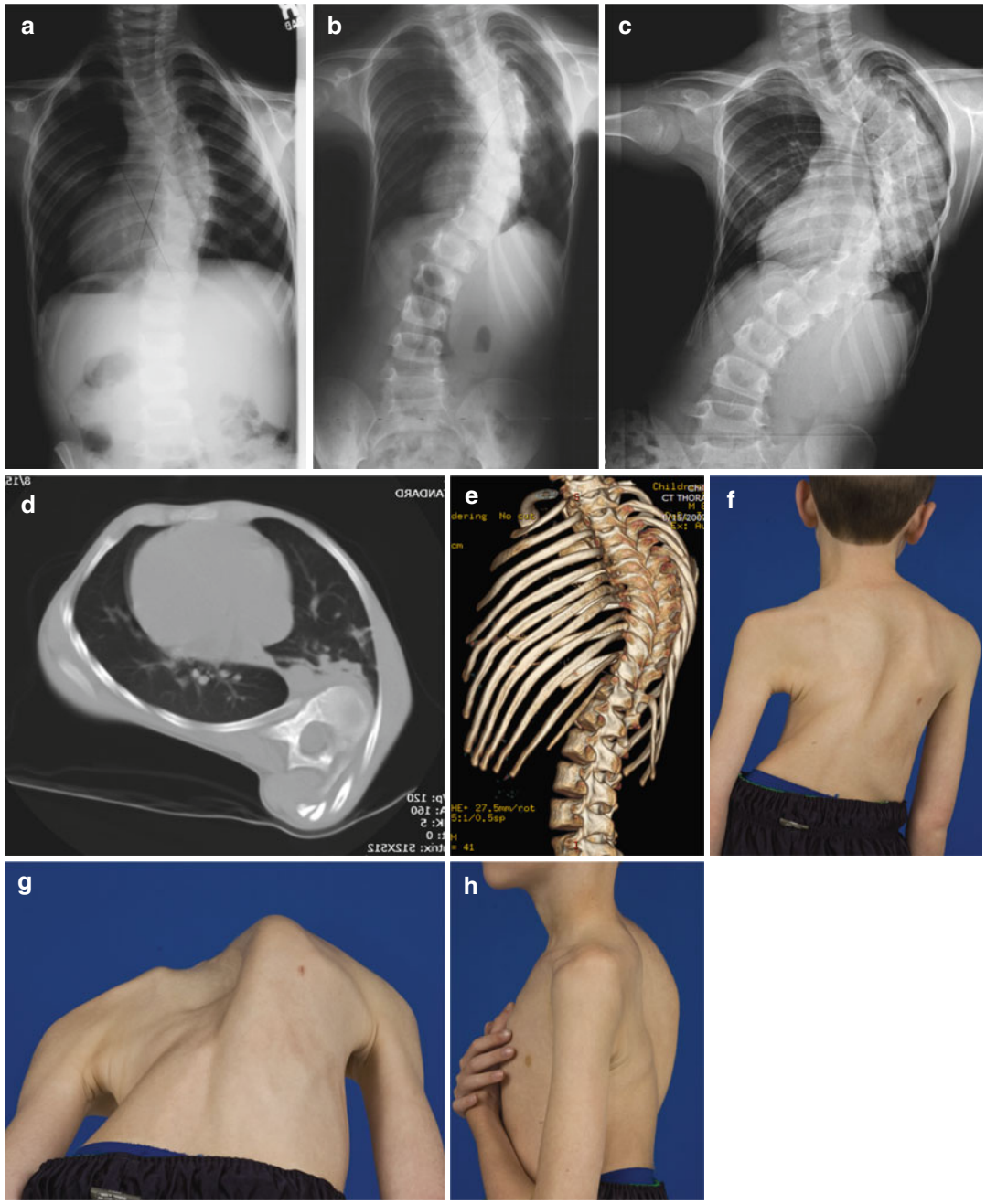


Fig. 28.4 Idiopathic early onset scoliosis treated with repetitive casting and bracing beginning at the age of 2 years. Initial thoracic deformity at the age of 3 years (**a**) was modest, worse at the age of 5 years (**b**), and severe at the age of 8 years (**c**), with pulmonary function tests approximately 50 % of predicted and early restrictive lung disease. The convex thorax is collapsed, with the ribs assuming a vertical orientation or "collapsing parasol deformity" as described by Campbell. Thoracic deformity is demonstrated on CT (**d**, **e**). An area of the posterior convex thorax normally occupied by lung is occluded. Thoracic deformity is clinically apparent (**f**–**h**). Spinal deformity can be controlled surgically at this stage, but the chest deformity will not be completely improved by surgical means. Earlier intervention with a growth-oriented technique such as dual growing rods or VEPTR might have controlled both spine and chest deformity and led to a better result

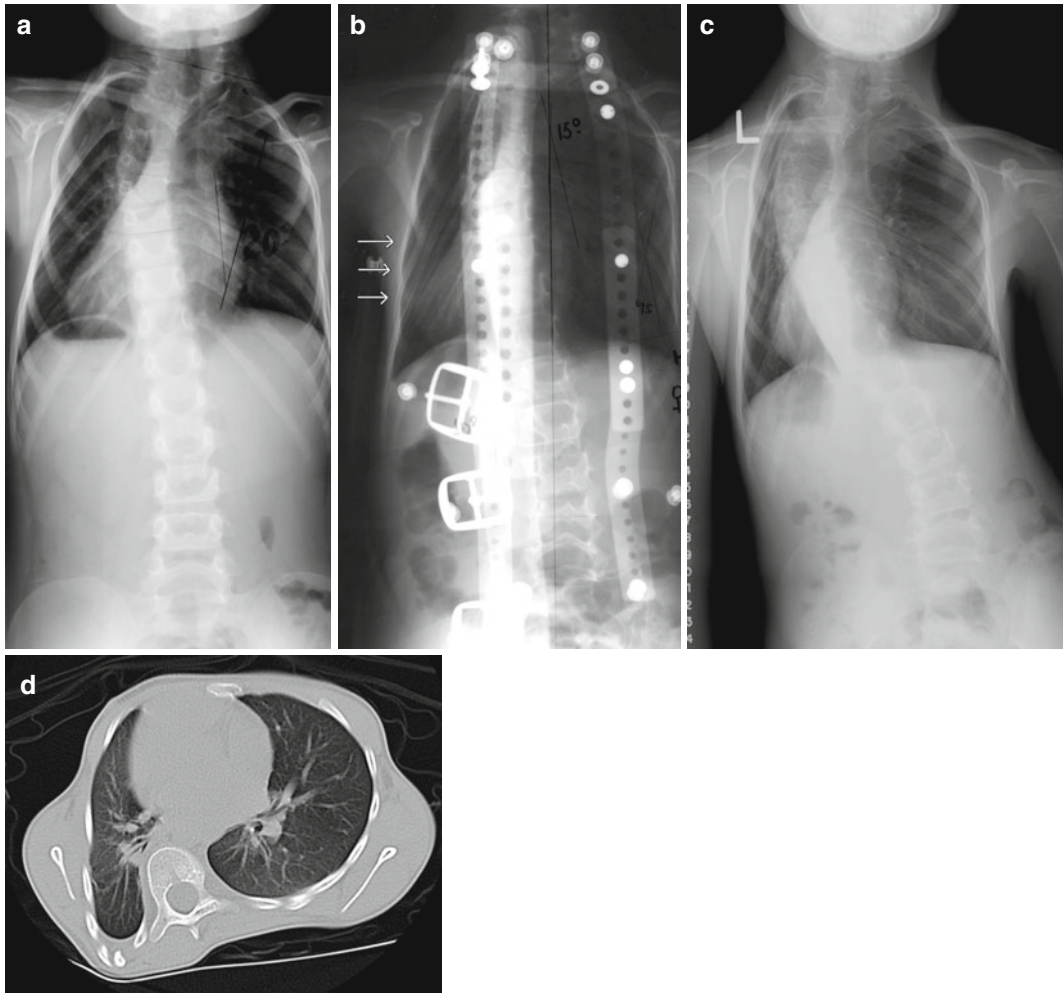


Fig. 28.5 Congenital early onset scoliosis in the upper thoracic spine was treated with circumferential *in situ* fusion at the age of 2 years (**a**). The normally segmented curve below was then treated with a full-time Milwaukee brace with a pad pressure applied (*arrows*) directly laterally over the convex chest wall (**b**). At the age of 9 years, the child was referred for surgical treatment because of curve progression (**c**) while still using the brace. Chest deformity with a severe collapse of the convex chest wall (**d**), however, has been evolving for years and is now irrevocable. Earlier growing rods would have been a better choice than persistent brace management

VEPTR should lower the threshold for discontinuance of bracing and initiation of surgery to a point *before* spine or chest deformity become too severe. Unfortunately quantifying the appropriate time for discontinuation of bracing is not defined, but is often determined by monitoring change of chest shape and spine deformity on serial radiographs and physical exam.

Effective brace treatment of early onset scoliosis demands appropriate indications, practical expectations, an effective brace, and committed care-givers. Commitment to bracing at the level of the physician (and the rest of the medical team), orthotist, and family is critical for success. Absence of dedication by any of the team will subvert the efforts of the others. Creating an effective scoliosis orthosis requires a skilled orthotist with experience in treating EOS patients. Although well-documented bracing systems of many types are available, it is difficult to be successful in early onset scoliosis without experience at some level. Not all techniques applicable

to adolescents are transferable to early onset scoliosis age group. Fortunately, the appropriateness of the specific orthosis and its potential effectiveness is easily assessed by radiographs taken in the brace. Radiographic confirmation of the effectiveness of bracing should complement clinical examination of the chest wall deformity. Patient compliance with requested brace usage is a major barrier to success in adolescents but much less problematic in younger children with early onset deformity if families are committed to brace wear. Often, the most likely member of the team to lack commitment is the physician, who may imply "try this brace for a while, it probably won't work; come back and see me when you need an operation."

28.2 Evidence for Efficacy of Bracing in Idiopathic Early Onset Scoliosis

Mehta's [10] experience with casting for idiopathic EOS is now well documented and shows remarkable, lasting correction of many patients in whom treatment was begun early and even some long-term improvement in many in whom referral was late. This experience clearly shows that the deformed growing spine can be guided through growth not just with stabilization of deformity but with actual long-term improvement in deformity, and that complete correction may be possible if the early infantile growth rate is harnessed to curve correction. The obvious advantage of casting includes full-time use without the need for adherence to bracing regimens. Mehta's [10] series included patients up to 48 months in age and her experience is relevant to bracing of early onset curves as it shows convincingly that with growth and appropriate application of external pressure, the deformed growing spine can be changed for the better. Mehta [12] has also advocated the use of serial plaster casts in older patients with idiopathic EOS, but this is less well documented. Experience with brace treatment alone for younger patients with idiopathic scoliosis is sparsely reported [12–17]. McMaster and Macnicole [18] documented Milwaukee brace treatment in 27 children with idiopathic EOS in young children, of whom only 5 did not require surgery during adolescence. However 70 % of the children in the study wore the brace a minimum of 5 years suggesting that bracing delayed the need for surgical intervention.

Experience reported with brace treatment of older patients with idiopathic EOS is encouraging. With notable exceptions [12, 19–24], this experience is blended into reports on success or failure with adolescent idiopathic scoliosis. Robinson and McMaster [24], in analyzing curve patterns in idiopathic EOS, reported 88 of 109 patients who were treated with a brace. Arthrodesis was needed in 67 of 84 thoracic curves but in only 3 of 20 thoracolumbar or lumbar curves. However, the mean Cobb angles were higher in the thoracic curve patterns at the initiation of treatment [25, 26] compared to the thoracolumbar and lumbar curve patterns [22, 27]. Curve correction in brace was best below the age of 6 years and early in bracing. Noonan et al. [28] discouraging report of bracing in idiopathic scoliosis included patients as young as the age of 8 years, but EOS patients are not distinguished from the rest, although the authors noted a higher failure rate in patients under the age of 12 years. The experience with the Boston Brace system in 295 patients [29] at our institution included 34 patients in the age of 10 years or less. When compared with adolescents in the study, patients less than 10 years old at initiation of bracing had a higher rate of surgery, but also a higher mean correction at the end of bracing in those who did not need surgery. There were very few patients whose curves remained the same by the end of growth, probably reflective of the large amount of growth and opportunity for change in either direction during treatment. We felt that bracing as a whole for this group was successful. Mean curve correction at the end of bracing was 25 % for patients who were successfully treated with bracing. Of all patients between the age of 4 and 10 years at initiation of brace treatment, only 5 of 34 went on to surgery. Of those less than 10 years of age starting bracing with curves between 30 and 49°, only 2 of 11 went on to surgery. Tolo and Gillespie [30] reported on 44 patients braced for EOS,

among whom, 16 went on to surgery and felt that part-time brace use might be effective. Jarvis et al. [20] reported on 23 patients and also felt part-time bracing was effective. Kahanovitz et al. [27] reported on treatment of 15 EOS patients with part-time bracing and noted success for patients who had curvatures less than 35° at the onset of part-time bracing and whose rib vertebrate angle difference remained less than 20°.

All these studies suffer from being retrospective selective reviews without either cohort controls or prospective controls. With the exception of Noonan, however, all observed encouraging outcomes for patients treated with braces. The Scoliosis Research Society prospective study of bracing in idiopathic scoliosis by Nachemson and Peterson [31] demonstrated efficacy in bracing for adolescent idiopathic scoliosis, but does not include patients below the age of 10 years. In a recent study by Weinstein et al., the rate of treatment success with bracing was 72 % after bracing, as compared with 48 % after observation in a prospective study of patients aged 10–15. This study also demonstrated a dose response with regard to hours of wear, but again whether these findings translate to the early onset population is not known [8]. In addition, the rate of successful treatment by observation or bracing was worse in young patients who were Risser 0. In Risser 0 patients, the calculated probability of failure ranged from 32 to 91 % depending on the initial Cobb angle compared to 7–53 % depending on the initial Cobb in Risser 1+ patients. Bracing reduced these probabilities to 13 % and 76 % and 2 % to 28 % with bracing, respectively [32].

Long-term studies of outcome after bracing for adolescent idiopathic scoliosis [33, 34] indicate a favorable long-term result with regard to pain and function. All show a cohort of patients functioning well with no report of major psychological impairment and no impairment of bone density when bracing begins in adolescence. The less optimistic functional outcome for early onset scoliosis as a group is well documented by Goldberg et al.[35, 36] and Pehrsson et al. [37]. Masso et al. [23] reported no difference in child health questionnaire results in braced patients with idiopathic EOS when compared with those only observed. No other reports of long-term functional outcome after bracing in early onset deformity are found. We may probably safely conclude that older EOS patients closer to age 10 with moderate curves at the end of growth have long-term outcomes similar to their adolescent counterparts, while those with more severe curves requiring early surgery are more likely to demonstrate respiratory insufficiency and functional deficits associated with a short, fused spine.

28.3 Decision-Making in Orthotic Treatment of Idiopathic Early Onset Scoliosis

28.3.1 Goal-oriented

Goal-oriented thinking is helpful in assessing patients with early onset spinal deformity. Broadly stated goals for early onset deformity patients include achieving maximum spine growth and length, maximum spine flexibility, optimal respiratory function and lung growth, and a minimum of hospitalizations and procedures. Some goals are frequently at odds with others, but utilizing these goals to assess patient status will often help make the choice between observation, bracing, and surgery more rational. Families should understand these goals, and the care of early onset deformity, as a logical progression toward a final, functionally acceptable spine at the end of growth and treatment.

28.3.2 Indications for Bracing

Indications for bracing are different dependent on age in EOS. In the youngest children, casting should be considered as the preferred treatment and the decision to observe or treat based on the criteria advocated by Mehta. The first 2 years of growth can be viewed as an opportunity to maximally correct the spine deformity. Indications for bracing in this group is probably restricted to bracing after serial cast treatment or infants who do not tolerate casting, or those with gastroesophageal reflux, severe eczema, severe sleep

apnea, or where casting is simply not available. Full-time brace treatment of progressive or persistent scoliosis may then be appropriate. If bracing is undertaken in infancy, great care should be taken to not apply pressure over the thorax, except as a part of a derotation maneuver and to allow adequate room for expansion of the thorax (Fig. 28.6b, c). Braces should follow the same principles outlined by Mehta for casting. An improperly designed brace applied full time can quickly create a new thoracic deformity in the infant in excess of or equivalent to that created by an improperly applied cast.

Indications for orthotic treatment of older patients with idiopathic EOS are suggested by published results of brace treatment, likelihood of curve progression, and biomechanical curve simulations. This center has utilized a Cobb angle in excess of 20° as a lower threshold for orthotic treatment in idiopathic EOS curves. We agree with Winter [38] and urge that curves over 20° should be considered for brace treatment in EOS, assuming that the curve has been persistent or progressive and is in a region of the spine accessible to bracing. Biomechanical models [25, 39, 40] of scoliosis suggest that at approximately 25° of curvature, the load required to deform the spine diminishes significantly and that conversely, if the curve can be diminished to well under 25°, then vertebrae may be loaded much more symmetrically. Stokes et al. demonstrated asymmetric growth of rat tail vertebrae in response to asymmetric loading [26]. The rationale for early bracing of moderate curves (those in excess of 20°) is the assumption that by placing the growing spine under straighter mechanical load, there is some chance for spine remodeling toward symmetry, and progression is less likely during the preadolescent period of rapid growth. Sanders et al. [41] and many others have shown the early adolescent growth phase to be the period of greatest risk for progression of scoliosis, while Lonstein and Carlson [42] and Charles et al. [43] quantified the relationship between growth phase, curve magnitude, and the risk of progression. The goal of early bracing of moderate idiopathic EOS should be to enter the rapid preadolescent growth phase, when risk of progression is highest, with as little deformity as possible.

Early onset scoliosis associated with syringomyelia, Chiari malformation, or a tethered spinal cord should also be considered for brace treatment. Although surgical treatment of the Chiari malformation, syringomyelia, or tethered cord often results in improvement in the associated spinal deformity, the spinal deformity may continue to worsen if there is established deformity or persistent neuraxis abnormality [44]. Surgeons and families may falsely assume that by alleviating the presumed etiologic cause of the deformity, the deformity itself will probably resolve spontaneously as it frequently does if the deformity is mild. Many patients with established kyphotic deformity or scoliosis in excess of approximately 30° may develop progressive deformity during the rapid growth of preadolescence, even though the neuraxis abnormality has been treated. Although a trial of observation following decompression of the Chiari malformation or syringomyelia or tethered cord is appropriate, if the deformity persists as excessive kyphosis or greater than 20° of scoliosis, treatment as in idiopathic EOS should be instituted.

28.3.3 Contraindications

Contraindications to bracing include certain curve locations, very large curves, associated thoracic lordosis, advanced chest deformity, and some medical and psychological conditions. Multiple reports of brace treatment of adolescent idiopathic scoliosis note the poor results of bracing in upper thoracic curves, triple curves, and curves at the lumbo-sacral junction. Although the Milwaukee brace is felt to be the most appropriate brace for curves with apices above T6, reported results [45] are not encouraging, leading most practitioners to observe rather than treat the upper thoracic curves. Jarvis et al. [20], Lenke and Dobbs [21], and McMaster and Macnicole [18], in their series of idiopathic EOS, did not specifically note curves with predominantly high thoracic apices, suggesting that this is an uncommon curve pattern in this age group.

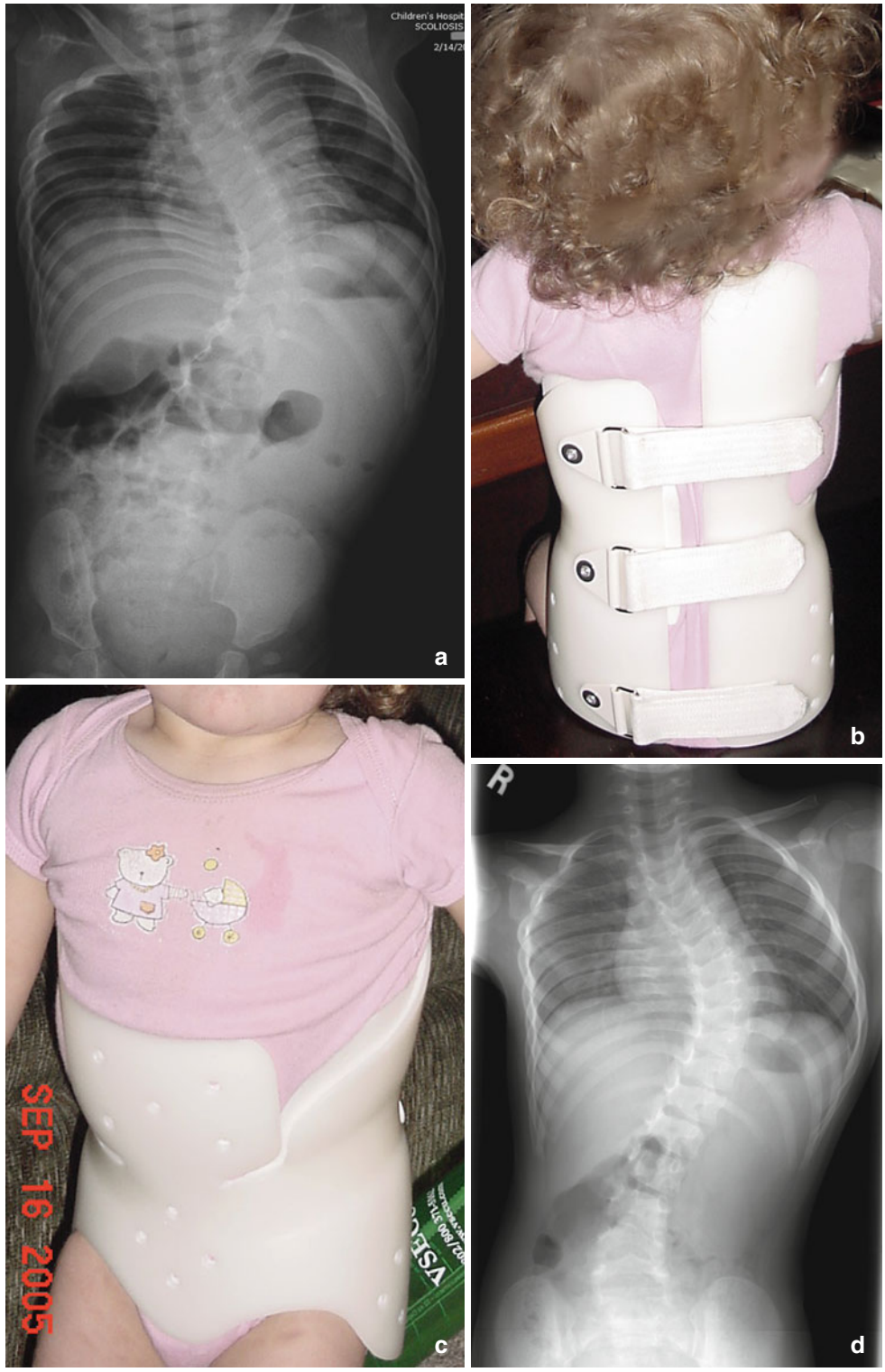


Fig. 28.6 EOS with tethered spinal cord. Scoliosis noted at the age of 6 months. No treatment was initiated and no MRI was done until the age of 2 years. In spite of detethering performed at the age of 2 years, scoliosis progressed at the age of 2 1/2 years (**a**) and bracing was initiated. Full-time custom-molded Boston brace is well-tolerated, asymmetric, and has large areas of relief opposite any area of pressure (**b**, **c**). At the age of 6 years, bracing continues with some progression of curve and mild thoracic deformity (**d**). Dual growing rods was planned, if worsening continued

Jarvis et al. [20] noted more success in EOS with single thoracic and thoracolumbar curves than with double major curves, mirroring our experience with adolescent idiopathic scoliosis and idiopathic EOS. Most curves with high thoracic apices are accompanied by secondary, less structural lower curves of lesser magnitude, which may be successfully treated by bracing. Surgical correction of the high thoracic curve followed by brace treatment of the lower curve(s) is also an option when the upper thoracic curve is rapidly progressive.

Thoracic hypokyphosis or thoracic lordosis is a nearly universal accompaniment of idiopathic thoracic curves, and is often cited as a contraindication to brace treatment, yet Mannherz et al. [22] found thoracic hypokyphosis in only 20 % of their series of idiopathic EOS curves. Although frequently mentioned [22, 23, 29, 45, 46], guidelines for the treatment of thoracic lordosis and hypokyphosis are not clear. Our practice has been to treat associated thoracic hypokyphosis with a brace modified to include posterior cephalad extensions of the brace intended to encourage thoracic kyphosis. For true thoracic lordosis (less than 0° of thoracic kyphosis), bracing may be counterproductive and produce more thoracic lordosis. Thoracic lordosis, in association with thoracic scoliosis, may however be the ideal indication for surgical guided-growth procedures such as anterior vertebral stapling, or tethering [47, 48].

Large curves (in excess of 60–90°) are rarely permanently stabilized by repetitive casting or bracing in EOS. Although bracing may be used for large curves to allow more growth before a planned surgical intervention, many large curves are best dealt with by surgical intervention such as dual growing rods. Moderately large curves in the adolescent may be successfully treated with bracing as demonstrated by Wiley et al. [49] and Katz and Durrani [50] and also in some EOS. In our Boston Brace series [29], none of the EOS patients, aged 4–10 years, with curves 40–49° at initiation of bracing needed surgery after bracing and follow-up. Our present practice is to attempt bracing in larger EOS curves between 40° and 60°, provided that there is acceptable chest deformity, but will switch to dual growing rods if the chest deformity worsens significantly.

Significant chest deformity [35, 51] often accompanies more severe EOS curves and may be a contraindication to bracing. Continued brace treatment may worsen the chest deformity while seemingly stabilizing the spine deformity. The more advanced the chest deformity is, the more likely that the patient will be left at the end of surgical treatment with a functionally significant thoracic deformity and increased risk of respiratory insufficiency as an adult [35, 37]. In most severe idiopathic EOS, final instrumentation and fusion is generally successful in achieving a balanced, stable, minimally deformed spine at the end of treatment. However, surgical treatment is rarely successful in restoring normal chest shape and normal chest compliance when there has already been a severe chest deformity. Therefore, chest deformity should not be allowed to worsen beyond a point at which it is irrevocable. Surgical treatment such as dual growing rods should be instituted earlier.

Some associated medical conditions are also contraindications to bracing. Severe gastroesophageal reflux [52] may be exacerbated by abdominal pressure from a brace and may be a contraindication. Failure to thrive or anorexia nervosa may be aggravated by a constrictive brace or any orthosis. Patients with severe asthma may not tolerate bracing during periods of exacerbation. Patients with difficulties with temperature regulation will also be affected by bracing, and those in very warm climates may not tolerate full-time orthotic use. Those with severe eczema or other skin conditions often will not tolerate the continued contact between skin and the brace. Adverse psychological reactions to bracing are commonplace in adolescents, less common in EOS patients, but may still be a contraindication to bracing. Family ambivalence toward treatment or failure to be supportive of the braced child is a relative contraindication to brace treatment. Similarly, if the physician and team do not really believe that bracing is beneficial, or have limited experience or insufficient skill to be successful with bracing, it is probably preferable to observe or operate rather than treat with a brace.

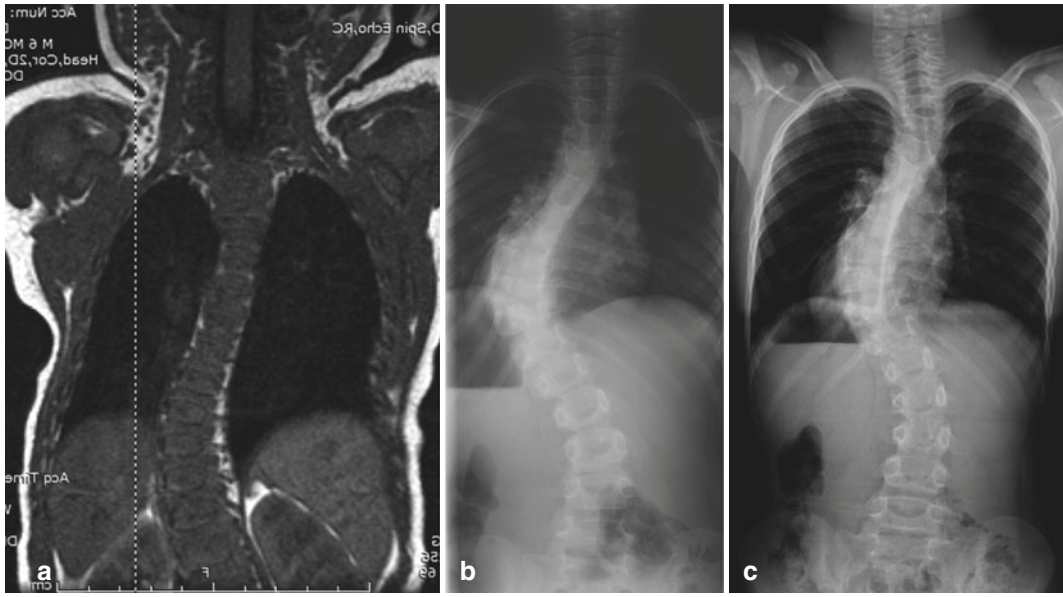


Fig. 28.7 Idiopathic early onset scoliosis. Bracing was begun at age 20 months with thoracic Cobb angle of 60° (MRI at 18 months of age (**a**)) and has been continued full-time in a TLSO (age of 6 years seen on (**b**)), with an emphasis on allowing chest expansion opposite the pressure pads. By the age of 11 years, the curve has increased minimally to 70° (**c**) with minimal thoracic deformity. If a rapid increase in either chest deformity or curvature starts in the preadolescent growth phase, definitive fusion or growing rods can be accomplished. Otherwise, fusion and instrumentation can wait until later in growth

28.3.4 When Should We Switch to Surgical Treatment from Bracing?

When should we switch to surgical treatment from bracing? A common and difficult issue is when to switch from brace treatment to surgical treatment for deformity in the growing spine. We suggest that the decision to switch to surgical treatment should be based more on progression and severity of the thoracic deformity than the magnitude of spinal deformity. A large thoracic curve stabilized by bracing, with minimal chest deformity may continue to be managed by bracing in anticipation of eventual definitive fusion (Fig. 28.7). At final operation, the spinal deformity can be stabilized, and the thoracic deformity and function will be acceptable. In contrast, if a moderate thoracic curve is associated with a severe or progressive chest deformity, brace treatment should be abandoned in favor of growing rods, or if old enough, definitive fusion (see Fig. 28.3). This attitude is based on the observation that surgical treatment is generally successful in correcting and stabilizing spine curvature, but poor at correcting severe chest deformity and restoring normal thoracic compliance and respiratory function. Brace treatment should be stopped and surgical treatment should begin before chest deformity becomes irrevocable or severe.

Bracing should also be abandoned in favor of surgical treatment if continued brace treatment will cause a more extensive spinal fusion at a later stage. Persisting with brace treatment of some progressive thoracic curves may cause increasingly larger lumbar curves and the need to eventually include the lumbar curve in the final fusion. Switching to growing rods or fusion for control of the thoracic curve may save the lumbar curve from eventual fusion.

28.4 Bracing Techniques

28.4.1 Brace Types

Brace types available for orthotic treatment of early onset scoliosis vary greatly. Available rigid braces include traditional CTLSO braces such as the Milwaukee brace, TLSO braces such as the Boston Brace, Wilmington brace, Cheneau or Rigo-Cheneau brace, and night time only, "over-correcting" braces such as the Charleston and Providence braces. Nonrigid braces such as the Kalabas, SpineCor, or dynamic movement orthosis systems are also available. Manuals and technical details for most North American bracing systems (Milwaukee, Boston, Charleston, Providence, and Charleston) are available online at the Scoliosis Research Society http://www.srs.org/professionals/bracing_manuals/. Most physicians will have experience with a limited number of bracing techniques, and it is probably preferable to use a familiar technique with which the team is skilled, than to attempt an unknown method for the first time. The authors' preference is to use a full-time Boston Brace for most patients with idiopathic EOS.

28.4.2 Principles of Correction

Principles of correction are remarkably similar among brace types and probably never completely achieved. Braces should be constructed based on principles of the particular bracing system, but applied and modified as needed to the individual patient. The orthotist and physician must communicate about the orthosis to be constructed and the orthotist must have access to and base the brace upon radiographs in both coronal and sagittal planes, as well as an examination of the patient. Common principles espoused by all systems include the recognition that idiopathic scoliosis is a three-dimensional deformity and that correction should be sought in all three planes by forces applied in all three dimensions. Brace construction should be planned in all dimensions. Coronal deformity is corrected by lateral pressure, rotational deformity by rotational pressure on both the front and back of the brace, and sagittal misalignment may be improved or should not be made worse. Every system, in one form or another, provides an area of relief, void, or window in the brace opposite the applied force, to both enhance the asymmetry of the force and provide an area into which the spine may shift as it moves toward a corrected position. Wherever possible, derotation forces are coupled, so that, for example, derotation of a typical right thoracic curve will include posterior to anterior pressure on the right posterior rib hump and anterior to posterior pressure on the left anterior rib prominence. No unnecessary constriction should occur, particularly of the chest. Continued application of force to the growing chest can create an irreversible thoracic deformity of greater significance than the underlying spinal deformity [51] (see Fig. 28.5).

28.4.3 Physical Therapy

Physical therapy is prescribed on a variable basis in North America, but in the opinion of the authors, specific, individualized physical therapy is an important adjunct to full-time bracing in the child, old enough to cooperate. The Milwaukee and Boston systems as well as the Rigo-Cheneau and other European systems place great emphasis on coordinated physical therapy. Purported benefits include reduction of associated lower extremity contractures, enhanced in-brace correction through active in-brace exercises, strengthening to counteract the inevitable weakening of trunk muscles by a full-time brace, and improvement of thoracic hypokyphosis or other sagittal malalignment. Contact with an informed and enthusiastic physical therapist on a regular basis further reinforces the team commitment to the individual patient. Physical therapy for nighttime only bracing may be much less important, but some patients with EOS as well as adolescents may exhibit pelvic obliquity associated

with infra-pelvic contractures, including the iliotibial band or tensor fascia femoris. Physical therapy alone has generally been considered in North America to be of little value in preventing progression or reducing scoliosis. However, there has been enthusiasm in Europe and increasing enthusiasm in North America for specific, individualized, intensive programs of physical therapy such as the Schroth technique [53] with some limited data to demonstrate its effectiveness in mild curves [54].

28.4.4 Assessment

Assessment of patient progress during bracing should include postero-anterior (PA) and lateral radiographs both in and out of the brace. However, in early onset scoliosis, radiographic assessment will likely continue for many years, with the potential for many radiographs during a potentially sensitive period of growth. Based on a large cohort of women treated for scoliosis, Hoffman et al. [55] and Morin Doody et al. [56] suggested that frequent exposure to low-level diagnostic radiation during childhood or adolescence might increase the risk of breast cancer. Reporting on an expanded cohort [57], recently, there appeared only to be a borderline-significant radiation dose response, not related to stage of development at first exposure, but increased significantly by a family history of breast cancer. Limiting the lifetime radiation exposure is nonetheless desirable in early onset patients. We generally require PA and lateral views before bracing, and obtain PA and lateral views in the brace once the brace is maximally adjusted, to assess brace construction, pad placement, and in-brace correction. The in-brace radiograph as well as the brace on the patient are assessed to see that maximum correction has been achieved and that the original plan, as agreed upon by the orthotist and prescribing physician, has been followed and is effective. Most follow-up radiographs are taken out of brace at intervals that are as widely spaced as feasible, usually when the data from the follow-up radiograph is needed for a treatment decision.

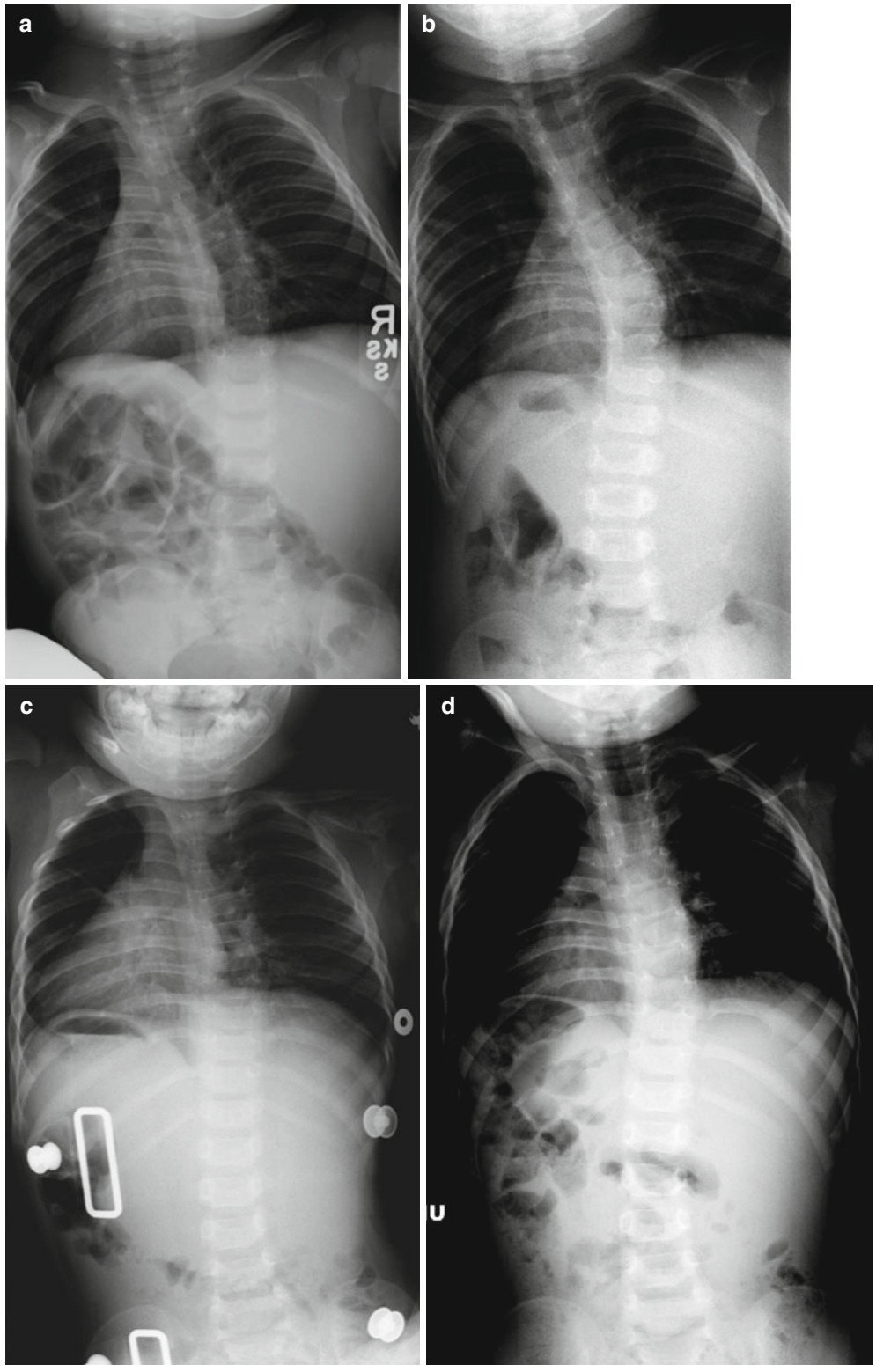
28.4.5 In-brace Correction

In-brace correction as seen on radiographs has been noted by multiple studies [10, 23, 24, 29, 33, 45, 49, 50, 58] to be a meaningful prognostic sign for eventual success or failure in bracing. In-brace correction is presumably related to both the inherent flexibility of the individual curve, brace construction, and strap tension [59]. The physician cannot influence the first factor, but the latter two can be improved by increased skill in brace construction and patient/family diligence. When in-brace correction is less than anticipated, a careful reassessment of the brace and plan for brace construction should be undertaken. We use 50 % correction in the brace as a goal, always expecting to see this in single thoracolumbar curves and most thoracic curves (see Figs. 28.1, 28.2, 28.6, 28.8, and 28.9).

28.4.6 Full- Versus Part-Time Bracing

Full- versus part-time bracing is often debated, with advocates for each. Definitions of "full-time" vary greatly; in our program, our "full-time" goal is 20 h daily, with additional time-out of the brace allowed for organized sports. Given the success of casting which is a full-time device, it makes inherent sense that the brace should be worn as much as possible. While traditional

Fig. 28.8 Idiopathic early onset scoliosis. This 18-month-old child (**a**) with idiopathic EOS did not tolerate an attempted cast treatment. By 24 months (**b**), the curve had increased further and a custom-molded TLSO was begun with substantial in-brace correction (**c**). Substantial in-brace correction should be expected and sought. At the age of 4 years, bracing has continued and some correction is maintained (**d**). Full-time bracing for idiopathic EOS can be employed if casting is not tolerated, or vice versa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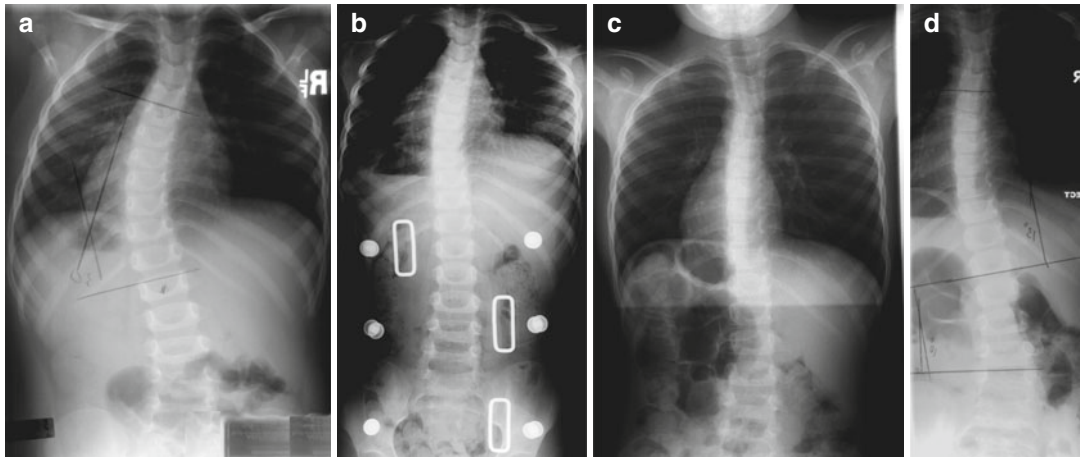

Fig. 28.9 Idiopathic early onset scoliosis. This child began full-time brace treatment at the age of 3 years for a persistent curve noted for 6 months (**a**). Good in-brace correction was achieved (**b**). At the age of 7 years, curve is minimal out of the brace (**c**) and brace was discontinued. By the age of 9 years, there had been some reoccurrence of the curve and part-time bracing was initiated (**d**)

bracing programs (MWB, Wilmington, Boston) have included "full-time" bracing, success has also been noted with part-time bracing systems (Charleston, Providence). For adolescent idiopathic scoliosis, meta-analysis [7] suggests full-time use is more effective than part-time use. Weinstein et al. demonstrated prospectively that increased time in the brace correlated with more success [8]. Katz et al. [58] showed that particularly for larger curves, full-time Boston Brace use was more effective than part-time Charleston bracing. Yet, they also demonstrated that for smaller single thoracolumbar or single lumbar curves, a part-time Charleston brace was as effective as a full-time orthosis. Two series of brace treatment of idiopathic EOS [20, 27] used part-time bracing and generally noted success. Tolo and Gillespie [30] began with patients with full-time, and then switched many patients to part-time when the curve was controlled, noting success with part-time use when RVAD fell toward zero or became negative with treatment. This has been similar to our practice; we encourage full-time use but if the curve as measured out of the brace is greatly reduced and below approximately 15°, part-time use will be instituted with close observation, particularly in the preadolescent growth phase. Many patients will need to shift back to full-time use during the preadolescent growth phase.

28.4.7 A Team Approach

A team approach to management of bracing of all ages is sought at most pediatric deformity centers and is felt by the proponents of Milwaukee, Wilmington, and Boston bracing to be germane to successful bracing. Typically, the "team" is composed of physician, orthotist, physical therapist, and nurse or other coordinator. We also view the family and patient as part of the team. Decisions and assessment of progress are made openly where possible, and if at all feasible, all members see the patient at each visit, assessing the patient's progress and the fit of the brace, a practice that we think fosters better brace wear compliance by the patient and family. All members of the team understand brace construction principles, and are encouraged to assess and critique the individual orthosis. This model is possible at a medical center, but more difficult at individual office practices or satellite centers. Still, it is possible to promote team communication with orthotist, physical therapist, and coordinator, even though they may be physically separated. Each patient represents a unique deformity, with individual physical findings, contractures, and curve patterns as well as specific lifestyle, activity, and emotional needs. Each patient deserves the best effort of each of these team members.

28.5 Current and Future Developments

Developments in imaging technology, computer-assisted modeling (see Chap. 9), compliance monitors, and genetics may improve the quality and specificity of bracing for idiopathic EOS. Dubousset et al. [60] and others have described the rapid low-dose acquisition of three-dimensional images. Surface topography is used by some centers to monitor for scoliosis and reduce radiation exposure. Nevertheless, the PA radiography remains the "gold standard" of assessment of the scoliosis patient. The availability of such images on a regular basis should help rationalize treatment decisions about early onset spinal deformity and permit brace construction based on three-dimensional data. Ogilvie et al. [61] have studied the genetic predisposition to idiopathic scoliosis in a limited population and suggested that it may be possible to predict progressive idiopathic scoliosis on the basis of genetic markers. The ability to identify patients at risk and initiate nonsurgical treatment early, in theory would facilitate earlier and more successful treatment of idiopathic EOS before curves have become too structural. Computer-aided acquisition of brace shapes from body contours [43, 57, 62, 63] has been reported to improve patient's acceptance and patient's fit of rigid TLSO braces. Aubin et al. [64] have been able to correlate real-time measurement of forces between pads and the patient with computer-generated models, and demonstrate improved brace correction with such designs. Effective braces presently require a maximum in orthotist skill and experience. Brace design and construction according to computer-generated guidelines offer the hope of more widely available, effective orthosis for the treatment of idiopathic EOS. The psychological impact of bracing and other treatments (casting, surgical) on the child and family is not known and must be studied. Instruments specific to this population (EOSQ) will be helpful in monitoring treatment success in conjunction with radiographic measurements [65, 66].

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Casting for Early Onset Scoliosis

29

James O. Sanders

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Key Points

- Among the types of early onset scoliosis, idiopathic early onset scoliosis is the most amenable to casting.
- Progressive idiopathic early onset scoliosis is potentially fatal.
- Proper derotational casting does not appear to be associated with increasing rib or chest wall deformity.
- Serial derotational casting in younger, particularly non-syndromic, children can result in a cure.
- Casting has an important role in older children in delaying the need for surgical intervention.

29.1 Introduction

Of the various types of early onset scoliosis (EOS), idiopathic EOS is the most amenable to casting and may be either resolving or progressive with resolving curves being far more common. Scoliosis presenting during the first year of life has a greater likelihood of resolving spontaneously, whereas curves developing after 1 year of age have a worse prognosis [1, 2]. Mehta was able to distinguish resolving from progressive scoliosis by using the rib vertebral angle difference (RVAD) on an early radiograph [3] with her later work noting this was

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obtained in the supine position [4]. Left untreated, the prognosis for curves that do progress is invariably poor; by age 5, 57 % of untreated children will have a curve greater than 70° [3]. These large thoracic curves can cause restrictive lung disease or thoracic insufficiency syndrome (TIS) characterized by decreased thoracic growth and lung volume, inhibiting alveolar development and lung function, which may cause respiratory failure, pulmonary hypertension and cor pulmonale, and death at an early age [3, 5–8] . Patients with progressive curves progress by about 5° per year reaching 70° or more by age 10 [9, 10]. Thoracic curves >70° in adolescence are associated with significantly lower forced expiratory volume in 1 s (FEV1) and forced vital capacity (FVC) values compared to patients with smaller deformities.

29.2 Radiographic Evaluation

Both the rib vertebral angle difference (RVAD) and rib phase are important for predicting which curves will progress [3]. The RVAD is the difference of the angles made between each rib and the corresponding vertebral body. This is measured at the vertebral level with the greatest angular difference between the concave and convex rib. Rib phase is classified as phase 1 or 2 depending on whether or not spinal rotation causes the rib head to overlap the vertebral body (phase 1 – no overlap, phase 2 – overlap). Eighty-three percent of resolving curves have a RVAD of <20° and 83 % of progressing curves have a RVAD of >20°. In progressive curves, the RVAD increases and the phase gradually transitions from 1 to 2. Phase 2 ribs are the hallmark of progressive curves as all phase 2 curves progress. Double curves present a special problem as most of them progress. The RVAD in double curves may be quite low, but an oblique 11th or 12th convex rib with lumbar rotation is a poor prognostic sign. Generally, the RVAD and phase are reliable, but the measurement error may make discernment difficult in marginal cases [11] in which case close observation, typically with repeat radiographs in 3 months, is the best course [3, 4].

29.3 Treatment

In the late 1950s and early 1960s, Harrington developed the first successful spinal instrumentation [12]. His earliest procedures were done without fusion and had a high rate of instrumentation failure leading him to supplement the instrumentation with a fusion, which subsequently created modern spinal deformity treatment. About the same time, James [13] identified a course of treatment for early onset scoliosis (EOS) persisting to the present – keep the spine growing while controlling the curve with whatever means you have (he had casts and the Milwaukee brace) until 10 years of age, and then do a definitive fusion. The current basic concept in most centers joins these two concepts by bracing, casting, or instrumentation without fusion so the spine can gain sufficient length to prevent pulmonary problems associated with fusion before age 10, after which a formal fusion is done if the curve magnitude warrants [14, 15]. Current treatments primarily the growing rods and VERTR [16], aim at delaying definitive fusion and almost never focus on a cure. Definitive fusion before 10 years of age is fraught with problems. Each of these methods has difficulties.

29.3.1 Orthotic Management

Bracing is the most common nonoperative treatment of EOS with variable success [17]. It remains an important adjunctive treatment in its treatment and plays an important role in delaying the need for surgery, but they are difficult to fit in young patients and have special problems in young children who have more pliable ribs than adolescents, and braces using a 3-point bend on the apical rib can create a chest wall deformation by pushing the ribs toward the spine. Bracing may be difficult to be properly applied each time to a young child's cylindrical shape. This is compounded by the need to make the brace sufficiently flexible for donning and doffing.

29.3.2 Growing Rod Techniques

There are currently three growing rod techniques – growing rods, VEPTR, and "Growth guidance – Shilla." Prospective studies of these techniques have shown that complications with all techniques are frustratingly frequent [18] and increase the longer a device is implanted. More frequent lengthenings appear beneficial in length gained, but more lengthenings also result in more complications, and there is not yet good evidence equating how much length must be gained for long-term pulmonary function. Our current estimate of 18–22 cm thoracic height (T1–T12) is based on data from those who had definitive fusions rather than growing rods [19]. Spontaneous fusion occurs with all current techniques [20–22]. The law of diminishing returns [23] is real and means you can often get 3–4 years of effective lengthening. This has led to the realization that if you can delay growing instrumentation until 6 or 7 years of age, you have a much greater chance of reaching 10 years of age before performing a final fusion.

29.3.3 Serial Casting

Casting represents another alternative for scoliosis which was quite common until the development of effective spinal instrumentation. Casting itself can create pressure sores, significant rib or mandibular deformities, and constrict the chest. "Cast syndrome" even denotes the historical term for superior mesenteric artery syndrome. However, many of these problems seem to be the result of indiscriminate casting of all types of scoliosis with use of improper techniques combined with a limited understanding of spinal and particularly of chest wall deformities. Scoliosis casting comes in several varieties. The most commonly used method in the United States was Risser casting [1], which uses a three-point bend for correction. While it is possible to obtain significant curve correction with this technique, it does not sufficiently account for rotational abnormality, and, especially in younger children with flexible bones, can cause significant rib deformities and chest constriction. Mehta [4] described her results of casting in 136 patients with IIS using the technique of Cotrel and Morel [24] with the philosophy that early rapid growth, if guided by the cast, would assist an initially curved spine to straighten. We have used serial casting in a select group of patients with success in curing the curve in younger patients with less severe curves and in delaying surgery in older children [25].

29.4 Influence of Age and Etiology

The age at treatment onset and etiology are crucial and significant factors in treatment success. Mehta found casting much more likely to be successful if started under age 2 years. Our results are nearly identical to Mehta's with the patients achieving nearly full correction starting at average age of 1.1 years and full correction rare in those started over age 18 months. Casting to resolution typically takes a year or more. Mehta identified four physiological patterns: a "sturdy phenotype" with good muscle mass and tone, a "slender phenotype" with more delicate features, ligamentous laxity, and more rapidly progressive curves, those with known syndromes, and those with unknown syndromes. In her series, all groups responded if treatment was initiated early with smaller curves. In older patients with larger curves, the prognosis worsened from the sturdy to the slender to named and unnamed syndromes. We have not found her classification of slender and sturdy reproducible in our hands and have divided the patients into the simple categories of idiopathic or syndromic. Older and syndromic patients usually have less correction. Our goal in these older and syndromic patients is to delay the need for surgery until the spine has achieved sufficient growth for good pulmonary function as an adult. Other investigators had since duplicated our results [26, 27] and found casting helpful in both resolving smaller curves and in delaying

surgery in the larger curves and patients with syndromic etiologies. In each of the published series, casting has markedly delayed or even eliminated the need for growing rods in a majority of patients. A matched cohort controlled study of growing rod to cast patients showed similar growth in both but with less complications in the cast group compared to the growing rod group [28]. The natural history following casting has not been well defined through the adolescent growth spurt, but we suspect many patients will need surgery at that time. Our recent results show 27 % of casted patients resolved their scoliosis, 56 % improved but did not resolve, 14 % remained stable, and 3 % progressed during casting. To date, only 10 % have had surgery, though this increased to 28 % of curves 50° or more at the start of casting. Among those who have had surgery, it was delayed by an average 2.7 years after the start of casting. We have not typically casted children with neuromuscular disorder being skeptical that casting has a major role in those curve types, though this hypothesis has not been widely tested.

29.5 Technique of Casting

Once the diagnosis of progressive scoliosis is made, based upon either a progressive major curve or a RVAD of more than 20° at presentation, casting is recommended. We have typically required an MRI of the spine before casting, but some centers initiate casting prior to the MRI. Mehta's program consists of cast changes under anesthesia in younger patients every 8–16 weeks until the curve was nearly resolved followed by an underarm brace which may be weaned if the patient's curve correction continues. We base our cast changes on the child's growth rate with changes every 2 months for those 2 years and under, 3 months for those aged 3 years, and 4 months for those aged 4 and above. We aim for curves less than 10° supine out of the cast and then use a brace molded under anesthesia just like the cast. Children are occasionally braced during the summer months with resumption of casting in the fall. We have also used a waterproof cast in some smaller curves that we are trying to hold stable during the summers to allow children to swim and bathe using a Gortex pantaloon used upside down as a shirt with waterproof padding.

A proper casting table is crucial. We have used both a Risser and a Cotrel frame, but found them quite large for small children. Mehta has designed a table marketed by AMIL which leaves the head, arms, and legs supported but the body free. The Salt Lake City Shriners Hospital uses a custom table which performs a similar function of supporting the child in traction while leaving the body free for the cast application. Patients are intubated since thoracic pressure during the cast molding can make ventilation temporarily difficult. Older children might be successfully managed without anesthesia, but it is impractical in very young children. A silver impregnated shirt is used as the innermost layer. Head halter and pelvic traction assists in stabilizing the patient and in narrowing the body (Fig. 29.1). Even though traction can correct the curve while applied, the position cannot be retained in the cast once traction is released and the body recoils unless the cast also supports the occiput or the mandible. For curves with an apex above T8, the shoulders are incorporate, and high thoracic curves may require a mandibular extension. A mirror slanted under the table is useful for visualizing the rib prominence, the posterior cast, and the molds. A thin layer of webril is applied with occasional felt on significant bony prominences. Mehta uses crape paper which is removed after the casting leaving plaster on the skin except at pressure points protected by felt. If there is a lumbar curve, the hips are slightly flexed to decrease lumbar lordosis and facilitate curve correction.

Plaster, or, in certain cases mentioned previously, fiberglass, is applied. The pelvis portion, as the foundation, is well molded. It important that the cast does not push the ribs towards the spine and consequently narrow the space available for the lung. Rather, the posteriorly rotated ribs are rotated anteriorly to create a more normal chest configuration and counter rotation is applied through the pelvic mold and upper torso (Fig. 29.2). While the Cotrel/Morel technique and

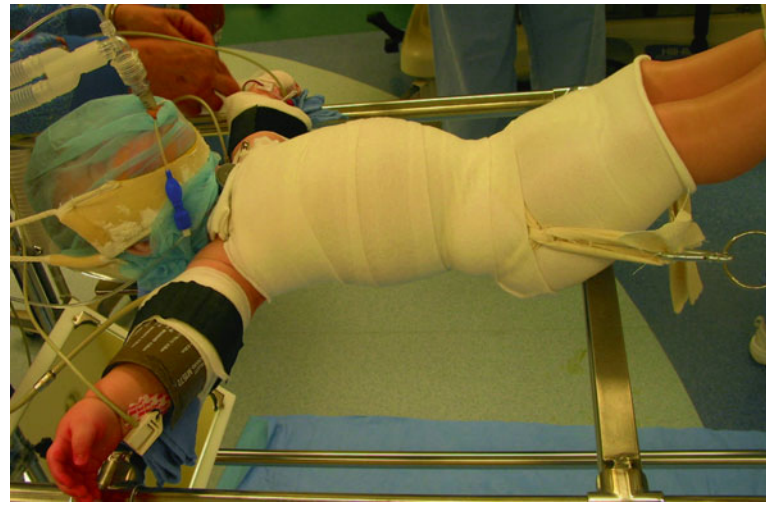

Fig. 29.1 Traction applied on the casting frame with the torso free

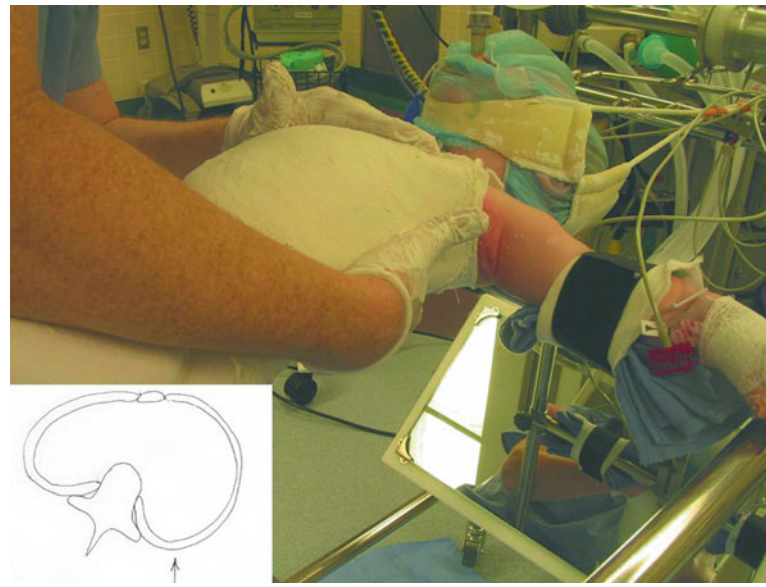

Fig. 29.2 Molding of the cast by derotation of the thorax

Mehta's modifications use an over-the-shoulder cast, we have had excellent success staying below the shoulders since most infantile curves have low apices, typically at T10 to 11 with nearly identical results to Mehta's. An anterior window is made to relieve the chest and abdomen while preventing the lower ribs from rotating (Fig. 29.3). A posterior window is made on the concave side allowing the depressed concave ribs and spine to move posteriorly (Fig. 29.4). A proper cast corrects the curve and the rotation without deforming the ribs towards the spine (Fig. 29.5).

Initiation of casting at a younger age, moderate curve size (<50°) and an idiopathic diagnosis carry a better prognosis than an older age of initiation, curve >50° and a non-idiopathic diagnosis. We have had no worsening of the space available for the lung (SAL) and no worsening rib deformities.

Serial casting for idiopathic EOS often results in full correction in young patients with idiopathic curves less than 50°. Casting for older patients with larger curves or non-idiopathic diagnosis still results in curve improvement and a long

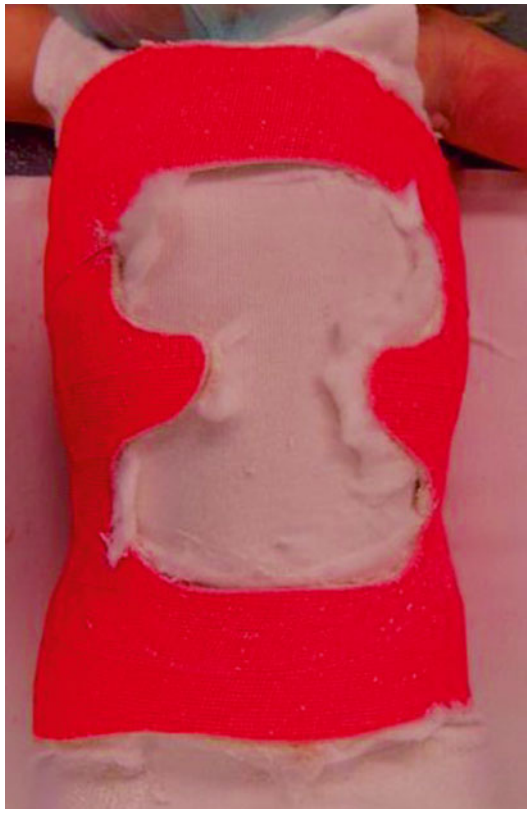

Fig. 29.3 Trim of the anterior windows leaving the abdomen open to expand

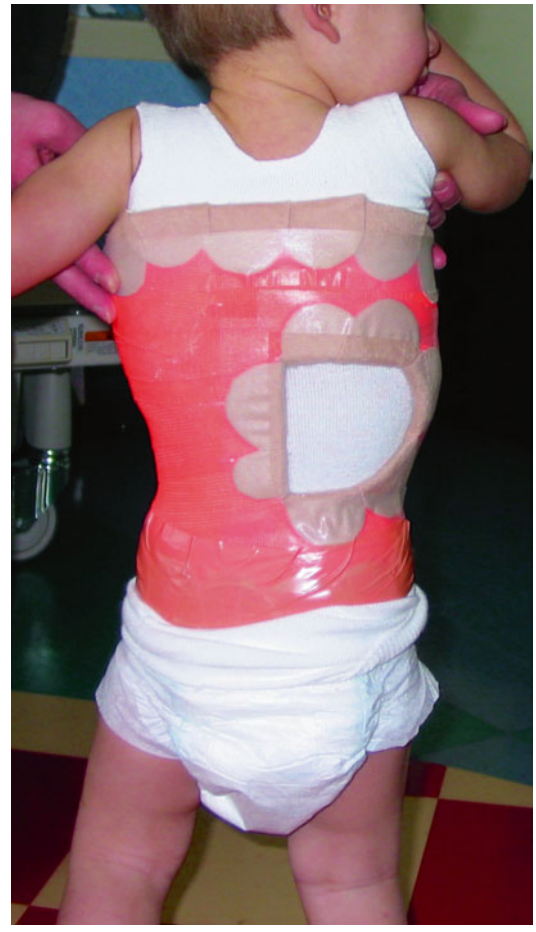

Fig. 29.4 Posterior window on the concavity allowing the spine to rotate into the window

delay in the time before surgery. The Cotrel technique of derotation casting appears to play a role in the treatment of progressive scoliosis with cures in young patients and reductions in curve size with a delay in surgery in older and syndromic patients.

29.5.1 Barriers to Successful Cast Treatment

While children tolerate casting very well, it can sometimes be difficult to convince the parents beforehand. Although casting is not difficult, it does require some training and proper equipment. We have found differences in the local compared to the jet set parents – the latter are often very vested in the treatment and will fly to the ends of the world to make it happen while the former may be nonchalant or even extremely disinterested. Parents may be disappointed if their child's curve is not cured but only stabilized or decreased, and early education and proper expectations on the pulmonary problems from early onset scoliosis is important for parental understanding. There is no avoiding the issue that casting requires anesthesia every few months and the uncertainty this entails for cognitive development, though non-magnetic growing rods face similar issues.

On the other hand, casting can certainly delay surgery and does not appear to cause troubles either with spine growth or the success of later surgery, is very effective in delaying surgery and is curative in younger children with smaller curves and has become a very important tool in treating early onset scoliosis.

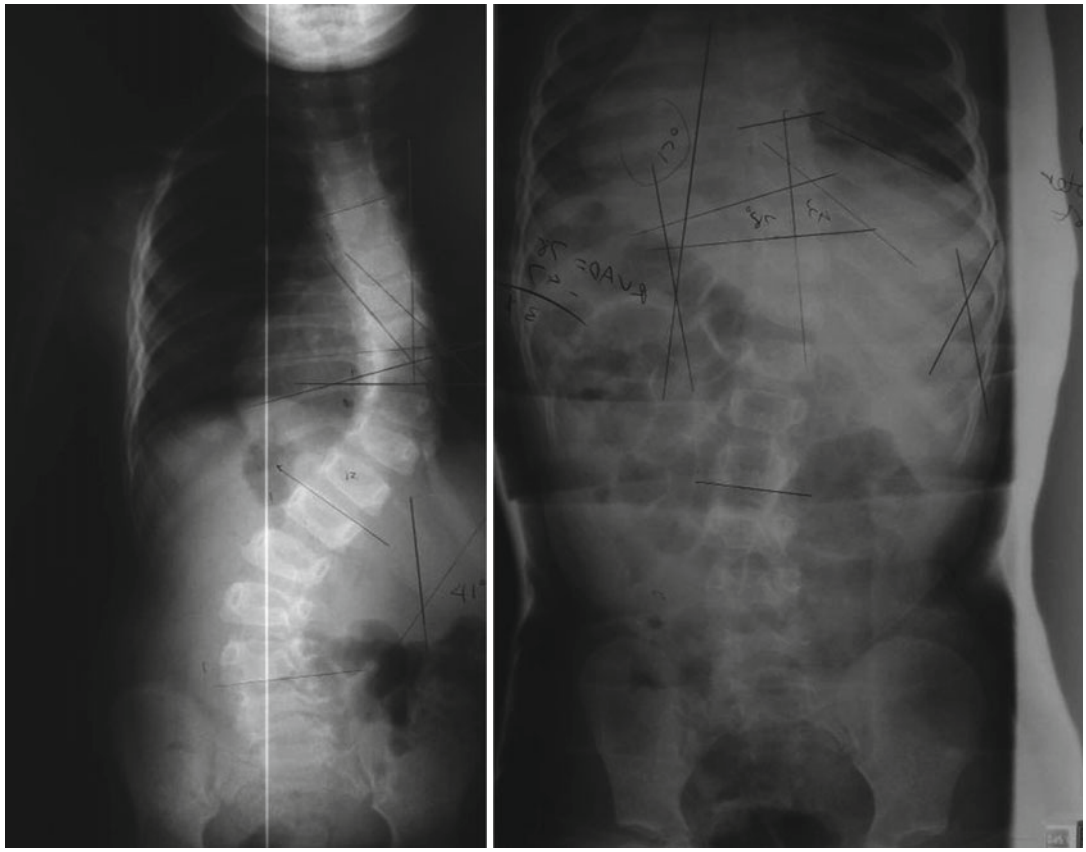

Fig. 29.5 Radiographs comparing the curve before and in the cast. A proper cast does not push the ribs into the spine

Conclusions

Idiopathic early onset scoliosis is the most likely type to respond to casting though we have also found an important role in syndromic scoliosis. We have not used it routinely for congenital or neuromuscular curves. Proper casting plays an important goal role? Both delaying and preventing the need for early surgery, and particularly in infantile idiopathic curves, can result in full curve correction.

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Halo-Gravity Traction

30

Charles E. Johnston

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Key Points

- HGT is indicated in diminutive, osteopenic patients with severe and rigid spinal deformity as preliminary corrective adjunct prior to operative management.
- HGT is valuable in patients with respiratory impairment due to upright positioning and improved diaphragmatic excursion.
- HGT is a safe (1–1.5 % incidence of neurologic complication) and effective (~30 % improvement in scoliosis, kyphosis, and thoracic length) method of non-operative deformity correction.
- HGT is contraindicated in patients with insufficient skull bone stock, space-occupying lesions of the spinal cord, or severe canal distortion with stenosis.

30.1 Introduction

Halo-gravity traction (HGT) is a time-tested orthopedic technique to produce, among other things, correction of spinal deformity. As applied to early onset spine deformity, it can be used effectively to obtain correction prior to operative treatment, or as a delaying tactic to produce correction so that a non-operative method can be

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applied. In the past, traction has been applied using various neck and head halters combined with pelvic or leg harnesses or by skeletal traction. Most methods require the patient to remain supine in bed with longitudinal traction being applied via attachments to the bed frame, thus immobilizing the patient in a non-upright and non-movable position. Not surprisingly, the duration of such traction is limited by physical, psychological, and medical complications which can befall a bedridden patient.

Because the HGT is applied using a halo ring device, discomfort or intolerance from external chin or occipital harness attachments are avoided. Head halter methods are notorious for producing chin and facial discomfort and irritation, thus limiting the effectiveness of the method. Because gravity is the method of force application to the lower body, the pelvis and the legs remain unrestrained, encouraging patient mobility. Most young patients seem totally unaware of the presence of the halo once they recover from the immediate pain of application – a process which often takes no more than 24 h following application. From that point on, the morbidity from HGT is so minimal that it can be continued for several months – and in exceptional situations, years if necessary (Fig. 30.1), to produce valuable deformity correction as well as improve respiratory status in anticipation of further surgical or non-operative treatment.

30.2 Indications

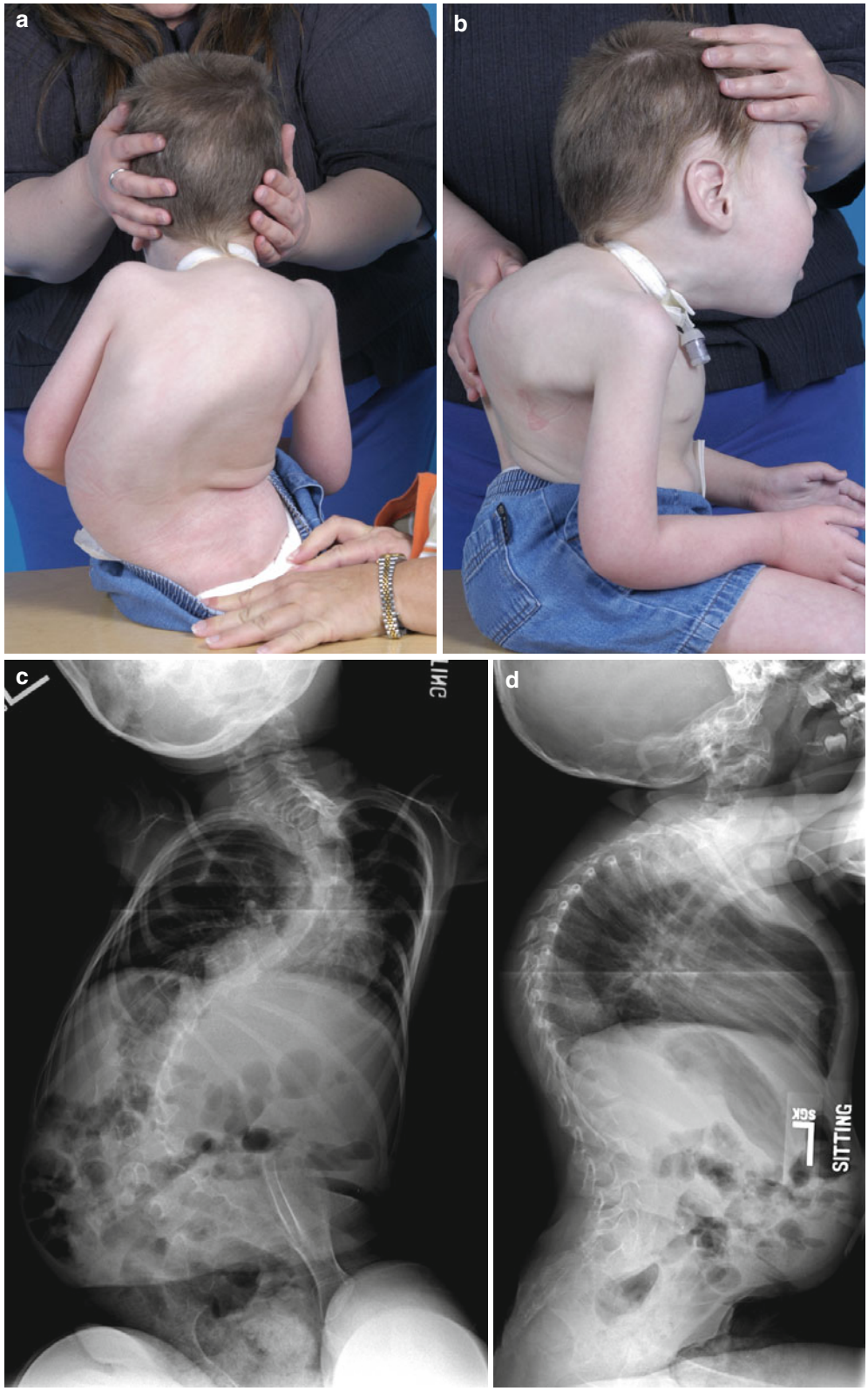
Patients with early onset spinal deformity often present with co-morbidities and physical characteristics which can significantly challenge and compromise *any* surgical treatment plan. For example, those with syndromic or "exotic" diagnoses possess diminutive osteopenic bony elements, which can severely limit acute deformity correction due to weakness of the bone-implant interface. Frequently, their deformity is rigid and/or kyphotic, in which case posterior distractive methods of correction (rod systems, VEPTR's) are compromised by the need for extreme contouring of the expandable device, leading to ineffective distraction and proximal anchor failure by posterior cut-out, if not peri-operatively, then later by fatigue "plowing" or fracture due to the unfavorable biomechanical forces – *especially* in kyphosis (Fig. 30.2).

Neurological injury from acute correction is always a concern, especially if severe deformity requires canal manipulation by osteotomy or vertebral resection to achieve it. Relative canal stenosis is another source of neurological risk with acute correction, especially if previous fusion produces cord compression at a junctional segment due to fusion mass overgrowth or a juxtafusion hypermobile segment [1, 2].

Patients with severe thoracic deformity, especially as a sequela of neglect or ineffective previous treatment, may present with respiratory impairment, in which case HGT is indicated as a preliminary step to improve respiratory mechanics and make them a more suitable surgical candidate. Such respiratory compromise, as well as hypotonia and weakness, chest wall defects, skin intolerance or anesthesia, and mental retardation may eliminate external means of deformity control, such as bracing or casting, from consideration.

Patients with severe deformity are also not candidates for bracing or casting purely from

Fig. 30.1 (**a**, **b**) A 4-year-old boy with collapsing kyphoscoliosis due to congenital myopathy (March 2007); (**c**, **d**) scoliosis curves measuring 95 and 90°, with 85° kyphosis; (**e**) correction after 2 months HGT; (**f**, **g**) placement of rib-pelvis and spine-pelvis construct (August 2007), followed by one interval lengthening; (**h**) rod erosion through skin (December 2008). Treatment by wound vac: (**i**) removal of right rods (January 2009); (**j**, **k**) removal of all implants (January 2010) due to unresolving wound sepsis. HGT was re-started: (**l, m**) 4 years later (January 2014). Continuous traction has stabilized deformity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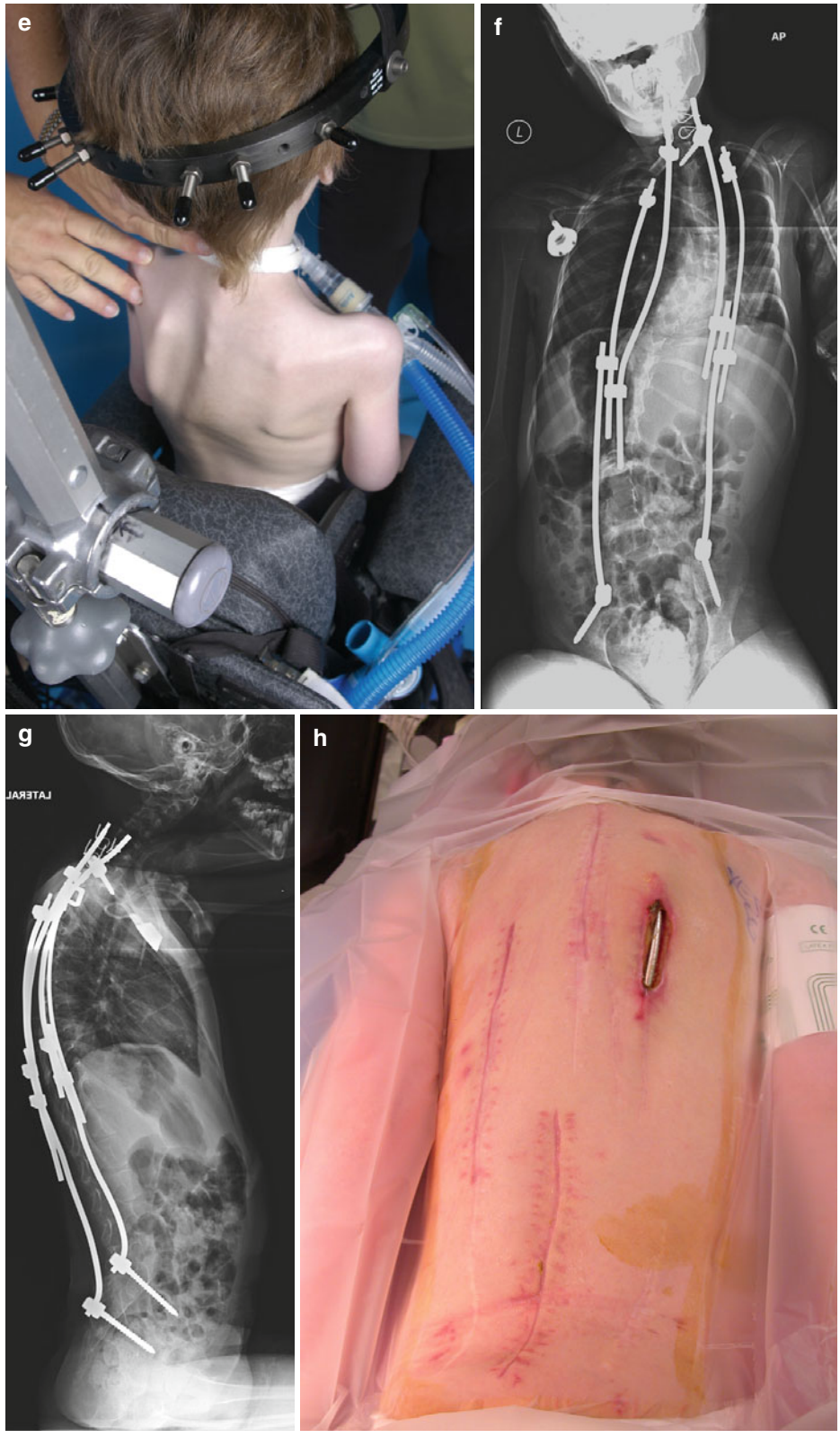

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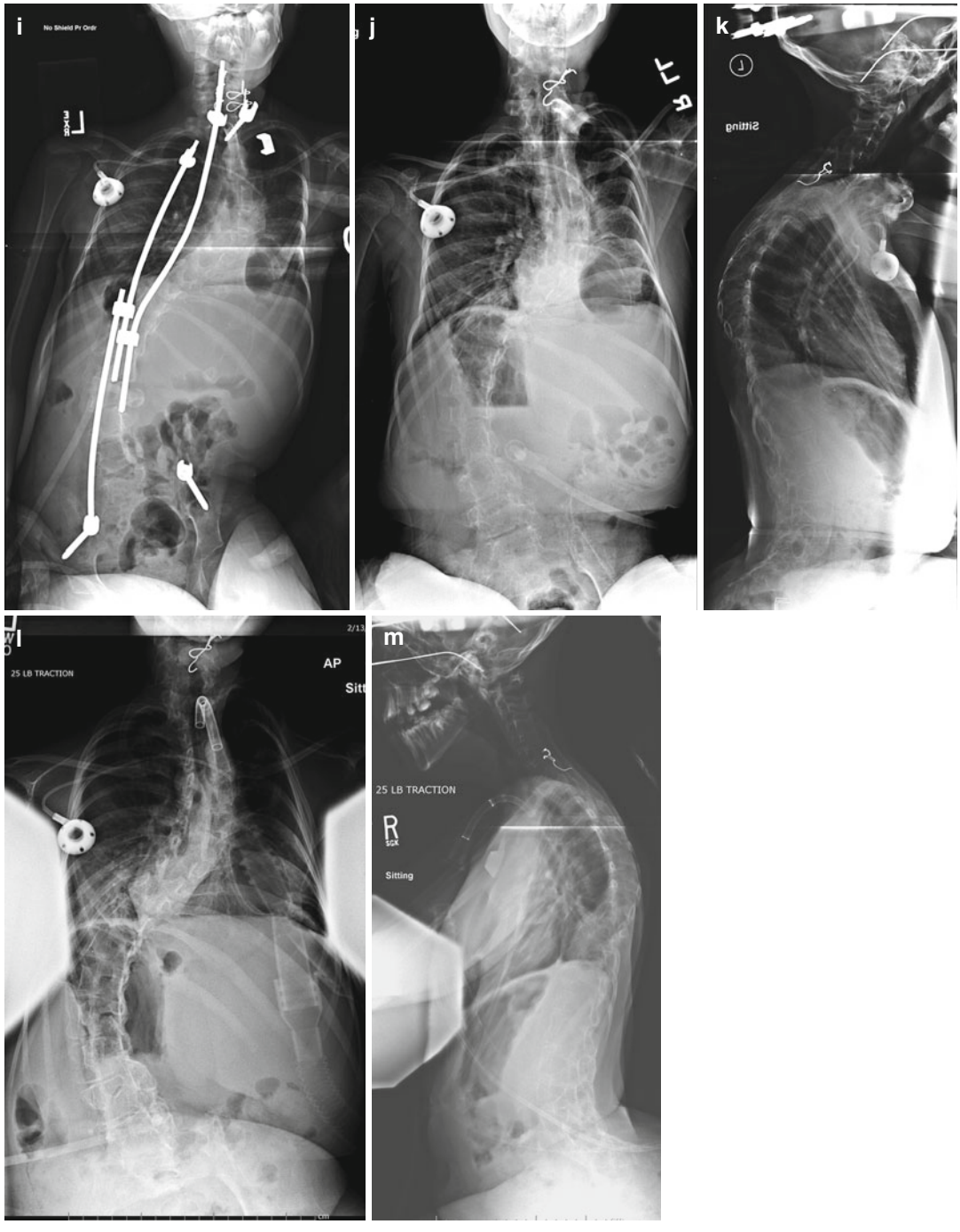

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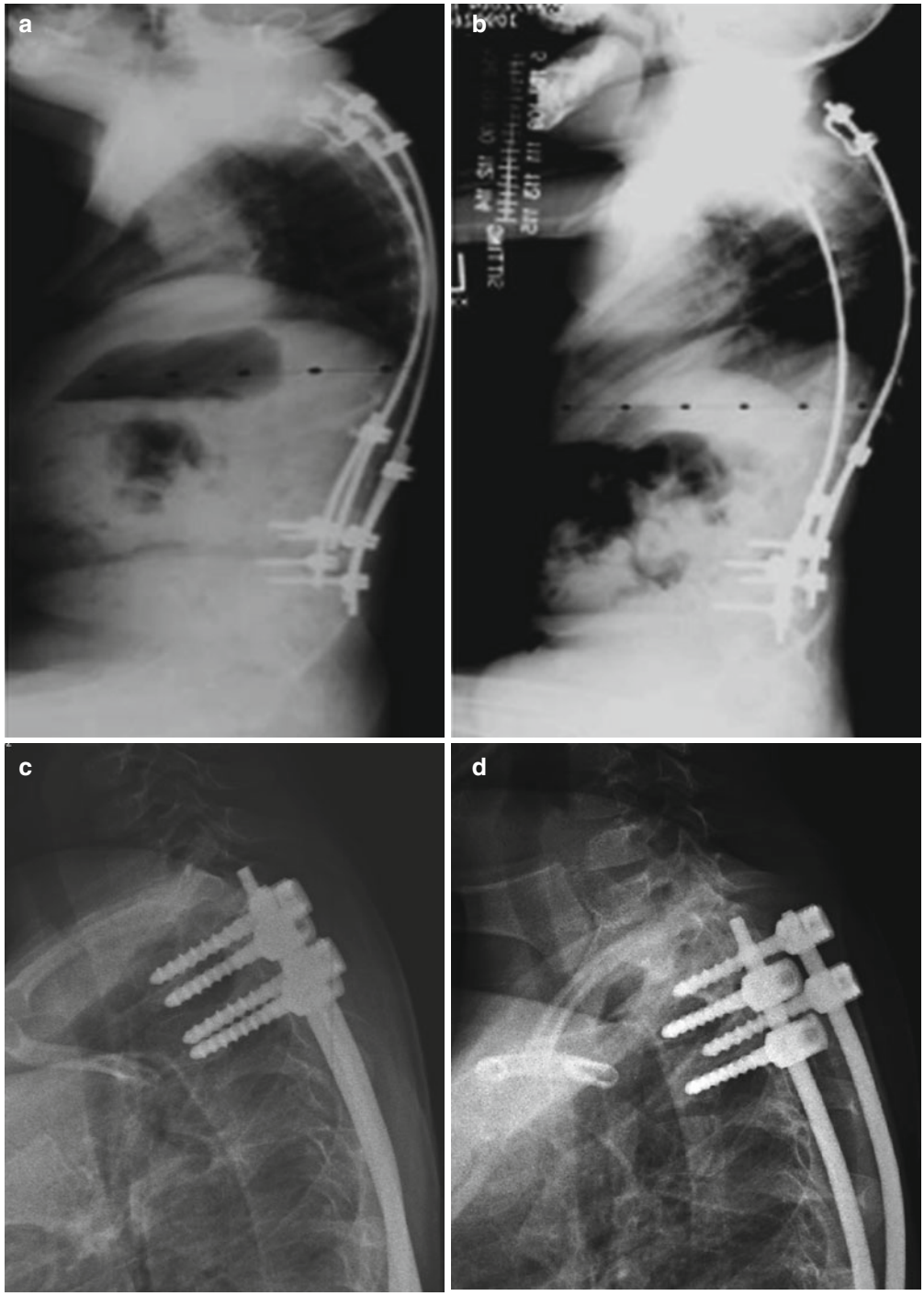

Fig. 30.2 (**a**, **b**) Proximal rib hook anchors failing over a period of 1 year in a collapsing kyphotic deformity; (**c**, **d**) thoracic pedicle screw pullout within the first year after implantation in an ambulatory patient

biomechanical considerations related to curve magnitude. Curves exceeding approximately 53° are corrected more effectively by longitudinal distraction forces [3], rather than laterally applied transverse forces realized with a cast or brace. Large, stiff curves thus may not benefit from use of the latter, being poorly tolerated due to excess skin pressure associated with inefficient transverse loads, as well as rib and chest wall deformation caused by the lateral rib pressure. In these instances, HGT is an effective method to achieve deformity correction, and indirectly, improve respiratory mechanics [4, 5]. We have noted up to 10 % increase in predicted vital capacity *acutely* in several patients who benefit from elongation of the chest wall associated with the spinal elongation/correction (Fig. 30.3). Improving the restrictive component of the deformity probably results from more efficient diaphragmatic excursion in the elongated trunk as well as from rib separation on the concavity, providing more effective inspiration and consequently respiration. This appears to be the physiologic explanation for this acute vital capacity increase during HGT.

30.3 Contraindications

HGT has been found to be almost uniformly safe [4–7]. The only absolute *contraindications* to its use would be bone stock in the skull insufficient to gain halo purchase due to underlying diagnoses such as osteogenesis imperfecta or fibrous dysplasia (Fig. 30.4); presence of an intra- or extra-medullary lesion (tumor, syrinx), with or without pre-existing neurologic deficit (Fig. 30.5); severe canal distortion with stenosis [1]. Otherwise, any patient with severe rigid deformity with or without kyphosis, potential or actual thoracic insufficiency syndrome, osteopenia, and increased potential neurological risk from acute instrumented correction is a candidate for HGT as a preliminary step before other operative treatment, to reduce the occurrence of instrumentation or neurological complications and to improve respiratory function and suitability for general anesthesia.

30.4 Technique

HGT is not a new method of treatment of spinal deformity, having been developed soon after the halo apparatus was first described in the 1960s at Rancho Los Amigos Hospital [8]. Stagnara [9] popularized the gravity-traction method, which was introduced at our institution after it was demonstrated to the author by Klaus Zielke in 1984 during a visit to the latter's clinic in Germany. The indications for which these early authors used HGT were essentially the same as its use for us today – neuromuscular "collapsing" deformity in the Rancho experience and as an adjunct to neglected deformity in older patients with respiratory insufficiency, as well as in the young patient with syndromic or exotic spinal deformity.

Halo application requires general anesthesia for children of this age group, using the maximum number of pins possible [5, 10] (Fig. 30.6a). Experience has shown that the use of numerous pins actually decreases the incidence of pin infection or loosening of any *single* pin. Pin direction should also be as perpendicular to the skull as possible [11, 12]. Pins are tightened to a torque approximately equaling the age of the child up to a maximum of 8 inch-pounds, e.g., a 4-year-old patient's pins are tightened to 4 inch-pounds of torque, using a calibrated torque wrench. Because of variation in skull thickness and location of sutures, computed tomography (CT) scanning of the skull has been recommended to control pin placement [13, 14], but in practice such thickness determination has not altered intended pin location when multiple pins are used and the penetration is controlled by torque determination. Frontal and occipital areas are commonly adequate for safe, secure placement. Upright overhead traction via a traction bale attached to a wheelchair or standing frame, using a spring-loaded fish scale or other dynamic traction device (Fig. 30.6b–d) is begun the following day, initially with 5–10 pounds of traction. The amount of time and weight is increased to tolerance under careful neurological surveillance. All patients should have cranial nerve testing once a shift while upright in traction, as well as motor and

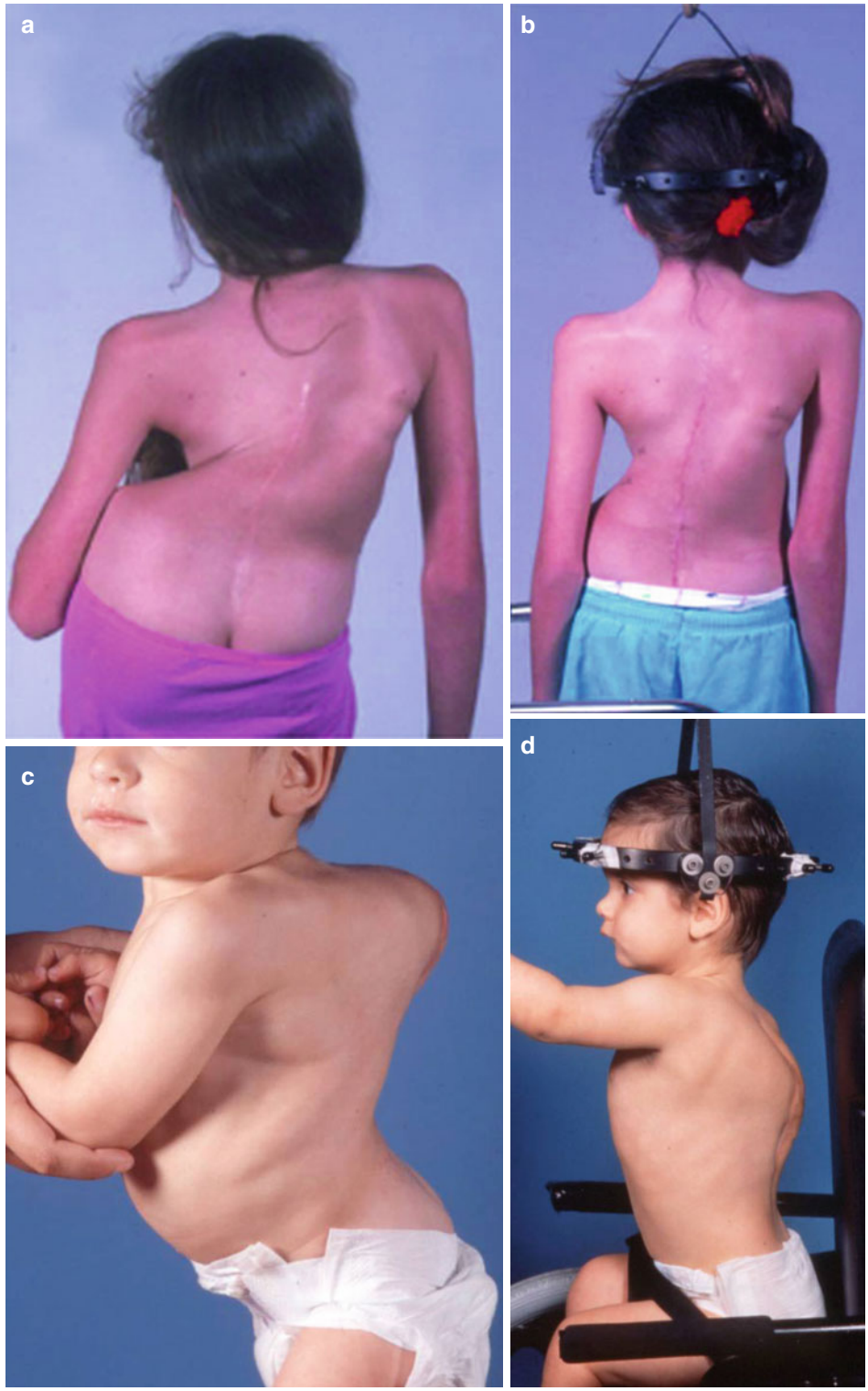

Fig. 30.3 (**a**, **b**) Elongation of the thorax by HGT following osteotomies of previous fusion mass; (**c**, **d**) elongation with kyphosis correction

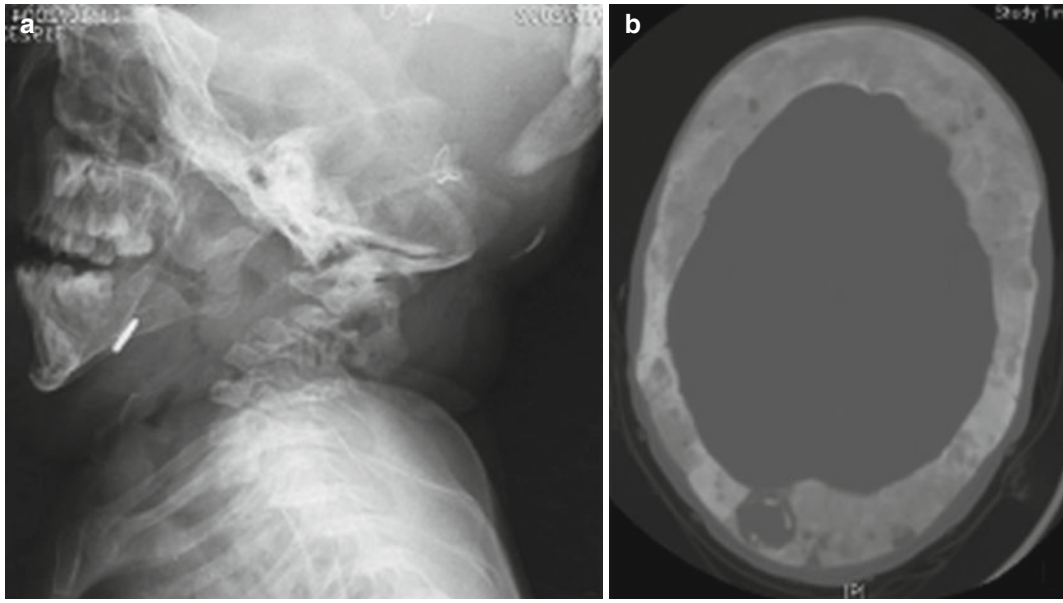

Fig. 30.4 (**a**) Absence of occipital bone in a patient with Loeys-Dietz syndrome. This patient is unsuitable for halo placement; (**b**) severe skull involvement with fibrous dysplasia, also unsuitable for halo placement

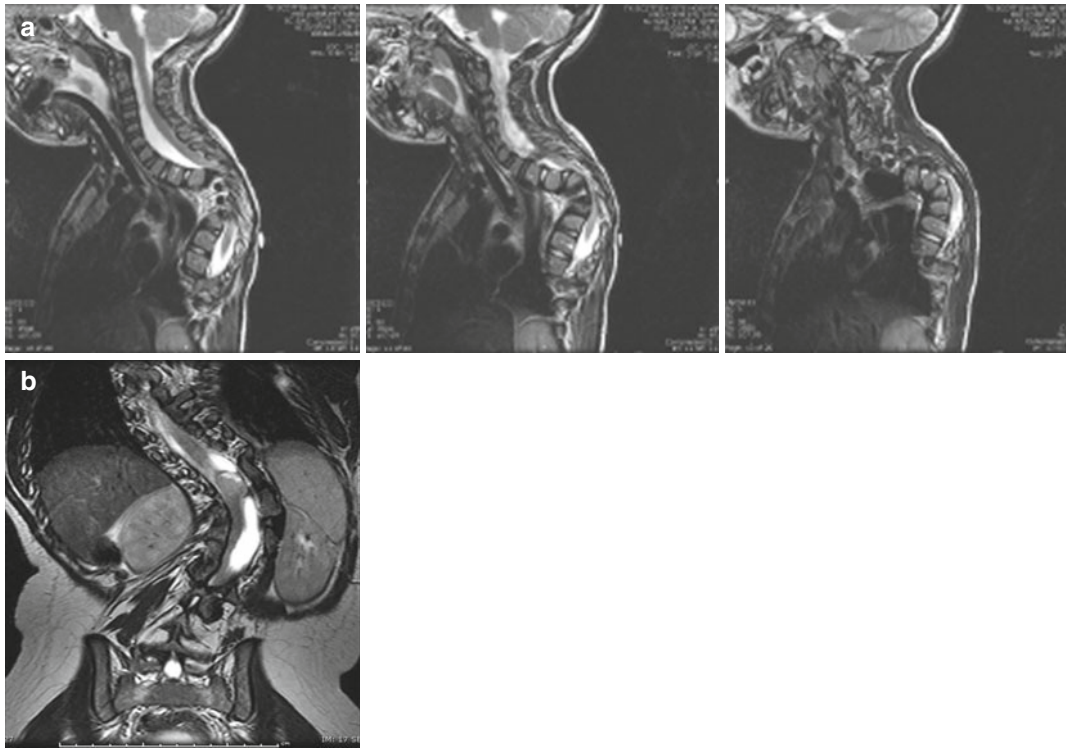

Fig. 30.5 (**a**) Cord compression with paraparesis in a 6-year-old patient with Pierre-Robin syndrome. Surgical decompression, not HGT, is indicated; (**b**) MRI revealing cystic astrocytoma in a 7-year-old male who was neurologically normal. HGT was started and he rapidly became paraparetic, which did not resolve with discontinuing the traction

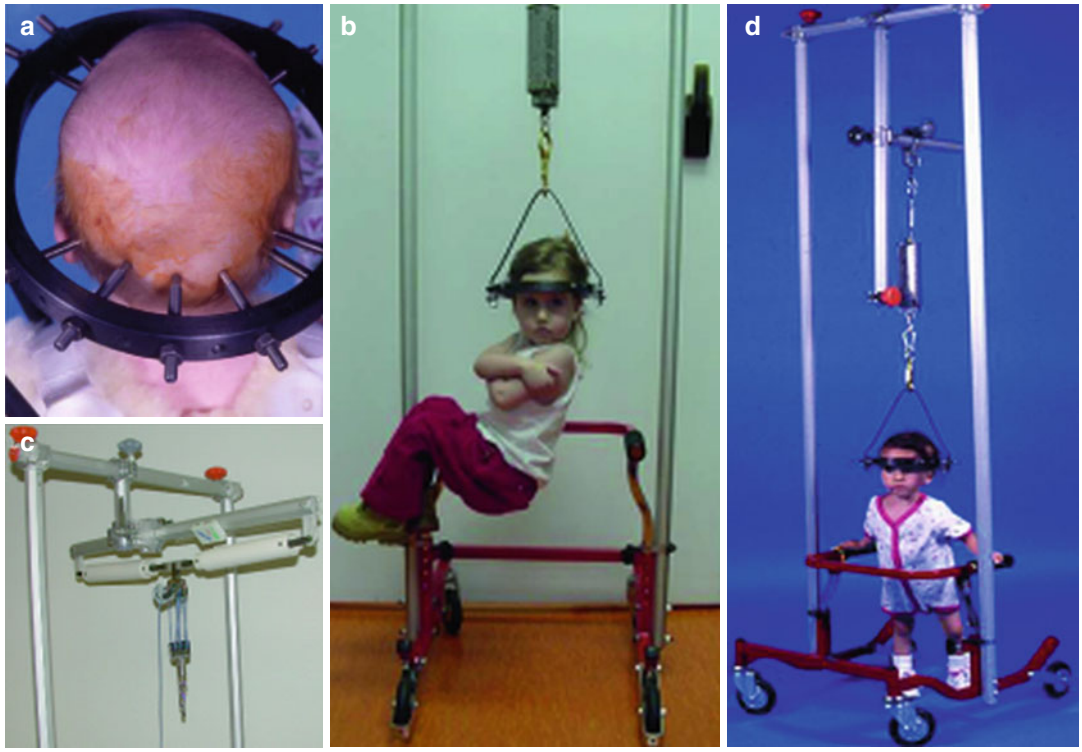

Fig. 30.6 (**a**) Ten pins in the skull of a 9-month-old infant. (**b**) Dynamic traction using fish scale spring-loaded device. Dangling is not encouraged; (**c**) currently used dynamic double-spring-loaded apparatus; (**d**) obligatory toe-walking while in overhead HGT. This is an appropriate upper limit of traction weight

sensory testing of upper and lower extremities, especially during the phase of increasing traction. Eventually traction force exceeding 50 % of body weight may be achieved, with cervical pain being the usual limiting factor. The goal of just lifting the patient's buttocks off the wheelchair seat while sitting, or being on tiptoes in the standing frame can usually be attained within 2 weeks (see Fig. 30.6d). The use of nighttime traction, making the treatment program more or less continuous, can be added by providing a cervical traction frame to the patient's bed, usually a gatched bed with the head portion elevated to act as a counter-traction [15]. Out-patient (home) traction can be attempted if caregivers are appropriately trained and vigilant. Radiographs are obtained every 3–4 weeks until a plateau of correction is reached. Remaining in traction without complication and with gradual deformity improvement over a period of 6 months is *not* unusual. In selected instances, in patients deemed *too* fragile for operative treatment or in whom operative treatment has failed and been abandoned, we have successfully treated severe uncorrectable deformity by extending HGT treatment *indefinitely* (Fig. 30.1).

30.5 Complications

Complications of traction include pin site infections, which are relatively common (up to 20 % incidence) [4, 5, 7, 15–17] but also usually controlled with oral antibiotics and pin care. As mentioned earlier, placement of extra pins at the time of halo application usually decreases the incidence of individual pin sepsis by presumably making the halo-cranial interface more stable. Intra-cranial abscess from septic pin penetration of the dura has previously been emphasized as an uncommon but serious halo complication [7, 17], although this complication is now rarely reported

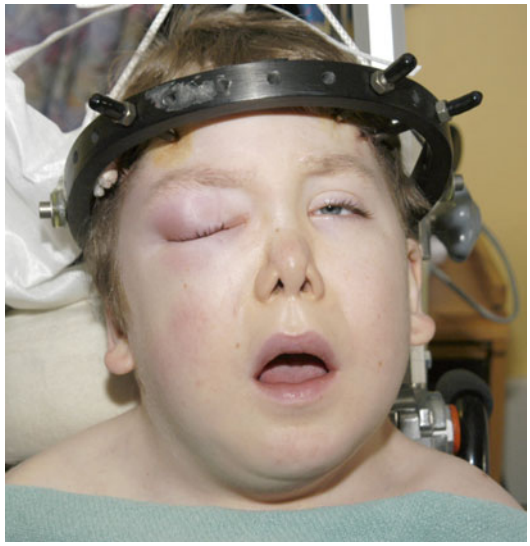

Fig. 30.7 Peri-orbital cellulitis associated with frontal pin infection

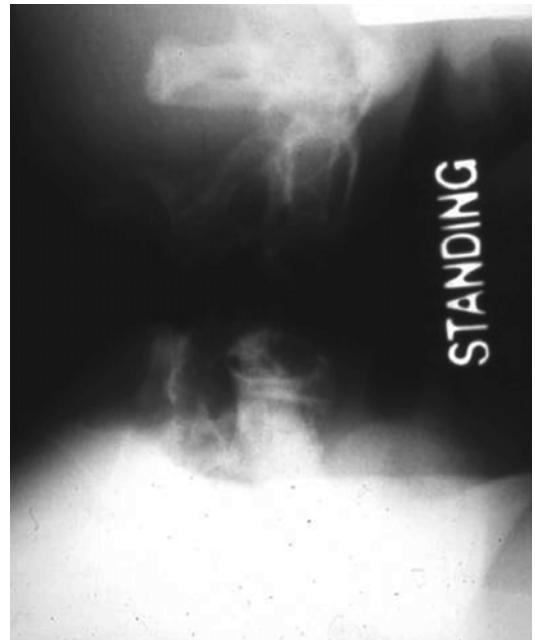

Fig. 30.8 A patient with known Klippel-Feil anomalies was undergoing HGT when she experienced a sudden increase in neck pain with facial numbness and peripheral dysesthesias. This separation of C2–3 occurred at the first non-fused segment cephalad to a long congenital fusion segment extending C3 to upper thoracic spine. HGT was immediately discontinued, with neurologic resolution

in more recent series and has never occurred at our institution. Pin sepsis can, however, produce subcutaneous cellulitis with alarming effect on peri-orbital structures, for example (Fig. 30.7).

Pin pain in the absence of pin tract infection suggests loosening, another relatively common occurrence, and mandates that the pin torque be checked by tightening the pin(s), under sedation if necessary. Cervical pain (axial) without radicular symptoms is also common, and probably indicates the limit of tolerable traction [15]. In older patients (>10 years), the pain associated with continual traction is a more frequent complaint, and while it may limit the amount of weight that can be eventually applied, it rarely produces symptoms which negate the efficacy of the method. Obviously any patient with a severe or unrelenting neck pain or a neurologic change in the cranial or upper extremity nerve function must be evaluated radiographically for cervical pathology (Fig. 30.8), which in our series has occurred but twice in >170 cases. Monthly surveillance by lateral cervical radiographs is recommended for any patient with known cervical abnormality (congenital fusion, previous surgery – see Fig. 30.9) or a hypotonic diagnosis. We have recently identified a XI cranial (spinal accessory) nerve lesion in a congenital myopathy patient which went undiagnosed during traction because of absence of neck pain in a situation where muscle weakness (shoulder shrug) could not be examined, but a radiograph reviewed after diagnosis demonstrated probable over-distraction in the cervical spine.

True neurological complications are rare and associated with rapid addition of traction force. Reversible cranial nerve lesions have been reported [15, 16], which respond to decreasing the amount of traction. Similarly, nausea and dizziness with nystagmus have been reported and are reversible with temporary traction relief. [4] On the other hand, motor paresis can occur rapidly at the onset of traction application, but is associated with pre-existing abnormalities of the cord or a stenotic spinal canal [1], and may not necessarily resolve with immediate discontinuance of traction. Thus *any* pre-existing cord (Fig. 30.5b) or canal abnormality producing local stenosis automatically constitutes a contraindication

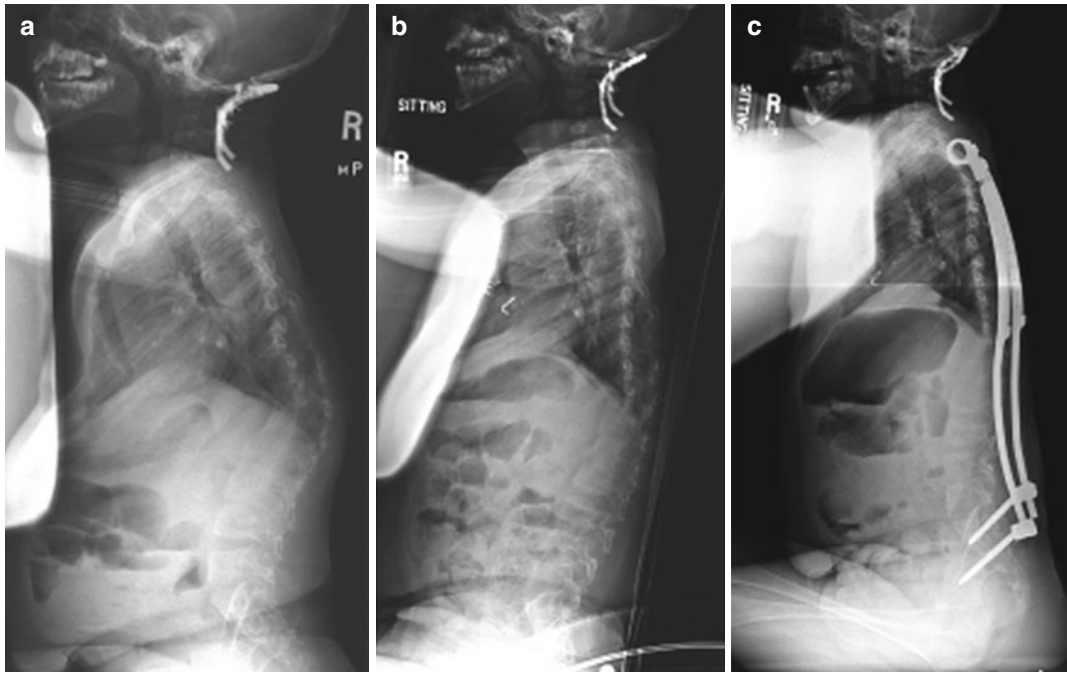

Fig. 30.9 (**a**) Paralytic kyphosis in a 3-year-old boy with cervical myelopathy due to basilar invagination associated with skeletal dysplasia. External bracing/casting was intolerable due to respiratory compromise in brace and gastro-esophageal reflux; (**b**) after 3 months in traction, kyphosis has improved enough to attempt expandable rib-pelvis devices. The patient also gained 5 kg in weight due to better nutrition; (**c**) rib-pelvis devices were implanted. Unfortunately due to continued weight gain and paralytic deformity, the upper cradles eventually required revisions

to HGT – indeed, *any* form of traction – especially if there is a pre-existing paraparesis (see Fig. 30.5a), which should be decompressed rather than elongated.

The *safety* of the method depends on the ability of the patient to auto-relieve the traction stretch by pushing up on the wheelchair arms or walker hand rails as necessary if uncomfortable and the "stretch" is excessive. Such safety is secured by using a spring-loaded traction appliance such as a common fish-scale or other device where spring tension provides the "weight," and shortening the spring decreases the weight amount (see Figs. 30.6b, c). Although classic orthopedic traction pulleys with weights are commonly used, these provide non-escapable traction, as the amount of weight is constant. Thus, the patient cannot auto-relieve distressing weight by pushing up on wheelchair arms (Fig. 30.10) or tip-toeing. If a series of pulleys in different directions are required, thus adding additional friction in the system, the patient may not be able to "bounce" in the traction, losing the dynamic feature, and again preventing auto-relief and safety. For this reason, spring-type devices are recommended whenever possible (see Figs. 30.6b, c).

30.6 Current Results

Radiographic improvement in spinal deformity is usually seen within 1 week of instituting traction, but as suggested earlier, longer duration of treatment is expected to produce more correction until a "plateau" is reached wherein no further improvement seems to occur. In all 30–40 % correction of coronal and sagittal plane Cobb deformity typically occur over periods of 2–21 weeks of HGT, whether for definitive correction and fusion [4–6] or as a preliminary treatment for use of VEPTR or growing rods [1, 2] (Figs. 30.9a, c). Equally important is the improvement in trunk

Fig. 30.10 Wheelchair traction using classic orthopedic pulleys and weights. Note the absence of arms on the wheelchair (*not* recommended). The patient will not be able to relieve traction tension by raising herself up in the seat, as the amount of weight is constant (as opposed to a spring device) and the absence of chair arms provides no point of effective push-off

length (5–6 cm average) and trunk shift [4–6] (Fig. 30.3). Literature review documents a 25 % increase in T1-12 length. A recent review of 50 patients showed that age (< or > 8 years) or previous treatment did not alter the efficacy of radiographic correction (Fig. 30.11) [6].

In addition to the radiographic improvement, other positive changes that are not easy to quantify have been observed. Although many patients do not have *objective* improvement in pulmonary function tests (PFTs) while in traction, there is often a sense of improved well-being and respiratory "reserve" during traction, which translates into postoperative ICU mechanical ventilation becoming unnecessary, even though it may have been predicted as part of the preoperative evaluation and one of the indications to proceed with traction. The surgical stabilization following traction also tends to be less technically difficult, since as much as 75–85 % of the eventual deformity correction has already been achieved by the time instrumented correction is attempted [1], and the instrumentation can be implanted with less contouring and much less stress on the anchors, especially if there is pre-existing kyphosis, which has been improved by the traction (Fig. 30.9). Finally, if there are any nutritional or metabolic issues preoperatively, which might jeopardize wound healing, these can easily be dealt with during the weeks in preparatory traction.

In extreme instances, we have treated selected patients in HGT *indefinitely* (Fig. 1). One current example involves a myopathy patient with collapsing deformity whose original surgical indication was to attempt improvement of respiratory function – he had been hospitalized repeatedly for pneumonia and lung abscess. More than 20 procedures (implantation, incision/drainage, explantation, re-implantation, etc) were performed trying to maintain rib-pelvis expandable devices, which eventually had to be abandoned due to recurrent dehiscence and sepsis. Now 4½ years after final explantation and institution of HGT, the patient has remained free of any infection, did not require any further hospitalization, and has gained 10 kg body weight, a 45 % increase from pre-HGT weight.

Conclusion

Halo-gravity traction is an invaluable treatment option for essentially any complex spine deformity patient, and in the EOS population becomes even more important as it treats both spinal as well as chest wall and nutritional issues simultaneously. Predictably HGT produces 30–35 % correction of both coronal and sagittal plane deformity *non-operatively*, while elongating the thoracic and lumbar spine, producing better

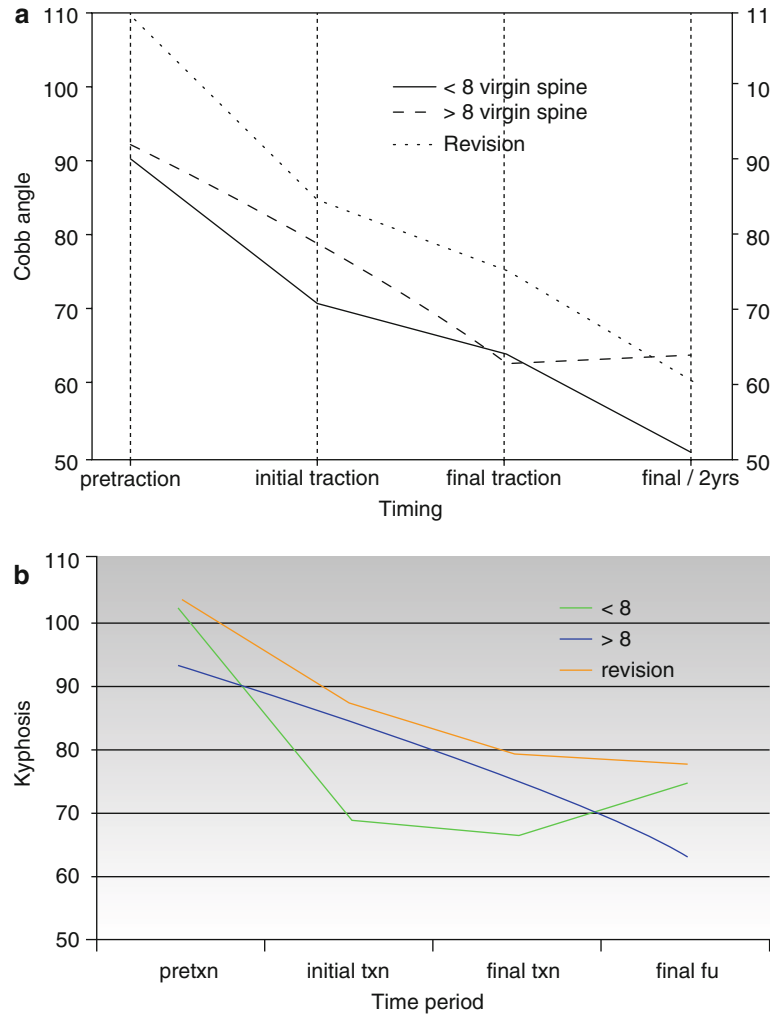

Fig. 30.11 (**a**) Response of coronal curve magnitudes to HGT and followup [6]. There is no difference in corrective outcomes between patients < age 8, > age 8, or undergoing traction after previous treatment; (**b**) response to HGT for sagittal plane correction. Patients < age 8 have the best kyphosis response to traction

respiratory function by improving volume and diaphragmatic function, and better nutrition by elongating the abdominal cavity. It can be used as a delaying tactic by producing deformity correction which then permits better continued non-operative management by brace or cast, or it can be used to produce enough deformity correction, especially kyphosis, to then facilitate efficacy of distraction-based growing constructs. Obviously it can also be utilized to gain deformity correction prior to definitive instrumentation and fusion, of significant advantage in the osteopenic, fragile neuromuscular or syndromic patient where simpler, faster surgical implantation is most attractive.

After 25 years of experience with the method, HGT has been noted to be exceptionally safe and free of complications, understanding that certain patients have definite contraindications to HGT – intradural lesions and stenosis at junctional areas. Further, it must be used with extreme caution and vigilance in patients with pre-existing paraparesis, hypotonic myopathies, and existing cervical abnormalities including congenital and post-surgical etiologies. In contrast to pin infections and loosening which are relatively common minor complications, the incidence of major complications – primarily neurologic – are less than 5 %, while the improvement in deformity, respiratory, and nutritional

morbidities is substantial, and leads to improved outcomes in the subsequent management of EOS patients.

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Part V

Management of Spinal Deformity in the Growing Child: Traditional Surgery

Hemivertebrectomy in Early-Onset Scoliosis

31

G. Bollini, P.L. Docquier, and Jean-Luc Jouve

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Key Points

- HV excision can be considered as soon as there is proof of evolutivity related to a free HV in a congenital scoliosis.
- In selected area such as the lumbosacral junction, HV responsible for trunk imbalance must be removed as soon as possible (ideally age 2–3).
- When performing HV excision by a single posterior approach use of "rod and screws" device is mandatory (associated "rod and hooks" device is a helpful tool to make the whole construct more effective).
- In complex deformities including HV or when the curvature induced by a single HV is already high, HV excision can be associated with other procedures such as growing rods or rib's distractor.
- For free lumbar HV, we still recommend HV excision by a combined anterior and posterior approach.

31.1 Background

The natural history of congenital scoliosis has been well documented. The degree of scoliosis produced by a hemivertebra (HV) depends on its type, its site, the number of HV and patient's age. When the HV is fully segmented or semi-segmented, progression of deformity is usually

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unavoidable. Depending on the HV location, evolution is different. Thoracolumbar and lumbosacral junctions represent transitional areas between the mobile lumbar spine and a less or nonmobile segment (thoracic spine or sacrum). The HV located in these two transitional areas results in a trunk shift. Moreover, in the thoracolumbar location, HV leads rapidly to torsional deformity so that an anatomical posterior convex compression can transform in a concave mechanical compression. In the thoracolumbar and lumbar location, sagittal component is important with risk of significant kyphosis. On the contrary, lumbosacral location does not typically produce a kyphotic deformity. Single, fully segmented HV located at the thoracolumbar junction can deteriorate at a rate of 2–3.5° per year, in the lumbar area (between L2 and L4) deterioration of 1.7° per year in cases of fully segmented HV and 1° in case of semi-segmented HV can be expected, and fully segmented HV located at the lumbosacral junction can deteriorate at a rate of 1.5° per year

For the surgical management of congenital scoliosis hemivertebral excision has a potential advantage over convex epiphysiodesis by addressing the deformity directly, thus allowing immediate correction of both the frontal and the sagittal planes.

Royle [40] in 1921 was the first to perform a hemivertebra resection.

Von Lackum reported on one case in 1924 and then five cases in 1933 [44]. He initially performed his surgery in one stage but later reported on a two-stage procedure consisting of an anterior vertebral body excision followed later by posterior excision and fusion. In 1932 Compere [17] performed a thoracolumbar hemivertebra excision followed by turnbuckle cast correction in two patients. Wiles [45] in 1951 reported good correction of scoliosis after thoracic hemivertebrectomies in two patients but severe kyphosis developed in both patients and paraplegia developed in one. Similar results were also reported by Winter [46].

Goldstein [20] in 1964 reported on one case of lumbosacral hemivertebra resection. Hodgson [24] in 1965 reported safe results using an anterior approach to remove hemivertebra. Carcassone et al. [13] in 1977 and Onimus and Michel [34] in 1978 reported on two cases each of hemivertebral resection by an anteroposterior approach in two stages to treat congenital scoliosis. In 1981 Bergoin et al. [3–5] reported in the French literature his technique to remove safely the hemivertebra using two separate approaches during the same operative session. In 2002 Ruf and Harms [39] reported HV resection by a posterior approach using a "screws and rods" construct. We have reported our series of HV resection, by a posterior and anterior approach, in a single operative procedure in different publications [6–10]. Banagan in 2007 [1] reported current concept on genetics and surgical treatment of congenital scoliosis.

31.2 Concept of Hemivertebra Resection

A hemivertebra is located laterally in the frontal plane (responsible for a scoliosis) and in the sagittal plane more or less posteriorly (responsible for a certain amount of kyphosis).

These two components, scoliosis and kyphosis, are important with growth in terms of major curve angles and moreover in terms of numbers of vertebrae included in the main curves (frontal and sagittal).

The reason it is mandatory, as soon as there is proof of progression, to perform HV excision before is that more than the two vertebrae adjacent to the HV become involved in the scoliotic process (i.e. torsional deformity), in our opinion before 3 years of age.

There are current controversies whether it is better to perform HV using a single posterior approach or an anterior and posterior approach in the same operative session.

There are also controversies concerning the use of hooks or screws together with a rod to stabilize the spine after such a resection.

31.2.1 Biological Concept

For a better understanding, we have to consider the HV resection in two parts.

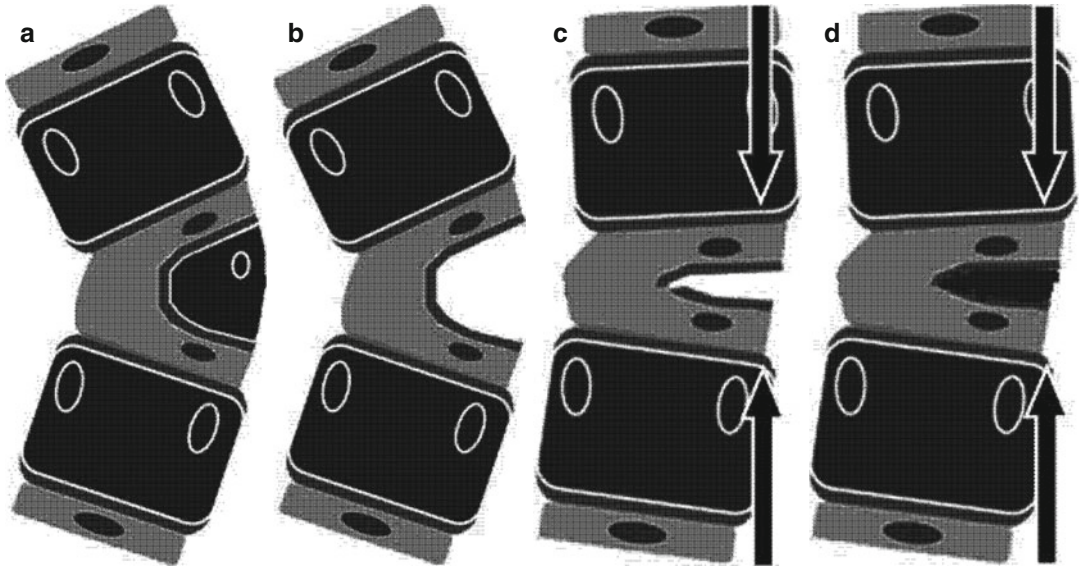


Fig. 31.1 *Black*: osseous part of the vertebrae. *Dark grey*: growing structures *Light grey*: disc structures. (**a**) The HV to be removed. (**b**) The surrounding growing structures of the HV remain after resection. (**c**) Compression force is applied. (**d**) The gap is spontaneously filled by new bone formation

31.2.1.1 First Part Is the Resection of the Posterior Aspects of the HV

Whatever the technique used this session is the same. Often the hemilamina is fused with either the lamina above or the lamina below. In such case, to determine which part of the lamina belongs to the hemivertebra, use fluoroscopy to determine the location of the pedicle of the hemivertebra. It is afterwards easy to answer that question.

Once the hemilamina belonging to the hemivertebra is identified, the resection starts removing the hemispinous process then the hemilamina.

The facets of the HV can be missing or poorly developed. When present, these facets must be removed, as well as the transverse process of the HV.

It is then relatively easy to circumscribe the pedicle of the hemivertebra.

This pedicle is then removed using a subperiosteal approach. The periosteum becomes the layer of tissue which protect, during this session, from the nerve roots above and below the pedicle.

Another option is, after removal of the transverse process in the thoracic area, to gain access to the lateral aspect of the pedicle and thus the vertebral body to remove the HV from its lateral to its medial aspect.

31.2.1.2 Second Part Is the Resection of the Anterior Aspects of the HV. Several Options Exist Regarding This Part

First Option: Removal of the Osseous Part of the HV (Fig. 31.1)

Whether the approach is single posterior or anterior and posterior and whether hooks or screws are used, correction is achieved through compression forces on the convexity. At the end of the procedure, the surrounding growth structure of the HV is still active.

The empty space will be filled with spontaneous new bone formation.

The positive consequence is the stability of the vertebral column in its anterior aspect, but a possible negative consequence is that there may be "recurrence," at least partial of the HV. That explains the observed "new bone mass at the site of the HV" as reported by Ruff and Harms [38].

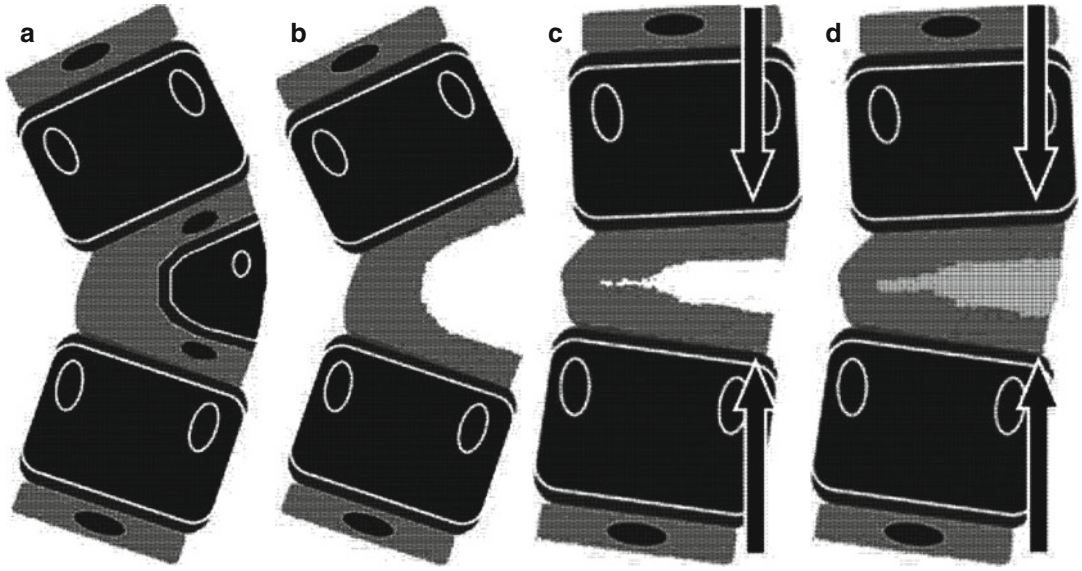


Fig. 31.2 *Black*: osseous part of the vertebrae. *Dark grey*: growing structures. *Light grey*: disc structures. (**a**) The HV to be removed. (**b**) The surrounding growing structures of the HV are removed. (**c**) Compression force is applied. (**d**) The gap is spontaneously filled by a fibrous scar (Courtesy of Gerard Bollini)

This option is the one used in the so called "eggshell procedure" and is usually performed by a posterior-only approach [32].

Second Option Is to Remove Both the Osseous Hemivertebral Body and the Surrounding Growth Structures of the HV (Fig. 31.2)

Whether the approach is single posterior or anterior and posterior and whether hooks or screws are used again, correction is achieved through compression forces on the convexity. At the end of the procedure, the growth structure of the adjacent vertebrae is still active.

The negative consequence of such a procedure is that the gap created anteriorly is filled with a fibrous scar whose mechanical stability is doubtful.

The relative positive consequence is that growth may occur between the resting endplates of the vertebrae adjacent to the HV.

The term "relative" is used because such a growth is correlated with distraction forces on the anterior aspect of the spinal column which could turn on posterior hardware breakage or dislodgement.

It is impossible to get an anterior convex arthrodesis between the two vertebrae adjacent to the HV and in the meantime to observe further growth coming from these same two vertebrae as stated by Ruf (Fig 31.3).

Third Option Is to Remove Both the Whole HV (Osseous Hemivertebral Body + Surrounding Growth Structures) and the Endplates of the Adjacent Vertebrae on the Convex Side of the Scoliosis (Fig. 31.4)

It is the only option which allows a true convex epiphysiodesis under the condition that bone grafting is performed between the two adjacent vertebral bodies on its convex aspect.

From these three options, whatever the option chosen, the main point to understand is that at the end of such procedures and after having applied compression forces on the posterior aspect of the vertebral column, there is still a gap between the two adjacent vertebral bodies.

The gap can be filled with a bone graft (Option 3) if an anterior and a posterior approach are performed in the same session and in such case hooks or screws can be indifferently used.

But in a posterior-only approach, screws rather than hooks are used to avoid a secondary

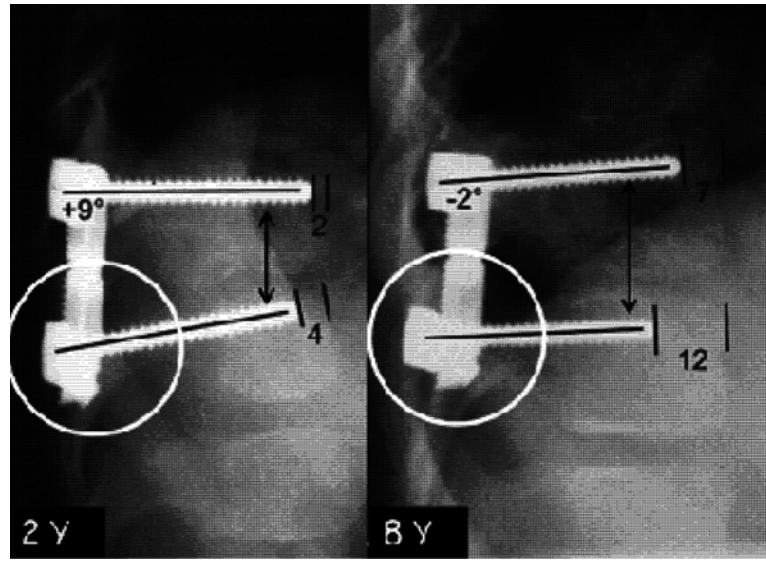


Fig. 31.3 Postoperative radiograph after HV resection, at 2 years of age on the left and at 6 years follow-up on the right. Although an arthrodesis has been performed posteriorly, there is still growth from the two adjacent vertebral bodies anteriorly (note the dislodgement of the bolt of the lower screw in the *white circles*) (Reprinted from Ruf and Harms [40]. With permission from Wolters Kluwer Health)

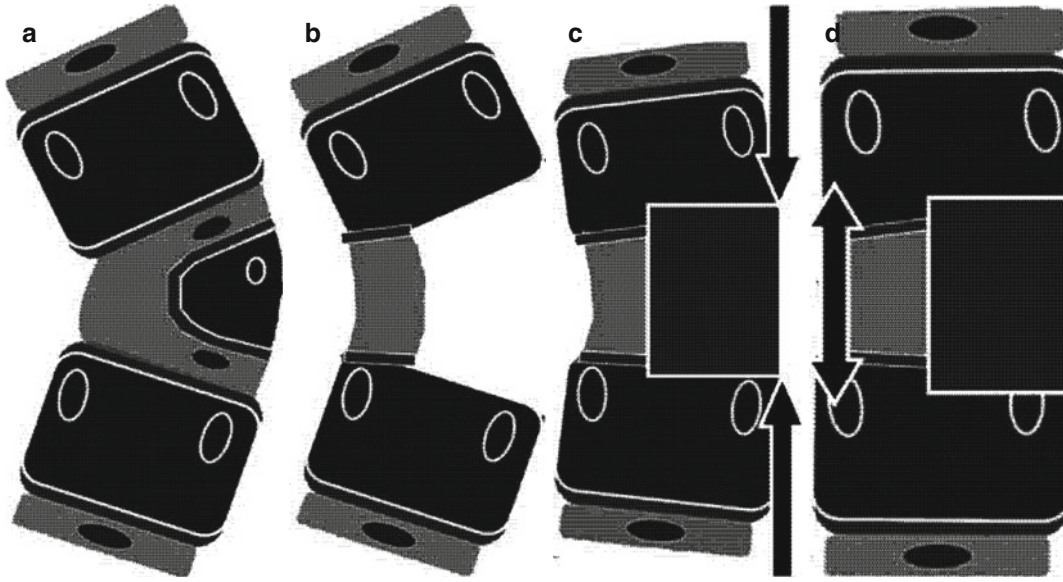


Fig. 31.4 *Black*: osseous part of the vertebrae. *Dark grey*: growing structures. *Light grey*: disc structures. (**a**) The HV to be removed. (**b**) The surrounding growing structures of the HV are removed as well as the disc and the convex growing structures of the adjacent vertebrae. (**c**) Compression force is applied. (**d**) The gap is filled by a bone graft (Courtesy of Gerard Bollini)

kyphotic deformity and/or a pseudoarthrosis. Hooks and rods do not provide significant enough stabilization as shown in Figs. 31.5 and 31.6, while screws and rods provide both a rigid construct and a "Blount staple effect", avoiding further growth as shown in Figs. 31.7 and 31.8.

The Fourth Option Is to Remove Everything Between the Two Vertebral Bodies of the Vertebrae Adjacent to the HV (Fig. 31.9)

This option allows to almost complete deformity correction and total arthrodesis between the two adjacent vertebrae.

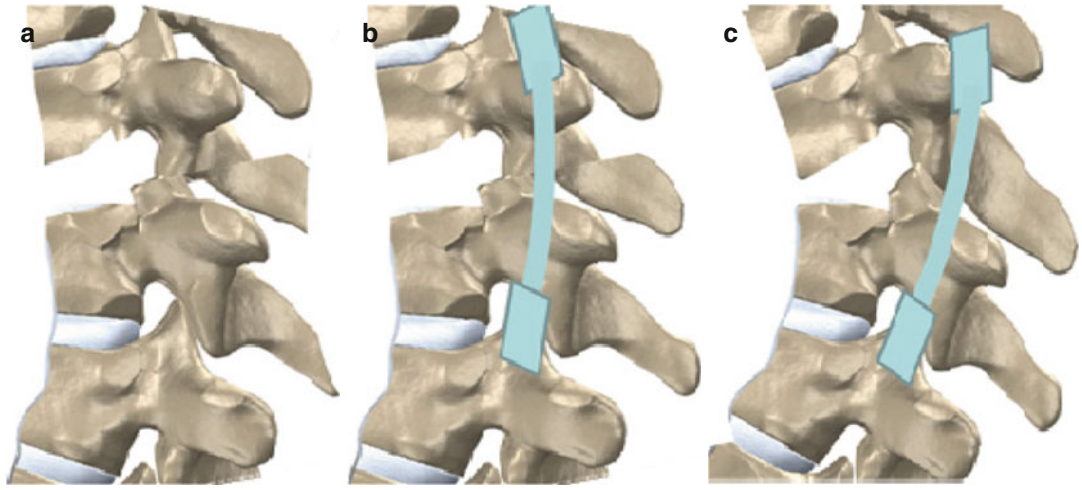


Fig. 31.5 (**a**) After removal of the HV. (**b**) Posterior compression with hooks and rods. (**c**) Kyphosis development (Courtesy of Gerard Bollini)

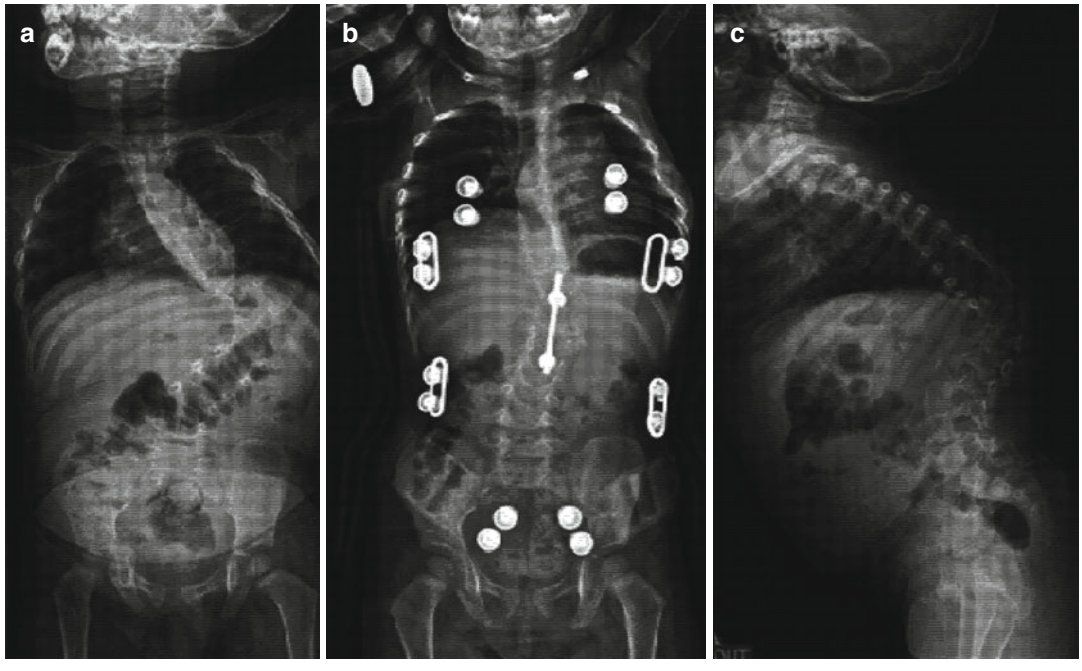
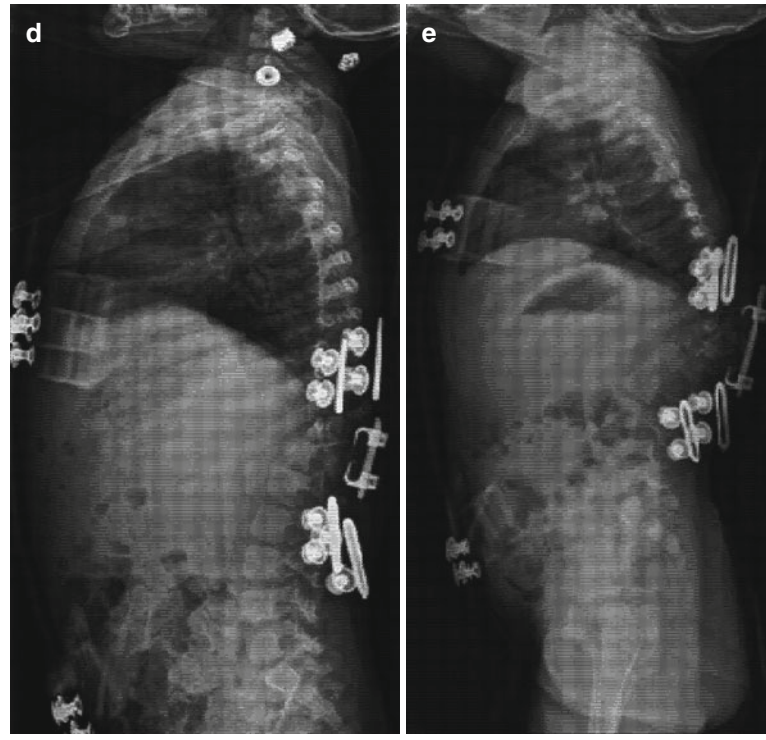


Fig. 31.6 (**a**) Thoracolumbar HV pre-op coronal. (**b**) Post-op coronal. (**c**) Thoracolumbar HV pre-op sagittal. (**d**) Post-op sagittal. (**e**) At follow-up sagittal with a kyphosis development

Fig. 31.6 (continued)



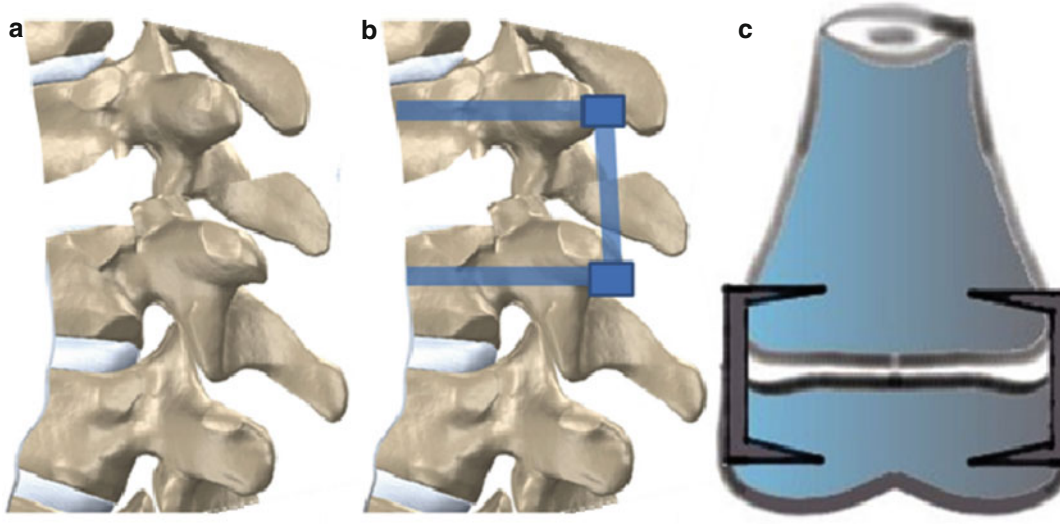


Fig. 31.7 (**a**) After removal of the HV. (**b**) Posterior compression with screws and rods construct. (**c**) The screws and rods construct acts like Blount staples avoiding further growth from the anterior aspect and providing stability thus avoiding kyphotic evolution (Courtesy of Gerard Bollini)

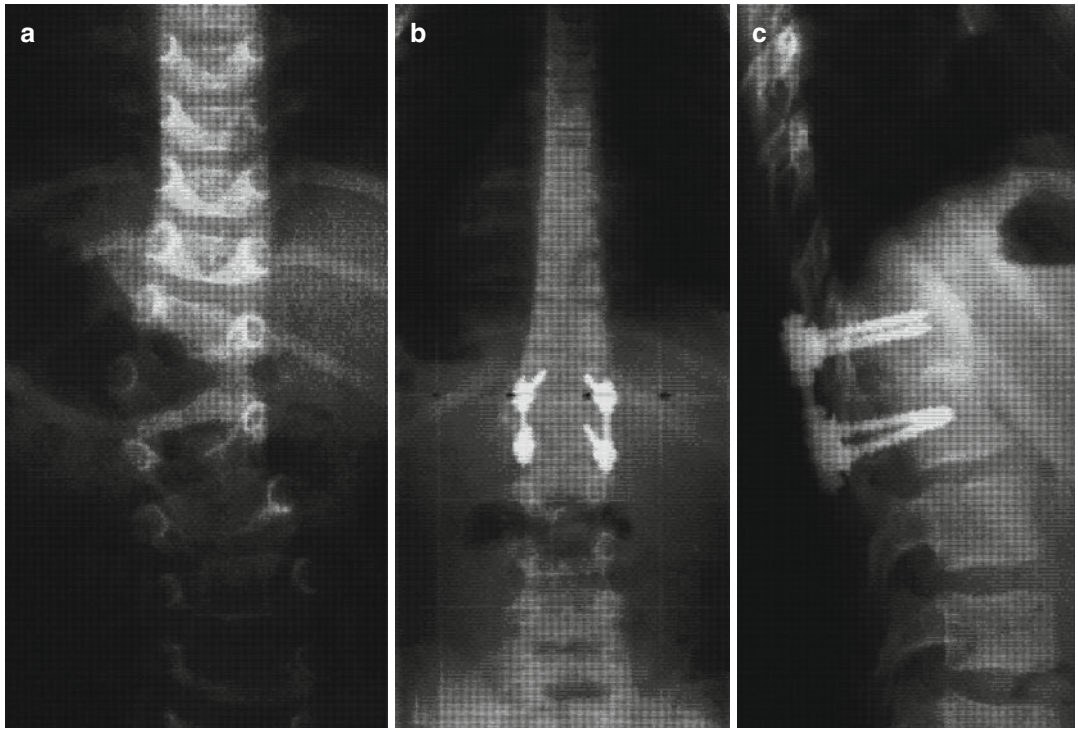


Fig. 31.8 (**a**) TL HV pre-op. (**b**) Post-op frontal view. (**c**) Post-op sagittal view

This fourth option is chosen for the older child with a markedly associated kyphotic deformity.

31.2.2 Stability Concept

Removal of a hemivertebra leaves an empty space posteriorly and anteriorly. To close the gap posteriorly and to ensure immediate posterior stability, compression instrumentation posteriorly is necessary.

Closing the posterior gap freed by the HV resection leaves an open anterior gap (otherwise you may observe a residual kyphotic deformity). This anterior gap decreases about 50 % after posterior compression. Therefore to obtain an immediate anterior stability it is necessary to use either a "screws and rods construct" or a "hooks and rods construct" associated with an anterior graft. In both cases posterior arthrodesis is mandatory.

31.2.3 Unexpected Issues

The closure of the anterior gap using a "screws and rods construct" can be tricky, in the young child, due to the small diameter of the pedicles and the fact that both the pedicle and the vertebral body are mainly cancellous bone. Loosening of the screws may occur because of the compression.

In such case we can use a "screws and rods construct" associated with a "hooks and rods construct", the later protecting the former of loosening (Fig. 31.10) [23].

31.3 Technique

31.3.1 Preoperative Evaluation

Radiologic imaging includes standing postero-anterior (PA) and lateral view of the full spine. Magnetic resonance imaging of the spine to

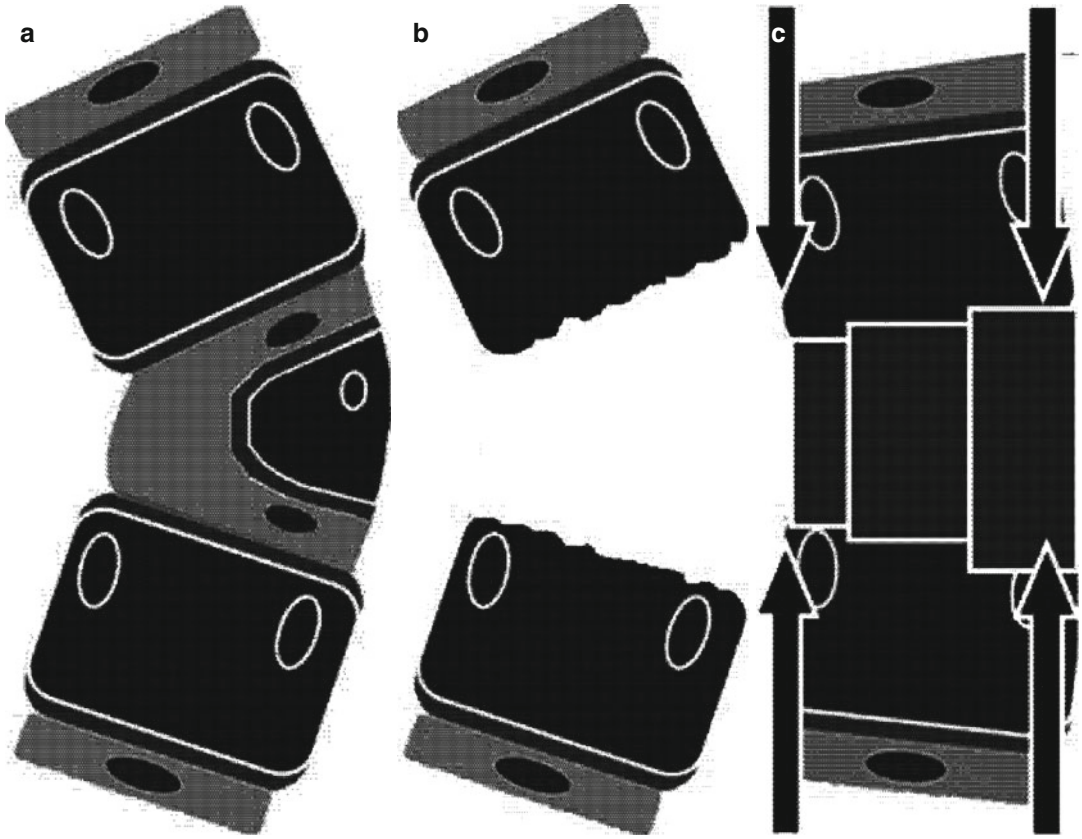


Fig 31.9 *Black*: osseous part of the vertebra. *Dark grey*: growing structures. *Light grey*: disc structures. (**a**) The HV to be removed. (**b**) The whole space between the two adjacent vertebral bodies is freed from all the structures. (**c**) Compression force is applied and the gap is filled with bone grafts (Courtesy of Gerard Bollini)

assess the neuroanatomy is performed to exclude intraspinal abnormalities and to study the segmentation of the HV and the growth plate. Renal ultrasound has to be done preoperatively to assess associated congenital renal system abnormalities. If the deformity looks complex on the radiographs, additional CT scan, with both 2D and 3D reconstruction, can be helpful.

31.3.2 Single Posterior Approach

The patient is positioned prone with the operative area freed for fluoroscopy. Neuromonitoring of both SSEP and MEP or NMEP is mandatory.

The pedicle of the HV is identified under fluoroscopy, the skin is marked, and a vertical midline incision is performed. The following has already been presented in Sect. 31.2.1.1.

31.3.3 Posterior and Anterior Approach

Posterior and anterior approach can be performed in the same anaesthetic session but require two different positions of the patient to make this surgery as safe as possible (a lateral position is not the best one to perform the resection of the posterior elements of the HV). For the posterior approach, the patient is in a prone position with the operative area freed for fluoroscopy. The procedure following the posterior approach has been described above (see Sect. 31.2.1.1). The further

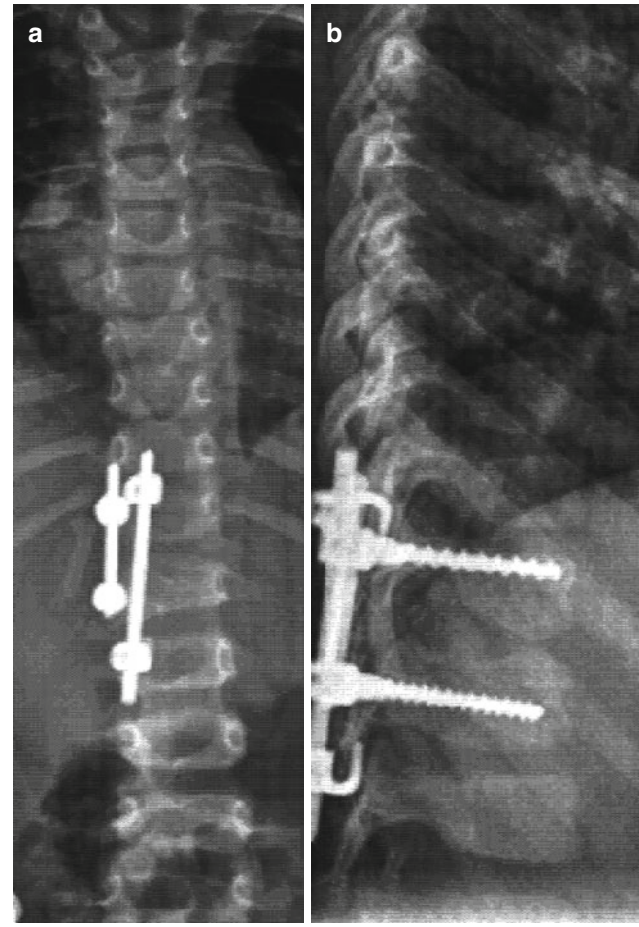


Fig. 31.10 (**a**) Frontal view of a hook and rod construct associated to a screw and rod construct. (**b**) Same patient. Lateral view

insertion sites for laminar hooks and/or pedicle screws in the adjacent proximal and distal vertebrae are prepared.

At the completion of the posterior procedure, the skin is temporarily closed with sutures and the patient is turned in a lateral position for a convex anterior approach.

The anterior approach depends upon the level of the hemivertebra, transthoracic for thoracic and thoracolumbar HV and a retroperitoneal approach for the lumbar or lumbosacral HV.

When the anterior spine is exposed, the posterior approach is reopened. The body of the HV lies very laterally and is easily found. Resection of the whole vertebral body plus the surrounding growth structures is performed as well as resection of the disc and growing structures of the HV adjacent vertebral bodies. In the young child, resection can be stopped at the midline of the vertebral column, avoiding any damage of the concave side if any further growth from the concave side is expected, allowing additional correction of the deformity.

Then, the hardware is inserted through the posterior approach. In front, the bone grafting needs to have a rigid structure to add a "pillar", thus avoiding any collapsing of the HV adjacent vertebrae.

Autogenous fibular bone graft harvesting is performed and the fibular graft is introduced in between the two HV adjacent vertebrae during the posterior compression in an order to control the kyphotic component of the deformity. Homologous fibular grafts can also be used. The two approaches are then closed. Postoperatively the patient has to wear a previously moulded brace for 3–6 months.

An alternative is to use vertebral body screws after this combined approach (Fig. 31.11).

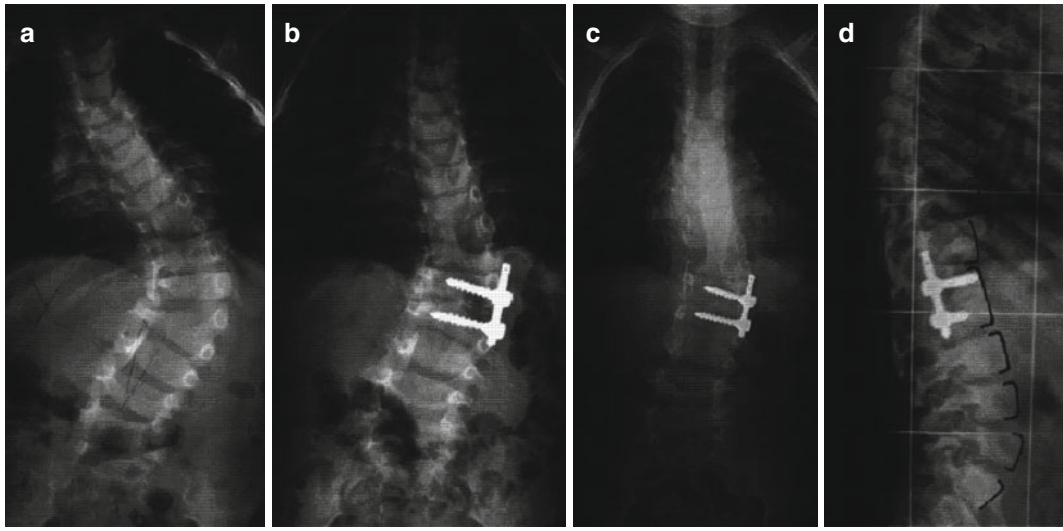


Fig. 31.11 (**a**) Thoracolumbar HV at 1 year 10 months of age. (**b**) Post-op. (**c**) Frontal view at 7 years follow-up. (**d**) Sagittal view at 7 years follow-up. Additional growth from the concave side improves the final result

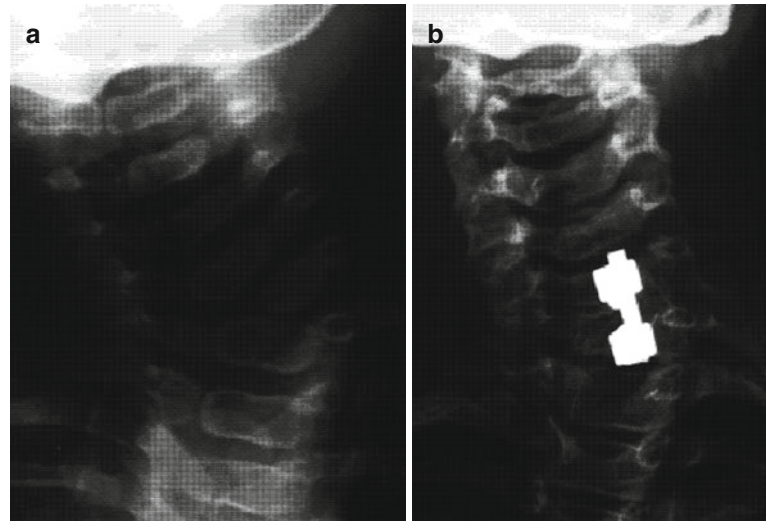


Fig. 31.12 (**a**) Free HV at the cervicothoracic junction in a 2-year-old girl. (**b**) Result at the end of the growth after resection at 2 years of age

31.3.4 Indication

The indication varies according to the location of the HV.

At the cervicothoracic junction, there are minimal indications for HV excision. Usually the HV is hemi-fused or totally fused with the adjacent vertebrae, producing mild deformity. The only indication is a free HV with severe deformity in a very young as shown in Fig. 31.12.

In the thoracic and the lumbar area, the indication depends upon the type of HV, the associated malformation and the observed evolution (Fig. 31.13).

In case of HV with associated malformation, CT scan provides information concerning the bone deformity which must be combined with the information provided by MRI which allows anticipating the evolution looking at the growing structures (Figs. 31.14 and 31.15).

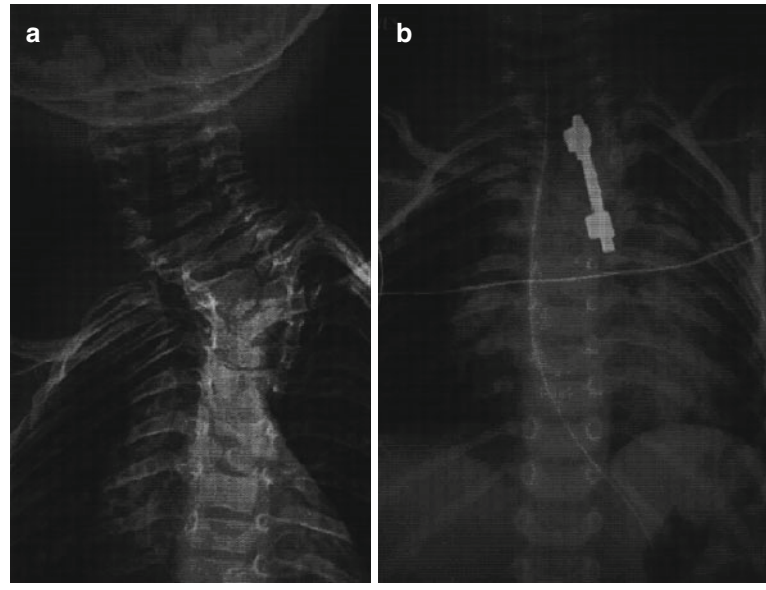


Fig. 31.13 (**a**) Free left T3 HV in a 3 years old boy. (**b**) Result after resection at 3 years of age

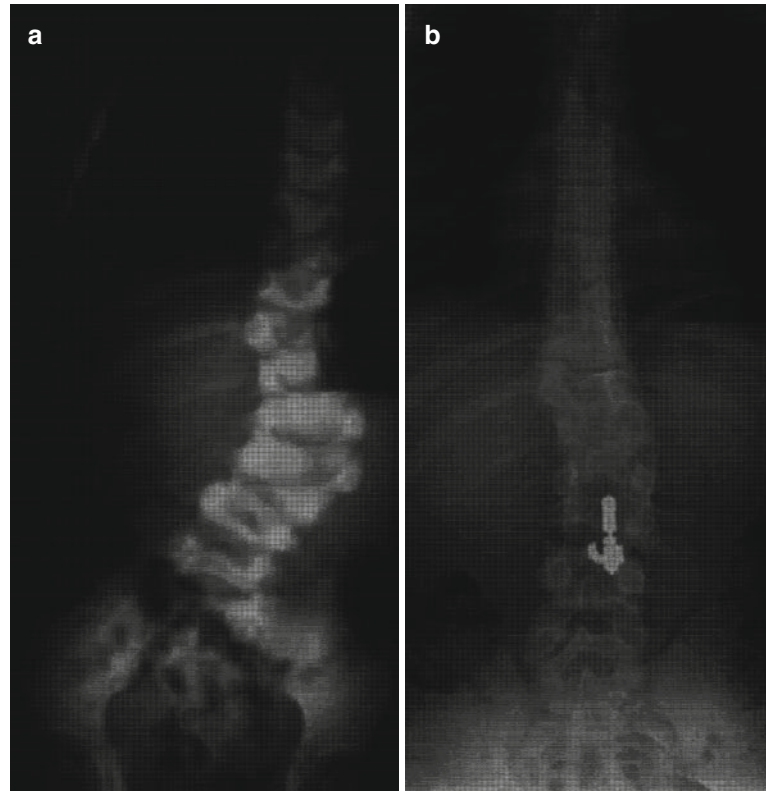


Fig. 31.14 (**a**) Complex lumbar malformations. Through MRI information a single HV excision was performed. (**b**) Result at the end of the growth

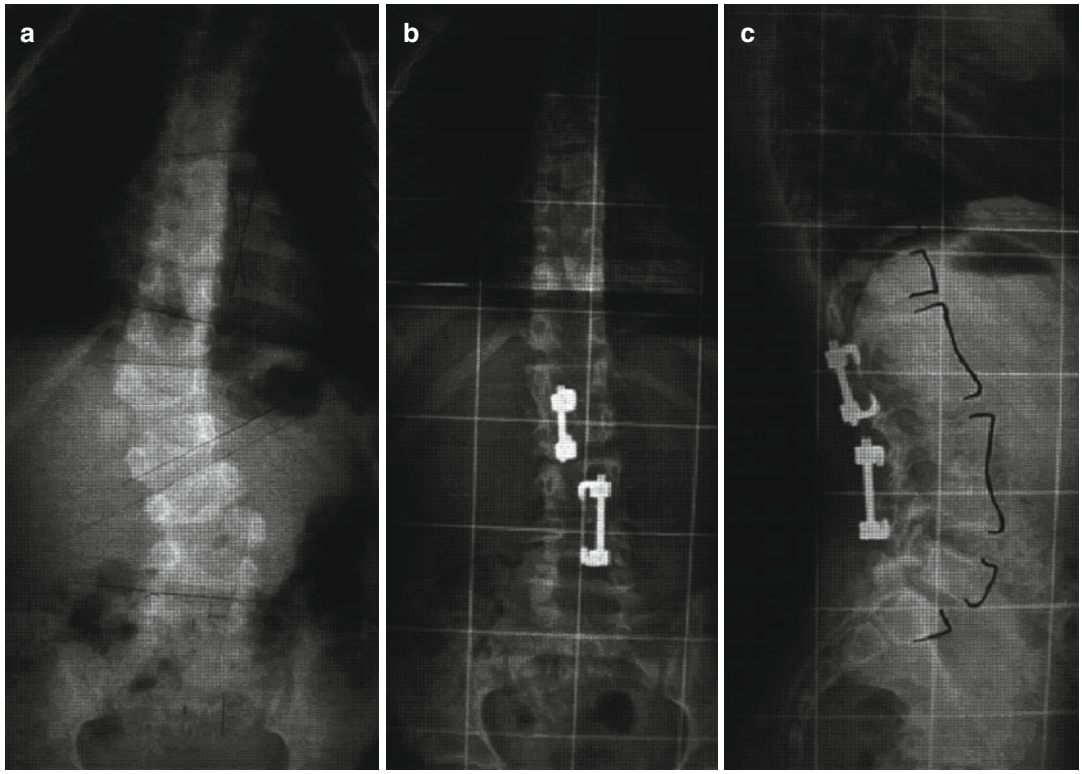


Fig. 31.15 (**a**) Coronal view of two contralateral HV operated on at 2 years of age. (**b**) Coronal view and (**c**) sagittal view at the end of the growth (Courtesy of Gerard Bollini)

At the lumbosacral junction, as soon as there is a trunk imbalance, HV excision is mandatory.

31.4 Discussion

Numerous authors have reported series of HV resection using successive or simultaneous anterior and posterior approaches [11, 12, 18, 22, 25, 27–31, 41, 43], posterior approach alone [33, 36–39, 42] or anterior approach alone (18 for two cases, [21]).

A multicentre retrospective comparison of three surgical techniques, i.e. hemiepiphysiodesis or in situ fusion (group 1), instrumented fusion without hemivertebra excision (group 2) and instrumented hemivertebra excision (group 3), showed that while group 3 had a higher complication rate than either group 1 or group 2, posterior hemivertebra resection in younger patients resulted in better percent correction than the two others techniques [47].

Among the complications, there were six neurologic complications (one in group 2 and five in group 3). No one resulted in definitive neurologic impairment.

Controversies still exist concerning the safety of pedicle screws in young child [2, 14–16, 19, 35]. Pedicle screw instrumentation performed before the age of 5 years does not cause a significant negative effect on the growth of pedicles, the transverse plane of the vertebral body or the size of the spinal canal.

For instrumentation at the lumbosacral junction, screws can be used or other devices such as the one described by Hosalkar et al. [26].

This chapter was dedicated to HV resection and the presentation was done dealing with a single evolutive HV in a growing child.

But HV can be part of a congenital scoliosis with associated malformation such as multiple

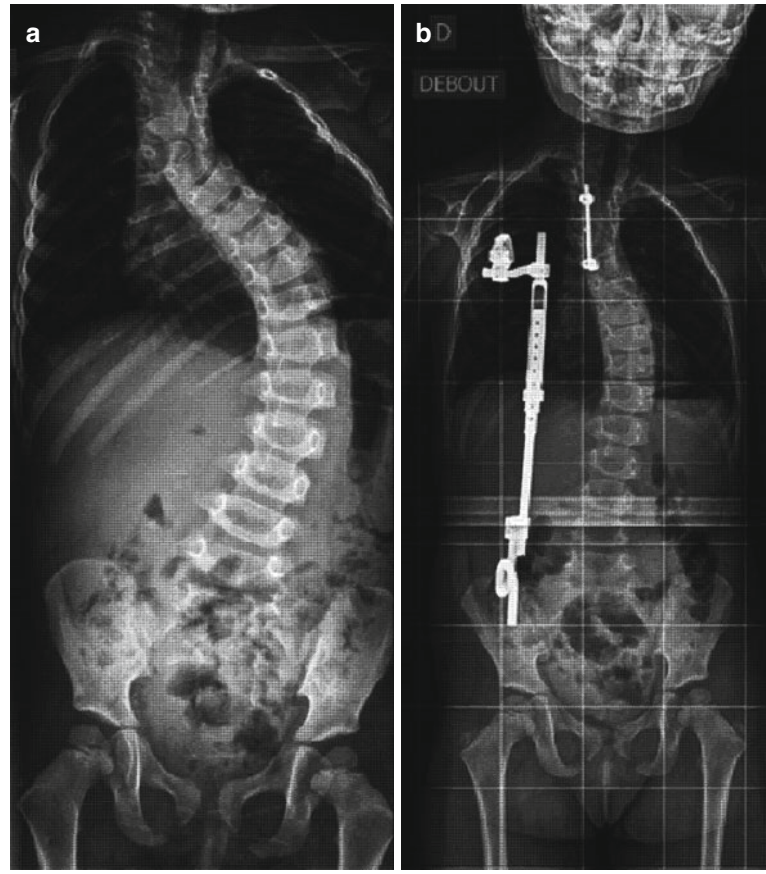


Fig. 31.16 (**a**) Frontal view of a congenital scoliosis. There is a right T5 HV and a long structural thoracolumbar compensatory curve. (**b**) The HV has been resected and a VEPTR inserted to address the compensatory curve

HV, contralateral bar, puzzle spine or rib's agenesis and/or fusion.

In such case HV resection can be part of a more extended surgical programme using new tools devoted for early-onset scoliosis patients as shown in Fig. 31.16.

HV can be also the single reason for a severe congenital scoliosis in an adolescent. In such case this chapter is no more adequate since the treatment must rather be a vertebral column resection (VCR) which is not the subject of our presentation.

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Vertebral Osteotomy

32

Tamás Fülöp Fekete, Daniel Haschtmann, and Dezső Jeszenszky

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Key Points

- Correction of spinal deformities in childhood is recommended as soon and as completely as possible to prevent the development of secondary structural changes and to ensure balanced spinal growth.
- Structural changes at the apex of the curve render it rigid. To allow for correction of the deformity in a harmonious way, apical soft tissue release with or without an osteotomy can be performed.
- Skeletally immature patients differ from adults in terms of the relative size of their vertebral body, spinal canal diameters, and disk height and also in terms of the elasticity of their connective tissue. This has a notable impact on the planning of osteotomies.
- Apical soft tissue release may include resection of the interspinous ligaments, ligamenta flava, joint capsule, and posterior and/or anterior longitudinal ligament.
- The Smith-Petersen osteotomy (SPO) or Ponte osteotomy can achieve more correction in children than adults due to the different vertebral and segmental size proportions and greater elasticity of the spine (Figs. 32.1 and 32.2).

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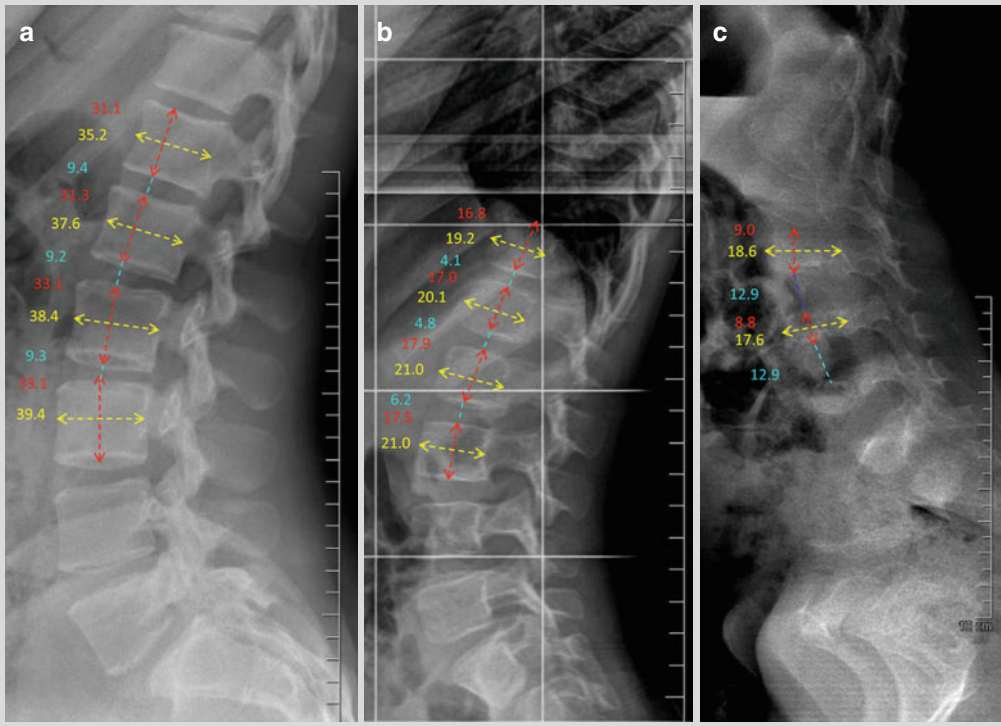

Fig. 32.1 This figure shows the differences visible on lateral radiographs in the size proportions within the lumbar spine in a 20-year-old adult (**a**), in a growing spine at the age of 5 (**b**) and in a 4-year-old child with spondyloepiphyseal dysplasia (**c**). The most important parameters to measure when selecting the appropriate type of posterior osteotomy are the vertebral body height (*red*), the sagittal diameter of the vertebral body (*yellow*), and the thickness/height of the intervertebral disk (*blue*). Note that in a syndromic spine deformity, substantial variations of size dimensions are possible which necessitates individual planning by measuring above parameters at the site of the osteotomy

- The pedicle subtraction osteotomy (PSO) can achieve in adults approximately 30° of correction in the sagittal plane at each spinal level at which it is performed. It is, however, rarely necessary in skeletally immature patients.
- Any posterior osteotomy, be it SPO, PSO, or posterior vertebral column resection (PVCR), can also be performed in an asymmetrical way, to allow coronal plane correction.
- A spinal deformity is always associated with a shortening of the spine. Conventional osteotomy techniques (some of the SPOs, PSO, VCR, hemivertebra resection) are coupled with resection and compression and thereby further shorten the spine.
- In the growing spine, distraction seems to be a more logical approach to increase (normalize) the length of the spine, although intuitively it is expected to be associated with higher risks. Nonetheless, for congenital deformities with a concave side bar, an opening wedge osteotomy can be performed. It is not necessarily coupled with fusion of the osteotomized region.
- Neurological injuries are the most feared complications following these procedures. There is an increased risk of neurological complications, compared with correction without any type of release or osteotomy. Therefore, multimodal intraoperative neuromonitoring is mandatory.

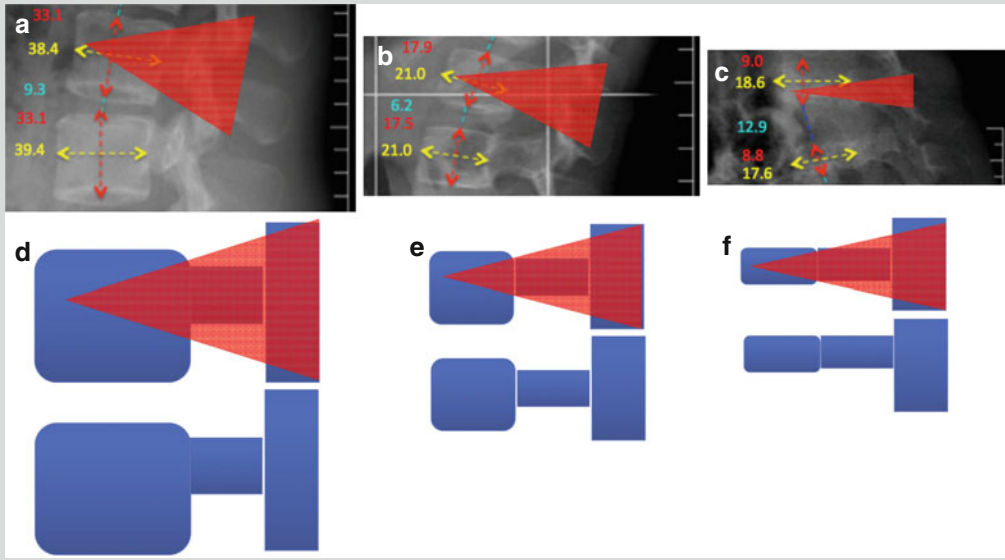

Fig. 32.2 The figure indicates the same parameters as in Fig. 32.1 as an example as measured in mm at the level of L2. The radiographs with illustrations (**a**–**c**) and illustrations (**d**–**f**) indicate what correction could be achieved by applying a PSO as indicated by the *red triangle*. Note that the spinal canal size in adults is almost reached at the age of 5 years, which means that the proportion in size of the anteroposterior diameter of the vertebral body and that of the spinal canal is smaller in children (**d** vs. **e** and **f**). These abovementioned size relations explain why less correction can be achieved when performing a PSO in children or patients with skeletal dysplasia patients compared to adults. Furthermore, a PSO in a child (**b**, **e** and **c**, **f**) would mean almost a vertebral resection. The consequences are twofold: the less competent anterior column (high disc space, low vertebral body) would then be even further weakened and one would loss an important anchoring point by removing the pedicles (no pedicle screws are possible at that level). Analyzing the size proportions becomes evident that a simple posterior SP-type osteotomy might be powerful enough to achieve sufficient correction in the growing spine

- The growth potential of the child has to be taken into consideration. Fusion should be avoided (or kept as short as possible, e.g., at the osteotomy site) in younger, skeletally immature patients in whom growing rods are considered to be a viable option for further guidance of spinal growth.

32.1 Introduction

The primary goal of spinal deformity surgery is to prevent further progression of the deformity and to improve three-dimensional balance. Generally, spinal osteotomy should be considered for deformities where instrumentation alone is unlikely to address the deformity adequately. A common assertion of pediatricians is that children are *not* small adults. This also holds true in spine surgical care and especially in the surgical management of severe spinal deformities of the growing spine.

There are many studies concerning osteotomies in adults and adolescents. Osteotomies are regarded as powerful tools but are also associated with increased risk of complications, especially neurological injury, as well as increased blood loss and operating time [1–3]. To the best of our knowledge, there are only few studies describing the use of osteotomies for spinal deformity in the pediatric patient population. Most of these simply describe the methods applied in adults and mention only incidentally—if at all—any differences between pediatric and adult populations. Only a few studies focus on skeletally immature

patients [4–6]. It is important to realize that different surgical principles apply when dealing with the growing spine as compared with the adolescent or adult spine. In this chapter we focus on these differences and attempt to highlight the specific factors that should be addressed when dealing with early-onset deformities in young and very young patients. We will limit our discussion to posterior-based osteotomies of the thoracic and lumbar spine.

32.2 Posterior Osteotomies: General Considerations and Indications

With the development of powerful segmental pedicle screw constructs that can now also be used in the pediatric patient population [7], posterior osteotomies are becoming more and more popular. Further, improved anesthetic techniques, including the use of antifibrinolytic agents, and advances in intraoperative neuromonitoring compensate for the increased blood loss, longer operating time, and potentially increased risk of neurological injury associated with posterior-only osteotomies. As a result, anterior release or combined anterior-posterior approaches have become much less popular. This is a major advantage for young children, since the negative effects of thoracotomy or other anterior approaches can be avoided. Anterior procedures, however, remain a useful tool in the armamentarium of the surgeon in some very difficult and complex deformity cases.

In order to achieve optimal correction over the shortest possible section, in a harmonious way, it is necessary to have similar elasticity over the region of the spine to be instrumented. Spinal segments with lower elasticity (stiff segments/curves) need more force to correct, which can exceed the maximum tolerable force that the instruments (pedicle screws) can sustain on that segment. To prevent plowing/pull-out of the screws, surgical steps are necessary to achieve more segmental elasticity. These steps are commonly recognized as osteotomy. Some of the osteotomies simply "normalize" the segmental elasticity (e.g., Smith-Petersen osteotomy, SPO), while others (e.g., vertebral column resection, VCR) destabilize the spine completely. These differences must be taken account when performing the correction. The greater the destabilization, the higher the risk of neurologic injury and blood loss.

In general, posterior osteotomies are recommended if the curve is large (or severe) and stiff, with or without fixed imbalance. It only makes sense to talk about balance of the spine if the patient is already ambulating (around age of 2 years). As a general rule, posterior osteotomies are useful when the curve does not reduce by at least half of its magnitude on side bending, lateral bolster, or traction films.

Osteotomies and soft tissue releases fall on a continuum ranging from the release of ligaments only to resection of multiple segments of the spine (vertebral column resection, VCR). Progressive segmental mobilization along this continuum is associated with increasing invasiveness and thus increasing risk.

In less rigid and smaller curves, apical soft tissue release alone might be sufficient to restore flexibility. This soft tissue release also serves as a basis for most osteotomies. Cutting and resecting the ligamentous structures that inhibit correction is the first step. Depending on the pathoanatomical situation, this may consist of resection of the interspinous ligament, the ligamentum flavum, and the facet joint capsule. If the correction-limiting rigidity lies around the intervertebral space, soft tissue release involves cutting the posterior (PLL) or anterior longitudinal ligament (ALL) and the lateral portion of the annulus fibrosus (especially on the concave side) through a posterior approach. If for any reason the ALL cannot be released from the back, an anterior approach can be considered. These maneuvers might suffice to provide adequate interbody elasticity; if soft tissue release is not enough, it is sometimes necessary to also resect the inferior articular process.

More extensive osteotomies include the Smith-Petersen or Ponte type osteotomies. An even more extensive procedure is the pedicle subtraction osteotomy (PSO), in which (compared with SPO) a much more extensive resection of the posterior

elements is carried out and a wedge-shaped resection of the vertebral body along with the pedicles is performed. PSO might be indicated in adolescent patients. However, for various reasons, it is rarely if ever used in patients who are skeletally less mature: probably the most important reason is that in skeletally immature patients, the ratio of the height of the elastic intervertebral elements to the height of the osseous vertebral body is greater. This means that a partial body resection along with the pedicles offers no/little better possibility for correction than a simple posterior soft tissue release or (partial) facet joint resection. However, the risks are higher, and the risk/benefit ratio drops substantially.

The most extensive of the spinal osteotomies is the posterior vertebral column resection (P-VCR) or combined vertebral column resection (C-VCR). For details regarding VCR, see Chap. 34. VCR is reserved for severe three-dimensional spinal deformities with significant sagittal imbalance. The progression from soft tissue releases to VCR must be considered in the context of increasing risk of complications, such as permanent neurological injury, increased bleeding, and increased likelihood of infection [8, 9]. Detailed preoperative planning with the anesthesiologist, pediatrician, neurologist, cardiologist, etc., is mandatory, as is preoperative discussion with the parents regarding the risks and benefits of osteotomies.

A comprehensive classification of osteotomies (SRS-Schwab classification of osteotomies) has recently been proposed [10], which to some extent can also be applied to the pediatric patient population. The aforementioned osteotomies are graded from 1 to 6, indicating increasing invasiveness. The differences in the characteristics of adult versus pediatric osteotomies are manifold. First, the types of deformity requiring surgery in young children are different from those in skeletally more mature (adolescent and adult) patients. Many of the deformities in young children are related to congenital abnormalities or are syndrome associated, which warrants a much more individual assessment and planning. In contrast, adolescents have mostly idiopathic deformities and, adults, a mixture of idiopathic and degenerative, with an increasing dominance of degenerative etiologies in elderly patients. Second, the remaining growth potential of the spine significantly influences decision-making when weighing up the risk of potential complications against the long-term benefit of more physiological development of the spine. Third, anatomical differences in the spinal structures (e.g., relative size of the vertebral body to the intervertebral disc height, spinal canal diameter, etc.) compared with adults influence the amount of correction achievable with resection of a given area of the spine (D. Jeszenszky, 2014 et al., unpublished data) (Table 32.1 and Fig. 32.1). Fourth, the area of the osteotomy is at risk of fusion or at least altered growth, and this risk is not necessarily related to the grade of osteotomy, as defined by the adult thoracolumbar osteotomy classification system of Schwab et al. but is related to the bony surface being exposed to perform the osteotomy.

Table 32.1 Important dimension relations to consider when planning posterior osteotomies in the growing spine

Vertebral body
Sagittal diameter
Height
Coronal diameter (by scoliosis correction)
Intervertebral disk height (thickness)
Spinal canal diameter

Osteotomies, although associated with a high risk, are very effective in providing correction. In our opinion, vertebral osteotomy can be even more effective in the growing spine than in adolescents and adults. It not only allows for rapid correction of the deformity but also guides further development. As such, the increased risk associated with surgery might be outweighed by the advantage of normalized development and growth of the rest of the spine, resulting from the rapid and extensive correction at the apex of the deformity.

32.3 Perioperative Measures and Surgical Techniques in General

The evaluation, diagnostic workup, and documentation of patients and the planning of surgery are all performed as usual in skeletally immature

children with spinal deformity. The freehand technique of pedicle screw insertion in children is described in detail in the chapter on VCR (Chap. 34). Pedicle screws are preferable to hooks or wires because of their greater biomechanical properties and thus better potential to achieve and maintain the correction [11, 12]. The placement of pedicle screws at strategically important sites prior to vertebral resection is of extreme importance, since it allows the control of spinal alignment and hence protection of the neural structures while the osteotomy is being performed. The use of pedicle screws in the growing spine does not seem to be associated with significant negative effects in the long term [7].

The patient is positioned (see Chap. 34) prone on a radiolucent operating table. Ideally, one should use a foam-like semielastic frame that allows the abdomen to float freely and allows gravity to assist in pulling the lumbar spine into lordosis but keeps the thoracic spine through the rib cage in kyphosis. During the exposure, meticulous preparation is recommended to preserve the periosteum and to minimize the chance of unwanted fusion at the surgical site. This keeps the options open for future surgical planning, both during and at the end of growth guidance.

Following the osteotomy, if compression is performed as part of the correction, special care must be taken to sufficiently undercut the osteotomy site above and below, to avoid neural compression and/or excessive dural buckling while closing the osteotomy site. Closure or opening of the osteotomy site may occur passively as a result of the positioning on the surgical frame and/or actively by applying the usual correction maneuvers (inserting pre-bent rods into the previously placed bone anchors, in situ bending, cantilever correction, compression, etc.).

Postoperative Management Pedicle screw instrumentation provides enough stability to mobilize patients without any external orthosis; therefore, children can ambulate immediately after surgery. The authors think that they do not need to be encouraged to get up and mobilize themselves, as this usually happens as soon as the wound pain subsides. According to other surgeons' practice, the children are usually mobilized the day after surgery and require considerable encouragement.

Radiographic and clinical follow-ups should be performed at regular intervals, dependent on the rate of growth.

32.4 Types of Posterior Osteotomies

Compared with the philosophy for the treatment of adolescents/adults, a different philosophy is needed in the treatment of the growing spine, since growth must be promoted. For this purpose, an osteotomy without relevant resection and with distraction is much more suitable. Furthermore, planning of an osteotomy in children is different compared to the adults due to the differences in size proportions of vertebral elements (Fig. 32.2). The SRS-Schwab classification of osteotomies is mostly (though not exclusively) associated with resection of an anatomical structure along with compression during correction and therefore is of limited use in grading osteotomies in early-onset deformities. Nonetheless, it will be mentioned below whenever applicable.

The surgical techniques of Smith-Petersen or Ponte osteotomies and pedicle subtraction osteotomies in adults have been described in the literature several times since 1945 and can be found in almost all spine surgery textbooks. The surgical techniques of such osteotomies are similar in the adolescent population, but there are certain differences when it comes to younger children. Here, we will focus mainly on the differences in technique.

Hemivertebrectomy is described in other chapters (see Chaps. 18 and 32.) Rib osteotomies should and can be avoided in the growing spine. The ribcage is usually elastic enough to perform correction of the spinal deformity. Only in rare cases (e.g., congenitally fused ribs) should a rib osteotomy be performed.

Some of the osteotomies, such as the concave side opening wedge osteotomy or hemivertebra resection, are almost exclusively done in pediatric spine surgery, while others (e.g., PSO) are

used infrequently. There are differences not only in the surgical technique but also, or even more importantly, in the philosophy of management and indication for one technique over another, in particular in regard to the growth potential of the child's spine.

32.4.1 Soft Tissue Releases Through a Posterior Approach

All posterior osteotomies begin with exposure of the bone and removal of the soft tissues. Such soft tissue release, usually at the apex of the curve, thus serves as the basis for all osteotomies. It may sometimes loosen up the spine sufficiently to allow deformity correction without the need for any further osteotomy. The ligamentum flavum, the interspinous ligaments, and also the joint capsule and cartilage are thicker in small children compared with adults or even adolescents. These structures can be resected with an appropriate instrument, a Kerrison or Luer rongeur, according the surgeon's preference. The flavum is opened most easily in the midline by a Luer rongeur, and as soon as the spinal canal is opened, the epidural fat and not infrequently a tiny vessel become visible through the narrow defect in the ligamentum. Epidural veins might interfere with dissection in the desired plane. Therefore a Woodson-like instrument or the tip of the Kerrison rongeur itself can be used to protect the dura and develop the plane just underneath the ligamentum. Resection of the flavum, which usually becomes thicker laterally toward the junction with the joint capsule, is carried out. During this maneuver, injury to epidural veins may cause vigorous bleeding, but this can usually be well controlled by gentle compression through cottonoid patties with or without fibrin sealant or a haemostatic matrix (e.g., thrombin-soaked gelfoam). Wide soft tissue resection allows for palpation in the spinal canal, which can be used on rare occasions also for orientation to place a pedicle screw. It also allows passage of sublaminar wires or cables, if necessary. Such soft tissue release can usually be done quickly; the additional blood loss is insubstantial and it adds minimal further risk to the surgery, but it offers several advantages. Segmental mobility can be examined before and after soft tissue release by distracting and relaxing the segment with an appropriate instrument (an osteotomy spreader). If necessary, the inferior articular process can also be resected (grade 1 osteotomy). If the release achieved proves to be insufficient, progression to higher grades of osteotomy should follow.

32.4.2 Posterior Osteotomies: Complete Facet Joint Resection

Smith-Petersen osteotomy (SPO) or Ponte osteotomy is a grade 2 osteotomy and consists of removal of the facet joint. They are indicated when the previously performed soft tissue release is insufficient to mobilize the motion segment. It allows reduction of the forces required for correction and facilitates correction of the deformity in the sagittal (kyphosis), coronal (scoliosis), and axial (rotation) planes. The latter means that derotation of the spine is facilitated, which results in better reduction of the rib hump.

In young children with a substantial growth potential (early-onset deformity patients), this technique is not recommended, especially if the implantation of growing rods are planned. Following a facet joint resection (SPO), there is a high chance of fusion, which generally should be avoided at all costs in surgery of the growing spine.

SPO is usually applied in adults and adolescents if the scoliosis is greater than 70–75° and does not reduce to less than 40° with bending or, in the case of kyphosis, that only corrects to greater than 40–50° in hyperextension. To our knowledge, there are no published data for resection-correction values in children, only for adolescents and adults. For the latter, each millimeter of resection in the setting of an SPO translates to 1 degree of correction, with a theoretical correction of 10–15° per level and a more practically achievable correction of 5–10° per level in the sagittal plane. To obtain an overall correction greater than 10–15°, SPOs on multiple levels may be performed, as in the case of rigid sagittal plane deformity caused by severe

Scheuermann's kyphosis or the loss of lumbar lordosis (flat back syndrome) after operative treatment of scoliosis [13, 14].

In the thoracic spine, the osteotomy runs in a transverse direction, and in the lumbar spine it is more chevron ("V") shaped, due to differences in the direction of the facet joints. After the posterior column osteotomy is performed, correction of the deformity follows by applying compression forces. This maneuver opens the anterior column (disk space) over a hinge on the posterior edge of the vertebral bodies (posterior longitudinal ligament) and the osteotomy site (posterior column) can be closed by the same amount as was resected. The posterior column is thus shortened and the anterior column is lengthened or unchanged depending on the location of the fulcrum between the vertebral bodies. The intervertebral space can be rigid for various reasons, including a collapsed or hypoplastic intervertebral disk or previously performed spinal fusion. Such rigidity of the intervertebral disk space of course limits the amount of correction achievable through posterior release only. This technique is usually applied on multiple levels.

Operative Technique The choice of osteotomy level depends on the level of the apex of the deformity. Pedicle screws are usually already in place when performing the osteotomy. The screw heads pose minimal, if any, obstruction to the osteotomy. For an SPO a complete facetectomy must be performed. This begins with a soft tissue release as described above, removing the entire ligamentum flavum, followed by removal of the caudal portion of the lamina, the whole spinous process, and the superior and inferior articular processes on both sides. Due to the orientation of the spinous processes in the thoracic spine, the spinous process of the vertebra just cephalad to the osteotomy site must be resected to the base too. The resection is ultimately wedge or chevron shaped, with the point oriented distally. Typically, the width of the osteotomy is 7–10 mm depending on the age and size of the child and also on the amount of correction that is aimed for. Depending on whether a coronal plane correction is desired, the limbs of the wedge resection can be asymmetrical. In this situation, one side of the wedge is widened more than the other. To promote de-rotation in the case of significant rotational deformity, the osteotomy should be more extensive (greater resection) on the side of the convexity. There is a risk of impingement of neural structures during correction (nerve roots laterally or dura/spinal cord in the midline) (see Sect. 40.3), but this is less pronounced compared with a higher-grade osteotomy. One should not forget that the site of an osteotomy is at risk of spontaneous fusion.

32.4.3 Concave Side Opening Wedge Osteotomy

A failure of segmentation (bar formation) can lead to inhibited growth ipsilaterally. Coupled with normal growth contralaterally, such a natural one-sided arrest of longitudinal growth leads to deformity. In such cases of congenital scoliosis, the hampered growth in the bar results in progression of the scoliosis during childhood (Fig. 32.3a, b). A logical step would be to promote growth in this area. To this end, the osseous bar on the concavity can be osteotomized and distracted to compensate for the relative (compared to the other side) or one-sided shortness (Figs. 32.3 and 32.4). Such a distraction maneuver is not without danger in terms of potential for neurologic injury, and therefore the use of multimodal intraoperative neuromonitoring (MIOM) performed by an experienced neurologist (or technician) is even more important than in other cases of deformity correction using osteotomy. The idea, the pathoanatomical observations, and the surgical technique of the opening wedge osteotomy were all developed by the senior author (DJ). The first report on the technique and later the results of a series of patients showing it to be a successful method have been presented previously [15–17].

When planning surgery, several imaging modalities should be used to analyze the pathoanatomy. The relative location and alignment of the spinal cord must be noted. Any clinical or radiological signs of myelopathy or compromise of the spinal cord itself must be ruled out. These would otherwise pose additional risk of

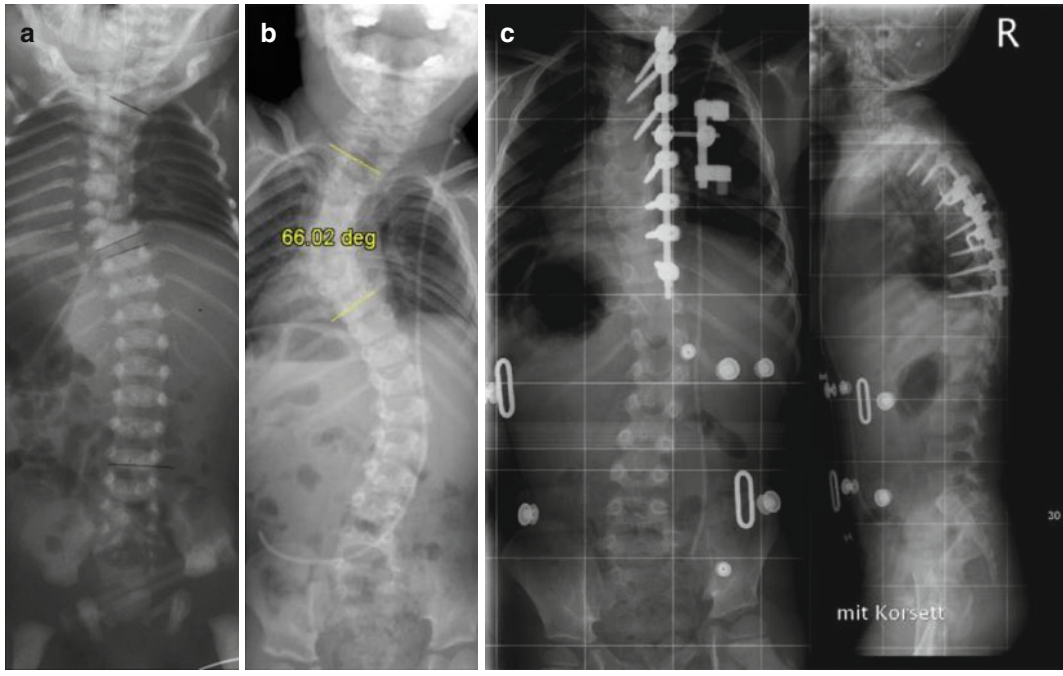

Fig. 32.3 Left convex congenital scoliosis due to failure of segmentation and formation between T3 and T9. The ribs 5–8 on the right are fused. (**a**) AP view at the age of 1 day. Note the normal alignment of the lumbar spine. (**b**) Significant progression occurred by the age of 5 years. (**c**) AP and lateral radiographs after concave side opening wedge osteotomy and instrumentation including additional rib hooks. Bracing was necessary for the lumbar curve which developed secondary structural changes during preoperative progression. Note the expansion of the rib cage pre- (**b**) and postoperatively (**c**) which is achieved virtually immediately. This expansion is very important for the development of the lungs

intraoperative spinal cord injury during manipulation and correction, and the technique would have to be abandoned. The failure of segmentation frequently involves the nerve roots and radicular vessels. One must check the preoperative imaging for nerve roots at the site of the planned osteotomy. An absence of these structures on the concave side, the site of the congenital bar, facilitates preparation of the osteotomy site and the correction maneuver itself; the spinal cord is not anchored to the concave side by the nerve roots and the segmental blood supply of the spinal cord is not exposed to risk. It is characteristic that the failure of segmentation is associated with some sort of failure of formation too. This results in a hypoplastic anterior column; the vertebral body or bodies are smaller on the concavity, and due to the deformity-associated rotation, they seem to move posteriorly over the course of development/growth. This renders the osteotomy technically less demanding.

Ideally, when performing the opening of the wedge, the fulcrum of rotation at the osteotomy level should lie on the contralateral side of the congenital bar but close to the expected position of the spinal cord following the correction maneuver. This means that the spinal cord itself undergoes negligible distraction. The spinal cord might even be indirectly decompressed by being moved away from the concavity such that it does not drape over the lateral wall of the spinal canal on the concave side. Nevertheless, this is the most critical step of the correction. Therefore, the correction maneuver (distraction) has to be done slowly and/or in a stepwise fashion over several minutes, allowing the timely recognition of any negative changes in the neuromonitoring and, if necessary, reversal of the correction. The use of a temporary rod might be

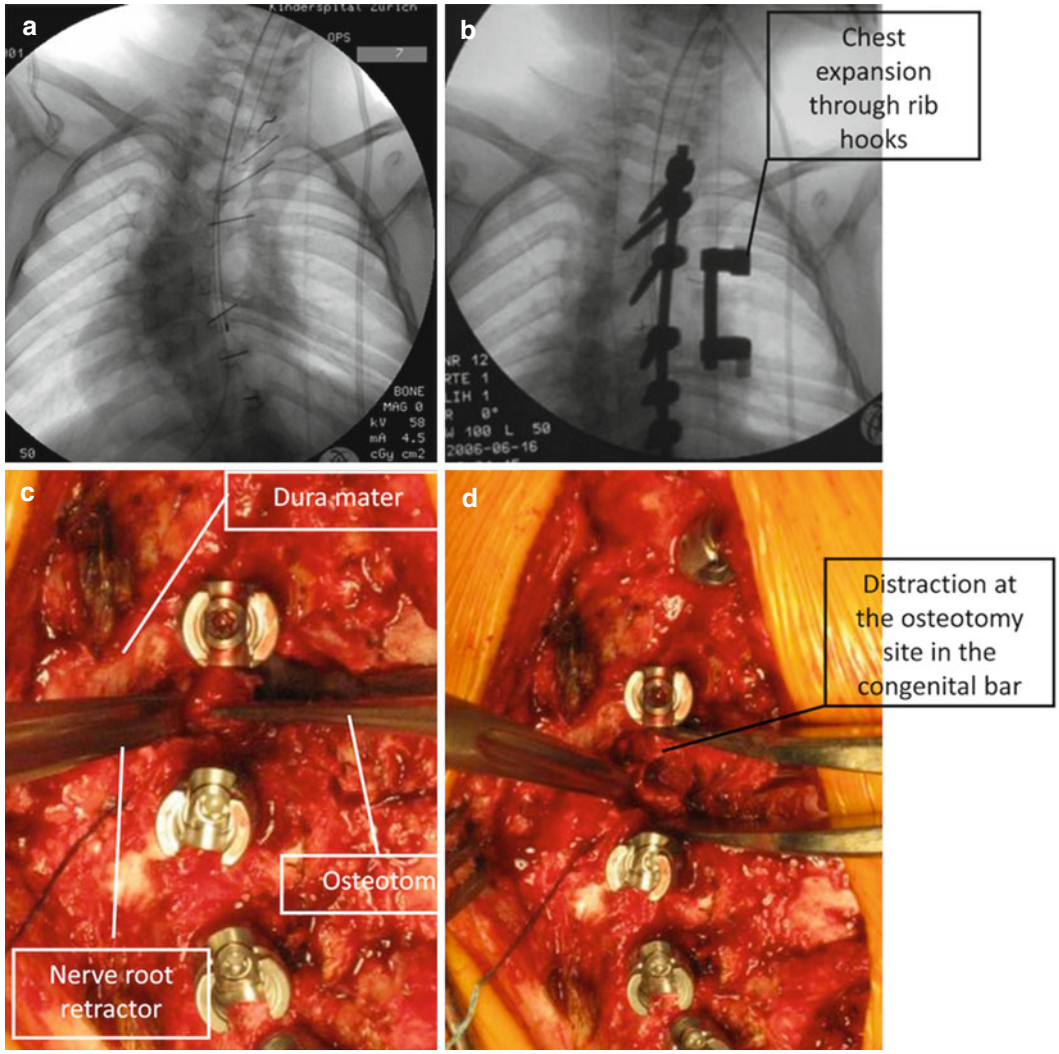

Fig. 32.4 Shows an opening wedge osteotomy through a posterior approach. (**a**, **b**) Intraoperative C-arm images in AP view; (**c**, **d**): the corresponding photos. (**a**) The k-wires are placed. (**b**) The pedicle screws are inserted and the osteotomy site is opened on the right side between the T5 and T6 pedicle screws. An additional rib osteotomy was done due to the fused ribs. The rib hooks and a rod provides additional stability

beneficial if the convex side does not seem to be stable enough. After the correction maneuver, the opened space can be left as it is. The children in whom this technique is used are usually not heavier than 10–15 kg and so the pedicle screw-rod construct alone provides sufficient stability. To avoid spontaneous fusion at the osteotomy site, a gore-tex patch or similar material can be inserted over the surfaces of the osteotomy. Avoiding fusion at the osteotomy site allows further correction to be performed at the same site at a later stage, several years down the line. The addition of, or conversion to, a growing rod construct can be performed along with, or after, the initial correction. During the growth guidance over the years, the convex side, which is considered to grow at a normal pace, is the indicator for the amount of lengthening.

Such an osteotomy can also be performed in a previously operated patient with a fusion mass, although the anatomical situation (presence of radicular vessels and nerve roots) and scar tissue may be associated with higher risks.

32.5 Summary

The most important aim of treatment in early-onset spinal deformities is to correct the deformity as soon and as completely as possible. This ensures a balanced growth of healthy regions of the spine. It also prevents the development of secondary structural changes. Ideally, the treatment focus should not extend beyond the site of primary deformity; the surgical correction should be kept as short as possible. This can be achieved by the careful planning of osteotomies. Since children have different body and vertebral proportions compared to adults, the principles of osteotomy in adults cannot be simply transferred. Instead, careful individual planning for each spinal segment involved in the deformity is required. Avoidance of fusion should be aimed for wherever possible if substantial growth potential remains, and the application of growing instrumentation should be considered.

Osteotomies provide excellent correction, although the increased operation time, blood loss, and transient destabilization of the spine is inherently associated with increased risk of neurological injury. Therefore, the use of multimodal intraoperative neuromonitoring is warranted.

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Vertebral Resection

33

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Key Points

- Severe, early-onset scoliosis (EOS) is likely to progress and can become life-threatening.
- Corrective surgery should be carried out as early and as completely as possible to prevent formation of secondary structural changes and to ensure a balanced spinal growth.
- Vertebral column resection (VCR), involving the removal of at least one vertebra with the two adjoining intervertebral discs, allows substantial correction of the deformity in a short section of the spine (e.g. in an angular deformity).
- Preservation of the periosteum remote from the VCR site is of utmost importance in skeletally immature patients, to prevent unwanted spontaneous fusion.
- Pedicle-screw constructs provide sufficient stability, do not influence spinal growth negatively and may be converted to a growing rod construct later.

33.1 Introduction

Progressive, severe, early-onset scoliosis (EOS) in younger children can be life-threatening if pulmonary development is impaired. By definition,

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EOS includes deformities with an onset of 10 years or less [1]. EOS is frequently encountered in complex congenital syndromes [2, 3]. Conservative treatment is often inadequate and surgical intervention becomes unavoidable.

Vertebral resection (VR) (also known as vertebral column resection (VCR), columnotomy, vertebrectomy or three-column osteotomy) has become established as a standard surgical procedure to correct severe deformities that cannot be adequately dealt with using alternative techniques such as multiple Smith-Petersen osteotomies (SPOs) or pedicle subtraction osteotomy (PSO). VCR is defined as a circumferential resection of at least one vertebra with all its anterior and posterior elements along with the two adjoining intervertebral discs. According to the comprehensive classification of osteotomies by Schwab et al., it represents the type 5 or 6 osteotomy [4]. VCR for treating spinal deformities was first described by Bradford and Tribus [5] as a combined anterior and posterior approach (and was later established as a single posterior approach by Suk et al. [6]). The reports published to date mostly concern adolescent and adult patients [5–14]. The surgical treatment of EOS differs from that of the deformities of skeletally more mature patients (adolescents and adults) for many reasons. Excessive extension of the instrumented fusion above and below the VCR site with the intention of improving safety/stability is not recommended. The subsequent arrest of growth in the operated area would lead to permanently impaired pulmonary function and an unacceptably short trunk and would pose a risk of the crankshaft phenomenon occurring.

In our opinion, VCR can be more effective in the growing spine than in adolescents and adults. It not only corrects the deformity effectively but also has the potential to guide further spinal growth, e.g. by the reduction of compensatory lumbar hyperlordosis with correction of thoracic kyphosis or vice versa, or the improvement of a compensatory scoliotic curve.

33.2 Evaluation

It is important to perform a thorough history and physical examination. Normally children do not complain of any pain. The main reason for seeking medical attention is usually the rapid progression of a visible spinal deformity and trunk asymmetry.

The identification and treatment of any concomitant medical problem is necessary prior to the surgical intervention, as some cases of EOS are elements of extremely rare or even unknown syndromes [2, 3].

The respiratory status of the patient should be evaluated thoroughly, which may be difficult in such young children. If respiratory function is compromised, it should be evaluated by a paediatric pulmonologist. Occasionally pulmonary function can be optimized prior to surgery, with halo traction.

The physical examination should focus on the flexibility of the spinal deformity as well as on the evaluation of coronal and sagittal plane balance and decompensation. It is important to evaluate the neurological status of the child including the sitting/standing and walking capabilities whenever possible. However, in the very early phase of development of motor skills (pre-sitting and preambulatory), the evaluation of balance is not worthwhile. A neurophysiological examination might also be justified. Furthermore, accompanying pathologies of the extremities, such as contractures or instabilities of major joints, should be looked for.

Radiological evaluation includes a posteroanterior (PA) and lateral standing radiographs in ambulatory patients and sitting anteroposterior (AP) and lateral radiographs in non-ambulatory patients. The magnitude of curves is determined in both the coronal and sagittal planes using the standard Cobb method. In some severe deformities, it may be difficult to measure the Cobb angles correctly. Trunk balance is determined by measuring the deviation from the midline at the sacrum of a plumb line dropped from the spinous process of C7 on the PA view and from the body of C7 on the lateral view. Additional radiographs are necessary for evaluating the flexibility of the deformity. AP supine maximally bending right and left radiographs are obtained with the pelvis fixed. Manual axial traction supine radiographs in the PA and lateral views are also performed. All patients planned for VCR should be evaluated by magnetic resonance imaging (MRI) and CT. 3D CT reconstruction helps to better understand

the morphology and the structural changes of the spine. If any intraspinal pathology such as syringomyelia, diastematomyelia or spinal cord tethering is found, its management should be considered prior to correction of the deformity. It should be noted that the presence of such a pathology involving the spinal cord increases the risk of neurological injury during correction.

33.3 Management

VCR is reserved for cases where other surgical techniques would not be sufficient to achieve an appropriate correction and balance of the spine (Table 33.1). VCR is applicable for all aetiologies with severe deformity including idiopathic, neuromuscular, syndromic or congenital deformities.

The ultimate goal of surgery is to halt progression and to achieve maximum correction of the deformity in order to allow for balanced growth and to improve pulmonary function. This is achieved with rigid internal fixation and fusion of the spine over the shortest possible section. Stable internal fixation mostly obviates the need for any postoperative orthosis. The short arthrodesis allows for motion preservation of the unaffected areas of the spine. However, in very severe cases with an extremely rigid spine, a longer non-fusion pedicle screw instrumentation may be necessary to avoid an early loss of correction with screw pullout. These long constructs can later be shortened, replaced by a growing rod system or even removed.

If the planned correction and instrumentation cannot be carried out due to the severity or configuration of the existing curve and/or insufficient lung function, preoperative halo traction is necessary (Figs. 33.1 and 33.2). Halo-gravity traction provides an acceptable means of continuous traction for several weeks or months. The traction devices are applied for each of the major body positions (halo-wheelchair, halo-bed and ambulatory halo-frame). For further details, see Chap. 30.

The collaboration with experienced paediatric anaesthesia teams and critical care teams is of paramount importance in the management of EOS patients. Preoperative anaesthesia evaluation several weeks prior to surgery is encouraged since many patients have multi-organ disorders as part of a syndrome and nutritional problems. Pulmonary function requires careful monitoring and support during surgery and in the immediately postoperative period. A tracheostomy and a percutaneous endoscopic gastrostomy (PEG) may become necessary. Blood loss during surgery is often substantial and the use of a cell saver system is recommended.

Table 33.1 Comparison of different surgical techniques for severe early-onset spinal deformities (authors' experience)

	In situ fusion	Stapling	Growing rods	Shilla	Vertebral resection	VEPTR
Applicability in severe deformities	+	+	++	++	+++	++
Correction, rate/speed	+	+	++	++	+++	+
Grade of immediate correction	0	0/+	++	++	+++	++
Ability to prevent secondary structural changes	+	+	++	++	+++	+
Surgical risks/complication rate	+	++	++	++	+++	+++
Immediate effect on thorax size after surgery	0	0	++	++	+++	++
Long-term effect on thorax function	0	+	++	++	++	+
Technically demanding	+	++	++	++	+++	+

Stapling and tethering can be considered in adolescents
0 no effect, + small effect, ++ moderate effect, +++ strong effect

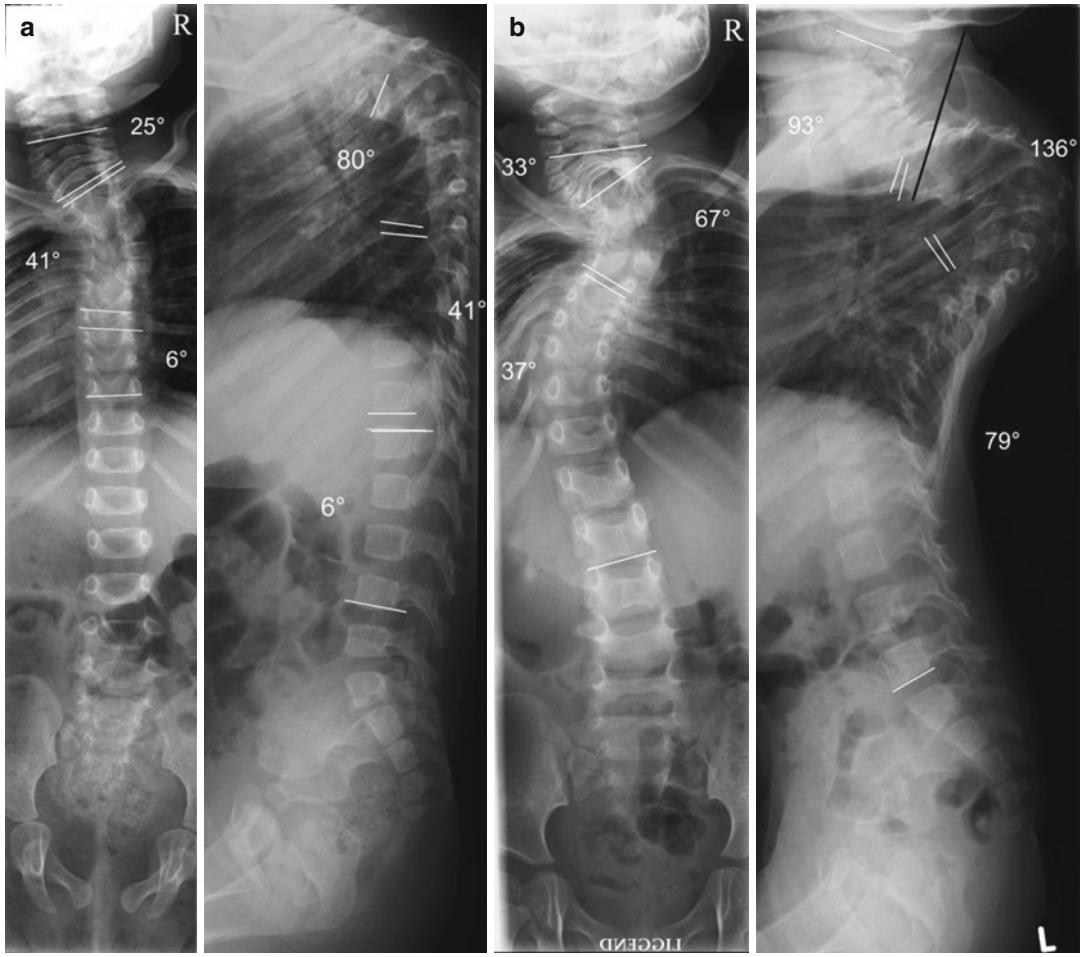

Fig. 33.1 **(a)** EOS caused by an unknown syndrome. Sitting lateral and posteroanterior radiographs of a girl 9 months of age demonstrate a high thoracic kyphoscoliotic curve pattern. The Cobb angles were as follows: left cervical scoliosis, 25° (C4–T1); right thoracic scoliosis, 41° (T2–T6); left thoracic scoliosis, 6° (T7–T10); cervical lordosis, not measurable on the first X-rays; thoracic kyphosis, 80° (T2–T6); thoracic lordosis, 16° (T7–T12); lumbar kyphosis, 10° (L1–L4) (**b).** The follow-up shows a rapid deterioration in 15 months time. The lying radiographs demonstrate a thoracic kyphosis of 136° (T2–T7) and a right thoracic scoliosis of 67° (T2–T6). Note how quickly the thorax collapses along with the spine. This results in a deterioration of lung function. At admission, at 2 years of age, the patient suffered from resting dyspnoea and had to lean on something to support her upper body and expand her chest in order to be able to breathe. Note the positions of the pedicles in the upper thoracic area. The *black line* in the lateral view indicates the trajectory of a planned pedicle screw. In this severe deformity, pedicle screws in this region cannot be inserted due to spatial limitations of the skull

Curve correction is considered to be associated with a high risk of neurological complications. The use of multimodal intraoperative monitoring (MIOM) is recommended as it reduces the risk of intraoperative neurological injury [15]. MIOM obviates the need for any form of wake-up test. A close collaboration between the anaesthetist and the monitoring neurologist or technician is important. Hypotensive anaesthesia helps to reduce intraoperative blood loss, but it can complicate the interpretation of MIOM.

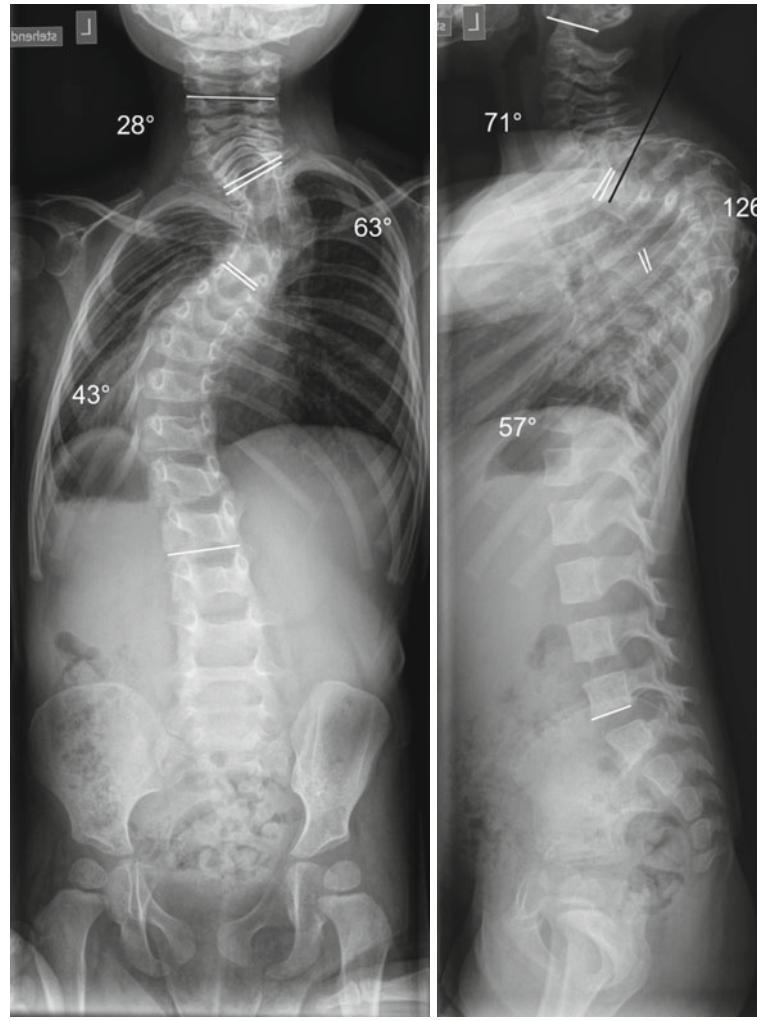

Fig. 33.2 Same patient as in Fig. 33.1. Notable correction in terms of spinal alignment and chest configuration is achieved after 2 months of halo traction. Lung function and the activity level of the child improved considerably. The cervical lordosis decreased from 93° to 71°, such that instrumentation with pedicle screws in the upper thoracic area became feasible (the *black line* again indicates the planned trajectory of the Th2 pedicle screw). It became obvious which part of the curvature was rigid and with a first correction the surgery became less demanding

33.4 Surgical Procedure

Other techniques for correction of EOS, including posterior spinal fusion, convex growth arrest, hemivertebrectomy, insertion of growing rods, thoracic expansion, vertebral osteotomies, non-fusion and motion preservation techniques, are described elsewhere in this textbook.

VCR is appropriate in the case of severe angular spinal deformity where pedicle subtraction osteotomy or a series of osteotomies would not be sufficient to achieve acceptable correction. Thorough preoperative planning is required to determine the number of vertebrae to be removed during surgery and the length of the pedicle screw segmental spinal instrumentation. Nonetheless, the preoperative plan may have to be modified according to the intraoperative findings, and the surgeon must have a certain amount of flexibility regarding the extent of resection. It is sometimes necessary to remove two or even three apical vertebrae, but the number of vertebrae removed should be kept as low as possible. It is therefore advisable to begin with resection of the apical

vertebra, with further resection being performed in a stepwise fashion, if necessary. In some cases, additional spinal osteotomies might be required.

33.4.1 Surgical Technique

Stand-alone posterior surgery posterior VCR is the recommended technique, as this offers more advantages compared with combined approaches [6]. With VCR, there is no thoracotomy and postoperative thorax wall scarring leading to thoracic deformities, which might impair the development of the lungs. For older children (above 10 years of age), the recommendations for treating deformities become increasingly similar to those for adults. The posterior approach allows for all the necessary steps for safe vertebral resection and correction. It allows continuous visual control of the neural structures during resection and correction. A costotransversectomy on one or on both sides in the thoracic area, or removal of the transverse processes in the lumbar area, provides good visualization and a capacious working area. This approach also allows for safe circumferential preparation around the vertebral body and control of the segmental vessels. One drawback is the difficulty in controlling the major vessels in the prevertebral area. In some cases, a combined anteroposterior or postero-anteroposterior approach cannot be avoided [5, 16, 17].

A conventional radiolucent orthopaedic surgery table is used, with the patient being placed in a prone position on specially made Maquet-like adjustable modular foam pillows that act as a frame. Children under halo-gravity traction are positioned with a traction weight of 1–3 kg.

Technically, there are small differences compared with the vertebral resections described elsewhere [5, 6]. In contrast to the usual subperiosteal preparation, preservation of the periosteal layer is attempted to avoid unnecessary spontaneous fusion and allow for unaltered or minimally altered growth. This also allows for conversion to a growing rod system or even for implant removal to regain mobility at a later stage (Figs. 33.3 and 33.4). This is followed by verification of the correct levels using fluoroscopy, usually in the PA projection. The next step is to identify the entry points of all planned pedicle screws by using anatomical landmarks. With the standard technique, a pedicle finder or probe is used to identify and prepare the pedicle followed by tapping or using a self-tapping screw. Alternatively, according to our practice, and especially in small pedicles, a 22 Gauge needle is first inserted followed by fluoroscopic verification in anteroposterior and lateral views. Then the pedicle is prepared using a 1.5 mm drill. Pre-bent K-wires are inserted into the holes, and fluoroscopic imaging is performed. Fine adjustment of the screw trajectories can be made in the next step using a 2.0 or 2.5 mm drill for final preparation. In most cases, pedicle screws with a diameter of 3.5 mm are inserted, though depending on the diameter of the pedicles, 2.7, 4.0, 4.75 or 5.00 mm diameter screws may also be used. Screws are preferable to hooks or wires because of their better stability and thus superior potential to achieve and maintain the correction (primary stability) [18, 19]. It also seems that pedicle screws in the growing spine do not result in clinically significant alteration in the development of the spinal canal [20]. The placement of pedicle screws at strategically important sites prior to vertebral resection is of major importance, since it allows spinal alignment to be controlled and the neural structures to be protected during the unstable phase of the resection and correction.

It is helpful to mark the exiting nerve roots using a vessel loop. A single nerve root may be sacrificed in the thoracic area between T3 and T9. However, a deficiency of T4 and/or T5 root function may lead to sensory loss in the mammary area, which may cause problems later in life (e.g. with breastfeeding).

The existence of distinctive tissue layers, due to the thick periosteum, facilitates the resection of the vertebral body. The intervertebral discs above and below the vertebral body are resected first. Bony resection does necessarily require the use of a burr, typical of an eggshell procedure carried out in adults. In contrast, the vertebral body can often be removed in one piece with preservation of the posterior wall. During the phase of resection, a unilateral rod is applied, to provide temporary stabilization and prevent

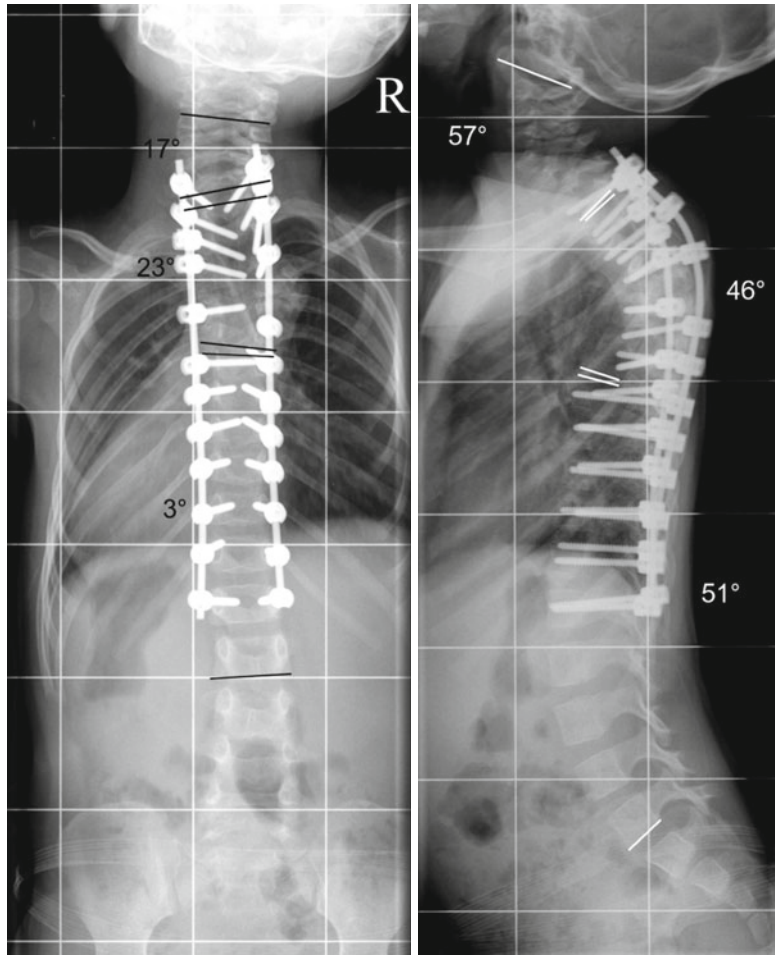

Fig. 33.3 Subperiosteal preparation was applied during the surgical procedure to avoid spontaneous fusion. Polyaxial pedicle screws with a diameter of 3.5 mm were inserted between T1 and L1 on both sides and were connected with 3 mm rods on either side. Such long instrumentation was required to correct the secondary hyperlordosis in the thoracic area. A bone-on-bone fusion was only performed between T4 and T6. The autologous bone material gained by the resection of the T5 vertebra was used to enhance fusion. Resection of the T5 vertebra was performed with costotransversectomy T5 and T6 on the right side and T5 on the left side through a posterior approach. The thoracic kyphosis was decreased to 46° and the right thoracic scoliosis decreased to 23° between T2 and T6. Note the near physiological sagittal and coronal alignment of the spine and the marked change of the chest wall immediately after surgery. The almost complete correction of the deformity resulted in correct alignment at the non-instrumented area. This allowed for undisturbed development of the spine, preventing secondary structural changes at the previously healthy spine segments

translational movements. Above and below the vertebral resection site, two blocks of several spinal segments are instrumented with pedicle screws. This ensures optimal control and an even distribution of forces during the correction manoeuvres (in situ bending, translation, rod rotation and compression/distraction). At this stage, during which the spine is substantially destabilized, it is essential to avoid translation, subluxation and dural impingement. The correction has to be performed slowly and carefully, with continuous inspection of the dural sac/spinal cord and nerve roots. In addition, careful and repeated circumferential palpation of the dura is necessary to assess any impingement, overdistraction or excessive shortening.

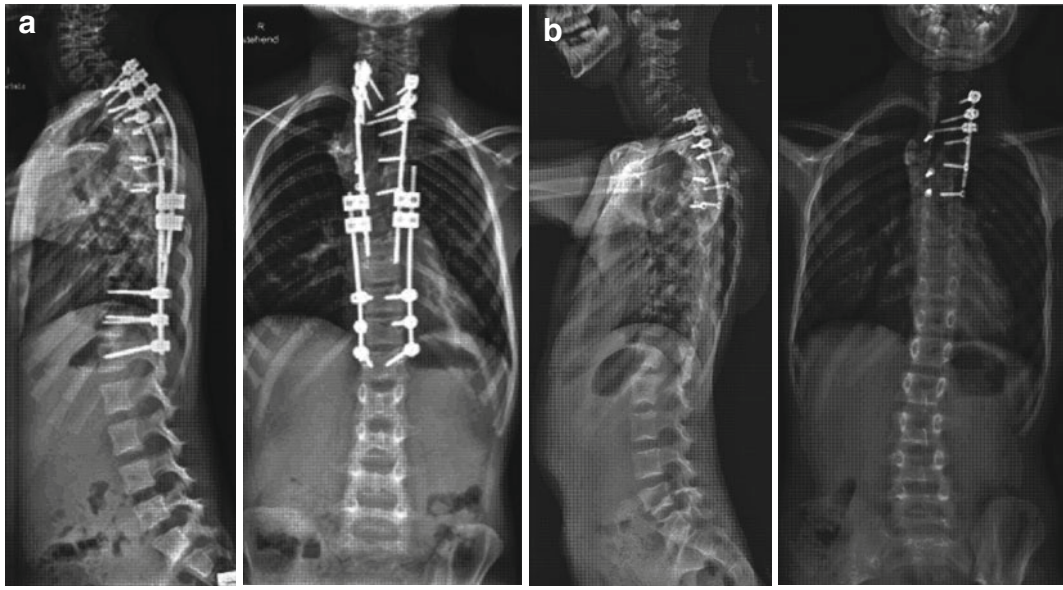

Fig. 33.4 Segmental instrumentation was replaced by a double growing rod construct after 3.6 years (**a**) followed by multiple distractions. The curvature improved during continued growth and the non-fusion instrumentation could be partially removed, i.e. shortened (**b**, radiographs at the age of 8 years). The motion segments were still functioning, as no fusion was intended in this area. Close follow-up is necessary to detect any deterioration, especially during the prepubertal growth spurt

The length of the spinal cord should ideally remain constant, with the theoretical pivot point of correction being at the given spinal level and within the confines of the spinal cord. This usually involves a shortening of the posterior elements and in most cases lengthening anteriorly. Anterior column support is achieved either by simple compression (bone-on-bone contact) or by inserting a cage or rarely a structural graft (rib, fibula or iliac crest).

Based on intraoperative findings and the achieved correction, additional osteotomies and further vertebral resection may be necessary.

In the authors' experience, rib resection (thoracoplasty) on the convex side and osteotomy on the concave side should be avoided. These structures are usually flexible enough in EOS patients.

Unfortunately, intraoperative radiographs in the prone position may fail to indicate whether the spine is correctly aligned with respect to the plumb line. Spinal balance can more reliably be evaluated in the postoperative standing or sitting radiographs. If a substantial residual imbalance is present, revision surgery for rebalancing the spine should be carried out as soon as possible.

33.4.2 Postoperative Management

In young children—just as in adults—pedicle screw instrumentation provides sufficient stability to allow patients to be mobilized without the use of a cast or orthosis. Children usually do not need to be encouraged to get up and move; they typically mobilize themselves as soon as the wound pain had subsided. Physiotherapy is carried out mainly for the improvement of lung function. Follow-up assessments are carried out at 8 weeks, 6 months and 1 year after surgery and include repeat radiographic and clinical assessment. Lung function tests might also be important. Depending on the stage of spinal growth, further follow-ups are typically planned at 6 months to 1 year intervals. If a significant residual deformity is present, conversion to a growing rod construct with repeated distraction may need to be considered.

33.4.3 Documentation

We recommend thorough documentation of deformities with radiological imaging supplemented with clinical photographs (Fig. 33.4). The photographs should be taken preoperatively and postoperatively in a standardized fashion. Video documentation might also be useful. Documentation should be repeated at all regular follow-ups and will allow for analysis of any changes in spinal deformity and the whole body. This is not only helpful for scientific or professional purposes, but it also demonstrates the surgical result to the patients and the parents.

33.5 Discussion

There are many aetiologies of EOS, which include idiopathic, neuromuscular, syndromic and congenital [2, 3].

Ideally one should not let a spinal deformity progress to a severe stage. If severe curves develop due to delay in treatment for any reason, severe chest deformity can develop, resulting in cardiopulmonary problems. In extremely severe cases, children can present with resting dyspnoea, due to their being at the limit of their respiratory compensatory mechanisms. Some surgeons advocate only incomplete correction, which is considered to reduce the risk of surgery [1]. However, the best long-term results in young children are probably obtained with full correction of the deformity, aiming for restoration of physiological curvatures. The proper management of children with progressive, severe EOS has proven difficult. Attempting any type of conservative treatment may be hazardous. Many of the well-established treatment methods for EOS such as growing rod procedures, hemiepiphysiodesis, VEPTR, Shilla procedures, growth modulation and others should be reserved for mild or moderate deformities due to technical issues or their slow effect (Table 33.1).

VCR is an established surgical treatment for severe and rigid spinal deformities over a short region. It is an extensive procedure that is indicated where less-invasive surgical techniques such as SPO and PSO would be insufficient to achieve optimal correction and restore balance. However, it is mostly used in adolescents and adults and its application in young children is not yet well established. For this reason, treatment standards are lacking. Only a few recent studies have included younger patients [9, 11], and in these the main focus was on patients 5 years of age and older. Basically, VCR in very young children can be compared to the vertebral resection technique described by Bradford and Suk [5, 21]. However, in our experience there are some small but important differences in the young patient population that should be considered during treatment. These are summarized in Table 33.2.

Table 33.2 Important differences between VCR in patients with EOS (≤10 years of age) compared with adolescents/adults

Parameters used in adults for assessment of global spinal balance cannot easily be transferred to preambulatory children
Surgery as early as possible and with maximal correction at the site of the main deformity prevents the development of nonstructural compensatory curves in intact spinal segments
Preservation of the periosteum away from the VCR site is of utmost importance to prevent unwanted spontaneous fusion
Long pedicle screw constructs (non-fusion) provide sufficient stability but do not influence spinal growth negatively
With a balanced growth, a shortening of a long pedicle screw construct may be considered to free-up spinal segments
The spine and the chest wall are more elastic: Allows for sufficient correction of the spinal deformity Thoracoplasty is usually unnecessary. Correction of the underlying spinal deformity helps to guide the development of the chest wall
Conversion to a growing rod construct for the remaining curve should be considered

In a recent study we analyzed the clinical and radiological results of VCR in a patient group with a mean age of 3.7 years. With VCR the kyphosis angle was reduced from 126° (87–151°) to 61° (47–75°). The mean operation time was 500 min (range, 463–541 min) with a mean blood loss of 762 ml (range, 600–1050 ml). There were no neurological complications. However, hardware failure with loss of correction, halo pin infection/dislocation and skin problems occurred [22].

Some authors would recommend the use of vertical expandable prosthetic titanium rib (VEPTR) [23, 24]; however, if the thoracic deformity is secondary to the spinal deformity, we do not believe that there is any rationale for addressing the secondary chest wall deformity.

Some studies have suggested that the shortening of the spine with VCR may be associated with a further decrease in pulmonary function [25]. However, the actual length of the spine is only one factor governing lung function and development: the curvature of the spine, and consequently the volume, configuration and function of the chest wall, should also be taken into account. Early spinal resection (with shortening) and correction of the deformity stops the unbalanced growth of the spine (Fig. 33.3) and thorax and results in an overall lengthening of the anterior trunk during growth. Compared with kyphosis correction using distraction techniques [26, 27], another advantage of shortening the spine is that it is better tolerated by the spinal cord. In this respect, it is similar to resection of a hemivertebra, another standard spine-shortening procedure. The latter is a less radical procedure, performed in less severe deformities, but it used to be regarded as a difficult and demanding procedure; in the meantime, it has become a standard treatment method [28].

The circumferential resection, or disconnection of the spine, allows correction in any direction. The realignment is limited mainly by the neural structures, i.e. the spinal cord. During realignment, the pivot point is defined in the spinal cord area. Thus, although VCR is a shortening procedure with regard to the spine, for the spinal cord, it is a neutral or only minimally shortening procedure. Therefore even with the large amount of freedom for realignment, it bears an acceptable risk of neurological injury.

In contrast to adolescent and adult deformities, balance of the spine is not the most important factor to consider in young children. More important are the severity of deformity and the rate of progression. In many cases these young patients have not developed any upright posture yet and therefore the notion of spinal balance is not yet relevant. If the young patient is already able to sit or stand, and if an imbalance is detected, it can reinforce the surgical indication. However, values describing imbalance in the adult spine are not easily transferable to children in the first 5 years of life because of their principally different body proportions.

For surgical indications in adolescent and adult deformity surgery, there are relatively clear-cut values to indicate the degree of deformity and the point at which surgery should be performed. Such limits cannot be applied to a severe early onset curvature. Instead, these have to be evaluated individually, in view of the progression and neurological impairment. Correction of severe deformities in young children must be considered a prophylactic intervention with respect to the remaining hitherto unimpaired sections of the spine.

Preoperative halo-gravity traction may be applied to decrease the spinal curvature, and in some cases it enables instrumentation which otherwise would not be possible. During traction, the abnormally shortened soft tissues including vessels, muscles, ligaments and neural structures are stretched. Furthermore, the chest volume and pulmonary function are increased, such that surgery may become an option in children who initially had severe respiratory insufficiency.

Reliable, strong, but also short anchoring is necessary to achieve and maintain correction, for which the pedicle screw–rod system seems to be the most appropriate.

Following a vertebral resection procedure, a subsequent change to a growing rod instrumentation might be necessary if there is any residual deformity or if longer (non-fusion) instrumentation is required.

Current experience with VCR in EOS in young children is still limited but the initial results are encouraging. It appears to be an effective surgical procedure for the treatment of severe deformities over a short spinal region in very young children, just as it is in adults. However, there are important differences between its use in skeletally immature children and in older patients. Additional surgical procedures may be necessary in the young, growing spine. The improved pulmonary function and optimized spinal balance result in an increased ability to perform daily activities and an improved quality of life, making it a worthwhile treatment for a subset of early-onset spinal deformities (Fig. 33.5).

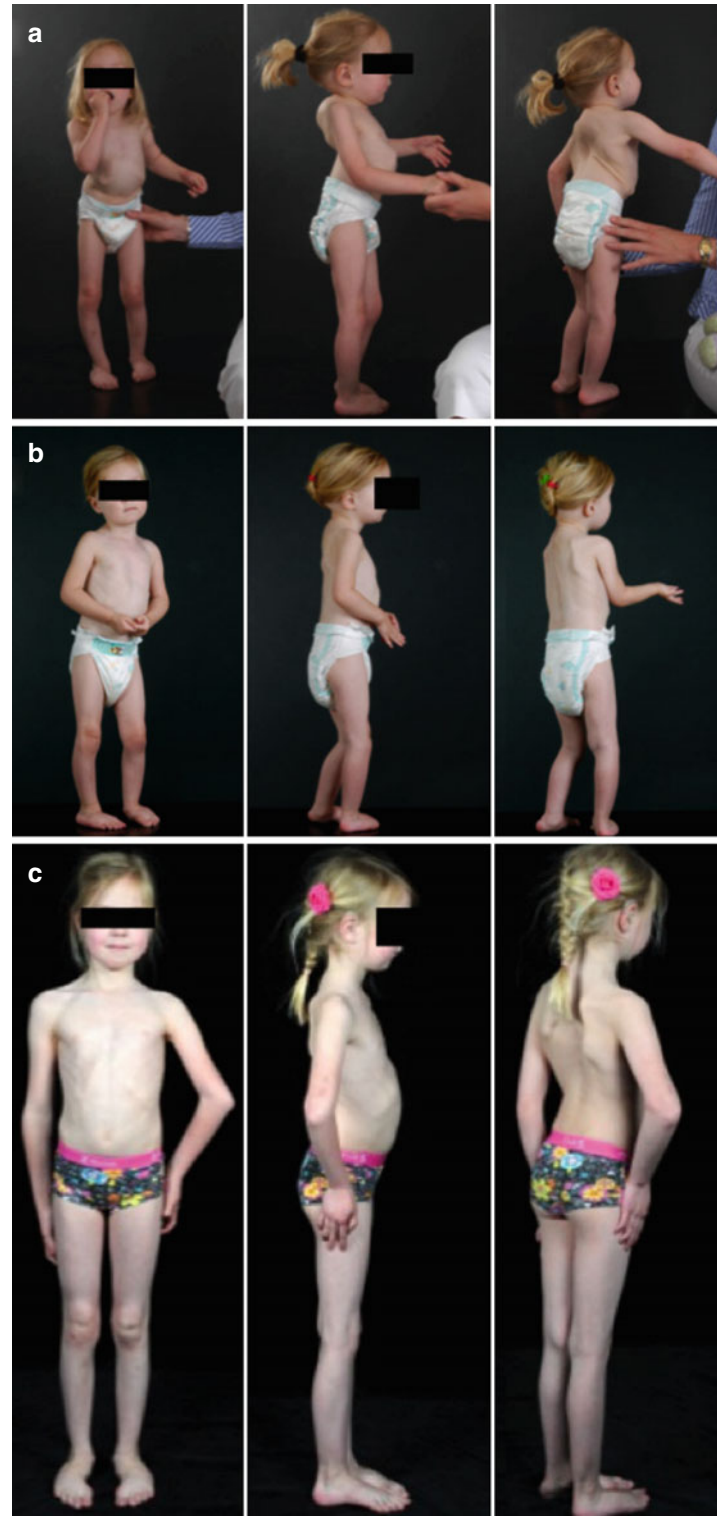

Fig. 33.5 Comparison of the preoperative (**a**) and immediately postoperative (**b**) images demonstrates a marked direct and immediate change of the whole-body morphology and symmetry caused by the radical spinal correction with vertebral resection. The child was able to stand alone due to the improved balance of the spine. This highlights the importance of the time factor: the earlier the correction is achieved, the less the secondary damage and abnormal development. The latest clinical pictures demonstrate a normal trunk at the age of 7 years (**c**)

33.6 Summary

The most important aim in the treatment of early-onset spinal deformities is to correct the deformity as early and as completely as possible. This ensures a balanced growth of the healthy regions of the spine and it prevents the development of secondary structural changes. Ideally, the treatment focus should not extend beyond the site of the primary deformity; the surgical correction should be kept as short as possible.

Severe EOSD is usually due to either a rapid progression or an unnecessary hesitation in introducing a corrective treatment. If the deformity is in a relatively short segment (angulated deformity), vertebral resection is indicated, i.e. a surgical procedure involving the removal of at least one vertebra with the two adjoining intervertebral discs. This allows for a short fusion whilst providing the fastest and maximum correction of the deformity. If a complete correction cannot be achieved, vertebral resection provides the option to substantially reduce the deformity such that other surgical techniques, e.g. additional osteotomies or a growing rod treatment, can later be used.

VCR is an extensive surgical procedure. Currently it is only used if other treatment modalities are considered likely to fail. However, with increasing experience, an extension to its indications can be expected, to include its application in less-pronounced deformities. Finally, the most important factor in the treatment of EOSD is the early onset of treatment.

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Surgical Management of Kyphosis in Meningomyelocele

34

Jozef Murar and John F. Sarwark

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Key Points

1. Congenital kyphosis can be a serious and difficult problem in patients with myelomeningocele.
2. Surgical intervention has been shown to be beneficial to the growing and developing child.
3. There are several surgical techniques, including vertebral resection and subtraction kyphectomy, which may be used to correct the deformity with good results.
4. The operating surgeon must be aware that these procedures have been associated with a high morbidity and mortality, and the family must be made aware of this prior to proceeding.
5. Treatment of the patient with myelomeningocele should be undertaken with a multidisciplinary approach in order to give the patient the best outcomes.

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34.1 Highlights/Background

Myelomeningocele is defined as a defect of the neural tube during embryonal development. With failure of closure of the caudad end of the neural tube, there is a resulting sac that may contain spinal cord, nerve roots, and meninges [1].

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Commonly the level of the defect correlates with the patient's neurologic deficits. Congenital vertebral anomalies, some producing scoliosis and kyphosis, have been associated with myelomeningocele secondary to the malformations that occur. Although not as common as scoliosis, kyphotic deformities occur in 8–20 % of myelomeningocele patients and are commonly associated with high motor level deficit involving thoracic or upper lumbar levels [2–6]. The kyphotic deformity in myelomeningocele patients is progressive, and as a result, one must consider early intervention. Kyphosis progression can approximate 8° annually in most children, and patients may already have a significant deformity present at birth [2, 3]. The underlying progressive nature of the curve has been attributed to the lack of muscular stabilization as a result of the absence of appropriate motor function as well as a lack of posterior bony elements. The erector spinae musculature is displaced anterior to the axis of the spine and, with a hypertrophied psoas and the tenodesis effect of the crus of the diaphragm, creates a strong flexor moment on the spine. Additionally, once patients become upright with sitting, the flexion moment arm is compounded by gravity and a progressive kyphotic deformity results. Ultimately the deformity becomes fixed and structural, and the vertebral bodies become wedged shaped [2, 7].

Intervention to restore sagittal balance has been well accepted in patients with severe kyphotic deformities to improve postural stability and sitting balance. Kyphectomy-type reconstructive surgery can be a useful treatment modality, specifically in patients with myelomeningocele, to achieve this goal. With significant sagittal plane spinal deformities, kyphectomy and instrumentation allows for satisfactory long-term correction.

The indications and surgical technique for kyphectomy have been well described in the literature [2, 8–10]. With recent and further advances in operative technique, the procedure has demonstrated safety and efficacy as an effective surgical management for the child with severe sagittal plane deformity and for the orthopedic surgeon.

34.2 Overview of Medical/ Neurosurgical Issues

Prior to early neurosurgical intervention, the mortality rate of patients with myelomeningocele was as high as 90–100 % [1]. With early intervention including sac closure and ventriculoperitoneal shunt placement, the survival rates have significantly improved. There is a very high incidence of associated hydrocephalus necessitating shunt placement. Additionally, due to an associated abnormal development of the cephalad portion of the neural tube, there is an association with Arnold-Chiari II malformation. Surgical repair of the Chiari II malformation is indicated in up to 15–35 % of patients. The procedures include an occipital craniotomy and upper cervical laminectomy for decompression [2].

Another commonly associated neurosurgical condition is tethered spinal cord syndrome. Symptomatic tethering of the spinal cord may occur in 25–30 % of patients with myelomeningocele. Given that most patients undergo sac closure, dural scarring is inevitable and almost always seen on magnetic resonance imaging. Therefore, tethered spinal cord syndrome is a clinical diagnosis with radiographic confirmation. Signs and symptoms include back and leg pain, motor and strength deterioration, urodynamic changes, rapid progression of scoliosis or kyphosis, and spasticity/contracture. This condition is treated with tethered spinal cord release (TSCR) neurosurgical exploration of the scarred dural sac with mobilization of the tight nerve roots and filum terminale. Patients may ultimately not recover their baseline motor or sensory function. Without appropriate recognition and early TSCR, an orthopedic surgeon may not adequately treat a kyphotic deformity if there is an underlying tethered cord syndrome [1, 11].

Associated medical issues with regard to kyphotic deformity in these patients include both urological and gastrointestinal. Patients may have neurogenic bladder dysfunction depending on the level of the spinal cord lesion. This may ultimately lead to urinary incontinence and impaired bladder emptying. Ultimate management may entail prevention of infections and

monitoring of renal function. Patients may also have an abnormal anal sphincter tone and anorectal sensation leading to problems with bowel incontinence and constipation [2].

Nonsurgical management of the kyphotic deformity usually proves futile. With the abnormal posture that results from the kyphotic deformity, patients often rely on the upper extremities for sitting stability and support. A patient may experience severe loss of independence as there is a dependence on use of the hands for sitting. Conservative, nonsurgical treatment including bracing has proven to be ineffective and ultimately does not prevent progression. Many times the patient is unable to lay prone due to the severe deformity. Additionally, there may be associated skin breakdown at the level of the spinal deformity, and this can lead to a vicious cycle of pressure sores. The ulceration over bony prominences combined with thin, scarred, and somewhat insensate skin leads to a difficult problem [2, 7, 8, 12, 13].

34.3 Perioperative Management

Patients require an extensive preoperative evaluation involving a multidisciplinary approach. The neurosurgeon should be actively involved, and the ventriculoperitoneal shunt should be tested [2]. Additionally, it can be worthwhile to determine the anatomic course of the abdominal aorta; however, it has been shown that the abdominal aorta is at little risk during kyphectomy correction surgical techniques since it does not follow the path of the kyphosis but rather spans it [2, 3]. Also, given the potential for chronic wound issues, it might be necessary to consult a plastic surgeon preoperatively to optimize the wound healing potential. It may be necessary to admit the patient preoperatively for wound care, including prone nursing care up to 2 weeks [14]. Postoperatively, the combined involvement of physical and occupational therapy is necessary to aid in the patient's recovery. Additionally, an orthotist may also be utilized to fabricate a thoracolumbosacral orthosis (TLSO).

34.3.1 Surgical Strategies: General

The objectives of a kyphectomy procedure are complete restoration of sagittal alignment, balance, and stability while simultaneously allowing the child to grow and achieve appropriate truncal height (Figs. 34.1, 34.2, 34.3, and 34.4). Surgical correction for congenital kyphosis is performed for the clinical manifestations of the kyphotic deformity rather than absolute radiological measurements. The primary indications for operative intervention include increasing spinal deformity,

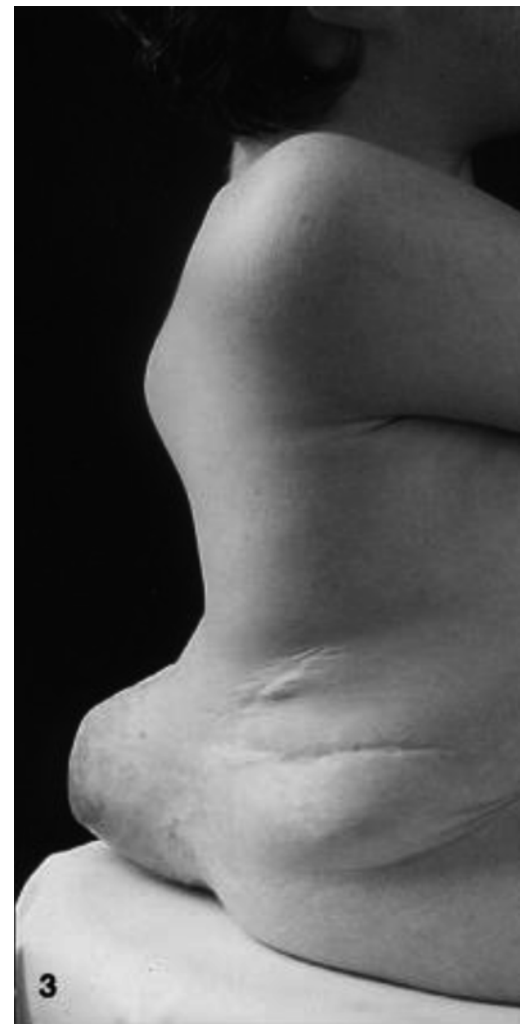

Fig. 34.1 14-year-old with a prominent gibbus and kyphectomy deformity (Reprinted from Furderer et al. [7] with permission from Springer Science)

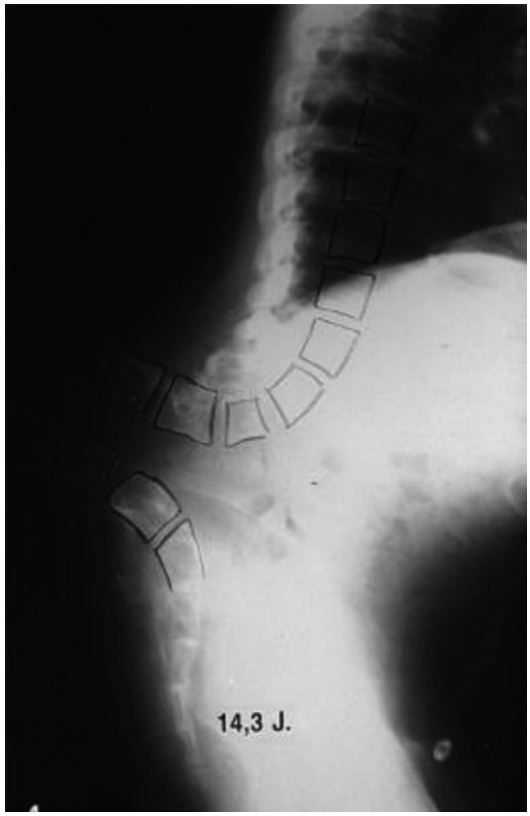

Fig. 34.2 Lateral radiograph demonstrating severe kyphotic deformity (Reprinted from Furderer et al. [7] with permission from Springer Science)

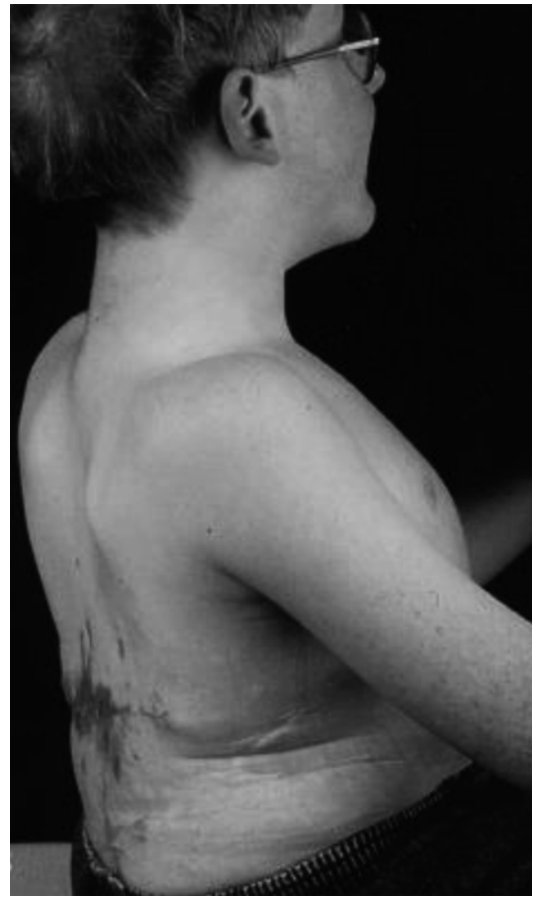

Fig. 34.3 Same patient from fig. 34.1 after surgical correction (Reprinted from Furderer et al. [7] with permission from Springer Science)

the need for primary skin closure over the spinal dysraphic defect and protuberant bone; recurrent decubitus skin ulceration over the kyphotic apex causing chronic debilitation; inability of placement in an upright sitting or standing position without the use of both upper extremities for support; and reduction in the anterior abdominal wall available surface area prohibiting necessary gastroenterology or urology procedures and fitting of appliances; significant compression of abdominal contents during upright posture creates upward pressure on the diaphragm and respiratory compromise. Costal margin impingement on the pelvis may cause pain and discomfort [15]. The major contraindication to surgical intervention is associated medical condition prohibiting a surgery of this magnitude [1, 11].

One of the most studied kyphectomy surgical procedures is vertebral resection with modified Luque fixation (resection kyphectomy). As shown by Lindseth in 1979, in addition to vertebral resection of the apex of the kyphotic deformity, resection of 1.5–2.5 vertebral bodies cephalad to the apical vertebra must also be performed to best correct the lumbar kyphosis and distal rigid compensatory thoracic lordosis [5]. The extent of resection must extend to the lordotic segment. One of the often-cited limitations of this procedure includes the potential inability to allow for complete preservation of the dural sac and subsequent potential life-threatening postoperative complications that may occur such as increased epidural bleeding and specifically acute hydrocephalus [16, 17]. Winston et al. also reported a case of sudden death from elevated

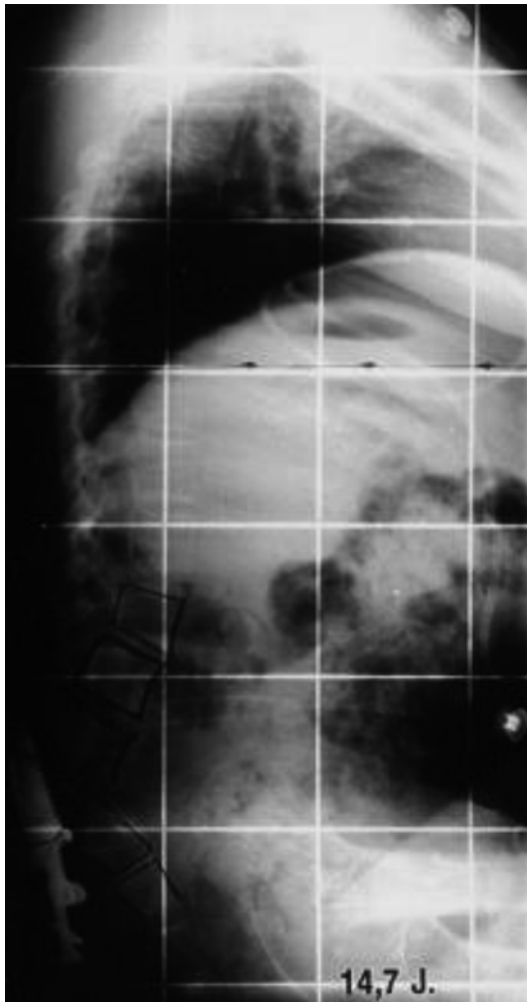

Fig. 34.4 Lateral radiographs after correction of kyphotic deformity (Reprinted from Furderer et al. [7] with permission from Springer Science)

intracranial pressure presumably due to CSF flow changes after ligation of the thecal sac [18]. Additionally, this procedure leads to vertebral shortening and an indirect reduction of tension on the spinal cord [3].

Another kyphectomy surgical technique is subtraction (decancellation) vertebrectomy. Conceptually, the decancellation kyphectomy technique is a lordosing intravertebral apical osteotomy over multiple lumbar levels. This procedure obviates the need to perform a cordotomy and the resulting associated morbidities of cerebrospinal fluid flow problems such as meningitis and acute hydrocephalus [3]. Additionally, this method is not a complete resection of a significant portion of the spinal column, allowing for preservation of spinal height. The lordosing kyphectomy is tethered along the length of the anterior longitudinal ligament, thus avoiding tension on neurovascular structures. The advantages of this procedure when compared to the vertebral resection include a satisfactory sagittal correction, preservation of the dural sac leading to fewer shunt complications, less blood loss, and decreased operative time [16]. The preferred age for surgery seems to be between 2 and 5 years of age, when the anteroposterior diameter reaches a 25 mm minimum [3].

With either the resection kyphectomy or decancellation kyphectomy, stable instrumentation is required to stabilize the osteotomies and sagittal correction, prevent recurrence, and allow for seating stability. The pelvis or sacrum should be included distally—in most instances—to prevent lumbosacral sagittal plane deformity. Additionally, the instrumentation must correct the developmental thoracic lordosis by including the thoracic spine to the level of T4–T6. A wide range of long-term results has been generated when performing a resection kyphectomy using different instrumentation techniques including the use of Harrington rods; plate fixation; Luque rods; combinations of cables, hooks, and wires; the Galveston technique; Dunn-McCarthy fixation; or the Warner and Fackler technique [14, 19–25]. However, the current consensus is that segmental posterior spinal instrumentation with inclusion of the sacropelvis is necessary to attain and maintain sagittal correction, which, again, can be achieved with multiple different techniques [8, 16, 17, 19, 20, 22, 25, 26].

34.3.2 Surgical Technique Specific

34.3.2.1 Vertebral Resection Kyphectomy Technique

The procedure is performed with the patient under general anesthesia, prone positioning, a radiolucent operating table, and a frame or chest rolls. A posterior midline longitudinal incision is

used and developed through the area of previous closure. If a tissue expansion procedure has been completed, the tissue expanders are removed at the conclusion and closure of the case. The thoracic paraspinal muscles are subperiosteally dissected from the thoracic posterior spinal elements. At the lumbosacral junction, the dural sac is dissected, and the proximal stump is oversewn. Evidence of a functioning shunt must be determined preoperatively. Of note, the visualization of cerebrospinal fluid during this portion of the procedure precludes closure of the neural plaque in fear of precipitating acute hydrocephalus [22]. Then, the dural sac is retracted proximal to the osteotomy site.

Dissection is performed laterally and anteriorly around the kyphosis in order to access the sinus of the kyphosis. The kyphectomy is then performed via vertebral excision of the proximal aspect of the apical vertebra and one to two vertebral bodies cephalad to the kyphotic apex [5].

Modified segmental instrumentation using the Luque technique as outlined by McCall may be utilized to correct and stabilize the kyphectomy (Figs. 34.5, 34.6, and 34.7) [22]. Contoured Luque rods are brought through the S1 foramen bilaterally, with the distal ends lying on the anterior aspect of the sacrum. The distal ends of the Luque rods are bent in accordance with the patient's sacral inclination, approximately 20–40°. A cross-link is placed distally near the sacral foramen to prevent rod migration and rotation. The rods are placed just medial to the lateral masses at the level of the osteotomy site and subsequently wired to the higher-level thoracic lamina sequentially starting at T4, progressively reducing the osteotomy site and creating more rigid fixation. The osteotomy site is augmented with bone from the vertebrectomy to perform a local arthrodesis [22].

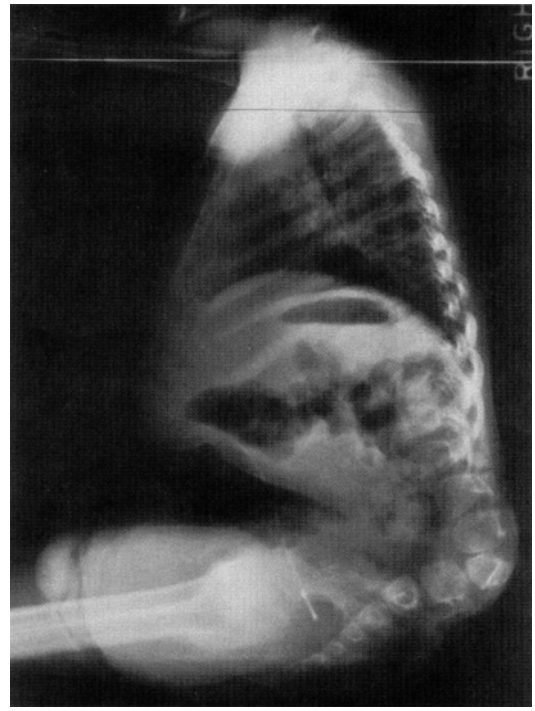

Fig. 34.5 5-year-old child with kyphotic deformity (Reprinted from McCall [22] with permission from Wolters Kluwer Health)

34.3.2.2 Subtraction (Decancellation) Kyphectomy Technique: Pedicle Subtraction Osteotomy

The operation is performed with the patient under general anesthesia and prone positioning using a radiolucent operating table and frame or chest rolls. The procedure is performed through a posterior midline incision. The laminar bars are subperiosteally exposed laterally and medially. The dorsal laminar bar overlying each respective neuroforamen is resected for isolation of each pedicle needed for decancellation. The thecal sac and nerve roots are mobilized medially in the subperiosteal plane. The pedicle is entered with a curette from one side, and the cancellous bone is progressively evacuated from the vertebral body to the midline. Subsequently, this canal is packed and the procedure is repeated on the contralateral side, resulting in a cortical shell of vertebral body. A fine curette is used to etch a line in the anterior cortex of the vertebral body from pedicle to pedicle. The osteotomy will close or hinge on this line. This decancellation procedure is then repeated over contiguous levels from caudad to cephalad, usually beginning at L4 and progressing to L1. A correction of 45° per vertebral level can be expected with restitution of lumbar lordosis, thoracic kyphosis, and sagittal balance [3].

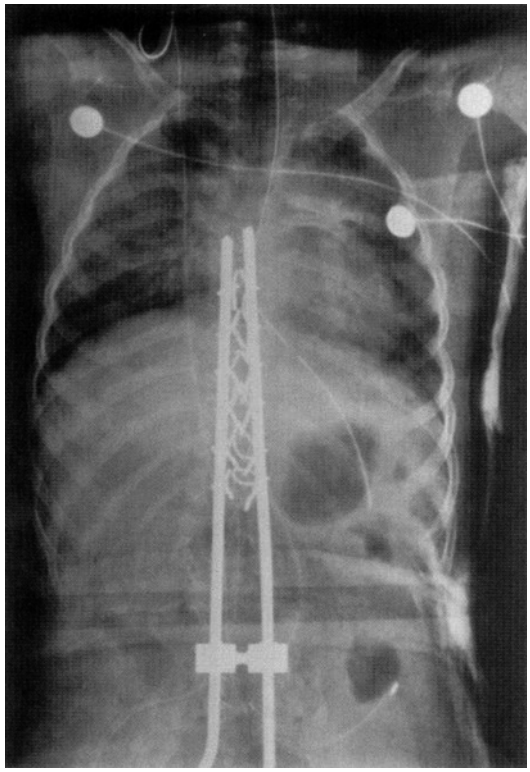

Fig. 34.6 Postoperative AP image showing rod construct (Reprinted from McCall [22] with permission from Wolters Kluwer Health)

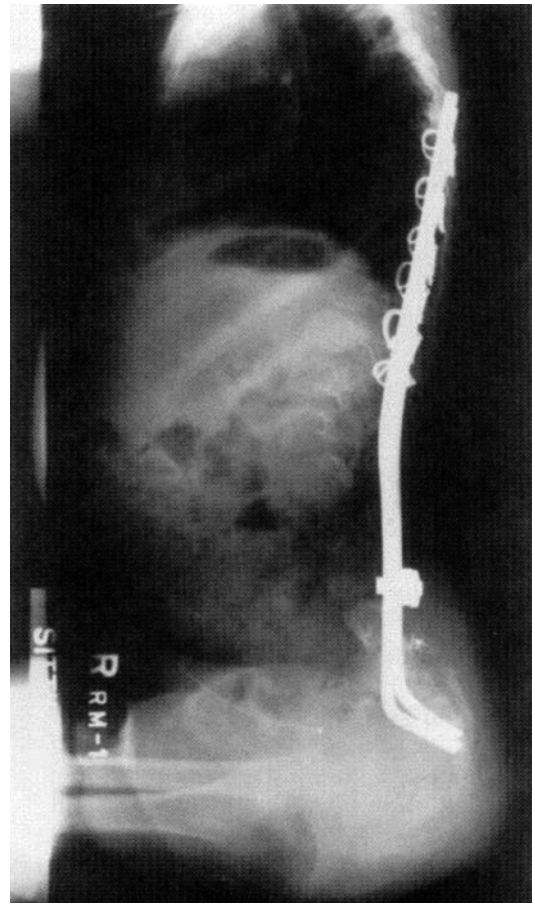

Fig. 34.7 6-month postoperative x-ray of same patient with demonstration of kyphotic reduction with Luque rods (Reprinted from McCall [22] with permission from Wolters Kluwer Health)

Stable instrumentation is required to stabilize the multilevel osteotomies and correction, prevent recurrence, and allow for seating stability and long-term stability. Again, the pelvis or sacrum must be included distally to prevent lumbosacral sagittal plane deformity. Additionally, the instrumentation must correct the rigid developmental thoracic lordosis by including the thoracic spine to the level of T4–T6. Instrumentation techniques include posterior stabilization with neutral or sagittally contoured, paired rods; segmental thoracic hooks or sublaminar wires; lumbosacral pedicle screw fixation with intrasacral distal rod insertion (Roger Jackson technique); and limited arthrodesis at the lumbosacral fixation points allowing growth proximally [16]. Furthermore, a growing construct can be attempted by extraperiosteal dissection for thoracic lamina exposure, limited lumbosacral arthrodesis, preservation of cartilaginous end plates at the decancellated levels, and utilization of segmental thoracic hooks, sublaminar wires, or cables (Fig. 34.8) [3].

34.3.3 Results

The results from the resection kyphectomy have varied dependent on the form of instrumentation. However, from these prior studies, it has been agreed upon that segmental spinal instrumentation with sacral fixation is important to obtain and maintain kyphectomy correction and stability.

Sharrard (1968) and Lindseth (1979) first described resection of vertebral bodies for treatment of kyphosis, which required significant

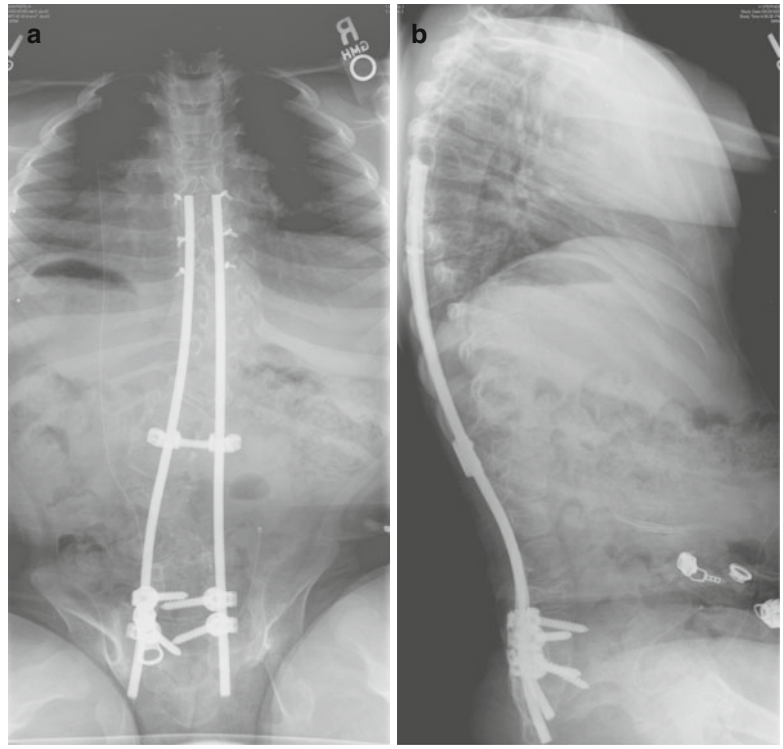

Fig. 34.8 (**a**) Postoperative AP image of subtraction kyphectomy and instrumentation. A growth construct is placed proximally to allow for column growth with arthrodesis only in the lumbosacral region. (**b**) Postoperative lateral image of subtraction kyphectomy and instrumentation. Lumbosacral fixation includes L5 and S1 pedicle screw fixation and intra-alar rod insertion. Rod contour is normal sagittal profile

postoperative immobilization [5, 27]. Since that time though, many different techniques for resection kyphectomy have been developed. Heydemann and Gillespie reported improved results using sublaminar wiring at the osteotomy site supplemented with Luque rods placed anterior to the sacrum distally, which no longer require postoperative external immobilization [20]. They showed a kyphosis correction from 124° to 33° postoperatively which was maintained at final follow-up. McCarthy in 1989 concluded that long posterior spinal fusion with Luque rod instrumentation achieved the best outcomes; and the Dunn McCarthy modification involved placement of Luque rods into the sacral ala [28]. Warner and Fackler also modified sacral anchoring and achieved improved correction and stability [25]. Their technique has since been a popular treatment method in institutions around the world. Lintner and Lindseth demonstrated that resection kyphectomy resulted in a mean correction to 40° kyphosis postoperatively and 62° at follow-up [4]. This also demonstrated how limited fixation might lead to loss of correction over a period of time as 34 of their 39 patients had a partial loss of correction. Huang and Lubicky illustrated that resection kyphectomy and posterior spinal instrumentation using Luque rods resulted in a mean correction of 21° postoperatively and 23.7° at follow-up [17]. McCall in 1998 showed a mean correction of 15° kyphosis postoperatively and 20° of kyphosis at follow-up assessment with a mean postoperative correction of 91° [22]. Overall, these studies have demonstrated mean postoperative corrections of 84–94 % and average final 5 year retained corrections of 81–93 % performing resection kyphectomies with rigid segmented fixation with Luque rods [17, 20, 22].

Several more recent studies corroborate these findings and demonstrate an excellent correction of kyphosis with vertebral resection kyphectomies [8, 12, 13, 29–31]. Altoik et al. reviewed 33 patients who underwent kyphectomy using the Warner and Fackler technique with distal pelvic fixation and showed a correction from 124° preoperatively to 22° at last follow-up, an 81 % mean correction [12]. Similarly Samagh et al.

achieved a mean correction of 88.7 % from 115° preoperatively to 13° postoperatively using the same technique [30]. Schroeder et al.'s cohort demonstrated a 90° correction with a mean postoperative curve of 22° using the Galveston technique [29]. Comstock et al. improved kyphosis averages from 123 to 60° at final follow-up using apical posterior kyphectomy with anterior placement of fixation [8]. Lastly, most recently in 2013, Gepp et al. improved mean kyphotic degree from 116 to 63 at one-year follow-up [31]. It should be noted that all of these authors, despite what technique was used, demonstrated excellent results with regard to improvement of the kyphosis and consequent ability to improve sitting balance and help with daily care of these patients.

With regard to the decancellation kyphectomy, the results as shown by Nolden, Sarwark et al. in 2002 from the author's institution also demonstrated a substantial improvement in sagittal correction [16]. In this study, the mean correction attained immediately postoperatively was 3° of lordosis, and at latest follow-up, the correction was stabilized at 20° kyphosis. These findings represent a mean postoperative correction of 91° and a mean final correction of 66°. When performing a decancellation kyphectomy, a mean immediate postoperative correction of 96 % has been attained and an average final 2-year retained correction of 87 % [16]. Furthermore, it has been stated that when compared to the resection kyphectomy, the decancellation kyphectomy resulted in diminished overall morbidity including substantially decreased intraoperative blood loss, decreased acute VP shunt malfunctions, and decreased mortality [16].

A more recent case report by Hwang et al. in 2011 also showed an adequate kyphectomy using pedicle screw only constructs without a complete vertebrectomy and without a cordotomy [32]. The authors were able to correct the kyphotic deformity from 130 to 142° preoperatively to 52 and 50°, respectively, with a mean correction of 63 %, which is similar to calculated means in the vertebrectomy literature.

Although decancellation kyphectomy appears to have similar results in sagittal corrections as the resection kyphectomy with significantly decreased morbidity, it is important to note that long-term studies of decancellation kyphectomy in the myelomeningocele population are still pending. Long-term studies examining decancellation kyphectomies (pedicle subtraction osteotomies) in the adult population and in the non-meningomyelocele pediatric population (such as congenital kyphosis) do exist and demonstrate pedicle subtraction osteotomies (PSOs) to be reliable and safe procedures for the correction of fixed sagittal imbalance [33–38]. The largest study to date was published by Kim et al. and reviewed 140 patients who underwent a PSO for the management of sagittal imbalance due to any etiology [33]. They found a mean correction of 36° with regard to kyphosis, which appears consistent with other authors' mean corrections from 29 to 49°. In the congenital kyphosis pediatric population, Spiro et al. found that vertebral body resection as well as PSOs achieved were successful correction with both procedures and improved the kyphotic deformity from 59.9 preoperatively to 17.5 postoperatively at the final follow-up [34].

Extrapolation of the decancellation kyphectomy data from the adult and non-meningomyelocele population demonstrates that the procedure seems to be an efficacious option with decreased morbidity when compared with the resection kyphectomy. However, further long-term studies are required to corroborate these results in myelomeningocele patients, and until then, caution should be exercised if attempting to draw conclusions from these results in the myelomeningocele population.

34.3.4 Complications

Complications are commonplace in the kyphectomy procedure in myelomeningocele patients no matter what kind of approach is used. Because these are major surgeries with long intraoperative time and significant blood loss, they are subject to a rate of complications up to 90 % [4, 8, 12–14, 16, 17, 19–22, 25, 31]. Major complications that may require reoperation include deep wound infections, osteomyelitis, skin ulceration leading

to recurrent deformity and prominent implants, cerebrospinal fluid flow dysfunction, and even death. Minor complications include delayed wound healing, superficial infection, urinary tract infection, postoperative lower extremity fractures, and asymptomatic pseudarthrosis. The most common complication appears to be wound healing overall. Surgery performed by a multidisciplinary team does permit the minimization of risk, increases the rate of arthrodesis, and reduces the chance of dehiscence, which can be a major risk factor for infection [31].

Despite the high risk of complications however, the results of the surgical correction for kyphosis correction have been excellent and have resulted in facilitating rehabilitation and improving daily care of these children. It appears that the benefits of this surgery greatly outweigh the risks despite the high perioperative complications [8, 12, 13, 30, 31].

34.4 Summary

Congenital kyphosis in children with myelomeningocele precludes significant functional and social development. It is associated with high rates of impairment including multiple shunt revisions, loss of ambulation, self-esteem issues, diminished skin integrity, and urologic problems. Surgical intervention has been shown to be beneficial to the developing child. Both vertebral resection kyphectomy and subtraction (decancellation) kyphectomy procedures have been described and utilized with successful results. Whether performing a resection kyphectomy or decancellation kyphectomy, it seems that the benefits of improved sagittal correction, better sitting balance, enhanced pulmonary, gastroenterologic, and urologic function, improved skin integrity, and overall superior function outweigh the risks. The surgical techniques for correction of the kyphotic deformity have historically been associated with high morbidity and mortality. Although long-term studies in the myelomeningocele patient are still pending, it is the author's experience that demonstrates that the decancellation vertebrectomy is a safe and efficacious procedure for correction and stabilization of myelomeningocele kyphosis in young patients. The cautious surgeon is well advised to seek assistance in performing either procedure during their learning phase. Additionally, with either surgical technique, it is important to have long-term follow-up to verify longitudinal growth after spinal instrumentation [3].

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Revision Spine Surgery in the Growing Child

35

Matthew E. Cunningham
and Oheneba Boachie-Adjei

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Key Points

- Pediatric spine deformity surgery is not common (1–10/100,000), and subsequently, revision spine deformity surgery is even less common. Common indications for such revision surgery include infections, failure of healing of the fusion, implant-related problems (prominence, breakage, pain), additional deformity ("adding on," crankshaft, and junctional problems), among others.
- The revision pediatric spinal deformity surgeon must thoroughly evaluate the patient clinically, with imaging, and with any other necessary means to accurately determine why the index surgery failed, and then develop a revision plan that will address this and any other structural spine issues.
- Revision surgery has a higher probability for longer operative time, increased blood loss, and neurological injury, and for these reasons, it is recommended that only surgeons very familiar with this type of surgery attempt these difficult cases and that the operative plan is prepared to include unanticipated intraoperative findings (infection, undiagnosed pseudarthrosis, incompetent bone stock).

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- Revision surgery commonly necessitates osteotomy procedures, which allow much more powerful corrections to be obtained, but come with their own set of bleeding and neurological risks.
- Excellent outcomes can be obtained with patient population with preparedness and application of sound surgical principles.

35.1 Introduction

The need for spinal surgery in pediatric patients is estimated at approximately 1 to 10 per 100,000 of all children, with surgical indications ranging from intra- or extradural neoplasms requiring laminectomies and excision to spinal deformity requiring correction and stabilization in adolescent idiopathic scoliosis [9, 33, 49, 57]. Intermediate and long-term (greater than 5 years) rates of problems following index procedures are reported to be as high as 25–50 % [21, 32, 42], while perioperative results have much lower rates of issues (2.1 %) and include diagnoses such as implant failure, wound infection, neurological deficit, pulmonary complications, and coronal plane imbalance [29]. Problems leading to repeat surgery for adolescent idiopathic scoliotic patients at late follow-up have been reported in several studies and show that 4.6–12.9 % of patients had one or more surgeries after their index procedure, most commonly for infection, symptomatic implants, deformity, and pseudarthrosis [10, 24, 36, 45]. This estimate is considerably lower than the reported 26–46 % rate of junctional kyphosis [21, 22, 24], and higher than the 1.3–1.5 % rate of pseudarthrosis reported for the group [3, 24], indicating that decision for surgical revision is made on a case by case basis. Due to good short-term surgical results, requirement for revision spine surgery while children are still in their growing phase is uncommon, if interval surgeries for "growing rod" lengthenings are not considered.

Patient diagnoses at the time of index spinal surgery include congenital deformity, neuromuscular conditions (e.g., cerebral palsy or muscular dystrophy), connective tissue disorders (e.g., Marfan's syndrome or Ehlers-Danlos), idiopathic deformity, Scheuermann's kyphosis, and neoplastic etiologies requiring laminectomy for intradural excisions (e.g., neuroblastoma, astrocytoma, or meningioma). The available English language literature does not definitively detail the relative rates of surgical intervention for these diagnoses, nor are there large studies for each of the diagnoses with sufficient follow-up to document the relative rates for required revision surgery. Smaller studies have reported rates for postsurgical problems and revisions for many of the common deformity diagnoses. In a series of 17 patients treated for Scheuermann's kyphosis with posterior fusion with multiple posterior element osteotomies, one case of each proximal and distal junctional kyphosis were reported that did not require revision surgery at minimum of 2-year follow-up [16]. Two similar studies of Scheuermann's patients ($n=23$ and 39) reported instrumentation problems requiring revision in 10–13 % of patients [28, 30]. Children with congenital spinal deformities had requirement for repeat surgery in 6.3–33 % of cases [18, 38, 39, 47, 52], with rates of pseudarthrosis [18, 38] and instrumentation problems [18, 47] that were approximately 10 %. Revision rates for skeletally immature patients with cerebral palsy range from 5.8 to 18 %, with indications including crankshaft phenomenon, deformity progression, and instrumentation problems [12, 14]. Perhaps the highest rates of surgical revision are in the myelomeningocele population where revisions for deformity progression average 10 %, but wound problems requiring removal of instrumentation range from 10 to 75 % [31, 41]. Overall, independent of diagnosis, pediatric spine deformity revision rates occur at about 10 % of the index case rates.

35.2 Clinical Presentation, Evaluation, and Diagnosis

35.2.1 Clinical Presentation

Children in need of revision spine surgery present with a limited number of symptoms [20, 54]. Namely, these presentations are for pain, obvious

deformity progression, and loss of clinical function [12, 14, 24, 45, 50]. Pain can be the result of pseudarthrosis with or without consequent implant fatigue failure, prominent implants either as suboptimally placed at the index procedure or after dislodgement [14, 18, 45, 51], or due to muscle fatigue in the setting of pain or decompensation after index procedures. Deformity progression can be in the setting of very young patients fused from posterior-only approach that have anterior element overgrowth and crankshaft phenomenon [46, 48], patients who have "adding on" of levels proximally or distally after being fused over too short of a spinal segment [55, 56], or those patients who have degeneration above or below a fusion that leads to instability and junctional kyphosis or complex junctional deformity [22, 32, 35, 42, 50]. Functional losses may be easily articulated by the patient, such as loss of walking tolerance in the setting of junctional stenosis below a fusion, but also may be more subtle as in the requirement for increased oxygen by nasal cannula in a patient who already has cardiopulmonary compromise in the setting of slowly progressive spinal deformity.

35.2.2 Evaluation

The evaluation should begin with a thorough history and physical examination, with emphasis placed on defining the timing and character of symptoms. While conducting the interview with young patients, be attentive to subtle information that is available in the examination room. For example, in the young or nonverbal child, observation of the patient in undisturbed activity or while they interact with their parent may demonstrate valuable information about asymmetry in extremity movements, gait or balance problems, and behaviors that indicate the location of the problem (rubbing the skin over a painful implant, or demonstrating dyspnea or tachypnea due to compromised pulmonary status). Obtain as detailed an account of the index procedure as possible, including preoperative radiographs, operative notes, and a thorough description of the postoperative course, along with serial radiographs. This information will document and quantify deformity progression if present and will demonstrate the chronology and potential associations of pertinent radiographic findings to the symptoms reported. A detailed account of what happened to the patient prior to presentation for revision will provide the physician with the best means to diagnose why the index procedure failed and what revision surgery can be offered to remedy the clinical situation.

Symptoms that occur acutely, either in the first few weeks after the index procedure or abruptly after a long symptom-free interval from the index surgery, suggest implant-related problems [12, 45]. Point tenderness over a prominent portion of the instrumentation may suggest irritation of superficial soft tissue. Radiographs will help to define the implant responsible, and examining serial x-rays from the index procedure and up to presentation will help define if this is newly dislodged instrumentation or a new inflammatory process in the overlying skin of chronically prominent implants. An injection of local anesthetic and a corticosteroid can be considered for further diagnosis and potential therapeutic intervention, but will likely be poorly tolerated in younger patients. Definitive management involves implant removal, but this should be delayed until definitive fusion is confirmed, unless overlying skin is tented and threatens breakdown. Patients and their families should be warned that deformity may progress after removal of instrumentation [43]. A febrile patient, or a patient with a diffusely tender wound, with or without signs of local infection, should be considered to have a deep wound infection and must be evaluated accordingly [17]. Complete blood count with differential and serology for inflammatory markers such as erythrocyte sedimentation rate and C-reactive protein should be obtained in addition to orthogonal radiographs of the instrumented area to evaluate for lucency around the implants that often is associated with infected instrumentation. If the white blood cell count and inflammatory markers are not sufficiently supportive of the diagnosis of infection, peripheral blood cultures can be obtained, the instrumentation can be cultured by image-guided

or open biopsy, or a gallium scan can be obtained to support a diagnosis of deep infection of the implants [1, 19, 34]. If infection is diagnosed in the setting of an unfused spine or immature fusion, irrigation and debridement will be needed followed by suppressive antibiotics; maturely fused spines with deep infections involving the instrumentation require implant removal, and proper management with surgical and chemotherapeutic modalities to clear the infection. Consultation with a pediatric infectious disease specialist is recommended.

Pain symptoms with a more gradual onset, or those that have a gradual onset that acutely worsen, could represent pseudarthrosis. On examination, the patient may have increased and painful motion through the region of the fusion and tenderness to palpation in specific locations of the spine. Pain symptoms may be the result of inflammation and painful motion at the site of a single or multiple pseudarthroses or may be associated with the instrumentation (increased strain at a nearby bone-instrumentation interface or fractured implants). Radiographs should be obtained in orthogonal planes to assess the entire length of the instrumented fusion, with coned down and oblique views obtained in areas where there is suspicion for nonhealing of the fusion [4, 25, 26]. If the radiographs do not demonstrate lucencies in the fusion mass or broken implants, then computerized tomography of the fusion or bone scan should be considered. Another etiology for gradual onset of lower back pain complaints is muscular deconditioning of the lumbar extensors. This is suggested on physical examination by tenderness within and spasm of the paraspinal muscles below the fusion segment and tenderness symptoms elicited with patients returning from a bent-forward position. It is an uncommon finding in children, due to their expected high activity levels, but typically is found in conjunction with other pain generators that limit general activity levels and predispose to muscular deconditioning. Addressing the primary source of pain and restoring general activity typically lead to spontaneous strengthening and conditioning of the musculature and resolution of symptoms.

Apart from pain, a common presentation for revision deformity surgery evaluation is continued deformity progression [12, 45]. This may take the form of shoulder asymmetry, enlarging thoracic rib hump or lumbar prominence, decompensation in the coronal or sagittal plane, or diagnosis based on follow-up radiographs from the index procedure without clinical complaints. Perhaps the most feared complication in spinal surgery of the growing child is that of crankshaft phenomenon. This is typically seen in skeletally immature children that are fused at a very early age (possibly with open triradiate cartilages and Risser 0 or 1) by a posterior-only approach [46, 48]. Continued growth of the vertebral bodies within the length of the posterior fusion leads to progressive deformity, which is typically seen in the coronal and axial planes due to residual scoliosis after the index procedure. Deformity progression can be recognized as progression of the major curve by 10° or more within the length of the index fusion as assessed through serial radiographs or by progressive clinical rotational deformity documented by photographs or quantified by increasing angle of trunk rotation measured by scoliometer. Loss of shoulder, coronal, or sagittal balance may be indicative of crankshaft phenomenon, but these are also encountered when postoperative deformity is due to "adding on" of segments outside of the index fusion and with degeneration of segments flanking the index fusion. Treatment of crankshaft phenomenon typically requires anterior spinal fusion (epiphysiodesis) to arrest the anterior spinal growth driving the deformity progression, along with posterior osteotomies and revision fusion to correct and stabilize the spine.

"Adding on" of segments proximal or distal to a fusion typically occurs in the setting of an index fusion that was not sufficiently long to correct and control the spinal deformity present [55, 56]. Deformity left untreated by the index procedure then progresses in the expected manner, worsening in until the completion of skeletal growth. The diagnosis is suspected when obvious deformity progression is seen by any of a multitude of clinical parameters and is confirmed with serial radiographs documenting progression of the major curve. Junctional kyphosis, or more broadly

junctional degeneration, is a different pathological process, involving accelerated degeneration and instability at segments proximal or distal to an index fusion [21, 22, 27, 35]. This is a diagnosis that is not commonly encountered in the growing child, but is seen with frequency in the older adolescent population and in the young adult population after spinal fusion. Clinical suspicion for junctional kyphosis should be raised when patients have a feeling of "falling forward" of either their head and shoulders or their trunk. Failure of the degenerating segment forward in the sagittal plane is thought to be more common due to the kyphogenic mechanics of posterior placement of the spine in the body; however, decompensation of the spine can also occur in the coronal plane with loss of balance laterally. Junctional degeneration is evaluated with orthogonal radiographs scrutinized for gross evidence of degeneration (arthrosis, disk height loss) and excessive angulation at the segment in question (measurement greater than 10°) [22, 35]. Typical management of added on segments or mild junctional degeneration involves extension of the posterior fusion to include the involved segments. For advanced junctional degeneration associated with central or neuroforaminal stenosis at the effected level, decompression and fusion of the level is required.

Functional losses may be the most difficult symptoms for the physician and patient to quantify and identify as symptoms of deformity progression or failure of an index spinal deformity surgery. Complaints may include increasing leg tiredness with walking associated with degeneration and spinal stenosis below an index fusion or can be as subtle as an increased percent inspired fraction of oxygen (FiO_2) requirement in a young patient with baseline pulmonary compromise that is being worsened by crankshaft phenomenon. Maintaining vigilance to the complaints reported by the patient and correlating these with observations made on imaging studies will often lead to expedient diagnosis of the pathology. A low threshold should be maintained for obtaining multidisciplinary consultations to further evaluate vague clinical functional losses including pulmonology, cardiology, gastroenterology, and physiatry/physical therapy.

35.2.3 Diagnosis

Complete evaluation of the patient requires a thorough history and physical examination characterizing the complaint and its chronology as described above and supplemental testing including imaging, routine laboratory workup, and possibly specialized testing. Orthogonal radiographs of the entire spine are a requirement and can be supplemented with oblique views, coned down views of specific areas of concern, and special views (e.g., Ferguson's view of the lumbosacral junction). Stress views may be added to assess for motion within a suspected area of pseudarthrosis or to assess the flexibility of a segment being evaluated for revision posterior fusion in the setting of "adding on." If further delineation of bony architecture is required, a computerized tomographic (CT) scan of the area of interest is the study of choice. To evaluate the neural canal and foramina outside of the index fusion, magnetic resonance imaging (MRI) is the study of choice, but due to interference of the magnetic field from instrumentation, evaluation of the neural canal within the instrumented segment is best done with a CT-myelogram. Evaluations for spinal infection can include tagged white blood cell scans, but increased sensitivity and specificity are obtained for spine infection with the combination of gallium and bone scans [34]. Routine blood work should be checked, and additional tests should be considered in the setting of very thin patients (prealbumin, transferrin, and total lymphocyte count), patients with potential metabolic bone problems (vitamin D level, NTX-1, osteocalcin, and bone-specific alkaline phosphatase), and if infection is suspected (erythrocyte sedimentation rate, c-reactive protein, and surveillance peripheral blood cultures). If a thoracic fusion is needed, and particularly if entry into the chest is required, pulmonary function testing should be performed, along with obtaining a pediatric pulmonology consultation. Additional pediatric multidisciplinary consultations should also be obtained as indicated.

Ultimate diagnosis for the index surgery failure may be obvious from the initial evaluation or may be only evident after review of the clinical

data including operative notes, serial radiographs, and notes documenting the postoperative clinical course. A critical appraisal of the diagnosis should be completed to be certain that the clinical complaints, testing, and other data are explained by the proposed etiology in their entirety. If symptoms or observations are not explained by the diagnosis, then alternate etiologies or combinations of problems should be considered. An example could be a patient with obvious crankshaft phenomenon that is also experiencing significant pain symptoms, or who has a fever, who may have a concurrent pseudarthrosis or deep infection. Suspicion for complete and accurate diagnosis should be maintained to optimize preparedness for the operating room and to minimize "surprises" that may adversely affect the level of care delivered. As much as is possible, the revision spine surgeon must be prepared for unrecognized pseudarthrosis or other issues in an effort to limit extended operating times and accompanied blood loss that are frequently encountered with revision surgery and to mitigate the elevated neurological risks of revision spine surgery evidenced in the recent publication of the Scoliosis Research Society Morbidity and Mortality database [44].

35.3 Goals for Treatment and Surgical Considerations

Goals for revision deformity surgery vary slightly depending upon the specifics of the revision surgery required. All revision surgeries will have the goals of stable reconstructions that lead to solid fusions, physiological coronal and sagittal balance postoperatively, and minimization of risks for curve progression, transfusion, neurological injury, infection, pulmonary problems, and future surgical requirements. In the setting of patients presenting with pain symptoms, an obvious goal is for relief of discomfort; this is predictable in the setting of pseudarthrosis or symptomatic instrumentation, but is less predictable when the pain generator is not obvious, such as in the patient with pain in the setting of "adding on" of deformity below or above an index fusion. Patients and family members should be counseled about the reasonable expectation for achieving postoperative goals in language that they are able to understand, to avoid dissatisfaction with surgical results in the postoperative period.

A special revision surgery situation involves the conversion of patients with growth-friendly implants that get converted to definitive fusions. These patients may have impressive kyphosis at the upper instrumented segment or in the non-instrumented segment of spine proximal to the upper fixation, multiple levels of scattered ankylosis and spontaneous fusion, as well as fixation points that have partially or completely lost ability to purchase in the vertebrae. Depending upon the amount of flexibility remaining in the spine when the implants are removed, and the magnitude and rigidity of the deformity present, the revision surgeon may need to consider "softening" procedures such as posterior column osteotomies to be described below, or possibly staging the definitive instrumentation and fusion procedure to allow interval halo traction and gradual correction of the deformity. Spine fixation may also be very challenging in the setting of altered bony landmarks or obliteration of normal landing sites for implants (i.e., deficient laminae and transverse processes following hook/claw pull-out, or pedicle wall deficiencies after pedicle screw fixation failure). If the revision surgeon has access to image-guided surgical instruments, this may be helpful; otherwise, it may be very beneficial to either consider performing small laminotomies to allow palpation of the pedicles to improve the safety of placing pedicle screws or to consider adding levels proximally or distally in the construct to ensure that adequate fixation is achieved. In the setting that pedicle screw and hook placement is simply not safe, then keep in mind the use of wires, cables, and newer mersilene tape options to segmentally attach the spinal laminae to the fusion rods. For rare patients with very rigid deformity that does not correct with the posterior column osteotomies or techniques mentioned here, the revision surgeon may need to consider 3-column osteotomies, which will be described below. Finally, as has been stressed throughout this chapter, keep in mind that these

children have been through many procedures, elevating their risk for infection, so consider sending surveillance cultures when doing the definitive instrumentation and fusion procedure.

Surgical considerations are different from case to case and are dictated by both the nature of the index procedure and etiology of the surgical failure. General strategies for revision fusions with correction include extension of posterior spinal fusion, revision of index fusion with or without posterior osteotomies, and revision anteroposterior spinal fusion. Posterior osteotomies include fracture of the posterior fusion mass for minor corrections (Smith-Petersen osteotomy) [53], resection of a posterior wedge of bone through all 3 spinal columns for major corrections (pedicle subtraction osteotomy) [7, 40], and resection of segments of the entire vertebral column from a posterior-only approach for debridement and decompression in addition to major deformity correction (posterior vertebral column resection) [5, 37]. Anteroposterior procedures are indicated for anterior spine growth arrest in the setting of crankshaft phenomenon, when prior anterior fusion was used but residual deformity is not acceptable, and when mobilization of a stiff spine is required for optimal correction [2, 6, 15, 23].

35.3.1 Spinal Osteotomies

Smith-Petersen osteotomies (SPOs) are performed by completing resections of the posterior fusion mass, sparing the lamina at the level of the transverse processes and pedicles [53]. Ponte osteotomies (POs) are similarly performed by resecting the facet joints, ligamentum flavum, and a portion of the lamina at one or more levels in spine that has not previously undergone fusion and achieve spinal mobilization similar to the SPO [16]. Revision fixation can then be obtained with hooks, wires, or pedicle screws into desired segments. Deformity correction is accomplished through relative translation, derotation, and compression/distraction of the vertebrae with reference to one another. Mean correction expected from a single SPO/PO is approximately 10° and is dependent upon the amount of bone resected from the prior fusion and the amount of mobility present in the intervertebral disk at the level being corrected [11]. Patients who have had prior anterior fusion at a particular spinal segment are not candidates for SPOs for deformity correction of that level at revision, but can have SPOs/POs performed proximal or distal to an anterior fusion mass if SPOs/POs will help to level the lowest instrumented vertebra or otherwise provide spinal balance.

35.3.2 Pedicle Subtraction Osteotomy

Pedicle subtraction osteotomies (PSOs) are performed through a posterior-only approach and compromise a complete transection of the bony spinal column through the vertebral level chosen [7, 40]. They are designed to remove a wedge of bone that will result in correction of the spine in one or multiple planes after apposition of the osteotomy site [8]. Ideally, the anterior cortex of the vertebral body involved in the osteotomy is left intact and acts as a hinge for the closing wedge resection. Laminotomies are performed proximally and distally to the osteotomized level, in order to allow the dura and contents adequate room to reposition after the osteotomy is closed. Rigid fixation during and after the osteotomy is essential to prevent spinal translation and neural injury. The osteotomy gets its name from the typical planes that are used in the bony posterior resection wedge, just above and below the level of the pedicles, which results in resection of the pedicles. Single-level PSOs result in corrections of approximately 30°, but can be modified to provide more or less correction by the size of the posterior wedge of bone removed [11]. Ultimate limitation of correction is limited by the posterior height of the vertebral body and the desire to preserve the disk integrity above and below the osteotomized level. Corrections in sagittal, coronal, or complex planes can be accomplished through asymmetric bone removal from side to side, effectively rotating the closing wedge from the anteroposterior plane toward one side or the other. PSOs are 3-column osteotomies, with

posterior or posterolateral shortening, and therefore do not depend upon the mobility of the intervertebral disks for the correction obtained.

35.3.3 Posterior Vertebral Column Osteotomy

Posterior vertebral column resection (PVCR) is similar to the pedicle subtraction osteotomy in that there is a complete resection of the spinal column, but differs in the exposure required, and extent of bone resected [5, 37]. The concept of PVCR is to complete a total vertebrectomy through a costotransversectomy approach (for the thoracic spine), allowing deformity correction and anterior decompression or debridement of the level(s) resected. It requires rigid fixation of the vertebral column during the resection and afterward, and the anterior spine requires reconstitution with a titanium mesh cage, or the equivalent, packed with bone graft [5, 13]. PVCR is a very powerful technique for focal correction of deformity and renders the spine reducible in coronal, sagittal, and axial planes.

Appropriate clinical use of these techniques requires the surgeon to critically assess the revision surgical problem and the structural changes required in the spine to obtain optimal results. For example, in the patient with crankshaft phenomenon, a PSO or PVCR could result in correction of deformity, but neither of these address the anterior vertebral overgrowth driving the deformity; a better option is multilevel anteroposterior revision surgery. For the patient with short-segment (focal) deformity, multiple-level SPOs can be attempted to provide global correction of balance, with lower expectation of correction of the focal deformity. Intraoperatively, if further correction of the focal deformity is desired, then a PSO can be added to the procedure with the posterior closing wedge osteotomy providing the focal deformity correction. Similarly, if the PSO is not able to obtain the amount of correction desired, then the osteotomy can be converted to a PVCR to maximally mobilize the spine for deformity correction. Use of the posterior osteotomies in a step-wise manner allows a "trial" of the less complex techniques to be attempted prior to exposing them to the increased potential for morbidity associated with the complex osteotomy procedures.

35.4 Author's Experience and Case Examples

In the period between 1995 and 2000, the senior author (OBA) treated 28 pediatric patients requiring revision spine deformity surgery. There were 9 boys and 18 girls with a mean age at revision surgery of 13 years (range 4–18 years). Twelve patients had diagnosis of congenital scoliosis, 8 with adolescent idiopathic scoliosis (Fig. 35.1), four patients with neuromuscular scoliosis (two with muscular dystrophy and one with cerebral palsy), and two each with Marfan's syndrome (Fig. 35.2) and post-laminectomy kyphosis (tumor resections). Index operations were instrumented posterior spinal fusions in 17, instrumented anteroposterior spinal fusions in three, instrumented anterior fusion in one, non-instrumented anteroposterior fusions in four, and posterior laminectomy in three. Each patient had a mean of three surgeries prior to the revision deformity surgery (range, 1–12 prior surgeries). All patients had increasing spinal deformity, and 10 patients had instrumentation-related problems, including three with broken rods. Spinal deformity encountered included crankshaft phenomenon in 11 patients, progressive kyphosis in six (Fig. 35.3), and pseudarthrosis in three. Pain was a major determinant of surgical indication in seven patients.

The type of revision surgical intervention was determined on a case by case basis, with 25 patients receiving anteroposterior fusions and three being treated with posterior spinal fusions. Instrumentation was used in all cases. Surgical revision for the eleven crankshaft phenomenon patients was anteroposterior fusion, as dictated by their disease. The pseudarthrosis patients were also managed with anterior spinal fusion to best guarantee solid arthrodesis across the segment and posterior instrumented fusion for correction and compression across the segment. Additional anterior procedures were indicated for osteotomy of unacceptable residual deformity and to mobi-

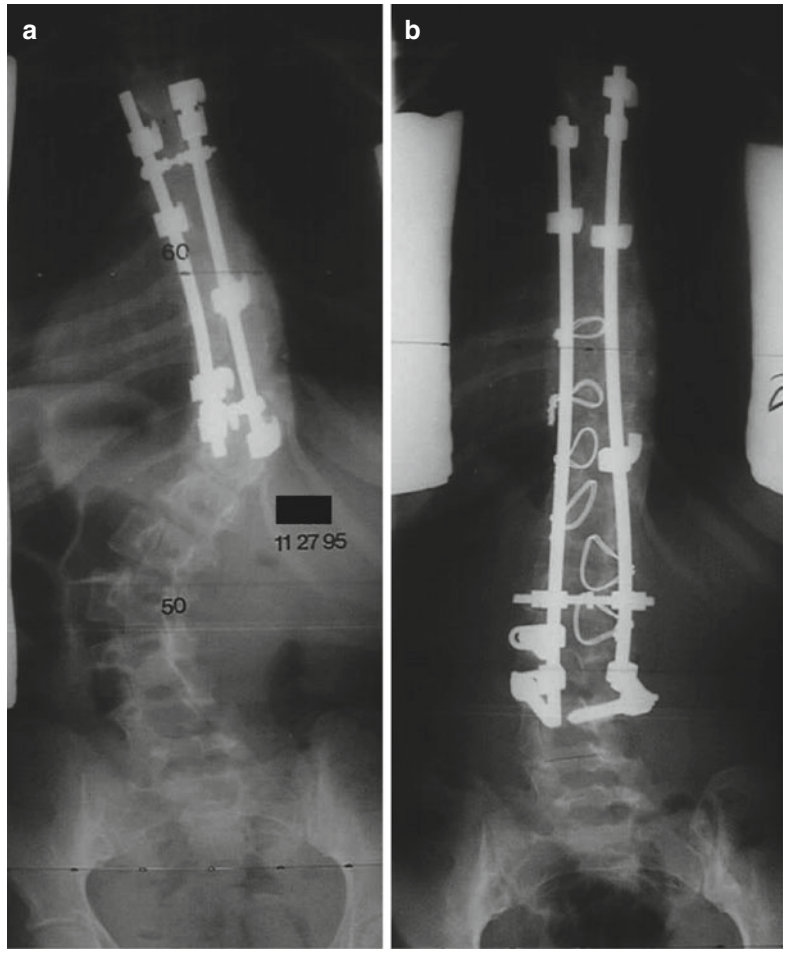

Fig. 35.1 A 17-year-old female with painful "adding on" below prior posterior selective thoracic spinal fusion for adolescent idiopathic scoliosis. Following the index surgery, she experienced progressive spinal deformity and pain symptoms throughout the spine and into the lower extremities that interfered with her ability to sit, stand, or walk for more than a few minutes. She was neurologically intact and otherwise very healthy. (**a**) A anteroposterior radiograph taken prior to the revision deformity surgery demonstrating a right thoracic curve of 60°, a left lumbar curve of 50°, and instrumentation T4–T11. Diagnosis of "adding on" below the prior fusion was made, and the patient was indicated for revision surgery for removal of instrumentation, posterior osteotomies (Smith-Petersen distally) to allow correction, and distal extension of posterior instrumented fusion to L4. The patient underwent the planned procedure with use of an ISOLA hybrid construct and distal pedicle screws, along with right-sided thoracoplasties and iliac crest bone graft harvest. By two months after the revision surgery, she required no pain medicine, and preoperative symptoms had resolved. (**b**) A 6-month postoperative anteroposterior radiograph showing revision instrumentation and deformity correction to 25° in the thoracic and 20° in the lumbar spines. She remains symptom-free in long-term follow-up and is managed by a physician located closer to her home

lize the spine for improved correction. Standalone posterior spinal fusions were performed as a simple extension in one patient and were combined with posterior osteotomies in two patients. Postoperative correction in the coronal plane was from 67° (range, 32–115°) preoperatively to 36° (range, 20–60°) postoperatively, for a mean correction of 43 % (range, 17–66 %). Sagittal measurements in the thoracic spine had a mean of 57° (range, 20–107°) of kyphosis preoperatively and 51° (range, 20–95°) postoperatively and in the lumbar spine were 66° (range, 40–106°) of lordosis preoperatively and 60° (range, 40–88°) postoperatively.

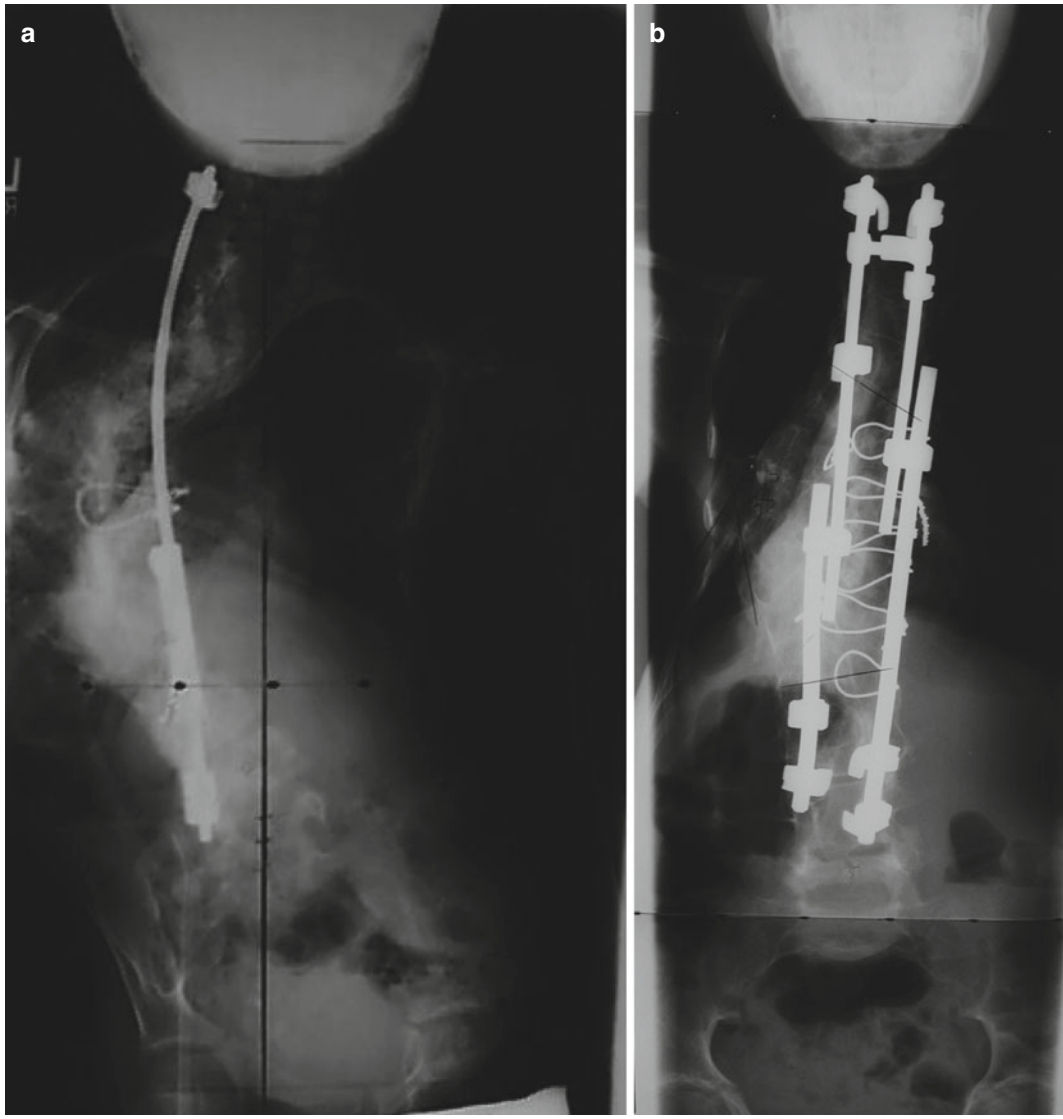

Fig. 35.2 A 7-year-old female with Marfan's syndrome diagnosed with scoliosis at 3 years old, who had undergone bracing and growing rods and then had loss of proximal implant fixation and crankshaft phenomenon after definitive fusion. Revision operative management was slightly delayed due to her emaciated state and need for nutritional optimization by gastroenterology consultation, along with placement of a gastrostomy tube and enteral supplementation. (**a**) An anteroposterior radiograph taken prior to the revision surgery showing loss of proximal fixation, main thoracic apex left curve from T5 to T12 measuring 128° and a 35° curve from T12 to L5. After application of preoperative traction, the T5–T12 curve was reduced to 108°. She had global kyphosis of 60°, was Risser 0, and had 3-dimensional imaging demonstrative of dural ectasia. She was indicated for revision anteroposterior fusion with posterior instrumentation and underwent anterior osteotomies from T5 to T12 and a staged posterior procedure. During the second-stage posterior procedure, multiple Smith-Petersen osteotomies were performed, and an ISOLA four-rod construct was used to instrument from T2 to L4 with claw and wire fixation. (**b**) A postoperative anteroposterior radiograph showing the revision instrumentation and correction of deformity with residual thoracic scoliosis of 40°. At most recent follow-up seven years after the revision surgery, she had gained 8 in. in height and 24 lb in weight and was pain-free and neurologically intact

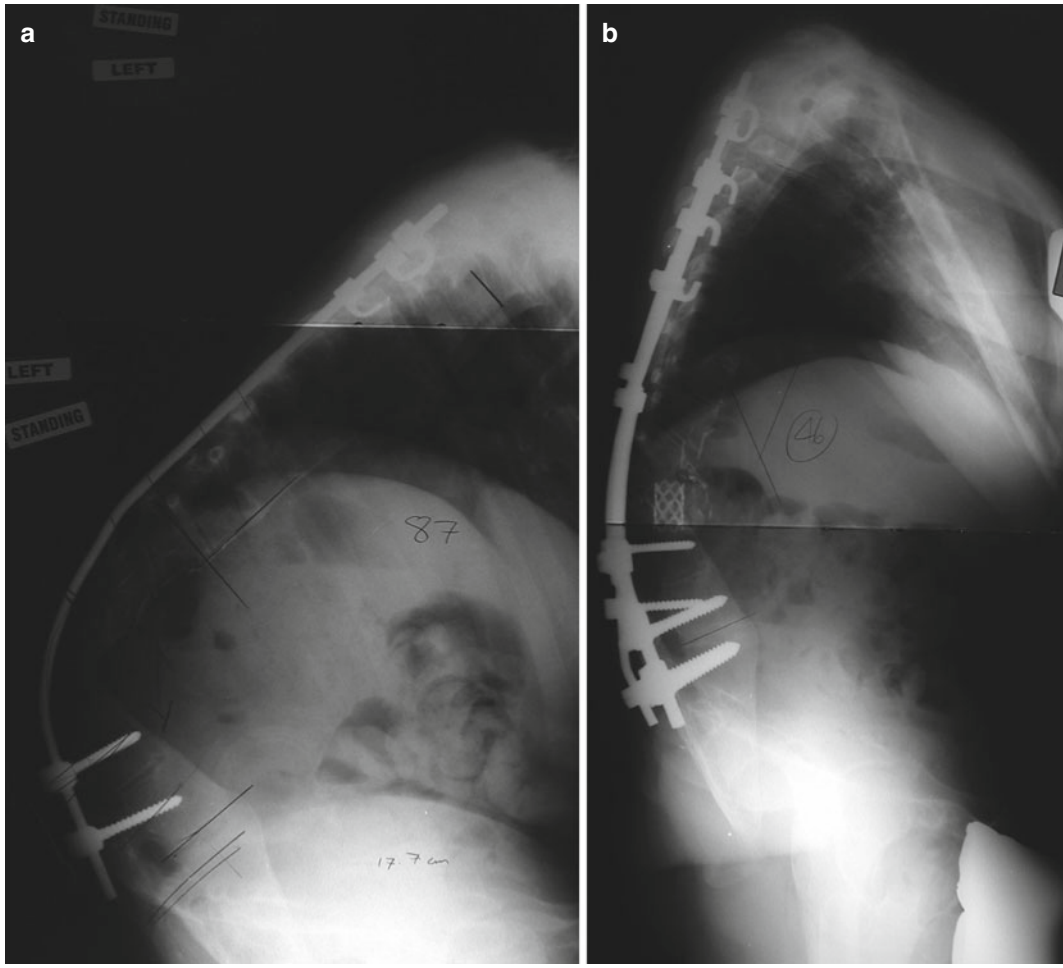

Fig. 35.3 A 13-year-old female with progressive kyphosis after "growing rods." She was diagnosed with neurofibromatosis type 1 at age 2 years and began growing rod management at age 3.5 years. She had 11 surgeries prior to presenting to our institution, the last of which involving infected implants, requiring implant removal and deformity management with a TLSO appliance. (**a**) A standing lateral radiograph taken prior to the placement of halo traction showing T3–L1 kyphosis of 125° with apex at T8/T9. She underwent a staged procedure where a halo fixator was applied; she had 3 months of axial traction (50 % of body weight), where the T3–L1 kyphosis decreased to 100°; and she underwent T2–L3 posterior spine fusion with instrumentation, including a T8–T10 posterior vertebral column resection. (**b**) A standing lateral radiograph taken at 1 year postoperatively following PSFI with PVCR demonstrating revision posterior instrumentation, anterior titanium mesh cage, posterior titanium mesh connected to the rods preventing soft tissue encroachment on the neuroelements, and overall kyphosis measurement of 55°. She had physiological coronal and sagittal balance, was neurologically intact, had no pain symptoms, and had returned to participation in her school play

Major complications occurred in three patients and included one superficial wound dehiscence, one pleural effusion/pneumothorax, one junctional kyphosis, and one proximal hook implant dislodgement. Each of these required invasive intervention, and each patient eventually fully recovered. No events of neurological deficit, deep infection, or death occurred. No patients required placement of tracheostomy, despite nine patients having preoperative forced vital capacity averaging 30 % of predicted (range, 20–40 %). None of the 28 patients have required further spine surgery.

Conclusions

Revision spine surgery in the growing child is technically challenging and not encountered

frequently except in growing rod patients. In approaching this complex problem, the surgeon must have a complete understanding of the primary deformity and reason for index surgery failure, comprehend the residual growth potential of the spine and how this affects the revision surgery plan, and have a realistic appreciation of technical ability required to perform the procedures safely. Remember: first do no harm, and if in doubt, do not hesitate to refer the patient to a more experienced surgeon or to ask for help. Indications and goals for revision surgery need to be agreed upon by the surgeon and the family of the child to assure that postoperative satisfaction is maximal, and preoperative planning must include a multidisciplinary evaluation to optimize both care delivered and surgical outcome.

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Instrumented Fusion in Early-Onset Scoliosis

36

Lawrence G. Lenke

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Key Points

- There are rare indications for fusion in patients with early-onset (infantile and very young juvenile) idiopathic scoliosis, except for considering periapical fusions, along with some form of growing instrumentation for those with >90–100° of scoliosis.
- Congenital spine deformities that are amenable to short resection and realignment procedures can often be best treated with concomitant short-segment instrumentation and fusion.
- Patients with actual/impending spinal cord neurological deficit should undergo appropriate spinal decompression along with instrumentation and fusion across the resected region, regardless of age.
- Although the goal should always be to obviate/minimize spinal fusion in the skeletally immature patient, there are still many patients/conditions were either a short or more extensive fusion may be in the best interest of the patient's overall health and development. Individual assessment and treatment will always be essential in this patient population.

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36.1 Introduction

The control of progressive spinal and chest wall deformity in young children is an extremely difficult problem. Intuitively, gaining control of a spinal deformity and limiting progression while maximizing the ultimate growth potential of the entire spinal column and rib cage would be beneficial. Thus, much effort has been expended over the last 15–20 years to apply various growth modulation techniques in order to avoid spinal fusion in young and highly skeletally immature children. However, there are circumstances where fusion may be the best option for these patients, and these limited but extremely important indications will be highlighted in this chapter. In addition, potential long-term consequences of spinal fusion in the young child will be discussed.

36.2 Patient Characteristics

Historically, the chronological separation of early-onset scoliosis (EOS) fell into two categories: *infantile* from birth to 2 years and 11 months and *juvenile* from the age of 3–9 years and 11 months. There was also an early and late onset scoliosis category which lists the age of 6 years as the distinction between these two groups [1]. More recently, the Growing Spine Study Group and Children's Spine Study Group have recommended that, based upon natural history and treatment, 10 years of age is a logical differentiation between early and late onset scoliosis. They defined EOS as "scoliosis of any etiology developing before the age of 10." Both the Scoliosis Research Society and Pediatric Society of North America have supported this view [2, 3]. As we know that spinal fusion limits axial spinal column growth and ultimate height, the younger the patient, the more detrimental the spinal fusion. In addition, as impeded spine growth also limits trunk and rib cage growth, fusion has a potentially significant detrimental effect on pulmonary development, chest volume, and function. We know that alveolar growth is rapid and continues until the age of 8 years; however, ultimate pulmonary function increases until the age of 18 years primarily due to the increased thoracic cavity volume [4]. Thus, every effort should be made to maximize spinal growth in the thoracic region, ultimate spinal column height, and chest wall and rib cage alignment in the very young child. In later juvenile years, this becomes especially important for males versus females, as males tend to develop later and their spine growth often continues until the late teenage years.

Besides chronologic age issues, patient diagnosis becomes extremely important as well. Although idiopathic early-onset scoliosis (IEOS) is the prototype diagnostic category for the young child with a spinal deformity, there are also quite a number of other diagnoses seen in children with EOS and progressive spinal deformity. These include congenital spine and/or rib deformities, various types of genetic syndromes such as neurofibromatosis, connective tissue disorders such as Marfan and Ehlers-Danlos syndromes, bone dysplasias such as Hurler's syndrome, neuromuscular disorders such as cerebral palsy and myelodysplasia, and those that fall under the "exotic" category of unusual spinal abnormalities and rare diagnostic categories such as prune-belly syndrome.

36.2.1 Idiopathic Early-Onset Scoliosis

In those patients who present with a spinal deformity under the age of 3 years, there are very few indications for the consideration of performing fusion surgery in these very young patients with otherwise idiopathic curves [5]. Today, many surgeons avoid any type of apical fusion in patients with rather severe IEOS deformities; however, in the past, this was a viable option and may still be so today [6]. However, this should be reserved for patients with curves over 90–100° and in very selective and individualized circumstances (Fig. 36.1a–h). If surgery is contemplated in an IEOS patient with a severe deformity, we strongly recommend a period of halo-gravity traction prior to any surgical intervention for maximal spinal correction as these patients have very small vertebrae and relative osteoporosis due to

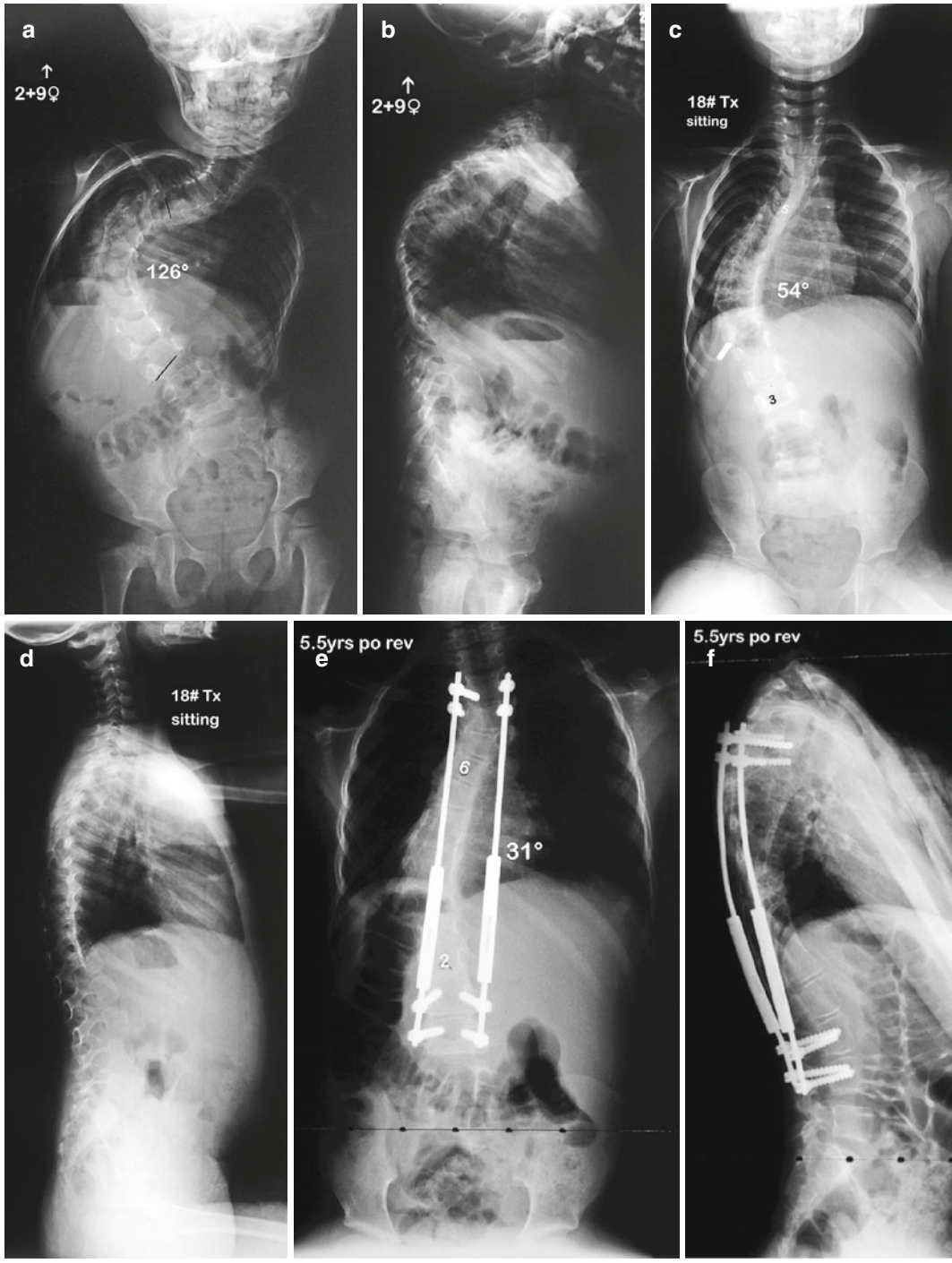


Fig. 36.1 Patient is a 2 + 9-year-old female with infantile idiopathic scoliosis. (**a**, **b**) She presented with a 126° scoliosis and thoracic kyphosis deformity. She had a normal total spine MRI and a negative genetics evaluation. She was placed in preoperative halo-gravity traction and underwent a five-level anterior apical release and fusion. (**c**, **d**) She then had further traction reducing her scoliosis to 54°. (**e**, **f**) Her 5.5-year postoperative x-rays show good maintenance of deformity correction. (**g**, **h**) Her preoperative and postoperative clinical photographs demonstrate her much improved appearance

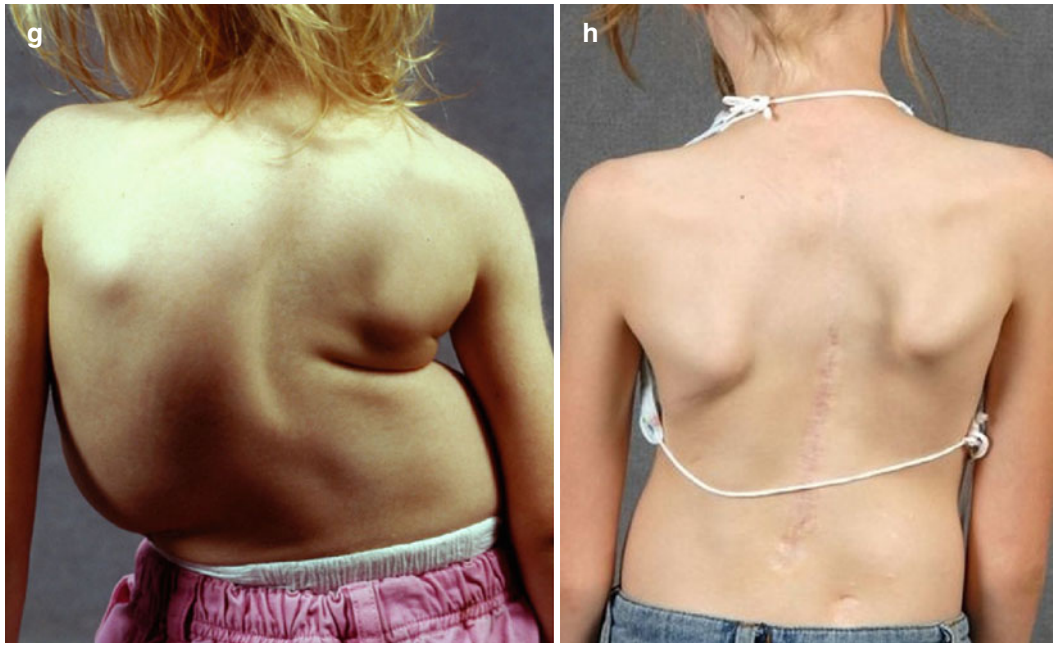


Fig. 36.1 (continued)

their small stature. Following several weeks of halo-gravity traction, a circumferential apical fusion of 3–5 levels may be performed with the posterior application of some type of growing rod construct covering the full extent of the scoliosis deformity. The goal of treatment should be continued growth of the areas cephalad and caudad to the apex following consolidation of the apical fusion. It is important to minimize the number of thoracic levels fused for reasons mentioned earlier, especially with reference to the pulmonary system and development. One can argue that a posterior-only growing rod or a chest wall growing system would be a better option for these patients. However, in patients that have a severe deformity where the rotated apex nearly touches the lateral rib cage, the risk of crankshaft phenomenon with posterior-only application of instrumentation is still extremely high. Another alternative would be a limited convex anterior hemiepiphysiodesis and a posterior hemiarthrodesis. However, in a severely rotated deformity, it is extremely difficult to know where the true "convexity" of the disk is at the apex. Thus, proceeding to a full growth arrest via a circumferential fusion may be the best option.

As the chronologic age of the patient increases, the indications for spinal fusion with progressive deformity increase as well. Although an EOS patient becomes an adolescent at 10 years of age, performing a spinal fusion for progressive scoliosis in a 9-year-old female would seem very reasonable for many surgeons. Certainly, the physiologic age of the patient needs to be further elucidated by various radiographic assessments, along with the basic knowledge of how tall the parents are in relation to the child (if known). One definite concern with a posterior-only procedure is whether a circumferential approach is needed to help prevent the crankshaft phenomenon in these skeletally immature patients [7]. This concern has led to three surgical techniques: a traditional anterior as well as posterior fusion with posterior instrumentation, an anterior instrumented fusion if feasible, and a posterior fusion utilizing segmental pedicle screw fixation. Although there are no good long-term studies demonstrating the efficacy of this last approach, it is performed in many centers by many surgeons throughout the world, who utilize pedicle screw constructs routinely. For an older EOS patient with a very severe deformity, it is probably best

to consider a circumferential fusion to control the progressive deformity prior to the adolescent growth spurt, rather than attempting to control the deformity during the growth spurt [8, 9]. Ideally, this should be performed after 8 years of age as alveolar development is complete, thus having less long-term detrimental effects on the pulmonary system.

36.2.2 Congenital Spinal Deformities

For patients with isolated congenital deformities, a unilateral, fully segmented nonincarcerated, hemivertebra resection with a short spinal fusion has become the preferred method of treatment. Often a near-complete correction of the congenital deformity can be obtained from a single operation, often performed via a posterior-only approach. The indications for a longer spinal fusion in congenital deformities are more controversial. However, a congenital dislocation of the spine (CDS), with or without neurological deficit, has been a nearly universal indication for spinal fusion as soon as the patient is old enough to tolerate the operation, usually in the first to second year of life. The spinal column is extremely unstable; thus, there is a high risk of spontaneous and/or progressive neurological deficit including full paralysis if left untreated (Fig. 36.2a–f). Traditional methods of treatment have included circumferential fusion with casting followed by brace immobilization. However, we have seen a very high pseudarthrosis rate with that method, so we now prefer to add posterior instrumentation, if at all possible, to stabilize the spinal column during the fusion process. Similar to the abovementioned CDS patient with a severe deformity, those with a severe congenital deformity may be best treated with a posterior-only or circumferential spinal fusion, rather than allowing the deformity to progress into adolescence.

36.2.3 Genetic Syndromes

Unfortunately, many genetic syndromes have musculoskeletal conditions involving progressive spinal deformity. Some of the more common are those involving the connective tissues, such as Marfan disease and Ehlers-Danlos syndrome. In a young patient, this becomes a detrimental combination involving a large deformity and very lax connective tissues. Growth modulation techniques should be utilized in the highly immature patient, but again, once patients are nearer to age 10, an apical if not complete spinal fusion should be considered. If left untreated, these patients may ultimately develop severe, life-threatening deformities because of their poor connective tissue; hence, surgical intervention can produce a secure and stable spinal column.

36.2.4 Neuromuscular Disorders

There is a spectrum of diagnoses involved in neuromuscular spinal deformity. Unfortunately, most patients do not benefit from a prophylactic orthosis, especially when their deformity reaches severe Cobb measurements of over 90–100°. In addition, many of them are not very healthy and thus may not be able to tolerate repeated surgical intervention necessary with treatments such as growing rods. Thus, entertaining a spinal fusion, especially during the EOS years, may be appropriate. One diagnostic category where this is certainly advantageous is with spinal muscular atrophy. These patients may develop a very severe deformity at a very young age and often are not very healthy from a pulmonary standpoint. Thus, it is doubly troubling as not only are they not healthy enough to tolerate an anterior procedure, they are also young enough to have a high risk of crankshaft phenomenon occurring if they undergo a posterior-only arthrodesis. However, this has been our approach, and a minimum 5-year postoperative review of these patients has shown reasonable radiographic and clinical results following early intervention for progressive, severe spinal muscular atrophy scoliosis [6].

Another category where spinal fusion has been performed at a very young age is congenital kyphotic deformities of the lumbar spine and those patients with a high-level myelomeningo-

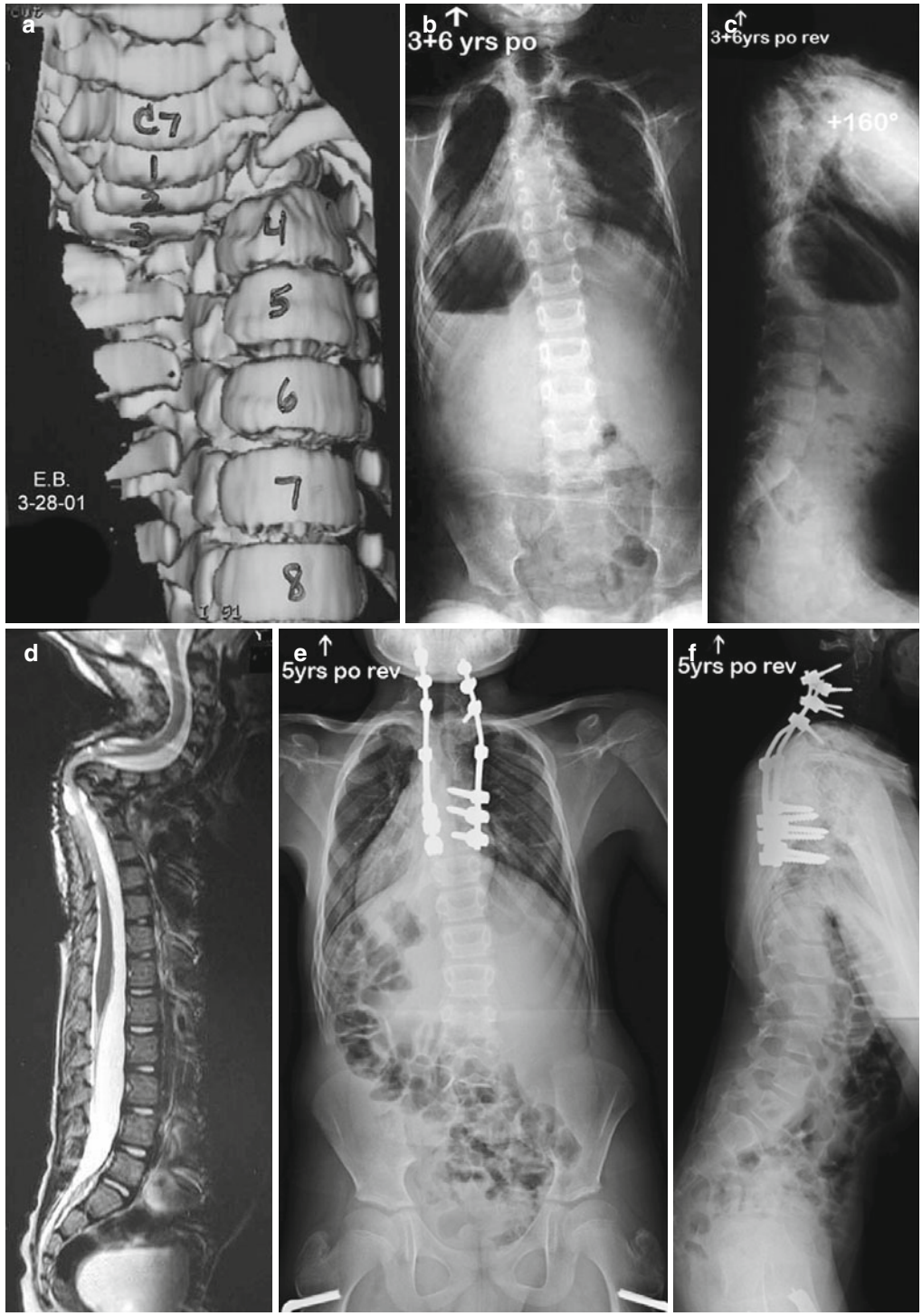


Fig. 36.2 Patient is a 2-year-old boy who presented with a congenital dislocation of T3/T4 as seen on (**a**) his preoperative coronal 3D CT scan. At three years of age, he underwent in situ fusion with subsequent deformity progression and myelopathy. (**b**, **c**) At the age of 5 (3 + 6-years postoperative), his coronal and sagittal x-rays somewhat hid his severe deformity. (**d**) However, on his sagittal MRI scan, the angular deformity of the cervicothoracic region with tenting of the spinal cord is obvious causing myelopathy. (**e**, **f**) He underwent a posterior revision reconstruction and a VCR of T3 and T4 for spinal cord decompression with ultimate midcervical to midthoracic instrumentation and fusion for stabilization of his deformity. His myelopathy completely resolved following decompression

cele. Once a lumbar congenital kyphosis reaches over 100° or so, nothing advantageous is going to happen to that region of the spine; hence, reconstruction via a kyphectomy and spinal fusion is indicated. Most surgeons would attempt to "grow" the middle and upper thoracic spine with implants such as a Luque trolley or other growth-guided techniques such as Shilla, if at all possible, to allow further thoracic growth despite the thoracolumbar and lumbar fusion. It is particularly advantageous to avoid the mid and lower thoracic lordotic deformity that often becomes fixed in response to a progressive lumbar kyphosis below.

Most patients with cerebral palsy with a scoliosis deformity can be observed and braced or undergo wheelchair modification during the EOS years, in order to avoid spinal fusion until adolescence. However, once again, in those with severe, relentlessly progressive deformities greater than 100–125°, continued observation seems inappropriate when so much growth remains. Obviously, if they are healthy enough to undergo multiple surgical procedures, some type of posterior growing rod construct may be advisable. However, many of these patients are frail and debilitated, and a better option might be a posterior spinal fusion utilizing intraoperative halo-femoral traction to maximize correction and sitting balance while minimizing complications [7]. It is essential to explain to caregivers the full ramifications of a spinal fusion at such a young age.

36.3 Salvage Procedures

Unfortunately, many young patients whether treated appropriately or inappropriately experience progressive deformity requiring revision surgery. Some patients treated with growing rods or actual fusion procedures develop severe and progressive deformities that can be worse than the primary deformity. These progressive deformities can be either in the coronal (Fig. 36.3a–k) or sagittal plane or both (Fig. 36.4a–h), in the location of prior short solid fusions resulting in single or multilevel pseudarthroses, which present a unique set of challenges. Their spine is invariably stiff, their pulmonary status is usually quite limited, and they have often undergone multiple surgical procedures either anteriorly and/or posteriorly rendering their spinal column very anatomically distorted. In addition, they have a much higher rate of impending or actual neurologic sequelae from their severe deformity and repeated surgical interventions. Revision salvage procedures must be well thought out and thoroughly performed with the goal being optimal spinal alignment and neurologic function for the rest of the patient's life. The use of halo-gravity traction is often very helpful in preparing patients for a definitive reconstruction, especially in these very young patients (Fig. 36.5a–m). We recently reviewed 44 patients treated with preoperative halo-gravity traction with early-onset spinal deformity less than age 10. We found significant improvement in radiographic parameters. We find halo-gravity traction especially effective for progressive kyphosis particularly in the cervicothoracic region.

One group of disorders notorious for having these types of issues is severe dysplastic neurofibromatosis making these patients very challenging at the outset. Most of these patients, initially treated at a young age, will require circumferential fusion. However, even after surgery, they are at a very high risk for pseudarthrosis and continued progression of their deformity [10]. Because of the number of prior procedures, there is a greater risk of spinal cord devascularization [11, 12]. Prior to revision surgery, we often find halo-gravity traction helpful to maximize gradual correction of their severe deformity, while optimizing pulmonary and nutritional health along the way. Many of these patients have already had anterior multilevel procedures; thus, repeating anterior work is fraught with difficulty either on the operated or unoperated side.

Recently, the use of posterior vertebral column resection procedures has been quite beneficial for these severe revision deformities (Fig. 36.6a–n). Obtaining secure pedicle screw fixation above and below the apex of the deformity followed by resection of one, two, or even three vertebrae from an all-posterior approach has allowed dramatic radiographic and clinical correction. Ultimately, a spinal fusion over the entire length of the posterior instrumentation has

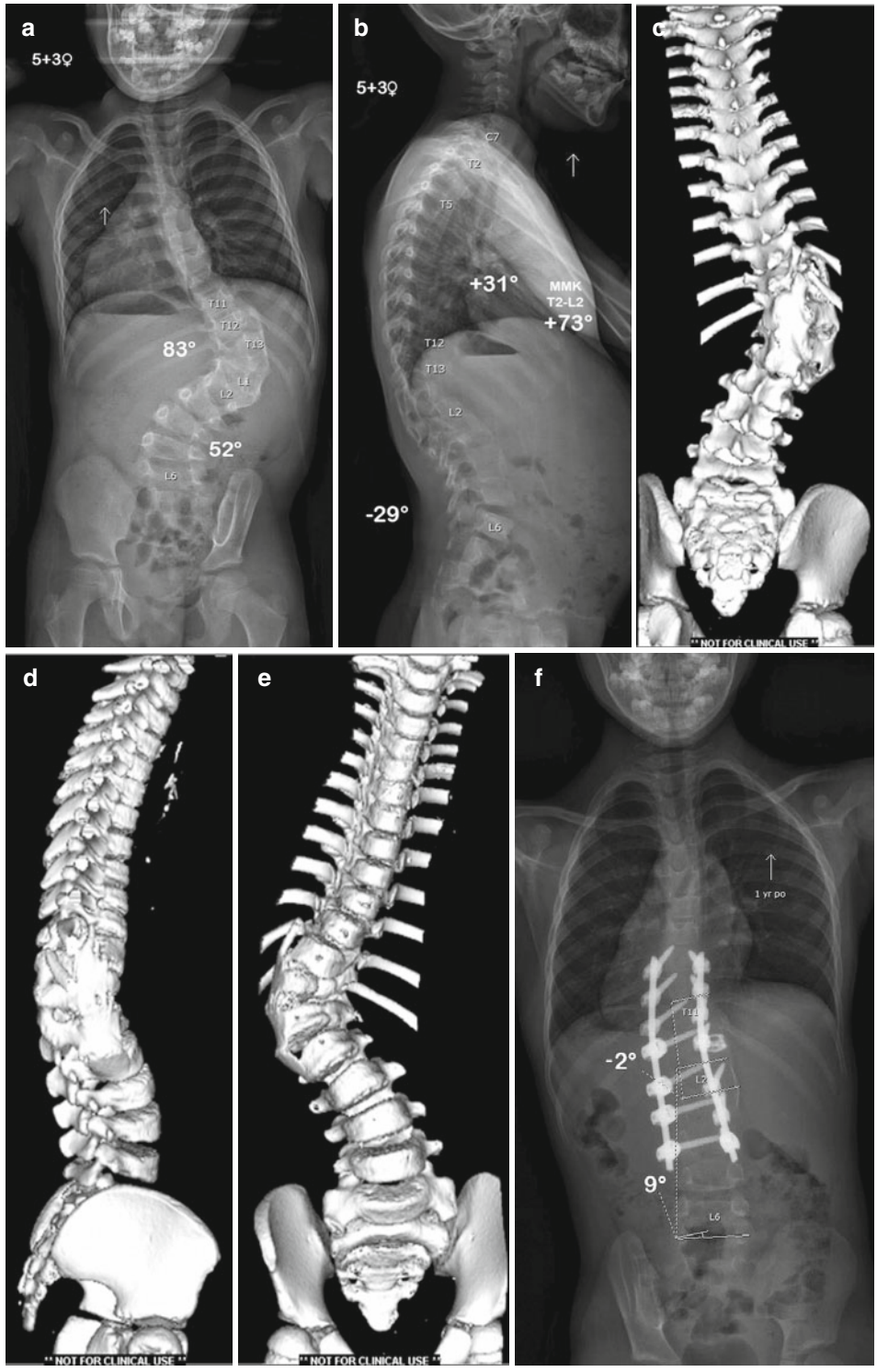
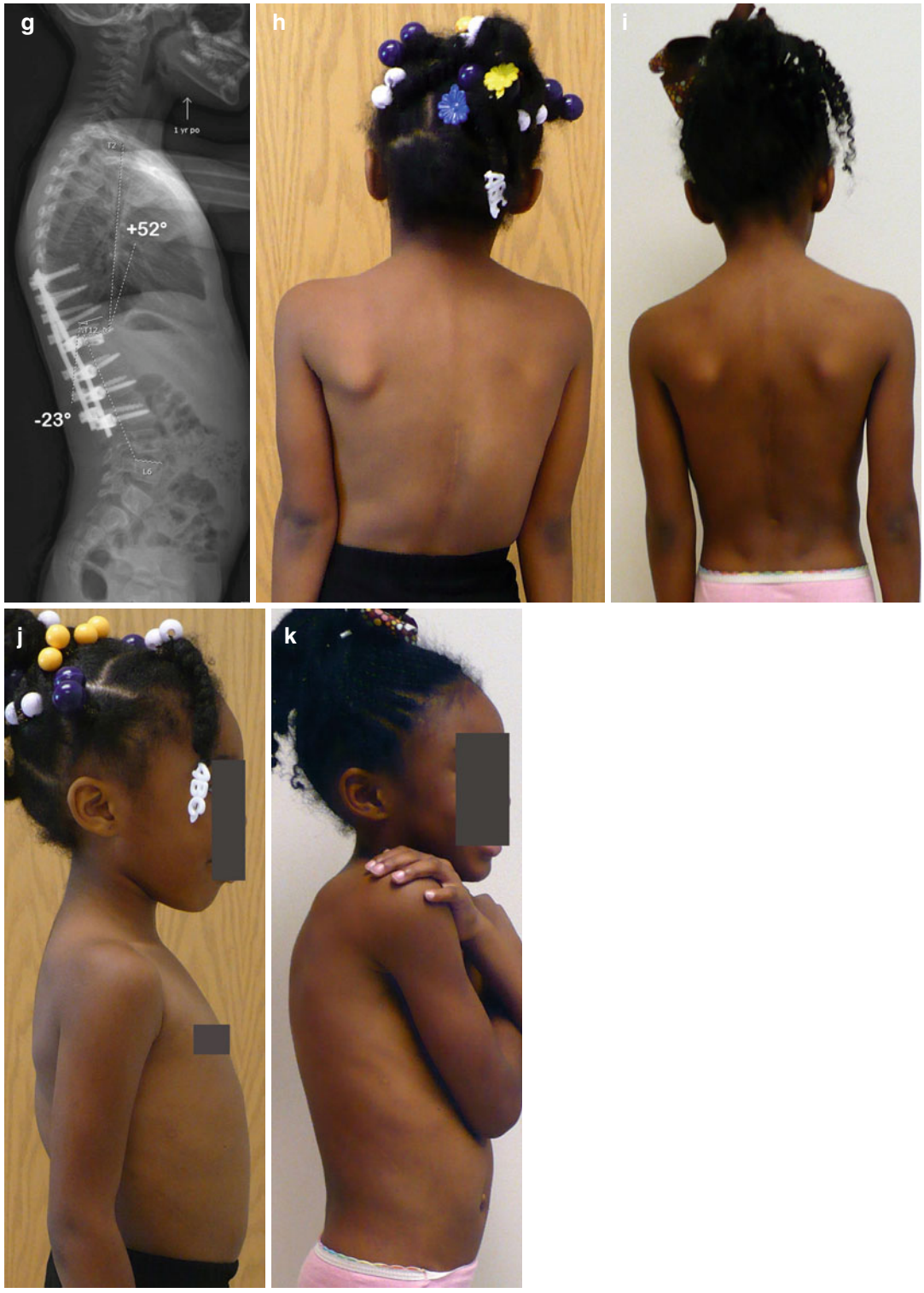


Fig. 36.3 Patient is a 5+3-year-old girl with congenital thoracolumbar kyphoscoliosis. She underwent a short anterior and posterior spinal fusion at another institution. (**a**, **b**) She presented to us with progression of her deformity to an 83° scoliosis and a +73° overall maximum kyphosis. (**c**–**e**) Her preoperative 3D CT scan shows the short posterior as well as anterior fusion. (**f**, **g**) She underwent a posterior revision reconstruction with a single-level apical VCR for realignment of her coronal and sagittal plane deformities. (**h**–**k**) Her preoperative and postoperative coronal and sagittal clinical photographs demonstrate improved truncal alignment



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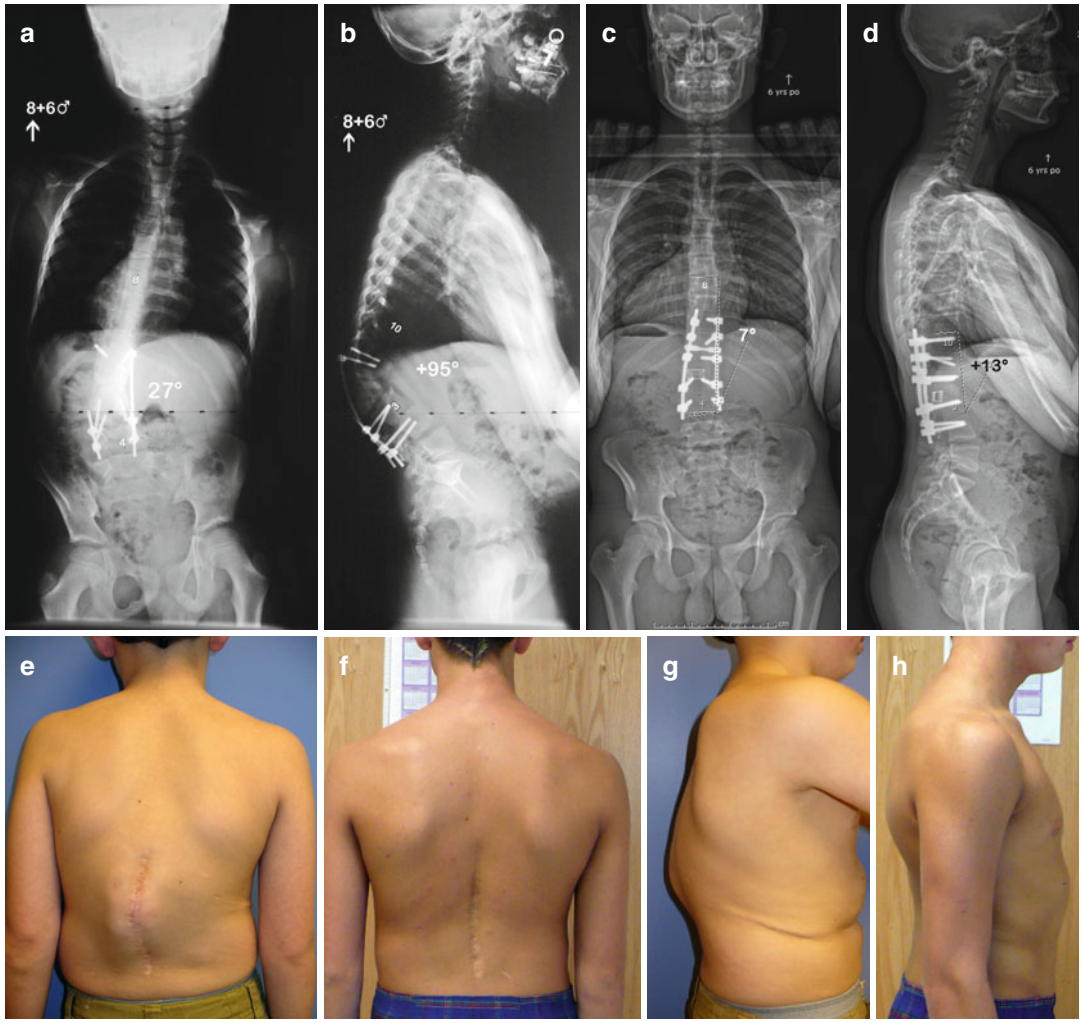
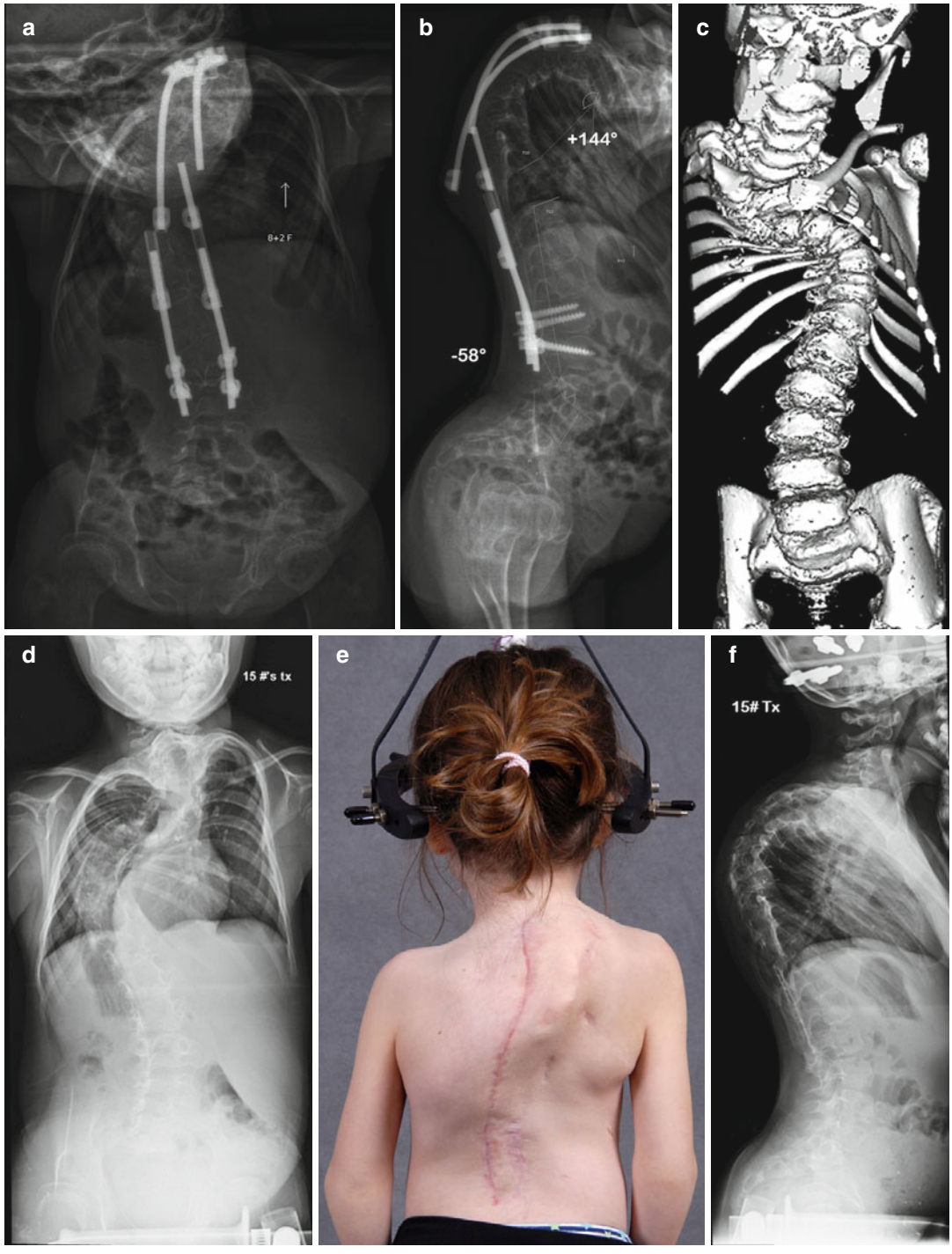


Fig. 36.4 Patient is an 8 + 3-year-old boy with congenital kyphosis treated elsewhere with posterior instrumentation and fusion. (**a**, **b**) He presented to us with subsequent pseudarthrosis and implant failure with a 27° scoliosis and a progressive kyphosis to +95° in the same region as the coronal plane deformity. (**c**, **d**) He underwent a posterior revision reconstruction with a two-level VCR for realignment of his spine. (**e**–**h**) Preoperative and 6-year postoperative clinical photographs show improved truncal alignment in both planes

Fig. 36.5 Patient is an 8 + 2-year-old girl with Desbuquois skeletal dysplasia treated elsewhere with growth-sparing techniques including VEPTR and current growing rod construct placement in an attempt to control progressive severe thoracic kyphoscoliosis. She presented to us with broken implants and subsequent severe progression of cervicothoracic kyphosis. (**a**, **b**) Her coronal and sagittal plane x-rays demonstrated a severe deformity which measured +144° in the sagittal plane. (**c**) The frontal view of her 3D CT scan detailed her severe coronal plane deformity much better than could be assessed on her plain x-ray. (**d**–**g**) Her implants were removed, and she was placed in preliminary halo-gravity traction to improve her radiographic and clinical alignment in preparation for surgery. She underwent a posterior revision reconstruction from C7 to L3 with several posterior column osteotomies to realign her deformity, gaining 4″ in height. (**h**, **i**) At one-year postoperative, she had excellent radiographic realignment of her coronal and sagittal planes. (**j**–**m**) Preoperative and postoperative clinical photographs demonstrate her much improved truncal alignment


a
8+2 F
b
+144°
-58°
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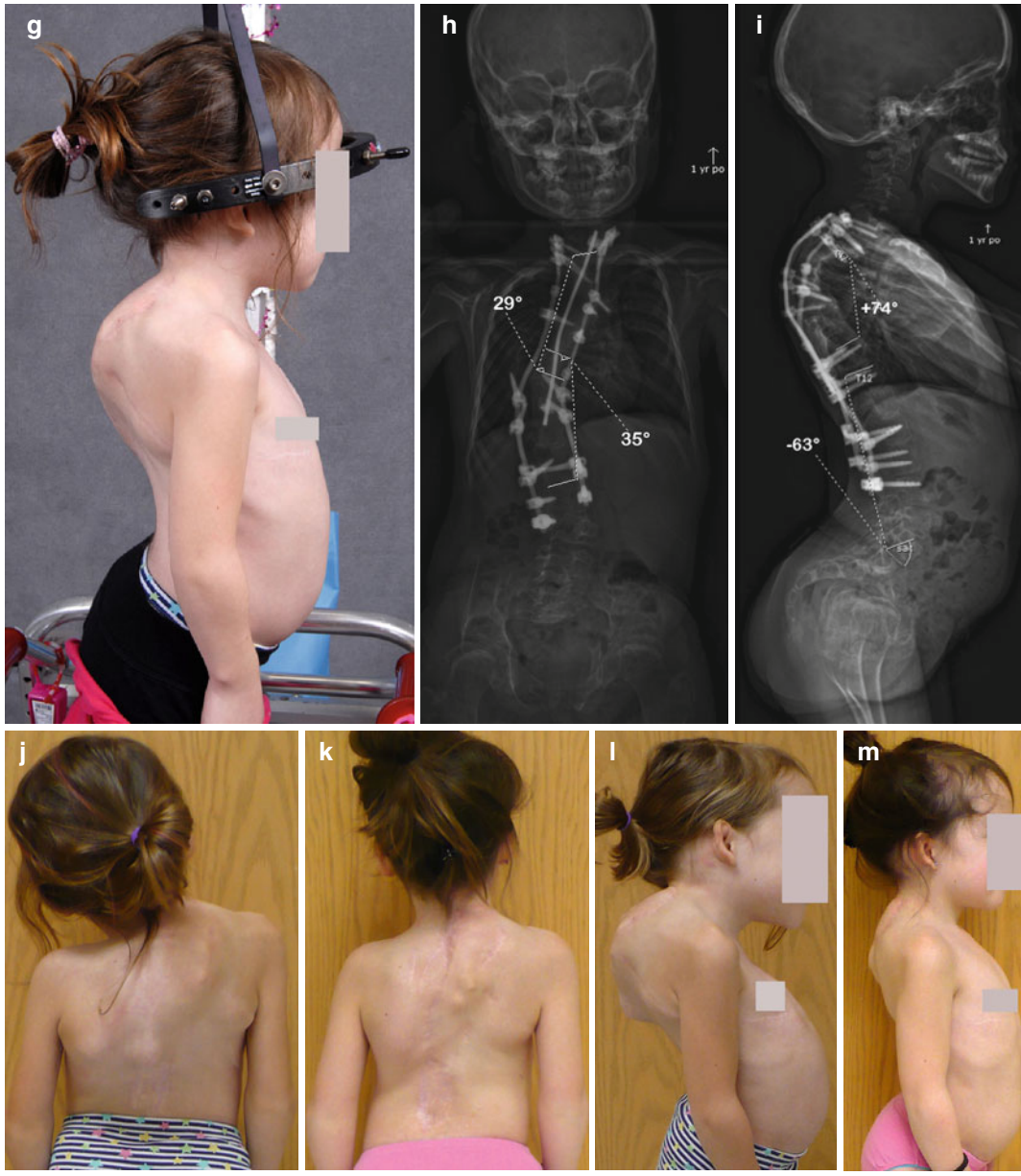

Fig. 36.5 (continued)

to be performed; however, that is usually the best option for these patients. These are very technically demanding operations that require not only the surgeon but the entire surgical team to be adept at very high-level spinal deformity care for successful execution. At our institution, this has revolutionized the care we provide these patients with quite encouraging results. We now have experience in over 210 consecutive posterior vertebral column resections for severe deformities, with 124 of the procedures performed in pediatric patients. We have obtained on average 60 % correction; the majority performed under one surgical setting, with no permanent paraplegia. We attribute this to the standard use of both somatosensory and some type of motor evoked

potential monitoring intraoperatively [11, 12]. There have been some root-level deficits in revision of upper lumbar procedures, but the two occurring in the pediatric revision setting were transient. Only one patient with a completely normal neurologic examination required revision for implant-related issues and/or pseudarthrosis thus far, with over 5-year follow-up available at present for nearly 40 of these patients.

Another rare but nearly universal indication for spinal fusion surgery in a skeletally immature deformity patient is one with impending or actual

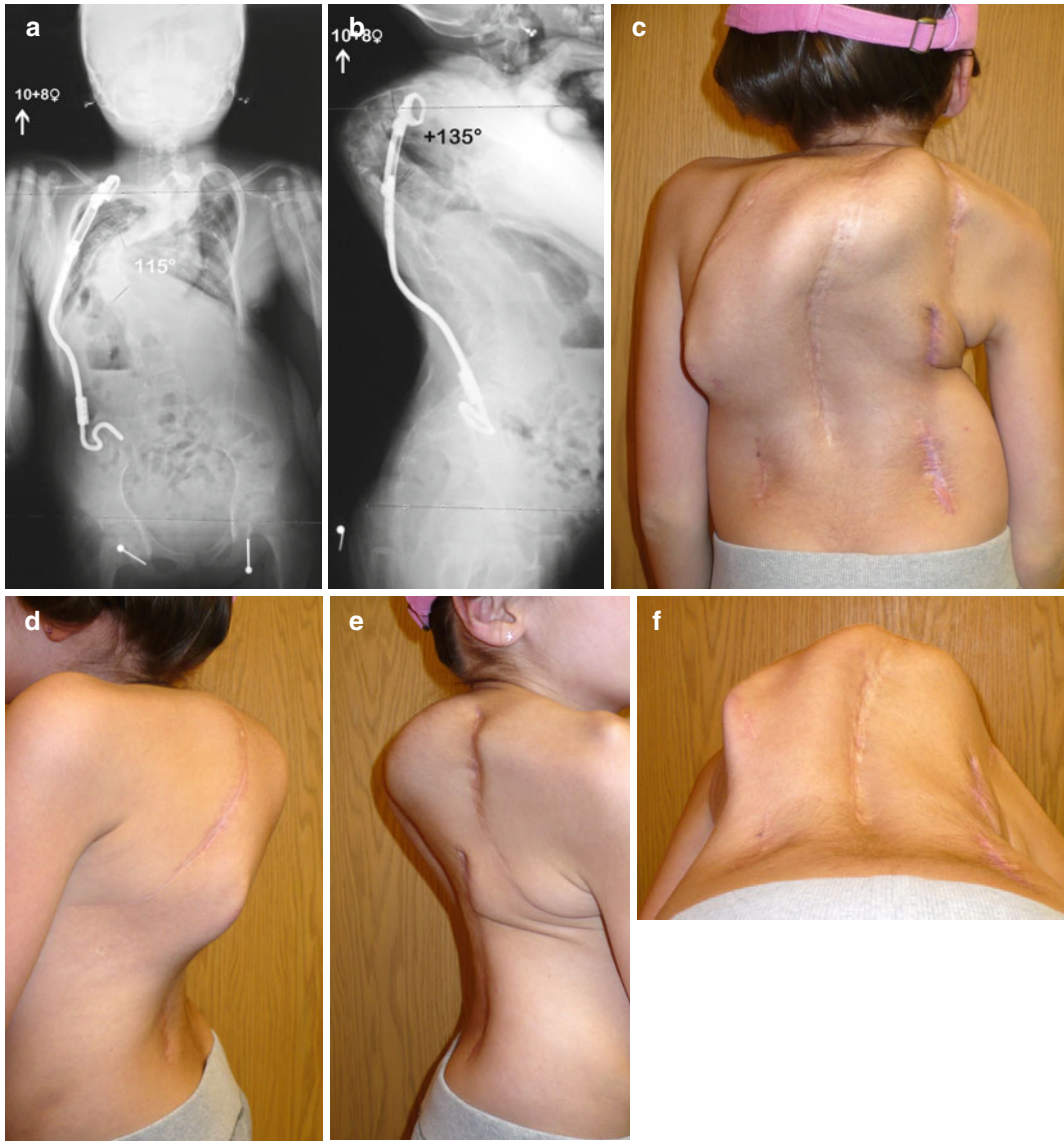

Fig. 36.6 Patient is a 10-year-old girl with severe early-onset congenital kyphoscoliosis. (**a**, **b**) She presented to us with a single VEPTR in place with a 115° thoracic scoliosis and +135° proximal thoracic kyphosis. (**c**–**h**) She had a history of 25 separate spinal surgeries all before age 10 as evidenced by the multiple scars seen on her clinical photographs. (**i**, **j**) She underwent preoperative halo-gravity traction demonstrating mild correction, followed by a two-level posterior VCR and ultimate C6–L4 spinal reconstruction to control her severe progressive kyphoscoliosis. (**k**, **l**) At 2 years postoperative, her coronal and sagittal x-rays show stable alignment. (**m**, **n**) Her preoperative and postoperative clinical photographs demonstrate improved, stable truncal alignment as well

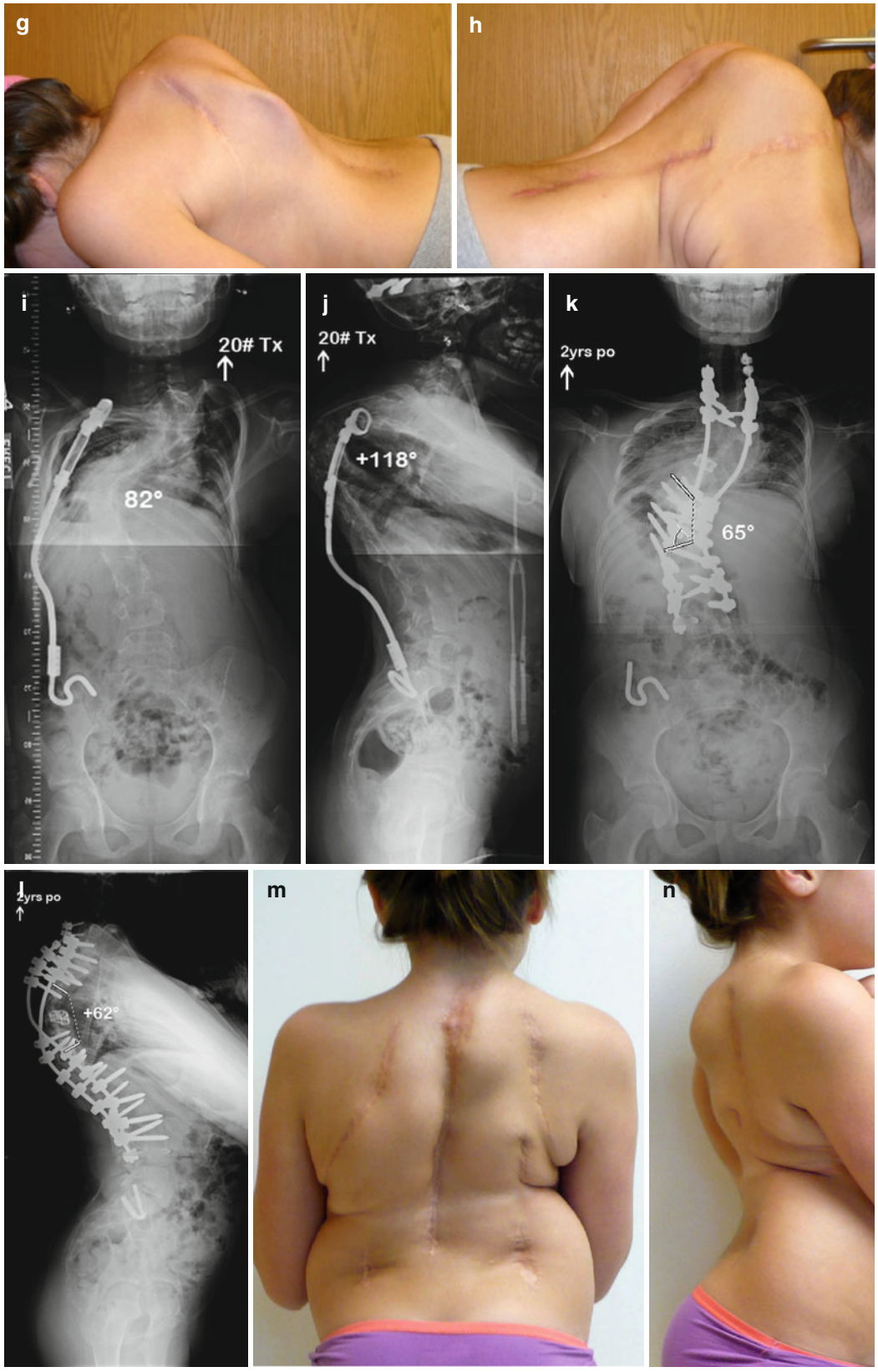

Fig. 36.6 (continued)

neurological deficit. In patients with a primary pediatric spinal deformity, this indication will be extremely rare, except in patients with a few types of congenital anomalies, specifically congenital dislocation of the spine or type 1 congenital kyphosis – failure of formation. In both of these congenital types of spinal deformities, an angular kyphosis may impinge upon the ventral spinal cord. In CDS, there is also a fair degree of spinal instability, which may lead to frank paraplegia even at a very young age. In addition, posttuberculous spinal infection can lead to severe angular kyphosis with neurological deficit. Although rare in North America, this is still somewhat common in underdeveloped countries. Fortunately, it is rare to see a primary EOS deformity patient present with a neurological deficit even with quite severe coronal Cobb angles.

Besides the few primary types of deformities discussed earlier, it is a bit more common to have patients present with actual or impending neurological deficit in a revision setting. In previously treated patients, a progressive kyphotic or kyphoscoliotic spinal deformity may lead to myelopathy, paraparesis or, in a very rare circumstance, paraplegia. The most common presentation is a patient with posterior multilevel pseudarthroses and an angular type of progressive kyphosis or kyphoscoliosis deformity causing ventral spinal cord impingement. Surgical treatment will necessitate spinal cord decompression and anterior as well as posterior spinal fusion. This can be performed via a formal circumferential approach versus an all-posterior approach with a costotransversectomy type of exposure. Even young patients will require fusion over at least the apex of their curve, with as many posterior fusion levels included to maintain ultimate coronal and sagittal balance. Potentially, performing an apical fusion with Luque trolley or Shilla-type pedicle screws at the end of the constructs may assist in the attempt to obtain some growth along the ends of the deformity while still securely stabilizing the apex to prevent further neurologic deterioration.

Conclusion

There has been much progress over the last decade in the attempt to avoid or minimize spinal fusion in very young children with progressive spinal deformities. However, as outlined in this chapter, there are certain conditions when performing at least an apical if not longer spinal fusion is quite appropriate. Each patient must be individually analyzed and treated. The analysis must include the patient's underlying medical condition, type and progression of deformity, radiographic presentation, pulmonary function, and chest wall alignment. Treatment will depend on the ability to provide adequate spinal stability, limit deformity progression, and maintain optimal chest wall alignment and function in the long term.

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Post-operative Infection in Early-Onset Scoliosis

37

Francisco J. Sanchez Pérez-Grueso

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Key Points

- The prevalence of deep postoperative surgical site infection associated with growth-friendly techniques is higher than that associated with standard spine fusion in adolescents.
- The reported post-operative infection rate is 5.3–30 % in patients treated with distraction-based systems.
- Significant risk factors for deep postoperative infection are repeated surgeries, neuromuscular diagnoses and stainless-steel implants.
- A deep infection will require aggressive irrigation, debridement and specific intravenous antibiotics.
- Even though traditional concept suggests the removal of the implants to clear the infection, new available data confirm that many patients with deep infection and implants left in place had completed the growing-rod treatment or continued the lengthening programme.

37.1 Introduction

Spinal surgery in children under 10 years of age is mainly used to treat deformities such as congenital scoliosis that requires early short fusion

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treatment with or without hemivertebra resection [11] or more commonly to treat deformities that affect a large part of the spine and that cannot be fused early due to a risk of serious repercussions on the length of the spinal column and the shape of the rib cage. Distraction-based growing systems (VEPTR, growing rods) [5, 21, 22] reduce the magnitude of the deformity and at the same time allow the spine and chest to grow using repeated lengthening surgeries.

There is hardly any information on the incidence of post-operative infections in children subjected to short arthrodesis [12], but it appears to be minimal and depends on the use of implants, the size of which may compromise the child's skin and the healing process of the surgical wound.

Information on post-operative infections in distraction-based systems is more abundant but mostly in relation to its incidence and not to its management and medium- and long-term results [1, 3]. A recent study has provided valuable information about how deep postoperative infection ultimately affects treatment outcomes in growth-friendly techniques [13].

37.2 Pathophysiology

The post-operative infection rate is 5.3–30 % in patients treated with distraction-based systems [15, 17] with the most significant risk factors being repeated surgeries and neuromuscular diagnoses such as myelodysplasia (poor quality of soft tissue) and cerebral palsy and non-ambulatory status. Other potential risk factors include low body weight in relation to age, a congenital absence of musculature in the rib cage and prominent implants [1, 7, 14, 20]. Stainless-steel implant material is also a significant risk factor for deep surgical site infection [13].

Subcutaneous or submuscular (sub-fascia) position of implants can also have a noticeable influence on the incidence of infection with 26 % on subcutaneous rods and 10 % for those submuscular rods [3].

Microorganisms most commonly identified as causal agents of infection have been coagulase-negative *Staphylococcus*, *Pseudomonas* and *Proteus mirabilis*. Some authors have emphasized the frequency of gram-negative organisms as causing infection in neuromuscular patients [4, 8].

37.3 Clinical Diagnosis

Deep postoperative surgical site infection has been defined by the Centers for Disease Control and Prevention (CDC) and modified by Horan et al. [10] as an infection occurring within 30 days after the operation if no implant is left in place or within 1 year if implant is in place and the infection appears to be related to the operation and involves deep soft tissues of the incision.

Clinical symptoms of infection are pain, redness and swelling around the wound and eventually exudate of the surgical wound accompanied by fever and an increase in white blood cells and CRP in blood tests. In general, imaging has little diagnostic value in severe post-operative infections.

The eradication of deep infection has been defined [13] as the return of ESR and CRP level to normal, a clean and intact wound with no drainage and no fever.

37.4 Treatment

Treatment for post-operative infection in children subject to definitive fusion should follow the same guidelines prescribed for adolescents [9]. A deep infection will require surgical intervention to carry out aggressive debridement and cleansing of the wound as well as instrumentation and bone grafting, all these accompanied by specific intravenous antibiotic treatment. The ultimate objective will be to try to maintain the instrumentation until obtaining a solid arthrodesis. The early removal of the surgical implant will cause instability and loss of the correction. Late removal once an apparently solid fusion is obtained will not guarantee that it will maintain the correction especially in cases with residual kyphosis [16].

The treatment of post-operative surgical site infections in children treated with distraction systems that require repeated surgeries tends to be

more controversial as success of the treatment depends on maintaining the instrumentation [1] without the necessity to terminate the planned course of treatment and undergoing a premature spinal fusion. The removal of the implant in a deep infection must therefore be the last option.

For superficial infections, a quick intervention for cleansing and direct cutaneous closure can reduce the risk of it becoming deep [18].

For deep infections, the only alternative is early debridement and cleansing accompanied by specific antibiotic treatment [1, 3, 8]. There is no data on the length of antibiotic treatment in deep infections in early-onset scoliosis treated with distraction systems, but it probably depends on the clinical course.

In the case of relapse, it is recommended to repeat the cleansing and debridement of the wound accompanied possibly by reconstructive plastic surgery [6], but ultimately it appears that no deep infection could be permanently controlled without the removal of the instrumentation [8, 15]. However, there is a debate regarding the need of implant removal for the resolution of deep wound infection in growth-friendly procedures.

Smith and Smith [18] reviewed the prevalence of post-operative infection after 678 VEPTR procedures in 97 patients to determine if infection could be managed without implant removal. Nineteen infections developed in 16 patients. Thirteen infections were classified as superficial and six deep. All patients were treated with initial irrigation and debridement and intravenous antibiotics for 58 days followed by oral suppressive therapy for 34 days. Three patients required more than one debridement to control the infection. No patient required VEPTR removal to resolve the infection.

Kabirian et al. [13] retrospectively reviewed the prevalence of deep postoperative infection in a multicentre international database. There were 379 patients treated with growing-rod technique and followed for a minimum of 2 years.

Twenty-two (52 %) of 42 patients who developed deep infection had removal of implants to control the infection. Nine of the 22 had only partial removal and routine lengthenings could continue. Thirty-one (74 %) of the 42 patients with deep infection had completed the growing-rod treatment or were still undergoing lengthenings at the latest follow-up.

Ideally, preventing the infection is the best method to use so that this type of treatment with growing rods and repeated surgeries can reach its ultimate objective [7].

It is of utmost importance to keep in mind all the risk factors that predispose an infection and try to control them. Best practice guidelines have been established in an attempt to prevent infection in high-risk paediatric spinal surgery [2, 19]. It is yet to be seen whether the application of these measures significantly reduces the rate of infection in patients with early-onset scoliosis treated with distraction systems.

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Part VI

Management of Spinal Deformity in the Growing Child: Growth Friendly Surgery

Single and Dual Traditional Growing Rods

38

George H. Thompson and Behrooz A. Akbarnia

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Key Points

- Traditional growing rod technique has evolved throughout the last 50 years.
- Both single and dual growing rods have their indications and are valuable techniques to correct and maintain deformity correction and allow the growth of the spine, thorax, and lungs but requires frequent surgical lengthening in the operating room.
- This treatment technique requires a long-term commitment by the surgeon and the family.
- Adherence to the technique detail is imperative.
- Complications are frequent but manageable when treated by an experienced spine or pediatric orthopedic surgeon.
- Cosmesis is less than ideal in single rods with "crankshaft" phenomena being a major problem.
- The results of dual-growing rods are superior when compared to single growing rod for correction and growth.

38.1 Introduction

Surgical treatment of early-onset scoliosis (EOS) is one of the most difficult challenges in modern pediatric spine surgery. This arises from the fact

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that young children with progressive spine deformity can have life-threatening cardiopulmonary complications and face severe consequences if left untreated. Ideal timing and type of intervention remain debatable. However, our understanding of the natural history and treatment options for this condition is steadily increasing and new techniques are being developed. It is understood that the development and progression of scoliosis at an early age will have a more significant impact on spinal growth, thoracic volume, and cardiopulmonary development. Appropriate treatment of the spinal deformity in these children in a timely manner is necessary to avoid these consequences.

Orthotics can be attempted [1–4] but are usually hampered by difficulties with compliance, expense, and suboptimal mechanical control of the deformity, particularly as the child gets older. Serial casts, Risser or Mehta, are very effective in young children [3–5]. At the other extreme, definitive surgical treatment using spinal fusion techniques can stabilize curve progression, even preventing crankshaft [6–8], but in such cases, this may result in a disproportionately short trunk that can further adversely affect thoracic cage and lung development [9] to bridge these options; traditional growing rods have been developed as a means to both control the spinal deformity and allow spinal and thoracic cage and lung growth and development.

Growth-friendly techniques for the treatment of EOS usually fit into three categories: distraction-based, compression-based, and growth-guided procedures [10]. The last two options are described in other chapters. The common procedures in the first group are chest wall distraction techniques such as rib devices (vertically expandable prosthetic titanium ribs [VEPTR]), traditional growing rods (single or dual) used primarily for the spine, and most recently the magnetically controlled growing rods. In this chapter, we will discuss indications, current surgical techniques, and complications of traditional growing rod technique in EOS.

From the time Harrington [11] introduced instrumentation without fusion in 1962, the growing rod techniques have evolved from one rod and two hooks placed subcutaneously [12] to the current dual-rod technique [13]. Historical background will be discussed in more detail for better understanding of the principles behind growing rod surgery.

38.2 Background

The indications and goals of surgery have changed significantly over the years. Currently the recommendation for surgery among children with idiopathic EOS is scoliosis of 45° or greater and documented progression. The trend to operate is a reflection of our improved understanding of the natural history of this disease and significant advances in surgical technology.

Historically, the goal of surgery has been to achieve a straight but shortened spine by early spine fusion when the curves could not be controlled. These goals quickly changed after Dubousset et al. [14] described the crankshaft phenomenon. This phenomenon is seen in skeletally immature patients following posterior spinal fusion. The posterior fusion mass acts as a tether to the growth of the anterior column, resulting in increase of rotational deformity. Sanders et al. [15] identified patients with Risser 0 and open triradiate cartilage as the population at maximum risk for crankshaft after posterior only fusion. To avoid this phenomenon, anterior spinal fusion was recommended in addition to posterior fusion. It was later discovered that limiting the growth of the immature spine with an anterior/posterior fusion had significant untoward effects on pulmonary development, chest growth, and height. Spine growth has been described by Dimeglio [16] who outlined two peaks in spinal growth: 0–5 and 11–15 years. Using his data, one can predict the expected height loss from an anterior/posterior fusion. Winter [17] similarly described a formula to calculate the expected height loss: multiply 0.07 × number of segments fused × number of years of growth remaining. Being able to predict this is a valuable tool in educating family and being aware of the potential ramifications of fusion at an early age. The negative consequences of early fusion on pulmonary

and thoracic development have been a strong motivating factor in the development of surgical techniques aimed at reducing morbidity and early mortality.

Advances in surgical treatment have been accompanied by a paradigm shift in the goals of surgical management of idiopathic EOS. No longer is a short but straight spine acceptable. Some spine deformity is acceptable if spine and chest growth can be maintained, allowing for more normal pulmonary development.

Harrington [11] was the first to describe internal fixation without fusion among 27 post-polio and idiopathic patients; however, his initial study did not describe the long-term follow-up among this subset of patients. His surgical technique included a subperiosteal dissection placing a single distraction rod on the concavity of the curve attached to hooks at both ends. The concept was to maintain spinal growth without fusion, correct deformity, and control residual deformity. He ultimately concluded that children under the age of 10 years were candidates for fusionless surgery and those older than 10 years needed definitive fusion.

Moe et al. [18] modified the technique described by Harrington and limited the subperiosteal dissection to the site of hook placement and passed the rod subcutaneously. The rod was also modified to have a smooth, thicker central portion to prevent scar formation to the threads and allow for sagittal contouring. Lengthenings were performed when there was a loss of correction of the major curve of 10° or more. Both the children treated for idiopathic scoliosis showed a considerable decrease in curve magnitude. They reported a 50 % complication rate including rod breakage and hook dislodgment from the rod or lamina.

Marchetti and Faldini [19] described their "end fusion technique" in 1977. The initial surgery included fusion of two vertebrae at each end of the curve. Six months later, these foundations were used as strong anchor points for hook placement. A rod was then placed connecting the two ends after subperiosteal dissection. Lengthening was then performed periodically. A Milwaukee brace was used as external support. The first patient treated by Marchetti and Faldini with this method was an 11-year-old girl with a thoracolumbar curve of 148°; the patient had a curve of 69° at the end of treatment 4 years later.

In 1977, Luque and Cardoso [20] described their technique of fusion-less surgery with segmental spinal instrumentation. Luque modified this technique by adding sublaminar wires and replacing the Harrington rod with an L-shaped rod. This technique was later coined as the "Luque trolley" technique. The initial series included 47 paralytic patients who grew a mean of 4.6 cm over the instrumented segment and an initial curve correction of 78 %. Although early results were promising, the system grew out of favor due to the extensive subperiosteal exposure and reports of early fusion. Sublaminar wire passage also created scar tissue and weakened the lamina resulting in more difficult definitive fusion surgery.

Pratt et al. [21] later performed a retrospective review of 26 patients treated with Luque trolley instrumentation with and without convex epiphysiodesis. Of the eight patients treated with Luque trolley alone, all showed significant curve deterioration. Of those treated with epiphysiodesis, 7 of 13 worsened, 4 remained unchanged, and 2 improved. Growth was 49 % as predicted in the Luque trolley alone group and 32 % among those undergoing combined surgery.

Mardjetko et al. [6] performed a retrospective review of nine patients who underwent Luque instrumentation without fusion. The mean preoperative major curve was 51°. All nine patients had at least one revision. All revisions were technically demanding due to extensive fibrosis and weakened laminar bone. Follow-up curves had a mean of 51° with a mean gain in spinal height of 5.8 cm. Only a small portion of this growth was derived from the instrumented levels. Spontaneous fusion was documented in all patients.

In 1997, Klemme et al. [22] reported their 20-year follow-up of the Minnesota experience. Sixty-seven children with various diagnoses were treated. Major curve magnitude improved from 67° at initial internal fixation to 47° at definitive fusion. They felt that the amount of correction

Table 38.1 Review and comparison of selected literature

Authors	Number of patients	Mean initial elongation pre- to post-initial (cm)	Mean growth of instrumented area (cm) during treatment	Mean total length increase of the spine (cm)	T1–S1 increase (cm/year)	Space available for lung (SAL) ratio (pre/final)	Number of complications per patient
Moe et al. [18]	20	Not reported	2.9	Not reported	Not reported	Not reported	1.1
Luque et al. [20]	47	Not reported	2.6	Not reported	Not reported	Not reported	0.3
Klemme et al. [22]	67	Not reported	3.1	Not reported	Not reported	Not reported	0.81
Blakemore et al. [23]	29	Not reported	Not reported	Not reported	Not reported	Not reported	0.3
Akbarnia et al. [13]	23	5.0	4.67	9.64	1.21	0.87/1	0.57
Akbarnia et al. [26]	13	5	5.7	10.7	1.46	Not reported	0.46

obtained declined with consecutive lengthenings (mean 6.1 procedures per patient). Curve progression was arrested or improved in 44 of 67 patients with a mean curve reduction of 30 %. Of the remaining 23 patients, 12 were neuromuscular and the curves progressed a mean of 33 %.

Blakemore et al. [23] reported on 29 patients with progressive kyphoscoliosis treated with a single submuscular Isola rod with or without apical fusion or convex hemiepiphysiodesis. Apical fusion was performed on curves >70° and on those curves that were stiff on bending radiographs. All used a Milwaukee brace postoperatively. Mean major curve improved from 66° to 38° immediately postoperative and 47° at latest follow-up. Overall complication rate was 24 % and included hook dislodgment [24], rod breakage [25], and superficial wound infection [13].

In 2005, Akbarnia et al. [13] building on the experience of McCarthy and Asher described the dual growing rod technique for EOS. The original series included 23 patients treated with dual growing rods using pediatric Isola instrumentation and especially made tandem connectors. All the patients had curve progression over 10° and unsuccessful treatment with bracing or casting. They reported on 189 procedures, of which 151 were lengthenings. Mean preoperative curve improved from 82° to 38° after initial surgery, and correction was maintained to 36° at last follow-up or final fusion. Mean growth from T1 to S1 was 1.21 cm/year excluding the initial correction. Space available for lung (SAL) ratio in patients with thoracic curves improved from 0.87 to a normal ratio of 1.0. Complications occurred in 11 of 23 patients (48 %) during the course of treatment. These included hook dislodgment, rod breakage, and superficial wound infections. This procedure was felt to be both a safe and effective technique for treating EOS [26]. See Table 38.1 for summary of the literature.

38.3 Indications

Principles governing the treatment of EOS are adhered to when applying traditional growing rod techniques. There is a trend in recent years to employ a more aggressive treatment strategy. In idiopathic EOS curves, progression greater than 10°, major curve greater than 35°, and RVAD greater than 20° should prompt consideration for active treatment. When curve magnitude reaches 45° or more, consideration should be given for surgical intervention. Older patients with idiopathic EOS follow principles similar to those with adolescent idiopathic scoliosis.

Table 38.2 Indications for treatment

Indications for operative vs. nonoperative treatment in EOS	
Nonoperative	Major curve <20°[a]
	RVAD <20°[a]
	Phase 1 rib head[a]
Operative	Major curve >25°[a]
	RVAD <20°[a]
	Phase 2 rib head[a]
	Failure of brace/casting treatment
	Documented progression of curve

[a]Idiopathic early-onset scoliosis only

Table 38.3 Comparison of single and dual-rod techniques

	Single rod	Dual rods
Unplanned trips to OR	Increased risk	–
Implant-related complications	Increased risk	–
Infection	No difference	No difference
Growth (mm/year)	6.8	11.3
Initial correction (major curve %)	38	47
Maintenance of correction (%)	14	40

Reprinted from Thompson et al. [27]. With permission from Wolters Kluwer Health

Documented curve progression and curve magnitude greater than 45° are indications for surgical intervention. However, curve angle alone should not be the main indication for surgical treatment. Deterioration of pulmonary function and other associated conditions leading to the impairment of quality of life may influence the decision to wait and accept some further progression or do an earlier intervention (Table 38.2).

Once the decision is made to treat the child surgically, the question should be asked as to which method would most consistently give good results with the least or most manageable complications for that particular child. The growing rod technique has the best indication in patients with idiopathic, neuromuscular, or syndromic scoliosis and absence of congenital anomalies. The dual growing rod has been found superior in clinical outcome to single-rod technique. A recent comparison of dual- vs. single-rod constructs by Thompson et al. [27] highlights this debate. The results quite convincingly favor the dual growing rod technique. Although the overall complication rate was slightly higher (19 %[3/16 patients] vs. 29 % [2/7 patients]), the amount of initial correction obtained and final correction sustained, as well as improved growth rate and T1–S1 length, make this a more effective treatment. The presence of a second rod allows some leeway for revision when one rod breaks or undergoes plastic deformation. In the single-rod construct, a broken rod or implant complication is urgent and cannot wait until the next scheduled lengthening (Table 38.3).

Bess et al. [24] further highlighted these differences from the Growing Spine Study Group (GSSG) database review of 910 growing rod surgeries. They found that the need for reoperation was lower in the dual vs. single group for complications. Also, the rate of implant-related complications was higher in the single-rod group. Superficial wound problems were greater in the dual-rod group due to the bulkiness of implants, but did not lead to a need for additional surgeries. The construct of the dual rod is also biomechanically favorable with improved initial curve correction and maintenance of correction.

Although the dual growing rod system may be somewhat more prominent than single rods, it certainly is lower profile than the current available rib devices such as VEPTR. Furthermore, dual growing rods more specifically address the pathology it treats compared to VEPTR. When comparing this technique to VEPTR, the main distinction that should be made is in the very indication. If a patient has thoracic insufficiency syndrome (TIS) with multiple congenital anomalies and stiff thorax, then the VEPTR may provide a better treatment modality. However, in children with scoliosis without chest wall anomalies and TIS, it could be argued that the growing rod constructs is lower profile and more directly addresses the spinal deformity. With its anchor points on the cephalad and caudal ends of the spine, it avoids attachment to the ribs and

Table 38.4 Indications for distraction-based devices

	Indications	Contraindications
Chest wall distraction (VEPTR[a])	*Thoracic insufficiency syndrome*[b]	Skeletal maturity
	Skeletally immature[b]	Poor rib bone stock
	Congenital scoliosis/fused ribs	Absence of proximal ribs
	Chest wall deficiency	
Spine distraction (growing rods)	IEOS	Skeletal maturity
	Neuromuscular scoliosis	
	Scoliosis associated with syndromes	

IEOS idiopathic early-onset scoliosis
[a]Vertically expandable prosthetic titanium rib (Synthes Inc.)
[b]FDA indications

Table 38.5 Predictable complications of two distraction-based nonfusion techniques

Growing rod	Chest wall distraction
Multiple surgeries, infection	Multiple surgeries, infection
Rod breakage	Drift of device attachments, brachial plexus injury
Spine stiffness or fusion	Chest wall stiffness

therefore avoids associated pulmonary complications such as chest wall stiffness. As the goal of this technique is growth modulation, it can be argued that placing anchors on a mobile segment away from the spine (on ribs) will result in a lower transmission of force on the spine, and consequently there is less stimulation of growth. For this reason, when growth modulation is the goal, growing rods are the most reliable way to accomplish this. No information on T1–S1 length is available following VEPTR procedures to date (Tables 38.4 and 38.5).

Traditional growing rods have been used for the treatment of EOS with a variety of etiologies such as idiopathic, neuromuscular, congenital, and syndromic disorders.

The growing rod may not be an effective procedure when there is no further growth potential or in patients whose primary problem is chest wall abnormalities and/or TIS. One also has to be careful in the presence of kyphosis to take precautions to avoid complications due to kyphosis, as discussed later in this chapter.

38.4 Surgical Technique

38.4.1 Surgical Technique for Single Growing Rods

38.4.1.1 Intraoperative

Our single growing rod construct consists of a proximal "claw" with hooks and a distal construct of either hooks or pedicle screws. The technique is similar to other spine procedures (Fig. 38.1a–d). After induction of general anesthesia, the patient is positioned prone on either the Hall-Relton four-poster frame or a Jackson table. Typically, a long, midline incision is made over that portion of the spine that is to be instrumented. The exact length is based on preoperative planning. The subcutaneous tissues are divided and the fascia exposed. One spinous process proximally and distally delineating the foundation sites is exposed and marked with a clamp. Intraoperative radiographs are obtained to identify the preplanned foundation sites. Once this has been accomplished, then the two or three adjacent vertebrae are exposed subperiosteally. The fascia between the foundation sites is also divided and the muscle thinned so that the rod will lie close to the spine. However, the laminae are not exposed or visualized. The distal foundation is usually constructed first. In the beginning, we used a claw construct with an over-the-top laminar hook proximally and a sublaminar hook distally. This allowed compression between the hooks. However, in the last 5 years, we have used a pedicle screw construct for the

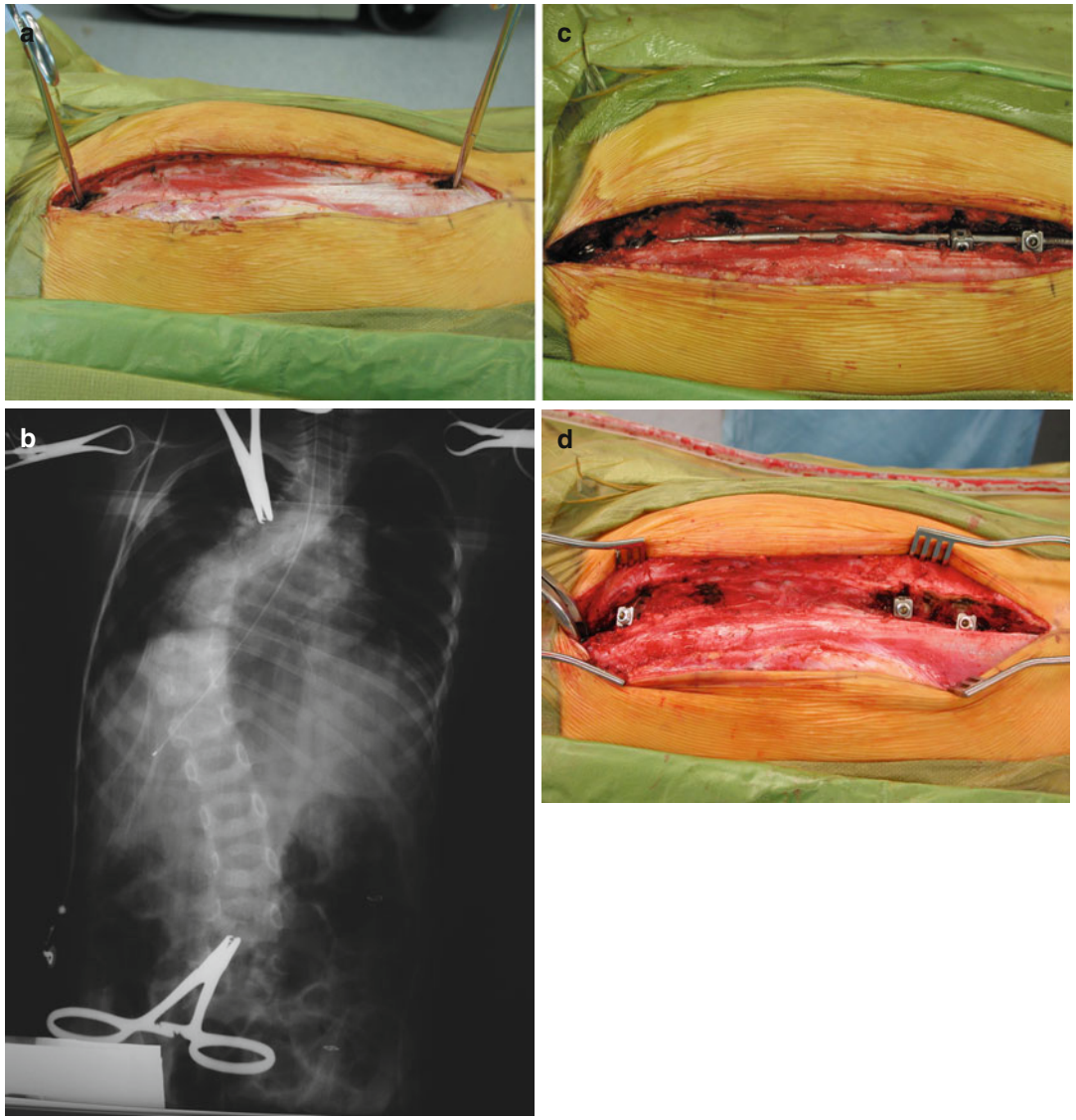

Fig. 38.1 (**a**) Intraoperative photograph demonstrating the incision and the exposure for the upper and lower foundations. The spinous processes at the planned levels are identified and marked. A radiograph is obtained to confirm the preplanned levels. (**b**) Intraoperative anteroposterior (AP) radiograph demonstrating identification of the foundation sites. (**c**) Hooks are typically used for the upper foundation and hooks and/or screws for the lower foundation. Hooks spanning two vertebral segments for the proximal and distal foundation were used in this patient. The muscle has been separated to allow the rod to lie as close to the bone as possible. However, the laminae, facets, and spinous processes are not exposed except at the foundation sites, which are fused. (**d**) Following rod insertion, a long segment of rod is *left* extending below the lower foundation. This is used to expand the rod and maintain spinal length. The fascia is closed over the instrumentation

distal foundation. The proximal foundation consists of an over-the-top laminar hook proximally and a sublaminar hook in the next distal vertebrae. Occasionally, this may extend one additional level distally. This also allows for compression once the rod has been inserted. The size of the rod is based

on the size of the patient. In most small children, a 5.5 mm rod is used, while in older children, a 6.35 mm rod is used. The interval between the foundations is measured with a silk suture and the rod measured. Typically, an additional 5 or 6 cm is added to this measurement to compensate for the initial distraction and to have enough extra length to allow it to be expanded at subsequent surgeries. The rod is appropriately contoured and inserted. When hooks are utilized for the distal foundation, they occasionally need to be open hooks to allow for the rod to be appropriately seated. Once the rod is inserted, it is grasped with rod holders and rotated to allow for a relatively normal sagittal plane alignment. The upper foundation is typically tightened first. The rod is distracted distally. This is always done under intraoperative neuromuscular monitoring. There is 4 or 5 cm of rod extending below the lower hook or screw. This is used for subsequent lengthenings. The wound is closed in layers. The fascia is closed over the entire construct. Typically, a drain is placed over the top of the fascia. The subcutaneous tissues and skin are closed in a standard manner. The latter is typically a subcuticular closure.

38.4.2 Technique for Dual Rod

38.4.2.1 Initial Dual-Rod Procedure

Patient is brought to the operating room and general anesthesia is induced. Prophylactic antibiotics are administered. Patient is placed on the table in prone position over chest roles for small children and over a frame or Jackson table for larger patients. The entire back is prepped and draped in usual fashion.

The procedure includes preparation of cephalad and caudal foundations for anchor placement and performing a limited fusion, rod contouring, rod passage, and application of connectors. The index procedure can usually be performed through one or two midline incisions [26].

38.4.2.2 Technique of Preparing Foundations

A foundation is defined as an assembly of at least two anchors and one or two rods that are stable and strong enough to accept corrective loads and to resist deforming loads without dislodgment of the anchors or plastic deformation of the rod [28]. In a typical dual-rod technique, we have used four anchors (hooks or screws) for each foundation for maximum stability (Fig. 38.2a–d). The exposures at the foundations are the only locations where the exposures are subperiosteal. Meticulous technique is employed to avoid a broad exposure and risking the occurrence of spontaneous fusion.

Selection of foundation sites is based on the type and location of the curve as well as the patient's age and diagnosis. Patients with neuromuscular scoliosis, for example, may require longer instrumentation compared to those with idiopathic curves. The upper foundation anchors are generally placed at the T2–T4 levels in a claw fashion. If hooks are used at the upper foundation, the author's preference is a supralaminar location for superior hooks. The inferior hooks are placed sublaminar under the lamina (facet), similar to the technique used with original Harrington hooks, in a "claw" construct (Fig. 38.3a, b). The superior hooks can be staggered over two or three levels to avoid crowding the spinal canal if this is a concern. Staggering hook levels, however, may interfere with the use of a transverse connector at the foundation level. In that case, the connector is attached just below the lower hooks. It is utmost crucial to achieve the best foundation stability at the initial surgery to reduce the possibility of failure, even at the expense of exposing three instead of two levels. A study by Mahar et al. [29] demonstrated the increased stability of a screw construct over hooks alone or hooks with cross-connectors showing the significance of adding a cross-link if all hook construct is used. We utilize both hooks and screws for foundations. It is our preference to use pedicle screws whenever possible; however, if anatomical considerations make this difficult, then hooks are another excellent option (Fig. 38.4a–c).

The caudal foundation is generally instrumented with four pedicle screws. The foundation levels are typically three levels below the lower end vertebra of the major curve. In the presence

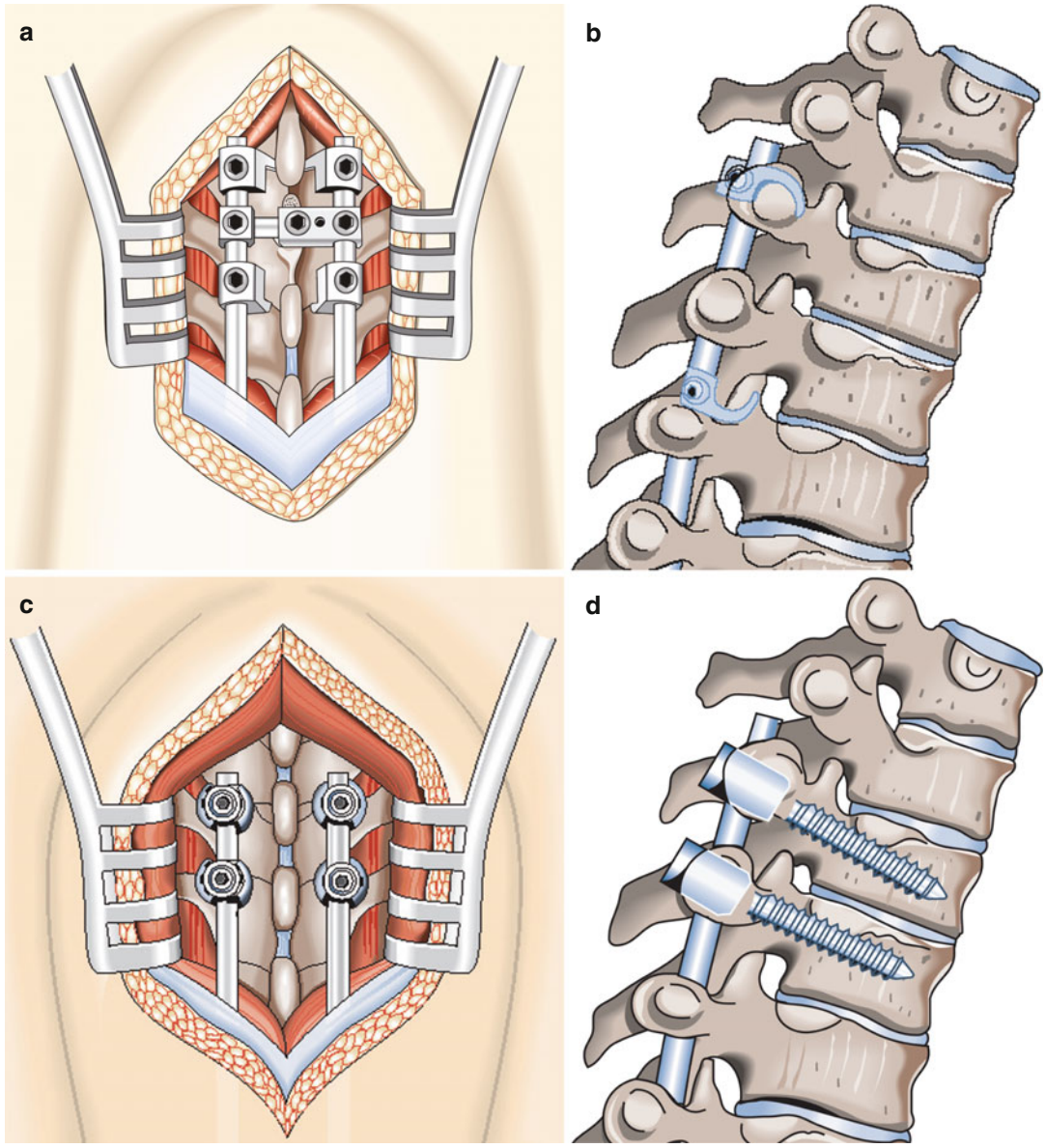

Fig. 38.2 Anteroposterior and lateral view of upper foundations using four hooks and a cross connector (**a**, **b**) or four screws (**c**, **d**)

of pelvic obliquity, such as neuromuscular scoliosis, the distal foundation may be extended to the sacrum or ilium using intrailiac fixations. It is extremely important that the foundations be stable and as strong as the bone quality permits. Bone graft or bone graft substitutes are used to augment bony fusion across at the foundation sites including a facet fusion and secure foundation anchors.

38.4.2.3 Insertion of Dual Rods and Anchors

A low-profile pediatric implant system, with appropriate dimensions, such as 4.5 mm diameter stainless steel or titanium rods, is usually used. The rods are measured and cut into four segments, two for each side, and contoured for sagittal alignment. When tandem connectors are used, the rods should be cut in the region where the rod

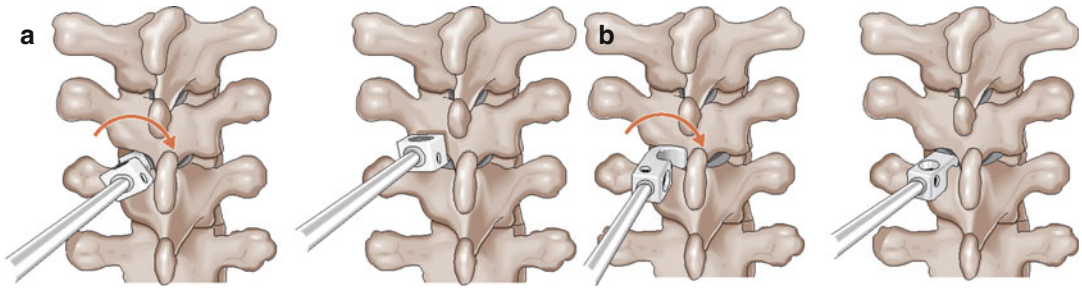

Fig. 38.3 Hook insertion maneuvers. Infralaminar hooks are inserted applying lateral to medial rotary motion (**a**); supralaminar hooks can be placed by medial to lateral rotation of hook holder (**b**)

ends meet mostly at the thoracolumbar junction as this will be the best site for the tandem connectors. Appropriate contouring may help to correct the kyphosis using a cantilever maneuver when the deformity is flexible. One has to avoid extreme sagittal correction in one session to avoid anchor failure. The individual rods are passed subfascially as the tip of the rod is felt at all times through the skin until it appears at the site of the upper foundation. They will then be secured to their respective anchors and a cross-link can be added if necessary. Mahar et al. did not show any benefit of cross-connecting if four pedicle screws are used for fixation, but did find significantly more strength associated with a cross-link and hook foundation. The lower foundation is then prepared and contour rods are connected to the caudal anchors. The tandem connectors are then placed at the thoracolumbar junction by first sliding them cephalad and then caudal. The thoracolumbar region is chosen for tandem connectors, as this is an area of the spine that is anatomically straight. The tandem connectors are rigid and do not bend; therefore, this location has the least effect on sagittal alignment. Lengthenings are done mostly by loosening the screw of the upper rod; therefore, only a short segment of the lower rod should be attached to the tandem connector leaving room for more of the upper rods to be inside the connector. This segment of the rod should not be contoured to allow entry into the straight connector. The tandem connectors are placed in their least prominent position to achieve the lowest profile possible. If there are set screws with connectors, they can be placed facing either medially or laterally. Turning the tandem connectors medially (set screw heads facing medial) makes the reach easier for the screws and possibly allows minimally invasive access during lengthening procedures. The lateral placement however may have a lower profile. Another option for rod connection is side-to-side connection. When side-to-side connection is used, the location of the connectors may vary and the rods in those segments can remain contoured for appropriate sagittal alignment.

At this point, an initial correction and lengthening is performed; however, extreme care is taken to avoid over distraction and immediate implant or neurological complication.

38.5 Lengthenings

The rods are lengthened periodically, usually every 6 months [13]. One method, usually in single-rod technique, is to leave the ends of the rod long. As long as there is enough rod available proximally or distally from the foundation for lengthening, this area alone can be exposed and lengthened.

In otherwise healthy children, this procedure can be performed on an outpatient basis at a hospital or ambulatory surgery center. In children with significant comorbidities, an inpatient stay or even intensive care may be needed for recovery and a children's hospital with specialized care is preferred. Spinal cord monitoring should be considered during lengthening procedures.

Fluoroscopy can be used to identify the location of the connector if one is used. The gap

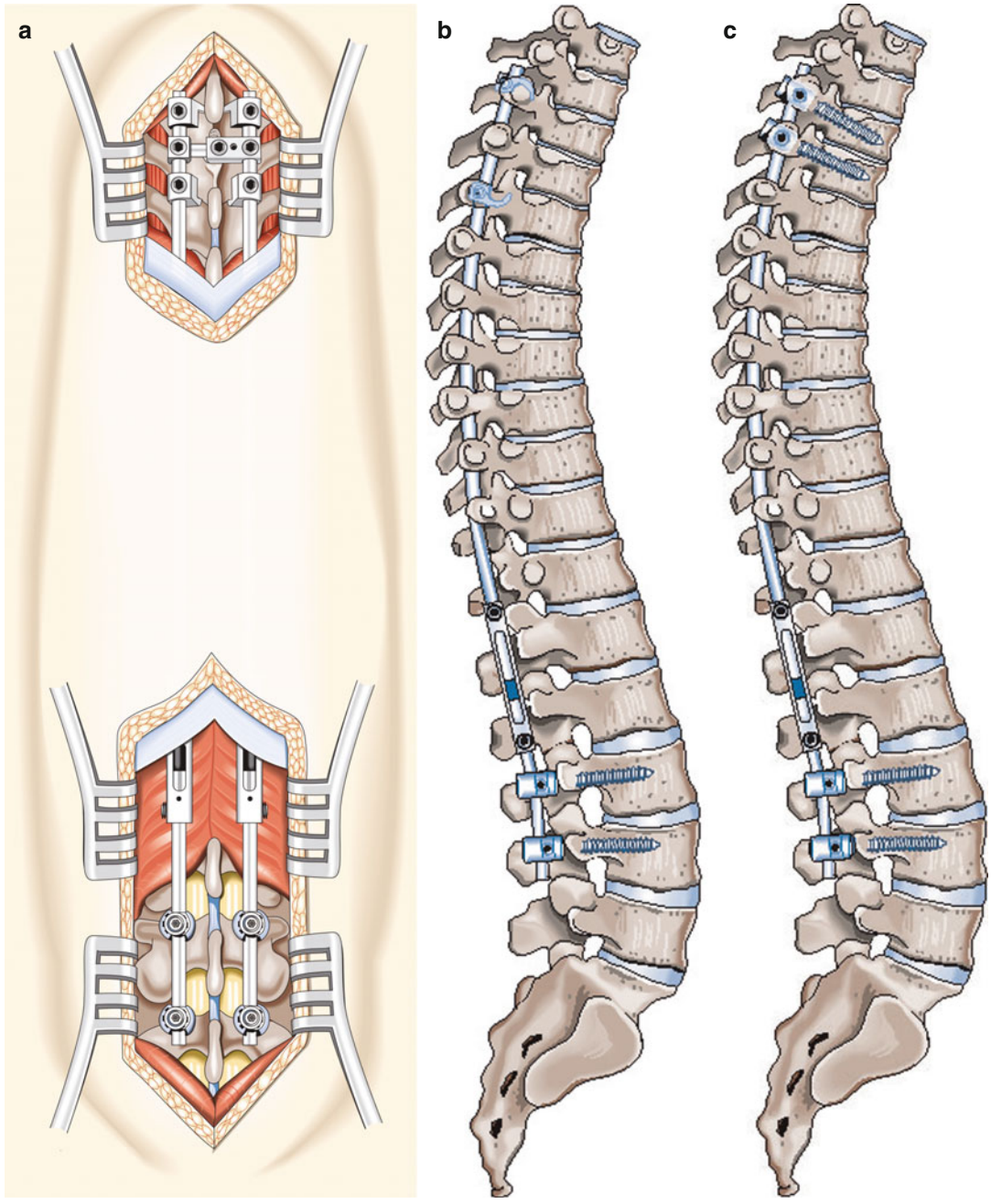

Fig. 38.4 Considering patient's age, anatomical provision and bone quality screws and hooks can be used as upper and lower anchors to provide strong foundations (**a**–**c**)

between the rods within the tandem connector and the site of the *proximal* set screws are identified. For dual-rod lengthening, a small midline incision is made, centered between the tandem connectors and at the site of the rod gap if all screws are medial or posterior. It is vital that the skin incision is taken to the depth of the tandem connectors prior to working laterally so that only one skin flap is created and to save good skin thickness and coverage. The gap on the side of

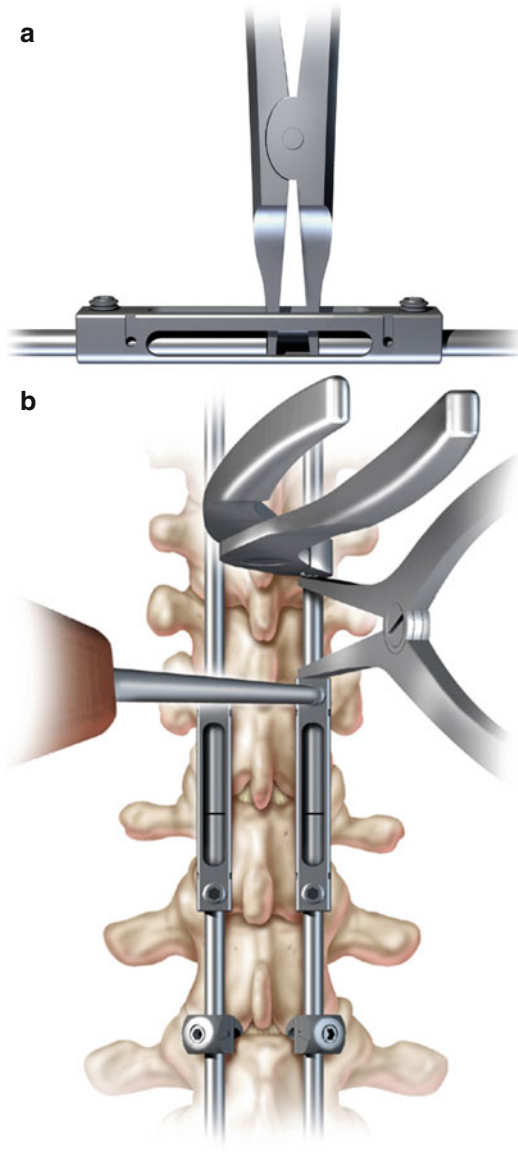

Fig. 38.5 Distraction can be done with special tandem connector distractor (**a**) or using a rod holder and a distractor outside the tandem connector (**b**)

the spine needing more correction (usually the concave side) is exposed and freed of fibrous tissue in order to fit the special distractor inside the connector and into the gap between the rods. Both the upper set screws are loosened (ensure that the distractor is already in place for lengthening to avoid any loss of length) and one side is lengthened (Fig. 38.5a). Excessive distraction is avoided. The set screw is then tightened. On the contralateral side, the distraction is performed to match the first side unless differential distraction is desired for improved coronal balance.

38.5.1 Lengthening Outside the Tandem Connector

A small midline incision is made just cephalad to the tandem connector. The incision must be long enough to reach the set screws and accommodate a rod holder. The same meticulous exposure is performed to approach the rods. The exposure is completed on both sides prior to lengthening. A rod holder is then placed cephalad to the tandem connector, with enough room to place a distractor between the rod holder and the connector (Fig. 38.5b). Both the set screws are released and sequential lengthening as described above is performed. The indications to employ this technique are when the rods are too close to each other within the connector and the distractor will not fit. If the rods are too far away from each other within the connector, then small pieces of rod can be placed within the connector to avoid a larger skin incision.

Excessive distraction force *must* be avoided especially at the first lengthening, to avoid implant complications. The timing of lengthening is universally at 6-month intervals. As the number of lengthenings increase and growth potential decrease, the interval can be longer. They are stopped when no further distraction is achieved and therefore are ready for final fusion. When the side-to-side connector is used, the rods can be lengthened by tightening the appropriate set screws of each connector and distracting between the connectors to distract the rods (Fig. 38.6). Based on which of the two screws are tightened, distraction or compression can be achieved using only distraction between two connectors.

38.6 Final Fusion

Final fusion usually necessitates the removal of growing rod system, complete exposure of the spine, identifying any possible fusion areas, correction of residual deformity, and reinstrumentation

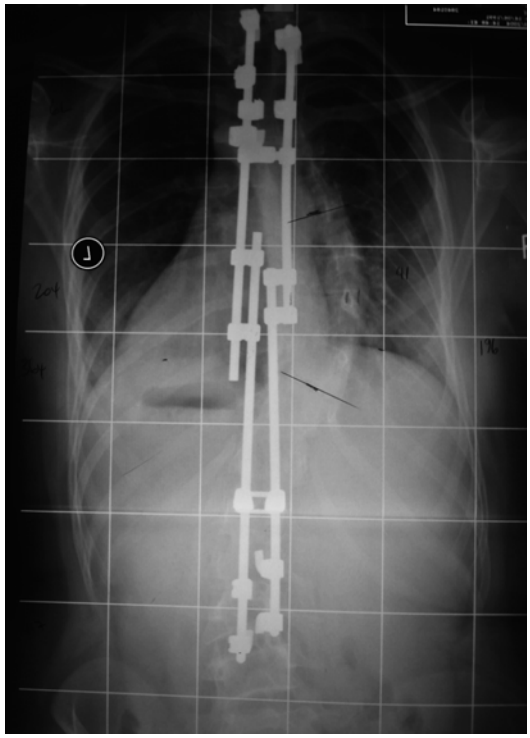

Fig. 38.6 When the side-to-side connector is used, the rods can be lengthened by tightening the appropriate set screws of each connector and distracting the connectors to achieve distraction or compression (Courtesy of David S. Marks, FRCS)

with arthrodesis. It may require multiple osteotomies to achieve correction. The levels of fusion are usually the same as the levels spanned in the growing rod construct (Fig. 38.7a–g). Exceptions to this include patients who have progressive curves above or below the foundations, an example being neuromuscular patients with pelvic obliquity. For more details regarding reduction maneuvers, instrumented fusion, and revision surgery, please see respective Chaps. 35 and 36 (Fig. 38.8).

38.6.1 Spinal Cord Monitoring

Although neurological risks are unlikely during growing rod surgery, neuromonitoring is commonly used in initial surgeries and often in lengthening/exchange procedures. Sankar et al. [30] reported a temporary neurological event among 782 growing rod surgeries (incidence rate = 0.1 %). This was caused by pedicle screw misplacement and resolved after 3 months. In their report, the incidence of intraoperative neuromonitoring changes was 0.9, 0.9, and 0.5 % in initial surgery, revision surgery, and lengthening, respectively. Recently, a case of delayed neurological deficit was reported by Akbarnia et al. [31], who had delayed neurological deficit after rod exchange procedure despite normal intraoperative somatosensory evoked potentials (SSEPs), Hoffmann reflexes (H-reflexes), and EMGs; the patient had full recovery after rod shortening. This highlights the need for more sensitive neuromonitoring techniques.

Despite the early historical success of SSEP monitoring, reliance on this one modality is no longer adequate for the patients who present for surgical correction of complex deformities. Hence multimodality neurophysiologic monitoring of the spinal cord including highly sensitive-specific transcranial electric motor evoked potentials (tceMEPs) has a definite place in the context of growing rod placement and lengthening/adjustment procedures. For more details on neuromonitoring, please see Chap. 53.

38.7 Complications (Fig. 38.9a–g)

Complications following growing rod techniques have been a problem since its first description by Harrington [11]. In his series, the complication rate was 48 % (32/67) for the duration of the treatment. Several other reports have addressed the high complication rate of this procedure and therefore Chapter 38 will address this separately [13, 26, 27, 32].

Bess et al. [24] reported the GSSG data on 897 growing rod procedures (single and dual growing rods) among 143 patients. Complication rate per procedure was 19 % (177/910). Eighty-one of 143 patients (57 %) had a minimum of one complication. Nineteen of 94 (20 %) patients with single rods had unplanned procedures due to implant complications compared to 7 of 83 patients with dual rods (8.4 %). Thirteen of 52 (13 %) patients with subcutaneous rods had

wound complications compared to 9/90 (10 %) patients with submuscular rods ($p<0.05$).

They were also able to demonstrate an increased risk for complications with increasing number of procedures. Complication risk increased 13 % with each successive procedure. More importantly, however, they demonstrated that not all complications require a separate surgical procedure. Furthermore, they identified factors under the control of treating surgeons to reduce complications and unplanned surgery. These factors include age at initial implantation, number of procedures performed during the treatment period, use of dual growing rods, and submuscular rod placement. Younger children are at higher risk for complications during the treatment period for several reasons: They have less soft tissue coverage, smaller bone, and less physiologic reserves than older children, and younger age at initial instrumentation implies a longer treatment course and increased number of operations needed until final fusion.

Dual growing rods reduced implant-associated complications and unplanned surgery for two

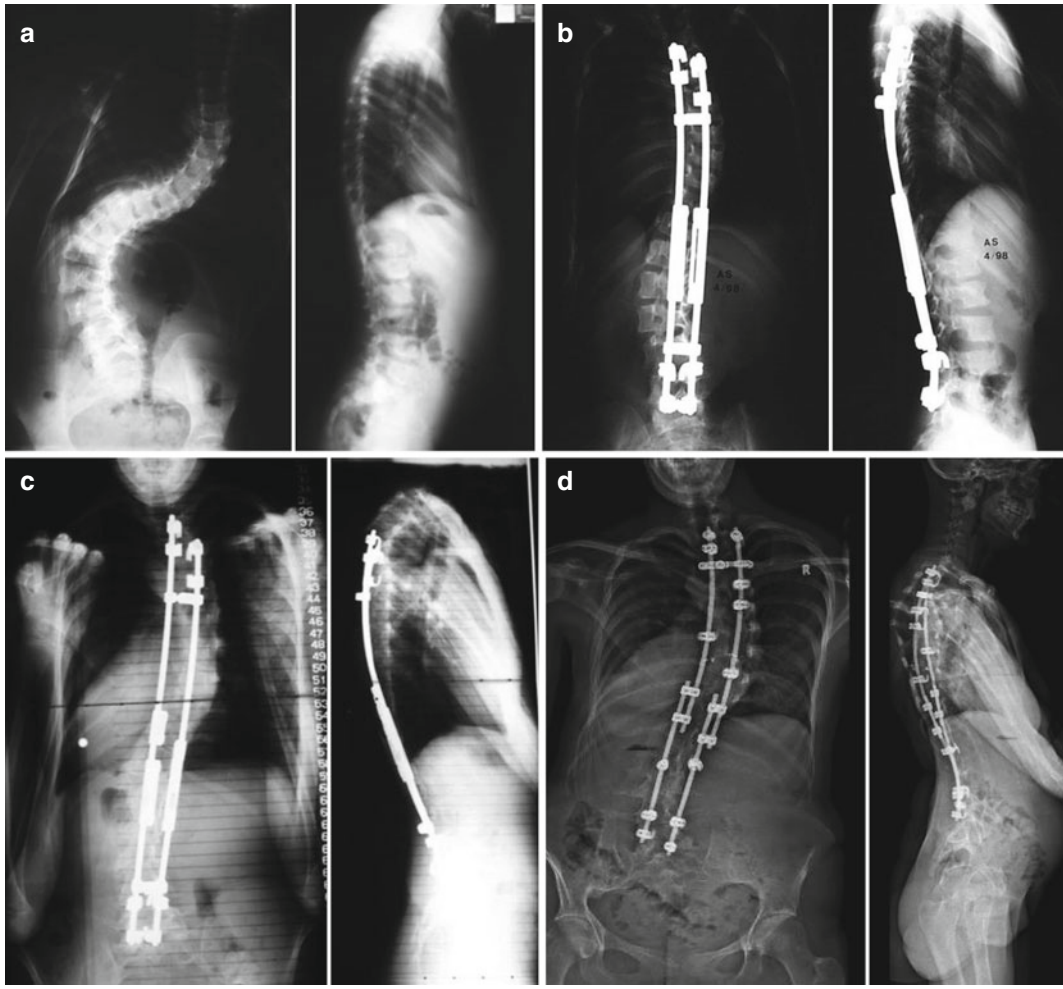

Fig. 38.7 A 4-year-old girl with Marfan syndrome (**a**). Patient underwent dual growing rod insertion for treatment of severe progressive scoliosis (**b**). Same patient after 7 years active treatment with frequent lengthening, before final fusion (**c**) and after final fusion (**d**). Clinical images showing the same patient, pre-initial surgery (**e**), post-initial surgery (**f**), and post-final spinal fusion (**g**)

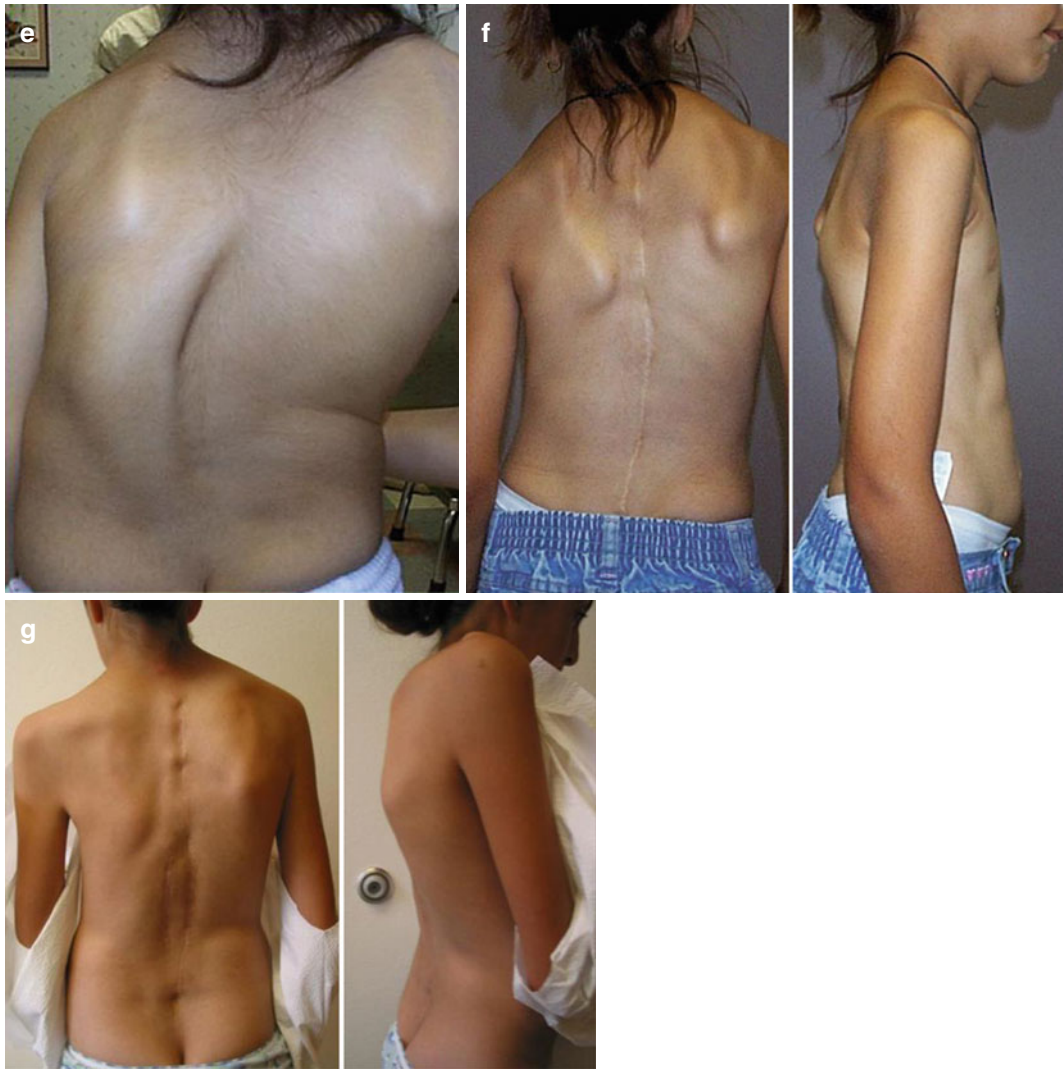

Fig. 38.7 (continued)

possible reasons. Dual rods reduce the mechanical stress on a rod compared to the single-rod construct. This becomes a very important factor when combining fusionless technology with instrumentation, as the construct will incur continued loading and micro motion. This leaves the implants susceptible to fatigue and mechanical failure. Another benefit of the dual growing rod construct is seen when one of the rods fail. In this instance, one rod remains, maintaining correction and stability, possibly delaying revision until the next planned lengthening procedure.

Another way to avoid further complication is by placing the rod submuscular rather than in the subcutaneous tissues. Subcutaneous rods were placed initially to reduce the risk of spontaneous spine fusion, due to subperiosteal exposure of the spine. It has been demonstrated however that there were more total and wound complications with subcutaneous rod placement. Bess et al. [24] also demonstrated that subcutaneous rods had more implant prominence as well as implant-related unplanned procedures compared to the submuscular dual growing rods. Furthermore, the

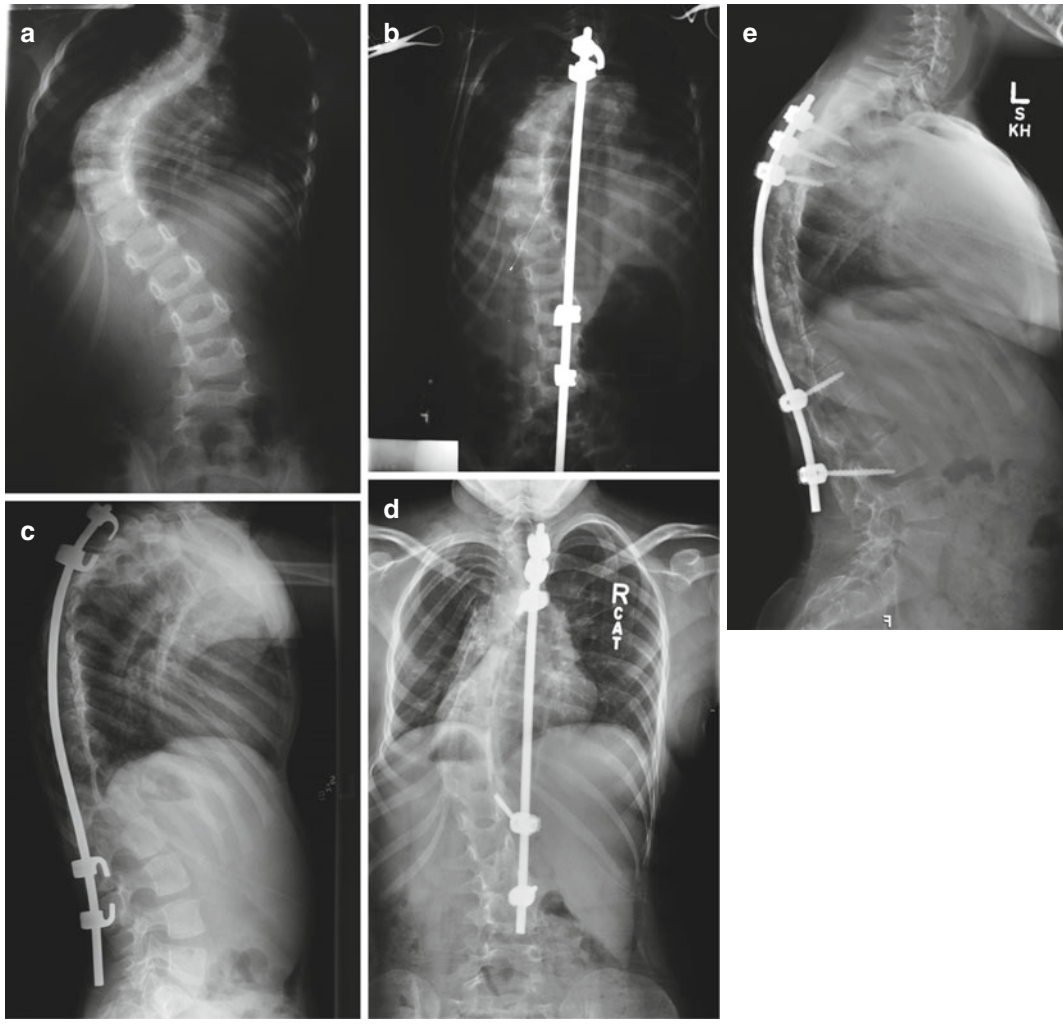

Fig. 38.8 (**a**) Preoperative posteroanterior (PA) radiograph of the patient in Fig. 38.1. She is a 4-year, 2-month-old female with infantile idiopathic scoliosis. She has a major 85° *left* thoracic curve between T6 and L2. (**b**) Postoperative PA radiograph showing that her curve has been reduced to 35°. Note the long length of rod below the lower foundation from which the rod will be lengthened in the future. (**c**) Postoperative lateral radiograph showing her sagittal plane alignment. She has a moderate increase in thoracic kyphosis and lumbar lordosis. (**d**) Anteroposterior radiograph 7 years postoperative at 11 years of age showing her final fusion and correction. Her major curve measures 46°. She has undergone one major revision to all pedicle screw construct. Because of the extensive fusions proximally and distally, a dual-rod system was not inserted. (**e**) Lateral radiographs showing maintenance of sagittal plane alignment. She has maintained her moderate increase in the thoracic kyphosis (72°) and lumbar lordosis (52°)

subcutaneous group had the highest rate of overall wound complications [33]. Unplanned surgery was reduced to the greatest extent in patients with submuscular dual growing rods where the planned to unplanned surgery ratio was 20:1. Patients with single growing rods placed subcutaneously had the worst planned to unplanned surgery ratio of 7.4:1.

In summary, the complication rate remains high, despite advances in technology and an improved understanding of the natural history of early-onset scoliosis. Though complications remain a problem, we have found ways to avoid unplanned surgery. Having the knowledge of the various complications and their incidence will allow us to communicate more effectively with

family and caregivers and allow for a more informed consent.

38.7.1 How to Avoid Complications

Indications: Proper patient selection is the key for improved outcomes. Consider the diagnosis, age, and clinical presentation especially the pulmonary condition. The younger the patient and the more surgical procedures, the greater the possibility of complications. If feasible, surgery can be delayed using nonoperative treatment such as casting, bracing, and sometimes traction.

Technique: When exposing for the placement of foundation, avoid subperiosteal dissection except

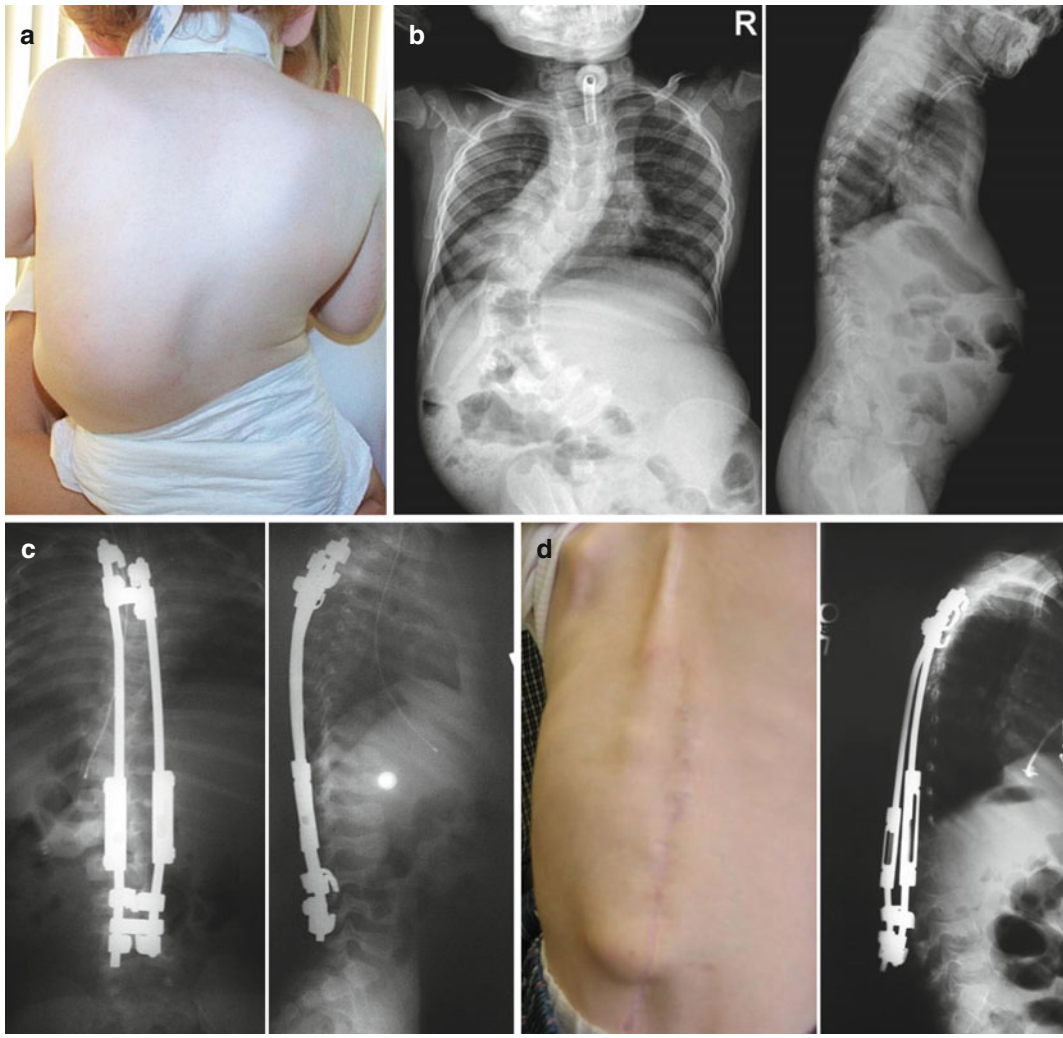

Fig. 38.9 (**a**–**g**) A 30-month-old boy with idiopathic scoliosis (**a**). Anteroposterior and lateral pre-initial surgery radiographs (**b**). The patient underwent dual growing rod insertion from T3 to L4. Hooks were used at upper foundation (T3–T4) and lower foundation (L3–L4) (**c**). During 7 years active treatment, patient had multiple complications, treated through planned/unplanned surgeries. Hooks pull out at lower foundation; hooks were replaced by pedicle screws (**d**). Infection and skin loss in the same patient happened between two lengthenings, treated in an unplanned surgery with irrigation, debridement, and secondary closure (**e**). Latest follow-up radiographs at the age of 10 years; all the hooks were replaced with screws throughout the treatment period (**f**). Clinical photos at the age of 10 and after 7.5 years of active treatment (**g**)

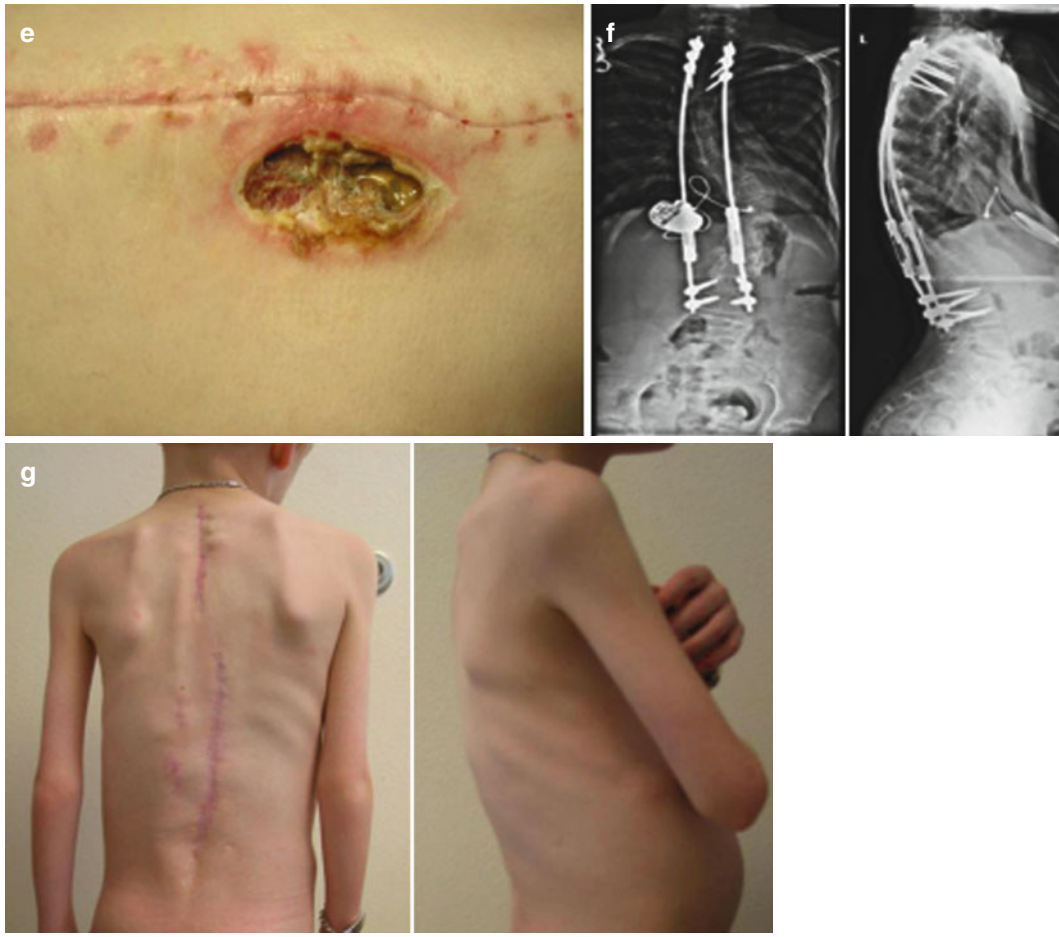

Fig. 38.9 (continued)

in foundation sites. Proper selection of foundation levels, proper position of connectors, and delicate handling of skin coverage are of utmost importance. Frequent lengthening assures maximum growth achievement. If the patient has a rigid curve and/or kyphosis, consider preoperative traction or annulotomy to improve flexibility. It is wise not to try to correct all the sagittal deformity in one sitting. Since the patient is brought to the operating room often for lengthening, the deformity can be corrected by gradual rod contouring to more normal alignment (Fig. 38.10a, b) (Table 38.6).

Complications should be diagnosed and treated early to avoid major catastrophes. Some complications are unavoidable and relates to the natural growth of the child. For example, if a small screw is placed in a very young child, it will be necessary to revise the foundation at some point in the future and should not be considered as complications and should be anticipated and planned. In case of rod breakage, replacement may be a better strategy than connecting two broken rods. If a rod breaks, one can wait until the time of scheduled lengthening and exchange the rods. We recommend changing both the rods if one breaks since the incidence of second rod breakage is significantly higher.

Skin problems and infections should be addressed aggressively to avoid long-term problems by debridement, antibiotics, and healthy skin coverage. Implant exchange is usually not necessary but long-term antibiotics are given until the laboratory work becomes normal.

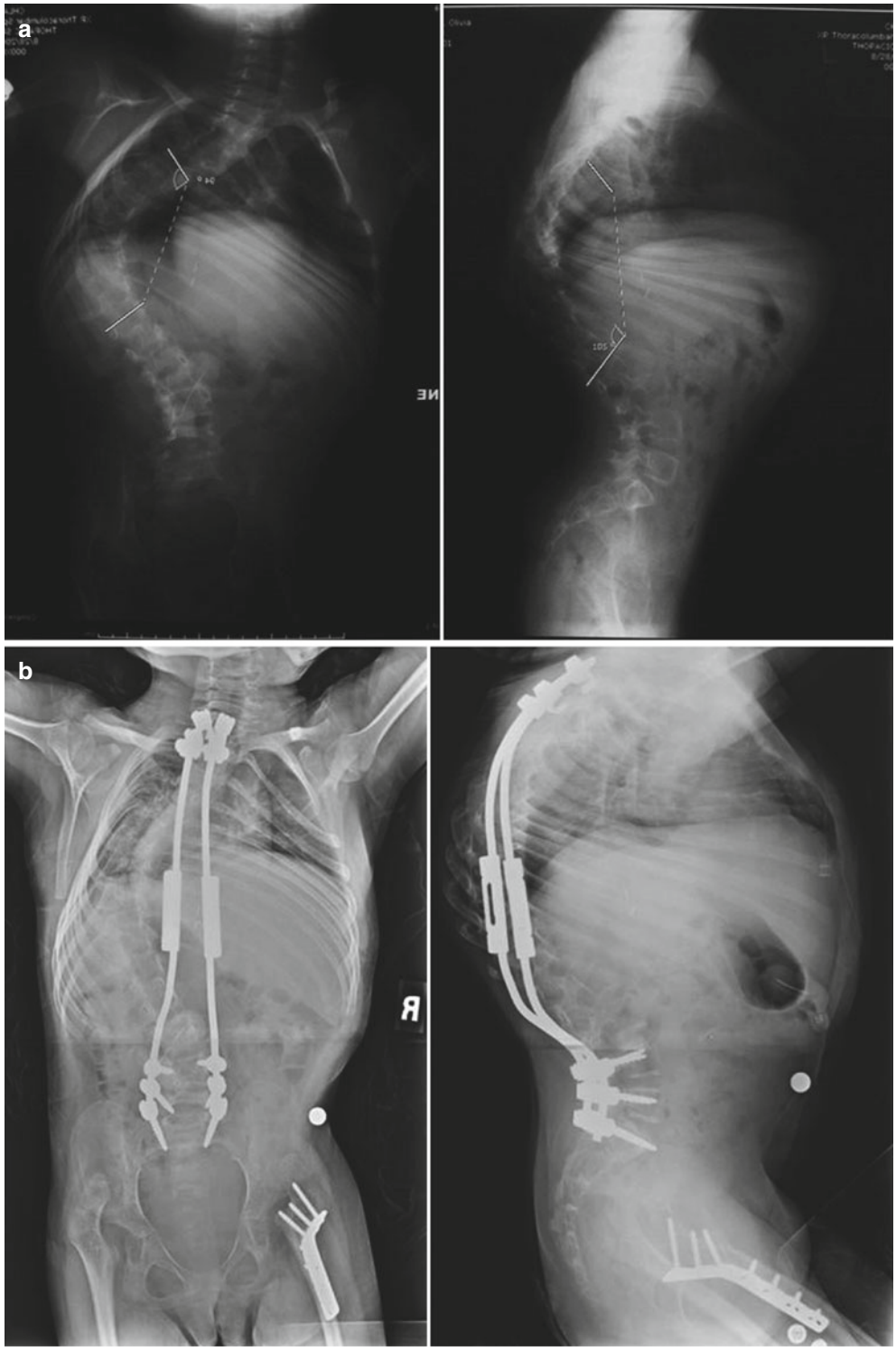

Fig. 38.10 Dual growing rods in a patient with severe kyphoscoliosis. (**a**) Postoperative radiographs show considerable correction after surgery and proper contouring to accommodate kyphosis (**b**)

Table 38.6 Recommendations for minimizing complications

Growing rod pitfalls and avoidance	
Pitfall	Avoidance
Proximal junctional kyphosis	Bend the proximal rods into appropriate kyphosis
	Preserve interspinal ligaments
	Proximal construct should be around T2 or T3; *do not* end construct in kyphotic segment
	Add more support in addition to foundations such as sublaminar wires or tape
Tandem connector problems	Place at thoracolumbar junction (T10–L2, a normally straight area of the spine) if possible and smaller size connectors if used in other spinal segments
Primary surgery failures	When in doubt instrument longer
	Construct usually ends at L3–L5
	Use pedicle screw instrumentation at caudal foundation
	Cephalad foundation uses either pedicle screws or hooks with a cross-link
	In rigid curves, consider preoperative traction and/or annulotomy
	With pelvic obliquity consider instrumentation to pelvis
Growth not occurring as expected	Ensure lengthening every 6 months but consider limitation
Premature fusion	Meticulous surgical technique
	Avoid overexposing the spine
	Apply less invasive techniques
	Frequent lengthenings
Wound problems	Avoid mishandling tissue
	Careful layered closure
	Avoid use of electrocautery near skin
	Use low-profile implants
	If tissue coverage cannot be obtained, involve plastic surgery for coverage

38.8 Results

In 1984, Moe et al. [12] reported on his outcomes after Harrington instrumentation. The major curve improved from 70° preoperatively to 38° at the time of last surgery. They also reported on T1–S1 growth and found a mean growth of 2.9 cm across the instrumented area for all patients and an overall growth of 3.8 cm. The gain achieved during initial curve correction was not counted. Klemme et al. [22] reported on 67 children with progressive scoliosis using a single-rod distraction-based technique. Curve magnitude improved from 67° at initial instrumentation to 47° (30 %) at definitive fusion. Growth across the unfused spinal segments had a mean of 1 cm/year (0.08 cm/segment/year). The mean number of spinal segments instrumented was 13.7.

Tello [34] described his experience with 44 children treated with Harrington instrumentation without fusion. He reported on 12 patients who went on to final fusion with a mean curve correction of 32 %.

Akbarnia et al. [13] in 2005 were the first to report the clinical outcomes following dual growing rod surgery. Their early results included data collected with a minimum of 2 years follow-up after initial surgery. Mean follow-up was 4.02 years with 6.6 lengthenings per patient at an interval of 7.4 months. Mean major curve improved from 82° to 38° after initial surgery and 36° at last follow-up or final fusion. T1–S1 increased by 1.21 cm/year. The SAL ratio among thoracic curves improved from 0.87 to 1.0. Complication rates were reported as 48 %.

In 2008, Akbarnia et al. [26] reported on the results of 13 patients with no previous surgery

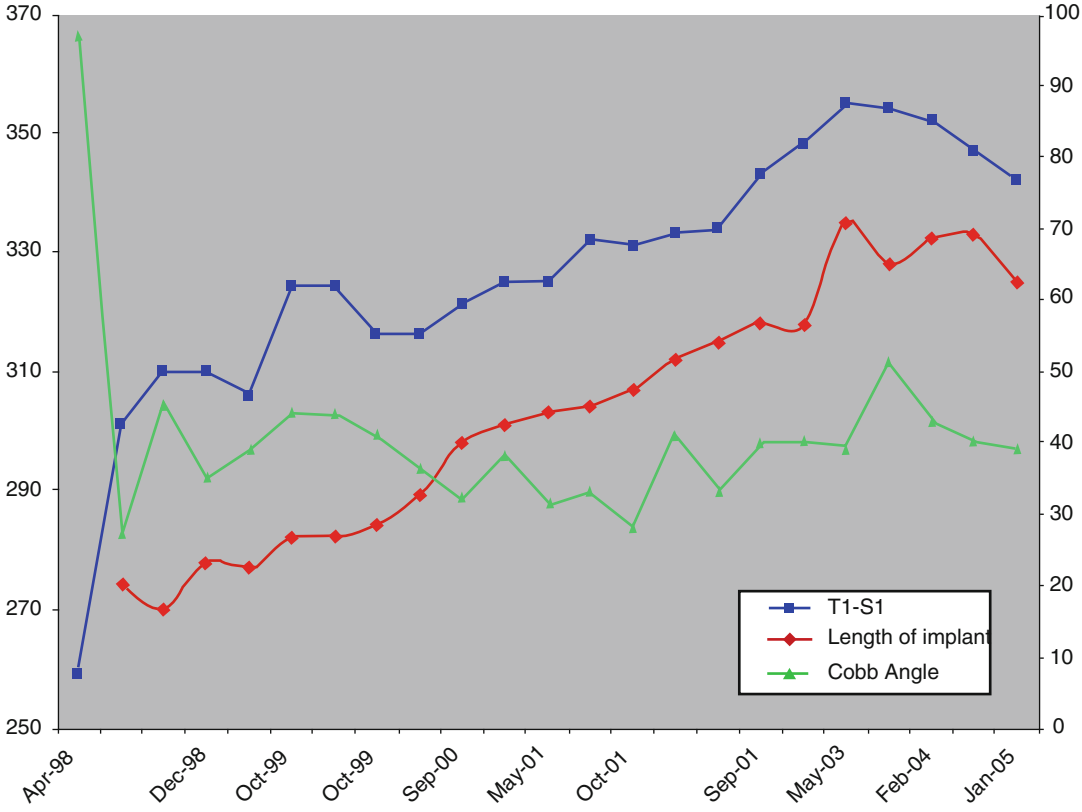

Fig. 38.11 T1–S1 and major curve changes in a 4-year-old patient with Marfan syndrome treated with dual growing rods (7 years follow-up)

and non-congenital curves who underwent dual growing rod insertion and were followed to final fusion. Mean age was 6.6 years at initial growing rod surgery and 11 years at final fusion. Patients underwent a mean of 7.8 surgeries including initial growing rods and final fusion. Major curve improved from a mean of 81° pre-initial to 36° postoperatively and 28° after final fusion. The patients underwent on a mean of 5.2 lengthenings at an interval of 9.4 months. Mean growth was 1.46 cm/year for a total of 5.7 ± 2.9 cm over 4.37 ± 2.4 years. When analyzing a cohort of children with more frequent lengthenings (≤6 months), he found a statistical improvement in growth rate (1.8 cm vs. 1 cm/year) and curve correction (79 vs. 48 %) (Fig. 38.11).

Sponseller et al. [35] recently reported the outcome of growing rods fixed to the pelvis. He included 36 patients, of which 30 were dual growing rods. A cross-link was used in the dual iliac fixation group to provide improved construct stability. Overall, there was significant improvement in coronal and sagittal balance. Among the six patients with final fusion, mean gain in T1–S1 was 8.6 cm, of which 4 cm occurred during the lengthening period. The dual iliac fixation group has a statistical advantage over single-rod fixation regarding correction of deformity (47 vs. 25 %) and pelvic obliquity (67 vs. 44 %). Iliac screws also showed overall superiority compared to sacral fixation with regard to correction of major curve and pelvic obliquity. Pelvic fixation, regardless of the technique, resulted in greater percentage improvement in pelvic obliquity than in correction of major curve. All the 12 patients who were expected to ambulate given their neurological status were able to do so after surgery.

There are several points worth reiterating regarding improved outcome in patients who

undergo dual growing rod surgery. The initial surgical treatment of scoliosis is by far the most important procedure to predict long-term success. This includes choosing which growing rod construct to use and levels to be included. In general, the upper thoracic spine is used as the proximal foundation, and caudally, the levels are chosen based on curve pattern but, in general, span the thoracolumbar junction (L2 or below). More frequent lengthenings (≤6 months) do seem to lead to more correction and improved radiographic outcomes but with a higher number of complications. Complication rates remain high; however, dual growing rods lead to fewer unplanned surgeries, with most complications addressed at routine lengthening. Pelvic fixation can be safely applied to a growing rod in cases where distal fixation is appropriate. Dual iliac screws with a caudal cross-link have the best outcome.

38.9 Discussion

The current results indicate that the use of both single and dual growing rods can be considered a useful adjunct in the surgical management of severe progressive spinal deformities in early-onset scoliosis. EOS represents one of the most complex conditions challenging the pediatric spine surgeons. These patients have a variety of underlying etiologies in addition to their spine and chest wall abnormalities that add to the complexity of their management. Many of these children die in their infancy if untreated. It is well known that even if they survive childhood, their life expectancy is much shorter. Furthermore, their quality of life is significantly impaired and may be even lower than that of children afflicted with asthma, heart disease, or childhood cancer [36]. Traditional methods for correcting spine deformity have included spinal fusion. This method is not appropriate for young children since it results in short spine and chest and lung underdevelopment and often associated with an increase in their spinal deformity. Over the past decade, there has been a renewed interest for improving the care of children with EOS. The goal of these new treatment options is to improve the care of these children. There are obstacles, however, from lack of evidence-based research and significant variability of treatment methods once the indication for treatment is established.

There has been increasing interest in the development of new technique and devices for surgical treatment of patients with EOS. Furthermore, there has been favorable legislature passed by US congress such as "The Pediatric Medical Device Safety and Improvement Act of 2007" which has improved the regulatory process; however, until recently, most of the new growth-friendly techniques for the treatment of patients with EOS involved off-label use of pediatric devices. There are also obstacles for proving the effectiveness of methods since it requires evidence-based studies that cannot be easily accomplished in this population.

Recent attempts have led to the establishment of Growing Spine Committee of Scoliosis Research Society and EOS Study Groups conducting multicenter studies. There is also a combined SRS/POSNA task force on pediatric devices that has been working with FDA in the USA to help the clearance for pediatric devices.

38.10 Future Direction

There has been significant progress in recent years in our understanding of the natural history and outcomes of treatment in children with EOS. However, there exists a great deal of variation in indications as well as treatment methods [37, 38]. This is partially due to a lack of standardized methods of categorizing the heterogeneous EOS population and difficulties in evaluating outcomes. The research by study groups using multicenter clinical information should help standardizing the data collection, management, and conduct quality research leading to improved quality of care for EOS patients.

The recent enthusiasm in developing new technology and devices such as remotely controlled distraction-based mechanisms [25, 39–41] will lead to less invasive procedures and minimize the complications associated with the current techniques (see Chap. 47).

Finally, to assess the effect of the treatment methods on the natural history of EOS requires long-term follow-up of these patients with the collection of meaningful clinical and other important data. We have come a long way in 50 years but certainly much more to learn in the next 50 years.

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VEPTR Expansion Thoracoplasty

39

Robert M. Campbell

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Key Points

- VEPTR expansion thoracoplasty procedures treat volume depletion deformities of the thorax which cause thoracic insufficiency syndrome.
- Thoracic insufficiency syndrome is the inability of the thorax to support normal respiration or lung growth.
- Conditions such as congenital scoliosis and fused ribs, infantile and juvenile idiopathic scoliosis, myelomeningocele, and other congenital anomalies/deficiencies of the chest wall can be addressed by VEPTR expansion thoracoplasty without inhibiting growth of the spine.
- Advances in imaging techniques such as dynamic MRI of the lungs will enable a better understanding of the biomechanical deficits of the thorax in thoracic insufficiency syndrome.

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39.1 Introduction

39.1.1 Thoracic Insufficiency Syndrome: Anatomic Basis

Thoracic insufficiency syndrome (TIS) [1] is the inability of the thorax to support normal respiration or lung growth. It is the primary indication for treatment by VEPTR (vertical expandable prosthetic titanium rib) expansion thoracoplasty. The natural history of TIS can be lethal in cases of untreated early-onset spine and chest wall deformity, but overall it is just beginning to be understood [2].

The thorax is a complex, dynamic chamber of respiration that both supports and rhythmically expands the lungs during breathing. Structurally, the thorax consists of the spine, as its posterior pillar, the rib cage [3], the sternum, and the diaphragm. As the respiratory pump [1], it must provide normal, stable volume for the underlying lung through rigidity of the chest wall, as well as the ability to change that volume. These are termed the two thoracic characteristics of breathing [1].

Abnormalities of the thorax, including congenital structural problems of the thoracic spine, the rib cage, and the diaphragm, result in *primary* thoracic insufficiency syndrome. The diaphragm may also be compromised unilaterally or bilaterally in *secondary* thoracic insufficiency syndrome [4] when there is collapse of the torso inferiorly through either lumbar kyphosis in myelomeningocele or when there is pelvic obliquity due to thoracolumbar scoliosis, both causing a relative obstruction to excursion of the diaphragm. Clinically these children have a Marionette's sign with the patient's head bobbing synchronously with respiration with the diaphragm, in effect, doing a push up against body weight [4].

The only practical aspect of TIS that is treatable is the volume reduction of the thorax. VEPTR expansion thoracoplasty can enlarge a constricted thorax, either unilaterally or bilaterally, with the assumption that the underlying diaphragm can make use of the new volume of lung that goes on to fill the expanded thoracic volume, but such an approach cannot restore chest wall motion, and it is not known whether such procedures enhance diaphragmatic function. The first step in VEPTR treatment is to classify the thoracic volume depletion deformity so that the proper VEPTR surgical strategy is chosen.

39.1.2 Volume Depletion Deformities of the Thorax

Three-dimensional thoracic deformity is defined in the three anatomic planes: coronal, sagittal, and transverse. These are as volume depletion deformities *(VDD)* of the thorax [2] (Table 39.1, Fig. 39.1). The VEPTR expansion thoracoplasty strategy for each volume depletion deformity of

Table 39.1 Thoracic volume depletion deformities

Type of volume depletion deformity	Thoracic deficit	Mechanism of lung volume loss	Examples
I. Absent ribs and exotic scoliosis	Unilateral thoracic hypoplasia	Lung prolapses into the chest with volume loss	VATER, absent ribs, and congenital scoliosis
II. Fused ribs and exotic scoliosis	Unilateral thoracic hypoplasia	Constriction of lung due to fused ribs shortening hemithorax	VATER, fused ribs, and congenital scoliosis, thoracogenic scoliosis from prior thoracotomy
IIIa. Foreshortened thorax	Global thoracic hypoplasia	Bilateral longitudinal constriction of lungs from loss of thoracic height	Jarcho-Levin syndrome
IIIb. Transverse constricted thorax	Global thoracic hypoplasia	Lateral constriction of lungs from rib deformity	Jeune's asphyxiating thoracic dystrophy, windswept deformity of the thorax in scoliosis

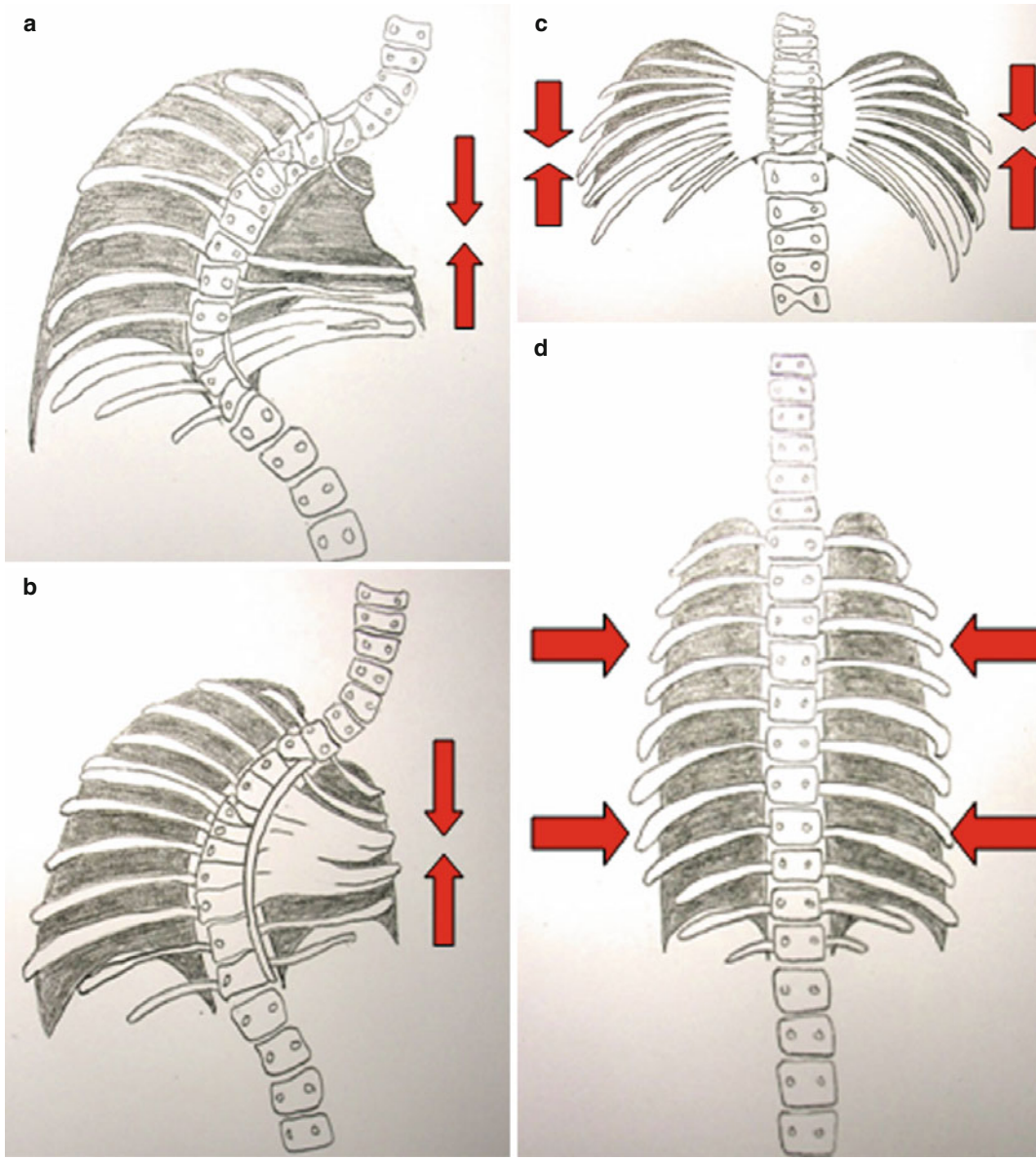


Fig. 39.1 (**a**) A type I thoracic volume depletion deformity: absent ribs and scoliosis. (**b**) A type II thoracic volume depletion deformity: fused ribs and scoliosis. (**c**) A type IIIa volume depletion deformity of the thorax: spondylothoracic dysplasia (Jarcho-Levi syndrome). (**d**) A type IIIb volume depletion deformity of the thorax: Jeune's asphyxiating thoracic dystrophy

the thorax is different. In mixed types of volume depletion deformity, VEPTR surgical treatment should address each individual segment of thoracic deformity with either appropriate longitudinal or lateral expansion of the constricted thorax.

39.1.3 FDA Indications for VEPTR Expansion Thoracoplasty

- *Presence of thoracic Insufficiency syndrome*
- *Skeletally immature patient*

Anatomic Diagnosis

- *Absent ribs*
- *Constrictive chest wall syndrome, including fused ribs and scoliosis*
- *Hypoplastic thorax*
- *Early-onset scoliosis of congenital or neurogenic origin without rib anomaly*

39.2 VEPTR Preoperative Assessment

39.2.1 Clinical Examination

It is important to obtain a detailed history in children with thoracic insufficiency syndrome: when was the onset of clinical deformity, what were past surgical treatments, and are there associated morbidities such as renal, gastrointestinal, central nervous system and cardiac system abnormalities? A good respiratory history should be taken to note past episodes of pneumonia, bronchitis, or asthma attacks or needs for respiratory support during illness. If the patient is on oxygen or dependent on more invasive respiratory support, the degree of respiratory insufficiency should be defined by the assisted ventilator ratings (AVR) [2, 5].

AVR Ratings

+0: no assistance, on room air
+1: supplemental oxygen required
+2: nighttime ventilation/CPAP
+3: part-time ventilation/CPAP
+4: full-time ventilation.

An increase in AVR suggests progressive clinical respiratory insufficiency, and this is a strong indication for treatment. Pulmonary function tests are practical in children age 5 years or older [6], so past testing, if available, would be helpful in determining any deterioration of vital capacity as determined by decreasing percent normal vital capacity. Clinical history, noting the child's ability to respond to pulmonary challenge such as play activities and running, can also be helpful.

On physical examination respiratory rate is assessed. Normal respiratory rate at birth is 40–80 breaths per minute and, up to age 5 years, 20–40 breaths per minute, with 15–25 breaths per minutes being normal from age 6–12 years, and adult values, 15–20 breaths per minute, are reached after age 15 years of age [6]. Respiratory rate at rest above these values suggest occult respiratory insufficiency [1]. The chest is assessed for clinical deformity and the circumference measured at the nipple line and compared to normal values for age to discern percentile normal [7].

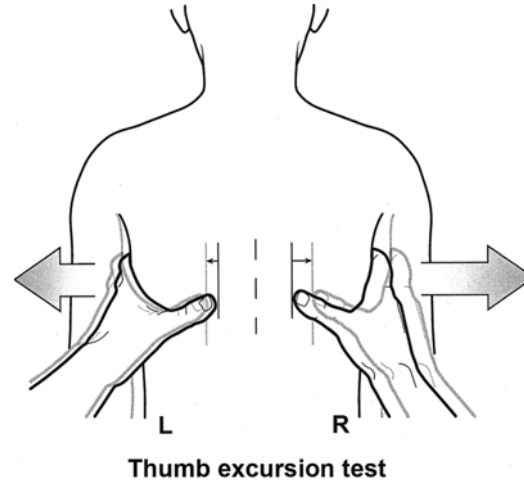


Fig. 39.2 The thumb excursion test

The thumb excursion test [1] is performed to clinically measure the ability of each side of the chest to contribute to respiration by rib cage expansion. In this test, the examiner's hands are placed around the base of the thorax with the thumbs posteriorly pointing upward at equal distances from the spine (Fig. 39.2). With respiration, the thumbs move away from the spine symmetrically because of the anterior lateral motion of the chest wall. Greater than 1 cm excursion of each thumb away from the spine during inspiration is graded as +3, and this is normal, 0.5–1 cm excursion is graded +2, motion up to 0.5 cm is graded as +1, and complete absence of motion is graded +0. Each hemithorax is graded separately. "Collapsing torso" deformities, resulting in secondary thoracic insufficiency syndrome, may raise pressure on the diaphragm by proximity to the pelvis and are assessed by presence of the Marionette sign [4]. Both the lips

and fingertips are examined for any signs of cyanosis and fingertips for evidence of clubbing, suggesting long-term clinical hypoxia.

39.2.2 Imaging Studies

Imaging studies should include weight-bearing AP/lateral radiographs of the entire spine, including the chest and pelvis, on the same radiograph. The radiograph is analyzed for Cobb angle, the height of the thoracic spine in centimeters, and the space available for lung [1]. The height of the thorax is determined by the radiographic height of the patient's thoracic spine, and this distance is divided by the normal thoracic spinal height for age [8], deriving a percentage normal. The lateral radiograph defines a loss of sagittal depth of the thorax either due to pectus excavatum or thoracic spinal lordosis.

CT scans of the entire chest and lumbar spine are performed at 5 mm intervals, unenhanced [9], with the scanner set for pediatric dosage to minimize radiation exposure [10, 11]. These provide CT lung volumes [12, 13] and anatomy details of chest and spine. Full chest CT scans may be taken at yearly follow-up if percent normal lung volumes are being followed to detect progressive thoracic volume loss. Both ventilation perfusion lung scans and 3 mm cut CT scans with airway reconstruction can define airway compression deformity, if necessary. All patients should also undergo MRI studies of the entire spinal cord to rule out spinal cord abnormalities. Either ultrasound or fluoroscopy of the diaphragm can be performed to document diaphragmatic function, but dynamic MRI study of the lungs (Fig. 39.3) will show great detail of diaphragm and chest wall function [14].

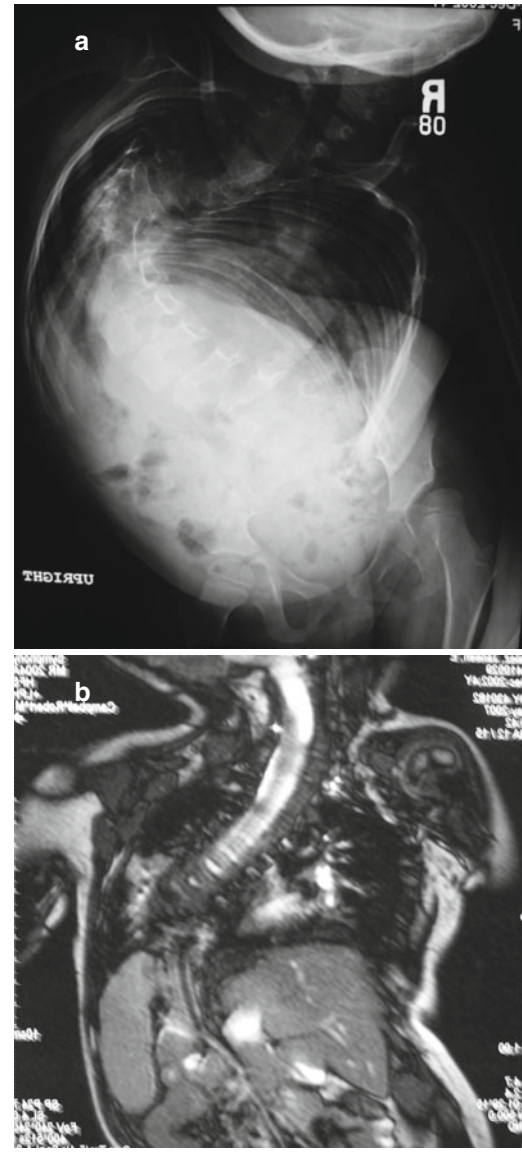


Fig. 39.3 (**a**) Severe scoliosis. (**b**) AP dynamic lung MRI of the thorax. Note intrusion of the liver into the chest from the iliac crest malposition, obstructing diaphragmatic motion

39.2.3 Specific Cardiopulmonary Studies

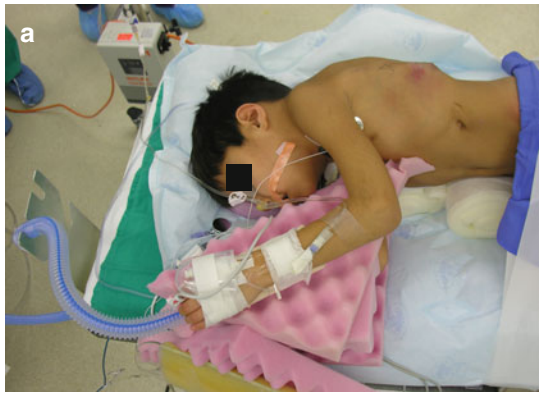
Routine spirometry pulmonary function studies are feasible for children age 5 years or older, and infant pulmonary function tests can be performed in younger patients, if available. When there is spinal deformity present, care must be taken to use arm span instead of height for normalization of pulmonary function test results. Pulse oximetry studies are useful to detect significant amount of hypoxia. When there is question of early cor pulmonale, echocardiograms are performed to detect tricuspid valve regurgitation.

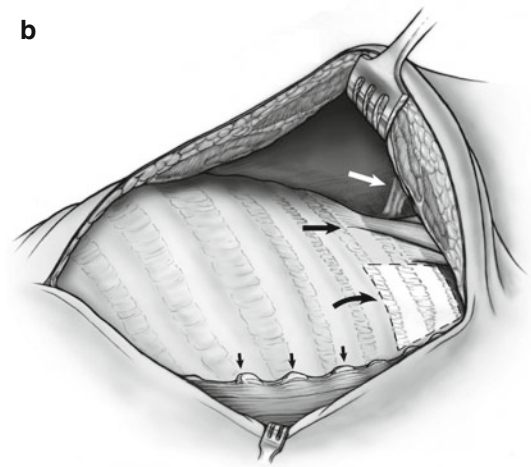
39.3 VEPTR Expansion Thoracoplasty Treatment Strategies

The VEPTR I and II devices are made by DePuy Synthes Spine Company of Raynham, MA, and is recently cleared as 510(K). Multiple types of expansion thoracoplasties have in common the ability to enlarge the constricted area of the hemithorax with the goals of restoring thoracic volume, stability, and symmetry. VEPTR procedures may be used in patients as early as 6 months and up to skeletal maturity. Contraindications for VEPTR treatment include an age of skeletal maturity. Another contraindication is poor rib bone stock or proximal absence of ribs for VEPTR attachment. In proximal rib absence, rib autografts and the use of a longitudinally osteotomized clavicle as a vascularized pedicle graft may provide a bony "first rib" for VEPTR attachment. Severe comorbidities that make repetitive surgeries impractical are also a relative contraindication. Soft tissue coverage is critical for VEPTR success, but commonly children with respiratory insufficiency have a calorie deficit from the work of tachypnea and may have percent normal body weight of less than 5 %. Diet supplements, or even G tube therapy, may be necessary to increase the soft tissue coverage for VEPTR implantation, and a minimum percent normal body weight of 25 % is recommended before proceeding to surgery. Poor lung function of itself is not a contraindication for VEPTR treatment.

39.3.1 Surgical Technique: General Approach

The patient is placed in a prone position (see Fig. 39.4a). Spinal cord and upper extremity status are monitored by both somatosensory evoked potentials and motor evoked potentials. A central arterial line is placed. Prophylactic IV antibiotics are given and maintained for 5 days or until drains are out. A modified curvilinear thoracotomy incision is used, extending anteriorly between the ninth and tenth rib. Once the chest wall flap is elevated, the common insertion of the middle and posterior scalene muscles is identified in order to determine the location of the neurovascular bundle just anterior to it. After complete exposure of the rib cage, the paraspinal muscles are next reflected by cautery medially up to the tips of the transverse processes of the spine. Care must be taken not to expose the spine in order to prevent inadvertent fusion. The underlying chest wall deformity is then assessed for the degree of instability, constriction of underlying lung by rib fusion, anomalous insertion of the ribs into the spine, and sites for device placement (see Fig. 39.4b). Surgical strategy depends on the specific type of volume depletion deformity to be addressed: unilateral constriction of the thorax is addressed by an expansion thoracoplasty, termed an opening-wedge thoracostomy. The lengthened hemithorax is then stabilized by a hybrid VEPTR device from proximal ribs to lumbar spine [15], sized so that the rib sleeve does not extend below the inferior end plate of T12. Hybrid devices are always inserted in a proximal to distal direction to avoid penetrating the chest and causing cardiopulmonary injury. In patients younger than 18 months, hybrid devices are impractical because of inadequate spinal canal width for a spinal hook, so a single rib-to-rib VEPTR is used instead. When such a child reaches age 2 or 3 years, the rib-to-rib device can be easily converted to a hybrid device which controls scoliosis better than the rib-to-rib devices. If space permits, a second device, rib to rib, is added laterally in the posterior axillary line to load share. If there are areas of chest wall instability more anteriorly, additional longitudinal rib-to-rib VEPTR devices are implanted as needed with care to place them well below the neurovascular bundle (see Fig. 39.4c). Once the thoracic reconstruction has been completed and VEPTR devices are in place, then the combined muscle and skin flapped are stretched to provide increased soft tissue coverage for the expanded hemithorax. The thorax should be equilibrated as much as possible in all planes, increasing space available for lung [1] on the concave side to 100 %, with symmetrical hemithorax width on radiograph and symmetrical hemithorax volumes in the transverse plane on the CT scan.





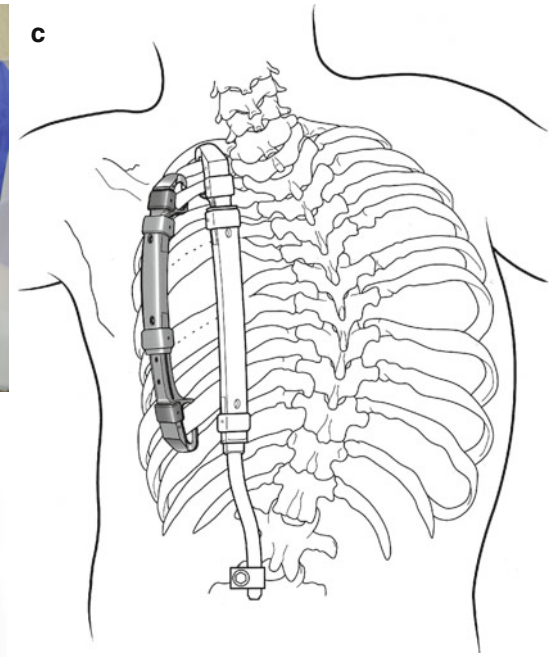
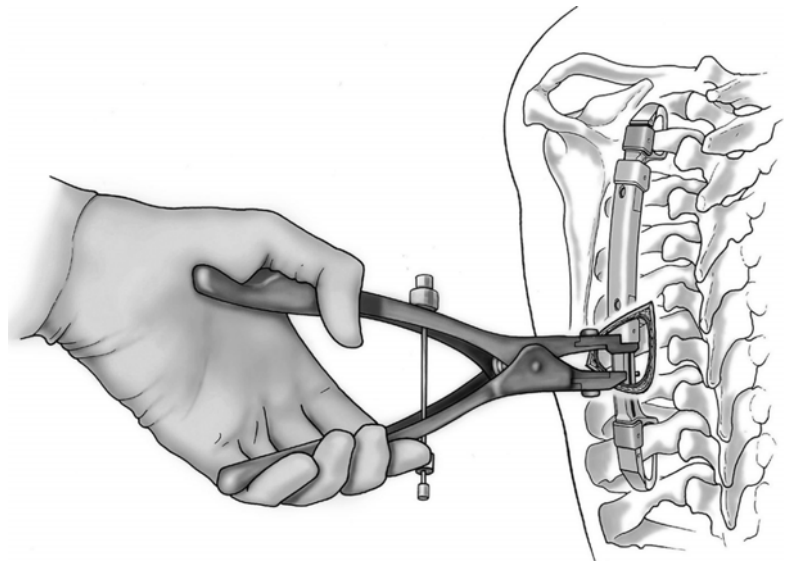


Fig. 39.4 (**a**) General patient positioning for VEPTR thoracoplasty. (**b**) To avoid damaging the neurovascular bundle in the VEPTR exposure: *white arrow* notes neurovascular bundle; hatched area is the safe zone for superior cradle attachment; *small arrows* show tips of the transverse processes. (**c**) Standard VEPTR construct

For closure, the scapula is first brought distally to the approximate anatomic position, and the pulse oximeter reading on the up arm and somatosensory evoked potentials are checked for signs of acute thoracic outlet syndrome. Patients with very anomalous proximal ribs, distracted into the area of the brachial plexus by VEPTR expansion thoracoplasty, are at risk for this and early signs are decreased in ulnar nerve tracings and diminished pulse. Usually relaxation of the position of the scapula, allowing a more proximal position, resolves this problem. If continued alterations in pulse oximeter and/or spinal cord monitoring are encountered, even with relaxation of the closure, it may be necessary to resect the anterolateral portion of the first and second rib, lateral to the devices, in order to provide clearance for the brachial plexus in the reconstructed thorax.

Two subcutaneous Jackson-Pratt drains are used. In patients when there is substantial defect in the pleura, greater than 4 cm, it is repaired with Surgisis® by Cook Medical [16]. Patients can be extubated in OR if doing well anesthetically or can be left intubated 24–72 h. The hematocrit is checked daily for 3 days. Although blood loss usually averages 50 ccs [4], continual oozing underneath the large flaps results in a 50 % risk for postoperative transfusion. Generally, a hematocrit of 30 % or greater is optimal for oxygen-carrying capacity for these patients. Fluid management should be on the restrictive side to prevent acute pulmonary edema.

Fig. 39.5 VEPTR expansion



Once weaned off the ventilator, the patient can be transferred to the surgical ward. Jackson-Pratt drains are removed when their individual drainage decreases to 20 cc or less over a 24-h period. Chest tubes are removed once their drainage equals 1 cc per kilogram of patient weight over 24 h. If the patient goes into respiratory distress after drains and chest tubes are removed, consider checking for acute reaccumulation of the pleural effusion with compression of the lung. Temporary chest tube drainage can address this through placement of an anterior "pigtail" chest tube. Vigorous pulmonary toilet, including percussion, is needed post operatively. The patients are mobilized as soon as possible. No bracing is used because of the potential constrictive effects. Specific postoperative care is detailed in other reports [15, 17].

39.3.2 VEPTR Expansion Procedures

Twice to three times a year, the devices are expanded under general anesthesia to accommodate growth of the patient [15]. Spinal cord monitoring is used for expansion procedures as well as for replacement procedures. Prophylactic IV antibiotics are given and maintained for 24 h. Each individual device is accessed by a 3 cm incision, with care taken to preserve a thick muscle flap over the devices by meticulous soft tissue technique in order to minimize the risk of skin slough. If the distraction lock is exposed through the thoracotomy incision, a freer elevator is inserted proximally along the top of the device and used to elevate the overlying muscle. Cautery is inserted into the soft tissue tunnel created by the freer elevator and is used to release the muscle deeply on each side of the device so that a thick muscle flap is mobilized with the free edge at the skin incision. The same approach is used distally. When the skin incision parallels the device, the muscle incision is made by cautery *along the side of the device* at the distraction lock site of the rib sleeve, then the cautery is turned sideways to release the muscle flap off the device (Fig. 39.5). The full thickness muscle flap is reflected by a freer elevator, the distraction locks of the device are removed, and the expansion procedure is performed. When there is a medial device, usually a hybrid, extending from proximal ribs down to lumbar spine, it is first expanded until the reactive force increases substantially and then the device is locked with a new distraction lock at its new length. The adjacent devices are then expanded approximately half that distance, the distraction lock replaced and locked into its new length. When there are bilateral devices to expand, first the concave hemithorax is expanded and locked and then the devices on the convex side are expanded and

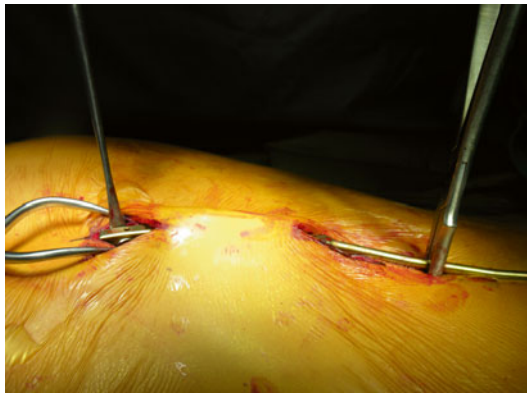


Fig. 39.6 VEPTR replacement through "skip" incisions

locked. The mobilized muscle flaps are closed without tension over the locks when device expansion is complete.

39.3.3 VEPTR Replacement Procedures

Once completely expanded, change out of the central rib sleeve portion and the inferior cradle is needed. This is usually accomplished through a limited access, central incision at the distraction point , a small incision over the superior cradle, and then a third incision over the lumbar hook or the inferior cradle [15] (Fig. 39.6). Prophylactic IV antibiotics are given and maintained for 3 days or until any drains are out. The device is unlocked from the spinal hook and the superior cradle, removed, and then replaced with a longer device. The new device is locked into place and then tensioned, much as is done during an expansion procedure.

39.4 Specific VEPTR Surgical Strategies

39.4.1 Type I Volume Depletion Deformity: Rib Absence and Scoliosis

The stabilization VEPTR expansion thoracoplasty for a type I VDD is performed through the usual thoracotomy incision with the goal of lateral and longitudinal expansion of the underlying collapsed hemithorax with stabilization of flail segment [5]. Care must be taken not to damage the lung when the skin incision is over the chest wall defect, and generally there is a large spine defect in the area of the chest wall defect, so care must be taken not to violate dura in the exposure. The initial VEPTR device is commonly placed adjacent to the spine. The first step is implantation of the superior rib cradle. Commonly it is attached to the proximal ribs above the chest wall defect, either on the bottom rib of the rib cluster or on a more proximal rib. In the latter case, a 1 cm incision is made by cautery in the intercostal muscle, immediately beneath the rib of attachment. Next a Freer elevator is then inserted, pushing through the intercostal muscle to the lower edge of the rib, stripping the combined pleura/periosteum layer off from the rib anteriorly. A second portal is the placed by cautery above the rib of attachment. A second Freer is inserted in this portal, pointing distally to strip off the periosteum of the rib anteriorly, and the two Freers should touch in the "chopstick" maneuver [4], to confirm that a continuous soft tissue tunnel has been made. The VEPTR trial instrument is then inserted into the portals to enlarge them superiorly and inferiorly. At least 1 cm of bone should be encircled by the superior rib cradle. If the rib chosen is too slender, then two ribs are encircled with an extended cradle cap added to the construct in order to encircle it. The rib cradle cap is inserted by forceps into the superior portal, facing laterally, to avoid the great vessels and the esophagus and then turned distally. Next, the superior rib cradle is then inserted into the inferior portal, mated with the cradle cap, and attached with a cradle cap lock. The superior cradle is gently distracted by forceps superiorly to test for instability. If unstable, the superior cradle can be moved another level distally to a stronger rib for attachment. Superior cradle insertion is similar when the attachment rib has fibrous adhesions instead of intercostal muscles linking it to the ribs above and below. When the superior cradle needs to be placed within a mass of fused ribs, however, then the inferior portal for the superior cradle is created by a bone burr, creating

a slot 5 mm by 1.5 mm, and a 5 mm superior portal is cut by burr for placement of the cradle cap.

An opening-wedge thoracostomy is then performed through the flail segment of the chest, usually requiring release of the fibrosed pleura, and then the proximal ribs are distracted upward so that they become horizontal in orientation. With the rib distractors holding the opening-wedge thoracostomy open to the corrected position, a correct length VEPTR rib sleeve and inferior cradle is then attached to the previously placed rib cradle and extended down to a stable rib near the inferior margin of the thorax, commonly the ninth or tenth rib.

When there is significant scoliosis, a hybrid VEPTR from rib to spine is often needed. With the rib distractor left in place to continue to lengthen the constricted chest wall through the thoracostomy, a separate paraspinous skin incision, 5 cm long, is then made 1 cm lateral to the midline at the level of the proximal lumbar spine (see Fig. 39.4a). A flap is elevated medially to expose the midline of the spine. Cautery is used to longitudinally section the apophysis of the two posterior spinous processes at the correct interspace and a Cobb elevator used to strip the spine laterally. The ligamentum flavum is then resected and the laminar hook inserted. Gelfoam is placed over the exposed dura. A bone block of autograft, usually from rib resection, is then placed from the superior lamina to the top of the hook, anchoring it with a single level fusion. Next, the size of hybrid lumbar extension rib sleeve needed is determined by measuring from the bottom of the rib of attachment encircled by the superior rib cradle down to the endplate of T12. This can usually be estimated by palpating the 12th rib clinically. The distance in centimeters should correspond to the number inscribed on the rib sleeve and the hybrid lumbar extension. The hybrid device is assembled and locked with a distraction lock. To estimate the proper length, the device is then placed into the field with the rib sleeve engaged into the implanted superior cradle proximally and the spinal rod marked by a skin marker approximately 1.5 cm below the bottom of the spinal hook. The hybrid is removed from the field and the rod cut smoothly by a rod cutter. Avoid using a bolt cutter because the resulting sharp edges may cut through the overlying soft tissues. The end of the rod is bent into slight lordosis and valgus by a French bender so that the rod will line up with the axis of the spine after implantation and conform to the lordosis of the lumbar spine.

Next a subfascial canal is created for safe passage of the sized lumbar hybrid extension. A long Kelley clamp is threaded from the proximal incision, through the paraspinal muscles, into the distal incision, with care taken not to violate the chest wall and the pericardium. A #20 chest tube is then attached to the clamp, and the tube pulled upward into the proximal incision. The end of the rod of the hybrid is then placed into the chest tube and the device carefully guided through the muscle by the chest tube into the distal incision. The tube is removed, and the rod threaded into the hook and then upward into the superior cradle. A distraction lock engages the superior cradle to the rib sleeve.

To perform the initial tensioning of the device, a DePuy Synthes C-ring is attached to the rod just above the hook, and a VEPTR distractor used to distract the device from the hook through the C-ring. The hook is then tightened. The rib distractor is then removed from the thoracostomy. If there is adequate distraction from the hybrid device, then the proximal ribs should remain horizontal, and the combined corrected opening-wedge thoracostomy and rib defect interval should be maintained. Additional VEPTR devices are then placed at a 3–4 cm intervals anteriorly, much like a "picket fence," to provide expansion for the thorax and stability for the underlying lung (Fig. 39.6). Proximally these additional devices should be placed well below the neurovascular bundle to avoid compression. To provide extra stability for the chest wall, a segment of distal ribs may be osteotomized apart inferiorly and then rotated upward than tied by nonabsorbable suture to the VEPTRs.

If there is scoliosis extending into the lumbar spine, or if there is considerable pelvic obliquity, then the hybrid can extend down to the iliac crest with attachment by an S-hook. This is termed an "Eiffel Tower" construct because the force vectors from iliac crests to proximal ribs have an

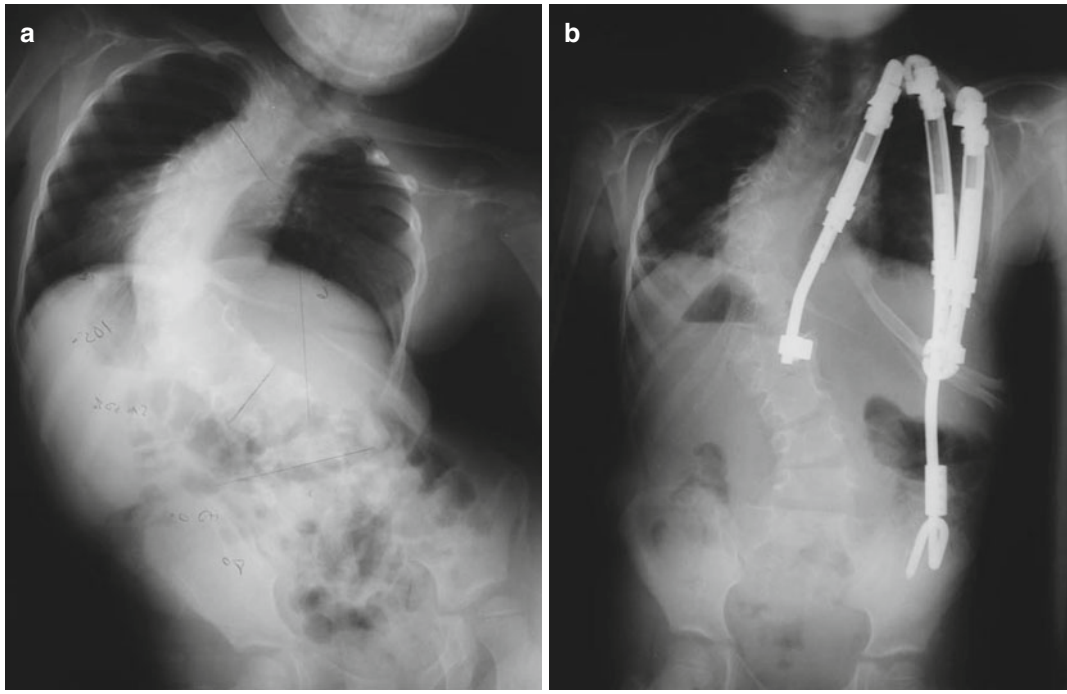


Fig. 39.7 (**a**) Preoperative x-rays of a rib absence patient. (**b**) Postoperative x-rays VEPTR procedure for rib absence

inferior upward and central orientation. The S-hook attachment to mid iliac crest is termed "iliac crest pedestal" fixation. This construct is also a powerful means to address pelvic obliquity [18] (Fig. 39.7).

Mal-insertion of the hemidiaphragm, usually attached too far proximally, is treated by circumferential release, with transposition distally to the most inferior circumference of the reconstructed thorax to provide both additional volume and also a better biomechanical dome shape.

39.4.2 Type II Volume Depletion Deformity: Fused Ribs and Scoliosis

The VEPTR expansion thoracoplasty for this volume depletion deformity is an opening-wedge thoracostomy [4, 15, 17]. The approach is very similar to that for absent ribs, but the volume depletion deformity of fused ribs commonly requires a transverse osteotomy at the apex of the constricted hemithorax from the transverse processes of the spine to the rib costochondral junction. A superior cradle anchor for the VEPTR device is first placed in an appropriate stable rib in the proximal segment of the constricted hemithorax, just lateral to the tip of the transverse process. The most proximal placement possible is the second rib; more proximal rib cradle location endangers the brachial plexus. Two ribs may be encircled by the cradle for enhanced stability. The opening-wedge thoracostomy is then performed. If there is a large bone plate of fused ribs, the section is cut apart transversely from anterior to posterior by a Kerrison rongeur. A no. 4 Penfield elevator is threaded underneath the line of bone resection to protect the lung. Sometimes an adjacent line of fibrosis between ribs just above or below the middle of the bone plate is identified and this can also be used as the cleavage point for the opening-wedge thoracostomy. If there is solid bone extending medially from the tip of the transverse process down to the spine at the posterior point of the thoracostomy, then it is resected with rongeur under direct vision, carefully pulling free the final fragment of bone

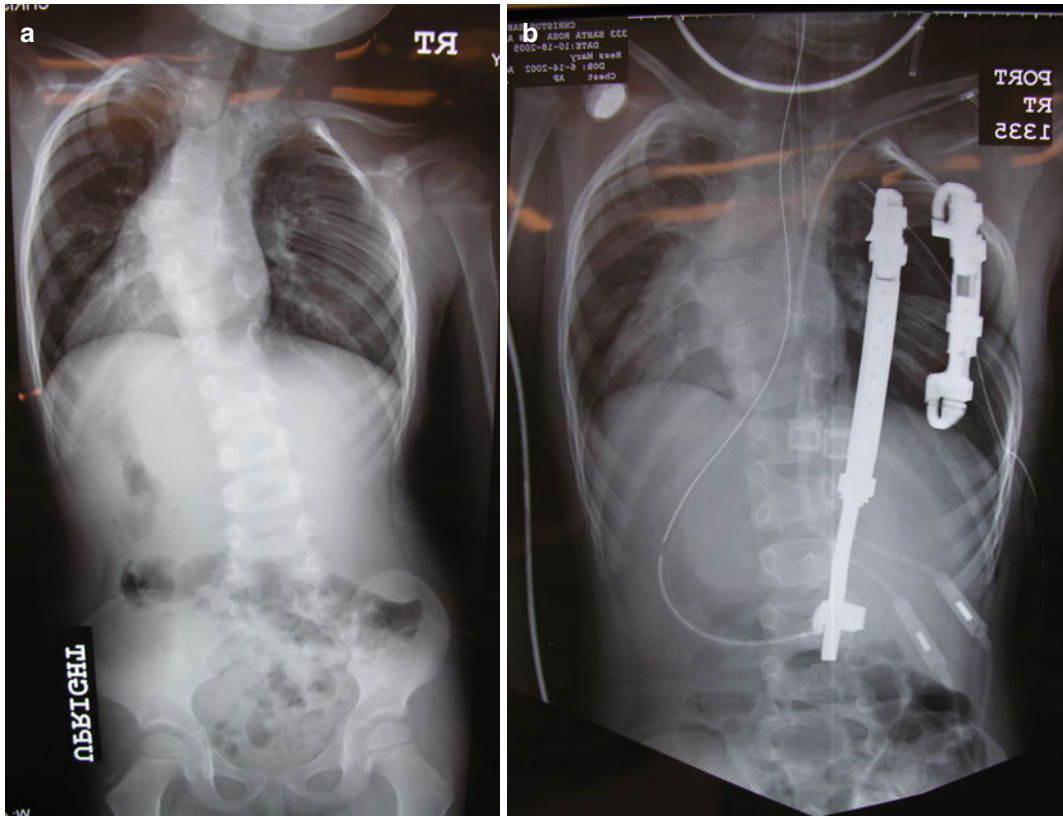


Fig. 39.8 (**a**) Preoperative x-rays of a rib fusion and congenital scoliosis patient. (**b**) Postoperative x-rays of a rib fusion VEPTR procedure

away from the spine with a curved curette to avoid canal violation.

AO bone spreaders are then inserted into the thoracostomy interval and used to widen it; then the rib distracters are next inserted to hold the hemithorax out to corrected length. The pleura is carefully stripped proximally and distally by a Kidner, often with only minimal tearing. When the proximal fused ribs are oriented in the horizontal orientation, then correction is felt to be adequate. Next, a rib-to-rib device is inserted in a younger patient, or, in an older patient, a hybrid device is placed down to lumbar spine [15]. Detailed operative technique is available in prior reports [15, 17]. An additional device may be added in the posterior axillary line, parallel to the more medial device, to share load and provide stability to the area of opening-wedge thoracostomy (Fig. 39.8). If there is flail chest present after the thoracostomy, then it is possible to perform a centralization transport of inferior fused ribs by osteotomizing them from the lower segment of the chest and then displacing them upward into the defect to increase stability of the construct, attaching them by nonabsorbable suture to the device. Closure is in the usual fashion.

Results of 27 children treated with this volume depletion deformity by VEPTR opening-wedge thoracoplasty, with average of 5.7-year follow-up, noted average correction of 25°, with space available for lung increasing from 63 to 80 % at follow-up [4] with evidence of growth in length of the unilateral and unsegmented bars on the concave side of rigid congenital curves [9]. The most common complications noted were asymptomatic migration of the hybrid devices superiorly through the ribs of attachment at an average of 3 years, requiring reinsertion into either a reformed rib or a rib more proximal or distal. Infection rate was 1.9 % per procedure; skin slough was present in 15 % of patients and was treated by debridement and local flap rotation. Two patients, early in the

series, had brachial plexopathy, which is treated by repositioning of devices, and one patient had acute respiratory distress syndrome. One patient died of postoperative pneumonia. Those patients operated on under age 2 years, when lung growth by alveolar cell multiplication is most rapid, had an average vital capacity of 58 % normal at last follow-up, those patients older than age 2 years at time of surgery had average 44 % vital capacity percent normal, while three patients with history of spinal fusion early in life had a vital capacity of 36 % predicted at time of follow-up [4]. VEPTR treatment of congenital scoliosis and fused ribs also improves truncal decompensation, head shift, and neck tilt [19].

39.4.3 Type II Volume Depletion Deformity: Myelomeningocele

Patients with myelomeningocele generally have progressive congenital scoliosis with fused ribs of the concave hemithorax, causing *primary* thoracic insufficiency syndrome, but these children also may have *secondary* thoracic insufficiency syndrome, when there is significant lumbar kyphosis placing the torso too close to the pelvis, blocking effective diaphragmatic excursion. There is the absence of posterior elements for hybrid VEPTR attachment in the myelomeningocele patients, as well as the presence of poor skin centrally from the myelomeningocele closure compromising central exposure. The former can be addressed by attachment of the hybrid VEPTR to the iliac crest, bypassing the deficit in posterior elements, and the latter by slightly skewing the incision laterally to avoid the poor skin. Secondary thoracic insufficiency syndrome in myelomeningocele from flexible lumbar kyphosis or rigid gibbus deformity can be addressed by bilateral VEPTRs extending from the proximal ribs to the iliac crests, an "Eiffel Tower construct." In a study of ten patients with myelomeningocele, treated by VEPTR with an average of 5.75-year follow-up, the average scoliosis was 73° and, at follow-up after VEPTR treatment, was an average of 46°. Six of these patients had flexible lumbar kyphosis, averaging 43°, with a positive marionette sign, indicating secondary thoracic insufficiency, but, with VEPTR treatment, the decrease of lumbar kyphosis was an average of 26° with resolution of their marionette signs [18]. The SAL improved from 66 to 83 % at follow-up, and the thoracic spine height increased 5.8 mm/year (Fig. 39.9).

39.4.4 Type IIIb/II Volume Depletion Deformity: Early-Onset Scoliosis

VEPTR opening-wedge thoracostomy, used to treat extensive thoracic congenital scoliosis and fused ribs of the concave hemithorax, can also address early-onset scoliosis with a similar opening-wedge thoracostomy approach, using intercostal muscle lysis rather than transverse rib osteotomy of the concave hemithorax. The type IIb volume depletion deformity is from the transverse constriction of the chest due to the windswept deformity from spine rotation into the convex hemithorax. The AP type II VDDD hemithorax constriction is identified by the area of multiple persistent intercostal space narrowing of the concave hemithorax in the bending films. The apex of the curve is often distal to this area of rib cage constriction. The superior distraction point, where the superior cradle is attached, should be at the proximal end of the curve. Care must be taken to not place it in the compensatory curve above the structural curve since the distraction force will just increase the compensatory curve without correction of the true curve. The soft tissue approach is identical to that for fused ribs and congenital scoliosis.

The superior cradle is placed in the usual fashion. Once the area of intercostal muscle narrowing is identified, then the central narrow interval is released by cautery, with a right angle clamp under the muscle to protect the underlying pleura. The pleura is mobilized by a Kidner, two to three ribs above and below the interval. Another opening-wedge thoracostomy can be made two ribs above or below the initial release, if the area of constriction is widespread. The ribs are distracted apart by the Synthes rib spreader to lengthen the constricted hemithorax. A unilateral rib to spine VEPTR hybrid is then placed. A second rib-to-rib VEPTR device is often added to load share (Fig. 39.10). Other constructs that can be used are bilateral rib to spine VEPTR hybrids, a unilateral rib to pelvis via Dunn-

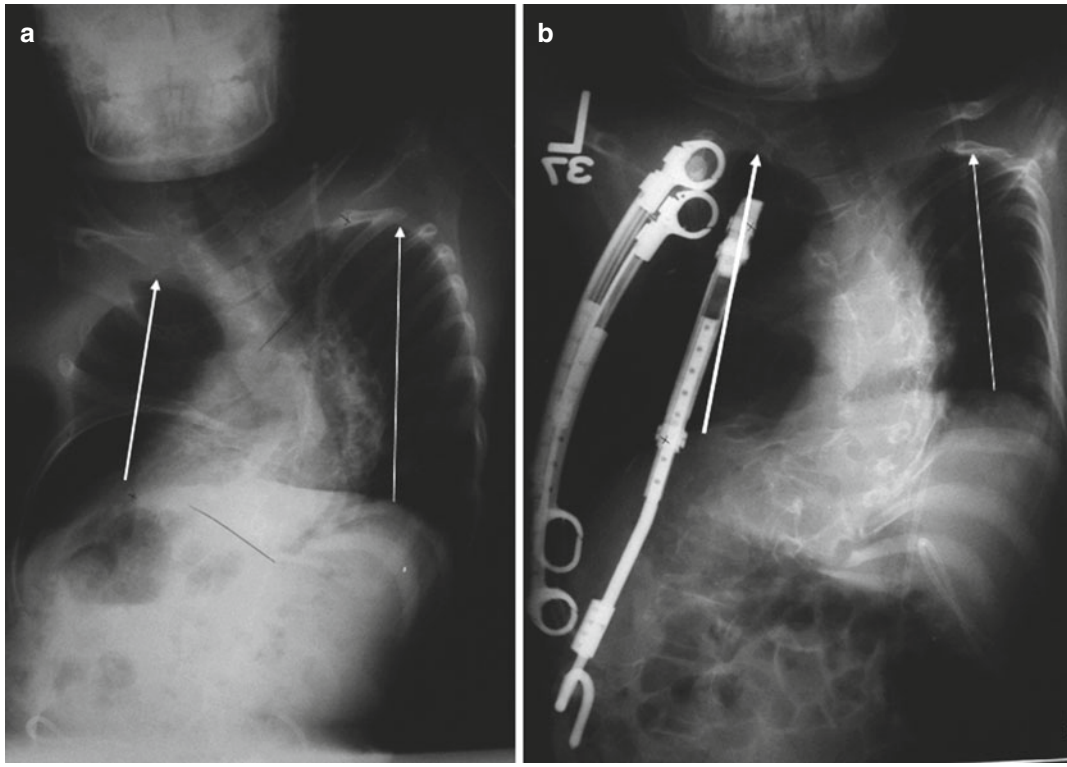


Fig. 39.9 (**a**) Myelomeningocele. The *white arrows* show the "space available for lung", with the concave lung shorter than the convex lung. (**b**) Postop VEPTR treatment of myelomeningocele

McCarthy hook VEPTR hybrids, or bilateral rib to pelvis via Dunn-McCarthy hook VEPTR hybrids.

VEPTR expansion thoracoplasty treatment of early-onset scoliosis remains controversial. Some argue that the presence of a VEPTR on the chest wall will eventually stiffen the chest, adversely affecting respiration, but since the chest wall is already irreversibly stiff preoperatively, it seems unlikely that the VEPTR will affect matters one way or the other. It is assumed that growing rods do not stiffen the chest wall because of their central placement, but often the rods extend over the ribs on the concave side of the curve.

39.4.5 Type IIIa Volume Depletion Deformity: Jarcho-Levin Syndrome

This bilateral hemithorax constriction is treated with bilateral opening-wedge thoracostomies in a staged fashion [20]. For Jarcho-Levin Syndrome due to spondylocostal dysostosis, the technique is very similar to that for the type II volume depletion deformity of fused ribs and congenital scoliosis. The concave hemithorax is addressed first with opening-wedge thoracostomy; then the other side 3–4 months later is expanded if there is significant longitudinal constriction. For patients due to Jarcho-Levin Syndrome due to spondylothoracic dysplasia with minimal, if any, scoliosis and crab-shaped fused chest wall, staged opening-wedge thoracostomies are performed, through a "V"-shaped osteotomy of the densely fused hemithorax with the apex adjacent to the tip of the transverse process in the midportion of the spine (Fig. 39.11a). A VEPTR is placed 2–3 cm lateral to the transverse process of the spine to provide maximum expansion of the hemithorax. The same procedure is performed on the contralateral side approximately 3 months after the first surgery (Fig. 39.11b). A recent report [21] of VEPTR treatment of 10 patients with spondylocostal dysostosis (SCD) and 19 patients with spondylothoracic dys-

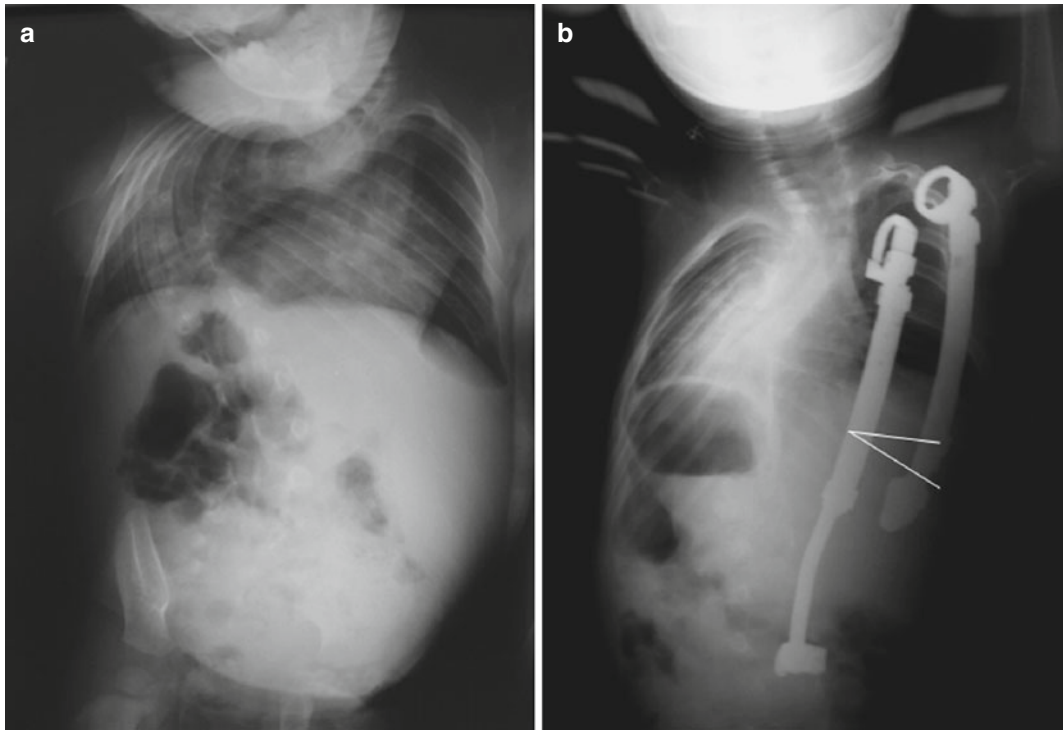


Fig. 39.10 (**a**) Early-onset scoliosis. (**b**) Postop VEPTR treatment of early-onset scoliosis

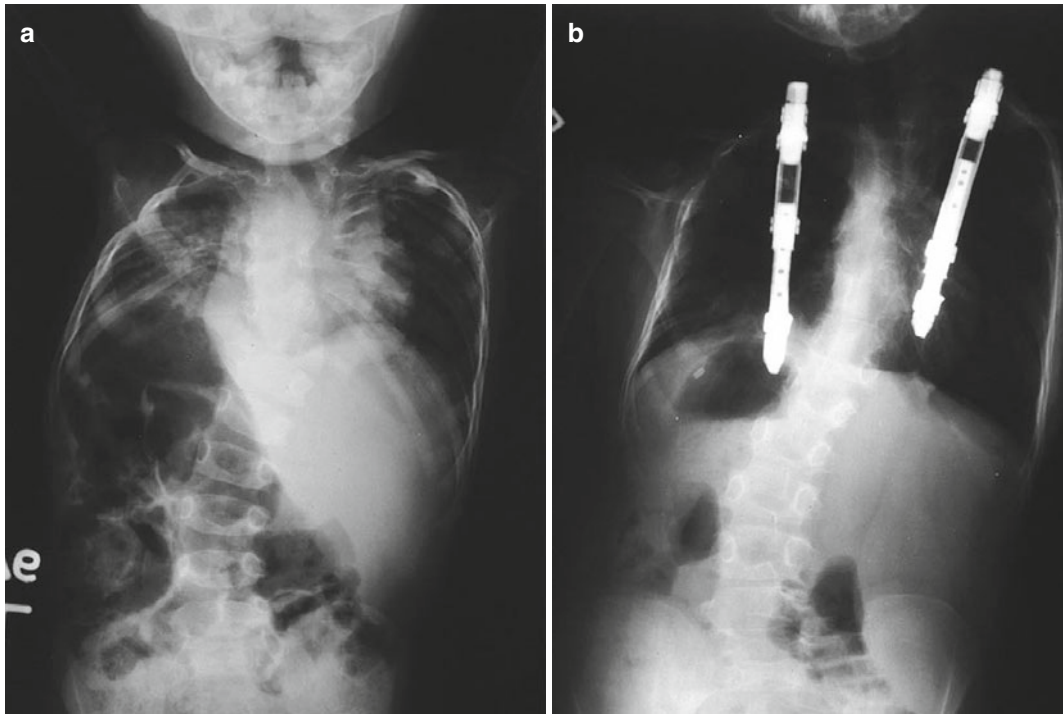


Fig. 39.11 (**a**) Jarcho-Levin Syndrome. (**b**) Treatment of Jarcho-Levin by bilateral VEPTRs after wedge osteotomies of the fused chest wall

plasia (STD) with 6–8 years of average follow-up noted improved thoracic symmetry, control of spinal deformity, and improved clinical respiratory function. Surgery has been controversial for the STD population, but the VEPTR-treated group had 100 % survival, with an increase of 42 % in height of the thoracic spine, and the average FVC percent predicted was higher than that reported in prior literature for natural history, suggesting there may be advantages to surgical treatment of both STD and SCD with VEPTR techniques.

39.4.6 Type IIIb Volume Depletion Deformity: Jeune Syndrome

Juene syndrome is known for the severe lateral constriction of the lungs from a congenitally narrowed chest, causing restrictive lung disease with a high natural history mortality rate of from pulmonary failure. Although in the past this disease seemed mostly just a constrictive chest wall disorder, new data from a soon-to-published large series of 24 patients with Juene syndrome [22] suggests that that spinal disease in these patients is common, with a 41 % incidence of scoliosis preoperatively, with remainder developing scoliosis during treatment, many requiring VEPTR devices to stabilize the scoliosis. In addition, congenital C1 stenosis (Fig. 39.12a, b) was seen in 16 % of patients, one requiring surgery. The chest in Jeune syndrome (Fig. 39.12c) is trilobar "three-leaf clover" shape on cross section (Fig. 39.12d), with the osseous ribs curling inward toward the mediastinum so that the lungs are mostly confined to the two posterior lobes of the chest. This is treated in severe cases of Jeune syndrome by a dynamic posterolateral 70 mm radius VEPTR expansion thoracoplasty [23] in which anterior/posterior osteotomies of ribs 3–8 are performed and then the mobilized chest segment is brought outward and attached by titanium slings to an acutely curved 70 mm radius VEPTR anchored to rib 2 and rib 9 (Fig. 39.12e). The right hemithorax is first expanded and then the left hemithorax 3 months later in a staged fashion. Full operative details are available in the report [23]. At an average follow-up of 8.4 years, survival in this group of treated patients was 68 %, a marked improvement over the 70–80 % death rate of natural history, and there was decreased dependence on respiratory support such as ventilator, CPAP, and nasal oxygen. Infection rate was 4.6 %/procedure. All patients diagnosed with Jeune syndrome should have a screening C-spine radiographs to evaluate for C1 stenosis, with consideration for decompression if significant cord compression exists, and also a CT scan of the chest, 5 mm intervals, unenhanced, to evaluate for lung volumes and to define the severity of the patho-anatomy of the volume depletion deformity of the chest. Severe clinical presentations of Jeune syndrome may benefit from VEPTR expansion thoracoplasty.

39.5 New VEPTR Constructs

39.5.1 VEPTR II Treatment of Kyphosis

The primary improvement of the new VEPTR II over the VEPTR I is the extension of the rib sleeve proximally into a 6 mm rod that can be cut to any length needed with the rib cradle attached to it by a compression fitting. This enables better fit of devices in kyphosis, contouring the rod downward on the kyphotic chest with ability to place the rib cradles at ribs 2–3 to better control kyphosis. Gradual correction of kyphosis can be accomplished by accessing the proximal rods through separate incisions during lengthening procedures to gently straighten the bent rods gradually to decrease the kyphosis (Fig. 39.13). In a study [23] of 26 kyphosis patients treated with this VEPTR II technique, average age 3 ½ years old, with 17.5-month follow-up, kyphosis was 70° preop, 42° immediately after surgery, and 52° at follow-up. Seven patients had a wound infection, but only five had proximal rib cradle migration. This approach may be useful for the control of kyphosis in the very young child.

39.5.2 The VEPTR Gantry

The collapsing parasol deformity (CPD) of Demiglio results in a volume depletion

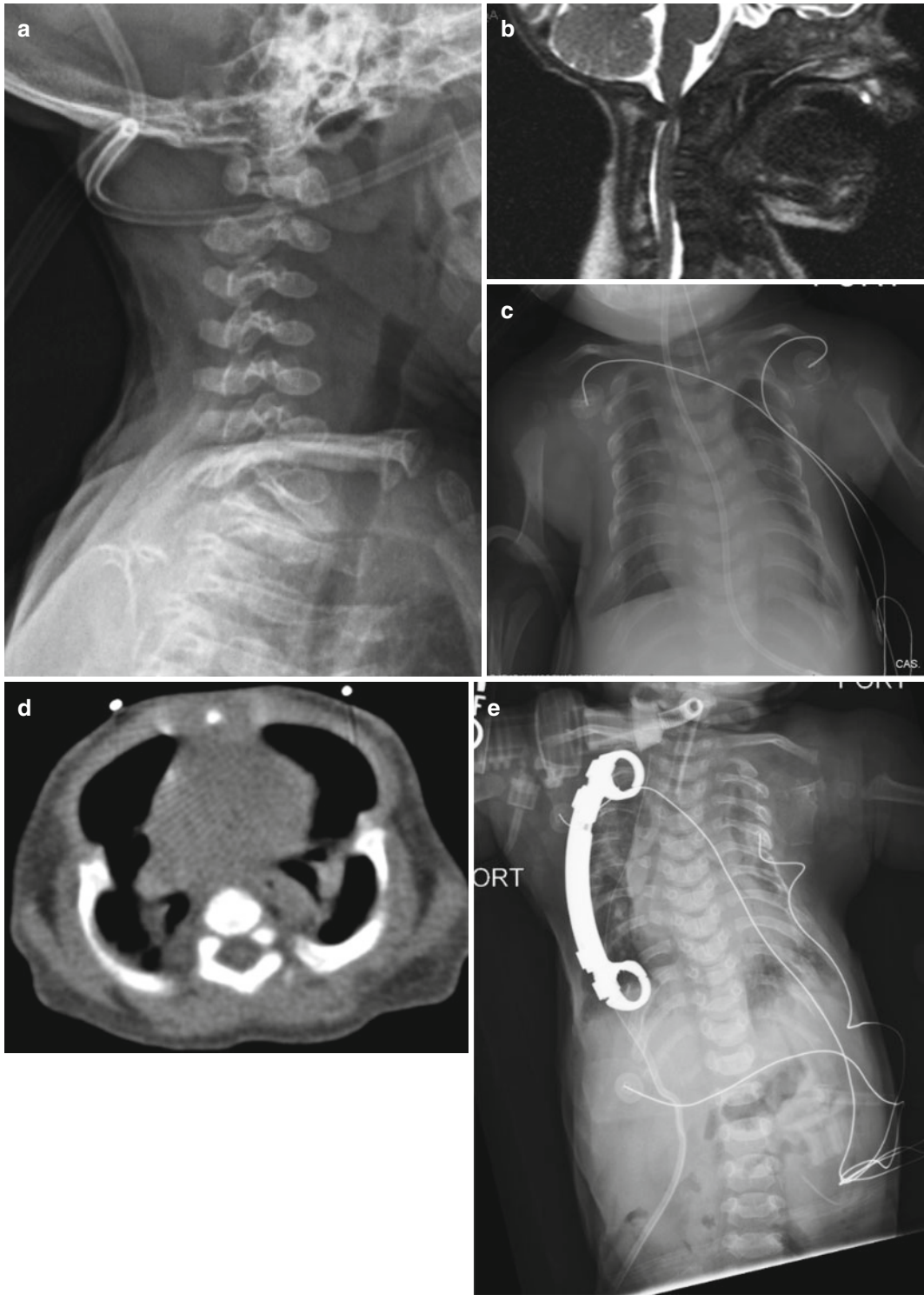


Fig. 39.12 (**a**) C1 stenosis on lateral radiograph of a Jeune syndrome patient with intact neurologic evaluation. (**b**) Significant compression of spinal cord on MRI. The patient underwent surgical decompression. (**c**) AP radiograph showing a narrow "stove pipe" chest of Jeune syndrome. (**d**) CT scan shows a trilobar "three-leaf clover" configuration of the chest in Jeune syndrome. (**e**) Post-VEPTR expansion thoracoplasty of the right hemithorax. The left hemithorax will undergo thoracoplasty approximately 3 months later

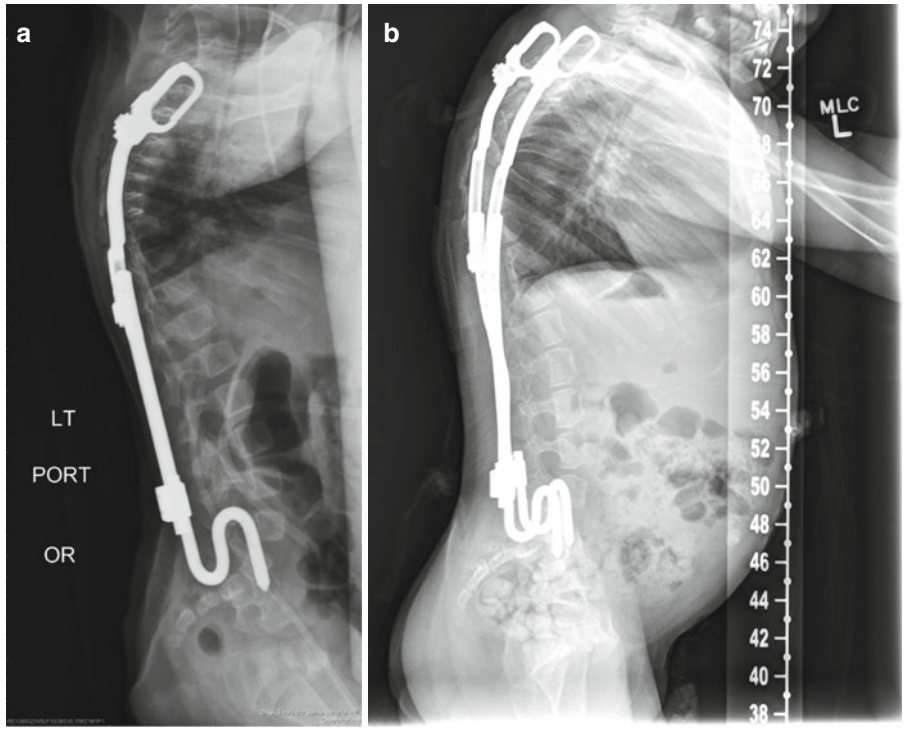


Fig. 39.13 (**a**) An infant with neuromuscular kyphoscoliosis has undergone implantation of a VEPTR II hybrid device on the concave side of the curve with the upper rod of the VEPTR II contoured down to the kyphotic portion of the chest for best fit. (**b**) At 2-year f/u, a second hybrid VEPTR has been implanted on the contralateral side to better control the kyphosis, and both VEPTR II proximal rods have been gently straightened during lengthenings to gradually correct the kyphosis partially

deformity of the thorax on the convex side of scoliosis, but what is even more serious is the thoracic volume loss from a bilateral CPD seen in neuromuscular disease such as spinal muscular atrophy (SMA) (Fig. 39.14a, b). With the VEPTR II, this deformity can be reversed with the use of transverse bars and right-angle rib cradles, a technique we term "the VEPTR gantry" construct. Bilateral VEPTR II hybrids are implanted from proximal ribs to pelvis in the usual fashion, but with at least 6 cm of proximal rod between the rib sleeve and the rid cradles. Two transverse bar rods are prepared for each side, contouring them with a French bender to fit the circumference of the chest, and then loosely attached to the proximal VEPTR II rods. Right-angle rib cradles are then placed in the posterior axillary line around ribs 3 and 5 and then the cradles are attached to the transverse rods. *Do not release the intercostal muscles; they are needed to help elevate the entire chest segment from proximal distraction of rib 3/5.*

C-rings are placed on the proximal VEPTR II rods, and the most proximal transverse bar is first distracted upward to elevate the chest wall segment, and then the second transverse bar is distracted upward to aid elevation of the entire chest wall segment (Fig. 39.14b, c).

Subsequent VEPTR lengthenings provide lateral support to the chest through the transverse bars. This approach has potential to improve chest volume when collapsing parasol deformity is present, both for unilateral or bilateral CPD. The instrumentation is somewhat bulky, so adequate soft tissue must be present over the right-angle rib cradles.

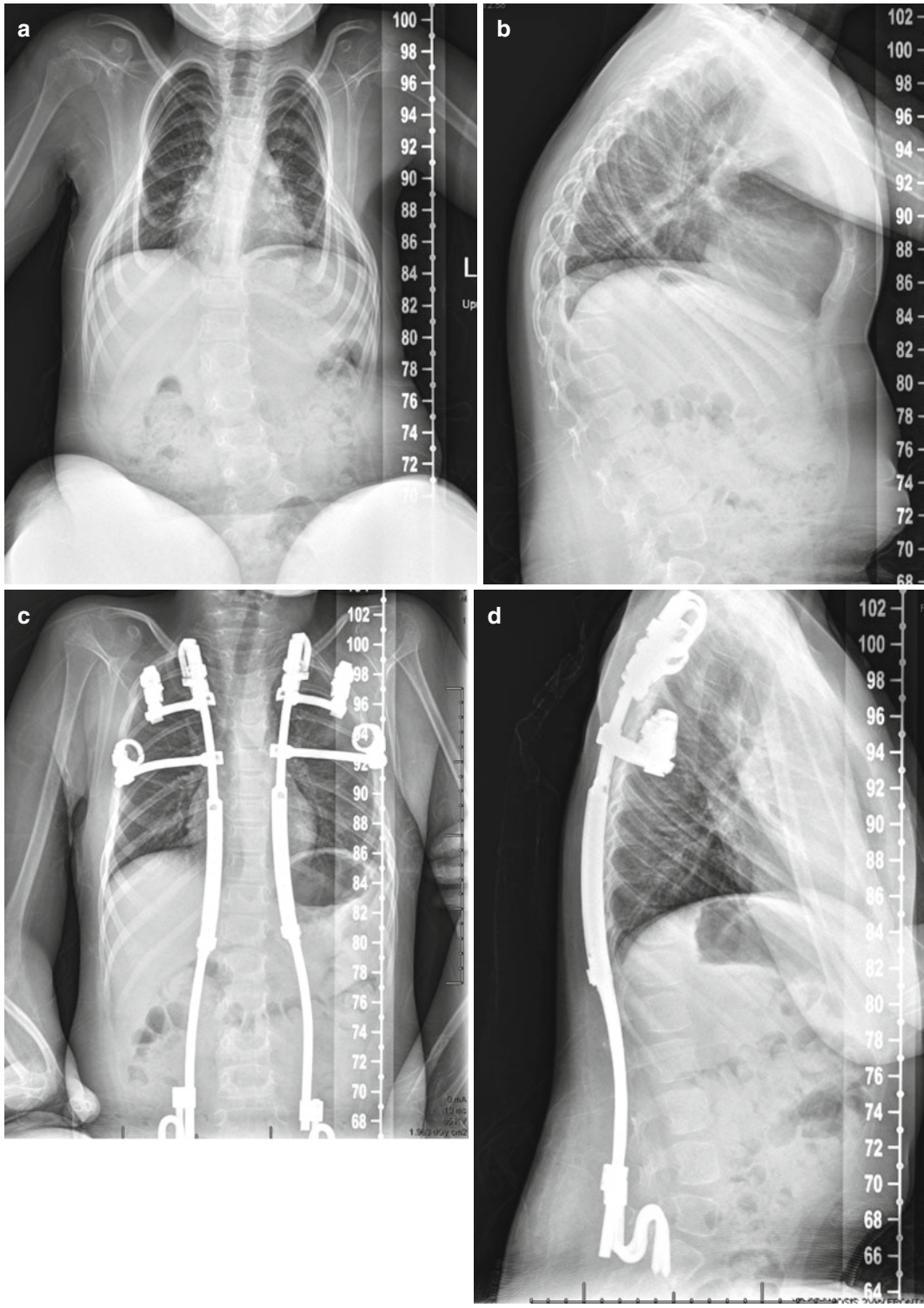


Fig. 39.14 (**a**) A 7-year-old male with SMA type II and collapsing parasol deformity of the thorax. (**b**) Flexible thoracolumbar kyphosis is present. (**c**) Expansion thoracoplasty with bilateral VEPTR gantry constructs, correcting the collapsing parasol deformity of the thorax. Patient clinically improved with decreased respiratory rate and went on to gain weight. (**d**) The procedure has also improved the thoracolumbar kyphosis

39.6 Complications

VEPTR surgeries have the tendency for complications inherent to all repetitive surgery approaches. The most common VEPTR complication is an asymptomatic upward migration of the superior cradle of the device into the rib of attachment over time. It is usually asymptomatic, and once completely migrated, reattachment can be accomplished during a scheduled VEPTR expansion surgery. Through a limited incision of the proximal portion of the thoracotomy incision, the rib cradle is re-implanted into the rib of attachment which is usually reformed, or a more distal rib. Curved curettes are extremely useful for shaving the hypertrophied rib down to acceptable size for reimplantation. Inferior cradle migration is treated in a similar fashion.

Lumbar hooks may migrate distally and can be reseated more distally. In the placement of lumbar hooks at original implantation, it is important not to violate the cortex of the lamina of attachment because this weakens its ability to withstand the distraction forces. If the interspace is too small for the hook, then a superior laminotomy of the interspace is performed. It is important to place the hybrid lumbar extension in the lumbar spine below any areas of junctional kyphosis seen on the lateral weight-bearing x-ray. The VEPTR hook should be placed at least two levels below any junctional kyphosis to prevent accentuating the kyphosis. Downward migration of an S-hook into the iliac crest greater than 4 cm can be addressed by removing the hook and placing it back over the reformed bone at the top of the crest.

Infection in VEPTR patients is often associated with skin slough. Many of these patients have comorbidities, such as myelomeningocele or syndromes, that probably make them prone to surgical infection. The infection rate is 3.3 %/per procedure [2]. For infection, debridement without removal of devices is generally performed and with irrigation by a dilute betadine solution. A wound Vac® is placed and then changed in the OR in 3–5 days. If no infection is seen, then primary wound closure is performed. The patient is maintained on 4–6 weeks of po antibiotics with culture results determining the specific antibiotic. Recurrent infections require removal of the rib sleeve and the lumbar hybrid extension or the inferior rib cradle, and the patient is maintained on 6 weeks of po antibiotics. When sedimentation rate/C-reactive protein has normalized, and the wound is healed, then reinsertion of the device can be considered.

Skin slough is treated by debridement and mobilization of flaps. Primary closure is possible, but loose approximation with proline suture is preferred. In patients with long-standing VEPTR devices, dense soft tissue scarring sometimes occurs over devices and recurrent skin slough becomes a problem. For these patients soft tissue expanders are placed laterally to mobilize skin, the scar is resected, and then the new skin is transferred posteriorly over the devices with the assistance of a plastic surgeon.

VEPTR breakage is rare. Devices with fracture can usually be replaced during a scheduled elective surgery.

VEPTR patients have complex, rare spine, and chest wall deformities, with a significant incidence of spinal cord abnormalities, but neural injuries have been rare. Brachioplexopathy in VEPTR patients can occur by distraction of proximal fused ribs into the plexus, but neural monitoring detects the problem during the procedure and decreasing distraction usually resolves the problem. To prevent spinal cord injury during VEPTR implantation, the preoperative CT scan should be checked carefully for areas of dysraphic spine within the area of approach. In these areas, dissection should be cautious, with medial exposure only above and below the areas of dysraphism. Particularly dangerous is an area of mid-thoracic spine dysraphism where the medial edge of the scapula lies within the spinal canal on the concave side of the curve. The usual thoracotomy approach in this anatomic variant, cutting through the rhomboid muscles with cautery, would result in direct spinal cord injury. To avoid this, the scapula should be retracted posteriorly with a rake to pull it out of the spinal canal, and the rhomboid muscles sectioned directly off the bone of the scapula, away from the area of dysraphism. There is also risk of lumbar spine

dysraphism of the posterior spinal elements in patients with extensive congenital scoliosis. Use a large Cobb elevator in stripping the paraspinal muscles to minimize the risk of violating the canal during exposure. Any monitoring changes during surgery should be addressed by decreasing distraction.

In summary, most VEPTR complications are treatable, and the frequency can be tolerated in view of the probable long-term pulmonary benefit.

39.7 The Future

The recent FDA clearance of the MAGEC® magnetic growing rod heralds a new era for growth-sparing instrumentation that could result in reduction of morbidity and possibly cost, but although early experience is favorable, long-term follow-up is needed to define the unique complications that may arise such as device jamming or breakage that may require additional surgeries or change in strategies. Another concern is the loss of MRI assessment for these patients because of the presence of the magnet in the device. Similar technology will be available for VEPTRs in the near future. The magnetic drive devices are compact in cross section but require considerable linear length in order to provide practical lengths of distraction, so while tolerated in older children, younger children may continue to need traditional VEPTR treatment until they are large enough for the magnetic option. While the ability to distract devices in an outpatient setting without surgery is a true advance, more advanced device intervention will require a deeper functional anatomic understanding of these deformities and their effect on spine, chest wall, and lung growth. Animal models may address the lung growth issues and dynamic lung MRI is helping reveal the patho-anatomic basis of thoracic dysfunction that causes thoracic insufficiency syndrome. It is important to understand the disease completely in order to best address its treatment. VEPTR as a device has made a positive impact at long-term follow-up on many complex spine and chest wall diseases, but with a deeper understanding of thoracic insufficiency syndrome and by using VEPTR concepts, even more effective surgical strategies and devices can be designed for the future.

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Convex Growth Arrest for Congenital Scoliosis

40

Muharrem Yazici and Ozgur Dede

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Key Points

- Controlling the growth of the convex side may stop the progression and also lead to further spontaneous correction, provided that the concave side has remaining growth potential.
- Convex growth arrest is a safe procedure; however, the unpredictability of the curve correction has prevented the common use of the technique.
- Various applications of convex growth arrest have been used in very young patients with congenital spinal deformity with some success.
- Recent techniques with convex hemiepiphysiodesis and concave distraction with instrumentation may result in improved deformity correction and continuing spinal growth.

40.1 Background

Growth arrest, as a technique to correct deformities and control growth, relies on the presence of localized growth center in the developing human skeleton. The physis (i.e., growth plate) is the cartilage zone between the metaphysis and epiphysis of the long bones that are responsible for the major part of longitudinal growth. Secondary to the

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imbalances of physeal growth, a variety of extremity deformities arise, such as limb length discrepancies and angular deformities. Modulation of the growth on one or both sides of a growth plate is a potent treatment method of such deformities in the growing skeleton. Thus, limb length discrepancy and deformity correction that interferes with the development at the growth plates (i.e., growth arrest) has long been tried and successfully practiced since the early 1930s [14, 24, 27, 35].

Congenital spinal deformities result from anomalous vertebrae that produce aberrations in the coronal and sagittal spinal alignment and may progress due to the longitudinal growth imbalances. In progressive curves, typically there are hemivertebrae or unilateral bars that cause an unbalanced growth of the vertebral column, causing convexity and concavity occurring simultaneously on opposite sides. Modulation of vertebral growth on either the convex or concave side of the curve (growth arrest or growth enhancement, respectively) theoretically seems to be an early and effective treatment alternative for the growing spine. Early attempts at arresting the growth of the convex side have been reported, notably that of Smith et al. [31] by a stapling method. In 1963 Roaf reported his surgical technique and corresponding patient outcome in utilizing unilateral growth arrest for congenital scoliosis [29]. His technique consisted of a posterior approach with removal of at least four ribs, transverse processes, and costovertebral articulations on the convex side of the curve, as well as removal of the part of the annulus and curettage of the disk and the epiphyseal cartilages. Convex anterior and posterior growth arrest has been used in patients with congenital spinal deformity, enabling the progression to cease or even reverse the deformity with subsequent growth of the spinal column [2, 4, 11, 16, 17, 32, 34, 38, 39]. Theoretically, controlling the growth of the convex side, which is relatively longer along the length of the vertebral column than the concave side, not only would stop the progression but also lead to spontaneous correction provided that the concave side has remaining growth potential. The convex growth arrest (CGA) procedure based on this concept has been popularized because of its safety and simplicity compared with other surgical alternatives [2, 4, 11, 16, 17, 21, 32, 34, 38, 39, 41].

40.2 Classical Indications and Contraindications

CGA is not suitable for every congenital spinal deformity, and several criteria have been defined in the literature. These commonly accepted criteria dictate a purely scoliotic curve comprised of less than five segments with a magnitude of less than 70° in a patient not over 5 years of age. Sagittal plane deformity, cervical involvement, intraspinal anomalies, posterior arch defects (e.g., myelomeningocele), and unilateral bars are considered to be contraindications for this procedure. Deformities caused by a single hemivertebra may be better treated with a hemivertebrectomy procedure.

40.3 Techniques

The original technique described by Winter [39] and Andrew and Piggott [2] consists of anterior and posterior interventions to the spinal column. Separate anterior and posterior exposures are performed; these surgeries can be done either during the same session or with a week in between. Initially, the cartilaginous end plates and the intervening disks are partially excised, and the gap is filled with bone grafts using an anterior approach to the spinal column. Then using a posterior approach, the zygapophyseal facet joints on the convexity of the curve are removed and filled with bone grafts in order to produce a fusion effect. A protective cast is applied 4–7 days after the operation. This provides some correction while the fusion occurs. It is then removed at 4–6 months when the fusion is evident on radiographs.

40.4 Results

One of the earliest studies by Smith et al., using anterior staples over the convexity [31], did not report any improvement, which was argued to be

due to the anatomy and the bony structure of the vertebrae. It was further argued that the growth rate of the vertebrae is much less than those of the long bones, as well as the soft cancellous bone not causing enough compression on the end plates. Roaf [29] reported his results of using anterior epiphysiodesis and posterior intra-articular fusion on 188 patients. There were no cases of complete correction, only those with limited improvement. These results were argued by Andrew and Piggott [2] to be partly due to the extrapleural approach that only provided a limited exposure and resection of the posterior ends of several ribs on the convex side.

Marks et al. [21] reported results for the use of anterior and posterior convex hemiepiphysiodesis for congenital scoliosis with a mean follow-up period of 8.8 years on 57 patients. Furthermore, the rate of change of the Cobb angle was decreased but not reversed when deformity was due to an unsegmented bar. For complex anomalies, they reported an increase in the final Cobb angle from a mean of 61–70°. The rate of progression reversed or decreased in 97 % of patients with hemivertebra, and the mean Cobb angle improved from 41° preoperatively to 35° postoperatively. Lumbar anomalies and the younger patient age resulted in better corrections. Additionally, Uzumcugil et al. [33] reported their results on 32 patients utilizing anterior and posterior approaches. Forty-one percent of the patients had true epiphysiodesis effect, 47 % of the patients had fusion, and only 12 % of the patients showed an increase in the curvature at a mean follow-up of 40 months.

The classically defined indications of the procedure have been commonly expanded by a number of authors. There are conflicting reports in the literature when the variables that are commonly accepted to affect the outcome of the convex hemiepiphysiodesis surgery are considered. Uzumcugil et al. [33] scrutinized these criteria in a series of 32 patients and also provided an extensive review of the series in the literature. Their literature review and clinical results imply that convex hemiepiphyseodesis can be performed for the balanced and cosmetically acceptable deformities of patients younger than 5 years of age regardless of the type, length, magnitude, and location of the curve. The existence of associated rib fusion or the presence of sagittal plane abnormality does not seem to negatively affect the results. Anomalies consisting of hemivertebrae instead of unsegmented bars have repeatedly been reported to yield a more favorable outcome [2, 11, 16, 18, 32, 39]. This may be originating from the belief that in the presence of an unsegmented bar, it is impossible for the concavity to grow. On the other hand it has also been shown that fusion of the one upper and one lower segment of the bar may result in an improvement of the deformity. From natural history studies, it is known that more severe and progressive deformities occur either in the thoracolumbar region [23, 30] or the thoracic spine [40]. However, Thompson et al. [32] reported that hemivertebra in the lumbar spine had the best prognosis when treated with CGA. Walhout et al. [34] reported that complex deformities in the thoracolumbar and upper thoracic regions were more favorable than those in the lower thoracic region when CGA was the treatment modality. These mixed results show that there is no preference between the upper and lower spine for CGA. For a successful CGA, the preoperative curve magnitude was reported to be less than 50–60° [23], whereas there are other studies showing that curves less than 70° also have favorable results [16, 17, 39]. The number of vertebrae included in the curve was also reported to have an effect on the outcome of the surgery. The best results were reported with curves that affect five consecutive segments or less [16, 17, 39]. On the other hand, longer curves have been successfully treated with CGA [33]. Majority of available literature indicate that the presence of an intraspinal anomaly, either treated or not, does not seem to have a negative effect on the progression of congenital curves [15, 22, 25, 36, 37]. However, Reigel et al. [28] reported that the release of spinal cord tethering results in a stabilized or improved scoliosis in lumbar curves, whereas it does not stop the progression of scoliosis in the thoracic level. The upper age limit set for an effective CGA was reported to be 5 years [2] since most of the vertebral growth occurs before this age. However, it

should be noted that another work has suggested that the procedure is effective in children older than 5 years without signs of advanced skeletal maturity [16, 33]. Although the existence of a sagittal plane abnormality (i.e., kyphosis or lordosis) is accepted as a contraindication for CGA, this issue has not been discussed or evaluated in detail [2, 16, 32, 34, 39]. Comprehensive analysis of the literature reveals two possible effects of a sagittal plane abnormality on the outcome of CGA: progression of sagittal plane deformity despite well-stabilized scoliosis or unsatisfactory control of the deformity in all three planes [2, 16, 34, 37]. Dubousset et al. [11] and Kieffer and Dubousset [17] reported that CGA could be used even in patients with kyphoscoliosis or lordoscoliosis. Their findings contradict with the assumption that the presence of a sagittal plane deformity negatively affects the outcome of the CGA procedures (Fig. 40.1a–d).

As can be learned from the above discussion, none of the variables including the age, type of the anomaly, presence of sagittal deformity or intraspinal anomaly, and length of the curve were found to significantly affect the outcome of the procedure.

40.5 Problems

CGA is accepted as a safe procedure that generally does not result in serious complications except for minor infections (wound or chest) and traction neuropraxias of either the intercostal or cutaneous thigh nerves which are related to anterior surgery [2, 4, 11, 16, 17, 29, 32, 34, 38, 39]. More significant problems include unpredictability of the curve behavior after the procedure and incapability to control the spinal balance.

40.6 Proposed Solutions and Modifications

For each of the drawbacks mentioned previously, potential solutions were proposed by a number of authors. Bandi et al. [3] reported a modified technique on two patients, which spares the segmental vessels during the anterior epiphysiodesis surgery in order to decrease the neurological complications. It has been argued that as the number of segmental vessel ligation increases, there is a corresponding increase in the risk of spinal cord ischemia and there are reports of such spinal cord injuries. In the proposed technique, segmental vessels are mobilized and elevated during the anterior epiphysiodesis procedure. This method provides a method of sparing segmental arteries however it does not address the other potential problems with anterior surgery and thoracotomy.

Keller et al. [16] and King et al. [18] reported an alternative method which avoids the anterior surgery and thus the risks related to it. This method consists of a posterior approach utilizing transpedicular curettage of the end plates anteriorly, from a posterior approach. Posterior hemiepiphysiodesis is done as in the original technique. Transpedicular approach has potential advantages over the standard two-staged operation in that it decreases the neurovascular complications by avoiding the anterior approach. On the other hand, a potential disadvantage is the chance for an incomplete hemiepiphysiodesis of the anterior end plates.

More recently transpedicular approach with short segment posterior instrumentation was reported by Ginsburg et al. [13]. Their series consisted of ten patients with a mean follow-up time of 29.7 months. They reported either no improvement or a decrease in the curves in seven of their patients. Conclusions were drawn that CGA with a transpedicular approach is an effective method for congenital scoliosis especially when done earlier in premenarchal patients and patients with open triradiate cartilages.

Cheung et al. [6] added a growing rod to the concave side and reported their results using posterior convex hemiepiphysiodesis with concave distraction. Distractions were not done in a regular basis but were done only when loss of distraction force was evident. The authors stated that the loss of distraction was manifested by the increasing space between the hook and the C-ring of the Harrington rod or between the hook and the lamina or when the hook became dislocated or the curve deteriorated. At a mean follow-up of

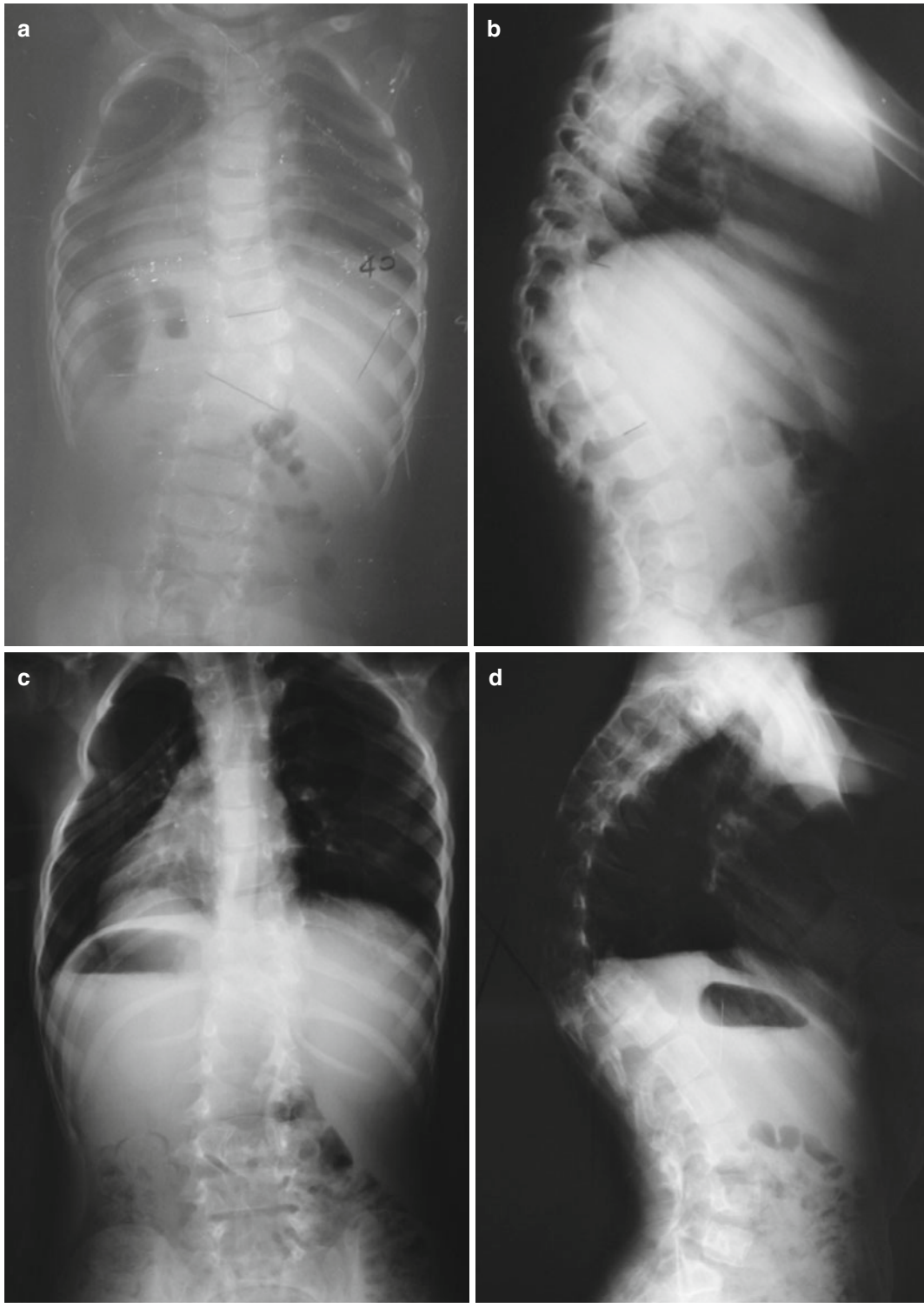

Fig. 40.1 (**a**) Thoracolumbar scoliosis and (**b**) concurrent sagittal plane deformity in a 4-year-old male patient who was treated with uninstrumented anterior convex growth arrest. Four-year follow-up radiographs show almost complete correction of (**c**) coronal spine deformity with (**d**) slight improvement of the sagittal deformity

10 years, the authors reported 41 % correction of scoliosis. The concave distraction produced immediate improvement in the coronal balance, and further correction with consecutive distractions was minimal. They suggested that this procedure could be recommended for children with severe deformities and decompensation in the lower thoracic spine.

Another modification that has been proposed by the Hacettepe group is the addition of posterior instrumentation with transpedicular screws to the convex hemiepiphysiodesis (Fig. 40.2a–f) [8]. Specifically, the technique exploits the concept that pedicle screws may control the growth of the vertebral column in both longitudinal [19] and transverse planes [7] as demonstrated in animal studies. Transpedicular screws were placed in all anomalous vertebrae to be hemiepiphysiodesed to eliminate the need for an anterior surgery. Compression-rotation maneuvers were employed to correct the deformity. Added posterior instrumentation provided an initial correction, thereby decreasing the unpredictability of the outcome; however, trunk balance was not achieved in every case.

The techniques that employ convex hemiepiphysiodesis depend on the growth potential of the concave side of the anomalous segment for further correction and growth. However, this potential may be very small in congenitally abnormal vertebrae. In order to achieve a better trunk balance and progressive correction, we added a concave instrumented distraction construct to convex instrumented compression and fusion [1], similar to that suggested by Cheung et al. [6]. The initial results of this latest modification showed that the technique may provide immediate deformity correction, trunk balance, and continued growth of the spine (Fig. 40.3a–f). Currently, longer follow-up of the technique with a larger cohort of patients has become available [9]. The initial coronal deformity was 60.5° and corrected to 40.7° postoperatively. Distraction of

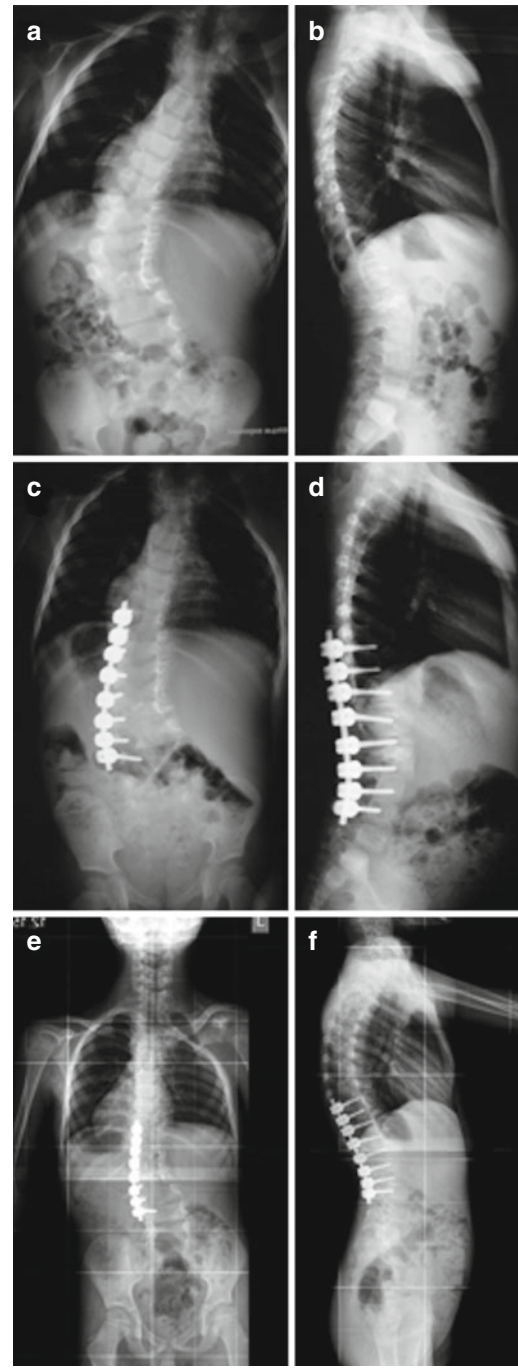

Fig. 40.2 A six-year-old male with (**a**) thoracolumbar coronal deformity and (**b**) normal sagittal alignment was treated with instrumented convex growth arrest. Postoperatively the patient had improved coronal alignment (**c**), and the sagittal alignment is positive on lateral radiograph (**d**). At 5-year follow-up, the coronal deformity further improved (**e**), and the sagittal contour appears to have reconstituted to normal (**f**)

the concave growing rod was performed every 6 months, and at a mean follow-up of 31 months, the final mean curve magnitude showed further correction at 35.5°. The sagittal plane was minimally affected from the procedure. Longitudinal growth of the T1–T12 segment has been noted in all patients in parallel with the improvement in deformity of mean 6.4 mm/year. Although our patients had multiple vertebral anomalies, the T1–T12 growth was only slightly less than the normative data published by Dimeglio for this age group [10]. This maybe due to the previously suggested growth stimulation by recurrent distractions of the growing construct [26].

The instrumented CGA with concave distraction appears to be more effective than the previously reported techniques of CGA, as there was no progression in the curve size but there was correction in all patients. The rationale for this technique is that the pedicle screws control the growth of the anomalous vertebral segments in the longitudinal [19] and transverse planes [7], and thereby obviating the need for an anterior fusion, while permitting spinal growth on the concave side of the curve through growth stimulation as a result of distraction [5]. Control of the whole curve by the concave growing rod also helped obtain immediate correction of coronal plane balance problems when compared with uninstrumented CGA. The potential advantages of this modification are that instrumentation on the convex side provides a complete hemiepiphysiodesis at the anterior and posterior convex sides, obviating the need for anterior surgery and enabling compression-rotation maneuvers for initial acute correction. Also, the instrumentation

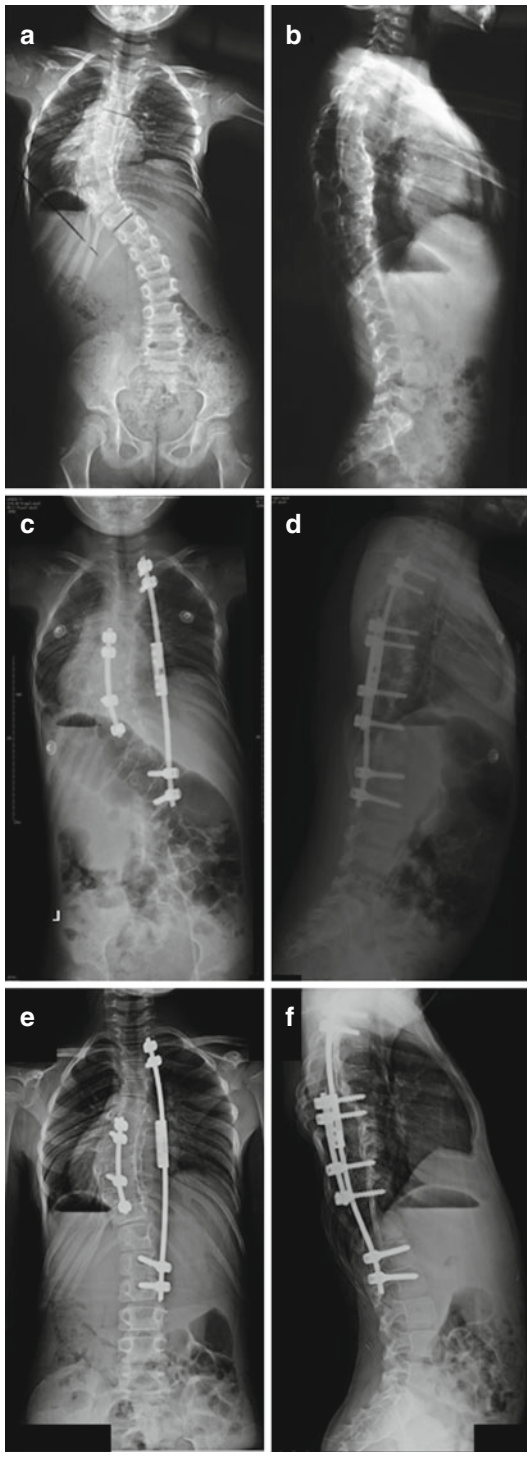

Fig. 40.3 A four-year-old female with (**a**) 60° thoracic scoliosis and coronal off balance and (**b**) hypokyphotic sagittal alignment underwent an instrumented convex growth arrest with concave distraction. Postoperative radiographs show that (**c**) coronal balance was achieved with substantial curve correction (40°), and (**d**) there appears to be a positive sagittal balance. At 2-year follow-up, the patient showed (**e**) 2 cm growth of the T1-S1 segment of the spine and further correction of the coronal deformity to 30°. The sagittal balance and alignment seems minimally changed from the preoperative radiograph (**f**)

makes the procedure reliable by preventing potential inconsistencies that might arise from surgeon-dependent factors such as amount of end plate preparation.

One of the main disadvantages of the technique is that recurrent trips to the operating room are required for distraction (lengthening). This disadvantage may be circumvented by the utilization of the new magnetically controlled growing rod (MCGR). Our first trials with this technique show promise; however, the follow-up period is too short to make any solid conclusions (Fig. 40.4a–e). Another disadvantage arises from the use of instrumented hemiepiphysiodesis (hemifusion) of the apical vertebrae. Growing rod treatment is an alternative method for treatment of young children with a long curve and with a relatively flexible apical deformity including congenitally deformed vertebrae [12, 42]. However, growing rods do not control the apex of congenital curves with stiff anomalous segments involving more than four vertebrae, as were the typical case samples in the current study [42]. Another option for rigid, long sweeping congenital curves may be vertebral column resection [20] with limited posterior fusion. However, for such curves involving multiple anomalous segments, this technique causes shortening of the thoracic spine and necessitates the fusion of at least four to six additional thoracic levels for fixation after resection of the anomalous segments, thus interfering with thoracic growth. Moreover, this procedure is technically difficult and carries more neurologic risk compared with less complex procedures. Therefore, a less invasive method that preserves growth is warranted, whenever possible.

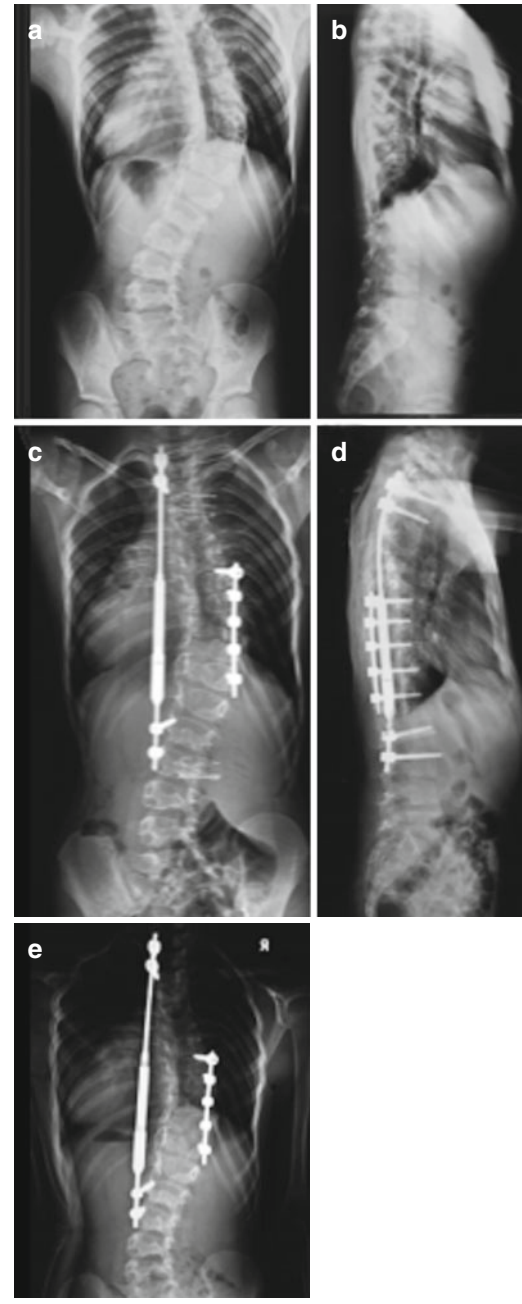

Fig. 40.4 Anteroposterior (AP) (**a**) and lateral (**b**) radiographs of a 6-year-old female with previous thoracic diastematomyelia spur excision. Her progressive long sweeping congenital curve was addressed with instrumented convex hemiepiphysiodesis and concave distraction using a magnetically controlled growing rod. Postoperative radiographs show satisfactory alignment in anteroposterior (**c**) and lateral (**d**) views. At 6-month follow-up, the AP radiograph showed further correction (**e**) with maintenance of sagittal alignment (not shown). The patient underwent three distractions during the 6-month follow-up at 2-month interval clinic visits using the magnetic rod remote controller

Conclusion

The main problems of CGA seem to be the unpredictability of the results and the variable ability to control the curve. In order to alleviate these issues, there have been several proposed solutions, and instrumented CGA with concave distraction appears to be an effective procedure to halt the progression of the congenital curves with an expected correction over time in patients with a substantial amount of remaining spinal growth.

In younger children with mild congenital curves, instrumented CGA with concave distraction is an alternative treatment modality and does not preclude future interventions such as a definitive instrumented fusion. CGA with concave distraction improves the coronal deformity with no negative effects on the sagittal alignment and allows for further deformity correction and spinal growth. The procedure is a less invasive alternative for complex congenital curves, which otherwise may require multiple osteotomies and longer thoracic fusions.

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Growth-Guided Instrumentation: Shilla Procedure

41

Richard E. McCarthy and Scott J. Luhmann

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Key Points

- The Shilla growing rod system is a growth-enabling system designed to direct spinal growth.
- The Shilla device harnesses the growth potential of the spine after correction and fusion of the curve apex.
- Growth continues at the ends of the curve by virtue of Shilla growing screws.

41.1 Introduction

Traditional "growing rod" systems are designed to produce a distractive force between the ends of a scoliotic curve and rejuvenate that distractive force on a regular basis. The latter usually requires a return to surgery every 6 months. The goal is to continue this distractive force throughout the period of juvenile and early adolescent growth until a time when sufficient vertebral column growth has been achieved. At that point, the implants are removed and replaced with a permanent rod system and posterior spinal fusion. The patient is sent into adulthood with this system in place.

With regard to early-onset scoliosis [1–3], the Shilla system begins with the ultimate "ideal goal" in mind, namely, sending patients into adulthood with a vertebral column as tall as possible in as neutral an alignment as possible, and

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as mobile as possible, free of implants. The Shilla system is a new and different way of thinking about the treatment of early-onset spinal deformities, which strives to achieve this ideal goal as closely as possible.

The Shilla growing rod system enables growth while directing it into a more normal alignment, harnessing the growth potential of the patient's vertebral bodies. The guiding principles include:

1. The maximum number of growth centers (34 exist between C7 and S1) should be preserved through the treatment process.
2. The more unfused segments maintained, the more normal the ultimate mobility.
3. The deformity is worst at the apex. This is where the maximum corrective forces need to be concentrated and maintained. Therefore, if a growth center has to be sacrificed, it should be at the apex of the deformity.
4. Bracing or casting compresses the chest wall, compromising pulmonary function while restricting a child's interaction with his/her environment and with others. It also stigmatizes the child as different.
5. Pedicle screws supply the best, most stable vertebral fixation and can be placed from an extraperiosteal position to maintain growth.

41.2 Historical Perspective

The growth guidance concept grew out of the sliding technique of the Luque Trolley. What was characterized as the "trolley" was an unfused spine instrumented with multiple levels of sublaminar smooth wires placed bilaterally and linked to smooth parallel rods placed along the lamina. Some patients with this device continued to grow in spite of the periosteal stripping of the spine used to place the implants, but the results were inconsistent, and the technique has fallen into disfavor [4–6].

The concept was good, namely, to guide the spine to grow straight. The problem was the anchors causing interlaminar ankylosis and eventually autofusion. This bone formation is avoided by the use of an extraperiosteally placed pedicle screw that allows continued spinal growth while fixing the vertebral body to a rod and guiding it to grow in a straight manner with normal sagittal curves.

41.3 Experimental Background

Early in its inception, Shilla was implanted into immature goats to address some basic questions. The implants had been tested using an Instron testing machine, and the device itself was shown to be sound. After a million compression cycles, the only instrument defect was metal filings from movement of the rods in the Shilla screws without instrument failures. The in vivo model consisted of 11 immature goats instrumented at approximately 2 months of age with explantation of their spines at 6 months postop. The questions addressed in this model were the following: (1) could the implant be safely inserted into small pedicles in a manner that would allow for growth? (2) would bilateral pedicle screws in the thoracic spine produce spinal stenosis?, and (3) what would be the effects upon the instrumented but unfused facet joints?

The examination at 6 months included gross and radiographic examination of the specimens, manual testing, and microCT studies. The results indicated that the spines grew and the implants slid along the rods as expected. No evidence of spinal stenosis occurred at the apex where the bilateral pedicle screws had been placed; although the facets at the level of the Shilla screws were degenerated, the adjacent levels survived and had preservation of the facet articular cartilage. One goat was paralyzed by the surgery and was sacrificed once this was evident soon after surgery. Autopsy showed one of the thoracic screws significantly encroaching the intrathecal space [7].

41.4 Method

41.4.1 The Implant Design

The Shilla "growing" screw is a polyaxial screw with a locking plug that fixes to the top of the screw and not the rod. It captures the rod allowing

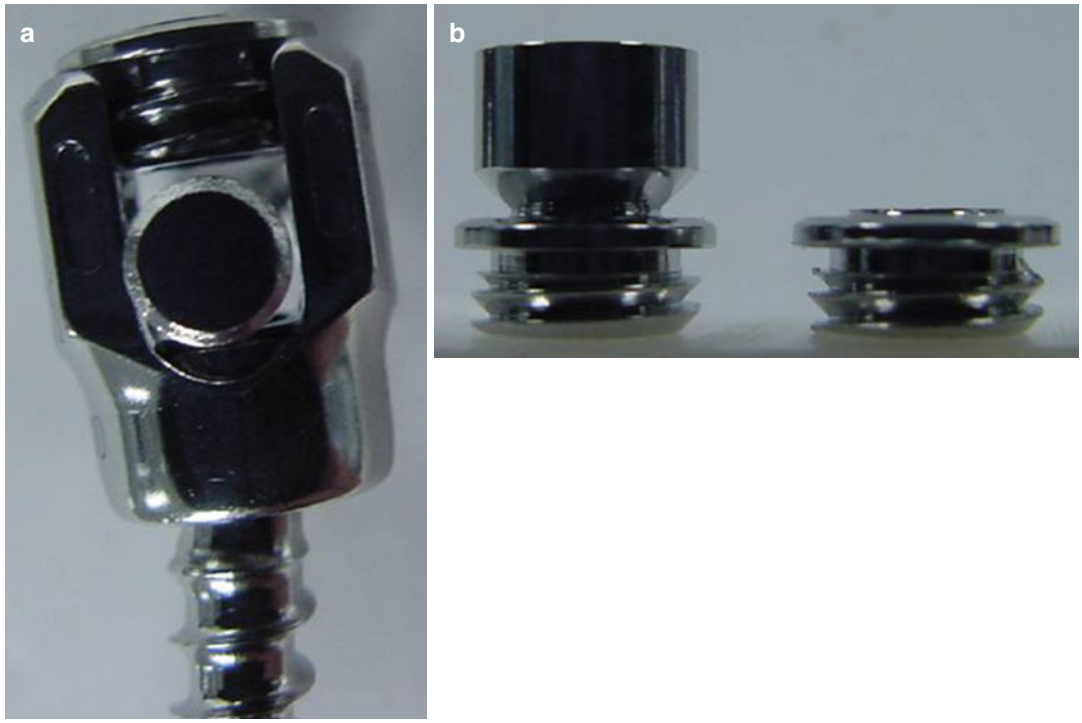

Fig. 41.1 (**a**, **b**) The Shilla growing screw: a polyaxial pedicle screw with a cap that fixes to the top of the screw and snaps off for a low profile (**b**) and allows for the rod to be captured and able to slide in the screw

it to slide in a longitudinal direction. The polyaxial head allows the rod a few degrees of side-to-side movement. This movement at the base of the screw head diminishes the stress on the bone/thread interface during normal movements (Fig. 41.1a, b). The screws placed bilaterally at the apex of the curve are fixed head type to provide maximal correction in all places. The tops of the locking plugs snap off to minimize contour of the implants (b).

41.4.2 Surgical Technique

41.4.2.1 Preoperative Planning

Preoperative patient planning consists of a careful assessment of the upright coronal and sagittal films coupled with analysis of the flexibility of the curve via supine bend films, fulcrum bend films, or traction films. Determination of the location of the apical vertebral segments is of key importance. Those apical three or four vertebral segments that are least corrected through flexibility testing are the ones that comprise the apical levels for fusion and maximum correction. The goal is to render these segments neutral in all planes. If the surgeon can achieve this through posterior techniques alone (Ponte osteotomies, pedicle subtraction, or vertebral column resection), then no anterior release is necessary. For very stiff curves, anterior disk and end plate excision of the interval levels may be necessary before the planned correction. The posterior placed fixation at the apex uses bilateral fixed head pedicle screws. The Shilla growing screws are placed above (cephalad) and below (caudad) the apex to guide the growth of the spine at the ends of the curve and maintain coronal and sagittal correction. These screws are placed through the muscle layer without taking down soft tissues except to cut the fascial planes on each side of the midline. The use of a blueprint based on preoperative planning is helpful for the operating room team (Fig. 41.2).

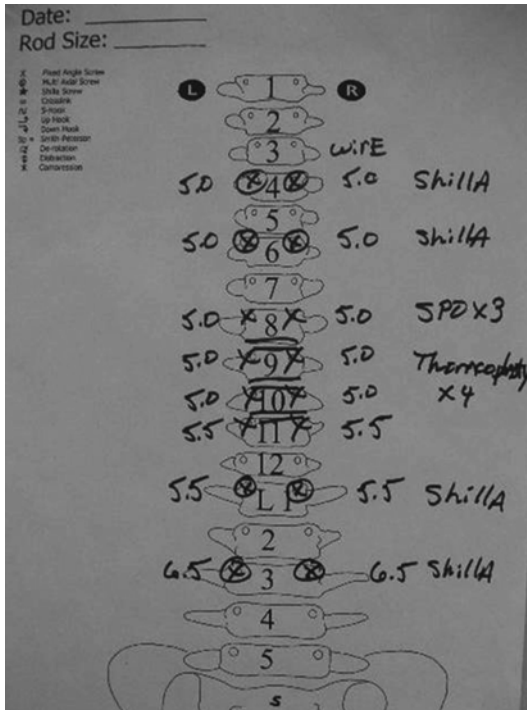

Fig. 41.2 Example of a blueprint planned preoperatively and placed in the OR where the operating room team can refer to it during surgery

A single midline incision has been used to approach the three areas of instrumentation. Before incision of the fascia, radiographs are taken with small needle markers placed in the spinous processes to identify levels. Subperiosteal dissection is isolated to the apical levels only.

The fascia is incised one centimeter off the midline on both sides of the spinous processes from cephalad to caudad merging with the subperiosteal dissection at the apex. With direct visualization and either a freehanded technique or using C-arm fluoroscopy, bilateral pedicle screws are placed throughout the apical levels with fixed head screws. If an apical thoracoplasty is being used to enhance correction of the rib hump or for bone graft harvesting, it can be accomplished from the midline incision through the paraspinal muscle layers, removing the medial 2 cm from the apical deformed ribs.

Ponte osteotomies are performed between the apical segments and will enhance correction in all planes. Apical decortication will be necessary for fusion of these levels.

The Shilla growth guidance screws are placed through the muscular layer without visualization of the bone except radiographically (Fig. 41.3a–f). A cannulated polyaxial screw of sufficient diameter to fill the pedicle is used. The location of the Shilla screws is curve dependent but should extend into the lumbar spine sufficient to control the lordosis and the coronal curve. Avoid stopping the caudal instrumentation at the thoracolumbar junction. The Shilla screws can be placed at bilateral locations or staggered but should be separated apart a sufficient distance on the rod to allow for sliding of the rod easily. The Shilla screws at the top of the construct are subject to pull out forces from kyphosis and are best protected with a sublaminar wire placed one level above the upper screws. The wire can be placed with minimal dissection leaving the inner spinous ligament intact and removing the ligamentum flavum with a small Kerrison rongeur. The double wire can be split after sublaminar passage and passed through the soft tissues to each side without lifting the periosteum. Fiberwire (5 mm) is a good alternative to wire (Fig. 41.4).

The rod diameter is chosen appropriate for the size of the child. For smaller-sized children, the 4.5 mm rods are satisfactory; smaller diameter rods will break prematurely. Larger children (greater than 30 kg.) can tolerate the 5.5 mm rods which are more resistant to stress fractures in the metal. The 4.5 rods generally last 4–6 years before fracturing; the 5.5 rods last longer. The rod is contoured with normal sagittal curves, and the rod is left one level long at each end for growth (Fig. 41.5a–d).

The apical levels are derotated with tube derotation devices or a vertebral column derotation device, while vice grips hold the rods in place to prevent rod rotation. The fixed head screws lock the rods at the apical screws via the locking set screws that press against the rods, while the Shilla caps capture the rods in the Shilla screws and press onto the polyaxial screw, not the rods. A cross-link is used to help counteract rod rotation. If the child is less than 5 years old, the cross-link should be avoided or a sliding type used to allow for growth in canal diameter. The torque/counter torque device snaps off the caps at a preset

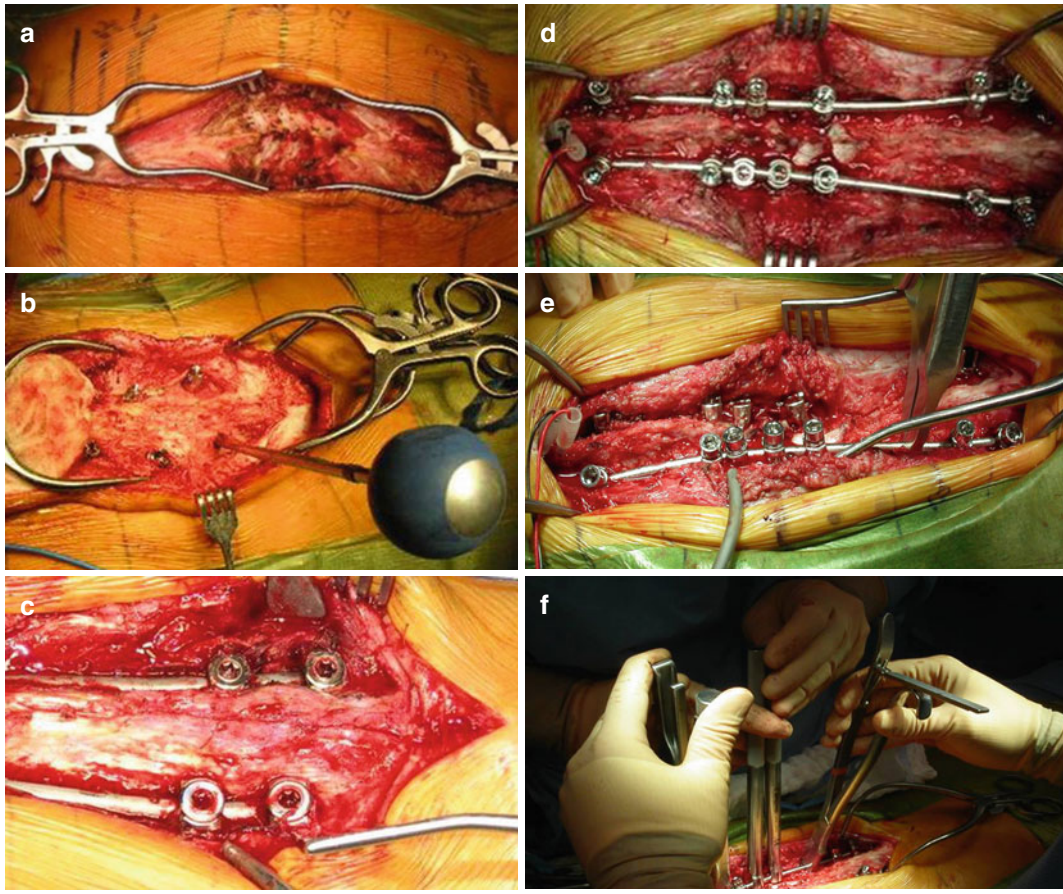

Fig. 41.3 (**a–f**) (**a**) A single incision is used, with levels identified using the C-arm, followed by subperiosteal dissection at the apex (cephalad on left, caudad on right). (**b**) Shilla screws are placed through the muscle. (**c**) And with C-arm guidance. (**d**) The rods are placed and rotated into the place with normal sagittal contours prebent into the rod. (**e**) Both rods in place. (**f**) Apical derotation with tube derotation

torque pressure. Bone graft is placed at the apex only. A small drain is often used.

A bivalved form fitting turtle shell brace for daytime use is recommended for 3 months until the apical fusion is established. Fragile skin may preclude use of the brace. After the initial period of immobilization, a protective brace is not necessary except if excessively vigorous activities are contemplated (Fig. 41.6a–i).

41.5 Clinical Experience

The Shilla Growth Guidance System has been used in the treatment of scoliosis 40° or greater (up to 115°, typically 60–80° range) in patients where spinal growth is anticipated for at least 3 years postoperatively. Single curve patterns (thoracic or thoracolumbar) are the most common curve pattern and the easiest to manage with a prototypical construct consisting of a bilateral apical fusion (2–4 vertebral levels) with Shilla pedicle screws cephalad and caudad to guide growth. Double major curves and single lumbar curves have been treated with Shilla constructs, but there is much less clinical experience in these curve patterns. Diagnoses for which the Shilla has been used include idiopathic early-onset scoliosis, congenital scoliosis, Beale's syndrome, myelomeningocele, Marfan's syndrome, neurofibromatosis, spinal muscular atrophy, arthrogryposis, multiple pterygium syndrome,

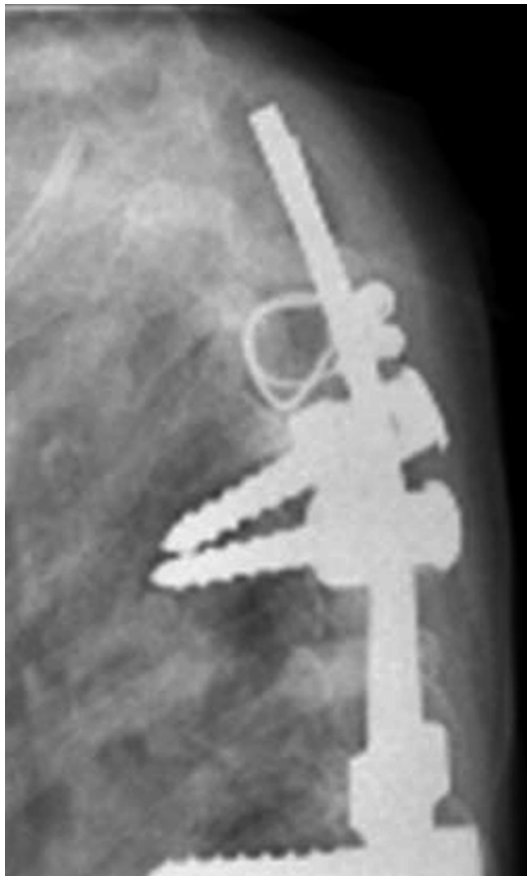

Fig. 41.4 Sublaminar wires can be placed cephalad above the Shilla screws to enhance fixation

and dwarfism. Ages at the time of surgery have ranged from 23 months to 11 years with the average patient being around 6–8 years. Curve magnitude, documentation of curve progression, and skeletal age have been the key determinants in the decision to operate. Figure 41.6 demonstrates a 70 % curve improvement at last follow-up 5 years postoperatively. She has returned to full activity (see Fig. 41.6) [8].

The Shilla spinal construct was first implanted in an animal model before widespread use in patients; the results at 6-month postimplantation demonstrated viable facet joints between the fused apex and the Shilla pedicle screws, as noted on microCT imaging. Degenerative facet changes were observed in the joints immediately adjacent to the Shilla pedicle screws. There was no evidence of apical stenosis over the 6 months of implantation (to skeletal maturity), and all the specimens documented longitudinal growth of the spine [7].

Comparison between Shilla spinal construct and growing rods (GR) in the treatment of children (<10 years of age) with progressive spinal deformity has recently been reported. [9] This was a multicenter retrospective study of the Shilla construct used as an alternative treatment to GR. Group demographics were similar between the two groups. The mean initial major curve magnitude was 70.3° for Shilla and 68.3° for GR, which decreased postoperatively to 22.4° (a 66.9 % improvement) and 32.2° (a 59.7 % improvement). The average decrease in Cobb angle at final follow-up was greater in the distracted GR group. At last f/u T1–S1 length was 35.4 for Shilla (6.4 cm increase from preoperative measurement) and 35.4 cm (8.7 cm increase from preoperative measurement) for GR.. Sagittal T2–T12 preoperatively was 36.3° for Shilla and 30.0° for GR. At 3-year follow-up, Shilla was 51.0° (14.7° increase) and GR 35.5° (5.5° increase). Sagittal T12–S1 preoperatively was −44.6° for Shilla and −55.0° for GR. At 3-year follow-up, Shilla was 57.0° (12.4° increase) and GR 52.0° (3.0° decrease). There were 29 reoperations in 12 of the 19 Shilla patients (63.2 %) and 43 reoperations in all 6 of the GR patients (100 %) related to the index procedure. The Shilla construct compares favorably with traditional GR constructs in terms of correction of the major curve, spinal length and growth, and maintenance of sagittal alignment. The greater than fourfold decrease in additional surgeries makes SGGS an attractive alternative to minimize comorbidities associated with additional surgeries.

The implant-related problems encountered with the Shilla construct include pedicle screw loosening, implant prominence, and rod fracture. Depending on the clinical presentation, these problems may necessitate implant revisions, which usually can be accomplished on an outpatient basis. However it must be noted that the reoperation rate for Shilla constructs is much less frequent (approximately 25 % as much) than the reoperation rate for traditional distraction-based growth rods. Symptomatic screw loosening

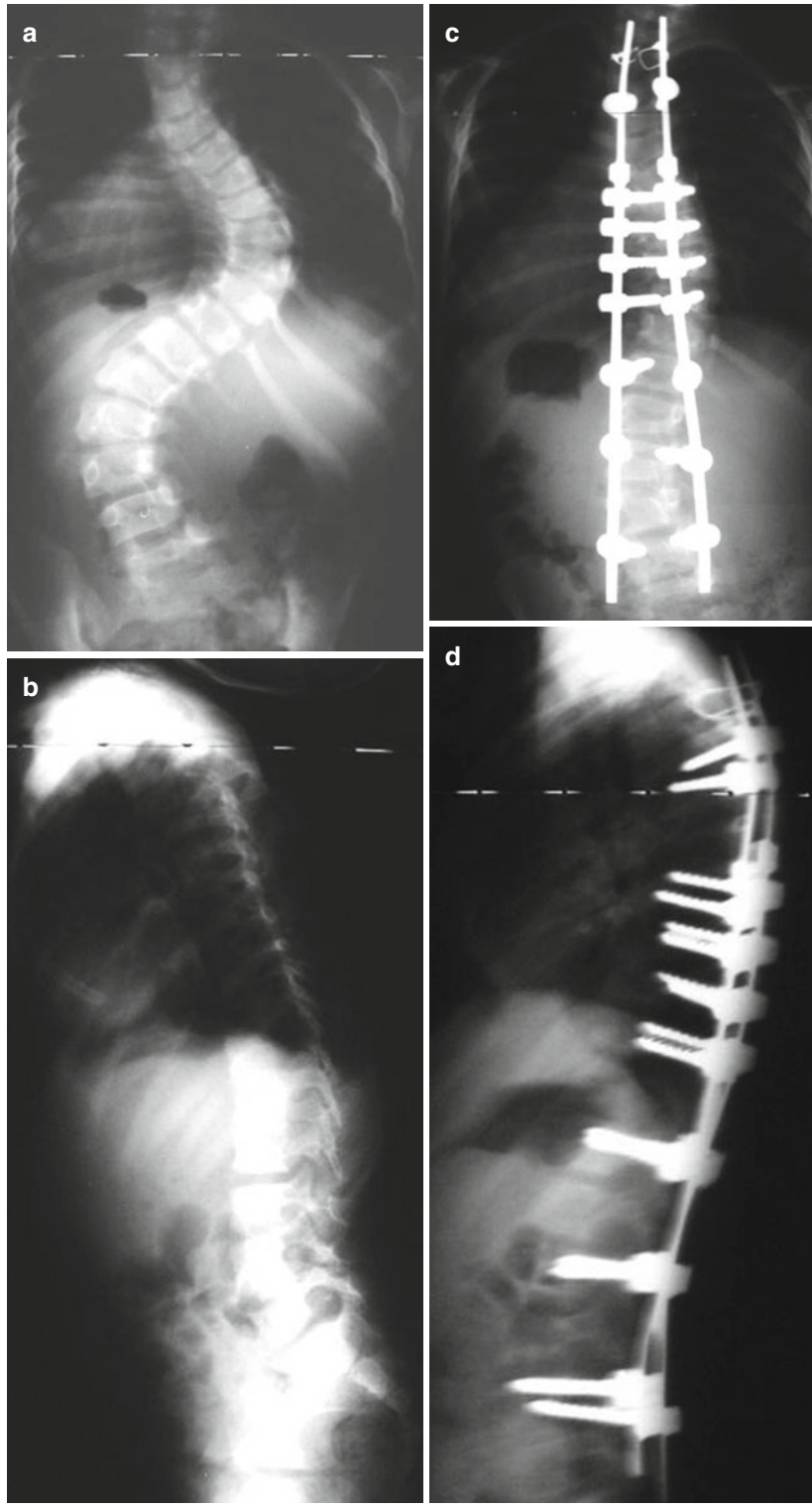

Fig. 41.5 (**a–d**) Preoperative (**a, b**) and postoperative (**c, d**) standing AP and lateral radiographs of a 4-year-old with Marfan's and scoliosis treated with Shilla procedure

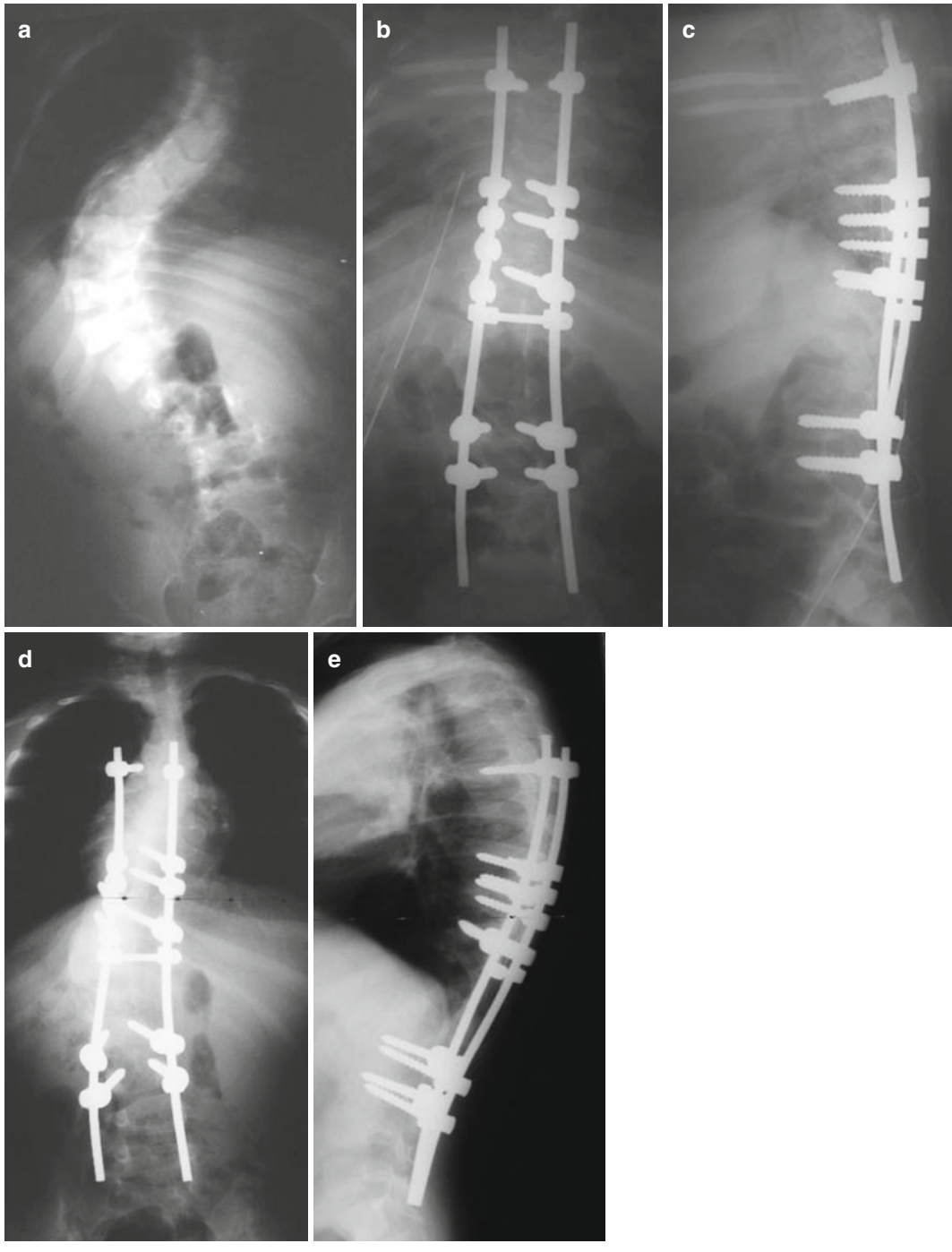

Fig. 41.6 (**a–i**) Preoperative (**a**), 6-week postoperative (**b**, **c**), and 5-year postoperative radiographs (**d, e**). Clinical photographs preoperatively (**f**, **g**) and 5 years postoperatively (**h, i**)

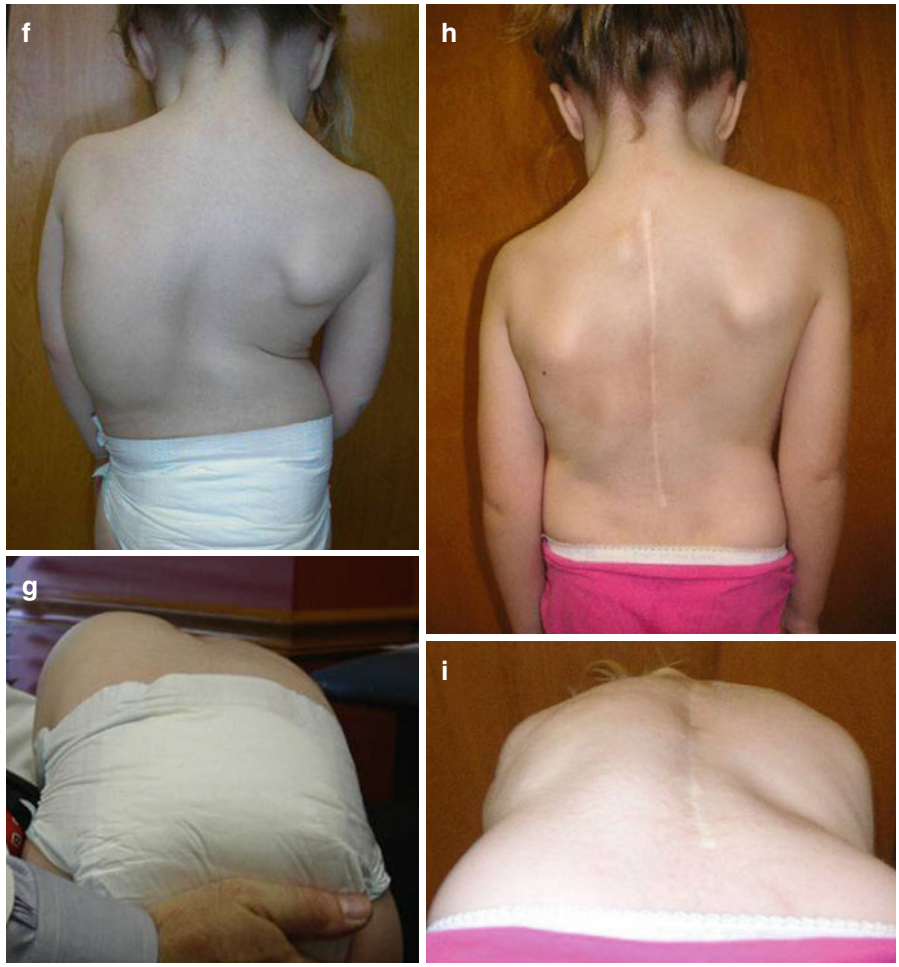

Fig. 41.6 (continued)

typically occurs at the most cephalad and caudal Shilla pedicle screws. This can be minimized by implanting the maximal pedicle screw diameter anatomically possible and as long as possible, which will optimize screw fixation by minimizing screw toggle and subsequent pullout. The smallest diameter Shilla pedicle screws used are 5.0 mm (at cephalad levels), but over 90 % of screws are a minimum of 5.5 mm (and up to 6.5 mm). In general, it is preferable to place pedicle screws which are at least the same diameter as the planned spinal rod. This is because it is much easier to surgically revise a broken rod than pedicle screw pullout, since the Shilla construct can only be optimally accomplished with intrapedicular pedicle screws.

Implant prominence can occur and is typically due to pullout (partial or complete) of the upper or lower pedicle screws, lack of adequate sagittal rod contouring, and/or patient size. Another method (in addition optimizing pedicle screw size) to minimize pedicle screw pullout is with the additional use of sublaminar wires or fiberwire (5 mm) placed cephalad to the cephalad-most screws. Also increasing the number of fixation points from the routine 4 Shilla screws (2 vertebral levels) to 6 Shilla screws (3 vertebral levels) at the cephalad and caudad end of the construct can be

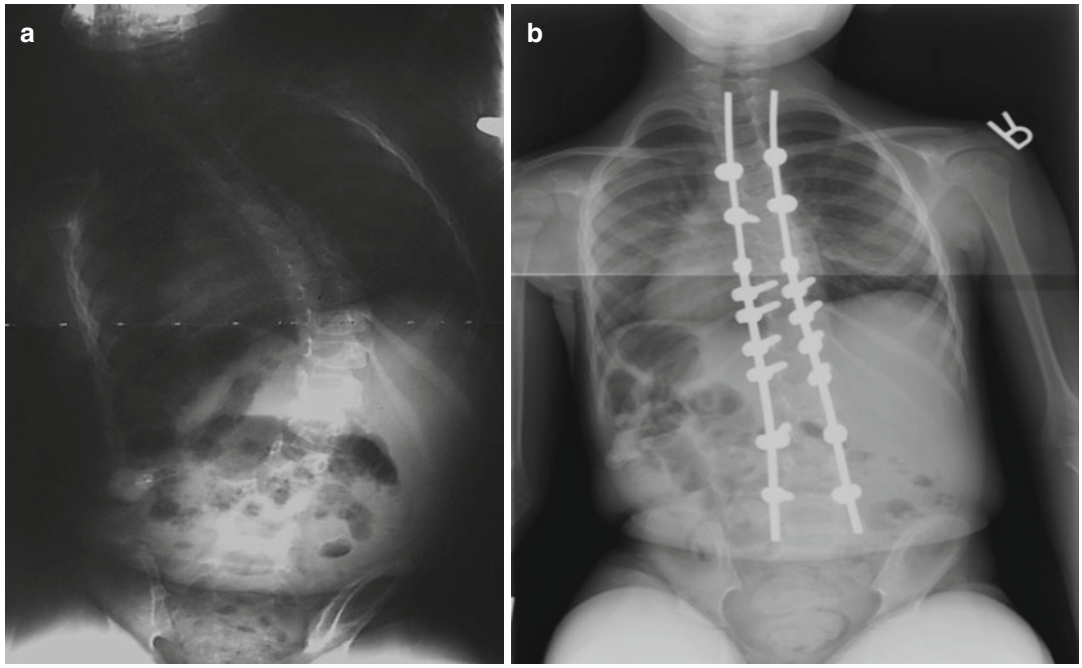

Fig. 41.7 (**a**, **b**) (**a**) Sitting preoperative AP spine radiograph of a 4-year-old with Dandy–Walker syndrome and spastic neuromuscular scoliosis with mild–moderate degree of flexible pelvic obliquity. (**b**) Postoperative sitting AP spine radiograph after Shilla procedure with instrumentation to L5 and a level pelvis

beneficial in cases with poor bone quality or proximal thoracic kyphosis. Sagittal rod contour is essential to minimize the ends of the rod from causes of painful dorsal prominence, specifically at the cephalad end. Extra kyphosis needs to be contoured into the construct since placement on the Jackson frame iatrogenically, and temporarily, decreases thoracic kyphosis. Inadequate cephalad rod kyphosis can become symptomatically prominent dorsally when the patient stands upright since the Shilla screws are only semi-constraining the vertebral position.

Rod fracture is not uncommon and typically occurs no earlier than 2 years of postimplantation. However, many patients never experience rod fracture during the time of Shilla implantation. When rod fractures do occur, surgical revision can be done with either a partial rod exchange (starting at apical screws and proceeding centripetally) of the fractured rod only or complete rod exchange of one or both rods. It is the authors' preference to completely exchange both rods when a single rod is fractured. By exchanging both rods in a single reoperation, the nonfractured, but fatigued rod, can be replaced and thereby minimize future contralateral rod fracture. Rod diameters of 5.5 mm are preferred and have fewer fractures than 4.5 mm rods, but it must be remembered that this system is a linked construct and screw failure should be the surgeon's primary concern. Also implant prominence must be considered.

Pelvic obliquity when flexible is best treated by placement of Shilla screws at L5 bilaterally, thereby balancing the pelvis (Fig. 41.7a, b). If a larger degree of pelvic obliquity is present especially when stiff, firm fixation must be used on the pelvis with screws at S1 coupled with iliac crest long screws bilaterally and linked to a cephalad directed rod. This is linked to an uncoupled domino attached to the rod coming inferiorly from the fixed apex. Once this is set up bilaterally, the pelvic obliquity is corrected and able to grow straight through the lower lumbar segments [9].

41.6 Summary

The Shilla growth guidance system allows for treatment of early-onset spinal deformities without the need for scheduled lengthenings. The Shilla harnesses the normal growth of the spine and maintains normal sagittal contours. It has the power to correct and maintain correction of pelvic obliquity and is applicable for a multiplicity of disorders causing spinal deformities.

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Growth-Guided Instrumentation: Luqué Trolley

42

Jean A. Ouellet, Catherine E. Ferland, and Hossein Mehdian

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Key Points

1. The key to successful management of EOS is the prevention of curve progression while maintaining spinal growth with the least amount of complications.
2. Self-guided growth surgical techniques have been developed to negate the need of repetitive lengthening required for the classic posterior distraction-based techniques (vertically expandable prosthetic titanium ribs/dual growing rods).
3. Implantation of self-guided growth construct is technically demanding and is best done in patients with flexible curves where the apex can be translated to midline, slightly older age group (6–10 years old) with underlying diagnosis of flaccid neuromuscular scoliosis such as spinal muscular atrophy.
4. There are two described self-guided growth constructs: the Shilla and the modern Luqué trolley. The main difference between the two constructs is that the Shilla procedure captures and fuses the apex of the deformity and allows the proximal and distal segments to grow away. The modern Luqué trolley construct consists of rigidly capturing the proximal and distal segments of the spine, while the apex of the deformity is

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translated and captured by gliding anchors.
5. Achieving apical translation is crucial to maximizing spinal height while minimizing the risk of curve regression as it realigns the axial forces of the spinal growth.
6. The gliding spinal anchors are inserted through muscle-sparing extraperiosteal "keyhole" dissections to avoid spontaneous fusion. At the apex of the deformity, gliding anchors are placed for maximal apical translation and deformity correction.

42.1 Introduction

The management of early onset scoliosis (EOS) carries significant challenges. Knowing that severe spinal deformities or early spinal fusion leads to poor lung development [1], new growth-sparing surgical techniques have evolved. The key to successful management of EOS is the prevention of curve progression while maintaining spinal growth [2, 3] with the least amount of morbidity. These new growth-sparing surgeries have been classified into three broad categories: distraction based, guided growth, and convex compression growth inhibition [4]. When deciding which of these growth-sparing procedures should be used, one must take into account the patients' underlying etiologies and their comorbidities. The most studied surgical options that have provided some hope for successful management of these challenging patients are the spine-based dual growing rods (DGRs) [5–8] and rib-based vertical expandable prosthetic titanium ribs (VEPTRs) [1, 9–11]. These two techniques carry a high complication rate with one major drawback: once implanted, the patients need to be returned to the operating room approximately every 6 months for lengthening procedures.

Recent literature revived interest in the previous concepts of Luqué of a spinal construct that allowed self-lengthening with growth [12–14]. The obvious advantage of this guided growth technique is that patients do not need repetitive surgical interventions to lengthen the implants. Both the Shilla procedure and the modern Luqué trolley consist of capturing the spine in such a way that gliding spinal anchors travel along fixed rods, preventing further spinal deformity while still allowing relatively normal spinal growth. The main difference between the Shilla and the modern Luqué trolley is that the Shilla procedure derotates and fuses the apex of the deformity and allows the proximal and distal segments to grow away, while the modern Luqué trolley consists of one pair of rods fixed proximally and one pair of rods fixed distally while the apex of the spine is translated and captured by the four rods. As the spine grows, the overlying rods glide away. The modern Luqué trolleys take advantage of modern spinal implants and of a better understanding of the physiology of the young growing spine. Patient selection is crucial when using the modern Luqué trolley treatment modalities to optimize successful management.

42.2 Philosophy

There is a general consensus among treating surgeons that conservative treatment consisting of serial casting, plus or minus bracing, is warranted as an initial treatment in all EOS cases [3]. It is true that casting can be successful in treating EOS in very young patients, particularly with small flexible curves [15]. It has been demonstrated that casting is also useful as a delay tactic buying time until the child is older to proceed to either a final fusion surgery or a growth-sparing procedure using DGRs or VEPTRs [16]. It has been demonstrated that by adopting such an approach, the overall complication rates in managing EOS will be decreased. By delaying the initiation of classic growth-sparing surgeries, one decreases the overall number of surgeries, delays the law of diminishing return [17], and decreases the overall potential for complications that have been quantified to be as much as 24 % for each additional surgery [5]. Currently, conservative treatment is just not feasible for certain patients

(respiratory compromise, neuromuscular etiology) or it is simply not successful (malignant curve progression despite casting). For such patients, only then, is surgery recommended.

When adopting growth guidance surgery such as the modern Luqué trolley, one must take a more proactive approach. Early surgical intervention is recommended rather than waiting until there are severe rigid spinal and/or chest wall deformities. However, such a philosophy must be based on strict guidelines as not to initiate unnecessary surgery. One needs to document curve progression in a child that remains skeletally immature and where there is a high likelihood that the curve will continue to progress. Thus, knowing that both the conservative treatment (serial casting) and classic posterior-based growth-sparing procedures require repetitive surgical intervention every 6 months, it is preferable to initiate self-growing rods to avoid these repetitive procedures, which carry a significant impact on the overall physical and mental health of the children. Pratt et al. concluded that the use of braces or plaster jackets for prolonged periods for EOS leads to an emotional scar [18]. They advocated the use of a self-lengthening construct such as the Luqué trolley, as a favorable option for EOS. They believed that the surgical scar could be more easily hidden and forgotten, in contrast to casting that is continually reminding the child of their abnormality. Therefore, they felt that the total physical and psychological trauma to the patient was smaller in children undergoing passive-guided growth surgery compared to bracing. Such surgery needs to be performed on curves that remain flexible and where the apex can be translated to midline. By achieving such correction, the axial forces of spinal growth will be "harnessed," maximizing spinal height while minimizing the risk of curve regression.

In addition to the benefit that the children do not need to be operated on serially, this growth-friendly surgery avoids the spinal elements (e.g., vertebral growth plates, disks, facets, and the spinal musculature) to be subjected to cyclical distractive and fixed constraints. Such unnatural loads across the spine during the classic repetitive lengthening may well contribute to the law of diminishing return seen with VEPTRs and DGRs [17]. Another physiological benefit of this guided growth surgery is that there are no posterior-based distractive force-inducing junctional kyphotic moments leading to sagittal imbalance. As the gliding anchors can travel up and down the rods matching the sagittal profile, there is also no set sagittal segment that needs to be straight for the growth to occur.

These self-guided growth constructs are particularly well adapted for patients with early onset neuromuscular scoliosis, particularly patients with spinal muscular atrophy (SMA). Type 2 SMA patients are particularly at risk of precocious severe spinal deformities, seeing the onset of the disease between 6 and 18 months and the onset of the spinal deformity by the age of 3 years [19]. These curves are at high risk of rapid progression resulting in significant deformity by the age of 7 years [19, 20]. The rationale for early surgical intervention in early onset neuromuscular scoliosis is to provide a straight and stable spine in order to allow proper-guided growth of the spine. Corrective spinal surgery protects the normal development of the lungs. In addition, it can help these patients to achieve a stable sitting balance and improved head control and overall posture, thus facilitating their caregivers' handling and improving their quality of life.

Patients with early onset or juvenile idiopathic scoliosis, congenital scoliosis, and to a lesser extent, spastic neuromuscular scoliosis are all candidates for guided growth. A key limitation behind this surgical technique is that if the spinal deformities require significant forces to straighten and maintain the spine straight, it will most likely not do well. For example, the spastic severely rigid neuromuscular patient may not grow as much as the flaccid collapsing neuromuscular scoliosis and its spinal deformity may return faster than the latter. Certain deformities require active distraction to ensure spinal growth, hence should be treated with classic DGRs and VEPTRs to maintain spinal correction and persistent spinal growth.

42.3 Background

The original Luqué trolley was described by Luqué and Cardoso in 1977 [21]. They developed the first self-growing rod construct consisting of

two L- or U-shaped rods fixed to the spine in a segmental fashion using sublaminar wires. Patients were selected for rigid internal fixation without fusion on the basis of young age (<11 years), severe long curves (e.g., wanting to avoid early long fusion), difficulty in casting (neuromuscular curves), and progressive curves [21]. As the spine grew, these rods were able to glide and "guide" the spine during longitudinal growth while maintaining the spinal correction. The short-term results of 2-year follow-up minimum were promising with mean major curve correction from 72° to 22° and spinal growth across the instrumentation of 2.5 cm. However, the use of the Luqué trolley has been abandoned as long-term result showed poor maintenance of spinal growth (range, 32–49 % of expected growth) [18, 22], high spontaneous fusion (range, 4–100 %) [22], and a high implant failure rate of 32 % [18].

Pratt et al. in 1999 published the long-term results of the Luqué trolley for the management of infantile and juvenile idiopathic scoliosis that were previously performed by Webb [18]. This retrospective study compared the Luqué trolley fixation with ($n=18$) and without ($n=8$) apical convex epiphysiodesis. In the Luqué trolley group without epiphysiodesis, the mean age was older (7 years old); the mean preoperative major curve was 48° and decreased to 25° immediate postoperatively. Over the next 5 years, all major curves worsened. Six of the seven patients underwent a second procedure consisting of the definitive spinal fusion with segmental spinal instrumentation. The major curves were corrected from 56° (range, 46–67°) to 43° (range, 24–55°), with a final major curve of 43°. With respect to spinal growth of the instrumented spinal segment at the 5-year follow-up (FU), it was 2.9 cm, representing 49 % (range, 31–71 %) of the expected growth for age- and gender-matched reference. For the other group of patients treated with the Luqué trolley with apical convex epiphysiodesis, the mean preoperative major curve was 65° (range, 40–95°). The mean major curve was 26° (range, 8–66°) after the combined anterior posterior surgery and 32° (range, 0–86°) at the 5-year postoperatively. Over a mean of 5 years postoperatively, the major curve worsened in seven patients, remained unchanged in four patients, and improved in two patients. While achieving better curve control (mean loss of correction of only 6°), spinal growth across the instrumented spinal segment at 5-year FU was only 2 cm, which represents only 32 % of that expected for age- and gender-matched norm groups. In the entire study group, there were three patients with broken rods and wires, two patients with broken wires alone, and three patients with rod prominence. A junctional kyphosis developed at the caudal end of two Luqué trolleys. At surgical revision, the instrumented vertebrae were found to be fused. One patient developed a postoperative pneumonia. There were no neurological complications. The authors concluded that there was a need for improved instrumentation and for new surgical measures to allow better spinal growth and curve control.

When choosing a growth guidance system, one needs to properly understand the shortfalls of the classic Luqué trolley. Patients who did poorly with the classic Luqué trolley were those with large rigid curves preoperatively and/or patients who had large residual postoperative curves. The usage of wires as the spinal instrumentation contributed directly to the causes of the high complication rates, including spontaneous fusion, implant failure, and poor deformity control. The dissection required to pass sublaminar wires at every level, and the binding of the rod down onto the lamina obviously led to a high rate of spontaneous fusion leading to growth inhibition. This posterior fusion, in turn, may have also contributed to a certain amount of curve progression in the form of crankshaft phenomenon. Despite such spontaneous fusion, previous authors have observed spinal growth across such extensive dissected spines [18]. Our belief is that the fusion mass is thin and does not impede the anterior spinal growth as long as proximal and distal fixation points are well anchored. Having converted Luqué trolley to final fusion, we have noted that these spontaneous fusions are generally thin and may explain persistent spinal growth. With respect to implant failure, it is not surprising that

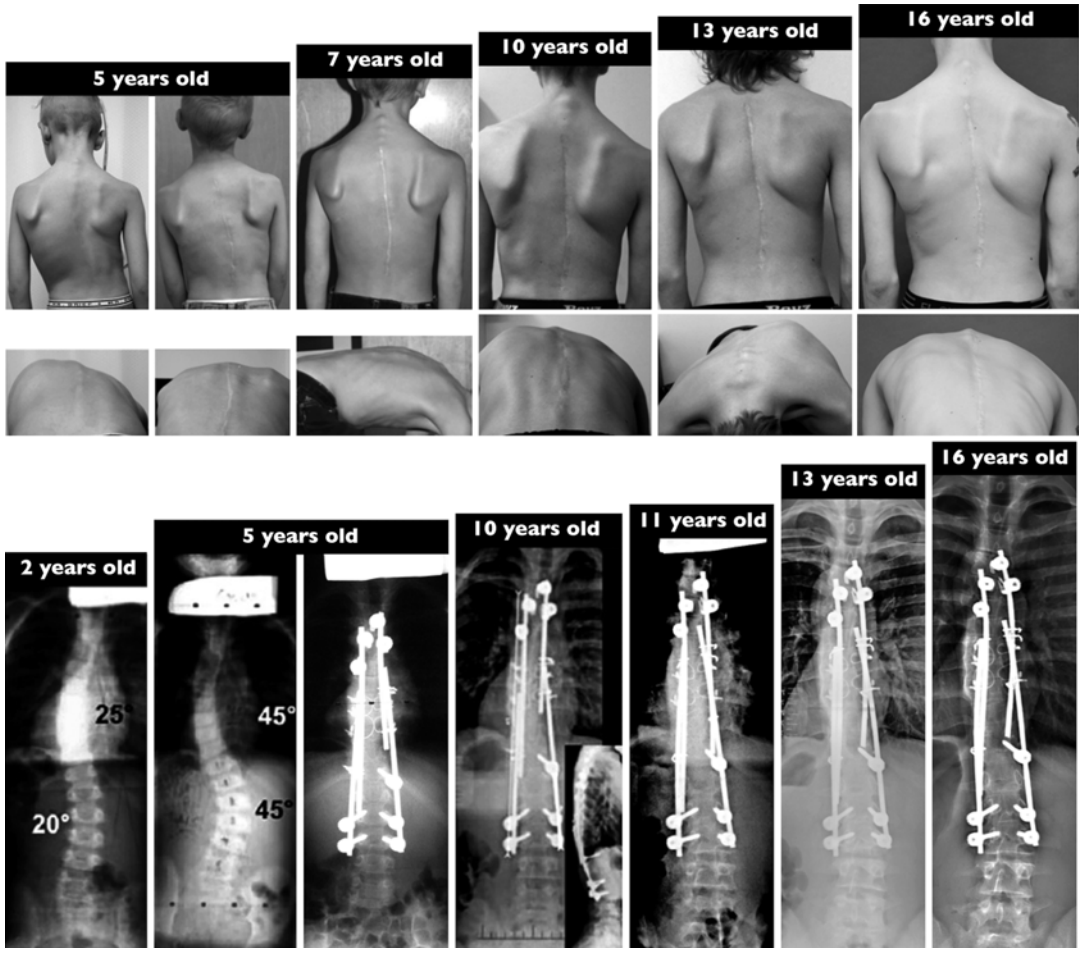

Fig. 42.1 Clinical example of a 2-year-old male with a progressive idiopathic early onset scoliosis undergoing a self-guided growth surgery. Despite serial casting from the age of 2–5, the deformity progressed. Patient was treated with a modern Luqué trolley construct, which grew over the next 10 years. He required only one revision surgery at the age of 5 as he outgrew the guided growth construct

there was a high rate of implant failure as the main implants used were simple wires. Rods could not be held in place solidly with only the wires; hence these had a tendency to migrate. With the use of wires, there was no ability to capture and control the anterior spinal column. Despite having every level "captured," the construct had to be loose to allow the rods to glide. With such fixation, the spinal stabilization was relatively poor, leading to poor curve control and therefore, contributing to the gradual loss of deformity correction. The patients in the study by Pratt et al. with the apical epiphysiodesis illustrated that curve control was improved significantly. However, it resulted in significant loss of spinal height, thus illustrating that apical control is indeed important for deformity control as long as one does not cause fusion across the apex.

In 2011, Ouellet published a small series of 17 patients with EOS of which 5 were treated with a modern Luqué trolley construct (Fig. 42.1) [12], reintroducing the concept of self-lengthening growth guidance systems [4]. The surgical technique consisted of using off-label modern spinal implants allowing for gliding spinal anchors and taking advantage of muscle sparing minimally invasive exposure to instrument the spine. The case series compared 12 patients treated with

conventional growth-sparing treatment (four patients treated with serial casting, four with DGRs, and four with VEPTRs) to 5 patients treated with a modern Luqué trolley. The etiologies of the deformities in these five patients were two patients with idiopathic EOS, two patients with syndromic scoliosis (Prader-Willi syndrome and a child with dysmorphic feature with global hypotonia of unknown etiology), and one patient with neuromuscular scoliosis (cerebral palsy). The mean age of the serial casting and distraction-based patients was 4.5 years old (range, 0.9–8.5 years) compared to 6.5 years old (range, 3–8.6 years) for the modern Luqué trolley group. Mean preoperative major curves were 61° (range, 38–94°) and 60° (range, 45–75°) and decreased to a mean of 21°(range, 10–33°) and 35° (range, 23–46°), respectively. Mean follow-up was 4.5 years (range, 2.5–6 years) and 5 years (range, 3–8 years) for the two groups. At the last follow-up, the mean major curve had increased to 31° (range, 14–54°) in both groups. At 5 years postoperatively, four out of five subjects (80 %) had required revision surgery. Three had their initial self-guided growth implants converted to new distraction-based implants as they had outgrown the initial construct. A fourth patient, with syndromic scoliosis, required final spinal fusion before reaching skeletal maturity because the curve had progressed (54°) and had minimal remaining spinal growth (26 % expected). The fifth patient was still immature and growing. Comparing the two groups, the first treatment group had a total of 89 procedures over a 4.5-year period, with a mean of 7 procedures per patient and 1.7 procedures per year, per patient. In contrast, the modern Luqué trolley had a total of 9 procedures over a 5-year period, resulting in 1.8 procedures per patient and 0.3 procedures per year. In respect to spinal growth, after the mean follow-up of 5 years, the spine grew on average 67 % (range, 26–91 %) of expected growth.

At the 2013 Scoliosis Research Society's Annual Meeting, Mehdian et al. presented their experience with the self-growing rod (SGR) system in patients with neuromuscular scoliosis (Fig. 42.2) [23]. Their SGR system is a growth guidance construct and is, in effect, equivalent to the modern Luqué trolley. They reported a total of 15 consecutive patients (Table 42.1). There were eight male and seven female patients, with a mean age of 7.4 years (range, 4–9 years). The instrumentation extended from T2 to the pelvis (including sacrum) in all patients. The diagnosis included SMA type 2 in six patients, SMA type 3 in three patients, hypotonia in two patients, and congenital muscular dystrophy in four patients. The mean blood loss and percentage of blood volume was 523 ml/19.7 % (range, 420 ml/17–640 ml/26 %). The mean follow-up was 3.5 years (range, 2–6 years). The mean length of pediatric intensive care unit stay was 2.7 days (range, 2–6 days) and mean hospital stay was 9 days (range, 7–11 days). The mean operation time was 5.3 h (range, 4–8 h). The mean preoperative major curve was 69° (range, 40–110°), 16° (range, 6–20°) immediately after surgery ($p=0.001$), and slight loss of correction with a 18° Cobb angle (range, 7–41°) at final follow-up ($p=0.001$). The mean preoperative thoracic kyphosis was 75° (range, 57–98°), 23° (range, 15–34°) immediately after surgery ($p=0.001$), and 28° (range, 22–38°) at final follow-up ($p=0.001$). Patients maintained their sagittal alignment without the appearance of any junctional kyphotic deformity. The maintenance of correction was statistically significant (Table 42.2). The mean preoperative coronal balance was 12 cm (range, 7.5–16 cm), 4 cm (range, 1–6.5 cm, $p=0.005$) postoperatively, and 8 cm (range, 3.5–15 cm, $p=0.036$) at final follow-up. The mean preoperative pelvic obliquity was 35° (range, 28–41°), 5° (range, 0–14°, $p=0.001$) postoperatively, and 12° (range, 3–21°, $p=0.005$) at final follow-up. The values for both measurements were statistically significant at final follow-up. In respect to spinal growth, the mean preoperative T1–S1 height was 25 cm (range, 22–30 cm), 32 cm (range, 28–35 cm) postoperatively, and 37 cm (range, 32–42 cm) at final follow-up. The T1–S1 height change was statistically significant ($p=0.002$). The mean yearly growth of the spine was 1.4 cm (range, 0.7–2.5 cm). There were no lengthening procedures performed in any of the cases. These

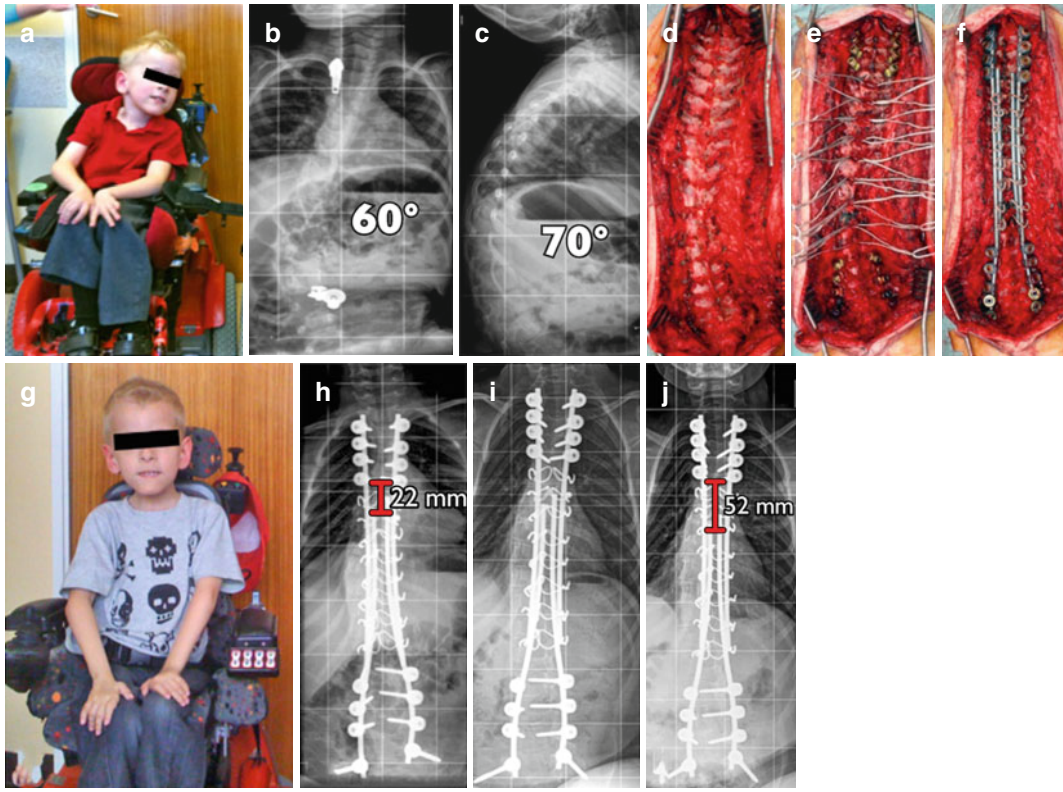

Fig. 42.2 Clinical example of a self-guided growth construct as described by Mehdian et al. treating an early onset neuromuscular scoliosis in a child with spinal muscular atrophy. (**a**) Preoperative clinical pictures. (**b**, **c**) Sitting AP and lateral x-rays pre-op. (**d**) Intraoperative pictures illustrating direction. (**e**) Multiple segmental sublaminar wires fixation points. (**f**) Intraoperative correction with four rods. (**g**) Postoperative clinical pictures. (**h**–**j**) Postoperative x-rays immediate, 1 year, 3 years, respectively, confirming spinal growth of 30 mm

results are markedly different than the unfavorable long-term result from Mardjetko et al. [22]. Mehdian et al. [23] also studied the impact of growth guidance surgery on the chest width and lung function. The mean preoperative chest width T6/T12 ratio was 0.8 (range, 0.7–0.9), 0.7 (range, 0.6–0.8, $p=0.004$) postoperatively and 0.7 (range, 0.6–0.8) at final follow-up. The mean preoperative functional vital capacity was 64 % (range, 59–74 %), 67 % (range, 61–77 %) postoperatively, and 57 % (range, 50–61 %) at follow-up. Two of the 15 patients (13 %) experienced complications. One patient had failure of fixation due to distal screw pullout from the iliac wing and required revision surgery. A second patient developed spinal infection that was treated with antibiotics for 6 weeks.

42.4 Surgical Technique

For the modern Luqué trolley, patients are positioned prone on a radiolucent table under a total intravenous general anesthetic compatible with multimodality spinal cord monitoring. Preoperative planning is mandatory to plan the skin incision as well as the location of the gliding anchors. Classic midline incisions are to be made ensuring that no prominent spinal implant will be directly below the skin incision. Either one single skin incision is made spanning the entire planed instrumented spine (Fig. 42.3a). Two or three separate skin incisions can be made over the proximal, apical, and distal segments (Fig. 42.3b). Currently, only the Shilla system has been FDA approved as gliding anchors; it has a

Table 42.1 Control of spinal deformity of patients with early onset neuromuscular scoliosis treated with the self-growing rod (SGR) system

ID	Sex	Diagnosis	Complication	Age	Follow-up (years)	Preoperative scoliosis (degrees)	Final scoliosis (degrees)	Preoperative kyphosis (degrees)	Final kyphosis (degrees)
1	F	Hypotonia	None	9	6	40	7	58	22
2	F	SMA	Failure of fixation due to pullout from the iliac wing	9	6	60	41	98	38
3	F	Hypotonia	None	9	5	92	8	76	31
4	M	SMA	None	9	5	78	38	71	32
5	M	Muscular dystrophy	None	9	3	71	7	76	30
6	M	SMA	None	8	5	73	7	81	29
7	M	Muscular systrophy	None	6	4	68	30	89	29
8	F	SMA	None	6	3	72	16	63	31
9	M	Muscular dystrophy	None	8	2	60	11	71	26
10	F	SMA	None	6	4	40	7	57	22
11	M	SMA	Superficial wound infection that was treated with antibiotics	7	2	110	21	81	23
12	M	SMA	None	6	2	59	25	93	31
13	F	SMA	None	4	2	70	17	68	27
14	M	Muscular dystrophy	None	8	2	72	20	74	29
15	F	SMA	None	7	2	77	23	76	27
Mean				**7.4**	**3.5**	**69.47**	**18.53**	**75.47**	**28.47**

SMA spinal muscular atrophy

Table 42.2 Mean (range) of preoperative, postoperative, and final measurements of patients with early onset neuromuscular scoliosis treated with the self-growing rod (SGR) system

	Preoperative	Postoperative	Final
Scoliosis	69° (40–110°)	16° (6–20°)	18° (7–41°)
Kyphosis	75° (57–98°)	23° (15–34°)	28° (22–38°)
Coronal balance	12 cm (7.5–16 cm)	4 cm (1–6.5 cm)	8 cm (3.5–15 cm)
Pelvic obliquity	35° (28–41°)	5° (0.3–14°)	12° (3–21°)
T1–S1 height	25 cm (22–30 cm)	32 cm (28–35 cm)	37 cm (32–42 cm)
T1–S1 height growth	Average yearly 1.4 cm (0.7–2.5 cm)		
T6–T12 ratio	0.8 (0.7–0.9)	0.7 (0.6–0.9)	0.7 (0.6–0.8)
FVC	64 % (59–74 %)	67 % (61–77 %)	57 % (50–61 %)

FVC functional vital capacity

special locking cap that does not bind to the rods. There is also a trolley-gliding vehicle that is currently only available in Europe that captures the rod with a cable tie mechanism allowing for gliding. Other implants can be used in such a fashion allowing for certain gliding properties. The

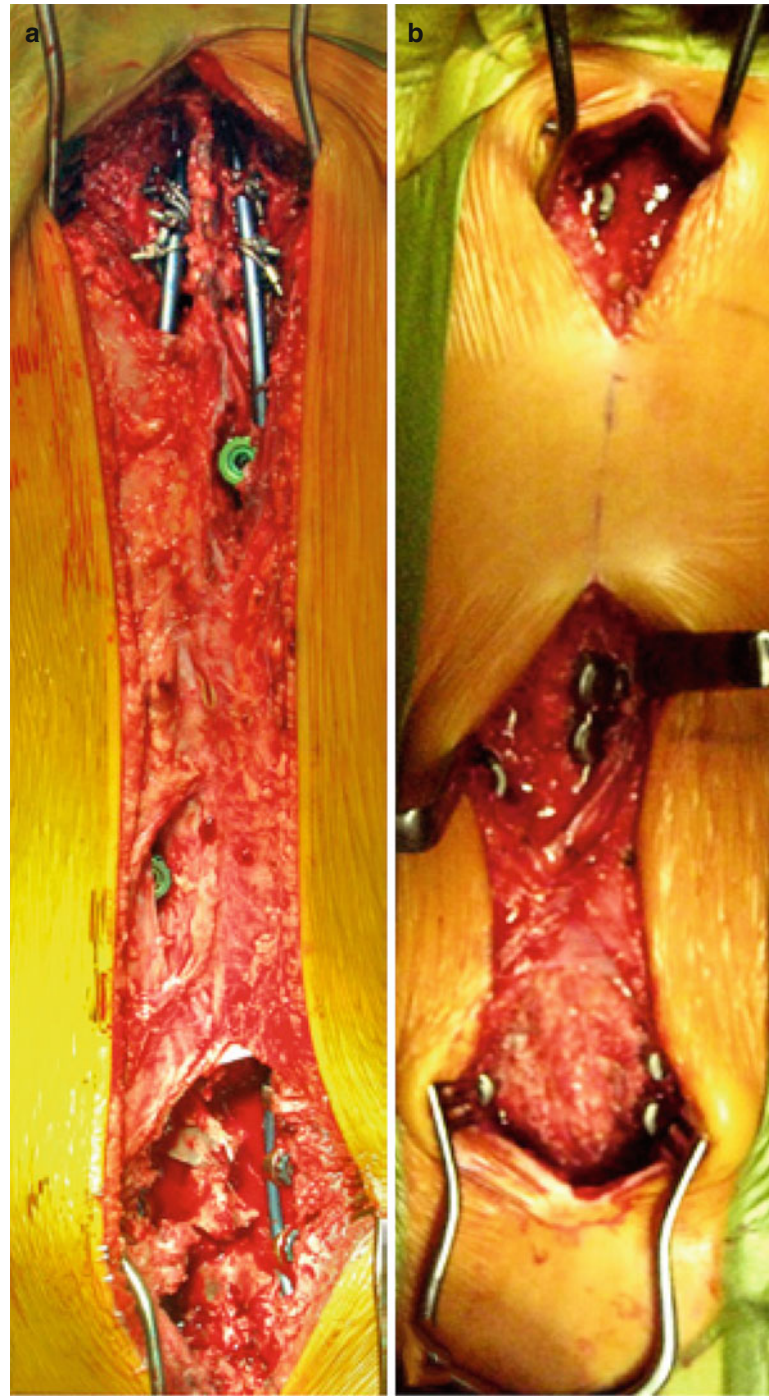

Fig. 42.3 Midline incisions: either one single skin incision spanning the entire planed instrumented spine (**a**), or two or three separate skin incisions over the proximally, apical and distal segments (**b**) can be performed

oldest segmental fixation is a sublaminar wire and it can be used as a gliding anchor. The other possibility is to purposely use a smaller diameter rod (5 mm) in a pedicle screw-based system designed to capture a larger diameter rod (6 mm). For example, the pedicle screws of the AO universal spine system can be used with its small stature AOUSS 5-mm rods. Obviously, using spinal instrumentation in this way is off-label and is not recommended by any of the manufacturers.

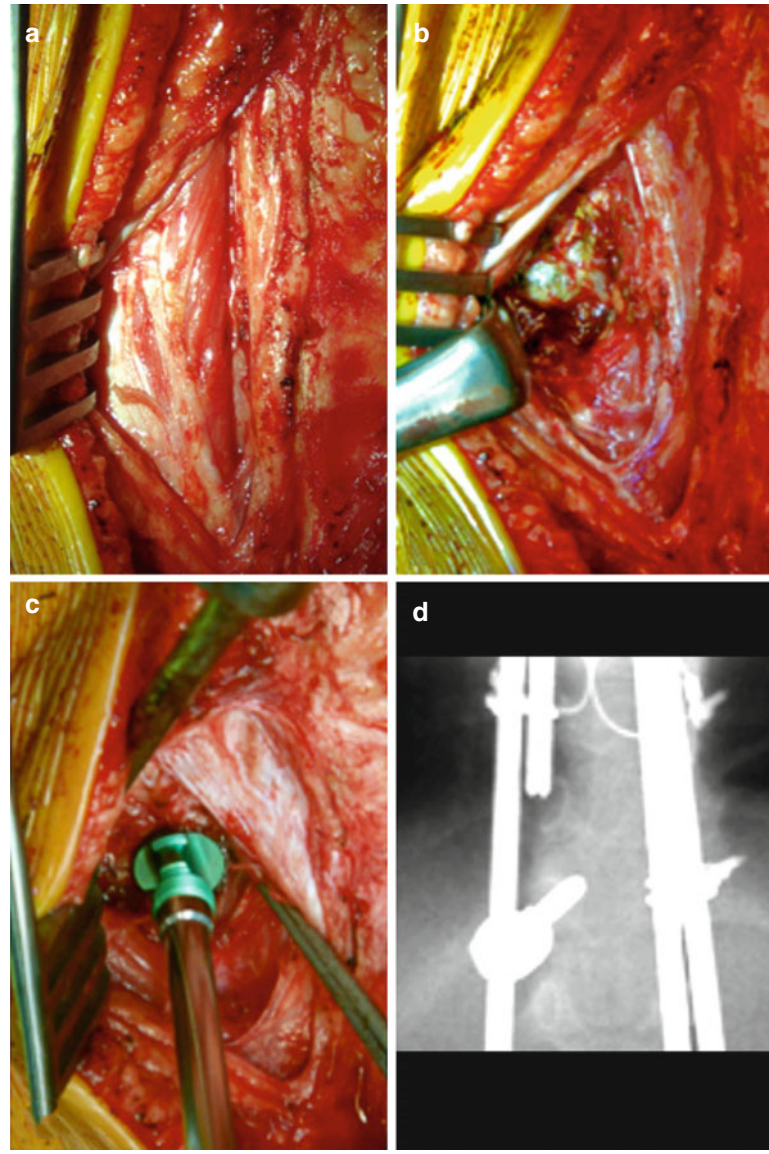

Fig. 42.4 The erector spinae are split with the multifidus and spinalis spinous process left medially with the longissimus and iliocostalis reflected lateral (**a**). Transverse process is visualized (**b**). Freehand or fluoroscopic-assisted gliding screws are inserted (**c**). Example of a gliding screw translating apex across midline (**d**)

The classic modern Luqué construct consists of fixed proximal and distal anchorage points. A classic subperiosteal dissection is performed at the proximal and distal segment, as these segments need to be fused to achieve long-term solid anchors. Fixed spinal anchors such as standard screws or hooks locked to the rods are inserted. The gliding spinal anchors (either gliding screws or sublaminar wires free to travel along the rods) are inserted through muscle-sparing "keyhole" dissections (see Fig. 42.3a, b). At the apex of the deformity, gliding anchors are placed for maximal apical translation and deformity correction. The dissection at the gliding anchors must be kept to a minimum using extraperiosteal and muscle-sparing techniques to avoid spontaneous fusion. In the lumbar spine, the gliding pedicle screws are inserted through a Wiltse approach sparing the joints and minimizing bony exposure. In the thoracic spine, the gliding pedicle screws are inserted laterally to the midline erector spinae, dissecting directly onto the transverse process avoiding exposure of the lamina (Fig. 42.4a, c). Pedicle screw insertion should be done with the

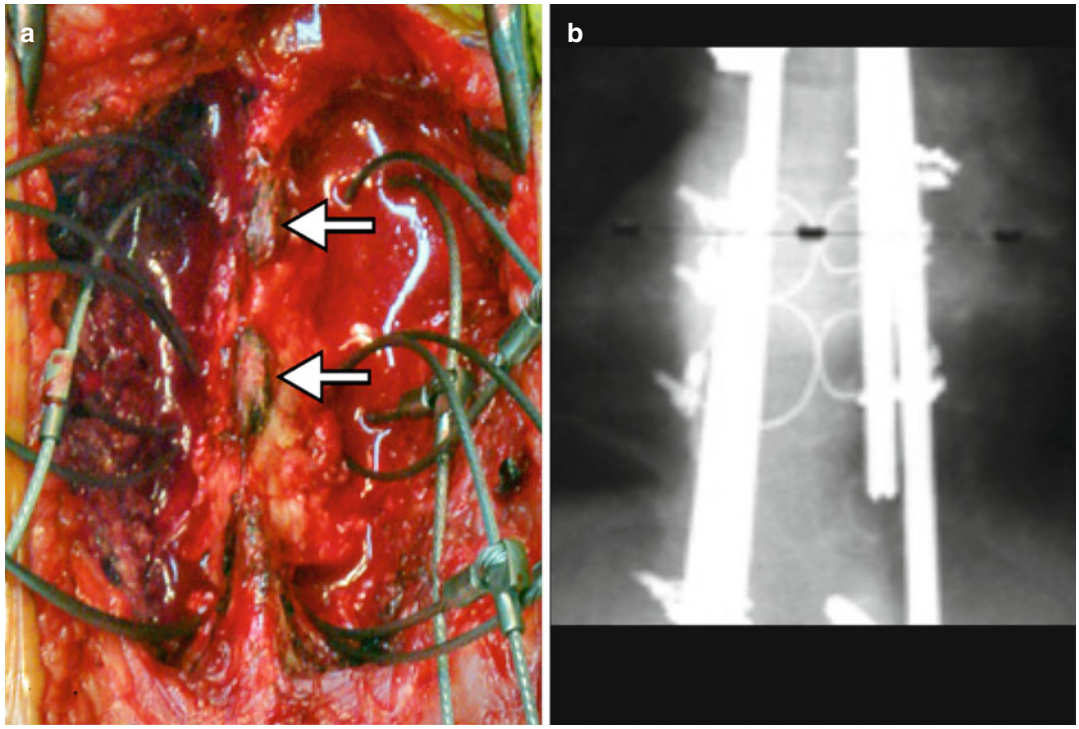

Fig. 42.5 Wires are inserted not via the standard midline ligamentum flavum resection but rather via small lateral laminectomy leaving the periosteum intact (*arrows*) (**a**). Example of apical sublaminar wires capturing the overlapping rods (**b**)

use of intraoperative imaging. Fluoroscopy can be used to confirm the pedicle entry point, and using a freehand technique, the gliding screws can be inserted at strategic points allowing for maximal apical translation. These gliding screws capture a 5-mm rods with a locking cap designed for 6-mm rods, thus permitting motion. At segments where sublaminar titanium cables are to be passed, the dissection is carried from midline to the medial border of the facet. Careful attention should be paid in order to leave the periosteum on the bone even with some muscle still attached. Dissection is to be performed with bipolar cautery and forceps at hand to control blood loss and minimize disruption of the periosteum. Avoid removing the spinous processes to prevent stripping the periosteum off the lamina and creating a raw bone surface. Small lateral laminectomies are to be done leaving the periosteum intact, while giving access to the ligamentum flavum. Once the central ligamentum flavum is removed, passage of sublaminar cables can be performed (Fig. 42.5). Once the fixed and gliding anchors are placed, two pairs of 5-mm titanium rods are tunneled in a subfascial/intramuscular fashion (below the fascia, above the periosteum) from the opened proximal and distal incisions. Each rod needs to only have one end rigidly anchored to the spine. In the intermediate segments, a series of gliding spinal anchors maintains the correction by keeping the rods parallel and engaged. As the spine grows, the rigidly proximally fixed rods will move away from the distally fixed rods (Fig. 42.6a). One can also only use two rods rather than four and have them fixed distally and have the spine grow off the proximal end (Fig. 42.6b). Correction of the spinal deformity is achieved with either a classic rod derotation maneuver (Fig. 42.7a) or an apical translation reduction maneuver (Fig. 42.7b) or in combination. As the rods are tunneled and partially engaged in the fixed and gliding anchors, and by rotating or translating

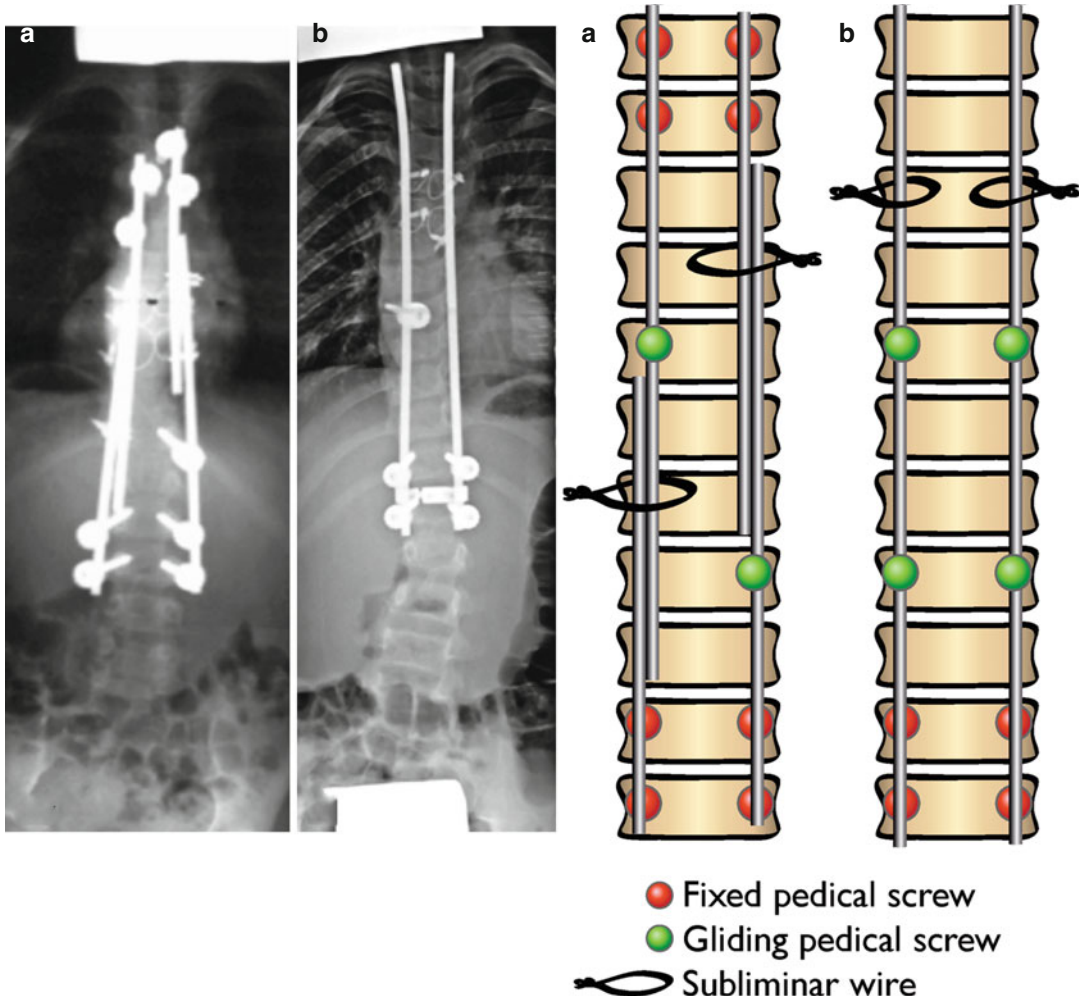

Fig. 42.6 Radiographic and schematic differences between two self-growing constructs: the modern Luqué trolley (**a**) and an alternative-guided growth construct (**b**). A series of gliding spinal anchors maintains the correction by keeping the rods parallel and engaged. As the spine grows, the rigidly proximal-fixed rods will move away from the distally fixed rods

the rods, the correction is achieved. The goal is to ensure that the four rods are parallel to each other. The number of gliding anchorage points will influence the ability to correct and maintain the deformity. If the number of the gliding anchors is kept to a minimum, the risk of spontaneous fusion is minimized. However, the risk of residual and recurrence of the spinal deformity is greater (Fig. 42.8). In contrast, if every spinal segment is instrumented then there is a lower risk of curve progression but a higher risk for growth retardation as spontaneous fusions may occur. The key is to have an adequate number of gliding anchors to translate the apex of the deformity toward midline, ensuring adequate correction and control of the spinal deformity without inducing spontaneous fusion. Different gliding constructs can be tailored to different spinal deformities (Fig. 42.9). This case illustrates the power of cantilevering a rod across the apex of a deformity. The spine was captured with fixed spinal anchors proximally (hooks and screws) and was then cantilevered across the two eggshell resections of the hemivertebra with an apical gliding screw

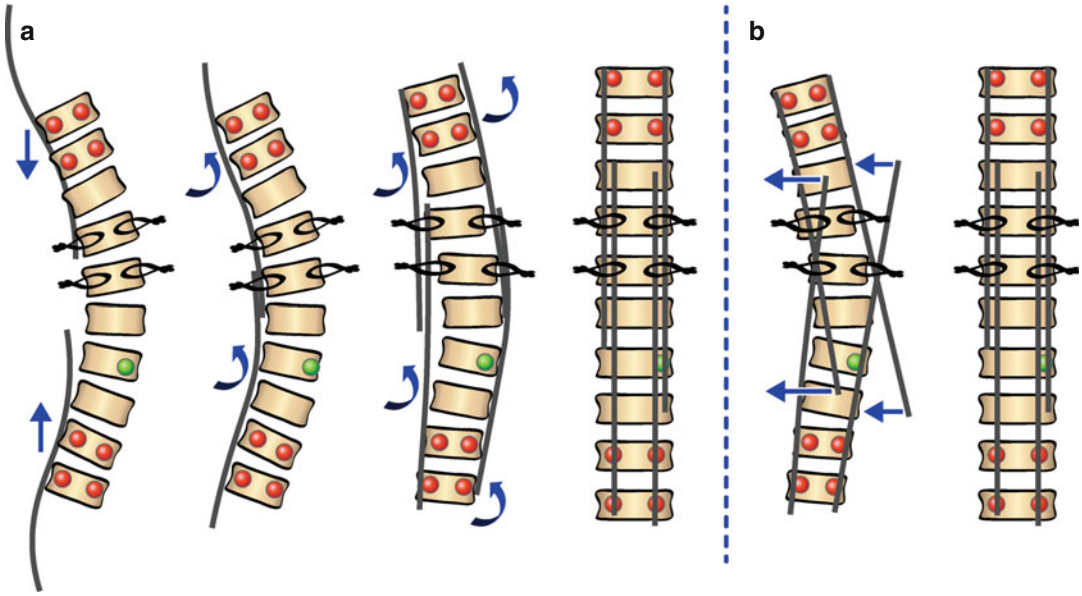

Fig. 42.7 Schemes of the technique of reduction. Correction relies on rod rotation and apical translation. Rods are attached to proximal and distal anchors. The latter are then cantilevered and/or rotated across the midline achieving parallel end vertebra

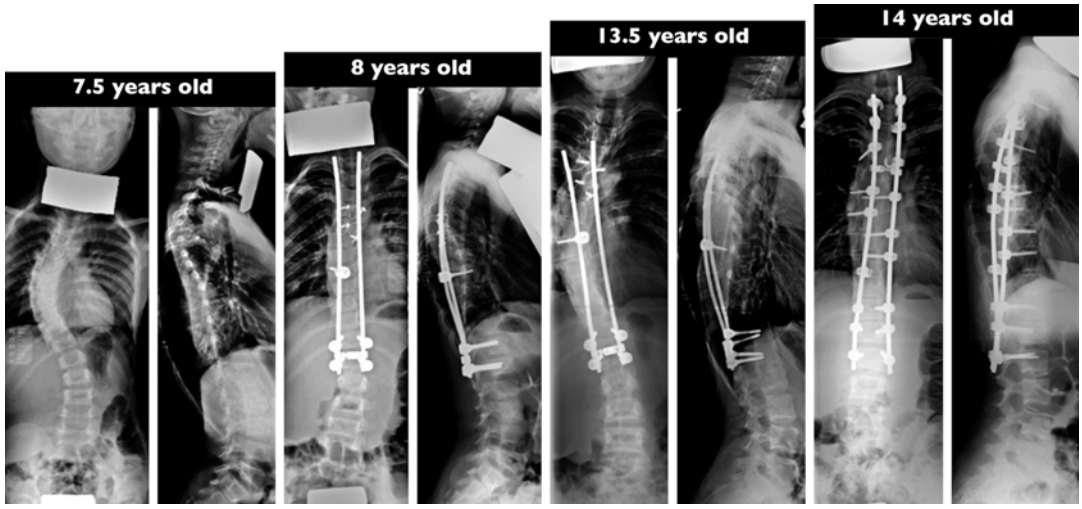

Fig. 42.8 Clinical example of a modern Luqué trolley with inadequate numbers of gliding anchors. Initially, deformity appeared under control. However, over the next 4 years, due to inadequate number of gliding anchors, deformity recurred requiring formal posterior spinal fusion observed 6 months following surgery (8 years old). Five years post-initial trolley (13.5 years old), a loss of proximal fixation, growth across the instrumentation, and a 75 % of normal growth without any lengthening surgery could be observed. Final fusion occurred at 14 years of age

and a set of gliding anchors distally. Follow-up radiographs confirm ongoing growth of the spine. Initially, the left rod extended below the disk of L5/S1 and now is at the level of the L5 pedicle screw. On the right side, a VEPTR 2 implant was used without the locking mechanism that allows for passive-guided growth. The gradual appearance of space within the male-female inlay of the

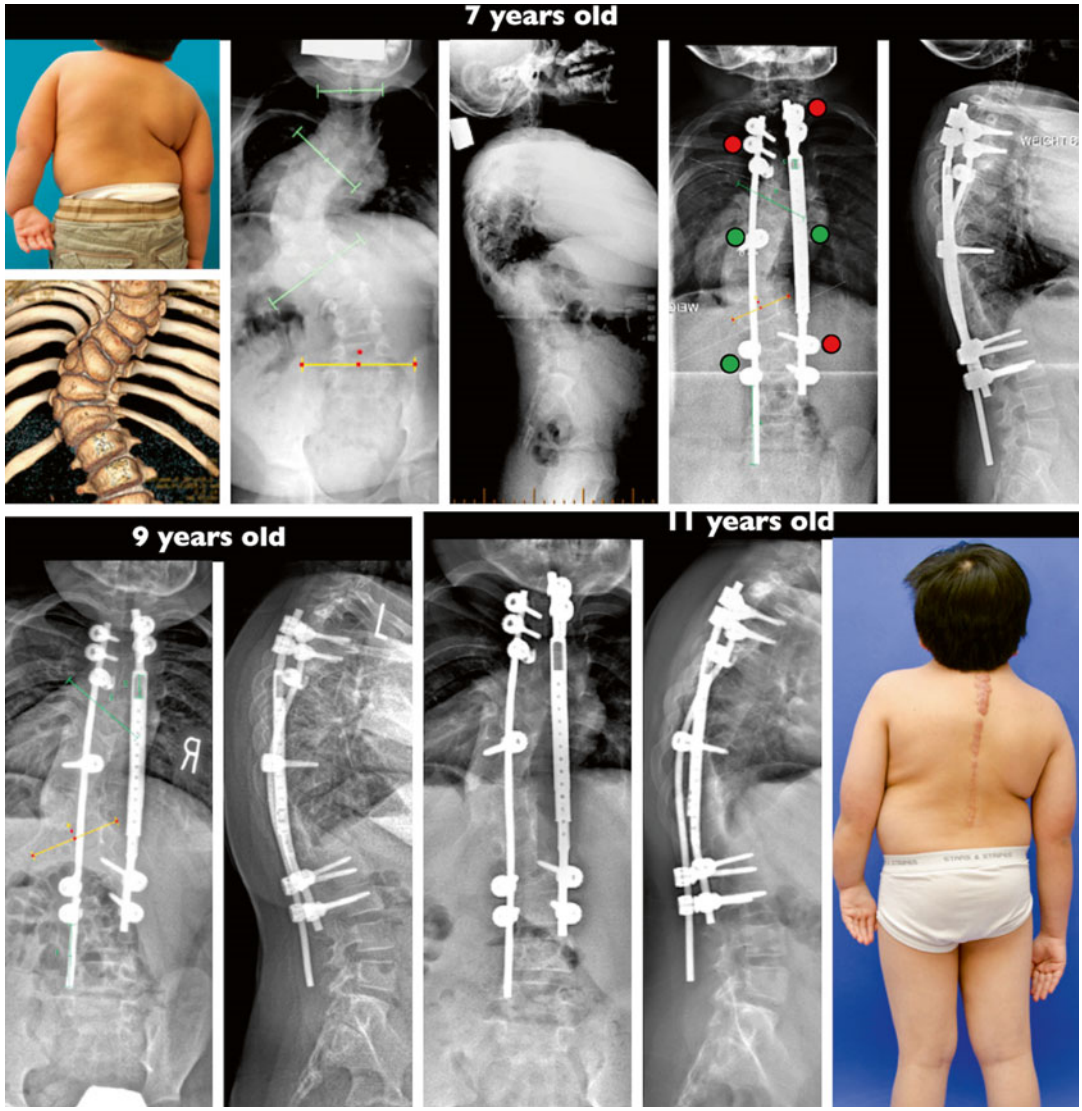

Fig. 42.9 Modified modern Luqué trolley treating early onset scoliosis in a 6-year-old male patient with severely rigid congenital scoliosis with radial hypoplasia. Hybrid construct with a left-sided proximally fixed rod with mid- and distal gliding screws. The right-side construct is a VEPTR used off-label that is not locked, thus allowing for self-growth

VEPTR implants represents the spinal growth across the instrumented spinal growth.

The surgical technique, as described by Mehdian et al. [23], has a similar four 5-mm rod construct with solid proximal and distal fixed anchor having more than six fixation points, which distally includes the pelvis. It differs from the modern Luqué trolley as to the more extensive classic spinal surgical dissection where every intercalated vertebra is exposed and captured with sublaminar wire (Fig. 42.2). Despite such extensive dissection, multiple cases have shown ongoing spinal growth (Fig. 42.10). Sublaminar wiring can be time consuming and possibly risky. Two 5-mm stainless steel rods are then contoured to accommodate thoracic kyphosis and lumbar lordosis. Rods are secured to the proximal and distal screws on either side or the middle section by sublaminar wires. Pelvic fixation should always

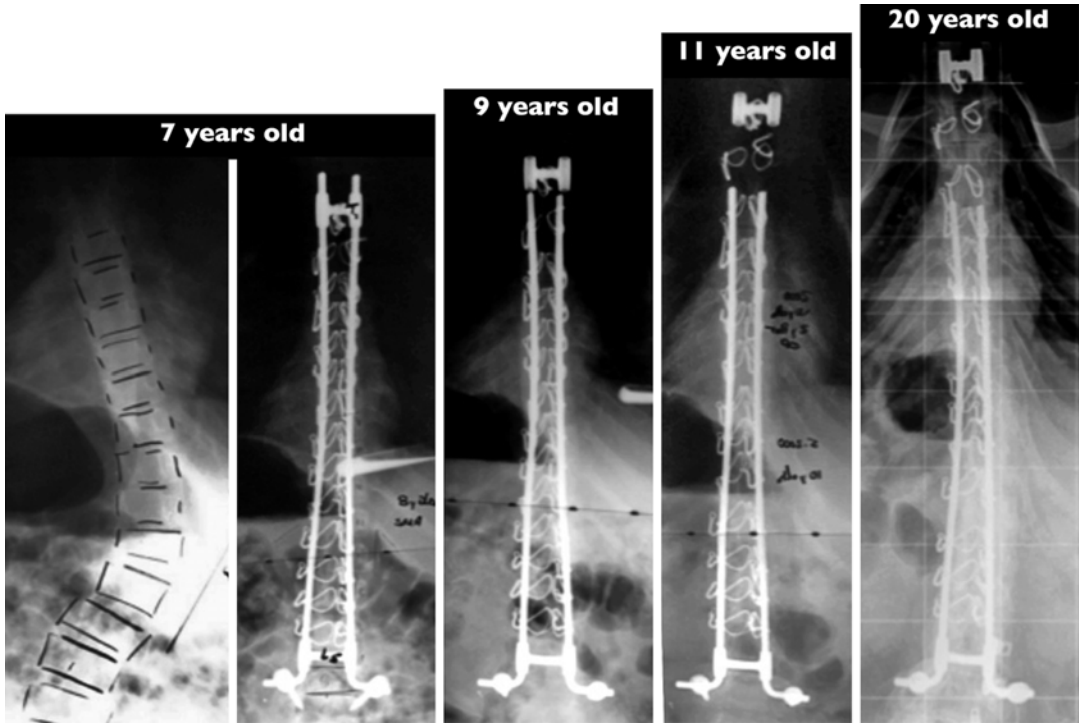

Fig. 42.10 Evidence of guided growth (10 cm) over a 13-year follow-up period

be considered in children with neuromuscular scoliosis due to the collapsing nature of the deformity and the propensity to pelvic obliquity [24]. The extension of the instrumentation from T2 to the pelvis not only corrects the pelvic obliquity but also prevents failure of distal fixation as it reduces the chances of loss of sagittal and coronal balance in the long term. Additionally, fixation to the pelvis in patients confined to the wheelchair is beneficial for maintaining the sitting balance during their life span. We feel that fixation to the pelvis is preferable in all patients with neuromuscular condition as this reduces the chances of loss of sagittal and coronal balance in the long term due to paralytic nature of the deformity.

42.5 Discussion

Guided growth construct is one among many surgical options for the management of EOS. This surgical technique is technically demanding and requires strict patient selection to ensure a predictable outcome. The use of sublaminar wiring can be time consuming and has possible risk in the hands of inexperienced surgeons. The risk of neurological complications has been well published in the literature [25–27], but in the hands of experienced surgeon, such complications are rare [28–30]. Passing the rods, engaging the fixed and gliding anchors through the muscle-sparing incision while achieving spinal correction, requires significant experience in deformity surgery. New gliding implants are starting to be available and may help to simplify the surgical technique and hopefully negate the need of sublaminar wires.

Patients with comorbid factors carrying additional risks associated with repetitive anesthesia are the ideal candidates for this technique. Patients with SMA and any other flaccid neuromuscular scoliosis are good candidates for this technique. Seeing that any attempt at prophylactic treatment with early bracing in these patients has not prevented curve development nor progression [7],

and that early spinal fusion impacts negatively on the development of the lungs and can cause death due to pulmonary failure [31, 32], this technique offers the best option to correct and control long c-shaped paralytic scoliosis during their growth and to an extended period.

Another favorable factor predicting good surgical outcome using this technique is the ability to translate the apex of the spinal deformity back to the midline and reestablishing the normal axis of spinal growth. The risks of add-on below the corrective growth-sparing implant are significant. Hence, having solid proximal and distal fixations is also very important. Even though we tend to try to keep our proximal and distal anchors to a minimum, we often regret not going just a bit longer to ensure no add-on occurs. If patient's morphology allows, the addition of cross-link is suggested across the fixed anchors, particularly if the pelvis is not incorporated into the distal anchor. In all patients with neuromuscular scoliosis, fixation to the pelvis is preferable as this reduces the chances of loss of sagittal and coronal balance in the long term due to the paralytic nature of their deformity. In such distal fixation, cross-links are not needed.

Passive-guided growth seems to be safe with a low complication rate. As predicted, there are fewer surgeries using this technique and fewer hardware failures. Despite no active distraction, all patients grow across the instrumented segments. We recommend that management of EOS, and particularly neuromuscular scoliosis, should be performed in a specialized center, where a high volume of procedures are carried out, in order to maintain safety and prevent significant complications. Having good medical support staff to deal with these high-risk patients is essential to achieve good results [33].

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Anterior Growth Modulation Techniques: Vertebral Body Stapling

43

Patrick J. Cahill, Justin Iorio, Amer F. Samdani, Joshua M. Pahys, and Randal R. Betz

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Key Points

- Vertebral body stapling (VBS) is a viable alternative to bracing for immature patients with moderate idiopathic scoliosis. Fusion can be avoided in >70 % of thoracic curves between 25 and 35° and in >80 % of lumbar curves between 25 and 45°.
- The technique for thoracic VBS is easily adopted by surgeons familiar with thoracoscopically assisted anterior spinal fusion surgery.
- Success of VBS can be predicted by the magnitude of the first standing radiograph after surgery; if the curve is <20°, success at avoiding surgery is very probable.
- Complications with VBS are rare and have little or no long-term sequelae. Complications encountered include implant failure, pneumothorax, and inability to control the scoliosis.

43.1 Introduction

Currently, the standard treatment for immature patients with moderate magnitude (20–45°) scoliosis is either observation or bracing with a thoracolumbosacral orthosis (TLSO). Despite the

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widespread use of bracing for controlling progression, results are variable [1–12] and are affected by compliance and poor self-image [13–15]. Curve progression in skeletally immature patients despite bracing is especially problematic for scoliosis with coronal deformity greater than 30° and often reaches a magnitude that requires fusion.

Compliance with recommended brace wear is an additional challenge; it is underreported by patients and their parents and is unpredictable, varying from 20 to 90 % [13, 16, 17]. Objective studies using a manometer or a temperature gauge find that compliance is, at best, around 60 % [18]. A study by Katz et al. [17] showed that 82 % of curves did not progress if the brace was worn for more than 12 h per day according to the compliance monitor. Bracing was initiated in these patients at or around peak height velocity (mean age greater than 12 years). Furthermore, the authors found that best results occurred when the brace was worn during daytime hours (while erect). Recently, the "Bracing in Adolescent Idiopathic Scoliosis Trial (BRAIST)" found that patients who complied with bracing for only 0–6 h per day had less favorable results (41 % success), which was similar to patients in the observation group (48 % success) [19]. In contrast, full-time bracing for an average of at least 12.9 h per day was associated with a success rate of 90–93 %. However, this wearing schedule can be difficult for many adolescents. Also, while brace treatment is noninvasive and preserves growth, motion, and function of the spine, it does not correct an established deformity. While most orthopedists, families, and patients agree that it is reasonable to wear a scoliosis brace for short periods if it means preventing an operation, a more difficult situation is encountered in the young child who faces the prospect of wearing a brace for more than 4 years with no guarantee of a favorable outcome. It is for these children that fusionless treatment options hold the most promise.

Vertebral body staples, which are made of a shape memory alloy, produce hemiepiphysiodesis at the curve convexity. The result is gradual curve correction via the Hueter-Volkmann principle as the spine grows. Advantages of vertebral body stapling (VBS) as an alternative to bracing include the elimination of compliance issues, direct spinal correction, consistent curve improvement, and prevention of future spinal fusion in properly selected patients. VBS also permits maintenance of spinal mobility and flexibility (Fig. 43.1).

Fig. 43.1 Demonstration of spinal mobility after T10–L3 VBS

43.1.1 Historical Overview

Stapling across physes of the long bones has been accepted as an effective method for treating limb malalignment in young children for over 50 years [20, 21]. Around the same time, the potential benefits were discovered for the spine. Animal studies using a rat tail model confirmed the ability to modulate vertebral growth plates with skeletal fixation devices [22]. In 1951, Nachlas and Borden [23] were initially optimistic about their ability to create and correct lumbar scoliosis in a canine model using a staple that spanned several vertebral levels. Many of the dogs exhibited some correction, and some of the animals exhibited arrest of their curve progression. Some of the staples failed because they spanned three

vertebrae. The enthusiasm for this new treatment was lost after the application of their stapling technique in three children with progressive scoliosis that yielded poor results. Other investigators have, similarly, been dissatisfied with convex stapling as a means of controlling progressive scoliosis.

Results for humans with congenital scoliosis were presented as early as 1954 [24], but the results were disappointing. The scoliosis correction was limited because the children had little remaining growth, and the curves were severe, with considerable rotational deformity. Some staples broke or loosened, possibly because of motion through the intervertebral disks. While the concept of stapling the anterior vertebral end plates/physes for growth modulation and curve stabilization seemed sound, the staples designed for epiphyseal stapling about the knee were prone to dislodging in the spine because they were not designed to function across the intervertebral disk and accommodate to the movement of the functional spinal unit.

In 2003, our institution published the results of a patient cohort that had undergone anterior VBS for moderate adolescent idiopathic scoliosis (AIS) with a newly designed staple [25]. The significance of the study was demonstrated by 87 % of the curves that were maintained using the stapling technique.

43.1.2 Basic Science Overview

Recent work has shown the efficacy of anterior growth modulation in animals. Despite the successful use of staples for epiphysiodesis of long bones in angular deformity, staples for growth modulation around the spine were not nearly as successful. The obvious issue was that staples designed for the long bones were prone to dislodge in the spine because they were not designed for movement which occurs in the spine.

Medtronic Sofamor Danek (Memphis, Tennessee) designed staples using nitinol, a shape memory alloy, which have 510(k) approval from the FDA specifically for fixation in the anterior spine within a single vertebral body or for fixation of hand and foot osteotomies. These staples are unique in that the prongs are straight when cooled but clamp down into the bone in a "C" shape when the staple returns to body temperature, thus providing secure fixation. The nitinol staple described in this text is considered "off-label" by the FDA. Nitinol is a biocompatible shape memory metal alloy composed of 50 % nickel and 50 % titanium. The temperature at which the staples will undergo the shape transformation can be controlled by the manufacturing process. Injury to surrounding tissues through the transformation temperature has not been seen in animal or human experience with cervical spinal fusions.

Nitinol has a very low corrosion rate and has been used in orthodontic appliances. Implant studies in animals have shown minimal elevations of nickel in the tissues in contact with the metal; the levels of titanium are comparable to the lowest levels found in tissues of titanium hip prostheses, and titanium is considered a biologically safe implant material. No method of sterilization used in operating rooms has been shown to have any effect on the metal's properties. Although sensitivity to nickel occurs in a very low percentage of the population, it is not anticipated to occur through the use of the nitinol staple. The crystal structure in nitinol is different than the small amount of nickel crystal structure in stainless steel such that the nickel does not leach out in nitinol compounds as it can on occasion with stainless steel. The nitinol staple has been tested in a goat scoliosis model applied across a disk space by Braun et al. [26] and has been shown to be safe and have utility for arresting iatrogenic curves of less than 70° in the goat.

43.1.3 Clinical Outcomes

In 2003, Betz and colleagues [25] reported on the use of the nitinol staples in 21 skeletally immature patients with AIS. Indications for the procedure were either brace noncompliance, often due to psychosocial reasons, or curve progression despite bracing. They found the procedure to be safe and effective, with the results comparable to

the expected results of bracing. In 2005, this same group [27] reported on 39 patients and their increased experience with the procedure. Stabilization of the curve was seen in 87 % of those patients older than 8 years at the time of stapling who had a curve of 50° or less with at least 1 year of follow-up. No curve less than 30° at the time of stapling progressed more than 10° at follow-up.

In 2010, Cuddihy et al. reported a retrospective study comparing VBS to bracing for patients with moderate idiopathic scoliosis using identical inclusion criteria [28]. In this comparison of two cohorts of patients with high-risk (Risser 0–1) moderate idiopathic scoliosis (measuring 25–44°), the results of treatment of smaller thoracic curves (25–34°) by VBS were statistically better than the results seen with bracing (82 % versus 54 %, respectively, $p=0.05$) when the cohorts were adjusted for mean age (10.5 years). For thoracic curves measuring 35–44°, the results were poor in both groups. The results of lumbar VBS and bracing were similar for curves measuring 25–44°. This study suggests that VBS can be used as an alternative or adjunct to bracing for patients with certain curve sizes who are noncompliant with bracing (Figs. 43.2 and 43.3).

Another study with 2-year outcome data consisted of 41 curves (26 thoracic, 15 lumbar) [29]. Thirteen patients had both curves stapled. The mean age was 9.4 years. Curves decreasing by greater than 10° were considered "improved." Curves within 10° of their preoperative measurement were considered "no change," and those progressing greater than 10° were considered "worse." Success was defined as "improved" or "no change." Thoracic curves measuring less than 35° had a 79 % success rate. Curves measuring less than 20° on first standing radiograph had an 86 % success rate. In patients with thoracic curves greater than 35°, 6 of 8 progressed past 50°. Seventy-one percent of patients with hypokyphosis showed improvement to a normal sagittal profile. One patient demonstrated worsening of kyphosis associated with coronal progression. Lumbar curves had an overall 87 % success rate, with only one patient with a preoperative curve of 40° progressing to 50°. Five patients lost greater than 10° of lordosis, but the final lumbar lordosis remained in the normal range. Complications were minimal, with a mean blood loss of 214 cc. A recent study by Auriemma et al. (unpublished data) reviewed the results of 63 patients who underwent VBS between the ages of 7 and 15 years for moderate idiopathic scoliosis (average Cobb, 30°). Of the 24 patients (36 curves) who reached skeletal maturity, defined as Risser grade 4 or 5, 71 % (12/17) of thoracic and 89 % (17/19) of lumbar curves were treated "successfully," defined as either greater than 10° improvement in Cobb angle or within 10° of preoperative curve magnitude.

VBS has shown superior outcomes when curves correct to less than 20° on first standing radiographs [29, 30]. Subanalyses of lumbar curves from these studies revealed higher success rates in curves that corrected to less than 20° on first standing radiograph (88.9 %) than those that did not (83.3 %) [29, 30]. As a result of these findings, the treatment algorithm for VBS has been transitioning from stabilization of preoperative curves to maintenance of intraoperative correction by the addition of postoperative nighttime bracing. Recently, a cohort of AIS patients with moderate (20–45°) thoracolumbar/ lumbar (TL/L) curves underwent VBS and adjuvant postoperative bracing (unpublished data). TL/L Cobb angle significantly improved from a mean of 34° preoperatively to 21° at a minimum of 2-year follow-up. Lateral trunk shift also improved from a mean of 1.8 cm preoperatively to 0.5 cm at most recent follow-up. Although health-related quality of life outcomes have not correlated with trunk shift [31], trunk imbalance has been shown to negatively impact self-image and is a major clinical concern of patients.

Based on this review, we have altered our strategy for when to use staples alone and when to use additional strategies, as follows: if the thoracic curve measures 35–45° and does not bend below 20°, then we will offer vertebral body tethering or a posterior hybrid distraction implant, a unilateral VEPTR, or a growing rod in addition to stapling (Fig. 43.4). If on the first standing radiograph the curve does not measure below 20°, we will brace the child until the curve measures less than 20°.

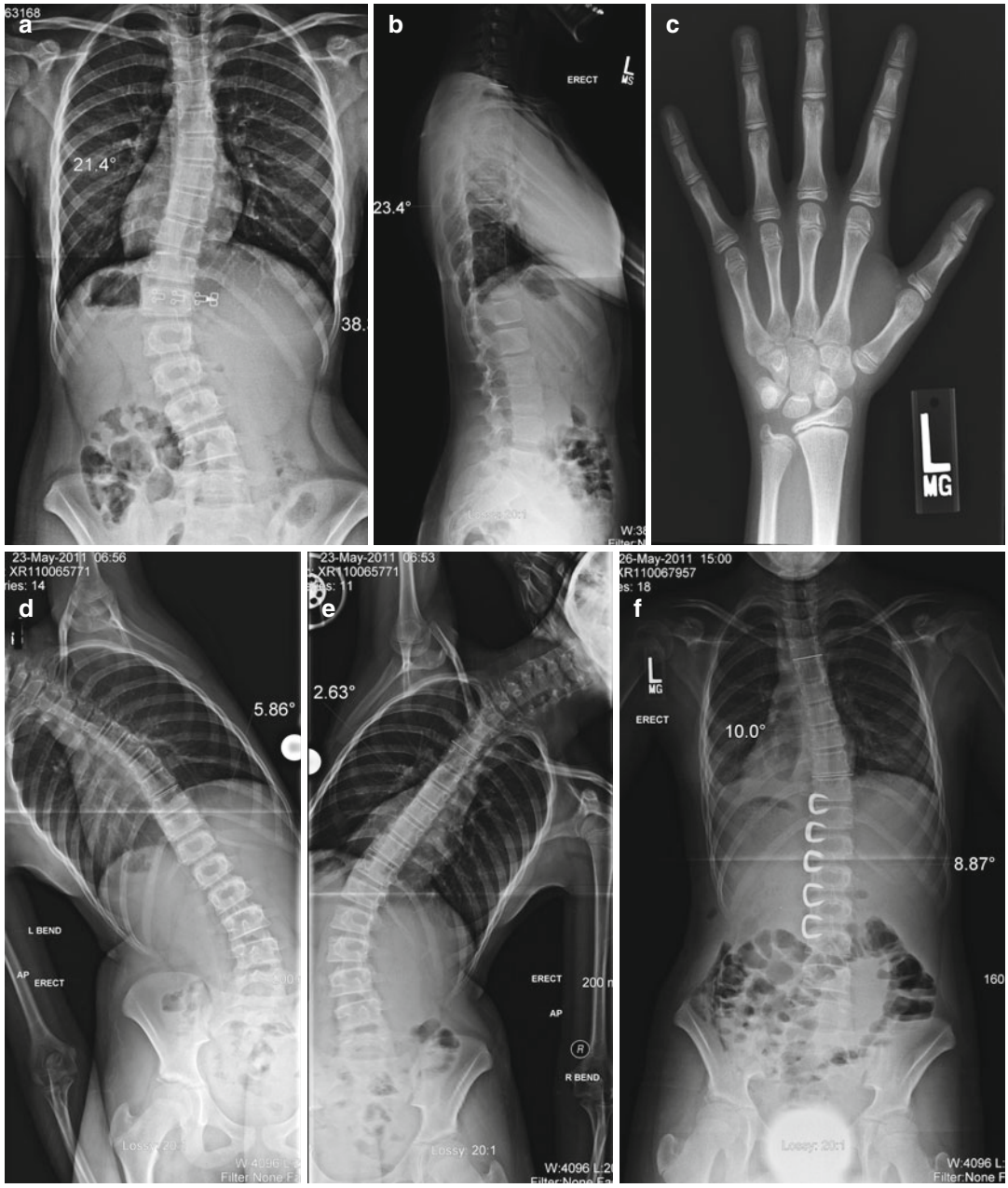

Fig. 43.2 PA (**a**) and lateral standing radiographs (**b**) of a 12-year-old female demonstrating a 21° thoracic curve and a 38° thoracolumbar curve. (**c**) Bone age shows the patient to be Sanders 3. Preoperative bending radiographs (**d**, **e**) demonstrate the flexibility of the curve. Patient underwent right-sided thoracoscopic VBS from T10 to L3. Her first standing radiographs (**f**, **g**) demonstrated thoracic curve correction to 10° and lumbar curve correction to 9°. Latest follow-up (**h**, **i**) at 3 years post-op demonstrates a thoracic curve of 14°, lumbar curve of 18° degrees, and improvement of thoracic kyphosis to 27°

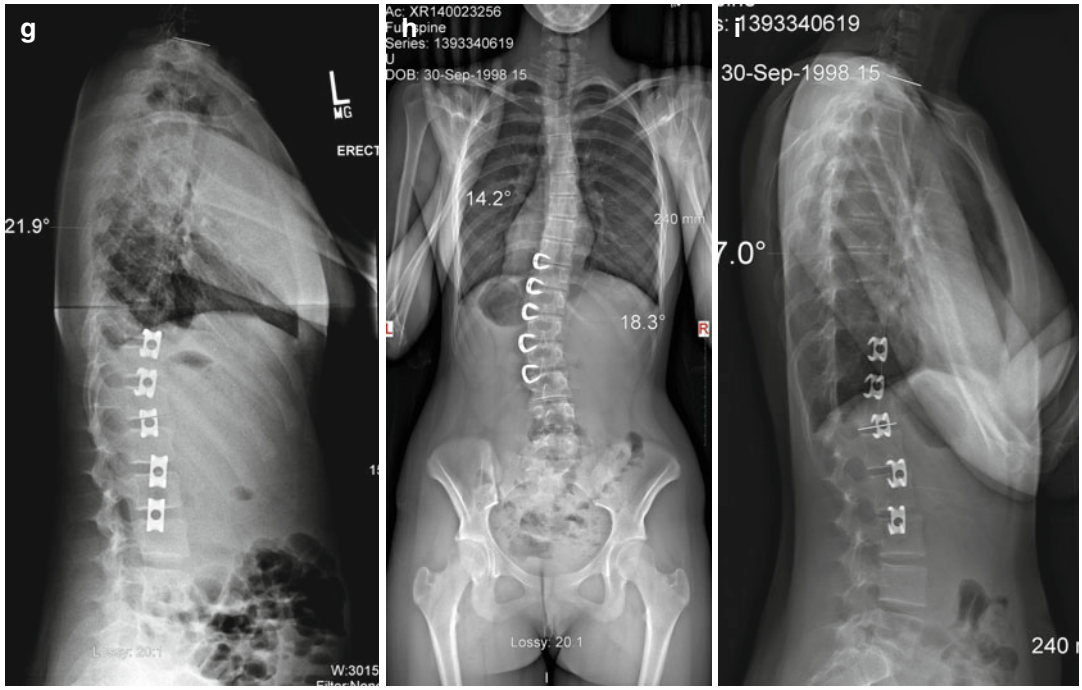

Fig. 43.2 (continued)

43.2 Clinical and Technical Overview

43.2.1 Indications and Contraindications

Patients who have at least 1 year of growth remaining, a scoliosis deformity for which brace treatment would be considered, or who may have failed or refused bracing, are good candidates for the stapling procedure. Lenke 1, 3, 5, and 6 scoliosis curves are ideal for treatment with vertebral stapling. Other indications are as follows: age less than 13 years for girls and less than 15 for boys; Risser 0 or 1, at least 1 year of growth remaining by wrist x-ray, or Sanders digital stage less than or equal to 4; thoracic curves 25–35°, and lumbar coronal curves less than or equal to 45°, with minimal rotation and flexible to less than or equal to 20°; and sagittal thoracic curve less than or equal to 40°.

Medical contraindications are the same as for any anterior spine or chest procedure and include systemic infection, active respiratory disease such as uncontrolled asthma, or conditions with increased anesthetic risk. Significantly compromised pulmonary function may be a relative contraindication.

We do not perform vertebral stapling for lumbar curves over 45° or for thoracic curves over 35° that do not bend to less than 20° because our early experience has yielded poor results. For these larger curves we now perform vertebral body tethering as described recently by Samdani et al. [32]. Over 100 vertebral body tethering procedures have been performed at our institution with 12 for primary lumbar curves. Initial results demonstrate 50 % initial correction with gradual improvement as the child grows. Also, if the curve on the first erect film does not measure less than 20°, the patient should wear a corrective nighttime brace until it does. Kyphosis greater than 40° is also a relative contraindication because of the potential for the creation of hyperkyphosis with growth. We occasionally do a lateral Stagnara view to confirm true kyphosis if there is a question.

Surgeons with experience in anterior spine surgery and especially minimally invasive techniques should be able to perform this procedure. It may be helpful to enlist the assistance of an experienced general or thoracic surgeon. With the use of thoracoscopic and minimally invasive techniques for lumbar curves, scoliotic vertebrae from T3 to L4 can be stapled while limiting the total scar length. Placement of instrumentation at other levels will depend on anatomic variances in the location

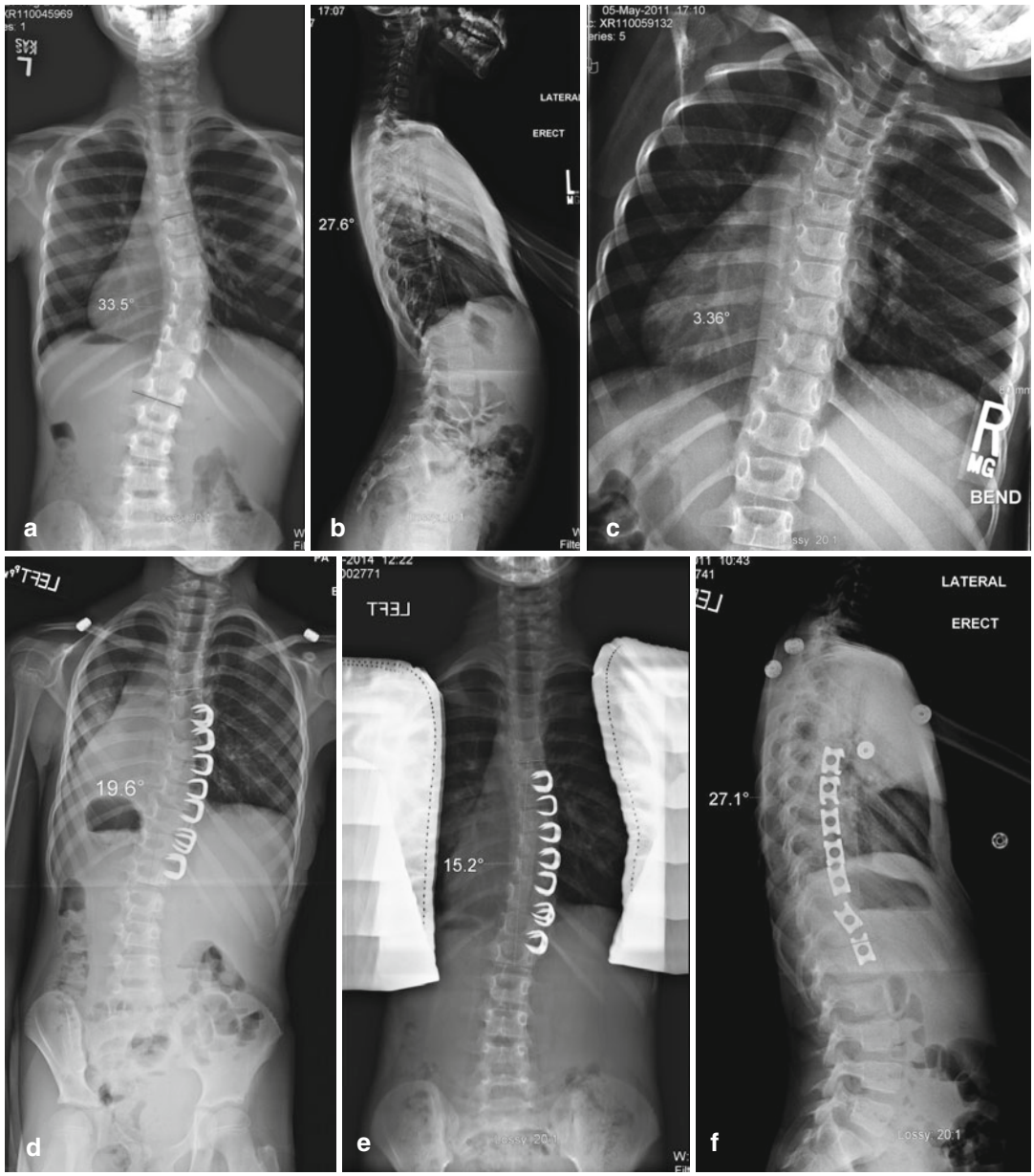

Fig. 43.3 AP (**a**) and lateral standing radiographs (**b**) of an 8-year-old female demonstrating a 34° thoracic curve. Preoperative standing radiograph (**c**) demonstrates the flexibility of the curve. Patient underwent right-sided thoracoscopic VBS from T6 to L1. Her first standing radiographs (**d**, **e**) demonstrated thoracic curve correction to 20°. Latest follow-up (**f**, **g**) at 2.5 years post-op demonstrates a thoracic curve of 15° and improvement of thoracic kyphosis to 36°

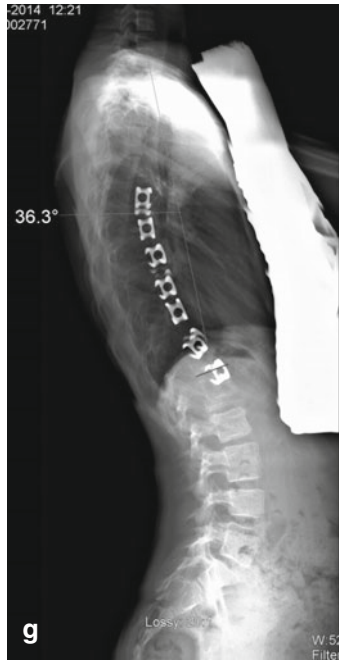

Fig. 43.3 (continued)

of the subclavian, azygous, or iliac vessels and the size of the psoas muscle. As a general rule, we try to avoid stapling the L3–L4 disk because of the risk to the nerve roots if a transpsoas approach is used or if retracting the psoas to get the staple posterior to midline of the body requires significant psoas mobilization and vessel ligation.

43.2.2 Technical Overview

43.2.2.1 Equipment/Instrumentation

- Bipolar cautery, monopolar cautery.
- Nitinol staples: staples are straight when cooled and clamp down into the bone in a C shape when achieving body temperature.
- Thoracoscope.
- Basin with sterile ice water.
- Fluoroscopy.

43.2.2.2 Anesthesia and Positioning

- General anesthesia and intubation with a double-lumen endotracheal tube for thoracic curves
- Lateral decubitus position with convex side of the scoliosis in the "up" position
- Soft pads under all pressure points
- C-arm under table for PA and lateral imaging.

All vertebral bodies included in the Cobb angle of the curve are instrumented. Under single-lung general anesthesia, patients are placed in the lateral decubitus position with the convex side of the scoliosis curve in the "up" position. The table is not flexed, and only a small axillary roll is placed. Patient positioning is critical and can be used to maximize correction. The axillary role is often positioned at the apex of the proximal thoracic curve, several centimeters lower than in the standard lateral decubitus position, in order to allow the main thoracic curve to partially correct.

This procedure lends itself to the use of minimally invasive surgical techniques. If video-assisted thoracoscopy is being utilized for insertion, then one-lung ventilation will be necessary, unless carbon dioxide (CO_2) gas insufflation is available to displace the lung for visualization of the spine and surrounding structures. Using fluoroscopy, a lateral image of the patient can be utilized to confirm the levels of the vertebra and also to center the ports over the midportion of the vertebral bodies (Fig. 43.5). The standing position is based on the surgeon's preference, but in our practice the surgeon usually stands in front of the patient with the access surgeon or the assistant holding a camera next to the surgeon. A second assistant stands on the opposite side to help with the retraction (Fig. 43.6).

In the thoracic spine, most incisions will be close to or within the area of the posterior axillary line. The first port is made in the fifth to seventh intercostal space along the anterior axillary line for visualization with the scope. Additional ports, generally 2 or 3, are made in the posterior axillary line for insertion of the staples. Two oblique incisions are usually required for placement of six or more staples (Fig. 43.7). The incisions are about 2.5–3 cm long and are oblique in such a way as to follow the slope of the ribs. Each incision can then be used to make two to three internal intercostal ports. This allows several levels to be stapled through each skin

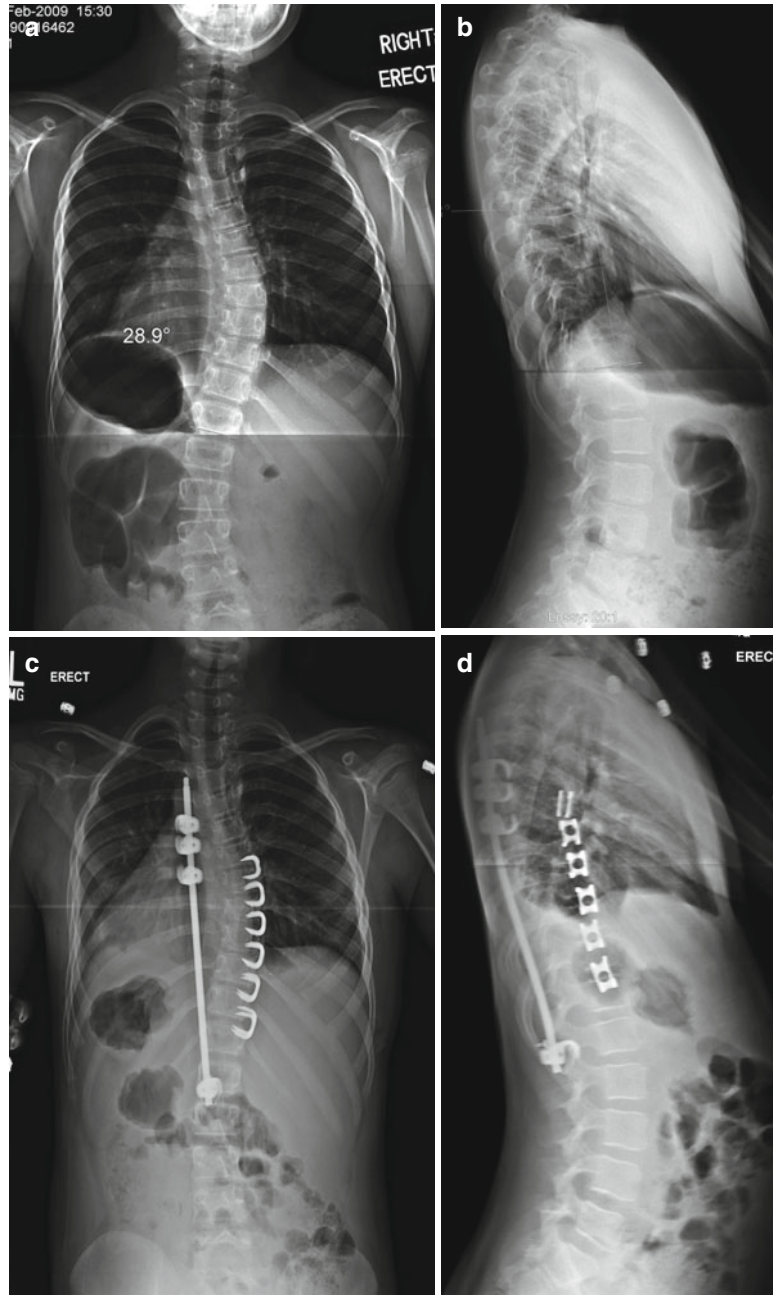

Fig. 43.4 PA (**a**) and lateral standing (**b**) radiographs of a 13-year-old female who underwent T7-T11 VBS and hybrid rod placement (**c**, **d**)

incision and accommodates the size of the instruments and implants. Fluoroscopy or direct visualization with the thoracoscope are both reliable methods for planning the incision.

Staples that cross the thoracolumbar junction require partial reflection of the diaphragm anteriorly. Lumbar disk spaces can usually be exposed with a retroperitoneal mini-open approach through a single incision. The incision length is 2.5–3 cm and, similar to the thoracic spine, is localized based on the image intensifier. During the approach, the psoas is either retracted posteriorly or carefully separated longitudinally directly over the posterior half of the disk under EMG control [33]. We generally place the staples posterior to the midline of the lumbar vertebral

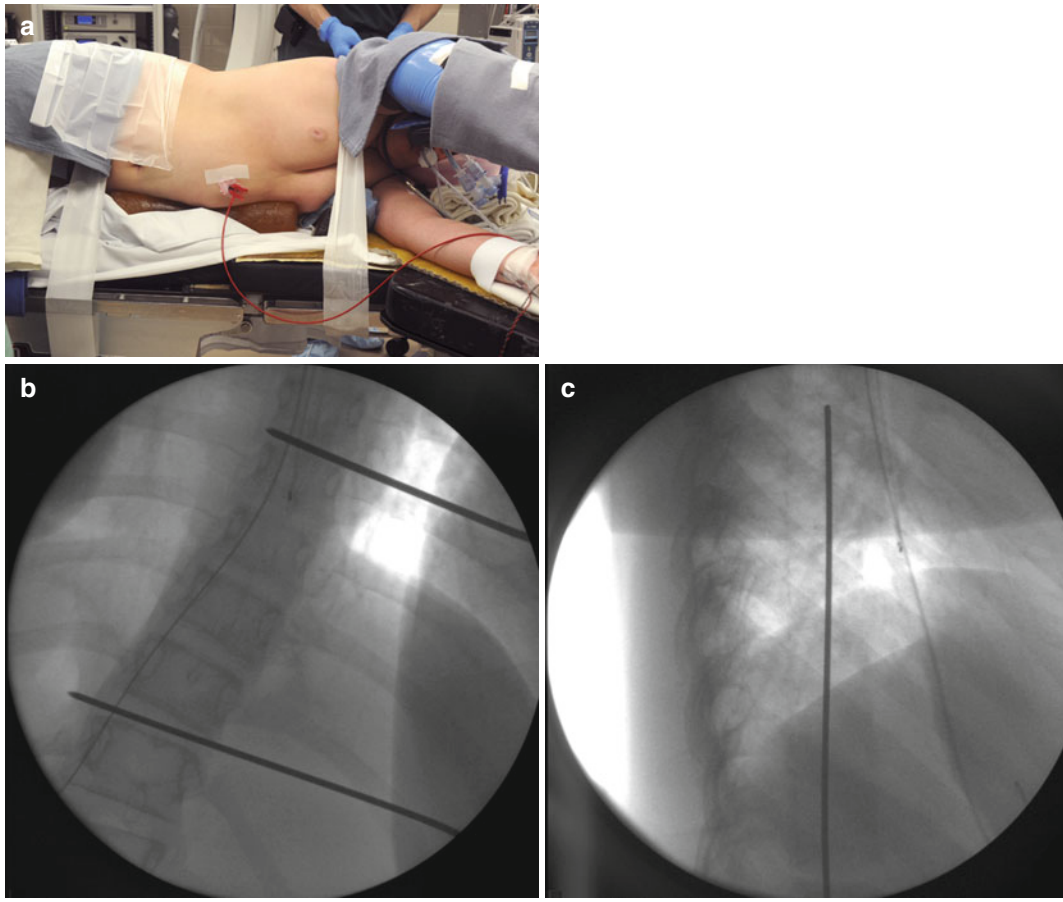

Fig. 43.5 (**a**) The patient is placed in a lateral decubitus position. (**b**, **c**) Fluoroscopy is used to confirm the levels of the vertebrae and to center the ports over the midportion of the vertebral bodies

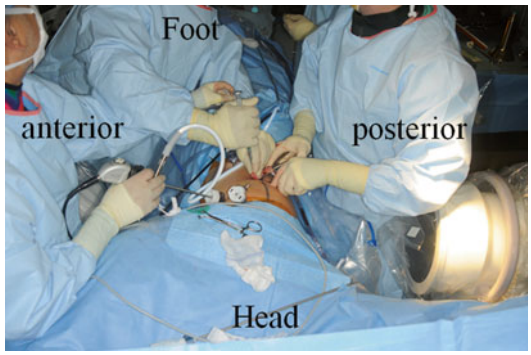

Fig. 43.6 Surgeon positioning during VBS. The surgeon usually stands anterior to the patient with the access surgeon or the assistant holding a camera next to the surgeon. A second assistant stands on the opposite side to help with the retraction

bodies, ligating or mobilizing the segmental vessels and retracting the psoas. A posterior staple will theoretically avoid diminishing lordosis of the lumbar spine. Position of the staples is reconfirmed using fluoroscopy at the end of the procedure.

While the patient is in the lateral decubitus position, often the flexible main thoracic curve reduces. To further reduce the curve while placing the staples, lateral pressure can be applied through an inserter affixed to staples previously placed in another level. This trial inserter can be used to push the spine straight, thus maximizing correction on the operating table. This may be important because patients who have less than or equal to 20° of curvature on the first erect

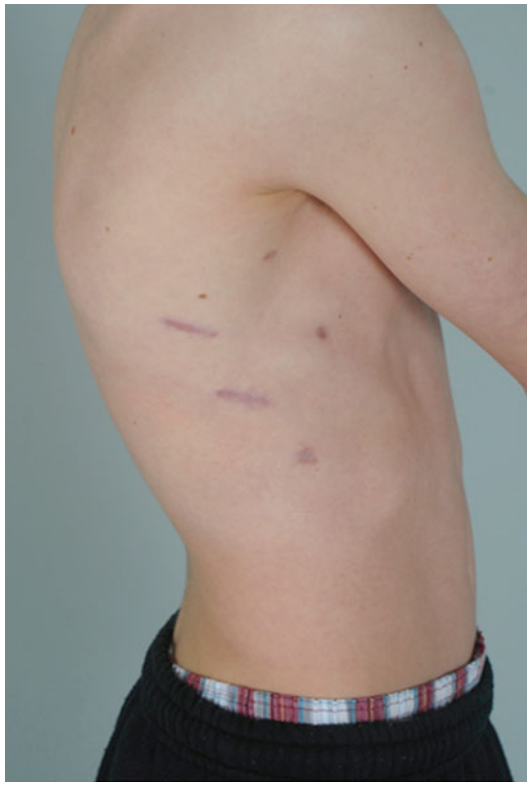

Fig. 43.7 Generally, 2 but up to 4 ports in the posterolateral line are used, with the thoracoscope being inserted in the anterior axillary line at the apex of the curve. Incisions are oblique and follow the slope of the ribs

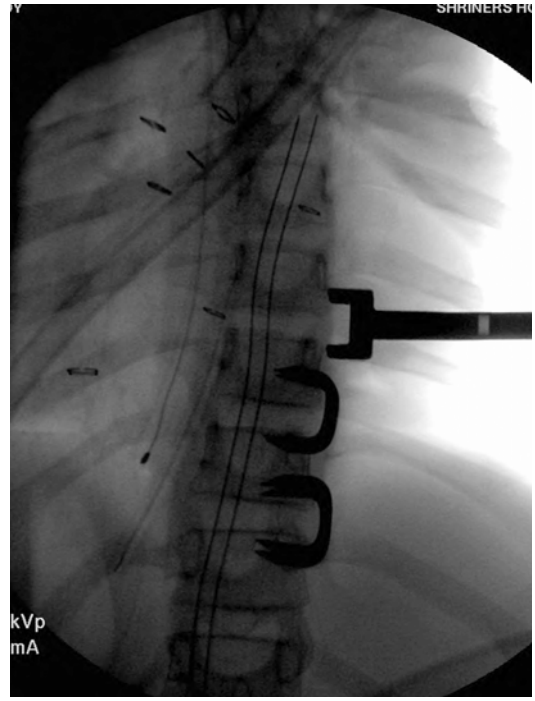

Fig. 43.8 Four-prong trial. The staple trial is passed through one of the posterolateral ports and centered over the intervening disk space for staple sizing. The surgeon should place the prongs as close to the end plates as possible

radiographs typically have better maintenance of correction.

The insertion device which holds the single two-prong staples is 10×14 mm wide. The staples come in many sizes, with the 12 mm four-prong double staple being the widest, longest object (at 14×12 mm) that has to pass between the ribs. They allow for maintenance of the intercostal port space, quick removal of the staple trial, and placement of the appropriate-sized staple while protecting the muscle and pleura from repeated trauma. Small pediatric Finochietto retractors or nasal speculum distractors can also be used to enlarge the intercostal ports and may be used in place of collapsible or rigid ports. These are ideal in the setting of CO_2 insufflation, since they are collapsible and better preserve intrathoracic pressure to maintain a collapsed lung.

A radiopaque trial inserter is used to obtain the dimension of the staple (3–8 mm) and to create pilot holes. Using fluoroscopy, the appropriate size trial is selected to span the distance across the disk, apophyses, and physes (Fig. 43.8). A thoracoscopic view of the trial inserter bridging a thoracic disk space is shown in Fig. 43.9a, and a fluoroscopic view of the trial inserter bridging a lumbar disk space is illustrated in Fig. 43.9b. Once the correct size for the trial is determined, it is tapped into place where the staple will be located. Two single staples (two prongs) or one double staple (four prongs) is placed at each level except when an additional two-prong staple is inserted anterior to these to induce further kyphosis. In very small children, the most proximal vertebra is often small, and only one single staple can be placed safely. The tines of the trial are used to create the pilot holes for the staple tines. If the tines of the trial come close to the segmental

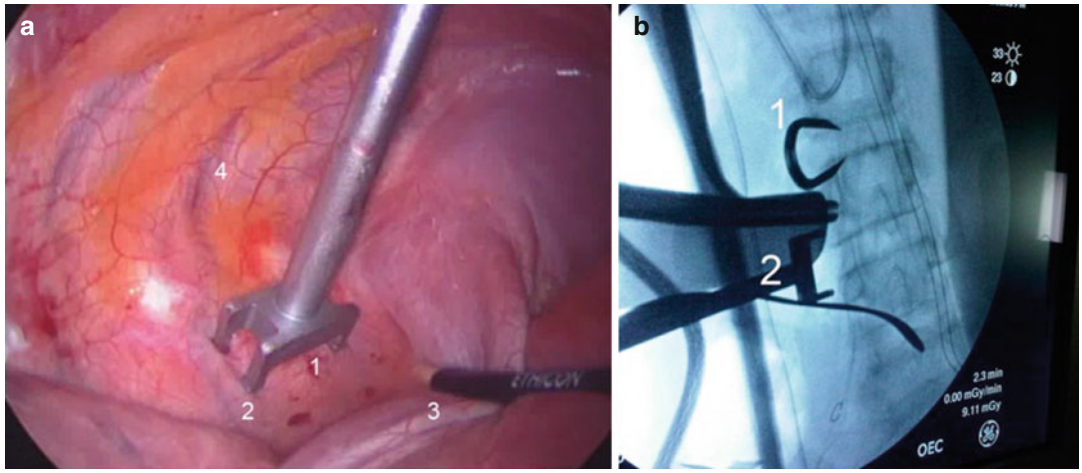

Fig. 43.9 Creating pilot holes with the trial inserter. (**a**) Thoracoscopic view. *1* disk space, *2* segmental vessels, *3* retracted lung, *4* intercostal vessels. (**b**) Fluoroscopic view of the same surgical step in the lumbar spine. *1* staple in the disk space above, *2* trial inserter

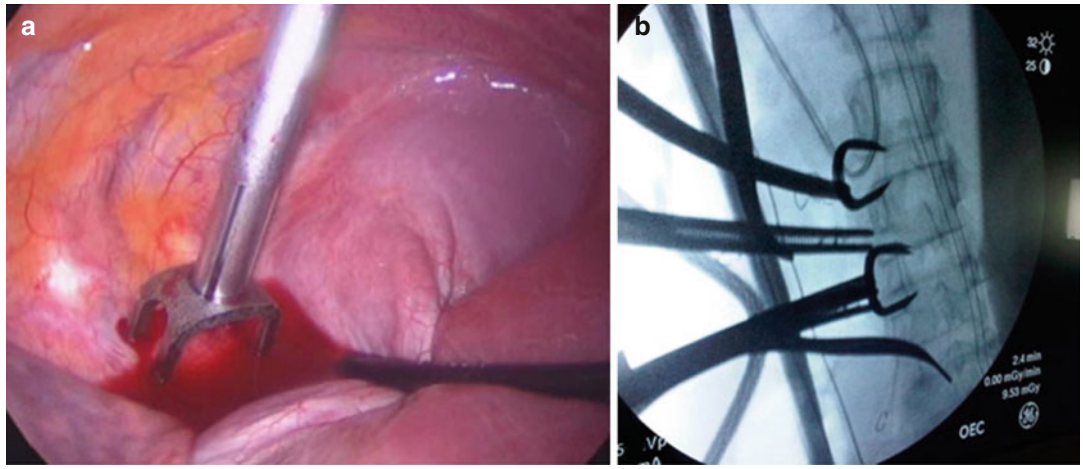

Fig. 43.10 Insertion of the staple in previously created pilot holes. (**a**) Thoracoscopic view prior to final insertion into the previously created pilot holes with the retracted lung on the right and the internal view of the rib cage on the left. (**b**) Fluoroscopic confirmation after the insertion of the staple bridging a lumbar disk space

vessels, then the pleura is incised, and the vessels are retracted gently, while the pilot holes are created and until the staple is seated in place. The authors prefer the use of a harmonic scalpel for dividing the pleura for this purpose. The smallest staple that spans the disk and growth plate is used.

The pilot holes will act as a guide for the staple tines to ensure correct placement. The trial is removed, and the staple, which has been cooled over a basin of ice, is placed over the pilot holes and quickly inserted (Fig. 43.10). The decision to insert a two- or four-prong staple is based on the width of the vertebral body as seen in the operating room. The four-prong staples provide the desired amount of compression with less time required for insertion and fewer instrument passes into the chest. Once the staple is in the desired position, the staple inserter is removed, and if the staple is not flush against the bone, an impactor is used to drive the staple deeper. This must be done quickly before the tines are fully

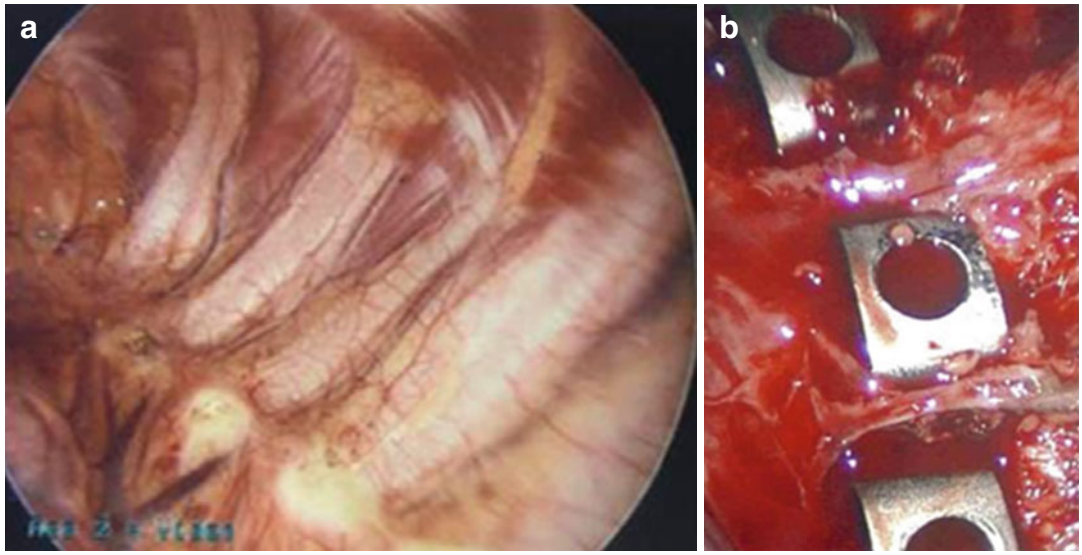

Fig. 43.11 (**a**) Thoracoscopic image demonstrating rib heads and adjacent ribs. (**b**) Proper staple placement is anterior to the rib heads

deployed. The dull staple trial or the inserter placed into a previously implanted staple (preferred) can be used to help push at the apex of the convexity to further reduce the curve while the staple is being inserted. This step of correcting the spine through translation is of critical importance to obtain correction on the OR table. If the patient has two curves greater than 25°, both curves should be stapled, which requires intraoperative repositioning.

The nitinol staple's sharp, curved prong design and shape-changing abilities allow for insertion parallel to the cartilaginous vertebral apophyses to provide end plate compression. The staple tines are sharp and are designed to pass easily through the bone. The staple's prongs are straightened manually and are then cooled by immersion in a sterile ice bath. The scrub nurse or technician can perform this ahead of time; it is important to have the staples on ice for a minimum of 45 min. The staples must pass quickly from the sterile ice water bath to the vertebral bodies to prevent staple warming and tine deployment. The tines will remain straight until the staple begins to return to normal body temperature at which point they deploy to their original curved shape. Complete tine transformation may take a minute. If the staple is completely seated within the vertebral bodies when the prongs deploy, then the staple position is secure. On a rare occurrence, if the staple does not deploy into its C shape, it should be replaced.

Staples are placed anterior to the rib heads (Fig. 43.11), and if the patient has severe hypokyphosis or thoracic lordosis, the staples can be placed more anterior on the vertebrae to help produce kyphosis with the patient's growth. If possible, a double and a single staple are placed across the two apical disks such that anterior growth can be further modulated to reduce the hypokyphosis. In the lumbar spine, the staples should be placed as far posteriorly on the vertebral body as possible, at least in the posterior half of the body, to maintain a normal lordosis.

If the staples are being placed thoracoscopically, the addition of CO_2 allows for collapse of the lung without single-lung ventilation. Specialized equipment and ports are needed for this technique. Low-pressure CO_2 also promotes hemostasis in the bleeding bone, but after it is discontinued, brisk bleeding is possible. Gas pressures should be kept low to prevent a lateral shift of the mediastinum, which may cause a drop in blood pressure. Vaseline gauze can be helpful to place over ports to reduce the leakage of CO_2.

A chest tube is placed to prevent pneumothorax and for drainage of any effusion.

43.2.3 Postoperative Care

All patients receive intercostal blocks, opioid analgesia, and ketorolac for pain relief. The chest tube is removed when less than 100 mL output in 24 h is achieved, which is usually on the first postoperative day. No brace is required for thoracic curves. A non-correcting soft corset can be prescribed for lumbar curves for 4 weeks to help with stabilization of the staples by decreasing the patient's motion. Standing posteroanterior radiographs are obtained from all patients prior to discharge. Postoperative nighttime bracing is implemented as soon as tolerated after surgery for (1) curves which measure greater than 20° on the first erect radiographs and (2) patients with thoracolumbar/lumbar curves who are aiming for complete resolution of all clinical deformity. Patients are asked to restrict activities for 4–6 weeks to allow for skin and muscles incised during the surgery to heal. Patients return at 3–6 weeks postoperatively for a wound check and radiographs to assess for implant stability. At 6 weeks, any remaining restrictions are lifted, and the patients are seen for routine follow-up every 6 months until maturity.

43.2.4 Complications

In our experience, there has been one documented major complication: a rupture of a preexisting, unrecognized congenital diaphragmatic hernia in a 4-year-old that ruptured at 6 weeks post-surgery and required emergency repair. Other, lesser complications have included 1 case each of bleeding from a nicked segmental vessel and conversion of the thoracoscopic ports to a mini thoracotomy, a chylothorax, development of mild pancreatitis, clinically significant atelectasis, and transient sympathectomy syndrome. Some patients early in the series had prolonged chest tube drainage beyond four days. However, in a vast majority of cases, we maintain chest tubes for less than 24 h. There have been no instances of damage to the great vessels, lung parenchyma, heart, abdominal organs, or kidneys. There have been no documented neuromonitoring changes.

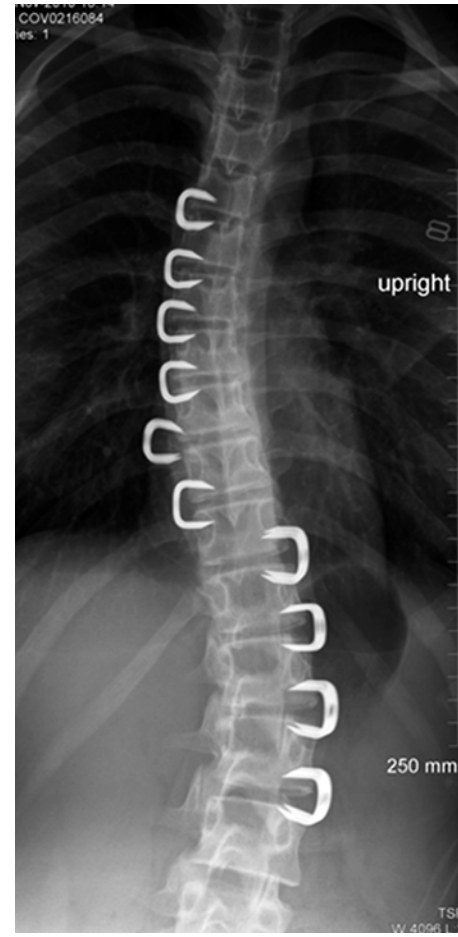

Fig. 43.12 AP standing radiograph of a patient who is 2.5 years status post VBS with staple loosening at T9–T10

Five staples (one in each of five patients) have shown evidence of movement, back out, or loosening. In four of the five patients, these changes were seen within 2 months of the initial procedure, and in one patient a loose staple was identified at 2.5 years postoperatively (Fig. 43.12). Loosening was asymptomatic in 3 of the 5 patients, and two of these patients underwent revision to remove and replace the loose staple. In one patient who experienced pain with a loose staple at 2.5 years, the pain was relieved after staple removal. Four broken staples (one in each of four patients) have been identified, all in the lumbar spine and all presenting from 6 to 12 months after the initial procedure. Two of the four patients experienced pain, and one of the two underwent staple removal with

Table 43.1 Results VBS versus bracing

	No change/improvement (%)	Progression (%)	*P* value (Fisher's exact test)
Thoracic curves 25–34°			
VBS (*N*=25)	80	20	0.2
Bracing (*N*=66)	63	36	
Thoracic curves 35–44°			
VBS (*N*=11)	18	82	0.08
Bracing (*N*=41)	51	49	
Lumbar curves 25–34°			
VBS (*N*=13)	77	23	0.5
Bracing (*N*=43)	63	37	
Lumbar curves 35–44°			
VBS (*N*=6)	67	33	1.0
Bracing (*N*=15)	60	40	

Based on data from Ref. [28]

subsequent pain relief. The curves have remained stable in these patients with further follow-up.

Thus far, four patients have experienced overcorrection of a stapled curve (two thoracic curves and two lumbar curves). Three of the four patients underwent staple removal between 1 and 4 years from the initial stapling procedure. We recommend staple removal if greater than 10° of overcorrection occurs. We also try to delay stapling until age 8 to minimize the risk of overcorrection.

Pain has been reported by one child, whose preoperative thoracic and lumbar curves measured greater than 50° when stapled. Her thoracic curve progressed and she required fusion. The stapled lumbar curve actually corrected with the thoracic fusion. Two distal lumbar segments that were stapled did not require fusion. Two months after fusion, she had pain in the lumbar spine, and a bone scan showed increased uptake at the staple-bone interface. Three weeks after removal of the two distal staples, the patient had no pain.

43.2.5 Current Outcomes

Our group presented the results of VBS versus bracing at the 2010 SRS annual meeting [28]. Inclusion criteria were (1) diagnosis of idiopathic scoliosis, (2) age at least 8 years at time of first visit, (3) curve magnitude of 25–45° at first visit, (4) Risser sign of 0 or 1 at first visit, and (5) minimum 2-year follow-up. Of the 49 patients with VBS identified, 43 (88 %) patients with 56 curves (13 patients had both thoracic and lumbar curves) were available and specifically reviewed for this paper. The mean age at the time of surgery was 10.5 years. The mean preoperative curve size was 32° (32° thoracic and 31° lumbar). The mean follow-up was 40.8 months.

The bracing cohort was comprised of a consecutive series of patients derived from the Göteborg bracing database who were treated between 1968 and 1994, meeting identical inclusion criteria as the VBS group. The Göteborg scoliosis database contains information about all patients with scoliosis (*n*=2655). From this bracing database, 165 curves in 129 patients (with 36 patients having both thoracic and lumbar curves) were identified who met our inclusion criteria.

The mean age at the time of treatment was 12.1 years. The mean preoperative curve was 32° (33° thoracic and 31° lumbar). The mean follow-up was 43.1 months.

The results were similar between stapling and bracing in all subgroups. For thoracic curves measuring 25–34°, VBS had a comparable success rate of 80 % versus 63 % for bracing. Thoracic curves measuring 35–44° showed bracing and VBS to have a poor success rate (18 % and 51 %, respectively). For lumbar curves measuring 25–34°, VBS had a 77 % success rate versus only 63 % for bracing, and for lumbar curves measuring 35–44°, VBS had a 67 % success rate versus 60 % for bracing (Table 43.1).

Table 43.2 Subanalysis of groups VBS versus bracing when matched for age

	No Change/improvement (%)	Progression (%)	*P* value (Fisher's exact test)
Thoracic curves 25–34°			
VBS (*N*=25)	80	20	0.09
Bracing (*N*=36)	58	42	
Thoracic curves 35–44°			
VBS (*N*=11)	18	82	0.21
Bracing (*N*=13)	46	54	
Lumbar curves 25–34°			
VBS (*N*=13)	77	23	0.27
Bracing (*N*=18)	56	44	
Lumbar curves 35–44°			
VBS (N=6)	67	33	0.16
Bracing (N=3)	0	100	

Based on data from Ref. [28]

Since the two groups were not comparable for mean age, a subanalysis was performed by changing the inclusion criteria for the bracing group by establishing an upper age limit of 12.5 years. This particular upper age limit was established so that the mean age at the start of treatment was not statistically different between the bracing group (average age = 11.1) and stapling group (mean age, 10.5 years), and the results were reanalyzed (Table 43.2). The results did not significantly change except in the subtype "lumber curves 35–44°" where the "success" rate in the bracing group was decreased from 60 to 0 % ($P=0.2$). However, the number of braced patients in the group was decreased from 15 to 3.

In a separate study, 63 patients (81 curves) who were at high risk of curve progression underwent VBS for idiopathic thoracic and/or lumbar scoliosis (Auriemma et al, unpublished data). Inclusion criteria were (1) idiopathic scoliosis, (2) age 7–15 years at time of surgery, (3) preoperative coronal curve magnitude of 20–35° for thoracic curves and 20–45° for lumbar curves, and (4) preoperative Risser grade 0 or 1. The average preoperative thoracic and lumbar Cobb angles measured 29.5° and 31.1°, respectively, and patients were followed for an average of 3.6 years (range, 2–8). Twenty-four patients (38 %) reached skeletal maturity by the most recent follow-up. "Success" was defined as either greater than 10° improvement or no change (+10 to −10°) in Cobb angle and "failure" as greater than 10° progression. Thoracic VBS had an overall success rate of 74 % (32 successes, 11 failures), and lumbar curves had an 82 % success rate (31 successes, 7 failures). An important finding was the high success rate at skeletal maturity: 71 % (12/17) of thoracic curves and 89 % (17/19) of lumbar curves. The successful results of VBS are in stark contrast to the findings of Dimeglio et al. [34] who reported that 75 % of idiopathic curves between 21 and 30° at the onset of puberty eventually required spinal fusion, as did 100 % of curves greater than 30° at the onset of puberty.

While a relatively successful treatment option, the curves of some patients who underwent VBS have progressed from moderate to a magnitude that classifies them as major (greater than 50°), requiring a spinal fusion. Many of these were patients in whom initial thoracic curves were greater than 35° who we would not staple today. In this small series of patients ($n=28$), posterior spinal fusion has been safely and effectively performed. Curve correction, surgical time, and blood loss have been comparable to our primary spinal fusion procedures. Staple removal is not required for the safe placement of posterior pedicle screw placement and subsequent curve correction. Thus, VBS does not preclude a patient from undergoing a future spinal fusion if necessary (Figs 43.13 and 43.14).

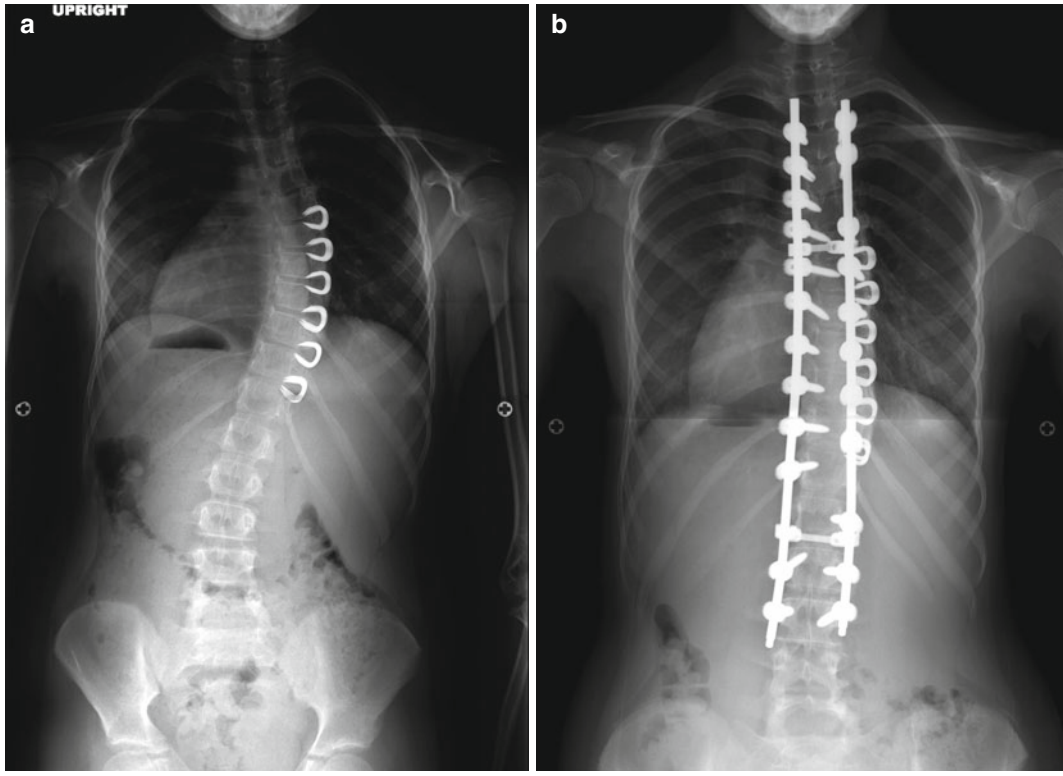

Fig. 43.13 Conversion of progressive deformity after VBS (**a**) to posterior spinal fusion (**b**)

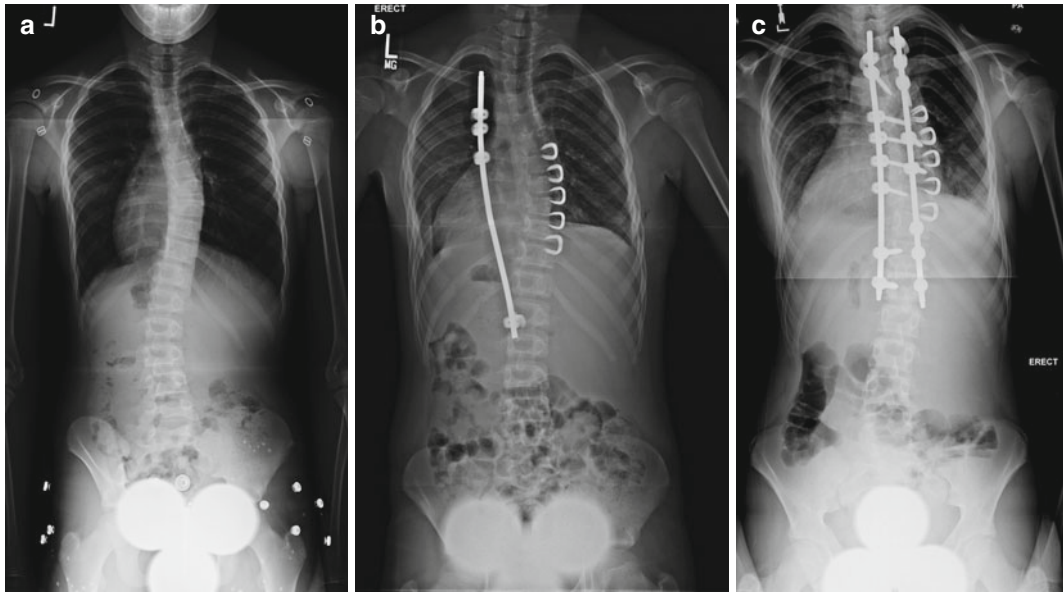

Fig. 43.14 (**a**–**c**) Pre- and post-conversion images of a patient who had progressive scoliosis despite posterior distraction rod treatment and who was ultimately converted to posterior spinal fusion

43.2.6 Summary

The recent investigations of convex anterior vertebral body stapling, both in animal models and in juvenile and adolescent scoliosis, offer solid early results with the use of improved implants and techniques. The use of a shape memory alloy staple tailored to the size of the vertebral body, the application of several staples per level, the instrumentation of the Cobb levels of all curves, and the employment of minimally invasive thoracoscopic and lumbar approaches all offer substantial improvements over previous fusionless techniques. Patient selection may also play a role in the current success of these fusionless treatments, with perhaps the ideal candidates for this intervention possessing smaller and more flexible curves.

Our indications currently are age less than 13 years for girls and less than 15 for boys; at least 1 year of growth remaining by wrist radiographs or Sanders digital stage less than or equal to 4 [35]; thoracic curves 25–25° and/or lumbar curves less than 45°, with minimal rotation and flexible on side bending to less than 20°; and a sagittal thoracic curve less than 40°. Following the VBS procedure, if on the first standing radiograph, the curve does not measure below 20°, we brace the child with a nighttime orthosis (preferably the Providence nighttime brace) until the curve measures less than 20°.

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Growth Modulation Techniques: Tethering

44

Peter O. Newton, Vidyadhar V. Upasani, and Christine L. Farnsworth

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Key Points

- Modulation of spinal growth using an anterolateral tether has the potential to become the standard in the treatment of scoliosis in children and adolescents.
- Anterolateral flexible tethering provides a technique to allow patients with spinal deformity and remaining growth to "grow out" of their deformity by redirecting the power of spinal growth.
- Anterolateral tethering allows motion through intervertebral disks during growth modulation treatment; thus, it may provide disk health for the long term.

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44.1 Introduction

Surgical spine fusion and instrumentation remains the definitive treatment for children with severe spinal deformities. Although midterm follow-up studies with modern instrumentation systems and techniques have demonstrated satisfactory

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outcomes in terms of deformity correction, maintenance of correction, and patient satisfaction, it is well accepted that spinal fusions sacrifice spinal flexibility, alter stresses on adjacent unfused segments, and may lead to problems with spinal imbalance in the long term. [1–6] Concerns with outcomes in the long term have motivated investigators to study more physiologic treatment options that would maximize axial growth and allow spinal motion, and maintain intervertebral disk health, while limiting or correcting the scoliotic deformity. The goal would be to provide a means for children to "grow out of" their deformity and end with a more normal spinal contour while preserving functional motion. This chapter discusses the rationale of applying an anterolateral flexible tether as a growth modulation technique, summarizes past and current research endeavors in this field, provides a preliminary assessment of the clinical application of this technology, and provides a perspective for further use of fusion-sparing growth-modulating surgery to treat childhood spinal deformities.

44.1.1 Scoliosis Deformity Development and Progression Theory Overview

Temporal relationships between scoliosis development and deformity progression with periods of rapid spinal growth have encouraged investigators to evaluate etiologic theories of vertebral and spinal growth abnormalities. The spinal deformity in main thoracic idiopathic scoliosis has been associated with relative anterior thoracic spinal overgrowth and a lordotic apical region [7, 8]. Dickson et al. [9] theorized that axial rotation and lateral deviation of the thoracic spine occurred in an attempt to balance asymmetric growth in the sagittal plane. When thoracic lordosis increases, global sagittal balance can only be achieved by a rotational buckling of the spine. In addition, several morphological studies have identified intravertebral deformities in patients with idiopathic scoliosis [10–12]. In 2003, Guo et al. [13] proposed that the pathomechanism behind vertebral body asymmetry was disproportionate endochondral versus intramembranous growth. Thus, asymmetric spinal growth and biomechanical imbalance, perpetuated by the Hueter–Volkmann effect, has been postulated as an etiology for the progressive deformity observed in idiopathic scoliosis. Consequently, treatment strategies have been developed that would correct the vertebral growth asymmetry while preserving spinal mobility and long-term function (Fig. 44.1a–d).

44.1.2 Spinal Growth Modulation Theory Overview

Spinal growth modulation provides an important alternative to treat spinal deformity in the young child [14]. As naturally occurring animal models with spinal deformity are not available, growth-modulating techniques are used to either correct iatrogenic deformities created in animal models [1, 15–18] or are used to create a deformity in a straight spine [19–26]. Scoliotic animal models created by posterior spinal tethering or rib tethering may not best mimic the human condition as the disks and growth cartilage have already experienced unusual forces prior to application of the growth-modulating treatment. Spinal growth modulation to create a scoliotic deformity from a normal immature animal spine may in fact be more similar to the clinical condition and more appropriate to the study the responses of the vertebral body and intervertebral disk to the forces created by growth modulation (Fig. 44.2a–d).

44.2 Growth Modulation Applications in the Spine

Nonsurgical applications of growth modulation of the spine (casting and bracing) have regained popularity recently. Bracing may be used for smaller curves as a means of loading the spine and allowing controlled distribution of forces within the spine, thus modulating growth if worn during a growth spurt. [27] The efficacy of bracing has been established by a multicenter randomized cohort by Weinstein et al., and the trial

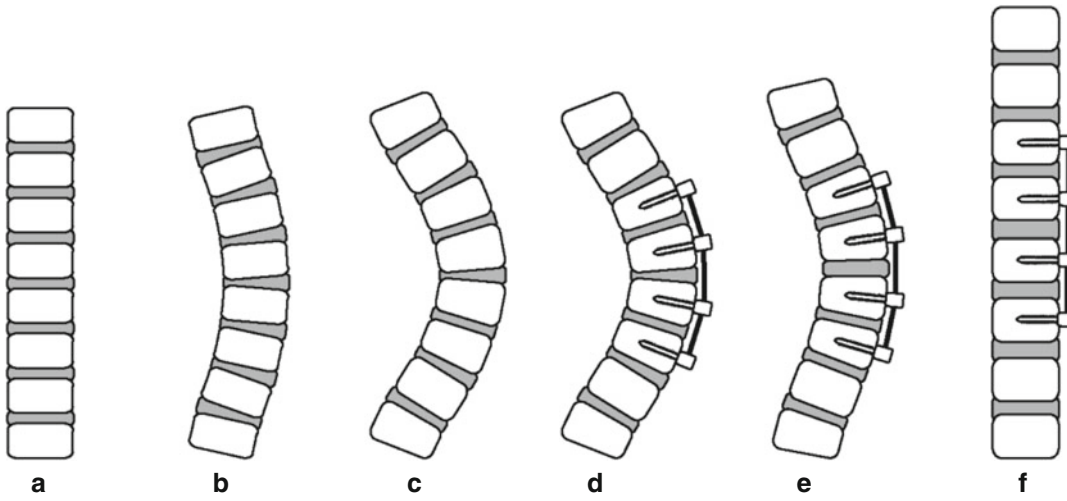


Fig. 44.1 Progression of scoliosis with growth from a straight spine (**a**), with disk wedging (**b**), and finally vertebral wedging (**c**). If the deformity is treated with a tether placed on the convex side to limit growth (**d**), disks straighten (**e**), then finally vertebrae grow straight (**f**). Once the tether is removed or cut, the spine has theoretically returned to a normal straight spine. A similar effect may be seen in the sagittal plane

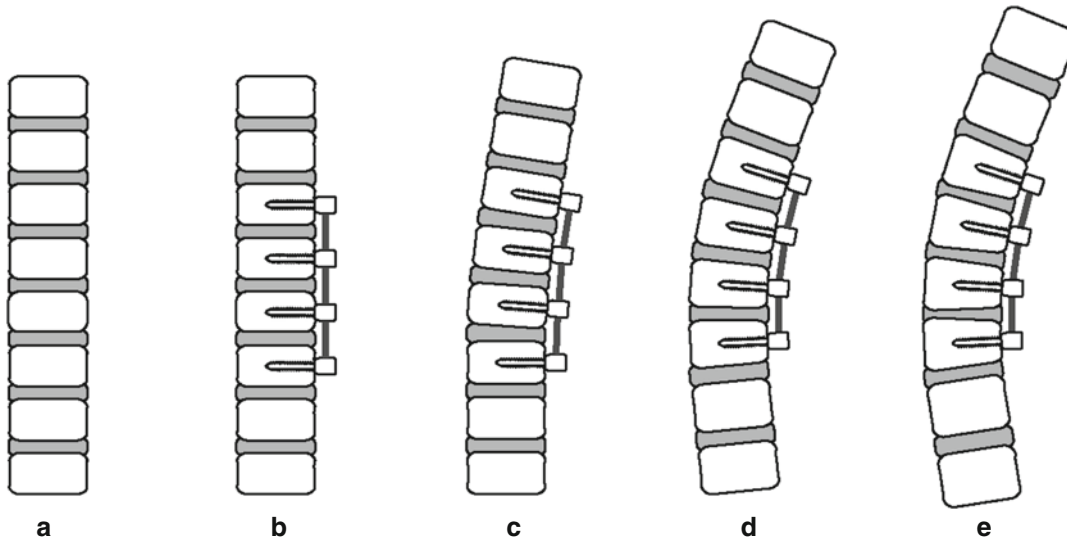


Fig. 44.2 Experimental growth modulation starting with a straight animal spine (**a**). A tether to growth is placed unilaterally (**b**), first resulting in deformity of IVD (**c**), then VB wedging with straightening of the IVD (**d**), and finally, VB wedging to create deformity with a metabolic response of the disk which results in "reverse" wedging of the disk (**e**)

was stopped early due to the documented efficacy of bracing [28]. However, the effectiveness of brace wear is strongly dose related, and there are clear limitations of brace use related to compliance of wear and in the selection of which patients actually benefit from brace wear.

Surgical methods that do not involve the use of implants, such as convex side costoplasty to treat the cosmetic deformity of the posterior rib hump, have provided evidence for the effect of rib asymmetry in the pathogenesis of idiopathic scoliosis [29]. Experimental studies have shown

that scoliosis can be corrected by rib length modulation (shortening or lengthening), taking into account the growth potential of the spine and rib cage [30–32]. These procedures, however, have yet to gain wide acceptance, as their biomechanical effects on the spine are not well understood, and other treatment options are increasingly available. A unilateral fusion, often termed a hemiepiphyseodesis, as a means of altering spinal growth has been used primarily in congenital scoliosis but may have a limited use in other forms of childhood scoliosis [33]. The technique has had mixed outcomes, but in some cases, deformity correction has been noted (if the age is less than 5 years, deformity less than 50°). The downside of this approach is the loss of motion associated with the arthrodesis, the loss of future growth, and the relatively low reliability of true growth modulation with the technique. Such a hemiarthrodesis likely acts as a boney tether to alter spinal growth.

Asymmetrical growth at the neurocentral synchondrosis (NCS) has also been considered as another possible etiology for the three-dimensional spinal deformity seen in AIS. In theory, an abnormality at the neurocentral junction is thought to produce pedicle length asymmetry, thus leading to vertebral rotation [34]. Vertebral wedging is then thought to result from rotation-induced increases in compressive loads on the vertebral growth plates [35]. Once the initial deformity is present, scoliosis is thought to develop due to the propagation of asymmetric loads by the Hueter–Volkmann effect. Theoretically, altering this pattern could halt progression and may provide a means for correction. Previous anatomic studies of normal and scoliotic vertebrae have found an association between a longer pedicle and vertebral rotation [36–38]; however, a causative relationship could not be established. A 2007 biomechanical simulation with asymmetrical pedicle geometry was not able to produce significant scoliosis, vertebral rotation, or wedging [39] and concluded that asymmetry of pedicle growth rate alone was not sufficient to cause scoliosis. On the other hand, asymmetric epiphysiodesis of the NCS was performed in a growing porcine model and was able to create a scoliotic deformity with axial vertebral rotation with the magnitude of the deformity created correlating with the degree of NCS physis closure [40].

44.2.1 Growth Modulation: Anterior Implants

Anterolateral implants are now being used as an internal brace (no patient wear compliance issues) to limit curve progression and ideally improve the deformity during the patient's remaining spinal growth. Such treatment may delay or eliminate the need for a definitive fusion procedure. In theory, the implants could be removed at maturity, leaving the patient with a "normal" spine with full flexibility and function.

Early attempts to provide such an internal brace used vertebral body staples, applying principles learned from growth modulation of the long bones as described in 1951 by Nachlas and Borden [41]. However, unlike the situation with long bones, the vertebral staples were required to cross a joint (intervertebral disk), and loosening of the implants was a common concern. Advancements in staple design, primarily the use of a temperature-dependent shape memory metal (nitinol), have led to more promising clinical results in terms of safety and efficacy to stabilize or modulate growth in moderate-sized curves [1, 15, 42–47]. A biomechanical study evaluating spinal flexibility after instrumentation with nitinol staples found that they staple significantly restricted motion, especially in axial rotation and lateral bending [48]. However the condition of the disks of these patients remains unknown and requires long-term clinical assessment.

Other designs allow more motion through the disk while providing selective force application required for spinal growth modulation [49]. The staple hemiepiphysiodesis technique consistently created spinal deformities in the coronal plane in a porcine model [26]. Driscoll et al. has reported using a hemi-staple which bridges vertebral body growth plates in a porcine model with the unique attribute that the staple does not cross a disk [50]. This model achieved growth modulation, evident

by histological changes in the physes, and the disk remained viable; however, it is not certain how this technology may be applied clinically as there is no epiphysis in the human vertebral body.

In a study by Braun et al. [51], a variety of fusionless scoliosis implant strategies were tested in the rat tail model. This study used both rigid and flexible implants to modulate vertebral body growth. The results from that study demonstrated that dynamic loading of the vertebrae provided the greatest growth modulation potential. Aronsson et al. similarly showed that alternating compression and distraction applied to adjacent vertebrae in the calf tail could modulate vertebral growth, suggesting that dynamic motion would be preferred [52].

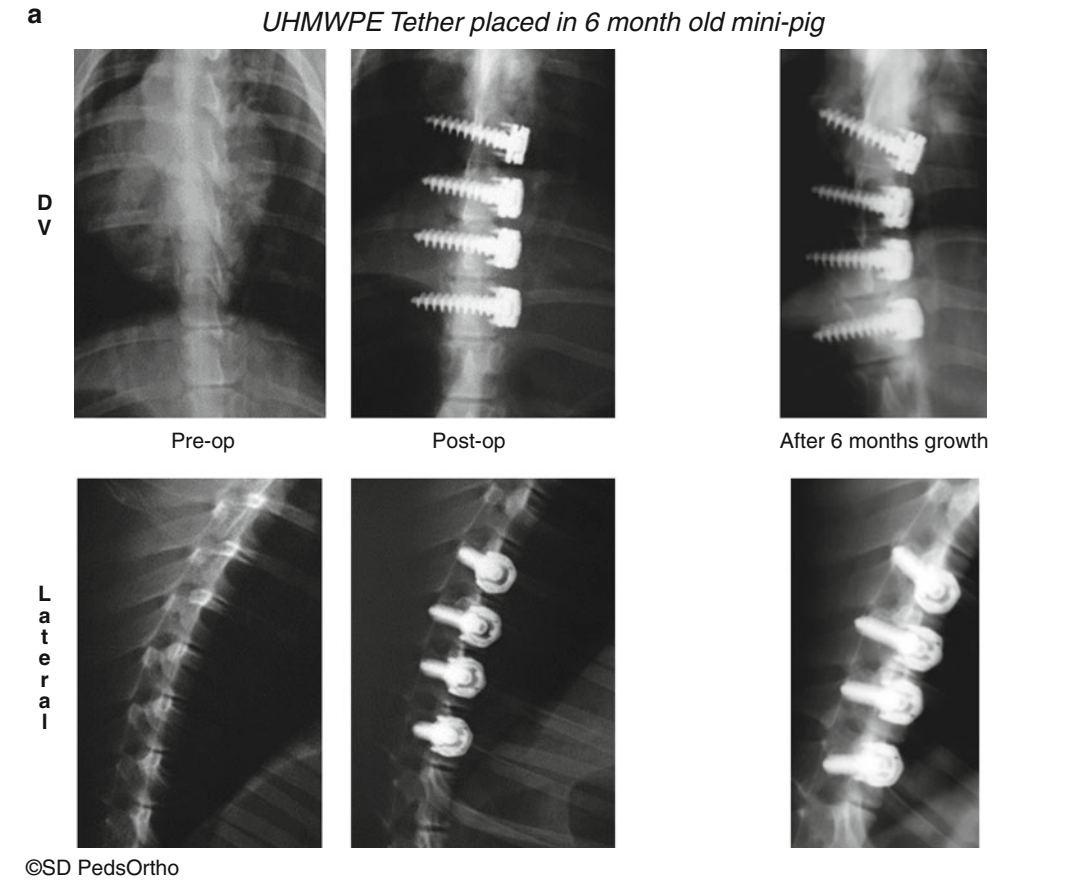
44.2.2 Basic Science Research Overview

Anterior vertebral body tethering provides an alternate approach for spinal growth modulation. Similar in theory to vertebral staples, the tether creates a compressive load on the anterior vertebral body and, through the Hueter–Volkmann principal, attempts to correct the asymmetric anterior spinal overgrowth [7–9, 53]. A theoretical advantage over vertebral staples, however, is that anterior tethering provides a less rigid construct in the directions of motion other than lateral bending away from the device and thus be less detrimental to long-term intervertebral disk health and spinal motion.

Anterolateral spinal tethering has been tested previously in various animal models. In 2002, Newton et al. [20] evaluated the effects of flexible mechanical tethering of a single motion segment. Eight immature calves were instrumented with anterior vertebral body screws over four consecutive thoracic vertebrae. Two screws were connected by a stainless steel tether, and two were left unconnected. After 12 weeks of growth, coronal and sagittal plane deformities were consistently created over the tethered motion segments, compared to control segments. In addition, vertebral body wedging was observed, indicating that physeal growth had likely decreased on the side of the tether. Biomechanical analyses revealed that the tether restricted lateral bending range of motion; however, this motion was found to return to control levels when the tether was removed. A follow-up study evaluating multilevel growth modulation in the bovine model concluded that given the adequate bony fixation, the flexible tether was able to consistently create a biplanar spinal deformity without having a detrimental effect on spinal motion [21]. Another study used an ultrahigh molecular weight polyethylene (UHMWPE) anterolateral spinal tether in a mini-pig model which more closely matched the size and growth rate of an adolescent patient. This study demonstrated the creation of vertebral body wedging in a tethered group of 14° after 6 months and 30° after 12 months, compared to a surgical sham group with no deformity creation (Fig. 44.3a, b) [25]. Additionally, anterior vertebral tethering of experimental scoliosis has been shown to alter growth, thus spinal contour and vertebral body shape, in three dimensions, decreasing coronal curvature, increasing kyphosis, and decreasing axial rotation [54].

In 2005, Braun et al. [17] compared the ability of shape memory alloy staples and bone anchor ligament tethers to correct an experimental scoliosis in 24 Spanish Cross-X goats. The flexible ligament tethers were found to improve scoliosis from an average of 73.4–69.9°, while scoliosis actually progressed in the goats treated with staples from an average of 77.3–94.3°. Pullout testing demonstrated that the bone anchors had improved integration into the vertebral bodies, while the staples were found to loosen. Histologic evidence of a halo of fibrous tissue around the staple tines was also presented and was thought to be responsible for staple loosening.

Intervertebral disk health has also been assessed after spinal growth modulation with anterior vertebral body tethers, as fusionless treatment strategies need to preserve the intervertebral disk if they are to be successful in the long term. Histologic and biochemical evaluation of intervertebral disk health after spinal growth modulation was reported by Newton et al. [22] Intervertebral disks from seventeen bovine instrumented with a multilevel flexible steel cable



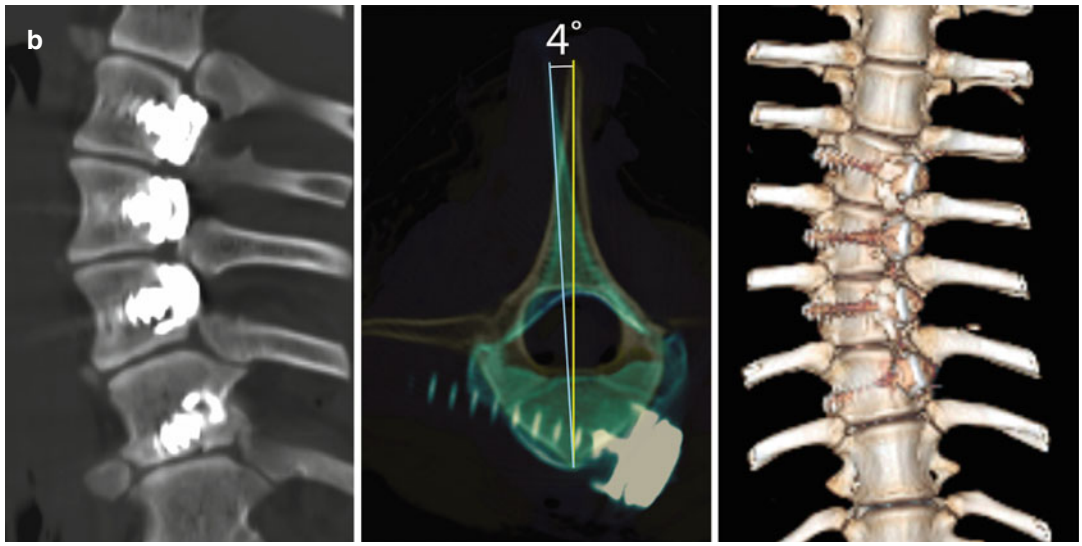


Fig. 44.3 (**a**) Radiographs showing creation of spinal curvature with a tether (radiolucent) in six months of treatment. (**b**) Representative computed tomography (CT) imaging done after 12 months of spinal growth modulation with a tether in a mini-pig. Midcoronal image demonstrates vertebral body wedging, axial image shows rotational deformity created, and anterior–posterior 3D CT image shows global deformity creation

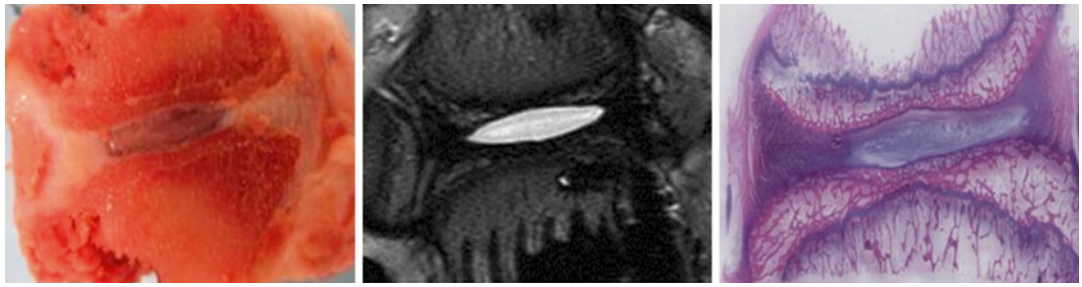


Fig. 44.4 Midcoronal views of intervertebral disks following tethering in a mini-pig model. Gross morphology demonstrates Thompson Grade 1 [83], T2 MRI demonstrates well-hydrated nucleus and intact annulus, and H&E-stained histology section shows all structures to be intact

were compared with disks from 19 bovine that underwent a control sham surgery (screw-only). A double screw-double tether construct was required in this rapidly growing model to achieve adequate bony fixation. No change in disk water content or gross morphological grading was observed between the two groups; however, decreased disk thickness, increased proteoglycan synthesis, and a change in collagen distribution were present in the tethered disks. 3D disk reconstructions showed that the tethered disks were significantly shorter than sham disks (24 % decreased on left and 34 % decreased on right) [55]. Disk narrowing and wedging is seen in clinical scoliosis and in animal models which mimic the clinical situation [56]. In the mini-pig model, the vertebral body wedging created (convex on the side of the tether) was accompanied by wedging in the disks opposite to that in the vertebrae (convex on the contralateral side of the tether). Disks from spines with tethers were found to be hydrated, with MR signals similar to sham surgical spines (Fig. 44.4a–c) [23, 25]. Another concern is how the disk would respond to a tether with an applied tension, as would be the case in a clinical surgery where the surgeon desired at least a partial deformity correction intraoperatively. Previous work comparing tensioned and untensioned anterolateral spinal tethers in a mini-pig model was done using a thoracotomy and a tensioning device [24]. Results showed that tensioning to 250 N caused 8° of deformity over four vertebral levels, between three disks. The tensioned group did develop a deformity more quickly initially, but after 7 months of tethered growth, the deformities of tensioned and not tensioned were the same. Pretensioning the tether did not result in disk injury or a decreased ability for the growth to be modulated over time. Fusionless scoliosis implants have been found to result in alterations in intervertebral disk cell density [57] and collagen content; [22] however, the clinical implications of these changes are unknown. Further studies of disk health with noninvasive imaging modalities for the detection of early degeneration and after long-term growth modulation are required.

The mechanism by which application of compressive forces unilaterally via vertebral tethering acts to modulate that the growth of vertebral bodies and of the global spinal contour is being explored and is just starting to be understood. Unilateral compression of a growth plate may result in changes to the growth plate. Indeed, Chay et al. found that tethering an experimentally created scoliotic curve in pigs resulted in a decrease in proliferative zone height on the side of the tether compared to the contralateral side of the vertebrae [58]. Others have found a decrease in hypertrophic zone height and cell numbers with a unilateral application of compression on the vertebral body [59]. Using 3D measurements of the growth plates from calves that had tethers placed, Newton et al. likewise showed a thinning of the physis on the side of the tether in the tethered spines [55]. In all of these studies, even though the growth plates were altered from their normal state, they remained open for the duration of the treatment, and thus potential future growth was preserved. Further work in animal models

involving growth plate response to changes in the mechanical environment continues and is particularly promising as growth cartilage in various bones across various species seem to respond similarly; thus, it could be especially appropriate to apply to growth in human vertebrae [60].

44.2.3 Posterior Spinal Tether

Vertebral tethers have also been used in the posterior spine to modulate spinal growth. In 2005, Lowe et al. [61] evaluated the ability of polyethylene cords to effect sagittal alignment in an immature nonfusion segmental pedicle screw sheep model. At 13 months after surgery, the tethered animals had significantly less kyphosis and vertebral body wedging than the control animals. The authors concluded that the posterior tether may be a potential treatment for adolescents with Scheuermann's disease. However, there are no clinical reports of use of a tether to correct this deformity, and the usefulness to modulate growth in Scheuermann's disease, which often presents after the adolescent growth spurt [62], is yet to be studied.

44.3 Anterolateral Spinal Tether: Clinical Application

44.3.1 Indications for Intervention

The specific indications and contraindications for juvenile spinal growth modulation are evolving with greater clinical experience. The ideal application is likely for the treatment of preadolescent idiopathic scoliosis with >40° curves that have a high likelihood of progression. However, it may be the best treatment choice in curves with higher magnitudes and in those that are progressing beyond the range that is considered to be braceable (>45°). Recently, it has been suggested that tethering may become the growth-modulating technique of choice for curves greater than 40° that are beyond the treatment range for vertebral body stapling effectiveness [63]. Our current indications to consider tethering are for patients with primary thoracic curves of 45–65° in females who are Risser 0 and males Risser 0 or 1. The thoracic spine should be hypokyphotic. Indications for tethering thoracolumbar curves remain unclear. It is very important that patients are carefully screened and their family situation is well known to be stable so that they remain available for close monitoring – as with all growth-modulating applications, there is a risk of overcorrection and the consequences can be serious.

44.3.2 Timing of Surgery

The timing of tether placement is crucial to its success. Treatment needs to be initiated in time to allow enough growth to modulate the vertebrae to become "straight." However, if the tether is applied too early, the risk of over correction increases. At this point, there are no clinical studies, as there are for lower limb length inequality [64, 65] that allow for definitive determination of what age/skeletal maturity stage is ideal, not too early or too late, to intervene. The determination of the amount of remaining growth in a candidate patient is essential and has been traditionally determined using the Greulich and Pyle method which uses a hand x-ray to determine skeletal age based on an atlas of skeletal development published in 1959 [66]. In routine clinical practice, the Risser grade of ossification of the ilium (grade 0–5) has been routinely used to determine skeletal maturity, and only children with Risser of 0 or 1 have been considered young enough to benefit for a spinal growth modulation procedure. The Risser sign remains 0 (no ossification of the iliac crest) until after a patient's peak spinal growth velocity has passed. Sanders et al. have found that open triradiate cartilage is a strong predictor of a child being at their peak or pre-peak of spinal growth velocity which is associated with continued anterior column growth [67]. The triradiate cartilage closes prior to the appearance of a Risser sign and therefore is an earlier predictor of the beginning of maturity. As such, open triradiate cartilage may be the best current criteria for determining that a patient has "enough" remaining growth to maximize the tether effect for

larger curves. The Tanner-Whitehouse-III RUS scores correlate skeletal age of the metacarpals and phalanges with scoliosis curve progression and can show maturity stages in the age range where triradiate cartilage is open; thus, it is an earlier predictor of the growth "spurt." [68] One method of determining skeletal growth remaining and the likelihood of scoliosis progression that is more practical in a clinical setting is proposed by Sanders et al. and uses a modification of the Tanner-Whitehouse-III score [69]. Another popular method for assessing skeletal maturity, particularly for patients with open triradiate cartilage, is the Sauvegrain method which correlates elbow ossification centers with peak skeletal growth [70, 71].

44.3.3 Intraoperative Considerations

In the 1990s, the use of the thoracoscopic approach to the spine [72] became popular as a safe [73] means for anterior thoracic discectomy to provide spinal flexibility for improved deformity correction [74–76] and to allow anterior fusion to prevent crankshaft deformities [77, 78]. Anterior instrumentation has also been successfully placed in main thoracic scoliosis, with 5-year postoperative outcomes comparable to those achieved for open anterior and posterior techniques. [79] Two decades later, the widespread safe use of pedicle screws as a powerful means for deformity correction and stabilization greatly decreased the need for anterior discectomies, and thus, the use of the thoracoscopic approach also decreased; however, it is still used for very severe and stiff curves and other select cases with specific indications for anterior release and/or fusion [80]. The thoracoscopic approach is experiencing recent renewal with anterolateral growth modulation techniques, including the tether, as new applications of the approach.

The current experience using anterolateral tethering techniques is discussed here; however, this technique remains novel, and there is very little published experience in its clinical application and no published reports on the clinical use or outcomes of the technique.

44.3.4 Thoracoscopic Approach

The authors prefer a thoracoscopic approach for tether insertion. Basic thoracoscopy skills are required in order to safely navigate the chest cavity (Fig 44.5a–c). Spinal deformity surgeons may find the skills of a general/thoracic surgeon with thoracoscopic experience a great value in performing this procedure. The details of performing thoracoscopic spinal instrumentation can be found in other sources [81, 82]. Briefly, the lungs are selectively ventilated allowing the lung on the convex side to collapse creating the working space. As with traditional instrumentation for fusion, three 15 mm ports are placed along the posterior axillary line in positions that will allow direct lateral access for screw placement. A single anterior axillary line portal at the apex of the curve is used for scope placement (See Fig. 44.6a). The pleura is incised longitudinally, and the segmental vessels are coagulated and divided using ultrasonic energy. The azygous vein and great vessels are dissected from the anterior spine by packing a sponge between these structures and the spine. This is an added step that creates space and protection when the vertebrae are drilled, tapped, and screws placed.

A pilot hole is created in the midportion of each vertebral body and the width of the vertebrae determined with a depth gauge. Pronged washers are used in combination with vertebral body screws to reduce the risk of the screws levering/plowing. Screws, augmented with hydroxyapatite (HA) coating, are placed with a direct lateral trajectory into each vertebra included in the scoliosis deformity (upper end vertebra to lower end vertebra). The screws are placed as parallel to the end plates as possible with bicortical purchase for stability. The image intensifier is utilized to confirm trajectory and screw length (See Fig. 44.6b). Once the screws are placed, the cord-like polymer tether is placed, connecting the screws. A locking set screw secures the tether to the screws (See Fig. 44.6c). Tension can be applied to the tether, between screws utilizing an external tensioning device, to provide an "on the table" tensioning and correction as desired, depending on the deformity severity and the growth remaining of the patient.

The senior authors' experience is that it is helpful to lay the tether in place and secure it to the proximal end of the construct first, then apply tension to the adjacent segment and secure the tether in place, continuing in this manner by tensioning individual segments. An attempt to bring the wedged disks (thinner on concave side and thicker on convex side of the scoliotic spine) to parallel without reversing the wedging may be the best criteria for tension application interoperatively. Less tension is required at the ends of the construct as less deformity is present in these segments. However, the actual amount of tension that is being applied is difficult to determine with current methods. Caution is advised against over tensioning as this may be detrimental to disk physiology as well as

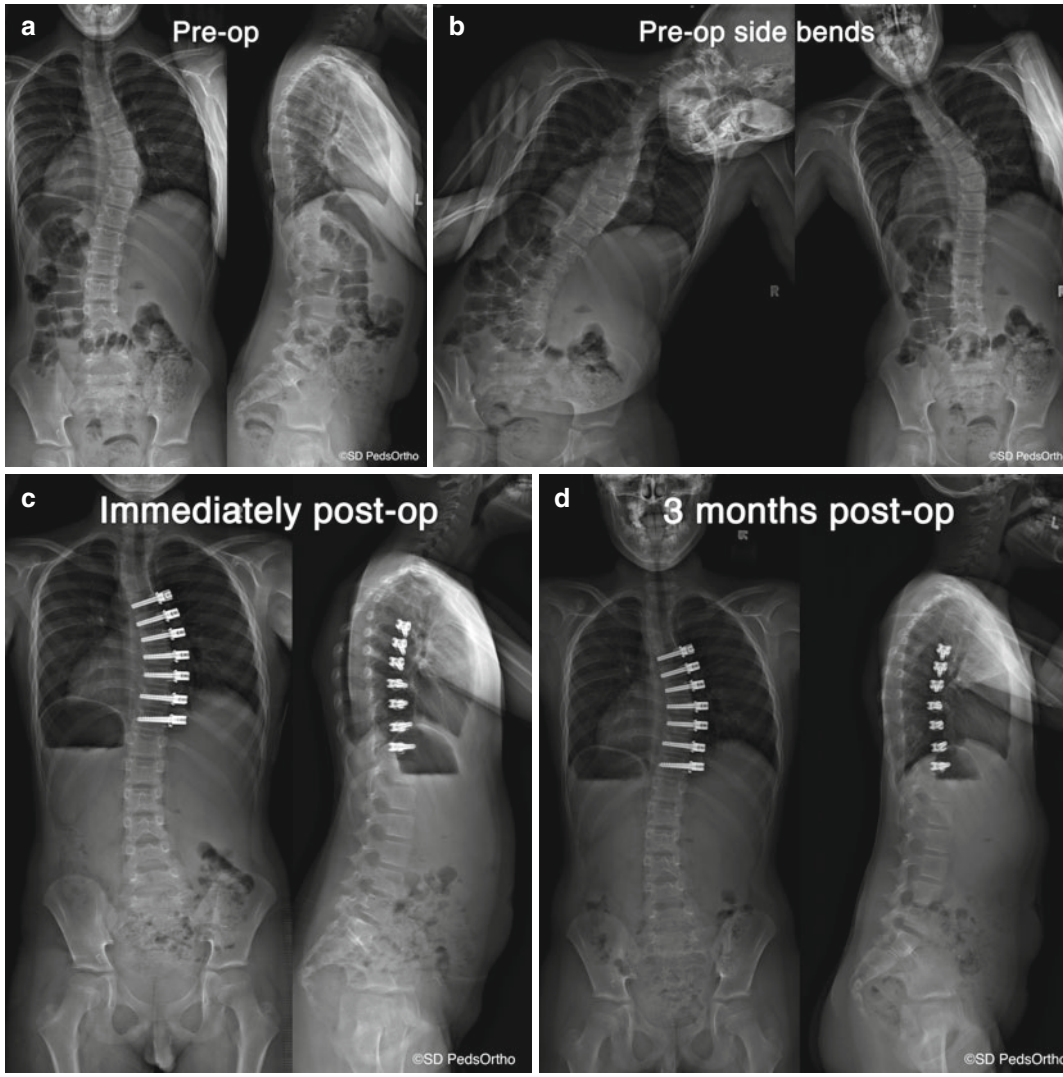


Fig. 44.5 An example case of growth modulation to treat scoliosis using an anterior tether. (**a**) Standing preoperative PA, lateral, and (**b**) side-bending radiographs of a 10-year, 2-month-old boy with 44° right thoracic curve. (**c**) Two weeks following anterior spinal tether placement from T5 to T11, PA image demonstrates a 28° right thoracic curve. The spine gradually straightens over time, demonstrated by radiographs (**d**) 3 months, (**e**) 6 months, (**f**) 12 months, (**g**) 18 months, (**h**) 24 months, (**i**) 2.5 years, and (**j**) 3 years following tether placement

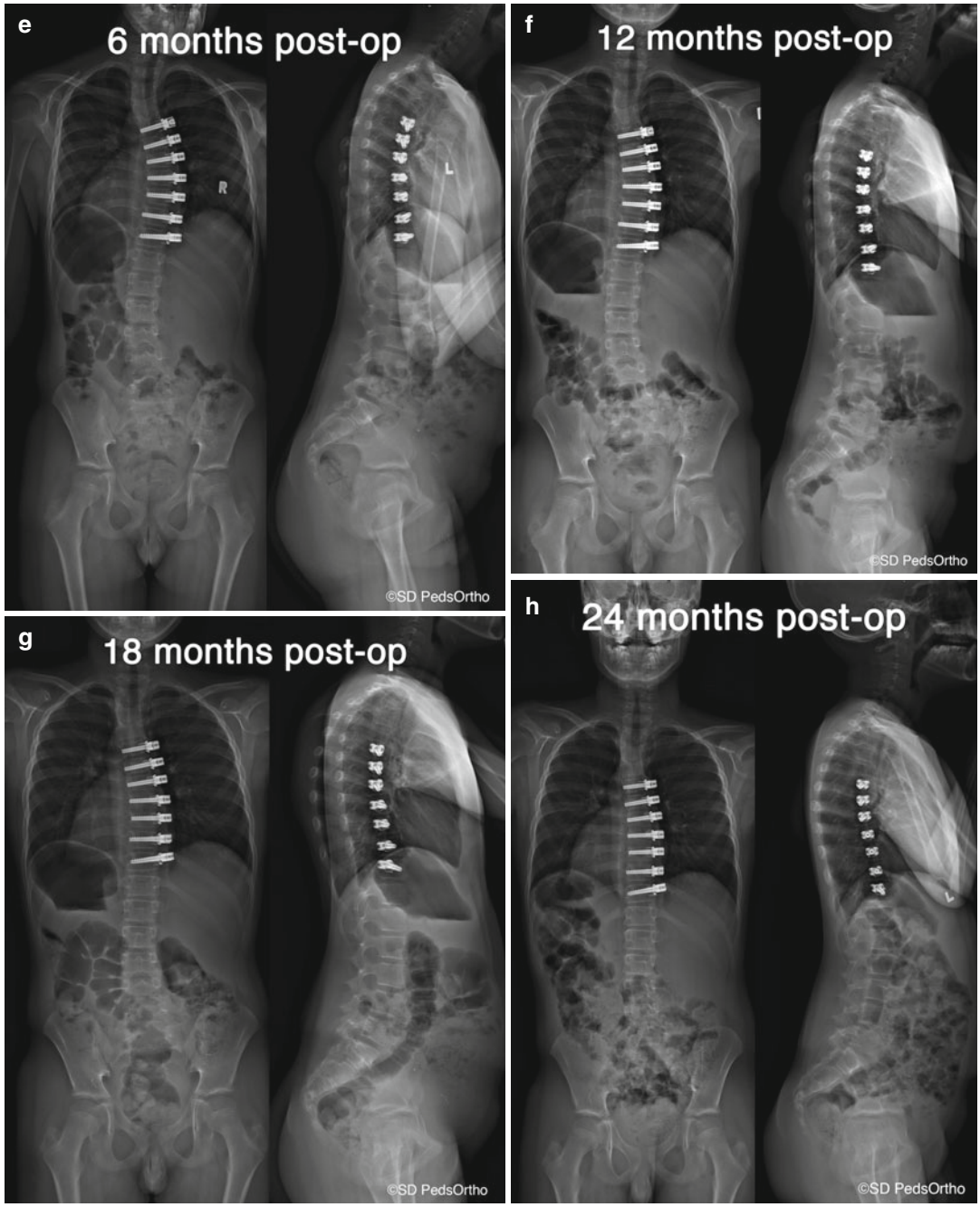
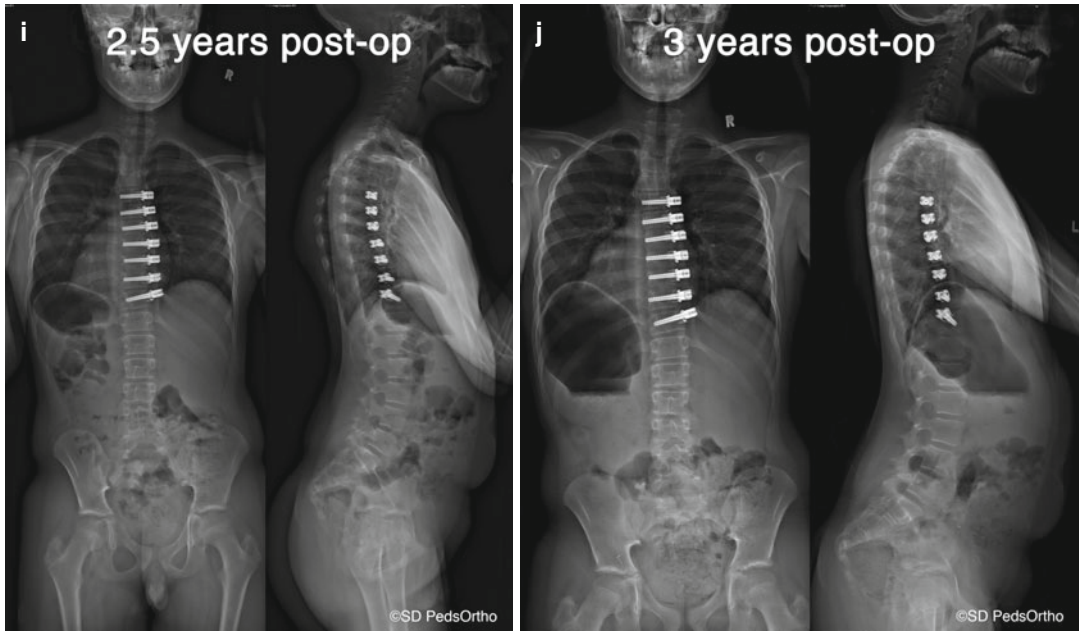


Fig. 44.5 (continued)



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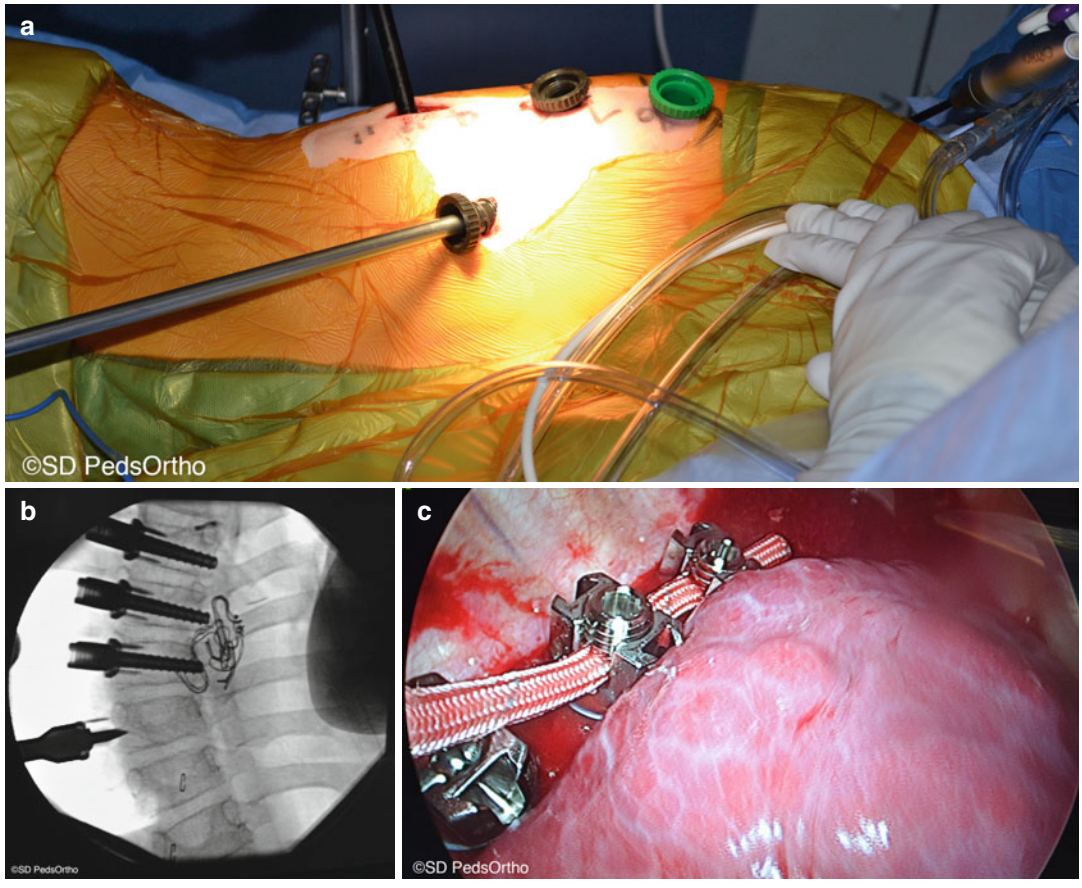

lead to rapid overcorrection particularly at the distal end where growth is the fastest.

Some technical issues may need to be considered as they could cause increased risk of complications to the patient. If a thoracolumbar curve is to be treated, the diaphragm may need to be divided and then repaired. If later revision of the device is required for loosening or removal, pleural adhesions to the device and chest wall should be anticipated. This may add to the challenge of the exposure and may necessitate conversion to an open or mini open approach.

44.3.5 Post-op Care

A chest tube is utilized for several days as required. Patients are placed in a brace for 3 months following surgery to allow for incorporation/stabilization of the HA-coated screws into the vertebral bone. Following surgery, biplanar radiographs should be performed every 6 months until skeletal maturity. Follow-up is extremely important as this device is a powerful modulator of spinal growth, and overcorrection is a clear risk. The patient and family must understand this risk from the outset with agreement of the follow-up requirement. A systematic method of tracking such patients is suggested.

44.3.6 Further Interventions

If and when the spinal deformity is fully or slightly overcorrected with the tether, the treatment needs to be terminated to prevent inducing a curve in the opposite direction. This may be done by cutting the tether and maintaining the implants, loosening the tether, or by removing the implants entirely.

44.4 Anterolateral Spinal Tether: Clinical Experience and Case Report

Clinical application of the anterior vertebral tether has been performed by a few surgeons in select patients over recent years. None of the growth-modulating devices are FDA approved for this use, and as such the implants are used "off-label" or "physician directed." Clinical studies will be needed to determine the true efficacy of these techniques.

An example case is presented of successful growth modulation using tether treatment in a boy with Prader–Willi Syndrome and progressive scoliosis (Fig. 44.5a–j). He has been treated with growth hormone currently and since infancy. Preoperative posterior–anterior (PA), lateral, and side-bending radiographs (See Fig. 44.5a, b) of a 10-year 2-month-old boy who has had progressive thoracic scoliosis despite TLSO brace treatment demonstrate a 44° right thoracic curve with some flexibility (see Fig. 44.5b, bend to the right). An anterior spinal tether was placed through a thoracoscopic approach from T5 to T11 (tether is not seen on x-ray as polymer material is radio lucent). Postoperative radiographs, taken standing 2 weeks post-op (See Fig. 44.5c), demonstrate a 28° right thoracic curve (36 % correction immediately post-op). At 1-year post-op, radiographs (See Fig. 44.5f) demonstrate a 20° right thoracic curve (44 % correction), 1.5-year post-op 10° (77 % correction), and 2.5-year postoperative radiographs show complete resolution of the curve, with an overcorrection of 5° (See Fig. 44.5i). At 3 years post-op (See Fig. 44.5j), the patient is 13 years old and remains skeletally immature. Disk space is preserved, and spine deformity is corrected. There is no evidence of

Fig. 44.6 Thoracoscopic lateral approach. (**a**) The patient is placed laterally recumbent with ports placed for scope, lung retractor, and working instruments. (**b**) Fluoroscopy image of the anterolateral vertebral body staple and screw placement in three vertebrae and one staple being placed. (**c**) Placement of a polymer tether into the screws, viewed through the scope (retracted lung on the right side of the image)

screw loosening. The treatment plan is to continue to monitor radiographically every 4 months and to remove the tether when/if he demonstrates progression of overcorrection. If this patient does not continue to be monitored closely, there is a very real risk of overcorrection and a reverse deformity being created by the tether. There are no clinical data that can help to determine whether this patient's deformity may return if the tether is removed while he is still skeletally immature with further spinal growth remaining. To prevent overcorrection, this patient could have received tether surgery at a later date, or perhaps the correction "on the table" could have been less. The ideal situation for a patient such as this one still remains to be determined.

Conclusions

Both spinal motion and spinal growth are unfortunately commonly sacrificed in the treatment of young patients with spinal deformity. New treatment solutions to overcome this current limitation are required. While several nonfusion treatment strategies to preserve motion and growth are under investigation, it is likely that more than one solution will be required to account for the numerous complexities of spinal deformity in the growing child. Although long-term clinical data are not available at this time, anterior spinal growth modulation via a flexible tether provides an exciting alternative to controlling progressive scoliosis, while maintaining spinal motion. By understanding normal and pathologic spinal growth, it is hoped that solutions to modulate spinal growth can be created to successfully correct spinal deformities in childhood and adolescence without the need for a spinal fusion.

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Growth Modulation Techniques: Titanium Clip-Screw Implant System (HemiBridge)

45

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Key Points

- Spinal growth modification using a thoracoscopically implanted clip-screw construct is described, including design objectives, theoretical advantages, mechanical function, clinical safety, surgical technique, and early radiographic and quality of life results.
- The titanium instrumentation system has been tested prospectively from first human use in a US FDA-approved clinical trial under an Investigational Device Exemption (IDE) for treatment of late juvenile and early adolescent idiopathic thoracic scoliosis. Eligibility criteria were chosen to include only those subjects at high risk of curve progression to >50° by published criteria.

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- Individual results were variable and included one subject who required instrumentation and fusion. However, proof of concept in humans was also demonstrated in one child whose curvature was corrected >60 % within 2 years.
- The device is approved for use in the European Union (CE mark). The pivotal-phase IDE clinical trial is approved by the US FDA.

45.1 Introduction

Asymmetrical compression of the vertebral body physes has long been considered a contributing factor in scoliosis [1]. Spine deformity progression is an example of a positive feedback loop or vicious cycle [2, 3] as a mechanism of disease. The concept of redirecting vertebral growth by redistributing stresses using anterior instrumentation as a reversal of the process of progression was discussed by Dwyer et al. [4]. Stapling of the spine for the purpose of modulating growth was attempted in the mid-twentieth century [5], but implant loosening and preoperative curve severity were later noted as problems.

Studies using an animal model to explore spine growth modulation began more than 60 years ago [6]. For the in vivo preclinical tests that led to the current system, the term "mechanical spinal hemiepiphysiodesis" was used to emphasize the application of a compression gradient to the growth plate using a device which continues to allow some intervertebral motion and to distinguish the method from a hemiarthrodesis or localized fusion of the amphiarthrotic disc.

45.2 Preclinical Studies

Preclinical studies beginning in the late 1990s showed that certain implant design factors were critical to the ability to alter spine growth [7]. Device features were related to the amount of curvature induced (Fig. 45.1) and to the degree of structural changes to the physis. The most successful device of the series, a staple-like implant with divergent inner tines fixed to adjacent vertebrae with two bone screws, was shown capable of producing asymmetric growth in otherwise straight spines in a quadruped model, the inverse analog of deformity progression.

To help quantify the Hueter-Volkmann principle, the relationship between physis function and mechanical stress, a study on a readily available clinical model of growth modification was performed [8]. Knee stapling in adolescents with genu valgum was analyzed (Fig. 45.2) under the assumption that the stress, i.e., force divided by cross-sectional area, that a growth plate can exert has a constant maximum value. The study showed that the force to correct knee angular deformities in adolescents was approximately 500 N, or slightly more than the normal body weight, which implied that a compressive stress of about 1 MPa is needed to stop the growth. At the same time, a study in scoliosis patients showed that vertebral body physes continue to exist even on the concave side of the apex of large curves in older adolescents [9] (Fig. 45.3).

Repeatable results were shown using the porcine inverse analog model, with curvatures increasing with postoperative time [10]. In order to determine if the mechanism of increasing curvature included changes to the vertebral physes, rather than consisting solely of asymmetric disc compression, a histomorphometric study was performed [11]. The study showed decreases in the size of the physeal structures which correlate strongly with growth rate, the height of the hypertrophic zone, and hypertrophic cell size on the treated side [12]. These side-to-side differences in growth (Fig. 45.4) indicated asymmetric vertebral growth. Related biomechanical, mechanobiological, and computational [13] studies helped determine initial displacements [14], stresses [15–17], and ranges of motion [18].

In preparation for human use, including implant design changes and titanium rather than the stainless steel implants, several preclinical evaluations were conducted. To align with proce-

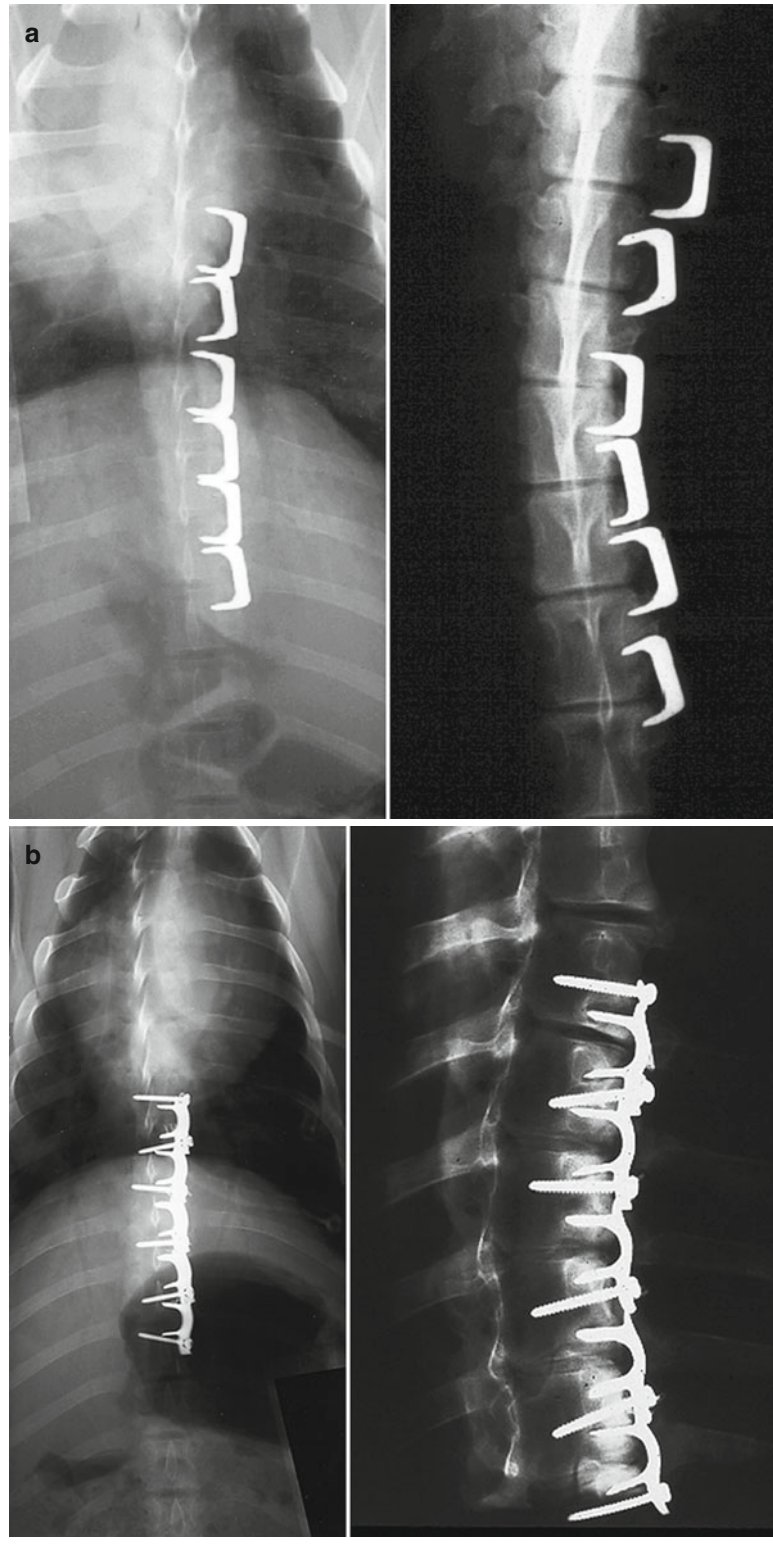

Fig. 45.1 (**a**) Radiographs from preclinical trial of first implant design, knee hemiepiphyseal staples with shortened tines. *Left*: Immediate postoperative. *Right*: 6 weeks postoperative. Bone fixation was obviously insufficient, but local curvature is suggestive at the center, stable, implant level. (**b**) Radiographs from preclinical trial of second implant design, custom stainless steel implant with one bone screw per implant. *Left*: Immediate postoperative. *Right*: 2 months postoperative

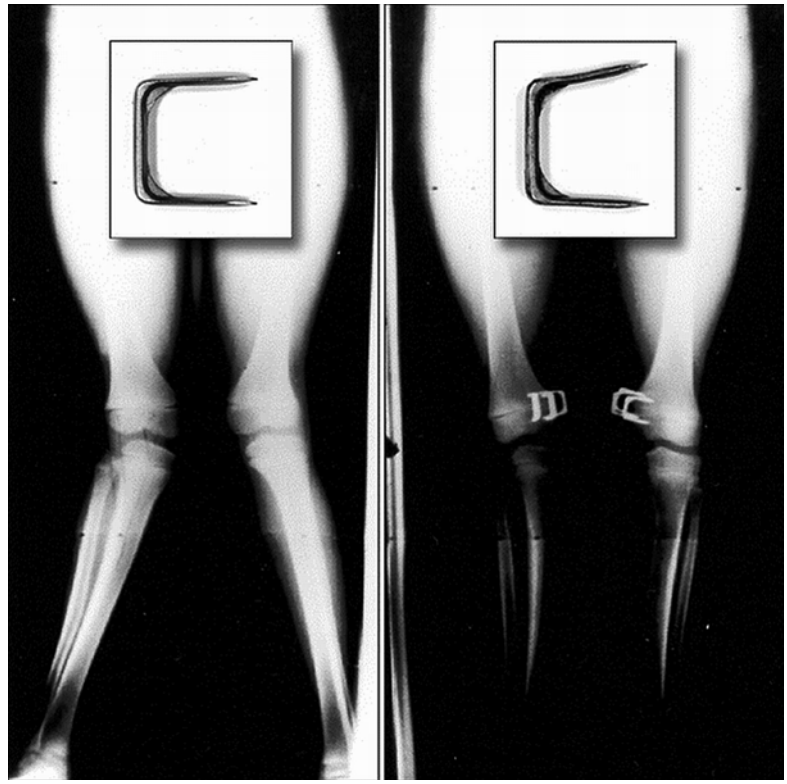

Fig. 45.2 Forces and stresses of growth were determined by experimentally reproducing deformations of hemiepiphyseal staples measured from serial radiographs of adolescents treated for genu valgum

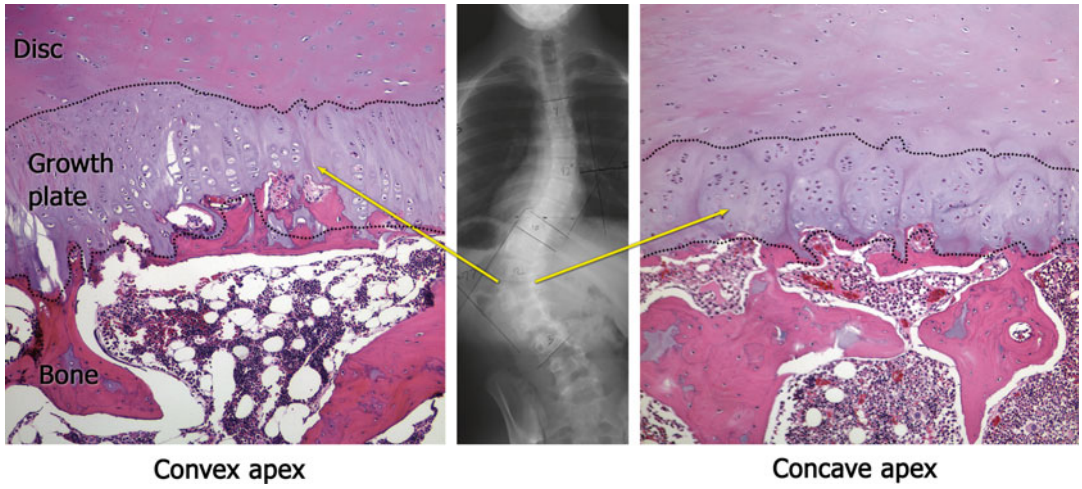

Fig. 45.3 Radiograph and histological sections of vertebral physis from scoliosis patient, 12-year-old male, 74° curve. *Arrows* indicate the side and level of curve sections. *Dotted lines* outline the physis. On the convex side both the hypertrophic cells and the hypertrophic zone of the growth plate are larger than those on the concave side

dural requirements of the US FDA, in vivo tests were conducted in an independent contract with Good Laboratory Practices (GLP) facility [19]. Curves again increased significantly. Results corroborated the repeatability of curve induction in this model (Fig. 45.5). The combined studies demonstrated that the system was effective in inducing asymmetric growth modulation in a

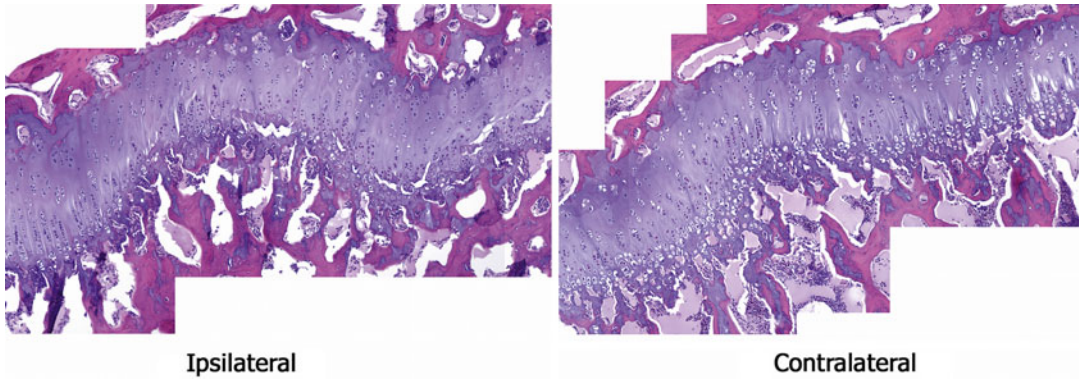

Fig. 45.4 Histological sections of vertebral physis from skeletally immature domestic pig from preclinical trial of last prototype implant of stainless steel with two vertebral body screws. Curvatures were induced in five consecutive animals. *Left*: Sample from treated side, within implant tines. *Right*: Sample from the opposite side of the same level. Qualitatively, the images suggested changes to the growth plate and bone at the instrumented level on the side ipsilateral to the implant. Quantitative structural differences were reported [11]. Similar to the human in Fig. 45.3, the hypertrophic cell height and the hypertrophic zone height of the stapled animals were greater on the side of the vertebrae contralateral (opposite) to the staple versus the side ipsilateral to the staple

quadruped model. Additional conclusions were that the clips and vertebral locking screws were able to be explanted, that the clip and screws remained secure in vivo, and that the clips showed no evidence of permanent deformation.

45.3 Clinical Study

45.3.1 Objectives and Design

A prospective, FDA Investigational Device Exemption (IDE), clinical safety trial was then initiated [20]. The novel intervertebral titanium clip device and surgical instrumentation were designed for the treatment of progressive idiopathic scoliosis (IS) by modulating spinal growth (hemiepiphysiodesis) without the need for spinal fusion. The device was designed as a fusionless, growth modulation system to provide an alternative to external bracing especially for patients who have a lower chance of success with brace wear [21–23]. The system is intended for patients who are diagnosed in the early stages of IS and have a high potential for curve progression.

The mechanical objectives of the clip/screw device are to provide compression on the convex side of the curve and mild distraction on the concave side. The implants are intended to redirect asymmetric growth by distributing vertebral growth-altering stresses on the main thoracic spine segment and by promoting the secondary curves to align and balance. Insertion of the implants realigns the vertebrae, as may be noted in the side-to-side change in disc height in the intraoperative fluoroscopic images (Fig. 45.6) and in the immediate postoperative thoracic curve reductions in the first standing radiographs. This slows growth on the convex side of the curve and may enhance growth or decompress the disc on the concave side. The mechanical design criteria incorporate evidence of the forces of growth and growth modulation stress gradients, as well as physiologic ranges of motion and loading. The construct consists of a clip and two vertebral locking screws (Fig. 45.7) which are placed on the lateral aspect of the thoracic and thoracolumbar spine. The device is designed and sized to bridge superior and inferior physes and intervening disc.

Like staples in long bones, evidence indicates that the device inhibits growth asymmetrically. The construct may be viewed as a combination of a knee hemiepiphyseal staple and an 8-plate. As vertebral growth gradually increases, the time-averaged compressive stress on the physis also increases, which slows growth on the side ipsilateral to the implants. If the average relative com-

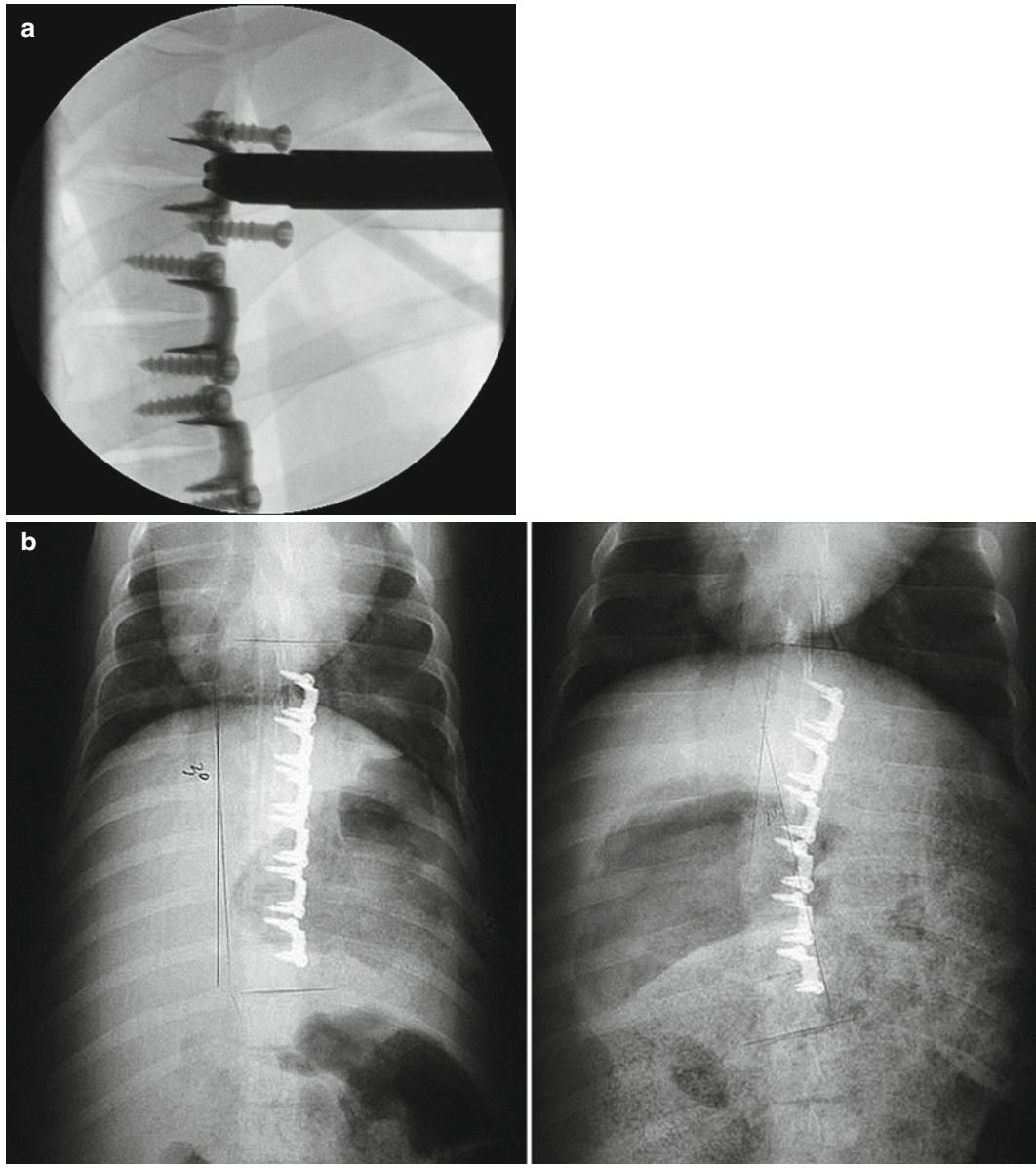

Fig. 45.5 Radiographs from preclinical in vivo study of titanium clip-screw construct in porcine model. (**a**) Intraoperative fluoroscopic image showing clip with two preloaded screws in insertion tool. (**b**) *Left*: Immediate postoperative curve is 3°. *Right*: At 8 weeks postoperatively, the major curve is 20°

pression on the concave side is lowered, the method may also allow for a growth rate closer to normal at the concavity. Disc motion is partially retained. Postoperatively, the compressive stress on the disc and the physis within the tines increases [17] and reaches a side-to-side gradient of a magnitude sufficient to inhibit growth asymmetrically, that is, a time-averaged spatial stress difference of between 0.1 and 1 MPa [8, 24]. Although scoliosis involves growth disturbances in three dimensions, it appears possible that if the thoracic coronal curvature is modified early and sufficiently, and if the initial curve is not too stiff or axially rotated, the otherwise inexorable pro-

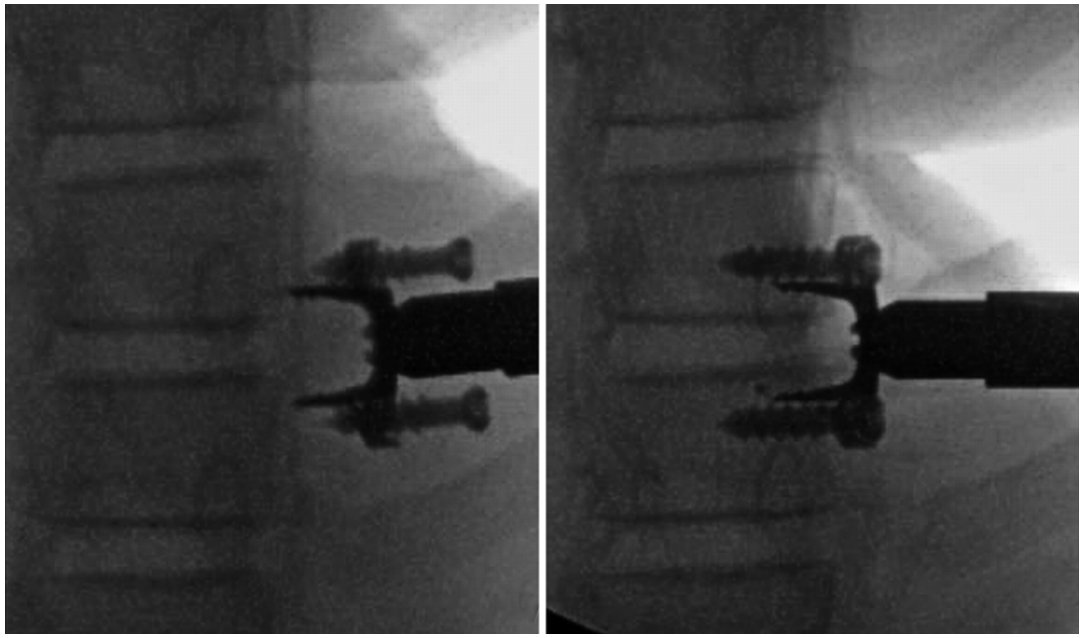

Fig. 45.6 Intraoperative fluoroscopic images from clinical trial show titanium clip with preloaded screws in insertion tool before (*left*) and after implantation (*right*). The disc is slightly compressed on the implant side and distracted on the opposite side

Fig. 45.7 Titanium clip-screw implant construct is shown with preloaded screws

gression of compensatory curves, rib deformities, and axial rotation may be interrupted or perhaps even corrected.

45.3.2 Therapeutic Goals and Theoretical Advantages

Titanium clip-screw implant constructs:

- Arrest or correct idiopathic spinal deformity without fusion or long-term bracing.
- Maintain quality of life; impose few activity restrictions.
- Eliminate compliance with brace dosing schedules.
- Avoid fusion, allow for continued growth of the thorax, and increase chest volume for pulmonary function.
- Apply forces at the vertebral body physes, as opposed to external braces that apply forces to the spine indirectly.
- Spare motion and disc height, which may allow for sufficient disc health maintenance.
- Allow for fusion instrumentation later if necessary.
- Use advantageous material and structural properties of titanium: biocompatibility, rarity of allergic reactions, flexibility in

bending, resistance to fatigue, and osseointegration.
- Allow for entirely thoracoscopic approach and implantation.
- Provide for reversible procedures for implant removal with thoracoscopic procedures
- Reduce operative time and blood loss compared to fusion and so avoid transfusion and keep anesthesia time low.
- Substantially reduce rehabilitation and recovery time compared to fusion.
- Provide strong safety profile: relatively small implants, no bi-cortical instrumentation, no risk to opposite side vessels, very low risk of spinal canal penetration, and low risk of disc encroachment during or after procedure.
- Ease surgical procedures compared to other types of growth modification systems: few parts, no large vertebral body screws, fewer possible variations in implant procedures, and no material phase change to induce implant deformation for shape change.
- Deliver relatively low dose of mechanical compression force and so low probability of overcorrection requiring reoperation. Largely limit biomechanical effects to individual motion segments: no direct interconnection between devices and no end-to-end ties so moments acting on implant fixation sites and laterally directed pull-out forces exerted on screws remain low compared to systems that connect several motion segments.

45.3.3 Patient Selection Criteria for Early Clinical Trial

The primary patient selection criteria for the pilot clinical trial in humans are summarized in Table 45.1

Table 45.1 Patient selection criteria for early clinical trial

Inclusion
Children with idiopathic scoliosis who are at nearly 100 % risk of progression to 50° Cobb angle [21, 23]
Single thoracic curves, Lenke Type 1A or 1B, Cobb angle ≥25° and ≤40
Before or at the beginning of puberty, not yet reached peak height velocity
≥8 years old, Risser 0, triradiate cartilages not closed
Atlas bone age <13 female and <15 male
Exclusion
Serious pulmonary or anatomical condition that would contraindicate an anterior endoscopic approach
Restrictive lung disease or short spine, conditions which may be better suited to rib- or spine-based growing devices
Neuromuscular, congenital, and syndromic scoliosis are relative contraindications
Curves >40°, which may be beyond the ability of localized, single-level, compression-based devices to correct a curve or stop progression

45.3.4 Processes for Study Conducted Under an Investigational Device Exemption

Processes and procedures of the US Food and Drug Administration (FDA) Investigational Device Exemption (IDE) trial were implemented in compliance with Good Clinical Practice (GCP) and ISO 14155 (International Standards Organization Clinical Investigation of Medical Devices for Human Subjects). These are international standards for the design, conduct, performance, monitoring, auditing, recording, analysis, and reporting of clinical trials. The purpose of GCP and ISO 14155 is to codify ethical standards and quality of data.

The clinical study involved spine surgeons who were selected based on their expertise, experience with the target population, and willingness to participate and adhere to the requirements of the IDE trial. Surgeons were required to certify that they would follow the Investigational Plan approved by the FDA and by their Institutional Review Board (IRB) by signing an Investigator Agreement. Clinical trial centers were monitored in compliance with 21 CFR 812 Good Clinical Practice. Obligations of the principal and other investigators included IRB considerations and reporting, informed consent and assent, device accountability, document recording, filing and retention requirements, reporting requirements, and confidentiality. The data monitoring committee (DMC) was comprised of three internationally recognized,

independent, pediatric orthopedic spine surgeons who helped determine if patients are qualified for the study, provided independent measurements of Cobb angles, reviewed and classified adverse events, and consulted with the sponsor on medical and strategic study goals and decisions.

Potential risks, complications, and anticipated adverse events were defined a priori, and potential risks were minimized by the sponsor's implementation of a risk management program to analyze, monitor, and mitigate risk identified in preclinical testing, during the clinical study, and in the commercial device. The program was based on ISO 14971, International Standards for Medical Device Risk Management. This program utilizes several methods to minimize risks to subjects, including the device design control, surgical implantation procedures, bench and animal testing, investigator training, and clinical protocol design. Specifically with respect to the implant, the construct was designed by the sponsor to mitigate device failure, including breakage, loosening, or migration. For the unlikely occurrence of breakage, the clip was designed so that any remnant would be held in place by the screws and not dislodge into the thoracic cavity.

45.3.5 Study Design for Early Clinical Research

The initial clinical safety trial design was a prospective, single-arm, single-cohort, pilot-phase study. Two sites were approved. The estimated sample size was four to six patients. The objectives were to evaluate the initial acute safety of the device and system and to determine the design and methods of a larger pilot study. Study stopping rules were defined a priori. The endpoint was completion of surgery of at least four cases with documented follow-up of 1 month, absence of any unresolved stopping rule, and data monitoring committee (DMC) recommendation to continue to a larger pilot study. Subjects were scheduled for evaluation at a minimum of eight distinct intervals to 24 months plus additional follow-up until each subject achieved skeletal maturity.

45.3.6 Surgical Technique

To prepare for surgery, the anesthesiologist performs single lung ventilation after bronchoscopic insertion of a double lumen tube and deflation of the lung on the convex side of the child's curve. Positioning of the patient is critically important (Fig. 45.8). The patient is placed on a radiolucent table in a perfect lateral position with the main thoracic curve pointing up. With the goal of providing maximum gravity-assisted thoracic curve correction, a padded axillary roll is placed just distal to the armpit, and a bumper is used to elevate the hip. This allows the chest to hang freely so that it just barely contacts the table. The surgeons and assistants stand anterior to the patient. The use of CO_2 gas insufflation helps to deflate the lung on the operative side and improve the view from the endoscope.

Using a thoracoscopic approach, the segmental vessels are cauterized with a harmonic scalpel to minimize bleeding. The titanium clips with preloaded screws are placed sequentially. Once an acceptable position is confirmed with fluoroscopy, with the clip straddling the disc and ring apophyses, and in the mid-vertebral body in the lateral view, the clip is tamped into place, and the screws are advanced using the screwdriver. The remaining clips are placed in similar fashion, in sequence, across adjacent discs.

A chest tube is placed, after which the deflated lung is expanded again under thoracoscopic visualization. Bronchoscopic lavage and suction are used to clear the lungs of mucus plugs. The opposite, dependent lung in particular is suctioned completely under bronchoscopic visualization prior to extubation to remove mucous plugs that may have drained into the lung during surgery due to the patient's positioning on the lateral side. A chest x-ray is obtained prior to extubation.

45.3.7 Postoperative Protocol

The patient must maintain aggressive pulmonary toilet with an incentive spirometer, coughing, and deep breathing. We have not used a brace or limited non-sports activities. The chest tube is

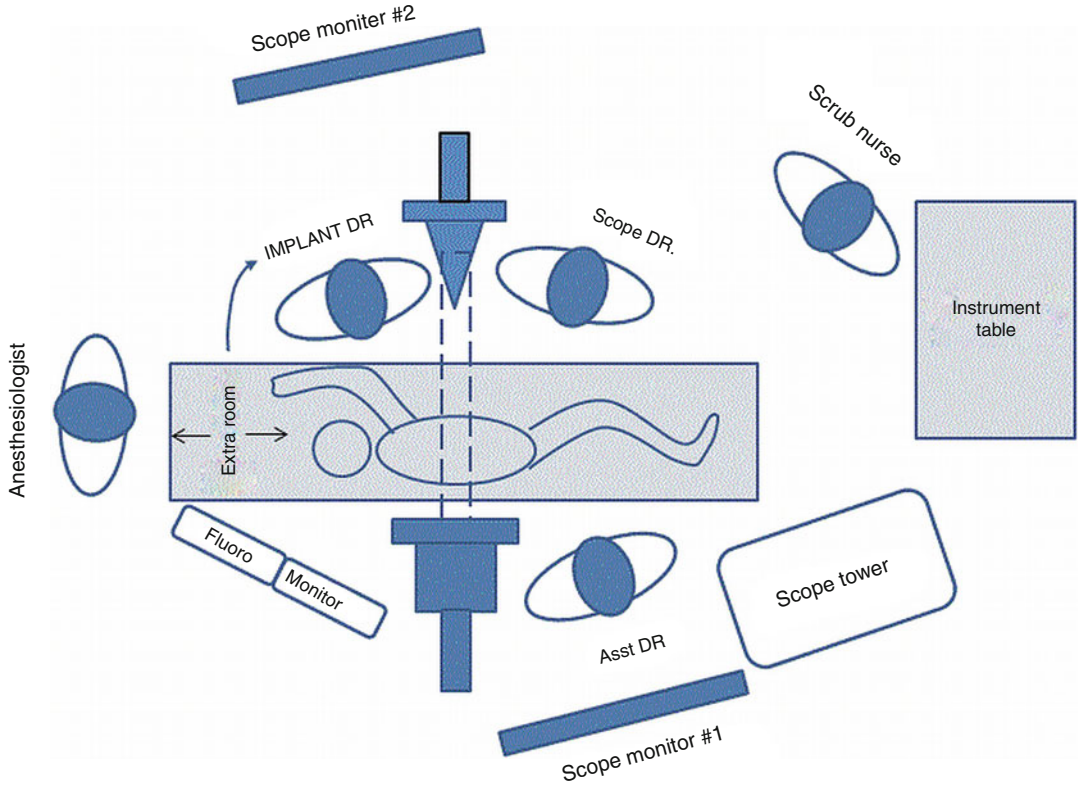

Fig. 45.8 Schematic showing operating room setup

removed when the output is approximately less than 50 ml over an 8 h shift. Team sports are restricted for 6 months, with the exception of cardiovascular conditioning.

45.4 Early Results of Clinical Safety Trial

The first six patients who met the eligibility criteria assented, with parental consent. These six were enrolled as subjects in the study and underwent endoscopic placement of the implants.

45.4.1 Clinical Trial One-Year Results

No device misplacement in spinal canal or disc space, no neuromonitoring changes or neurological deficits, and no device breakage were noted. A procedure-related mucous plug secondary to single lung ventilation in one patient resolved after bedside bronchoscopy. A chylous effusion in one resolved with pigtail catheter and nonfat diet. Mean postoperative hospital stay was <4 days. Surgical implantation time had a mean of 90 min (range, 57–124). Mean blood loss was <75 ml. All curves were right sided. Immediate postoperative curve correction had a mean of 11° (±8°). Mean major curve was 34° (±3°) preoperatively, 28° (±9°) at 6 months, and 30° (±13°) at 1 year. The greatest thoracic curve increase was 14° from preoperative baseline in one patient. The greatest thoracic curve correction was 71 %. In this initial safety study of a clip/screw implant in a small prospective cohort of highly skeletally immature patients at very high risk of progression, blood loss was minimal, surgical times low, and no device misplacement occurred. FDA IDE approval was granted for the next 30 subjects in a pivotal clinical study. Curve changes were variable, but included proof of concept of growth modification in humans by this method.

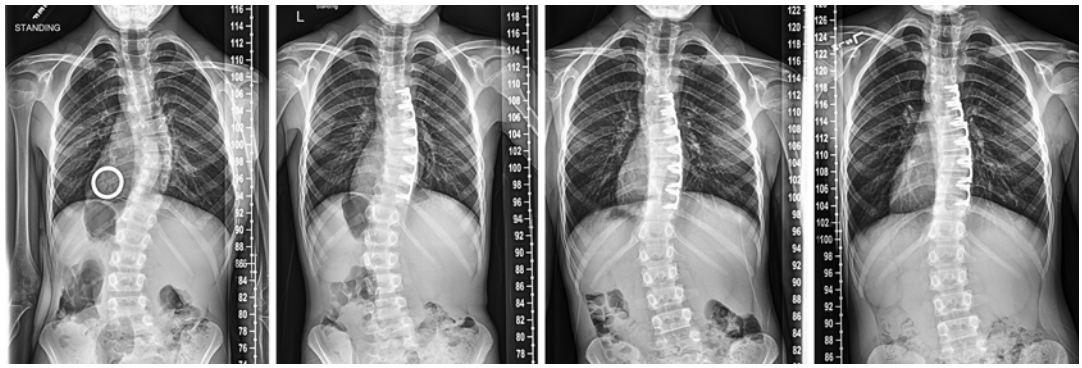

Fig. 45.9 Radiographic time series of most successful subject result to date. *Left*: Immediate preoperative baseline film with calibration *ring*. *Left central*: First postoperative standing radiograph at 1 month PO. *Right central*: 12 months PO. *Right*: 21 months PO

45.4.2 Clinical Trial Results at Latest Follow-Up

At the longest follow-up of all 6 subjects to date, quality of life showed no significant differences between screening and 18 months; SRS-22r mean scores were 4.4 at screening and 4.2 at 18 months (range 3.5–4.7, both at screening.) Radiographic results at 18 months continued to show no mean difference from immediate preoperative Cobb angles, albeit with increasing variation (34° ± 3° *versus* 34.5° ± 16°). The patient with the least flexible curve continued to increase and was fused at 22 months using posterior instrumentation without clip removal. In the mid-range, the mechanism of loss in initial correction was largely migration of the tines in vertebral bone, such that the distance between implants gradually increased. In the most successful case to date, the curve reduction continued to hold, with 67 % correction at 21 months (Fig. 45.9). Between 12 and 18 months, the corrected thoracic curve, coupled with a simultaneously developing compensatory lumbar curve, started to decrease coronal alignment. By 21 months both the lumbar curvature and the misalignment had improved (Fig. 45.10). This time course suggests that the method has the potential to both correct a thoracic curve and to alter the natural history of progression of compensatory curvatures and misalignment. A decrease in the wedge angle of the apical disc appeared to be one of the more significant components of the correction, whereas mean disc heights were not decreasing. In right side bending, the thoracic curve bent out to <5°, indicating at least some continuing curve flexibility at instrumented levels.

In retrospect, the patient whose curve progressed and required spinal fusion was the tallest, had the least flexible curve, and had the flattest sagittal profile. This subject was also among the heaviest, with notable axial rotation and rib hump. Further, this patient was the only one of the cohort in which little to no initial correction was achieved. We would have been delighted to have achieved success in the case. However this case has caused us to reconsider some selection criteria based on the cumulative effect of these factors. By contrast, the best results in this small cohort to date occurred in the subjects of the lowest height and weight and the greatest curve flexibility. Other factors that may be related to variation in results include relative fit between implant and motion segments and aspects of surgical technique. Changes to implant system and processes to improve success rates are planned.

45.5 Technique and Implant Comparison

The clip/screw device technique is a minimally invasive method that many spine surgeons would likely find relatively simple to implant. Patients did not wear a brace postoperatively and returned to school as tolerated. Exercise restrictions were

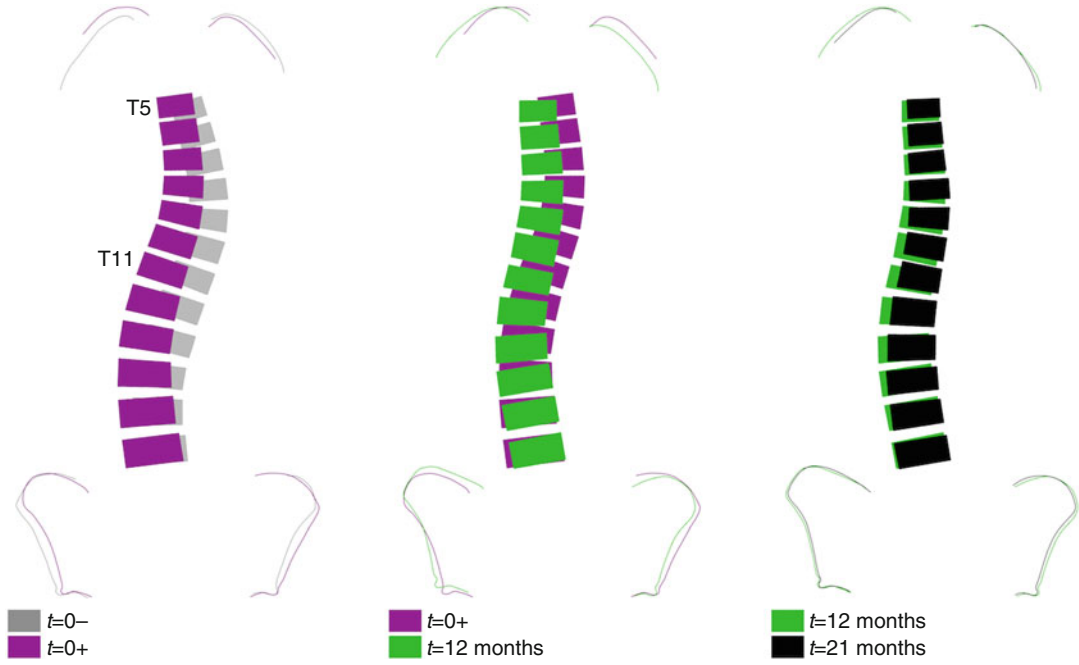

Fig. 45.10 Schematics showing time series of overlays derived from most successful subject results to date. Radiographic images from time points after *t*0-, the immediate pre-operative time, were scaled by ≤12 % to overlay shoulders and hips in order to most directly compare curvature changes

recommended through 8 weeks postoperatively. This is currently the only compression-based growth modulation scoliosis device with European CE mark certification and undergoing a study in the United States of America under a US Food and Drug Administration (FDA) Investigational Device Exemption.

Conclusion

After a history of research dating back to the mid-1900s, a spine growth modulation device is under investigation in a US FDA IDE trial in humans. Radiographic results in this small initial cohort showed that curves in children who had a very high probability of curve progression to >50° did not, on average, progress from baseline values to date. Longitudinal results were variable and have so far included one progression to posterior instrumentation and fusion. Proof of concept in humans, however, was demonstrated in at least one patient whose thoracic curve correction was 71 % at 1 year and 67 % correction at latest follow-up at 21 months. The time course of curvatures and alignment in this case suggests that the method may not only arrest a thoracic curve but also has the potential to correct the main curve and alter the natural history of compensatory mechanisms. Longer-term results and larger cohort are essential to document device performance. If successful, this non-fusion, minimally invasive technique of scoliosis arrest may offer a viable choice for treating late juvenile and early adolescent idiopathic scoliosis while avoiding years of bracing and obviating the need for fusion.

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Apical Control in the Management of Severe Early-Onset Scoliosis

46

Charles E. Johnston

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Key Points

- Apical segment penetration into the convex hemithorax is a primary axial plane deformity which leads to extrinsic thoracic/chest wall dysfunction and thoracic insufficiency syndrome.
- Distraction-based constructs have limited efficacy to improve axial plane deformity.
- Convex apical control methods permit serial rod re-contouring to correct spinal penetration without disturbing concave growth.
- Apical control of thoracic lordoscoliosis is important to control axial plane deformity and pulmonary sequelae.

46.1 Introduction

Progressive thoracic deformity associated with early-onset scoliosis (EOS) has long-term deleterious effects on cardiopulmonary function, including increased mortality rates in untreated patients [3, 6, 15]. The three-dimensional *extrinsic* deformity involving the chest and spine is thought to be largely responsible for impaired pulmonary function, due to the narrowing and rigidity of the convex hemithorax as the axial plane deformity progresses

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and the lack of expandability of the concave hemithorax as the ribs grow progressively more compressed. The axial deformity, sometimes referred to as the "windswept" thorax, is only partially controlled by current distraction-based growing instrumentation techniques [1, 5], due to biomechanical limitations of simple distraction through end-vertebral anchors, and the possibility of "crankshaft" phenomena occurring even though serial distraction is being performed [1, 7] (Fig. 46.1a–e). Should spontaneous ankylosis [4] or ineffective distraction due to the "law of diminishing returns" [16] also become clinically evident, the only available method to correct the axial plane deformity may be definitive fusion, which if performed prior to an adequate thoracic length being achieved is known to produce deleterious pulmonary effects separate from any impairment caused by poorly controlled deformity [9, 11, 17].

Techniques to control apical deformity, and at the same time avoid the serial lengthening procedures required by "traditional" growing rod instrumentation, have been added to the EOS treatment armamentarium. Growth-guidance procedures [12, 14], described in detail elsewhere in this textbook, provide direct control of the apical deformity

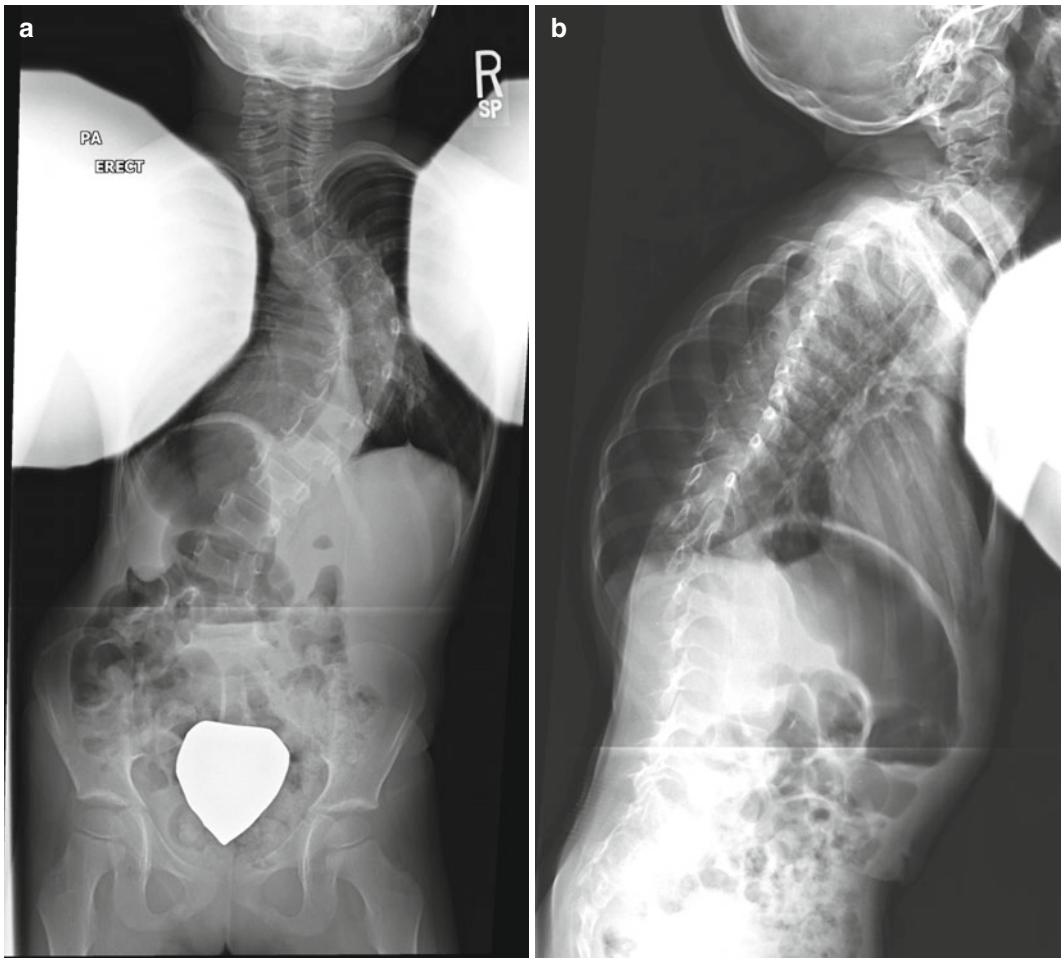

Fig. 46.1 (**a**, **b**) Preoperative radiographs of a 5-year-old female with neglected 75° lordoscoliosis and Marfan syndrome. T1–12 length = 16.5 cm. (**c**) Apical CT of thorax. Significant spine penetration into the convex hemithorax, as well as narrowing of the ventral-dorsal lung space due to lordosis, is seen. (**d**) AP radiograph after 3 years of distraction-based growing rod instrumentation. T1–12 length = 23.6 cm, scoliosis reduced to 46°. (**e**) Apical CT after 3 years of distraction treatment. In spite of significant length gain, there is little change in the spinal penetration, apical rotation, or ventral-dorsal lung space

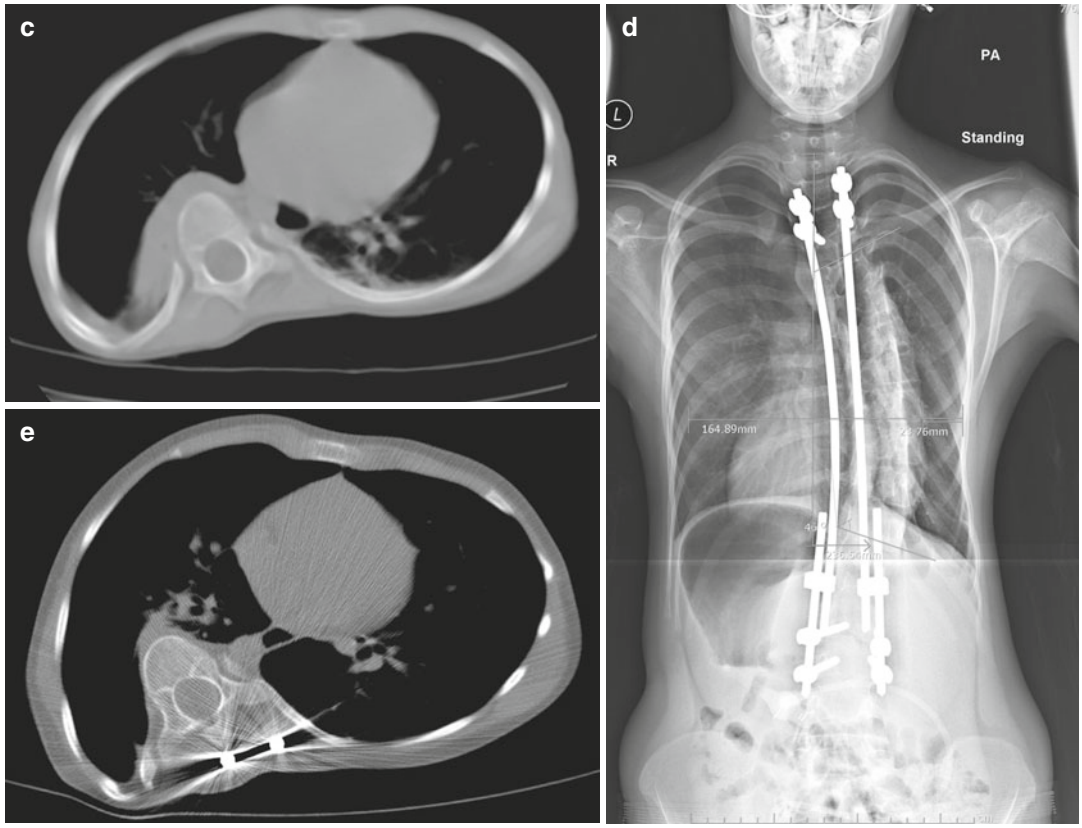

Fig. 46.1 (continued)

by vigorous correction of apical segments, and then by extending instrumentation to the end vertebrae with non-constrained, "sliding" anchors as part of the construct, permit continued spinal growth along a path dictated by the non-constrained longitudinal members of the construct . In the "Shilla" technique, growth occurs outside the fused apical segments, elongating as the spine grows away from the apex (Fig. 46.2a). In the modern "trolley" technique, growth occurs in the non-fused but "guided" apical segments between the end-vertebral anchors which are fused in place (see Fig. 46.2b). Both methods enjoy the theoretical advantage over conventional growing rod instrumentation (GRI) of *not* requiring serial surgical lengthening procedures in order to accomplish deformity control while permitting continued spine growth.

Recent reports [2, 12, 13] have elucidated the limitations of the Shilla method, namely, technical complications requiring frequent unplanned revisions while achieving less length gain and deformity reduction than matched cases treated by conventional GRI procedures. Results of the modern trolley are too limited to make outcome comparisons with conventional methods valid. However, the importance of apical control of EOS deformity has been reaffirmed in a series of cases described in this report, combining the use of apical segment anchors with conventional GRI technique to achieve axial plane correction progressively with elongation of the spine.

46.2 Rationale

The concept of the windswept thorax was originally introduced by Dubousset referring to the penetration of the lordoscoliotic apex into the convex hemithorax [8], narrowing the space for

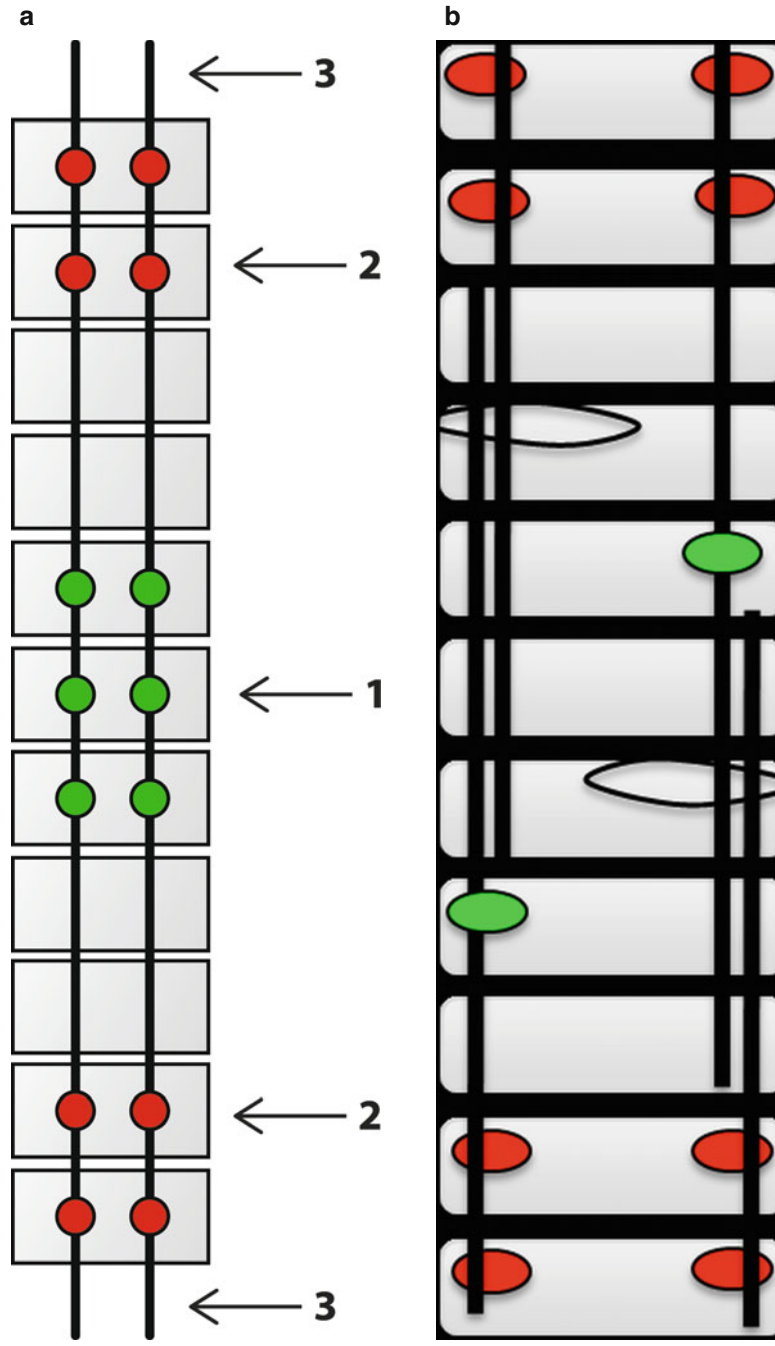

Fig. 46.2 (**a**) Shilla construct. Apical anchors (*1*) are formally locked to the rods after vigorous short-segment deformity correction. End-vertebral anchors (*2*) are "sliding" anchors, captured by the set screws but not locked to the rods, permitting "guided" growth of the non-fused segments within the construct. Extra length of rods (*3*) outside the end vertebrae must be available in order for guided growth to occur (**b**) modern "trolley" construct. End-vertebral anchors (*red*) are fused into place to provide stable anchoring points for the rods. Sliding anchors (*green*) and sublaminar wires which capture the overlapping rods as well as the spine segment control the apical deformity by cantilever correction as well as permit longitudinal growth with the rods sliding apart (**a**: Courtesy R.E. McCarthy, MD; **b**: Courtesy J.A. Ouellet, MD)

lung volume – already narrowed by the rotatory rib deformity – to the most peripheral area between the spine and the chest wall (Fig. 46.1c). In severe cases, the space remaining for convex lung volume is reduced to a narrow sliver as the spine lateral translation into the convex hemithorax squeezes the lung against the chest wall. The term "collapsing parasol" has been used to describe the relative vertical position, or "drooping," of ribs in primarily hypotonic neuromuscular patients [7], but this description can certainly be applied to the convex rib deformity

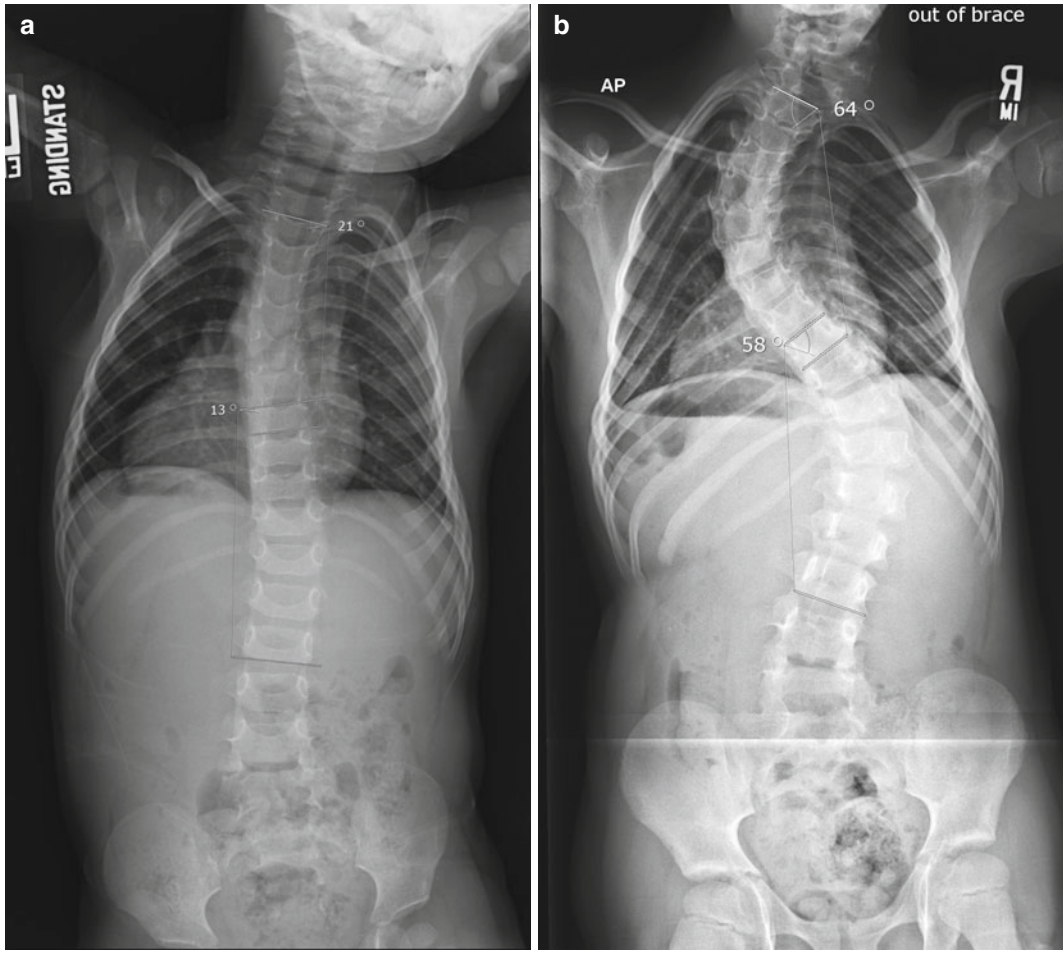

Fig. 46.3 (**a**) A 20-month-old male with syndromic, hypotonic scoliosis. First standing radiograph. Rib morphology is unremarkable. (**b**) Age 7, spine deformity has progressed along with apparent chest wall narrowing and "drooping" of the ribs ("closed parasol"), possibly due to casting and brace treatment

in any form of EOS (Fig. 46.3a, b). Because the anatomic volume of the hemithorax is so diminished by the combined apical rotation and translation, direct control of this portion of the spine is mandatory if more effective corrective measures to reverse the windswept deformity are to be undertaken. The use of apical pedicle screw anchors, for example, placed only on the convexity to allow further concave growth, permits direct derotation *and* translation of the apex toward the concave hemithorax (Fig. 46.4a, b). By fusing only the apical segment to stabilize the convex anchors, and then progressively recorrecting the lateral translation toward the concavity by in situ rod contouring at each subsequent scheduled lengthening, the goal of controlling – indeed improving – deformity while permitting or driving growth, fundamental to the management of EOS, can be achieved [10]. The procedure described here combines progressive apical correction with serial lengthening – the best effects of traditional GRI and the growth-guidance concepts.

Apical control is attained by achieving stable anchors, usually pedicle screws, at two or three apical convex segments. Initial correction of the apex may be achieved by a vetrebrectomy or decancellation in a congenital deformity or by aggressive short-segment fusion following anterior release of the apical segments. The short

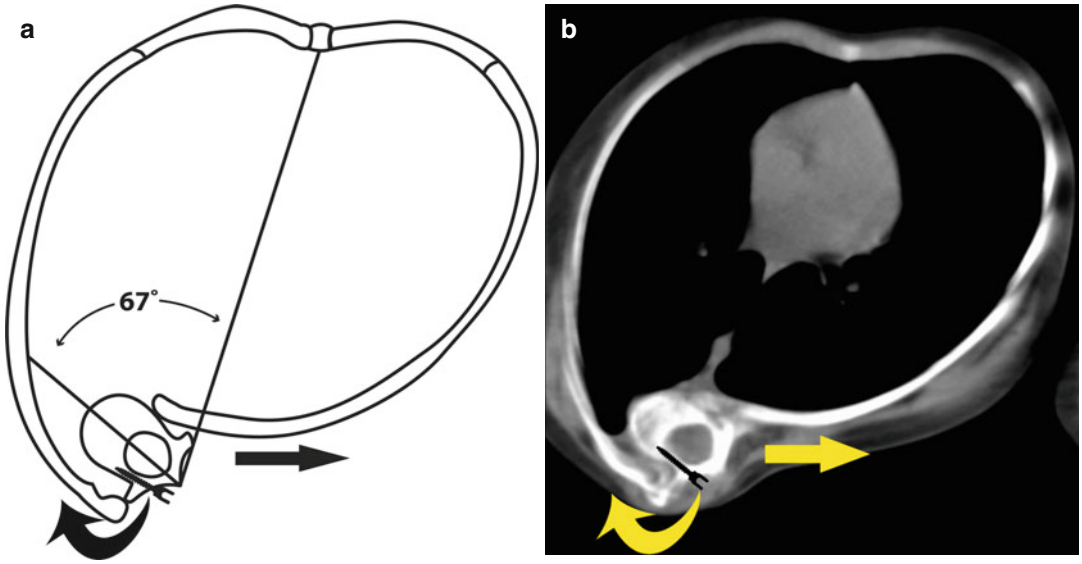

Fig. 46.4 (**a**, **b**) Direct derotation indicates direction of (*curved arrow*) and translation of the apex indicates (*arrow*) is possible by the use of apical convex pedicle anchors. Further correction can be implemented by in situ bending at subsequent scheduled lengthening

instrumented segment is compressed for correction, and then the apical anchors are connected to the end-vertebral anchors of the "standard" growing rod segment, either cephalad or caudal, and then dominoed to the rod connecting to the other end vertebrae. The apical construct is "locked" to achieve initial fusion of this short segment. At the first scheduled lengthening (e.g., 6 months later), the apical construct is exposed, the locking caps loosened or removed, and the apex is further corrected by in situ bending to translate the apex to the concavity. The lengthening procedure is then carried out, with the rod connecting the apical anchors to their respective end vertebra free to slide in the apical anchors.

46.2.1 Case Example

A 6-year-old male with Dubowitz syndrome and short stature had a progressive scoliosis measuring 78° in spite of previous bracing (Fig. 46.5a–c). T1–12 length was 14.5 cm (significantly <3 SD length [7]). Significant spine penetration was seen on axial chest CT [10] (Fig. 46.5d). He underwent anterior apical release/discectomy T8–10 and posterior GRI with apical convex anchors at T9 and 10 with fixed head pedicle screws (see Fig. 46.5e), with immediate correction to 41°. The patient subsequently underwent 6 lengthenings with apical translation toward the concavity by in situ rod contouring, with one anchor revision over the next 4 years. At age 10, the curve had been reduced to 27° and the T1–12 length increased to 17.5 cm. (still <3SD, see Fig. 46.5f). Apical spine penetration, sagittal concave ratio, and axial rotation were all markedly improved (Fig. 46.5g). Two further lengthenings were performed, and at most recent follow-up (age 13 years), the patient has acceptable curve maintenance and a T1–12 length of 20.1 cm, thought to be appropriate for his diminished stature syndrome. He is currently being observed annually.

46.3 Discussion

Currently nine patients with EOS (out of a total of more than 50 operative EOS cases) have been selected for apical control technique based on perceived windswept deformity magnitude. Common indications for this approach included large curves (mean 86°; range, 57–109°), short

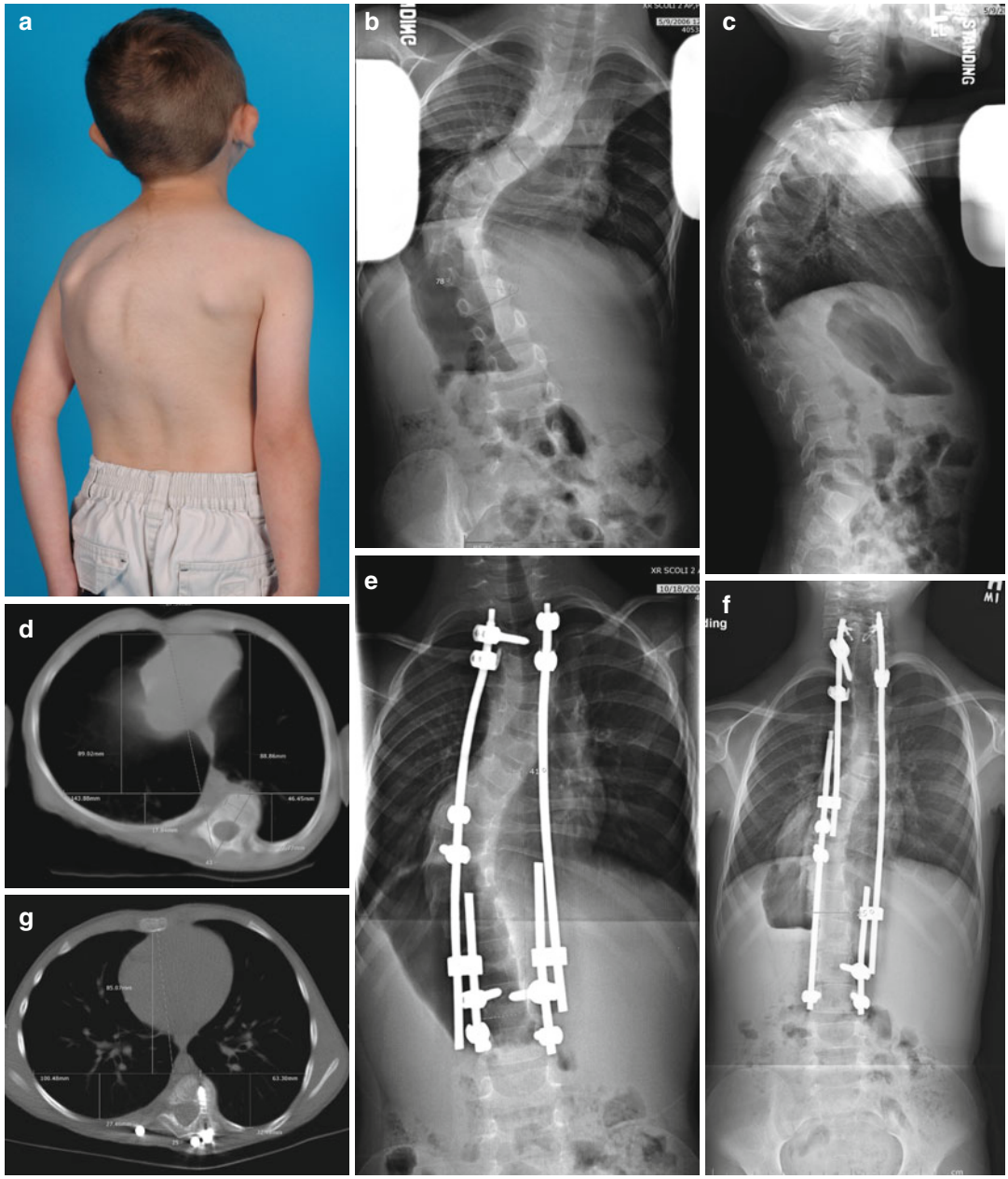

Fig. 46.5 (**a**) Clinical appearance at age 6 years. (**b**) A 78° scoliosis with a short 14.5 cm T1–12 length, significantly <3rd percentile. (**c**) Lateral radiograph at age 6 years. The patient underwent preliminary traction due to the kyphosis. (**d**) Axial CT of chest showing spine penetration into the convex hemithorax. Convex to concave ratio [10] 46.5/143.9 = .32 (normal = 1). Concave anterior-to-posterior ratio [10] 89/17.7 = 5 (normal at T10 = 2.5). Axial rotation = 43°. (**e**) Immediate postoperative radiograph following GRI with apical control. Scoliosis reduced to 41°. (**f**) Age 10, after 4 years of serial lengthening, the curve now measures 27°, and the T1–12 length = 17.5 cm. (<3SD below mean). (**g**) Noticeable improvement in symmetry between the hemithoraces. Axial plane apical penetration is partially corrected to 63.3/100.5 = .63, while sagittal concave penetration is 81.1/27.5 = 3.1. Apical rotation is improved to 25°. (**h**, **i**) Most recent follow-up age 13 + 6. The curve is maintained at 27°, while T1–12 length is now 20.1 cm (still <3SD). Sagittal balance is acceptable

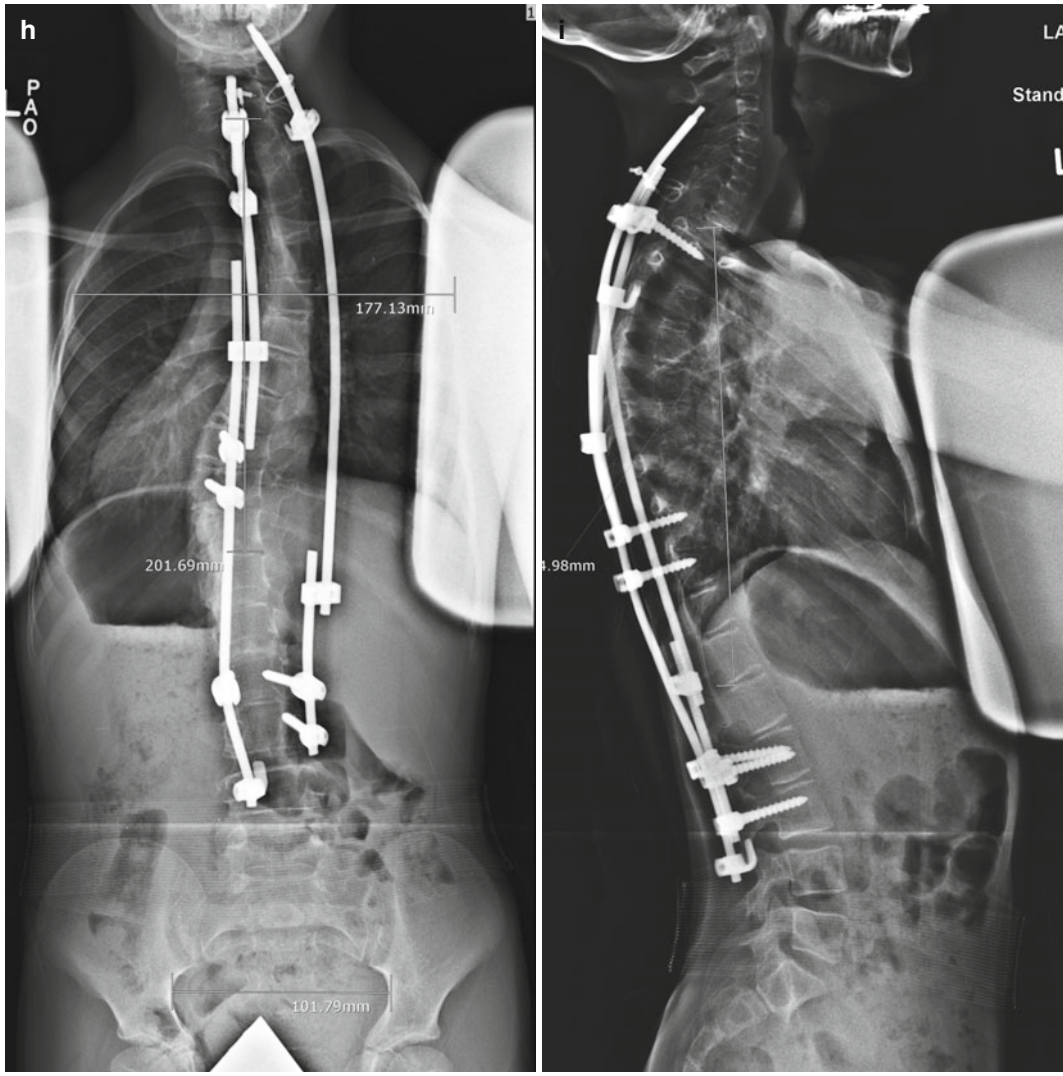

Fig. 46.5 (continued)

T1–12 segments (range, 8.5–14.4 cm) in patients 5 years of age or less. All had significant axial plane deformity, with minimum 40° apical rotation (see Figs. 46.4 and 46.5d). In three patients with greater than 5 years follow-up, correction of apical windswept deformity (rotation and convex penetration) has been effective and worthwhile in restoring symmetry between hemithoraces (compare Fig. 46.5b with Fig. 46.5h). Apical control was ineffective in two patients and abandoned early (within 2 years of initiation) due to apical anchor failure in one patient with osteogenesis imperfecta and to spontaneous ankylosis in one patient with Larsen syndrome. In the remainder, there have been no complications or unscheduled returns to surgery, and thoracic parameters and CT- or MR-calculated thoracic volumes have increased 50 % or greater [10]. Within this small series from a single institution, one can find clear evidence from imaging studies that the ability to control or correct the apical deformity in these more severe cases appears to be a valuable addition to the EOS management armamentarium.

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Magnetically Controlled Growing Rods (MCGR)

47

Behrooz A. Akbarnia and Navid Arandi

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Key Points

- Multiple surgeries and younger patients have higher risk for complications.
- Indications and surgical technique for MCGR is similar to TGR; however, it requires attention to some details.
- Contouring properly, location of actuator, and direction of the rod (s) are important factors.
- There are fewer planned surgeries for lengthening; however, as in TGR, re-operations still occur.
- There would be more frequent lengthenings possible but the optimal interval still unclear.
- There is evidence that, in the long term, the procedure is economically feasible.
- Ultrasound is promising to reduce the radiation exposure.
- Improvement of pulmonary function and quality of life should be the primary goal.
- New technology has certainly helped to improve the quality of care for these children but more research and development needed to define meaningful practice guidelines.

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47.1 Introduction

Traditional growing rod (TGR) surgery is a commonly used technique in the treatment of progressive EOS. TGR requires periodic surgical lengthening typically every 6 months – to maintain curve correction and allow continued spinal growth [1]. Recent studies on TGR have shown that frequent surgical lengthening significantly increases the risk of complications [2, 3].

Magnetically controlled growing rod (MCGR) has been developed with the aim to reduce the number of planned open surgical procedures and to lessen the burden of repeated surgeries for EOS patients while maintaining the benefits of distraction and curve control. Preclinical and early clinical results on MCGR surgery have been promising [4, 5].

47.2 MAGEC Device

The only MCGR device currently available in the USA is MAGEC® (Ellipse Technologies, Inc., Irvine, California, USA). This device has been available outside of the USA since 2009. However, the device received 510(k) clearance from the US Food and Drug Administration (FDA) in February of 2014, and at the time of this writing, more than 1500 procedures have been performed worldwide with over 300 in the USA.

The MAGEC System includes an implantable rod, a MAGEC Manual Distractor (MMD), the MAGEC Magnet Locator (MML), and the External Remote Controller (ERC) (Fig. 47.1). The implantable rod includes an adjustable actuator portion with an enclosed magnet. The ERC includes two larger magnets that can be rotated when the physician activates the ERC. The implant can be lengthened through a magnetic coupling between the enclosed magnet of the adjustable actuator portion and the magnets of the ERC. When the ERC is activated in proximity to the implanted magnet, the rotation of the ERC magnets causes the implanted magnet to also rotate. The mechanism within the actuator converts this rotational motion to linear motion, and the length of the implant changes. The implantable rod is offered in two different actuator lengths, a 90 mm actuator and a 70 mm actuator. The 90 mm and 70 mm actuator bodies allow for a total of 48 mm and 28 mm of distraction, respectively. The actuator portion of the MAGEC rod cannot be contoured. The 70 mm actuator body allows for a longer overall portion of the implantable rod that may be contoured as compared to the 90 mm actuator body. The 70 mm actuator may be chosen for specific anatomical considerations or in smaller stature children who might not be able to accommodate the longer 90 mm actuator body.

In this chapter, we will focus on the indications, surgical technique, and early results of this procedure and compare the clinical outcomes of the MCGR and TGR techniques for the treatment of EOS.

47.3 Indications

The device has FDA clearance for the treatment of progressive EOS in immature patients with or at risk of thoracic insufficiency syndrome (TIS).

47.4 Contraindications

- Patients with infections or pathologic conditions of the bone which could impair the ability to securely fix the device (e.g., osteoporosis, osteopenia)
- Patients with metal allergies and sensitivities to the implant materials (e.g., titanium).
- Patient with a pacemaker or other active, electronic devices (e.g., implantable cardioverter defibrillator)
- Patient requiring MR imaging during the expected period of device implantation
- Patients younger than 2 years old
- Patients weighing less than 25 lbs (11.4 kg)
- Patients and/or families unwilling or incapable of following postoperative care instructions
- Patients with stainless steel wires or other implants containing incompatible materials

Fig. 47.1 MAGEC System: (**a**) MAGEC rods (*1*) standard and (*2*) offset (**b**) MAGEC Manual Distractor (MMD) (**c**) Magnet Locator (MML) and (**d**) External Remote Controller (ERC model 1) (Courtesy of Behrooz A. Akbarnia, MD)

47.5 Surgical Technique

The technical principles of this procedure are similar to those of traditional growing rod (TGR) described elsewhere in this book. Following the proper patient positioning and preparation of the surgical site, the selected levels of foundations are approached through one or two midline incisions, and anchors are implanted using screws, hooks, bands, or a combination, per patient's needs and the surgeon's choice over two or more levels if needed. Usually, hooks and/or pedicle screws are used as proximal anchors and screws for the distal foundation. The selection of the levels and preparation of the foundations is similar to that of TGR [6] (see Chap. 38).

47.5.1 Rod Preparation and Contouring

A template (e.g., a typical anesthesia stylet) can be used to determine the length of the rod with consideration of extra length based on anticipated intraoperative correction and distraction. Preoperative flexibility radiographs can help in planning the location of the actuator, size, and length of the rods. The concave rod is usually

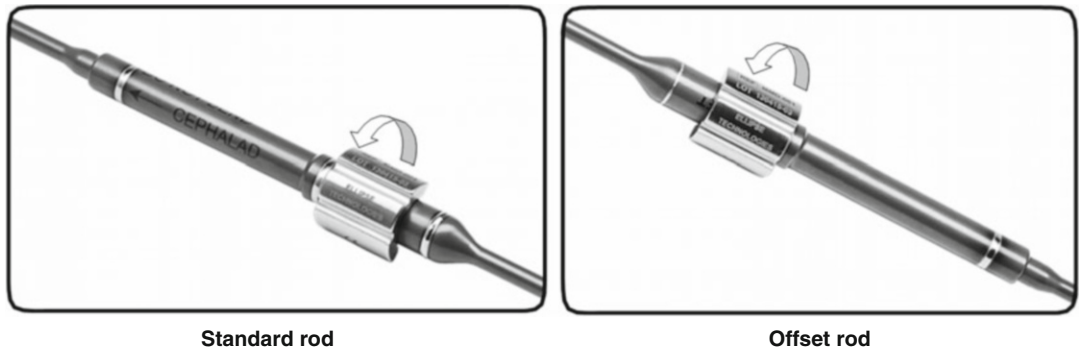


Fig. 47.2 Distraction and testing by MMD. Note the counterclockwise direction of the MMD (Courtesy of Behrooz A. Akbarnia, MD)

prepared first. It is cut and contoured to fit the desired sagittal alignment. Some surgeons apply a temporary convex rod first to maintain the distraction and more accurately estimate the rod length. Care should be taken to avoid any bending of the actuator or within 20 mm of it. For extensive contouring or for smaller stature children, the 70 mm actuator may be an alternative to provide more rod length available for bending.

47.5.2 Rod Testing

It is very important to verify that the rod distracts correctly after contouring and prior to implantation. The MAGEC® Manual Distractor (MMD) will self-align over the zone marked "MAGNET." Mark the rod where it first exits the actuator with a sterile marker to aid in visualizing movement of the rod. Next, slide the MMD over the implant zone marked with the letters "MAGNET" while maintaining standard sterile technique. Rotate the MMD by hand about the centerline axis of the actuator counterclockwise when viewed from the distal end of the implant with the arrow pointed up (cephalad). This will cause the implant to distract (lengthen) (Fig. 47.2). It is recommended that four full counterclockwise rotations are performed to ensure the rod is properly functioning. After confirmation, three full clockwise rotations should be done to return the rod almost to its neutral position and avoid jamming.

47.5.3 Rod Orientation

When using dual rods, a combination of two single standard rods or a single standard rod and an offset rod combination can be used. Standard rods may be chosen if the surgeon's preference is to distract the rods at the same time in the same direction. A standard and an offset rod combination may be chosen if the surgeon's preference is to distract the rods individually and in opposite directions since the magnets are located at opposite ends of the actuator portion. Regardless of the type of the rod, it has been suggested that they are placed with the actuators at the same level for the best function (Fig. 47.3). Anatomically, the best location for the rods is the thoracolumbar junction, but it could be placed in other levels with consideration of sagittal alignment.

47.5.4 Rod Insertion

A standard chest tube may be used to tunnel the rod subfascially between the two foundations. Then, preliminarily attach the rods to the anchor sites at first proximally and then distally. Insert the concave rod first for single curves. Tunneling the rod can be assisted by first passing a chest tube subfascially and then use the same tunnel for passing the rod or using a long clamp (Fig. 47.4). The second rod is

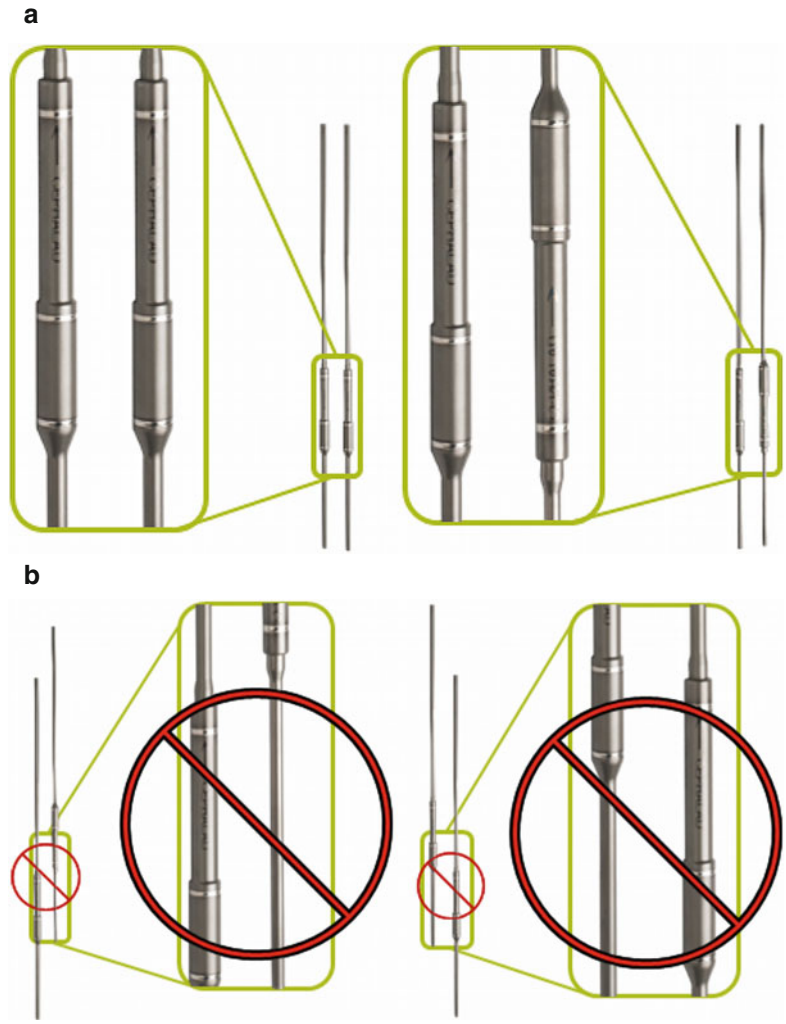


Fig. 47.3 Implant location in dual-rod technique: actuators are placed at the same level for the best function. Note (**a**) correct and (**b**) incorrect positions (Courtesy of Ellipse Technologies, Inc.)

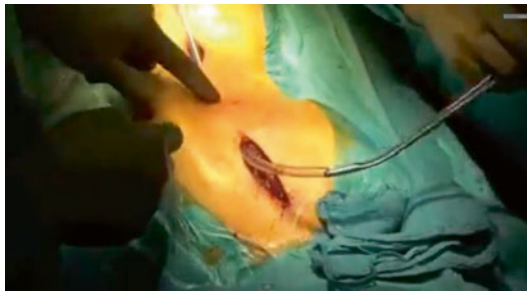


Fig. 47.4 Passing the rod subfascial: the rod is being introduced using a chest tube (Courtesy of Behrooz A. Akbarnia, MD)

then cut, contoured, and passed in the same manner. Some surgeons prefer to use a temporary rod on the convex side and then use the permanent rod on the concave side for more accurate rod measurement. Both rods are then attached to the upper foundation first making sure that they are in proper sagittal orientation. The entire upper foundation anchors are preliminarily tightened, and a cross-connector is attached between two rods if needed to complete the upper foundation construct. The lower foundation is then constructed, where both rods are attached to the lower anchors loosely and the concave rod followed by the convex rod is distracted. No cross-link is applied distally unless hook anchors are used. After radiographic check, the wounds are irrigated, foundations are prepared for fusion, bone graft is placed, and wounds are closed.

47.6 Lengthenings

47.6.1 Method of Lengthening

There are two methods of distraction, incremental mode and continuous mode. The External Remote Controller (ERC) contains two large magnets that are rotated when the ERC is activated. The magnets of the ERC couple to the magnet in implanted actuator portion of the rod. When the ERC magnets rotate, the coupling causes the implanted magnet to rotate, and the implanted rod to change in length. If the first method of distraction is selected, the ERC is placed in the "incremental" mode and the desired distraction amount is entered in the control panel. The patient is placed in the prone position on the examining table. The MAGEC Magnet Locator (MML) is held vertically near the distal end of the device at the location of the actuator (Fig. 47.5a, b). It is place just above the skin surface of the patient that approximates the location of the internal magnet and allows the locator to be pulled to the strongest point of attraction. The locator will naturally attract itself to the internal magnet. Mark the point on the skin and position the implant-locating window of the ERC over the magnet area that is marked. The ERC should be oriented along the axis of the implant with its orientation arrow pointing towards the patient's head (Fig. 47.5c). Lastly

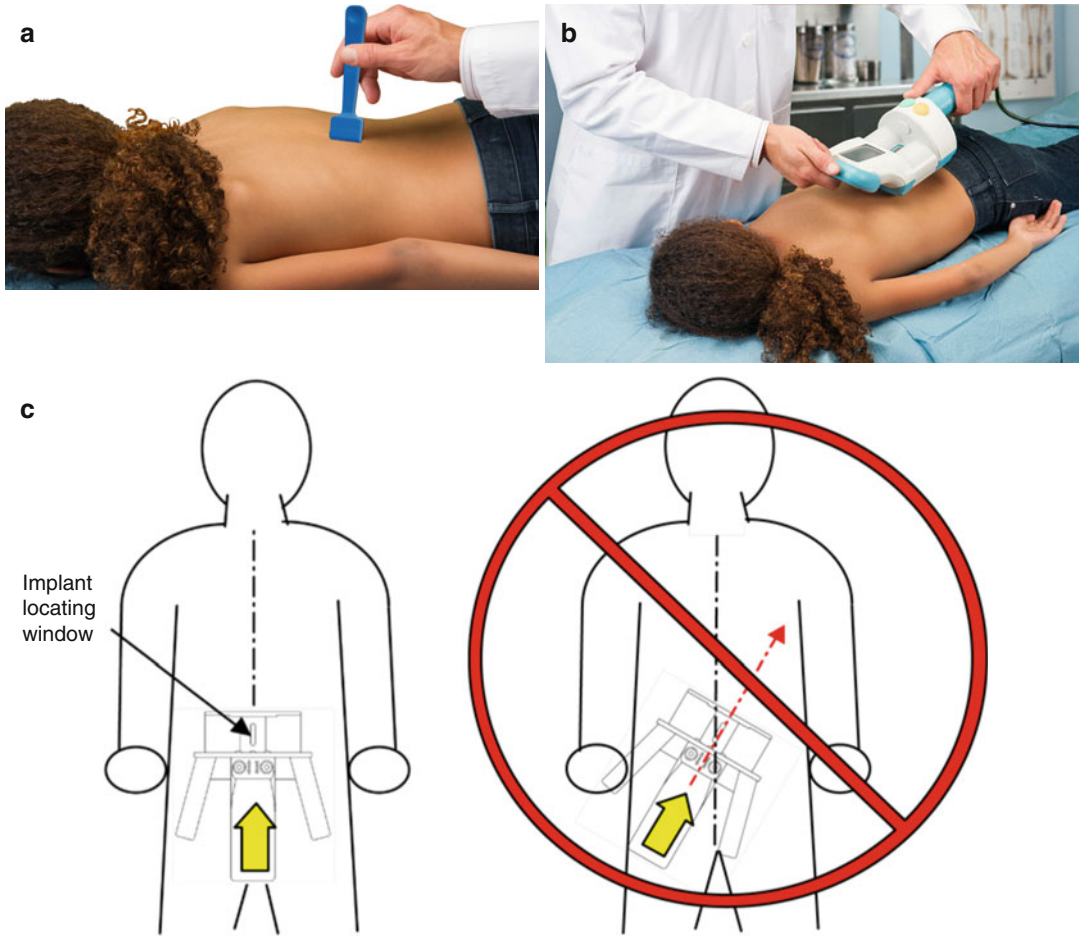


Fig. 47.5 Lengthening: (**a**) MML skin placement on the site of magnet. (**b**) ERC 2 was used for distraction. (**c**) Correct placement of ERC in relation to the magnet (**a**, **b**: Courtesy of Behrooz A. Akbarnia, MD; **c**: Courtesy of Ellipse Technologies, Inc.)

the "on" button is pushed and the device is lengthened as the ERC is activated. Since the amount of the lengthening has already been entered, the ERC will stop when the distraction is complete.

Alternatively, continuous mode can be used for distraction. The ERC is placed in continuous mode, and distraction is done as the previous method until the device stalls. Distraction is then stopped. This indicates that the maximum distraction force of the implant has been reached.

47.6.2 Frequency of Lengthenings

The lengthenings in MCGR can be accomplished more frequently than in TGR since MCGR distractions are noninvasive and lengthening can be performed in the clinic. The ideal interval is not yet known, but finite element analysis (FEA) studies have suggested that more frequent lengthenings may put less stress on the rod and possibly reduce rod fractures [7]. Cheung et al. in a study of 30 patients with minimum of 2 years follow-up has also shown that more frequent lengthenings (1 week to 2 months) is associated with less implant complications but increased incidence of proximal junctional kyphosis and difficulty in lengthening compared to less frequent lengthenings (3 months and longer) [8].

The authors' current practice is to obtain radiographs prior to each lengthening and to confirm desired lengthening was achieved with ultrasound study post lengthening (Fig. 47.6). Ultrasound has been reported as a valuable and accurate tool for demonstration of lengthening, therefore reducing ionizing radiation exposure [9].

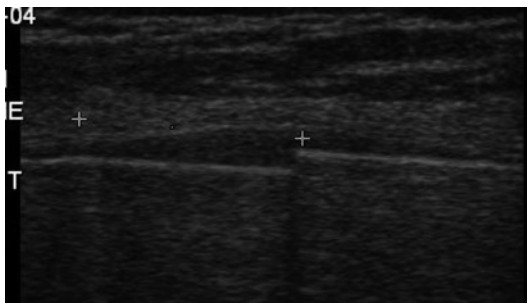


Fig. 47.6 Ultrasound examination during lengthening: accurate readings can be obtained by using noninvasive ultrasound method (Courtesy of Behrooz A. Akbarnia, MD)

47.7 Complications

Complications could be the result of MCGR rod or due to the nature of patients with EOS and underlying etiology. In the first report by Cheung et al., five patients were implanted with MCGR, two of which had a 24-month follow-up. One patient showed a loss of distraction related to the MCGR device, which resulted in the exchange of the device and the correction of the spinal height and the major curve [10].

In a recent multicenter retrospective study of 54 patients with 24 patients having 2 years of follow-up, complications were categorized into wound related and implant related as well as early (<6 months) and late (>6 months). Twenty-one patients of 54 had at least one complication. Fifteen had a revision surgery. Six had rod fractures (two 4.5 and four 5.5 mm rods); two 5.5 mm rods failed early (4 months) and 4 late (mean = 14.5 months). Six experienced 1 episode of lack or loss of lengthening of which 4 lengthened subsequently. Seven had either proximal or distal fixation-related complication at an average of 8.4 months. Two patients had infections requiring I & D, one early (2 weeks) with wound drainage and one late (8 months). The late case required explantation of one of the dual rods [11].

In another retrospective multicenter study, Cheung et al. reported a rate of reoperation of 42 % in a group of 26 EOS patients. This study highlights the inherent risks of unplanned operations in EOS patients. Further studies with MCGR with longer follow-up are required to assess the outcomes of this technique [12].

47.8 Discussion

The idea of remotely controlled distraction-based systems as well as the use of a magnet is not new. Takaso et al. in 1998 were the first to use a remote-controlled growing rod spinal instrumentation in a scoliotic canine model. Mean major

curve improved from 25° to 3° after 4 distractions over a span of 12 weeks. The authors were able to show the efficacy of this magnetic system in spinal distraction and curve correction [13].

Soubeiran, Miladi, and Dubousset developed the Phenix device. Albeit there is very limited data, all authors reported on the use of this magnetically expandable GR for distraction between ribs, vertebrae, and pelvis [14–16].

In 2012, Akbarnia et al. reported on the use of a new magnetic spinal device in an animal study to assess its safety and efficacy [17]. The authors were able to demonstrate that MCGR provided 80 % of predicted spinal height via remote distractions in the porcine model. No MCGR-related complications were reported.

Cheung et al. published the first preliminary study of MCGR surgery [10]. The authors describe five patients, two with 2 years follow-up who had syndromic scoliosis and significant major curve correction (overall mean, 57 % correction) and acceptable gain in T1–S1 spinal height (overall mean, 46 mm) and T1–T12 thoracic height (overall mean, 30 mm) after a 2-year course of monthly noninvasive lengthenings. One of the patients experienced loss of distraction that ultimately resulted in a surgical revision to exchange the device and restore curve correction and spinal height. The same patient also experienced a superficial surgical site infection (SSI) that was treated with medical management. The report also described a presumed economic benefit with the use of the MCGR, citing the high cost of repeated surgical lengthenings.

In a separate study, Akbarnia et al. reported their results of 14 MCGR patients with a mean follow-up of 10 months (range, 6–18 months) [5]. At the latest follow-up, the major curve corrected an average of 48 % and spinal height increased an average of 9 mm for single-rod constructs and 20 mm for dual-rod constructs. Complications included superficial SSI and prominent implants. There was a partial loss of distraction noted in 14 of the 68 noninvasive lengthenings for the entire cohort.

In 2013, Dannawi et al. reported a series of 34 patients with minimum 1-year follow-up [18]. Mean major curve correction was 41 % and overall gain in spinal height was 44 mm. There were two patients with rod breakages, two patients with superficial SSI, two patients with loss of distraction, one patient with a hook pullout, and one patient with prominent implants.

Most recently, Hickey et al. [19] studied clinical and radiographic outcomes in eight MCGR patients with minimum 23-month follow-up. Major curve correction averaged 43 % for primary MCGR patients and 2 % for patients who were converted from TGR to MCGR. Annual spinal growth was 6 mm/year for primary MCGR patients. Typical complications associated with growing rod surgery occurred in this study included anchor failure and rod breakage.

In the most recent publication [20], MCGR patients were compared with a matched cohort of TGR patients from a multicenter EOS database. MCGR patients who were followed for a minimum of 2 years had similar overall percent major curve correction at latest follow-up: 32 % versus 31 %, respectively. The corrective techniques used to reduce the scoliosis are essentially the same between MCGR and TGR. It is worth noting the MCGR is limited by the amount of sagittal contour that can be achieved due to the current length and the position of the magnetic actuator. The smaller actuator (70 mm) in MAGEC device may allow more rods for contouring with less effect on sagittal alignment.

MCGR patients had a greater T1–S1 and T1–T12 lengths than TGR patients prior to initial surgery, but these modest differences evened out immediately after surgery. After at least two years of lengthenings, TGR patients ended up with a greater mean T1–S1 (342 mm) and T1–T12 (210 mm) compared to MCGR patients (307 mm and 189 mm, respectively); however, TGR patients had an average 1.6 years longer follow-up than their MCGR-matched cohorts. In an attempt to control for the significantly different lengths of follow-up between treatment groups, annual T1–S1 growth and annual T1–T12 growth were calculated. Annual T1–S1 and annual T1–T12 were found to be similar between MCGR and TGR patients suggesting both techniques provided comparable gains in spinal and thoracic height per year during the lengthening period (Fig. 47.7).

Differences in spinal and thoracic height between the groups at latest follow-up bring into question several technical differences between the two procedures. The amount of distraction applied at the initial surgery and at each lengthening are important factors that are difficult to quantify in a retrospective study. Additionally, we expect the time interval between lengthenings to have an effect on changes in spinal and thoracic height. Since MCGR lengthenings can be performed in a clinic setting, the lengthening intervals are often much shorter than the 6-month intervals used typically in TGR surgery.

All MCGR patients in our study received the first-generation device. In an attempt to address the reports of loss of distraction as seen in other MCGR studies, a second-generation MCGR device has been developed and is currently being used. Furthermore, due to the novelty of the MCGR device, there is likely a learning curve effect at play as many of the MCGR patients were among the earliest cases ever performed.

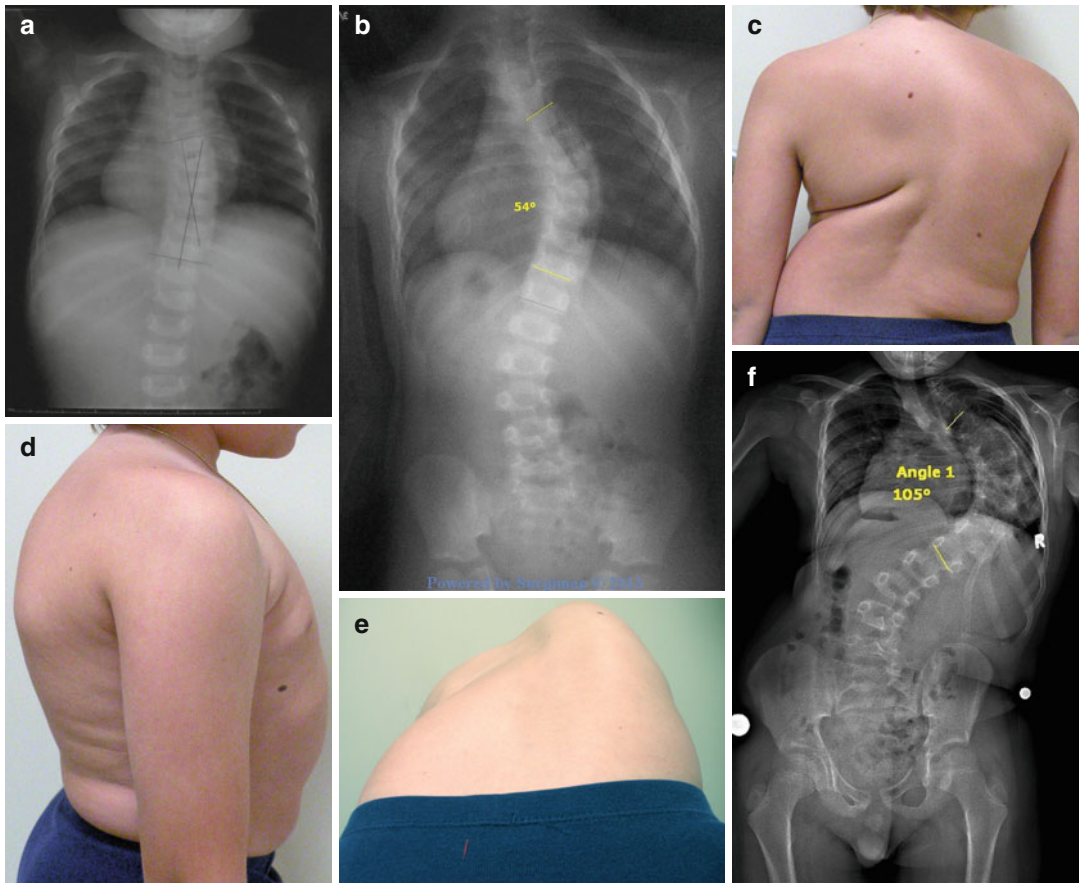


Fig. 47.7 An 8-year-old boy with the history of idiopathic EOS diagnosed at the age of 9 months when he had a 20° curve. He was observed but his curve progressed to 54°, at the age of 3 years, when he was started with orthotic treatment. (**a**) Posteroanterior radiograph at the age of 9 months showing a 20° curve. (**b**) Progression to a 54° curve at the age of 3 years. (**c**, **d**) Preoperative clinical photographs. (**e**) The patient exhibited a right-sided thoracic rotational prominence measuring 30°. (**f**, **g**) PA and lateral radiographs demonstrating a right thoracic curve from T5 to L1 measuring 105°, with a maximum thoracic kyphosis and lumbar lordosis of 77° and −69°, respectively. (**h**) Push prone flexibility radiographs showed a 44 % curve correction. (**i**, **j**) Immediately following insertion of MCGR, the major curve improved to 67° and patient had satisfactory sagittal alignment. (**k**, **l**) Postoperative clinical photographs. (**m**) Showing distraction obtained (*arrow*). (**m**, **n**) Follow-up PA and lateral radiographs demonstrating maintenance of the major curve at 65° as well as maintenance of sagittal alignment (Courtesy of Behrooz A. Akbarnia, MD)

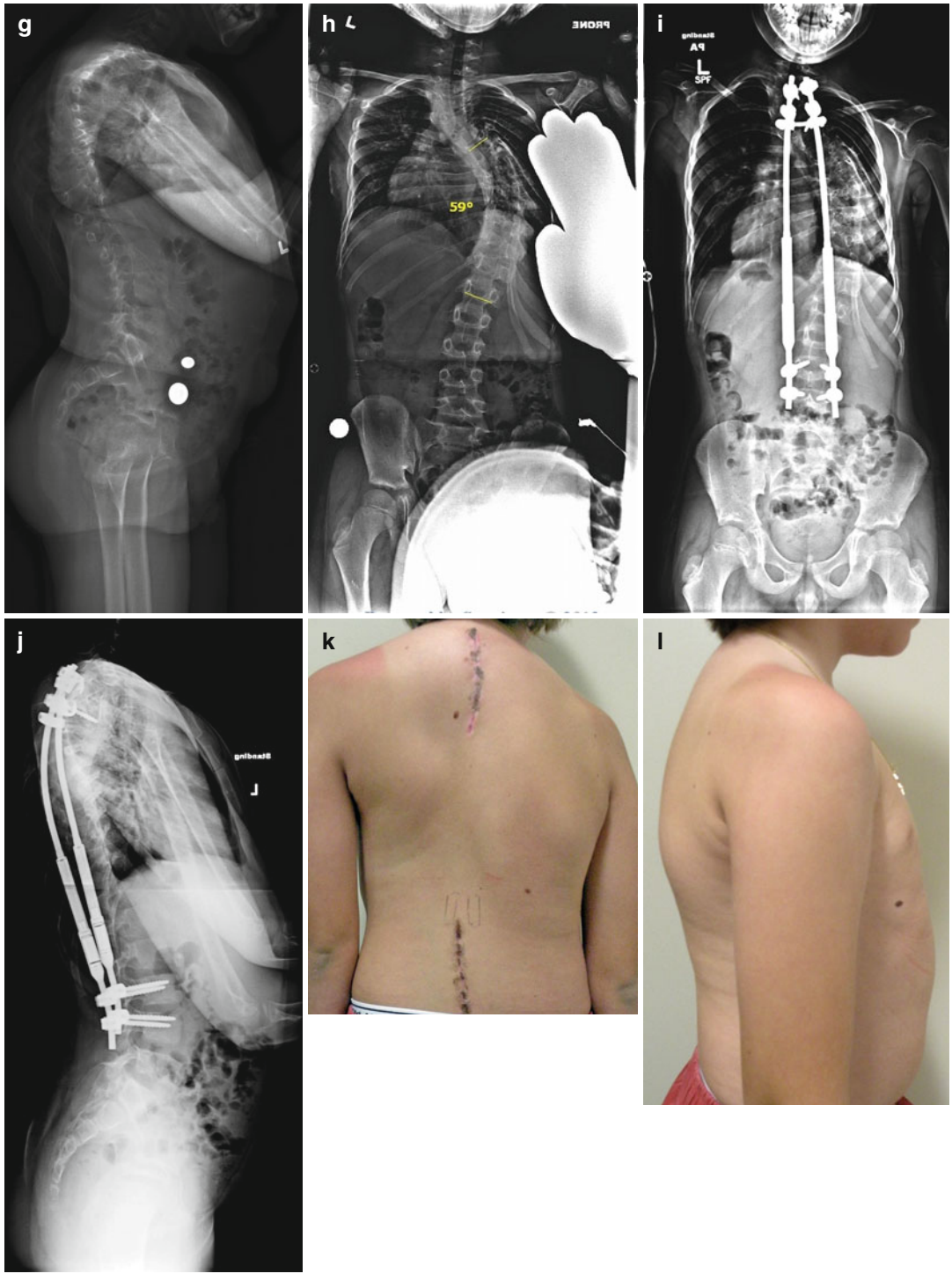
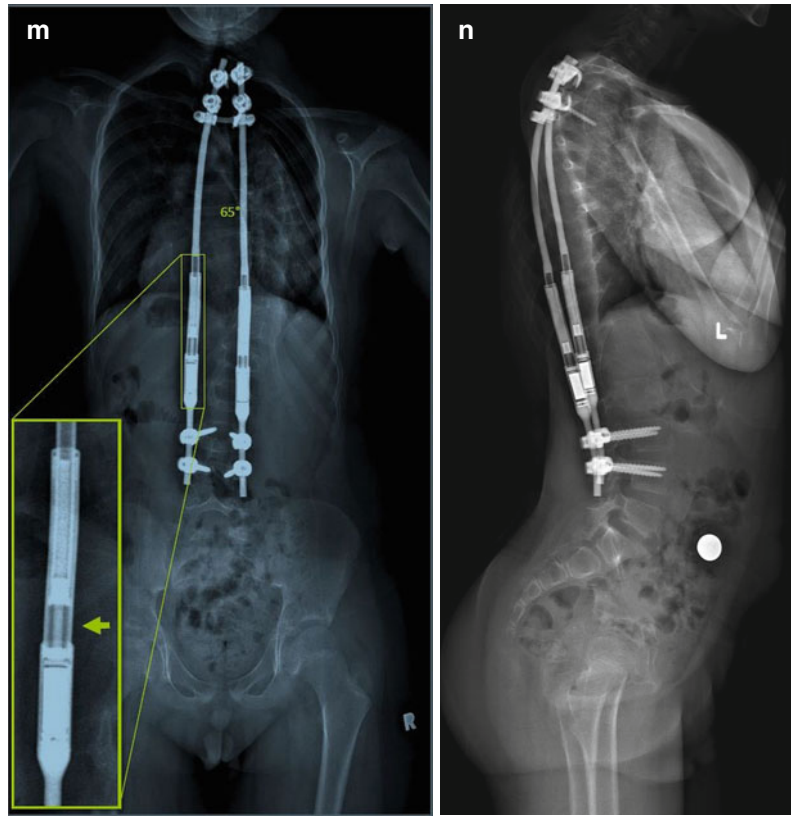


Fig. 47.7 (continued)

Fig. 47.7 (continued)



We expect surgeon variability to diminish over time once consensus is developed and practice guidelines are established.

The majority of complications for both groups were implant related. Loss of distraction made up the majority (63 %) of the MCGR complications. One of the three patients who experienced loss of distraction required revision surgery. Anchor pullout and rod breakage were the most common TGR implant-related complications. TGR patients also had longer follow-up compared to MCGR patients and thus had more time to experience postoperative complications. The majority of implant-related complications in TGR patients (10 of 13) were treated during routine surgical lengthenings.

Due to the retrospective nature of this study, there are limitations. This study included patients from multiple centers; therefore, the variability in surgical technique, postoperative care, and lengthening regimen introduced several confounding variables that were not accounted for in analysis. Patients were not matched by curve pattern or levels of instrumentation, and these were not factored into the radiographic analysis. Furthermore, growing rod surgery typically requires several years of lengthenings until patients reach skeletal maturity. Therefore, there may be clinically relevant differences between the groups that may not be evident within the 2-year follow-up period. Additional follow-up until skeletal maturity will be required to truly determine if differences exist between these two surgical techniques.

In summary in this small yet carefully case-matched series of MCGR and TGR patients, there was no statistically significant difference in major curve correction or gain in spinal and thoracic height after 2 years of treatment. Both groups experienced similar rates of implant-related complications that necessitated unplanned revision surgery; however, MCGR patients were subjected to significantly less open surgical procedures thus reducing the burden of repeated surgery associated with the TGR technique. While the 2-year results of MCGR compared to TGR is

encouraging, this study emphasizes the need to develop MCGR practice guidelines in order to achieve optimal and reproducible results with the use of this novel technique.

Conclusion

The MCGR procedure is a safe and effective distraction-based growing rod technique with similar results in curve correction and spinal growth compared to traditional growing rods while allowing noninvasive lengthenings and significantly reducing the number of surgeries associated with this treatment. It can be safely and effectively used in outpatient settings, minimizing surgical scarring and psychological distress that is associated with multiple surgeries. It is hoped this technique will improve the quality of life of children undergoing growth friendly surgical treatments and their families. It is also shown that the procedure is cost-effective in the long run, in comparison with traditional growing rod surgery [21].

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Distraction-Based Growth Friendly Implants with Rib Anchors

48

Lindsay Andras and David L. Skaggs

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Key Points

- Distraction-based growth friendly implants with rib anchors (hybrid) constructs are useful in the management of thoracic and cervicothoracic spinal deformities.
- The use of rib anchors avoids intentional fusion of the upper thoracic spine, which is important for pulmonary development.
- As ribs are mobile, a theoretical benefit of rib anchors is motion preservation, as opposed to spontaneous autofusion observed frequently after extended treatment with rigid standard growing rod constructs.
- Hooks from standard spinal instrumentation systems can be used for this technique.
- In most cases, a thoracotomy is not needed.
- Neuromonitoring of the upper extremities is imperative when distracting on rib anchors and positioning the arms in adduction is advised if there is particular concern for brachial plexus injury.

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48.1 Introduction

Our specialty and patients are indebted to Dr. Robert Campbell (Fig. 48.1) for pioneering treatment of the growing child with thoracic and spinal deformity. He not only first described thoracic insufficiency syndrome [1] but also first described the treatment of this condition with thoracic expansion [2]. A unique feature of his treatment was attachment of distraction-based implants on the ribs. The use of traditional spine hooks on the ribs is considered "off-label" by the Food and Drug Administration of the United States. Whether this is of any significance is questionable, as off-label usage of spine implants in pediatrics is commonplace. While this technique has gained popularity, the indications for rib-based proximal anchors compared to spine-based anchors were recognized as an area of the greatest uncertainty in treatment of early onset scoliosis in a recent survey of 14 experienced pediatric spine surgeons [3].

There are many potential benefits for using ribs as anchors (Table 48.1), the first of which is that the rib attachments may allow for motion preservation. The ribs are attached to the spine via the costotransverse joints (neck of rib to anterior portion of transverse process) and the costovertebral joints (head of rib to vertebral body). The costovertebral joints constitute a series of gliding or arthrodial joints formed by the articulation of the rib head with the facet on the contiguous vertebrae. Ribs 1, 10, 11, and 12 articulate with single vertebral bodies; the remaining ribs attach to two vertebrae [4] (Fig. 48.2a, b). These joints permit a gliding motion. During normal respiration, there is just over 10° of bucket-handle motion of the ribs relative to the spine [5]. In addition, the interface of a hook on the rib is not rigid and allows for some "slop." Yamaguchi et al. reviewed a series of 176 patients treated with growing rods. In this series, proximal rib anchors were protective against rod breakage with a 77 % decreased incidence compared to proximal spine anchors [6]. Additionally, rib hooks may have an advantage in preventing anchor failure. Akbarnia et al. performed the first study to evaluate the properties of rib hooks used as an upper growing rod foundation in a porcine cadaver model [7]. They showed that rib hooks had a significantly higher load to failure than transverse process–lamina hook pairs and lamina hook–lamina hook pairs. Rib hooks had a higher load to failure than pedicle screws but this was not significant.

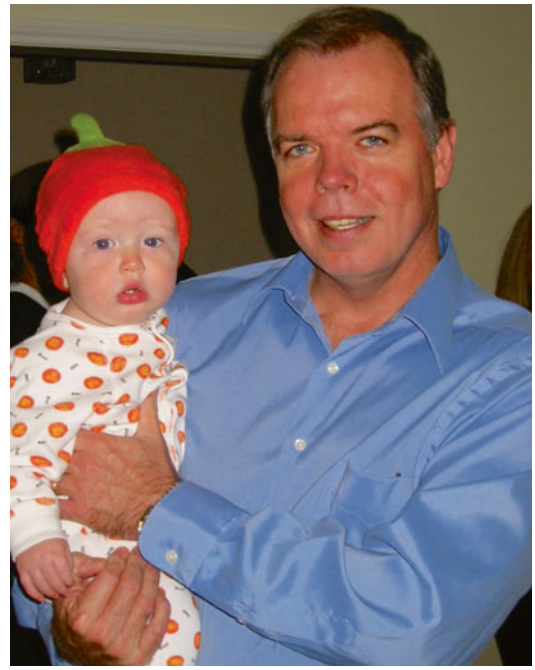

Fig. 48.1 Dr. Robert Campbell holding Clay Skaggs, 2001, Los Angeles (Reproduced with permission of Children's Orthopaedic Center, Los Angeles)

Table 48.1 Possible benefits of spine implants as rib anchors

Motion preservation
No dissection of spine
Good soft tissue coverage
No special equipment, training, or institutional approval needed
Load sharing over multiple ribs
No fusion of the upper thoracic spine, which is important for pulmonary development

In contrast, traditional growing rods attached to the spine permit little motion of the vertebrae within the construct. As with any diarthrodial joint, prolonged immobilization decreases motion and can lead to spontaneous fusion. It is a common experience upon converting growing

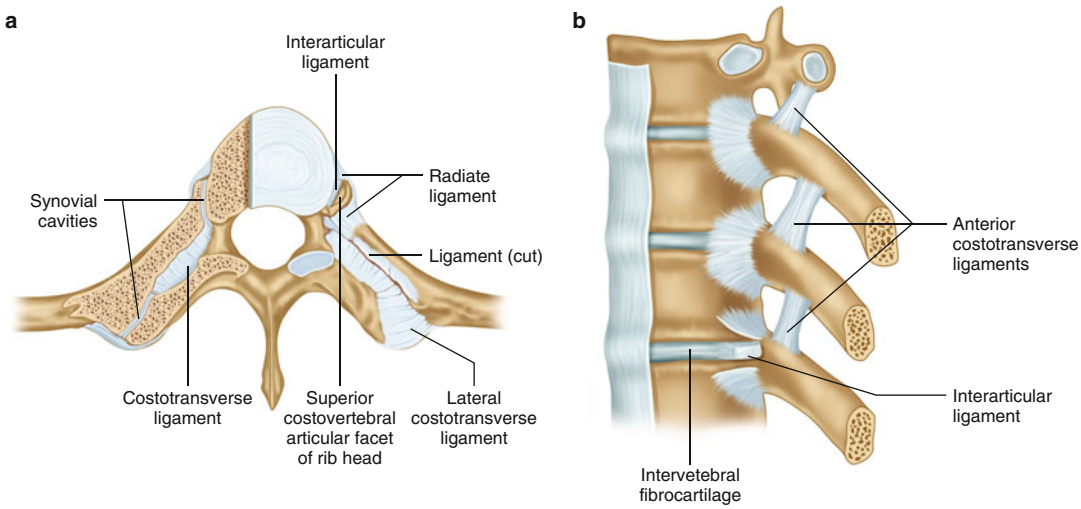

Fig. 48.2 (**a**, **b**) Demonstration of the costotransverse and costovertebral joints

rods to a final fusion construct to discover spontaneous fusion of most, if not all, of the vertebrae within the construct. Sankar et al. substantiated this theory demonstrating in patients treated with dual growing rods that over time there was less increase in T1 to S1 distance with each subsequent growing rod lengthening [8]. This "law of diminishing returns" is a particular concern when growing rods are placed in young children, i.e., if dual rods are placed in a 2-year-old, the spine may be fused by age 7. In theory, the use of the ribs for anchor points will permit motion between vertebrae and prevent or delay spontaneous fusion. This technique is too new to have adequately tested this theory.

Another advantage of using ribs as anchors is to stay out of the spine and preserve a virgin spine for future surgeries. While traditional growing rods aim for a fusion at the top of the construct, methods that attempt nonfusion, such as Luque-Trolley (sublaminar wires and rods without fusion), have been shown to cause fusion in 100 % of patients in one series [9]. Fusion in the upper thoracic spine (T1–T3) in young children is particularly harmful to long-term pulmonary function, so avoiding fusion in this region may be an important benefit of this technique [10].

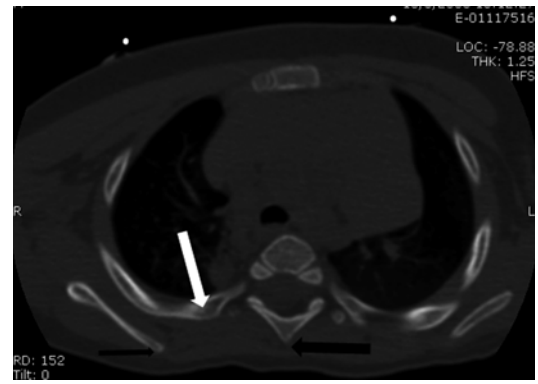

Fig. 48.3 CT of the chest. Note that the attachment point of the rib anchor (*white arrow*) is in a trough with good soft tissue coverage between the spinous process (*thick black arrow*) and scapula (*thin black arrow*) (Reproduced with permission of Children's Orthopaedic Center, Los Angeles)

It has been shown that children with thoracic insufficiency are nutritionally depleted, with 79 % being below the fifth percentile for weight [11]. Soft tissue coverage of traditional spine implants can be challenging in this population. A further advantage of rib anchors is that they tend to have good soft tissue coverage, as they are located deep to the rhomboids and trapezius, in a valley between the more prominent spine and scapula (Fig. 48.3).

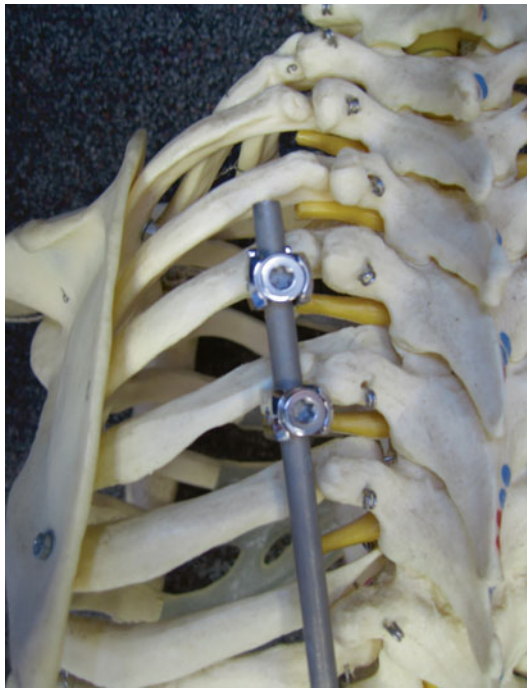

Fig. 48.4 With hooks of standard spine implants, multiple ribs may be engaged to share the load over multiple ribs. Note the hooks are immediately adjacent to the transverse process (Reproduced with permission of Children's Orthopaedic Center, Los Angeles)

A significant practical advantage of using traditional spine implants on ribs rather than VEPTR is that no special equipment is needed. Hooks that fit the ribs are readily available on all spine implant systems. There are, thus, no special equipment needs and no special training needed for the surgeon or operating room staff. In addition, there is no need for institutional or research approval as traditional spine implant hooks are FDA approved. A particular advantage over the original VEPTR design is that load sharing over multiple ribs is quite simple (Fig. 48.4).

48.2 Indications

The indications for use of ribs as anchors in growing systems continue to evolve. This remains an area where further investigation is needed and a particular area of uncertainty even among experienced early onset scoliosis surgeons [3]. The argument may be made to use rib attachments in children under the age of 5 years, as they would be expected to have growing implants for at least 5 years, and rib attachments may decrease the risk of spontaneous fusion as discussed earlier. Another indication is when there is already a substantial fusion of the mid thoracic spine present (whether from previous surgery or congenital), and one wants to minimize further fusion in the upper thoracic spine. In cases of previous infection of growing implants, the ribs provide an area of new, uninfected tissue as salvage. Using rib anchors also allows one to avoid sites of previous surgery such as laminectomies.

Another indication for using ribs as anchors is in cases of significant cervicothoracic scoliosis and head tilt, in which the upper ribs are fused. This is discussed in detail near the end of the chapter.

48.3 Contraindications

Rib attachments tend to function poorly in cases of kyphosis as the ribs tend to pull backwards over time as the spine falls forward. In particular, upper thoracic kyphosis is poorly controlled with rib attachments, and this may be a time where traditional growing rods bent into kyphosis and attached cephalad to the region of kyphosis are more appropriate (Fig. 48.5a, b).

48.4 Thoracotomy Generally Unnecessary

It is very uncommon that a formal thoracotomy is used with this technique. It has been shown that a thoracotomy in the treatment of scoliosis leads to a disruption of pulmonary function and is simply not needed to improve spinal and thoracic deformity in the great majority of cases [12]. Soft tissue or osseous release between ribs is rarely needed, except in the truly rare case of multiple rib fusions limiting thoracic expansion. It is natural that ribs are closer together in the concavity of a curve than the convexity, and that is not an indication for tissue destruction between the ribs.

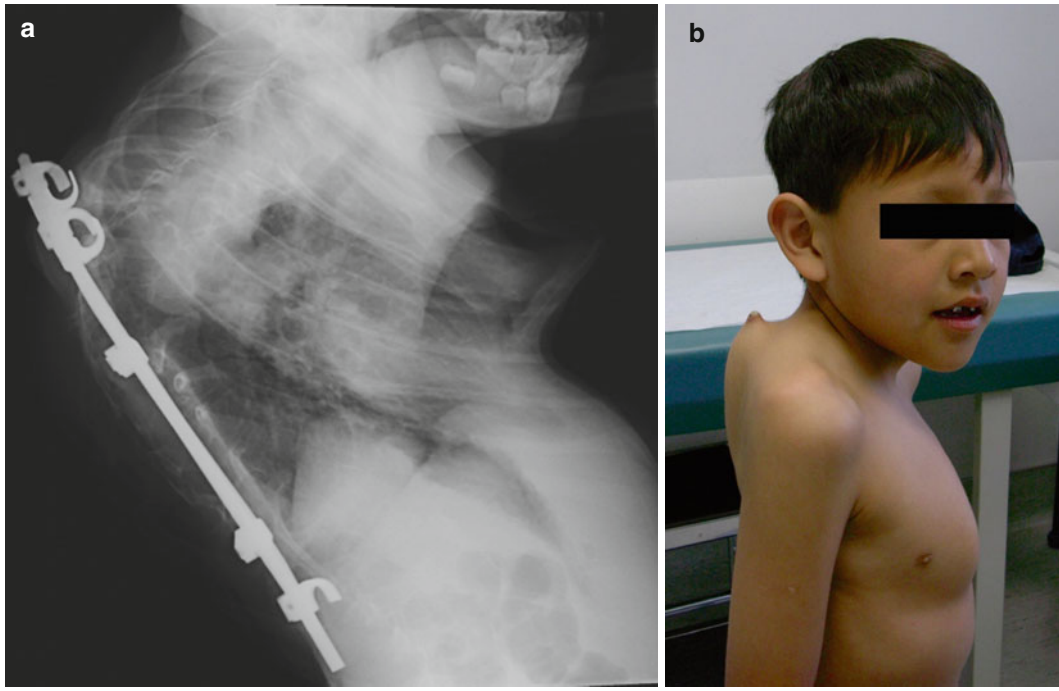

Fig. 48.5 (**a**) Lateral radiograph demonstrating progressive kyphosis despite rib implants. (**b**) Clinical photograph demonstrating rod coming through the patient's skin (Reproduced with permission of Children's Orthopaedic Center, Los Angeles)

When the scoliosis is improved by distraction implants, the spaces between the ribs open up in a harmonious fashion (Fig. 48.6a, b). In contrast, distraction across a formal thoracotomy distracts between the two ribs at the site of the thoracotomy (Fig. 48.7). Any tissue lysis between ribs cannot help but leave scar tissue, which is less mobile and functional than virgin intercostal muscle.

48.5 Surgical Technique

Neurologic monitoring should include both the upper and lower extremities. Joiner et al. have described the brachial plexus injuries that may occur in this setting [13]. Noted in this study were two cases in which the neurologic symptoms were present when adducted but resolved with the arms abducted in the typical positioning for a prone patient undergoing spine surgery. Consequently, positioning of the child with arms adducted is recommended to allow for accurate intraoperative monitoring (Fig. 48.8).

The spine is approached through a midline skin incision as it is likely that this incision will be used for the final fusion in the future. Depending on the specifics of the surgery, a long midline incision or separate incisions at the top and bottom of the construct may be made. As the figure demonstrates, the top incision for the rib attachments is 4 cm in length (Fig. 48.9). The skin is undermined laterally past the transverse processes. The transverse processes are generally palpable as a point of resistance through the muscles. If there is any question, fluoroscopic imaging over a needle placed into bone clarifies the location. A combination of muscle splitting and cautery in a vertical incision should bring one quickly to the ribs with minimal blood loss.

Care should be taken to make certain the dissection is immediately adjacent to the transverse process only. There is a tendency for the hook to slide laterally if soft tissues are dissected laterally. In addition, the implants exert the most control of the spine when they are adjacent to the transverse process, as opposed to a more lateral

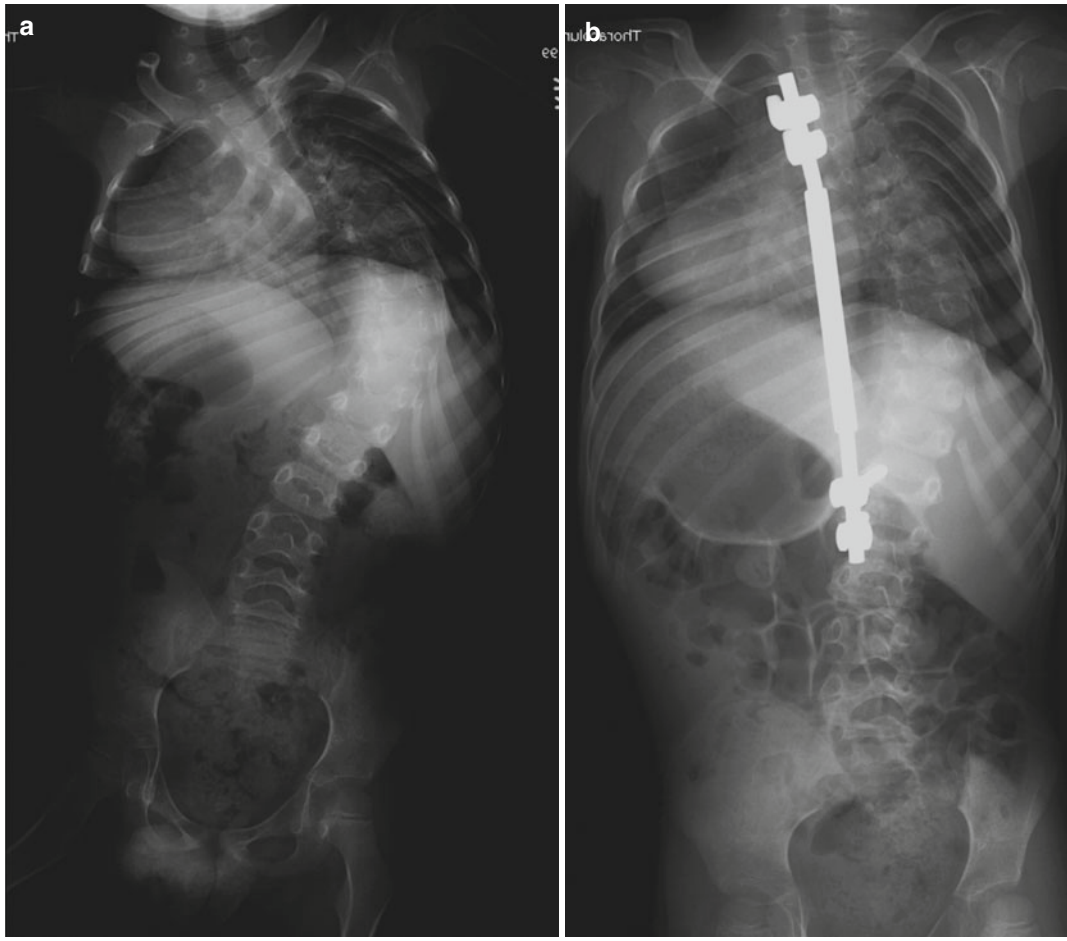

Fig. 48.6 (**a**) A preoperative AP radiograph of 91° scoliosis with ribs on the concave side appearing constricted. (**b**) Evidence of harmonious distraction of the ribs through distraction instrumentation without scapula elevation, thoracotomy, or any lysis of tissue between ribs (Reproduced with permission of Children's Orthopaedic Center, Los Angeles)

placement in which the ribs tend to move cephalad in a bucket-handle fashion independent of correction of the spine.

Ideally, the periosteum is preserved around the rib to allow the rib to hypertrophy over time in response to stress. Surprisingly, the neurovascular bundle caudad to each rib is really of no consequence and can be ignored in this surgery. To place a hook, a 5-mm transverse incision with cautery is made just distal to the neurovascular bundle and immediately adjacent but lateral to the transverse process. A freer elevator is then used to dissect soft tissue anterior to the rib, aiming to exploit the plane between the periosteum and the pleura (Fig. 48.10a). In reality, this technique evolves over time to the point where one simply pushes the hook into position without any preliminary dissection. Usually, a second hook is placed around a second rib as well. Both hooks are upgoing. There is no need for a down-going hook, as distractive forces keep the rib engaged in the hook, and a properly sized hook extends a bit cephalad to the rib (Fig. 48.10b). We are not aware of any ribs migrating off the top of hooks.

Attention is then turned to the distal anchor point. Through a midline incision, the lamina of the intended vertebrae is subperiosteally dissected. Either one-level fixation with a down-going

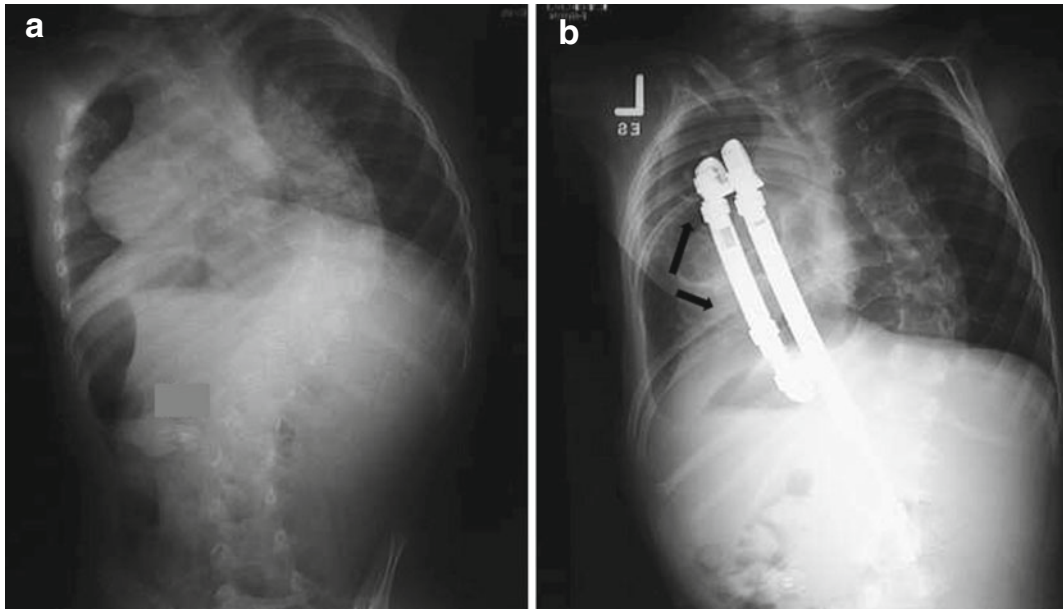

Fig. 48.7 (**a**, **b**) Pre- and postoperative radiographs demonstrate large opening of two ribs at a thoracotomy site (between *black arrows*) and compression of rib spaces above the thoracotomy (Reproduced with permission of Children's Orthopaedic Center, Los Angeles)

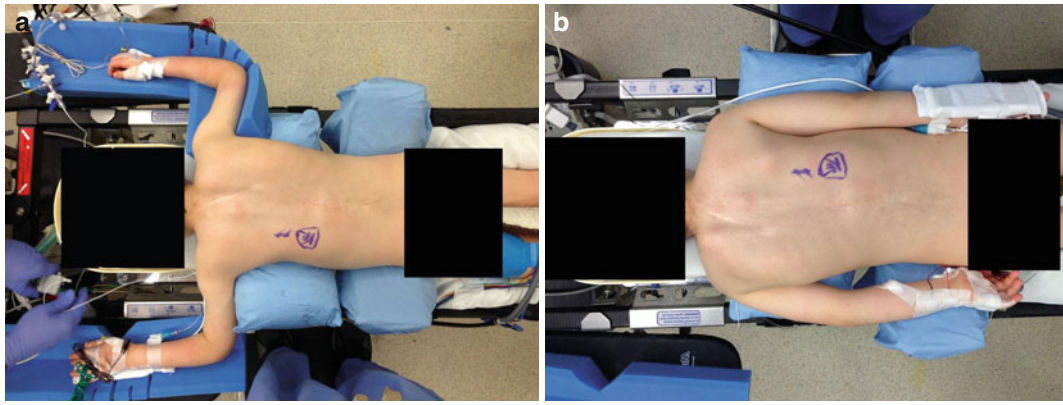

Fig. 48.8 Clinical photo showing typical prone position (**a**) and recommended adducted position (**b**)

supralaminar hook, or two-level fixation with pedicle screws may be used. The theoretical advantage of a one-level hook construct is that no fusion is intended, and there is a certain amount of "slop" that may translate to motion and less spontaneous fusion. The disadvantage to using a single hook is that it often migrates posteriorly over time, leading to either a bump that is concerning to the parents or migration through the lamina and need for revision. A potential pitfall here is to make sure the interspinous ligament is left intact when placing a supralaminar hook or distraction may lead to kyphosis of the distal segment. When using pedicle screws, always place them in at least two segments (unilateral or bilateral) as distal migration over time through pedicle screws could injure nerve roots along the inferior border of the pedicle. The facet joint between the two pedicle screws is destroyed with a narrow rongeur and corticocancellous crushed allograft is placed in the joint.

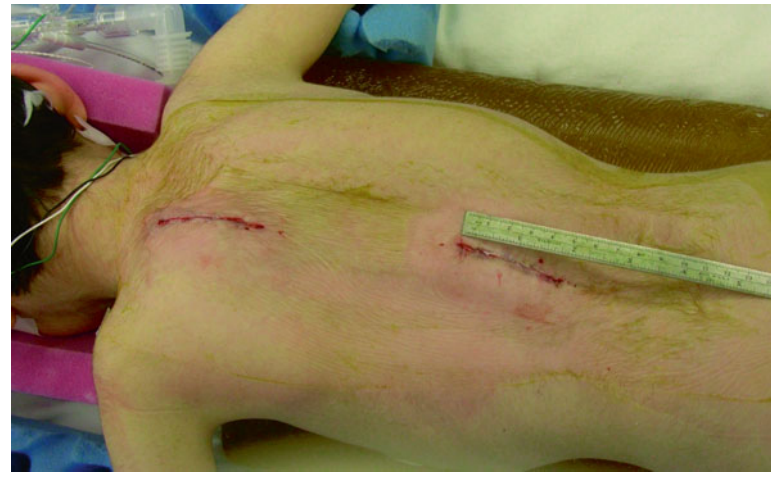

Fig. 48.9 Intraoperative photograph demonstrating a 4- and 5-cm incision for growing rod implants. Same case as Fig. 48.6 (Reproduced with permission of Children's Orthopaedic Center, Los Angeles)

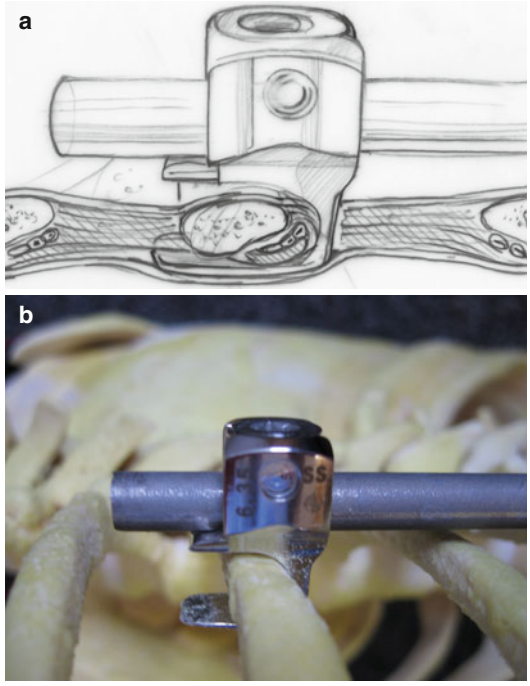

Fig. 48.10 (**a**) Cross-sectional drawing of a lumbar hook engaging a rib. The exact location of the neurovascular bundle within or outside of the hook is not important. (**b**) Side view of lumbar hook engaging a rib. Note the anterior portion of the hook extends superiorly to the rib, minimizing the possibility of superior migration of the rib away from the hook. The rib is not likely to fall out in a cephalad direction as the hook is distracting the rib (**a**: Reprinted from Skaggs et al. [11]. With permission from Wolters Kluwer Health. **b**: Reproduced with permission of Children's Orthopaedic Center, Los Angeles)

Table 48.2 Pearls and pitfalls

Keep the chest wall intact! It is rarely necessary to cut between ribs, do a thoracotomy, or elevate the scapula
Keep the dissection and hook as close to the transverse process as possible, or the hook may slide laterally
If using a supralaminar hook for the distal anchor, leave the supraspinous ligament intact, or risk kyphosis of this vertebrae
If in doubt, use a longer construct, especially in younger children, as the curve is likely to add on over time if a short construct is used

The exposed bone of the level of the distracting is decorticated and bone graft placed before the rod is inserted to maximize osseous contact. See Table 48.2 for additional pearls and pitfalls in anchor placement.

The upper and lower rods may be connected with either a traditional longitudinal growing rod connector or a side-to-side connector with the rods overlapping (Table 48.3, Fig. 48.11). If using a side-to-side connector do not rely on only one for the whole system or it may fail over time, perhaps as a result of bending moments in addition to compression forces.

Make a soft tissue tunnel between the two anchor sites with a tonsil clamp, being deep to muscle directly on the ribs. Pull a chest tube through this tunnel, and attach the end of the rod into the chest tube, which is then used to safely pass the rod from one anchor to the other in a submuscular fashion.

Plan on cutting rods at least 2–3 cm longer than the anchors to permit intraoperative distraction. Once distraction is performed, fill the upper anchor site with warm saline, and ask the anesthesiologist to perform a Valsalva maneuver to look for a pleural leak. If one is present, we place a small hemovac in the chest to function as a chest tube for a few days.

48.6 Unilateral or Bilateral Rods

This technique may be used with unilateral rods or bilateral rods. Unilateral rods are minimally invasive but have less anchor points to share load. Both acutely, and more so over time, balancing the curve can become problematic with a single unilateral rod (Fig. 48.12a–c). We rarely use unilateral rods except to span a unilateral bar. In general, bilateral rods seem more stable and less prone to loss of fixation. Bilateral rods also make balancing the spine easier, especially over time, with the ability to preferentially distract one side more than the other. A minor disadvantage of bilateral rods is that more dissection is needed, particularly at the bilateral rib attachments, though this does not affect the spine and is probably of little importance (Fig. 48.13a, b).

Table 48.3 Comparison of side-to-side and end-to-end connectors

Type of connector	Advantage	Disadvantage
Longitudinal connector	• Less bulky, only one rod	• No sagittal contouring • Limited expansion potential
Side-to-side connector	• Sagittal contouring possible • More expansion potential	• More bulky, two overlapping rods

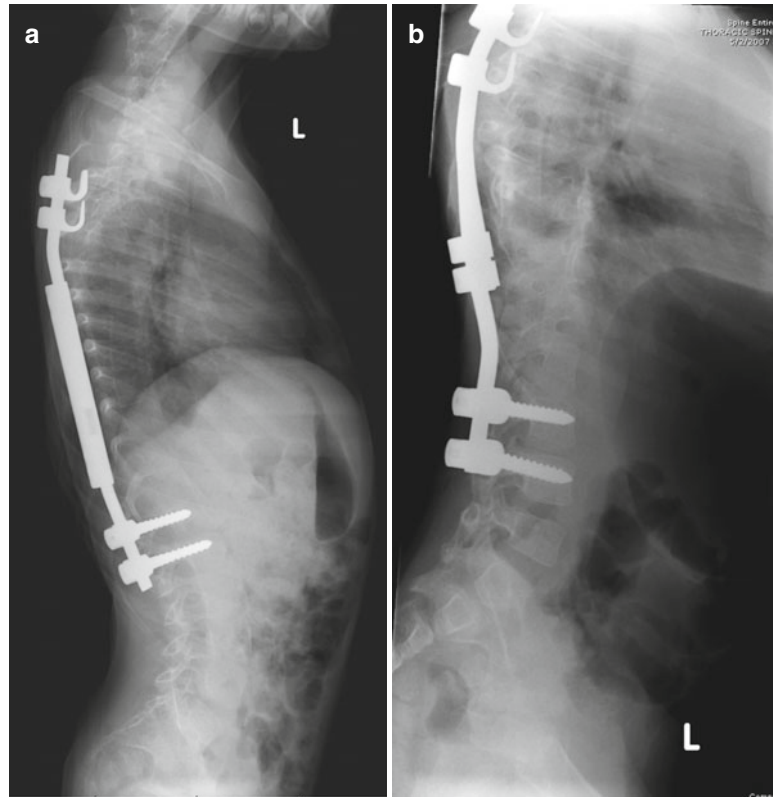

Fig. 48.11 (**a**) Lateral view of a longitudinal connector. (**b**) Lateral radiograph demonstrating a side-to-side connector with physiological sagittal contouring of rods (Reproduced with permission of Children's Orthopaedic Center, Los Angeles)

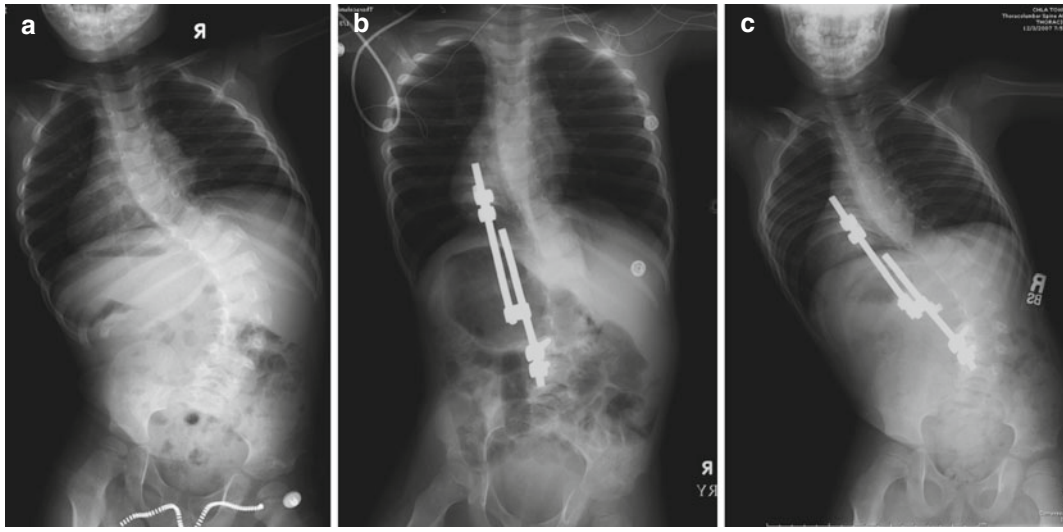

Fig. 48.12 (**a**) Preoperative AP radiograph. (**b**) Postoperative unilateral growing rod. (**c**) Two years later, significant decompensation has occurred (Reproduced with permission of Children's Orthopaedic Center, Los Angeles)

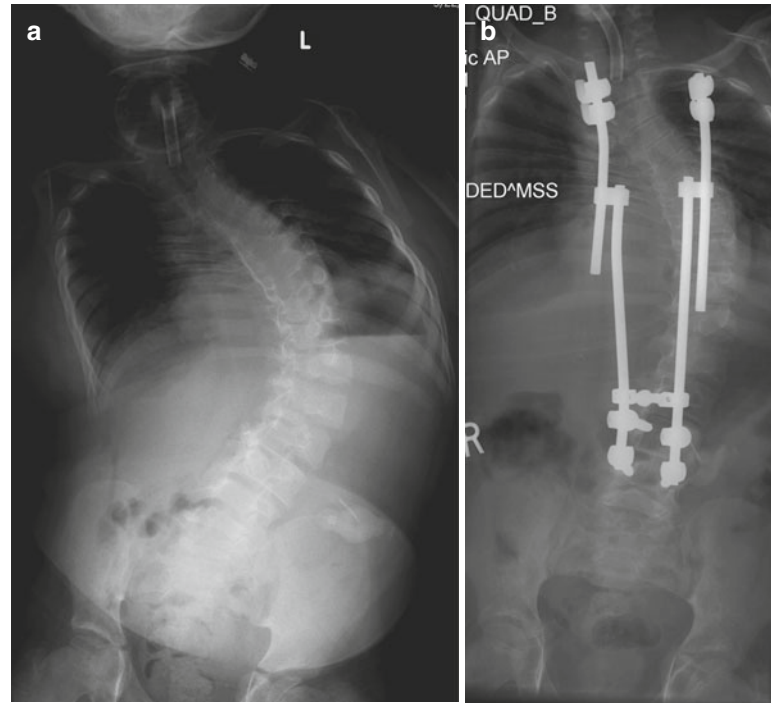

Fig. 48.13 (**a**) AP radiograph demonstrates an 83° curve in this 8-year-old with neuromuscular scoliosis and pulmonary problems. (**b**) Postoperative demonstration of bilateral growing rods with hooks as rib anchors (Reproduced with permission of Children's Orthopaedic Center, Los Angeles)

48.7 Complications

A unique feature of using ribs as anchor points in distraction-based growing instrumentation is the possibility of neurologic injury to the upper extremity. A multicenter prospective study of the VEPTR study group found that neurologic injury to the upper extremity was six times more frequent than to the lower extremity. The authors conclude the rate of potential neurologic injury (neurologic injury plus monitoring change) during primary device implantation (2.5 %) and device exchange (1.3 %) justifies the use of intraoperative neuromonitoring of the upper and lower extremities during primary and exchange VEPTR surgeries [14].

Joiner et al. have described mechanisms by which neurologic injury to the upper extremity occurs. The three mechanisms observed were [13]:

1. Brachial plexus injury by pushing the first rib superiorly during distraction of rib-anchored growing instrumentation
2. The superior pole of the retracted scapula causing direct injury to the brachial plexus
3. Pulling the scapula inferiorly during Sprengel's deformity reconstruction causing brachial plexus injury

Avoiding placing an anchor on the first rib (unless it is fused to other ribs – see below) is recommended to avoid this complication (Fig. 48.14). Direct impingement of the superior tip of the scapula into the brachial plexus is another mechanism by which brachial plexus injury occurs. Therefore, care should be exercised when the scapula is elevated off the chest wall. In general, this mechanism is easily avoided as there is not a reason to elevate the scapula if a thoracotomy is not being performed, which is the case in the great majority of surgeries. Additional brachial plexus injury may occur from pulling the scapula inferiorly as is performed during a Sprengel's reconstruction. Consequently, patients with concomitant Sprengel's deformities are at increased risk.

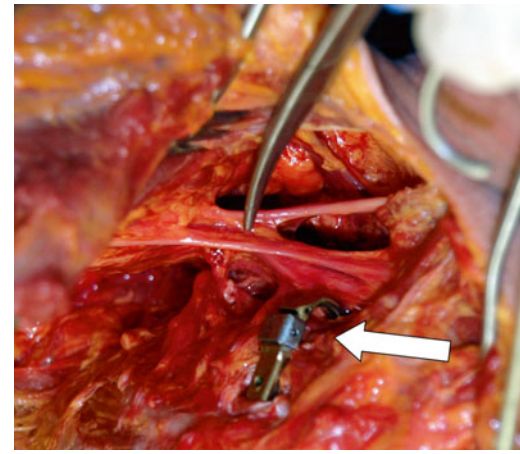

Fig. 48.14 Cadeveric dissection demonstrates the brachial plexus draping over the first rib. *White arrow* shows a VEPTR cradle on the first rib–rib anchors on the first rib alone should be avoided (Reproduced with permission of Children's Orthopaedic Center, Los Angeles)

The hook–rib articulation is mobile, and plowing of the hook through the rib may take place over time. In single unilateral rod constructs with one hook on one rib, this is likely over many years, as has been demonstrated in the case of VEPTR. However, we have not seen a spinal hook plow through a rib when multiple hooks and bilateral rods have been used. If this occurs, it is generally not much of a problem, as the rib grows back, often with more and stronger bone than before, and the same rib can be used again. Often times, there is so much new bone about the rib that a power bur may be needed to cut a slot into the bone mass to allow the hook to be properly seated.

A study at Children's Hospital of Los Angeles compared 36 children with spinal deformity treated either by dual growing rods, VEPTR, or the hybrid technique described in this chapter with spinal hooks on ribs. The

children were a mean of 4 years and 10 months at first surgery with a mean of 51 months of follow-up and a mean of four lengthenings each. The rate of having major complications requiring an unplanned surgery was 230 % for growing rods, 237 % for VEPTR, and 86 % for the hybrid group with spinal hooks on ribs (Table 48.4) [15].

48.8 The Special Case of Cervicothoracic Congenital Scoliosis

Cervicothoracic congenital scoliosis presents a number of unique problems. The head is usually much more tilted than it would be if the same size curve were more caudal, because there are no vertebrae above the curve to create a compensatory curve, and the head tilt is often a very noticeable deformity. In addition, this is an area where spine surgeons do not commonly perform an anterior exposure. The good news is that there are frequently multiple ribs fused at the top, which present a solid fixation point. Although the first rib is generally avoided as a fixation point to prevent migration of the rib into the brachial plexus, the fused rib mass is not mobile and can be used as a fixation point with little risk of brachial plexus injury. Of course, neuromonitoring of the upper extremity is mandatory at the time of the primary surgery if the fused ribs are used. This is the one situation where we will intentionally place the rib hook a distance lateral to the transverse process to take advantage of the moment arm and maximally improve the pathologic tilt of T1 (Fig. 48.15).

In the case of hemivertebrae opposite a unilateral bar, consideration should be given to an instrumented compression and fusion for the hemivertebrae with pedicle screws. This may obtain modest correction acutely and hopefully prevent future growth anteriorly and posteriorly.

Table 48.4 Complications of growing spine surgery

Major complications	Ccx rate (%)	Ccxs/cm growth	Ccxs/year treatment	Ccxs/planned surgeries
Dual growing rods	230	0.20/cm	0.52/year	0.47
Spine hook on rib hybrid	86	0.19/cm	0.36/year	0.29
VEPTR	237	0.97/cm	0.52/year	0.44

Based on data from Ref. [15]
Ccx complication

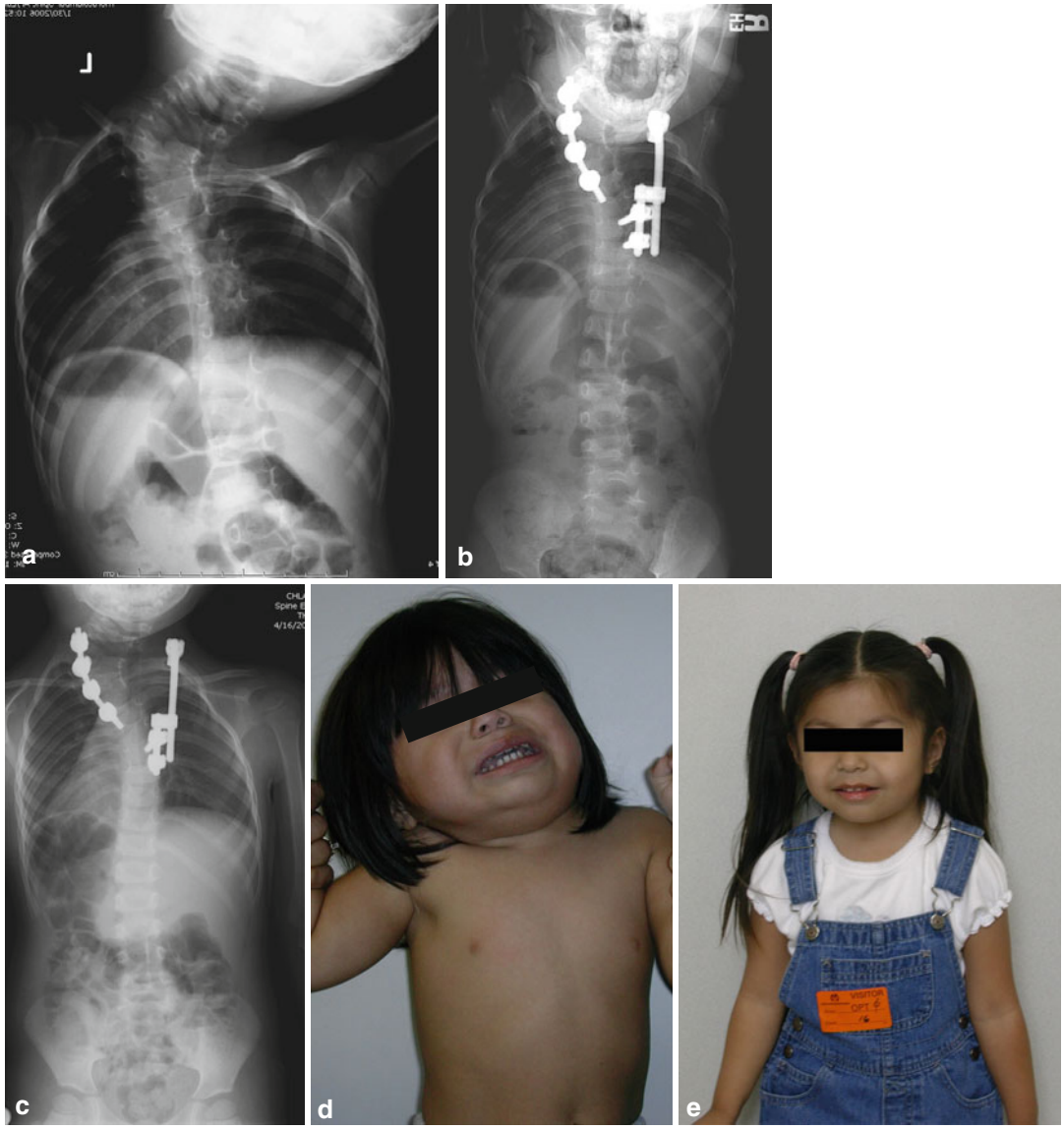

Fig. 48.15 (**a**) Preoperative AP radiograph demonstrates a unilateral bar opposite six pedicles. (**b**) Immediate postoperative AP radiograph. Note improvement in head position. (**c**) AP radiograph 1 year after initial surgery, with one surgical lengthening having been performed in the interim. Note a little migration of the hook on the upper rib, with new rib growing behind it. If this migrates through in the future, the hook could most likely be replaced on the same rib. The head position is significantly improved from preoperative position. (**d**) Preop clinical photo. The child's ear almost rested on her shoulder. (**e**) Postop clinical photo demonstrating no significant deformity. The child has no pain or complaints (Reproduced with permission of Children's Orthopaedic Center, Los Angeles)

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49 Plate-Rod System in the Management for Progressive Scoliosis in Growing Children

Qibin Ye, Zhengda Kuang, Guanjun Wang, Jin Lin, Xinyu Zhang, and Mingkui Du

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Key Points

- PRSS is a growing rod that was developed in PUMC Hospital in 1998. Several experiments have confirmed PRSS possesses the function of modulating asymmetrical growth of the scoliotic spinal segments.
- The PRSS is implanted in a single surgery without fusion and is thereafter lengthened by its own modulating efficiency which is expressed in the X-ray evidence of more longitudinal growth on the concave side than convex side, and the wedging vertebrae were gradually remodeled to normal contour in the lateral growing years.
- This technique is an effective and safe instrumentation for correcting growing scoliosis, especially for EOS.

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49.1 Introduction

Although many growth friendly techniques are effective in correcting the deformity in EOS, they do not stop progression of scoliosis curvature. Scoliotic curve progression after surgical correction in growing children continues to be a problem. The rapid progression is thought to be due to

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asymmetrical growth after surgical treatment of childhood scoliosis which results asymmetrical stress on both sides of the scoliotic spine. Aronsson [1] stated that the convex side grows 10 % more length than concave side in 30° scoliotic spine. A small lateral curvature of the spine can produce asymmetrical spinal loading that causes asymmetrical growth and a self-perpetuating progressive deformity during skeletal growth [2]. Many factors can affect the longitudinal growth of vertebrae, but the most important one is stress [3].

In recent years, all the researchers are looking for a way to modulate the asymmetrical growth on both sides of the cartilage end plate of the scoliotic spine to reverse such Hueter-Volkmann principle. In the laboratory (animal model), scoliosis with vertebral wedging has been created by asymmetrical mechanically loading and has been corrected by reversing the loading. The mechanical modulation of vertebral growth in the presumed asymmetrically loaded scoliosis spine with scoliosis was described by Stokes and Roaf as a "vicious cycle" [4–6]; they addressed the concept of mechanical modulation of vertebral body growth in the pathogenesis of progressive adolescent scoliosis generally attributed to the Hueter-Volkmann principle in which constant pathologic strong pressure inhibits endochondral longitudinal growth while reduced compression accelerates growth. We think that transference of idea from controlled animal studies to the clinical application is possible if a new device which has modulating function be able to provide two types of effects: either to modify the vertebral growth asymmetrically or to modify the forces acting on the spinal column and end-plate physes. Currently, several growth friendly instrumentation systems are adopted in the clinical practice. These include distraction based (growing rods [7] and VEPTR [8]), guided growth (Luque trolley and Shilla [9]), and compression based (Staple [10] and Tether [11]) instrumentation systems. All these devices have the merit to correct progressive EOS, but they need repeated instrument-lengthening operations (every 6 months on average). Besides, a definitive spinal fusion will be required later. Ideally, the surgical management should be able to correct the deformity without fusion and maintain the correction results in one-stage operation For this purpose, plate-rod system for scoliosis (PRSS) was developed in PUMC Hospital in 1998 [12].

49.2 Design Rationale and General Principles of PRSS Instrumentation

The PRSS instrumentation is designed for the treatment of scoliosis in young children, and this system is composed of four components: the plate rod, the rod, the screw hook, and the connectors. The components are made of titanium alloy (Fig. 49.1).

The screw hooks are fixed on the lamina, which minimizes the risk of dislodgement of the hooks. Plate rod is placed on the convex side by way of lateral sidewise push to provide asym-

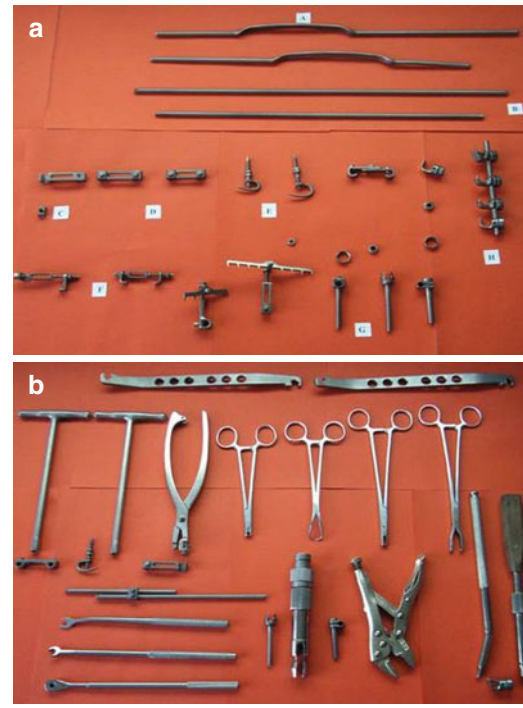

Fig. 49.1 PRSS instrumentation: (**a**) implant components; (**b**) surgical tools

Fig. 49.2 A set of PRSS

metrical stress on both sides of the vertebral end plates, and a cylindrical rod is placed on the concave side and they are connected by the upper and lower connector thus the system forms a strong frame-like construct (Fig. 49.2). The middle and bottom parts of the rods are not fixed; therefore, the rods are free from the hole inside of the lower connector, which allow the rods upward on migration with the growth of the spinal column after operation. They have no distraction force during the operation; this decreases the risk of paraplegia. Bone fusion procedure is not required; essential normal spine can be obtained after removing the implants when the skeletal growth terminated [13].

49.3 Therapeutic Mechanism of PRSS in the Treatment of Early-Onset Scoliosis

PRSS due to its characteristics of elasticity and curve structure, and the mobility of the middle and bottom part of the instrumentation as well as absence of fusion in the operation, lacks the tether effect on the concave side of the scoliosis; this allows PRSS rod migration upward and downward during patient's activity. In this manner, the concave side is constantly being stimulated by tensile stress, and the convex side is constantly being pushed by the lateral sidewise force, which will stimulate the growth of vertebral cartilage end plate, and at the same time it increases compression on the convex side, which inhibits the growth in this side; the subsequently asymmetrical growth over both sides of the vertebrae occurs in the subsequent growing year. Several experiments were studied which confirmed that PRSS has a function of modulating asymmetrical growth of the scoliotic spine.

The components of the PRSS system were tested with MTS machine (Material Testing System Bionex 858, Minneapolis Minnesota, USA) at the laboratory of the University of Hong Kong and Peking Union Medical College Hospital (Fig. 49.3). The results indicated higher spinal stiffness following instrumentation than those of the intact porcine spine models in vitro. Thus, bony fusion procedure is not necessary for maintaining the correction. When PRSS is implanted, asymmetrical stress is created by the lateral sideway push of the plate rod and can be calculated according to the Hooke's law [14–16].

49.3.1 Radiographic Analysis

When PRSS is applied in the spine, asymmetrical stress is created by the lateral sideway push of the plate rod, compressive stress is exerted over the convex side, while tensile stress is exerted over the concave side of the curvature which is expressed in the form of change of the width of the disc spaces (Fig. 49.4a, b). In this manner, asymmetrical growth over both sides of the vertebral cartilaginous end plate occurs, wedging is corrected, and the vertebrae are remodeled to normal shape (Fig. 49.4a, c) [17].

Fig. 49.3 Biomechanical test using porcine spine model

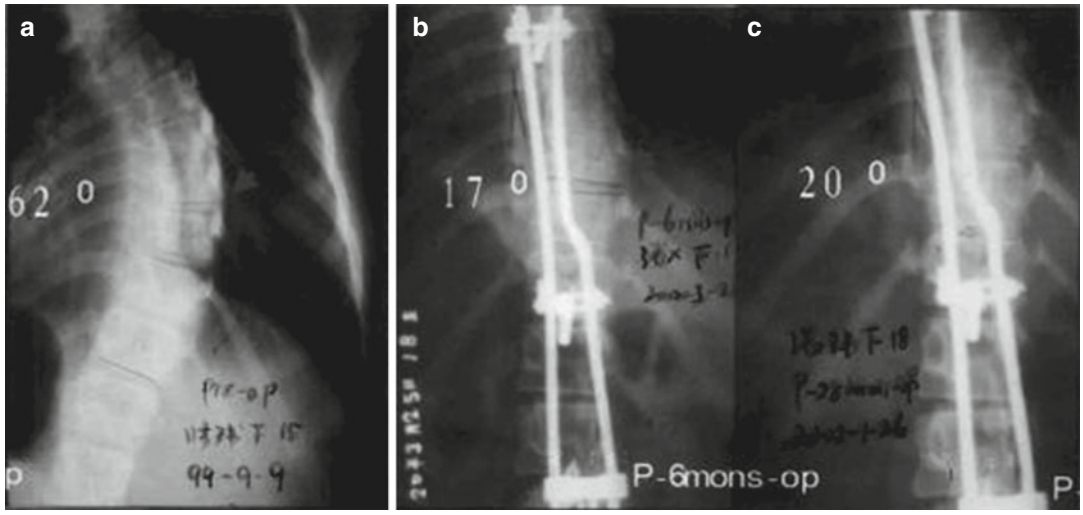

Fig. 49.4 Preoperative radiograph shows disc spaces open up on the convex side and close up on the concave side all along the apical region (**a**), which is reversed on both side after PRSS was applied (**b**). Another A–P view taken 28 months after operation shows wedging vertebrae were remodeled to normal shape (compare both **a** and **c** at the *arrow* site), indicating asymmetrical growth on both sides

49.3.2 Photoelastic Test

Photoelastic study was used to show the mechanical properties of PRSS [18]. A model of scoliosis and correcting clamp was built on the base of similar principle (Fig. 49.5a).

According to the factual situation, the in situ stress was measured by using photoelastic and strain gage method. ANSYS (a finite element analysis software) method was also applied to simulate the experiment process and evaluate the reliability of measurement.

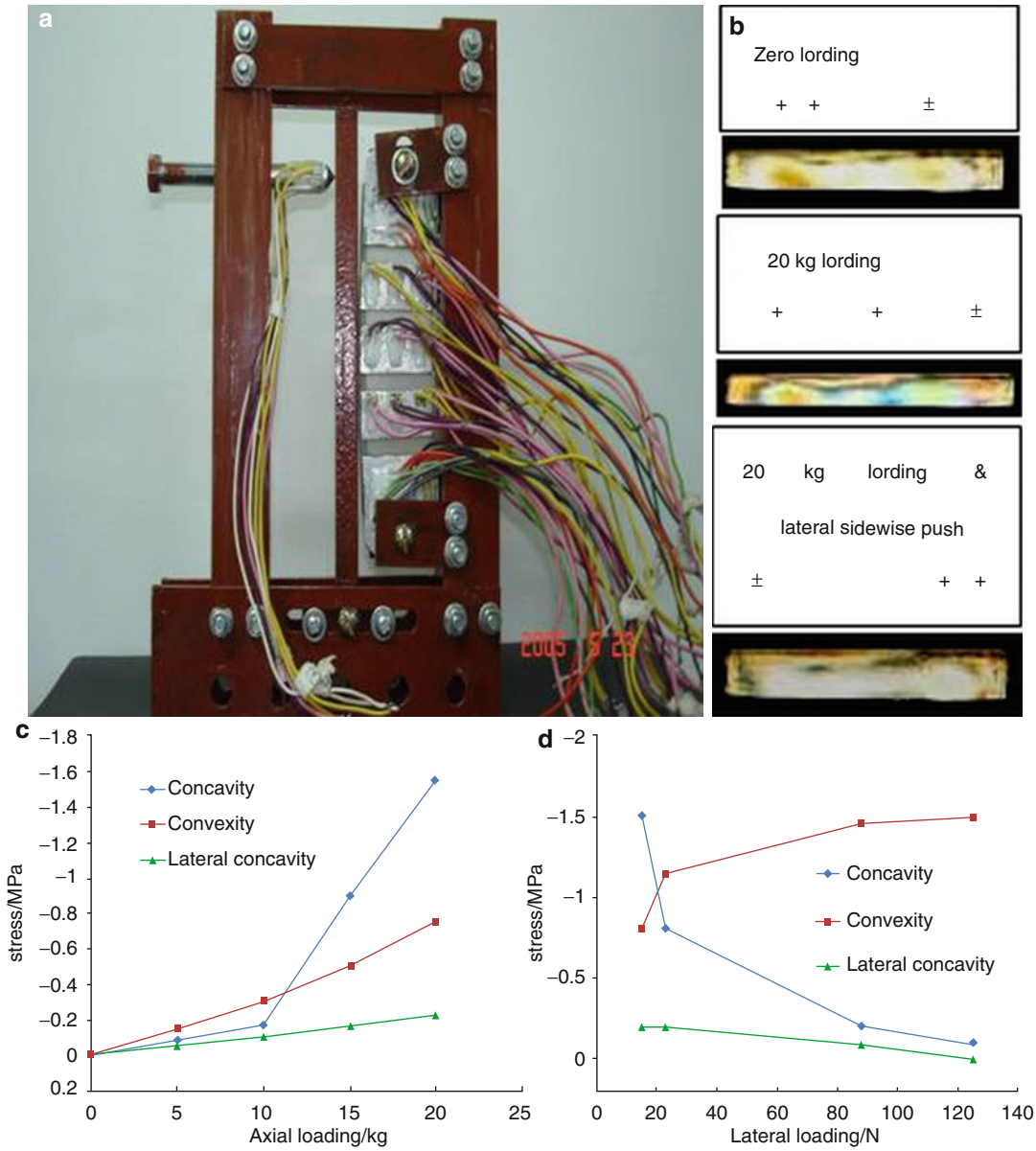

Fig. 49.5 Photoelastic study shows PRSS provides asymmetrical stress on both sides of the end plates of the vertebrae: (**a**) A model of scoliosis and correcting clamp. (**b**) Photoelastic stress under different corrective loading along spine. (**c**, **d**) The asymmetrical stress data were expressed in a linear formulation of *green line* as change of compressive stress on the concave side; *red* one on convex side

The results indicated that a small lateral curvature of the spine can produce asymmetrical spinal loading; the relational changes and variation of stress on both sides of scoliotic spine were demonstrated by the change on the color band in the photoelastic test. Color stripe showed more color stripe on the concave side than the convex side in the scoliotic spine (Fig. 49.5b at the top site) indicating that more compress stress in this side, and it was increased when 20 kg vertical load was given (Fig. 49.5b at the middle site); when the lateral push was applied over the convex side, more concentration of color stripe appeared on the convex side indicating compressive stress increase on this side and compressive stress decrease, and tensile stress was produced on the concave side (Fig. 49.5b at the bottom site). It was also shown that the stress value was in proportion to the correcting load. The asymmetrical stress data were expressed in a linear formulation of green line as change of compressive stress on the concave side and red one on the convex side (Fig. 49.5c, d), indicating PRSS has a significant efficiency to alter the asymmetrical mechanical loading on both sides of the scoliotic spine.

49.3.3 Type X Collagen Study

Type X collagen has also been studied to express the therapeutic mechanism. Type X collagen is used to reflect chondrogenesis, subchondral bone formation, and cartilage degeneration [19]. Type X collagen was studied as growth mark of cartilage end plate using semiquantitative RT-PCR method. The fact that more type X collagen was expressed on the convex side (Fig. 49.6-CL) than concave side (Fig. 49.6-CR) suggests that compressive stress leads to increase earlier cartilage degeneration of the convex side end plate, correlating as well with decreased growth of the end plate of this side and resulting in maximum spinal realignment.

49.4 Surgical Technique

49.4.1 Surgical Procedures

The patient is placed in the prone position. A standard posterior approach and exposure as for Harrington rod procedure is made. But exposure of the subperiosteal laminae is limited to the hook sites. In this way, the unwanted spontaneous posterior ankylosis or "autofusion" is minimized.

Hook insertion: The upper and lower hooks are inserted on the lamina of the selected end vertebrae, fixed with a screw and linked with the connector (see Fig. 49.7a, b), which forms two attached points to accept the rod and plate rod. An elastic prebent plate rod is first placed on the convex side by way of the lateral sideway push. This maneuver provides asymmetrical stress on both sides of the end plate of the vertebrae all

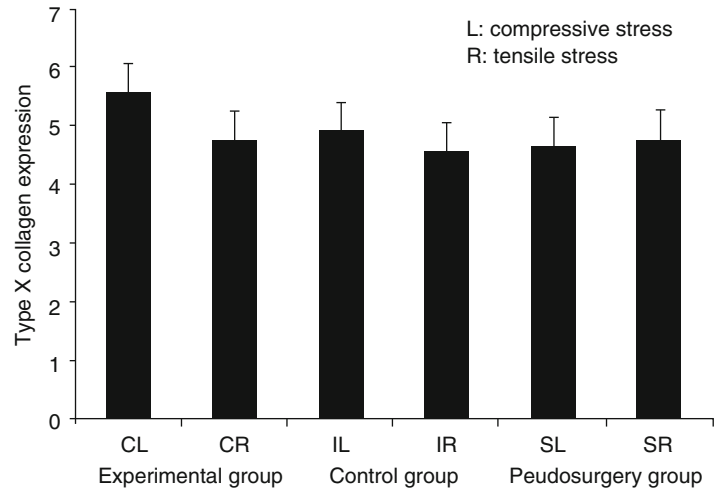

Fig. 49.6 Type X collagen expression level

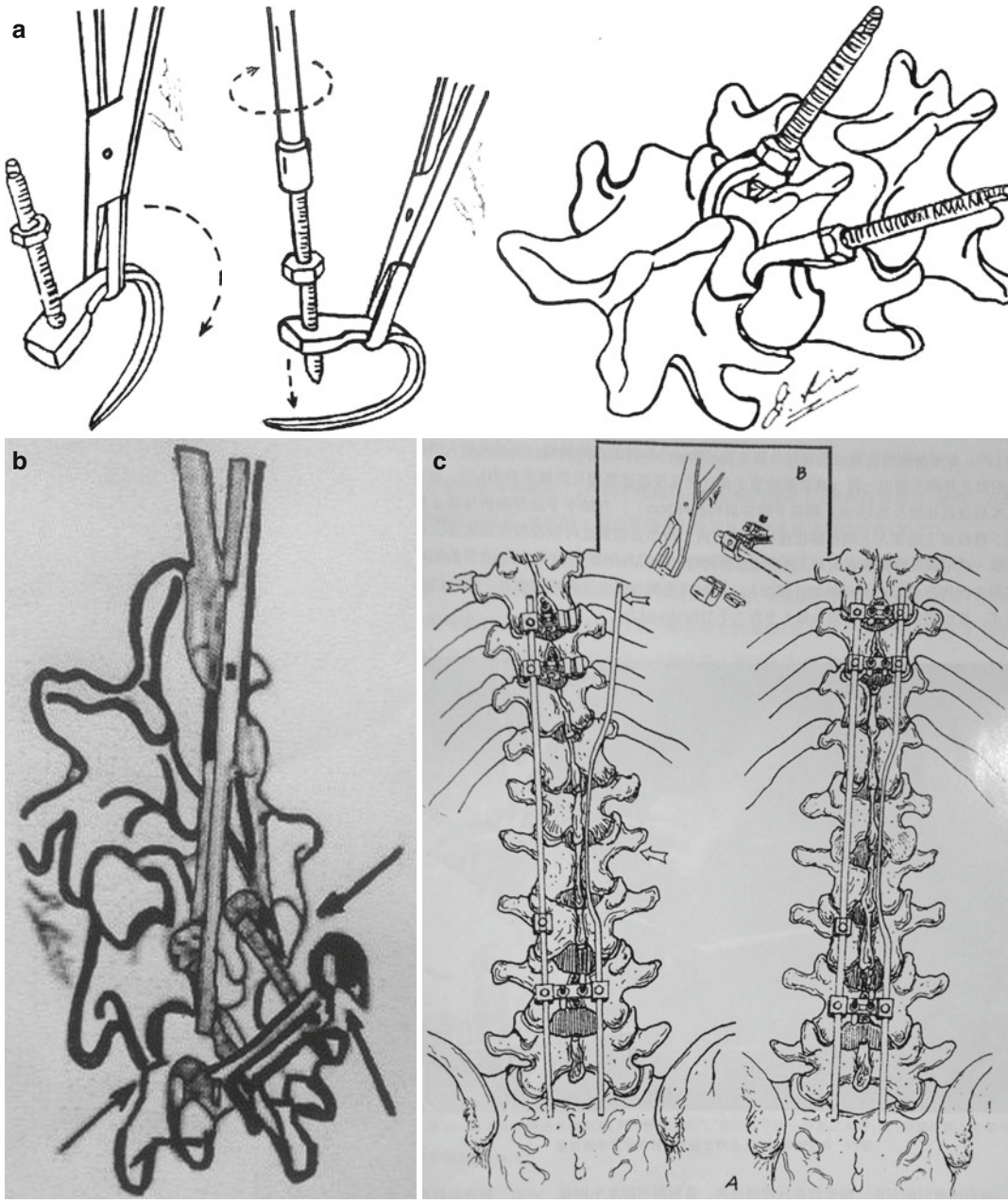

Fig. 49.7 Sketch map of correction of scoliosis with PRSS. (**a**) Screw hooks are placed under laminae and fixed onto the laminae. (**b**) Linked with the connector. (**c**) Plate rod is placed on the convex side and rod is placed on the concave side. This forms a frame-like spinal construct (Courtesy of Jin Lin)

along the apical region. The lower end of this plate rod is first passed through the hole of the lower connector to an appropriate length, and free from the inside of the hole, the upper end of the plate rod is then introduced into the open head of the upper connector and fixed with the setscrew. The rod (5.5 mm in diameter) is placed on the concave side after contouring to fit the profile of the spine. This forms a frame-like spinal construct (Fig. 49.7c). The upper and the lower

intermediate hooks are inserted in place in a manner similar to that of CD system. The hooks are linked with the interconnector and fastened with the rod and plate rod using the U-shaped ring. This allows additional correction of scoliotic curve. The screw caps of the intermediate screw hook on the concave side are tightened to bring the screws and its linking lamina backward for derotation of the vertebral bodies over the apical area (Fig. 49.8a, b).

There is no distractive force during surgical correction; hence, the risk of cord damage caused by overcorrection is unlikely. PRSS is a sliding instrumentation; only the upper end of the plate rod and rod are tightly fixed in place with the set-screw in the upper connecter, while the lower and middle part of the rods are left alone and free from the hole inside the lower connector, which allow the plate rod and rod upward migration to keep up with the longitudinal growth of the instrumented spinal column after operation and to obviate repeated operation. No bony fusion is performed.

49.4.2 Clinical Materials and Results

We have reported the outcome of a series of 23 juvenile scoliosis [20] and a group of 66 adolescent scoliosis [21] treated with PRSS showing that PRSS has a significant efficiency to lower the risk of scoliotic curve progression (Fig. 49.9a–f). In the latest follow-up, there was no significant loss of correction. Most of these patients in our hospital who were treated with PRSS had one-stage operation without fusion and repeat procedure obviated and spinal movement preserved after removing PRSS when children growth terminated (Fig. 49.10a–i).

Our clinical practice have shown that 20 cases had curve correction continued improvement after PRSS instrumentation in the later growing year (Fig. 49.11a–f); it is an evidence that PRSS has modulating efficiency.

In a recent study, we compared the standing longitudinal radiographs of patients with documented significant progressive and nonsignificant progressive scoliosis after PRSS treatment in 31 consecutive patients with the age less than 10 years old at surgery to determine the relationship between the curve progression and relative amount of disc wedging [22]. Each patient was studied with radiographs which were taken immediately after operation and latest follow-up. Scoliotic major curve angle and the wedge disc angle(WDA)adjacent above apical vertebrae were measured and analyzed. The result showed that in 28 cases without significant loss of correction, the WDA distributed between −10° and 5°. In the other three patients with significant loss of correction, the WDA dis-

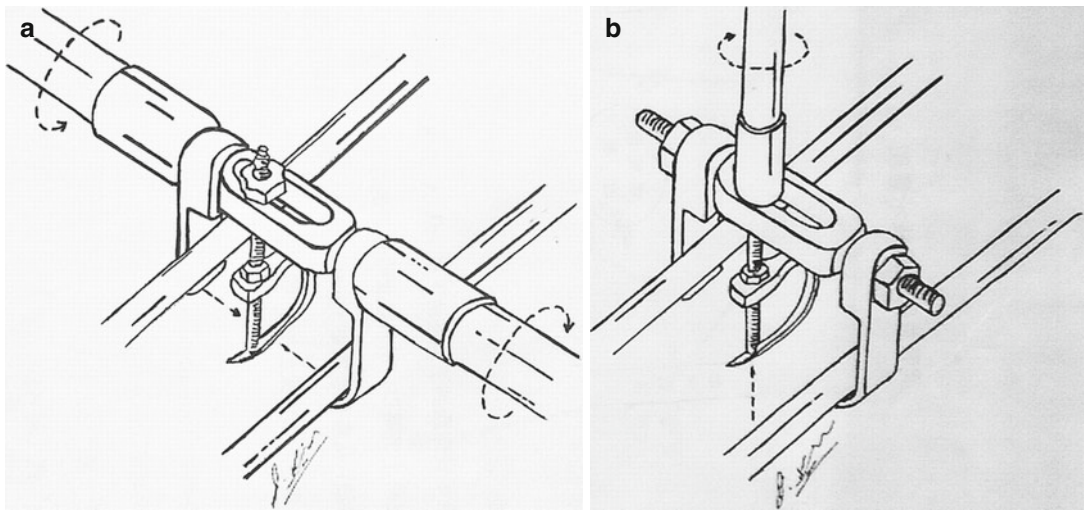

Fig. 49.8 (**a**) additional correction and (**b**) derotation (Courtesy of Jin Lin)

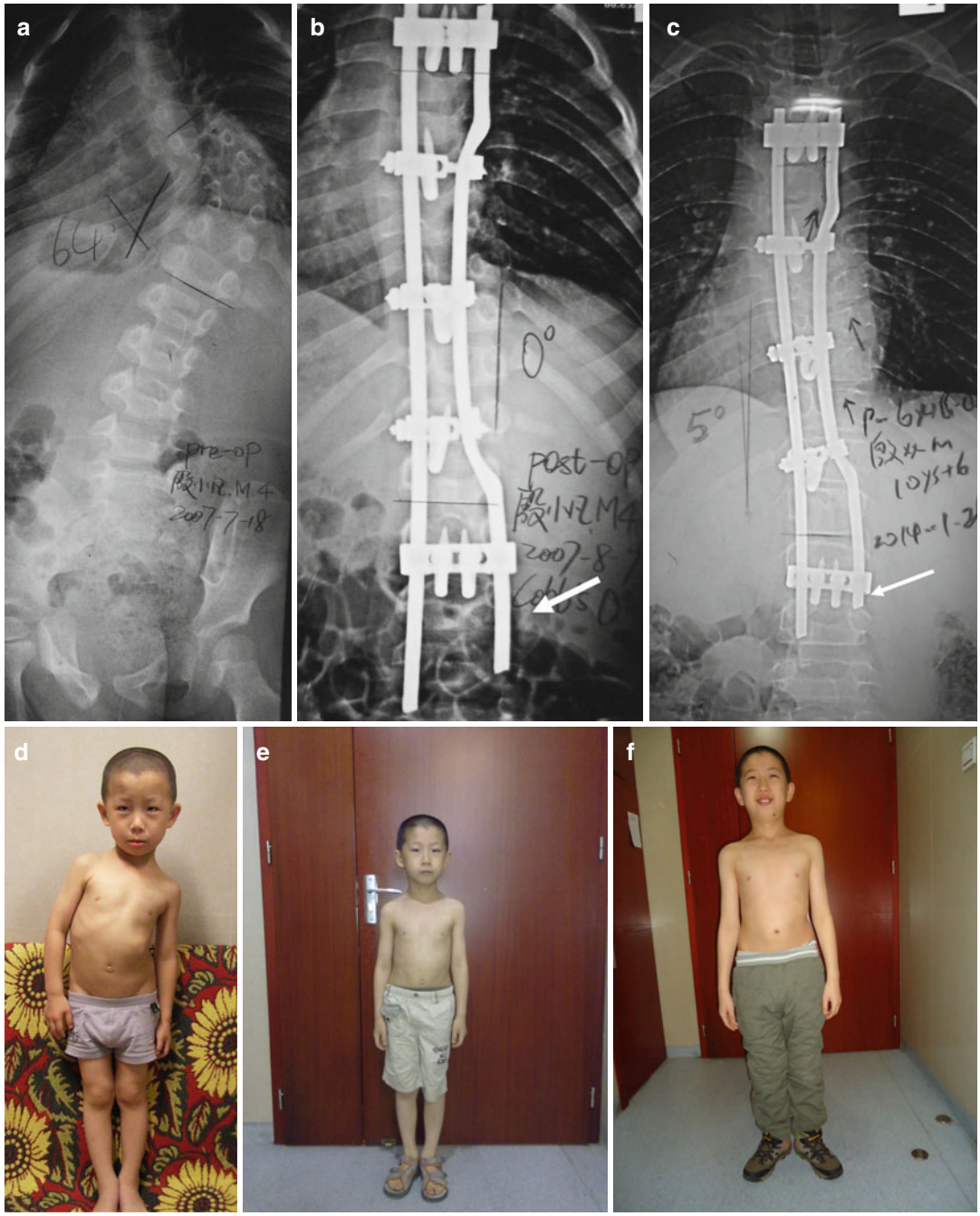

Fig. 49.9 YXF, a 4-year-old boy with a major curve of 64° (**a**) that was corrected to 0° (**b**), at recent follow-up, 6-year post-op scoliotic curve was 5° (**c**), instrumented spinal column increased to 4.5 cm (compare both **b** and **c** at the *arrow* site). Good posture achieved after operation (**d**) 4 years old preop; (**e**) post-op; (**f**) 10 years old

tributed between 6° and 11°. They all received surgical intervention. Desirable curve correction was obtained again. One of them is a 3-year-old boy with a multiple scoliotic curve of 98° (Fig. 49.12a) that was treated with PRSS in 2000-6-9. Scoliotic curve was corrected to 37 (Fig. 49.12b), but imme-

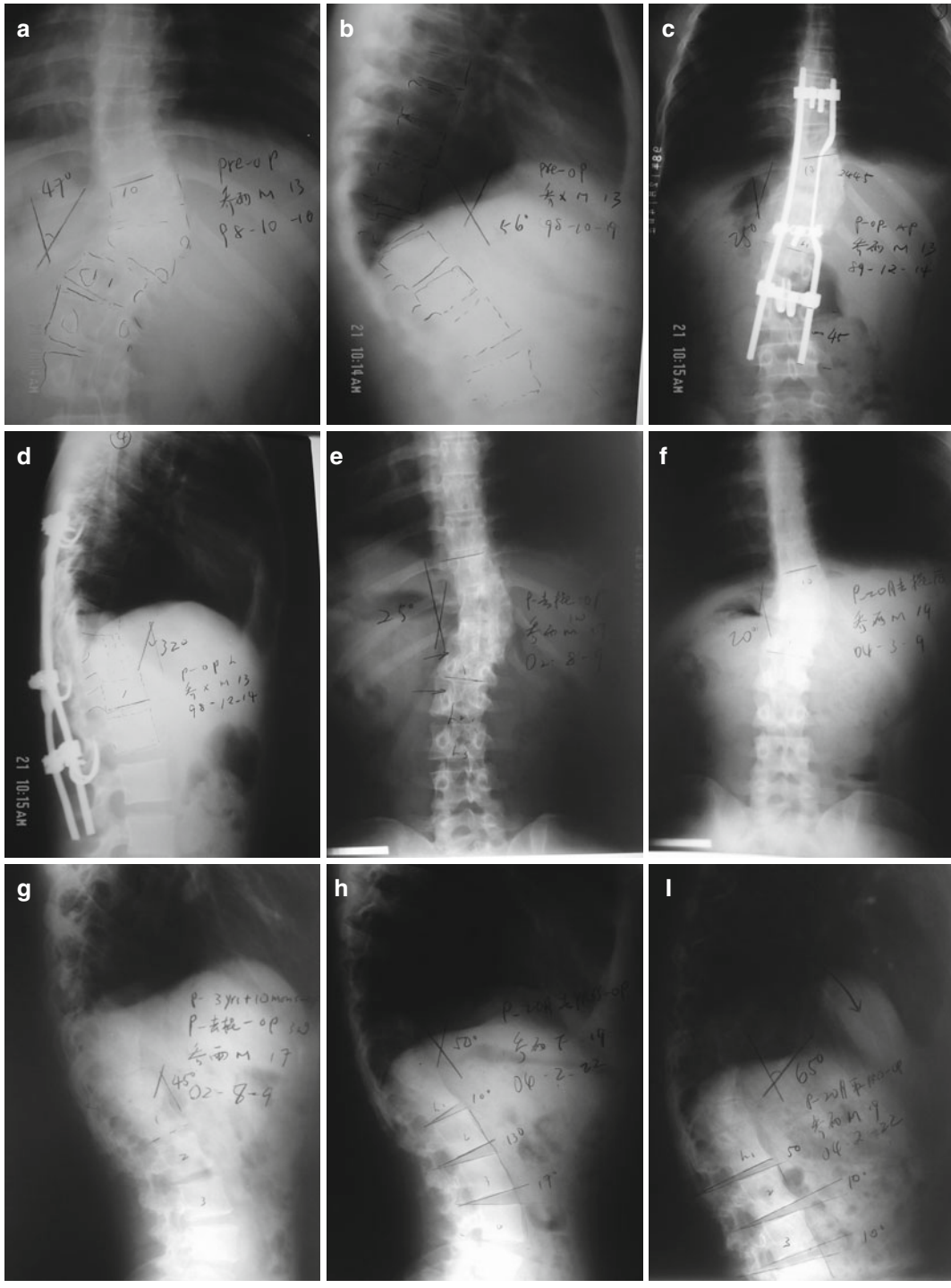

Fig. 49.10 QX, a 13-year-old boy with T10 hemivertebral scoliosis of 47° (**a**) and kyphosis of 56° (**b**); PRSS was used to correct scoliotic curve to 25° (**c**) and kyphosis of 32° (**d**) followed resection of hemivertebrae. At 46 months after surgery, a radiograph taken immediately after PRSS was removed and showed that curve correction maintained at 25° (**e**); another radiograph taken at 20 months after removing PRSS showed the scoliotic curve continued to correct to 20°, and the spinal flexibility improved gradually (**f**). P-removing PRSS – 20 months (**g**). P-removing PRSS. (**h**) Three weeks after removing PRSS l. (**i**) P- removing PRSS – 20 months post-op

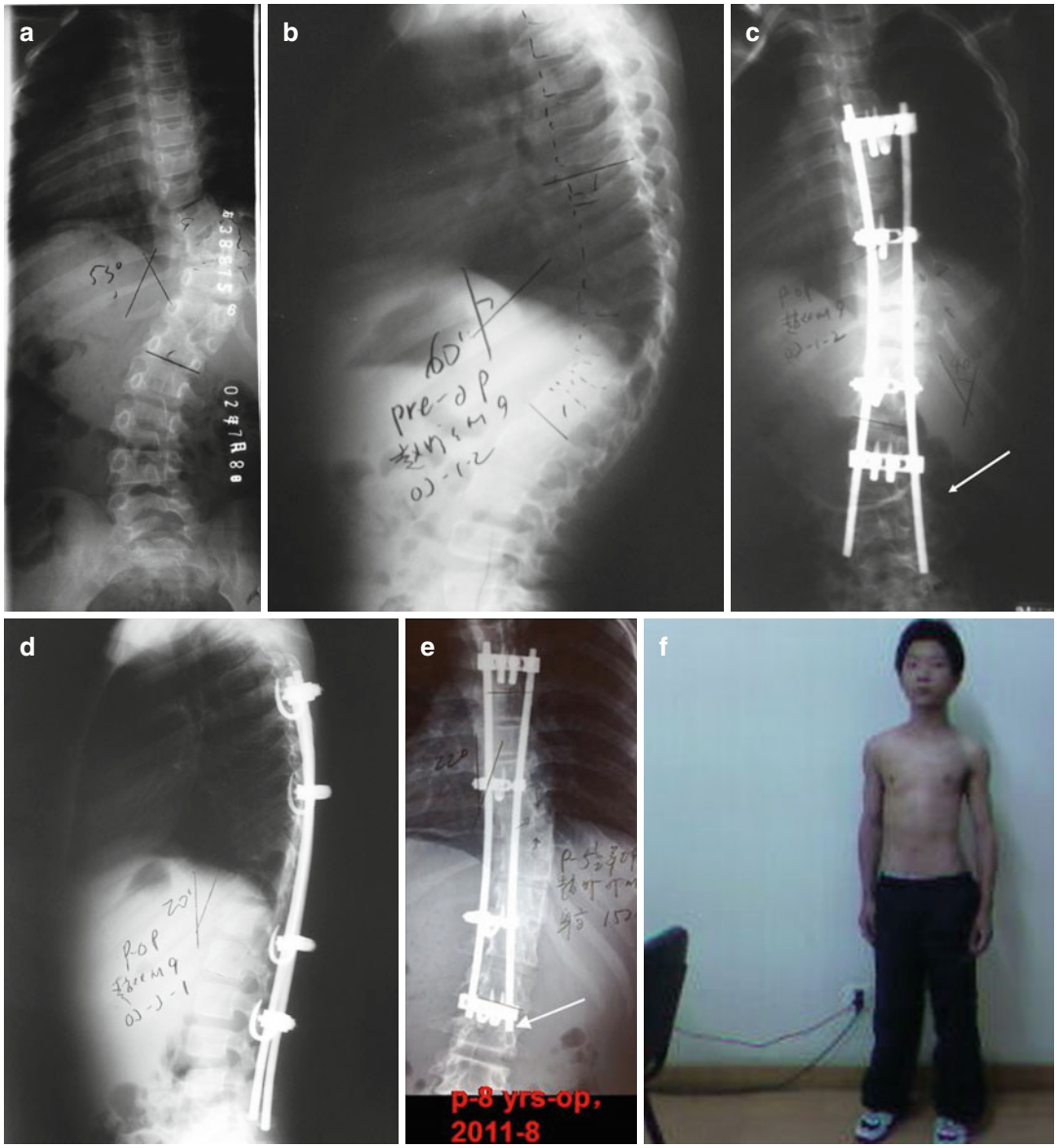

Fig. 49.11 A 10-year-old boy with congenital scoliosis of 58° (**a**), kyphosis of 60° (**b**), and T9 hemivertebrae; posterior operation was performed to remove the hemivertebrae using transpedicular approach and fixed with PRSS. Major curve correction continued improvement from initial 58–40° post-op (**c**), kyphosis of 20° (**d**), and to 22° (**e**) in the resent followed-up 8 years post-op, instrumented segments growth 5.5 cm (compare both **c** and **e** *arrow* site) and (**f**) good posture obtained

diate postoperative x-rays showed the WDA adjacent above apical vertebrae was 10° (Fig. 49.12c black arrow), indicating there is asymmetrical stress on both sides of the scoliotic spine. Seven years after operation when patient was 10 years old, the major curve increased to 63° (Fig. 49.12d). Surgical intervention was given; two PRSS hooks were added in the convex side to increase the compress stress in this side and to increase tensile stress on the concave side; therefore, major curve was corrected to 40° (Fig. 49.12e), and the WDA was reduced to −5° (Fig. 49.12f; see arrow), indicating the initial asymmetrical mechanical loading was reversed. X-ray showed bony fusion developed on

the convex side over the apical area (Fig. 49.12g, white arrow); the major curve was gradually improved to 28° after second operation. In the recent follow-up, when patient was 17 years old, radiographs showed the major curve correction maintained at 28° (Fig. 49.12h), and 3-D CT showed bony fusion on the convex side clearly (Fig. 49.12i, white arrow), while in the concave side, disc space is still intact (Fig. 49.12i, yellow arrow), indicating that there is a potential growth ability in the concave side. Good posture appearance (Fig. 49.12j/k/l) was seen.

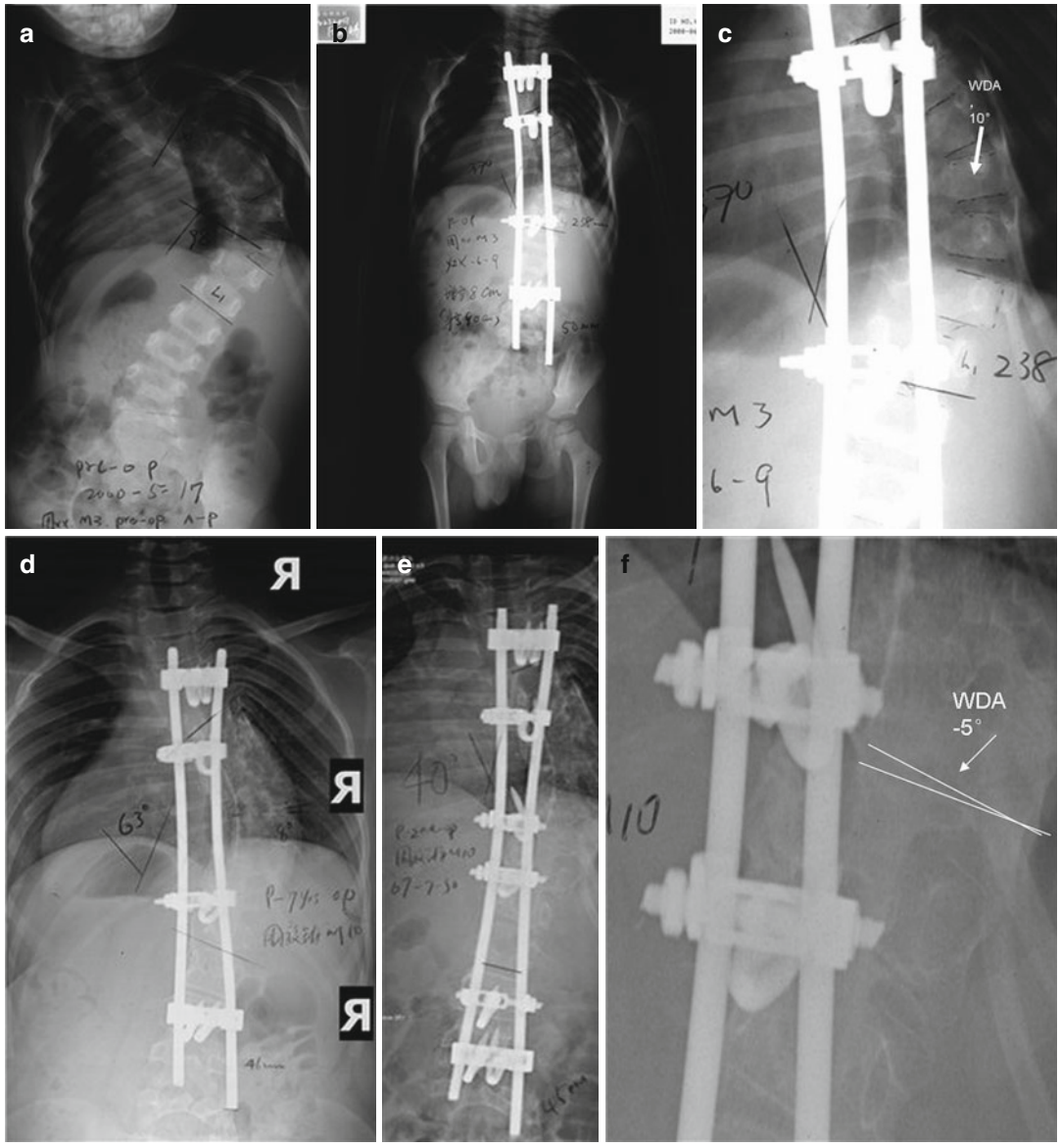

Fig. 49.12 Zu xx, a 3-year-old boy with scoliotic curve of 98° (**a**), post-op scoliotic curve was 37° (**b**). The WDA was 10° (**c**; see *arrow*). Seven years after operation, the scoliotic curve was 63° (**d**). After second operation, scoliotic curve was reduced to 40° (**e**). WDA was reduced to −5° (see *arrow*). (**f**) X-ray showed bony fusion on convex side (**g**, *white arrow*). In the recent follow-up, 14 years after operation, scoliosis was 28° (**h**) and curve correction maintained; 3-D CT (**i**) showed bony fusion on the convex side clearly (*white arrow*) and good posture obtained (**j**) 3 years old preop; (**k**) post-op; (**l**) 14 years old

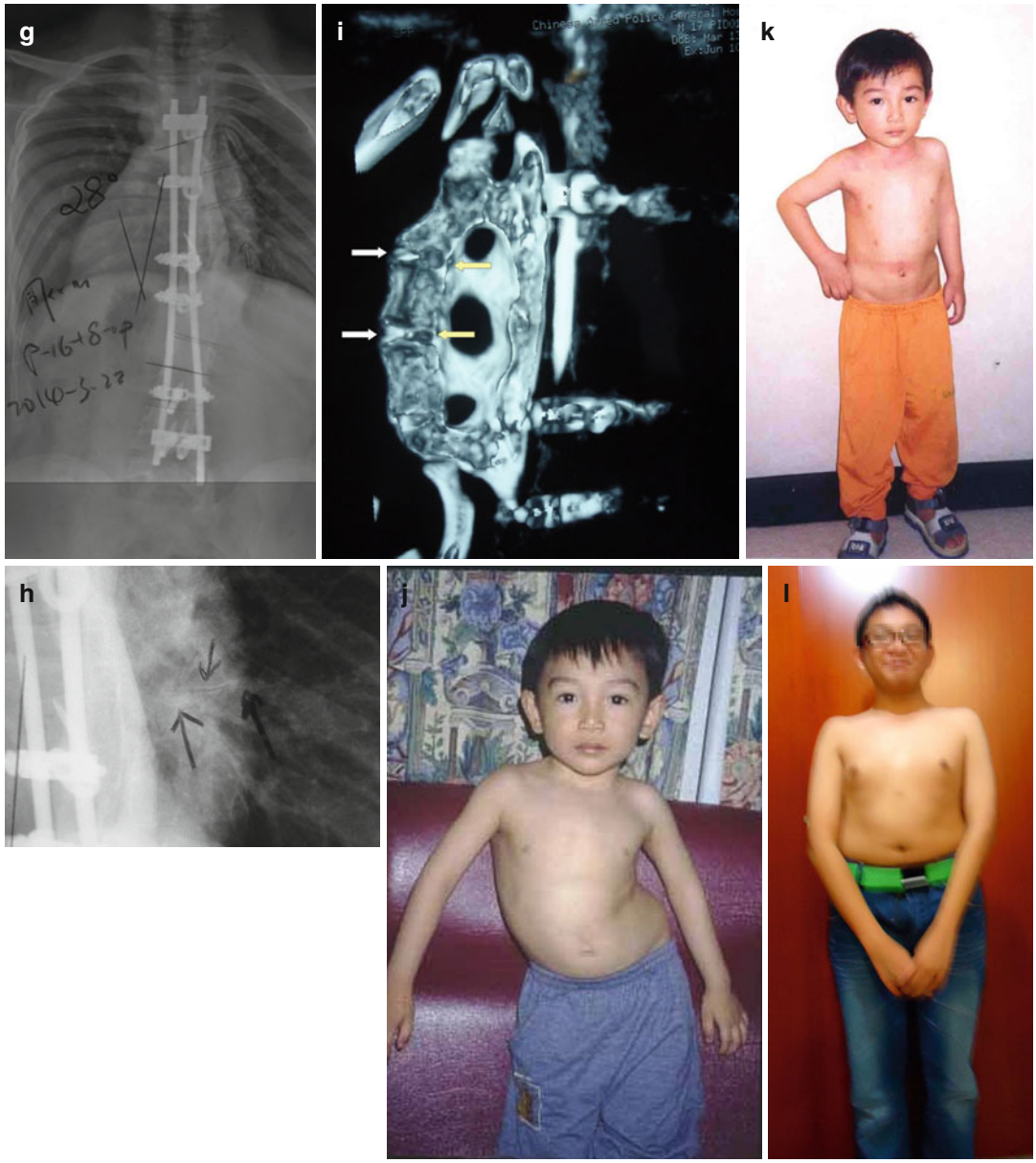

Fig. 49.12 (continued))

It is probably that WDA reaches a certain magnitude (more than 5°), the laws of mechanical modulation growth take place, and then, curve progression occur in the later growing years which could be gradually corrected if appreciate approach (such as PRSS) was applied to alter the IVDA for reversing the Hueter-Volkmann effects [23, 24].

49.4.3 Measurement of the Growth of the Instrumented Spine

The growth of the instrumented spinal segments can be determined by measuring the upward shift of the lower end of the plate rod below the lower connector (see Figs. 49.9b, c and 49.11c, e). An average of 13.13 mm spinal lengthening under

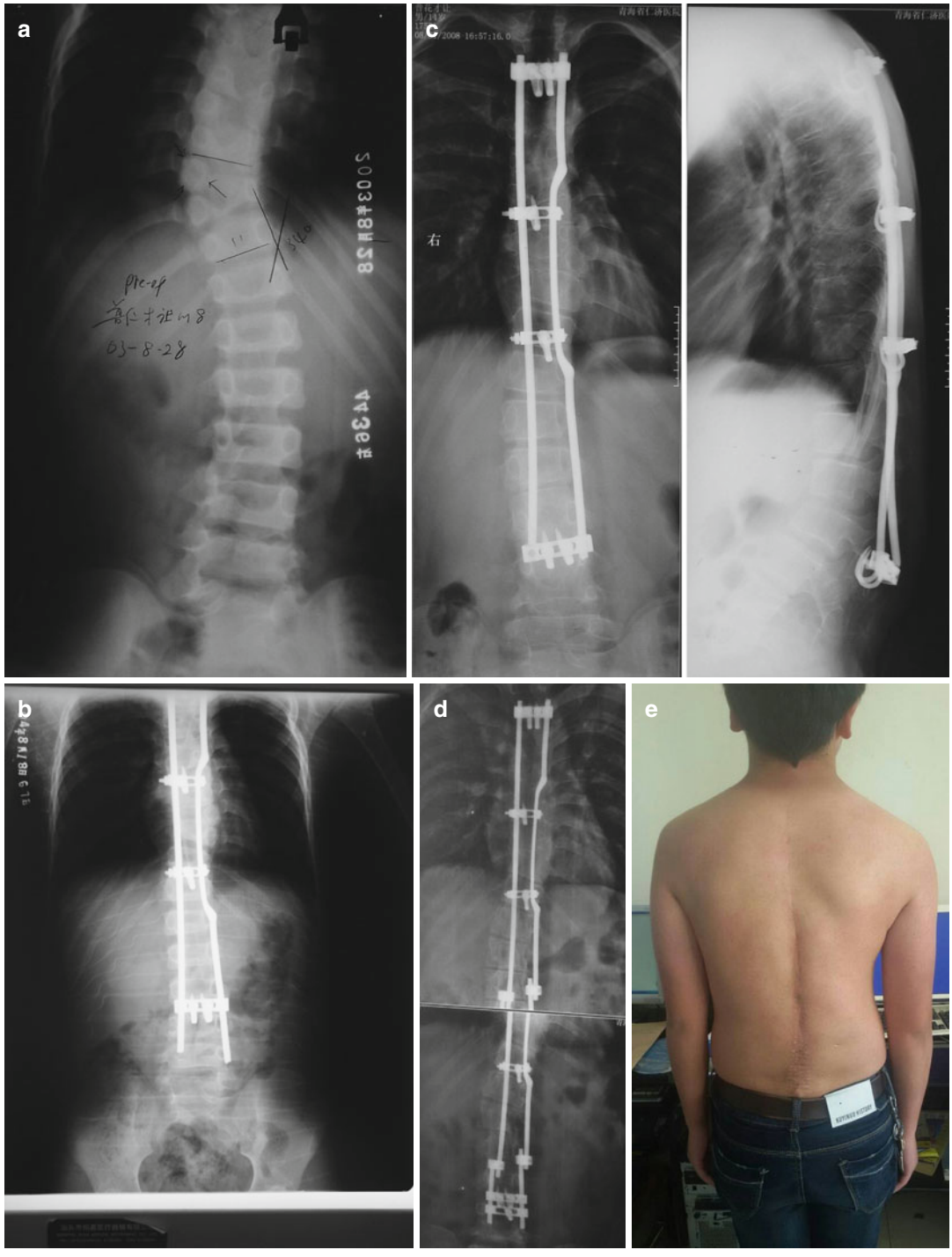

Fig. 49.13 PRCY M 9, hemivertebral scoliosis: preop 34° (**a**), post-op 0° (**b**). Another radiograph taken 5 years later shows curve correction maintained, the lower end of rods sliding out of the lower connector (**c**), which was elongated with tandem connector (**d**). Good posture was seen (**e**)

instrumented segments was achieved, the longest one being 5.6 cm.

49.4.4 Complications

There were no intraoperative complications. To date, no infection or neurological deficit has occurred. Skin irritation was seen in four cases; two cases had skin pressure sores in the early period after operation. For this reason, low-profile connector is designed to reduce the prominence of implants. When the growth of the instrumented spinal segments exceeds the length of the lower end of the rods to elongate, a small operation is needed for connecting a rod by a tandem connector on each side (Fig. 49.13).

Our studies have introduced a new idea for the treatment of children scoliosis, that is, a new device should have no tethering on the concave side. Correction mechanism for growing scoliosis should be changed from distraction on the concave side to the lateral sidewise push on the convex side and from static lording to dynamic lording.

Conclusion

The PRSS technique which dispenses with spinal fusion and allows extension along with the children's growth is able to provide and maintain desirable correction of scoliosis in the later growing years due to its modulating character. This new technique is an effective instrumentation for correcting scoliosis in growing children, especially for EOS. In order to assess the real merit of this technique, it is necessary to collect long-term follow-up period until the completion of bony growth.

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50 Complications Following Distraction-Based Growth-Friendly Surgery in Early-Onset Scoliosis

John B. Emans and Behrooz A. Akbarnia

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Key Points

- Growth-friendly surgery in EOS is associated with a high rate of complications.
- A variety of factors are responsible for complications in EOS surgery, including the age at surgery, etiology, multiple surgical procedures, and technical factors such as anchor stability and rod location.
- Optimization of patient health, nutrition, and surgical technique including soft tissue handling will minimize complications.
- Early recognition of complications and appropriate and timely treatment is necessary for a successful outcome.
- Spontaneous posterior intervertebral fusion can occur with growth-friendly treatments and can limit further spine growth or contribute to progressive deformity.

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50.1 Introduction

Early-onset scoliosis (EOS) is now defined as spinal deformity diagnosed before the age of 10 years [1]. Unlike adolescent spinal deformity, untreated progressive spine deformity in this age group can cause significant health problems for young children and later in their adult life, particularly significant pulmonary compromise. Any treatment of EOS must focus not only on the spine deformity but also on the development of the thorax to improve the child's long-term quality of life. The surgical options for EOS are complex. Complications are a common, seemingly inevitable part of the surgical treatment of EOS. Children with EOS may also have associated morbidities and are at risk for even more complications. Most importantly pulmonary growth and development can be affected when spine and chest deformities start very early in life.

The complexity and potential complications associated with surgical treatment of EOS begin with the varied etiologic diagnoses for EOS. EOS may be associated with congenital vertebral anomalies, bone dysplasias, connective tissue disorders, neuromuscular disorders, or idiopathic spinal deformities. The list of known etiologies is long, each with its own set of unique potential problems. Additionally EOS can be associated with lordosis, kyphosis, or any combination. All these variations in etiology and deformity share the same potential problems associated with untreated EOS. Lung development and pulmonary function are at risk [2–4]. Campbell has termed the potential pulmonary compromise associated with severe EOS as "thoracic insufficiency syndrome" defined as "the inability of the thorax to support normal lung growth and respiration." [5] Spine length is at risk. In addition to spinal deformity, which detracts from spine height, EOS has previously been treated with early fusion, to the detriment of final spine height [2–4]. Spine mobility is also at risk in EOS and less well documented [3]. Based on these adverse outcomes for EOS, objectives in the treatment of EOS should include the maximization of pulmonary function, spine length, and residual mobility, while minimizing hospitalizations, family burden, cost of care, and complications.

Surgeons and families who are facing these decisions should balance the risk versus long-term benefits before starting any treatment. Because of significant complications of untreated spine and chest wall deformity in this age group, treatments, even with higher complication, are often chosen to improve the natural history. Some of the difficulty in treatment is due to the very nature of the growing spine and chest wall and the need for periodic revision, while other difficulties represent true complications. Understanding the nature of the difficulties associated with the treatment of EOS as well as possible complications will allow early detection and timely management of complications.

The goal of treatment in children with EOS is to control the deformity, allow spinal and chest wall growth and lung development, improve pulmonary function, and therefore provide a better quality of life. Treatment options such as casting or bracing are commonly used methods for early intervention, but when the deformity is progressive and severe, surgical treatment is often indicated. Definitive spinal fusion with or without instrumentation has been used as standard approach for a long time, but fusion results in a short trunk with possible lung underdevelopment and subsequent pulmonary complications if performed while the child is growing [2–4]. There is also a possibility of progression of spinal deformity through adding on to the deformity in unfused segments or within the fusion by the "crankshaft" phenomenon [6, 7].

Growth-friendly procedures, which allow or encourage spine and chest growth rather than inhibit them, have gained popularity in the treatment of EOS. Skaggs divided these procedures into distraction-based, growth-guided, and tension-based procedures [8]. The most common spine-based distraction techniques are "growing rods" (GR) [9], and rib-based distraction techniques include the use of "vertical expandable prosthetic titanium rib" (VEPTR) [10] with or without expansion thoracoplasty. These techniques follow the concept of periodic distraction to allow the spine and thorax to grow. Like all

other procedures for EOS, growing rods and VEPTR have been shown, each in their own way, to have a high risk of complication. The high rates of complication may be the result of both multiple surgeries and the presence of critical health issues in these patients. In this chapter we will discuss common complications and necessary actions to decrease the risk of complications for both growing rods and VEPTR procedures. Growth-guided procedures are discussed in other chapters, and no long-term data is available for tension-based techniques in very young children.

50.2 General Complications in Growth-Friendly Techniques

Frequent surgeries increase the risk of general complications caused by surgery, such as undesired events due to anesthesia, postoperative period, and hospitalization. Other complications include failure to successfully control progressive spine deformity, ending growth with an unacceptably short or stiff spine, an unacceptably deformed, stiff or small chest, or failing to avert thoracic insufficiency syndrome as described by Campbell. Catching up and keeping pace with growth demands repetitive lengthening procedures. Current attitude has evolved into periodic lengthening to "drive" the growing spine [11]. Lengthening schedules vary depending upon the extent of the spine instrumented and growth rate but may be as frequent as every 4 months in very small children, every 6 months in most children, and every 9 months when only a short section of spine is involved [12]. Children exposed to multiple trauma are also more likely to experience severe symptoms of posttraumatic stress disorder (PTSD) and depression than those who experience a single event [13]. Additionally from a financial point of view, more time, surgeries, and complications lead to enormous direct and indirect cost for the patient's family. To achieve satisfactory results with these techniques, the treatment period is often long and may take many years to complete.

50.3 Complications in Growing Rods

Traditional growing rod (TGR) treatment necessitates regular rod lengthening; therefore, children receiving initial growing rod implants at a younger age are likely to undergo more procedures than children initially treated at an older age. Bess et al. [14] reported a 13 % decrease in the likelihood of suffering a complication for each year increase in age at initial surgery. Additionally, it is possible that younger children are less healthy and have more severe scoliosis than older children and accordingly are at greater risk for complications. Klemme et al. [15] reported multiple complications in a group of 67 children, including one death, but felt fusion without instrumentation is appropriate for severe EOS. In 2002, Mineiro and Weinstein [16] questioned the worthiness of growing rod techniques. In their report on 16 patients, the complication rate was notably high. Implant-related complications were the most common complications in their series, with rod fracture as the most common implant complication. They also reported skin breakage, wound complication, and misalignment in their patients. Most recently Bess et al. [14] reported complications in 897 growing rod surgeries from Growing Spine Study Group (GSSG). Overall complication rate per procedure was 19 %. Fifty-eight percent of patients had minimum of one complication (mean of 1.2 complications per patient). Fifty-eight percent of complications were managed at the time of planned surgery. Analysis of total complications demonstrated a linear decrease in survivorship (complication-free rate) for each surgery performed, indicating increasing complication rates with increase number of surgeries. At seven procedures, there was a 49 % chance of having a complication. At 11 procedures, the complication risk increased to 80 %. They concluded that high complication rate in this group of patients is a function of sustained treatment duration and the number of procedures required during the treatment period. The new technologies, such as MCGR, have reduced the number of surgeries since lengthenings are done noninvasively.

However, it has not affected all complications risks inherent with any surgical intervention for children with EOS.

50.3.1 Complications Related to Foundation Level Fusions

In traditional growing rod technique, limited fusion is carried out within the two upper and lower foundations [17]. This is usually performed at two or three adjacent levels at each end of the construct. Like any other fusion techniques, although very low, there is always a possibility of nonunion within the foundation levels. Foundations are usually designed to carry on distraction load. Motion within foundation anchors can increase the failure rate, which includes wide variety of complications, from screw/hook loosening, implant prominence, to rod fracture.

There is also a risk of unwanted fusion of adjacent levels if subperiosteal exposure extended beyond foundation levels.

50.3.2 Skin-Related Complications

Multiple surgeries, mostly through the same incision site, make the skin tissue susceptible to infection and other skin problems. Impaired nutritional status also can increase the risk of skin-related complications and should be attended to preoperatively. Tissue handling is extremely important during these multiple surgeries for providing adequate coverage and to reduce skin complications. Skin retraction should be minimized, and skin flaps should have developed for full-thickness skin coverage. At closure, skin should be under minimal tension. Occasionally, flaps are necessary for coverage. Implant prominence may be unavoidable in very thin and small child. Postoperative padding may be helpful to minimize pressure and possible skin dehiscence. Skin has to be carefully watched for any redness and signs of skin breakdown, and if seen, aggressive treatment should immediately be initiated. Applying the rod submuscular is also shown to decrease wound complications [18].

50.3.3 Superficial and Deep Wound Infection

This complication may affect the treatment outcome significantly by increasing the number of unplanned surgeries and difficulties of controlling infection. Implants are an essential part of the treatment, and implant removal in deep wound infections should be the last resort. The rods can usually stay if the infection is diagnosed early and treated with debridement and IV antibiotics. Occasionally with the dual rod technique, one rod can be removed if prominent, with plans for reinsertion later (Fig. 50.1a, b). There is no data available at this time for the length of antibiotic therapy needed in a child with EOS following postoperative deep wound infection. With superficial wound infections, more aggressive surgical intervention and skin closure can reduce the chance of becoming a deep wound infection [19]. Kabirian et al. [19] retrospectively studied the prevalence of deep postoperative surgical site infection in a multicenter growing spine database. There were 379 patients treated with traditional growing rod technique and followed for a minimum of 2 years. Twenty-two (52 %) of forty-two patients who developed deep infection had removal of implants to control the infection. Nine of the twenty-two had only partial removal, and routine lengthenings could continue. Thirty-one (74 %) of the forty-two patients with deep infection had completed the growing rod treatment or were still undergoing lengthenings at the latest follow-up.

50.3.4 Implant-Related Complications

Implant-related complications are the most common complications in growing rod surgeries. These include rod fracture, anchor failure, or prominent implant, which can cause skin breakdown and even infection (Figs. 50.2, 50.3, and 50.4). Among the implant-related complications, rod fractures are the most common problem.

Yang et al. [20] reported the GSSG experience of 86 rod fractures in 46 patients. The overall rate of rod fracture was 15 %; however, the risk was

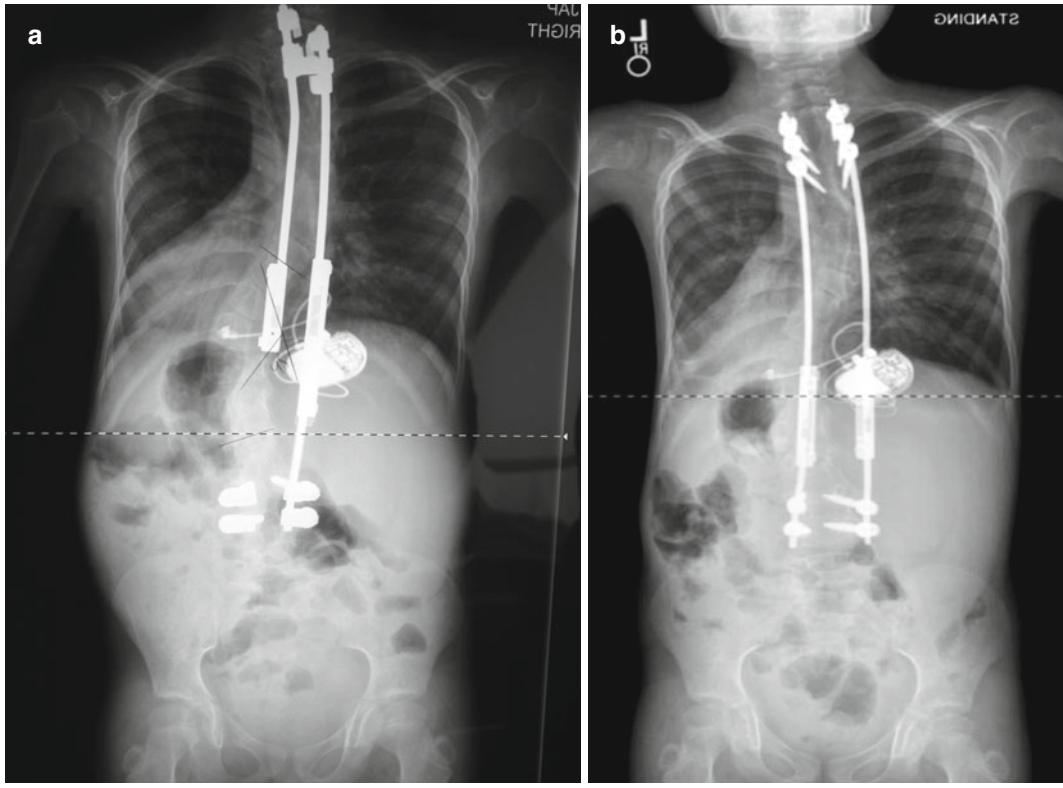

Fig. 50.1 (**a**, **b**) A 9-year-old boy with idiopathic EOS treated with dual growing rods. Seven years after beginning treatment, he had a deep wound infection and was treated with irrigation and debridement. (**a**) The implant removed partially and temporarily on one side to help the healing process. (**b**) After complete recovery, the revision surgery was done to complete the treatment with dual growing rod technique

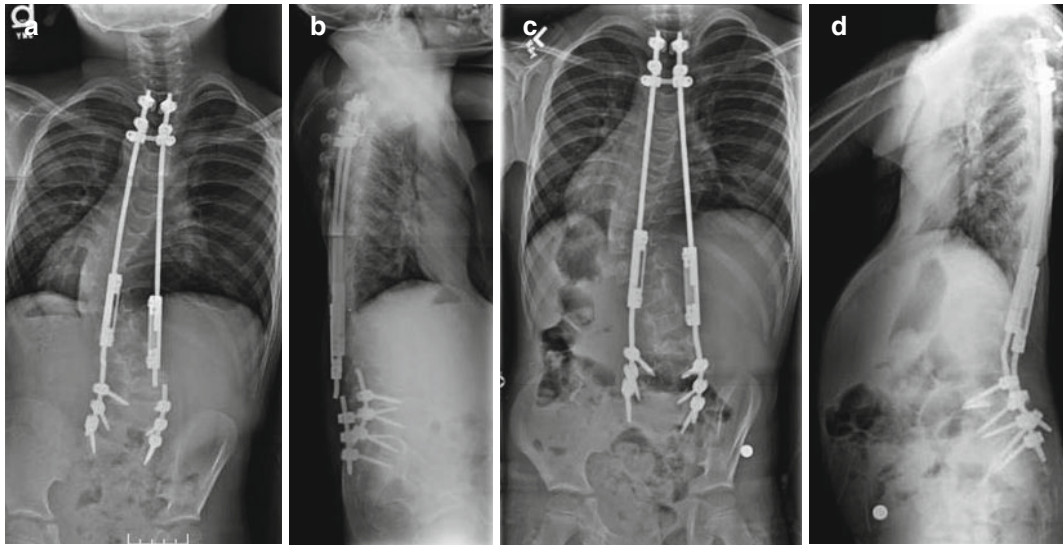

Fig. 50.2 A 6-year-old boy with Beal's syndrome treated with dual growing rods for progressive scoliosis presented with low back pain. (**a**, **b**) Posterior-anterior (PA) and lateral radiographs confirmed rod fracture. (**c**, **d**) Revision surgery was done and the broken rods were replaced (Courtesy of Burt Yaszay, MD San Diego)

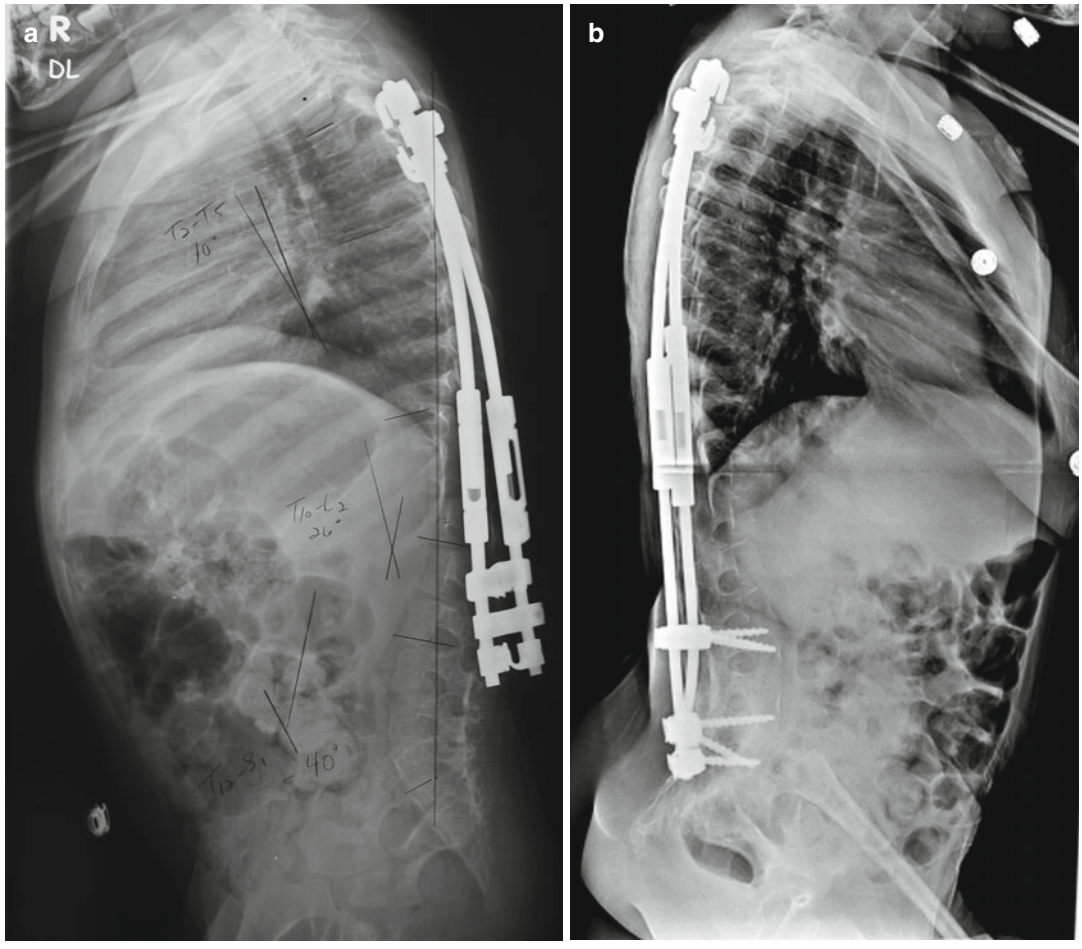

Fig. 50.3 (**a**) Hook dislodgement in the lower foundation, in a patient with EOS treated with dual growing rods. (**b**) Hooks were replaced with pedicle screws

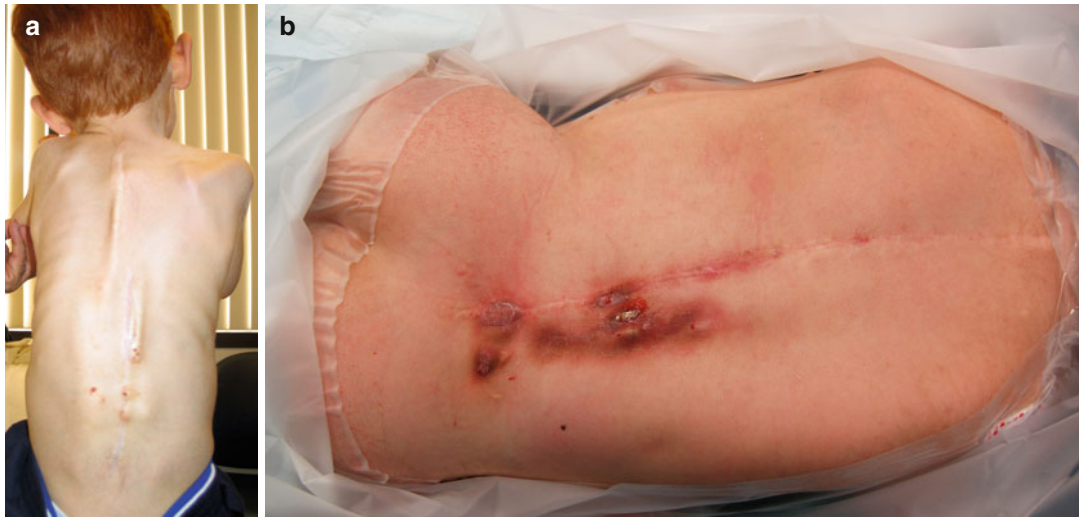

Fig. 50.4 (**a**) Prominent implants in an 8-year-old patient treated with dual growing rods. (**b**) Prominent implants can cause skin breakage and wound infection if not treated properly

increased in patients with single rods, history of previous fracture, small diameter rods, stainless steel rods, proximity to tandem connectors, and smaller tandem connectors and in ambulatory patients. The rate of rod fracture did not correlate with anchor type or degree of the curve. It is advised that replacing the rod may be a preferred strategy over connecting the broken rods.

Asymptomatic implant failure may be revised at the time of planned lengthening surgery. If only one rod in a dual rod construct is broken, it is recommended to change both rods if possible to prevent early fracture of the second rod.

There are changes of the anchor sites that are expected because of normal spinal growth and are not considered true complications. These include hook and screw migrations due to vertebral growth, requiring revision, which can be usually performed at the time of planned surgeries.

50.3.5 Alignment Complications

It is important to obtain and maintain acceptable coronal and sagittal alignment at initial surgery. Multiple studies have shown improvement of coronal and sagittal plane deformity after initial TGR surgery in both single and dual GR techniques [21]. To avoid proximal junctional kyphosis, the rods should be contoured into kyphosis at the top of the construct and the interspinous ligaments kept intact as much as possible. The upper foundation is usually extended to T2 and occasionally even higher to reduce the risk of proximal junctional kyphosis. This is especially true in children with non-idiopathic scoliosis. If there is thoracic hyperkyphosis, the rods should be contoured into kyphosis since excessive correction may lead to implant/anchor site failure postoperatively. The tandem connectors should be placed at the thoracolumbar junction and not at the lordotic or kyphotic segments of the spine unless it can be contoured. Short instrumentation, especially in patients with non-idiopathic scoliosis, should be avoided to prevent adding on to the curve as the child grows. In cases of MCGR, the actuator cannot be contoured, so the rod proximal to it needs to accommodate for the straight segment. If more rod length is needed for contouring, a smaller actuator (e.g., 70 mm) can be used which leaves more rod for contouring (see Chap. 47) [22, 23].

Another possible complication is curve decompensation. If the levels are selected carefully and initial instrumentation done accordingly, curve decompensation is unlikely. In the original report on dual growing rods, Akbarnia et al. [17] reported only two cases of curve decompensation following final fusion, both treated by extension of the instrumentation and fusion (Fig. 50.5).

50.3.6 Neurological Complications

Neurological complications are uncommon in growing rod surgeries without associated procedures. Neurological deficit may occur with excessive distraction or with significant deformity correction. The incidence of intraoperative neurological injury is 0.1 % in index surgeries, revision, and lengthenings [24]. Intraoperative neuromonitoring is a reliable way to monitor changes during surgery and is recommended for primary insertion and exchanges, but not used routinely by everyone for lengthening [24].

Careful lengthening to avoid over distraction at initial surgery and at lengthening procedures will reduce the risk of complications. In revision and exchange surgeries using dual rods, it is helpful to maintain a baseline length by keeping one side of the construct intact. Two rare cases of delayed neurological event are reported [25, 26], and both recovered after immediate shortening of the rods. Therefore, the child should continue to be closely monitored during the immediate postoperative period for development of any late neurological deficit.

It is necessary to follow proper surgical technique to reduce the rate of complications and to achieve the best long-term results. This is especially important during the initial surgery to pay special attention to the details of selection of the levels, exposure, anchor insertion, and rod contouring and placement to reduce the complication rate in these complex surgical procedures.

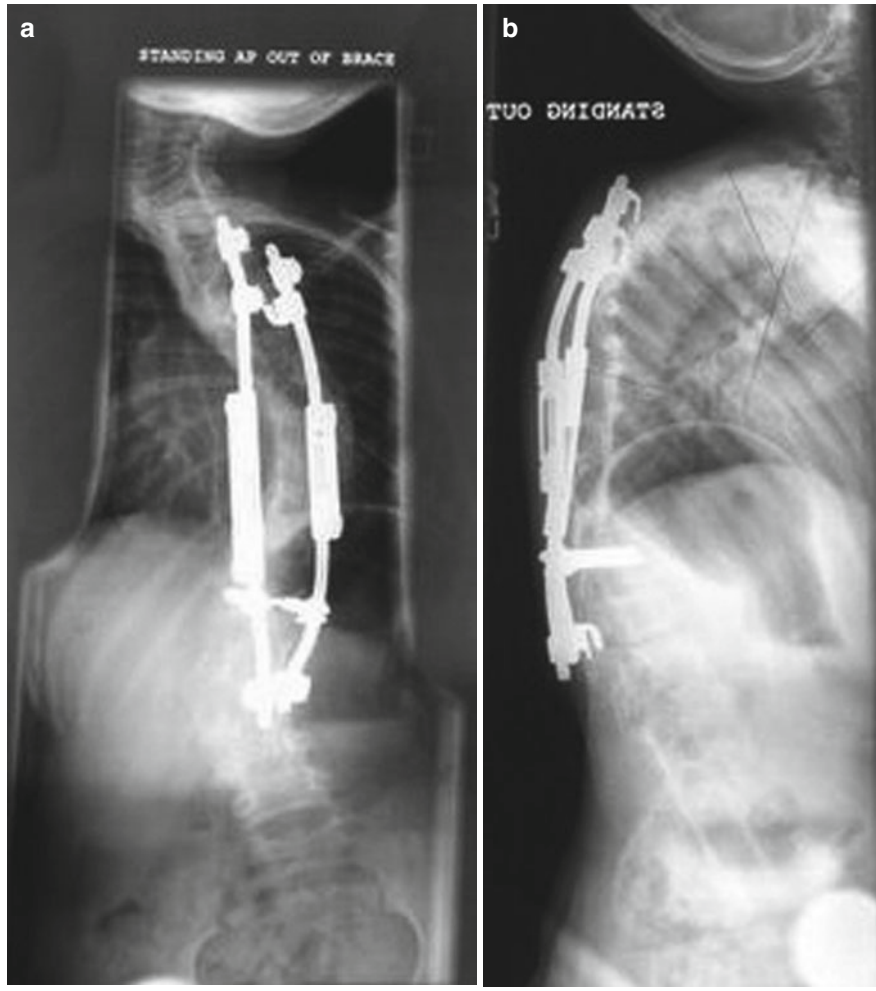

Fig. 50.5 (**a**, **b**) Curve decomposition in an early-onset scoliosis patient treated with a short dual growing rod construct. If the levels are selected carefully and initial instrumentation done accordingly, curve decomposition is unlikely

50.4 Complications in VEPTR and Rib-Based Distraction Devices

VEPTR and growing rods are both distraction-based growth-friendly techniques and therefore share many of the same complications associated with repeat surgeries [27] The advent of self-lengthening devices (MAGEC) may result in fewer complications owing to the decreased number of surgeries. Some types of complications are unique to rib-based distraction and VEPTR [28–31]. Familiarity with VEPTR-related complications will help the surgeon choose the most satisfactory growth-sparing surgical technique for a particular EOS and proactively avoid complications as much as feasible.

50.4.1 Rib-Based and VEPTR Anchor Point Problems

The cephalad attachments of VEPTR devices are circumferential rib "cradles," while the caudal attachments may be ribs, spine, or pelvis [10]. Non-VEPTR rib-based techniques generally use upward-directed distraction spine hooks as rib anchors beneath the caudal surface of the ribs.

Acute rib anchor failure is usually rib cutout or fracture and associated with excessive stress or poor quality of rib bone such as bone dysplasia or nutritional osteopenia [32] (Fig. 50.6). Preoperative assessment of bone density and treatment of osteopenia or osteoporosis will minimize this complication. Rarely acute dislocation of the entire rib at the costovertebral articulation occurs if excessive distraction force is applied to the rib. Acute loss of rib fixation is best avoided by distributing distraction force to multiple anchor points ("load sharing"), either by encircling more than one rib (VEPTR I) or staggered multiple anchors (VEPTR II) [33] (Fig. 50.7) or similar configurations with multiple spine hooks used on the ribs. Where appropriate, an expansion thoracostomy [28] can diminish the necessary distraction force needed to achieve control of

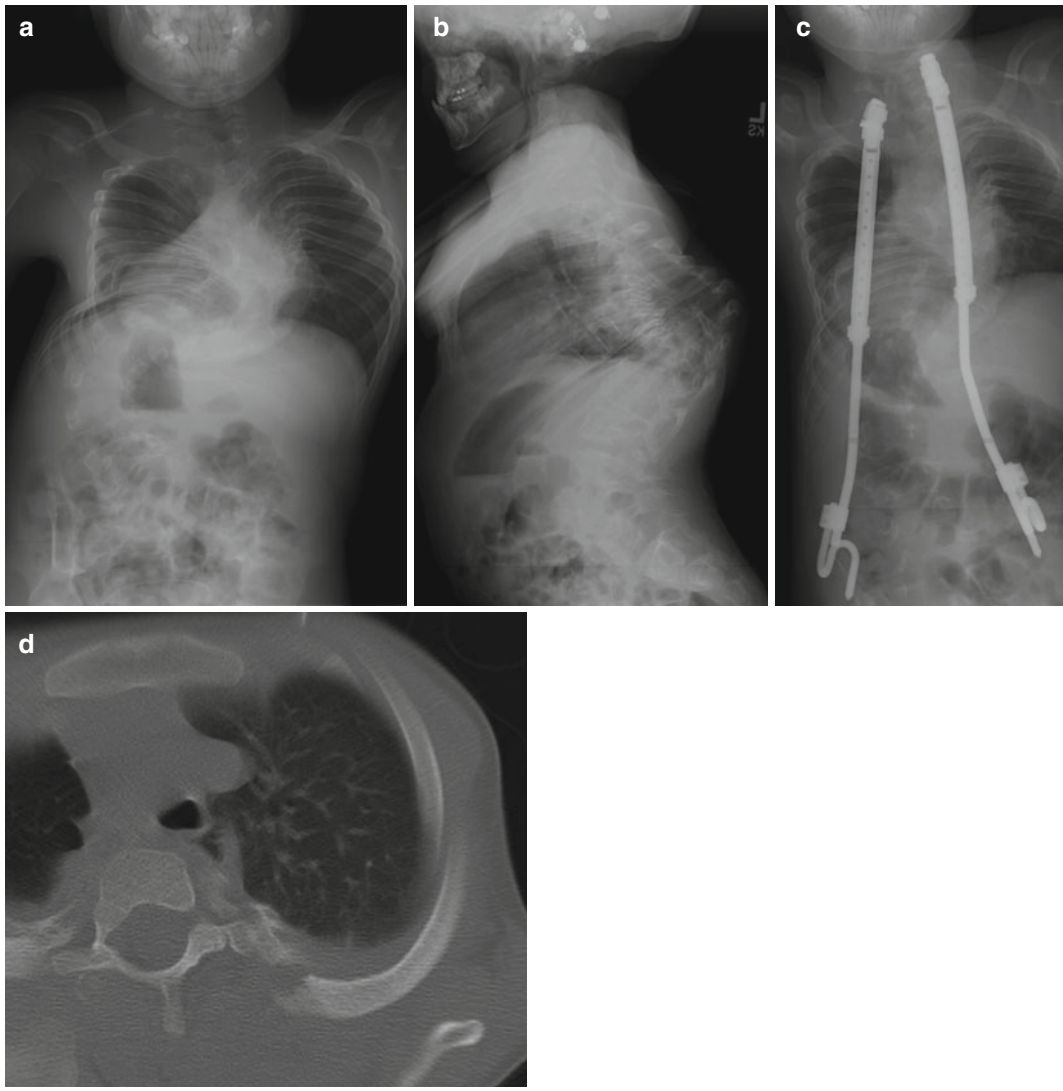

Fig. 50.6 Acute rib fracture in VEPTR rib-based distraction. An 11-year-old with uncharacterized bone dysplasia and severe kyphoscoliosis (**a**, **b**) was treated with 2 months of halo gravity traction followed by bilateral rib-to-pelvis VEPTR devices. Postoperatively he turned in bed and felt pain. X-rays (**c**) and CT (**d**) revealed bilateral rib fractures and cephalad displacement of both rib anchors. Revision to new rib anchor points restored fixation but with less correction (**e**, **f**). Bone dysplasia and osteopenia contributed to weakness of the rib attachments

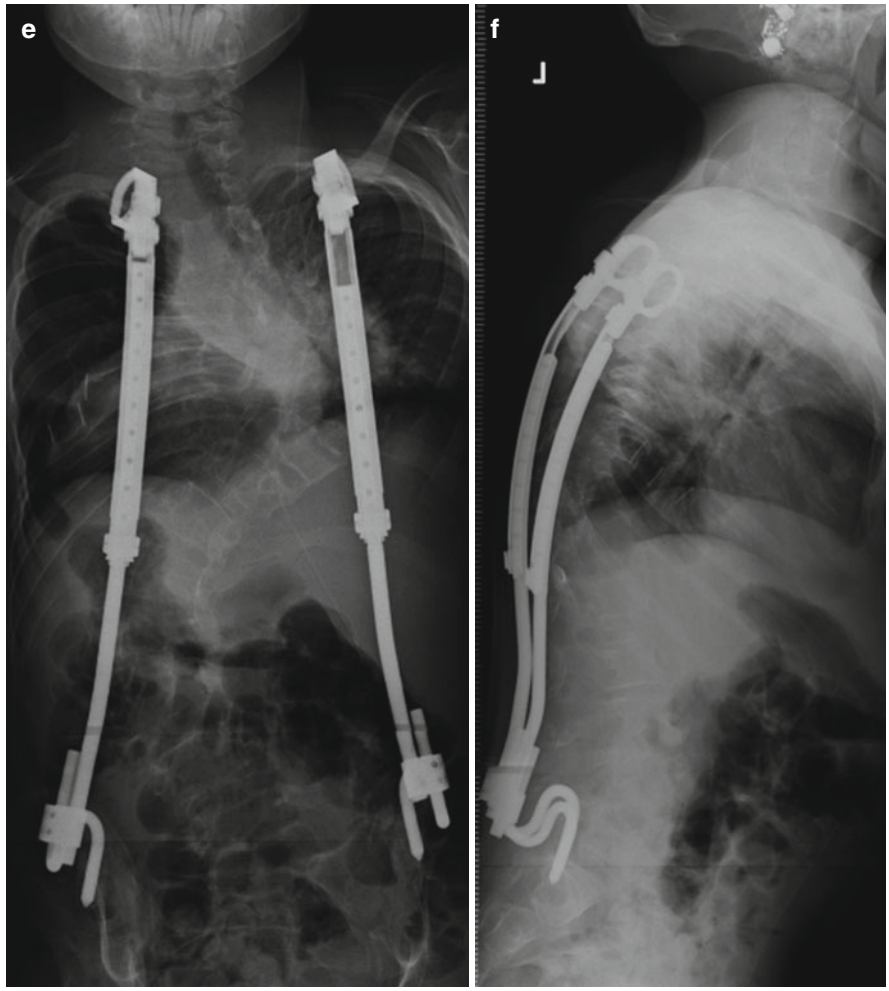

Fig. 50.6 (continued)

combined chest and spine deformity. Kyphotic deformities corrected by cantilever forces applied via the upper rib attachment may fail acutely if too much deformity is corrected. Chronic migration or "drift" of rib anchors is a common phenomenon, particularly in rib-to-spine more than rib-to-rib distraction constructs [10, 28–31, 34, 35]. Campbell [31] reported drift of rib attachments in 7 of 27 patients. Usually rib anchor drift is not functionally significant, as the drifting rib attachment gradually pulls with it a solid bone attachment and remains functionally connected to ribs, but some lose functional connection and require revision, which can usually be done at the time of a planned lengthening. Distal migration of VEPTR iliac S-hooks is common over time, particularly in ambulatory patients or unilateral devices [36, 37]. Distal drift of an S-hook can be troubling, as the S-hook becomes buried in the ilium, gradually drifting toward the acetabulum. Earlier revision of drifting S-hooks should be considered to avoid the need for extensive bone removal to access buried S-hooks.

50.4.2 Brachial Plexus Injury

Injury to the brachial plexus is unique to VEPTR and rib-based distraction techniques which commonly use the uppermost chest wall for attach-

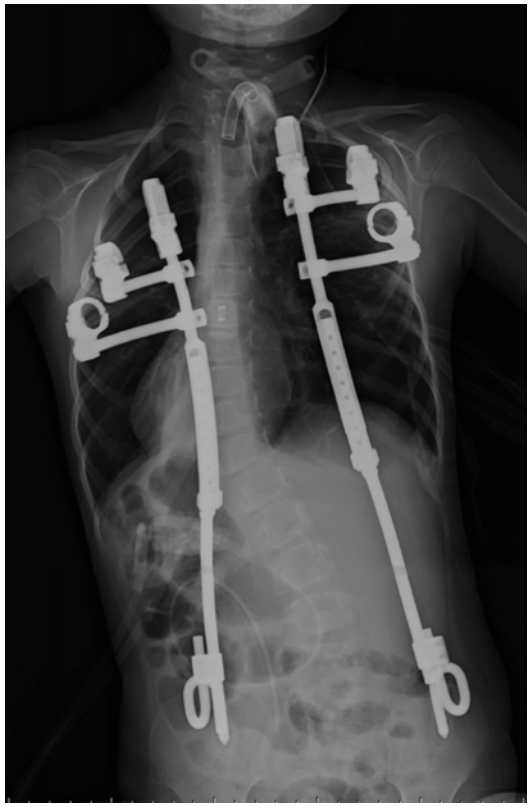

Fig. 50.7 Multiple rib anchor points facilitate load sharing and chest expansion. A 6-year-old with collapsing spine and chest deformity and osteopenia was treated with bilateral rib to pelvis constructs. Multiple rib anchor points were used to distribute force and normalize chest shape

ment points [30, 38, 39]. Concomitant congenital rib and shoulder anomalies may contribute to the occurrence of injury, but two definite etiologies for brachial plexus injury in primary VEPTR or rib-based surgery are recognized. The brachial plexus can be injured directly by an implant placed too cephalad and laterally in the uppermost thorax. Campbell has described the boundaries for safe upper rib cradle placement, suggesting devices should remain medial to the scalene muscles and never cephalad to the second rib [10]. Probably the most common etiology for brachial plexus injury is compression of the plexus between the acutely cephalad-displaced upper chest wall and the clavicle or upper humerus at the time of the initial distraction and expansion procedure. Nassr et al. [38] has validated this explanation experimentally. Brachial plexus palsy may be delayed, as the compression gradually takes effect and postoperative swelling occurs. Awareness of the possibility of brachial plexus palsy and attention to upper extremity motor and sensory monitoring intraoperatively can provide early warning [39]. If extensive displacement of the thorax is planned or the chest wall soft tissue covering is stiff, preliminary clavicular osteotomy, as done for correction of Sprengel's deformity, and preliminary implantation of a tissue expander may help avoid brachial plexus compression. VEPTR when used as a pure spine distraction device, such as in the minimally incisional technique described by Smith [34, 35], is not associated with brachial plexus injury, as there is much less acute chest expansion.

50.4.3 Chest Wall Scarring and Heterotopic Ossification

Rib-based attachments such as VEPTR or chronic contact between the chest wall and either rib-based or spine-based rods can produce local chest wall scarring and fusions between otherwise normal ribs. This phenomenon is readily seen clinically at the time of revision or on CT and has been documented [40, 41]. The clinical significance of chest wall scarring and fusions between ribs in the EOS patient is not clear. If the preoperative condition included a congenitally stiff, small thorax such as that seen with congenital rib fusions, spondylocostal or spondylothoracic dysplasia, or some myopathies, then the stiffness created by rib-based devices, or spine-based devices contacting the ribs, is likely not significant, as the result of treatment remains the creation of a larger albeit still stiff thorax. However, if the chest wall was mobile preoperatively and there were not extensive congenital rib fusions, then the scarring, rib fusions, and stiffness associated with treatment may be relatively detrimental to thoracic function when compared to techniques that do not directly affect the chest wall. The leather-like scarring on the chest wall beneath spine- or rib-based devices is readily apparent to the surgeon at the time of device

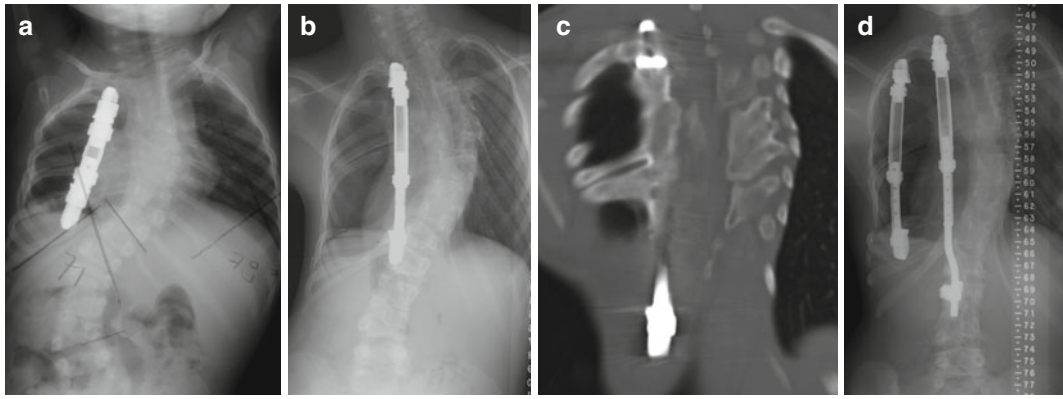

Fig. 50.8 Unintended rib fusion is common in rib-based techniques. Congenital rib fusions and scoliosis were treated with expansion thoracostomy and VEPTR devices at age 13 months (**a**). At age 8 (**b**) device distraction became difficult and CT (**c**) revealed extensive recurrent rib fusions beneath the VEPTR device. Revision with resection of rib fusions and repeat expansion thoracostomy regained control of chest and spine deformity and has continued to withstand repeated lengthening through 4-year (**d**) follow-up. When lengthening of VEPTR or other rib-based devices becomes difficult, CT may reveal underlying rib fusions, which may be amenable to repeat expansion thoracostomy

exchange. Re-fusion of previously separated congenitally fused ribs can occur and may be a cause of deformity progression and inability to continue with lengthening. Repeat osteotomy of rib fusions can lead to improvement in deformity and continued lengthening [41]. Rib re-fusions are typically medial in location or beneath VEPTR devices. Excision of the bony bridge and release of the adjacent chest wall scar usually permits resumption of device lengthening (Fig. 50.8). If VEPTR lengthening becomes increasingly difficult, a search for rib fusion by CT is appropriate.

50.4.4 Scapulothoracic Scarring

Scapulothoracic stiffness, subscapular bursa formation, and spontaneous fusion of the scapula to the VEPTR device and ribs can occur. The location of upper rib-based anchors beneath the scapula stimulates bursa formation and may contribute to shoulder stiffness. If the original procedure included a thoracostomy, the incision of the scapular stabilizing muscles may contribute to scapulothoracic stiffness or dysfunction. Repetitive incisions for lengthening in the area also contribute to scapulothoracic scarring. Attention to surgical technique, early encouragement of range-of-motion exercises, and placement of lengthening incisions away from the scapula may help preserve scapulothoracic function in rib-based devices. If scapulothoracic motion is limited, a CT may reveal bridging bone between scapula and ribs at the location of the subscapular rib-based attachment (Fig. 50.9). At the time of device exchange or as a separate procedure, it is possible to mobilize the scapula from the underlying thorax, freeing scar and adhesions, and excising bony bridges. Postoperative physical therapy is needed to retain mobility.

50.4.5 Wound Integrity and Infection

Wound problems may limit the duration and success of rib-based distraction devices. Integrity of the wound is particularly important for the initial procedure as well as multiple subsequent lengthening procedures. Wound dehiscence or superficial wound infection may lead to deep infection involving the implant, a difficult problem at best with implant removal sometimes needed (Fig. 50.10). Experience with implant infection in VEPTR and rib-based devices is well documented by Campbell, Smith, and others [42, 43].

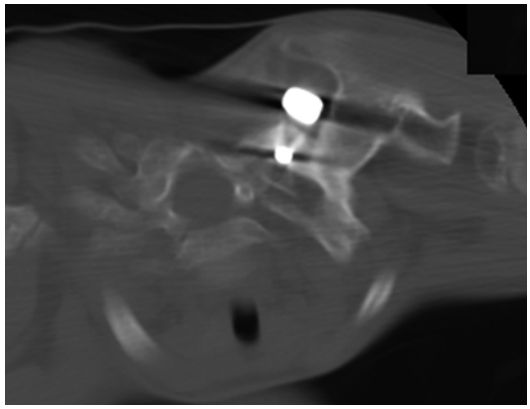

Fig. 50.9 Unintended scapula to thoracic fusion. Scapulothoracic stiffness is common, and occasional fusion between the ribs and scapula in the area of the upper thoracic rib attachment can occur. In this example, expansion thoracostomy and VEPTR insertion were done for severe congenital scoliosis and extensive unilateral rib fusions. Increasing shoulder stiffness was noted at age 6. CT showed bony fusion between the scapula and ribs and upper portion of the device. Revision with resection of the bony bridge and physical therapy improved scapulothoracic motion somewhat

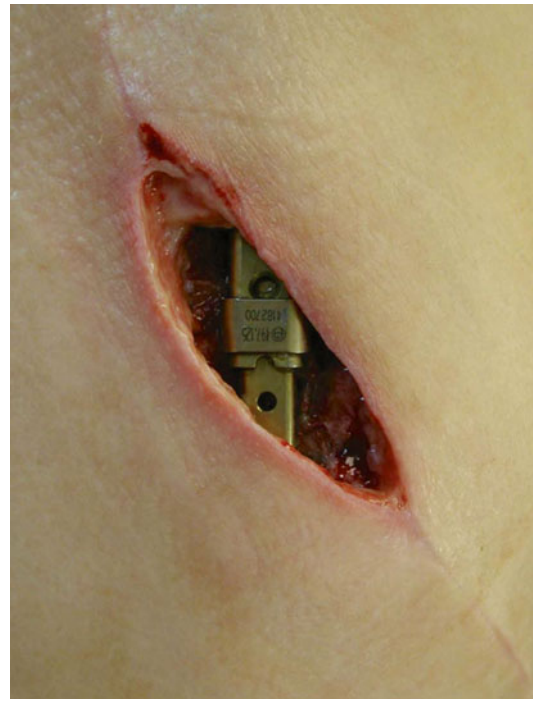

Fig. 50.10 Wound dehiscence. Wound dehiscence occurred in the setting of primary expansion thoracostomy and VEPTR insertion in a 9-year-old. A prior full-thickness wound used for multiple tracheoesophageal fistula surgeries was used for the VEPTR procedure. Dehiscence occurred with minor trauma. Attention to preoperative nutrition, preoperative tissue expansion, and postoperative protective padding of the wound might have made this complication less likely

Preoperative nutrition, soft tissue health, and soft tissue handling are important deterrents to perioperative infection. Predisposing factors to wound problems include poor nutrition, prior infections, or prior incisions in the area such as in myelodysplasia and many neuromuscular deformities, where there may be insensate skin or an uncooperative patient. Nutrition should be assessed and must be optimal preoperatively, even to the point of establishing enteral feeding to encourage weight gain. Preoperative planning of skin flaps and use of tissue expanders can help avoid leaving surgical incisions directly over prominent devices. For expansion thoracotomies, the author prefers to create flaps in which the muscle layer is longer than the overlying skin, making dehiscence less likely (Fig. 50.11). Excessive tension on the wound must be avoided. Prominent devices need to be protected from pressure in the post-op period, and a donut-like padding is incorporated into the post-op dressing.

Each operative device lengthening represents another chance for a wound problem or infection. For lengthening, we try to avoid full-thickness incisions over the device, making separate superficial skin and deep muscular incisions (Fig. 50.12) to access the device so that should there be partial wound dehiscence, the device is less likely to be exposed. We try to handle the soft tissues carefully and avoid excessive trauma, emphasizing restoration of muscle coverage and obliteration of wound dead space with each closure.

50.4.6 Proximal Junctional Kyphosis and Loss of Normal Sagittal Contour

Spine- and rib-based distraction treatments for EOS both rely on repetitive distraction force to maintain alignment between cephalad and caudal

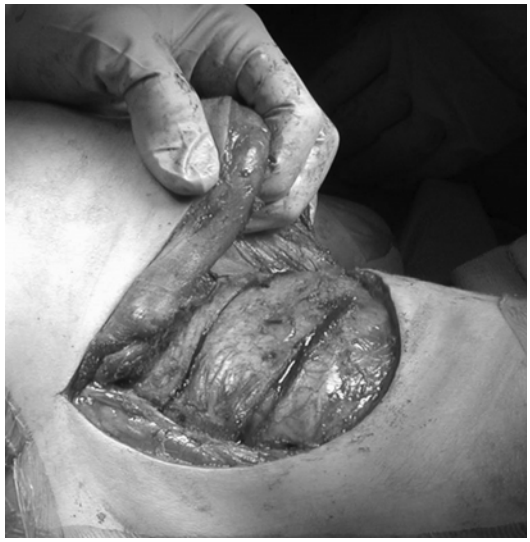

Fig. 50.11 A longer distal muscle than skin flap is used when primary expansion thoracostomy is done. This enables closure under less tension after primary thoracic expansion and lessens the chance of dehiscence by staggering wound layers

anchors. Sagittal alignment is commonly disturbed. Distraction across the normally lordotic lumbar spine inevitably diminishes lordosis, particularly if the caudal end of the distraction is the pelvis. Pelvic S-hooks, because of their more dorsal position, may adversely affect lordosis more than iliac screws. Contouring of the lumbar portion of the rods may mitigate this anti-lordosing effect somewhat, but typically lumbar lordosis is progressively lost with continued distraction treatment. Normal thoracic kyphosis is fostered by the curved contour of the VEPTR device, but with continued elongation of the device, the arc of the device increases, sometimes beyond the desired thoracic kyphosis. Revising with shortening the expandable portion of the device or using a device with a longer radius of curvature mitigates this problem. Proximal junctional kyphosis including the upper anchor points remains problematic for rib- and spine-based devices.

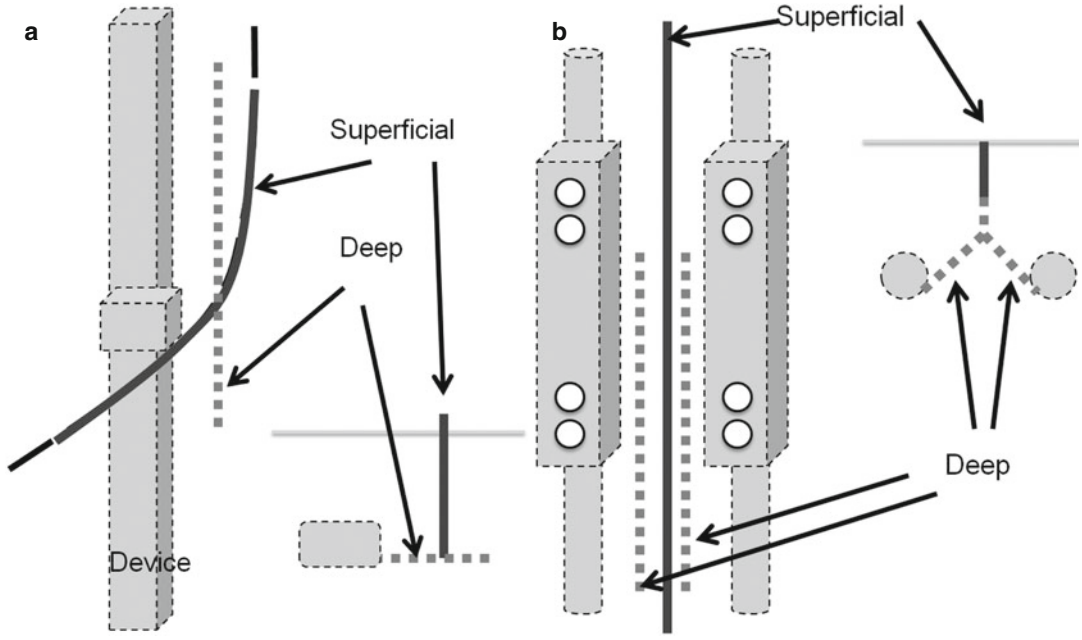

Fig. 50.12 Separate superficial and deep incisions are used wherever possible for device lengthening. Repetitive lengthening procedures create local scar and provide recurring opportunities for wound problems including infection or dehiscence. Careful treatment of soft tissues may make these complications less likely. One approach to minimize wound problems with lengthenings is to use separate superficial skin and deep wound incisions (for VEPTR) (**a**) or for GR (**b**), so that if either layer is compromised, the device is still covered by one of the layers

VEPTR devices can correct some mild upper thoracic kyphosis by cantilever correction at the time of insertion, but they are unable to extend above the second rib as a point of purchase and are less effective at controlling severe upper thoracic kyphosis than a spine-based construct. Although the upper ribs may be kept from deforming further into kyphosis by the VEPTR, the spine may independently deform and continue to collapse into an unacceptable progressive upper thoracic kyphosis, particularly in the setting of poor or absent upper thoracic or cervical musculature or neuromuscular deformity. In the author's opinion, severe upper thoracic kyphosis cannot be treated successfully by rib-based devices and is better managed by spine-based anchors which can continue up into the cervical spine for better purchase past the severe kyphosis.

50.5 Discussion

50.5.1 Dual Versus Single Growing Rods

Several studies have compared single and dual traditional growing rods. In the study by Bess et al. [14], patients with a single growing rod were 1.2 times more likely to experience complications compared to those with dual rods. Dual growing rods also reduced implant-related complications and unplanned surgeries. Mechanical stress is reduced when two rods are used compared to a single rod. This is especially important in growth-friendly techniques, because the construct incurs continued loading and micro-motion that makes the implants susceptible to fatigue and mechanical failure. Dual rods likely dissipate the amount of stress seen by the construct compared to single rods. Additionally, if one rod fails, the intact rod is likely to maintain stability and delay construct revision until the next subsequent planned lengthening procedure. Unplanned surgery is reduced the greatest among patients with submuscular dual growing rods and increased the greatest among patients with subcutaneous single rod. Notching the rods with set screws or benders may create a stress riser and promote fatigue and failure.

50.5.2 Subcutaneous Versus Submuscular Placement

The rationale behind subcutaneous rather than submuscular (subfascial) placement of growing rods was to reduce the risk for unwanted spine fusion, by minimizing subperiosteal exposure of the spine, thereby reducing the incidence of spontaneous fusion. However, Bess et al. [14] reported more total complications and more wound complications following subcutaneous growing rod placement compared to submuscular placement. Patients with subcutaneous rods were 1.8 times more likely to experience complications compared to those with submuscular rod placement.

Submuscular placement of the growing rods reduced wound complications because the implants had superior soft tissue coverage compared to subcutaneous growing rods. The mechanical benefits provided by a dual rod construct were negated by soft tissue problems when they were placed subcutaneously. Patients who had subcutaneous dual rods demonstrated more wound complications, more prominent implants, and more implant-related unplanned procedures compared to submuscular dual growing rods and demonstrated the greatest wound complication than all other treatment groups (11 of 37 patients or 31 %). Unplanned surgeries were reduced the greatest among patients with submuscular dual growing rods (planned to unplanned surgery ratio = 20:1). It appears that children treated with submuscular dual rods benefited from stable constructs that had adequate soft tissue coverage. Conversely, patients treated with single subcutaneous growing rods demonstrated the worst planned to unplanned surgery ratio (7.4:1). These patients were at greatest risk for a complication, either due to tenuous construct stability or poor soft tissue coverage.

In an attempt to reduce the number of surgical procedures during the treatment period and therefore reduce the complications, in posterior growth-friendly distraction-based techniques, the MCGR technology for external, remotely controlled growing rods has been recently cleared by FDA and has shown that the number

of surgeries is significantly reduced [44–46]. Growth-guided treatment modalities and the remote lengthening techniques theoretically allow for even less invasive treatment than was available using traditional growing rod procedure. These techniques are promising; however, there are no long-term outcomes available on their use.

50.5.3 Spine-Based Versus VEPTR (Rib-Based) Distraction Technique

How should potential complications influence the choice between spine-based (GR) and rib-based treatments? Each technique has its "ideal indications," and decision-making for GR and VEPTR is intertwined as the indications for each overlap in some areas (Table 50.1). Perhaps the best indication for GR is a progressive deformity in a normally segmented spine such as infantile idiopathic scoliosis not controlled by bracing or casting. In this example, bone quality and soft tissue coverage should be optimal for GR and chest deformity will likely improve with control of the spinal deformity. By contrast, the "ideal" indication for VEPTR (and expansion thoracostomy) is thoracogenic scoliosis or multiple fused ribs in association with congenital scoliosis. VEPTR may have an advantaged when intraspinal anatomy is unusual such as skeletal dysplasia or congenital scoliosis [47] and makes spine-based anchors difficult or where there is poor soft tissue coverage over the spine such as spina bifida [48]. VEPTR also may make final spinal fusion easier by not creating scar over the spine. However, rib-based devices contacting the chest wall result in chest wall scarring and stiffness, certainly a major drawback if the original deformity had a mobile, unscarred chest wall.

50.6 Conclusions

The goals of EOS treatment can be broadly stated and will help the surgeon in decision-making. EOS treatment should seek to achieve maximum spine length, maximum pulmonary function, and as much residual spine motion as feasible, yet minimize operations, hospitalizations, overall complications, and family burden and cost by the end of the growth.

Table 50.1 GR and VEPTR for EOS compared: indications, treatment, and complications

	Growing rods	VEPTR
Optimum indication	Normally segmented spine (idiopathic, syndromic, neuromuscular) Flexible thoracic deformity	Thoracogenic scoliosis or fused ribs Chest wall constriction is primary deformity Volume depletion diagnoses (Jeune, Jarcho-Levin, etc.) Myelodysplasia
Relative contraindication?	Chest wall deformity is primary	Inadequate soft tissue coverage
Multiple operations needed?	Yes	Yes
Upper thoracic kyphosis?	Possible control	Poor control (author opinion)
Spine growth?	+++	+++
Chest deformity correction?	When flexible	Direct, invasive
Ease of final fusion	Difficult, spine scarred	Easier? Spine less scarred
Final fusion needed?	Variable, depends on residual deformity and spontaneous fusion at maturity	Variable, depends on residual deformity and spontaneous fusion at maturity
Most common mechanical problem:	Rods break	Rib attachments drift
Most common severe complication	Spontaneous posterior spine fusion	Chest wall stiffness

50.6.1 Balancing Risk and Benefit of Growth-Sparing Treatment

Complications are a prominent feature of the treatment of EOS by growth-friendly techniques. The surgeon and family may be faced with a difficult choice between growth-friendly surgical intervention and continued nonoperative management of the deformity. In each case a definitive fusion will likely be needed near maturity. A discussion of the options must include a discussion of complications as they relate to GR or VEPTR. Families need to understand that there will be unexpected events such as rod breakage or loss of anchor points or need for revision or exchange and that complications may cause the premature cessation of treatment of EOS. Treating surgeons must find a balance for the number of procedures during the treatment period. On one hand one must perform lengthenings in a timely basis to allow spinal growth and thoracic development; on the other, an excessive number of lengthening procedures may lead to an increased number of complications. This is especially applicable as growth velocity decreases and duration between lengthening can be extended. In a report by Sankar et al. [49], length achieved progressively diminished with subsequent lengthenings. Previous recommendations by the authors have advocated lengthening the growing rod construct every 6 months to facilitate growth and prevent spontaneous spinal fusion; however, future research is needed to determine the optimal interval for individual construct lengthening. The new MCGR technology will allow more frequent lengthenings without the need for surgery and may prove to be beneficial in reducing complications.

50.6.2 When to Start?

Longer experience with growth-friendly treatment of EOS suggests that many patients will suffer treatment-halting complications. Infection, adverse patient reaction, extensive rib re-fusion in VEPTR, or spontaneous posterior fusion in GR patients may force lengthening to stop well before the planned final fusion near maturity. Campbell has shown greater spine growth and pulmonary function in patients whose treatment began earlier [28], yet Sankar's report [49] from the GSSG database for TGR suggests less and less length is achieved with subsequent lengthenings. Spontaneous posterior fusions beneath growing rods or VEPTR devices are presumed more common when procedures start earlier in life. The surgeon is thus faced with a dilemma: early operation may yield the best chance for lung growth and curve control, but also more operations, greater risk of complications, and a risk that if lengthening is halted because of infection or spontaneous fusion, cessation of growth-sparing treatment will occur at an early age, while large amounts of growth still remain. Although often there may be no choice as to when to intervene, if a deformity is worsening slowly, it may be preferable to wait before starting the cycle of initial growth-friendly surgery followed by repeated lengthening. Early results with MCGR technology have shown that the more frequent lengthenings (1 week to 2 months) may be associated with less implant-related complications but more PJK and failure to distract. Long-term studies needed to determine when is the right age to start MCGR treatment for different group of patients. Two factors other than curve magnitude may help in decision-making: thoracic kyphosis and chest wall deformity. Because growth-friendly treatments are problematic for the treatment of kyphosis, particularly upper thoracic kyphosis, treatment should not be delayed if kyphosis is worsening or severe. Because no growth-friendly treatment is entirely successful in reversing chest wall deformity, the severity and evolution of chest wall deformity should be considered in the decision as to when to initiate treatment. Treatment should start before the chest wall deformity becomes so severe that reasonable thoracic shape and function can no longer be anticipated at the end of treatment.

50.6.3 Minimizing Complications

In this chapter we have tried to set forth the common complications associated with the management of EOS by growth-friendly procedures.

Although not always achievable, complications should be minimized by selecting the optimal surgical technique for the individual disease and deformity. Preoperative patient nutrition is critical for soft tissue health. Careful soft tissue handling techniques minimize complications in both VEPTR and GR surgery. Submuscular/subfascial rod placement, careful creation of anchor bases, and stable foundations will minimize complications related to the implant. In all instances, doing the initial surgery correctly the first time is advantageous, as revisions are less successful. Multiple lengthenings will be needed, and lengthening operations should be done with the same care and respect for tissues as the initial procedure. One needs to remember that even if repeated lengthening can be avoided by using remote control technology, complications related to EOS will remain a challenge for the surgeons caring for these complex patients.

Early recognition of complications, particularly infection, may mitigate their effect. Experience with both GR and VEPTR suggests that early aggressive treatment of implant-related infection can often allow retention of the implants and subsequent continued lengthening [19].

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Part VII

Patient Care and Outcomes

Anesthesia and Postoperative Management of Spinal Deformity Surgery in Growing Children

51

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Key Points

- Because scoliosis surgery is multidisciplinary, several teams must work together to assure the best outcome.
- Early-onset scoliosis may be associated with progressive restrictive lung disease, which increases the risks of pulmonary complications following surgical correction.
- The increased prevalence of malnutrition in patients with neuromuscular scoliosis is a significant concern and one that needs to be evaluated preoperatively.
- Mean blood loss per vertebral level correlates with the number of vertebral levels fused and has been reported to be as high as 500 mL.
- In view of the ischemic nature of spinal cord injury, it is now suggested that mean arterial pressure (MAP) should be maintained in the low normal range.

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51.1 Introduction

The number of pediatric scoliosis surgeries is increasing each year due to recent advances in spinal instrumentation, surgical techniques, and improved perioperative monitoring. A clear understanding of the disease processes with their associated changes in cardiovascular and respiratory function, the preservation of spinal cord blood flow, and techniques in monitoring spinal cord integrity are essential for a good outcome.

Anesthesia during correction of scoliosis in children must address surgical requirements for positioning and monitoring in addition to taking into consideration the associated comorbidities, age-related pathophysiology, and the potential for blood loss and vascular injury of the spinal cord. Children undergoing correction of spinal deformities present a significant challenge to the pediatric anesthesiologist due to not only the wide spectrum of underlying pathology but also the variable range of age and size.

Awareness of the risk of spinal cord injury (SCI) that will affect the function is critical. Expertise in the management of the patients in various positions, prevention of hypothermia secondary to exposure of a large surgical field for a prolong period of time, and severe hemorrhage, which can sometimes exceed the patient's total blood volume, are required. In the case of correction of spinal deformities, all of those situations may converge, demanding attentive intraoperative monitoring, particularly of spinal function, and an anesthesia plan tailored to maintain appropriate spinal cord perfusion, minimize blood loss, and allow for early awakening and extubation.

This chapter reviews spinal blood flow and autoregulation; preoperative assessment; anesthesia care, including one-lung ventilation and blood preservation techniques; and postoperative management.

51.2 Spinal Cord Blood Flow and Regulation

The spinal cord vascular anatomy comprises separate anterior and posterior circulations that arise from the vertebral arteries and are supplemented by intercostals and lumbar vessels from the descending aorta. A single anterior spinal artery supplies the ventral two-thirds of the spinal cord, which includes the corticospinal tracts and motor neurons. Paired posterior spinal arteries form a plexus-like arrangement on the surface of the cord and supply the dorsal one-third of spinal cord parenchyma, which transmits proprioception and light touch. There is essentially no collateral flow between the anterior and posterior circulations [1].

The anterior spinal artery, which supplies motor neurons and tracts, is of uneven caliber and is not functionally continuous. The blood flow to the anterior spinal cord is supplemented by collateral flow through radicular arteries arising from the aorta. Only 6–8 of the 62 radicular vessels that present during development persist into adult life. The large distance between these radicular arteries leaves watershed areas at the upper thoracic and lumbar levels, making the spinal cord particularly vulnerable to ischemia. The great radicular artery of Adamkiewicz arises from the aorta between T8 and L3 nerve roots and supplements the blood flow to the anterior portion of the distal thoracic spinal cord and lumbar enlargement [2]. It provides up to 50 % of the entire spinal cord blood flow and may be injured during aortic or spinal surgery or after spinal trauma [3].

The venous outflow of the spinal cord is divided into two systems, called the vertebral and venous plexuses. These internal and external plexuses communicate with each other and with the segmental systemic veins.

Animal data suggest that the spinal cord blood flow is controlled by the same factors and the same general physiological principles as the cerebral blood flow (CBF) [4]. Spinal cord blood flow is lower than CBF, because absolute spinal cord metabolism is lower than that of the brain. Blood flow to the spinal gray matter is about half that to the cerebral cortex, and the flow to the white matter is about one-third of that to spinal gray matter.

Spinal cord perfusion pressure (SCPP) is equal to MAP minus extrinsic pressure on the spinal cord. Pressures exerted by local extrinsic mechanical compression, such as tumor, hematoma,

spinal venous congestion, and increased intraspinal fluid pressure, can be important determinants of the SCPP. Spinal blood flow is maintained constant by vasodilatation or vasoconstriction of the cord's vasculature to accommodate for changes in MAP.

Limits of spinal cord blood flow autoregulation are 45–180 mmHg. Conditions that affect this autoregulatory mechanism include severe hypoxia, hypercapnia, and trauma [4, 5]. The spinal cord vasculature and cerebral vasculature react to changes in oxygen and carbon dioxide concentrations in a similar fashion.

51.3 Anesthesia Management

51.3.1 Preoperative Evaluation

Patients presenting for correction of spinal deformities present as two distinct groups: those with adolescent idiopathic scoliosis, who are usually well with good cardiorespiratory reserve, and those with secondary scoliosis, who may have very limited reserve and for whom the risks of surgery and anesthesia are greater. The preoperative anesthesia begins with a detailed assessment to identify patients at risk with the aim of decreasing morbidity and mortality related to surgical correction of deformities. The initial step is a thorough history and physical examination. Because of the known association of spinal column deformity with potentially serious alterations of the cardiovascular and respiratory systems, the preoperative anesthesia evaluation is directed primarily to these systems in order to exclude any preoperative factor that could make the surgical intervention too dangerous unless corrected (e.g., cardiac failure, severe pulmonary hypertension, clotting abnormalities, respiratory insufficiency, etc.). When such factors have been ruled out, the preoperative evaluation progresses to detect other factors that, although perhaps not life threatening, could cause significant morbidity (e.g., malnutrition, muscle weakness, abnormal cough reflex, chronic aspiration secondary to gastroesophageal reflux). The outcome of these patients depends, to a significant degree, on their physiological reserves to endure the high demands that spinal surgery entails. During their preoperative evaluation, it will be important to identify any physiological abnormality and assess the degree of severity. Patients must be in optimal medical condition before planned surgery if uncomplicated intraoperative and postoperative courses are to be realized.

Because scoliosis surgery, especially early-onset scoliosis, is multidisciplinary, several teams must work together to assure the best outcome. Patients with comorbidities will require an evaluation by pulmonary, cardiology, pediatric, and anesthesia services. Neurology should be involved in patients with significant underlying neurological conditions, and gastroenterology needs to be involved for patients with poor nutritional status or suspicion of a previously undiagnosed gastrointestinal disorder. In the physical examination during the preoperative evaluation of the patient with scoliosis, special attention needs to be given to the airway anatomy, which can be severely deformed by abnormalities of the thoracic and cervical spine and may require special airway instrumentation devices like video laryngoscopes and/or fiberoptic assistance at the time of surgery.

Most cases of idiopathic scoliosis occur in otherwise healthy adolescent females, with only mild abnormalities that do not affect their intraoperative management. Preoperative assessment in these patients includes hemoglobin and coagulation function testing and a type and crossmatch. Autologous blood or directed donor blood donation is highly recommended. Preoperative pulmonary function tests (PFTs) are only indicated for curves 60–80° or greater, if there is a history of reactive airway disease, or if a combined anterior and posterior spinal fusion is being performed.

51.3.2 Cardiopulmonary Involvement

Scoliosis may be associated with progressive restrictive lung disease, which increases the risks of pulmonary complications following surgical correction. Patients with preexisting respiratory disease have an increased risk of developing postoperative complications [6, 7]. The spinal

deformity significantly affects the respiratory mechanics, gas exchange, pulmonary vasculature, and chemical regulation of ventilation. As the curvature worsens, the severity of pulmonary and cardiovascular involvement increases, often resulting in respiratory failure, pulmonary hypertension, and cor pulmonale. Several factors are thought to play a role in the changes of lung volumes and decrease respiratory compliance. These include the abnormal development of the thoracic cage with a direct effect on the elastic properties of the respiratory system. In addition, the deformity will also affect the development of inspiratory and expiratory muscle forces [8, 9]. These changes occur earlier and are more pronounced in patients with congenital scoliosis than in those with adolescent scoliosis, which make age of onset a significant factor for treatment, because curves that demonstrate a major thoracic deformity before a child is 5 years of age are more likely to be associated with significant cardiopulmonary morbidity in addition to other growth abnormalities. Long-term cardiopulmonary complications of untreated scoliosis can include hypoxemia, hypercapnia, cor pulmonale, and pulmonary hypertension.

The goal of surgical treatment of early-onset scoliosis (EOS) is to stop the progression of the curve while allowing maximum growth of the spine, lungs, and thoracic cage, preventing the chronic hypoxemia and pulmonary hypertension that presents as the curve is left untreated. The literature supports surgical treatment of spinal deformities in pediatric patients in order to prevent cardiopulmonary morbidity; surgical treatment of infantile idiopathic scoliosis is recommended for progressive curves of 45° or greater in an immature child, indicating the current trend toward less tolerance of curve progression prior to operative intervention.

The effect of surgery on lung function depends on the surgical approach and type of surgery. Thoracic surgery decreases lung volumes, expiratory flow rates, and oxygenation after surgery as a result of the surgery itself, anesthesia, pain, and immobilization [10]. In adult studies, thoracic and upper abdominal surgery may reduce lung volumes and flow rates to 50–75 % from their preoperative baseline values.

Because of developmental immaturity of the respiratory system and possibly decreased ability to cooperate with respiratory care, the decrements in children may be even worse [11]. This can lead to serious pulmonary complications, including pneumonia, respiratory failure requiring prolonged mechanical ventilation, bronchospasm, atelectasis, and exacerbation of the underlying chronic disease. Pulmonary function decreases immediately following scoliosis surgery, and the extent of this decrease has been documented in children. Yuan et al. [12] evaluated the immediate change in PFTs values in 24 children following scoliosis surgery and showed that PFT values fell by 60 % after surgery. The PFT nadir was at 3 days. PFT values remained significantly decreased at 1 week, with values half of preoperative baseline. Most PFT values were near the preoperative baseline by 1–2 months postoperatively. No statistical significance was found between the degrees of decline in PFT with etiology of scoliosis. Based on their findings, the authors concluded that patients are still at risk for postoperative complications as long as 1 week after surgery. Neuromuscular scoliosis (NS) includes a wide variety of disorders, including muscular dystrophies, spinal muscular atrophies, and cerebral palsy. When taken as a group, patients with NS have increased postoperative complications [13, 14]. Although the role of preoperative PFT in patients with idiopathic scoliosis is controversial, PFT should be routinely performed in NS patients to assess the need for prolonged postoperative mechanical ventilation, as accurately as possible. In a study evaluating risk factors for prolonged ventilatory support in 125 patients with idiopathic and nonidiopathic scoliosis on whom PFTs were obtained in the preoperative visit, patients with neuromuscular disease were more likely to require prolonged mechanical ventilation than other diagnosis groups ($P < 0.0001$). After removal of the neuromuscular patients, no significant difference was observed in the risk of prolonged ventilation among other diagnosis groups ($P = 0.631$). The combination of age >13 years, neuromuscular disease, and FEV1 <40 % almost undoubtedly

predicted the need for prolonged mechanical ventilation in this study [15].

In addition to the restrictive lung deficit caused by the spinal deformity, children with neuromuscular disease have impaired pulmonary function from progressive muscle weakness and recurrent chest infections, as a consequence of poor cough and impaired airway protective reflexes. Of these conditions, Duchenne muscular dystrophy (DMD) is the most common disorder, with an incidence of 1 in 3300 male births. It is inherited as an X-linked disorder, which presents as weakness during the fourth to eighth years of life. The genetic defect results in a deficiency of the protein dystrophin in the skeletal, cardiac, and smooth muscle. As these patients enter their second decade of life, the myocardium is progressively affected; patients develop dystrophy cardiomyopathy and may present with arrhythmia, ventricular dilatation, and heart failure. Preoperative assessment in patients with muscular dystrophies should routinely include a 12-lead electrocardiogram and an echocardiogram.

The increased prevalence of malnutrition in patients with NS is a significant concern and one that needs to be evaluated preoperatively. Systemic implications of a suboptimal nutritional status include poor wound healing and immunological depression; their presence makes these patients highly susceptible to postoperative complications [16].

Poor intake of vitamin K will cause depletion of vitamin K-dependent coagulation factors, resulting in preoperative coagulation dysfunction. Preoperative screening of coagulation function is required to detect any abnormality that may require optimization previous to surgery.

51.3.3 Conduct of Anesthesia

It is important during the planning of the anesthesia technique to take into consideration all the potential problems associated with spinal reconstruction. Aside from consideration of the cardiorespiratory risk, other important challenges during the administration of anesthesia for this operation in children and adolescents include management of a patient in the prone or lateral positions, prevention of hypothermia secondary to a long procedure with an extensive exposed area, potential for severe blood loss, the provision of one-lung ventilation when required for surgical access, and the preservation of adequate spinal cord perfusion.

Premedication should be guided by the patient's physical examination status. Midazolam administered orally or intravascularly in patients with intravenous access provides adequate anxiolysis, facilitating the transition to the operating room. For patients with abnormal airway anatomy on whom fiberoptic-assisted endotracheal intubation is planned, an anticholinergic agent such as glycopyrrolate will dry secretions and facilitate visualization. Following premedication, patients are transported to the operating room, where placement of ASA standard monitors is followed by induction of general anesthesia. The choice of induction of general anesthesia will be guided by the age of the patient, clinical status, and patient's preference. Inhalation induction with sevoflurane and combined nitrous oxide/oxygen mixture is the preferred method at our institution. In children and adolescents with intravenous access and appropriate cardiac reserve, propofol is the agent of choice; in those without optimal cardiac function, ketamine is a good option. After completion of induction and establishment of vascular access, muscle relaxation is normally achieved with a nondepolarizing muscle-blocking agent (NMBA). A single dose of the NMBA, administered during the induction of general anesthesia, provides optimal conditions for endotracheal intubation and will allow return of neuromuscular function to obtain baseline motor evoked potentials (MEP) before surgery begins. Monitoring muscle relaxation with a nerve stimulator is the standard of care in any patient in whom muscle relaxation is used as part of the anesthesia technique; in patients with muscular disorders, the duration of action of these agents may be prolonged. Succinylcholine is contraindicated in Duchenne muscular dystrophy, along with other dystrophic myopathies, because of increased risk of malignant hyperthermia; in patients with neuropathies, the potential

for rhabdomyolysis, hyperkalemia, and cardiac arrest also precludes its use.

Immediately after the endotracheal tube is secured, additional venous access needs to be obtained in all children. Because of the potential of severe hemorrhage during spinal reconstruction, a minimum of two large peripheral intravenous lines will provide for adequate and prompt intravascular volume resuscitation in case of severe bleeding and will also allow for the infusion of medications. Patients with poor intravenous access and those with significant comorbidities that could prolong hospital stay require central venous cannulation for appropriate intravascular access.

Monitoring during spinal surgery should routinely include ECG, pulse oximetry, capnography and anesthetic agent monitoring, temperature (core and peripheral), neuromuscular block, continuous neuromonitoring by somatosensory and motor evoked potential, and blood pressure. Owing to the probability that hemodynamic instability could occur during the course of the surgery, intravascular monitoring of blood pressure is required. Hemodynamic instability could be caused by either a reduction in preload, an increase in afterload, impaired contractility, or any combination of the above. The prone position alone can reduce the cardiac output from decreased venous return and increased intrathoracic pressure [17]. Major blood loss is a significant factor for reduction in preload and, in patients with impaired contractility, may not be well tolerated; inotropic support may be required, particularly in boys with DMD, to maintain appropriate hemodynamic parameters. Invasive intravascular monitoring is routinely obtained in most cases of pediatric spinal surgery by placement of an arterial cannula allowing for continuous monitoring of blood pressure and frequent evaluations of acid–base status, blood gases, hemoglobin, hematocrit, and coagulation function. Monitoring for air embolism is also recommended in patients undergoing scoliosis repair. Measurement of unconsciousness by bispectral index (BIS™) allows titration of medication for the maintenance of anesthesia. A BIS value of <60 reflects depression of the brain function adequate to ensure unconsciousness during surgery [18, 19]. Titration of anesthesia agents using the BIS monitor to maintain an appropriate level of unconsciousness results in decreased drug utilization and more rapid recovery [20, 21]. In this regard, BIS monitoring serves as a useful intraoperative monitor for guiding drug administration during correction of spinal scoliosis.

When appropriate vascular access and monitoring have been established, and before positioning the patient on the operating table, children with idiopathic scoliosis are turned in a lateral position for an intrathecal administration of 5–10 μg/kg preservative-free morphine via a 24-gauge pencil point spinal needle. The addition of intrathecal morphine decreases MAP, blood loss, and anesthetic agent requirements, without affecting somatosensory and motor evoked potentials; in addition, it provides for up to 18 h of postoperative analgesia in children undergoing posterior spinal fusion [22, 23]. The final position of the patient on the operating table depends on the specific type of surgery but is generally prone or lateral. In the prone position, the patient in placed face down with supports placed beneath the upper chest, shoulders, and iliac crest, allowing freedom of abdominal movement to facilitate ventilation by preventing restriction of diaphragmatic movement.

The surgeon and the anesthesiologist must coordinate patient position, whether this involves a specific positioning frame (e.g., Relton–Hall or chest rolls) or type of headrest. Placement of the patient and appropriate padding of pressure points should be performed by the surgeon with the assistance of both nursing and anesthesia staff (Fig. 51.1).

The patient's arms should be placed in a well-padded position of no more than 90° of combined abduction and forward flexion, and care should be taken to avoid pressure in the axilla. The elbows should remain free of compression, with particular attention paid to the ulnar nerve. In the female patients, the breasts should be moved toward the midline, and generous padding should be placed over the anterior–superior iliac spine in all patients to decrease the risk of injury to the lateral femoral cutaneous nerve. The knees

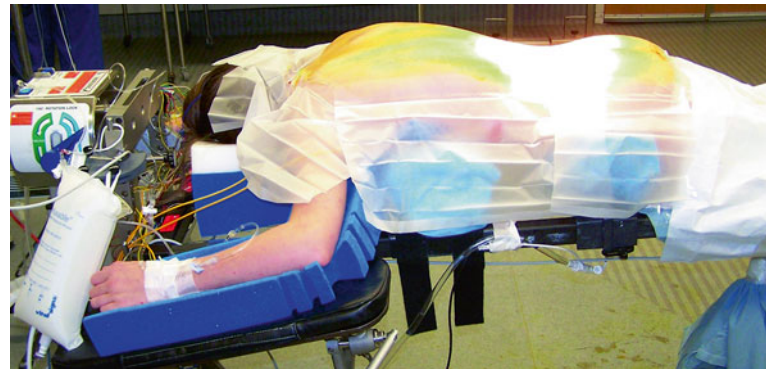

Fig. 51.1 Prone position with a Jackson table. The arms are extended less than 90° whenever possible. Pressure points are padded, and the chest and pelvis are supported to preserve pulmonary compliance and minimize intra-abdominal pressure

should be flexed and the feet be supported, but the toes should be allowed to hang freely.

The most important factor that influences the choice of anesthesia agents is the use of evoked potentials to assess spinal cord integrity. Only small concentrations of inhalation agents are used. Maintenance of general anesthesia usually consists of continuous infusions of propofol and remifentanil. In patients with intrathecal morphine, the addition of remifentanil is not necessary.

51.3.4 One-Lung Ventilation (OLV)

Pediatric scoliosis surgery may require single-lung ventilation for surgical access. Current methods of lung isolation are inadequate for some or all of these children. Spinal access in pediatric scoliosis correction surgery may require lung collapse for several hours and is traditionally achieved in larger children with a double lumen tube (DLT) or with specially designed selective endobronchial blockers that are placed with the assistance of a fiberoptic bronchoscope like the Univent endotracheal tube (Fuji Systems, Tokyo, Japan) and the Arndt endobronchial blocker (Cook Critical Care, Birmingham, IN). Other alternatives to providing bronchial blockade, such as a Fogarty embolectomy catheter or main bronchus intubation with a conventional endotracheal tube, are limited by nonspecific design [24]. They can result in inadequate isolation that requires direct lung compression, which is potentially traumatic for lung tissues. Although DLT is the standard technique for lung isolation in thoracic surgery, its use in scoliosis patients is limited for several reasons. The smallest size available is 26 Fr, which prevents its use in patients <8–10 years of age and in those who are difficult to intubate. In patients with abnormal airway anatomy, placement of a DLT may not be possible and may be contraindicated due to the potential traumatic injury to the airway.

For younger patients and those in whom the DLT or the Univent endotracheal tube is not indicated, the Arndt endobronchial blocker (Cook Critical Care, Birmingham, IN) is an alternative for providing lung isolation. There are three commercially available sizes of this device: 5, 7, and 9 Fr. Its application and successful use in small patients undergoing scoliosis surgery has been reported [25].

In small children, use of the smallest blocker is limited by its external diameter and can be placed only via an endotracheal tube with an internal diameter of 4.5 mm or larger, requiring a thin pediatric bronchoscope.

Dexterity in fiberoptic bronchoscopy and familiarity with these devices are essential for their successful use. Although single-lung isolation provides the optimal surgical access, it is not without risk of potential serious complications due to migration and tracheal occlusion by endobronchial balloons, resulting in inadequate ventilation. Constant vigilance is required, including uninterrupted auscultation of breath sounds on the nonisolated lung and monitoring of airway resistance, in order to identify this problem promptly and avoid serious complications.

51.3.5 Antibiotic Prophylaxis

The prophylactic administration of antibiotics is indicated during scoliosis surgery in order to decrease the risk of a surgical site infection, which is associated with increased morbidity, prolonged hospital stay, and added health care cost [26]. These infections are difficult to treat and often require multiple surgical debridements, long-term parenteral antibiotics, and hardware removal.

51.3.6 Hypothermia Prevention

Prevention of hypothermia secondary to a long procedure with an extensive exposed area is also very important. The goal is to prevent its vicious circle of coagulopathy and acidosis. There are several other consequences of hypothermia, those include impaired drug metabolism, impaired SSEP and MEP signals, prolonged recovery from anesthesia, cardiac irritability, wound infections, and postoperative shivering.

Routine use of environmental factors by changing the room temperature, forced warm air blankets, fluid warmers, and warmed gases will help maintain the temperature of the patient.

51.4 Neurological Risk

Paraplegia resulting from the operative treatment of scoliosis is the complication most feared by surgeons, anesthesiologists, and patients [27, 28]. Neurological injury is most often due to ischemic injury caused by spinal cord distraction or direct spinal cord compression by a hook or wire. The areas of the cord most vulnerable to ischemic injury are the motor pathways supplied by the anterior spinal artery. Rapid interventions, such as adjustment or removal of the hardware, can reverse neurological deficits and prevent permanent injury. Prevention of spinal cord injury (SCI) begins with maintaining spinal cord perfusion with reasonable MAP and agreed transfusion thresholds [29].

Recognition of the high-risk case is essential. Congenital kyphosis, neurofibromatosis, skeletal dysplasias, and postinfectious scoliosis carry higher neurological risk [30]. Congenital scoliosis also increases risk due to a higher incidence of occult spinal cord anomalies [31, 32]. Neurological deficit prior to the onset of treatment indicates an increased possibility of additional injury [28].

Intraoperative spinal cord monitoring is an integral part of almost all surgeries for scoliosis in pediatric patients. For a more comprehensive understanding, please refer to Chap. 53.

51.4.1 Intraoperative Management of Neurological Insult

If spinal cord injury (SCI) is suspected, immediate confirmation and appropriate action are necessary to reduce the likelihood of permanent damage. In general, the following events should occur in a timely and coordinated fashion:

1. The anesthesiologist should be informed and the patient's blood pressure, hematocrit, and oxygenations should be optimized [33].
2. Wake-up test. The Stagnara wake-up test remains the gold standard to determine the presence or absence of injury to the anterior (motor) portion of the spinal cord. It requires two important elements: an anesthesiologist familiar with the procedure and a patient who can understand and follow directions. If the patient cannot follow directions due to mental retardation, significant preoperative weakness, or profound hearing loss, the surgeon will not be able to evaluate any abnormal result. Because the patient may struggle during this test, there is a risk of self-extubation [34]. Therefore, a gurney should be available in the room to turn the patient quickly into the supine position so that reintubation can be performed without delay.
3. Remove instrumentation if there is no change with previous maneuvers.

In the past, methylprednisolone had been administered for acute SCI; there is insufficient evidence to support the prophylactic administration of methylprednisolone as a standard treatment in acute SCI [35].

51.5 Anesthesia Techniques in Blood Conservation

Perioperative blood loss remains a significant concern for orthopedic surgeons performing spinal fusion and instrumentation. Mean blood loss per vertebral level correlates with the number of vertebral levels fused and has been reported to be as high as 503 mL per segment [36]. Many factors affect blood loss in patients undergoing spinal fusion and instrumentation; the surgical technique employed, duration of surgery, number of vertebrae fused, site of autologous bone graft harvest, MAP, the pressure in the inferior vena cava, and patient position affect the total blood loss. In addition, there may be other factors influencing blood loss during scoliosis surgery that are not affected by current techniques to decrease intraoperative bleeding. Yarom et al. [37] described abnormal platelet in vitro function and ultrastructure in patients with idiopathic scoliosis, and Udén et al. [38] noted both an increased bleeding time and decreased ability of collagen to aggregate platelets in patients with scoliosis when compared with nonscoliotic controls. These factors are exacerbated in scoliotic patients with an underlying neuromuscular disorder. In one study comparing neuromuscular scoliotic patients with idiopathic scoliotic patients, the former were found to have a nearly sevenfold risk of losing over 50 % of their estimated blood volume during scoliosis surgery, after adjusting for age, weight, number of levels fused, and coagulation profile [39]. Mean estimated blood loss associated with surgical procedures for neuromuscular scoliosis has been reported to range from 1000 mL for anterior procedures to 2000–3000 mL for posterior approaches [40]. Disseminated intravascular coagulation has also been described in patients undergoing surgery for scoliosis, suggesting that extensive decortication may stimulate the intrinsic system of the coagulation cascade, promoting the production of kallikrein, bradykinin, and plasmin, thereby increasing fibrinolytic activity, which may ultimately lead to a consumptive coagulopathy and increase perioperative blood loss.

There is considerable evidence that transfusion of allogeneic blood products is associated with serious complications, including transfusion reactions, transmission of infectious diseases, graft-vs. -host disease, acute lung injury, and immunosuppression.

Because major blood loss is to be expected, proper positioning, optimal ventilatory pressures, autologous blood donation, intraoperative hemodilution, the use of a cell saver, induced hypotension, and the use of antifibrinolytic agents should be considered. Transfusion decisions should be based on clinical judgment rather than reliance on a predetermined hemoglobin concentration as a "transfusion trigger."

51.5.1 Positioning and Ventilation

Proper positioning plays an important role in blood conservation in patients in the prone position. Placing the patient with support below the pelvis and shoulder leaves the abdomen free. It has been shown that, by preventing pressure on the abdominal wall, the pressure on the vena cava is minimized, thus reducing blood flow through collateral vertebral venous plexuses, known as Batson's plexus [41]. During mechanical ventilation, airway pressure increases, resulting in an increase in mean intrathoracic pressure. Because venous return to the thorax is dependent on the difference between peripheral venous pressure and intrathoracic pressure, venous return is, consequently, impeded during the inspiratory cycle of mechanical ventilation [42].

There is evidence that elevation in intrathoracic pressures during mechanical ventilation raises the peripheral vascular pressure to adequate level to affect blood loss. Spontaneous ventilation, on the other hand, assists venous return because of reduced mean intrathoracic pressure with inspiration. Therefore, the hemodynamic differences between spontaneous and mechanical ventilation can reduce intraoperative blood loss.

Another aspect of ventilation affecting venous return is expiratory and inspiratory resistance. Maintaining expiratory resistance as low as possible assists venous return by reducing intrathoracic

pressure [43]. Appropriate management of reactive airway disease, appropriate setting of the inspiratory-to-expiratory ratio, allowance of adequate expiration time, and maintenance of unobstructed expiratory flows (e.g., avoidance of kinks or buildup of secretions in the endotracheal tube) may be beneficial in reducing blood loss.

51.5.2 Preoperative Autologous Blood Donation and Acute Normovolemic Hemodilution

Although preoperative donation of autologous blood was first suggested by Fantus in 1937, when he founded the first blood bank in the United States, the technique did not became popular until the 1980s. Advantages of this technique include reduced exposure to allogeneic blood, the availability of blood for patients with rare phenotypes, reduction of blood shortages, avoidance of transfusion-induced immunosuppression, and the availability of blood to some patients who refuse transfusions based on religious beliefs. There are no limitations in regard to a patient's weight or age. Patients who weigh 50 kg or more can donate a standard unit of blood (450 mL), while those who weigh less than 50 kg can donate proportionately smaller volumes. The hematocrit (Hct) should be ≥33 % prior to each donation. Red blood cell production can be augmented by iron supplementation and the administration of erythropoietin. Donations may be made every 3 days, but the usual practice is to donate 1 unit per week. The last unit should be donated at least 5–7 days before surgery to allow plasma proteins to normalize and to restore intravascular volume. Autologous blood donation in pediatric patients undergoing spinal fusion is an efficient blood-saving technique, especially in idiopathic scoliosis. In some centers, almost 80 % of children and adolescents undergoing spinal fusion participate in the autologous blood predonation program; almost 90 % of the participants avoid receiving allogeneic blood. Patients with neurological causes of scoliosis less often participate in the predonation program and usually need transfusion of allogeneic blood.

Acute normovolemic hemodilution (ANH) involves removing and temporarily storing 2–4 units of a patient's blood just before major elective surgery in which major blood loss is anticipated. The blood that has been withdrawn is then reinfused into the patient during or after surgery. Simultaneous infusions of crystalloids (3 mL of crystalloids per 1 mL of blood withdrawn) have been recommended. The rationale for the use of hemodilution is that, if intraoperative blood loss is relatively constant with or without preoperative normovolemic hemodilution, then it is better to lose blood at a lower rather than at a higher level of Hct. This procedure lowers the patient's preoperative Hct to 28 %. If the perioperative Hct level falls to 24 %, the ANH blood units are reinfused in reverse order of their collection (i.e., last unit collected is the first unit transfused). The first unit of blood collected, and therefore the last unit reinfused, has the highest Hct, contains the most platelets, and has the highest concentration of clotting factors [44].

Clinical observations show that ANH reduces allogeneic blood use in 20–90 % of patients with no difference in postoperative outcomes [45, 46]. Furthermore, ANH is substantially more cost-effective than transfusion. ANH has been shown to decrease perioperative transfusion requirements of adolescents undergoing extensive spinal surgery. By allowing patients to arrive at surgery with a higher preoperative hemoglobin and Hct levels and by decreasing the quantity of predonated autologous blood collected and therefore used, the hemodilution method may indirectly decrease the quantity of postoperative autologous transfusion in this population.

51.5.3 Controlled Hypotension

Controlled hypotension involves the use of pharmacological agents to lower the MAP to 50–65 mmHg. This method significantly decreases both intraoperative blood loss and blood requirement. Blood loss during controlled hypotension is at least in part dependent on venous pressure [47, 48].

The potential contribution of venous pressure to blood loss can be further understood from studies using epidural anesthesia. Modig and Karlstrom [47] demonstrated that both intraoperative and postoperative blood loss are significantly lower during epidural anesthesia when compared with general anesthesia in patients undergoing total hip replacement.

In view of the ischemic nature of spinal cord injury, it is now suggested that MAP should be maintained in the low normal range, and hypotension should be quickly corrected if there is a loss of MAP [29]. It has been shown that the spinal cord is more sensitive to distraction and/or compression during controlled hypotension than at normotension as measured by reduction in somatosensory-evoked potentials [49, 50].

Several different agents and methods are used in spinal surgery to provide controlled hypotension, including direct-acting vasolidators (sodium nitroprusside, nitroglycerine), calcium channel blockers, and intrathecal opioids.

Sodium nitroprusside produces a reliable decrease in blood pressure and at least initially increases spinal cord blood flow; however, it may be associated with tachyphylaxis, rebound hypertension, and toxicity. Nitroglycerin, which has been used successfully for controlled hypotension in adults, may be ineffective in children. Nicardipine is the first calcium channel-blocking agent for intravenous administration. It was introduced to prevent and treat spasm of cerebral arteries in patients with subarachnoid hemorrhage, but it has also been employed in adults to control perioperative hypertension. It does have some intrinsic negative chronotropic effects, which may limit the rebound tachycardia. Like other direct-acting vasodilators, nicardipine and other calcium channel antagonists may increase intracranial pressure. Studies comparing SNP with nicardipine have demonstrated several potential advantages of nicardipine, including fewer episodes of excessive hypotension, less rebound tachycardia, less activation of the rennin–angiotensin and sympathetic nervous systems, and, in some studies, decreased blood loss. One disadvantage of nicardipine is that its effect is somewhat prolonged (20–30 min) following discontinuation of the infusion.

51.5.4 Hemostatic Drugs

Desmopressin acetate (DDAVP) is a synthetic analog of vasopressin with decreased vasopressor activity. DDAVP therapy causes a 20-fold increase in plasma levels of factor VIII and stimulates vascular endothelium to release von Willebrand factor (vWF). Factor VIII is a plasma glycoprotein that speeds up activation of factor X by factor IXa in the presence of a phospholipid surface and calcium ions. vWF mediates platelet adherence to vascular subendothelium by functioning as a protein bridge between glycoprotein Ib receptors on platelets and subendothelial vascular basement membrane proteins.

Intravenous DDAVP has been shown to reduce blood loss during scoliosis surgery in some patients. In patients with neuromuscular diseases undergoing spinal fusion, the overall blood loss was reduced in the treatment group when compared with placebo group, but the results were not statistically significant [51].

51.5.5 Antifibrinolytics

Epsilon-aminocaproic acid (EACA) and tranexamic acid (TA) are omega aminocarboxylic acid analogs of lysine. The antifibrinolytic effect of these drugs is due to the formation of a reversible complex with plasminogen, which prevents the fibrinolysis that would normally occur with activation of plasminogen to plasmin. As a result of this inhibition, fibrin is not lysed, which allows for the formation of a more stable clot. EACA is administered at an intravenous loading dose of 100–150 mg/kg, followed by infusion of 10–15 mg/kg/h. Ninety percent is excreted in the urine within 4–6 h of administration. TA is six to ten times more potent than EACA and may be used at lower doses (loading dose of 10 mg/kg followed by an infusion of 1 mg/kg/h). Ninety percent is present in the urine after approximately 24 h. Adverse effects of EACA or TA may be related to the effect on coagulation function and the route of excretion. As these agents are cleared by the kidneys, their administration in the presence of renal or ureteral bleeding is not

recommended because ureteral clot formation and possible obstruction may result. In 2001, Florentino-Pineda et al. [52] administered EACA or placebo (100 mg/kg followed by 10 mg/kg/h) to 28 adolescents undergoing posterior spinal fusion. Patients who received EACA had decreased intraoperative blood loss (988 ± 411 mL vs. 1405 ± 670 mL, $P = 0.024$) and decreased transfusion requirements (1.2 + 1.1 U vs. 2.2 ± 1.3 U, $P = 0.003$). TA has been found to be similarly effective in decreasing blood loss in spinal fusion [53].

51.6 Postoperative Care

Children returning from surgery directly to the intensive care unit have a number of unique concerns. It is incumbent upon the care team to systematically address the needs of the patient and provide support during this critical phase.

51.6.1 Central Nervous System

Scoliotic patients should be monitored closely after surgery; the patient needs to be placed in an intensive care unit or a "step-down" unit for approximately 24 h. Postoperative monitoring should include close assessment of MAP and overall hemodynamic status. Anecdotal reports have hypothesized that the late-onset neurological changes within the immediate postoperative period may be the result of spinal cord ischemia in patients due to relative hypotension [54].

Postoperative analgesia must not overly sedate the patient or in any way mask the timely discovery of delayed or evolving neurological dysfunction. Family presence is often vital to assess children's level of activity and cognition, especially in the preverbal years or in the special-needs population.

Pain Control Pain after spinal surgery usually requires the use of an opioid-based technique. In younger children, this may be by using a morphine infusion or a nurse-controlled analgesia. Children >7 years of age may be able to use patient-controlled analgesia. Infiltration of the wound at the end of surgery with local anesthetic will improve pain relief in the immediate postoperative period. Opioids should be supplemented with acetaminophen of 15 mg/kg/dose given every 4 h around the clock for the first 48 h. Muscle spasms present a unique challenge after posterior spinal fusion. Diazepam (0.05–0.2 mg/kg/dose Q 2–4 h, max dose 0.6 mg/kg within an 8-h period) administered intravenously may prove a useful adjunct in the first 48 h as the musculature of the rib cage and back adjust to the new contour afforded after corrective surgery. Scoliotic surgery pain could also be managed with the use of an epidural infusion of opioids–local anesthetic combination with the catheter inserted by the surgeon at the end of the procedure [32]. Intrathecal morphine administered as part of intraoperative anesthesia management will provide for up to 18 h of postoperative analgesia in children undergoing posterior spinal fusion [22].

51.6.2 Respiratory System

Many of these patients have restricted lung disease secondary to rib cage distortion that has led to decreased maximum voluntary ventilation over time. The goal is to extubate the children in the operating room or at least in the first 24 h. This goal is met with ease if the child had good presurgical respiratory function as demonstrated through pulmonary function testing. Often children are successfully extubated and require only face shield oxygen immediately, with noninvasive positive pressure ventilation reserved for the children who are unable to effectively ventilate despite optimal positioning and low sedation requirement.

51.6.3 Cardiovascular System

Blood pressure lability is frequently seen in the first 24 h after surgery, and thus, arterial line monitoring of blood pressure is recommended. Shock may develop secondary to hypovole-

mia from severe fluid losses during prolonged surgeries. Inotropic support may be required if contractility seems impaired, to ensure adequate perfusion in the patients.

51.6.4 Fluids/Electrolytes

Fluid losses during a prolonged surgery require appropriate volume replacement. It is not unusual for a child to receive up to 100 mL/kg of fluid during surgery. Excellent hand-off communication must occur between the anesthesia staff and intensive care team to ensure adequate understanding of the fluid balance concerns for each patient. Children are often managed with Dextrose 5 % + normal saline and potassium chloride 20 meq/L at maintenance rate.

51.6.5 Gastrointestinal

Enteral nutrition is often not started for the first 48–72 h, but should be engaged as soon as possible. Adequate nutrition promotes healing and will aid in the total recovery of the child. While under NPO, it is prudent for the child to begin taking prophylactic Zantac of 2 mg/kg/dose, given every 8 h intravenously, to prevent stress ulcer development.

51.6.6 Hematological

Because blood loss may be profound, it is important to obtain a baseline hemoglobin and coagulation panel when the child returns from the OR. Monitoring of these parameters every 6 h for the first 24 h is not unreasonable, as many patients require packed red blood cell transfusion (over time, the threshold for transfusion has lowered, but many would agree that a hemoglobin <7 with a symptomatic patient is worthy of transfusion). Care should be taken to note the presence of a Jackson–Pratt drain and the amount of drainage per hour. Drainage that exceeds 3–5 mL/kg/h is excessive, and these children will often require transfusion or correction of coagulation factors.

51.7 Summary

Care for children with spinal deformities starts well before their admission for surgery.

The anesthesiologist must address surgical requirements for positioning and monitoring, in addition to taking into consideration the associated comorbidities, age-related pathophysiology, the potential for blood loss, and vascular injury of the spinal cord. Care for these patients requires a number of pediatric subspecialists, and close communication and dedication are essential to providing these children the best opportunity for a safe operation and recovery.

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Repetitive Anesthesia Concerns in Early-Onset Scoliosis

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Key Points

- Children requiring repeated visits to the operating room or imaging suite may develop severe anxiety and a host of behavioral responses.
- Repeated surgical procedures and negative memories of previous hospital experiences are among the most significant predictors of stress at the time of anesthesia induction.
- Multiple sources are available to prepare children for their visit: non-pharmacologic tools (parental presence, distraction, explanation), premedication, and hospital-based resources (preoperative visit, web-based information).
- In recent years, there has been growing concern regarding the potential neurotoxic effects of anesthesia on the developing brain. No study has provided evidence of a cause and effect relationship between anesthesia exposure and long-term neurocognitive impairment. The evidence available so far does not justify changing current practice, as the risk of delaying surgery or altering anesthetic management is also largely unknown.

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52.1 Perioperative Stress and Anxiety in Children

Although it was underappreciated in the past, children experience a significant amount of perioperative stress and many consider anesthesia to be the most terrifying period of their hospital stay. In one study of healthy children, undergoing mostly minor operations, 42 % displayed distress at induction, and close to 17 % displayed significant stress with at least three distressing behaviors including crying, screaming, verbal resistance, verbal expression of fear, and seeking emotional support [1]. Another study of healthy children observed 53 % to have high anxiety at induction [2]. These behaviors interfere with a smooth mask induction, decrease parental satisfaction, and place the child at risk for maladaptive behaviors postoperatively, including poor compliance with future medical therapy, especially anesthesia [3].

In anticipation of a surgical procedure, an adult may worry about awareness under anesthesia, postoperative pain, and nausea [4]. Children, however, have a very different set of stressors – threat of discomfort, parent separation, unknown and strange environment, losing autonomy and control, and uncertainty of acceptable behavior [5]. In addition to verbal expressions of fear, children exhibit behavioral changes such as agitation, increased muscle tone, shivering, cessation of playing, silence, spontaneous urination, and active attempts to escape. They also differ from adults in that it is difficult to identify those children who may become uncooperative or may have sudden and unexpected behavioral changes [6].

Children's perioperative concerns and behaviors are based on their developmental age. Infants form attachment with caregivers usually by 9 months of age but can start as early as age 3–6 months. Separation from parents may therefore cause stress in these infants. Their manifestations of stress include irritability, sleep disturbance, and decreased food intake [7]. Coping strategies are largely limited to crying and sucking. Toddlers have a sense of autonomy and want control but are difficult to inform about medical treatments. They are most likely to show "acute stress behaviors" that peak at mask induction including crying, screaming, and nonverbal resistance [1]. They may even feel that hospitalization is a punishment for bad behavior. Immobilization, along with pain, and separation are stressors at this age. Stress may manifest as feeding problems, sleep disorders, irritability or hyperactivity, loss of control of motor and bodily functions, and delay in language development. Preschool-aged children start to develop magical thinking. Like younger children, they fear separation from parents and "the needle." Stress in this age group can be manifest by somatic symptoms such as a stomachache. Hyperactivity or quiet anxiety may also emerge depending on the child's temperament [7]. Children at this age may display more "anticipatory distress" – verbal resistance and negative verbal emotions – peaking at the time they are notified that the mask will be placed [1]. Cognitive coping skills start to emerge such that role-play can become a powerful tool.

Older school-aged children, 7–12 years, are more reality based in their thinking and can understand direct information given to them about surgery and what to expect. They think more specifically about anticipated negative effects of surgery and their illness and how it will effect separation from friends and other activities. Stress manifestations become more varied as children age and can include anxiety, depression, stomachache, headache, hyperactive symptoms, and bed-wetting. At this age, regulatory behaviors that can reduce anxiety such as humor, nonprocedural talk, and information seeking start to become more common [1]. Finally, adolescents may feel their disease threatens their identity, as this is a time when body image is of utmost importance. They should be active participants in discussions about their disease and procedure. While teens normally have an outward appearance of being calm, over 80 % report significant anxiety at induction [8].

Children are a part of a family unit, and parents may experience even more stress and anxiety around their child's hospitalization than their children do. Parents caring for an acutely ill child may be faced with uncertainty, environmental constraints, and decreased confidence in

parenting [9]. Some parents may then become immobilized and paralyzed by fear while others overly anxious and overprotective.

52.1.1 Predictive Factors for Perioperative Anxiety

Identification of children at high risk for anxiety at induction may aid in tailoring a preventive strategy that is effective and avoids overmedication. In one Australian study, the negative memory of a previous hospital experience was the strongest factor in predicting preoperative anxiety in children. Other factors included number of people in the induction room and waiting time in the holding area [2]. A US prospective cohort study found that younger age, frequent hospital admissions, previous behavioral problems in healthcare settings, and anxious parents were the factors associated with anxiety at induction [10]. Other studies have identified additional risk factors, such as a child's temperament, coping style, and parent's coping skills, and underlying developmental and behavioral disorders [3, 11, 12].

Children aged 1–5 years have the strongest tendency towards anxiety at induction. Temperament, a child's innate personality, has been shown to influence how a child reacts to the stress of induction. One temperament scale, the EASI – Emotionality, Activity, Sociability, Impulsivity Temperament Scale [13] – has been used to study children and their reactions to the perioperative environment. Children with low scores of emotionality (shy, take a long time to warm up to strangers), activity (less energetic, prefer non-active games), and impulsivity (difficult to distract once they get upset, do not calm down quickly, tolerate frustration poorly) are more likely to experience anxiety at induction [11]. Traits that lead to better reactions to stress include intelligence, optimism, and creativity [7]. Coping style and support network also play a large role in shaping a child's reaction to stress. Children who have a passive coping style – being more withdrawn and quiet – are also at higher risk. Studies show no consistent prevalence towards gender and perioperative anxiety [11].

Parents' anxiety has been consistently shown to affect a child's response to hospitalization and surgery [10, 14, 15]. During the perioperative process, parents experience fear, discomfort, helplessness, and disorientation. Even young children can pick up on their parents' nonverbal cues, and these negative emotions can impact the induction process and subsequent recovery period [16].

Frequent hospitalizations can have a tremendous impact on the whole family. Each medical encounter provides us with the ability to shape the family's reaction to new and possibly painful situations, in either a negative or positive way. Early encounters are specifically important in shaping expectations and easing fears for future encounters. Clear communication between medical staff on what has worked in the past and communication of parents' and child's perception of previous encounters are all important. Of note, while respecting parent wishes and doing what is best for the child are often one in the same, this is not always the case. One example of an autistic teenager having multiple traumatic inductions highlights this fact. This patient presented eight times for general anesthesia over 4 months, with each induction becoming more combative and distressing. While the parents of the teen were comfortable fighting and holding him down for mask inductions or intramuscular (IM) injections of ketamine, the anesthesiologists felt uncomfortable with this process and sought a more comprehensive solution to decrease his anxiety [17]. Overall, repeated interventions leads to increased anxiety and should always be minimized when possible.

52.1.2 Behavioral Consequences of Perioperative Anxiety

Poorly controlled perioperative anxiety can lead to both short- and long-term behavioral changes. Separation anxiety, problems with sleep, nightmares, bed-wetting, aggression, regression, chronic anxiety, depression, and problems with long-term memory have all been reported negative postoperative behaviors. While most of these

will be short lived lasting less than 2 weeks and completely reversible, effects lasting up to 1 year have been demonstrated in some children. Increased postoperative pain, increased use of sedative medications, lowered defenses to infection, and increased incidence of emergence delirium are some of the immediate effects that can be seen. While preoperative anxiety and difficult induction play a large role, other factors that can contribute to these negative behaviors include previous mental health problems, low maternal participation in child's care, and lower support from parents and the healthcare team [11].

52.1.3 Decreasing Perioperative Anxiety: Non-pharmacologic Approaches

Parental presence is the most widely practiced non-pharmacologic technique to decrease preoperative stress. Data on parental presence is conflicting, most likely because each parent/child pair is different in terms of their quality of interaction in the OR and recovery room. A recent Cochrane review concluded that parental presence did not improve child anxiety scores or cooperation at induction [18]. Downsides of parental presence include increased stress for the anesthesiologist if a complication occurs, possible prolonged induction, and the potential for increased anxiety in the child if the parent is unsupportive [19]. Despite these concerns, parental presence is still a common practice and does tend to ease the trip to the OR especially for older children. Another reason to support parental presence is to respect the parents' wishes and keep them engaged in the decision-making process. This increases parental satisfaction and decreases parental anxiety [3].

Anesthesiologists and parents can help to distract the child and encourage coping behaviors during the induction period. One study trained anesthesiologist and nurses in "desired behaviors" and "undesirable behaviors" and taught parents to model after the anesthesiologist's lead. After implementing these behavioral modifications, they showed reduced anxiety in children at induction that equaled that of premedication. "Desired behaviors" included non-procedural talk (talking about friends, music, a favorite game, sports), humor, medical reinterpretation (reframing medical equipment as something fun – such as the mask as an "astronaut mask"), and providing developmentally appropriate procedural information. "Undesirable behaviors" that increased stress included reassuring statements, empathizing, and apologizing – which may all focus the child on their emotions, as well as excessive medical talk, and implying control in a situation where the child has none ("Are you ready to go?") [20].

Other effective strategies include creating a low sensory environment with dim lighting, having the least number of people necessary present for induction, allowing a comfort object in the OR, minimizing unnecessary clothing changes, and utilizing a video game device or television for distraction. Reduced wait time and ensuring an on-time OR are also important [2].

52.1.4 Decreasing Preoperative Anxiety: Premedication

Premedication is the most reliable way to decrease preoperative anxiety. It reduces anxiety in children and parents, increases parent satisfaction, and decreases postoperative maladaptive behaviors. After any premedication, medical staff must monitor the child and motion should be limited. Additional resources required for administering the medication and monitoring the child and time required to wait for the medication to take effect are all factors that limit their use in some settings. Leaving patients sleepy and potentially prolonging discharge can also be problematic in a high turnover recovery area.

Midazolam is the most commonly used premedication in children. It is a short acting benzodiazepine that provides sedation within 10 min and peaks in 20–30 min when given orally. Oral doses range from 0.25 to 0.75 mg/kg, the usual starting dose being 0.5 mg/kg to a maximum of 20 mg. Because it tastes bitter, a commercially available sweetened formulation is used. A low pH leads to burning if given nasally or intravenously

(0.05–0.1 mg/kg). In addition to anxiolysis and anterograde amnesia, mild sedation and loss of balance are commonly seen. The duration of action is 90 min. If cases are significantly delayed, an additional 0.25 mg/kg can be given [21].

Ketamine is another medication that can lead to calm separation from parents and good induction conditions. Its main advantage over midazolam is that an induction can be accomplished via intramuscular route within 5 min. It should therefore be given in a highly monitored setting, usually the OR or induction area. The IM dose is 4–5 mg/kg, and if used in combination with IM midazolam 0.1 mg/kg, the dose is lowered to 2–3 mg/kg. IM induction with ketamine is usually reserved for highly uncooperative children [21].

Dexmedetomidine and clonidine are alpha-2 adrenergic agonists that cause sedation, reduce emergence delirium, and have analgesic properties. Clonidine 2–4 mcg/kg given orally produces similar sedation to midazolam but has a slow onset (60–90 min). Dexmedetomidine has a shorter half-life and faster onset time than clonidine. It is usually given nasally as it is non-irritating and has poor bioavailability when given orally. A nasal atomizer is used to improve medication spread. Because onset time ranges from 25 to 45 min, it should be given at last 45 min before induction and lasts 90 min. Bradycardia and hypertension followed by hypotension may occur but are uncommon after premedication doses. For the child who is uncooperative or unable to receive an oral medication, nasal dexmedetomidine is a less traumatic alternative to IM ketamine, the main disadvantage being slow onset time [3].

52.1.5 Decreasing Perioperative Anxiety: Hospital-Based Programs

Hospital-based programs vary but many include a website or pamphlets dedicated to educating parents and children about the perioperative process, OR visits, and child life specialists who can do coaching over the phone and introduce the child to the mask and other hospital items. Toys in the holding area and a "surprise" given to the child at induction are used in some institutions. Especially for the older child, any interventions should occur early – ideally weeks before the surgery. In a toddler or infant, giving information to parents early but preparing the child on the day of surgery is usually sufficient. Hospital-based programs to help reduce anxiety are often costly. Although they are not proven to be superior to midazolam premedication, many parents favor a non-pharmacologic approach. It may be more beneficial to parents and patients who will have repeated procedures because they can learn coping skills that can be used for future inductions [22].

52.2 The Combative Child

Occasionally, a child is uncooperative and combative despite all efforts. While this is sometimes predictable based on risk factors discussed earlier, a seemingly cooperative child may simply be overwhelmed and become combative at any time. In these cases, restraint for an inhalation or IM induction is common. When parents believe that restraint is in the best interest of their child, they are likely to support its use. When parents are experienced, they may assist in restraining the child, but under normal circumstances, staff should perform the restraint so that it can be done speedily and decisively. A survey of pediatric anesthesiologists in the USA found that restraint was more commonly needed for infants and that practitioners' comfort level decreased with the age of the child such that 75 % would restrain a 6-year-old child and only 9 % would restrain a 15-year-old. The mean age at which anesthesiologists complied with the child's wishes to refuse induction was 12 years [6]. Cancellation is uncommon but does occur on occasion to allow the child and family to regroup and more adequately prepare for the induction and procedure.

Optimization of care for a child whose combativeness is predictable includes a preoperative visit, scheduling as first case of the day, premedication, distraction and play by the anesthesiologist and parents at induction, rewards for cooperative behavior, and the use of appropriate restraint for a mask or IM ketamine [6].

52.3 Anesthesia Neurotoxicity

In recent years, there has been growing concern regarding the potential neurotoxic effects of anesthesia on the developing brain in the lay media (FDA to Study Whether Anesthesia Poses Cognitive Risks in Young Children, New York Times, March 9, 2011). Initial concerns came from an animal study, which showed impaired learning in anesthesia-exposed rat pups [23]. Subsequently, many more animal studies have been published providing evidence that exposure of the immature brain to commonly used general anesthetic agents during a vulnerable age can lead to neuronal apoptosis, neurodegeneration, abnormal neurogenesis, and other cellular changes. In addition, these animals were found to have abnormal behavior and deficits in memory, learning, and motor function as adults. However, the mechanisms of anesthesia neurotoxicity remain to be elucidated, but a growing body of research is actively trying to explain it. Interestingly, data from recent animal studies suggests that vulnerability to anesthetic neurotoxicity may persist into adult life because it is not the age of the organism but the age of the neuron that is important in determining susceptibility to the neurotoxic effects of anesthetics [24].

Several cohort studies have been published to address the question: is anesthesia toxic for the developing brain? The data are conflicting with regard to the association of anesthetic exposure and impaired neurodevelopment. Results derived from a birth cohort of Olmstead County reviewed the medical and educational charts of 5,357 children. Five hundred and ninety three of them had received anesthesia before age 4. They assessed the risk of development of a learning disability before age 19 years by looking at individually administered IQ and achievement tests. They found that children having received a single exposure to anesthesia were not at increased risk of having a learning disability compared to unexposed children, but those receiving two or three anesthetics had an increased risk (OR 1.59 and 2.6, respectively). In this cohort, the incidence of learning disability diagnosed by age 19 year was almost twice as high (35.1 %) in children with multiple exposures to anesthesia and surgery compared to unexposed children (20 %). Of course, the requirement for anesthesia may be associated with another unknown factor associated with an increase in learning disabilities [25].

A birth cohort of 10,450 siblings derived from children who were enrolled in the New York State Medicaid program between 1999 and 2005 compared 304 exposed children to 10,146 unexposed to anesthesia before age 3 years. In this study that excluded many birth complications, previous diagnoses of developmental or behavioral disorder, and surgical interventions associated with neurodevelopmental disorders (i.e., bilateral myringotomy and tubes, neurosurgical procedures, etc.), multiple exposures to anesthesia were associated with a 60 % increase in subsequent diagnoses of developmental or behavioral disorders. The estimated hazard ratio for developmental or behavioral disorders associated with anesthesia exposure before age 3 was 1.1 for one surgery, 2.9 for two surgeries, and 4.0 for three or more surgeries [26].

These retrospective cohort studies do not provide causal inference between anesthesia exposure and impaired neurodevelopment, and continuing research in this area is still needed. While there is no evidence to support any change in practice at this time, these results are sufficiently concerning. They underscore the need to avoid unnecessary exposure to anesthesia and surgery at a young age if possible.

A recent study compared the different outcome measures used in the published studies to date [27]. Using data from the Western Australian Pregnancy Cohort (Raine), this study specifically compared children exposed to anesthesia prior to age 3 and unexposed peers using three different outcome measures: neuropsychological testing; International Classification of Diseases, 9th revision (ICD-9) codes; and academic achievement tests. When comparing 112 exposed children to 669 unexposed children, there was an increase in deficit when assessed by direct neuropsychological language assessment and ICD-9 codes, but not when comparing academic achievement scores. This may help explain the variation in the literature and emphasizes the importance of

understanding the outcome measures utilized to identify disabilities of cognitive function.

There are three large ongoing studies that are worth mentioning. The GAS trial is a large international multicenter randomized controlled trial comparing patients having inguinal hernia repair under regional or general anesthesia. The goal is to look at neurodevelopment outcomes with neuropsychological testing at ages 2 years and 5 years and postoperative apnea. The results of this trial will help elucidate whether there is a difference between regional and general anesthesia with respect to neurodevelopmental outcome. Additionally, it will also provide data for the comparative risks of postoperative apnea between general and regional anesthesia.

The Pediatric Anesthesia NeuroDevelopment Assessment (PANDA) study is a multicenter ambidirectional cohort study comparing siblings exposed or unexposed to anesthesia before age 3 year. Neuropsychological testing will occur between ages 8 years and 15 years. The use of siblings as the control comparison group will eliminate some of the known important confounding factors in neurodevelopmental outcome studies such as socioeconomic status and genetic background.

The Mayo Anesthesia Study of Kids (MASK) is also an ambidirectional cohort study that will perform direct neuropsychological assessment in a total of 1,000 children. The study will assess children with exposure before age 3 years, at two different age ranges: 6–11 years and 15–19 years. The study will examine both single and multiple anesthesia exposures. The results of the study will address both frequency of exposure and persistence of changes, if any.

It is worth noting that multiple confounding factors such as patient comorbidity, stress, and surgery are difficult to control in any study. No study has provided evidence of a cause and effect relationship between anesthesia exposure and long-term neurocognitive impairment. The evidence available so far does not justify changing current practice, as the risk of delaying surgery or altering anesthetic management is also largely unknown.

SmartTots, a nonprofit research initiative that is a public private partnership of FDA and the International Anesthesia Research Society (IARS), released a consensus statement in December 2012 stating: "it would be unethical to withhold sedation and anesthesia when necessary. Instead, healthcare providers should do the following:

Discuss with parents and other caretakers the risks and benefits of procedures requiring anesthetics or sedatives, as well as the known health risks of not treating certain conditions.
Stay informed of new developments in this area.
Recognize that current anesthetics and sedatives are necessary for infants and children who require surgery or other painful and stressful procedures."

This consensus statement, endorsed by the IARS and the American Academy of Pediatrics (AAP), is in the process of being revised, and any updates will be made available at www.smarttots.org.

The Food and Drug Administration (FDA) issued a consumer update in August 2013, "Anesthesia: Is it safe for young brains?" detailing the currently ongoing research efforts and specifically provided warning to parents against postponing necessary surgery. The conclusions were that the available data have been inconclusive, and more research is needed before we change our current practice.

Conclusions

Children requiring repeated visits to the operating room or imaging suite may develop severe anxiety and a host of behavioral responses. Multiple sources are available to prepare children for their visit: non-pharmacologic tools (parental presence, distraction, explanation), premedication, and hospital-based resources (preoperative visit, web-based information) may help alleviate some of the stress involved with these encounters.

The discussion surrounding anesthesia neurotoxicity has been increasing and most surgeons are now faced with a host of questions from parents regarding the risk to their children. Most experts agree that if surgery or imaging is required, we must remember that there is no definitive causal evidence that

neurotoxicity will result from being exposed to anesthesia. Withholding necessary treatment though has definitive risks. For completely elective procedures, it would seem prudent to consider the risks and benefits of delaying until the child is older. However, this age has not yet been defined. Research is ongoing and the results of three large multicenter studies, the GAS trial, PANDA study, and MASK study will help guide future management.

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Intraoperative Neurophysiological Monitoring During Corrective Spine Surgery in the Growing Child

53

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Key Points

- Multimodality neurophysiological monitoring of spinal cord and brachial plexus function has a definite place in the context of growing rod and vertical expandable prosthetic titanium rib (VEPTR) placement and lengthening/adjustment procedures.
- Transcranial electric motor-evoked potential (tceMEP) recording is the only viable method for monitoring the corticospinal tracts.
- Monitoring brachial plexus function with tceMEP and ulnar nerve SSEP recordings should be considered routine during VEPTR placement and adjustment.
- Total intravenous anesthesia and absence of neuromuscular relaxation optimize neurophysiological signal amplitude and reduce interpretation ambiguity.

53.1 Introduction

Operative management of scoliosis has undergone dynamic evolution over the course of the previous half century, particularly in the immediate past decade. Advances in multisegmental instrumentation and improved surgical technique have facilitated treatment of complex deformities,

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even in young children with progressive early-onset scoliosis who are unresponsive to nonoperative treatment. These developments in spinal instrumentation and surgical management have been complemented by dramatic improvements in anesthesia care and intraoperative neurophysiological monitoring (IONM) of spinal cord and spinal nerve root function. Both of these latter clinical specialties have played vital roles in making scoliosis surgery safe and effective across a broad range of patient populations.

This chapter discusses the role of intraoperative neurophysiological monitoring during surgical treatment of rapidly progressing spinal and/or thoracic cage deformity in young children, using temporary internal bracing.

53.2 What Neural Structures and Pathways Are at Risk?

In contrast to formal instrumented fusion of the spine involving placement of permanent fixation devices and application of multidimensional corrective forces, the insertion/adjustment of growing rods and other internal braces would seem to pose fewer risks of iatrogenic neurological injury. Although the neurological risks are likely to be diminished during these latter, less-extensive surgical procedures, they cannot be discounted completely. Assessment of risk to underlying neural structures follows from systematic analysis of the patient's pre-existing pathology in the context of the proposed surgical intervention. For example, treatment of early-onset scoliosis with growing rods requires anchoring of the rods to the spine, commonly with pedicle screws. Medial misdirection of pedicle screws in the lumbar spine poses risk of contusive injury to the spinal nerve roots, whereas in the thoracic spine, there is a risk of injury both to the roots and the spinal cord. Similarly, lengthening of growing rods, particularly in the presence of abnormal vasculature, spinal cord lesions, or hypotension, may compromise normal blood supply to the spinal cord and predispose it to hypoxic injury. Other techniques, such as those that use internal bracing to expand the chest wall, can inadvertently stretch the brachial plexus [1]. Consequently, lessons learned from monitoring the spinal cord, spinal nerve roots, and brachial plexus during traditional surgical correction of spinal deformity appear to have direct application during surgical treatment of deformities in the growing child.

Neurological injury to the spinal cord usually has a mechanical or vascular etiology. Mechanical insults in the form of direct contusion or distortion of a neural element by a surgical instrument or spinal implant, such as a sublaminar hook, tend to manifest globally, producing alteration of anterior and lateral motor and posterior sensory column function. By comparison, vascular insults due to stretch of critical vessels following lengthening or distractive maneuvers can present either as focal compromise to the motor tracts alone or more globally to include the sensory tracts. As a result, spinal cord monitoring *must* be a multimodality technique to allow for neurophysiological surveillance of both pathways [2].

Figures 53.1, 53.2, and 53.3 show a typical intraoperative setup for multimodality monitoring of neurological function during vertical expandable prosthetic titanium rib (VEPTR) and growing rod surgery. These electrode composites provide neurophysiological monitoring coverage of the spinal cord sensory and motor pathways, brachial plexus, neuromuscular junction, and adequacy of

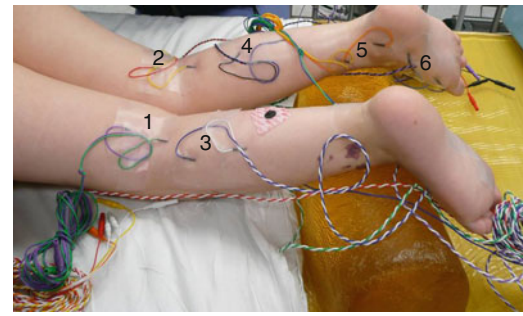

Fig. 53.1 Electrode positions for posterior tibial nerve SSEP, H-reflex and train of four (TOF). (TOF is monitored to ensure clearance of the neuromuscular junction for optimal tceMEP recordings.) (*1*) *Left,* popliteal fossa (H-reflex stimulation site); (*2*) *right,* popliteal fossa (H-reflex stimulation site); (*3*) *left,* gastrocnemius muscle (H-reflex and tceMEP recording site); (*4*) *right,* gastrocnemius muscle (H-reflex and tceMEP recording site); (*5*) *right,* posterior tibial nerve (TOF and SSEP stimulation site); (*6*) *right,* abductor hallucis muscle (TOF and tceMEP recording site). The *left* posterior tibial nerve stimulation electrodes are not shown

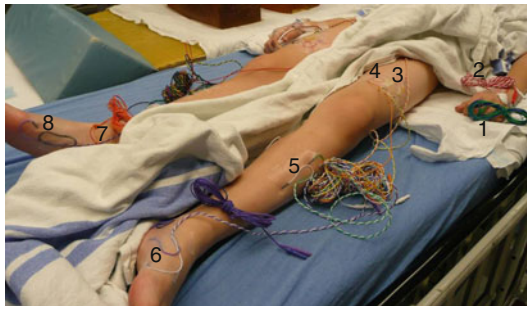

Fig. 53.2 Electrode position for recording upper and lower extremity tceMEPs. (*1*, *2*) *Left,* first dorsal interosseous muscle; (*3*, *4*) *left,* quadriceps muscle; (*5*) *left,* tibialis anterior muscle; (*6*) *left,* abductor hallucis muscle (also used for TOF recording); (*7*) *right,* posterior tibial nerve SSEP and TOF stimulating site; (*8*) *right,* abductor hallucis muscle (also used for TOF recording)

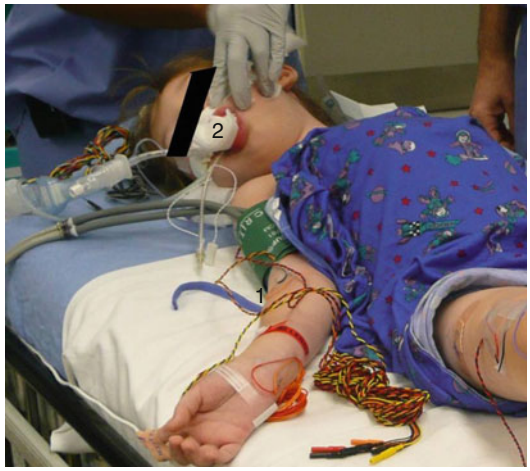

Fig. 53.3 (*1*) Recording electrode over the *right* deltoid and biceps muscles for upper extremity tceMEP recordings during VEPTR procedure. (*2*) Position of the bite block for tongue bite protection

anesthesia. The specific recording electrodes for cortical and subcortical somatosensory-evoked potentials (SSEP) and EEG as well as the stimulating electrodes for transcranial electric motor-evoked potential activation are not shown.

53.3 Neuromonitoring Modalities

Despite the early historical success of SSEP monitoring during surgical correction of scoliosis, reliance on this modality is no longer adequate for the growing population of patients who present for surgical correction of increasingly complex deformities. As the SSEP is mediated by the posterior sensory columns and reflects integrity of spinal cord white matter, it provides no direct information about the condition of the descending motor tracts or spinal cord gray matter structures which are particularly susceptible to vascular insult. Hence, when used alone to monitor spinal cord function, SSEPs carry a definite risk of false-negative findings, even among patients with adolescent idiopathic scoliosis [1, 3, 4]. Because of the distractive or lengthening maneuvers needed for deformity correction both with traditional fusion and contemporary nonfusion techniques, there is increased opportunity for excessive vascular stretch and ischemic spinal cord injury. This may not manifest in the SSEP, either at all, or within the critical period necessary to initiate timely intervention for injury reversal.

In response to the limitations of SSEP monitoring, other techniques have been introduced to assess the descending spinal cord motor tracts and anterior horn function. These include transcranial electric motor-evoked potentials (tceMEPs) and the Hoffmann reflex (H-reflex). The highly debated and often misunderstood neurogenic "motor"-evoked potential (NMEP) is discussed separately in this chapter because of several seminal studies that point to its sensory origin.

53.3.1 Transcranial Electric Motor-Evoked Potentials

tceMEPs are neuroelectric events elicited from descending motor pathway structures including the corticospinal tract (CST), spinal cord interneurons, anterior horn cells, peripheral nerves, and skeletal muscles. These potentials are triggered by delivering electric pulse trains to the brain through subdermal scalp electrodes over the motor cortex, as illustrated in Fig. 53.4. Following depolarization of the cortical motor neurons, efferent neural signals course through the internal capsule to the caudal medulla where CST fibers decussate and descend into the spinal

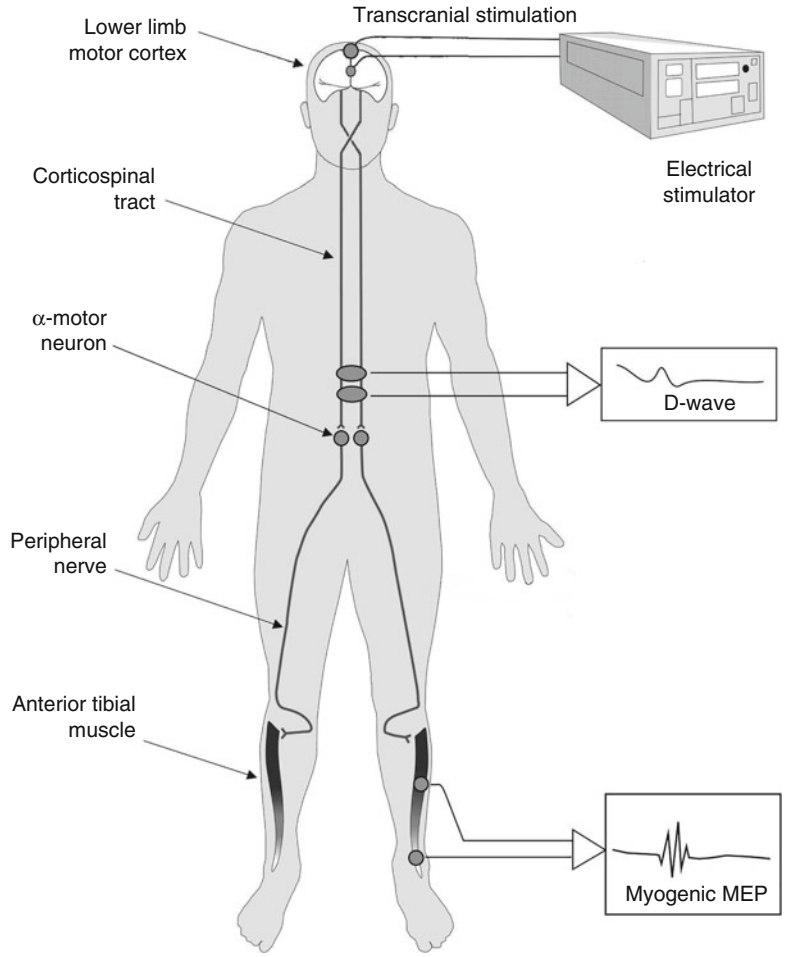

Fig. 53.4 Schematic of tceMEP stimulation and recording

cord motor tracts. CST axons enter the spinal cord gray matter, interact with spinal interneurons, and go on to synapse with alpha motor neurons that innervate peripheral muscle.

Compound muscle action potentials representing motor-evoked potentials can thus be recorded from upper and lower extremity peripheral muscle with subdermal needle electrodes at the end of this neural chain. Because of the high sensitivity–specificity of tceMEPs for the identification of spinal cord and spinal nerve root injury [3–7], they should now be considered as the gold standard for monitoring spinal cord motor function during complex spine surgery.

The sensitivity of tceMEPs to motor pathway insult is illustrated in Fig. 53.5 which shows the time course of tceMEP monitoring in a 9-year-old female undergoing revision of growing rod for the treatment of neuromuscular scoliosis. This child presented preoperatively with bilateral upper and lower extremity weakness, though she was weight-bearing and capable of taking several steps with support. Reference to Fig. 53.5 shows that soon after placement of pedicle screws at T2–3, there was acute tceMEP amplitude diminution at left tibialis anterior (TA) and right abductor hallucis (AH) recording sites and complete loss of the right tibialis anterior muscle response. Moments later, the patient became hypotensive with a mean arterial pressure (39 mmHg) well below the threshold level for spinal cord ischemia (see Chap. 51 for further discussion). At this time, there was bilateral loss of lower extremity tceMEPs, with the exception of a barely observable response (10 % of baseline amplitude) from the left AH muscle. Note,

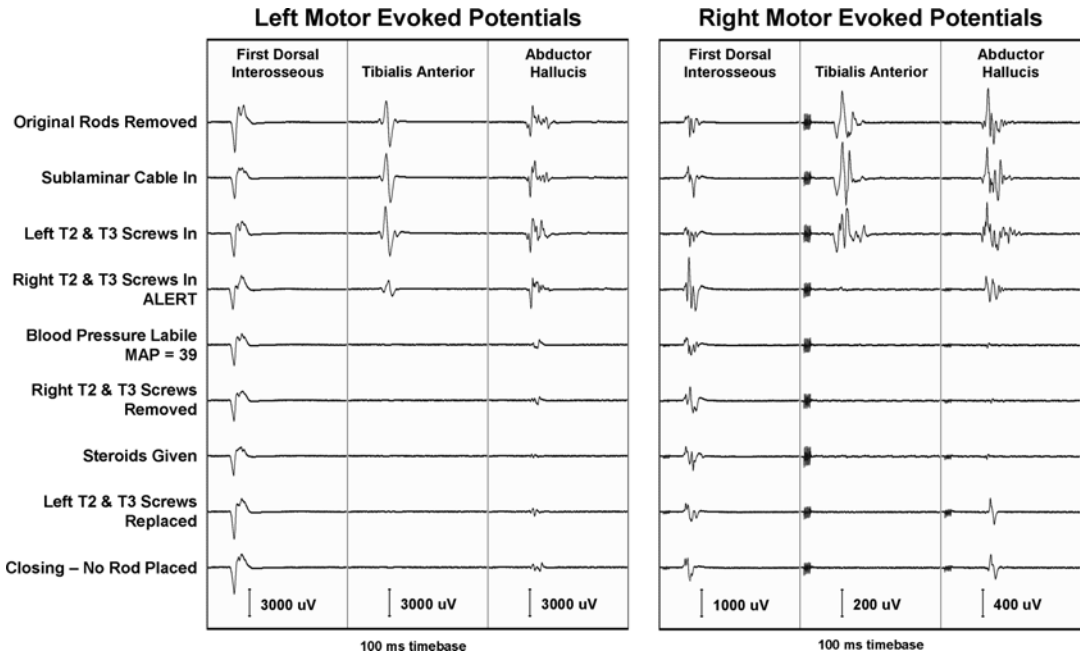

Fig. 53.5 Time course showing acute tceMEP loss following pedicle screw placement at T2–3 in a 9-year-old female undergoing revision of growing rod for treatment of neuromuscular scoliosis

however, that control responses from the left and right upper extremities (first dorsal interosseous muscle) remained unchanged.

Despite all attempts to raise the blood pressure, replace screws, and begin a spinal cord injury course of methylprednisolone, the tceMEP amplitudes never improved. Predictably, the child emerged from anesthesia exhibiting further deterioration of lower extremity motor function.

53.3.2 The Hoffman Reflex

The H-reflex, recorded from gastrocnemius muscle following stimulation of the tibial nerve at the popliteal fossa, is a neurophysiological correlate of the ankle stretch reflex influenced by upper motor neurons and other supra-segmental structures of the spinal cord and brain. The H-reflex reflects the functional integrity of complex motor control subsystems in the highly integrated ascending, descending, and interneuronal pathways of the spinal cord. It is thought that severe, acute spinal cord injury results in suppression of the H-reflex, which is observable within moments of insult. When amplitude suppression has exceeded 90 % and has persisted for the rest of the surgery, patients have awakened with profound postoperative neurological deficit consistent with spinal shock [8].

Because of its neurophysiological underpinnings and the fact that the anesthesia requirements for recording an H-reflex are not as restrictive as those for tceMEP monitoring, a minority of clinical neurophysiologists have proposed H-reflex monitoring as an equivalent substitute for tceMEPs [9]. Hicks [10] compared the results of H-reflex monitoring to the postanesthesia ankle clonus test in a diverse population of 292 spine surgery patients. The H-reflex predicted the outcome of the ankle clonus test in 80 % of the cases, leading Hicks [10] to conclude that the H-reflex was a reliable predictor of spinal cord injury during spine surgery. Of the three patients in the series who exhibited loss of the H-reflex intraoperatively, all had subsequent recovery to baseline prior to closure. There were no patients with permanent loss of the H-reflex and none with postoperative deficits, making comparison of results with known sensitivity of tceMEPs to spinal cord injury difficult at best.

Our experience with the H-reflex has not been as positive as that reported by others, particularly in very young children. All too often, the responses are either variable or there is significant inter-leg amplitude asymmetry for recorded H-waves. Moreover, we have observed dissociations between H-reflex responses and tceMEPs on several occasions, suggesting that the former may be less sensitive than the latter to predisposing factors for spinal cord injury.

An example of such dissociation is illustrated in Fig. 53.6. This figure shows intraoperative transcranial electric motor-evoked potentials and H-reflex responses recorded from a 42-month-old female during a third lengthening of growing rod to treat a 68° kyphotic deformity of the thoracic spine. The patient's history is significant for the resection of thoracic teratoma at 1 month of age, initial placement of growing rod at 10 months with removal at 15 months, and subsequent biopsy of spinal cord lesion with reinsertion of growing rod at 23 months of age.

Following lengthening of the growing rod, tceMEPs disappeared from multiple lower extremity myotomes, including bilateral tibialis anterior, gastrocnemius, and abductor hallucis muscles. There were no concomitant changes in the H-waves recorded from bilateral gastrocnemius muscles during this period. Upon decrease in distraction and elevation of mean arterial blood pressure, motor-evoked potential amplitudes returned to baseline range. There were no new postoperative neurological sequelae. In light of such examples, where H-reflex responses have been insensitive to surgical and physiological conditions known to be risk factors for postoperative deficit, we have come to view its role in neuromonitoring somewhat differently from

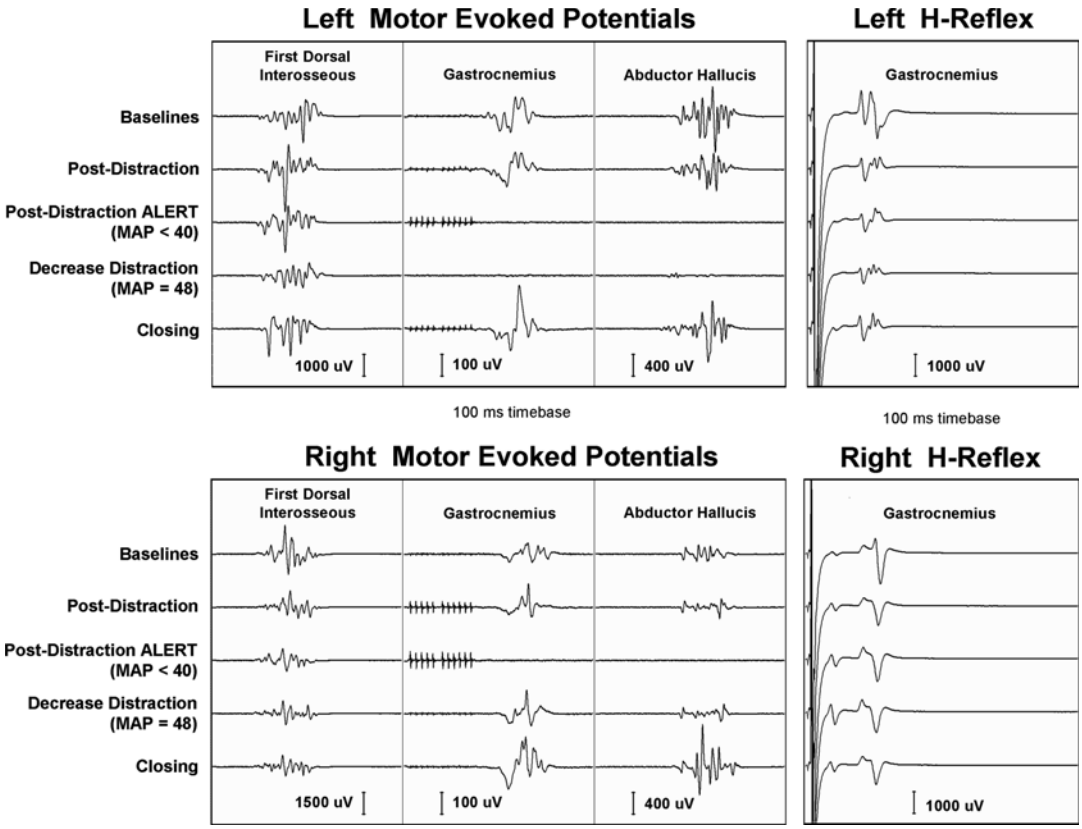

Fig. 53.6 Example of the dissociation between transcranial electric motor-evoked potentials and H-responses recorded from a 42-month-old female during a third lengthening of growing rod. Note the loss of tceMEPs with no significant change in H-responses

that described by Hicks [10] or Toleikis and Toleikis [9].

Unlike tceMEPs which are both highly sensitive and specific for evolving spinal cord injury, the H-reflex appears to be more specific than being sensitive. That is, while presence of the H-wave cannot be equated to a prediction of no neurological injury, acute loss of the response could point to spinal shock. For that reason, we would caution not to view the H-reflex as a substitute for tceMEP monitoring, but rather as an adjunct. Moreover, in the presence of discrepancy between the results of the two monitoring modalities, greater interpretive weight should be given to tceMEP findings. To be sure, it is not a "new standard" as posited by Toleikis and Toleikis [9].

53.3.3 Neurogenic-Evoked Potentials are Sensory Not Motor

There is considerable evidence dating back to 1992 that the NMEP predominantly reflects the functional integrity of posterior column sensory tracts rather than lateral and anterior column motor pathways [11–18]. The general conclusion drawn from these studies is that the NMEP is triggered by antidromic activation of posterior column sensory fibers that communicate with alpha motor neurons via collateral branches at a segmental level. Thus, NMEPs are insensitive to a wide variety of surgical and vascular insults to the motor tracts of the spinal cord by virtue of the sensory pathways that mediate them. As such, it may be more appropriate to label these responses NSEPs (neurogenic sensory-evoked potentials) or DASEPs (descending antidromic sensory-evoked potentials).

Despite the body of evidence pointing to a sensory origin for the NMEP, it continues to have its proponents as a viable technique for monitoring the spinal cord [19, 20]. While there is a general consensus in the surgical neurophysiology community that its use in place of the tceMEP provides a false sense of protection for spinal cord motor tract function, the ultimate decision to continue monitoring with NMEPs lies with the neurophysiological monitoring personnel and surgeons at each hospital facility [21].

In our experience, neurogenic-evoked potentials have played a limited role as an adjunctive modality for monitoring spinal cord sensory function, since we began the routine use of tceMEPs to monitor the motor tracts over 15 years ago. The contribution of neurogenic-evoked potentials is further limited by observations that they can disappear in the presence of rigid spinal instrumentation due to current shunting [22]. This creates interpretative ambiguity for the neuromonitoring professional and can result in a false-positive result just at the time of deformity correction, leading to unnecessary intervention.

53.3.4 Monitoring Spinal Nerve Roots

To the extent that growing rods are anchored to the spine with pedicle screws, individual spinal nerve roots may be at risk of injury from medial pedicle breaches and should be monitored using spontaneous and electrically stimulated electromyography, as well as tceMEPs [7]. The principles and techniques for testing pedicle screw placement with electrical stimulation described in considerable detail for the mature spine by Schwartz et al. [7] are equally applicable to the growing spine. Note that stimulation threshold criteria for detection of medial pedicle breaches may have to be adjusted downwards from those reported for adults to account for smaller pedicles in children. An increase in the use of image-guided navigational placement of pedicle screws coupled with either O-arm or fluoroscopic post hoc analysis of screw placement has also gained popularity among spine surgeons. This technique allows the surgeon to visualize the complete path of the pedicle screws to rule out medial, lateral, or other potentially unsafe trajectories at a sensitivity equivalent to traditional pedicle screw testing [23]. In some centers, pedicle screw placement under O-arm navigation has supplanted stimulated EMG for detection of medial pedicle wall fracture from an improperly directed screw, while in others, stimulated EMG serves as an adjunctive test to the O-arm gold standard.

53.3.5 Monitoring the Brachial Plexus

There has been recent interest in the evaluation and treatment of patients who have a congenital spinal deformity together with chest-wall abnormalities leading to thoracic insufficiency syndrome. The VEPTR has been effectively utilized for the treatment of these challenging patients by expanding the chest during the growing years to allow for maximum lung development, while also treating the spinal deformity. As is common with a new technique, there is a debate on the value of intraoperative neuromonitoring during VEPTR surgery.

The most commonly reported neurological complication during VEPTR procedures is brachial plexus injury [24–26]. In a multicenter investigation on the use of neuromonitoring, Skaggs et al. [26] reported that 8 of 299 (2.3 %) children presented with new-onset postoperative neurological sequelae. Of these, 6 patients showed deficits limited to the upper extremities, which resolved within 12 months for 5 patients and were incompletely resolved after 4 years for the sixth. They emphasized two potential underlying causes. First, the brachial plexus often drapes over the first rib and may be subject to compression or entrapment during VEPTR expansion; therefore, they recommended that the device not be placed at this level. Second, while lifting the scapula, pressure may be applied inadvertently to the brachial plexus, particularly in patients with a hypoplastic chest, requiring adjustment of retractive forces when alerted by neuromonitoring changes.

Risk to the brachial plexus and other upper extremity peripheral nerves is not limited to VEPTR procedures. Figure 53.7 shows one example of unilateral tceMEP and upper extremity SSEP loss in a 5-year-old female undergoing growing rod lengthening. Her status was post Chiari decompression and repair of myelomeningocele. During hook placement, there was an acute tceMEP amplitude decrease followed by complete loss of the response from the left first dorsal interosseous muscle (FDI), accompanied by a >60 % attenuation of the left ulnar nerve SSEP. Unchanged tceMEPs from both legs and the right hand pointed to the left upper extremity as the site of emerging injury. Upon repositioning the left arm, both the tceMEP and SSEP responses returned to baseline.

In our experience, one of the tangential benefits of intraoperative neuromonitoring is the detection of impending brachial plexopathy or other peripheral neuropathies [1, 27]. As in the case of the spinal cord, monitoring sensitivity for peripheral nerve injury is improved by using both sensory- and motor-evoked potential modalities [28].

53.3.6 Anesthesia Considerations

The role of anesthesia for optimized delivery of IONM care has been both underestimated and misunderstood by many. IONM personnel all too often have an inadequate understanding of the goals of anesthesia and the pharmacodynamics of different anesthetic agents, as well as how each agent interferes with the generation of neurophysiological signals. Likewise, it is not uncommon for the anesthesiologist or nurse anesthetist to have an equally poor appreciation of the influence of such agents and their aggregate effect on the neurophysiological signals.

All general anesthetic agents depress synaptic function in the brain and spinal cord gray matter, resulting in amplitude suppression of neurophysiological signals that cross those synapses. The goal, then, is to meet the conventional anesthetic requirements of amnesia, hypnosis, analgesia and akinesis without compromising neurophysiological signals to the point where they are too small or variable for meaningful interpretation.

Inhaled volatile agents, such as isoflurane, desflurane, sevoflurane, as well as nitrous oxide, present the greatest challenges of synaptic suppression and consequent depression of cortical SSEP and tceMEP amplitudes [29–36]. While some claim to be able to record neurophysiological signals in the presence of these potent anesthetics, they do so under suboptimal conditions, even when the agents are present at low levels of concentration. In these situations, there is

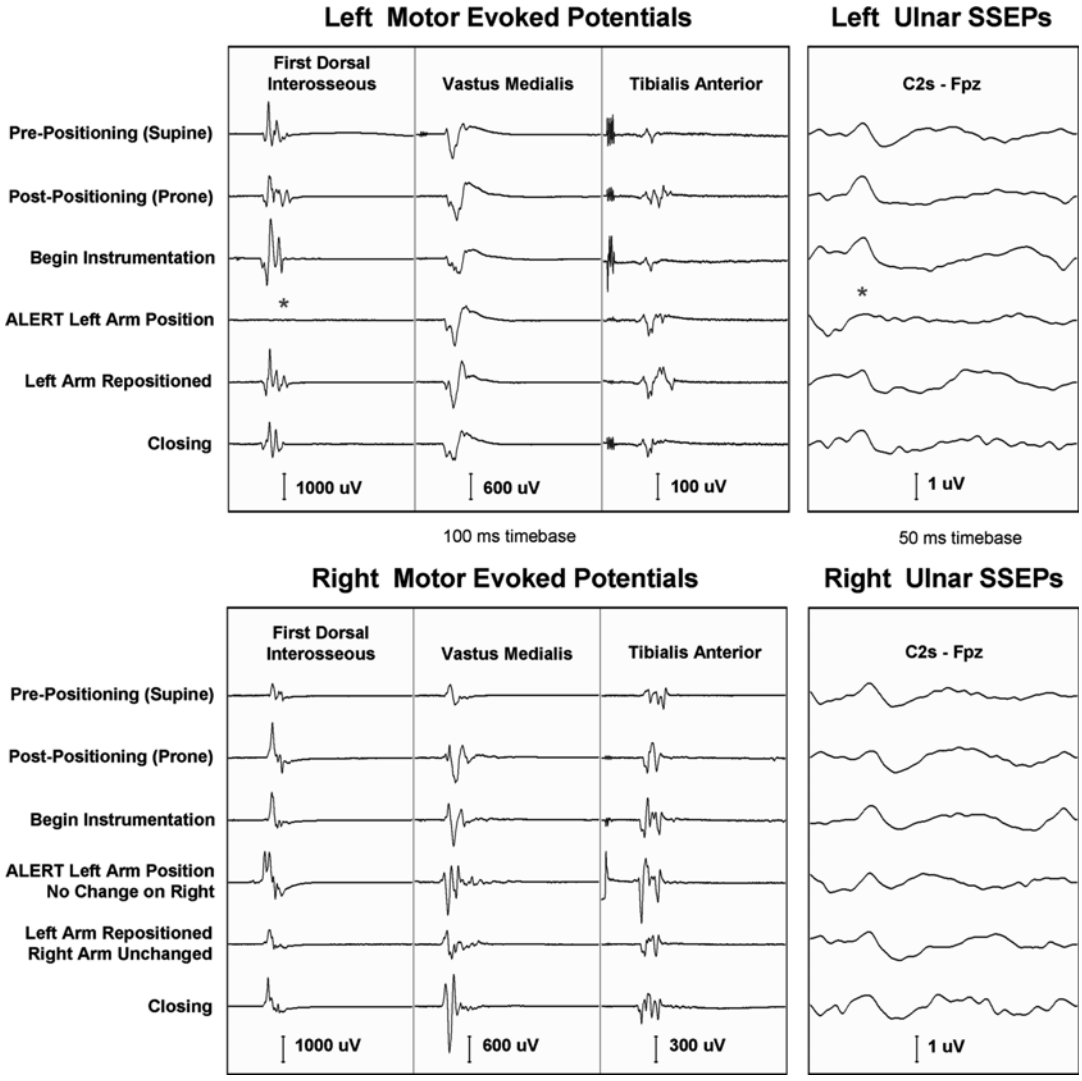

Fig. 53.7 Example of unilateral tceMEP and upper extremity SSEP loss in a 5-year-old female undergoing growing rod lengthening indicative of emerging brachial plexopathy

increased risk of interpretive ambiguity or overall inability to monitor, owing to the fact that the signal amplitudes are either near the physiological noise floor or completely absent [37]. Schwartz et al. [35] have stated that the use of these potent anesthetic agents (and nitrous oxide) is perhaps the biggest reason why many surgeons and neuromonitoring personnel complain of an inability to record stable and acceptably large-amplitude tceMEPs.

To circumvent the amplitude-suppressive effects of these inhalational agents and nitrous oxide, we transitioned to a propofol-narcotic total intravenous anesthetic technique soon after propofol was introduced some 25 years ago, initially, to optimize cortical SSEPs and, later, transcranial electric motor-evoked potentials. Using constant infusion delivery, average propofol concentrations of 150 μg/kg/min, in combination with remifentanil (0.2–0.5 μg/kg/min) and intermittent dosing of midazolam (0.1–0.2 mg/kg) provide adequate amnesia, hypnosis, and analgesia for the majority of spine patients, both pediatric and adult, without compromising tceMEP and

cortical SSEP amplitudes. (Caveat: propofol infusion rates that cause burst suppression on EEG will have significant depressive effects both on cortical SSEP and tceMEP amplitudes, similar to inhalational anesthetics; therefore, it is best to maintain a range between 125 and 175 μg/kg/min).

Drugs which act at the neuromuscular junction have profound effects on tceMEP and EMG monitoring, both of which depend on nerve depolarization and innervated muscle contraction. Any use of neuromuscular blockade either will diminish tceMEP amplitude significantly or abolish the response completely. In keeping with the theory of maximizing neurophysiological response amplitudes to optimize interpretation of signal change, no muscle relaxants should be used except to facilitate intubation.

Recently, dexmedetomidine, an α-2 agonist, has been suggested as an anesthetic adjuvant during spine surgery because of its sedative, analgesic, and neuroprotective properties [38]. Adding dexmedetomidine also enables reduction of propofol requirements, which helps facilitate more rapid emergence at the conclusion of surgery. We have been evaluating the effects of dexmedetomidine on tceMEP and cortical SSEP amplitudes over the last few years. In general, a marked dose-dependent suppression of tceMEP amplitudes without concomitant changes in cortical SSEP amplitudes has been observed when holding propofol steady at 100 μg/kg/min and varying the dexmedetomidine infusion rate between 0.2 and 0.7 μg/kg/h [38]. Until the effects of dexmedetomidine on tceMEP amplitudes are better understood, it is best to titrate infusion rates for each individual patient by establishing the threshold at which tceMEP amplitudes begin to decrease.

53.3.7 Hemodynamic Considerations

Nonanesthetic factors that can influence the size of neurophysiological signals include spinal cord perfusion pressure, mean arterial blood pressure, hematocrit, and blood volume. Although regulation of spinal cord perfusion is not as well understood as its cerebral perfusion counterpart, similar principles seem to apply (for further discussion see Chap. 51). Hence, decreases in mean arterial blood pressure below the autoregulation threshold for adequate spinal cord perfusion will result in a significant decrease in tceMEP amplitudes. We believe that if spinal cord blood flow and the associated delivery of oxygen and nutrients to neural tissue are reduced around the same time when spinal corrective maneuvers (i.e., growing rod lengthening) are applied, there may be increased risk of spinal cord injury.

All too often, we have noticed that children undergoing surgery for scoliosis correction are volume-depleted both prior to and during surgery. This challenges maintenance of a recommended mean arterial blood pressure, preferably 65 mmHg or more, to ensure adequate spinal cord perfusion pressure during deformity correction. Treatment of the ensuing hypotension with an alpha-agonist such as phenylephrine usually offers only temporary relief by elevating blood pressure transiently and thus increasing tceMEP amplitudes. What follows, however, is a picket-fence phenomenon, where the mean pressure drops again, as do tceMEP amplitudes, until pharmacologic intervention is reinitiated. It is usually more productive to address issues related to volume depletion, blood loss, and hematocrit proactively rather than reactively. Given the sensitivity of the spinal cord to ischemic injury, avoidance of prolonged controlled hypotension as a strategy for minimizing intraoperative blood loss is highly recommended.

53.3.8 Neurodevelopmental Considerations and Technical Challenges

The developing nervous system in the pediatric population poses its own clinical and technical challenges when utilizing IONM. The two main factors that the surgical team (surgeon, neurophysiologist, and anesthesiologist) must address are: (1) the effect of neurodevelopment on axonal depolarization and conduction and (2) presence and progression of neuromuscular diseases or

muscular dystrophies which affect the ability to monitor functional motor activity.

The primary factors affecting axonal depolarization and conduction are quite simply the normal progression of neural development and myelination, which in the pediatric population is expectedly incomplete. Consequently, when challenges arise in acquiring reliable baseline sensory- and/or motor-evoked potentials in very young patients, neuronal immaturity should be considered as the underlying cause. Oftentimes, this obstacle can be overcome by optimizing anesthetic technique as described in Sect. 53.3.6 and adjusting the stimulation and recording parameters.

The tceMEP is a response triggered by high-frequency electrical stimulation and as such is sensitive to slower axonal conduction secondary to smaller diameter axons and incomplete myelination in the pediatric population. Typical technical adjustments for improved triggering of tceMEPs include increasing the number of stimulus pulses, pulse width, voltage, and inter-pulse interval from that normally used with adults. Additionally, responses can be facilitated by using a double-train stimulation technique, incorporating inter-train intervals ranging from 10 to 15 ms. It may also be necessary to increase the duration of the time base used to record tceMEPs in infants and toddlers, given that their response latencies may exceed those of adults.

As an averaged response, the SSEP is also susceptible to the level of myelination in the growing child. Incomplete myelination results in slower and asynchronous axonal conduction, reducing the effectiveness of averaging. Recording SSEPs from the cervical spine is one solution that preserves the ability to evaluate dorsal column conduction and avoids signal degradation secondary to conduction delays and variability at the level of medulla oblongata, thalamus, and cortex. Additionally, decreasing stimulation rate provides additional time for recovery from prolonged refractory periods which can contribute to degradation of the averaged SSEP. [Caveat: decreasing stimulation rate increases the time required to obtain an averaged somatosensory response].

Overall, studies of IONM feasibility and utility in very young children, while relatively few in number, are promising [39–41]. Changes in intraoperative neuromonitoring data have been shown to be predictive of outcome in infants as young as 2–5 months of age [39, 41].

Neuromuscular diseases and muscular dystrophies affecting the spine of the growing child pose their own challenges in obtaining reliable baseline IONM data for effective monitoring during corrective spinal surgery. The techniques employed to improve reliability of IONM baseline signals in these patients are similar to those described above for the immature nervous system. DiCindio et al. have reported that while most patients with mild or moderate cerebral palsy (CP) or non-CP-related neuromuscular scoliosis can be monitored successfully, those CP patients who do not have weight-bearing ability and have significant neurological involvement are less likely to have monitorable sensory- and motor-evoked potentials [42]. Given the difficulty of predicting preoperatively which severely compromised children will have monitorable neurophysiological signals, some surgeons elect to test these patients in the operating room prior to incision to determine feasibility of continued neuromonitoring. Future research focused on preoperative identification of good candidates for neuromonitoring in the severely compromised pediatric population is necessary to improve surgical planning and appropriate IONM resource utilization.

Conclusions

The use of intraoperative neuromonitoring during growing rod and VEPTR procedures is still debatable. Our experience has been that neuromonitoring serves a valuable purpose and is well justified for routine use both during initial implant placement and subsequent adjustment. The sensitivity of tceMEPs to emerging spinal cord injury has been well established; however, the value of neuromonitoring for the detection of brachial plexus and other positionally related peripheral nerve injuries during these procedures should not be underestimated.

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Nursing Care

54

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Key Points

1. Patient and family education by the nurse or similar health-care professional is critical to improve the quality of care.
2. To be a valuable member of the team caring for children with complex early-onset scoliosis, nurses should attempt to learn as much as possible using all available resources.
3. Well-informed nurses can help surgeons provide early detection of potential complications.
4. Treatment of early-onset scoliosis is a long-term commitment by the family and the medical team. Nursing support is essential.

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54.1 Introduction

The care for children with early-onset scoliosis (EOS) is demanding not only on the child, but the parents, family members, and health team members who must deal with the long-term aspects of not only the treatment, but, in many cases, the other medical problems stemming from their disease process. The role of the nurse or similar health-care professional is critical in assessing, evaluating, and providing appropriate care at the appropriate time.

54.2 Family Education

Often, families of children with progressive EOS have obtained multiple surgical opinions for their child's care. It is not uncommon for these surgical opinions to vary, thus leading to frustration on the parents' part due to the lack of evidence-based information available to them. Fortunately, the number of scientific presentations and publications continues to grow, increasing the knowledge of the results and of the current treatment methods available. Families should strongly consider going to a center that is actively involved in the evaluation and treatment of EOS, as well as participation in the research needed to answer the complex questions stemming from this medical problem. Once the family has been evaluated by a surgeon at such a center, family education can then be initiated by both the surgeon and his or her educational liaison.

Regardless of the recommended treatment plan (nonoperative vs. operative), the family must remember that EOS treatment is a long-term commitment on the part of the child and the family. Whether the treatment involves serial cast applications, bracing, surgical placement of growing rods or a vertically expandable titanium rib (VEPTR) device, etc., the care of the child must be consistent until the end of the treatment period. This can be quite daunting to the family, especially if treatment is recommended at a very young age. In addition, the child's family must be fully aware of the treatment options, the benefits and risks (potential complications) of each option, and the anticipated treatment period (in years). Willingness to commit to long-term care must be expressed by the parents in order to move forward with any treatment program.

Many of the EOS centers are now study sites of the Growing Spine Study Group (GSSG), a group of international spine surgeons who are committed to learning the most effective and safe ways to treat EOS and other early-onset spinal deformities. As a result, part of the educational process will be to ask the family to consent to having their child participate in prospective studies (involving an ongoing review of the child's medical records and diagnostic studies). These centers emphasize that participation in the GSSG's studies will in no way change the care offered to their child, but allows the group to collect observational data on many children over time to better study various treatments, especially by subgroups such as diagnosis, age, etc.

Education for the family must always consider the child first and then the spinal deformity. Growth and development milestones are critical at very young ages. Treatment and the education about treatment should keep growth and development in mind, especially in the cognitively normal and ambulatory patient populations. As many of the treatments either can be repetitively invasive or impose upon body image problems (long-term casting or bracing), keeping the family focused on its child's developmental milestones as well as attempting to maintain balance in the family is important. Other siblings may feel that they are not getting attention due to their "sick" brother or sister. Parents should be reminded to keep the family in focus to avoid this pitfall. Also, many children with EOS may be quite active physically and cognitively, and every effort should be made to allow the child to engage in age-appropriate activities. This will help reduce the child's image of being "sick" or appearing "sick" to others. For example, a child having growing rods inserted may need to wear a special thoracic-lumbar-sacral orthosis (TLSO) brace for the first 6 months to allow the proximal and distal bone anchors to heal. After that, the child is weaned out of the brace for the duration of the treatment, which may be many years, and is

allowed to participate in most physical activities. This will help improve the child's self-esteem and self-image. Parents are reminded that potential risks of this relatively unrestricted activity could result in a problem such as a rod fracture, which is anticipated as a possible treatment outcome and is often treated at the time of the next scheduled lengthening of traditional growing rods (TGR) to avoid unnecessary surgery.

Each surgeon and his or her team must establish rapport with the patient and family for the best long-term success. Families who are well informed about commitment and potential risks are far more cooperative than those who have not been allowed to have an active role along with the treatment planning team.

It is also important to note that families will likely to go through financial, social, and emotional stress at any time when they deal with a long-term health problem of a family member. The treatment of EOS is no different. Families may find themselves reaching their maximum insurance coverage and having to seek state aid to continue receiving care. The doctor and EOS team need to be very in tune with the family's needs and concerns and be ready to assist. Counseling may be necessary for those requiring financial, social, or other assistance.

54.3 Nonoperative Care

54.3.1 Bracing

A brace or orthosis is named for the part of the body in which it supports. A TLSO may be prescribed as a nonoperative treatment for scoliosis. The purpose of the brace is to stop the curve from progressing.

To clean, condition, and prevent skin breakdown, rubbing alcohol and cornstarch are applied before each brace application. Rubbing alcohol is applied at the waistline and where the edges of the brace touch the skin, and it dries quickly. Cornstarch is applied to the same areas where the rubbing alcohol was applied to help absorb skin moisture. Do not use creams, lotions, or Band-Aids under the brace because they may cause a rash or skin breakdown. The skin should be observed for areas of redness, irritation, discoloration, or puffiness. This may indicate a need for the brace to be adjusted.

Children can wear any clothes in which they are comfortable. Pants or shorts may need to be one to two sizes larger in order to fit. In general, clothing with elastic waistbands or drawstrings is easier to fit. Loose-fitting clothing will usually not reveal the outline of the brace.

Physical activity and playing are important for children. All activities are encouraged if the brace is being used as a nonoperative treatment. However, the brace should be removed before swimming, showering, and tub bathing.

54.3.2 Serial Casting

Serial body casts, such as Risser or Mehta derotation casts, are another form of nonoperative treatment. The purpose is to stop the curve from progressing or delay surgical intervention [1, 2]. Sometimes, the curve may improve with casting [3, 4].

Parents are usually pleasantly surprised on how well a child adapts to wearing the body cast once the initial adjustment is completed. Parents have commented that the cast is not only more comfortable for their child; it is better tolerated than wearing a brace.

If the cast is utilized as a nonoperative treatment, there are very few restrictions. Most pertain to water: tub baths, showers, or swimming with the cast is not permitted as the cotton web roll or padding would absorb the water and act as a sponge. Playing around sand is not advised as the sand can easily lodge under the cast and become abrasive. Other restrictions may be disorder specific.

Stockinet, layers of cotton web roll, and plaster or fiberglass are applied to make a well-molded body cast. Extending the stockinet and web roll over the edges secured to the outside of the cast with staples helps to keep the edges of the cast well padded. This also eliminates the time and need for pedaling the cast with adhesive tape.

Creating an adequate opening in the abdominal area of the cast makes a difference in comfort, tolerance, respirations, digestion, and keeping the skin clean. As children are predominately "abdominal breathers," having an opening in this area is important. It is not uncommon for the abdomen to protrude from the cast, especially after eating.

Common concerns of parents include keeping their child clean and preventing the cast from smelling and itching. Although keeping the cast clean and dry can be a challenge for parents of those children who are not toilet trained, it is possible. Each concern can be addressed by teaching proper skin care.

Moisten a long, narrow cloth with rubbing alcohol. Alcohol is used instead of soap because it cleans, toughens the skin, dries quickly, and does not leave any residue on the skin. Thread the cloth under the cast from the top to the abdominal opening or from the abdominal opening to the bottom of the cast. Move the cloth back and forth over the skin. If the child lies on their abdomen, it creates extra space to clean the skin on the back. Itching under the casts is caused by dead, flaking skin and moisture. Cleaning the skin on a regular basis with rubbing alcohol can prevent itching, as it removes the dead skin. Lotions are not applied because they tend to soften the skin. Loose powders are avoided as they can cake and cause a rash under the cast. Some centers encourage wearing a special T-shirt under the cast.

One of the most important aspects of cast care is preventing the cast from becoming wet. The inside of the cast has layers of cotton web roll for padding. If the cotton becomes wet, it is difficult to dry; the child's skin can break down, develop a rash, or become macerated.

Padding the bottom of the cast with disposable underpads along with tucking the diaper inside the cast goes a long way to prevent wetting and soiling of the cast:

- Tuck a 4-in.-wide × 12-in.-long disposable underpad strip three quarters of the way inside the bottom opening of the cast, between the child's skin and the cast, with the absorbent side next to the skin.
- Fold the remaining quarter of the disposable underpad over the outside of the cast.
- Repeat these steps all the way around the opening of the cast.
- Tape the edge of the disposable underpad to the outside of the cast.
- Change the disposable underpad strips any time they become wet or soiled.

Changing the diaper frequently and placing a sanitary pad in the diaper to absorb the urine also is helpful.

The skin around the cast is observed for redness, irritation, discoloration, and puffiness, which indicates the need for cast adjustment. Unusual odor, burning, or discomfort needs to be investigated. If the cast becomes wet, dry the inside using a hair dryer set on low or cool. If the cast has a strong odor, rub the outside of the cast with deodorant powder or baking soda to help absorb the odor.

There are several ways to shampoo the child's hair:

- Place the child on the kitchen counter, with their head over the sink.
- The child may lean over the bathtub and use a spray hose.
- The child may lie across a bed with the head hanging over the side of the bed.

Arrange a large sheet of plastic (a trash bag cut open) under the head to form a trough for the water to run into a tall wastepaper basket next to the bed. Cover the cast with plastic to prevent it from becoming wet while washing the child's hair.

Clothing tips are the same as for brace use. It is important to note that the child should always wear a T-shirt over the cast to cover the opening at the top of the cast. This will help prevent food or toys from falling inside the cast by accident.

54.4 Operative Care

Traditional posterior spinal fusion (PSF) and segmental spinal instrumentation (SSI) can pose problems when performed on the growing spine (and indirectly, the thorax). SSI with PSF may correct the curve, but in very young children, it

will also retard spinal and lung growth. In some, serious pulmonary compromise may result.

The concept of growing rods is not a new one. Harrington [5], the developer of Harrington rod instrumentation (HRI) for the treatment of scoliosis caused by the polio epidemic, recognized the risks inherent in fusing the young pediatric spine. Early attempts of using spinal rods without fusion proved difficult, demonstrating a high rate of complication.

With the evolution of better spinal implants with anchor designs, which are more capable of maintaining spinal fixation than their predecessors, traditional growing rod (TGR) spinal constructs are being used around the globe.

Originally developed as a single-rod system, growing rods are most commonly being used as a dual (two)-rod system. There are advantages of using the dual-rod over single-rod systems, including better maintenance of the curve and better spinal growth [6]. For this reason, the dual growing rod technique is described in this chapter.

The magnetic controlled growing rod (MCGR) is another approach used to surgically control large curves in young patients with early-onset scoliosis. The children will have the spinal implants including magnetic growing rods placed in the operating room. The initial surgical placement of MCGR and the care after are the same as TGR. These rods are then magnetically controlled with an external remote control device and lengthened in the office every 1–3 months or longer, depending on each child's unique circumstances, without the need for general anesthesia or surgery.

Chest wall deformities can also cause scoliosis and TIS. For rib distraction-based procedures, VEPTR will be described as the representative technique for treatments directed specifically to chest wall-related deformities.

The operative techniques for both growing rods and VEPTR are beyond the scope of this chapter (see Chaps. 38, 39, and 47).

54.5 Complications

The treatment of EOS, both nonoperative and operative, carries risks. Cast and bracing can cause some skin issues but rarely stops the casting procedure. The discontinuation of nonoperative treatment is a result of curve progression or restriction on the thorax during critical growth.

Likewise, surgical treatments such as growing rods, VEPTR, and other growth-friendly treatments that are intended to control the deformity while allowing growth can also have surgical or medical complications. The complication rate associated with the treatment of EOS is a function of the long duration of treatment and the number of procedures required during the treatment period. It is possible for one child to have multiple problems during the treatment period over many years [7]. Implant problems can be minor or major; wound problems, such as deep wound infection, may require long-term antibiotic treatment to avoid removal of the implants. To prevent surgical site infections (SSI), the OCHSPS National Children's Network developed and utilized preventative SSI protocols. Well-informed nurses can help surgeons to prevent and provide early detection of potential complications.

54.6 Preoperative Teaching

54.6.1 TGR and MCGR

Traditional growing rods and magnetically controlled growing rod treatments have been recently compared by Akbarnia et al. [8]. This comparison should be considered when educating the family regarding all treatment options.

The surgeon and his/her liaison should attempt to initiate education regarding treatment options with the family well before preoperative planning, if possible. This will help build trust between the surgical and patient/family team members. A close rapport between the medical team and the family is the goal. This will be helpful to all over time as care progresses and especially if a complication should develop.

Education should be ongoing throughout the treatment plan but is critical prior to the first surgery. Written material, if available, should be provided to the family as well to assist in the retention of the complex information provided.

The goals of treatment need to be emphasized throughout the treatment plan. For the growing rod treatment, curve control and allowance of spinal and lung growth over time are the main objectives. Details of the initial surgery include the implant components; what they are; their function initially and over time; the need for temporary external immobilization (surgeon's choice); what to expect before, during, and after the surgery; the length of the surgery (approximately 5 h for the initial TGR or MCGR surgery and 30–60 min for a TGR lengthening); the length of the hospital stay (4–5 days for the initial surgery and up to 23 h for a TGR lengthening); or lengthening in the office every 1–3 months for MCGR. Home care instructions, benefits, risks, and anticipated outcomes of the initial and all treatment procedures need to be communicated at this time. Attention to any unique patient/family needs should also be addressed and the family assured that its needs are being considered. Any necessary medical clearance requirements from other specialists should be obtained and discussed with the family as well. If the family requires any other special services, financial or otherwise, it is best to set this up prior to the first surgery.

Discussion regarding ongoing care is equally important, including the need for frequent lengthenings until the child is no longer benefiting from the treatment, or otherwise must be stopped for medical reasons. If a final fusion is likely needed at the end of the treatment period, this must also be included in the preoperative education with the family. Knowledge of the complete treatment plan is critical for trust between the surgeon and the family. Complication risks, especially those that are anticipated such as rod fracture, need to be explained to the family and, depending on the circumstances, will be addressed during a planned or unplanned surgery. The surgeon needs to communicate that it is likely for a complication to occur at some point during the treatment and rapid communication from the family to the surgeon followed by complete evaluation and explanation of the problem and the anticipated treatment from the surgeon to the family is very important.

If the patient/family is being asked to participate in a clinical study, this is the time to present the details of the study to the family, including all required HIPAA and IRB consent documents, so the family will feel at ease about the nature of the study and will not feel they are being pressured to participate. All educational and study consents provided to the family for signature should be given to the family, in copy form, to keep as part of their educational records.

Lastly, having one person on the team as the liaison between the surgeon and family is critical to promote trust, rapport, and communication as the child's treatment progresses over time.

54.6.2 VEPTR

Campbell and Smith developed the VEPTR [9]. It was designed to treat children with congenital scoliosis with or without fused or absent ribs. The chest wall deformity due to anomalies of the ribs causes thoracic insufficiency, which leads to lung failure. There are a variety of disorders that cause thoracic insufficiency including Jeune's syndrome, VATER syndrome, and more. Surgery includes treating the congenital scoliosis and addressing the chest wall deformity.

The population of children that this procedure is designed for includes patients who may be very ill. Some children are ventilator dependent and may have a tracheostomy as well as have feeding problems. The procedure, which includes cutting the ribs and placing rods for future expansion, is difficult and requires specialized training by the physician. Children undergoing VEPTR will have multiple hospitalizations.

Presented with the potential complexity of the orthopedic intervention in the treatment of these children, the nurse becomes acutely aware of the importance of her first-line observations. Management of the patient at any phase of treatment demands knowledge and skills so that complications are reduced and therapeutic effect can be achieved.

From the moment of introduction to the family regarding hospitalization, the nursing approach

determines the degree of patient and family participation, understanding, and cooperation.

Reduction of fear through a well-thought-out orientation can ease the patient and family through what otherwise could be a very traumatic experience. Physiologic responses have a direct relationship to compliance. Stressful experiences do not elicit patient or family compliance.

A surgical plan of care that introduces sophisticated mechanical devices and equipment requires a nursing practitioner, or similar liaison, who is skilled in the intricacies of observation, maintenance of equipment, and problem-solving. Corrective action must be superseded by correct application of principles. Problems should be solved by appropriate and timely intervention.

Nursing must emerge as a body of knowledge that supplements assessment, develops a standard of care outlining the individuals needs with the task at hand, and continually seeks to reassess the consumer's responses and anticipate outcomes.

The VEPTR patient population will have repeated procedures over the course of their lifetime. Planned actions need to be established, assuring that the surgical corrections and hospitalization are both seen by the patient and family as growth experiences. The treatment will span years. Sacrifice and cooperation will be needed by all members of the family unit. The nurse becomes the pivotal part of the team by helping the family and patient bridge the gap between knowledge and inexperience.

The preoperative evaluation and conference become the vehicles to bridge the knowledge deficit and secure trust in the team and anticipate outcomes in relationship to understanding, acceptance, and follow-up.

54.7 Preoperative Planning (All Techniques): See Table 54.1

54.7.1 Preoperative Testing: Initial Surgery

Thorough preoperative planning, teaching, and testing are important for surgery and recovery and to reduce the risk of complications.

Table 54.1 Preoperative planning

	Placement of growing rods/VEPTR	Lengthening TGR/VEPTR
Length of stay	4–7 days	[a]ASU – <23 h
Length of surgery	4–5 h	30–60 min
Pre-op labs	CBC, basic chemistry, nutrition panel, T and S, MRSA screen, UA, urine C and S (neuromuscular patients)	MRSA screen
Pre-op tests	PA and lat spine, side bending, traction; CT of the thorax, MRI entire spine, PFT, EKG	PA and lat spine
Tubes placed	2 IVs, Foley catheter, ET tube, central or arterial line, Hemovac, chest tube (VEPTR)	1 IV, ET tube
Pain meds post-op	Dilaudid/MSO4, OnQ, Ativan/Valium	Marcaine in surgery, oxycodone, Tylenol, Zofran for nausea
Return to school	2–4 weeks	3–7 days

[a]Ambulatory surgical unit

Preoperative testing can be completed 1–3 weeks before surgery, in most cases.

Preoperative testing usually includes lab work: complete blood count (CBC), type and screen (T and S), and methicillin-resistant *Staphylococcus aureus* (MRSA) screen. Other labs including basic chemistry, coagulation, nutritional panel, urinalysis (UA), and urine culture and sensitivity (C and S) are usually driven by the patient's disease process and need.

Radiological examinations include posterior-anterior (PA) and lateral views of the entire spine (AP sitting if the patient is unable to stand), supine right and left maximum bending views, and a traction radiograph before or after the patient is anesthetized. Magnetic resonance imaging (MRI) of the entire spine should be obtained (if it was not completed prior) to rule out intraspinal abnormalities such as tethered

cord, congenital components, Arnold-Chiari malformation, or syrinx [10]. The MRI may need to be obtained under sedation or general anesthesia as many of the patients are very young and/or unable to cooperate or hold still for extended period of time. Computed tomography (CT) scan of the thorax assists in assessing for thoracic insufficiency [11].

Consultations and tests from other medical services such as cardiology or pulmonary may be necessary depending on the child's comorbidities. Surgery may need to be postponed if the patient is not medically cleared for surgery until the cause is determined and treated.

To prevent surgical site infections (SSI), the OCHSPS National Children's Network developed and utilized a preventative SSI protocol. This consists of MRSA screen (and treatment with mupirocin if warranted), preoperative bathing at home the night before surgery with Dial soap or chlorhexidine, use of chlorhexidine wipes in the pre-op area, antibiotic selection and timing in the operating room with re-dosing schedule, and use of ChloraPrep to prepare the surgical site.

54.7.2 Preoperative Testing: Lengthening Procedures

Most cases are usually considered outpatient; therefore, no lab work except for MRSA nasal screen is necessary; however, this will be up to the individual surgeon's discretion based on comorbidity issues.

54.8 Perioperative Care

54.8.1 Growing Rods or VEPTR: Initial and Complex Revision Surgery

Once the child is asleep, an endotracheal tube, two large-bore intravenous lines, an arterial central line, and a Foley catheter are placed. After the surgery is completed, a Hemovac, Jackson-Pratt, or chest tube may be placed.

Patients having growing rods placed are positioned prone, usually on a Jackson table. A folded blanket with a gel pad under the thighs keeps the knees free. Two to three blankets with a gel pad are placed under the lower legs to keep toes free and knees flexed. Arms are abducted and the elbows are flexed 90°. Padding or a folded blanket placed under each arm protects the elbows and arms.

Patients having VEPTRs inserted are positioned in the lateral decubitus position with the thoracic area prepped and draped free.

After the initial placement of the growing rods or VEPTR, the orthotist can mold the patient for a TLSO in the operating room while the child is under anesthesia. This allows time for the brace to be completed by the time the patient is ready for discharge.

The nursing care before and after the initial insertion of growing rod(s) or VEPTR is very similar to the nursing care after a posterior spinal fusion. The patient may recover in the pediatric intensive care unit (PICU) or on a patient division, depending on the comorbidity of the patient or at the discretion of the surgeon.

54.8.2 Growing Rods, VEPTR: Lengthening Procedures

Surgical lengthening of the TGR or VEPTR is completed approximately every 6 months. The perioperative care is simpler than the initial placement. Since the procedure is short and there is usually very little blood loss, preoperative lab work or other special tests are usually not necessary except for the MRSA nasal screen. The surgical site infection protocol is also utilized. Only one IV is required along with an endotracheal tube. A Foley catheter is not needed. Positioning in the operating room is the same as with the placement. Injection of ¼–½% Marcaine in the surgical incision after closure makes a significant difference with pain management. Occasionally, one dose of morphine may be needed in the post-anesthesia care unit (PACU). Oxycodone and Tylenol are started at home the evening after surgery.

Patients are usually discharged home from the PACU. Patients who have a tracheostomy and ventilator are observed in the PACU for respiratory issues immediately following surgery. If none develop, they are also discharged from the PACU. If extensive revision of the implants is needed, observation (23 h admit) would be indicated for pain management. Another reason a patient might need to stay overnight or be observed would be related to nausea and vomiting. Administering Zofran during surgery often prevents this. Delaying clear liquids for a few hours after surgery also helps.

54.9 Postoperative Care

54.9.1 Growing Rods, VEPTR: Initial Surgery

Postoperatively, the patient will be transferred to the intensive care unit or the patient care division depending on their preoperative level of health. The patient participates in a routine every 2 h, which consists of log rolling from side to back to side, isometric exercises, and incentive spirometer or similar respiratory care appropriate for the child's age and ability.

Children, especially those under the age of 5 years, may have a difficult time with pulmonary toilet postoperatively. It is especially difficult using an incentive spirometer. Having the child blow bubbles has the same effect and the child will embrace this treatment.

Depending on the patient's preoperative ability to ambulate, the patient will be sitting up 1 day after surgery and ambulating with the assistance of physical therapy 2–3 days post-op. The patient may have a brace for ambulation that can usually be removed at night when sleeping or for showering.

The patient may be NPO for up to 3 days waiting for the GI system to resume normal activity. Once the patient passes flatus, clear liquids are started and the diet is advanced as tolerated. IV hydration will be required until fluid intake is adequate. If a patient is at nutritional risk, hyperalimentation is advised.

It is common for patients to have decreased appetite for a week after discharge. As a result, temporary weight loss often occurs. Small frequent meals can help encourage adequate nutritional intake. Supplemental milk shakes made with Carnation Essentials and ice cream provides increased calories and nutrition.

The patient will be placed on a bowel regimen using MiraLax or Dulcolax suppository until bowel movement is achieved. The Foley catheter is used for 2–3 days post-op.

IV antibiotics are administered for 24–48 h. Standard pain management for patients having posterior spine surgery is indicated for patients having initial placement of the growing rods or VEPTR. The pain management team is involved with postoperative pain medication ranging from intrathecal Duramorph, morphine or Dilaudid IV, OnQ, or PCA for the older child. Ativan or Valium is beneficial to relieve muscular tension or spasms caused by straightening the spine. PO pain medications consisting of oxycodone and Tylenol are initiated once the patient tolerates clear liquids.

Patients are usually discharged 5–7 days postoperatively. Prior to discharge, the patient may take a shower. Tub bathing is usually permitted 7–10 days after discharge or when the skin incision is healed. Increasing activity with the brace on is encouraged. Ambulating up and down stairs is permitted. They may sleep in their regular bed when they go home. Patients often return to school 2 weeks post discharge. Patients should not use a backpack over 20 lbs. or 10 % of their body weight.

Parents are instructed to call if there is swelling, redness, drainage from their incision, or open areas along their child's incision, an increase in back pain, a fever over 101°F, or other symptoms that were not present previously. Calls related to questions and concerns are always encouraged.

A brace or cast may be prescribed after growing rods are first inserted. Skin care and clothing tips are the same whether the brace/cast is utilized, nonoperatively or after surgery. Body contact sports or activities in which one can be pushed, bumped, or jarred should be avoided.

54.9.2 TGR and VEPTR: Postlengthening Care

Patients may ambulate (or if nonambulatory, be up in a wheelchair) at home the first post-op night. Progression to normal activity is encouraged. Many patients return to school within 2–5 days after lengthening. Physical education class is permitted except for body contact sports.

The back dressing is removed 4 days after surgery. Some patients' skin may be sensitive to certain tape. Alternative tapes and dressings can be used.

Showering is permitted approximately 4–7 days after surgery, tub bathing approximately 1 week later, or when incision is healed.

The family is instructed to observe for possible wound infection, implant failure, and/or retractable pain. Parents are instructed to look for change in height or in shoulder level and whether the implant is prominent. Families have access to their nurse daily and can call with any concerns regarding post-op recovery.

54.9.3 Growing Rods: Post Final Fusion Care

The nursing care post final fusion is the same as posterior spinal fusion and similar in experience to the patient's initial surgery for implantation.

Caring for patients and parents having growing rod treatment can be challenging, yet rewarding. Nurses can make a significant difference in the patients' treatment, attitude, and team's problem-solving. As the treatment continues for a long time, the nurse becomes close to the patient and family, sharing its struggles and successes.

54.10 Future Treatment: Evolution from Knowledge

With improved techniques including less invasive means of allowing for controlled growth and curve maintenance, such as growth-guided techniques or MCGR which allow growth without repeated, frequent surgeries, it is believed that children with EOS may be treated earlier than before (40–50° curves at start of treatment vs. 80–90° curves). A question with regard to earlier treatment is: will a final fusion be necessary at the end of treatment? It is hypothesized that if early brace treatment for AIS can potentially prevent spinal fusion surgery, early growth-friendly procedures might also prevent the need for definitive spinal fusion for those EOS patients in the idiopathic scoliosis population or, possibly, all diagnosis groups.

So far, only retrospective study data points to effective curve maintenance and spinal growth over time. Only prospective, long-term analysis will determine if certain children treated with the growing rod technique and similar techniques may not require final fusion.

The GSSG, acknowledging that more needs to be learned about the care of children with EOS, hopes to address this and many other questions in the future.

Conclusions

The nursing care and treatment of patients with early-onset scoliosis is challenging for all involved. It requires long-term commitment by the family and health-care team. Nurses have an important role in improving the quality of care for this patient population. They can help surgeons to prevent and provide early detection of potential complications. They can also be instrumental with patient and family education to improve quality of care. To be a valuable member of the team caring for children with early-onset scoliosis, nurses should learn as much as possible about the care involved with nonoperative and operative treatment of these patients.

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Outcomes in Children with Early-Onset Scoliosis

55

Howard Y. Park, James G. Wright, and Michael G. Vitale

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Key Points

- In last two decades, there has been a rapid expansion of treatment options for children with early-onset scoliosis (EOS).
- The evidence base has not kept pace with the expansion of treatments, and there exists some relative uncertainty regarding optimum treatment options.
- Traditional radiographic measures inadequately reflect outcomes in the area of early-onset scoliosis.
- There is reasonable evidence that early fusion results in negative long-term pulmonary consequences in children with scoliosis.
- Measurement of pulmonary function may require a different set of measures than used in older populations.
- Health-related quality of life (QOL) is an important endpoint in children with early-onset scoliosis.
- We have witnessed an expansion in available nonfusion options for patients with early-onset scoliosis.
- Although significant obstacles with regard to high-level research exist, high-level research should be strived for.

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- All of these factors, including EOS patients' underlying disease, contribute to the variability in decision-making in the treatment of these patients.
- Research in this area should be prioritized and incentivized in order to drive innovation and provide optimal care to these patients.

55.1 Introduction

The term "orthopedics" comes from the Greek roots "ortho" and "pedics" meaning "straight child." Intuitively, a straight bone seems to imply a good outcome, and radiographic measures have been the historical standard in assessing surgical outcomes. However, in the last two decades, there has been a fundamental shift in thinking about how best to measure and describe outcomes in order to assess efficacy for the treatment of early-onset scoliosis (EOS). We now look at more broadly defined outcomes including functional status, health-related quality of life (QOL), and cost-effectiveness. We utilize more rigorous methods of research design including randomized trials, and we apply more sophisticated means of analysis of data in an attempt to decrease bias and better compare treatment options.

In the last two decades, there has been a rapid expansion in the treatment options for EOS. As a result, the treating physician now faces a complex palette of often overlapping and mutually exclusive choices, yet both experience and knowledge base appear insufficient to optimize decision-making. When reviewing the literature, the bulk of evidence in EOS is level 3 or 4 with no level 1 or 2 studies, forcing physicians to rely on retrospective experience, intuition, and consensus for the basis of treatment decisions [1]. As a result of this dearth of evidence, variation in decision-making in the area of EOS treatment is abundant. In a survey of members of the Children's Spine Foundation, Vitale et al. [2] demonstrated substantial intra-observer and inter-observer variability in decision-making where the indications for surgery were in large part agreed upon but the surgical technique and constructs were in contention. Furthermore, areas of equipoise within EOS surgery have been studied formally, which cited rib versus spine proximal fixation, treatment options at skeletal maturity following growing spinal instrumentation, and intervals of lengthening as three topics of highest uncertainty among a collection of pediatric spine surgeons [3] (Fig. 55.1).

The obstacles to producing high-level studies to answer the numerous questions regarding EOS treatment are multiple and substantial, not least of which are the difficulties in assessing outcomes in the EOS population. First, patient populations are small and heterogeneous, making it difficult to accrue sufficient numbers of patients and to compare treatments. Second, the pace of evolution of treatment options has been rapid, creating a moving target for patient assessment. Third, altering the natural history implies following a patient cohort longitudinally over time, and the long periods of follow-up that are necessary represent a major challenge to research in this area. Fourth, there has been little consensus about how best to measure pulmonary outcomes in this population. In fact, we are just learning how complex the relationships are between EOS, treatment, and pulmonary function [4, 5]. In summary, a primary goal of the treatment of EOS is to improve the natural history of the disease by permitting pulmonary maturity and, in turn, decreasing early mortality. The outcomes to assess pulmonary function and mortality require difficult pulmonary assessments with decades' long follow-up within a heterogeneous pediatric population.

Furthermore, new technologies need to be evaluated and approved by the FDA, but the regulatory burdens present significant obstacles. As discussed earlier, it is difficult to amass sufficient patient populations for study, and it is challenging to follow patients in the long term to meet the requirements of post-market surveillance imposed by the FDA. While industry plays a role in promoting the necessary research to obtain approval for new technologies in orthopedics

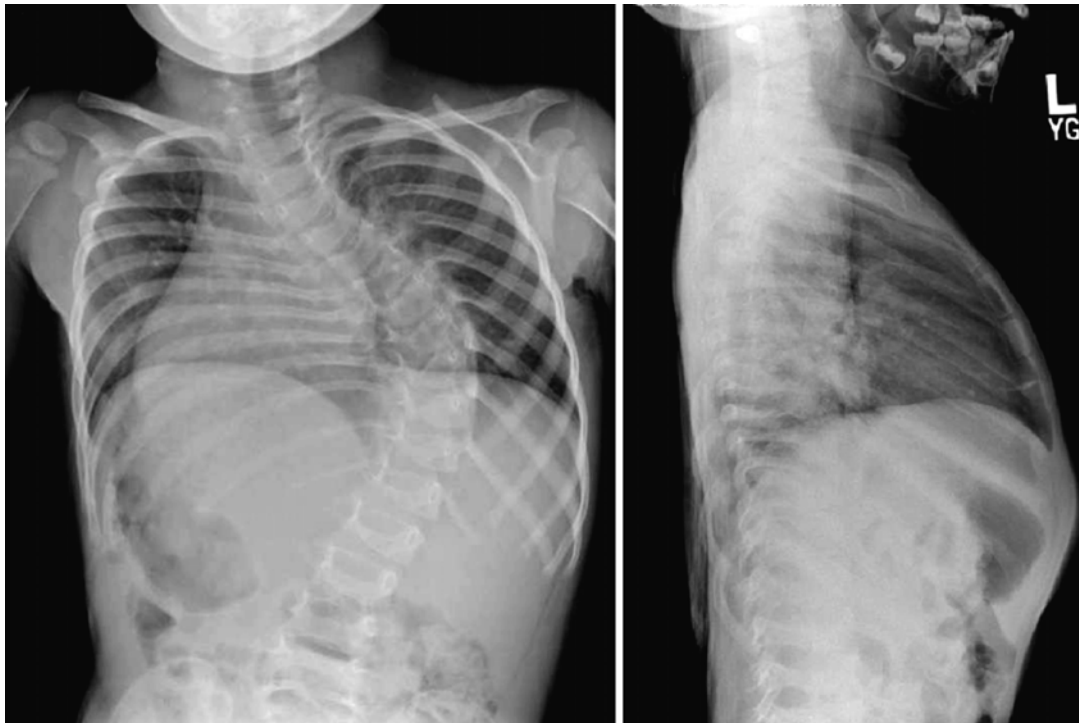

Fig. 55.1 A representative case highlighting difficulty and variability in decision-making in patients with early-onset scoliosis. Members of the Children's Spine Foundation exhibited significant inter- and intra-observer variability in treatment recommendations for this child, an 18-month-old Jehovah's witness with an undiagnosed mitochondrial disorder and progressive scoliosis

surgery, the small patient populations affected by early-onset scoliosis do not draw sufficient interest and may limit available funding.

All of these factors create a "perfect storm," making the design and conduct of high-quality, important clinical evaluative research in this area significantly challenging. Nonetheless, an increasing number of higher-level studies are being undertaken, and groups like the Children's Spine Foundation and Growing Spine Study Group are answering the call for higher-level evidence in EOS.

55.2 Natural History as a Framework for Outcomes

The ultimate goals of treatment for children with EOS are to alter the natural history in the short and long term, which inform researchers of pertinent outcomes. The onset of spinal deformity from birth to age eight can be deleterious to pulmonary maturation, the long-term consequences of which contribute to a fourfold mortality rate at middle age compared to patients with adolescent idiopathic scoliosis [6]. In addition, spinal deformity in the early childhood years can have significant quality-of-life (QOL) consequences for the child and caretaker [7, 8]. Therefore, the natural history can be utilized as a framework when assessing outcomes to quantify the effects of treatment. Radiographic measures and scoliometer readings to assess spinal deformity, pulmonary function testing and thoracic measurement on radiograph to assess pulmonary development, caregiver burden and functional status surveys to assess health-related quality of life, and mortality rates decades following treatment to assess survival are all outcomes of interest.

55.2.1 International Classification of Functioning as a Framework of Measurement

The immediate goals of treatment for children with EOS are to minimize physical disability including respiratory symptoms and relieve pain to improve appearance. The long-term aim of treatment is to prevent the onset of new symptoms, minimize future decline in function, and prevent premature death. While several useful frameworks have been proposed to organize outcomes, the International Classification of Functioning [9] has particular relevance to surgery.

The pathway (Fig. 55.2) begins with a disease, which in turn leads to impairment. Impairment is defined as an abnormality of structure or function. Impairments lead to disabilities, defined as the lack of ability to perform an activity in a manner considered normal. Finally, activity restrictions lead to handicap or role limitation, the inability to fulfill roles, which for children refer to family, friends, and school. In the ICF pathway, the disease could be congenital scoliosis leading to spinal, chest wall, and lung deformity. These impairments lead to physical and respiratory disability, limitations in school and play, and, possibly, early death. Such a pathway is particularly relevant to the area of early-onset scoliosis because our treatments are often directed at reducing spine and chest-wall deformity. If treatments reduce impairments, the hope is that, in turn, we will ameliorate activity restrictions, enhance role function, and prevent early death. However, the pathway is clearly not linear. Furthermore, children with EOS often have associated conditions that moderate the pathway, influence the success of treatment, and lead to other impairments with additional consequences. Nevertheless, this framework provides a way to categorize measures and draw attention to the important question of what treatment is best at reducing impairments, thereby positively affecting the consequences of disease.

55.3 Available Endpoints and Measures of Outcome

55.3.1 Radiographic Measures

The Cobb angle is a well-accepted primary endpoint in adolescent idiopathic scoliosis. However, Campbell has highlighted the complex three-dimensional relationships between EOS thoracic deformity and pulmonary function, which are poorly reflected in the unidimensional Cobb measurement [10–12]. Sagittal deformity on radiograph can provide a second dimension of the complex spinal deformity, and it is an often-utilized outcome in EOS studies. Campbell and others [10, 11] have described a variety of other radiographic measures including the space available for the lung, thoracic height, the spinal penetration index, and the posterior thorax symmetry ratio. Other potential radiographic measures include the interpedicular line ratio, decompensation, shoulder level, and vertebral rotation. Mehta [13] has brought attention to the importance of the position of the ribs in relation to the spine in the idiopathic infantile scoliosis population. A rib vertebral angle difference of 20° or

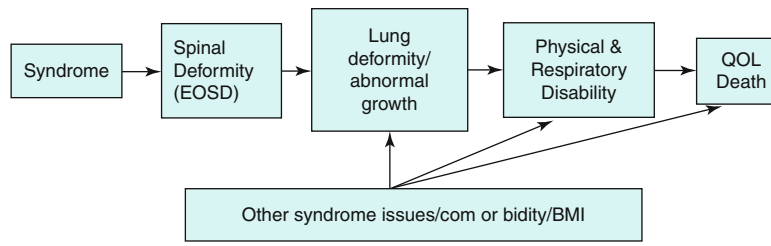

Fig. 55.2 Disease leads to impairment, then disability, and then loss of role function (Adapted from the international classification of functioning)

more strongly predicted progression in this group. However, even within the diagnostic category of "infantile idiopathic scoliosis," it is not clear that we are examining a single clinical entity. As Fernandes and Weinstein [14] questioned, "Is resolving scoliosis, as opposed to progressive scoliosis, a different entity with a different etiology, or does infantile idiopathic scoliosis have a full spectrum of presentations."

55.3.2 Pulmonary Function

While pulmonary function is of central importance in the EOS population, the measurement of pulmonary function in very young children requires special equipment and technique. Furthermore, it is not clear that traditional measures of pulmonary function such as vital capacity and expiratory volumes sufficiently describe the pathology relevant to the children with EOS. Redding [15] has shown significant alterations in lung function in children with congenital scoliosis and has hypothesized that scoliosis affects lung function by incursion into the hemithorax, as well as by reducing chest-wall compliance and excursion, producing asymmetry in lung size and function. However, this study of 39 patients with EOS did not demonstrate a correlation between Cobb angle and pulmonary measures. Computerized tomography with three-dimensional reconstructions provides the ability to estimate lung volumes and characterize the intrathoracic deformity [16]. Work by Adam [17] has shown strong correlations between lung volumes as measured by CT and pulmonary function tests. This study highlights the complex nature of spine, thoracic, and lung deformity, and it discusses the limitations of traditional pulmonary function tests that ignore the significant asymmetries in right versus left lung. However, concerns about the increased risk of malignancy associated with routine and perhaps repeated CT scans limit its use as a research tool [18]. Magnetic resonance imaging may have similar potential, though investigations in this area are still in their infancy.

55.3.3 Health-Related Quality of Life

It is clear that many patients with early-onset scoliosis have significant issues with a variety of aspects of health-related QOL, but measurement of QOL is fraught with difficulties in this population. Vitale et al. [7] have documented significant perturbations in health-related QOL as measured by the Child Health Questionnaire [19] in older patients who had undergone expansion thoracoplasty. However, generic measures of QOL have been developed and validated primarily in patients >5 years of age limiting the usefulness in this population. Children develop and mature, creating a moving target for functional assessment in this population. Finally, health issues in this heterogeneous population can be quite specific and may not be fully captured with available "off-the-shelf" measures of health status.

Therefore, collaborative efforts between the Children's Spine Foundation and Growing Spine Study Group have yielded the Early-Onset Scoliosis Questionnaire (EOSQ), which is a validated instrument in EOS patients. It is now being utilized in various studies as a novel and valid outcome for EOS patients.

55.3.4 What Do We Know About the Outcomes of Treatment?

Despite the considerable obstacles to meaningful clinical research in this area, research efforts have afforded clinicians some initial understanding of important issues in this area. Following is a brief summary of the available evidence pertinent to the field of early-onset scoliosis.

55.3.4.1 Negative Effects of Early Fusion

Several studies have now been published that collectively demonstrate untoward effects of early fusion in children with scoliosis. Karol et al. [20] examined pulmonary function tests of 28 patients who had undergone a spinal fusion prior to the age of 9 and demonstrated pulmonary function

testing with values of 50–60 % predicted when compared with norms. Patients at highest risk of restrictive disease were those who had a more extensive thoracic fusion, especially proximal thoracic fusion. Similar results have been reported by other authors including Vitale et al. [8], Goldberg et al. [21], and others. While none of these articles prospectively measured pulmonary function before and after fusion, collectively, this body of work provides substantial evidence that children with early-onset scoliosis who have been treated with traditional fusion techniques are at significant risk for considerable problems with pulmonary function. The onus is now on current researchers to demonstrate that new techniques result in an improvement from this dismal picture presented by natural history and early fusion.

55.3.4.2 Outcomes in Patients Treated with "Growing Rods"

In an attempt to avoid fusion at all costs, "growing rods" or expandable spinal implants have become the standard of care for children with early-onset scoliosis. In a multicenter study of 23 patients who underwent dual growing rod constructs for early-onset scoliosis, Akbarnia et al. [22] demonstrated significant improvements in Cobb angles and space available for the lung and spinal growth. Other studies corroborate these findings with a Cobb correction range of 29–63.7 % as a result of growing rod surgery. Greater growth and correction are seen in patients who undergo lengthening at interval of less than 6 months, and dual growth rods seem to deliver better radiographic results than single rod constructs [23]. There exist only a few studies that assess pulmonary function in growing rod constructs that utilize various outcome measures that have resulted in conflicting results in terms of pulmonary benefit. No studies report on quality of life.

A recent development within growing rod surgery is the magnetically controlled growing rod, which circumvents repetitive surgery for lengthenings. These externally controlled growing rods are investigational, but the initial Cobb correction and sagittal correction data suggest that these devices are similar in efficacy to their nonmagnetic counterparts, and patients experienced minimal pain and good function and were satisfied with the procedure [24–26].

55.3.4.3 VEPTR and Expansion Thoracoplasty in Children with Fused Ribs

As described by Campbell [27], expansion thoracoplasty implies surgical separation of rib fusion and placement of the VEPTR device. Campbell followed a cohort of children with scoliosis and rib fusion who underwent surgery at a mean age of 3.2 years and with mean follow-up of 5.7 years who underwent expansion thoracoplasty [10]. Significant improvements in radiographic measures such as the Cobb angle, space available for the lung, and thoracic spinal height were noted, though pulmonary function at follow-up continued to demonstrate significant perturbations from norms. Vital capacity at follow-up ranged from 44 % for those older than 2 years of age at the time of surgery to 58 % for those children younger than 2 years of age. In a subset of patients whose lung function was followed longitudinally, there was some improvement in vital capacity, but no significant improvement in percent predicted vital capacity. Of note, 52 complications were encountered in 22 patients through the course of treatment. Motoyama and collaborators [28] in Pittsburgh showed that PFT during and after expansion thoracoplasty kept up with growth, but did not increase in percent predicted. Emans et al. [29] studied 31 patients with fused ribs and thoracic insufficiency syndrome prospectively. Significant spinal growth and improved lung volumes as measured by CT and pulmonary function tests were noted in this cohort, supporting the role of early nonfusion surgery in this patient cohort.

Two studies have explicitly looked at quality of life. Significant differences were found between healthy patients and preoperative VEPTR patients in physical function domains, and significant differences in time burden and emotional domains were found between parents of healthy children and parents of VEPTR surgery patients. The second study found

differences in adaptability and resiliency domains when calculated by total number of surgeries. Various trends were found in differing domains when analyzed by the number of instances of VEPTR surgery (implantation and lengthenings), age at initial implantation, and age at initial assessment [30].

55.3.4.4 Growth Modulation: Vertebral Stapling and the Shilla Technique

In an effort to improve upon the outcomes of children with scoliosis, clinicians have developed strategies to attempt to modulate or control growth. Betz et al. have pioneered the use of anterior vertebral staples [31, 32]. While this technique has great theoretical appeal and continues to gain popularity, clinical research that is necessary to document that such a technique positively impacts natural history has been sparse. McCarthy has developed the Shilla technique, a novel technique employing an apical fusion with expansion of the rest of the instrumented spine using a specially designed setscrew. The Shilla technique's advantage is the avoidance of repetitive surgery for lengthening. A recent study of 10 patients with 2-year follow-up shows adequate deformity correction, acceptable complications rate, and the ability to guide growth that with other growth systems would have necessitated 49 operations for lengthenings [33].

55.4 Opportunities for Future Progress

Despite the challenges, there are a number of opportunities to better organize processes that will facilitate clinical research and innovation in this area. In a review of the literature currently underway, endpoints of Cobb angle, kyphosis correction, pulmonary function, quality of life, and complications were reported in various ways prohibiting pooled statistics or meta-analyses. A first step towards uniform outcome reporting would involve some consensus among experts in this area about what should constitute a minimum common set of outcomes and a protocol for reporting them.

In order to efficiently conduct clinical research in this area, multicenter studies will be critical. Existing today are the Growing Spine Study Group and the Children's Spine Study Group, both of which contain members with significant interest and experience in the area of EOS. These groups serve to centralize multicenter efforts in this area, but opportunities to further capitalize on this important infrastructure exist. Such groups should require a minimal common dataset for all studies that are performed by the members. One example of this is a study that is sponsored by the Scoliosis Research Society, and it takes advantage of both study groups to explore the differences between rib versus spine proximal fixation in EOS, a topic of significant equipoise [3]. This study represents one of the first high-level prospective comparative studies of EOS to date, and it will analyze outcomes of Cobb angle, kyphosis, quality of life through the use of the Early-Onset Scoliosis Questionnaire (EOSQ), and complications.

However, despite the efforts of these two study groups, many patients with EOS are not part of these research efforts, which is a lost opportunity for research. In contrast, the majority of children who receive chemotherapy for childhood cancers are captured as part of a national protocol, which has facilitated meaningful clinical research in this area. Perhaps use of new investigational techniques and implants on "index" patients with EOS should be contingent on contribution of results to a multicenter registry.

Children with EOS should be considered an "orphan population." The small markets and regulatory burdens involved in obtaining approval of new devices in this field hinder innovation. Society must therefore create incentives for innovation in this area. Our national societies and subspecialty organizations can help by funding requests for proposals for research in this area.

These issues have not gone unnoticed by the federal authorities. The Pediatric Medical Device Safety and Improvement Act of 2007 provides several incentives to innovation, including allowing manufacturers to profit from devices approved under the Humanitarian Device Exemption and

providing funding through consortia to support innovation in the area of pediatric devices. Recently, we have seen progress including the FDA approval of growing rods in patients with EOS, pedicle screw use for adolescent idiopathic scoliosis, and the magnetic implantable growing rod. Nonetheless, much remains to be done.

Conclusion

The treatment options for early-onset scoliosis are varied and numerous with relatively few studies corroborating the conceptual promise of these treatment modalities. The rapid expansion of treatment options has not been accompanied by rapid clinical study necessary to rigorously compare and contrast various treatment options. In addition, outcomes in EOS are challenging to establish and collect, and the decades-long natural history of EOS is difficult to study. Therefore, rigorous study through the two established study groups for EOS as well as others is required to optimize decision-making for children with EOS, minimize risks and complications, and maximize benefits. However, given the current state of EOS study, treatment in the absence of rigorous evidence may nonetheless be justified based on the consensus expert opinion that currently available treatment strategies are likely to provide superior outcomes in contrast to the rather dismal natural history.

In this regard, an analogy can be made to the days of solid organ transplantation. Patients at the beginning of the era of heart transplantation had few viable options, yet techniques of heart transplantation and management of posttransplantation were rapidly evolving and not well defined. In the course of time, complications decreased dramatically and outcomes improved. Indications became more uniform based on acquired data of outcomes in various populations.

The same challenge confronts those focused on improving outcomes of children with early-onset scoliosis. An improved understanding of patient outcomes in this area is a prerequisite to the timely evolution of care of these children.

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Part VIII

Current and Future Considerations

Regulatory Policies Regarding Pediatric Spinal Devices

56

Vincent J. Devlin

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Key Points

- The introduction of transformative medical devices often presents new scientific and regulatory challenges.
- Collaboration between the medical device industry, surgeons, and regulatory authorities is integral to the introduction of innovative pediatric orthopedic medical devices.
- Regulatory processes for medical devices vary worldwide and are subject to control by the regulatory body of the specific country or region.

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- In the United States, the Food and Drug Administration (FDA) is committed to assuring that patients have timely access to important new pediatric medical devices safely and based on sound science.

56.1 Introduction

Devices for treatment of pediatric spinal disorders have been identified as a critical unmet need in the United States as well as globally. Of the estimated 1.24 million patients who utilized health-care resources to receive care for health problems associated with spinal deformity in the United States during 2007, 44 % were under the age of 18 years [1]. As highlighted in its *Survey on Pediatric Device Development*, the American Academy of Orthopaedic Surgeons (AAOS) identified self-expanding spinal deformity control devices for the growing child and minimally invasive growth modulation instrumentation for spinal deformity as specific unmet device needs in the pediatric population [2]. Introduction of new medical device technologies directed toward care of pediatric patients with disorders affecting the spine and thorax has the potential to advance treatment of these challenging clinical conditions. The regulatory process influences all phases of device development and subsequent device use. Knowledge regarding regulatory processes is required to successfully navigate the available pathways to market for innovative pediatric orthopedic medical devices in order to provide timely access to potentially life-enhancing and lifesaving orthopedic devices.

56.2 Pediatric Populations for Medical Devices

The age limits of pediatric populations have been variably defined in the medical literature. The American Academy of Pediatrics defined that the purview of pediatrics begins with the fetus and continues through 21 years of age [3].

Regarding pediatric medical devices in the United States, Section 520(m)(6)(E)(i) of the Federal Food, Drug, and Cosmetic Act (FD&C Act) defines pediatric device patients as persons aged 21 years or younger at the time of their diagnosis or treatment (i.e., from birth through the 21st year of life up to but not including the 22nd birthday). Pediatric subpopulations are defined to be neonates, infants, children, and adolescents (Table 56.1). The Food and Drug Administration (FDA) recognizes, however, that the defined pediatric subpopulations are somewhat arbitrary and that, in fact, the subject's weight, body size, physiological development, neurological development, and neuromuscular coordination may often be more appropriate indicators than chronological age [4]. In contrast, the AAOS considers that the transition to adulthood with regard to orthopedic devices is defined by skeletal maturity, which is attained at approximately ages 14–16 years for females and ages 16–18 years for males [5]. Recently, professional spine societies have defined the early-onset scoliosis population including patients with spine deformity that is present before 10 years of age and recognize various diagnostic categories including idiopathic, congenital, thoracogenic, neuromuscular, and syndromic types.

Table 56.1 Pediatric subpopulations for medical devices and related ages as defined by the FDA

Pediatric subgroup	FDA guidance on approximate age range for this subpopulation
Newborn (neonate)	From birth to 1 month of age
Infant	Greater than 1 month to 2 years of age
Child	Greater than 2–12 years of age
Adolescent	Greater than 12–21 years of age[a]

This table presents FDA guidance on the approximate age ranges for four main pediatric subpopulations as reported in the FDA document Guidance for Industry and FDA Staff: Premarket Assessment of Pediatric Medical Devices (2004, 2014)

[a]For internal review purposes, the FDA has identified another subpopulation, transitional adolescent, to include those aged 18–21 years

56.3 Challenges Related to Advancing Pediatric Spinal Devices to Market

Challenges encountered in the development of pediatric spinal devices relate to economic factors, target population considerations, liability concerns, regulatory processes, and clinical trial design (Table 56.2).

56.3.1 Economic Factors

The economic incentives for development of medical devices for pediatric populations differ fundamentally from those in adult populations. In general, device developers consider pediatric medical devices as small volume product categories due to small target population sizes and the existence of substantial patient heterogeneity within these populations. Such factors limit future profitability and create financial disincentives to direct resources toward development of pediatric devices. Additional challenges include difficulties related to obtaining insurance reimbursement and lack of billing codes.

Table 56.2 Challenges related to development of pediatric spinal devices

Potential market for pediatric devices smaller than for adult devices
Single-device design and device size may be inadequate across all pediatric subpopulations due to anatomic and physiologic differences among pediatric subpopulations
Pediatric devices are associated with additional liability concerns for clinicians and device manufacturers compared to adult devices
Lack of pediatric device trial infrastructure results in increased burdens for large-scale studies
Difficulty in recruitment of adequate numbers of subjects for pediatric clinical device trials
Challenges regarding adequate study design for pediatric clinical device trials compared to adults (i.e., treatment outcome measures are difficult to define; suitable control population may not exist)

56.3.2 Target Population-Related Considerations

The pediatric population consists of different subpopulations which are accompanied by differences in size, growth rate, metabolism, and activity levels. Consequently, a manufacturer may need to develop and test a different device for each subpopulation, even though the disease or condition treated is the same. For example, a device which is appropriate for an infant may not be appropriate for use in an adolescent with the same disease or condition. It is a common experience that devices designed for adults are not adequate for use in children and require redesign. An additional consideration relates to limited knowledge regarding the etiology of specific pediatric spinal deformities such as idiopathic scoliosis, which occurs for unknown reasons [6].

56.3.3 Liability Concerns

Device manufacturers have expressed reluctance to pursue development of devices in pediatric populations due to concerns regarding potential future product liability. In the United States, due to the limited number of devices specifically cleared or approved for pediatric use, clinicians are likely to use medical devices for indications not contained in the approved labeling for a specific device. Such use is referred to as "off-label use," practice of medicine, or physician-directed use. This practice is considered to be associated with higher risks to patients as there is less information available to physicians and patients regarding the safety and efficacy of such use. Although the FDA regulates the indications and intended uses of medical devices, it does not regulate the practice of medicine. Physicians may legally use devices for an indication not in the approved labeling if such use represents good medical practice and is in the best interests of the patient according to the physician's best knowledge and judgment. Physicians have the responsibility to be well informed about the product, to base product use on firm scientific rationale and

on sound medical evidence, and to maintain records regarding the product use and its effects [7, 8]. In contrast, medical device manufacturers are prohibited by law from promoting off-label use. The FDA has outlined specific practices which manufacturers should employ if they choose to disseminate scientific and medical journal articles, scientific and medical reference texts, and clinical practice guidelines (CPGs) that include information on unapproved new uses of approved, cleared, or 510(k) exempt products [9, 10]. Physicians are responsible to be informed regarding regulations and guidances concerning off-label use and should be aware of the potential for their specific actions to be perceived as promoting off-label use for a company, especially if the clinician enters into any type of consulting arrangement with the manufacturer of a specific product.

56.3.4 Regulatory Process Related to Pediatric Medical Devices

The regulatory requirements for medical devices in the United States affect all phases of the product development cycle.

The regulatory process currently used in evaluating pediatric devices is not different than for adult devices [11]. The regulatory process implements the legislative framework established by Congress and includes device clearance and approval pathways that do not differ on the basis of whether a device is intended for use in a pediatric population or an adult population. Regulatory challenges related to pediatric device development include conservative criteria for pediatric device approval, lack of device-specific FDA guidance documents, limitations related to the number of devices eligible for the Humanitarian Device Exemption (HDE) pathway, restrictions on the use of off-label data in clinical studies, and challenges associated with the Institutional Review Board (IRB) oversight of pediatric clinical trials [12, 13]. As the FDA considers pediatric subjects as a vulnerable population, additional safeguards for pediatric patients involved in clinical trials are required for sponsors, investigators, and IRBs (21 CFR, Part 50, Subpart D).

56.3.5 Issues Related to Design of Clinical Trials for Pediatric Devices

Pediatric devices present unique challenges related to the design of clinical trials. Classic randomized double-blind comparative studies are often not feasible or ethical in small pediatric populations with limited alternative treatment options. Innovative trial designs and statistical methods are often necessary. Additional challenges arise due to mandated safeguards for pediatric patients involved in clinical trials (21 CFR, Part 50, Subpart D).

56.4 Overview of Medical Device Regulation in the United States

56.4.1 Responsible Regulatory Agency

In the United States, the FDA is the scientific, regulatory, and public health agency responsible for protecting and promoting public health through the regulation and supervision of a wide range of products including most food products (excluding meat and poultry); prescription and over-the-counter drugs; medical devices; radiation-emitting products for consumer, medical, and occupational use; vaccines, blood, and biologic agents; veterinary products; cosmetics; and tobacco products.

56.4.2 Legislative Overview

The FDA is an agency of the US Department of Health and Human Services, a part of the executive branch of the federal government of the United States. The FDA is responsible for implementing the laws which are written and enacted by the legislative branch of government (the US

Congress). The legal authority of the FDA to regulate medical products, including medical devices, stems from the FD&C Act which contains regulatory requirements that define the FDA's level of control over medical devices. To fulfill the provisions of the FD&C Act that apply to medical devices, the FDA develops, publishes, and implements regulations which are published in the Federal Register (FR) and codified in the Code of Federal Regulations (CFR). Most of the FDA's medical device and radiation-emitting product regulations are in Title 21 CFR Parts 800–1299. The regulations most relevant to spinal devices are located in Title 21 (Food and Drugs), Chap. 1 (Food and Drug Administration), Subchapter H (Medical Devices), Part 888 (Orthopedic devices).

The regulatory authority of the FDA was established with passage of the Federal Food and Drug Act (1906) which granted authority to prohibit the sale or distribution of adulterated food or drug products in the United States. The FD&C Act of 1938 was a precedent for regulation of medical devices, but its scope was limited to regulation of marketed devices considered to be unsatisfactory, unsafe, or misbranded. Premarket regulation of medical devices was formally initiated in 1976 when the Medical Device Amendments to the FD&C Act expanded the FDA's authority to include regulation of devices. Under the Medical Device Amendments, the FDA established panels of experts in individual specialties to develop definitions and classifications that would apply to all commercially available medical devices. The Medical Device Amendments defined a medical device as a product intended for diagnosis, cure, mitigation, treatment, or prevention of a disease or condition, or an entity intended to affect the structure or function of the body, which does not achieve its primary intended use through chemical action or metabolism. The mandate of the 1976 Medical Device Amendments provided the FDA with authority to ensure the safety and effectiveness of medical devices prior to marketing, established a classification system for medical devices, and outlined pathways to market for medical devices. Modifications and updates to regulations have been made in response to subsequent advances in medicine and technology. In 1990, the Safe Medical Devices Act (SMDA) required institutions to report any adverse events involving a medical device to the manufacturer and, under certain situations, to the FDA. In addition, the SMDA initiated the Humanitarian Use Device/Humanitarian Device Exemption (HUD/HDE) program. The Medical Device User Fee and Modernization Act of 2002 (MDUFMA) provided the FDA authority to collect user fees from sponsors to offset the expense of premarket review of devices in exchange for adherence to established timelines for agency review of specific types of premarket device applications. The Food and Drug Administration Amendments Act of 2007 (FDAAA) focused on pediatric safeguards, research, and innovation, in addition to strengthening the transparency of clinical study results. The Food and Drug Safety and Innovation Act (FDASIA) of 2012 included specific provisions to improve the safety and effectiveness of pediatric medical devices, drugs, and biological products.

56.4.3 Organizational Structure Relevant to Spinal Devices

The FDA is organized into multiple centers and offices. The Center for Devices and Radiological Health (CDRH) is responsible for review of medical devices through its eight offices. Within the CDRH, the Office of Device Evaluation guides the premarket regulation of orthopedic medical devices, while in vitro diagnostics and radiologic medical devices are evaluated in the Office of In Vitro Diagnostics and Radiological Health. The Office of Device Evaluation consists of seven divisions, each of which focuses on a different medical specialty area, and each division consists of multiple branches. Most devices used in the treatment of pediatric spinal disorders are under the authority of the Division of Orthopedic Devices. Responsibility for regulation of spinal devices is shared between the Posterior Spine Devices Branch (e.g., pedicle screw spinal systems) and the Anterior Spine Devices Branch (e.g., intervertebral body fusion cages, anterior spinal plate systems). Other products used in the

spine (e.g., bone morphogenetic proteins, bone void fillers, bone cements) are reviewed by the Restorative and Repair Devices Branch. Additional offices within the CDRH other than the Office of Device Evaluation also participate in the regulation of orthopedic spinal devices. For example, the Office of Surveillance and Biometrics monitors adverse events and possesses the authority to mandate postmarket studies. Additionally, the Office of Compliance is responsible for conduct of inspections and audits to ensure adherence to quality system regulations and good manufacturing practices.

56.4.4 Classification of Medical Devices

Medical devices are classified by the FDA based on their level of risk to patients and the ability to mitigate these risks. Three classes of risk control are identified based on the level of evidence required to demonstrate the safety and effectiveness of a specific medical device. The classification assigned to a device determines the pathway required for marketing of a device.

56.4.4.1 Class I: General Controls

Class I devices are those devices for which general controls are sufficient to provide reasonable assurance of the safety and effectiveness of the device. General controls apply to all medical devices. These consist of basic standards for ensuring consistent safety and effectiveness and include a requirement for annual registration by manufacturers, importers, distributors, repackagers, and relabelers; annual listing of devices to be marketed and activities to be performed using specific devices; strategies for prevention of device mislabeling; and Premarket Notification. General controls also include quality systems regulations related to good design, manufacturing practices, record keeping, and reporting. Examples of class I devices include manual surgical instruments which are not system specific, such as scalpels, retractors, drills, soft tissue elevators, and general surgical instruments (e.g., Cobb elevator). While most class I devices are exempt from premarket review, a limited number of devices (e.g., surgical gloves, removable skin staples) require Premarket Notification through the 510(k) process.

56.4.4.2 Class II: Special Controls

Devices which cannot be classified into class I because general controls by themselves are insufficient to provide reasonable assurance of safety and effectiveness may be classified into class II if special controls are adequate to provide this assurance. Special controls are evaluations which are specific to the device. Special controls may include specific labeling requirements, preclinical performance testing based on FDA guidance or recognized standards such as those described by the American Society for Testing and Materials (ASTM) International or International Organization of Standards (ISO), patient registries, postmarket surveillance, or clinical data. Most orthopedic spinal devices are designated as class II (e.g., posterior thoracic and lumbar pedicle screw systems for scoliosis and kyphosis). Premarket Notification through the 510(k) process is generally required for marketing of class II devices.

56.4.4.3 Class III: Premarket Approval

Class III devices are those for which general controls are not adequate to provide reasonable assurance of safety and effectiveness and the available data are insufficient for establishment of special controls. Class III devices are usually intended to support or sustain life or prevent substantial health impairment but are accompanied by substantial risk of causing harm to the patient. Such devices may include technologies which raise new questions regarding safety or effectiveness as a consequence of associated novel technologies and/or novel indications.

Such devices are subjected to the highest level of scrutiny which generally includes a complete preclinical and clinical review conducted during the premarket approval (PMA) application. Some class III devices also require post-approval clinical studies. Examples of class III spinal devices include total disc replacement devices and facet replacement devices.

56.4.5 Additional Considerations Related to Device Classification

56.4.5.1 Preamendments Versus Postamendments Devices

The Medical Device Amendments of 1976 divided medical devices based on when they were introduced into commercial distribution. A *preamendments device* is one that was in commercial distribution before May 28, 1976, the date the Medical Device Amendments to the Federal Food, Drug, and Cosmetic Act were signed into law. These devices were initially classified as class I, II, or III. A device that was not available on the market before the passage of the Medical Device Amendments of 1976 is generally referred to as a *postamendments device*.

56.4.5.2 Classified Devices Versus Reclassified Devices Versus Unclassified Devices Versus Not-Classified Devices

Classified Devices

The FDA has established classifications for approximately 1700 different generic types of devices and grouped them into 16 medical specialties referred to as *panels*. Each of these generic types of devices is assigned to one of three regulatory classes based on the level of control necessary to assure the safety and effectiveness of the device. Searchable databases located on the FDA website provide information on how various devices are regulated by the CDRH.

Reclassified Devices

Based on new information, the FDA may, on its own initiative or upon petition by any interested person, change a device classification. The new information used in this determination must be publicly available valid scientific information as defined in Section 513(a)(3) of the FD&C Act and 21 CFR 860.7(c)(2).

Unclassified Devices

An unclassified device is a preamendments device for which a classification regulation has not been promulgated. Unclassified devices require submission of a 510(k) Premarket Notification to the CDRH. Until the unclassified device type has been formally classified and a regulation established, marketing of new devices within this type will require submission of a 510(k) Premarket Notification to the FDA. Once classified, these devices may require submission of a PMA application, a 510(k), or be exempt from any premarket submission requirement.

Not-Classified Devices

A not-classified device is a postamendments device for which the agency has not yet reviewed a marketing application or for which the agency has not made a final decision on such a marketing application.

56.4.6 Classification Product Code

Classification product codes are a method of internally classifying and tracking medical devices used by the FDA and are also used for import/export purposes [14]. CDRH-regulated medical device product codes consist of a 3-letter combination which associates a device's type with a product classification. Classification product codes help to delineate technology and indication subgroups within a regulation and may be identified using the product classification database on the FDA's website [15]. Classification product codes are a key element with respect to many aspects of device regulation including the reporting of adverse events and product problems in medical device reports (MDRs).

56.4.7 Major Regulatory Pathways for Medical Devices

The pathways to market for a medical device in the United Sates [16] are often complex from the perspective of someone outside of regulatory

Table 56.3 Comparison of 510(k), PMA, and HUD/HDE regulatory pathways

	510(k)	PMA	HUD/HDE
Approval benchmark	Substantial equivalence	Safety and effectiveness	Safety and probable benefit
Clinical data requirements	Requirement limited to a small percentage of submissions	Clinical data generally required Randomized controlled trial design is common	Clinical data generally required Often limited to single-arm trial with safety and feasibility assessments
Population	Unrestricted	Unrestricted	Disease affects less than 4000 patients per year No similar device available through 510(k) or PMA
Requirement for IRB approval after marketing	None	None	Yes
User fees required	Yes	Yes	No
Profit restriction for device manufacturers	No	No	Device may be sold for profit if the number of devices distributed in calendar year does not exceed the ADN[a]
FDA review cycle time	90 days	180 days	75 days

[a]Annual distribution number (ADN) is determined by multiplying the number of devices reasonably needed to treat, diagnose, or cure an individual multiplied by 4000

affairs and depend on multiple factors including device classification, existence of similar devices previously classified for use by the FDA, the identified target population, and the date at which a device was first legally marketed (Table 56.3).

56.4.7.1 510(k) Pathway

The most common mechanism by which medical devices are cleared for commercial distribution in the United States is by Premarket Notification through the 510(k) program [17]. In the absence of a specific exemption, sponsors must notify the FDA regarding intent to introduce a new class I or class II device into interstate commerce. This is achieved by submission of a Premarket Notification under Section 510(k) of the FD&C Act. Sponsors are required to demonstrate that their medical device is *substantially equivalent* to a *predicate device*. A predicate device is a device that was legally marketed prior to May 28, 1976 (preamendments device), a device which has been reclassified from class III to class II, or a device which has previously been found to be substantially equivalent (SE) through the 510(k) Premarket Notification process. A medical device is determined to be substantially equivalent in comparison to a predicate device if the new device has the same intended use, and the FDA has determined that it has the same technologic characteristics as the predicate device. If the new device has a new intended use or different technologic characteristics, the FDA may determine the device as substantially equivalent if the information in the 510(k) does not raise different questions regarding safety and effectiveness or change the therapeutic effect, and the information submitted, including appropriate scientific or clinical data, shows that the device is at least as safe and effective as the predicate device. If a device is determined as not substantially equivalent (NSE), the device is automatically considered class III. Applicant options following an NSE determination include resubmission of another 510(k) with new data or pursuit of an alternate pathway to market (i.e., PMA, HUD/HDE, or de novo pathways).

In limited specific situations, class III devices may be the subject of a 510(k) Premarket Notification according to Section 515(b) of the

FD&C Act. The devices covered by 515(b) requirements fall into two categories:

1. Devices in commercial distribution before May 28, 1976 (preamendments devices), that were subsequently classified by the FDA as class III devices by means of classification regulations promulgated under Section 513 of the Act.
2. Devices offered for commercial distribution on or after May 28, 1976 (postamendments devices), that are determined through the 510(k) process to be substantially equivalent to class III preamendments devices.

   After classification, in the case of category (1), or after substantial equivalence has been established by 510(k) Premarket Notification submissions, in the case of category (2), these devices may continue to be marketed commercially without an approved PMA until the FDA publishes a final rule under 515(b) to require the filing of a PMA application.

56.4.7.2 PMA Pathway

Premarket approval (PMA) is the FDA process for scientific and regulatory review to evaluate the safety and effectiveness of most class III medical devices. These devices are considered to require the highest level of regulatory scrutiny since insufficient information exists to allow use of general or special controls to provide reasonable assurance that such devices are *safe and effective* for their intended use, based on sufficient valid scientific evidence, such as well-controlled clinical studies or significant detailed data on long-term clinical experience with a device. Medical devices which require premarket approval include most class III preamendments devices, class III postamendments devices, and devices found not substantially equivalent to class I and class II predicate devices through the 510(k) process.

The requirements for PMA applications are mandated by statute (21 CFR Part 814.20). These requirements include an indication for use statement, device description, summary of nonclinical and clinical studies, details regarding principles of device operation and manufacturing, sterility information, proposed device labeling, financial certification and disclosure, and environmental assessment [18]. Sponsors are encouraged to obtain the FDA feedback prior to submission of a PMA application through the agency's Pre-Submission Program [19]. The majority of PMA applications involve clinical data, and clinical studies which are performed to collect such data are required to be conducted under the investigational device exemption (IDE) regulations (21 CFR Part 812). PMA applications for first-of-a-kind devices require review by an FDA advisory panel prior to authorization to market such devices. Additionally, a manufacturing facility inspection is required to assess compliance with good manufacturing practices prior to approval.

56.4.7.3 Humanitarian Use Device (HUD)/Humanitarian Device Exemption (HDE) Pathway

The Humanitarian Use Device (HUD)/Humanitarian Device Exemption (HDE) pathway is an alternative regulatory pathway intended to create incentive for development of devices for use in the diagnosis or treatment of diseases that affect or are manifested in a small number of individuals (<4000 patients in the United States per year). This pathway involves a two-part process. Part 1 requires submission of an HUD designation request to the Office of Orphan Product Development in the Office of Special Medical Programs under the Office of the FDA Commissioner. If an HUD designation is granted by the Office of Orphan Product Development, the sponsor may submit an HDE application to the Center for Devices and Radiological Health (CDRH) or the Center for Biologics Evaluation and Research (CBER) for marketing review [20].

The HDE regulatory pathway differs from the PMA pathway in that the requirement to demonstrate reasonable assurance of effectiveness required for PMA application approval is waived. For an HDE, a sponsor is required to demonstrate

that their device does not pose an unreasonable or significant risk of illness or injury and that the probable benefits (in lieu of effectiveness) to health outweigh the risks of injury or illness from using the device. This exemption from the effectiveness requirements of the FD&C Act lessens the time and cost burdens associated with performing large-scale, controlled clinical trials typically required to demonstrate safety and effectiveness. The HDE pathway permits approval for marketing of devices that demonstrate a favorable benefit-risk profile, based on clinical experience in combination with relevant nonclinical testing. The sponsor is also required to demonstrate that their device would not otherwise be available without this exemption and that no comparable devices are available (other than another device with an approved HDE application or a device currently under study under an approved IDE). Use of an HDE device requires initial and continuing IRB approval. Until recently, manufacturers had restrictions on the ability to make a profit on HDE devices, but this restriction has been modified through subsequent legislation including the FDA Safety and Innovation Act (FDASIA, 2012). An example of a pediatric spinal device approved for marketing under an HDE was the Vertical Expandable Prosthetic Titanium Rib® (VEPTR, DePuy Synthes). In 2014, the Vertical Expandable Prosthetic Titanium Rib® (VEPTR/VEPTR II, DePuy Synthes) was cleared by the FDA through the 510(k) pathway.

56.4.8 Research Studies Related to Pediatric Medical Devices

A clinical trial may be required before marketing approval of a PMA submission or clearance of a 510(k) application. Clinical studies are most often conducted to support PMA submissions, whereas only a small percentage of 510(k) submissions require clinical data. All clinical evaluations of investigational devices, unless exempt, must have an approved investigational device exemption (IDE) before the study is initiated [21]. An approved IDE permits a device to be shipped lawfully for the purpose of conducting investigations of the device without complying with other requirements of the FD&C Act that would apply to devices in commercial distribution. Prior to initiation of testing in human subjects, proof of concept and relative safety are required to be established through preclinical studies. Additional regulatory requirements include an investigational plan approval by an IRB, informed consent, labeling stating that the device is for investigational use only, study monitoring, and submission of required records and reports. In addition, for pediatric device studies, the investigator must make adequate provisions for soliciting the assent of a child participant when children are capable of providing assent.

56.4.9 Postmarket Regulatory Requirements

Postmarket regulatory requirements for medical devices serve an important role in identifying issues which arise in the period after initial device marketing when the device is used in a "real-world" setting [22]. These issues include rare, serious, or unexpected adverse events, adverse events that occur with long-term device use, and adverse events associated with vulnerable populations. The medical device reporting (MDR) regulations (21 CFR Part 803) contain specific mandatory requirements for manufacturers, importers, and device user facilities to report certain device-related adverse events and product problems to the FDA, such as death, serious injury or illness, and device malfunctions or failures. Health-care professionals, patients, caregivers, and consumers are also encouraged to submit voluntary reports about serious adverse events that may be associated with medical device products to MedWatch, the FDA's Safety Information and Adverse Event Reporting Program or through the MedWatcher mobile app [23]. The Manufacturer and User Facility Device Experience (MAUDE) database houses the MDRs submitted to the FDA.

The SMDA of 1990 provided the FDA with two additional postmarketing tools: *postmarket surveillance* for the monitoring of products after their clearance to market and *device tracking* for

maintaining traceability of certain devices to the user level. Section 522 of the FD&C Act authorizes the FDA to require postmarket surveillance studies for class II and class III devices reasonably likely to have a serious adverse health consequence, expected to have significant use in pediatric populations, intended to be implanted in the human body for more than 1 year, or intended to be a life-sustaining or life-supporting device used outside of a user facility. These studies are separate from post-approval studies required at the time of approval of a PMA or HDE. Medical device tracking is intended to facilitate notification and recall in the event a device presents a serious risk to health that requires prompt attention. No pediatric spinal devices are currently subject to tracking requirements.

56.5 Regulatory Status of Common Spinal Devices Intended for Use in Pediatric Populations in the United States

56.5.1 Spinal Devices Intended to Achieve Fusion for Treatment for Pediatric Spinal Conditions

A limited number of spinal devices are cleared or approved specifically for use in pediatric populations to promote spinal fusion. Currently, no intervertebral body fusion devices are cleared for specific use in pediatric patient populations. Certain types of posterior spinal systems intended for fusion indications are cleared for use in specific pediatric populations. The indications for use statements and regulations are silent regarding the pediatric use of anterior spinal systems to achieve fusion for treatment of spinal deformities.

Modern posterior spinal instrumentation systems consist of rods and bone anchors used to create a system that can span from the upper thoracic spine to the sacrum through use of a variety of components and configurations to accommodate individual patient anatomy. Spinal anchors form the bone-implant interface, longitudinal members connect the anchor points, and transverse connectors link the longitudinal members for additional stability (Table 56.4). In the United States, Harrington [24] advanced the concept of posterior internal fixation of the spine which led to marketing of the Harrington system, comprised initially of hooks anchored at the proximal and distal end vertebra and connected to longitudinal rods. Subsequently, Harrington [25, 26] described use of pedicle screws in combination with fusion for treatment of severe spondylolisthesis in adolescent patients. As posterior screw systems for various spinal indications were first marketed in the United States before the 1976 Medical Device Amendments, these devices are considered preamendments devices. The original Harrington hook and rod system and systems using other non-pedicle bone anchors (e.g., sublaminar or spinous process wires) were subsequently classified into class II by the FDA. However, pedicle screw spinal systems remained unclassified, until the FDA published a final rule classifying certain previously unclassified preamendments pedicle screw spi-

Table 56.4 Posterior spinal systems intended for fusion indications for treatment of specific pediatric spinal conditions stratified by spinal anchor types

Anchor type	Location	Intended use/device type	Product code	Device classification	Regulation
Hooks	Cervical, thoracic, lumbar	Posterior/ non-pedicle	KWP	II	21 CFR 888.3050
Wires/clamps/cables Sublaminar or facet location	Cervical, thoracic, lumbar	Posterior/ non-pedicle	OWI	II	21 CFR 888.3010
Screws	Thoracic, lumbar, sacral	Posterior/pedicle	OSH	II	21 CFR 888.3070

nal systems in the July 27, 1998, FR (amended in a Technical Amendment dated May 22, 2001). According to CFR Title 21, Part 888, Subpart D, Section 888.3070, pedicle screw systems were classified as:

Class II (special controls), when intended to provide immobilization and stabilization of spinal segments in skeletally mature patients as an adjunct to fusion in the treatment of the following acute and chronic instabilities or deformities of the thoracic, lumbar, and sacral spine: severe spondylolisthesis (grades 3 and 4) of the L5-S1 vertebra; degenerative spondylolisthesis with objective evidence of neurologic impairment; fracture; dislocation; scoliosis; kyphosis; spinal tumor; and failed previous fusion (pseudarthrosis).

Class III (premarket approval), when intended to provide immobilization and stabilization of spinal segments in the thoracic, lumbar, and sacral spine as an adjunct to fusion in the treatment of degenerative disc disease and spondylolisthesis other than either severe spondylolisthesis (grades 3 and 4) at L5-S1 or degenerative spondylolisthesis with objective evidence of neurologic impairment.

Expansion of the indications for use for these previously cleared pedicle screw systems to pediatric populations occurred via the 510(k) process as these devices were found substantially equivalent to other class II products already marketed. Specific spinal systems are currently cleared for use in pediatric populations for treatment of adolescent idiopathic scoliosis, spondylolisthesis, and fracture/dislocation (due to tumor and/or trauma).

56.5.2 Spinal Devices Intended for Nonfusion Indications for Specific Pediatric Spinal Conditions

Development of spinal systems for nonfusion treatment for progressive pediatric spinal conditions is an area of active scientific, clinical, and regulatory interest. The Harrington spinal system, comprised of stainless steel rod(s) fixed to the spine with hooks at both ends and implanted through a posterior spinal approach, was developed for both nonfusion and fusion treatment of lateral (coronal) plane spinal curvature or scoliosis. In 1962 Harrington [24] stated that "progressive scoliosis in a child less than 10 years old could be managed with the apparatus alone without fusion, whereas in a child more than 10 years old fusion should usually be done at the time of the initial correction." He further cautioned that no form of treatment with or without spinal fusion can be considered definitive in a child whose axial skeleton is still growing. Since the introduction of the Harrington system, multiple implant systems have evolved for nonfusion treatment of a wide range of conditions affecting the pediatric spine (Table 56.5).

In 2003, the VEPTR device (DePuy Synthes) was approved by the FDA (H030009) through the HUD/HDE regulatory pathway for treatment of thoracic insufficiency syndrome (TIS). TIS is a condition in which severe deformities of the chest, spine, and ribs prevent normal breathing and lung growth and development. The regulatory approval of the VEPTR device was based on feasibility and multicenter IDE clinical trials conducted between 1990 and 2004. The VEPTR device is attached perpendicularly to the patient's natural ribs (vertically) and/or to the lumbar vertebrae or sacrum. It mechanically stabilizes the chest wall and enlarges the thorax to improve respiration and lung growth. Once the VEPTR is in place, its design allows for expansion, anatomic distraction, and/or replacement of component parts through less invasive surgery than the initial implantation surgery.

In 2014, the CD HORIZON® Growth Rod Conversion Set (Medtronic Sofamor Danek, USA) received clearance (K133904) through the 510(k) process for use in treatment of patients with potential for additional spinal growth under 10 years of age who require surgical treatment to obtain and maintain correction of severe, progressive, life-threatening, early-onset spinal deformities associated with thoracic insufficiency, including early-onset scoliosis.

Table 56.5 Cleared or approved spinal device types intended for nonfusion indications for specific pediatric spinal conditions

Device type	Location	Product code	Device classification	Regulation
Vertical Expandable Prosthetic Titanium Rib (VEPTR)	Thoracic, lumbar, sacral, thoracic (rib) cage	MDI	Preamendments, unclassified	None
Growth Rod Conversion Set (device actuation requires open surgical procedure, i.e., mechanical mode of action)	Thoracic, lumbar	PGM	Preamendments, unclassified	None
Spinal Growth Guidance System	Spinal Growth Guidance System	PGM	Preamendments, unclassified	None
Growth Rod Expandable Device (device actuation achieved without an open surgical procedure, i.e., noninvasive mode of action, e.g., magnetic actuation)	Thoracic, lumbar	PGN	Preamendments, unclassified	None

Nonfusion spinal procedures using mechanically based expandable distraction instrumentation, also described as "growing rods," provide a method for controlling thoracic spinal deformity while permitting spinal growth to occur. This technique requires periodic open surgical procedures in order to achieve lengthening of the spinal implant construct through manual methods. The CD HORIZON® Growth Rod Conversion Set consists of various connectors designed to convert a rod-based spinal system utilizing screw and/or hook anchors into a spinal construct which enables growth. The agency determined the device as substantially equivalent for the indications for use noted above by comparison to a legally marketed predicate device marketed in interstate commerce prior to May 28, 1976, the enactment date of the Medical Device Amendments. The FDA designated the Harrington rod system (9/11/13) as a preamendments device for nonfusion surgical correction in this patient population. The CD HORIZON® Growth Rod Conversion Set is currently designated as an unclassified device since a classification regulation has not yet been promulgated (see Table 56.5).

Subsequently in 2014, the MAGEC® Spinal Bracing and Distraction System (Ellipse Technologies, Inc.) received clearance (K140178) through the 510(k) process for use in skeletally immature patients less than 10 years of age with severe progressive spinal deformities (e.g., Cobb angle of 30° or more; thoracic spine height less than 22 cm) associated with or at risk of TIS. The MAGEC® Spinal Bracing and Distraction System consists of a sterile single-use titanium spinal rod system that is surgically implanted in the posterior aspect of the spine, using spinal anchors consisting of specific pedicle screws and/or hooks (Stryker® Xia® system components). The implant system, used as a single- or dual-rod construct, braces the spine during growth to minimize progression of scoliosis. The titanium rod includes an actuator portion that holds a small internal magnet which can be rotated noninvasively to lengthen or shorten the rod(s) by use of a nonsterile, handheld external remote controller (ERC), thereby providing adequate bracing of the spine and minimizing progression of scoliosis. MAGEC® Spinal Bracing and Distraction System received clearance through the 510(k) process based on nonclinical and clinical data and was determined to be substantially equivalent to the preamendments Harrington rod system. The MAGEC® Spinal Bracing and Distraction System is also currently designated as an unclassified device as a classification regulation has not yet been promulgated (see Table 56.5).

Recently, the SHILLA™ Growth Guidance System (Medtronic Sofamor Danek, USA) received clearance (K140750) through the 510(k) process for use in skeletally immature patients less than 10 years of age with the potential for additional spinal growth, who require surgical treatment for correction and maintenance of severe, progressive, life-threatening early-onset deformities, including early-onset scoliosis, which are associated with or at risk of thoracic insufficiency syndrome. The SHILLA™ Growth Guidance System was determined to be substantially equivalent to the CD HORIZON® Growth Rod Conversion Set (K133904) and to the pre-amendments Harrington rod spinal system. Additional recent pediatric spinal device clearances via the 510(k) pathway include K142114 (Xia® Growth Rod Conversion Set, Stryker Spine) and K141509 (ISOLA® and EXPEDIUM® Growing Spine Systems, DePuy Synthes Spine).

56.6 Future US Regulatory Considerations Regarding Pediatric Spinal Devices

A variety of growth-sparing spinal implant systems for treatment of spinal deformities in skeletally immature patients have been developed over the past decade. Recently, these devices have been classified by surgeons according to the type of corrective forces exerted on the spinal column by the implant system [27] and include posterior distraction-based systems (e.g., VEPTR, growing rods), posterior guided growth systems (e.g., SHILLA system), and anterior compression-based systems (e.g., spinal staples, spinal tethers). The current and future regulatory status of these various device types in the US market is complex and varies according to device type as well as relevant clinical and nonclinical data. As new pediatric spine devices progress into the US market in the future, these emerging growth-enabling technologies will expand the armamentarium of the spinal surgeon for treatment of various pediatric spinal disease states (Table 56.6).

Table 56.6 Present and future spinal device options for treatment of various pediatric spinal disease states

Disease state	Pediatric spinal device treatment options
Early-onset scoliosis	Orthoses, rigid spinal systems intended to achieve fusion, VEPTR, growing rods (mechanical and noninvasive types), Spinal Growth Guidance System, spinal tethers[a], vertebral body staples[a]
Thoracic insufficiency syndrome	VEPTR, growing rods (mechanical and noninvasive types), Spinal Growth Guidance System
Adolescent idiopathic scoliosis	Orthoses, rigid spinal systems intended to achieve fusion, spinal tethers[a], vertebral body staples[a]
Spondylolisthesis	Orthoses, rigid spinal systems intended to achieve fusion
Fractures/dislocations (due to tumor and/or trauma)	Orthoses, rigid spinal systems intended to achieve fusion

[a]These device types have not been cleared or approved by the FDA

Regarding distraction-based and guided growth nonfusion spinal systems currently available for on-label use in the US market, the CD HORIZON® Growth Rod Conversion Set (Medtronic Sofamor Danek, USA) and other cleared distraction-based systems, the MAGEC® Spinal Bracing and Distraction System (Ellipse Technologies, Inc.), the Vertical Expandable Prosthetic Titanium Rib® (VEPTR/VEPTR II, DePuy Synthes), and the SHILLA™ Growth Guidance System (Medtronic Sofamor Danek, USA) are unclassified devices and require future FDA classification. This will require (1) recommendation from a device classification panel (an FDA advisory committee); (2) publication of the panel's recommendation for comment in the FR, along with a proposed regulation classifying the device; and (3) publication of a final regulation classifying each device in the lowest regulatory class consistent with protection of the public health and the current statutory scheme for device regulation.

Regarding anterior compression-based spinal systems for pediatric patients, no devices in this class are currently cleared or approved in the United States. A US-based IDE study to evaluate the use of spinal stapling (HemiBridge™ Clips, Spine Form, LLC) in a subpopulation of pediatric idiopathic scoliosis patients is currently underway.

56.7 International Device Regulation Perspectives

Processes for regulation of medical devices worldwide are variable and ultimately remain subject to control by the regulatory body of the specific country. In 2010, approximately 30 % of countries possessed a developed framework for regulation of medical devices; approximately 30 % of countries possessed a partial framework for medical device regulation; and approximately 20 % of countries either lacked regulations or were in the initial stages of development of a regulatory framework for medical devices [28]. Diversity regarding regulatory processes creates challenges for medical device manufacturers who wish to market their medical devices globally, as well as for health-care professionals who desire access to state-of-the-art medical device technologies.

Common principles across the regulatory framework of various countries have been identified by the World Health Organization (WHO) and correspond to the various phases of a medical device's lifespan [29]. In the premarket phase, medical devices are required to meet specific safety and performance requirements, quality system requirements, and labeling requirements. A classification strategy which ranks medical devices according to level of potential risk associated with their use and determines the appropriate level of regulatory control proportional to these risks is an essential component of device assessment during this phase. Classification systems generally stratify devices into three or four risk levels or classes. For example, in the European Union (EU), medical devices are classified into four categories (class I, class IIa, class IIb, and class III) based on risk, considering factors such as invasiveness and duration of contact with the body [30, 31]. In contrast, the China Food and Drug Administration (CFDA) utilizes a three-tier risk-based medical device classification system. Regulatory bodies authorize medical devices for market entrance through specific processes. For example, in the EU, a compliance label, or CE mark, is issued and permits marketing across all EU member states. Transition to the next phase of the regulatory process occurs as a medical device is placed on the market. Important regulatory activities during this phase include establishment registration to permit vendor tracking, as well as prohibitions against fraudulent or misleading advertising. The postmarket phase is the final phase in the regulatory framework. This phase, also termed postmarket surveillance, includes postmarket surveillance studies and adverse event reporting and is interrelated with quality system requirements and good manufacturing practices (GMPs).

International efforts have been undertaken to foster consistency and standardization regarding laws and regulations relating to medical devices across various countries through publication of guidance documents (Table 56.7). The Global Harmonization Task Force (GHTF), consisting of voluntary representatives from medical device regulatory authorities including the European Union, the United States, Canada, Japan, and Australia, initiated efforts toward international harmonization of the regulation of medical devices. Subsequently, these initial efforts have been assumed by the International Medical Device Regulators Forum (IMDRF), whose stated goal is to accelerate international medical device regulatory harmonization and convergence. The IMDRF regulatory members include Australia, Brazil, Canada, China, Europe, Japan, Russia, and the United States. The World Health Organization is an official observer. The Asian Harmonization Working Party and Asia-Pacific Economic Cooperation (APEC) Life Sciences Innovation Forum (LSIF) Regulatory Harmonization Steering Committee are both affiliate organizations with IMDRF. Expansion of such efforts to involve additional countries is a stated goal of this organization.

Table 56.7 Representative global medical device regulatory bodies

Country or region	Medical device regulatory body
United States	Food and Drug Administration, Center for Devices and Radiological Health (CDRH) http://www.fda.gov/MedicalDevices/default.htm
European Union	European Commission http://ec.europa.eu/health/medical-devices/index_en.htm
China	China Food and Drug Administration (CFDA) http://eng.sfda.gov.cn/WS03/CL0755/
Australia	Australian Therapeutic Goods Administration (TGA) https://www.tga.gov.au/
Japan	Pharmaceutical and Medical Devices Agency (PMDA) http://www.pmda.go.jp/english/
Canada	Health Canada http://www.hc-sc.gc.ca/dhp-mps/md-im/index-eng.php
South Korea	Korean Ministry of Food and Drug Safety (MFDS) http://www.mfds.go.kr/eng/index.do;jsessionid=1ndhfPwEQJxyWink8pmVEOh6CzYdhUTePmRww2JGboC1kiXUSVRhZtoHB4pW7Tyo

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A Journey from the Past to the Future

57

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57.1 Yesterday

Although first attempts at treating spinal deformities date as far back as many years before Christ, those groundbreaking changes that define our contemporary practice of spinal surgery were accomplished by senior spine surgeons still actively practicing today. If we recall that the history of the first universal implant, pedicle screw, multisegmentary instrumentation, or instance of intraoperative neuromonitoring was only 50–60 years ago, we can better grasp the amount of distance that has been traversed in such short time.

Paul R. Harrington, M.D., does not hold historical significance because he invented a type of spinal implant that would solve the problems of innumerable patients, but because he took the first and most crucial step in making surgery imaginable, possible on the previously untouchable human spine. Harrington's implant today may be only a curiosity of the past. However, the inspiration and courage it kindled endowed contemporary spine surgeons with the resolve to go beyond the acceptable and aim for better, the best results. With the demonstration of this kind of resolve, a wide range of patients from children and adolescents to octogenarians have been able to benefit from the rewards of corrective spinal surgery.

Despite these rapid developments in spinal deformity surgery, early-onset spinal deformity (EOSD) remained an orphan area that did not receive the attention it deserved. The causes for

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this paucity of interest can be traced back to the rarity of this condition, the lack of sufficient infrastructure and superstructure in many clinics to handle this oft-complicated patient group, and a lack of knowledge regarding the natural history and effective management of the pathology.

Fortunately, today the field of EOSD has changed drastically, becoming an area of passionate discussion, generation of tremendous evidence, and rapid discovery of knowledge. The incendiary effect of two factors that serve to accelerate and facilitate this striking change cannot be denied. The first is the definition of the thoracic insufficiency syndrome by Dr. Robert Campbell and the subsequent development of his implant to correct the problem. The second is the establishment of new standards for a well-known technique that had fallen into disfavor due to its previously disappointing results: the growing rod.

Many other advances have contributed to the invention of new treatment techniques, the design of new implants, and, more importantly, a better understanding of the disease and the conception of more realistic solutions. Among these are the formation of a committee on growing spine problems within the Scoliosis Research Society (SRS) that inspired new research and contributed to its coordination; the International Congress on Early Onset Scoliosis (ICEOS), the first of which convened in Madrid in 2007, bringing together all interested parties (surgeons, nurses, pulmonologists, basic scientists, and others) and providing a consistent scientific platform for the exchange of ideas; the founding of the Growing Spine Study Group (GSSG) and Children's Spine Study Group (CSSG), the collection of multicenter data; and, last but not the least, the publication of this book, dedicated exclusively to spinal disorders in young children.

57.2 Today

Early-onset spine deformities are no longer orphan diseases. In the last few years, a number of high-evidence level studies have been completed, addressing a multitude of topics in EOSD, from classification to the definition of risk factors, from the development of patient-specific outcome analysis instruments to how the natural history affects the spine and non-spine organs, and from sophisticated and objective clinical and radiographic evaluation methods to efficient and safe treatment options. The EOSD literature has proven itself to possess a trend for exponential expansion. Through all these developments, surgeons who find themselves facing a young child with a crooked spine and a distorted chest cage can now feel more knowledgeable regarding treatment and are able to assist these children's families in looking more hopefully upon brighter future. However, although we know we are on the right path, it is obvious to us that the road is long and beset with many obstacles. It undoubtedly remains important to achieve for these children sufficiently long, functionally, and cosmetically aligned spines comparable to those of their peers. However, the questions that loom before us are these: what is the price to be paid for the attainment of this goal, and how close can we get, at the end of the road, to achieving normalcy regarding the spine in particular and the patient's physical and mental well-being in general?

57.3 Tomorrow

After emphasizing the significance of the juncture at which we have arrived, and highlighting the greatness of the distance traversed, we can summarize the points that are still in requirement of intense research as such.

Today, spinal deformity surgeons can perform operations that as short a time as 10 years ago appeared to be no more than figments of the imagination. The safe three-dimensional reconstruction of the spine, regardless of severity of the deformity or etiology, is now a realistic expectation, and fortunately, thousands of children around the world can now look forward to better health due to it. Despite these truly remarkable advances, two significant abilities still elude us: recognize early those patients with a predisposition to deformity, and reliably prevent progression of, or even reverse, existing deformity.

Preventative medicine is the successful resolution of conflict between disease and physician

without the spillage of blood. However, spinal surgeons can only enter the picture at the conclusion of the process, and the treatment they apply cannot bring back what has been lost up until that point or prevent damage before it occurs. Although the genetic background of spinal deformities has been on the forefront of scientists' agendas for many years, it is impossible to say that great strides have been made regarding this subject. Genetic research regarding EOSD is at the time almost nonexistent. However, the necessary knowledge for early recognition of individuals under risk, perhaps during the intrauterine period or even before pregnancy, would provide a unique opportunity for the development and institution of preventative measures.

Studies on the genetic background of EOSD are required, which would command intensive effort and determination, especially as this is not an area where short-term projects and quick results abound.

The possible relationship between melatonin and adolescent idiopathic scoliosis (AIS) discovered during the late 1990s and early 2000s caused much excitement among spinal surgeons. The possibility of medically controlling adolescent curves resounded with many. Since then, the lack of progress that was hoped for in this area may have caused disappointment. However, persistent research into the mechanisms that create deformity will undoubtedly result, one day, in the discovery of methods that enable us to reverse them. Through this kind of advances, it will become possible to engage these curves in a more humane manner, without the bloodshed, and to be proactive by nipping the deformity in the bud before it gains a hold of the spine.

Studies regarding the etiology of deformity and the management/modulation of these etiological factors utilizing medical, biological, and other such methods are the most critical advances expected in the near future.

Contemporary techniques utilized in the treatment of EOS deformities such as surgery and casting expose young children to the potentially detrimental effects of repetitive anesthesia. Strong evidence exists regarding these negative effects on the immature brain, indicating that children undergoing repetitive anesthesia sessions grow up to suffer from learning disabilities and abnormal behavior. Understanding this matter will continue to grow with the conclusion of some large-scale, prospective, multicenter trials. If these concerns are legitimate, the recent popularity of cast treatment, which has reemerged as an alternative to growth-friendly surgical methods because of its ability of controlling curve progression without affecting spinal growth and mobility, will need to be reexamined.

There exists a great need for data on the effects of undergoing anesthesia during childhood on growth and development. Detailed studies examining the effects of the various types of anesthetic agents and number of applications on the same should be conceived.

Childhood is a period of life that should be experienced without anxiety or worry about the future. However, the existence of a condition that precludes a profoundly joyful experience of this life chapter, one that brings with itself a necessity for serious medical and surgical intervention, creates a massive stress generator for the child and parents regardless of the innate effects of this condition on the body. This stress will be greatly magnified if, during the treatment of this condition, the child is forced to leave their social circle for extended periods of time, to be imprisoned in cold and desolate hospital environments full of frightful strangers, and, more importantly, to undergo multiple painful procedures and bouts of immobilization afterward.

Treatments such as the traditional growing rods, vertical expandable titanium ribs (VEPTR), and cast applications under anesthesia require long-term, repetitive dependency on hospitals and doctors, causing children to spend many birthdays and holidays in the hospital. Should complications occur, the process becomes even more difficult on the children and they must endure greater stress. This shifts the focus from

problems in the present to possible, but not certain, implications of the disease in the future.

Although the psychological effects of other childhood disorders have been the subject of many previous studies, a paucity of information exists regarding health problems specific to EOSD, the treatments intending to decrease these problems, their effects on the psychologies of the individual and the family, or the variation in such effects as pertains to the severity of disease and success of treatment.

It is entirely insufficient to evaluate the long-term effects of EOSD and their treatments on the individual child, solely by clinical and radiographic methods. Ability to see the complete picture will emerge only if psychological aspects of the situation are also considered and included within the study design. Evaluation with instruments specifically developed for this disease with inclusion of the special situations that may arise during its treatment is essential.

Historically, the measurement of the success of treatment for spinal deformities was based on radiological parameters alone. Improvements in angles were considered evidence of treatment success. Once it was recognized that clinical improvement and radiology did not always correlate, the necessity arose to include other parameters such as cosmetic determinants (shoulder balance, rib hump, waistline asymmetry, etc.) and those of functional capacity as well. The introduction into our daily practice of health-related quality of life tools that aim to determine objectively the perception of the patient of the clinical and radiological improvements imparted upon their body with or without treatment is relatively new. Studies have been performed on the pediatric age group aiming to determine the impact on quality of life of some disorders both in childhood and later in adulthood and the outcomes of treatment. The parent is relied upon for the acquisition of data during childhood that can, in adolescence and adulthood, be obtained from the patient themselves. The reliability of this secondhand information is subject to controversy. Again, evaluations performed at the beginning and at the end of the process when the child is able to communicate will be obtained from two different people, who, even though sharing a common living space, are still two different people with different experiences, making an objective and reliable comparison impossible. Having never experienced the normal, the expectations of children with chronic disease are shaped by their afflictions, introducing new and fundamental difficulties for the comparison of the data obtained from them to that obtained from healthy individuals.

Furthermore, EOSD includes a fairly heterogeneous group of patients, ranging in etiology from congenital deformities with serious pulmonary comorbidities to serious muscular dystrophies with a life expectancy of only one or two decades and from spastics with profound mental retardation to spina bifida patients burdened to go through life with immobile, insensate lower extremities. Even though it may be a good start, it will soon be obvious that lumping these children afflicted with this huge spectrum of problems together, into the subject heading of "early-onset spine deformity" and hoping to evaluate them effectively, utilizing a single questionnaire, is insufficient.

The development of new, detailed questionnaires specific to EOSD that take into account the child's comorbidities that are sensitive enough to distinguish the negative effects of disease from those of treatment complications and show high compatibility between information collected from child and/or parents and caregivers is required. Studies utilizing such questionnaires will profoundly affect the future direction of treatment.

The concept of magnetically controlled growing rods (MCGR) that aim to halt the progression of young children's spinal deformities while protecting them from repeated surgical trauma and preserving their growth potential is undoubtedly going to become a new keystone in EOSD treatment. It is remarkable progress that this concept has become reality today, when very recently

it was only a dream for pediatric spinal deformity surgeons. This ability to adjust an implanted device painlessly and bloodlessly with a mechanism outside the body will enable us to reach our treatment goals more easily than with the often problematic traditional methods. This advance is significant not only for our current patients treated with this treatment today, but the great group of future patients not even born yet who will benefit from yet newer, perhaps even less invasive, methods inspired by the magnetic rod itself.

While the concept is groundbreaking and hopefully inspiring, the MCGR treatment is far from being ideal from technological and design points of view. This subject is virgin territory, with a great need for the conception and completion of many research projects.

The noninvasive ability of adjusting from outside the body the instrumentation inside it that aims to control spinal deformity, investigations on the feasibility of yet more different technologies other than magnetism such as smart metals, the creation of technologies that will in addition to pure distraction allow correction based on translation and derotation, and the design of a new instrumentation system more friendly to physiological spinal alignment are topics awaiting those researchers equipped with curiosity and drive.

Conclusions

In summary, judging by developments in the field of EOSD that have taken place in the last decade and today give rise to the most passionate discussions, we can expect even more fascinating, thought-provoking, and powerful advances in the near future. Under the light of healthy and objective analysis of the past, walking to a brighter future ceases to be a dream. Evidence-based guidance engendered by study groups, scientific associations, and discussion platforms such as the ICEOS will create a more productive and efficient work environment for surgeons dedicated to EOSD, allowing us to come together and, for our young children afflicted with spinal deformities, provide a better and brighter future.

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