The Management of Disorders of the Child's Cervical Spine

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This book is dedicated to all children who have disorders of the cervical spine and to the people who care for them.

Foreword

In the early 1970s, I became interested in the cervical spine, specifically congenital anomalies. That led to the publication of a report on the Klippel-Feil syndrome. I was fortunate to find a monograph entitled *Upper Cervical Spine* published in 1972. The authors, Detlef von Torklus and Walter Gehle, were from the Orthopedic Clinic and Outpatient Department of the University Hospital in Hamburg, Germany. They had done an extensive review of the literature and pathoanatomy of the cervical spine, and, importantly, nearly half of their book was devoted to children. The authors identified many normal physiologic and anatomic variations that frequently mimic pathology. Unlike the extremities, in spine issues, one cannot use a comparison X-ray of the opposite side. Their work identified variations in the pediatric spine and how they differed from the adult. This text became my go-to source for insight in complex cervical spine problems.

The Management of Disorders of the Child's Cervical Spine edited by Jonathan Phillips, Daniel Hedequist, Suken Shah, and Burt Yaszay continues that legacy. This text is comprehensive and includes an extensive review of previous literature by individuals knowledgeable in the management of children with complex cervical spine problems.

Part I, Basic Medical Science, is essential to effective diagnosis and treatment. This section contains important chapters on anatomy, biomechanics, radiology, advanced imaging, and current diagnostic techniques.

Part II, Clinical Aspects of Disorders of the Child's Cervical Spine, contains an extensive discussion of trauma to the immature spine and its potential for serious morbidity and mortality. There is a special section on cervical injury in the young athlete. The clinical aspects of many of the disorders that can affect the child's spine are presented in detail. This list is comprehensive and includes inflammatory conditions, infection, tumors, congenital anomalies, metabolic disorders, and bone dysplasias.

Part III, The Medical and Surgical Treatment of Cervical Disorders in Children, covers management—including conservative techniques such as immobilization and rehabilitation. Also included are surgical approaches, including current instrumentation, anesthesia, and neurological monitoring. There is a unique section on complications and revision surgery.

The strength of this text is that it is the product of an international panel of experts, all of whom are recognized authorities. This is coupled with the skillful oversight of Dr. Phillips and his colleagues to create a powerful text that will be an important clinical resource for many years. This will be exceedingly helpful to those involved in the management of cervical spine problems of children, and it continues the legacy of von Torklus and Gehle.

Ann Arbor, MI, USA Robert N. Hensinger, MD

Preface

There is no one reason why we wrote this book. It came about, as so many different things do, by way of a conversation at the dinner table. Suken Shah, MD; Burt Yaszay, MD; and I were talking at such a dinner table in Orlando at a meeting on early onset scoliosis. We all had a big interest in children's cervical spine problems, but agreed that they were pretty rare and there wasn't much of a forum for talking about them among us orthopedic surgeons who specialize in pediatric problems.

I give Burt the credit for the statement that "peds cervical spine is the last black hole in kids' spinal knowledge" or something like that. And with that prophetic statement the seed was sown.

Suken polled the membership of the Pediatric Orthopedic Society of North America (POSNA), and within a very short time, we had a small but enthusiastic group of interested surgeons who formed the nidus of a new study group which, for now at least, is called the Pediatric Cervical Spine Study Group (PCSSG). The members of this international group have contributed most of the chapters in this text, along with their fellows and other associates. We meet a few times a year at POSNA and Scoliosis Research Society (SRS) and International Congress on Early Onset Scoliosis (ICEOS) meetings and have been supported by these organizations. I'm very happy to acknowledge their support.

One of the early topics we discussed at PCSSG meetings was the possibility of writing a text that could guide the novice surgeon in this rare but dangerous area. Both Fran Farley, MD, and Haemish Crawford, FRACS, were the initial proponents of the idea and contributed chapters. Dan Hedequist, MD, already was involved in writing a book for our publisher, Springer, and put me in touch with Kris Spring in their New York office who has been beyond patient in waiting for a long overdue final draft. Dan, Suken, Burt, and I took on editorial responsibilities for this text, so the four of us are responsible for its content.

There are many others who have put up with the long process of writing, notably our families, of course. But I would also like to acknowledge the help of my colleagues at Arnold Palmer Children's Hospital in Orlando in disciplines apart from orthopedics, namely, neurosurgery, ENT, general surgery, and physiatry, who have written chapters which complete the scope of this book.

The final and most important thank you of all goes to my secretary and friend of 20 years, Mary Regling, BA, who has been the “den mother” of the PCSSG from its inception and the driving force behind getting this work published. Without her, the project would have foundered and failed.

Orlando, FL, USA Jonathan H. Phillips, MD

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Introduction

This book was written for a wide audience. Some of its readers will be familiar, or even expert, in the care of children with neck and cervical spine disorders. Others will be completely new to the subject. Though its emphasis is on the orthopedic and neurosurgical approach to children's cervical spine, there are chapters that are contributed by other disciplines. Thus, an ENT surgeon who may be called upon to perform an anterior trans-oral approach to the dens will be reassured by the account of this technique in Chapter 19. Chapter 21 focuses on non-spinal disorders which may present to physicians and others encountering children with neck problems in their clinics. Knowing what their significance is and which consultant to engage with in their management is important.

While it is unwise to try to be all things to all people, it is hoped that this is a reference that can be dipped into by the occasional reader looking for something specific and also be a comprehensive guide to the young surgeon embarking on a career which may include pediatric cervical spine surgery.

The area we cover is quite rare and quite dangerous for the unprepared. Yet with the changing demographics of childhood trauma and increasing survival of children with previously lethal syndromes, we are encountering these rare diagnoses with greater frequency.

The reader is encouraged to approach the text in a traditional fashion. We are all anxious to know "how to do it," but such enthusiasm must be tempered by acquiring the building blocks of "why." Thus, we start with basic science, and it cannot be overemphasized how important a thorough knowledge of the anatomy (both normal and abnormal), pathology, biomechanics, and radiology is to treating these rare disorders. The chapters on clinical assessment and presentation of the multitude of problems in this area follow. Only after these basic areas are covered do we embark on accounts of the surgical and nonsurgical management of the problems encountered.

Each chapter is written by experts in the area and can be taken as standalone treatises. It is hoped, however, that the whole will be greater than the sum of its parts.

Jonathan H. Phillips, MD
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Part I

Basic Medical Science

Embryology and Anatomy of the Child's Cervical Spine

1

Jonathan H. Phillips

Embryology and Definitions

The process of embryological development and maturation of the fetus can be described in various stages known as Carnegie stages. These refer to levels of development rather than gestational age or crown-rump length in millimeters. Though the three systems overlap, we will use the Carnegie stages as much as possible in this discussion.

The terms rostral and caudal and ventral and dorsal—while intuitive in embryology—are used less often in descriptive surgical anatomy, and the terms superior and inferior and anterior and posterior are used interchangeably in this chapter. In addition, descriptive names such as basiocciput, atlas, and axis are interchanged with skull, C1, and C2, which better describe the approach of the surgeon in the operating theater to ensure accurate surgical instrumentation at correct levels.

Metamerism is an important concept that relates to the general pattern of segmental repetition of similar structures in the developing embryo. It is this basic symmetrical template which is modified by localized gene expression to form region-specific structural changes which result in highly differentiated anatomical areas in vertebrates. Nowhere is this specialization more apparent than in the upper cervical spine of the human. The formation of the skull base and upper two cervical vertebrae is unique in the axial human skeleton and departs quite markedly from the lower cervical, thoracic, lumbar, and sacral morphology where there are more structural similarities than differences. We will see that the particular embryology of this rostral area of the spine has highly complex origins.

Segmentation describes a phenomenon of division of building blocks of tissues into repeating units and is similar in concept to metamerism. There is a further twist to the idea of segmentation in the human spine, however, because a process of resegmentation occurs during embryogenesis in which the caudal and rostral parts of adjacent segments fuse together to form the completed vertebrae. When this process is corrupted, the vertebrae are malformed. In the so-called hemimetameric shift, for instance, the process of resegmentation can fail unilaterally, resulting in the appearance of a hemivertebra and resulting in congenital scoliosis. This occurs most frequently in the thoracic spine, where coronal plane decompensation is an expected outcome for a fully segmented coronal hemivertebra, depending on the specific pattern of malformation. It occurs also in the cervical spine, both in the coronal and sagittal planes. Sagittal plane abnormalities are more common than coronal, the prototypical example of which is that seen in

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Klippel-Feil syndrome, a failure of segmentation rather than a hemimetameric shift, though this last can and does occur in the child's neck, resulting in cervical congenital scoliosis.

Somites, or more properly their derivatives, sclerotomes, are the building blocks of the spine. They appear in increasing numbers during embryogenesis, and the number of these segmental tissue blocks correlates with the anatomical staging of the embryo. Somites are just one part of the mesoderm layer of the three-layered early embryonic disc. This disc, a few days old, has an outer epidermal layer facing the amniotic cavity, a middle layer of mesoderm, and an endodermal layer facing the yolk sac. This pattern is apparent by about 3 weeks postfertilization. The mesoderm is itself divided into three parts, medial, intermediate, and lateral mesoderm. The most medial band is called the paraxial mesoderm and once again divides into three, this time from dorsal to ventral. The area nearest the dorsal surface is the dermatome, next the myotome, and further to the center of the embryo is the sclerotome. All of these areas are arrayed surrounding two structures which carry powerful molecular signaling properties—the notochord in the very center of the embryo and the neural tube which by now (stage 10 or about 4 weeks) has formed from the original neural plate and which lies right behind the notochord on its dorsal aspect. The notochord will regress quickly, but not before the ventral cells of the somitic mesoderm have spread toward this structure, which induces the formation of the sclerotome. The sclerotomal segments (and this tissue mass is segmented) will form the vertebrae, whereas the notochord, under the negating influence of the neural tube, remains in the mature human only as the nucleus pulposus of intervertebral discs and the alar and apical ligaments of the craniocervical junction. This segmented system develops in a rostral to caudal direction (Fig. 1.1).

Somite count increases from about one to four at age 20 days, first appearing at the head of the embryo, to 34–35 at age 30 days toward the tail. Ultimately, 44 pairs of somites occur and form the left and right half of the sclerotome. The other two parts of the somites go on to form muscle and skin. The remaining parts of mesodermal layer lateral to the somites form splanchnic structures. These include gut, vascular, and urological structures. Insult to the embryo at this stage can affect all these systems and explains the concomitant appearances in clinical practice of multi-system congenital formation failure. The best known example of this is VACTERL syndrome in which heart, gut, renal, and vertebral malformations coexist.

At about the 5- to 8-week period, or Carnegie stages 15–22, the emerging pattern of spinal formation is becoming evident. However, the contribution of somites to their sclerotomal structures is

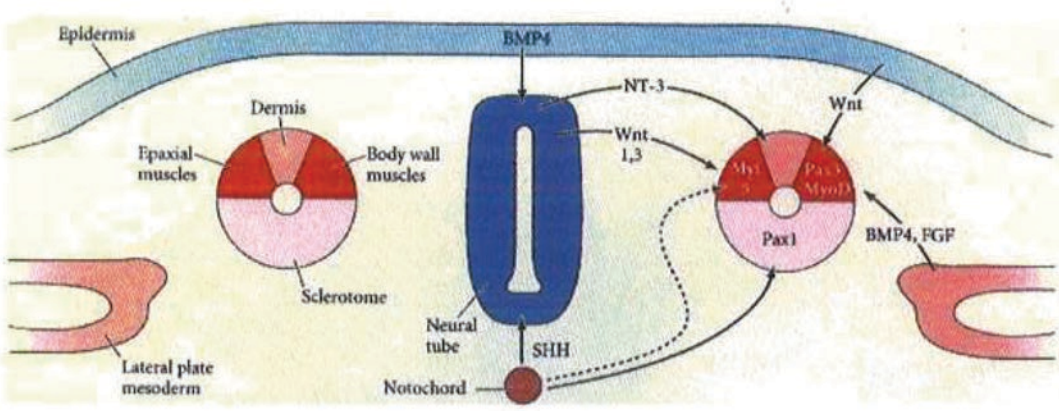

Fig. 1.1 The relationship and control of somatic mesoderm to the notochord and neural tube (Reproduced with permission from Gilbert [7]; © Sinauer Associates, Sunderland, MA)

highly complex at the cranial extent of the vertebral column. There are designated pairs of sclerotomes inasmuch as the upper four form the basiocciput, the next eight form the cervical vertebrae (of which there are only seven, but with eight spinal nerves), and the more caudal pattern (12 thoracic, five lumbar, and five sacral, variable coccygeal) is more easily understood based on the gross anatomy of the fully formed human skeleton. It is the complex variation from the typical pattern of vertebral development which gives rise to the unique shape and function of the atlas and axis. These two vertebrae share a common origin with the basiocciput, and as such should be considered as an embryological, anatomical, and functional unit very different from the subaxial spine. This unit is uniquely designed to transmit the termination of the brainstem and emerging spinal cord in a highly flexible protective tube that allows very roughly 50% of the total movement of the skull on the spinal column. The remaining cervical motion is distributed over the five lower cervical segments. All of these cervical vertebral segments except C7, however, carry the responsibility of transmitting the vertebral arteries, a function solely attributed to neck vertebrae. Once again the pattern of the vertebral arterial anatomy is radically different at the atlas and axis, and a thorough understanding of this arterial anatomy is fundamental to safe posterior surgical approaches to the upper cervical spine.

The relative somatic contributions to the spinal column are shown in Fig. 1.2. The upper four sclerotomes form the basiocciput but also borrow from somite five, which is a cervical one, thus the intimate relationship of the atlas to the skull base embryologically. Sclerotome five (a cervical one) forms both the posterior arch of the atlas and occipital condyles. The anterior arch of the atlas has an origin in the hypochordal bow which appears ventral to the notochord and undergoes chondrification and fusion with the posterior neural arch elements. There is a transient proatlas centrum which is dissolved. There is no vertebral body in C1 under normal circumstances. Teratogenic influences at this stage have been shown in mice. Interference with Hox genes by retinoic acid (most commonly used in the human for the treatment of acne) has been shown to

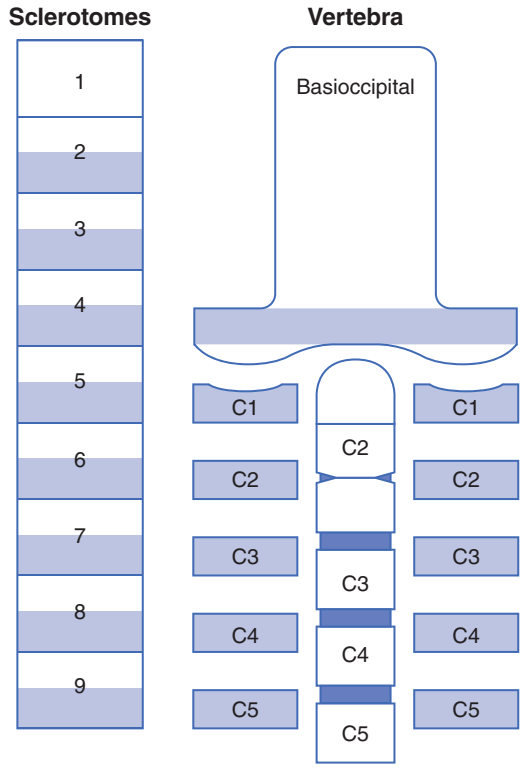

Fig. 1.2 Relative somatic contributions to the spinal columns (Redrawn with permission from Muller and O'Rahilly, © 1994 Wiley Publishing)

cause caudal or rostral homeotic transformations [1]. The Hox-4.2 gene expression can transform occipital bones into neural arches [2]. Finally, transgenic mice can be found to exhibit a third occipital condyle fusing the skull base to the dens [3], and rostral vertebral shifts have been seen after heat exposure.

Though murine and avian genetic models should be interpreted with caution in the human, it is easy to imagine that altered expression of these homeobox genes may be the basis for well-known malformations at the upper cervical spine such as assimilation of the atlas, which can occur posteriorly and anteriorly.

The formation of the axis is in many ways perhaps the most bizarre in the human axial skeleton. A review by Muller and O'Rahilly in 2003 explains the process well [4]. The fact that two, not one, sclerotomes form the posterior neural arch of C2 explains why it is so massive

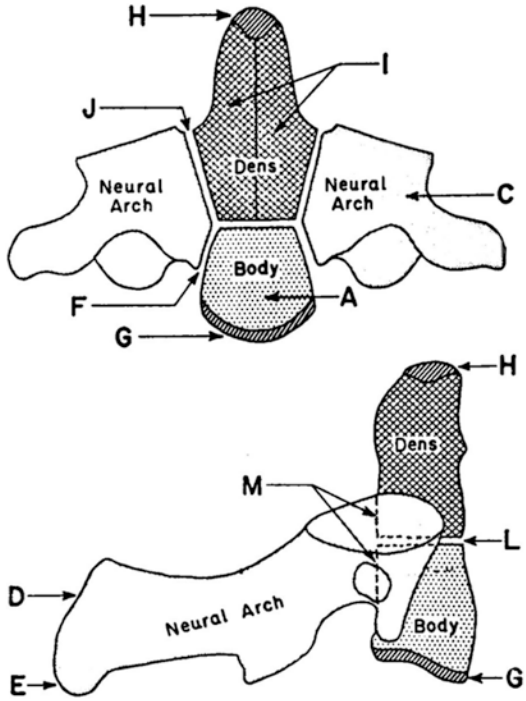

Fig. 1.3 Ossification centers of C2. *L* is the dentocentral synchondrosis; *J* is the neurocentral synchondrosis. There are two *I*s, two *C*s, and one *A*, totaling five primary centers. *G* and *H* are secondary centers of ossification (Reproduced with permission from Bailey [8]; © The Radiological Society of North America)

(and therefore ideally suited to the placement of translaminar screws during cervical instrumentation). It also helps us to understand the sometimes confusing radiological appearance of the synchondroses of C2 in the immature child (Fig. 1.3), an important goal to achieve since these areas are often misinterpreted as fractures. Perhaps mutations of gene expression in this area can also explain the retroflexed dens seen in Chiari malformation and congenital types of basilar invagination.

The third cervical vertebra and its subjacent levels exhibit the so-called typical cervical morphology. As noted above, this is still distinguishable from thoracic and lumbar vertebrae but approximates more closely to the general pattern of vertebral development.

There are three primary ossification centers at C1. The anterior center, derived from the hypochordal bow, fuses with the posterior/dorsal elements of the neural arches at the neurocentral synchondrosis. This junctional area fuses completely around age 7. The spinous process uniting the left and right neural arch growth centers unites at age 3. Thus, radiographically there appears to be a spina bifida occulta present in the toddler, though usually the laminae meet at a complete cartilaginous bridge. The same appearance may be present in more caudal vertebrae also.

At C2 there is predictably a much more complex arrangement consequent upon its development from three sclerotomes. Five ossification centers appear and there are also two secondary centers (the tip of the dens and the ring apophysis of the inferior/caudal aspect of the body of the axis). These centers result in two radiographically significant synchondroses (see Chap. 4). The dentocentral synchondrosis represents the fusion of two sclerotomes at the level of the future body of C2. However the fusion level, though less distinct, may also be apparent in the young child, most commonly on CT scan or MRI reformatted in the sagittal plane. As mentioned above, this may be a source of concern in the injured child as a potential fracture line [5]. The possible relationship of these synchondroses to later formation of an os odontoideum is discussed elsewhere (see Chap. 4). The neurocentral synchondrosis represents the junction of two sclerotomes anteriorly (ventrally) with the left and right neural arches derived dorsally from the same tissue. There are therefore two of these junctions left and right, and they are best seen in coronal imaging modalities. The growth centers of C1 and C2 are represented graphically in Figs. 1.3, 1.4, and 1.5. These describe the prototypical arrangements, but it must be emphasized that many anatomical variations can occur, which may be confusing on imaging studies of the young child. This point is well made by Karwacki and Schneider in their 2012 analysis of atlas and axis growth center variability based on 550 CT scans of children aged 0–17 years [6].

Growth centers are present at various stages in the human embryo. Initially seen as chondrification centers, they become ossified and visible

radiographically by birth and early childhood, though the adult pattern is not achieved until final vertebral physeal closure in the 20s.

Muscles of the Neck

The musculature of the neck has a complex arrangement predicated on the function of high mobility of the skull on the spinal column. The most superficial muscle posteriorly is the trapezius. This huge triangular muscle arises from the superior nuchal line of the skull all the way to the spinous process of T12. Its lateral attachment is on the spine of the scapula. Thus it is, strictly speaking, a muscle of the upper limb. The true deep muscles of the neck lie deep to trapezius and comprise five groups. The groups are splenius, erector spinae, transversospinal, interspinal, and intertransverse muscles.

Splenius covers the deeper muscles of the back of the neck and has capitis and cervicis divisions (Fig. 1.6).The splenius capitis and cervicis arise from the ligamentum nuchae and spinous processes C7 to T6. Cervicis inserts into the posterior tubercles of the upper two or three cervical vertebrae, and capitis has a more proximal insertion on the mastoid process and the lateral part of the superior nuchal line. Contraction of the splenius rotates the head toward the side of the muscle acting, and bilateral contraction extends the head and neck. Innervation is from dorsal rami of C2 to C6. Deep to it lie erector spinae and semispinalis. Anteriorly the sternocleidomastoid inserts more superficially to the capitis division at the mastoid. This last muscle arises from both clavicle and sternum and opposes splenius rotating the head to the opposite side of the muscle contracting, flexes, and laterally flexes the neck.

The erector spinae muscle group is represented in the neck as iliocostalis cervicis, longissimus cervicis, and spinalis cervicis and capitis—in other words, three subgroups (Fig. 1.7). Iliocostalis is lateral; spinalis medial and longissimus are in between the other two. The muscles lie in the costovertebral groove.

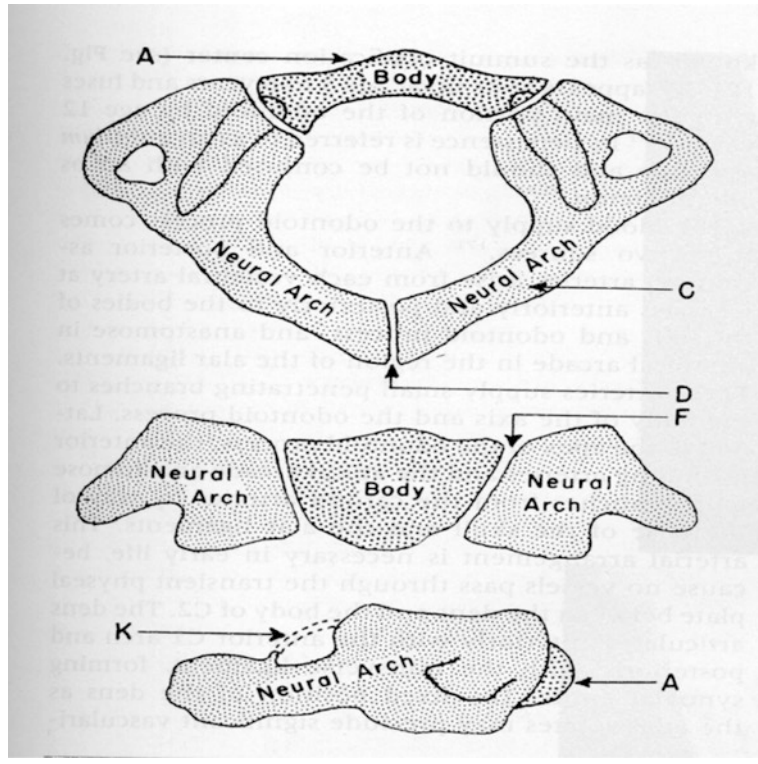

Fig. 1.4 Ossification centers of C1. Not unusually, the body center is bipartite and occasionally occurs in three or other multiple parts (Reproduced with permission from Bailey [8]; © The Radiological Society of North America)

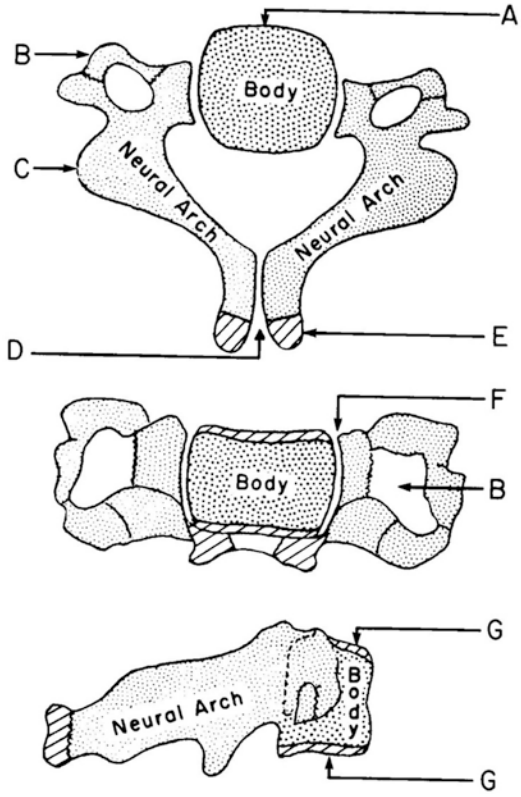

Fig. 1.5 Primary (stippled) and secondary (striped) ossification centers of the typical cervical vertebra. There are three. Note the ring apophyses (*G*) at superior and inferior parts of the body. These fuse late in life, sometimes in the early 20s (Reproduced with permission from Bailey [8]; © The Radiological Society of North America)

Iliocostalis cervicis arises form upper ribs and inserts onto transverse processes of the lower cervical vertebrae. Longissimus cervicis arises from the uppermost ribs and inserts into the C2 to C6 transverse processes. Spinalis cervicis is a variable muscle often not well defined. The erector spinae muscles laterally flex and extend the neck.

Of the transversospinal group, one muscle is important and forms the largest single muscle of the posterior neck. It is the semispinalis capitis and arises from transverse processes of the upper thoracic and seventh cervical vertebrae and the articular processes of C6 to C4 (Fig. 1.7). It inserts onto the undersurface of the skull base posteriorly and is a powerful extensor of the neck. Semispinalis cervicis is contiguous with its thoracis component and passes from thoracic transverse processes to spinous processes several levels higher, ultimately reaching the posterior axis.

Interspinal and intertransverse are small segmental muscles represented by such groups as multifidus and rotatores arising from transverse processes of adjacent vertebrae. All are segmentally innervated and perform functions of local stabilization and small rotations at segmental levels.

At the base of the skull lies a unique triangular arrangement of muscles which form the suboccipital triangle (Fig. 1.8). These suboccipital

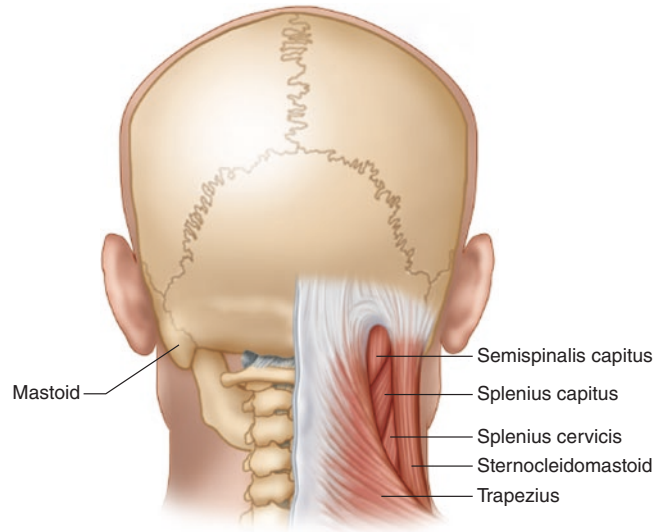

Fig. 1.6 Dorsal neck muscles, superficial layer

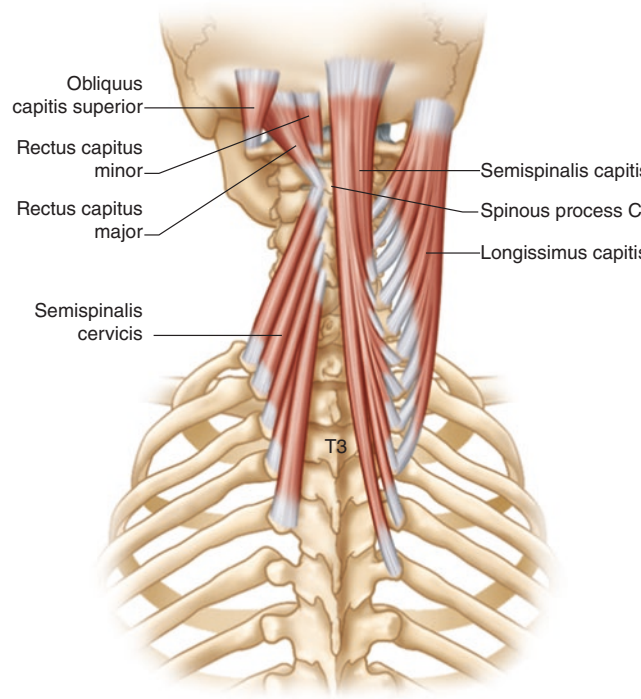

Fig. 1.7 Dorsal cervical musculature, deep layer

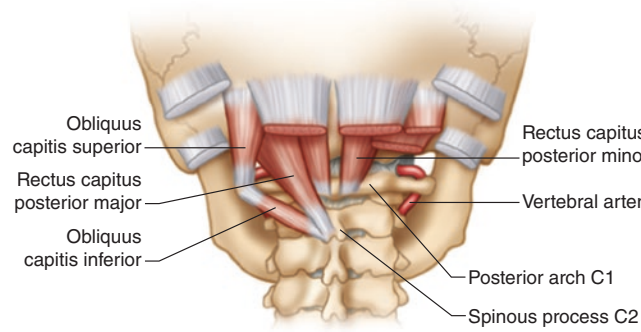

Fig. 1.8 Suboccipital musculature

muscles are part of the transversospinal group. The four muscles are rectus capitis posterior major and minor and superior and inferior oblique muscles of the head, as seen in Figs. 1.2 and 1.5. In the floor of this triangle lies the posterior atlanto-occipital membrane deep to which the vertebral artery passes of the arch of the atlas into the foramen magnum. The suboccipital (C1) and greater occipital nerve (C2) arise, respectively, above and below the posterior arch of the atlas. The C2 root overlies the lateral mass of the atlas and can obstruct the placement of a screw in this structure during posterior C1 instrumentation (Fig. 1.9). Occasionally the nerve must be sacrificed for this reason. A prolific venous plexus also lies in this region and can cause troublesome bleeding during C1 lateral mass instrumentation.

The vertebral artery arises as the first branch of the subclavian. It passes upward in the posterior part of the pyramidal space above the apex of the lung. It enters the cervical spine through the foramen transversarium of C6, not C7, and ascends to C2 where it passes backward then medially and then forward in a wide loop that

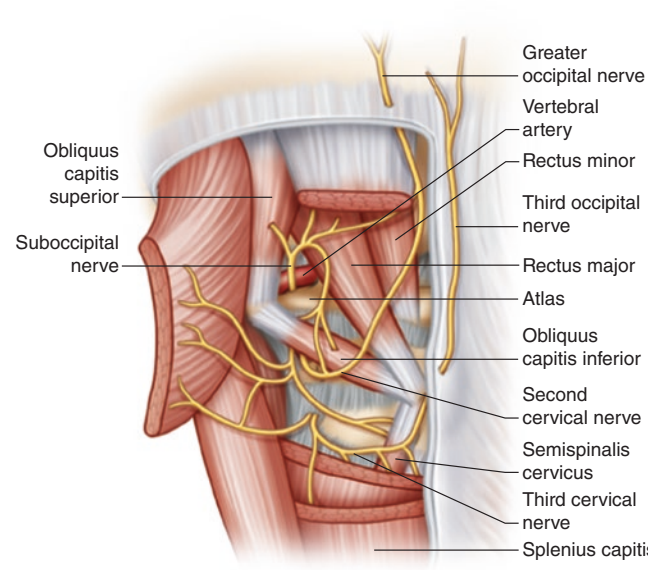

Fig. 1.9 Cervical nerves: first through third

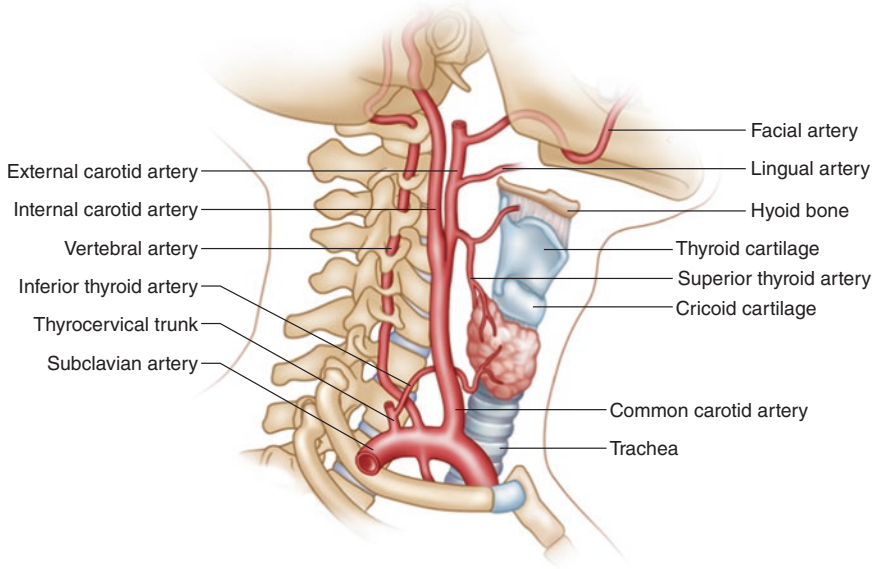

Fig. 1.10 Arteries of the anterior and lateral neck

allows for movement between atlas and axis (Fig. 1.10). As it passes anteriorly toward the foramen magnum, it leaves a groove on the superior surface of the atlas. It is highly vulnerable in this area to damage during surgical exposure of the occipital and atlantoaxial region. Its position lateral to the midline plane effectively limits the lateral dissection in surgery of this area. Accompanying the vertebral artery is the vertebral vein or more properly a plexus of veins which pass both inside and outside of the foramina transversaria. One branch exits at C6 and another at C7 transverse foramen, and both drain into the subclavian vein.

Anterior vascular anatomy dictates the surgical approach to the anterior cervical spine

(see Chap. 18). The carotid sheath contains the common carotid artery, the internal jugular vein, and the vagus nerve. It extends from the base of the skull to the aortic arch and is tightly opposed to the posterior surface of the sternocleidomastoid muscle above the sternoclavicular joint. At C3 level the common carotid bifurcates. The internal carotid has no branches in the neck and passes up into the skull through the carotid foramen just anterior to the jugular foramen, which itself contains the internal jugular vein and lies just deep to the external acoustic meatus. The carotid sheath and its contents, lying deep to the anterior border of sternocleidomastoid, form the posterior border of the anterior surgical approach to the mid cervical spine. Anteriorly the esophagus and trachea are retracted laterally to allow exposure of the anterior cervical vertebral bodies, and their discs thus form the anterior border of this exposure (see Chap. 14, 18).

Cervical Osteology

The atlas C1 is a gracile, almost circular, ring of bone with articular facets above for the occipital condyles and below for the axis (Fig. 1.11a, b). The neural arches are massively enlarged to form the lateral masses, which constitute the only substantial bony elements allowing surgical screw purchase. Their axes pass from posterior lateral to anterior medial. Above are the deeply concave kidney-shaped articular facets for the occipital condyles; below are the more circular facets for articulation with the axis. Lateral to the masses lies the foramen transversarium, formed from both neural and costal elements. Anteriorly on the arch of the atlas is a tubercle to which the anterior longitudinal ligament attaches. There is no centrum; the proatlas has dissolved. In addition, the anterior arch of the atlas is formed not from the centrum remnant as would be imagined, but rather from the hypochordal bow. This structure is important in cervical spine embryology, but exists elsewhere only as the ligamentous fascicle running deep to the anterior longitudinal ligament joining two rib heads. Thus the hypochordal bow of the atlas is the ossified ligament joining its two costal elements. It often shows failure of complete ossification in the child, as does the posterior arch of the atlas, which is more conventionally formed from neural arch elements. The course of the vertebral artery over the superior surface of the posterior arch has been described. One other anomaly is of interest. The articular elements of the upper two cervical vertebrae are in series with the tiny synovial uncovertebral joints on the lateral aspects of the subaxial cervical vertebrae and not their more massive and functional cervical articular facets aligning more posteriorly from C3 to C7. Thus the first and second cervical nerves send their anterior primary rami behind the joints and not in front as the lower vertebrae do. The resulting obstruction to surgical approaches to C1 has been mentioned.

The axis C2 has much more massive proportions than its cephalad neighbor (Fig. 1.12a, b). We have seen that it is derived from a larger number of sclerotomes and not only has retained, but also co-opted, a greater centrum contribution embryologically. Its characteristic features are the upward pointing dens which articulates with the posterior surface of the anterior arch of C1,

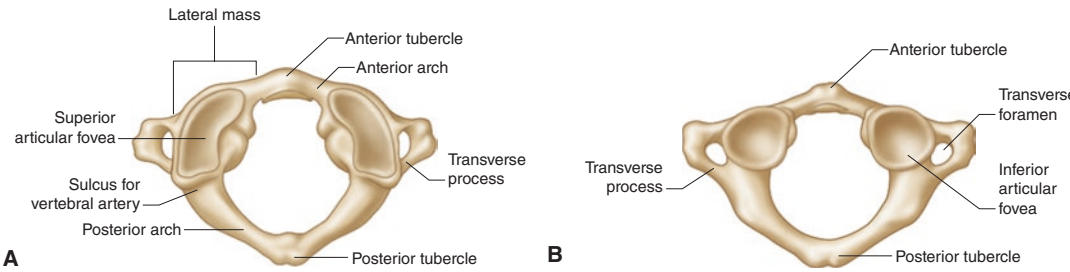

Fig. 1.11 (A, B) First cervical vertebra (atlas). (**a**) Cranial and (**b**) caudal

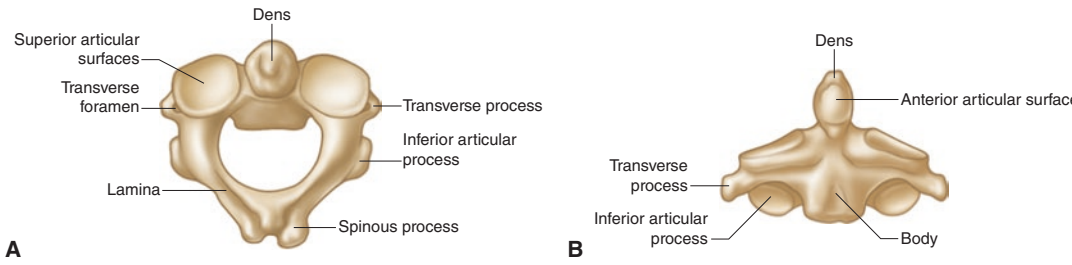

Fig. 1.12 (A, B) Second cervical vertebra (axis). (**a**) Cranial and (**b**) anterior

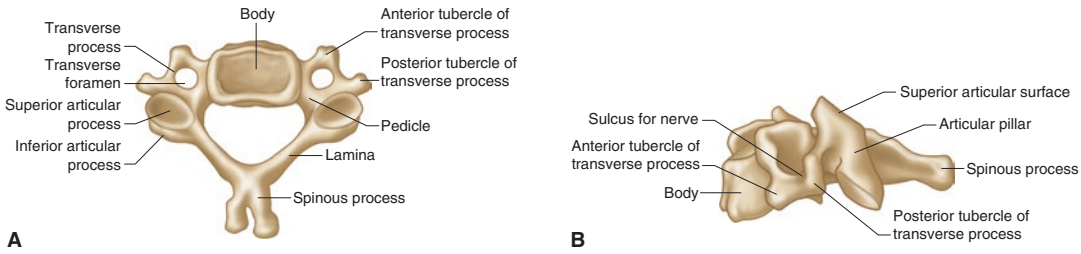

Fig. 1.13 (A, B) Typical cervical vertebrae showing a body, neural arch, spinous, and complex lateral processes

the large lateral masses, and the huge spinous process, which even in small children may be big enough to allow passage of translaminar screws. In contrast to the atlas, it may have discrete pedicles, though their size in children is variable and often does not allow for accommodation of a true pedicle screw. Alternative techniques of C1-C2 arthrodesis, such as a Magerl screw, are discussed in later chapters of this text. In addition, sublaminar wiring techniques have a long history in orthopedics and utilize the posterior arch of C1 and the lamina or spinous process of C2. Again, see later chapters.

The orientation of the articulations between occiput and atlas and atlas and axis allows for a very large range of motion, nodding at the former and rotation at the latter. Approximately 50% of rotation is lost by atlantoaxial arthrodesis, but the effect on atlanto-occipital fusion is less obvious because of the large flexion extension range of the subaxial spine.

From C3 to C7 the vertebral morphology is more typical and reproducible. Usually the costal elements are limited to the contribution to the foramen transversarium, but occasionally true cervical ribs appear at C7 or are represented by sometimes troublesome fibrous bands, causing thoracic outlet symptoms. In the case of properly formed cervical ribs, the brachial plexus may exhibit precession arising from C4 to C8 instead of C5 to T1.

Typical cervical vertebrae show a body, neural arch, spinous, and complex transverse processes which contain the vertebral artery and its veins (Fig. 1.13a, b). The lateral part of the body at the interface with the intervertebral disc shows an upward turn into the uncus, and it is here that tiny uncovertebral synovial joints exist. They limit lateral flexion and, described by Lushka, only exist in the cervical spine. Pedicles are much better formed in the typical cervical vertebrae and allow screw instrumentation. In addition, the greatly broadened lateral masses, which exhibit superior and inferior articulations that sandwich the bony masses, allow for strong screw fixation, again explained later in this text. The choice of lateral mass or medial pedicle trajectory is predicated on the position of the vertebral artery which lies directly anterior to the lateral masses and thus precludes a straight anteriorward approach in surgical fusions. However, purchase under the substantial laminae of C3 to C6 is available, if not as stable in fusion constructs. At C7 the pedicle is usually so well formed, even in children, that it has become the preferred location for spinal instrumentation at this level. Between C3 and

C6, careful analysis of the specific anatomy with advanced imaging is mandatory for safe placement of instrumentation, indeed all levels should be examined with CT preoperatively, and this idea is discussed in Chap. 4.

Ligamentous Support of the Child's Cervical Spine

In the lower cervical spine, a familiar pattern is found. The anterior longitudinal ligament supporting the anterior vertebral bodies, with intervertebral disc annulus and posterior longitudinal ligament at the posterior margin of discs and bodies, is very similar to the thoracic and lumbar pattern. At C2 and above a very different construct exists, uniquely suited to the previously mentioned function of allowing large degrees of motion between the head and neck. The cruciform ligament joins the posterior body of the dens to the base of the occiput at the anterior margin of the foramen magnum, bypassing the atlas. Its strong transverse ligament component captures the dens axis against the posterior part of the anterior arch of the atlas, where a synovial joint and significant bursa exists. The apical ligament is the continuation of the cruciform into the skull. Inferiorly the stem of the "cross" is adherent to the inferior posterior body of the axis below the dens. One additional and important connective tissue structure also adds stability to the skull-to-atlas-to-axis complex. The tectorial membrane passes from the posterior rim of the anterior lip of the foramen magnum to the posterior axis body. It is continuous with the posterior longitudinal ligament and blends with the dura on its deep or posterior surface. It can be imaged with MRI and if ruptured in this modality implies instability at the occipito-atlantal level (Figs. 1.14 and 1.15).

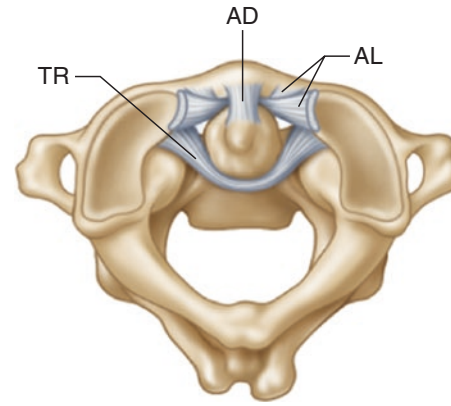

Fig. 1.15 Axial view showing transverse, alar, atlanto-dental ligaments

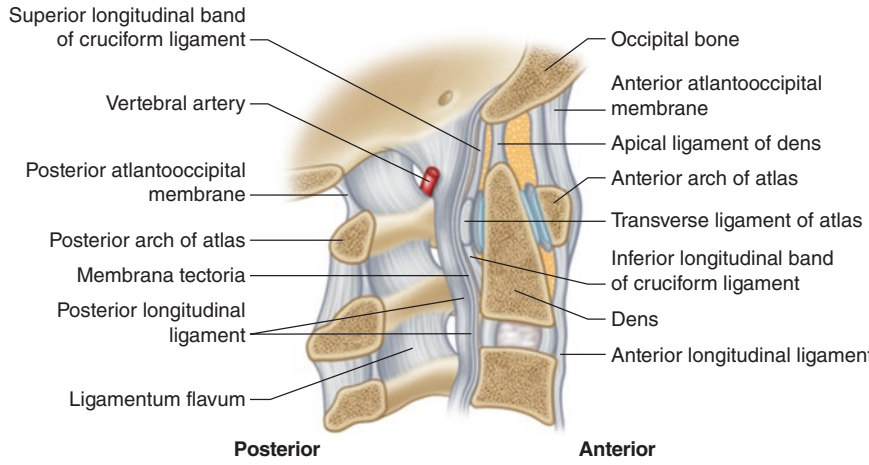

Fig. 1.14 Occipito-cervical level, showing ligaments and membranes, sagittal view

Summary

The development of the human neck shows unique aspects of specialization to fulfill the functions of great flexibility and range of motion not seen elsewhere in the spinal column. The uppermost two vertebrae show major departures from the pattern seen subaxially. A common embryological origin of the skull base and parts of the atlas and axis explains their unique shape and the fact that these parts function as a unit.

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Biomechanics of the Growing Cervical Spine

2

John Kemppainen and Burt Yaszay

Normal Cervical Spine Biomechanics

The cervical spine functions to provide motion of the head atop the axial skeleton and to protect the neural elements of the spine as they traverse the neck. The cervical spine can be divided into two distinct segments, the occipitocervical junction extending from the occiput to C2 and the subaxial cervical spine from C3 to C7, each having distinct anatomic and biomechanical features. Together, they provide several degrees of motion for the head on the axial skeleton, including flexion, extension, and lateral rotation and bending to the right and left; distraction and compression are theoretical and not desirable. The normal active range of motion of the child's cervical spine is slightly greater than that of an adult, with average values of 60° of flexion, 90° of extension, 45° of lateral bending in each direction, and 70° of axial rotation in each direction [1, 2]. Reasons for this include maturing osseous structures and increased ligamentous laxity in children, which will be discussed further throughout the chapter. Because of their distinct differences, the two regions of the cervical spine are discussed separately. A thorough discussion of the anatomy and embryology of the growing cervical spine can be found in Chap. 1, but here we discuss the osseous and ligamentous structures of the cervical spine as they relate to its kinematics and stability.

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Functional Anatomy and Normal Biomechanics

Occiput to C2

Osseous Anatomy

The craniocervical junction is made up of the base of the occiput, C1 (the atlas), and C2 (the axis) that function as a unit to control head movement on the subaxial spine. The primary motion of the occipitoatlantal joint is flexion and extension, with the atlantoaxial joint contributing primarily axial rotation. The underside of the occipital bone includes the foramen magnum, through which the spinal cord passes into the cervical spine. The anterior midline of the occiput is known as the basion, and the posterior margin is known as the opisthion. The transverse diameter of the foramen magnum is slightly less than the anterior posterior diameter. On the lateral side, just anterior to the midline are the occipital condyles, which are convex in the sagittal plane but oblique and rest on the concave lateral mass of

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C1, or the atlas, allowing for flexion and extension at the O–C1 articulation. In the coronal plane, the articulation is angled slightly medially, allowing for a small amount of lateral bending while resisting lateral translation.

The atlas is a ring-shaped bone, lacking the vertebral body and spinous process of other vertebrae and acting as a dished washer between the occiput and C2. The two thick lateral masses (which are the morphologic corollary to the transverse processes in the sub-cervical spine), act as the articulating surfaces of C1. The superior surface articulates with the occipital condyles as previously mentioned and the inferior surface with C2. The inferior facets are relatively flat, with a slight convexity and lateral tilt to allow axial rotation between C1 and C2. This rotation occurs around the odontoid process, which is a cephalad projection of the body of C2 and is a significant stabilizer of the C1–2 articulation, as discussed below. The body of C2 is larger than that of C1 and is connected on each side by a neural arch that includes an inferior and superior facet. The superior facets sit lateral and just posterior to the dens, are slightly concave, and receive the convex inferior facets of C1. The lateral tilt limits lateral translation while allowing significant amounts of rotation. The inferior facet of C2 sits posteriorly on the neural arch and has an orientation similar to the subaxial articular facets of the cervical spine.

Ligamentous Anatomy

The limited articular and osseous constraints of the craniocervical junction place significant importance on the ligamentous structures to provide stability while still allowing for a very extensive range of motion. The tectorial membrane is a cranial continuation of the posterior longitudinal ligament that travels posterior to the body of C2 and anchors to the base of the skull at the anterior rim of the foramen magnum (see Chap. 1). It controls extension by becoming taught when the head is extended and limits flexion at C1/2 when it is tightened as the skull tilts anteriorly on C1 [3]. A recent investigation argued that the tectorial membrane may act less as a true stabilizing structure and more as a reinforcement to prevent impingement of the odontoid on the neural elements, which secondarily stabilizes the occipitocervical junction [4]. The alar ligaments extend from the dorsolateral surface of the dens to the medial aspect of the occipital condyles, each one limiting lateral rotation to contralateral side. They also act as a check ligament to limit the amount of axial rotation between C1 and C2, further discussed below. The cruciate ligament consists of a transverse and ascending/descending portion. The transverse ligament is the thickest and most important portion and connects between the two condyles of C1, stabilizing the dens between them. The ascending/descending portion extends from the anterior edge of the foramen magnum to the body of C2. The apical ligament, the atlantodental ligament, and the anterior and posterior atlantooccipital membrane are biomechanically insignificant [5, 6].

Kinematics

The occipitocervical complex provides approximately 40–50% of flexion and extension and 60% of axial rotation of the cervical spine. Much less lateral bending is allowed at these two articulations, most of which comes from the lower cervical spine. The primary motion between the occiput and C1 is flexion and extension, contributing approximately 25° total [7]. The cup-shaped articulation limits rotation with reports ranging from 0° to 8° [7–9] and lateral bending ranging from 2° to 8° [7, 10, 11].

Axial rotation is the primary motion of the C1–C2 articulation, with up to 65° of motion in one direction in adults [5]. The joint also contributes approximately 20° of flexion and extension and, similar to the occipitoatlantal joint, contributes only approximately 5° of lateral bending [7, 10]. In their attempts to further understand lateral axial rotatory subluxation in children, Pang and Li have performed a thorough CT evaluation of the kinematics of the C1–2 articulation in normal children. In the early phase of lateral rotation, C1 moves with the head, while C2 is left stationary, a phase which they termed the single motion phase. At approximately 23° of rotation, the alar ligaments begin to tighten and C2 rotates with C1, but at a different rate, termed the double

motion phase. In other words, C1 continues to rotate with the head at a faster rate than C2, and the resulting angle between C1 and C2 continues to increase. Beyond 65° of C1 rotation, C1 and C2 move in exact unison (the unison-motion phase), and the remainder of head rotation originates from the subaxial spine. Interestingly, they noted that the relationship between C1 and C2 when returning to neutral follows a nearly identical path and inferred that ligamentous tension is not the only mechanism coupling the motion between C1 and C2 (if this were the case, C1 would have to pass the C2 neutral point and place tension on the contralateral alar ligament to pull C2 back to neutral). The authors surmised that the identical reverse rotation is likely a result of dragging between irregular bony surfaces at the C1–C2 joint.

Determinants of Stability

As discussed above, the occiput to C1 articulation is comprised of cup-shaped facet joints with a slight medial angulation that provides stability against lateral translation, but very little in the AP direction. Overall, the O–C1 joint is likely inherently very unstable, relying on the ligamentous attachments of the occiput and C1 to C2 and muscle strength for stability. What little intrinsic stability that does exist derives primarily from very weak ligamentous structures including the joint capsules of the occiput to C1 joints and the anterior and posterior atlantooccipital membranes.

At C1–2, the articulations are covered by loose articular capsules that allow the freedom of movement discussed above, lending little stability from the facet joints. Therefore, the odontoid process provides much of the stability of the C1–2 articulation, with its relationship to the anterior ring of C1 and the surrounding soft tissue structures, primarily the transverse portion of the cruciate ligament (see Chap. 10).

Consequently, underdevelopment, injury, or dysmorphism of the odontoid process can lead to substantial atlantoaxial instability. For example, Morquio syndrome results in hypoplasia of the odontoid process (Fig. 2.1a–d). The resulting instability at the atlantoaxial articulation can lead to myelopathy of sometimes rapid progression, quadriparesis, and a risk of sudden death from respiratory failure [2]. The treatment of myelopathy in these patients consists of fusion of the occipital cervical junction, and some have advocated a prophylactic approach as soon as instability is noted, given the detrimental effects and the rapid progression of myelopathy that can be seen in these patients [12, 13]. However, the optimal timing of surgery in children with asymptomatic instability due to odontoid hypoplasia is debated.

Likewise, insufficiency of the transverse ligament and other stabilizing ligamentous structures from trauma or other pathologic entities can produce instability of the craniocervical junction. In the pediatric population, this commonly occurs in conditions with associated connective tissue abnormalities. Down syndrome, for example, results in generalized ligamentous laxity throughout the musculoskeletal system, and laxity in the stabilizing ligamentous structures of the craniocervical junction can lead to hypermobility, often in the presence of normal bony anatomy (Fig. 2.2a, b). For this reason, patients with Down syndrome should be monitored for craniocervical instability, and, if present, careful evaluation for myelopathy is required.

Aside from pathologic entities that may result in upper cervical instability, the craniocervical junction is vulnerable in normal young children compared to adults. Reasons for this include smaller flatter occipital condyles, shallow morphology of the articulation with C1, large head size, and an odontoid synchondrosis that may be susceptible to fracture. These differences are discussed further later in this chapter.

Subaxial Cervical Spine C3–C7

Osseous Anatomy

Below C2, the vertebral morphology becomes similar to the rest of the spine, with an anterior vertebral body and neural arch consisting of a pair of pedicles and laminae. Anteriorly, the vertebral segments articulate through an intervertebral disc and posteriorly though the articular cartilage of the facet joints. The shape of the vertebral body accommodates the motion seen in the

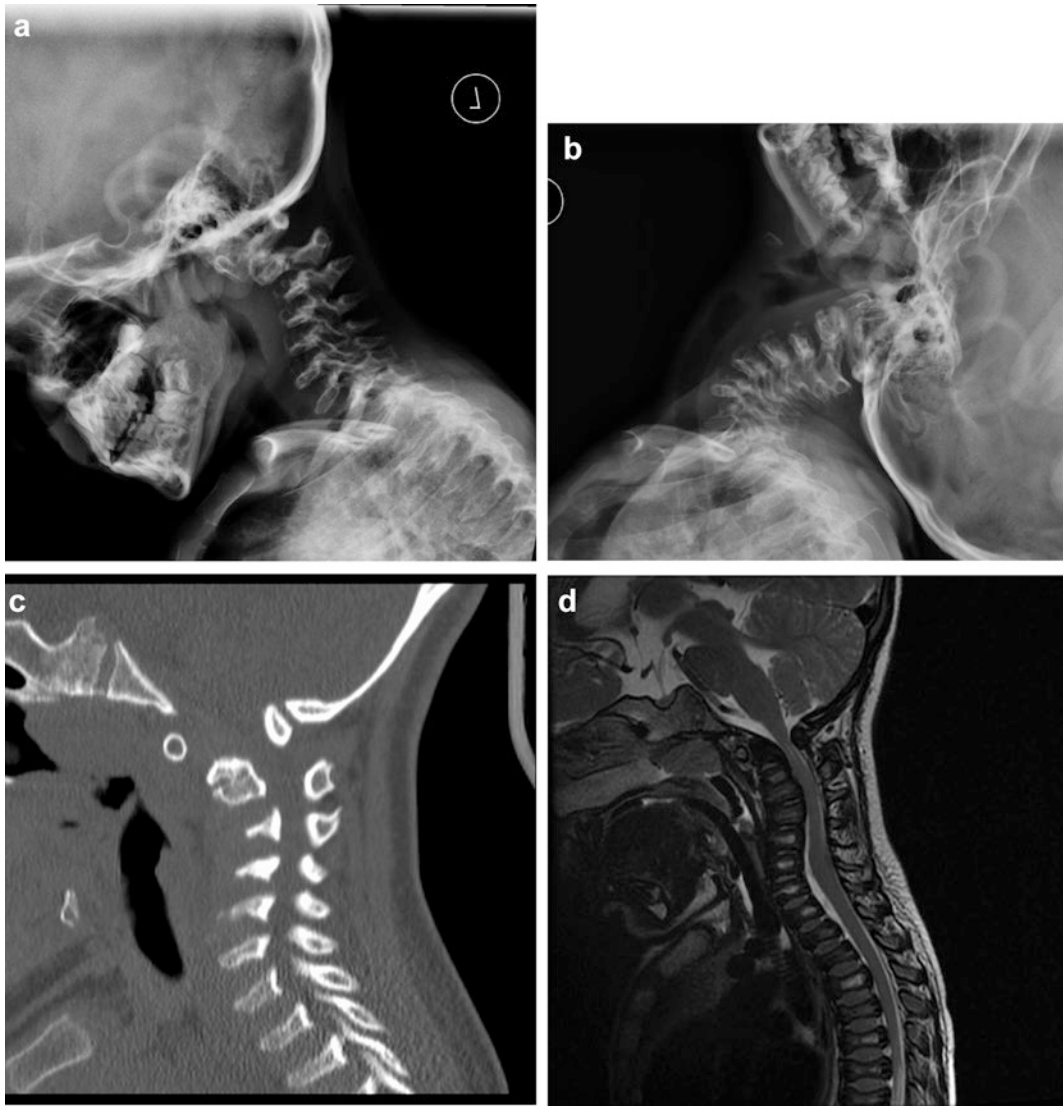

Fig. 2.1 (**a**–**d**) Morquio odontoid hypoplasia that has resulted in instability. Dynamic MRI demonstrates cervical cord compression

subaxial cervical spine, predominately lateral bending and flexion and extension. Inferiorly, the endplate is convex in the coronal plane being received by a concave superior endplate of the level below, allowing for lateral bending through the disc. In the sagittal plane, the inferior endplate is slightly concave, allowing for flexion and extension. Posterolaterally, the uncinate process extends from the superior endplate to articulate with the inferior endplate of the level above and is an important feature in coupled motion of the lower cervical spine [14]. The uncinate process is not well developed in children younger than 10 years old [15, 16], which contributes to the increased mobility of the cervical spine in young children. Posteriorly, the facet joints are angled to accommodate flexion and extension, with that angulation increasing with skeletal maturity to an eventual slope of approximately 45° in adulthood. Particularly in children, the upper cervical facet joints are oriented more horizontally than caudal levels in older necks and contribute to

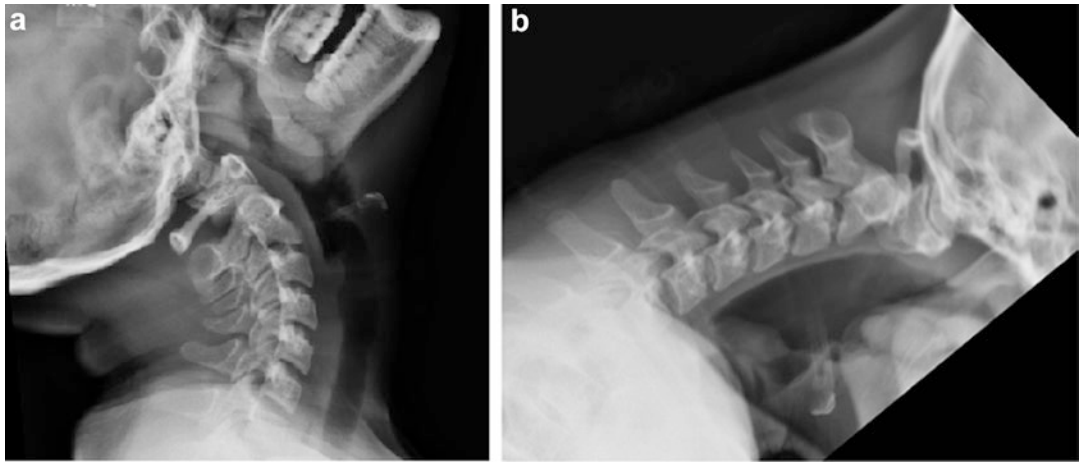

Fig. 2.2 (**a**, **b**) Nine-year-old with atlantoaxial instability secondary to Down syndrome

pseudosubluxation seen in some normal children, which is most commonly seen at C2/3 and discussed later in this chapter [15], and in chapter 4.

Ligamentous Anatomy

Like the upper cervical spine, the stability of the subaxial spine is highly dependent on the integrity of its ligamentous structures, particularly in tension. The anterior longitudinal ligament (ALL) attaches securely to the anterior aspect of the vertebral bodies and loosely to the intervertebral disc. On the posterior portion of the vertebral body, the posterior longitudinal ligament (PLL) is secured firmly to the disc and loosely to the vertebral body. Both have similar material properties. Posteriorly, the ligamentum flavum passes between the laminae of each level. It is pretensioned in vivo to limit bunching of the ligament when the spine is extended [17]. The interspinous ligaments and facet joint capsules also contribute to the stability of the posterior elements in flexion. The ALL and PLL have been shown to have a higher ultimate strength than the posterior ligamentous structures, but the elongation to failure is higher posteriorly, allowing for greater distraction and displacement posteriorly before failure [18]. As in the upper cervical spine, the relative hyperlaxity of immature soft tissues contributes to increased mobility of the subaxial spine in children [19]. Intervertebral discs also contribute to stability of the spine and have a high water content at birth, which gradually decreases with age and may also contribute to the overall hypermobility of the spine in children.

Kinematics

The most prominent motion in the lower cervical spine is flexion and extension, with the middle segments having the most significant contribution [20]. At any given segment, the axial rotation of the lower cervical spine is much smaller than in the upper cervical spine, but together, the lower cervical segments contribute approximately 30° of axial rotation of the head on the axial skeleton [9, 21, 22], which is coupled with other movements. Coupling is the consistent association of one motion with another in a different plane, such that one does not easily occur without the other. In the lower cervical spine, axial rotation is associated with lateral bending and, to a lesser degree, flexion and extension.

Determinants of Stability

As in the craniocervical junction, the stability of the subaxial spine is dependent on both bony and ligamentous factors. In compression, the vertebral bodies, the intervertebral discs, and the facet joints stabilize the spine. In tension, stability relies on the ligamentous structures, including the annulus fibrosis, anterior longitudinal ligament, posterior longitudinal ligament, facet capsules, and the interspinous ligaments. The relative

contribution to stability in all planes of motion of the discoligamentous structures has recently been evaluated in vitro. Richter et al. sequentially sectioned ligamentous structures beginning anteriorly with the ALL, anterior portion of the disc, interspinous ligaments, ligamentum flavum, and, finally, the facet capsules [23]. Flexion/extension showed a significant increase in range of motion (ROM) with each sequential dissection. In axial rotation, no significant difference was noted until the facet capsules were sectioned, and in lateral bending the soft tissue sectioning did not result in a significant increase in ROM.

Of course, rarely is a physiologic load purely compression or tension in both the anterior and posterior spine; for example, axial pressure in the flexed cervical spine creates compression anteriorly and tension on the posterior structures. The result is a complex interaction between the bony and ligamentous anatomy needed to provide stability to the cervical spine. Significant injury to, or malformation of, either the bony or ligamentous structures can result in pathologic instability that is important to recognize.

Unique Biomechanical Characteristics of the Immature Cervical Spine

As alluded to in the preceding sections, there are substantial differences between the immature and mature cervical spine, which can have a profound effect on the normal biomechanics and response to loads applied to the spine. Here we will discuss some of the unique features of the growing spine, with a focus on those relevant to clinical practice.

Growth

An obvious difference between the adult and pediatric spine is remaining growth in children, which is biomechanically important for two reasons: (1) forces exerted by growth centers affect the morphology of the spine (see Chap. 1), and (2) the growth centers act as a weak points that are susceptible to fracture.

As expansion occurs at growth centers, forces are exerted on the spine that must be well balanced for normal development. These forces are small, but imbalance applied over long periods of time can result in structural changes that can have a profound clinical effect on the growing child. Although more applicable to the sub-cervical spine, a classic example of this is the crankshaft phenomenon seen after posterior-only fusion in very young children [24, 25]. As a result of the fusion, the posterior forces of growth are halted, leaving unbalanced growth between the anterior and posterior columns. As the anterior growth continues, the forces overcome the restraints of posterior fusion, and the spine rotates around the fusion, resulting in the characteristic deformity. As expected, younger patients are at higher risk for this phenomenon as they have more growth remaining and the potential for greater forces anteriorly. Thus early fusion should be approached cautiously.

In the cervical spine, the effect of asymmetric growth can be seen in postlaminectomy kyphosis. The incidence of deformity following laminectomy for disease not usually related to spine deformity is well documented, and skeletal immaturity appears to be one of the greatest risk factors for its development [26–28]. Yasuoka, et al. showed that the postlaminectomy kyphosis is much more likely in growing patients, showing an incidence of 46% in children younger than 15 years old, 6% in patients 15–24 years old, and in no patients older than 24. In the younger age group, all patients who received cervical laminectomies developed kyphosis [26]. In the normal lordotic cervical spine, the weight-bearing axis lies posterior to the vertebral bodies [29]. With the loss of the posterior tether from laminectomy, kyphosis places the weight-bearing axis along the anterior column of the cervical spine. Additionally, decreased lordosis of the cervical spine is a common finding in children [16, 30], putting them at risk for kyphotic positioning. As kyphosis begins, compression anteriorly increases, resulting in decreased anterior growth

based on the Heuter-Volkman principle of decreased growth with compression across a growth center. Additionally, the decreased posterior compression allows for less inhibited growth posteriorly, resulting in a self-perpetuating deformity that can lead to neurologic compromise if not recognized and treated.

The forces of spinal growth can also be used as a clinical advantage when treating disorders and injuries of the spine. Long utilized in the lower extremities for the correction of angular deformity [31], guided growth techniques are being explored in the spine. Tethering inhibits growth of the convexity of a scoliosis, allowing for relative increased longitudinal growth of the concave side of the deformity and resulting in a gradual correction of the scoliosis [32–34]. In the cervical spine, the effect of asymmetric growth can be harnessed in treating traumatic injuries of the posterior elements of the spine. Hyperemia after a cervical injury results in increased growth of the growth centers throughout the spine. The resulting overgrowth of the anterior structures may counterbalance the potential risk for progressive kyphosis and limit the need for anterior stabilization in children with a significant amount of growth remaining. In the extreme case, this anterior growth associated with a posterior tether can result in hyperlordosis of the cervical spine (Fig. 2.3a, b).

Additionally, like physes elsewhere in the growing skeleton, the syncondroses and endplates of the spine are potential weak points and focus for fracture (Fig. 2.4). The most common example are odontoid fractures, which occur through the base of the odontoid at the physeal plate [35]. Traumatic injuries are further discussed in chapter 6.

Structural Differences

Even in normal individuals, certain anatomic and physiologic differences between the child and adult result in hypermobility of the cervical spine. These include the following differences, as articulated by Brockmeyer [36]:

1. Increased ligamentous laxity [37].
2. Disk and annulus are elastic and allow for more distraction.
3. Facet joints are horizontally oriented and shallow, especially at higher levels [15].
4. Bony vertebral bodies are wedge shaped and allow for increased flexion.
5. Uncinated processes are not present in children under 10 years old—resulting in increased lateral and rotational movements between the bodies.
6. Growth centers are a weak point for fracture.
7. Large head size relative to the thorax.
8. Weak neuromuscular control.

These anatomic and physiologic differences combine to allow greater displacement of the immature cervical spine under normal physiologic loads. Clinically, this is appreciated as pseudosubluxation—anterior translation of one vertebral level on another in normal individuals (See also chapter 4). Pseudosubluxation occurs most commonly between C2 and C3, likely due to the more horizontal orientation of the facets at more cranial cervical levels [15]. Cattell and Filtzer in 1965 performed flexion and extension lateral X-rays on 160 normal children and found 46% of children younger than 8 years old had pseudosubluxation at C2/3 of 3 mm or more. It is important for physicians evaluating pediatric spines to recognize that this is a normal entity and to be able to differentiate it from pathologic subluxation [30].

Swischuk developed the posterior cervical line to aid in this evaluation [38]. A line connecting the anterior aspect of the spinous processes of C1 and C3 is drawn on the lateral radiograph. If pseudosubluxation of C2 on C3 exists, this line will lie on the anterior aspect of the C2 spinous process. It is important to note that this line only applies if there is subluxation of C2 on C3, and if no subluxation is present, the spinous process will commonly be posterior to the posterior cervical line. If subluxation is present and the posterior cervical line misses the spinous process of C2 by 2 mm or greater, true instability should be suspected. Keep in mind that if the posterior

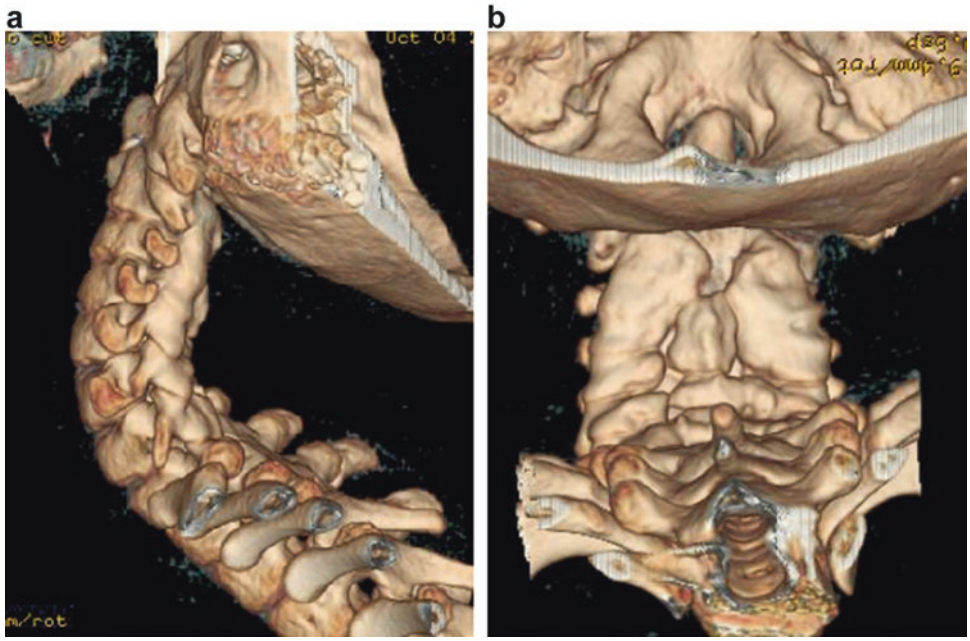

Fig. 2.3 (**a**, **b**) Cervical hyperlordosis secondary to anterior growth with an associated posterior tether. The posterior tether is secondary to autofusion of the posterior elements following surgery at the age of 2

cervical line is anterior to the lamina of C2 and there is anterior displacement of the vertebral body of C2 on C3, a fracture of the neural arches of C2 should be suspected (see Chap. 4).

Fortunately, cervical spine fractures and dislocations are uncommon in children, with only approximately 2% of all cervical fractures/dislocations occurring in children younger than 15 years old [39]. When cervical fractures do occur in children, the injury pattern is predictable based on the patient's age. Children younger than 8 years old have a tendency for injury of the craniocervical junction, most likely explained by biomechanical differences between young children and the older child or adult [40, 41]. These differences include the horizontal orientation of the facets and relative laxity of the cervical ligaments, as discussed previously. Additionally, young children have a larger and heavier head relative to the size of their thorax and undeveloped neuromuscular control, combining for a "bowling ball" effect above the hypermobile cervical spine. These features combine to produce a cranially positioned fulcrum that progresses caudally with age from C2–3 in infants to C3–4 by 5 years old, C4–5 at 10 years old, and C5–6 by 15 years old. This places the craniocervical junction at risk in young children, leading to an increased incidence of upper cervical injuries in younger children. After age 8, the child's cervical spine begins to mature into a more adult-like

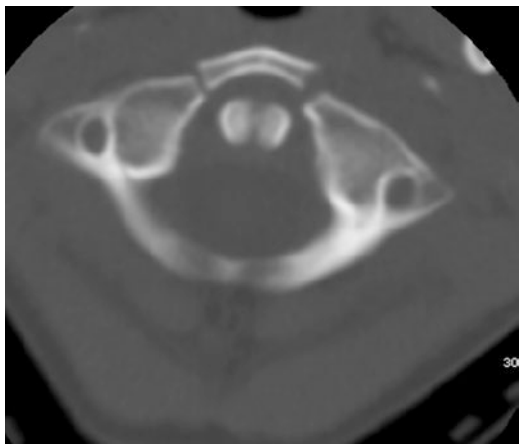

Fig. 2.4 Axial CT image of a C1 fracture through the neurocentral synchondroses of a 6-year-old boy, illustrating the potential for injury at the growth centers of the spine

morphology, and injury patterns begin to resemble that of adults by the age of 12, with a higher incidence of osseous injury and more injuries in the lower cervical spine [37, 39, 42, 43].

The effect of the child's large head size relative to his or her thorax is well taught among a wide range of specialties treating traumatic injuries in children, and immobilization on a flat backboard will result in flexion of the cervical spine if it is not corrected with the placement of an appropriately sized cervical collar [44]. Although the clinical effect of this flexion is not clear, recommendations for immobilization include the use of a cut out backboard to allow the head to fall into a more posterior position or elevating the torso for the same effect. This may be particularly important in children younger than 8 years old, who tend to have a larger head-to-torso ratio than older children [45]. However, other authors have demonstrated that deviations from a neutral C-spine is common in all age groups, and appropriate vigilance should be maintained to avoid unnecessary flexion or extension of the cervical spine and when choosing the appropriately sized cervical collar when immobilizing children all ages [46].

Perhaps the most clinically relevant biomechanical difference between children and adults is the increased elasticity of the ligamentous structures in the immature spine. As discussed, anatomic differences such as horizontally oriented facets, large head size, and immature neuromuscular control place the cervical spine in children at risk. When combined with the ligamentous elasticity, the immature cervical spine can undergo significant displacement without complete osseous or ligamentous disruption. Cadaveric studies have shown that the child's cervical spine can undergo displacements beyond the limits of the allowable strain for neurologic elements, without disruption [19]. Pang and Willberger et al. evaluated the tensile properties of immature cervical spine specimens and found that the tensile stiffness to failure and load to failure increased nonlinearly with age. The displacement at failure decreased nonlinearly from birth to adulthood, indicating that significant amount of displacement can occur in young children without structural failure, potentially exceeding the allowable strain of the enclosed neural elements. In cases of trauma, these factors can combine to produce significant neurologic injury without structural damage to the cervical spine, known as a spinal cord injury without radiographic abnormality, or SCIWORA. In young children, SCIWORA is more likely in the cervical spine and tends to be more severe than in older children, where it commonly occurs at lower levels [47, 48]. SCIWORA is further discussed in Chap. 6 of this text.

Biomechanics of Cervical Spine Instrumentation

Surgical technique and implant design for instrumentation of the cervical spine has advanced greatly in recent years, but remains a technically demanding procedure, especially in children, where osseous anatomy may be underdeveloped or morphologically altered. A thorough understanding of the relative biomechanics of the available instrumentation construct is important for a surgeon treating cervical spine pathology. The following discussion addresses clinically relevant biomechanical considerations when choosing instrumentation for the pediatric cervical spine. As has been true for discussions throughout this chapter, the majority of data presented comes from adult specimens or patients. Clinical indications and surgical techniques for the instrumentation discussed are reserved for other chapters within this text.

Occipitocervical Instrumentation

The anatomy and biomechanics of the occipitocervical region present unique challenges for achieving adequate fixation, especially in the pediatric or dysmorphic spine. Instrumentation must resist the significant motion in the sagittal and rotational planes and the weight of the head, as well as the lever arm created by the angle of the occiput and the cervical spine.

Foerster [49] is attributed with the first description of surgical fusion of the occipitocervical junction, using fibular strut graft, and most

early fusion techniques involved bone graft onlay with immobilization in a halo vest. Since then, techniques have evolved to include wire fixation, hook constructs, and plate and rod constructs with screw fixation to the spine. As instrumentation has advanced, the ability for internal fixation to adequately immobilize the fused segments has improved, and the need for prolonged halo immobilization has diminished.

Summary

For those who treat the pediatric spine, it is important to understand the biomechanical properties of the cervical spine. They influence the pathology and injuries pediatric patients may sustain, as well as the treatments that are applied. Unfortunately, when compared to the adult cervical spine, there is limited literature evaluating the biomechanics of the pediatric cervical spine. Even within the pediatric population, age-related changes in the anatomy and tissue strength influence the biomechanical properties. This ultimately prevents a universal principal for managing pediatric cervical spines that can be applied to all pediatric patients.

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Pathology of the Child's Cervical Spine and Its Clinical Implications

3

Ehsan Saadat, Daniel J. Hedequist, and Patrick Wright

Connective Tissue Disorders

Immature cervical vertebrae and their surrounding joints provide little in the way of bony support in the neck of a child; thus, the integrity of the pediatric cervical spine relies heavily on the soft tissues that surround it. This chapter discusses conditions that are associated with cervical spine pathology due to weak connective tissues, including syndromes of Marfan, Loeys-Dietz, Ehlers-Danlos, and Down. Later we touch on bone dysplasias, inflammatory conditions and infections.

Marfan Syndrome

Introduction

In 1896, the French pediatrician Antoine Marfan described a 5-year-old girl with arms and legs that were long and slender and were associated with spiderlike fingers [1]. Continued observation of this patient and others in the early 1900s led to the classic Marfan triad of arachnodactyly, ocular, and cardiac abnormalities [2]. Currently, this syndrome is the most commonly seen connective tissue disorder, and it is one of a few orthopedic conditions associated with tall stature.

Classification and Pathology

Marfan syndrome is caused by mutations in the fibrillin gene, localized to the long arm of chromosome 15 (15q21.1) [3, 4]. The mutant gene product has a dominant negative effect, which means production inactivates the normal fibrillin function [5]. Seventy percent of cases are inherited in an autosomal dominant pattern, while 30% of cases are sporadic [6].

Fibrillin is a protein found in the microfibrils of cartilage extracellular matrix, periosteum, ligaments, and others. Primarily, fibrillin helps maintain a normal soft tissue response to cyclic stress [7]. A defect in this protein leads to laxity in joints and subsequent subluxation or dislocation, as well as weakness in other tissues, such as arterial walls. Another function of fibrillin is to maintain a normal extracellular matrix and bind growth factors. Loss of this function may cause increased release of transforming growth factor β and lead to long limbs and tall stature [8].

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Demographics and Presentation

Patients usually present to an orthopedic surgeon in their second decade with a complaint of joint laxity or scoliosis. The characteristics noted on the physical exam include tall stature, arachnodactyly (long, slender digits), and dolichostenomelia (long, slender limbs). Pectus deformities, such as pectus carinatum or excavatum, are often seen, and up to 70% of patients will have some evidence of scoliosis [9, 10]. Known ocular findings are myopia and superior dislocation of the lens, and cardiovascular manifestations include aortic regurgitation, dilatation, arterial aneurysms, and mitral valve prolapse. Recognizing that your patient may have an underlying diagnosis of Marfan syndrome warrants a referral to a geneticist for a complete workup, as some of these manifestations may be life-threatening [11].

Diagnosis and Imaging

The diagnosis of Marfan syndrome is typically based on clinical findings and a family history. Major diagnostic criteria include a positive family history, cardiovascular and ocular involvement, and the presence of dural ectasia. There are other findings commonly seen in patients with Marfan syndrome which can alert the physician to the possibility of the diagnosis but are not pathognomonic of the disease. They include an arm span greater than height, an upper segment over lower segment ratio of 0.85 or less, a positive Steinberg sign (thumb extends past the ulnar border of the hand when adducted), and thumb and index finger overlapping when wrapped around the opposite wrist [12, 13].

Imaging studies are useful in the diagnosis of Marfan syndrome [14]. Dural ectasia remains a major criteria, and signs of it can be seen on plain radiographs. These include a transverse process-to-vertebral width ratio of greater than or equal to 2.25 at L3, a sagittal diameter of greater than or equal to 13.5 mm at L5, and an interpedicular distance greater than or equal to 36 mm at L5. A better way to confirm dural ectasia, however, requires CT or MRI imaging. It is defined as a dural volume of greater than or equal to 7 cm^3 below the inferior L5 endplate. Unfortunately, this has only been defined in adults, as norms in growing children are not known.

Scoliosis should be evaluated and imaged similar to patients with an idiopathic type, and patients symptomatic of neck instability should undergo a series of images that include flexion and extension laterals of the cervical spine. Cervical spine X-rays, however, are not routinely recommended for screening the asymptomatic patient due to the low rate of clinically significant findings.

Treatment

Before any skeletal surgery is undertaken on a patient with Marfan syndrome, a full genetics workup should confirm the diagnosis. Type I homocystinuria has a phenotype similar to Marfan syndrome but is associated with coagulopathies that could complicate surgical intervention. Also, evaluation of the cardiovascular system and appropriate treatment of any life-threatening manifestations, although rare in children, should be performed [15].

A number of cervical spine abnormalities have been reported in the setting of Marfan syndrome and are more common than in the general population. These include focal kyphosis, atlantoaxial hypermobility, subaxial hypermobility [16], Chiari malformation [17], basilar invagination, and syringomyelia. Despite this, neurologic compromise in these patients is rare [18]. There are case reports of atlantoaxial instability leading to rotatory subluxation after surgery [19], postoperative kyphosis due to halo traction [20], and even sudden death due to atlantoaxial dislocation in a young man performing forestry work [21]. These case examples indicate that, although catastrophic injury is rare, the sustained injury is worse due to ligamentous hypermobility than that which would have been sustained by a person without Marfan syndrome.

Surgical treatment in Marfan syndrome is associated with higher complication rates than in the general population [22]. Scoliosis surgery is associated with increased rates of pseudarthrosis, postoperative kyphosis, infection, blood loss, dural tears, instrumentation failure, and coronal

decompensation. Managing patient expectations and careful preoperative planning is essential.

Loeys-Dietz Syndrome

Introduction

Named for Bart Loeys and Harry Dietz, their syndrome is a recently described inherited collection of findings which have a phenotype similar to Marfan syndrome [23]. It affects the cardiovascular system, craniofacial appearance, neurocognitive abilities, and skeletal development. Some key differences to Marfan syndrome are widely spaced eyes, a split uvula, characteristic skin findings, and a similar height compared to an unaffected parent.

Classification and Pathology

Currently, there are two different types of Loeys-Dietz syndrome. The first type has craniofacial findings of widely spaced eyes, bifid uvula, and craniosynostosis. The second has systemic manifestations without significant craniofacial findings. Four genes have been found to contribute to this syndrome: $TGFBR_1$ (~20%), $TGFBR_2$ (~70%), $SMAD_3$ (~5%), and $TGFB_2$ (~1%) [24]. This disorder is inherited in an autosomal dominant fashion. Twenty-five percent of parents inherited the disorder and 75% have de novo mutations.

Both Loeys-Dietz and Marfan syndromes are associated with increased transforming growth factor beta activity in the body. In the soft tissues, this leads to an impaired response to cyclic stress and subsequent vascular aneurysms, joint laxity, and pregnancy-related complications. Arachnodactyly, pectus deformities, scoliosis, and cervical spine instability are skeletal manifestations also found in patients with Loeys-Dietz syndrome. The natural history is characterized by aggressive arterial aneurysms and average life expectancy of just over 26 years [25].

Demographics and Presentation

There are four major groups of clinical findings in Loeys-Dietz syndrome: vascular, skeletal, cutaneous, and craniofacial. Vascular anomalies include aortic dilatation or dissection and other cerebral, thoracic, or abdominal aneurysms. Pectus excavatum or carinatum, scoliosis (20%), dural ectasia, joint laxity, arachnodactyly, clubfoot, and cervical spine instability (7%) are the noted skeletal deformities. The skin demonstrates a velvety texture, is translucent, and will heal from trauma with dystrophic scars. Those with Loeys-Dietz type II will be spared from the following, but patients with type I will have widely spaced eyes, a bifid uvula or cleft palate, and craniosynostosis (19%). Retinal detachment and blue sclera may also be noted on exam. Patients will present to an orthopedic surgeon as infants if a clubfoot is noted. Otherwise, the presentation is similar to Marfan syndrome, with complaints of pectus deformities, scoliosis, joint laxity, and cervical spine hypermobility.

Diagnosis and Imaging

The diagnosis of Loeys-Dietz syndrome is suspected based on family history and clinical findings. Characteristic craniofacial deformities, velvety translucent skin, and the lack of an abnormally tall height can help differentiate this from Marfan syndrome. Definitive diagnosis is based on molecular genetic testing for mutations in the $TGFBR_1$, $TGFBR_2$, $SMAD_3$, or $TGFB_2$ genes.

Multiple evaluations are recommended to establish the extent of involvement in these patients. A genetics consult should include surveillance for life-threatening aneurysms, as well as craniofacial and eye exams [26, 27]. Standing radiographs of the spine for scoliosis and flexion/extension views of the cervical spine for potential instability are indicated in symptomatic patients.

Treatment

Flexion and extension cervical spine imaging is recommended before undergoing surgical procedures, due to high rate of cervical instability. Any symptomatic cervical instability should be stabilized to prevent damage to the spinal cord. Spinal deformity correction and fusion is indicated for progressive scoliotic curves and should be approached in a similar fashion to Marfan syndrome.

Down Syndrome

Introduction

The previous conditions associated with connective tissue disorders and hyperlaxity of joints are all disorders of single genes. Trisomy 21, or Down syndrome, is a chromosomal disorder and by far the most common one of orthopedic importance. Chromosomal disorders are more common than single-gene defects, and it is estimated that 1/660 newborns has Down syndrome. This condition is associated with underlying collagen defects leading to joint hypermobility and cervical spine instabilities. The most common form of Down syndrome is a true Trisomy 21 or three inherited copies of chromosome 21.

Demographics and Presentation

Down syndrome is likely the most recognizable genetic abnormality due to its frequency and characteristic appearance [28]. Potential skeletal manifestations are many, and joint hypermobility with ligamentous laxity is common. The characteristic pelvis deformity demonstrates flattened iliac wings and flat acetabulae. Hip dysplasia, slipped capital femoral epiphysis (possibly due to hypothyroidism), valgus knees, and pes planus are other lower extremity deformities associated with Down syndrome [29].

Multiple types of spine deformity are noted with this syndrome, especially with respect to the cervical spine [30]. Cervical spine hypermobility is very common and can occur at the atlanto-occipital joint, the atlantoaxial joint, or the subaxial spine. The atlanto-occipital joint in Down syndrome patients has less condylar curvature than age-matched controls; thus, there is very little intrinsic bony stability [31]. Without bony stability, the joint relies heavily on the intrinsic properties of the apical and alar ligaments, the tectorial membrane, and the joint capsules, which are lax in children with Trisomy 21. This same ligamentous laxity is also noted at the atlantoaxial joint, where an atlanto-dens interval (ADI) of 4–5 mm can be normal in this population. In addition to ligamentous laxity in the spine, bony deformity is also noted, to include os odontoideum, persistent dentocentral synchondrosis of C2, laminal defects at C1, and C1 hypoplasia causing canal narrowing [32]. Despite the potential pathology of the cervical spine in this setting, neurologic compromise is quite rare. Presenting symptoms of myelopathy in Down syndrome are torticollis, gait disturbance, exercise intolerance, loss of bladder control, hyperreflexia, clonus, or the presence of primitive reflexes [33]. Other, but less common, spine findings include scoliosis and spondylolisthesis.

Diagnosis and Imaging

The clinical findings associated with Down syndrome are the mainstay of diagnosis. Cytogenetic analysis can be performed for confirmation, and an AP pelvis radiograph can be helpful to demonstrate the characteristic findings of the flattened iliac wings. If not previously performed, referral for cardiac and endocrine workup is indicated.

There exists some controversy about whether the cervical spine should be screened in patients with Down syndrome [34]. The most important distinction to be made is the difference between asymptomatic hypermobility and true instability that can cause neurologic compromise. A good history and physical exam is likely the best way to determine the difference. Regardless, an exam and dynamic cervical images are required for participation in the Special Olympics. An ADI of less than 4.5 mm is automatic clearance to participate. An ADI greater than 4.5 mm but less than 10 mm is restricted from participation unless parental consent is given, and two physicians other than the treating physician allow participation. The Special Olympics makes no recommendations about the occipitocervical junction and does not require Power's ratio to be recorded. Patients with radiographically significant cervical instability should be further imaged with flexion and extension CT or dynamic MRI to fully evaluate the space available for the cord and the extent of cord compression [30].

Treatment

Any patient with Down syndrome, cervical hypermobility, and signs of neurologic compromise should undergo stabilization of the cervical spine [35]. Reported radiographic indications for fusion

have included an ADI greater than 10 mm, a Power's ratio greater than 1, and a space available for the cord of 13 mm or less. It should be mentioned here that instability at multiple levels (e.g., occipitocervical, atlantoaxial, and subaxial) can create an additive effect on the underlying neural elements and lead to chronic cord injury. MRI can be helpful to identify signs of Chiari malformation, basilar invagination, syringomyelia, or signal change in the cord due to instability. Asymptomatic patients with a lesser degree of instability should definitely avoid collision sports.

The surgeon should remember the comorbid conditions with this syndrome: cardiac disease, higher rate of infection, and endocrine abnormalities that may impair healing. Despite this, with improving medical technology, the life expectancy of those with Down syndrome is increasing, and attention to preserving quality of life is important.

Metabolic Conditions

Many skeletal deformities in children can be attributed to a specific genetic mutation. Each mutation alters the protein produced and thus alters the metabolism within the cells of the body. This can lead to a buildup of waste metabolites, unchecked cellular growth, or prevention of growth altogether. This section addresses conditions where altered metabolism within the body can lead to cervical spine disorders.

Mucopolysaccharidoses

Introduction

The mucopolysaccharidoses (MPS) are a group of genetic disorders leading to a common phenotype of short stature, coarse facies, and stiff joints [36]. Over a dozen types have been described, each with a specific enzyme defect that leads to excretion of mucopolysaccharide metabolites in the urine. For most of the known types, the deficient enzyme and the resultant glycosaminoglycan that builds up over time have been described. As these glycosaminoglycans increase in number, they are stored around the body, leading to pathology in the brain, vicera, and joints. Thus, mental retardation, skeletal deformities, and soft tissue problems are common with MPS.

Classification and Pathology

These disorders are classified based on their clinical similarities and enzymatic defect, if known. The most common types are Hurler syndrome (MPS I) and Morquio syndrome (MPS IV). MPS I is characterized by progressive mental retardation, hepatosplenomegaly, and dwarfism. The enzymatic defect is α-L-iduronidase, which leads to a buildup of heparin sulfate (HS) and dermatan sulfate (DS) [37]. MPS IV is characterized by normal intelligence, joint hypermobility, and significant skeletal deformities. The phenotype is caused by a buildup of keratin sulfate (KS) and, in one subtype, chondroitin sulfate (CS). There have been two enzymes identified in Morquio syndrome, N-acetyl galactosamine-6-sulfate sulfatase and β-D-galactosidase [38].

These disorders are a part of a larger group of lysosomal storage disorders. Specific to the mucopolysaccharidoses, the genetic defects are in lysosomal enzymes that degrade the sulfated glycosaminoglycans and lead to a buildup of HS, DS, KS, and CS in the lysozymes. The lysozymes are unable to rid the body of these sugars, which are subsequently deposited around the body, causing progressive, unrelenting pathology.

Demographics and Presentation

The overall incidence of mucopolysaccharidoses is approximately 1/25,000 live births [39]. All are inherited in an autosomal recessive manner, except for MPS II (Hunter syndrome), which follows X-linked recessive inheritance. Although children are born with the enzymatic defect, clinical disease is not evident at birth because the glycosaminoglycans must accumulate over time. In severe cases, the diagnosis may be made in infancy, but in cases where the defective enzyme retains some ability to process the sulfated sugars, clinical symptoms may not present until the teenage years.

An underlying MPS should be considered in a short-statured patient with varying levels of mental retardation, dental changes, and hepatosplenomegaly. Presenting complaints to the orthopedic surgeon include carpal tunnel syndrome, small joint stiffness, large joint laxity, joint pain, limb deformity, or spine deformity [40, 41]. Hip dysplasia and early arthritis are also commonly found but late in the disease [42, 43]. Myelopathy may be present due to cervical compression or instability. Glycosaminoglycan deposition occurs in the dura, ligamentum flavum, and surrounding ligaments. Congenital vertebral anomalies have also been described, including small thickened posterior elements, odontoid dysplasia/hypoplasia, and hyperkyphosis leading to a gibbus deformity [44]. Hydrocephalus, pseudotumor cerebri, and empty sella syndrome have also been described and can present with upper motor neuron dysfunction.

Diagnosis and Imaging

If an underlying diagnosis of one of the mucopolysaccharidoses is suspected, the patient should be referred to a geneticist for definitive blood tests and subtyping. The orthopedic surgeon should consider a skeletal survey to better characterize any skeletal deformities. Dysostosis multiplex is a collection of radiographic findings seen in patients with MPS that are caused by defective endochondral and membranous growth throughout the body [45]. These findings include small ovoid vertebral bodies that result in a kyphotic or gibbus deformity, large shallow acetabuli, coxa valga, genu valgum, broad clavicles and ribs, rounding of the iliac wings, a hypoplastic distal ulna with ulnar curvature to the distal radius, and short, thin metacarpals that appear bullet shaped.

Dedicated cervical spine radiographs, including lateral projections with flexion and extension, are always indicated due to an association with odontoid hypoplasia and instability [46]. A patient with significant myelopathy, however, may have relatively normal-appearing radiographs. This is due to the deposition of sugars into the soft tissues and compression of the spinal cord by thickening of the posterior longitudinal ligament, the ligamentum flavum, and the dura mater itself. The clinician must rely on a careful history and physical exam, as a complaint of decreased physical endurance could be the first sign of myelopathy [47]. This can be confirmed by static MRI, although flexion and extension MRI may best define the extent of cord compromise [48]. Quadriparesis due to cervical cord compression is possibly the most common cause of premature death in MPS patients; thus, a comprehensive neurologic exam every 6 months has been recommended [49, 50].

Treatment

The ultimate treatment for MPS would be early total body enzyme replacement. Bone marrow transplant has been used with some success [51]. It has demonstrated short-term improvement in non-osseous manifestations in the most severe forms of MPS. The long-term results, however, have not been as promising. Musculoskeletal deformities persist despite successful transplantation and will require treatment as the patient ages. Intravenous targeted enzyme replacement may prove beneficial in the future, although the enzymes cannot cross the blood-brain barrier and do not slow the neurocognitive decline [52, 53]. In addition, the cost of treatment is massive.

Orthopedic interventions for patients with MPS are directed at correcting deformities to improve function and decrease or eliminate pain. Due to craniofacial deformities, difficult airways should be expected for any surgical intervention [54]. Spine deformities should be corrected early to prevent spinal cord compromise [55, 56]. Sudden death has been described in MPS patients, and atlantoaxial instability is probably the most common cause. Unfortunately, achieving a stable fusion and predictable long-term result can be difficult [57, 58]. Rib graft should be considered when performing a cervical spine fusion, and long-term surveillance is needed due to high rate of junctional instabilities in the future [59].

Neurofibromatosis

Introduction

Neurofibromatosis (NF) is the most common single-gene disorder in humans and classically identified by smooth-edged café au lait spots, neurofibromas, skeletal anomalies, and autosomal dominant inheritance. Over 2 million people worldwide have some type of NF. There are several types, made different by their clinical presentations and genetic causes. Despite their clinical differences, they are all caused by the lack of a tumor suppressor gene product, thus allowing an oncogene pathway to proceed unchecked.

Classification and Pathology

There are four types of neurofibromatosis, each with its own diagnostic criteria. The first and most common is NF1, originally known as von Recklinghausen disease, after Friedrich Daniel von Recklinghausen, who was the first to characterize the disorder in 1882. The genetic defect in NF1 is located on chromosome 17 and causes a reduced amount of neurofibromin to be produced, which is a negative regulator of the Ras oncogene signal transduction pathway. Thus, this pathway is not turned "off" and neurofibromas are produced [60].

NF2 is also called central neurofibromatosis and is classically associated with bilateral vestibular nerve schwannomas. The genetic defect is found on chromosome 22 and causes reduced amount of schwannomin, or Merlin, protein to be produced [61]. Schwannomin is also a tumor suppressor protein, and the result in NF2 is schwannomas, meningiomas, gliomas, or neurofibromas.

The two least common forms of NF are NF3 and NF4. Patients with NF3 present with findings consistent with NF1, but acoustic neuromas are also present (as with NF2). This type has the highest rate of cervical spine deformity, with a relatively rapid and fatal course due to central nervous system tumors. Patients with NF4 are also very similar in presentation to NF1, but Lisch nodules (hamartomas of the iris), which are seen in every adult patient with NF1, are not present in patients with NF4 [62].

Demographics and Presentation

Neurofibromatosis affects 1/3000 newborns and is inherited in an autosomal dominant pattern with 100% penetrance. Sporadic cases, however, do occur and may be associated with advanced paternal age. Orthopedic manifestations are most commonly seen in patients with NF1, and cervical spine abnormalities are more commonly seen with NF3.

Spine deformities are also common in NF and generally present later in life. Although idiopathic-type curves are seen, the hallmark of spine deformity in NF is the dystrophic, short-segment single curves with associated rib penciling [63]. There is a significant risk of paralysis with these curves, especially with short-segment kyphosis [64].

Diagnosis and Imaging

Although genetic mutations for NF1 and NF2 have been identified, the diagnosis remains based on clinical findings. Characteristic skin markings of NF1 are multiple café au lait spots with smooth edges or a "coast of California" appearance. Axillary and inguinal freckling may also be present. Lisch nodules will be present in 50% of children and 100% of adults with NF1; thus, referral for a comprehensive eye exam is warranted.

If an underlying diagnosis of NF is considered, spine X-rays may identify evidence of scoliosis, and if abnormalities are noted in the cervical spine, flexion/extension lateral X-rays would help identify any instability. MRI and CT scan should be obtained for preoperative workup to identify neurofibromas, dural abnormalities, and vertebral deformities [65].

Treatment

Cervical spine compression can be caused by neurofibromas, ganglioneuromas, or ependymomas, necessitating decompression and fusion [66–69]. Dystrophic cervical kyphosis is particularly challenging in this population [70–73]. Continued follow-up is required after spinal surgery in patients with NF, as late complications are common. In a series of 22 cervical spine surgeries for compression or instability, 14 patients required a second surgery, and 3 required even

more surgery to achieve a stable outcome [67]. Patients should be educated about the importance of follow-up and likelihood of subsequent surgeries. Junctional problems adjacent to fusions are almost to be expected and are more a manifestation of the disease than a complication of surgery.

Skeletal Dysplasias

Introduction

Genetic disorders that lead to skeletal deformity present a challenge to the orthopedic surgeon because of their variety. The 2010 revision of the nosology and classification of genetic skeletal disorders is a list of 456 conditions in 40 categories [74]. The underlying genetic mutations are known in 316 of these disorders and include 226 individual genes. Not all of these disorders, however, are associated with significant cervical spine or foramen magnum pathology. Those of interest to the cervical spine surgeon include achondroplasia, spondyloepiphyseal dysplasia congenita (SED), spondyloepimetaphyseal dysplasia (SEMD), spondylomegaepiphyseal dysplasia (SMED), diastrophic dysplasia (DD), Larsen syndrome (LS), pseudoachondroplasia, campomelic dysplasia (CD), chondrodysplasia punctata (CDP), and osteogenesis imperfecta (OI).

Classification, Pathology, and Presentation

The aforementioned skeletal dysplasias are classified according to the ninth International Skeletal Dysplasia Society meeting and include those groups and disorders of interest to the spine surgeon.

Achondroplasia is caused by mutation in a gene on the short arm of chromosome 4 that codes for fibroblast growth factor receptor-3 (FGFR3) [75]. It is inherited in an autosomal dominant manner, but spontaneous mutations are frequent. FGFR3 limits endochondral ossification in the proliferative zone of the physis, and the "gain-of-function" mutation in achondroplasia causes rhizomelic shortening of the limbs and resultant dwarfism [76]. Arguably the most recognizable form of dwarfism, patients have fontal bone bossing, relatively normal trunk length, short humeri and femurs, flexion contractures at the elbows, bowed legs, and a "trident" hand. Associated spinal pathology includes symptomatic kyphosis, foramen magnum and spinal stenosis, and os odontoideum [77]. Apart from foramen magnum stenosis however, intrinsic cervical problems are rare in achondroplasia and it is the thoracic and lumbar spine which are affected in the main.

Spondyloepiphyseal dysplasia congenita (SED) is another form of dwarfism with severe deformities of the spine and the epiphyses of the long bones. It is caused by mutations in the *COL2A1* gene; thus, it is a type 2 collagen disorder [78]. The trunk, neck, and limbs are shortened, but hand size is usually average. Kyphosis and scoliosis are common and present early in life. The cervical spine may have findings of os odontoideum, hypoplastic odontoid, hypoplasia of C1, and surrounding ligamentous instability [79]. Atlantoaxial instability is common and often requires fusion. Spondyloepimetaphyseal dysplasia (SEMD) is very similar to SED and is in the same group of collagen disorders. It is associated with multiple joint dislocations, spine deformities, and short stature. An elongated odontoid and small atlas has been described [80]. Spondylomegaepiphyseal dysplasia (SMED) is also caused by a collagen deficiency but differs in that type 11 collagen is involved. Short stature and spine deformity are present, and radiographs demonstrate large "balloon-like" epiphyses. Severe cervical kyphosis and instability associated with cervical cord compromise are common in this disorder [81].

Diastrophic dysplasia (DD) is a member of the sulfation disorder group and is specifically caused by a defect in the sulfate transporter gene (DTDST gene) located on chromosome 5 [82]. The mutation leads to the production of proteoglycans without the proper compliment of sulfate, thus causing buildup in the cartilage cells [83]. The characteristic findings are clubbed feet, multiple joint contractures, swelling of the pinnae of the ears, and abducted thumbs called

"hitchhiker" thumbs. Scoliosis is common in DD and is either an idiopathic type or a dystrophic type with sharp curves and kyphosis. DD is also associated with a congenital kyphosis of the mid-cervical spine and hypoplastic posterior vertebral elements of the cervical and thoracic spines [84]. This kyphosis can improve with time, and observation is justified.

The filamin group of skeletal dysplasias includes Larsen syndrome (LS). LS is caused by mutations in the filamin B gene on chromosome 3, which is a cytoskeletal protein that facilitates communication between the cell wall and the internal structure. The phenotype is usually obvious at birth, with dislocated hips, knees, and elbows and concomitant clubfeet. Cleft palate, heart defects, and tracheomalacia are also common [85]. Cervical spine kyphosis and instability are the most concerning spine deformities associated with LS, due to their progressive nature and eventual cord compromise [86]. Its near cousin, atelosteogenesis type III, affects the same allele and is clinically almost indistinguishable.

Pseudoachondroplasia (PSACH) is a part of the multiple epiphyseal dysplasia group, because they all involve mutations in the cartilage oligomeric matrix protein (COMP) [87]. This group is a spectrum of disorders, with PSACH occupying the more severe end. This form of dwarfism is not readily identifiable at birth, because COMP must accumulate in the cartilage cells over time to cause the phenotype. Intelligence is unaffected by COMP, but a disproportionate dwarfism eventually develops, with joint laxity and early osteoarthritis. Cervical pathology includes os odontoideum and atlantoaxial instability [88].

A member of the bent bone dysplasias, campomelic dysplasia (CD), is characterized as bowed legs, dislocated hips, clubfeet, and females with male genotype (46, XY) [89]. Vertebrae are typically short and flat. There is a lack of correlation between genotype and phenotype, and life expectancy is highly variable. Cervical instability and kyphosis leading to quadriparesis and death have been described [90].

Chondrodysplasia punctata (CDP) is a heterogeneous group of disorders characterized by stippled epiphyses during infancy. There are multiple genetic causes, but the common phenotype is a midface hypoplasia, hearing loss, distal phalangeal hypoplasia, and radiographic evidence of epiphyseal stippling [91]. Atlantoaxial instability has been reported to get progressively worse with this disorder, and long-term monitoring was recommended. Cervical stenosis has also been described [92].

Osteogenesis imperfecta (OI) is caused by mutations in genes that code for type I collagen [93]. More than 90% of cases are related to mutation of either *COL1A1* or *COL1A2*. Patients usually present with multiple fractures at a young age. Blue sclera, dental abnormalities, and hearing loss may also be present. Due to lack of normal type I collagen, bones demonstrate poor density and ligaments are lax. This combination predisposes these patients to spinal compression fractures, progressive kyphotic deformities, and progressive scoliosis [94].

Diagnosis and Imaging

The diagnosis of a skeletal dysplasia is based on characteristic physical findings and, for many, confirmatory genetic testing. Patients who present directly to the orthopedic surgeon without an underlying diagnosis should be referred to a geneticist. Genetic counseling should be offered to the parents of a newly diagnosed child, as well as to the patients themselves as they reach child-bearing age.

Radiographs are of value to characterize the level of skeletal involvement. In some cases, such as CDP, they are needed for an accurate diagnosis. Whole-spine radiographs, as well as flexion/extension lateral cervical spine radiographs, should be used as a screening tool to monitor scoliosis, kyphosis, and cervical instability. MRI is helpful in determining the extent of spinal cord involvement should any neurologic findings be present, and it can be used in conjunction with CT scans for preoperative planning. Basilar invagination will be found in the severe dysplasias.

Summary

Bone dysplasias represent a highly heterogeneous group of pathological disorders. The genetics and biochemistry have been described here, and further clinical discussion is to be found in Chap. 10.

Infections and Inflammatory Conditions (See also Chapter 7 for the Clinical Discussion of these Conditions)

Juvenile Rheumatoid Arthritis

Introduction

Juvenile rheumatoid arthritis (JRA) encompasses a heterogeneous group of idiopathic disorders, the hallmark of which is chronic synovitis, predominantly of the peripheral joints. JRA refers to children under 16 years of age; this age distinction is somewhat arbitrary, as adult rheumatoid arthritis (RA) can present in children, particularly in older female adolescents.

Classification and Pathology

JRA is subdivided into three main subtypes based on the clinical course 6 months after onset of symptoms: systemic onset, polyarticular (five or more joints), and pauciarticular (four or fewer joints). The subtypes that usually involve the cervical spine are the polyarticular and systemic onset types; only rarely does the pauciarticular type affect the cervical spine. Some pediatric rheumatology centers divide the subtypes based on presence or absence of HLA-B27 antigen.

Cervical spine complications in JRA can include fractures, erosions, subaxial subluxation, and atlantoaxial subluxation (which can rarely lead to basilar invagination in JRA), resulting in neurologic impingement. Rare spinal complications of JRA include spinal lipomatosis, developing as a complication of increased weight gain from glucocorticoid use [95] and rotatory subluxation of the cervical spine [96]. Severe complications are rare in JRA cervical spine disease but should be watched for, especially in systemic or polyarticular cases with severe and/or advanced disease in peripheral joints. Any history of neck pain, neurologic symptoms, or trauma to the neck (however trivial) should be taken seriously in these patients, as they are at higher risk for fracture or dislocation because of concurrent osteopenia or ligamentous damage from chronic inflammation and/or steroid use.

Demographics and Presentation

In the United States and Europe, JRA occurs in approximately 1–22 per 100,000 children, with an estimated prevalence of 86.1 per 100,000, of which half would represent inactive cases [97]. The typical child with JRA will be a preschool-age girl who presents with an asymmetrical inflammatory arthritis, with mildly elevated acute phase reactants, slight elevation of white blood cell count, and mild anemia. Children with systemic JRA present with systemic signs and symptoms, such as rash, fever, lymphadenopathy, serositis, leukocytosis, and marked increase in acute phase reactants and may be quite ill at presentation.

Cervical spine disease is common in JRA and occurs in 50–60% of children with polyarticular and systemic disease but virtually absent in children who have a pauciarticular course [98]. Cervical spine involvement usually occurs 1–2 years from the disease onset. The child will usually present with neck stiffness and decreased range of motion on examination. Pain and torticollis is rare, and when they occur in a patient with JRA, other causes such as trauma, infection, or tumors should be ruled out. Abnormal neurologic findings are also rare. Usually, peripheral arthritis presents first, and the cervical spine disease appears later in the clinical course; rarely, however, cervical spine disease can be the sole initial presentation of JRA, and the child will develop signs of peripheral arthritis later in the course of the disease [99]. Isolated cervical spine arthritis is extremely rare in JRA and careful evaluation will reveal involvement of other joints. Thoracic or lumbosacral involvement is extremely uncommon in JRA.

Diagnosis and Imaging

Standard radiographs are normal at onset of JRA joint disease, and typical destructive changes may take years to develop. Radiographs are obtained at onset for baseline studies for comparison and, more importantly, to rule out other causes of articular symptoms.

The radiographic features of cervical spine involvement in JRA are classified into seven types:

1. Anterior erosion of the odontoid process (common)
2. Anteroposterior erosion of the odontoid process ("apple-core odontoid")
3. Subluxation of C1 on C2
4. Focal soft tissue calcification appearing adjacent to the ring of C1 anteriorly (common)
5. Ankylosis of the apophyseal joints (common)
6. Growth abnormalities
7. Subluxations between C2 and C7

Although there may be hypermobility of C1–C2 with flexion and extension, true instability and myelopathy are rare. Basilar invagination, which often occurs in adult rheumatoid arthritis, is rare in JRA.

In a patient with JRA and neck pain, other imaging modalities are required to rule out fractures, infections, and tumors. A bone scan is used for pinpointing the exact anatomic location of activity, and CT scanning is very helpful to study the anatomy. MRI is particularly helpful for the evaluation of neurologic impairment or cervical spine fractures (Figs. 3.1 and 3.2).

Treatment

All children with JRA need an individualized treatment plan and should be managed by a practitioner skilled in the treatment of these disorders. For children with cervical spine involvement, symptomatic treatment is similar to management of peripheral arthritis and would include analgesics, immobilization (cervical collar and, rarely, traction), and various physical therapy modalities. A short course of glucocorticoids may be helpful for immediate relief of symptoms in the child with new onset of neck disease. Patients rarely develop flexion deformities; early in the course of the disease, a cervical collar may prevent this deformity. A cervical collar is also recommended for patients with involvement of the odontoid process or subaxial subluxation whenever they are in an automobile or other mode of travel. Indications for surgery are neurologic abnormalities, intractable pain, and impending neurologic deficits. In general, outcome is favorable, and early identification of neurologic impingement and prompt surgical treatment provide the best outcome [98, 100].

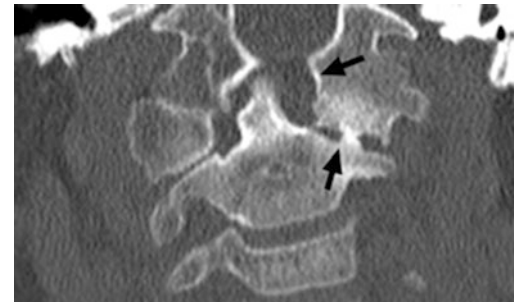

Fig. 3.1 CT scan in a patient with JRA. Note the arrows pointing at the dens showing the erosive nature of the disease as well as at the C1–C2 joint depicting the bony ankylosis

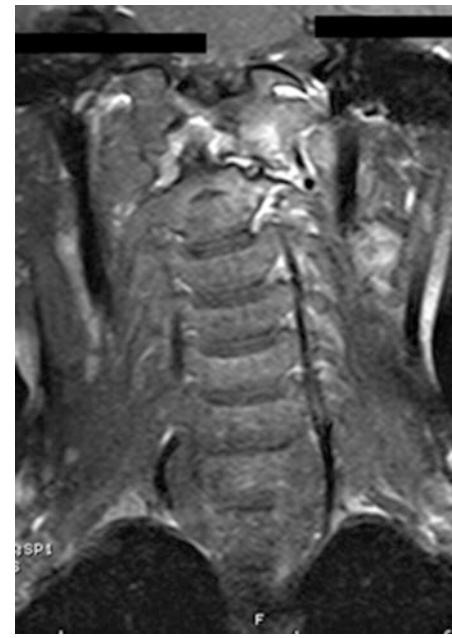

Fig. 3.2 MRI of the patient with JRA shown in Fig. 3.1 depicting the synovitis and erosive nature of the disease process

Chronic Recurrent Multifocal Osteomyelitis

Introduction

Chronic recurrent multifocal osteomyelitis (CRMO) is a rare and unusual inflammatory process that mimics pyogenic osteomyelitis. It is often a diagnosis of exclusion in patients who present with lytic or sclerotic bone findings and periosteal reaction with serological and histological findings negative for infection and tumor. Spinal CRMO is rare but documented and can

initially be confused with pyogenic vertebral osteomyelitis. However, unlike pyogenic vertebral osteomyelitis, the intervertebral disc is spared in CRMO, and typically several vertebral bodies are involved at different levels, with one or several normal intervening vertebrae.

Classification and Pathology

CRMO is an unusual inflammatory process occurring in children and adolescents. It remains contentious as to whether this entity is separate from the SAPHO (synovitis, acne, pustulosis, hyperostosis, osteitis) syndrome, which occurs in adults, or whether these two entities are part of a continuum [101–103]. No causative agent has yet been identified. Proposed theories include a post-inflammatory reaction, as some authors report prior throat infections and elevated antistreptolysin O titers [101, 104]. There is no definite evidence of an autoimmune disorder, and serological tests for rheumatoid factor and antinuclear antibodies and HLA-B27 are usually negative. Occasionally, immunoglobulin levels are mildly elevated, but their significance is uncertain [101]. The differential diagnosis includes Langerhans cell histiocytosis X and lymphoma, malignant metastatic diseases (such as leukemia, neuroblastoma, Ewing's sarcoma), bacterial infection, benign bone tumors (such as osteoid osteoma, osteoblastoma), trauma, Gorham's vanishing bone disease, and avascular necrosis [103–105].

The pathological appearances of CRMO depend on the disease phase [103, 106]. The histological appearance is practically indistinguishable from acute and chronic conventional bacterial osteomyelitis, with the exception that no organisms are found. Early, more acute lesions contain polymorphonuclear (neutrophilic) leukocyte marrow infiltration. Neutrophils may collect in groups surrounded by a lymphocytic infiltrate forming pseudoabscesses. Osteoclasts with marked osteolysis are a common feature. More chronic lesions show fibrosis and lymphocytic infiltrates with new reactive bone in later phases. There may be granuloma formation consisting of collections of neutrophils surrounded by epithelioid histiocytes [106]. Bacterial, viral, and fungal cultures and stains are negative. Specific immunoperoxidase tests (T6 and S-100) can confirm that any histiocytes present are not of the type strongly implicated in Langerhans cell histiocytosis [103].

Demographics and Presentation

CRMO is recognized worldwide, accounting for 2–5% of all osteomyelitis cases [107]. It is primarily a disease of childhood, with the mean age at presentation around 10 years (with range from 6 months to 55 years) [108]. A recent German study estimates an annual incidence of 0.4 per 100,000 children for the condition, with approximately 60 new patients diagnosed annually [109]. There appears to be a female predominance in the disease, with a female-to-male ratio of 2:1 [110]. Initial presentation with primary spinal involvement has been reported to occur in only about 3% of patients with classic CRMO [101], but vertebral lesions accompany other bone lesions in up to 24% of CRMO patients at initial presentation [109].

Patients with CRMO typically present with insidious onset of localized bone pain, associated with soft tissue and bone swelling. Although most patients present with a single symptomatic site, other sites of disease become apparent at imaging or during follow-up, ranging from 1 to 18 [108], with an average of 5 sites of involvement [111]. The common sites of skeletal involvement include the long tubular bones and clavicle, but lesions have been described throughout the skeleton. Involvement of the lower extremity has been reported to be three times more common than disease in the upper extremity [108].

Patients with spinal involvement of CRMO complain of localized back pain but can also present with anterior chest pain from referred pain. Up to 40% may develop vertebral compression fractures [109]. The condition can have a fluctuating clinical course, with exacerbations and remissions lasting for years.

Diagnosis and Imaging

The diagnosis of CRMO is generally one of exclusion by means of laboratory, microbiology, and pathology investigations, which permit the

exclusion of infection (osteomyelitis or bacterial discitis), tumor (histiocytosis X, leukemia, neuroblastoma, and rhabdomyosarcoma), and systemic diseases. Due to the varied clinical and radiological appearances, it is generally recommended that two biopsies be performed from two different sites [103].

Combining the criteria proposed by King et al. with Jurik and Egund [112, 113], the following criteria should be met before a diagnosis of CRMO is made:

1. Multifocal (two or more) bone lesions, clinically or radiographically diagnosed
2. Prolonged course (over 6 months) characterized by varying activity of disease and with most patients being healthy between recurrent attacks of pain, swelling, and tenderness
3. Lack of response to antimicrobial therapy given for at least 1 month
4. Typical radiographic lytic regions surrounded by sclerosis with increased uptake on bone scan
5. Lack of an identifiable organism
6. No abscess, fistula, or sequestrum formation
7. Atypical site for classic bacterial osteomyelitis, such as clavicles and multiple sites
8. Nonspecific histopathological findings and laboratory results compatible with osteomyelitis
9. Occasionally, acne and palmoplantar pustulosis
10. Symmetry of the lesions has been reported as a helpful feature [114]

Radiographic evaluation of spinal CRMO may demonstrate vertebral erosion, osteolytic lesions with a "square outline" within the vertebral bodies, sclerosis, and mild collapse with reduction of disc space [115, 116]. Complete vertebral collapse (vertebra plana) is rare [117]. All vertebrae from the mid-cervical spine to the sacrum can be potentially affected. A characteristic MRI finding is a subchondral endplate fracture-like line associated with increase in signal in the vertebral marrow [118]. Multifocal disease is common, with spontaneous healing and new lesions presenting over a period of years. MRI is particularly useful in determining the activity of lesions, as healed lesions have abnormal contour but normal marrow or show fatty replacement of normal red marrow.

Spinal CRMO can initially be confused with pyogenic vertebral osteomyelitis. However, unlike pyogenic vertebral osteomyelitis, CRMO involving the vertebrae is not associated with involvement of the intervertebral disc, and typically several vertebral bodies are involved at different levels with one or several normal intervening vertebrae.

Treatment

Once the diagnosis of CRMO is established, pain relief and supportive therapy are required if there is minimal and partial vertebral body collapse. By definition, no antibiotics are necessary in treatment of CRMO. Severe vertebra plana may be associated with marked kyphosis, rarely requiring fusion [119]. Rare presentation with cord compression requiring anterior decompression and fusion has been documented [120]. Many medications have been tried, but no single medication has proven to be optimal.

Vertebral Osteomyelitis

Introduction

Pyogenic vertebral osteomyelitis and discitis in children is a spectrum of disease, defined as symptomatic narrowing of the disc space associated with fever and infection-like symptoms. It can affect all pediatric age ranges. Diagnosis and treatment are aided by bone scans and MRI, and a short course of intravenous followed by oral antibiotics will resolve the infection. Surgery is only necessary if there is no response to nonsurgical management.

Classification and Pathology

The current understanding of pathophysiology of vertebral osteomyelitis is that bacteria gain hematogenous access to the vertebrae through spinal arteries that enter through the intervertebral foramen at the level of the disc. These arteries send off ascending and descending branches, creating a rich arterial anastomosis with end arterioles in the

"metaphyseal" region [121]. Once microorganisms lodge in the low-flow vascular arcades in the metaphysis, infection spreads. Spread of the infection to the disc in the child is facilitated by orderly arranged "cartilage canals" within the endplates that contain vascular organs resembling glomeruli [122, 123]. The disc may also be destroyed by bacterial enzymes in a manner similar to the destruction of cartilage in septic arthritis. These changes can be followed radiographically as a narrowing of the disc space, followed by eventual destruction of the vertebral body in later stages.

The upper cervical spine has a peculiar blood supply. Parke and colleagues [124] have demonstrated a venous plexus around the odontoid, called the "pharyngeal vertebral vein," which frequently has lymphovenous anastomoses. This venous plexus may be responsible for hematogenous spread to the upper cervical spine [124, 125].

Abscesses may drain into the soft tissues surrounding the spine or into the spinal canal itself. In the cervical spine, a retropharyngeal abscess may invade the mediastinum [126].

The pathogenesis of neural compromise in pyogenic vertebral osteomyelitis may be related to direct compression by epidural pus, granulation tissue, or bone and disc from the development of spinal deformity and instability. In addition, the cord or nerve roots may suffer ischemic damage from septic thrombosis or may be damaged by inflammatory infiltration of the dura [127, 128].

Demographics and Presentation

Vertebral osteomyelitis remains an uncommon entity among pediatric patients. It affects all pediatric age ranges and is more common in boys than girls. Young children with osteomyelitis are unable to verbalize their complaints, making the presentation vague and nonspecific. Vertebral osteomyelitis in children may present as inability or refusal to walk; a limp; or back, neck, or abdominal pain; fever; and malaise. The child may be febrile and quite ill-appearing or may have a low-grade fever and appear otherwise well. Physical exam may reveal tight hamstrings or a positive straight-leg raising test. The child may also have a characteristic loss of normal "spinal rhythm"—when asked to pick up an object from the floor, the back or neck will be held in an abnormally stiff posture rather than the normally smooth flexion and extension.

Diagnosis and Imaging

Laboratory studies, such as CBC, ESR, and CRP, should be obtained on every patient with suspected vertebral osteomyelitis but may provide nonspecific information. Blood cultures should be obtained in every suspected case, and biopsy should be strongly considered in patients with suspected vertebral osteomyelitis, where definition of a causal agent is particularly important in selection of the appropriate antimicrobial therapy and in defining the duration of therapy. Interestingly, Fernandez et al. [129] found *B. henselae* to be the causative agent in 2 of 14 patients with vertebral osteomyelitis.

The findings on plain radiographs are characteristic, but may not be apparent on presentation. The earliest and most constant radiographic finding, narrowing of the disc space, is present in 74% of patients at presentation [128]. CT scans show abnormalities earlier than plain films. Widening of the retropharyngeal space in the cervical spine, enlargement of the paravertebral shadow in the thoracic spine, or changes in the psoas shadow in the lumbar spine may indicate either abscess or granulation tissue surrounding the infection. After 3–6 weeks, destructive changes in the body can be noted, usually beginning as a lytic area in the anterior aspect of the body adjacent to the disc, and diffusely in the endplate. In children, the appearance of pyogenic vertebral osteomyelitis will rarely progress beyond this point.

The nonspecific presentation of vertebral osteomyelitis in children makes radionuclide studies useful for early detection and localization of infection, before plain films become positive. Clinical and experimental studies have shown that gallium scans become positive before technetium scans do [130, 131]. Technetium scans show increased uptake diffusely in the region of the infection, whereas gallium scans may show increased uptake in a butterfly area around the infected spine [132]. Gallium scanning has been found to have a sensitivity of 89%, a specificity of 85%, and an accuracy of 86% in the diagnosis of disc space infections [130]. In a separate study, technetium scans were found to have a sensitivity

of 90%, a specificity of 78%, and an accuracy of 86%. The accuracy of combined technetium and gallium scans was 94%, and the combined scans are the nuclear medicine study of choice [133].

The imaging modality of choice for the evaluation of spine infections is MRI. MRI permits early diagnosis of infection and recognition of paravertebral or intraspinal abscesses without the risk associated with myelography. MRI has the advantage of providing more anatomic information than radionuclide studies and is capable of differentiating degenerative and neoplastic disease from vertebral osteomyelitis. The changes on MRI occur at about the same time as the changes on gallium scans [133].

On T1-weighted sequences, there is a confluent decreased signal intensity of the vertebral bodies and adjacent disc, making the margin between the two structures indistinct. On T2-weighted sequences, the signal intensity of the vertebral bodies and the involved disc is higher than normal, and there is generally an absence of the intranuclear cleft normally seen within the adult disc [133, 134]. The extent of the infection is best seen, however, using gadolinium contrast enhancement: the disc and the involved portions of the vertebral bodies reveal a marked increased signal intensity that delineates the margins of the infection (Fig. 3.3).

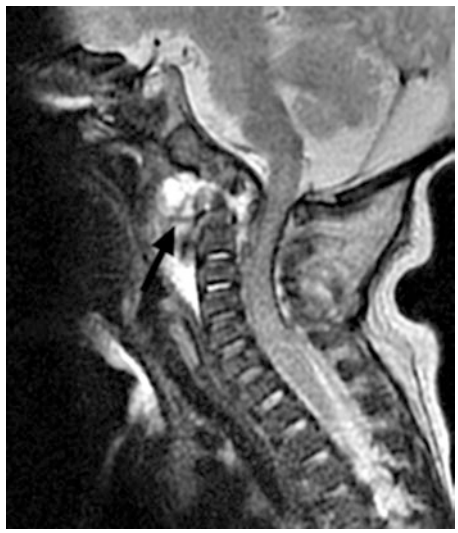

Fig. 3.3 MRI of a neonate with osteomyelitis at the craniovertebral junction. Note the arrow pointing at the abscess surrounding the dens

Treatment

The goals of treatment are to establish tissue and bacteriologic diagnoses, prevent or reverse neurologic deficits, relieve pain, maintain spinal stability, correct symptomatic spinal deformity, eradicate the infection, and prevent relapses. Once the diagnosis of vertebral osteomyelitis is made with laboratory tests, imaging studies, blood cultures, and possibly a biopsy, empiric intravenous antibiotic therapy directed against *S. aureus* should be initiated, and antibiotics later modified as bacteriologic data becomes available. Intravenous antibiotics are continued until the symptoms are largely relieved and laboratory values approach normal. The erythrocyte sedimentation rate (ESR) is a reasonable guide to the response to therapy [127, 128, 135, 136] and can be expected to decrease to half to two-thirds of pre-therapy levels by the completion of successful treatment [128]. If ESR does not decrease with treatment, consideration should be given to a (repeat) biopsy. CRP is also a useful laboratory test to follow resolution of the infection. However, it is still unclear at what point to discontinue antibiotics on the basis of CRP levels. Intravenous antibiotic therapy is typically followed by a 4–6-week course of oral antibiotics.

Surgery is indicated when a clinically significant abscess is present (spiking temperatures and septic course); in cases refractory to prolonged nonoperative treatment, where the ESR and/or CRP remain high or pain persists; in cases with spinal cord compression causing a neurologic deficit; and in cases with significant deformity or with significant vertebral body destruction, especially in the cervical spine [137–139].

Intervertebral Disc Calcification

Introduction

Intervertebral disc calcification (IDC) is a benign condition of unclear etiology, with an abrupt presentation of neck pain related to calcific deposits in the intervertebral discs that resolves over several weeks with conservative measures. Neurologic symptoms are rare, and long-term sequelae of the condition are not well defined.

Classification and Pathology

Pathologic examination of the pediatric intervertebral disc in IDC reveals calcification in the nucleus pulposus. This is in contrast to adults with IDC, where calcification is seen in the annulus fibrosus instead [140, 141]. It is not known why IDC appears in children, and the etiology of IDC is unclear. Proposed theories invoke antecedent trauma (present in 30% of patients) or recent upper respiratory infections (present in 15% of patients). No evidence of inflammation, malignancy, or neovascularization has been identified on histopathological investigation. Recognized causes of calcification in adults, including hyperparathyroidism, hypervitaminosis D, chondrocalcinosis, hemochromatosis, ochronosis, pseudogout, gout, and degenerative diseases have not been reported in any case of pediatric disc calcification [141–146].

Demographics and Presentation

Intervertebral disc calcification is rare. Because most cases of IDC are asymptomatic, it is difficult to estimate the incidence of the entity. Calcifications can occur throughout the cervical and thoracolumbar spine, but are most frequent in the cervical spine, and are especially symptomatic when located there [140, 141, 143–145, 147]. In 30–40% of the patients, multiple discs may be involved. Boys seem to be more often affected than girls. The average age at presentation is 7–8 years old (range: 7 days to 20 years) [141–144, 146, 148, 149].

The most common clinical presentation of IDC is neck pain and stiffness, which may be abrupt in onset over a 12–48-h interval. Twenty-three percent of those presenting with IDC will be febrile, and a quarter will present with torticollis. Radiculopathy may occur if the calcification herniates through the annulus fibrosus. No protrusions have been seen in the asymptomatic patients, but 38% of symptomatic patients have had detectable protrusions in one series [150]. Myelopathy is rare. Dysphagia has been described associated with an anterior protrusion [146, 151, 152].

Diagnosis and Imaging

Radiographic examination will reveal oval or round calcifications that are often located in the middle portion of the disc; occasionally the entire disc will be calcified. An indentation of the adjacent vertebral endplate is also seen occasionally [141, 143, 144]. In two series, associated spinal deformity such as decreased height of the adjacent vertebral bodies was also noted [144, 148].

MRI scans will reveal regions of reduced signal intensity on both the T1- and T2-weighted images, consistent with calcification. If any neurologic symptoms are present, an urgent MRI of the cervical spine is indicated. CT scans can confirm disc calcifications (Fig. 3.4a, b); however, given the radiation dose, MRI is the preferred cross-sectional imaging modality in the pediatric population.

Laboratory tests (complete blood count and inflammatory markers) are typically normal or may reveal a moderate inflammatory state [141, 144, 146, 153].

Treatment

The natural history of IDC is one of symptom resolutions within weeks to months, and the treatment of choice is conservative and symptomatic. Treatment modalities include oral analgesics, nonsteroidal anti-inflammatories, muscle relaxants, a soft cervical collar, and traction, depending on severity of symptoms. Avoidance of contact sports is prudent. Surgical intervention is warranted in cases of progressive neurological deficit and will usually involve an anterior cervical discectomy and fusion.

Two-thirds of children are reported to be symptom-free within 3–4 weeks, and 95% will be free of symptoms by 6 months. Radiographic evolution will consist of regression or disappearance of the calcific deposits in 90% of patients, with half of the improvement occurring within 6 months. However, in asymptomatic children, calcifications may persist for much longer periods. In children with multiple lesions, each lesion may regress at a different rate. Persistent flattening of the vertebral bodies may continue into

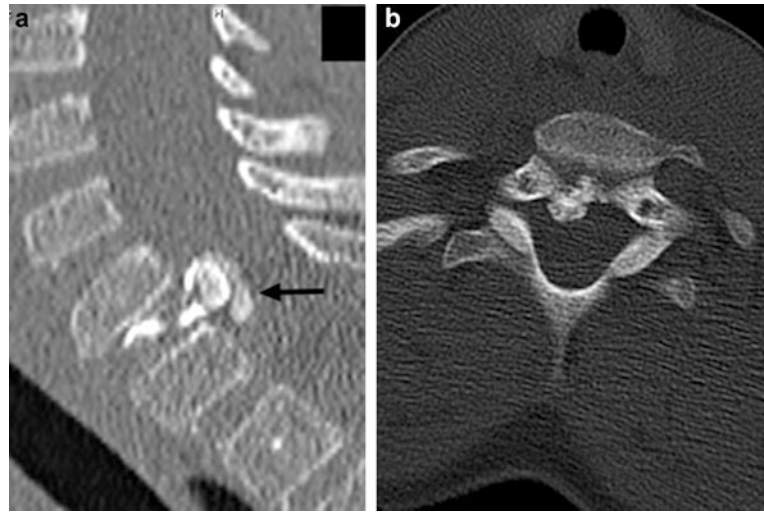

Fig. 3.4 (**a**, **b**) (**a**) Sagittal CT cut in a 10-year-old patient with an intervertebral disc calcification. The arrow points at the extrusion into the canal. (**b**) Axial CT cut demonstrating the disc calcification and protrusion

adulthood but whether these alterations are the cause of severe degenerative vertebral disease in adult life is unclear [141–143, 149, 154].

Atlantoaxial Rotatory Subluxation

Introduction

Atlantoaxial rotatory subluxation (AARS) is a common cause of acquired torticollis in children. Most common causes of AARS include trauma and infection, although congenital and iatrogenic causes also exist. If the subluxation persists for more than 3 months, the child will present with a resistant and unresolving torticollis that is referred to as atlantoaxial rotatory fixation. Patients present with headaches, neck pain, and a "cock-robin" position after trauma or an episode of upper respiratory infection. Diagnosis is made by fine-cut dynamic CT scans, and treatment outcomes differ depending on duration of symptoms since onset.

Classification and Pathology

Nearly 50% of the rotation in cervical spine is owed to the atlantoaxial articulation. There are several unique aspects of the pediatric spine that may predispose children to hypermobility of the C1–C2 articulation and AARS. In children, the ligaments and joint capsules possess sufficient elasticity to allow for hypermobility without disruption. The facet joints, especially at C1–C2, are shallower and more horizontally oriented in children, allowing greater freedom of motion. The uncinate processes are virtually absent in the cervical spine of young children, allowing excessive lateral bending and rotation. Additionally, the disproportionate size of a child's head and the underdeveloped neck musculature allow for excessive rotation and bending at the atlantoaxial junction. The condition also appears to be correlated to Down syndrome, Morquio syndrome, and juvenile rheumatoid arthritis, all of which include congenital atlantoaxial anomalies and various degrees of ligamentous laxity.

Fielding and Hawkins classified rotatory subluxation into four types, which remains the most commonly used classification to date [155]. In type 1, the most common type, the rotation is within the normal range of C1–C2 motion. The transverse ligament is intact and the dens acts as the pivot. Type 2, the second most common type, refers to rotatory fixation with 3–5 mm of anterior displacement of the atlas. This is associated with deficiency of the transverse ligament and unilateral anterior displacement of one lateral mass of the atlas while the opposite, intact joint acts as the pivot. In type 3, the atlas is translated more than 5 mm anterior to the axis and is associated with failure of both the transverse and secondary ligaments. Both lateral masses of the atlas are displaced anteriorly, one more than the other,

producing the rotated position. Type 4 involves rotatory fixation with posterior displacement, where a deficient dens allows for posterior shift of one or both lateral masses of the atlas, one of them shifting more than the other so that the atlas rotates on the axis.

Demographics and Presentation

Atlantoaxial rotatory subluxation is characteristically a pediatric problem. Typically, the child will present with neck discomfort, limited neck range of motion and torticollis, and occasional headaches. Initially, patients with AARS may simply appear to have a stiff neck, with truncal rotation ameliorating the loss of cervical rotation. Soon after, the patient assumes the "cock-robin" position, with the head rotated to one side and the neck tilted laterally to the contralateral side. History will usually reveal a preceding infection such as pharyngitis or otitis media, previous neck surgery, or trauma to the neck or upper trunk.

The infectious etiology for AARS is known as *Grisel syndrome* and most commonly occurs after an upper respiratory tract infection but can also occur after tonsillectomy, pharyngoplasty, or retropharyngeal abscess [156]. Because of the anastomoses between the veins and lymphatics draining the pharynx and periodontoid plexus, inflammation in the pharynx can lead to attenuation of the transverse ligament or synovium, or both, surrounding C1–C2 and resulting in subluxation.

While the neck discomfort typically decreases over time, the torticollis worsens; the condition is called rotatory *fixation* if the malalignment persists for more than 3 months.

Diagnosis and Imaging

Distinguishing AARS from other causes of acquired torticollis in children requires the combination of a detailed history, physical examination, and imaging studies. History can be helpful by identifying predisposing factors such as trauma or infection, as well as any prior occurrences and their duration. Neurologic exam is typically normal in children with AARS, and any abnormality should raise suspicion for other causes of torticollis. A careful neurologic exam is also necessary to establish baseline prior to any intervention. There are three clinical signs that help to distinguish muscular torticollis from true AARS. Normally, neck rotation beyond 20° will result in deviation of the C2 spinous process to the *contralateral* side. In true AARS, there is often a palpable deviation of the spinous process of C2 in the *same direction* as the rotation of the head. This abnormal rotation of C2 may be a compensatory mechanism to restore the normal mechanical axis of the neck in AARS. Second, in AARS, the sternocleidomastoid muscle *ipsilateral* to the direction of head rotation will exhibit spasm in an effort to correct the deformity. The third clinical sign of true AARS is the inability of the patient to rotate the head past midline in the direction opposite that of the subluxation [157].

The radiographic diagnosis of AARS is difficult to establish with plain radiography alone. Anteroposterior and open-mouth odontoid images should be obtained in as close to a neutral position as possible. The lateral masses appear different in size because one is rotated anteriorly and one posteriorly and the distances from the lateral masses to the dens are asymmetrical. The markedly abnormal head and neck positions make the radiographic views difficult to obtain and interpret, and its usefulness mainly lies in ruling out other concomitant injuries to the cervical spine. Likewise, static CT scans of the upper cervical spine, while revealing the C1–C2 resolution in greater resolution (Fig. 3.5), may also fail to differentiate between normal rotation and true AARS.

Because AARS implies an element of rotation fixation between C1 and C2, dynamic CT scanning offers a more clinically relevant assessment of C1–C2 rotation. First introduced by Rinaldi et al. in 1979, the technique involves three fine-cut axial CT scans through the upper cervical spine with patient's neck in neutral (presenting) position, followed by a scan with neck maximally rotated to either side, limited by discomfort [158]. The relative positions of C1 and C2 in normal conditions and in AARS can be quantified by the methodology described by Pang and Li in a series of three papers in 1995: the angle formed between anteroposterior lines drawn through the

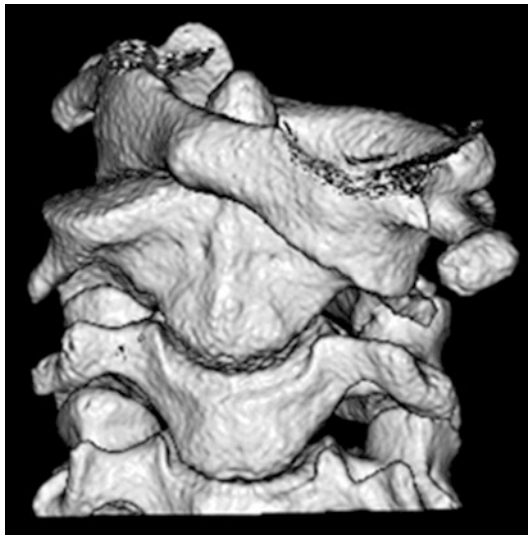

Fig. 3.5 Three-dimensional CT image of a patient with a fixed rotary subluxation of C1–C2. Note the rotation of C1 in relation to C2 with joint incongruity

midline of C1 and C2 vertebrae determines their relative position. Normally, C1 may rotate independently of C2 for the first 20° of rotation from midline. With rotation beyond 20°, C2 begins to rotate in the same direction. With normal rotation and muscular torticollis, the angle reduces to nearly zero when C1 is returned to midline and may crossover C2 when the head is turned to the contralateral side [159]. With AARS, the C1–C2 angle *fails to reduce* with bringing the head toward midline and *never* crosses over C2 [160].

Treatment

Once AARS is diagnosed, treatment should be undertaken expeditiously. The length of time prior to reduction seems to correlate with the likelihood of recurrence and failure of closed reduction techniques. In particular, patients whose AARS remains unreduced for longer than 3 weeks have a significantly higher frequency of persistent or recurrent deformity after attempted reduction. This may be because of chronic changes in the ligament and joint structure of the atlantoaxial complex.

In cases of AARS with a short duration of symptoms prior to presentation (1–2 weeks), a trial of cervical immobilization, anti-inflammatories, and close clinical follow-up is indicated. If spontaneous reduction does not occur after a week, the patient should be placed in head halter traction and started on muscle relaxants and analgesics, which usually relieves the symptoms if they have persisted less than 1 month. In cases of atlantoaxial rotatory subluxation that have endured more than 1 month, reduction with head halter traction is unlikely [161], but halo traction can be used to attempt reduction. Documentation of the reduction should be performed by CT scan.

In a study by Subach et al. in 1998, no child experienced a recurrence if reduction was achieved within 21 days of symptoms [162]. After reduction, all children are best immobilized for 6 weeks in a collar or Guilford orthosis, based on their age and likelihood of compliance. In the case of recurrence, some authors advocate a second trial of closed reduction followed by prolonged immobilization for up to 3 months [162]. In the case of irreducible subluxation or second recurrence, if the subluxation has been present for more than 3 months or if the patient has instability or neurologic compromise, open reduction and posterior C1–C2 fusion is warranted [163].

Untreated AARS leads to pain and progressive deformity. Changes in vocal character may result from chronic pharyngeal compression. The patient may have difficulty opening the mouth, may experience elevation of one shoulder, and may demonstrate changes of the subaxial spine.

Summary

Many different pathologies affect the child's cervical spine. Genetic, syndromic, inflammatory, and other processes all contribute to instability and potential spinal cord compromise. Often there is more than one disorder contributing to the clinical picture. Thus a child with Down syndrome may have an acute infection of the pharynx, compounding underlying ligamentous instability. A diligent search for all contributing pathologies is essential in the evaluation of these children.

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Radiology of the Growing Cervical Spine

4

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Introduction

The evaluation of the pediatric cervical spine presents certain diagnostic dilemmas that are not encountered in adults. There are several well-documented radiographic pitfalls that can lead physicians to misinterpret normal anatomical variations for true pathologic changes. Physeal variations, unique vertebral architecture, incomplete ossification, and hypermobility of the cervical spine may all cause uncertainty when interpreting radiographs of a child with a history of trauma, pain, or stiffness [1]. Furthermore, the limited ability of children to cooperate during flexion and extension films compounds the problem. There are various radiographic measurements that can be utilized to differentiate true pathology from normal variants; however, knowledge of the embryological development and postnatal ossification patterns of the cervical spine is paramount in recognizing traumatic spinal injuries in children. Although cervical spine injuries in children are rare, they are associated with high morbidity and mortality. Therefore understanding the presentation of the developing pediatric cervical spine on plain films, CT, and MRI is essential in ensuring early and accurate diagnosis.

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Postnatal Development of the Pediatric Cervical Spine

Evaluation of the pediatric cervical spine, particularly in the setting of trauma, often becomes problematic because of the unique radiological anatomy of children [2]. Without proper knowledge of normal anatomical variants encountered in the pediatric population, physicians may misinterpret normal radiological anatomy as a pathological condition. The aspects that make pediatric cervical spine radiology so challenging are ossification centers and synchondroses. Within the cervical spine, different vertebral levels exhibit different phases of ossification and synchondrosis fusion at different ages of life (Table 4.1). These normal variants can be misinterpreted as fractures if the radiologist has limited knowledge of the age of ossification and closure of the various synchondroses. The following will attempt to provide a helpful guide in the developmental

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Table 4.1 Ages of visibility and fusion of ossification center and closure of synchondroses in pediatric cervical spine

	Anterior OC visible	Posterolateral OC (neural arches) visible	Anterior OC complete	Neural arch OC complete	Anterior NCS	Posterior SC	Subdental SC	Intradental SC
C1	6 mos–2 yrs	7th fetal wk	8–10 yrs	5 yrs	7 years	3–5 yrs	–	–
C2	Birth	Birth	3–4 yrs	2–3 yrs	5 yrs	4–6 yrs	3–5 yrs	3 mos
C3-C7	Birth	Birth	1 yr	1 yr	3–6 yrs	2–3 yrs	–	–

OC ossification center, *NCS* neurocentral synchondrosis, *SC* synchondrosis

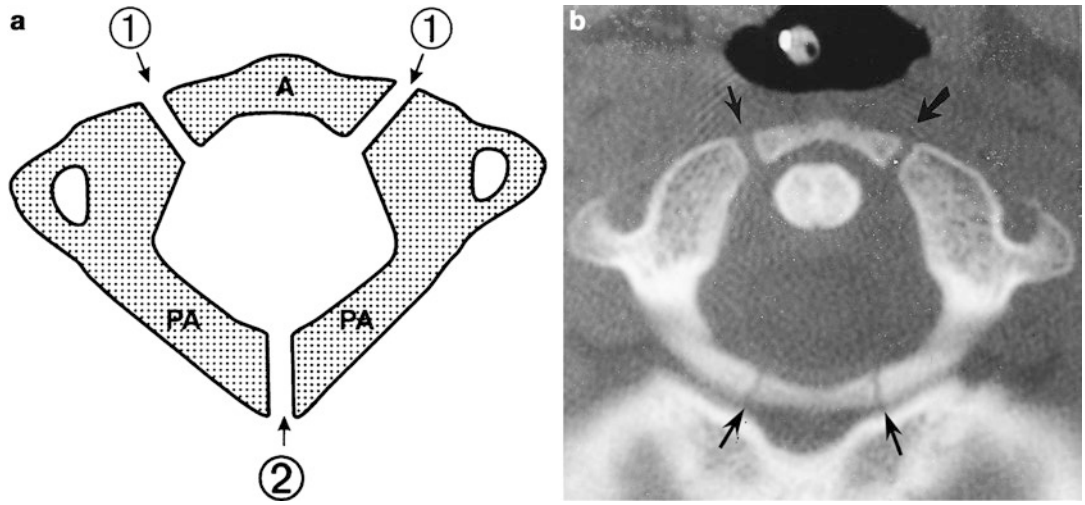

Fig. 4.1 (**a, b**) Synchondroses of C1. (**a**) Diagrammatic representation of the usual anterior arch synchondroses (1) and normal posterior arch synchondrosis (2); anterior arch (A), posterior arch (PA). (**b**) Axial CT study showing the same anterior synchondroses (anterior arrows). In this patient there also are two anomalous, posterior synchondroses (posterior arrows) (From Imaging of the Cervical Spine in Children by Leonard E. Swischuk. © 2013 Springer Science + Business Media, New York. With permission of Springer Nature)

anatomy of the growing pediatric cervical spine. Both the pediatric and the adult cervical spine can be discussed as three distinct units: C1, C2, and C3–7. The discussion can be separated in this fashion due to the unique radiographic anatomy, embryological development, and biomechanical characteristics that each unit displays.

C1

C1, also known as the atlas, has several morphological characteristics that distinguish it from other cervical vertebrae, including an absence of a body, its unique ring-shaped structure surrounding the dens of C2, a unique articulation with the cranium on its cephalad aspect, and a largely cartilaginous structure at birth [3]. Although formed embryologically, as mentioned above, C1 is unique in that it is derived from the basioccipital somites. It also yields a portion of the initial somite cell mass to form the dens [4].

C1 has three primary ossification centers: two posterolateral ossification centers that contribute to the neural arches and one anterior ossification center which becomes the anterior arch (Fig. 4.1).

The anterior arch becomes visible radiographically between 6 months and 2 years but usually around 1 year of age [3–5]. In about 20% of cases the anterior arch may be ossified at birth [3]. The relative delay in the radiographic appearance of the anterior ossification center may contribute to misinterpretation of plain radiographs involving an infant or young child, particularly in the setting

of acute trauma. Therefore CT and MRI are becoming increasingly relied upon for more accurate evaluation and diagnosis of the pediatric cervical spine [4]. The neural arches appear in the seventh fetal week and are therefore present at birth. Ossification of the cartilaginous arch proceeds in a centrifugal manner from within each of the three ossification centers. The cartilaginous bridges that remain between the anterior arch and two neural arches, as well as posteriorly between the neural arches, are known as synchondroses. The closure of these synchondroses by ossification will eventually result in the formation of a continuous osseous vertebral ring [3].

By 1 year of life there are three synchondroses visible: two neurocentral synchondroses and one posterior synchondrosis [3]. There are numerous studies evaluating the age at which the C1 vertebral synchondroses fuse on plain film, CT, and MRI, and all have different results. The age at which C1 synchondroses close remains controversial. However, there has been an established range, which may help guide the radiologist when interpreting images. The posterior midline synchondrosis closes between three to 5 years of age, whereas the anterior neurocentral synchondroses usually close around 7 years of age. This ambiguity can lead to misinterpretation of unfused synchondroses as fractures in the pediatric trauma case. Although sometimes difficult to differentiate persistent cartilaginous synchondrosis from a fracture, there exist a few general rules that the radiologist can follow to help guide interpretation of the images, particularly in the setting of trauma. Generally synchondroses (or physeal plates) are smooth and regular, are seen in predictable locations, and have sclerotic lines. In contrast, acute fractures can occur in unpredictable locations and are irregular and non-sclerotic [3]. With this knowledge the radiologist can avoid misinterpretation of radiographic images.

On CT the posterior synchondrosis closes by 4–5 years of age, while the neurocentral synchondrosis closes by 7 years of age, similar to that seen on plain film [3, 6]. However, there was still evidence of lucency on CT scan of the posterior synchondrosis at 8 years of age in a small percentage of patients [7, 8]. At 1 year of age, the transverse foramen of C1 is visualized. By 2 years of age the superior articular facet appeared with posterolateral sclerotic margins, while the inferior articular facet was not well visualized at this point. Also at 2 years of age, the tubercle for the transverse ligament appears as a density change on CT [7].

Although the aforementioned development of C1 is the normal, there are three major variations in development of the anterior arch of C1 that are essential to know when evaluating the cervical spine radiograph in a pediatric patient. Normally only a single anterior ossification center forms, becoming the anterior arch of the C1 vertebrae; however, sometimes the anterior arch of C1 arises from two ossification centers which ultimately fuse with each other (Fig. 4.2) and then the neural arches. Occasionally the ossification center fails to appear at all, and in its place there are forward extension and fusion of the anterior portions of each neural arch. Failure of posterior fusion of the neural arches results in a posterior rachischisis [9]. Finally, the center of the anterior arch may be absent, and the anterior portions of the neural arches fail to fuse anteriorly, resulting in a cleft [4, 5].

One of the unique characteristics of C1, as previously mentioned, is its articulation with the cranium. Development of this articulation, known as the craniovertebral junction (CVJ), is a complex process. The presence of large unossified cartilaginous parts and numerous synchondroses creates problems for radiologists and may be potential pitfalls when attempting to diagnose fractures in children. Knowledge of the normal radiographic anatomy of the CVJ is exceedingly important, as the CVJ is the most common site of osseous and ligamentous injury in pediatric cervical spine trauma [10].

C2

C2, also known as the axis, has the most complex and unique development of all the cervical vertebrae, in part due to the existence of the dens. It is for this reason that most difficulties arise in

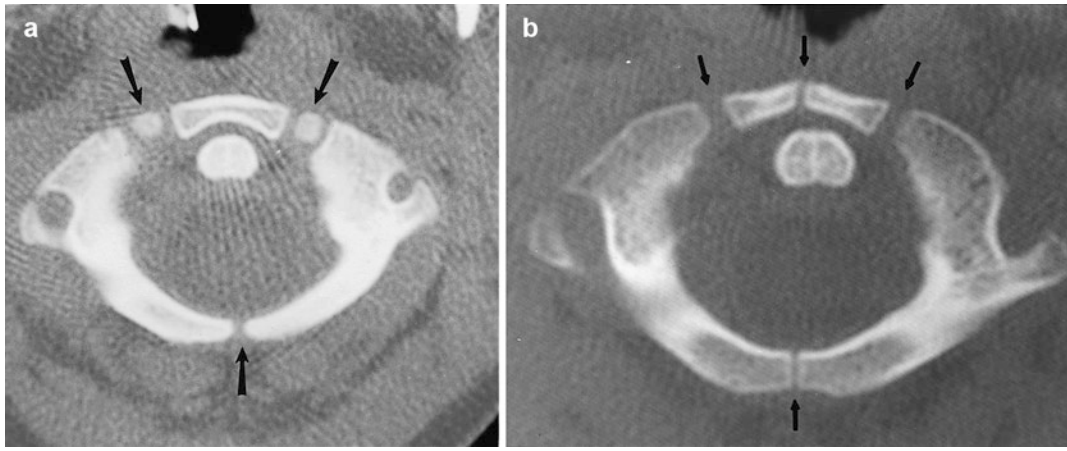

Fig. 4.2 (**a, b**) Variable synchondrotic configurations of C1. (**a**) Two extra ossicles (anterior arrows) are present in this patient. Also note the posterior synchondrosis through the posterior neural arch (posterior arrow). (**b**) Three separate anterior syndroses are present (arrows) in this individual. Normal posterior arch synchondrosis (posterior arrow) (From Imaging of the Cervical Spine in Children by Leonard E. Swischuk. © 2013 Springer Science + Business Media, New York. With permission of Springer Nature)

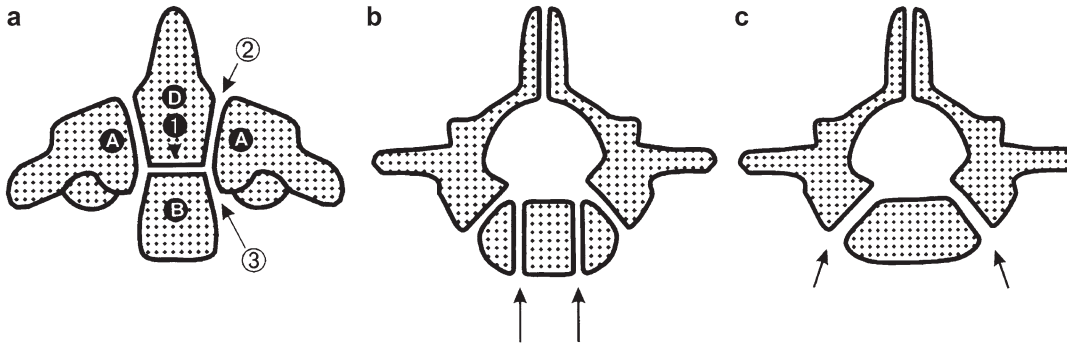

Fig. 4.3 (**a–c**) Normal synchondroses of C2, diagrammatic representation. (**a**) Coronal plane. Note the dens (D) and body (B) of C2. Also note the synchondroses between the dens and the body (B) of C2. Also note the synchondroses between the dens and the body (1), the dens and the neural arch (A) (2), and the body and the neural arch (3). (**b**) Axial plane. Note the parallel configuration of the synchondroses (arrows) between the dens and the neural arch. **(c)** Axial plane. The synchondroses (arrows) between the body and neural arch of C2 are more divergent (From Imaging of the Cervical Spine in Children by Leonard E. Swischuk. © 2013 Springer Science + Business Media, New York. With permission of Springer Nature)

differentiating normal anatomy from pathological change (Fig. 4.3a–c) [2, 5]. Ogden et al. have nicely illustrated the timeline of radiological development of the axis. The dens, or odontoid process, develop two primary ossification centers in utero, which normally fuse in the midline by the seventh fetal week but may persist as two separate centers up to 3 months after birth. This is known as "dens bicornis" [11]. If fusion of the dens occurs in utero, there are four primary ossification centers in C2 at birth: one for each neural arch, one for the body, and one for the dens. As the ossification of C2 is so complex, we will first discuss the chronologic changes seen on transverse radiography and then discuss standard anteroposterior radiography.

In the neonate the midline intradental synchondrosis is usually not visible as was discussed above. This synchondrosis may persist until 3 or 4 months of age. The neurocentral and posterior synchondroses are clearly open and evident on plain film. It is important to notice the more circular appearance of

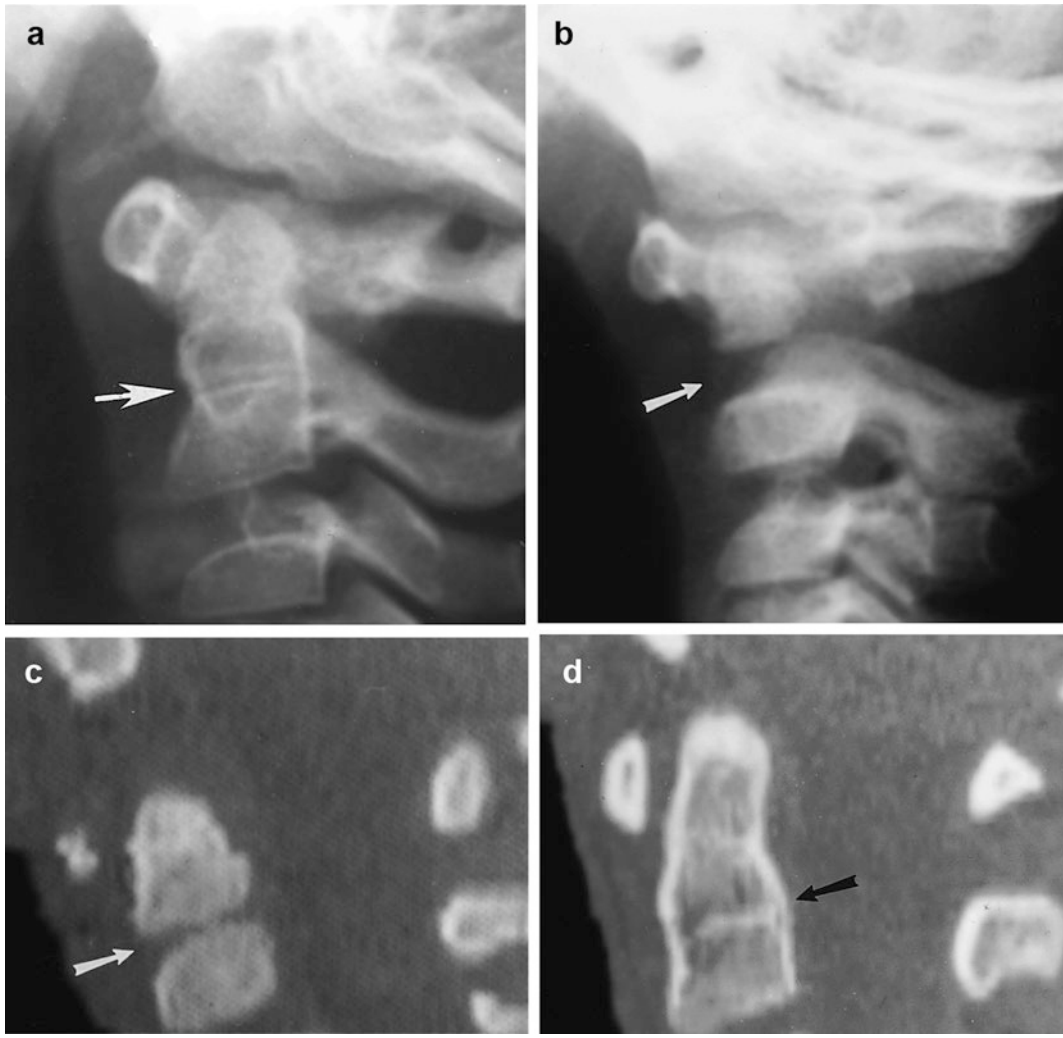

Fig. 4.4 (**a–d**) Normal synchondrosis between the dens and the body of C2. (**a**) Note the typical radiolucent synchondrosis (arrow) between the dens and the body of C2. (**b**) Young infant with a very wide, and potentially confusing, but still normal synchondrosis (arrow) between the dens and body of C2. (**c**) Sagittal reconstructed CT image demonstrates the normal synchondrosis (arrow) between the dens and body of C2 in an infant. (**d**) CT study in an older patient demonstrates virtual obliteration of the synchondrosis (arrow) between the dens and the body of C2 (From Imaging of the Cervical Spine in Children by Leonard E. Swischuk. © 2013 Springer Science + Business Media, New York. With permission of Springer Nature)

the spinal canal before closure of these synchondroses. The posterior ossification centers extend from the neurocentral synchondroses within the eventual vertebral body laterally to the facets and then around to the posterior synchondrosis.

By 3 months of age (Fig. 4.4a–d) the intradental synchondrosis is fused, all three synchondroses have enlarged, and there is clearer visualization of the vertebral artery foramina. There is circumferential thickening of the inner cortex, and ossification is extending posteriorly into the spinous processes.

At 1 year ossification has extended further posteriorly, more clearly defining the paired spinous processes. By 3 years the posterior ossification centers have fused at the spinous processes, closing the posterior synchondrosis and effectively stopping further internal canal growth. There is increased sclerosis of the inner aspects of the posterior elements, which extends anteriorly to the

neurocentral synchondroses; however, there is no comparable thickening of inner surface of the dens and centrum. By 5 years closure of the neurocentral synchondroses is almost complete. One variation that may be encountered around five or 6 years of age is one where the neurocentral synchondroses have closed; however, the posterior synchondrosis remains open. This is an illustration of a biologic variation and should be recognized by the radiologist as a normal variant and not a fracture. By 9–10 years of age closure is complete anteriorly and posteriorly, and the vertebral artery foramina are well visualized. By 14 years the vertebra is in its final, skeletally mature shape.

Throughout the maturation and ossification of the C2 vertebra, there is marked variation in the shape of the spinal canal, which begins as circular but ends as triangular. Similar to the canal of C1, that of C2 reaches its maximum sagittal and coronal diameters by four to 5 years of age, when the posterior and neurocentral synchondroses close. However, it is important to note that the vertebra continues to increase in size by periosteal osteogenesis along the external borders and vertebral height increases through the ring apophysis and chondrum terminale [11].

Along with transverse imaging, it is also important to elucidate the anteroposterior radiographic development of the C2 vertebra due to the existence of the dens or odontoid process. The radiographic anatomy of this unique structure can only be truly appreciated with anteroposterior views of the cervical spine. In the pediatric population the dens presents its own unique set of problems for the radiologist in differentiating true fracture from normal anatomical variant. Although sometimes already fused at birth, the two primary ossification centers of the dens may still be visible in the neonate, particularly on CT scan [10]. The dens is clearly separated from the vertebral body by a cartilaginous structure known as the dentocentral, or subdental, synchondrosis. As in the transverse views of the spine, the neurocentral synchondroses are visible, separating the centrum ossification center from the two lateral ossification centers. Also in the neonate, one can appreciate the chondrum

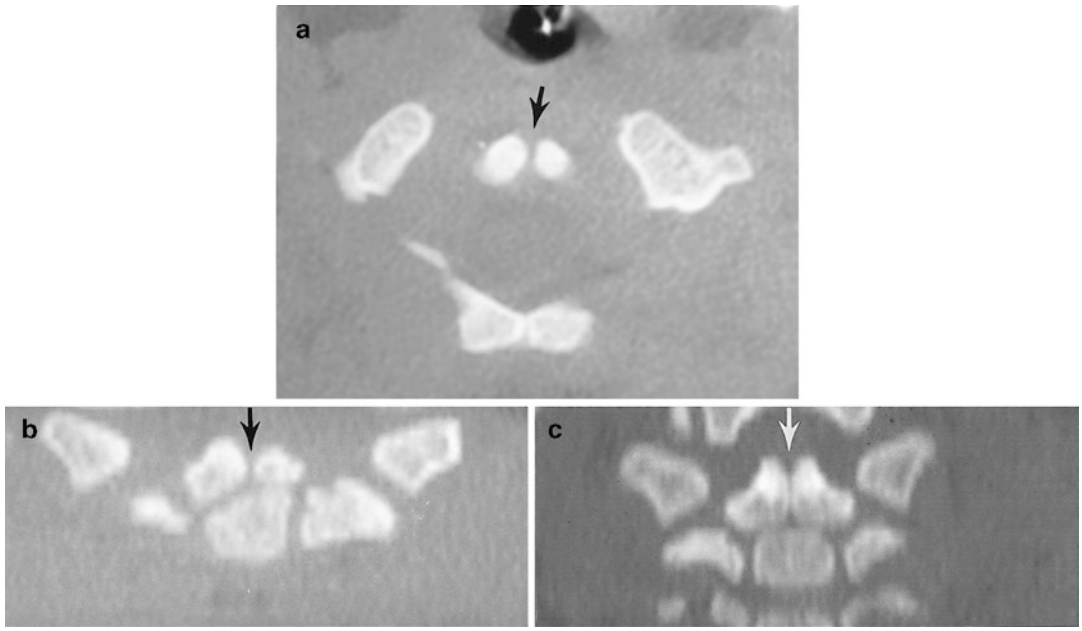

Fig. 4.5 (**a–c**) Bifed dens. (**a**) Note the bifed dens (arrow). (**b**) Coronal reconstruction again demonstrates the bifed dens (arrow). (**c**) Another patient with a bifed dens (arrow) (From Imaging of the Cervical Spine in Children by Leonard E. Swischuk. © 2013 Springer Science + Business Media, New York. With permission of Springer Nature)

terminale, which is the cartilaginous region at the cephalad tip of the dens. This structure usually has a cleft or "V"-shaped junction with the bipartite dens ossification centers (Fig. 4.5a–c). It is important to note anatomically that the vertebral body is actually a composite of ossification from the lower portion of the dens as well as the centrum. Therefore the superior margin of the eventual vertebral body is above the lower level of the dens. At 3–4 months of age the two ossification centers of the dens have fused together. All other synchondroses are still visible at this point, including the one between the dens and the posterior ossification center, which extends from the facet to the junction of the dentocentral and neurocentral synchondroses.

At 1 year there is minimal change that can be appreciated, other than increased radiodensity and enlargement of all ossification centers. By 3 years there is evidence of closure of the dentocentral synchondrosis as well as its W communication with the facet regions. By 5–6 years closure of all synchondroses is underway. As early as 5 years (Fig. 4.6a–d) there may also be evidence of an ossiculum terminale, which is the ossification of the chondrum terminale at the cephalad tip of the dens; however, this typically is not evident until 8 years of age and usually completely ossifies by 10.5 years of age [10, 11].

By 9–10 years the synchondroses have all fused. By age 12 the os terminale fuses with the dens. There are a few essential points to be aware of when evaluating the AP radiograph of the pediatric cervical spine. The first is that it is important to realize that the dentocentral synchondrosis is a bipolar growth zone responsible for longitudinal growth of the upper part of the C2 centrum and lower part of the dens. Secondly,

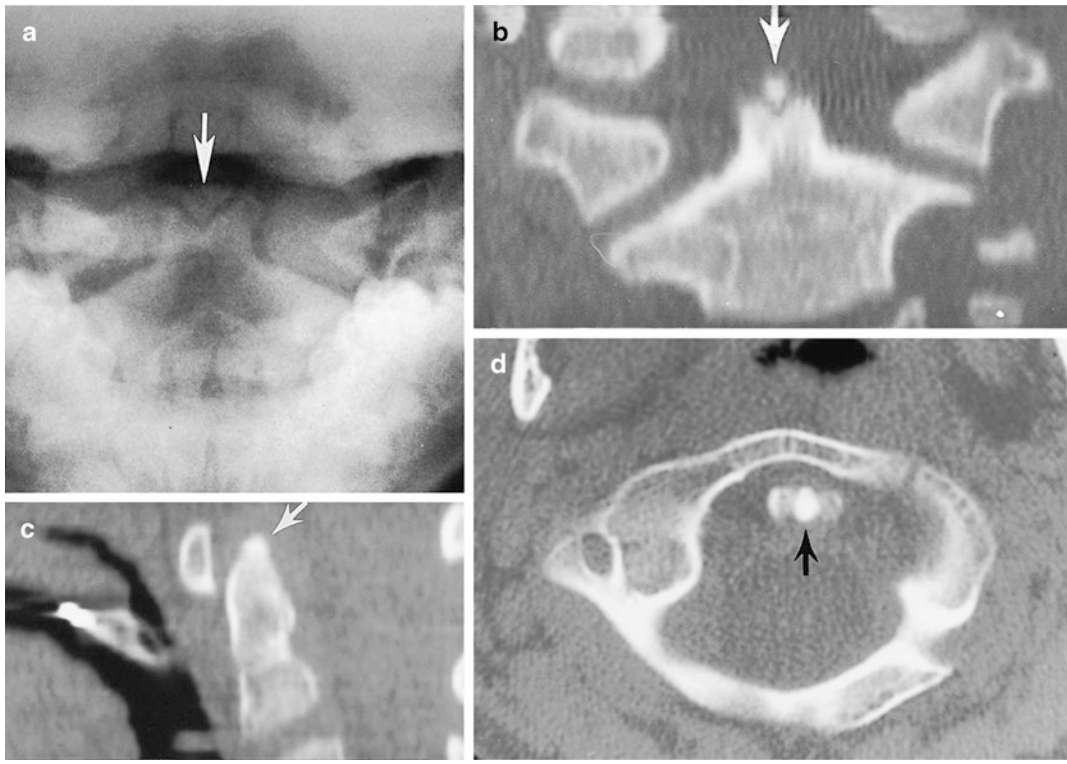

Fig. 4.6 (**a–d**) Normal os terminale. (**a**) Note the os terminale (arrow) embedded into the V-shaped notch of the dens. (**b**) Similar findings; os terminale (arrow) on a coronally reconstructed CT study. (**c**) Sagittal CT reconstruction demonstrates the typical position of the os terminale (arrow). (**d**) Axial CT study demonstrates the usual central position of the os terminale (arrow) (From Imaging of the Cervical Spine in Children by Leonard E. Swischuk. © 2013 Springer Science + Business Media, New York. With permission of Springer Nature)

as mentioned above, the dentrocentral synchondrosis is located below the level of the articular facets, within the eventual C2 vertebral body. A third point to recognize is that although the dentocentral cartilage begins to ossify by ages 5–7 and disappears completely by age 10, a sclerotic ghost of this structure may persist into adolescence and even adulthood and should not be confused with a fracture [11, 12].

CT imaging of the axis provides a few unique characteristics that are not seen on plain film. On CT, two separate ossification centers of the dens can be visualized in patients up to 2 years old, with an apparent deficiency of the posterior cortex of the dens visible in patients as old as 7 years [7]. By 2 years of age the superior articular facet is visible, with indistinct cortical borders seen between ages 6 and 15 [9]. Because of the greater sensitivity to density differences, CT may show unfused synchondroses at a later age than seen on plain film; however, with knowledge of the location and patient history, they should not be confused with fracture.

C3–7

The remaining cervical vertebrae, C3 through C7, should be discussed as a unit, as they exhibit the same developmental pattern [2]. The vertebrae of the lower cervical spine develop from three primary ossification centers: one for the body and one for each of the two neural arches. Similar to C2, the neural arches fuse posteriorly by age 2–3 years, and anteriorly the neurocentral synchondroses fuse between 3 and 6 years of age [2, 9]. As with C1 and C2, secondary ossification centers may exist. These are usually seen at the tips of the transverse processes and spinous processes, persisting until the third decade of life, mimicking fractures. Secondary ossification centers may also appear in the superior and inferior aspects of the vertebral bodes and remain unfused until early adulthood, also simulating fractures [2].

MRI imaging of the development of the pediatric cervical spine reveals a similar timeline as that seen on plain film and CT; however, the unique appearance of the images requires a brief description. On T2-weighted images, the neurocentral synchondroses appear as a low-intensity, posteriorly directed, concave, black line between the neural arch and the centrum [13]. In young children the line is easily visualized on sagittal views, as it appears continuous with the synchondrosis in the adjacent vertebrae. The synchondroses could be visualized on T1-weighted imaging as well; however T2 weight imaging provided better clarity and resolution [13].

Normal Anatomical Variants Found in the Pediatric Cervical Spine

Evaluation of the pediatric cervical spine can be very difficult, owing to the presence of several normal anatomical variants not encountered in the adult cervical spine. With the knowledge of the development of the cervical spine as discussed above, and typical variants which will be emphasized below, physicians can avoid confusing anatomical variants with pathological conditions.

Pseudosubluxation

Physiologic laxity of the cervical spine in children may give the false impression of subluxation. In children the C2–C3 space and, less commonly, the C3–C4 space have a normal physiologic displacement, termed pseudosubluxation, where C2 is anteriorly displaced on C3 (Fig. 4.7a, b). In some cases the displacement can be so profound that it resembles true injury. In a study of 161 patients, Cattell and Filtzer discovered that 46% of children younger than 8 years old had pseudosubluxation of C2 on C3 on lateral flexion and extension radiographs [1]. In order to differentiate pseudosubluxation from true pathologic change, Swischuk described a differentiating line drawn from the anterior cortex of the posterior arch of C1 to the anterior cortex of the posterior arch of C3 called the *posterior cervical line* (Fig. 4.8a–c) [14]. The anterior edges of the spinous processes of C1, C2, and C3 should line up within 1–1.5 mm of each

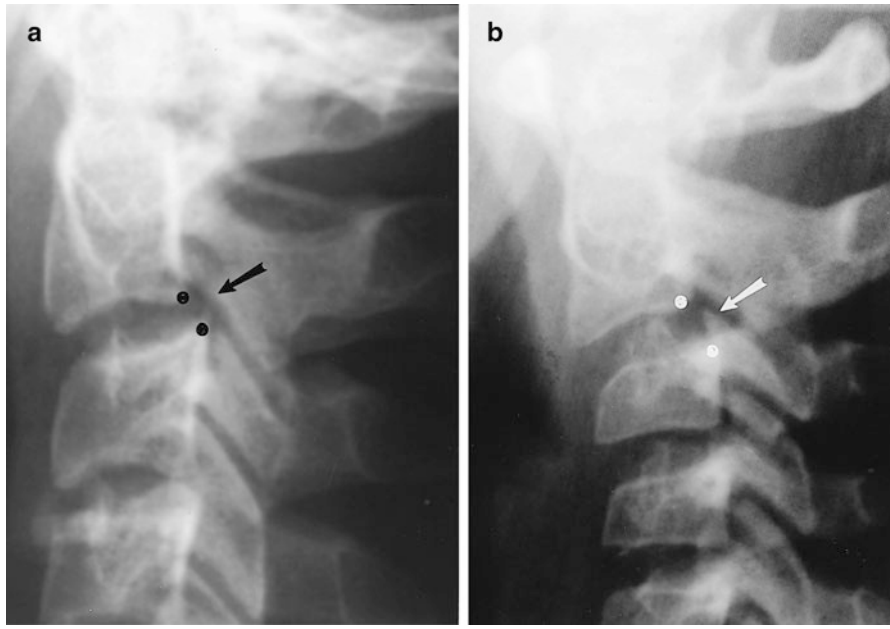

Fig. 4.7 (**a, b**) Physiologic anterior subluxation of C2 on C3. (**a**) Note that C2 is anteriorly displaced on C3 (dots and arrow). (**b**) Another patient with similar findings (dots and arrow). Note that the apophyseal joint faces are parallel (From Imaging of the Cervical Spine in Children by Leonard E. Swischuk. © 2013 Springer Science + Business Media, New York. With permission of Springer Nature)

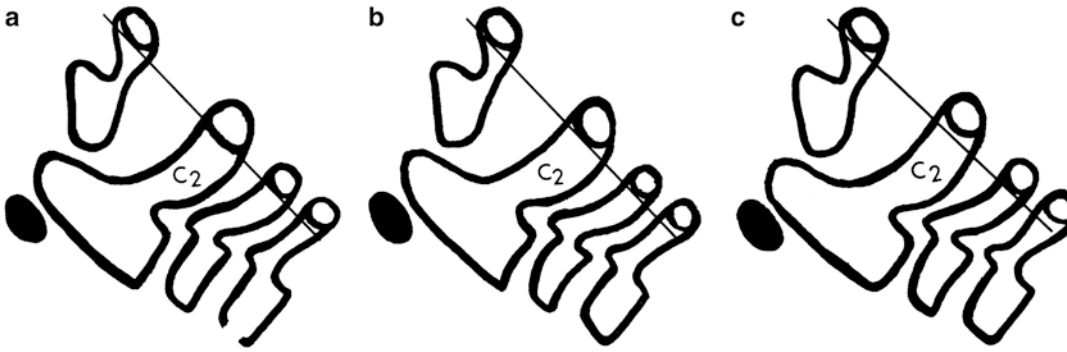

Fig. 4.8 (**a–c**) Posterior cervical line: normal limits. (**a**) The posterior cervical line touches the anterior cortex of C2. (**b**) The line passes through the anterior cortex of the spinous tip of C2. From Imaging of the Cervical Spine in Children by Leonard E. Swischuk. © 2013 Springer Science + Business Media, New York. With permission of Springer Nature. (**c**) The posterior cervical line misses the anterior cortex of C2 by only 1 mm. Used with permission of Leonard E. Swischuk

other on both flexion and extension radiographs [2, 9, 14]. Pathological or abnormal change is evident when the posterior cervical line misses the posterior arch of C2 by 2 mm or more. This should raise concern for true subluxation or bilateral pars fractures of the axis, termed Hangman's fracture.

Pseudospread of the Atlas

In adults, a Jefferson fracture is characterized by abnormal separation of the lateral masses of the atlas with fractures of its anterior and posterior arches. The most common mechanism of injury is vertical compressive force applied to

the top of the head, driving apart the wedge-shaped lateral masses. In children, particularly up to the age of 7, the lateral masses of the atlas may appear laterally offset relative to the lateral masses of the axis, mimicking a Jefferson fracture. Up to 6 mm of displacement is common in children up to 4 years old and may be seen in patients up to 7 years of age. However, this is a normal anatomic variant and almost always does *not* represent any pathological change, as Jefferson fractures in children are extremely rare. In the setting of trauma, without knowledge of the anatomical variants encountered in children, the radiologist may misinterpret this finding. Pseudospread of the atlas is found in most children 3 months to 4 years of age, with a prevalence of over 90% in the second year of life [15]. This developmental phenomenon is thought to arise due to a discrepancy in the growth rate of the atlas vs. the axis, as the former follows a faster neural growth pattern, while the latter follows a somatic growth pattern, particularly during the first year of life [16]. The subsequent disappearance of pseudospread between 3 and 6 years of age indicates a faster pattern of growth during this time. True Jefferson fractures are very rare in children prior to their teens. The reason for this is fourfold: children weigh less, their skulls are more plastic and absorbent of force, their necks are more flexible, and the synchondroses of C1 may serve as an elastic buffer [15].

Posteriorly Tilted Dens

The dens, or odontoid process, is prone to numerous developmental anomalies. Although many anomalies may be bizarre and quite obvious to the physician, the posterior tilting of the dens tends to be more subtle and may be confused for pathological change, including fracture [17]. The normal posteriorly tilted dens represents an anomalous growth of the dens and is not part of a generalized pattern of anomalous growth of the cervical spine. However, posterior tilting of the dens may be severe in some cases and may prove difficult for the radiologist during interpretation of the image (Fig. 4.9a, b). Fracture of the dens usually occurs through the base and can result in either anterior (hyperflexion injury) or posterior (hyperextension injury) tilting of the dens [18]. If

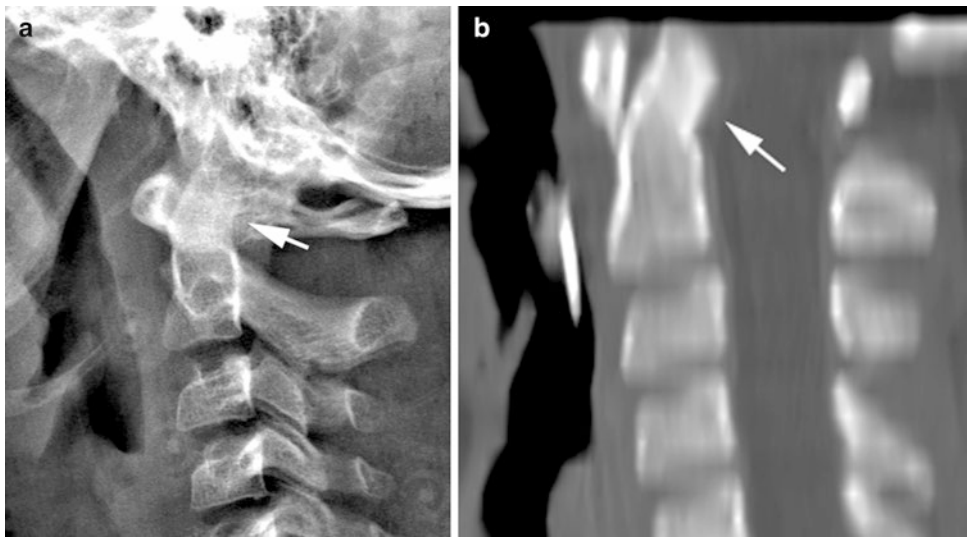

Fig. 4.9 (**a, b**) Posteriorly tilted dens: normal finding. (**a**) Note the posteriorly tilted dens (D). This patient was normal. (**b**) CT study, sagittal reconstruction. Note the posteriorly tilted dens (arrow) (From Imaging of the Cervical Spine in Children by Leonard E. Swischuk. © 2013 Springer Science + Business Media, New York. With permission of Springer Nature)

no fracture line is visible, one should look for posterior displacement of C1, which only occurs if there is a true fracture of the dens [17]. Interpretation is made even more difficult when there is an associated notch-like defect present at the base of the tilted dens. This notch is most likely representative of the former dens-body synchondrosis; however, this finding can make ruling out underlying fracture almost impossible on plain films alone. CT scan would be necessary in these cases [17].

Atlas-Dens Interval

Measurement of the atlas-dens interval (ADI) is a useful tool in evaluating the pediatric trauma patient, as an increased ADI is consistent with transverse ligament injury. The spinal cord is protected from the dens by the transverse ligament, and any injury to the ligament can result in lethal injury to the cord. Therefore, quick and accurate diagnosis is essential. ADI is defined as the distance between the posterior inferior margin of the anterior arch of the atlas and the anterior margin of the dens [19]. In adults normal ADI should be less than 3 mm; however in children ADI can be as high as 3.5 mm [19] or even 5 mm [20], with no evidence of ligamentous injury. If there is continued suspicion of ligamentous injury in the pediatric patient after plain films, MRI should be performed.

Vertebral Body Wedging

Early in infancy the vertebral bodies of the cervical spine have an oval shape, which become more rectangular as the patient becomes older [21]. There is a generally predictable sequence of development from oval to rectangular, particularly of the lower cervical spine. However, the developments of the vertebral bodies of C3 and C4 are less predictable and can be a source of confusion for the physician. In children up to 7 years of age, wedging deformity of the vertebral bodies of up to 3 mm, particularly at the C3 level, is a normal variant and should not be confused with a compression fracture (Fig. 4.10) [9, 21, 22]. The wedging occurs at the anterior superior corner of the vertebral body. The exact cause of wedging is unclear, but it is postulated that hypermobility of the cervical spine in children causes chronic, repetitive impaction of the vertebral body of C3 by C2. If there is doubt as to whether a compression fracture is present, one can perform a CT for verification. In true anterior compression fractures an axial CT scan will reveal fracture lines through the vertebral body. In patients with normal wedging, no fracture line is seen.

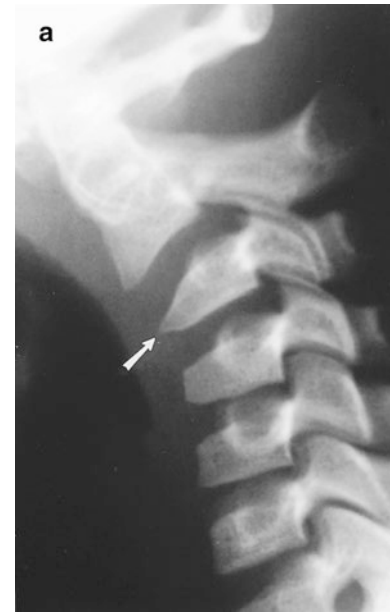

Fig. 4.10 Anterior wedging of C3 with pseudodislocation. Note the pronounced wedging of C2 (arrow). Dislocation at the C2–C3 level might be suggested (Used with permission of Leonard E. Swischuk)

Retropharyngeal Soft Tissue Anomalies

Prominence of prevertebral soft tissues (PVST) on lateral radiographs of the cervical spine in adults is often due to cervical spine injury and subsequent edema and hemorrhage. This finding is also true in children; however, thickening of up to 6 mm of the prevertebral soft tissues at the

level of C3 is considered normal [2]. On lateral radiographs, PVST thickness varies depending on the level at which it is measured. At the level of C2, in children less than 14 years of age, PVST thickness is usually between 4 and 5 mm [23, 24]. At the level of C6, the thickness increases as the patient gets older, with an average thickness of 6 mm in patients younger than 2 years of age and over 9 mm in patients older than 10 years. What may be more important than the measurement of the thickness is the understanding of patient positioning and its effect on PVST thickness. In pediatric patients, widening of the prevertebral soft tissues can be a result of flexion of the neck or expiration when the film was taken. Therefore, in the setting of suspected cervical spine injury in a child, repeat the radiograph with the neck extended and during inspiration. A reversal of this finding supports interpretation of a normal variant and not pathologic change [2].

Absence of Lordosis

Children younger than 16 years of age can demonstrate localized absence of lordosis of the cervical spine when the neck is in a neutral position, a pathological change if seen in an adult [2, 9].

Secondary Ossification Centers

As mentioned above, the ossification centers of the cervical vertebrae in the developing child can lead to misinterpretation of radiographs, particularly in the setting of trauma. In the cervical spine, secondary ossification centers most commonly appear in the spinous processes and the ring apophyses at the superior and inferior border of the vertebral bodies. In the setting of acute trauma, these centers may be confused with fractures. Acute fractures are irregular and nonsclerotic and can occur at unpredictable locations, whereas normal centers of ossification are smooth, regular structures with subchondral sclerotic lines [9]. CT scan shows evidence of hematoma and swelling with fracture.

Assessing Images of the Pediatric Cervical Spine

As the evaluation of the pediatric cervical spine can be difficult owing to the numerous anatomical variants that make it different from the adult, there are a variety of lines, angles, and measurements that have been described to make radiological assessment easier and more accurate. These measurements apply to plain radiographs as well as CT and MRI (Table 4.2).

Wackenheim Clivus Baseline

This is formed by drawing a line along the clivus and extending it inferiorly into the upper cervical spine (Fig. 4.11). Normally, this line lies tangential to the posterior aspect of the dens. It may be disrupted in basilar invagination with atlanto-occipital dislocation and atlantoaxial dislocation [25].

Atlanto-Dental Interval (ADI)

Discussed earlier, the ADI represents the space between the posterior inferior margin of the anterior arch of C1 and the anterior border of the dens. In children younger 8 years old, it can measure up to 5 mm. In children older than 8 years, old it should be less than 3 mm [20]. Increased distance and widening of this space is consistent with atlantoaxial instability and transverse ligament disruption as well as basilar invagination.

Basion-Dens Interval (BDI)/Basion-Axial Interval (BAI)

The BDI is the distance between the basion and the tip of the dens. This can measure up to 12 mm [26]; however, this may not be reliable in young children whose dentes are not yet ossified. Harris et al. described another relationship using the basion-axial interval (BAI). The BAI is the distance between the basion and a line drawn along the posterior aspect of the dens (the posterior

Table 4.2 Normal relationships and radiological measurements at the craniovertebral junction

Line/angle	Anatomic landmark	Normal value	Pathology
Wackenheim clivus baseline	Drawn along posterior surface of clivus, extended inferiorly to upper C-spine	Lies tangential to posterior aspect of dens	Atlanto-occipital dislocation and atlantoaxial dislocation
Atlanto-dental interval (ADI)	Distance between posterior inferior margin of anterior arch of C1 and anterior border of dens	<8 yrs: up to 5 mm >8 yrs: up to 3 mm	Atlantoaxial instability and transverse ligament disruption
Basion-dens interval (BDI)	Distance between the basion and the tip of the dens	Up to 12 mm	Atlanto-occipital dislocation
Basion-axial interval (BAI)	Distance between the basion and line drawn along the posterior aspect of the dens	Up to 12 mm	Atlanto-occipital dislocation
Powers ratio	Ratio of the distance between the basion and the posterior arch of C1 to the distance between the opisthion and the anterior arch of C1 (BC/OA)	Less than 1	Atlanto-occipital dislocation
Condylar gap (Kaufman's method)	Distance between the occipital condyle and facet of atlas	Less than 5 mm	Atlanto-occipital dislocation
Swischuk's line	Line drawn from anterior cortex of posterior arch of C1 to anterior cortex of posterior arch of C3	Less than 2 mm	Hangman's fracture and true spondylolisthesis

axial line). In children younger than 13 years old, the BAI should not exceed 12 mm [27]. A value greater than 12 mm can indicate atlanto-occipital dislocation and basilar invagination.

Powers Ratio

In 1979 Powers et al. described another radiographic tool to determine the relationship between the occiput and the atlas for accurate diagnosis of atlanto-occipital dislocation. This became known as the Powers ratio. The Powers ratio is defined as the ratio of the distance between the basion and the midpoint of the spinolaminar line of C1 (posterior arch) to the distance between the opisthion and the midpoint of the posterior margin of the anterior tubercle of C1 (anterior arch). This ratio should be less than 1, with average being 0.77 [9, 28]. A ratio of more than one is indicative of atlanto-occipital dislocation. Although useful in conjunction with other measurements, by itself the Powers ratio is not helpful in diagnosing longitudinal and posterior dislocation, particularly in children, due to the difficulty in reliably identifying the opisthion [29].

Condylar Gap Method (Kaufman's)

Kaufman's method is defined as simply measuring the gap between the occiput and the atlas, known as the atlanto-occipital junction. In children younger than 15 years of age, the distance should not exceed 5 mm at any point in the joint

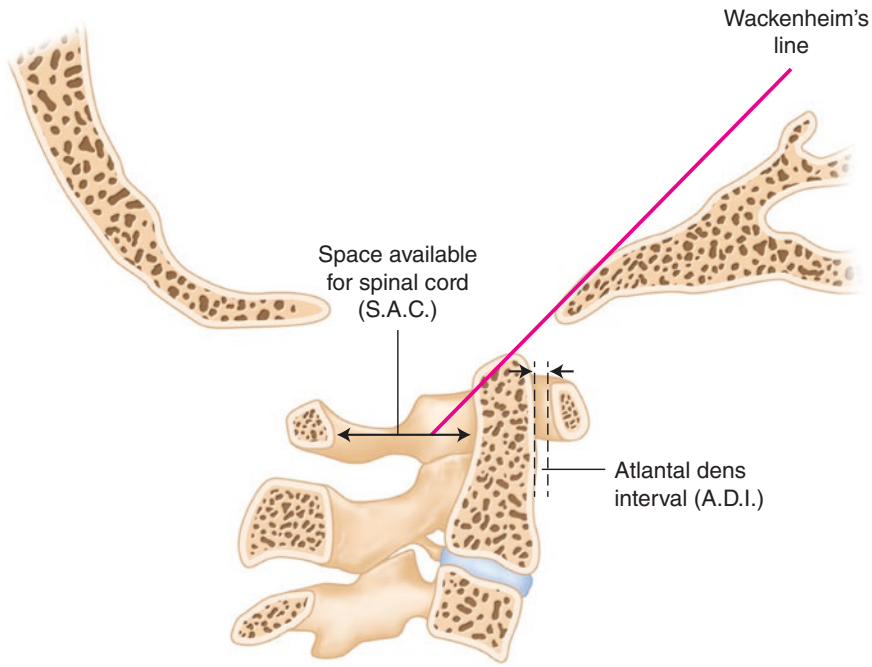

Fig. 4.11 Wackenheim's line is a line extending along the posterior surface of the clivus downward and posteriorly. The tip of the dens should lie below and anterior to this line

[30]. Widening of the atlanto-occipital junction raises concern of a distraction-dislocation injury at the craniovertebral junction [9, 30].

Pediatric Cervical Spine Injuries

Pediatric patients younger than 8 years old are susceptible to ligamentous injuries involving the upper cervical spine, with 87–100% of injuries occurring at C3 level or higher [31]. In children over the age of 8 years, nearly 80% of injuries are fractures involving the lower C5–C6 cervical level [32]. The incidence of injuries to the cervical spine in children younger than 11 years is estimated to be 1.19 per 100,000, compared with adolescents, who have a reported incidence of 13.24 per 100,000 [31]. Sixty percent of injuries occur in boys, with motor vehicle accidents, falls, and sports injuries being generally causative [33]. The mortality associated with these injuries varies from 4–41% [34]. Neurological deficits, concomitant head injuries, and significant abdominal injury may be associated with up to 67% of these injuries, underlying the importance of accurate and timely diagnosis.

Atlanto-Occipital Dislocation (AOD)

Atlanto-occipital dislocation is rare and is associated with a mortality rate of 50% [35–37]. Deployment of motor vehicle airbags has been identified as a risk factor for this injury in children. Many of the surviving children have complete loss of neurological function below the brain stem and are ventilator dependent. Occasionally, some children present without any neurological deficit, and MRI is necessary to detect the injury to the tectorial membrane and alar ligaments if plain radiographs or CT scans are inconclusive (Fig. 4.12) [38]. It is important to understand that the Powers ratio assessment may miss these distraction injuries, in addition to

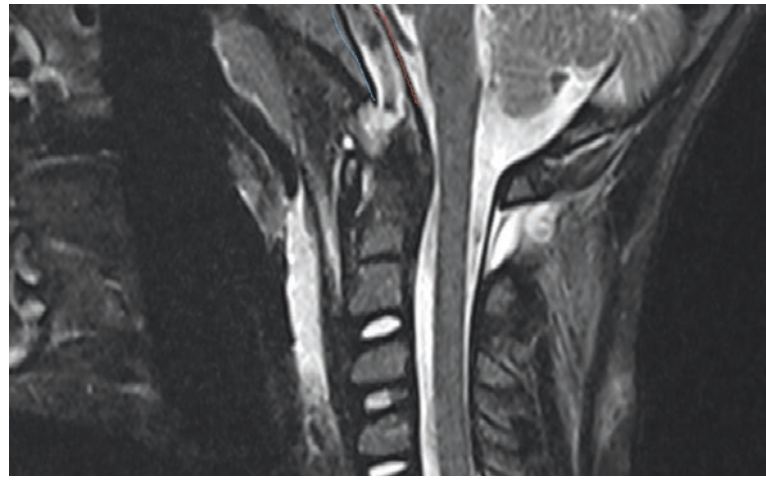

Fig. 4.12 T2-weighted sagittal MRI of a 6-year-old boy injured in a car accident. Note the elevation of the tectorial membrane off the clivus. The apical ligament is disrupted, and there is posterior ligamentous disruption. Image courtesy of Jonathan Phillips MD

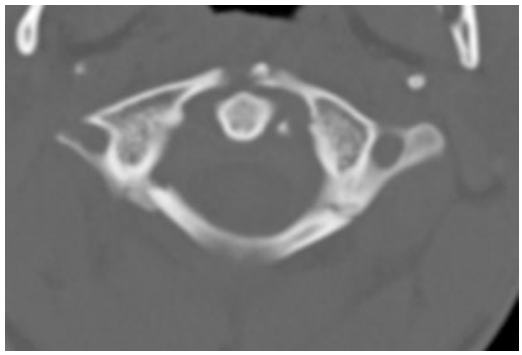

Fig. 4.13 Axial CT of Jefferson fracture in 15-year-old male injured in American football. Note an anterior fracture, two posterior fractures, and an avulsion fracture just lateral to the dens. Image courtesy of Jonathan Phillips MD

posterior atlanto-occipital injuries and anterior atlanto-occipital dislocations that have spontaneously reduced. As a consequence, Kaufman's gap method is now regarded as the most sensitive technique to diagnose this injury [39–42].

Atlas Fracture

The Jefferson fracture, or burst fracture of C1, was first described in 1920 and is an uncommon pediatric injury, with less than 20 cases in the literature [43, 44]. It is caused by axial loading and is more common in adolescents (Fig. 4.13). In contrast to adults, who may present with comminution, children may present with an isolated fracture of a synchondrosis [9]. If the fracture is undisplaced, the diagnosis can be difficult even with CT scans, and an MRI scan may be required. As discussed previously, lateral mass displacement up to 6 mm, or "pseudospread," may be seen in children up to 7 years old. Offset greater than 6 mm is suggestive of transverse ligament disruption, and further imaging is indicated [15]. MRI can also help visualize the transverse atlantal ligament which, if compromised, represents an unstable injury [45, 46].

Odontoid Process (Dens) Fracture

In children, fracture of the odontoid process is the most common fracture of the cervical spine, with the subdental and neurocentral synchondroses being the most susceptible to injury (Fig. 4.14) [47–49]. Unfortunately, the persistence of the subdental synchondrosis in children up to 10 years old may delay the diagnosis of this injury, and adjuvant imaging either with CT or MRI scans is often necessary to confirm this fracture. Typically in odontoid process fractures, the dens is tilted posteriorly on the lateral plain radiographs, and concomitant PVST swelling

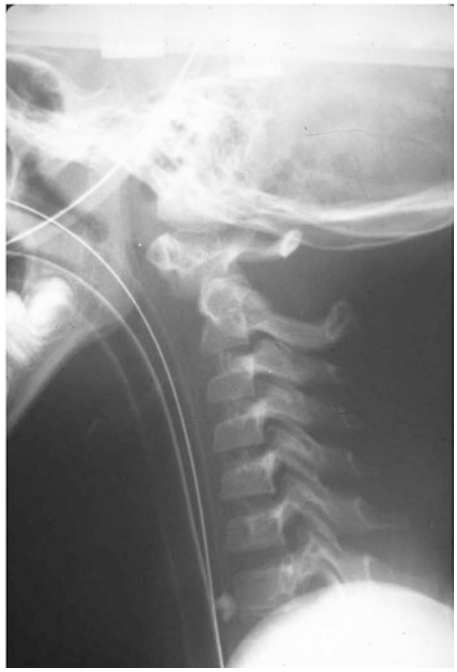

Fig. 4.14 A 7-year-old female with a type II dens fracture sustained on a trampoline. Image courtesy of Jonathan Phillips MD

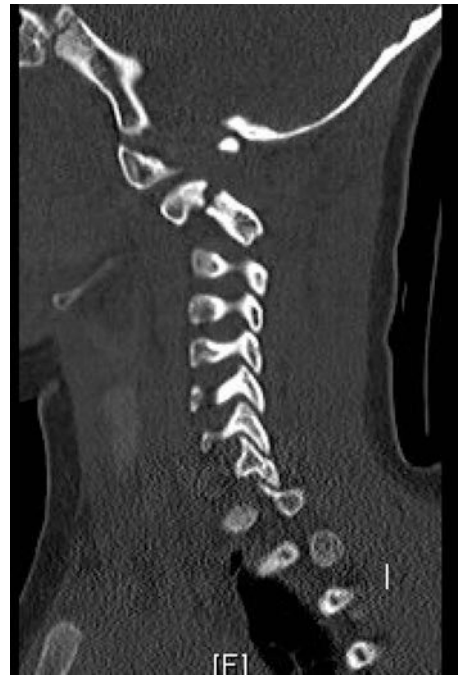

Fig. 4.15 Hangman's fracture in a 6-year-old female sustained by head impact on a waterslide. CT scan with sagittal reformat. Note the flexion position of the fracture, not the classical extension position attributed to the Hangman's noose. Image courtesy of Jonathan Phillips MD

facilitates the diagnosis of this injury. Open-mouth views tend to be of little diagnostic value, as the fracture line tends to run through the epiphyseal plate and in a different plane to the radiograph [50–52]. Delay in diagnosis may compromise the vascularity to the fracture site and result in the development of an os odontoideum [53].

Hangman's Fracture

Hangman's fracture is classically described as a hyperextension injury of the cervical spine with bilateral pars fractures of the axis, horizontal tearing of the C2–3 disk, and anterior displacement of C2/C3 level [46, 54]. This unstable injury usually results from abrupt deceleration of the head and is visible on lateral cervical spine radiographs in approximately 90% of cases [55]. Hangman's fracture is most commonly confused with pseudosubluxation of the C2/C3 level. Displacement greater than 2 mm is pathognomonic of a true spondylolisthesis, and Swischuk's posterior spinolaminar line is very useful to differentiate between these two entities (Fig. 4.15) [14].

Atlantoaxial Rotary Fixation

Atlantoaxial rotary fixation may occur spontaneously, post inflammation, post-minor trauma, or may occur with congenital anomalies. At initial presentation, atlantoaxial rotary fixation may be indistinguishable from muscular torticollis using plain radiographs. Clinically, torticollis resolves spontaneously within a few days or weeks, whereas true rotary fixation does not unless treated. Using CT scans, Kowalski et al. have described a technique to distinguish the two conditions on initial presentation. Atlantoaxial rotary

fixation on plain radiography demonstrates a rotated appearance of C1 on C2, with asymmetry of the distance between the odontoid and C1 lateral masses [56–58]. Using a functional examination with CT, in which patients turned their head to the maximum contralateral rotation, Kowalski et al. were able to demonstrate reduction and reversal of rotation of C1 on C2 in patients with torticollis. Patients with atlantoaxial rotary fixation demonstrated no motion at C1–C2 during this maneuver [58].

SCIWORA

SCIWORA was first defined by Pang and Wilberger as "spinal cord injury without evidence of radiological abnormality" [59]. As previously discussed, due to certain biomechanical and anatomical factors, children younger than 8 years old are more susceptible to spinal cord injury than older children. As the name implies, initial plain radiographs are normal. An MRI scan is indicated in patients who have had normal plain films or CT and who have exhibited transient or persistent neurological symptoms [54]. In patients with SCIWORA, an MRI will show signs of spinal cord injury, including increased signal intensity on T2-weighted images representing edema and decreased signal intensity representing areas of hemorrhage.

Radiographic Assessment of Trauma

In pediatric trauma patients, imaging is frequently necessary, due to the difficulty in detecting cervical spine injuries based on history and physical examination. Indeed, the American College of Surgeons advocates plain radiographs of the cervical spine in all children as part of the initial major trauma evaluation. The standard trauma series for a pediatric trauma patient includes a cross table lateral (CTL), anteroposterior neck, and anteroposterior open mouth. While numerous radiographic anomalies may be present in children, 85% of cervical spine injuries may be visualized on the CTL view alone. Ninety-eight percent of cervical spine injuries may be identified by completing the trauma series [60, 61].

However, many pediatric trauma centers are now routinely utilizing CT instead of plain radiographs to assess the cervical spine because CT has reduced the time to diagnosis of injury and the time to disposition from the emergency department and is more cost-effective than plain radiographs in moderate- to high-risk adult trauma patients. CT has also provided more adequate imaging, with only 1.4% of CT scans needing repeat imaging, in comparison with 30% of plain radiographs [51]. While CT can induce neoplasia, particularly of the thyroid gland, modern "high-end" pediatric trauma CT systems have strategies that reduce the CT dose by adhering to the ALARA (as low as reasonably achievable) principles [47]. Pediatric radiologists can significantly decrease the scanner's radiation dose by a factor of 4–5 by modulating the fixed tube current. It is also worth remembering that the radiation dose associated with a typical adult CT scan (1–14 mSv) is comparable to the annual dose received from natural sources of radiation, such as radon and cosmic radiation (1–10 mSv), or the equivalent dose received from a single transatlantic flight [35].

MRI plays a significant role in the diagnosis of cervical spine injuries. MRI is better than plain radiographs and CT scan at visualizing soft tissue injury and identifying intervertebral disk herniation, ligamentous injury, and spinal cord compression. The American Academy of Neurological Surgeons (AANS) recommends incorporating MRI to exclude spinal cord or root compression. Flynn et al. highlighted the utility of MRI in cervical spine trauma by identifying unrecognized ligamentous and soft tissue injury in nearly 25% of patients with normal plain radiographs that necessitated cervical orthoses [53]. MRI has also been demonstrated to reduce time to clear the cervical spine, leading to a reduced inpatient hospital stay and significant cost saving per patient [53].

Summary

The unique formation of the children's cervical spine, particularly at C1 and C2, results in significant variations in radiographic appearances as compared with adults. Knowledge of these variations and the complex physeal patterns is important in interpretation of routine radiographs. Dynamic studies, such as flexion and extension films and advanced imaging, are often required to distinguish pathological conditions from normal.

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Part II

Clinical Aspects of Disorders of the Child's Cervical Spine

Clinical Presentation and Physical Examination of Children with Cervical Spine Disorders

5

William C. Warner and Ilkka Helenius

Introduction

Evaluation of the pediatric cervical spine presents a unique challenge for the treating physician. The cervical spine in infants, children, and adolescents has different characteristics than those of an adult cervical spine. A variety of diseases and congenital anomalies may affect the pediatric cervical spine. Knowledge of the anatomy, growth, and development of the pediatric cervical spine is required to aid in the understanding of these conditions. Multiple anomalies of the upper cervical spine are common within a single patient, so when a single anomaly is seen, other anomalies should be sought. An average of 3.4 cervical spine osseous anomalies per patient has been reported [1]. In addition, an adequate examination in children may be difficult, particularly in those who are preverbal, nonverbal, frightened or in pain, or are unable to describe their symptoms or localize pain. Knowledge of known associations of cervical spine anomalies and instability patterns seen in pediatric patients with syndromes, skeletal dysplasias, and congenital anomalies is needed because these are not often seen in the adult population.

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Why the Immature Spine Is Different

Developmental Anatomy

At birth, the atlas is composed of three ossification centers, one for the body and one for each of the neural arches. The posterior arches usually fuse by the age of 3 years; however, occasionally the posterior synchondrosis between the two arches fails to fuse, resulting in a bifid arch. The neurocentral syncondroses that connect the neural arches to the body close by 7 years of age [2]; the internal diameter of the atlas reaches its final size by this time, although the external diameter continues to grow through appositional bone growth [3].

The axis develops from at least four separate ossification centers: one for the dens, one for the body, and two for the neural arches. The body is connected to the adjacent lateral masses by the neurocentral syndondrosis and to the dens by the dentocentral synchondrosis. The dentocentral synchondrosis closes by the age of 6–7 years, although it may persist as a sclerotic line until 11 years of age. The neural arches of C2 fuse at 3–6 years of age. Occasionally, the tip of the

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odontoid is V shaped (dens bicornum), or a small separate summit ossification center may be present at the tip of the odontoid (ossiculum terminale) [4, 5].

The third through seventh cervical vertebrae share a similar ossification pattern: a single ossification center for the vertebral body and an ossification center for each neural arch. The neural arch fuses posteriorly between the second and third years and the neurocentral syncondroses between the neural arches, and the vertebral body fuses by 3–6 years of age. These vertebrae are normally wedge shaped until 7–8 years of age (Fig. 5.1) [6].

Anatomic and developmental factors in a young child result in unique characteristics of the pediatric cervical spine. Children younger than 8 years of age tend to have increased neck motion compared to adults [7, 8]. This may be the result of children younger than 8 years of age having more horizontally oriented facet joints. These become more vertical with growth. The facet orientation from C5 to C7 increases from approximately 55–70° during growth. The facet joint orientation in the upper cervical spine changes from 30° of vertical orientation to 70° of vertical orientation during growth [7, 8]. Infants and younger children also have generalized ligamentous laxity and weaker neck muscles compared to older children, allowing for increased range of motion and decreased muscular resistance to these increased ranges of motion [8]. Children between the ages of 3 and 8 years have 50% more flexion and extension at C2–C3 than adults. The center of rotation of the cervical spine in children varies with age, with the level of greatest motion descending with age (C2–4 is most mobile between 3 and 8 years of age, C4–C5 between 9 and 11 years, and C5–C6 between 12 and 15 years) [9, 10]. This is the reason that adolescents present with adult patterns of injuries, and younger children present more often with craniocervical injuries.

The head in younger children accounts for a greater proportion of the body weight than in older children and adolescents. This is part of the reason for the higher center of rotation of the cervical spine in younger children. The proportionately larger head with respect to the body in young children can place the cervical spine in flexion if the child is immobilized on a standard backboard. A board with an occipital recess for

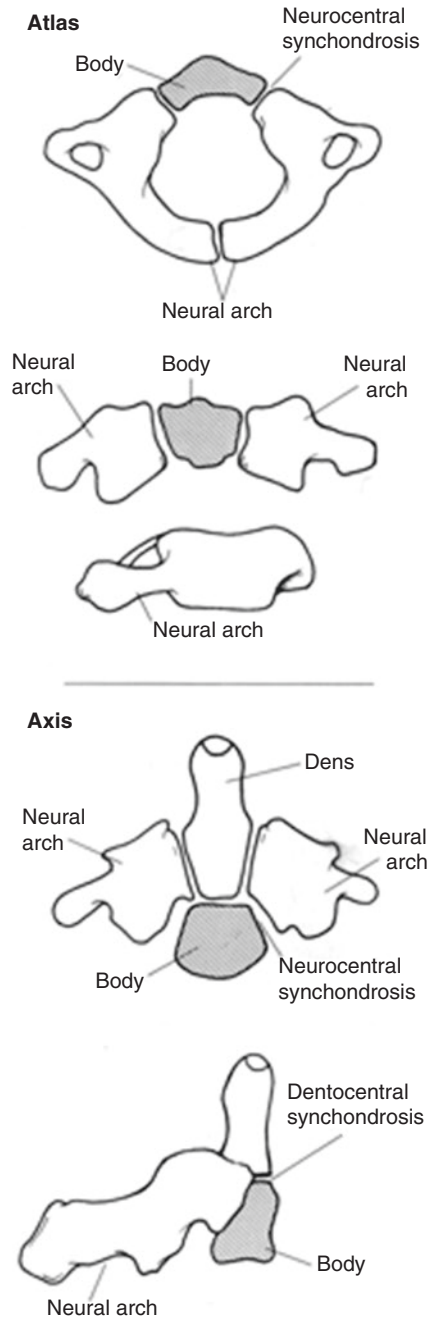

Fig. 5.1 Ossification of the atlas and axis (Reproduced from Copley and Dormans [59]; with permission of WoltersKluwer.

the head places the cervical spine in a neutral position (Fig. 5.2) [10, 11].

History

Evaluation of the pediatric cervical spine should always begin with a thorough history and physical examination. The "chief complaint" will guide the treating physician in regard to the history and physical examination. A chief complaint can range from deformity, pain, neurological deficits, delay in developmental milestones, or trauma. These are clues as to the underlying etiology of the cervical spine problem.

A chief complaint of deformity, usually described as a head tilt or asymmetry of the face and neck, leads the examining physician to explore the reasons for the deformity (Fig. 5.3). Congenital muscular torticollis is the most common cause of deformity and head tilt in an infant. Muscular torticollis usually is noticed by the parents shortly after birth. Often they report a nodule or palpable mass in the neck during the second to sixth weeks of life.

Ballock and Song found that almost 20% of their patients with torticollis had nonmuscular causes [12]. The diagnosis of nonmuscular causes of torticollis or cervical spine deformity is probably one of the most important aspects in treatment.

Deformity that comes and goes or is intermittent may be consistent with ocular torticollis or Sandifer's syndrome (torticollis caused by gastroesophageal reflux).

Osseous causes for deformity can be the result of congenital anomalies such as Klippel-Feil syndrome, a congenital osseous fusion of two or more segments of the cervical spine. If Klippel-Feil syndrome is suspected, the examiner should ask about other anomalies, such as in the heart, genitourinary, auditory, or musculoskeletal systems. A history of prolonged neonatal care and operative treatment of a tracheoesophageal fistula and esophageal atresia should be recorded [13–17]. Occipitalization and congenital hemiatlas also can be causes for nonpainful osseous deformity of the cervical spine [18].

Painful causes for cervical deformity or acquired torticollis may be traumatic or inflammatory in origin or may be the result of a tumor. Atlantoaxial rotatory subluxation or fixation is the most common cause of painful acquired torticollis. This occurs in otherwise healthy children after minor cervical trauma or upper respiratory tract infection (Grisel's syndrome) [19–22]. Other inflammatory conditions, such as juvenile rheumatoid arthritis, may present early in the course of the disease with neck pain and painful torticollis [23]. Infectious discitis and calcific discitis also can result in painful torticollis [24]. Posterior fossa tumors account for about 50% of intracranial tumors in infants and may present as deformity with pain in the upper cervical spine. Occipital headaches can be an initial finding in posterior fossa tumors (Table 5.1).

When the chief complaint is a neurological deficit, then the history should include discovery of any occult trauma, instability, or any other cause that may compress or compromise the spinal cord or brainstem. Neurological deficits can be subtle, such as a delay in developmental milestones, easy fatigability, or clumsiness, or they can be obvious such as motor and sensory deficits or complete paralysis. Brainstem or upper cervical spine anomalies should be looked for when a

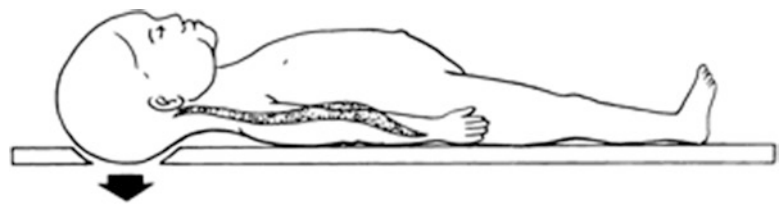

Fig. 5.2 Occipital recess in backboard, placing the pediatric cervical spine in neutral position (Reproduced with permission of JBJS Inc., from Herzenberg et al. [11])

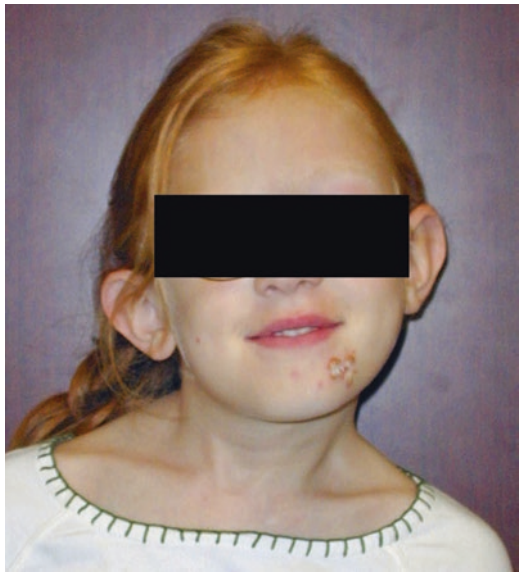

Fig. 5.3 Head tilt deformity

patient has difficulty with respiration or cannot be weaned from a ventilator.

A history of a difficult or breech delivery that is associated with delayed motor development, hypotonia, or later hypertonia may indicate a perinatal injury to the upper cervical spine [25, 26].

Certain syndromes and skeletal dysplasias are known to have upper cervical spine anomalies (Table 5.2) [27–29]. Thus, clinical recognition relies on the knowledge of a high incidence of cervical instability in these conditions such as Morquio disease [30–33] and other mucopolysaccharidoses [34, 35], Q22 deletion syndrome, spondyloepiphyseal dysplasia [36, 37], diastrophic dysplasia [38–42], Down syndrome [43–46], and Larsen syndrome [47]. Children with skeletal dysplasia and associated cervical spine instability present with short stature, and mapping the child's growth on a chart may be helpful in detecting any growth abnormalities if characteristic features of the involved syndrome are not present. Patients with Larsen syndrome and diastrophic dysplasia typically have midcervical kyphosis. This may resolve in patients with diastrophic dysplasia if the cervical kyphosis remains below 60° [39, 40]. In contrast, patients with Larsen syndrome may develop spinal cord compression and quadriparesis early during infancy [47].

Table 5.1 Differential diagnoses in torticollis

Congenital	Congenital muscular Vertebral anomalies Failure of segmentation Klippel-Feil Occipitalization of C1 Failure of formation Congenital hemiatlas Combined failure of segmentation or formation Ocular
Acquired, painful torticollis	Traumatic Atlantoaxial rotatory displacement Os odontoideum C1 Fracture Inflammatory Atlantoaxial rotatory displacement (Grisel) Juvenile rheumatoid arthritis Discitis or osteomyelitis Other infection in the neck Tumors Eosinophilic granuloma Osteoid osteoma or osteoblastoma Calcified cervical disk Sandifer syndrome
Acquired, nonpainful torticollis	Paroxysmal torticollis of infancy Tumor of the central nervous system Posterior fossa Cervical cord Acoustic neuroma Syringomyelia Hysterical Oculogyric crisis (phenothiazine toxicity) Associated with ligamentous laxity Down syndrome Spondyloepiphyseal dysplasia or mucopolysaccharidoses (MPS) dysplasia

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Respiratory difficulty may be the result of cervical or foramen magnum stenosis or basilar impression. This is seen in conditions such as mucopolysaccharidosis [35], achondroplasia [48–50], and osteogenesis imperfecta [51, 52]. It is important to be familiar with basilar impression and its presentation because this spinal deformity often goes unrecognized or is misdiagnosed as a

Table 5.2 Cervical spine disorders in patients with skeletal dysplasias

Syndrome	Associated cervical spine disorders
Achondroplasia	Stenosis Craniocervical joint Subaxial spine
Diastrophic dysplasia	Kyphosis Hypoplasia vertebral body Hypotonia Spina bifida
Hunter syndrome	Stenosis Cervical canal Thickened dura Posterior odontoid
Hurler syndrome	Dysplasia Odontoid Malformation Tip of odontoid
Kniest dysplasia	Instability Atlantoaxial Occipitoatlantal
Metaphyseal chondrodysplasia	Instability Atlantoaxial Ligamentous laxity
Morquio syndrome	Instability Atlantoaxial Hypoplasia odontoid Os odontoideum Thickened dura
Spondyloepiphyseal dysplasia	Instability Atlantoaxial Hypoplasia odontoid Os odontoideum Ligamentous laxity
Spondylometaphyseal dysplasia	Instability Atlantoaxial Hypoplasia odontoid Ligamentous laxity

posterior fossa tumor, bulbar palsy of polio, syringomyelia, amyotrophic lateral sclerosis, spinal cord tumor, or multiple sclerosis. The clinical presentation varies, and symptoms are usually weakness, fatigue, and paresthesia in the limbs. Ataxia, dizziness, and nystagmus may be present. Involvement of the lower cranial nerves also may occur. Compression of the vertebral arteries as they pass through the foramen magnum is another source of symptoms [53] and may consist of dizziness, seizures, mental deterioration, and syncope.

Trauma also may result in neurological deficits. If a neurological deficit is noted after minor trauma, an underlying osseous or occult instability syndrome should be suspected [54]. In major trauma, associated injuries are common. Facial and cranial injuries are the most frequent. Other musculoskeletal injuries and, most importantly, other noncontiguous spine injuries can occur and often are missed if not specifically sought.

Upper cervical spine injuries are more common in children younger than 8 years of age. Cervical spine injuries become similar to adult patterns as the child grows into adolescence.

Physical Examination

A thorough and systematic examination should be performed when a cervical spine disorder is suspected. Multiple physical examinations at different times may be needed in young children who are not cooperative or are nonverbal, to obtain an accurate and reliable examination. When possible, the examination of an infant should be done in a warm examination room in a quiet and calm atmosphere. It usually is helpful to examine infants and toddlers in the lap of a parent or caregiver. In a child who is just old enough to speak, the examination may be complicated by his inability to follow directions or to describe or understand relevant signs secondary to the physical examination; for example, the patient may not understand what numbness means when asked.

The initial portion of the examination consists of observation. This may be the most important part of the examination, especially in infants and children who often are not cooperative during examination. The examiner should note the shape of the neck and hairline. The classic clinical presentation of Klippel-Feil syndrome is the triad of a low posterior hairline, a short neck, and limited neck motion (Fig. 5.4). The face and skull should be inspected for facial asymmetry or plagiocephaly.

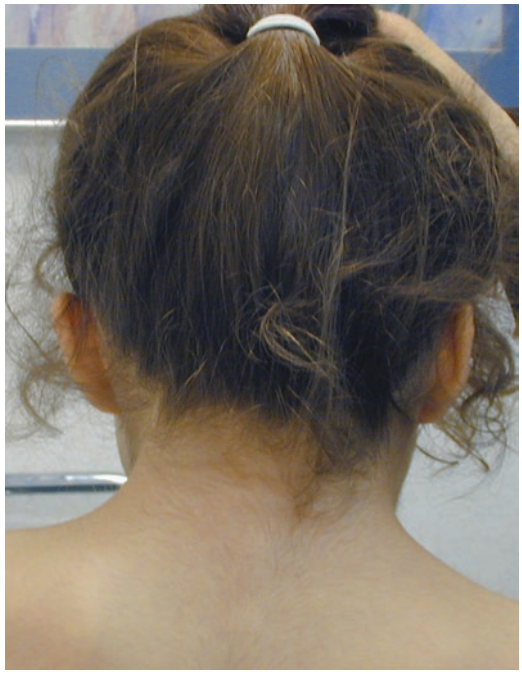

Fig. 5.4 Low posterior hairline in patient with Klippel-Feil syndrome

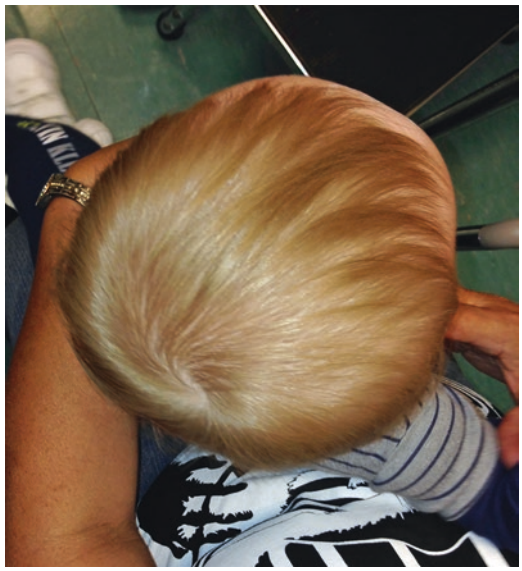

Fig. 5.5 Plagiocephaly can be easily seen observing the child from above

The examiner should observe the child from the front, rear, and top while the patient is supine or seated in the caregiver's lap. This will help in detecting minor degrees of asymmetry that otherwise may not be apparent (Fig. 5.5). Any tilt or rotation of the head in relationship to the body should be noted. Watching the child's head and neck movements while he is tracking a toy or an object will give an indirect measurement of the range of motion of the neck. A "hanging head" posture (forward flexion of the neck) may be indicative of a tumor in the brainstem or cervical spine [55, 57]. Evaluation of the gait in an ambulatory child is important, and any gait abnormalities, posturing, or abnormal position of the upper extremities exhibited while walking should be noted.

Palpation of the head and neck is the next portion of the examination. The head should be palpated for the presence of any masses or defects. The anterior fontanel of infants should be palpated, looking for any fullness that may indicate occult hydrocephalus. Premature closure of the fontanel may suggest craniosynostosis; the anterior fontanel normally closes between 4 and 26 months of age. Plagiocephaly usually indicated long-standing torticollis. The posterior cervical region should be palpated for any masses or areas of tenderness. Any tenderness or paraspinal muscle spasms should be noted. The spinous processes are palpated for deviations or defects. The length of the cervical spine is estimated. The C2 spinous process is large and is therefore the uppermost spinous process that can easily be palpated.

Next the anterior neck should be evaluated for any areas of swelling or tenderness. The sternocleidomastoid muscle should be palpated for spasm, fibrosis, or masses. Pseudotumors of the sternocleidomastoid muscle often are seen in congenital muscular torticollis during the second and sixth weeks of life. The neck should be checked for both active and passive range of motion. Active flexion and extension, rotation, and lateral bending should be evaluated. Seventy-five degrees

of horizontal rotation to both sides, extension of 80°, flexion to 60°, allowing the chin to touch the chest, and lateral bending of 45° [56] are approximate normal active ranges of motion in children (Fig. 5.6a–e). Passive range of motion also should be gently measured, and any guarding, pain, or limitation of motion should be noted.

The cranial nerves may be involved in upper cervical spine disease, such as basilar impression, Chiari malformation, or any upper cervical spine congenital anomalies or instability, and should be tested. Cranial nerves III, IV, and VI control ocular function such as extraocular movements and pupillary reaction to light. Cranial nerve IV (trochlear) specifically controls the superior oblique muscles of the eyes, which can be used as a clinical finding. Cranial nerve V (trigeminal) controls corneal reflex and facial sensation. Cranial nerve VII (facial) controls both motor power and sensation in the face. It can be tested by observing facial symmetry during smiling or crying. A hearing screening examination will test cranial nerve VIII, and cranial nerves IX and X can be tested by evaluating the patient for hoarseness, deviation of the uvula, and gag reflex. Cranial nerve XI is a spinal accessory nerve that controls motor power to the trapezius muscle. Assessing shrug strength of the shoulder will test this nerve. Cranial nerve XII (hypoglossal) can be evaluated by listening to the patient's pronunciation of words and by observing the tongue for protrusion, asymmetry, or deviation [57].

Motor and sensory testing of the upper and lower extremities may be challenging in an infant or child. Observation of the child walking, running, crawling, or playing may provide clues regarding neuromuscular function of the upper and lower extremities. Having the child play with toys and reach and grab objects overhead also will give the examiner an idea of muscle strength and tone. Muscle bulk and tone should be assessed for atrophy or any asymmetry. Hypertonicity and spasticity should be looked for in the upper and lower extremities. Observing the patient's gait and checking the patient's ability to toe-and-heel walk will serve as a screening test for plantarflexion and dorsiflexion strength, as well as balance.

Testing of the C1–T1 nerve roots also should be done. Sensation of the occipital area is provided by the C1–C3 nerve roots and should be recorded. The C4 nerve root innervates the levator scapulae, trapezius, and rhomboid, and if affected, causes difficulty in elevating the shoulder (Fig. 5.7a, b). Testing biceps and wrist extension motor strength provides information about the C5 and C6 nerve roots. The C5 nerve root innervates the axillary nerve and provides sensation over the lateral deltoid muscle, and the C6 nerve root provides sensation over the lateral forearm, thumb, and index finger. C7 motor function is tested by triceps and wrist flexion strength, and C7 sensory function is tested by sensation over the middle finger (not always reliable). Finger flexor strength tests C8 motor function, and the sensory test for C8 is over the medial forearm and the ring and middle fingers. Motor testing of the interosseous muscles and sensory testing of the medial portion of the arm provide information about the T1 nerve root (Fig. 5.8).

When examining the cervical spine, a neurological evaluation of the lower extremities also is necessary and should include hip flexion strength for T12–L3; quadriceps strength for L2–L4; tibialis anterior muscle strength, patellar tendon reflex, and sensation in the medial aspect of the foot for L4; extensor digitorum longus muscle strength and sensation of the dorsum of the foot for L5; peroneal muscle strength, Achilles tendon reflex, and sensation on the lateral side of the foot for S1; and perianal sensation for S2–S4 [57].

Imaging

Radiographic evaluation should include lateral, anteroposterior, and open-mouth odontoid views of the cervical spine. Lateral flexion and extension

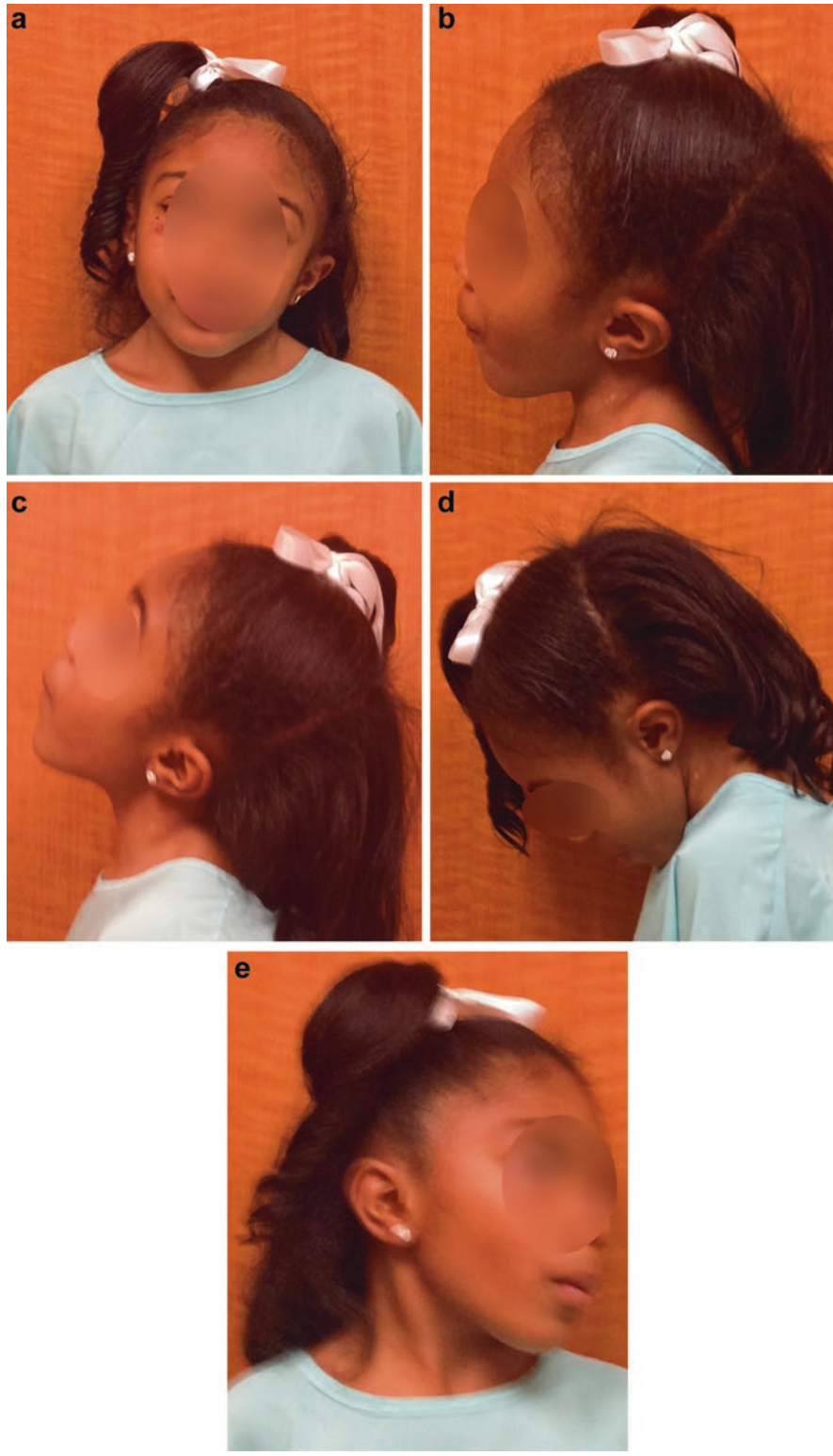

Fig. 5.6 (**a**, **b**) Examination of range of motion in child with head tilt. (**c**) Extension, (**d**) flexion, (**e**) rotation

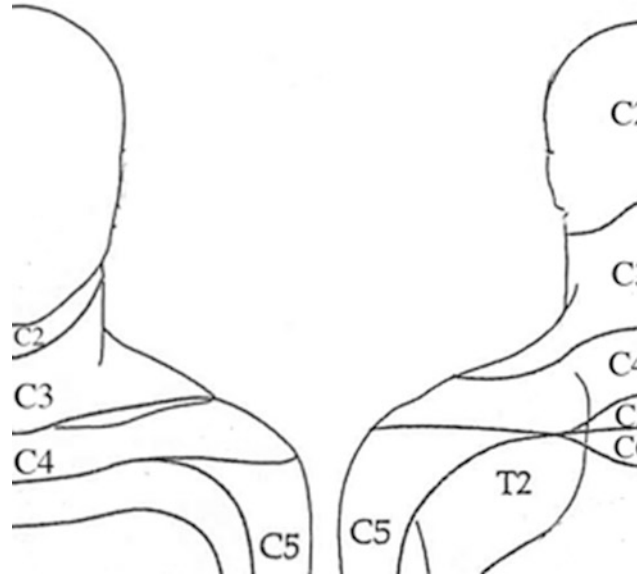

Fig. 5.7 (**a**, **b**) C1 through C5 nerve root dermatomes: (**a**) anterior, (**b**) posterior

radiographs that are supervised by the treating physician may be helpful in evaluating cervical instability. Further imaging studies that include CT or MRI evaluation of the cervical spine often are invaluable in defining cervical spine anomalies and spinal cord or soft-tissue pathology about the cervical spine. Radiographic and further imaging of the cervical spine will be covered in another chapter.

Electrodiagnostic Studies

Nerve conduction studies and electromyography can sometimes be of diagnostic help in patients with cervical spine pathology. These electrodiagnostic test may be helpful in differentiating any peripheral nerve compression or pathology from any central or spinal cord pathology, although obtaining these test in a child or adolescent may be difficult.

Nelson has reported the use of short-latency somatosensory evoked potentials in the management of upper cervical spine problems in patients with achondroplasia. Obtaining this study on an outpatient basis in an awake child may be challenging and therefore limits its clinical usefulness [58].

Summary

Clinical evaluation of a child presenting with cervical pathology requires a comprehensive history and careful examination. Depending on the age of the child, cooperation may not be good. Subtle clues in the history and repeated physical examinations using "tricks" to gain insight into the physical signs are important. A thorough knowledge of comorbidities in the various congenital, syndromic, and traumatic etiologies will ensure that no important symptoms or signs are missed.

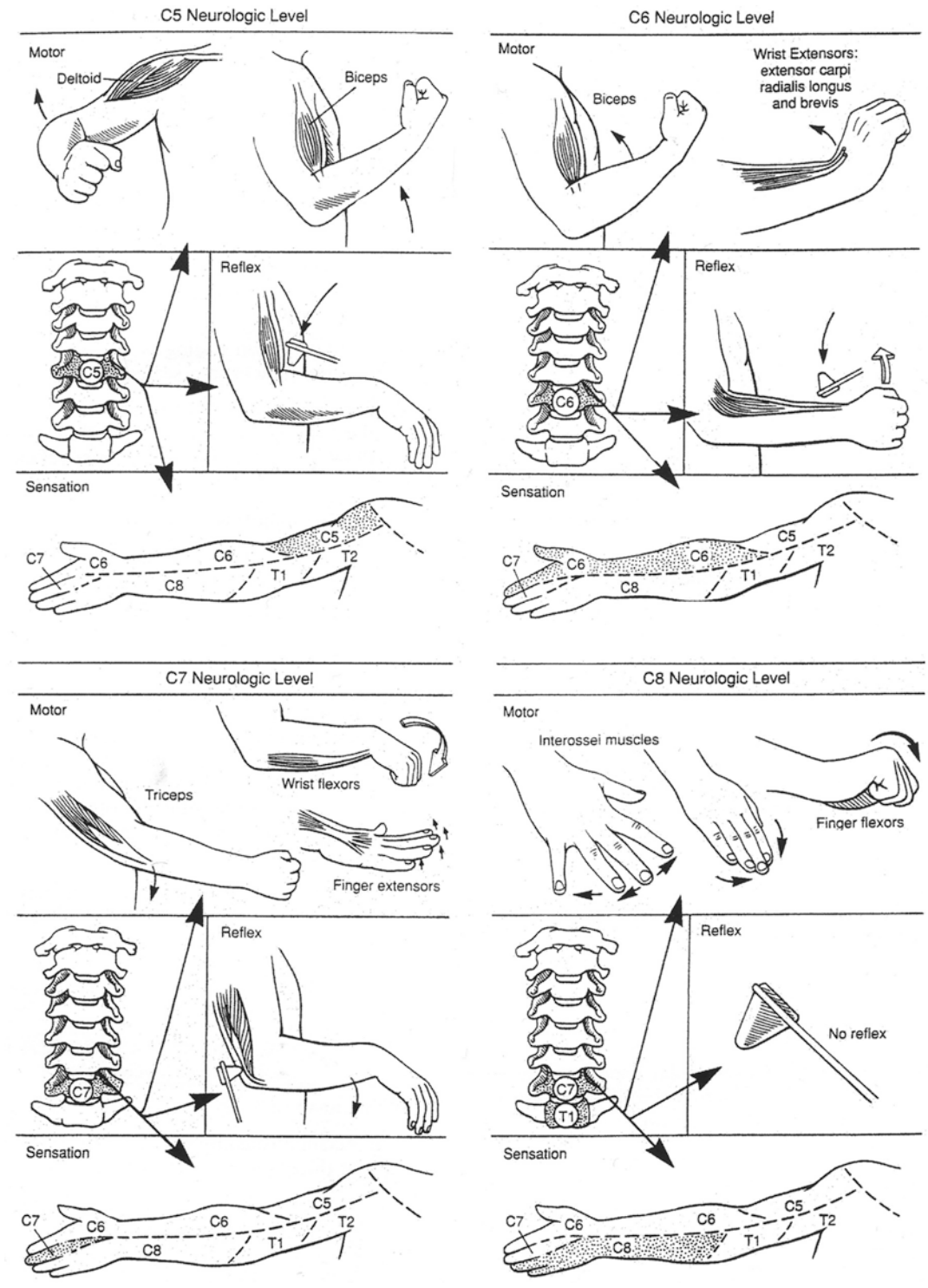

Fig. 5.8 Neurological evaluation of the upper extremity (C3–T1) (Reproduced with permission of Wolters Kluwer Health, from Deyo [60])

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Pediatric and Adolescent Cervical Spine Trauma

6

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Part I: Patient Evaluation

Introduction

While cervical spine injury in the pediatric trauma population is rare, the diagnostic and treatment challenges entailed can be formidable.

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With an incidence of approximately 1–2%, pediatric cervical spine trauma is relatively infrequent but may have devastating consequences if not diagnosed and treated in a timely manner [1, 2]. Cervical spine trauma constitutes 80% of spine trauma in the pediatric patient, compared to 30–40% in adults [1]. The average age of the pediatric cervical spine trauma patient is 10–11 years, and 60% of injuries occur in boys. The incidence of cervical spinal injury increases after the age of 9. Compared to children 9 years of age or younger, cervical spine injuries are two-fold more common in preadolescents and five-fold more common in adolescents [3]. Patients younger than 8 years old most commonly have injuries which involve the upper cervical spine, compared with adolescents whose injury patterns resemble those of adults [1, 4]. The incidence of upper cervical spine injuries (C1–C4) compared with lower cervical spine injuries (C5-C7) is approximately 70% vs. 25% in children younger than 9 years. In adolescents, upper cervical injuries account for approximately 40% of cervical fractures, while subaxial injuries account for 45% of fractures [3].

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Mechanism of Injury

The most common mechanism of injury for children and adolescents is a motor vehicle trauma (44–68%) [4], predominantly passengers in motor vehicle crashes, with approximately one-half of victims injured while unrestrained or inappropriately restrained [1, 5, 6]. The mechanisms of cervical spine injury vary according to the age of the patient. Birth injuries account for a very small percentage of cervical trauma related to difficult delivery (1 in 60,000) [7], often requiring the use of a vacuum or forceps. Infants and toddlers are susceptible to cervical spine injury from child abuse and falls. In older children and adolescents, pedestrian or bicyclist versus motor vehicle trauma, motorcycle and ATV accidents, gunshot wounds, and sports-related injuries are more common etiologies [5]. Of the sports activities (16% of total cervical spine injuries) [6], collision sports such as football and rugby, cheerleading, and diving are the highest risks for cervical spine trauma.

Cervical Spine Characteristics: Child Versus Adult

Anatomical

Important anatomical and physiologic differences exist between the cervical spine of a child younger than 8 years of age and an older child or adolescent whose cervical spine is structurally similar to that of an adult. The younger child exhibits greater neck range of motion (ROM) and underdeveloped neck musculature, is relatively ligamentous lax, has an increased cartilage-to-bone ratio within the spinal column, and has more horizontally oriented facets. Additionally, the size of the head compared to the size of the torso is proportionally larger in young children compared to those older. As a result of these differences, children younger than 8 years of age are more susceptible to upper cervical injuries, with one large study showing that 87% of injuries in this population occurred at C3 level and above [4]. By contrast, the injury pattern of older children and adolescents is similar to that seen in adults, with the majority of injuries occurring in the lower cervical (subaxial) spine.

Radiological

Evaluation of pediatric cervical spine radiographs can be challenging due to developmental changes during growth (Fig. 6.1a–e). Pseudosubluxation at C2–C3, and less frequently at C3–C4, is often encountered in the pediatric cervical spine. Examination of the spinolaminar line, drawn from the posterior arch of C1–C3, provides guidance in appropriate alignment of the upper cervical spine. A distance of >1.5 mm between the line and the posterior arch of C2 suggests a true injury. The atlanto-dens interval (ADI) is the distance between the anterior aspect of the dens and the posterior aspect of the anterior ring of the atlas. This measurement ranges from 3 to 5 mm in approximately 20% of children younger than 8, while the normal distance in adults should be ≤3 mm. While easy to overlook, attention must also be paid to the soft tissues. The retropharyngeal space, which can appear increased in the setting of trauma due to soft tissue edema, is variable in children compared with adults. The retropharyngeal space is typically <7 mm, and the retrotracheal space is <14 mm in children [4]. This may be difficult to assess in the distraught child, but any abnormality should heighten suspicion for cervical spine injury.

The multiple growth centers and incomplete ossification at different ages can cause confusion and be mistaken for fractures. The atlas is formed from three ossification centers, the two lateral masses and one for the body, which does not appear until 1 year of age, leading to an apparent absence of the anterior arch. The axis is comprised of five ossification centers, the two lateral masses, the body, and the two halves of the dens. The basilar synchondrosis separates the odontoid from the body and can be mistaken for a fracture until its fusion around age 5–6 years (Fig. 6.2a–c). Unfused ring apophyses anteriorly and secondary ossification centers of the spinous processes can also appear suspicious for fractures. The physes can be differentiated from the typical fracture lines by their smoother appearance and predict-

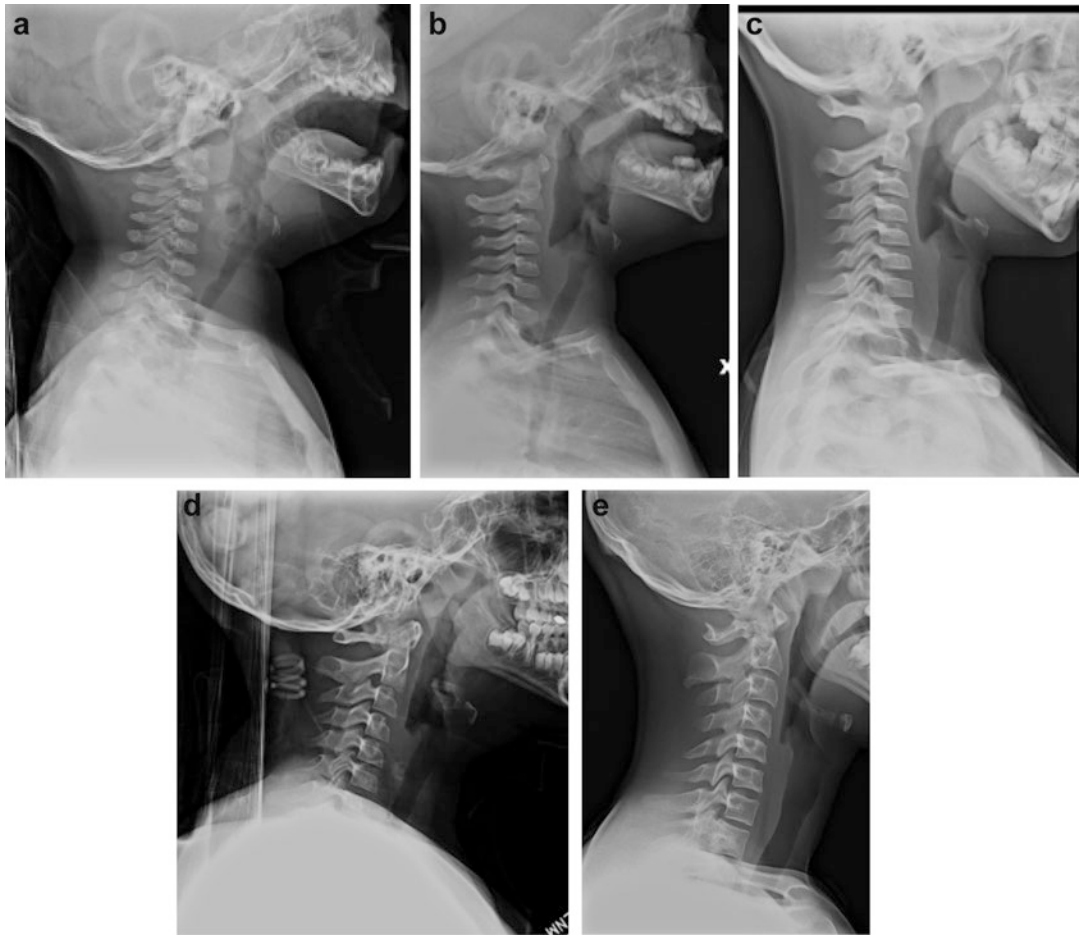

Fig. 6.1 (**a**) Lateral radiographs as children skeletally mature. Age 6 months, (**b**) 2 years, (**c**) 6 years, (**d**) 9 years, (**e**) 16 years

able locations. Morphology of the vertebral bodies also changes through maturation, transitioning from oval to rectangular shaped. Anterior wedging of the vertebral bodies ≤3 mm is within normal limits and not necessarily a compression fracture [1].

SCIWORA

In 18–38% of pediatric cervical spine injuries [1, 4, 8], the patient may exhibit myelopathic signs in the setting of trauma without any evidence of fracture or ligamentous instability on plain radiographs or CT. This phenomenon is termed spinal cord injury without radiographic abnormality (SCIWORA) . There is a higher incidence in younger children, likely reflecting the underdeveloped nature of the pediatric spine. Increased elasticity and range of motion at the bony articulations allow greater movement, while the spinal cord remains tethered cephalad by the brain, caudad by the cauda equina, and at the nerve plexuses. Children may present with neurologic injury ranging from transient paresthesia to complete injury, with mild to moderate injuries having the greatest potential for recovery [9]. Where plain radiographs and CT may fail, MRI often elucidates the soft tissue and spinal cord injury, which may correlate with neurologic injury. However, 30–35% of children with SCIWORA have no evidence of spinal cord injury on MRI [10].

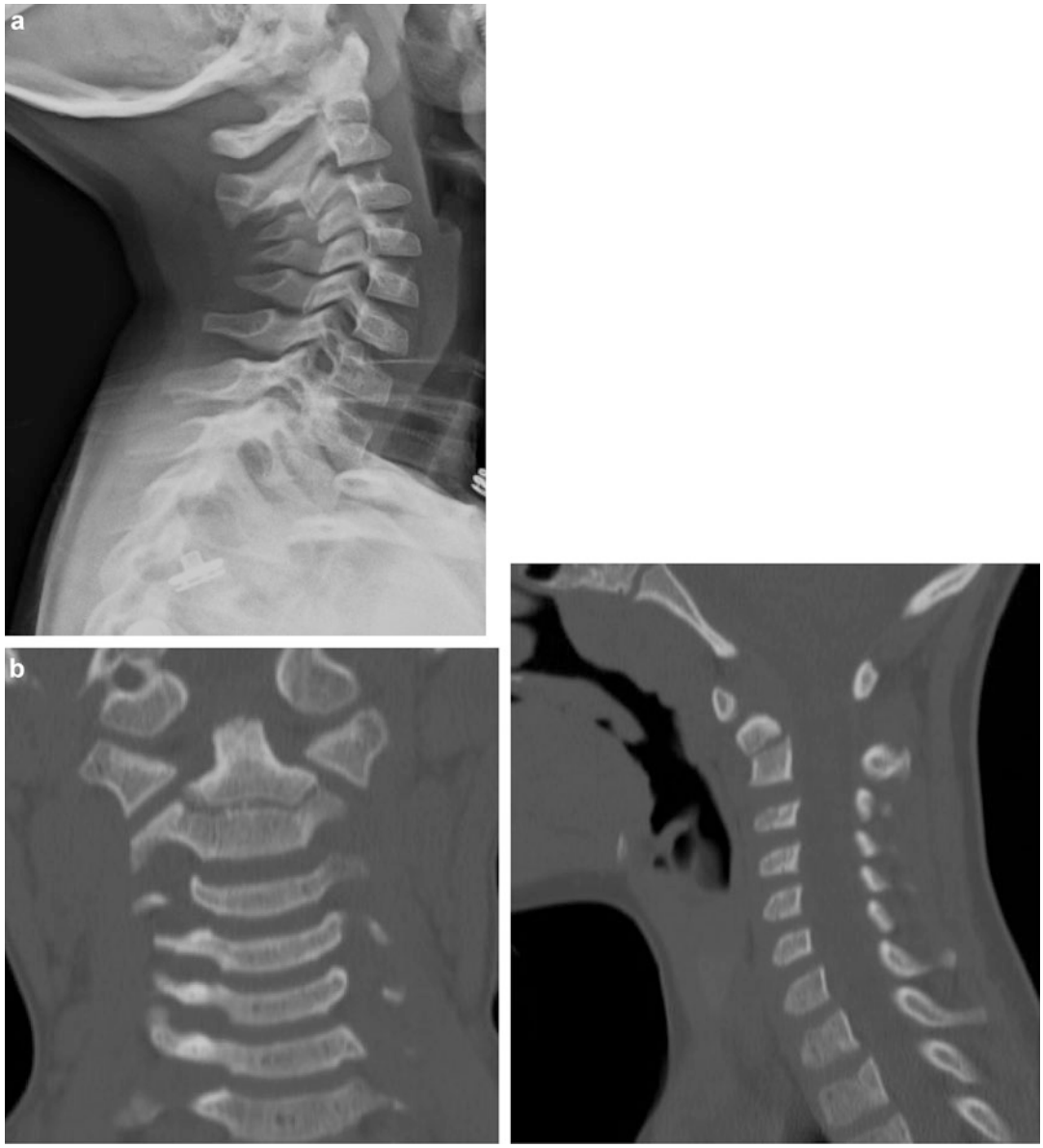

Fig. 6.2 (**a**) A 7-year-old boy in a motor vehicle accident complaining of neck pain. Lateral cervical view was interpreted as possible fracture of C2 versus a normal synchondrosis. (**b**) CT clarifies the diagnosis of a minimally displaced fracture through the synchondrosis of C2. (**c**) Normal appearance of C2 in a 7-year-old

Concomitant Injuries

Due to the significant forces imparted by the mechanisms of injury associated with cervical spine trauma, approximately 60% of patients with cervical spine fractures sustain concomitant injuries, as listed below [11]. In one large study of associated injuries occurring in children with spinal trauma of any type, including 36% with cervical spine fractures, pulmonary injuries such as contusions, rib fractures, and pneumothoraces were most common. While brain injuries and facial trauma were seen in all groups, children ages 3 years and younger were more likely to sustain these injuries compared to older children and adolescents. Neurologic injury, most commonly spinal cord injury, was more likely to occur in the youngest patients as well. Other associated injuries, especially in older children

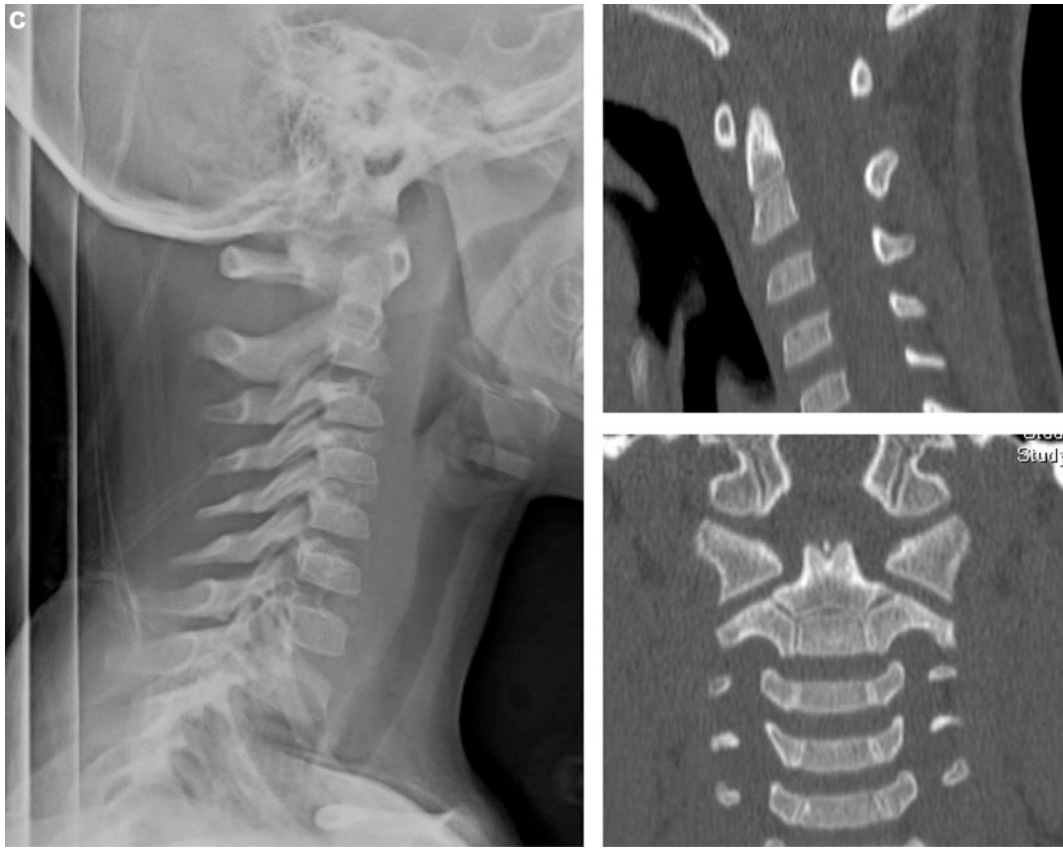

Fig. 6.2 (continued)

and adolescents, include abdominal trauma and orthopedic injuries, most commonly upper extremity fractures. The possibility of multiple spinal fractures must also be considered. As many as 45% of spinal injuries in children and adolescents are multilevel [5, 12, 13], and 6–17% are noncontiguous, most commonly a combination of cervical and thoracic spine fractures [5].

Top Four Concomitant Injuries Seen in Patients with Cervical Spine Trauma

1. Traumatic brain injury
2. Pulmonary contusion, rib fracture, and pneumothorax
3. Abdominal injuries
4. Fractures of the upper extremity

Mortality

While most cervical spine injuries are not life-threatening, mortality in patients with cervical spine injury ranges from 4% to 41%, though most commonly found to be 16–17%. [1, 4]. Mortality is correlated with the level of cervical injury, ranging from 17% at C1 to 3.7% at C4, with occipito-atlantal dislocation and complete spinal cord disruptions being the most common lethal injuries. The cause of death in children and adolescents, however, is infrequently the cervical spine injury. Severe concomitant traumatic brain injury is the most common reason for death in patients who present with cervical spine trauma and is seen in as many as 40% of trauma patients who die with a cervical spine injury.

Spinal Cord Injury (SCI)

The incidence of spinal cord injury (SCI) is relatively rare, ranging from 0.65% to 10% of all reported spinal injuries, peaking at ages less than 5 years and greater than 10 years [14, 15]. At ages ≤6 years, the rate of injury is similar between boys and girls; however, in older children boys are two to three times more likely than girls to sustain spinal cord injury [16]. The level of spinal cord injury also varies significantly with age. Two-thirds of spinal cord injuries in patients younger than 6 years of age occur at the thoracic, lumbar, or sacral segments, resulting in paraplegia; however, more than half of spinal cord injuries in older children and teenagers occur in the cervical segment, resulting in quadriplegia [16, 17]. SCIWORA accounts for up to 67% of all spinal cord injury in children, most commonly affecting children under 5 years of age, and results in a complete cord injury in 40% of cases [18, 19].

Thirty-five to 60% of cervical spine-injured patients sustain neurologic injury [1]. A neurologic injury may be difficult to diagnose promptly in the pediatric patient due to difficulty with exam and history or the subtly of the injury itself. Motor vehicle collision is the most common cause of spinal cord injury in all children older than 1 year [17, 20, 21]. In all age groups, paraplegia from the spinal cord injury is most common after a motor vehicle collision injury, although with increasing age, the rate of tetraplegia from these collision injuries increases [17]. Among teenagers, sports-related injuries are the second most common cause of SCI, similar to that of cervical spine injury, including football, ice hockey, horseback riding, and diving [17, 22]. Violence from firearms, stabbing, and assault are common causes of SCI, particularly for minority groups [17].

Birth trauma and child abuse are the most common causes of SCI in children younger than 1 year. SCI secondary to birth trauma (Fig. 6.3) has been reported as the result of difficult cephalic or breech delivery, with distraction and hyperextension of the head and neck resulting in injury to the spine [23]. The infant and young child who presents with an SCI or any traumatic injury to the spinal column not consistent with the mechanism of history provided by the caregiver should be carefully evaluated to rule out child abuse.

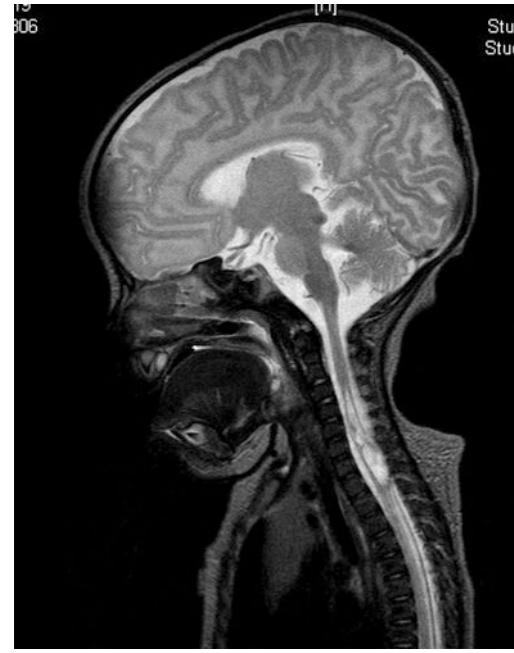

Fig. 6.3 A 3-month-old with history of difficult delivery. Note the spinal cord lesion at the C3-T1

Emergency Management

Prehospital Care

In the pediatric trauma patient with a high index of suspicion for cervical spine injury, strict immobilization must be initiated in the field and maintained until an injury to the cervical spine has been ruled out. The goal of immobilization is to maintain spinal alignment and stability to protect the spinal cord from injury. Prevention of neurologic injury in the intact patient and prevention of further spinal cord damage in the neurologically compromised patient with a cervical spine injury are critical factors in the outcomes of these injuries. Ideally, an appropriately sized cervical collar is applied at the scene of the accident or injury prior to transportation to the hospital. For patients with obvious deformity of head-neck alignment, gentle positioning of the head into an anatomically aligned position prior to collar

placement may be necessary. For those patients wearing athletic equipment such as shoulder pads or a helmet, removal of equipment acutely is not recommended to prevent inadvertent flexion of the cervical spine. Instead, the face mask part of the helmet should be carefully removed to allow access to the airway while the head is stabilized. If the properly sized collar is not available, any movement of the patient at the scene by one of the first responders must be performed with the head held aligned with the torso (manual in-line stabilization) until a reliable method of external spine immobilization can be applied prior to transport.

After application of a cervical collar, the patient is then secured with straps on a spine board that is the length of the patient from the head to the feet to facilitate transfer of the patient to the hospital and protect the entire axial spine if contiguous injuries have occurred. In a patient younger than 8 years of age, immobilization on a flat spine board should be avoided. Because of the proportionally larger head, a child lying supine on this type of spine board will lie with the cervical spine in flexion, potentially compromising the airway and worsening spinal cord compression if a cervical spine injury is present. Instead, immobilization is best achieved on a spine board with an occipital cutout or on a traditional spine board that has additional padding 2–3 cm thick that elevates the shoulders and thorax. These modifications place the cervical spine in a more normal sagittal alignment of slight extension. The stability of cervical spine immobilization on a spine board with a cervical collar in place may be augmented by placing head and neck blocks or sandbags, secured by taping [24].

Emergency Room Resuscitation

The initial evaluation of all pediatric trauma patients involves a thorough adherence to ATLS protocol. The ABCs of trauma should be addressed and cardiovascular stabilization achieved as priority one. For patients with a compromised airway or with respiratory failure, intubation is ideally performed with the cervical spine immobilized in a collar or while manual in-line stabilization is applied. Axial traction is not recommended during stabilization of the spine for intubation. In a patient with an unstable fracture, axial traction has been shown to cause distraction at the injury site. Manual ventilation may be necessary for those patients with a spinal cord injury. Placement of two large-bore intravenous catheters facilitates cardiovascular resuscitation. Simultaneous, a primary survey is undertaken to identify concomitant life-threatening injuries such as a pneumothorax, abdominal injury, sites of severe hemorrhage, and obvious long-bone fractures.

Patient Evaluation

History

The physician evaluating the awake patient ideally should inquire about the details of the pertinent past medical history (Fig. 6.4), mechanism of injury, the specific area of neck pain and its duration, limits of cervical motion or paracervical muscle spasm, and neurological symptoms such as difficulty breathing, arm and leg tingling or weakness, and altered walking ability or loss of coordination. For patients with altered mental

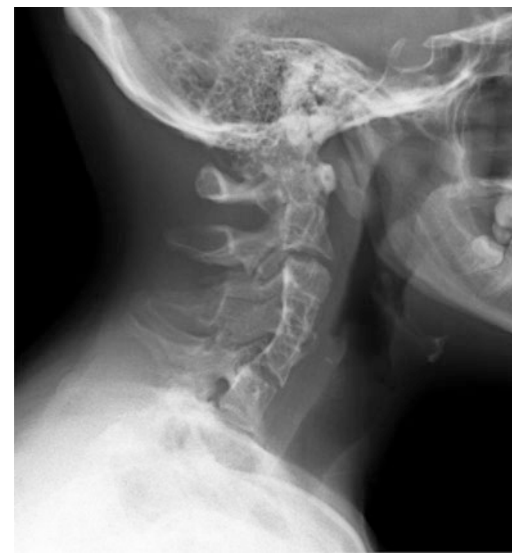

Fig. 6.4 A 10-year-old with neck pain after a fall from the bleachers. The patient had a past history of "dysplasia" and one kidney. The diagnosis was Klippel-Feil syndrome, underscoring the importance of taking a history in all trauma patients

status, gathering information about the accident or injury from family members or emergency transport personnel also provides useful information for the care provider, especially if high-energy mechanisms such as a motor vehicle accident or a fall from a significant height are the causes of injury. All victims of high-energy trauma, regardless of the mechanism described, should be initially treated as if he or she has a cervical spine injury with spinal precautions until an evaluation is completed.

Physical Exam

The exam begins with careful inspection of the head, neck, and torso for signs of trauma to the cervical area such as skull and facial trauma, lap belt marks, abnormal chest movements, abrasions, or contusions. If a cervical collar had been placed in the field, it should be removed for the examination and then replaced after its completion. The neck is assessed for posterior spinal step-offs or abnormal head positioning, such as torticollis, and is palpated along the midline and the paracervical muscles for tenderness or spasm. In the awake patient, the physician then requests that the patient actively move the neck, noting limitations of flexion-extension, lateral bending, and rotation, as well as any particular movement that is especially painful or worsens spasm. Passive assessment of cervical motion is not recommended in the acutely injured but awake patient because of the potential to cause discomfort or possibly even further injury. In the patient with altered mental status, assessing active motion may be difficult, and possibly harmful, if the patient's behavior is aggressive or unpredictable and should be avoided. Assessing range of motion passively in the obtunded patient is dangerous and should never be done, potentially causing further injury if an unstable cervical spine fracture or spinal cord injury has occurred.

A thorough neurological exam should be performed on all patients with a potential cervical spine injury. Assessment of cranial nerve function, upper extremity sensation and motor strength, and upper extremity reflexes provides basic information about nerve root function of the upper cervical spine. Acute clinical manifestations of a spinal cord injury include respiratory failure with no obvious pulmonary injury, generalized flaccidity, bradycardia with hypotension, absence of sacral reflexes, and urinary retention. If a spinal cord injury is suspected, complete dermatomal sensory testing and motor root strength testing must be documented and repeated to identify evolving changes. For a more in-depth discussion of SCI assessment and emergency management, please see the chapter elsewhere in this book.

Radiological Evaluation

AP, Lateral, and Odontoid Views

Initial imaging for most patients with suspected isolated cervical spine injuries is plain radiographs. The complete cervical spine trauma series includes the AP, lateral, and open-mouth odontoid views. On an adequate lateral view, the most critical view, alignment and angulation are assessed. Using the spinolaminar line, proper alignment can be determined, and any listhesis or subluxation can be identified and differentiated from pseudosubluxation. Relationships of the atlantoaxial and atlanto-occipital joints are measured using Power's ratio, the rule of 12, basion-axial interval (BAI), and basion-dental interval (BDI). Power's ratio is measured using the distance from the tip of the basion to the posterior arch of the atlas divided by the distance from the opisthion to the posterior aspect of the anterior arch of the atlas. Atlanto-occipital dislocation may be suspected in a ratio >1.0. Other lines and relationships including BAI/BDI, McGregor, McRae, and Wackenheim can all be useful as reference to assess for injury. Angulation may be difficult to assess in the normal child's cervical spine due to ligamentous laxity inherent in younger children; however angulation >11° in the sagittal plane is suspicious for injury [4]. The usefulness of the open-mouth odontoid view for diagnosing injuries, especially in children younger than 9 years of age [25], has been called into question. Obtaining an adequate radiological view is difficult in a child who has neck pain, is uncooperative, or is unable to understand instructions. Additionally, the open-mouth view, and

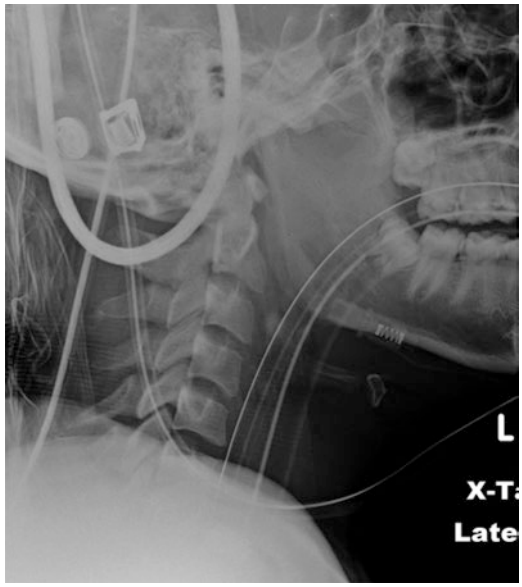

Fig. 6.5 Inadequate lateral radiograph in a 12-year-old with a head injury and possible cervical spine fracture. The entire cervical spine is not visualized from the occiput to the endplate of C7, and overlying structures obscure the details. CT is indicated to evaluate the cervical spine

cervical radiographs in general, can be difficult to interpret in younger children with incomplete ossification of the cervical spine (see above).

Flexion-Extension Lateral Radiographs

Lateral cervical flexion-extension radiographs are useful to assess segmental instability from congenital abnormalities, fracture, or ligament disruption. In the acute setting, however, neck pain and muscular spasm may inhibit any pathologic motion at an injured site, creating a false-negative examination [5]. In our practice, flexion-extension views are used only in delayed fashion, ordered a minimum of 2–3 weeks post-injury after the neck pain and spasm have subsided, to assess for latent instability.

Computed Tomography (CT)

At many institutions, including our own, CT is used primarily to assess trauma patients with a potential cervical spine injury. At others, CT of the upper cervical spine from the occiput to C2 is combined with plain radiographs of the cervical spine to make the primary assessment [4, 8, 25]. This shift has occurred because of two important reasons: (1) In the setting of level one trauma centers, many patients are evaluated with CT to identify head injuries, thoracoabdominal injuries, and pelvic injuries. Including a CT of the cervical spine, while associated with some increased radiation risk, is a more expeditious way of evaluating the cervical spine compared to plain radiographs. (2) CT is potentially a more reliable modality for identifying cervical spine injuries in children and adolescents [26–28]. In a comparison between CT and plain radiography, one study reviewed 413 high-risk pediatric patients. CT diagnosed 71 of 74 cervical spine injuries, where plain radiography found only 50 of 74 [27]. Additionally, CT is commonly used in conjunction with plain radiography when the three-view cervical spine series has not adequately imaged the entire cervical spine (Fig. 6.5) and is inconclusive, when the radiographs are normal but a high suspicion for cervical injury exists, or when a fracture has been identified and requires more detailed analysis to fully delineate the fracture pattern and extent of injury. For patients younger than 5 years of age who have incompletely ossified cervical spines and for those who have sustained non-osseous injuries such as ligament disruptions or SCIWORA, CT may, however, potentially miss some diagnoses. CT is a more definitive diagnostic modality for adolescent patients whose injuries more closely resemble adult patterns, which are in most case osseous in nature and more readily diagnosed by CT [1].

Magnetic Resonance Imaging (MRI)

In theory, MRI is an ideal modality for evaluating the cervical spine in the pediatric spinal trauma patient because it can simultaneously identify fractures and osseous injuries, soft tissue and ligamentous disruptions, and spinal cord and nerve root trauma without radiation risks. Several limitations, however, make it impractical to use MRI currently as the primary diagnostic modality for assessing acute cervical spine injury. MRI is a highly sensitive imaging modality with a relatively high false-negative rate and low specificity [29] compared to CT and plain radiographs, making the clinical significance of some of the findings

questionable. While MRI has a much greater sensitivity for soft tissue and ligamentous injuries than CT and plain radiography, its usefulness for assessing osseous injuries is limited and not as reliable as CT for diagnosing fractures [29, 30]. Additionally, difficulties in obtaining MRI compared to CT in an emergency setting, including the need for sedation or anesthesia for young or uncooperative patients who cannot remain motionless for the MRI, the prolonged length of time for the study compared to CT, and the excessive cost of MRI and its limited availability, are other factors that make its use prohibitive for trauma patient evaluation in most institutions.

MRI is primarily used in many institutions as an adjunct modality to plain radiographs and CT when evaluating cervical spine injuries acutely. For patients exhibiting neurological deficits, MRI imaging may identify abnormalities in the spinal cord indicative of trauma as well as disc, cartilage, or bone that is impinging upon the neural elements. For pediatric trauma patients with normal radiographs and CT of the neck but who are intubated, obtunded, or unable to fully understand or cooperate with an examination, MRI may also facilitate cervical spine clearance [31, 32]. MRI is also used frequently for patients with long-term complaints of pain or slow, progressive neurological changes from occult instability, skeletal dysplasias, and congenital abnormalities, among other diagnoses, to identify chronic cord changes, canal stenosis, and cord impingement.

Clearing the Cervical Spine in Children

Difficulties with Pediatric Cervical Spine Protocols

Cervical spine clearance protocols differ from one institution to the next. No clear consensus has been reached among institutions because limited evidence-based literature exists to guide development of these protocols, particularly for pediatric patients [4, 29, 32]. Additionally, staffing and other resources differ across institutions, making the details of each institution's method of operation unique. There are also inherent difficulties in clearing the cervical spine in children when compared to adults. In general, orthopedic surgeons have less experience evaluating children with cervical spine injuries, especially very young children, because of the relative rarity of these diagnoses in this population compared to adults. Similarly, interpretation of diagnostic imaging of the cervical spine is difficult for radiologists who infrequently evaluate these studies, especially given the developmental changes that occur in the cervical spine from birth until age 8 years.

Finally, because clearance protocols rely primarily on aspects of the clinical history and physical exam, obtaining reliable information from children, regardless of age or types of the injuries, is challenging. Fear, anxiety, and discomfort may make the child unable to effectively cooperate with the examination. This difficulty is compounded in children younger than 5 years of age, who may not developmentally be able to understand or communicate with the examiner.

Patient Stratification

Low-Risk Child

Trauma centers frequently use similar principles in the formation of protocols. Stratifying patients into categories based on risk of having a cervical spine injury is the basis of most protocols. National Emergency X-Radiography Utilization Study (NEXUS) [33] established five criteria in adults that correlate with a low risk for cervical spine injury. The goal of establishing these criteria was to allow the physician to clear the cervical spine by clinical examination, reducing or even ideally eliminating the need for diagnostic imaging:

NEXUS Criteria for Clinical Clearance of the Cervical Spine without Imaging

1. No midline cervical tenderness on palpation
2. Normal alertness
3. No intoxication
4. No neurologic abnormality
5. No distracting injury

Because the NEXUS criteria were established from data on adult patients, however, their applicability for children is questionable. Studies of the correlation in children are few and are hampered by low numbers of very young children in the study populations and a relatively low number of injuries diagnosed [4, 34]. Because of the limitations of the evidence, different criteria or additional considerations are used to identify those children at low risk for a cervical spine injury who do not need imaging. In one study, additional criteria were added to the NEXUS guidelines including any of the following: high-energy mechanism, transient neurological changes, physical signs of neck trauma, head or facial trauma, and inconsolability of the child [35].

Two important concerns, among others, are frequently raised regarding the application of NEXUS criteria. The developmental age of the child at which he or she can reliably cooperate with the cervical spine evaluation is variable and may be confounded by a learning disability or language delays. Finally, the definition of "distracting injury," and when it ceases to be distracting, is also not clear. It is our opinion, for example, that a child with a significant long-bone fracture may not be able to focus specifically on the cervical spine exam until a day or two after definitive treatment of that injury.

Based on our analysis and experience, it is our practice to use several criteria, coupled with clinical judgment about the developmental capabilities of the injured child and assessment of possible distractions, to determine who requires cervical immobilization and diagnostic imaging. Children who meet these criteria are able to have their cervical spine cleared based on the clinical examination without the need for cervical spine imaging.

At-Risk Child

As the above discussion reflects, many considerations must be taken into account when evaluating the conscious child who is developmentally appropriate and capable of cooperating with the exam before clinical cervical spine clearance can be done (Table 6.1). In this group of patients, clinical judgment is employed to weigh the risks of imaging versus the risk of missing a cervical spine fracture, erring always on the side of caution by imaging when in doubt. No debate is needed, however, when evaluating a potential injury in the patient who is younger than 5 years of age, has developmental disabilities, is under the influence of toxic substances, or has an altered level of consciousness for any other reason [32]. All who meet any of these criteria are considered unreliable and must be evaluated with diagnostic imaging.

Table 6.1 Factors in children and adolescents considered at risk for an acute cervical spine injury [3, 30, 34]

History of…	Examination findings	Unreliable patient[a]
Neck pain	Signs of trauma to the head, neck, or face	Unconscious
Mechanism of injury consistent with a cervical spine injury	Midline neck tenderness and limited ROM	Altered mental status secondary to intoxicants, head trauma, metabolic derangement
Transient neurological abnormalities	Abnormal neurological findings referable to the spinal cord or sensorimotor nerve roots	Unable to be reliably be examined secondary to young age, developmental disability, or a distracting injury[b]
Inconsolability after an injury (infants and young children)		

[a]A reliable patient is one who meets none of the three criteria for an unreliable patient

[b]A distracting injury is one that requires routine narcotic pain management for adequate comfort and is defined based on the judgment of the examiner

A trauma patient who meets any of these three criteria for an unreliable patient is considered at risk for a cervical spine injury. He or she should be immobilized in a hard cervical collar and undergo diagnostic imaging

Imaging and Care Algorithm

Imaging

While maintaining immobilization, all patients deemed at risk are imaged. As the above discussion shows, the ideal algorithm has not been established and is based on several factors including age, developmental status, and level of consciousness, the presence of neurologic injury, and the interpretation of prior studies. In the acute setting for patients deemed at risk who are awake and reliably able to cooperate with an examination, the standard three radiographic views are ordered primarily. In some centers, the open-mouth view is not done for children younger than 5–10 years of age because of the difficulty in obtaining an adequate odontoid view [32, 36]. CT of the cervical spine is the secondary study if the radiographs are inadequate or require further elucidation of a finding. Most other patients in the at-risk category, especially those who are unreliable or who are being evaluated with CT for concomitant injuries, however, are evaluated with CT scan to rule out a cervical spine injury.

Care Plan

Positive Findings

Immobilization in a hard cervical collar is maintained, and the Spine Service is consulted for all patients with positive findings on imaging. MRI may be acutely performed as a secondary study in the acute management phase for those with neurological deficits.

Negative Findings in a Reliable Patient

For those at-risk patients whose imaging is negative, initial treatment is determined primarily based on the reliability of the patients. The collar may be removed for reliable patients, except for those whose comfort is improved by immobilization. If the collar is prescribed, follow-up with a spine surgeon is indicated within one week or so of injury. Secondary imaging at follow-up may include flexion-extension lateral views (Fig. 6.6a, b) and MRI of the cervical spine.

Negative Findings in an Unreliable Patient

For unreliable at-risk patients with negative radiographs and CT scan, the collar is maintained. For

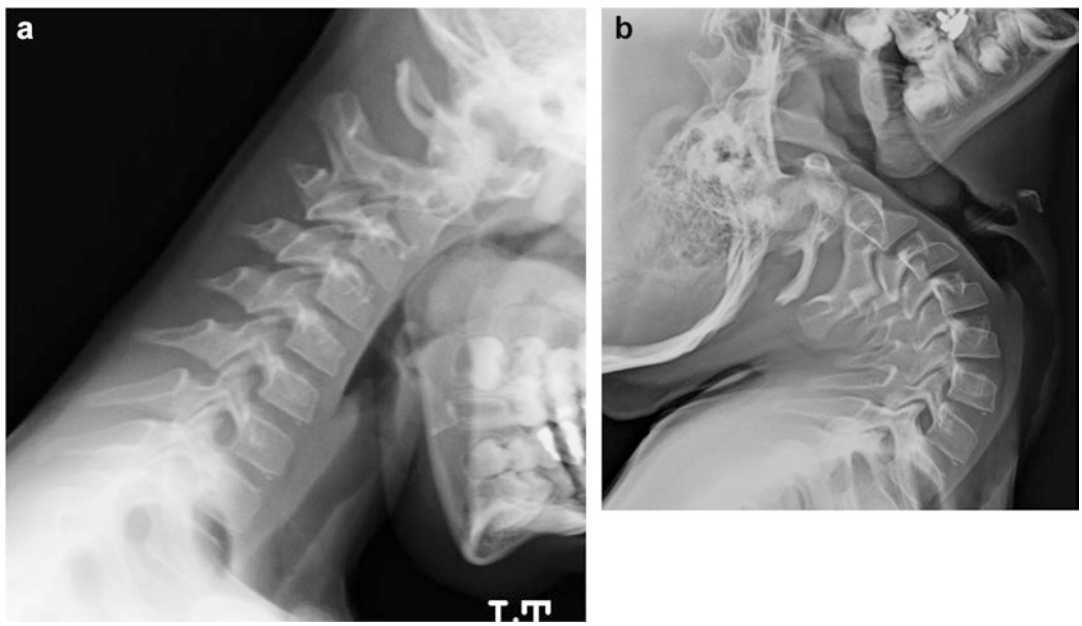

Fig. 6.6. An 8-year-old boy with neck pain 1 year after a fall on his head. Flexion lateral (**a**) and extension lateral (**b**) views of the cervical spine show instability at C1-C2 with an atlanto-dens interval measuring 7 mm

those unreliable patients who have negative radiographs and CT, MRI may be used as a secondary study to rule out soft tissue and ligament disruptions, osteocartilaginous injuries, and SCIWORA. MRI has been shown to facilitate clearance of the cervical spine in pediatric trauma patients, and its removal in an expeditious fashion is important to prevent skin ulceration and maceration, complications that may develop as early as 5–7 days after application. It is our opinion that flexion-extension views and fluoroscopy of the cervical spine have limited indications in this setting. If the unconscious or intubated can be reliably reassessed within 3–5 days, the collar may be removed at that time if it not necessary for comfort, with follow-up as described for the reliable patient.

Part II: Pediatric Upper Cervical Spine Injuries

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Cervical Vascular Injury

The incidence of cervical vascular injury (CVI) after blunt cervical trauma in children and adolescents is low. However, potential harm from missed injury is substantial; thus screening for CVI is important. Accepted screening methods include noninvasive angiography, such as computed tomographic angiography (CTA) and magnetic resonance angiography (MRA). Tolhurst et al. [37] examined 61 patients aged 4–18 years with cervical spine injury. Seven patients had a cervical vascular injury, for an overall incidence of 11.5%. High-risk criteria (fracture extension to transverse foramina, fracture/dislocations, severe subluxations, or C1-C3 injury) were associated with increased rates of CVI. Neurologic injury was found in six of seven patients with CVI; two of seven patients underwent anticoagulation due to documented CVI, and there were no delayed-onset ischemic neurological events.

Occipital Condyle Fractures

Occipital condyle fractures are rare in children [38–40]. Most of occipital condyle fractures are associated with head injuries with loss of consciousness and occur secondary to motor vehicle accidents or sporting injuries [39]. Presenting symptoms range from depressed level of consciousness to pain at the cranial/cervical junction, cranial nerve deficits, and paresis of the limbs. Plain radiographs may not clearly delineate the fracture, although they may reveal prevertebral soft tissue swelling from C1-C4. Computed tomography (CT) scan confirms the diagnosis, and fine-cut reconstructed images should be obtained in any pediatric patient presenting with basilar skull fracture, head injury, or neck pain.

Occipital condyle fractures were classified by Anderson et al. [41] into three categories. In Type I fractures, the mechanism of injury is an axial force which results in an impacted and comminuted fracture, without displacement of the fragments into the foramen magnum. Type II fractures occur via a direct blow to the cranium and are skull base fractures which extend into the condyle. In Type III fractures, the occipital condyle is avulsed by a combined lateral inclination and rotation force, which results in a free fragment which can be displaced in the foramen magnum. Type I and II fractures are generally considered stable, whereas Type III fractures are potentially unstable injuries.

Type I and II fractures are typically treated with the use of a cervical collar for 6–8 weeks. Type II fractures, where the occipital condyle is entirely separated from the occiput, or Type III fractures involving avulsion of the alar ligament, require halo immobilization or instrumented occipitocervical arthrodesis [41, 42].

Occipital Cervical Dislocation

Injuries to the atlanto-occipital (A-O) junction are unstable injuries which occur most commonly following high-energy trauma. This injury has been seen in up to 25% of fatal pediatric trauma cases and has a reported mortality rate of over 50% [7]. The rate of survival is improving

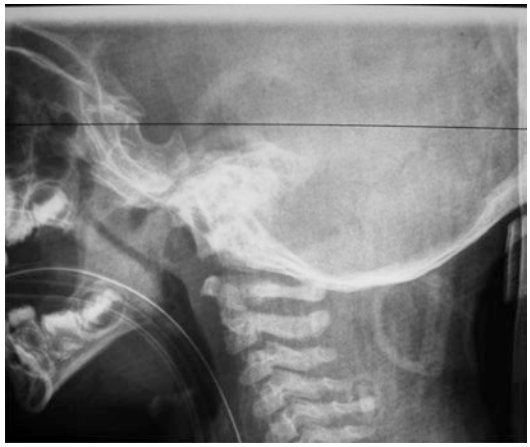

Fig. 6.7 Atlanto-occipital dislocation injury: 2-year-old male found unresponsive in the field following motor vehicle collision was intubated and transferred to the emergency department. Lateral C-spine radiographs reveal atlanto-occipital dislocation. Patient never regained consciousness and ultimately expired

due to advanced emergency management, cervical immobilization, and earlier diagnosis [43, 44]. The mechanism of injury involves a sudden deceleration, throwing the head forward on a relatively fixed trunk, such as in motor vehicle collisions or pedestrian versus auto injuries (Fig. 6.7). Younger children are at a higher risk for this injury pattern due to their larger head size, horizontal orientation of the A-O joint, underdeveloped occipital condyles, and overall ligamentous laxity. Clinical stability at the cranial/cervical junction is dependent on the alar ligaments, joint capsules, and tectorial membrane, as there is minimal inherent chondro-osseous stability of the A-O articulation. At this level, the tectorial membrane is a continuation of the posterior longitudinal ligament that connects C2 to the clivus and anterior rim of foramen magnum.

A number of radiographic parameters have been defined to diagnose A-O dissociation (Table 6.2) [45–50]. The usefulness of many of these measurements is limited, as they are based on changes in normal relationships between bony landmarks, which are not part of the occipital condyle-C1 articulation. Given the instability of this injury, minor changes in positioning can profoundly affect reliability of these measurements

Table 6.2 Normal radiographic parameters on lateral C-spine imaging in pediatric patients <8 years of age

Difference compared to adult spine	Normal value
Larger atlanto-dens interval	<5 mm
Pseudosubluxation: C2/C3	<4 mm
Larger retropharyngeal space	>6 mm at C2; >22 mm at C6
Neurocentral synchondrosis	Closure by 7 years of age

Adapted from Li and Hedequist [136]

on plain images. A high index for suspicion is necessary for any child with a closed head injury or significant facial trauma.

Pang et al. [51] proposed a radiographic measurement of the occipital condyle-C_1 (CC_1) interval as measured on high-resolution CT scans. The authors found a normal CC_1 interval of less than 2 mm, with strong left-right symmetry and no significant changes with age. In a follow-up study, the CC_1 interval was found to have a diagnostic sensitivity of 100% as compared to Wholey's dens-basion interval (50%), Power's ratio (37.5%), Harris' basion-axis interval (31%), and Sun's interspinous ratio (25%) in 16 patients with A-O dislocation [52].

Advanced imaging with CT or MRI should be obtained on patients with suspected occipital cervical dislocation and may reveal incongruent occipital cervical joints, tectorial membrane injury, perimedullary blood, and C1-C2 extra-axial hematoma [53]. Management of this injury includes instrumented surgical stabilization from occiput to C2 (or lower if needed to obtain adequate fixation) and immobilization in a pediatric halo. Fewer than 80 survivors have been reported in the literature with full neurological recovery in fewer than 10 cases [53].

Pediatric C1 Burst Fracture (Jefferson Fracture)

Fractures through the ring of C1 are uncommon in children and are most often the result of axial loading injuries [54]. The axial load is transmitted

from the head through the occipital condyles to the lateral masses of C1. Disruption of the ring of C1 most commonly involves failure in two places, which may occur through bone, synchondrosis, or both. Closure of the posterior synchondrosis occurs at approximately 3 years of age, whereas the neurocentral synchondrosis typically fuses by 7 years of age.

Fractures of C1 may involve a single arch (Type I) or include both arches (Type II) or involve the lateral masses (Type III) [54]. Plain radiographs may reveal the fracture; however CT is more accurate in defining the fracture pattern, determining the displacement of the lateral masses, and evaluating the integrity of the transverse atlantal ligament. Fractures through a synchondrosis can be difficult to appreciate radiographically and can contribute to a high diagnostic error rate for cervical spine injury in young children [55]. In adults, on AP radiographs and coronal CT images, displacement of the lateral mass of the atlas of >2 mm with respect to the superior articular facet of the axis indicates disruption of the ring of the atlas. In very young children, a "pseudo-spread" is commonly seen because of the more rapid growth of the atlas compared to the axis, and up to 6 mm of pseudospread can be seen in children younger than 4 years of age [54].

Spinal cord injury associated with Jefferson fractures are rare, given the large space available for the cord at this level, and disruption of the ring results in further expansion [56]. Stability of the transverse ligament is typically preserved unless wide displacement of the lateral masses (>7 mm) beyond the lateral margin of C2 is seen [57].

With an intact transverse ligament and minimal displacement of the lateral masses, treatment consists of immobilization in a hard collar, Minerva cast, or halo vest. If the lateral masses are displaced more than 7 mm, attempted fracture reduction with cervical traction should be considered, followed by halo immobilization. Healing may be confirmed by repeat CT scan and C1-C2 stability documented with flexion-extension radiographs. Residual instability on flexion-extension radiographs is an indication for posterior instrumented C1-C2 fusion with a cross connector used to compress the fracture (Fig. 6.8a–f) [58, 59].

Pediatric Odontoid and Synchondrosis Fractures

Odontoid fractures are among the most common injuries to the pediatric cervical spine. Most often these fractures occur through the synchondrosis at the base of the odontoid and are Salter Harris I fractures. The synchondrosis normally fuses at approximately 6 years of age, although delayed fusion is common (Fig. 6.9). This fracture is most commonly seen in patients with an open synchondrosis, usually occurring in patients younger than 6 [60, 61]. Odent et al. [62] reported on 15 patients over a 22-year period. In greater than 50% of the cases, the patient was involved in a motor vehicle collision with the child seated in a forward-facing car seat [62]. Other mechanisms of injury include falls, sports, and child abuse [63]. In over 90% of cases, the dens displaces anteriorly [61]. These factors suggest that the mechanism of action involves sudden deceleration with forced flexion of the head. Spinal cord injury occurs in approximately 25% of cases, with 20% of these being complete injuries [61].

Common symptoms include neck pain and resistance to extension, yet these may be missed during the initial assessment. Lateral plain film radiographs may reveal anterior displacement of the dens, unless spontaneous reduction has occurred. A CT scan with coronal and sagittal reconstructions may demonstrate the fracture if there is any displacement. MRI is also helpful in assessment of odontoid fractures to assess for edema and injury at the level of the synchondrosis and may reveal associated ligamentous injury.

Treatment consists of closed reduction in extension followed by immobilization in a halo or Minerva cast for 8–12 weeks. Healing potential is excellent, with up to 90% achieving union with external immobilization, likely due to an intact anterior periosteal sleeve [61]. Fulkerson et al. reported a complication rate of 43% and

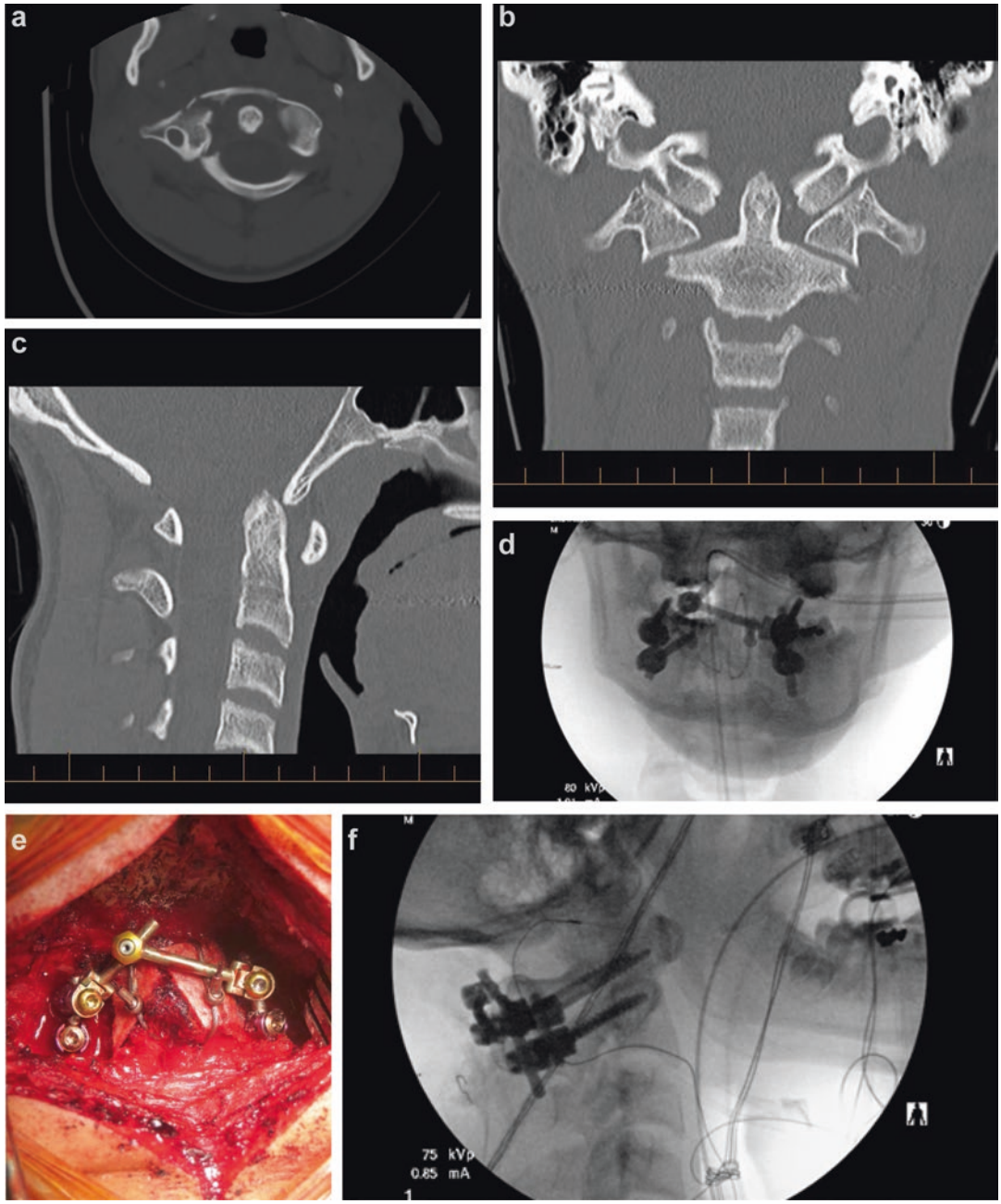

Fig. 6.8 (**a**–**f**) A 16-year-old male with neck pain following football tackle with C1 burst fracture seen on CT with axial (**a**), coronal (**b**), and sagittal (**c**) reconstructed images. The patient was treated with posterior C1-C2 instrumented fusion with C1 cross connector used to compress across the fracture. AP intraoperative fluoroscopy images (**d**), intraoperative photograph (**e**), and intraoperative lateral fluoroscopy (**f**) reveal improvement of C1-C2 alignment

nonunion rate of 11% with external immobilization for treatment of pediatric synchondrosis fractures. The authors recommended surgical fixation with posterior C1-C2 fusion for synchondrosis fractures with angulation >30° or significant displacement [64].

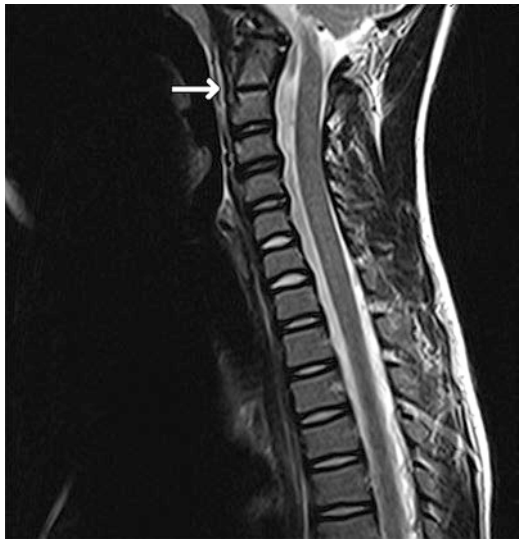

Fig. 6.9 Odontoid synchondrosis: 8-year-old female with neck pain following collision at soccer practice. MRI was obtained, revealing normal findings of persistent odontoid synchondrosis (arrow)

Os Odontoideum

Os odontoideum is characterized by a hypoplastic dens which is completely separate from the C2 body, with smooth cortical margins appearing as a separate ossicle. There is controversy about whether this finding represents a developmental abnormality or a nonunion after traumatic injury to the odontoid [65, 66]. Regardless of the etiology, the diagnosis and management are similar.

Patients may present with a wide range of clinical signs and symptoms, or the diagnosis may be made as an incidental finding on radiography. Three common presentations of os odontoideum may occur: (1) patients with occipitocervical pain only, (2) those with myelopathy (transient, static, or progressive), and (3) patients with intracranial symptoms or signs from vertebrobasilar ischemia [67].

Plain lateral images of the cervical spine clearly delineate the os odontoideum. CT scan may be helpful to differentiate an os from a fracture in the setting of trauma. Flexion-extension radiographs can reveal instability if present.

Two anatomic types of os odontoideum have been described: orthotopic and dystopic [67]. In the orthotopic type, the ossicle moves forward with the anterior arch of C1 on flexion-extension radiographs. In the dystopic type, the ossicle is functionally fused to the basion and does not move forward. Instability occurs anteriorly most commonly with neck flexion; however, posterior instability may be seen with extension [66]. Displacement of the ossicle by 8 mm or more with respect to the C2 body indicates instability.

Neurologic symptoms with evidence of cord compression or instability demonstrated on flexion-extension radiographs are indications for surgical treatment. Several case reports have documented catastrophic neurologic injuries after minor trauma when an os odontoideum is present [68–70]. Treatment should include instrumented posterior atlantoaxial arthrodesis. Several techniques have been described in the literature, including posterior wiring or transarticular screw fixation. Posterior wiring is associated with a higher nonunion rate, and overtighting of the wires may pull the os posteriorly into the spinal canal. Postoperative immobilization is often necessary with a pediatric halo. However, fusion rates are improved with transarticular screw fixation with reported union rates of 100%, without the need for postoperative immobilization in a halo device [71–74]. Other fixation options include C1 lateral mass screws with C2 pedicle screws (Fig. 6.10a–f).

Pediatric C2 Spondylolisthesis (Hangman Fracture)

Traumatic spondylolisthesis of the axis (Hangman fracture) occurs rarely in children and is usually associated with child abuse in children younger than 2 years of age [75–78]. The mechanism of injury is usually hyperextension with axial loading. Lateral plain radiographs demonstrate the fracture with associated anterolisthesis of C2 on C3 in greater than 90% of cases but must be distinguished from a persistent synchondrosis or congenital arch defect, which can mimic a fracture [79, 80]. CT scan can help delineate the osseous detail of the fracture, and MRI is useful for revealing hematoma or edema associated with

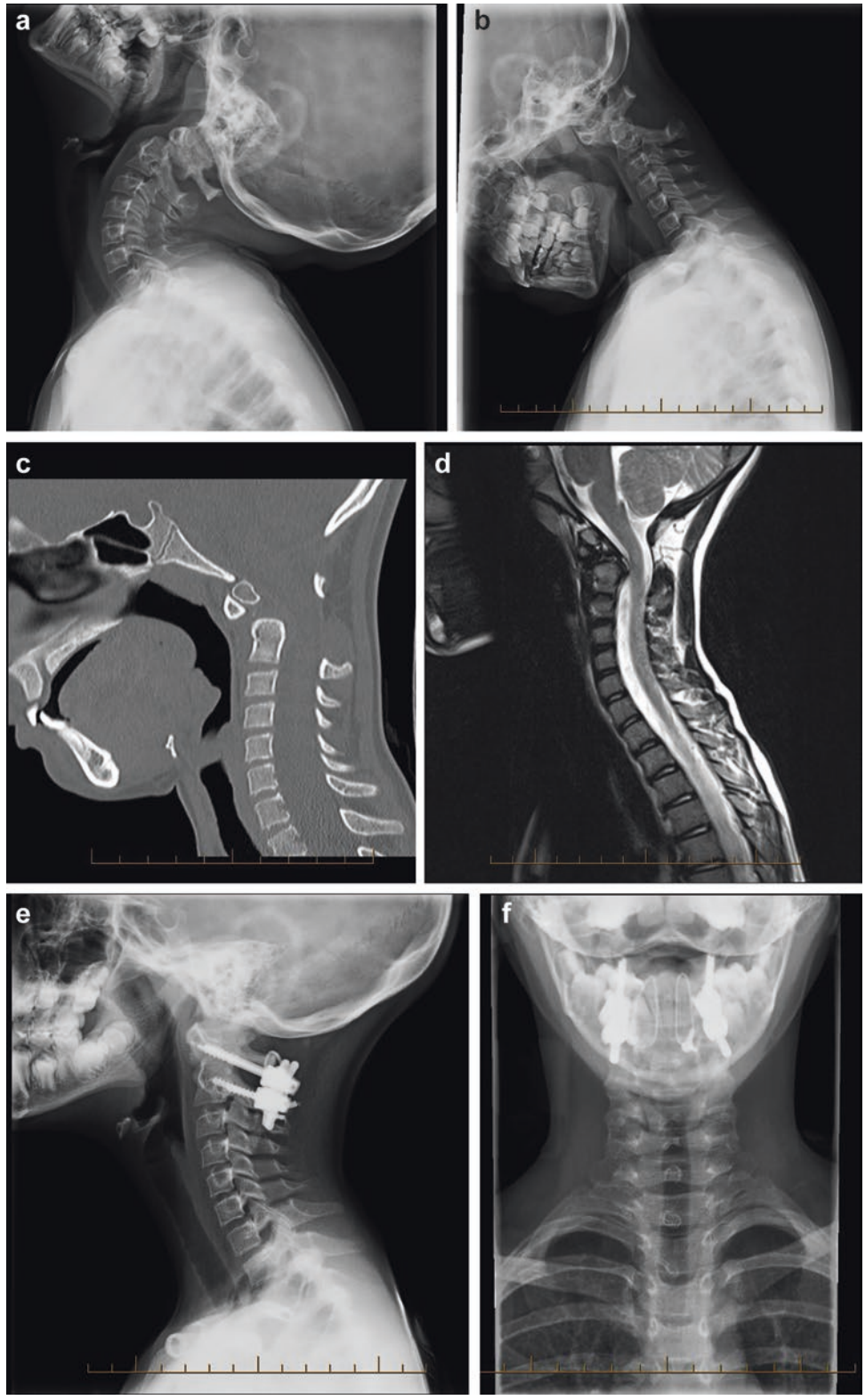

Fig. 6.10 (**a**–**f**) An 11-year-old male with neck pain. Flexion/extension radiographs revealed an os odontoideum with atlantoaxial instability (**a**, **b**). CT (**c**) and T2-weighted sagittal MRI (**d**) indicate severity of instability at rest and neural compression. Patient was treated with posterior instrumented fusion with C1 lateral mass screws and C2 pedicle screws (**e**, **f**)

an acute injury. Neurologic injury rarely occurs, as this fracture often increases the space available for the spinal cord.

Treatment of Hangman fractures includes closed reduction by extension of the neck, followed by 8–12 weeks of immobilization with a Minerva cast or halo fixator [77, 78, 81]. Instrumented arthrodesis via a posterior approach (C1-C3) or an anterior approach (C2-C3) may be indicated if persistent instability is documented or if a nonunion occurs.

Traumatic Atlantoaxial Rotatory Subluxation (AARS)

Atlantoaxial rotary subluxation (AARS), also called atlantoaxial rotary fixation, was originally described by Bell in 1830 and involves a fixed rotational deformity of C1 on C2 with significant loss of motion. AARS has been described after minor trauma, upper respiratory infection (Grisel syndrome), or following otolaryngologic procedures. The patient typically presents the "cock robin" position (head tilted to one side and rotated toward the contralateral side), has severely limited rotation, and has sometimes pain with movement. Neurologic symptoms are rare. Diagnosis may be delayed due to the subtle presentation of the patient's symptoms or unrecognized mechanism of injury (minor trauma or upper respiratory infection). Congenital torticollis can be distinguished from atlantoaxial rotary subluxation in that torticollis is passively correctable and is not typically associated with pain, and there is tightness in the sternocleidomastoid muscle on the side opposite to the deviation of the chin. In atlantoaxial rotary subluxation, the sternocleidomastoid muscle on the ipsilateral side is often in spasm, suggesting an attempt to stabilize or reduce the deformity. Patients are unable to rotate the head past the midline in the direction opposite to which the head is turned.

AARS is classified into four types, based on the direction and degree of rotation and translation as seen on axial CT images [82]. In Type I, which is most common variant, there is unilateral rotation of the C1 without displacement, which suggests an intact transverse ligament. Type II involves deficiency of the transverse ligament allowing anterior displacement of one lateral mass of 3–5 mm with the contralateral lateral mass serving as the pivot point. In Type III, there is deficiency of both transverse and secondary ligaments allowing anterior displacement of both lateral masses >5 mm. With a Type IV injury, a deficiency or injury of the dens allows posterior displacement of C1 on C2.

Plain radiographs can be difficult to interpret given the abnormal positioning of the head and cervical spine [83]. A lateral radiograph of the skull (with the cassette parallel to the head and the X-ray beam directed perpendicular to the head) will allow evaluation of the occupt-C1 relationship. Asymmetry of the lateral masses may be seen on an open-mouth odontoid view. Dynamic CT scan allows evaluation of the relationship of C1 on C2 and may demonstrate a fixed malalignment. Pang and Li utilized dynamic CT scans to identify and plot a template of the normal motion between C1 and C2 in 21 children [84]. The authors found this normal template could be compared to dynamic CT scan, with the head in neutral position and rotated both directions, to effectively diagnose AARS [85, 86].

Treatment strategies depend upon the duration of symptoms prior to initial presentation [83, 87]. Symptoms which are present less than 1 week can typically be treated with a soft collar, anti-inflammatory medications, and physical therapy [87]. Other authors have recommended all acute and subacute AARS patients be managed with halter traction and muscle relaxants and sedation immediately to prevent chronic AARS. Halter traction should be initiated in an inpatient setting with scheduled benzodiazepines until reduction is achieved. Following reduction, the child should be immobilized in a cervicothoracic orthosis for 3 months. Acute and subacute AARS while in a brace are considered "recurrent AARS" and are treated with traction in calipers followed by a placement of a halo device or instrumented posterior atlantoaxial arthrodesis (Fig. 6.11a–d). Irreducible AARS is also treated with posterior atlantoaxial arthrodesis [82, 83, 87, 88]. Preoperative traction may be useful in reducing the amount of subluxation prior to fusion or open reduction [87, 88].

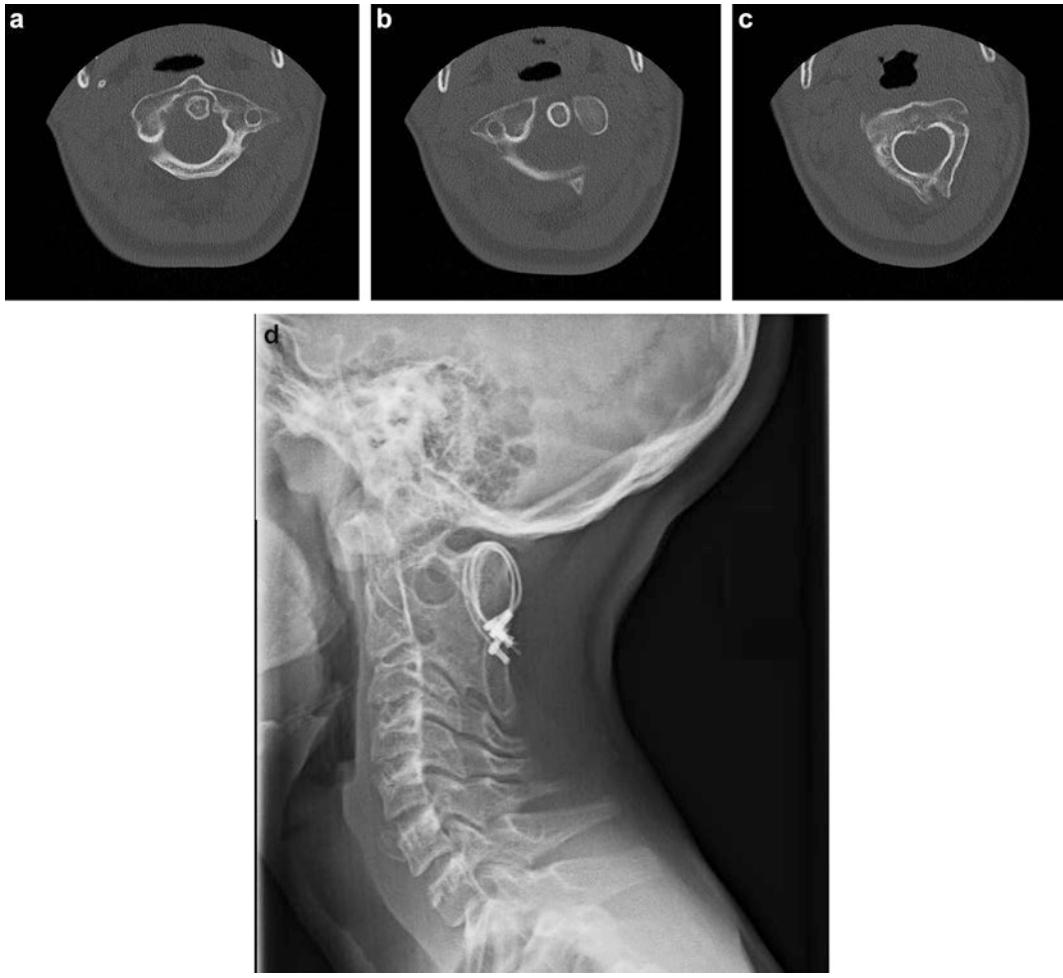

Fig. 6.11 (**a**–**d**) Atlantoaxial rotatory subluxation: 11-year-old male presented 3 months following minor neck trauma with a tilted head. CT examination revealed rotatory subluxation of C1 on C2 (**a**–**c**). Initial treatment with halo traction improved head alignment, although persistent rotatory subluxation was present. Posterior wired C1/C2 arthrodesis was performed with solid fusion present on lateral radiography (**d**)

Part III: Subaxial Cervical Spine Injuries in Children

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Introduction

After the age of 8 years, cervical spine injuries in children resemble patterns seen in the adult population. Thus, subaxial injuries become more prevalent. Subaxial cervical spine injuries represent 23–73% of pediatric cervical spine injuries [6, 89–102]. The distinction between what is considered subaxial is variable, however. This can range from C3 to C7, C4 to C7, or C5 to C7, depending on the series. Although it is clear from the literature that subaxial injuries occur more commonly in older children [96, 98, 99, 103, 104], this region

of the cervical spine should be evaluated thoroughly in younger patients, as injuries in this region are not that uncommon. For example, Brown et al., in their series of 103 patients, found that injuries at the C5-C7 levels were almost equally distributed between patients younger than 8 years of age (24%) and those older than 8 years of age (25%) [101]. Additionally, Osenbach et al. [94] and Bilston [103] found injuries at the C4-C7 levels in children younger than 8 years of age in 26% and 21% of their subjects, respectively.

Demographics

The most common mechanism of injury is motor vehicle collisions (MVC). In a review of 51 subaxial injuries in children, Dogan et al. reported that MVC was responsible for 50% of injuries [104]. However, in the older age group, more commonly afflicted with subaxial cervical spine injuries, sports and recreational activities become more common [6, 93, 95, 97, 98, 101]. McGrory et al. [96] found that 43% of cervical spine injuries were due to sports-related activities in the 11–15-year-old age group versus only 8% in those younger than 11 years.

The average age for subaxial cervical spine injuries ranges from 12.4 years to 16 years [6, 93, 98, 100, 104]. Males are injured more commonly than females. In Dogan's review of 51 pediatric subaxial cervical spine injuries, 75% of the cases involved males [104].

Classification

Unfortunately, a classification system specific to pediatric subaxial cervical spine injuries does not exist. Allen et al. developed a system that categorizes injuries based on mechanism of injury [105]. The system involves six phylogenies: (1) compressive flexion, (2) vertical compression, (3) distractive flexion, (4) compressive extension, (5) distractive extension, and (6) lateral flexion. Each phylogeny is designated an injury stage numerical value to describe injury severity, e.g., stage I, stage II, etc. This system was based on the review of 165 cases, 26 of which were patients under the age of 18 years. More recently, Vaccaro et al. [106] developed the Subaxial Injury Classification and Severity Scale (SLIC) (Fig. 6.12). The system takes into consideration three injury characteristics: (1) morphology, (2)

	Points
Morphology	
No abnormality	0
Compression	1
Burst	+1 – 2
Distraction (*e.g.,* facet perch, hyperextension)	3
Rotation/translation (*e.g.,* facet dislocation, unstable teardrop or advanced staged flexion compression injury)	4
Disco-ligamentous complex (DLC)	
Intact	0
Indeterminate (*e.g.,* isolated interspinous widening, MRI signal change only)	1
Disrupted (*e.g.,* widening of disc space, facet perch or dislocation)	2
Neurological status	
Intact	0
Root injury	1
Complete cord injury	2
Incomplete cord injury	3
Continuous cord compression in setting of neuro deficit (Neuro Modifier)	+1

Fig. 6.12 SLIC system (From Vaccaro et al. [106]. With permission of Lippincott Williams & Wilkins)

integrity of the discoligamentous complex, and (3) neurologic status. Each of these three categories contains subgroups that are given a numerical value depending on severity. The sum of each numerical value is then used to guide the surgeon's decision to treat the patient nonsurgically or surgically. A score of 1–3 favors nonsurgical treatment, whereas a score of 4 or greater favors surgical stabilization. Given that subaxial injuries occur more commonly in older children, with anatomy similar to that of an adult, this system seems to provide a reasonable option for surgeons caring for these particular injuries. Obviously, this is a topic that requires additional research. What is not known is whether children and adolescents are capable of healing injuries that typically fail nonsurgical care in adults.

Injury Patterns

Dogan et al. [104] described four injury patterns seen with pediatric subaxial injuries: (1) fractures (63%), (2) fracture with dislocation (19%), (3) dislocation (12%), and (4) purely ligamentous injury (6%). They also found that injury to the C6-C7 level was the most common (33%) followed by C5-C6 (23%). Both Nitecki and Moir [96] and Brown et al. [101] found C7 to be the most commonly injured vertebrae. In fact, Brown et al. found that of their 26 children with subaxial injuries, 85% had a fracture of C7. This underscores the importance of visualizing C7 on imaging studies. Likewise, the possibility of a purely ligamentous injury cannot be overemphasized as these injuries might not be immediately recognizable in a child. Pennecot et al. [91] in a pre-MRI era study reported eight cases of delayed diagnosis. The authors reported no radiologic evidence of injury at the time of initial assessment. The common feature of these patients was a lack of physiologic cervical lordosis and a stiff neck. The injuries were not discovered until 2 weeks to 4 months post-injury after the muscle spasms had resolved and dynamic radiography was performed.

As with purely ligamentous injuries, the possibility of injury to more than one level needs to be considered. Patel et al. [100] reported a 7% incidence of combined upper and lower level injuries in their review of 1098 pediatric cervical spine injuries. Eighty-eight percent involved children older than 8 years of age. Dogan et al. [104] found that 18% of their 51 pediatric patients with subaxial injuries had injury to more than one level. All nine of these patients were 8 years of age or older.

As with any injury to the spinal column, neurologic injury must be ruled out. In Birney and Hanley's [93] review of 84 consecutive pediatric and adolescent patients with cervical spine injuries, 19 (44%) had injuries to the lower cervical spine, defined as C3-C7. Of the 19 patients, 10 had neurologic injury. Three of the ten were complete injuries, one of ten was permanent incomplete, and six of ten were transient deficits. Patel et al. [100] reported a 5% incidence of spinal cord injury in patients with C4-C7 level injuries. Dogan et al. [104] reported a 16% incidence of incomplete neurologic injury and 14% for complete spinal cord injury.

Treatment

The available evidence to support the various treatment methods for pediatric subaxial cervical spine injuries is minimal. Unfortunately, most of the published literature is inconsistent in regard to injury classification/description and outcome measurements. As most studies combine patients with craniocervical and subaxial injuries, it is often difficult to evaluate the outcomes specific to subaxial injuries. As a result, some of the recommendations for treatment must be extrapolated from the adult literature. The following will be the author's recommendations for common pediatric subaxial cervical spine injuries, based on personal experience and the available evidence.

Posterior Element Fractures

A majority of isolated posterior element injuries can be treated nonoperatively. In the series reported by Dogan et al. [104], 29% of their pediatric subaxial injuries were isolated posterior element injuries. These were treated nonsurgically.

The authors did not elaborate, but most patients were treated with either a cervicothoracic orthosis or cervical orthosis. At an average follow-up of 9 months, none of their nonsurgically treated patients required surgical intervention for instability. McGrory et al. [95] included ten subaxial posterior element fractures in their series of pediatric/adolescent cervical spine injuries. An interesting finding of this study was that the injuries were primarily considered secondary injuries, associated with a more severe primary injury. In fact, all five spinous process fractures reported were associated with a nonadjacent, more severe, primary injury.

Of spinous process fractures, a well-known injury is the so-called clay-shoveler's fracture (Fig. 6.13), which often involves the spinous process of C7 [107]. Typically, this injury entails an avulsion of the spinous process from forceful trapezius and rhomboid contraction. Since the spinous process apophysis does not fully ossify until the third decade [108, 109], an obvious fracture may not be visible on radiographs. Yamaguchi et al. [110] reported two cases of clay-shoveler's equivalent injury in two male athletes, involving a soft tissue avulsion of the spinous process. One of the injuries was at C7. The injury was identified on MRI. The patient was treated with activity restriction for 8 weeks.

Before making the diagnosis of clay-shoveler's fracture, it is important to critically review the imaging. Matar et al. [111] reported six cases of cervical spinous process fractures that displayed the sign of "spinolaminar breach" (Fig. 6.14). The average patient age was 31 years, with a range of from 8 to 59 years. Five of the six patients sustained their injuries from either an MVC or motorcycle accident. The injuries occurred at C6 (n = 5) and C5 (n = 2). In their report, four of the patients developed anterior subluxation, and two developed a neurological deficit.

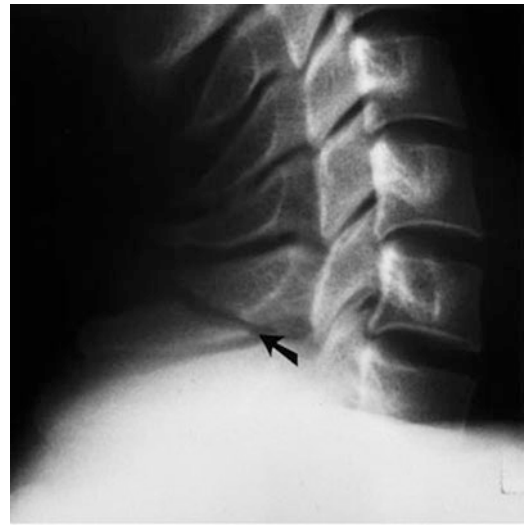

Fig. 6.14 Spinolaminar breach of C5 spinous process. Skeletal radiology by International Skeletal Society (From Richardson et al. [111]. Reproduced with permission of Springer-Verlag)

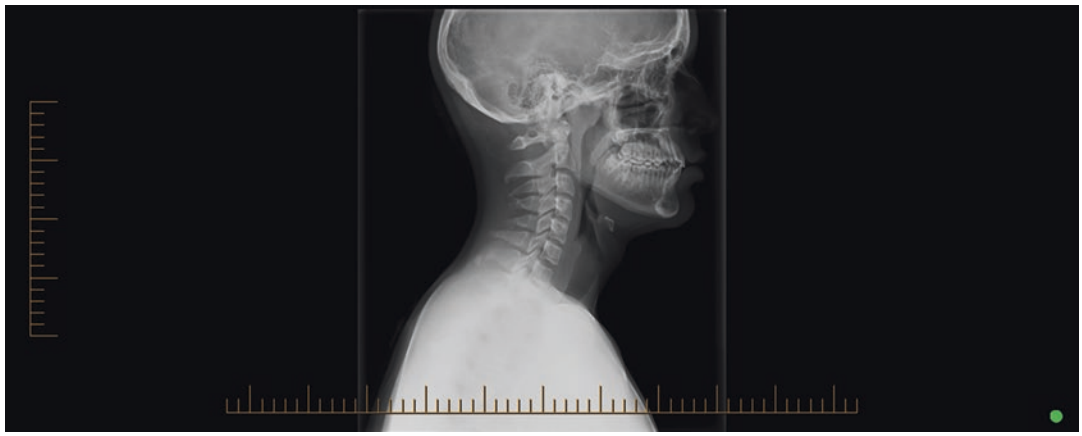

Fig. 6.13 Clay-shoveler's fracture of C7

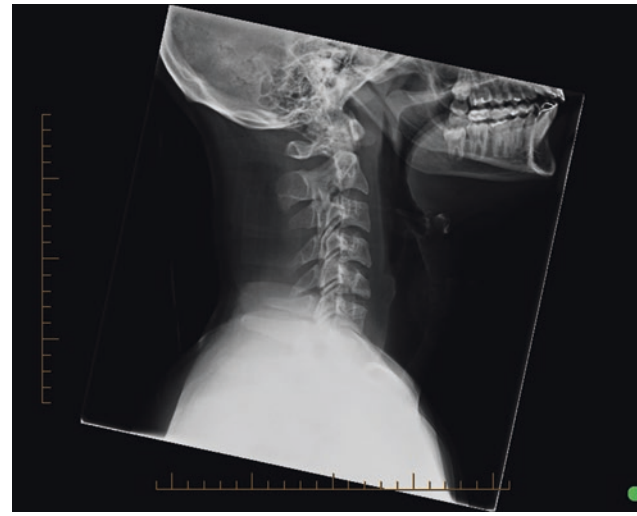

Fig. 6.15 C5 compression fracture

Compression Fractures

By definition, a compression fracture (Fig. 6.15) usually entails a fracture involving only the anterior column of the spine, with blunting of the anterosuperior vertebral body and no evidence of ligamentous injury. Of the six phylogenies described by Allen et al. [105], this would represent a compressive-flexion stage I injury. Unfortunately, when reviewing the pediatric spine literature, what is defined as a compression injury is not clear. Therefore, treatment recommendations have to be taken with caution. Dogan et al. [104] reported 10 compression fractures in their series of 51 pediatric subaxial cervical spine injuries. In two of the patients, they performed corpectomies and anterior fusions. It is not possible to ascertain what exactly is defined as a compression fracture. One of the patients had a complete spinal cord injury, which would lead one to infer that the patient had a severe injury. Although the exact number of subaxial cervical spine compression fractures in their study of 122 pediatric neck injuries was not disclosed, Hill et al. [112] recommended external immobilization for stable compression fractures. They did not report the results of treatment rendered.

Based on the SLIC classification system [106], an isolated compression fracture without a disrupted discoligamentous complex, and in a neurologically intact patient, would produce a score of 1. In this setting, the patient would qualify for nonsurgical treatment. Usually, a rigid cervical orthosis will suffice. Obviously, it is important to take the entire clinical and radiological picture into consideration prior to determining definitive treatment.

Burst Fractures

Unlike compression fractures, burst fractures (Fig. 6.16) typically involve at least two of the three spinal columns, often with retropulsion of the posterior vertebral body into the spinal canal. This injury is categorized by the vertical compression phylogeny in the Allen-Ferguson classification system [105]. None of their patients under the age of 18 years was in this phylogeny. Since these injuries are felt to be secondary to axial load, in theory, the posterior ligaments should be intact. However, Allen et al. [105] noted that in stage III injuries, the posterior ligaments may be disrupted.

Treatment of these injuries is controversial, for both children and adults, as no high-level evidence exists to guide surgeons. An age-old problem is defining exactly what is meant by instability

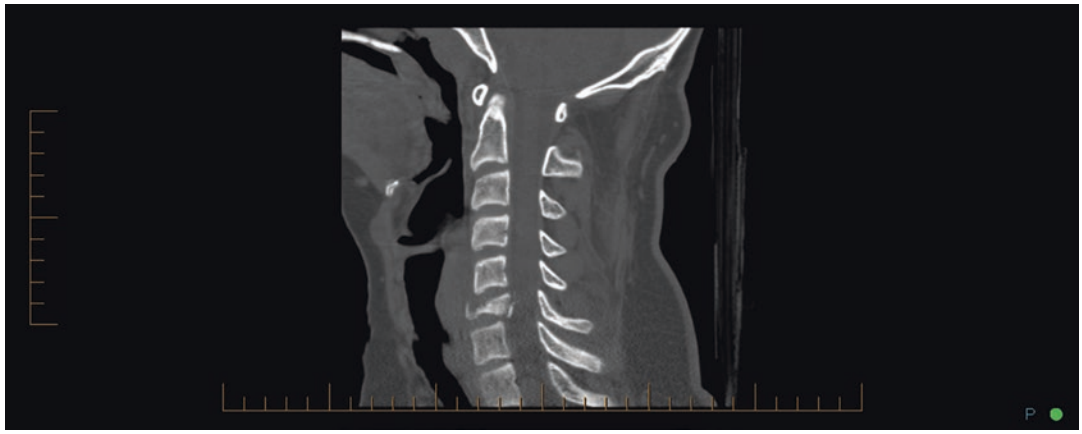

Fig. 6.16 C6 burst fracture

[113]. As with all pediatric subaxial injuries, the available literature pertaining to this particular injury is sparse. Dogan et al. [104] reported only one burst fracture in their review of subaxial pediatric injuries. The patient was a 16-year-old male with a complete spinal cord injury that underwent an anterior fusion without corpectomy. Finch and Barnes [97] reported five patients with subaxial burst fractures. They reported treating three of the five with a posterior fusion with a wire construct for instability and kyphosis. Both patients had more than a one level of injury and went on to fuse. The details of how the other two burst fractures were treated are not given, but the authors state that all nonoperatively treated patients went on to achieve radiographic stability.

Based on the SLIC classification system [106], a burst fracture without a disrupted discoligamentous complex, and in a neurologically intact patient, would produce a score of 2, thus suggesting nonsurgical care as a viable option. However, if the patient has any disruption of the discoligamentous complex or neurologic injury, the score could easily reach 5, which would favor surgical treatment (Fig. 6.17a–c). Although data comparing one form of surgical treatment to another is lacking, it seems well accepted that a burst fracture with concomitant neurologic injury would be a reasonable indication for an anterior cervical corpectomy, strut grafting, and instrumented fusion [114–116]. If significant posterior ligamentous injury is present, such in the case of a teardrop fracture, a concomitant posterior instrumented fusion also seems reasonable [114]. Fisher et al. [116] reviewed their results of treating unstable flexion teardrop fractures with either anterior corpectomy and plating ($n = 21$) or halo-thoracic vest immobilization ($n = 24$). Anterior corpectomy and plating produced better results in regard to residual kyphosis and failure of treatment. Five patients in the halo-thoracic vest group were considered to have treatment failure; four of whom went on to surgical treatment. Whether or not this study is relevant to the pediatric/adolescent population is debatable, as it involved adult patients only.

Koivikko et al. [117] reported on 69 patients with burst fractures treated either nonsurgically ($n = 34$) or by means of anterior decompression and instrumented fusion ($n = 35$). The age range for the nonoperative group was 15–64 years (average = 30.3) versus 17–83 years (average = 32.9) for the surgically treated group. The authors found better results in the operatively treated cohort in regard to neurological recovery, spinal canal narrowing, and kyphotic deformity. Caution must be taken before extrapolating the results of this study to the pediatric/adolescent population, as no power analysis was performed, the patients are primarily adults, and some element of bias may have been introduced due to the retrospective design of the study.

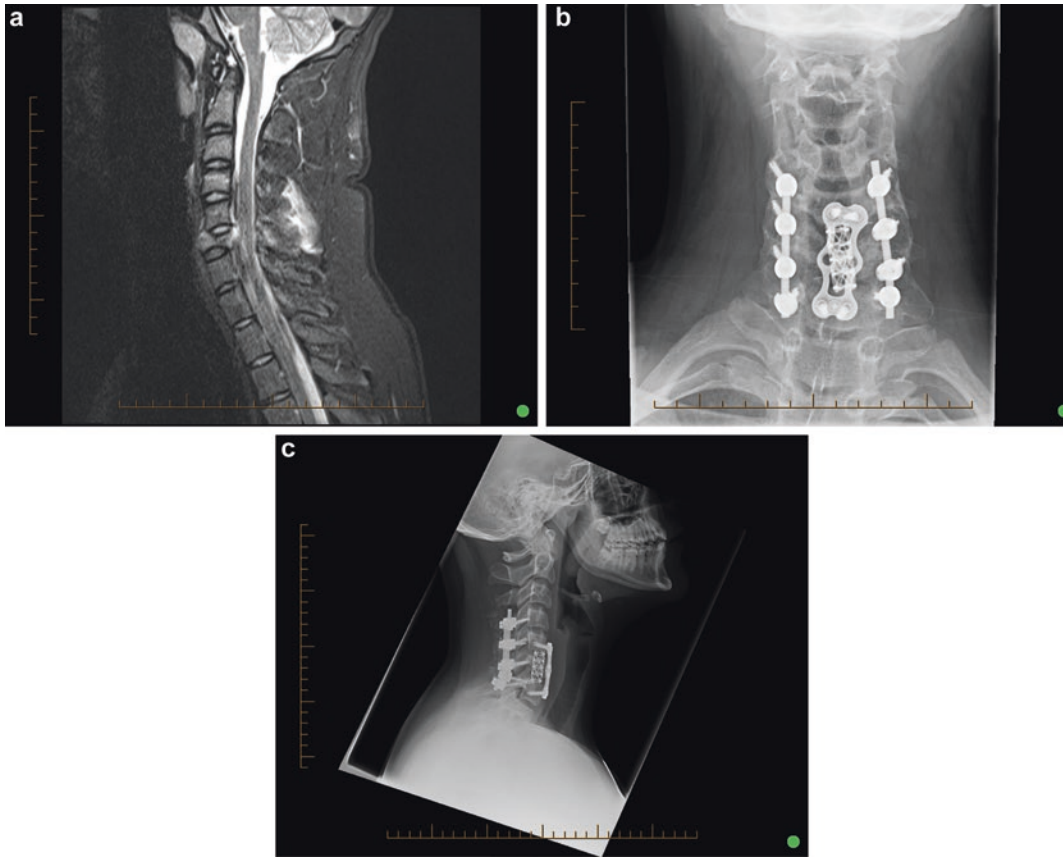

Fig. 6.17 (**a**–**c**) (**a**) Sagittal MRI image of the same patient in Fig. 6.16. Note the posterior ligamentous injury cephalad to burst fracture; (**b**, **c**) postoperative X-rays following C4-C7 posterior instrumented fusion and C6 corpectomy with plate fixation

Facet Dislocations

Unilateral facet dislocations are categorized under the flexion-distraction phylogeny of the Allen and Ferguson classification system [105]. Unilateral dislocations are considered stage II injuries, whereas bilateral dislocations are designated stage III injuries. Bilateral dislocations are usually associated with at least 50% displacement. To no surprise, treatment recommendations based on pediatric data are extremely limited. Dogan et al. [104] reported five unilateral facet dislocations in their review of pediatric subaxial cervical spine injuries. Three of them were treated with an anterior fusion. No complications were reported. Details of the nonoperatively treated patients were not given, but the manuscript would suggest that they underwent closed reduction with stability obtained at follow-up. As with unilateral dislocations, the literature pertaining to bilateral dislocations is also limited. Finch and Barnes [97] and Dogan et al. [104] each reported one patient with a subaxial bilateral facet dislocation. Both patients were treated surgically.

For unilateral or bilateral facet dislocations, a closed reduction should be attempted. The weight that should be utilized is controversial. I generally prefer to start with 10 pounds for the head and adding 5 pounds per level of injury. It is helpful to maintain the bed in reverse Trendelenburg. The total amount of weight that can be safely utilized is unknown. A previous study by Cotler et al. [118] reported using up to 140 pounds.

The youngest patient in this study was 16 years old. What is not controversial, however, is the importance of repeat radiographic assessment and neurological evaluation after the weight is increased. The issue to whether an MRI should be obtained prior to reduction is a matter of debate [119]. Some feel that a closed reduction can be performed in an awake, alert patient who is capable of cooperating with a neurological examination [120]. Most would agree that a patient with a complete spinal cord injury should probably undergo a reduction prior to MRI [120]. If the patient is unable to cooperate with a neurological examination, an MRI should probably be performed prior to reduction. If a disc herniation is noted on MRI, an open decompression and instrumented fusion are generally recommended.

The definitive method of treatment is controversial for unilateral facet dislocations. Unfortunately, surgeons treating these injuries in children will largely have to rely on the adult literature for guidance. Rorabeck et al. [121] felt that successfully reduced unilateral dislocations could be successfully treated in a halo-thoracic orthosis. Shapiro et al. [122] reported two patients in their series of 51 unilateral facets that they unsuccessfully attempted to treat with external immobilization following closed reduction. Both patients developed resubluxation. Dvorak et al. [123] found in their review of the literature a higher incidence of treatment failure, pain, and neurological deterioration in the nonoperatively treated unilateral facet dislocations.

Based on the SLIC classification system [106], most dislocations, unilateral or bilateral, will produce a score of at least 5, thus favoring surgical treatment (Fig. 6.18a–e).

Spinal Cord Injury without Radiographic Abnormality (SCIWORA)

Pang and Wilberger [124] were the first to define SCIWORA as myelopathy secondary to trauma without evidence of fracture or ligamentous instability on plain radiographs or tomography. The paper was published prior to the use of MRI. The injury is thought to be possible due to the relative flexibility of the pediatric spinal column compared to the less flexible spinal cord. The spinal column is capable of stretching up to 2 in, whereas the spinal cord can rupture with less than 1 cm of stretch. Pang and Wilberger [124] found that children younger than 8 years of age sustained more serious neurologic injury and injuries to the upper cervical cord. Bosch et al. [125] also found that children under the age of 8 sustained more serious injuries. However, contrary to some reports [94, 98], but similar to that of Brown et al. [101], they found SCIWORA more commonly in children older than 8 year of age, with an average age of 10.7 years. The reported incidence of SCIWORA, in literature pertaining to pediatric cervical spine injuries, is 18–38% [6, 93, 94, 98, 100, 101]. Males sustain SCIWORA more commonly than females [101, 126]. Brown et al. [101] reported that 72% of their cohort with SCIWORA was males. Although motor vehicle accidents are a common cause of SCIWORA, recreational/sports-related injuries are also quite common [93, 101, 125]. Brown et al. [101] reported that 75% of their sports-related injuries involved SCIWORA. Fairly trivial episodes of trauma have also been reported [126].

The prognosis for recovery seems directly related to the neurologic status at presentation [93, 125–127]. Those with complete neurologic injury at presentation appear to have a poor prognosis for neurological recovery. Findings on MRI also seem to be predictive of neurological recovery. Of 60 MRIs reviewed by Bosch et al., 51 were negative [125]. All of these patients made a full recovery even if they presented with a severe neurological deficit. Similarly, Mahajan et al. [127] reported that 67% of children with an abnormal MRI had persistent neurological deficits versus only 6% with a normal MRI. Overall, they reported that 78% of their cohort had normal MRIs.

Treatment of SCIWORA is controversial because the exact meaning of the term is somewhat less clear in our era of frequent MRI use. It seems logical, however, to follow standard support measures in those presenting with a spinal

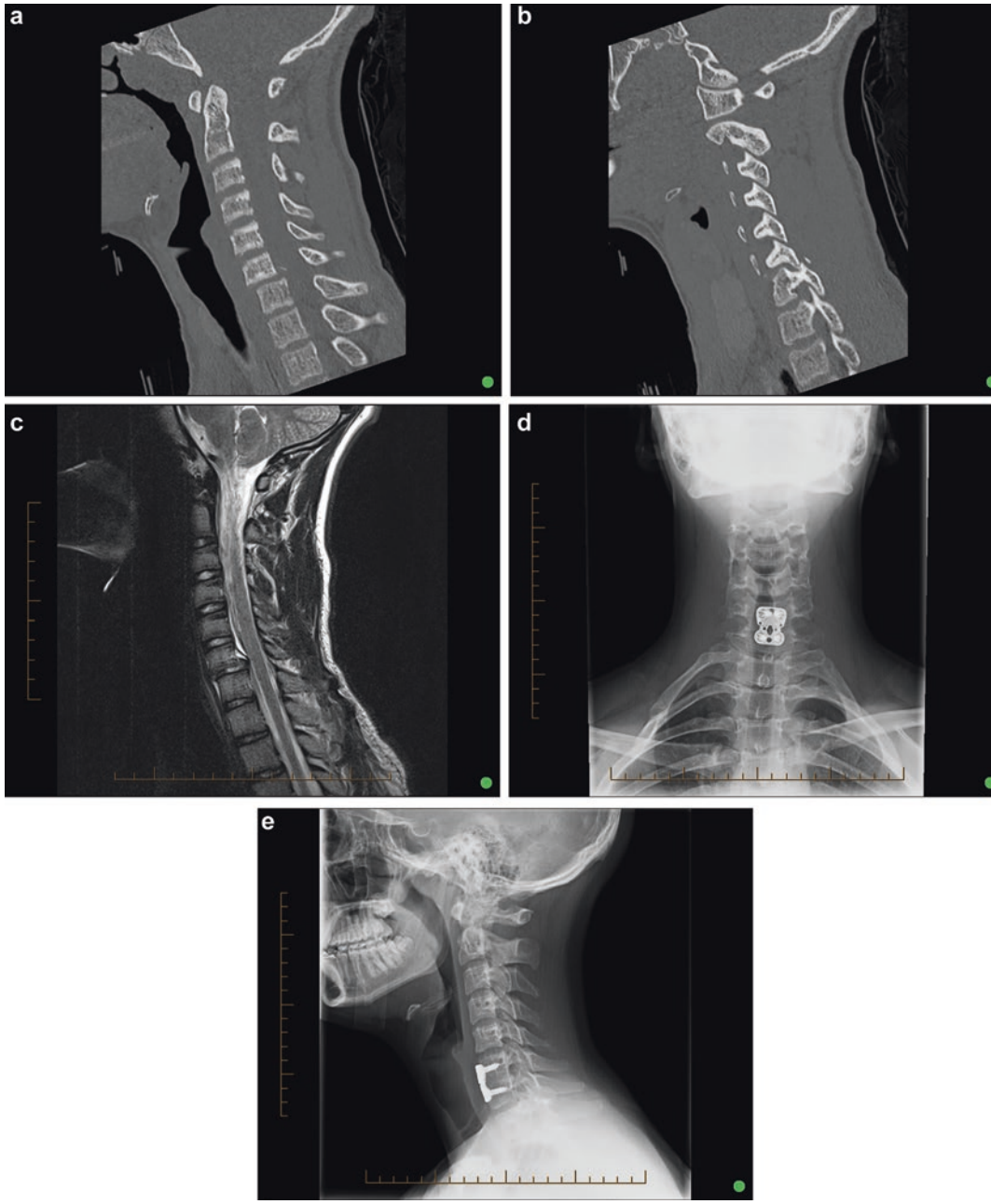

Fig. 6.18 (**a**–**e**) (**a**, **b**) Sagittal CT images depicting unilateral right-sided facet dislocation; (**c**) sagittal MRI revealing discoligamentous disruption; (**d**, **e**) postoperative X-rays 2 years after undergoing a C6-C7 anterior cervical discectomy and fusion with iliac crest autograft

cord injury pattern. What is debatable is the need for external immobilization in patients with normal MRI. Although advocated by some [1], this practice has been challenged. Bosch et al. [125] found the incidence of recurrent SCIWORA to be 4% when strict criteria were used to establish the diagnosis. In all cases, the injury occurred in an older patient, was the result of a low-energy mechanism, and was associated with a normal MRI and mild-transient neurological deficits.

All patients made a complete recovery. They found that bracing did not prevent reinjury. In fact, the group that was braced experienced a 15% recurrent injury rate versus 7% in those treated without a brace.

Spinal Cord Injury

Fortunately, spinal cord injury (SCI) in children is rare. However, it can have devastating long-term consequences. Hwang et al. [128] in their longitudinal study of adult patients that sustained pediatric-onset spinal cord injury found an increasing risk of developing hypertension/cardiac disease, upper extremity joint pain, and activity-limiting upper extremity pain with time. More acutely, children with SCI are prone to many medical complications, including those related to the pulmonary system, deep venous thrombosis, autonomic dysreflexia, hypercalcemia, heterotopic ossification, spasticity, neurogenic bladder/bowel, decubitus ulcers, scoliosis, and pain [129]. Clinicians caring for these children must obviously be diligent in taking measures to prevent some of these complications and be knowledgeable regarding treatment strategies when they occur.

An area of controversy that continues to spark much discussion is the role of corticosteroid administration to patients with SCI. This topic is controversial in adult circles and even more so in the pediatric medical community. This is the result of the paucity of data to support the use of corticosteroids in children with SCI. The recommendation to administer methylprednisolone to patients with SCI is based largely on the results of the second and third National Acute Spinal Cord Injury Studies [130–133]. The results of these studies are difficult to extrapolate to the pediatric population, as the number of pediatric patients was very small. In fact, no patients under the age of 13 were included in the studies. Pettiford et al. [134] recently conducted a literature review with an emphasis on pediatric SCI. They concluded that methylprednisolone cannot be recommended for children with SCI.

The timing of surgical decompression is also controversial. More recently, the trend has favored early decompression. The results of the Surgical Timing in Acute Spinal Cord Injury Study have supported this trend [135]. The authors conducted a multicenter, international, prospective cohort study to evaluate the outcomes of early (<24 h after injury) versus late (>24 h after injury) decompressive surgery after cervical SCI. The authors found that 19.8% of the patients that underwent early decompression improved two or more ASIA grades versus only 8.8% in the late group. It is not known if the results of this study can be applied to the pediatric population. This is clearly a topic that is deserving of a well-designed study specific to the pediatric population.

Summary

The incidence of traumatic cervical conditions in children is rare, but catastrophic results can occur. Injury pattern is different in the young child compared with older children and adults. Radiographic assessment is challenging and will often require advanced imaging. Spinal cord injury approaches 10% in some series. The unique problem of SCIWORA in children is commonest under the age of 5. As children grow injury levels descend from the skull base, C1 and C2 to the subaxial spine. The need for operative fixation depends on fracture stability and presence or absence of neurological compromise.

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Infections and Inflammatory Conditions of the Pediatric Cervical Spine

7

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Introduction

When a child presents with complaints of pain, limited neck motion, or the suggestion of neurologic compromise, comprehensive evaluation includes history, physical examination, laboratory studies, and appropriate imaging of the spine. When compared to the mature cervical spine, images of the immature cervical spine are more difficult to evaluate due to incomplete ossification and increased mobility of spinal segments. Knowledge of the normal growth and development of the immature cervical spine is necessary to avoid misdiagnoses and to expedite treatment.

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One percent of bone infections involve the spine, and only 4% of spine infections affect the cervical spine [1]. Cervical spine infections in children are rare and are not frequently considered in the differential diagnosis. Lack of experience with the evaluation and treatment of children with cervical conditions is an additional factor that may delay diagnosis and treatment [2]. Inflammatory conditions involving the immature cervical spine are varied and occur more frequently than infections of the pediatric cervical spine. This chapter provides information to allow early diagnosis and treatment of infections and inflammatory conditions of the cervical spine in the pediatric population.

Bacterial Infections of the Cervical Spine

Non-Tuberculous Discitis/ Osteomyelitis

The presenting complaints of insidious, ill-defined shoulder, neck, or upper back pain in otherwise healthy-appearing children and adolescents suggest a relatively benign cause, such as a cervical strain or sprain. Unfortunately, this constellation of complaints may also be expressed by children with infection of the cervical spine [3–5]. While this condition is rare, early diagnosis and treatment of cervical discitis/

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vertebral osteomyelitis are crucial to avoid neurologic consequences and attendant disability. In the past, controversy existed over the differences between discitis and vertebral osteomyelitis. While the former may present solely with pain in a healthy child, the latter is more likely associated with signs of sepsis [6]. Discitis and vertebral osteomyelitis are currently thought to be in the spectrum of the same disease process.

The spread of infection to vertebrae in the absence of an invasive procedure involving the spine is via hematogenous spread [6]. Hassler elegantly demonstrated that blood vessels penetrate disc tissue in the human fetus and neonate and provide a route for infection to spread directly to the disc and the vertebral body [7]. During early periods of growth and development, disc tissue is cellular and has a high rate of metabolic turnover. This is facilitated by thin-walled vessels that exist in a terminal glomerular configuration within cartilage canals. These complexes provide for metabolic exchange and for nutrition of the growing vertebra and disc. With maturation, the vertebral cartilaginous end plates become thinner, and the cartilage canals are incorporated within the vasculature of the mature vertebral body [8]. With hematogenous infection, bacteria have access to the immature cartilage end plates and can establish foci of infection that can then spread to the disc and vertebral body. Delay in treatment allows destruction of bone and disc and may result in neural compression due to accumulating purulent debris.

Vertebral involvement accounts for only 1% of all childhood cases of osteomyelitis, and the cervical spine is involved in 4–5% of these patients. A survey of 28,722 pediatric ED visits revealed 170 patients with neck complaints. Of the 170 patients, 28 had viral infections of the neck and only 5 had bacterial infections [9]. Inflammatory conditions, including juvenile idiopathic arthritis and intervertebral disc calcification, have a much higher incidence than infections in this population. The consideration of potential infection of the cervical spine in children and adolescents is not often included in the differential diagnosis of the abovementioned complaints with resulting significant delay in diagnosis and treatment [10].

Clinical Presentation

Patients with cervical discitis/osteomyelitis typically present with neck pain or headache with concomitant shoulder and upper back pain and no history of trauma or signs of sepsis. Neurologic examination is usually normal except in advanced stages of the disease, in which abscess formation may produce radiculopathy or myelopathy [11]. Less frequent presentations of cervical spine infection include acute torticollis, dysphagia, and respiratory distress. The affected neonate may present solely with irritability or anorexia, normal laboratory values, and no systemic signs of sepsis.

Physical examination, with discrete palpation of the cervical spine, will most commonly reveal local tenderness and muscle guarding with significant loss of neck motion. Torticollis and neurologic compromise are infrequently present.

Diagnostic Tests

Radiographic changes involving the cervical spine will lag behind the onset of clinical complaints by 10 days to 2 weeks and may then only demonstrate narrowing of an involved disc space. In the absence of appropriate treatment, progressive destruction of the disc and adjacent vertebrae is observed [12]. Multiple levels of vertebral involvement with at least two involved segments may be seen [13]. Serial radiographs often demonstrate vertebral collapse, new bone formation, and occasionally progression to spontaneous fusion of vertebral segments when healing occurs. With the formation of an abscess, root and cord compromise may ensue due to volume effect of purulent material or contiguous infection of the meninges but is rare.

MRI is the study of choice when advanced imaging is indicated. Not only does MRI define early bone involvement, but it will also provide earlier evidence of disc involvement [6, 10]. MRI elegantly provides evidence of contiguous soft tissue infection, abscess formation, and signal changes within the cord.

Initial laboratory studies include CBC with differential, ESR, CRP, and blood cultures. The CBC may be normal or may suggest acute infection. As with other sites of bone infection, both

the ESR and CRP are elevated. Recent trends in the evaluation of treatment efficacy suggest that CRP is more reliable in following a treated patient's clinical course. Positive blood cultures are reported in 10–60% of patients [14]. While identification of specific bacteria and antibiotic sensitivity are valuable in guiding treatment, in the absence of positive cultures, empirical antibiotic therapy is indicated. Needle or open biopsy of the spine is rarely performed but is indicated in the face of clinical deterioration despite antibiotic treatment.

Organisms

The most common organism involved in spinal infection is *Staphylococcus aureus*. Other bacteria include *S. epidermidis*, Group B *Streptococcus*, *Pasteurella multocida*, and *Bartonella henselae*. Cases of neonates with cervical vertebral osteomyelitis have been reported secondary to Group B *Streptococcus* and *Pasteurella multocida* [1, 15]. Two five-year-old patients with cat scratch fever have been reported with vertebral osteomyelitis due to *Bartonella henselae.* One patient had thoracic spine involvement, and the other had cervical spine involvement with epidural abscess formation [6].

Treatment

Ideally, treatment success requires identification of the offending organism and the administration of appropriate intravenous antibiotics, based on sensitivities, for 4–6 weeks. While the absolute duration of antibiotic treatment remains controversial, duration is determined on the basis of the patient's clinical response and improvement in laboratory values, especially the CRP [14]. When a prolonged clinical response is noted, long-term oral antibiotic therapy may be empirically indicated to reduce the risk of recurrent infection.

Immobilization of the neck with an appropriately sized pediatric cervical collar or with a pinless halo device will provide relief of pain and maintain the neck in neutral alignment. When an aggressive infectious process results in significant anterior destruction of vertebral bodies, the threat of secondary cervical kyphosis or instability requires more secure immobilization in a halo vest. Surgical intervention to debride infected tissue, drain an abscess, or to stabilize the cervical spine is rarely indicated in the pediatric population. As opposed to the adult spine, spontaneous fusion of the involved cervical vertebrae has been observed in more than 50% of treated patients. In the absence of fusion, serial physical and radiographic evaluations are indicated to detect evolving deformity that may require stabilization [16, 17].

Tuberculosis of the Cervical Spine

Tuberculosis (TB) remains a problem in most of the world, including the United States. According to the Centers for Disease Control, tuberculosis cases have been declining in the United States since 2008, although there were still 440 cases in children younger than age 15 in 2015, with 21.6% occurring in foreign-born children. When the younger group was analyzed, those younger than 5 years of age were most frequently infected [18]. This is of special concern, since infants and younger children are more likely to develop life-threatening forms of TB. Lee et al. recently reported a 13-month-old child who presented with weakness of the right upper and lower extremities. MRI revealed destruction of cervical vertebrae and abscess formation with spinal cord compression [19].

While not all infected individuals become ill, children are more likely to become ill when compared to adult populations. Infants, young children, and immunocompromised children are at highest risk for developing TB meningitis or disseminated TB [20]. The lung is still the most commonly involved organ in tuberculosis, but other areas of the body, such as the spine, may also be involved [21]. The constellation of cough, fever, and night sweats would suggest the possibility of TB, but it should be noted that children may merely present with lethargy or failure to thrive without constitutional signs. The initial clinical presentation of myelopathy in a young child is rare.

Clinical Presentation

The pediatric patient with cervical spine involvement may present with neck pain, limited neck motion, respiratory distress, alteration in voice quality, and neurologic compromise [22, 23]. If a child is foreign-born or has a foreign-born parent, the diagnosis of TB must be considered. Clinical presentation may be classic for pulmonary TB or may be nonspecific. Sputum is difficult to obtain from the infant and young child and is less likely to yield a positive result, since a smaller number of bacteria may initiate infection in this population when compared to adults [24]. Radiographic evaluation of the chest may demonstrate findings consistent with the diagnosis of TB; however, when clinical signs are few and do not suggest the need for radiographic evaluation, TB skin test is indicated in children. The differential diagnosis of radiographic findings includes eosinophilic granuloma, metastasis, osteoblastoma, chordoma, pyogenic osteomyelitis, neurofibromatosis, and changes secondary to retropharyngeal abscess.

Imaging

When the TB skin test is positive, a complete medical evaluation is mandatory. Pain or limited neck motion indicates the need for radiographic evaluation of the cervical spine. Tuberculous infection of the cervical spine is initiated in the vertebral body with destruction of the bone and then involvement of disc as seen on plain radiographs; adjacent vertebral involvement is common. On MRI, soft tissue abscess may be noted anterior to the spine but rarely in the vertebral canal [25]. MRI is best for defining the extent of infection, and in some cases may be diagnostic; especially early on in the process, the bone involvement is evident but the disc spaces are spared, and the opposite pattern forms nontuberculous infections.

Treatment

Treatment involves drug therapy including INH (isoniazid), rifampin, and pyrazinamide [1]. In the presence of abscess or significant vertebral destruction, surgical drainage and debridement with spinal stabilization are indicated [22, 26]. Since the most common site of vertebral involvement is anterior to the spine, laminectomy is not effective in debridement or drainage and will further destabilize the spine [23].

Inflammatory Conditions of the Cervical Spine

Juvenile Idiopathic Arthritis

Juvenile idiopathic arthritis (JIA) refers to a spectrum of chronic arthritis of childhood that are defined by the absence of an apparent cause, duration of clinical problems greater than 6 weeks, and onset of disease prior to 16 years of age. The term JIA has replaced the more familiar terms of juvenile rheumatoid arthritis (JRA) and juvenile chronic arthritis (JCA). JIA occurs in 4–14 per 100,000 children, with a significantly increased incidence in female patients [27]. Involved individuals frequently have a positive family history of autoimmune disease, with a concordance rate of 25–40% in monozygotic twins [28]. Polyarticular JIA and systemic onset JIA will commonly affect the cervical spine. A review of radiographs of 158 patients with JIA revealed significantly smaller vertebrae from C3 to C6, due to earlier growth maturation, when compared with controls [29]. Laiho et al. observed radiographic signs of inflammation of the cervical spine in 98 of 159 adults who had onset of JIA between 6 months and 15.9 years of age. The most commonly observed radiographic change involves ankylosis of the apophyseal joint, with the less common findings of atlantoaxial impaction, atlantoaxial subluxation, and subaxial subluxation [30]. Juvenile onset ankylosing spondylitis is rare but has been reported in a 12-year-old with acute neck pain and limited motion. Nuclear medicine evaluation of the patient revealed significant increased uptake in the cervical spine, at bilateral sacroiliac joints and at the right ankle [31].

Evaluation

Typically patients with JIA present with neck stiffness and limitation of motion. Neurologic

evaluation is usually normal, with no signs of radiculopathy or myelopathy. Radiographs of the cervical spine will initially appear normal. With progression of disease, erosion of the base of the odontoid, settling of the occiput, subluxation of C1–C2, subluxation of the subaxial spine, and diffuse ankylosis of the cervical spine may be noted [32, 33]. MRI is helpful in evaluation of the spinal cord by visualizing hypertrophic soft tissue that may encroach on the cord (Fig. 7.1a–c) [34].

Treatment

The treatment of JIA is primarily medical, with concomitant physical and occupational therapy. Over the past two decades, a host of medications have been developed to more specifically address clinically different patients with JIA. Symptomatic relief of neck discomfort with the use of anti-inflammatory medication and a cervical collar is helpful. Surgical interventions, such as decompression of the spinal canal or surgical stabiliza-

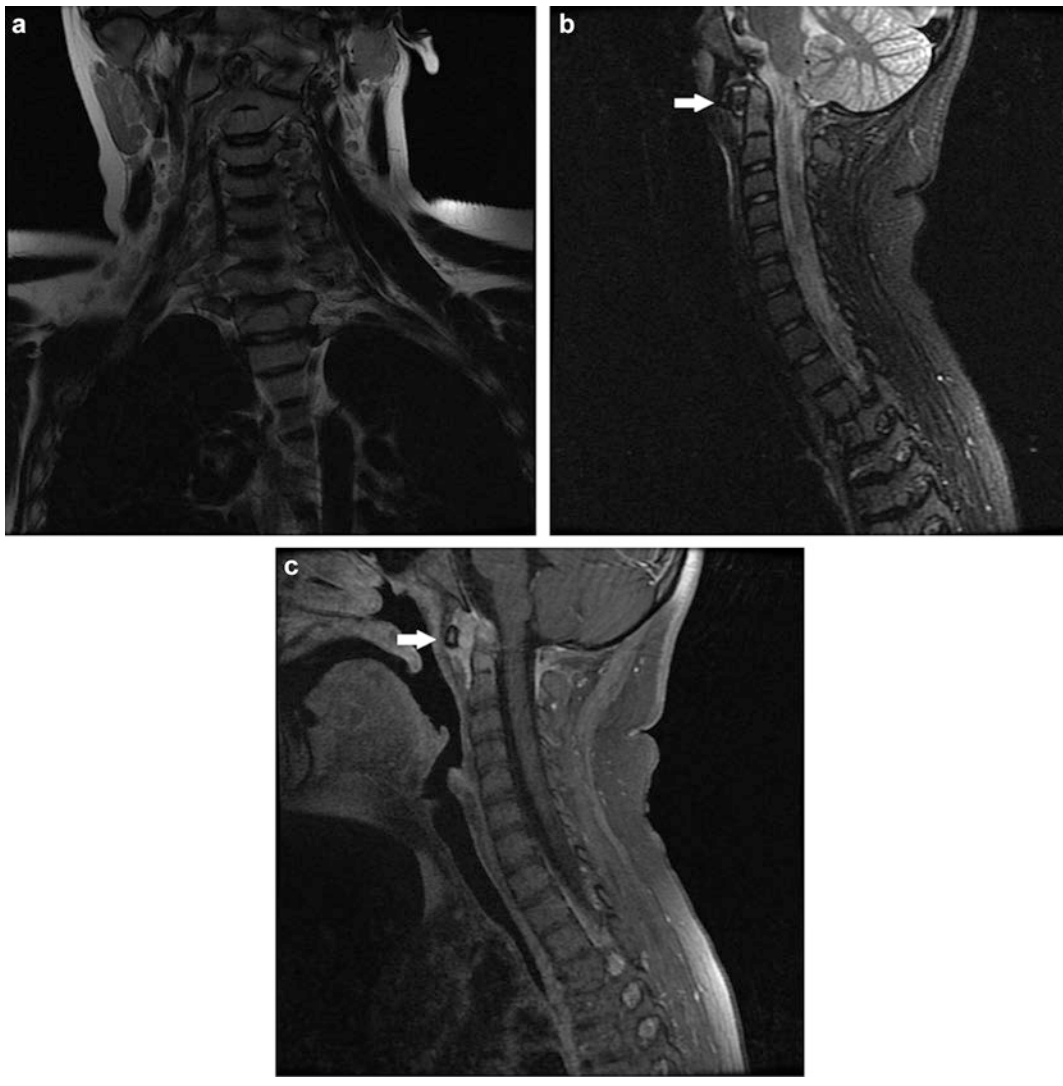

Fig. 7.1 A 12-year-old male with history of rapidly progressive scoliosis and torticollis secondary to juvenile idiopathic arthritis. (**a**) Coronal T2 FRFSE demonstrates left-sided torticollis. (**b**–**c**) Sagittal and axial STIR sequence depicts widening of the predental space with heterogeneous signal and avid enhancement on post-contrast sequences, consistent with pannus. There is also mild increased T2 signal within the odontoid process compatible with the bone marrow edema-like signal and patchy enhancement, concerning for erosive changes. The constellation of findings is consistent with JIA

tion, are more commonly indicated in the adult with rheumatoid arthritis and are rarely necessary in the pediatric and adolescent populations.

Intervertebral Calcification of the Disc

Intervertebral calcification of the disc or calcific discitis of childhood refers to a clinical condition in which patients, with a mean age of 8.6 years, experience the acute onset of neck pain that may be associated with torticollis [35]. Neither trauma nor pharyngeal inflammation has been observed in association with this condition, and the etiology of intervertebral calcification of the disc remains unknown. None of the suggested pathologic processes, including tissue necrosis due to vascular insult, trauma, viral infection, inflammation, vasculitis, and hypercalcemic state, have been supported [36–38].

Evaluation

Physical examination typically reveals limitation of neck motion, with pain at the extremes of motion, occasional torticollis, and normal neurologic examination. Fever, leukocytosis, and elevated inflammatory markers are infrequently observed in this population [36].

Intervertebral calcification of the disc most commonly occurs in the cervical spine but may also be seen in the thoracic spine [36]. Radiographic evaluation of the cervical spine reveals soft, fragmented calcifications within one or more disc. The nucleus pulposus is universally involved, and, less commonly, the annulus fibrosis may be calcified [39]. In addition to calcification within the disc, radiographs reveal biconvex expansion of the disc into the adjacent vertebral bodies. MRI of involved discs exhibit swelling of the disc with loss of signal on T1- and T2-weighted sequences [36].

Treatment

The natural history of intervertebral calcification of the disc indicates it is a self-limited condition with progressive resolution of the calcification within the disc. Pain and limitation of neck motion usually resolve with symptomatic support within 6 months, while the gradual resolution of calcification may take years [37]. Of interest, intervertebral calcification of the disc may be observed as an incidental radiographic finding in completely asymptomatic individuals.

Grisel's Syndrome

Grisel first reported the association of acute torticollis with infection of the neck and attributed this to dislocation of the atlas on the axis as the result of muscle spasm of the occipital muscles in response to cervical lymphadenitis [40]. Medical literature is replete with reports of spontaneous or nontraumatic subluxation/dislocation of the atlantoaxial complex associated with inflammatory conditions. Posterior pharyngitis, upper respiratory infections, mastoiditis, sinusitis, mumps, adenoidectomy, tonsillectomy, cochlear implant insertion, parotiditis, acute rheumatic fever, rheumatoid arthritis, and retropharyngeal abscess (Fig. 7.2a–c) have all been implicated as initiating factors in patients with atlantoaxial subluxation/dislocation [41]. The term "Grisel's syndrome" is appropriately applied to the pathology in all of these patients.

The atlantoaxial joints allow for a great degree of freedom in rotation of the cervical spine. Parke et al. demonstrated the existence of a venous-lymphatic system from the posterior pharynx that drains over the periodontal area. This anastomosis provides a direct route for inflammatory products to travel from the pharynx to the upper cervical spine and results in hyperemia that decalcifies the insertions of the transverse atlantal ligament on the ring of the atlas [42]. The combination of swelling of adjacent soft tissues, such as facet joint capsules, and stretching of the transverse atlantal ligament allows translation of the lateral masses of the atlas on the axis. In extreme cases, rotary subluxation or dislocation results with the unilateral mass of the atlas rotating and translating anteriorly on the lateral mass of the axis. Bilateral displacement of the lateral masses of the atlas on the axis may occur but is rare. The majority of patients remain neurologically normal [1].

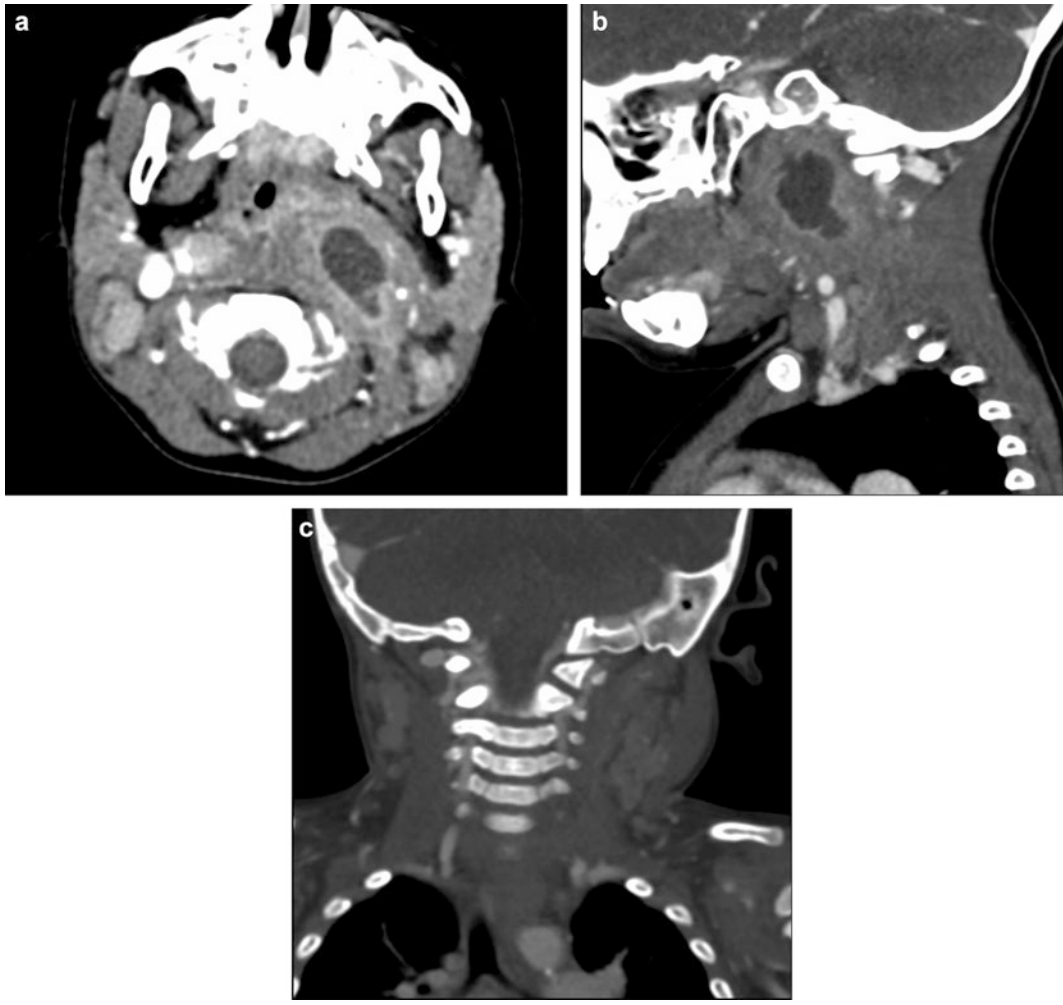

Fig. 7.2 A 15-month-old male with fever, neck pain, swelling, and torticollis. (**a**, **b**) Axial and sagittal images from contrast-enhanced CT neck demonstrate a rim enhancing collection in the left retropharyngeal soft tissues, compatible with a retropharyngeal abscess. (**c**) Coronal image depicts leftward torticollis without osseous abnormality

Evaluation

The patient with Grisel's syndrome will usually present with the acute onset of torticollis that is typically associated with antecedent pharyngitis or a surgical procedure, most commonly tonsillectomy and adenoidectomy. The patient is usually in good health but has marked stiffness and inability to move the neck. The "cock robin" attitude, with the head tilted to one side and the chin rotated to the opposite side, is a classic finding [43]. As opposed to congenital muscular torticollis, in which contracture of the sternocleidomastoid muscle is noted on the side opposite that of chin rotation, the patient with Grisel's syndrome will demonstrate spasm of the sternocleidomastoid muscle on the same side as the chin rotation [44]. Anterior or posterior palpation of the neck does not produce pain; however, attempts to passively rotate the head and neck to a neutral position are resisted and result in distress [45]. Physical examination reveals that the spinous process of the axis is rotated to the same side as the chin rotation. Complete physical examination with documentation of neurologic status is

complemented by laboratory studies to rule out inflammatory conditions and infection. Laboratory studies, including CBC, ESR, and CRP, are usually normal or may display mild elevation of inflammatory markers.

Radiographic evaluation of the upper cervical spine may be difficult to interpret due to the rotation and tilt of the head on the neck. Sagittal view of the cervical spine may reveal true or apparent widening of the atlantodens interval in association with malrotation of the upper cervical spine [43]. CT scan or MRI evaluation will more critically demonstrate partial or complete anterior displacement of the lateral mass of the atlas on the axis on the side opposite the chin [46]. While CT scan has been the most commonly used modality for definitive evaluation of the cervical spine, recent concern in regard to radiation exposure of young children has resulted in an increasing trend toward the use of MRI. The disadvantage of MRI in this young population is the need for sedation or general anesthesia. MRI evaluation is especially important in the absence of trauma or an obvious concomitant inflammatory or postsurgical state because the differential diagnosis of new-onset torticollis includes a posterior fossa tumor [47].

Treatment

When the patient with Grisel's syndrome is examined in the first week of onset of symptoms, treatment with a cervical collar is usually effective in resolving the clinical problem. While spontaneous resolution of the Grisel's syndrome may occur, it is prudent to place the neck at rest, limit activities, and follow the patient closely in 1 week to determine the efficacy of treatment. If the patient is able to demonstrate full motion of the neck in lateral rotation and lateral side bending, progressive weaning from the collar is indicated [1]. If the patient has persistent findings of limited neck motion, halter traction in the inpatient setting will usually result in return of normal motion in less than 48 h. Since the integrity of the transverse atlantal ligament may be difficult to assess with imaging modalities until the subluxation is reduced, inpatient observation is indicated in order to ensure compliance with the traction and for regular neurologic evaluations. When the integrity of the transverse atlantal ligament is compromised, unauthorized removal of the traction device or excessive rotation of the neck within the confines of the halter traction may result in injury to the vertebral arteries or cord impingement.

The loss of integrity of the transverse atlantal ligament requires surgical stabilization of the atlas and axis [48]. Surgical stabilization involves posterior fusion with internal fixation of the atlas and axis. When the transverse atlantal ligament maintains its integrity, immobilization in a firm cervical collar, Minerva cast, or brace is indicated for 6 weeks to allow soft tissue healing. If subluxation recurs, surgical stabilization is indicated.

If the patient has had more than 4 weeks of symptoms when first evaluated, it is likely that surgical stabilization will be required to maintain reduction. Reduction of the atlantoaxial rotary subluxation has been accomplished with halter traction or halo traction followed by immobilization in a halo cast or brace [44]. Case reports suggest success in closed reduction of fixed atlantoaxial subluxation under general anesthesia, but this technique has not been widely accepted as a safe means of reduction [49, 50]. After alignment is achieved, posterior C1–C2 fusion with instrumentation, often supplemented with a halo to control rotation and reinforce the fixation, is applied.

Chronic Recurrent Multifocal Osteomyelitis (CRMO)

Chronic recurrent multifocal osteomyelitis is more appropriately referred to by its recent designation as nonbacterial osteitis (NBO). Clinicians have encountered patients who have presented with symptoms of discomfort and radiographic bone lesions that suggest the diagnosis of bacterial osteomyelitis; however, these

patients have no growth on bacterial cultures and fail to respond to antibiotic therapy. The course of disease may be acute, recurrent, or chronic in nature and may prove confusing to the clinician. Unfortunately, radiographic images that may initially reveal lytic bone lesions that progress to sclerotic or hyperostotic lesions suggest a more aggressive process and warrant biopsy of the lesion for more specific diagnosis. Biopsy of early bone lesions reveals nonspecific inflammatory changes with fibrosis and new bone formation noted in older lesions.

The etiology of nonbacterial osteitis appears to be autoinflammatory. Thirty-one percent of individuals with nonbacterial osteitis have concomitant generalized inflammatory disorders; 12% of their first- and second-degree relatives also carry a diagnosis of nonbacterial osteitis, and 52% have other autoinflammatory diseases [51]. This information suggests a genetic basis. A German National Surveillance Study reported that two-thirds of reported patients were female. The mean age at diagnosis of the 148 reported patients was 11.4 years (1.9 years to 17.9 years of age). The range of time between diagnosis and treatment was a median of 1.7 months (0 months to 9.3 years) [53].

Evaluation

Systemic signs of disease, such as fever, weight loss, or lack of appetite, were noted in one-third of patients, with the remaining patients appearing to be normal. Clinical presentation did not differ when patients with single lesions were compared to those with multiple lesions. Laboratory evaluations, including ESR, CRP, or blood count, were elevated in 78% of patients. Interestingly, ESR was elevated in 72% of patients, versus CRP levels, which were elevated in 40% of patients. Inflammatory markers were higher in patients with multiple lesions or with constitutional signs of inflammation when compared to those with single lesion and no constitutional signs. Cultures of blood or bone aspirate were negative in 87 tested patients [52].

Imaging techniques documented a single bone lesion in one-third of patients and multiple lesions in the remainder of the reported group. At presentation, vertebral lesions were observed in 24% of patients, and lesions of the pelvis, legs, or feet were noted in 90% of patients [52]. Vertebral lesions may be asymptomatic or result in fractures and deformity that require symptomatic treatment. MRI or bone scan will aid in diagnosis of the lesion when performed in conjunction with radiographs.

Treatment

When a patient, especially a very young patient, presents with constitutional signs, including fever, antibiotics have routinely been administered. Since NBO is a self-limited inflammatory process, patients who appear to have been successfully treated with antibiotics may be expressing the natural course of the process. A presumed clinical response to antibiotics would lead the clinician to conclude that the primary diagnosis was bacterial osteomyelitis. Therefore, the true incidence of NBO is unknown and is likely underestimated.

Once the diagnosis of NBO has been confirmed, treatment involves administration of NSAIDs, which usually improve the patient's clinical condition [53]. Recently, early reports of the use of bisphosphonates have been reported to be successful in NBO patients who have not responded to NSAIDs [54]. Bracing or surgery may be indicated to treat spinal or bony deformity that arises from the lesions but, in the majority of cases, neither of these is necessary.

Summary

A variety of inflammatory conditions and infections may affect the pediatric cervical spine. Presentation of patients may vary considerably and is not pathognomonic. Comprehensive evaluation, including history, physical examination, and laboratory studies, is complemented with appropriate imaging of the cervical spine and allows for early diagnosis and treatment.

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Tumours of the Cervical Spine

8

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Introduction

The most common tumours that affect the spine are metastatic in origin. Roughly 7500 primary spinal tumours are detected annually in the USA and account for less than 10% of all spinal neoplasms among people of all age groups [1]. The estimated prevalence of spinal tumours is reported to be 3.6 out of 100,000 people [2]. However, benign lesions are predominant in the first two decades of life. Benign tumours of the cervical spine are less common than their malignant counterparts from the third decade onwards. Metastatic lesions in children are most commonly haematological malignancies (i.e. leukaemia, etc.). Spinal neoplasms may remain entirely intraosseous or affect the paraspinal soft tissues or the dura/thecal sac and neural structures. The tumours affecting the cervical spine in the paediatric age group could be broadly classified into:

- Benign
- Malignant

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The benign tumours are further classified into:

- Bony
- Cartilaginous
- Vascular

Those which affect the neural elements/dura are further classified into:

- Intradural—meningioma, astrocytoma, etc.
- Extradural—neurofibroma, schwannoma, paraganglioma, etc.

The common primary spinal tumours (both benign and malignant) to affect the paediatric cervical spine across different age groups (i.e. <5 years/5–15 years/>15 years) are summarized in Table 8.1 [3].

Some lesions have a predilection to affect the posterior elements (i.e. lamina, pedicle, spinous process), while others tend to affect or involve anterior elements (vertebral body). Haemangioma of the bone and GCT predominantly affect the anterior elements. There exists a wide spectrum of pathological behaviour by benign neoplasms ranging from latent/asymptomatic lesions that are detected incidentally to locally aggressive ones with destructive pattern, and pathological fractures that can potentially cause spinal instability and/or neurological deficit. Yet, these benign neoplasms share a common, clinical presentation, and a majority of patients present with

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Table 8.1 Common musculoskeletal tumours affecting children's cervical spine

Age (years)	Benign	Malignant
0–5	Langerhans cell histiocytosis	Ewing sarcoma Leukaemia Neuroblastoma (metastatic) Wilm tumour (metastatic) Rhabdomyosarcoma
5–10	Langerhans cell histiocytosis Aneurysmal bone cyst Osteoblastoma Osteoid osteoma	Ewing sarcoma
10–20	Aneurysmal bone cyst Giant cell tumour Osteochondroma Osteoid osteoma Haemangioma Fibrous dysplasia	Ewing sarcoma Osteosarcoma Leukaemia
>20	Giant cell tumour Haemangioma Fibrous dysplasia	

pain localized to the area of involvement. This chapter reviews the principles of diagnosis, staging, treatment with case examples, and outcomes for common benign and malignant tumours of the cervical spine in the paediatric age group.

Clinical Examination and Preoperative Workup

The common presentations for tumours of the musculoskeletal system in children are:

- Pain
- Imminent fracture on X-ray or oedema/hyperintense lesion on MRI scan
- Pathological fracture
- Soft tissue mass

However, the most common presentation is pain, and it is present in 76% of benign and 95% of all patients with malignant lesions in the vertebral column [4]. Night pain is characteristic of certain pathologies (i.e. osteoid osteoma and osteoblastoma), and 30–80% of those may also have a co-existent torticollis and radicular pain (instead of a scoliotic list, which is seen with a thoracic or lumbar osteoid osteoma). Torticollis is due to a protective paraspinal muscle spasm, and it resolves with a definitive treatment of the lesion and with time [5]. The incidence of radicular pain varies from 20% to 40% in published literature [6]. Significant relief with an intake of prostaglandin inhibitors like acetylsalicylic acid (ASA) and nonsteroidal anti-inflammatory drugs (NSAIDs) is pathognomonic of osteoid osteoma. Pain relief in up to 70% of osteoblastomas was found to be refractory to NSAIDs in one reported series [7]. The pain of malignant lesions can be acute and severe, with rapid onset of symptoms accompanied by soft tissue involvement or swelling, unlike benign pathologies that are chronic and indolent and are of mild to moderate pain severity without any soft tissue mass. Pathological fractures are uncommon in benign conditions, though their presence may cause instability and/or neurological deficit. The periosteum is highly innervated by nociceptors and has the lowest pain threshold of all the deep sensate structures. The initial evaluation of any child's cervical spine lesion should include:

- Detailed history and physical examination
- Plain X-rays: AP/lateral and open-mouth views (for odontoid peg)
- Dynamic X-rays: Cervical spine lateral in flexion and extension (supervised by a physician)

- Special imaging studies, viz.,
  - Computed tomography (CT)
  - Magnetic resonance imaging (MRI)
  - Single-photon emission computed tomography (SPECT) and positron emission tomography (PET) scans
  - Nuclear medicine (NM) bone scans

History and Physical Examination

A detailed history and physical examination is of paramount importance in all patients. A thorough neurological examination and evaluation of sphincteric function (i.e. the bladder and bowel) is vital and has prognostic significance. Meticulous palpation for any soft tissue mass over the spinous processes, paraspinal area, and front and back of the neck and evaluation for pressure effects on the vital structures (i.e. trachea, oesophagus, and carotid bundle) need to be undertaken. Elicitation of power, tone, and deep tendon reflexes and an assessment of gait are mandatory, and any pathological findings (i.e. upgoing Babinski's sign, exaggerated deep tendon reflexes, clonus or Hoffman's sign) must be recorded diligently. A palpable bony lesion was seen or felt in only 6% of primary benign bone tumours, in comparison to up to 47% of malignant neoplasms [8]. Roughly 1/5 (19%) of all cervical spine tumours are referred to oncologists as soft tissue masses or abnormal swellings. Locally aggressive lesions can cause myelopathy with canal encroachment, as observed with an ABC (aneurysmal bone cyst), GCT (giant cell tumour), and osteoblastoma.

Imaging Studies

The initial method of investigation is always good quality, plain X-rays (anteroposterior and lateral views) with adequate exposure (i.e. covering O–C_1 junction to C_7–T_1 disc space). In a series of 127 consecutive patients with suspected cervical spine pathology, 98% had abnormal radiographs [9]. The most commonly observed findings were:

Table 8.2 Common paediatric cervical spine tumours affecting anterior and posterior elements

Anterior elements	Posterior elements
Langerhans cell histiocytoses (LCH)	Aneurysmal bone cyst
Haemangioma	Osteoid osteoma
Giant cell tumour	Osteoblastoma
Aneurysmal bone cyst	Osteochondroma
Gorham disease	Metastatic lesions
Leukaemia	
Metastatic lesions	

- Presence of a soft tissue shadow
- Destruction of a pedicle—*winking owl* sign

A small subset of patients (especially those with osteoid osteoma) may have normal plain radiographs. The most common region of involvement (i.e. anterior vs. posterior elements) for common paediatric bone tumours of the cervical spine on plain radiographs is summarized in Table 8.2 [10]. The specific radiological features, as well as some unique attributes/characteristics and their significance, are discussed in greater depth under individual pathologies. The dynamic radiographs (i.e. lateral views in flexion and extension) demonstrate spinal instability and the need for MRI-compatible spinal instrumentation at the time of definitive stabilization surgery.

Magnetic resonance imaging (MRI) facilitates an excellent visualization of the soft tissues and the extent or evidence of neural involvement. Their role in evaluation of intradural/extradural and intramedullary tumours cannot be underestimated. MRI scans are the investigation tool of choice in the evaluation of compartmental containment vs. extracompartmental spread. It provides accurate staging, especially in primary malignant neoplasms (i.e. Ewing sarcoma) presenting with soft tissue mass. It is widely used in the preoperative planning of surgical excision, assessment of response to chemo- and/or radiotherapy, as well as monitoring for recurrence. The greatest advantage lies in its noninvasive method of investigation, unlike angiography or myelography, but it requires a great deal of competence in interpreting them. They are also helpful in differentiating pathological fractures

from compressive fractures (hyper intense on T_2 and STIR images).

Computed tomography (CT) scans are especially helpful in early detection of lesions when plain X-rays are normal, as up to 40% of the trabecular bone must be destroyed for X-rays to be abnormal. The central nidus and peripheral sclerosis of osteoid osteoma are well demonstrated on CT (2 mm fine-cut axial bone window). The CT, in addition, would also give a wealth of information regarding the involvement of the neurovascular structures (i.e. vertebral/carotid vessels and nerve roots/spinal cord). The corduroy pattern of haemangioma in the vertebral body and granular ossification in lytic areas of osteoblastoma is well demonstrated on CT scans.

A nuclear bone scan is of great value in detecting the tumour activity and ruling out infections. The old adage *biopsy all infections and culture all tumours* cannot be overemphasized. Infections form the number one differential diagnosis for musculoskeletal neoplasms, especially in the paediatric age group. The most common benign neoplasm that may be picked up on a bone scan and may get missed on plain rays is *osteoid osteoma*. The only benign cervical spine neoplasm that may not exhibit any tumour activity on a bone scan may be *Langerhans cell histiocytosis (LCH)*. Though a bone scan is sensitive, it is not specific enough for a diagnosis of neoplasms, as any condition that affects the metabolic activity or turnover may have increased uptake (i.e. infections, osteoarthritis, fractures, etc.). The predictive value of bone scans for metastasis appears almost 100% [11].

SPECT and PET scans are usually used for the staging of newly diagnosed malignant tumours and to evaluate the response to treatment. The areas of increased glucose uptake in tumour areas are monitored by a positron-emitting radionuclide tracer introduced into the body on fluorodeoxyglucose. A recent study found PET to be a very accurate screening test in diagnosis for all vertebral metastasis in patients with cancer and particularly accurate especially for patients harbouring a non-selective vertebral lesion [12].

Histological Diagnosis: Biopsy

The biopsy provides a definitive diagnosis of a lesion with histological confirmation. However, it is not without complications and should never be taken lightly (including benign pathologies). The key principles one should always adhere to when performing a biopsy are as follows:

- To be performed by a surgeon who will eventually carry out a definitive surgery.
- Direct approach to the lesion with violation of least number of compartments to avoid seedling of tumour cells that would make definitive surgery difficult.
- When using a drain (in open biopsies), the drain should exit in close proximity to the main biopsy surgical incision, which could be included in the final surgical incision at the time of definitive surgery.
- Obtain adequate tissue for frozen section, immunohistochemistry/special stains, and microbiological cultures.
- Secure adequate haemostasis and ideally have a frozen section result before the patient leaves the OR (operating room).

The above generic criteria for musculoskeletal biopsies may be difficult to adhere to in spinal neoplasms, owing to the vital neurovascular structures in the neck. Fortunately, benign lesions of the cervical spine are forgiving to these violations, and malignant ones are very rare, few, and far between! A needle biopsy, either by fine-needle aspiration cytology (FNAC) or core biopsy, can be undertaken and be performed by anterior or posterior approaches. It can be clubbed with definitive excision (especially with benign lesions) as a therapeutic procedure and is called an excisional biopsy.

When performing a needle biopsy, a CT-guided biopsy may provide an accurate representation of the area of interest and reduces false-negative rates. The biopsy could be performed for both anterior and posterior lesions in the neck. For anterior lesions located in the upper cervical spine, the biopsy can be performed through the

thyroid gland, and for lower cervical spine lesions, it is usually performed posterior to the sternocleidomastoid muscle. The success rate of such needle biopsies vary from 50% to 90%. The most common cause of failure in getting a definitive diagnosis with needle biopsies is obtaining (i) nonrepresentative and (ii) non-diagnostic tissue. The use of CT fluoroscopy reduced the operative time in obtaining a biopsy by 50% compared to a conventional CT [13]. More recently, the use of intraoperative 3D fluoro-navigation (O-arm image intensifier) technology has reduced the recurrence rate and allows the confirmation of complete excision of nidus in small lesions (i.e. osteoid osteoma).

Oncological Staging and Treatment Principles

The staging of any neoplasm helps in defining the extent of involvement or spread and in planning treatment options. It is based on histological grade, size, degree of metastasis, and the local spread of the lesion. The Enneking staging system is most commonly used in musculoskeletal oncology and is non-specific to spinal neoplasms. Separate staging systems exist for benign and malignant tumours [14].

The benign tumours are classified into three categories by the Enneking staging system, and their recommended treatments with examples are as follows:

- Inactive (latent): treated by observation (haemangioma of the bone)
- Active: treated by intralesional excision (osteoblastoma)
- Locally aggressive: treated by excision with wide margins (aneurysmal bone cyst)

Most benign tumours are treated by either observation or intralesional excision (i.e. excision through the pseudocapsule and macroscopic tumour residues left behind). The tumour edges are treated with a high-speed burr, electrocautery, dilute 5% phenol (as a chemical cauterization agent), and/or cryotherapy to reduce the risk of tumour recurrence [15]. Utmost care to prevent their contact with the neural tissues (i.e. the dura/nerve root and spinal cord) should be taken to minimize iatrogenic neurodeficit. When a spinal instability is created as a part of the surgical resection of a lesion, MRI-compatible spinal instrumentation and reconstruction of the anterior/posterior column are undertaken to optimize function and outcome [16]. In general, when more than 50% of the facet joints are sacrificed at a single level or when a unilateral facet joint is sacrificed to facilitate the clearance of a tumour mass, posterior instrumented fixation is highly recommended to prevent the development of deformity, secondary to segmental instability. Similarly, laminectomy, especially in children, is notorious for the development of post-laminectomy kyphosis, and instrumented spinal fusion is recommended [17].

Anterior column reconstruction is recommended when more than 50% of the vertebral body is either resected or destroyed by the lesion. Strut grafts (tricortical iliac crest/fibula autografts or allografts) and/or commercially available metallic/carbon cages are routinely used in the reconstruction of the anterior column, supplemented with plate-screw fixation. An external orthosis may be advocated for 6 weeks to 3 months until solid arthrodesis occurs. The surgical stabilization is also influenced by other intrinsic factors like the bone quality, location of the lesion (occipitocervical junction vs. lower cervical spine), and individualization, driven by the patients' unique characteristics and functional needs. Intralesional excision is always preferred over wide excision with the risk of potential instability for all benign neoplasms, except with recurrent giant cell tumours (GCT) and aneurysmal bone cysts (ABC).

An en bloc resection technique may be required for malignant neoplasms (e.g. Ewing and osteosarcoma) [18]. A multidisciplinary, dedicated, paediatric oncology team comprised of medical oncologists and surgeons, coupled with neoadjuvant chemo−/radiotherapy, is a prerequisite for surgical success. An open and

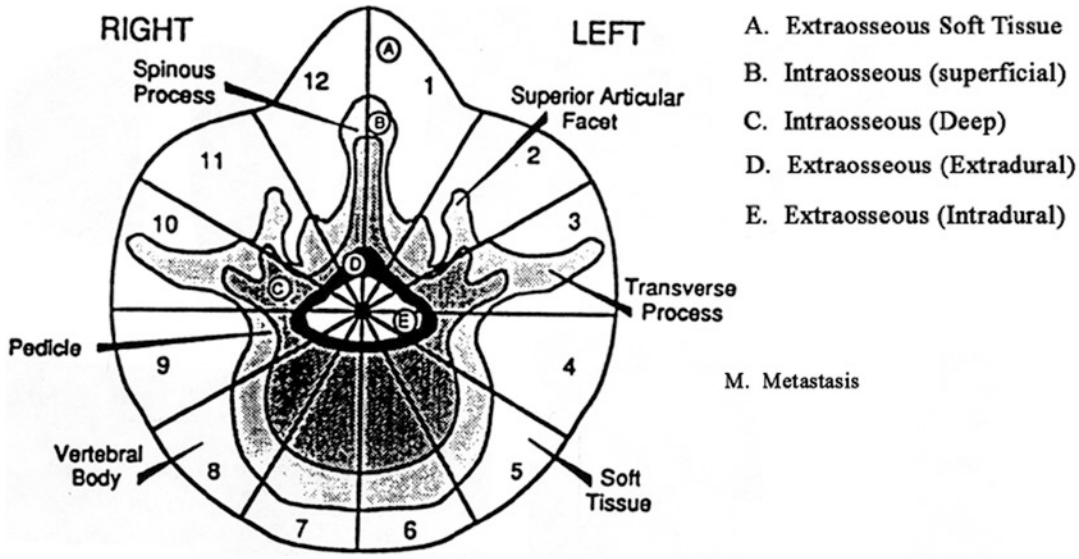

Fig. 8.1 The Weinstein, Boriani, and Biagini (WBB) staging system and representation of 12 transverse zones of a vertebra on axial plane

honest discussion with the parents or guardians about the potential risks, complications, alternative treatment options, and the natural history of the underlying complex pathology is of paramount importance in having realistic expectations and optimizing the patient's experience.

The Weinstein, Boriani, and Biagini (WBB) staging system divides the vertebra on an axial plane into 12 zones that are used to define the borders of a tumour (Fig. 8.1) [6]. This also provides guidelines for the approach and recommendation on the type of reconstruction following surgical excision of the tumour. Lesions involving anterior areas (i.e. zones 4–8 and 5–9) require a vertebrectomy, and those affecting the posterior areas (i.e. zones 10–12 and 1–3) require a posterior approach and the removal of posterior elements. Lesions affecting zones 2–5 or 7–11 would warrant removal of half of the vertebra (i.e. hemivertebrectomy).

Tomita et al. have proposed a two-part numeric classification to accurately express the location of a tumour in the spine, along with the extent of vertebral involvement [19]. It is a refinement and builds on the existing Enneking classification by incorporating a description of affected anatomic site and extent of metastatic involvement. The first set of numbers ranges from 1 to 5 with 1 representing vertebral body and 5 paravertebral area. The second set of numbers ranges from 1 to 7 with lesions 1–3 being intracompartmental, 4–6 being extracompartmental, and subtype 7 representing multiple/noncontiguous/skip lesions (Fig. 8.2) [19]. The main purpose of Tomita classification was to help the treating surgeon in planning curative vs. palliative resection options. More recently, the Tomita classification is incorporated into the Tokuhashi scoring system to form a new algorithm that predicts life expectancy following surgical management in the metastatic disease of the spine [20].

Complications

The salient complications that can occur in cervical spine tumour surgeries are:

- Missed/incorrect diagnosis
- Undertreatment/overtreatment
- Infection
- Recurrence
- Development of post-operative deformity (especially post-laminectomy kyphosis)
- Development of post-operative instability

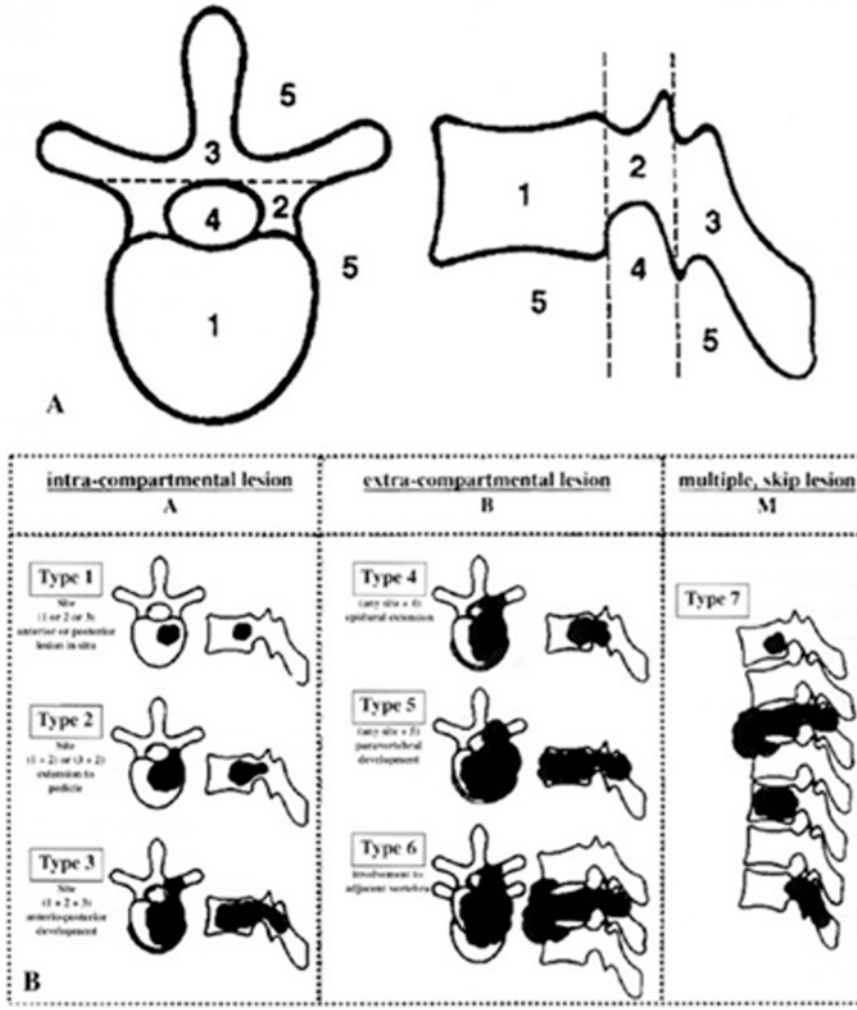

Fig. 8.2 Tomita's two-part numeric classification system with first set of numbers from 1 to 5 and second subset from 1 to 7 (1–3, intracompartmental; 4–6, extracompartmental; and 7, multiple noncontiguous lesions)

The recurrence can be minimized by a multidisciplinary team approach and the judicious use of adjuvant/neoadjuvant treatment (i.e. chemo- and/or radiotherapy). Higher-grade tumours, increased cellular atypia with reactive tumour bed, and intralesional excision are associated with increased recurrence. Irradiated tissues have a risk of undergoing malignant transformation, especially a decade or two after initial treatment, and regular long-term surveillance is mandatory [21].

The overall complication rate reported in literature varies widely from 12% to 92%, and the mortality rate is in range of 2.6–7.7% [22]. Other complications unique to the cervical spine include:

- Airway oedema: treated with definitive airway
- Dysphagia: usually self-limiting and improves in 3–6 months post-op
- Palatal dysfunction
- Injury to vertebral artery
- Risk of dural tear with CSF leak and risk of CSF fistula/secondary infection

Benign Tumours

Osteoid Osteoma

Osteoid osteoma (OO) is the most common benign primary tumour affecting the cervical spine and was seen in 18/41 cases in the series from the Rizzoli Institute (Bologna, Italy) and accounted for 13.5% of all benign lesions of the spine seen at the Mayo clinic [22, 23]. It accounts for up to 9% of primary bone tumours of the spine, and 11–25% of all osteoid osteoma are found in the axial skeleton. It is more common in males, and the mean duration from onset to diagnosis was 19 months in one reported series. The cervical spine is a less common site for OO, in comparison to the thoracic and lumbar spine. OO and osteoblastoma (OB) are histologically identical lesions and have a predilection to affect/involve the posterior elements (i.e. lamina, pedicle, or spinous processes). Often the lesion is characterized by a central nidus surrounded by a sclerotic rim. They are usually ≤15 mm in diameter. OO is at least four times more common than OB, and plain X-rays may be entirely normal. A technetium bone scan is the most sensitive imaging technique in detecting them. Pain is the predominant feature, and torticollis is usually seen when the cervical spine is affected. The involvement of the thoracic/lumbar spine is associated with the development of scoliosis, and OO is usually found in the concavity of the deformity. This torticollis/scoliosis may persist indefinitely despite adequate excision/clearance of nidus, especially when the symptoms are long-standing (i.e. ≥15 months). CT scans are highly diagnostic, and MRI paints an overly aggressive appearance, owing to the surrounding oedema. Night-time pain is also a characteristic feature that can be relieved by intake of NSAIDs/ASA. Histologically, the nidus is composed of a dense layer of osteoblastic cells surrounded by vascular fibrous tissue and ultimately a peripheral layer of mature reactive cortical bone.

The surgical treatment of choice is an intralesional excision of the nidus. The use of intraoperative 3D fluoro-navigation (Iso-C three-dimensional image intensifier) is beneficial to confirm adequate or complete excision [24]. Incomplete excision is associated with recurrence. Kneisl et al. observed that long-term NSAIDs were as effective as a surgical excision of the lesion, though pain relief with NSAIDs was reported to be as low as 30% in one published series [7, 25]. Medical management with NSAIDs is also fraught with the risk of gastric irritation, ulceration, and duodenal perforation. Alcohol, with its vasodilator properties, may precipitate *acute pain crises*. Pain relief is rapid following successful/complete excision of the nidus. Gamma camera scintigraphy facilitates the accurate localization of the nidus, and radiofrequency ablation is also employed in treating OOs. Spontaneous resolution of the nidus over 2–4 years is also reported by some investigators. Figure 8.3a–e depicts an index case of osteoid osteoma arising from right C5 posterior element in an 8-year-old child. The pre- and postoperative axial CT scan images along with histological appearance (in low- and high-power magnification) are shown.

Osteoblastoma

Osteoblastomas (OB) are histologically identical to osteoid osteomas (OO) but are very vascular lesions, and they attain a larger size (i.e. ≥20 mm) [6]. They commonly affect the posterior elements and are less common than OO. The spine is the most common site for OB, and up to 50% of them are found in the lumbar spine. They are equally distributed in the cervical and thoracic spine (i.e. 25%), and they comprise 1% of all spinal neoplasms. They are most commonly seen in children and young adults (i.e. ≤30 years), and they most commonly affect the posterior elements. Vertebral body involvement is rare (<3% of all OBs), and the male/female ratio is 2:1 [4].

Microscopically, the OBs are composed of vascular spindle cell stroma with abundant irregular spicules of the osteoid and mineralized bone, with areas of cystic degeneration or haemorrhage. The features overlap with those of an aneurysmal bone cyst (ABC). However, ABCs may also affects anterior elements (i.e. vertebral body) as

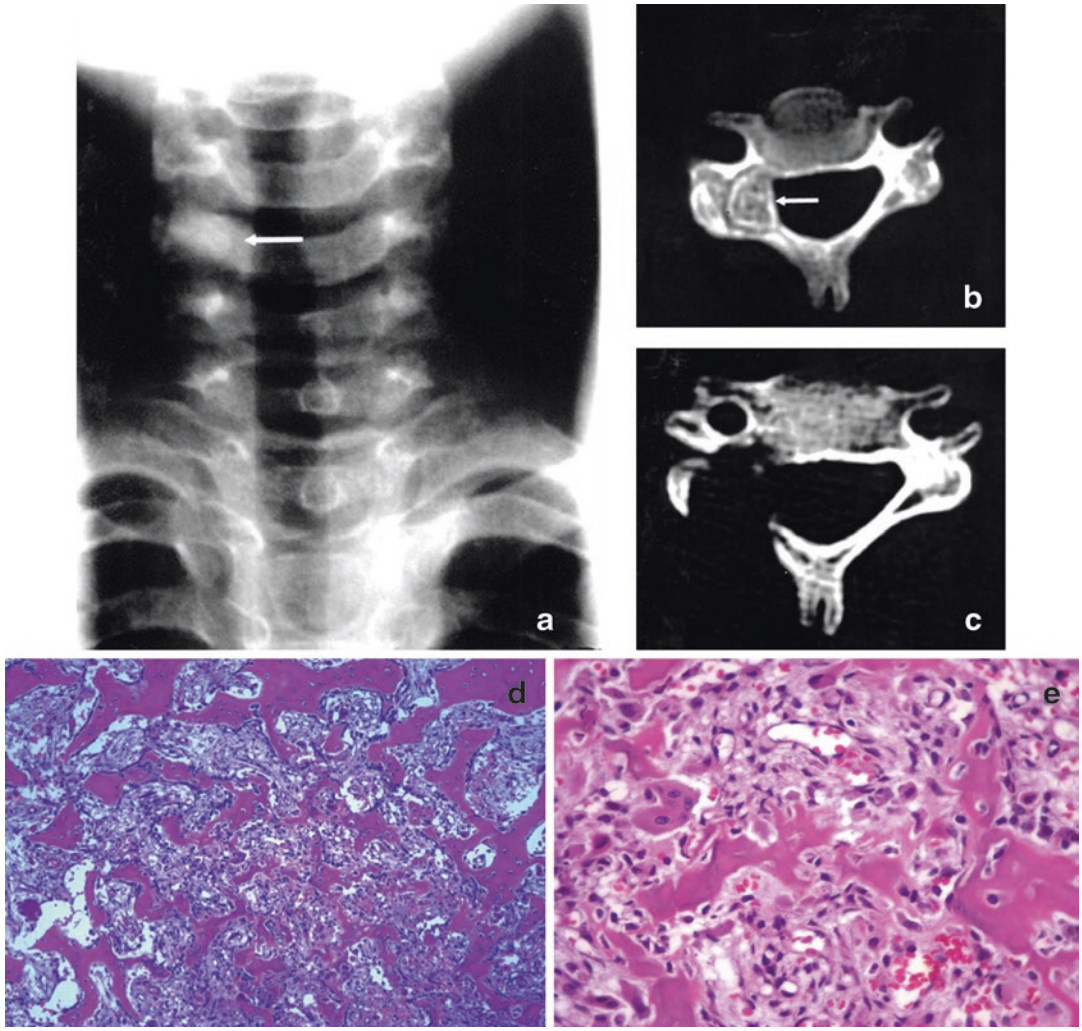

Fig. 8.3 (**a**–**e**) Osteoid osteoma of right C5 posterior elements (lamina-lateral mass junction). (**a**) AP X-ray. (**b**, **c**) Axial CT images before and after resection of the lesion. (**d**, **e**) Low- and high-power magnification views of histology showing osteoid matrix and osteoblastic cells

eccentric, expansile, locally aggressive lesions with cortical destruction and multiple fluid-filled levels. OBs can also be locally aggressive lesions with cortical penetration and adjacent soft tissue involvement. Finally OBs should predominantly contain osteoid elements/cells, and the presence of cartilage cells (i.e. chondroid matrix/chondrocytes) should raise suspicion of malignant transformation into osteosarcoma.

The predominant clinical feature is pain, and its response to NSAID intake is variable and unpredictable. OBs can also cause neurological compression, with local spread into the epidural space by canal encroachment of the neoplastic mass. Plain X-rays may reveal a destructive expansile lesion with a central hyperdense nidus. The destruction is best seen on a CT scan, and an MRI may reveal any impingement of the thecal sac or nerve root.

En masse surgical excision or extended curettage/resection of the lesion is the treatment of choice, and it often warrants spinal instrumentation because facet joints/lamina may need to be sacrificed to facilitate tumour removal [26].

MRI-compatible spinal instrumentation by lateral mass screws and rods is recommended to prevent post-laminectomy kyphosis and/or postoperative instability [16, 17]. Complete excision may not always be possible, owing to the proximity of the OB to the vital structures, and an intralesional excision is fraught with recurrence. Surveillance MRI scans are recommended to monitor for any recurrence. OBs are very vascular tumours, and preoperative embolization of the main/feeder blood vessel is recommended prior to surgical excision [27]. The Rizzoli Institute had one mortality due to excessive haemorrhage in their series of 41 patients with benign cervical spine neoplasms, 38 of whom were treated by surgical excision [23]. The risk of recurrence is strongly correlated with the grade of the lesion. Aggressive high-grade OBs have a 50% risk of recurrence, as opposed to a 10–15% risk for low-grade OBs. Post-op adjuvant radiotherapy, though advocated by some, has not been shown to reduce the risk of recurrence and, therefore, should be used very judiciously. Brachytherapy is recommended for multifocal recurrences [6].

Osteochondroma

Osteochondromas are the most common primary bone tumours in children and adolescents. They constitute 8% of all bone tumours and 35% of all benign bone tumours [1]. They are also referred as osteocartilaginous exostosis, and they most commonly affect the appendicular skeleton. Only 2.5% of OCs are found in the spine [28]. They are hamartomas that develop from aberrant germ cells of the feotal cartilage. They can be either solitary or multiple and sessile or pedunculated. Both males and females are affected equally. They can be a part of multiple hereditary exostosis (MHE), and co-existent exostosis in the appendicular and axial skeleton is common [29]. A majority of OCs in the axial skeleton are asymptomatic and are detected incidentally on a workup for MHEs. They become symptomatic by causing compression on the thecal sac or by causing nerve root pressure/mass effect, and in general, they are usually painless lesions. OCs are most common in the cervical spine, and Dahlin et al. found only eight cervical OCs in a series of 615 patients with exostosis [30]. They most commonly arise from posterior elements but could also affect the anterior elements (i.e. arise from the anterior or anterolateral aspect of the vertebral body). Axial skeletal OCs should be suspected strongly and actively looked for in a patient with MHE presenting with neurological signs or symptoms.

The natural history of OCs is usually growth until skeletal maturity, followed by either a plateau phase or minor regression with time. Spontaneous regression during pubertal growth and adolescence has also been described. They are capped by cartilage cells from the growth plates that are left on the surface of the bone. Rapid increase in the size of an OC with a significant increase in the thickness of the cartilage cap (esp. >20 mm) and acute onset of pain is usually suggestive of a malignant transformation into chondrosarcoma [6]. The presence of calcification in the cartilage cap is suggestive of malignancy. Such a malignant transformation is rare (i.e. <1%) and usually happens in the background of MHE, where the tumour acquires a cauliflower appearance. Acute precipitation of pain may also be seen in stalk fractures of pedunculated OCs.

Asymptomatic OCs do not require any treatment. Surgical excision (intralesional or marginal) is recommended for symptomatic OC, and a complete removal of the cartilage cap is desired to reduce the risk of recurrence. Figure 8.4a–d represents a giant osteochondroma arising from posterior elements of C2. The clinical photograph, sagittal, and axial MRI images, along with histological appearance postexcision, are shown. The perichondrium covers the cartilage cap overlying on the osteoid matrix. Rose et al. have reported an OC of the odontoid peg that caused sudden death by a partial transection of the spinal cord [31].

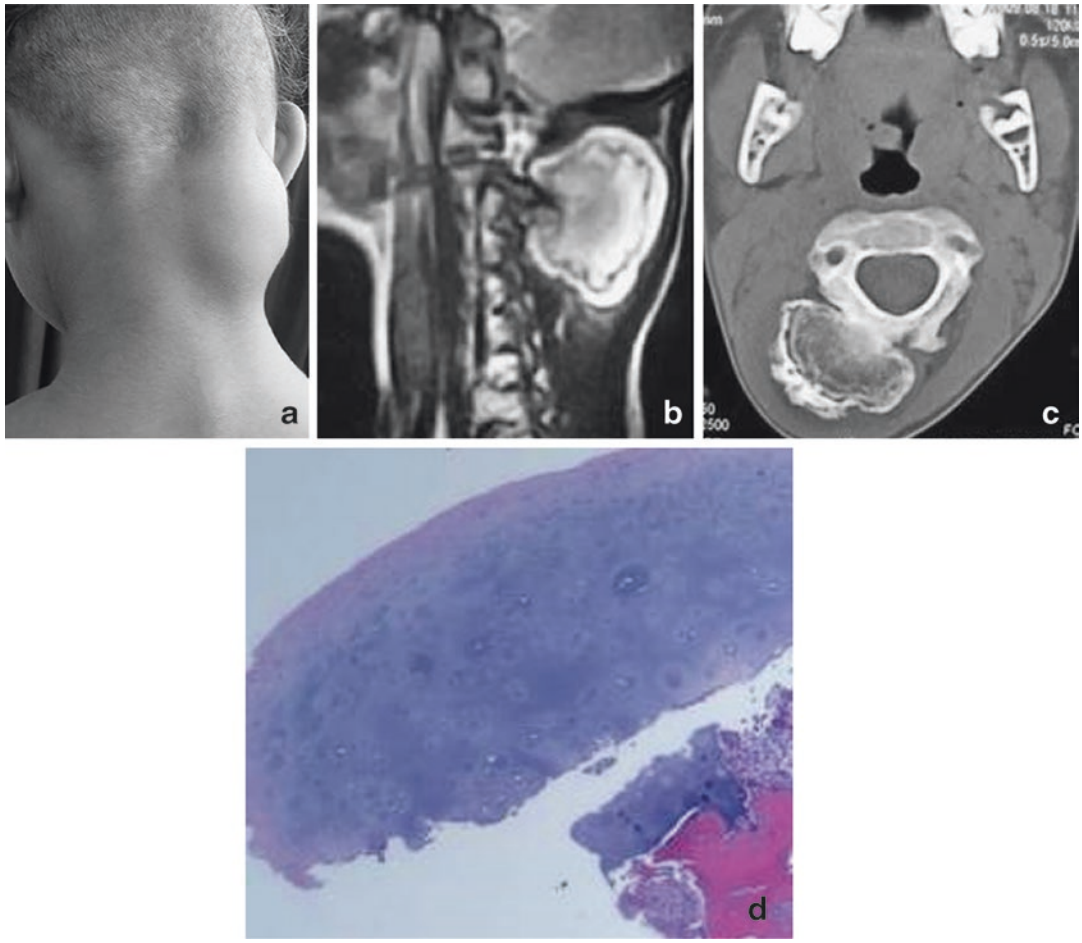

Fig. 8.4 (**a**–**d**) Osteochondroma arising from right-sided lamina of C2 (*axis*) vertebra in a patient with MHE (multiple hereditary exostosis). (**a**) Clinical photograph; (**b**, **c**) MRI sagittal and axial images; and (**d**) histology showing osteoid matrix covered by bluish cartilage cap

Haemangioma

Most haemangiomas are incidental findings detected or observed on evaluation for other pathologies. The axial skeleton (i.e. spine) is the most common site for haemangiomas, and 28% of all haemangiomas are located in the spine [32]. They are most common in the thoracic spine and least common in the cervical spine. They can be broadly categorized into three types:

- Asymptomatic
- Symptomatic
- Aggressive/compressive

From a pathological perspective, haemangiomas of the spine can be of three types:

- Capillary
- Venous
- Cavernous

They differ from aneurysmal bone cysts (ABC) by having an endothelial lining of their walls. A majority of haemangiomas are asymptomatic (>99%), and they most commonly affect the anterior elements (i.e. vertebral body). They were found in as many as 10–12% of people in a cadaveric dissection/autopsy study [4]. Though they

affect males and females equally, the incidence of symptomatic haemangiomas (which are less than 1% of all haemangiomas) is more common in females. Up to 20% of patients may have multifocal, noncontiguous haemangiomas elsewhere in the axial skeleton [23]. In a series of 148 symptomatic haemangiomas, Nyugen et al. found ten of them in the cervical spine [33]. Aggressive haemangiomas were most commonly found to involve the T3–T9 vertebrae.

The symptoms are usually either due to a localized haematoma formation/mass effect or a pathological fracture of the expanded trabeculae. They most commonly present with pain localized to the area of the underlying haemangioma. Neurological symptoms are rare and can be seen when there is compression of anterior radiculomedullary artery, epidural haematoma, or/and expansion of the vascular bone. The senior author (JPD) has reported a case of an aggressive haemangioma presenting as acute cauda equina syndrome in an adolescent [34]. Plain X-rays demonstrate characteristic trabecular striations and are colloquially referred to as *honeycomb* or *corduroy* in appearance. On CT scans, they have a characteristic *polka dot* (i.e. stippled) appearance. The characteristic appearance on an MRI is a hyperintense lesion on both a T_1- and T_2-weighted image, owing to the high water and fat content in the tumour (Fig. 8.5). Aggressive lesions may have an expanded or poorly defined cortex with a soft tissue component.

Asymptomatic haemangiomas do not require any treatment. The natural history of an untreated haemangioma is that of ossification of the affected vertebra. Treatment is recommended only for symptomatic or painful lesions. Treatment options include:

- Vertebroplasty
- Radiotherapy
- Surgical stabilization (for aggressive lesions)

Vertebroplasty provides marked to complete pain relief in most of the patients and has the added advantage of reinforcing the structural integrity of vertebra, preventing collapse. Selective arterial embolization of the feeding vessel can also be undertaken and provides consistent pain relief. Radiotherapy at a dose of 20–30 Gy resulted in complete pain relief in a majority of haemangiomas [35]. Corpectomy with anterior column reconstruction using a strut graft/cage, augmented by anterior plate or screw fixation, may be needed for aggressive haemangiomas causing neural compression. Preoperative embolization of the main feeder vessel is recommended prior to surgical fixation to minimize intraoperative blood loss.

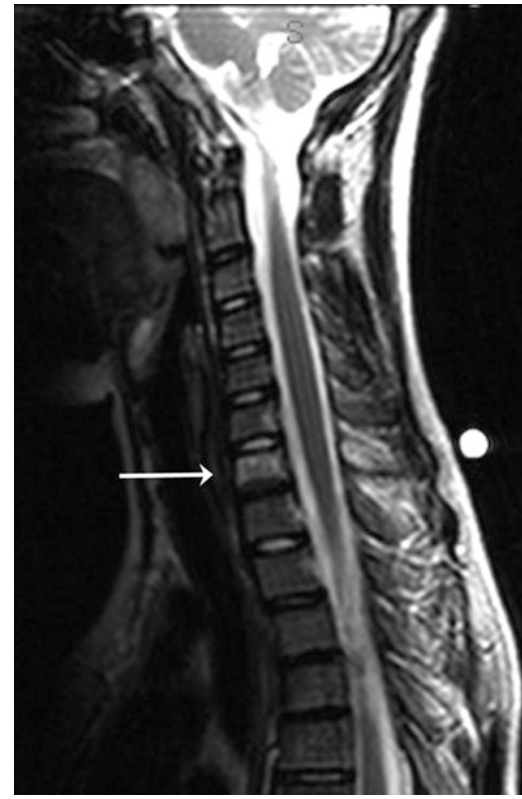

Fig. 8.5 Haemangioma of C7 vertebral body. Haemangiomas are hyperintense on both T_1- and T_2-weighted MRI sequences

Langerhans Cell Histiocytoses (LCH)

LCH is the most well-known childhood histiocytoses and was previously known as *Histiocytoses X*. The hallmark in all three forms of LCH is the presence of a clonal proliferation of cells from

monocyte-macrophage lineage containing Langerhans cells. 10–20% of children with LCH will have vertebral involvement at a mean age of 8 years [36, 37]. LCH constitutes class I histiocytoses and is comprised of a spectrum of three main clinical entities:

- Eosinophilic granuloma
- Hand-Schüller-Christian disease
- Letterer-Siwe disease

Eosinophilic granuloma is a self-limiting condition associated with a focal destruction of the bone and may be either monostotic or polyostotic. Extra-skeletal involvement is not seen in EG, unlike in the other two variants of LCH, and carries the best prognosis. It most commonly affects males aged 5–10 years. Skull bones are most commonly affected. There exists some controversy as to the commonest region in the spine to be affected by LCH, with some studies reporting cervical vs. others as thoracic. Multiple vertebrae in the spine can be involved, and usually a technetium bone scan and/or a skeletal survey are performed to identify other involved sites.

It commonly presents with pain and a local rise in temperature. It most commonly affects the anterior elements and is the most common cause of *vertebra plana*. The vertebra plana is either a partial or complete collapse of the vertebra with a *coin on edge* appearance. The natural history of LCH is that of a complete resolution by conservative management (i.e. watchful observation and symptomatic therapy). However, a biopsy should be performed to confirm diagnosis and rule out other sinister pathologies (mainly Ewing sarcoma and lymphoma). The pathognomonic feature on histology is the presence of *Birbeck granules*. They are tennis racquet-shaped bilamellar granules seen in the cytoplasm of lesional cells (i.e. Langerhans giant cells). Other features on histology include coffee bean-appearing liquid containing histiocytes of the reticuloendothelial system and eosinophils, in addition to Langerhans cells. Letterer-Siwe disease represents the acute disseminated form of LCH with visceral involvement and carries a poor prognosis.

In a series of 26 LCH patients treated by the senior author (JPD), three grades of vertebral collapse were observed: [37]

- Grade I <50% of collapse
- Grade II 51–100% of collapse
- Grade III with Grade II plus involvement of posterior elements

Each grade was further subdivided into two subtypes: (a) symmetric and (b) asymmetric. Grades Ib and IIb (i.e. those with asymmetric collapse) may manifest with spinal deformity (i.e. scoliosis/kyphosis/both), and 4/26 patients did develop one. Neurological involvement in the form of radicular pain in the upper extremities was seen in 3/26 patients, and all three of them had a cervical spine lesion.

The treatment of symptomatic LCH lesions manifesting with persistent pain, and/or spinal instability, includes intralesional curettage with bone grafting with, or without, instrumentation. External immobilization, in the form of a hard cervical collar or custom made cervicothoracic orthosis (CTO), may be needed for 6–12 weeks until sound arthrodesis occurs. There exists no evidence to support that intralesional steroids change or alter the natural history of LCH. Recurrence is rare, and radiotherapy is not recommended for solitary lesions. Systemic multiagent chemotherapy may be needed for disseminated forms of LCH and may also be combined with low-dose radiotherapy (5–10 Gy) for better efficacy. Figure 8.6a is an index case of *vertebra plana* of C3 vertebra in a 9-year-old child. The reconstituted vertebra at 6 years post-op is shown in Fig. 8.6b. The histological features and immunohistochemistry are depicted in Fig 8.6c, d. Birbeck granules are as shown in Fig 8.6e.

Aneurysmal Bone Cyst (ABC)

An ABC is a misnomer, as there is nothing aneurysmal about them. ABCs represent 1.4% of all primary bone tumours, and 5–20% of them are located in the spine [38]. The prevalence of ABC is 1.4 per million population, and they are rare

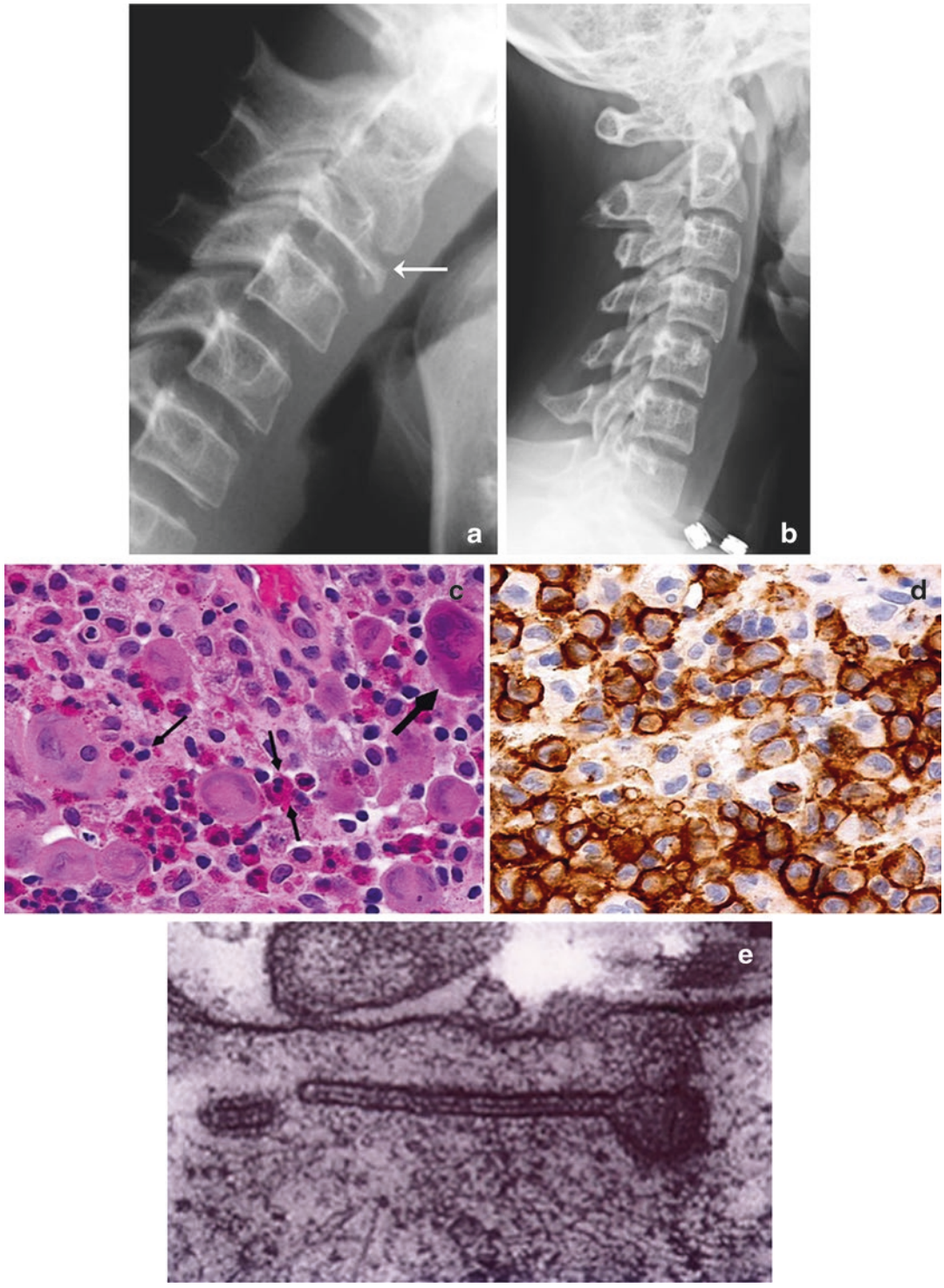

Fig. 8.6 (**a**–**e**) Langerhans cell histiocytoses: plain radiographs of cervical spine lateral view in a child with *vertebra plana* of C2 vertebra. (**a**) Before biopsy; (**b**) 6 years after biopsy showing complete reconstitution; (**c**, **d**) histology showing coffee bean liquid containing histiocytes and immunohistochemistry showing CD1a positivity; and (**e**) characteristic *Birbeck* granule

after the third decade of life. The most commonly affected vertebrae are thoracic. They are locally aggressive pseudotumourous lesions that predominantly affect the posterior elements (in 60–70% of instances) causing eccentric vertebral expansion and cortical destruction with soft tissue involvement [4, 6, 23].

ABCs may affect contiguous 2–3 vertebrae spanning intervertebral discs and may cause regional instability. Unilateral pedicle destruction produces a *winking owl* sign and may potentially cause spinal instability. An MRI scan reveals a characteristic multiloculated, septated, expansile lesion with fluid-fluid levels that produce low intensity on T1-weighted and high intensity on T2-weighted images. The cyst walls lack endothelial lining, in contrast to haemangiomas. Biopsy with an intraoperative frozen section would confirm haemorrhagic fluid with haemosiderin laden macrophages. A CT scan would delineate the accurate extent of vertebral involvement. ABCs may co-exist with other conditions, namely chondromyxoid fibroma (CMF), Langerhans cell histiocytoses (LCH), giant cell tumour (GCT), and chondroblastoma. The solid variant of ABCs does not have a fluid-filled cavity and consists of spindle cells. Both the solid variant and the conventional ABCs can also co-exist together. The soft tissue variant of ABCs does not have osseous involvement, and there are sporadic case reports published of such lesions affecting the appendicular skeleton. However, such soft tissue variant ABCs have not been observed in the axial skeleton/spine. ABCs are characterized by three distinct stages/phases: [4]

1. Growth phase: associated with bony destruction and subperiosteal blowout pattern
2. Stabilization phase: characterized by a distinct peripheral bony shell with an internal bony septa and trabecular meshwork imparting a *soap bubble* appearance
3. Mature phase: bony healing with progressive calcification/ossification of cyst

ABCs are usually symptomatic, causing localized pain either due to a pathological fracture or spinal instability. Neurological symptoms are seen in up to 40% of all patients with ABCs. Historically they were treated by intralesional curettage and the bone grafting. However, this technique is fraught with an increased incidence of recurrence. The senior author's (JPD) four-step approach is comprised of: [15]

1. Aggressive intralesional curettage
2. High-speed diamond burr and electrocauterization of cyst wall
3. 5% dilute phenol application to cyst wall lining (chemical cauterization)
4. Bone grafting (synthetic osteoconductive bone graft substitutes)

This approach was not associated with any evidence of recurrence at a minimum follow-up of 1 year (0/8 recurrence with 4-step approach vs. 4/4 recurrence with mere intralesional curettage and bone grafting) [15]. As ABCs are vascular, preoperative selective arterial embolization is beneficial in keeping intraoperative blood losses to a minimum [39]. An index case of ABC affecting C7 vertebra treated by the four-step approach and combined anterior + posterior spinal stabilization, along with gross macroscopic appearance of excised lesion and histological appearance, is shown in Fig. 8.7a–g.

Calcitonin is also found to be effective in suppressing osteoclastic activity and stimulating trabecular bone formation within the fibrous septa of an ABC [40]. However, external beam irradiation was found to be associated with a risk of recurrence (at least 25%), in addition to the risk of sarcomatous transformation and is currently not advocated for the management of ABCs [41].

Giant Cell Tumour (GCT)

GCTs represent 5% of all primary bone lesions, and vertebral lesions, especially in children, are rare. They are twice as common as ABCs and have a predilection to affect females (the male/female ratio being 1:2) [42]. They tend to predominantly affect the anterior elements (i.e. vertebral body) and are locally aggressive (especially recurrent GCTs) [4, 23]. They are less common

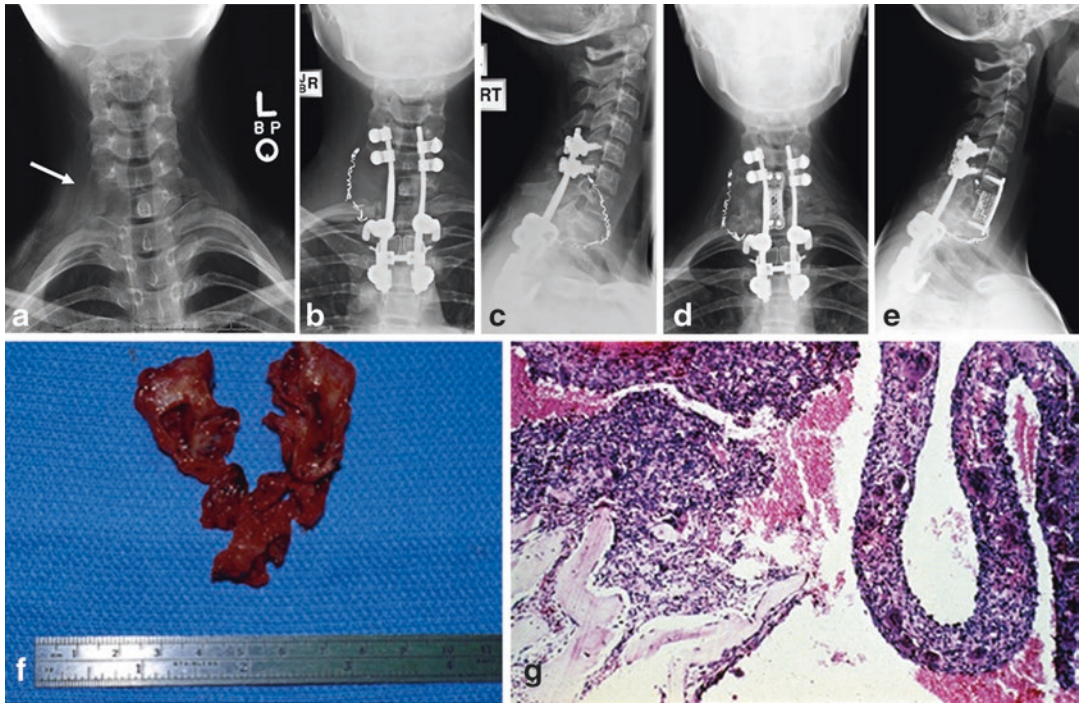

Fig. 8.7 (**a**–**g**) Radiographs of recurrent aneurysmal bone cyst (ABC) with soft tissue mass arising from C7 vertebra. (**a**) AP X-ray showing destruction of right-sided pedicle; (**b**, **c**) AP and lateral X-rays of posterior tension-band fixation following selective arterial embolization of thyrocervical trunk; (**d**, **e**) AP and lateral X-rays following anterior corpectomy, reconstruction of anterior column, and plate/screw fixation; (**f**) macroscopic appearance of cyst wall lining; (**g**) histology showing numerous hemosiderin laden macrophages

in skeletally immature individuals, and they most commonly affect young adults and the middle aged (third–fifth decades). Spinal GCTs account for roughly 15% of all GCTs and are most commonly found in the sacrum. Cervical spine involvement is less common, and the characteristic radiological appearance is that of an expansile lytic cavity in the vertebral body without septation or a mineralized matrix surrounded by the reactive bone. A CT scan accurately delineates cortical destruction or thinning. A co-existent soft tissue mass may be present in many of the GCTs. The characteristic feature on histology is the presence of multinucleated giant cells within the tumour mass, found by the fusion of mononuclear cells. Giant cells are not pathognomonic of GCTs and are also found in other lesions (ABCs, non-ossifying fibroma, chondroblastoma, and osteosarcoma).

Staging of GCTs is done by using a combination of X-rays, bone scans, CT, and MRI scans. Stage II (active) lesions are best treated by intralesional excision and curettage with adjunct therapy, using phenol, liquid nitrogen, and/or methyl methacrylate, whereas stage III (locally aggressive) lesions may need marginal or en bloc resections. Mere intralesional excision without any adjunct treatment is associated with a recurrence of at least 50% and is not recommended. Wide excision is associated with a significant reduction in incidence of recurrence to $\leq$10%. GCTs of the cervical spine are treated by wide excision with corpectomy and anterior + posterior spinal stabilization, using MRI-compatible titanium implants. Selective preoperative embolization of the feeder vessel is desired, as GCTs are highly vascular tumours. Radiotherapy is reserved for unresectable sacral, and/or recurrent, lesions.

Advances in the molecular and cellular biology of GCTs have led to an identification of RANKL (a nuclear factor required for osteoclast formation and expressed by a variety of GCTs). Denosumab, a monoclonal antibody to RANKL, has promising early results in the treatment of GCTs [43].

The biological behaviour of GCTs is unpredictable, and the transformation of stage II and III lesions to overt malignancy is also observed (5–15%). Rapid malignant transformation may cause a pathological fracture with cord or nerve root compression. Pulmonary metastasis, up to 10%, is reported with GCTs, and a high degree of suspicion is needed in detecting them [44]. A CT scan of the chest is the investigation of choice in ruling out the lung involvement.

Recurrent lesions that are not amenable for surgery are treated by:

- Cryosurgery
- Regular/repeated embolization
- Use of interferons
- Bisphosphonate infusion
- High-dose radiotherapy (25–45 Gy).

Miscellaneous Lesions

Gorham Disease It is an extremely rare benign condition characterized by massive regional osteolysis with a proliferation of capillaries. It is a slowly progressive and self-limiting pathology occurring commonly in children and arrests spontaneously by 2 years [6]. It predominantly affects the anterior elements, and the diagnosis is confirmed by histological evaluation. Less than 20 cases are reported in English literature to this day, and the end result is partial restitution with incomplete fusion. The pathogenesis is unknown, and treatment is usually conservative with a halo-vest or collar immobilization. Close monitoring and supervision are needed during the active phase to evaluate for instability and/or new neurology.

Fibrous Dysplasia It is a hamartomatous condition characterized by a *ground-glass appearance* of the affected skeleton on plain X-rays. It could be monostotic (i.e. solitary involvement) or polyostotic. They are largely asymptomatic (i.e. stage I) lesions and are discovered incidentally. Neck pain and torticollis with spasm is the main symptom, owing to the weakening of the trabecular architecture and vertebral collapse. Intense lytic process in stage II and III lesions (i.e. active and locally aggressive forms) may mimic the *vertebra plana* of eosinophilic granuloma. Spinal stabilization is needed when mechanical failure occurs or is impending. Fibrous dysplasia in children is particularly notorious for recurrence, and a bone graft may get resorbed [44].

Malignant Tumours

Primary malignant tumours of the cervical spine are uncommon in children and have a distinct male preponderance, with the male/female ratio being 3.2:1 [6]. The most common tumour is Ewing sarcoma, which is characterized by the presence of numerous small, round blue cells on histology. C2 and C5 appear to be the most commonly affected vertebrae, though chondrosarcoma most commonly affects the C6 and C7 vertebrae [6]. Other lesions that can potentially affect the cervical spine are:

- Osteosarcoma
- Chondrosarcoma
- Metastasis from leukaemia and Wilm tumour neuroblastoma (i.e. PNET family)

Lymphoma and metastasis are also uncommon in children. Chordoma of the craniovertebral junction and sacrum usually occurs in adulthood, though some sporadic cases in the second decade of life (i.e. adolescence) are also reported.

The WBB (Weinstein, Boriani and Biagini) staging system is helpful in describing the zones of involvement and planning surgical resection or spinal stabilization [6]. En bloc resection is the most commonly exercised option in a selective group of patients whose lesions are amenable for surgery. As these are very rare tumours, their diagnosis, preoperative workup, and management

are best performed in tertiary centres with dedicated multidisciplinary oncology teams for surgical success and tumour-free survival. Surgery is often combined with adjuvant/neoadjuvant chemo- and/or radiotherapy, to optimize patient outcomes. A tissue biopsy and a histological confirmation of the underlying pathology are mandatory prior to any definitive en bloc resection. The potential risk of iatrogenic injury to vital structures includes:

- Spinal cord
- Sympathetic trunk
- Vertebral artery and carotid vessels

Ewing Sarcoma (ES)

Of all cases of Ewing sarcoma, 3.5–10% originate in the spine, and up to 75% of all ES occur between 5 and 15 years of age. The average age of ES in the axial skeleton is 16.5 years. ES of the cervical spine is rare, and it most commonly affects the sacrum [4]. Cytogenetic studies typically reveal a translocation of chromosome 11 and 22. ES forms one part of a large family of primitive neuro-ectodermal tumours (PNET), comprising of lymphoma, rhabdomyosarcoma, etc. The mRNA transcript and expression of proto-oncogene '*dbl*' are helpful in distinguishing ES from other PNETs [4].

The most common presentation in symptomatic individuals is pain (seen in 90% of affected individuals), and torticollis, muscular spasms, and a soft tissue mass are usually present. It usually manifests as a painless soft tissue mass with no obvious neurodeficit to begin with (91%), and neurological involvement was reported in only 9% of 344 cases of ES, as reported by Venkateswaran et al. [45]. Another series has reported the incidence of neurological involvement to be 58–64% at the time of diagnosis [4, 6]. Some may have constitutional symptoms in the form of fever, weight loss, and raised biochemical inflammatory markers (i.e. ESR and CRP). An elevated lactate dehydrogenase (i.e. LDH) is a useful blood marker and indicator of the tumour load during follow-up.

The plain X-rays reveal a big soft tissue mass in addition to *moth-eaten appearance* with a permeative pattern of destruction and an aggressive periosteal reaction with a wide zone of transition. Two characteristic of radiological appearance patterns observed were:

(i) Ivory vertebra: Sclerotic appearance with periosteal reaction
(ii) Vertebra plana: Variable osteolysis from focal lesion to complete collapse

The MRI is very sensitive in evaluating soft tissue involvement and neural canal compromise. Metastatic ES is seen in roughly 25% of children, and they most commonly metastasize to the lungs. Other areas of dissemination include the viscera, lymph nodes, brain, and other long bones.

ES is both a radio- and chemo-sensitive tumour, and nonoperative management with these modalities is the first line of treatment. The 3-year survival rate is at least 80% with newer anticancer medications [4]. Local treatment of ES has included high-dose irradiation of the affected area, and the incidence of local recurrence is as high as 25%, despite optimum treatment [46]. The cumulative risk of developing radiation-induced soft tissue sarcoma at 20 years was 42.8% (±17.2%) [47]. En bloc resection is usually advocated for recurrent cases, as survival rates are impressive. Neoadjuvant therapy is usually instituted prior to surgical resection. The prognosis of axial ES is better than its appendicular counterpart. Multiagent chemotherapy has resulted in impressive survival results, and the common agents used include vincristine, actinomycin-D, and cyclophosphamide (VAC) regimen or vincristine, actinomycin-D, cyclophosphamide, and Adriamycin (VAC-A) with, or without ifosfamide/etoposide [48]. Pulmonary irradiation and chemotherapy have markedly reduced the incidence of metastasis to the lungs. Factors associated with poor prognosis include:

- Metastasis at the time of presentation
- Large soft tissue mass/tumour
- High-grade tumour

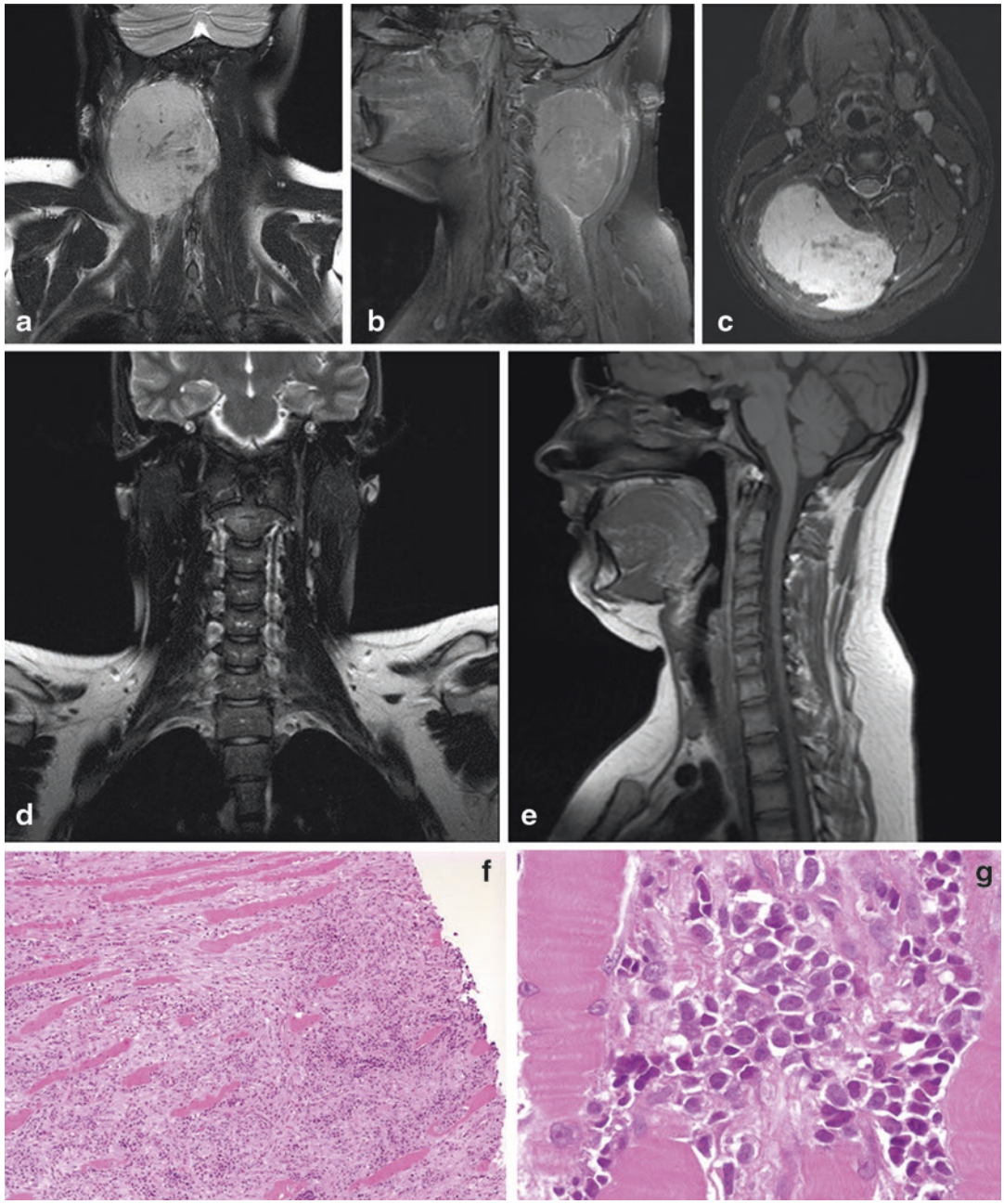

Fig. 8.8 (**a**–**g**) MRI of child with Ewing sarcoma. (**a**, **b**, **c**) Coronal, sagittal, and axial MRI showing isointense T_1 and hyperintense T_2 soft tissue mass arising from right posterior intermediate muscles of the neck; (**d**, **e**) coronal and sagittal post-op MRI following exploration of tumour bed, intraoperative biopsy with frozen section and flap mobilization with closure; (**f**, **g**) histology showing small, round blue cells (low- and high-power magnification)

- Poor response to chemotherapy
- Primary ES arising from epidural space without any osseous involvement (very rare)

An index case of ES of the neck presenting as a soft tissue mass treated by surgical excision and adjuvant chemotherapy by the senior author

(JPD) is depicted in Fig. 8.8a–e. The pathognomonic histological finding is numerous small, round blue cells (Fig. 8.8f, g).

Osteosarcoma (OS)

Though osteosarcoma is the most common primary malignant bone tumour, it is less commonly seen in the axial skeleton and constitutes only 4–5% of all spinal tumours [4]. They are aggressive lesions that cause a permeative pattern of destruction with a wide zone of transition. An abnormal radiodense osteoid matrix is produced by neoplastic cells with periosteal reaction imparting a *sunburst appearance* and causes distant metastases. It is uncommon in children, and most commonly seen in the third decade of life. Neurological abnormality is present in 40%. Mutations in the tumour suppressor gene (*p53*) and retinoblastoma (*Rb*) gene locators on chromosome 17 and 13 are contributory (25–80%). OS commonly occur in families with *Li-Fraumeni* and *Rothmund-Thomson* syndromes. Malignant transformation of the pagetoid bone into OS is uncommon in vertebrae.

An MRI is the investigation tool of choice in the evaluation of OS, and neural encroachment was seen in 84% (i.e. 38/45 patients) in a recently reported series of spinal osteosarcomas [49]. The different histological types/patterns which are reported are:

- Parosteal
- Periosteal
- Telangiectatic
- Dedifferentiated/anaplastic

In a series of 30 cases with the spinal OS by Shives et al., only four were found in the cervical spine [50]. The entire vertebra was affected at the time of diagnosis. En bloc surgical resection is the treatment of choice, and neoadjuvant chemotherapy has eliminated the risk of micrometastasis. The 5-year survival rate with en bloc resection and chemotherapy is 30% [51].

Leukaemia and Haematological Malignancies

The peak incidence of leukaemia is between 2 and 5 years of age, and it is the most common form of cancer to affect young children. The diagnosis is challenging, as the cells are physiologically metastatic from an early phase of the disease process, and surgery has no role in the treatment of leukaemia. Constitutional symptoms are very common, and bone pain is present in up to 25% of affected patients. Peripheral smear, blood tests, and immunohistochemical studies confirm the diagnosis. It is treated by using chemotherapy and radiotherapy.

The vertebral lesions are non-specific and do not help in a definitive diagnosis. Vertebral collapse may be seen in 6% and pathological fractures in 10–15% of affected children [52, 53]. The plain radiographs may show features of osteolysis, osteosclerosis, a permeative pattern of destruction, and a mixed pattern of involvement (i.e. areas of lysis and sclerosis).

Metastasis in Cervical Spine and Miscellaneous Tumours

These include other primitive neuro-ectodermal tumours (i.e. PNET family) which are Wilm tumour, neuroblastoma, lymphoma, and soft tissue sarcoma (e.g. rhabdomyosarcoma and teratoma) [54]. They most commonly present as pathological fractures, and neurological involvement is rare and uncommon. The treatment options are dictated by an underlying diagnosis and are specific to the type of malignancy and stage at the time of diagnosis.

Summary

Tumors of the child's cervical spine are rare and in the first 2 decades of life are usually benign. Metastatic lesions are often part of a hematological disorder. Staging and grading of malignant

tumors follow biopsy and advanced imaging principles as in other tumors. Surgical resection follows principles of anatomical zones.

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Congenital Disorders of the Child's Cervical Spine

9

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Introduction

Congenital cervical spine abnormalities are uncommon and frequently asymptomatic, going undetected throughout life, particularly when they are not associated with a genetic syndrome or with multiple malformations. There is not a reliable true incidence, as many of these abnormalities go undetected [1]. With growth, these abnormalities have the potential of causing progressive deformity and neurologic compromise, so early detection is critical.

The diagnosis of these abnormalities is hindered by the complexity of radiographic evaluation of the cervical spine, since there are variable patterns of vertebral ossification and growth. When there is suspicion of a possible congenital anomaly, the evaluation may include the use of dynamic studies, computed tomography (CT), magnetic resonance imaging (MRI), arteriography, bone scans, and myelography. Physicians treating children must be familiar with the wide variability of normal radiographic findings and have an understanding of possible associated abnormalities related to vertebral formation.

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Concurrent incidence of abnormalities in different organs and systems is 30–60% [2], mostly affecting the urogenital system, followed by the cardiopulmonary system. This strong association stems from the fact that the mesodermal tissue forming the spine is also responsible for the formation of the mesonephros, which in turn gives rise to the mature genitourinary system [3].

The etiology of cervical spine malformations is poorly understood. Genetic anomalies, metabolic conditions, and connective tissue disorders have been postulated as causing vertebral anomalies; nevertheless, most of the time the anomaly is isolated, which is the reason why it has also been proposed to be multifactorial and to have a vascular contribution [4, 5]. Congenital abnormalities of the cervical spine can be found as isolated anomalies or as a part of well-established syndromes. The most commonly associated syndromes to congenital cervical spine abnormalities in which there should be a high suspicion of anatomic and dynamic abnormalities include Klippel-Feil syndrome, skeletal dysplasias, mucopolysaccharidoses, Weaver syndrome, Goldenhar syndrome, Hajdu-Cheney syndrome, Larsen syndrome, Down syndrome, Ehler-Danlos type IV, and Marfan syndrome [6].

The most comprehensive classification to categorize spinal malformations is the one proposed by Tsou et al. [7] and later modified by Tanaka and Uhthoff [8], which is based on the abnormal embryogenetic mechanism involved (Table 9.1) [9].

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Table 9.1 Embryogenetic malformations

Embryogenetic abnormal mechanisms	Anatomic abnormality
Gastrulation	Dysraphic spinal cord malformations
Alignment of sclerotomal rest	Hemivertebrae
Formation	Vertebral wedging, hemivertebrae, caudal agenesia
Segmentation	Block vertebrae, Klippel-Feil syndrome
Vertebral alignment	Congenital vertebral dislocation
Assimilation of sclerotomal cells	Butterfly vertebrae
Ossification and fetal growth	Central vertebral defects, spondylolysis

Congenital Abnormalities of the Occipitocervical Joint

Congenital abnormalities of occiput-C1 are rare in children. It is important to identify the anomalies because they can cause a fixed torticollis, cervical instability, spinal cord encroachment, and myelopathy, and the children can have life-threatening comorbidities. The atlas (C1) ossifies throughout early childhood. Occipitalization of C1 is an occiput-C1 fusion which can be anterior, posterior, or lateral. Basilar invagination is a developmental anomaly of the craniovertebral junction in which the odontoid prolapses into the foramen magnum [10]. This section will define these craniocervical congenital anomalies and discuss associated comorbidities.

Upper cervical spine anomalies are described individually, but clinically they occur in groups and they are highly associated with neurologic signs and symptoms. Hosalkar et al. [11] examined 68 children with upper cervical anomalies (occiput-C3) and found a high incidence of neck pain, torticollis, and neurological changes. Dysmorphic C1 was seen in 42 children, occipitalization of C1 was seen in 22 children, basilar invagination was seen in 25 children, and instability (O-C1 and C1–C2) was seen in 44 children [11]. Most children had 3.4 of these and other upper C spine abnormalities [11]. Myelopathy was present or the child was at risk for myelopathy in 58 children [11]. Forty-four children had a decompression and/or occiput-C2 fusion [11].

Imaging of occiput-C1 is important to define the abnormal elements and any associated neurological abnormalities. Imaging of the base of the skull from plain radiographs is difficult, as the landmarks are indistinct. Chamberlain described a line from the opisthion to the hard palate (Fig. 9.1) [12]. The tip of odontoid should be no more than 5 mm above this line [13]. McGregor modified Chamberlain's line to be a line drawn from the hard palate to the lowest point of the occipital squamosal surface [12, 13]. The tip of odontoid should be no more than 7 mm above this line. McGregor and Fishgold have radiographic measurement techniques at occiput-C1 [11]. Occipital condylar hypoplasia has been diagnosed using a helical three-dimensional (3D) CT scan [14]. Occipital hyperplasia can been diagnosed on MRI and CT [15, 16].

Basilar invagination is often imaged on MRI where the same lines can be drawn. Kulkarni and Goel [17] defined a vertical atlantoaxial index (VAAI). This index is drawn by placing a horizontal line through the endplate of the axis. A second parallel line tangential to the lower border of the anterior arch of the atlas is drawn. A third line parallel line tangential to the superior dens is drawn. The shortest distance between the first two lines is divided by the shortest distance between the first and third lines to determine the

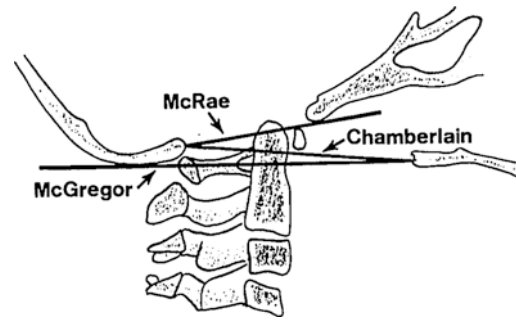

Fig. 9.1 Lateral occiput-C1. Chamberlain's line is from the opisthion to the dorsal hard palate. McGregor's line is from the hard palate the most caudal point of the occipital curve. McRae's line is the opening of the foramen magnum. Used with permission from Hensinger [89]

VAAI [17]. The authors found a VAAI range of 0.2–0.67 in preoperative patients with basilar invagination and 0.76–0.85 in the normal population [17]. The cleidoaxial angle or clivus canal angle is the angle formed by Wackenheim's line and a line constructed along the posterior margin of the dens and the axis body [13]. The normal angle is 150 degrees in flexion and 180 degrees in extension [13, 18].

The first cervical vertebra is best imaged by CT scan. Three-dimensional CT scan has been used to delineate pathology of occiput-C1 [19]. Occipital cervical fusions are often imaged by CT to determine bony anatomy and MRI to determine compression of neurologic structures. Four types of lateral atlantoaxial articulations have been described associated with occipitalization of the axis based on 3D CT scans [19]. Upper cervical spine anomalies are seen in 22q11.2 deletion syndrome, Down syndrome, spondyloepiphyseal dysplasia, and Goldenhar syndrome [11].

Ossification of C1

Ossification of C1 is absent at birth and complete by 7–10 years of age [12]. Ossification occurs around birth in the lateral masses [12]. The anterior portion ossifies at about 1 year of age and the posterior portion ossifies at 4 years of age [12].

Occiput-C1 Fusion

Children with occiput-C1 fusions have the triad of short neck, low hairline, and torticollis similar to the physical findings in Klippel-Feil syndrome. These patients can also present with headaches and a variety of neurological findings [20, 21]. Gholve et al. [22] classified occiput-C1 fusions into three groups based on C1 anatomic zones: zone 1 is a fusion of the anterior arch, zone 2 is a fusion of the lateral masses, and zone 3 is a fusion of the posterior arch (Fig. 9.2a–c). Their study found a high incidence of associated myelopathy. Cervical instability is also common, particularly when associated with C2–C3 fusion [22]. The highest prevalence of spinal canal encroachment was in zone 2 [22]. The incidence of occipitalization of the atlas is 0.32% [23].

The course of the vertebral artery in occipitocervical fusions is well studied [24, 25]. Wang et al. [25] described four patterns of the course of the vertebral artery in patients with occipitocervical fusions. In two of these patterns, the vertebral artery travels posterior to the lateral masses of C2 [25]. This puts the vertebral artery at risk for injury when posterior instrumentation is used. Surgeons may consider a computed tomography angiography (CTA) as preoperative planning when fusing C1–C2 in children with an occiput-C1 fusion.

Basilar Invagination

Basilar invagination is a developmental anomaly of the craniovertebral junction in which the odontoid prolapses into the foramen magnum [10]. Basilar invagination can be caused by a combination of basiocciput hypoplasia, occipital condylar hypoplasia, and absent elements of C1 with spreading of the lateral masses [10]. Twenty-five to thirty three percent of patients with basilar invagination have associated neural axis abnormalities, including Chiari malformation, syringomyelia, and hydrocephalus [10]. Basilar invagination is distinguished from basilar impression as basilar impression is caused by softening of the bones of the skull base [10]. Platybasia is flattening of the skull base [10].

Patients with basilar invagination are stratified into those with and without Chiari malformation [10, 26]. Patients without Chiari malformation, group I in Goel et al.'s series [26], present in the second decade [10]. Common signs and symptoms are weakness, neck pain, posterior column dysfunction, and bowel and bladder disturbance. Focal signs include low hairline, torticollis, and short or webbed neck [10]. Patients with Chiari malformation, group II in Goel et al.'s series [26], presented in the second to fourth decade with

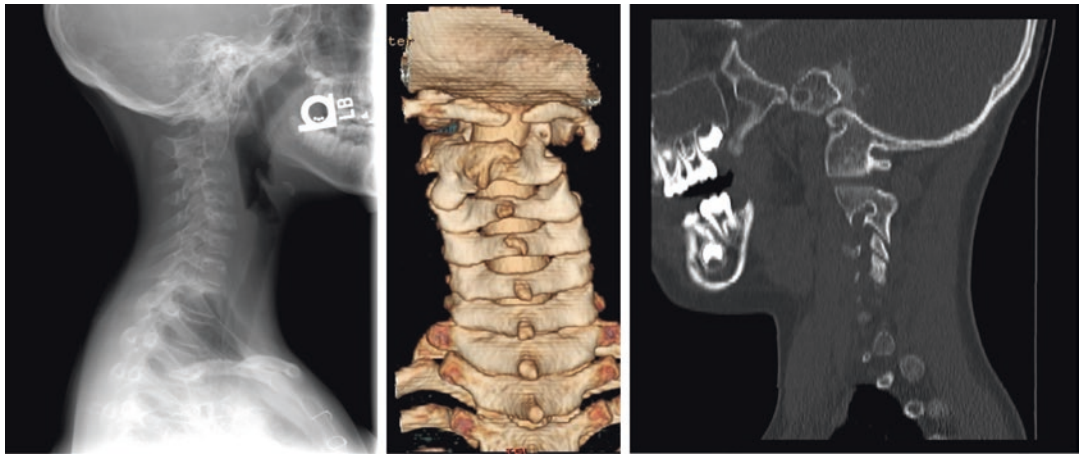

Fig. 9.2 **(a–c)** Radiographs of a 10-year-old female with a fixed torticollis. (**a**) Lateral cervical spine radiograph shows occipitalization of C1 and a C2–C3 fusion. (**b**) CT with three-dimensional reconstruction showing rotatory subluxation at C1–C2 and a posterior cleft at C1. (**c**) CT with two-dimensional reconstruction showing the occipitalization of C1 to be due to an occipital condyle-lateral mass fusion (Gholve Zone 2)

weakness, paresthesias, posterior column and spinothalamic track dysfunction, and ataxia [10]. Focal signs and symptoms are the same as for patients without Chiari malformation. Bollo et al. [18] found that a group of children with Chiari I malformations who failed to respond to decompression had craniocervical anomalies which required reduction and occipitocervical fusion [18]. Specifically, children with a Chiari malformation and brainstem herniation and a clivoaxial angle less than 125 degrees are at higher risk of requiring an occipitocervical fusion [18].

Treatment of basilar invagination can include preoperative traction. Goel et al. [26] recommend a transoral approach with fixation for group I and decompression of the foramen magnum for group II. If the basilar invagination is reducible with traction, posterior fusion is done typically from the occiput to C3 [10, 27–30]. If traction fails to reduce the basilar invagination, a transoral anterior approach is undertaken by an experienced surgeon with a posterior fusion and possible posterior decompression [10]. The combination of basilar invagination and occipitalization of the atlas has been treated with a transoral decompression of the anterior arch of C1 and the odontoid [20].

Atlantal Arch Defects

Defects in the atlantal arch are considered to be a benign anatomic variation, even though it has been associated with neurologic deficit during neck extension. Atlantal arch defects are commonly asymptomatic but occasionally can appear with headache, chronic neck pain, and Lhermitte's sign [31]. These defects can be divided into anterior and posterior defects. Anterior defects occur in the absence of an anterior ossification center that fails to fuse with the lateral masses or by nonfusion between the two anterior ossification centers [32]. For those occurring in the posterior arch, the disturbance is attributed to an abnormal formation, rather than to an abnormal ossification [33].

Imaging and cadaveric studies have estimated the incidence of atlantal defects to be 3.3–4% for posterior and 0.09–0.1% for anterior [34–36]. Associated conditions are gonadal dysgenesis, Klippel-Feil syndrome, Arnold-Chiari malformation, and C6–C7 fusion [33, 37].

The most commonly used system to classify posterior arch defects is the one proposed by Currarino et al. [38], which categorizes these abnormalities into five groups: (A) failure of posterior fusion with resulting midline cleft, (B)

unilateral cleft defects, (C) bilateral cleft defects, (D) complete absence of posterior tubercle, and (E) complete absence of the entire posterior arch. The clinical presentation of atlantal arch defects includes weakness, paresthesia, transient quadriparesis, and atlantoaxial (AA) instability, the latter more frequent in the context of anterior defects, while the presence of myelopathic and neurologic symptoms is more commonly associated with posterior defects [39–41].

It is also important to consider this abnormality in the setting of a traumatic event and distinguish between a Jefferson fracture and a congenital defect. This can be achieved through CT imaging, which shows soft tissue swelling, an irregular cortical wall, and a translation of the lateral masses of >3 mm in a fracture [42]. The role of MRI should focus on the evaluation of the spinal cord and adjacent neural structures and identification of the transverse AA ligament integrity, whose disruption can predispose to instability [31]. If the ligament is intact and there is no instability, consideration can be given to primary fracture repair.

If neurologic symptoms are present, treatment should aim to decompress the neurologic structures and stabilize C1–C2, because minimal trauma can precipitate severe neurological involvement. Methods of fusion include sublaminar wiring, transarticular screw, and screw-and-rod fixation.

Os Odontoideum

Os odontoideum is an uncommon condition which frequently leads to AA instability and risk of spinal cord compression [43]. It is defined as an independent ossicle with smooth circumferential cortical margins separated from the foreshortened odontoid process.

Os odontoideum can be anatomically divided into two groups [44]:

- Orthotopic: Ossicle in an anatomic position that moves jointly with the anterior arch of C1
- Dystrophic: Ossicle migrated toward the clivus and functionally fused to the basion

There is still controversy regarding the etiology of os odontoideum; the two theories most widely accepted are the congenital and acquired. The congenital hypothesis states that the anomaly results from a failed fusion between the dens and body of the axis. It has been supported by reports of identical twins and a familial case with autosomal dominant inheritance [45, 46]. Its weakness is that the location of the nonfused segment rarely is found at the level of the neurocentral synchondrosis, which corresponds to the fusion site between the first and second sclerotomes [47]. The traumatic hypothesis states that an unrecognized traumatic event or repeated microtrauma followed by avascular necrosis and subsequent remodeling of the dens fragments caused the os odontoideum. It is supported by frequent history of a traumatic event, an appearance undistinguishable from an old type II odontoid fracture nonunion, and the location of the gap, which usually extends above the superior facets of the axis [48, 49]. Os odontoideum is associated with Down syndrome, Klippel-Feil syndrome, achondroplasia, and skeletal dysplasias [44].

The clinical presentation of this disorder ranges from being asymptomatic to severe neurological involvement. It has been reported by Fielding et al. [49] and Klimo et al. [50] that, at initial presentation, radiographic evidence of anterior instability is demonstrated in 67–70% of patients, posterior instability in 10–15%, and no instability in 9–13%. The presence of instability has been shown to have poor correlation with the neurologic status, and the space available for cord (SAC) of <13 mm is strongly associated with clinical symptoms [51].

Treatment of incidental os odontoideum is controversial. Spierings and Braakman [52] showed in their series that of the 20 patients treated conservatively, 15 never developed neurologic symptoms and the remaining five developed minor neurologic symptom deterioration [52]. Fielding et al. [49] reported that patients at initial presentation without instability did not develop neurologic sequelae within the next 3 years. Conversely, reports exist of severe neurologic sequelae after minor trauma in patients with

stable os odontoideum [53] and development of late instability and neurologic symptoms. Currently, no predictive factors have been identified to determine which patients will develop instability or myelopathy (Fig. 9.3).

CT and MRI have shown no increased diagnostic accuracy compared to X-rays, but prove to be useful in defining the anatomy and determining if cord compression exists [54]. There are no guidelines for treatment. Currently, there is no strong evidence to support prophylactic surgical management, and it has been shown that asymptomatic patients without instability can be managed nonoperatively with no morbidity over time; however, an individualized risk assessment should be done [55].

It is our opinion that conservative treatment could include asymptomatic patients with no AA instability. This conservative treatment should include counseling about the risks of contact sports, and may restrict contact sports, and encourage annual cervical spine films to include flexion extension lateral cervical spine films. All patients with radiologic criteria for instability, regardless of symptoms, and those who develop symptoms, should be offered surgical treatment. Klimo et al. [5] recommend surgical management for asymptomatic patients who are under 20 years of age because of increased risk of suffering minor trauma. The goals of surgical treatment are to stabilize the atlantoaxial joint and to avoid neurological involvement. The suggested method of stabilization is posterior cervical fusion of C1–C2, which can be achieved with sublaminar wires, transarticular screw fixation [50, 56, 57], and screw rod constructs. More rigid internal fixation techniques have been associated with higher rates of fusion compared with posterior wiring. Dickman and Sonntag [58] showed a 98% fusion rate with transarticular screws versus 86% with posterior wiring. Farey et al. [59] also showed a 33% rate of instability with the Gallie technique with halo-vest immobilization versus 100% fusion rate with transarticular screws. Neural decompression should address anterior bony or soft tissue impingement of the spinal cord or compression posteriorly toward the dorsal arch of C1. The approach should be based on the direction of the compression.

Klippel-Feil Syndrome

Klippel-Feil syndrome (KFS) was originally described by Klippel and Feil [60] with the characteristic triad of short neck, low posterior hairline, and limited neck range of motion. It is now known that less than 50% of patients present with

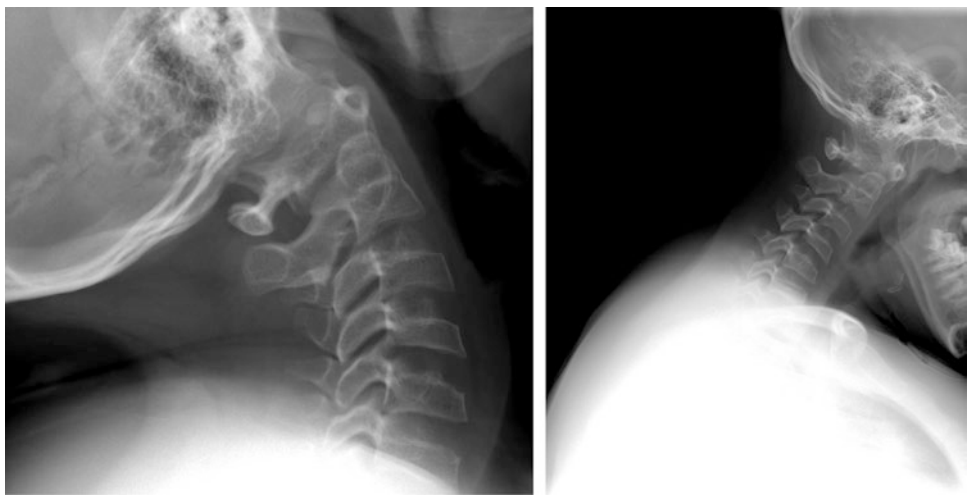

Fig. 9.3 Lateral cervical spine extension and flexion radiographs: os odontoideum associated with C1–C2 instability characterized by displacement of >2 mm of anterior arch of C1 in relation to the odontoid process

all three signs. It includes a heterogeneous group of patients unified by the presence of a congenital fusion of the cervical spine. It is known to be the result of a segmentation defect in the cervical spine; familial occurrence and the association with various chromosomal aberrations support a genetic origin [61, 62].

Feil classified the syndrome into three categories based on the extent and location of the fusion: (I) block fusion of all cervical and upper thoracic vertebrae, (II) fusion of one or two pairs of cervical vertebrae, and (III) cervical fusion in combination with lower thoracic or lumbar fusion [63]. Later, Samartzis et al. [63] developed a classification taking into account only the cervical segment and the number and location of the fused segments, finding that types II and III have an increased prevalence of radicular and myelopathic symptoms:

I: Single congenital fused segment

II: Multiple noncontiguous fused segments

III: Multiple contiguous congenitally fused cervical segments

Genes PAX1, GDF6, and GDF3 have been strongly related to the development of KFS, since they play important roles in somite development. The latter two are known to be causative for an autosomal dominant trait responsible for KFS1 and KFS3, respectively. Recently, a mutation in gene MEOX1was found in chromosome 17q21.31, which plays an important role in maintaining sclerotome polarity and the formation of craniocervical joints, and has been reported to be the cause of the autosomal recessive subtype KFS2 [61, 62].

KFS has a wide spectrum of associated anomalies, but the most frequently encountered are the following [12, 64, 65]:

- Congenital scoliosis (50–70%)
- Rib abnormalities (33%)
- Deafness (30%)
- Genitourinary abnormalities (25–35%)
- Sprengel's deformity (16–20%)
- Synkinesis (15–20%)
- Cervical ribs (12–15%)
- Cardiovascular abnormalities (4–29%)

The presentation of patients with KFS depends on the extent of the affected vertebral segments. KFS can be apparent at birth, but more commonly, affected children tend to present in their teens with complaints of pain, decreased neck range of motion, or deformity [66]. KFS can be asymptomatic and diagnosed incidentally, as affected children often maintain a close to normal range of motion, even though this can be deceptive, since the unfused vertebral segments can become hypermobile and compensate for the fused segment, showing a "normal" range of motion [12].

Up to 68% of patients exhibit cervical symptoms related to mechanical and neurological abnormalities [67]. They originate from open segments which compensate by being hypermobile, which in turn increases the biomechanical demands leading to instability and a faster progression of degenerative changes [68, 69]. The study by Samartzis et al. [63] showed that patients with long-segmental fusions and those with nonconsecutive fusions are at increased risk of developing radicular and myelopathic symptoms. A study by Guille et al. [70], which evaluated KFS patients at a mean age of 35 years, found that 100% had disc degeneration changes and that the MRIs of 86% of patients showed anatomic abnormalities, including disc protrusion, osteophytes, syringomyelia, and narrowing of the craniovertebral junction.

Regardless of the presentation of KFS, high-quality radiographs should be obtained [71]. Flexion and extension lateral cervical views are important, as they provide an assessment of stability. Radiographic images of the thoracic and lumbar spine should be obtained to look for associated anomalies and monitor the development and progression of scoliosis [72]. The radiographic hallmark of KFS is a fused vertebral segment of variable length, even though other features that are also present include flattening and widening of the vertebral body, increased SAC, and hypoplastic or absent disc spaces [73]. Samartzis et al. [74] found that patients with KFS had a higher mean superior odontoid migration (SOM). They also found that

four or more fused segments constitute a factor associated with increased risk of a SOM >4.5 mm. Limitations of radiographic studies for evaluation of the craniovertebral junction in the context of KFS are due to the difficulty of a proper identification of anatomic landmarks, which has been reported to be impossible in up to 18% of patients [75]. For this reason the use of MRI has been advocated due to its high sensitivity in identifying the tip of the odontoid and the various anatomic landmarks [76].

Computed tomography defines the bony anatomy and confirms the extent of the fusion. CT angiography is an important tool preoperatively. Being aware of an abnormal course of vessels in the context of surgical treatment can prevent disastrous complications such as bleeding and posterior circulation stroke [77]. Magnetic resonance is indicated in the setting of a clinical diagnosis that suggests instability or neurological involvement and to determine if associated central nervous system abnormalities are present. Flexion and extension images are particularly useful to directly visualize the SAC and the degree of spinal stenosis.

The most frequent anomaly found in association with KFS is scoliosis, presenting in 50–70% of the cases [12, 72]. Samartzis et al. [78] demonstrated that the most strongly associated risk factor for the development of cervical scoliosis was the presence of vertebral malformations, especially at the low cervical and high thoracic regions, and that the number of congenitally fused segments does not seem to dictate an increased risk for the presence of cervical scoliosis (Fig. 9.4).

Two types of scoliosis have been recognized in KFS: congenital, in which vertebral anomalies are present, and compensatory, in which the curve is found below the area of vertebral involvement. Hensinger et al. [12] found the congenital type to be more common (55%) and more than half required treatment due to the nature of the progression, which consisted of a posterior spinal fusion in the majority of cases.

As previously mentioned, cervical ribs or the fusion of them can be present in 12–15% of cases; it has clinical importance since neurovascular symptoms can develop as a result of nerve root compression or a thoracic outlet syndrome [79].

Sprengel's deformity high association (16–20%) is due to the coinciding time at which scapular descent and KFS abnormalities happen at 3–8 weeks of gestation. It contributes to the abnormal appearance of the neck; when approaching this deformity, it should be done taking into account its complex nature as a combined deformity consisting of an undescended scapula and vertebral fusion. Treatment results can be less effective in this context, compared with isolated Sprengel's deformity patients, particularly when treatment is pursued with a cosmetic purpose.

Hearing loss can be present in up to 30% of patients; it has been suggested to be the result of ankylosis of the ossicles, footplate fixation, an absent external auditory canal, or sensorineural abnormalities [1, 80]. Because of the high incidence of hearing abnormalities, all patients with KFS should have an audiologic evaluation [81].

Genitourinary anomalies have also been highly associated with this syndrome, in up to 35% of patients, probably due to a synchronic development with the cervical spine. The most common manifestation is unilateral renal agenesis. Other renal abnormalities include kidney malrotation, ureteral agenesis, hydronephrosis, double collecting system, and horseshoe kidney. Renal ultrasound scanning offers a good noninvasive diagnostic and screening tool, with the advantage of allowing examination of the reproductive system, which can frequently be affected as well in these individuals. Intravenous pyelography can complement the aforementioned study to better define the abnormal findings and when ultrasound yields inconclusive results [82].

Cardiovascular abnormalities are found less frequently but can represent a source of morbidity if they have clinical impact. The most frequent is a ventricular septal defect, isolated or in combination with other cardiac defects [12].

Synkinesis refers to involuntary mirror movements, generally of the upper extremities. The etiology is unknown; autopsy specimens have shown an incomplete pyramidal decussation, which has been suggested as the cause. Twenty

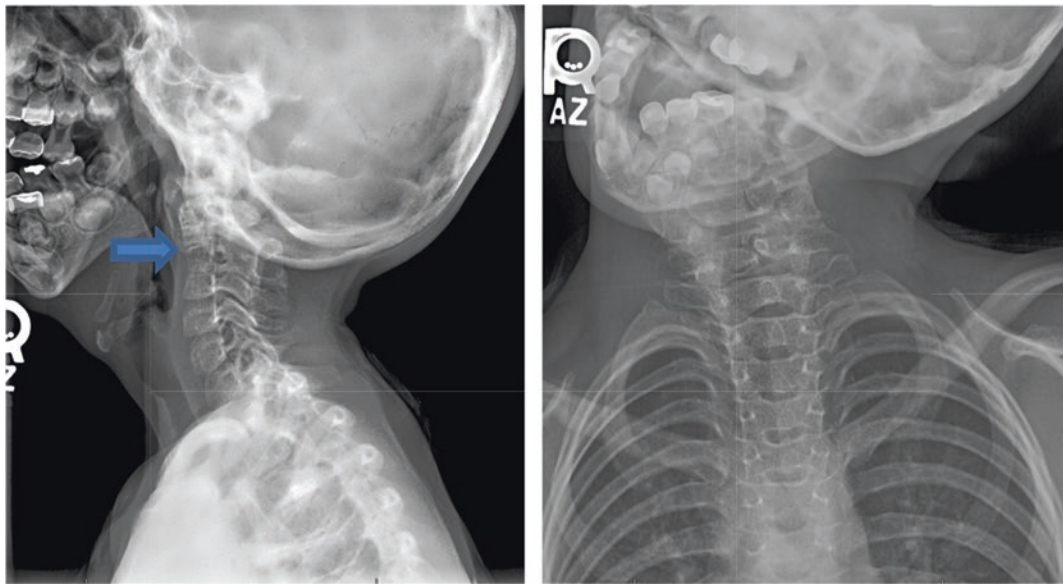

Fig. 9.4 *Left, right*—Lateral and AP cervical spine radiographs of a patient with Klippel-Feil syndrome, with fused C2, C3, and C4 segment (arrow) and cervical scoliosis

percent of KFS patients have this condition, although it has been seen in normal children as well. Synkinesis is more pronounced under 5 years of age and has a tendency to subside with aging and maturation of the nervous system. It can be treated effectively with occupational therapy [1, 12, 83].

Treatment of patients with KFS should be tailored and individualized, depending on the extent of the fused segment, presence of instability, and frequency of symptoms. Patients with three or more levels of cervical fusion should be advised to avoid contact sports and tumbling. The decision on when to stabilize a cervical spine which already has limited motion should be weighted with the risk of developing complications due to instability. Mechanical problems, which usually present during adulthood, originate from degenerative osteoarthritis changes and can be managed with symptomatic relief, which can include gentle traction and use of a cervical collar and analgesics. Progressive symptomatic segmental instability or neurologic compromise warrants surgical treatment.

Surgical treatment should focus on decompressing neurologic structures, stabilizing hypermobile segments, and correcting or avoiding the progression of spine deformities. The surgical procedure should be tailored to the type of deformity and the direction of the compression and instability. A variety of techniques to decompress the neurologic structures exist. Transoral decompression should be used for anterior and brainstem impingement, whereas posterior craniectomy or C1 laminectomy should be used for posterior instability and compression. Instrumentation of the occipitocervical junction is achieved using the traditional techniques of wiring and corticocancellous bone grafting or rods and screws [84]. These techniques should be supplemented by the use of a halo vest to provide additional stability to the fusion site (Fig. 9.4) [85].

When atlantoaxial instrumentation is indicated, traditional techniques such as those of Gallie, Brooks, and Magerl, or using transarticular screw fixation (Fig. 9.5), can be used [86]. Transarticular screw fixation has been proven to be the most biomechanically stable method, and its use is encouraged whenever possible to ensure adequate fusion [86]. Alternatively, C1 lateral mass and C2 pedicle screws connected by rods can achieve excellent stability for arthrodesis. The subaxial segmental instrumentation of the

anterior cervical spine can be done with a plate and screws and an interbody fusion and/or posterior with lateral mass screws and rods. Wedge osteotomies and hemivertebrae resection have been used with variable results to correct deformities but carry a high risk of injury to the neural structures.

Cosmetic treatment can also be done in properly selected patients by means of soft tissue procedures and Z-plasty, providing the neck with a more natural contour and length [87]. It should be kept in mind that the scar might be extensive, and motion most likely will not improve, especially in those patients with presence of an omovertebral bone connection to the clavicle [88]. If this anomaly is detected, resection will allow a higher degree of mobility to the scapula.

Summary

In summary, anomalies of the cervical spine are rare and often are undetected. The anomalies can be life threatening so it is important to be vigilant. The classic triad of low hairline, webbed neck, and limited cervical motion is the presentation of a variety of cervical spine anomalies. Many anomalies are difficult to detect because of apparently normal cervical spine motion. A multidisciplinary approach is essential for optimal surgical management. Multicenter studies are required to determine optimal treatment for os odontoideum and basilar invagination.

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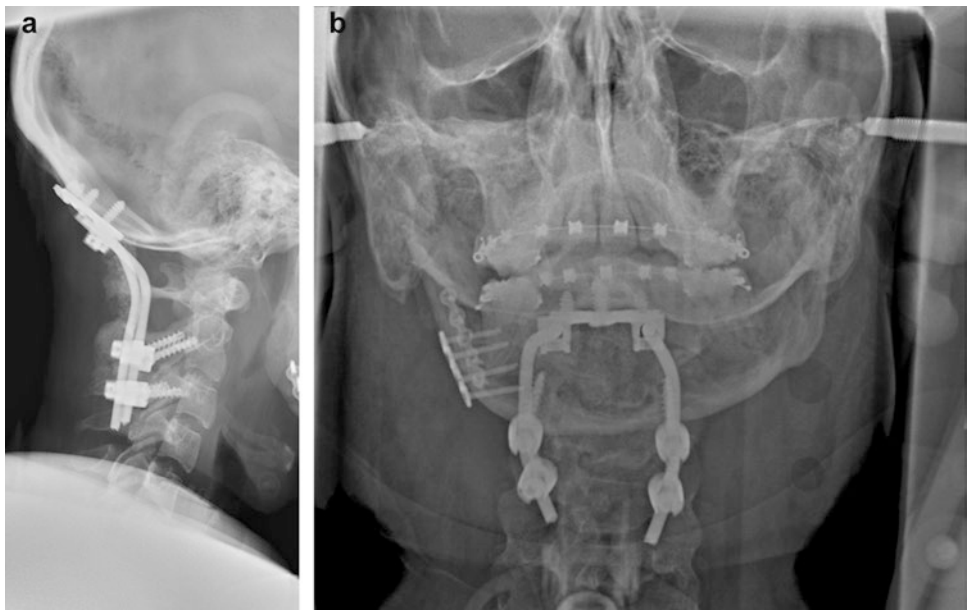

Fig. 9.5 (**a**) Lateral cervical spine and (**b**) AP cervical spine radiographs of a patient with an occiput to C3 fusion with craniovertebral instrumentation

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Dysplasias in the Child's Cervical Spine

10

Jennifer M. Bauer and William Mackenzie

Introduction

Skeletal dysplasias are a diverse group of conditions with altered cartilage and bone development, therefore presenting unique cervical spine pathology and management challenges. Cervical spine involvement, including sagittal and coronal deformity, instability, and stenosis, is routine; resultant spinal cord compression and myelomalacia is common, making screening important. Some deformities are transient, such as cervical kyphosis in diastrophic dysplasia. Others, like cervical instability and stenosis in mucopolysaccharidoses, are progressive and require early recognition and intervention to prevent or reverse neurological deterioration. An understanding of the diagnosis and natural history of these various disorders guides the clinician in their care of the child. This chapter aims to provide a framework to assess and manage cervical spine problems of the more common skeletal dysplasias.

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Skeletal Dysplasia Nomenclature and Classification

Skeletal dysplasias are organized into 33 groups, under which more than 450 types are subclassified [1]. 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, but others may be less clear in presentation.

Dwarfism, or adult height less than 4′10″ (about 147 cm), is a common unifying characteristic of many dysplasias. The children can be proportionate or disproportionate. Although often the entire skeleton is affected, there can be a predominant involvement of the axial skeleton (such as brachyolmia) or the appendicular skeleton. In the appendicular skeleton, disproportionate shortening of the hand, forearm, or arm is termed acromelic, mesomelic, or rhizomelic, respectively. Dysplasias typically reference the location or source of altered development, such as cartilage ("-chondro-") or the vertebrae ("spondylo-"). Table 10.1 includes common dysplasias that present with typical cervical spine pathology. Significant overlapping can occur within the diagnoses. Precise diagnosis is determined in consultation with an experienced geneticist.

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Table 10.1 Common skeletal dysplasia with cervical spine pathology

Dysplasia	Typical characteristic features	Cranio-cervical stenosis	Occipito-atlanto instability	Atlantoaxial instability	Odontoid hypoplasia	Cervical stenosis	Cervical kyphosis	Basilar invagination	Cervical vertebral body hypoplasia
Achondroplasia	Disproportionate rhizomelic dwarfism, frontal bossing, genu varum, midface hypoplasia	+++	–	–	–	–	+	–	–
Pseudoachondroplasia	Normal facies, limited hip and elbow ROM, ligamentous laxity especially hands and wrists, short fingers	–	–	+++	+++	–	–	–	–
Spondyloepiphyseal dysplasia congenita	Short-trunk dwarfism, sad-appearing facies, severe lumbar lordosis balanced with hyperextended neck	–	–	+++	+++	–	–	++	–
Spondyloepiphyseal dysplasia tarda	Similar to SED congenita with later and milder presentation	–	–	++	++	–	–	+	–
Diastrophic dysplasia	Severe rhizomelic dwarfism, rigid clubfeet, hitchhiker thumb, scoliosis, cauliflower ears	–	–	+	–	++	+++	–	+++
Kniest syndrome	Short trunk dwarfism, dumbbell-shaped long bones, sight and hearing problems	–	++	+	–	–	+	+	–
Chondrodysplasia punctata	Rhizomelic dwarfism, intellectual disability, cataracts	–	–	+	–	+++	+	–	–
Metatropic dysplasia	Short-trunk rhizomelic dwarfism, thoracolumbar kyphosis, platyspondyly, restrictive chest	–	–	+++	–	–	–	–	–

Dysplasia	Typical characteristic features	Cranio-cervical stenosis	Occipito-atlanto instability	Atlantoaxial instability	Odontoid hypoplasia	Cervical stenosis	Cervical kyphosis	Basilar invagination	Cervical vertebral body hypoplasia
MPS I – Hurler	Progressive hepatosplenomegaly and intellectual disability, deafness, clouded cornea, obstructive airway disease, short lifespan	++	–	++	++	–	–	–	–
MPS IV – Morquio	Short-trunk dwarfism, pectus, thoracolumbar kyphosis, genu valgus, joint hypermobility	+++	–	+++	+++	–	+	–	–
MPS VI – Maroteaux-Lamy	Normal intelligence, cardiac valve dysfunction, restricted joints, excessive lumbar lordosis, deafness, clouded cornea	++	–	+	–	–	–	–	–

(+) at least one case reported; (++) common; (+++) universal or nearly so
MPS mucopolysaccharidoses

Atlantoaxial-Occipital Complex Abnormalities

Base of Skull Abnormalities

The base of the skull is composed of the ethmoid, sphenoid, petrous part of the temporal bone, and the occiput, which are formed by endochondral ossification. Abnormal development at the base of the skull occurs in several skeletal dysplasias and causes foramen magnum or cranio-cervical stenosis, hydrocephalus, platybasia, occipitalization of C1, and basilar invagination.

Cranio-Cervical Stenosis

Achondroplasia classically exhibits foramen magnum stenosis, as well as cranio-upper cervical stenosis which is a trait shared with the mucopolysaccharidoses (MPS). Defective endochondral ossification is to blame in achondroplasia, while deposits within surrounding tissue at the cranio-cervical junction in Morquio syndrome (MPS IV) and a thickened posterior longitudinal ligament in Maroteaux-Lamy syndrome (MPS VI) are the causes in MPS [2–4]. The symptoms and signs may be subtle and 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 [5] and has been reported at up to 7.5% in the first year of life [6]. The risk is highest in the first 2 years of life and diminishes with the expansion of the spinal canal. The American Academy of Pediatrics' 2005 recommendations call for close neurologic exam, a sleep study investigating central apnea, and a CT to measure the foramen size for all patients with achondroplasia [7]. However, surgeon-led studies since then have found poor correlation of CT findings with neurologic symptoms and central sleep apnea, the latter two being the best indicators for needing surgery [8, 9]. We prefer to follow their consensus recommendations and obtain an MRI if a sleep study and physical exam are positive. Foramen magnum decompression is then indicated in symptomatic patients with documented stenosis (Fig. 10.1a, b).

Hydrocephalus

Children with achondroplasia are also at risk for hydrocephalus in early life. Symptoms include irritability, lethargy, and vomiting. Head circumference measurements should be done regularly and plotted on the normative achondroplasia chart [10]. Any upward crossing of the percentile would warrant further investigation. The defective endochondral ossification from the FGFR-3 mutation and early closure of synchondroses which leads to the above foramen magnum stenosis also causes jugular foramen stenosis. It is thought that diminished jugular return due to jugular foramen stenosis results in hydrocephalus [11]. With growth, the risk of spinal cord compression and hydrocephalus is diminished significantly. This realization has led to a decreasing trend in the number of shunts placed in this population [12].

Platybasia

Platybasia literally means flattening of the skull base. It has been reported in patients with Kniest dysplasia [13] and cleidocranial dysostosis [14]. 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 widens. 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). Figure 10.2 shows a patient with Kniest dysplasia with a clivus-canal angle of 139. 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 [15].

Basilar Invagination

In addition to that which is secondary to platybasia in Kniest and cleidocranial dysostosis, basilar invagination is also seen in spondyloepiphyseal dysplasia (SED) congenita [16], which is a type II collagen defect. Typical signs and symptoms

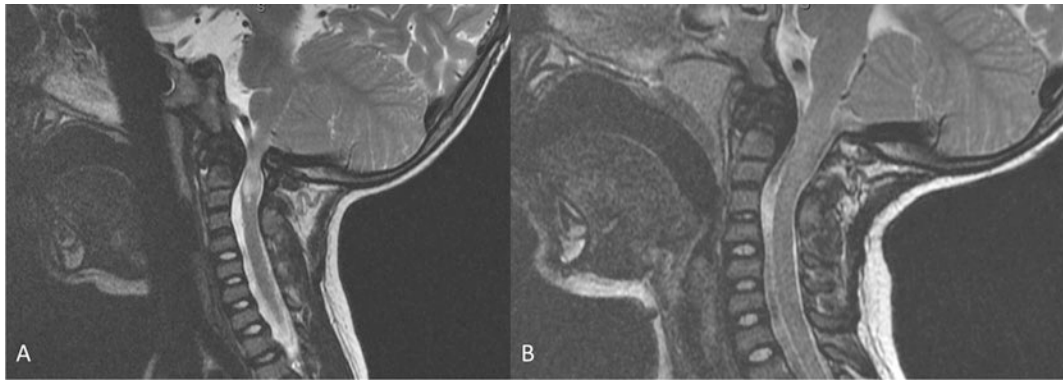

Fig. 10.1 (**a**, **b**) Achondroplasia patient with foramen magnum stenosis and myelomalacia before (**a**) and after (**b**) foramen magnum surgical decompression

are similar to those previously described for stenosis at this level, including hypotonia, and hyperreflexia, with the addition of characteristic nystagmus and cranial nerve dysfunction. A treatment algorithm starts with Minerva-type orthotic immobilization, followed by traction if symptoms persist. If traction reduces the invagination, then posterior occipitocervical decompression and fusion is sufficient; if irreducible, a ventral decompression followed by posterior fusion may be necessary.

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 results in occipitalization of C1 and potential basilar invagination. Occipitalization is reported in patients with Goldenhar syndrome and Russell-Silver syndrome [17].

Upper Cervical Spine Instability

Upper cervical spinal instability includes occipitocervical 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 manifestations range from asymptomatic to quadriparesis. In older children, the earliest symptom may be gradual loss of physical endurance. In toddlers and younger children, gross and fine motor developmental delay can occur.

Occiptocervical Instability

O-C1 stability is determined by ligamentous strength and the shape of the articulation. Uniquely, there is no intervertebral disc at this segment. The bony articulation of atlas and occipital condyles are shallow and broad, with relatively lax capsules to allow motion. The cord-like alar and apical ligaments, which arise from the odontoid to the anterior and medial aspects of the occiput, and the tectorial ligament, a continuation of the posterior longitudinal ligament to the 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 [18]. Figure 10.3 demonstrates this instability.

Atlantoaxial Instability

C1-C2 stability depends on combinations of bony and ligamentous restraints. The associated delay in ossification often seen in skeletal dysplasias results in odontoid abnormalities that range from odontoid aplasia, to hypoplasia, to os odontoideum [19]. Failure of closure at 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

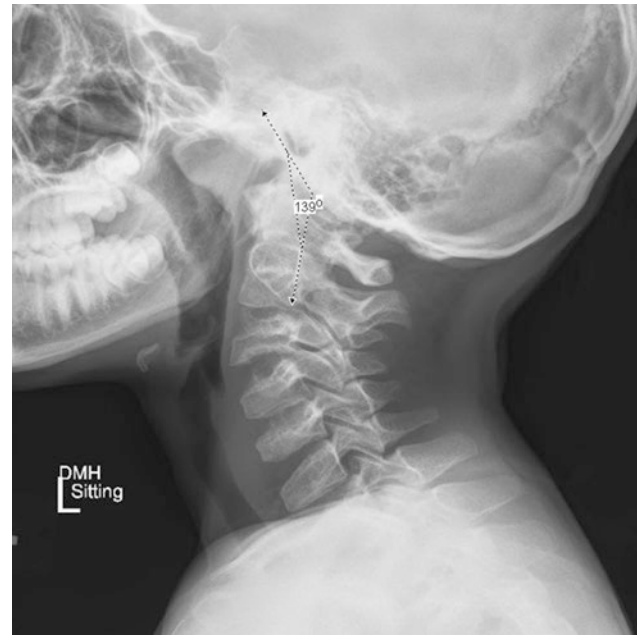

Fig. 10.2 Kniest patient, 12 years old, with clivo-cranial angle of 139°, with less than 150° indicating platybasia

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 [20]. 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. Moreover, normal odontoid morphology does not preclude one from instability, as ligamentous laxity also plays a role.

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 os odontoideum, there is mobility. Anterior subluxation can be due to odontoid abnormalities, ligamentous laxity or absence of ligaments (Fig. 10.4). In a lordotic cervical spine, a fixed posterior subluxation is probably more tolerable than an immobile anterior subluxation.

Multiple skeletal dysplasias are associated with odontoid abnormalities and atlantoaxial instability. Pseudoachondroplasia has a reported 60% incidence of os odontoideum which leads to worsening instability with age, yet commonly without myelopathy [21]. The family of MPS has a high rate of instability, with Morquio syndrome (MPS IV A) in particular demonstrating universal findings of odontoid dysplasia [19, 22, 23]. This, combined with a common extradural soft tissue thickening [3, 19], leads to stenosis with myelopathy (Fig. 10.5) and in some cases quadriparesis and death, prompting some to recommend prophylactic occipitocervical fusion [24, 25]. Because symptoms start at 4–6 years old, we favor clinical and radiographic monitoring and fusion at initial appearance of symptoms or MRI cord changes. It is our experience that cervical myelopathy presents earlier in patients with spondyloepiphyseal dysplasia congenita (Fig. 10.6) compared to patients with Morquio syndrome, often in infancy, though may appear radiographically similar [23]. Kniest dysplasia, a type 2 collagenopathy like SED, and Larsen syndrome, a filamin disorder, can have similar instability, as can cartilage-hair dysplasia, metatropic dysplasia, and congenital nasopharyngeal abnormalities such as cleft palate [26]. Diastrophic dysplasia's upper C-spine instability is less common than its characteristic cervical kyphosis but

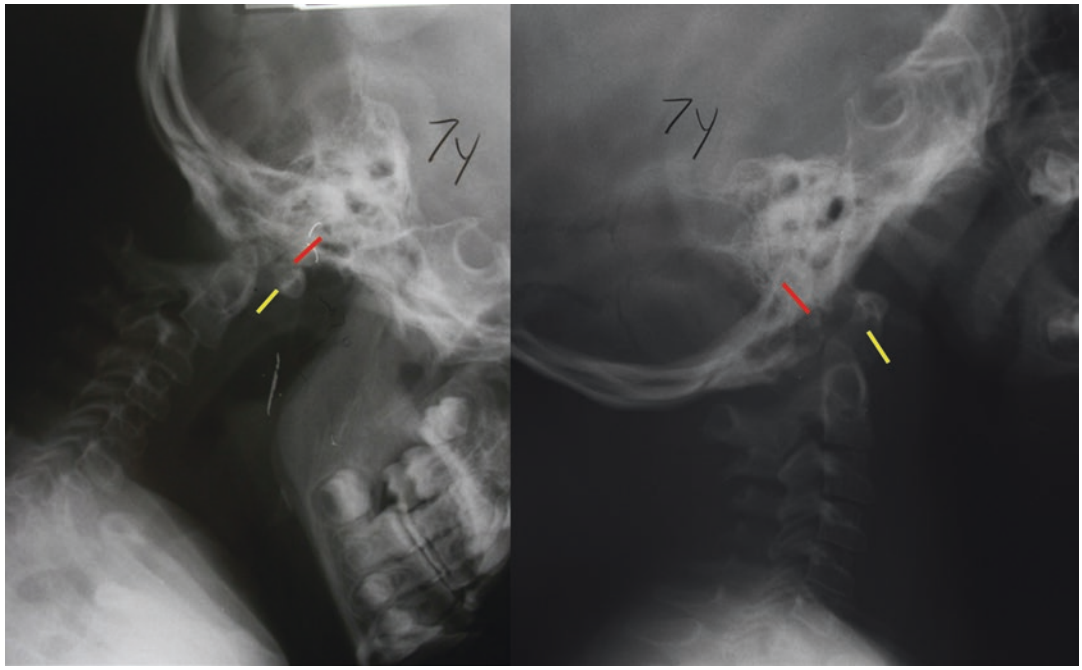

Fig. 10.3 Occipital-C1 instability in a 7-year-old patient on flexion/extension radiographs. On extension, the skull moves posterior (red line), relative to the posterior edge of the C1 arch (yellow line)

may be associated with spina bifida occulta and posterior arch defects, an important preoperative consideration [27, 28].

Cervical spine instability may not cause symptomatic spinal cord compression. In a normal spinal canal at the C1 level, Steel's rule of third dictates that 1/3 of the space is occupied by dens and 1/3 by spinal cord, and 1/3 is a reserve space that may be sufficient to buffer the instability created by pathology. In Down syndrome, for instance, symptomatic atlantoaxial instability is only present in 18% of the patients with atlantoaxial instability [29]. 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. However, our practice is to not perform fusion for asymptomatic instability, nor do we recommend prophylactic fusion in populations prone to instability such as Morquio syndrome (Fig. 10.7). The most important factor is spinal cord compression, and instability without spinal cord compression in a capacious canal does not require surgical management.

Physicians 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 [26] or metatropic dysplasia [30]. In these cases, laminectomy of C1 may be necessary to decompress the spinal cord. The earlier noted findings of extradural soft tissue thickening contributing to stenosis in Morquio syndrome has been shown to dissipate after fusion [19].

Radiological Signs of Upper Cervical Spine Instability

The occipitocervical junction is difficult to visualize. Plain radiography can be unclear in young children with skeletal dysplasia due to motion, delayed ossification, odontoid abnormalities, abnormal anatomy, and failure to cooperate for flexion/extension films. MRI is ideal for assessing

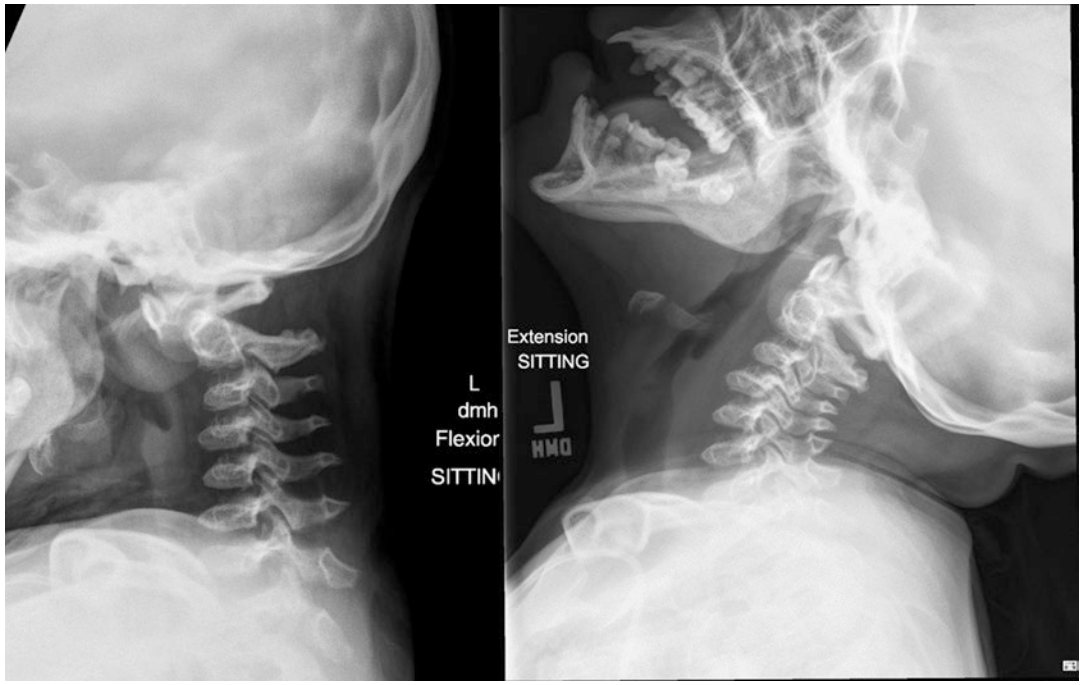

Fig. 10.4 Atlantoaxial (C1–2) cervical instability. Flexion-extension radiographs of a patient with Morquio syndrome showing large anterior subluxation of C1 on C2 in flexion, which reduces in extension from an underdeveloped dens and ligamentous laxity. Here you can see the space available for the cord (SAC) greatly decrease

the spinal cord and should include flexion/extension positioning [31]. In very young children, this entails general anesthesia. MRI is done in neutral position initially. Compression seen at this position may obviate the need for further dynamic positioning. Common measurements are as follows:

- The standard measurement for O/C1 horizontal instability includes Power's ratio,

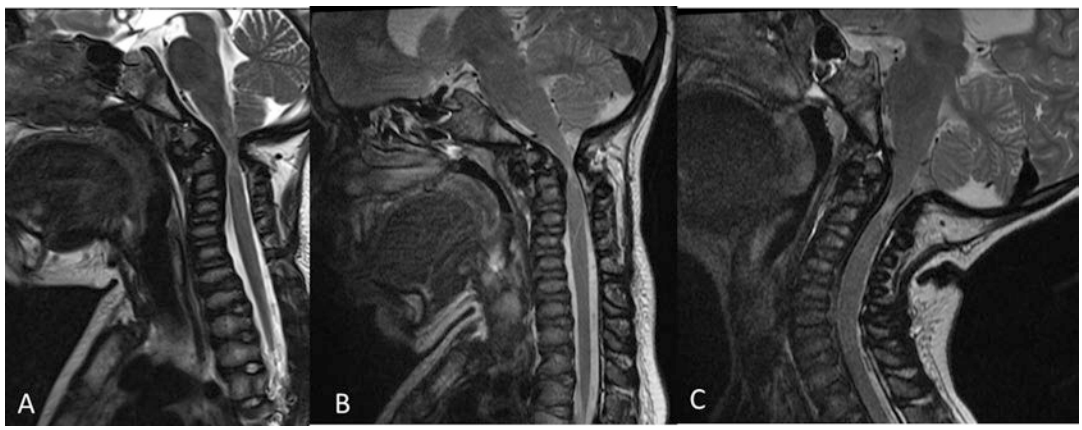

Fig. 10.5 (**a–c**) C1-C2 instability causing stenosis and myelomalacia in an MPS patient (Hurler). MRI neutral (**a**), flexion (**b**), and extension (**c**) views. Note the extradural soft tissue thickening from mucopolysaccharide deposition causing the stenosis

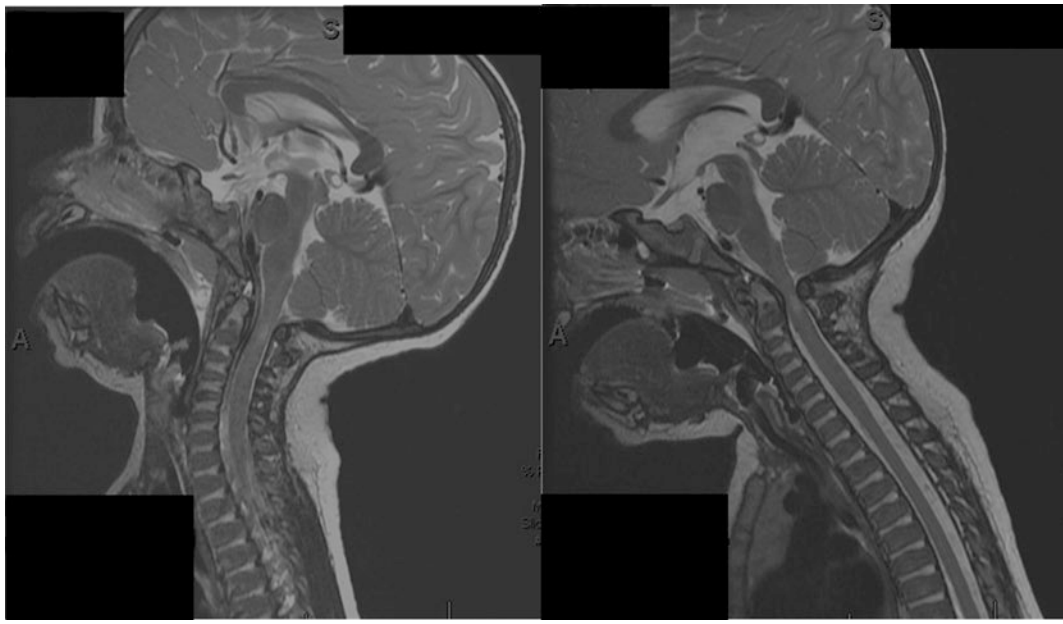

Fig. 10.6 Flexion and extension MRI of a 16-month-old child with spondyloepiphyseal dysplasia tarda. In contrast to the MPS example in Fig. 10.5, dynamic stenosis and myelomalacia of the spinal cord is from pure instability, without extradural tissue thickening seen

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 Wackenheim'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, or posterior atlantoaxial distance (also known as space available for the cord, SAC).

Surgical Options

Cervical arthrodesis at C1-C2 include Gallie's technique, Brooks-Jenkins' technique, Sontag modification of Gallie's technique, Magerl's transarticular screw [32], and Harm's modification of Goel's technique [33]. 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 preop evaluation of the vascular anatomy needs to be performed to avoid iatrogenic injury to the vertebral artery.

The Brooks-Jenkins' technique [34] and the Sontag modification of Gallie's technique [35] 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 dural impingement in a reduced position or if it is secondary to an irreducible subluxation. Fixation is ideally achieved with the Magerl's or Harm's 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 occipitocervical fusion after decompression, we have used a modified occipital-based technique with a fashioned autologous iliac bone graft notched between O and C2 using cable fixation. Immobilization is performed using halo-vest postoperatively. Sitoula and Mackenzie have reported this occipital wiring technique with fashioned iliac crest bone graft

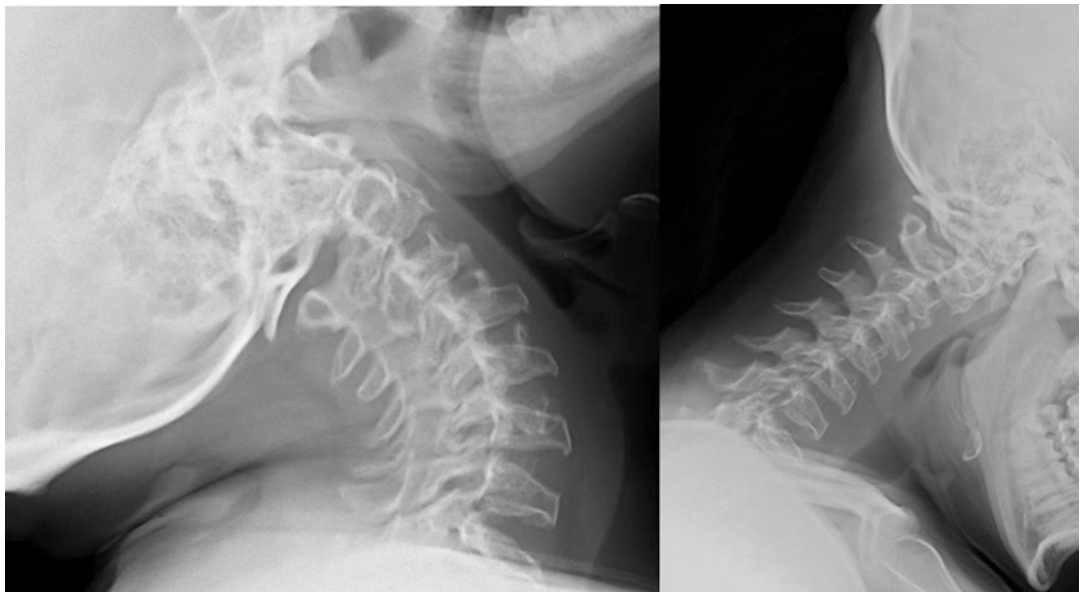

Fig. 10.7 22-year-old Morquio syndrome patient without instability, who therefore would not have benefitted from a prophylactic C1-C2 fusion at a young age. Note the occipitalization of C1

that resulted in no nonunions [36]. Other techniques for O to C2 fusion include the use of rib graft and wire-based fixation of a fashioned iliac crest bone graft. Occipitocervical fixation is often too bulky in young children but does provide very rigid fixation in older children.

Torticollis: Atlantoaxial Rotatory Fixation/Subluxation/Dislocation

In patients with metatropic dysplasia, torticollis may occur as part of a possible mechanism to protect the airway [30]. Patients present with torticollis in extension. This can also develop postoperatively in dysplasia 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 [37].

Subaxial Cervical Spine Abnormalities

Abnormal Extradural Impingement

Extradural impingement of the spinal cord may be observed in mucopolysaccharidoses (Fig. 10.5). This is almost universal in mucopolysaccharidoses type IVA (Morquio syndrome) [3, 19] and to a lesser extend in type IH (Hurler syndrome) and type VI (Maroteaux-Lamy syndrome) [4]. The glycosaminoglycan accumulation intracellularly results in mechanically incompetent cartilaginous and ligamentous tissue that encourages reactive tissue formation. In cases where biopsy was performed via trans-oral approach, reactive fibrocartilaginous tissue accumulated extradurally without evidence of meningeal involvement [19]. It was also noted that the reactive tissue could extend caudadly to the dorsal aspect of the centrum of axis (C2) and distally to C3 and C4.

Kyphosis and Stenosis

Subaxial cervical spine kyphosis should be screened for in patients with diastrophic dysplasia and Larsen syndrome [38]. Other skeletal dysplasias with cervical spine kyphosis include Kniest dysplasia, chondrodysplasia punctata (Fig. 10.8), and campomelic dysplasia. Rarely, Morquio syndrome may present with cervical kyphosis which is associated with its distinct severe platyspondyly [30, 39]; however, stenosis

can be present without such kyphosis, as above with extradural impingement.

With 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. Further 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 elements deficiency, and the loss of the posterior tension band.

In diastrophic dysplasia, a quarter of the patients have cervical kyphosis at birth. The kyphosis has its apex at C3 to C4, less commonly at C5. With the preservation of lordosis at the caudad level, an 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 corners" [40]. Concurrent spinal bifida occulta may be present from C3 to upper thoracic spine [41]. Most of the cases were resolved spontaneously (Fig. 10.9), and if this is the case, they will do so by the age of 5 [42]. In the largest series to date, cervical kyphosis of more than 60 at presentation had a poor prognosis with either progression of the kyphosis or respiratory failure from associated severe tracheal and bronchomalacia in early life [40].

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. The findings include a hypoplastic anterior column at the apical vertebra, segmentation defects, and spinal bifida occulta (Fig. 10.10). In the most severe cases, the associated ligament laxity, hypotonia, and posterior column deficiency result in spondylolisthesis and spondyloptosis [43]. 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 in situ posterior arthrodesis without instrumentation in milder deformity without cord compression. Halo immobilization is essential [44]. 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 have been described [45]. In young children, the posterior elements are not sufficient for rigid instrumentation.

Cervicothoracic Abnormalities

Cervicothoracic stenosis has been reported in Morquio syndrome and chondrodysplasia punctata [46, 47]. The stenosis is usually associated with junctional kyphosis; however, stenosis has also been reported without associated kyphosis due to dysplastic vertebrae. With 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 commonly affected level is C7-T1, but it can extend to T4 [47]. A whole spine MRI is necessary to screen for pathology. The age of onset can start at 2 years. The majority of the patients with Morquio syndrome are diagnosed from 2 to 5 years old [48]. The most common presenting symptoms include an unsteady gait or failure to walk. Upper motor neuron signs are often seen. Treatment to decompress the kyphosis is recommended once symptoms are present.

Summary

Because of the altered cervical anatomy and development in skeletal dysplasias, the pathology and best manner for treatment are particular to each syndrome. By understanding the diagnoses, a practitioner will not miss screening for what are sometimes life-threatening cervical pathologies. Once found, treatment can be individualized to the patient and his or her anatomy.

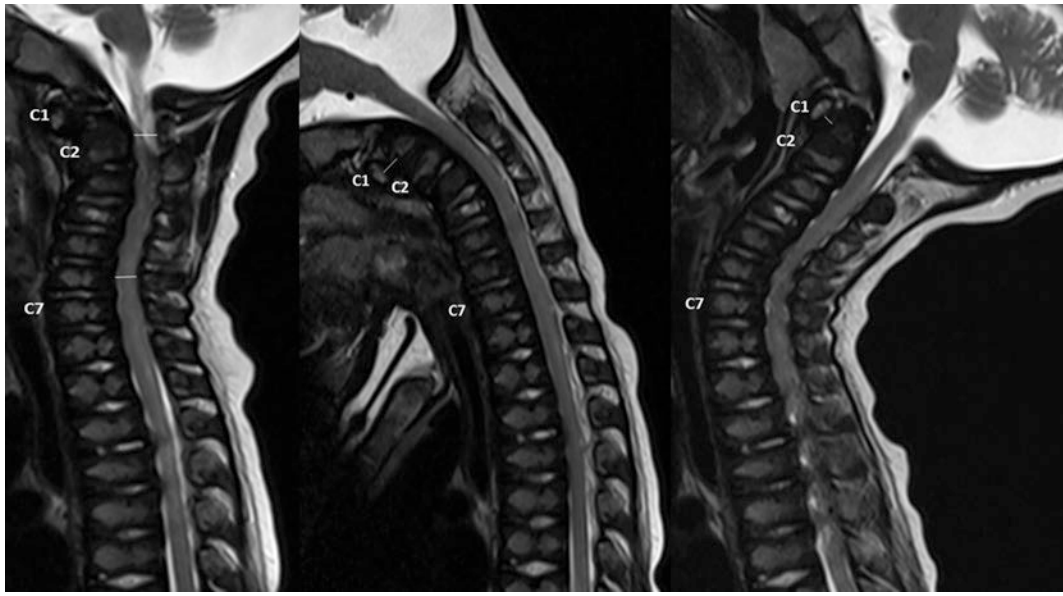

Fig. 10.8 Diffuse subaxial cervical stenosis in a 20-month-old with chondrodysplasia punctate. The stenosis, which is present in neutral and extension positioning, is somewhat relieved in flexion

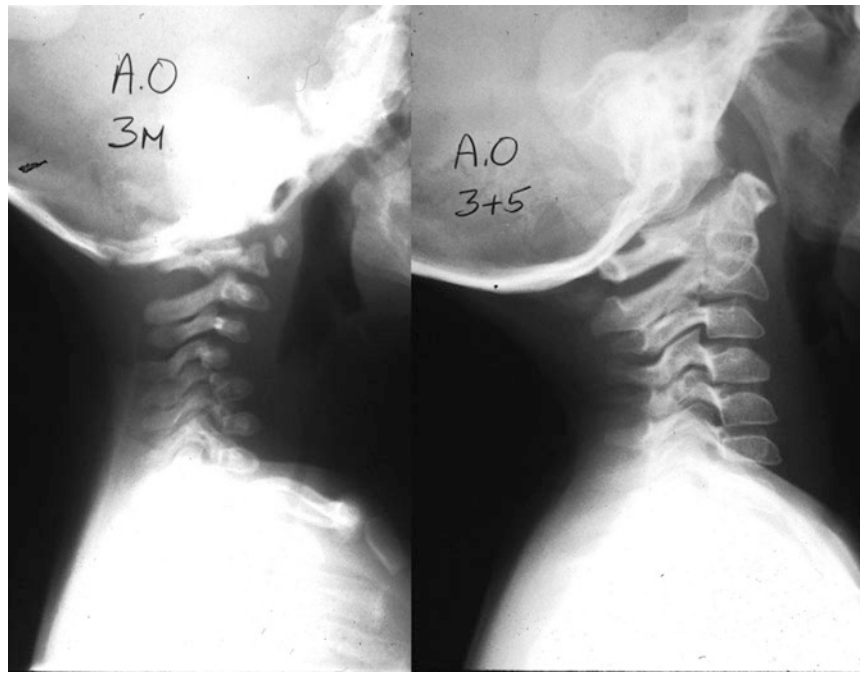

Fig. 10.9 Resolution of kyphosis with age in a patient with diastrophic dysplasia. The radiograph on the left shows characteristic C5 kyphotic apex at 3 months of age. The radiograph on the right is of the same patient at 3 years and 5 months with resolution

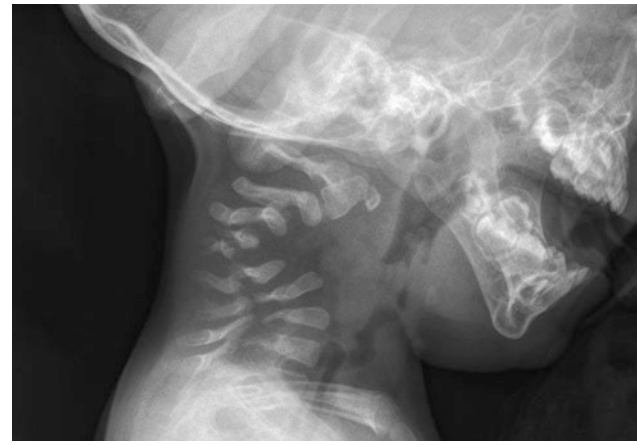

Fig. 10.10 Patient with Larsen's syndrome showing kyphosis with poorly developed anterior spinal column

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Congenital Muscular Torticollis

11

Kaela Frizzell, Archana Malik, Martin J. Herman, and Peter Pizzutillo

Introduction

Derived from the Latin terms "tortus" (twisted) and "collum" (neck), torticollis is the term utilized to describe the clinical finding of a lateral head tilt with rotation of the chin to the opposite side of the tilt. Torticollis is a nonspecific term applied to this entity. It is important to note that torticollis is a clinical symptom or sign of pathology, but is not a specific diagnosis [1]. It can be classified as congenital, which presents in the first few months of life, or acquired, which appears later in the child's development. The most common type of congenital torticollis is known as congenital muscular torticollis. Approximately 82% of children of all ages presenting to a tertiary care pediatric orthopedic facility were found to have congenital muscular torticollis as the cause of their torticollis.

Etiology and Epidemiology

Congenital muscular torticollis occurs in approximately 1–2% of children [2]. It has a slight male predominance with a 3:2 incidence and tends to affect the right sternocleidomastoid muscle more than the left [3]. The exact cause of the disease is unknown; however, several theories of etiology exist. It has been proposed that intrauterine positioning and perinatal events can lead to contractures and fibrosis of the sternocleidomastoid muscle. Intrauterine overcrowding has been suggested as a root cause, since congenital muscular torticollis is more likely in infants who were firstborn or one of multiple gestation, as well as those who were involved with complications of pregnancies such as oligohydramnios or breech presentation. Further, infants who experienced a difficult delivery may also have developed congenital muscular torticollis as a result of compartment syndrome of the sternocleidomastoid muscle from prolonged time in the birth canal. In this case, local compression during birth may have compromised venous outflow of the sternocleidomastoid muscle, resulting in ischemia, hemorrhage, and subsequent fibrosis [4–6]. In Cheng's review of infantile torticollis, 62% were reported to have had a difficult birth history [3].

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Clinical Features and Associations

The most common presentation of a child with congenital muscular torticollis is the head tilt. Again, the head will assume a lateral tilt toward the side of the affected sternocleidomastoid muscle, with a rotated chin away from the head tilt due to the contracture of the muscle. Although rare, bilateral involvement may occur and makes the diagnosis particularly challenging. Torticollis typically develops by 2–4 weeks of age. After that time, the infant prefers to hold their head tilted to one side, and plagiocephaly and facial asymmetry may exist. A pseudotumor comprised of fibrous tissue may be present in the muscle and persists for approximately 2–3 months. This fibrous mass usually abates by 4–6 months of age (Fig. 11.1a, b).

Congenital muscular torticollis is associated with other pathologies such as developmental dysplasia of the hip and metatarsus adductus of the feet. They may also have talipes equinovarus and C1–C2 subluxation. These findings help to corroborate the theory of torticollis as a "packaging" disorder. The hips of all infants presenting with torticollis must be evaluated carefully to rule out hip dysplasia. In their study, Walsh and Morrissy found the rate of hip disease in those infants with torticollis to be approximately 8% [7]. More recent literature has revisited the association of congenital muscular torticollis and hip dysplasia. In a retrospective review by von Heideken et al., it was noted that in infants with a primary diagnosis of congenital muscular torticollis, 3.7% also had developmental dysplasia of the hip. Further, those infants that were less than

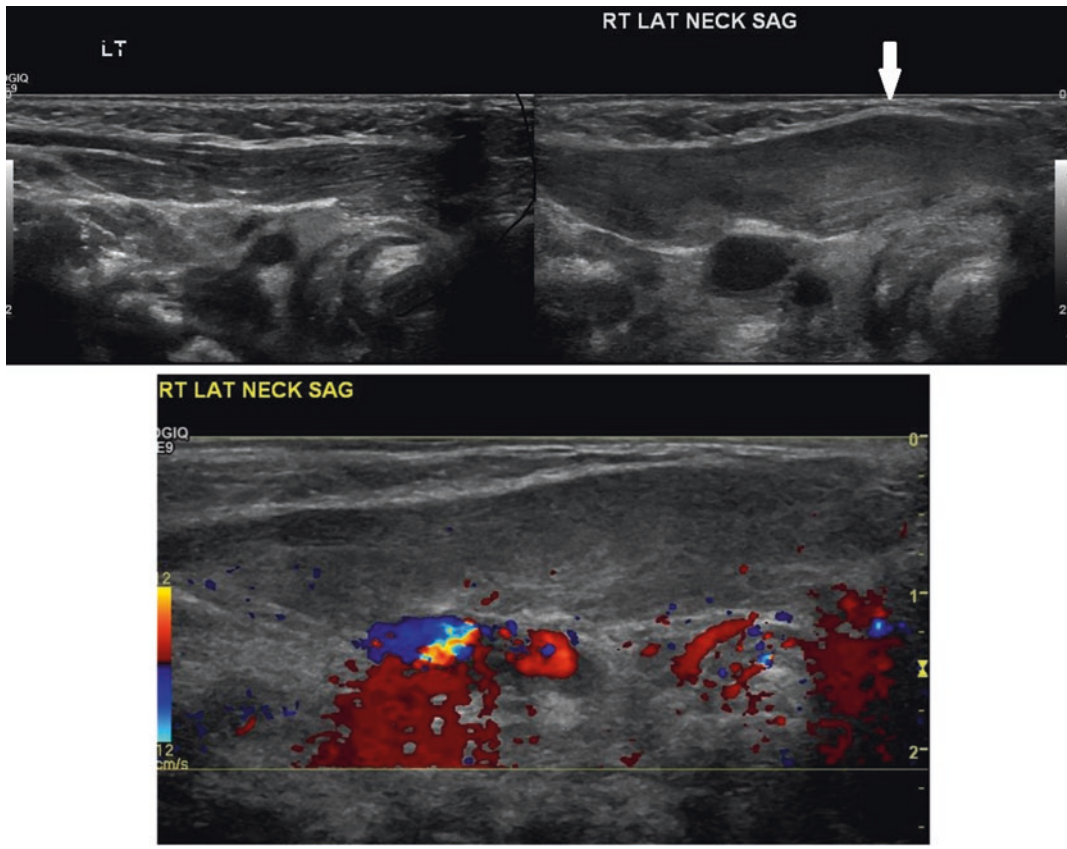

Fig. 11.1 (**a, b**) 1-month-old male with right-sided torticollis. (**a**) Dual longitudinal US images of the left and right neck demonstrate focal mass (arrow) in the lower portion of the right sternocleidomastoid (SCM) muscle, which is mildly echogenic. Note the normal left SCM for comparison. (**b**) Longitudinal US image of the right neck with color Doppler does not depict increased internal flow within the enlarged right SCM. These findings are compatible with congenital muscular torticollis, also known as fibromatosis colli

1 month of age and diagnosed with developmental dysplasia of the hip had a 9% risk of developing congenital muscular torticollis. Therefore, they strongly recommended screening infants with torticollis for hip dysplasia as well [7–9].

Because torticollis causes the child to maintain a certain head position with the face turned to one side, skull deformation and facial asymmetry may develop. In one study, all infants who presented with torticollis had some degree of craniofacial asymmetry with reduction of the vertical facial height on the affected side [10]. The treatment of the torticollis usually improves the facial asymmetry in most cases, and early treatment is associated with a more favorable prognosis. In severely affected children with delayed presentation, surgical correction was needed for the skull and facial deformities.

Studies regarding the prevalence of developmental delay in children with torticollis demonstrate conflicting results. In one study, Ohman et al. concluded that children with congenital muscular torticollis are not at a higher risk for delay in motor development at preschool age. Conversely, Schertz et al. have described a higher risk of neurodevelopmental delays associated with torticollis and found that these conditions presented at varying stages of development [11, 12].

History and Physical Examination

In congenital torticollis, it is essential to not only obtain a pertinent history of present illness, but a detailed birth history, as well. This should include birth weight and complications during pregnancy and delivery, the age at which the torticollis was first noted, and a history of feeding difficulties or other signs of gastroesophageal reflux. Along with a detailed history, the child who presents with congenital torticollis must be evaluated with a documented physical examination for abnormalities of the sternocleidomastoid muscles, shoulder and neck birth trauma, and vision, hearing, or neurologic deficits. The skull should also be evaluated for asymmetric flattening, as well as the face for asymmetry. This can help confirm the diagnosis of congenital muscular torticollis, which is the most common cause of these findings [13].

In congenital muscular torticollis, a well-circumscribed, firm mass may be palpated in the inferior one-third of the affected sternocleidomastoid muscle. The mass reaches maximum size in the first few months of life and then gradually abates. Generally, the mass is not present by 6 months of age. Again, patients with congenital muscular torticollis will present with the top of their head tilted toward the affected sternocleidomastoid muscle, with the chin rotated toward the unaffected side. They have limited lateral bending away from the abnormal sternocleidomastoid muscle. Further, they have limited rotation toward the same side as the abnormal muscle. The occiput tends to be flat on the side of the skull that is opposite to the abnormal sternocleidomastoid muscle [14].

Diagnosis and Imaging

The diagnosis of congenital muscular torticollis is usually made clinically. Further work-up of the neck component is usually not needed in an infant who developed the torticollis within the first 2–4 weeks of life. This is especially the case if there is a palpable mass present in the inferior portion of the sternocleidomastoid muscle. However, if there is a need to differential congenital muscular torticollis from other pathologies in the neck, such as if there is a delay in presentation or a difference in location or firmness of the mass, then ultrasound is the modality of choice. Ultrasound will assist in confirming the location of the mass and may also help to corroborate improvement in the torticollis, as it can confirm disappearance of the pseudotumor within the muscle when repeated after several months. It is generally regarded as a safe procedure as it does not require the infant to be sedated [15, 16]. Plain radiographs are usually unnecessary in infants who display characteristic features of congenital muscular torticollis and typically are low yield, since so much of the child's spine at that age is cartilaginous [17].

However, in certain atypical cases, radiographs may be necessary to evaluate potential bony anomalies. An anterior-posterior and lateral X-ray of the entire cervical spine is a quick way to determine bony abnormalities in the patients who may present with such findings as craniofacial asymmetry, a short neck or low hairline, elevation of the scapulae, or asymmetry of the size and position of the scapulae or those who do not have characteristic tightness of the SCM muscle. Imaging may also be necessary to evaluate those patients who do not demonstrate improvement in neck range of motion despite consistent physical therapy for a month or two. Advanced diagnostic imaging of the cervical spine, including CT and MRI, is indicated if congenital anomalies of the spinal column, spinal cord, or brain are suspected [18].

Ultrasound of the bilateral hips is recommended for any child with congenital muscular torticollis and for any infant with torticollis of any etiology and clinical signs of hip dysplasia. Some surgeons prefer to routinely perform hip ultrasounds on all infants with a diagnosis of congenital torticollis because of the strong association between this diagnosis and acetabular dysplasia without clinical signs of hip instability. A patient with congenital muscular torticollis should be evaluated for developmental dysplasia of the hip via ultrasound if under 4–6 months of age and with radiographs of the pelvis if older than 6 months of age.

Differential Diagnosis

Not all cases of congenital torticollis are caused by pathology within the sternocleidomastoid muscle; therefore, careful evaluation and consideration of the whole patient is expected, and sometimes a multidisciplinary approach is required [19].

Congenital torticollis may result from other neurologic, muscular, or bony abnormalities of the cervical spine (Fig. 11.2a–d). Again, these entities should be considered in patients who present atypically or in those who did not have a history of a difficult birth. Alternative abnormalities that can cause congenital torticollis are various skeletal abnormalities such as craniosynostosis, Klippel-Feil syndrome, and pterygium colli. The patient may present with neurologic issues such as syringomyelia (Fig. 11.3a, b), brachial plexus palsy, or ophthalmologic disorders driving the torticollis. Other entities causing torticollis may be acquired.

Treatment

Congenital muscular torticollis may resolve spontaneously; however, if left untreated and it does not resolve, it can leave the child with significant loss of motion and craniofacial asymmetry. Treatment of the child with congenital muscular torticollis includes positioning and handling of the infant for midline orientation, with passive stretching for symmetric range of motion of the neck. The mainstay of treatment for congenital muscular torticollis is centered on physical therapy and a home stretching program for the infant. With proper physical therapy and stretching, the majority of patients with congenital muscular torticollis resolve by 1 year of age [2, 20]. Early recognition of the condition and prompt initiation of the physical therapy and stretching program play a large role in the prognosis of the patient. Some studies report that if a physician-guided home exercise program for congenital muscular torticollis is started prior to 4 months of age, the average length of treatment is approximately 3 months [2]. Also, it is noted that some children who are referred for physical therapy after 1 month of age required significantly longer treatment.

Lee et al. have demonstrated that initial cervical range of motion is an important factor in predicting the rehabilitation outcome in patients with congenital muscular torticollis [10]. Other studies report that 95% of children with less than ten degrees of deficit in passive rotation on presentation have successful outcomes with a home stretching program. If no improvement was noted after 4 weeks of home stretching, the patients were switched to formal physical therapy [2].

Monitoring of the patient with congenital muscular torticollis should occur every month.

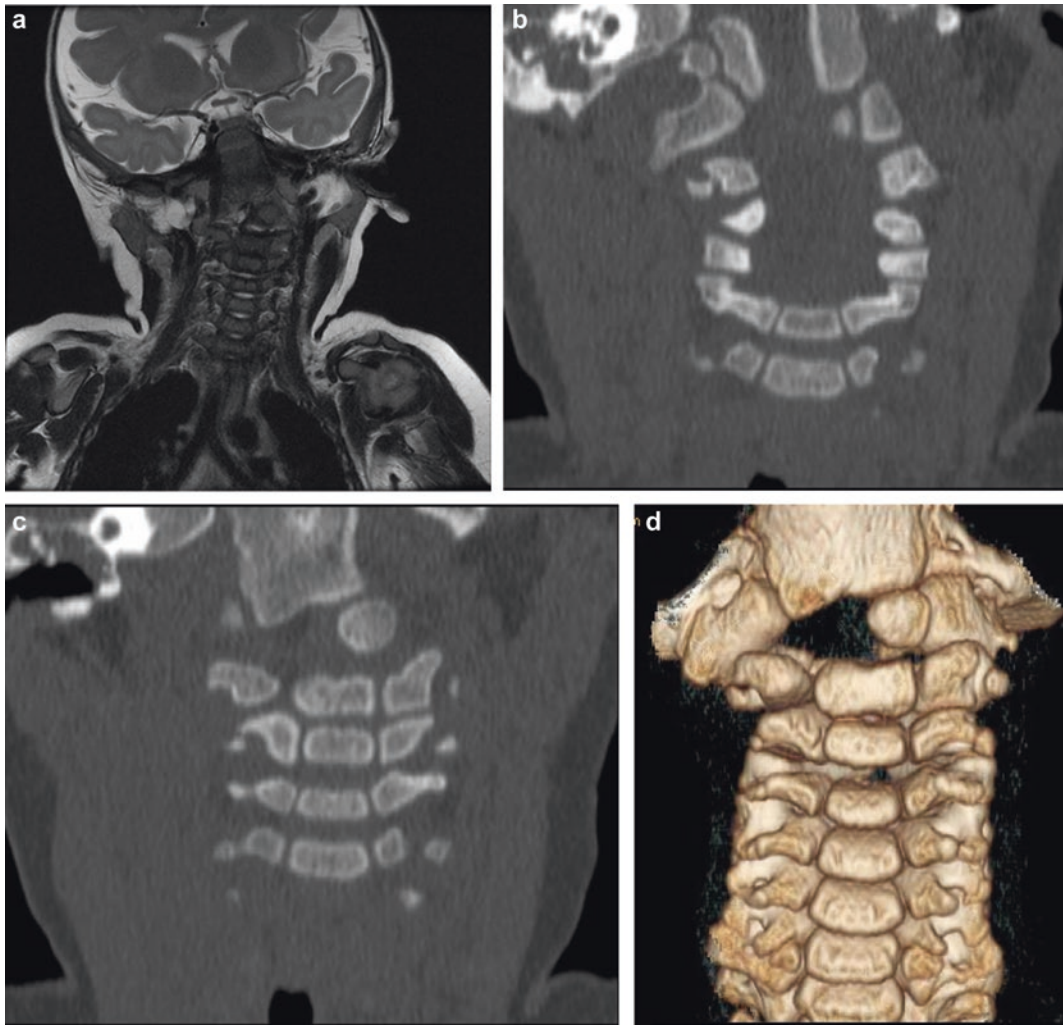

Fig. 11.2 (**a**–**d**) 6-month-old male presented with left-sided torticollis. (**a**) Coronal T2 FRFSE image from MRI of the cervical spine shows leftward torticollis with suspected osseous anomalies at the craniocervical junction and upper cervical spine. (**b**–**d**) Subsequently obtained CT cervical spine coronal bone reformats and volume-rendered 3D image demonstrate left-sided atlantooccipital fusion, dysmorphic left C1 hemivertebra, and dysmorphic C2 with absence of the odontoid process

This allows the clinician to determine improvement of the patient's torticollis and compliance with home stretching or formal physical therapy. Further, it may highlight the development of any neurologic or other abnormalities that could represent an alternative cause to the patient's diagnosis. Patients with congenital muscular torticollis refractory to consistent stretching and physical therapy may benefit from botulinum toxin A injection to the affected sternocleidomastoid muscle. While some studies state that good range of motion can be achieved with this method, obviating the need for surgical intervention, botulinum toxin should be utilized with extreme caution due to the small anatomic structures about the neck [21]. If attempted, the use of ultrasound guidance is suggested. The use of orthoses, cervical collars, and helmets for patients who do not benefit from physical therapy is controversial. The primary contraindication for stretching exercises is bony abnormalities, which may be ruled out with cervical radiographs.

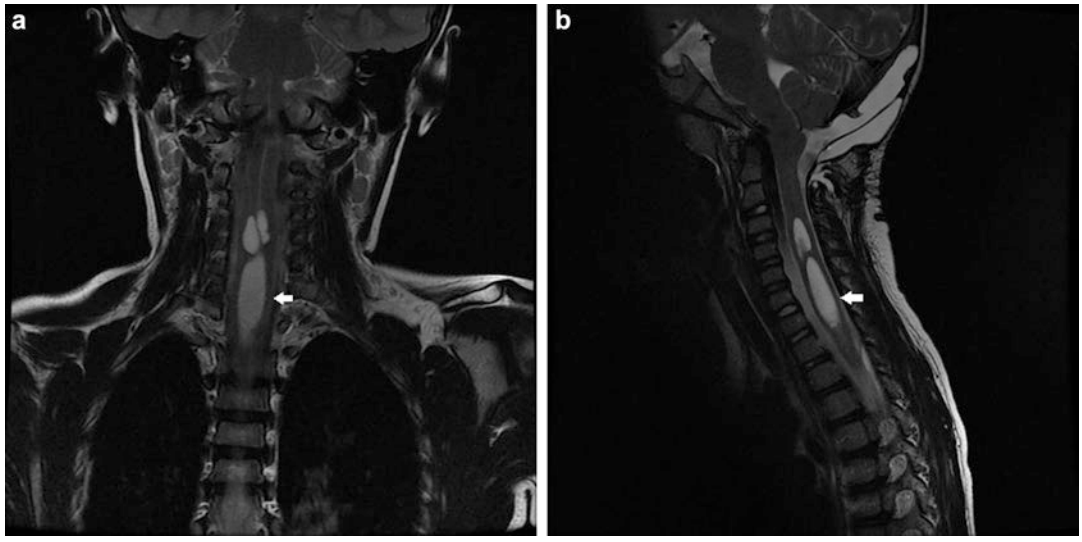

Fig. 11.3 (**a**, **b**) 6-year-old female presenting with torticollis. Patient has history of Chiari I malformation status post suboccipital craniectomy and syrinx. (**a**) Coronal T2 FRFSE sequences demonstrate expansile, mildly septated, intramedullary T2 hyperintense signal abnormality within the cervical and upper thoracic spinal cord compatible with syrinx (arrow). (**b**) Sagittal T2 FRFSE sequences demonstrate the same signal abnormality (arrow) in this patient with history of Chiari I malformation status post decompression surgery

If nonoperative treatment for congenital muscular torticollis fails, surgical intervention may be indicated due to the potential for residual plagiocephaly, facial asymmetry, and restricted cervical motion. The most common surgical intervention for children younger than 3 years of age is a unipolar release of the sternal or clavicular head of the SCM muscle, with intensive postoperative physical therapy (Fig. 11.4a–c). For older children or those with more unrelenting disease, a bipolar release, z-lengthening of the muscle, or, rarely, radical resection of the sternocleidomastoid muscle may be utilized. These patients generally have good results with improvement of neck motion and head position, but relapse is common in those whose postoperative protocol is inadequate.

The remodeling potential of the facial and skull asymmetry is variable and dependent upon the patient's age at diagnosis and degree of deformity, as well as success of treatment. The role of surgery in improving facial and skull deformity is controversial. Studies have shown that surgical release of the sternocleidomastoid muscle before the age of 5 years has been shown to have greater success. These patients also appear to have more cervical motion, less scarring, and less head tilt if they had undergone surgery prior to age five. There is data to suggest that improvements can be made with surgery in patients who are older than 8 years of age. In one study, Shim et al. demonstrated that satisfactory cosmetic and functional results could still be achieved with surgical treatment of congenital muscular torticollis in these older children [22].

Other Entities of Congenital Torticollis

Any patient who presents with signs or symptoms of congenital torticollis that are inconsistent with the congenital muscular variety, or those who do not demonstrate improvement with stretching exercises, should be reevaluated and another underlying condition considered. A more in-depth work-up and referral to other specialties may be warranted. Congenital torticollis

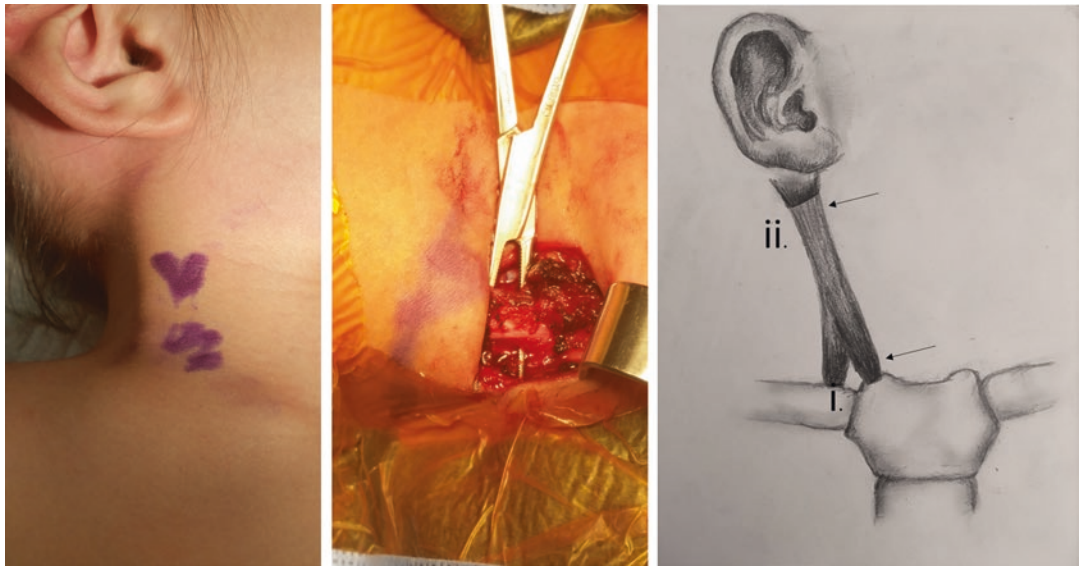

Fig. 11.4 (**a**) A 6-year-old girl was diagnosed with right congenital muscular torticollis and, after 9 months of formal stretching program with a physical therapist, had a significant head tilt and limited range of motion. Intraoperatively, the child is supine with a small bump between the scapulae, and the sterile field includes the sternal notch and the full length of the sternocleidomastoid muscle, SCM. The incision is made superior to the clavicle at the insertion the SCM for a unipolar release. (**b**) After incision of the skin and platysma, the distal part of the SCM is isolated and transected. The tighter of the two heads of the SCM is released first; if motion and head position are restored, the other head is left uncut. If the SCM remains tight, the other head is released. (**c**) A schematic drawing of the SCM. For a unipolar release, only the insertions of the SCM are released (arrow i). For bipolar release, indicated in older children, a second incision is made along the SCM proximally and the SCM is divided at this level in addition to the distal release (arrow ii)

with abnormalities of the basic eye exam should be referred to the ophthalmologist, as the torticollis may be resulting from an ophthalmologic issue such as strabismus. Children with torticollis and an abnormal neurologic exam should be referred to neurology for evaluation of benign paroxysmal torticollis. Further examination by a neurosurgeon is required if the skull asymmetry is atypical, extreme, or if there is perhaps a suggestion of a structural abnormality of the brain or spinal cord. A gastroenterology referral may be indicated for children with persistent episodes of torticollis from suspected gastroesophageal reflux that is not improving with medical treatment. A genetics consult is warranted for patients in whom torticollis appears as part of a syndrome. Congenital torticollis secondary to abnormalities of the spine may also have associated congenital abnormalities of the cardiac and genitourinary systems.

Summary

Congenital muscular torticollis remains the most common form of congenital torticollis. It has the characteristic features of early presentation in the first few weeks of life, with the infant posturing the head tilted to the side of the affected muscle, with the chin rotated to the opposite side. There may be a palpable mass in the lower portion of the affected sternocleidomastoid muscle. The diagnosis is usually made clinically, but can be corroborated with ultrasound and advanced imaging, as necessitated in atypical cases. The majority of cases of congenital muscular torticollis will resolve after several months of conservative management, with manual stretching and physical therapy as needed. Surgical correction may be required in patients with residual deficits in range of motion of the neck, plagiocephaly or facial asymmetry, and late presentation.

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Cervical Spine Injuries in Athletes

12

Firoz Miyanji

Epidemiology

Approximately 3.5 million children suffer sports- or recreational-related injuries annually. Athletic activities are one of the leading causes of cervical spine injuries in the pediatric population and account for the second most common cause of spinal cord injury in people less than age 30 [1]. In children aged 10–14, sports are the most common cause of cervical spine injuries [2, 3]. Although football and ice hockey largely account for the majority of these injuries, other sports offering significant risk include rugby, wrestling, diving, skiing, snowboarding, cheerleading, and equestrian sports. The mechanics of cervical spine injury in athletes has been extensively studied; however, the true mechanism of injury remains controversial.

Approximately 1.2–1.5 million athletes play high school football annually. While football has a lower rate of catastrophic cervical spine injuries than hockey or gymnastics, the larger number of participants has resulted in football being associated with the highest number of severe cervical spine injuries per year for any high school or collegiate sport [4, 5]. The pattern of cervical spine injury in football has evolved over time. With the advent of protective helmets, head-related fatalities declined in the early 1970s; however, the number of cervical quadriplegia cases increased. This was a direct result of "spearing" which was a technique that used the top of the helmet as the initial point of contact for blocking or tackling, a style of play that placed the cervical spine at increased risk of injury, as demonstrated by Torg and colleagues [6]. Between 1959 and1963, Schneider documented 56 cases (1.36 per 100,000) of cervical spine injuries, of which 30 (0.73 per 100,000) had permanent quadriplegia [7]. From 1971 to 1975, the National Football Head and Neck Injury Registry compiled 259 cervical fractures/dislocations (4.14 per 100,000) and 99 cases of quadriplegia (1.58 per 100,000) [8]. In 1976, intentional spearing was banned, and subsequently from 1976 to 1987, the rate of cervical injuries decreased 70%, from 7.72 per 100,000 to 2.31 per 100,000 at the high school level. At the time of the rule change in 1976, the annual rate of permanent quadriplegia was 2.24 per 100,000 participants in high school football. By 1984, the rate of this neurologic injury had decreased to 0.38 per 100,000 [9].

Current data indicates a relatively stable incidence of traumatic quadriplegia in football players. In 2002, the incidence of this injury was 0.33 per 100,000 in high school football [9]. Cantu and Mueller's [10] review of catastrophic cervical spine injuries in American football from 1977 to 2001 found 223 football players sustained a

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severe cervical spine injury with no or incomplete recovery. One hundred and eighty-three of these injuries occurred in high school athletes, giving an incidence rate of 0.52 per 100,000 high school participants over the past 25 years. In a more recent review of 500,000 high school football injuries during the 2005–2006 season, 4.1% of all injuries involved the cervical spine, further defining this at-risk population [11].

When compared with other sports, the overall number of catastrophic injuries in high school and college ice hockey players is lower, but the incidence is comparatively higher. A longitudinal Canadian survey by Tator et al. [12] uncovered 241 spine fractures and dislocations between 1966 and 1993, 90% of which involved the cervical spine. Permanent spinal cord injury (SCI) occurred in 108 of the 207 athletes in the registry (52.2%), and 52 (25.1%) had complete lesions. Eight players died as a result of complications from their SCI. The incidence of major spinal injuries in ice hockey began increasing in the early 1980s. From 1982 to 1993, there was an average increase in 16.8% cases per year, peaking in 1995 with 26 injuries, as reported by the Canadian ThinkFirst-SportSmart Sports and Recreational Injuries Research and Prevention Center registry [13]. Although football is associated with a higher number of catastrophic cervical injuries each year, the annual incidence of SCI is three times higher in hockey than football. The annual rate of catastrophic cervical spine injuries in the USA at the high school level is 2.56 per 100,000 in ice hockey, compared to 0.68 per 100,000 in football [3, 9].

The vast majority of cervical fractures and major ligament injuries in wrestlers occur at about a rate of 1 per 100,000 annually at the high school and college levels [14]. Most injuries occur in match competitions in the low- and middleweight classes. The most common wrestling maneuver associated with cervical spine injuries in wrestlers is a takedown of a standing opponent in a defensive position.

Race diving into the shallow end of a pool is the most common cause of swimming-associated catastrophic spinal injuries [15]. This is a direct result of diving headfirst into a shallow end of a pool, causing a cervical axial compression injury. There have been rules now implemented to help prevent such injuries during the racing dive both at the high school and college levels. Many recreational diving injuries, however, go unreported, limiting attempts at awareness and water safety. Schmitt and Gerner's retrospective review of traumatic spinal cord injuries presenting to their center noted diving accidents were the cause in 7.7% of cases, with 97% being males [16]. Others have noted inadequate supervision, alcohol use, shallow water, and inexperienced divers to be risk factors for injury [17].

The incidence of severe spinal cord injury is 0.01 injuries per 1000 skier days [18]. These are evenly distributed at all spinal levels, without a preponderance to the cervical spine. The injuries have been increasing over the last two decades, especially in young males. Although spinal injuries are not the etiology of all skiing fatalities, they have been well documented as a significant cause of death in skiers. Spinal injuries in snowboarders have been reported to be three- to fourfold higher than in skiing, and this ratio seems to be growing [19, 20]. Up to 80% of snowboarding-related injuries are due to jumping accidents [20]. The other population of snowboarders is the inexperienced riders sustaining falls [19–21].

Approximately 10% of serious injuries in rugby involve the cervical spine, with cord contusions constituting 25% of these injuries [22, 23]. Cervical spine injuries most frequently occur during a scrum, with the hooker suffering the most injuries. During the scrum, each side may generate weights up to 1.5 tons, and the hooker may encounter almost 50% of this weight [24, 25].

Developmental Anatomy (see also Chapter 1)

By the fourth week of gestation, there are 42–44 pairs of somites (four occipital, eight cervical, twelve thoracic, five lumbar, five sacral, and eight to ten sacrococcygeal), each of which differenti-

ates into a dermatome, myotome, and medial sclerotome. After the division of each sclerotome, the individual vertebral bodies will be formed by the union of the superior half of one sclerotome to the lower half of its neighbor.

The occiput, atlas, and axis are formed by different mechanisms. The first spinal sclerotome gives rise to the atlas. The atlas is formed from three ossification centers: the anterior arch and two lateral masses. The anterior arch may not be ossified at birth but becomes visible as one or two ossification centers during the first year of life. The lateral masses must be present at birth; however, they remain separated by a remnant cartilaginous cleft posteriorly which usually ossifies by 3–4 years of age to complete the ring.

The axis is formed from three separate sclerotomes: the terminal portion of the dens is formed by the fourth occipital sclerotome (the "proatlas"), the dens is formed from the first spinal sclerotome, and the axis body is formed by the second spinal sclerotome. At birth, the body of the axis and the dens are separated by a vestigial disc called the neural central synchondrosis. It is important to recognize that the synchondrosis is located below the anatomic base of the dens and does not represent the anatomical base of the dens. The synchondrosis is present in most children younger than 3–4 years but disappears by the age of eight. The tip of the dens is not ossified at birth but is represented by a separate ossification center, which usually is apparent by the age 3 years. It fuses to the remainder of the dens by age 12 years (Fig. 12.1).

An understanding of developmental anatomy is important, as certain conditions predispose the pediatric athlete to the injury of the cervical spine. These include patients with os odontoideum, Down syndrome, Klippel-Feil syndrome, skeletal dysplasias, mucopolysaccharidosis, and Marfan syndrome.

Radiographic Parameters

Several radiographic parameters are useful in interpreting pediatric cervical radiographs and avoiding the potentially devastating consequences of a missed diagnosis. These parameters include measurements of the craniocervical junction, defined as the region from the basiocciput to the second cervical interspace (Fig. 12.2). Atlanto-occipital instability can be assessed on plain radiographs using the Powers ratio, the basion-axial interval (BAI), the basion-dens interval (BDI), and the atlanto-occipital joint space. The Powers ratio is the relationship between the occiput and C1 through a ratio of the distance between the basion and C1 posterior arch divided by the distance between the opisthion and anterior C1 arch. A Powers ratio less than or equal to one is considered normal. BAI and BDI values less than or equal to 12 mm are considered normal. Although one researcher specified that the atlanto-occipital joint space in children should measure less than 5 mm at any point in the joint [26], others arbitrarily chose 2 mm as normal [27]. The reported sensitivity of the Powers ratio is 33–60% and that of the BDI is 50%. The BAI is reported to be as sensitive as 100% for detecting true atlanto-occipital injury [28]. In children in particular, these radiographic parameters have been difficult to assess because of the difficulty of accurately observing the involved anatomic structures. A comparison of the interobserver and intraobserver reliability of three different techniques used to measure atlanto-occipital instability found that measurement of atlanto-occipital translation by any of the methods was not reproducible in children with Down syndrome [29], and the authors challenged the reliability of plain radiographs for quantifying instability at the craniocervical junction. A recent study encouraged the use of CT for revealing the craniocervical junction anatomy [30]. Improved sensitivity, specificity, and positive and negative predictive values were reported with CT for the radiographic parameters commonly used in diagnosing atlanto-occipital instability. A BDI of more than 10 mm on CT was recommended as the diagnostic test of choice for confirming atlanto-occipital dissociation because of its ease of use and accuracy for this purpose [31].

Atlantoaxial instability has commonly been assessed using the anterior atlanto-dens interval

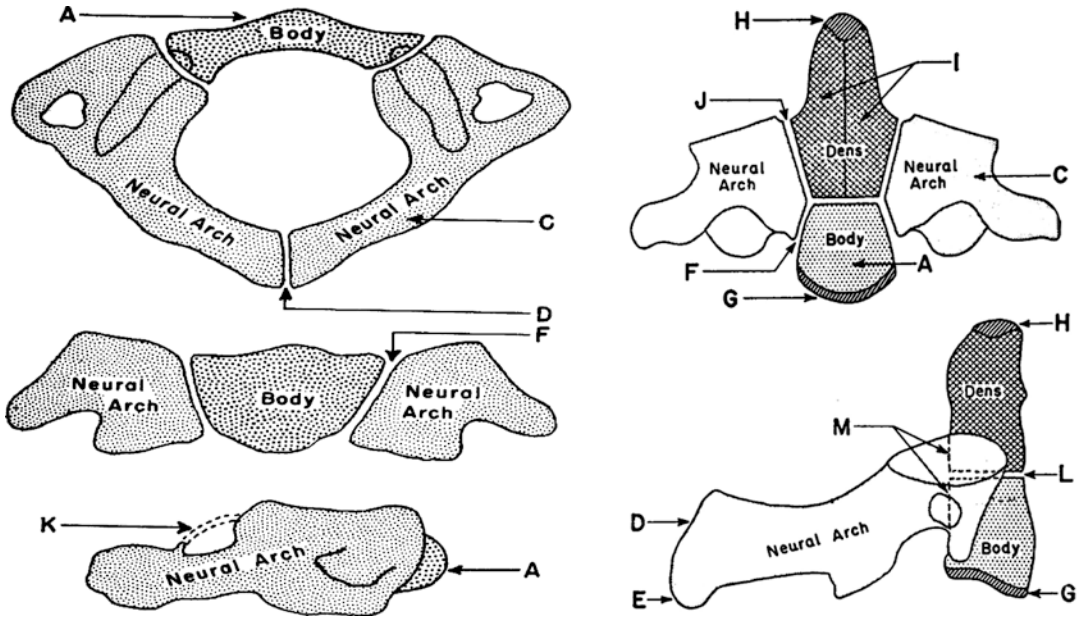

Fig. 12.1 Schematic drawing of the developmental anatomy of C1 and C2: cross-sectional view of C1 and anterior view of C2. Schematic drawing of the upper cervical spine showing a lateral view of common radiographic parameters

(AADI or ADI). In addition to its wide use as a tool for evaluating motion, the AADI is valuable as a marker for predicting cord compression at the atlantoaxial junction. The AADI was found to average 1.9 mm in very young children and to reach 2.45 mm in adolescents [32]. An AADI of less than 3 mm is considered normal between flexion and extension excursion in patients older than eight years; in younger patients the limit is 5 mm. It is important to note that all of these values were obtained from patients with normal cervical anatomy. No current literature has determined whether AADI values beyond these are predictive of cord compression or subluxation. Some authors have suggested that minimal sagittal diameter is an important factor in developing myelopathy, but others believe that the degree of instability is more significant [33, 34]. Although cord compression in adults has been reported when the canal diameter is 14 mm or less, no similar age-related criteria exist for children [35].

The posterior atlanto-dens interval (PADI), particularly in flexion, may be more useful than the AADI for identifying an increased risk of significant neurologic symptoms, particularly in patients with a chronic condition. The PADI represents the space available for the spinal cord (SAC) at the atlantoaxial junction. Absolute stenosis is defined as flexion PADI of less than 10 mm, and relative stenosis is reported to be 10–13 mm. In children, there is uncertainty as to the narrowing of the canal diameter beyond which cord compression occurs, but this measurement can be inferred from the age-related diameter of the

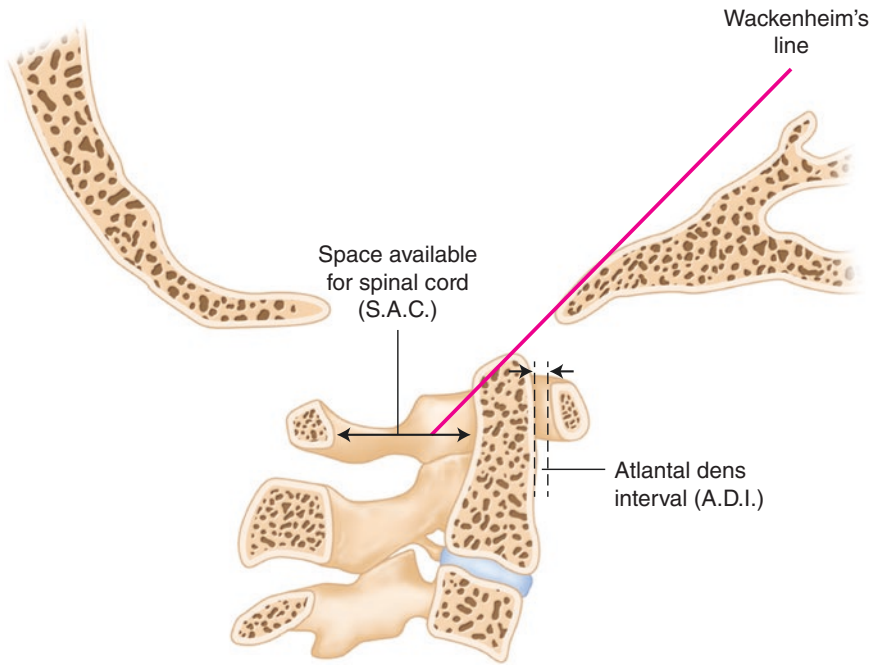

Fig. 12.2 Wackenheim's line is a line extending along the posterior surface of the clivus downward and posteriorly. The tip of the dens should lie below and anterior to this line

cord. The AP cord diameter was reported to range from 4.5 mm in newborns to a maximum of 9 mm in older children [32]. A longitudinal prospective study found average canal diameters in the upper cervical spine of approximately 13 mm at age 6 months and 16 mm in adolescence [32].

The use of CT or MRI is the best means of assessing and confirming spinal stenosis. Spinal stenosis is represented by the direct measurement of the SAC. Although initially described, the Pavlov-Torg ratio for assessing cervical spinal stenosis has phased out as a screening tool. This ratio represented by the cervical spinal canal diameter divided by the anteroposterior width of the vertebral body was reported to identify significant stenosis if the ratio was less than 0.8. Many athletes, however, have normal canal dimensions but large vertebral bodies, therefore artificially decreasing this ratio below 0.8. Functional spinal stenosis defined by the loss of the normal amount of CSF around the spinal cord identified on MRI may be a more accurate measure of risk for cervical spine injuries. Because CT and MRI are static examinations, an emphasis on flexion-extension plain radiographs should be maintained to help identify those injuries that may have dynamic instability. There is an emerging interest in using dynamic MRI for assessing cervical instability [36].

The unique biomechanics and anatomy of the pediatric cervical spine help to explain why upper cervical spine injuries are most common in young children. As the pediatric spine matures, the fulcrum of motion of the cervical spine descends. In children younger than 8 years of age, the maximal mobility occurs between C2-C3, whereas in children between 8 and 12 years of age, the fulcrum changes to C3-C5. Beyond 12 years of age, the fulcrum moves to C5-C6 where it remains through adulthood. In

addition, the immature spine is hypermobile because of ligamentous laxity, shallow and horizontally angled facet joints, underdeveloped spinous processes, and physiologic anterior wedging of the vertebral bodies, leading to high torque and shear forces acting on the pediatric cervical spine. Head size and muscle tone at time of injury can also predispose to upper cervical instability.

Mechanism of Injury

Although mechanisms of injury have been categorized by previous authors into multiple subtypes, controversy continues to exist surrounding which mechanism of injury dominates a particular sporting activity. In football, it is generally agreed that axial loading/compression has attributed to the noted compression fractures, intervertebral disc injuries, and ligament injuries. With the head lowered, the loss of the cervical spine's normal lordosis significantly reduces the cervical spine's ability to absorb and dissipate compressive forces. With continued loading, compressive forces result in the spine "buckling," resulting in disruption of the osteoligamentous complex [37]. Nightingale [38] first documented axial buckling caused by headfirst impact as a mechanism of injury in contact sports. The classic spear tackling in football was felt to result in catastrophic cervical spine injuries by this mechanism. Others have challenged axial buckling as the predominant injury mode in athletes [39–43]. Ivancic et al. [39] and Panjabi et al. [41] have both shown that flexion moments combine with axial compression and shear to produce facet dislocations. Others have noted that several factors such as the angle of the head impact, initial neck posture, and head constraint are all relevant to cervical spine injury from headfirst impact [43]. Pinter et al. [44] and Liu and Dai [42] showed that straightening of the cervical spine away from its normal lordotic posture prior to dynamic compression resulted in stiffening of the spine and compression injuries. Finite element simulations by Hu et al. [45] suggest that considerable muscle activation prior to headfirst impact (as may occur in a player preparing for tackle) significantly increases the injury potential to the cervical spine.

Hyperflexion injuries can cause distraction and stress on the posterior ligamentous structures and lead to ligament rupture. In children, the range of motion throughout the cervical spine is greater than in late adolescents or adults. The pediatric atlanto-occipital joint allows 10° of flexion and 25° of extension, with the remainder of flexion and extension coming from the vertebral segments below. Catastrophic cervical trauma caused by a flexion vector generally results from either a direct blow to the occipital region or rapid deceleration of the torso. Despite Nightingale's initial description of axial buckling, most retrospective studies and systematic reviews have suggested hyperflexion to play a significant role in sports-related injuries to the cervical spine [46–49]. Most authors suggest that this is the primary mechanism of injury in rugby players [50].

Hyperextension injuries commonly are due to falls and often associated with a whiplash injury in which the forehead is struck. These injuries often involve the anterior longitudinal ligament and can also cause transient quadriplegia due to a pincer mechanism [51].

Other described mechanisms of injury include lateral bending, producing traction injuries on peripheral nerve roots. In addition, any combined rotational moment to axial loading, flexion/extension, or lateral bending can be exceptionally dangerous, as these tend to cause dislocation or subluxation injuries in this setting.

The range of injuries produced includes ligamentous and muscle strains, vertebral fractures, intervertebral disc injuries, ligamentous injuries, subluxations, facet dislocations, fracture-dislocations, and spinal cord injuries. It is worth emphasizing that the mechanics of these injuries in sports are multifactorial and that the range of injury mechanisms described above is all possible, and likely present, in some combination for any particular injury.

Types of Injury

Cervical Sprain and Strain

These are common entities in contact sports and characteristically present as localized neck pain with the absence of radiation or neurological deficit. Mildly limited range of motion may be present.

A strain involves injury to the cervical muscle or musculotendinous junction. These injuries generally occur as a result of a force applied to the head or neck during eccentric muscle contraction, causing tissue shearing. The healing process for these injuries has been described to occur in three phases, with hematoma formation and inflammatory response, followed by repair phase and finally remodeling phase.

Acute cervical sprains are most frequent in collision sports. Athletes will often complain of a jammed neck, with localized cervical pain and limited neck range of motion. A restricted range of motion precludes immediate return to play, and if an athlete is unable or unwilling to perform a range-of-motion exam, a more significant injury should be ruled out.

The initial treatment of these injuries should be tailored to the severity of the injury. Rest, ice, and NSAIDS as needed are appropriate interventions. Plain radiographs to rule out osseous injury and instability should be considered. Although White and Punjabi's [52] instability criteria for the subaxial spine are relevant for the adult population, they can serve as a guideline for the pediatric cervical spine. Pseudosubluxation of C2 on C3 should be recognized as a normal variant in the pediatric population up to the age of eight.

Stingers and Burners

Stingers and burners are likely the most common peripheral nerve injury in contact sports. Up to 65% of collegiate football players report a stinger-type injury at least once, and there has been as much as an 87% recurrence rate reported [53]. The C5 and C6 nerve distributions are most commonly involved. Typically athletes present with unilateral upper extremity burning, paresthesias, pain, or weakness after a contact or collision event. Patients lack midline tenderness on palpation and have a full and pain-free range of motion of the neck. The spectrum of burning, tingling, or lancinating sensory disturbance may vary. The symptoms are usually short-lived and resolve spontaneously within a few minutes. Strength is usually restored within 24–48 h. Permanent motor deficits are rare; however, a variable degree of muscle weakness can last beyond 2 weeks and up to 6 weeks in a small percentage of patients [54].

Stingers result from either a traction of the brachial plexus or compression of the cervical nerve root. There is disagreement in the literature as to the exact location and therefore the predominant mechanism of the nerve injury [55]. Although a few studies suggest that nerve root compression may be the more responsible mechanism, the debate is hampered by the failure of imaging or electromyography to accurately identify nerve lesions corresponding with symptoms in affected athletes [56].

The mechanism of traction injury occurs as a result of shoulder depression, with concomitant lateral flexion and deviation of the head and neck to the contralateral side. This motion places the brachial plexus under tensile forces, leading to nerve injury. This traction-type injury seems to occur more frequently in young athletes with less experience and weak neck and shoulder musculature who are without a history of cervical spine injury and who lack radiographic evidence of cervical spondylosis.

Stingers may also result from compression of a nerve root within the neural foramen in response to forceful extension and rotation of the neck toward the affected side. There is continued debate, but this extension-compression may be the most causative mechanism resulting in the stinger presentation. Several authors have described this mechanism to be associated with older college-aged and professional athletes, especially those with preexisting cervical disc disease, cervical stenosis, or other related conditions [55].

There is currently no good evidence that cervical collars or neck rolls protect against stingers. There is also no strong evidence that recurrent stingers increase the risk of more serious injury; however, some investigators propose that a third stinger during a season should prompt consideration for removal of the athlete from play [54, 57, 58].

Cervical Cord Neuropraxia and Transient Quadriplegia

Cervical cord neuropraxia (CCN) is defined as a transient neurologic deficit following spinal cord trauma. Approximately 80% of such injuries involve all four extremities, with variable weakness and combined sensory deficits. Clinically CCN presents with tingling or pain and paresthesias in more than one extremity. A common complaint is the "burning hands syndrome," with painful paresthesias in both hands. The bilateral presentation of CCN represents a central SCI and should not be confused with the single upper extremity presentation of a stinger or burner. A spectrum of complete to partial muscle weakness may be present in CCN, and neck pain is usually not present at the time of injury.

Transient quadriplegia is CCN resulting in paralysis in all four limbs. It is estimated to occur in 0.2 per 100,000 high school football players [59]. By definition, neurological symptoms are temporary in this condition. Complete and rapid resolution of symptoms generally occurs from 15 min to 48 h in adults, but symptoms have persisted for up to 5 days in children [51]. The rate of recurrence has been reported as high as 56% in adults; however, there have not been any reported recurrences in the pediatric population [60].

From a mechanical perspective, momentary cord compression can occur at the extremes of the neck extension or flexion owing to a pincer mechanism. Penning [61] described this as forced hyperextension of a lower cervical motion segment causing an approximation of the posterior margin of the endplate of the cranial vertebral body to the spinolaminar line of the subjacent vertebra. Enfolding of the ligamentum flavum may also contribute to this dynamic narrowing of the spinal canal. In response to hyperflexion force, the pincer effect is created by the endplate of the caudal vertebral body and the lamina of the cranial vertebra. The pathophysiology of spinal cord dysfunction in this condition is thought to be due to a physiologic conduction block without true anatomical disruption.

Management of CCN should be the same as for a suspected catastrophic cervical spine injury. Although imaging is initially with plain radiographs, CT can more readily diagnose fracture or dislocation and is more cost-effective. MRI should be performed to evaluate the cervical spinal cord for injury or impingement. No fractures or frank cord injuries should be seen on imaging in patients with suspected CCN spectrum, although some authors have described the MRI to occasionally show some cord edema.

Certain preexisting conditions are closely associated with CCN and transient quadriplegia, namely, cervical spinal stenosis—either congenital or acquired. Although Torg [62] initially described spinal stenosis by a plain radiographic ratio measurement of the spinal canal to vertebral body length, several studies have subsequently shown that this method is unreliable in assessing cervical spinal stenosis [63–66]. Although in one study a Torg ratio of 0.8 or less was found in 93% of football players with CCN [67], its low positive predictive value (0.2%) for CCN left it ineffective as a screening tool [68, 69]. Although some authors feel that the Torg ratio may be useful in identifying those adult athletes at risk for recurrent CCN after an initial event, the Torg ratio defining cervical spinal stenosis in the pediatric population may not be applicable. A retrospective study of 13 pediatric patients with CCN showed that none of these patients had cervical spinal stenosis [64].

More recently MRI has been used to describe the anatomy and mechanics of spinal stenosis and has surpassed standard radiographs in this regard [70]. The ability of MRI to directly image herniated discs and allow measurement of the spinal column, vertebral discs, and spinal canal and cord makes it the preferred method of evaluation. Spinal stenosis is represented by the direct measurement

of the space available for the cord (SAC). Recent publications place emphasis on quantifying an amount of functional stenosis observed on cervical imaging [71]. There is emerging interest in using dynamic MRI for assessing functional cervical spinal stenosis and possibly aid with return-to-play decision-making.

Disc Disease/Herniation

Cervical disc disease, although uncommon in young individuals, does occur with increased frequency in those participating in contact sports and may be associated with neck, shoulder, or arm pain. Symptomatic impingement most commonly affects the C4–C7 nerve roots [72]. An acute disc herniation can also impinge the spinal cord, causing varying degrees of transient or permanent paralysis. Cervical disc disease can either be classified as soft disc disease or hard disc disease, with the latter being a chronic degenerative process producing a spectrum of spondylotic cervical changes and is most common in adult athletes [73, 74]. Soft disc disease describes an acute annular rupture with extrusion of the nucleus pulposus due to a collision or contact event. Acute neck pain and paraspinal muscle spasms often result, and depending on the cervical level and the presence of foraminal nerve root impingement, a spectrum of neurological symptoms may manifest.

MRI is standard to evaluate suspected disc herniations. It is important to note that imaging must be correlated with clinical findings, as disc protrusion on MRI can be a common finding [75]. In the lumbar spine, the reported prevalence of disc degeneration on MRI or autopsy in individuals between 10 and 19 is 6–16% [76]. The association with low back pain, however, is not fully understood. In a case-control study, no significant difference was found between the incidence of degenerative disc changes in symptomatic and asymptomatic adolescents [77]. Although most of the literature concentrates on degenerative disc disease of the lumbar spine, the data can be extrapolated to understand cervical disc disease to some extent, in particular that MRI changes and symptomatic disc disease does not have an exact correlation.

MRI is useful in confirming a disc herniation, defining its size and level, and determining whether it is posterolateral or far lateral. A short course of NSAIDS, rest, and activity modification usually are successful. Although physiotherapy is also commonly recommended, a recent evidence-based review found insufficient evidence for the effectiveness of physical therapy in this setting [78]. Patients with persistent arm-dominant radicular pain may be candidates for a diagnostic and therapeutic selective nerve root injection with a local anesthetic and steroid. Surgery is indicated for a patient whose radicular symptoms persist after nonsurgical modalities are exhausted or for a patient with progressive neurology.

Fractures and Fracture/Dislocation

As noted earlier, the injury patterns are a direct result of the unique anatomy and kinetics of the pediatric cervical spine. A large head-to-body ratio and intrinsic elasticity, especially in children younger than eight years, commonly lead to injury between the occiput and C2. In young children, the fulcrum for flexion is at approximately C2-C3. With increasing age, the head-to-body mass ratio decreases, and the fulcrum moves caudally to the C5-C6 level. The horizontal orientation and shallow nature of the facet joints in children increase translational mobility and movement during flexion-extension. Pediatric and adult cervical spine kinematics were compared using the helical axis of motion (a three-dimensional analogue defined as an axis along which an object translates and about which it rotates) [79]. Cervical spine kinematics was found to vary with both age and sex; young girls had a more anterior helical axis of motion in flexion-extension and axial rotation than either adult women or adult men. These differences in cervical spine kinematics may account for the greater incidence of upper cervical spine injury in children compared to adults.

The movement of the fulcrum of flexion distally to C5-C6 in adolescents explains the distribution of traumatic subaxial spine injuries. Sports-related cervical spine injury most commonly occurs in the subaxial spine for older children because their biomechanics more closely resemble that of adults. Some authors believe that more stringent radiographic criteria than normally used should be applied for cervical instability in children [80, 81]. Angulation greater than 7°, translation of more than 4.5 mm in younger children, and subluxation of more than 3.5 mm in children older than eight years should raise suspicion of injury (Fig. 12.3a–c).

In the young athlete, fracture patterns normally encountered can include stable avulsion fractures (e.g., spinous process "clay shoveler's," odontoid avulsion injury, etc.), compression fractures of the anterior column (e.g., wedge compression injuries), or burst fractures involving both the anterior and middle columns (e.g., c1 Jefferson fracture). Unstable patterns involving all three columns include flexion-distraction injuries, which may be bony, purely soft tissue, or a combination (Fig. 12.4a–d), the flexion teardrop injury (Fig. 12.5a–c), and unilateral or bilateral facet dislocations (Fig. 12.6a–c).

Management should be based on stability of the injury, neurology, and patient factors. Although most injuries can be managed with external immobilization, modern instrumentation techniques for internal fixation with posterior lateral mass screw-rod constructs and anterior plating systems are increasingly being used. They have the advantage of obviating the need for postoperative immobilization and avoiding the frequent complications of halo orthosis.

Return to Play

The decision on whether the young athlete can return to playing sports is complex and must be considered on an individual basis. Most published data offer expert opinion with limited objective data. The literature supports the concept of a gradation of increasing risk with different sports [82]. Torg, Albright, Cantu, and others [83–91] have established some guidelines in regard to return-to-play decisions in contact sports. Cantu [90] recommended that prior to return to play, the athlete should have no neck tenderness or spasm, neck or arm pain, paresthesias, or weakness, whether at rest, with neck range of motion, or on axial compression. Watkins [91] proposed a point

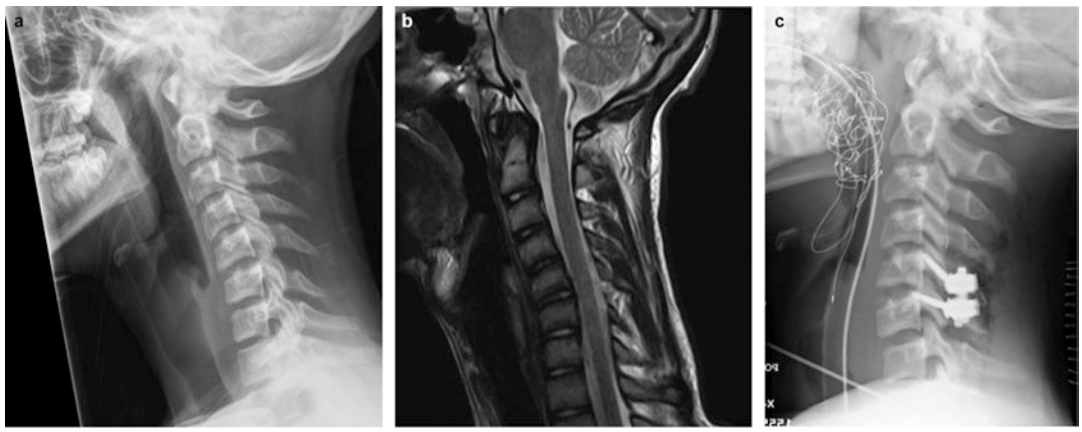

Fig. 12.3 (**a–c**) 13-year-old male following scrum-related rugby injury. (**a**) Lateral X-ray showing acute kyphosis at C5-C6 measuring greater than 7 degrees. (**b**) MRI confirming significant flexion-distraction injury with complete disruption of the ligamentum flavum. (**c**) Postoperative X-rays showing rigid single-level fixation with lateral mass screw-rod construct and restoration of normal cervical lordosis

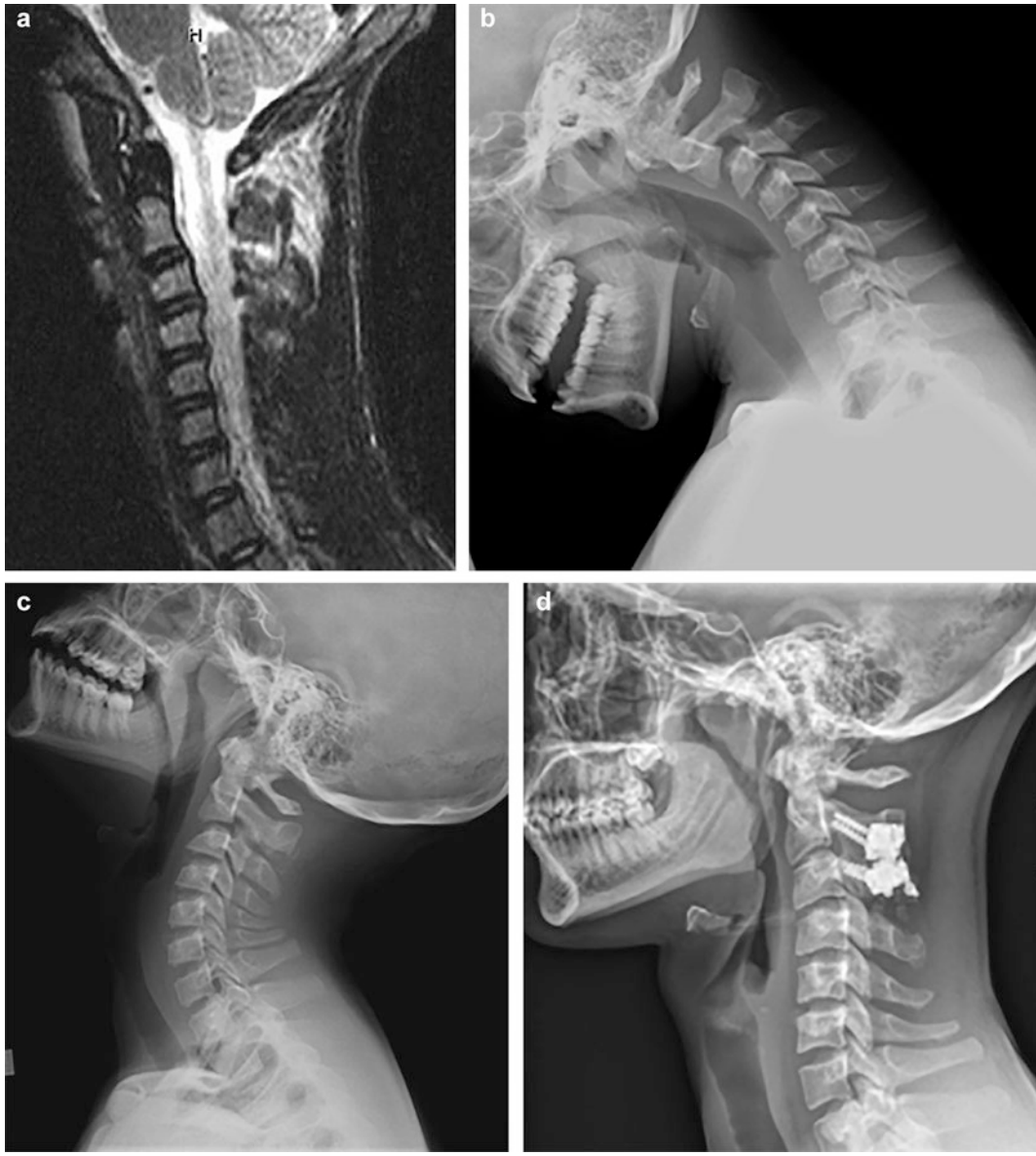

Fig. 12.4 (**a–d**) (**a**) MRI of a 14-year-old male who sustained an injury to cervical spine wrestling. Note the posterior signal change at the C2 interspace suggestive of a ligamentous injury. (**b**, **c**) Flexion-extension radiographs 3 months after injury. Note the translation and facet subluxation upon flexion and reduction in extension. (**d**) Postoperative lateral radiograph after the patient underwent C2–3 instrumented fusion for pain and instability

grading system to quantify the patient's clinical situation and offer an objective guideline for return to play. The investigators looked at three topics and assigned point value systems based on the extent of the neurological injury, the time from injury to treatment, and the narrowing of the central canal diameter. Patients with 0–6 points were thought to have minimal risk of injury with return to play; 6–10 points represented moderate risk; and 10–15 points were associated with high risk. Unfortunately, guidelines for return to play in sports with less inherent risk have not been established. Most guidelines do not categorize

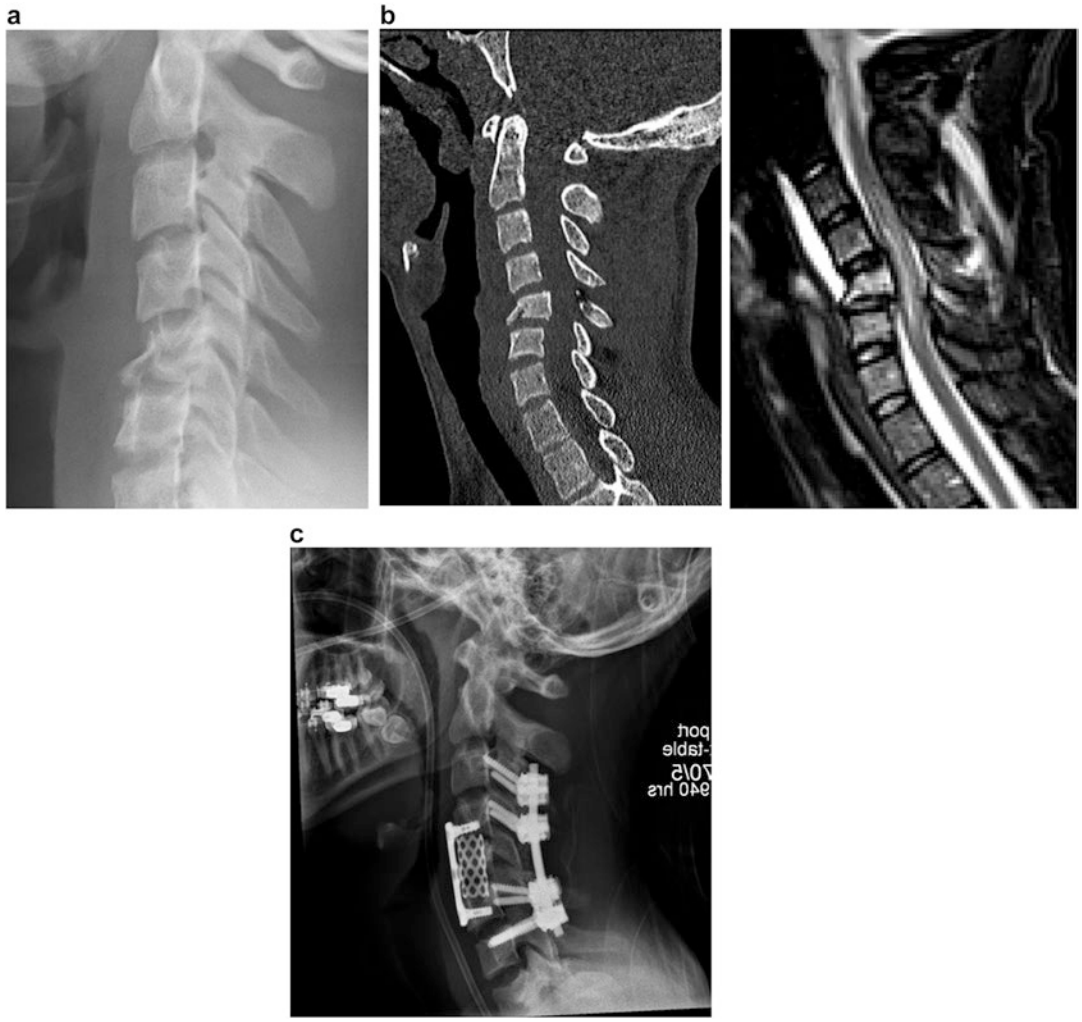

Fig. 12.5 (**a–c**) 14-year-old male with cervical unstable flexion teardrop variant injury following snowboarding. (**a**) Preoperative Lateral X-ray. (**b**) CT and MRI showing extent bony injury and spinal cord injury. (**c**) Patient was treated with a circumferential fusion

sports beyond "contact activity," which is not specifically defined [92]. Athletes in noncontact, high-velocity sports are at a similar, if not greater, risk for cervical injury, and Torg's criteria may be appropriate [93].

Some experts currently advocate return to play in collision sports for asymptomatic athletes after a single-level anterior cervical discectomy and fusion (ACDF) as seen in Fig. 12.7a–c [94, 95]. Although traditional recommendations have been to reserve surgery for patients failing conservative measures for up to 12 weeks, many authors are now recommending early surgical intervention rather than nonsurgical therapies. A recent study of NFL athletes looked at performance-based outcomes of football players undergoing ACDF compared to those opting for conservative treatment [96]. Seventy-two percent of those who underwent surgery successfully returned to play, whereas 46% of those treated with conservative measures alone returned to play successfully [96].

It remains unclear how the study of NFL players can inform the decision-making process for young athletes. The study was retrospective,

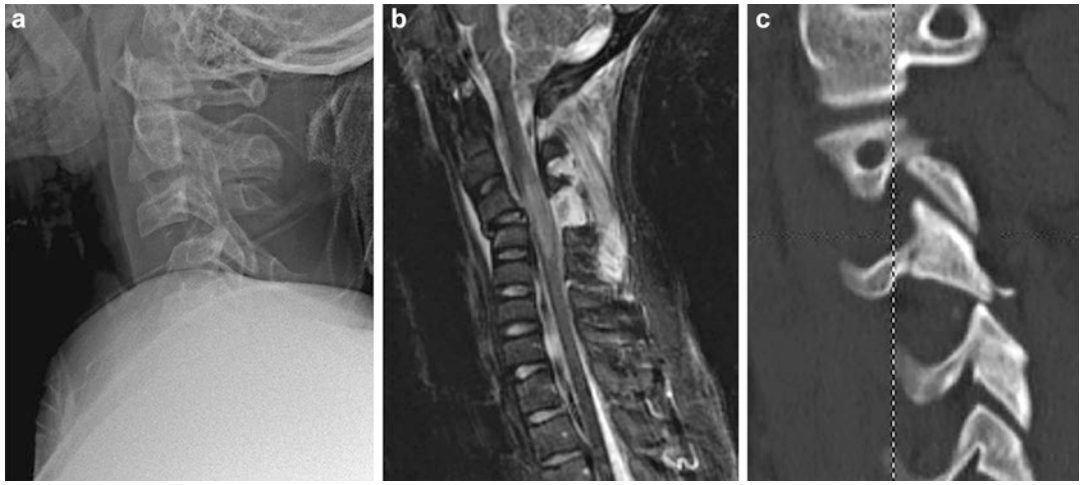

Fig. 12.6 (**a–c**) (**a**) Lateral radiograph of the cervical spine in a 13-year-old male who sustained a neck injury while playing hockey. Note the displacement suggestive of a facet dislocation. (**b**) MRI showing the significant signal changes at the facets and posterior interspinous region as well as disc injury at C3–4. (**c**) CT scan after attempted reduction of the dislocated facets with awake traction; note the persistent subluxation of the facet and incongruity of the joint

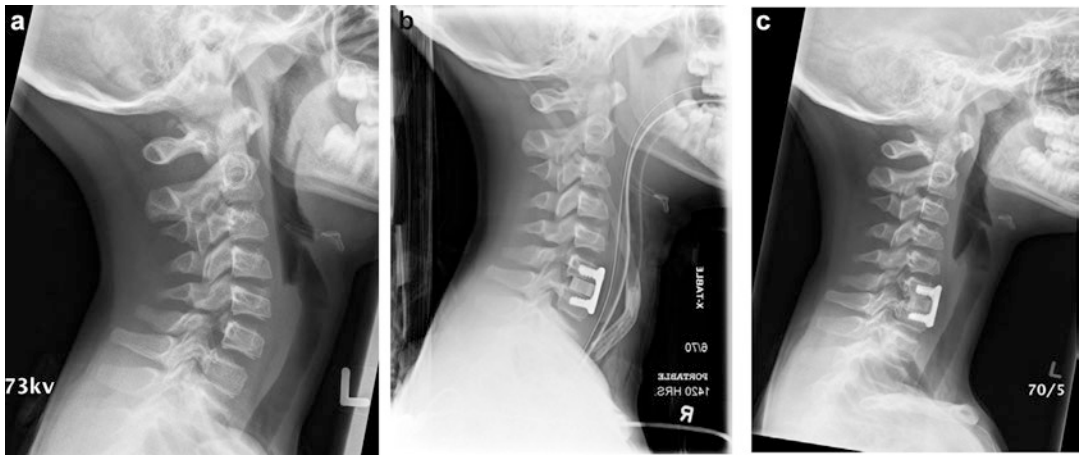

Fig. 12.7 (**a–c**) 8-year-old male with C6 pars defect and C6 on C7 anterolisthesis presented with neck pain after soccer-related injury. (**a**) X-rays on presentation showing C6 pars defect with listhesis C6 on C7. (**b**, **c**) Patient treated with ACDF and returned to play once graft incorporated

nonrandomized, did not evaluate concomitant cervical spine stenosis, and did not detail the type or grade of disc disease between the two groups. Professional athletes who have spent more time involved in contact sport could be expected to have a higher degree of cervical spondylosis than high school athletes.

Morganti and colleagues [92] attempted a questionnaire survey study to evaluate which factors such as published guidelines, type of sport of the patient, number of years in practice, subspecialty interests, and sport participation of the respondent had a role in the return-to-play decision-making process. The authors found the

opinions to vary regarding the decision to allow an athlete to return to play after a cervical spine injury. Collision and high-velocity noncontact sports were perceived to hold greater risk for further injury following a previous cervical spine injury.

Summary

Cervical spine injuries in athletes can be potentially devastating. A high index of suspicion must be maintained for underlying cervical instability and soft tissue injuries that frequently accompany collision and high-velocity sporting injuries. Plain radiographs remain useful as a first-line screening tool for assessing instability and osseous injury; however, MRI is gaining favor in assessing functional stenosis and is warranted in the presence of neurologic symptoms or radiographic evidence of instability. Although the majority of sports-related injuries to the cervical spine can be managed conservatively, newer instrumentation techniques providing rigid fixation for stabilization and arthrodesis are becoming increasingly popular for children and adolescents.

The decision to allow an athlete to return to play can be difficult for the physician, the player, and his or her family. Risk factors are not well defined, and the risk-benefit ratio differs depending on the value system and biases of both the physician and athlete. The literature remains inconclusive in providing firm, objective guidelines for sport participation following a cervical spine injury, with most authors recommending that this decision be made on an individual basis.

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Part III

The Medical and Surgical Treatment of Cervical Disorders in Children

13 Medical and Rehabilitative Techniques in Cervical Disorders of the Child

Katrina M. Lesher

Congenital Torticollis

Congenital muscular torticollis (CMT) is a condition in which the sternocleidomastoid (SCM) muscle is shortened on the involved side, leading to an ipsilateral tilt of the head and a contralateral rotation of the face and chin. It is a finding, not a specific diagnosis, is relatively common, and is known by many names including wry neck and loxia. The term torticollis is derived from the Latin words *tortus* meaning twisted and *collum* for neck. It can be further classified and described based on the resultant positioning of the head and neck. The most well-known form of CMT involves rotational and lateral bend of the head due to shortening of the SCM muscle on one side [1–3]. The SCM muscle originates from both the manubrium of the sternum and the medial third of the clavicle. These two tendons then combine to form a single muscle bundle that runs superiorly and posteriorly to its insertion at the mastoid process of the temporal bone and the superior nuchal line of the occipital bone. It is innervated by the ipsilateral accessory nerve, and its vascular supply is from the occipital artery and superior thyroid artery. The actions of the SCM muscle rotate the head to the opposite side in an oblique fashion, as well as laterally flexing the neck to the ipsilateral side. The SCM also acts as an accessory muscle of inspiration in conjunction with the scalenes. In utero positioning can lead to shortening of the SCM muscle, and this is the most common etiology for uncomplicated CMT. Most patients present early in infancy, and cases are generally identified in the primary care office and then are referred to appropriate specialists for further treatment and management.

It is important to identify any underlying craniocervical vertebral anomalies or ocular abnormalities, such as strabismus or congenital nystagmus, which may be contributing to the abnormal head positioning prior to initiating therapy. Other nonmuscular causes can include Sandifer's syndrome resulting from gastroesophageal reflux, neural axis abnormalities, and benign paroxysmal torticollis (discussed later in this chapter). Imaging is warranted if there are any concerning clinical findings such as abnormal eye positioning, facial asymmetry, asymmetric use of upper extremities, or other abnormalities on physical exam [4]. In newborns or infants with congenital torticollis, ultrasonography (US) is the imaging modality of choice. Sonographic findings are typical and include an enlarged SCM with heterogeneous texture [5]. Recent studies

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have discussed the use of adding sonoelastography to conventional US for the diagnosis of congenital muscular torticollis and to aid in predicting treatment outcomes [6–9]. Most cases of uncomplicated congenital muscular torticollis are successfully treated with conservative management with physical therapy, including range of motion and stretching exercises [10].

Diagnosis begins with a detailed history and physical examination. Clinical findings can include a fibrous band within the SCM muscle mass, limited cervical range of motion, head positioning of cervical rotation away from the side that is shortened, and lateral tilt toward the shortened side. Secondary findings may include positional plagiocephaly or a flattening of the ipsilateral parieto-occipital region. Plagiocephaly is frequently present, due to the head being positioned in one direction for the majority of the time. Measurements of cranial symmetry including ear positioning, facial asymmetry, and 3D scanning are useful to determine severity. Cranial molding helmets may be prescribed for moderate to severe cases of uncomplicated positional plagiocephaly without underlying craniosynostosis [11]. Hip dysplasia is associated with CMT in about 15% of cases [12, 13]. Always assess the hips in patients with torticollis and perform imaging as appropriate, based on history and physical exam findings.

Although torticollis is a sign of an underlying disease process, its presence does not imply a specific diagnosis, and the cause should be sought if torticollis persists or is associated with other symptoms [5].

Once the diagnosis is made, therapeutic stretching exercises and appropriate positioning are the mainstay of treatment. Most cases improve and resolve with this conservative approach. Supervised therapy sessions with a licensed, qualified physical therapist should focus on instructing the caregivers on proper techniques for stretches and recommendations for positioning. Evidence-based clinical practice guidelines were published in 2013 for physical therapists to guide referral, screening, examination and evaluation, prognosis, first-choice and supplemental interventions, consultation, discharge, and follow-up of patients with congenital muscular torticollis [14]. Outpatient physical therapy treatment should involve passive stretching and range of motion, with an emphasis on education to caregivers in order to ensure carryover therapy in the home. Carryover of appropriate positioning techniques and consistency with stretching exercises outside of formal supervised therapy are essential for successful outcomes. Unfortunately, compliance with any recommended home program is less than ideal [15].

Caregivers should be instructed that the best place to perform the stretching exercises is on a stable, supportive surface, such as a changing table or carpeted floor. The child should be supine to perform the cervical side bend and rotation stretches. For left SCM shortening, the caregiver should hold the child's left shoulder down with their right hand. They then place their left hand on top of the left side of the child's head. They can then slowly bend the child's right ear toward their right shoulder (Fig. 13.1). Hold the position for 15 s, repeat three times, and perform three times a day. For right-sided torticollis, bend the ear toward the left shoulder (Fig. 13.2).

The next stretch for left-sided torticollis involves placing the caregivers' right hand on the child's right shoulder. They should then cup the child's head with their left hand and slowly turn the child's nose toward their left shoulder (Fig. 13.3). Hold the position for 15 s, repeat three times, and perform three times a day. It is important to position the child in side-lying or prone position for playtime to take advantage of gravity in the stretching process. Parents should be instructed to place all things of visual interest to the ipsilateral side to encourage rotation back toward side of shortening (Fig. 13.4). This is often confusing for parents, as they think the shortened muscle is the one on the side that the patient turns their head toward, when in fact it is the opposite side that is tight. Additional instructions can be provided to have the caregivers carry the child in a forearm-supported side-lying carry position (Fig. 13.5). Conservative management with stretching and therapy should always be the initial treatment regimen for uncomplicated CMT. These stretching and positioning techniques can be supplemented with modalities including kinesiotaping, massage,

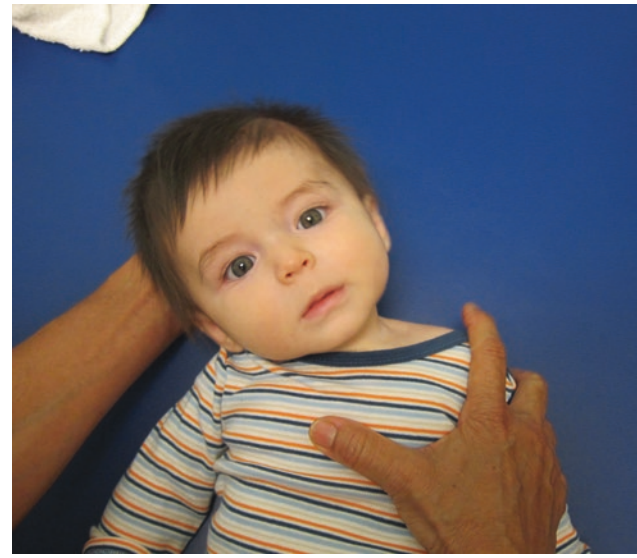

Fig. 13.1 Lateral bend stretch to the left

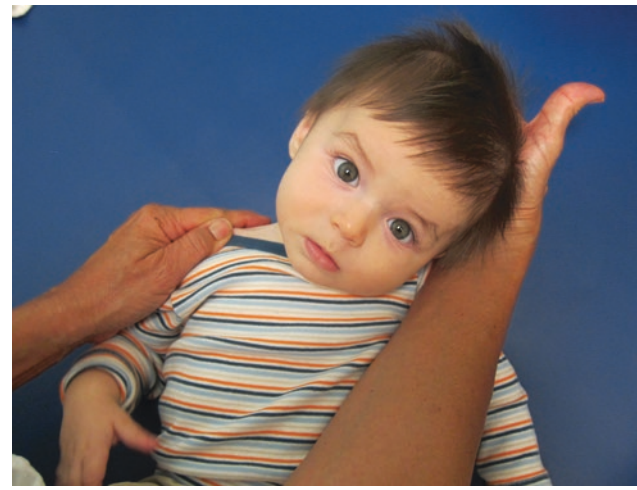

Fig. 13.2 Lateral bend stretch to the right

and cervical orthoses such as the TOT collar [16]. Positioning devices such as the TOT collar can be helpful for decreasing the frequency of abnormal head positioning in smaller infants but can be difficult for parents to apply correctly, and as children get older and stronger, they are able to pull the collar off, rendering it essentially ineffective. Education and counseling should be provided to parents who are considering alternative therapy options for CMT, including chiropractic intervention. There is no role for cervical spine manipulation in the treatment of CMT in infants and children [17].

Only after failure of conservative management options should interventional options, including focal botulinum toxin injections or, as a last resort, surgical release, be considered. Focal botulinum toxin injections can be performed to facilitate relaxation of the spastic sternocleidomastoid muscle and allow improved stretch and range of motion (ROM) in patients with persistent limitations with ROM after a trial of more conservative treatment measures. Botulinum toxin decreases muscle activity by blocking the release of acetylcholine from the presynaptic nerve terminals. Risks and benefits of the injections should be discussed with the care-

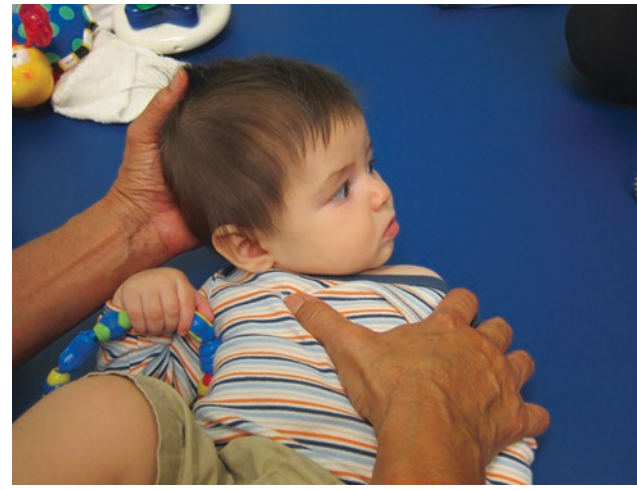

Fig. 13.3 Cervical rotation stretch to the left

Fig. 13.4 Active play positioning

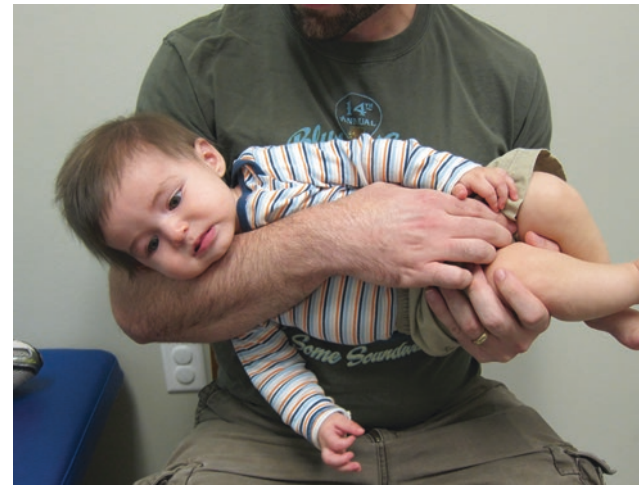

Fig. 13.5 Carrying position for right torticollis

givers, and after informed consent is obtained, the injections can be performed in the outpatient clinical setting. The child should be positioned in the supine position with extremities wrapped tightly and close to their body, on a papoose board or using linens. Trained medical staff (not the caregivers) can assist in holding and positioning the child's head for optimal results and minimizing risk of complications. A small amount (units vary depending on which preparation of botulinum toxin is used) of botulinum toxin is injected just distal to the insertion of the SCM into the mastoid and a second injection proximal to the origin of the clavicular head of the muscle above the supraclavicular fossa. An additional injection in the ipsilateral trapezius is often beneficial if the child is demonstrating signs of asymmetrical shoulder elevation. It is of utmost importance for the child to be positioned and restrained accordingly. Aspiration prior to injection at each site is essential to ensure proper placement of the needle and to avoid accidental intravascular injection. Care must be taken during the injection process to minimize movement of the child and ensure proper instillation of toxin into the targeted muscles, while avoiding key structures, including the carotid artery and jugular vein, nearby. Intravascular injection of botulinum toxin can lead to systemic effects including respiratory depression and potential paralysis. The injections are only mildly uncomfortable and generally well tolerated. There are no special aftercare instructions following the injections other than monitoring for signs of bruising or infection. Ultrasound or EMG guidance is not necessary for muscle localization and can cause unnecessary discomfort for the patient. Anatomic localization of the targeted muscles is sufficient. The toxin begins its therapeutic effect in about 3–5 days, peaks at 1 month, and continues to work for about 3–4 months. The goal during this time is to optimize therapies, including stretching and positioning, in order to maximize the benefits of the toxin. In an ideal clinical protocol, patients would come in to an outpatient clinic for an evaluation, return in 1–2 weeks for the injection (allowing time for insurance authorization), come back for a 1-month follow-up to determine clinical effectiveness, and then return again 3–4 months later after the botulinum toxin has worn off. Continued supervised physical therapy sessions, along with carryover of the home stretching and positioning program, are paramount in maximizing the effectiveness of the botulinum toxin injections. Injections should not be performed more frequently than every 4 months. Generally, one to two rounds of injections will provide significant improvement and help identify patients who may be candidates for surgical release.

If a patient has persistent limitations in range after an appropriate trial of conservative management and botulinum toxin injections, consider evaluation for a surgical release. Occasionally, patients present with congenital muscular torticollis later on in childhood, or even as adults, at which time conservative management is rarely successful. Only after patients have failed conservative management should surgical intervention be considered. Congenital anomalies of the occipital condyles and upper cervical spine must be ruled out before performing a release of the sternocleidomastoid muscle in a child who fails to improve with physical therapy [18–20].

If left untreated, congenital muscular torticollis can lead to persistent deformational plagiocephaly and progressive facial asymmetry, and these children may be at risk for later neurodevelopmental issues [21–23]. Infants with CMT who are diagnosed earlier and have earlier intervention tend to have a shorter duration of rehabilitation. Initial cervical ROM seems to be an important prognostic factor for predicting the rehabilitation outcome of patients with CMT [24]. In summary, it is important to differentiate muscular from nonmuscular torticollis. Congenital muscular torticollis is benign; and missing a case of nonmuscular torticollis could be potentially life threatening [25].

Acquired Torticollis

Torticollis or abnormal neck positioning that develops after the first few weeks of life (not congenital) always warrants evaluation and workup for underlying etiology. Acquired torticollis can be secondary to trauma, infection, or tumors [26–30]. It can be due to underlying congenital bone

malformations (Klippel-Feil syndrome), GE reflux (Sandifer syndrome), hemifacial microsomia, oculomotor nerve palsies, and syringomyelia. Other causes can include meningitis, upper respiratory infections, otitis media, mastoiditis (Bezold's abscess), cervical adenitis, retropharyngeal abscess, CNS or bone tumors, vertebral osteomyelitis, and epidural abscess.

Benign paroxysmal torticollis of infancy (BPTI) is a disorder characterized by recurrent episodes of head tilt secondary to cervical dystonia. Symptoms present around 3 months of age, with intermittent episodes lasting from minutes to weeks. Shorter episodes can be accompanied by vomiting, pallor, and ataxia, resolving spontaneously within hours or days. Episodes begin within the first 12 months of life and resolve spontaneously by age 5 [31].

Post-traumatic torticollis can occur due to muscle spasm, unilateral interfacetal dislocation (UID), and occipital condyle fracture. In younger infants, fibromatosis colli is a relatively rare cause of acquired torticollis.

Fibromatosis colli (FC) is a rare form of fibromatosis involving the sternocleidomastoid muscle, resulting in diffuse enlargement of the SCM muscle [32, 33]. It is also known as pseudotumor of the SCM. Though the exact etiology is not known, it is most likely due to birth trauma and can be seen following a difficult delivery requiring the use of vacuum extraction or forceps delivery. Infants usually appear normal at birth but present with abnormal head positioning at 2–3 weeks of life. Parents report noticing a swelling on the side of the neck about 2 weeks after birth. The diagnosis can be made by ultrasound, which shows spindle-shaped thickening of the SCM on the affected side in contrast to the normal contralateral side. There is no cervical lymphadenopathy and no vascular invasion or bony involvement, as may be seen with other neck masses. Real-time ultrasonography shows synchronous motion of the mass with the sternocleidomastoid muscle, thus confirming the diagnosis. FC is a self-limiting condition and usually resolves within 4–8 months [33]. It is important, therefore, to quickly identify fibromatosis colli as such in order to avoid unnecessary expenditures of resources and to promptly begin conservative treatment.

In cases of acquired torticollis, imaging should be used as a general screening tool after a complete medical history and examination have been obtained. In cases of acquired torticollis resulting from trauma, conventional radiography (lateral and anteroposterior views) should be the first-line imaging modality. In older children with post-traumatic or acquired torticollis, CT of the neck and cervical spine is needed to evaluate for fracture or malalignment. If the CT is positive, consider further MRI/MRA studies to evaluate for associated spinal cord, ligamentous, or arterial injuries. In nontraumatic acquired torticollis, CT of the neck or cervical spine is the initial imaging study. If there is no history of trauma and the CT is negative, perform a brain and spinal cord MRI to exclude an underlying CNS cause [5]. Management of acquired torticollis focuses on identification and treatment of the underlying cause.

Soft Tissue Injury

The pediatric cervical spine is at risk for soft tissue injury due to a variety of factors, and cervical musculature strains or sprains do occur in childhood, although less commonly than in adults. Children younger than 8 years tend to have more pure ligamentous injuries rather than fractures and more involvement of the occipitoatlantoaxial complex. Subaxial soft tissue injuries have been reported in children under the age of 8 but are relatively rare. Children under 8 years have a relatively large head compared to the size of their shoulder girdle and torso, and with hypermobility of the upper cervical spine, this results in a higher-placed fulcrum of flexion compared to older children [34]. Cervical soft tissue injuries can be difficult to identify, particularly in younger, nonverbal children.

Motor vehicle accidents account for the largest number of injuries, and sports injuries are more common in older children compared to younger

ones. These soft tissue injuries generally involve muscle strain or ligament sprain. A ligamentous sprain is usually caused by a sudden flexion or extension of the neck. Symptoms frequently involve neck pain and muscle spasm and can lead to decreased range of motion. Nonverbal children may display increased irritability, a head tilt, and signs of tenderness to palpation over the affected muscle group. Any neurologic signs including numbness, tingling, or weakness should be evaluated thoroughly. Management of more serious soft tissue injury or cervical spinal cord injury is outside of the scope of this chapter.

For patients with negative imaging and no neurologic signs, conservative measures including heat, massage, and stretching are usually sufficient for complete resolution of symptoms. Instruct caregivers to avoid immobilization. Acutely, they can apply cold or ice packs for 15–20 min. four times daily for 2–3 days and then can transition to moist heat. Scheduled NSAIDS and, in severe cases, a short course of a muscle relaxant generally not to exceed 2 weeks can be prescribed for pain control in the acute period. Outpatient physical therapy can be utilized to perform range of motion, stretching, and strengthening exercises. Modalities including massage, heat, TENS, and traction may be incorporated into the therapy sessions. Strengthening exercises should only be initiated once the child is pain-free. Therapeutic exercises and modalities will focus on relaxation of the spastic muscles and strengthening of the cervical spine stabilizers to prevent reinjury.

Summary

Non operative techniques for congenital torticollis and soft tissue injuries include physical therapy and pharmacologic management. A thorough evaluation of the child with neck pain thought to be due to these etiologies must be undertaken to ensure that an underlying congenital or other cause is not missed. Symptoms usually resolve and persistent complaints indicate more advanced investigation.

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Surgical Approaches to the Child's Cervical Spine

14

Haemish A. Crawford and William Warner

Anterior Approach

An anterior approach to the pediatric cervical spine may be utilized to treat infections, congenital abnormalities, and tumors, as well as for spinal stabilization. These conditions may be associated with neurologic compromise, and decompression of the injured spinal cord may be done from an anterior approach. The level of the cervical spine that needs exposure will dictate what type of anterior approach is used. The cervical spine exposures can be divided into three levels: (1) base of the skull (clivus), atlas, and odontoid; (2) C3 to C7; and (3) C7 and T1.

Approaches to the Upper Cervical Spine (C1-C2)

Anterior approaches to the upper cervical spine (C1-C2) include the lateral retropharyngeal approach, transoral approach, and transoral combined tongue and mandible-splitting approach. The retropharyngeal approach usually is preferred, when possible, because of the increased incidence of wound complications and infection associated with the transoral approaches.

Endoscopic approaches also have been described for anterior resection of the odontoid. The endoscopic approaches to the odontoid and anterior ring of C1 may be intranasal, sublabial, or from an anterior cervical approach (Fig. 14.1a–c) [1–4].

Fang and Ong described a transoral approach to the anterior arch of C1 and the odontoid [5]. This surgical approach gives the most direct approach to pathology involving the clivus and anterior arch of the C1 and C2 vertebral bodies. This dissection should not go beyond the lateral border of the lateral mass of the C1-C2 articulation to avoid damage to the vertebral arteries.

Because of the increased risk of infection with this approach, prophylactic antibiotics are given based on preoperative nasopharyngeal cultures. Endotracheal intubation is achieved with a noncollapsible tube and cuff, and if extensive dissection is anticipated, a tracheostomy should be performed. The vertebral bodies are identified by palpation, and a mouth gag is used to provide retraction of the tongue.

The ring of the first vertebra has a midline anterior tubercle, and the disc between the second and third vertebrae is prominent, providing localizing landmarks. Through a longitudinal incision in the midline of the posterior pharynx, the soft palate can be divided in the midline,

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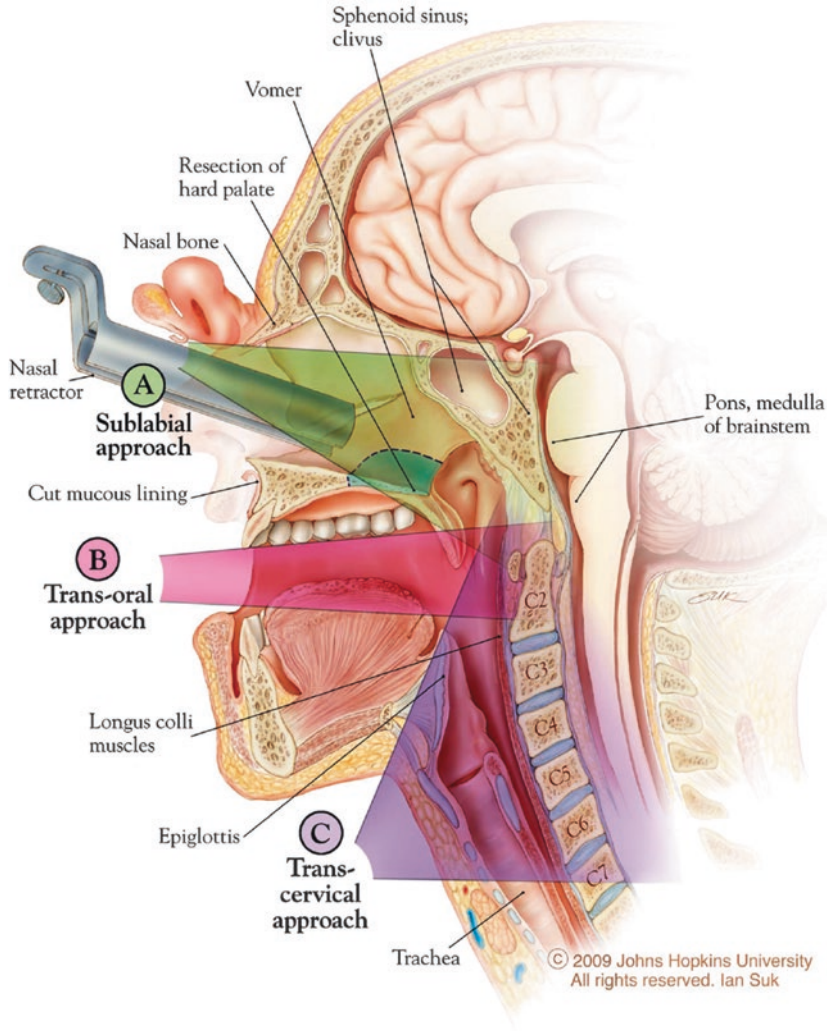

Fig. 14.1 Relevant normal anatomy and operative angles for endoscopic approaches to the upper cervical spine: (**a**) sublabial, (**b**) transoral, (**c**) transcervical (Reprinted with permission from Bettegowda et al. [1])

making post-retraction paresis less likely, or it can be folded back on itself.

The midline dissection is carried down to bone, with the soft tissue reflected laterally to the outer margin of the lateral masses of the axis. Any dissection beyond these margins will place the vertebral arteries at risk. The soft-tissue flap can be retracted with long stay sutures (Fig. 14.2a, b).

After the procedure is complete, the wound is irrigated and closed loosely with interrupted absorbable sutures. Because of the frequency of postoperative infections with this approach, continuing antibiotics for at least 3 days after surgery is recommended.

Spetzer et al. [6, 7] recommended using a self-retaining McGarver three-ring retractor to allow the lips and gums to be spread apart and the tongue and endotracheal tube to be displaced caudally. Two small, red rubber catheters also are inserted through the nostrils into the oropharynx and sutured to the uvula. The uvula and soft palate are retracted into the nasopharynx by placing traction on the two rubber catheters. This combination of retraction provides wider operative

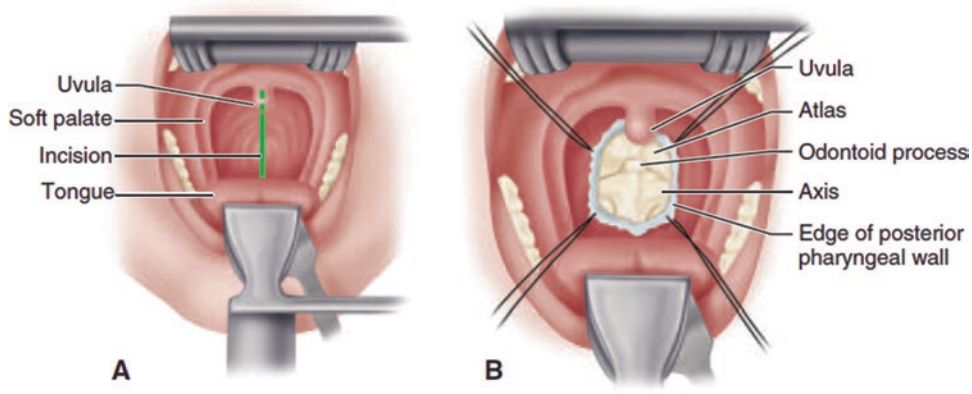

Fig. 14.2 (**a**, **b**) Transoral approach to upper cervical spine for exposure of anterior aspect of atlas and axis (Reprinted from Warner [17]. With permission from Elsevier)

exposure of the posterior pharyngeal wall and often may eliminate the need for a tracheostomy.

Transoral Combined Mandible-Splitting and Tongue-Splitting Approach

Hall, Denis, and Murray [8] described a mandible-splitting and tongue-splitting transoral approach to the cervical spine. This approach gives a more extensive exposure of the upper cervical spine than the transoral approach. Hall et al. recommended that this approach be used only in special circumstances, given the significant patient morbidity and technical challenges of the approach.

Halo immobilization and tracheostomy usually are required before the procedure. An incision is made from the anterior gum margin through both surfaces of the lower lip and down over the middle of the mandible to the hyoid cartilage. Traction sutures can be used to allow better exposure of the midline raphe. The tongue is divided longitudinally to the epiglottis through its central raphe with the electrocautery. The lower incisor is removed, and a step cut is made in the mandible with an oscillating saw. The uvula can be folded on itself and sutured to the roof of the soft palate. The mandible and tongue are retracted down on each side to improve exposure. The mucosa is opened over the posterior wall of the oral pharynx to expose the anterior cervical spine from the first cervical vertebra to the upper portion of the fifth cervical vertebra.

The anterior longitudinal ligament is divided in the midline and reflected laterally to allow enough exposure of the anterior portion of the cervical spine.

During closure, the posterior pharyngeal flap is repaired with 3–0 chromic suture, and a suction drain can be placed through the nose and inserted deep to the pharyngeal flaps. The tongue is repaired with 2–0 and 3–0 chromic sutures, and the mandible is fixed with wires inserted through drill holes on each side of the osteotomy. The infralingual mucosa is closed with 3–0 chromic sutures, and the subcutaneous tissue and skin are closed. The use of preoperative and postoperative antibiotics is recommended (Fig. 14.3a–c).

Anterior Retropharyngeal Approach

McAfee et al. [9] used a superior extension of the anterior approach of Robinson and Smith to the cervical spine. This approach provides exposure from the atlas to the body of the third cervical vertebra without the need for posterior dissection of the carotid sheath or entrance into the oral cavity.

Gardner-Wells tongs are applied with 4–5 kg of traction, and the neck is carefully extended with the patient awake. The maximal point of

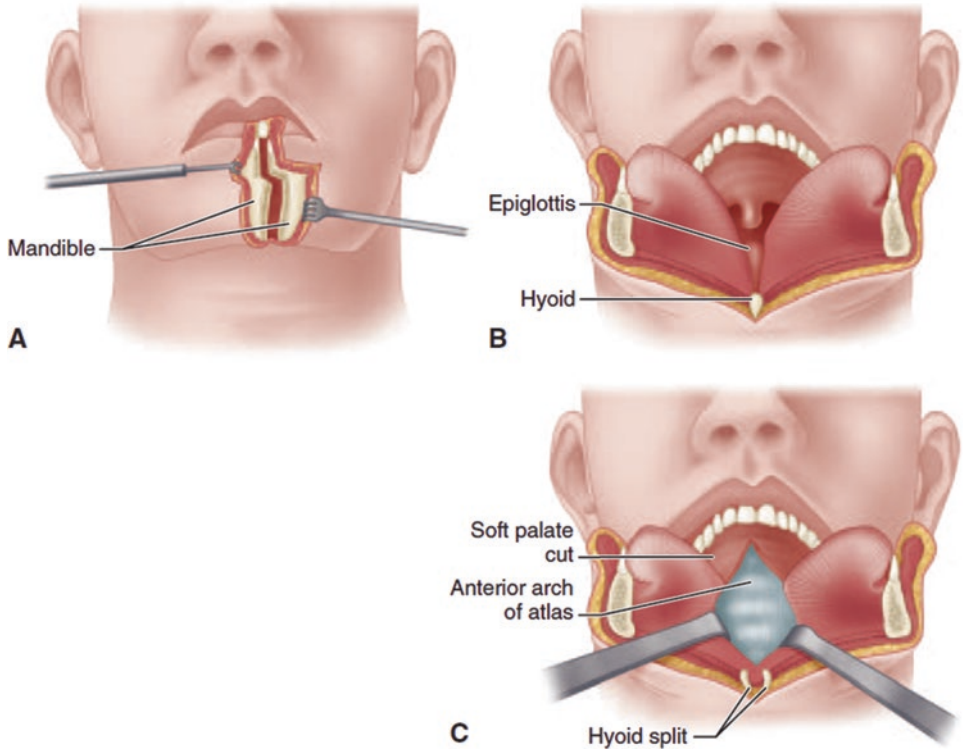

Fig. 14.3 (**a**–**c**) Mandible-splitting and tongue-splitting transoral approach (Reprinted from Warner [17]. With permission from Elsevier)

safe extension is marked, and this point should not be exceeded at any time during the operative procedure. A fiberoptic nasotracheal intubation is done with the patient under local anesthesia. When the airway has been secured, the patient is placed under general anesthesia. The patient's mouth must be kept free of all tubes to prevent any depression of the mandible inferiorly that may compromise the operative exposure. A modified transverse submandibular incision is made; the incision can be made on the right or left side depending on the surgeon's preference. As long as the dissection does not extend caudal to the fifth cervical vertebra, this exposure is sufficiently superior to the right recurrent laryngeal nerve to avoid damage to this structure. The incision is carried through the platysma muscle, and the skin and superficial fascia in the subplatysmal plane are mobilized. The marginal mandibular branch of the facial nerve is identified with the aid of a nerve stimulator and by ligating and dissecting the retromandibular veins superiorly. Branches of the mandibular nerves usually cross the retromandibular vein superficially and superiorly. By ligating this vein as it joins the internal jugular vein, and by keeping the dissection deep and inferior to the vein as the exposure is extended superiorly, the superficial branches of the facial nerve are protected. The anterior border of the sternocleidomastoid muscle is freed by longitudinally transecting the superficial layer of deep cervical fascia. The carotid sheath can be identified by palpation.

The submandibular salivary gland is resected, and its duct is sutured to prevent a salivary fistula. The posterior belly of the digastric muscle and the stylohyoid muscle is identified. The digastric tendon is divided and tagged for later repair. Division of the digastric and stylohyoid muscles allows mobilization of the hyoid bone and the hypopharynx medially. The hypoglossal nerve is freed from the base of the skull to the anterior border of the

hypoglossal muscle and retracted superiorly throughout the remainder of the procedure.

The dissection is continued between the carotid sheath laterally and the larynx and pharynx anteromedially. Beginning inferiorly and progressing superiorly, several arteries and veins may need to be ligated for exposure: the superior thyroid artery and vein, the lingual artery and vein, and the facial artery and vein.

The superior laryngeal nerve is freed from its origin near the nodose ganglion to its entrance into the larynx. The alar and prevertebral fascia are transected longitudinally to expose the longus colli muscles. Midline orientation can be confirmed by noting the attachment of the right and left longus colli muscles as they converge toward the anterior tubercle of the atlas. The longus colli muscles are detached from the anterior surface of the atlas and axis, and the anterior longitudinal ligament is divided to expose the anterior surface of the atlas and axis. The dissection should not be carried too far laterally to avoid damage to the vertebral artery.

Closure begins with approximation of the digastric tendon. Suction drains are placed in the retropharyngeal space and the subcutaneous space. The platysma and skin are sutured in the standard fashion. If the hypopharynx has been inadvertently entered, the anesthesiologist should insert a nasogastric tube intraoperatively, and the hole is closed in two layers with absorbable sutures. Parenteral antibiotics effective against anaerobic organisms should be added to the routine postoperative prophylactic antibiotics. The nasogastric tube is left in place for 7–10 days. Skull traction is maintained with the head elevated 30 degrees to reduce hypopharyngeal edema. Nasal intubation is maintained for 48 h. If extubation is not possible in 48–72 h, a tracheostomy can be performed (Fig. 14.4).

Cocke et al. [10] described an extended maxillotomy and subtotal maxillectomy for excision that gives excellent anterior exposure to the base of the skull and upper cervical spine. This approach, like the mandibular- and tongue-splitting approach described by Hall, should be used only in special circumstances.

Lateral Retropharyngeal Approach

The lateral retropharyngeal approach described by Southwick and Robinson [11, 12] is a modification of the classic approach of Henry to the vertebral artery. This approach exposes the anterior vertebral bodies from C3 to T1. In this approach, the sternocleidomastoid muscle is everted and retracted laterally. The remainder of the dissection follows a plane medial and posterior to the carotid sheath.

The recurrent laryngeal nerve is at risk during this approach to the anterior cervical spine. The right recurrent laryngeal nerve passes from lateral to medial in the lower part of the neck to reach the midline trachea, making it slightly more vulnerable during exposure than the left recurrent laryngeal nerve. This is why some surgeons prefer a left-sided approach.

With the patient supine, a small roll can be placed between the shoulder blades to ensure extension of the neck. Head-halter traction also can be used to aid exposure. Palpable anterior landmarks will aid in identifying the appropriate vertebral levels in the neck. The lower border of the mandible is at the level of C2-C3. The hyoid bone is located at the level of C3. C4-C5 is located at the level of the thyroid cartilage, and C6 is at the level of the cricoid cartilage and the carotid tubercle.

A transverse incision is more cosmetic, but a longitudinal incision along the anterior margin of the sternocleidomastoid muscle can be made if more extensile exposure is needed. The platysma muscle is divided to expose the anterior border of the sternocleidomastoid muscle. The fascia overlying the sternocleidomastoid muscle is divided, and the sternocleidomastoid muscle is retracted laterally to expose the carotid sheath that consists of the carotid artery, vein, and vagus nerve. A plane is developed between the medial edge of the carotid sheath and the midline structures. Retraction of the carotid sheath and the sternocleidomastoid muscle laterally, and the sternohyoid and sternothyroid muscle, with the trachea and underlying esophagus medially, exposes the longus colli muscles and the prevertebral fascia. After the vertebral level is identified, a

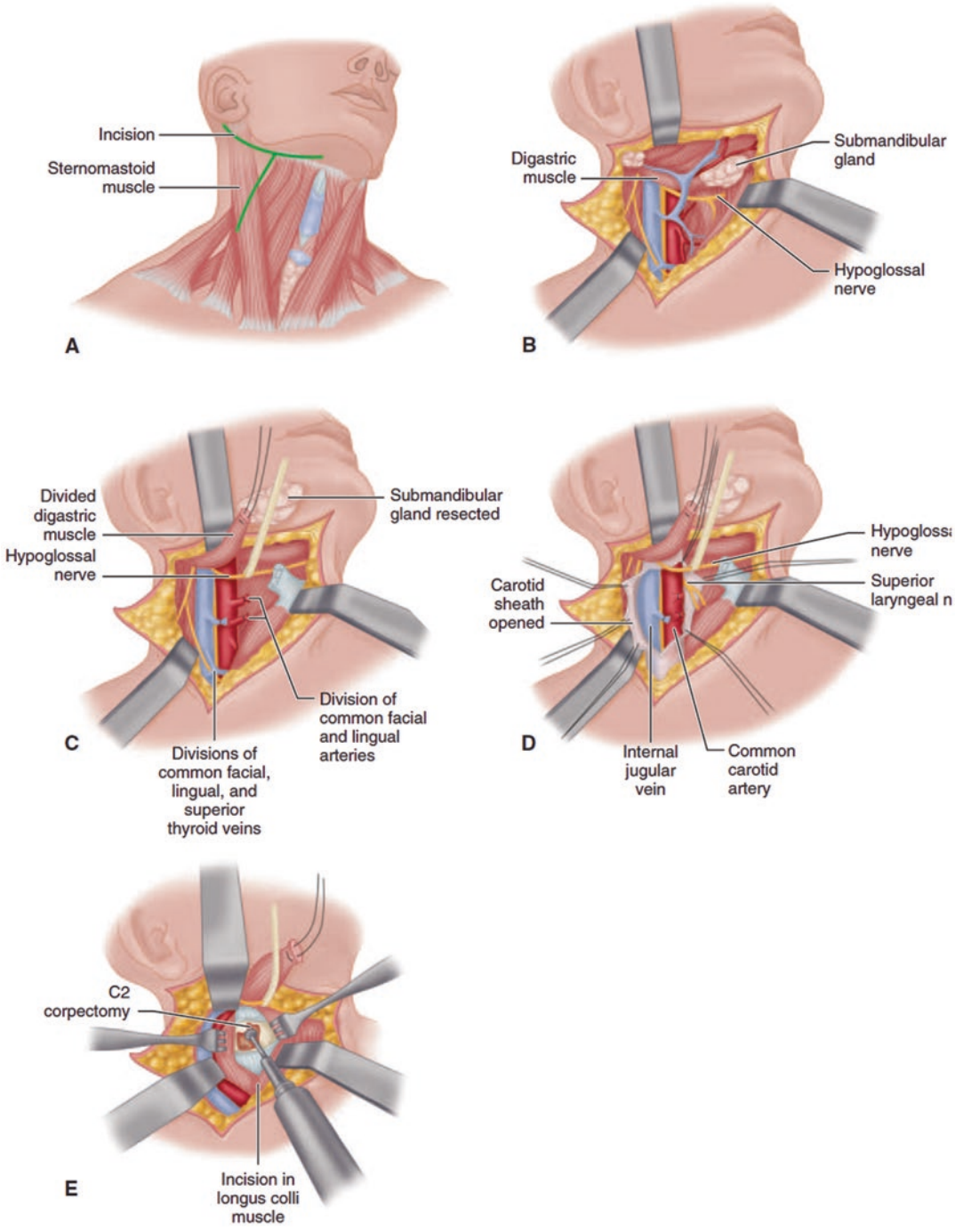

Fig. 14.4 Anterior retropharyngeal approach to the upper cervical spine. (**a**) Submandibular incision. Lower limb of incision is used only if midcervical vertebrae must be exposed. (**b** and **c**) Submandibular gland is resected, and digastric tendon is divided. Superior thyroid artery and vein also are divided. (**d**) Hypoglossal nerve and superior laryngeal nerve are mobilized. Contents of carotid sheath are mobilized laterally, and hypopharynx is mobilized medially. (**e**) Longus colli muscle is dissected laterally to expose the anterior aspect of the atlas and axis (Reprinted from Warner [17]. With permission from Elsevier)

longitudinal incision is made down to the bone through the anterior longitudinal ligament. A subperiosteal dissection is then performed to expose the vertebral bodies. At the conclusion of the procedure, the wound is irrigated and closed in layers over a suction drain in the retropharyngeal space (Fig. 14.5a, b).

Hodgson [13], Whitesides [14], and Henry [15] have described slightly different anterior approaches to the cervical spine. In Henry's classic approach and Whiteside's approach, the sternocleidomastoid muscle is retracted laterally and the carotid sheath medially with the plane of dissection between the sternocleidomastoid muscle and carotid sheath. Hodgson described a similar approach to the cervical spine that begins at the posterior border of the sternocleidomastoid muscles, with retraction of this muscle and the carotid sheath medially (Fig. 14.6a, b).

Sternal-Splitting Approach

Mirpuri et al. [16] described a sternal-splitting approach to the cervicothoracic junction that is useful for complex spinal deformities around the cervicothoracic junction in children. The approach requires the assistance of a cardiothoracic surgeon. A standard extensile anterior cervical spine approach is made incorporating an anterior sternal extension. The superior part of the exposure is a standard lateral retropharyngeal approach to the lower cervical spine using a longitudinal incision. The incision is continued along the medial border of the sternomastoid muscle, extending down to the sternal notch. The sternomastoid muscle is retracted laterally with the neurovascular sheath, including the carotid artery, the jugular vein, and the vagus nerve. Division of the omohyoid, sternohyoid, and sternothyroid muscles will facilitate this extensile exposure. The distal portion of the incision is extended as a midline sternotomy approach. The retrosternal soft tissues are mobilized by blunt digital dissection. The sternum is split with a sternal saw. Once the sternum is divided, the thymus gland is resected to provide additional exposure of the brachiocephalic trunk. At this point, the anterior cervical spine and upper thoracic spine can be accessed contiguously. If necessary, the pericardium can be opened to increase mobility of the brachiocephalic trunk; however, the dissection of the brachiocephalic trunk usually can be done down to the pericardial reflection without opening the pericardium. The trachea and esophagus are retracted away from the mid-

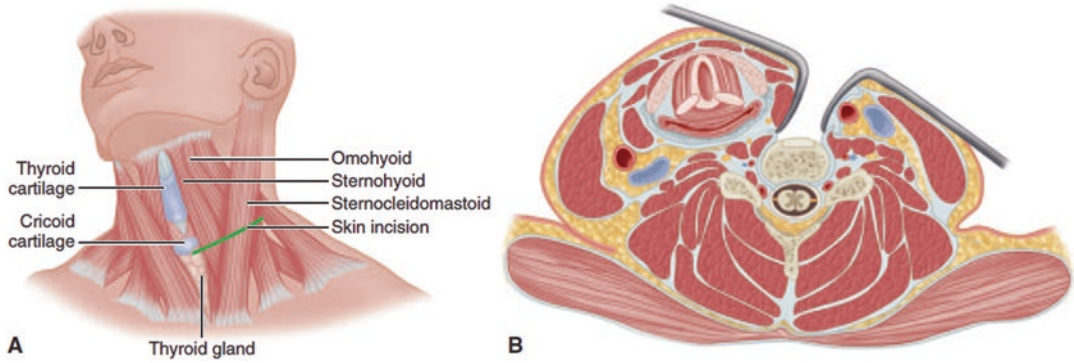

Fig. 14.5 Smith-Robinson approach. (**a**) Incision. (**b**) Thyroid gland, trachea, and esophagus have been retracted medially, and carotid sheath and its contents have been retracted laterally in the opposite direction (Reprinted from Wood [18]. With permission from Elsevier)

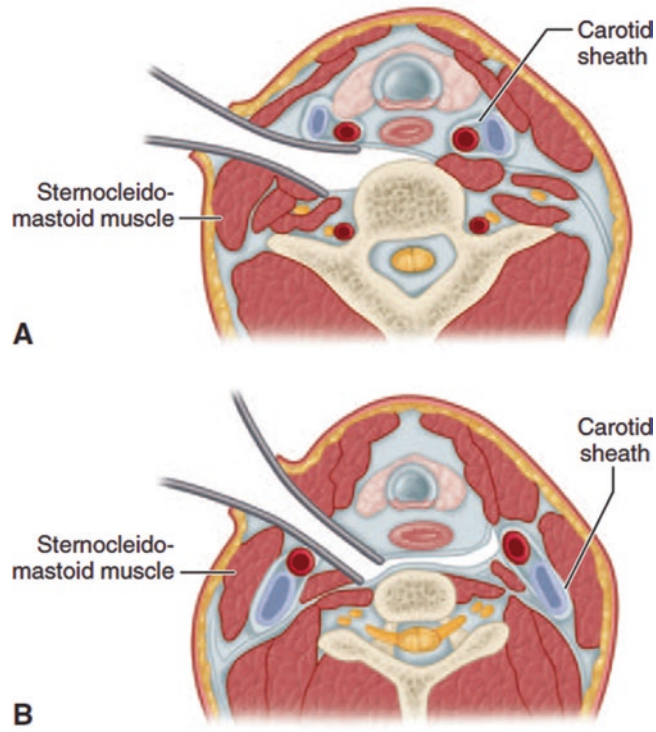

Fig. 14.6 (**a**) Whitesides and Kelly approach anterior to sternocleidomastoid muscle and posterior to carotid sheath. (**b**) Approach anterior to sternocleidomastoid muscle and anteromedial to carotid sheath (Reprinted from Warner [17]. With permission from Elsevier)

line with a right-angle retractor. Another right-angle retractor is placed under the innominate artery to pull it forward and downward as necessary to provide access to the lower cervical and upper thoracic spine. The distal extent of the exposure at this point depends on the patient's anatomy and deformity; in most patients, T4 can now be accessed and disc removal and instrumentation can be done safely. Aggressive distal exposure places the recurrent laryngeal nerve under traction and must be done carefully. Although left-sided anterior cervical approaches typically are preferred because of the distal course of the recurrent laryngeal nerve on that side, Mulpuri et al. used a right-sided approach with mobilization of the brachiocephalic trunk because medial displacement of the trunk exposes more segments of the thoracic spine on the right side. After completion of the orthopedic procedure, the sternum is approximated with wires or sutures, depending on the age of the child. The sternothyroid and omohyoid muscles are reattached, and the neck is closed in the usual fashion (Fig. 14.7a–d).

Posterior Approach Cervical Spine

The posterior surgical approach to the child's cervical spine can be extremely challenging. There are anatomic and pathologic features of the pediatric spine that make it slightly different from the adult cervical spine. Naturally the cervical spine in children is smaller, the vertebrae can be largely cartilaginous, congenital abnormalities can be present, and bone dysplasias can make the anatomy look quite peculiar. The head is disproportionately large compared to the rest of the body, making positioning of the child for the exposure of the upper cervical spine more difficult.

There is a wide range of spinal pathologies in the pediatric population that must be appreciated before embarking on exposure of the cervical spine. Many of the deformities are syndromic, and there are important anesthetic considerations that must be considered. Thorough preoperative evaluation of these patients is essential to maximize perioperative care. Imaging of the cervical spine preoperatively can help greatly. A fine cut (2 mm) CT scan with 3-D reconstruction can

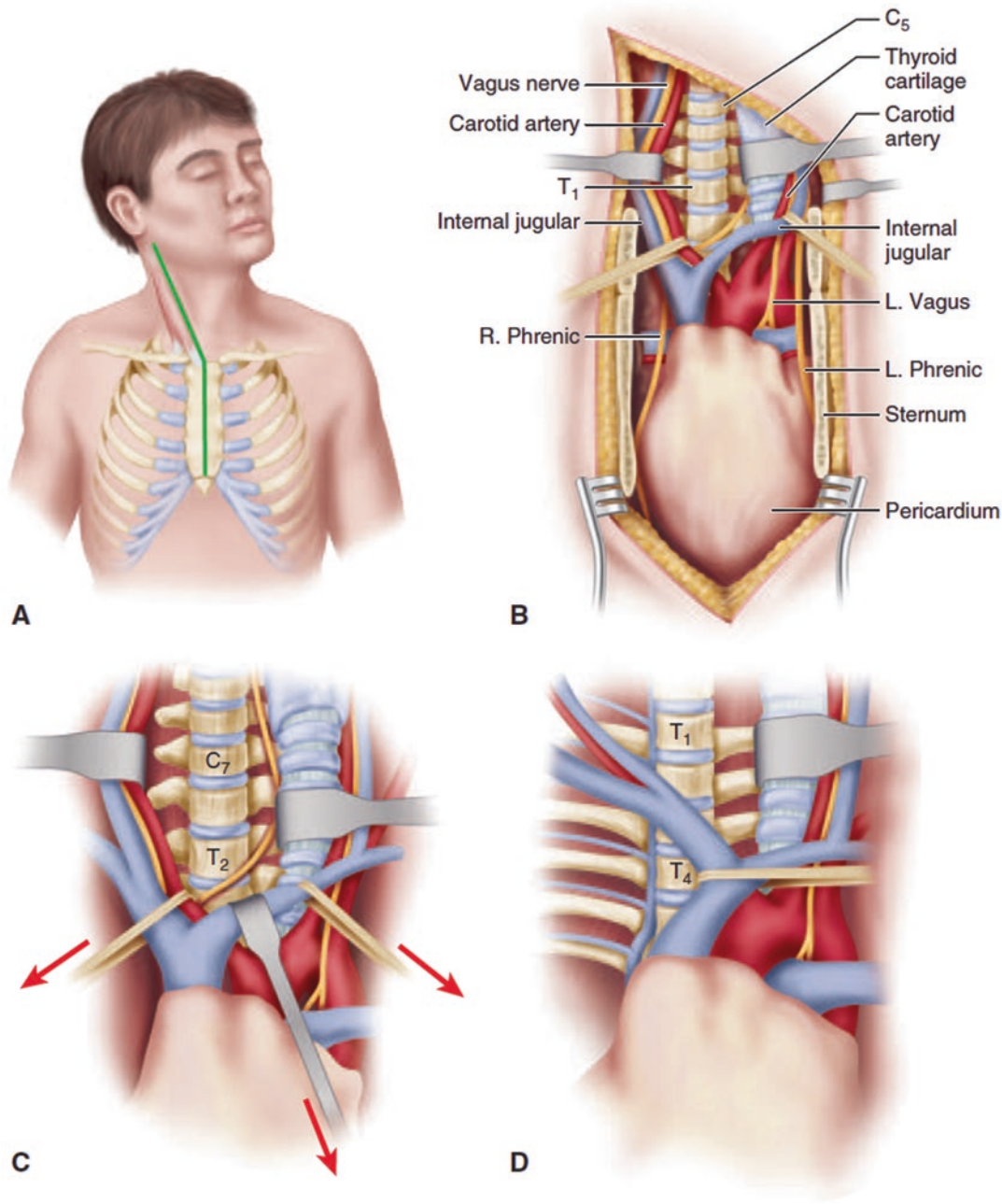

Fig. 14.7 Mulpuri et al. sternal-splitting approach to the cervicothoracic junction. (**a**) Incision for right-sided approach. (**b**) Sternum is opened, thymus gland is resected, and brachiocephalic trunk is mobilized to allow contiguous access to anterior cervical spine and upper thoracic spine. (**c**) Retraction of trachea, esophagus, and innominate artery provides access to lower cervical spine and upper thoracic spine. (**d**) Medial displacement of brachiocephalic trunk allows more distal access to thoracic spine (Reprinted from Warner [17]. With permission from Elsevier)

help define the bony architecture accurately (Fig. 14.8a, b). The CT can identify bifid lamina, aberrant foramen transversarium, congenital fusions, rotational abnormalities, vertebral subluxation, thin lamina, depth of the occipital bone, and angles of pedicles and facet joints. MRI scans are more useful for looking at the soft tissues, especially the space available for the spinal cord (SAC). Flexion/extension MRI scans can define the best position to put the neck in to minimize cord compression if instability and stenosis is present. CT or MRI angiography is also very useful to define vertebral artery (VA) position in patients with rotatory subluxation, congenital abnormalities, and bone dysplasia patients.

Exposure is such an important part of any surgical procedure. Meticulous handling of tissues and precise positioning of retractors allows the surgeon to safely proceed with surgical procedures and internal fixation in challenging, often life-threatening conditions of the pediatric cervical spine. Positioning of the patient is crucial, the anesthetic conditions must be optimal, the correct surgical equipment immediately available, and a headlight and loupe magnification aid in maximizing visualization. All these factors need to be considered and brought together to execute the best operation. Sometimes it may be helpful to have a colleague assist, especially when some difficult intraoperative decision-making is required. Likewise, a neurosurgeon can bring a whole different surgical skill set that may be useful in more unusual cases, especially when the spinal cord or nerve roots are compressed.

Positioning

When neuromonitoring is being used, this should be attached prior to positioning the patient and baseline recordings made. Neuromonitoring is paramount in order to minimize any risk of neurologic injury. This then allows immediate feedback of neurologic change once the child is turned prone. Positioning the child can be difficult: however, it is an extremely important initial step in the surgical procedure. It is the surgeon's responsibility to organize the positioning of all the personnel within the operating room. This includes the anesthesiologist, surgical assistants, scrub nurse, neuromonitoring technician, and radiology technician. Space around the pediatric spine is very limited, and maximizing efficiency, especially with the fluoroscopic unit, is helpful prior to making an incision.

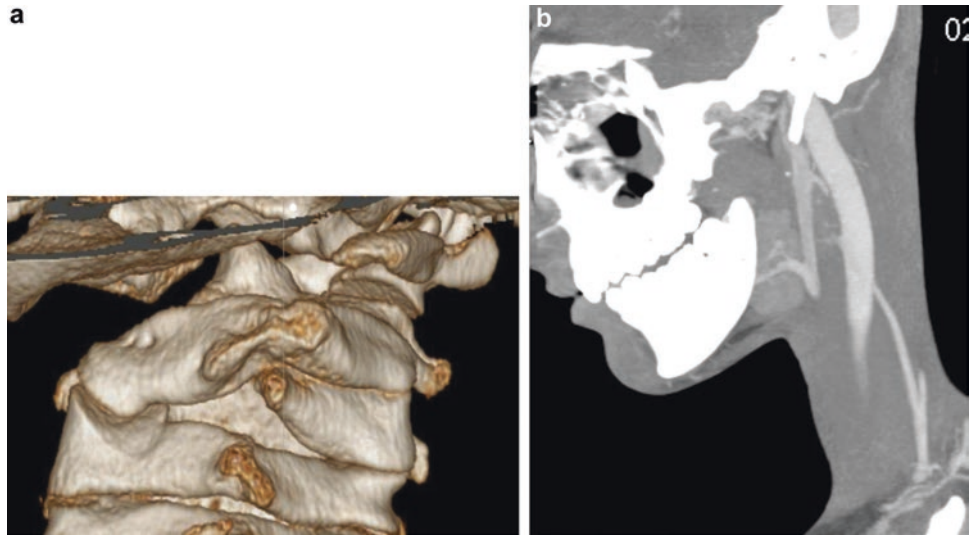

Fig. 14.8 (**a**) Three-dimensional CT scan of a 12-year-old boy with a hemivertebrae of C1 and C3 and painful torticollis. (**b**) A CT angiogram can be performed at the same time to assess vertebral artery course and size

The head needs to be held securely with either a Mayfield clamp (Integra Corporation) or a halo (Fig. 14.9a–c). The Mayfield clamp should not be used in children younger than 5 years of age and when the skull bones are too thin and soft. These younger children should have a halo fitted with multiple (eight or more) pins, which should be placed only to finger-tight pressure. These are then attached to the operating table, and the head and neck positioned in the correct position. This position is particularly important when a cervical fusion is being performed. An alternative is to place the child in a halo crown and apply the vest component, which then can be used to safely turn the patient prone with the back part of the vest then being removed for surgery. This is a very safe way to position the patient, especially in cases of instability or pre-existing spinal cord deficit.

There are three components to the positioning of the head in relation to the body: the sagittal plane (flexion and extension), the coronal place (sideways tilt), and rotation. All of these aspects must be considered with final tightening of the external holding device. In practical terms, the child's ear should line up with the shoulder tip, the eyes should point directly forward, and there should be no lateral tilt of the head or neck. Preoperative imaging, especially the flexion/extension MRI scan, may help with positioning the neck in the position where the spinal cord is least compressed. The Mayfield holder does allow the position of the neck to be manipulated intraoperatively. This can be useful in upper cervical fusions, when the head can be flexed slightly to allow easier exposure of the upper cervical vertebrae and occiput. Fluoroscopy or plain radiographs should be used to confirm the correct alignment of the head and neck before proceeding with surgery. When adjusting the Mayfield frame, it is best to have the surgeon hold the head and the assistant loosen all the tightening apparatus, so the head and neck can be positioned appropriately. Releasing only part of the securing device makes manipulating the head more difficult.

The child's head is best held securely with a Mayfield clamp or halo. The Mayfield clamp is a temporary device for children over 5 years of age. It is applied in the "sweat band" region of the skull caudal to the greatest diameter. This helps prevent slippage of the device intraoperatively. The pins should be placed to avoid the temporalis muscle. The amount of torque applied to the clamp depends on the age of the child and the strength of the bones. In an adult or adolescent patient with good bone stock, 60 in. lbs. can be applied; however, this may need to be less in younger children. It is important to assess the stability of the clamp once it has been tightened to make sure it will not slip during the surgical procedure. If the intention of the surgeon is to use a halo vest or cast postoperatively for immobilization, then it is advisable to apply the halo while the child is still supine and before turning them prone for positioning. A crown halo is prefer-

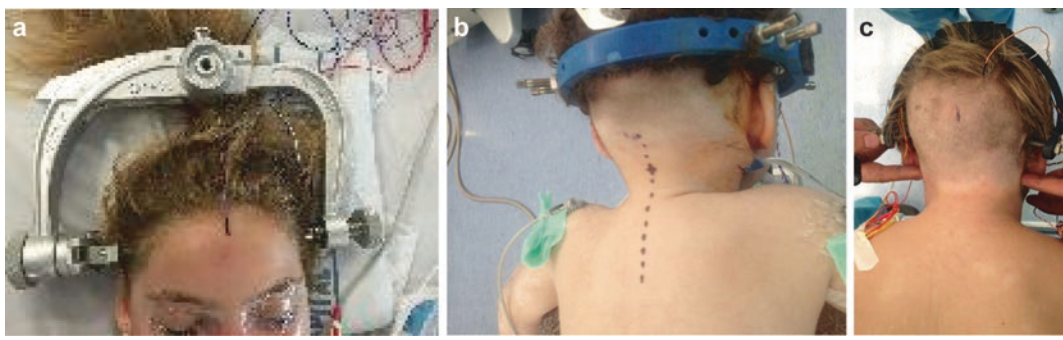

Fig. 14.9 (**a**) The Mayfield Clamp applied in the "sweat band" area, avoiding the temporalis muscle. (**b**) A halo ring with eight finger-tight screws on a 21-month-old child with Larsen's disease. This was converted into a halo cast jacket after the cervical decompression and fusion for the kyphotic deformity. (**c**) A halo crown on a 12-year-old boy with congenital torticollis. Note the excellent exposure possible with a halo crown. Placing your fingers on the mastoid process helps assess rotational alignment

able to a ring halo, as it allows a greater area of skin to be exposed and provides easier access, especially to the upper cervical spine (Fig. 14.9). For a child younger than 6 years of age, 10–12 screws are often required to achieve adequate halo fixation. The pins should only be finger tightened or no more than 2 in. lbs. in infants and young children. Over the age of 8 years, the child can be treated like an adult unless there is unusually soft or thin bone. Occasionally traction can be applied to the skull intraoperatively to help with reduction of the cervical spine. One has to be very careful when using this technique, as the head is more unstable than when it is held rigidly. Exposure can be made easier by the traction; however, the head and spine is more mobile, and great care must be taken with exposure, especially when pressing on the lamina and lateral processes with periosteal elevators and curettes.

In children who require an occiput-thoracic fusion or children who have significant global sagittal plane deformity and require a significant occipital-cervical fusion, then trialing the position of the head may prevent placing them in an untoward position at fusion. This may be done by placing them preoperatively in a halo crown vest in the desired position and making sure this is clinically functional by trialing them in that position for a period of time preoperatively. If the position is well tolerated by the patient, then at the time of the fusion you may place them prone in that locked position and remove the posterior vest while still assuring correct fusion alignment.

Posterior Approach to the Craniocervical Junction

The child is intubated and all cannula are secured to the patient before turning prone. Spinal cord monitoring electrodes are placed and a baseline reading recorded before moving the patient. The halo is securely applied and then the patient is turned prone. The halo is then attached to the holder, and the patient's head is supported on the operating bed with a gel horseshoe head support. The optimal head and neck position is assessed clinically and then confirmed radiographically and any adjustments made. The neuromonitoring is then further checked before proceeding with surgery to confirm no neurologic deterioration has occurred with positioning the patient.

The posterior scalp and neck are prepared along with the bone graft harvest site. Even if autologous bone graft use is not anticipated, it is advisable to prepare the site in case an unexpected need arises. It is easier to prepare access to either a rib donor site or posterior iliac crest preoperatively. The posterior scalp is shaved with an electric razor to avoid the microtrauma seen with standard razor blades and the area thoroughly cleaned with chlorohexidine and betadine wash. The patient is prepped and draped, ensuring the edges of the drapes are securely fastened to the skin with opsite, surgical clips, or sutures. Prior to prepping, cleaning the edges of the drapes at the skin-drape interface with an alcohol-soap scrub is advisable to further diminish chances of infection.

A midline incision is made from the external occipital protuberance to the midcervical region. A longer incision than anticipated is usually required. The dermal layer and subcutaneous tissue are then infiltrated with lidocaine and epinephrine to help with hemostasis. Sharp dissection or electrocautery is then used through the subcutaneous tissue and deep fascia to the median raphe. The occiput bone is exposed relatively easily with subperiosteal dissection. Bleeding from the multiple small emissary veins can be controlled with bone wax. Exposure of the atlas and axis is occasionally difficult, due to the hypermobility of the cervical spine. Stabilization to make this dissection easier can be achieved by passing a sharp towel clip through the lamina of C2 and having an assistant hold it. Also, placing two deep Mayo retractors at either end of the incision helps by tensioning the soft tissues, which indirectly stabilizes the spinal column.

Exposure of the foramen magnum may be necessary if suboccipital wires are going to be passed through the occiput. Likewise, good visualization is required if a foramen magnum decompression is required. In some conditions (achondroplasia, osteogenesis imperfect) there may be invagination of the occipital bone, and the foramen magnum can only be exposed after a

large piece of occipital bone has been drilled out. On other occasions, the posterior part of the ring of C1 has to be excised to gain access to the foramen magnum. The foramen can then be further exposed by elevating the dura off the anterior surface of the occipital bone using a combination of Penfield elevators and angled curettes. Occasionally there is a vestigial venous sinus in the dural leaves at the level of the foramen magnum which, if torn, can be controlled with compression. The bone can be removed from the foramen magnum with kerrison rongeurs once the dura has been elevated. All locally harvested bone should be kept for latter grafting.

Dissection of C1 vertebra is probably the most difficult, especially if the entry point for C1 lateral mass screws is being exposed. The posterior arch of C1 sits a variable distance anterior to the C2 lamina. Slight flexion and traction of the head can aid in the exposure of the C1 vertebra. The vertebral arteries (VA) also run in the foramen transversarium groove on the ventral surface of the posterior arch. The medial edge of this grove is an important anatomical landmark, as dissection lateral to it can result in perforation of the vertebral artery. The medial aspect of the VA is approximately 1.5 cm from the midline in adults and can be as close as 1 cm in children. The VA courses from this lateral position in front of the C1 arch to the midline when they enter the foramen magnum. There is often a small tubercle at the most posterior aspect of the C1 ring that helps in identifying the midline. The vertebral artery lies in the cranial surface of the C1 posterior ring, and as dissection proceeds laterally, care must be taken to avoid dissecting cranially. The lamina is often very thin and soft, so dissection must be carried out delicately. Too much pressure with a dissector can fracture the lamina and damage the spinal cord. Finger dissection can be used to sweep the soft tissues laterally off the C1 lamina safely (Fig. 14.10).

If sublaminar wires are to be passed at C1, the most posterior arch can be exposed subperiosteally with elevators and curettes, and the ligamentum flavum does not need to be excised. Below C2, a small window is made in the ligamentum flavum to allow the passage of cerclage wires, if required. The ligamentum is best opened with a scalpel and then a series of kerrison rongeurs to widen the hole.

Exposure of the C1-C2 facets joints is another area where the VA is at risk to injury. The VA runs between the foramen transversarium of C2 and C1 immediately lateral to the C1-C2 facet joints. Exposure of the C1-C2 joint is important both for the safe excision of the facet joint cartilage and also the accurate insertion of the C1 and C2 screws. Additional to the VA being immediately lateral to the facet joint, the C2 nerve root and its ganglion traverse the facet capsule (Fig. 14.11). The main branch of this posterior nerve root is the greater occipital nerve, and lacerating it causes a loss of sensation in the occipital region. Painful neuromas can also occur. Preserving the nerve and ganglion is ideal; however, resecting it is also acceptable if safe exposure cannot be achieved by just retracting it. At the dorsal aspect of the C2 lamina, the C2 pars, and C1-C2 facet joint is a venous plexus that can cause considerable bleeding. Often confused with VA hemorrhage, this darker lower pressure bleeding is usually controlled with diathermy, Gelfoam (Pharmacia and Upjohn), and cotton patties. It is useful to sometimes pack one

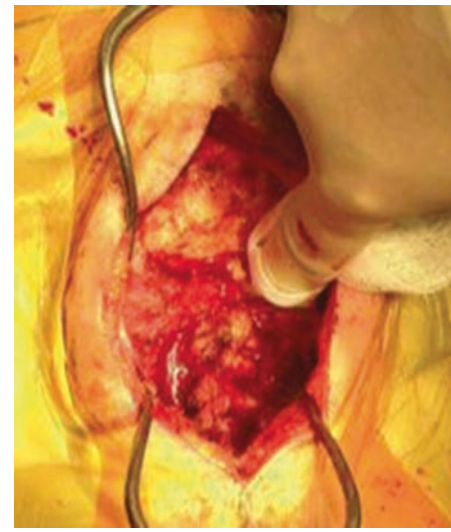

Fig. 14.10 Finger dissection of the soft tissue from the posterior aspect of C1 is an effective and safe way of exposing the lamina

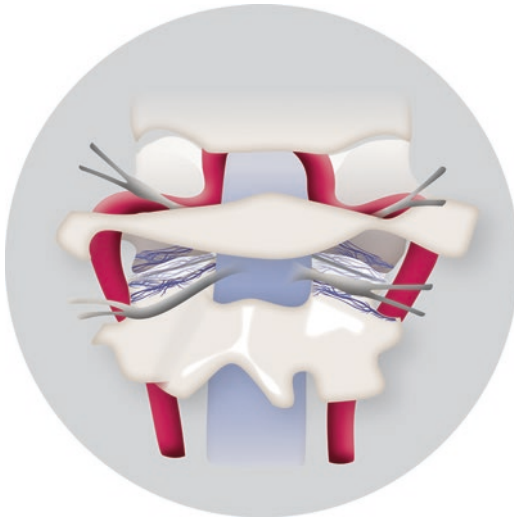

Fig. 14.11 The upper cervical spine showing the tortuous course of the vertebral arteries. Note the venous plexus around the C2 nerve roots and ganglion. The C2 nerve root and venous plexus lie over the C1-C2 facet joint

side if profuse bleeding is making further exposure difficult and start dissecting the opposite C1-C2 facet joint. IsoCool (Codman) diathermy tips that minimize tissue sticking to the end of the forceps are very useful in dissecting the venous plexus and achieving hemostasis. Repeated pinching and diatherming the venous tissue with the sharp-pointed forceps can be rewarding. This is best done prophylactically prior to any bleeding by judicious and meticulous diathermy of any venous structures. Once hemostasis is achieved, the exposure needs to allow the positioning of three retractors. A blunt Penfield or MacDonald retractor is placed lateral to the facet joint protecting the VA. A small nerve hook retracts the posterior C2 nerve root inferiorly, and a blunt retractor is placed medial to the facet joint to protect the dura. This allows adequate exposure of the joint for resection of the articular cartilage and the safe passage of instrumentation.

In congenital abnormalities, bone dysplasias, and rotatory subluxation, this upper cervical anatomy can be very distorted, and a preoperative CT as well as occasional MRI with angiography is important in defining the anatomy before the surgical exposure.

C2 usually has a bifid spinous process and a relatively thick lamina. Subperiosteal dissection is aided by stabilizing the C2 vertebra with a pointed towel clip in the lamina. Either the surgeon or assistant can hold this as the ligaments and muscles are dissected subperiosteally.

Subaxial Exposure

After prone positioning and skin and drape preparation as described above is performed, a midline incision is made from the base of the occiput superiorly to C7 inferiorly. Local anesthetic and epinephrine is infiltrated into the subcutaneous tissue to aid hemostasis. A scalpel or electrocautery is then used to dissect down to the avascular median raphe. This raphe is not as well defined as in the thoracic and lumbar spine. Its course can also be more tortuous in the cervical spine, and care needs to be taken to follow it closely, as deviation into the paraspinal muscles can lead to heavy bleeding. Once the spinous processes are exposed, a pointed towel clip or metal hub needle can be inserted into the lamina and fluoroscopy used to check the level. Care must be taken to expose only the levels required, as excessive exposure will lead to spontaneous fusion above and below the desired area.

Subperiosteal dissection is carried out on both sides of the lower cervical spine down to the base of the lamina. In infants and children with soft bone, this is more easily performed with electrocautery rather than periosteal elevators. A fine-tipped diathermy is more effective on the soft cartilaginous bone and puts a lot less pressure on the cervical spine. The interlamina distance is disproportionately wider in children compared to adults, so care must be taken to avoid inadvertently entering the spinal canal. Be careful to recognize any bifid lamina, which has hopefully been visualized on preoperative imaging. Failure to do so can lead to inadvertently entering the spinal canal and potentially damaging the spinal cord. Dissection is carried out to the lateral margin of the facet joints. A narrow, smooth dissector can be placed beside the lateral mass to protect the VA when the facet joint is

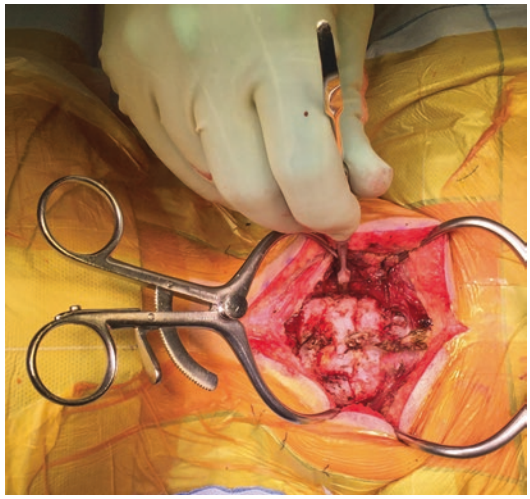

Fig. 14.12 Subaxial dissection of the cervical spine. A smooth dissector can be placed beside the lateral processes and facet joints to protect the soft tissues

excised (Fig. 14.12). The facet joint capsules are easily removed for clear visualization of the articular cartilage. This cartilage can be removed with the drill, burr, or curettes to help with a fusion. A smooth dissector can be inserted into the facet joint to help with orientation when inserting lateral mass screws.

Summary

Surgical treatment of pediatric cervical spine problems may be undertaken from an anterior, posterior, or combined approach. Surgical success is dependent on safe and effective exposure, which allows for adequate visualization of the anatomy. Knowledge of the surgical anatomy allows for the successful implementation of decompression, instrumentation, and fusion.

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Spinal Cord Monitoring in Pediatric Cervical Spine Surgery

15

Jonathan H. Phillips and Michael Isley

Introduction

Since the efforts by Nash and Brown using cortical-evoked potentials as an approach to spinal cord monitoring during scoliosis surgery, the role of sensory and motor, intraoperative, and neurophysiological monitoring of spinal cord physiology at multiple, neural levels has become an established routine [1]. It was Nuwer's study based on a SRS database that first addressed the overall benefit in reducing postoperative neurological deficits by monitoring somatosensory-evoked potentials (SSEPs) during scoliosis surgery [2]. This profoundly shaped and helped ensure the widespread acceptance and proliferation of this neuromonitoring concept. However, the technological advances that have accrued still beg the question of what constitutes significant alarm criteria for predicting postoperative neurological deficits, which surgical cases warrant monitoring, and how lessons learned from one patient age group apply to another. There is a dearth of research data regarding spinal cord monitoring in children's cervical spine surgery. By contrast, in the arena of adult cervical spine surgery, numerous studies have been generated regarding the routine use of neuromonitoring. However, agreement is not universally supported by the peer-reviewed literature about these techniques. Thus, what follows is, of necessity, a review of the principles and concepts that need further study in this very specialized area of pediatric cervical spine surgery.

Relative Importance of Sensory and Motor-Evoked Potential Monitoring and Electromyography

Alarm criteria for changes in waveform morphology developed for intraoperative intervention, as well as the techniques for elicitation, recording, and measurement, are well established. As described above, the first widely accepted technique for evaluating spinal cord function during spinal surgery and purports to assess incipient injury was that of monitoring SSEPs. SSEPs do, however, have some known limitations involving "alleged," false-negative results (e.g., selective, anterior, spinal cord injury can occur with persistently normal SSEP recordings) [3, 4]. However, many do not regard this as a false-negative result

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since SSEPs directly monitor dorsal column function and only infer, at best, the function of motor pathways. Indeed, motor-evoked potentials (MEPs) are the primary neuromonitoring modality for assessing the likelihood of impending injury to the motor tracts arguably perhaps, a more dismal outcome compared to sensory deficits. However, the inherent polyphasic and often capricious nature related to the morphology of these neural signals, particularly in the face of volatile anesthetic agents [5] and blood pressure changes, has made a consensus position on significant alarm criteria more difficult to reach.

Nevertheless, a thorough understanding of the current thinking in this area is germane to surgery of the child's cervical spine and generalizable to any children's spinal surgery. The subtleties of monitoring technique, the various thresholds for alarm, and the evolution of thinking in this area are justifiably important. In particular, both SSEPs and transcranial MEPs have become common neuromonitoring modalities for spinal deformity surgery, in part, because they can potentially monitor the most rostral extent of the spinal cord, an area frequently visited by the surgeon involved with cervical surgery. Another technique of neuromonitoring, which has gained popularity in cervical surgery, is the use of free run and triggered electromyography (EMG) such as pedicle screw stimulation technique.

Definitions

The spinal cord constitutes a contiguous array of nervous tissue extending from the medulla oblongata above the foramen magnum to the conus medullaris, usually at the L1 or L2 level. From here it flares into a "brush" as the cauda equina of lower nerve roots. The neural pathways of the spinal cord are characterized by multiple tracts with various sensory, reflex, and motor functions. Traditional anatomical descriptions divide these spinal tracts into primarily anterior and posterior elements, although this is an oversimplification. The primary motor tracts are those located in the anterolateral corticospinal tracts, while the primary sensory tracts are located in the dorsal columns. It is these two pathways, functionally if not precisely anatomically, which are the focus of intraoperative spinal cord monitoring.

Somatosensory-Evoked Potentials

SSEPs have for about 20 years been the first standard in spinal cord monitoring for deformity surgery of the spine. Other surgeries (including aortic aneurysm) have benefitted from their use, but this discussion will center on spinal surgery. The name implies that a stimulus is necessary for their elicitation, and, indeed, it is the nature of this stimulus applied over time which results in one drawback to this technique.

The stimulus is applied via transcutaneous electrodes placed at standard locations in the lower and upper extremities. Typically, a bipolar square wave stimulus is applied for 100–300 microseconds, sufficient to elicit a visible twitch in local muscle groups. The tibial and median nerves are usually stimulated with two electrodes about 2 cm apart. A constant train of electrical stimuli is applied to these electrodes, and ascending impulses are detected at standard positions proximally (subcortical responses, cortical responses) by reading electrodes. Several standard recording positions on the scalp (e.g., N25, P60, N80) are monitored.

A computerized algorithm averages the values of the waveforms over many minutes, and the signals are displayed on a computer screen showing the composite of these impulses. Many thousands of waves are averaged over many minutes. Thus, if a deficit of function occurs, it is not seen in real time but may appear long after the inciting event. This can make it difficult to decide which, if any, maneuver of spinal deformity correction is responsible for the observed change, and a stepwise reversal of surgical manipulations may be necessary for the recovery of the responses, which may in turn take many minutes to reverse, further disguising the etiology of the observed electrical event. In addition, this technique represents a more whole-spine overview of function, with less anatomical localization than other modalities. Monitoring of many levels cortically

is possible, but stimulation of a few levels distally is the norm. Combining these signals with motor-evoked potentials is a standard technique, but limitation in the number of channels available on the monitoring equipment limits practically the number of anatomical points which are monitored, especially if pedicle stimulation EMG is also being evaluated during the surgery.

Motor-Evoked Potentials

Like SSEPs, MEPs are elicited waveforms to an inciting electrical event. Initially stimulated at the spinal level, the technique has evolved to include a motor cortex level of surveillance, which allows one to distinguish between limbs, which was not afforded by spinal cord stimulation. The stimulus is applied between two scalp electrodes over the motor cortex and the waveforms recorded by intramuscular electrodes in upper and lower extremity muscles. Typical muscle groups monitored include the abductor pollicis brevis for the upper extremities, the medial gastrocnemius, and the abductor hallucis brevis for the lower extremities. One major difference between MEPs and SSEPs is that the former are a single event created by a non-summated technique which has to be triggered after critical events in the surgical procedure whereas the latter are an ongoing averaged electrical paradigm running more or less continuously in the background. An analogy might be the difference between a ship's radar set to constantly scan the surroundings compared to the sonar of a submarine which needs to be made to elicit a "ping" to get an echo response from its target. Both instruments can be turned on and off, and both need skilled interpretation of the waveforms on the screen, but the sonar is only activated as needed to avoid revealing the submarine's location.

Comparatively, stimulation of motor cortex by an electrical shock is also limited, for reasons of concern for destabilizing possible epileptic foci, a relatively rare event (<0.03% chance of inducing a seizure). Typically, a train stimulus of 5–9 at an intensity of a few hundred volts is applied for 50–75 milliseconds. The compound muscle action potentials elicited by such techniques may range from simple, biphasic responses recorded from the most distal, fine muscle groups to very complex, polyphasic potentials recorded from the more proximal, superficial larger muscle groups. The alarm criteria for intervention are discussed below.

Alarm Criteria

The criteria for concern in SSEP monitoring are well established—the "50/10" rule which was recently reviewed in the American Society for Neurophysiological Monitoring Position Statement. Simply stated, a > 50% decrease in amplitude and a > 10% increase in latency of the subcortical and/or cortical waveforms are considered a potentially clinically significant change. However, any persistent and distinct change in waveform morphology from established baselines should always be reported immediately to the surgical team. Of concern at times is elucidating the etiology of such changes, e.g., a particular mechanical maneuver, a slow decrease in hematocrit, or an unstable blood pressure. The response checklist for the alarm is discussed later, but the dilemma in SSEP monitoring is the averaging over time and how the alarm relates to the steps in children's spinal deformity correction.

Several alarm criteria have been suggested for MEPs, although there is currently no consensus. The various criteria have included the following:

1. The amplitude criterion which involves the application of the presence or absence ("all-or-none") criterion of abMEP waveform or decreases in amplitude of greater than 80% in one or more muscle groups [6].
2. The threshold level criterion, i.e., ≥100 V increase over baseline thresholds for eliciting MEPs for a period of 1 h [7].
3. The waveform criterion identifies significant changes in morphology, e.g., a change from

multiphasic to biphasic morphology along with changes in voltage threshold [8].

Our interpretation of normal versus abnormal will depend primarily on the absence or presence of waveforms, the all-or-none criteria. This is the only criterion for which there seems to be a general consensus [9]. However, any persistent change from baselines should be immediately reported to the surgical team.

Contraindications for transcranial MEPs may include recent onset of epilepsy, cortical lesions, skull defects, intracranial electrodes, pacemakers, and other implanted bioelectric devices. The estimated incidence of a seizure with pulse train stimulation is 0.03% [10]. Lastly, great care should be taken to appropriately place soft bite blocks bilaterally, since jaw contractions during stimulation may cause oral trauma such as tongue bites, lip injuries, and/or cheek lacerations (0.63%) [11]. Appropriate placement of the bite blocks should prevent the teeth from being able to close on the tongue, and the block is secured so as to minimize the chance of shifting.

As far as MEPs are concerned, however, providing that appropriate events are recognized and a stimulus elicited, each step in the surgery can be checked for safety. Changes in the motor responses typically take only a few minutes and can be more confidently related to the surgical events. The dilemma with this technique is one of consensus in what constitutes a significant alert. An amplitude decrease of 70–80% is typically thought to be significant. Some believe that an all-or-nothing approach is appropriate, and this standpoint is further complicated by opinions which attribute weight to the number of different muscle groups that lose responsiveness. In both these approaches, the question remains what part of the polyphasic motor waveform to measure. Is it the maximal peak-to-peak deflection, or should the total area under the curve be summated? We noted above the school of thought that attributes an increase in the needed voltage to elicit responses distally. An increase of 100 volts sustained for an hour or more was proposed by Calancie to be significant [8]. This clearly has the limitations of late reporting of a potentially disastrous event. Finally, the waveform criteria are thought by some to have significance, including a reduction in the complexity and length of the response recorded. An excellent review of these concepts is provided by DD Langeloo et al. [9].

The preceding discussion underlines the great complexity of both sensory and motor monitoring in spinal surgery. It is, however, incumbent upon a surgeon embarking upon a complex cervical spine operation to have a good understanding of the parameters which are being reported to him by his neurophysiological monitoring team.

Here we illustrate an example of highly significant intraoperative alarm criteria in a 6-year-old female with duplication of chromosome 1 (Figs. 15.1, 15.2, 15.3, and 15.4) . She had undergone several uncomplicated lengthenings of a vertical expandable prosthetic titanium rib (VEPTR) construct placed for severe early-onset scoliosis (EOS). The remaining details are in the legends of the figures. Ultimately, a posterior occipito-cervical fusion was necessary to stabilize her cervical spinal cord to allow further treatment of her subjacent progressive scoliosis (Fig. 15.5).

Response Strategies to Alarm Criteria

As evidence-based level 1–2 studies become more frequent in the literature, we are able to more reliably make decisions regarding therapeutic safety. Ethical dilemmas, however, preclude the true randomized trial of spinal cord monitoring alert responses, since allowing a patient to suffer neurological injury whose alarm criteria are met intraoperatively is untenable. Thus, retrospective and animal studies and surgeon experience are the best we have to rely on. This constitutes level 4 and 5 evidence, hardly reassuring to the patient undergoing complex spinal surgery. There are several of these studies recently reported [12–14], but these, though large in number of patients undergoing cervical

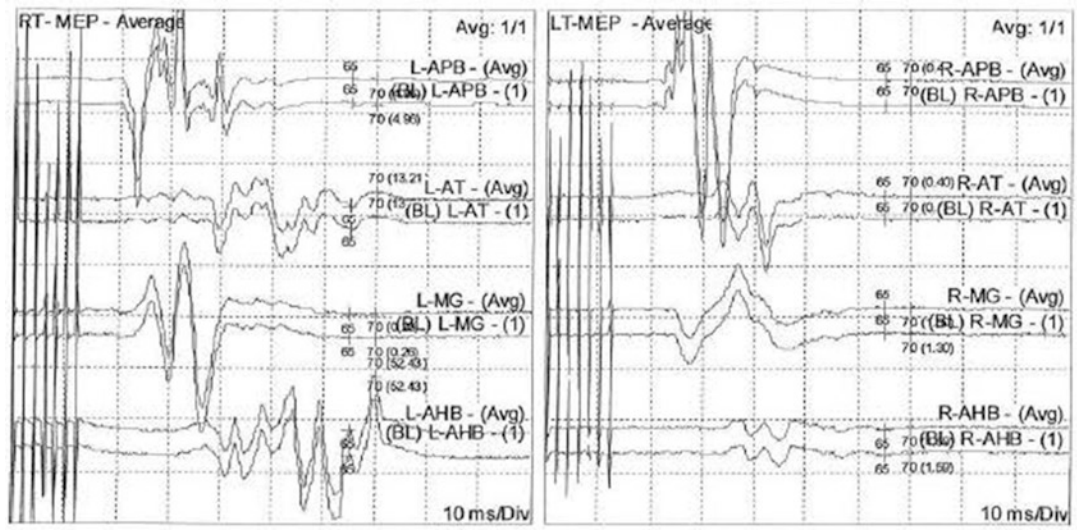

Fig. 15.1 Baseline MEPs of a 6-year-old with severe EOS at routine VEPTR lengthening. *APB* abductor pollicis brevis, *AT* anterior tibialis, *MG* medial gastrocnemius, *AHB* abductor hallucis brevis. Note the robust large deflection of these polyphasic waveforms. The sequence of seven spikes to the left of each frame is the transcranial stimulus applied at the scalp electrodes, and the subsequent waveforms are the peripheral responses recorded by transcutaneous muscle needle electrodes

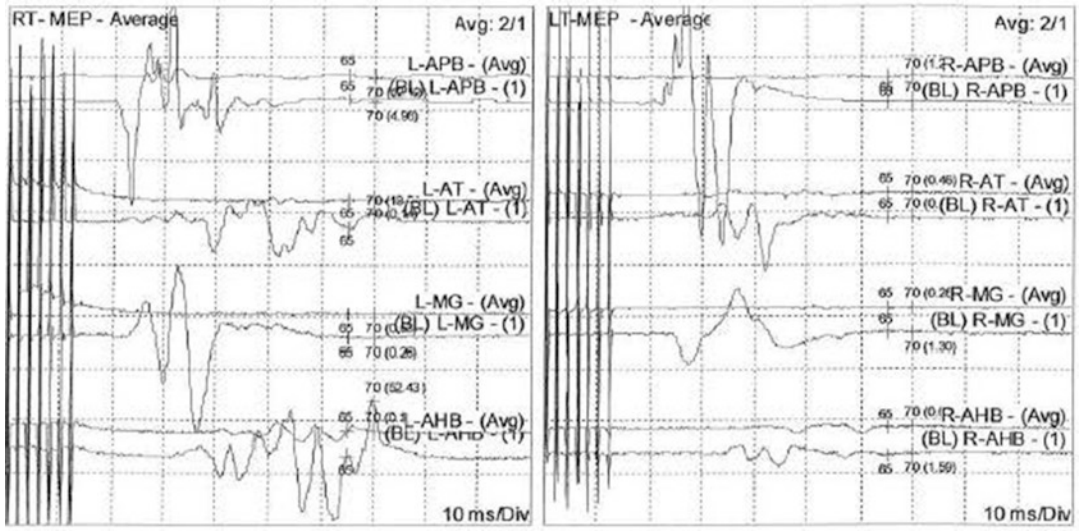

Fig. 15.2 The potentials have disappeared completely at APB, AT, and MG bilaterally, with barely a flicker of response in either foot. The visible waveforms seen on this screen are the baselines recorded earlier against which the new data, now largely absent, is recorded. The "flatline" is best seen in the MG trace on the right. This data was obtained upon prone positioning of the patient and before any surgical procedure was attempted

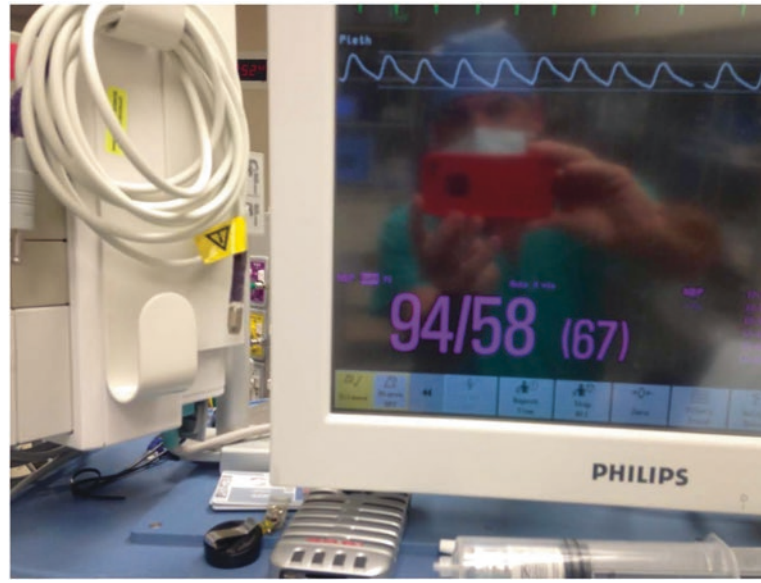

Fig. 15.3 Despite checking the patient's position, ensuring blood pressure was adequate, and checking for technical monitoring issues, the MEPs did not return. The operation was aborted

surgery, are reports of adults in the mid-1950s age range, and applicability to children's cervical surgery is questionable.

An algorithmic approach to alarm criteria is often posited, with reference to the checklists routinely used in the airline industry. The less-experienced surgeon embarking on a rare surgery in the child's cervical spine would be well served by this approach, as the extreme stress invoked by sudden loss of potentials during such a procedure can rapidly lead to a disorganized recovery strategy. Such an algorithm is presented in several articles. The one referenced earlier is recommended [9]. However, airplanes still crash or land at the wrong airports, despite these checklists, and they are no substitute for familiarity with the procedure and an experienced team of anesthesiologists, neurophysiologists, and surgeons. The literature is replete with evidence of better surgical outcomes in high-volume centers and with experienced surgeons. The particular problem in children's cervical spine surgery is that the caseload is extremely small, but the risk is very high. It is therefore logical to apply these recovery strategies as suggested in deformity surgery in general and not look for specifics in this rare field because they don't exist.

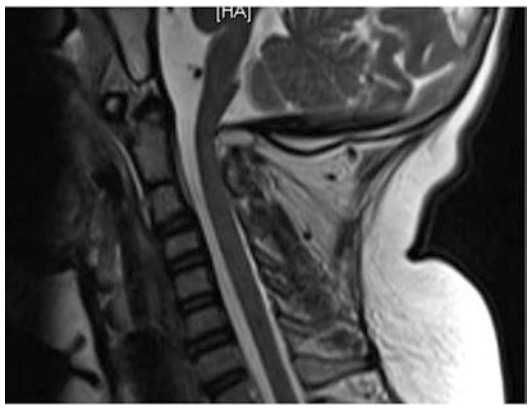

Fig. 15.4 Immediately after reversal of anesthesia, with no neurological deficit, an MRI was performed showing posterior impingement on the cord by the edge of the foramen magnum (A previous MRI 3 years before had been normal)

There is one other potential shortcoming of an algorithm for recovery of spinal cord monitoring potentials. By definition, an algorithm has a stepwise and presumptively logical approach to solving a problem (if x, then do y). It is unclear how long a child's cervical spine can suffer critical ischemia and still recover. It may be minutes or longer. If the recovery algorithm takes longer than the time tolerance of the spinal cord, then a stepwise approach is potentially damaging. At our institution, the three components of the team (anesthesia, neurophysiology, and surgery) act

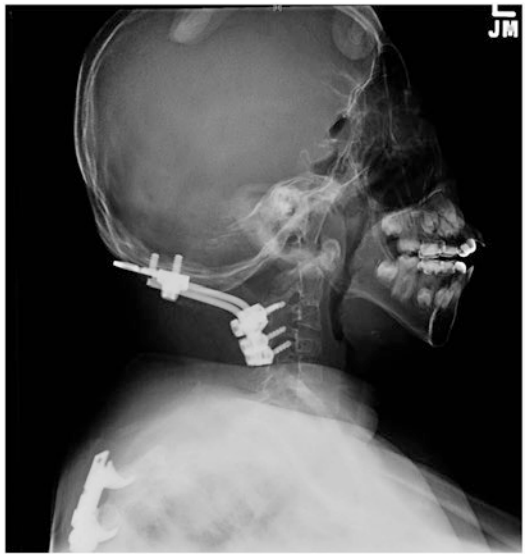

Fig. 15.5 Despite foramen magnum decompression and two successful VEPTR lengthenings after this, the monitoring alerts returned 18 months later, necessitating occipito-cervical decompression and fusion. Further VEPTR surgery is anticipated at the time of this writing

immediately and simultaneously when a monitoring alarm is called. Indeed, when there is downtrending monitoring data, a proactive strategy is followed almost intuitively before any threshold is met. This potentially lessens both the incidence and severity of the insult to the cord and diminishes the stress to the whole team. The strategies are listed here:

Anesthesia Elevate blood pressure. Do this proactively before any aggressive orthopedic manipulation that could cause cord ischemia. Check hematocrit, assess blood loss, and look at anesthetic agents; are there volatile agents still in the circuit?

Neurophysiology Is there a loose wire? Cross-reference SSEPs with MEPs (usually requires changing from one computer program to another). Is this a false-positive or has there been a gradual decline over minutes or hours?

Surgery Undo the last manipulation, remove retractors from the field, and irrigate with warm saline. Check the images on fluoroscopy; has the neck fallen into flexion? (This is usually, but not always, the position of danger.)

The advantage of the simultaneous approach is rapidity of recovery of potentials. The disadvantage is that the inciting event may be obscured by multiple concomitant interventions. Given the potential severity of the neurological injury resulting from cervical spine surgery, a simultaneous rather than stepwise approach is probably justified. Prior assessment of imaging studies by the surgeon, a thorough understanding of the medical comorbidities of the patient (e.g., congenital cardiac issues) from the anesthesia team, and meticulous technique in establishing the neurophysiological monitoring will all minimize both the rate of alerts and the false-positive rate. Recovery can then be expected to be predictably achieved through the interventions outlined above.

Should recovery of spinal cord monitoring potentials not be achieved by appropriate recovery strategies, then a difficult decision has to be made. Usually any instrumentation has to be removed. If no recovery supervenes, the surgery is abandoned. This is a bitter pill for the surgeon to swallow and is derived mostly from experience in thoracolumbar scoliosis surgery, where little doubt exists as to its advisability. The situation in children's cervical spine surgery may, however, be different. Much of what is done in this admittedly rare area is to treat spinal instability. Removing stabilizing instrumentation may actually worsen the potential for non-recoverable cord injury. The immediate availability of MRI under the same anesthetic, assuming a wake-up test that shows neurological loss, may shed light on an undetected mechanical problem. Alternatively, the child can be allowed to fully reverse from anesthesia and be reassessed both clinically and radiologically. This is much more palatable when a halo vest has been placed preoperatively to aid in stabilizing the cervical spine, and it is partly for this reason that the majority of these patients are placed in a halo vest as part of the surgical approach in the authors' institution. The vest can easily be removed at the end of a successful cervical fusion, but a potentially unstable cervical spine with no adjunctive external immobilization and lost spinal cord potentials is more of a management dilemma.

The foregoing thus addresses responses to alarm criteria in both a proactive and reactive

way. Post-alarm checklists are helpful, but meticulous preparation can avoid the need for their use.

Risks of Spinal Cord Monitoring

The risks of the techniques we are discussing are far outweighed by the risks of the surgery itself. There are, however, two main areas of risk which should be assessed. The first is a matter of quality and accuracy, and the second is a question of usually minor complications of the monitoring itself.

The latter concerns were the reason that MEPs were originally being neurogenically generated. Concern over inducing an epileptic seizure resulted in initial reluctance to stimulate the brain transcranially. As time has passed, the incidence of this potential complication has been found to be acceptably low, and the advantages of transcranial stimulation of the motor cortex have resulted in this technique's wide acceptance. In addition to this potential complication, local burns from stimulating electrodes, bleeding, and infection might result. Tongue bites are very rare if adequate bite blocks are used.

The bigger issue is one of quality, and therefore accuracy, of the data and how it is analyzed. There is good evidence that the greater the monitoring team's experience, the better the interpretation of the data and the better the surgical outcomes can be expected to be as a result. In particular, a multicenter survey by Nuwer and colleagues [2] addressed whether there is an overall benefit of SSEP monitoring during scoliosis surgery. Specifically, they evaluated outcomes for surgeons with much experience and frequent use of neuromonitoring compared to others who had limited or no use. Surveys derived from 173 US surgeons (all members of the Scoliosis Research Society, SRS) over a 15-year period compared historical data. 88% (153) of the surgeons who responded to the survey used SSEP monitoring for some or all of their cases, which numbered 97,586. Severe persistent deficits, such as paraparesis or paraplegia, were reduced by more than 60% by interventions based on intraoperative SSEP monitoring. Surgeons who use SSEP monitoring the most produced the lowest rate of neurological deficits, according to this survey [2]. Attesting to the efficacy of this intraoperative neuromonitoring technique, Schwartz et al. remarked that "SSEPs have had a remarkable success in identifying impending neurological injury, particularly when viewed in perspective of the thousands of cases monitored by an extremely divergent group of monitoring personnel…." [15]. For example, in the Nuwer et al. study, a false-negative rate of 0.063% was reported for 51,263 spinal cases in which SSEPs were the only modality monitored. Furthermore, the negative predictive value (i.e., the likelihood of normal spinal cord function in the presence of stable SSEPs) was an impressive 99.93% [2].

Assuming good experience, the modalities chosen for monitoring are limited next by their intrinsic accuracy, the nature of which we have already discussed. Deterioration of accuracy can be influenced by poor electrode placement, mechanical uncoupling of monitoring cables, or poor baseline electrophysiology of the patient (such as in cerebral palsy or an already myelopathic child). Usually, mechanical issues are very obvious, and the baseline poor physiology can be assessed after induction of anesthesia and elimination of volatile anesthetic agents. A decision can then be made to rely or not on the limited data available in the neurologically impaired patient, though abandonment of monitoring is rarely needed, and often one modality may be preserved (e.g., SSEPs) though the other is unreliable. It then remains to be cautious of the data and interpret it in the face of the clinical scenario. In such situations, it is best to avoid hypotensive anesthetic techniques for blood loss limitation, to attend assiduously to patient positioning, and to have a very low threshold for pausing the surgical process should any alarms occur. Occasionally, electrophysiological data actually improves during the procedure, perhaps due to decompression of spinal elements. At least we should aim for preservation of the baseline thresholds and address changes that are seen, as discussed above.

The Ideal Situation

In an ideal world, the child undergoing cervical spinal surgery—whether anteriorly or posteriorly—would have an experienced operating theatre team of anesthesiologists, neurophysiologists,

nurses, and surgeons. Baseline SSEPs and transcranial MEPs would be obtained in the supine position after induction of anesthesia and fiber optic intubation. A halo vest would be applied and, when extreme spinal instability is a concern, a second set of baseline potentials recorded. Care would be taken to stimulate for MEPs with the lowest possible voltage to minimize the myoclonic jerks and thus any irritation of the cord that results. Assuming monitoring is satisfactory, the patient would be turned prone for a posterior approach and positioned on the table. Another baseline recording would be made, since the mere act of prone positioning is known to cause loss of potentials [16]. The surgery would proceed uneventfully, cervical stability would be achieved and fusion performed. The halo vest, if any, would be removed at the end of the procedure and the child awoken neurologically intact.

Summary

The foregoing addresses current thinking in spinal cord monitoring for spinal surgery in general, children's spinal deformity surgery more specifically, and children's cervical spinal surgery by implication. In the literature search for the preparation of this chapter, only one reference specific to spinal cord monitoring in pediatric cervical spine surgery could be found in the English language [16]. However, the principles of neurophysiological monitoring of the spinal cord during orthopedic and neurosurgical procedures are well established and, indeed, should be followed in the management of disorders of the child's cervical spine.

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External Immobilization of the Child's Cervical Spine

16

Michael B. Johnson and Leah McLachlan

Introduction

A range of external devices has been used over centuries to immobilize—or in some cases to correct or alter—neck shape in children. This chapter seeks to describe the known applications, contraindications, and complications of modern orthotics used in the management of children's cervical spine conditions. Children have specific anatomical differences of size, proportions, and biomechanics that have direct implications for the choice of devices.

Where there is some paucity of data specific to children's cervical spine, some extrapolation from the adult literature is necessary. The adult literature itself is limited in data and evidence, particularly with regard to effectiveness of specific devices. There is, as a result, little consensus in indications, duration, or choice of devices.

There are two categories of devices: those that provide a support or immobilization and those that seek to provide a corrective force on the growing cervical spine. Orthoses are applicable in a wide range of traumatic, developmental, and acquired conditions of the cervical spine, but the details of indications are discussed elsewhere in this book, under the specific conditions.

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Anatomical and Biomechanical Considerations

Appropriate application of cervical orthoses requires an understanding of the anatomical and biomechanical differences present in children. The size and proportion of the head and neck change throughout childhood (Fig. 16.1).

Table 16.1 gives some data for the change in size of the skull. The child's skull makes up a much larger portion of the whole body. By age 5, the skull has reached 95% of the size of the adult skull dimensions.

The impact of a larger skull is to increase the amount of occipital offset, i.e., the protrusion of the skull posteriorly with respect to the posterior aspect of the torso. Both the occipital offset and the occipital neck angle change with growth (Fig. 16.1). Positioning a child's cervical spine in a neutral position is a key principal of immobilization, requiring increased amounts of occipital offset in younger children. The average offset required was found to be 25 mm [1]. The neutral position also ensures an appropriate shape for the airway. Cervical orthoses applied without accounting for offset will produce a kyphosis of the cervical spine and obstruct the airway.

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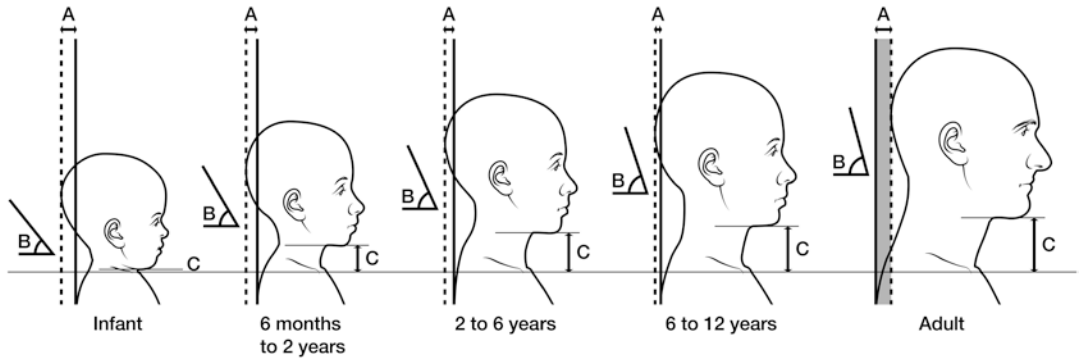


Fig. 16.1 Skull size and occipital offset during childhood. (**a**) indicates the occiput offset with respect to the posterior aspect of the trunk. This diminishes from 2.7 cm in infancy to become a negative offset in adults (Indicated by shaded area) [1]; (**b**) the angle of the occiput forms an acute angle in infancy; (**c**) the chin offset with respect to the sternum steadily increases over childhood

Bone strength is a function of bone dimensions (overall thickness, cortical thickness) and bone density/composition. These parameters are considerably reduced in children (Table 16.1). Skull thickness increases through childhood, as seen in measurements from CT scans (Fig. 16.2) [4, 5]. Children do not acquire adult skull thickness until age 16. Similarly, bone density does not significantly increase until the onset of puberty. Orthoses, which utilize skeletal fixation, need to account for the reduced bone strength in younger children.

Children have greater joint laxity overall. In the cervical spine, there is additional range of motion because of relatively flat facet joints and lack of uncinate processes [8]. There is greater flexibility to distraction force through the joints compared with the spinal cord, with implications for the use of traction. Cranial nerves, which traverse from upper spinal cord to eyes, are at the greatest risk for traction injury.

Biomechanically, the fulcrum of motion for the cervical spine is higher than that for adults. For infants this is at the C2/C3 level, at age 5 at C3/C4, and by adolescence at C5/C6 which is the same as for adults [8]. Cervical instability in younger patients is more often at the upper cervical levels, with the implication that standard rigid cervical collars will not control this level. Immobilization at this level will require involvement or control of the skull, and extension to the thorax.

The role of an orthosis in general is to maintain an anatomical neutral position of the cervical spine against the forces that are threatening to displace it. These forces are the static gravitational force of the skull, the voluntary muscle force, and the involuntary forces applied by falls, car travel, etc. Cervical orthoses are limited in the ability to apply a direct force against the individual motion segments of the cervical spine, largely because of the surrounding soft tissues. As a result, the immobilization forces of the orthotic are applied against the more rigid structures of the occiput, mandible, and thorax. Rigid collars achieve control through the connection of the two halves of the orthotic. For example, a flexion motion on a rigid collar is resisted by the stiffness of the anterior half of the orthosis running from mandible to sternum. An additional anterior displacement force is resisted by the connection to the posterior half by Velcro or buckle strap [9].

A cervicothoracic device that extends to above the occiput and onto the trunk increases the contact surface area and places the rigid columns of the orthotic farther from the center of rotation (within the spine). This achieves an increased moment and a resultant increase in rigidity. There are two methods of extension to a cervicothoracic orthoses. Some orthoses, such as the SOMI and Minerva, employ posts to a separately fitted thoracic vest. Many prefabricated collars now have prefabricated extensions. There is little evidence of the effectiveness of such braces over traditional cervicothoracic orthoses (SOMI, Minerva).

Table 16.1 The skull dimensions in childhood compared with mean adult size

	Mean transverse diameter cm (%)[a]	Mean longitudinal diameter cm (%)[a]	Skull thickness (mm)[b]	Bone density (% of adult normal)[c]	
Age				Male	Female
0	10.2 (72%)	13.7 (72%)	1.8	N/A	N/A
1	12.5 (88%)	16.3 (86%)	2.0–3.7	N/A	N/A
5	13.4 (96%)	17.9 (95%)	3.0–4.8	65%	69%
10	13.8 (98%)	18.3 (97%)	3.0–6.1	74%	74%
15	14.1 (99%)	18.6 (98%)	6.0	92%	97%
17	14.2 (100%)	18.9 (100%)	>6.0	99%	100%

[a]Mean cranial vault region diameter [2, 3]
[b]Mean skull thickness (inner and outer table). Note considerable variability [4–6]
[c]Total body bone mineral density (DEXA), not available under age 4 [7]

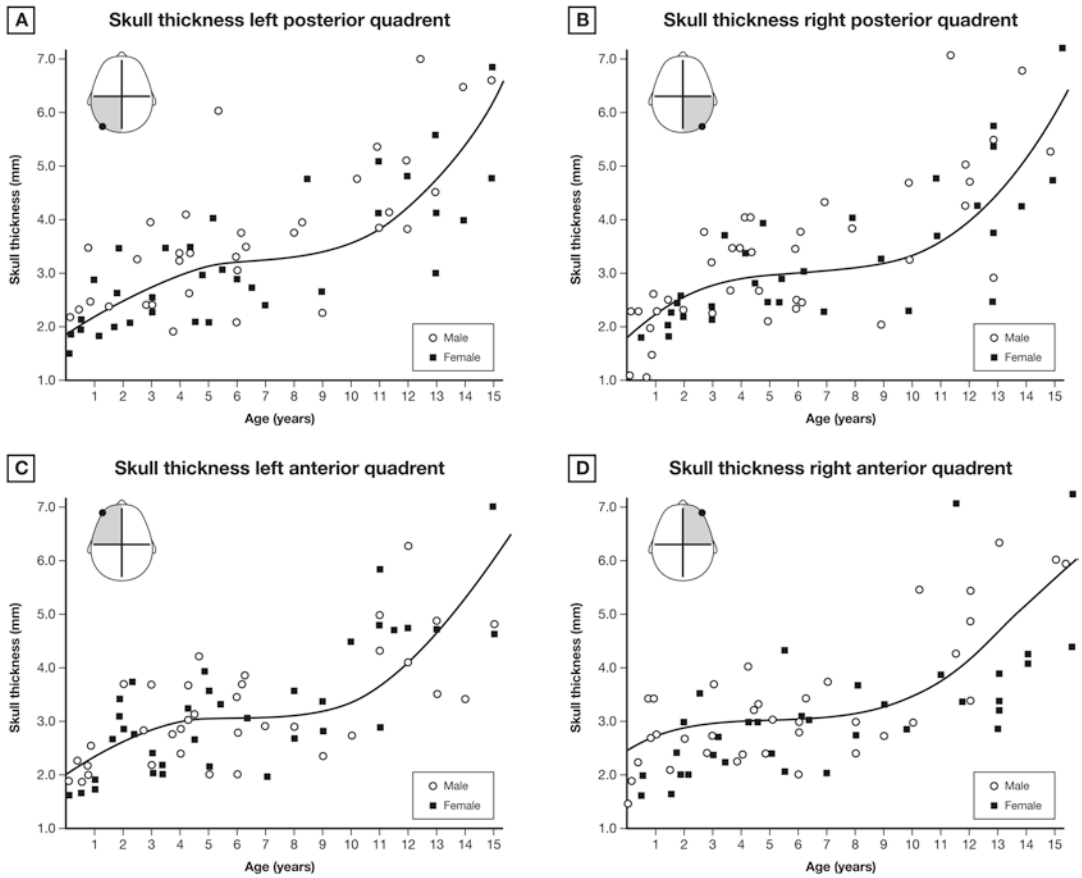


Fig. 16.2 Skull thickness on CT scan during childhood. Skull thickness in millimeters as measured from CT scans, stratified by age, gender, and location: (**a**) left posterior, (**b**) right posterior, (**c**) left frontal, (**d**) right frontal. Reproduced with permission and copyright © of the British Editorial Society of Bone and Joint Surgery [4] (Fig. 16.4)

A phenomenon common to many cervical orthoses is described as "snaking." For example, a flexion movement of the head is resisted by the collar, causing extension in the upper cervical spine, while permitting flexion at the cervicothoracic junction [10] (Fig. 16.3). Snaking may be reduced by collars that encapsulate some of the soft tissues of the neck or by extension of the orthotic from the skull to the torso, e.g., in CTLSOs [9].

A variety of methods have been used to assess the biomechanical effectiveness of cervical orthoses. Some studies have used cine-radiology (or more recently 3-D motion analysis [11, 12] and electrogoniometry [13]) on adult normal volunteers to describe the effect of a brace on the motion of each segment. There are, as a result, important limitations on the applicability of this knowledge to children. A further limit of applicability is that the motion is generated by the volunteer in a single plane of motion, without ability to measure the force applied to the orthosis. Other studies avoid this limitation by the use of anatomical models to perform in vitro analyses of brace function [14]. A final group of studies use cadaver studies to simulate cervical spine instability.

Specific Orthoses: Implant Types/Materials

- Soft collar
- Dynamic support: MBrace
- Rigid collar (cervical orthoses)
  - Aspen™
  - Philadelphia™
  - Miami J™
  - Stifneck
- Cervicothoracic orthoses
  - SOMI
  - Minerva
  - Pinless halo
  - Halo-thoracic vest
  - Papoose™

Soft collars are typically constructed of foam covered by cotton stockinette. They provide a very small amount of restriction to motion (Table 16.2). As such, the main role for such braces is as a proprioceptive aid or as a step-down from a more rigid orthosis. As an analgesic device, it may offer some warmth and comfort.

The *MBrace* is a collar designed specifically to restrict hyperextension while allowing normal

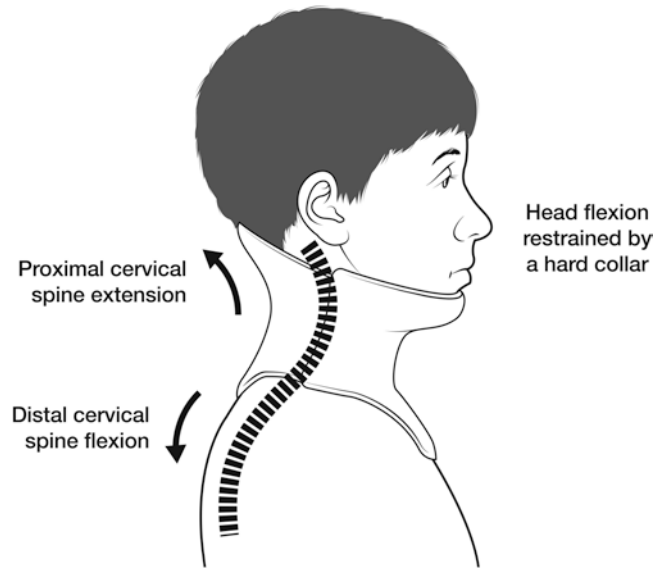


Fig. 16.3 "Snaking" movement in a cervical orthosis. When the subject attempts head flexion, the restraint of the collar produces a reciprocal upper cervical extension and a distal cervical flexion

Table 16.2 Comparison of common prefabricated orthoses with respect to percentage restriction of occiput to C7 motion

Orthosis	Flexion-extension C0–T1	Flexion-extension C0–C1	Lateral bending	Axial rotation	Reference
Normal range of motion (degrees)	135 ± 16	20	99 ± 12	145 ± 20	[11, 15]
Soft cervical collar	5–20%		5–14%	10–17%	[6, 16–20]
Rigid cervical collars					
Philadelphia	49–71%		26%	29–56%	[11, 16, 17, 21]
Aspen	62–77%		34–62%	38–59%	[10, 11, 21–23]
Miami J	68–73%		31–43%	50–65%	[21–23]
NecLoc	83%		51–60%	73%	[22]
Miami J with Occian back	63%		35%	64%	[21]
Cervicothoracic orthoses					
SOMI	44–89%	Paradoxical increase	34–80%	50–66%	[16, 17, 19, 23]
Minerva	87%		50–79%	69–82%	[17, 23, 24]
Pinless halo	86%		84%	81%	[23]
Halo	96%		96%	99%	[16, 17]

SOMI sterno-occipital-mandibular immobilizer

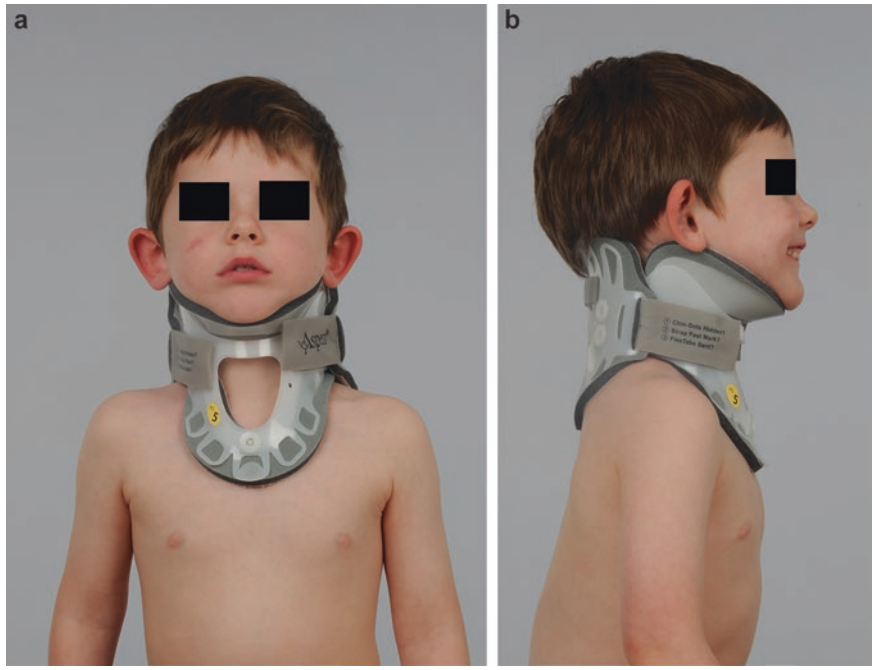
flexion movements. This is only indicated in whiplash hyperextension injuries and is therefore only likely to be applicable in the older adolescent group [25].

Rigid collars (Aspen™, Miami J™, Philadelphia™, Malibu™) achieve increased stability as a result of stiffer materials, creating a rigid column from the upper thorax to the occiput (Fig. 16.4a, b). Most of these devices have a midline anterior cutout, which alleviates pressure on the trachea with attempted extension in the brace and provides space available for a tracheostomy. Collars prevent flexion-extension movement but are relatively poor in preventing lateral flexion and rotation. The fit of a rigid collar has an effect on the amount of immobilization achieved. In particular, increasing height of collar increases the stiffness in all planes at the expense of increasing extension of the cervical spine and increasing pressure on the chin [13]. The make and design of collar show variability in overall stiffness: in increasing order of stiffness, they are Philadelphia, Aspen, Miami J, and NecLoc (Table 16.2) [22]. The Miami J and Aspen collars produce less contact pressure on the occipital skin, which is desirable in the prevention of ulceration and may be a cause for improved comfort scores [21, 23, 26, 27].

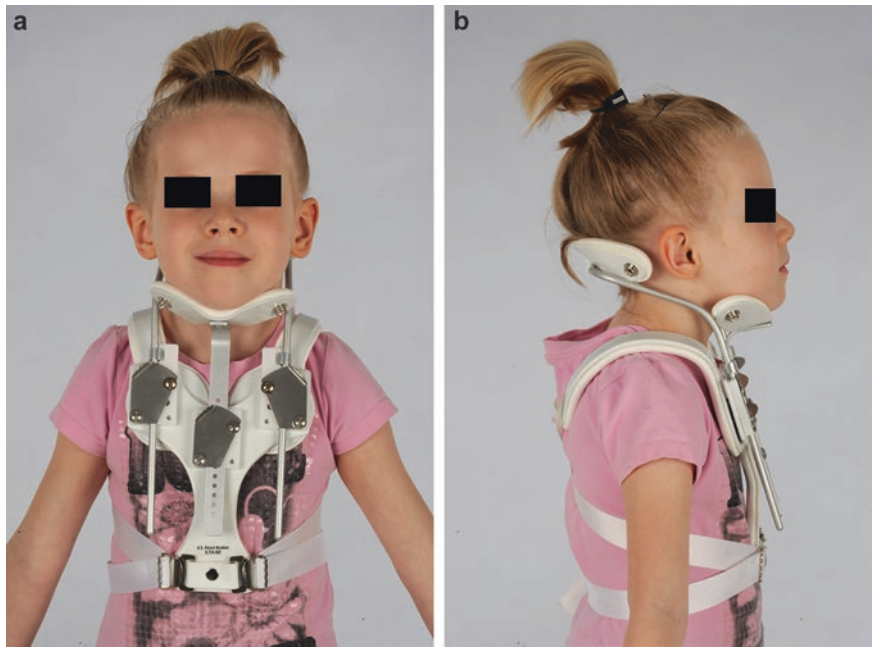
Cervicothoracic orthoses include the SOMI (sterno-occipital-mandibular immobilizer) and Minerva (Figs. 16.5a, b and 16.6a, b) and rigid collars with extension to a thoracic component [14]. There is an increase in the restriction of motion achieved with more rigid materials and with extension onto the thorax. These devices perform less well against cervical rotation forces. The fit of the chest piece, especially the strength of connections of the anterior and posterior halves of the chest piece, determines the effectiveness against rotation.

The SOMI has an anterior single strut and two posterior struts that run from occiput to the anterior chest piece. Some "snaking" at the C0–C1 level has been reported with the SOMI, such that it does not restrict motion at this level as it does in the sub-axial spine [28, 29]. Despite the extension onto the thorax, there is no effective immobilization below C7–T1. The SOMI provides less resistance to extension moments because the posterior struts are being subjected to a bending load rather than an axial load [9].

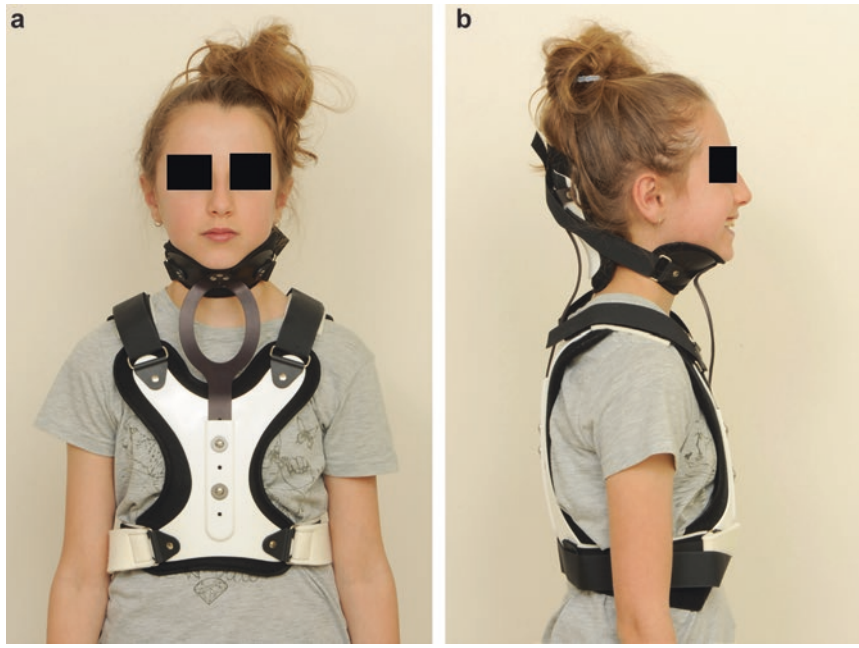
The Minerva is an example of a four-poster cervicothoracic orthosis. In contrast to the SOMI, the posterior struts run to a more rigid posterior thoracic piece. This provides a better extension resistance. The original design of Minerva braces utilized a cast mold thoracic



Figs. 16.4 Aspen cervical orthosis. Illustrates a typical two-piece prefabricated cervical orthosis with rigid and soft foam elements. (**a**) Frontal view; (**b**) side view



Figs. 16.5 SOMI (sterno-occipital-mandibular immobilizer). (**a**) Frontal view and (**b**) side view illustrate that the posterior post connects with the anterior portion of the separately fitted vest



Figs. 16.6 Minerva cervicothoracic orthosis. (**a**) Frontal view and (**b**) side view show improved resistance to extension, compared to the SOMI, by the posterior strut connection of occipital component to the molded posterior cervicothoracic shell

component. Modern materials use a prefabricated thoracic component equivalent to that used in halo-thoracic devices, connected to chin and occiput pieces with rigid uprights.

The infant with a need for cranio-cervical stabilization presents a particular challenge. The Papoose™ is a prefabricated molded shell that incorporates the skull, cervical spine, and torso in one unit, with an appropriate occipital offset. There are options for an anterior cervical orthosis piece to fit to provide further restraint against flexion, in addition to the strap across the forehead. This device is suitable from 0 to 3 months old, but similar custom molds can be made for even younger premature infants up to the age of about 4–5 years (Fig. 16.7). It is most ideal in nonambulatory patients. There is no data on the biomechanics of this device. In theory it would be similar to the results achieved with the Minerva brace.

The *halo-thoracic device* provides the greatest overall restriction of motion at all levels and in all directions, particularly in the uppermost cervical motion segments (Fig. 16.8a, b). The stability achieved is the result of pins inserted through the

Fig. 16.7 Custom-made Papoose. This custom-made posterior shell has been fitted to an anterior Aspen component for a 2-year-old. Note the occipital offset incorporated in the mold of the posterior shell

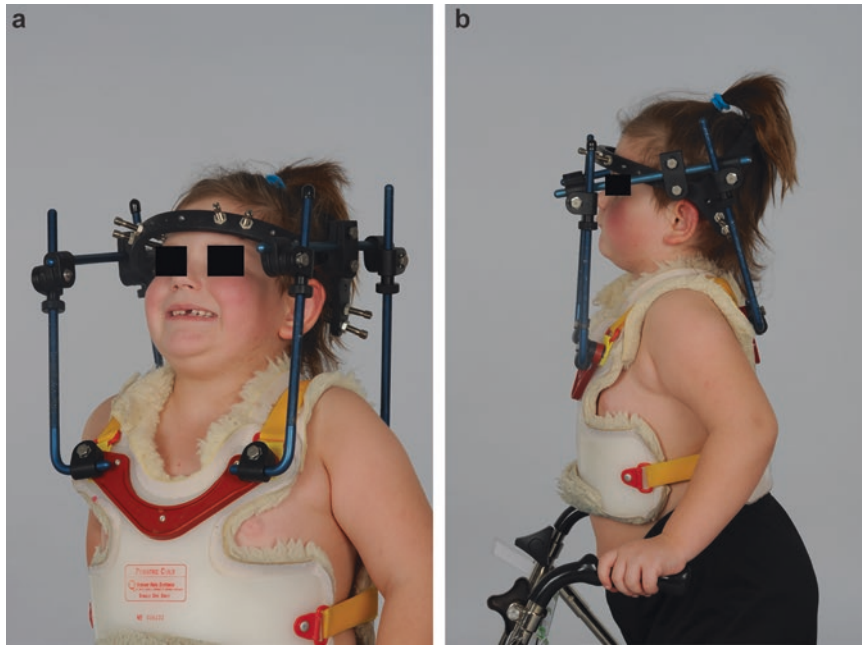


Fig. 16.8 Halo-thoracic vest. Pictured is a typical 8-pin construct for age 5–12 years. (**a**) Frontal view shows pins inserted above the lateral 2/3 s of the orbit; (**b**) side view in an ambulant patient. The posts are adjusted to produce the desired position with respect to flexion/extension

skin and pressed against the outer table of the skull. These translate bending forces to axial loading on struts attached to the chest wall, which are well away from the axis of rotation and thus provide very powerful moments. Rotational control is much improved compared with other orthoses but is still dependent on the quality of fit of the thoracic component. Snaking motion can occur, especially with a loose vest or in prone or supine positions [30]. See the technique for halo application below.

A pinless halo involves a ring around the head with contact on the skin, with silicone composite pads on the forehead and occiput. A ring around the mandible controls rotation. This has been successfully used in children as management for fractures and postoperative treatment (fusion, torticollis, atlantoaxial rotatory subluxation). Skaggs et al. reported successful treatment in 29 out of 30 children [31]. Complications included skin ulceration, noncompliance, and self or family removal. The one failure led to questioning whether this orthosis provides adequate support in the most severely unstable spine. Sawers et al. reported 1 of 19 adults with occipital ulceration and two cases of noncompliance with self-adjustment of straps [32].

In many cases there are prefabricated braces in each of the categories listed, which would suit a wide range of patient age and size. There are, however, extremely limited prefabricated cervicothoracic orthoses for children between the ages of 3 months and 2 years of age. Halo vests can be fitted for children older than approximately 18 months. For the very young infant and child, there is a need for customized individual solutions. Even when there is a prefabricated brace of appropriate size, there can be a significant need for brace modification to achieve appropriate fit. This emphasizes the need for a good orthotic service.

Applications for Orthoses

Provision of External Support: Instability

Key questions that need to be addressed in the choice of external support in the case of instability

included the cause, location, direction, and likely duration of instability, patient size and compliance, and potential complications. In many cases there are a wide range of options available, and there may be local factors of experience or availability of orthoses that influence the choice of external support.

Initial Trauma Management

In the setting of trauma, the requirement for cervical spine immobilization can begin in the prehospital setting. Adequate immobilization involves the use of spine boards with an occipital offset for elevation of the torso. Sandbags taped to a short board can provide the most effective immobilization in the prehospital setting. An evaluation of three methods showed that significant lateral movement of the neck can still occur, as a result of difficulty securing the thorax [33–35]. A rigid cervical orthosis applied without consideration for the size of skull can apply a significant flexion and distraction force to the skull. Distraction and flexion forces are especially undesirable in the very young because of the significant incidence of occipito-cervical dissociation in high-speed deceleration injuries [36]. There is some debate in the literature as to whether rigid orthoses should be applied at all in this setting [37]. These counterarguments point to the paucity of any data showing benefit and the creation of other difficulties such as delaying extraction from vehicles and difficulty with intubation [38]. Some orthoses can cause genuine harm through excessive kyphosis, airway compromise, and excessive skin pressure.

At hospital level the requirement for occipital offset or torso elevation continues (Fig. 16.9a–d). This can be provided according to size and age of child with a proprietary product, e.g., Occian AirWay PAD™ or simply an additional thin mattress under the torso. A sizing chart, such as the Broselow-Luten system, can assist. A rough rule

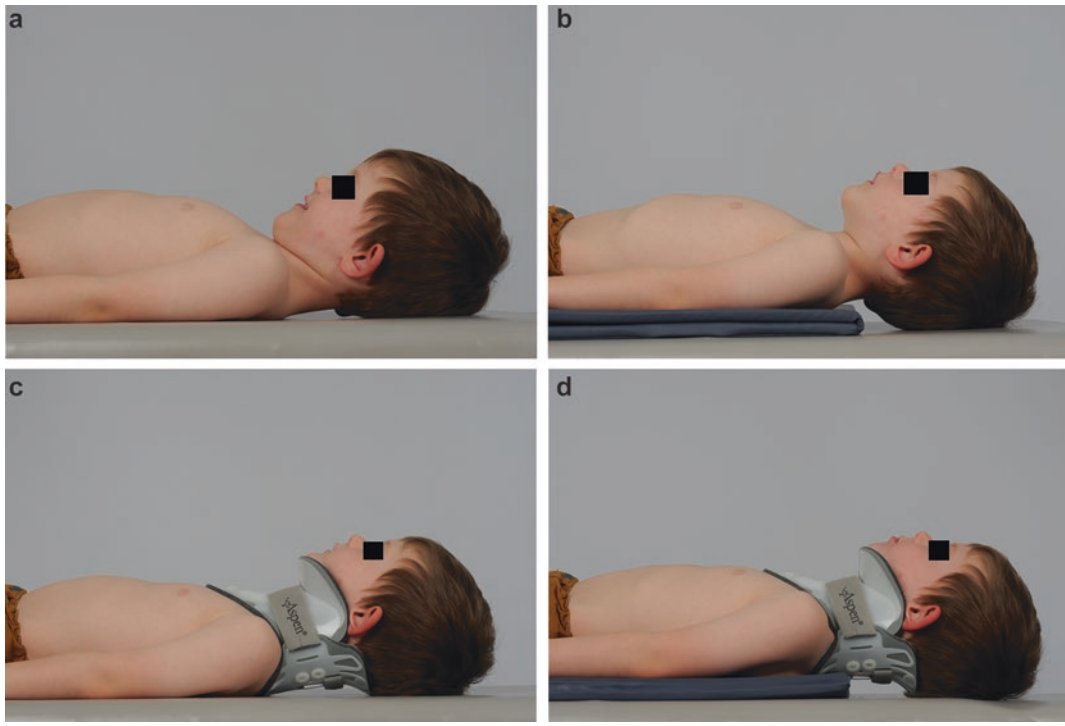


Fig. 16.9 The importance of occipital offset in the neutral positioning of the child's cervical spine. Illustration of the cervical spine of a 5-year-old lying on a firm surface (**a**) without a mat and (**b**) with a mat, accounting for occipital offset. The provision of a (**c**) cervical orthosis alone is insufficient to avoid excessive cervical flexion and kinking of airway; (**d**) shows the improvement of positioning with provision for occipital offset

of thumb is to provide enough offset to align ears with shoulders. The offset is frequently forgotten in the acute resuscitation setting and particularly in CT and MRI scanners. Full spinal precautions include the addition of a rigid collar and sandbags or equivalent to prevent lateral or rotational movement. Hospital protocols are required to expedite the removal of unnecessary spinal precautions and immobilization in order to reduce the complications associated with the supine position and rigid cervical orthoses.

Later Trauma Management

Cervical orthoses are used as definitive treatment for traumatic instability where there is an expectation of healing. The level of instability, whether traumatic or nontraumatic, strongly influences the potential direction of instability. Biomechanical studies assist in determining the best orthotic choice to reduce movement in the desired direction. Distraction forces are particularly difficult to counteract. Even halo vests produce significant distractive forces in the supine position [39].

The required duration of orthotic treatment for posttraumatic injury is dependent on the rate of healing, on the restoration of normal stability, and on the onset of any complications.

Postsurgical Treatment

The use of orthoses as an adjunct to surgical stabilization of the cervical spine is controversial in adults. Problems associated with collars used in the postoperative setting include increased surgical site infections [40], dysphagia [41], and respiratory compromise.

Proponents for the use of orthoses in children following surgery point to the difficulty of behavioral control in children, who may mobilize far too early, placing excessive loads on limited internal fixation implanted in relatively weak bone [42]. External immobilization may be essential in bone dysplasias [43].

Torticollis has many causes, many of which are not amenable to brace treatment. A common indication for an orthosis is in the immediate postoperative phase after sternocleidomastoid release for muscular torticollis. Orthoses used for this purpose typically have an asymmetric design with more rigid materials in the previously concave side.

- Atlantoaxial rotatory subluxation
- Developmental instability
- Congenital anomalies of the cervical spine
- Dysplasias

A variety of orthoses have been used in the treatment of atlantoaxial rotatory subluxation. In most cases orthotic immobilization has been used after reduction is achieved with the use of traction and muscle relaxants [18]. Successful maintenance of reduction has been described with the use of the Philadelphia collar [44]. The authors, however, prefer a device with better control of axial rotation. A modification can be made to the Minerva brace with the use of a side strap, or the pinless halo is a suitable alternative [31]. The halo vest provides the most rigid control of axial rotation and has been reported to be successful in treating refractory atlantoaxial rotatory subluxation with observation of remodeling of C1-C2 facet joints [45, 46]. The fit of the halo vest has been critical in the control of rotation.

While significant nontraumatic upper cervical spine instability is generally an indication for operative stabilization, there may however be a temporizing role for bracing. In such situations the brace is used to control the instability, preventing further neurological deterioration while gaining further growth. Caution should be exercised, as not all conditions will be adequately stabilized. Neurological deterioration may still occur [47].

Nonoperative treatment of kyphosis has been described with combinations of traction and orthotics. Katz et al. concluded with their experience of kyphosis secondary to Larsen's syndrome that the results were very unpredictable [48]. Prevention of post-laminectomy kyphosis with the use of a brace remains controversial, with little supportive evidence [49].

Corrective Forces: Traction

Corrective (mainly traction) forces can be applied to the cervical spine. These can be applied via a halter under chin/occiput, or through tongs, or a

halo device. In considering the role for traction, consideration needs to be given to the patient age and size, the nature of the deformity, the likelihood of correction, the time required to achieve a reduction, and the possible complications. Often a period of traction is followed by a period of support in an orthosis.

The nature of the deformity determines the angle of traction to be applied. For example, the correction of atlantoaxial rotatory subluxation requires a neutral angle with respect to flexion/extension. This further requires that the occipital offset be taken into account and appropriate torso elevation applied. A kyphotic deformity would require a component of extension to the angle of traction.

Halter traction is only suitable for short-term, low-tension force application such as that required to manage spasmodic torticollis. Maximum forces applicable through the halter are 1–2 kgs. Care needs to be taken of the skin under the occiput and chin.

Much greater tension forces can be applied through a halo ring or tongs. Indications include the correction of atlantoaxial rotatory subluxation [46], unilateral or bilateral facet joint dislocation, and slow correction of kyphotic or scoliotic deformities. Additional care needs to be taken with pin sites and neurological assessment especially when tension forces are being increased. A maximum weight of 1/3 body weight should not be exceeded. Cranial nerves are at risk for neuropraxia, especially the abducens CN VI and oculomotor IV. Lateral gaze palsy should be regularly checked for during traction [46].

Techniques

Collar Sizing

Individual products will come with sizing recommendations and techniques. The Broselow-Luten system is a commonly used pediatric measuring guide used in emergency departments to avoid common drug dosage mistakes. In the setting of acute trauma, it can be used to provide a guide for the amount of occipital offset required, as well as the correct size for cervical orthoses. Several orthotic providers base their sizing recommendations on this system.

Halo Application in Children

Important precautions need to be taken in the application of halos in children. In particular, attention needs to be given to size, shape, and thickness of the skull. MRI-compatible materials are readily available and are preferable to ferromagnetic materials. Most halo designs account for the shape of the skull at the level ideal for placement. Longer pin lengths may be necessary if there is a significant mismatch between ring size and skull diameter.

Skull dimensions rapidly approach adult numbers, permitting the use of a small adult ring in the average 5-year-old (see Table 16.1). It is recommended in the adult population that four pins inserted with a torque pressure of 8 lb./inch lower the rate of pin loosening and infection [50, 51]. There is a significant variability of the thickness and bone density of the pediatric skull (see Fig. 16.2). This requires an increase in the number of pins and reduction of torque pressure [5, 52]. Mubarak et al. recommended up to 10–12 pins inserted at 2 lb./inch torque in the under-3-years-of-age group [52]. A pre-application CT scan is recommended to identify areas of significant thinness and assist in planning pin sites [4].

Increasing the number of pins increases the probability of pin positions in non-ideal locations. The halo ring should be below the maximum diameter of the skull and above the ears and orbits [50]. It remains desirable to insert anterolateral pins at 1 cm above the lateral 2/3 of the orbit. These pins should avoid the zone above the medial 1/3 of the orbit to avoid the supraorbital nerve and frontal sinus and should avoid the temporal bone to avoid interference with the temporalis muscle and zygomaticotemporal nerve. In children under 18 months of age, the patent anterior fontanelle is an additional hazard to be avoided. Posterolateral pins are placed behind the pinna on the occipital bone.

The halo ring is connected to the chest piece with four struts. Positioning should take into account the occipital offset by placing the ears in alignment with the shoulders. The quality of fit

of the chest piece will determine the degree of rotational control achieved.

Particular steps to note in halo application in children:

- Obtain limited CT scan.
- Measure/template size of ring.
- Measure chest size, estimate size of vest.
- Estimate pin length and number required.
- General anesthesia except in the most compliant older age patients.
- Secure eyes shut.
- Shave hair if necessary over mastoid processes.
- Position halo (temporary pins).
- Prep solution (avoid alcohol or soap containing prep around the eyes).
- Introduce pins through desired holes, to the skin.
- Local anesthesia injection.
- Sequential tightening to reduce torque (diagonal alternates).
- Lock pins with locking nuts.
- Recheck torque on second day, once only, and cease tightening if screw fails to achieve desired torque after a full turn of screw.

Complications

The use of rigid cervical orthoses can have adverse effects, including pressure necrosis of the skin, increased intracranial pressure, effects on swallowing and respiration, paraspinal muscle atrophy, loss of flexibility, and failure to achieve the aims of treatment through incorrect fitting, loss of position, or noncompliance. The psychological aspect of prolonged brace wear should not be ignored.

Common areas of increased pressure are under the chin and mandible, sternum, and occiput. Patients at greater risk for development of pressure areas are recumbent or with reduced conscious state, particularly with closed head injury [26]. This emphasizes the importance of early removal of unnecessary immobilization [53].

Hunt et al. described a small but significant elevation of intracranial pressure secondary to rigid orthoses, which would potentially be clinically important in head injury patients [54]. This has been corroborated with more recent findings recommending removal or, at minimum, readjustment of collar in the setting of elevated intracranial pressures [55, 56].

Complications of halo treatment are common with reported incidences in the literature of up to 44%. Complications reported include pin site infection, scarring, nerve injury, dural penetration with CSF leak, brain abscess [57], seizures, dysphagia, fungal or lice infection, muscle atrophy, and hearing loss [58]. In a large retrospective review, Bransford et al. reported on 12 halo vests used in children (<16 years old) out of a total of 342 patients. They reported a completion of halo vest treatment (survivorship) of 71% in pediatric patients (to about 12 weeks) with a 44% complication rate [59].

The most common complication of halo treatment is pin site infection (13%); one-third are manageable with oral antibiotics and pin track care alone and 50% can be managed by pin removal and debridement. Pin removal can result in a compromise to the stability of the construct. Serious infections requiring surgical debridement and IV antibiotics are less common (5%). Pin infections become more common after 6 weeks of halo application [59].

Pin loosening can occur as a result of infection or can be aseptic. Fleming et al. measured the compressive force at the tip of the pin through the life cycle of a halo vest, demonstrating an 83% loss of compressive force [60]. Re-tensioning of pins can be hazardous in children, increasing the risk for pin penetration.

The pediatric airway is compromised by excessive flexion. Intubation in general anesthesia is more hazardous with a rigid collar applied [61]. The rigidity of the halo creates greater problems for intubation. Awake, fiber-optic intubation may not be able to be tolerated by pediatric patients [62]. Other options described include fluoroscopically guided scopes with less resulting motion to the upper cervical spine [63].

Ono et al. described atrophy of the sternocleidomastoid and nuchal muscles on CT scan. After

12 weeks of halo vest application, the muscular atrophy amounted to 15–22% but was found to be reversible [64].

Complications of halos leading to loss of reduction, failure of union, and/or persistent instability are the most serious. Reported incidences of this type of failure range from 9–15% [59]. In addition, incorrect fitting can result in significant loss of effectiveness in immobilization.

Summary

An understanding of anatomical and biomechanical differences is extremely important in the correct application of orthoses in children for both cervical spine and airway alignment. Halo-thoracic devices can be used in children but require specific care and attention to details to avoid complications. In short, there is an absence of an evidence base to guide the use of devices in children.

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Posterior Cervical Arthrodesis in Children

17

Daniel J. Hedequist and Anthony Stans

Introduction

Posterior fusion and instrumentation for pediatric cervical spine problems may be performed for deformity or instability that is related to a wide variety of pathologic processes from trauma to congenital anomalies to connective tissue disorders. Classically, fusions have been aided by placement of internal fixation, which historically had been done via a variety of cable or wiring techniques. These techniques may be done by passage of cables through or around the spinous process, through the facet joints, or underneath the lamina. The pediatric cervical spine is inherently osteogenic, making wiring successful most of the time, although biomechanically wiring techniques are not stable in multiple planes of motion. Application of a halo may be used as a backup measure for stability either in younger children or in cases where wiring techniques alone are not sufficient for stability until fusion ensues.

Rigid screw-rod fixation has been described by many authors for treatment of pediatric cervical spine problems [1–4]. Techniques that had been developed for adult patients have been adapted and are especially useful, given the disorders encountered in children. The craniocervical junction and atlantoaxial complex are frequent sites of pathology in the pediatric population and are less likely to fuse than the subaxial spine, given the inherent ligamentous laxity at those areas and the greater surface area required to obtain successful fusion. The use of screw-rod constructs is feasible in most children; however, there are constraints to using modern screw techniques based on anatomical limitations, which are present throughout the cervical spine. This chapter focuses on the use of spinal instrumentation in children.

Preoperative Considerations

Successful surgical treatment requires a thorough understanding of the patient and preparation for different instrumentation options. Plain radiographs remain a standard part of the diagnostic evaluation of children with cervical spine problems and are required in all patients not only to diagnose the condition but also to have a gross understanding of the overall anatomy and alignment of the cervical spine. Flexion-extension radiographs are generally useful to understand stability at both the area being operated on as well as to avoid missing instability that may be present in junctional regions. This is paramount in patients with congenital anomalies and Klippel-Feil,

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where instability may be present adjacent to congenitally fused segments of the cervical spine.

Magnetic resonance imaging is required in most patients, especially in order to assess the cervical cord. MRI is useful in documenting areas of neurologic compression as well as documenting the presence of signal changes in the cord that may be present as a result of cervical instability. The use of MRI is important in tumor cases to understand the anatomical soft tissue extension of the tumor. MRA (angiography) can be a useful adjunct in patients with severe instability, connective tissue disease, or tumor load encroaching on the vertebral artery. MRA is also useful in trauma patients with foraminal injuries in order to assess the possible injury to the vertebral artery.

Computed tomography scanning remains the most important study when evaluating a patient prior to surgery. The anatomical detail of the bony architecture is important for determining the ability to place screws. Fine-cut CT scanning (1 or 2 mm), done under a pediatric protocol to minimize radiation, should be utilized in all younger patients undergoing surgery with potential screw fixation and should include axial and sagittal reformats. Three-dimensional reconstructions are helpful to completely understand the anatomy prior to surgery.

Operative Considerations

Prior to surgical incision, there are a few factors that must be addressed, the first being the stability of the spine for intubation and for positioning. Intubation techniques for patients with disorders requiring surgery can range from standard intubation to awake fiber-optic techniques. Understanding the stability of the spine and having a discussion with anesthesia prior to the operating room is critical to avoid unnecessary harm to the patient or delay in the procedure. Once intubation has been completed and appropriate lines have been placed, then the patient is ready for positioning, which is important for safety and to facilitate intraoperative imaging. Patients with significant instability should have a halo crown and vest placed for both turning and positioning. Once the halo and crown has been placed, they can then be safely turned prone without worrying about losing a stable position of the neck. Standard pin placement can be done if the halo is being used postoperatively; if it is only being used as a positioning device, then placing the anterior pins in the anterolateral hairline is useful for cosmetic concerns while still affording stability. The posterior portion of the vest may be removed and the surgery performed with the patient wearing the crown and anterior portion of the vest. The struts may be loosened to a small extent and patient placed in a gentle amount of traction for stability. Other options for positioning include a Mayfield headrest or cervical tongs.

Positioning the patient with arms tucked down at the sides and taping the shoulders creates room for C-arm positioning and allows for imaging of the cervical spine (Fig. 17.1). Care must be taken not to tape the shoulders down with too much force, as this can lead to brachial plexus palsy. Fluoroscopic setup should be done in order to have adequate visualization of the spine, as well as having monitors placed that are in a surgeon-friendly location while placing screws. Checking fluoroscopic views and making sure adequate visualization is possible should be confirmed prior to prepping and draping, in order to assure technical feasibility.

Neuromonitoring is required in all cases, and both motor-evoked potentials (MEP) and sensory-evoked potentials should be used. Continuous EMGs are useful as well. Baseline signals should be obtained prior to positioning with the patient lying supine on the hospital cart and then rechecked after positioning to confirm that the spinal cord is not compromised during the positioning process and to confirm that adequate signals are present prior to initiation of the surgical procedure.

C1 Screw Fixation

The lateral mass of C1 is a quadrangular area of the bone, which is optimal for screw purchase given its size, even in the youngest of patients [5–7]. The lateral mass is bordered by the spinal canal medially, and the anterolateral region is bordered by the vertebral artery, which descends out of the cranium and traverses along the

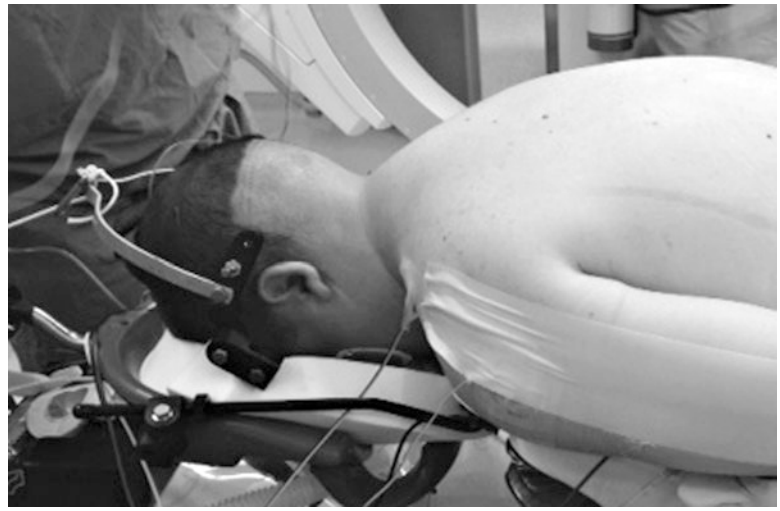

Fig. 17.1 Intraoperative photograph documenting correct positioning. Note the position of the hands with the shoulders taped down to allow for adequate fluoroscopic visualization during screw fixation

superior aspect of the lateral ring of C1. Morphologic studies utilizing computed tomography scans to describe the anterior-posterior, medial-lateral, and cranial-caudal dimensions have been performed [8, 9]. In a recent study, Geck found that the lateral mass is able to accept a screw in children even as young as 2 years of age, with predictable anatomical parameters [9].

Placement of lateral mass screw is completely dependent on the exposure of the lateral mass. The C1 lateral mass entry point lies significantly more anterior than screw entry points for C2 and subaxial screws, thus making dissection more difficult. Paramount to exposing the starting point for the screw is identification and bipolar cautery of the venous plexus, which is diffused and must be controlled for adequate visualization (Fig. 17.2). The lateral mass entry point may be found by dissecting out the inferior border of the posterior ring of C1 and following this laterally as it turns and heads ventral into the lateral mass of C1. The vertebral artery lies on the superior surface of the ring of C1, and as surgical dissection is carried laterally, it is imperative to remain on the inferior border to avoid vertebral artery injury. The starting point for a C1 lateral mass screw is directly beneath the intersection of the arch and the lateral mass, occasionally slightly medial, depending on the anatomy seen on computed tomography scan (Fig. 17.3). The C2 dorsal nerve root may overlie the lateral mass screw starting point, and there is no consensus among surgeons as to whether the nerve root should be resected or retracted out of harm's way [10].

Once the starting point has been exposed, the trajectory of screw placement is fairly predictable. Prior to drilling, we recommend using a burr to create a starting point which will aid in controlling the drill bit during initiation of the drill hole. Occasionally, the inferior surface of the ring of C1 must be burred away to allow for placement of the drill bit. Burring a starting point is also helpful because bone at the entry point tends to be dense cortical bone where the ring intersects the lateral mass. Creating a starting point with a burr avoids the need for excessive anterior pressure when using the drill. The drill bit should be guided in the cranial-caudal direction by fluoroscopy, using the middle of the C1 anterior ring projection, which is clearly outlined on the lateral fluoroscopy views. The drill bit should be aimed approximately 15° cephalad and toward the middle 20–40% point of the anterior ring projection. Aiming superiorly to that projection risks placement of the screw into the occiput-C1 articulation [11]. Medial-lateral trajectory should be inward 15° [11]. Anterior and laterally, the vertebral artery is at risk and thus the path is always directed medially.

The depth of the drill bit should not pass the posterior border of the anterior projection of the ring of C1, as the screws are not bicortical. Bicortical perforation puts the hypoglossal nerve and internal carotid artery at risk, and so fluoroscopy should be

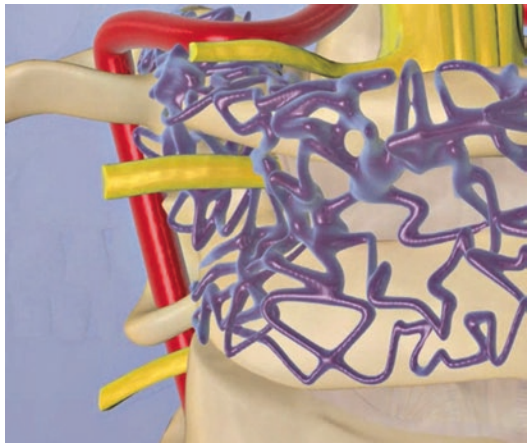

Fig. 17.2 Schematic representation of the diffuse venous plexus encountered during exposure for C1 and C2

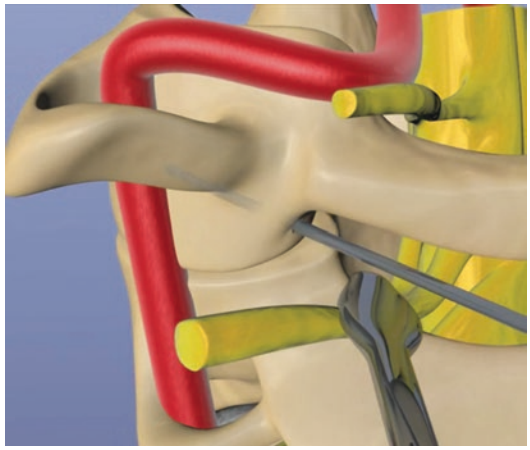

Fig. 17.3 Schematic representation of the starting point for C1 lateral mass screws. Note the retraction of the C2 nerve root and the position of the starting point relative to the C1 arch

used to avoid over-penetration [12]. Once drilling has been done, tapping the dense cortical bone at the starting point is crucial to create a secure entry into the bone and to direct the screw down the correct path. Screw length within the lateral mass is measured with a depth gauge, and then, depending on the size of the patient, 6–10 mm of additional length must be added to account for the anterior position of the C1 lateral mass starting point compared to C2 and other screws. Thus, the tulip of the C1 screw sits out of the bone 6–10 mm in order to be aligned with other screws in the construct (Fig. 17.4). There are

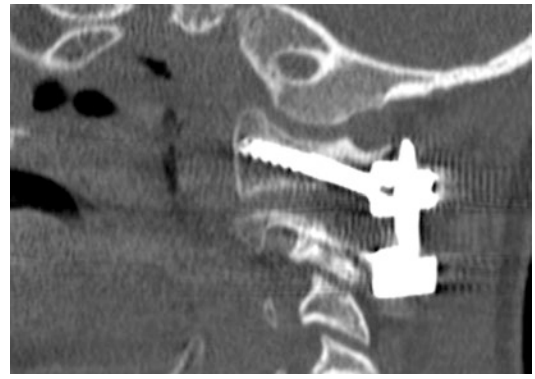

Fig. 17.4 Sagittal CT cut of a patient who had undergone C1-C2 instrumentation. Note the position of the C1 screw underneath the C1 arch, the partially threaded screw used to avoid C2 nerve irritation, and the distance of the tulip from the screw entry point

partially threaded screws available in order to have a smooth shank resting against the C2 nerve root, theoretically minimizing potential neuralgia complaints in patients where the dorsal C2 nerve root has been preserved during exposure.

C2 Screw Fixation

There are multiple screw options for fixation at C2, depending on the individual anatomy of the patient. Screw fixation to C2 is important in the pediatric population, as it is a key anchor point for fixation in craniocervical constructs as well as in atlantoaxial instability [2, 13–15]. The isthmus of C2 is a tubular region of bone that medially is bordered by the spinal cord and has a variable relationship to the vertebral artery, which plays a major role in dictating screw choice. Placement of "pars" screws and "pedicle" screws is frequently interchanged, as both screws share the isthmus of bone but differ in starting points as well as trajectories. The vertebral artery has a variable course, and the anatomy must be determined by preoperative studies, as a higher riding or more medial course of the vertebral artery may make screw placement hazardous (Fig. 17.5) [15].

Paramount to placement of a C2 pars or pedicle screw remains the surgical dissection of the dorsal and medial borders of the C2 isthmus.

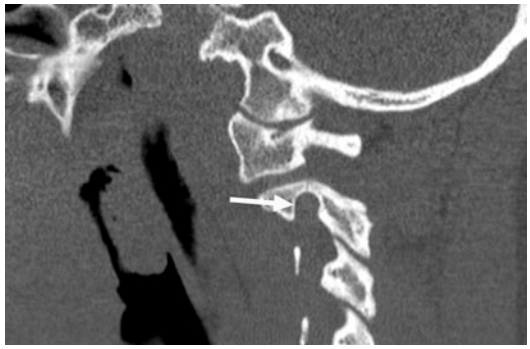

Fig. 17.5 Sagittal CT cut of a patient undergoing preoperative evaluation of cervical anatomy. Note the *arrow* pointing at the vertebral artery representing the high course in relationship to the C2 isthmus prohibiting transarticular screw placement

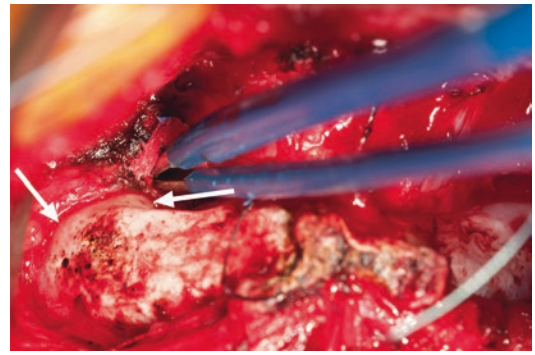

Fig. 17.6 Photo of intraoperative dissection of the C2 isthmus. The *arrows* outline the pars, and the bipolar device seen in the picture is at the proximal C1-C2 joint

This is done by following the lamina down to the region of bone between the C1-C2 facet and the C2-C3 facet and then dissection proximally and medial on the bony surface while coagulating the venous plexus (Fig. 17.6). Complete visualization of the isthmus will allow for protection of the medial canal while allowing for direct visualization of the medial-lateral trajectory of the screw. The rostral-caudal direction of the screw must be done with fluoroscopic visualization. Once adequate visualization of the isthmus is achieved, then the starting point and trajectory will vary, dependent on the screw types. A "pars" screw starts just rostral to the C2-C3 joint and is directed in a rostral direction toward the C1-C2 joint, with medial-lateral direction being straightforward thru the isthmus. A "pedicle" screw has a more cranial and lateral starting point, with the direction hugging the medial isthmus and directed into the C2 body on lateral fluoroscopy. Both screws must be placed using direct visualization of the pars, as well as lateral fluoroscopic imaging, to avoid inadvertent medial penetration of the canal and to avoid vertebral artery injury (Fig. 17.7). After drilling the screw path, the path must be probed and tapped prior to placing the screw (Fig. 17.8). Vertebral artery injury would be manifested by profuse bleeding, and placement of the screw would be the appropriate step in order to tamponade the injury. If vertebral artery injury is suspected on the first side, then no attempt should be made to place a screw on the other side, as injury to both vertebral arteries would be catastrophic.

Placement of intralaminar screws into C2 has been described in clinical series, in morphometric studies, and in biomechanical studies [16–18]. These screws are easily placed, have good construct strength, do not have the potential for vertebral artery injury, and are feasible in the majority of pediatric and adult patients. Intralaminar screws have a starting point at the base of the contralateral spinous process. Typically, bilateral screws are placed in a crossing manner, so the starting point must be more rostral on one side and caudal on the other. The starting point is created using a burr, and then an awl may be used to cannulate down the intralaminar space in a trajectory guided by the dorsal lamina. Error in screw placement can be minimized by placing a small freer on the undersurface of the C2 ventral lamina as a guide (Fig. 17.9). The path of the awl can then be tested with a probe, followed by tapping the starting region and placing the screw. Screw length can be measured, but should be estimated to a reasonable degree by the preoperative studies. CT analysis of children has revealed that the C2 intralaminar bone is of reasonable width and length to allow for screw placement in the majority of children (Fig. 17.10) [9, 19]. The tulips of

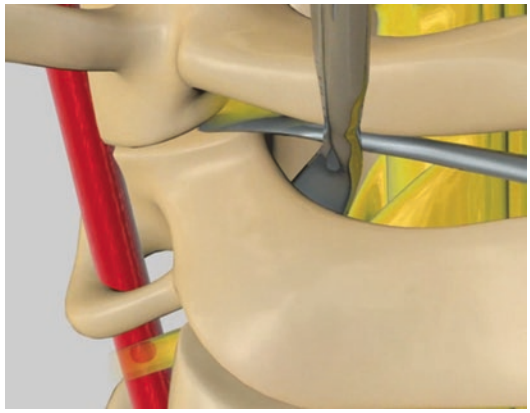

Fig. 17.7 Schematic representing identification and visualization of the C2 isthmus. Intraoperative use of a freer elevator on the medial pars allows complete identification of the anatomy and is paramount to C2 screw placement

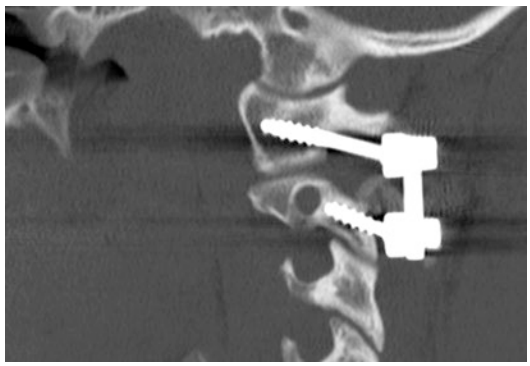

Fig. 17.8 CT scan of a patient who underwent C1-C2 Harms construct. Note the pars screw at C2 and how it stops short of the vertebral artery

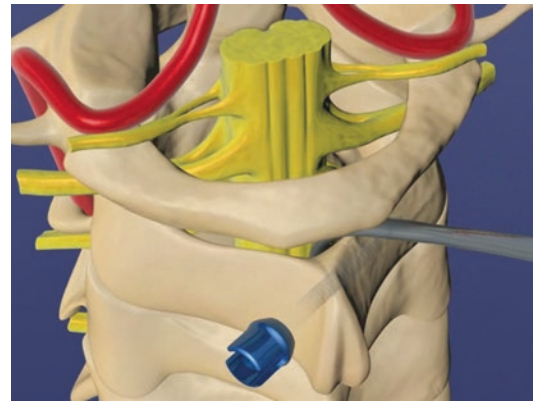

Fig. 17.9 Schematic representation of a translaminar screw at C2. Note the freer elevator placed to help determine the ventral aspect of the C2 lamina

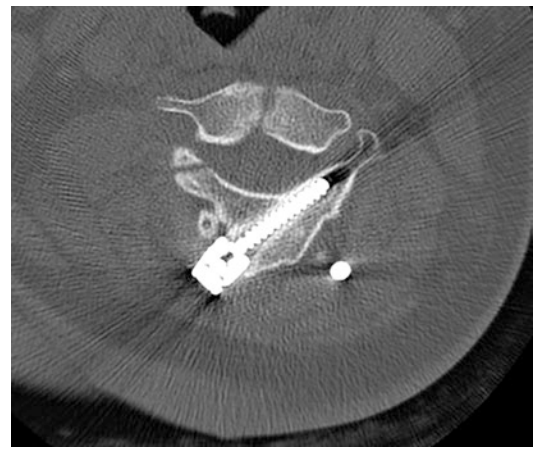

Fig. 17.10 Postoperative CT analysis of C2 intralaminar screw position depicting containment and appropriate length and position

the screws lie posteriorly next to the spinous process, and so offset connectors may be needed if connecting rostral or caudally to other screws which are more lateral, given the starting points of lateral mass screws.

C1-C2 Transarticular Screws

The use of transarticular screw has been shown to be efficacious in carefully selected patients [1, 20]. These screws obtain excellent biomechanical strength, as they utilize the bony isthmus of C2 and cross the C1-C2 articulation into the lateral mass of C1. The screw path must be meticulously studied on preoperative imaging tests, as there is variability regarding the path of the vertebral artery [21]. The sagittal cuts give an idea of the vertebral artery course in relationship to the isthmus, and the coronal CT cuts can give an idea of the direction of the C1 landing point. Multiple studies have shown the anomalous nature of the vertebral artery make placement of screws unsafe in many patients. Some patients have variable anatomy, making screw placement possible on one side and not the other. The C1-C2 joint must

be reduced for correct placement of transarticular screws, and if the joint is either anomalous or the joint is not reduced (as in rotary subluxation), then other fixation methods should be used.

Once the preoperative scans demonstrate anatomy adequate for safe screw placement, exposure is performed by meticulously dissecting the isthmus of C2 and the C1-C2 relationship. Visualization of the isthmus is paramount for correct screw placement, just as when placing a C2 pars screw which has the exact same trajectory but stops short of the C1-C2 joint. Dissection of the C1-C2 joint and the ability to put a freer elevator in the joint is a key step when placing transarticular screws. The initial step in transarticular screw placement is positioning a guidewire across the C1-C2 joint, and correct guidewire position is confirmed by palpating the guidewire crossing the joint (Fig. 17.11).

The preferred method of screw placement is by first placing guidewires across the reduced joints. The starting point is created using a burr and is just rostral to the C2-C3 joint. The rostral-caudal direction of the screw is guided by the lateral fluoroscopic images, with the trajectory aiming for the midline of the outline of the middle of the C1 anterior ring. Placing the screw above or below this risks malposition. Frequently the angle of ascent of the screw path dictates that distal stab wounds are required in order to achieve the optimal trajectory for screw placement. Direct visualization of the isthmus will guide medial-lateral orientation with the rostral direction being guided by fluoroscopy. Correct position of bilateral guidewires is confirmed by palpating the wires in the facet joints and by checking the AP fluoroscopic view to assess correct position in the C1 lateral mass. After measuring for screw length, the guidewires are over-drilled and the screws placed (Fig. 17.12). It is best to drill one side first and place that side screw first, in the rare event that injury to the vertebral artery occurs. If suspected vertebral artery injury occurs, then placement of the screw should be performed to tamponade bleeding, and a screw should not be placed on the contralateral side to avoid the possibility of bilateral vertebral artery injuries [22].

The clinical and biomechanical studies do suggest that placement of one screw is efficacious if either the anatomy permits only single-screw placement or if difficulty is encountered in placing a screw (Fig. 17.13). The current transarticular screw sets contain 4.0 mm screws and do not have a tulip, so if the screws are being used as part of a screw-rod construct, then screws from standard cervical implants can be used; however, these screws are not cannulated. When using a screw-rod construct, it is useful to use the can-

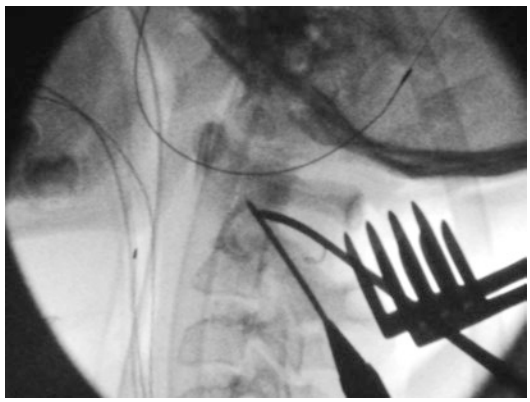

Fig. 17.11 Intraoperative fluoroscopic view demonstrating correct trajectory of guidewire placement for transarticular screw placement. Not the freer elevator in the C1-C2 joint allowing for palpation of the guidewire

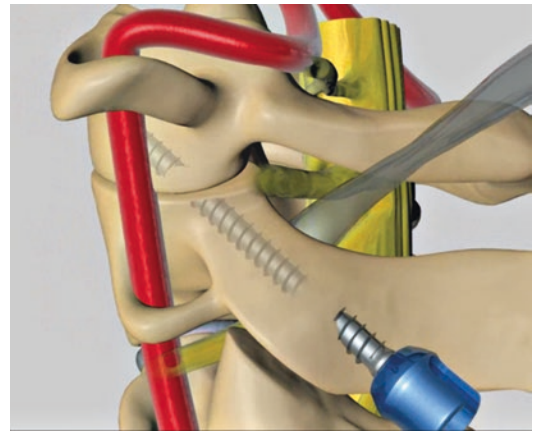

Fig. 17.12 Schematic representing correct placement of a transarticular screw. The freer elevator is placed on the medial pars as protection and as a guide to correct screw trajectory

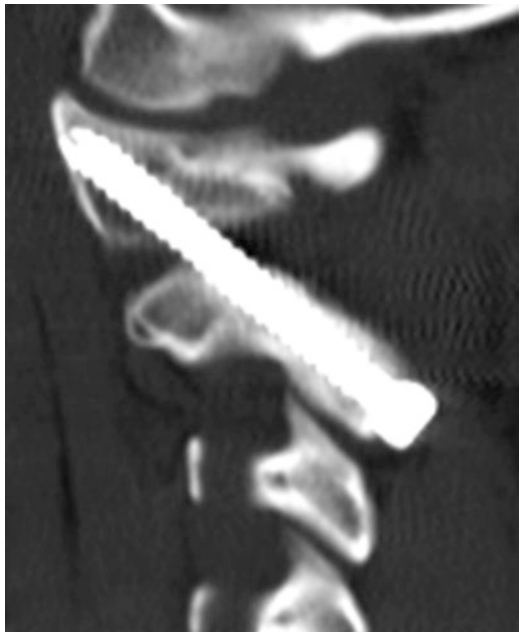

Fig. 17.13 CT scan documenting correct position of a C1-C2 transarticular screw

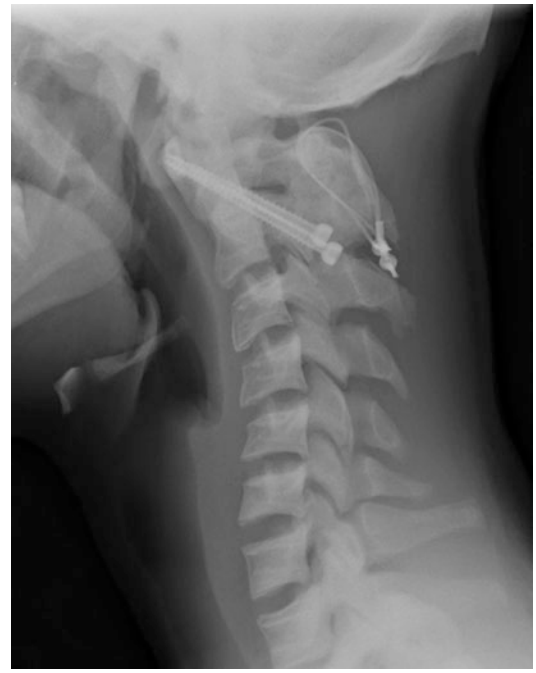

Fig. 17.14 Lateral radiograph 1 year post-transarticular screw placement and cable grafting of structural iliac crest for a patient with osteopetrosis and an odontoid nonunion. Note the radiographic evidence of fusion between the posterior elements of C1 and C2

nulated system for guidewire placement, drilling, and tapping, followed by removal of the guidewire and placement of a noncannulated screw with a tulip. Fusion rates as well as biomechanical construct stiffness can be improved with the addition of cable grafting a structural graft between C1 and C2.

C1-C2 Cable Fixation

Cable fixation at C1-C2 can be done either as a primary means of fixation or as a supplemental means of fixation and fastening down a structural graft between C1 and C2. The term "cable" refers to titanium cables, which can be tensioned; wiring in this text refers to stainless steel wiring techniques. Preference will be given to cabling techniques because the titanium cables are MRI compatible and compatible with titanium screw-rod instrumentation. Biomechanically, cable grafting alone is not as inherently stable as screw constructs in all planes of motion. However, in very young or small patients, the use of cables may be the only technical option. In addition, using posterior structural bone graft secured with cables has been demonstrated to improve stability and enhance fusion in patients who are undergoing screw fixation at C1-C2 (Fig. 17.14) [23].

Cables are carefully passed beneath the posterior lamina and posterior arch of C1. Care must be taken to evaluate the preoperative anatomy to make sure C1 is not bifid, as this greatly affects cable stability and increases the risk of pullout, especially if cables are being used as a primary form of fixation rather than for augmenting screw fixation. The ring of C1 is completely exposed, with care taken laterally and superiorly to avoid potential vertebral artery injury. Complete subperiosteal exposure will allow placement of a fine right angle clamp underneath the lamina. Safe passage of #0 silk suture can then be performed which, in turn, is used to pass cables beneath the posterior ring of C1. Multiple options exist with regard to cables being secured to C2 and include

sublaminar passage, passage around the spinous process, and passage through the spinous process. Passage of cables underneath the lamina of C2 puts the spinal cord at greater risk than at C1 because there is much less space available for the cord at C2 compared to C1.

Typically cables are passed on both the right and the left side of the lamina to increase fixation strength. In a classic Brooks-type fusion, the cables are passed on either side of the midline, first under the arch of C1 and then beneath the lamina of C2. Structural iliac crest grafts are then wedged between C1 and C2 and secured with cables. Care must be taken not to excessively tension the cables and over-reduce C1 on C2. Using bone graft of sufficient size will help prevent over-reduction.

The bifid spinous process of C2 is a large structure through which cables can be passed. In a Gallie-type fusion, the cables from underneath the arch of C1 are passed through the spinous process of C2 and are used to secure a horseshoe-shaped structural graft between C1 and C2. Cables are passed through the C2 spinous process by making a small burr hole on either side of the spinous process and using a towel clamp to open up the hole. The cables are then easily threaded thru this channel.

Complications

The inherent risks of using modern instrumentation primarily relate to the nearby anatomical structures. No large clinical series reporting implant-related complications in children currently exists, but the use of screw fixation does have potential risk. The most commonly reported complication in pediatric cervical spine patients treated with screw fixation is vertebral artery injury. The vertebral artery exits the cranium and courses along the superior aspect of the lateral ring of C1 and then turns rostral with an intimate relationship to the lateral mass of C1 and foramen transversarium of C2 (Fig. 17.15). Hence, vertebral artery injury is described mostly commonly in upper cervical fixation, notably with transarticular screws [22]. Risk of this complication can

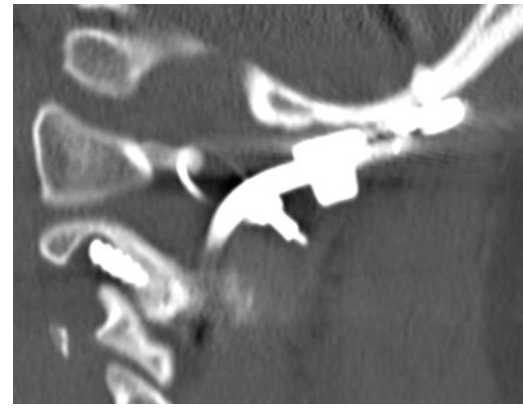

Fig. 17.15 CT scan documenting malposition of a C2 pars screw with impingement into the vertebral artery

be decreased by careful preoperative CT imaging and study of each patient's anatomy. Vertebral arteries which course more medially at C2 or have a high riding relationship to the inferior aspect of the C2 isthmus are at risk with screw placement. If the anatomy on the preoperative imaging is favorable for screw passage through the isthmus, then surgical dissection of the isthmus is paramount to minimizing aberrant screw paths. The dissection of the isthmus for placement of either transarticular screws or C2 screws is also important to avoid medial malposition of the screw. Fortunately, pseudoarthrosis following instrumented cervical fusion in children is uncommon, but when it occurs nonunion is often manifested by implant failure or breakage (Fig. 17.16).

Neurologic complications related to screw placement are rare and, in multiple series in children, have not been described. Penetration of screws into canal is a rare complication and can be avoided with meticulous surgical technique [12, 24]. The placement of subaxial lateral mass screws requires an understanding of the anatomy and familiarity with correct surgical technique [25]. Placement of screws in a bicortical manner is not needed for fixation to be adequate. Anterior penetration as well as lateral penetration of the screws puts the exiting nerve root at risk as well as the vertebral artery at risk. These risks can be minimized by probing the drill path to assure containment prior to placing the screws. The use

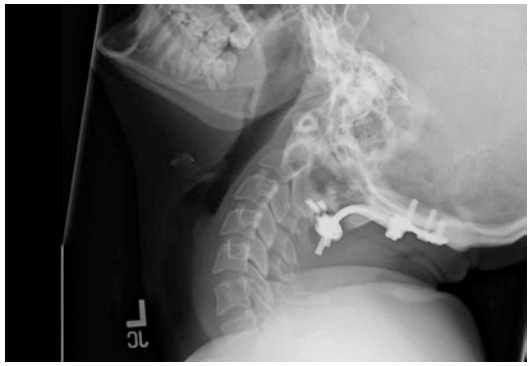

Fig. 17.16 CT scan of a patient with implant failure and a pseudoarthrosis. Not the broken C2 translaminar screws

of fluoroscopy during screw placement is helpful to ensure that screws are accurately placed within the small pediatric lateral mass. In addition to neurologic injury, inaccurate screw placement can lead to adjacent facet joint penetration and late pain or arthritis.

Subaxial Screw Fixation

Instrumenting the subaxial cervical spine in children is more challenging than in adult patients because children have smaller anatomic structures, increased segmental motion, increased ligamentous laxity, and decreased bone ossification [3]. Many pediatric patients undergoing cervical arthrodesis have congenital or genetic abnormalities, resulting in abnormal anatomy. Cervical spinal pathology affecting C3 to T1 in children and adolescents differs from disorders affecting the pediatric cervical spine from occiput to C2. Whereas disorders affecting the occiput to C2 are often congenital or developmental in their etiology and tend to affect younger children, pediatric cervical spine pathology from C3 caudal tends to affect older children and adolescents and is more often the result of acquired disorders such as trauma [26]. Instrumentation of the caudal cervical spine may also be indicated in patients where the primary disorder is in the upper thoracic spine, and instrumentation across the cervicothoracic junction is necessary to maintain a well-balanced and stable spine.

Anatomy and CT Morphology

Al-Shamy and coauthors conducted a morphometric analysis for lateral mass screw placement in children and adolescents [8]. Cervical spine CT scans were reviewed in 56 boys and 14 girls ranging in age from 2 months to 16 years. No significant differences were noted between males and females. The largest dimension was the sagittal diagonal measurement from the posterior edge of the inferior facet to the anterior edge of the superior facet. This sagittal diagonal measurement was at least 10 mm at all levels in all children age 4 and older and at most levels in children age 1 and older. Considering that the shortest commercially available screw is 10 mm, nearly all children age 4 and older have cervical spine morphology which will accept lateral mass screws. The mean facet angle decreased and facet height increased moving caudally from C3, suggesting a more steep screw trajectory may be appropriate when placing lateral mass screws in pediatric patients compared to adults. For young children with very small osseous anatomy, non-spinal screws and plates including implants from foot or mandibular systems have been used successfully.

Technique

CT scanning is very useful for preoperative planning to ensure that the patients' lateral mass will accommodate commercially available lateral mass screws. In patients with congenital abnormalities, the CT scan is especially useful for identifying lateral mass screw starting points and for identifying vertebrae with atypical anatomy that will not accept screw fixation. In select cases of congenital deformity or revision surgery, creating a rapid prototype three-dimensional model using CT scan data and a three-dimensional printer is very helpful for understanding abnormal cervical spine anatomy and may facilitate safe and efficient screw placement intraoperatively. In patients undergoing fusion to C2, fine cuts should be performed in this region to demonstrate the relationship between the vertebral artery and pars isthmus [4].

Prior to positioning on the operating table, motor-evoked potential (MEP) and somatosensory-evoked potential (SEP) signals are obtained. EMG monitoring may also be used. Patients are positioned prone on the operating table, with the head secured using either a halo or pinion whenever possible. MEP and SEP signals are again checked prior to proceeding with the operation. A midline skin incision is made, and posterior elements are exposed to the lateral border of the lateral masses using subperiosteal dissection.

A modified Magerl technique is used for screw placement [27, 28]. The medial, lateral, superior, and inferior borders of the lateral mass are defined, outlining a rectangular box. At a point 1 mm medial and 1 mm inferior to the center of the box, a burr is used to create a starting point for the screw. Fluoroscopy can be used in the lateral plane to confirm the correct starting point and to confirm that the drill hole parallels the plane of the facet joint. On low speed, using a stop mechanism to control depth, a drill is used to create a hole angled 30° laterally and 30° cephalad. The stop mechanism is typically set to limit screw hole depth to 10 mm in small children and 14 mm in adolescents. The screw hole is probed to confirm depth and to confirm the hole is contained within bone. Unicortical screws afford sufficient fixation in virtually all patients. The drill hole is tapped and a 3.5 mm diameter polyaxial screw is placed. To allow full utility of the polyaxial screw, the screw heads are not placed flush with the bone surface. If screw hole depth is measured to be 8 mm, then a 10 mm screw is typically used. Screw position and length can then be checked using lateral fluoroscopy. Once all screws are safely placed, a rod of appropriate length is contoured and secured to the screws. The posterior elements are decorticated over the instrumented segment and autologous bone graft is used to achieve fusion. A cross-link is used whenever possible [4] (See Figs. 17.17 and 17.18).

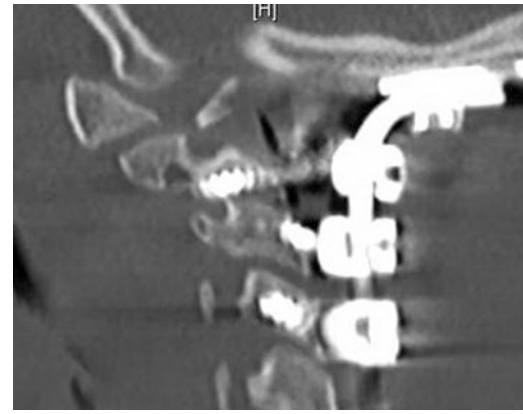

Fig. 17.17 C4 lateral mass screws lateral CT

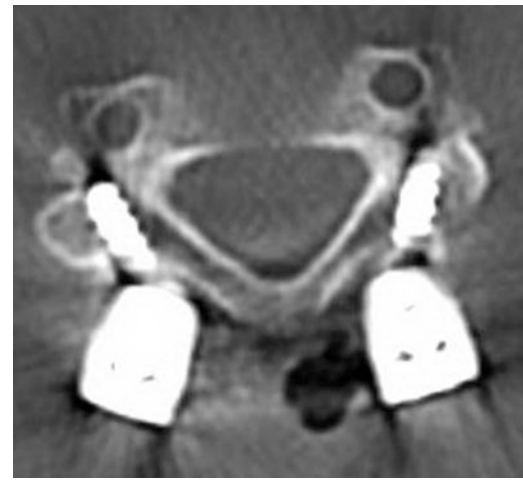

Fig. 17.18 Lateral mass screws postop axial

Clinical Studies

Hwan and colleagues reported their treatment results, in addition to reporting a comprehensive literature review, including posterior instrumented cervical spine fusion in over 880 pediatric patients [29]. Overall fusion rates were high at 94%, but significant differences were identified between fusion techniques using wires or cables compared to screw constructs. Differences were especially apparent in the subaxial surgery patients. While occipital cervical fusion rates were high in both wire (95%) and screw (99%) construct groups, patients undergoing subaxial instrumentation had statistically significant lower fusion rates in the wire (83%) group compared to the screw (99%) construct group. The frequency and severity of complications were statistically higher ($p < 0.05$) in the wire-construct group (54%) compared to the rod-and-screw patients

(15%). Complications in the screw and rod patients included paresthesia, unintended fusion extension, kyphotic deformity, pseudoarthrosis, rod migration, and infection, while complications in wire-construct patients included death, spinal cord injury, quadriparesis, transient radiculopathy, graft resorption, pseudoarthrosis, wire migration, seroma, CSF leak, and mechanical instability. Less external immobilization was needed in patients undergoing rod-and-screw construct instrumentation compared to patients undergoing arthrodesis with a wire construct, with wire-construct patients more likely treated with halo immobilization compared to rod-and-screw construct patients [29].

Cables and Wiring

Cables and wire fixation have been virtually completely replaced by screw-rod instrumentation in the subaxial cervical spine. The most simple posterior cervical wiring technique uses the tension band principle and interspinous wiring. Fixation is not rigid and is most successful when limited to fusion of one motion segment. The ideal indication is the child who has experienced trauma with posterior ligament disruption or facet dislocation.

After identification of the correct fusion level, a drill is used to create a hole toward the base of the spinous process of the upper level to be fused [30]. A 0.8–1.2 mm diameter wire is passed through the drill hole and then around the base of the spinous process of the adjacent lower level to be fused, leaving the interspinous ligaments intact. Posterior elements are decorticated with a high speed burr. An "H" graft of cortical-cancellous autograft may be fashioned to lie beneath the interspinous wire. The wire is tightened by twisting the ends, and cancellous autograft is applied to the decorticated lateral masses.

Complications of posterior wiring technique include wire breakage, cutout, pseudoarthrosis, and extension of fusion beyond intended levels. The technique cannot be used in patients with fractures or pathology that compromises the posterior vertebral arch.

Bone Graft

Autologous bone graft is the gold standard to which other graft materials are compared. Because case numbers are relatively small and confounding variables are numerous, statistically significant comparisons between grafting materials are difficult to make.

The potential benefits of recombinant human bone morphogenic protein are numerous, including decreased blood loss, decreased surgical time, reduced donor site morbidity, elimination of potential infection transmission with allograft, decreased pseudoarthrosis rate, unlimited quantity, and immediate availability. Potential impediments to its use include cost, cancer risk, unintended fusion extension, exuberant bone formation compromising the spinal canal, interaction with exposed dura, local toxicity, and potential effect on distant organs. Because no statistical comparisons are available, the selection of graft material is left largely to the discretion of the surgeon.

Summary

The use of spinal instrumentation in the pediatric cervical spine has allowed for improved stabilization, deformity correction, and fusion of cervical disorders in children. Posterior cervical instrumentation techniques rely on the surgeon's understanding of the anatomy as well as familiarity with different fixation options at the upper cervical spine and lower cervical spine. Complications with instrumentation may be avoided by knowledge of the anatomical structures at risk, and meticulous surgical technique.

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Anterior Cervical Arthrodesis in Children

18

Emmanuel N. Menga, Michael C. Ain, and Paul D. Sponseller

Introduction

Cervical spine arthrodesis is most often used for the treatment of pediatric cervical spine instability, with the majority of patients undergoing an instrumented posterior cervical spinal arthrodesis for instability secondary to traumatic or congenital causes. The main indications for anterior arthrodesis in children are pathology or deformity that is primarily anterior in location, when there is the potential inability to obtain posterior fixation or when bone quality is poor, mandating circumferential fusion. Examples include severe focal kyphosis, postlaminectomy deformity, and neurofibromatosis. Anterior cervical arthrodesis is most commonly used in the adult population for the treatment of cervical disc herniation and myelopathy, but many authors have also reported good clinical outcomes with this procedure in pediatric patients [1–6].

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Anterior cervical spine arthrodesis with plate fixation as an adjunct to posterior spinal arthrodesis may provide more rigid fixation and limit the use of external immobilization. The goals of anterior surgery include decompression, enhancing bony fusion, and restoring alignment while minimizing the risk of neurologic compromise. The standard anterior approach, originally described by Smith and Robinson [7], is most often used for exposure of the anterior cervical spine and is described in detail elsewhere in this textbook.

Indications for Anterior Cervical Arthrodesis in Children

The small vertebral body size, the immature and less ossified bone, and the lack of adequately sized instrumentation are all reasons for the limited use of anterior cervical spine arthrodesis for the management of pediatric cervical spine abnormalities, especially in younger children (less than 8 years old) [2, 3]. However, by age 10, the bony anatomy of the pediatric cervical spine is similar to that in the adult, with similar expected postsurgical outcomes [5, 8]. Although uncommon in the pediatric population, stand-alone anterior cervical arthrodesis may be indicated in children with disc herniation, trauma, cervical spine deformity, tumor, and infections. The number of levels (single- versus multiple-level discectomy or corpectomy) incorporated in the fusion,

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as well as the use of instrumentation, depends on the location and extent of the abnormality and the feasibility of instrumentation. Structural bone graft or cages filled with morselized bone graft may be used to augment the fusion bed and restore alignment. There is limited literature on the use of bone morphogenic protein (BMP) in pediatric anterior cervical surgical arthrodesis; however, given the evidence of potential adverse consequences of airway swelling and compromise in adults, the use of BMP in anterior cervical surgery in children is not recommended.

Disc Herniation

Intervertebral disc herniation is rare in children [9], although intervertebral disc herniation at the unfused segments in pediatric patients with Klippel-Feil syndrome has been reported [10]. The true cause of disc herniation in children remains unknown, with infection and trauma implicated as possible causes [9].

The treatment for pediatric disc herniation is often nonoperative, with surgery indicated for patients with worsening radiculopathy and/or myelopathy. Cervical and thoracic disc herniation in the pediatric population accounts for less than 1% of all surgically treated disc diseases [11]. As in the adult patient, anterior cervical discectomy and arthrodesis is a viable option for the treatment of cervical disc herniation in the pediatric patient. Two case reports were found in the literature: one patient with Klippel-Feil syndrome and symptomatic herniated cervical intervertebral discs underwent a combined posterior-anterior arthrodesis with anterior cervical discectomy, interbody tri-cortical autograft, and a posterior plate [10], and a 5-year-old girl with Klippel-Feil syndrome underwent an anterior cervical discectomy and arthrodesis with autograft [12].

Trauma

Most spinal injuries in children younger than 8 years old occur in the upper cervical spine between the occiput, C1, and C2 vertebral levels. Traumatic cervical spine injuries in children older than 10 years old are similar to subaxial injuries observed in the adult population (C3-C7) and may be treated in a similar fashion.

For pediatric patients, most cervical spine trauma is treated without surgery; for those who require surgical intervention, an instrumented posterior cervical spine arthrodesis is the most common procedure. However, anterior cervical spine arthrodesis can be used as an adjunct to posterior fixation or in isolation. For the surgical treatment of traumatic cervical spine lesions, Shacked et al. [13] recommended the anterior approach when possible, which has the advantages of excellent anterior column visualization, effective repair and stabilization, early patient mobilization, and potential long-term neurologic improvement in patients with deficits due to canal compromise. The injuries in their series involved the anterior column and ranged from severe hyperflexion injury with crush fracture and avulsion of the vertebral body, fracture-dislocation of the vertebral body with involvement of the posterior elements and the disc, to injuries that caused major anatomic deformity of the cervical spine with cord compression.

Anterior cervical arthrodesis has been successfully used in pediatric patients older than 10 years of age. In a case series and review of the literature, Eleraky et al. [14] described 30 patients who underwent surgical management of cervical spine injury for instability, neurologic deterioration, irreducible fracture/dislocations, and failed nonoperative treatment. Of those 30 patients, 18 patients (10–16 years old) had anterior cervical spinal fixation and arthrodesis, including odontoid screw fixation, corpectomies, and anterior cervical discectomy and arthrodesis. Those authors reported solid fusion at the 6-month follow-up with no report of surgery-related deaths or complications. We have successfully used instrumented anterior spinal arthrodesis with iliac crest structural autograft in our trauma patients requiring decompression, stabilization, and fusion (Fig. 18.1a–d).

In the more immature pediatric spine, the smaller vertebral body size and wedge-shaped vertebra render anterior cervical spine arthrodesis even more challenging than in the adolescent patient. There are no current large series of patients reported on in the

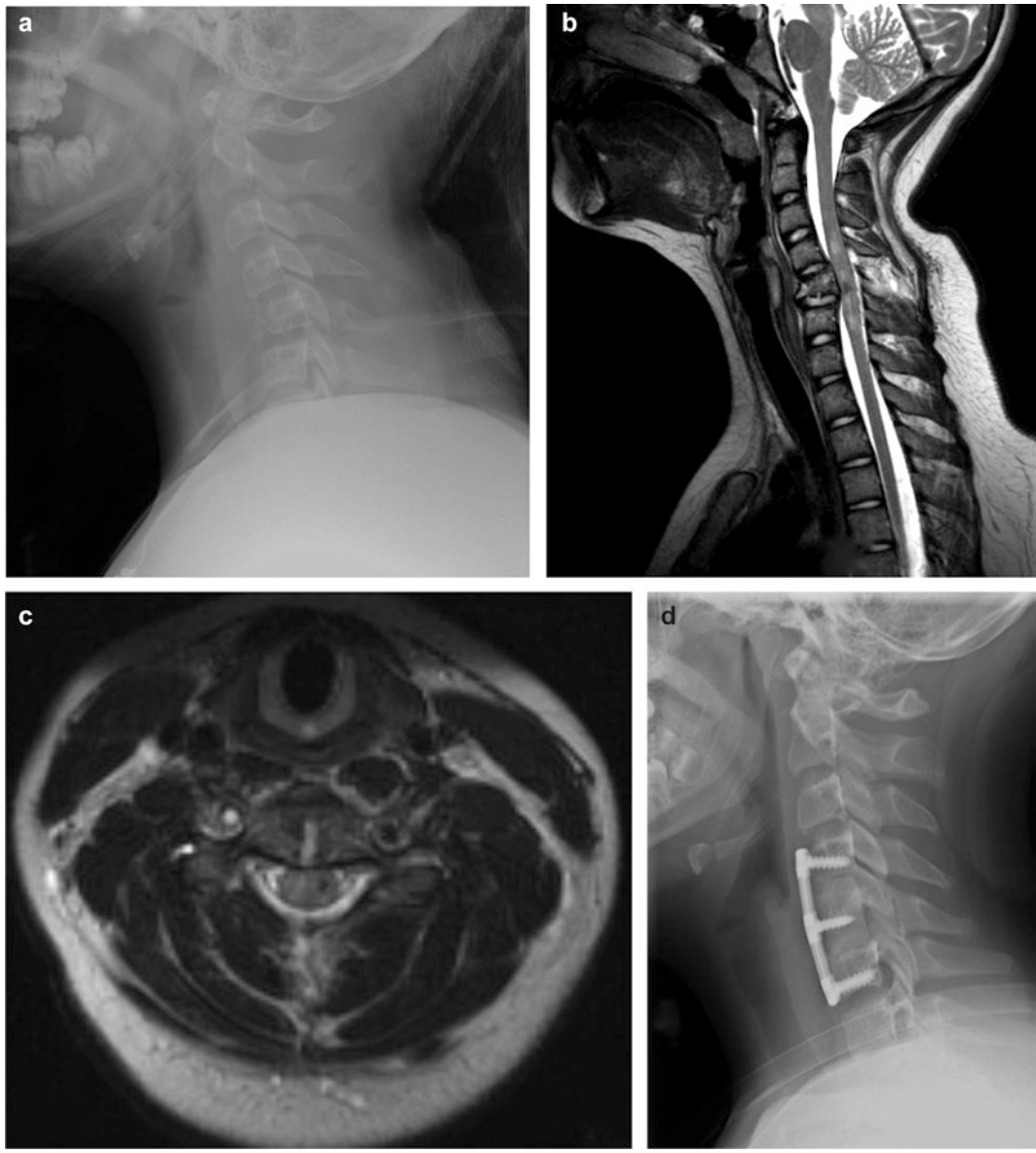

Fig. 18.1 (**a**–**d**) Imaging studies of a 12-year-old girl sustained a burst fracture C5 with quadriplegia and underwent a C5 corpectomy, adjacent discectomies, and spinal decompression with instrumented anterior spinal arthrodesis using iliac crest structural autograft. (**a**) Lateral preoperative radiograph shows the C5 burst fracture with kyphosis at the fracture level. (**b**) Preoperative sagittal and (**c**) axial magnetic resonance images indicate cord contusion and compression, fracture, posterior ligamentous injury, and right vertebral artery occlusion. (**d**) Postoperative lateral radiograph at 3-year follow-up shows anterior fusion with good sagittal alignment

literature, given the rarity of this injury pattern in smaller children. Li et al. [3] described the successful use of a craniofacial miniplate with interbody allograft as an anterior cervical fixation device, combined with a posterior C3-C4 arthrodesis, iliac crest autograft, and sublaminar bone sutures in a toddler with a grossly unstable C3-C4 injury after a motor vehicle accident. They reported good cervical alignment and fusion with stable neurologic function at 2-year follow-up.

Although the majority of odontoid fractures in children and adolescents are usually successfully treated with external immobilization via a halo vest orthosis, there are a limited number of patients with type II fractures who may require surgical treatment. Wang et al. [15] described anterior cervical arthrodesis of odontoid fractures with single-screw fixation. In their series of 16 patients with atlantoaxial instability, they reported successful screw treatment of three children (3, 10, and 14 years old) with unstable type II odontoid fracture after trauma from motor vehicle accidents. They reported no major complications; one patient (age 14 years) experienced dysphagia that completely resolved in 2 weeks.

Cervical Spine Deformity

Cervical spine deformity in the pediatric population has been associated with a variety of disorders including trauma, dysplasia, congenital spinal anomalies, and iatrogenic disorders [16]. There is limited literature on the anterior surgical management of pediatric cervical spinal deformity. Anterior, posterior, and combined anterior and posterior surgical approaches have been described for the management of various cervical spinal deformities [17, 18].

Anterior cervical surgery may include anterior release by single or multilevel discectomy with interbody grafting. Single or multilevel corpectomy with strut grafting or a cage may also play a role in the treatment of cervical spinal deformity. Deburge and Briard [19] reported a successful 25° correction of a cervical spine deformity in a 14-year-old girl with a C7 hemivertebra. This correction was achieved through staged procedures, including a single-level anterior partial C7 hemivertebra excision with discectomy and arthrodesis with an anterior plate, followed by a posterior procedure [19].

Francis and Nobel [1] described various approaches for the treatment of cervical kyphosis in children, including (1) traction and a single-stage anterior release with strut arthrodesis for patients with loss of posterior elements and (2) preoperative traction, followed by posterior osteotomies with intraoperative traction and a staged second procedure for an anterior release and strut arthrodesis for patients with intact posterior elements. They reported satisfactory outcomes in six patients with primary posterior incompetency secondary to previous laminectomies and in three patients with anterior insufficiency (one patient with multiple anterior subluxations, one with dislocations secondary to Larsen's syndrome, and one with cervical spondyloptosis). All patients showed fusion 18 weeks after surgery, with improved functional levels [1]. We have found in our experience the anterior approach is helpful in the management of cervical kyphosis, especially in children with neurofibromatosis-1 (Figs. 18.2a, b and 18.3a–d).

Tumor and Infection

The surgical approach for the treatment of cervical spinal tumors and infection depends on the location and extent of the pathologic process. Anterior cervical surgical stabilization and arthrodesis after resection and debridement of the abnormality remain viable options in the pediatric population. Despite the limitations in treatment, most commonly attributed to the small size of the immature pediatric cervical vertebral body, multiple studies have reported successful treatment of cervical spinal tumors and spinal epidural abscess in the pediatric cervical spine through anterior cervical decompression or debridement and arthrodesis with or without instrumentation [4, 6, 17, 18, 20]. Treatment should be tailored to each individual patient, with the goal of achieving adequate excision or debridement, neurologic decompression, and anatomic spinal alignment with stability. We have successfully used anterior arthrodesis to treat adolescents with symptomatic eosinophilic granulomatosis involving the anterior, middle, and posterior elements (Fig. 18.4a, b) as well as for the treatment of a symptomatic osteoid osteoma (Fig. 18.5a–e).

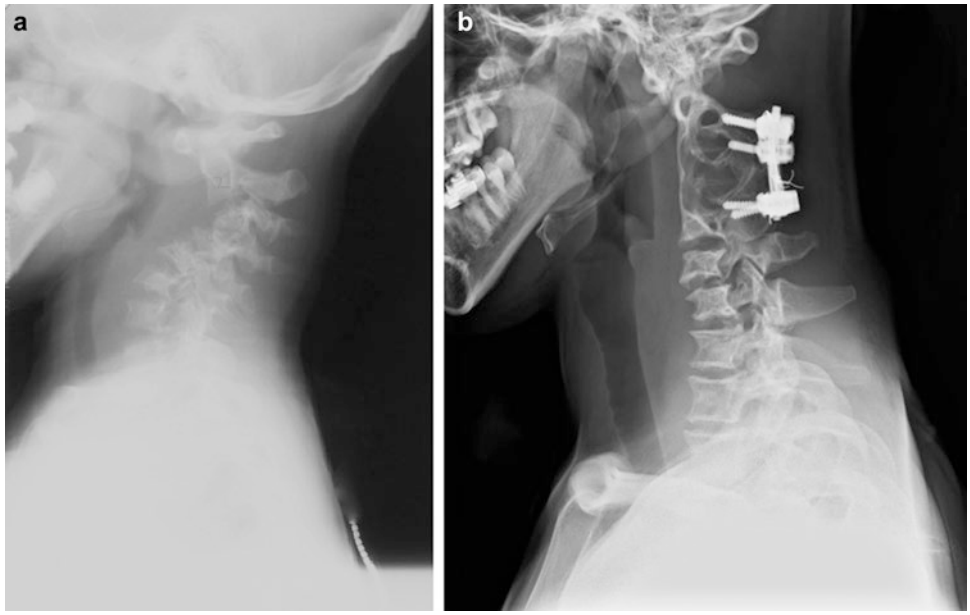

Fig. 18.2 (**a**, **b**) Imaging of a 6-year-old girl with neurofibromatosis-1 and severe progressive cervical kyphosis who underwent an anterior noninstrumented C2-C5 arthrodesis after partial C4 and C5 corpectomy with structural rib autograft supplemented with a C2-C5 instrumented posterior spinal arthrodesis, autologous iliac crest bone graft, and a halo Minerva orthosis. (**a**) Lateral preoperative cervical spine radiograph. (**b**) Lateral 10-year postoperative radiograph shows maintenance of appropriate sagittal alignment with solid anterior and posterior arthrodesis

Approach

The standard anterior approach applies for most patients and is described in the chapter covering surgical anatomy. The approach is more extensile distally (to T2) because of smaller neck muscles and ease of retraction. Micheli and Hood [21] described a combined anterolateral cervical and posterior transpleural transthoracic approach that allows exposure from C3 to the T9 for management of severe structural deformities of the cervicothoracic spine. They reported successful fusion, with adequate correction of kyphoscoliosis, in all six of their pediatric patients undergoing the procedure. Mulpuri et al. [22] also reported successful access to the cervicothoracic junction in six children with severe cervicothoracic kyphosis requiring instrumentation. They used an anterior neck approach and a sternal splitting approach that allowed access from C5 to T6.

Materials

Bone Grafting and Biologics

Autologous bone graft is the most commonly used graft for anterior cervical surgical arthrodesis in children. Multiple studies have reported successful arthrodesis using iliac crest autograft, rib autograft, and fibular strut autograft [4, 6, 17, 18]. However, donor-site morbidity, including hematoma, acute and chronic pain, injury to the lateral femoral cutaneous nerve, peroneal nerve injury, motor deficit, stress fracture, and the quality (rigidity and strength) of the bone graft, should be taken into consideration when contemplating the use of autograft. The osteogenic, osteoinductive, and osteoconductive nature of autograft as well as the minimal risk of disease transmission and infection renders it a more favorable option than allograft.

Nevertheless, allograft bone graft remains a viable option, with fusion rates in nonsmokers comparable to that of autograft [7]. The integrity

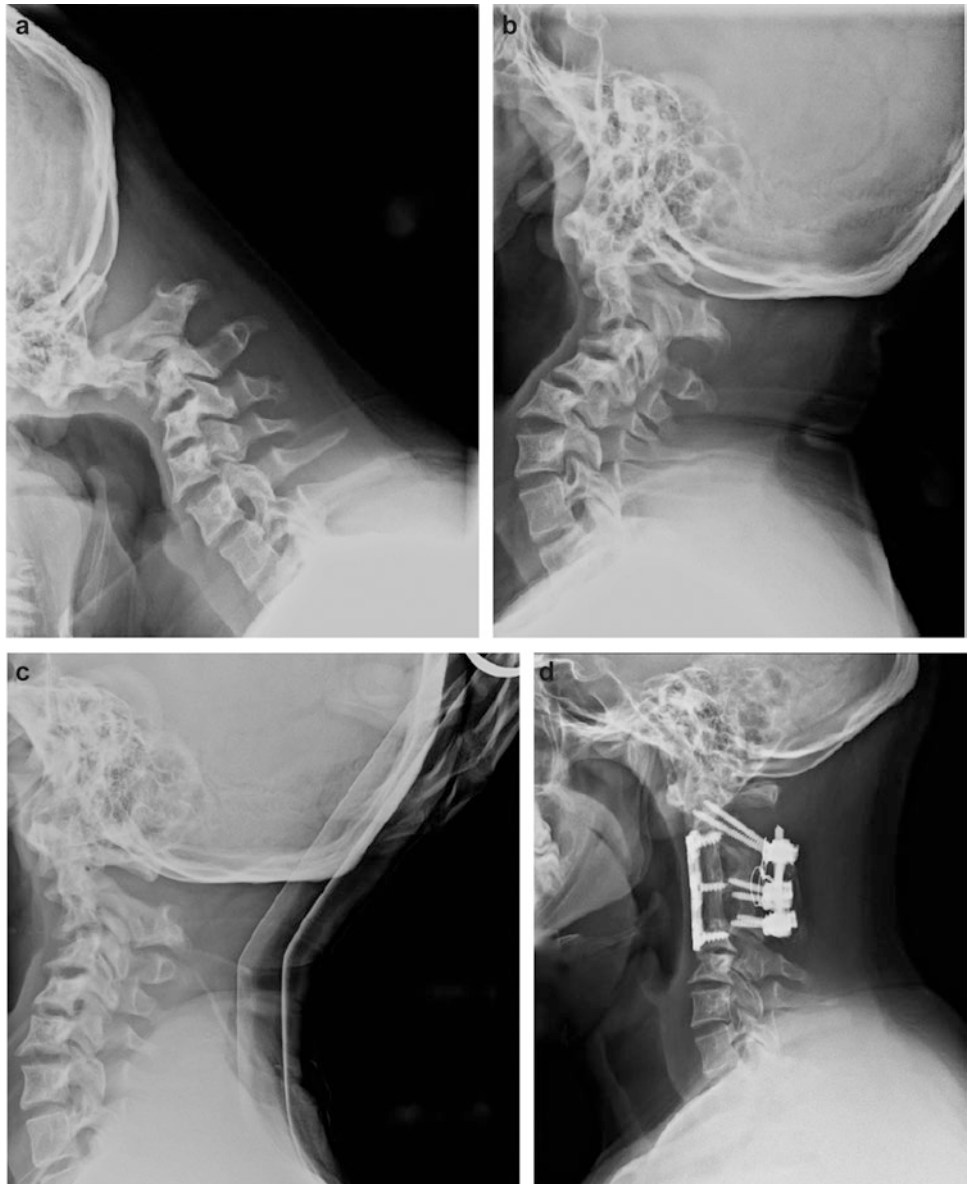

Fig. 18.3 (**a**–**d**) Radiographs of a 16-year-old male with neurofibromatosis-1 and dysplastic cervical kyphosis underwent successful anterior C3 corpectomy and arthrodesis with autologous structural rib graft and combined posterior instrumented arthrodesis with rib autograft. (**a**) Preoperative lateral flexion, (**b**) extension, (**c**) traction radiographs. (**d**) Postoperative 5-year lateral radiograph shows intact cervical fixation and alignment

(bending and torsional strength, as well as the compression stress) of allograft bone is compromised by the processing technique: fresh-frozen allograft has a 10–20% decrease in compression strength, and freeze-dried allograft has a 55% reduced bending strength and a 39% reduced torsional strength [16]. The use of autograft is also associated with an overall higher fusion rate in the adult population [23].

Decision-making with regard to the use of morselized or cancellous bone graft versus structural bone grafting depends on the structural support needed to ensure a rigid construct and fusion bed. Morselized bone grafts are

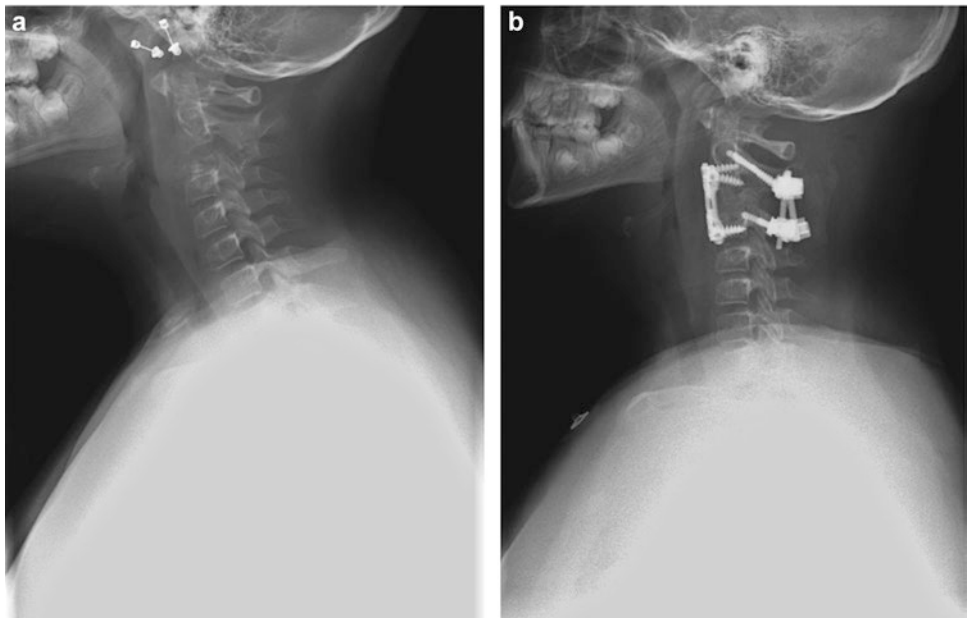

Fig. 18.4 (**a**, **b**) Radiographs of a 9-year-old female who successfully underwent a C3 corpectomy with an instrumented C2-C4 anterior arthrodesis, autologous structural iliac crest bone graft, and posterior spinal arthrodesis for her symptomatic eosinophilic granulomatosis involving the anterior, middle, and posterior elements of C3. (**a**) Preoperative lateral radiograph of the cervical spine shows vertebra plana at C3. (**b**) Postoperative 3-year lateral radiograph shows adequate anterior and posterior arthrodesis with instrumentation with appropriate sagittal alignment

porous and favor ingrowth but provide limited structural support compared with corticocancellous and cortical structural graft [24].When spanning multiple levels, especially in patients with compromised fixation, structural autograft may be preferred, as well as consideration of a posterior procedure with bone grafting, given the osteogenic nature of posterior cervical spine.

There is limited literature on the use of recombinant human bone morphogenetic protein-2 (rhBMP-2) in pediatric spine surgery. Jain et al. [25] reported a significantly increased (greater than 9%) use of rhBMP-2 from 2003 through 2009. Boakye et al. [26] reported a good clinical outcome and solid fusion in 24 adult patients undergoing anterior cervical discectomy and arthrodesis using polyetherketone filled with rhBMP-2. They also provided an anecdotal report of anterior neck soft tissue swelling in patients undergoing anterior cervical discectomy and arthrodesis using rhBMP-2-filled polyetherketone. Multiple studies have reported on the adverse effects attributed to use of rhBMP-2 in the pediatric spine [27–29]. Further studies are necessary to make recommendations on the use of rhBMP-2 in the pediatric cervical spine.

Instrumentation

The number of levels incorporated in the anterior cervical spine arthrodesis and the decision-making on single- versus multiple-level discectomy or corpectomy for the management of cervical spine abnormality depends on the extent of the disease. The small nature of the growing pediatric vertebra presents a technical challenge for instrumentation to the spine surgeon, especially in younger children. Anterior instrumentation enhances cervical stability and fusion and prevents graft extrusion, which is especially important in patients undergoing multiple-level decompression and

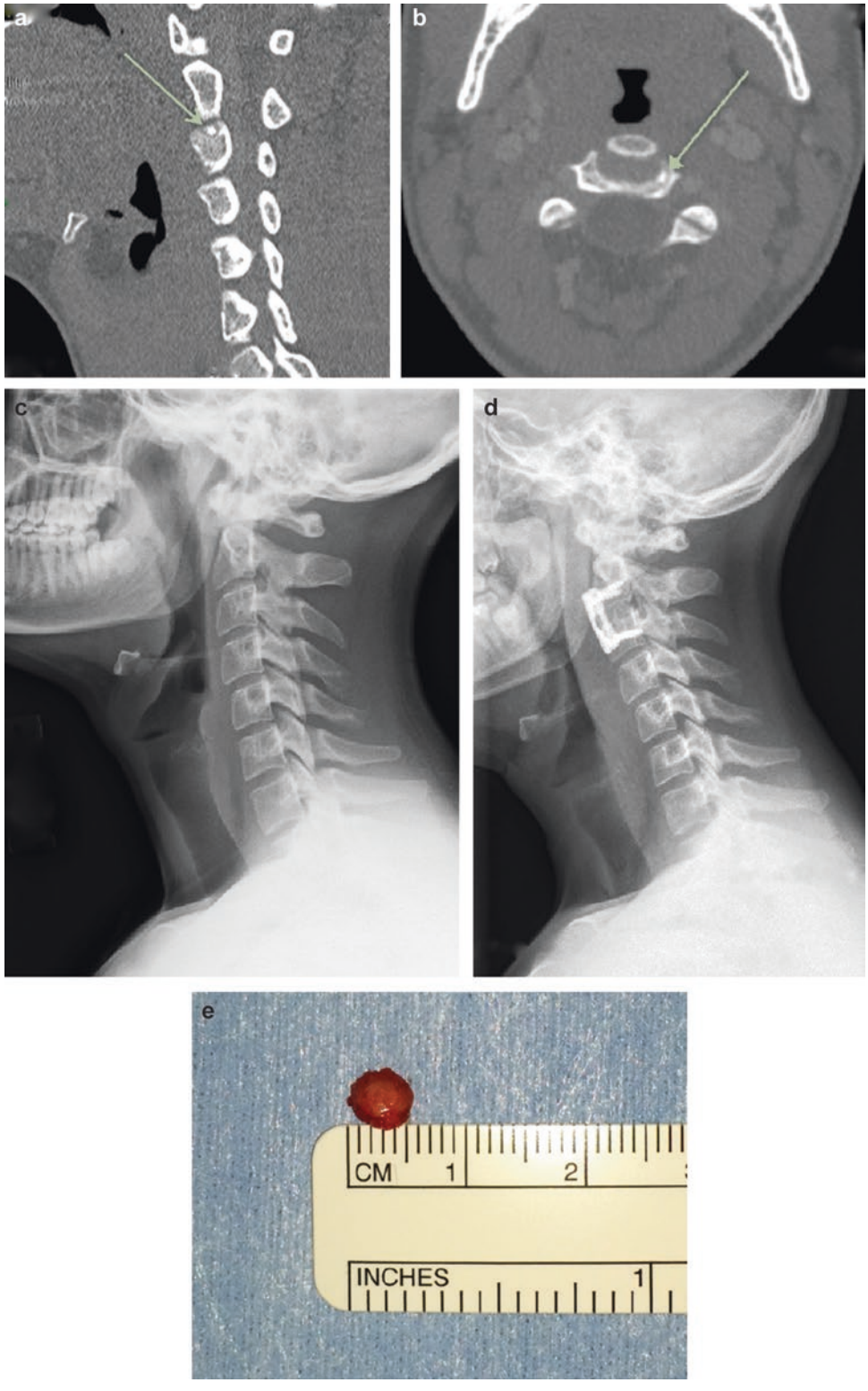

Fig. 18.5 (**a**–**e**) Imaging of an adolescent male patient with a diagnosis of symptomatic osteoid osteoma. (**a**) Sagittal and (**b**) axial preoperative computed tomography images of the cervical spine show an osteoid osteoma of the C3 vertebral body (see *arrows* on each). (**c**) Preoperative lateral radiograph. (**d**) Postoperative lateral radiograph after osteoid osteoma excision, anterior C2-C3 discectomy, and instrumented arthrodesis with autologous iliac crest bone graft. (**e**) Gross image of the excised osteoid osteoma

arthrodesis. Posterior cervical spine arthrodesis may be indicated, in addition to anterior arthrodesis, to enhance the stability of the cervical spine and minimize potential complications associated with multilevel anterior cervical spine arthrodesis.

There are no standardized implants for anterior arthrodesis of the pediatric cervical spine. Hedequist et al. [2] reported successful results (a 100% fusion rate) with children older than 12 years undergoing isolated anterior cervical arthrodesis (one patient) or combined anterior-posterior cervical arthrodesis (seven patients) using the standard adult anterior plate and anterior cages. The successful use of a craniofacial miniplate in a 2-year-old [3] and a 2 mm-thick miniplate in a 7-year-old [6] has been described for the treatment of cervical instability and for anterior cervical corpectomy and arthrodesis, respectively, to accommodate the small vertebral body in the young child. Multiple fixation options should be available in children younger than 10 years of age and should include an array of craniofacial and orthopedic miniplates with consideration of augmenting the fusion bed with posterior implants and/or halo vest application.

Summary

Anterior cervical surgical arthrodesis offers a valuable option for the treatment of anteriorly based cervical spine abnormalities in the pediatric population. Appropriate patient selection and preoperative planning are keys to the approach to overcome the limitations associated with the growing pediatric cervical spine and small vertebral bodies. The use of modern cervical spinal instrumentation in the adolescent patient and the availability of miniplates in the younger patients have proven to be successful treatment techniques, with good clinical outcomes and fusion rates. Consideration of halo vest for postoperative treatment should be done in younger patients, regardless of fixation, given the size and quality of their vertebra.

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19 Anterior Transoral Approaches to the Upper Cervical Spine in Children

Joshua Gottschall and James Kosko

Introduction

First described by Kanavel in 1919 for removal of a bullet from the cervical spine, the transoral approach to the cervical spine is now a favored approach for many extradural cervical spine pathologies [1]. Otolaryngologists frequently use the transoral approach for drainage of retropharyngeal abscesses and resection of oropharyngeal cancers. The transoral approach provides a direct route to the cervical spine with less morbidity than open approaches through the neck. In this chapter we describe the anatomy, indications for surgery, surgical approach, and complications of the transoral approach to the cervical spine.

Anatomy

The anatomy of the nasopharynx, oral cavity, and oropharynx is a complex and intricate passageway which contains many important structures. Surgical intervention in this region requires a complete understanding of the surrounding blood vessels, nerves, and musculoskeletal landmarks. The nasopharynx begins at the posterior choanae and extends inferiorly to the soft palate, which serves as the separation between the nasopharynx and the oropharynx. The oropharynx is bounded superiorly at the soft palate, posteriorly at the posterior oropharyngeal wall, anteriorly by the palatoglossus (anterior tonsillar pillar), and inferiorly by the base of the epiglottis.

The blood supply of the oropharynx is primarily from the ascending pharyngeal branch of the external carotid artery, with minor contributions from the superior thyroid artery, tonsillar branch of the facial artery, and the sphenopalatine artery. The glossopharyngeal and vagus nerves supply the primary innervation to the oropharynx.

There are multiple layers of the posterior oropharyngeal wall which serve to help protect the important structures of the head and neck. If bacteria are allowed to gain access and multiply in the deeper fascial layers, infection can propagate rapidly, leading to severe problems such as mediastinitis. The oropharynx is covered by a mucosal layer made up of nonkeratinized stratified squamous epithelium. Deep to the mucosal layer lies the pharyngobasilar fascia, which extends to the skull base. The next soft tissue structure is the superior pharyngeal constrictor muscle, which is located between the pharyngobasilar fascia superficially and the buccopharyngeal fascia deep. The superior pharyngeal muscle does not directly attach to the skull base, and because of

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this, there is a gap between these two structures called the sinus of Morgagni. Deep to the buccopharyngeal fascia is the alar fascia. The loose connective tissue layer between these two fascial layers lies the retropharyngeal space which is at risk for potential infection. The next fascial layer is the prevertebral fascia. It is between the alar fascia anteriorly and the prevertebral fascia posteriorly which lies the "danger space." The danger space is a potential space where infection can travel to the mediastinum. The prevertebral fascia covers the anterior longitudinal ligament, which travels anterior to the vertebral bodies [2].

The cervical spine is made up of seven bony vertebral segments (C1–C7), most unique being C1 and C2. The atlas (C1) helps form the joint between the skull base and the cervical spine. The bodies of C1 and C2 are fused, which allow C1 to rotate on C2, the axis. The axis (C2) has a prominent odontoid process (dens), which extends superiorly from the body and is the focus of most transoral cervical spinal surgical intervention. The cervical structures which comprise the posterior oropharynx are primarily C1–C2; sometimes, depending on the development, C3 can be included as well.

Lateral to the oropharynx are some important neurovascular structures which are typically not at risk during transoral approaches to the cervical spine, but are important to recognize, as complication from infection of inadvertent iatrogenic injury could potentially occur. Contained within the carotid sheath posterolateral to the oropharynx are the internal jugular vein, internal and external carotid artery, and vagus nerve (CN X) and superiorly also the glossopharyngeal (CN IX), the spinal accessory (CN XI), and the hypoglossal nerve (CN XII).

Preoperative Evaluation

A multidisciplinary approach (ENT, orthopedic surgery, neurosurgery) serves the patient well in the preoperative planning. Preoperative evaluation with imaging (CT, MRI, or both) is imperative to help identify the underlying cervical spine abnormality. Pathologies amenable to the transoral approach include os odontoideum, rheumatoid disease, atlantoaxial instability secondary to Down's syndrome, traumatic dens dislocation, pseudogout of C1/C2, ossification of the posterior longitudinal ligament, Arnold-Chiari I basilar invagination, and neoplasms (chordoma, giant-cell tumor, chondrosarcoma) [3]. During the physical exam, it is necessary to document any and every cranial nerve deficit present. If there are significant cranial nerve deficits, it is our opinion that a tracheostomy is more likely necessary prior to the procedure, due to the combined insult with surgery. During the postoperative care, the patient will not be able to eat by mouth and will have a nasogastric feeding tube for their primary nutrition. Wound healing requires nutritional optimization in order to aid in a timely closure. The posterior oropharyngeal incision is at risk for wound dehiscence and infection, with subsequent severe sequelae (explained later in the postoperative complications), which requires a proper nutritional evaluation and optimization prior to surgical intervention.

Surgical Procedure

Preoperative antibiotics (Ancef) are recommended 15–30 min prior to incision and continued for 24 h postoperatively. Intraoperative steroids are not routinely recommended. The airway is secured with an oral or nasal intubation, depending on the surgeon's preference. The patient is then secured in a supine position on the table, with the head slightly extended in a padded horseshoe headrest or Mayfield headrest. The oral cavity is not considered a sterile surgical site due to the multiple bacterial floras that exist, and as such, prepping the oral cavity is unnecessary. A self-retaining retractor (Crowe-Davis, Dingman, FK retractor) is then inserted into the oral cavity and then suspended from either a Mayo stand or other arm extension (Figs. 19.1 and 19.2). Catheters are then placed through each nostril and used to retract the soft palate anteriorly and clamped with a hemostat and a 4 × 4 gauze in between each catheter edge to prevent alar necrosis. The posterior oropharynx and

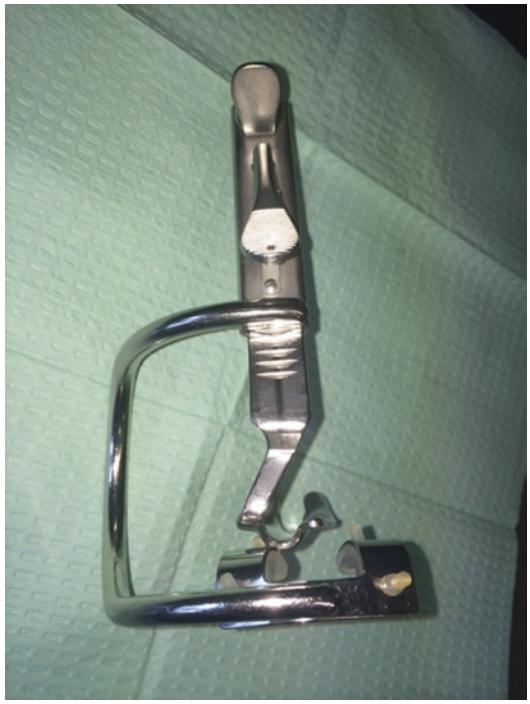

Fig. 19.1 Self-retaining retractor

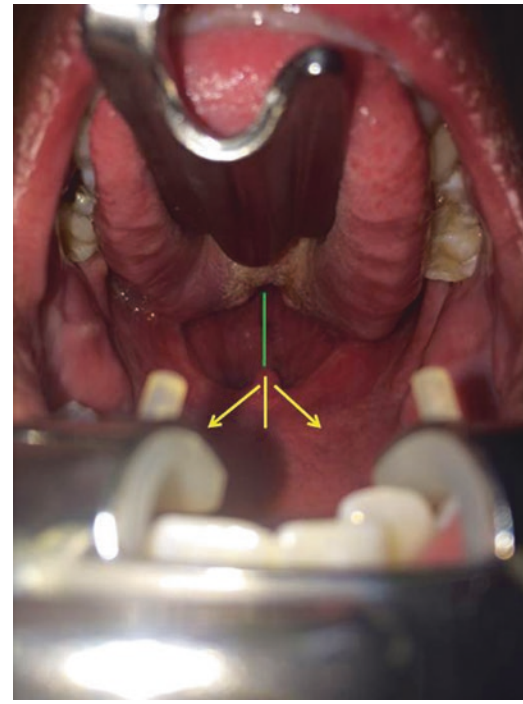

Fig. 19.2 Patient in suspension

cervical spine are then palpated to identify landmarks. If the superior exposure is not adequate, then the soft palate is split to provide further access. A 12-blade (sickle knife) is used to make an incision in the midline from the base of the hard palate to the uvula, staying just lateral to the uvula to avoid splitting it. A 3-0 silk suture is then placed through each of the soft palate flaps and used to retract them laterally (Fig. 19.3). The cervical spine is then again palpated to identify landmarks. The posterior oropharynx is then injected with 1% lidocaine with epinephrine 1:100,000 in the midline. Multiple different incisions have been described, including U-shaped and vertical (Fig. 19.4) [3, 4]. We recommend an I-shaped incision. The incision should be made approximately 1–2 cm superior and inferior to the cervical spine pathology. Following incision, the posterior oropharyngeal musculofascial layers are identified, and 3-0 silk suture is placed through the flaps to retract them laterally. Periosteal elevators are then used to dissect the anterior longitudinal ligament from the underlying vertebral bodies.

Once the cervical spine is exposed, the operating microscope is then brought into the field for further manipulation of the cervical spine. After the cervical spine work is complete, the oral cavity is copiously irrigated with saline or bacitracin-instilled solution. Closure of the pharyngeal flaps with 3-0 Vicryl suture is performed in a two-layer approach to reapproximate the muscular and mucosal layers and obtain a watertight closure. If the soft palate was split, the soft palate is reapproximated in a three-layer approach (nasopharyngeal mucosa, muscular layer, oral cavity mucosa) with 3-0 Vicryl suture. Nasogastric tube is then placed and secured in place for postoperative nutrition.

Recently the da Vinci robot has been described as another tool in the surgical armamentarium. Lee et al. [5] described a case report of using the robot during a transoral odontoidectomy for basilar invagination. Based on their result and the emergence of robotic usage, this is another tool to be considered in the transoral approach to the cervical spine, due to the increased precision, field of view, and range of motion that the robotic approach affords the surgeon.

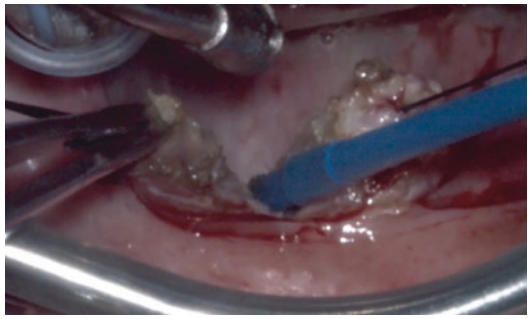

Fig. 19.3 Soft plate retracted laterally

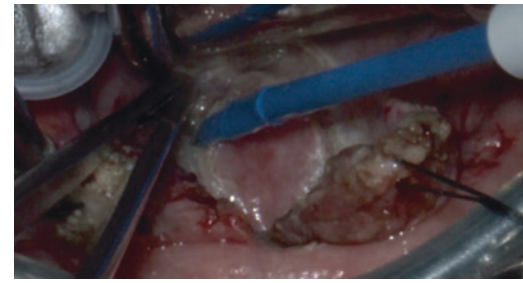

Fig. 19.4 U-shaped flap being elevated

Airway Management

The decision for tracheostomy versus prolonged postoperative intubation is an important part of the surgical planning process that needs to be made prior to surgical intervention. With many of the pathologies of the cervical spine, cranial nerve involvement is common. If there are significant glossopharyngeal, vagal, or hypoglossal nerve deficits, there is a need for tracheostomy, as these individuals will not be able to adequately protect their airway postoperatively. Due to the physical constraints from getting to the airway and neck in those patients with external hardware, bag-mask ventilation, direct laryngoscopy, and emergent tracheostomy are impaired, and thus a tracheostomy is needed. Other factors which will prompt a tracheostomy include poor oral opening, high Mallampati grade, increased neck circumference, and micrognathic patients. Postoperative intubations have been described ranging from extubation in the operating room to 7 days postoperatively, with most postoperative intubations ranging from 24 to 48 h postoperatively [3, 4]. We recommend tracheostomy at the time of surgery for any patient requiring external fixation hardware postoperatively (i.e., halo). In patients without the need for postoperative external hardware, we advocate testing extubation parameters, especially a strong cuff leak (depending on the size of endotracheal tube in place) with the first 24 h postoperatively. Once the patient is ready to be extubated, we recommend removing the endotracheal tube, as this will rub and irritate the posterior pharyngeal wall incision, putting it at higher risk for dehiscence.

Postoperative Nutrition

Postoperatively the patient will feed exclusively via a nasogastric tube with enteral feeds. We recommend nothing to eat by mouth for 7 days postoperatively to allow the posterior pharyngeal wall incision to heal. On postoperative day 7, we recommend bedside swallow evaluation to assess for aspiration in those patients who are eligible for trial of oral diet. If there is no evidence of aspiration, we recommend slow advancement of diet from clear liquid to soft diet, with a soft diet recommended for 1 week.

Complications

Velopharyngeal insufficiency (VPI) is the most reported complication of this approach, with up to 50% of patients having VPI with the transpalatal approach reported [3, 6]. Most cases of VPI resolved within 1 month postoperatively [3]. Dysphagia is another common complaint of patients, which is usually transient. Dehiscence of the pharyngeal wall incision is a rare complication, usually the result of inappropriate manipulation of the posterior pharyngeal wall (misuse of instrumentation in the mouth), infection, or malnutrition. If infection is suspected, intravenous antibiotics are initiated and, if necessary, surgical drainage of abscess is performed. Most infections are derived from oral flora such as bacteroides, streptococci, staphylococci, and corynebacterium. Aspiration pneumonia has been reported and is more common in Down's syndrome patients who are already at risk for

aspiration [2]. Cerebrospinal fluid (CSF) leak is a potential complication if the dura is violated during surgery. As most lesions are extradural, this is a rare complication but one to be particularly aware of. Emergent tracheostomy due to inability to secure an airway postoperatively has also been observed and is the reason preoperative identification of patients who potentially might need a tracheostomy needs to be identified before surgical intervention.

Summary

Disorders of the craniocervical junctions due to compression, inflammatory conditions, and tumors present management dilemmas when the problem is anterior to the spinal cord. The exposure of this area is technically demanding and may be done through a transoral approach. The exposure is fraught with complications; however, a detailed understanding of the anatomy and potential pitfalls with the exposure can lead to excellent visualization and surgical treatment in this challenging area.

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Chiari Malformations and Foramen Magnum Stenosis

20

Christopher A. Gegg and Greg Olavarria

Introduction

In the early 1890s, Professor Hans Chiari developed a four-type classification for the malformation entity while working as an anatomic pathologist at the German University in Prague. His description was based on more than 40 autopsies, noting varying degrees of hindbrain herniation and malformation of hindbrain structures [1, 2]. Since the advent of modern imaging, we have a better understanding of these tiers, which has allowed more distinct descriptions based on Chiari's original classifications, along with associated findings as syringomyelia, tethering of the lumbar spinal cord, and scoliosis with other potential boney spinal disorders. For the scope of this chapter, types 1 and 2 will be discussed, while types 3 and 4 will be mentioned.

Epidemiology

Type 1 Chiari malformation consists of caudal displacement of the cerebellar tonsils into the upper cervical canal; 6 mm caudal displacement of the cerebellar tonsils below the foramen magnum edge has been chosen by many, including the authors, as the cutoff extent of displacement that distinguishes Chiari type 1 malformation from cerebellar tonsillar ectopia, less than 6 mm. Since in many cases there are no symptoms, the true incidence is unknown, but since MRI technologies have advanced, the diagnosis is certainly more common, and a familial pattern has been observed in about 3–12% of cases depending upon the study (Fig. 20.1) [3, 4, 5].

In Chiari type 2 malformations, the foramen magnum herniated structures include the cerebellar tonsils, cerebellar vermis, brainstem, and involvement of the fourth ventricle. The dural venous sinus, typically the torcula (sinus confluens), is also caudally displaced and is an important consideration at time of surgical decompression when indicated. Type 2 malformations have much more severe anomalies with great health impact. Uniformly these include myelomeningocele "spina bifida" and hydrocephalus, as well as boney anomalies of the spine, ribs, and hips. Frequently these lead to surgical corrections being needed.

Chiari 3 and 4 lesions are quite unusual. Chiari 3 represents a suboccipital encephalocele with contents of the cerebellum and brainstem; this is the most severe form of Chiari, and ethical decisions are necessary, since this form can be lethal or associated with severe neurologic dysfunction. It is important to distinguish between Chiari 3 and high cervical

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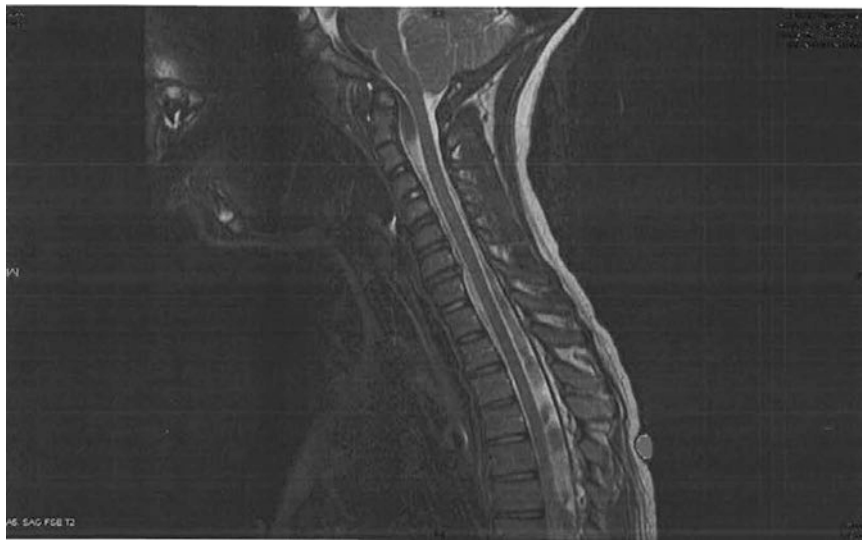

Fig. 20.1 Chiari I malformation, crowding of foramen magnum "retroflexed" odontoid

myelomeningocele, which has a prognosis similar to the lumbar variant, rather than the severity of Chiari 3. Chiari 4 malformations are simply manifested by hypoplasia or aplasia of the cerebellum without hindbrain herniations (Table 20.1).

Signs and Symptoms of Chiari 1

The diagnosis of Chiari 1 is made earlier now with MRI being so available. Most patients who present to our clinic already have an MRI in hand with the suspected problem already diagnosed. It is estimated that in almost half of patients, the diagnosis is made within their first 10 years, while the rest are between 10 and 15 years of age [6].

The most common presentation is discomfort; in our experience, frontal headaches are the most common symptom and may be exacerbated by forceful maneuvers—the described Valsalva headache during a sneeze, cough, or loud scream. This may be accompanied by neck discomfort, but that is less common. In younger children, who cannot express themselves, unexplained irritability or crying may be the only evidence of discomfort.

Other symptoms or signs may include upper back or shoulder discomfort, arm discomfort or sensory changes, and ataxia/dysmetria, especially if syringomyelia is present. Cranial nerve irregularities may occur; nystagmus is, in our experience, the most common of these, while dysphagia or vocal cord paralysis with Chiari 1 is rarely encountered. In children or adolescents, respiratory irregularities may be discovered during monitored sleep studies. Tubbs et al. found approximately 5% of pediatric patients with associated sleep apnea [3].

Progressive scoliosis is becoming a frequent finding in conjunction with Chiari 1 malformation. Scoliosis is reported in 25–50% of Chiari 1 cases [1, 7–9]; this is more common in the presence of syringomyelia, which can be present in 50–76% of Chiari 1 patients and in up to 82% of patients with associated scoliosis. Scoliosis may also occur in absence of syringomyelia [10]. Improvement or stabilization of curve is recognized after Chiari decompression [10].

The causality of scoliosis coexistent with Chiari 1 is not clear but may involve vector forces aggravated by Valsalva maneuvers and downward displacement of the hindbrain; there is good evidence that early decompression may avoid a larger scoliosis operation [11–18].

Table 20.1 Findings in Different Chiari Malformations

Chiari 1	Tonsillar herniation >5 mm	No brainstem herniation	Low frequency of hydrocephalus	No supratentorial irregularities	Syringomyelia may occur
Chiari 2	Caudal herniation of cerebellum/vermis and tonsils, 4th ventricle	Frequent hydrocephalus and supratentorial irregularities	Frequent syringomyelia		
Chiari 3	Occipital/suboccipital encephalocele containing most of the hindbrain	Associated with severe neurologic dysfunction or lethality	Need to distinguish from high cervical myelomeningocele		
Chiari 4	Hypoplasia or aplasia of the cerebellum				

Lower back discomfort may also be present; it is important to screen for coexistent tethering of the lumbar spinal cord, especially if scoliosis is present, which, in this case, would have the radiographic appearance of a thickened "fatty" filum terminale or low conus medullaris with tight filum terminale. Although this is not usual, we have seen a relationship between the two (Fig. 20.2).

Among the theories to be addressed, with or without syrinx, the most common pathogenesis is the pressure gradient differential across the foramen magnum that translates into altered CSF circulation [11–14, 17–21]. This presumably results in intermittent vector forces that transmit caudally; this may affect axial alignment and result in scoliosis as well as the other described signs and symptoms. Evidence suggests that a scoliosis curve convex to the left is more likely associated with Chiari 1 [6, 20, 21].

We have also seen a strong association of Chiari 1 in patients who have pseudotumor cerebri, a condition of elevated intracranial CSF pressures without ventricular enlargement. This commonly presents with chronic headaches and may be associated with papilledema. Other associated diagnoses can include hydrocephalus in about 9.6% of patients and neurofibromatosis type 1 in about 5%, while craniosynostosis and Klippel-Feil anomalies are also occasionally encountered. Basilar invagination was encountered in 3% of pediatric patients in a Tubbs et al. study; this is an indication of instability, and surgical intervention should be appropriately modified, as listed here:

Signs and symptoms of Chiari 1 malformations

- Headaches (occipital or frontal, may be exertional)
- Nystagmus
- Arm or shoulder pain with clumsiness
- Sleep apnea
- Ataxia
- Dysphagia
- Hoarseness scoliosis
- Recurrent aspiration
- Lower cranial nerve irregularities

Imaging Findings of Chiari 1

Pathologic position of cerebellar, tonsillar caudal displacement is generally accepted as 5 mm below the foramen magnum. Barkovich et al. in a study of 200 healthy adults and children found normal cerebellar tonsils lie 1.0 mm above the foramen magnum and virtually all symptomatic patients had tonsillar herniation beyond 3–5 mm range [3, 8]. It is also not uncommon to find one cerebellar tonsil longer than the other.

Some have observed that normal cerebellar tonsils may be displaced up to 6 mm during the first decade of life, with gradual ascent with growth. This has been the occasional experience in our practice as well. This is one factor to take into account in the asymptomatic patient.

One radiographic finding that we have found important is a crowded foramen magnum with

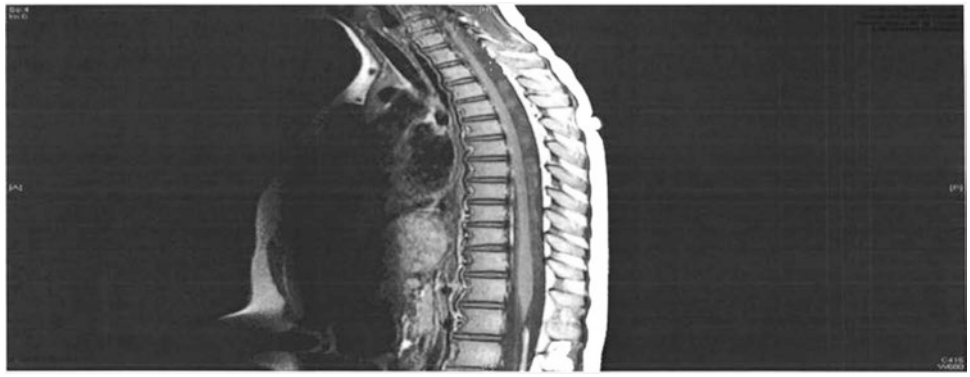

Fig. 20.2 Fatty filum terminale in patient with Chiari I malformation, tethering of lumbar spinal cord

CSF displacement. This is best seen on MRI axial images at the level of the foramen magnum (Fig. 20.3). This has helped with surgical decision-making, especially when tonsils may be borderline in caudal displacement.

Some have advocated obtaining flexion and extension radiographs preoperatively, but a large series by Oakes et al. did not discover any instability in Chiari 1 decompression patients. The odontoid process is frequently discovered to be elongated and "retroflexed" on MRI, but if basilar invagination is absent, surgical decompression has not been associated with causality of instability, as seen in Fig. 20.1 [3, 9].

Posterior fossa volume may also be small in a minority of patients. Schady et al. discovered this in 23% of patients. Our experience has shown that this can be unapparent on Chiari-type 1MRI imaging, but discovered at time of surgical decompression. A study by Badie et al. compared 20 Chiari 1 patients to 20 control patients and found that the posterior fossa to supratentorial space ratio was smaller in the Chiari 1 group; this correlated with clinical symptoms (Fig. 20.4) [6, 22]. Syringomyelia can be seen in 50–76% of patients; thus it is important to include cervical and thoracic MRI imaging when working up Chiari 1 patients. This also assists in discovering the occasional associated tether of the lumbar spinal cord that may aggravate scoliosis and require a separate procedure.

The cause of syringomyelia is not fully understood, but one classic and interesting theory was by Gardner in 1965. This was the hydrodynamic theory, which postulated that the persistent pulsing communication between the fourth ventricle and central canal of the spinal cord (obex) could lead to syrinx development, a "water hammer" effect. Oldfield and coworkers similarly developed a theory based on cine MRI. In their theory, they postulated there is a pistonlike motion of the cerebellar tonsils that produces a CSF systolic pulse wave that acts on the spinal cord and induces CSF leakage through the interstitial and perivascular spaces [23, 24]. Frequently the syrinx can be quite large with thinning of the spinal cord, but despite this the spinal cord is usually strikingly unaffected throughout much of the initial diagnosis. This finding though dictates the need for decompression.

Surgical Planning in Chiari 1

There is no recognized alternative to surgical decompression for symptomatic Chiari 1 malformations. Caudal displacement of the cerebellar tonsils from 5 mm to 6 mm below the foramen magnum qualifies as a Chiari 1 malformation, but this as an isolated finding without syringomyelia or other symptoms as headaches (see the box on Signs and Symptoms, above) and is followed conservatively. We recommend yearly follow-up with imaging until it is evident symptoms are not likely to occur. When one adds signs or symptoms to the above finding, such as syrinx, head-

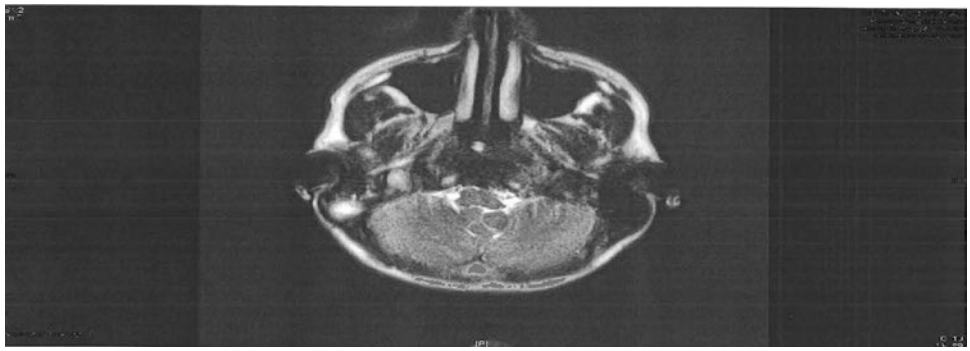

Fig. 20.3 Note asymmetry of the brainstem secondary to crowding of cerebellar tonsils, right greater than left

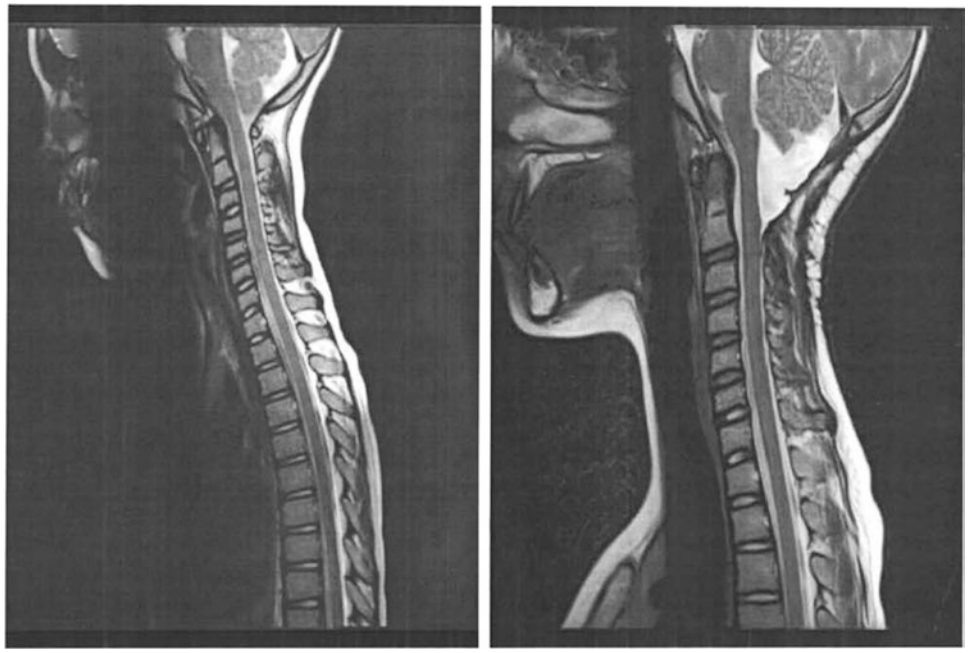

Fig. 20.4 Before and after Chiari 1decompression (sutureless duraplasty technique) demonstrating improved posterior fossa volume; note obex visualized after decompression

aches, nystagmus, or neck discomfort, surgical decompression becomes the safer alternative.

In the unusual case of a patient having hydrocephalus and Chiari 1 with a syrinx, the hydrocephalus should be treated first and observed for improvement over 6–8 months. If signs or symptoms persist, then it is prudent to proceed with decompression.

As mentioned earlier, it is important to evaluate the MRI for ventral compression that might destabilize the upper cervical spine with suboccipital decompression. This finding is unusual in most patients, and severe ventral compression would be mostly categorized as basilar impression, where the odontoid tip is above the clival line; this would be more common in patients with associated segmental anomalies as Klippel-Feil syndrome or ligamentous laxity of Down syndrome. These patients would require a concomitant fixation/fusion procedure. Patients with mild to moderate ventral compression from an elongated "retroflexed odontoid" without basilar impression are treated best with standard

suboccipital decompression and C1 laminectomy alone; this has been our experience also [3, 9].

Once the decision to decompress is made, while under general endotracheal anesthesia, the patient is positioned prone with enough flexion to assess the suboccipital space—we use a padded horseshoe head rest. The incision is midline from mid C2 to the occipital protuberance, followed by a subperiosteal exposure of dorsal C1 ring, and the subocciput from occipital protuberance to the C1 exposure across the foramen magnum. The foramen magnum is exposed by periosteal dissection out to the lateral edge where it makes it turn on both sides; craniectomy is performed with a combination of drilling and rongeurs. In the younger patient, we attempt 3 cm of side-to-side craniectomy of the foramen magnum; in older patients, 3.5–4 cm can be achieved safely. A penfield #2 is used to dissect periosteum and palpate the lateral edges.

Once the bone work is complete and bone dust is washed away, treatment of the dura is the next decision. In our practice, we have developed a dural splitting technique that thus far over 5 years has yielded good results. In our dural treatment, we have considered it a "sutureless duraplasty" that involves splitting and peeling the dura back one layer at a time until the thin transparent dura is left. In this manner, working under the microscope, we can see dural and tonsillar pulsations without the need of ultrasound. It is important to split the dura past the thick dural band at the middle of the lower cerebellar hemispheric protrusions and down to top of C2. Development of pseudomeningocele with this procedure has not occurred, and improvement in symptoms or signs is greater than 97%. Litvack et al. describes a similar approach to the dura with over 90% improvement rate [25].

Decompression for foramen magnum stenosis is performed in a similar manner. Most of the work, in such cases as achondroplasia, is expanding the foramen magnum by craniectomy to release tension on cervical medullary junction (Fig. 20.5). This can be done safely without causing instability. This also can be lifesaving in such cases to prevent central sleep apnea. Sleep studies are beneficial in decision-making and to look for postoperative improvement [26–28].

Stabilization or improvement with syringomyelia or scoliosis has been achieved in most patients; return to surgery for open patching has been about 1% (Fig. 20.6a, b). Brockmeyer et al. reported a decrease in syrinx size did not necessarily correlate with scoliosis improvement [7, 29]. In general, rates of scoliosis improvement without dural opening are still under investigation, and a larger series is needed, but our early results have been satisfying [7].

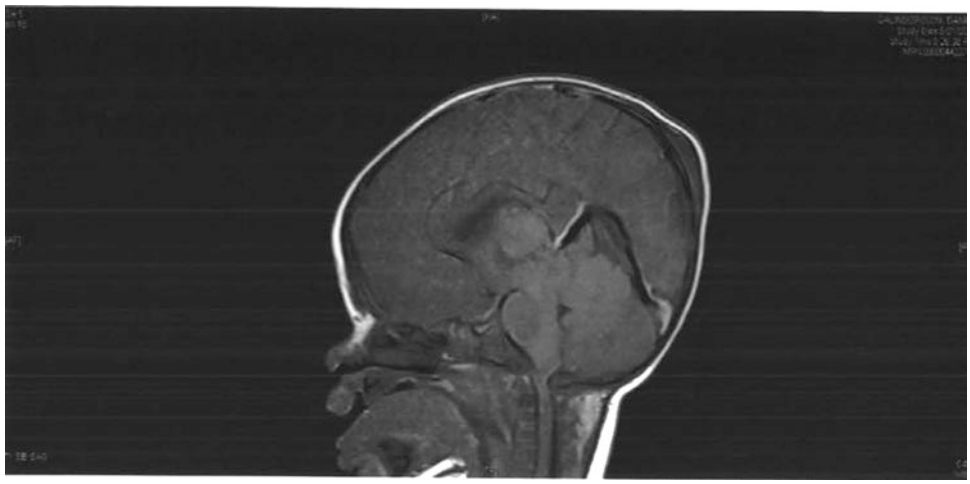

Fig. 20.5 Achondroplasia with severe stenosis of foramen magnum with compression of cervical medullary junction

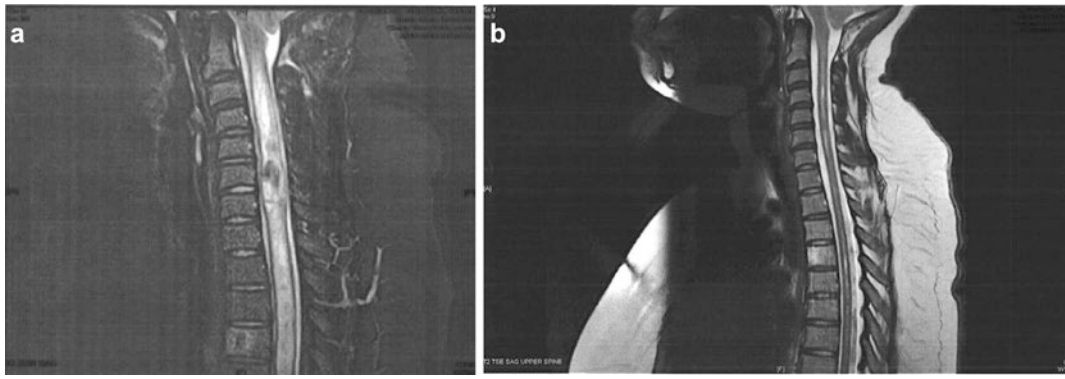

Fig. 20.6 (**a**) Severe syringomyelia secondary to Chiari 1 in patient with pseudotumor cerebri; (**b**) shows significant improvement of syrinx after repeat decompression with more thorough dural splitting "sutureless"; patient did not require open patch graft

In more severe cases of tonsillar displacement and syringomyelia, the decision to open patch and inspect for arachnoid adhesions may be necessary. In Litvack et al., the rate of chemical meningitis was 6%, and symptomatic pseudomeningocele was 1.5% in the cohort of open dural patch graft patients with more severe presentation. There were none in the splitting group. In Hankinson et al., the open dural patch group had a rate of 17% of radiologic evident pseudomeningocele and 7% of other CSF-related complications [30].

The graft material is typically pericranium or bovine pericardium and is sutured to the open dural edges to create a sack appearance; the dural venous sinuses are cauterized with the bipolar. When dura is open, one can inspect for dural adhesions and release these working under the microscope.

Follow-Up

The typical patient's hospital course is 3 days with office follow-up at 2 weeks and post-op MRI of the brain and cervical/thoracic spine at a 3-month follow-up. Syringomyelia may not show improvement at this early stage and may take up to a year to show improvement, but stability can be demonstrated, as well as clinical improvement. Infection without pseudomeningocele is very rare and, when present, is usually a stitch abscess or superficial infection treated with oral antibiotics. Cerebellar tonsils may not ascend, but we have seen this. More importantly, the appearance of improved CSF spaces at the posterior fossa and cervical medullary junction and the obex within the fourth ventricle can be seen, indicating a more open and relaxed fourth ventricle, as seen in Fig. 20.4. Boney regrowth is not common and is more likely in the younger child under 5 years of age. In all, the proper approach to treating Chiari 1 patients should be individualized to each patient, taking into account the expectations of the patient and/or family.

The Chiari Type II Malformation: Presentation, Diagnosis, and Surgical Management

Presentation and Pathophysiology

The Chiari Type II malformation differentiates itself from the Chiari Type I malformation by its association with myelomeningocele. Tonsillar herniation, as in Chiari Type I, is the hallmark of the disorder, but this is just one manifestation of a complex series of brain and spinal cord abnormalities. A unified theory has been proposed for the constellation of findings. The theory involves the open lumbar defect with escape of cerebrospinal fluid and therefore lack of ventricular distension at a critical stage of embryological development, causing herniation downward of the posterior fossa contents. This ventricular dis-

tension is critical to development of not just the brainstem and cerebellum but the supratentorial brain as well [31]. The lack of ventricular distension also prohibits the normal development of the cranial mesenchymal tissue, leading to a smaller than normal posterior fossa and crowded contents therein. Hydrocephalus develops from lack of fourth ventricular outflow. The type II malformation and, in my opinion, the hydrocephalus caused by the Chiari II have the most potential to cause the affected child serious neurological morbidity (Figs. 20.7 and 20.8).

Symptoms on presentation differ with the affected child's age. Hydrocephalus presents in infancy with separated cranial sutures, rapidly enlarging head circumference, and a full fontanel. It usually progresses after closure of the lumbar defect but can be present and symptomatic in the first hours of life. Bradycardia and respiratory instability can ensue when severe. Shunting the ventricles is the mainstay of treatment (see below). Older children can present with shunt malfunction and headaches, nausea, vomiting, and mental status changes. Though rare for an infant to display symptoms of the Chiari malformation itself, it may happen and can present with apnea, stridor, swallowing problems, and the risk for aspiration pneumonitis. Upper extremity hypotonia and weakness usually accompany the above findings. These are thought to be causally related to either the lower cranial nerve/brainstem downward herniation and compression or an intrinsically abnormal brainstem physiology. These competing theories have an impact on surgical decision-making. Attention and treatment of the hydrocephalus often ameliorate the above symptoms, obviating surgical intervention for the Chiari II. Older children present with headache, ataxia, upper and lower extremity weakness, and visual disturbance, a very rare phenomenon because most patients present symptomatic in infancy [32]. Shunt malfunction with increased intracranial pressure is often the cause of late deterioration in this population. Older children with myelomeningocele can also present with tethering of the spinal cord years after initial closure. Symptoms include back and leg pain, lower extremity weakness, and bladder function changes.

Diagnostic Findings

At birth, an ultrasound of the head is often the first test ordered to assess ventricular size, after a complete neurological and physical examination of the infant. MR imaging of the brain and

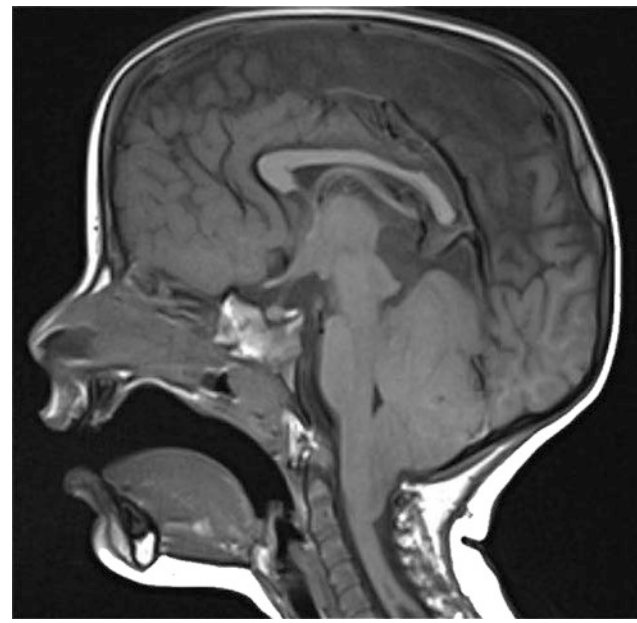

Fig. 20.7 Representative case of Chiari II malformation

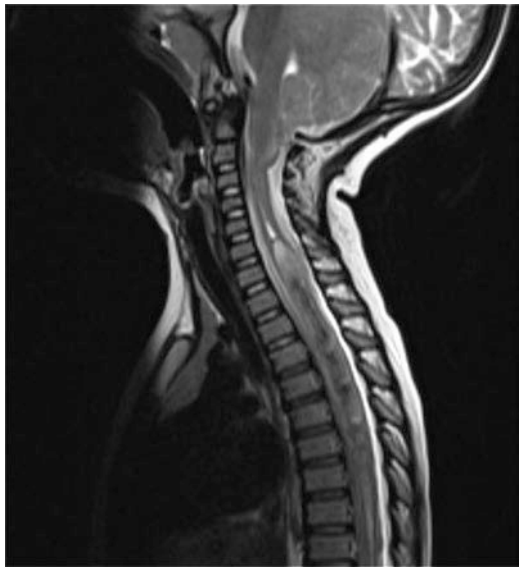

Fig. 20.8 Representative case of Chiari II malformation

spinal cord can follow, especially if symptoms of the Chiari II arise early in life. Apart from tonsillar herniation, patients with Chiari II exhibit herniation and kinking of the brainstem downward into the foramen magnum. The torcula and sinuses are lower than normal and can be near the foramen magnum, which has surgical implications. Other findings include hydrocephalus, beaking of the midbrain tectum, an enlarged third ventricle massa intermedia, and an enlarged foramen magnum. Hydro- or syringomyelia can occur, especially in the presence of untreated hydrocephalus, a shunt malfunction, or a tethered cord. Disorders of the skull include Luckenschadel or copper beaten thinning, a small posterior fossa, a low-lying tentorium cerebelli, and shortening of the clivus. Cerebral abnormalities include polymicrogyria, heterotopias, and dysgenesis of the corpus callosum, with the attendant lifelong risk of seizures. Hydro- or syringomyelia of the cord can result from shunt malfunction but can also be attributed to the Chiari II malformation or tethered spinal cord and requires artful diagnostic skill and attention to differentiate the various causes [33].

Surgical Management and Outcomes

Surgical management addresses the findings described in the above sections. Initial closure of the myelomeningocele defect is usually the first neurosurgical procedure, and sometimes infants are shunted at the same time. If not initially shunted, close surveillance, preferably by ultrasound or MRI, is especially important, and a multidisciplinary approach with specialists in neurosurgery, orthopedics, and urology leads to optimal outcomes.

In the shunted child, ruling out a shunt malfunction when new neurological symptoms appear is critical and, if overlooked, can lead to unnecessary surgical interventions. Once shunt malfunction is ruled out, decision-making turns to the Chiari/posterior fossa itself, whether a syrinx is present, and possible spinal cord tethering. The surgical approach to the Chiari involves cervical laminectomy, usually without suboccipital craniectomy, as the foramen magnum is enlarged in these patients. The standard approach involves upper cervical laminectomy with dural opening, lyses of adhesions to open the fourth ventricle, and duraplasty [32]. Some have argued for bone-only decompression without dural opening, lyses of adhesions to open the fourth ventricle, and duraplasty [32]. Some have argued for bone-only decompression without dural opening and section of epidural bands as a valid option [34, 35]. Complications such as hemorrhage from the venous sinuses, intradural scar formation, and cerebrospinal fluid leak can be avoided with good outcomes (the majority of patient's symptoms improved in these series).

Syringomyelia and scoliosis management deserve special attention and often present together. La Marca and McLone elegantly described presenting symptoms, location, and type of syringes in their series. Management (after ruling out shunt malfunction) is directed at the Chiari for stridor, swallowing difficulties, and neck pain; tethered cord for bladder function changes, back pain, or lower extremity weakness; and a creative approach for mixed symptoms based on imaging findings and type (holocord versus segmental) of syrinx. Scoliosis surgery by orthopedic surgeons,

especially in the presence of a syrinx, requires neurosurgical attention to the shunt, Chiari, and tethered cord, depending on patient symptoms. Progressive scoliosis in the absence of a syrinx may not require neurosurgical intervention unless the patient is symptomatic [36]. Neuromonitoring is highly recommended during scoliosis surgery. Postoperative laminectomy can sometimes lead to cervical kyphosis if a wide laminectomy and facet disruption is performed.

Fetal surgery for myelomeningocele repair has shown promise in reducing both the need for shunting hydrocephalus and Chiari Type II imaging findings [37]. In theory, early closure in utero can prevent the brainstem and tonsillar descent that initiates a harmful cascade of pathophysiology in these children. Long-term results are forthcoming and eagerly awaited.

Summary

Compression of the upper cervical spine can lead to brainstem dysfunction, myelopathy, and, ultimately, death if left untreated. Chiari malformations and foramen magnum stenosis may be caused by a wide array of conditions and may lead to significant patient morbidity if untreated. A clear understanding of the pathoanatomy, natural history, and treatment options will allow the surgeon to successfully manage these disorders.

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Miscellaneous Conditions of the Head and Neck in Infants and Children

21

David Miller

Introduction

Head and neck abnormalities occur throughout the entire pediatric age spectrum, from the newborn to the child and adolescent. Primary causes include embryologic vestigial remnants (i.e., thyroglossal duct cyst, branchial remnant anomaly, thymic cyst); infections (i.e., lymphadenitis, lymph node abscess, cat scratch disease, Epstein-Barr virus lymphadenopathy); neoplasms, both primary and metastatic (i.e., lymphoma, rhabdomyosarcoma, neuroblastoma); and metabolic disorders (i.e., hyperthyroidism with goiter). The diagnosis at times is straightforward, made simply by history and physical examination. Many of these lesions can be accurately diagnosed by their characteristic location, appearance, and age at presentation. However, the most sophisticated radiographic studies, scans, and laboratory investigations may be necessary, and involvement of a surgeon may be required to arrive at the correct diagnosis.

In general, the majority of these lesions are histologically "benign," but they can certainly behave in a "malignant" or life-threatening manner, with impingement on adjacent vital structures. This is well illustrated by a newborn infant with a large, congenital teratoma causing upper airway obstruction (Fig. 21.1).

The neck occupies a relatively small area of the child's total body, but within this small, defined space, the anatomy is complex, and the unprepared surgeon can rather quickly be led astray. An understanding of the anatomy, embryology, and development of the cervical area is critical. Excision of a thyroglossal duct cyst without removal of the central portion of the hyoid bone is almost certain to result in recurrence.

The following features and criteria are more likely to be associated with a benign process: (1) midline location; (2) cystic in nature; (3) inflammation, i.e., tenderness, erythema, pain, and swelling; (4) chronic long-standing history of the lesion; and (5) history of cat scratch.

The time-honored approach to workup and diagnosis begins with the patient's history. Prior fever and a short or long history of the lesion's presence point to a benign process. On exam, the lesion may be fluctuant or cystic. If unsure, an ultrasound study can be helpful to distinguish cystic from solid tumors; the latter is more likely to be malignant. While most head and neck lesions are benign, malignant tumors are seen in infancy and childhood. The following features suggest a malignancy: (1) low cervical or supraclavicular nodes; (2) persistent fever >38 C (101 F) for over a week; (3) weight loss; (4) mediastinal lymphadenopathy on chest X-ray (CXR) or computerized axial tomography (CAT) scan; (5) local characteristics, i.e.,

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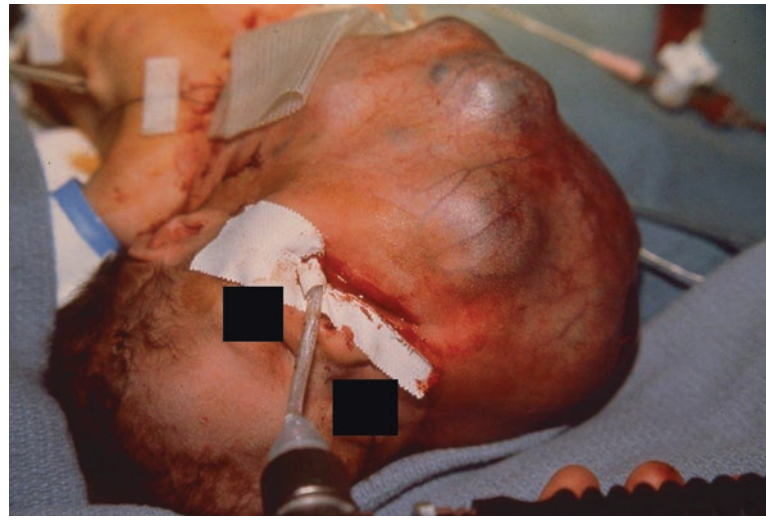

Fig. 21.1 Large cervical teratoma with upper airway occlusion at birth

mass non-tender, fixed to surrounding structures, and firm to hard on palpation; (6) no decrease in size after 4–6 weeks of observation ± treatment with antibiotics; (7) hepatosplenomegaly; and (8) lymph node >3 cm in diameter.

Radiographic Studies

Color Doppler ultrasound can demonstrate blood flow of the commonly seen hemangiomas and the occasional arteriovenous fistula. Multicystic lesions without significant vascularity are more likely to indicate a lymphangioma. Although CAT scan is an excellent tool in the workup, it should be used judiciously due to the latent effects of ionizing radiation, with the development of secondary malignancies later in life. The benefits should be weighed against the risk. MRI (magnetic resonance imaging) scans are becoming increasingly popular in the evaluation of a child with a complicated head and neck lesion. It is excellent in evaluating soft tissue lesions, with the downside of requiring deep sedation or even general anesthesia for the study, due to the problem with motion artifact. Radionucleotide thyroid scan can assist in the evaluation of thyroid masses, especially when used in conjunction with a Doppler ultrasound. One must not forget the basic CXR, which can be the first clue of lymphoma, presenting with mediastinal lymphadenopathy or a metastatic lesion of thyroid carcinoma.

Benign and Malignant Neck Lesions

Benign Midline Neck Lesions

- Thyroglossal duct cyst
- Median ectopic thyroid
- Epidermoid cyst
- Submental lymphadenopathy
- Thyroid nodule-isthmus
- Midline cervical fissure

Benign Lateral Neck Lesions

- Branchial remnant anomaly
- Lymphadenopathy, lymphadenitis, lymph node abscess
- Thyroid and parathyroid adenomas/cysts
- Lymphangioma
- Cervical teratoma
- Torticollis

Other Benign Neck Lesions

- Hemangioma
- Cervical thymus
- Ranula
- Parathyroid tumor
- Salivary gland tumor
- Thyroid cyst, adenoma, goiter

Malignant Neck Lesions

- Lymphoma
- Rhabdomyosarcoma
- Neuroblastoma
- Thyroid carcinoma
- Salivary gland tumor

Thyroglossal Duct Cyst

Midline neck masses in the neonate are rare and generally benign but can produce airway obstruction from tracheal compression. A thyroglossal duct cyst is usually not clinically evident until the child is 6–12 months old, since the cyst gradually enlarges as it accumulates the characteristic mucus-like fluid. The embryogenesis is intimately involved with the development of the thyroid gland, hyoid bone, and tongue. The thyroid gland begins as a diverticulum which extends from the foramen cecum at the tongue base down to the midline of the lower neck. This occurs while the hyoid bone is forming from the second branchial arch. As it descends, the thyroglossal duct passes closely to the hyoid bone (not through it). Once it reaches its final descent, the thyroglossal duct or "tract" regresses and disappears. Complete failure of descent results in a lingual thyroid, present at the base of the tongue, as the only functioning thyroid tissue. It is often inadequate in production of thyroid hormone, and the patient is usually "hypothyroid." Partial descent can lead to a median ectopic thyroid, lying across the trachea as a solid midline mass, which also may represent the only functioning thyroid tissue. Thyroid scan or ultrasound may be helpful, and evaluation of thyroid function-serum levels of T3 (triiodothyronine), T4 (thyroxine), and TSH (thyroid stimulating hormone) may help to guide management. Finally, during descent, elements of the thyroglossal tract may persist, forming a cyst. It typically is located at or near the midline and passes in a cephalad direction to the base of the tongue (foramen cecum area). The cyst is lined with epithelium and secretes a gelatinous, mucus-like material [1].

Diagnosis is suggested by location and findings on physical exam. A soft, cystic lesion is found near the midline, at the level of the hyoid bone, and it moves up and down with deglutition and protrusion of the tongue (Fig. 21.2). This may not be easily discernable or obvious in the struggling, uncooperative 2-year-old child! Ultrasound study may be helpful to distinguish it from a sold mass, such as a lymph node or median ectopic thyroid. It is usually non-tender, with the overlying skin clear and intact. However, due to its communication with the posterior pharynx, the cyst can become infected and present as an abscess.

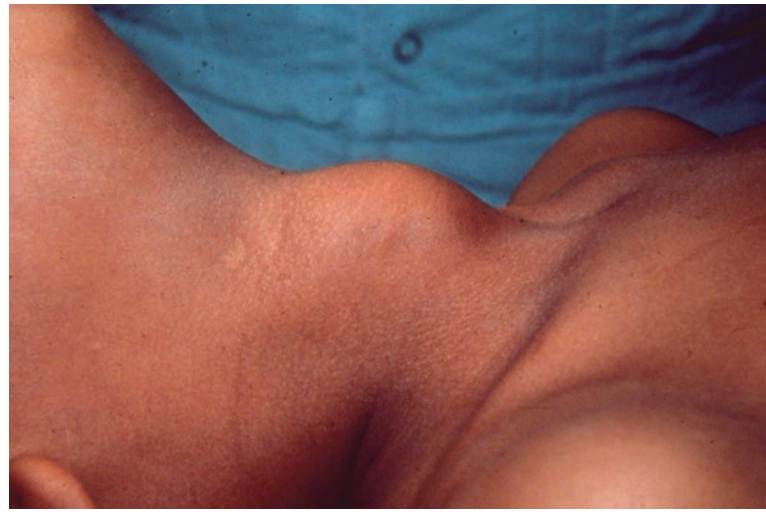

Fig. 21.2 Thyroglossal duct cyst

Treatment consists of complete excision of the cyst and tract to the base of the tongue (Fig. 21.3). Sistrunk (Mayo Clinic) recognized the high recurrence rate following surgery and advocated concomitant excision of the middle third of the hyoid bone, which is now recognized as the standard approach [2]. Locating the tract cephalad to the hyoid bone is not always easy. In fact, there may be more than one tract, and recurrence rates following excision approach 10%, especially with a history of previous infection [3, 4]. If the patient presents with an abscess, it should be drained and antibiotics administered, with excision postponed until the infection has resolved.

A thyroglossal duct cyst can also contain ectopic thyroid tissue, which later in life (in the fifth or sixth decade) has been associated with thyroid cancer—another indication of the need for excision [5–8].

Median Ectopic Thyroid and Lingual Thyroid

A solid midline neck mass may also represent median ectopic thyroid. This usually is the patient's only functioning thyroid tissue, and the patient is often hypothyroid. Preoperative assessment of thyroid function can determine if the patient requires supplemental thyroid hormone. If the patient is euthyroid, one can expose the ectopic thyroid; divide it in the midline, preserving the blood supply which enters the gland from the lateral aspect; and reposition each "half" of the gland beneath the respective sternocleidomastoid muscle [9]. In the hypothyroid patient, excision of the gland, followed by lifetime replacement of thyroid hormone, is not an unreasonable approach. Similarly, ectopic lingual thyroid can be found at the base of the tongue and, as above, represents the only thyroid tissue.

Dermoid and Epidermoid Cysts

During development, ectodermal elements may be buried beneath the skin surface, along the lines of fusion, leading to formation of inclusion cysts. They may contain hair follicles, sweat glands, and connective tissue. Located near the midline adjacent to the hyoid bone, an epidermal inclusion cyst may not be discernable preoperatively from a thyroglossal duct cyst. At surgical exploration, the contents of the cyst are quite different, containing sebaceous material and lacking intimate connection to the hyoid bone. Simple excision of the cyst will suffice. Similarly, one may find a pretracheal or submental lymph node which is removed for histologic confirmation and diagnosis. If during exploration, the lesion is found to represent a thyroid nodule, and then biopsy is warranted. If a thyroid lesion is suspected, then preoperative fine needle aspiration

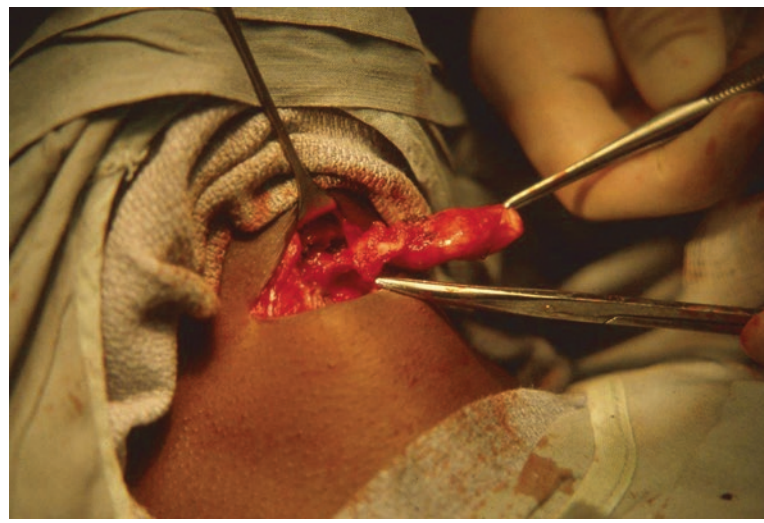

Fig. 21.3 Sistrunk operation: excision of thyroglossal duct cyst and middle portion of the hyoid bone

(FNA) is the preferred approach, as frozen section histology cannot differentiate follicular carcinoma from a benign follicular adenoma [10].

Midline Cervical Cleft

These congenital clefts represent failure of fusion of the branchial arches, resulting in incomplete epithelialization of the skin of the anterior neck near the midline. It is immediately apparent in the neonate. The vertically oriented cleft varies in length, and often contains underlying fibrosis, which can lead to limitation of neck extension and create a significant cosmetic deformity and contracture (Fig. 21.4). The skin basically sits directly on the underlying platysma muscle. There is usually a red to violaceous hue to the cleft. Initially it can "weep" and remain moist, but this tends to dry out over time. Treatment is by excision and primary closure in the smaller lesions and excision and closure by Z-plasty in the larger ones [11, 12].

Branchial Remnant Anomaly

Branchial remnants in the neck result from embryonic structures that fail to mature or persist in an aberrant fashion [13]. Postnatal remnants of the embryonic branchial apparatus produce a variety of abnormalities in the ear and down the lateral aspect of the neck. Sinuses, fistulae, and cartilaginous remnants are often noted in infancy. Cysts may not appear until early childhood, when they are filled with fluid to become a distinct mass. During gestational weeks 4 through 8, pairs of well-developed ridges (branchial arches) dominate the lateral cervicofacial area of the fetus. The arches are separated by paired external grooves (branchial clefts), matched internally with pharyngeal outpouchings (pharyngeal pouches). Each arch contains mesenchymal tissue (mesoderm) that develops into muscle, blood vessels, bone, and cartilage. Arches are covered externally by squamous epithelium (ectoderm) and by cuboidal epithelium (endoderm) internally. A complete fistula occurs more commonly than a blind-ending sinus tract. A true branchial cleft cyst is not seen that often.

The first branchial arch anomaly is rare, as illustrated by a large series of patients collected by Dr. Robert Gross, in which only one of the 308 patients had true first branchial remnants [14]. Preauricular cysts, sinuses, and fistulae are not included, as they represent an abnormal infolding with epithelial entrapment, as the branchial arches coalesce. A first branchial fistula passes from the neck, just below the angle of the mandible, to open in the external auditory canal. Surgical excision requires careful attention to avoid injury to the facial nerve and parotid gland, which lie in proximity [15].

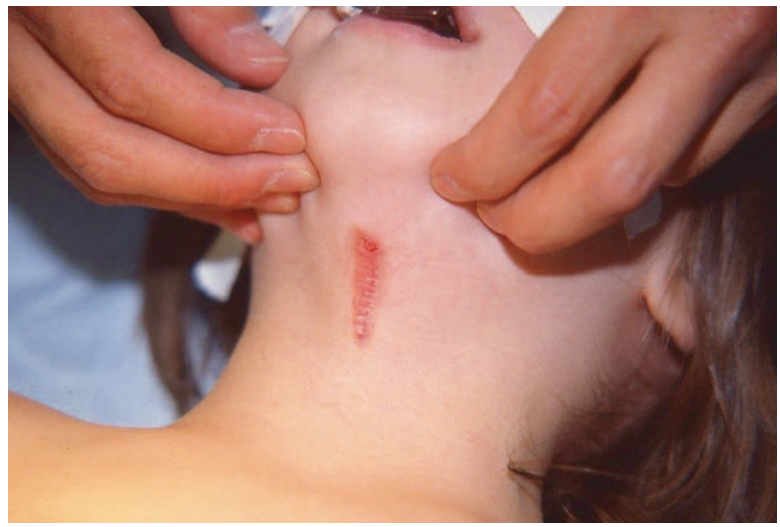

Fig. 21.4 Congenital cervical cleft

By far, the second branchial remnant is the most prevalent, with fistulae being more common than cysts [16]. In the infant, a fistula presents as a tiny opening along the anterior border of the sternocleidomastoid muscle (SCM), near the junction of the middle and lower third of the muscle (Fig. 21.5). It then dives deep to the muscle to ascend alongside the carotid sheath, passing between the branching of the internal and external carotid arteries, to enter the posterior pharynx at the tonsillar fossa. Intermittent drainage of mucus, or fluid resembling sputum, is common. Occasionally, the branchial remnant may become infected and require incision and drainage. Excision of the tract is curative and may require two separate parallel or "stepladder" incisions in order to carry the dissection cephalad, to reach the pharynx. The carotid vessels are never seen, as one maintains the dissection on the surface of the fistula. Injection of methylene blue dye into the tract may assist in the dissection.

Third branchial remnant anomalies are being found more frequently. The pyriform sinus fistula is a peculiar anomaly in that it is almost always found on the left side of the neck [17]. The fistula begins at the pyriform sinus or recess and extends inferiorly toward the upper pole of the ipsilateral thyroid lobe. It often presents as "suppurative" thyroiditis, causing inflammation as it terminates in or near the gland. A thyroid scan typically shows decrease uptake in the gland on the involved side, and the patient is clinically euthyroid with normal thyroid function studies (Fig. 21.6). Frequently presenting as an abscess, incision and drainage are followed by a course of antibiotics and then a period of waiting until the inflammation resolves, at least 6 weeks. Subsequently, a barium esophagram study, with attention focused at the level of the hyoid bone and pyriform sinus, can demonstrate the small sinus tract extending inferiorly toward the thyroid gland (Fig. 21.7). Surgical exploration with excision of the entire tract to the pyriform sinus is necessary to prevent recurrence. Extreme caution is exercised to preserve the external branch of the superior laryngeal nerve and recurrent laryngeal nerve. Partial thyroidectomy or lobectomy may at times be required to ensure complete removal [18–20].

Lymphadenopathy and Lymphadenitis

Cervical lymph nodes are readily palpable in most children over a year of age. The goal of the clinician is to recognize and diagnose those patients with significant or serious illnesses. Lymphadenopathy is defined in the child as a node greater than 1 cm in diameter. The most common cause is reactive hyperplasia, followed by granulomatous disease (i.e., cat scratch disease and atypical mycobacterial lymphadenitis),

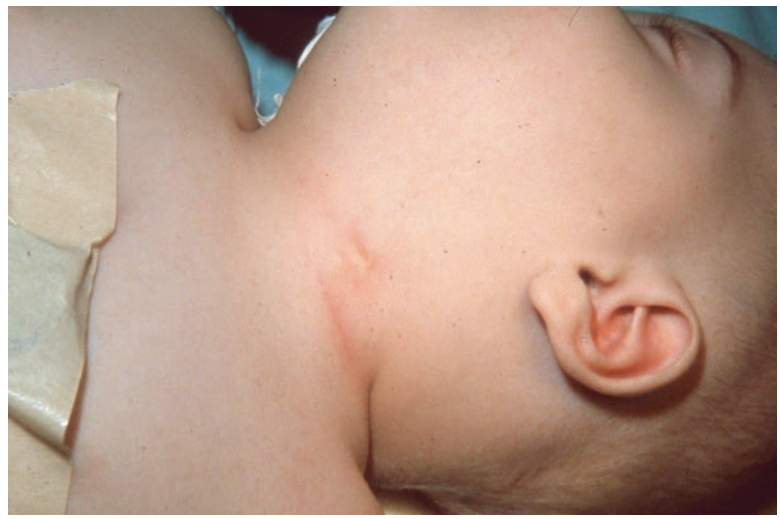

Fig. 21.5 Second branchial cleft fistula along anterior border of the SCM muscle, junction of middle and lower third

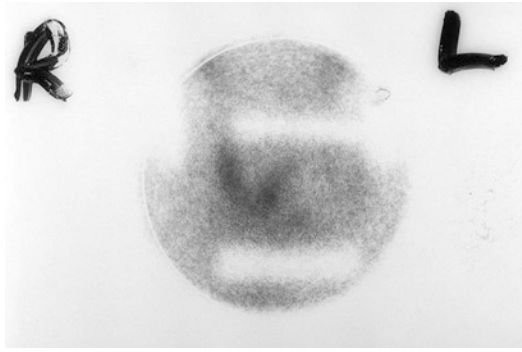

Fig. 21.6 Infected pyriform sinus fistula with secondary inflammation involving the left thyroid lobe; thyroid scan showing decreased uptake on the left

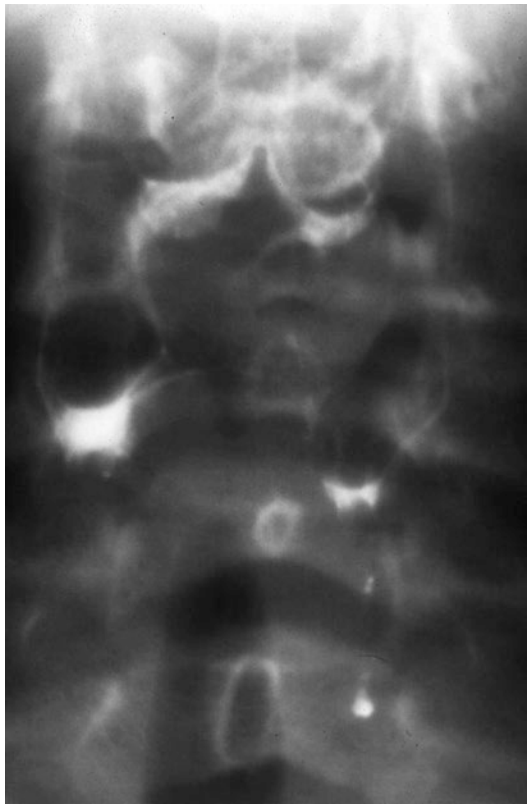

Fig. 21.7 Pyriform sinus fistula-barium esophagram weeks later, after inflammation resolved, showing tract extending from pyriform sinus to area of upper lobe of the thyroid

neoplasms (i.e., lymphoma), and chronic lymphadenitis [21–25]. It is impractical to biopsy every child with enlarged cervical nodes. Specific criteria help to identify those patients with a high probability of having serious underlying disease [26]. Firstly, any enlarged supraclavicular lymph node is of concern for possible lymphoma (Hodgkin's disease or non-Hodgkin's lymphoma). A CXR should be done initially to rule out mediastinal widening due to tumor mass and lymphadenopathy (Fig. 21.8).

Secondly, a fever exceeding 38 ° C for over a week suggests lymphoma or a disseminated bacterial or fungal infection. Thirdly, in the absence of systemic symptoms, a specific cause should be sought. Serologic tests are readily available for Epstein-Barr virus, cytomegalovirus (CMV) histoplasmosis, and cat scratch disease (*Bartonella* titers). When a skin test is used for *Mycobacterium tuberculosis*, be sure to also test the patient for anergy, to avoid a false-negative result [27]. Methicillin-resistant *Staphylococcus aureus* (MRSA) has become a major infection problem in hospitals as well as in the general population [28]. Skin infections often lead to lymphadenitis and enlarged nodes along the drainage route. An acute, cervical lymph node abscess is treated by incision and drainage and appropriate antibiotics [29]. The use of a small-vessel loop drain can minimize the postoperative care and simplify the treatment by avoiding daily packing changes. The abscess which has localized with suppuration and parchment-like changes in the skin is easily recognized (Fig. 21.9). Deeper lymph nodes require ultrasound study to determine if liquefaction is present, amenable to drainage or needle aspiration. Atypical mycobacterial lymphadenitis is a slowly developing, indolent process that tends to involve submandibular nodes. The infection can develop over a period of weeks and even months as it erodes through the overlying skin (Fig. 21.10). Surgical excision of the involved lymph node and skin is curative but should be approached cautiously to avoid injury to the mandibular branch of the facial nerve [30–32]. If the enlarged node or mass is indurated, non-tender, and fixed to the surrounding tissues, there is real concern for malignancy. In addition, any solid node which has increased in size over a 2-week period, or persists beyond 4 weeks without a decrease in size, should be considered for biopsy [33].

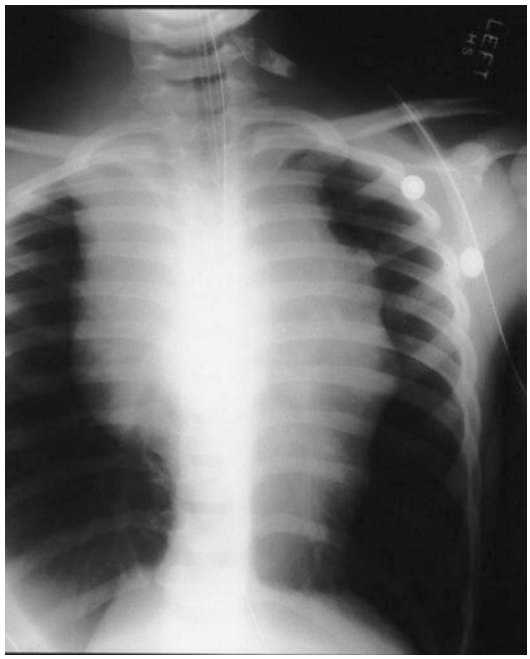

Fig. 21.8 Lymphoma with mediastinal lymphadenopathy, tracheal compression, and hyperexpansion of the lungs due to air trapping; positive supraclavicular lymph node biopsy

Lymphangioma

From the Greek "hygroma" meaning water tumor, this benign, uncommon cystic lesion arises from primitive lymphatic sacs which fail to coalesce, leading to sequestered lymphatic spaces that lack venous communication. Previously known as "cystic hygromas," they are often evident at birth or even by prenatal ultrasound. They can become large enough to produce dystocia, requiring delivery by C-section. Airway compression can also occur if they develop in the floor of the mouth and tongue (Fig. 21.11) [34]. They present as soft, discrete cystic masses which are non-tender and transilluminate readily. They are found in the lateral neck (75%) and axilla (20%), and the remaining develop in the mediastinum, retroperitoneum, groin, and pelvis (Fig. 21.12). Males and females are affected equally. Sudden enlargement may follow an upper respiratory infection, and spontaneous resolution is rare. Excision offers the best opportunity for cure and is generally delayed several months to a year after birth [35]. Occasionally, tracheostomy may be necessary to provide a secure airway. Lesions can be microcystic, macrocystic, or a combination of the two. Some lesions displace adjacent structures and are more easily removed, while others tend to infiltrate and insinuate, wrapping around major nerves, vessels, and muscles and rendering complete excision improbable without significant morbidity. Recurrence following an operative approach is common, and repeated operations are the norm. These operations are often difficult and challenging, and postoperative complications are common, including recurrence, infection, and nerve damage. A select group of patients, with small lesions, can sometimes be followed with observation, without surgical intervention [36]. Sudden enlargement of a mediastinal lymphangioma can occur if only the cervical portion is removed. CXR, MRI scan, and/or CAT scan will help to identify those cases where the cervical component extends into the mediastinum. If needed, the cervical excision can be extended into the chest via a median sternotomy.

Because of the recurrent nature of the disease and the frequent inability to excise the lesion for cure, other treatment modalities have been tried. Used either as a primary approach or for recurrent disease, these include sclerotherapy with bleomycin and with OK-432 (lyophilized incubation mixture of group A *Streptococcus pyogenes* of human origin or "Picibanil") [37–40]. Recently, the LigaSure vessel sealing system (LVSS) has been used with good success, sealing the lymphatics and decreasing lymphocele formation afterward [41].

Cervical Teratoma

From the Greek term meaning "monstrous growth," this disfiguring tumor of the neck in the infant can be so large as to cause dystocia and life-threatening airway obstruction immediately in the delivery room, as shown in Fig. 21.1. If detected antenatally, one can plan for an elective delivery by an EXIT (ex utero intrapartum treatment) procedure [42–45]. This requires a multidisciplined approach, coordinated with the anesthesiologist, neonatologist, obstetrician, and pediatric surgeon. The infant undergoes a

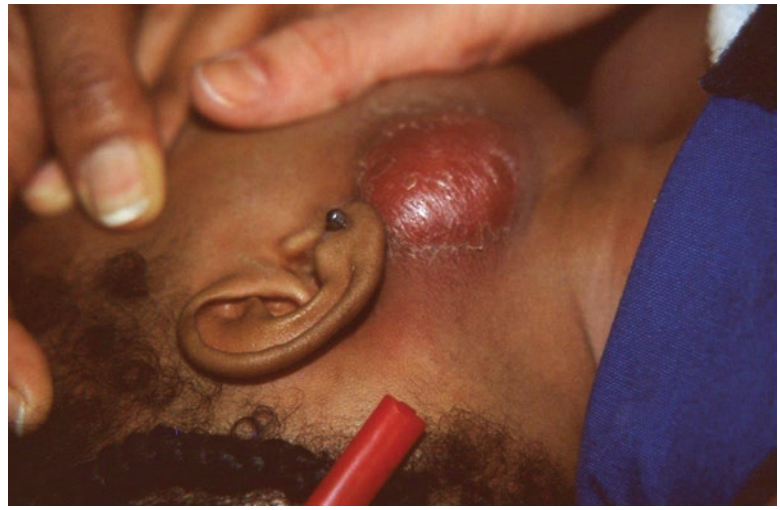

Fig. 21.9 Suppurative lymph node abscess secondary to MRSA

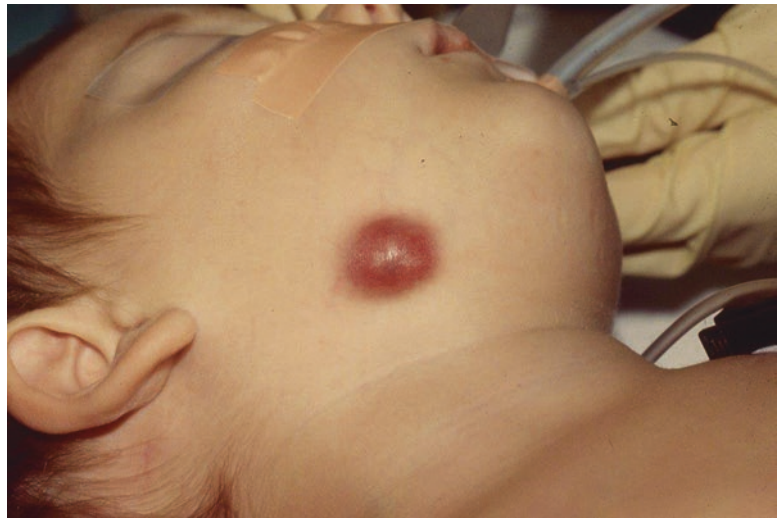

Fig. 21.10 Atypical mycobacterial lymphadenitis developing over 4–6 weeks, with involvement of the overlying skin

Cesarean section with partial delivery of the head and shoulders, without clamping of the umbilical cord. This maintains placental circulation and "buys time" for establishment of a critical airway either by intubation or by surgical tracheostomy. These tumors contain elements of all three germ cells (endoderm, ectoderm, and mesoderm); therefore, virtually any tissue in the body can be found. Calcifications are common due to bone and cartilaginous remnants. Although a teratoma does not represent a malignant tumor by histology, its presence can lead to an inability to secure an airway and even result in death in the delivery room. The underlying etiology is unclear, possibly related to aberrant development of stem cells from the thyroid gland. Formerly, these were known as "thyroid teratomas." Surgical excision is curative [46].

Torticollis (See also Chapter 11)

From the Latin "tortus" (twisted) and "collum" (neck), this deformity was first mentioned by Plutarch in describing Alexander the Great. Many causes exist in childhood, such as cervical hemivertebrae, ocular muscle imbalance [47], cervical lymphadenitis [48], acute fasciitis, Sandifer's syndrome [49], neural axis abnormalities, atlantocervical rotary displacement [50],

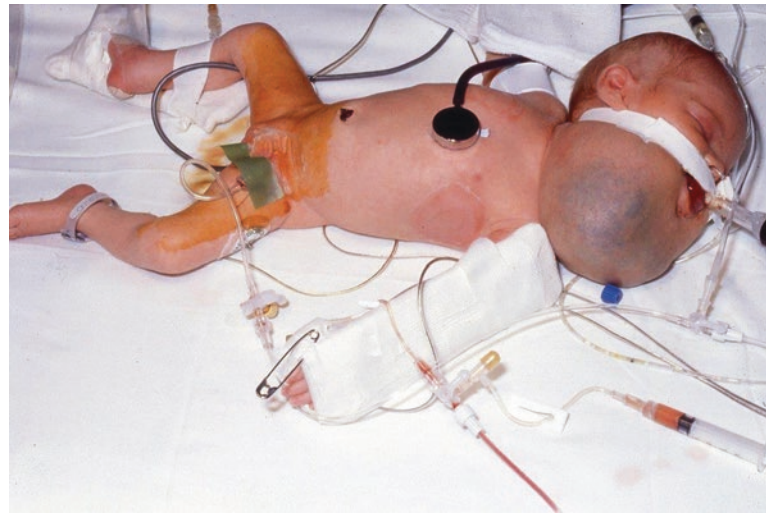

Fig. 21.11 Infiltrating lymphangioma involving floor of the mouth and tongue

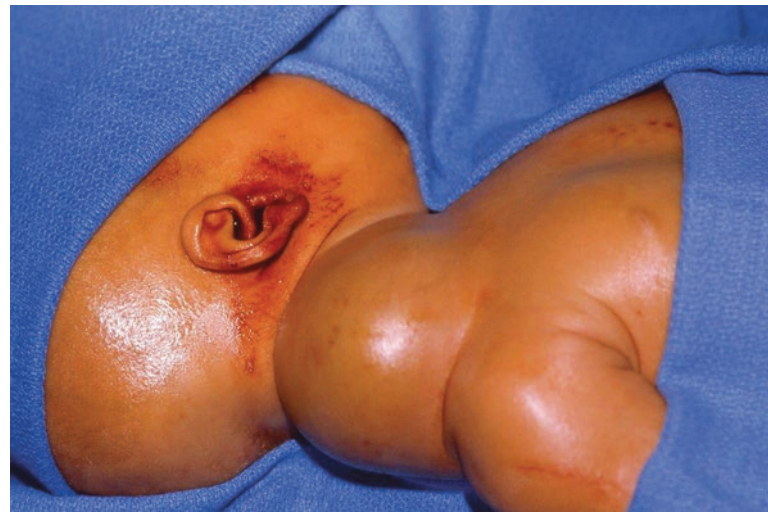

Fig. 21.12 Lymphangioma: typical location in posterior cervical triangle

cervical spine injury [51, 52], epidural hematoma [53], oropharyngeal inflammation, craniosynostosis [54], pseudotumor cerebri [55], posterior fossa tumors [56], and cervical spine tumors [57]. However, the most common type seen in the pediatric age group presents in the first 2–8 weeks of life, and the underlying cause is unknown. Breech presentation is present in only 20% of these patients, so traumatic delivery cannot be postulated as an underlying cause in the majority of patients [58]. Another theory is that of antenatal injury to the sternocleidomastoid muscle which leads to bleeding, muscle atrophy, and scarring, with subsequent fibrosis and shortening of the muscle [59]. It can present as a discrete mass or diffuse scarring throughout the muscle (Fig. 21.13). Other associated conditions related to formation of torticollis in the neonate include multiple births, small maternal pelvis, and oligohydramnios. Clinically, the head and ear on the affected side are pulled toward the shoulder, and the chin and face are rotated to the contralateral "normal" side. Left untreated, this can lead to plagiocephaly and hemihypoplasia of the face. Early on, physical therapy should begin with passive stretching and range-of-motion exercises [60]. The parents are encouraged to change the infant's sleeping position, increase "tummy time," and position the bed in the room to encourage the infant to look toward the affected side.

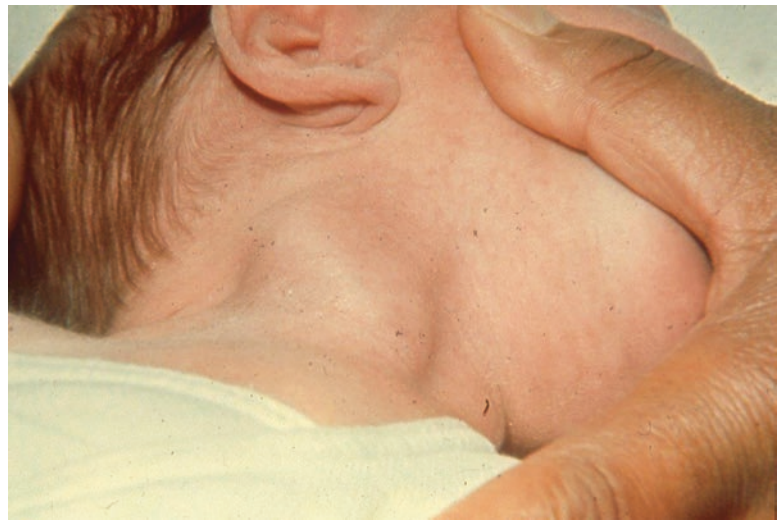

Fig. 21.13 Torticollis: discrete tumor mass in the SCM muscle

The use of a helmet cast or brace has been reported to be beneficial, used either as a primary treatment or following surgery [61]. Results, however, are mixed [62]. The helmet gives the best results if used early and is usually in place for about 6 months. The absolute indication for surgery is failure of a nonoperative approach with the development of hemihypoplasia [63–65]. Transection of the middle third of the SCM muscle and underlying fascia colli (Fig. 21.14) is followed by postoperative physical therapy and/or helmet casting. In the older child, it is important to rule out cervical spine instability, and the myriad other causes listed above, before proceeding to the operating room.

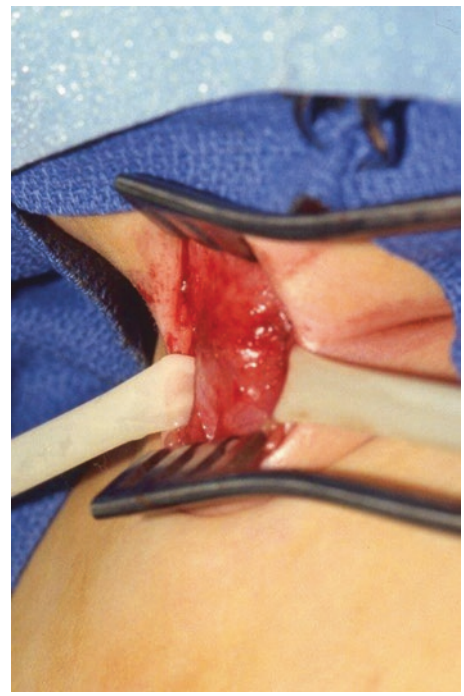

Fig. 21.14 Torticollis: division of SCM and fascia colli

Hemangioma

Vascular anomalies are among the most common lesions of the head and neck region in infants and children. The management of these lesions has been inconsistent and confusing, mainly due to a lack of a coherent and organized classification system. Finn et al. [66] and Mulliken et al. [67] have been instrumental in putting forth such a system that helps to identify these lesions correctly and thus improve the likelihood that they will be properly treated. Greene and associates have divided the lesions into vascular tumors, which include the common infantile hemangioma and vascular malformations illustrated by capillary malformations, venous malformations, lymphatic malformations, and arteriovenous malformations [68]. It is beyond the scope of this chapter to delve into this complicated issue in depth. But it is helpful to the orthopedic surgeon to recognize that there is a wide variety in these lesions and they each have unique characteristics. A long list of treatment modalities in the past

points to the difficulty in their management and their unpredictable behavior and response to these management schemes. These include surgical excision, radiation therapy, ligation or embolization of afferent vessels, injection of sclerosing solutions, pulse-dye laser, compression, propranolol, and alpha interferon.

The clinical behavior of congenital vascular lesions varies widely, from an innocuous, small infantile hemangioma that will resolve spontaneously to the large congenital vascular malformation, which can be present at birth and be associated with platelet entrapment, thrombocytopenia and hemorrhage from disseminated intravascular coagulation, the Kasabach-Merritt syndrome [69, 70]. Other vascular lesions can develop in or about the trachea, causing critical airway obstruction. Antenatal ultrasound has been useful to identify these patients, and an EXIT procedure may be necessary to establish an airway in the immediate postnatal period [71].

The most common vascular lesion is the "benign" infantile hemangioma. Usually not discernable at birth, they appear within 2–4 weeks of life. They initially may resemble a cutaneous stain or vascular marking. And over the next couple of months, a deeper component becomes noticeable. Draining veins are prominent, one of their hallmarks, and they go through a period of proliferation up to a year of age and then begin a slow process of involution, which can last up to 5–10 years (Fig. 21.15). Initial management is that of observation, to identify the lesion and determine if it will resolve spontaneously or follow a more treacherous course. Even the infantile hemangioma can be a serious management dilemma if it is located near the eye or face. In this instance, earlier intervention may be required to avoid disfigurement and interference with the visual axis. Intralesional steroids have proved to be helpful to shrink the lesion and allow for excision if needed [72–74]. During the period of involution, ulceration, bleeding, and infection are common problems. Use of simple measures such as local pressure, topical antibiotics, and judicious observation may suffice. Propranolol has also been used with excellent results [75]. With a failed response to corticosteroids, interferon alpha 2 is the second-line treatment strategy [76–78]. The child should be followed closely for adverse side effects. The effectiveness of steroids and interferon appears to be related to their inhibition of the angiogenic peptides—vascular endothelial growth factor (VEGF) and basic fibroblast growth factor (bFGF) [79].

If an infant or child presents with stridor and has an accompanying hemangioma elsewhere on the body, one should suspect an underlying hemangioma or vascular lesion involving the airway, usually in the subglottic trachea. Laryngoscopy and bronchoscopy are diagnostic, and repeated treatments with laser therapy, along with steroids, are indicated. The radiologist can be helpful to diagnose these lesions as well. Color Doppler ultrasound, CAT scan, and MRI scan can identify the anatomical extent and help form a treatment plan [80].

Surgical excision certainly has its place in the management of these lesions. It can be curative if the lesion is localized and able to be removed without sacrificing vital structures or causing significant deformity. It also is an important adjunct used with the other various methods of treatment [81].

In summary, the majority of these vascular lesions can be managed by the primary physician, with the occasional involvement of a specialist such as a hematologist, dermatologist, or surgeon. Observation is the first approach, followed by the use of corticosteroids, either orally or by intralesional injection. Then one enlists the other methods of treatment including propranolol and interferon, and in the case of the arteriovenous malformation, occlusion of the afferent "feeding" vessels may be necessary [82]. In the most serious cases, total circulatory arrest with deep hypothermia may be required [83]. The more complicated lesions will require a multidisciplinary team approach which can include most of the specialty areas of pediatrics. Successful outcome demands patience, an understanding of the natural behavior of each vascular lesion, its correct identification, and a willingness to involve specialists in the field and to refer the patient to a national vascular anomalies center when appropriate [79].

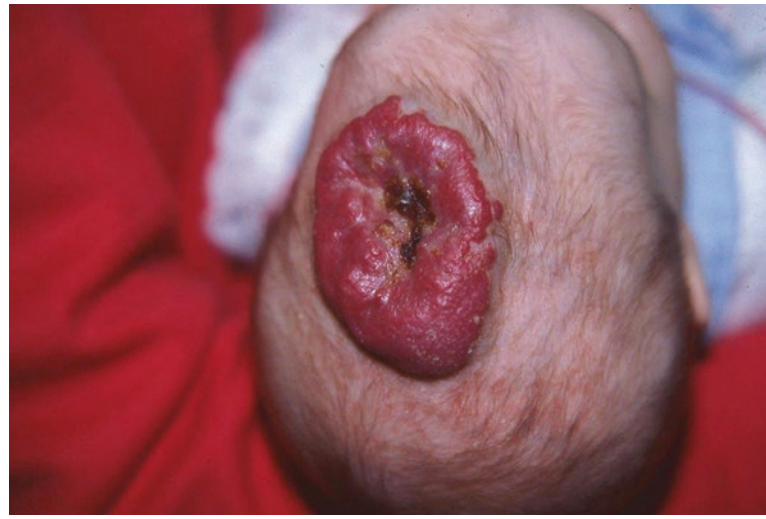

Fig. 21.15 Infantile hemangioma of scalp undergoing involution

Ectopic Cervical Thymus

By the sixth week of fetal development, the third pharyngeal pouch begins to divide into dorsal and ventral wings. The dorsal wing gives rise to the inferior parathyroid glands, while the ventral wing is the precursor for the thymus gland. Solid thymic buds form and migrate caudally along a thyropharyngeal duct to reach the anterior mediastinum. Various theories exist as to embryogenesis. Most embryologists believe that ectopic thymic cysts form from cystic degeneration of Hassall's corpuscles [84]. Ectopic solid thymus develops from failure of the gland to descend and therefore give rise to sequestered cervical thymus [85]. Cystic ectopic thymus in the neck is much more common than the solid variety, and it is usually asymptomatic, unless associated with underlying infection or hemorrhage within the cyst. These lesions are usually non-tender and can be found anywhere in the neck, from the level of the mandible down to the thoracic inlet. They tend to increase gradually in size as the child approaches puberty, but rapid enlargement is possible. Rarely, the mass can be large enough to cause stridor, dyspnea, dysphagia, or neck pain. They are more commonly located in the left neck and often have some form of communication or connection to the thymus in the mediastinum. Solid ectopic thymus has even been described in infants. Preoperative imaging studies should be obtained, including CXR, MRI scan, and/or CAT scan. CAT scan is used less often due to concern with exposure to ionizing radiation. The diagnosis may not be clear until final histologic review of the resected tissue. Excision is recommended, due to reports of latent malignant degeneration in cervical ectopic thymus [86, 87].

Ranula

Ranula is a form of mucocele or mucus retention pseudocyst (lacking epithelial lining) that forms in the floor of the mouth following rupture of the excretory duct of the sublingual gland. It can also arise as a congenital lesion from aberrant embryogenesis. The solitary cystic mass accumulates saliva and mucus and is generally asymptomatic but can become infected. The simple form is confined to an intraoral location beneath the tongue. The plunging ranula extends through the mylohyoid muscle to present as a bulging cystic mass beneath the mandible in the anterolateral neck (Fig. 21.16). The term comes from the Latin *Rana* meaning "belly of the frog," an apt description [88]. CAT scan is helpful in the diagnosis, especially to distinguish it from a branchial remnant or lymphangioma [89]. Excision of the pseudocyst and accompanying sublingual gland through an intraoral approach is curative [90, 91]. Recently, direct injection of sclerosing agents, such as O-K

432, has been used as initial treatment and may obviate the need for an operation [92].

Parathyroid Gland

The orthopedic surgeon is very familiar with calcium and phosphorus metabolism and with parathyroid hormone produced by the parathyroid gland under influence of the pituitary gland [93]. Hypercalcemia can be due to a number of causes, such as sarcoidosis, chronic renal insufficiency, and prolonged immobilization, to name a few [94]. Primary hyperparathyroidism, leading to hypercalcemia, can also result from a solitary parathyroid adenoma, which functions autonomously and is refractory to the normal feedback regulation of the pituitary-parathyroid axis. Neck exploration to locate a small, non-palpable parathyroid gland can be a daunting operation. With the introduction of the 99 m Tc-sestamibi scan, a small incision can be made over the offending adenoma, and using the gamma ray detector, the involved parathyroid gland can be removed [95]. Intraoperative measure of PTH is confirmatory. Occasionally, extensive remineralization of the bone, or the "hungry bone syndrome," can develop postoperatively, leading to profound hypocalcemia and even tetany. Close monitoring of the ionized calcium levels is important following surgery [96]. If all four of the parathyroid glands are enlarged and hyperplastic, then three glands can be removed and the remaining gland transplanted into a forearm muscle [97, 98]. Primary hyperparathyroidism due to hyperplasia has even been described in infancy [99].

Salivary Gland Disorders

Salivary gland disorders are uncommon in the pediatric age group and, when present, are usually benign. The most common benign lesion is the hemangioma, followed by lymphangioma and pleomorphic adenoma (benign mixed tumor). Most of these tumors involve the parotid gland, but less commonly the submandibular gland [100]. Hemangiomas can present early in infancy or early childhood and are readily diagnosed by color Doppler ultrasound. Most gradually involute and resolve spontaneously without intervention. Occasionally, they can be associated with rapid growth and sudden enlargement. These respond well to systemic steroids and interferon alfa-2a [101]. Lymphangiomas can infiltrate the parotid gland and involve the surrounding soft tissues and neurovascular structures. Surgical excision is often required; they do not involute and may require parotidectomy. Extreme care and an intimate knowledge of anatomy are requisite for a good outcome and to avoid damage to the facial nerve and its branches [102].

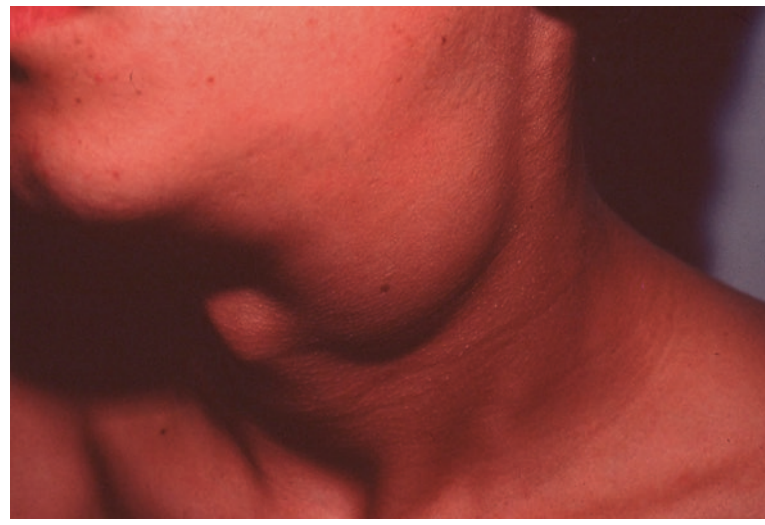

Fig. 21.16 Plunging ranula: left submandibular area

Pleomorphic adenomas present as a firm, slowly growing, non-tender mass in the parotid gland. Treatment is by surgical excision. The vast majority of these tumors are benign, although malignant transformation has been described. If the lesion is superficial, then superficial parotidectomy with removal of all gland external to the facial nerve is curative [103]. Preoperative workup may include a detailed history and physical exam and various radiologic studies including ultrasound, MRI scan, CAT scan [104], and percutaneous FNA biopsy [105]. Other benign lesions include inflammatory disease (both viral and bacterial sialadenitis) and chronic sialadenitis [106]. Viral parotiditis usually is self-limiting and usually resolves in a few weeks. Bacterial infections require administration of antibiotics and drainage of any abscesses (Fig. 21.17).

As uncommon as salivary gland tumors are, malignant tumors of the parotid and submandibular glands are even less common. Most malignant salivary tumors involve the parotid gland and on histology are mucoepidermoid carcinomas and less often acinar carcinomas [107]. These have been associated with radiation treatment of an earlier malignancy such as leukemia or lymphoma. Sudden enlargement of a solid, nonvascular parotid tumor, which is non-tender and associated with dimpling of the skin, arouses one's suspicion for a malignant process. Involvement of the facial nerve with paresis or paralysis is even more concerning. Treatment requires removal of all involved tissue, which usually means a total parotid resection (both superficial and deep lobes), with critical attention to avoid injury to the facial nerve and its branches [108]. If involved, a portion of the nerve may need to be resected, with intraoperative nerve grafting. Rhabdomyosarcoma may also arise within the parotid gland and is managed by biopsy for confirmation of diagnosis, followed by chemotherapy and/or radiation therapy [109]. Complications of treatment include facial nerve injury, fistula, Frey's syndrome (gustatory sweating of the face on the affected side), and recurrent tumor. The quality of life in pediatric patients surviving parotid tumors is excellent for benign disease. Outcomes for adolescent patients with malignant parotid tumors are not as encouraging. Allan and associates reported 96% survival rate at 5 years, decreasing to 83% by 20 years, with worse prognosis for adolescents compared to children younger than 15 years of age with parotid malignancies [107].

Lymphoma and Non-Hodgkin's Lymphoma

Hodgkin's disease, or lymphoma, usually develops in the lymph nodes or the primary lymphoid organs (spleen, thymus, tonsils, intestinal tract, and bone marrow). First described in 1832, it is character-

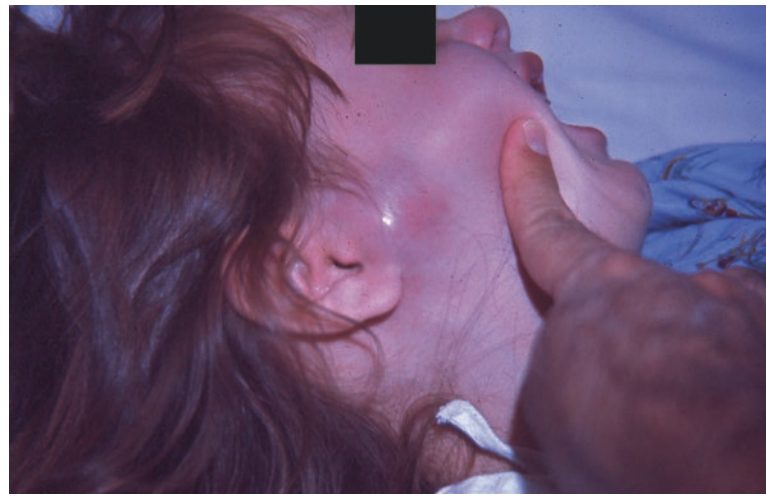

Fig. 21.17 Parotid gland abscess

ized by a multinucleated giant cell with prominent nucleoli, the so-named Reed-Sternberg cell [110]. It accounts for approximately 5% of malignancies in childhood and has been associated with prior Epstein-Barr (EB) virus infection. It can arise from lymphoid tissue nearly anywhere in the body, but in the majority (75%), it begins in the mediastinum with extension to the cervical and supraclavicular nodes. On palpation, the nodes typically are firm and rubbery in consistency, non-tender, and often fixed or matted, and the overlying skin is clear and normal appearing. The presence of such a node in the supraclavicular fossa is almost diagnostic of lymphoma. An open surgical biopsy is necessary, as a percutaneous biopsy does not provide adequate tissue for diagnosis [111, 112]. Prior to biopsy, a thorough workup is undertaken, beginning with a CXR and CT scan of the chest, abdomen, and pelvis. Mediastinal disease can compress the heart, and tumor encasement of the trachea can lead to disastrous consequences as the patient undergoes induction of anesthesia [113]. With inability to expel CO2 via passive recoil of the chest, the PCO2 rises to fatal levels, with cardiac arrhythmia and arrest. This is more common with non-Hodgkin's lymphoma. With severe upper airway obstruction, it may be necessary to obtain a biopsy under local anesthesia or even forego biopsy altogether and initiate treatment (steroids and chemotherapy) [114, 115].

The treatment protocol is predicated by staging. Historically, patients underwent a staging laparotomy, which included splenectomy, biopsy of multiple intra-abdominal lymph nodes, and bone biopsy [116, 117]. Currently, since nearly all patients receive chemotherapy, staging laparotomy is no longer required, as clinical staging is determined by radiographic studies (CXR and CAT scan of the chest, abdomen, and pelvis). Recently, PET (positron emission tomography) scan has been advocated for staging [118]. A central venous device (Broviac-type catheter or Infusaport) for administration of chemotherapy is usually placed during the biopsy procedure. Patients are further divided into staging subgroups: substage A (no symptoms) and substage B (symptoms present—fever > 38 C, drenching night sweats, pruritus, malaise, and weight loss of greater than 10% of body weight during the previous 6 months) [110].

The role of surgery is to confirm the diagnosis [119]. Hodgkin's lymphoma is represented by four distinct histologic presentations: nodular sclerosing (most common), lymphocyte predominant (best prognosis), mixed cellularity, and lymphocyte depletion (worst prognosis). Prognosis for cure is excellent, with 5-year survival rates approaching 90%. There are, however, particular risks and concerns related to the toxicity of treatment, manifested by cardiac toxicity, secondary malignancies, growth retardation, and bone growth retardation. Some treatment regimens also include radiation therapy which can cause lung damage, sterility, secondary thyroid cancer, and myocardial injury [120]. Postsplenectomy sepsis is no longer an issue, as staging laparotomy has been discontinued [121].

Non-Hodgkin's lymphoma is also found in childhood, and the workup and evaluation are similar. Burkitt's lymphoma more commonly presents as an intra-abdominal mass and requires laparotomy for diagnosis. Cure rates are excellent, as the tumor responds well to chemotherapy. Lymphoblastic lymphoma arises in the mediastinum and spreads to the supraclavicular and cervical nodes. This type of tumor may present with the superior vena cava syndrome (edema of head and upper extremities, dilated veins, dyspnea, and stridor). As above, if airway compression is severe, one may even need to initiate treatment prior to obtaining a biopsy [110].

Rhabdomyosarcoma

Rhabdomyosarcoma is the most common soft tissue sarcoma in children, with the majority presenting in the preteen years, occurring primarily in the head and neck region and in the genitourinary tract. It can arise from nearly every site except the bone. The extremities are involved in 15–20% of cases. Orbital tumors typically present with proptosis and ophthalmoplegia. Tumors arising in the sinuses and nasal passages can cause obstruction to mucus drainage. Workup includes diagnostic imaging, bone scan, bone marrow evaluation, and open biopsy of the tumor for diagnosis. Rhabdomyosarcomas of the head and neck are usually embryonal cell-type histology with excellent prognosis, especially true for

orbital lesions. These tumors are extremely sensitive to chemotherapy, and major surgical resection is to be avoided, especially if it would result in significant morbidity. Spread to regional nodes is uncommon. This is to be differentiated from those tumors arising from the parameningeal tissues, with early lymphatic metastases and a poorer prognosis. Incisional biopsy is followed by chemotherapy and, in some cases, radiation therapy. A second-look surgical excision may be required [122–124].

Neuroblastoma

Neuroblastoma is the most common, extracranial, solid malignancy of childhood, most diagnosed by age 10 years. Arising from neural crest tissue, in 70% of cases, it is found in the adrenal gland, paraspinal ganglion tissue, and retroperitoneum near the aortic bifurcation (islands of Zuckerkandl). It is also found in the cervical (2%) and thoracic (20%) regions. In the chest, it appears in the posterior mediastinum, near the thoracic inlet, and can present with Horner's syndrome (ptosis, miosis, and anhydrosis), indicating tumor involvement of the stellate ganglion [125]. If not present initially, it will almost certainly be present following surgical resection of a neuroblastoma in the upper, posterior mediastinum [126]. Most neuroblastomas in the head and neck area are metastatic from another site; therefore, a thorough evaluation is essential to find the primary and to detect other secondary lesions. Workup includes CXR, bone marrow biopsy, CAT scan, bone scan, and evaluation of urine for the presence of catecholamines [127]. These lesions typically present as a solid, non-tender, lateral neck mass. Other clinical features may include opsomyoclonus ("dancing eyes") and hypertension. Although the ultimate goal is removal of all tumor, this should not lead to damage or sacrifice of structures such as the brachial plexus, phrenic and vagal nerves, and major blood vessels (carotid and subclavian arteries). Tumors that arise in the thoracic and head and neck areas have a much better prognosis than those found below the diaphragm [128]. Second-look surgery is helpful following adjuvant therapy (chemotherapy and radiation therapy).

Outcome is best for those neuroblastoma lesions arising in the head and neck areas. Prognostic factors include age at presentation (the younger the better), staging (bad prognosis with metastases), and the presence of specific genetic alterations. N-myc amplification (>10 copies), allelic loss (1p36 locus), and allelic gain (17 q locus) are associated with a poor prognosis [129].

Thyroid Neoplasms

Relatively uncommon in the past, thyroid neoplasms are being recognized with increasing frequency, most likely due to the advancement in radiographic imaging techniques. The presence of a solid thyroid nodule in a child carries a real concern for a possible malignancy [130]. The evaluation begins, as always, with a detailed history and physical exam. One should then decide if the patient is euthyroid, and laboratory determination of thyroid function with measurement of T3, T4, and TSH is the initial step. If there is a family history of multiple endocrine neoplasia (MEN) syndrome, then there is particular concern for possible medullary carcinoma of the thyroid, so serum calcitonin levels are also measured [131]. A history of radiation therapy for a previous malignancy such as rhabdomyosarcoma or lymphoma, in the presence of a palpable thyroid nodule, arouses the suspicion for a secondary thyroid cancer [132, 133]. The nuclear plant disaster at Chernobyl led to a greatly increased number of Russian children who developed thyroid cancer [134]. A CXR is a useful screening test to determine if pulmonary metastases are present, but this is not as accurate as MRI or CT scan. A thyroid ultrasound can determine if the lesion is cystic or if solid-solid tumors are much more likely to be malignant. A thyroid scan will distinguish functioning from nonfunctioning nodules.

In adults, the accuracy of fine needle aspiration (FNA) biopsy is well proven. However, in younger children, its role is less clear [135]. Children tend to be more frightened and apprehensive. Deep sedation or even general anesthesia may be necessary for a FNA biopsy. A solid thyroid nodule is more likely to be malignant in a

child than in an adult. Also, many nodules are complex and contain both solid and cystic components. FNA may miss the malignant portion and falsely label the nodule as benign.

Thyroid surgery has a definite role in the treatment of both benign and malignant diseases [10, 136, 137]. Children with Graves' disease who are refractory to medical management, or the patient with Hashimoto's thyroiditis, respond well to thyroid resection. Increasingly, more extensive thyroid operations are being performed, both lobectomy and total thyroidectomy [138]. This can be performed safely in an institution with a good volume of these patients and with skilled, experienced surgeons. One must always be careful to avoid injury to the superior and inferior laryngeal nerves and to the parathyroid glands.

Papillary carcinoma, if unilateral, can be treated by lobectomy, but follicular thyroid carcinoma is frequently multifocal and should be managed by total thyroidectomy or subtotal thyroidectomy and postoperative I-131 ablation. Also, a follicular thyroid tumor is diagnosed by the presence of vascular invasion on histology, which cannot be determined on a frozen section during the neck exploration.

A unique situation exists in children who have the MEN-2 syndrome (medullary carcinoma of thyroid, parathyroid hyperplasia, and pheochromocytoma). Medullary carcinoma develops from the parafollicular C cells and presents at a very early age and has been described in the first few years of life. Prophylactic thyroidectomy should be done early in childhood to prevent this carcinoma, which eventually develops in 100% of these patients. Recent reports have even advocated thyroidectomy in the first year of life [139–141]. Determination of the RET proto-oncogene is useful to identify the patients at risk [142, 143].

Regardless of the type of malignancy, patients with differentiated thyroid cancer do well, even in the presence of metastases [144, 145]. Removal of the thyroid gland followed by I-131 ablation of any residual thyroid tissue is well tolerated. If there is any concern for possible damage to its blood supply, the parathyroid gland can be transplanted into the adjacent SCM muscle or a muscle of the forearm.

Summary

Many different pathologies arise in the neck of children whose diagnosis may present difficulties to the paediatric orthopaedic or neurosurgeon. Whilst otolaryngologists and paediatric general surgeons will be familiar with most of these lesions which primarily present in the anterior neck, this chapter serves as a primer for other disciplines. With awareness of their significance, appropriate referral and treatment can be achieved.

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Complication Types and Rates in Pediatric Cervical Spine Surgery

22

Urvij M. Modhia and Paul D. Sponseller

Introduction

Many types of surgery are performed in the pediatric cervical spine. The most common procedure [1, 2] is segmental arthrodesis for cervical instability, which may be caused by trauma, congenital malformations, atlantoaxial rotary subluxation, juvenile rheumatoid arthritis, tumors, iatrogenic causes, epiphyseal dysplasias, and various syndromes [1–4]. These abnormalities usually involve the upper cervical spine in younger children and the lower cervical spine in adolescents. In addition to bony abnormalities, some children are affected by disorders that require decompression of the spinal cord or removal of space-occupying lesions in or around the spinal cord, which may render the spine unstable and thus necessitate the need for arthrodesis.

Although pediatric cervical spine surgery has a high rate of success [1, 5], it may be associated with various complications [1, 3, 5, 6]. This chapter addresses the types, rates, and management options of these associated complications.

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Complications

The main factors that determine the rate of complications are the type of surgery performed, the method of instrumentation, and the patient comorbidities. The complication rates are affected due to the following factors: arthrodesis extending to the occipitocervical junction (OCJ), decompressive surgeries which involve more than three cervical spine segments without instrumentation, nonrigid fixation rather than screws as the method of instrumentation, and allograft rather than autograft is used when fusion involves the upper cervical spine.

Complications encountered in pediatric cervical spine surgeries include wound infection, wound dehiscence, cerebrospinal fluid (CSF) leak, vertebral artery injury, neurologic decline, fixation failure, pseudarthrosis, unwanted extension of the fusion, postlaminectomy kyphosis, and adjacent segment disease [3, 6]. Recently published studies by Elliot et al. [6], Hwang et al. [3], Anakwenze et al. [7], and Liu et al. [8] have addressed the rates of these complications (Table 22.1).

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Table 22.1 Types and rates of complications [3, 6–8]

Complication: wound infection	Rate, %
If arthrodesis extended to occipitocervical junction	3
If arthrodesis not extended to occipitocervical junction	1
Wound dehiscence	5
Pin site infections	32
Cerebrospinal fluid leak	1–7
Vertebral artery injury	0.6
Neurologic decline	1–4
Implant-related complications: fixation failure	
Screw pullout	2
Screw malpositioning	0.5
Pseudarthrosis	6
Postlaminectomy kyphosis	20
Adjacent segment disease	5
Unwanted extension of fusion (hyperostosis)	1

Surgical Site Infection

The cause of wound infection after cervical spine surgery is multifactorial. Most of the patient-related factors (e.g., age, sex, ethnicity, family history, and medical history) cannot be modified for orthopedic procedures, with the exception of improvement in nutritional status [9, 10]. Among surgery-related factors, wound infection rate increases if the arthrodesis extends to the OCJ (approximately 3% versus 1% if it does not) [3] and if there is an overt CSF leak during surgery (13% versus 2% without) [8]. The use of a halo crown, a common method of immobilization in children, is also associated with infections at the pin sites; the pin tract infection rate has been reported as approximately 32% [11, 12].

Superficial surgical site infections may be managed by dressing changes and a course of oral antibiotics, depending on the microorganism. Some superficial wound infections may require parenteral antibiotics and wound debridement. Deep surgical site infections are usually managed by wound debridement and parenteral antibiotics, with every attempt being made to retain instrumentation for stability [13, 14]. The use of antibiotic powder locally in the surgical bed is still variable and surgeon dependent, with additional studies needed to determine the efficacy.

The use of a halo crown for traction or a halo crown with vest for immobilization is a common treatment method in children; however, halo use is associated with a pin track infection of about 30% in some series [11, 12]. This is best prevented by using an adequate number of pins, usually eight to ten in younger children, due to the diminished skull thickness and the need to load share pins. Routine pin care should be done on a daily basis. Treatment of pin tract infection includes pin tract care and oral antibiotics for superficial infections. Deeper pin tract infection, which may lead to loosening, needs to be treated with intravenous antibiotics, with removal of the affected pin if loose, and potential new pin reinsertion at a different location.

Wound Dehiscence

Wound dehiscence occurs more commonly with extension of the arthrodesis to the OCJ and with overt CSF leak (approximately 5%) [8].

Management of a wound dehiscence consists of debridement and then soft tissue mobilization and closure of the incision, if it can be done without undue tension. However, if skin edges are difficult to approximate, then occasionally it is advisable to close the deep fascia and allow the skin to heal by secondary intention. The use of the vacuum-assisted wound closure is also justified to manage wound dehiscence [14]. A severe wound dehiscence may require soft tissue reconstructive procedures, and plastic surgical colleagues are invaluable in this situation, with early consultation recommended.

CSF Leak

The incidence of CSF leak in pediatric spinal procedures is approximately 1–7% and is related to the type of procedure performed [8]. Leaks occur most commonly in patients with reoperations, connective tissue disorders, or procedures requiring decompression. CSF leaks may become complicated by pseudomeningocele formation or may manifest as a CSF leak from a surgical

incision (termed overt CSF leak) [8]. Patients may present with postural headaches, wound dehiscence, wound infection, chronic subdural hematomas, and/or meningitis.

There are nonoperative and surgical treatment options when a CSF leak is suspected or present. Nonoperative modalities include bed rest with the head of the bed down and/or placement of a lumbar drain (CSF diversion by subarachnoid catheter). Operative interventions include dural repair with continuous locking suture, use of muscle/fat graft, and use of dural graft for complicated dural defects [8].

Vertebral Artery Injury

The incidence of vertebral artery injury is approximately 0.6% and is most commonly seen during C1-C2 arthrodesis [6]. The risk of vertebral artery injury secondary to screw fixation is greatest with attempted placement of C2 screws. Bilateral vertebral artery injury may lead to vertebrobasilar stroke and brainstem infarction [6, 15]. If a suspected vertebral artery injury occurs during C1-C2 instrumentation, it is best managed by creating a tamponade effect via the insertion of a screw, use of fibrillar mesh, and use of various hemostatic agents such as gel foam or FloSeal (Baxter BioSurgery, Westlake Village, CA). If the vertebral artery on one side is suspected of being injured, then a method of fixation on the contralateral side such as an intralaminar C2 screw is required to negate the catastrophic possibility of a second side vertebral artery injury. Postoperative angiography is then recommended to study the extent of arterial patency, in order to guide treatment recommendations for prevention of injury/clot propagation.

The use of contemporary screw fixation into the upper cervical spine mandates a preoperative CT scan in order to study the anatomical feasibility of screw placement in C1 and/or C2. Anatomical variants of the vertebral artery course are present, and understanding the relationship of the vertebral artery to the isthmus of C2 is paramount prior to attempting transarticular screws or C2 pars screws.

Iatrogenic Neurologic Complications

Potential neurologic complications, which can occur during any spinal procedure, include paresthesia, radiculopathy, quadriparesis, quadriplegia, cord syndromes, and death (Fig. 22.1a, b) [3, 6, 8, 15–17]. The rate of these complications ranges from 1% to 4%. Neurologic complications are more common with the use of wiring techniques than with screws in the pediatric cervical spine, given the passage of wires into the spinal canal [3, 6].

Management of neurologic complications includes maintaining a mean blood pressure of more than 85 mmhg, improved oxygenation, intravenous fluids, and removal of any implant that could have caused neurologic decline [18]. Additional imaging via CT and/or MRI might be useful to identify any compressive abnormality that could be treated by additional surgical intervention and decompression.

Implant-Related Complications

Spinal implant-related complications include screw pullout, malposition of implants, wire cutout, and screw-rod-wire failures. In the pediatric cervical spine, these complications may be attributed to a variety of reasons, such as smaller anatomical structures, increased segmental motion, and increased ligamentous laxity [3]. Modern screw techniques with the associated potential complications will be discussed elsewhere in this book.

Pseudarthrosis

Pseudarthrosis is a commonly seen complication in pediatric cervical spine surgery. Factors mentioned above (surgical site infection, dural leak) are frequently associated with this complication. In addition, arthrodesis in the cervical spine is frequently performed for various congenital syndromes that are associated with osseous, vascular, and neurologic abnormalities [3]. Wiring techniques of fixation results in a significantly

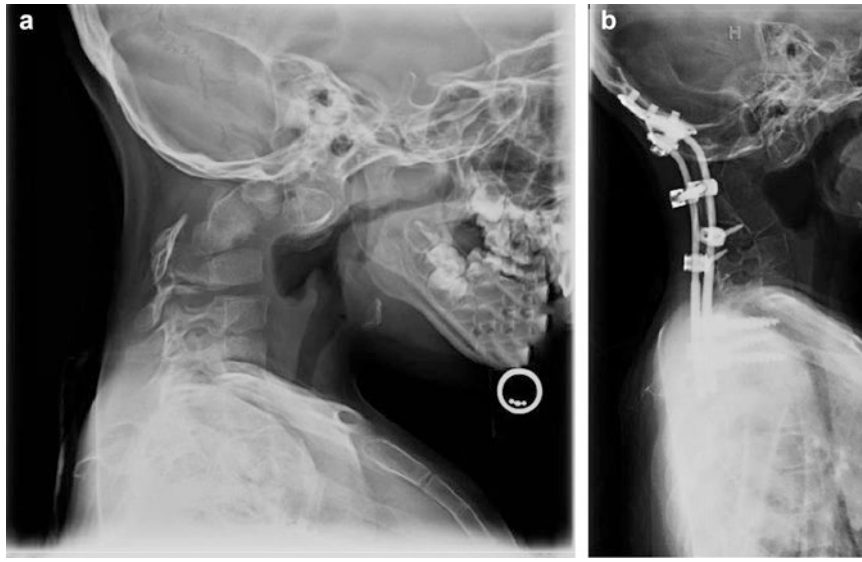

Fig. 22.1 (**a**) Radiographs of a 6-year-old child with severe congenital cervical kyphosis. (**b**) Postoperative radiograph after patient underwent posterior spinal arthrodesis with kyphosis correction, which was complicated by left-side C5 nerve root palsy after surgery that resolved after 6 months

higher incidence of pseudarthrosis (6%) than does rigid fixation (1%) [3]. The incidence of pseudarthrosis has been shown to be higher with the use of allograft than with autograft [19]. However, this may be mitigated by more rigid fixation, especially if the fusion is in the subaxial cervical spine. Fusions which involve the craniovertebral junction or C1-C2 are best treated with autograft.

Recent technological advances have made it feasible to insert smaller profile screws and rods in the pediatric cervical spine, allowing better fixation with safe insertion [3, 6].

The management of pseudarthrosis includes revision arthrodesis with improved fixation and bone grafting (Fig. 22.2a–c) [3]. The use of biologic substances such as bone morphogenic protein (BMP) in the pediatric cervical spine is controversial and generally not needed, given the osteogenic potential of children. BMP may be considered in posterior cervical cases where there is a higher incidence of pseudarthrosis such as trisomy 21, NF-1, and revision surgery.

Postlaminectomy Kyphosis

Laminectomy, often performed in the pediatric spine for intraspinal abnormalities [7], is usually associated with removal of the lamina and interspinous ligaments with subsequent loss of posterior tension. In the pediatric cervical spine, the incidence of postlaminectomy kyphosis, a well-known complication in the spine literature, may be as high as 20%. Because of this high incidence, simultaneous arthrodesis may be needed if decompression includes three or more cervical spine levels or crosses the cervicothoracic junction. If arthrodesis is not done at the time of laminectomy, then consideration of orthotic support for a period of time may be warranted to prevent deformity. Laminoplasty is also used as a method to avoid arthrodesis and prevent kyphosis but with varied results [7, 20]. Recommended treatment of this complication is kyphosis correction via posterior instrumented spinal arthrodesis with/without anterior spinal arthrodesis (Fig. 22.3a, b).

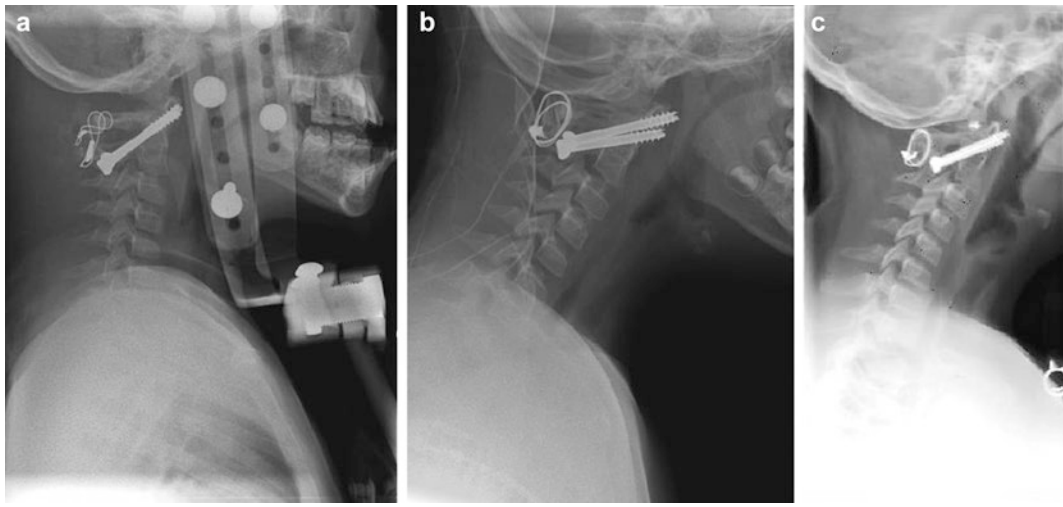

Fig. 22.2 (**a**) Postoperative radiograph of a 5-year-old child with Down syndrome with C1-C2 instability who developed a deep wound infection after instrumented spinal arthrodesis and later developed pseudarthrosis requiring reoperation. (**b**) Radiograph after with implant removal, revision instrumentation, and repeat bone grafting. (**c**) Follow-up radiograph documenting successful C1-C2 fusion

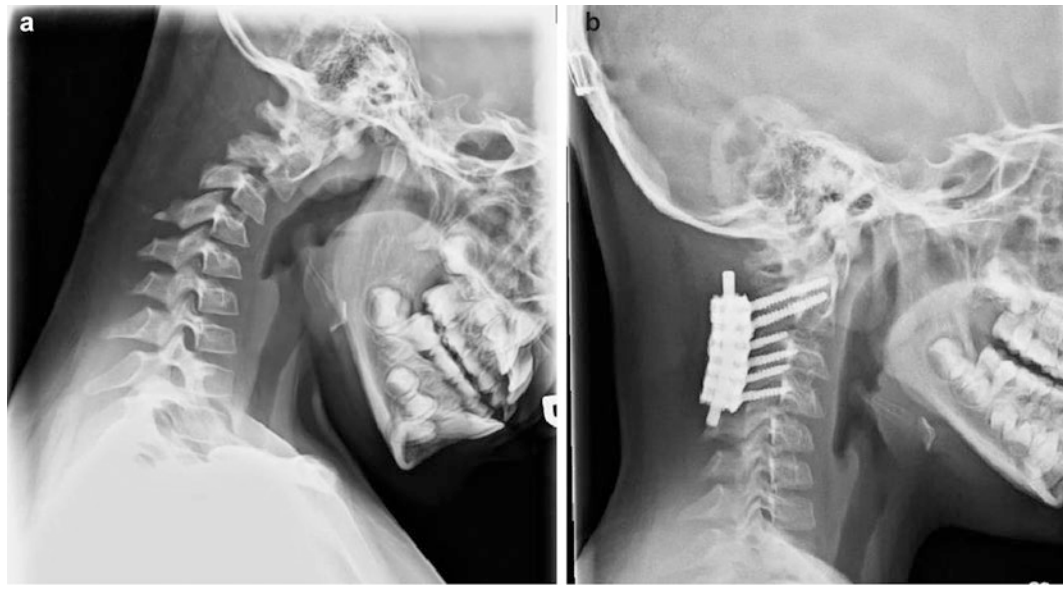

Fig. 22.3 (**a**) Radiograph of a 6-year-old child who had an Arnold-Chiari malformation treated by decompression that later resulted in kyphosis. (**b**) Postoperative lateral radiograph after the patient underwent posterior instrumented spinal arthrodesis with kyphosis correction

Adjacent Segment Disease/ Junctional Kyphosis/Fracture of Adjacent Vertebrae

This disease spectrum encompasses adjacent disk degeneration, junctional kyphosis, and fracture of adjacent vertebrae. The literature reports a 3–46% incidence of radiographic adjacent segment degeneration and a 21–26% incidence of proximal junctional kyphosis, after spinal fusion for adolescent idiopathic scoliosis [21, 22]. However, data in the literature are insufficient for identifying the rates of adjacent segment disease, proximal junction kyphosis, or adjacent vertebral fracture in the pediatric cervical spine. Additional studies are needed to quantify the rates of this complication in the pediatric cervical spine. Most patients with these complications are asymptomatic and do not require intervention [3]. Patients with junctional problems which are symptomatic may benefit from nonoperative management, including physical therapy with extensor muscle strengthening and pain management interventions. Operative management may include proximal/distal extension of the fusion with/without decompression (Fig. 22.4a, b) [23].

Hyperostosis

Hyperostosis (unwanted extension of fusion) is another common complication in the pediatric cervical spine [3, 6, 24]. Risk factors include exposure of unnecessary levels and postoperative halo immobilization. Extension of fusion is more commonly seen in C1-C2 arthrodesis that extends to C3, rather than in cases of subaxial cervical spine arthrodesis. This complication may potentially be avoided by prevention of overexposure at the junctions of the surgical site. However, children are osteogenic and autofusion at adjacent subaxial levels may occur, which in general are asymptomatic and require no intervention.

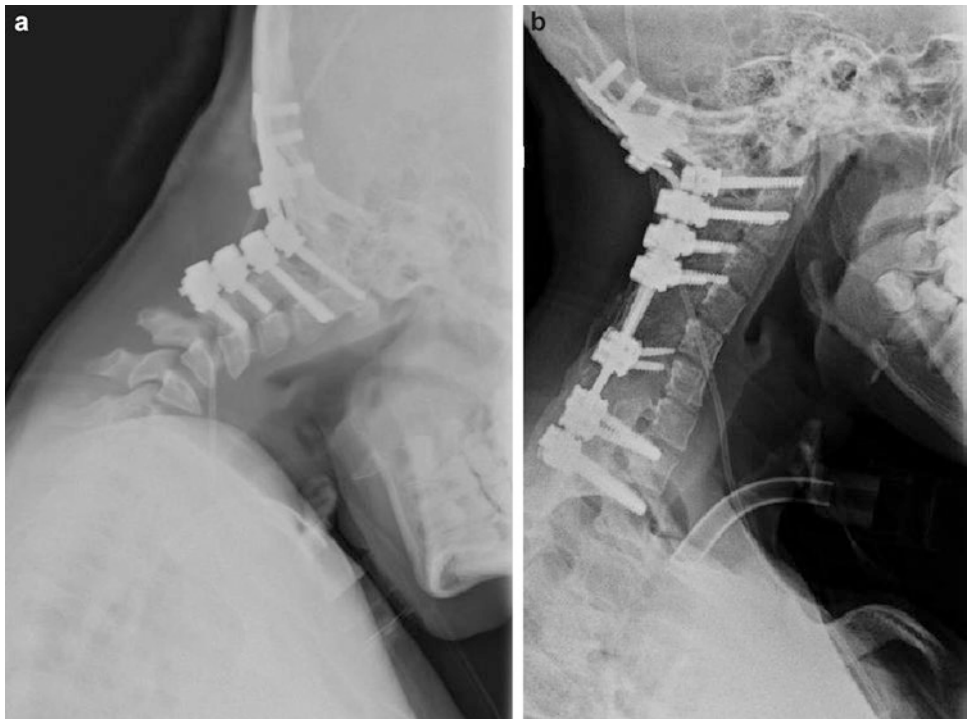

Fig. 22.4 (**a**) Lateral radiograph of an 8-year-old child with junctional kyphosis after occiput to C4 fusion for traumatic occipitocervical dissociation and spinal cord injury. (**b**) Lateral radiographs documenting improved alignment and fusion after extension of instrumentation with arthrodesis

Summary

Complications of cervical spine surgery that are unique to children encompass failure of fixation, unintended extension of the fusion, and additional deformity because of growth. Many of these events do not require revision surgery. However, discussion of these possible outcomes may help to provide realistic expectations for the patients and their families.

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