Essentials of Spine Surgery

Alpaslan Şenköylü
Federico Canavese
Editors

Foreword by
Jean Dubousset

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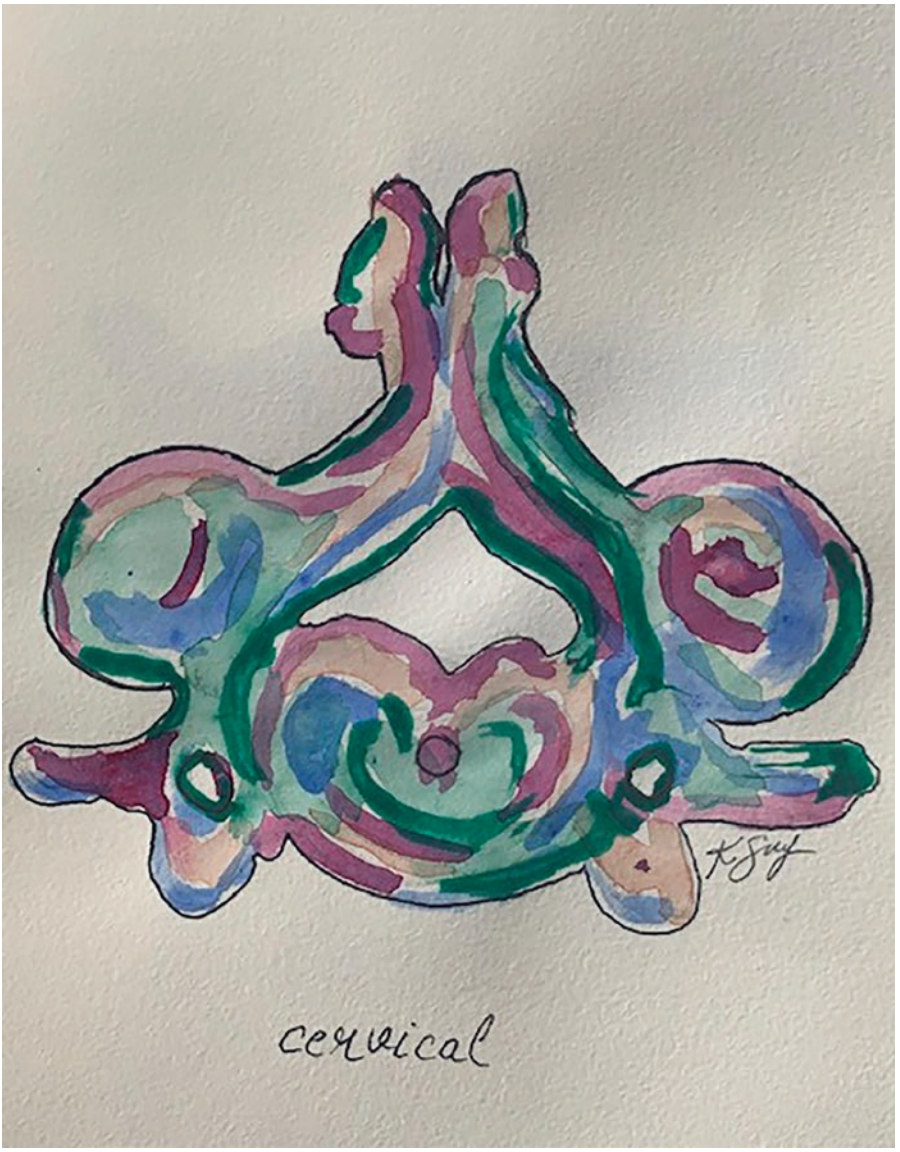

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"To our families and mentors..."

A.S. and F.C.

"Science is the only true guide in life"

Mustafa Kemal Ataturk

Foreword

The very beautiful, generous, and ambitious goal of the editors, Alpaslan Şenköylü and Federico Canavese is to relay information to the young generation of spinal surgeons. Pediatric and adult orientation, the basic spinal knowledge, pathology at any age, and major treatment principles as well as the clinical and imaging aspects regarding multiple presentations are introduced in a concentrated manner.

Hoping for both patients and the physicians to get the full benefit, each chapter is didactically introduced to the readers and supported by numerous educational videos and illustrations. I am sure you will find this very useful and practicable.

Each patient is unique and can be assessed in numerous approaches. With that being said, it is vital to follow a "check list" during an examination to obtain a detailed and an organized patient report ensuring an efficient treatment.

About diagnosis, a complete clinical and neurological examination is frequently better than many sophisticated imaging devices or even histopathological interpretations. An effective diagnosis includes concordant signs and symptoms narrowing down possible disorders. This book will help you narrow down these possibilities as well as providing you with suitable advice.

Most of the spinal disorders are clearly defined in this book enabling fellows and trainees to make a plausible diagnosis. According to my experience, instead of asking a patient or their parents, it is better to make the choice yourself and present it as clearly as you are convinced. They can accept or refuse but will never feel guilty if some difficulties occur postoperatively. This is for me one of the ethical bases in our professional life.

In any case the spinal surgeon must always evaluate advantages and disadvantages for each specific patient. When the decision is made, the surgeon will perform his surgery in the best conditions.

In this book, important key points are frequently presented to prevent a major pitfall, which is why I am convinced that this book will give you an excellent introduction for your future work in spinal surgery.

Paris, France — Jean Dubousset

Preface

The spine surgery subspecialty has a long and steep learning curve. Throughout this path, it is sometimes difficult to find a compact source that contains fundamental information on every aspect of this discipline. The purpose of this unique book is to give you the basis to understand paediatric and adult spine disorders including trauma.

We have deliberately asked the authors of the various chapters to respect a fixed scheme. In fact, the structure of each chapter is the same throughout the whole book, so that you can easily orient yourself between clinical, imaging, and instrumental examinations, differential diagnosis and treatment principles. We hope the concise and easy-to-read structure of each chapter will allow you to avoid major diagnostic or treatment pitfalls.

The chapters have been enriched with clinical videos and with a series of appendices that present the most commonly used classifications in spine surgery.

Be aware that the realization of this book has been possible thanks to the enthusiastic participation of some of the most eminent spine surgeons of our era, both orthopaedic- and neuro-surgeons, who have synthesized their incredible experience in the chapters you are preparing to read. This book would have not been possible without their diligence and commitment, and we would like to thank all of them for their enthusiastic contribution. Our special thanks go to Jean Dubousset, one of the legends of spine surgery who peer-reviewed all chapters and certainly further improved the overall quality of this book.

We are sure that you will be able to make good use of the authors' suggestions and advice.

We wish you a good reading!

Ankara, Turkey — Alpaslan Şenköylü
Lille, France — Federico Canavese

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Part I

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Pediatric Cervical Injuries

1

Moyo C. Kruyt and F. Cumhur Öner

1.1 Definition

Pediatric cervical spine injury is an uncommon but significant condition, present in 1% of the very young and 3% of the adolescent trauma patient admissions. The cause of these injuries is predominantly due to transportation accidents and sports [1]. However child abuse should be considered especially in the presence of other skeletal injuries.

1.2 Natural History

The cervical spine is the most vulnerable part of the pediatric spine with about 80% of the spinal fractures occurring in this region compared to <50% in adults. The younger the child the more vulnerable the cervical spine and the more proximal the lesions occur. This is mainly due to the relatively huge head and weak ligaments. Nevertheless, high forces are still needed for these injuries which are reflected by the high rate of concomitant spine fractures [2] and the relatively high mortality (5%) in these young patients [3]. Also spinal cord injuries (SCI) are relatively frequent with up to 16% in young children; the neurological symptoms fortunately are often temporary.

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1.3 Physical Examination

In the emergency setting, the basic principles of ATLS apply. Children suspect of cervical trauma should be immobilized with head-blocks, and for physical examination, the log roll should be used. The use of seatbelts should not argue against injury, as these are used incorrectly in >50% [4]. Physical and specifically neurological examination of a young child can be difficult because of limited attention and cooperation. It should be performed systematically by an experienced physician. Since neurological symptoms may worsen, the neurological exam should be repeated until stable (Video 1.4).

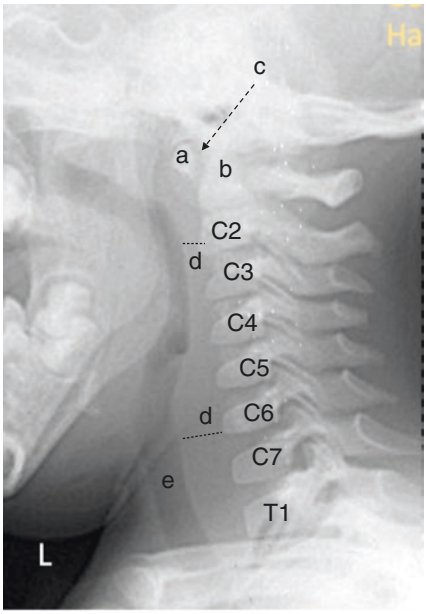
1.4 Imaging

After vital stabilization, radiological examination of the cervical spine can be done. For that purpose, plain radiographs have the advantage of less radiation; however, subtle fractures can be missed. In the case of unconsciousness, a multitrauma survey, or the need for a computed tomography (CT) scan of the brain, including a CT of the cervical spine is a logical step (Video 1.4). Be aware of the relatively large head that causes forward position and anteflexion. A cushion between the scapulae or at least removal of the extra elevation of the head should be standard in children. In case of neurological symptoms, magnetic resonance imaging should be used to visualize ligament injury, potential SCI, and follow-up of these lesions; according to the mechanism of injury, and clinical presentation, SCIWORA syndrome must be ruled out (Chap. 4).

For initial assessment, the alignment of the vertebrae should be verified including T1. This can be done by drawing lines anterior and posterior to the vertebral bodies and spinous processes in the sagittal plane. The atlanto-dental interval and dens to basion (skull) distance should also be assessed, as well as soft tissue swelling (Fig. 1.1). Specific features of pediatric cervical spine are discussed below.

1.4.1 Anatomical Considerations

The development of cervical spinal vertebrae is complex and varies greatly in the age and order of the events. All vertebrae initially consist of three ossifying nuclei, two posterior and one in the front, linked with neurocentral cartilage. The posterior arches fuse around 3–5 years of age, and fusion with the vertebral body usually occurs 1–3 years later. During puberty, secondary nuclei develop consisting of the transverse and (two) spinous processes and the annular ring apophyses. A complete fusion of the vertebra may occur after the age of 20. Typical exemptions are C1 and C2. C1 does have three primary nuclei, but no vertebral body, and the position of the synchondrosis varies greatly. C2 is actually a fusion between C2 and the original vertebral body of C1. The dens consists of two primary nuclei that usually fuse in utero, but can be interpreted as a fracture in the first years. The dens fuses with the



a. Anterior arch of C1
b. Dens axis
c. Atlantodental interval (ADI) (should be <5 mm in children)
d. Prevertebral soft tissue
e. Tracheal shadow

Dens must be below the foramen magnum
Synchondroses should be fused >6 years
Global alignment
Soft tissues
T1 must be visible

Fig. 1.1 Radiographic examination of the cervical spine. (By courtesy of Prof. Senkoylu)

body of C2 between 6 and 11 years. This synchondrosis can fracture or fail to fuse, resulting in an os odontoideum that can cause instability. The dens has an additional nucleus at the tip that ossifies around 3 years and normally fuses with the dens around 12 years. If not, it is referred to as ossiculum terminale. The cervical facet orientation is relatively horizontal and becomes more oblique with age. Together with some wedging in C3, the relatively loose ligaments, and weak muscles, this allows for kyphosis and translation up to 4 mm especially between C2 and C3 (Fig. 1.2).

1.5 Differential Diagnosis

For children, the same strategy as used for adults can be applied (Chaps. 5 and 6). Many classification systems have been developed, but none of them has proven to be perfect in terms of reliability and accuracy to predict instability and the need for stabilization. For the cervical spine, a distinction between high cervical (C0–C2 = axial) and lower cervical (= subaxial) is helpful. The presence of neurological symptoms and (congenital) anomalies are important modifiers. For the axial region, an increased distance between dens and basion (>10 mm) is indicative of instability like rupture of the alar ligaments. C1 ring fractures are difficult to recognize because of the synchondroses; in extreme cases, the tip of the dens can subside into the foramen magnum, which can cause a dangerous basilar impression. Most important of C1 is the integrity of the transverse ligament, which may be torn or avulsed when

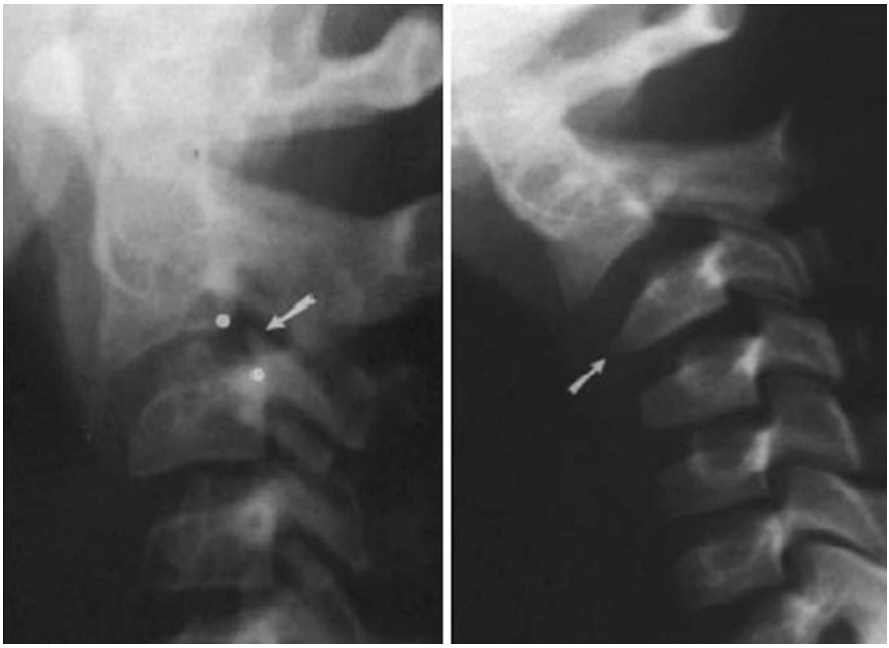


Fig. 1.2 Physiological anteposition of C2–C3 and wedging of C3

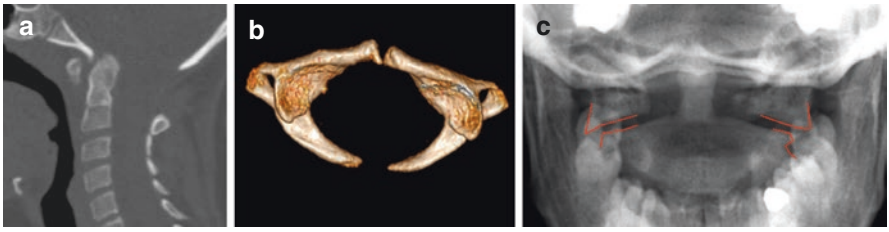


Fig. 1.3 (**a** and **b**) Subsidence of dens in foramen magnum, due to a (congenital) non-union of posterior laminae and a fracture through the anterior synchondrosis; (**c**) assessment of lateral position of C1–C2. When >7 mm in total, the transverse ligament may be insufficient

the lateral masses of C1 project >7 mm lateral from C2 in the anterior-posterior plane, or if the atlanto-dental interval (ADI) exceeds 5 mm (Fig. 1.3). Dens fractures can most easily be seen on the lateral radiograph. Be aware of the subdental synchondrosis. C2 hangman fractures are uncommon and usually a type of spondylolysis/olistesis through the synchondrosis [5].

Subaxial injuries are less common in children and behave comparable to in young adults (Chap. 6). We prefer the AO classification that makes a distinction between compression-only (Type A), tension band injury (Type B), and translation in any plane (Type C) [6]. In addition the facet fracture is considered an important

modifier in this classification, especially in the case of a floating lateral mass or (sub) luxation.

SCIWORA syndrome to be ruled out (Chap. 4).

1.6 Treatment Options and Expected Outcomes

For treatment decisions, the most important, but also controversial, parameter is (in)stability. Instability may be acute, which means that the mechanical integrity will fail with loading; this one is easy to recognize, usually with CT imaging. Neurological instability is more difficult; it means a condition where cord or nerve injury has occurred, and (intermittent) compression can worsen the neurological outcome. MRI is usually needed as well as regular follow-up. Most difficult to recognize and treat is the long-term instability which may cause severe deformities, pain, or even neurological problems. In children, the remodeling potential should be considered at the same time, which can improve the posttraumatic deformity considerably. Instability is always a reason for stabilization. The younger the child the less invasive this stabilization can be achieved. Halo fixation and especially Minerva casts are well tolerated and treatment of 1 to 2 months is usually sufficient. In the case of deformity, closed reduction techniques should be considered such as tong or halo-gravity traction (Videos 1.1 and 1.4) or transoral reduction of dens fractures. Open reduction with or without fixation is a valuable option which can usually be performed with standard cervical spine instrumentation even if this restrains the further development of the canal diameter which has largely developed by age 4 [7].

A painful neck without a radiological sign of fracture should be followed until normalized especially in the case of torticollis. If this takes more than 1 or 2 weeks, advanced imaging is recommended. After external stabilization with halo or cast, we typically wean with a soft collar. After surgical stabilization, this is usually not required.

1.7 What Family and Patients Should Know?

The prognosis is related to the severity and the location of the cervical spine injury.

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Thoracolumbar Spine Injuries in Children

2

Mehmet Kaymakoglu and Muharrem Yazici

2.1 Definition

Pediatric spine injury constitutes a small percentage (4%) among all trauma cases, and thoracolumbar injuries (30% to 40%) are even rarer compared to cervical injuries among all pediatric spine trauma [1]. Although most of the cases can be managed with conservative treatment, a careful physical exam and adequate knowledge about the features of pediatric spine injury are essential to prevent devastating complications.

Pediatric patients should not only be considered as "little adults" and it should be taken into account that they have many anatomical and physiological differences. This has also an importance in thoracolumbar injuries, and there are many differences in the management compared to adults (Chap. 7). The spinal column of children starts carrying adult properties at 8 years of age, and the injury pattern changes before and after that age. The children have a growing spine and the decision of fusion should be made very carefully, as it diminishes the growing potential. Additionally, the ligamentous laxity and the superior location of the pivot point of the head, which is larger relative to the body, lead up to a higher risk for distraction

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injuries. More horizontal and immature facets and immature paraspinal muscles also reduce the stability of the spinal column. Studies revealed that an elongation up to 2 cm is possible during trauma in children and this elongation protects the bony structure from fracture but the spinal cord has more limited flexibility up to 5–6 mm. Thus, a clinical entity called SCIWORA (spinal cord injury without radiographic abnormality) with neurological symptoms may occur without any sign of radiological finding in the pediatric population, and this increases the importance of the physical examination of the whole spine in the Emergency Department (ER). SCIWORA syndrome can also have a delayed onset, up to 4 days, and neurological follow-up of patients is therefore important (Chap. 4).

2.1.1 Mechanism of Injury

The main mechanisms of injury are distraction and compression. Besides more elastic properties of the pediatric spine, high-energy trauma tends to damage the vertebra through its relatively weak ossification centers such as vertebral endplates and facet joints. Biomechanical studies showed that in case of high bending stresses, Salter-Harris type 1 fracture occurs through the weakest point of the spinal column: the growth plates. Motor vehicle accidents constitute the most common reason (>50%) of pediatric spine trauma, especially in children older than 10 years old. Falling from height is also an important reason for those under 10 years old. Since the thoracic spine is biomechanically supported by the rib cage, the injury rate is less than the cervical and lumbar regions [2].

2.2 Physical Examination

The management of pediatric spine trauma necessitates a multi-disciplinary trauma team. After the evaluation of Glasgow Coma Score (GCS) (Appendix F) and Advance Trauma and Life Support (ATLS) protocol for children, immobilization of the spine and a cervical collar should be provided immediately. Neck hyperflexion should be avoided when using a spine board to prevent airway obstruction and spine injury. A complete neurological examination must be performed (Video 2.4); including motor and sensory evaluation, genital and rectal examination, and reflexes. Palpation of the entire spinal column should be made; and any abnormalities such as bruising, step-offs, or open wounds should be noted. Fatal intra-abdominal and head injuries often accompany pediatric spine trauma; thus, additional injuries should be studied and excluded before any treatment intervention.

2.3 Imaging

Anterior-posterior (AP) and lateral “full spine” radiographs are mandatory for the initial radiographic evaluation. Unlike the adults, in case of any neurological findings or in the need for further evaluation, a full spine computed tomography (CT)

scan is not recommended because of the excessive radiation exposure. Magnetic resonance imaging (MRI) should be the preferred study in such cases. Furthermore, MRI gives the clinician the advantage of the ability to assess the ligamentous structures (e.g., posterior ligamentous complex) and soft tissues around the spinal column. It also enables to classify the patients with SCIWORA according to MRI abnormalities of the spinal cord. If the patient is unable to get in MRI, a very short section CT for the suspected/injured spinal levels can be done for further evaluation.

2.4 Treatment Options

Most of the spinal injuries in children are managed conservatively, as bony fractures and neurological deficits are rarely seen. The treatment algorithm is dependent on the neurological status of the patient and the degree of instability of the spine as adults. The Thoracolumbar Injury Classification and Severity Score (TLICS) system has been used for adults to provide better outcomes for both conservative and surgical management, and a new study approved its validation among pediatric patients (Table 2.1) [4]. For those for whom surgical management is mandatory, the surgeon must be familiar with the possible effect of multilevel spinal fusion which disturbs the growth potential of the spine, especially in children younger than adolescence [5]. It should be considered that spinal alignment of the spine evolves during the child's growth, and a patient-specific sagittal alignment should be restored. The Risser sign can be used for the remaining growth and remodeling potential of the child (Appendix M). According to Pouliquen et al., [6] stable compression fractures have excellent outcomes with

Table 2.1 Thoracolumbar injury classification and severity score system

Type of injury	Points
Morphology	
Compression	1
Burst	2
Rotation/translation	3
Distraction	4
Disruption of the posterior ligamentous complex	
Intact	0
Suspected	2
Disrupted	3
Neurologic status	
Intact	0
Nerve root	2
Cord, conus medullaris: complete	2
Cord, conus medullaris: incomplete	3
Cauda equina	3

The sum of all points: 0–3: Conservative management, ≥4: Surgical management. If the sum is equal to 4, patient-specific management can be chosen (adapted from Vaccaro et al. [3])

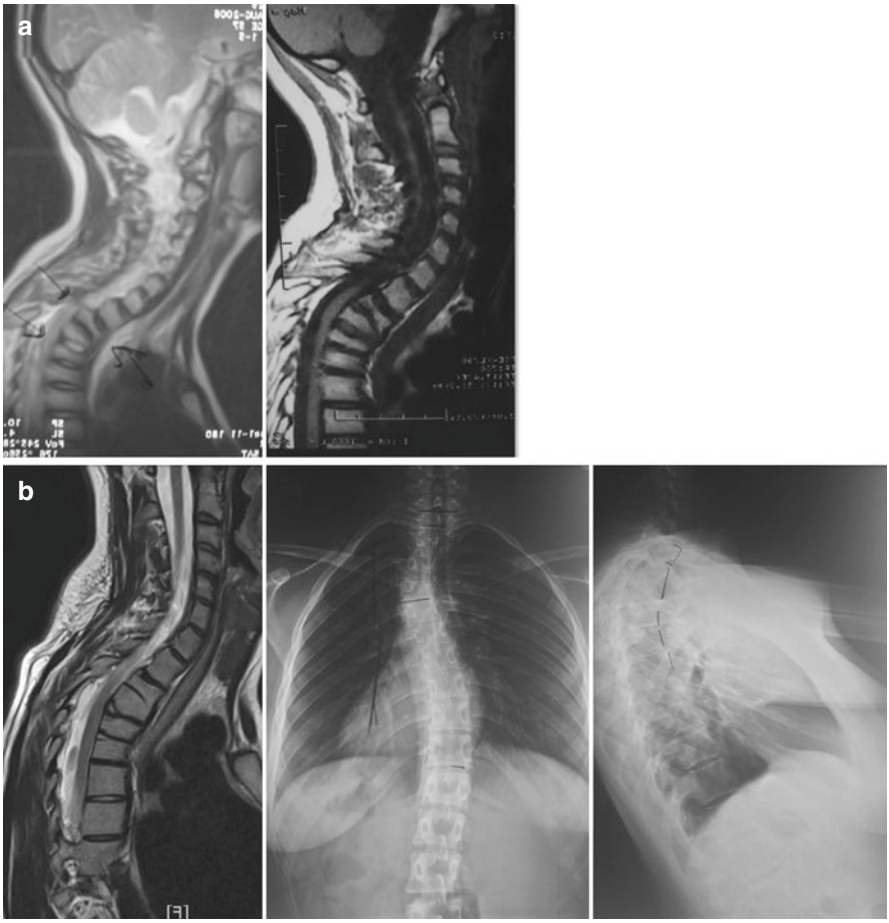

Fig. 2.1 A 7-year-old female patient with multiple proximal thoracic vertebral fractures was treated conservatively (**a**). After more than 10 years of follow-up, coronal and sagittal alignments were restored despite mild scoliosis and vertebral heights were regained (**b**)

nonsurgical management, especially in children with Risser sign of 0–1. Another study with 33 years of mean follow-up showed that single-column compression fractures in children have good remodeling potential with the ability to regain the vertebral height (Fig. 2.1) [7]. Fracture dislocations, injuries with neurological deficits, and unstable fractures according to the TLICS scoring system should be managed with surgical intervention (Fig. 2.2) (Video 2.4). High-dose corticosteroid treatment in children is controversial, and there is not any randomized trial with a high level of evidence suggesting the use of corticosteroids.

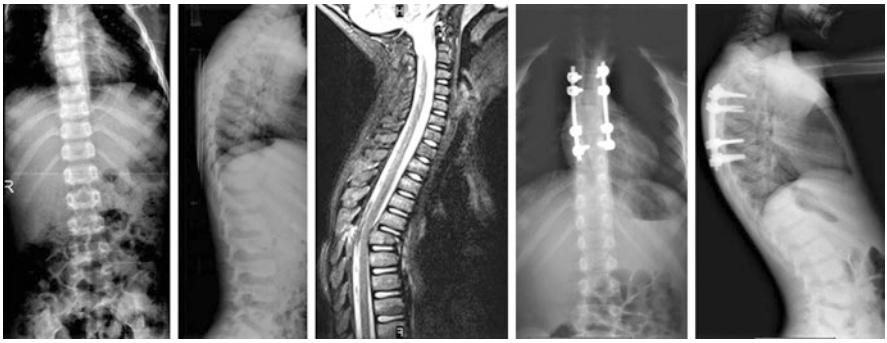

Fig. 2.2 An 8-year-old male patient with T6 fracture and posterior ligamentous complex injury. Mechanical instability was treated with a short segment posterior instrumentation and fusion (Video 2.3)

2.5 Expected Outcomes

Outcomes of non-surgical and surgical treatment in neurologically injured patients mainly depend on the initial severity of spine injury. Mild-to-moderate injuries recover normal or nearly normal, whereas severe injuries are difficult to heal regardless of the choice of surgical or non-surgical treatment. Burst fractures without neurological injury can be treated with hyperextension bracing relying on the remodeling potential of the canal in children. The surgical intervention seems to provide better radiological outcomes, but there is not any difference in functional scores between non-surgical and surgical groups. However, surgeons should consider the risk of spinal deformity after mild spinal injuries or SCIWORA syndrome (Chap. 4) in patients treated conservatively (Fig. 2.3). Thus, the follow-up of pediatric patients with deformity is crucial in conservative management and growth-sparing instrumentation has to be the treatment of choice if the patient has growth potential. Non-fusion spine stabilization as internal bracing is another treatment strategy in the growing spine (Fig. 2.4). Excellent radiological and functional outcomes have been reported by several authors with internal bracing. The only disadvantage of this technique is the need for the second stage to remove the hardware after 1 year.

In case of permanent neurological deficit, the risk of developing paralytic scoliosis with/without pelvic obliquity is extremely high, close to 100%.

2.6 What Should Patient and Family Know?

Spine fractures in children rarely need surgical intervention unless there is a high-energy trauma with fracture-dislocation or neurological deficits. Outcomes are good at experienced spine centers, and a growth-friendly strategy is considered if a

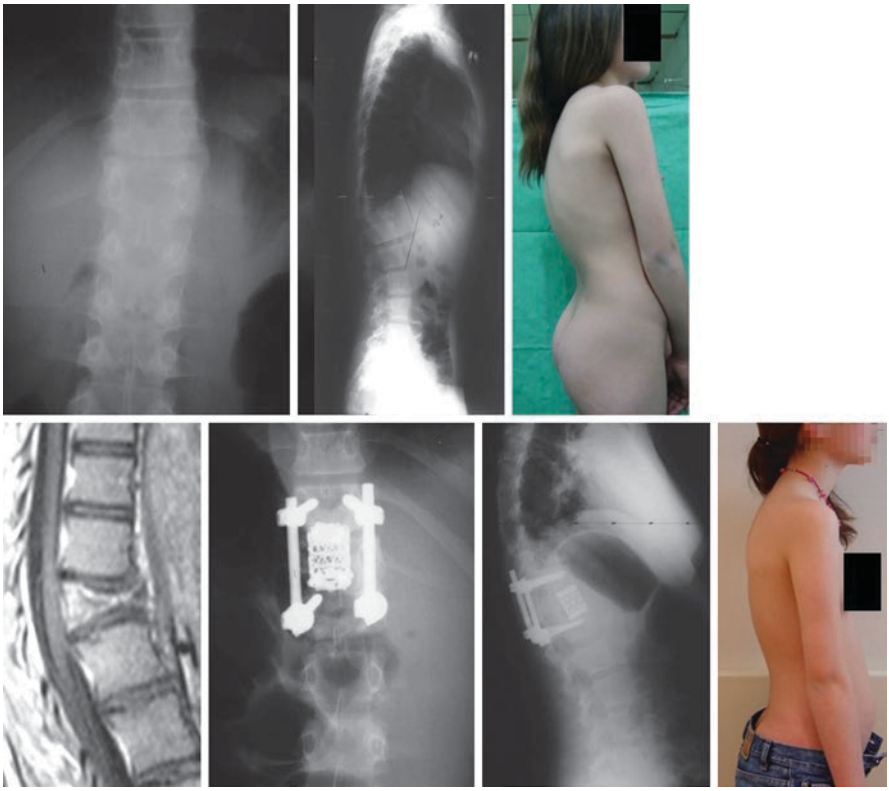

Fig. 2.3 An example of possible complications in conservatively treated patients. A 16-year-old female admitted to our hospital after 4 years of a T12 burst fracture. Her kyphosis was corrected with anterior corpectomy and posterior instrumentation and fusion (Video 2.3)

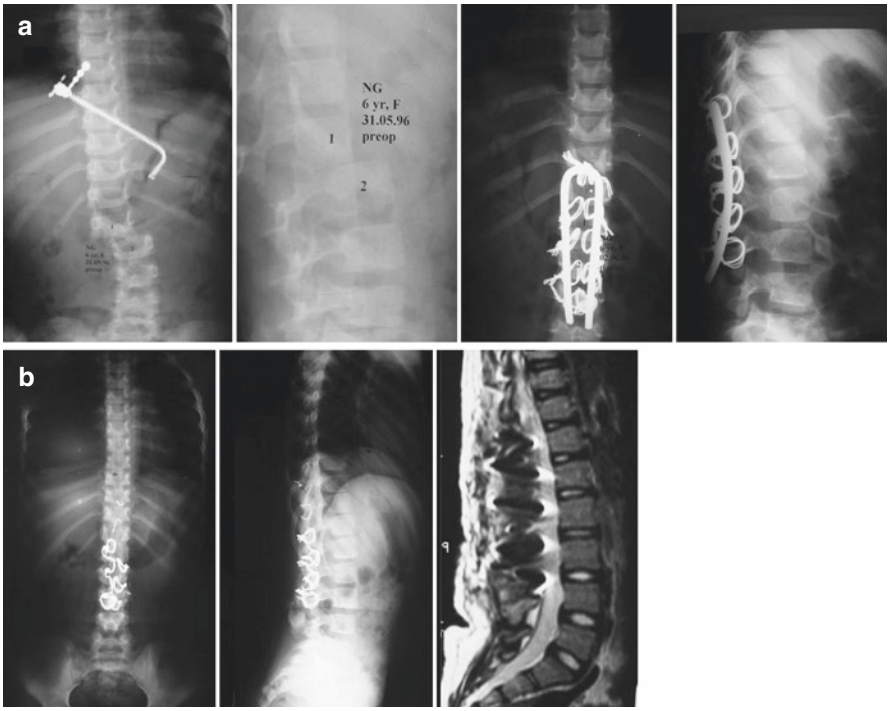

Fig. 2.4 Six-year-old female patient after a vehicle accident. L1–2 fracture-dislocation with an incomplete neurological injury was managed with open reduction and fixation without fusion (**a**). Instruments were removed after 1 year (**b**)

surgical intervention is needed. The neurological status at the initial trauma is the main determinant of the treatment success.

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Atlanto-Axial (C1–C2) Subluxation and Dislocation

3

Federico Canavese

3.1 Definition

Atlanto-axial dislocation (AAD) is the loss of stability between C1 (atlas) and C2 (axis), which can be secondary to traumatic, inflammatory, idiopathic, or congenital abnormalities, although the cause is most commonly multifactorial (Fig. 3.1).

According to the direction and plane of the dislocation, AAD can be divided into four types: anterior-posterior, rotatory (frequent in children), central, and mixed.

3.2 Natural History

If not diagnosed and treated in a timely and appropriate manner, AAD can cause permanent neurologic deficits and sagittal plane deformity.

In particular, sagittal deformity develops when upper cervical spine lordosis decreases and, as a compensatory mechanism, sub-axial cervical lordosis increases. Some patients with end-stage changes (mostly adults) can develop kyphosis at the occipito-axial (C0–C2) segment together with severe sub-axial lordosis (C3–C6), resulting in swan neck deformity.

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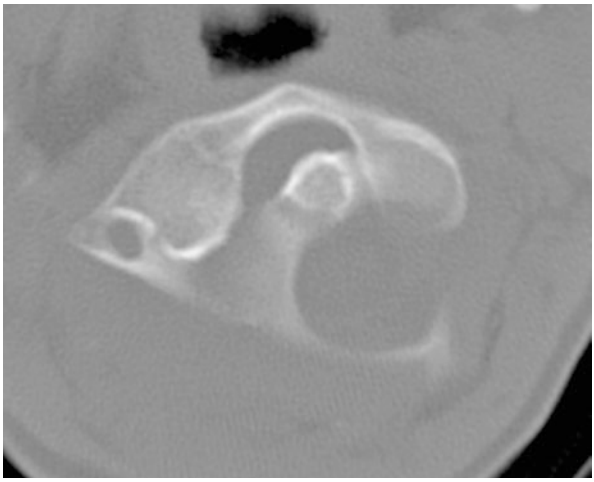

Fig. 3.1 Atlanto-axial dislocation (CT scan; transverse section); patient from GuangzZhou Women and Children Hospital, Guangzhou, China

3.3 Physical Examination

Clinical presentation varies according to the type of injury, the underlying pathology, and the severity of the dislocation. Approximately 50% of patients present with neck pain and/or neck movement restriction, 70% with weakness and/or numbness, and 90% with pyramidal signs (Videos 3.4 and 3.9).

Some patients may also develop muscle weakness, dizziness, tinnitus, blurred vision, sphincter disturbances, lower cranial nerve dysfunction, and respiratory distress.

3.4 Imaging

Lateral flexion-extension (dynamic) and anterior-posterior (AP) open mouth radiographs of the spine should be performed in all patients.

The atlas-dens interval (ADI) is an important radiographic parameter to evaluate the severity and direction of dislocation. ADI is measured from a line projected superiorly along the anterior border to the axis body to the anterior arch of the atlas, and it does not change during flexion-extension movements (normal values: 2–3 mm in adults, and 4–5 mm in children). Increased ADI is suggestive of disruption of transverse ligament of C1.

Three-dimensional computed tomography (CT) scan provides useful information about the osseous anatomy of C1–C2 while magnetic resonance imaging (MRI) helps to assess soft tissues, joints, and the spinal cord. In particular, MRI is indicated when myelopathic symptoms are present or when plain radiographs show

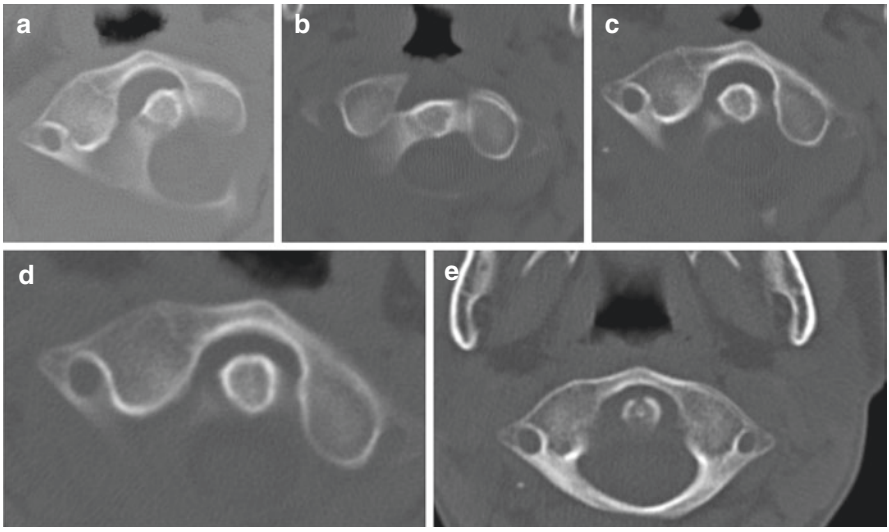

Fig. 3.2 Gradual correction (CT scan; transverse section; (**a**–**e**))

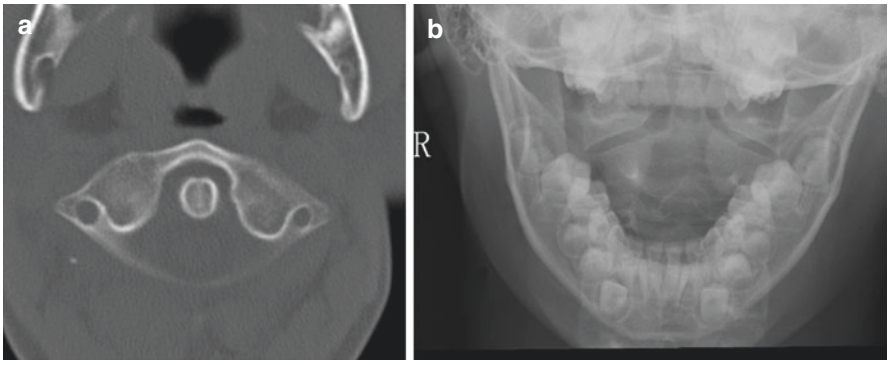

Fig. 3.3 End of treatment ((**a**) CT scan; transverse section; (**b**) open mouth radiograph)

increased ADI and decreased space available for the spinal cord (sub-axial stenosis if the space available for the cord is less than 13 mm) (Figs. 3.2, 3.3, 3.4, and 3.5). It is important to remind here CT scan can have false positives (images suggestive of subluxation but normal anatomical variant); careful imaging assessment is warranted.

In case of AAD secondary to tumor, CT scan and MRI are very important to assess size, range, density and involvement of the tumor, as well as the damage of the body and the pedicle of the vertebra. MR angiography can also be performed to identify the course of the vertebral artery.

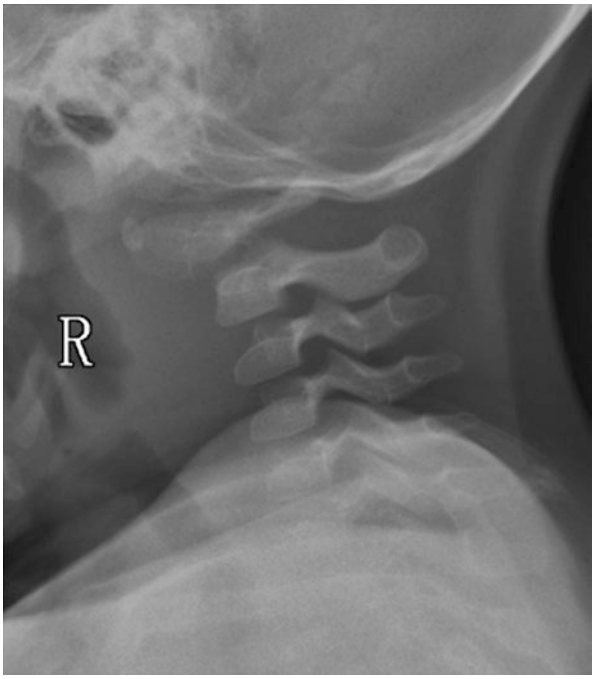

Fig. 3.4 Odontoid (C2) disruption (lateral radiograph)

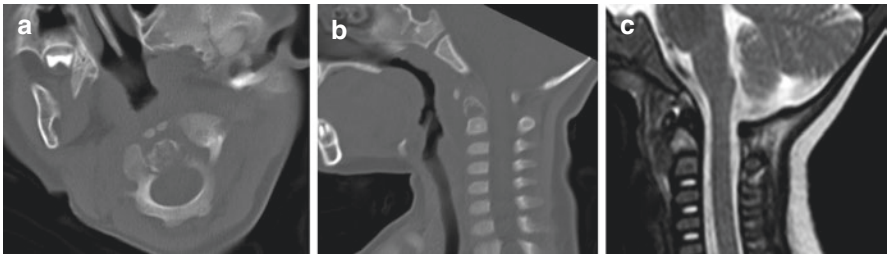

Fig. 3.5 Odontoid (C2) tumor ((**a**, **b**) CT scan; (**c**) MRI)

3.5 Differential Diagnosis

AAD should be considered in a child with the inability or unwillingness to turn the head (evidence of torticollis) when history and physical examination are inconsistent with congenital torticollis (Chap. 28).

The most frequent and relatively benign form of AAD in children without associated fracture is rotatory dislocation; most children with rotatory dislocation have underlying pathologic conditions.

A purely traumatic AAD in the absence of another predisposing risk factor is extremely rare; traumatic osseous injuries may also result in atlanto-axial instability (e.g., type II odontoid fractures).

Table 3.1 Causes and relative incidence of atlanto-axial dislocation

Causes of atlanto-axial dislocation		Incidence
Traumatic	Lesion of the transverse ligament (C2)	Rare
Chromosomal disorder	Trisomy 21	15–20%
	Sandifer syndrome	
Skeletal dysplasias	Goldenhar syndrome	Not known
	Spondyloepiphyseal dysplasia	30%
	Mucopolysaccharides type IV (Morquio syndrome)	40–90%
Congenital osseous abnormalities	Occipitalization of the atlas	Not known
	Congenital abnormality of the odontoid	
Inflammatory disease	Chronic rheumatoid arthritis	Adults 20–86%
	Gastroesophageal reflux (GERD) and chronic oesophagitis	Not known
Tumor	Benign and malignant	Not known
Infection	Retro-pharyngeal abscess (tuberculosis) Grisel's syndrome	Not known

Chromosomal disorders, skeletal dysplasia, congenital osseous abnormalities, inflammatory disorders, and tumors such as aneurysmal bone cyst (Chap. 38), osteochondroma (Chap. 34), chordoma (Chap. 62), osteoblastoma (Chap. 33), fibrous dysplasia, eosinophilic granuloma (Chap. 35), and Ewing's sarcoma; Chap. 39) can be frequently associated with AAD (Table 3.1; Figs. 3.4 and 3.5).

Grisel's syndrome, also known as nasopharyngeal torticollis of inflammatory origin, is a non-traumatic subluxation of the C1–C2 joint, caused by the contracture of the muscles in contact with an infection. Treatment includes antibiotics and immobilization of the neck; early treatment is essential to prevent long-term sequelae (fixed C1–C2 dislocation). Surgical fusion may be required for residual instability of the joint or to correct a rigid deformity.

3.6 Treatment Options

The treatment of AAD varies according to the severity of symptoms and the presence/absence of neurological involvement. Children presenting acutely with AAD can be treated conservatively, in the absence of neurologic injury; orthopedic treatment consists of cervical halter traction in the supine position until AAD is reduced; traction is then followed by orthotic immobilization (rigid brace including the head, neck, and chest, or halo-vest) (Video 3.1) and subsequent active range-of-motion exercises until free-motion returns (Figs. 3.1, 3.2, and 3.3).

Surgical treatment for patients with symptomatic AAD is indicated in children when one or more of the following clinical and/or radiological signs are present: (1) neurologic involvement; (2) ADI greater than 4–5 mm with the reduction of the

space available for the cord; (3) deformity present for more than 3 months; (4) recurrence of deformity following 6 weeks of immobilization.

The goals of surgery are to reduce and stabilize the C1–C2 complex and to decompress the spinal cord. Posterior C1–C2 fusion can be used alone for reducible AAD or in conjunction with anterior trans-oral decompression to treat certain types of irreducible AAD (Videos 3.2 and 3.4).

3.7 Expected Outcomes

In patients treated conservatively, the outcome is variable; resolution of symptoms should be expected in patients treated surgically, although neurological signs may not completely resolve after decompression if treatment is performed too late. Surgical stabilization aims to protect against potential respiratory failure, progressive neurologic symptoms, and death (rare).

3.8 Potential Complications

Serious sequelae include myelopathy, respiratory failure, vertebral artery dissection, neurologic compromise, and rarely quadriplegia or death if left untreated.

3.9 What Should Patient and Family Know?

AAD in children is rarely caused by trauma; congenital abnormalities C1–C2, space-occupying lesions, genetic disorders, and syndromes are significant risk factors for AAD in children.

Children with trisomy 21 are predisposed to it; they should be screened for AAD between 3 and 5 years of age (cervical radiographs) although it is unclear if asymptomatic trisomy 21 patients with an ADI greater than 4 to 5 mm are at higher risk for neurologic sequelae; similarly, patients with Morquio syndrome and Goldenhar syndrome must be advised against sports participation (in particular contact sports) although they may not require surgical treatment.

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Spinal Cord Injury Without Radiographic Abnormality

4

Federico Canavese

4.1 Definition

Spinal **c**ord **i**njury **w**ithout **r**adiographic **a**bnormality (SCIWORA) is a syndrome characterized by clinical symptoms of traumatic myelopathy with no radiographic or computed tomography (CT) scan features of spinal fracture or instability; it is responsible for up to 20% of spinal cord injuries in children (mostly aged less than 10 years of age). SCIWORA is typically seen in the cervical spine although it can also occur at the level of the thoracic and lumbar spine.

4.2 Natural History

The injury of the spinal cord seen in SCIWORA syndrome is caused by a contusion or ischemia of the cord due to temporary occlusion of vertebral arteries, followed by a spontaneous return of vertebrae to their original position. Specific biomechanics of the vertebral column in children allows the musculoskeletal system to move beyond the normal physiological range of motion without the risk of fracture.

Children under the age of 8 years have the most unfavorable prognosis, which is associated with a large head-to-body ratio, increased mobility of the cervical spine, inherent ligamentous laxity, immaturity of neck musculature, incomplete ossification of the vertebrae, and shallow angulation of facet joints during childhood.

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In case of permanent neurological compromise, 100% of patients develop scoliosis (paralytic scoliosis) with or without pelvic obliquity.

4.3 Physical Examination

History of trauma is important as SCIWORA lesions are often caused by hyperextension forces (e.g., during a rear-end car accident) or from a direct frontal impact to the face (e.g., diving, rugby, wrestling, and baseball).

Neurological examination is of paramount importance as patients with SCIWORA can develop a broad spectrum of neurological deficits, from mild to extremely severe symptom; in particular, tetra/para/hemiparesis/plegia, paresthesia, changes in tendon reflexes, loss of bladder and bowel function, signs of anterior/central/posterior cord or Brown-Séquard syndrome in addition to local pain, sensitivity, abrasions, and bruising around the vertebral column. Moreover, neurological deficits can be delayed from a few minutes to 48 h after injury in about 50% of cases. This latency is associated with repeated micro-insults to the spinal cord from striking against the unstable vertebrae. Typically, neurological signs/deficits are more severe in the upper extremities than in the lower extremities (Video 4.4).

The level of spinal cord injury corresponds to the location of the SCIWORA lesions. It is advisable to use the ASIA scale during a clinical examination (Appendix G).

4.4 Imaging

Conventional radiographs are of limited help due to the presence of muscle spasms. In particular, lateral radiographs of the cervical spine alone have low sensitivity and specificity; diagnostic accuracy can be improved if anterior-posterior, lateral, oblique, and open mouth or odontoid radiographs are performed. The stability of the cervical spine can also be assessed by flexion and extension dynamic radiographs. Moreover, the interpretation of cervical radiographs in children can be difficult due to incomplete ossification and the presence of normal anatomical variants (e.g., pseudo-subluxation of C2–C3).

CT is most accurate in detecting bony pathology. SCIWORA should be suspected, and magnetic resonance imaging (MRI) performed, in patients with neurologic symptoms and a positive history of blunt trauma with plain radiographs and CT scans without evidence of fracture.

MRI (sagittal plane) is helpful in identifying the location and extent of the injury as it can detect the signs of acute spinal cord injury (Fig. 4.1), including edema, hematoma, loss of continuity (transection) of the spinal cord, and prolapsed nucleus pulposus. MRI can highlight important prognostic factors of SCIWORA lesions. In particular, hematomas less than 30% of the spinal cord diameter or edema have a favorable prognosis and resolve over time in most cases; on the other hand, transection of the cord or hematomas greater than 50% of the spinal cord diameter have a poor prognosis (Fig. 4.2) and manifest clinically as paresis or paralysis.

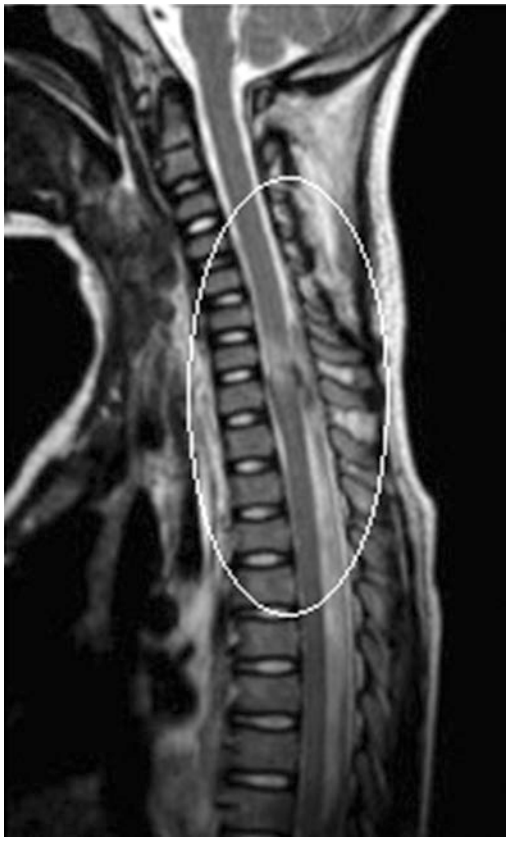

Fig. 4.1 SCIWORA. MRI shows a lesion of the spinal cord at the cervicothoracic junction

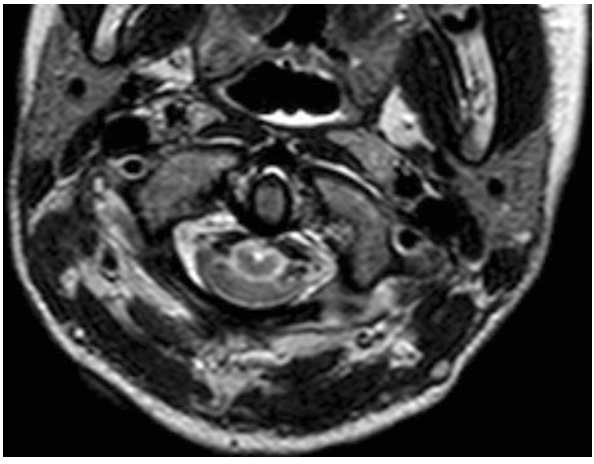

Fig. 4.2 SCIWORA. MRI (transverse section) shows a lesion of the spinal cord at the upper cervical spine

4.5 Differential Diagnosis

Differential diagnosis should include embolism from vertebral artery occlusion associated with cardiovascular diseases such as endocarditis, cardiac arrhythmia, persistent foramen ovale, arteritis, or bleeding disorder. Acute or chronic myelitis should also be excluded (Chap. 1).

4.6 Treatment Options

External immobilization of the spine (brace, collar, or halo vest) for up to 3 months is the mainstay of treatment as most cases show the absence of bony involvement and malalignment (Video 4.1); moreover, the majority of published reports suggest a significant improvement in neurological status without operative treatment. In selected cases with MRI evidence of ligamentous injury, instability, spinal cord compression, worsening, or not-improving neurological findings should be indications for surgical decompression with or without fusion.

For asymptomatic patients who obtained stable spine fixation as assessed by flexion and extension dynamic radiographs, external immobilization devices can be removed earlier. Patients must avoid increased-risk activities for 6 months after diagnosis to prevent acute exacerbations of symptoms and reduce the risk of another injury.

4.7 Expected Outcomes

The two main predictors of prognosis after SCIWORA are the initial neurological status and MRI findings. Improvement of neurological symptoms following conservative treatment should be expected in patients with incomplete neurological injury (absence of osseous lesions and instability); patients with instability or anatomical transection of the spinal cord (complete neurological deficit at initial presentation) have the poorest prognosis with permanent neurologic deficits.

4.8 Potential Complications

Permanent disabilities and possible long-term deformities are among the complications encountered by SCIWORA patients.

4.9 What Should Patient and Family Know?

The upper cervical spine is more commonly affected in younger children while the lower cervical spine is more commonly affected in older children and adolescents as the fulcrum of movement is between C2 and C4 in younger children, and between C5 and C6 in adolescents and adults.

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Part II

Adult Spine Trauma

Upper Cervical Spine Injuries

5

Sander P. J. Muijs and F. Cumhur Öner

5.1 Occipital Condyle and Cranio-Cervical Junction Injury

The cranio-cervical junction is the most mobile part of the spine, formed by the condyles of the occiput and the first two cervical vertebrae; its stability is largely dependent on ligamentous structures. Of all cervical spine injuries, approximately one-third involves the cranio-cervical junction. Due to the great improvement of onsite management (pre-hospital care) of trauma patients, cranio-cervical injuries, which in the past were often fatal on-site, are becoming increasingly common in-hospital trauma care.

5.1.1 Physical Examination

Symptoms may include altered consciousness, high cervical or occipital pain, loss of cervical spine motion, torticollis, and cranial nerve dysfunction (most frequently the 12th cranial nerve). Directly lateral to the condyles, the jugular foramina are located and contain the jugular vein and the 4th, 5th, and 6th cranial nerves, which provide innervation to the throat and to sternocleidomastoid and trapezius muscles. Acute traumatic palsy of cranial nerves rarely shows complete recovery while delayed onset palsy tends to have a more favorable outcome.

In Type B and C injuries neurologic deficit is frequent: only 20% of the patients have no neurological compromise while 38% of patients have quadriparesis or

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quadriplegia, 34% hemiparesis or hemiplegia, and 10% lower cranial nerve palsies (n. hypoglossus, n. vagus) (Videos 5.3 and 5.4).

5.1.2 Imaging

Multiple radiologic measurements have been proposed for the assessment of the atlanto-occipital region (Appendix A). The cranial tip of the odontoid, the caudal tip of the basion (clivus), the midpoint of the posterior arch of C1, the opisthion (the midpoint on the posterior margin of the foramen magnum), and the spinous processes are the commonly used landmarks.

Most commonly used are computed tomography (CT) scan–based measurements such as the Pang's occipital Condyle-C1-Interval (CCI) [1], the Harris' Basion-Axial Interval (BAI) [2] combined with the Wholey's dens-basion interval (DBI) [3], the Powers ratio, and the Sun's C1–C2/C2–C3 interspinous ratio [4, 5].

Magnetic resonance imaging (MRI) should be performed to visualize compression, myelopathy, and hemorrhage and to rule out ligamentous injury in case of any signs of neurologic involvement. MRI is also indicated to rule out SCIWORA (Chap. 4).

Atlanto-occipital dislocation can reduce spontaneously and therefore radiologic measurements as described above may underestimate the injury.

On MRI, extensive ligamentous injury of the cranio-cervical junction can be found even in the presence of a (almost) normal CT scan. Information concerning the mechanism of trauma and the kinetics of the injury is of paramount importance in the workup of all trauma patients with suspected cervical spine injury.

5.2 Occipital Condyle and Cranio-Cervical Junction Injury

5.2.1 Occipital Condyle Fractures (AO Type A)

High-energy blunt compression trauma is the most common trauma mechanism for occipital condyle fractures. The occipital condyles form the lateral border of the foramen magnum.

5.2.1.1 Treatment Options

The majority of condyle fractures are without any signs of ligament injury and hence are calssified as Type A injuries.

5.2.2 Atlanto-Occipital Injury (AO Type B and C)

Atlanto-occipital injuries are often fatal as a result of neurogenic shock and respiratory arrest and are hence rare although nowadays more patients tend to survive and reach the emergency department due to the improvement in pre-hospital (on site).

The normal relation between C0 (occiput) and C1 is (potentially) unstable. Atlanto-occipital dislocation (AO Type C) results from extreme high forces, in which the cranial extension of the posterior longitudinal ligament ruptures due to a combination of axial distraction and extreme hyperextension. The injury is more frequently seen in children after a high-energy trauma (Chap. 1), which results in a combination of axial, translation, and rotational forces. In some cases where there is no dislocation, ligamentous injury can only be detected on MRI (AO Type B injuries).

5.2.2.1 Treatment Options

Atlanto-occipital injuries are highly unstable. In case of manipulation, respiratory arrest and neurologic deterioration can occur. Traction should be avoided at all times.

Operative Treatment

AO Type C injuries require an occipital-cervical fusion (Fig. 5.1) (Videos 5.2 and 5.7). Immobilization in a halo-jacket (Videos 5.1 and 5.2) is a safe option for transport, intubation, and positioning of the patient on the operation table.

Conservative Treatment

Atlanto-occipital dislocations are highly unstable. Halo-vest immobilization for 6–8 weeks (Video 5.1), with a weekly radiologic follow-up, is an option only for AO Type B injuries without displacement.

5.2.2.2 Expected Outcomes

The expected outcomes are largely dependent on neurological symptoms and the presence of concomitant (e.g., vascular) injuries. A C0–C2 fixation will lead to a

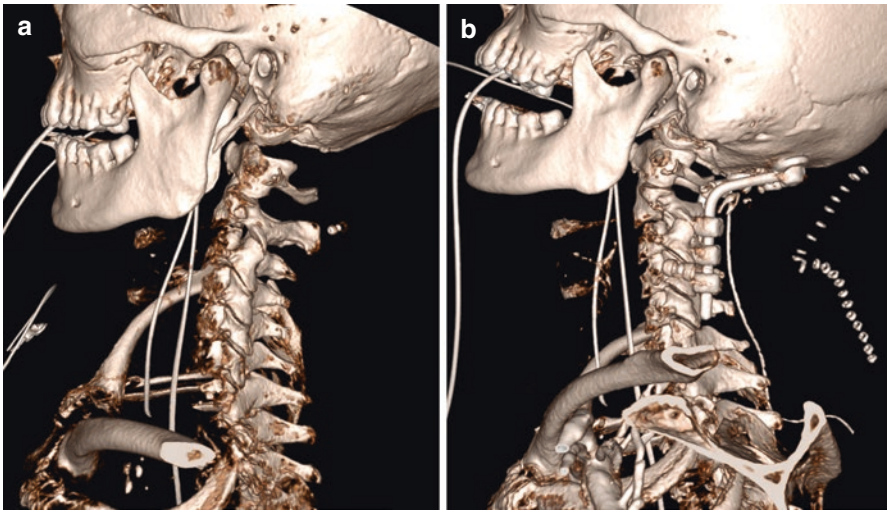

Fig. 5.1 (**a**) 3-D CT reconstruction of an evident C0–C1 dislocation. (**b**) Imaging after reduction in halo and surgical C0–C4 fixation

decrease of flexion-extension of at least 25°, and loss of approximately 50% of cervical rotation.

5.3 C1 Ring and C1–C2 Joint Injuries

5.3.1 Definition

C1 is a closed ring formed by the lateral masses and the anterior and posterior arch. The odontoid process of C2 articulates with C1 and ensures most of the rotation of the cervical spine (all other segments combined together contribute significantly less to the rotational range of motion of the cervical spine).

Fractures of C1 account for 25% of atlanto-axial injuries, and for 10% of all cervical spine injuries. If a C1 fracture is diagnosed, a second fracture of the spine is seen in approximately 50% of the cases (Appendix A).

Preservation of a range of motion of the atlanto-axial segment should be considered when treating atlanto-occipital or C1–C2 injuries.

The isolated C1 lateral mass or posterior arch fracture is in most cases stable (AO Type A). The most classical form is the Jefferson fracture which is a burst fracture of the Atlas and is seen in axial compression trauma. The fracture typically has bilateral fractures at the junction of the lateral masses and the posterior and anterior arches of C1 [6].

The assessment of the integrity of the transverse ligament is essential in the evaluation of isolated C1 fractures [7]. Injury to the transverse ligament makes the C1–C2 complex potentially unstable (AO Type B). Although most patients with isolated C1 fractures do not have any neurological deficit, secondary displacement with neurologic deterioration can occur.

Frank dislocations of the C1–C2 complex (AO Type C) are rare injuries and are commonly seen in children and young patients (Chap. 1).

5.3.2 Treatment Options

All injuries without any sign of transverse ligament disruption are considered to be stable and external immobilization for 6–8 weeks is generally sufficient although there is no consensus on the type of immobilization (halo-jacket vs. collar) due to the lack of good scientific evidence.

In injuries in which a bony avulsion of the transverse ligament is present, a good clinical outcome with non-surgical treatment can be obtained. These injuries can be treated with halo-immobilization for 12 weeks (Video 5.1). In case of MRI evidence of a mid-substance tear of the transverse ligament, the prognosis is less favorable when non-surgical treatment is chosen.

Surgical treatment options in order to fuse C1 and C2 include C1–C2 transarticular screw fixation and the technique described by Harms (C1 lateral mass and the C2 pedicle, pars interarticularis or translaminar screw fixation using polyaxial screws) (Videos 5.2 and 5.7).

5.3.3 Expected Outcomes

Conservative treatment can preserve some of the C1–C2 rotational range of motion although there is a higher risk of long-term pain due to osteoarthritis and/or pseudoarthrosis. If non-surgical management is chosen, and radiologic follow-up shows signs of instability after a period of 8 weeks, surgical treatment should be re-considered.

5.4 C2 and C2–C3 Injuries

C2 fractures represent approximately 20% of all cervical fractures and are common in the elderly population. The most common fracture types (Appendix A) are odontoid fracture (dens fracture) and the *hangman* fractures (traumatic bi-lateral spondylolisthesis of C2). The incidence of neurologic deficit is less than 10% due to the wide diameter of the spinal canal at this level. C2–C3 injuries show a higher incidence of neurologic involvement (up to 25%), especially in (bi-lateral) facet dislocation. Associated injury is seen in the form of paravertebral soft tissue injury, injury of the pharynx, trachea and esophagus, and Horner's syndrome; injury of the vertebral artery is described in fractures passing through the transverse foramen (Chap. 4) [2, 6–8].

5.4.1 Imaging

CT scan imaging is routinely performed when any cervical fracture is suspected. An MRI should be considered in case of a suspected combined injury or ligamentous injury.

5.4.2 Odontoid Fractures

Odontoid fractures are the most common fractures of the axis and are also the most common osteoporotic fractures of the cervical spine. Historically three types are recognized (Fig. 5.2) [8] (Appendix A).

In ***Type I*** odontoid fractures atlanto-occipital dislocation (AO occiput-to-cervical junction fracture Type C) should be ruled out. If there are no signs of an atlanto-occipital dislocation (AO C2 Type A), treatment with a stiff collar for 6 weeks is appropriate.

Type II odontoid fractures are the most common type and have a significantly higher non-union risk [9]; non-union rates for conservative management can be as high as 75% in elderly patients with displacement of more than 6 mm. The symptomatic non-union rate is much lower but still approximately 20%.

There is an ongoing discussion among spine surgeons concerning the need for early surgical fixation of Type II fractures, especially in the elderly. Surgeons supporting early fixation (with anterior odontoid screw) argue that halo treatment has a high morbidity and even mortality in the elderly population. On the other hand,

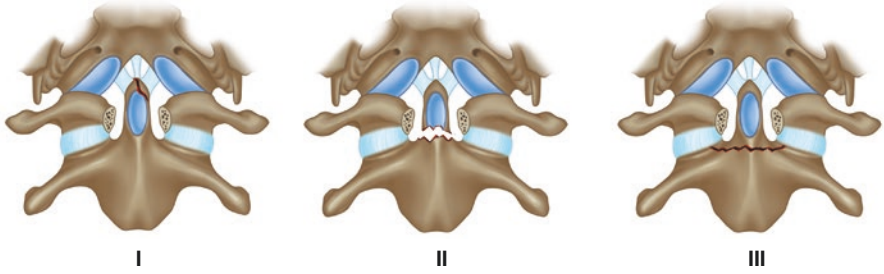

Fig. 5.2 Type I, II, and III odontoid fractures

surgeons supporting treatment by halo-jacket (Video 5.1) argue the bone quality is often sub-optimal, and the risk of non-union and secondary dislocation of the screw is also high. If surgery is chosen, primary C1–C2 fixation is preferable especially in the elderly. C1–C2 fusion for Type II odontoid fractures should be considered in case of secondary dislocation or symptomatic non-union.

Type III odontoid fractures are usually stable and may be treated with cervical immobilization with a union rate above 80%. Although Type II and III odontoid fractures have historically been treated with halo vest immobilization, evidence that cervical collars give a higher non-union rate compared to halo-jackets is lacking [10]. Considering the high rate of complications in the elderly population following surgery or aggressive conservative treatment with a halo-jacket (Video 5.1), and the benign nature of many of the non-union, supportive treatment with a collar and a "benign neglect" policy can also be considered.

Surgical options for odontoid fractures include posterior fusion or anterior odontoid screw in some cases (Video 5.7). In some rare cases, a dislocation through a fractured odontoid may lead to AO Type C injury.

5.5 Traumatic Spondylolisthesis of the Axis (Hangman's Fracture)

The classical *hangman* fracture describes a fracture of the pedicles or pars interarticularis of C2 (Appendix A). The term is partially misleading due to the fact that the most common trauma mechanism leading to traumatic spondylolisthesis of C2 is caused by compression and hyperextension, and not by distraction and hyperextension as occurs in hanging. The most frequent mechanisms of injury are diving in shallow water, motor vehicle accidents, and falls.

5.5.1 Other C2 Fractures

Not all C2 fractures can be classified with the systems described above. Especially more comminuted C2 vertebral body fractures that are neither odontoid nor

hangman type fractures. These fractures (often flexion type injuries) can be difficult to reduce with halo-traction. The extension needed to reduce the fracture anatomically makes it impossible for the patient to see the horizon when fixed in the halo (Video 5.1). In our experience, a horizontal bar between the two posterior-vertical bars of the halo-jacket, with a pad in the neck to stabilize the caudal cervical vertebral column, gives a lever to reduce and maintain reduction. Surgery is reserved for cases with neurologic deficit or painful non-union following halo-treatment.

5.5.2 Combined C1–C2 Injuries

Combined fractures of C1 and C2 are responsible for approximately 4% of all cervical fractures. Fractures of C1 are found in up to 53% of Type II or III odontoid fractures and in up to 26% of hangman fractures. Combined C1–C2 injuries have higher rates of neurologic deficit and death than isolated C1 or C2 fractures; this is probably due to the high-energy trauma mechanism needed for these fractures to occur.

5.5.2.1 Treatment Options

Most hangman fractures are AO Type A or B and can be managed conservatively by 6–12 weeks of cervical immobilization with a rigid cervical collar or halo-jacket (Video 5.1).

Surgery for hangman fractures is necessary in case of nonreducible facet dislocations. When surgery is indicated, options include C1–C3 posterior fusion (Videos 5.2 and 5.7) or C2–C3 anterior cervical fusion.

In the treatment of combined C1–C2 fractures, the type of treatment needed for the C2 injury dictates the surgical option. Most of these combined injuries can be managed, with external immobilization using a collar or halo-jacket. Surgery for combined C1–C2 injuries is reserved for Atlas-Dens Interval >5 mm with signs of mid-substance injury (AO C1 ring and C1–C2 joint Type B and C) of the transverse ligament on MRI (without avulsion fragment), or painful non-union after nonsurgical management. When instrumentation of C1 is impossible due to the fracture type of C1, a C1–C2 transarticular screw fixation or occipital-cervical fusion can be considered (Video 5.7) [11].

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Sub-axial Cervical Spine Injuries

6

Luiz R. Vialle and Emiliano N. Vialle

6.1 Definition

The sub-axial area is the one starting at the third cervical vertebra (C3) and ending in the C7/T1 disc and joints. Any injury occurring from C3 to the C7/T1 disc space is considered to be a sub-axial cervical spine injury.

6.2 Natural History

Sub-axial injuries account for about 3% of all blunt trauma, with an incidence of 64 per 100,000 cases, according to the most recent data. The majority is due to traffic accidents in the younger population while falls represent the main cause in the elderly; diving in shallow water is also one of the main causes of sub-axial cervical spine injury and spinal cord injury (SCI). When associated with SCI, the treatment of such injuries is more demanding and expensive. Unfortunately, some cases are underdiagnosed and may lead to secondary deformity and permanent neural damage; it is of utmost importance to manage all injuries during the index surgical procedure.

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6.3 Physical Examination

The simple inspection tells the examiner how severe the cervical injury could be; an ambulatory patient obviously differs from a tetraplegic one. The physical examination starts with palpation of the neck; the examiner must look for edema, tenderness, crepitation, or hematoma. Even minor pain with neck movements must be an alert to the examiner. Neurological evaluation (Video 6.4) should be performed in all cases followed by established protocols if a SCI is present. It is imperative to annotate all clinical and neurological findings as changes frequently occur during the first posttraumatic period (hours).

6.4 Imaging

Not all cervical spine trauma cases should be screened by all sorts of imaging. There are two well-established protocols: the Canadian C-spine rule [1] and the North American National Emergency X-Radiography Utilization Study Low-Risk (NEXUS) Criteria. The C-spine rule is slightly more precise, as it evaluates local pain or movement limitations during the examination; patients without major clinical/neurological signs do not need to undergo imaging assessment. This measure can save time and it is economically sound.

However, any major trauma or symptomatic case should be sent to radiology. Standard radiographs—anterior-posterior (AP) and lateral, flexion-extension projections—may not be always trustful. Most of the time, the presence of muscle spasms or pain can hide minimal dislocations. A flexion-extension radiograph is not recommended in the emergency room (ER); an unstable spine may put the patient at risk for spinal cord compression or spine dislocation. Similarly, the cervicothoracic junction may be difficult to assess on plain radiographs (due to tissue superposition) in obese patients, patients with large shoulders, and obtunded patients (Fig. 6.1a). In particular, most subtle injuries of the cervicothoracic junction are often misdiagnosed. In order to avoid missing a sub-axial cervical spine injury, the computed tomography (CT) scan is the imaging of choice (Fig. 6.1b, c); it is fast reliable, and highly sensitive, and it can provide three-dimensional reconstruction.

In the case of a negative CT scan, or if any doubt, magnetic resonance imaging (MRI) should be performed to evaluate ligaments, facets, discs, and spinal cord. However, in the ER setting, the first-line imaging studies are radiographs and CT scans while MRI has a limited place. In the presence of pain without radiological abnormalities, a second radiograph should be requested within 2 weeks from trauma, once muscle spasms have resolved; in this case, instability may be detected (Fig. 6.2).

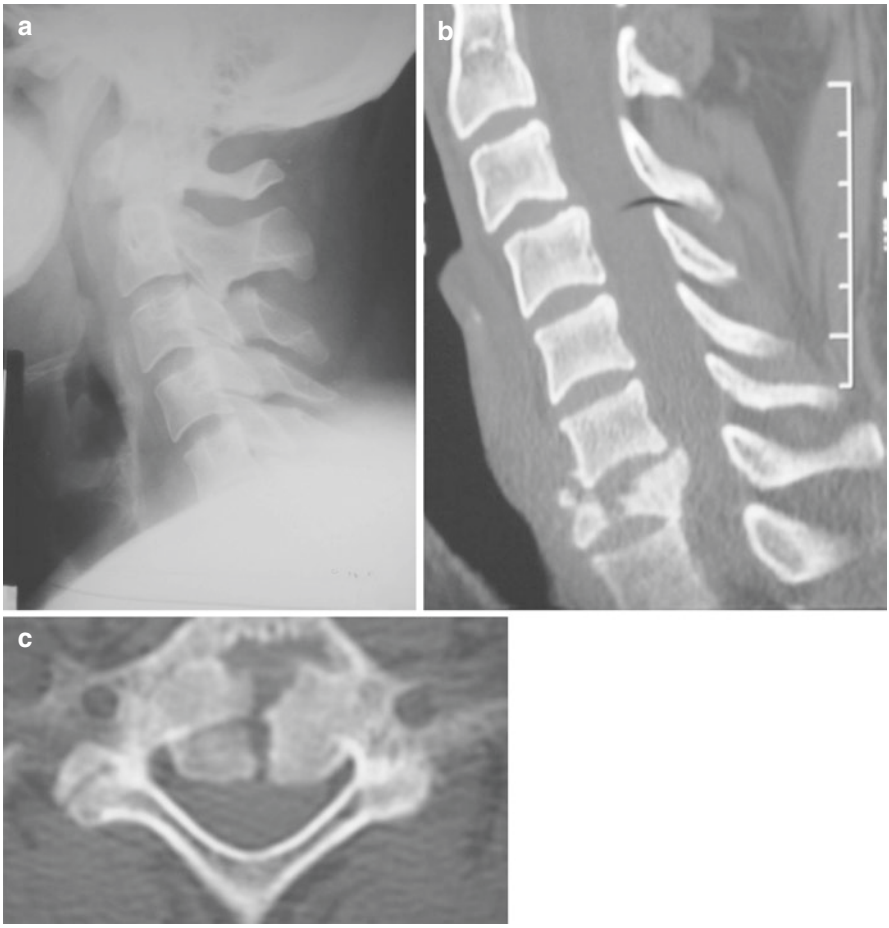

Fig. 6.1 (**a**) Shoulders masking the sub-axial cervical spine (lower cervical spine); (**b**, **c**) CT scans of the same patient

6.5 Classification

Over the years, several classifications have attempted to characterize cervical injuries according to force vectors, mechanisms of injury, and type of deformity. Recently, the AO Spine Cervical Spine Injury Classification System was published and it is the only validated one, so far [2, 3]. The neurological assessment in the ER and the modifiers included in this system are useful tools to evaluate the injury and to drive the decision-making process. The main innovation of this system is the independent analysis of the facet injury; it helps the surgeon to correctly understand the lesion and its treatment (Appendix B).

6.6 Differential Diagnosis

The objective is to evaluate, depending on the classification system, the severity of the injury, and its potential instability. Ligaments are not apparent on plain radiographs and a simple Type A1 fracture may hide a more complex unstable Type B. The modifiers suggested by the classification system highlight this possibility. Palpation with pain or tenderness is a "red flag," and further investigation is warranted (radiographs, CT scans, and eventually MRI) (Fig. 6.3). The ligamentous injury needs to be identified in order to avoid complications due to dislocations or later deformities due to misdiagnosed injuries.

6.7 Treatment Options

As a rule of thumb, stable injuries can be managed conservatively, with a rigid collar. On the other hand, unstable injuries as well as those with neurological deficits should be treated surgically (Videos 6.1, 6.2, 6.4, 6.7 and 6.9). The anterior approach is our preferred option, especially in dislocations; it is a relatively rapid surgery that allows an effective decompression by removal of the disc/bone fragments and also allows a stable plate-screw fixation. Some cases, with gross instability or with the involvement of the cervicothoracic junction, may need a circumferential approach (Videos 6.2, 6.3 and 6.7).

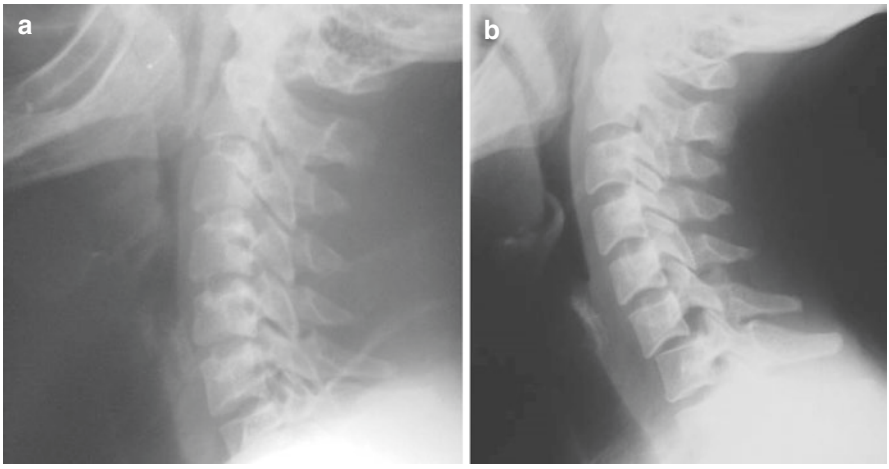

Fig. 6.2 (**a**) Direct lateral radiogram taken in the emergency room shows normal findings; (**b**) C6–C7 injury seen in control X-ray taken at 2 weeks follow-up

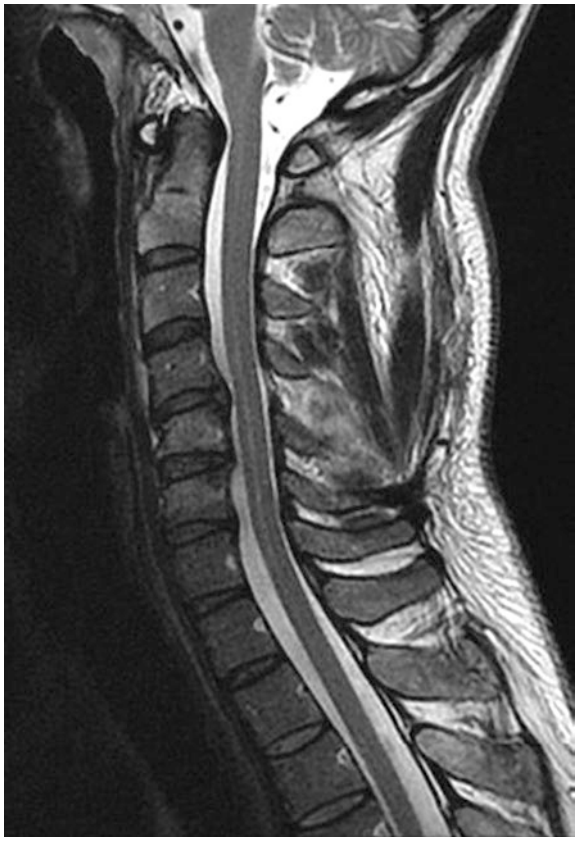

Fig. 6.3 The C5 injury was classified as AO Type A1; further investigation showed C4–C5 interspinous process opening and signs of ligamentous-injury (AO Type B2 lesion)

6.8 Expected Outcomes

Without the presence of neurological damage, the recovery is usually fast. Injuries adequately classified and treated, with a well-performed surgery, will heal in around 3 months. Adjacent segment disc degeneration must also be considered in the long term.

6.9 Potential Complications

Other than the neurological deficit, complications may include pseudoarthrosis, surgical site infection, and postoperative deformity. The management of complications is usually challenging for the surgeon.

6.10 What Should Patient and Family Know?

Patients and family need to be warned about potential complications and follow-up during the postoperative period. In particular, they should understand the importance of limiting in activities of daily living, the time needed to heal; obviously, neurological involvement demands a multidisciplinary approach.

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Thoracolumbar Injuries

7

Alpaslan Şenköylü

7.1 Definition

About half of spine fractures are seen at the thoracolumbar (TL) junction (T10–L2). The TL junction is a transitional area between the flexible lumbar spine and relatively less mobile thoracic spine due to coronally oriented facet joints, rib cage, and thin intervertebral discs. Therefore, this area is subjected to significant biomechanical stress. Most of TL injuries occur as a result of high-energy blunt trauma (traffic accidents or falling from height) in individuals with normal bone quality.

7.2 Natural History

Natural history varies according to the severity of the injury. Many classification systems have been developed to predict both the natural history and the management options of such fractures. Of these, the AO Thoracolumbar Classification System has shown to be reliable and reproducible (Appendix C). The prognosis worsen from Type-A to Type-C fractures. In particular, Type-C injuries are associated with neurological compromise in up to 50% of cases.

TL burst fractures, including AO Type-A3 (stable) and Type-A4 (unstable) injuries without neurological deficit, have different prognosis (Appendix C). In particular, AO Type-A3 fractures are biomechanically stable and rarely require surgery while AO Type-A4 fractures commonly need operative treatment due to the risk of

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progressive kyphosis (unstable fracture pattern) (Chap. 51). The AO Type-A2 injuries ("pincer" fractures) are at risk of pseudarthrosis as the coronally oriented split of the vertebral body can be associated with leakage of disc material between bony fragments that prevents bone from healing.

7.3 Physical Examination

After prioritizing the life-threatening injuries, a careful history of trauma should be obtained. A palpatory screen is also needed to gauge the location of pain and to rule out concomitant injuries of the spine (not uncommon).

Patients must be positioned in lateral decubitus to palpate the spine from top down. The back of the patient should be inspected meticulously for possible bruises, tenderness, and a palpable gap between spinous processes that is a sign for a posterior ligamentous complex injury.

A thorough neurological examination of the extremities is essential (Video 7.4). Since the spinal cord ends at L1–L2 level, TL junction injuries can cause spinal cord, conus medullaris, and/or lumbar nerve root lesions.

Spinal cord injury with upper motor neuron signs should be classified according to the American Spinal Cord Injury Association (ASIA) for prediction of prognosis (Appendix G).

Conus lesions occur with sphincter dysfunction with loss of perianal sensation and preserved lower limb motor function.

Nerve root lesions are seen with lower motor neuron signs including flask paralysis of related myotome, decreased deep tendon reflexes, and absent pathologic reflexes (Video 7.4).

7.4 Imaging

Standard radiographic examination consists of anterior-posterior (AP) and lateral radiographs of the affected area. AP view may demonstrate rotation and widened inter-pedicular and inter-spinous interval whereas lateral view may show wedging of vertebral body, local kyphosis, translation, and widened inter-spinous distance (Fig. 7.1a).

A computed tomography scan provides good-quality images of calcified tissues and is the best imaging modality to characterize a bony lesion previously observed on plain radiographs. Two-dimensional and three-dimensional reformatted images provide even better elaboration for the classification of injury (Fig. 7.1b).

In patients with associated spinal cord or nerve root injury, magnetic resonance imaging (MRI) is indicated to assess the posterior ligamentous complex; MRI is also indicated to identify noncontiguous injuries by screening the whole spine.

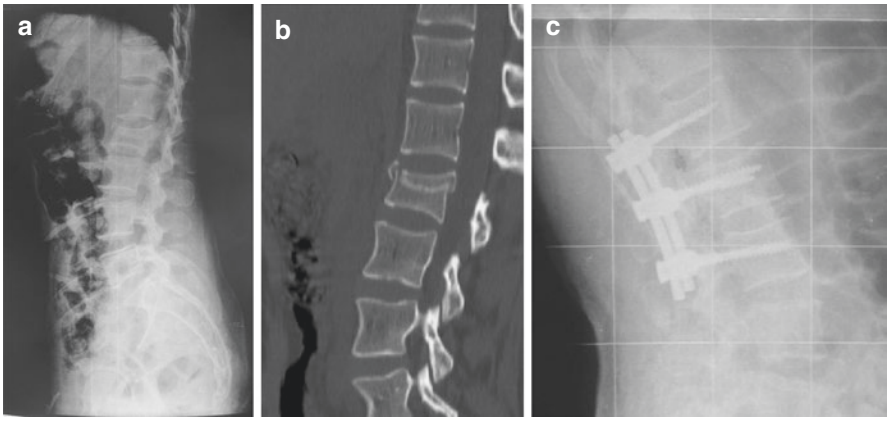

Fig. 7.1 A 32-year-old female admitted to the hospital after traffic accident. Her direct lateral X-ray examination (**a**) revealed a wedging because of collapse of upper endplate at L2 level. Sagittal 2D reformatted CT scan images (**b**) showed an AO Type-A3 (incomplete burst) fracture at L2 vertebra. After discussing treatment options with the patient, posterior short segment instrumentation procedure was decided for early ambulation (**c**)

7.5 Differential Diagnosis

Pathologic and osteoporotic fractures should be excluded in patients with negative history for trauma and in those with a history of low-energy trauma (fall from a standing height).

MRI is helpful for the differential diagnosis of malignant lesions. Whole vertebral body involvement, convex posterior wall, endplate erosions, multiple lesions, soft tissue, and/or involvement of posterior elements are suggestive of pathologic fracture due to malignancy.

7.6 Treatment Options

Management of spinal trauma includes transportation of the patient, care in the emergency room, treatment of the fracture, and the rehabilitation period.

Careful transportation from the site of the accident to the hospital is important to prevent secondary injuries to the spinal cord. Upon arrival at the emergency room, the general condition of the patient and the presence of concomitant injuries must be assessed. If the general condition is stable, treatment of spine injury can be performed.

If the injury is stable, conservative treatment is indicated. The concept of stability is defined by Panjabi and White as follows: “Maintain the spinal alignment under the physiological loading without neurologic deficit and pain.” Fortunately, most of the TL fractures are stable and can be treated conservatively with a rigid brace such as thoraco-lumbo-sacral orthosis (TLSO). Simple compression (AO Type-A1) and

stable burst fractures (AO Type-A3) can be treated by brace for 8 weeks (brace can be removed at night).

AP and lateral radiographs are useful to assess consolidation and during follow-up. Early rehabilitation is important for preventing complications.

Generally, AO Type A4, Type-B and Type-C injuries are unstable and surgical treatment can be needed. Presence of neurologic deficit is considered as clinically unstable and should be treated surgically. Urgent surgical treatment including decompression and stabilization increases the chance of recovery of the patients with neurological deficit. Pedicle screw fixation (Video 7.3) is effective to provide biomechanical stability; short versus long fixation and anterior versus posterior instrumentation options should be decided upon the type of fracture, the number of affected levels, the presence of comorbidities, and the neurological status (Fig. 7.1c). Decompression is only indicated if the patient has a neurological deficit due to canal compromise. There is no need to use a rigid brace postoperatively if stable fixation can be obtained (Type-A and Type-B fractures). Early postoperative rehabilitation is important.

7.7 Expected Outcomes

Pseudoarthrosis is unlikely to occur since vertebrae have a good healing potential.

The expected outcome of nerve root injury is relatively better compared to spinal cord injury, which is discussed particularly in Chap. 9.

7.8 Potential Complications

Post-traumatic kyphosis can be related to the misdiagnosis of the type injury or to neglected injury in case of polytrauma (Chap. 51). Other complications are generally related to the surgical approach and to the instrumentation.

7.9 What Should Patient and Family Know?

TL fracture should be considered as a major injury which may need a multidisciplinary treatment (surgery and/or rehabilitation). Return to normal activities of daily living may take more than 3 months.

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Sacral Injuries

8

Luiz R. Vialle and Emiliano N. Vialle

8.1 Definition

Sacral injuries do affect the sacrum and its relationships with the pelvic girdle. As a result, sacral injuries can be isolated or in association with fractures of the pelvis and/or the sacroiliac joint.

8.2 Natural History

Sacral injuries are generally due to high-energy trauma and are frequently associated with injury of other organs and are characterized by hemodynamic instability. The majority of sacral injuries are a consequence of traffic road accidents although other causes such as fall from height are also possible. The concomitant occurrence of other pelvic and long bone fractures is not uncommon. Neuro-urological or anorectal sequelae are frequent.

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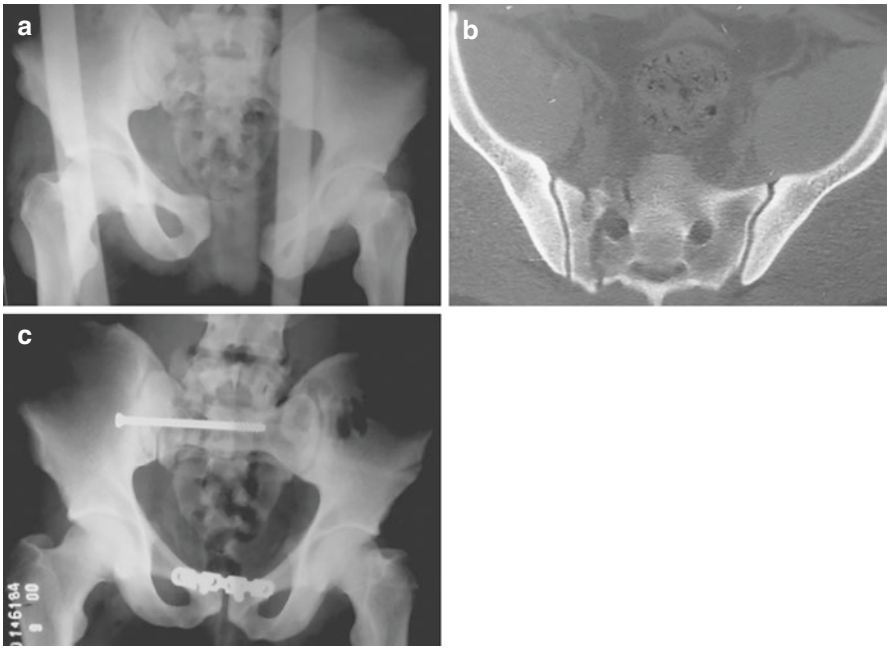

Fig. 8.1 (**a**) "open book" lesion; (**b**) a Type B3 pattern; (**c**) surgical treatment of the same patient

8.3 Physical Examination

As sacral injuries are associated to high-energy trauma, all the ATLS principles must be applied. Bleeding can occur when the pelvic ring is disrupted ("open book" injury, in particular), so special care must be taken to avoid hemodynamic failure (Fig. 8.1a–c). Once abdominal and urogenital evaluations are cleared, and the bleeding has been controlled, the examiner can proceed with the clinical examination. Essentially, a neurological examination is performed to identify any loss of motor or sensory function. Territories from L5 to S2 should be evaluated with as much care as possible (Video 8.4).

Pathological fractures due to osteoporosis or tumor lesions are also described in the elderly population. In particular, it is important to identify osteoporotic fractures (frequently caused by a fall).

8.4 Imaging

After the initial pelvic trauma radiographic protocol—anterior-posterior (AP), inlet, and outlet views—the next imaging study, or the first one in some emergency room departments, is the computerized axial tomography (CAT) scan. In polytraumatized

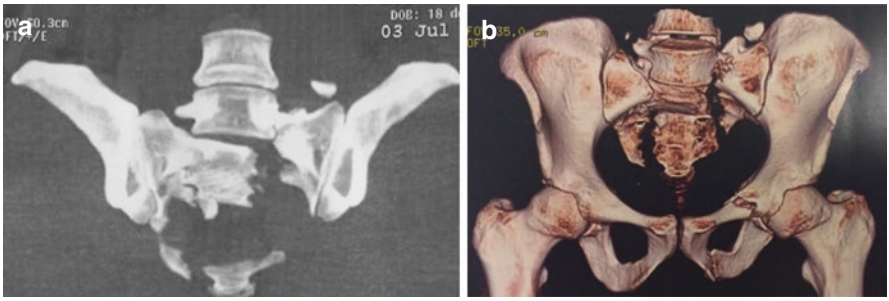

Fig. 8.2 Type C3 injury; (**a**) CAT scan (**b**) CT 3D reconstruction

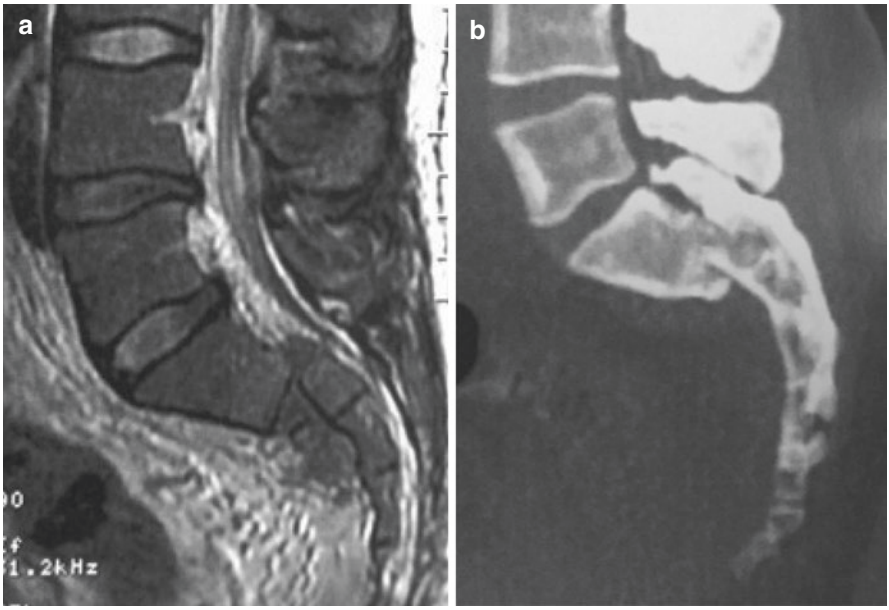

Fig. 8.3 Type C3 injury; the information was given by MRI (**a**) is less precise than the one provided by the CT scan (**b**) which should guide the treatment algorithm

patients, delayed diagnosis of a sacral injury is common. The routine CAT scan clearly identifies minor and major injuries, allowing the application of a classification system that can help the surgeon with the decision-making process (Fig. 8.2). Magnetic resonance imaging (MRI) is rarely needed upon the admission of the patient in the ER although it may be helpful to evaluate nerve roots or in the case of pathological fractures (Fig. 8.3).

8.5 Classification

Traditionally, sacral fractures were included in the pelvic trauma classifications, due to their frequent association. Few classifications started to prioritize the sacrum, although none were inclusive nor validated. The AOSpine sacral classification offers a definitive tool for the correct understanding of the displacements resulting from the injury [1]. The analysis of the morphological alterations defines the degree of severity of the deformity and the associated amount of instability. A neurological evaluation at admission and the application of modifiers help with the treatment algorithm. A detailed description can be found online [2] Appendix D highlights this classification. The correct interpretation of the injury allows the surgeon to differentiate between stable and unstable injury, to discard or confirm the spinopelvic involvement, and to suggest the best treatment option.

8.6 Differential Diagnosis

There is not a significant differential diagnosis to keep in mind although an adequate assessment of the amount of instability is of paramount importance. The analysis of high-quality images and a correct application of the classification are needed to rule out complex spinopelvic dissociation. Not displaced Type C0 fractures are potentially unstable, and without proper identification, a secondary displacement can occur. The small signs of a more severe injury must always be on the surgeon's mind, as the minor L5 transverse process avulsion may be a warning sign for vertical instability.

8.7 Treatment Options

1. Conservative treatment: This is an option applicable to all stable injuries, without joint disruption or pelvic ring dissociation. It consists of bed rest and progressive mobilization without full weight-bearing. The patients must be regularly followed with weekly radiographs so that any misdiagnosis or minor displacement could be promptly identified. The treatment option should then be reevaluated.
2. Surgical treatment: This option is indicated when there is uncontrolled pain, instability, or gross displacement and fixation or reduction is deemed necessary [3]. As a rule of thumb, it is important to reduce and fix the sacrum first and then to extend the instrumentation if needed (Video 8.3). The options are:
   (a) Cement injection for pathological fractures is effective to control pain.
   (b) Percutaneous iliosacral screws for trans-alar fractures, mainly Type B injuries (Fig. 8.1c).
   (c) Spinopelvic fixation for unstable injuries (Type C; Fig. 8.4).

According to the clinical status, the anatomy of the injury, and the amount of displacement, the goal of treatment is to reduce and stabilize the injury with a sound fixation; not infrequently, this means spinopelvic instrumentation from L4 to the iliac ala (uni or bilateral) (Fig. 8.4c, d).

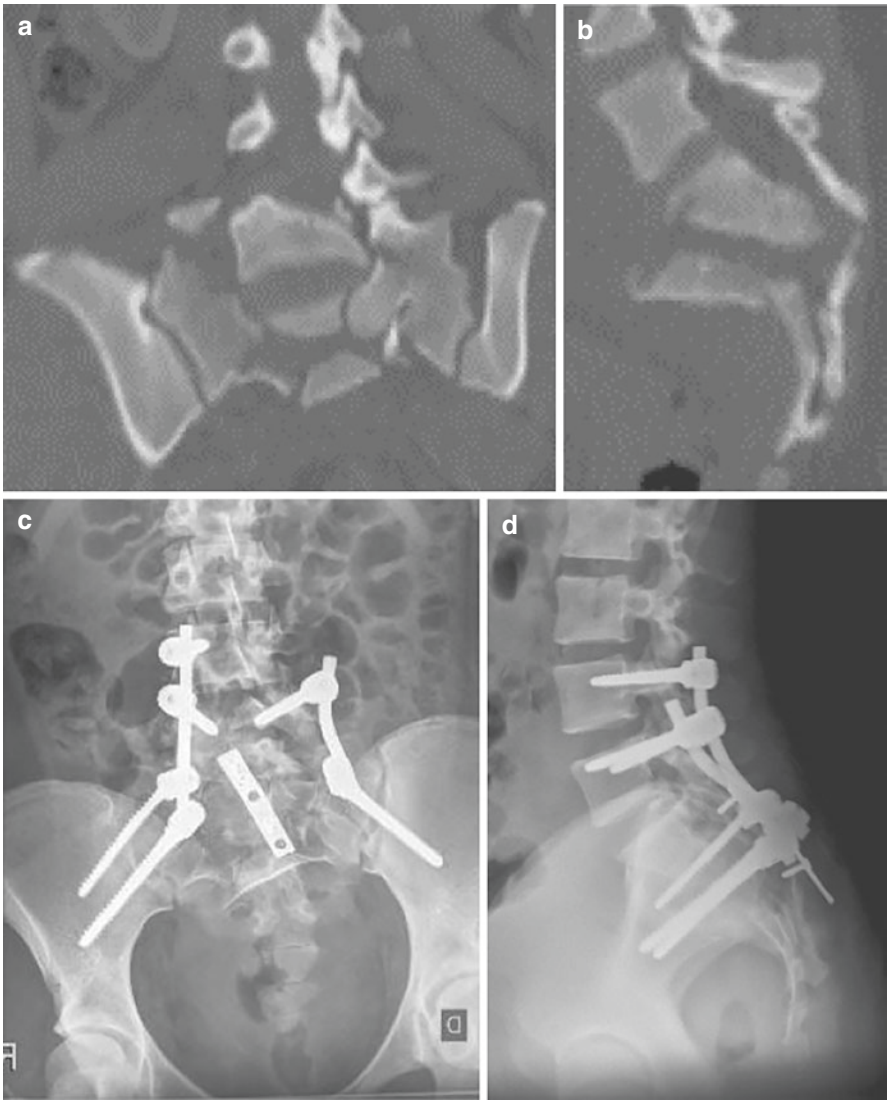

Fig. 8.4 Type C3 injury; (**a**, **b**) CT scan showing displacement and instability; (**c**, **d**) surgical treatment with spinopelvic bilateral fixation and additional posterior plate

8.8 Expected Outcomes

Outcomes depend on the severity of the injury. In Type A injuries, the return to normal daily life activities is possible in about 2 months while complete recovery from a severe sacral injury implies a long-term rehabilitation program.

8.9 Potential Complications

Patients must be aware of the potential complications such as neural deficits, residual deformity, lower limb discrepancy, pseudoarthrosis, and postoperative infection (not uncommon; perineum close to the surgical approach). The initial treatment must consider all these issues, and the surgeon must use all resources to achieve the best possible reduction and fixation.

8.10 What Should Patient and Family Know?

The consequences of such injury must be stressed to patients and families before starting any treatment. It may be a life-threatening situation in the ER, a demanding procedure at the operating room with numerous potential postoperative complications and risks; all potential complications should be carefully explained and detailed. Last but not the least, patients and family should be prepared for a long-term rehabilitation process.

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Spinal Cord Injury and Related Conditions

9

Paula Valerie ter Wengel and F. Cumhur Öner

9.1 Definition

Traumatic spinal cord injury (tSCI) is defined as neurologic dysfunction as a consequence of spinal trauma. Its incidence ranges between 10 and 84 cases per million per year, with wide variations between countries. The cervical spinal cord is the most commonly affected level, followed by the thoracic and lumbar spinal cord. The severity of the injury is generally categorized according to the American Spine Injury Association (ASIA; Appendix G) Impairment Scale (AIS: AIS A-E). Around 30% to 55% of patients with tSCI will present with complete neurological injury (AIS A).

9.2 Natural History

The severity of initial neurological injury negatively affects the neurological outcome, where patients with complete tSCI AIS A and incomplete AIS D are less likely to recover neurologically compared to incomplete AIS B and C patients. One year after trauma, approximately 70% to 85% of AIS A patients and 85–88% of AIS D patients will not recover in AIS grade. In contrast to AIS B and C patients, patients with less severe incomplete tSCI (AIS D) are also less likely to recover

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neurologically. Injuries at the cauda equina level, in general, have a better potential for substantial recovery.

9.3 Physical Examination

When feasible, patients should be evaluated neurologically according to the International Standards for Neurological Classification of Spinal Cord Injury (ISNCSCI). This systematic neurological examination is used to determine the motor, sensory impairment, severity, and level of tSCI (Video 9.4). The severity of the neurological injury is classified in AIS grades, where AIS A is sensorimotor complete tSCI and AIS B-D refer to incomplete tSCI. AIS B describes motor complete lesions with preserved sensation below the level of injury, which at least should include the lower sacral segments. AIS C describes motor incomplete lesions ranging from only the presence of voluntary anal contraction to preservation of some motor function below the level of injury, where at least half of the key muscles have less than antigravity function (<MRC 3). AIS D refers to incomplete motor lesions where at least half of the key muscles have antigravity function (≥MRC 3). When there is a disproportionately greater motor deficit in the upper extremities compared to the lower, this is defined as a central cord–type injury (TCCI). While TCCI is the most frequent incomplete tSCI, there are still inconsistencies in the definition and treatment of TCCI patients. Some argue that TCCI patients have a more favorable prognosis compared to incomplete tSCI. However, the initial severity of the injury, as categorized by the AIS grade, plays a more important role in neurological recovery potential rather than having a TCCI-like injury.

9.4 Radiographic Examination

Imaging should start with a high-quality CT scan with multiplanar reconstructions. In cases with tSCI, an MRI should be obtained even if there are no injuries seen on CT scans. MRI should be preferentially performed preoperatively to adequately assess the extent of spinal cord compression, hemorrhage, traumatic disc herniation, epidural hematoma, or ligamentous injury.

9.5 Differential Diagnosis

Complete spinal cord injury can be difficult to distinguish initially from spinal shock. Spinal shock is characterized by the temporary reduction or loss of reflexes, motor, and sensory function following tSCI and can last from hours to weeks. Spinal shock is more pronounced in severe spinal cord injury and at higher neurological levels of injury. There is debate regarding the end of the spinal shock phase; usually,

it starts with the gradual return of reflexes. While it can be difficult to distinguish in the acute phase, only about 3% of patients with complete cervical tSCI will show some spontaneous neurological recovery within a week indicating recovering spinal shock.

9.6 Neurogenic Shock

Injuries above T6 can cause sudden loss of autonomic tone. This can cause a life-threatening distributive shock with hypotension, bradycardia, and peripheral vasodilatation. In the acute phase, hypovolemic shock should be ruled out. In contrast to hypovolemic shock, patients with neurogenic shock will have bradycardia due to the loss of sympathetic innervation instead of tachycardia.

9.7 Treatment Options

Surgery aims at restoring the spinal alignment and stability while decompressing the injured spinal cord; early treatment increases the chances of recovery. Life-threatening injuries should be managed first. When there is ongoing spinal cord compression, it should be decompressed to prevent further damage to the spinal cord and enhance potential neurological recovery. Dislocation injuries can be treated by promptly closed reduction, while other compressive injuries should be treated operatively. There is debate on the optimal surgical timing. While surgical decompression within 24 h from trauma has been shown to increase the chances of neurological recovery and to prevent secondary deterioration, the effect of earlier timeframes (less than 8 to 12 h) is still under investigation though early treatment increases the chances of recovery. The surgical plan depends on the type of injury and the compression of the spinal cord. Nonetheless, laminectomy appears to have a greater potential to adequately decompress the spinal cord than anterior surgery only.

9.8 Expected Outcome

The neurological outcome is dependent on the level and severity of the injury as well as the surgical timing. When surgical decompression is performed within 24 h, patients with complete cervical tSCI (AIS A) appear to have a significantly greater likelihood to improve ≥2 AIS grades compared to surgery performed thereafter, namely, 22.6% versus 10.4%. About 27.2% of patients with cervical tSCI (AIS A-D) will recover ≥2 AIS grades or recover to normal when surgery is performed within 24 h, compared to 26.5% when surgery is performed later. For thoracic and thoracolumbar tSCI, this is 42% when surgery is performed within 24 h, compared to 27.3% when it is performed later (Tables 9.1, 9.2, 9.3, and 9.4).

Table 9.1 The impact of surgical timing on >2 ASIA grade improvement in the complete cervical tSCI

first author of study	total nr of patients	nr of patients with improvement	percentage improvement	lower 95%CI	higher 95%CI
Fehlings [30]	44	8	21.7	14.2	29.3
Levi [34]	22	5	22.7	14.9	31.8
Umerani [41]	12	3	23.0	14.3	34.3
Newton [37]	24	7	23.7	16.0	34.8
Randle [39]	20	3	21.7	12.7	30.0
Bourassa–Moreau [28]	14	4	23.4	15.1	35.5
Papadopoulos [38]	38	8	22.4	15.2	30.5
Jug [33]	26	4	21.5	12.8	29.4
Mattiassich [36]	20	5	23.0	15.0	33.0
Hansebout [32]	14	4	23.3	15.1	34.2
Grassner [31]	14	5	24.2	16.0	37.4
Early surgery	**248**	**56**	**22.6**	**16.6**	**28.7**
Fehlings [30]	27	3	10.6	5.3	17.1
Levi [34]	14	4	11.9	5.9	21.8
Umerani [41]	20	2	10.5	5.2	17.1
Randle [39]	12	1	10.5	5.1	17.2
Liu [35]	66	8	10.8	5.7	16.3
Benzel [23]	35	0	9.2	3.5	15.1
Late surgery	**174**	**18**	**10.4**	**5.6**	**15.8**

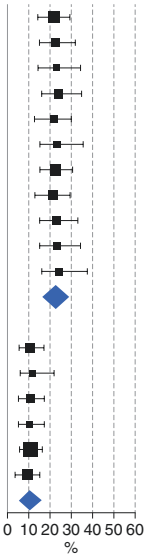


Table 9.2 The impact of surgical timing on >2 ASIA grade improvement in the incomplete cervical tSCI

first author of study	total nr of patients	nr of patients with improvement	percentage improvement	lower 95%CI	higher 95%CI
Fehlings [30]	87	30	33.4	25.1	43.2
Umerani [41]	19	8	35.6	22.1	53.9
Newton [37]	17	6	32.5	18.8	49.6
Papadopoulos [38]	28	6	26.4	13.6	38.3
Jug [33]	16	8	38.3	23.6	59.5
Mattiassich [36]	29	9	30.7	18.9	44.2
Grassner [31]	21	1	20.3	5.7	35.2
Early surgery	**217**	**68**	**30.4**	**19.8**	**41.6**
Fehlings [30]	64	17	28.8	19.0	38.2
Umerani [41]	41	8	25.3	13.8	36.4
Randle [39]	12	6	38.6	22.9	59.7
Liu [35]	251	86	34.0	28.6	39.6
Benzel [23]	51	21	37.9	27.4	50.8
Late surgery	**419**	**138**	**32.5**	**21.4**	**45.8**

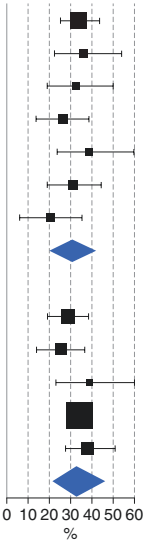


Table 9.3 The impact of surgical timing on >1 ASIA grade improvement in the complete thoracic/thoracolumbar tSCI

first author of study	total nr of patients	nr of patients with improvement	percentage improvement	lower 95%CI	higher 95%CI
Cengiz [23]	6	4	58.4	25.1	89.4
Bourassa [5]	24	4	19.5	7.0	36.1
Rahimi [22]	7	1	22.1	3.3	51.8
Payer [26]	6	5	69.3	34.6	95.4
Dobran [32]	16	6	37.6	17.6	60.1
Early surgery	**59**	**20**	**40.0**	**11.2**	**79.4**
Cengiz [23]	7	1	18.7	2.3	47.2
Bourassa [5]	9	2	23.6	5.4	49.7
Rahimi [22]	9	1	16.5	2.1	40.7
Rahimi [33]	11	8	63.5	34.7	87.8
Rahimi [29]	10	2	21.6	4.7	46.7
Late surgery	**46**	**14**	**27.4**	**4.8**	**61.9**

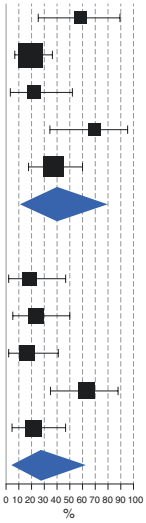
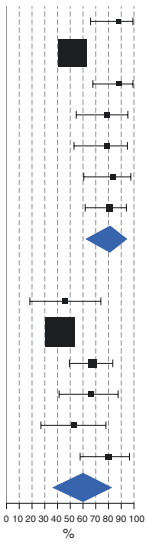


Table 9.4 The impact of surgical timing on >1 ASIA grade improvement in the incomplete thoracic/thoracolumbar tSCI

first author of study	total nr of patients	nr of patients with improvement	percentage improvement	lower 95%CI	higher 95%CI
Cengiz [23]	6	6	87.8	65.9	99.2
Du [24]	331	170	51.9	46.3	57.3
Rath [27]	7	7	88.6	67.9	99.2
Rahimi [22]	9	7	78.5	54.7	94.9
Clohisy [34]	9	7	78.5	52.9	94.8
Payer [26]	8	7	83.5	60.3	97.5
Dobran [32]	16	13	80.7	62.1	94.0
Early surgery	**386**	**217**	**81.3**	**62.2**	**94.3**
Cengiz [23]	8	3	45.6	18.3	73.8
Du [24]	380	158	41.8	37.0	46.9
Rath [27]	26	18	67.5	49.6	83.2
Rahimi [22]	10	7	66.3	41.1	87.6
Clohisy [34]	10	5	53.0	26.9	78.1
Wang [25]	11	10	80.4	57.7	96.4
Late surgery	**445**	**201**	**60.2**	**36.4**	**82.6**



The severity of the injury not only affects the functional outcome in tSCI patients but also negatively affects the lifelong incidence of long-term complications such as pulmonary, urogenital complications, pain, and pressure ulcers. These complications can lead to frequent re-hospitalization and are the cause of morbidity and even mortality.

9.9 Potential Complications

Early surgery does not lead to a higher postoperative complication rate nor mortality compared to delayed surgery. On the contrary, early surgical management appears to decrease the overall complication rate during the acute hospital phase. Nevertheless, there is a chance that a patient can deteriorate neurologically after surgery independently from surgical timing. After the acute phase, late complications due to tSCI can occur and are essentially related to the severity of the injury. These late complications include bowel, bladder, and pulmonary dysfunction, pressure ulcers, pain, spasticity, sexual dysfunction, and even increased mortality.

9.10 What Should Patient and Family Know?

The severity of the initial injury directly affects the neurological outcome. There is growing evidence that early decompression within 24 h has a positive effect on neurological recovery. An additional beneficial effect of an ultra-early intervention is not clear and yet to be elucidated. Not only is the severity of injury related to the ability to improve neurologically, but it is also related to the prevalence of long-term complications which can cause great morbidity and even mortality.

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Osteoporotic Fractures

10

Luiz R. Vialle and Emiliano N. Vialle

10.1 Definition

An osteoporotic fracture occurs when the bone trabeculae cannot support the normal compression forces under physiologic conditions or slight trauma; such injuries are also named "stress fractures," "frailty fractures," or "insufficiency fractures."

10.2 Natural History

Osteoporotic fractures are becoming more frequent due to the increased life expectancy, combined with metabolic alterations such as Vitamin D deficiency, poor calcium intake, sedentarism, and genetic influence. It is a common problem in women after menopause related to hormonal changes, even though it can also affect about 20% of men.

Patients may admit with minor trauma, coughing or sneezing, or even no trauma. Without detection and adequate care, the fractured vertebra may collapse, leading to increased kyphosis and eventually compression of the neural elements.

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10.3 Physical Examination

In general, a patient's main complaint is pain over the affected area, sometimes so intense that clinical examination may become difficult. Local tenderness can help to localize the affected vertebra (or vertebrae). This is an important step because initial imaging must be requested precisely; otherwise, the osteoporotic fracture may be missed. Some patients present with pain at the lumbo-sacral junction (irradiated pain), thus leading to wrong imaging level and potential misdiagnosis.

10.4 Imaging

Plain radiographs centered over the most painful area are needed. Sometimes the fracture is not visible on plain radiographs. However, persistent pain or worsening clinical picture may require a new set of radiographs; at this stage, the fracture will become visible, together with the associated deformity (collapse) (Figs. 10.1, 10.2, and 10.3). In this case, a magnetic resonance imaging (MRI)

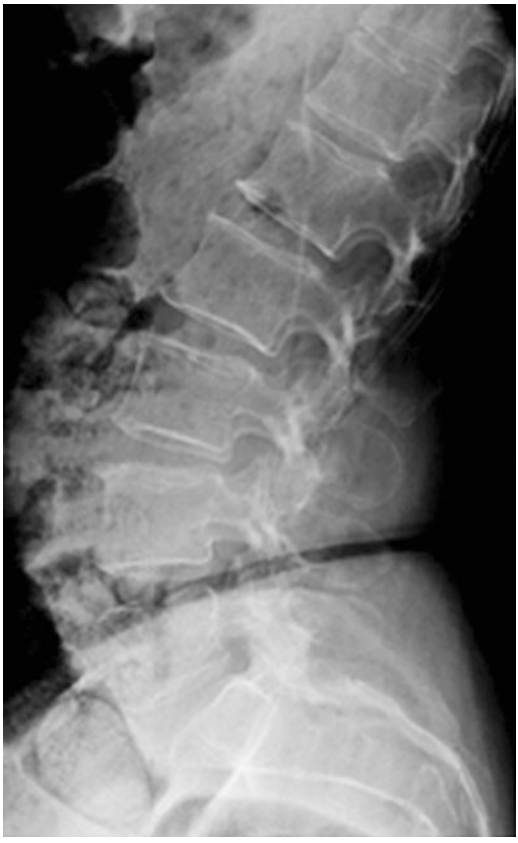

Fig. 10.1 Lateral direct radiogram reveals no signs of fracture of lumbar spine after a minor trauma

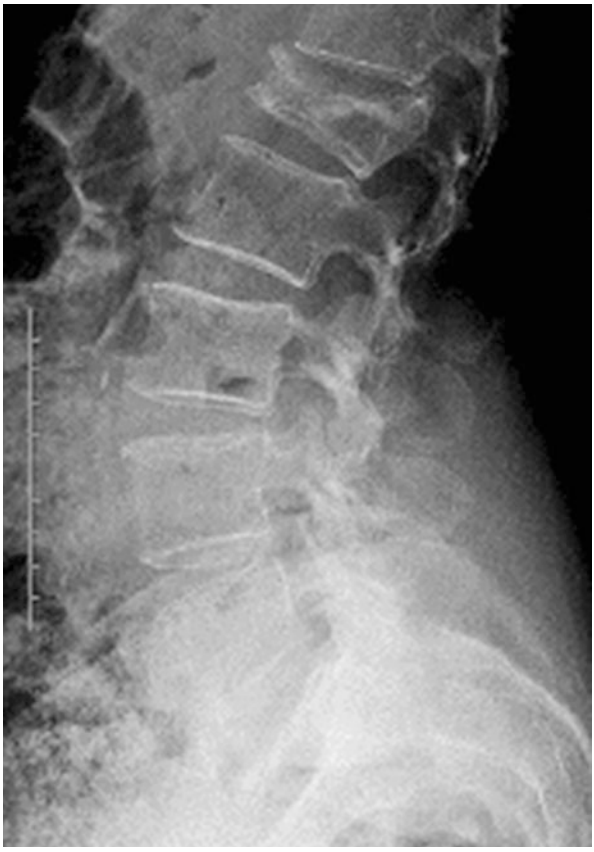

Fig. 10.2 Follow-up X-ray of the patient in Fig. 10.1 that taken 2 weeks later shows an L1 osteoporotic fracture

should be requested to better assess the fracture and to differentiate recent from old fractures (Fig. 10.4).

10.5 Differential Diagnosis

Insufficiency fractures are due to the weakness of the bone (trabeculae) and are indeed osteoporotic. However, the cause of osteoporosis must be thoroughly investigated in order to rule out a potential underlying disease; a full physical examination and laboratory data are needed. Primary tumors of the spine (mainly myeloma) are frequently related to secondary osteoporosis and insufficiency fractures (Chap. 61) though metastatic lesions can share the same clinical picture (Chap. 63). The practitioner must be aware of this condition in order to avoid misdiagnosis and subsequent complications.

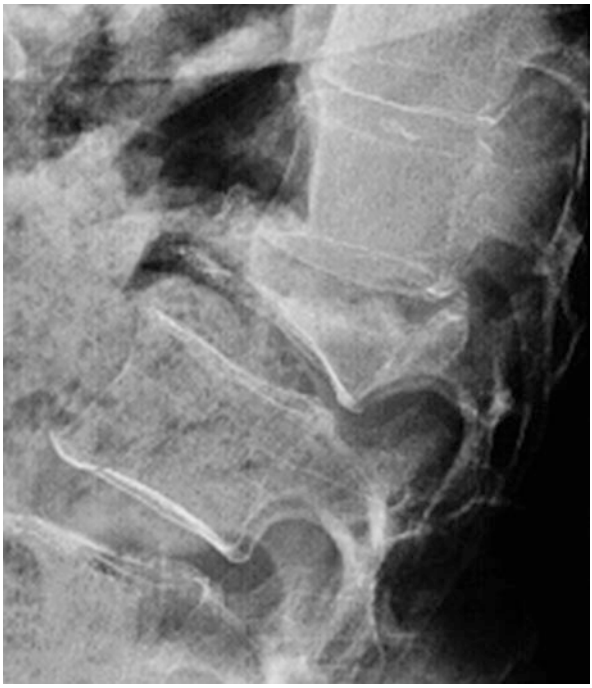

Fig. 10.3 Follow-up X-ray of the patient in Fig. 10.1 that taken 1 month later shows kyphotic deformity at L1 level

10.6 Classification

A classification for osteoporotic fractures was proposed by the German Spine Society and has been accepted by surgeons as a guideline for treatment together with other criteria [1] (Appendix E).

10.7 Treatment Options

Two actions are needed once the diagnosis of osteoporotic fracture is confirmed: treatment and prevention.

(a) *Treatment* is related to the fracture and aims to provide the best possible pain relief to the patient.

Conservative treatment is based on oral analgesics; non-steroidal anti-inflammatory drugs should be administered carefully while opioids should be avoided. Opioids represent an additional risk factor in this patients' population, decreasing their cognitive ability. The patient should be stimulated to walk whenever possible and, despite the pain, should be trained on daily life activities such as how to get out from bed or seating, among others. A physical

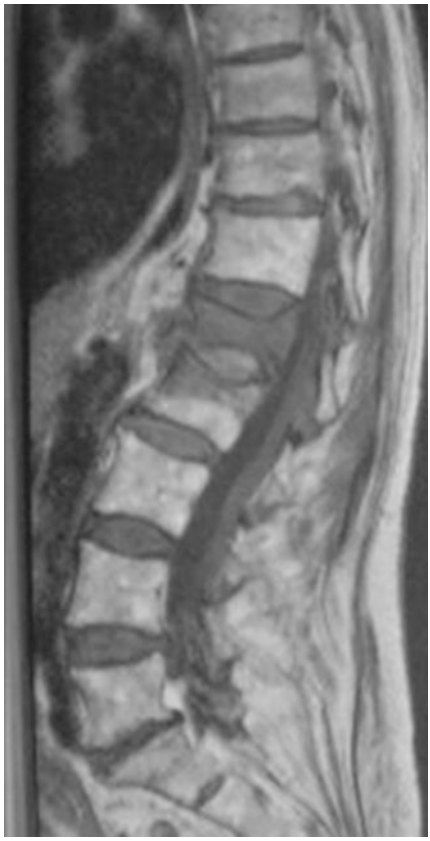

Fig. 10.4 T1-weighed MRI image showing signs of recent L1 and L2 fractures

therapist can be helpful at this stage. In some cases, a Jewett brace, forcing hyperextension, may be prescribed. With a careful clinical approach, effective medication, and psychological and family support, the majority of the patients can tolerate the pain during the first few weeks following injury. Conservative approach is the first option; the only reason to switch to an invasive method is unbearable pain for more than 3 weeks.

Surgery is indicated in case of persistent pain and/or neurological involvement. For pain control, cement injection (vertebroplasty) is a powerful method. When well indicated and performed, the pain relief is immediate, allowing the patient to return to normal life activities. Vertebroplasty is a non-demanding technique, with several companies producing many tools for safe injection of acrylic cement. To avoid cement leakage and correct the vertebral deformity, other techniques such as kyphoplasty and stents can also be performed. Recent literature reviews [2, 3] showed no essential differences among the various techniques, all being effective on pain treatment. Considering all procedures, techniques, and costs, vertebroplasty is the procedure of choice.

Some patients may present with neurological involvement. It may be from the initial trauma, with a posterior wall fragment compressing the dural sac. Depending on the level, some may present a spinal cord compression. Other cases may develop a kyphotic deformity, where the neurological deficit is progressive. In both cases, more aggressive intervention is needed to decompress the cord and to correct the deformity. The best surgical approach is a combined procedure (anterior decompression and mechanical support with a cage or mesh followed by percutaneous posterior screws) (Video 10.3); short fusions are better than long fusions: the osteoporotic spine when operated on with long fusions needs the association of augmentation techniques, making the procedure more riskier. However, in such cases, even with the associated morbidity, there is no other way to get a full decompression and segmental stabilization.

(b) *Prevention* is the second action to be taken; it requires evaluating the amount of osteoporosis and timely beginning the medical treatment, to avoid further bone loss and to improve bone quality and stock. Delaying the medical treatment increases the risk of new fractures. As soon as the pain is under control, the patient needs a lifestyle change and a continuous medical check of the osteoporosis status.

10.8 Expected Outcomes

Following the recommended protocols, conservative treatment is able to control pain in 3–4 weeks, with a progressive reduction in pain severity day after day.

Patients treated with vertebroplasty will experience significant pain improvement immediately after the procedure. However, new fractures should be expected, sometimes during the early post-procedural period. When open surgery is indicated, recovery is longer, and systemic complications can develop.

10.9 Potential Complications

The most dangerous complication of an osteoporotic fracture is the neurological deficit which can be secondary to a retro pulsed bone fragment of the posterior vertebra wall pushing on to the spinal cord, or to progressive kyphosis leading to spinal cord compression (Fig. 10.5); sagittal malalignment may be the result of multiple osteoporotic fractures (Fig. 10.6).

Intra-canal cement leakage may occur during vertebroplasty. The majority of cases will not have any consequence even though it may cause neural damage. In such cases, an open surgical decompression is recommended. Leakage into the intervertebral disc space is not uncommon and is not necessarily a complication, as it is usually asymptomatic.

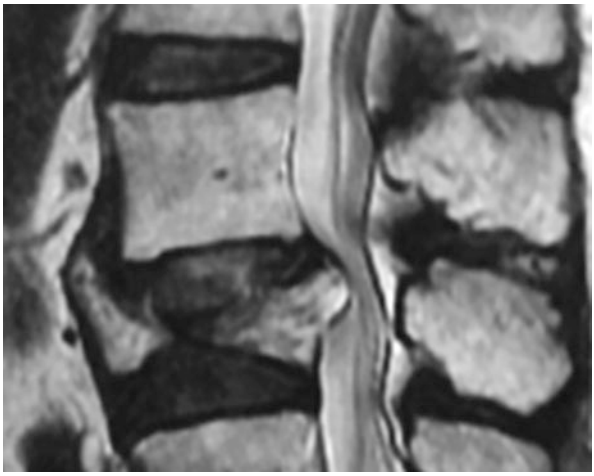

Fig. 10.5 T2-weighed MRI image showing a retro pulsed posterior wall due to an osteoporotic fracture

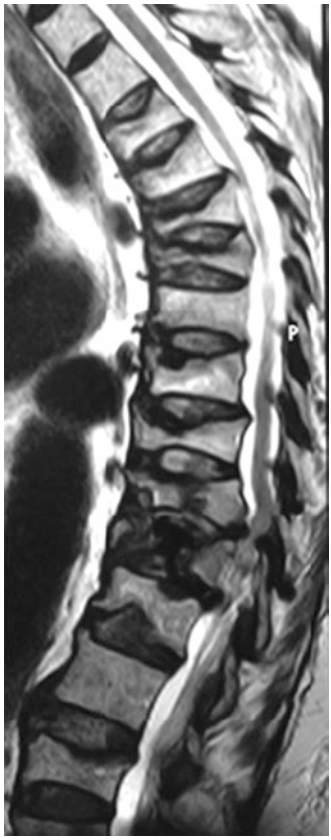

Fig. 10.6 T2-weighed MRI image demonstrating multi-level fractures and overall kyphosis

10.10 What Should Patient and Family Know?

Fractures related to osteoporosis mostly affect the aged population. The pain gradually improves during the first 3 to 4 weeks; most patients are generally pain-free after 3 to 4 months of medical treatment. Patients with co-morbidities or bedridden due to excruciating pain will benefit from surgery (and immediate pain relief).

Lifestyle changes, physical exercises, nutrition, and medical treatment are necessary to avoid new fractures.

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Part III

Pediatric Spine Pathology

Back Pain in Children

11

Federico Canavese

11.1 Definition

Painful sensation along the spine or back, sometimes with radiation down to one or both lower extremities.

11.2 Natural History

Pediatric dogma suggests that children with serious back pain (BP) may have serious pathology, including malignancy and infection. In the past, BP was considered a rare event highly associated with significant pathology. More recently, BP has been presented as a common occurrence unlikely caused by a significant underlying pathology.

However, there is still a small group of children with BP associated with a significant pathology; this is particularly true if the child is 4 years old or younger, or if a child of any age has BP associated with fever or weight loss, weakness or numbness, abnormal gait, pain that radiates down one or both legs, bowel or bladder dysfunction, and night pain that keeps the child from sleeping (*red flags*).

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11.3 Physical Examination

The history, the physical examination, and the search for *red flags* guide the clinician to rule out the most likely cause of BP and to perform the most appropriate diagnostic studies.

The onset, character, and location of BP, and any associated radiation of pain must be investigated. Although the majority of causes of BP are benign, clinicians must be alert to the potential *red flags* indicating serious pathology; red flags are warning signs that indicate the need for further work-up should not be missed: (1) age less than 4 years; (2) persistent pain; (3) muscle spasms/stiffness; (4) patient is unable to touch the ground with the knee extended; (5) stiff back or painful scoliosis; (6) pain not related to effort; (7) night pain; (8) significant functional repercussion; (9) neurological symptoms; (10) weight loss, fever, and sweating.

The examining physician must carefully observe the patient's posture, gait, and movement as the abnormalities may suggest a neurologic disorder.

Palpation is needed to gauge the degree and the location of any muscle spasm, as well as for any changes in skin texture. In particular, the skin should be inspected for cutaneous lesions (dimples, hair patches, and hemangiomas) (Fig. 11.1) frequently associated with intraspinal disorders (Chap. 31).

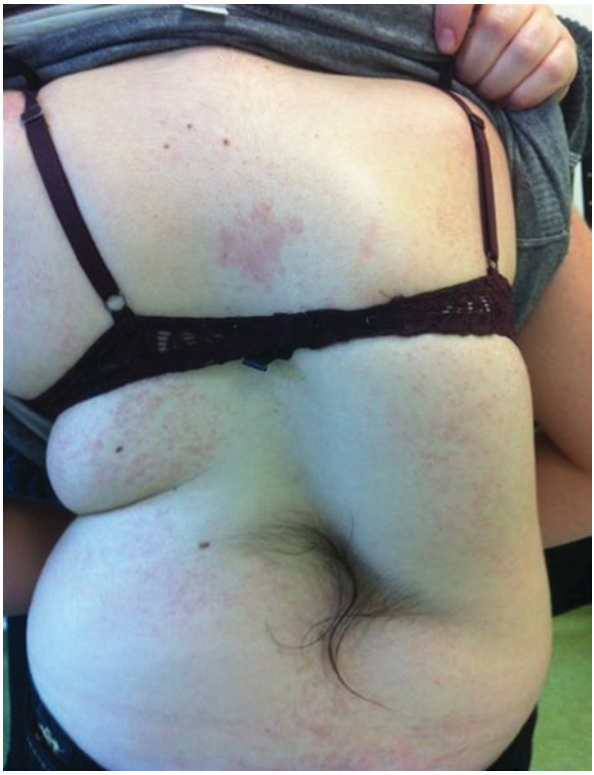

Fig. 11.1 Hair patches (lower back) and hemangioma (upper back) associated with underlying spine deformity (congenital) and spinal dysraphism

Body habitus, asymmetry of shoulder height, scapular prominence, flank crease, spinal curves, muscle bulk, or abnormalities on the skin should be recorded. Flexion, extension, lateral flexion, and rotations are tested bilaterally; and any gross limitations of movement, and pain on gross movement, must be recorded.

An extremely careful neurological examination is essential in patients with BP. Motor and sensory function and deep tendon and abdominal reflexes should be assessed in all patients. Absence or asymmetry of abdominal reflexes can be associated with syringomyelia (Chap. 31; Video 11.4) (Fig. 11.2).

Fig. 11.2 Left thoracic scoliosis and back pain; an MRI should be requested to rule an underlying pathology

11.4 Imaging

Plain radiographs are often the initial diagnostic imaging test. In general, anterior-posterior and lateral views are done initially to assess vertebral alignment, to detect vertebral endplate irregularities and disc space narrowing and/or vertebral scalloping, and to identify lytic or blastic lesions. Standard radiographs are not enough to detect soft tissue lesions.

Magnetic resonance imaging (MRI) is the most useful screening test for the evaluation of patients with BP; it is mostly used to evaluate the spinal cord, neural elements, intervertebral discs, and soft tissues. In particular, MRI provides accurate images of neoplasms, syringomyelia, infections, herniated discs, and disc bulges. MRI should complete the work out of every patient with abnormal findings on neurological examination.

A computed tomography (CT) scan provides good-quality images of calcified tissues and is the best imaging modality to characterize a bony lesion previously observed on plain radiographs or bone scintigraphy. It is particularly useful to evaluate fractures, deformities, tumors, infection, and herniated or ruptured disc of the spine.

Bone scintigraphy (BS) is a very sensitive diagnostic tool in localizing pathologic processes that affect the bone such as benign (e.g., osteoid osteoma) and malignant tumors, infections, and stress fractures. However, its specificity is quite low as it cannot define the precise nature of a lesion. BS is also helpful to assess the healing process of established lesions (e.g., stress fracture).

Single-photon emission computed tomography (SPECT) can be used when BS is non-diagnostic or equivocal.

11.4.1 Laboratory Tests

Laboratory tests should be ordered for patients with warning signs. Complete blood cell count with differential and peripheral smear, erythrocyte sedimentation rate (ESR), C-reactive protein (CRP), serum glucose, and immune-electrophoresis (IEP) should be obtained as an initial screening study. If the rheumatologic disease is included in the differential diagnosis, human leukocyte antigen (HLA)-B27, rheumatoid factor, antinuclear antibody, and Lyme titers should complete the workup. Urinalysis may complete the workup to screen for urologic conditions that may contribute to the patient's symptoms and signs.

11.5 Differential Diagnosis

The causes of BP in children should be ruled out: be suspicious! (Table 11.1)

Table 11.1 Causes, frequency, and type of back pain in children and adolescents

Cause and frequency of back pain	Age range	Pain
Common		
Muscle strain/overuse	>10 years	Related to activity
Postural		Related to posture
Spondylolysis		Lumbosacral
Spondylolisthesis		Lumbosacral; neurological symptoms in most severe cases
Trauma/micro-fracture		Usually mild; increases with activity; can radiate to the buttocks/legs
Less common		
Infection (discitis, spondylodiscitis, vertebral osteomyelitis)	<10 years	Low-back/abdominal pain, stiffness, gait abnormalities (limping)
Scheuermann disease	>10 years	Thoracic spine; increases with activity (most cases)
Trauma/fracture		Mild; increases with activity; can radiate to the buttocks and legs
Uncommon		
Disc bulging (herniated nucleus pulposus)	>10 years	Persistent; neurological compromise is rare
Bone tumor	All ages	Progressive; unrelated to activity and/or at night
Intracanal tumors	<10 years	Progressive; unrelated to activity and/or at night; neurological symptoms can be present
Juvenile rheumatoid arthritis	>10 years	Persistent
Ankylosing spondylitis		Persistent
Psychogenic[a]		Persistent without any organic cause

[a]In case of "psychogenic BP," a neurological exam performed by a neurologist must be requested

11.6 Treatment Options

The treatment of BP in children is heterogeneous and can include physiotherapy, medical treatment, infiltration, and surgery, depending on the underlying cause.

11.7 Expected Outcomes

In children and adolescents, BP prognosis is related to the type and severity of underlying pathology. Younger patients with BP associated with warning signs (*red flags*) require careful assessment to rule out serious pathology. BP in older patients, not related to activity (sport) or posture, and without any functional repercussion, is generally benign and is resolved with physiotherapy.

11.8 Potential Complications

Potential complications are related to missing the diagnosis of life-threatening pathology in a patient with BP.

11.9 What Should Patient and Family Know?

Painful spine in children should be taken seriously. Symptoms can be mild, even in presence of serious underlying pathology.

Backpack-related factors, type of transportation, and presence of lower limb discrepancy less than 2.5 com are not related to BP in children and adolescents.

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Congenital Scoliosis

12

Michael Ruf

12.1 Definition

The term *congenital scoliosis* includes deformities of the spine in the coronal plane caused by malformations occurring within the early embryonic period. The formation of the somites (precursors of the spinal segments) starts between 3 and 5 weeks of gestation while segmentation occurs between 6 and 8 weeks of gestation. Failures of formation result in hemivertebrae which may be fully segmented with two adjacent disc spaces, semi-segmented, or non-segmented (incarcerated) (Fig. 12.1).

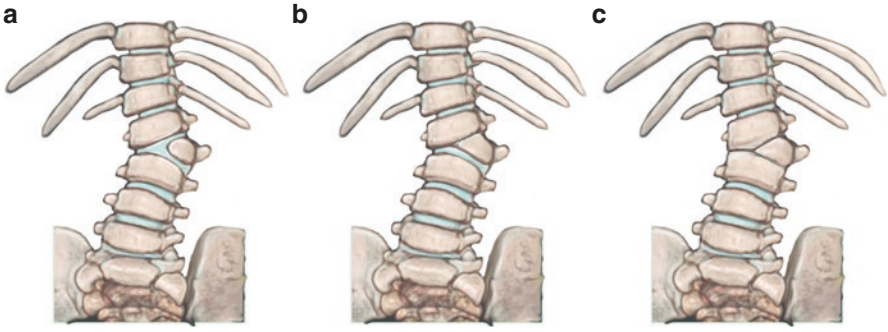

Fig. 12.1 Failures of formation: fully segmented (**a**), semi-segmented (**b**), non-segmented (incarcerated) (**c**)

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Segmentation defects include bar formations spanning one or more segments that may be located lateral, anterior, or posterior. Delay or cessation of growth within the bar usually results in asymmetric growth with progressive scoliosis, kyphosis, or lordosis (Fig. 12.2). Mixed deformities are frequent, often combined with abnormalities of the rib cage. Rib deformities, especially rib synostosis, may increase the scoliotic deformity of the thoracic spine (Fig. 12.3).

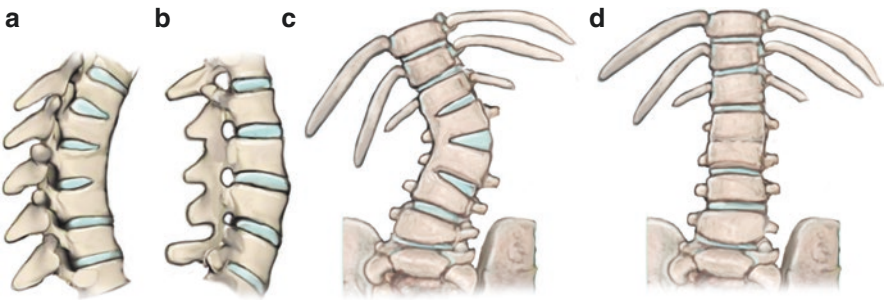

Fig. 12.2 Failures of segmentation: anterior bar (**a**), posterior bar (**b**), lateral bar (**c**), bloc vertebra (**d**)

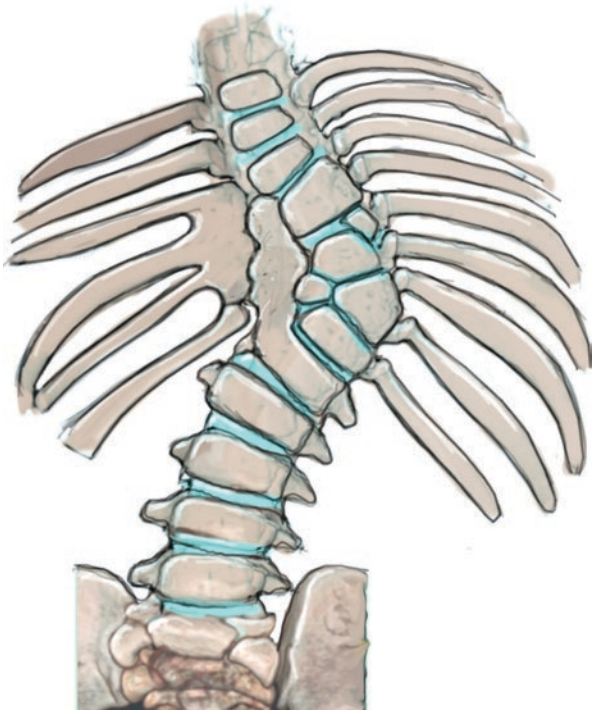

Fig. 12.3 Mixed deformity with hemivertebra, bar formations, and rib synostosis

12.2 Natural History

The natural history of congenital scoliosis depends on the type and the location of the congenital abnormality. Scoliosis progression may be expected during the growth spurt. The worst prognosis is usually seen in hemivertebrae with contralateral congenital bar (up to 14° of progression/year), followed by unilateral congenital bar, fully segmented hemivertebrae (1° to 5° of progression/year), and by non-segmented hemivertebrae which may remain stable [1].

The unaffected vertebrae adjacent to the congenital deformity are subject to asymmetric loading; in particular, asymmetric loading during growth can ultimately cause wedge-shaped deformity of the vertebral body. Over time, the stiffness of the main curve tends to increase. At the same time, secondary curves develop to promote trunk balance. These curves are flexible in the beginning but become structural with time.

12.3 Physical Examination

Physical examination includes documentation of

- Any deformity in the coronal plane.
- Pelvic/shoulder/head obliquity.
- Trunk balance/plumbline.
- Rib hump/lumbar prominence.
- Trunk height/ relation thoracic spine length - lumbar spine length.
- Any sagittal deformity.
- Flexibility of the curves/compensation mechanisms.

Neurological examination (Video 12.4) is mandatory although neurological compromise is rare; the examiner has to pay attention to pigment disorders, hairy patches, and other signs of dysraphism (Chap. 31). Associated malformations (cardiac, urogenital, gastro-intestinal; VACTERL syndrome) must be ruled out.

Special attention should be paid to congenital dislocation of the spine (rare malformation) secondary to a developmental failure of the spine and the spinal cord at a single spinal level (Chap. 24). The goal of treatment is to stabilize the spine in order to avoid secondary neurological damage (early cast immobilization and an early instrumented decompression-stabilization with circumferential fusion).

12.4 Imaging

Radiographs of the whole spine, anterior-posterior and lateral projection, in a standing position show the deformity secondary to the congenital malformation as well as compensatory curves (Video 12.6); radiographs are also needed to assess trunk balance and during follow-up. Bending films are obtained for preoperative planning, particularly to evaluate the flexibility of the secondary (compensatory) curves.

A computed tomography (CT) scan with three-dimensional reconstructions is important to evaluate the morphology and the location of the congenital hemivertebra as well as of the adjacent vertebrae and to provide information on bar formation or ribs synostosis. Three-dimensional reconstruction is helpful for surgical planning especially in mismatched abnormalities of the anterior and posterior structures; the size and diameter of the pedicles are measured to select proper implants.

Magnetic resonance imaging (MRI) is mandatory to evaluate the spinal cord and other soft tissue structures; malformations of the spinal cord must be detected or excluded.

The deformity is then classified according to the type and degree of the malformation.

12.5 Treatment Options

The goal of treatment is to achieve a straight spine with a physiological sagittal profile, with limited loss of motion, and limited growth deficit. To achieve these goals, early diagnosis and early surgical intervention in young children are needed due to the natural history of the disease: aging usually results in fixed, structural deformities (secondary curves), necessitating more aggressive surgical treatment usually resulting in longer instrumented fusions. If there is any doubt about further progression, especially in semi-segmented or incarcerated deformities, or hemivertebrae in the upper thoracic spine, frequent radiographic follow-up is mandatory.

In case of hemivertebra, resection of the hemivertebrae is the mainstay of treatment. Posterior resection with short instrumentation is the standard treatment option (Video 12.3). Surgery should be performed early before the adjacent vertebrae get deformed, and before secondary curves become structural.

Pedicle screws are inserted in the adjacent vertebrae (Videos 12.3 and 12.6). The hemivertebra and the adjacent discs are exposed and resected from the convex side. A cage may be inserted to ensure lordosis in the lumbar spine. In young children, the spine is highly flexible; thus, correction requires little force and is achieved by compression on the convexity via the instrumentation. With the early and complete correction of the focal deformity, the development of secondary changes can be avoided. Thus, the primarily healthy segments are allowed to grow physiologically: with short fusion, the overall growth deficit is minimal (Fig. 12.4) [2].

In the thoracic and lumbar spine, a posterior approach is usually sufficient. An additional anterior approach is required in the cervical spine, and it can also be performed at the lumbosacral junction in order to facilitate complete resection of the hemivertebra.

In patients with synostosis around the hemivertebra or congenital bar, especially older children or adults, an additional osteotomy at the concave side or a complete apical vertebrectomy is usually required; this procedure involves a complete disconnection of the spine [3].

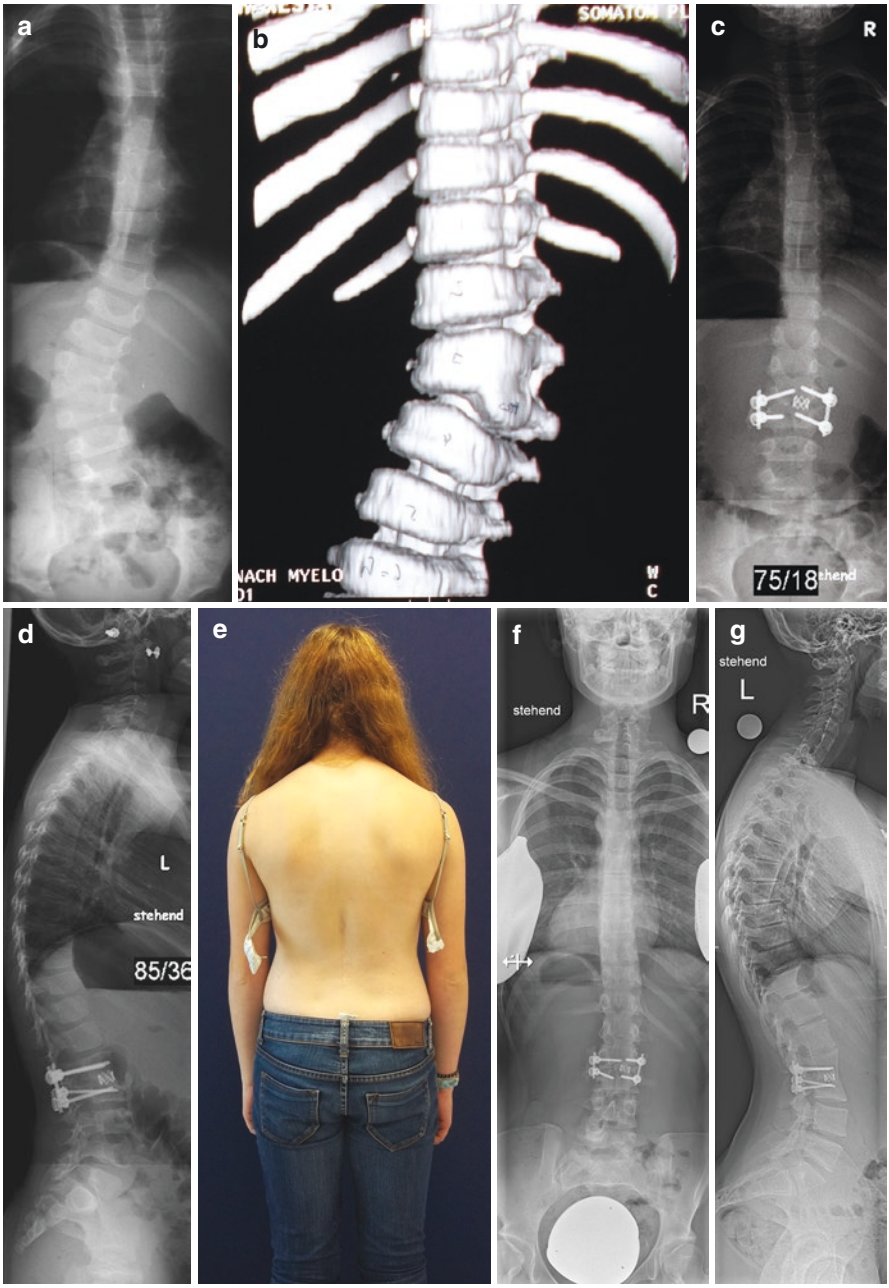

Fig. 12.4 A 4-year-old girl with semi-segmented hemivertebra L3a (**a**, **b**). Postoperative (**c**, **d**) at the age of 12 (**e**) and 17 (**f**, **g**) years

Multi-segmental deformities with a congenital bar in young children are often rapidly progressive. To avoid a long instrumented fusion potentially resulting in a short trunk, a combination of apical correction and "guided-growth" instrumentation of the adjacent segments may be useful.

12.6 Expected Outcomes

In patients with single hemivertebra, early resection with short fusion allows for complete and stable correction, and physiological growth. Complex deformity with congenital bar and rib synostosis may result in longer instrumented fusions and residual rib cage deformities. Depending on the primary growth plate abnormality, the trunk height may be short. Delayed treatment requires correction of the concomitant compensatory curves (usually stiff) resulting in longer fusion with the functional deficit.

12.7 Potential Complications

Neurologic compromise is rare. Progressive scoliosis may result in severe and very severe deformity; trunk imbalance, short trunk, pulmonary restriction, and loss of function are also possible.

12.8 What Patient and Family Should Know?

Patients must undergo regular follow-up until the end of growth. In the case of deformity progression, or new deformity, further surgery may be needed.

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Early Onset Idiopathic Scoliosis

13

Barlas Goker and Muharrem Yazici

13.1 Definition

Early onset scoliosis (EOS) is a progressive childhood spinal disorder which may potentially cause severe deformity, respiratory problems, psychological disturbances, and in most severe cases death. The management of EOS is a challenge for clinicians as the spectrum of treatment can vary between observation and complex surgical procedures. The management of children with EOS requires a multidisciplinary approach.

EOS is a spinal deformity regardless of the etiology with a curvature ≥10 degrees seen in children under 10 years of age. This age group is unique in that the spine is still growing (Appendix J). EOS can be further classified according to the etiology: *congenital scoliosis* results from abnormalities in the development of the spine (Chap. 12); *neuromuscular scoliosis* is caused by disorders of the central nervous system (Chap. 15); scoliosis due to an underlying syndrome (Marfan, Ehlers-Danlos, neurofibromatosis, Prader-Willi, etc.) are called *syndromic scoliosis* (Chap. 14). EOS without a specific etiology is defined as *infantile idiopathic scoliosis* for children 3 years or younger, and *juvenile idiopathic scoliosis* for children between 4 and 10 years of age.

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13.2 Natural History

EOS should not be considered as a sole cosmetic deformity (Appendix P). Without treatment, some deformities tend to progress. Severe progressive curves and early fusion may lead to decreased lung volume and function, a condition termed thoracic insufficiency syndrome (TIS). Normally, rapid thoracic growth begins before the age of 5, and it continues up to the adolescent growth spurt (Chap. 17). Alveoli continue multiplication during the rapid growth phase until about 8 years of age. Therefore, premature spinal fusion may hinder lung development and may cause lowered forced vital capacity (FVC). Maintenance of spinal growth and an upright posture by non-surgical and surgical methods (growth-friendly surgery) allows pulmonary development and it is of paramount importance.

Early fusion may also lead to deformity progression (e.g., "crankshaft phenomenon" and "adding-on" deformity). Even in series of anterior and posterior combined fusion, deformity progression has been reported in up to 40% of cases. Thoracic spinal growth inhibition is another issue; 57% of patients who were fused before 8 years of age had a thoracic height lower than a 5-year-old child's [1]. To avoid these spinal and pulmonary complications, growth-friendly techniques have been used in clinical practice.

13.3 Physical Examination

The physical examination of the EOS patient must start with the inspection of clues that indicate underlying associated conditions. The presence of hairy patches and dimples may suggest an abnormality of the spinal cord. Shoulder and waist asymmetry, truncal shift, and leg length discrepancies should be evaluated for global spinal alignment. A full neurological examination should be performed, and pathological reflexes must be noted (Video 13.4). The pulmonary examination includes auscultation and thumb excursion tests to compare hemithorax involvement in rib expansion.

13.4 Imaging

Deformity evaluation, Risser staging for skeletal maturation (Appendix M) and Cobb angle measurement (Video 13.6) is done over full-length anteroposterior and lateral images of the spine. Cobb angles more than 20°, rib-vertebral angle differences more than 20°, and convex rib head overlap with the apical vertebra (rib phase 2) are associated with curve progression. Hand and wrist radiographs are helpful in determining skeletal maturation according to Tanner-Whitehouse and Sanders classification systems (Appendix M). Computed tomography (CT) is utilized for patients with rib fusions and other structural abnormalities. Magnetic resonance imaging screening for intraspinal pathologies is recommended for most EOS patients, even in the absence of neurological examination findings.

13.5 Differential Diagnosis

As idiopathic EOS is a diagnosis of exclusion, other possible etiologies for EOS (congenital, neuromuscular, syndromic) must be ruled out. The age of onset determines whether it should be considered infantile or juvenile idiopathic scoliosis.

13.6 Treatment Options

Conservative treatment (brace, serial casting) is mainly performed to postpone surgery as much as possible (time buying strategy) although occasionally it may work as the definitive treatment, particularly in mild to moderate deformities. Braces are commonly used, and to be effective they must be worn for up to 23 h a day. Young children may experience discomfort and irritation due to a cylindrical trunk anatomy, leading to lowered patient compliance, and treatment failure.

Serial Risser casting (Fig. 13.1a–d) is applied under general anesthesia and traction (Video 13.5). Axial rotation of the curve is corrected with counter-rotation while the cast is molded. Care must be taken to avoid pushing the ribs toward the spine as this would compromise the lung volume. The cast is changed every 3 to 4 months (Video 13.5).

Numerous techniques for growth-friendly surgery have been developed. These techniques have been classified into three groups, according to the predominant forces exerted on the spine: *distraction-based systems* [dual growing rods, vertical expandable prosthetic titanium rib (VEPTR)] in which distractive forces are applied on the curve between proximal and distal anchors; *compression-based systems* (tethers, staples) that apply compression on the convexity of the curve; and *guided growth systems* (Luque Trolley, Shilla) where multiple anchors gradually slide over rods due to spinal growth.

The traditional dual growing rod (TGR) construct is a distraction-based nonfusion instrumentation system composed of a dual set of rods and anchors interconnected by a domino or telescopic connector. The children undergo lengthening in the operation room (OR) under general anesthesia, biannually. Lengthening is achieved by applying distraction on the rod systems. Until reaching maturity, children mostly undergo a multitude of lengthening procedures, often finalized with new instrumentation and spinal fusion.

Magnetically controlled growing rods (MCGR) theoretically overcome the need for repetitive surgeries that are required in TGRs. Instead of utilizing a telescopic connector, a remote-controlled magnet provides the lengthening (Fig. 13.1e–i). This allows the lengthening to be performed as an outpatient procedure, thus saving the patient from undergoing numerous surgical procedures which pose the risks of general anesthesia and surgical complications.

VEPTR was introduced by Campbell et al. in 1989 to counter the detrimental effects of progressive scoliosis on the respiratory system and TIS, by providing thoracic expansion alongside spinal deformity correction [2]. In the past, VEPTR

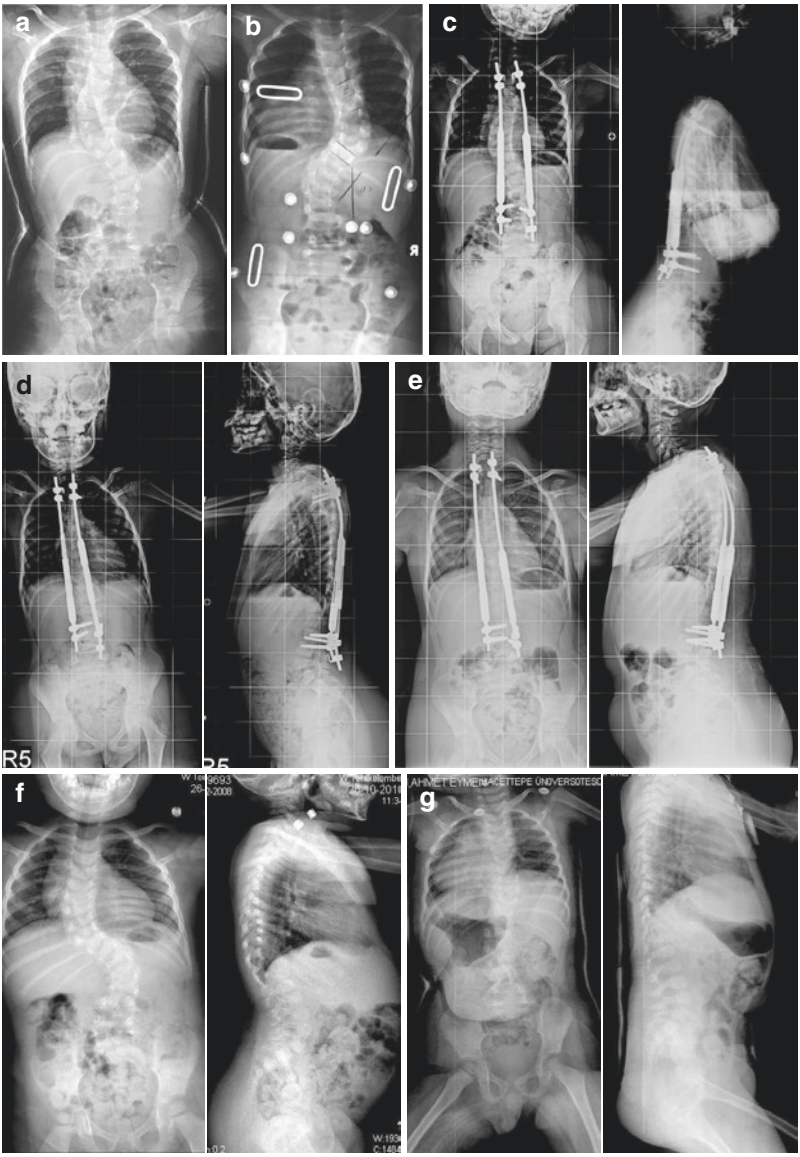

Fig. 13.1 A 3-year-old male patient with generalized ligamentous laxity and left hemihypertrophy (**a**) was started on serial casting (Video 13.5) (**b**). After 3 months in the cast, the parents refused treatment. 6 months later, he presented with deformity progression, and casting was repeated (**c**). At 5 years of age, after two attempts at casting, the family opted for a brace treatment (**d**). Although bracing provided balance, the amount of correction was inadequate. At 6 years, he underwent MCGR surgery (**e**) and was started on noninvasive lengthenings. There had been no complications for 2.5 years and the rods were exchanged due to reaching the lengthening limits (**f** and **g**). At 9 years, the rods were exchanged for a second time (**h**). As of writing, the patient is 11 years old and lengthenings are continuing (**i**). A definitive fusion with traditional spinal instrumentation is planned after removing the MCGRs at age 13–14

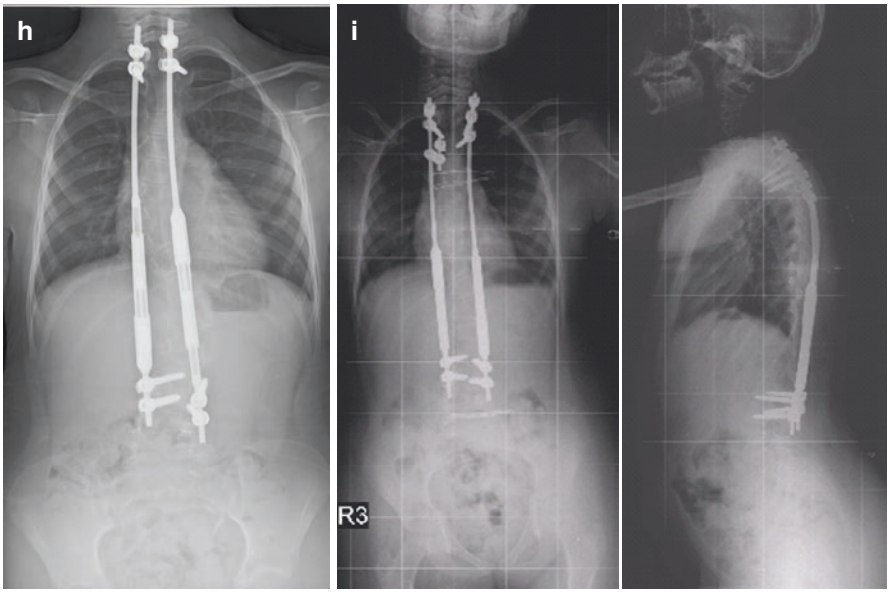

Fig. 13.1 (continued)

was used to treat some cases of idiopathic EOS; however, it has been mostly abandoned due to spontaneous rib and vertebral fusions.

Guided growth systems (Luque-Trolley, Shilla) are constructs in which rods glide over spinal anchors (wire, cable, or screw), thus allowing gradual deformity correction without additional surgeries. The pioneering guided growth technique is the Luque-Trolley construct. This method acts as an internal brace with sublaminar wires, fixed and gliding pedicle screws (Video 13.3). However, this method lost its popularity due to high rates of heterotopic ossifications and spontaneous fusions, and technically difficult revision surgery. The Shilla technique was developed to avoid the complications of Luque-Trolley related to extensive subperiosteal dissection. Subperiosteal dissection and fusion are limited to the apex of the curve with Shilla, and gliding anchors or screws are placed distally and proximally.

Anterior vertebral body tethering (VBT) is a thoracoscopic method of growth modulation in which the convex side of the spine is anteriorly instrumented by screws and staples connected with a tether for preadolescent IS. The use of tethers and staples allows gradual deformity correction by a "growth modulation" effect. The main advantage of this system is the ability to correct the curve without a final fusion surgery. Due to concerns over questionable resistance of vertebral body screws in cartilaginous vertebral bodies in young children and a high likelihood of overcorrection, its use is limited in EOS treatment.

13.7 Expected Outcomes

Initiation of casting at a younger age, moderate curves (<60°), and idiopathic scoliosis have shown better outcomes with serial casting. A "phase 1" rib phase and rib vertebral angle difference (RVAD) lower than 20° are also associated with higher success with serial casting. According to a recent meta-analysis, serial casting provides an average of about 25° Cobb angle improvement [3].

TGR systems are considered safe and effective for EOS treatment in children. Curve correction of 30° to 45° and a T1-S1 length increase of over 1.2 cm per year could be achieved by the time of final follow-up. The number of lengthening could be quite variable with a reported average between 4 and 7, which may sometimes go over 14–15 [4].

Although the number of total surgeries is lower in MCGR, most studies have found similar health-related quality of life outcomes, numbers of unplanned surgeries, and rates of complications between MCGR and TGR [5]. MCGR provides solutions to most of the issues seen in TGR; however, it is yet far from being an ideal treatment option.

13.8 Potential Complications

Skin irritation and wounds are common complications of nonoperative treatment that may occasionally require a brace or cast removal. Rib deformities, pressure sores, and rarely vascular complications such as subclavian vein thrombosis are other potential complications.

In contrast to overall successful results, TGR is also prone to a slew of complications. Among these, the most commonly encountered are implant-related complications (rod fracture, hook and screw dislodgement, prominent implants, etc.) and wound complications (dehiscence, superficial/deep infection). Systemic complications include pulmonary, cardiovascular (heart failure, inferior vena cava thrombosis), and neurological problems (dural tear, epidural bleeding). Repetitive surgeries and frequent hospital visits for TGR treatment might also lead to psychosocial impairment in children.

Migration of the curve apex due to initial apical correction and fusion, similar to an adding-on deformity, has been reported with the Shilla technique [6]. VBT is prone to complications including overcorrection, tether breakage, and curve progression. Intraoperatively, the patient must be closely monitored for possible pulmonary complications (pleural effusion, atelectasis, chylothorax).

13.9 What Should Patient and Family Know?

Follow-up until skeletal maturity is needed as the deformity can progress. Surgical risk is high.

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Syndromic Scoliosis

14

Krishna V. Suresh and Paul D. Sponseller

14.1 Definition

Syndromic scoliosis refers to the development of scoliosis secondary to a larger underlying disease process. Due to the large number of genetic and non-genetic syndromes involved, the clinical symptoms of these patients can be highly variable. Common syndromes associated with syndromic scoliosis include Marfan syndrome, Ehlers-Danlos syndrome, Loeys-Dietz syndrome, Trisomy 21 (Down syndrome), Rett syndrome, Achondroplasia, Neurofibromatosis, and Prader-Willi.

14.2 Natural History

The natural history of syndromic scoliosis varies from that of idiopathic scoliosis (Chap. 13). The age of presentation can be variable depending on the severity of the associated syndrome but is typically earlier in life compared to idiopathic scoliosis patients. Curve progression is more rapid in patients with syndromic scoliosis and can restrict pulmonary development especially in younger patients, leading to thoracic insufficiency syndrome. Early screening for scoliosis in pediatric patients with known syndromes should occur. In infants, this typically first starts when the child is able to sit independently, although no clear guidelines have been established.

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14.3 Presenting Signs

The presentation of patients with syndromic scoliosis is highly variable depending on the underlying disease process. Clinically important syndromes that should not be missed due to the high risk of associated morbidity and mortality without intervention include the following.

Marfan Syndrome: Defect in Fibrillin-1, a component of elastin polymer. Spinal deformity includes scoliosis >20° with increased kyphosis (Fig. 14.1). Patients are classically tall and thin. They are at a high risk of superolateral lens dislocation and retinal detachment. Patients are an elevated risk of dilation of the aortic root with the concurrent ascending arch aneurysm.

Loeys-Dietz: Defect in TGF-Beta receptor protein 1 or 2. Patients present with hypertelorism, arterial aneurysms, and a bifid uvula. Spinal deformity includes scoliosis but can also present with bifid anterior/posterior arch of C1, dural ectasia, significant C2-C3 instability with associated C3 body hypoplasia, and focal cervical kyphosis.

***Neurofibromatosis Type 1 (NF1)*:** Defect in a tumor suppressor gene that produces neurofibromin. Patients can present with café au lait spots, axillary or inguinal freckling, or Lisch nodules of the iris. Skeletal abnormalities include decreased bone mass density, bone dysplasia, tibial pseudarthrosis/bowing, dural ectasia, and early onset scoliosis (Fig. 14.2); C1-C2 rotatory dislocation can also be present and traction allows for neurological deficit recovery, if any. Rarely, patients can develop malignant peripheral nerve sheath tumors near or within the spinal cord, which can result in compressive neurologic symptoms (Chap. 23).

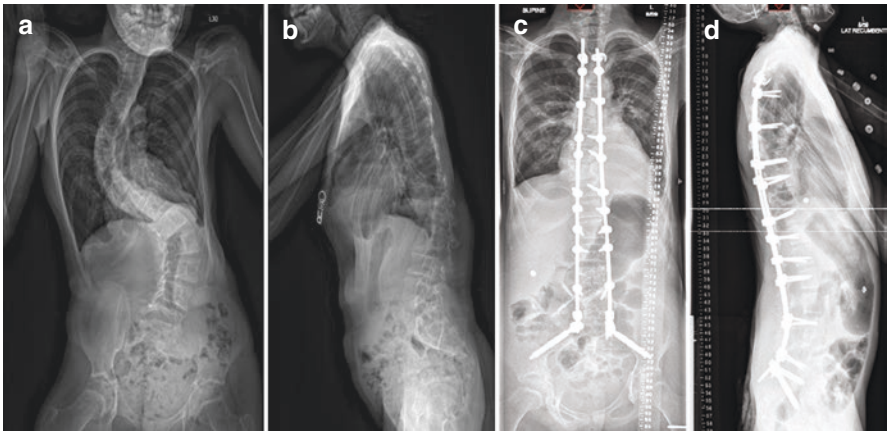

Fig. 14.1 Marfan's Syndrome–associated scoliosis in a 10-year-old female: (**a**) *AP view*: Significant right thoracic and left lumbar scoliosis. (**b**) *Lateral view:* Flattening of both thoracic kyphosis and lumbar lordosis. (**c**) *AP view status post-T3-pelvis posterior spinal fusion:* Curve correction of both right thoracic and left lumbar curves. (**d**) *Lateral view status post-T3-pelvis spinal fusion:* Restoration of adequate thoracic kyphosis and lumbar lordosis

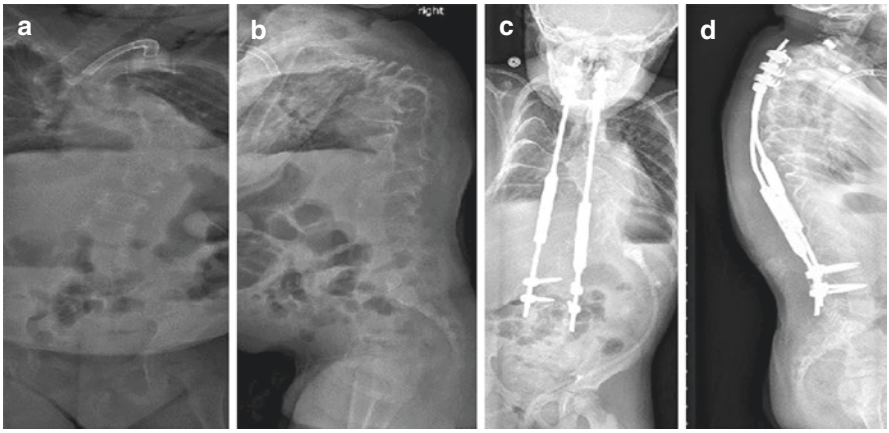

Fig. 14.2 NF Type 1–associated scoliosis treated with growth-friendly instrumentation in a 6-year-old male: (**a**) *AP view:* Significant right-sided thoracic curve and left-sided thoracic curve. (**b**) *Lateral view:* Significant proximal thoracic kyphosis (**c**) *2-year follow-up AP view*: Magnetically controlled growth rods placed with the patient receiving routine distraction every four months. (**d**) *2-year follow-up lateral view*: Restoration of adequate thoracic kyphosis

14.4 Physical Examination

Physical examination for syndromic scoliosis should involve evaluation of overall appearance, skin, and neurologic system in addition to the shape of the patient's back (Video 14.4). Thorough skin examination should be performed, looking for any cutaneous abnormalities such as café-au-lait spots, freckles, or neurofibromas. Height should be measured at each visit to monitor the patient's skeletal growth. Tanner's pubertal development should be assessed to evaluate pubertal maturity or associated delay. Patients with long, thin fingers and increased arm to height ratio should raise suspicion for Marfan syndrome and warrant the additional evaluation, including cardiac imaging. Joint laxity, range of motion, and skin hyperelasticity should be noted. A thorough neurologic examination should be performed, noting any focal neurologic deficits or radicular pain. Evidence of neuropathy in combination with significant cavus may suggest Charcot-Marie Tooth disease. When examining the back, the presence of any hair patches or skin dimples should be noted as these may be signs of spinal dysraphism (Chap. 31). Pelvic height should be measured for any inequality of lower-limb length, along with any inequalities in shoulder elevation.

14.5 Radiographic Examination

Radiographic evaluation of syndromic scoliosis involves determining the severity of the curve (Video 14.6), evaluating for abnormal vertebrae morphology or number, and obtaining additional imaging to assess complications of the associated

syndrome. Standing posterior-anterior radiograph should be obtained and assessed for two pedicles at every level, extent of deformity in the coronal plane, and severity of rotation. If no rotational deformity is present, additional evaluation for spinal masses, syrinx, or bony tumors should be performed. Spinal magnetic resonance imaging is also indicated in young patients under 10 years of age, those with neurologic deficits, and in patients who present with spinal dysraphism. In patients with suspected Marfan, obtain a cardiac echocardiogram. For patients with Marfan, biconcave vertebrae, transitional vertebrae, and increased transverse process distance are common, in addition to smaller pedicle widths and decreased laminar thickness.

Cervical spine instability must be ruled out in patients with Trisomy 21; in particular, atlantoaxial instability affects 10% to 20% of individuals with Down syndrome and it is mostly asymptomatic; the condition can be diagnosed on plain radiographs by an enlarged anterior atlanto-odontoid distance. Symptomatic atlantoaxial instability (about 2% of cases) manifests with spinal cord compression (Chap. 3).

14.6 Treatment Options

First-line treatment for syndromic scoliosis depends on the severity of the curve upon initial patient presentation. On presentation, Cobb angle >20° (Video 14.4), rib-vertebral angle difference (RVAD) >20°, and rib-vertebral overlap are predictors of curve progression. In infants, an RVAD greater than 20° or a curve greater than 35° warrants Mehta casting (Video 14.5). If the curve continues to progress to approximately 60° to 70° despite casting, consider growth rod placement. Similarly, in older children, curves of 25° or greater are managed initially with bracing, with definitive growth rod placement if the curve continues to progress to 50° to 60° (Videos 14.1, 14.3, 14.5 and 14.7). In syndromic scoliosis patients with thoracic insufficiency syndrome due to significant curvature, vertical expandable prosthetic titanium rib (VEPTR) can also be considered to aid lung development, till definitive growth rod placement or fusion can be performed.

14.7 Expected Outcomes

Syndromic scoliosis, compared to idiopathic scoliosis, is less likely to respond to bracing and casting measures. Surgical intervention in these patients is associated with greater risk of deep surgical infections, longer length of hospital admission, and increased risk of pulmonary embolism and neurologic injury. Device-related complications including failure and rates of pseudarthrosis are significantly higher in syndromic scoliosis patients.

14.8 What Should Patient and Family Know?

Syndromic scoliosis is a type of scoliosis that is secondary to an underlying disease process. The presentation of syndromic scoliosis is highly variable. Scoliosis in these patients starts earlier, progresses more rapidly, generally does not respond well to bracing, and has a higher risk of complications with surgery.

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Early Onset Neuromuscular Scoliosis

15

Kareem Kebaish and Paul D. Sponseller

15.1 Definition

Early-onset scoliosis is associated with a variety of neuromuscular disorders. In comparison to other forms of scoliosis, neuromuscular scoliosis often has an early onset with rapid progression during growth, which may continue to progress following skeletal maturity. It is more likely to present with severe spinal deformities that frequently involve the sacrum, and may have associated pelvic obliquity; respiratory compromise is frequent. The associated disorders are broadly classified as either neuropathic or myopathic (Chap. 20).

15.2 Natural History

Progression throughout life is common as well as a respiratory compromise; depending on the underlying disorder, progression can be extremely rapid.

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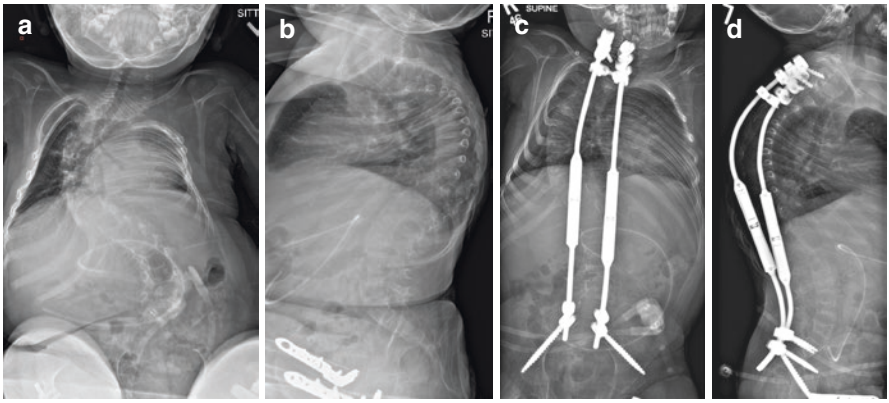

Fig. 15.1 A four-year-old patient with spinal muscular atrophy Type 1 with severe progressive kyphoscoliosis (**a** and **b**), treated with MCGR (**c** and **d**)

15.3 Physical Examination

Neuropathic disorders, including cerebral palsy (CP), spinal muscular atrophy (SMA), myelomeningocele, and Rett syndrome, are more commonly associated with early onset (Videos 15.4 and 15.5). In contrast, some patients with myopathic disorders such as Duchenne's muscular dystrophy are most likely to develop scoliosis after the age of 10 years old. Curve progression in all may continue after skeletal maturity, and a respiratory compromise is frequent. Patients with SMA are notable in that age of scoliosis onset may occur as early as less than 2 years of age (Fig. 15.1). Earlier onset is predictive of more severe development of scoliosis. Patients with myelomeningocele may develop tethered cord syndrome following surgical repair, which may be a cause of progressive scoliosis. Management of these patients is complicated by associated disorders including renal anomalies, and neurogenic bowel and bladder. Successful management of patients with myelomeningoceles requires a multi-disciplinary approach. Patients with Rett syndrome who present for management will be female, as the X-linked mutation leads to death in males within the first 2 years of life. As with other causes of neuromuscular scoliosis, patients with Rett syndrome can develop rapidly progressive scoliosis, which may continue after skeletal maturity.

15.4 Imaging

Radiographic evaluation of scoliosis involves determining the severity of the curve (Video 15.6), evaluating for abnormal vertebrae morphology or number, and obtaining additional imaging to assess complications of the associated syndrome. Spinal magnetic resonance imaging is also indicated in young patients under 10 years of age, those with neurologic deficits, and in patients who present with spinal dysraphism.

15.5 Treatment Options

Nonoperative management. Several goals of care, including maintenance of function and delay of surgical intervention (e.g., casting; Video 15.5), can be assisted by non-operative interventions. Bracing can be used to control the progression of curves during growth periods and delay the need for surgery until it can be performed safely. Patients with pulmonary conditions should be closely monitored, as bracing can cause decreased chest wall expansion. Wheelchair modifications, such as trunk support and posterior contouring, allow patients to sit in a functional position and maintain more independence. Frequent skin evaluations in patients treated with bracing or wheelchair modification are important in preventing the development of skin breakdown and pressure ulcers.

Intrathecal Nusinersen injections have improved the prognosis of SMA Type 1 and 2 patients. The medication is delivered directly to the central nervous system, which allows it to be distributed to the spinal motor neurons located in the spinal cord.

Surgery. Surgical intervention may be considered in patients with rigid curves greater than ~60° to 70°. A curve may be considered rigid when it has <50% of flexibility. Similar to conservative management, the goals of surgical intervention are to control curve progression and provide stability in the coronal and sagittal plane. Benefits include decreased pain, improved pulmonary function, improved seat positioning, and increased functional independence. Surgical options can be broadly categorized into definitive fusion and growth-friendly non-fusion techniques. In DMD patients cardiac function is an important parameter to consider when surgery is planned; anticipation is the best strategy.

Compared to idiopathic and congenital scoliosis, risks of surgery are greater in patients with neuromuscular scoliosis as a result of their underlying disorders. A preoperative assessment including a detailed history and evaluation of cardiac, respiratory, neurological, and urologic function should be performed. Furthermore, nutritional status and possible concomitant metabolic bone disease should be assessed.

The use of non-fusion surgical techniques has become favored in the treatment of early-onset scoliosis before a skeletal age of about 9 years as, compared to fusion, non-fusion techniques allow for the continued spine and chest growth. In addition to traditional growing rods, many non-fusion options are available, including magnetically controlled growing rods (MCGR), vertical expandable prosthetic titanium rib (VEPTR), Shilla growth guidance, self-expandable domino mechanical growing rods, and localized fusion in myelomeningocele (Videos 15.3 and 15.7). Compared to traditional growing rods, MCGR and the Shilla technique reduce the need for multiple operations to expand the construct as the patient grows. VEPTR constructs may be helpful in improving pulmonary function but require surgical adjustment every 6–8 months, increasing the risk of complications.

The extent of pelvic obliquity should be assessed in patients with neuromuscular scoliosis. Pelvic obliquity can cause uneven weight distribution while patients are sitting, leading to pressure ulcers and decreased sitting tolerance due to pain. Pelvic

fixation may be performed with fusion or as a part of a growing rod construct. The typical criterion for pelvic fixation is pelvic obliquity >10° to 15°, as measured on the anteroposterior radiograph.

Definitive surgical fusion is commonly viewed as the endpoint of treatment for neuromuscular scoliosis. Nevertheless, with the increased use of growth-friendly techniques, definitive instrumentation and fusion may not be necessary for all patients (Videos 15.1 and 15.5). Many patients treated with growth-friendly techniques undergo varying degrees of autofusion. Thus, patients with evidence of autofusion, minimal gain in length during their final distraction, and no implant-related complications can potentially be observed rather than undergo definitive fusion.

15.6 Expected Outcomes

Neuromuscular scoliosis, compared to idiopathic scoliosis, is less likely to respond to bracing and casting measures. Surgical intervention in these patients is associated with a greater risk of deep surgical infections, a longer length of hospital admission, and increased risk of pulmonary embolism and neurologic injury.

In comparison to idiopathic and congenital scoliosis patients, patients with neuromuscular scoliosis have high rates of complications associated with both treatment and their underlying disease state. Neuromuscular scoliosis patients, especially those with CP, have high rates of infection. Furthermore, patients may experience pulmonary insufficiency due to severe scoliosis curvature or associated deformities such as kyphosis. Uneven sitting posture and treatment with bracing increase risks of skin breakdown and pressure ulcers. Poor nutritional status in these patients is associated with decreased immune function, increasing the risk of surgical complications such as wound infections and sepsis. A goal serum albumin of >3.5 mg/dL should be reached prior to surgical intervention. Patients treated with growing rods that necessitate repeated distraction may experience progressive stiffness. In addition, instrumentation failure such as rod breakage or loosening of implants may occur. Finally, some curves may rapidly progress despite attempts to control them with growth-friendly techniques, leading to a more severe deformity that necessitates definitive surgical correction and fusion.

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Idiopathic Scoliosis

16

Jean Dubousset

Two important points should be remembered when dealing with patients with idiopathic scoliosis:

1. Stiff deformity, painful deformity, or abnormal abdominal reflexes are not suggestive of idiopathic scoliosis.
2. Maximum reduction of the deformity by surgery is not always the optimal for balance, harmony, and function.

16.1 Definition

Idiopathic scoliosis (IS) is a deformity of the axial skeleton that develops from head to pelvis (Videos 16.6 and 16.10); IS is in relation with the erect posture of the human and it is a three-dimensional deformity located in the sagittal, coronal, and horizontal (rotation) plane [1]; the torsion of the vertebral column is the key deformity. It is called idiopathic because until now no clear etiology has been established. Most advanced research converges to a genetic neuro-hormonal cause in relation to the erect posture in otherwise healthy individuals.

IS involves up to 3% of the population (>90% females), and it must be distinguished from a scoliotic attitude where torsion does not exist.

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16.2 Natural History

IS can be artificially classified according to age into infantile (0–3 years) (Chap. 13), juvenile (3–9 years), adolescent (9–end of growth) (Chap. 17) and adult (neglected case or de novo deformity) (Chaps. 48 and 49). The majority of IS cases show typical deformity progression (worsening of the torsion) during the adolescent growth spurt (Appendix M). The rate and severity of deformity progression during childhood and adolescence is extremely variable. Generally, the end of skeletal growth stabilizes the deformity even though the deformity tends to progress mainly at lumbar level during adulthood.

The three-dimensional architecture of the deformity [2] and the anatomical location of the deformity are responsible of the major related problems:

Thoracic spine: Apart from the cosmetic issue of the hump, the respiratory problems are predominant if significant coronal or sagittal deformity exists.

Thoraco-lumbar and lumbar spine: cosmetic issues (waist asymmetry) and balance problems are predominant; moreover, the rotatory disorders (degenerative) become predominant during adulthood and are responsible for pain (Chap. 45).

Physical Examination (1) Patients do not complain of pain nor stiffness. A painful and stiff scoliosis is from tumor, trauma, or infection until proven the contrary. (2) The patient is clinically checked in a standing posture (Fig. 16.1a): (a) to rule out lower limb discrepancy; (b) the plumb line dropped from C7 help to detect and

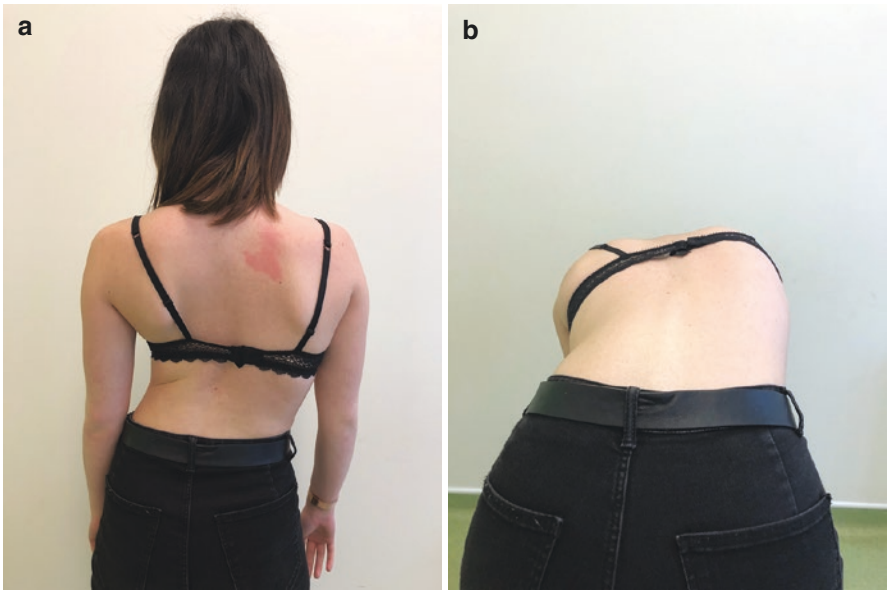

Fig. 16.1 Clinical view of a typical adolescent idiopathic patient. She has an apparent truncal shift and waistline asymmetry (**a**). Adam's forward bending test of the patient shows a right rib hump (gibbosity) which represents the axial rotation of vertebrae at the thoracic spine (**b**). (Courtesy of Prof. Jean Dubousset)

quantify right/left imbalance; (c) the Adam's forward bending test is used to detect, locate and measure the gibbosity (with the aid of the "scoliometer") (Fig. 16.1b); (d) to detect and record the severity of waist asymmetry (window sign); (e) additional signs: shoulder balance (elevated/symmetric/asymmetric), sagittal plane abnormality (kyphosis/lordosis; anterior hump. (3) Inspect the skin of the entire body to detect "café au lait" spots suggestive of Neurofibromatosis type-1 or McCune-Albright syndrome; search for angioma (midline or a tuft hair predicting spinal cord anomaly (Chap. 31). It is wise to check the elasticity of the skin and of the joints of the hand to rule out connective tissue diseases such as Ehlers Danlos, Marfan; if such diagnosis is suspected, echocardiography is more important than the radiographs of the spine in order to detect a latent aneurysm of the aorta! ***Complete neurological examination*** is mandatory: the examiner must check the cranial nerves, both upper and lower extremity osteo-tendinous reflexes, spasticity and clonus, Babinski, and extrapyramidal function (Videos 16.4 and 16.9); abdominal reflexes must always be texted as the absence of *cutaneous abdominal reflex* may lead to suspecting a spinal dysraphism (Chiari malformation, syringomyelia) (Chaps. 30 and 31).

Respiratory and pulmonary function (pulmonary function tests) must be assessed at rest and during effort. It is important to remember that *growth of the spine and growth of the lungs are linked together from birth to 7 years of age when alveolar multiplication stops.*

A precise evaluation of secondary sexual characteristics is needed though it requires a lot of tact; breast and pubic hair development in girls as well as knowledge about the menarche status: growth spurt starts with the first pubic hairs and stops with the full horizontal pubic hair development; the first menarche is the end of the ascending phase of the pubertal diagram (Appendix M). In boys the augmentation of testis size highlights the beginning of the pubertal growth spurt which ends with the appearance of the facial hair (beard).

Finally, it is wise to ***check the family history*** and to rule out if other family members have the disease because of the frequent genetic inheritance.

16.3 Imaging

Remember radiographs are only the shadows of the three-dimensional reality as they can show the spine in two planes only. The essential imaging is the full spine standing (anterior-posterior and lateral projections); the cervical spine up to the skull and the entire pelvis should be included (if possible with low dose radiation allowing a 3D computer reconstruction of the spine; EOS system). The full spine radiographs allow locating and measuring all the curves (Video 16.6) and identifying the apex of the deformity, the "junction" between curves and between spinal segments and the axial rotation of each vertebra. The Cobb angle with its perfect reproducibility is still the basic measurement; the evaluation of the apex and of the junctional levels must be done on both projections.

The flexibility of the curve can be assessed on traction or bending films or fulcrum bending films; the examiner must evaluate the Cobb angle (Video 16.6) and the rotation of the vertebrae. This allows differentiating between structural and compensatory curves; in particular, axial rotation does not disappear in structural curves while it disappears completely in compensatory curves.

A computed tomography (CT) scan and a bone scan have limited indications in patients with idiopathic scoliosis; CT is useful to measure the spinal penetration index.

Magnetic resonance imaging is highly recommended in the infantile group and every time a spinal dysraphism is suspected (left thoracic curve, painful scoliosis, abnormal neurological exam).

Three-dimensional reconstruction of the spine with vertebral vectors representation and view from the top help to evaluate in a glance the deformities and help to choose the levels to be treated [3]. Axial and intervertebral rotation can be measured by the Nash and Moe or the Perdriolle method. Bone maturation is assessed with the Risser sign, the hand and elbow maturation, and the vertebral listel ossification (Appendix M).

16.4 Treatment Options

At any age, treatment options can be observation, exercises, cast, brace, or surgery. Exercises alone are ineffective and the brace is often the treatment of choice (Chap. 18).

Surgery cannot be performed at any age to the negative effects of early arthrodesis on spine and thoracic cage growth and function; modern techniques allow the spine to grow even though autofusion and subsequent stiffness are frequent (Chap. 13).

For adolescents and adult patients, goals of treatment are Cobb angle correction, cosmetic improvement, balance and harmony of the spine, and the best possible function in relation to the three-dimensional mobility of the unfused segments above and below the fusion mass (major importance of selecting properly the limits of fusion) (Chap. 19). Surgery is indicated in case of deformity progression not responsive to brace treatment (>45°); however, current orientation is not to wait too much for worsening of the deformity but to get a reliable prognosis for progression and to treat by brace as soon as possible even with very low Cobb angle in order to prevent surgery (Video 16.6).

16.5 Potential Complications

Potential complications may occur following serial casting or surgical treatment.

Complications related to casting are pressure sores and brachial plexus injury; such complications can be prevented by good technical expertise and proper advice

to the patient and parents. Surgical complications can be (1) **neurological** (roots or cord): usually secondary to over-stretching of the cord or misused instrumentation; prevention: preoperative progressive awake traction, spinal cord monitoring (during surgery), wake-up test (during surgery); (2) **infection:** mostly early infection (<3 months) (Chap. 66); prevention: perioperative antibiotics; surgical setting. Treatment requires early reoperation in order to keep the hardware in place. Late infection (after fusion obtained) can also occur; the treatment consists of hardware removal and antibiotic treatment; (3) **nonunion or pseudarthrosis**: revision surgery (localized).

16.6 What Should Patient and Family Know?

Families and patients must be aware that (1) when brace (or cast) treatment is prescribed, compliance is mandatory, and patient and parents' collaboration is required; (2) a straight spine on post-operative radiographs is not the ultimate goal of surgical treatment: balance, harmony, and residual motion are the main goals.

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Progression Risk of Adolescent Idiopathic Scoliosis During Puberty and Natural History

17

Alain Dimeglio and Federico Canavese

17.1 Definition

The pubertal growth spurt (puberty) is the turning point in the natural history of idiopathic scoliosis; almost all curves progress during this period.

17.2 Natural History

Deformities less than 45° at skeletal maturity tend to remain stable during adulthood, while those over 50° tend to progress gradually, and surgery is generally recommended during adolescence (Video 17.6). Special attention must be paid to lumbar spine deformity; in this case, clinical examination plays an important role (vertebral rotation and waist asymmetry) while Cobb angle (45° or more) is not the only parameter to take into account as some deformity may require surgery below this threshold.

Patients with untreated scoliosis do not experience an increased mortality rate although on rare occasion (<1%) the deformity can progress to the point of causing death by *cor pulmonale* (pulmonary hypertension and right heart failure); the rate of

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dyspnea is slightly increased and it is associated with thoracic curves exceeding 80° (heart compromise can be seen in curves exceeding 90°). Most patients with untreated scoliosis function at or near normal levels, even though pain is more prevalent.

17.3 Physical Examination

In order to monitor growth, biometric measurements should be repeated at regular intervals.

Standing height is a global marker and it can be divided into two parts: sitting height and sub-ischial height. Because these two components often grow at different rates and times, standing height does not always correlate with trunk height loss. Final height is reached 2.5–3 years after menarche, which occurs at Risser I (Appendix M).

Sitting height correlates strictly with trunk height and the loss of sitting height is related to the severity of the deformity.

The measurement of *arm span* is an indirect evaluation of standing height (about 97% of arm span), and it is particularly helpful to estimate standing height in non-ambulatory patients.

Weight increases by 20-folds from birth to skeletal maturity. At age 5 years, the weight is about 20 kg, it becomes 30 kg by age 10 years, and it reaches 60 kg or more by age 16 years; during the pubertal spurt, weight usually doubles, and each year of puberty corresponds to a weight increase of about 5 kg. In children with low weight, pubertal spurt changes are moderate as the weight has to be at least 40 kg for the pubertal spurt to be normal.

17.4 The Pubertal Cycle

The pubertal cycle is the same regardless of ethnic origin and it follows a stereotypic course.

Growth velocity provides the best indicator of the beginning of puberty, on which so many decisions rest. The first sign of puberty is the rapid and significant increase of the standing height (more than 0.5 cm/month or more than 6–7 cm/year; this period is called *peak height velocity* or *acceleration phase*). At the onset of puberty, boys and girls will have about 22.5 cm and 20.5 cm to grow (standing height), respectively.

The appearance of secondary sexual characteristics signals the beginning of the pubertal growth spurt (Tanner stage 2); in particular, the first appearance of pubic hair, the budding of the nipples (93% of girls; 2 years before menarche), and the swelling of the testes (77% of boys; 3.5 years before attaining adult height) are the first physical signs signaling the beginning of puberty.

The first 2 years of puberty end with menarche and are followed by 3 years of gradual reduction of growth rates (*deceleration phase;* Risser I through V). During

this phase, the average remaining growth on the standing height is about 6 cm and 5.5 cm for boys and girls, respectively.

17.5 Radiographic Examination

Bone age assessed on hand, elbow, and pelvis radiographs is an essential parameter to evaluate remaining growth (Appendix M).

Puberty starts at the bone age of 11 and 13 years in girls and in boys, respectively; at this stage, the sesamoid bone of the thumb (left hand radiograph) is clearly ossified. The Y-cartilage ossifies about 1 year after the onset of puberty (mid-point of the acceleration phase; 12 and 14 years of bone age in girls and in boys, respectively) (Fig. 17.1).

The acceleration phase ends at age 13 and 15 years of bone age in girls and in boys, respectively. At this point the elbow (radial head, medial and lateral condyle, trochlea, olecranon) and the distal phalanx of the thumb are completely ossified; remaining growth is about 6 cm in boys and 5.5 in girls (Fig. 17.1). It is important to note that during the whole acceleration phase, the left iliac crest is still not ossified (Appendix M).

The deceleration phase follows the complete ossification of the elbow. At this stage the distal phalanx of the left-hand is ossified, Risser sign is I (onset of menarche in girls). In particular, 42% of the girls experience menarche before Risser I; 31% at Risser I (remaining growth: 4 cm; lower extremities growth: completed); 13% at Risser II (remaining growth: 3 cm); 8% at Risser III (remaining growth: 2 cm); 5% at Risser IV (remaining growth: 1 cm). Two years after menarche, there is usually no more growth (Risser V). The ossification of the greater trochanter (mid-point of the deceleration phase) occurs at Risser II-III (14.5 and 16.5 years of bone age in girls and in boys, respectively) (Fig. 17.1).

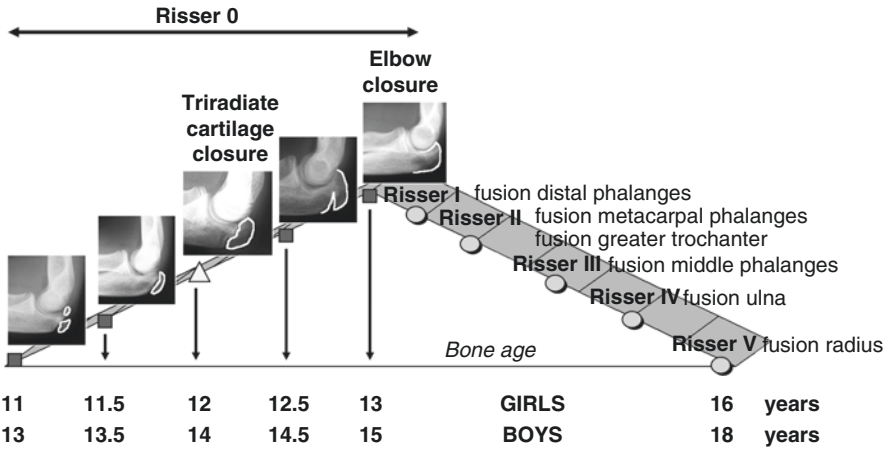

Fig. 17.1 Pubertal diagram. The first 2 years of puberty represent the acceleration phase while the last three (Risset I through V) the deceleration phase

17.6 The Scoliotic Risk: Anticipation Is the Key to an Effective Strategy

The scoliotic risk varies with growth, and it is proportional to both the amount of remaining growth and the severity of the deformity (Video 17.1). It is high during the *acceleration phase* (11–13 and 13–15 years of bone age in girls and in boys, respectively), and it decreases progressively from Risser I through V (*deceleration phase*). About 75% of curves 20–30° at the onset of puberty end up with surgery; the percentage rises to almost 100% if the deformity is more than 30° at the onset of puberty.

During the acceleration phase, a 5° curve is associated with a 10% risk of progression, a 10° curve has a 20% risk of progression, a 20° curve carries a 30% risk of progression, and a 30° curve raises the risk of progression to virtually 100%; moreover, any spinal curve increasing by more than 1°/month (or more than 12°/year) is likely to be a progressive curve requiring treatment. Any curve that increases by 0.5–1°/month (between 6 and 11°/year) during this phase must be monitored closely while any curve increasing less than 0.5°/month (or less than 6°/year) can be considered mild.

On the other hand, during the deceleration phase, the scoliotic risk decreases progressively (Risser I: 10% risk for a 20° curve and 60% risk for a 30° curve; Risser II: 2% risk for a 20° curve and 30% risk for a 30° curve; Risser III: 12% risk of progression for a curve of more than 20°; Risser IV: risk of progression is markedly decreased; Risser V: skeletal maturation is completed) [1–3] (Figs. 17.2 and 17.3).

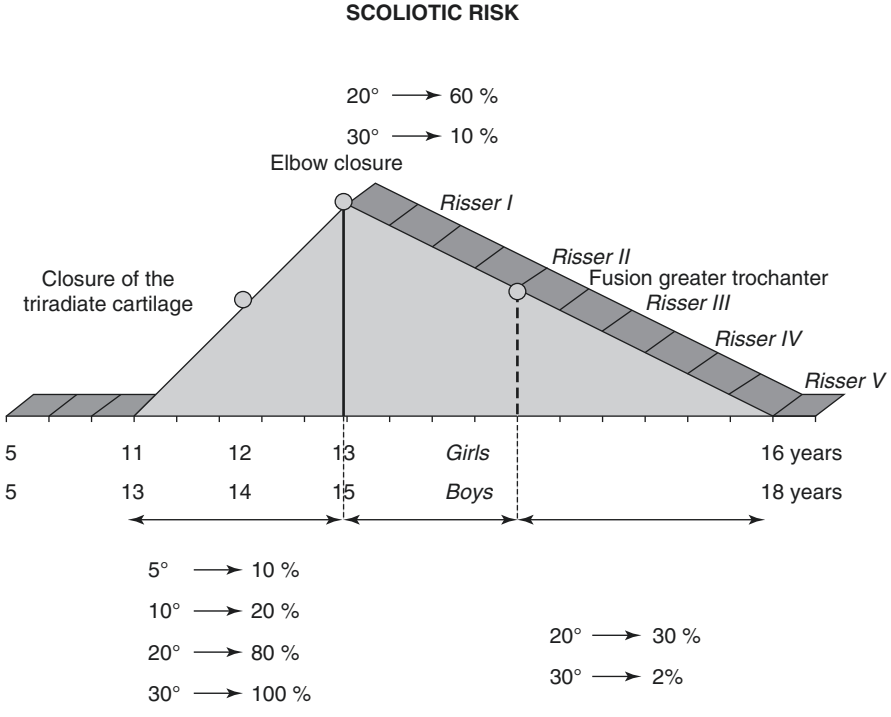

Fig. 17.2 Scoliotic risk

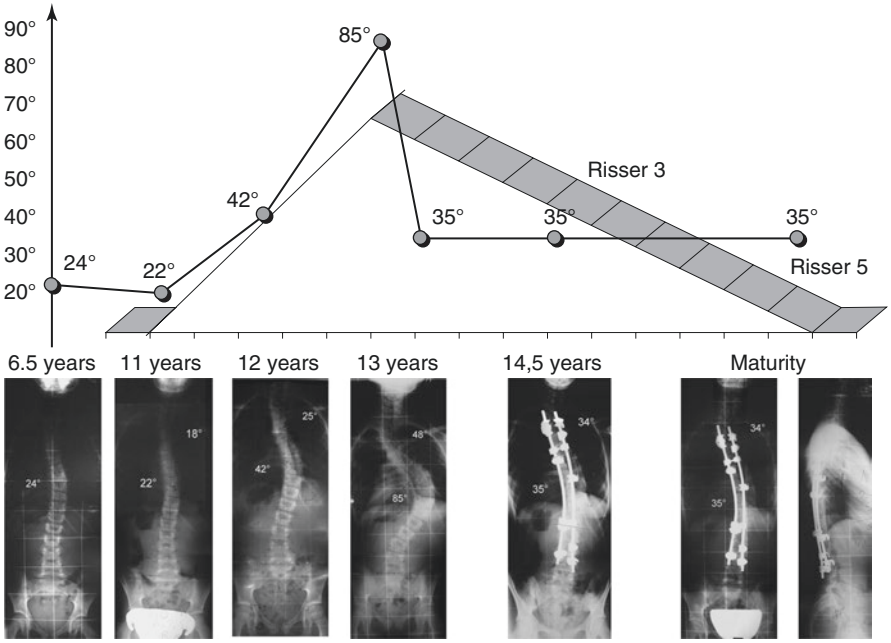

Fig. 17.3 Deformity progression during pubertal growth spurt

17.7 Treatment Options

Most curves require observation. About 10% and 0.1% of curves progress to the point that treatment with bracing and/or surgery is warranted, respectively (Chap. 18).

17.8 Expected Outcomes

Untreated adolescent idiopathic scoliosis patients do not have an increased mortality rate. The outcome of brace treatment is good in about 70% of cases. The result of surgery is usually satisfactory although there is a lack of long-term studies.

17.9 What Should Patient and Family Know?

Adolescent idiopathic scoliosis affects about 3% of the general population. Many of these children will not have curves progressing beyond 20° and do not require active treatment. Almost all curves progress during puberty; in particular, thoracic curves have the highest risk of progression. The risk of progression is related to the severity of the deformity at the onset of puberty.

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Conservative Treatment for Adolescent Idiopathic Scoliosis

18

Altug Yucekul, Caglar Yilgor, and Ahmet Alanay

18.1 Definition

Conservative treatment is the first-line, and the most common treatment approach for adolescent idiopathic scoliosis (AIS). Conservative treatment includes a wide spectrum of devices and activities including orthotics, physiotherapy scoliosis-specific exercises (PSSE), and electrical stimulation. Treatment should be individualized. The magnitude (Video 18.6) and location of the curve and the patient's skeletal maturity at initial evaluation help decision-making and the determination of the frequency of follow-up visits.

18.2 Treatment Options

Observation is the first step of an active approach. In general, no treatment is needed for curves less than 25° (Video 18.6), regardless of the patient's skeletal maturity. For small curves (<20°) in growing children, follow-up should be scheduled approximately every 6 months and 5° to 6° increase is indicative of curve progression. It must be remembered, however, that waiting for curve progression might be time lost for treatment.

Supplementary Information The online version contains supplementary material available at (https://doi.org/10.1007/978-3-030-80356-8_18).

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18.2.1 Orthotic (Brace) Treatment

Bracing consists of using a corrective orthosis for a specific period of time, each day. The corrective forces are passive, the predominant corrective component being the transverse loading of the spine through the use of corrective pads. The overall primary mechanism of action of a brace can be bending, detorsion, elongation or three-points, although many brace designs use combinations of such. General indications for brace treatment are: age >10 years at brace prescription, with significant growth potential (Risser 0 to 2/3) (Appendix M), Cobb angle between 25° and 40° (Video 18.6), and no prior treatment and potentially progressive curves [1]. These indications, however, are alterable in regards to factors such as remaining growth potential, documentation of progression and location of deformity.

A wide variety of braces with different treatment strategies have been described. They can be classified as soft (e.g., SpineCor) or rigid; or symmetrical (e.g., Milwaukee, Boston) or asymmetrical (e.g., Rigo-Cheneau, Gensingen); monocot, bivalved or multisegmented. Treatment protocols can comprise nighttime (8 h to 12 h per day), part-time (12 h to 20 h per day), and full-time bracing (20 h to 24 h per day).

Although the use of such thoraco-lumbo-sacral orthoses (TLSOs) is common today, they are more effective in patients whose curve apex is at T7 or below. Thoracic curves with an apex above T7 and cervicothoracic curves are difficult to control with a brace; hence they are not ideal candidates.

Contraindications of brace treatment are large curves (>45°; except in very immature adolescents and/or as a delaying tactic), extreme thoracic hypokyphosis, skeletal maturity, and patients that emotionally cannot tolerate wearing a brace.

Low body mass index, osteopenia, flattened physiological kyphosis, high initial trunk rotation, lower reduction of in-brace vertebral rotation, and lumbar pelvic relationship (LPR) angle greater than 12° and high angle trunk rotation have been shown as possible reasons for treatment failure. Compliance, hump magnitude, and in-brace correction have been proposed as the most important predictive factors of successful treatment. There is a dose-effect response of bracing and full-time bracing (~20 h) is more effective than part-time brace (<12 h) (Fig. 18.1).

The brace should be worn until the maturation is completed. Brace wean off criteria are (1) no height gain for the last 6 months, (2) closed distal radial/ulnar epiphysis, (3) Risser grade-5, (4) and 3 years after menarche.

18.2.2 Physiotherapy Scoliosis-Specific Exercises

PSSE are curve pattern exercises and therapeutic interventions, which can be used alone or in combination with bracing or surgery according to the individual indication. There are different approaches to PSSE (e.g., Schroth, Lyon, SEAS, Barcelona, Dobomed) with the same goal: auto correction in the three dimensions, training in

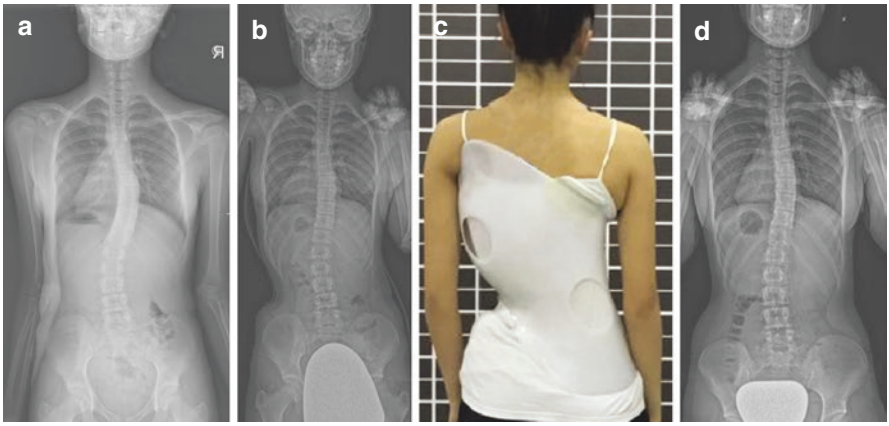

Fig. 18.1 (**a**) A 11-year and 8-month-old patient was diagnosed with adolescent idiopathic scoliosis with curves measuring 30° in the main thoracic and 25° in the thoracolumbar areas. Brace treatment with asymmetric dynamic Rigo-Cheneau Brace was initiated along with Schroth exercises. (**b**) Both curves were measured as 15° in the in-brace radiographs taken 3 weeks after first fitting. (**c**) Good elongation and midline alignment were achieved and breathing mechanics were observed to well function clinically. The brace was renewed twice as the patient overgrew her brace. (**d**) Three and a half years later, radiographs displayed two balanced curves measuring 20° in the main thoracic and 15° in the thoracolumbar area with good midline alignment

daily living activities, stabilizing the corrected posture, and patients' education [2]. The current level of evidence is that PSSE along with bracing can improve results better than brace treatment alone, especially in terms of health-related quality of life, and functional and psychological outcomes. Thus, combining PSSEs with brace treatment is highly recommended. However, specific PSSE have been associated with different brace designs (e.g., Schroth and Cheneau brace, SEAS and Sforzesco brace). This creates limitations in evaluating the effectiveness of the results and making comparisons.

18.2.3 Other Conservative Treatments

Electrical stimulation was used as an alternative; however, today it is no longer considered as an effective method for the management of patients with AIS. Performing general aerobic activities and respiratory training are recommended to regulate abnormal ventilation, improve exercise capacity, and control asymmetric movement of the chest wall via respiratory muscle functioning. Most PSSE approach use specific breathing techniques as an integral part of the exercise treatment.

In order to achieve the best possible outcome, conservative treatment should be delivered by an experienced therapeutic team including a physician, a physiotherapist, an orthotist, and possibly a psychologist.

18.3 Expected Outcomes

The main goals of conservative treatment are: (1) to have better results than natural history (Chap. 17), (2) to halt curve progression or to reduce curve magnitude to avoid fusion, (3) to improve aesthetics via postural correction, and (4) to prevent respiratory dysfunction and spinal pain.

The treatment can be considered as successful if <5° curve progression is observed (including curves that are stable and improving), or the curve measures less than 45° Cobb angle (surgical threshold; Chap. 19) at maturity. Different treatment methods and techniques have different results in obtaining the expected outcomes. Rigid braces are more effective than soft ones, especially for patients at high risk of progression during the pubertal growth support. There is no clear evidence regarding whether asymmetric braces are more effective than symmetric braces; however, greater correction in single curve patterns can be achieved with asymmetric braces. Recently, CAD/CAM braces came to the fore. Although such braces cannot "a priori" guarantee successful outcomes, they offer the possibility to achieve a better in-brace correction with lighter and more comfortable devices. At brace weaning, some loss of correction should be expected. PSSE can markedly reduce the loss of correction, so the implementation of PSSE at this stage is of utmost importance. Supervised PSSE exercises have been shown to have better results compared to home exercises.

The methods are interrelated (dynamic continuum) and can be adjusted during treatment according to the change in the patient's clinical status; however, widely recognized conclusions on the type and amount of treatment, as well as predictive factors leading to successful outcomes, are not known completely.

18.4 Potential Complications

Choosing the correct method of conservative treatment is mandatory, in order to avoid over-treatment (too much burden) or under-treatment (treatment without efficacy). Physical changes related to compression of the brace and psychological disturbance due to appearance are the two major concerns. Skin color changes, pressure sores, and cutaneous nerve impingement are common side effects of bracing. Prolonged symmetrical rigid brace wearing can cause tubular thorax deformity. Reflex esophagitis and decrease in total lung capacity have also been noted. In Milwaukee brace, temporomandibular joint problems can be seen. However, the most difficult effect to manage is the psychological impact of brace treatment.

18.5 What Should Patient and Family Know?

When diagnosed and treated early, AIS can be managed conservatively [3]. A common feature of all forms of conservative treatment is the need for the active involvement of the patient and caregivers. Therefore, education, psychotherapy, systematic

monitoring, assessment of compliance, and verification and modification of methods during the course of the therapy are deemed crucial elements for the successful outcome of conservative treatment. Willingness to actively participate in the different stages of the treatment plays the most important role. Besides families, support groups and Internet forums can help increase compliance.

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Determination of the Fusion Levels in Adolescent Idiopathic Scoliosis

19

Aaron F. Zhu and Kenneth M. C. Cheung

19.1 Definition

The goals of surgical management of adolescent idiopathic scoliosis (AIS) include maintaining coronal and sagittal alignment, producing level shoulders, correcting the deformity, and saving motion segments. Failure to select proper fusion levels may lead to coronal imbalance, sagittal junctional problems, and even revision surgery (Chap. 16).

19.2 Imaging

Before learning to select fusion levels, some radiographic parameters should be clarified (Video 19.6). End vertebra (EV) is defined as the vertebra that is most tilted from the horizontal apical vertebra. Substantially touched vertebra (STV) is defined as the most proximal lumbar vertebra where the central sacral vertical line (CSVL) either intersected the pedicle outline or was medial to the pedicle outline [1].

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19.3 Treatment Options

The first step in planning the fusion levels (Videos 19.3 and 19.6) is determining which curve should be fused, especially in Lenke 1C and Lenke 5C AIS patients with compensatory curves (Appendix I). Selective fusion refers to fusing only the major (structural) curve, and the compensatory curve could experience a spontaneous correction. Non-selective fusion would fuse both structural and compensatory curves. Selective fusion for Lenke 1C AIS would bring lumbar motion preservation but a less corrected lumbar curve. It is still unknown which of the following would be a better clinical outcome: an unfused lumbar spine with moderate residual curvature or a straight, partially fused lumbar spine.

It is generally agreed that a spinal fusion should be as short as possible. Some authors have observed a correlation between distal fusion length and severity of back pain and disk degeneration. Thus, selection of the lowest instrumented vertebra (LIV) is critical in the surgical treatment of AIS. In the classic article, King et al. recommended stable vertebra (the distal vertebra most closely bisected by the CSVL) as LIV (Appendix H). With the modern Lenke classification, Lenke 1A/B and 2A/B curves in which the lumbar curve does not cross the CSVL could select STV as LIV, saving one fusion level compared to those who were fused to SV (Appendix I) (Fig. 19.1). For Lenke 1C AIS patients, the surgeons should

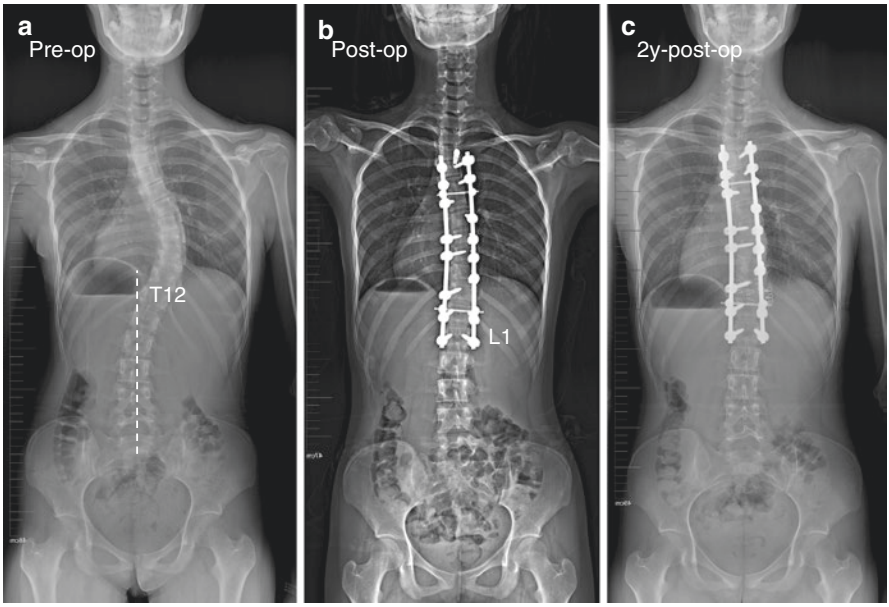

Fig. 19.1 This is a Lenke 1A AIS patient, the CSVL touched L1 vertebra (**a**); according to the Touching vertebra principal, L1 was selected as LIV (**b**); at 2-year follow-up, no distal complication was found (**c**)

communicate with patients and their families at first regrading to choosing selective fusion or not. Selective fusion will leave a curved but mobile lumbar curve; the patients also need to face the risk of adding-on or coronal decompensation. Thus, surgeons should be careful when they apply selective fusions to the patients who are at the preadolescent period and/or with a Risser grade 0–1 (Chap. 17). Some surgeons specifically suggested under-correcting the thoracic curve to match the residual lumbar curve, or to leave the LIV tilted after selective thoracic fusion.

In Lenke curve types in which the lumbar curve is to be included in the fusion, including Lenke 3C, 4C, 5C, 6C, and non-selective fusion for Lenke 1C/2C, the selection of LIV should consider more about the flexibility of the disk. The disk below the proposed LIV should be reversed or neutralized on convex bending radiographs, and the vertebra below the proposed LIV has a rotation of $\leq 15°$ in the transverse plane on convex bending films. The LIV is usually one disk cephalad to the EV of the lumbar curve.

For the selection of upper instrumented vertebra (UIV), motion preservation is less important since the thoracic spine is more rigid than the lumbar spine and the rib cage could help to stabilize the thoracic spine. The primary concern for UIV selection is the post-operative shoulder balance, especially in Lenke 2 AIS patients. In Lenke 1 patients, the proximal thoracic (PT) curve is non-rigid and could be left unfused. It should be noted that the fusion of the main thoracic curve alone would elevate the left shoulder since almost every thoracic AIS patient has a right curve. Thus, Lenke et al. proposed an easy method to determine UIV for Lenke 1/2 AIS patients: if the patient shows elevated left shoulder before surgery, the fusion should extend to T2; if the baseline shoulder is balanced, the UIV could be T3; if the patient shows elevated right shoulder, the UIV should be T4 or below, leaving PT unfused. Ilharreborde et al. recommended fusion of structural PT curves and of nonstructural PT curves if T1 tilt and shoulder balance are in the same direction and would be worsened with correction of the main curve. If they are in the opposite direction, then partial inclusion of the thoracic curve (T2 or T3) is possible [2] (Fig. 19.2).

Cheung KM and colleagues proposed a method for decision-making in selecting fusion levels in thoracic AIS patients using fulcrum bending radiographic (FBR) [3]. FBR can very closely predict the surgical outcome of posterior correction for thoracic curves. The method of selecting fusion levels in thoracic AIS using FBR is as follows: (1) determine the estimated UIV and LIV; (2) draw a line parallel to the inferior endplate of the estimated LIV on the FBR, and draw a line perpendicular (central line, CL) to the above line from the center of the LIV; (3) measure the Cobb angle between the estimated UIV and LIV on FBR; the ideal angle should be smaller than 20°; (4) measure the shift of the UIV from the CL; this distance should be smaller than 20 mm; (5) if the shift from the UIV was greater than 20 mm from the CL, the next caudal vertebra was chosen as the LIV and these were the selected levels for instrumentation; if the shift was less than 20 mm and the Cobb angle was greater than 20°, then the next cranial vertebra was chosen as the estimated UIV.

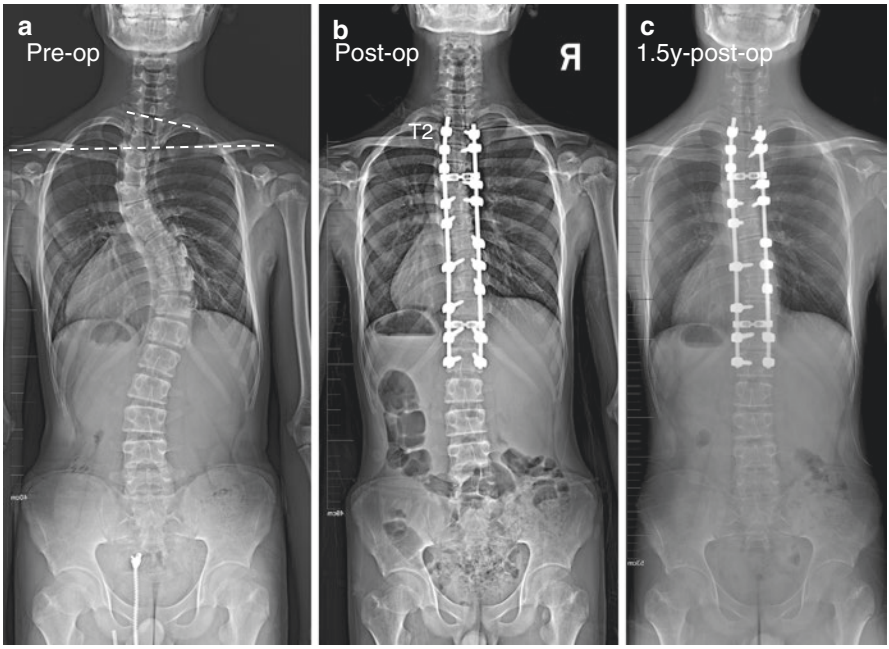

Fig. 19.2 This is a Lenke 2A AIS patient; T1 tilt and the shoulder balance was in the opposite direction (**a**), so it is not necessary to fuse up to T1; we selected T2 as UIV (**b**); after 1.5 years, no shoulder imbalance was found (**c**)

For Lenke 5C AIS patients, selective lumbar fusion is more common than non-selective fusion. Traditionally, fusion level extends from the upper-end vertebra (UEV) to the lower end vertebra (LEV) of the main curve (Cobb-to-Cobb fusion) in both anterior and posterior fusion. Dubory et al. reported the equivalent clinical outcome of Cobb-1 to Cobb anterior fusion (UIV at one level caudal to UEV and LIV at LEV) compared to Cobb to Cobb fusion [4]. Shu et al. recently reported a hyper-selective fusion strategy (Cobb-1 to Cobb posterior fusion) is a reasonable alternative to conventional fusion strategy in the treatment of Lenke 5C AIS (Fig. 19.3). They recommended that the Cobb-1 to Cobb posterior fusion could be performed in Lenke 5C patients with Risser more than grade 2 and with a thoracic compensatory curve over 15° [5]. It has been shown that thoracic curves did not progress after selective anterior fusion in Lenke 5C curves [6].

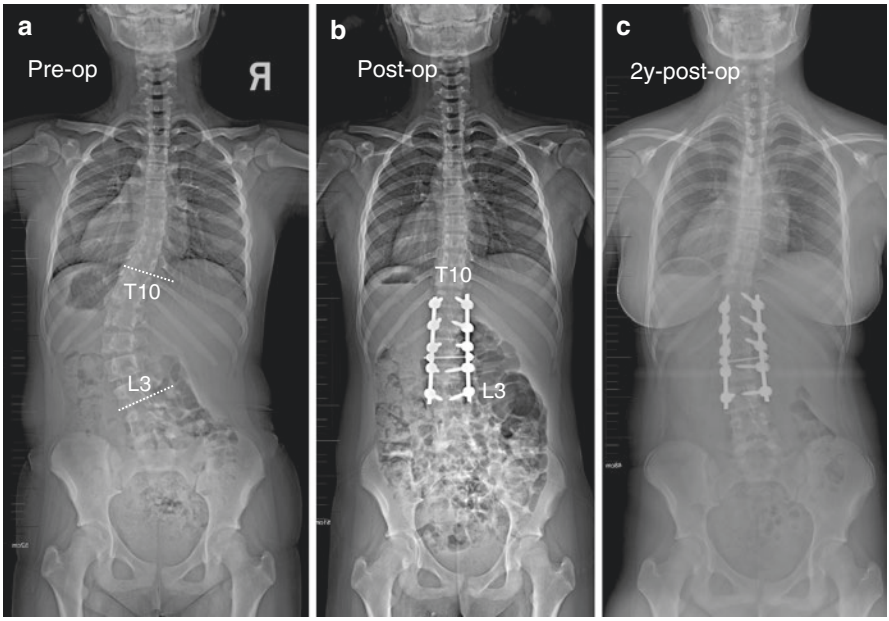

Fig. 19.3 This is a Lenke 5C AIS patient; the main curve was from T10 (upper EV) to L3 (lower EV) (**a**); according to the "Cobb to Cobb" principal, UIV should be T10; however, we selected T11 as UIV following the "Cobb-1 to Cobb" fusion principal (**b**); at 2-year follow-up, coronal balance was well maintained (**c**)

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Late Onset Neuromuscular Scoliosis

20

Federico Canavese

20.1 Definition

Children with neuromuscular disorders affecting the neuro-musculoskeletal system frequently develop progressive spinal deformities, also known as neuromuscular scoliosis (NMS); in such patients, respiratory compromise is common.

20.2 Natural History

Patients with neuromuscular disorders tend to develop scoliosis at younger ages than patients with idiopathic scoliosis; a large proportion of NMS are progressive and often non-responsive to orthotic management even though conservative treatment during infancy is important to control, at least partially, deformity progression. Unlike idiopathic scoliosis, NMS can progress beyond skeletal maturity, particularly in wheelchair-bound patients, and be associated with pelvic obliquity. Also, NMS is often characterized by a greater decrease in lung volumes compared to idiopathic curves, which, in contrast, have normal muscle function (Chaps. 14 and 15).

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20.3 Physical Examination

NMS is characterized by long collapsing C-shaped thoracolumbar and lumbar curves, pelvic obliquity, and changes in sagittal plane alignment that can affect sitting balance and cardio-respiratory function. In most severe cases, patients with NMS can develop painful impingement between concave ribs and the same side iliac crest.

Pulmonary, neurologic (Video 20.4), genitourinary, nutritional, and gastroenterological comorbidities are common in patients with NMS and must be managed (pre- and post-operatively) by a multidisciplinary team of care providers. A multidisciplinary approach is the key to a successful outcome.

20.4 Imaging

Full standard anterior-posterior (AP) and lateral radiographs of the spine (standing or supine) allow locating and measuring the magnitude of the curve (Video 20.6), measuring the amount of pelvic obliquity, and evaluating the changes in the sagittal plane; long C-shaped thoracolumbar, with associated thoracolumbar kyphosis, and lumbar curves are common. Lateral bending films and fulcrum radiographs are useful to assess the reducibility of the curve (Figs. 20.1 and 20.2).

Magnetic resonance imaging (MRI) of the whole spine is useful to rule spinal cord abnormalities and spinal dysraphism (Chap. 31).

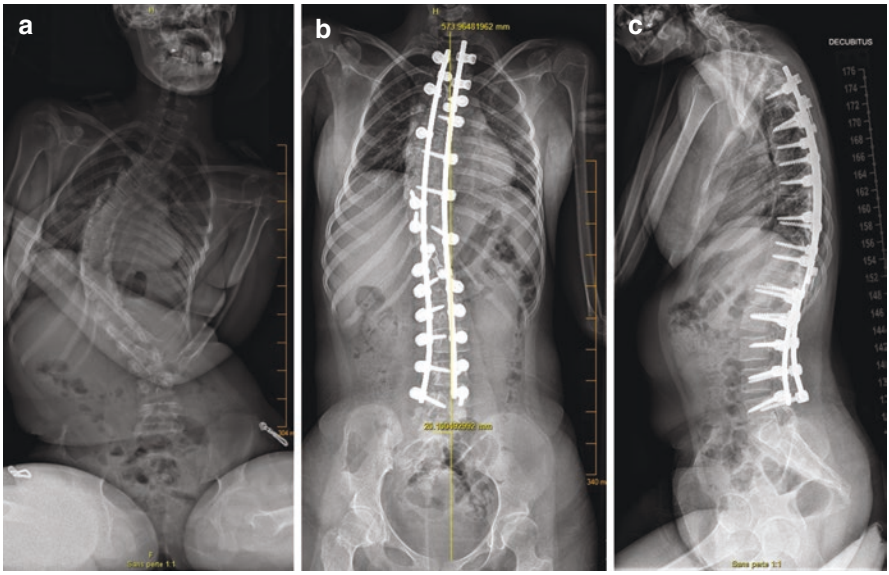

Fig. 20.1 Patients with cerebral palsy (GMFCS IV; **a**); instrumentation stopped at L5 (even pelvis; **b** and **c**)

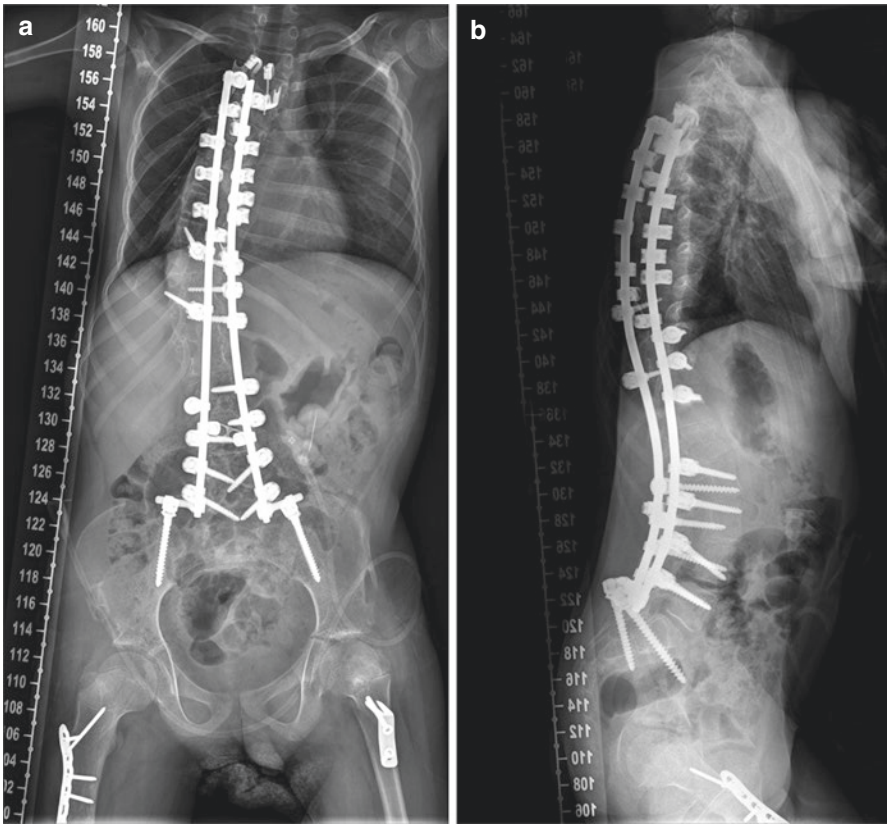

Fig. 20.2 Patient with cerebral palsy (GMFCS V); instrumentation included the pelvis (uneven pelvis; **a** and **b**)

A computed tomography scan of the spine (with or without 3D reconstruction) is indicated to study spine, pelvis, and thoracic cage anatomy.

Clinical pictures of the patient before and after surgical treatment are also important.

20.5 Differential Diagnosis

Differential diagnosis is important in the context of the underlying neuromuscular pathology. Conditions associated with NMS include cerebral palsy, Duchenne muscular dystrophy, spinal muscular atrophy, Friedreich's ataxia, spina bifida, and several genetic syndromes (e.g., arthrogryposis, osteogenesis imperfecta, Larsen syndrome) (Chaps. 14 and 15).

20.6 Treatment Options

In patients with NMS, bracing is usually not effective, and surgery is the primary treatment option (Videos 20.1 and 20.4). The indications for surgery are (1) a significant curve (exceeding 50°) resulting in functional disturbance, uncomfortable sitting (pelvic obliquity), and/or cardio-respiratory compromise; (2) a progressive spinal deformity not controllable with orthosis; (3) a small curve with inevitable progression; and (4) painful deformities.

Patients with neuromuscular disorders have many similarities in curve patterns, despite different etiologies of the main disease; therefore, similar strategies and common goals are applied to patients with NMS. The goals of surgical treatment are: (1) to prevent curve progression; (2) to maintain the spine balanced on the coronal and sagittal planes, with a level and upright trunk position; (3) to provide a balanced and comfortable sitting position to reduce repositioning; (4) to reduce pain; (5) to reduce the discomfort caused by the impingement of the ribs against the iliac crest on the concave side of the curve; (6) to maximize patients' health and function; and (7) to maintain walking ability in ambulatory patients.

Surgery can be challenging due to the presence of comorbidities, the need for longer fusions, the frequent need for fusion to the pelvis, the increased bleeding, the longer operative time, the relatively poor bone quality stock, and the higher complication rate.

Luque rods, or variations on the Luque technique, often remain the preferred instrumentation for neuromuscular curves; pedicle screws, hooks, and/or sublaminar polyester bands can also be used (Videos 20.3 and 20.7).

Instrumentation and fusion should be extended to the pelvis in non-ambulatory patients with pelvic obliquity. In contrast, instrumented fusion can stop at L5 or above when the patient is still ambulatory and shows minimal or no signs of pelvic obliquity. Small amounts of pelvic obliquity (less than 10° to 15°) are compatible with comfortable sitting. In contrast, larger fixed obliquities are not compatible with comfortable sitting and must be corrected surgically or, if not fixed, with wheelchair modifications.

20.7 Expected Outcomes

The success of treatment depends on the maintenance of a balanced spine on the coronal and sagittal planes over a level pelvis.

In children with NMS, puberty can begin earlier or, more frequently, later than the puberty of children with idiopathic scoliosis. Depending on the neuromuscular disease, the rate of progression of the scoliotic deformity during pubertal growth spurt can increase by 2°–4° per month, especially in patients who are wheelchair-bound. NMS continues to progress beyond skeletal maturity at a rate of approximately 1°–4° per year if the curvature is greater than 50° at the end of growth, compared to approximately 0.5°–1° per year for curves of less than 50°. The

risk-to-benefit ratio is an important parameter that must be considered before surgery as the results can be gratifying if patients are properly selected.

20.8 Potential Complications

The postoperative complication rate is much higher (approximately 30%) in patients with NMS, compared to patients with idiopathic scoliosis.

Early postoperative complications include deep spine infections (risk 10–20 times higher than in idiopathic scoliosis), cardio-respiratory, neurologic and nutritional issues, prolonged ileus, constipation, fluid overload, skin breakdown, bleeding, and death. Late postoperative complications include chronic infections, non-union, coccygodynia, crankshaft phenomenon, implant-related issues, loss of correction, and inadequate correction.

20.9 What Should Patient and Family Know?

Although spinal surgery can restore proper spinal alignment, it has some potential disadvantages. In particular, spinal fusion and instrumentation can adversely affect those patients with neuromuscular disorders who have developed functional compensation techniques requiring a short and mobile trunk. Moreover, surgery stops any further growth over the fused segments, and it can accentuate hip deformity.

In patients with neuromuscular disease, the likelihood and severity of scoliosis increase with the degree of neuromuscular involvement.

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Postural Kyphosis

21

Federico Canavese and Alpaslan Şenköylü

21.1 Definition

Postural kyphosis (PK) is the most common type of increased thoracic kyphosis (greater than 40°). It is a benign condition characterized by an abnormal rounding of the thoracic spine due to the lack of rigidity in the spine, and the lack of structural abnormalities of the vertebral bodies on plain radiographs.

PK is reducible (flexible deformity) by the patient (active correction) or by orthopedic maneuvers (passive correction).

21.2 Natural History

Patients not improving their posture can develop persistent back pain, and rigid kyphosis; the concern is mainly esthetic.

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21.3 Physical Examination

PK often occurs in adolescents and it can be the result of poor posture and/or muscle weakness. Clinical examination reveals abnormal rounding of the thoracic spine (hyper-kyphosis); the deformity is flexible (Fig. 21.1a, b). A neurological evaluation includes an assessment of pain, numbness, paresthesia, extremity sensation and motor function, muscle spasm, weakness, and bowel/bladder changes (Video 21.4).

21.4 Imaging

The radiographic diagnosis of PK is straightforward. Usually, imaging is not needed. However, standard anterior-posterior and lateral radiograph of the spine allows investigating the structure of the vertebrae (vertebral wedging, end-plate irregularities, disk height) and assessing the overall sagittal alignment of the spine (Video 21.6). To evaluate curve flexibility, radiographs may be taken with the patient supine (positive response to positional changes).

Complementary investigations, such as magnetic resonance imaging (MRI) and computed tomography scan of the spine, are rarely needed unless different etiology is suspected (presence of warning signs (*red flags*; Chap. 11)).

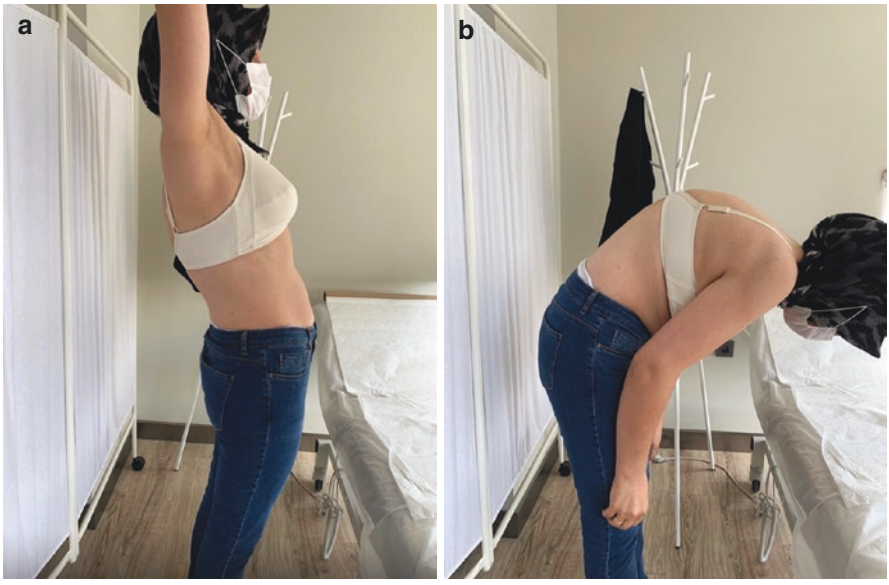

Fig. 21.1 A patient's hyperkyphosis (**a**) can be easily corrected with hyperextension maneuver (**b**). This is not possible for the Scheuermann's kyphosis

21.5 Differential Diagnosis

The other causes of abnormal kyphosis are Scheuermann's kyphosis (Chap. 22), congenital kyphosis (Chap. 24), kyphosis associated with achondroplasia or neuromuscular disorders (Chap. 20), post-traumatic kyphosis (Chaps. 4 and 9), and kyphosis post-laminectomy; moreover, PK is among the causes of back pain in children and adolescents (Chap. 11).

Most of the above-mentioned pathologies are rigid whereas PK is quite flexible (Fig. 21.1a, b).

21.6 Treatment Options

The treatment of PK in children aims to prevent the curve from worsening and to restore normal posture. The goal of physical therapy is to strengthen the back and abdominal muscles in order to share the load bearer by the spine, improve posture, and reduce discomfort/pain. Some adolescents are resistant to perform exercises given by physiotherapists. In this case, the patient should be encouraged to play sports regularly that he/she enjoys. Especially sport activities such as Pilates, yoga, swimming, and dancing are useful to strengthen paravertebral muscles and help to correct the posture. The key is to choose the type of activity by the patient him/herself since motivation is the most important variable for continuing the workout.

In skeletally immature patients with more severe deformity and significant amount of remaining growth (Risser 0–2) (Appendix M), a brace is indicated to support the spine growing into the correct posture (good sagittal alignment). A Milwaukee-type cervico-thoraco-lumbo-sacral brace is the most common orthosis in patients with a thoracic hyper-kyphosis, and it is usually very effective. However, the most substantial drawback of the Milwaukee brace is poor patient compliance due to self-appearance problems.

Nonsteroidal anti-inflammatory drugs or pain-killers can help to reduce any discomfort related to abnormal kyphosis.

21.7 Expected Outcomes

Most patients with PK can achieve a good outcome (correction of the kyphosis to within the normal range) with education in proper posture and participation in an exercise program (strengthening of the back and abdominal muscles to reduce discomfort and improve posture).

21.8 Potential Complications

PK is a benign condition. The development of rigid kyphosis and persistent pain not controlled with medication are potential complications related to poor posture and/or lack of treatment. Breathing difficulties and neurological symptoms (numbness

or weakness in the extremities, problems with balance, loss of normal bladder or bowel control) are possible, although rare, complications; if neurological symptoms are detected, MRI of the whole spine is mandatory (Chap. 11).

21.9 What Should Patient and Family Know?

PK can be prevented by maintaining good posture. Parents should encourage their child to avoid slouching, to sit correctly, to avoid carrying heavyweights, and to take regular physical activities, including breathing exercises. But the behavior of the parents should be more supportive rather than commanding. Backpacks are also another biomechanical factor often implicated as a reason for back pain and poor posture in children and adolescents. Although there is no convincing evidence that supports this hypothesis, children should use school bags properly by arranging straps and putting both handles evenly to their shoulders.

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Scheuermann's Disease

22

Alpaslan Şenköylü

22.1 Definition

Scheuermann's disease (SD; also known as Scheuermann's kyphosis) is the rigid hyper-kyphosis of the thoracic or thoracolumbar region due to multilevel vertebral wedging which occurs during the adolescent period (Fig. 22.1). According to the Scoliosis Research Society, hyper-kyphosis is defined by a sagittal Cobb's angle measurement between T1 and T12 exceeding 45° (Video 22.6). The etiology of SD is not fully known and several pathogenetic mechanisms have been hypothesized, including genetics, hormonal, biomechanical, and environmental factors. Histopathological studies revealed a disorganization of vertebral endplate due to the modification of the collagen/proteoglycan ratio at affected levels. This structural change causes a growth disturbance and subsequent anterior vertebral wedging (anterior column). SD is not a rare disorder with an incidence ranging between 1 and 8% of the general population. However, mild and moderate cases can be neglected through a perfect compensation mechanism of the spinal column which develops a compensatory increase of cervical and/or lumbar lordosis.

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22.2 Natural History

SD is a benign condition in most patients. Disabling pain can be seen in about one-third of adolescents although it tends to get frequently better during adult life. Cardio-pulmonary problems may develop in severe kyphotic patients with sagittal Cobb's angle measurement exceeding 100° (Video 22.6). Neurologic complications of SD are not common.

22.3 Physical Examination

SD typically presents by the painful, sharp, and rigid dorsal hyper-kyphosis during adolescence (Fig. 22.1). Although pain is generally located at the apex of the kyphosis, it can also be seen at lumbar and interscapular regions due to the paravertebral muscle contracture. Spondylolysis should be kept in mind in patients complaining of low back pain (Chap. 11). The apex of the kyphosis can be thoracic or thoracolumbar. As mild scoliosis can commonly be associated with SD, patients may have a shoulder imbalance and/or a rib hump with positive Adam's forward bending test (Chap. 16). The patient, generally, compensates the excessive thoracic hyper-kyphosis with cervical and lumbar hyper-lordosis. Hamstring and iliopsoas contracture are frequent findings.

Neurologic examination including motor and deep tendon reflex evaluation is essential even though neurologic problems are rare in SD (Video 22.4).

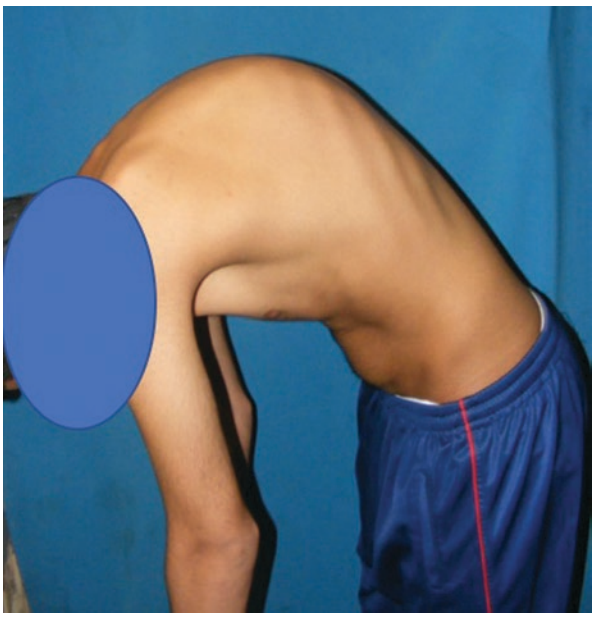

Fig. 22.1 Clinical picture of an adolescent patient with a sharp-angled hyper-kyphosis (forward bending position)

22.4 Radiographic Examination Including Advanced Imaging and Classification

Standard anterior-posterior and lateral long cassette radiographs should be requested in patients with SD (Fig. 22.2). Anterior wedging more than 5° in three or more adjacent vertebrae on lateral radiographs is the characteristic finding of SD as defined by the Sorensen, although one level of wedging has been appreciated enough for the diagnosis according to recent studies. Associated findings are Schmorl's nodules (Chap. 27), irregular endplates, and decreased disc height (Fig. 22.3). However, these findings have low specificity and Sorensen's criteria are enough to make the correct diagnosis. Coronal Cobb angle (Video 22.6) should be measured if there is associated scoliosis which can be found in about one-third of patients with SD. Fulcrum extension radiograph is a useful technique for the evaluation of the kyphosis flexibility during preoperative planning. The fulcrum is applied at the apex of the kyphosis with the patient in the supine position, and a lateral radiograph of the spine is taken (Fig. 22.3).

Magnetic resonance imaging (MRI) should be performed in order to rule out a possible thoracic disc herniation that may induce neurological symptoms during surgery (correction of the hyper-kyphosis with an extension maneuver).

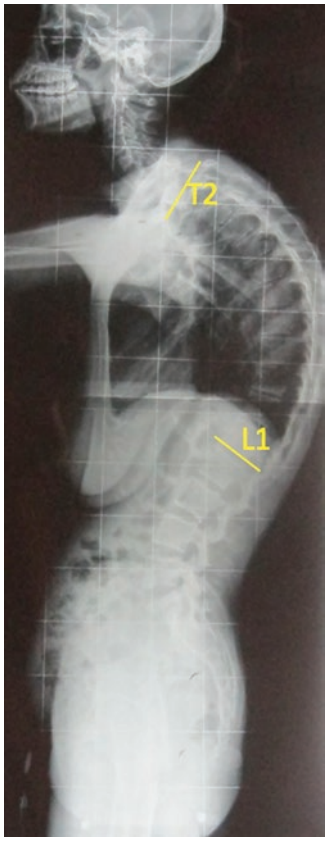

Fig. 22.2 Full spine radiograph of a 17-year-old girl complaining of pain and deformity; sagittal Cobb's angle measurement (T2-L1) is 92°

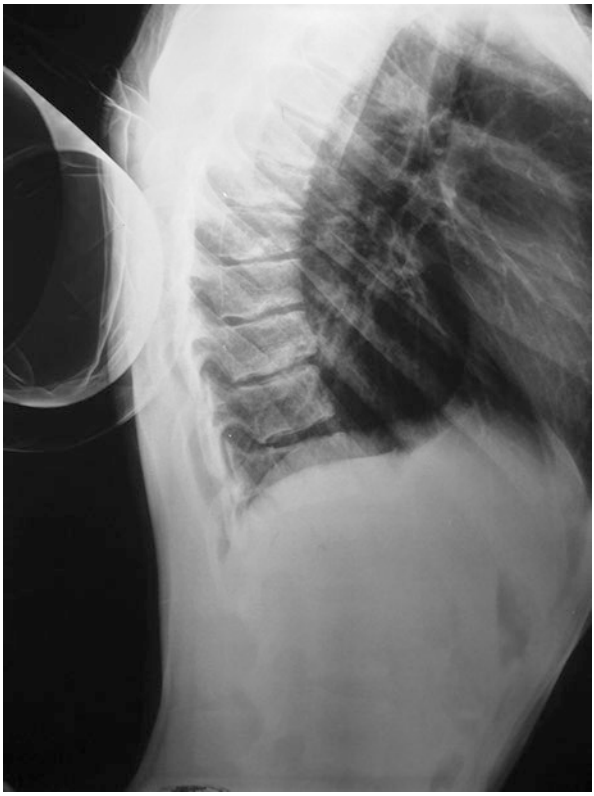

Fig. 22.3 Fulcrum extension radiograph. This radiograph shows typical findings of SD, including >5° of anterior wedging of three consecutive vertebrae, Schmorl's nodules and endplate irregularities

There are two types of SD: type-1 has the typical curve pattern with the apex located between T6 and T8. On the other hand, in type-2 SD the apex is located at the thoracolumbar junction; type-2 SD is more likely to progress and to become symptomatic.

22.5 Differential Diagnosis

Postural kyphosis (Chap. 21) or adolescent round-back is another common sagittal plane deformity that can easily be confused with SD; postural kyphosis is also seen during adolescence. However, it is more flexible, more round-shaped, without any morphological changes such as wedging of the vertebrae and Schmorl's nodules. Pain is relatively uncommon in postural kyphosis.

Other causes of thoracic hyper-kyphosis should be excluded, including old compression fractures (Chap. 7), kyphosis post-laminectomy, ankylosing spondylitis (Chap. 54), destructive lesions such as tumor, and infection (Chap. 52).

22.6 Treatment Options

Non-operative treatment including back exercises and brace can be applied to symptomatic patients with sagittal Cobb's angle measurement between 45 and 75°. There is no evidence regarding the improvement of the kyphosis with exercises. However, pain relief is possible with an effective exercise program, and it should be prescribed to all patients with pain a chief complaint. Brace treatment should be reserved to growing patients (Risser 0–3) as it generally fails if Risser sign is grade 4 or 5. Thoraco-lumbo-sacral orthosis (TLSO) is indicated if the apex of the kyphosis is below T8, and it is most effective in type-2 SD deformity. In type-1 patients, Milwaukee brace should be prescribed to type-1 SD patients although its efficiency is somehow reduced due to inadequate patient compliance.

Operative treatment is indicated in patients with pain and/or unacceptable cosmesis, with sagittal Cobb angle exceeding 75° and unresponsive to conservative treatment; however, surgery must be deeply discussed with patients and family because of the extension of the instrumented fusion (very often from T2 to L3) (Videos 22.3 and 22.6). Neurologic complications are exceptional in SD and this is a straightforward indication for the surgical treatment. Surgical planning should be done after having requested an MRI to exclude a possible thoracic disc herniation and fulcrum extension radiographs to assess the flexibility of the deformity. Posterior correction, instrumentation with pedicle screws and fusion is the standard surgical technique (Video 22.3). Multimodal intraoperative neuromonitoring must be performed during surgery to avoid neurological complications. Schwab type-2 osteotomies which provide posterior column shortening can be done for rigid curves (Fig. 22.4). Proper fusion level selection and avoidance of overcorrection are important points to prevent junctional problems. The most cranial end vertebra within the Cobb angle should be included in the fusion; the sagittal stable vertebra (proximal vertebral body touched by the vertical line drawn from the posterior superior corner of the sacrum) should be identified to select the distal end of the fusion as well as the disc space perpendicular to the horizontal plane on fulcrum lateral radiograph.

22.7 Expected Outcomes

Pain relief can be achieved in a significant number of patients by physiotherapy, and it should be given as the first step of the non-surgical treatment. Surgical treatment is an effective method for a satisfactory correction and prevention of curve progression though it stiffness almost the whole spine.

22.8 Potential Complications

SD itself can be complicated with chronic back pain, poor cosmesis, and pulmonary function problems. Possible surgical complications are proximal or distal junctional kyphosis, wound infection, pseudoarthrosis, neurological deficit, and superior mesenteric artery syndrome.

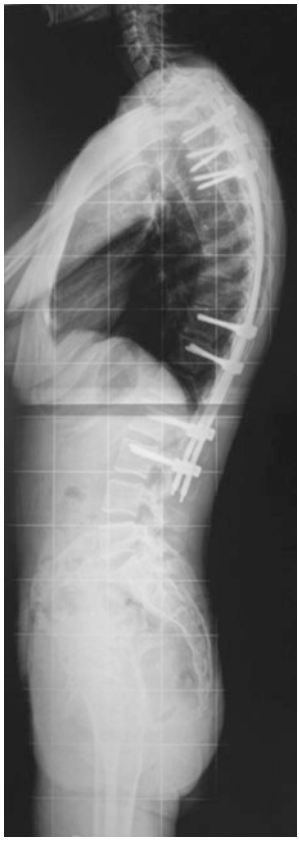

Fig. 22.4 Postoperative lateral radiographs of the patient shown in Fig. 22.2. Correction was achieved with three levels Schwab type-2 osteotomies at the apex of the deformity

22.9 What Should Patient and Family Know?

SD appears during adolescence and it is characterized by pain and poor posture. Pain can occasionally affect daily life and professional activities. Neurologic complications are rare during the natural course of disease.

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Cervical Kyphosis in Neurofibromatosis Type I

23

Federico Canavese

23.1 Definition

Children with neurofibromatosis type I (NF-1) can develop cervical kyphosis (CK). Severe CK in the setting of NF-1 is a rare manifestation of the disease in the pediatric population, and it is often associated with vertebral dysplasia.

23.2 Natural History

The natural history of patients with CK secondary to NF-1 is not well known due to the rarity of the condition within this patient's population.

23.3 Physical Examination

Clinical examination reveals a short and hypo-mobile neck. Symptomatic patients with CK can experience pain or neurological deficits although some of them may have a surprisingly high tolerance for deformity with few symptoms even if spondyloptosis (dislocation of one vertebral body over another) is present.

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A neurological evaluation includes an assessment of pain, numbness, paresthesia, extremity sensation and motor function, muscle spasm, weakness, gait disturbance, and bowel/bladder changes (Video 23.4).

23.4 Imaging

The radiographic diagnosis of CK is straightforward. Lateral radiographs of the cervical spine show severe CK with dystrophic changes of the vertebrae (Fig. 23.1). The kyphosis can be at the upper or at the sub-axial cervical spine, and it is always rigid in flexion-extension (Video 23.6). Characteristic dystrophic changes are scalloping of the posterior vertebral margins, spinal canal widening, enlarged neural foramina, defective pedicles, and spindling of the transverse processes.

Computed tomography (CT) scan provides useful information about the osseous and three-dimensional anatomy of the cervical spine, and it allows estimating the severity of dystrophic changes in the cervical spine.

Magnetic resonance imaging (MRI) helps to obtain information on soft tissues and the spinal cord. In particular, MRI can detect coexisting paraspinal and spinal cord tumors and can evaluate the anatomy of the spinal canal and its relationship with the cord, the status of the spinal cord, and the presence of dural ectasia (Fig. 23.1). It is important to stress out that MRI must include the brain and the whole spine, including the sacrum.

It is possible that the patients with NF-1 may have an aneurysm at the vertebral arteries. Thus, before starting traction, patients should be evaluated by MR

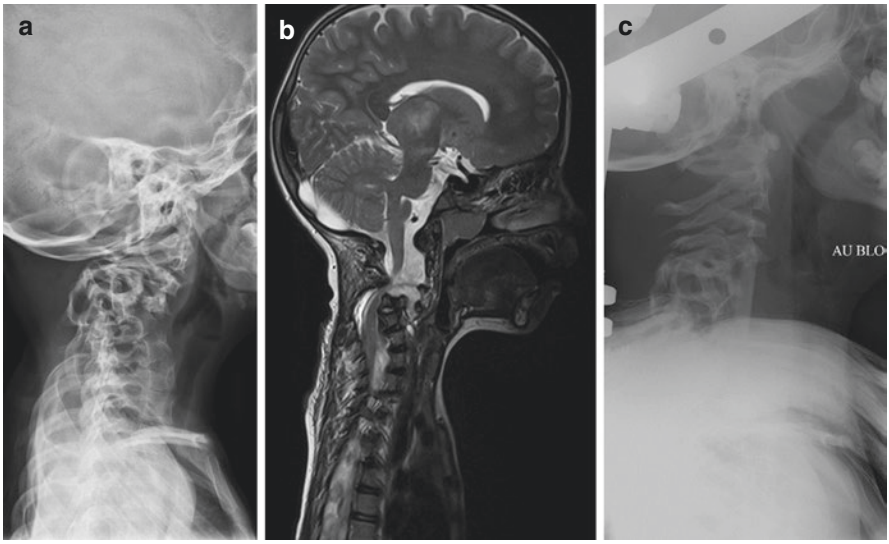

Fig. 23.1 A patient with NF-1 and cervical kyphosis (**a**); MRI shows kinking of the cord and dural ectasia (**b**); anterior strut graft (tibia; **c**)

angiography or contrast-enhanced CT to rule out an aneurysm at the vertebral arteries due to the risk of rupture during traction.

23.5 Differential Diagnosis

None.

23.6 Treatment Options

Patients with progressive deformity or symptoms should be offered surgery; combined anterior-posterior fusion is indicated in most skeletally immature NF-1 patients with severe CK. Preoperative halo-gravity traction or halo-vest (Video 23.1) is useful to achieve a gradual, although partial, and safe correction of the CK. In patients with NF-1 smaller traction forces should be used because of the fragility of the cervical spine due to dystrophic changes.

Fusion can be associated with vertebral osteotomy to restore the sagittal alignment of the cervical spine (20% to 25% risk of new neurologic deficits), or it can be achieved "in-situ" with an anterior (strut graft) and posterior bone graft (Fig. 23.1). Halo should be maintained postoperatively up to 10 to 12 weeks as it is possible that the anterior strut graft can partially dislodge at halo-removal (Videos 23.1, 23.2, 23.3 and 24.4).

23.7 Expected Outcomes

Significant radiographic correction can be achieved with corrective surgery although it carries a relatively high risk of new neurologic injury. On the other hand, fusion "in-situ" does not attempt to correct the deformity above the correction obtained with traction, and it has almost no risk of new neurological injury. Whatever the technique, erosion of bone usually continues after surgery and bone graft can get resorbed leading to revision surgery with new bone graft application (posteriorly in most cases, though anterior surgery may be necessary too).

23.8 Potential Complications

The onset of pain and neurologic symptoms are possible. The presence of coexisting paraspinal and spinal cord tumors increases the risk of hemorrhaging and spinal cord injury during corrective surgery. Post-operative hematoma, causing breathing difficulties by compressing the airways, is possible and therefore patients should be monitored carefully.

Malignant transformation of neurofibroma into neurofibrosarcoma (nerve sheath tumors) is possible throughout the whole life of the patient (Chap. 59).

23.9 What Should Patient and Family Know?

Close follow-up is extremely important because complications and deformity progression are frequent.

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Congenital Kyphosis

24

Michael Ruf

24.1 Definition

Congenital kyphosis is a sagittal plane deformity caused by malformations taking place during the early embryological period (Chap. 12). Posterior hemivertebra (failure of formation) leads to short angulated kyphotic deformity while anterior bar (failure of segmentation) leads to anterior vertebral body coalition. As a consequence, the anterior and posterior growth of the spine is not synchronous (unbalanced; posterior > anterior) and induces progressive kyphosis. Mixed deformities are frequent. A further cause of congenital kyphosis is the hereditary absence or weakness of the posterior structures of the spine (failure of the posterior tension band).

24.2 Natural History

Congenital kyphosis and kyphoscoliosis usually tend to progress rapidly, resulting in severe deformity (2.5°-5°/year in deformity secondary to failure of formation, more rapid in mixed type deformity). Progression of the curve accelerates during the adolescent growth spurt while it slows down at skeletal maturity [1]. The thoracolumbar (T-L) junction is the most affected spinal segment; in particular, increased kyphosis at the T-L junction leads to thoracic hypo-kyphosis and lumbar hyper-lordosis needed to maintain trunk balance.

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Congenital kyphosis has a much higher risk to develop neurological deficits compared to congenital scoliosis: the short angulation acts as a hypomochlion impairing the spinal cord over time resulting in myelopathy. It is important to stress out that patients with congenitally dislocated spine (CDS) can develop severe neurologic symptoms following minor trauma; a proportion of patients with CDS may injure the spinal cord in utero.

24.3 Physical Examination

Physical examination includes documentation of

- Location and severity of the kyphosis.
- Tenderness on palpation/percussion.
- Compensatory lordotic spinal segments.
- Trunk balance (plumbline).
- Flexibility of the kyphotic segment (active and passive correction).
- Neurological examination (Video 24.4); if necessary, the realization of motor-evoked (MEP) and somatosensory-evoked potentials (SSEP) is recommended.
- Pulmonary function test.
- Clinical pictures of the patient.

24.4 Imaging

Full-length standing anterior-posterior and lateral radiographs of the spine (including the hips) demonstrate the kyphotic deformity as well as the compensatory changes (thoracic hypo-kyphosis and lumbar hyper-lordosis) (Video 24.6). Lateral radiographs allow measurement of the local kyphosis due to the malformation as well as the total thoracic kyphosis, the lumbar and cervical lordosis, the pelvic incidence and pelvic tilt, the sacral slope, and the plumbline. Lateral radiographs with a fulcrum at the apex of the kyphosis are useful to evaluate the flexibility of the deformity.

Magnetic resonance imaging is mandatory to rule out compression of the spinal cord as well as signs of myelopathy.

Computed tomography scan with three-dimensional reconstruction is important to evaluate the anatomy and the location of the malformation.

24.5 Treatment Options

Early treatment is required, as congenital kyphosis is usually progressive and the risk of neurologic impairment is high. Conservative treatment is not effective: neither a brace, or cast, nor physiotherapy can stop the progression of the deformity;

delay in treatment will allow progression of the deformity and of the compensatory curves.

Surgery is the treatment of choice. It should be performed early in order to avoid rigid deformity as well as neurologic impairment.

Surgical options include posterior-anterior and single-posterior procedures. Anterior-only correction without posterior instrumentation will result in progressive kyphosis during growth.

In the case of the posterior hemivertebra causing kyphosis, the abnormal vertebra is resected through a posterior approach. The adjacent discs are removed, and an anterior spacer/cage is inserted. Stabilization and correction are achieved by posterior compression instrumentation (Chap. 12); the anterior bar can be resected from posterior or an additional anterior approach is performed (Fig. 24.1).

The apex of the congenital deformity tends to fuse over time. Thus, delayed treatment is more challenging, requiring an osteotomy to achieve correction. A posterior wedge osteotomy or vertebral column resection can usually be performed via a single-posterior approach, since the apex of the deformity is shifted backward [2, 3]. An anterior approach may be necessary in revision cases or in case of abnormal course of the great vessels. The spinal cord has to be handled with extreme care; the use of intraoperative monitoring is strongly recommended. Tension or translation must be avoided. Posterior instrumentation in combination with stable anterior support provides safe and reliable stability (Fig. 24.2) (Videos 24.1 and 24.3).

24.6 Expected Outcomes

Without corrective surgery, reduction of pulmonary function and progressive neurologic compromise have to be expected without treatment in most cases. Early treatment can restore a physiological sagittal profile, thus avoiding the development of neurologic impairment. Delayed treatment requires more extensive surgery, with a higher risk for neurological deficits and poorer outcomes with respect to trunk balance.

24.7 Potential Complications

Neurological risk is substantial in severe cases requiring extensive osteotomies for correction, especially in combination with pre-existing myelopathy, or in revision cases.

24.8 What Should Patient and Family Know?

Congenital kyphosis is a serious disorder with a potentially dramatic outcome. In mild deformity, regular follow-up is needed; in case of progression, early surgery is recommended.

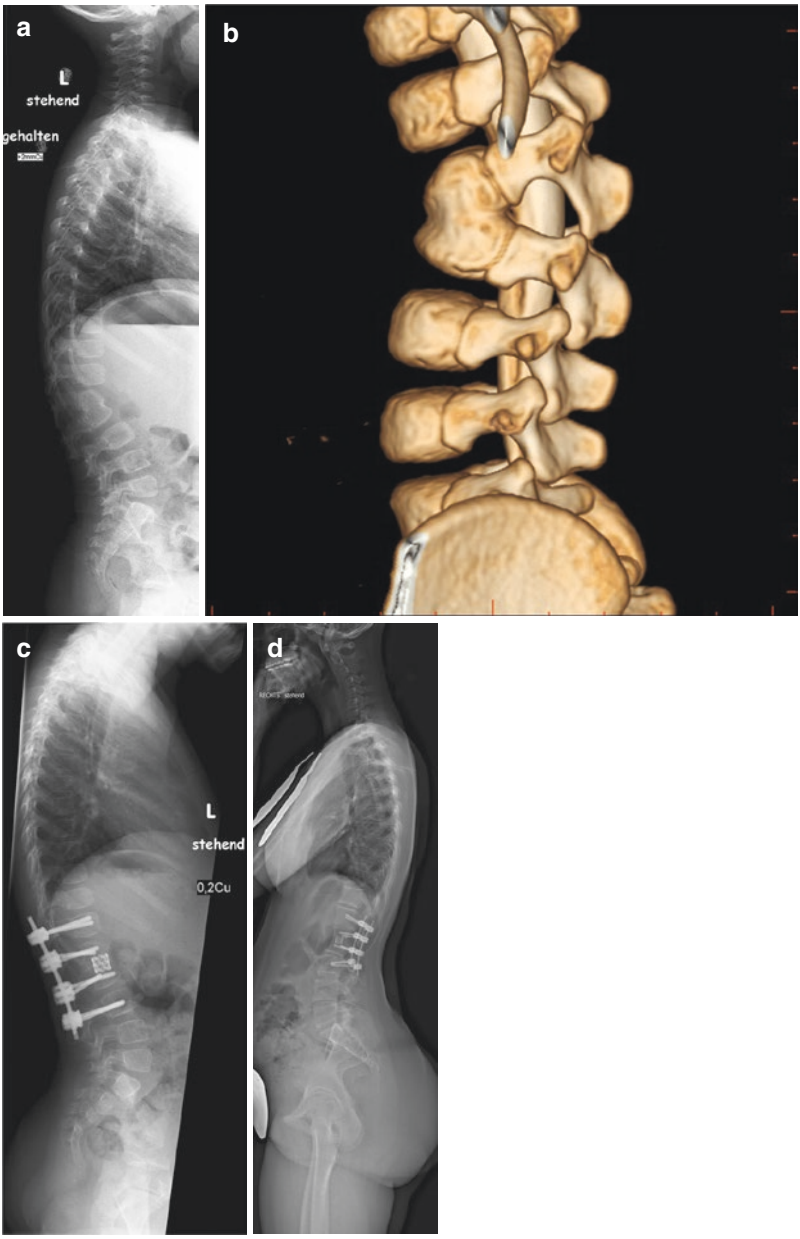

Fig. 24.1 A 2-year-old girl with the anterior bar at L2–3 (failure of segmentation); lateral radiograph (**a**) and 3D-CT (**b**). Postoperative lateral radiographs following osteotomy of the bar through a posterior approach, insertion of a cage and instrumentation L1–4 (**c**); follow-up at the age of 14 years (**d**). Please note the correction to a physiological sagittal profile and the further growth of the spine. Lordosis increases by anterior growth against the posterior instrumentation (tension band)

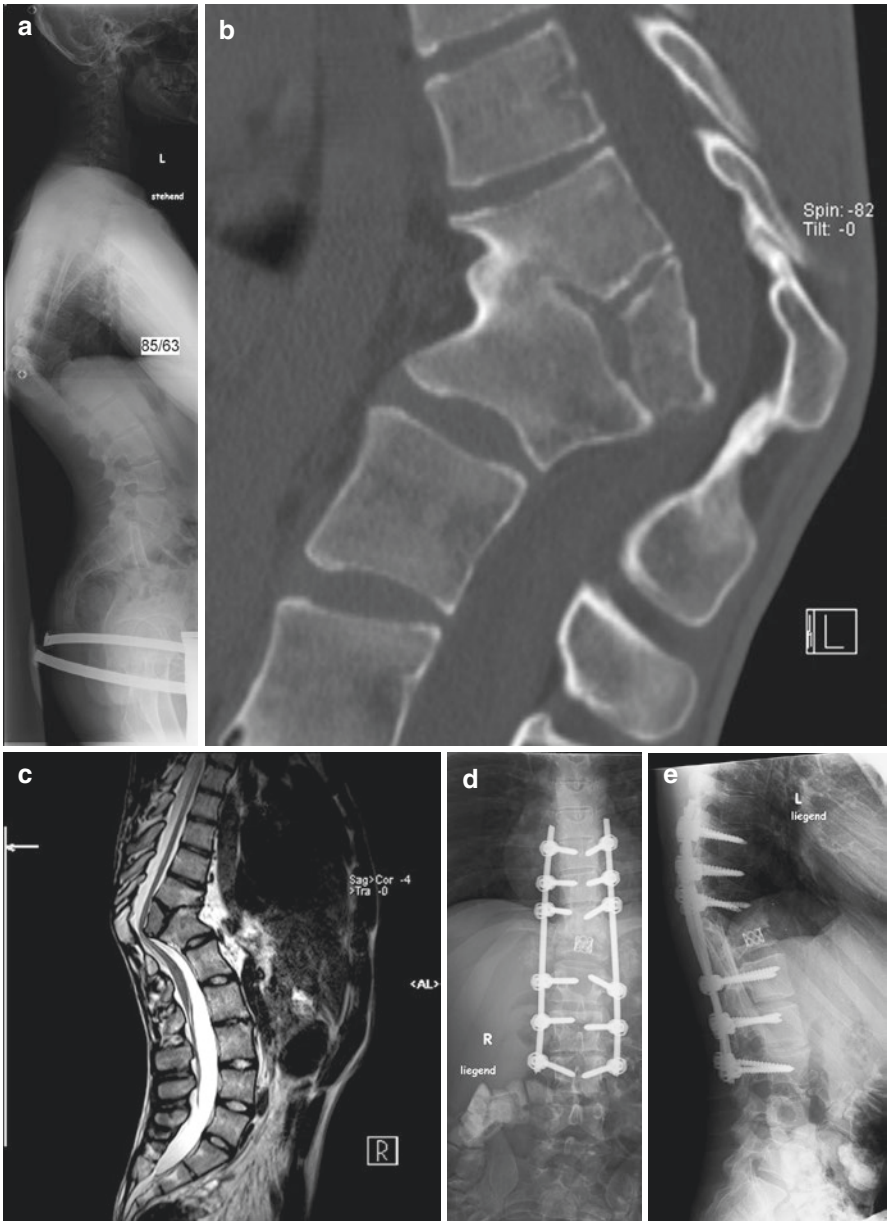

Fig. 24.2 A 22-year-old patient with posterior hemivertebrae T10 and T11, completely fused (**a**, **b**). The patient developed progressive myelopathy with spastic paraparesis (**c**). Postoperative radiographs following posterior hemivertebrectomy and fusion (**d**, **e**); slow neurological recovery. Early surgery in childhood would probably have avoided the neurologic impairment

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Spondylolysis and Spondylolisthesis

25

Yann Philippe Charles

25.1 Definition

Spondylolysis represents the non-union of the isthmus (inter-articular portion) of lumbar vertebrae. The most frequent level is L5, but spondylolysis may also occur at L4 or L3. Spondylolisthesis is defined as anterior slippage (olisthesis) of the cranial vertebra over the caudal vertebra, mostly at L5-S1 (Fig. 25.1); it is the source of localized kyphosis L5-S1 (prognostic factor). The severity of spondylolisthesis is classified according to Meyerding: <25% (grade 1), 25–50% (grade 2), 50–75% (grade 3), and >75% (grade 4) (Appendix K). Grades 1 and 2 are defined as low-grade spondylolisthesis, whereas grades 3 and 4 account for high-grade spondylolisthesis. Complete anterior luxation of the cranial vertebra is defined as spondyloptosis.

25.2 Natural History

The pathophysiology of spondylolysis is assimilated with a fatigue fracture of the inter-articular portion (isthmus), which occurs during growth. Different factors might explain the non-union of the isthmus. Dysplastic isthmus shapes can lead to spondylolysis (Fig. 25.2). Repeated microtrauma such as hyperextension (gymnastics) during the growth period has been described as a risk factor. However, a genetic

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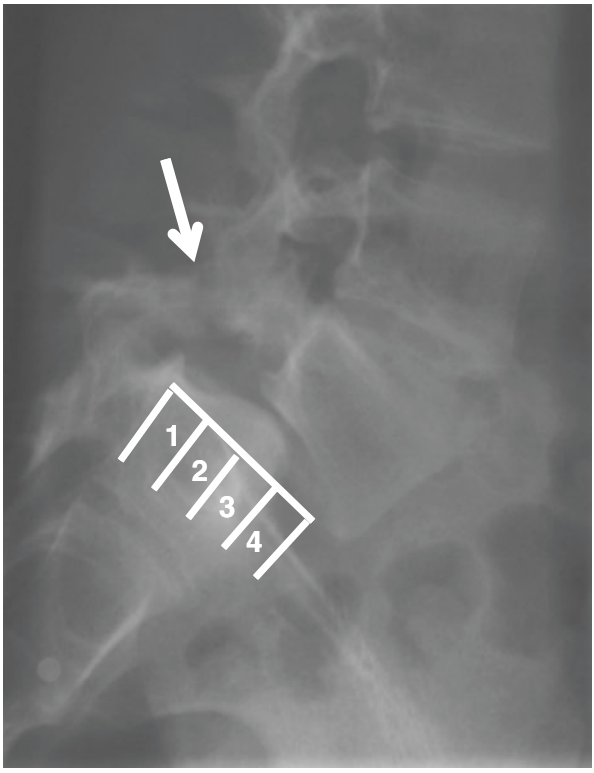

Fig. 25.1 Spondylolisthesis at L5-S1with an anterior slippage of L5 (Meyerding grade 2). The arrow indicates spondylolysis at L5 (Appendix K)

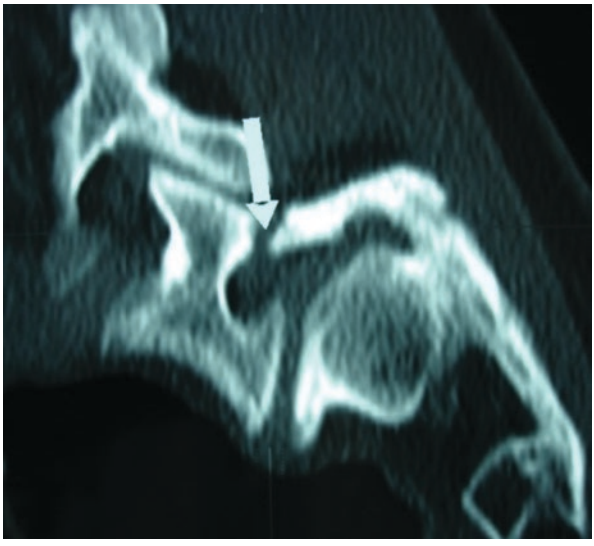

Fig. 25.2 Sagittal CT view demonstrating spondylolysis of a dysplastic L5 isthmus

predisposition might explain the spondylolysis prevalence of 7% in Caucasian populations. A large sagittal pelvic width (high pelvic incidence) is associated with large lumbar lordosis, which enhances stress at the L5 isthmus and thus predisposes to spondylolysis. The severity of spondylolisthesis can progress during growth and create a kyphotic deformity at the lumbosacral junction if high-grade olisthesis is reached. In spondylolisthesis grades 3 and 4, and in spondyloptosis, L5 and S1 nerve roots are stretched and radicular symptoms can occur. At the adult age, spondylolisthesis often leads to premature intervertebral disc degeneration, which can cause low-back pain (Chap. 41).

25.3 Physical Examination

Orthopedic examination of the spine, the hips, and the lower limbs should be carried out in addition to neurologic examination. The patient is asked if low-back pain is present, which is assessed on a visual analog scale (VAS 1–10). Leg pain and paresthesia can also be present. It should be checked if leg pain follows a typical pattern of the L5 or S1 nerve root dermatomes, also known as sciatica or radicular pain. Incomplete posterior leg pain (e.g., stopping at the thigh, knee, or calve level) can mimic sciatica and is described as pseudo-radicular pain. This type of pain represents muscular pain originating from the back or pain from shortened hamstring muscles.

Palpation of the painful level in the lumbosacral area and of paravertebral muscle tightness is performed. In high-grade spondylolisthesis, lumbosacral kyphosis might be observed clinically. The lumbar range of motion is then measured using the Schober method by drawing two points: one at the upper sacral level and another one 10 cm above. The normal range of motion of the lumbar spine is observed if the distance between both points increases to 15 cm in maximal anterior trunk flexion (Schober 10/15 cm). Any shorter distance indicates the limited range of motion of the lumbar spine.

The neurologic examination focuses on radicular sensory and motor testing (Video 25.4). The sensory examination should investigate paresthesia and a pinprick test should be performed. The motor examination requires muscle strength testing according to the Medical Research Council scale from 1 to 5. In high-grade spondylolisthesis, weakness can be observed in some patients, typically at L5. Furthermore, the sphincter function should be investigated by asking the patient if voiding abnormalities are present (Video 25.4).

25.4 Imaging

The standard radiographic examination consists of anteroposterior (AP) and lateral radiographs of the lumbosacral spine, and sagittal alignment should be measured on lateral full spine radiographs (EOS); lateral radiographs also allow the surgeon to

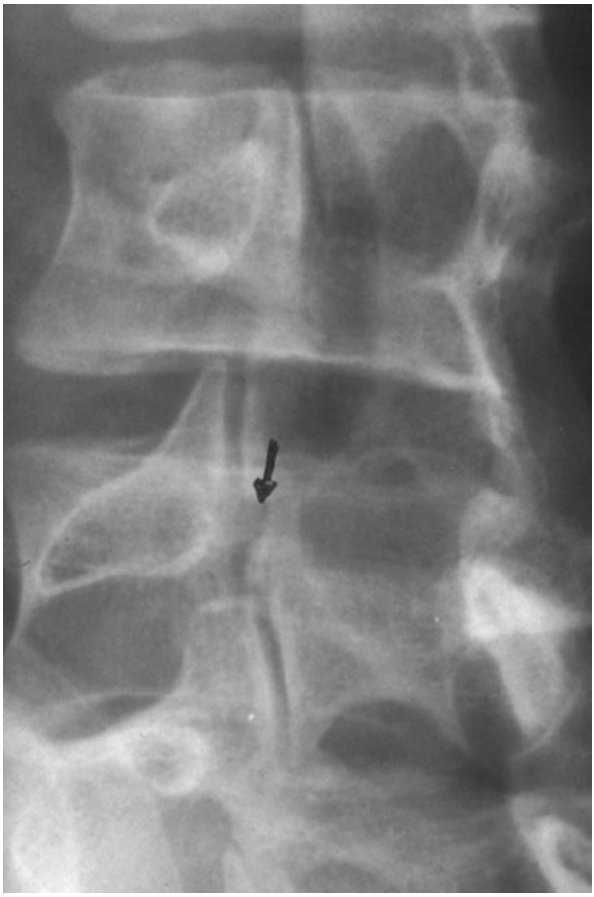

Fig. 25.3 Oblique radiograph of the lumbosacral spine demonstrating the typical "Scottie dog" image, where the collar represents the spondylolysis

quantify the amount of L5-S1 kyphosis. The lateral view usually shows spondylolysis and the amount of spondylolisthesis can be evaluated (Fig. 25.1). In spondylolysis without olisthesis, oblique radiographs of the lumbosacral spine can be helpful in identifying the non-union, appearing as a typical "collar of the Scottie dog" image (Fig. 25.3).

Computed tomography (CT) scan provides good-quality images of bony lesions. If the presence of spondylolysis is unclear on previous plain radiographs, CT represents a helpful diagnostic tool to clarify the diagnosis (Fig. 25.4). However, radiation caused by CT should be avoided whenever possible in children and adolescents. Therefore, this exam should not be considered in the first line.

In patients with low-back pain, magnetic resonance imaging (MRI) is indicated to evaluate the intervertebral disc (Fig. 25.5). Disc degeneration can contribute to

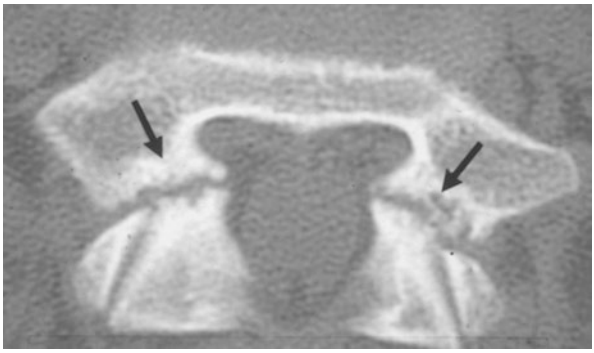

Fig. 25.4 Axial CT scan showing bilateral spondylolysis (non-union) at the isthmus of L5

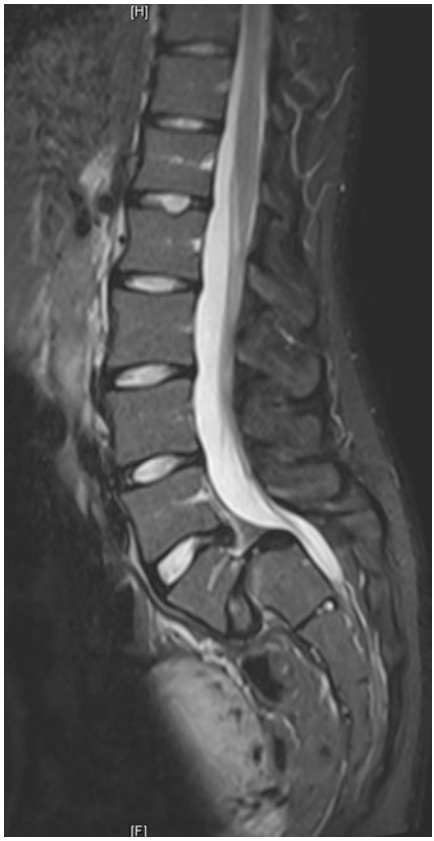

Fig. 25.5 T2-weighted sagittal MRI demonstrating intervertebral disc degeneration at the level of spondylolisthesis L5-S1

low-back pain in adolescents and adult patients. Furthermore, MRI is indicated when evaluating lateral recess and foraminal stenosis. At L5-S1, the L5 nerve root is stretched by the olisthesis and the fibro-cartilaginous nodulus (Gill nodulus) at the level of the spondylolysis narrows the intervertebral foramen.

25.5 Differential Diagnosis

In children and adolescents, non-specific low-back pain (muscular strain) might be considered (Chap. 11). Among deformities, Scheuermann's disease (Chap. 22) and lumbosacral congenital malformations can cause back pain (Chap. 32). Vertebral trauma should be investigated. Inflammatory disorders (arthritis), primary or secondary neoplasms, and infections (spondylodiscitis, Pott's disease) are rare in children.

Degenerative spondylolisthesis represents another entity which is encountered in elderly patients. It usually develops at L4-L5 and is associated with degenerative changes of the lumbar spine such as facet joint osteoarthritis and spinal canal stenosis (Chaps. 45 and 46). Spondylolysis is not present in this type of olisthesis.

25.6 Treatment Options

The management of spondylolysis and low-grade spondylolisthesis is mainly conservative. During the growth period, sports should be stopped, temporarily. Analgesic and anti-inflammatory drugs might be prescribed and a brace can be prescribed to patients who complain of pain. In adult patients, CT-guided infiltration of the spondylolysis can be performed. Physical therapy mainly focuses on paravertebral muscle reinforcement and hamstring muscle stretching. In young patients, surgical isthmus reconstruction and fusion are rarely indicated. In adult patients with intervertebral disc degeneration and low-back pain despite conservative measures, surgical treatment by posterior or anterior (ALIF) fusion might be indicated.

High-grade spondylolisthesis usually represents a surgical indication for L5 nerve root decompression (posterior arch resection of L5) combined with posterior and interbody fusion; surgery should reduce the amount of slippage and the local L5-S1 kyphosis. Non-instrumented fusion can only be applied to children. Adult patients usually require a fusion from L4-S1 (Fig. 25.6). Different interbody fusion techniques exist, including interbody fusion cages (PLIF, TLIF) or trans-sacrum-lumbar grafts. The amount of required reduction should be assessed preoperatively (Videos 25.3, 25.4, 25.6 and 25.7). In situ fusion represents a reasonable approach in balanced spinopelvic alignment. The reduction should be performed with caution in unbalanced alignment (retroverted pelvis with high pelvic tilt).

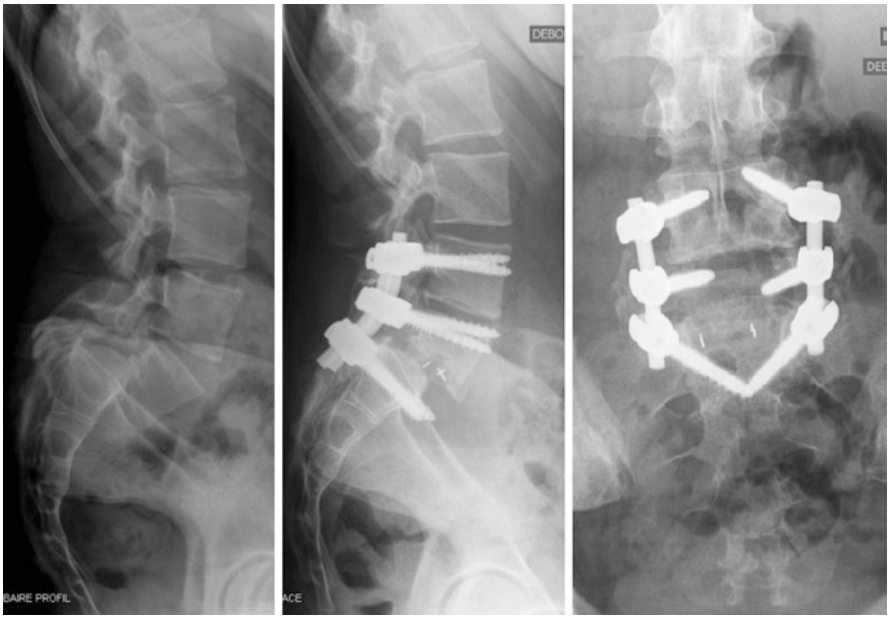

Fig. 25.6 Pre- and postoperative radiographs demonstrating the principle of surgical treatment in high-grade spondylolisthesis by nerve root decompression, posterior instrumentation L4-S1, spondylolisthesis reduction, and posterolateral and inter-body fusion

25.7 Expected Outcomes

Conservative treatment usually provides an improvement in the quality of life in spondylolysis and low-grade spondylolisthesis. Surgical treatment is appropriate in high-grade spondylolisthesis, providing improved spinopelvic alignment and good functional outcomes.

25.8 Potential Complications

Neurologic complications represent the main risk of surgical treatment in high-grade spondylolisthesis. Complete reduction carries a risk of L5 nerve root stretching and paralysis.

25.9 What Should Patient and Family Know?

Spondylolysis and low-grade spondylolisthesis are common in the population and only symptomatic patients require treatment. High-grade spondylolisthesis represents a rarer condition that requires surgical intervention.

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Disc Bulging and Herniation in Children and Adolescents

26

Federico Canavese

26.1 Definition

A bulging disc must be differentiated from a herniated disc; in particular, a bulging disc is a condition in which the nucleus pulposus remains contained within the annulus fibrosus, unlike a herniated disc in which the nucleus pulposus extrudes through the fibers of the annulus fibrosus.

26.2 Natural History

Although intervertebral disk herniation is common in adults, it is relatively uncommon in children (rate in the pediatric population: 5%).

The loss of disc height with subsequent posterior bulging or herniation of the disc into the spinal canal (cervical, thoracic, or lumbar) as well as bulging of the ligamentum flavum can lead to stenosis (dynamic or permanent) with pain that can radiate through the back and sometimes down the arms (if the herniation is in the cervical spine; rare) and legs (if the herniation is in the lumbar spine; more frequent).

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26.3 Physical Examination

Compared to adult patients, children (in particular) and adolescents often will have less specific descriptions of the pain or complain of other symptoms, which leads to a longer duration before diagnosis. Importantly, children and adolescents are less often seen with neurological symptoms such as numbness and weakness.

History of trauma (prior back injury) or sports-related injury, activities with repetitive or excessive axial loading, poor conditioning, decreased range of motion, presence of spine deformity (scoliosis, transitional vertebral abnormalities, spondylolysis, and olisthesis), and obesity can be related to disc herniation.

Presentation is usually acute low-back pain and/or lower extremity radiculopathy. Pediatric patients often will have a less specific description of the pain or complain of other symptoms, which leads to a longer period before diagnosis.

Neurological examination is of paramount importance as patients with disc bulging and disc herniation can develop a broad spectrum of neurological deficits, from mild to extremely severe symptom; depending on the location of disc pathology, radicular pain (upper or lower extremity; debilitating in some cases), progressive neurological deficit (upper or lower extremity), paresthesia (numbness or tingling; motor deficit are rare), changes in tendon reflexes, loss of bladder, and bowel function (Video 26.4). Other symptoms include pain that decreases with rest; increased pain after sitting for long periods of time, or after bending, picking up heavy objects, or twisting; and pain that is relieved with walking or changing positions.

In children and adolescents with lumbar disc herniation, the straight-leg raising test is positive in more than 90% of cases.

26.4 Imaging

Conventional radiographs of the whole spine represent the initial imaging and may allow identification of narrowing of the intersomatic disc space; moreover, plain radiographs of the spine will allow the treating surgeon to rule out other causes of back pain (Chap. 11).

If disc pathology is suspected, magnetic resonance imaging is optimal (a) to determine the morphological changes of the intervertebral disc and ligaments and (b) to evaluate the soft tissue and neural constituents.

Although such degenerative changes are rare in children and adolescents, the computed tomography (CT) can show moderate narrowing of the articular facet space (axial bone window images) as well as subchondral sclerosis.

26.5 Differential Diagnosis

Disk disease is uncommon in the pediatric population. Therefore, other common causes of back pain should usually be sought first (Chap. 11).

26.6 Treatment Options

Accurate clinical examination and imaging are mandatory to identify the level of disc bulging or disc herniation and to propose an adapted treatment option.

Treatment options are always conservative first and usually include rest, physical therapy to improve mobilization, flexibility and strength, limitation of physical activities (sports), and anti-inflammatory medication. Physical therapy is of paramount importance since strengthening the paravertebral and abdominal musculature may improve spine posture and decrease pain. Bracing should be restricted to severe symptomatology, and infiltrations may be indicated if nerve root irritation increases (rare). Bracing is also indicated to reduce pain before starting physiotherapy.

However, pediatric patients respond less well than adults to conservative management and are more likely to require operative treatment. Because pediatric intervertebral disks are more elastic and have a higher water content than adults who often have more dried-out or degenerative discs, surgical treatment can be more difficult to perform; in addition, the pediatric disc does not dry up and resorb like a degenerated adult disc might (Chaps. 42, 44, 45). As a result, open procedures (discectomy) are usually preferred to endoscopic techniques.

Surgery should only be indicated after failure of all conservative treatment options. Indications for surgery are (1) severe pain refractory to more than 4–6 weeks of conservative treatment, (2) disabling pain affecting daily activities, (3) cauda equina syndrome, (4) progressive neurological deficits, and (5) presence of associated spinal deformities.

26.7 Expected Outcomes

Surgical treatment has a good short-term prognosis (resolution of radicular pain and of neurological symptoms); children and teens often return to school and activities quickly after surgery. Most patients report satisfaction and have few complications, but in 12% to 28% of cases, repeated surgical treatment may be required later in life.

26.8 Potential Complications

A symptomatic and severely herniated disc can lead to permanent nerve damage (rare in skeletally immature patients).

26.9 What Should Patient and Family Know?

Although intervertebral disk herniation is common in adults, it is relatively uncommon in children. Studies have shown that between 13% and 57% of adolescents with disc herniation have a first-degree relative with the same disorder; herniation is

less frequent in patients younger than 10 years of age but increases in the adolescent population. Surgery has a satisfactory short-term prognosis, but up to 28% of patients may require repeated surgical treatment later in life; moreover, degenerative changes are still possible in adulthood.

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Schmorl's Nodes

27

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27.1 Definition

Schmorl's nodes (SNs), or vertical intra-vertebral disc herniations, are protrusions of the nucleus pulposus of the intervertebral disc through the vertebral body endplate into the adjacent vertebral body; inflammation can develop if SNs contact the bone marrow of vertebral body.

27.2 Natural History

SNs are quite common and most frequently asymptomatic. Post-mortem studies have found SNs in around 75% of autopsies, at all ages, more frequently in males.

27.3 Physical Examination

SNs are one of the potential causes of back pain in young patients (Chap. 11) although they often cause no symptoms (chronic SNs). Symptomatic patients complain of back pain and stiffness (acute SNs). There is a limited range of motion in the lumbar and/or thoracolumbar area. Neurological exam should be performed (Video 27.4).

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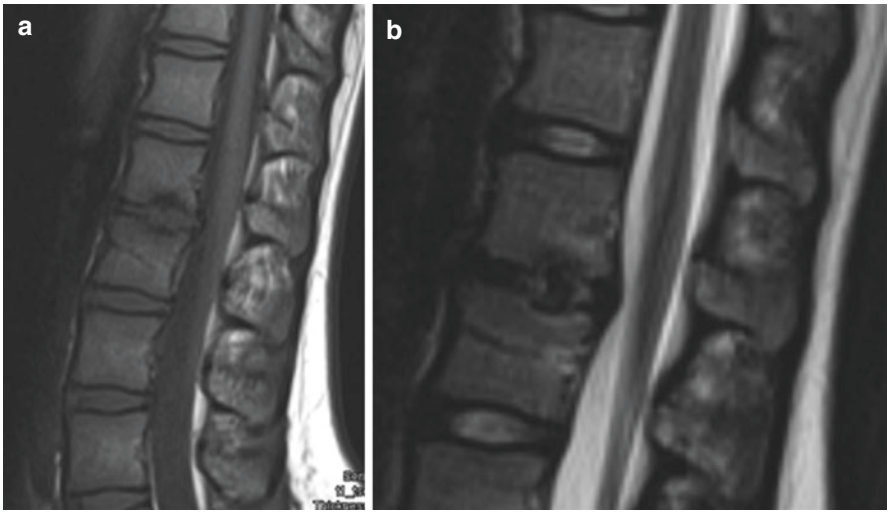

Fig. 27.1 Schmorl's node and disc degeneration (lumbar spine, L1-L2 space) (**a**: MRI T1, **b**: MRI T2)

27.4 Imaging

The best imaging modality for diagnosing SNs is magnetic resonance imaging (MRI) although plain film radiographs of the spine (lateral view) can also detect them. Radiographically, SNs appear as relatively small round-shaped, radio-lucent lesions with/without sclerotic margin, involving the inferior and/or the superior endplate of lower thoracic and lumbar vertebral bodies (Fig. 27.1).

MRI best identifies SNs on the sagittal sequences. SNs usually have the same signal characteristics of the adjacent intervertebral disc, with a thin sclerotic margin. Acute herniations usually show surrounding bone marrow edema and peripheral enhancement.

SNs identified on computed tomography scan have the same characteristics observed on plain radiographs.

27.5 Differential Diagnosis

In symptomatic patients, the other causes of back pain (Chap. 11) should be ruled out, in particular infection and malignancy (different radiographic appearance).

27.6 Treatment Options

Although most lesions are asymptomatic, some SNs can be symptomatic, causing chronic back pain. Most symptoms related to SNs tend to resolve spontaneously or respond to conservative treatment (non-steroidal anti-inflammatory drugs, muscle relaxants, and/or physiotherapy). In patients with chronic and disabling back pain resistant to conservative treatment, fusion surgery with/without removal of the herniated disc is possible in order to alleviate the symptoms. Surgery is possible in patients with Scheuermann's disease (Chap. 22) with associated SNs (source of pain).

27.7 Expected Outcomes

Most symptoms related to SNs resolve spontaneously, or following conservative treatment (over a variable period of time, up to one year in some cases). In the very few cases with debilitating pain, surgery can alleviate symptoms.

27.8 Potential Complications

SNs tend to occur more often in cases of Scheuermann's disease (Chap. 22) with the spine that has become more rigid and hypo-mobile. As a result, forces normally distributed all over the nucleus pulposus are concentrated in a limited area causing upper and/or lower endplates to deform in a concave manner.

27.9 What Should Patient and Family Know?

There is a relatively strong heritability of SNs (>70%). Painful or symptomatic SNs can lead to a significant decrease in quality of life. Currently, there is no established treatment modality.

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Congenital Torticollis (Torticollis Not Related to Trauma)

28

Federico Canavese

28.1 Definition

The congenital torticollis (C-TO), present from birth, is characterized by an inclination and rotation of the head in relation to the trunk, due to contracture or fibrosis of the sternocleidomastoid (SCM) muscle.

Inclination occurs on the same side of the affected SCM muscle, and it is localized across the entire cervical spine while rotation of the face and chin is contralateral and it is largely localized at C1 and C2 levels.

According to the etiology and the reducibility, three types of C-TO can be identified: postural (pTO), muscular (mTO), or osseous (oTO).

28.2 Natural History

C-TO can lead to a malformation of the skull (plagiocephaly), and in untreated cases, it can induce asymmetry of the face (facial scoliosis) characterized by a loss of alignment between the eyes and the mouth (normally parallel). In older children, C-TO is often accompanied by plagiocephaly, a difference in height of the ears and shoulders, and facial scoliosis. The treatment done too late does not allow the correction of distorted craniofacial skeletal structures.

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28.3 Physical Examination

The clinical examination should look for a misalignment of the head and cervical spine, and test the mobility of the cervical spine. The severity of the clinical picture depends on the age of the child at the time of diagnosis; neurological examination should be performed (Video 28.4).

As a rule of thumb, pTO is completely reducible (no muscle tightness or restriction to passive range of motion), mTO is incompletely reducible (tightness of the sternocleidomastoid muscle and restricted passive range of motion), and oTO is irreducible (osseous malformation; Chap. 29).

In case of mTO, the clinical examination should look for an induration or swelling on the lateral side of the sternocleidomastoid muscle, at the level of its distal third (Fig. 28.1). This swelling is rarely present at birth and appears between the second and fourth weeks of life; it disappears a few months after birth and can be replaced either by definitive fibrosis or by a complete recovery of the elasticity of the muscle fibers.

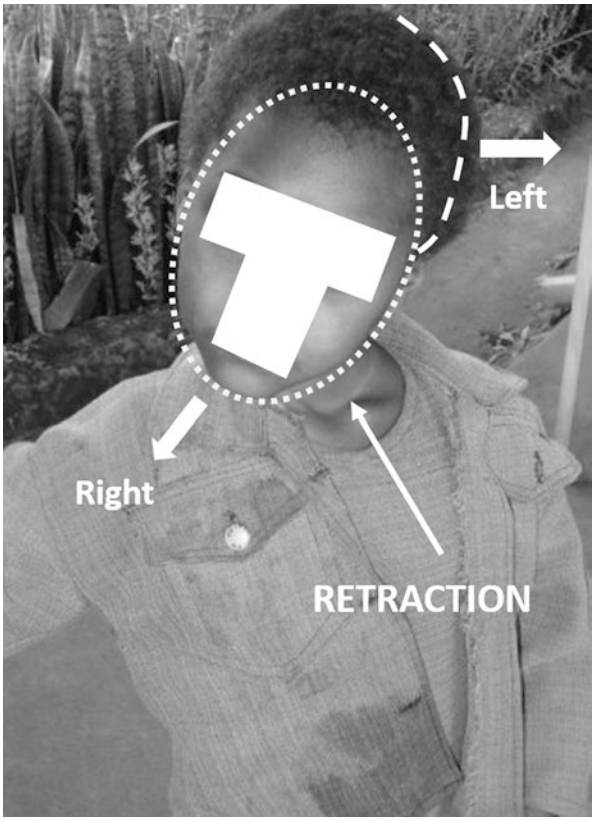

Fig. 28.1 Congenital torticollis (untreated). Left retraction of the sternocleidomastoid muscle; occiput rotates towards the left, chin towards the right

- C-TO diagnosed at birth requires examination of both hips to rule out developmental dysplasia of the hip (DDH); C-To is a malposition whose presence increases the risk of DDH (association C-TO + DDH in up to 15–20% of cases).
- C-TO can correlate with other musculoskeletal abnormalities such as metatarsus adductus, calcaneovalgus feet, and brachial plexus palsy.

28.4 Imaging

Imaging is usually not necessary. In case of oTO, standard anterior-posterior (AP) and later radiograph of the spine allow identification of the underlying bone malformation.

Complementary investigations, such as magnetic resonance imaging (MRI) and computed tomography (CT) scan of the cervical spine, are usually not necessary and are not urgent.

MRI of the whole spine and skull can identify underlying spinal cord abnormalities or spinal dysraphism. CT scan can confirm the osseous anomaly.

28.5 Differential Diagnosis

The following conditions should be ruled out: brain tumor (posterior fossa tumor, in particular), congenital scoliosis (Chap. 12), Klippel-Feil syndrome (Chap. 29), Arnold-Chiari malformation (Chap. 30), syringomyelia (Chap. 31), cervical spine tumor, unilateral congenital absence of the sternocleidomastoid muscle, ocular torticollis, visual disturbances, and Sandifer syndrome (dystonic movements causing torticollis and scoliosis; episodes usually last between 1 and 3 min and can occur up to 10 times a day, and are most often associated with ingesting food. Vomiting, feeding difficulties, anemia, epigastric pain, hematemesis, and abnormal eye movements were also reported. Reflux esophagitis is common).

Radiology can differentiate causes of oTO from causes of mTO (Fig. 28.2).

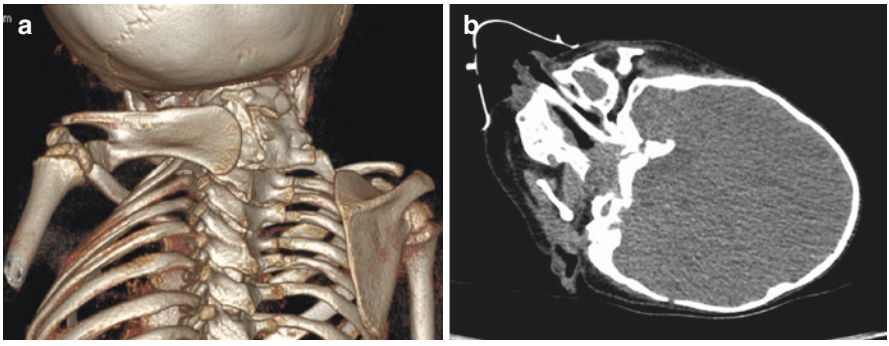

Fig. 28.2 Osseous torticollis in a patient with Klippel-Fiel syndrome and Sprengel's deformity (CT scan; **a**); severe plagiocephaly (CT scan; **b**)

Torticollis due to problems other than SCM muscle abnormalities is characterized by a normal neck examination with a full range of motion. To correct the torticollis, underlying cause needs to be evaluated and treated.

Grisel's syndrome and traumatic atlantoaxial dislocation are also important in the differential diagnosis of torticollis in older children (Chap. 3).

28.6 Treatment Options

Initial treatment focuses on manual stretching, positioning, and close follow-up.

Surgery is indicated if no improvement is seen after 4–6 months of conservative treatment, if there are >15° loss of passive rotation and lateral bending, the presence of a tight sternocleidomastoid muscle, or induration or swelling at the distal third of the sternocleidomastoid muscle (poor prognostic factor). The procedure includes unipolar/bipolar sternocleidomastoids muscle lengthening, or Z-lengthening. Bipolar release and Z-lengthening are indicated for correcting neglected cases.

28.7 Expected Outcomes

With proper treatment, 90% to 95% of children improve before the first year of life, and 97% of patients improve if treatment starts before the first six months. After the age of five, the form and efficacy of treatment are controversial.

Postoperative outcome can be assessed according to Lee et al. (excellent: 17–18 points; good: 15–16 points; fair: 13–14 points; poor: <12 points) (Table 28.1).

28.8 Potential Complications

Untreated patients can develop fascial asymmetry including recessed eyebrow and zygoma, deviation of the chin point and nasal tip, inferior orbital dystopia on the affected side, commissural canting toward the affected side, inferiorly and posteriorly positioned ipsilateral ear, and distorted craniofacial skeletal structures. Early

Table 28.1 Postoperative outcome according to Lee et al.

Points	Neck movement	Head tilt	Scar	Loss of SCM column	Loss of SCM lateral band	Facial asymmetry
3	Full	None	Fine	None		
2	Loss of rotation or side flexion <10°	Mild	Slight			
1	Loss of rotation or side flexion 11°–25°	Moderate		Obvious, cosmetically acceptable		Moderate
0	Loss of rotation or side flexion >25°	Severe	Unacceptable			Severe

treatment is important for the prevention of fascial asymmetry, which is irreversible.

28.9 What Should Patient and Family Know?

When diagnosed and treated early (<6 months), C-TO can be managed conservatively with good or excellent results as most cases tend to resolve spontaneously. In older patients (1 to 4 years), corrective surgery has both cosmetic and functional benefits. In neglected cases (>5 years), the form and efficacy of treatment are controversial and correction of bone malformations should not be expected.

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Klippel-Feil Syndrome

29

Mehmet Çetinkaya and Alpaslan Şenköylü

29.1 Definition

Klippel-Feil syndrome (KFS) is primarily characterized by the presence of congenital union or fusion of two or more cervical vertebrae secondary to failure of segmentation of the developing axis during the first 3–8 weeks of embryogenesis. The prevalence is 1/40.000 newborns with females (60%) more frequently affected than males.

The fused vertebrae can limit the range of movement of the cervical spine, cause torticollis (secondary to bone abnormality; Chap. 28), and lead to chronic headaches and muscle pain in the neck and back; the extension of the affected and fused segments may determine the intensity of the pain.

Three morphologic subtypes were identified and described by Feil in 1919; type 1 includes patients with the massive fusion of many cervical vertebrae without thoracic vertebrae that develop contiguous or noncontiguous bony blocks. Type 2 includes those with two or more cervical vertebrae in a single fusion segment although they sometimes have accompanying hemivertebrae, occipito-atlantal (O-C2) fusion, or other abnormalities. Patients with cervical fusion associated with lower thoracic or lumbar fusion are included in type 3 (Fig. 29.1).

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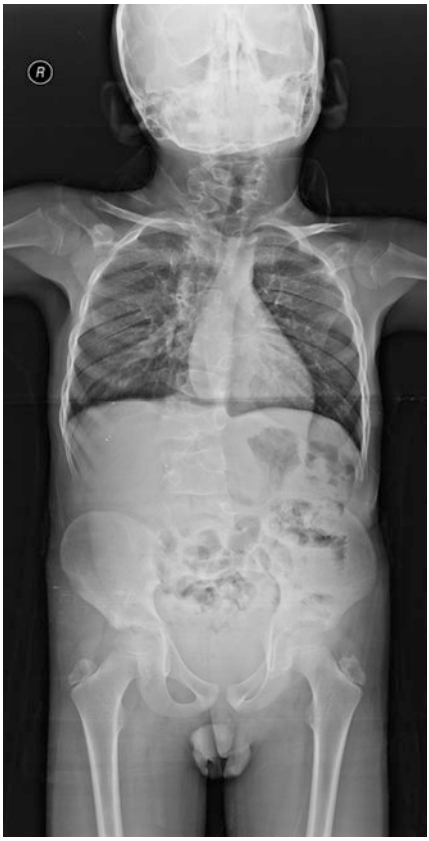

Fig. 29.1 A 14-year-old male patient with Type 3 Klippel-Feil syndrome. Please note the presence of scoliosis, torticollis, and fusion at cervical, thoracic, and lumbar levels

Clarke et al. (1988) have classified KFS according to the position of cervical fusion. According to this classification, KFS type 1 has C1-C2 fusion and autosomal recessive trait; KFS type 2 has C2-C3 fusion and autosomal dominant trait; C3 fusion is found in KFS type 3 characterized by reduced penetrance or autosomal recessive trait; KFS type 4 has an X-linked trait and eye abnormalities associated with cervical fusion.

GDF6, GDF3, MEOX1, SGM1, Notch, and Pax genes have been linked to KFS.

29.2 Natural History

Patients with KFS are often asymptomatic. KFS is a congenital malformation present at birth although mild cases might go diagnosed only later in life once symptoms have worsened; incidental diagnosis is also possible in asymptomatic cases. Patients with KFS may develop spontaneous neurologic sequelae as a result of bony abnormalities. The axial neck symptoms are highly associated with type 1, whereas predominant radicular and myelopathic symptoms are seen mostly in KFS type 2 and

3 (according to Fiel's classification). According to Jovankovicova et al., a wide spectrum of associated abnormalities can be associated with cervical fusion, such as scoliosis (60% of cases, mostly congenital), spina bifida occulta (45%), renal abnormalities (35% to 55%), rib deformity (20% to 30%), deafness (30% to 40%), synkinesia (20%), Sprengel deformity (20%), and congenital heart disease (8% to 14%).

Patients with KFS can develop spinal stenosis and myelopathy in the long term. Though, fusion of the vertebrae itself can damage the nerve in the head, neck, or back. Minor low-energy trauma can aggravate the clinical picture or even cause spinal cord injury, particularly in the setting of the existing stenotic spinal canal.

29.3 Physical Examination

The classic clinical triad includes the short neck, low posterior hairline, and a limited range of neck movement (present in less than 50% of patients). Patients may also present with Sprengel deformity, jaw abnormalities, partial loss of hearing, torticollis (Chap. 28), and scoliosis (Chap. 12) (Fig. 29.2). The torticollis can cause facial asymmetry and plagiocephaly.

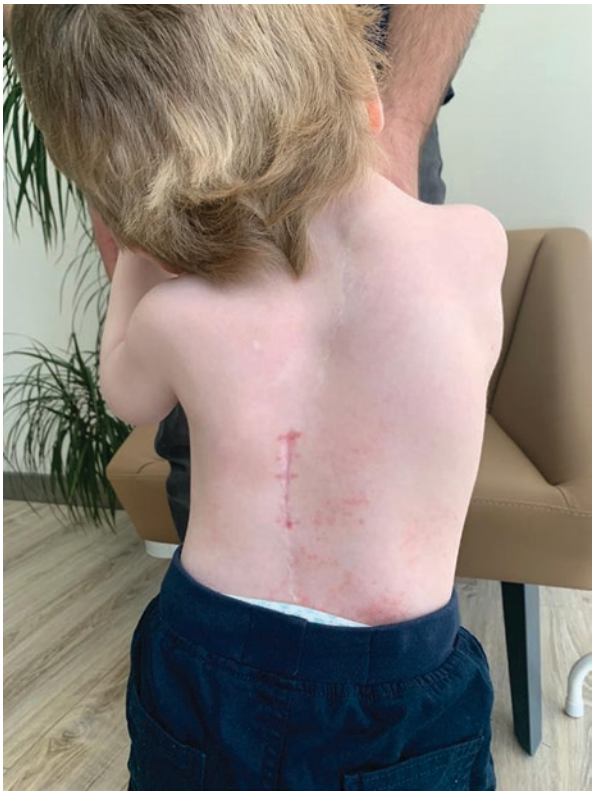

Fig. 29.2 A 2-year-old male patient with Klippel-Feil syndrome presented with scoliosis and torticollis

Approximately 30% of patients have additional skeletal abnormalities. When Sprengel deformity is present, elevation of the underdeveloped shoulder blade (scapula), a lump at the base of the neck, and a limited range of movement of the shoulder complete the clinical picture. In some individuals, an exposed spinal cord may be present due to incomplete posterior arc closure, or a tuft of hair or dimple over the underlying closure defect which is sometimes accompanied by neurologic impairment (Chap. 31).

Some patients have deafness secondary to conductive hearing loss, sensorineural hearing loss, or mixed. Various ocular disorders may also be associated with KFS, such as cross-eye or convergent strabismus, nystagmus, and colobomas. Other abnormalities are torticollis and cleft palate (up to 17% of cases).

Rarely, patients may have ventral septal defects, hypoplasia or agenesis of one or both of the kidneys, abnormal renal rotation or ectopia, and hydronephrosis due to ureteral narrowing. Neurological complications due to associated spinal cord injury mostly result from instability of cervical vertebrae. The neurologic deficit can range between root symptoms and complete para- or tetraplegia. Associated neurological complications tend to develop between the second and third decades of life and may occur spontaneously or following minor trauma. Such complications may include pain, synkinesia or mirror movements, hyperreflexia, hemiplegia, paraplegia, and cranial nerve palsies (Video 29.4).

29.4 Imaging

Radiographs are taken as anterior-posterior, lateral, and odontoid views. Further imaging techniques are not necessary for diagnosis. However, magnetic resonance imaging or computed tomography scan may be needed to evaluate the presence of basilar invagination, spinal stenosis, or other central nervous system abnormalities.

On plain radiographs, cervical fusion is seen most commonly at C5-C6 and C2-C3 level. Any sign of instability and any degenerative change must be evaluated carefully. Typical associated findings are basilar invagination, atlanto-axial instability, degenerative changes, and calcifications within the intervertebral disc space.

29.5 Differential Diagnosis

Diagnosis can be challenging if history, examination, and radiologic investigation are not made properly. Several entities with fused cervical vertebrae should be differentiated from KFS. These include ankylosing spondylitis, juvenile idiopathic arthritis, the chronic complication of discitis, post-traumatic fusion, surgical fusion, spinal anomalies, and congenital muscular torticollis. It may also be a part of other anomalies including MURCS (Mullerian duct aplasia, renal agenesis cervicothoracic somite dysplasia), Sprengel deformity, and Wildervanck syndrome.

29.6 Treatment Options

KFS is often diagnosed during childhood. The diagnosis may be delayed because of the associated anomalies. When diagnosis is made, the orthopedic surgeon must recognize the high-risk skeletal abnormalities (cervical spine, in particular), find out all the associated abnormalities, and make the proper referrals.

The initial treatment is conservative; and it includes cervical collars, braces, non-steroidal anti-inflammatory drugs, or other analgesics. However, patients must be aware of the associated spinal cord abnormalities, the increased risk for spinal cord injury with low-energy trauma, and the potential need for early intervention in case of other organs' abnormalities.

Otherwise, the prognosis of the KFS is relatively good. Patients must be advised to avoid neck injury which may cause spinal cord injury easily. Congenital fusions above C3 are mostly asymptomatic and require abstention from contact sports, especially when occipitalization of the atlas is present. Fusions distal to C3 are most likely to be symptomatic during their normal life span if there is no severe associated anomaly of vital organs. Associated disorders like basilar invagination, adjacent segment disease, and spinal instability are managed individually.

29.7 Expected Outcomes

The life expectancy of patients with KFS is similar to the general population. The factors affecting the quality of life and survival depend on associated anomalies. Most patients are managed conservatively or require lifestyle adjustments and changes. Surgical intervention is rarely indicated. However, symptomatic basilar invagination, high-grade degenerative changes, adjacent segment disease, spinal injury, stenosis, and instability need surgical stabilization with or without decompression.

29.8 Potential Complications

Spinal stenosis, spinal cord injury, spinal instability, degenerative changes, basilar invagination, and adjacent segment disease are reported complications of KFS.

29.9 What Should Patient and Family Know?

The life expectancy of children with KFS is similar to the general population. In some circumstances, contact sports are permitted. However, in the presence of fusion rostral to C3 or long fusion segments (more than 3 levels), any activity that may cause trauma to the cervical spine must be avoided as the risk of injuring the spinal cord is high in this group of patients. Therefore, contact sports, gymnastics,

heavy or traumatic activities must be replaced with moderated daily life activities, and light and non-contact sports.

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Chiari Malformations

30

Robert Mertens, Majd Abdulhamid Samman, and Peter Vajkoczy

30.1 Definition

The Chiari malformations are a group of congenital or acquired hindbrain abnormalities, named after the Austrian pathologist Hans Chiari. The majority of Chiari malformations are types 1 and 2, whereas the remaining types occur very rarely.

- Type 0: patients with syringohydromyelia without hindbrain herniation that respond to surgical posterior fossa decompression. Rare occurrence [1].
- Type 1: mildest form, defined as congenital or acquired caudal displacement of the cerebellar tonsils below the foramen magnum more than 5 mm, with 3–5 mm being borderline.
  - Type 1.5: herniation of cerebellar tonsils and obex below the foramen magnum.
- Type 2 (also known as *Arnold-Chiari malformation*): characterized by caudal displacement of the cerebellar tonsils, hemispheres, vermis, pons, medulla, and fourth ventricle through the foramen magnum and usually associated with neural tube defects such as meningomyelocele.
- Type 3: most severe form, associated with herniation of the cerebellum and sometimes supratentorial tissues into a suboccipital encephalocele below the foramen magnum. Usually not compatible with life.
- Type 4: cerebellar hypoplasia without caudal herniation of the hindbrain below the foramen magnum.

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30.2 Physical Examination

Patients with CM1 present with symptoms caused by hydrocephalus, syringomyelia (Chap. 31), or compression of the brainstem. Nevertheless, patients may be asymptomatic, and scoliosis may be the only symptom; magnetic resonance imaging to check the spinal cord and the cranio-cervical junction must be requested for all children with infantile idiopathic scoliosis. CM1 tends to be diagnosed in the second or third decade of life. The most common symptom is a suboccipital headache and neck pain, followed by mostly unilateral sensorimotor deficits. Characteristic signs in the physical examination are downbeat nystagmus, hyperactive reflexes with atrophy or weakness, gait disturbance, and cerebellar signs like ataxia (Video 30.4).

Patients with CM2 present with symptoms caused by brainstem compression, hydrocephalus, myelomeningocele, and lower cranial nerve dysfunction. Onset is usually in childhood and uncommon in adulthood. Neonates tend to develop severe brainstem dysfunction with rapid neurological deterioration (Video 17.6), whereas symptoms in older children are rarely as severe. Significant findings in the physical examination are swallowing difficulties with aspiration, respiratory distress, apnea, stridor, and signs like downbeat nystagmus and opisthotonus. Weakness progressing to quadriparesis may occur (Video 30.4).

30.3 Imaging

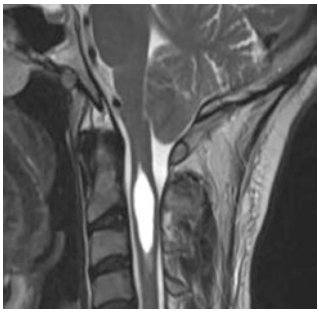
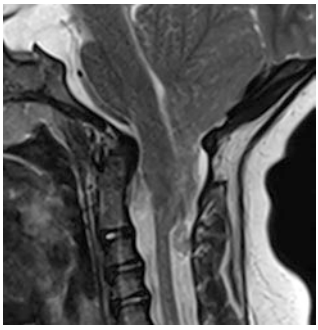
The neuroradiological investigation is crucial to diagnose patients with Chiari malformation and to rule out any associated anatomical pathologies.

- Magnetic resonance imaging (MRI) of the brain and cervical spine is the diagnostic test of choice.
- Cerebrospinal fluid flow study (Cine MRI) using a variety of MRI techniques to visualize the CSF flow through the foramen magnum, which is typically impaired.
- Computed tomography (CT) and CT-myelography: Myelography is used when MRI is not possible or unavailable. Due to bony artifacts, unenhanced cCT is deficient in the evaluation of posterior fossa pathologies.
- Ultrasonography is widely used as a routine examination during the prenatal examination to follow up the intrauterine development and to detect congenital anomalies such as neural tube defects or ventriculomegaly.
- Skull films may demonstrate disproportion from hydrocephalus in infants.

30.4 Differential Diagnosis

Table 30.1 outlines differential diagnosis (Table 30.1; adapted after [2]).

Table 30.1 Differential diagnosis between CM1 and 2

Findings	Chiari malformation type 1	Chiari malformation type 2
Caudally dislocated structures	Cerebellar tonsils	Cerebellar hemispheres, vermis, tonsils, pons, medulla, fourth ventricle
Caudal dislocation of the medulla	Unusual	Yes
Syringomyelia	May be present	May be present
Spina bifida (myelomeningocele)	May be present	Present in most
Hydrocephalus	May be present	Present in most
Age of presentation	Adolescent/adult	Infancy
Symptoms	Suboccipital headache, cervical pain	Hydrocephalus and brainstem/lower cranial nerve dysfunction: swallowing difficulties, respiratory distress, apnea, stridor, downbeat nystagmus
MRI T2 sagittal		

30.5 Treatment Options

In symptomatic patients with CM1, early surgical posterior fossa decompression (PFD) is recommended since patients respond best when operated early (<2 years after onset [3]). Asymptomatic patients or patients with stable long-lasting symptoms may be observed and operated on when they become symptomatic or when showing signs of deterioration. However, when syringomyelia is present in asymptomatic patients, most neurosurgeons recommend surgical intervention.

In symptomatic patients with CM2, PFD is recommended. In the presence of hydrocephalus, implantation of a shunt system is recommended before PFD as soon as the patient can tolerate surgery. In severe cases with feeding difficulties, respiratory insufficiency, and apnea, patients need ICU management. If patients develop stridor due to laryngeal palsy, temporary tracheostomy must be considered.

Suboccipital craniotomy with C1 (sometimes C2/C3) laminectomy with or without duraplasty and tonsillar resection to decompress the cervicomedullary junction, reestablish the CSF flow, and reduce the size of the syrinx is the surgical treatment

of choice. The general surgical technique of PFD is shown in the pictures below, from top left to bottom right (Fig. 30.1): prone position in Mayfield clamp and flexion of the neck (a); midline skin incision from C4 spinous process to inion (b); incision of fascia and preparation of the muscle (c); preparation of occipital bone (d); bone removal above the foramen magnum 3 × 4 cm and laminectomy of C1 (e, f).

Then open the posterior atlantooccipital membrane and cut dural contraction bands, followed by splitting and resection of the outer layer of the dura (Fig. 30.2).

In severe cases, a duraplasty with dural patch graft +/− shrinkage of tonsils can be performed (PFDD). The procedure of duraplasty is shown below from left to right (Fig. 30.3): wide Y-opening of the dura (beware of inferior sagittal sinus; keep arachnoidal layer intact); suture of the patch graft (autograft like fascia or galea patch/xenograft/synthetic substitute).

PFD without duraplasty produces comparable clinical and radiological outcomes and is associated with a lower risk of complications compared to PFDD (pain,

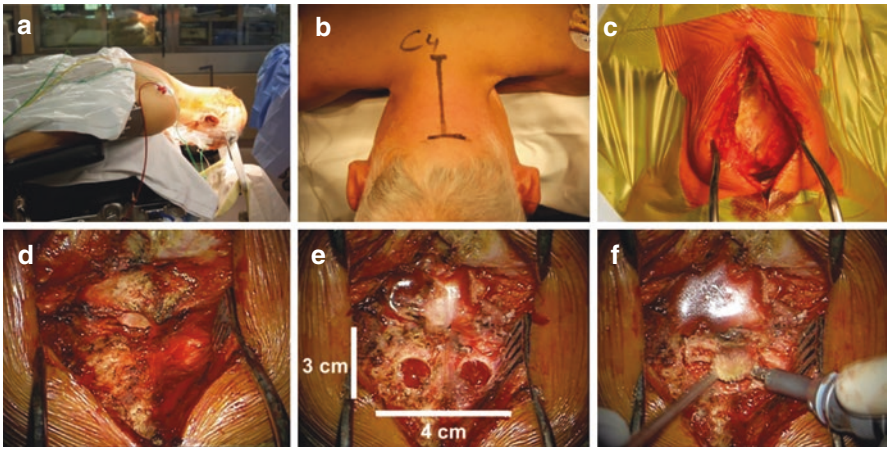


Fig. 30.1 Posterior fossa decompression (PFD) by suboccipital craniotomy: (**a**) Prone position, (**b**) Midline marking of the incision from C4 spinous process to inion, (**c**) Skin and fascia incision and preparation of the muscle, (**d**) Preparation of occipital bone, (**e**) Bone removal above the foramen magnum 3 x 4 cm, (**f**) Laminectomy of C1

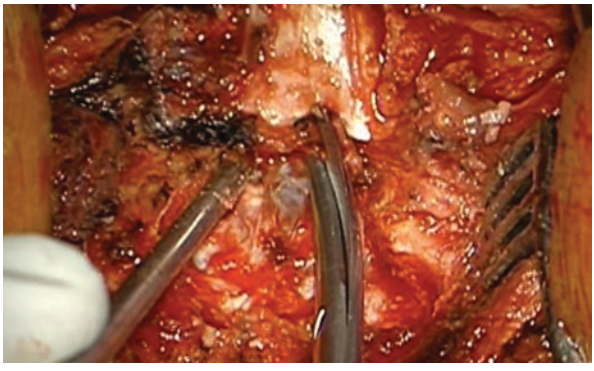


Fig. 30.2 Dissecting and cutting the posterior atlantooccipital membrane

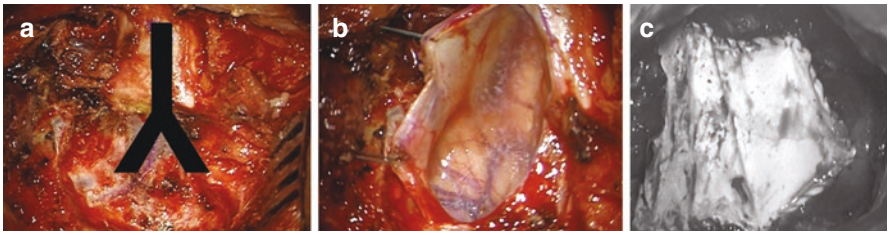


Fig. 30.3 Duraplasty with dural patch graft (PFDD): (**a**) Y-shaped opening of the dura, (**b**) Intact arachnoidal layer after opening the dura, (**c**) Dural closure using patch graft

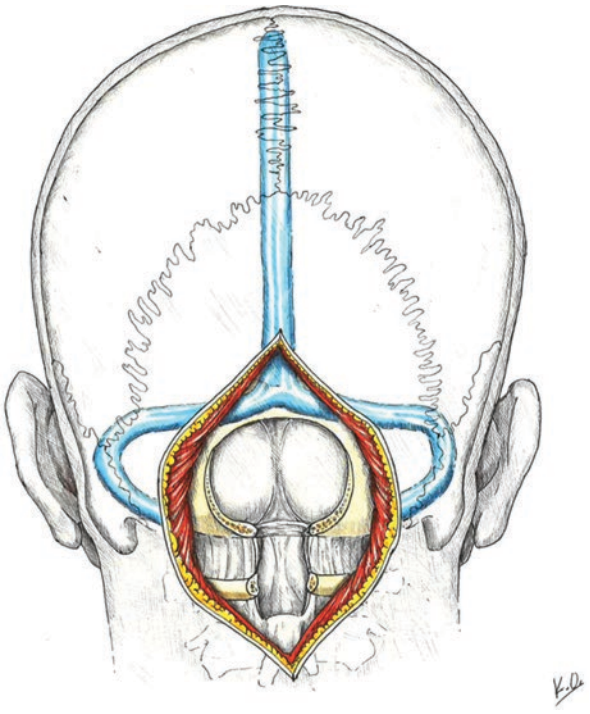


Fig. 30.4 Posterior Fossa Decompression (PFD) with suboccipital craniotomy and laminectomy of C1 and intact dura. Illustration by Kimberly Ohm

morbidity, and CSF fistulas) [4]. Therefore, PFD with closed dura should be preferred to PFDD (Fig. 30.4).

In case of ventral brainstem compression, some authors recommend an additional transoral clivus-odontoid resection [3].

30.6 Expected Outcomes

In general, the postoperative outcomes are considered to be good in symptomatic older children and adults. Successful interventions with almost complete remission of symptoms have been reported; spontaneous regression of thoracic infantile

scoliosis can be observed in some patients if decompression is performed early enough, before the pubertal growth spurt.

In CM1, the pain usually improves significantly after surgery with the most favorable results in patients with cerebellar symptoms, whereas weakness is less responsive [5]. Symptoms lasting longer than two years are correlated with worse outcomes [3]. In CM2, 2/3 of patients showed a (near-) complete resolution of symptoms, whereas 20% had no benefit [6]. The most important prognostic factors are preoperative deficits and rapidity of worsening with respiratory arrest and laryngeal palsy being associated with a worse outcome.

30.7 Potential Complications

General surgical complications include CSF leak, subdural hygroma, wound infection, meningitis/ventriculitis, vascular injuries, and hydrocephalus.

Specific surgical complications related to the posterior fossa decompression include cerebellar herniation through the craniotomy with recurrence of symptoms and brainstem injury/stroke.

Some patients can develop kyphosis at the craniocervical junction, which can be prevented by simultaneous posterior craniocervical junction instrumented fusion.

30.8 What Should Patient and Family Know?

Patients with CM1 without symptoms need follow-up; when developing symptoms, surgery is recommended within two years after onset with good postoperative results [3]. Symptomatic patients with CM2 need surgery with postoperative results depending on the severity of deficits and the rapidity of deterioration.

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Spinal Dysraphism

31

Burak Karaaslan and Alp Özgün Börcek

31.1 Definition

Spinal dysraphism (SD) is an umbrella term that includes congenital midline neural tube defects. Midline closing of bone, neural, or other mesenchymal tissue is defective. Spinal dysraphism can be classified as open type (spina bifida aperta; SBA) and closed type (spina bifida occulta; SBO) dysraphism. Open-type dysraphism includes hemimyelocele, meningocele, myelomeningocele, and hemimyelomeningocele. Closed dysraphism includes lipomyelomeningocele, dermal sinus, diastematomyelia, slit notochord, tight filum terminale, myelocystocele, neurenteric cyst, and developmental tumors such as spinal lipomas (Table 31.1).

31.2 Physical Examination

Clinical symptoms of SBO are often due to tethering of the spinal cord. Tethering of the spinal cord may cause back pain, perineal sensation loss, myelopathy of lower extremities, incontinence, and neurogenic bladder (Video 31.4).

Spinal bifida occulta can be associated with a cutaneous stigma (hypertrichosis, dimple, capillary hemangiomas, or sinus tract; Fig. 31.1).

SD is a congenital spinal malformation, so its occurrence accompanied by other congenital system anomalies is not rare. The most common accompanying

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Table 31.1 Classification of spinal dysraphism

	Spina bifida occulta	
Spina bifida aperta	*With subcutaneous mass*	*Without subcutaneous mass*
• Myelomeningocele	• Lipomyelomeningocele	*Simple*
• Meningocele	• Lipomyelocele	• Spinal lipoma
• Hemimyelocele	• Terminal myelocystocele	• Tight filum terminale
• Hemimyelomeningocele	• Meningocele	• Dermal sinus
		• Persistent terminal ventricle
		Complex
		Disorders of midline notochordal integration
		• Diastematomyelia
		• Neurenteric cysts
		Disorders of segmental notochordal formation
		• Caudal agenesis
		• Segmental spinal dysgenesis

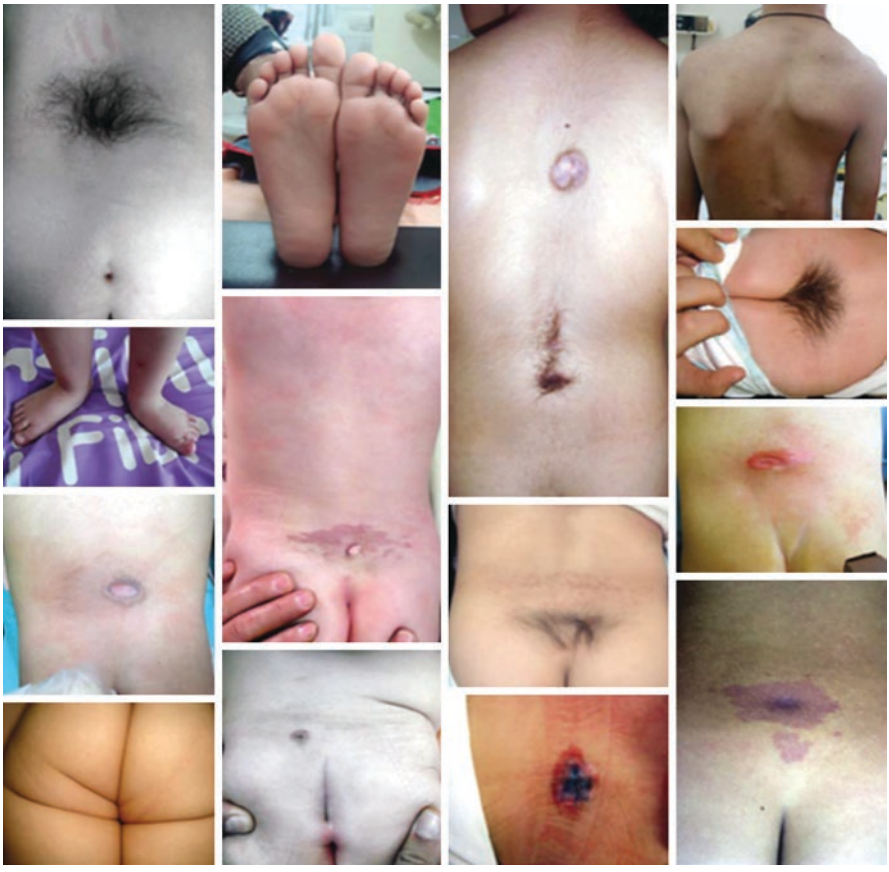

Fig. 31.1 Patients' photographs demonstrate different types of cutaneous stigmata and orthopedic anomalies associated with spinal dysraphism. (GUFM Division of Pediatric Neurosurgery archive)

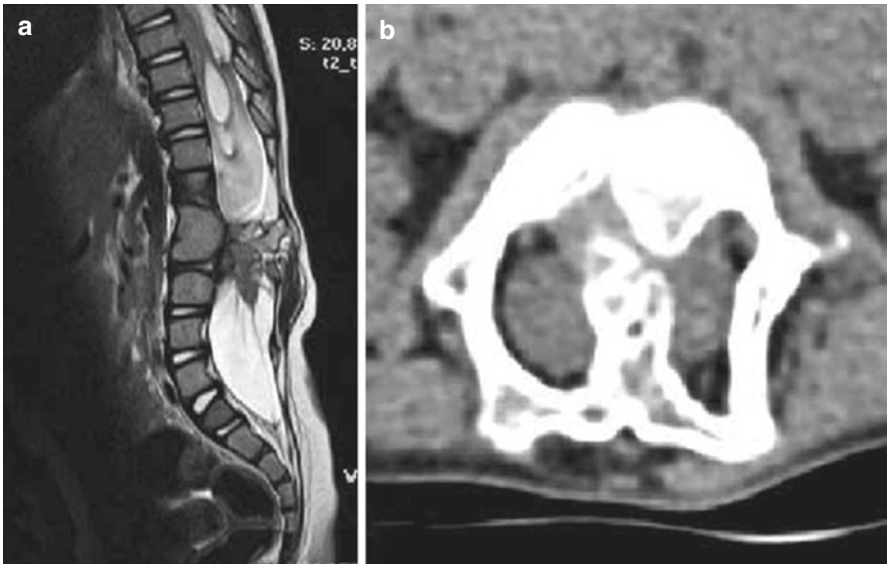

Fig. 31.2 (**a**) T2-weighted sagittal MRI images of an SCM patient. (**b**) Axial CT scan shows a midline osseous spur

congenital anomalies with SD are urologic problems. Studies showed that nearly 25% of SD patients have urologic pathologies. Also, different types of cardiovascular, renal, and skeletal (particularly in lower extremities) malformations are reported.

31.3 Imaging

Computerized tomography is helpful in the assessment of bony malformations (Fig. 31.2). Magnetic resonance imagining (MRI) is the gold standard radiological modality. Complete spinal and cranial MRI scanning is mandatory for these patients, because multiple congenital malformations are not rare in these patients.

31.4 Treatment Options

Surgical treatment is indicated as soon as possible for open-type spinal dysraphism. Local infection and central nervous system infection are the main potential risks for these patients. The main surgical aim is to close the spinal opening which exposes the neural tissue to infectious risks.

Clinical symptoms of occult spinal dysraphism are usually due to tethering of the spinal cord. Neurological deficits usually progress slowly and coincide with body growth. Surgical untethering is indicated as soon as possible for patients with neurological findings. Surgical treatment is aimed to release the tethering of the spinal cord.

There is a debate in the literature about the management of asymptomatic SBO as spinal lipomas, including lipomyelomeningocele. Some studies advocate early prophylactic untethering surgery. However, Kulkarni et al. reported follow-up results of conservative management of asymptomatic spinal lipomas. This study showed no statistical difference in the follow-up result of neurological deterioration between early surgery and conservative management.

Since most of the closed-type patients have subtle neurological problems, surgery in those patients puts them at great risk for further deterioration, so every precaution should be exercised during the surgery of those patients. Intraoperative neuromonitorisation (IONM) is the most important of those measures. Whenever possible, surgeons should rely on IONM both for their (for medicolegal aspects) and patients' safety.

31.5 Expected Outcomes

The most important prognostic parameter for symptomatic SBO is early diagnosis and treatment. Because severe neurological deficits may not be reversible after surgery. On the other hand, the most important prognostic parameter for SBA is the location of the defect and related level of the neurological deficit. Low-level lesion location and good spontaneous movement of lower extremities have a good functional outcome.

31.6 Potential Complications

Hydrocephalus is usually seen in SBA patients. CSF shunting is usually necessary. Other systemic disorders associated with SBA are vertebral deformities, genitourinary and gastrointestinal dysfunctions.

Spinal deformities, such as scoliosis, can be seen with spinal dysraphism. Scoliosis is the result of spinal cord tethering. So surgical correction of scoliosis without untethering the spinal cord will be a disaster.

Latex hypersensitivity is commonly seen in spina bifida aperta, especially in myelomeningocele patients. Frequent and early exposure to latex products is suspicious. Latex-free surgical instruments should be used to avoid latex allergy for these patients.

31.7 What Should Patient and Family Know?

Although surgical closure of SBA and surgical untethering of symptomatic SBO is usually necessary, spinal dysraphism is a congenital embryological disorder. No single intervention is expected to correct all of the problems in a particular patient. Anatomical, urological, cosmetic, and neurological problems require different management techniques and lifelong medical follow-up with a multidisciplinary team is

mandatory. Prevention plays an important role; in particular, folic acid and folates supplementation before conception have been proven useful to prevent congenital midline neural tube defects.

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Sacralization of the fifth Lumbar Vertebra

32

Federico Canavese

32.1 Definition

Bertolotti syndrome (BS), or sacralization of the fifth lumbar vertebra, is a congenital transitional anomaly of the lumbosacral spine characterized by a transverse mega-apophysis of L5 merging or articulating with the iliac wing and/or sacrum; it can be unilateral or bilateral.

32.2 Natural History

BS is linked to a congenital transitional anomaly of the lumbosacral spine. Ossification of the transverse mega-apophysis occurs at the end of skeletal maturation, at which time the impingement may become symptomatic. As a consequence, malformations that were tolerated during childhood may become symptomatic in young adults. In particular, BS is responsible for about 5% of low-back pain cases in the general population (15% to 20% in individuals younger than 30 years of age).

Untreated patients can develop disabling chronic low-back pain and neurological complications (lower-limb pain) related to impingement between the mega-apophysis and the underlying nerve root.

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32.3 Physical Examination

BS is a common cause of low-back pain in young patients (Chaps. 11 and 41). The transitional anomaly of the lumbosacral spine is generally associated with lumbosacral or gluteal pain. Neurological examination completes the physical examination (Video 32.4).

32.4 Imaging

The radiographic diagnosis of BS is straightforward. Standard anterior-posterior (AP) radiograph of the lumbosacral spine allows identification of the transverse mega-apophysis of L5 (Fig. 32.1), in most cases; AP radiograph of the lumbosacral spine with a 30° cranial angulation (Ferguson) has a higher sensitivity in detecting the transitional anomaly. The Castellvi Classification identifies four types of lumbosacral transitional vertebrae, according to the shape of the transverse process and the presence/absence of neo-joint (Appendix L).

Complementary investigations, such as magnetic resonance imaging (MRI) and computed tomography scan of the lumbosacral spine can confirm the lumbosacral transitional anomaly with uni- or bilateral sacralization of L5 (Fig. 32.2), the transverse apophysomegaly, and the presence/absence of transverse process-sacrum/iliac

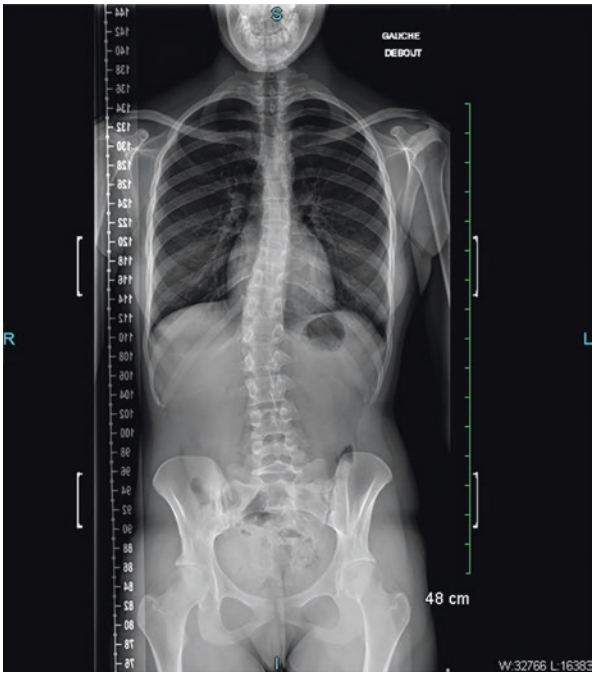

Fig. 32.1 AP radiograph of the spine showing the L5-S1 abnormality (left side) and scoliosis related to back pain

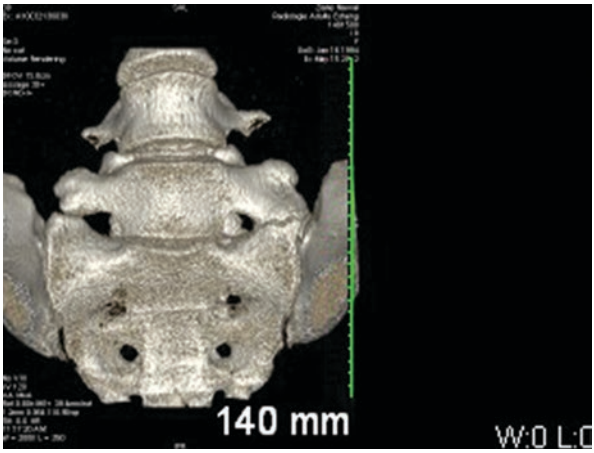

Fig. 32.2 CT scan with 3D reconstruction (sacralization of the fifth lumbar vertebra; left side)

wing neo-joint (pseudo-joint). MRI can show intramedullary bone edema of the involved transverse process and, frequently, a hypoplastic L5-S1 intervertebral disc.

Bone scintigraphy can show hyper-fixation at the level of the neo-joint, and signs of transverse process-sacroiliac impingement, providing additional support for a diagnosis of BS.

32.5 Differential Diagnosis

The other causes of low-back pain in children should be ruled out (Chap. 11).

32.6 Treatment Options

The treatment of BS in children is controversial and includes physiotherapy, medical treatment, infiltration, and surgery. Nonsteroidal anti-inflammatory drugs and/or muscular relaxants are initially recommended. Cases resistant to medical treatment or rehabilitation, and cases with transient efficacy of infiltration, are managed surgically by resection of the transverse mega-apophysis. In case of pain related to degeneration of the intervertebral disc or overlying instability, posterior segmental fusion is recommended.

To confirm the suspicion of BS, infiltration with corticosteroids and local anesthetic of the transverse process-sacrum/iliac wing neo-joint can be performed under fluoroscopic control; in particular, infiltration is used to differentiate patients with pain originating exclusively at the neo-joint (complete pain relief following injection) from those with additional pain due to the irritation of the L4 or L5 nerve root.

32.7 Expected Outcomes

In patients treated conservatively, the outcome is variable; resolution of symptoms should be expected in patients treated surgically, although neurological signs may not completely resolve after decompression if treatment is performed too late.

32.8 Potential Complications

Untreated patients can develop disabling low-back pain and neurological complications (lower-limb pain) related to impingement between the mega-apophysis and the underlying nerve root.

32.9 What Should Patient and Family Know?

The presence of transverse mega-apophysis affects about 20% of subjects; on the other hand, the presence of a neo-joint (pseudo-joint) between the transverse mega-apophysis of L5 and the sacrum and/or iliac wing is found in less than 10% of patients with BS.

BS can be a source of chronic low-back pain; in some cases, neurological signs can develop and may not completely resolve if they are treated too late.

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Osteoid Osteoma and Osteoblastoma

33

Krishna V. Suresh and Paul D. Sponseller

33.1 Definition

Osteoid osteoma is a benign bone-forming tumor, characterized by a small, radiolucent nidus typically less than 2 cm, which produces high levels of prostaglandins and osteocalcin (Fig. 33.1). Osteoblastoma is morphologically and genetically similar to osteoid osteoma but differs primarily on the size of the lesion (more than 2 cm), areas of involvement, patient presentation, and treatment (Fig. 33.2). Osteoid osteoma and osteoblastoma account for approximately 10% and 3% of all benign bone tumors, respectively, and in the spine, primary osteoblastoma accounts for 10% of all osseous spinal neoplasms.

33.2 Natural History

Untreated osteoid osteoma spontaneously resolves over several years. In contrast, untreated osteoblastoma is progressive and will continue to grow, with worsening pain, damage to adjacent healthy bone, and impingement on nearby neurovascular structures.

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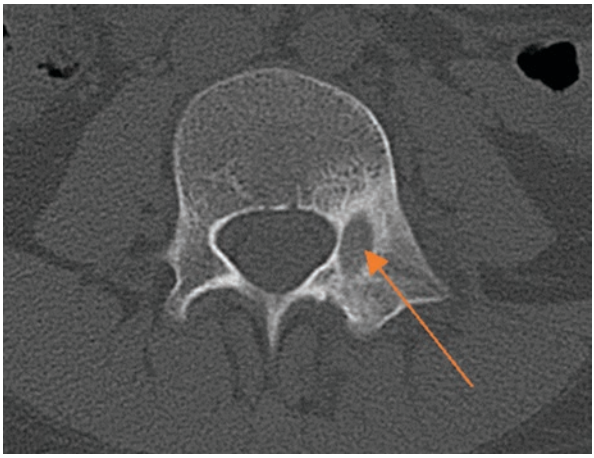

Fig. 33.1 Axial CT view of lumbar osteoid osteoma: 1.5 × 0.7 × 0.7 cm lytic lesion located in left L4 pedicle. Note the rim of reactive sclerosis. Orange arrow indicates lesion

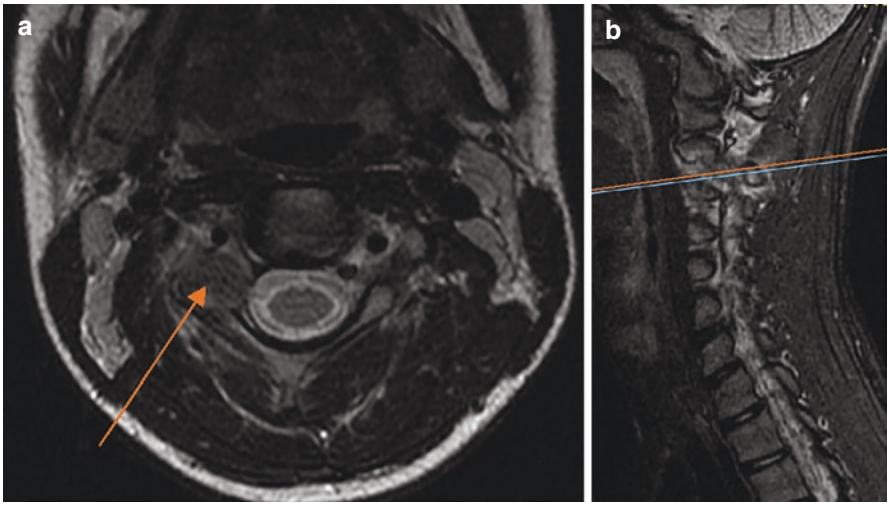

Fig. 33.2 T2 MRI view of cervical osteoblastoma: (**a**) Axial cut: Orange arrow indicates lesion at right C2 pedicle. (**b**) Sagittal cut: Orange line indicates the level of lesion. Lesion appears as hyperintense signal surrounding a central nidus

33.3 Physical Examination

Physical examination for osteoid osteoma and osteoblastoma is similar (Table 33.1). The examination may show a palpable area of bone that is tender to touch. The mass may be swollen and erythematous, and may be accompanied by deformity, effusion, contracture, or atrophy in the affected extremity. Thorough neurologic examination is essential, especially in patients with known or suspected spinal involvement

Table 33.1 Osteoid osteoma versus osteoblastoma

	Osteoid osteoma	Osteoblastoma
Incidence	10% of benign tumors	3% of benign tumors 10% of all spinal osseous lesions
Size	<2 cm	>2 cm
Location	Proximal femur > tibia diaphysis > spine	Spine > proximal humerus>hip
Natural history	Spontaneously resolves	Progressive
Symptoms	Nocturnal pain, relieved by NSAIDs	More chronic pain, dull aches, and not relieved by NSAIDs. Spine involvement more likely to have neurologic symptoms
Management	First line: medical therapy (NSAIDs)	First line: surgery (RFA or surgical resection)

(Video 33.4). Spinal involvement can manifest as postural scoliosis accompanied by muscle spasms at rest or activity. Focal neurologic symptoms can be highly variable depending on the level of the lesion and the extent of soft tissue extension, bony destruction, and cord compression.

Presenting signs. Osteoid osteoma typically presents in the second decade of life but can also be found in pediatric patients. The most common complaint is progressively dull and aching pain that is particularly increased at night and may or may not be related to the activity (Chaps. 11 and 41). Characteristically, the pain resolves with the administration of nonsteroidal anti-inflammatory medications (NSAIDs). Osteoid osteoma is most commonly found in the lower extremities (50%), spine (15%), hand (10%), or feet (5%). Patients can present with limp, muscle atrophy, leg-length discrepancy, and point tenderness at the site of the lesion. In patients with spine lesions, scoliosis (Fig. 33.3), paraspinal muscle spasms secondary to localized inflammatory reactions, and a restricted range of motion may also be present. Osteoblastoma may present similarly to osteoid osteoma but is significantly less responsive to NSAIDs. More commonly than osteoid osteoma patients, osteoblastoma patients have isolated spinal involvement, with 40% of patients having isolated spinal lesions. Symptoms can involve any part of the spine, with approximately one-third of patients complaining of neurologic symptoms such as paraplegia or paraparesis. Radicular symptoms, secondary to soft-tissue involvement and mass effect, occur in approximately 50% of patients. Paraspinal muscle spasms and restricted spinal range of motion may also be present.

33.4 Imaging

On plain radiographs, osteoid osteoma appears as a <2 cm radiolucent nidus surrounded by reactive sclerosis. Osteoblastoma appears very similar to osteoid osteoma but is a larger lesion (>2 cm). Lesions may not be visible on radiographs due to location in spine or increased cortical thickening around the nidus. In these cases,

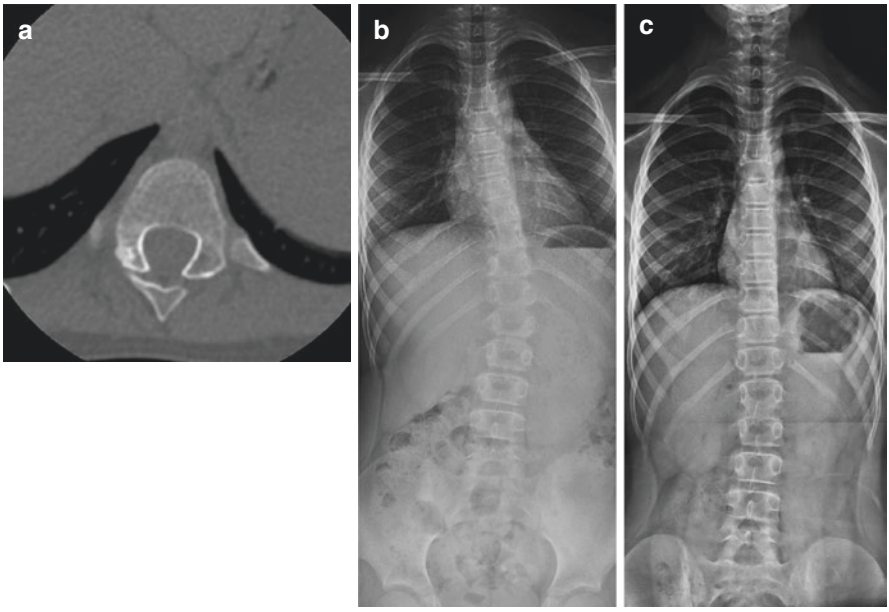

Fig. 33.3 Scoliosis status post T11 osteoid osteoma resection in female patient: (**a**) Preoperative axial CT: it demonstrates osteoid osteoma in right super articular process of T11. Orange arrow indicates lesion. (**b**) Preoperative AP film: moderate left-sided curve measuring approximately 24°. Orange lines represent end vertebrae. (**c**) AP view film taken six months postoperatively: Resolved curvature

Computed tomography (CT) scan is the next preferred imaging modality for spinal lesions, to identify the size and location of the lesion (Fig. 33.1). Magnetic resonance imaging (MRI) is generally not recommended due to the *flare phenomenon* which refers to soft-tissue swelling and edema adjacent to the lesion that may falsely suggest more malignant or infectious etiologies. If MRI imaging is obtained, lesions appear as high-intensity signals on T2 sequences due to calcification and vascularity of the lesions. For osteoblastoma, lesions are typically found in posterior elements of the vertebrae, including laminae, pedicles, transverse processes, and spinous processes (Fig. 33.2).

33.5 Differential Diagnosis

Differentials for osteoid osteoma include osteoblastoma (Table 33.1), stress fracture, and osteomyelitis. Features in favor of osteoid osteoma are: (1) pain at rest or activity and lesions parallel to the cortex (versus pain relief with rest and perpendicular or oblique fractures lines in stress fractures); (2) <2 cm in size (versus >2 cm in size for osteoblastoma); and (3) absence of systemic signs of infection with no radiographic signs of periosteal reaction/elevation, periosteal purulence, or significant cortex destruction (all seen in osteomyelitis) (Fig. 33.3). The differential for

osteoblastoma overlaps with osteoid osteoma but can also include osteosarcoma (Chap. 38) and aneurysmal bone cyst (Chap. 56).

33.6 Treatment Options

For osteoid osteoma, first-line treatment is clinical observation and NSAID treatment. Approximately 50% of these cases can be managed with NSAIDs alone. In patients with osteoid osteoma with spinal involvement, NSAIDs can be used for pain control in patients without concurrent scoliosis. In patients with pain refractory to NSAIDs, osteoblastoma must be considered and operative management is recommended. Medical management is not effective in patients with osteoblastoma. Operative management can be performed with CT-guided radiofrequency ablation (RFA) or superficial resection. RFA is first-line therapy in patients who fail medical therapy and who have periarticular extremity lesions. RFA is generally avoided in patients with lesions immediately adjacent to spinal cord or nerve roots due to a high risk of thermal neurovascular injury. In these patients, surgical resection, either with curettage/biopsy (Video 33.8) or total en-bloc resection (preferred option), is indicated. Scoliosis associated with these lesions rarely requires additional treatment and typically resolves after resection.

33.7 Expected Outcomes

Most commonly, the removal of the radiolucent focus results in total pain relief. For osteoblastoma, the prognosis following surgical resection is good, although recurrence rates can be as high as 20% if the lesion has grown outside the bone. A locally aggressive subtype of osteoblastoma has been identified and is more common in older patients. Aggressive subtypes can display more significant vertebral and epidural involvement.

33.8 Potential Complications

Ten to 15% of patients treated with RFA can experience recurrence of lesions. In patients who underwent superficial resection with curettage, recurrence rates are typically 10% to 20%. Successful operative treatment is dependent on complete resection of the nidus, with no remnants.

33.9 What Should Patient and Family Know?

Osteoid osteoma and osteoblastoma are benign bone tumors that both have excellent prognosis. Some of osteoid osteomas respond well to medications, can be managed conservatively, and resolve spontaneously, while osteoblastoma is usually progressive and requires surgical intervention.

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Osteochondroma and Multiple Hereditary Exostosis

34

Krishna V. Suresh and Paul D. Sponseller

34.1 Definition

Osteochondroma is a benign lesion that is derived from aberrant cartilage, typically appearing as a cartilaginous cap overlying a bony spur in the metaphysis of long bones. Lesions are only rarely found in the spine, making up approximately 3% of all benign spinal tumors (Figs. 34.1 and 34.2). Solitary osteochondroma can be secondary to sporadic mutations, fracture involving growth plate, or radiation therapy.

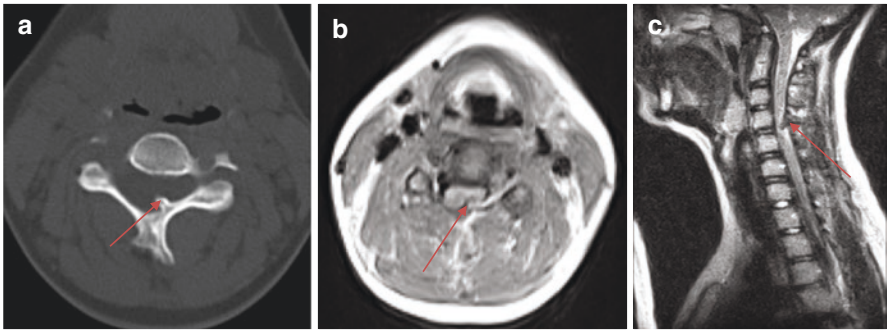

Fig. 34.1 A 14-year-old female with a history of MHE and spinal osteochondroma at C4 lamina, preoperative: (**a**) Preoperative axial CT view reveals a lesion on left C4 lamina, (**b**) preoperative axial MRI view, and (**c**) preoperative sagittal MRI view. Orange arrows indicate the location of the lesion. [Courtesy of Prof. Alpaslan Şenköylü]

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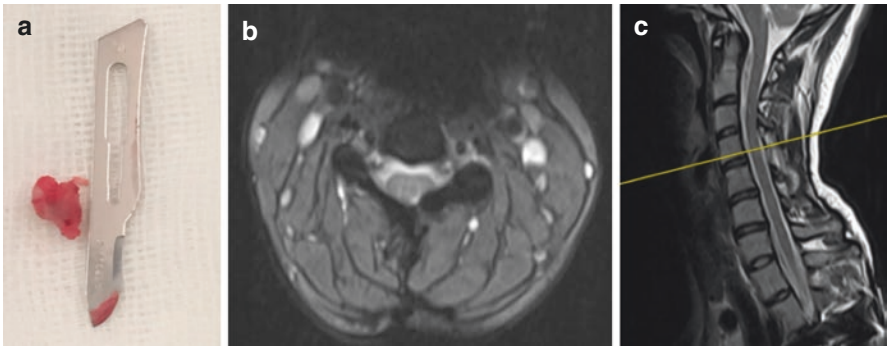

Fig. 34.2 A 14-year-old female with a history of MHE and spinal osteochondroma at C4 lamina, postoperative: (**a**) Image of resected spinal osteochondroma. (**b**) Axial MRI four years after resection does not show recurrence. (**c**) Sagittal MRI four years after resection does not show recurrence. [Courtesy of Prof. Alpaslan Şenköylü]

Multiple hereditary exostosis (MHE; autosomal dominant inheritance of germline mutations in EXT1, EXT2, or EXT3 tumor suppressor genes), also known as hereditary multiple osteochondromas (HMO), is a disorder characterized by two or more osteochondromas in appendicular or axial skeleton (Fig. 34.3). Approximately 9% to 11% of spinal osteochondromas are secondary to MHE (prevalence is approximately 1:50.000).

34.2 Natural History

Osteochondromas grow throughout childhood and cease growth once physes have closed. Patients with spinal osteochondromas develop neurologic symptoms secondary to mass effect and have good outcomes when the lesion is excised appropriately. Prognosis is excellent with disease-specific mortality considered negligible.

34.3 Physical Examination

Spinal osteochondromas may be completely asymptomatic or may present with pain on palpation, a decreased spinal range of motion, or neurologic deficits. These symptoms depend on the location of the lesion, which is most commonly in the cervical spine. A thorough neurologic examination should be performed to evaluate for any motor or sensory abnormalities (Video 34.4). A spinal range of motion, as well as any focal tenderness on palpation, should be noted. A thorough examination of all joints and extremities should be performed to evaluate for the presence of palpable lumps or associated angular deformities.

Presenting signs. The majority of spinal osteochondromas are outside the spinal canal, with the incidence of intracanal osteochondroma being approximately 27% among MHE patients. In patients without spinal canal involvement, symptoms are

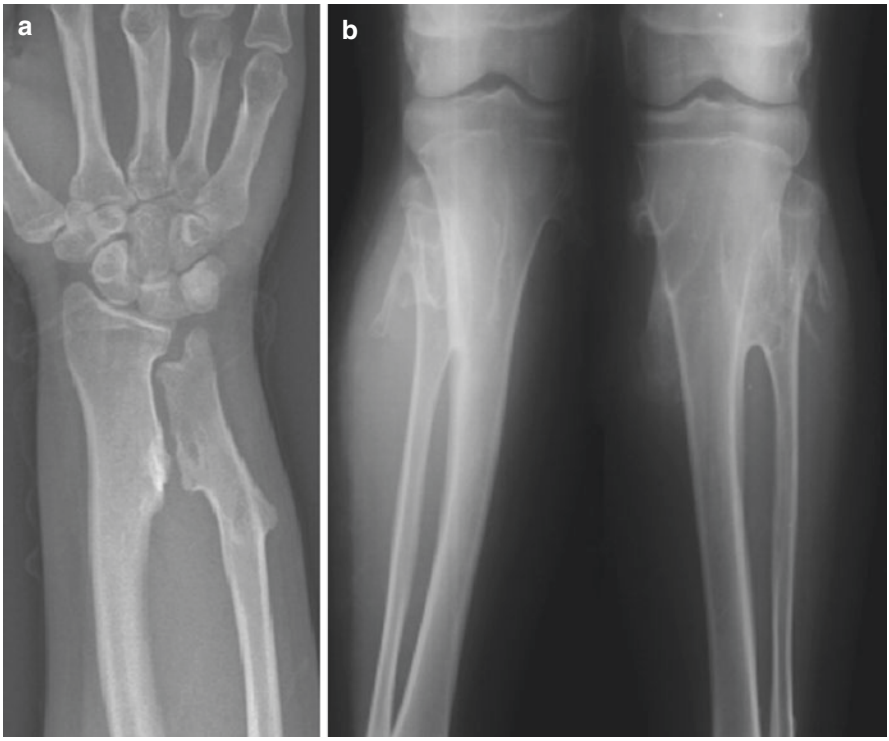

Fig. 34.3 Upper and lower extremity osteochondromas in an MHE patient: (**a**) AP X-ray or right-hand demonstrating lesions on distal ulna and radius. (**b**) AP X-ray of lower extremities demonstrating lesions in bilateral proximal tibias. Valgus deformity of the right tibia can be appreciated. [Courtesy of Prof. Alpaslan Şenköylü]

primarily limited to mechanical pain in the region of the lesion. In patients with canal involvement, sciatica and compressive myelopathy are the most common findings. Spinal osteochondromas have been previously documented to present as severe cervical myelopathy, involving severe gait disturbances, loss of sensation, and diffuse hyperreflexia, though such presentations are rare. Symptomatic presentation is most commonly due to the mass effect of lesions, fractures through lesion, or malignant transformation. Local impingement of nerves, ribs, and tendons can cause significant pain in active motion and limit the range of motion. In the extremities, a palpable lump is typically present, with most lesions presenting in the distal femur (30%), proximal tibia (20%), or humerus (10%).

34.4 Imaging

Magnetic resonance imaging (MRI) is the imaging of choice in patients with spinal involvement, typically revealing lesions in the posterior elements of the vertebrae, including lamina, transverse processes, and spinous processes. Lesions are typically

found in cervical and thoracic vertebrae and particular care should be taken to identify any spinal canal involvement or nerve root impingement (Fig. 34.1). On MRI, the appearance of the cartilaginous cap can vary significantly based on the extent of calcification but is generally less than 2 cm. On T2 sequences, the central fatty marrow appears hyperintense, with a hypointense rim of cortical bone. There are no established cost analyses regarding systematic MRI spine screening of asymptomatic MHE patients, although surveillance screening is typically performed once in children who do not require sedation. MRI is indicated in patients with neurologic symptoms (Fig. 34.2). Plain radiographs are generally not utilized to evaluate spinal osteochondromas, as lesions may be difficult to identify. Bony spinal protrusions in posterior elements of the spine may be identified. Lesions can appear as sessile or pedunculated lesions and cartilaginous cap can present as rings of calcification. Evidence of bony destruction with developing cortical irregularities or increasing thickness of the cartilaginous cap >3 cm after skeletal maturity may indicate malignant transformation.

34.5 Differential Diagnosis

Both clinical and radiographic differential diagnoses include a wide range of benign and malignant bone tumors including periosteal chondroma, parosteal and periosteal osteosarcoma (Chap. 38), and enchondroma. In favor of osteochondroma is evidence of continuous medullary involvement between the lesion and host bone and absence of significant periosteal reactions or destructive lytic lesions.

34.6 Treatment Options

For most cases, observation with routine follow-up is sufficient. Operative resection/biopsy (intra and extra-canal) can be considered only when patients have symptoms associated with mass effect or if the lesion is cosmetically unappealing (Video 34.2). En bloc or intralesional resection can be performed, as radical resection is typically avoided in the spine. Due to the potential malignant transformation later in life, all lesions should be monitored carefully, especially in patients with MHE. Ideally, operative intervention should be delayed until after skeletal maturity has been reached.

34.7 Expected Outcomes

The majority of osteochondromas stop growing after skeletal maturity. Patients who undergo operative resection of symptomatic spinal osteochondromas typically experience significant symptom relief.

34.8 Potential Complications

The risk of malignant transformation to chondrosarcoma is under 1% for solitary osteochondromas and 5% for osteochondromas associated with MHE. The continued growth of lesions after skeletal maturity should raise suspicion for malignancy. There is a chance of recurrence following resection, if osteochondromas are not completely excised. MHE patients are also at higher risk for keloid development from incision sites.

34.9 What Should Patient and Family Know?

Osteochondromas are benign tumors derived from cartilage. Most cases of osteochondroma do not require active treatment and typically stop growing after the patient reaches their growth potential. Spinal involvement is rare. Screening MRI may be obtained in asymptomatic patients and is indicated in patients with neurologic symptoms.

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Eosinophilic Granuloma (Vertebra Plana)

35

Federico Canavese

35.1 Definition

Eosinophilic granuloma (EG) is a benign form of Langerhans cell histiocytosis (LCH). It is usually a solitary lesion (= monostotic disease) that can affect any bone of the human skeleton. Spine involvement may progress to vertebra plana (VP) which is characterized by the almost complete loss of anterior and posterior vertebral body height. Synonyms of VP are *pancake* or *silver dollar* or *coin-on-edge* vertebra [1] (Figs. 35.1 and 35.2).

35.2 Natural History

Solitary lesions spontaneously resolve; there are a 100% survival rate and low rates of recurrence for patients with the monostotic disease.

35.3 Physical Examination

The physical examination of the child may be essentially normal as most EGs are incidental findings.

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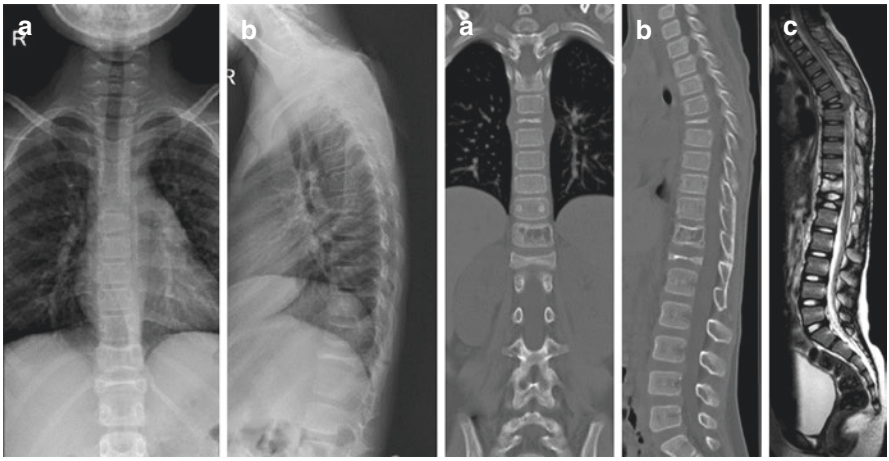

Fig. 35.1 Eosinophilic granuloma (T12). Standard radiographs (**a** and **b**, left side), CT scan (**a** and **b**, right side), and MRI (**c**, right side)

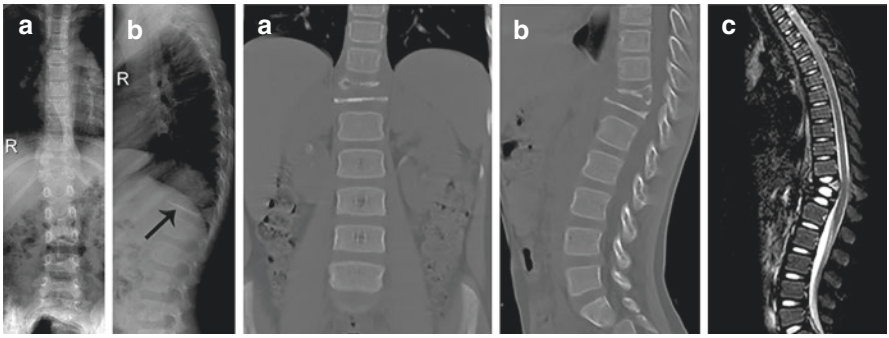

Fig. 35.2 Eosinophilic granuloma (T11 and T12). Standard radiographs (**a** and **b**, left side, black arrow), CT scan (**a** and **b**, right side), MRI (**c**, right side)

EGs of the spine are primarily asymptomatic but can be occasionally painful. Symptoms such as pain, tenderness on palpation, restricted spine motion, and torticollis can be severe and depend on spinal location. Neurologic symptoms and spinal instability are uncommon. The most common location is the thoracic spine followed by the lumbar and the cervical spine.

A thorough exam, including a complete neurologic evaluation, is necessary for patients with known or suspected cranial or spinal involvement (Video 35.4). Laboratory studies are also important, including a basic laboratory panel, inflammatory markers, coagulation studies, and urinalysis in order to differentiate EG from infection (Chap. 65) and other causes of lytic bone lesions [1–3]. Laboratory findings are usually non-specific except for a moderate and inconsistent rise in the erythrocyte sedimentation rate.

35.4 Imaging

Vertebral EG in skeletally immature patients appears as osteolytic on plain radiographs and results in VP with sparing of the posterior elements and disc spaces. Cervical spine EG more often manifests with osteolytic lesions, rather than VP.

Once a lesion has been identified, it is important to rule out the presence of additional lesions; for this purpose, a skeletal survey or bone scintigraphy can be requested even though the skeletal survey should be preferred as some lesions may not be visualized with scintigraphy.

Computed tomography scan is helpful to confirm the diagnosis and to estimate the amount of cortical disruption; magnetic resonance imaging is highly sensitive although nonspecific.

35.5 Differential Diagnosis

Symptomatic EG of the spine is one of the potential causes of back pain in children and adolescents (Chap. 11). It is necessary to rule out multiple lesions and multisystem involvement, and other forms of LCH, which are Letterer-Siwe disease and Hand-Schüller Christian disease. If a simultaneous skull lesion is present, its biopsy is generally easier than the spine and allows faster diagnosis (Video 35.8).

The radiographic differential diagnosis should include plasmacytoma (Chap. 61), multiple myeloma, lymphoma, Ewing's sarcoma (Chap. 39), and other sarcomas (Chap. 38), tuberculosis (Chap. 64), osteomyelitis (Chap. 65), osteochondritis, and osteogenesis imperfecta [1, 3]. In favor of the EG diagnosis are the isolated spinal disease, the lack of constitutional symptoms, and minimal laboratory abnormalities.

35.6 Treatment Options

Brace treatment has been shown to be sufficient to allow remodeling and reconstitution of the vertebral height and shape, and to avoid kyphosis; however, the process of reconstruction can be extremely long. Observation alone or biopsy to confirm the diagnosis of EG has also been recommended as a treatment strategy. Nevertheless, symptomatic patients are good candidates for the percutaneous biopsy for histopathological evaluation. This also facilitates differential diagnosis. Intralesional methylprednisolone injection seems to be an effective and safe treatment method. EG rarely causes spinal instability which needs a surgical stabilization [1–3].

35.7 Expected Outcomes

In children, vertebral EGs resolve spontaneously with time (one year or more). The biopsy itself can help with treatment by triggering the inflammatory response.

35.8 Potential Complications

Spinal instability and neurological compromise are extremely rare.

35.9 What Should Patient and Family Know?

LCH is a spectrum of diseases ranging from simple, solitary lesions of bone to leukemia-like disorders. EG is the most common expression of LCH; it is a benign lesion and usually solitary.

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Medulloblastoma and Other Seeding Tumors

36

Aydemir Kale and Hakan Emmez

36.1 Definition

Medulloblastomas are malignant embryonal tumors of the cerebellum that tend to spread in the cerebrospinal fluid (CSF). They are among the most common central nervous system (CNS) tumors of childhood, accounting for 10–15% of pediatric CNS tumors [1]. Although relatively rare, medulloblastoma can also affect adults. While leptomeningeal involvement is the most common in the cerebrospinal spread, intramedullary metastasis is rare. Typically cerebrospinal fluid seeding is seen in, e.g., medulloblastomas, ependymomas, high-grade astrocytomas, germinomas, or choroid plexus tumors [2]. In the management of these lesions, there are still difficulties in whether to continue with surgery, radiotherapy, and chemotherapy. Evaluation of the disease and determination of response to treatment is mainly based on the evaluation of magnetic resonance imaging (MRI) picture. However, clinical and CSF examination of the patients are also very important.

36.2 Natural History

Medulloblastoma often metastasizes throughout the CNS. Multiple lesions, both nodular and laminar, can be found in the brain and in the spine. The presence of metastases affects prognosis and treatment decisions. Patients are generally

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evaluated for metastatic spread by spine MRI with contrast-enhanced examinations and also CSF cytology [1, 3].

36.3 Physical Examination

The clinical findings of the patients can be divided into two groups. The first belongs to the region where the disease is primarily located: focal neurological deficits, signs of increased intracranial pressure, and epileptic seizures. The second is similar to what can be observed in other spinal cord lesions: pain, motor deficit, sensory changes, and sphincter dysfunction (Video 36.4). The pain usually fits the location of the tumor and tends to increase at night and with movement (Chap. 11). Motor and sensory changes can vary considerably, depending on the location and extent [2, 3].

36.4 Imaging

Spinal metastases can be intramedullary or extramedullary. Intramedullary localization is less common and occurs by hematogenous or direct spread from the leptomeninges. Extramedullary, intradural metastasis is much more frequent and usually results from CSF seeding from primary CNS neoplasms which are in close proximity or within the CSF compartments. MRI is considered the gold standard imaging modality to diagnose spinal intramedullary tumors. The lumbosacral region is most frequently affected with a nodular and irregular, contrast-enhancing thickening of the thecal sac and nerve roots. The surface of the spinal cord may be coated, also known as "sugar-coating" [2]. Contrast-enhanced T1-weighted sequences are mandatory. Figure 36.1 shows a patient with lumbar intradural medulloblastoma.

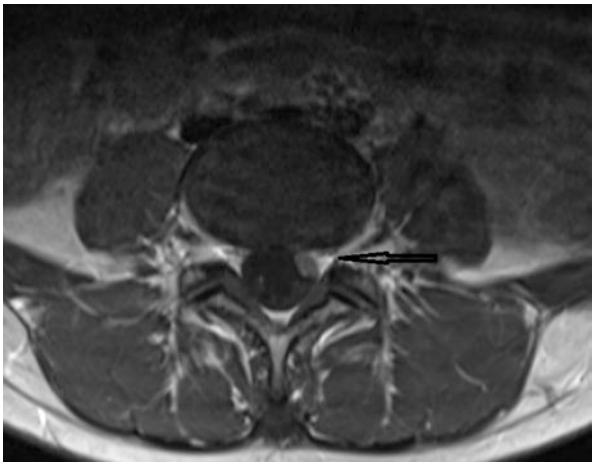

Fig. 36.1 MRI of patient with lumbar intradurally medulloblastoma (white arrow)

Medulloblastoma patients were classified as average or high risk by disease staging using the Chang classification, which included age, post-resection tumor size, CSF cytology, and CNS and extra-CNS metastases (Fig. 36.1). Also, it has recently been subclassified with histological and genomic findings [1].

The sensitivity of MRI for the detection of disseminated tumor is 83% compared with 60% for individual contemporaneous CSF samples and 78% for multiple CSF samples, over time.

In the assessment of leptomeningeal metastases, CSF collection for cytology or flow cytometry differs between studies in terms of timing, collection site, and volume of CSF collected.

36.5 Differential Diagnosis

Intramedullary and extramedullary pathologies should be kept in mind in the differential diagnosis of spinal medulloblastomas. The most common ones are astrocytoma (Chap. 37), ependymoma (Chap. 60), and seeding choroid plexus tumors and germinomas. Spinal involvement due to hematological malignancies should not be forgotten, especially in the childhood age group. Spinal masses observed in patients with a previous diagnosis of primary CNS tumor should be considered in favor of metastasis.

36.6 Treatment Options

Aggressive treatment in medulloblastoma results in better survival for these patients. After gross total tumor resection, radiation is given to the posterior fossa. In children older than 3 years of age, a lower dose of radiotherapy is given to the rest of the head and spinal cord to treat a macro- or microscopic tumor that has spread along the CSF pathways. In children younger than 3 years of age, radiation therapy is usually delayed until after the first postoperative chemotherapy to reduce neurotoxicity. In addition to adjuvant chemotherapy, bone marrow transplant and high-dose chemotherapy have also been shown to improve survival in some patients [1, 3].

It has been reported that medulloblastoma surgery often causes tumor cells to spill into CSF. The presence of tumor cells seen in early postoperative CSF samples, however, does not always indicate that the cells are capable of establishing distal implants. Therefore, spinal MRI and CSF samples obtained more than 2 weeks after surgery will reduce the incidence of false-positive samples.

Given the paucity of literature, the management of intramedullary spinal metastasis remains controversial. The case reports available so far only suggest biopsy and chemotherapy with or without radiation.

Early diagnosis of disseminated tumor is important for the initiation of prompt treatment that may prevent neurologic deterioration, produce symptomatic improvement, and improve or prolong survival.

36.7 Expected Outcomes

Widespread spinal involvement is observed in 33% of the patients followed up with MRI after medulloblastoma surgery. This spread occurs predominantly in the leptomeninges and rarely in the vertebral marrow. Spinal MRI has been reported to have higher diagnostic accuracy than CSF cytological analysis for early detection of the disseminated tumors [1]. The presence of disseminated disease seen with MRI is associated with a poor prognosis.

36.8 Potential Complications

In addition to general complications of the surgery, the progression of neurological deficit and development of instability/deformity may occur after removal of the tumor (kyphosis post laminectomy, in particular). Besides, the negative effects of adjuvant chemotherapy and radiotherapy should be kept in mind, including the loss of spinal growth in younger patients.

36.9 What Should Patient and Family Know?

It should be noted that the prognosis of the patients mainly depends on the extent of the disease. In diseases spread by spinal insemination, the treatment process is quite difficult, and tumor control is generally not possible. The possibility of recurrence should be kept in mind, and strict follow-up and controls should not be neglected.

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Spinal Astrocytoma

37

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37.1 Definition

Spinal cord tumors represent 6% to 8% of all central nervous system tumors combined and are relatively rare compared to intracranial neoplasms. Astrocytomas are the most common intramedullary spinal cord tumor in childhood that develops from astrocytic glial cells [1]. The peak incidence of spinal astrocytomas occurs in the third decade and is more common in males. It is usually diagnosed a few months after symptoms begin. The thoracic and cervical regions of the spinal cord are more frequently affected, and usually multisegmental involvement occurs [1, 2]. It can be diffuse or limited, and cystic areas can be found in half of the cases.

37.2 Natural History

Three-quarters of spinal astrocytomas are low grade. They tend to grow quickly or slowly depending on the aggressiveness of the tumor [3]. If left untreated, they can cause serious disability. The severity of preoperative neurological deficits is correlated with poor postoperative outcomes. An increased incidence is observed in patients with NF-1 (Chap. 23).

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37.3 Physical Examination

Symptoms in intramedullary spinal cord tumors usually develop slowly, over a long period of time. The most commonly reported symptoms are pain, motor deficit, sensory changes, and sphincter dysfunction [1] (Video 37.4). The pain usually fits the location of the tumor and tends to increase at night and with movement. Scoliosis may accompany the thoracic region. Motor and sensory changes can vary considerably, depending on the location and extent.

37.4 Imaging

Astrocytomas are usually intramedullary masses that cause a diffuse expansion in the spinal cord. If osseous remodeling has not developed yet, radiographs and computed tomography (CT) scans are normal. Magnetic resonance imaging (MRI) is considered the gold standard imaging modality to diagnose spinal intramedullary tumors (Fig. 37.1). They are generally detected as hyperintense lesions in T2-weighted images and as iso-hypointense lesions in T1-weighted images in MRI. Its borders are not clear, and there is heterogeneous contrast enhancement [2]. The true size of the mass may not be understood due to edema in the cord, and contrast-enhanced examinations help in the distinction. There may be a cystic component as well as intratumoral hemorrhage. Figure 37.1 shows a patient with cervical intramedullary grade 2 astrocytoma.

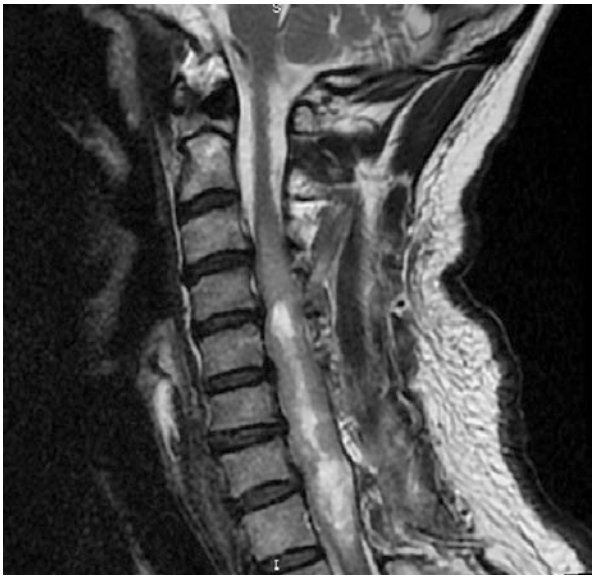

Fig. 37.1 MRI of patient with astrocytoma (cervical spine)

37.5 Differential Diagnosis

The most important differential diagnosis is ependymoma (Chap. 60). It is observed more frequently in adults. More frequent observation of scoliosis and bone remodeling, location of the central part of the spinal canal, well-circumscribed nature, high tendency to hemorrhage, homogeneous contrast involvement in focal intense, and more frequent and prominent cystic components are characteristics of ependymomas [3]. In addition, other intramedullary masses such as hemangioblastoma, epidermoid cyst, ganglioglioma, and metastasis should be kept in mind as potential differential diagnosis.

37.6 Treatment Options

Treatment protocol choices for intramedullary astrocytomas remain controversial but usually include surgery, radiotherapy (RT), and chemotherapy [1–3]. Among surgical options are gross total resection, subtotal resection, and biopsy (Video 37.8). The main treatment for spinal astrocytomas is surgical removal. Figure 37.2 shows the surgical view of cervical intramedullary grade 2 astrocytoma (Fig. 37.2). However, it is generally not possible to remove the tumor completely due to its infiltrative nature. Concerning the surgical approach, some spine surgeons recommend the following cleavage, while some others recommend removing the tumor from the inside to the outside of the glial plane. CUSA is extremely helpful in the removal of astrocytomas because low-grade astrocytomas tend to be hard and sticky. Intraoperative ultrasonography is an extremely helpful method for both surgical strategies. Somatosensory evoked potentials taken with the help of electrodes placed on the dorsal columns are another functional auxiliary method during surgery. Since the motor pathways can be damaged independently of the sensory system, motor

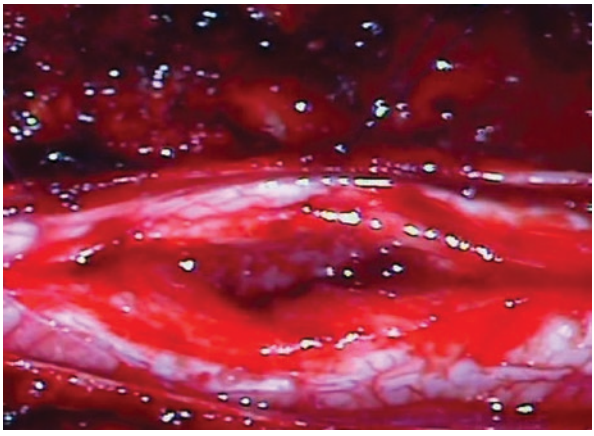

Fig. 37.2 Surgical view of cervical intramedullary grade 2 astrocytoma

evoked potentials can be monitored by taking transcortical stimulation and epidural recording to eliminate this risk. Survival rates of patients with gross total or subtotal excision are much better than those with biopsy. Therefore, postoperative adjuvant chemotherapy and RT are used for tumor control. RT can significantly prolong survival time. On the contrary, the spinal cord is sensitive to the effects of radiation. Overdosage of RT treatment has been proven to lead to the occurrence of radiation-related tumors. Considering this, overdosage must be avoided to reduce the risk of radiation-related tumors. Although temozolomide and bevacizumab are the most widely used chemotherapeutic agents, their effect on survival in high-grade astrocytomas is controversial.

37.7 Expected Outcomes

Histological grading is the most important predictor of survival in spinal cord astrocytomas. Mortality risk increased 14 times in high-grade gliomas compared to low-grade gliomas. Young patients have a better prognosis than the elderly population. The prognosis of spinal glioblastoma cases was extraordinarily gloomy, with a mean survival of 14.3 months, and only 14.1% of patients still survive 24 months after initial diagnosis. Also, gender does not have a significant effect on prognosis. Since total resection rates of lesions located in the cervical region are better, the prognosis is better, while the possibility of permanent neurological deficit in the thoracic region is much higher. Surgery performed with mild neurological deficits at the onset of symptoms has better results. As much tumor resection as possible improves the prognosis. Postoperative RT increases mortality rates in low-grade gliomas, while the opposite prolongs survival in high-grade gliomas. While chemotherapy is within the standard treatment protocol for high-grade gliomas, its effectiveness has also been reported in low-grade gliomas, recently. Although there is not much data about the use of RT in combination with chemotherapy, it is reported that it has little effect on long-term survival [3].

37.8 Potential Complications

In addition to general complications of the surgery, the progression of neurological deficit and development of instability and/or deformity may occur after removal of the tumor. Besides, the negative effects of adjuvant chemotherapy and RT should be kept in mind.

37.9 What Should Patient and Family Know?

It should be known that the prognosis of the patients depends on many factors. Very good results can be obtained, or the rest of their life can be maintained as a nursing patient. The possibility of recurrence should be kept in mind, and strict follow-up and controls should not be neglected.

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Osteosarcoma

38

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38.1 Definition

The most common malignant bone tumors of the spine are metastasis (metastatic disease; Chap. 63). However, OS is the second most common primary malignant bone tumor after multiple myeloma, of which only 3% to 5% occur in the spine. OS mostly affects patients during their second and seventh decade of life (peak incidence). OS is a very aggressive high-grade malignant bone tumor with a poor prognosis. Radiotherapy, chemotherapy, surgery, and combined treatment options have been developed to manage patients with OS. Recent advances in treatment, including aggressive en bloc resection and the use of adjuvant and neoadjuvant chemotherapy, provided longer survival times and better local control of the disease. Inadequate excision leads to high rates of metastasis and local recurrence since OS is extremely aggressive locally. The optimal excision technique is the en bloc resection of the vertebral body with tumor-free margins. However, because of the local aggressiveness of the tumor and the anatomical constraints, en bloc total resection is not always possible.

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38.2 Natural History

OS of the spine has a poor prognosis with relatively short-term survival. It ends up with death when untreated in the early term. Even with the appropriate treatment and maximum precautions, poor outcomes are not unexpected in highly aggressive tumors. Mukherjee et al. [1] reviewed the National Cancer Institute's Surveillance, Epidemiology, and End Results (SEER) database in 2011 for 1892 patients with spinal neoplasms. Their report included 430 patients with OS who underwent surgical treatment and radiotherapy. According to the results of their study, 78% of those patients had died during their SEER follow-up period and 28% developed metastasis. In agreement with Mukherjee's study, Shives et al. [2] documented 27 cases with spinal OS and found that 26 (96.3%) of the patients had died of the disease 1 to 18 months after surgery.

38.3 Physical Examination

On physical examination, inspection mostly gives no information unless there is an obvious spinal deformity, invasion to the skin, fistula, constitutional symptom, or any symptom secondary to second organ metastasis. The spinous process of the affected vertebral level may show some tenderness on palpation, particularly when there is significant posterior element invasion. Since the structural integrity and weight-bearing capacity of the vertebral body is lost, mechanical pain is provoked by anterior/posterior/side bending, spinal column rotation, and long-distance walking (Chap. 41). Neurologic functions can be impaired due to canal violation by the tumor, and it may range from single nerve root pathology to para/tetraplegia (Videos 38.4 and 38.9).

38.4 Imaging

Plain radiographs of the spine usually show a blastic lesion that occasionally appears as an "ivory body." However, OS can also present as a lytic lesion. The pure lytic pattern is seen in various subtypes, such as telangiectatic OS with predominant cystic architecture simulating aneurysmal bone cyst (ABC; Chap. 56). In 20% of the cases, mixed type may be found, while 5% has no typical radiographic features.

Computed tomography (CT) is obviously superior to plain radiographs in depicting the cortical destruction and the matrix mineralization pattern of lytic lesions which is found in about 80% of osteolytic cases.

Magnetic resonance imaging (MRI) can show the dense mineralization as a low signal intensity on all pulse sequences. Particularly in the telangiectatic OS, fluid-fluid level finding can be demonstrated on T2 sequences which differs from that of ABC with thick, solid tissue surrounding the cystic spaces and matrix

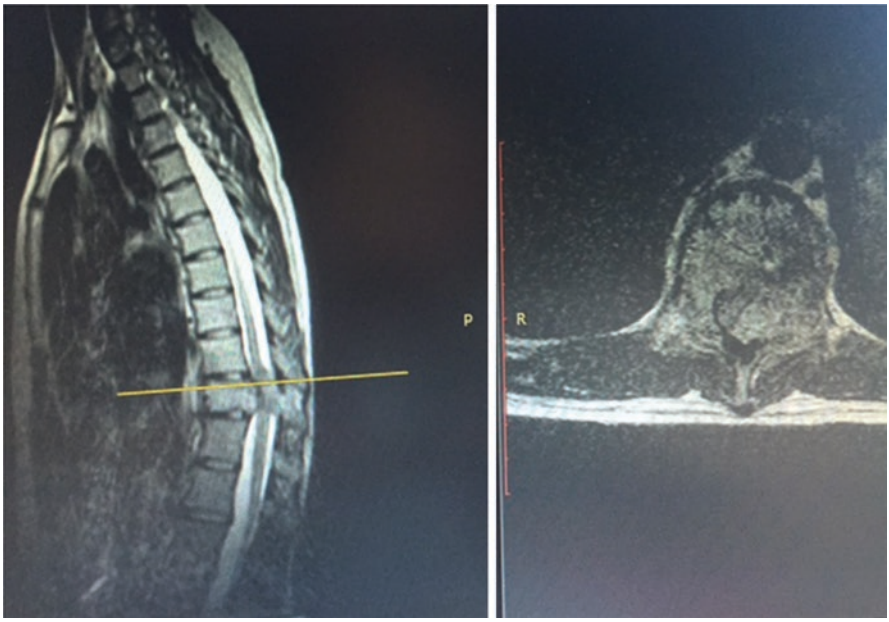

Fig. 38.1 A 19-year-old female patient's T2-weighted sagittal and axial thoracic MR images revealed a significant canal involvement by a soft tissue mass originated from vertebral body

mineralization (Chap. 56). In addition, aggressive growth patterns with surrounding soft tissue invasion, expansile remodeling, periosteal reaction, cortical destruction, associated peritumoral soft tissue mass, spinal canal violation, and pathologic fractures can be seen (Fig. 38.1).

38.5 Differential Diagnosis

Diagnosis is made with biopsy (Video 38.8). Metastatic lesion to the bone (Chap. 63), Ewing's sarcoma (Chap. 39), aneurysmal bone cyst (Chap. 56), leukemia, lymphoma, eosinophilic granuloma (Chap. 35), osteomyelitis, and spondylodiscitis (Chap. 65) and spondylodiscitis are the most common entities in differential diagnosis.

38.6 Treatment Options

Musculoskeletal sarcoma is best treated with wide resection and neoadjuvant/adjuvant therapy if the tumor histology is compatible. Currently, OS is treated with multimodality therapy including neoadjuvant therapy followed by surgery and then adjuvant therapy.

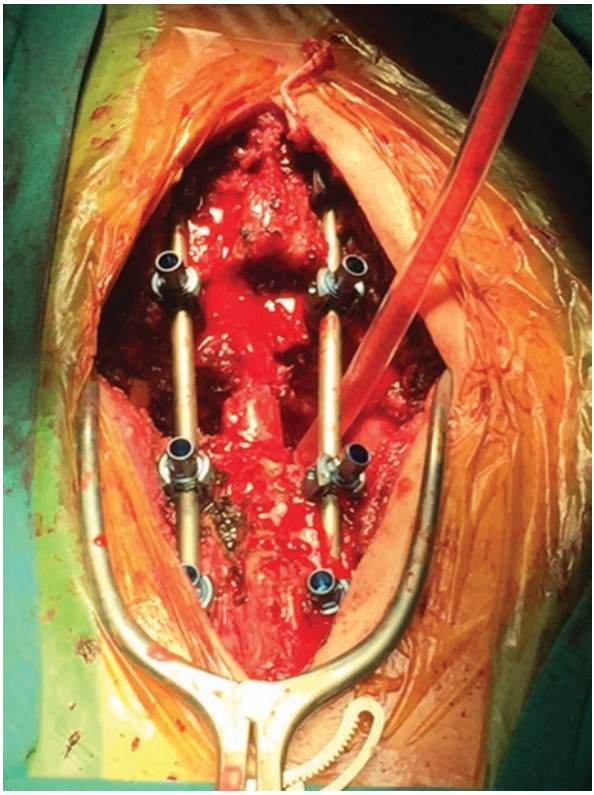

Fig. 38.2 After confirmation diagnosis with a percutaneous biopsy, en bloc vertebrectomy was done

After the diagnosis with biopsy (percutaneous or open), the primary goal of the surgical treatment is complete resection of the tumor with tumor-free margins (Video 38.8). However, complete tumor resection may cause significant local and systemic morbidity, depending on the tumor size, location, and the basal health status of the patient. Aggressive resection is known to be improving the neurological and functional status, local tumor control, and long-term survival. As well, inadequate excision leads to high rates of metastasis and local recurrence since osteosarcomas (OS) are locally very aggressive. The optimal excision technique is the en bloc resection of the vertebral body with tumor-free margins and reconstruction of the defective level(s) with an appropriate size cage (Figs. 38.2, 38.3). However, because of the local aggressiveness of the tumor and the anatomical constraints, en bloc total resection is not always possible. Radiotherapy is rarely used since OS are almost always resistant to it.

38.7 Expected Outcomes

Despite aggressive treatment, the expected outcome (survival) is frequently poor.

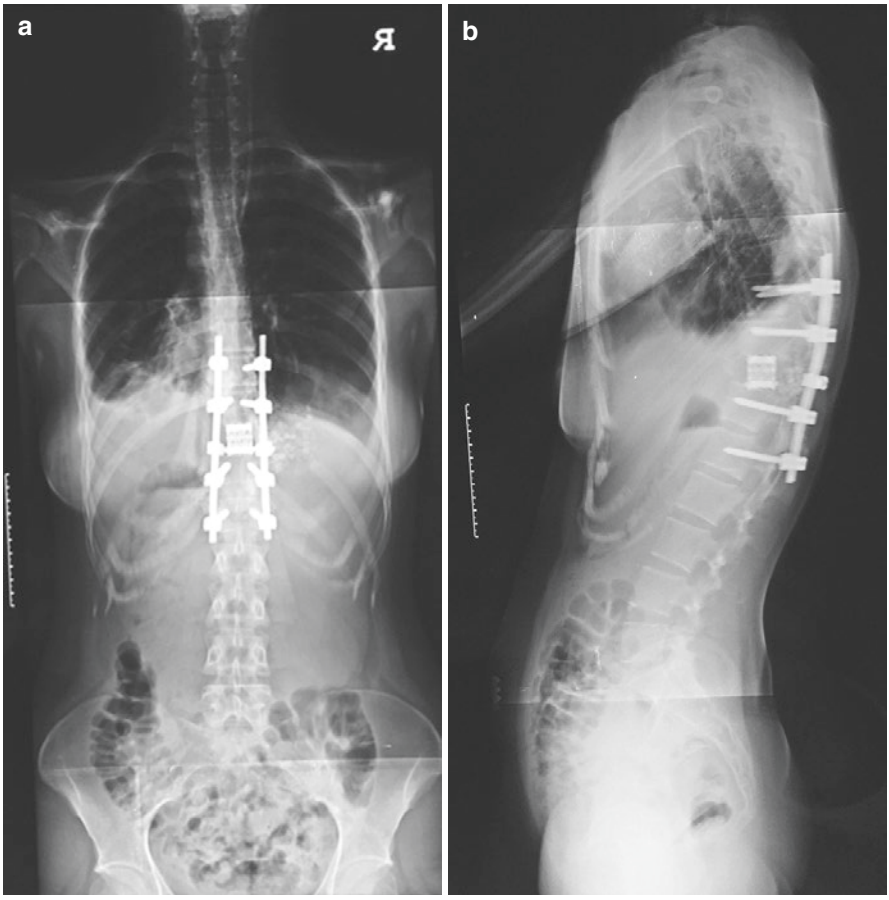

Fig. 38.3 A-P (**a**) and lateral (**b**) X-ray view after reconstruction of the vertebral column with a mash cage and posterior pedicle screw fixation

38.8 Potential Complications

Besides the natural poor prognosis and the fatal progress of the disease, there are multiple complications secondary to the aggressive treatment options which include chemotherapy, surgery, and radiotherapy. Radiotherapy is rarely used in OS as it has local side effects like surrounding healthy tissue damage, postsurgical bony fusion delay, and wound healing trouble, while chemotherapy has mostly systemic effects. Surgical intervention has several potential complications as well including spinal cord injury, dural tears and cerebrospinal fluid leakage and fistula, nonunion or delayed union, implant failure, and major vascular structure injury.

38.9 What Should Patient and Family Know?

OS of the spine has a poor prognosis. Even with appropriate treatment, poor outcome is not unfrequent. All caregivers involved in the care of patients with OS (spine surgeon, oncologist, other specialists) must warn patient and family about the prognosis of the disease and what are the available treatment options, the outcome, and the need for additional surgical procedures.

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Ewing's Sarcoma

39

Peter Pal Varga and Aron Lazary

39.1 Definition

Ewing's sarcoma (ES) is the second most common primary malignant bone tumor in children and adolescents although the spinal localization is not frequent. The tumor arises from bone marrow-derived mesenchymal stem cells, and a genetic translocation between chromosomes 11 and 22 (EWS and FLI1 genes) is found in most (>80%) cases. Extraskeletal (lung, kidney, etc.) localizations of primary ES are also possible. Therapy for ES includes aggressive multimodal therapy with chemotherapy, surgery, and radiation which allows 50% to 60% of those without metastases to achieve long-term relapse-free survival. The role of neoadjuvant chemotherapy is essential, even in a spinal case with neurological compromise, but the timing and type of surgery and dose of radiation are still questioned.

39.2 Natural History

The median age of diagnosis is approximately 15 years. The spine can be affected by the disease both as the primary site and more commonly as a metastatic progression (Chap. 63); prognosis is poorer in the latter case. Depending on the extent and localization of the primary tumor, the spinal lesion can be symptom-free and can be

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discovered during follow-up screening imaging studies of limb ES. The most common symptom of a spinal tumor is pain which can be nonmechanical tumorous pain, mechanical pain in case of spinal neoplastic instability, or pain related to neurological compression (Chaps. 11 and 41). Functional and neurological deterioration can be caused by the tumor mass or pathological fracture. Alarming symptoms are intractable pain, progressing neurological deficit (Video 39.4), cauda equina syndrome, or signs of spinal cord compression.

39.3 Physical Examination

Standard spinal physical examination of the patient is crucial to identify conditions requiring emergency surgery; however, it is rare in ES. Laboratory findings are nonspecific.

39.4 Imaging

In imaging studies, ES is an aggressive lesion with mostly mixed lytic-sclerotic pattern with large soft tissue component. Periosteal reactions like onion-skinned, spiculae, "sunburst," or Codman's triangle are rarer in spinal localization. For bony structures, computed tomography (CT) scan is more sensitive than standard radiographs, but the primary imaging modality for local staging and surgical planning is magnetic resonance imaging (MRI). Bone scintigraphy and positron emission tomography (PET) are useful to detect distant metastases which occur in 25% of all cases at the time of the initial diagnosis. Regular imaging studies (MRI, CT) are advised during the follow-up postoperatively to assess local control.

39.5 Differential Diagnosis

Tumorous lesions and tumor-like lesions of the spine are the most common differential diagnostic issues. Following the first and most important oncological principle, namely, "tissue is the issue," the cornerstone of the differential diagnosis is the histological examination of the lesion. In case of a primary spinal tumor, imaging-guided percutaneous biopsy or open biopsy can provide adequate tissue for the detailed histopathological studies (Video 39.8).

39.6 Treatment Options

Treatment of ES is multimodal. Except for emergency cases, where an urgent decompression of the spinal cord can be required, the treatment process is planned by the oncologist and started with chemotherapy (vincristine, doxorubicin, and cyclophosphamide; VDC), alternating with ifosfamide and etoposide (IE) or VDC/

IE. Surgical resection of the remnant tumor mass and radiotherapy are crucial to achieve local control. Based on Enneking's principles, a wide en bloc resection of the high-grade malignant bone tumor would be required for the optimal oncological outcome; however, it is extremely difficult to achieve such outcome in spine ES without dramatic functional loss. On the other hand, intralesional resection of the tumor is associated with a higher rate of local recurrence and shorter survival. The recommended surgical treatment for such lesions is an en bloc resection with wide or marginal margins, if technically possible, followed by radiotherapy. It requires advanced technical skills and experience in spinal tumor surgery; as it; these patients must be treated in spinal tumor centers. Meticulous surgical planning is mandatory: resection, stabilization/reconstruction, and soft tissue procedures must be meticulously planned. Involvement of professional partners—as vascular surgeon, chest surgeon, and plastic surgeon—can be required depending on the localization of the ES and the surgical plan. Ideally, treatment should be done in a few dedicated centers of the country gathering all competencies in one unique place (Fig. 39.1).

39.7 Expected Outcomes

Survival in spinal ES is poorer compared to extremity cases. A 5-year survival is about 60% in spinal ES, and it is related to the surgical resection (according to Enneking's principles) and timing of chemotherapy. The local recurrence rate is high, about 30% within 5 years after the index procedure. Local recurrence is associated with intralesional margins and previous tumor surgery. The functional outcome depends on the neurological functional loss caused by the tumor itself, the consequence, or the complication of the surgery. Loss of spinal stability can result in chronic, progressing pain decreasing the function and quality of life of the patient.

39.8 Potential Complications

The most frequent perioperative complications are wound healing problems, deep surgical site infections, and neurological deterioration. Long-term complications are implant loosening and development of secondary spinal instability.

39.9 What Should Patient and Family Know?

ES is a high-grade malignant disease, where survival is relatively poor also in case of effective multimodal therapy. Survival, local recurrence rate, and functional outcome (quality of life) are related to the outcome of surgery (proper resection, margins); however, performing adequate surgery is frequently challenging and is characterized by a high complication rate. The proper oncological treatment before and after the surgery is essential.

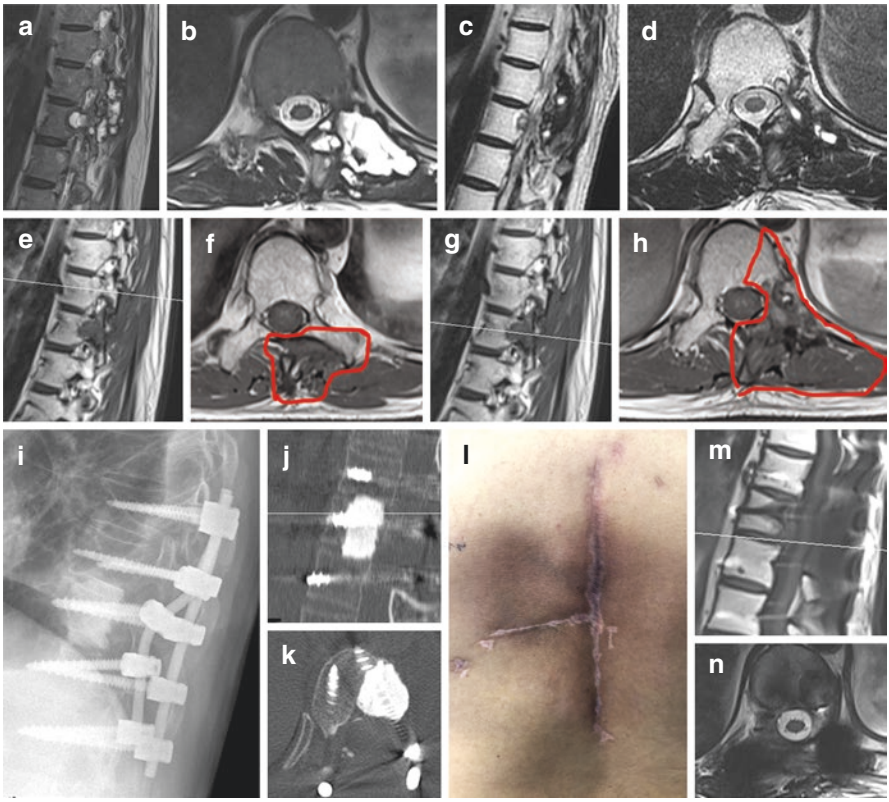

Fig. 39.1 Thoracic ES surgically treated with en bloc resection, stabilization, and reconstruction. The 48-year-old male patient was operated 3 years ago because of a left humerus ES. Follow-up imaging showed a thoracic tumor, affecting the Th9–11 level causing mild pain without neurological deficit nor spinal instability (**a**, **b**). Neoadjuvant chemotherapy and irradiation resulted in the shrinking of the tumor mass (**c**, **d**). Planning of the Enneking's appropriate en bloc resection (**e**–**h**) and stabilization. Postoperative X-ray after the surgery (**i**). A PMMA bone cement spacer anchored to the posterior rod was used to reconstruct the anterior column (**j**, **k**). A "sliding" latissimus flap was used for soft tissue reconstruction that resulted in perfect wound healing (**l**). The surgery and adjuvant radiation therapy provided local control (**m**, **n**—1-y follow-up)

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Discitis in Pediatric Spine

40

Yat-Wa Wong

40.1 Definition

Discitis, vertebral osteomyelitis, and spondylodiscitis refer to the infection of the intervertebral disk, vertebral body, and bony vertebrae with the intervening disk, respectively. They are the spectrums of spinal infections that are named according to the primary site of infection. However, pure discitis is rare because most infections start in the metaphyseal region of the vertebral body as a result of rich blood supply. Pediatric spinal infections can also be divided into pyogenic (most common), granulomatous, fungal, or parasitic origins.

40.2 Natural History

Pediatric spinal infections are relatively rare [1]. It is notoriously difficult to catch the causative microorganisms, and inflammatory markers do not usually elevate to high levels. Some patients may even recover spontaneously. Therefore, in the past, many clinicians did not believe the existence of discitis in children. Missing the diagnosis may sometimes be catastrophic. The mean delay in diagnosis is 28 weeks [2].

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40.3 Physical Examination

Infants or very young children cannot present their symptoms well, and it imposes diagnostic difficulties. Isolated spinal infections in infants are rare, but the isolated spinal infection may be a part of disseminated sepsis. Although toddlers and young children may complain of back pain, they usually present with irritability and refusal to walk. Many such patients are afebrile on admission. Neurological deficit is uncommon until at a late stage (Video 40.4). The diagnosis of spinal infections in older children and adolescents is similar to that in adults, because older children and adolescents can present their symptoms well and have a higher incidence of fever. Non-pyogenic spinal infections tend to have a subacute or chronic course. In the tuberculous (TB) spine, the children may present with round or angular kyphosis, torticollis, dysphagia, stridor, or respiratory obstructions due to huge cold abscesses.

40.4 Imaging

Radiographic changes may not be apparent in the first two weeks except for soft tissue edema. Typical radiological findings of spondylodiscitis, regardless of pyogenic or TB infections, are narrowing of the intervertebral disk space and adjacent vertebral end plate destruction. For spondylitis, the only destruction of the vertebral body with a normal looking intervertebral disk is seen. Cold abscesses of TB spine appear as paraspinal soft tissue shadow, but patients are typically not septic looking. Other radiological features are described in the chapter on TB spine (Chap. 63). Magnetic resonance imaging is the most sensitive in diagnosing spinal infections. Its sensitivity and specificity are approximately 96% and 93%, respectively. For patients with atypical presentations, FDG-PET may distinguish infections from neoplasms.

40.5 Differential Diagnosis

For pyogenic spinal infections, an elevation of white blood cell count, CRP, and ESR was only found in 41%, 57%, and 86% of the cases, respectively [2]. Positive blood culture only appears in 8% of the cases. The positive yield from spinal biopsies or aspirations is 40%. With such a low detection rate, some clinicians recommend empirical antibiotics and reserving image-guided biopsies (Videos 40.7 and 40.8) or aspirations for patients who do not respond to empirical antibiotics, who may be infected by atypical microorganisms, or who have features of tumoral lesions.

Staphylococcus aureus is the most common cause of pediatric infections for all age groups. Spinal infections in the first 6 months of life are rare because of the

inherited immunity from the mother. *Staphylococcus aureus* accounts for 80% of pyogenic spinal infections in infancies less than 6 months old. Other organisms include coagulase-negative *Staphylococcus*, alpha-hemolytic *Streptococcus*, *Streptococcus pneumoniae*, and Gram-negative rods. For children between half a year and 4 years old, clinicians should specifically look for *Kingella kingae*, which is very difficult to be found by traditional culture. Using aerobic blood culture vials or polymerase chain reaction (PCR) for biopsy specimens is recommended. The diagnosis is further supported if throat swabs were positive for *Kingella kingae*. In the regions where tuberculosis is endemic, an elevation of the procalcitonin points to a pyogenic infection, since its level should be normal or only slightly increased in a TB spine.

40.6 Treatment Options

In principle, antibiotic therapy should be given according to the sensitivity tests of the causative agent. However, the positive detection rate of blood culture and biopsy is low. Empirical broad-spectrum antibiotics can be given while waiting for investigation results, and the patient's response should be closely monitored. For community-acquired infections, cloxacillin can be used for young children below 6 months old, since the most common organism is MSSA. Amoxicillin-clavulanate is used when the patients are between 6 months and 4 years old, because it covers both MSSA and *Kingella kingae*. Covering *Kingella kingae* is important if PCR on throat swabs is positive. For hospital-acquired infections or immunosuppressed patients, empirical antibiotics, such as vancomycin and third-generation cephalosporin against MRSA and Gram-negative microorganisms, are necessary. If the patients have a history of visiting farms or consuming unpasteurized milk products, rifampicin or trimethoprim-sulfamethoxazole are antibiotics of choice. There is no standard recommendation on the duration of antibiotics for pyogenic infection, but in general the antibiotics should be given until serum infective markers return to normal, and there should be a minimum duration of three weeks.

For TB spine, four anti-TB drugs (isoniazid, rifampicin, pyrazinamide, and ethambutol) are given for the first 8 weeks. If the patients are responding well, or the sensitivity test confirms effective isoniazid and rifampicin, they should be continued for a total of 9 months, and the other two anti-TB drugs can be stopped. Children with TB spine after adequate antituberculosis treatment should still be carefully followed up for any progression of kyphosis, particularly if multiple apophyseal rings of vertebral bodies are destroyed. This may lead to the progression of kyphosis during growth. Rajasekaran described four radiological risk features with which patients may develop progressive kyphotic deformity [3] (Fig. 40.1).

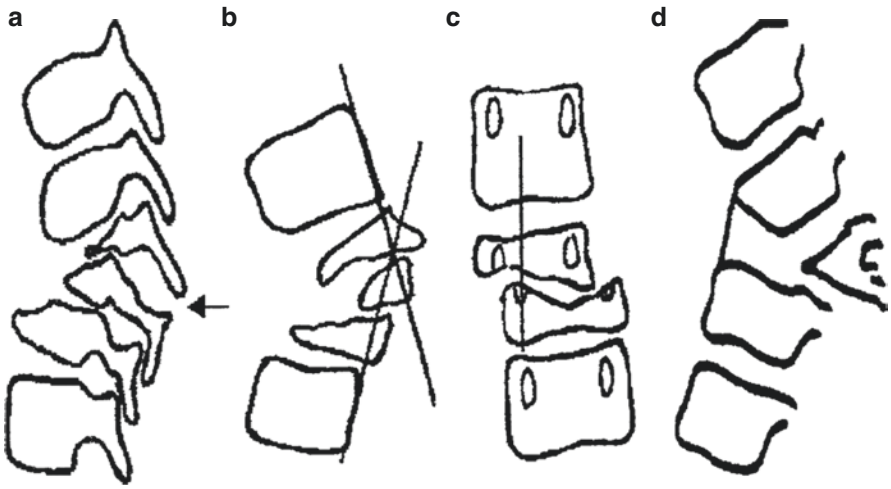

Fig. 40.1 The schematic drawings [3] from Dr. S. Rajasekaran (with permission): (**a**–**d**) illustrating the radiological signs of the high risk of progression of kyphosis. (**a**) Separation of facet joint at the apex. (**b**) Retropulsion of the intervening vertebral bodies posterior to two lines along the posterior surfaces of the first upper and lower normal vertebrae. (**c**) Lateral translation between the first lower normal vertebra and the first upper normal vertebra. (**d**) Toppling sign. When a line drawn along the anterior surface of the first lower normal vertebra intersects higher than the middle of the anterior surface of the first normal upper vertebra

40.7 Expected Outcome

Most pyogenic spinal infections respond to antibiotics alone. It is only very occasionally that surgery is indicated for neurological deterioration. Correction of severe deformity is mandatory in TB spine to prevent Pott's paraplegia of late onset. Surgery for persistent symptoms not responsive to anti-TB treatment also gives favorable short- and long-term results (Fig. 40.2).

In case of established kyphotic deformity with persistent infected tissues or abscess, anterior debridement and fusion together with posterior instrumented fusion are indicated.

40.8 Potential Complications

Disseminated pyogenic infections, paraplegia, or spinal instability.

40.9 What Should Patient and Family Know?

Compliance with antibiotics or anti-TB treatment.

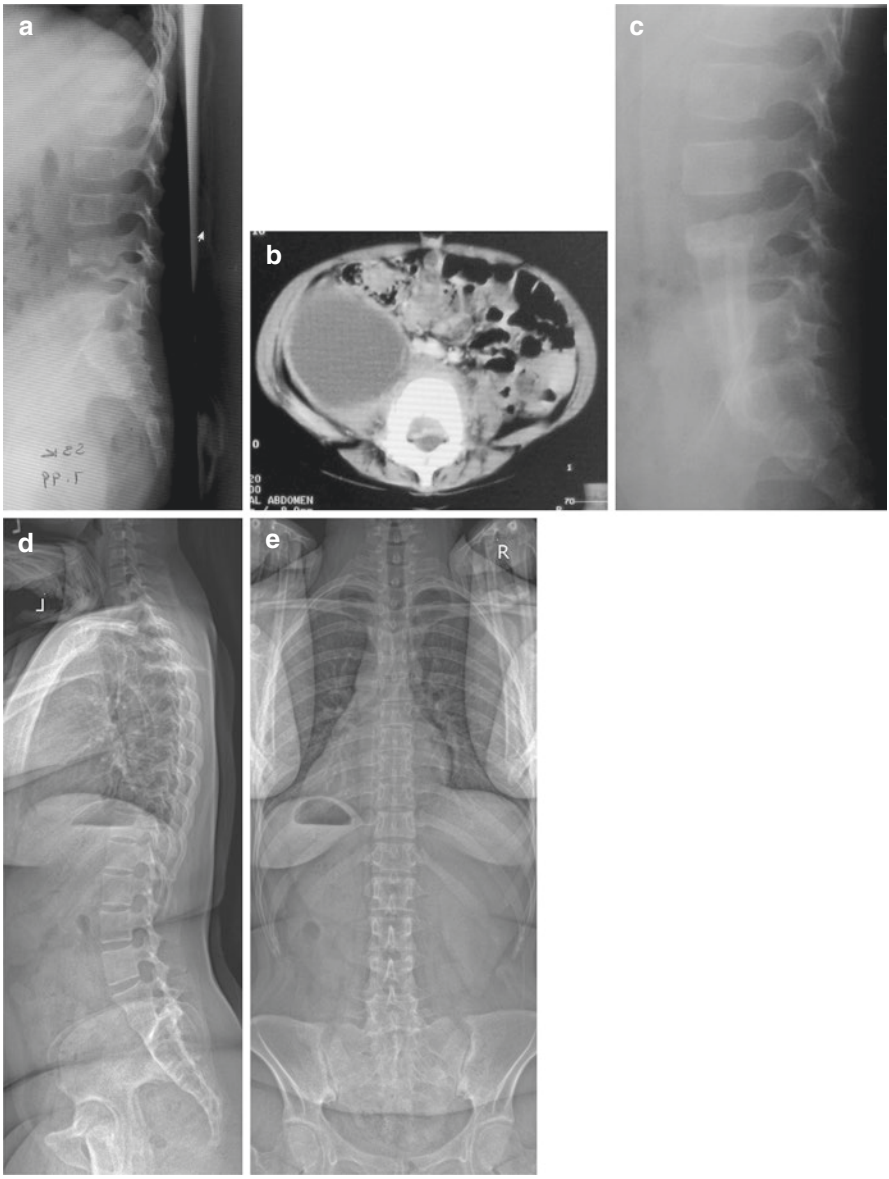

Fig. 40.2 This 6-year-old patient suffered TB spine. (**a**) Lateral X-ray showing the complete collapse of L5 and partial collapse of L4. (**b**) Axial CT scan demonstrating a huge right psoas abscess. (**c**) Lateral X-ray after the anterior debridement and fusion L3 to S1 using a fibular graft. (**d**, **e**) Standing whole spine X-ray 20 years after surgery. The global spinal alignment was well maintained

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Part IV

Adult Spine Pathology

Back Pain and Its Generators

41

Philip K. Louie and Todd J. Albert

41.1 Definition

Back pain is one of the most common health complaints among adults, resulting in pain and disability. Musculoskeletal pain in adults ranges from 65% to 85% with 36% to 70% suffering from back pain. The 1-year prevalence of back pain in seniors (aged 65+) ranged globally from 13% to 50% in population-based studies. Up to 80% of older residents in long-term care facilities experience substantial back pain which is often underreported and inadequately treated.

41.2 Physical Examination

Research has shown that less than 50% of primary care physicians have strong confidence in diagnosing the causes of chronic back pain in adults (Appendix N). A stepwise approach to history collection can lead to a cost-effective and cost-efficient process of treating the patient. The clinician must determine whether the pain is

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Table 41.1 Physical exam: mechanical versus nonmechanical and axial versus radicular

Mechanical	Nonmechanical
• Rest improves symptoms • Pain often worsens progressively throughout the day	• Rest/immobilization does not improve symptoms • Pain is independent of activity and can be worse at night
Axial	**Radicular**
• Diffuse • Referred pain patterns include: – Cervical spine: scapula or shoulder – Lumbar spine: buttock or posterior thigh	• Dermatomal distribution • Can present with paresthesia, numbness, or weakness • Pain can be associated with tension signs

Table 41.2 Inspection

Coronal plane evaluation	Sagittal plane evaluation
– Scoliosis – Pelvic obliquity – Shoulder imbalance – Scapular protuberance – Rib prominence	• Note normal spinal curves – Cervical lordosis: 20–40° – Thoracic kyphosis: 20–45° – Lumbar lordosis: 40–60°

Table 41.3 Palpation

Bones	Soft Tissue
– Spinous processes – Posterior superior iliac spines – Scapula and ribs – Iliac crests – Sacrum and coccyx – Trochanter – Ischial tuberosity	– Trapezius muscle – Rhomboid/levator muscles – Gluteus muscles – Piriformis muscle – Sciatic nerve

mechanical or nonmechanical and axial or radicular (Tables 41.1, 41.2, 41.3). Paresthesia, numbness, or weakness that follows a dermatomal distribution and is associated with tension signs is more indicative of radicular pain. A comprehensive physical exam should include inspection, palpation, movement, neurological examination (Video 41.4), and other special exams. While assessing for coronal or sagittal imbalance, also evaluate for walking aids that the patient may use for mobility.

41.3 Imaging

Table 41.4 outlines the indications for imaging during the workup of back pain vary (Table 41.4). Standing plain radiographs of the thoracic and/or lumbar spine are the initial diagnostic test of choice in the setting of trauma, malignancy, infection, deformity, and degenerative spine disease and offer many benefits due to ease in the acquisition and relatively low cost. However, plain radiographs are limited in the

Table 41.4 Indications for imaging of the thoracolumbar spine

Plain radiographs	MRI	CT
First diagnostic test of choice in the setting of trauma, malignancy, infection, deformity, and degenerative spine disease	Radicular symptoms (pain, weakness, and numbness/tingling which radiates to the extremities)	Concern for fracture
Back pain that does not improve with conservative measures for more than 6 months (all ages)	Myelopathic signs and symptoms (altered gait, difficulty with fine motor tasks, hyperreflexia, upper motor neuron signs)	History or concern for present malignancy
Evaluate for dynamic instability of the spine	Neurogenic claudication	Assess bone mineral density
Assess overall coronal and sagittal alignment (concern for deformity)	Bladder dysfunction (usually urinary retention or overflow incontinence) with leg pain and weakness	Assess status of fusion
Evaluate for arthritic changes in neighboring joints (sacroiliac, hips, etc.)	History or concern for present malignancy	Preoperative planning
	Infection (both rule out and assess current status/progression)	Acute postoperative evaluation of hardware placement
	If the back pain is accompanied by constitutional symptoms (such as loss of appetite, weight loss, fever, chills, shakes, or severe pain when at rest)	*Myelography:* diagnostic test of choice in patients unable to receive an MRI; provides additional diagnostic evaluation for foraminal/lateral recess stenosis and disk lesions

ability to identify soft tissue, three-dimensional characteristics, and high image resolutions. Plain radiographs can also evaluate stability and flexibility in the form of flexion-extension, sitting, supine, and bending views to provide dynamic feedback.

Magnetic resonance imaging (MRI) is the diagnostic procedure of choice for most primary spinal pathologies. MRI provides relatively high sensitivity and specificity for infections, tumors, disk degeneration, pathologic fractures, and herniations. Some visceral pathology may also be initially evaluated. However, MRI is relatively expensive and has varying degrees of utility in obese, claustrophobic, and pacemaker-dependent patients.

Computed tomography (CT) provides a more detailed evaluation of the osseous structures of the spine, largely due to its ability to reconstruct three-dimensional and multiplanar images. Although not as commonly utilized at this point, CT myelography has some utility to improve the diagnosis of foraminal/lateral recess stenosis and disk lesions, especially in patients that cannot undergo MRI.

Discography has largely fallen out of favor due to controversial evidence and risks associated with the procedure, specifically iatrogenically induced disk degeneration.

Angiography provides clear visualization of specific structures due to variations in vascular flow. This form of imaging is particularly useful for the characterization of many spinal tumors and can be applied preoperatively to evaluate vascular anatomy.

41.4 Differential Diagnosis

While the vast majority of nonspecific complaints can be self-limited and treated conservatively, notable findings can present that aid with diagnosis and may drastically alter the time course of evaluation and treatment, such as weight loss, bowel or bladder incontinence, saddle anesthesia, weakness, or gait imbalance. A differential diagnosis of common back pain etiology is listed below.

Nonspecific or mechanical low back pain: Often multifactorial, disk degeneration, facet joint pain, spondylolisthesis, spondylosis, sacroiliac joint disorders, and myofascial sources.

Radiculopathy or neurogenic claudication (compression of nerve roots): Disk herniation, lumbar spinal stenosis from secondary to degenerative changes (hypertrophic ligamentum flavum, facet arthropathy, disk-osteophyte complexes), and epidural lipomatosis.

Facet joint syndrome: Can also be a source of chronic low back pain in older individuals. As the degenerative process in the spine progresses, kinematics and load transmission throughout the motion segments are impacted, which can stimulate nociceptive fibers in the facet joints. Studies have not found reliable referral patterns of facet joint pain. Symptoms can range from local soreness to "pseudoradicular" pain that typically radiates uni- or bilaterally along with lumbar nerve root distributions without neurological deficits.

Trauma/vertebral body fractures: Can be a result of varying impact levels of trauma or poor bone mineral density.

Scoliosis (previous adolescent idiopathic scoliosis or de novo degenerative lumbar scoliosis): The asymmetric loading on vertebral end plates on the concavity of the curve may elicit inflammatory responses in the end plate and adjacent bone marrow of the vertebral body.

Tumors: Common metastatic sources of LBP are prostate and kidney; however, primary malignant tumors are also found in older adults (plasmacytoma, lymphoma, or chordoma). Primary benign tumors (e.g., aneurysmal bone cysts, eosinophilic granuloma, osteochondroma, osteoma, and osteoblastoma). Clinically, the patient can describe progressive, unremitting, localized, or radiating pain aggravated by activity, worse at night, and does not improve with rest.

Visceral disease: Several visceral diseases (e.g., urinary tract infection, prostatitis, dissecting abdominal aortic aneurysm, pelvic inflammatory disease, cholecystolithiasis, and nephrolithiasis) can present as back pain.

Spinal infection: Bacteria may be disseminated hematogenously from a distant infected source and multiply at the metaphyseal arterioles of vertebral bone that causes abscess formation, bone necrosis, and fistula within the bone. Specifically, aerobic gram-negative bacilli in men with urinary tract infection may travel to the lumbar spine through Batson's plexus. Rarely, tubercular osteomyelitis may occur in those who have contracted tuberculous infection earlier in life.

41.5 Treatment Options, Expected Outcomes, and Potential Complications

Treatment options are based on the etiology of the back pain. Ultimately, improper treatment or nontreatment of adults with back pain may result in impeded cognition, malnutrition, withdrawal from social and recreational activities, sleep disturbances, psychological distress, rapid deterioration of functional ability, and falls. Thus, a comprehensive diagnostic workup is recommended, especially in presentations that do not directly correlate with symptoms, exam findings, and early imaging results.

The surgical treatment options for spine-specific pathology will be discussed at length throughout this book. There are several well-established algorithms and evidence-based approaches for surgical treatment. However, conservative management for nonspecific back pain can be a bit challenging and nuanced (Table 41.5).

Table 41.5 Treatment options

Treatment option	Description
Activity modification	Common recommendation is to avoid activities that cause pain (until acute symptoms decrease), but evidence is lacking. Bed rest no longer indicated. Patients with chronic low back pain should be encouraged to remain physically active
Exercise/physical therapy	The general consensus is that low-impact cardiovascular and aerobic exercises provide other benefits (as no significant impact on clinical outcome), such as improved mood, increased pain tolerance, and prevention of deconditioning. Symptoms can be improved by a focus on aerobic fitness, restored normal lumbosacral motion, strengthening of trunk muscles, and emphasis of correct body mechanics
Specific modalities	Physical therapists and chiropractors often provide modalities that include cold packs, superficial heat, short-wave diathermy, and ultrasound
Education	Provide information regarding correct posture, biomechanics of the spine with activities of daily living, and simple methods that can reduce symptoms. Inform patients on the expected outcome and favorable natural history of low back pain
Manipulation/traction	There is no evidence that spinal manipulative therapy or traction is superior to other treatments of patients with acute or chronic low back pain

(continued)

Table 41.5 (continued)

Treatment option	Description
Acupuncture	Overall effectiveness remains unclear. Not recommended as a first-line treatment for back pain. May be a small part of a comprehensive program in selected patients with chronic back problems but should not be used in lieu of established effective treatment methods
Medications (analgesics, antidepressants, corticosteroids, muscle relaxants, narcotics, non-narcotics, nonsteroidal anti-inflammatory, topical)	Consideration of medication class, dose, and duration is important. Potential side effects and risk for dependence
Injections (epidural, facet joint, sinuvertebral, trigger point, sacroiliac)	Therapeutic injections without a reasonable presumptive diagnosis should not be provided. Otherwise, the listed injections should target specific presumptive diagnosis. Oftentimes, the injections can serve diagnostic and therapeutic purposes. Patients should be instructed on recording the degree/percent of symptom improvement as well as the duration
Orthoses (brace, corset)	There is no evidence that supports the effectiveness of orthoses in the treatment of acute and chronic low back pain. Orthoses may act as proprioceptive reminders to correct spine mechanics during lifting and bending activities
Transcutaneous electrical nerve stimulation	Conflicting evidence supports the use of TENS as treatment of acute and chronic low back pain

41.6 What Should Patient and Family Know?

Back pain and its generators are incredibly multifactorial. As outlined in this chapter, the literature suggests that the prevalence of severe and chronic back pain increases with age. Various age-related physical and psychological changes as well as multiple medical risk factors may affect the prognosis and management of back pain in adults. Constant research is being performed to better understand the impacts of various factors on the assessment and treatment of back pain as clinicians are working toward evidence-based evaluation and treatment that is personalized to the patient and their specific presentation.

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Cervical Degenerative Disc Disease (Including Cervical Disc Herniation)

42

Michael H. McCarthy and Todd J. Albert

42.1 Definition

Cervical degenerative disc disease refers to age-related changes within the intervertebral disc space typically found in tandem with the facet and uncovertebral joint degeneration resulting in spondylosis. Symptoms of cervical spondylosis manifest as axial neck pain, radiculopathies, and cervical myelopathy.

42.2 Natural History

Cervical disc degeneration is common among patients presenting with axial neck pain and upper extremity symptoms, especially within the elderly population. Loss of cervical disc height results in bulging of the annulus, infolding of the ligamentum flavum, and facet hypertrophy reducing the volumetric area of the spinal canal and foramen. Disc desiccation and collapse occur concomitantly with facet arthrosis contributing to focal and multifocal cervical spondylosis. Structural changes within disc's proteoglycan typically occur within the third decade of life diminishing the hydration of the disc; specifically, keratin sulfate increases and chondroitin sulfate

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decreases altering the viscoelasticity [1]. Focal and multilevel disc degeneration can cause neurologic impairment and deformity, most commonly with progressive loss of cervical lordosis leading to kyphosis. Degenerative changes within the nucleus pulposus and annulus fibrosus predispose patients to disc herniations. Disc degeneration can lead to a cascade of arthritic changes within the intervertebral space, uncovertebral joints, and facets. Neurologic impingement within the cervical spine can manifest as upper extremity radiculopathies, myelopathy, and a combination of both termed myeloradiculopathy.

42.3 Physical Examination

After a thorough history, the physical exam should entail a comprehensive assessment of the location of pain, gait, neurological exam, and cervical-specific maneuvers. The neurological examination must include a detailed motor exam, assessment of reflexes (Table 42.1), and a sensory exam (Videos 42.4 and 42.9). Motor evaluation should assess upper and lower extremities with the evaluation of each muscle group supplied by its respective nerve root (Table 42.2). Unilateral sensory or motor deficits within a specific nerve root are indicative of a radiculopathy, whereas bilateral weakness or sensory disturbances, hyperreflexia, gait instability, and pathologic reflexes are more commonly found in the setting of myelopathy. Spurling's test, head in extension and lateral bending with applied axial compression, and

Table 42.1 Reflexes

Nerve root	Reflex
C5	Biceps
C6	Brachioradialis
C7	Triceps
L4	Patellar
S1	Achilles

Table 42.2 Myotomes

Nerve root	Myotome
C5	Shoulder abduction/elbow flexion
C6	Elbow flexion/wrist extension
C7	Elbow extension/wrist flexion
C8	Finger flexion
T1	Finger abduction
L2	Hip flexion
L3	Knee extension
L4	Ankle dorsiflexion
L5	Hallux extension
S1	Ankle plantar flexion

Lhermitte's sign, "shock-like" sensation with passive cervical flexion, are a few examples of sensitive exam maneuvers delineating cervical radiculopathy and myelopathy, respectively. Additional exam findings, such as Hoffman's and Babinski signs, provide further insight on the impact of the offending pathology and are useful in gaining a thorough understanding of the patient's neurologic status.

42.4 Imaging

Radiographic imaging of the cervical spine is a quick and cost-effective initial study for patients presenting with neck pain, radiculopathy, and myelopathy (Chap. 43). Radiographs can offer critical assessments of alignment, degree of spondylosis, and/or possible trauma. Anterior-posterior and lateral views provide a static understanding of the state of the cervical spine; supplementary views in flexion and extension can be beneficial when there is a concern for dynamic instability. Additionally, radiographs are useful in the initial assessment of disease states such as diffuse idiopathic skeletal hyperostosis (DISH) (Fig. 42.1) (Chap. 54) and ossification of posterior longitudinal ligament (OPLL) (Fig. 42.2), which are better defined with computed tomography (CT). CT is the imaging of choice to assess bony architecture providing granular information on fused or potentially fused intervertebral segments and compressive osseous pathology. Magnetic resonance imaging (MRI) is an excellent imaging modality to assess neural elements, intervertebral disc, and

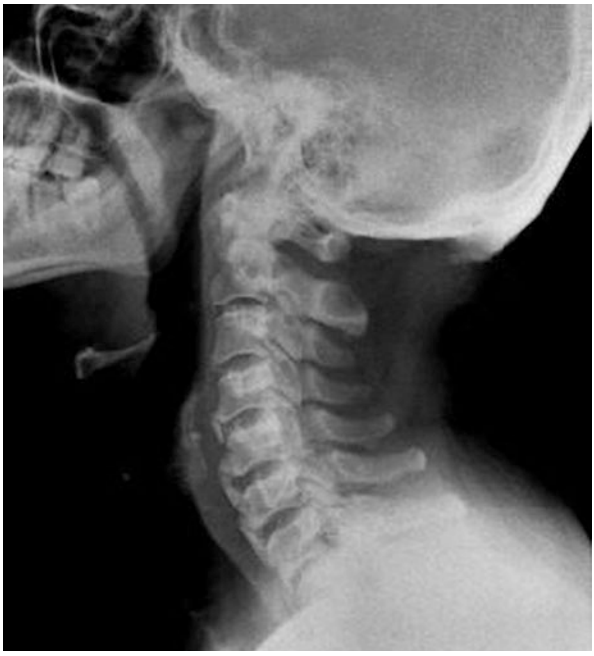

Fig. 42.1 Diffuse idiopathic skeletal hyperostosis (DISH)

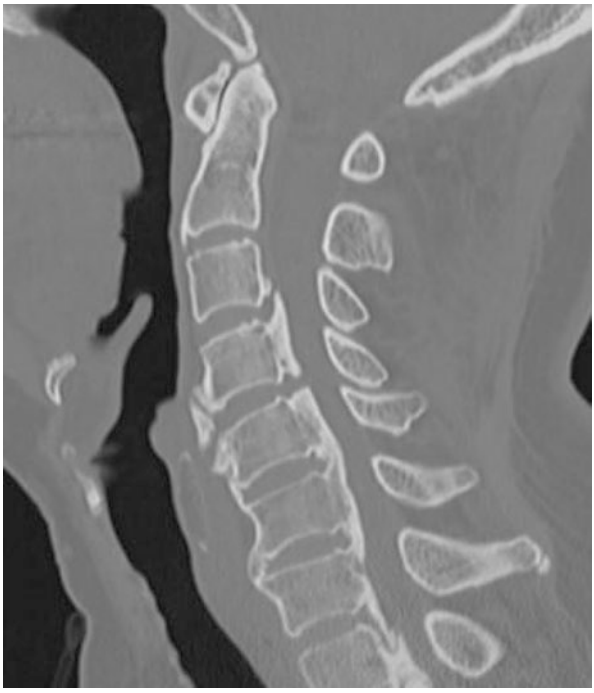

Fig. 42.2 Ossification of posterior longitudinal ligament (OPLL)

ligamentous structures and is highly sensitive at detecting degenerative changes, such as bulging discs and cervical spondylosis. It is important for providers to recognize the high prevalence of degenerative changes on MRI among asymptomatic patients. Therefore, it is imperative that diagnostic imaging be utilized as an adjunct to the history and clinical examination.

42.5 Treatment Options

Cervical degenerative disc disease is a continuum from focal disc herniation leading to radiculopathy and/or myelopathy to multilevel cervical degeneration causing sagittal malalignment. Treatment plans should be individually tailored targeting clinically relevant pathology in order to optimize outcomes. Patients with cervical myelopathy commonly present with stepwise deterioration intermixed with static periods of unchanged symptoms. Overall prognosis is highlighted by progressive compression and dysfunction of the spinal cord leading to gradual deterioration in functional status over time. Surgical intervention, in the form of decompression with possible fusion, is commonly required to halt the progression of disease and preserve function. Non-myelopathic cervical patients, specifically those presenting with radiculopathies, typically have a favorable prognosis with the majority noting improvement with nonoperative care. Nonoperative treatment modalities for cervical radiculopathies include nonsteroidal anti-inflammatory drugs, oral

corticosteroids, and cervical steroid injections. Surgeons should consider the constellation of preoperative symptoms, including the degree of spinal cord dysfunction, duration of symptoms, and general health of the patient, prior to proceeding with surgery. Surgical treatments primarily focus on decompression of neurologic elements with possible fusion if there is a disruption of the spinal column. Situations addressing sagittal malalignment may require osteotomies with multilevel instrumentation and arthrodesis. Specific treatments of degenerative disc disease, such as anterior cervical discectomy and fusion (ACDF) or cervical laminectomy and fusion (Videos 42.2 and 42.7), are predicated on the location of compression and the surgeon's decision regarding optimal construct.

42.6 Expected Outcomes and Potential Complications

Goals of intervention in the setting of degenerative disc disease are twofold, decompression of the spinal cord and/or roots and maintenance or reestablishment of normal sagittal alignment. Typically, successful surgical outcomes are more common among patients with duration of symptoms less than 1 year, younger age at presentation, pathology limited to fewer vertebral segments, and the presence of unilateral symptoms. Results of anterior cervical surgery are highly favorable both in terms of providing durable relief of symptoms and rates of fusion. Although fusion rates tend to decrease with multilevel ACDFs, these surgeries, when performed on appropriately selected patients, are highly reliable treatment options for cervical radiculopathy and myelopathy. Posterior decompression and/or fusion is considered for patients with multilevel cervical myelopathy, OPLL, kyphotic deformities, or congenital stenosis. Laminoplasty, laminectomy alone, and laminectomy and fusion offer a variety of surgical options in treating multilevel disease while achieving neurologic decompression. Surgeons should consider preoperative cervical global alignment prior to performing a posterior-only decompression since kyphotic alignment is associated with poor outcomes. Additional consideration should be given to those patients at risk of developing post-laminectomy kyphosis. Combined anterior and posterior surgeries are thought to provide rigid stabilization through a circumferential fusion affording surgeons correction of alignment and decompression of the spinal cord. These surgeries tend to entail greater blood loss, longer operative times, and overall increases in morbidity.

42.7 What Should Patient and Family Know?

Degenerative changes within the cervical spine are consequences of aging with the majority of the population remaining asymptomatic. Patients with cervical disc degeneration and spondylosis present with a myriad of symptoms most commonly grouped as radiculopathies, myelopathies, and/or axial neck pain. A thorough knowledge of the natural history of disease and determining the level of pathology through clinical findings and advanced imaging are critical in determining ideal

treatments. Treatment of radiculopathy is centered around nonsurgical interventions with surgical decompression and stabilization reserved for those with persistent symptoms despite conservative measures. Clinically evident myelopathy requires close monitoring and possible surgical intervention to prevent the progression of disease and maintain functional status.

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Conflict of Interest: The authors declare no conflict of interest.

Further Reading

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Cervical Spondylotic Myelopathy

43

Yoshiharu Kawaguchi

43.1 Definition

Cervical spondylotic myelopathy (CSM) is a degenerative disease caused by cervical cord compression that is associated with clinical symptoms. In the degenerative stage, spinal canal narrowing is caused by intervertebral disk bulging, posterior bony spur, deformity of uncovertebral joints, and hypertrophy of facet joints and the ligamentum flavum. Based on these pathologies, spinal cord compression occurs. Patients with CSM exhibit symptoms, such as clumsiness or a loss of fine motion skills in the hand, gait disturbance (spastic gait), and vesicourethral disturbance.

43.2 Natural History

CSM is usually a slowly progressive disease. The severity of symptoms gradually increases after the initial onset. In general, 5% of patients display rapid symptom onset followed by a long period of quiescence, 20% of patients exhibit gradual but steady progression of signs and symptoms, and 75% of patients exhibit stepwise deterioration of clinical function with intervening variable periods. An effective conservative treatment has not yet been established.

Supplementary Information The online version contains supplementary material available at [https://doi.org/10.1007/978-3-030-80356-8_43].

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43.3 Physical Examination

Cervical myelopathy is characterized by the presence of segmental signs and long tract signs. Segmental signs indicate lower motor findings that occur at the level of the compressive lesion. Long tract signs describe upper motor findings that are observed below the lesion. Neurological findings vary depending on the level and nature of compression (Video 43.4). The finding of "myelopathy hand" strongly suggests the existence of cervical myelopathy. The finger escape sign and rapid grip and release test are important for judging the presence of myelopathy hand. The finger escape sign is positive when the patient does not keep all digits in adduction and extension, and the little finger falls into flexion and abduction within 30 s. In the rapid grip and release test, a healthy person can make a fist and rapidly release it 20 times in 10 sec, but patients with cervical myelopathy are unable to do this rapidly (Video 43.9). Hyperreflexia is found as a long tract sign in the upper and lower extremities below the lesion. Hyporeflexia in the upper extremity is attributed to a segmental sign of cord compression. A "cervical line," the sensory change around the clavicle, is typical for cervical myelopathy. Patients have sensory disturbance below the level of the clavicle and normal sensation above this level. The modified Japanese Orthopaedic Association (mJOA) score is usually used to evaluate the severity (Table 43.1).

Table 43.1 Modified JOA scoring system[a]

Motor function	
Fingers	
0	Unable to feed oneself w/ any tableware including chopsticks, spoon, or fork, &/or unable to fasten buttons of any size
1	Can manage to feed oneself w/ spoon &/or fork but not w/ chopsticks
2	Either chopstick feeding or writing is possible but not practical, &/or large buttons can be fastened
3	Either chopstick feeding or writing is clumsy but practical, &/or cuff buttons can be fastened
4	Normal
Shoulder & elbow : evaluated by MMT score of the deltoid or biceps muscles, whichever is weaker	
−2	MMT2 or below
−1	MMT3
−0.5	MMT4
0	MMT5
Lower extremity	
0	Unable to stand & walk by any means
0.5	Able to stand but unable to walk
1	Unable to walk w/out a cane or other support on a level
1.5	Able to walk w/out support but w/ a clumsy gait
2	Walks independently on a level but needs support on stairs
2.5	Walks independently when going upstairs, but needs support when going downstairs

Table 43.1 (continued)

3	Capable of fast but clumsy walking
4	Normal
Sensory function	
(I) Upper extremity	
0	Complete loss of touch & pain sensation
0.5	≦50% normal sensation &/or severe pain or numbness
1	>60% normal sensation &/or moderate pain or numbness
1.5	Subjective numbness of slight degree w/out any objective sensory deficit
2	Normal
(II) Trunk	
0	Complete loss of touch & pain sensation
0.5	≦50% normal sensation &/or severe pain or numbness
1	>60% normal sensation &/or moderate pain or numbness
1.5	Subjective numbness of slight degree w/out any objective sensory deficit
2	Normal
(III) Lower extremity	
0	Complete loss of touch & pain sensation
0.5	≦50% normal sensation &/or severe pain or numbness
1	>60% normal sensation &/or moderate pain or numbness
1.5	Subjective numbness of slight degree w/out any objective sensory deficit
2	Normal
Bladder function	
0	Urinary retention &/or incontinence
1	Sense of retention &/or dribbling &/or thin stream &/or incomplete continence
2	Urinary retardation &/ or pollakiuria
3	Normal

The maximum score is 17 points
[a]Total score for a healthy patient = 17. Abbreviation: *MMT* manual muscle testing

43.4 Imaging

The size of the spinal canal plays an important role in the development of cervical myelopathy. The normal canal diameter from C3 to C7 is 17 to 18 mm in Caucasians and 15 to 17 mm in Japanese subjects. The canal size is smaller in patients with cervical myelopathy than in their healthy counterparts. A sagittal diameter of 12 mm (or 13 mm in some reports) or less is a critical factor in the development of CSM. Magnetic resonance imaging (MRI) is important for judging spinal cord compression (Fig. 43.1). Computed tomography is available for detecting ossification of the posterior longitudinal ligament (OPLL, Fig. 43.2) and calcification of the ligamentum flavum (Fig. 43.3). Movement of the cervical spine could affect spinal cord compression. In extension of the cervical spine, the ligamentum flavum buckles, thereby narrowing the spinal canal. Dynamic MRI clearly reveals the pathology of cervical cord compression in flexion and extension (Fig. 43.4).

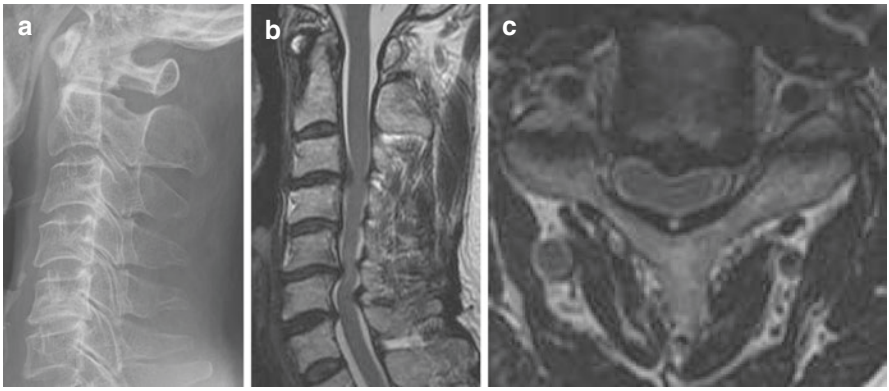

Fig. 43.1 Cervical plain radiograph (**a**) and MRI (**b** and **c**) in a typical case of CSM

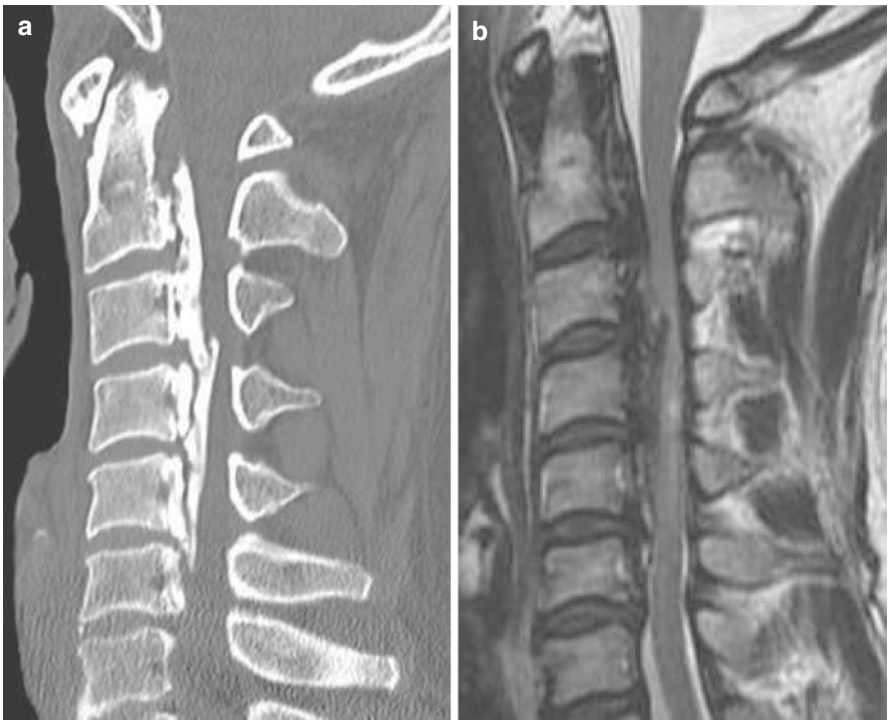

Fig. 43.2 CT (**a**) reveals OPLL. OPLL is one of the causes of spinal cord compression by MRI (**b**)

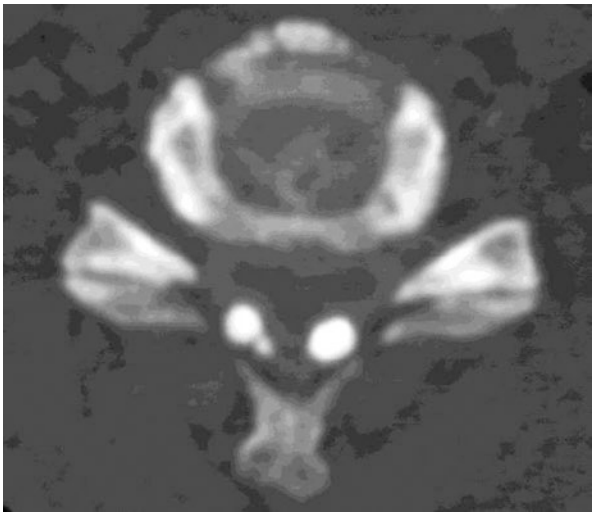

Fig. 43.3 CT reveals narrowing of the spinal canal by calcification of the ligamentum flavum

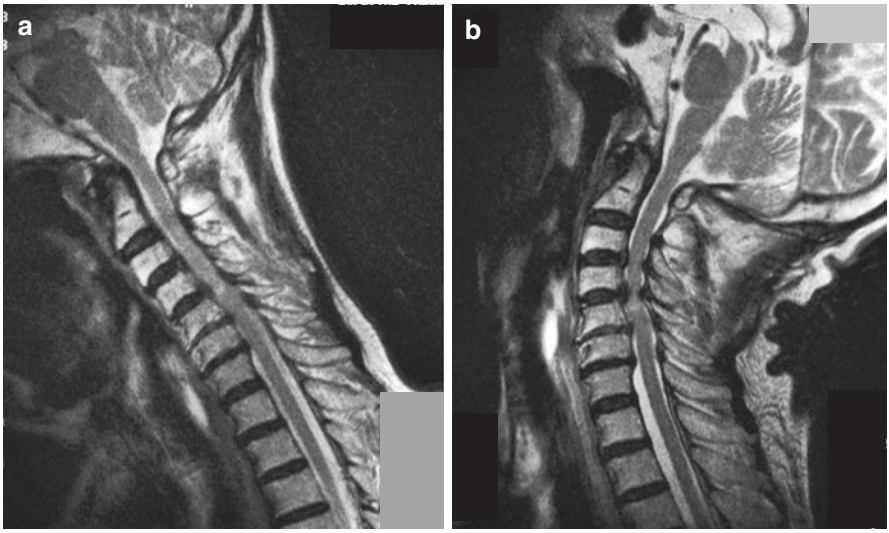

Fig. 43.4 Dynamic MRI reveals spinal cord compression in extension position. (**a**) MRI in flexion, (**b**) MRI in extension

43.5 Differential Diagnosis

Cervical cord compression is caused by spondylosis of the cervical spine and various other pathologies, such as OPLL, spinal tumors, and epidural abscess (Chap. 42). In addition, intrinsic neurogenic conditions are based on the primary pathology in the spinal cord. These intrinsic pathologies, such as motor neuron disease and multiple sclerosis, are important as differential diagnoses of compressive myelopathy.

43.6 Treatment Options

The therapeutic strategy should be decided on the basis of the symptoms, severity of cervical myelopathy, and general conditions. A narrative review revealed that surgery is not superior to conservative treatment for mild and moderate forms of CSM, whereas surgery is more suitable for patients with a clinically worse status. Guidelines for the surgical management of cervical degenerative disease published in the *Journal of Neurosurgery Spine* in 2009 stated that more severe CSM (mJOA score $\leq$ 12) should be considered for surgery (Video 43.4). There is controversy concerning whether the anterior or posterior technique is preferable. The important factors for decision-making are (1) sagittal cervical alignment, (2) the width of the spinal canal, (3) the number of affected segments, (4) the location of the compressive abnormality, and (5) the presence of instability. Patients with one or two levels of involvement should be managed using anterior cervical discectomy or corpectomy and fusion (Fig. 43.5), whereas those with the involvement of four or more levels should be treated using a posterior approach (Fig. 43.6).

43.7 Expected Outcomes

Both anterior and posterior approaches produce favorable surgical outcomes when effective decompression of the spinal cord is achieved. Various factors, such as age and the preoperative severity of spinal cord dysfunction as revealed using T2-weighted MRI, affect the surgical outcome. Patients with lordotic alignment have better clinical results after posterior decompression than those with neutral or kyphotic alignment. Poor surgical outcomes were reported after posterior decompression in patients with local kyphosis exceeding 13°.

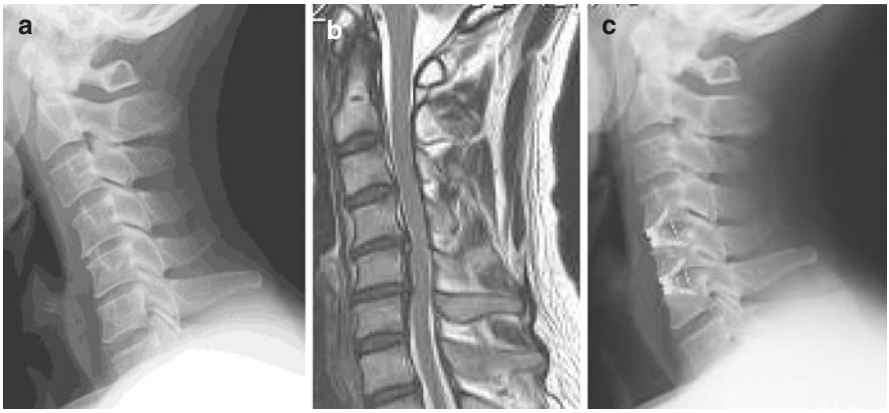

Fig. 43.5 Anterior decompression and fusion surgery at C4–C5 and C5–C6 levels

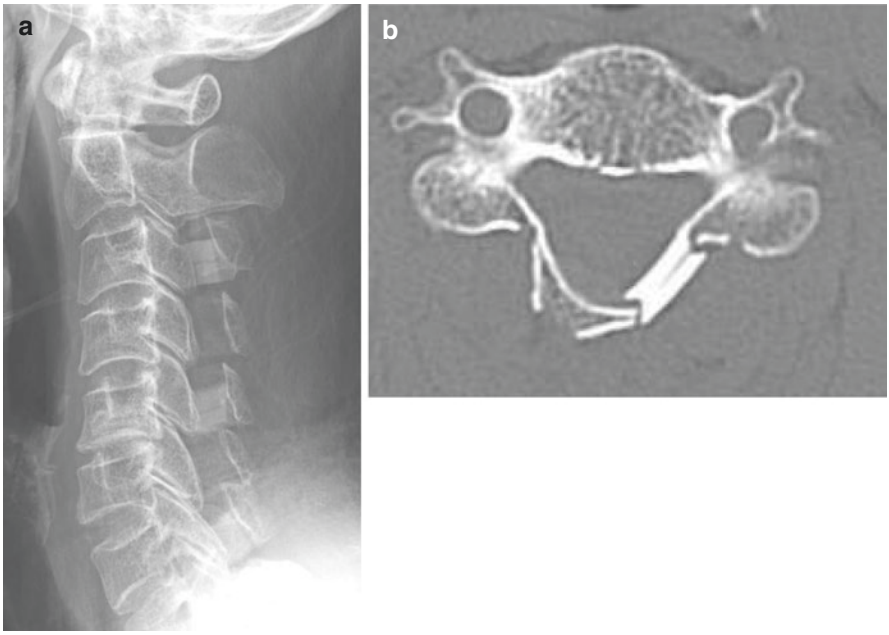

Fig. 43.6 Posterior decompression surgery using cervical laminoplasty from C3 to C7

43.8 Potential Complications

One of the disadvantages of the anterior approach is the long-term application of a rigid orthosis. Further, recurrent laryngeal nerve palsy, sympathetic nerve injury resulting in Horner syndrome, dysphagia, and vertebral artery injury have been cited as postoperative complications related to the anterior surgical approach. Dislodgement of grafted bone might occur after anterior surgery. In long-term follow-up, adjacent segment disease above or below the fusion levels should be considered. A few possible complications are related to the posterior approach. However, when posterior instrumentation is used, especially fixation using pedicle screws, care must be taken to not penetrate vertebral artery. Postoperative radiculopathy, or motor palsy of the C5 nerve root, is well known to occur occasionally after posterior decompression of the spinal cord. This complication can be found after anterior surgery. Postoperative C5 palsy is reported to occur in 5–8% of patients on average after surgery. Other common postoperative problems of the posterior approach are axial symptoms, such as axial pain and a limited range of motion of the cervical spine.

43.9 What Should Patient and Family Know?

The therapeutic strategy should be based on the severity of the symptoms. Patients with severe or progressive cervical myelopathy associated with concordant radiologic findings are candidates for operative treatment. Anterior or posterior decompressive surgery effectively improves the symptoms of cervical myelopathy. Both the anterior and posterior approaches have their own advantages and disadvantages.

Further Readings

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Thoracic Disk Herniation

44

Yoshiharu Kawaguchi

44.1 Definition

Thoracic disk herniation (TDH) causes spinal cord compression due to the prolapse of the thoracic disk and is associated with numbness and other clinical symptoms of thoracic myelopathy. The incidence of TDH is 0.25% to 1% of all spinal disk herniations. TDH is a degenerative disease, rarely related to trauma, and occurs equally in men and women, most often during middle age. The lower thoracic spine, an area where the ribs do not attach to the sternum, is the most common injury site.

44.2 Natural History

Patients with TDH can have ill-defined back pain and a range of symptoms, depending on the location of the herniation. Involvement of the intercostal nerve results in radicular girdle pain in the thoracic region. The symptoms of thoracic myelopathy progress slowly and include numbness of the lower extremities, gait disturbances (spastic gait), and vesicourethral involvement. The weakness of the lower extremities can progress rapidly after onset in some cases, and minor trauma can cause a sudden onset of a neurological deficit.

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44.3 Physical Examination

A neurological examination is checked by deep tendon reflexes, motor functions, and the existence of sensory disturbances. Patients with thoracic myelopathy have hyperreflexia of the lower extremities. The Japanese Orthopedic Association has developed a scoring system that can evaluate the severity of thoracic myelopathy (Table 44.1). Pathological reflexes, such as Babinski and Chaddock, are positive, and motor palsy and weakness are found in some cases. Sensory disturbance can also be found below the affected thoracic disk (Video 44.4). Patients with TDH in the lower thoracic region can have conus syndrome, a collection of symptoms that includes back pain, bowel and bladder dysfunctions, spastic or flaccid weakness, and bilateral sensory loss in the lower extremities.

Table 44.1 Modified Japanese Orthopedic Association scoring system for thoracic myelopathy[a]

Motor function	
Lower extremity	
0	Unable to stand & walk by any means
0.5	Able to stand but unable to walk
1	Unable to walk w/out a cane or other support on a level surface
1.5	Able to walk w/out support but w/ a clumsy gait
2	Walks independently on a level but needs support on stairs
2.5	Walks independently when going upstairs, but needs support when going downstairs
3	Capable of fast but clumsy walking
4	Normal
Sensory function	
(I) Trunk	
0	Complete loss of touch & pain sensation
0.5	≦50% normal sensation &/or severe pain or numbness
1	>60% normal sensation &/or moderate pain or numbness
1.5	Subjective numbness of slight degree w/out any objective sensory deficit
2	Normal
(II) Lower extremity	
0	Complete loss of touch & pain sensation
0.5	≦50% normal sensation &/or severe pain or numbness
1	>60% normal sensation &/or moderate pain or numbness
1.5	Subjective numbness of slight degree w/out any objective sensory deficit
2	Normal
Bladder function	
0	Urinary retention &/or incontinence
1	Sense of retention &/or dribbling &/or thin stream &/or incomplete continence
2	Urinary retardation &/or pollakiuria
3	Normal

[a]Total score for a healthy patient = 11

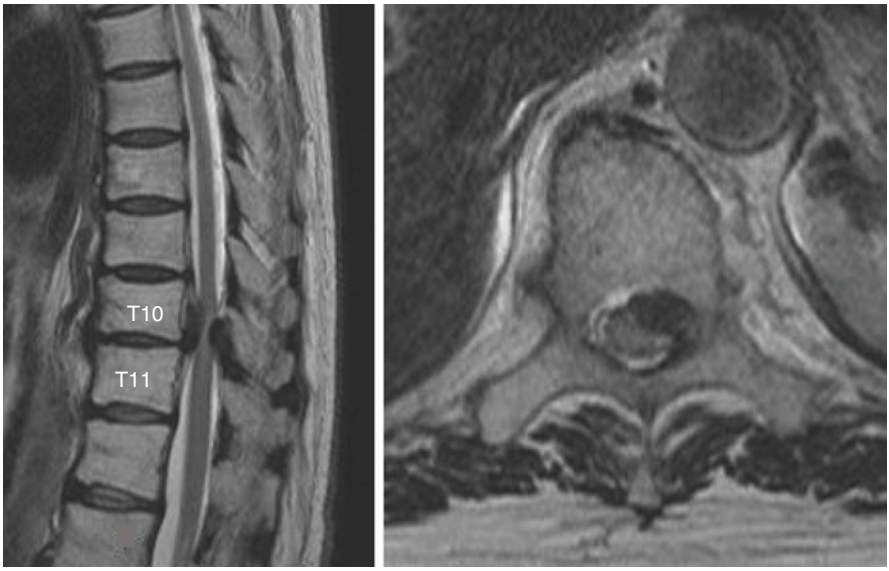

Fig. 44.1 A thoracic MRI in a typical TDH case. The patient is a 77-year-old female with T10–T11 DH suffering from incomplete paraplegia (Case 1)

44.4 Imaging

Magnetic resonance imaging (MRI) is used to check for the existence and severity of TDH and spinal cord compression (Fig. 44.1). MRI can identify the extent of disk degeneration, endplate damage, intramedullary changes of the spinal cord, and appearance of Schmorl nodes (Chap. 27). Knowing the extent of spinal cord compression allows decisions to be made about decompression surgery. The herniation type can be seen in MRI and classified as a protrusion (the most common), extrusion, or sequestration. Computed tomography (CT) is used to detect calcified lesions (Fig. 44.2), posterior bony spurs, and coexistence of ossification of either the ossification of the posterior longitudinal ligament (OPLL) (Fig. 44.3) or ligamentum flavum (OLF) (Fig. 44.4). TDH is not effectively detected with the radiographic examination. However, radiographs might identify disk space narrowing and the calcified lesions associated with TDH. Spinal alignment can also be checked with radiographs. Patients with TDH have high lumbar lordosis and low thoracic kyphosis.

44.5 Differential Diagnosis

Pathologies such as OPLL, OLF, spinal tumors, and epidural abscesses can also cause thoracic cord compression. Patients should be checked for common disorders of the spinal cord such as motor neuron disease and multiple sclerosis. Patients with TDH can also have coexisting spinal lesions in the cervical and lumbar spine.

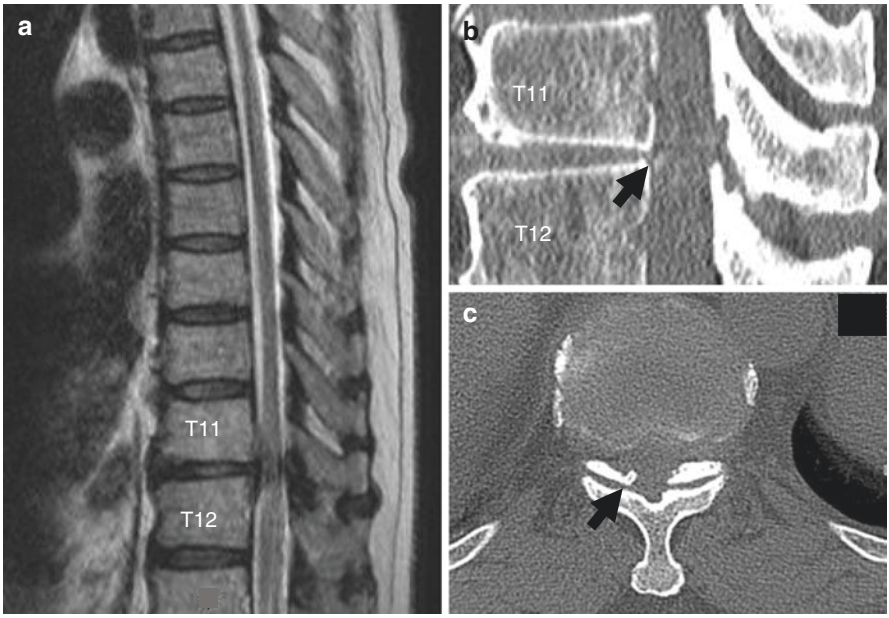

Fig. 44.2 A thoracic MRI in an 80-year-old male patient with T11–T12 DH (Case 2). (**a**) A sagittal MRI showing TDH. (**b**) TDH has a calcification shell (sagittal CT). (**c**) OLF is associated with TDH (axial CT at T11–T12)

44.6 Treatment Options

Patients with TDH whose only symptoms are back and/or girdle pain should receive conservative treatment, and pain-relief medication is the first option. Surgical treatment is indicated when a patient's persistent, severe pain cannot be relieved by medication and/or when patients have progressive neurological symptoms. Early surgery is necessary and urgent if thoracic myelopathy is evident, before irreversible spinal cord damage and loss of function occurs due to compression of the spinal cord. There are three surgical approaches: posterolateral, lateral, and anterior (Fig. 44.5), each of which decompresses the spinal cord by removing the TDH. A posterolateral approach with posterior fusion (Videos 44.3 and 44.7) is the most common procedure to remove the TDH. This approach avoids retracting the vulnerable spinal cord (Fig. 44.6). Liquorrhea should be avoided by leaving in place any occurrence of a calcified herniated disk that is firmly attached to the dura mater. A retropleural costotransversectomy, performed by a lateral approach, was popular in the 1980s but is less so now because it invades bone and soft tissue. This approach requires resection of the lateral part of the unilateral facet joint and a partial resection of the rib. More recently, minimally invasive surgery (MIS) has been introduced although it is a technically demanding procedure. An anterior approach might be considered when a large TDH is centrally located and the patient's general

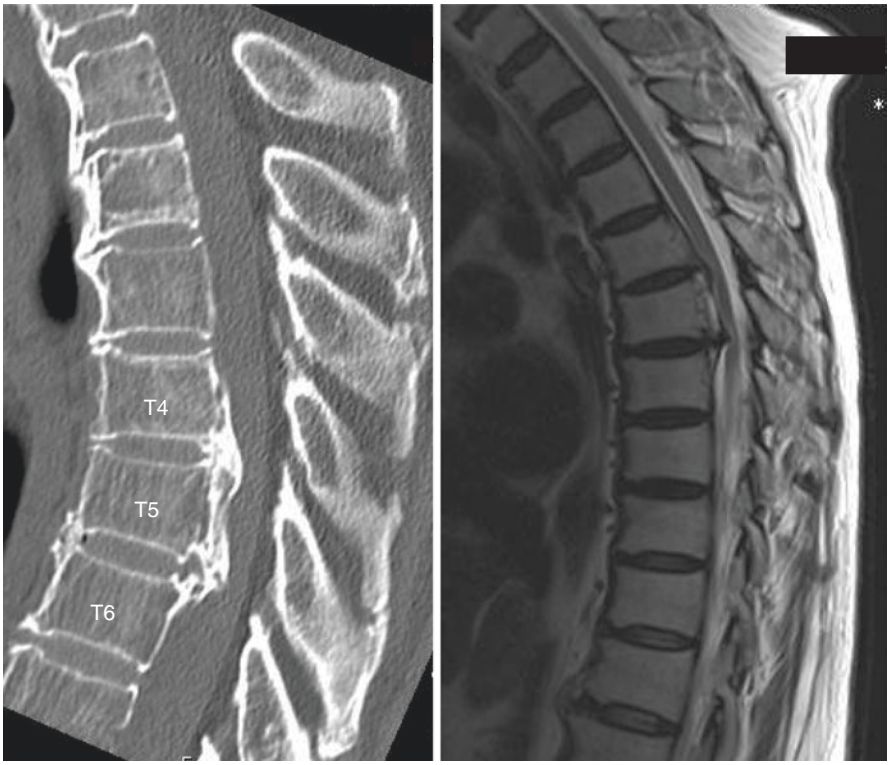

Fig. 44.3 CT of the thoracic spine showing ossification of the OPLL. MRI reveals spinal cord compression by OPLL. A 74-year-old female with T4–T6 OPLL

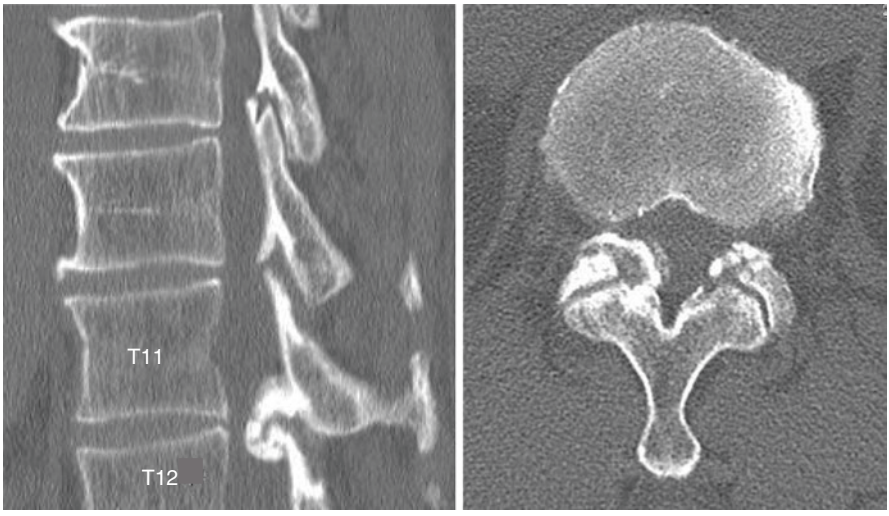

Fig. 44.4 CT of the thoracic spine showing OLF. A 55-year-old male with T11–T12 OLF

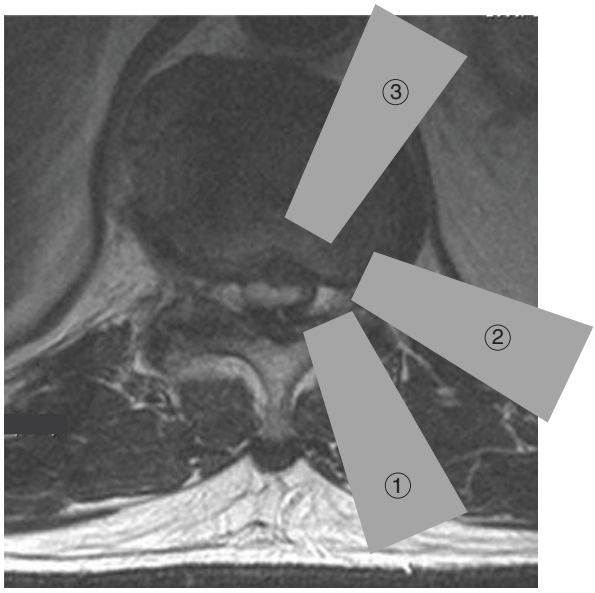

Fig. 44.5 Three options of surgical approach: ① posterolateral, ② lateral, and ③ anterior approaches (Case 3, 60-year-old male with T11–12 TDH)

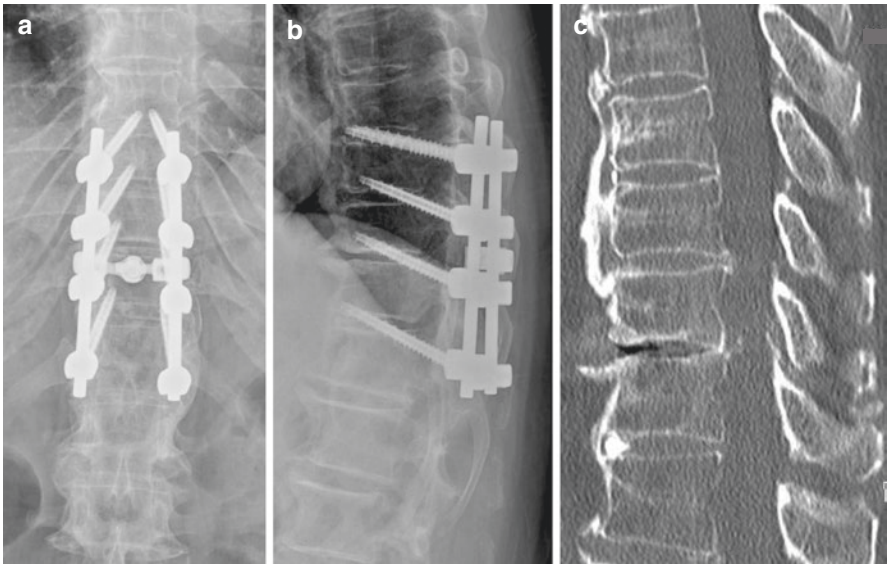

Fig. 44.6 Posterolateral approach with posterior fusion. This patient had T8–L1 posterior fusion with transpedicular screws because the patient had diffuse idiopathic skeletal hyperostosis (Case 1; Chap. 54)

condition is acceptable. With this approach, however, fusion carries a risk of injury to the major vessels, lungs, and diaphragm. Fusion surgery is usually performed in most of patients with TDH, but fusion is not required in cases where a single-level

thoracic discectomy is treated with MIS. Neuromonitoring is performed by motor-evoked potentials and/or somatosensory-evoked potentials. There is no consensus about whether intraoperative neuromonitoring is useful for preventing spinal cord damage because neuromonitoring does not always prevent spinal cord dysfunction after surgery.

44.7 Expected Outcomes

The surgical outcome is considered satisfactory when the TDH is removed and adequate decompression of the spinal cord is achieved at an early stage. Factors such as age and preoperative severity of the spinal cord dysfunction affect the surgical outcome. There can be postoperative neurological deterioration after surgery. The pathogenesis of this deterioration is unknown, but spinal shock, medullary contusion, and vascular impairment might be considered as causes.

44.8 Potential Complications

Neurological deterioration can be a major complication. Dural tears are encountered during surgery in about 40% of patients who had a massive calcified TDH and a thoracoscopic resection. Lung-related complications can occur in both anterior and posterior approaches. In addition, injury to the intercostal nerve might cause postoperative pain syndrome and neuralgia.

44.9 What Should Patient and Family Know?

The incidence of TDH is rare, compared to herniation in the cervical and lumbar disks. TDH has the potential to cause the severe neurological deficits, including paraplegia. The prognosis after any of the treatments for TDH can be favorable, but the surgical procedures have a risk of complications, including neurological deterioration.

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Lumbar Degenerative Disc Disease and Lumbar Disc Herniation

45

James E. Dowdell III and Todd J. Albert

45.1 Definition

Low back pain (LBP; Chap. 41) can have a variety of generators including degenerative disc disease (DDD) and lumbar disc herniation (LDH; Chap. 46). DDD can be defined as an age accelerated structural failure of the intervertebral disc (IVD), and thus disc degeneration does not necessarily equal DDD. To make a diagnosis of DDD, the clinical picture must be evaluated in addition to the radiographic presence of disc degeneration. LDH can occur in both degenerated and non-degenerated disc segments. LDH occurs when the nucleus pulposus is able to herniate through the annulus fibrosis. LDH can cause back pain, radicular pain, and sensory/motor disturbances.

45.2 Natural History

LBP is the most common cause of disability in young adults with an indirect loss of >100 billion dollars per year in decreased productivity. Over 80% of young adults will experience an episode of back pain in their lifetime, and an additional 2% to 3% of these patients will get radiculopathy as well. Typically, patients with DDD will

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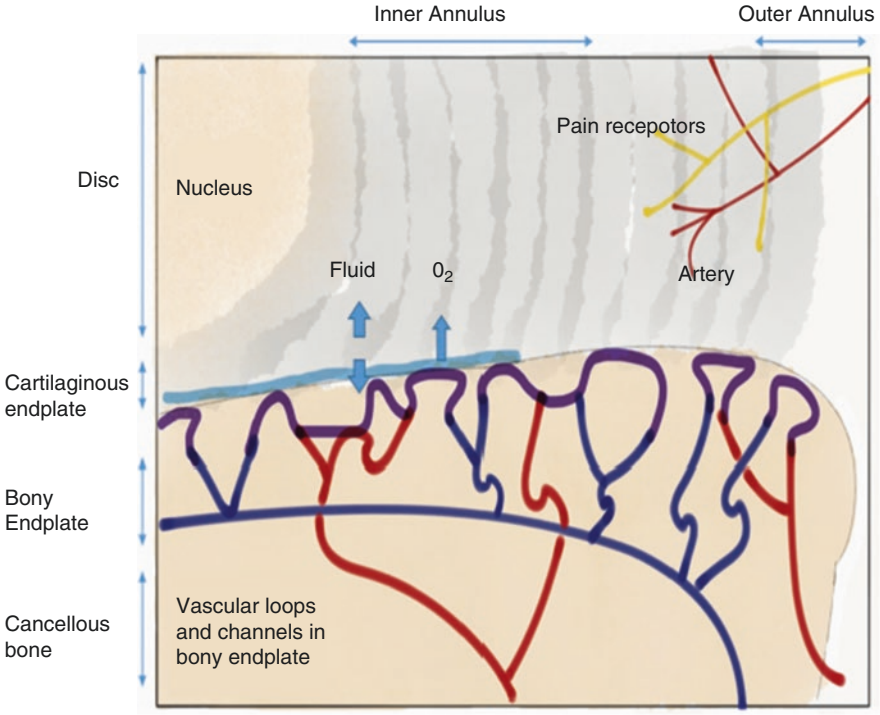

Fig. 45.1 Nutrition of the IVD

have a self-limited course of back pain, with 95% experiencing a recovery in 3 months with appropriate nonoperative treatment modalities. However, chronic back pain is a possibility for these patients. The pathway that leads to DDD is a complex interplay between nutritional, environmental, and genetic factors. Decreased nutritional supply limits the IVD from responding to load (Fig. 45.1), while genetic polymorphisms can affect genes that contribute to IVD structure and upregulate pro-inflammatory pathways (Fig. 45.2). Environmental factors that contribute to DDD include obesity and cigarette smoking. Understanding the degenerative cascade is important for the development of future treatment modalities. For LDH, the vast majority of patients (~75%) will experience relief at 1 year. However, in patients who remain symptomatic beyond 6 weeks, there is a small but statistically significant benefit to undergoing a direct decompression of the affected nerve root.

45.3 Physical Examination

Observation of behavior, gait, and muscle atrophy is important when evaluating patients with DDD and LDH. Observation of Waddell's signs is important for this patient population as well. Patients with a large LDH may be very uncomfortable and

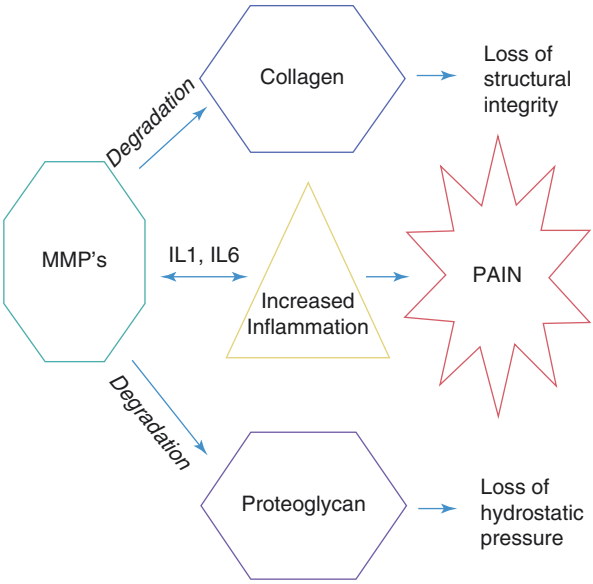

Fig. 45.2 Inflammatory pathways in DDD

be unable to sit down. Patients with DDD will present most commonly with back pain. Midline spinous process tenderness is very common. The range of motion will be limited secondary to pain (Appendix N). Pain that worsens with extension is likely facet-oriented, while worsening of pain with lumbar flexion suggests a discogenic mediator of pain. Radicular pain is very common with disc herniation. There are commonly motor, sensory, and reflex deficits along the affected nerve root as well (Video 45.4). The straight leg raise test can reproduce radicular limb pain. A positive contralateral leg raise test usually indicates a massive or extruded disc fragment.

45.4 Imaging

Radiographs of the lumbar spine should include anteroposterior (AP) and lateral radiographs of the affected area (Fig. 45.3). The AP view may show arthritic changes in the facets or osseous bridging between the disc spaces. The lateral view can show a variety of pathology including loss of disc height, spondylolisthesis, and vacuum disc (Fig. 45.4). Computed tomography (CT) scan will clearly show arthritic changes throughout the spine including in the facet joints, end plate sclerosis, and calcified disc herniation. However, a CT scan is not always required for these patients unless it is to address a specific question or if the patient is unable to obtain magnetic resonance imaging (MRI).

MRI is indicated for patients who fail to recover after 6–12 weeks of proper nonoperative management and for those with any neurological deficit (sensory/

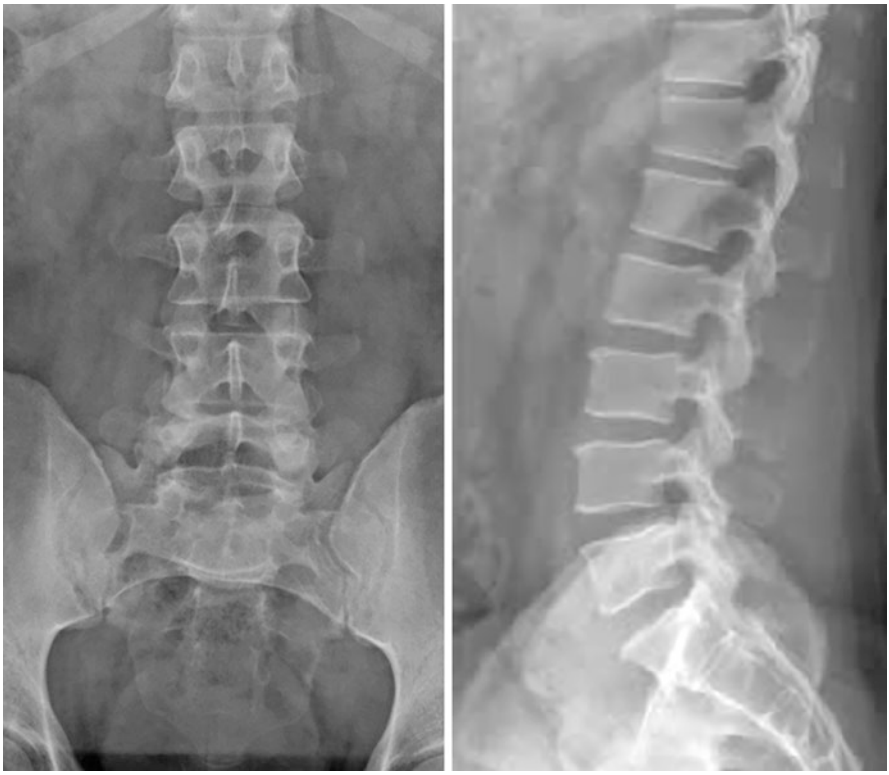

Fig. 45.3 Normal lumbar radiographs (normal disc height, no vacuum phenomenon in the disc, no arthritic changes)

motor) in a radicular pattern. Patients with DDD will typically have a loss of signal within the nucleus pulposus and disc space collapse on MRI (Fig. 45.5). Annular tears can be appreciated as high-intensity zones in the posterior annulus on T2-weighted imaging. Some MRI classifications include the Modic system and the Pfirrmann classification (Fig. 45.6; Tables 45.1 and 45.2). The clinical utility of these classification systems is uncertain, but there is evidence to suggest a higher failure of nonoperative treatment in those with Modic changes on MRI.

45.5 Differential Diagnosis

There is a broad differential diagnosis for DDD and LDH (Table 45.3). Potential pain sources could be viscerogenic (renal/abdominal causes), neoplastic (primary or metastatic bone tumors; Chap. 63), neurogenic (spinal cord tumors/cysts), inflammatory disease (ankylosing spondylitis; Chap. 54), infectious (discitis, osteomyelitis; Chap. 65), psychogenic, and spondylogenic (muscle strains, spinal stenosis, disc disease, facet arthropathy; Chap. 41; Chap. 46). Radiculopathy is most commonly

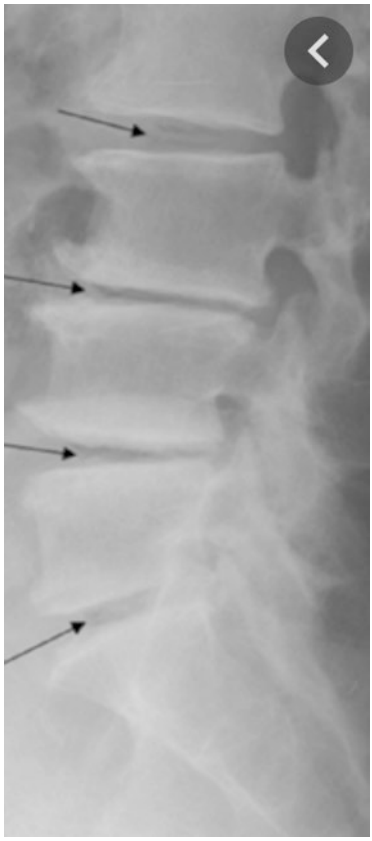

Fig. 45.4 Lumbar lateral radiograph showing vacuum disc phenomenon with accumulation of air in the disc space (black arrows)

caused by a disc herniation but may be mimicked from peripheral nerve compression (e.g., piriformis syndrome) or intraneural pathology (e.g., peripheral nerve sheath tumor).

45.6 Treatment Options

The vast majority of patients undergo nonoperative care for DDD. Physical therapy is a mainstay of treatment, along with lifestyle modification (smoking cessations/weight loss/core strengthening/back education). Anti-inflammatory medications are very effective for treating pain related to DDD and LDH. Steroid medications are useful for radiculopathy. Cognitive behavioral therapy is effective at treating back pain in patients with chronic pain. Alternative therapies are frequently attempted as well, including yoga, CBT, chiropractor, CBD oil, and oral CBD with varying success. In patients with radiculopathy, transforaminal epidural injections can be effective in reducing the chemical/inflammatory component of the radiculopathy but do not change the natural history of the disc herniation.

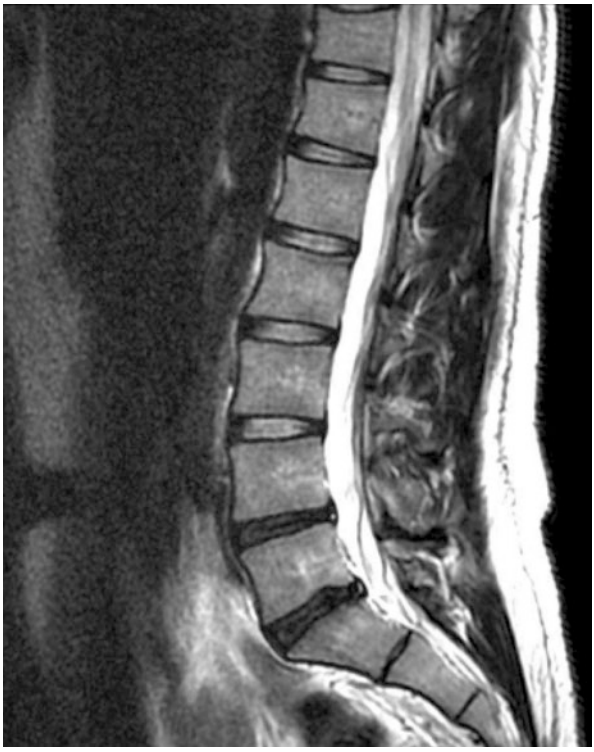

Fig. 45.5 T2 lumbar MRI showing degenerative disc at L4-5, L5-S1 (loss of hydration, disc collapse), no Modic changes

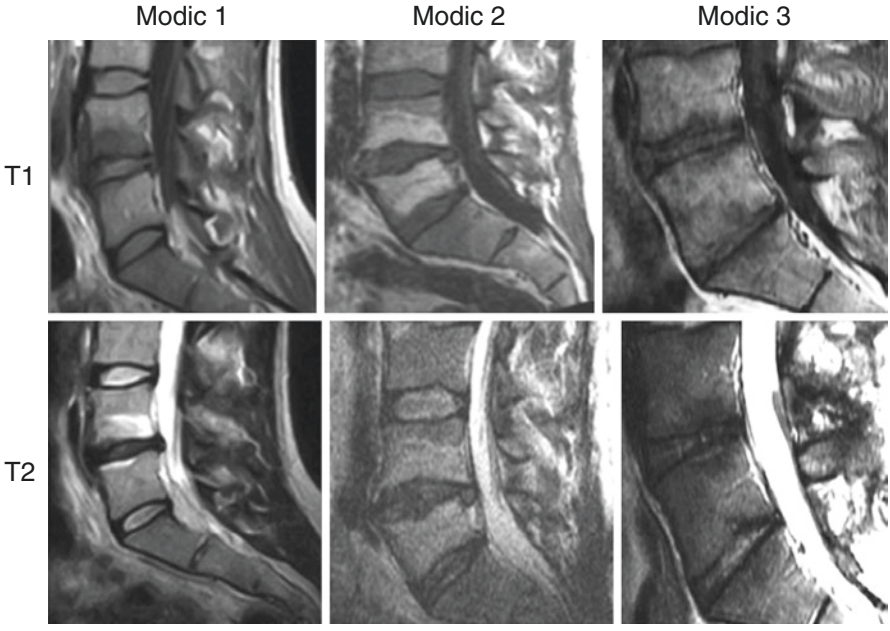

Fig. 45.6 Modic changes (Reproduced from Jones et al.)

Table 45.1 Modic changes as illustrated

Type	T1 MRI signal intensity	T2 MRI signal intensity
I	Hypointense	Hyperintense
II	Hyperintense	Iso or hyperintense
III	Hypointense	Hypointense

Reproduced

Table 45.2 Pfirrmann grades as illustrated

Grade	Structure	Distinction (nucleus and annuals)	T2 MRI signal intensity	Disc space height
I	White, homogenous	Clear	Isointense to cerebrospinal fluid (hyperintense)	Normal
II	Inhomogeneous, with banding	Clear	Isointense to cerebrospinal fluid (hyperintense)	Normal
III	Gray, inhomogeneous	Unclear	Intermediate	Normal to decreased
IV	Gray to black inhomogenous	No distinction	Intermediate to hypointense	Normal to decreased
V	Black inhomogenous	No distinction	Hypointense	Collapsed

Reproduced

Table 45.3 Differential diagnosis of low back pain

Type	Differential
Viscerogenic	Abdominal and renal
Neoplastic	Primary and metastatic bone tumors
Neurogenic	Spinal cord tumors or cysts
Inflammatory diseases	Ankylosing spondylitis, Reiter's syndrome, inflammatory bowel diseases, and psoriatic arthritis (sacroiliitis)
Infectious	Diskitis, osteomyelitis, psoas abscess
Spondylogenic	Myofascial syndromes Iliolumbar syndrome, piriformis syndrome, quadratus Lumborum syndrome, and fibrositis (trigger point syndrome) Motion segment disorders Disk disease, facet syndrome, spinal stenosis Bony problems Fractures, including osteoporotic compression fractures Spondylolisthesis Sacral lesions, coccyx pain Psychogenic

For LDH, in patients who fail conservative treatment for at least 6 weeks, surgical intervention can lead to improved outcomes. For any patient with a progressive neurological deficit or neurological findings (radicular pain/straight leg raise positive), operative management is appropriate (Video 45.4). Any method of lumbar decompression and stabilization is appropriate (open or minimally invasive) for these patients if a direct nerve decompression can be achieved (Video 45.3).

45.7 Expected Outcomes

The majority of patients recover very well from disc herniation. Patients with DDD can have a more variable clinical course due to the complexity of treating LBP. Nonoperative modalities of treatment can provide significant relief for these patients, but for those with chronic unrelenting pain, surgical treatment without any accompanying neurological symptom is very controversial with no guarantee it will relieve pain.

45.8 Potential Complications

The major complication risk profile for lumbar decompression surgery is continued symptoms, nerve root injury, dural tear, re-herniation, and infection. However, the complication of nonoperative care for an LDH in a patient with neurological compromise is chronic pain and permanent loss of muscle function.

45.9 What Should Patient and Family Know?

DDD is a lifelong condition that will require significant efforts in lifestyle modifications and physical therapy for the best chance of a good outcome. Patients with LDH with symptoms lasting longer than 6 weeks or with neurological compromise have better outcomes with surgery. These patients have a great chance for a full functional recovery.

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Lumbar Spinal Stenosis

46

Jason Pui Yin Cheung and Kenneth M. C. Cheung

46.1 Definition

Lumbar spinal stenosis should be differentiated into clinical and/or radiological stenosis. Clinical stenosis refers to the presence of radiculopathy, neurogenic claudication, or lower limb neurological deficit. Radiological stenosis refers to narrowing of the spinal canal with the compromise of the dural sac and the nerve roots, due either to developmental narrowing or progressive degeneration or a combination of both. Developmental stenosis (Fig. 46.1) is due to maldevelopment of the neural arch, while degenerative stenosis is a result of progressive spinal segment degeneration, starting with the loss of disc height, disc bulging, facet joint, and ligamentum flavum hypertrophy. These features contribute to canal stenosis, and the location of pathology can be classified and is illustrated in Table 46.1 (Table 46.1).

46.2 Natural History

The natural history of spinal stenosis is quite variable. Radiological stenosis is not necessarily accompanied by symptoms. A general trend observed is that over a period of 2 to 5 years after the initial presentation of symptoms secondary to spinal stenosis, approximately 20% of patients worsen with nonoperative treatment, 40%

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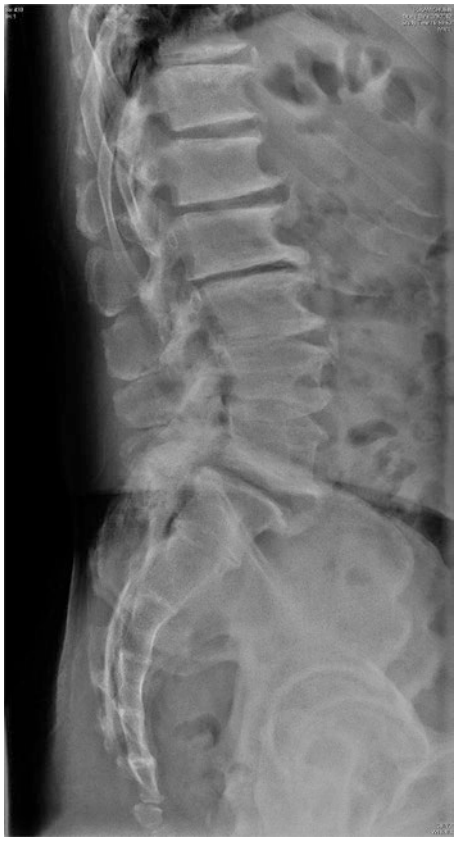

Fig. 46.1 Developmental spinal stenosis as evidenced by multilevel short pedicles

Table 46.1 Classification of spinal stenosis by location

Classification	Location	Compression	Pathology
Central stenosis	Narrowed central zone of the spinal canal	Dural sac	Anterior: central disc herniation Posterior: inferior articular facet hypertrophy and ligamentum flavum hypertrophy
Lateral recess stenosis	Narrowing of the subarticular recess causing nerve root compression	Traversing nerve root	Anterior: posterolateral disc herniation Posterior: Superior articular facet and ligamentum flavum hypertrophy
Foraminal stenosis	Neuroforamen anteriorly bounded by the disc and end plate, posteriorly by the pars interarticularis, and superiorly and inferiorly by the pedicles	Exiting nerve root	Far lateral disc herniation, overriding superior articular process

Table 46.2 Pain generators in lumbar spinal stenosis

Leg pain/ radiculopathy	Direct mechanical compression of nerve root by osteophyte or disc herniation Vascular embarrassment of blood supply
Neurogenic claudication	Cauda equina irritation from exercise-induced ischemia Small intraneural arterial occlusion Venous congestion

stay the same, and 40% improve [1]. Typically, over a period of 2–3 years, patients with moderate stenosis may be treated without surgery, because acute deterioration is uncommon. Regardless of the decreased walking tolerance and gait disturbances, most patients still lead active lives.

46.3 Physical Examination

Patients may present with a combination of radiating leg pain and/or back pain, weakness, and sensory change, with loss of ankle or knee jerk reflexes due to compression of segmental nerve roots (Table 46.2) (Video 46.4) [2]. Lumbar extension on physical examination can elicit back or leg pain, but the extension position should be held for at least 30 s in order to provoke such symptoms. Objective motor and sensory loss should pinpoint the specific nerve root involved.

46.4 Imaging

Plain standing radiographs can demonstrate spondylolisthesis, disc space narrowing, end plate sclerosis, osteophytes, and facet hypertrophy. Lateral dynamic flexion and extension radiographs can determine whether spondylolisthesis (Chap. 47) is demonstrating instability**.** Whole spine standing radiographs are also used to assess the sagittal alignment. Lateral recess and central canal stenosis can be easily assessed on axial T2-weighted magnetic resonance imaging as a trefoil-shaped spinal canal (Fig. 46.2). Foraminal nerve root compression is identified as a complete obliteration of the fat in the foramen on T1-weighted images. Nerve root sedimentation (Fig. 46.3) to the dorsal section of the dural sac as a result of gravity is expected for a patient in supine. The lack of sedimentation is a positive sign of lumbar spinal stenosis.

46.5 Differential Diagnosis

Lower limb ischemia is an important differential diagnosis. The vascular examination should be performed including observation for trophic changes in the skin and nails of the lower limbs and diminished distal pulses. Other conditions such as cervical myelopathy (Chap. 43) and hip osteoarthritis should be ruled out. Examination of the upper limb neurology, gait instability, and brisk reflexes should distinguish

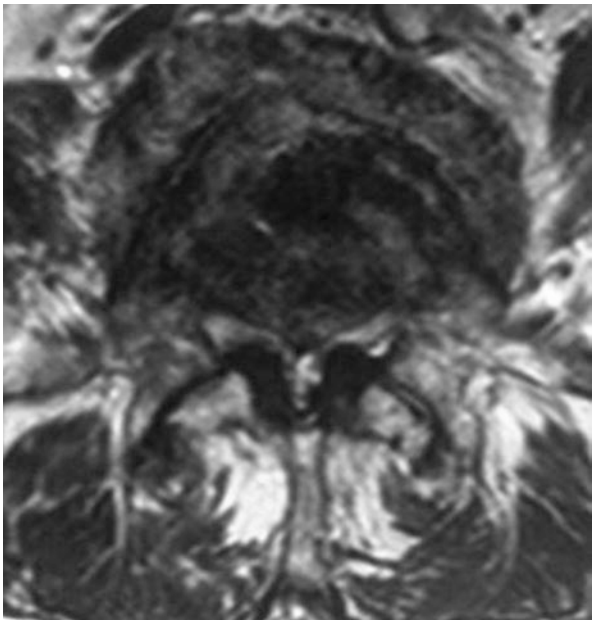

Fig. 46.2 Obliterated cerebral spinal fluid signal with a trefoil-shaped canal. Compression is by disc herniation anteriorly and hypertrophic ligamentum flavum posteriorly

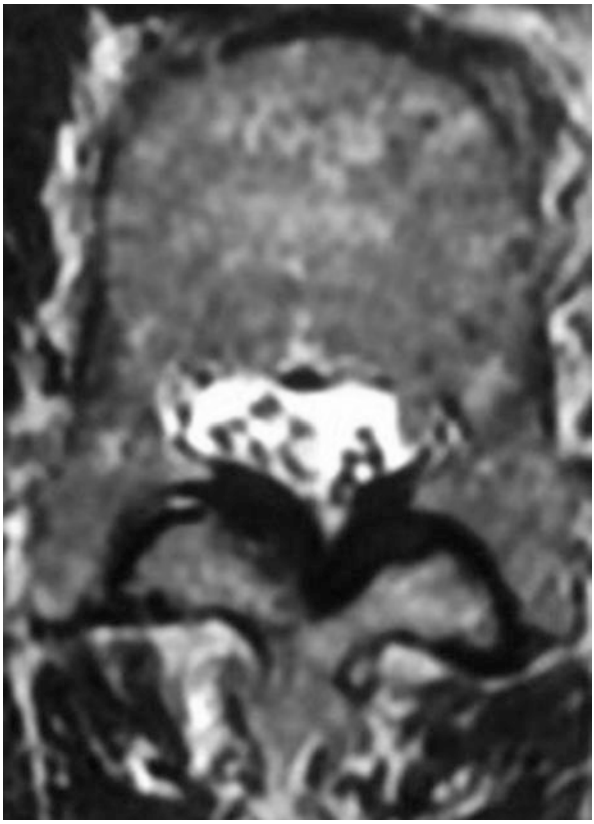

Fig. 46.3 Nerve root sedimentation sign

any cervical cord lesions. Patients should be observed while walking to detect any unusual limping, and the hip should be examined.

46.6 Treatment Options

Management of spinal stenosis involves relief of back and leg pain, prevention of deterioration and recurrence of symptoms, and improvement of function such as quality of life and walking distance. Conservative management is commonly offered first, and surgery is indicated if conservative measures fail to improve symptoms or if they persist and in cases of cauda equina syndrome where a delay of treatment may cause irrecoverable neurological, bladder, and bowel deficits.

Nonoperative management includes avoidance of spine extension posture, core muscle strengthening, and physiotherapy. Some well-known physiotherapy treatment regimens include elastic lumbar binder, physical therapy, cryotherapy, heat therapy, acupuncture, ultrasound, transcutaneous electrical nerve stimulation, and lumbar traction, although most only provide temporary relief. Medications include nonsteroidal anti-inflammatory drugs (NSAIDs), gabapentin, vitamin B_{12}, and calcitonin, but evidence for their utility is weak. Epidural and selective nerve root blocks can be diagnostic and therapeutic.

Surgery is usually helpful for leg symptoms, but back pain associated with spondylosis or root compression may not always improve. The actual surgical procedure is dependent on the pathoanatomy. Decompression is by laminotomy or laminectomy. Removal of the ligamentum flavum, medial facetectomy, and removal of osteophytic ridge adjacent to the disc space is usually sufficient to decompress the central canal and traversing nerve roots (Fig. 46.4). Usually, the medial one-third to half of the facet is removed. Any further lateral can result in instability of the spinal segment. Discectomy is also performed for extrusion or protrusion that compresses the neural elements. Foraminal decompression is achieved by undercutting hypertrophic facet joints. The nerve root and thecal sac should be visualized to avoid nerve root or dural injury. Patency of the entire nerve root should be confirmed through the neuroforamen.

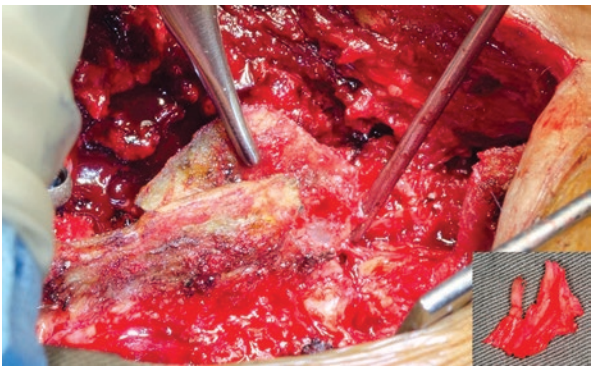

Fig. 46.4 Ligamentum flavum excision with its characteristic yellow appearance in the lower right corner

46.7 Expected Outcomes

Nonoperative management is helpful for symptom control, especially pain. Surgery is often required in patients with neurogenic claudication and neurological deficit. Surgery usually leads to good improvement of pain, function, and quality of life (Video 46.1).

46.8 Potential Complications

Inadequate decompression and wound infections are the most common complications. Dural tears may occur and should be managed intraoperatively. Injury to the nerve root is rare. Reoperation rates at the adjacent segment are common (21.7%) in patients with developmental spinal stenosis [3].

46.9 What Should Patient and Family Know?

Lumbar spinal stenosis is a common disease affecting an aging population. Many patients lead functional lives despite the disease. Surgery is recommended if there are neurological deficits or if conservative treatment fails. Surgical treatment is not complex, and patients often experience good relief of symptoms.

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Degenerative Instability

47

Yat-Wa Wong

47.1 Definition

Degeneration of the intervertebral disc reduces its capability to resist loading in all directions. Extra stress is then imposed on the facet joints, resulting in degeneration. With the progression of disc and facet joint degeneration, segmental instability develops, and the proximal vertebra slips anteriorly on the adjacent distal vertebra, known as degenerative spondylolisthesis. It commonly occurs at L4/L5, but it may also occur at other segments.

47.2 Natural History

Radiological instability is not necessarily symptomatic. With the same radiological findings, patients may be asymptomatic or present with back pain, radicular pain, spinal claudication, or any of these combinations. Back pain can also be a continuous dull ache or mechanical in character. The latter is more suggestive of instability but not pathognomonic. Long-term follow-up by Matsunaga (*J Neurosurgery* 2000) showed that only 30% had progression of slip and 24% of patients without neurological deficit had deterioration of symptoms. Osteophytes, a complete collapse of

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the intervertebral disc leading to proximal vertebral body resting on the distal vertebral body, facets hypertrophy, and ligamentum flavum hypertrophy may stabilize the motion segment (Fig. 47.1a–d). For severely symptomatic patients, Weinstein (*N Engl Med* 2007) reported better outcomes by surgery. In summary, natural history varies among patients. The treatment given should be tailor-made.

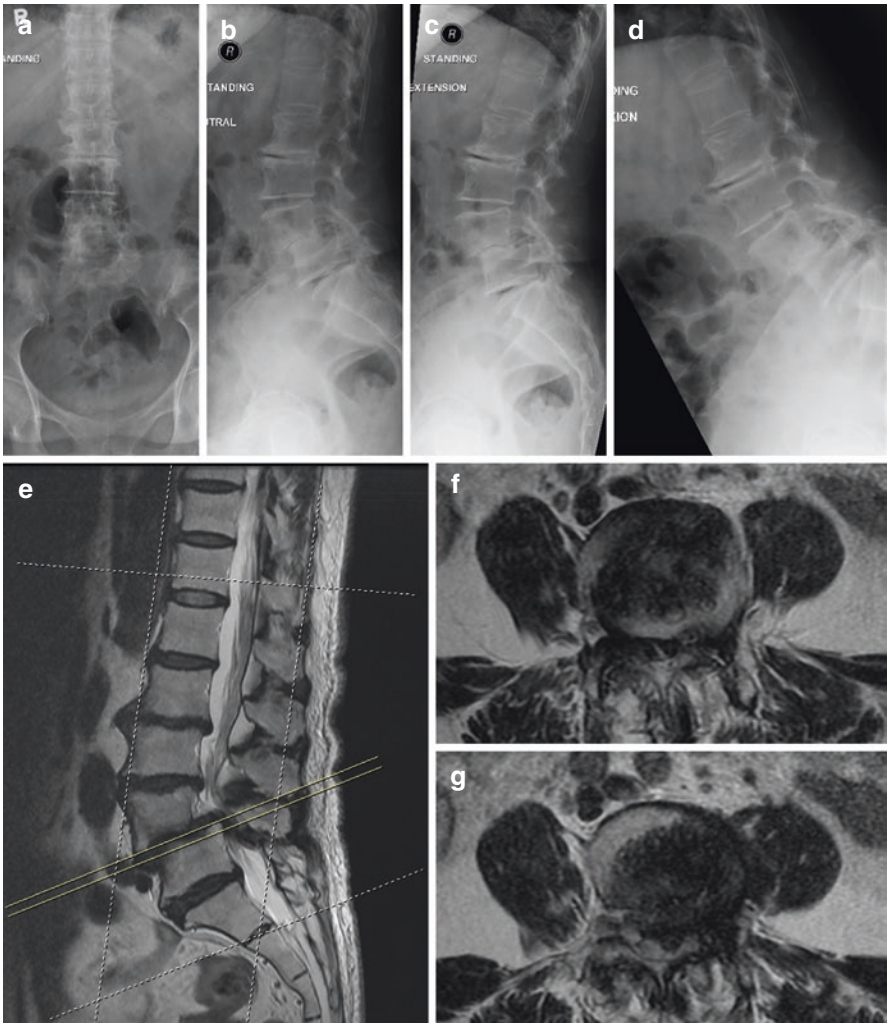

Fig. 47.1 (**a** and **b**) Standing anteroposterior and lateral radiographs showing degenerative changes such as reduced intervertebral disc height and osteophytes. (**c** and **d**) Standing extension and flexion radiographs demonstrating no significant instability although there was L4/5 grade II spondylolisthesis. (**e**–**g**) T2-weighted sagittal and axial MRI showing the L4/5 spinal stenosis. The patient presented with spinal claudication without symptoms at rest. L4/5 decompression without fusion gave good symptomatic relief with improved walking tolerance (**e**–**g**)

47.3 Physical Examination

If patients complain of lower limb pain (or numbness/paresthesia), clinicians should differentiate whether they are referred pain, radicular pain, or spinal claudication (most evocative sign). Referred pain does not go beyond the knees. Radicular pain is distributed along the dermatomes. The pain may be persistent or immediately after ambulation. For spinal claudication, the pain or paresthesia is also distributed along the dermatomes, but patients are relatively asymptomatic within claudication distance. Patients tend to lean forward, and their lumbar extension movement may be reduced due to customary lumbar flexion to relieve claudication. To distinguish spinal from vascular claudication, patients have palpable peripheral pulses, no skin ischemic changes at the distal lower limbs, and symptoms (pain or sensory disturbance) in a dermatomal distribution.

Neurological examination is mandatory although the neurological deficit is a late presentation (Video 47.4). Genuine positive nerve tension sign is uncommon in spinal stenosis due to degenerative spondylolisthesis. A positive test suggests the sciatica is due to prolapsed intervertebral disc (Chaps. 44 and 45), which is less common in the elderly.

47.4 Imaging

Standing anteroposterior and lateral radiographs of the lumbar spine may reveal degenerative changes, including decreased intervertebral disc height and spondylolisthesis (Fig. 47.1a, b). The degree of slip is usually Meyerding classification grade I (less than 25% slip). Grade II (25–50% slip) is uncommon, and grade III (50% to 75% slip) or above seldom occurs in degenerative spondylolisthesis (Appendix K). Other important findings include congenitally short pedicles which reflect narrow spinal canal, pars defect which may indicate additional instability apart from degeneration, and sagittal malalignment; moreover, it is important to rule out rotatory dislocation between two successive spinal units. Rotatory dislocation can be "open" when facet joints widen on one side or "closed" when facet joints narrow on the opposite side. For patients with a loss of normal lumbar lordosis, conducting a whole spine radiographs is important to evaluate the global spinal alignment (Chap. 50).

Dynamic radiographic examination is used to assess the degree of instability (Fig. 47.1c, d)**.** However, there are different ways to take the dynamic views. Lateral flexion/extension radiographs can be taken in recumbent or standing position. Luk (*Spine* 2003) demonstrates that the maximal slip occurs at standing flexion radiograph, whereas the maximal reduction occurs at prone traction radiograph.

Magnetic resonance imaging is useful for confirming the diagnosis and how the cauda equina or nerve roots are compressed (Fig. 47.1e–g). It can evaluate disc pathology, hypertrophy of the ligamentum flavum and facets, synovial cyst, and facet joint inflammation. However, correlation with the clinical picture is important for management since not all patients with positive radiological findings are symptomatic.

47.5 Differential Diagnosis

Typical spinal claudication due to degenerative spondylolisthesis is not difficult to diagnose. It can be easily differentiated from vascular claudication. Radicular pain occurs at rest or immediately after walking without pain-free claudication distance. Occasionally, it can be caused by acute disc prolapse or inflammation of the nerve root itself. Their natural history and treatment strategies are different from degenerative instability. It is difficult to locate the pain source of patients with predominantly back pain and multilevel degeneration (Chap. 41). Understanding the characteristics of patients and conducting a careful analysis of the clinical features are the keys to avoiding poor surgical results.

47.6 Treatment Options

Most patients have the self-limited disease and do not require active intervention. The majority of symptomatic patients respond to nonoperative treatments, such as activity modification, simple analgesics, anti-neuropathic pain drugs (such as gabapentin and pregabalin), and physical therapy. Having an appropriate amount of strengthening exercise, stretching, and mobilization may increase the functional capability and pain tolerance. Injections such as epidural steroids or facet joints may serve diagnostic and therapeutic purposes, although controversy exists in the published literature.

Surgery is indicated if there is a significant neurological deficit. Failed nonoperative treatment and severe symptoms are relative indications. Treatment options include direct decompression, direct decompression plus fusion, and indirect decompression with fusion. Degenerative instability is a spectrum of disease with a wide clinical presentation and varying degrees of spinal segmental instability. The choice is based on patients' symptoms, radiological findings, and surgical expertise. The limitation of published literature is due to the lack of outcome comparison for cohorts with similar symptoms, radiological findings, and surgical techniques (Videos 47.3, 47.4 and 47.7).

Direct decompression by fenestration and preserving more than 50% of facets does not destabilize the motion segment significantly. Less invasive techniques such as unilateral approach with bilateral decompression or endoscopic decompression further reduce destabilization. Decompression alone is suitable for patients presenting with predominant spinal claudication or radicular pain without significant mechanical back pain. It is also suitable for spondylolisthesis slip less than 5 mm from lateral standing radiograph or segments having been self-stabilized (Fig. 47.1c, d).

Decompression with fusion is appropriate for patients with severe mechanical back pain and gross radiological instability. Since instrumentation improves fusion rate and sagittal alignment, modern fusion is commonly done with instrumentation. Common fusion techniques include posterolateral fusion and posterior or transforaminal interbody fusion +/− posterolateral fusion (Fig. 47.2a–f). Indirect

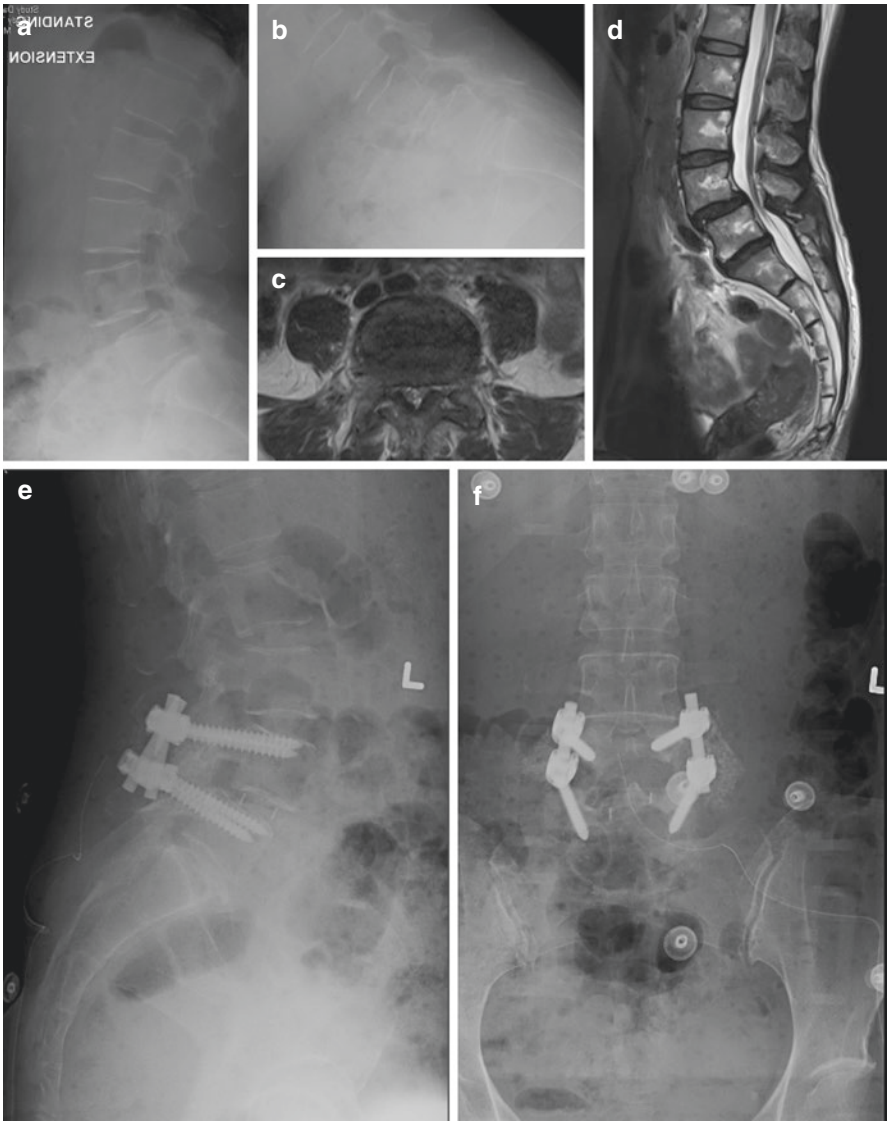

Fig. 47.2 This patient presented with severe back pain and bilateral L5 radicular pain. (**a** and **b**) Extension and flexion radiographs demonstrating unstable L4/5. (**c** and **d**) Axial and sagittal T2-weighted MRI showing L4/5 spinal stenosis. (**e** and **f**) Postoperative radiographs after direct decompression, transforaminal interbody fusion, and posterolateral fusion

decompression by restoring the intervertebral disc height (e.g., by anterior, oblique, or lateral interbody fusion) is another alternative, but additional fixation is necessary to avoid nonunion (Fig. 47.3a–h). The prerequisite of indirect decompression and fusion is minimal facet hypertrophy.

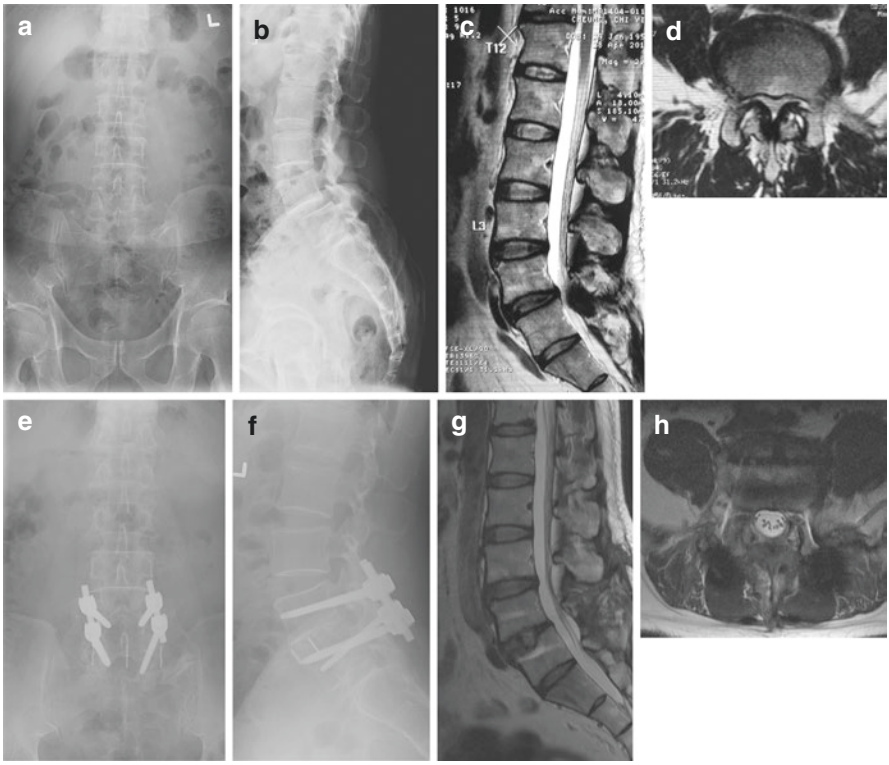

Fig. 47.3 (**a** and **b**) Standing anteroposterior and lateral X-ray showing grade 1 L4/5 spondylolisthesis before surgery. (**c** and **d**) T2-weighted MRI demonstrating L4/5 spinal stenosis mainly due to ligamentum flavum hypertrophy. (**e** and **f**) Anteroposterior and lateral radiographs after surgery showing the restoration of disc height. The construct was reinforced by posterior instrumentation. (**g** and **h**) Postoperative MRI illustrating the bigger spinal canal at L4/5. The spinal canal was enlarged by ligamentotaxis, and it is regarded as indirect decompression since the spinal canal was not entered during surgery

47.7 Expected Outcome

The surgical outcome is satisfactory for most patients with appropriate indications.

47.8 Potential Complications

Neurological complications are not common. Progression of instability may occur if excessive decompression without fusion is done, with the consequence to perform a more extended fusion area later on. For fusion, nonunion and implant malposition may happen.

47.9 What Patient and Family Should Know?

The treatment selected depends on the patients' symptoms. The natural history is favorable for most patients with or without operation.

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Adult Idiopathic Scoliosis

48

Ali Eren and Alpaslan Şenköylü

48.1 Definition

A lateral spinal plane deformity with a curve greater than 10° in a skeletally mature individual is defined as adult idiopathic scoliosis (AdIS). It is a combination of neglected adolescent scoliosis and progressive spine degeneration related to aging (Video 48.10). An asymmetrical degeneration of the discs or facet joints and osteoporosis mainly lead to the deformity. However, iatrogenic, post-traumatic, congenital, post-paralytic, or secondary to infectious or neoplastic issues can also be the other causes of AdIS which are relatively less frequent. Despite prevalence ranging from 1.4% to 32%, the rate is on the rise due to increased life span.

48.2 Natural History

While adolescent idiopathic scoliosis progresses by 0.5° to 1° per year on average (Chap. 17), AdIS is likely to progress more rapidly by 1° to 6° per year due to its degenerative and imbalanced nature. Thoracic curves progress more rapidly compared to lumbar and thoracolumbar curves that are more disabling for patients with pain, imbalance, and radicular impingement.

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48.3 Physical Examination

Unlike adolescent idiopathic scoliosis, back pain (Chap. 41) is a common symptom in AdIS, and the main reason for the pain is mostly spinal stenosis which is located particularly on the concave side of the curve. The stenosis typically does not relive with sitting or forward flexion. Radicular pain and neurologic claudication are also common symptoms caused by the impingement of nerve roots (Video 48.4).

Following meticulous evaluation of spinal geometry including kyphosis, curve magnitude (Video 48.6), flexibility, and pelvic obliquity, a complete neurological exam should be carried out.

48.4 Imaging

Anterior-posterior and lateral radiographs should be the first step after a proper physical examination. In these radiographs, clavicle and pelvic vertebra, involving both femoral heads, should be included for a proper evaluation of the spine as a whole. Flexibility can also be evaluated with lateral bending radiographs and/or longitudinal traction radiographs (standing or supine). The Cobb angle (Video 48.6) should be measured together with central sacral vertical line (CSVL) and C7 plumb line.

A computed tomography scan can be requested to evaluate bone and rotational anatomy in more detail and to detect rotatory dislocation (Chap. 47); it is particularly helpful for preoperative planning and operation.

Based on neurological examination, magnetic resonance imaging can be done for either a specific level or entire spine evaluation in order to identify any related pathologies such as disc degeneration, hernia, and intraforaminal and central stenosis.

The degree of osteoporosis, one of the contributors to AdIS, can be evaluated by dual-energy X-ray absorptiometry (DEXA); it should be suspected if there are associated compression fractures.

48.5 Differential Diagnosis

AdIS is a combination of idiopathic deformity and degeneration of skeletally mature patients and should be differentiated from adolescent idiopathic scoliosis in which curve starts developing in an immature skeleton (Chap. 16). This differentiation is crucial since the treatment of both conditions varies considerably.

In addition to this, the patient's main concern and its source should be differentiated from other reasons. In other words, back pain, for example, may not stem from a spinal deformity but from something else like a spinal tumor, infection (Chap. 65), Schmorl's node (Chap. 27), or myalgia and this should be addressed meticulously. Otherwise, the further intervention might not relieve the symptoms and result in unsatisfying outcomes.

48.6 Treatment Options

The majority of patients can be managed by conservative means such as anti-inflammatory medication, pain medication, exercises and activity modification, epidural steroid injections, and bracing. NSAIDs and non-opioid pain killers can be used for pain relief. The ideal initial treatment should be full-body exercises with strengthening but not stretching exercises. Exercises can delay curve progression while decreasing the symptoms. Epidural steroid injections could be reasonably beneficial for those who have spinal stenosis or radiculopathy signs together with contribution to the diagnosis. Even though bracing can alleviate symptoms, it does not affect curve progression. In some patients, radiofrequency denervation of the facet joints could be an alternative treatment despite not being enough support regarding long-term results.

The main reasons for surgery are persistent pain despite conservative treatment, rapid increase in curve magnitude, symptomatic coronal and sagittal imbalance, and cardiopulmonary limitations.

The surgical treatment can be divided into three main categories, considering the nature of the deformity, the symptoms, and the surgical planning.

1. Patients with isolated spinal stenosis with an acceptable and stable spinal balance can be managed with decompression without fusion. Patients should be selected carefully since decompression alone can cause further instability in the unstable spine which may lead to localized fusion.
2. Patients with stenosis, instability and moderate malalignment, no kyphosis, and plumb line no more than 5 cm from S1 can be treated with decompression, instrumentation, and fusion with minimally invasive surgery (Videos 48.3 and 48.7). Slight correction can be sufficient for this type of deformity.
3. Patients with severe curvature and instability should be managed with decompression, correction of the sagittal-coronal alignment, and long fusion, regardless of the presence of spinal stenosis (Videos 48.3 and 48.7). Osteotomies (pedicle subtraction and Smith-Petersen technique) can also be performed when necessary. Due to higher complication rates of osteotomies, the use of intraoperative neuromonitoring and high-level experience are strongly recommended (Fig. 48.1).

Correction of the deformities is performed through anterior and posterior approaches. Despite solely posterior approach along with osteotomies being sufficient in the vast majority of cases, extremely rigid curves may require previous anterior release through multiple levels discectomy and hemivertebrectomies before performing posterior osteotomies and fusion. Besides, overcorrection of huge curves, greater than 80°, might end up with severe tension on spinal cord, which may lead to intraoperative neuromonitoring issues and postoperative disabilities. In

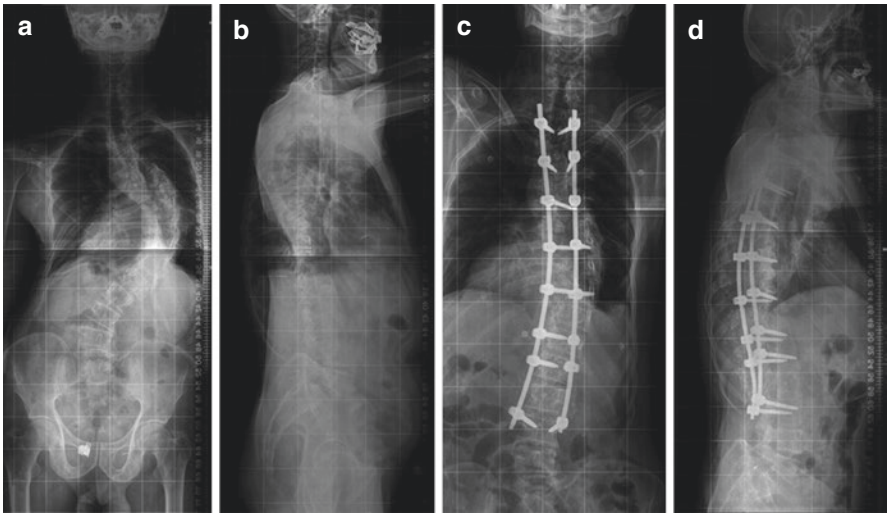

Fig. 48.1 (**a** and **b**) Preoperative X-ray images of a patient with AdIS with the thoracic Cobb angles of 82°. (**c** and **d**) After surgical correction, the thoracic Cobb angle was decreased to 22°

order to prevent this potential complication, anterior spinal column shortening through multiple level anterior discectomy and hemivertebrectomies should be considered before going through posterior fusion surgery. In certain cases, even only discectomies could suffice for appropriate release without additional vertebrectomies. Anterior release through multiple level discectomies is also viable through thoracoscopic approach in thoracic levels, making it advantageous in terms of lowering the morbidity on patients.

Thoracic curves should be treated with selected thoracic fusion with avoiding fusing lumbar level as much as possible to preserve lumbar motion. Proximally, as high as T2 or T3 should be included in a fusion in order to provide better shoulder balance.

Fusion level in thoracolumbar and lumbar curves should be extended to the neutral vertebra proximally, while L3–L4 levels being included distally if they are not part of the curve. In case L3–L4 levels are part of the curve, the fusion should be extended to L5 level.

Whether S1 is included or not in the fusion level is a controversial issue. It is a well-known fact that preserving the L5–S1 level provides retention of lumbosacral motion, lowers pseudoarthrosis rate, reduces operation time, and stress on the SI joint. However, in case of severe disc degeneration, spinal stenosis, spondylolisthesis, and laminectomy at L5–S1 level, including this level in the fusion is mandatory.

When L5–S1 is included in the fusion, anterior fusion may be necessary, and this can also be utilized through the posterior approach with posterior (transforaminal) lateral interbody fusion.

Selection of levels to be fused is of utmost importance, and proximal junctional kyphosis (PJK) is possible.

48.7 Expected Outcomes

The outcomes of AdIS surgery are highly satisfying as the pain relief occurs postoperatively in most patients.

48.8 Potential Complications

General postoperative complications such as pneumonia, atelectasis, ileus, delirium, and pulmonary embolism can be seen. Spinal surgery-related complications include hardware failure, failure of fusion (pseudoarthrosis) (5% to 25%), infection (0.5% to 8%), and neurological compromise (0.5% to 5%). On the other hand, the involvement of neuromonitoring intraoperatively has decreased complications considerably.

48.9 What Patient and Family Should Know?

Surgical treatment methods of AdIS are promising and sometimes very satisfying in carefully selected patients who do not respond to conservative treatment.

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De Novo Deformity

49

Kenny Y. H. Kwan and Kenneth M. C. Cheung

49.1 Definition

De novo deformity refers to spinal deformity in patients with no history of scoliosis during childhood or adolescence. Although traditionally this refers to a deformity in the coronal plane that spontaneously develops during adulthood, we now understand that adult spinal deformity can affect all three planes (Video 49.10). De novo deformity arises from degenerative changes in the spine, and aging, osteoporosis (Chap. 10), degenerative disc disease (Chaps. 44, 45, and 47), facet joint arthrosis, and sarcopenia can all contribute to its development.

49.2 Natural History

Little information is available on the prevalence, incidence, and risk factors associated with curve progression in de novo degenerative scoliosis in the general population. In a community-based cohort study with 12-year follow-up, de novo deformity has developed in 29.4% of patients during the study period, and the risk factors for de novo deformity development were smaller L4 size, unilateral osteophyte formation, and lateral disc wedging. In terms of it natural history, in a systematic review of the literature in which 12 studies were included, strong evidence indicated that increasing intervertebral disk degeneration, lateral vertebral translation ≥6 mm, and an intercrest line through L5 (rather than L4) were associated with curve progression.

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Moderate evidence suggested that apical vertebral rotation grade II or III was associated with curve progression. However, no clinical risk factors were identified. It is also unknown if these risk factors are directly applicable to individual patients.

49.3 Physical Examination

Patients with de novo scoliosis present with complaints ranging from debilitating back or lower extremity pain and spinal imbalance to incidental findings on lumbar radiographs (Chap. 41). Hence, physical examination should focus on what the clinical complaints are. With the patient standing, an inspection of the patient's back, noting any coronal imbalance, abnormal posture including the sagittal plane, and presence of rib-on-pelvis deformity that may lead to inefficient breathing and decreased appetite (Fig. 49.1). The general muscle mass and nutritional status should be noted, and palpation of the back should be performed to identify the site(s) of pain. Gait and peripheral nerve examination and special tests for sciatica should be performed (Video 49.4).

The purpose of the physical examination is to identify any clinical problem that can be correlated with the relevant imaging, hence formulating a targeted management plan.

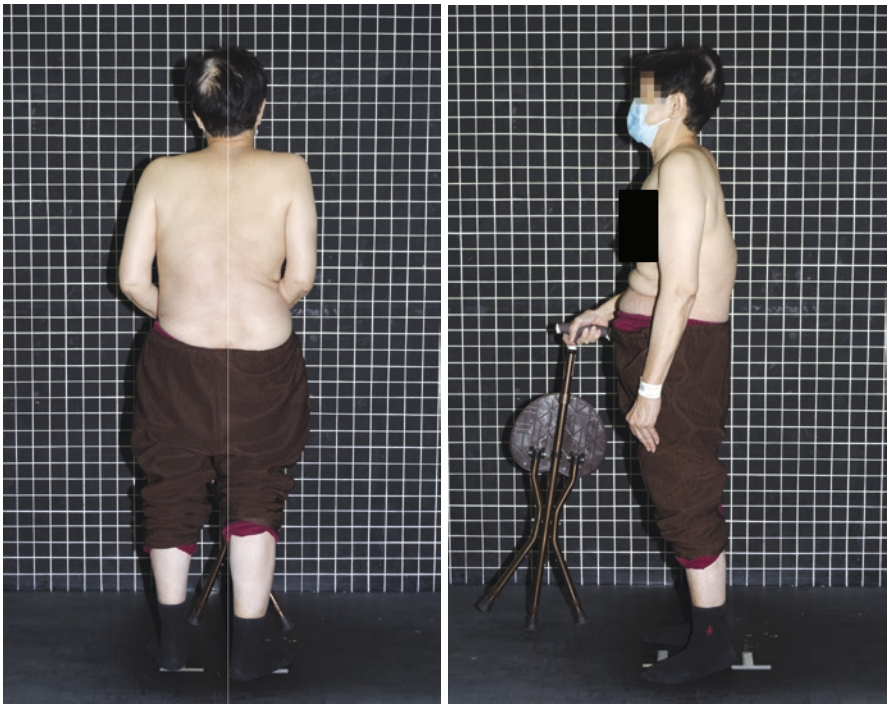

Fig. 49.1 Clinical appearance of a female patient with de novo deformity. The patient has typical sagittal alignment problems such as flat back besides the coronal deformity

49.4 Imaging

Although the lumbar spine is typically involved, the standard radiographic examination should consist of whole spine 36-inch long-cassette standing radiographs in the anteroposterior and lateral views (Fig. 49.2). This allows an accurate assessment of the true magnitude of the coronal deformity, presence of lateral subluxation of the

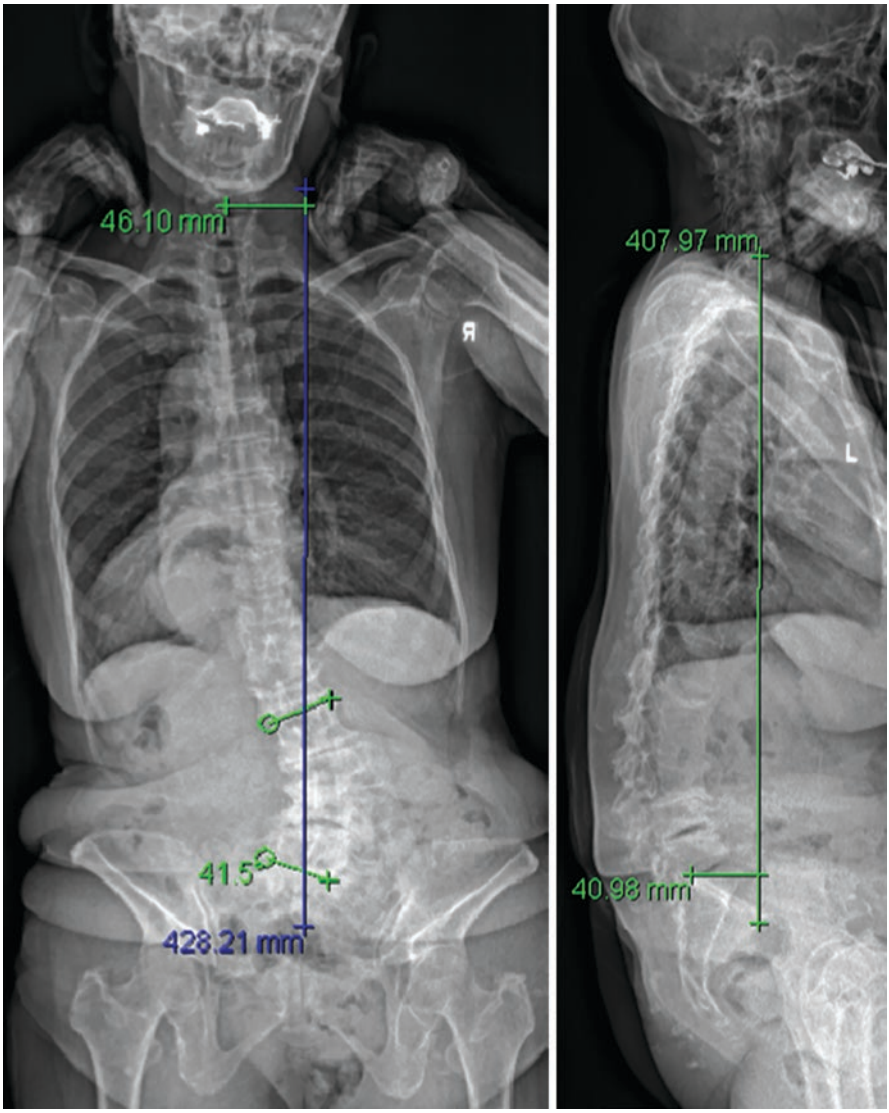

Fig. 49.2 Anteroposterior (AP) and lateral (Lat) long-cassette radiogram of a de novo deformity. The patient has a lateral subluxation of the apical vertebra and osteophytes around the apex in the AP view. The Lat view shows a lumbar kyphosis due to axial rotation. This mechanism is demonstrated with an animation in Fig. 49.1

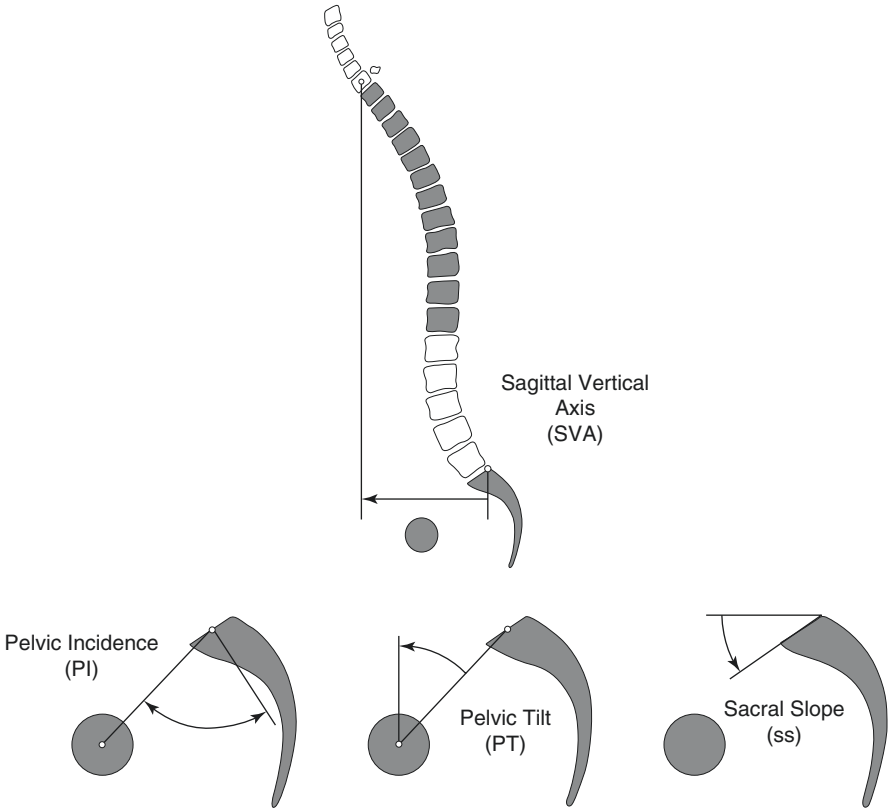

Fig. 49.3 Sagittal and spinopelvic parameters which can be used in deformity analysis and preoperative planning

vertebra, and tilting of the lower lumbar vertebrae and pelvis (Video 49.6). On the lateral view, both femoral heads should be included in the film to allow the determination of the pelvic incidence. In addition, regional and global sagittal balance parameters can be determined (Fig. 49.3). Using these parameters, these deformities can be classified using the Aebi classification or the now more commonly used SRS-Schwab classification (Fig. 49.4) (Chap. 50).

In instances where the anatomy is not very clear on plain radiographs, computed tomography can be requested. In patients where the status of the disc and the spinal canal needs to be visualized, magnetic resonance imaging should be obtained.

49.5 Differential Diagnosis

Curve patterns and locations may indicate if the deformity is due to neglected adolescent idiopathic scoliosis, and they will often present younger than the de novo curves. Other diagnoses of spinal deformities such as neuromuscular, syndromal,

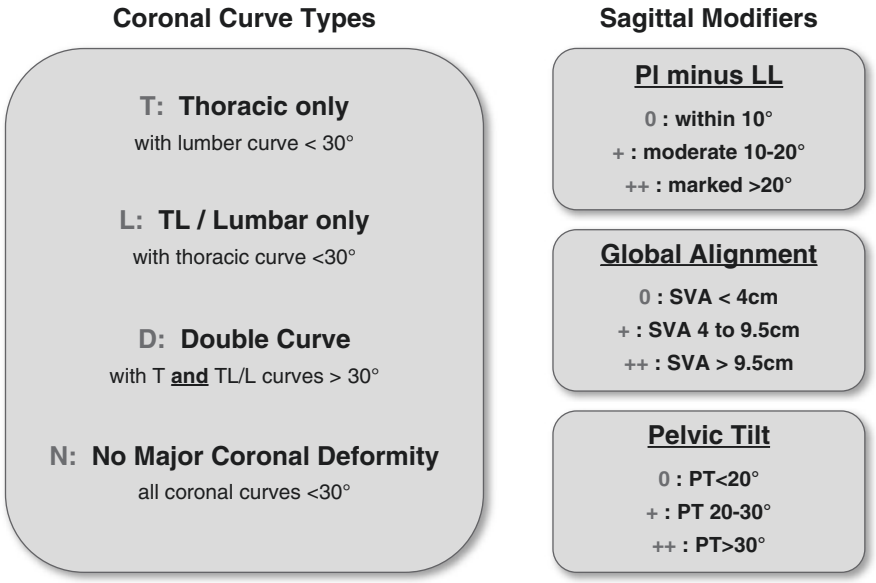

Fig. 49.4 SRS-Schwab classification for the adult deformity

traumatic, and congenital should be obvious from the patient's history and physical examination.

49.6 Treatment Options

De novo deformity of the spine per se in adulthood does not require surgical correction if the patient is completely asymptomatic. Hence, a comprehensive clinical history including the chief complaints, functional limitation, and presence of neurologic deficit is key in determining the treatment regimen. It is established that coronal deformity and imbalance are better tolerated to a certain degree than sagittal imbalance.

Nonoperative treatment includes pharmacotherapy, lifestyle modifications, isometric core, and back muscle strengthening exercises that have their roles in alleviating patients' symptoms and improving quality of life. The use of brace preceded by plaster cast to test correction (of lumbar kyphosis, in particular) may work sufficiently well to avoid surgical treatment in some cases. Nonetheless, most research has shown that patients with severe sagittal imbalance do not improve with conservative care.

Diagnostic nerve root and facet joint blockades are useful adjuncts in the decision-making of complex de novo deformity, where the pain generators are not immediately obvious from the clinical history or physical examination. It allows the

surgeon to determine the goals of any surgical intervention that is required and, at the same time, can give the patients some temporary relief to maximize the efforts of conservative care.

The aims of any surgical interventions are to improve patients' quality of life by decompression of neural elements, stabilization of spinal segments, and correction of spinal deformities in the coronal and sagittal planes (Videos 49.2, 49.3, 49.7 and 49.10). However, the alignment goals, particularly of the sagittal plane, are still under intensive investigation. Both the SRS-Schwab (Fig. 49.4) and Roussouly/Global Alignment and Proportion (GAP; it evaluates the following parameters: (a) sacral slope, (b) L1–S1 lordosis, (c) L4–S1 lordosis, and (d) global tilt) classifications give some guidance on the degree of deformity correction is needed, but long-term results are difficult to assess due to the heterogeneity of this patient population (Chap. 50). Preoperative dual-energy radiograph absorptiometry should be obtained to assess the bone mineral density.

49.7 Expected Outcomes

For patients with significant limitations in the quality of life with progressive deformity, nonoperative treatment with standard conservative care has not shown to be effective although brace treatment may provide pain control during daytime. Surgical correction in this group of patients leads to significant improvements in their quality of life, which also applies to patients who suffer from postoperative complications and unplanned return to the operating theaters. However, potential surgical candidates should be made aware of the high complication profile of such procedures (both neurologic and non-neurologic events) during the shared decision-making process.

49.8 Potential Complications

It should be noted that many patients will not suffer from any degree of functional limitation and will not experience any complications. Nonetheless, untreated de novo scoliosis may result in curve progression, neurologic deficit, pain, and limitation in functional activities.

49.9 What Patient and Family Should Know?

De novo scoliosis may be an incidental finding or the cause of back pain and functional limitation in the adult population. Treatment options depend on patients' symptoms and neurologic status. Although surgical interventions carry high complication rates, most studies suggest a significant improvement in the quality of life and a high rate of patient satisfaction.

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Sagittal Plane Malalignment

50

Caglar Yilgor, Altug Yucekul, and Ahmet Alanay

50.1 Definition

While the spine, in the ideal standing posture, is straight in the frontal plane, it has various physiological curvatures in the sagittal plane such as cervical lordosis, thoracic kyphosis, and lumbar lordosis. The magnitude and location of these curvatures differ from one person to another (Video 50.6). Sagittal plane malalignment refers to the condition where the magnitude and/or shape of these curvatures has deviated from the normal range of values. A range of pathologies may lead to sagittal malalignments such as scoliosis (Chap. 16), spondylolisthesis (Chap. 25), degenerative disease (Chap. 47), and iatrogenic flat back.

50.2 Natural History

Once bones, joints, discs, and ligaments lose their anatomical and physiological qualities via trauma, various spinal diseases, and through the normal aging process, spinopelvic alignment begins to deteriorate. The natural evolution of the sagittal plane is increased kyphosis, loss of lordosis, pelvic retroversion, and

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positive global tilt. Yet, human beings, through subconscious mechanisms, have a tendency (1) to stand in the most ergonomic position that leads to the lowest energy consumption while standing and walking, (2) to position their heads over the pelvis, and (3) to maintain a forward gaze. In cases where the spine deviates from this "ideal" ergonomic, so-called compensatory mechanisms are activated. These mechanisms use "reserves" located in the spine and in non-spinal body segments (particularly legs) in order to maintain the upright posture. The use of these reserves requires active muscle contraction and increases energy consumption related to mobilization.

50.3 Treatment Options

Today, there are three comprehensive analysis models based on different approaches: the SRS-Schwab criteria focusing on the clinical impact, the Roussouly back types based on shapes and biomechanics, and the Global Alignment and Proportion (GAP) score using a PI-adjusted mathematical approach.

50.3.1 SRS-Schwab Classification

The criteria for the SRS-Schwab classification are pelvic tilt (PT), pelvic incidence (PI) minus lumbar lordosis (PI–LL), and sagittal vertical axis (SVA) [1]. Measured values are categorized as normal, moderate, and severe. For these three criteria, target values are SVA <5 mm, PT <25°, and PI-LL ±9°. It has the advantages of relating to patient-reported outcomes and ease of use. Conversely, disadvantages include the absence of anteversion, negative alignment and distribution of lordosis, and the position-dependent nature of the use of SVA. More importantly, these target values have been determined by considering population-based averages and are the same for all individuals.

50.3.2 Roussouly Back Types

Roussouly classification describes the spinal shapes of the curvatures by defining the apex and the number of spinal segments within each curvature and the inflection point between opposing curvatures. Five different spine types with distinguishable characteristics have been defined, in normative databases [2]. This approach is the first to classify patients instead of using universally constant targets. It accounts for the distribution of loads and defines compensatory mechanisms and degeneration processes. Disadvantages include difficulty of determining the original spine type once degeneration/pathology occurs and difficulties experienced in terms of interpretation and explication since this is an analysis method based on visuals.

50.3.3 GAP Score

The concept of chain of correlations forms the basis of the GAP Score. It implies that the amount of every spinal curvature affects the amount of the next curvature. The pelvis acts as the link between the spine and legs, and it is considered to be the foundation of the spine. Thus, sagittal angular width (PI) and tilt of the pelvis is in close relation with the sagittal spinal curvatures. For example, a patient with a larger horizontal diameter of the pelvis will have a more tilted pelvis while standing; a patient with a more tilted pelvis will have a deeper lumbar lordosis; a patient with a deeper lordosis will have a larger thoracic kyphosis; and a patient with a larger kyphosis will have a deeper cervical lordosis, and vice versa.

The PI, having a wide distribution within the population that does not change during adult life, is considered as a signature of a given individual. Since all other curvature measurements are affected by diseases and degeneration, their absolute values cannot be directly used. In GAP analyses, all sagittal plane parameters are subject to assessment in proportion to PI and calculated as deviations from the "ideal," in a PI-adjusted manner. This personalized approach is developed from a spinal deformity database, which includes patients treated for spinal disorders, by taking mechanical complications into consideration [3].

Besides the concept of using PI-adjusted relative angular values for the pelvic, lordotic, and global alignment measurements, the GAP score also comprises the mathematical formulation of the distribution of lordosis. Similarly, ideal lordosis distribution has different thresholds depending on the PI. The fifth parameter contributing to the GAP score, age factor, is a surrogate for the biology of aging such as osteoporosis, neurodegenerative diseases, and sarcopenia. The numerical value of the GAP score indicates the deviation from ideal and the amount of compensation used and defines the spinopelvic state into proportioned, moderately disproportioned, and severely disproportioned (Fig. 50.1).

The main advantage of this approach is that it does not categorize patients but instead calculates their standing posture as a continuum of states. It denotes normal and pathologic in a single score and helps differentiate between deformity and compensation. More importantly, it correlates with the mechanical complications and satisfaction related to the treatment. Disadvantages include controversy in external validation efforts and lack of prospective evaluation.

50.4 Expected Outcomes

Malalignment in the sagittal plane affects the health-related quality of life and presents generally with pain and disability (Chap. 41). It is anticipated that spinal diseases will be of concern to >60% of people who are above 60 years of age. When compared to cardiac diseases, diabetes, and chronic obstructive pulmonary disease, it was reported that spinal conditions affect the public health at least as much as

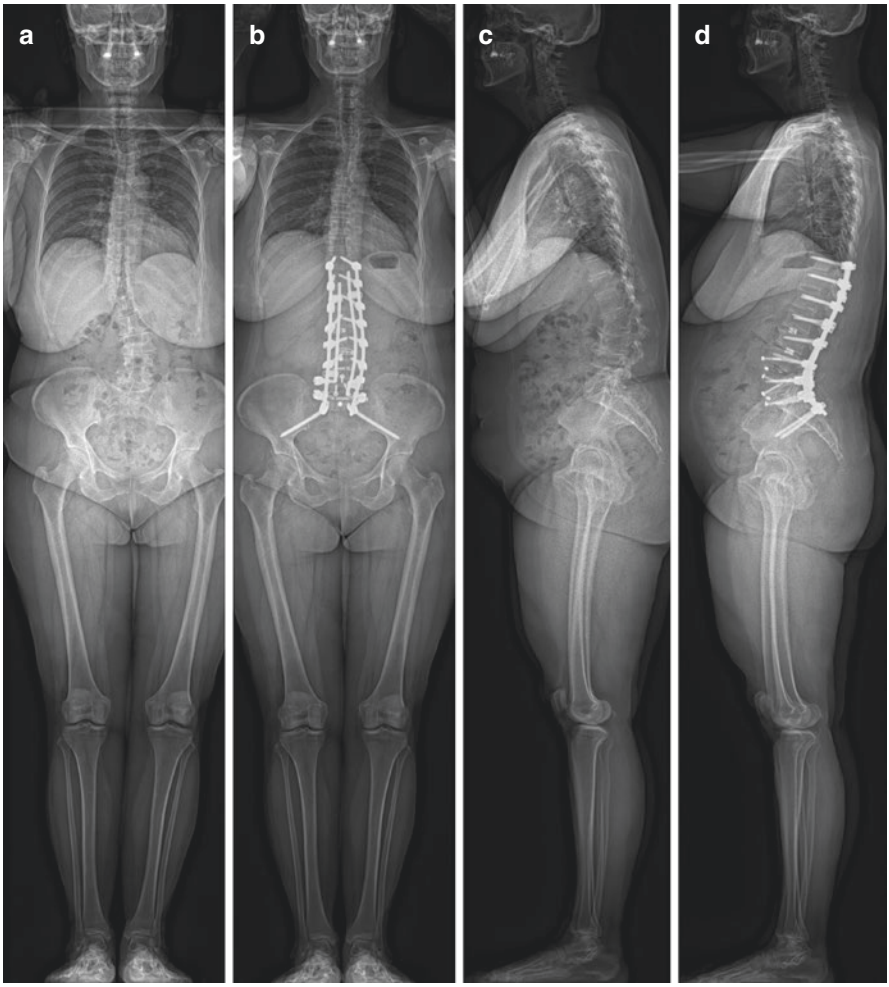

Fig. 50.1 (**a**) A 56-year-old patient applied to the clinic with complaints of back pain, left leg pain, and claudication. Her neurological examination revealed 3/5 strength in ankle plantar flexion and 2/5 strength in extensor hallucis longus. She had a history of numerous conservative treatment sessions over the past couple of years. (**b**) She was treated with two-staged operation, in which L4–L5 and L5–S1 anterior lumbar interbody fusion and L2–L3 and L3–L4 transforaminal lumbar interbody fusion with T10 to pelvis posterior instrumented fusion was performed. (**c**) Preoperatively, lumbar vertebrae showed a kyphotic orientation with sagittal malalignment. (**d**) Segmental and total lordosis correction magnitudes were planned according to GAP score and interbody cage sizes, and wedge angles were decided accordingly

these chronic diseases. Therefore, the preservation of spinal health gains more importance for a person's quality of life.

Further importance of the sagittal alignment lies in its relation to the occurrence of mechanical complications. Various studies indicated patient-related factors,

technical factors, and sagittal plane to be the most important factors affecting the development and prevention of mechanical complications. The most common errors in the sagittal plane are being unable to achieve personalized curvatures and, consequently, performing under- or overcorrection (Video 50.3).

Most complications that result in mechanical problems consist of proximal and distal junctional kyphosis and failures, adjacent segment degeneration, implant-related complications (screw loosening, cage and screw pullouts, etc.), nonunion, and rod fractures. Approximately 50% of patients who experience a mechanical complication require a second intervention.

50.5 What Should Patient and Family Know?

Sagittal alignment is complex and hard to interpret. It is obvious that every individual has unique anatomy that is regulated mainly by the pelvic shape. Sagittal malalignment and accompanying compensatory mechanisms may be the underlying cause of back pain for many patients with degenerative conditions. Thus, treatment success and avoidance of complications rely on the subject-specific evaluation and adapting a personalized treatment planning.

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Post-Traumatic Kyhphosis

51

Meric Enercan and Azmi Hamzaoglu

51.1 Definition

Post-traumatic kyphosis (PTK) is defined as painful kyphotic angulation of the post-traumatic spine. PTK most often presents after major trauma resulting in spinal column fracture but can also present following minor trauma in patients with poor bone quality.

51.2 Natural History

PTK can involve untreated patients, patients with failed conservative treatment, and patients with unsuccessful surgery. Regardless of the etiology or the initial treatment, the most common complaints are (i) progressive deformity, (ii) increasing neurologic deficit, (iii) sagittal and/or coronal plane imbalance, (iv) increasing pain, and (v) cosmetic and functional deterioration. There are many factors that define the characteristics of PTK deformity. The level and the magnitude of the injury are the main contributors. The PTK deformity typically occurs at the thoracic and thoracolumbar spine. Compression fractures are unlikely to produce a progressive deformity since the middle and posterior columns are left intact. If the local kyphosis exceeds 20°, concomitant injury of the posterior ligamentous structures may lead to progressive deformity. If the injury is more severe, such as a severe burst fracture or a flexion-distraction

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injury in which the anterior, middle, and posterior columns are disrupted, PTK is likely to be greater and progressive in nature (Chap. 7). PTK patients present with two different types of sagittal deformity. Type I sagittal deformity is characterized by normal overall sagittal balance with a focal kyphosis; type II sagittal deformity is characterized by focal kyphotic deformity with global sagittal imbalance (Chap. 50).

The progression of PTK can cause a new or worsening neurologic deficit by direct compression of the neural elements, tenting of the neural elements across the angular kyphosis, and mechanical stress on the spinal cord. The development of post-traumatic syringomyelia also leads to the development or progression of the neurologic deficit. Up to 50% of patients develop spinal cystic changes, and 21–28% of the patients may develop a syrinx even 30 years after the initial spinal cord injury. Post-traumatic syringomyelia usually presents with segmental pain and sensory loss, followed by progressive asymmetrical weakness.

Pain is one of the most common symptoms of PTK, and it is generally secondary to abnormal spine biomechanics at the apex of the kyphotic deformity; it results in altered forces acting on the soft tissues and surrounding structures. Patients with a focal kyphotic deformity greater than 20° are at increased risk to develop chronic pain with poor functional tolerance. Pain may also be secondary to canal stenosis or neural foramen compromise, nonunion, instability, and adjacent compensatory changes that occur above or below the kyphotic deformity. Hyperlordosis in the lumbar spine or hypokyphosis/thoracic lordosis above the deformity may be a source of chronic fatigue and pain.

PTK often presents in two different patterns, malunion or nonunion, with different courses. Malunion is essentially a stable late deformity that carries the risk of spinal stenosis. Nonunion or pseudoarthrosis is more similar to unstable late deformity with risks of acute instability (Chap. 47). Table 51.1 outlines fracture-related risk factors for malunion and nonunion (Table 51.1).

51.3 Physical Examination

A detailed history and physical examination must be performed in every patient. Injury patterns and previous treatment interventions should be noted. The patient should be observed undressed while standing and walking. Any compensatory

Table 51.1 Fracture-related risk factors for PTK with malunion and nonunion

Fracture-related risk factors for PTK with malunion	Fracture-related risk factors for PTK with nonunion
• Sagittal index >20°	• Burst fractures with vertebral body height loss >50%
• Vertebral body height loss >30%	• Extension-type fractures effecting three columns
• Local kyphosis angle >30°	• Dead bone sign in traumatic osteoporotic fractures
• Posterior ligamentous complex and posterior osseous injuries	• Severe posterior ligamentous ligament injury
	• Significant posterior distraction or lateral translation on plain radiographs

alignment changes and contractures of the hips and knees should be noted. Neurologic examination should include sensory, motor power, deep tendon reflexes, nerve root tension signs, and gait pattern (Video 51.4). Video recording of patients' gait gives more details about the gait pattern and enables comparison with postoperative gait. Photographs should be taken from the front, back, and sides in standing position for preoperative coronal and sagittal plane balance analyses (Chap. 50).

51.4 Imaging

1. Sagittal and coronal spinal parameters should be assessed with standing anteroposterior and lateral full-length spine radiographs. Local kyphosis angle should be measured between the superior end plate of the adjacent cranial vertebra and the inferior end plate of the adjacent caudal vertebra (Video 51.6). The sagittal index is obtained by the difference in degrees of the kyphotic deformity minus the normal contour in the corresponding area. Hyperextension fulcrum or prone lateral radiographs can be used to assess the flexibility of the kyphotic deformity. Dynamic lateral radiographs can be helpful in detecting instability, pseudoarthrosis, hardware failure, and adjacent segment disease.
2. Computerized tomography (CT) is used for defining the bony anatomy, including the degree and pattern of fracture healing and fusion status. CT is also helpful to assess bony anatomy with previous surgery. Three-dimensional (3D) CT scan reconstruction can be helpful in preoperative planning.
3. Magnetic resonance imaging (MRI) is used to evaluate the posterior osteoligamentous complex, to determine the integrity of the spinal cord, and to assess the integrity of the surrounding intervertebral discs and the presence of disc herniation.
4. CT myelography is indicated when MRI is contraindicated (e.g., cardiac pacemaker) or not possible due to previous instrumentation.

51.5 Differential Diagnosis

Other etiologies of the kyphosis including postinfectious kyphosis (Chap. 52), congenital kyphosis (Chap. 24), and Scheuermann kyphosis (Chap. 22) should be excluded.

51.6 Treatment Options

The goals of surgery are to obtain a satisfactory balance in both sagittal and coronal planes, to attain a solid fusion with balanced spine, to relieve pain, to improve neurologic deficits if present, and to prevent further deformity. In order to successfully address a PTK, the surgeon must consider the areas of neural compression, the magnitude of the focal PTK, the overall sagittal and coronal balance/alignment, the flexibility and character of the deformity, and the location of the deformity.

Surgical options are all-anterior, simultaneous combined anterior-posterior surgery, or posterior-only surgery. All anterior approaches allow direct access to the anterior and middle column and anterior decompression. Morbidity related to anterior surgery is the major limitation. In the presence of the significant and rigid deformity, the correction will be limited. Combined anterior-posterior surgery imposes three stages; the first stage includes posterior instrumentation, posterior release, and temporary posterior fixation. The second stage includes anterior decompression and anterior column restoration with or without anterior instrumentation. The third stage involves final posterior reduction and fixation. In recent years, posterior-only surgery is being preferred to combined approaches. Posterior osteotomy options are posterior-column osteotomy (Ponte) or three-column osteotomies. Three-column osteotomy options are pedicle subtraction osteotomy (PSO), bone-disc-bone osteotomy (BDBO), and posterior vertebral column osteotomy (PVCR). Flexible global kyphosis can be managed with single or multiple posterior-column osteotomies, whereas patients with sharp angular kyphosis with major sagittal imbalance require three-column osteotomies (Videos 51.3 and 51.6). The PVCR osteotomy provides the most complete mobilization of the spine for deformity correction in all planes, enables anterior column reconstruction, and is useful for rigid deformities with very significant sagittal or coronal imbalance.

51.6.1 Correction Technique for Rigid Sharp Angular Kyphosis Following PVCR

The correction should be applied in a stepwise manner. Placing a temporary cage will prevent sudden shortening, dural buckling, and iatrogenic neurologic deficit. When the sharp angular kyphotic deformity is corrected by 30% to 50%, temporary mesh cage is removed and replaced by a lamina spreader. The osteotomy site is distracted and lengthened anteriorly with the lamina spreader, while simultaneous compression is applied posteriorly. When the anterior gap is lengthened 50% or more compared to the initial status, an expandable cage is placed anteriorly at the osteotomy site. The expansion of the expandable cage provides additional anterior lengthening, and final correction is achieved by posterior compression maneuvers. Anterior lengthening and posterior compression will be continued until ideal sagittal alignment is achieved (Fig. 51.1). PTK secondary to malunion and nonunion can be managed successfully with the PVCR technique (Figs. 51.2 and 51.3).

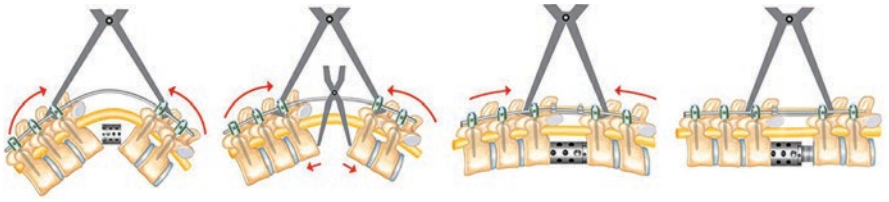

Fig. 51.1 Correction technique of PTK correction following PVCR with sequential posterior compression and simultaneous anterior column lengthening technique

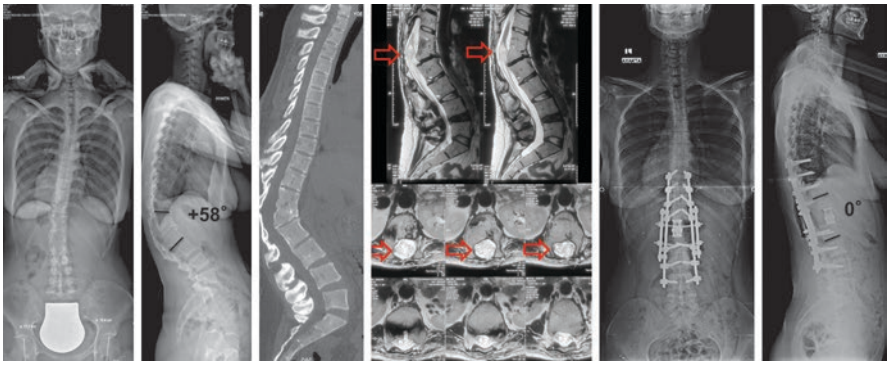

Fig. 51.2 A 30-year-old female patient with PTK due to malunion. She had a previous failed surgery and significant neurologic deficit due to post-traumatic syringomyelia. Following L1 PVCR, sequential posterior compression and simultaneous anterior column lengthening technique was performed for deformity correction

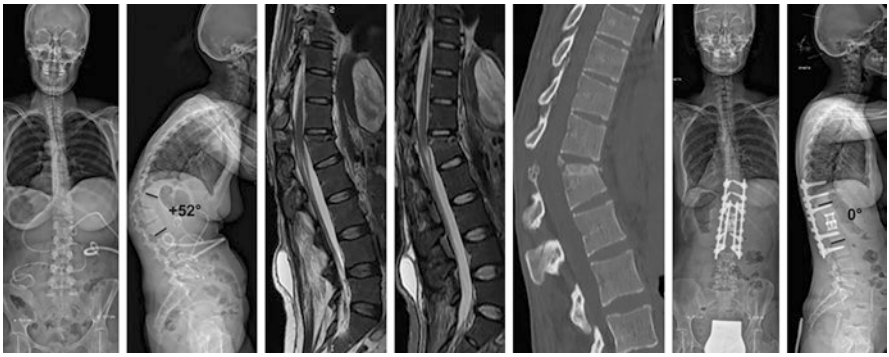

Fig. 51.3 A 42-year-old female patient with PTK presented with nonunion. PVCR was performed, and an expandable cage was used in order to reconstruct anterior column support and restore the ideal thoracolumbar sagittal alignment

51.7 Expected Outcomes

If a post-traumatic kyphosis is left untreated, there is a high risk of chronic back pain, and this risk increases in the patients whose kyphosis is located in the lumbar region. A progressive post-traumatic kyphosis can be related to neurologic compromise. The outcome of the surgical treatment is satisfactory if the treatment is performed in highly specialized centers.

51.8 Potential Complications

PTK surgery is technically challenging and prone to complications. Various studies reported a more than 60% rate of overall complication rate with a 10% reoperation rate. The most commonly reported complications are neurological complications,

pulmonary and urinary complications, bleeding, dural tear, pseudoarthrosis, implant failure, and infection (Chap. 66). The neurological complications occur in 6.3–15.8% and are transient in most cases. Patients with preoperative neurologic deficits had a higher risk of postoperative permanent deficits.

51.9 What Should Patient and Family Know?

Treatment of PTK is often challenging, and successful treatment depends on careful patient selection and appropriate surgical intervention. Surgical treatment should address decompression of neural elements and restoration of ideal sagittal alignment and achieve solid fusion. In recent years, posterior-only surgery is being preferred to anterior or combined approaches. Among posterior osteotomies, PVCR provides the most complete mobilization of the spine for deformity correction. Proper correction techniques should be performed to achieve ideal correction and prevent complications.

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Post-Infectious Deformity

52

Shahnawaz Haleem

52.1 Definition

Post-infectious kyphosis is the loss of sagittal alignment caused by destruction of intervertebral discs or adjoining vertebral bodies due to infective process.

52.2 Natural History

Spinal infections (pyogenic and non-pyogenic) can occur either due to blood-borne infections or local inoculation peri-operatively (Chap. 66), followed by increased pain in the affected area. Haematogenous infection in capillary loops induces the inflammatory cascade which leads to bony destruction and collapse. The infection can spread in all directions leading to paravertebral and epidural abscesses and adjacent disc involvement. The patients' symptoms and signs will depend specifically on the primary site of infection, its area of spread, the duration of infection, any ongoing infection, the organism involved and immune status of the patient.

While pain and fever are usually present, involvement of the epidural space can affect neural structures and loss of stability due to softening of vertebral bones causing a collapse (due to pathological fracture) and resultant cord compression which can lead to a neurological deficit. The onset of paralysis may occur early or late. Delayed onset of paralysis may be due to the vertebral destruction causing either bony impingement or a kyphotic deformity.

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The clinical picture may be varied and lead to a delay in diagnosis and therefore correct appropriate antibiotic management and surgical intervention. The most common organisms are *S. aureus* and *S. epidermidis* with *Pseudomonas aeruginosa* being most commonly seen in intravenous drug users (Chap. 65). *Mycobacterium tuberculosis* is the most common non-pyogenic organism (Chap. 64).

52.3 Physical Examination

Symptoms and signs depend on any ongoing infection. The majority of patients present with pain (on any form of movement), night sweats, intermittent fever, anorexia and weight loss. Paralysis is not an early presentation in most patients but when present is rapidly progressive. Occasionally, patients present with myelopathy due to the stretched spinal cord at the apex of sharp kyphosis so-called gibbus. Gibbus is a sharp-angled kyphosis and pathognomonic for post-tuberculous kyphosis.

Physical signs included localised tenderness, muscle spasm (paraspinal and torticollis depending on site of infection), hamstring spasm and generalised weakness. A pointing abscess is rare, and straight leg raise (SLR) test may elicit back pain and rarely radiculopathy (Video 52.4).

The location/level of the infective process may dictate the physical signs as deformity occurring in the cervical and thoracic spine with cause increased neurological deficit as compared to the thoracolumbar spine. The patient therefore may present with upper motor neurone (UMN) signs in higher lesions with a sensory level or flaccid weakness in lower motor neurone (LMN) lesions depending on the level of cord compression. Previous cord injury due to infection can lead to central cord/anterior cord syndromes with their features on examination. Central cord syndrome presents with increased weakness in the upper limbs as compared to the lower limbs, and anterior cord syndrome affects the anterior two-thirds of the spinal cord resulting in motor paralysis below the level of the lesion and loss of pain and temperature at and below the level of the lesion.

It is therefore imperative to include ASIA (American Spinal Cord Injury Association) chart scoring from the first examination as a written record to ensure any improvement or deficit is correctly monitored (Appendix G). Any new deficit will then require repeat imaging to gain further information and guide appropriate management.

52.4 Imaging

Radiographs are not very sensitive to early spinal involvement, and the first radiographic finding may be seen 2 weeks to 3 months after infective onset. These are usually endplate erosions. Other early changes include loss of endplate normal contour, subchondral bone defects and sclerotic bone formation. Late radiographic changes may reveal collapse of vertebrae, bony column deficits, segmental

kyphosis and bony ankylosis (fusion). Standing long-cassette anteroposterior and lateral radiograms should be taken for better evaluation of the spinal alignment (Video 52.6).

Computed tomography (CT) can identify paravertebral collections, show bony changes (endplate destruction) and lytic/sclerotic features and define neural impingement (Appendix Q). CT-guided biopsy may help obtain deep specimens to rule out any ongoing infection. Magnetic resonance imaging (MRI) can reveal any ongoing infection (especially with gadolinium enhancement), neurological injury (including cord infarction) and ongoing impingement due to gibbus. T2-weighed images are valuable for demonstrating myelopathy as a hyperintense lesion. All neurological deficits need to have up-to-date scans.

52.5 Differential Diagnosis

The differential diagnosis of post-infectious kyphosis should include congenital kyphosis, primary and metastatic tumours (Chap. 63), pathological fractures caused by metabolic bone disease, rheumatoid arthritis and ankylosing spondylitis (Chap. 54), Charcot spinal arthropathy and any other infective process causing bony destruction and kyphosis.

52.6 Treatment Options

The first step is to make sure the causative pathogen has been correctly identified and proper treatment (especially for tuberculosis, TB) initiated. It is presumed that the patient has no ongoing infective issues in this treatment plan which would then depend on the presence of pain, neurological deficit (time since onset and progression), patient mobility status and comorbidities. Nonoperative options in a neurologically intact patient presenting only with pain and no or limited deformity would include pain relief, brace application, physiotherapy and mobilisation. Operative indications are due to presenting and progressive neurological deficit. Here the priority is to relieve the cord compression, improve malalignment (Chap. 50) and stabilise the spine. This may involve surgical decompression, realignment with osteotomies and stabilisation (posteriorly and/or anteriorly) (Videos 52.3, 52.4, 52.6 and 52.7). The length of the pedicle screw and rod construct (number of levels fused) should be carefully planned preoperatively to ensure optimal outcomes depending on the neurological status and degree of kyphosis to ensure that optimal spinopelvic parameters are achieved.

Figure 52.1a and b show postinfectious deformity in a 14-year-old boy suffering from TB since the age of 3 years. He presented with no neurological deficit and only progressive deformity. Figure 52.2a and b show deformity on CT and MR imaging. He underwent a successful stabilisation and remains neurologically intact at latest follow-up (Fig. 52.3a and b).

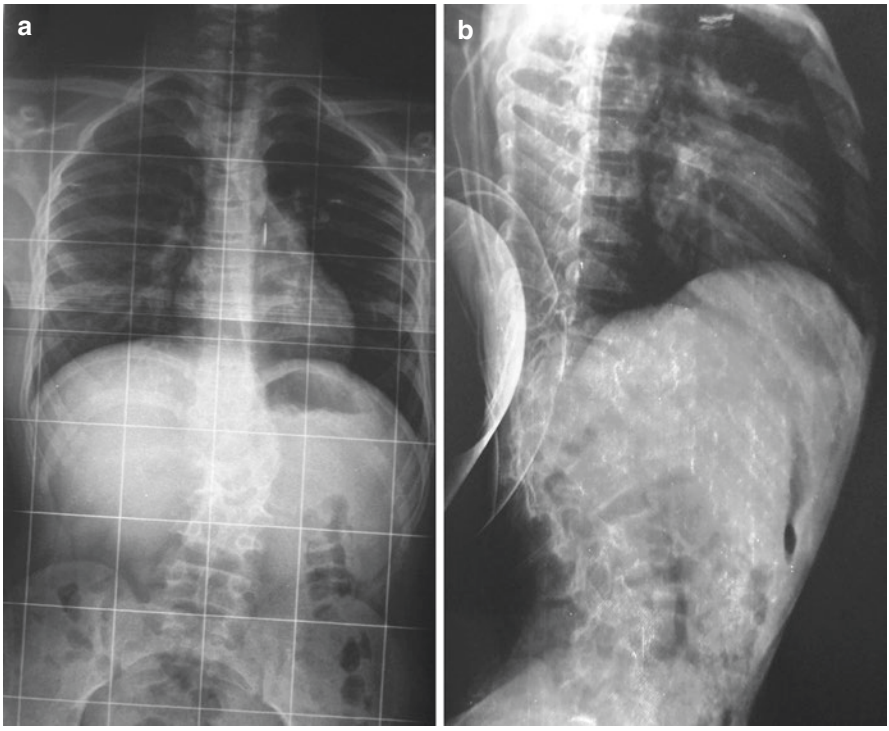

Fig. 52.1 (**a** and **b**) Show a kyphoscoliotic deformity in a 14-year-old boy with a history of TB at first presentation in 2014 [Courtesy of Prof. Alpaslan Şenköylü]

52.7 Expected Outcomes

This depends on the preoperative neurological status and degree of kyphosis. Early surgical intervention is required if the patient presents with neurological deficit. Early surgical intervention involving judicious decompression, optimal realignment and stabilisation (with front and back surgery) will ensure a good outcome. However, once paralysis sets in with a patient who has other comorbidities, then recovery can be poor, and further management will involve rehabilitation in a spinal cord injury centre.

52.8 Potential Complications

These can include infection, bleeding, nerve injury, paralysis, spinal fluid leak, blood clots (legs and lungs), malpositioned metalwork, failure to fuse, metalwork failure, continued symptoms, post-junctional kyphosis and risks of anaesthesia, COVID infection and death.

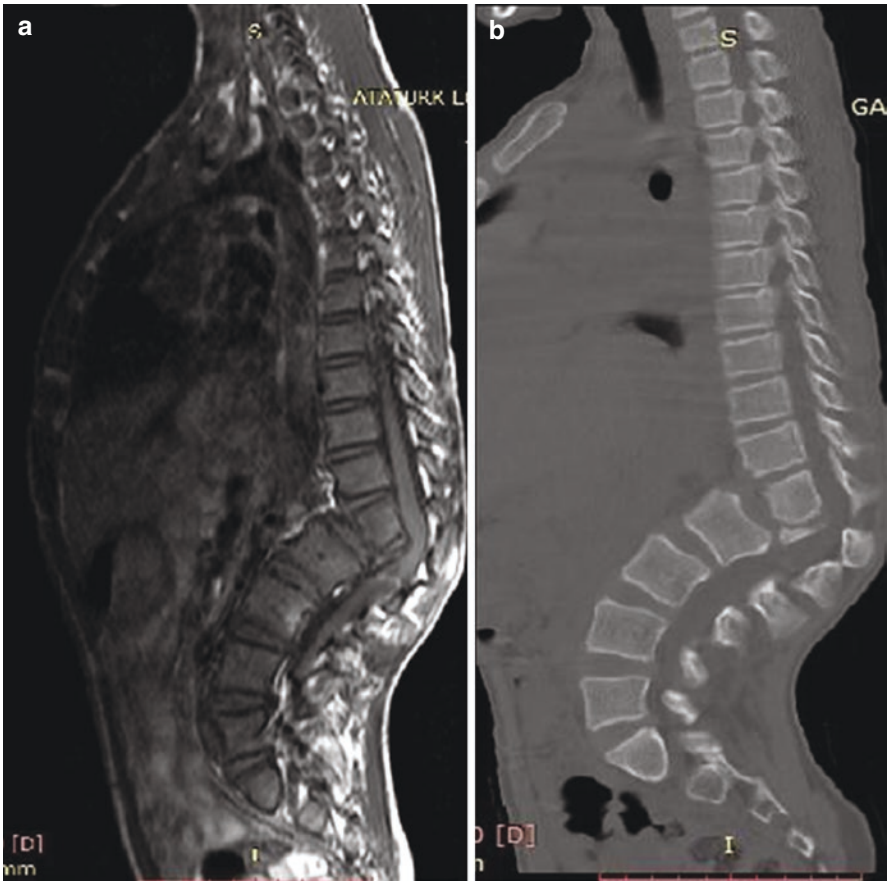

Fig. 52.2 (**a** and **b**) Show CT and MRI imaging revealing a L1 vertebral body collapse and kyphotic deformity. It can be classified as Kyphosis Classification (Rajasekaran) type IIIB (Appendix P) [Courtesy of Prof. Alpaslan Şenköylü]

52.9 What Should Patient and Family Know?

Postinfectious kyphosis may present with pain, deformity and neurological deficit (mild to complete). Treatment will depend on the amount of pain, reduction in activities of daily living, degree of deformity (kyphosis), patients' comorbidities and patients' aspirations. Surgical intervention can carry its own risk of complications including paralysis, rod breakages and a higher risk of infection. While the surgical team will optimise patients preoperatively and plan accordingly, patient factors also play a major role in the recovery process. A patient with multiple comorbidities and poor immune status will not do well with major surgical intervention. Each patient needs to be assessed independently prior to any surgical intervention.

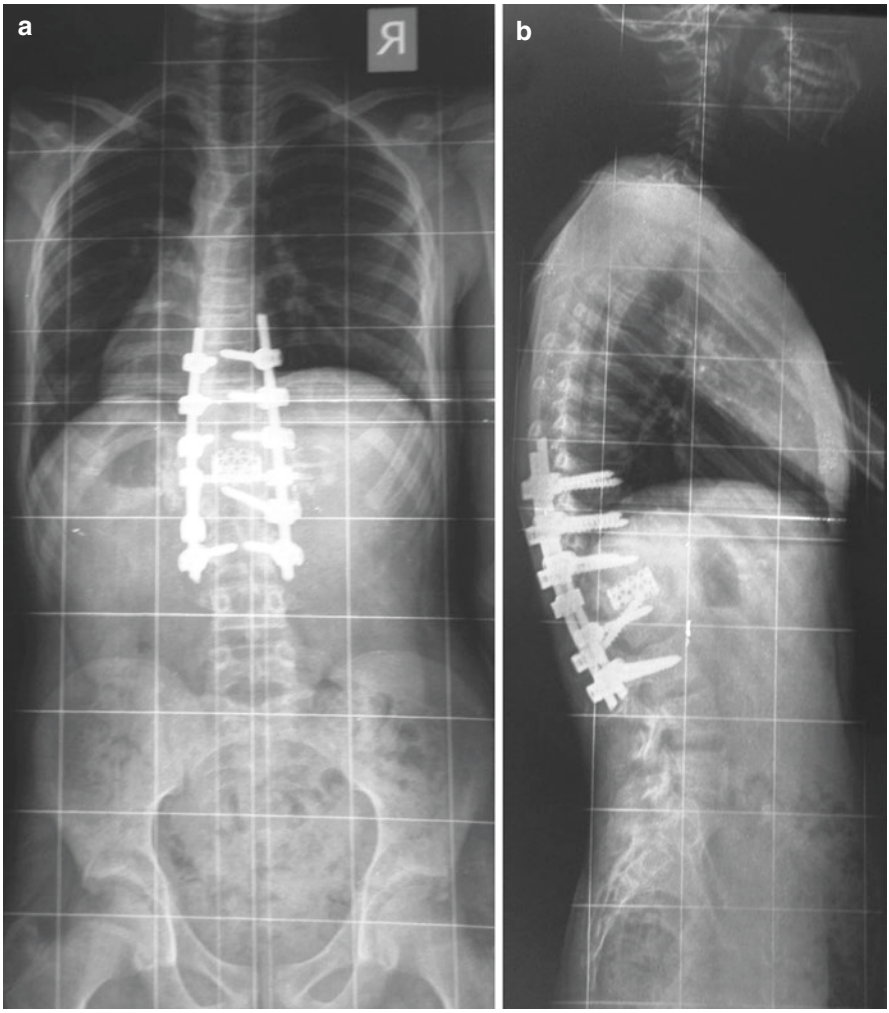

Fig. 52.3 (**a** and **b**) Show satisfactory coronal and sagittal alignment after posterior vertebral column resection [Courtesy of Prof. Alpaslan Şenköylü]

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Paget's Disease

53

Yann Philippe Charles

53.1 Definition

Paget's disease, also known as *osteitis deformans*, represents a metabolic bone disorder. The spine represents the second most commonly affected part of the skeleton after the pelvis. Monostotic and polyostotic patterns exist. Paget's disease is usually diagnosed after the age of 50 years, and the overall prevalence is reported around 2% to 3%, although ethnic differences have been described. The prevalence increases in elderly patients. The etiology and exact pathophysiology remain unclear. On the one hand, viral infections (paramyxovirus, syncytial respiratory virus) have been incriminated. On the other hand, different genetic mutations (sequestosome) might play a role.

53.2 Natural History

The pathophysiology of Paget's disease is characterized by dysregulation between osteoblastic and osteoclastic activity. Three main phases of the disease exist when the spine is involved. The first phase represents the initial osteolytic phase which is represented by mainly osteoclastic activity. The second phase is characterized by a combination of osteoblastic and osteoclastic activities. The third phase represents

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the last stage which is characterized by new bone formation. An additional inactive late stage has further been described as sclerotic phase, where stimulation of new osteoblasts and osteoclasts ceases. During this late stage, the bone is metabolically inactive, but it maintains a sclerotic coarsened architecture.

53.3 Physical Examination

Clinical symptoms of Paget's disease depend on the localization of the skeleton and the stage of the disease. However, patients can be asymptomatic, and pathologic findings may be discovered incidentally on imaging or blood tests. In patients with spinal involvement, back pain represents the most common symptom (Chap. 41). The spinal level is determined by palpation, and pain intensity is assessed on a visual analog scale (VAS 1-10).

In some cases, excessive bone formation can lead to narrowing of the spinal canal. In the thoracic spine, this might lead to slowly progressive spinal cord compression and paraparesis. In the lumbar spine, spinal canal narrowing can lead to radicular pain and neurogenic claudication. Physical examination should include neurologic radicular sensory and motor testing (Videos 53.4 and 53.9). The sensory examination should investigate paresthesia, and a pinprick test should be performed. The motor examination requires muscle strength testing according to the Medical Research Council scale from 1 to 5. If spinal cord compression is suspected, pyramidal tract signs such as hyperreflexia and positive Babinski sign should be investigated.

Blood Tests. Alkaline phosphatase (ALP) is typically elevated at stages with increased bone metabolism. This biomarker correlates with the active bone resorption and formation by osteoclasts and osteoblasts. It is therefore used for follow-up in patients that are treated with bisphosphonates. Hepatic enzymes should be checked in parallel, since ALP might also increase in diseases of the liver. Osteocalcin is secreted solely by osteoblasts. This protein hormone has no diagnostic value in patients with Paget's disease.

53.4 Imaging

The standard radiographic examination consists of anteroposterior (AP) and lateral radiographs of the spine. Radiographic findings depend on the stage of the disease. During the initial phase, osteolysis can be present at the level of the vertebral body. During the course of the disease, periosteal and endosteal bone apposition will result in characteristic sclerotic lines parallel to the end plates (Fig. 53.1). Finally, cortical thickening will result in a framed picture and an enlargement of the vertebral body in the sagittal and coronal planes.

Computed tomography (CT) scan provides good-quality images of bony lesions. In the initial phase, osteoclastic activity will result in osteolytic images (Fig. 53.2). In the second phase, periosteal apposition and endosteal resorption can be present

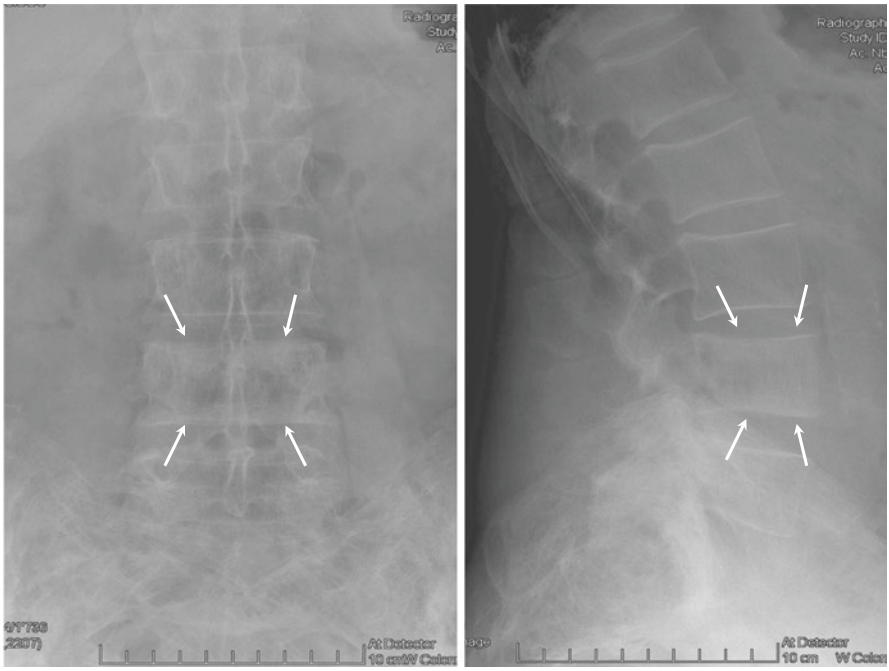

Fig. 53.1 Radiographs showing periosteal and endosteal bone apposition parallel to the end plates of L4

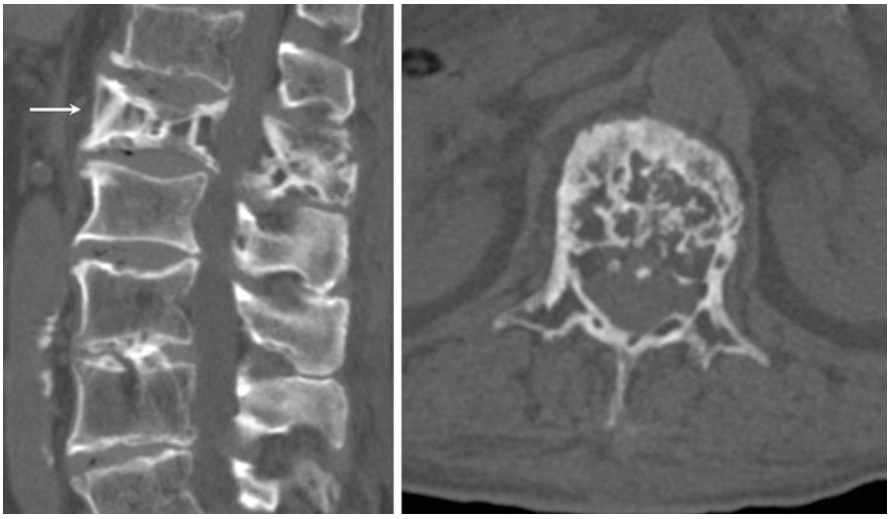

Fig. 53.2 Sagittal and axial CT demonstrating osteolytic images of the L2 vertebral body and posterior elements during the initial phase

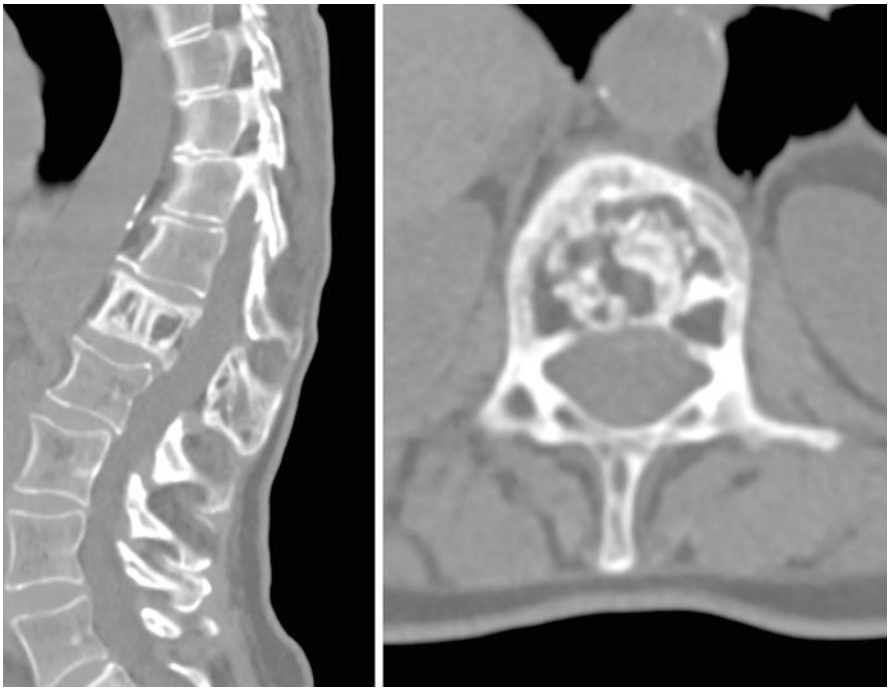

Fig. 53.3 Sagittal and axial CT images showing periosteal apposition and endosteal resorption at L1 during the second phase of Paget's disease

simultaneously (Fig. 53.3). In the third phase, sclerotic bone formation can be observed, also described as dense "ivory vertebra" (Fig. 53.4).

Magnetic resonance imaging (MRI) is indicated to evaluate the spinal cord or cauda equina if spinal stenosis is present. The coarsened aspect of the vertebra can be observed on T1-weighted images (Fig. 53.5), and T2-STIR sequences might help to identify an active spot, but CT is preferred as first-line imaging when evaluating osteolysis and bone formation. Bone scintigraphy using 99-technetium substrates and SPECT can aid the diagnosis of Paget's disease and demonstrate the distribution of different spots on the skeleton (Fig. 53.6).

53.5 Differential Diagnosis

Osteolytic images can be present in osteoporotic fractures and in vertebral metastases (breast, kidney, lung cancer) (Chap. 63), aggressive hemangioma (Chap. 55), giant cell tumor (Chap. 57), myeloma (Chap. 61), chordoma (Chap. 62), and chondrosarcoma. Differential diagnoses of dense sclerotic bone images include metastases (prostate cancer), osteosarcoma (Chap. 38), carcinoid, and Hodgkin's lymphoma.

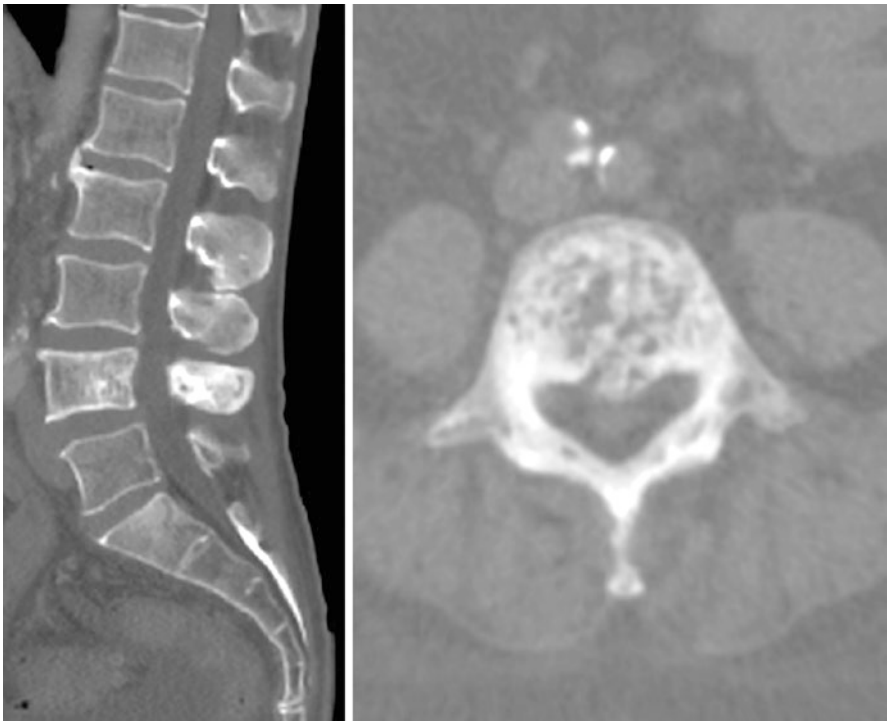

Fig. 53.4 Sagittal and axial CT images showing sclerotic bone formation at L4 during the third phase of Paget's disease

53.6 Treatment Options

The management of Paget's disease relies on normalization of bone remodeling and the treatment of pain. Analgesics, nonsteroidal anti-inflammatory drugs, and anti-neuropathic drugs are prescribed. Calcitonin was historically used in the treatment of bone metabolism. Today, different bisphosphonates (etidronate, clodronate, pamidronate, risedronate, zoledronate) were compared in clinical trials and represent the most efficient treatment. Surgical treatment is rarely indicated. Decompression and fusion might only be considered in neurologic complications and major osteolysis.

53.7 Expected Outcomes

Conservative treatment using bisphosphonates usually stabilizes bone remodeling and improves the quality of life for the patient. Clinical and radiologic follow-up is mandatory since complications might develop.

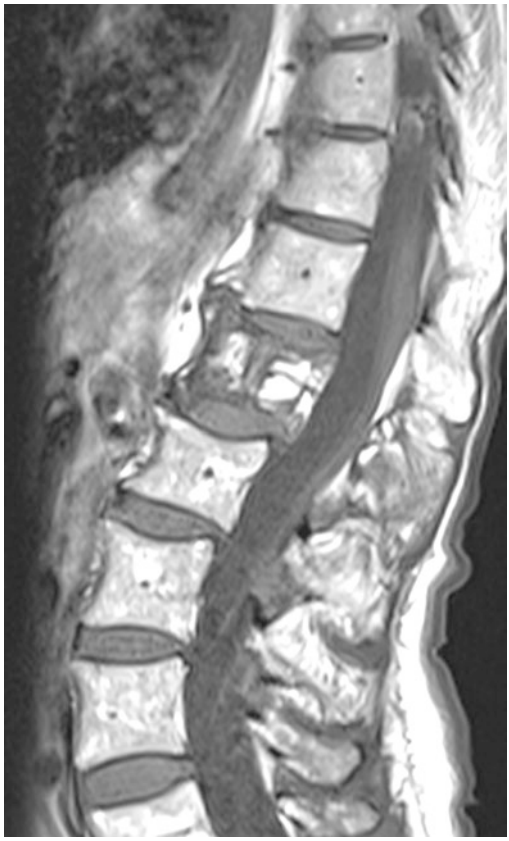

Fig. 53.5 T1-weighted sagittal MRI demonstrating the coarsened structure of the vertebral body at L1

53.8 Potential Complications

Bone is structurally weak in the osteolytic phase, and vertebral compression fractures can occur. Neurologic complications might appear in severe spinal canal stenosis due to excessive bone apposition. Neoplastic transformation is very rare in the spine (0.7%) and represents only 7% of all sarcomatous degeneration in Paget's disease.

53.9 What Should Patient and Family Know?

Paget's disease is mostly asymptomatic, but pain and neurologic symptoms can occur, usually after the age of 50 to 60 years. The entire skeleton needs to be initially checked using scintigraphy. Bisphosphonates represent the main antiresorptive treatment of bone metabolism. ALP dosage is required for monitoring bone metabolism.

Fig. 53.6 Bone scintigraphy demonstrating polyostotic Paget's disease with involvement of the spine (T11) and the pelvis

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Ankylosing Disorders of the Spine: AS and DISH

54

Yann Philippe Charles

54.1 Definition

Among ankylosing spinal disorders, ankylosing spondylitis (AS) and diffuse idiopathic skeletal hyperostosis (DISH) represent two distinct etiologies. Both entities will be presented in parallel in the following chapter, since certain aspects, such as stiffness and the risk for unstable spinal fractures, are similar for both diseases.

AS belongs to the group of axial spondyloarthropathies, which refers to a group of inflammatory rheumatic diseases. AS, also known as Bechterew's disease, is characterized by chronic inflammation of the joints and ligaments of the spine which leads to pain and stiffness. Vertebrae may fuse and result in a rigid spinal deformity such as cervical and thoracolumbar kyphosis. The prevalence of AS ranges between 0.1% and 1.4%. AS affects males two to three times more often than females. The onset occurs before the age of 45 years. A positive family history is often present. A gene known as the HLA-B27 is thought to be a risk factor.

DISH, initially described by Forestier and Rotés-Querol as senile ankylosing hyperostosis, represents a noninflammatory disorder that is characterized by ossification of ligaments and joint capsules. It typically occurs in the axial skeleton where progressive ossification of the anterior longitudinal ligament and bone formation between vertebral bodies results in spinal ankylosis. The prevalence of DISH ranges between 3% and 6% in the population over 40 years and increases with age. The sex ratio between males and females is 2:1.

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54.2 Natural History

Axial spondylarthropathies such as are associated with back pain and inflammation of sacroiliac joints. Additional clinical features of inflammatory rheumatic diseases such as skin psoriasis, heel enthesitis, dactylitis, uveitis, or colitis can be present. Over time, vertebrae may fuse and result in spinal deformity. Cervicothoracic kyphosis can lead to loss of horizontal gaze. Global thoracolumbar kyphosis is the most common deformity, which can induce anterior trunk malalignment. In severe cases, kyphosis increases pressure on the abdominal cavity and reduces lung capacity.

The exact pathogenesis of DISH remains unclear to date. An occasional familial incidence of DISH raised suspicion of genetic predisposition. Diabetes mellitus and obesity represent frequent comorbidities of DISH (23% to 40%). It is also associated with diminished glucose tolerance, gout, hypertriglyceridemia, and hyperretinolemia.

54.3 Physical Examination

Inflammatory pain and alternate buttock pain are typical in AS. Pain is usually present for more than 3 months. Morning stiffness, which improves with exercise, is common, whereas rest does not improve pain. Some patients report pain during nighttime. Decreased lumbar range of motion can be measured using the Schober method by drawing to points: one at the upper sacral level and another one 10 cm above. Normal range of motion of the lumbar spine is observed if the distance between both points increases to 15 cm in maximal anterior trunk flexion (Schober 10/15 cm). Any shorter distance indicates limited range of motion of the lumbar spine. The patient should further be examined for cervical and thoracolumbar sagittal plane deformities (Fig. 54.1). Pelvic retroversion and knee flexion indicate the compensation mechanism of anterior malalignment of the trunk.

Many patients with DISH have only mild clinical symptoms. They include stiffness in the cervical spine and eventually the lumbar spine. Ossifications might restrict range of motion in these mobile vertebral segments. Pain is reported by some patients, but its intensity is variable, and it is not constantly present. In severe cervical anterior ossification, dysphagia might be present.

Blood Tests. There are no specific lab tests to identify ankylosing spondylitis. Inflammatory markers such as C-reactive protein (CRP) can check inflammation, but CRP is nonspecific for sacroiliitis progression, and it is not always elevated. The HLA-B27 gene is associated with AS. However, it is also positive in 8% of the normal population.

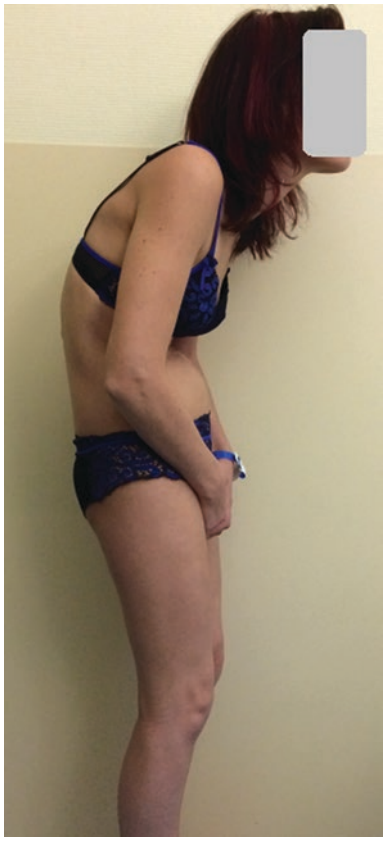

Fig. 54.1 The clinical aspect of a fixed thoracolumbar kyphosis in AS

54.3.1 Imaging

The standard radiographic examination consists of anterior-posterior (AP) and lateral radiographs of the spine (Video 54.6), the pelvis, and sacroiliac joints. During the initial phase of AS and DISH, anterior shiny corners of vertebral bodies can be observed. Syndesmophytes then turn out the fusion between vertebral bodies. Radiologic findings of DISH are typically found in the thoracic and cranial lumbar spine first (Fig. 54.2).

Computed tomography (CT) scan provides good-quality images of bony fusion. In AS, posterior facet joint fusion develops over time and results in a typical "bamboo spine." Bone mineral density can decrease within vertebral bodies in AS, resulting in a trabecular aspect of cancellous bone.

Magnetic resonance imaging (MRI) is indicated in AS to evaluate sacroiliac joints on T2-STIR sequences. A hypersignal indicates an active inflammatory process (Fig. 54.3).

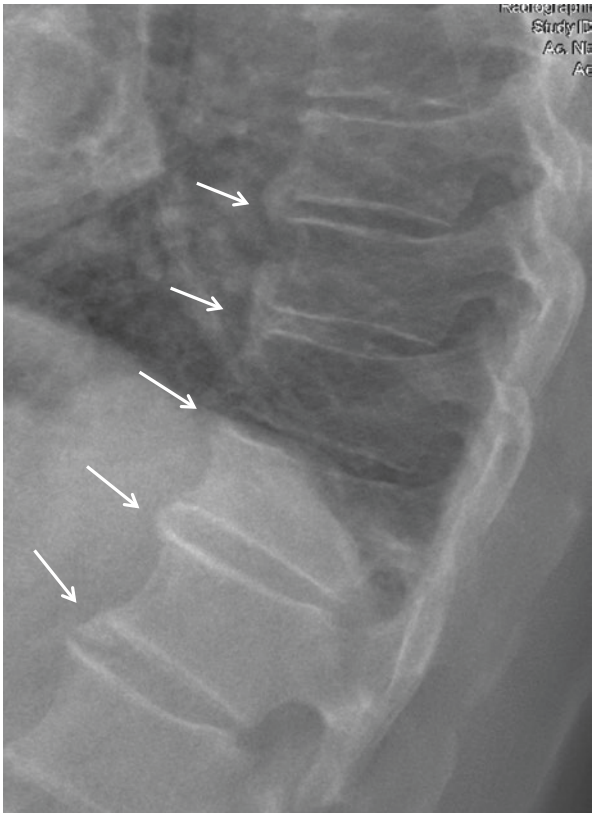

Fig. 54.2 The radiographic aspect of ossification of the anterior longitudinal ligament in the caudal thoracic and cranial lumbar spine in DISH

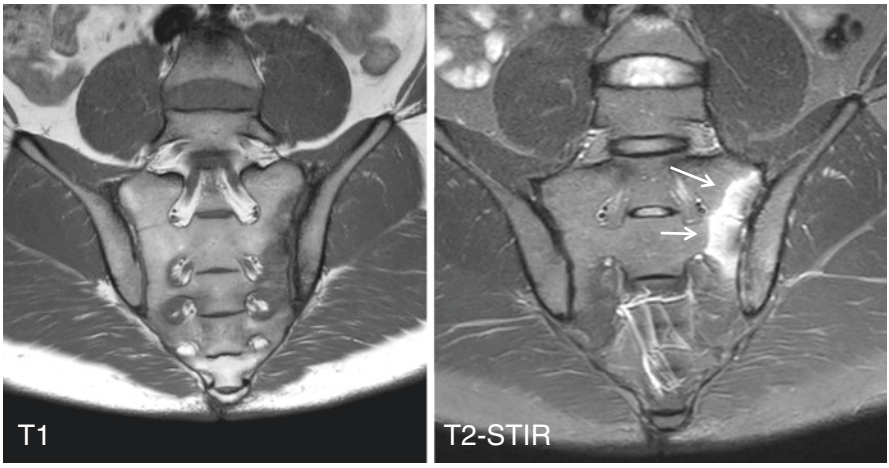

Fig. 54.3 MRI images showing an inflammatory sacroiliac joint on the T2-STIR sequence

54.4 Differential Diagnosis

Differential diagnoses of AS are nonspecific low back pain (Chap. 41), degenerative changes of the intervertebral disc, facet joints and sacroiliac joints, or inflammatory diseases such as rheumatoid arthritis.

A possible link between DISH and ossification of the posterior longitudinal ligament of the spine (OPLL) has been genetically investigated. However, it seems that both entities are different. DISH occurs more often in Caucasian patients, whereas OPLL is more frequent in Asian patients. Ossification of the posterior longitudinal ligament in OPLL typically leads to clinical signs of myelopathy or radiculopathy, which is rare in DISH (Chaps. 42 and 43).

54.5 Treatment Options

The management of AS relies on education, physical therapy, rehabilitation, and the treatment of pain using analgesics and nonsteroidal anti-inflammatory drugs. Tumor necrosis factor (TNF) blockers or an interleukin-17 (IL-17) inhibitor should be prescribed to reduce pain and stiffness. The following TNF blockers can be used: adalimumab, certolizumab pegol, etanercept, golimumab, or infliximab. Surgical treatment is indicated in severe kyphotic deformities. The indication for surgical correction of the fixed sagittal deformity is to improve global alignment and to restore a horizontal visual axis if necessary. It has been shown that deformity correction by single- or two-level pedicle subtraction osteotomy (PSO) might improve disability, pain derived from muscle fatigue, respiratory function, and quality of life (Fig. 54.4).

The treatment of DISH remains symptomatic using analgesics. Cervical osteophyte resection might be indicated in rare cases of dysphagia, and neurologic decompression might be considered in neurologic symptoms.

54.6 Expected Outcomes

Conservative treatment of AS using TNF blockers in combination with regular exercise improves the inflammatory process and the course of the disease. Surgical treatment should be carefully planned in severe deformity only, but it can improve the quality of life in severe rigid/fixed deformities.

54.7 Potential Complications

Ankylosed spines in AS and DISH are fragile, and low-energy trauma can cause unstable vertebral fractures. The cervical spine between C5 and C7 accounts for 75% of traumatic spinal injuries and carries a high risk of neurologic complications such as tetraparesis or tetraplegia (Fig. 54.5). Non-displaced thoracolumbar

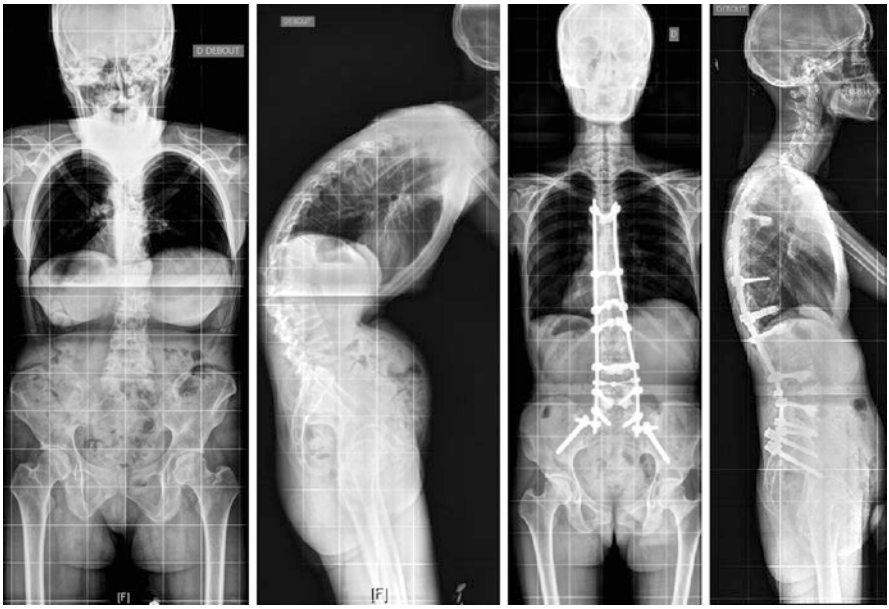

Fig. 54.4 Pre- and postoperative radiographs of fixed sagittal thoracolumbar kyphosis correction in AS by two-level pedicle subtraction osteotomy at L2 and L4

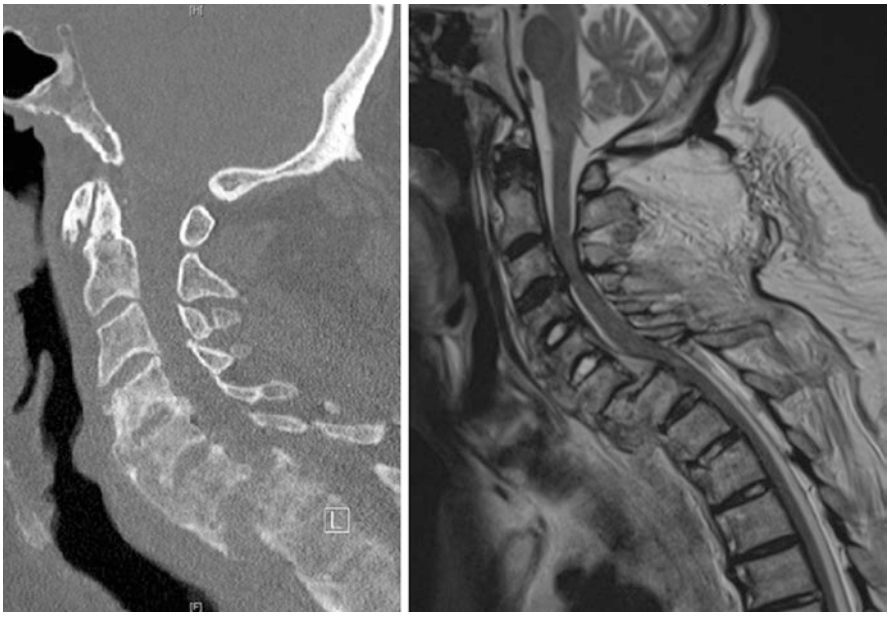

Fig. 54.5 CT and MRI of a displaced C6-C7 fracture in DISH showing spinal cord compromise

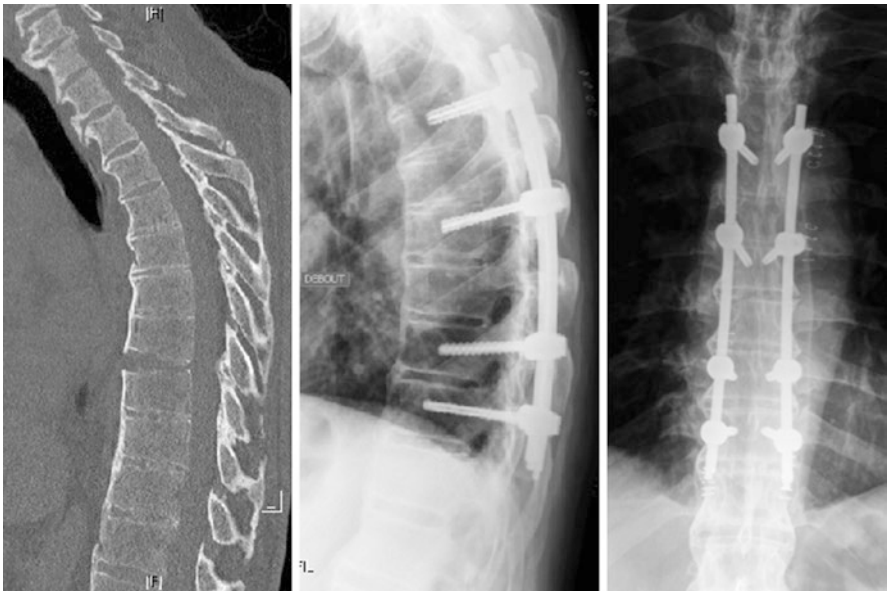

Fig. 54.6 Preoperative CT and postoperative radiographs of a patient with unstable thoracic fracture treated by minimal invasive percutaneous instrumentation

fractures caused by low-energy trauma may be challenging to diagnose. There is a high risk of secondary displacement and neurologic complications. Therefore, surgical treatment using percutaneous instrumentation should be preferred (Fig. 54.6) over conservative brace treatment.

54.8 What Should Patient and Family Know?

AS is an inflammatory disease that belongs to the group of axial spondyloarthropathies, which occurs in younger patients and requires lifelong treatment. DISH is a noninflammatory ankylosing disorder of the axial skeleton of the elderly patient, who often remains asymptomatic. Both entities lead to stiff ossified spines that are at high risk of secondary displacement and neurologic complications in the event of trauma.

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Vertebral Hemangioma

55

Burak Karaaslan and Alp Özgün Börcek

55.1 Definition

Pathologically, hemangiomas are benign, unencapsulated proliferation of vascular channels. Pathologically, there are three types, capillary, cavernous, and mixed. They can occur anywhere, but the most common locations are the skin of the face, scalp, chest, or back. Vertebral hemangiomas have an incidence of 11% on autopsy series, and most of them are asymptomatic and discovered incidentally. İncidence increases with age, and about 30% of patients have multiple lesions. There is a female predominance (3:1).

55.2 Natural History

Vertebral hemangiomas (VH) may be either progressive or silent. Most of the asymptomatic patients remain so for several years. In case of a painful lesion, the amount of involvement of the vertebra and the location should be considered. Lesions confined to the vertebral body of a lumbar or cervical region are not expected to progress; in those patients, other pain sources should be evaluated (Chap. 41). On the contrary, a thoracic lesion extending outside the vertebra body with soft tissue expansion should be considered as a potentially progressive lesion. Especially in younger patients, even if the lesion is discovered incidentally, the patient should be followed carefully.

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55.3 Physical Examination

There is no physical examination finding for pure VH. Only 1% to 1.2% of all hemangiomas are symptomatic, pain being the most common symptom. Young adults present with cord compression or radiculopathy symptoms more commonly. Neurological findings change related to the level of the lesion and presence of expansile character or soft tissue involvement (Video 55.4). Deficits can occur due to either direct compression of the spinal cord or a change in blood flow dynamics. Additionally, fractures, spinal epidural hematoma, or direct vascular compression may cause spinal deficits. The clinical picture is usually progressive; however, there are sudden changes that can also be observed. Of note, pregnancy—probably due to both increased venous tonus and hormonal activity—may aggravate symptoms. Additionally, there are reports of the fluctuating clinical pictures in premenopausal women.

55.4 Imaging

HVs are usually found incidentally. In rare cases, they may show findings of pathological fractures or deformative changes. On plain radiographs, the presence of coarse vertical striations—"honeycomb" appearance within the vertebra body—is the main finding. On T1- and T2-weighted magnetic resonance imaging (MRI) pictures, they have increased signal intensity. Soft tissue parts have usually lower signals. HV enhances intensely with contrast medium. More than 70% of the lesions are found in the thoracic spine, and they can involve every part of the vertebra including the posterior arcs, and they can also have soft tissue components with expansile behavior. In up to 65% of cases, all parts of the vertebra are involved. The presence of adipose tissue in hemangiomas may be a sign of a benign course. For cases that are not diagnosed easily with MRI, angiography may show dense opacification of the vertebral body (Fig. 55.1).

According to their clinical and radiological features, they can be divided into three groups:

- Type A: lesion presenting with cord compression due to the extraosseous extension of the tumor.
- Type B: lesion presenting with local pain without extraosseous extension.
- Type C: most common type, asymptomatic. However, in young patients, those lesions should be followed up.

55.5 Differential Diagnosis

Symptomatic VH is a potential cause of back pain (Chaps. 11 and 41). In cases with incompatible radiographic findings, other causes of back pain should be ruled out. Paget disease (Chap. 63), metastasis mainly from osteosarcoma (Chap. 38), prostate

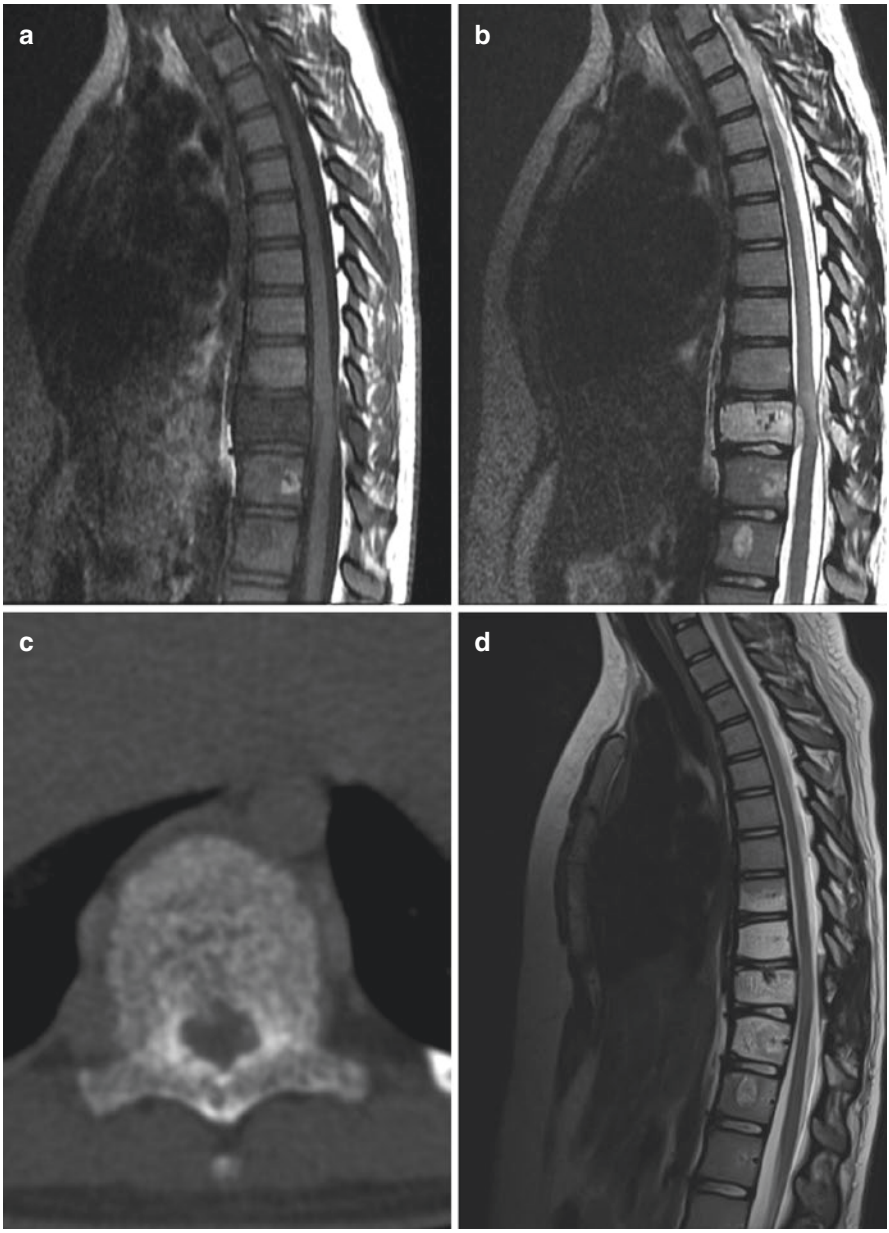

Fig. 55.1 MRI and computed tomography pictures of a 9-year-old patient with symptomatic thoracic vertebral hemangioma managed with radiotherapy. (**a**) T1-weighted image of the vertebral hemangioma. (**b**) T2-weighted image of the lesion with significant anterior and posterior cord compression. (**c**) Axial CT image of the lesion. (**d**) Post-radiotherapy image 3 months after the treatment showing complete resolution of the extraosseous lesions and the cord compression

carcinoma, and medullary thyroid carcinoma should be considered in the differential diagnosis (sclerotic lesions; Chap. 63) (Video 55.8). Additionally, hematological malignancies such as lymphoma and myeloma should also be considered.

55.6 Treatment Options

As in every pathology, treatment should be tailored to the patient. Asymptomatic patients can be followed up with no intervention. In case of neurological deficits or significant pain, there are various treatment options. Radiotherapy, endovascular embolization, percutaneous polymethyl methacrylate (PMMA – vertebroplasty), and ethanol injection are the main management options. In case of significant cord compression and acute findings, surgical resection of the lesion and spinal stabilization can be considered although significant blood loss can complicate the surgical procedure. In symptomatic cases, embolization techniques and surgical decompression may be combined according to the patients' needs. Of note, young patients with asymptomatic lesions should be followed clinically and/or radiologically. Treatment options should be considered according to the extension of the involvement, clinical picture, and age of the patient.

55.7 Potential Complications

Acute cord compression, pathological spinal fractures, and related deformity are potential complications. In case of surgical intervention, surgeons should be prepared for intraoperative bleeding that can cause significant morbidity and in some cases mortality.

55.8 What Should Patient and Family Know?

VHs are usually benign and indolent lesions; successful treatment with the current medical intervention techniques is possible. Only a small fraction of VH poses clinical difficulties, and successful treatment with the current medical intervention techniques is possible.

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Aneurysmal Bone Cyst

56

Murat Songür and Alpaslan Şenköylü

56.1 Definition

Aneurysmal bone cyst of the spine (ABC) was formerly accepted as a pseudotumoral lesion of unknown origin; it is now considered to be a benign bone tumor. ABC is characterized as multiple cysts containing blood and separated by a fibrous septae. ABC is a rare tumor consisting of about 1% of primary bone tumors. Although being a naturally benign lesion, some ABCs may have aggressive behavior. Success of index treatment is extremely important, because recurrence frequently results in a high level of structural problems and neurological deficit.

56.2 Natural History

ABC has a benign nature without the risk of sarcomatous transformation. The cervical spine and sacrum (Fig. 56.1) are the most frequent locations of ABCs. Pathological fracture, neurological involvement, and instability are rare although possible.

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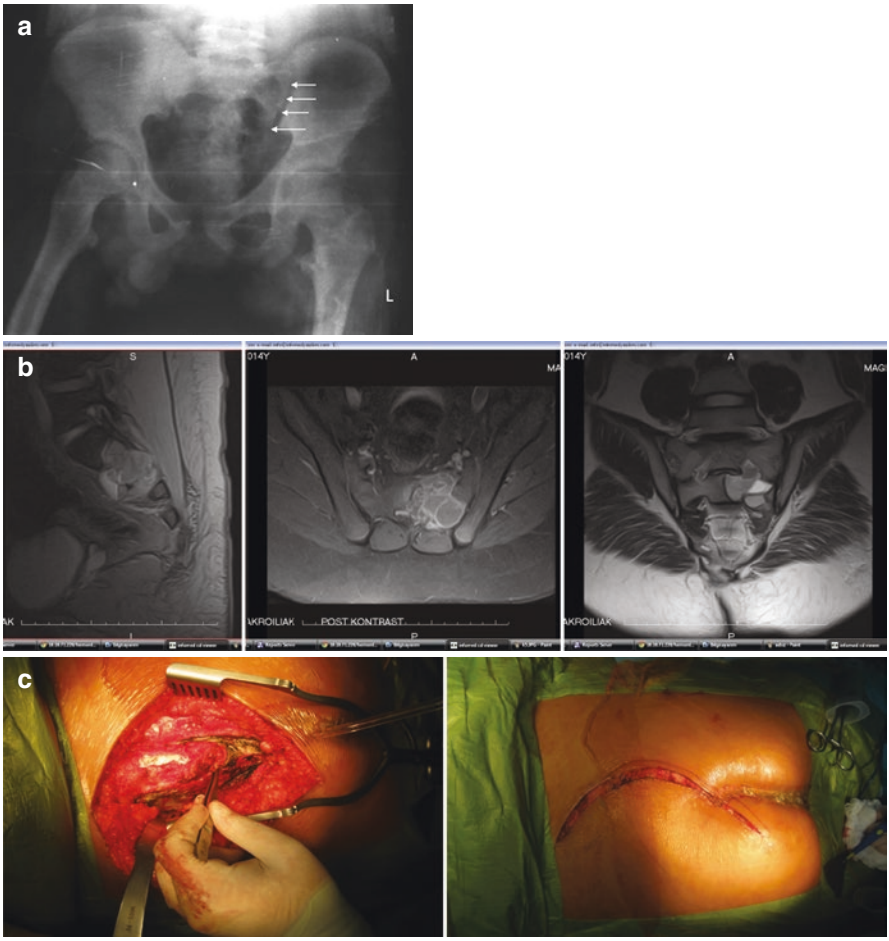

Fig. 56.1 ABC of left S1–S2 (**a**) in a 14-year-old boy. T2-weighted and post-contrast MRI images (**b**) reveal multiple cysts with fluid-fluid image at left S1 and S2 growing ventrally and dorsally. Partial sacrectomy with sacral canal decompression followed by autogenous bone grafting was performed (**c**)

56.3 Physical Examination

ABCs usually involve children and young adults. Patients present with localized pain and swelling, with or without tenderness (Chap. 11). Spine motion can be restricted at the involved region. Neurological impairment is rare (Videos 56.4 and 56.9). Signs and symptoms of myelopathy or single root involvement should be evaluated carefully in cervical lesions, since these findings may be overlooked in healthy teens.

56.4 Imaging

Plain radiographs reveal cystic-lytic soap bubble-like lesions with smooth non-sclerotic margins. Computerized tomography (CT) shows similar imaging. T2-weighted magnetic resonance imaging scan typically demonstrates multiple separated cysts with vertical fluid-fluid levels. Cervical lesions tend to involve posterior elements but may progress anteriorly interfering vertebral artery and involve epidural space. Therefore, axial scans help involvement of vertebral artery and epidural space and also facilitate staging and decision-making.

56.5 Differential Diagnosis

Although ABCs have unique radiological features, tissue sampling is mandatory, and it can be facilitated as CT-guided or open biopsy (Video 56.8). Telangiectatic osteosarcoma may present with a similar radiographic pattern although it usually progresses with completely different clinical course and outcome.

56.6 Treatment Options

Several treatment options are available for ABCs. Successful outcome is directly related to the success of the index procedure. Therefore, selection of appropriate initial treatment is of paramount importance. All treatment modalities have inherent advantages and disadvantages. En bloc resection of the lesion may provide safe margins with low risk of recurrence; the increased surgical morbidity is usually accepted by many authors as "overtreatment." Intralesional curettage (with or without adjuvant) has about a 30% risk of recurrence, and it usually necessitates preoperative embolization in order to avoid intraoperative hemorrhage. Selective arterial embolization (SAE) can be considered as a first-line treatment for lesions not involving neural structures and without risk of pathological fracture. Preoperative angiography is required to prevent sclerosing agent leakage into the spinal canal or nervous system. Intra-cystic injections with acrylic sclerosing agents or biologics are other treatment options. Especially bone marrow-derived mesenchymal stem cell (MSC) concentrate injection under fluoroscopy control gives promising results although it is generally avoided in children because of the risk of late malignancy. Radiation therapy (RT) is another treatment option with variable outcomes.

Since most of the literature on spine ABCs is represented by case series involving different treatment protocols, such studies outline favorable outcomes that are not related to the type of treatment. Therefore, the least morbid type of treatment should be performed.

Eventually, decision-making involves size, location, stability, neural involvement, and surgeon's and interventional radiologist's preference. Each case should be evaluated separately, and treatment options should be tailored individually.

SAE is a common method for most lesions, especially extending a wide portion of spine at both mobile segments and sacrum. If equipment and interventional radiologist are available, this treatment approach may be accepted as the first-line treatment option in lesions carrying a high recurrence rate and requiring morbid surgical approach. Angiography is indicated for almost all ABCs of the spine requiring treatment. Principles of embolization for ABCs may be summarized as microcatheterization of feeding vessel and injection of sclerosing agent to not only feeding level but also segmental arteries bilaterally two levels above and below the lesion (for thoracic and lumbar lesions). If the catheterized segmental artery is associated with anterior spinal artery, embolization is aborted at that level, since this approach will eventually result in cord ischemia. Selective arterial coil embolization is a method of choice for cervical lesions. Unlike sclerosing agent injection embolization performed at thoracic and lumbar levels, this procedure is performed unilaterally. Vertebral artery embolization from C7 to posterior inferior cerebellar artery is common practice with good results. As the patient is awake during the procedure, temporary obstruction of vertebral artery is performed to evaluate the patency of the contralateral vertebral artery. Patients are followed for progression-regression of the disease, ossification of the lesion, or revascularization of the lesion by CT scan and/or angiography. SEA can be repeated until signs of healing are achieved. Despite efforts, an increase in tumor size, deterioration of neurological status, and development of instability may indicate surgery. Another advantage of SEA is decreased blood loss during surgery.

Intralesional bone marrow-derived MSC aspirate is another minimally invasive procedure (Video 56.7), usually reserved for lesions not suitable for SAE. Percutaneous puncture of the cysts and MSC aspirate injection theoretically can decrease the intra-cystic pressure and facilitate osteoblastic regeneration. Multiple injections may be required to achieve healing. MSC concentrates can be mixed with demineralized bone matrix and other scaffolds to enhance osteo-inductivity within the cystic cavity.

The acute neurological deficit, responsiveness to minimally invasive procedure, and instability are the common surgical indications for ABCs. Two types of surgical treatment are available: (1) wide resection and (2) curettage and bone grafting. Wide resection is usually performed in posterior element-only cases, and it causes minimal surgical morbidity after resection. However, post-laminectomy kyphosis is an important complication of posterior element wide resection in the growing spine, reported between 37 and 95% of cases; in such cases, anterior and posterior surgery is recommended. Another indication of surgical resection is sacral lesions. Since achievement of complete embolization is extremely difficult in sacral lesions, surgery is the only viable method (Fig. 56.1). Intralesional curettage has up to 30% recurrence rates; therefore, it is not preferred in spinal lesions.

RT promises successful result in terms of tumor control, but radiotherapy-induced sarcoma, radiation myelopathy, and other side effects (especially in children) limit its use. RT should be reserved when other treatment options fail.

Therefore, multimodal approach is necessary to successfully manage ABCs of the spine, with collaboration between interventional radiology and spine surgery.

Future studies will focus on osteoclastic inhibition via RANKL inhibitors for ABCs. Promising reports are available but not enough for routine use.

56.7 Expected Outcomes

Favorable results are reported with minimally invasive procedures such as selective angiographic embolization and intralesional injections. Usually necessitating multiple applications, healing can be expected within 12 to 18 months. Surgery, even if indicated, has a high rate of recurrence; wide resection may have unfavorable effect on growing spine.

56.8 Potential Complications

Minimally invasive procedures seem to be resulted minimal complications. Open surgery may have a high rate of complications such as kyphosis and recurrence, unless it is performed by an experienced surgical team.

56.9 What Should Patient and Family Know?

Minimally invasive approaches are evolving, and a favorable outcome is possible. Multiple interventions may be required to achieve complete healing.

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Giant Cell Tumor

57

Peter Pal Varga and Aron Lazary

57.1 Definition

Giant cell tumor (GCT) is a benign bone tumor with locally aggressive behavior. High risk of local recurrence (LR) and low risk for distant (lung) metastasis characterize the disease. Histologically, GCT is composed of multinucleated, osteoclast-like cells, mononuclear cells, and stromal spindle-shaped cells. These latest are the neoplastic components of the tumor.

According to the Enneking classification, GCT is S2 (active benign) or S3 (aggressive benign) lesion. In S2 tumors, the Enneking appropriate treatment is intralesional excision, while, in S3 cases, only en bloc resection can provide long-term local control. Both cases can be challenging in the spine because of the proximity of neural elements, the rich blood supply of GCT, and the consequent spinal instability. The possible complexity and morbidity of the surgical treatment and the high rate of LR resulted in the development of adjuvant therapeutic options like local adjuvants (phenol, hydrogen peroxide, cryosurgery, or PMMA) and systemic drugs (denosumab). Postoperative radiation therapy can be also used for local control; however, it can increase the transformation of GCT to radiation-induced sarcoma.

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57.2 Natural History

GCT is most common in young adults, and females are more frequently affected. GCT is usually localized in the epiphyseal area of long bones; its spinal appearance is rare although the aggressive nature of the tumor can cause spinal instability and/or neurological deficit. The most common symptom of a spinal tumor is pain which can be nonmechanical tumorous pain, mechanical pain in case of spinal neoplastic instability, or pain related to neurological compression (Chap. 41). Functional deterioration can be related to the pain of neurological deficits caused by the tumor mass or pathological fracture. Alarming symptoms are intractable pain, progressing neurological deficit, cauda equina syndrome, or signs of spinal cord compression (Videos 57.4 and 57.9).

57.3 Physical Examination

Standard spinal physical examination of the patient is crucial to identify conditions requiring emergency surgery; however, it is rare in primary spinal tumors. Laboratory findings are nonspecific.

57.4 Imaging

GCT is an aggressive, lytic lesion. Results of osteolysis can be visible on radiographs and computed tomography (CT) studies, while soft tissue expansion is diagnosed on magnetic resonance imaging (MRI) (Fig. 57.1). In spinal GCT, MRI is crucial to describe the relationship of the tumor to the spinal canal, nerve roots, and spinal cord. Contrast-enhanced imaging studies are also important to explore the main vessels and sometimes the feeding artery. Chest CT is indicated to exclude lung metastases, especially in an advanced stage of the disease. Regular imaging studies (MRI, CT) are advised during the follow-up postoperatively to assess local control. Spinal GCT with extraosseous involvement can be categorized as an aggressive tumor.

57.5 Differential Diagnosis

Tumorous lesions and tumor-like lesions of the spine are the most common differential diagnostic issues. Following the first and most important oncological principle, namely, “tissue is the issue,” the cornerstone of the differential diagnosis is the histological examination of the lesion. In case of a primary spinal tumor, imaging-guided percutaneous biopsy or open biopsy can provide adequate tissue for the detailed histopathological studies (Video 57.8).

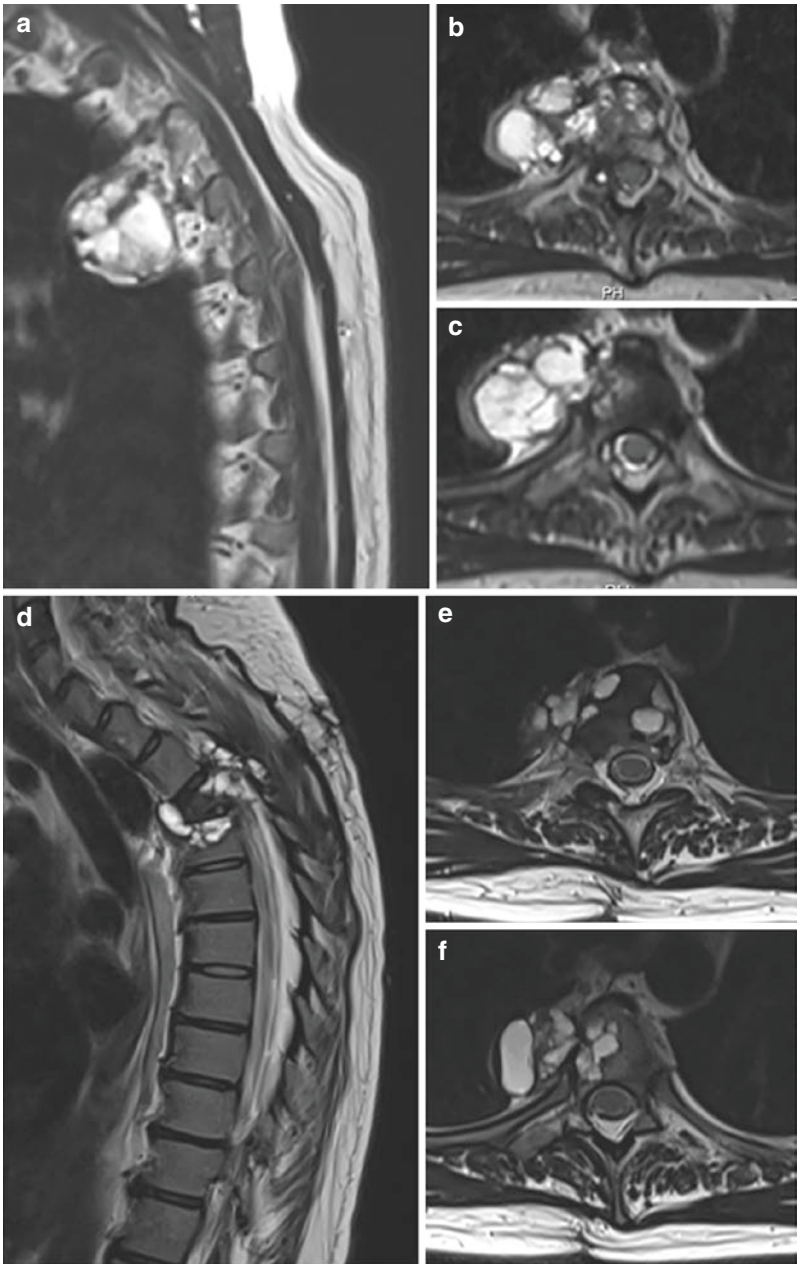

Fig. 57.1 Thoracic GCT treated with denosumab. The 38-year-old female patient had back pain for months. Thoracic spine MR showed a large soft tissue mass and lytic tumor at ThIII (**a**–**c**). Biopsy proved the GCT diagnosis. Denosumab treatment was initiated with excellent clinical result and local regression of the mass; however, a progressive kyphotic deformity developed 8 months after the diagnosis (**d**–**f**)

57.6 Treatment Options

57.6.1 Surgery

Enneking appropriate surgical treatment (intralesional resection in S2 and en bloc resection in S3 GCTs) provides significantly better oncological outcome compared to Enneking inappropriate treatment where LR rate is higher. Management of LR can be difficult, and LR is associated with shorter survival. The en bloc resection of aggressive spinal/sacral GCTs can be challenging and result in significant neurological loss of function. Not only tumor resection but the management of spinal instability and soft tissue reconstruction are important issues of preoperative planning (Fig. 57.2).

57.6.2 Embolization

GCTs are vascularized tumors; high blood loss is one of the most common complications in intralesional surgeries. That is why preoperative embolization is strongly

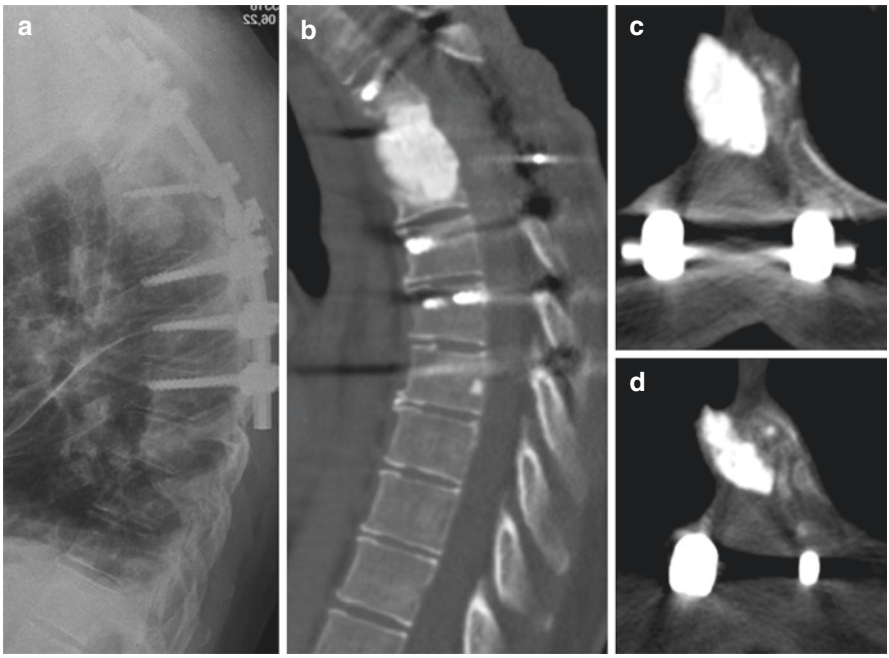

Fig. 57.2 Surgical resection, reconstruction, and adjuvant denosumab treatment. The case above with stable disease suffered from severe back pain because of the increasing kyphosis and pathological fracture (Fig. 57.1d–f). An intralesional surgical resection after embolization was performed followed by anterior column reconstruction with PMMA and posterior stabilization (**a**). Denosumab has been followed providing good local control 2 years after the surgery (**b–d**)

advised. On the other hand, embolization itself can cause the shrinking/sclerotization of the tumor mass, so in spinal cases, where there is no instability or urgent neurological symptom, embolization is the first line of management. The effect of the embolization should be follow-up by CT imaging studies, and the procedure can be repeated several times. In case of a surgical treatment, preoperative embolization (the day before the surgery) is strongly advised to reduce intraoperative blood loss.

57.6.3 Denosumab

Denosumab is a human monoclonal antibody against osteoclast activator RANKL. The inhibition of osteoclast activation via RANKL inhibition prevents bone resorption. Denosumab is primarily used in the treatment of osteoporosis, but a growing body of evidence shows its effect and safety in the management of GCT. Calcium levels should be monitored and calcium/vitamin D supplementation is advised during denosumab treatment. A dental checkup is also essential to screen for osteonecrosis of the jaw. Stop of tumor growth and the sclerotization of the lytic GCT mass can be seen on follow-up images as results of the treatment. "Stand-alone," pre- and postoperative administration of denosumab can be supported in spinal GCT after the individual evaluation of the situation, but the length of the treatment is still an open question.

57.7 Expected Outcomes

The LR rate is about 20% to 30% in spinal GCT, especially in case of intralesional resections. Functional outcome is strongly associated with biomechanical stability and neurological function which can be compromised by surgical resection. Even in huge tumors, denosumab treatment can stop the progression and improve function.

57.8 Potential Complications

Intralesional surgeries especially in sacral locations result in fatal intraoperative blood loss. Spinal cord injury is a serious complication in thoracic/cervical GCT resections. Denosumab treatment can have significant side effects such as hypocalcemia or jaw osteonecrosis.

57.9 What Should Patient and Family Know?

Patients and their families should understand that spinal GCT can be a life-threatening condition especially if the management is not individually planned and flawed by reckless steps. The surgical treatment can result in massive blood loss and serious complications, while inadequate surgery leads to LR, increasing morbidity,

and mortality. On the other hand, well-timed adjuvant therapies especially embolization and denosumab treatment increase the success of the management.

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Spinal Meningioma

58

Marcel Ivanov

58.1 Definition

The spinal meningiomas are usually well-defined, benign, and slow-growing tumors with dural attachment. If untreated, they may cause significant morbidity.

Meningiomas constitute approximately 30% of intradural extramedullary spinal tumors. Their incidence is approximately 3:100,000 and they are more common in women than in men (3:1).

The meningiomas are arising from the layers around the spinal cord (arachnoidal cap cells).

Radiation and NF are recognized risk factors of meningiomas.

The meningiomas are located predominantly in the thoracic area (80%). The rest are distributed in the cervical segment (15%) and less frequently in the lumbosacral area (5%).

58.2 Natural History

Meningiomas are usually slow-growing tumors. The tumors tend to be diagnosed between the fifth and the seventh decade of life. Diagnosis of meningioma at an earlier age suggests more aggressive tumor behavior which has an incidence of <2% [1].

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Meningiomas may reach considerable size with severe radiological spinal cord compression before becoming symptomatic.

Once the compression on the spinal cord and/or nerves becomes significant, the patients may present various degrees of sensory and motor dysfunction with progressive deterioration of balance and coordination, occasional falls. Occasionally, the patients may describe vaguely localized pain in the spinal area, which is more pronounced at night (Chap. 41).

In advanced stages, if untreated, the mobility may be lost, and the patients may develop sphincter dysfunction.

58.3 Physical Examination

The clinical examination should look for signs of spinal cord/nerve compression. It is essential to perform careful neurological examination with assessment of gait, muscle power, tonus, reflexes, proprioception, and sensation (Video 58.4).

Positive long tract signs with positive Romberg probe, brisk reflexes, abnormal proprioception, and clonus should be a strong indication for magnetic resonance imaging (MRI) scan of the spine.

58.4 Imaging

MRI scan is the method of choice for the diagnosis of spinal meningiomas. On the MRI scan, the meningiomas appear as well-defined intraspinal lesions isointense with the spinal cord on T1- and T2-weighted images. They homogeneously enhance contrast on T1WI with gadolinium.

The presence of dural tail with a wider dural base helps to differentiate meningiomas from the nerve sheath tumors [2] (Fig. 58.1a). The MRI scan will confirm the level and degree of spinal cord compression.

Computed tomography (CT) can help to assess tumor calcification (Fig. 58.1b). Myelo-CT can be an imaging alternative in cases when MRI is contraindicated.

58.5 Differential Diagnosis

Biopsy is important; the following conditions should be considered (Video 58.8):

- Nerve sheath tumors.
- Myxopapillary ependymoma.
- Dermoid/epidermoid tumors.
- Calcified thoracic disc herniation.
- Myelopathy of another cause.

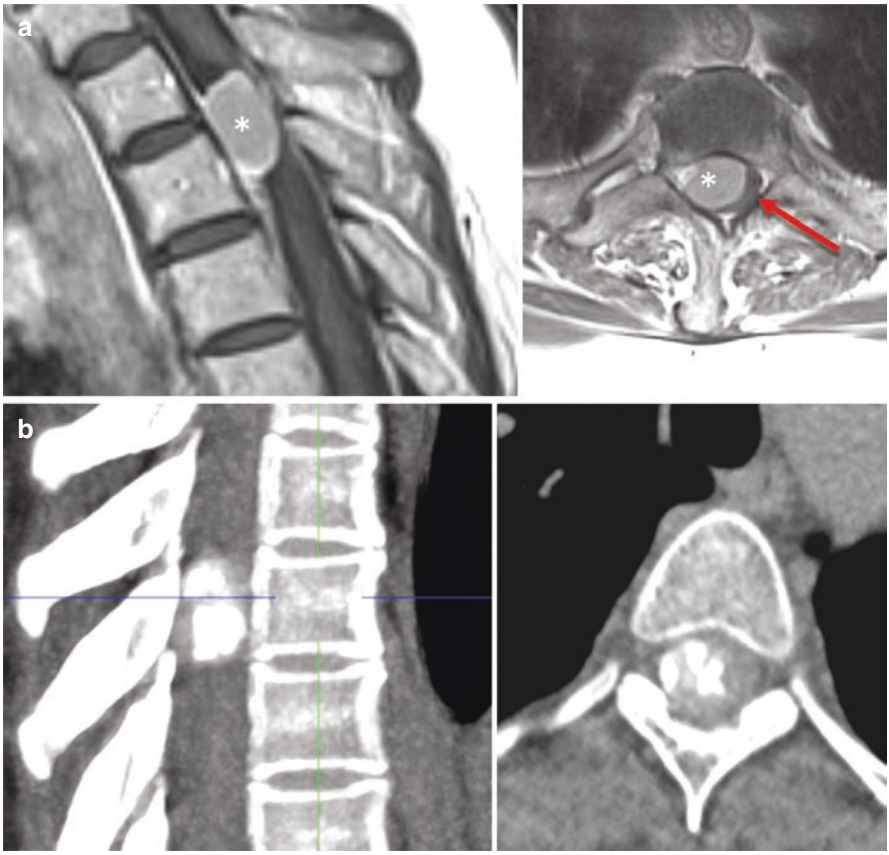

Fig. 58.1 (**a**) T1WI MRI with contrast. Spinal meningioma (*) located anterolateral to the spinal cord (arrow). Tumor has a visible dural tail. (**b**) CT thoracic spine demonstrated a highly calcified intradural tumor. This can be particularly challenging if located anterior to the spinal cord

58.6 Treatment Options

Surgery remains the mainstay of treatment of meningiomas and can be curative in the vast majority of patients.

The surgery has two main goals:

1. Spinal cord decompression.
2. Prevention of tumor recurrence.

The patient is placed prone. Even if the tumor is located anterior to the spinal cord, it still can be resected via the standard posterior approach. Either en bloc laminotomy using a craniotom (with the goal of its re-attachment after tumour is removed - laminoplasty) or standard laminectomy is performed (Laminoplasty as

described by Raimondi has the benefit of lower risk of postoperative kyphosis comparing to laminectomy [3]. Laminotomy is the surgical removal and subsequent reinsertion of laminae and spinous process; the term laminotomy is often incorrectly used to describe a partial laminectomy. A laminectomy is defined as complete removal of the lamina.

Surgery of the tumors located anterior or anterolateral to the spinal cord (40%) is usually more challenging due to narrow corridor and the potential need for gentle manipulation of the spinal cord. This could be particularly challenging when the tumor is calcified (Fig. 58.1b). Intraoperative neurophysiological monitoring is helpful in cases when the spinal cord needs to be mobilized.

We recommend dural exposure of at least 5 mm cranial and caudal to the tumor poles. This can be confirmed with intraoperative ultrasound [4, 5] (Fig. 58.2).

Under microscopic magnification, meticulous hemostasis is performed before opening the dura. After midline dural opening, the dural edges are stitched laterally. At this stage, the tumor should be visible (Fig. 58.3).

In most cases, the spinal meningiomas are fleshy, red-purple, sometimes fibrous, and occasionally calcified tumors. There is normally a thin arachnoid layer between the tumor and the neural structures, which prevents adherence to the spinal cord and allows tumor mobilization and separation. Dissection of the tumor base with coagulation of the vessels helps to reduce the intraoperative bleeding.

By debulking the tumor (either with an ultrasonic aspirator or piecemeal removal), the pressure on the spinal cord is decreased (Fig. 58.3). It allows mobilization of the tumor capsule away from the spinal cord. If the tumor is located anteriorly, the dentate ligament can be divided, and the spinal cord can be gently rotated laterally with the guidance of intraoperative neurophysiological monitoring.

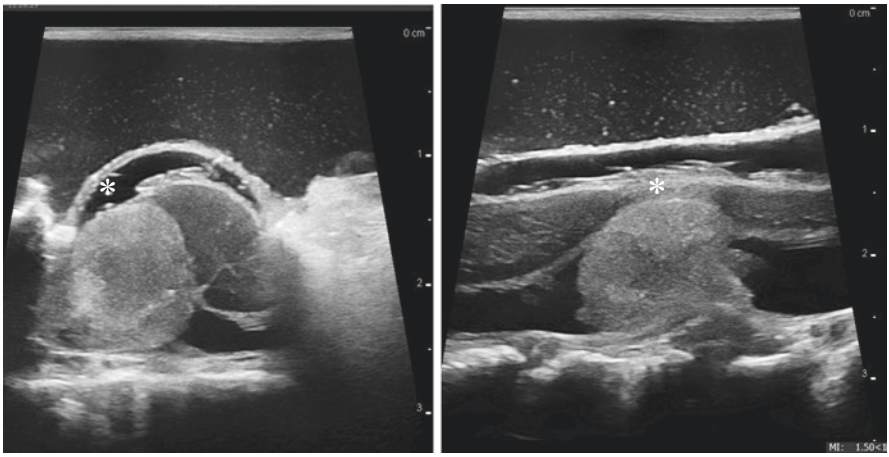

Fig. 58.2 Intraoperative ultrasound can be helpful to confirm tumor location and adequate dural exposure before opening the dura. The ultrasound demonstrates tumor (*) located anterolateral to the spinal cord

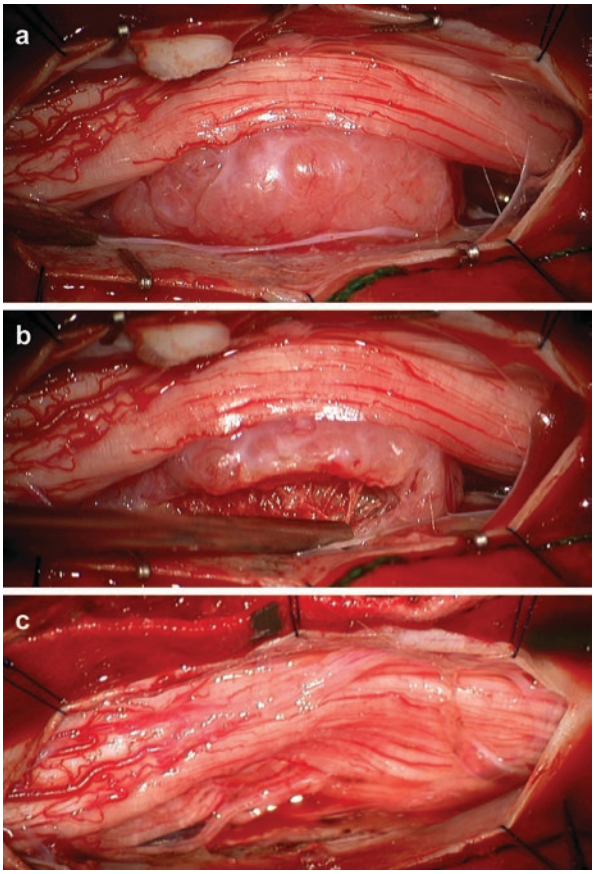

Fig. 58.3 Intraoperative image – spinal meningioma located anterolateral to the spinal cord. Surgical steps: view after (**a**) dural opening, (**b**) tumor debulking, (**c**) tumor excised, and spinal cord decompressed

After the tumor is removed and hemostasis achieved, the dura is inspected for any residual which should be excised. The dural base is coagulated (Fig. 58.3).

Dura is closed in a watertight fashion with non-resorbable stitches. When laminotomy is performed, posterior instrumented fusion is recommended; on the other hand, in case of laminoplasty, the "posterior shutter" is placed back and fixed with stitches, and the patient is immobilized (cast) until fusion is achieved.

58.7 Expected Outcomes

Surgery for excision of spinal meningioma is usually a gratifying procedure with generally good outcomes. Once the tumor is removed, the progressive neurological deficit is halted. Patients that had incomplete neurology before surgery will usually notice some improvement or even return to normal. Most of the improvement will happen within the first 2 to 3 months from surgery, and usually by 12 months it will reach a plateau.

The risk of recurrence is generally low. However, it is possible, even after many years; therefore, surveillance MRI is recommended. The proliferation index Ki-67 and the arachnoid invasion are the risk factors for recurrence of spinal meningiomas [6].

The postoperative outcome may be negatively influenced by the anterior location of the tumor with regard to the spinal cord, increased size, transdural expansion, degree of spinal cord compression, calcification of the tumor, poor preoperative neurological status, and aggressive histological status.

58.8 Potential Complications

Untreated patients can develop progressive neurological deficit and potentially loss of neurological function below the level of compression.

Surgical risks can be divided in:

- Intraoperative and early postoperative:
  - Spinal cord or nerve injury.
  - Bleeding.
  - Infection—superficial or deep wound infection and meningitis.
  - CSF leak.
  - General/anesthetic risks.
- Delayed complications:
  - Arachnoiditis.
  - Tumor recurrence.
  - Spinal deformity or possible osteoporotic fractures at the level of laminectomy.

58.9 What Should Patient and Family Know?

Tumors may be incidental findings. If asymptomatic – they may stay dormant for many years without obvious progression. Therefore, "watch and see" attitude is acceptable in such cases, in particular in patients with low life expectancy.

Nevertheless, in most cases, the tumor will continue to slowly grow, and in symptomatic patients with good life expectancy, it is recommended to excise the tumor early.

Usually the tumor is benign, and in the vast majority the surgery is curative. However, recurrence is possible; therefore, long-term MRI surveillance is recommended.

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Spinal Nerve Sheath Tumors (NST): Schwannoma and Neurofibroma

59

Marcel Ivanov and Ion Poeata

59.1 Definition

Spinal nerve sheath tumors, e.g., schwannomas and neurofibromas, are slow-growing, usually benign tumors, which arise from the nerve root (Figs. 59.1 and 59.2).

The tumors affect equally men and women. If untreated, they may reach a considerable size and may cause significant morbidity.

Although in most of the cases solitary, they may be multiple and be part of more complex conditions—neurofibromatosis or schwannomatosis. These are separate entities that are not discussed in this chapter.

NSTs are equally distributed along the entire spinal neuraxis. Of NSTs, 60% to 80% are intradural, 10% are both intradural and extradural, and occasionally NSTs can be pure intramedullary or completely extradural or even extraspinal when they can reach impressive size before being diagnosed (Fig. 59.3).

Macroscopically schwannomas are more likely to grow on the periphery of the nerve with the preservation of some neural filaments which are displaced and remain

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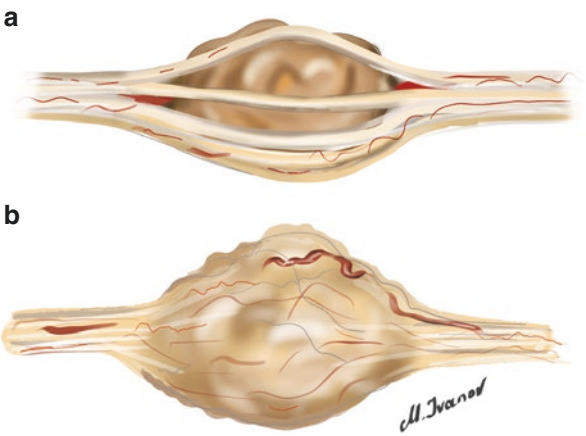

Fig. 59.1 Macroscopic appearance of a nerve sheath tumor. (**a**) schwannoma—the tumor is displacing normal fascicles to the periphery; (**b**) neurofibroma—the tumor appears as fusiform dilatation of the nerve. Surgical preservation of the nerve is usually not possible

intact. Neurofibromas appear as diffuse fusiform enlargement of the nerve (Fig. 59.1).

Histologically schwannomas have densely packed Antoni A areas in conjunctions with Antoni B areas and do not have cytologic atypia, which differentiates them from neurofibromas.

59.2 Natural History

The benign NST may remain dormant for many years. However, the tumor may continue to slowly grow. If untreated, they may cause initially some intermittent irritation and subsequently compression of the transiting neural structures inside the spinal canal with resulting symptoms of spinal cord or nerve root compression. When growing outside the spinal canal, the tumor may reach impressive size before becoming symptomatic [1].

In approximately 2% to 6% of cases, the tumors may suffer malignant transformation. This risk is much higher in patients with neurofibromatosis (NF-1)—up to 50%.

59.3 Physical Examination

The symptoms in patients with NST may result initially from the nerve root affected by the tumor. With tumor progression, it may compress other-transiting neural structures and the patients may develop symptoms from compression of the bypassing nerve roots or spinal cord.

Clinical examination should look for signs of the spinal cord or nerve compression with an assessment of gait, muscle power, sensation, reflexes, plantars, sphincters and Romberg probe (Video 59.4).

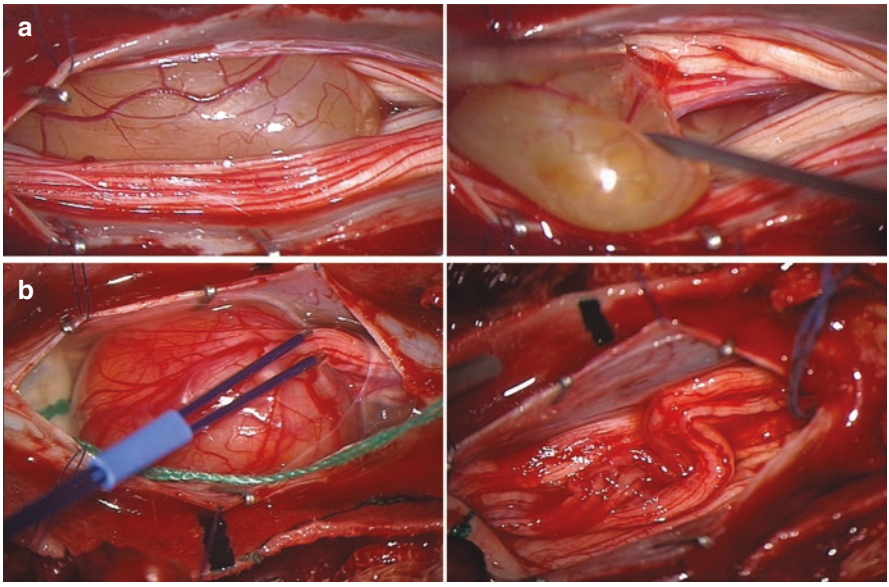

Fig. 59.2 (**a**) Intraoperative image demonstrating cystic nerve sheath tumor arising from the nerve root which is otherwise intact. (**b**) Left—direct intraoperative nerve root stimulation. Right—image after excision of the tumor with preservation of the integrity of the nerve root

In addition to detailed neurological examination (Videos 59.4 and 59.9), the doctor should perform a careful inspection with the assessment of signs of NF-1 (café au lait skin lesions, skin neurofibromas, spinal deformity, axial or inguinal freckles, hamartomas of the iris) (Chap. 23).

59.4 Imaging

Magnetic resonance imaging (MRI) scan is the method of choice for diagnosis of spinal NST, which appear as roundish lesions, located either entirely inside the spinal canal or extending into neural foramina as well outside the spinal canal.

On T1-WI and T2-WI, they usually have intensity similar to spinal cord and show moderate gadolinium enhancement. The tumors sometimes may have a cystic component, which is hypointense. Cystic tumors are more common in the lumbar spine and can be easily missed if the tumor wall is thin (Figs. 59.2 and 59.3).

Computed tomography (CT) scan may show bone remodeling. Myelo-CT can be an alternative imaging technique when an MRI scan is contraindicated; however, it will demonstrate only intradural tumor component.

It is recommended to scan the whole neuraxis and rule out other lesions.

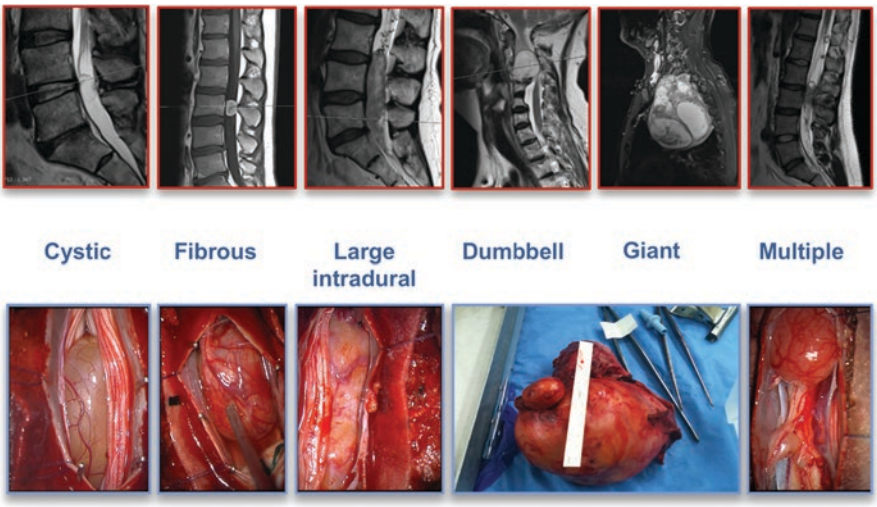

Fig. 59.3 MRI and intraoperative characteristic of the nerve sheath tumors

59.5 Differential Diagnosis

Other lesions that should be considered are meningiomas (Chap. 58), myxopapillary ependymomas (Chap. 60), and dermoid/epidermoid tumors. Cystic tumors should be differentiated from the arachnoid cysts (Video 59.8).

Nerve sheath tumor, when compared to meningiomas, showed a significantly higher frequency of cystic change (96% vs. 24%), neural foraminal extension (29% vs. 3%), and lumbar location (41% vs. 5%) [2].

59.6 Treatment Options

NST can be incidental findings. If completely asymptomatic, it is reasonable to keep them under surveillance. The tumor may remain dormant for decades; however, malignant transformation is possible.

In patients with significant or progressive symptoms, surgical resection is recommended.

After dural opening, the surgeon can see tumor arising from one nerve root. The tumor is generally mobile and not attached to dura. The transiting nerve roots or spinal cord appears displaced. If the spinal cord is severely compressed, the tumor can be initially debulked before manipulating it (not always necessary). The proximal and distal poles of the tumor should be identified.

We recommend the use of intraoperative neurophysiological monitoring with direct nerve stimulation (Fig. 59.2b). It can help to confirm whether the affected

nerve is still functional. In case of positive motor response on direct nerve root stimulation, the effort should be made to preserve its integrity. Careful microscopic inspection can help to identify the thinned and unaffected by the tumor neural filets on the surface/capsule of the tumor. With microscopic dissection, these filets can be separated from the capsule with preservation of their anatomical integrity (Fig. 59.2b).

When located in proximity to the spinal cord, the tumor may originate from the nerve root entry zone and may have intramedullary extension, which adds to the complexity of surgery.

In case of a dumbbell tumor, the surgery is more challenging. The challenges result from:

(a) Large dural defect that needs to be repaired.
(b) Need of removal of the intra- and extraspinal components, which may require two separate procedures.
(c) Need for spinal stabilization in case of excessive bone removal.

59.7 Expected Outcomes

The outcome after removal of a solitary nerve sheath tumor is usually good. The symptoms from compression of transiting neural structures would normally improve.

Due to slow tumor growth, the function of the affected nerve root frequently is taken by other nerve roots (functional reorganization), and in many patients with dumbbell tumors, transection of the affected nerve root does not always produce a new postoperative deficit.

Some new neurological deficit is nevertheless possible. As the tumor is arising predominantly from the sensory filets, 30% of patients may have new postoperative sensory deficit (usually well tolerated) and 5% new motor deficit [3].

Long-term follow-up reveals that 20% of patients considered themselves free of symptoms. The most common late complaint was local pain (46%), followed by radiating pain (43%), paraparesis (31%), and radicular deficit (28%) [4].

The recurrence rate is low, but possible therefore surveillance MRI scan is recommended [4].

59.8 Potential Complications

Surgery may cause the new neurologic deficit, CSF leak, infection, and bleeding. Transection of the T8-T12 nerve roots may lead to abdominal hernia.

In the late stages, the patients may develop spinal deformities such as scoliosis and kyphoscoliosis (6%), spinal arachnoiditis (6%), and persisting local or referring pain [4].

59.9 What Should Patient and Family Know?

When the tumor is asymptomatic or minimally symptomatic, it is reasonable to keep it under surveillance. The tumor may remain unchanged for decades or the rest of life; however, a small risk of malignant transformation exists.

The surgery is recommended in symptomatic patients or if the surveillance imaging shows tumour progression. The goal of surgery is to prevent further neurological decline. It can help to relieve symptoms from compression of the transiting neural structures; however, due to potential need to sacrifice one of the nerve roots, new postoperative deficit is possible.

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Spinal Ependymoma

60

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60.1 Definition

Ependymomas are rare glial tumors that arise from ependymal cells, which form the lining of the ventricles of the brain and the central canal in the spinal cord. Spinal ependymomas usually affect adults, with an incidence of 0.21 per 100,000, whereas intracranial ependymomas mostly develop in children.

60.2 Natural History

The median age of diagnosis is mainly reported as mid-40s ±15 years and a slight predominance in males with a male/female rate of 1.15. Histologically, ependymomas are classified in WHO grades I–III, with grade II being the most common in the spine. With 60%, spinal ependymoma represent the most common intramedullary tumor and are mostly of tanycytic histological type. They can also arise from the terminal thread (filum terminale), associated with the histological type of myxopapillary ependymomas, which account for 90% of tumors in this location (Table 60.1).

Supplementary Information The online version contains supplementary material available at (https://doi.org/10.1007/978-3-030-80356-8_60).

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Table 60.1 Distribution of location of spinal ependymoma

Location in the spine	
Cervical	35%
Thoracic	21%
Lumbar	69%
Intramedullary	64%
Extramedullary or filum	36%

They present as intradural, extramedullary lesions; however, by definition, they are intramedullary lesions, as they arise from the central canal, too.

An important characteristic of ependymoma is the possibility of leptomeningeal drop metastasis. In children, ependymomas primarily manifest intracranially, although leptomeningeal drop metastasis can occur in the spine. Therefore, contrast-enhanced magnetic resonance imaging (MRI) of the brain and the whole spine is the gold standard for the diagnosis of intraspinal tumors. Additionally, at least two lumbar punctures and cerebrospinal fluid (CSF) examinations for drop metastasis are advised.

60.3 Physical Examination

Overall, cervical and lumbar are the most common locations of spinal ependymoma, and symptoms are primarily determined by the location, possibly leading to sensorimotor deficits, impairment of coordination and/or vegetative symptoms, such as bladder and bowel dysfunction. Clinically, spinal ependymoma often presents as incomplete paraparesis, although pain, sensory and motor impairment can also occur in a radicular pattern when nerve roots are affected. Thus, a thorough neurological exam should be performed, consisting of motor function tests, sensory evaluation, straight leg raise, Romberg's test, heel-knee test, and digital rectal exam (Video 60.4).

60.4 Imaging

On spinal MRI, ependymomas show a homogeneous contrast enhancement on the T1 sequence. Intramedullary manifestations typically cause symmetric widening of the spinal cord. Syringohydromyelia can be present. In the T2 sequence, the so-called cap sign, hypointense hemosiderin areas adjacent to the lesion caused by hemorrhage, are suggestive of ependymoma (Figs. 60.1, 60.2, and 60.3).

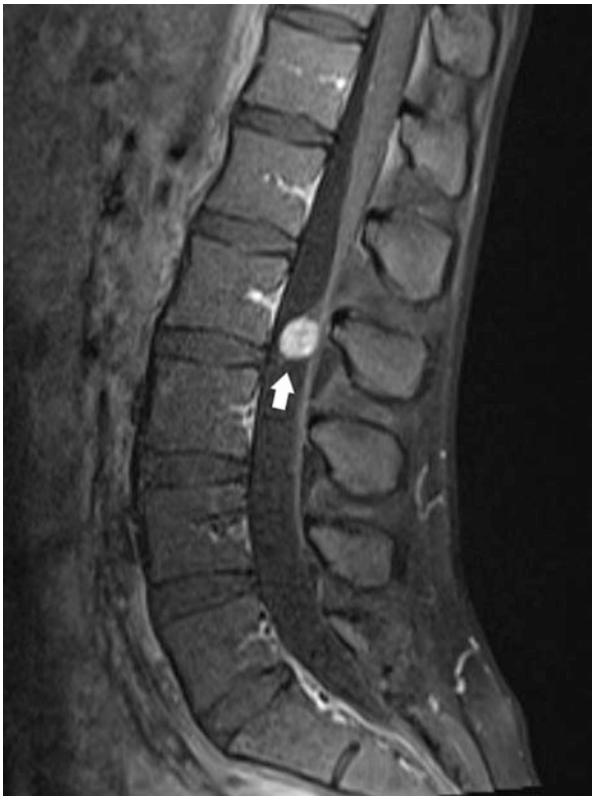

Fig. 60.1 MRI T1 sequence post-contrast of the lumbar spine in sagittal view: a white arrow pointing at a myxopapillary ependymoma in the lumbar spine, on level of lumbar vertebrae 2–3 in a 31-year-old female

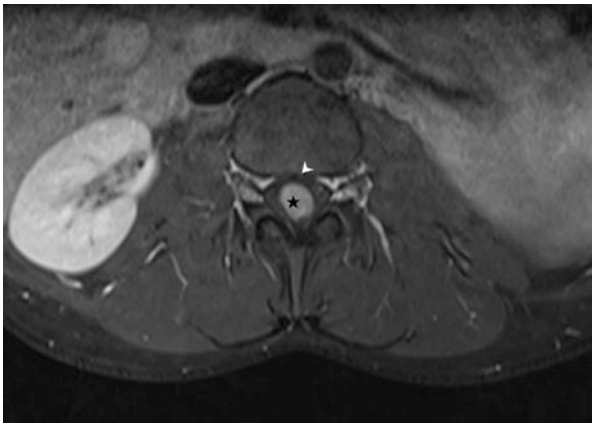

Fig. 60.2 MRI T1 sequence post-contrast in axial view on the level of lumbar vertebrae L2–3: Myxopapillary ependymoma (black star), white arrowhead pointing at the remaining cerebrospinal fluid-filled dural sac

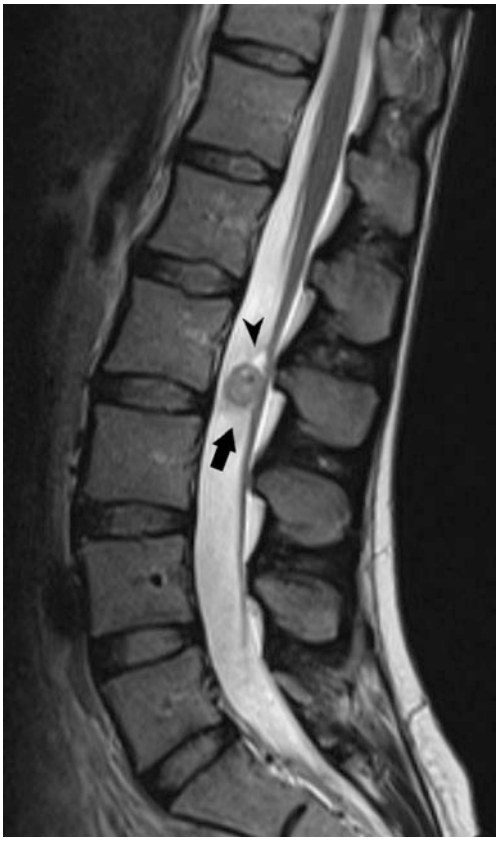

Fig. 60.3 MRI T2 sequence of the lumbar spine in sagittal view: Arrow pointed at the myxopapillary ependymoma, arrowhead pointed at "cap sign"

60.5 Differential Diagnosis

Differential diagnoses of intraspinal, extramedullary lesions are schwannoma, spinal meningioma, metastasis, and lipoma. The differential diagnoses of intramedullary lesions are hemangioblastoma, astrocytoma (Chap. 37), dermoid, epidermoid, teratomas, and rarely intramedullary metastasis (Chap. 63). In clinical practice, astrocytoma of the spinal cord is in fact the most frequent and most relevant differential diagnosis since a clear separation of both tumor types is not always possible via MRI, especially when cysts are missing and the contrast uptake is atypical. Here, the intraoperative appearance and biopsy will only lead to a final diagnosis (Video 60.8).

60.6 Treatment Options

Most spinal ependymomas are growing slowly and encapsulated, tending to displace the spinal cord rather than to infiltrate it. There is usually a dividing layer between the tumor and the spinal cord tissue allowing for a better separation. Thus, first-line treatment of spinal cord ependymomas aims at gross total resection (GTR)

with preservation of healthy tissue and neurological function. GTR is defined as resection without visible residual tumor, confirmed in the postoperative MRI, and can be achieved in the vast majority. It is the main prognostic factor for a progression-free survival (PFS). In order to maintain neurological function and reduce postoperative complications, intraoperative neurophysiological monitoring (IONM) is mandatory. However, feasible GTR depends on various factors including location, growth pattern, and size. As mentioned above, spinal ependymomas sometimes cannot be distinguished well from intramedullary astrocytomas. In this case, the surgical strategy (aiming for GTR in ependymomas versus biopsy/decompression only for astrocytomas) depends on the results of intraoperative fresh frozen biopsies. However, the surgeon has to be aware that the reliability of intraoperative biopsy assessment is only around 70–80%. Thus, in some cases, a staged strategy has to be applied with biopsy first and then secondary GTR if ependymoma is confirmed.

If GTR of ependymoma cannot be achieved, postoperative radiation therapy can be performed. However, there are no guidelines for when radiation therapy should be performed, and every case should be evaluated individually. Generally, radiation therapy is recommended for patients with incomplete resection, anaplastic ependymomas, and leptomeningeal spread. For completely resected ependymoma, the controversy about postoperative radiation therapy is even more prevalent.

60.7 Potential Complications

In pretreated or infiltrating ependymomas, surgical removal is challenging, and the risk of spinal cord irritation due to intraoperative traction and manipulation is high. Infiltration also increases the risk of tumor recurrence due to incomplete resection. Theoretically, tumors located in the higher cervical spinal cord are associated with a higher risk of severe neurological impairment including respiratory dysfunction and quadriplegia, which can cause significant functional disability. However, in our experience, the cervical spinal cord is more forgiving than the thoracic spinal cord, where deficits tend to be more complete and more permanent. Further complications include wound healing problems, CSF spread of tumor cells, and CSF leak.

Potential orthopedic complications (spinal deformities) secondary to the surgical approach must be kept in mind.

60.8 Expected Outcomes

While most patients suffer from sensory deficits, increased neurological impairment including worsening of motor function and spinal ataxia only occurred in about 37% with 9% experiencing a new significant deficit. In the long term, however, 41% improved and 35% recovered to the preoperative state, leaving only 2% with new and permanent severe deficits. Increased age appeared to be the only independent predictive factor for permanent neurological deficits. In general, the outcome is better in patients with fewer preoperative deficits and smaller tumor size, making an early diagnosis essential.

60.9 What Should Patient and Family Know?

Whereas most of the neurological deficits are transient, postoperative disability might prolong in-hospital stay and delay return to normal life. Especially older age is associated with a higher rate of permanent deficits.

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Plasmacytoma

61

İsmail Daldal, Aliekber Yapar, and Alpaslan Şenköylü

61.1 Definition

There is a wide range of clinical highlights, from monoclonal gammopathy of obscure importance to multiple myeloma (MM) to plasma cell leukemia. Most of the plasma cell tumors are analyzed as MM and will in general influence a more established grown-up population. In any case, a minority (<5%) of patients with plasma cell malignancies present with either a solitary bone mass called solitary bone plasmacytoma (SBP) or less ordinarily a soft tissue mass of monoclonal plasma cells called solitary extramedullary plasmacytoma (SEP).

SBP is a plasma cell problem described by a localized collection of neoplastic monoclonal plasma cells in the bone. SBP has a male/female ratio of 2:1, with an average age of 55 years. Solitary type essentially involves axial skeleton, particularly the vertebrae.

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Malignant bone tumors of the spine are extremely uncommon. SBP is the most common separate tumor inside this gathering, representing around 30% of the aggregate. These tumors place in the spinal bony structures twice as frequently as other bones.

61.2 Natural History

SBP has a high incidence of progression to MM, and on the magnetic resonance imaging (MRI) assessment, at any rate, 25% of patients with an evident solitary mass have proof of disorder somewhere else. Interestingly, SEP is almost in every case genuinely restricted and has a high healing rate with local treatment options. Most of the patients with obvious SBP progress to myeloma, with a middle opportunity to the progression of 2 to 4 years. The middle by and large endurance in various studies fluctuates from 7.5 to 12 years.

If a MM patient has features including low levels of uninvolved immunoglobulins, axial disease, older age, lesion size >5 cm, and persistence of the M protein after treatment, it will result in disease progression. The presence of M protein has been accounted for in 24% to 72% of patients in various studies. The recurrence presumably relies upon the degree of affectability of the tests utilized.

61.3 Physical Examination

The most common manifestation is the pain at rest (Chap. 41); however, it can likewise present spinal cord or root compression findings (Video 61.4). Inclusion of the base of the skull can give cranial nerve compromise (paralysis).

61.4 Imaging

Just as the proper blood and urine tests, imaging of the spine is required and best accomplished by MRI and computed tomography (CT) scan. A mini-brain appearance has also been described in MRI (Fig. 61.1). It is seen as curvilinear low-signal-intensity areas within the lesion, giving an appearance of sulci in the brain [1]. Indeed, this appearance is a trademark to such an extent that it might block the requirement for a symptomatic biopsy. The biopsy is generally conceivable percutaneously, guided either by fluoroscopy or CT (Video 61.8). Positron emission tomography (PET) has as of late been assessed in the organizing of patients with myeloma and plasmacytoma. PET, similar to MRI, seems to be valuable in identifying occult disease in patients with SBP.

There are no detailed rules to characterize contribution on MRI scans with regard to evident SBP. Be that as it may, the MRI appearances in MM have been very much portrayed. The presence of at least one foci of anomalous sign force, for example,

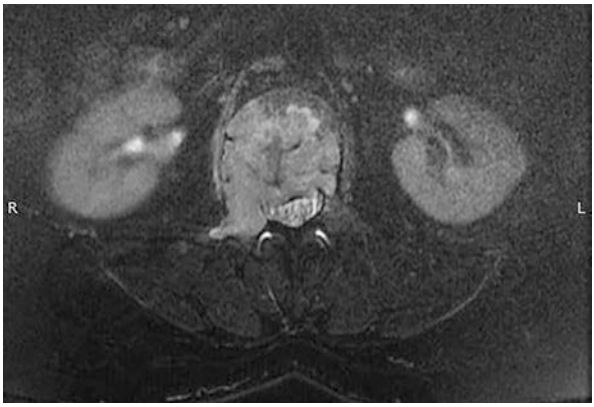

Fig. 61.1 A case with "mini-brain" appearance in axial MRI (L2 vertebrae)

low on T1-weighted imaging and high on T2-weighted or STIR (short TI inversion recovery) images, which upgrade after the organization of paramagnetic differentiation without realized late pressure cracks, other essential danger, or average attributes of generous or harmful essential bone tumors, is viewed as proof of removed association in patients with obvious SBP.

61.5 Differential Diagnosis

The accompanying measures are suggested:

- Single region of bone destruction because of plasma cells.
- Histologically <5% plasma cells in bone marrow suction.
- Totally normal outcomes on skeletal overview including radiology of long bones.
- No hypercalcemia or renal debilitation due to plasma cell dyscrasia.
- Missing or low serum or urinary degree of monoclonal immunoglobulin (level of >20 g/l) dubious of MM.
- No extra lesion on MRI of the spine.

SBP is for the most part diagnosed by biopsy or fine-needle aspiration. Percutaneously guided biopsy of the spine is typically conceivable either by fluoroscopy or CT (Videos 61.7 and 61.8). As these tumors are uncommon, pathology survey by a pathologist with an exceptional interest in either bone tumors or lymphoproliferative issues is firmly suggested.

61.6 Treatment Options

Although the mainstay treatment for MM is systemic chemotherapy, radiotherapy often has an important supportive role, offering very effective symptom relief for plasmacytomas in bone or soft tissues. Radical radiotherapy is the preferred

treatment for SBP. As with other rare tumors, the evidence base for treatment consists largely of retrospective studies of a small number of patients. Progression to MM is common, despite the high local control rates of 83% to 96% achieved with medium-dose radiotherapy. Based on the results of retrospective case studies, a dose of 40 Gy in 20 fractions is recommended for SBP. For SBP larger than 5 cm in size, a higher dose up to 50 Gy in 25 fractions should be considered [2]. An alternative strategy for tumors larger than 5 cm is chemotherapy followed by radiotherapy. This is theoretically attractive, but there is little published evidence to support it.

Radiotherapy remains the treatment of choice of the primary pathology, and surgery is contraindicated in the absence of structural instability or neurological compromise. However, early diagnosis and referral for a neurosurgeon/orthopedic surgeon opinion is advised in most cases with spinal involvement. Due to the development of modern spinal fixation systems over the last decade, surgical treatment is now a viable and successful option for patients who develop pain caused by structural compromise within the vertebra, vertebral instability, neurological compromise, or a combination of these.

Close contact between hematologist, radiotherapist, and neurosurgeon/orthopedic surgeon is along these lines essential in planning ideal treatment options for patients.

61.7 Expected Outcomes

As a radiotherapy and/or chemotherapy-sensitive neoplasm, the prognosis of plasmacytoma with conservative treatment is satisfactory in general.

61.8 Potential Complications

Spinal instability and neurological compromise are extremely rare.

61.9 What Should Patient and Family Know?

SBP is a rare disease in the spectrum of plasma cell dyscrasias. Solitary lesions have a high potential to transform into MM.

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Chordoma

62

Mehmet Ali Deveci and S. Aykın Şimşek

62.1 Definition

Chordoma is an aggressive, locally invasive tumor representing 1% to 4% of bone tumors; the prognosis is generally poor. Although it is classified as a low-grade tumor, its clinical course resembles malignant tumors because of its high recurrence rate and infiltrative growth pattern [1]. It was first described by Virchow in 1857, but Ribbert used the term "chordoma" for the first time in 1890 [2]. Its incidence is <0.1/100.000, and it is mostly seen in the fifth decade of life; the tumor is hardly ever seen in individuals younger than 30 to 40 years of age, and it is more common among men; about 5% of all chordomas develop during adolescence [1].

Chordomas originate from the remnants of the notochord. Undifferentiated notochord remnants in the form of microscopic foci can be found within the vertebral bodies at the cranial and caudal ends of the embryo. Chordomas can rarely develop from normal notochord remnants such as the nucleus pulposus. Malignant transformations from these residues occur during the third to fourth decade in sphenooccipital lesions and during the fifth to sixth decades in sacrococcygeal lesions. The most important evidence proving the notochord origin of the tumor is the 6q27 duplication detected in the transcription factor-T gene (brachyury) of familial chordomas. Brachyury is found in the normal, undifferentiated embryonic notochord of the axial skeleton [3].

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62.2 Natural History

The prognosis is better in patients aged <40 years; the overall survival rate is 6 years on average, and the 5-year survival is 70%, while 10-year survival is 40%. Local recurrence is extremely frequent.

62.3 Physical Examination

Chordomas are mostly midline lesions, and 32% of them are located in the skull base, 32% in the mobile vertebrae, and 29% in the sacrum [2]. Since chordomas are slow-growing, locally aggressive, and invasive tumors, the disease frequently concerns bone and neural structures. For this reason, effective treatment is very difficult because of the large lesions at the time of diagnosis, the involvement of neural structures, compression of the surrounding soft tissues, inability to obtain wide margins during surgery, and resistance to radiotherapy.

Since chordomas are insidious and slow-growing tumors, they tend to remain asymptomatic until the later stages of the disease.

Depending on their size and involvement of the sella, they may cause endocrinopathies, rarely epistaxis, and intracranial bleeding. Cervical chordomas can cause airway obstruction and dysphagia and appear as oropharyngeal masses.

Involvement in the mobile spine presents symptoms according to deep-seated pain (Chap. 41) and the level of the involved nerve. Constipation and sphincter paralysis, bladder dysfunction, gait disturbances, and hypoesthesia can be seen with the growth of the mass. Neurological deficits are more common in mobile vertebral involvement than in sacrococcygeal involvement (Video 62.4).

Sacrococcygeal chordomas often involve S4–S5, and the masses usually enlarge into the pelvis. The invasion of the pelvic structures is limited by the presacral fascia. Rarely, it may present as masses that grow into the postsacral area and are palpated externally.

62.4 Imaging

Midline-located chordomas appear radiographically as destructive bone lesions, and typically the destruction starts from the midline. While chordomas spread to the adjacent spine, they may locally invade the intervertebral discs. Lateral radiographs are more useful in sacrococcygeal chordoma (Fig. 62.1).

Computed tomography (CT) is more useful in showing lesions located within the midline spine. It may show lesions containing scattered calcifications and extending into the bone with soft tissue involvement (Fig. 62.2).

Magnetic resonance imaging (MRI) depicts the tumor as iso- or hypointense in T1-weighted images and as hyperintense in T2-weighted images; soft tissue

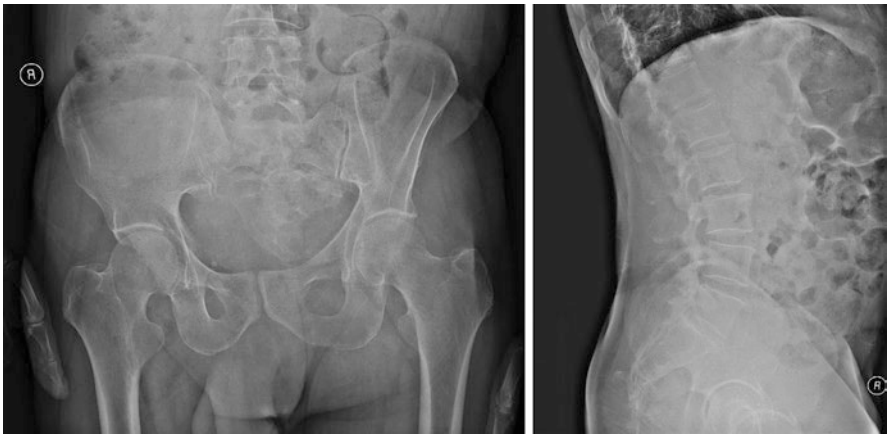

Fig. 62.1 Sacral chordoma. Radiography of the anteroposterior pelvis and lateral lumbosacral vertebra

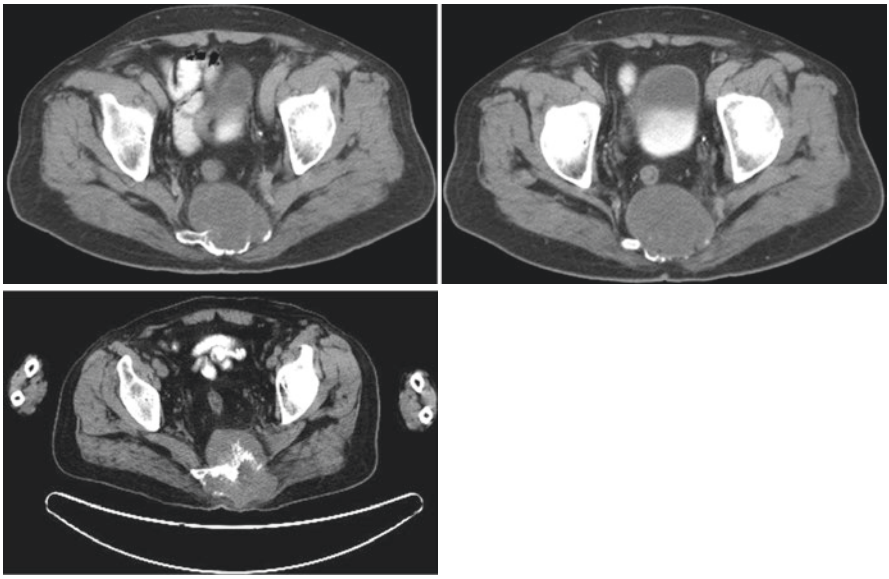

Fig. 62.2 Sacral chordoma. CT findings show destruction of sacrum with soft tissue lesion

component heavily retains gadolinium. MRI gives a better idea about the soft tissue involvement (Fig. 62.3).

Chordomas show normal or decreased uptake than other bone tumors with the Tc99m bone scintigraphy.

The PET/CT with 18-FDG shows an increased systemic uptake values (SUV) highlighting the aggressive nature of the tumor (Fig. 62.4).

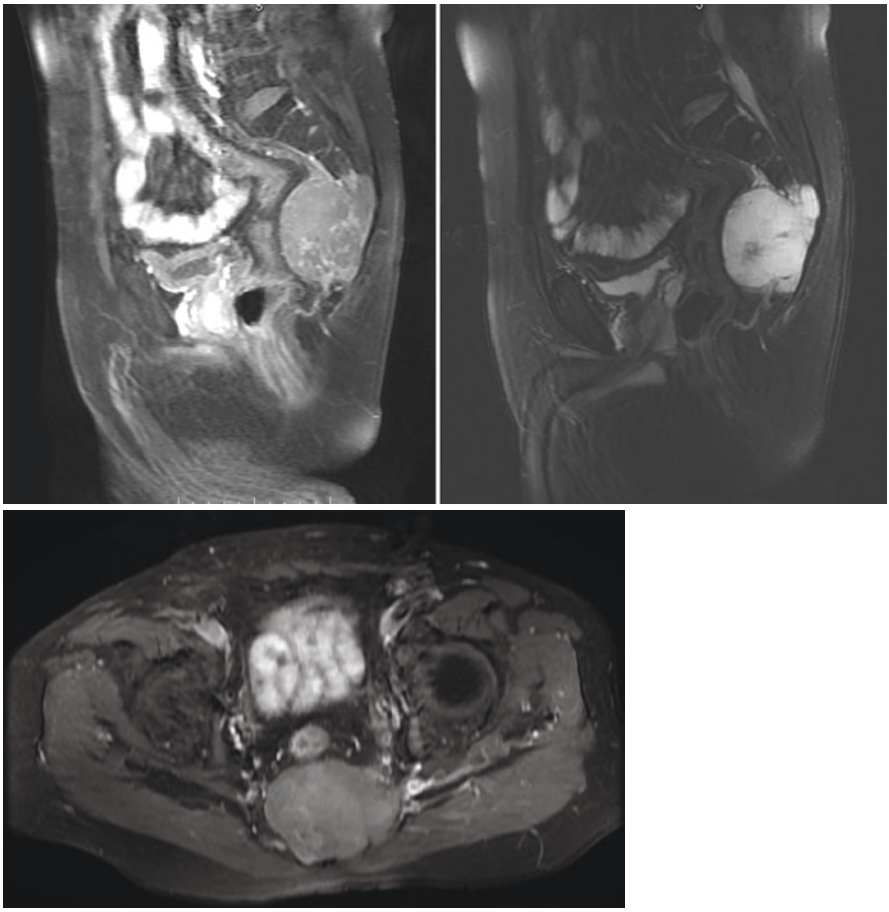

Fig. 62.3 Sacral chordoma. Wide presacral soft tissue involvement of the tumor in T1 sagittal, T2 fat-suppressed sagittal, and T1 axial sections

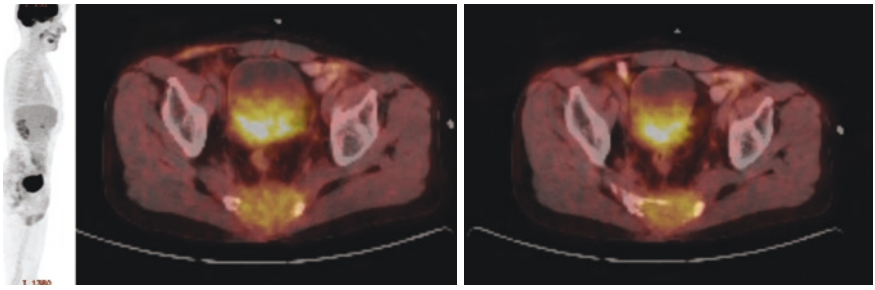

Fig. 62.4 PET/CT images of sacral chordoma

62.5 Differential Diagnosis

The histological diagnosis can be done with a needle biopsy under CT scan guidance (trans-pedicular) in about 80% of cases (Video 62.8); open biopsy is exceptional, and it should be performed if two successive needle biopsies are not conclusive.

Pathology. Chordomas show various degrees of histological atypia, and the relationship between these histopathological findings and tumor behavior is the subject of many studies. There are three separate histological subgroups: classical (conventional), chondroid, and dedifferentiated.

The classic chondroma is a soft, gray-white, lobulated tumor (Fig. 62.5). Cell groups settled in a myxoid stroma are separated from each other by fibrous septa. Cells are round, have regular nuclei, and show little nuclear atypia. They are characterized by prominent vacuole cytoplasm. These vacuoles give the cell a physaliphorous (bubbly) appearance (Fig. 62.6). On the other hand, chondroid chordomas contain cell structures belonging to both classical chordoma and

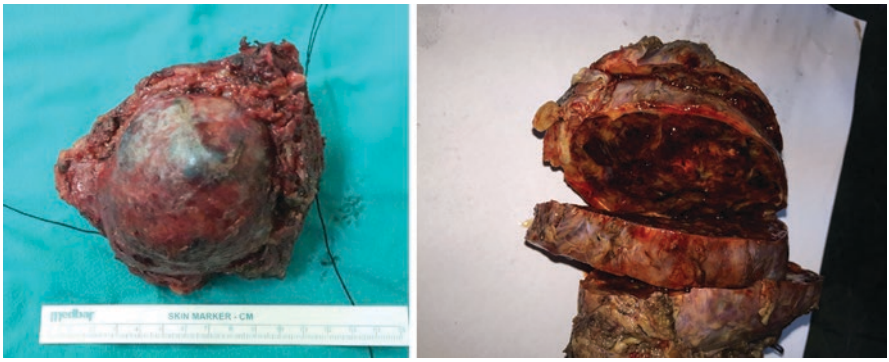

Fig. 62.5 Macroscopic views after chordoma resection; interior surface shows lobulated, gelatinous nature of the tumor

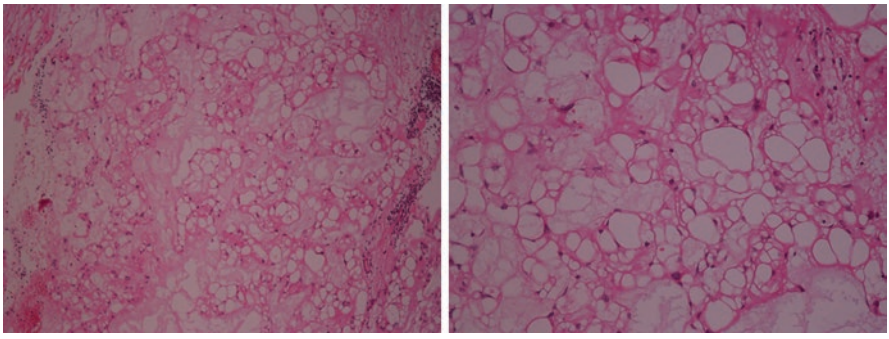

Fig. 62.6 Chordomas appear as lobulated cells separated by fibrous septa within the mucinous matrix. The vacuole structure in the cytoplasm of the cells is remarkable

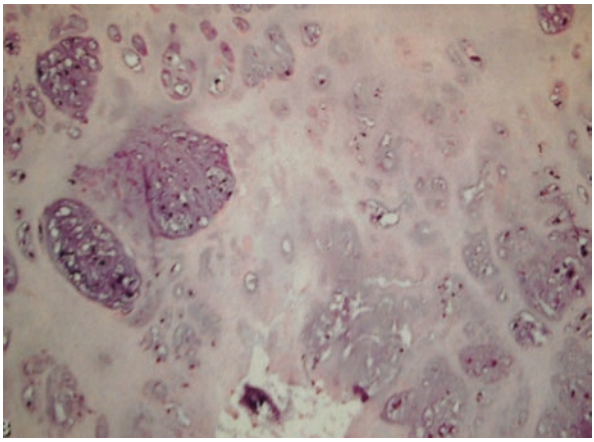

Fig. 62.7 Chondroid chordoma. Chondroid tumor islets are seen among the tumor cells

chondrosarcoma forming malign cartilage (Fig. 62.7); chondroid chordomas are common in lesions located in the skull base [1]. Besides its classical physaliphorous appearance, it is characterized by ability to stain immunohistochemically with epithelial markers such as chordoma S-100 and epithelial membrane antigen (MUC1) and cytokeratins [4]. In some studies, the transcription factor for notochord development, brachyury, has been shown to be a good marker for chordomas.

The differential diagnosis of chordoma include chondrosarcoma, myeloma (Chap. 61), lymphoma, giant cell tumor (Chap. 57), aneurysmal bone cyst (Chap. 56), and metastases (Chap. 63). Since both chondrosarcomas and chordomas are S-100 positive, it should not be forgotten that if cytokeratin evaluation cannot be made clear in small biopsy samples, there may be problems in differentiating chondrosarcoma from chordoma.

62.6 Treatment

*Surgical Treatment***.** In the 1970s, Stener and Gunterberg introduced the idea of en bloc resection for the treatment of sacral tumors [2]. It is very difficult to obtain a wide surgical margin due to the complex anatomy and the challenges of reconstruction, in particular filling the dead space and the potential neurological deficits (especially above S2 level). Risk of recurrence is twice as high if the tumor capsule is damaged during resection; the local recurrence of sacral chordomas is about 2 years in patients undergoing radical resection, and it is 8 months in the other patients.

The posterior transperineal approach alone is often sufficient for chordomas located below the sacroiliac joint. Resection is more difficult in lesions located in the caudal part of the sacroiliac joint which require both anterior and posterior approaches. If S2 roots can be preserved, it is possible to obtain normal bladder and anal sphincter functions in about 50% of cases. Loss of more than 50% of the

sacroiliac joint causes vertical and rotational instability in the lumbopelvic region and requires lumbopelvic stabilization.

In lesions located in the sacrum, flaps such as gluteus maximus flap and vertical rectus abdominis muscle (VRAM) flap should be used to fill the dead space after total resection (Fig. 62.8).

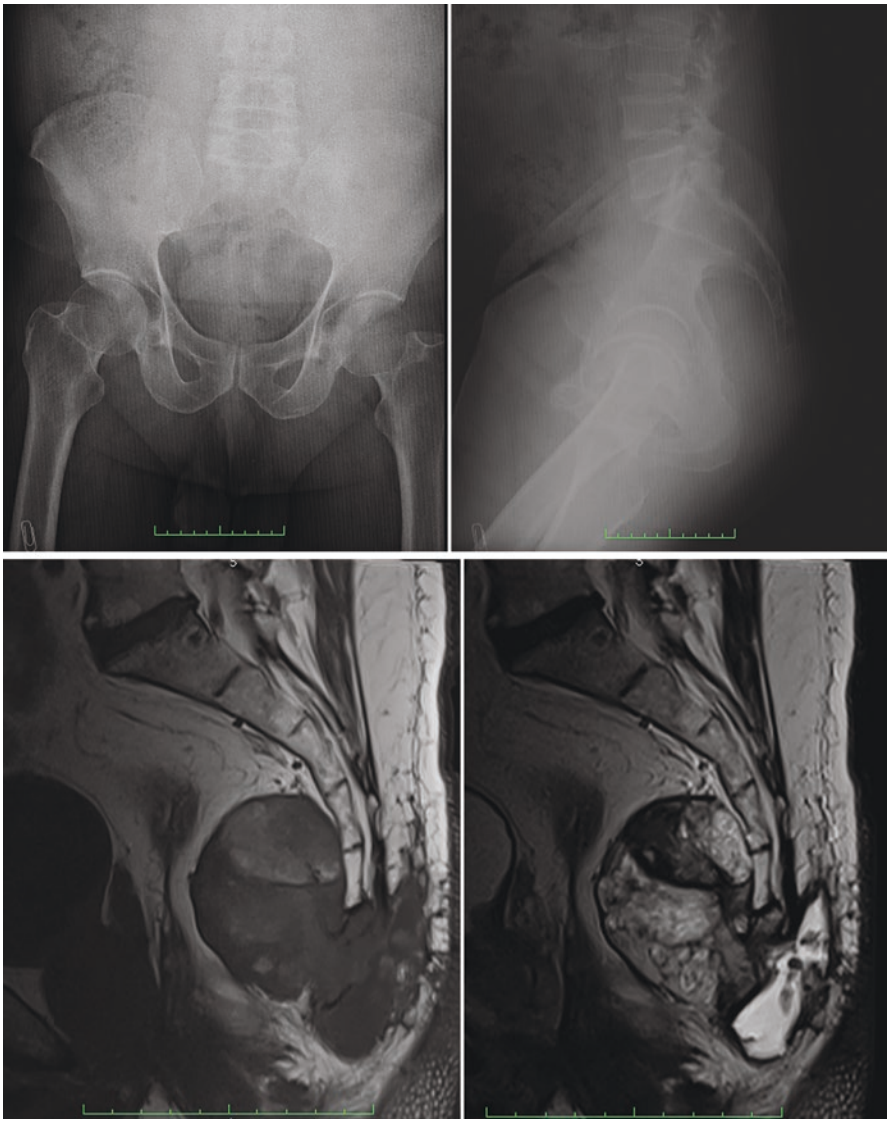

Fig. 62.8 A 69-year-old male with sacral chordoma. Wide resection with closure of the defect with gluteal flap

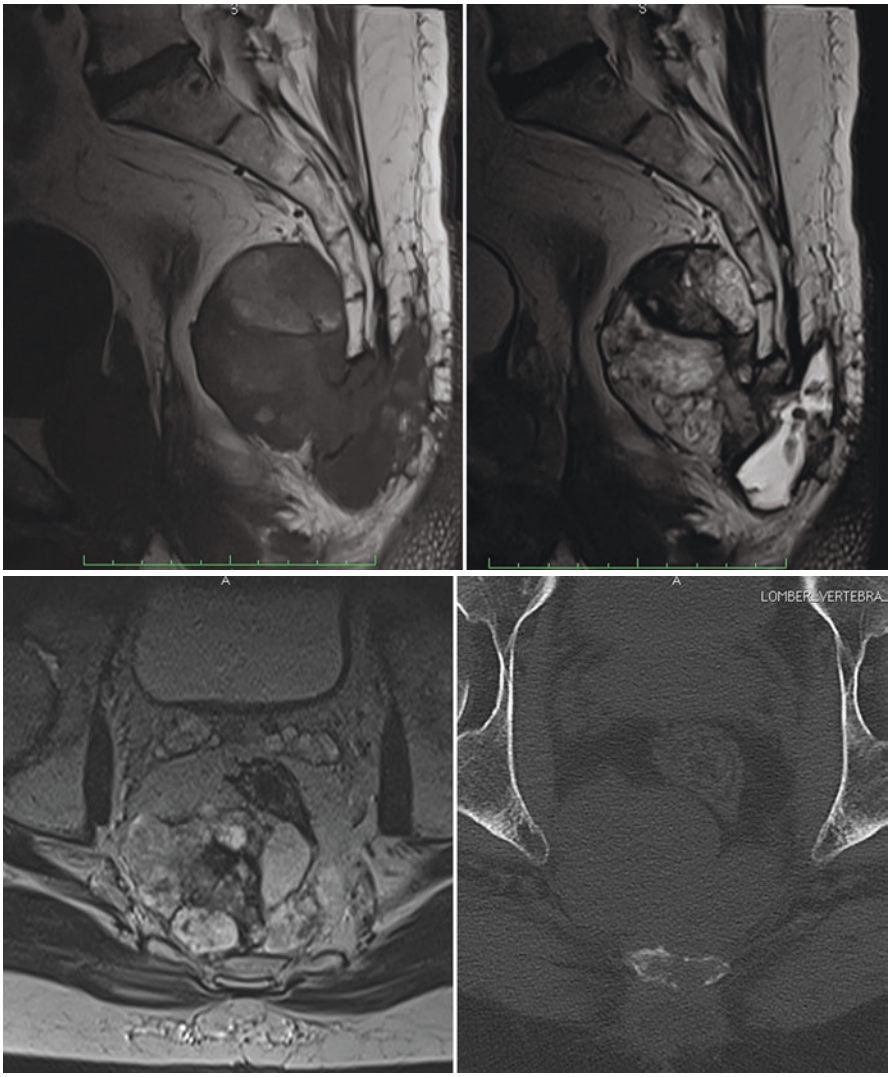

Fig. 62.8 (continued)

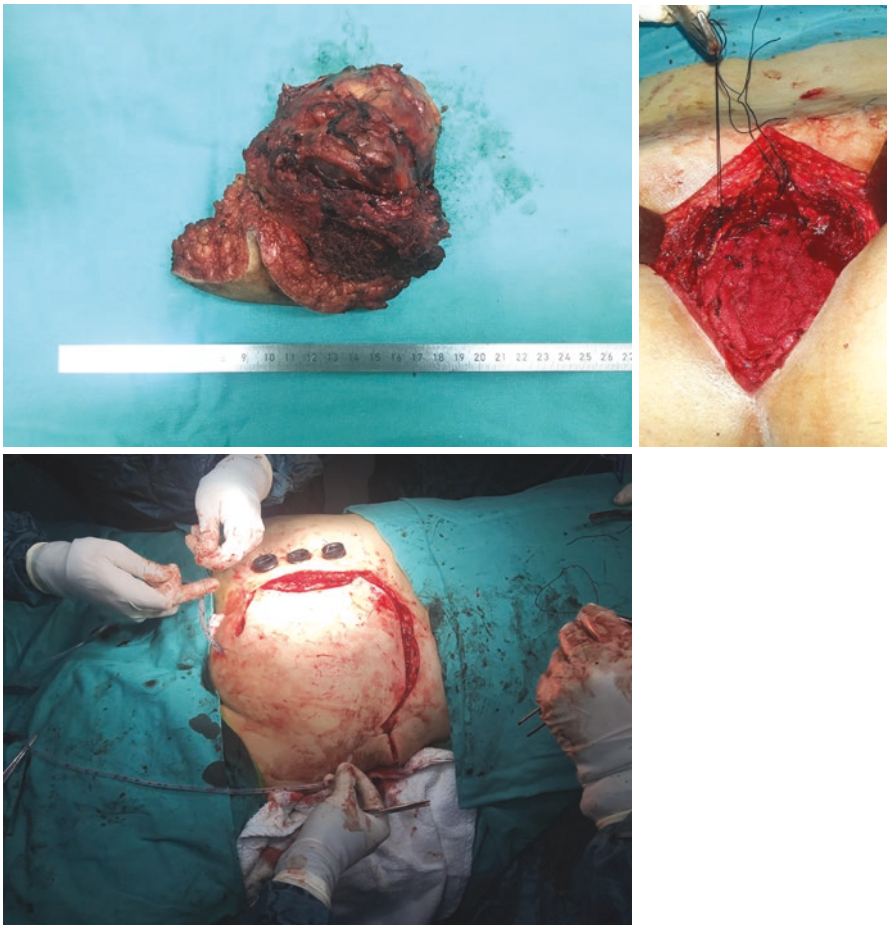

Fig. 62.8 (continued)

Medical Treatment. Despite the advances in surgical treatment, en bloc resection is only possible in only 50% of sacral tumors; this percentage is lower in tumors of the mobile spine and the skull base.

Chordomas are known as radioresistant tumors and require doses above 60 Gy. Conventional radiotherapy applied with high-energy photons can only achieve 50–55 Gy and cannot provide local control. With the development of methods using particle radiation (especially protons) and improvements in the preservation of normal structures by imaging, planning, and directing radiotherapy to target tissues, better tumor control has been achieved with fewer side effects [4]. Especially in cervical lesions or lesions located at the skull base, where extensive surgeries are not possible, better results are obtained with razoxane use, which increases radiosensitivity. There are also limited studies on radiotherapy with particles heavier than protons such as helium and carbon ions. Compared with helium ions, carbon ions

have been shown to have more biological advantages and are more effective in the treatment of chordoma [5].

Chordomas are not sensitive to chemotherapy like all other low-grade tumors. The effectiveness of chemotherapy has been demonstrated in dedifferentiated chordomas, which constitute only 5% of chordomas.

In recent years, molecular-targeted agents have also begun to be tried in the treatment of chordomas. It has been shown that chordomas use the platelet-derived growth factor receptor beta (PDGFR-ß) and its phosphorylated form for activation, and tumor shrinkage has been shown as a result of targeted treatments. With the use of 850 mg of imatinib mesylate, which is a tyrosine kinase inhibitor, a decrease in tissue activity has been demonstrated [6]. The epidermal growth factor receptor pathway may also constitute another target for drugs. Studies are carried out on the combination of cetuximab and gefitinib.

62.7 Expected Outcomes

Chordomas have metastasized in 5% of cases at the time of diagnosis. Lung, bone, skin, and brain are the most common areas for metastasis. The metastasis rate is up to 65% in advanced disease or recurrent lesions. The survival rate is affected by the presence of metastases rather than local progression. It has been shown that the average survival after the development of distant metastasis is less than 12 months. Metastases are often stable, and the patient is lost due to local tumor spread and invasion of vital organs. Although it has been shown that the prognosis is not affected by the histological type, there is information proving that chondroid chordomas have a better prognosis.

62.8 What Should Patient and Family Know?

Although chordoma is a low-grade tumor, its clinical course resembles malignant tumors because of its high recurrence rate and infiltrative growth pattern; the prognosis is generally poor. The overall survival rate is 6 years on average, and local recurrence is extremely frequent.

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Metastatic Lesions of the Spine

63

Vanessa Hubertus, Peter Vajkoczy, and Julia Sophie Onken

63.1 Definition

Metastatic lesions of the spine commonly occur in advanced malignant disease. More than 50% of tumor patients suffer from bone metastases, with the spine presenting the most common site. Spinal metastases can be symptomatic or asymptomatic. At autopsy, 30% to 90% of tumor patients present with previously undiagnosed spinal metastases. Around 5% of patients with spinal metastases present with acute neurological deficits due to metastatic epidural spinal cord compression (MESCC), thus drastically reducing their health-related quality of life (HRQOL) and their statistic life expectancy [1].

63.2 Natural History

Malignancies underlying spinal metastases present a heterogenic group; the most common are prostate, breast, kidney, and lung cancers. A significant role also play hematologic diseases such as multiple myeloma. In the spinal column, the thoracic spine is the most frequent localization for metastatic lesions, followed by the lumbar spine. The least frequent localization presents the cervical spine, although

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clinically highly relevant due to a higher occurrence of neurological deficits in this region. Special attention in the surgical treatment of spinal metastases must be paid to metastases at the junctional zones—the craniocervical, cervicothoracic, thoracolumbar, and lumbosacral junctions, for their role in spinal stability.

63.3 Physical Examination

In the examination of a patient potentially suffering from spinal metastases, specific *red flags* must be assessed: Does the patient suffer from a new neurological (motor or sensory) deficit, bladder or bowel dysfunction, ataxia, new axial or radicular pain, or paraparesis / paraplegia? It is important to assess the acuity of the symptoms in order to speed up the diagnosis process accordingly. In MESCC, a classic spinal emergency, "time is spine." The faster surgical decompression is applied, the better is the neurological outcome of the patient. Next to the neurologic examination (Video 63.4), assessment of clinical status using the Karnofsky performance status (KPS) and comorbidities is necessary, as is the clinical assessment, a thorough history of tumor diagnosis, staging, and prior treatment.

63.4 Imaging

Magnetic resonance imaging (MRI) is the gold standard with the highest sensitivity in detecting spinal metastases. Usually, contrast-enhanced T1-weighted protocols are used to display the extent and limits of the metastatic lesion and their invasion into the spinal canal. In patients with spinal metastases, MRI of the whole spine should be performed, as metastatic lesions often occur multifocally. To assess bone involvement, with differentiation of osteoblastic and osteoclastic lesions, additional high-resolution computed tomography (CT) is necessary. Alternative imaging like bone scintigraphy or positron emission tomography (PET) is commonly used in staging examinations and shows a lower resolution than CT or MRI; therefore, they cannot replace these imaging modalities. The use of spinal angiography is restricted to the preoperative display and embolization of highly vascularized metastases, such as originating from renal cell or thyroid carcinomas.

63.5 Differential Diagnosis

One important differential diagnosis to rule out before surgery is a primary bone tumor, such as chordoma (Chap. 62), Ewing sarcoma (Chap. 39), and chondro- or osteosarcoma (Chap. 38). These tumors are quite rare but necessitate a gross tumor resection due to their aggressive growth behavior, radioresistance, and potential for tumor cell distribution during surgery. In suspected cases, tumor biopsy should precede definite surgery (Video 63.8).

63.6 Treatment Options

The primary goals of surgery for spinal metastases are the preservation of neurological function, pain relief, mechanical stabilization, and local tumor control (Video 63.1). Ideally, surgical decision-making is based on an interdisciplinary approach, considering four factors: *n*eurological and *o*ncological status, *m*echanical stability, and *s*ystemic disease (the NOMS-decision framework by Laufer et al., Table 63.1) [2]. Evaluation of oncological status should include the expected response to nonsurgical tumor treatment and radiation sensitivity according to the tumor's histopathology. Spinal stability is assessed according to the Spinal Instability Neoplastic Score (SINS) [3], using CT and MR imaging of the spinal lesion (Table 63.2). Systemic disease assessment includes the calculation of expected patient prognosis according to tumor and treatment history and comorbidities. Different prognostic scores were established for this purpose, like the Tokuhashi and the Tomita scoring systems—their clinical applicability though remaining limited.

Surgical options include decompression and stabilization surgery, dependent on spinal stability according to SINS. Biopsies are functional for differential diagnosis and can be combined with either decompression, stabilization, kyphoplasty or vertebroplasty. In case of extensive lytic disease or disease at junctional zones, 360° stabilization and decompression with vertebral body replacement might be necessary (Fig. 63.1). To reduce surgical risks, minimally invasive percutaneous stabilization using spinal navigation is trending. When instrumentation and fusion of the

Table 63.1 The Neurologic Oncologic Mechanical Systemic (NOMS) decision framework by Laufer et al. [2]

Neurologic	Oncologic	Mechanical	Systemic	Decision
Low-grade MESCC without myelopathy	Radiosensitive	Stable		EBRT
		Unstable		Stabilization + cEBRT
	Radioresistant	Stable		SRS
		Unstable		Stabilization + SRS
High-grade MESCC with/ without myelopathy	Radiosensitive	Stable		EBRT
		Unstable		Stabilization + cEBRT
	Radioresistant	Stable	Able to tolerate surgery	Decompression + Stabilization + SRS
		Stable	Unable to tolerate surgery	cEBRT
		Unstable	Able to tolerate surgery	Decompression + stabilization + SRS Stabilization (MIS/ cement-augmentation) + cEBRT
		Unstable	Unable to tolerate surgery	cEBRT

Table 63.2 The SINS by Fisher et al. with SINS 0–6 points stable, SINS 7–12 points potentially unstable, and SINS 13–18 points defined as unstable lesions [3]

Element of the SINS	Characteristics	Score
Location	Junctional (C0–C2; C7–Th2; Th 11–L1; L5–S1)	3
	Mobile (C3–C6; L2–L4)	2
	Semirigid (Th 3–Th 10)	1
	Rigid (S2–S5)	0
Mechanical pain	Yes	3
	No (occasional nonmechanical)	1
	Pain free lesion	0
Bone lesion	Lytic	2
	Mixed	1
	Blastic	0
Alignment	Subluxation/translation	4
	De novo deformity	2
	Normal	0
Vertebral body involvement	>50% collapse	3
	<50% collapse	2
	No collapse with >50% vertebral body involvement	1
	None of the above	0
Posterolateral involvement	Bilateral	3
	Unilateral	1
	None	0

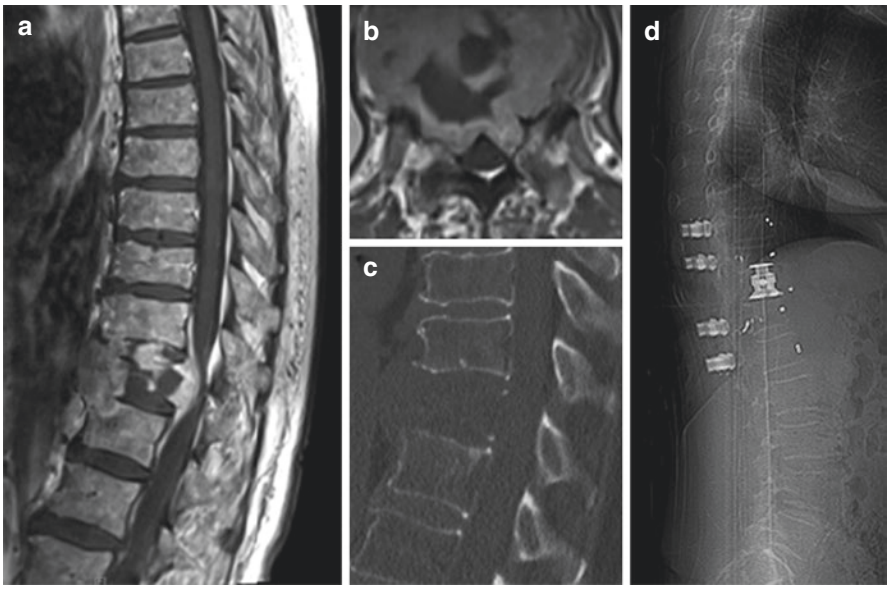

Fig. 63.1 Example of a 67-year-old female suffering from metastatic renal cell carcinoma with osteolytic spinal metastasis at Th11, clinically presenting with pain and ataxia. Following angiographic embolization of the metastasis, 360° decompression and stabilization were performed via vertebral body replacement Th11 with cage implantation and posterior stabilization with navigated pedicle screw placement Th9–L1 using PEEK carbon pedicle screws for better MRI follow-up and adjuvant radiation planning. Preoperative images **a**–**c**, postoperative image **d**

spine are employed, bone allograft, autograft, and graft substitutes are available. Hardware with low artifact potential (carbon fiber, polyetheretherketone PEEK) should be used to facilitate planning for adjuvant radiation and MRI follow-up.

Adjuvant treatment following surgical therapy is essential for local tumor control. The standard therapy is conventional external beam radiation therapy (cEBRT). Advances in technology have brought other techniques, such as stereotactic radiosurgery (SRS) into the field, with the advantage of a higher biologically effective dose (BED) on the tumor field while reducing the dose delivered to tissues surrounding the radiation site. Moreover, SRS has shown to overcome the radioresistance of some tumor types. In selected palliative cases, cEBRT for pain control alone can be evaluated. Systemic tumor therapy should be planned in interdisciplinary oncological settings according to the guidelines of the underlying malignancies.

63.7 Expected Outcomes

With the occurrence of spinal metastases in advanced malignant disease, the therapeutic setting is mostly palliative. Primary goals of surgery are neurological stabilization, pain control, spinal stabilization, and local tumor control. Thanks to evolving oncologic therapies like immunotherapy, the statistical life expectancy of patients suffering from metastatic disease are increasing. Patients suffering from spinal metastases present a very heterogenic group. Overall, the prognosis is limited and dependent on systemic metastases, underlying malignancy, previous therapies, clinical status, and comorbidities.

63.8 Potential Complications

The most dreaded complication of spinal metastases is MESCC. Fast-developing neurological deficits decrease statistic life expectancy and HRQOL for the patient. The perioperative complication rate in surgery of spinal metastases is high. Most complications arise from surgical site infections or systemic infections and cardiovascular complications, increasing patient morbidity and mortality. In case of surgical instrumentation, material displacement or dislocation and hardware failure during follow-up is possible, potentially necessitating revision surgery. Local tumor recurrence is common, and adjuvant radiation and frequent follow-up imaging is therefore necessary.

63.9 What Should Patient and Family Know?

Spinal metastases frequently occur in advanced malignant disease. Mostly dreaded is MESCC, leading to fast-developing neurological deficits and a decreased statistic life expectancy. Fast diagnosis and interdisciplinary decision-making in the

treatment of spinal metastases are essential. Therapy options have evolved towards an interdisciplinary concept including surgery, adjuvant radiation therapy, SRS, and systemic treatment according to the underlying malignancy, with a trend towards prolonged overall survival, thanks to evolving oncologic therapy options.

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Tuberculosis of Spine

64

Yat-Wa Wong

64.1 Definition

Tuberculosis of the spine (TB spine) is caused by *Mycobacterium* species, of which *Mycobacterium tuberculosis* (*M. tuberculosis*) is the most common. Respiratory and genitourinary systems are frequent primary foci. The TB infections spread to the spine through hematogenous or lymphatic routes. In contrast to pyogenic infections (bacteria destroy local tissues by proteolytic enzymes), tuberculosis infection induces type IV delayed hypersensitivity reaction which is a cell-mediated response. Macrophages engulf *M. tuberculosis* bacilli and are subsequently activated to recruit monocytes which differentiate into epithelioid cells and then Langerhans giant cells. The resulting granulomatous inflammation causes local tissue destruction, caseous necrosis, and cold abscess formation. TB spine originates from pulmonary or renal tuberculosis as a secondary infection.

64.2 Natural History

Most patients present with a chronic course with an insidious onset of pain and constitutional symptoms. Vertebral column destruction, cold abscesses, and debris compressing on the spinal cord or nerves, pachymeningitis, and meningomyelitis may cause neurological deterioration. The cold abscesses may find their ways to be

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discharged such as to the paraspinal muscles or externally through sinus tracts. Without effective treatment, TB spine tends to affect multiple vertebral levels and results in severe kyphosis. In adults, the severity of kyphosis relates directly to the severity of vertebral body destruction. In children, kyphosis may deteriorate during their growth even the TB spine is adequately treated.

Pott's paraplegia refers to the neurological deterioration related to the TB spine. It may occur at the acute phase (early onset) or many years after apparent quiescence (late onset) due to reactivation of tuberculosis or spinal cord atrophy as a result of chronic stretching over the internal kyphosis, healed bony bars, calcified caseous material, fibrosis, increasing kyphosis, or instability.

64.3 Physical Examination

Depending on the stage and extensiveness of the disease, the physical examination may reveal local tenderness, paraspinal swelling indicating cold abscess, discharging sinus, sinus scars, sharp kyphosis (gibbus), or neurological deficit (Video 64.4).

64.3.1 Imaging

There is no pathognomonic radiological feature to distinguish TB spine from other spinal infections (Chap. 65). The initial radiographic finding is osteopenia due to the chronic nature of the disease. Later, fusiform paravertebral soft tissue swelling and psoas shadow may cold abscesses (Fig. 64.1). Reduction of the intervertebral disc height and destruction of the adjacent vertebral bodies are similar to pyogenic spondylodiscitis. However, it is relatively more common for TB spine to have vertebral body destruction without affecting the intervertebral disc and multiple levels of involvement due to the affinity of the bacteria to the well-perfused tissues such as bone marrow (Fig. 64.2). Plain radiographs are useful to detect instability and assess spinal alignment. Computed tomography (CT) scan or magnetic resonance imaging may show elevation of the anterior or posterior longitudinal ligament with subligamental spread of abscesses. Scalloping of the anterior or posterior walls of the vertebral bodies is a common feature.

64.4 Differential Diagnosis

Neoplasms or other spinal infections are common differential diagnoses. There is no clinical, serological, and radiological feature that is diagnostic of TB spine. Biopsy under fluoroscopic, CT, or ultrasonic guidance is frequently required to confirm the diagnosis and guide antituberculous drug treatment (Videos 64.7 and 64.8).

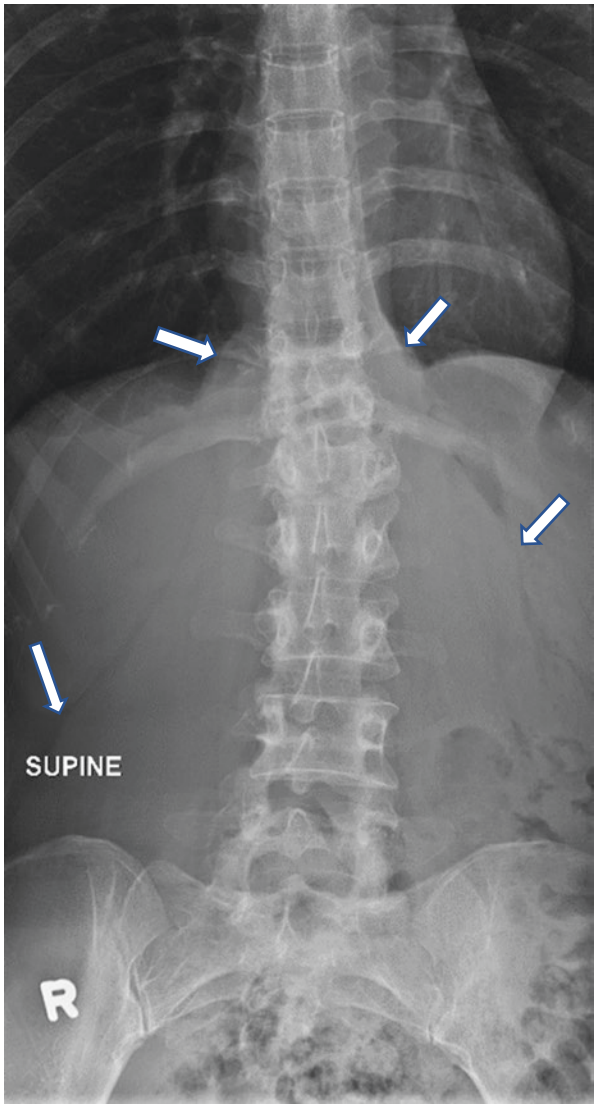

Fig. 64.1 Anteroposterior radiograph showing the fusiform paravertebral soft tissue swelling and bilateral psoas shadow enlargement due to TB cold abscesses (and bilateral psoas shadow **enlargement** due to TB cold abscesses)

It is diagnostic of TB spine if biopsy specimens show mycobacterium bacilli with Ziehl-Neelsen staining, positive TB polymerase chain reaction, or typical histological features of epithelioid granuloma, Langerhans multinucleated giant cells, and caseation. Identification of mycobacterium bacilli is only possible in about 50% of cases, and it takes 6 to 8 weeks to get the culture and sensitivity results.

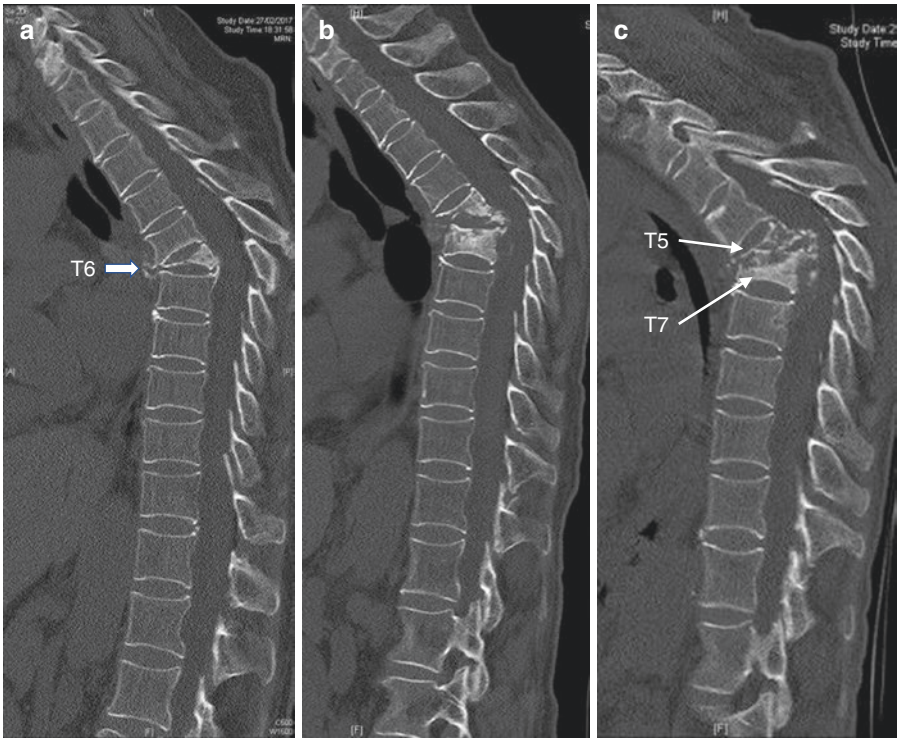

Fig. 64.2 (**a**–**c**) Sagittal reconstructed CT scans showing the progressive anterior column destruction. (**a**) Only T6 was infected by TB and collapsed. (**b**) T5–T7 collapsed 1 year later. (**c**) T5–T7 collapsed further with increasing kyphosis in 2 years

64.5 Treatment Options

Most TB spine can be successfully treated by nonoperative treatment. The first-line anti-TB treatment at author's region is isoniazid 300 mg oral daily 9 to 12 months; rifampicin 450 mg (body weight < 50 kg) or 600 mg (body weight ≥ 50 kg) oral daily for 9 to 12 months; pyrazinamide 25–30 mg/kg/day oral for 2 months; and ethambutol 15 mg/kg/day oral for 2 months. Apart from regular monitoring of complete blood count, liver function and renal function, clinicians should look for specific side effects such as peripheral neuropathy (isoniazid), high serum urate (pyrazinamide), and decreased vision (ethambutol). Apart from regular monitoring of complete blood count, liver function and renal function for active TB spine, surgery is indicated if there are significant neurological deficits, severe spinal deformity, huge cold abscess not responding to anti-TB drugs and percutaneous drainage, or intractable mechanical pain not responding to nonoperative treatment. Surgical options include anterior radical debridement and strut graft fusion (Fig. 64.3, Hong Kong operation), posterior decompression and instrumented fusion with or without

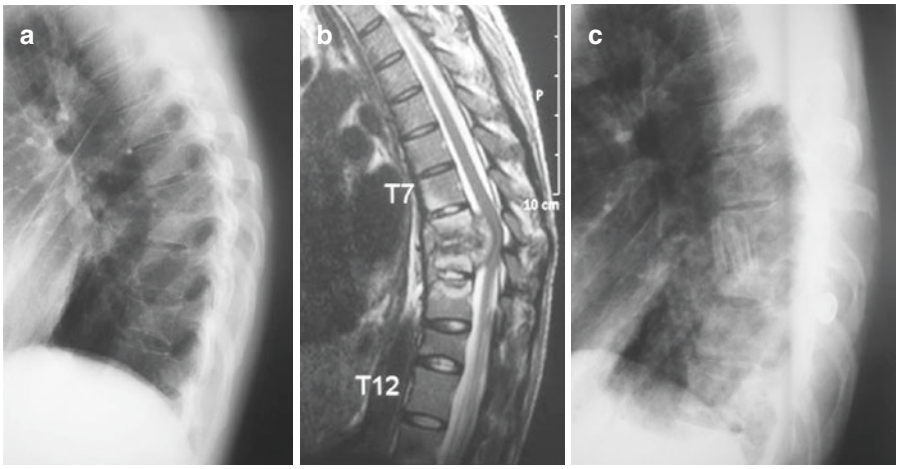

Fig. 64.3 (**a**–**c**) This T8–T9 TB spine patient received anterior debridement and fusion using autogenous rib grafts through left thoracotomy. (**a**) Lateral thoracic spine radiograph. (**b**) T2-weighted sagittal MRI. (**c**) Postoperative lateral thoracic radiograph

anterior column debridement and support, and combined anterior and posterior surgery (Fig. 64.4). The choice is based on the location, extensiveness, indications of the surgery, and expertise. Application of metallic implants is safe provided that adequate anti-TB treatment is given (Fig. 64.5). Instrumentation is generally indicated for the more extensive disease to restore spinal stability and alignment. Combined approaches are the preferred option if the disease is extensive with multilevel involvement.

Patients may develop severe kyphosis in the TB spine even they have good functional status. Apart from cosmetic issues, it may compromise cardiopulmonary function, limit the abdominal volume, and cause pain by rib on iliac crest impingement. Deformity correction is feasible in selected cases. Preoperative halo traction may help the correction. Pedicle subtraction osteotomy or vertebral column resection (VCR) is the surgical options depending on the severity of the kyphosis. To avoid excessive shortening or lengthening of the spinal cord, VCR is usually followed by lengthening of the anterior column and shortening of the posterior column during the kyphosis correction.

For Pott's paraplegia of late onset, it is sometimes associated with severe rigid kyphosis, compromised cardiopulmonary function, myelomalacia, and spinal cord atrophy. Correction of severe rigid kyphosis is difficult if not impossible for older patients, but adequate decompression can be achieved through costotransversectomy approach with or without fusion. Prognosis of neurologic recovery is poor even with adequate spinal cord decompression and deformity correction if there is associated spinal cord atrophy. Most of these patients acquire TB spine in childhood, and their deformities progress during growth. Early stabilization and deformity correction may prevent late-onset paraplegia.

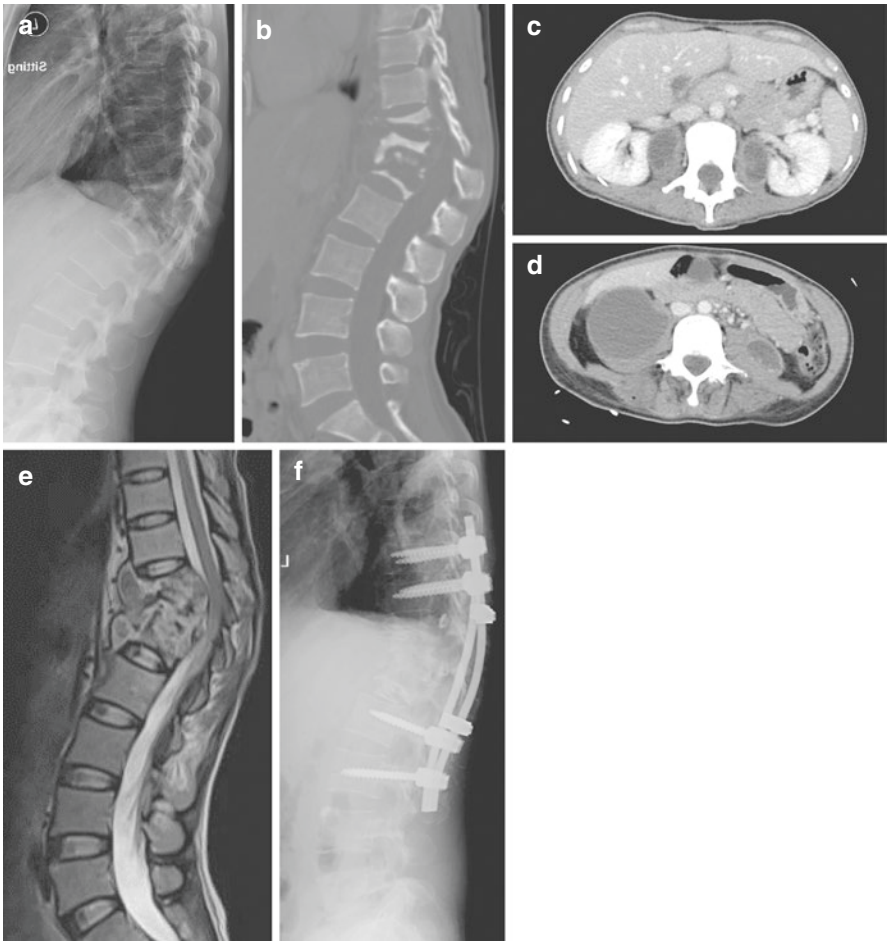

Fig. 64.4 This patient suffered from tuberculous thoracolumbar spine affecting multiple levels and bilateral psoas abscesses. (**a**) Lateral standing radiograph showing the thoracolumbar kyphosis. (**b**) Sagittal reconstructed CT spine demonstrating the multilevel involvement. (**c**, **d**) Axial CT scans showing the bilateral psoas abscesses. (**e**) T2-weighted sagittal MRI demonstrating the thoracolumbar TB spine causing conus compression. (**f**) Lateral postoperative standing radiograph after anterior debridement, drainage of abscesses, anterior fusion using rib grafts, and posterior instrumented fusion

64.6 Expected Outcomes

Most patients respond to anti-TB drug treatment alone with a favorable outcome. Spinal decompression and stabilization give favorable results for patients with neurological deficits in acute disease. The surgical difficulty and risk increase with increasing deformity, chronicity, and extensiveness of diseases. Pott's paraplegia of late onset carries a poor prognosis for neurologic recovery.

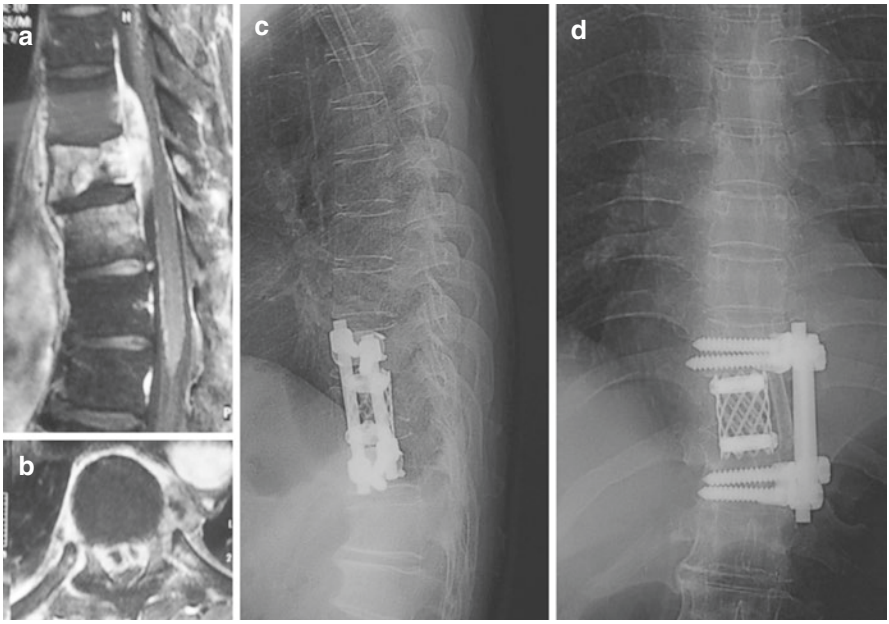

Fig. 64.5 (**a**, **b**). T1-weighted sagittal and axial contrast MRI showing the subligamental spread of abscess in TB spine. (**c**, **d**) Standing lateral and anteroposterior radiographs after anterior radical debridement, fusion by titanium mesh cage packed with autogenous bone graft, and anterior instrumentation. There is no contraindication for applying metallic implants in TB

Prevention of severe kyphosis and nonunion reduces the risk of late-onset neurological deterioration.

64.7 Potential Complications

Severe complications include death due to disseminated tuberculosis, paraplegia, angular kyphosis (Appendix Q), and persistent discharging sinuses.

64.8 What Should Patient and Family Know?

Compliance with anti-TB drug treatment is the key of successful treatment.

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Pyogenic Spondylodiscitis

65

Yat-Wa Wong

65.1 Definition

Pyogenic spondylodiscitis is the bacterial infection of the intervertebral disc and its adjacent vertebral bodies. It is the most common form of spinal infection. Other forms of spinal infection include spondylitis (infection of the vertebral body), discitis (infection of the intervertebral disc alone), epidural abscess, and facet joint septic arthritis. Spinal infection can also be classified according to the causative microorganisms. They are bacterial (pyogenic), mycobacterial (granulomatous), fungal, or parasitic.

Hematogenous spread is the most common route of transmission in pyogenic spondylodiscitis. Bacteria seed on the bony end plate and migrate into the intervertebral disc causing infection. Skin infection, dental caries, genitourinary, and gastrointestinal tract infections are potential sources.

65.2 Natural History

Most patients respond to appropriate antibiotics if they are treated early. Diagnostic and treatment delays are potentially devastating. Death due to systemic sepsis, neurological deterioration, spinal instability, and deformity are possible consequences. Risk is higher for immunocompromised or debilitated patients.

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65.3 Physical Examination

The most common presentation is pain (Chaps. 11 and 41), and if there is any one of the following additional features, it should raise the suspicion:

1. Fever of unknown source.
2. Intravenous drug abusers or immunocompromised patients.
3. Elevation of infective markers such as CRP, ESR procalcitonin, and WBC.
4. Radiological features of pyogenic spondylodiscitis.

Pain at rest is the typical complaint of pyogenic spondylodiscitis. In the presence of fever and back pain, spondylodiscitis should be excluded. There may be local tenderness at the affected segment. Deformity if any is usually not severe especially in early cases. Detailed neurological assessment is mandatory (Video 65.4). The severe neurological deficit at presentation or deterioration is an indication for surgery. Any skin lesions or other septic foci should be treated accordingly. At least two sets of blood cultures (especially when patients have a fever, or chill and rigor) and one midstream urine for urinalysis/culture should be done. Other specimens (sputum, wound swaps) for culture should also be obtained, if clinically appropriate. Prolonged incubation during culture is needed if low virulence pathogens such as *Propionibacterium* and diphtheroids are suspected. Brucella serology must be checked if there is history of exposure such as farm visit, raw milk, or placenta consumption. Elevation of procalcitonin suggests pyogenic rather than tuberculosis (TB) infection.

65.4 Imaging

Spine radiographs may show decreased intervertebral disc height with adjacent bony end plate erosion (Fig. 65.1). Bony destruction is not apparent in the first or second week. The only radiological features may be soft tissue edema and loss of spinal sagittal alignment due to muscle spasms. In chronic infection, radiographs may show osteosclerosis and deformity. Clinical diagnosis is usually not difficult with compatible clinical, serological, and radiographic features. Magnetic resonance imaging is the most useful tool for early diagnosis before radiographic features develop. It also delineates the extent of the disease for surgical planning.

Blood culture can only catch less than 50% of the causative bacteria. Computed tomography scan or fluoroscopy-guided biopsy (Videos 65.7 and 65.8) is frequently necessary to get the bacteriological diagnosis. If there is a paraspinal abscess, percutaneous drainage can be done at the same time of the biopsy. Specimens should be sent for aerobic and anaerobic culture, AFB smear and culture, TB PCR, fungal culture, and histological examination. Special culture may be necessary if rare organisms are suspected.

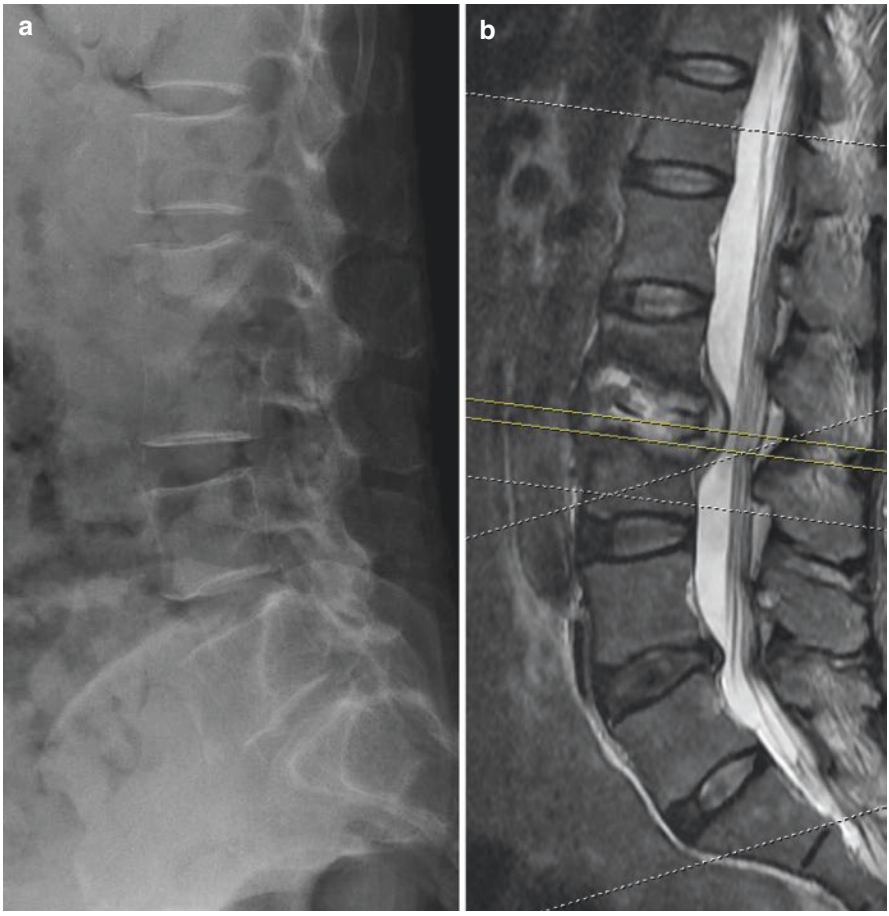

Fig. 65.1 (**a**, **b**) Showing the typical radiological features of destruction of intervertebral disc space and adjacent vertebral bodies in spondylodiscitis

65.5 Differential Diagnosis

Spinal tumors, both primary and secondary, tend to affect the vertebral bodies alone without intervertebral disc involvement. TB is a great mimicker that can present with either spondylitis or spondylodiscitis (Chap. 64). The lymphocyte monocyte ratio is a cheap and good indicator for differential diagnosis of TB spine and pyogenic spondylodiscitis. The ratio is typically elevated in specific infections like TB.

65.6 Treatment Options

Most patients respond well to antibiotic treatment, and surgery is not required for most patients. Antibiotics are withheld until the bacteriological diagnosis is confirmed. However, empirical antibiotics can be started early after essential diagnostic workups especially if the patients are septic looking and ill. For community-acquired infection, cloxacillin 2 g IV every 6 h will cover the most common organisms such as methicillin-sensitive staphylococci and streptococci. For hospital-acquired infection or immunocompromised patients, ceftriaxone 1–2 g IV daily plus vancomycin 15–20 mg/kg/dose IV every 12 h provides broader coverage including Gram-negative bacilli and MRSA. The subsequent antibiotic regime is adjusted according to the culture result. Duration of antibiotic treatment varies but normally ranges from 3 to 12 weeks depending on the clinical, serological, and radiological responses. Typically, antibiotics can be stopped if patients' symptoms improve; infective markers return to normal; radiographs showed evidence of stopping new vertebral destruction. A regular blood examination is essential, not just to check infective markers but also to monitor patients' liver and renal function because of the prolonged antibiotic treatment. Like other infections, spondylodiscitis is catabolic, and adequate nutritional support promotes early recovery. Spondylodiscitis may be very painful. Adequate symptomatic treatments including analgesics, corset or collar, and appropriate rest relieve patients' suffering.

The sizable paraspinal abscess should be drained, preferably image-guided percutaneous drainage. Loculated abscess or thick pus may need open drainage. Surgery is generally indicated if there is neurological deterioration, persistent instability, unacceptable deformity, debilitating pain not under control by nonoperative treatment, an uncertain diagnosis, and unfavorable response to medical treatment. The aims of treatment are to decompress the spinal cord or nerves, debride necrotic tissues, drain abscesses, correct deformity, and stabilize the spine.

Anterior or anterolateral approach accesses the infective focus directly and provides wide exposure for thorough debridement and spinal canal decompression. Reconstruction can be done by autologous iliac crest tricortical, rib, or fibular graft. Alternatively, titanium cage or mesh cage filled with a bone graft can be used. Anterior instrumentation at the cervical and thoracic spine provides additional stability, while anterior fixation at the lumbar spine is less secure, and additional posterior instrumentation may be necessary (Fig. 65.2).

The posterior approach alone is effective for decompression of the spinal canal and allows rigid instrumented fusion (Fig. 65.3). It is particularly useful for the upper thoracic spine since the anterior approach there is difficult (may need split manubrium approach). However, surgeons cannot normally debride the infective focus as thoroughly as the anterior approach.

The choice of surgical approaches is dictated by surgeons' expertise and the indications for surgery. Regardless of the approaches, metallic cages and implants can be safely used to reconstruct the spine if patients are covered by adequate antibiotics and the surgical bed is relatively clean. PEEK cages should be avoided because it is more difficult to eradicate the bacteria on the PEEK surface.

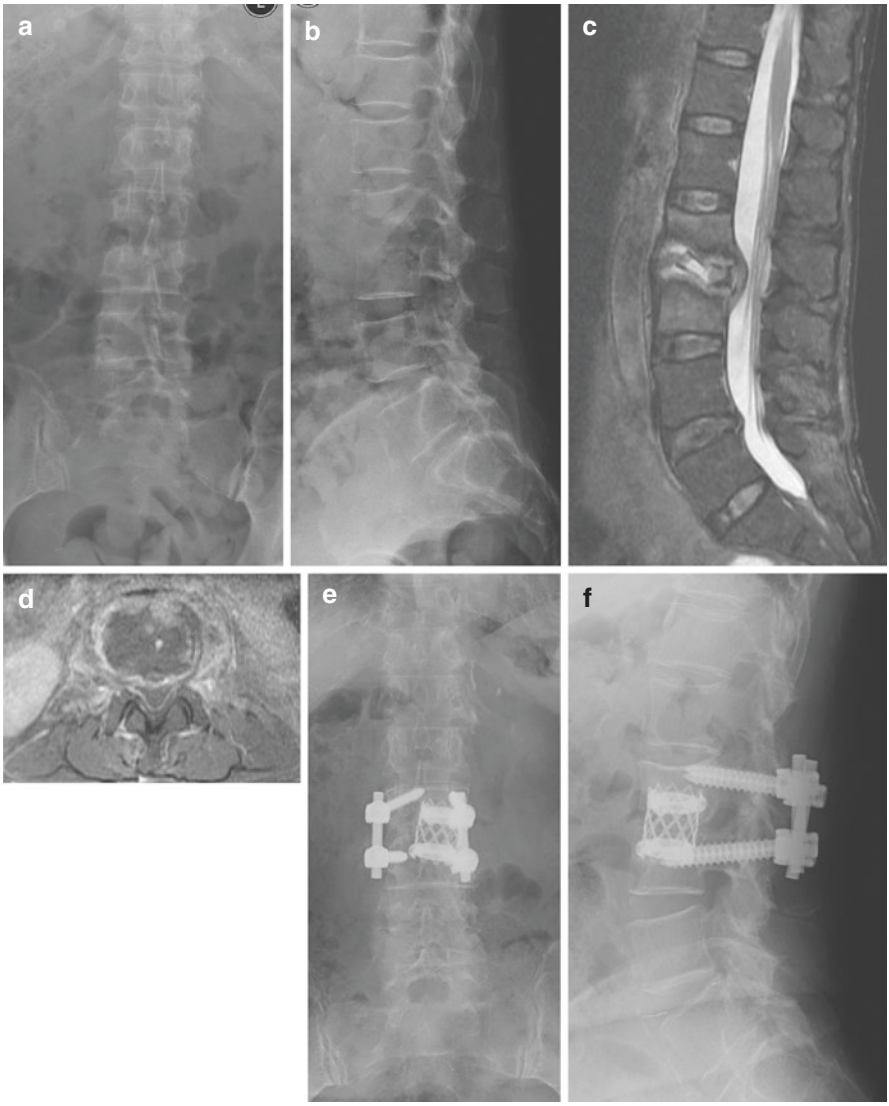

Fig. 65.2 (**a**, **b**) Anteroposterior and lateral radiographs showing the L2/3 spondylodiscitis. (**c**, **d**) T2-weighted sagittal and T1-weighted contrast axial MRI demonstrating the active infection. Patient did not respond to empirical antibiotics after negative biopsy. (**e**, **f**) Radiographs after anterior debridement and fusion followed by posterior stabilization

65.7 Expected Outcomes

Most patients can recover completely with early treatment. The mortality and morbidity rates become higher if the patient has many comorbidities.

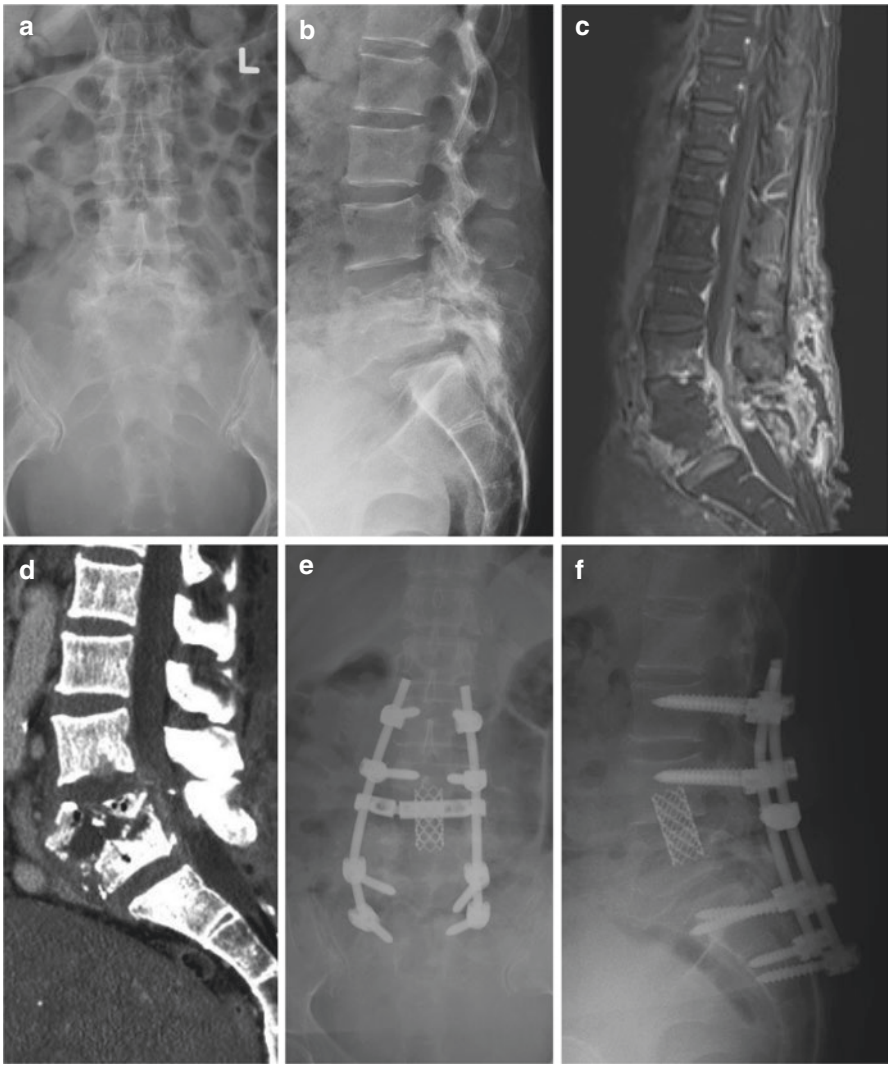

Fig. 65.3 (**a**, **b**) Radiographs showing L4/L5 spondylodiscitis. (**c**) T1-weighted contrast sagittal MRI demonstrating the extensiveness of the infection. (**d**) Sagittal CT scan illustrating L4 vertebral body fragmentation. Due to neurological deterioration and poor response to antibiotics, anterior debridement, fusion, and posterior fixation via posterior approach alone were done. (**e**, **d**) Postoperative radiographs

65.8 Potential Complications

- Uncontrolled sepsis leading to mortality.
- Neurological deterioration.
- Spinal instability (Chap. 47).

- Kyphosis.
- Secondary degeneration and chronic pain (Chap. 41).

65.9 What Should Patient and Family Know?

Prolonged antibiotic treatment and close monitoring of treatment responses are necessary.

Further Reading

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Postsurgical Spinal Infection

66

Alain Dimeglio and Federico Canavese

66.1 Definition

According to the surgical site and the time since surgery, postoperative spine infections can be divided into (a) superficial or deep and (b) acute (early-onset) or chronic (late-onset). It is also important to distinguish between infections occurring in adult and pediatric patients.

Superficial infections typically develop above the fascial layer and involve subcutaneous tissues and skin; on the other hand, deep infections do extend below the fascial layer (lumbodorsal fascia or ligamentum nuchae for posterior surgery and abdominal fascia or platysma for anterior surgery).

Acute infections are usually diagnosed within 3 to 4 weeks of the procedure, while chronic infections are diagnosed more than 4 weeks since surgery; for some authors, the cutoff between acute and chronic infection is 6 weeks. However, it is important to recognize the very late infection (sometimes years after the index procedure) usually secondary to low-grade pathogens such as *Propionibacterium* species and that can be cured by hardware removal.

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66.2 Natural History

The incidence of postoperative spine infection is highly variable; it ranges from 0% to 18% depending on the type of surgery (surgeries without bone grafting or instrumentation have the lowest rate of infection) and of surgical approach (posterior cervical fusion > posterior lumbar surgery > anterior surgery).

Overall, the development of a postoperative infection put the patient at increased risk for pseudoarthrosis, chronic pain, adverse neurological sequelae, return to the operating room, worsened long-term outcomes, and – in most severe cases – even death.

66.3 Physical Examination

The typical physical signs of surgical site infection are pain, erythema, swelling of the incision or wound dehiscence, and purulent drainage from the wound; importantly, wound drainage for more than 1 week is a risk factor for deep infection. Other signs and symptoms are fever (present in about half of the patients), fatigue, and, in some cases, weight loss (depending on the chronicity of the infectious process).

Laboratory Tests: If surgical site infection is suspected, white blood count (WBC), erythrocyte sedimentation rate (ESR), and C-reactive protein (CRP) should be requested by the treating surgeon. However, used in isolation, WBC is a poor marker, and ESR is a nonspecific marker—though more sensitive than WBC—to rule out a surgical site infection. CRP is the most sensitive indicator currently available to diagnose postoperative infection. The combination of CRP and ESR values is the most predictive method for diagnosing and monitoring treatment response of postoperative spinal infections; however, no laboratory method has demonstrated excellent specificity/positive predictive value.

The use of procalcitonin (PCT) for the evaluation of spinal infection has shown lower sensitivity than CRP. Other relatively novel markers such as serum amyloid A (SAA) and presepsin, although promising, have limited clinical validation and require to be studied further.

66.4 Imaging

Plain radiographs are of limited use in postoperative spine infection although they should be obtained to assess for any hardware failure.

Computed tomography (CT) scan is the imaging of choice to evaluate bone, and it also provides information on soft tissue collections. Early bony changes include erosion and destructive changes at the level of the end plates and disk space narrowing. CT can identify (earlier than radiographs) lucencies around orthopedic implants.

Magnetic resonance imaging (MRI) with gadolinium contrast is considered to be the most sensitive modality for the assessment of postoperative infection. MRI

findings usually associated with postoperative spine infection are rim-enhancing fluid collections, ascending epidural collections, evidence of bony destruction, and progressive marrow signal changes; the presence of hardware can create artifacts making the interpretation of the images more difficult.

Nuclear medicine (gallium-67 or technetium-99) can be used as an adjunct for the diagnosis of postoperative spine infection although it has limited sensitivity and is not used regularly in the diagnostic process.

66.5 Differential Diagnosis

Currently, the gold standard for the diagnosis of postoperative spine infection is a positive deep culture. Differential diagnosis focuses on the type of pathogen involved in the infectious process (Videos 66.7 and 66.8).

66.6 Treatment Options

Multiple debridement procedures of necrotic and infected tissue and long-term antibiotics are required for treatment. Exploring below the fascia is recommended for all but the most superficial infections. Bone graft that is loose at the time of debridement should be removed, but any graft material that is adherent to bony structures should be left in place; similarly, all necrotic, infected, and foreign material, such as sutures, must be completely debrided.

For early postoperative infection (acute or chronic <3 months), in cases where spinal instrumentation is present, the current recommendation is not to remove the hardware to avoid destabilizing the spine. For late postoperative infection (chronic >3 months), if fusion has occurred, hardware removal is usually needed to allow adequate debridement of the wound. The vacuum-assisted closure (VAC) system can be used in patients with acute infection; it has been shown to be a useful tool in the armamentarium of the spinal surgeon dealing with patients susceptible to deep postoperative infections, especially those with neuromuscular diseases (Fig. 66.1). VAC system is changed every 48 to 72 h.

Equally important to multiple debridements and lavages is antibiotic therapy. Importantly, antibiotics should not be administered prior to culture results; if the patient is septic or unstable, antibiotics should be administered empirically to help prevent further clinical decline. Broad-spectrum antibiotics should be initiated prior to obtaining final culture results and adjusted depending on the results of the cultures. Although the duration of antibiotic therapy is controversial, short antibiotic course should be reserved only for patients without any hardware in place. For patients with deep infection and hardware in place, the length of antibiotic treatment is much longer, including 4 to 6 weeks of IV antibiotics followed by at least 4 to 6 weeks of oral antibiotics. Postoperative discitis/osteomyelitis is generally treated with >3 months of antibiotics depending on the inflammatory markers (Fig. 66.2).

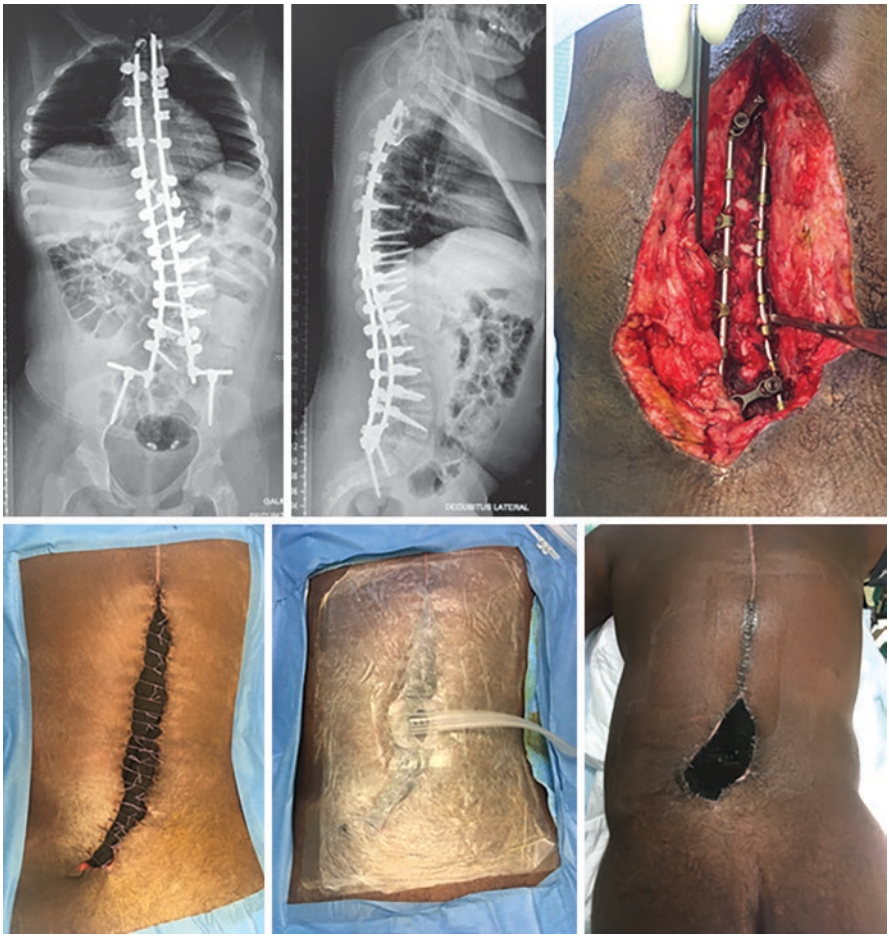

Fig. 66.1 Deep postoperative infection in a patient with spinal muscular atrophy. Use of VAC system. Hardware is left in place, and superficial and deep layers are progressively closed

66.7 Expected Outcomes

A high index of suspicion is needed to make an early diagnosis; if treated promptly through debridement and lavage in association with targeted antibiotic therapy (according to deep culture results), the outcome is generally good. Older age, presence of comorbidities, smoking history, and obesity can be associated with poorer outcomes (Table 66.1).

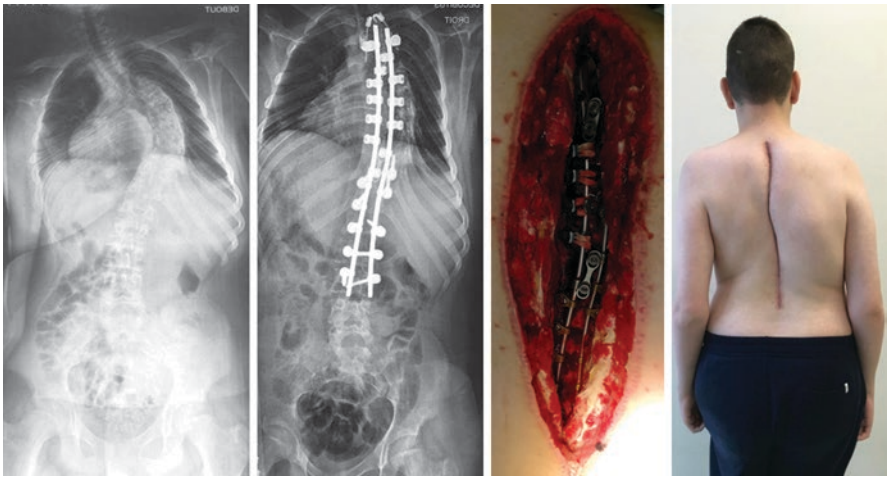

Fig. 66.2 Deep postoperative infection in a patient with severe deformity. Throughout debridement and lavage. Skin closure

Table 66.1 Risk factors

Patient related	Comorbidities	Past history	Surgery related
Older age	Diabetes	Prolonged hospitalization	Increased blood loss
Obesity	Urinary incontinence	Prior infection	Use of blood transfusion
Poor nutritional status	Complete neurological deficit	Prior spine surgery	Prolonged surgical time
Alcoholism	>3 comorbid diseases	Surgery after spine trauma	Posterior approach > anterior
Tobacco		Tumor surgery (resection)	Use of grafting and instrumentation
		Radiation therapy	Fusion extending to the sacrum
			Spinal fluid leak

66.8 Potential Complications

Potential complications are pseudoarthrosis, chronic pain, adverse neurological sequelae, return to the operating room, worsened long-term outcomes, and—in most severe cases—even death; in case of deformity surgery, the removal of hardware may cause the recurrence of the deformity (if fusion has not occurred).

66.9 What Should Patient and Family Know?

Before each surgery, patients and their families must be informed of the infectious risks, their treatment modality, and possibly short-, medium-, and long-term complications.

Further Readings

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Appendix A: Classification of Upper Cervical Injuries

There have been many classification systems for different injury patterns. AO Spine has introduced as part of the comprehensive classification system of all spine injuries a simple and unified classification of injuries to the region between occiput and C2–C3 junction.

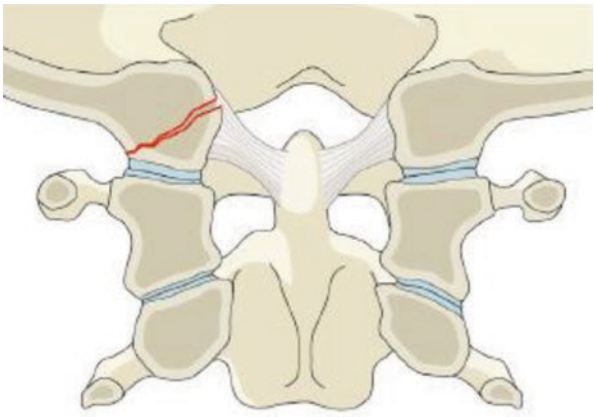
AO Spine Upper Cervical Classification System:

Three parts of this anatomical area:

1. Occipital condyle and occipitocervical junction
2. C1 ring and C1–C2 joint
3. C2 and C2–C3 joint

1.1 Occipital Condyle and Occipitocervical Junction

Type A: Simple or comminuted occipital condyle injuries



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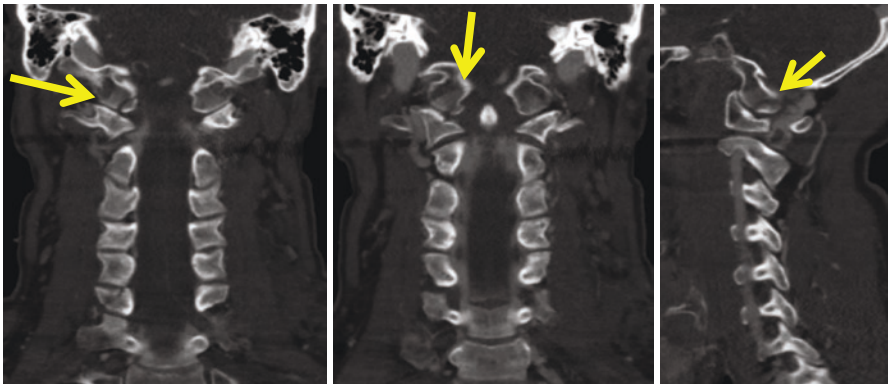
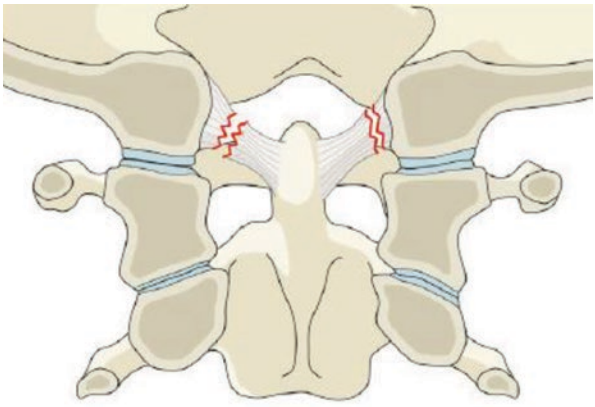
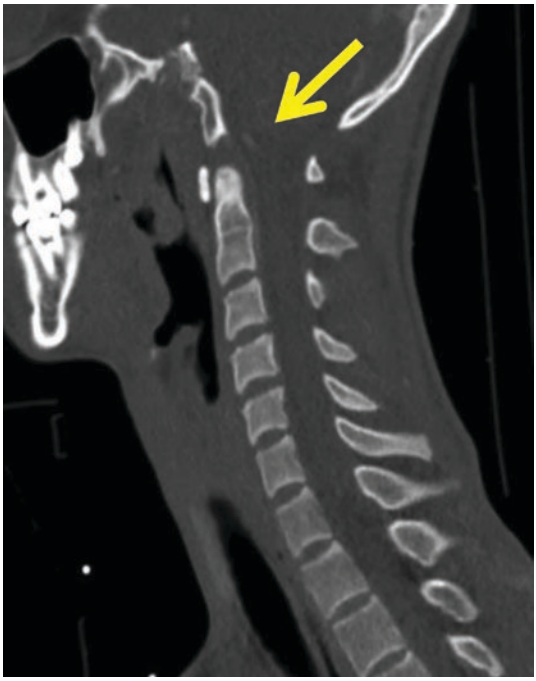
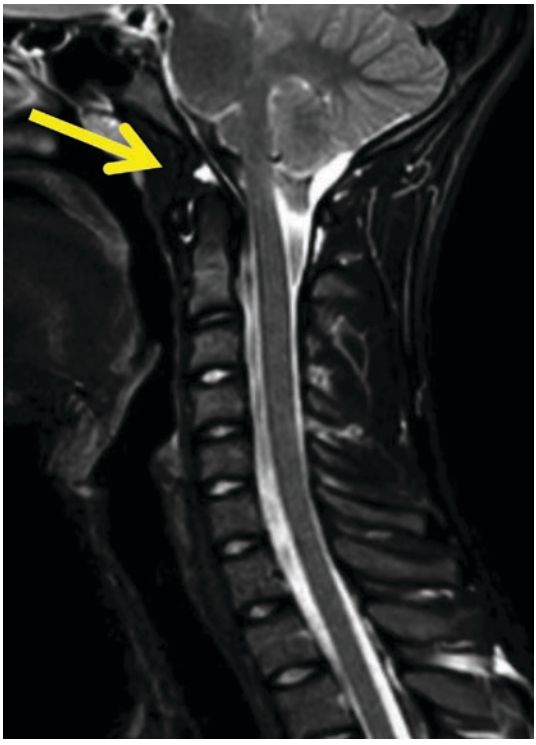


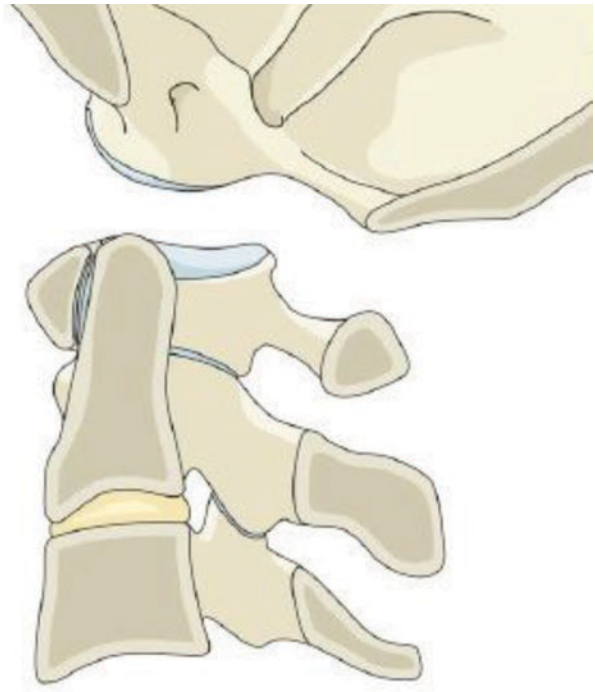
Type B: Nondisplaced ligamentary injury

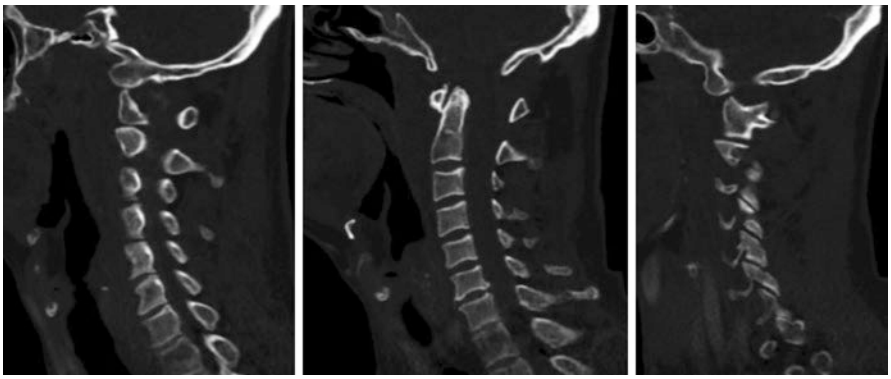






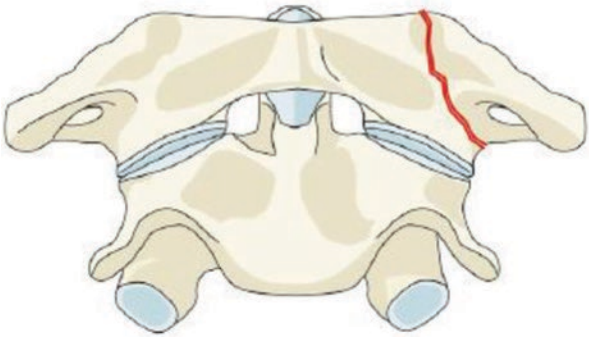
Type C: OC dislocation

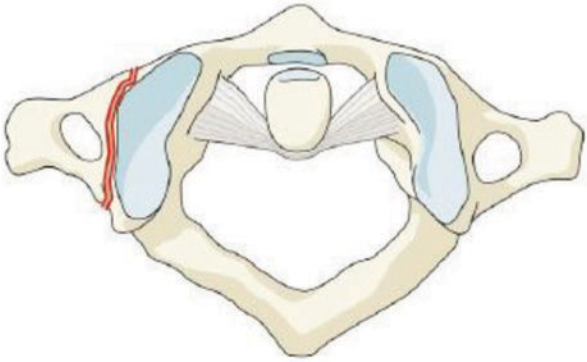


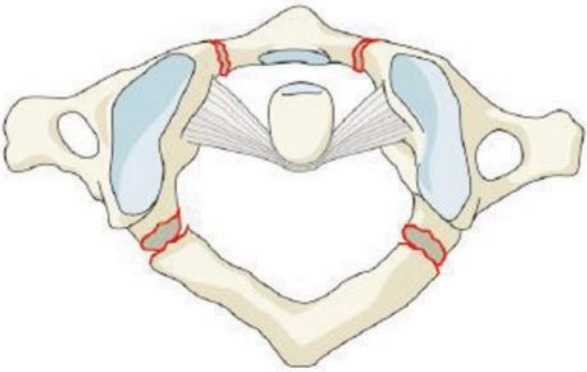


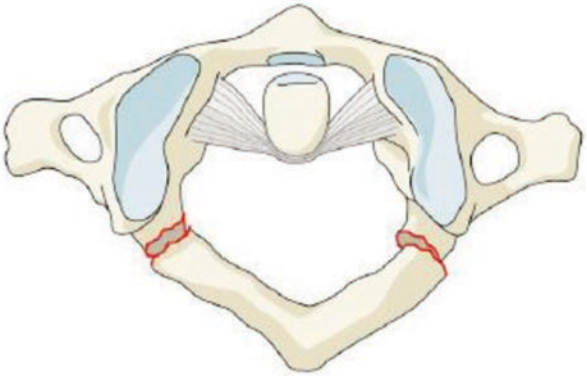
1.2 C1 Ring and C1–C2 Joint

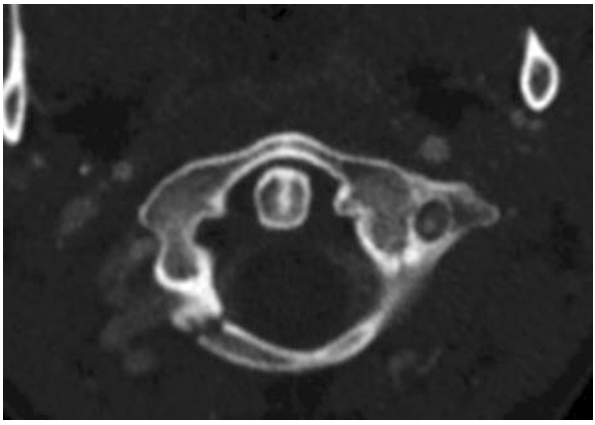
Type A: Isolated bony arch of C1:



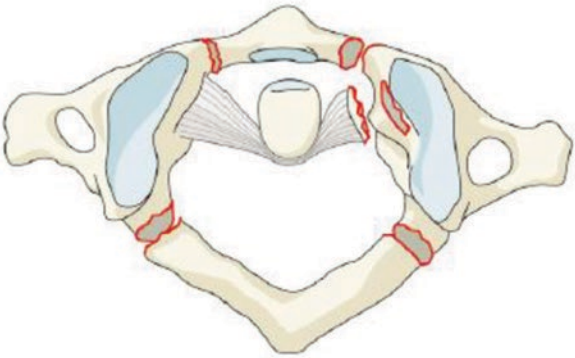


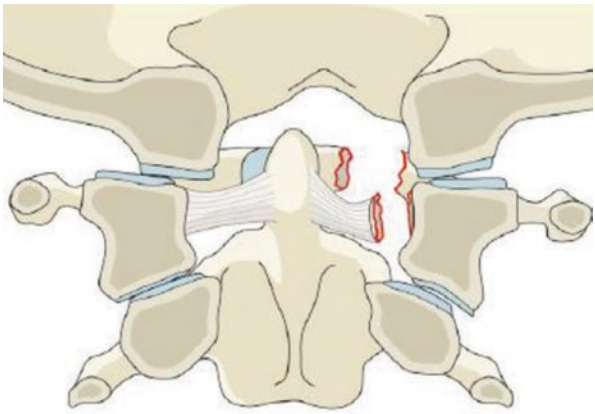


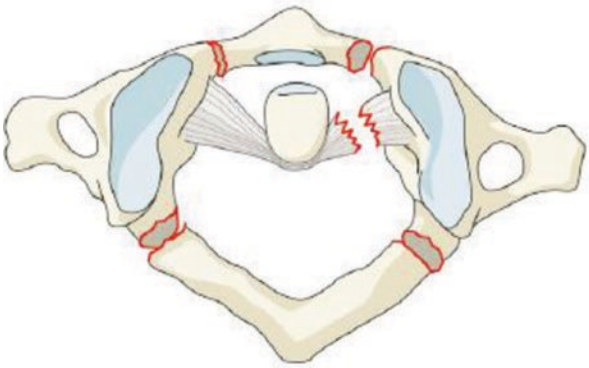


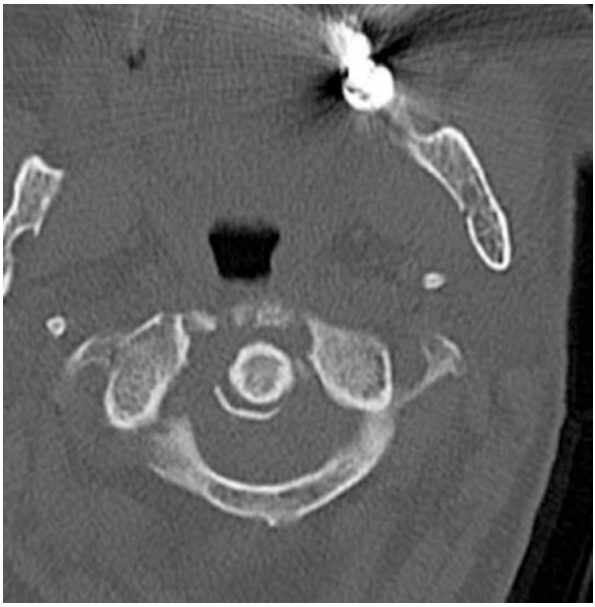


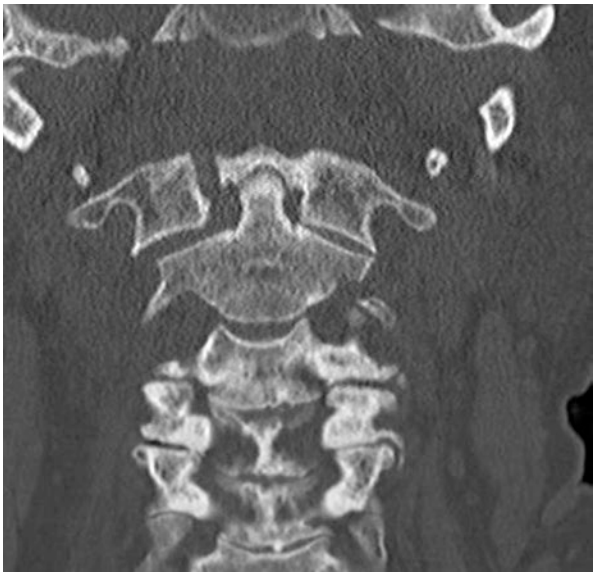
Type B: With transverse atlantal ligament (TAL) injury



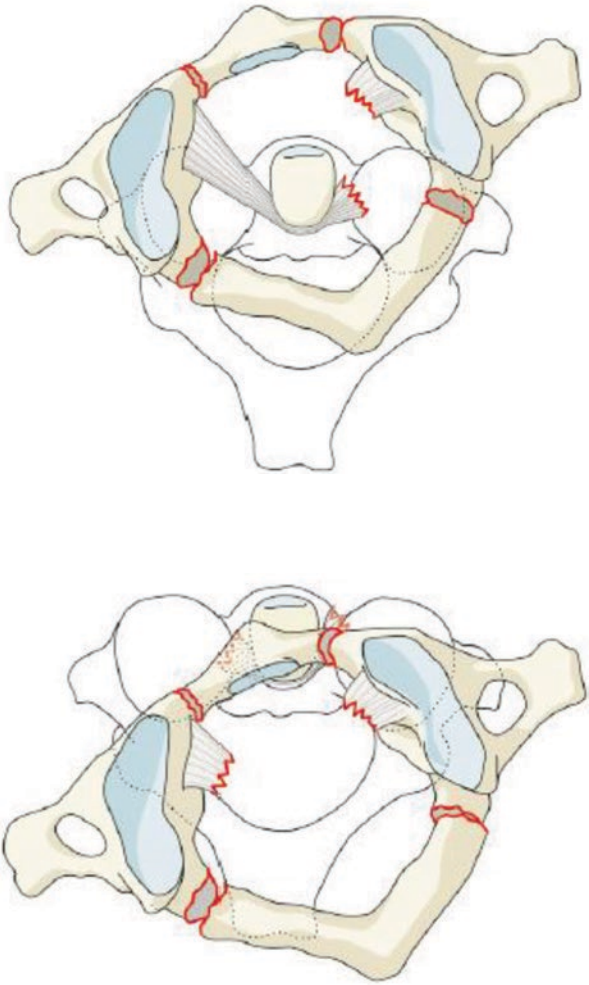


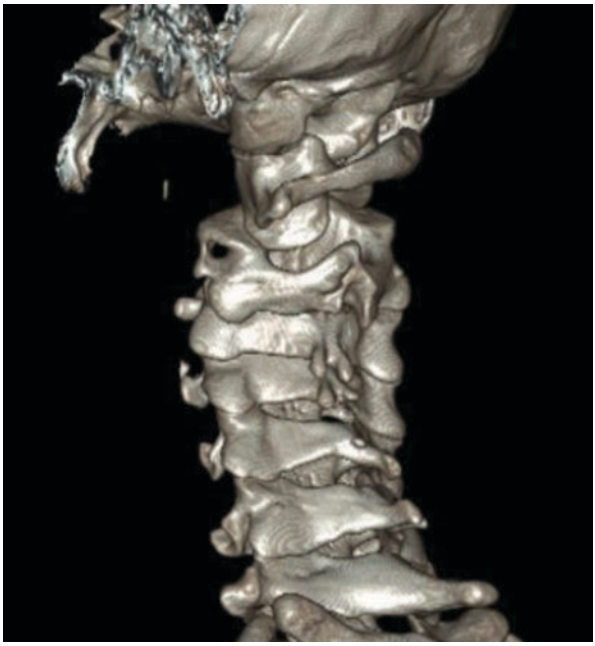


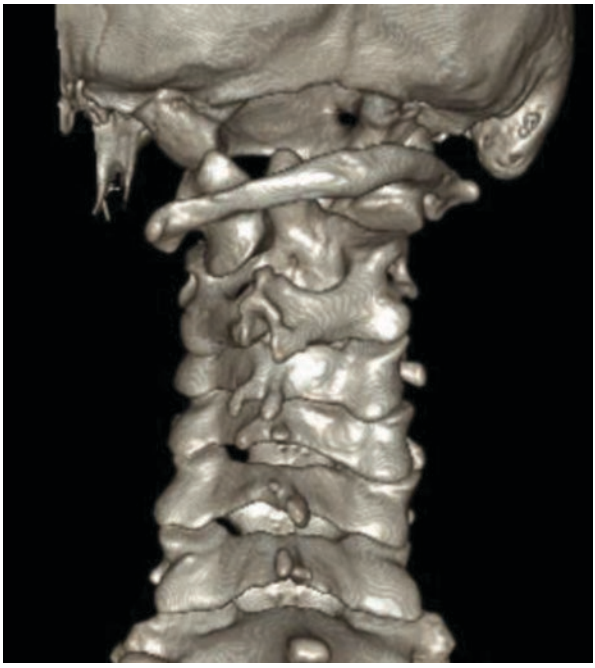


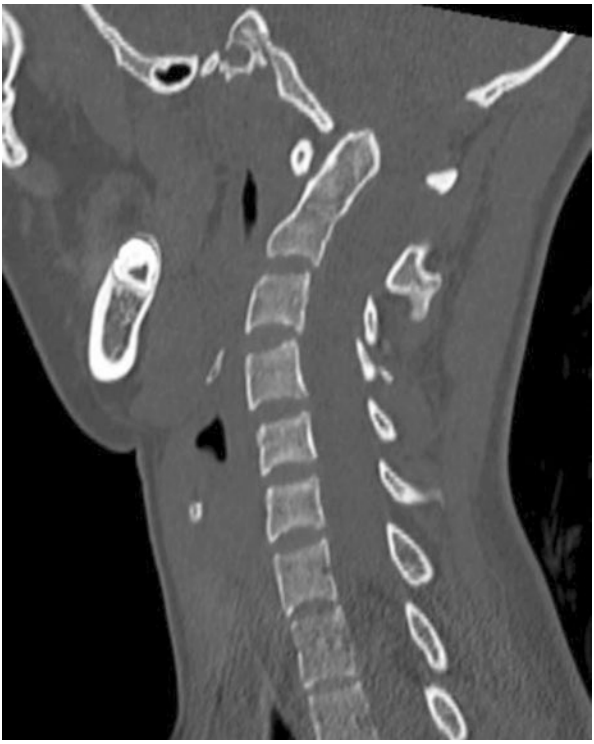


Type C: Atlantoaxial dislocation



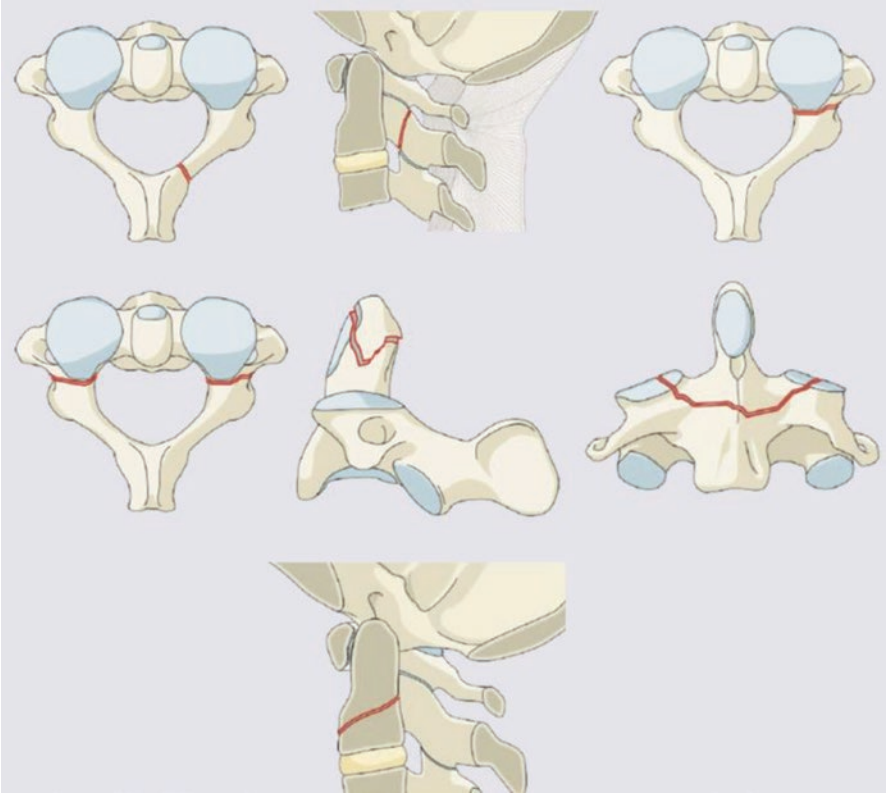


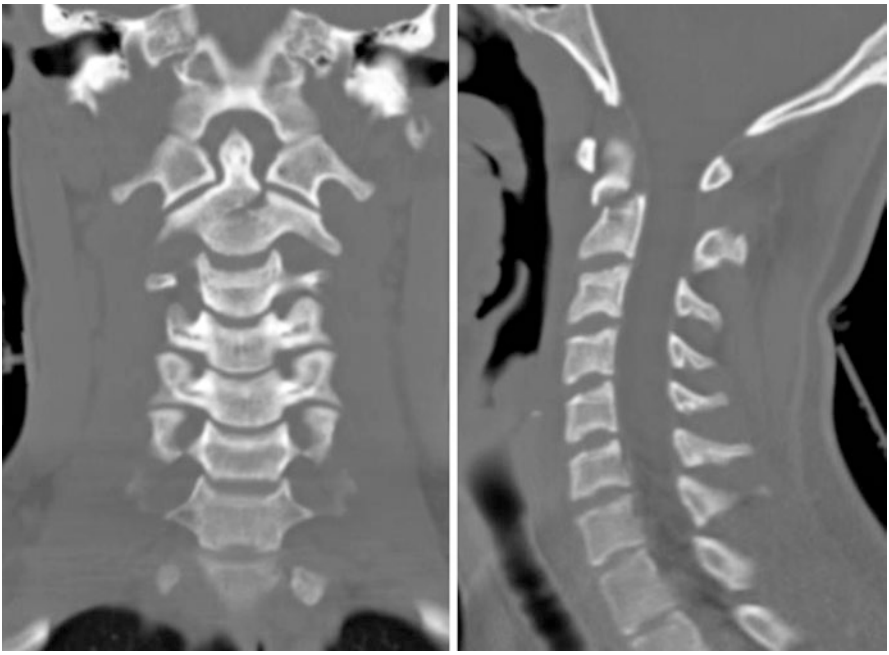




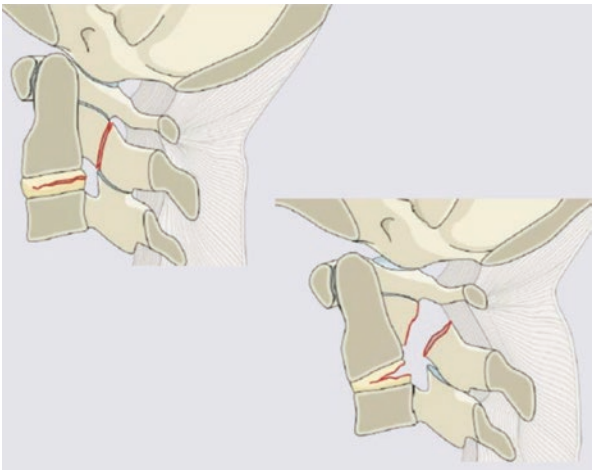
1.3 C2 and C2–C3

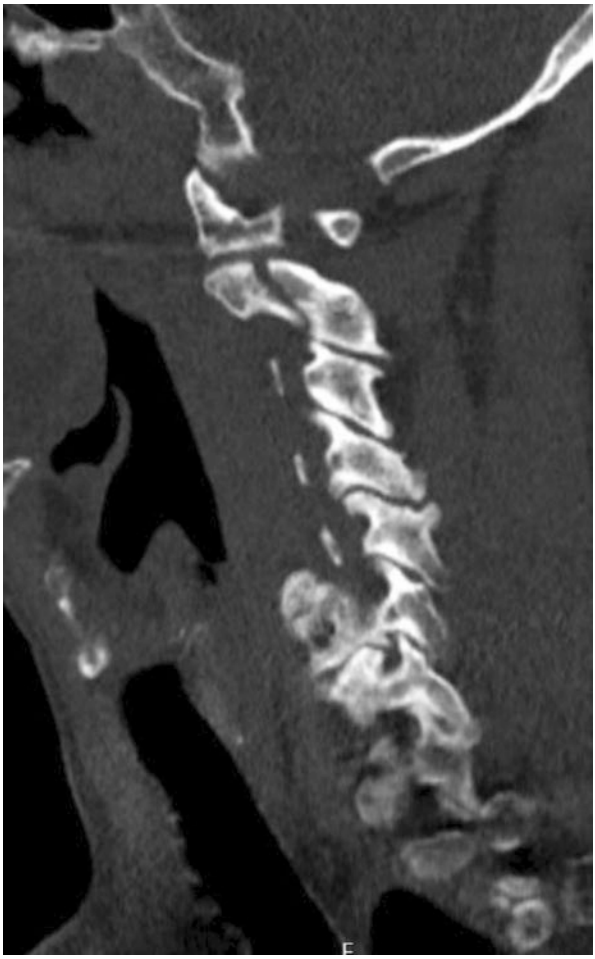
Type A: Bony injury without ligamentous, tension band or discal injury



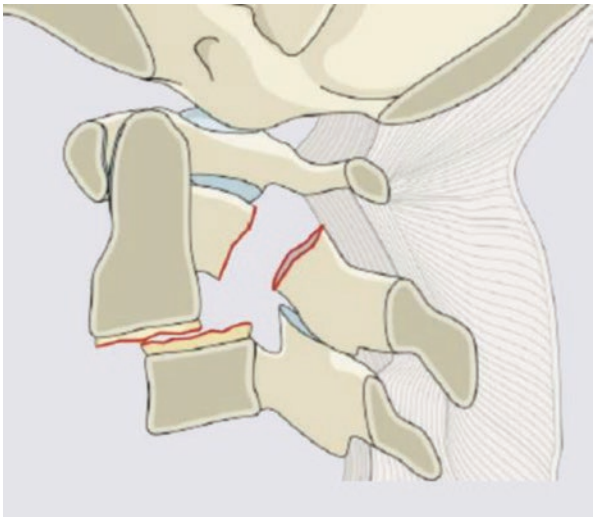


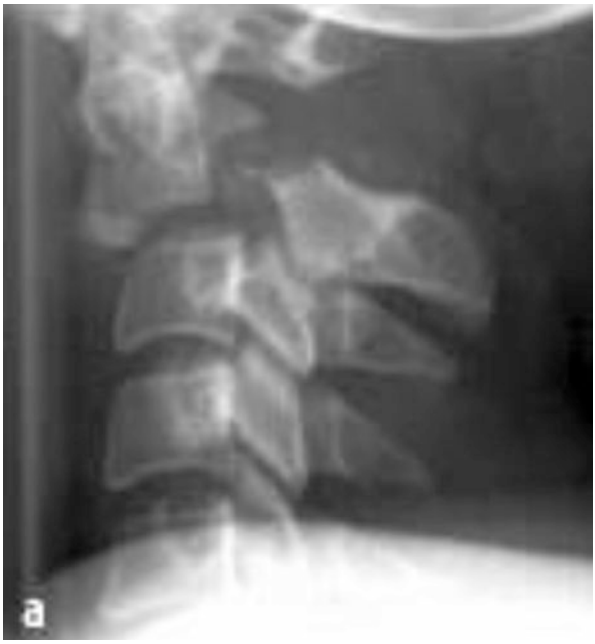
Type B: Tension band injury

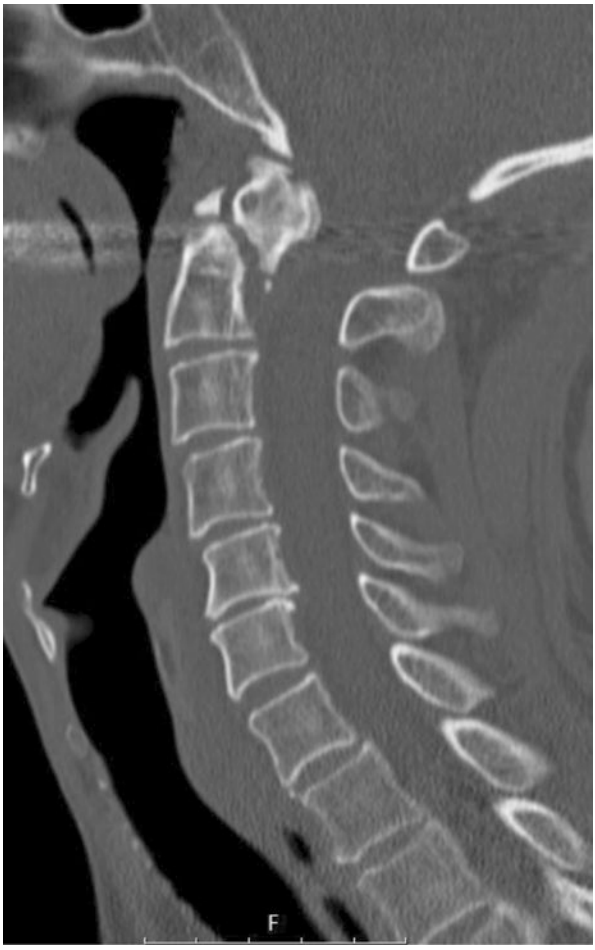




Type C: Any injury leading to translation in any plane



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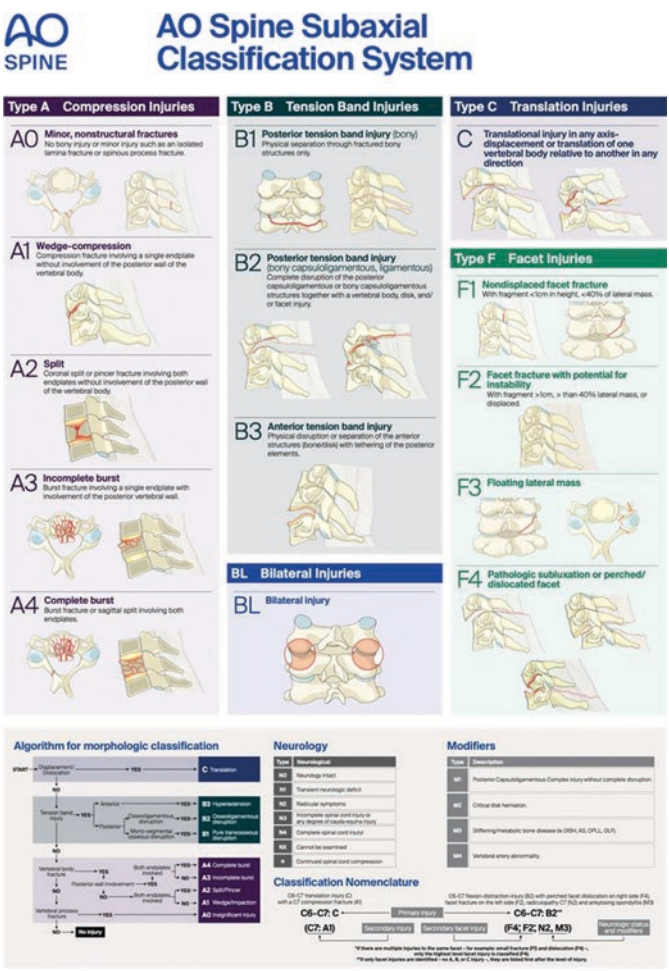
Appendix B: Subaxial Cervical Spine Classification

https://surgeryreference.aofoundation.org/spine/trauma



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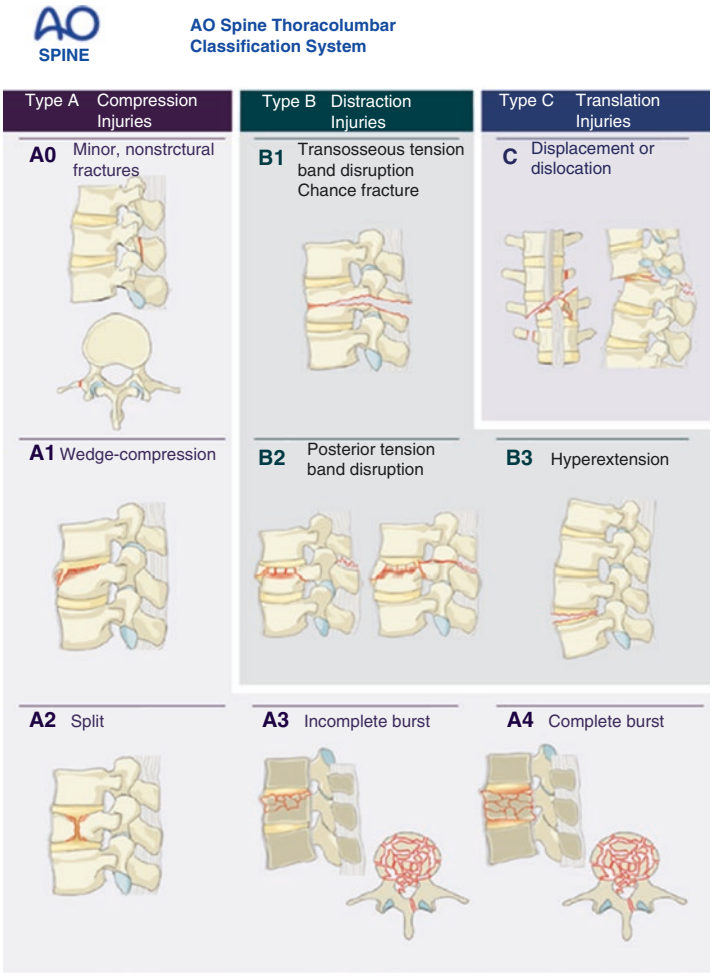
Appendix C: AO Spine Thoracolumbar Fracture Classification



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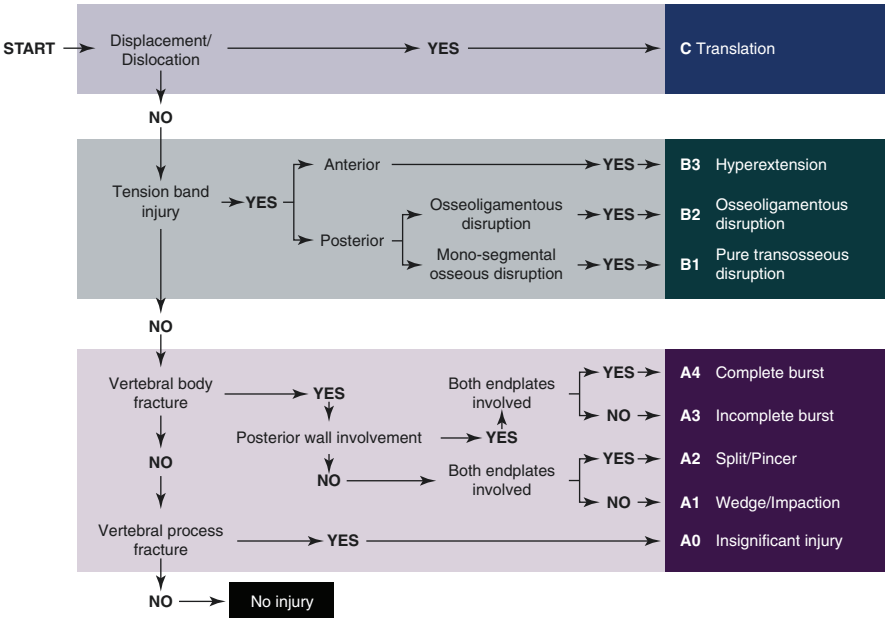


Neurology

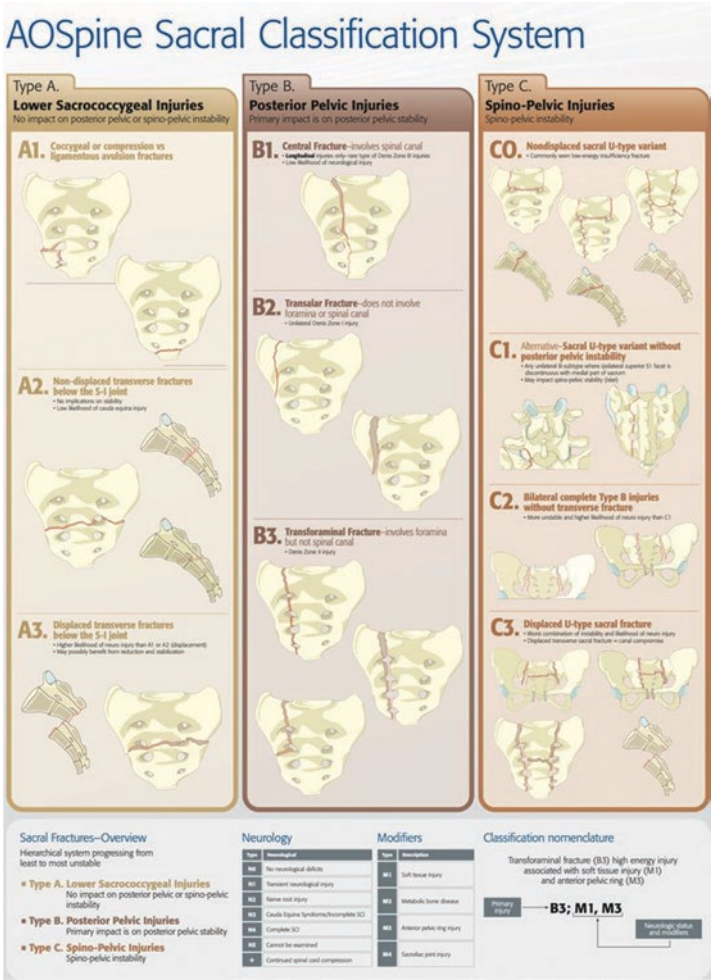
Type	Neurological
N0	Neurology intact
N1	Transient neurolocic deficit
N2	Radicular symptoms
N3	Incomplete spinal cord injury or any degree of cauda equina injury
N4	Complete spinal cord injuryl
NX	Cannot be examined
+	Continued spinal cord compression

Modifiers

Type	Description
M1	This modifier is used to designate fractures with an indeterminate injury to the tension band based on spinal imaging with or without MRI. This modifier is important for designating those injuries with stable injuries from a bony standpoint for which ligamentous insufficiency may help determine whether operative stabilization is a consideration.
M2	Is used to designate a patient-specific comorbidity, which might argue either for or against surgery for patients with relative surgical indications, Examples of an M2 modifier include ankylosing spondylitis or burns affecting the skin overlying the injured spine.

https://aospine.aofoundation.org/clinical-library-and-tools/ao-spine-classification-systems

Appendix D: Sacral Fractures (AO Spine Classification System)



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Appendix E: Osteoporotic Vertebra Fracture Classification

Definition: The Spine Section of the German Society for Orthopaedics and Trauma (DGOU) described a classification for osteoporotic thoracolumbar fractures.
Content: The classification includes five groups: OF 1 no vertebral deformation (vertebral edema); OF 2, deformation with no or minor (<1/5) involvement of the posterior wall; OF 3, deformation with distinct involvement (>1/5) of the posterior wall; OF 4, loss of integrity of the vertebral frame or vertebral body collapse or pincer-type fracture; OF 5, injuries with distraction or rotation (Table E.1 and Fig. E.1).

Table E.1 Detailed explanation of the subgroups in the classification

OF 1	No vertebral deformation (vertebral body edema in MRI-STIR only). This type is rare. The stable injury is clearly visible on MRI-STIR sequence only. X-rays and CT scans do not show vertebral deformation.
OF 2	Deformation with no or only minor involvement of the posterior wall (<1/5). This type of fracture affects one endplate only (impression fracture). OF 2 are stable injuries.
OF 3	Deformation with distinct involvement of the posterior wall (>1/5). This type of fracture affects one endplate only but shows distinct involvement of the anterior and posterior wall (incomplete burst fracture). The fracture can be unstable and may collapse further over time.
OF 4	Loss of integrity of the vertebral frame structure, or vertebral body collapse, or pincer-type fracture. Both endplates and posterior wall are involved (complete burst fracture). A vertebral body collapse is typically seen as a final consequence of a failed conservative treatment and can impose as a vertebra plana. Pincer-type fractures involve both endplates and may lead to severe deformity of the vertebral body. OF 4 are unstable fractures and intravertebral vacuum clefts are often visible.
OF 5	Injuries with distraction or rotation. This group is rare but shows substantial instability. The injury includes both anterior and posterior columns consisting of ligamentous structures. OF 5 injuries can be caused either by trauma directly or by ongoing collapsing of an OF 4.

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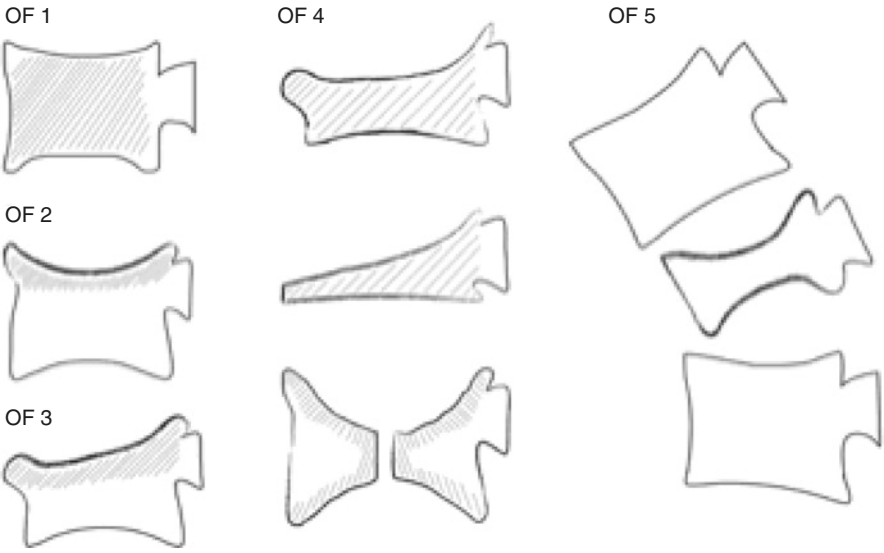

Fig. E.1 Representation of subgroups

Clinical Application: Types OF 1 and OF 2 can be treated conservatively whereas types OF 4 and OF 5 are good candidates of surgical treatment. Type OF 3 may be treated either surgically or conservatively.

Further Reading

1. Schnake K, Blattert T, Hahn P, et al. Classification of osteoporotic thoracolumbar spine fractures: recommendations of the spine section of the German Society for Orthopaedics and Trauma (DGOU). Global Spine J. 2018;8(2 Suppl):46S–49S.

Appendix F: Glasgow Coma Scale

Definition: The Glasgow Coma Scale (GCS) is a widely accepted neurological scale that is used to measure a patient's level of neurological status. Additionally, it eases communication between health professionals regarding their patients' neurological situation.
Content: The GCS is composed of three layers. Eye, verbal, and motor responses of a patient either to pain or verbal commands are recorded (Table F.1). It can range from 3 points to 15 points.

Table F.1 Glasgow Coma Scale, response, and score

Glasgow Coma Scale		
	Response	Score
Eye opening response	Spontaneous	4
	To speech	3
	To pain	2
	None	1
Best verbal response	Orientated	5
	Confused	4
	Inappropriate	3
	Incomprehensible	2
	None	1
Best motor response	Obeying	6
	Localizing	5
	Flexing	4
	Abnormal flexion (decorticate)	3
	Abnormal extension (decerebrate)	2
	None	1

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Clinical Application: Although it was designed for and best used after head trauma, today it is one of the main scales used to evaluate a patient with any kind of cranial neurological insult.

Further Reading

1. Teasdale G, Jennett B. Assessment of coma and impaired consciousness. A practical scale. Lancet. 1974;2(7872):81–4.

Appendix G: ASIA Scale

Definition: Based on the sensorimotor scores, the level and the severity of the spinal cord injury can be determined. The scale most commonly used to classify the severity of the injury is the American Spinal Injury Association (ASIA)/International Spinal Cord Society (ISCoS) neurological standard scale (AIS), better known as the ASIA Impairment Scale.

Content: The AIS is a standardized examination consisting of a myotomal based motor examination, dermatomal based sensory examination, and an anorectal examination. Based on the findings of these examinations, an injury severity or grade and level are assigned (Fig. G.1). The AIS further classifies injuries as a complete or incomplete spinal cord injury (Table G.1).

Clinical Application: The purpose of the AIS is to standardize careful, detailed documentation of an injury; guide further radiographic assessment and treatment; and determine whether injuries are complete or incomplete, an important and sometimes subtle neurologic distinction that has tremendous prognostic implications.

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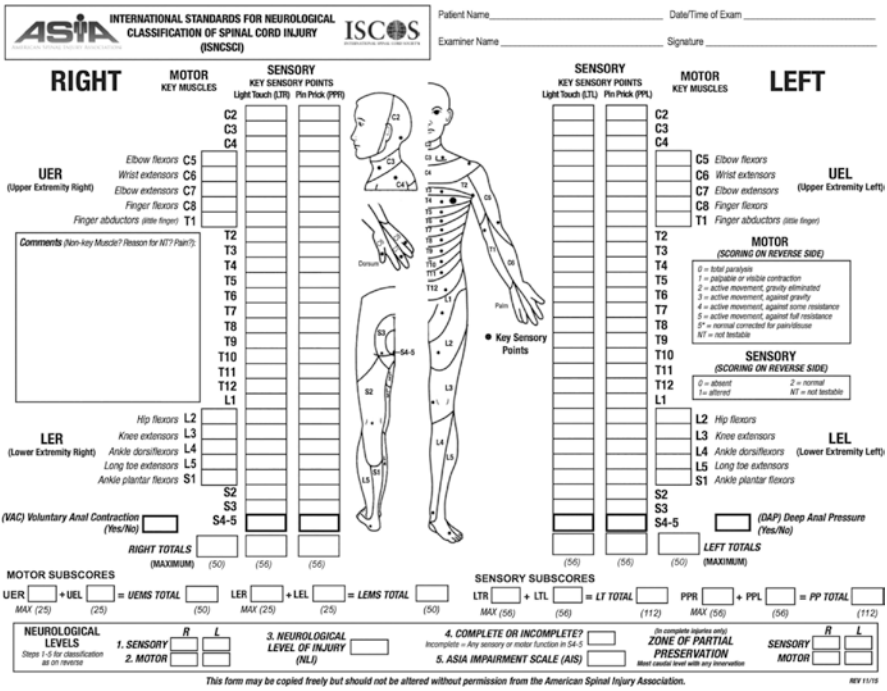

Fig. G.1 The American Spinal Injury Association International Standards for Neurological Classification of Spinal Cord Injury form

Table G.1 The American Spinal Injury Association/International Spinal Cord Society Neurological Standard Scale (better known as the "ASIA Impairment Scale")

ASIA Impairment Scale		Lesion
A	No motor or sensory function is preserved in the sacral segments S4–S5	Complete
B	Sensory but not motor function is preserved below the neurological level and includes the sacral segments S4–S5	Incomplete
C	Motor function is preserved below the neurological level, and more than half of the key muscles below the neurological level have a muscle grade less than 3	Incomplete
D	Motor function is preserved below the neurological level, and at least half of the key muscles below the neurological level have a muscle grade of 3 or more	Incomplete
E	Motor and sensory functions are normal	Normal

Further Readings

1. American Spinal Injury Association. International standards for neurological classification of spinal cord injury, 2002 revision. Chicago, IL: American Spinal Injury Association; 2002.
2. Roberts TT, et al. Classifications in brief: American Spinal Injury Association (ASIA) Impairment Scale. Clin Orthop Relat Res. 2017;475(5):1499–504.
3. van Middendorp JJ, et al. Diagnosis and prognosis of traumatic spinal cord injury. Global Spine J. 2011;1(1):1–8.

Appendix H: King et al. Classification System of Adolescent Idiopathic Scoliosis

Definition: King et al.'s classification identifies five types of curve patterns in adolescent idiopathic scoliosis and it helps to properly select the level of fusion and instrumentation in order to preserve motion as much as possible.

Content: King et al. introduced for the first time the concept of *stable vertebra* (the vertebra most closely bisected by the center sacral vertical line) and of *structural/compensatory curves* depending on their flexibility on side-bending films.

Clinical Relevance: This system was developed on the experience with Harrington rod instrumentation and with the advent of segmental instrumentation it failed to give accurate and reliable guidelines for choosing the proper levels for fusion.

Type I: S-shaped or double curve in which both the thoracic and lumbar curves cross the center sacral vertical line; the lumbar curve is larger and stiffer than the thoracic curve.

Treatment: Fuse down to L4 (via a posterior approach) or down to L3 (anterior approach).

Type II: S-shaped or double curve in which both the thoracic and lumbar curve cross the center sacral vertical line; the thoracic curve is larger (or equal) and stiffer than the lumbar curve; type II curves are also called "false double major."

Treatment: Often a thoracic fusion (or down to L1) is indicated.

Type III: Single (major) thoracic curve; only the thoracic curve is structural and crosses the center sacral vertical line. The lumbar curve does not cross the midline.

Treatment: Thoracic fusion.

Type IV: Long C-shaped thoracic curve in which L5 is centered over the sacrum and L4 is tilted into the thoracic curve.

Treatment: Fuse down to L4.

Type V: Double-thoracic curve; T1 tilts to the upper curve.

Treatment: Make sure that the upper (left) thoracic curve is nonstructural; if not, fuse the upper thoracic spine as well as the lower thoracic spine.

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Further Reading

1. King HA, et al. The selection of fusion levels in thoracic idiopathic scoliosis. J Bone Joint Surg Am. 1983;65:1302–13.

Appendix I: Lenke et al. Classification System of Adolescent Idiopathic Scoliosis

Definition: Lenke et al.'s classification identifies six types of curves based on the curve type, coronal lumbar modifier, and thoracic sagittal profile. Four series of plain radiographs are needed to apply Lenke et al.'s classification system: upright anteroposterior, lateral, supine right-bending, and supine left-bending.
Content: Lenke et al. introduced for the first time the concept of *coronal lumbar (A, B, C) and sagittal plane (−, N, +) modifiers* and of *main/minor curves* depending on their magnitude and flexibility on side-bending films (structural/not structural). The main curve is the largest curve; minor curves are defined as structural if coronal plane rigidity is greater than 25° upon side-bending or kyphosis greater than 20° on sagittal radiographs.
Clinical Relevance: This system aims to give accurate and reliable guidelines for choosing the proper levels for fusion. Only main curve and structural minor curves should be included in the instrumented fusion.
Type I: Main thoracic (MT)
Type II: Double thoracic (DT)
Type III: Double major (DM)
Type IV: Triple major (TM)
Type V: Thoracolumbar/lumbar (TL/L)
Type VI: Thoracolumbar/lumbar-main thoracic (TL/L-MT)

Lumbar coronal modifier: (A) the central sacral vertical line (CSVL) is between the pedicles of the apical vertebra; (B) the CSVL is between the medial border of the concave pedicle and the lateral edge of the apical vertebra; (C) the CVSL is medial to the lateral edge of the apical vertebra.

Sagittal plane modifier: Normal (N): T5–T12 kyphosis is 10°–40°; minus (−): T5–T12 kyphosis <10° (hypokyphotic); plus (+): T5–T12 kyphosis >40° (hyperkyphotic).

In total, 42 different types of curves can be identified (type 5 and type 6 curves are associated with a lumbar coronal modifier type C) (Table I.1).

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Table I.1 Lenke et al.'s classification system

Curve type		Proximal thoracic	Main thoracic	Thoracolumbar/ lumbar	Lumbar modifier	Sagittal modifier
I	MT	NS	S	NS	A, B, C	−, *N*, +
II	DT	S	S	NS		
III	DM	NS	S	S		
IV	TM	S	S	S		
V	TL/L	NS	NS	S	C	
VI	TL/L-MT	NS	S	S		

Further Reading

1. Lenke LG, Betz RR, Harms J, et al. Adolescent idiopathic scoliosis: a new classification to determine extent of spinal arthrodesis. J Bone Joint Surg Am. 2001;83:1169–81.

Appendix J: Early Onset Scoliosis Classification (C-EOS)

Definition: The Classification of Early-Onset Scoliosis (C-EOS) was developed to provide a comprehensive, practical, and predictive novel classification system designed exclusively for young patients with scoliosis.
Content: The C-EOS is based on five parameters: etiology, major curve, curve flexibility, total kyphosis, and annual progression (Table J.1).
Clinical Application: The treatment options can be classified based on the mechanism of action as distraction-based technique, guided-growth procedures, and compression-based techniques; to these methods, serial casting can be added or selected as a first-choice treatment option (Table J.2).

Table J.1 C-EOS

Etiology	Major curve (Cobb angle)	Flexibility modifier (optional)	Maximum total kyphosis	Progression modifier (optional)
Idiopathic	**1**: <20°	Flexible (**F**)	**Negative (−)**: <20°	**P0**: <10°/year
Syndromic	**2**: 21°–50°	Rigid (**R**)	**Neutral**: 21°–50°	**P1**: 10°–20°/year
Neuromuscular Low tone	**3**: 51°–90°		**Positive (+)**: >51°	**P2**: >20°/year
Neuromuscular High tone	**4**: >91°			
Congenital or *structural*				

Table J.2 Classification of EOS according to the treatment mechanism of action

Treatment: Mechanism of action			
Distraction based	*Guided growth*	*Compression based*	*Serial casting*
Growing rods	Luque trolley	Tether	
VEPTR	Shilla procedure	Staples	

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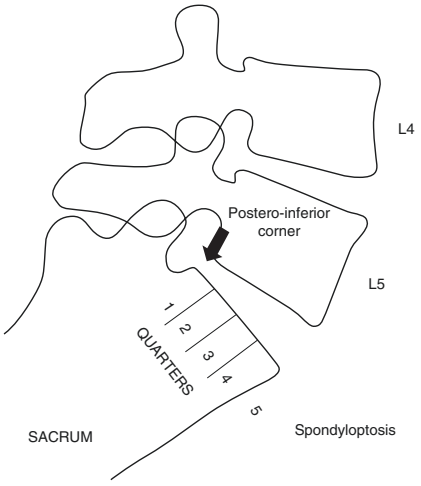
Further Readings

1. Williams BA, et al. Development and initial validation of a novel classification system for early-onset scoliosis: Classification of Early-Onset Scoliosis (C-EOS). J Bone Joint Surg Am. 2014;96(16):1359–67.
2. Skaggs, et al. A classification of growth friendly spine implants. J Pediatr Orthop. 2014;34(3):260–74.

Appendix K: Classification Systems for Spondylolisthesis

Meyerding Classification for Spondylolisthesis

Definition: The Meyerding classification is the most commonly used method of assessing the amount of olisthesis of one vertebra (forward displacement) over the vertebra below. In particular, Meyerding defined the slippage on plain radiographs (lateral view) in accordance to the vertebra below.
Content: The classification divides the superior endplate of the vertebra below into four quarters; the grade depends on the location of the posteroinferior corner of the vertebra above.
Grade I: 0–25%
Grade II: 26–50%
Grade III: 51–75%
Grade IV: 76–100%
Grade V**:** >100% or spondyloptosis (complete displacement)
Clinical Application: Grading can help dictate the type of treatment depending on whether it is a low (grades I–II) or high grade (grades III, IV, and V).



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Spinal Deformity Study Group (SDSS) Classification

Definition: The SDSS classification is based on the sacropelvic morphology, slip grade, and spinal balance.
Content: The classification identifies two main groups: low- and high-grade spondylolisthesis; each group has three subtypes (I–III and IV–VI).

Low grade: *Type I*: Pelvic incidence <45° (nutcracker)
Type II: Pelvic incidence 45°–60°
Type III: Pelvic incidence >60°

High grade: Balanced pelvis: *Type IV*
Retroverted pelvis: *Type V*: balanced spine
Type VI: unbalanced spine

Clinical Application: SDSS classification can identify spondylolisthesis at the risk of progression and those requiring treatment. In particular, type 1 and type 2 spondylolisthesis have a lower risk of progression compared with type 3; reduction is indicated for type 5 and type 6.

Further Readings

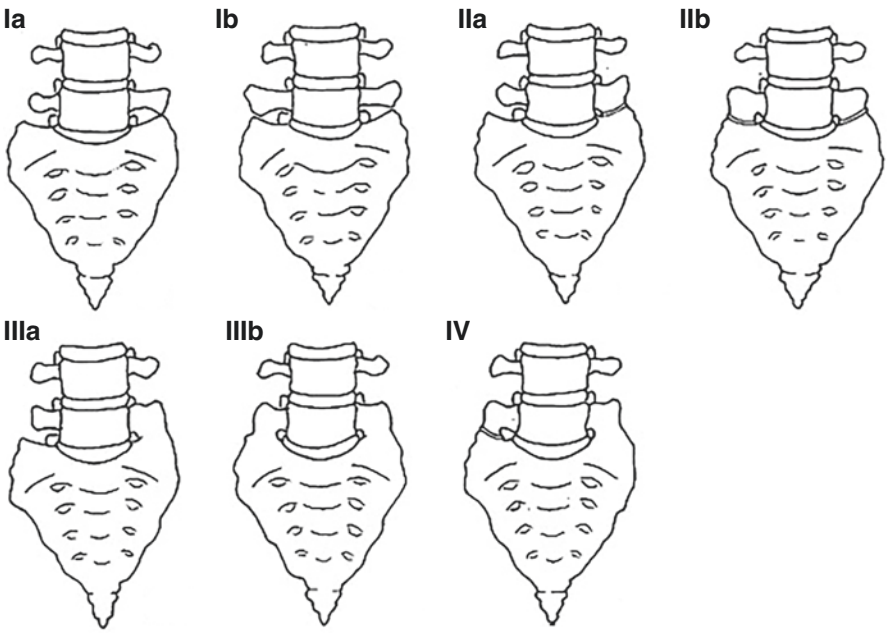
1. Meyerding HW. Spondylolisthesis. Surg Gynecol Obstet. 1932;54:371–7.
2. Mac-Thiong JM, Labelle H. A proposal for a surgical classification of pediatric lumbosacral spondylolisthesis based on current literature. Eur Spine J. 2006;15:1425–35.

Appendix L: Castellvi Classification of Lumbosacral Transitional Vertebrae

Definition: Castellvi identifies the lumbosacral transitional vertebrae according to the shape of the mega-transverse process and the presence/absence of a neo-joint (pseudo-joint).

Content: The classification identifies four types of lumbosacral transitional vertebrae:

- ***Type I***: enlarged and dysplastic transverse process (≥19 mm); ***Ia***: unilateral, ***Ib***: bilateral
- ***Type II***: pseudo-joint between the mega-transverse process and sacrum (incomplete sacralization); ***IIa***: unilateral, ***IIb***: bilateral
- ***Type III***: mega-transverse process fused with the sacrum (complete sacralization); ***IIIa***: unilateral, ***IIIb***: bilateral
- ***Type IV***: type IIa on one side and type III on the contralateral side



A. Şenköylü, F. Canavese (eds.), *Essentials of Spıne Surgery*,
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Further Reading

1. Castellvi AE, Goldstein LA, Chan DP. Lumbosacral transitional vertebrae and their relationship with lumbar extradural defects. Spine (Phila PA 1976). 1984;9(5):493–95.

Appendix M: Bone Age—Ossification of Hand, Olecranon, and Iliac Apophysis

1.1 Ossification of Hand (Sanders et al.) and Ossification of the Olecranon (Dimeglio et al.)

1.1.1 Ossification of the Hand

Definition: The ossification of the hand is evaluated on standard anteroposterior radiograph of the hand.
Content: The ossification of the hand correlates highly with scoliosis behavior; the method is rapid and reliable in clinical practice.

1.1.1.1 Ossification of the Olecranon

Definition: The ossification of the olecranon is evaluated on standard lateral radiograph of the elbow.
Content: The ossification of the olecranon correlates highly with scoliosis behavior; the method is rapid and easy to use in clinical practice.
Clinical relevance: Both methods allow to assess skeletal maturation, and help to estimate the risk of progression of several orthopedic disorders, including idiopathic scoliosis (Table M.1). The methods can complement each other for more precise bone age assessment. The olecranon method offers detailed information during the pubertal growth spurt, while the digital method is as accurate but less detailed, making it more useful after the pubertal growth spurt once the olecranon has ossified (Table M.2).

Ossification of the Iliac Apophysis (Risser Sign)

Definition: The Risser sign is the ossification of the iliac apophysis (left side); it progresses from the anterior iliac spine (lateral on anteroposterior radiographs of the pelvis) to the posterior end of the iliac crest (medial).

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Table M.1 Summary of olecranon, hand, and iliac apophysis methods of assessing bone age

Dimeglio	Olecranon	Sanders	Hand	Risser	Iliac apophysis
1	Double-ossific nucleus	1 (juvenile slow)	Distal epiphysis not covered	0	No ossification
2	Single-ossific nucleus (half-moon)	2 (adolescent slow)	All epiphyses are covered	1	<25% ossification
3	Single-ossific nucleus (quadrangular)	3 (adolescent rapid, early)	Most digits are capped Metacarpal epiphysis is wider than metaphysis	2	25–50% ossification
4	Partial fusion	4 (adolescent rapid; late)	Any distal phalangeal physes are beginning to close	3	50–75% ossification
5	Complete fusion	5 (adolescent steady; early)	All distal phalangeal physes are closed; others are open	4	>75% ossification
		6 (adolescent steady; late)	Middle or proximal phalangeal physes are closing	5	Complete fusion
		7 (early mature)	Only distal radial physis is open		
		8 (mature)	Distal radial physis is closed		

Table M.2 Correlation between Dimeglio et al., Sanders et al., and Risser methods. The pubertal growth spurt is in gray

Dimeglio	Sanders	Risser
0	1	0
0	2	
1-2-3-4-5	3	
	4	
5	5	
	6	>1
	7	4
	8	5

Content: The ossification of the iliac apophysis is divided into five stages (1–5):
Risser 0: no ossification of the iliac apophysis; greatest remaining growth (greater risk of progression for scoliosis).
Risser 1: less than 25% of the iliac apophysis is ossified; menarche occurs in girls; it is the end of the pubertal growth spurt (ascending side).
Risser 2: between 25% and 50% of the iliac apophysis is ossified.
Risser 3: between 50% and 75% of the iliac apophysis is ossified.
Risser 4: more than 75% of the iliac apophysis is ossified.
Risser 5: the apophysis has fused with the iliac crest (skeletal maturation is completed) and the risk of progression for curve <40° is minimal.
Clinical relevance: The Risser sign is a marker of skeletal maturation, and it helps to estimate the risk of progression of several orthopedic disorders, including idiopathic scoliosis.

Further Readings

1. Sanders J, Khoury JC, Kishan S, et al. Predicting scoliosis progression from skeletal maturity: a simplified classification during adolescence. J Bone Joint Surg Am. 2008;90(3):540–53.
2. Canavese F, Charles YP, Dimeglio A, et al. A comparison of the simplified olecranon and digital methods of assessment of skeletal maturity during the pubertal growth spurt. Bone Joint J. 2014;96(11):1556–60.
3. Risser JC. The Iliac apophysis: an invaluable sign in the management of scoliosis. Clin Orthop. 1958;11:111–19.

Appendix N: Oswestry Disability Questionnaire (ODI)

Abstract The Oswestry Disability Index (ODI) is among the most widely used patient-reported outcome measures for the assessment of spinal conditions. Traditionally, the ODI has been administered in outpatient clinics on a face-to-face basis, which can be expensive and time consuming.

Keywords Oswestry Disability Questionnaire (ODI); assessment of disability; outcome measurement

Definition

Oswestry Disability Index (ODI), also known as the *Oswestry Low Back Pain Disability Questionnaire*, is a patient questionnaire which assesses the amount of restriction imposed by pain on ten domains: pain intensity, personal care, lifting, walking, sitting, standing, sleeping, sex life, social life, and traveling. Each question offers six answers, which allows the assessment of subtle differences of disability [1, 3].

Content: ODI was developed by Fairbank et al. [2]. It is one of the most commonly used questionnaires in the field of low back pain. It is available in several languages and has proven good internal consistency and test-retest reliability. The test is considered the "gold standard" of low back functional outcome tools.

Clinical Application: Assessment of the degree of disability caused by low back pain and leg pain.

Scoring Instructions

For each section there are six possible answers—between 0 and 5. The patient is asked to identify which of the six statements in each domain applies to them at the time of evaluation. The sentences are arranged from no impairment (0) to maximum impairment (5).

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If all ten sections are completed the score is calculated as follows:
Example: 18 (total scored) of 50 (total possible score) × 100 = 36% (18:50 × 100 = 36)
The alternative option is to calculate the total score and multiply by two
Example 2: 12 (total scored) × 2 = 24
If one section is missed or not applicable the score is calculated: 16 (total scored)
45 (total possible score) × 100 = 35.5%

Minimum detectable change (90% confidence): 10% points (change of less than this may be attributable to error in the measurement)

Interpretation of Scores

0–20%	Minimal disability	The patient can cope with most living activities. Usually no treatment is indicated apart from advice on lifting, sitting, and exercise.
21–40%	Moderate disability	The patient experiences more pain and difficulty with sitting, lifting, and standing. Travel and social life are more difficult and this has an impact on their travel and social life, which can result in being signed off work. Personal care, sexual activity, and sleeping are not grossly affected. The patient can usually be managed by conservative means.
41–60%	Severe disability	Pain is reported as the main problem for this particular group of patients. However, travel, personal care, social life, sexual activity, and sleep are also affected. These patients require detailed investigation to rule out possible red flags.
61–80%	Crippled	Back pain affects all aspects of the lives of these patients in their home and working environment. They require active intervention.
81–100%	Bed-bound or exaggerating symptoms	Patients require careful observation during their physical examination to discriminate between patients who are experiencing severe pain or exaggerating their symptoms.

1.1.1 Oswestry Low Back Pain Disability Questionnaire

Instructions
This questionnaire has been designed to give us information as to how your **back or leg pain** is affecting your ability to manage in everyday life. Please answer by checking ONE box in each section for the statement which best applies to you. We realize that you may consider that two or more statements in any one section apply but please just shade out the spot that indicates the statement which most clearly describes your problem.

Section 1: Pain intensity

□ I have no pain at the moment.
□ The pain is very mild at the moment

- ▫ The pain is moderate at the moment
- ▫ The pain is fairly severe at the moment
- ▫ The pain is very severe at the moment
- ▫ The pain is the worst imaginable at the moment

Section 2: Personal care (washing, dressing, etc.)

- ▫ I can look after myself normally without causing extra pain
- ▫ I can look after myself normally but it causes extra pain
- ▫ It is painful to look after myself and I am slow and careful
- ▫ I need some help but manage most of my personal care
- ▫ I need help every day in most aspects of self-care
- ▫ I do not get dressed, I wash with difficulty and stay in bed

Section 3: Lifting

- ▫ I can lift heavy weights without extra pain
- ▫ I can lift heavy weights but it gives extra pain
- ▫ Pain prevents me from lifting heavy weights off the floor, but I can manage if they are conveniently placed, e.g., on a table
- ▫ Pain prevents me from lifting heavy weights, but I can manage light to medium weights if they are conveniently positioned
- ▫ I can lift very light weights
- ▫ I cannot lift or carry anything at all

Section 4: Walking

- ▫ Pain does not prevent me from walking any distance
- ▫ Pain prevents me from walking more than one mile
- ▫ Pain prevents me from walking more than 1/2 mile
- ▫ Pain prevents me from walking more than 1/4 mile
- ▫ I can only walk using a stick or crutches
- ▫ I am in bed most of the time and have to crawl to the toilet

Section 5: Sitting

- ▫ I can sit in any chair as long as I like
- ▫ I can only sit in my favorite chair as long as I like
- ▫ Pain prevents me from sitting more than one hour
- ▫ Pain prevents me from sitting more than 30 minutes
- ▫ Pain prevents me from sitting more than 10 minutes
- ▫ Pain prevents me from sitting at all

Section 6: Standing

- ▫ I can stand as long as I want without extra pain
- ▫ I can stand as long as I want but it gives me extra pain
- ▫ Pain prevents me from standing for more than one hour
- ▫ Pain prevents me from standing for more than 30 minutes
- ▫ Pain prevents me from standing for more than 10 minutes
- ▫ Pain prevents me from standing at all

Section 7: Sleeping

- ▫ Pain does not prevent me from sleeping well
- ▫ I can sleep well only by using medication

- ▫ Even when I take medication, I have less than 6 hours' sleep
- ▫ Even when I take medication, I have less than 4 hours' sleep
- ▫ Even when I take medication, I have less than 2 hours' sleep
- ▫ Pain prevents me from sleeping at all

Section 8: Social life

- ▫ My social life is normal and gives me no extra pain
- ▫ My social life is normal but increases the degree of pain
- ▫ Pain has no significant effect on my social life apart from limiting my more energetic interests, e.g., dancing
- ▫ Pain has restricted my social life and I do not go out as often
- ▫ Pain has restricted my social life to my home
- ▫ I have no social life because of pain

Section 9: Travelling

- ▫ I can travel anywhere without extra pain
- ▫ I can travel anywhere but it gives me extra pain
- ▫ Pain is bad, but I manage journeys over 2 hours
- ▫ Pain restricts me to journeys of less than 1 hour
- ▫ Pain restricts me to short necessary journeys under 30 minutes
- ▫ Pain prevents me from traveling except to the doctor or hospital

Section 10: Employment/homemaking

- ▫ My normal homemaking/job activities do not cause pain
- ▫ My normal homemaking/job activities increase my pain, but I can still perform all that is required of me
- ▫ I can perform most of my homemaking/job duties, but pain prevents me from performing more physically stressful activities (e.g., lifting, vacuuming)
- ▫ Pain prevents me from doing anything but light duties
- ▫ Pain prevents me from doing even light duties
- ▫ Pain prevents me from performing any job or homemaking chores

Further Readings

1. Fairbank JC. Oswestry disability index. J Neurosurg Spine. 2014;20:239–41. https://doi.org/10.3171/2013.7.SPINE13288.
2. Fairbank JC, Couper J, Davies JB, O'Brien JP. The Oswestry low back pain disability questionnaire. Physiotherapy 1980;66:271–3.
3. Fairbank JC, Pynsent PB. The Oswestry Disability Index. Spine. 2000;25:2940–52; discussion 2952. https://doi.org/10.1097/00007632-200011150-00017.

Appendix O: SRS-22 Questionnaire

Definition: The Scoliosis Research Society-22 (SRS-22) questionnaire [2] is a well-accepted health-related quality-of-life assessment instrument that has been validated in many languages. It measures how idiopathic scoliosis affects the quality of life.
Content: The SRS-22 includes 5 domains and 22 questions related to the quality of life (Table O.1).
Clinical Application: The SRS-22 is a valid instrument for evaluating the patient-reported outcomes. It also helps to assess the effectiveness of the treatment in idiopathic scoliosis patients.

Table O.1 Domains of the SRS-22 questionnaire

Domain	Relevant questions
Function	5-9-12-15-18
Pain	1-2-8-11-17
Self-image	4-6-10-14-19
Mental health	3-7-13-16-20
Satisfaction with management	21-22

Further Readings

1. Asher M, Lai SM, Burton D, Manna B. The reliability and concurrent validity of the scoliosis research society-22 patient questionnaire for idiopathic scoliosis. Spine 2003;28(1):63–9.
2. https://www.srs.org/UserFiles/file/outcomes/srs-22.pdf

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Appendix P: Early-Onset Scoliosis Questionnaire

The 24-item Early-Onset Scoliosis Questionnaire (EOSQ-24) is a disease-specific instrument to measure health-related quality of life (HRQoL) of patients with EOS, and parental and financial burden of their caregivers.

The EOSQ-24 has shown to be reliable and sensitive to changes before and after surgical interventions, and to intra- and postoperative complications.

The EOSQ-24 has been validated for children aged 0–18 years, and their parents. The questionnaire has 11 domains and 24 items:

- General health (2 items)
- Pain/discomfort (2 items)
- Pulmonary function (2 items)
- Transfer (1 item)
- Physical function (3 items)
- Daily living (2 items)
- Fatigue/energy level (2 items)
- Emotion (2 items)
- Parental burden (5 items)
- Financial burden (1 item)
- Satisfaction (child and parents; 2 items)

Each item scores 1–5 points (1 point: worst scenario to 5 points: best scenario). The score of each domain can be obtained by the transformation of the algebraic mean of the items answered to standardized 0 (worst) to 100 (best) scores using the following algorithm: (algebraic mean of items answered − 1)/4 × 100.

For domains with one item only, the following algorithm should be used: (value of item choice − 1) × 100.

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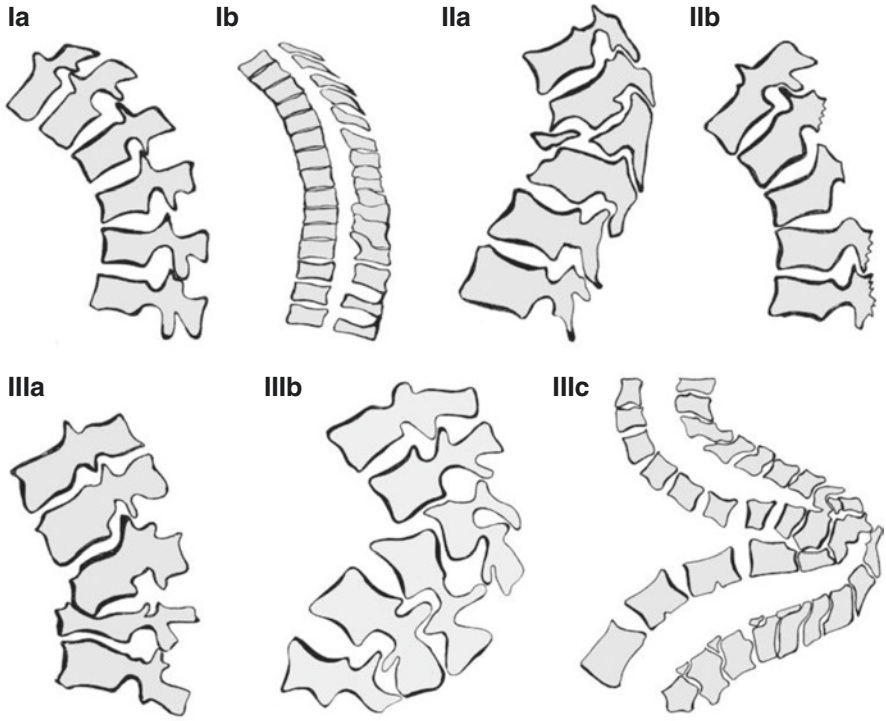
Further Reading

1. Matsumoto H, Williams B, Park HY, et al. The final 24-item Early Onset Scoliosis Questionnaires (EOSQ-24): validity, reliability and responsiveness. J Pediatr Orthop. 2018;38(3):144–51.

Appendix Q: Post-Infectious Kyphosis Classification

Definition: Post-infectious kyphosis involves loss of alignment due to bony destruction. This classification (Rajasekaran et al.) is based on spinal column deficiency, spinal flexibility, and curve magnitude and proposes surgical intervention.
Content:



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Type I: Intact anterior and posterior columns (IA: mobile disk spaces, IB: ankylosed segments)
Type II: Single-column deficiency (IIA: anterior column, IIB: posterior column)
Type III: Both column deficiency (IIIA: kyphosis of ≤60°, IIIB: kyphosis of >60°, and IIIC: buckling collapse)
Clinical application: The classification provides guidance on the surgical intervention required for each grade.

Further Reading

1. Rajasekaran S, et al. A classification for kyphosis based on column deficiency, curve magnitude, and osteotomy requirement. J Bone Joint Surg Am. 2018;100(13):1147–56.