INTRODUCTION

Scoliosis can arise from a variety of causes and is defined as a lateral curvature of the spine greater than 10° on an anterior-posterior standing radiograph (Fig. 1). However, in reality, it is a 3-dimensional structural deformity that includes a curvature in the anterior-posterior plane, angulation in the sagittal plane, and rotation in the transverse plane. This 3-dimensional deformity differentiates scoliosis from nonstructural spine deformities, which arise as compensation for abnormalities in other regions (eg, lower limb disorders resulting in limb length discrepancy), in which case the deformity is mono-planer and resolves when the primary abnormality is treated.

IDIOPATHIC SCOLIOSIS

The most common cause of scoliosis is idiopathic, which accounts for up to 80% of scoliosis in children.1 The cause of idiopathic scoliosis is unknown and is a diagnosis...
Scoliosis Classification

**Neuromuscular**

- **Neuropathic**
  - Upper motor neuron
  - Cerebral palsy
  - Spino cerebellar degeneration: Friedrich’s Ataxia
  - Charcot-Marie-Tooth Disease
  - Syringomyelia
  - Spinal Cord Tumor
  - Spinal Cord Trauma

- **Myopathic**
  - Arthrogryposis
  - Muscular dystrophy: Duchenne’s
  - Limb-girdle
  - Fascioscapulohumeral
  - Congenital hypotonia
  - Myotonia dystrophica

**Idiopathic**

- Infantile
- Juvenile
- Adolescent

**Congenital**

- Failure of formation: Wedge vertebra
- Hernivertebra
- Failure of segmentation
- Unilateral bar
- Block vertebra
- Mixed

**Miscellaneous**

- Neurofibromatosis
- Connective tissue: Marfan’s syndrome
- Ehlers-Danlos
- Osteochondrodystrophies: Diastrophic Dysplasia
- Mucopolysaccardiosis
- SED
- MED
- Achondroplasia
- Metabolic: Rickets
- OI
- Homosystinuria
- Tumors

**Fig. 1. Classification of scoliosis.**

of exclusion. It is classified based on age of onset into infantile (0–3 years), juvenile (3–10 years), and adolescent (>10 years). These 3 periods mark the different periods of growth velocity during childhood; hence, the curves behave differently.

A different classification, first used by Dickson, separates idiopathic scoliosis into early onset (<5 years) and late onset (>5 years), given that the natural history, prevalence, and treatment methods for patients with scoliosis when younger than 5 years is significantly different from patients presenting with scoliosis when older than 5 years. Another advantage of this classification is that it separates scoliosis into 2 distinct periods of pulmonary development; from 0 to 5 years of age is the period of major pulmonary development, and a thoracic deformity during this period will have a greater impact on pulmonary function than one developing in later years. Early onset scoliosis includes all patients with an age of onset of less than 5 years regardless of the cause; however, more recently, there is a growing trend toward changing this definition to less than 10 years of age regardless of cause.

**Infantile Idiopathic Scoliosis**

Infantile idiopathic scoliosis accounts for less than 1% of idiopathic scoliosis. It is more common in boys (ratio: 3:2); most are convex left curves (75%–90%); most tend to resolve spontaneously and often can be associated with plagiocephaly (80%–90%).

**Juvenile Idiopathic Scoliosis**

Juvenile idiopathic scoliosis makes up between 12% and 21% of patients with idiopathic scoliosis. Juvenile idiopathic scoliosis is a transition between infantile and adolescent idiopathic scoliosis. There is a slight female preponderance ranging from 1.6:1.0 to 4.4:1.0, which tends to increase with increasing age of onset. A right thoracic curve is predominant in this category. Because the juvenile period is a period of slow spinal growth, the natural history is that of slow progression until about
10 years of age when curve progression is more rapid, coinciding with the period of accelerated spine growth. Because of the earlier age of onset compared with adolescent idiopathic scoliosis, they are more likely to progress to severe deformity and less likely to respond to nonsurgical treatment.

**Adolescent Idiopathic Scoliosis**

Adolescent idiopathic scoliosis (AIS) is the most common type of scoliosis with an overall incidence in the population of 2% (Fig. 2). The female-to-male ratio tends to increase with increasing magnitude of the curve: 1:1 for curves less than 10°, 1.4:1.0 for curves between 11° and 20°, 5.4:1.0 for curves 21° and greater, and 7.2:1.0 for curves requiring treatment. The natural history and risk of progression of AIS depends on several factors, including skeletal maturity, sex, and curve magnitude. Curves in girls are more likely to progress and are more likely to require treatment. The curve magnitude increases with skeletal growth; hence, the more skeletally immature a patient is, the greater the likelihood is for the curve to progress. Another determinant of curve progression is the curve magnitude at presentation. Patients with curves greater than 20° who are skeletally immature are at a greater risk for curve progression.

**Fig. 2.** (A) Clinical photograph of a 14-year-old girl with AIS. Note the convex right main thoracic scoliosis with resultant elevation of the right shoulder and trunk shift to the right. (B) Standing posteroanterior radiograph of the same patient indicating a scoliosis of greater than 90°. Radiographs are oriented as if looking at the patient from behind.
CONGENITAL SCOLIOSIS

Congenital scoliosis arises as a result of congenital malformations of the spine that are present at birth; however, because of the effects of growth, the deformity may not be apparent until later in childhood. Congenital scoliosis is classified as a failure of formation of a vertebral body (hemivertebrae), failure of segmentation between 2 or more vertebrae (bar), or a failure of segmentation in combination with a failure of formation (Fig. 3). The natural history of congenital scoliosis depends on the type of malformation, with the combination of a unilateral hemivertebrae and a contralateral bar having the worst prognosis. A significant percentage (61%) of patients with congenital scoliosis have an associated anomaly in other organ systems, which may appear independently or as part of a syndrome. VACTERL syndrome (vertebral, anorectal, cardiac, tracheoesophageal, renal, and limb) is often found to be associated with congenital scoliosis. It is important to screen for these other potential abnormalities when assessing patients with congenital scoliosis. The authors routinely order renal ultrasound and echocardiograms on all patients with a diagnosis of congenital scoliosis.

NEUROMUSCULAR SCOLIOSIS

Neuromuscular scoliosis is scoliosis arising as a result of neurologic or muscular disorders. The Scoliosis Research Society has classified it into neuropathic and myopathic causes. Neuropathic causes include upper motor neuron lesions, such as cerebral palsy, spinocerebellar degeneration (Fredrick ataxia, Charcot-Marie-Tooth disease), syringomyelia, spinal cord tumors and trauma, and lower motor neuron lesions, such as poliomyelitis, spinal muscular atrophy, and myelomeningocele. Myopathic conditions include arthrogryposis, muscular dystrophies (Duchene,
limb-girdle, facioscapulohumeral), congenital hypotonia, and myotonia dystrophica. The underlying cause for all neuromuscular scoliosis is the lack of muscular support to the spinal column, which allows gravity and posturing that leads to deformity of the spine. The age of onset of neuromuscular scoliosis and the natural history vary depending on the cause.

**MISCELLANEOUS CAUSES**

Other causes of scoliosis include tumors, neurofibromatosis, connective tissue disorders (Marfan syndrome, Ehlers-Danlos syndrome), osteochondrodystrophies (dystrophic dysplasia, mucopolysaccharidosis, spondyloepiphyseal dysplasia, multiple epiphyseal dysplasia, achondroplasia), and metabolic causes (rickets, osteogenesis imperfecta).

**EVALUATION OF SCOLIOSIS**

The evaluation of patients with scoliosis starts with a detailed history and thorough physical examination, which are aimed at identifying nonidiopathic causes and at identifying features that are associated with rapid progression of the scoliosis.

**HISTORY**

Keys in the history include the age of onset, history of progression of the spinal deformity, and how the curve was noticed and by whom (child, parent, school screening, or by the primary physician). The usual presenting complaint is chest wall or back asymmetry. Adolescent girls will sometimes complain of breast asymmetry, unequal shoulders, uneven waistline and difficulty with fitting clothes.

Associated symptoms, such as pain, neurologic, or respiratory, should be sought. Although pain is not a prominent feature of scoliosis, about one-quarter of patients with AIS will present with pain, which is mostly benign and nonspecific. Some patients will complain of posterior chest wall pain around the area of rib prominence. Persistent and severe back pain, with red flags signs like fever and constitutional symptoms, might be related to infection and should be investigated further. Back pain that is worse at night and relieved by nonsteroidal antiinflammatory medication might suggest osteoid osteoma of the spine, which may create a deformity of the spine.

History of breathing difficulty and failure to thrive in a child presenting with scoliosis and chest wall deformity might suggest pulmonary insufficiency syndrome and warrants further pulmonary evaluation.

The evaluating physician should seek to identify neurologic symptoms, such as sensory or motor weakness and difficulty with coordination, gait, and balance. Any bowel or bladder symptoms may be secondary to intraspinal diagnoses like syringomyelia, tumor or tethered cord.

For a child presenting with scoliosis, a detailed perinatal history including any illness during the pregnancy, medications taken, length of gestation, mode of delivery, and birth weight should be obtained. Developmental history, both motor and cognition, should also be noted as these may indicate a neuromuscular or syndromic causes of scoliosis.

For adolescent patients, a history of adolescent growth spurts and other maturity indicators like menarche status in girls (both onset and duration) are important. The risk of curve progression and methods of treatment depend on the amount of spinal growth remaining. Psychosocial history is important in assessing an adolescent with
scoliosis, as patients are often not happy with the cosmetic deformity. The patients’ main desire to treat the scoliosis may be related to the cosmetic deformity.

Past medical and surgical history are important in ruling out any syndromic features. A history of heart disease might prompt one to look at other syndromic features (Marfan syndrome, for example). Any family history of scoliosis should be noted, as there is often a genetic component associated with AIS.18 For neuromuscular patients, it is important to note the associated comorbidities, including medications that could impact any planned surgical treatment.

EXAMINATION

The physical examination should include a general head-to-toe examination, including assessment for pubertal development in adolescents. Patients’ height should be documented and plotted on a growth chart to monitor for peak growth velocity. Clinicians should evaluate for syndromic features and associated deformities: The presence of plagiocephaly, bat ear, or torticollis may suggest infantile idiopathic scoliosis; skin manifestations, such as café au lait spots and axillary freckles, may suggest neurofibromatosis; sacral dimple, hairy patch, or lipoma at the lower back may suggest spinal dysraphism (ie, myelomeningocele); cavus foot may suggest a sensory-motor abnormality (ie, Charcot-Marie-Tooth Disease or a spinal cord tumor). Tall patients, with an increased ratio of arm span to height, should prompt examination for other features of Marfan syndrome, including cardiac and ophthalmologic examination. The presence of joint hyperlaxity and poor skin tone might point to a connective tissue disorder, such as Ehlers-Danlos syndrome.

Examination of the deformity begins with inspection (from behind the patients) for shoulder and flank asymmetry. Care should be taken to ensure that the pelvis is as level as possible. If the pelvis remains unlevel, this might suggest a leg-length discrepancy, which may be the cause of the scoliosis. If this seems to be the case, the patient should be examined while in the seated position. A curve that is caused by a leg-length discrepancy disappears with patients sitting.

The Adam’s forward bending test assesses the curve rotation (Fig. 4).19,20 By assessing patients from behind, while they forward flex, the axial plane rotation associated with a structural scoliosis will be evidenced by a rib prominence in the thoracic spine and/or paraspinal muscle prominence in the lumbar spine. In a child that is too young for an Adam’s forward bending test, laying the child prone may help in assessing the rotational deformity. The curve flexibility can be assessed at the same time by rotating the child to a lateral position while still supine. Suspending the child under the arm of the examiner can also assess the flexibility of the spinal deformity. Also, an evaluation for asymmetry or limitation in chest excursion, may suggest thoracic insufficiency syndrome.

The clinician should perform a complete neurologic examination including a cranial nerve examination as well as sensory, motor, and reflex evaluation of the upper and lower extremities. The abdominal reflex should be performed in order to evaluate for potential neural axis abnormalities in the thoracic spine. Absent abdominal reflex is also seen in some patients with Chiari malformation.21

SCREENING FOR ADOLESCENT IDIOPATHIC SCOLIOSIS

School screening of healthy asymptomatic adolescents for AIS has been a controversial issue over the years, with arguments for and against the benefit of routine screening. However, the Scoliosis Research Society, the Pediatric Orthopedic Society of North America, the American Academy of Orthopedic Surgeons, and the
American Academy of Pediatrics all agree that girls should be screened twice, at 10 and 12 years of age, whereas boys should be screened once at 13 or 14 years of age. Clinical signs used in screening programs include shoulder asymmetry, asymmetry of scapular prominence, greater space between the arm and the body on one side compared with the opposite side with arm hanging loosely by the side, head not centered over the pelvis (examining patients from the back), and the Adam’s forward bending test. A scoliometer is used to measure any rotational deformity on the Adam’s forward bending test, and a trunk rotation of $7^\circ$ or greater is an indication for referral. Modern scoliometers are now readily available as applications for smartphones. The clinical benefit of screening is thought to be that it leads to the early detection of curves, and the institution of early brace therapy can alter the natural history of the deformity.

**RADIOGRAPHIC EVALUATION**

**Plain Radiograph**

Initial radiographic evaluation of patients with suspected scoliosis is with a standing posterior-anterior (PA) and lateral radiograph of the whole spine include the hip joints in a single 3-ft film (see Fig. 2B). PA radiography minimizes radiation to organs including the breast and thyroid. Recently, a low dose radiographic system has been introduced (EOS Imaging, Paris, France), which can capture these images of standing patients in a single scan in both frontal and sagittal views simultaneously without having to stitch the images together and without vertical distortion. If there is an

**Fig. 4.** A 7-year-old girl with congenital scoliosis. (A) Standing evaluation evaluates the lateral curvature of the spine. (B) Adam’s forward bending test evaluates the axial plane rotation of the spine. This patient’s preoperative and postoperative computed tomography scans are demonstrated in Figs. 3B and 10B, respectively.
associated leg-length discrepancy, it should be corrected by placing an appropriately sized wooden block under the short leg to level the pelvis while the standing radiographs are obtained. A supine radiograph is obtained if a patient is too young to stand independently, and a sitting radiograph is obtained for wheelchair ambulators.

PA radiographs are conventionally viewed with the heart on the left side as if looking at patients from behind. This position mimics the view that the clinician has during the clinical assessment for scoliosis and is also the position of patients during posterior spinal fusion and instrumentation surgery for scoliosis. In addition to evaluating the curve magnitudes by measuring the Cobb angles, look for any obvious vertebral or rib malformations, which could suggest congenital scoliosis. The curves are described by the direction of the convexity and the location of the apex. For instance, most AIS curves are right main thoracic curves (see Fig. 2B).

On the lateral radiograph, there is generally apical hypokyphosis associated with idiopathic scoliosis. Lack of vertebral rotation or lack of hypokyphosis at the apex may suggest a nonidiopathic cause of the deformity, such as tumor (osteoid osteoma) or intraspinal abnormality (syringomyelia). For patients with AIS, examine for features of skeletal maturity, including the Risser sign (maturity of iliac crest apophysis) and open versus closed triradiate cartilage of the acetabulum. These features are used in predicting the growth remaining, hence, the curve progression, which influences the choice of treatment. The curve magnitude is evaluated using the Cobb method of measuring the angular deformity from the upper end vertebrae to the lower end vertebrae.

For patients with infantile idiopathic scoliosis, the likelihood of curve progression is determined radiographically by measuring the rib-vertebral angle difference (RVAD) of Mehta (Fig. 5). An RVAD of 20° or less indicates that the curve is unlikely to progress, whereas an RVAD of 20° or more indicates a curve is likely to progress.

![Fig. 5. Two methods of determining risk of scoliosis curve progression in patients with infantile idiopathic scoliosis. (A) Evaluation of the rib vertebral angle. At the apex of deformity, draw a line along the end plate of the vertebrae. Draw another line perpendicular to the first line. Draw a line along the rib. Measure the angle between the second and third lines. Repeat this for the contralateral rib. The difference between the 2 angles is the RVAD. If the RVAD is greater than 20°, this indicates significant rotation of the scoliosis and a high likelihood of curve progression. (B) Example of a phase-2 rib head. It also indicates significant rotation of the scoliosis and a high likelihood of curve progression. RVA, rib vertebral angle.](image)
An additional method of predicting curve progression also described by Mehta is the relationship of the convex rib head with the apical vertebra body (phase of the rib head). In phase-1 rib, there is no overlap of the rib head of the convex rib of the apical vertebra with the vertebral body; such curves have a low risk of progression. However, in phase-2 rib, there is an overlap; hence, there is a high risk of progression (see Fig. 5).

ANCILLARY INVESTIGATION

These tests are recommended for further evaluation of scoliosis and to help with surgical planning.

**Computed Tomography Scan**

A computed tomography (CT) scan may be used to further define the anatomy, including assessing congenital abnormalities or investigating for suspected tumor cause. CT can also be used in evaluating 3-dimensional lung volume in young patients who may not be able to comply with pulmonary function testing. Supine bending and stretch radiographs are used to assess the flexibility of the curve.

**MRI**

MRI is indicated for all patients presenting with early onset scoliosis. It is not routinely performed for patients with AIS, except for those with pain, atypical curve pattern (Fig. 6), large curve on presentation, rapidly progressive curve, or for patients with abnormal neurologic examination. There is an increasing trend to obtain an MRI scan for any patient requiring operative treatment of their scoliosis to rule out any unexpected intraspinal abnormality (see Fig. 6).

![Fig. 6](image-url) (A) MRI of a 9-year-old boy with scoliosis. The sagittal plane image demonstrates associated Chiari malformation and resultant cervical syrinx. (B) PA radiograph of the same patient demonstrating a left main thoracic curve pattern. This atypical curve pattern has a higher rate of associated neural axis abnormality and is an indication to obtain an MRI.
Other Investigations

Other investigations that are useful for a preoperative workup include an echocardiogram and a renal ultrasound for patients with congenital scoliosis; assessment of pulmonary function in patients with early onset scoliosis; and assessment of overall nutritional status, especially for patients with neurologic disorders.

MANAGEMENT OF SCOLIOSIS

Management of Adolescent Idiopathic Scoliosis

Because the natural history of AIS at skeletal maturity is for continued progression of the curve into adulthood only if the deformity is greater than 50°, the ultimate goal of the treatment of AIS is to keep the scoliosis less than 50° at maturity. The treatment choices are based on several factors, including curve magnitude, type and location of the curve, level of maturity, remaining growth, cosmetic appearance, and patient psychosocial factors. The options include observation, bracing, and surgery. In broad terms, the treatment guidelines for AIS are as follows:

Observation

Observation is recommended for curves that are 25° or less, regardless of the level of skeletal maturity. These patients require close radiographic monitoring for evidence of curve progression (5°–6° change in Cobb angle). The follow-up interval should be 3 to 6 months depending on the size of the curve and the level of skeletal maturity. Patients who are Risser grade 0 or 1 (immature) with curves close to 25° should be seen more frequently (3 monthly), whereas those who are Risser 3 and greater (more mature) with curves that are 20° or less are seen every 6 months.

Brace

Brace treatment is recommended for patients with curves between 25° and 45° who are Risser 2 or less. The goal of bracing is to prevent curve progression and to keep it below the surgical range at skeletal maturity. The most common type of brace used currently is the thoracolumbosacral orthosis (TLSO), which includes the Boston (Fig. 7), Charleston, and Providence braces. These braces are only suitable for curves with an apex at T7 or lower. The specific indication for the brace type depends on the type of curve. For the brace treatment to be successful, patients must be willing to comply with the prescribed amount of time in the brace. A recent randomized controlled trial by Weinstein and colleagues25 demonstrates that brace treatment was effective in decreasing the curve progression to the surgical threshold in AIS. They also showed that the benefit of brace wear increases with longer hours of wear. Patients should also be willing to accept their cosmetic deformity before treatment, as this is unlikely to improve. Hence, careful patient counseling before bracing is important.

Surgical treatment

Surgical treatment is recommended for patients with curves greater than 45° who are Risser 2 or less or for curves greater than 50 who are Risser 3 and greater. The goal of surgical treatment is to arrest the curve progression while improving spinal balance and alignment. This goal is achieved by inducing fusion of the spine by way of instrumentation and bone grafting. Fusion techniques have evolved over the years from Harrington’s26 introduction of the hook and rod construct in the 1960s to Luque’s27 segmental fixation with wires and the current third-generation segmental fixation with pedicle screws (Fig. 8). The underlying principle of all fixation techniques involves the placement of bony anchors, including hooks, wires, or pedicle screws to the
vertebrae and connecting them to a dual rod construct. Fusion can be performed anteriorly, posteriorly, or both depending on the curve type, magnitude, skeletal maturity, and the available skill set of the surgeon. The factors to consider in preoperative planning include the curve type and magnitude, spinal balance, curve flexibility, and the level of skeletal maturity.

Management of Early Onset Scoliosis

Recently, there has been a realization that regardless of cause, young patients with scoliosis have an increase risk of developing a pulmonary insufficiency syndrome that can lead to increased morbidity and mortality. This increased risk is because the bronchial tree and the alveolar are only fully developed by 8 years of age and the thoracic cavity is 50% of adult volume by 10 years of age. Also, the spine has its most rapid growth during the first 5 years of life (2.2 cm/y) before it slows down during the following 5 years (0.9 cm/y) and peaks again at puberty (1.8 cm/y). It has been shown that gaining a thoracic height of least 18 cm at maturity is associated with a better pulmonary function. These factors are important to consider in the treatment of early onset scoliosis. The goal of the treatment of early onset scoliosis is not only to stop progression of the spine deformity but to also allow for continued growth and development of the spine, thoracic cavity, and lungs. The treatment options include observation, nonsurgical treatment (bracing, casting, halo traction), and growth-friendly surgery.
Observation
Observation is for patients whose curves have a low risk of progression based on Mehta’s criteria. These curves have a curve angle of 25° or less and an RVAD of 20° or less and are followed up with serial radiographs every 4 to 6 months. Treatment should be commenced if there is curve progression of 10° or more. Curves with an RVAD of 20° or more or a phase-2 rib relationship are likely to progress and require treatment.

Serial casting
Serial casting is one of the nonsurgical methods used to delay fusion surgery in patients with early onset scoliosis (Fig. 9). It has been shown to be a viable alternative to growth-friendly surgery in early onset scoliosis32 and has been shown to cure some small idiopathic curves.33 Its attractiveness is that it is a nonsurgical treatment, hence, avoiding the potential complications with surgical treatment; however, patients
still require general anesthetics for its application and during routine cast changes every 3 to 4 months. There has been a recent increase in the use of casting in the management of early onset scoliosis with the realization of the high complication rate associated with growth-friendly surgical techniques (spinal growing rods and rib-based distraction surgery). It is indicated for patients with documented curve progression of 10° or greater, patients with curves of 25° or greater at presentation, those with an RVAD of 20° or greater or phase-2 rib rotation. Casting may be poorly tolerated in patients with poor pulmonary function or those with a neuromuscular disorder.

**Bracing**

Bracing is another nonsurgical method of delaying a curve. Bracing is an alternative to serial casting in patients who cannot tolerate casting and can also be used as a step down from casting after a satisfactory improvement in the curve with casting. It has an advantage over casting in that it is removable; however, this may contribute to lack of compliance in patients treated with a brace.

**Surgery**

Surgery is indicated for patients with progressive deformity or when casting/bracing has been ineffective or are contraindicated. Historically, surgical treatment of progressive early onset scoliosis has been spinal fusion similar to those performed in adolescence. Unfortunately, early surgical fusion ultimately led to restrictive lung disease because of the lack of growth of the spine and the pulmonary system, which resulted in early mortality from pulmonary insufficiency syndrome.31,34,35 These outcomes

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**Fig. 9.** A patient with early onset scoliosis who is being treated with a derotation cast. (A) Note the anterior chest and abdominal windows to allow for chest wall and abdominal excursion during respiration. (B) Viewed from behind, a window along the concavity of the scoliosis allows for the deformity to derotate in the axial plane. (Courtesy of J. d’Astous, MD, Salt Lake City, Utah, USA.)
have led to a shift toward growth-friendly surgery with the goal of arresting curve progression while allowing for spine growth and pulmonary development. Several growth-friendly surgical techniques have been developed over the years. Currently, the most commonly used technique is that of posterior distraction-based surgeries, which can be either spine based or rib based (Fig. 10). Spine-based distraction (spinal growing rods) involves the placement of anchors in the spine proximal and distal to the curve, which are connected to 2 rods. Rib-based distractions are similar to spine-based distraction; however, the proximal anchors are attached to ribs. The curve is controlled by serial distraction procedures approximately every 6 months, which grows the spine through the unfused segment. VEPTR is more commonly used in patients with associated chest wall deformity, such as absent or fused ribs. Given that posterior distraction treatment requires repetitive surgical interventions, the complication rates are high. Although these complications are a relative improvement as compared with the grim natural history of early onset scoliosis and to the poor long-term pulmonary outcomes associated with early surgical fusion, the quest for newer surgical techniques that do not require repetitive surgical interventions continues.

Fig. 10. Radiographic examples of posterior distraction based implants used for early onset scoliosis. (A) Image of an 8-year-old boy with juvenile idiopathic scoliosis who was treated with spinal growing rods. Note the superior and inferior foundations of spinal implants, which anchor the dual telescopic growing rods. (B) Three-dimensional CT reconstruction of the 7-year-old girl with congenital scoliosis and fused ribs who is featured clinically in Fig. 4. She has been treated with a rib-based distraction surgery, which is anchored to the ribs as well as to the spine. This type of device works well if there is an associated chest wall deformity. Note the improved spread between her left-sided ribs as compared with her preoperative CT scan demonstrated in Fig. 3B.
One such technique, still in its infancy, is growth guidance surgery (Shilla and Luque trolley systems), which involves a limited fusion at the apex of the curve with rods that are linked to anchors proximal and distal to the curve (Fig. 11). Because the rods are not completely constrained by the proximal and distal anchors, the rods are able to glide along the spine, which allows for continued spinal growth without periodic distraction surgeries. A variation on this theme is the magnetically controlled growing rod. This technique is similar to the commonly used spinal growing rods; however, rather than periodic distraction surgeries to lengthen the rods, the rods can be lengthened with the application of an external magnet in the clinic environment.

Another new technique that is evolving is convex side growth inhibition (staples and tethers). This technique involves applying a compression device on the convex side of the curve, producing inhibition of spine growth on that side and allowing the concave site to continue to grow, thereby strengthening the spine over time (Fig. 12). The advantage of this technique is also the avoidance of frequent anesthesia for distraction surgeries.

Management of Neuromuscular Scoliosis

Neuromuscular scoliosis includes a heterogeneous group of patients with different multisystem involvement; therefore, the treatment varies with the individual condition. In general, most patients with neuromuscular scoliosis are nonambulatory and are

![Fig. 11. Radiographs of a 6-year-old boy with syndromic scoliosis (Marfan). (A) Preoperative radiographs demonstrate significant thoracic and lumbar scoliosis. (B) Radiographs obtained after a growth guidance procedure. Note the anchors at the apex of deformity as well as at the superior and inferior ends of the constructs. The anchors at the upper and lower ends of the rods are designed in a way that they allow for the patient’s spine to continue to grow along the rods, which are purposefully left long to allow for this.](image-url)
wheelchair ambulators. The goal of treatment is to obtain a balanced spine over a level pelvis in order to maintain wheelchair seating balance.

**Nonsurgical treatment**
Nonsurgical treatments include wheelchair modification and bracing. These treatments can be technically demanding and require an experienced wheelchair specialist and orthotics department. Brace treatment is less effective in neuromuscular patients than in patients with AIS; however, the goal of brace treatment in neuromuscular patients is not to stop curve progression as in AIS but rather to maintain an upright posture in their wheelchair.

**Surgical treatment**
Surgical treatment and indications vary depending on the type of condition. For patients with cerebral palsy, most surgeons will consider surgery for a progressive curve of $50^\circ$ or greater or when there is deterioration in functional sitting. Other factors, like
medical comorbidity and caretaker concerns, should be taken into consideration. In patients with Duchene muscular dystrophy, surgery is advocated once the curve is more than the 20° to 30° range in nonambulatory patients because the natural history is that of rapid curve progression once patients are nonambulatory. In addition, the pulmonary and cardiac function of these patients generally worsens over time. There is increased surgical complication in patients with neuromuscular scoliosis; hence, a thorough preoperative workup and optimization is essential for these patients.

In terms of surgical planning, factors to consider include fusion levels, fixation type, and approach (anterior, posterior, or both). Most neuromuscular patients require long fusion from T2 to the pelvis (Fig. 13). Fixation to T2 is necessary to prevent proximal

Fig. 13. Sitting radiographs of a 13-year-old boy with scoliosis secondary to a neuromuscular condition (cerebral palsy). (A) Preoperative images demonstrate the typical neuromuscular curve pattern with a long thoracolumbar scoliosis extending to the pelvis with a resultant pelvic obliquity. (B) Radiograph obtained after posterior spinal fusion and instrumentation surgery. Note the extension of the implants to include the pelvis, which allows for correction of pelvic obliquity, which should improve wheelchair sitting balance.
junctional problems like kyphosis and screw pullout, whereas fixation to the pelvis is necessary to address pelvic obliquity that is often associated with neuromuscular curves. The exception to this is in ambulatory patients whereby pelvic fixation is thought to impair the ability to ambulate. Because of a combination of a high pseudoarthrosis rate and osteoporotic bone in these patients, segmental fixation is required. The choice of instrumentation depends on the surgeon’s preference, ranging from pedicle screw fixation to a hybrid of screws, wires, and hooks.

SUMMARY

Scoliosis is a 3-dimensional structural deformity of the spine. Although the causes are many, most patients have idiopathic scoliosis. A thorough clinical assessment and radiological evaluation is required to identify the nonidiopathic causes and to institute appropriate treatment. The treatment of scoliosis varies depending on the cause and ranges from observation, bracing, and casting to surgery.

REFERENCES