ADOLESCENT IDIOPATHIC SCOLIOSIS

Adolescent idiopathic scoliosis (AIS) is a structural deformity of the spine that is most often diagnosed just before or during puberty. The term idiopathic denotes that the disease or process has an unknown pathogenesis. AIS can only be diagnosed once other causes of spinal curvature and deformity (i.e., congenital, neuromuscular, infectious, or pathologic sources) have been ruled out. Screening occurs in the primary care office as well as in school starting in the preadolescent years.

Relevant Anatomy

The typical spine is nearly straight in the coronal plane, and has a sagittal profile with normal thoracic kyphosis (20 to 45 degrees) and lumbar lordosis (40 to 60 degrees). The spinal complement includes 7 cervical, 12 thoracic, and 5 lumbar vertebrae which connect to the sacrum. In scoliotic curves there is deviation from the midline in the coronal plane and axial rotation that is greatest at the apex of the curvature(s). In addition to this, most AIS is associated with a relative hypokyphosis in the thoracic spine.

Pathogenesis

There have been many theories as to the pathogenesis of AIS, and at this point it is commonly believed that there is a strong, although not fully understood, genetic component. Genealogy studies have shown an increased incidence of scoliosis in the family members of affected individuals. Paul Harrington showed a 27% incidence of scoliosis among female children born to mothers with a scoliotic curvature greater than 15 degrees. More recent data suggest that 10% of patients diagnosed with AIS will have a first-degree relative with the disease. Twin studies have shown that monozygotic twins have a concordance rate of 73% whereas dizygotic twins have much lower concordance rates. Despite a general consensus that scoliosis patients have increased global flexibility, many genes commonly associated with soft tissue disorders have been ruled out in the pathogenesis of AIS (e.g., genes that code for type I and II collagen, fibrillin, and elastin). Promising advances in hormonal and growth factor research have been made, but, as of yet, no study has shown conclusive evidence as to the underlying cause of AIS. As such, this form of scoliosis remains idiopathic until satisfactory evidence is presented.

Epidemiology

Studies of prevalence indicate that AIS is present in 0.5% to 4% of adolescents when defined curve value is a Cobb angle ≥10 degrees. Rates of small curves, not requiring treatment, are similar for males and females, but larger curves have a 4:1 female-to-male ratio. In addition, females have a 10 times greater risk of curve progression when compared to male counterparts. The most common curvature found is a right thoracic curve and/or a left lumbar curve, and patients with left thoracic curves should undergo additional screening to rule out nonidiopathic causes of scoliosis. Standardized screening is performed in many countries, often in school, beginning in the preadolescent years. Approximately 10% of children with a positive school screening will require treatment. A positive screening is generally defined as an inclinometer (scoliometer) reading of 7 degrees off center in the axial plane of the thorax or abdomen.

Classification

Before the advent of advanced imaging and surgical treatment options the King and Moe classification was the standard by which scoliotic curvatures were classified. This classification system has been supplanted by the Lenke system which takes into account multidimensional deformity and delineates six coronal curve patterns (one through six) which are then subdivided based on lumbar curve midline deviation (A, B, or C) and, finally, sagittal balance in the form of thoracic kyphotic profile (−, N, or +) (Table 22.1). The apex of each curve is defined by the most laterally deviated vertebral body or disk from the central vertical sacral line, and is classified as thoracic, thoracolumbar (T12–L1), or lumbar.

- The lumbar curve modifier is determined by where the central sacral vertical line intersects the lumbar apical vertebral body.
TABLE 22.1 CURVE PATTERNS AS DESCRIBED BY LENKE et al.

<table>
<thead>
<tr>
<th>Type</th>
<th>Proximal Thoracic</th>
<th>Main Thoracic</th>
<th>Thoracolumbar/Lumbar</th>
<th>Curve Type</th>
</tr>
</thead>
<tbody>
<tr>
<td>1</td>
<td>Nonstructural</td>
<td>Structural (major)</td>
<td>Nonstructural</td>
<td>Main thoracic</td>
</tr>
<tr>
<td>2</td>
<td>Structural</td>
<td>Structural (major)</td>
<td>Nonstructural</td>
<td>Double thoracic</td>
</tr>
<tr>
<td>3</td>
<td>Nonstructural</td>
<td>Structural (major)</td>
<td>Structural</td>
<td>Double major</td>
</tr>
<tr>
<td>4</td>
<td>Structural</td>
<td>Structural (major)</td>
<td>Structural</td>
<td>Triple major</td>
</tr>
<tr>
<td>5</td>
<td>Nonstructural</td>
<td>Nonstructural</td>
<td>Structural (major)</td>
<td>Thoracolumbar/lumbar</td>
</tr>
<tr>
<td>6</td>
<td>Nonstructural</td>
<td>Structural</td>
<td>Structural (major)</td>
<td>Thoracolumbar/lumbar; main thoracic</td>
</tr>
</tbody>
</table>

Modifiers

<table>
<thead>
<tr>
<th>Lumbar Spine Modifier</th>
<th>Location of Central Sacral Vertical Line at Lumbar Apex</th>
<th>Thoracic Sagittal Modifier</th>
<th>Sagittal Profile T5–12</th>
</tr>
</thead>
<tbody>
<tr>
<td>A</td>
<td>Between pedicles</td>
<td>–</td>
<td>Hypokyphotic &lt;10 degrees</td>
</tr>
<tr>
<td>B</td>
<td>Touches apical body</td>
<td>N</td>
<td>Normal 20–40 degrees</td>
</tr>
<tr>
<td>C</td>
<td>Lateral to body</td>
<td>+</td>
<td>Hyperkyphotic &gt;40 degrees</td>
</tr>
</tbody>
</table>


- The stable vertebra is the most proximal lower thoracic or lumbar vertebra that is bisected most closely by the central sacral vertical line (Fig. 22.1).
- Curves are considered structural if the Cobb angle is greater than 25 degrees on side bending films.
- Each patient is described by a designation, such as 1B+ (curve type/lumbar modifier/thoracic sagittal modifier), that thoroughly describes the curve.

Based on a multicenter review, the most common curve type is main thoracic, comprising 51% of curves, followed by double thoracic (20%), thoracolumbar/lumbar (12%), and double major curves (11%).

Diagnosis

Screening

Universal screening for scoliosis is not an absolute practice in the United States of America, but most states and school systems support screening of asymptomatic preadolescents and adolescents. In 2004, the U.S. Preventive Services Task Force (USPSTF) had an acute change in their position and recommended against the routine screening of adolescents for idiopathic scoliosis. In the wake of this decision many medical societies (American Academy of Pediatrics, American Academy of Orthopaedic Surgeons, Pediatric Orthopaedic Society of North America, and the Scoliosis Research Society) formed consensus statements further recommending scoliosis screening. All four societies recognize the benefits that can be provided by effective clinical screening programs, including (1) the potential prevention of

Figure 22.1 Radiograph showing the central sacral vertical line (CSVL), apical vertebra, stable vertebra, and end vertebra. The stable vertebra is the most proximal lower thoracic or lumbar vertebra that is bisected most closely by the CSVL.
deformity progression by brace treatment and (2) the earlier recognition of severe deformities requiring operative correction. With recent evidence, from the Bracing in Adolescents with Idiopathic Scoliosis Trial (BrAIST) study, supporting the efficacy of bracing in the treatment of moderate curves, there may be increased support for school screening programs.

Screening is performed using the Adams forward bending test. With the patient bending forward from the waist, the examiner views the patient from behind for thoracic asymmetry, from in front for lumbar asymmetry, and from the side for assessment of kyphosis. A rib prominence seen with the forward bending test is from rotation of the spine and can be measured with an inclinometer (Fig. 22.2). An angle of trunk rotation (ATR) measurement of 7 degrees is the current recommendation for referral to an orthopaedist; this was established from screening data gathered by Bunnell. The data from his scoliosis screening research also showed that 98% of patients with an ATR 5 degrees or less will have a Cobb angle <20 degrees on x-ray, but that figure decreases to 94% at 6 degrees, 88% at 7 degrees, and 50% at 10 degrees. With this level of screening, approximately 10% of children referred require treatment. With newer evidence outlining the importance of awareness in radiation exposure to the growing patient, it is important to note that not all patient referrals lead to the procurement of radiographs. However, each patient should have a thorough examination by a medical professional before radiographs of the spine are obtained. The ATR cutoff of 7 degrees on the inclinometer favors a type I error, but prevents missing patients with curves who are likely to progress.

History
As in all medical conditions, an accurate history is vital in the diagnosis and management of idiopathic scoliosis. In addition to routine questions (age, gender, medical history, family history, and social history) the following points should be queried in the initial patient encounter:

- Pubertal/menarchal status: onset of secondary sexual characteristics, menarche in females, and voice changes in males
- Family history of scoliosis: 3x greater risk if parent has scoliosis and 7x greater risk if sibling has scoliosis
- Current patient height and height of parents and skeletally mature siblings (if available)
- Back pain and/or neurologic symptoms: warning signs that scoliosis may not truly be idiopathic in nature
- Other medical problems such as Marfan’s, Ehlers-Danlos, or neurofibromatosis

Figure 22.2  A: PA standing radiograph of a 28-degree right thoracic idiopathic scoliosis. B: Adams forward bend test with 11 degrees of ATR measured on inclinometer. (Reprinted with permission from Lovell and Winter's pediatric orthopaedics seventh edition, Wolters Kluwer Health, Figure 17.8.)
Physical Examination

The examination in scoliotic deformity should include current height and weight, presence of secondary sexual characteristics, dermatologic examination (café au lait spots, which may indicate neurofibromatosis), neurologic examination, limb length, and assessment for signs of spinal dysraphism (patch of hair or dimple along the spine).

The standing patient should be assessed for asymmetry of the trunk, back, shoulder, and neckline. The Adams forward bend test can then be used in conjunction with an inclinometer to evaluate rotational deformity. The patient should be viewed from the side, while performing the forward bend, to assess for any kyphosis in the thoracic spine. An inclinometer reading of 7 degrees is roughly correlated with a radiographic Cobb angle measurement of 15 to 20 degrees (Fig. 22.2). Leg length discrepancy is a common reason for spurious diagnoses of spinal curvature. Leg lengths can be measured supine or standing, but if a leg length discrepancy exists then it should be taken into account (typically with a measured wooden block that levels the pelvis) before further physical examination or radiographic evaluation.

The standard neurologic assessment for scoliosis patient includes strength testing, deep tendon reflexes, straight leg raise, Babinski sign, abdominal reflexes, and examination for clonus. In patients where stenosis or intrathecal anomaly is suspected more specific tests may be performed.

Radiologic Features

In scoliosis, radiographs allow for diagnosis, quantification of curve, serial observation, and treatment planning. The standard screening radiograph is a standing posteroanterior (PA) radiograph on a 36-in cassette which should measure from the occiput to the pelvis in most patients. The PA view allows for assessment of coronal plane deformity as well as some inference of the axial plane based on rotation. If the pelvis is included in the screening PA radiograph then the surgeon can make commentary on the Risser sign (Fig. 22.3) as well as the status of the triradiate cartilage, which are indicators of bony maturity. An approximation of any leg length difference can also be made based on a PA radiograph that includes the iliac crest or top of the femoral heads. Lateral radiographs, also taken on a full size cassette, allow for examination of the sagittal profile. Often the lateral radiograph is omitted during the screening process and acquired once a diagnosis of scoliosis is made or if there is concern for kyphosis or spondylolisthesis.

The apical vertebra is used to describe the location of the curve or curves (thoracic, thoracolumbar, or lumbar), and the Cobb method is used to measure the coronal plane magnitude of the curve. The Cobb method is performed by marking the end plates on the most tilted vertebrae at the top and bottom of the curve. The angle of intersection between the lines (formed by the end plates) is the Cobb angle (Fig. 22.4). Measured angles of less than 10 degrees are considered to be normal.
physiologic variation, but curves greater than 10 degrees are considered scoliotic curvatures and should be followed for progression. The timing and frequency of follow-up appointments are different for each patient and surgeon, but, as a general rule, immature patient with larger curves should be followed closely (every 4 to 6 months), whereas more mature patients with smaller curves can be followed with less frequency.

Assessing maturity with regard to growth remaining is an important factor that may influence potential treatment. In addition to the Risser sign and the status of the triradiate cartilage, hand x-ray for bone age and use of the Sanders epiphyseal staging method may prove useful and may be more accurate than pelvic findings. The peak height velocity is the time when risk of curve progression may be greatest, but unfortunately many commonly used methods for skeletal maturity assessment such as the Risser sign are not visible until after the peak height velocity has passed. Other systems such as assessing the ossification of the proximal ulna has been shown to be useful prior to peak height velocity, however this tool has not been widely accepted.

During surgical planning, in addition to standard PA radiographs, a lateral x-ray of the spine and bending films is obtained. The lateral will be used to assess thoracic kyphosis and overall sagittal balance. It is also possible to make commentary on rib-hump deformity from the lateral radiograph, but this is more reliably viewed during the physical examination. Bending films assess the flexibility of a given curve. The patient will bend into the curve (often over a bolster) and the radiograph is taken. Curves that remain greater than 25 degrees on bending films are considered structural in nature.

When following young patients over a long period of time it is worthwhile to consider minimizing radiation exposure and risk either through radiographic technique (low exposure methods and anatomically placed shields) or by obtaining fewer x-rays in stable curves. Data from the 1960s and 1970s suggest a 1% to 2% increase in lifetime risk of breast and thyroid cancer for patients exposed to multiple spinal radiographs during treatment of spinal curvatures. Since that time x-ray protocols and techniques have changed drastically to reduce radiation exposure. New x-ray technology, such as the EOS (Cambridge, MA) machine, is emerging with the capability to decrease radiation exposure to an even greater extent.

Magnetic resonance imaging (MRI) is not required for all cases in the treatment of AIS. MRI should be obtained if the curve is atypical (apex to the left), if there is associated back pain (concern for infection or malignancy), if there has been a heightened pace of curve progression, or if there is any neurologic abnormality (concern for intraspinal process, such as syrinx, tethering, or tumor).

**Treatment**

Patients with scoliosis may be observed, treated nonoperatively, or treated with surgical intervention. Contributing factors in decision-making are curve magnitude, patient age and maturity, and curve progression. In most treatment centers, curves greater than 20 to 25 degrees are believed to necessitate an increase in monitoring and treatment. A general treatment algorithm is presented in Table 22.2.

**Nonoperative Treatment**

**Observation.** Patients who have a spinal curvature that is less than 25 degrees can be serially observed either by their primary care physician or orthopaedist. In general, curves smaller than 10 degrees can be followed clinically without radiation exposure from serial radiographs. Patients with true scoliosis (curve >10 degrees) should be monitored clinically and radiographically, particularly immature patients. This group of patients should be evaluated every 4 to 6 months until such time as a decision for surgical intervention is made, or until they reach skeletal maturity.

**Bracing.** Historically, there has been controversy in the brace management of scoliosis secondary to a dearth of reliable data in the bracing literature. The data are confounded by use of different brace types, materials used, hours in-brace prescribed, quality of brace fabrication, and general opinion of bracing by the treating physician. In 2013, the BrAIST results were

<table>
<thead>
<tr>
<th>Curve Magnitude (Degrees)</th>
<th>Documented Progression &gt;5 Degrees</th>
<th>Treatment</th>
</tr>
</thead>
<tbody>
<tr>
<td>≤25</td>
<td>No</td>
<td>Observe</td>
</tr>
<tr>
<td>≤25</td>
<td>Yes</td>
<td>Brace</td>
</tr>
<tr>
<td>25–45</td>
<td>No</td>
<td>Brace</td>
</tr>
<tr>
<td>25–45</td>
<td>Yes</td>
<td>Revise brace/consider fusion</td>
</tr>
<tr>
<td>≥45</td>
<td>n/a</td>
<td>Consider fusion</td>
</tr>
</tbody>
</table>
published in The New England Journal of Medicine. This was a prospective randomized clinic trial that confirmed the clinical efficacy of bracing using a Boston style thoracolumbosacral orthosis (TLSO) brace (Fig. 22.5). Another important finding from the BRAIST data was the prescribed (and worn) hours of bracing; patients who wore the brace >13 hours per day did not require posterior spinal fusion (PSF) 90% of the time, while patients who wore the brace <6 hours per day had similar surgery rates as patients treated with observation alone.

Bracing should be considered for skeletally immature patients with curves that have reached a Cobb angle of 25 degrees or, in some centers, measure 20 degrees and have shown to be progressive. Brace wear is not offered to patients who are skeletally mature because, although, braces prevent progression of scoliosis, they do not correct scoliosis in the mature patient. Several types of braces have been described and prescribed, but the most commonly used current brace is a TLSO with small variations based on their place of origin (Boston, Wilmington, Miami) (Fig. 22.5). This style of brace can prevent progression of curves with an apex below the T7 vertebral level. Curves with a more proximal apex are better managed by a cervicothoracolumbosacral orthosis (CTLSO) like the Milwaukee brace (Fig. 22.6) designed by Walter Blount in 1946. This style of brace is rarely prescribed as compliance is quite poor with modern adolescents. Most comparative studies between full-time bracing and nighttime bracing (Providence, Charleston) have demonstrated inferior results of nighttime bracing, although these studies generally are not high quality investigations.

Newer fabrication methods and materials have led to thinner, more breathable braces that can often be worn under than patient's clothing. There are many opinions on the prescriptive hours of brace wear that should be encouraged. Data from the BRAIST study exhibit a considerable increase in efficacy with brace wear greater than 13 hours per day. Ideally, a brace is worn as much as possible.

Bracing should be continued until the patient has reached skeletal maturity clinically and radiographically. During treatment with a brace, patients should be evaluated every 4 to 6 months with clinical and radiographic examination. In patients who have curve progression to 50 degrees despite brace wear, surgical stabilization should be considered.

**Surgical Intervention**

Indications for surgery differ in each practice but general principles include the following:
Curve progression despite appropriate brace wear
- Curve greater than 45 degrees in a skeletally immature patient
- Curve greater than 50 degrees in a skeletally mature patient

These are not established rules and will often be broken based on surgeon experience, patient input, and sagittal profile. Many patients with well-balanced 50-degree curves will defer surgery, as the cosmetic appearance is acceptable in their eyes whereas, many thin patients with unbalanced 40-degree curves will ask for surgical intervention. Each patient must be treated individually as there is a great deal of emotional and social psychology associated with body morphology, brace wear, and spinal fusion.

Once it has been mutually agreed upon that a patient will have spinal fusion for scoliosis, the treatment goals include the following:
- Prevention of further curve progression
- Coronal, sagittal, and axial plane realignment
- Formation of a fusion mass
- Achieving the above goals without neurologic compromise or other complication

The most common method of achieving these goals is PSF with use of instrumentation. Current popular instrumentation systems include rods that are affixed to the spine with wires, hooks, and pedicle screws. Correction of scoliosis can also be performed with anterior spinal fusion (ASF) techniques and a combination of ASF and PSF.

**Posterior Spinal Fusion.** In the technique for PSF a subperiosteal dissection is performed and excision of the facet joints at the proposed fusion levels. The original described technique involved the placement of bone graft and then casting the patient in a corrected position until fusion takes place. This treatment left adolescent patients in body casts for months at a time and still had an unacceptable rate of pseudarthrosis. In the early 1960s, in Houston, Texas, Paul Harrington introduced the Harrington rod system for PSF. By fixing rods to the spine with hooks Harrington was able to correct deformity and immobilize the spine during the time that a fusion mass was forming. Many iterations of wire, hook, and rod systems have been used, but the greatest advances since the original Harrington instrumentation have come from the use of transpedicular fixation. Pedicle screws (Fig. 22.7) allow for fixation in all three columns of the spine and have superior pullout strength and ability to maintain fixation during aggressive reduction maneuvers. The three-column fixation of pedicle screw constructs has allowed surgeons to manipulate

![Figure 22.7](image-url) Preoperative and postoperative x-ray of a patient with 63-degree Lenke 1a curvature who underwent segmental posterior spinal instrumentation for a selective thoracic fusion.
axial plane deformities as well as the coronal and sagittal deformities that were corrected with older instrumentation systems.

There is currently controversy in choosing fusion levels for different curve types, but there is a general consensus that it is best to fuse the fewest levels possible while attaining a balanced spine. Structural curves (those with bend film Cobb measurements >25 degrees) should always be fused, but many secondary curves are compensatory in nature and can be left unfused. This is the nature and reasoning behind selective fusion constructs (Fig. 22.7). There is no current long-term data on PSF with current fixation strategies. There is a general belief that the moment arm of long fusions (thoracolumbar fusion to the level of L4 or L5) will lead to a preponderance of distal segment disk disease and pain. For this reason it is a reasonable strategy to make attempts to stop fusion short of these segments in the idiopathic patient.

**Anterior Spinal Fusion.** ASF techniques have decreased in popularity over the last decade with the onset of three-column fixation in the posterior spine. Anterior fusion can be used to correct single thoracic, lumbar, or thoracolumbar curves, but cannot be used for double major curve types (Fig. 22.8). ASF can be performed through an open incision or thoracoscopically/laparoscopically. There is a steep learning curve associated with scope-assisted techniques and extra training or assistance from a knowledgeable colleague is recommended when beginning to use this approach. Because ASF can increase kyphosis, it should not be used for patients with excessive preoperative thoracic kyphosis, and it must be used cautiously when trying to preserve or enhance lordosis. All levels within the curve should be fused; instrumentation and fusion should extend from the transitional neutral vertebra above to the transitional neutral vertebra below the curve. This extension may allow a shorter segment of fusion than PSF, which

**Figure 22.8** Preoperative (A) and postoperative (B) x-rays of a patient treated with anterior instrumentation and fusion for scoliosis. This thoracic curve showed hypolordosis, which was corrected by the kyphogenic anterior instrumentation.
is preferable in many young athletes who wish to keep as much flexibility as possible. ASF can be accomplished through an open thoracotomy, retroperitoneal approach, anterior thoracolumbar approach, or endoscopically. After disk removal and structural bone grafting at each level (with autograft ribs, allograft, or cages), a bicortical screw is placed in each vertebral body, and a single rod is used for correction. Newer systems use double screws at each level and double rods to improve stability and possibly to decrease the pseudarthrosis rate, which has been shown to be higher with anterior fusions. A double-rod system also may obviate the need for postoperative bracing. Anterior tethers and staples are not yet proven but may offer promising opportunities for smaller curves.

**Combined Anterior and Posterior Spinal Fusion.** Reasons for combined ASF and PSF in idiopathic scoliosis currently include the following:

- Large, stiff curves
- Operative curves in skeletally immature patients (Risser 0 or 1) in whom there is concern for crankshaft phenomenon

In curves with a high Cobb angle and little to no correction on bend films (no flexibility) anterior release can be used to create greater flexibility. This can be performed through either open or endoscopic techniques. In skeletally immature patients who undergo PSF, the anterior spine can continue to grow and create a crankshaft phenomenon as the anterior spine rotates around the posterior fusion mass. Young patients (Risser 0 or 1) who are at risk of developing problems from the crankshaft phenomenon frequently undergo a combined ASF and PSF. This procedure can be done in a single setting or staged days to months apart, depending on the condition of the patient and surgeon preference.

**Surgical Risks.** The most devastating complication associated with scoliosis surgery is neurologic damage, ranging from a mild neuropathy to paraplegia. Intraoperative monitoring during surgery can help protect a child from permanent injury. Most centers use motor-evoked potential (MEP) or somatosensory-evoked potential (SSEP) monitoring intraoperatively. MEPs monitor the motor pathways and are more sensitive and reliable than SSEPs, which monitor only the sensory pathways in the dorsal columns of the spinal cord. The Stagnara wake-up test is the gold standard for monitoring motor function if there is any intraoperative concern; this allows alteration of hardware or curve correction with the patient still under anesthesia.

Surgery can be complicated by early or late infection. There also is a risk of pseudarthrosis, hardware failure, superior mesenteric artery syndrome, syndrome of inappropriate antidiuretic hormone, and postoperative back pain. As the patient ages, there is a concern for degenerative changes proximal and distal to the levels fused.

**Postoperative Management.** Postoperative plans can vary depending on the patient, surgeon, and treatment facility. After posterior fusion, with good fixation and bone quality, most patients do not require bracing. ASF alone frequently requires external support with a custom-molded TLSO for up to 12 weeks postoperatively. In many institutions, patients are mobilized the day after surgery. If bracing is necessary, the patient is molded and fitted with the brace before discharge from the hospital. Patients with spine fusion for AIS typically are discharged by postoperative days 3 to 5. They are encouraged to walk as much as tolerated for the first 6 weeks, with no lifting, twisting, or bending allowed. After 6 weeks, activity usually is progressed to jogging and bicycling. After 6 to 12 months, if x-rays show adequate fusion, patients are allowed to return to noncontact sports. It is recommended that patients avoid contact sports for at least another year.

**Scheuermann Kyphosis**

Scheuermann kyphosis, also known as Scheuermann disease, was originally described in 1919 and refers to a rigid juvenile or adolescent kyphosis with wedging of the vertebral bodies over the affected segments in the thoracic spine. The apex of the kyphosis is typically located between T7 and T9. It is often associated with back pain and is believed to be caused by a growth disturbance at the end plates of the vertebral body. Much like AIS, the age of onset of Scheuermann kyphosis is most often during the prepubertal growth spurt and clinical detection is typically shortly after this. It is difficult to diagnose Scheuermann kyphosis before the prepubertal growth spurt as the characteristic vertebral wedging cannot be detected radiographically until the ring apophysis is ossified (approximately 10 years).

**Relevant Anatomy**

The standard complement of spinal gross anatomy is present in patients with Scheuermann kyphosis. Anatomic changes include anterior vertebral wedging with irregularity of end plates. These changes can be seen on gross specimens, or, in the living patient, on radiographic examination. The criteria for diagnosis of Scheuermann kyphosis are more than 5 degrees of wedging on three or more adjacent vertebral bodies, and/or a rigid kyphosis in a patient meeting the age and maturity level associated with this type of kyphosis.

**Pathogenesis**

Much like idiopathic scoliosis, the exact etiology of Scheuermann kyphosis is unknown. Genetic, vascular, and hormonal disturbances have been put forth as the cause of Scheuermann kyphosis, but none of these theories, on their own, has stood up to rigorous
experimentation. Scheuermann originally hypothesized that aseptic necrosis of the ring apophyses led to an arrest of growth and anterior wedging of the vertebrae, but this hypothesis has not been confirmed and is unlikely, as the ring apophysis does not contribute to longitudinal growth of the spinal column. Schmorl and Junghans noted the radiographic finding of Schmorl nodes and hypothesized that a weakening of the cartilaginous end plate allowed the intervertebral disk to penetrate the bone and disrupt normal growth. It since has been shown, however, that Schmorl nodes are present in many different types of spinal deformity and are not limited to Scheuermann kyphosis. Furthermore, they are often present in completely asymptomatic patients. At present, the most widely held belief is that this type of kyphosis is secondary to the upright, bipedal posture of the different hominid species. This is supported by the fact that no cases of similar wedging have been found in the spine of quadrupedal or quadrumanous species. Lambrinudi proposed these effects of upright posture with his observation that many patients had a tight anterior longitudinal ligament. This would create a mechanical force at the anterior aspect of the vertebral body and effect growth following the Huetter-Volkmann principle, much like current growth modulation techniques.

Scheuermann kyphosis is associated with several disease processes, including endocrine abnormalities, hypovitaminosis, inflammatory disease, neuromuscular disorders, dural cysts, and spondylolysis, but no direct cause-and-effect relationship between these conditions and Scheuermann kyphosis has been established.

Irrespective of the true etiology, it is known and accepted that there are histologic alterations at the endochondral ossification centers of the affected vertebrae. It is not known whether these ossification center changes are secondary to Scheuermann disease or are the cause of the kyphotic deformity. It is likely that there is a complex interaction of hereditary and environmental factors that result in kyphotic deformity. Further studies need to be done to determine more definitively the etiology and biology of Scheuermann kyphosis.

**Epidemiology**  
A range of incidence from 0.4% to 10% has been reported with varying rates of male-to-female ratios. As previously stated, age of onset is during the prepubertal growth spurt and diagnosis is most often after 10 years of age. An associated mild scoliosis is noted in 20% to 30% of patients, but this lateral curve rarely progresses to require treatment. Progression of kyphosis has been documented during the growth spurt and later in adult life. In contrast to idiopathic scoliosis, the risk for kyphosis progression currently is unknown and warrants further study. Data reported by Lowe, Conti, Travaglini, and Murray all suggest that the natural history is fairly benign, except for a known increase in back pain. Patients with thoracic kyphosis below 100 degrees actually have an increase in pulmonary function due to the increase in chest circumference. Once there is progression past 100 degrees of kyphosis, patients will exhibit restrictive cardiopulmonary pathology.

**Classification**  
Scheuermann kyphosis is divided into typical and atypical forms. The typical form is classically seen and has been described in the above sections. Atypical Scheuermann kyphosis is a deformity at the thoracolumbar junction or in the lumbar spine. Atypical forms of Scheuermann kyphosis are much less common and rarely require surgical intervention.

**Diagnosis**

**History**  
The kyphotic patient may present to the clinician’s office shortly after onset of kyphosis, in the peripubertal years, or anytime thereafter as deformity and pain dictate. Patients with kyphosis have a wide range of body morphology and body mass index; therefore, cosmetic deformity will be evident at a lower degree of kyphosis in some patients. In patients who have not yet developed pain at the level of kyphosis or in the lower back, the presenting symptoms is often poor posture or rounded shoulders that have alarmed the parents or primary care giver. Patients who present with pain will commonly complain of pain at the apex of the deformity as well as in the lower back; particularly if there is a concomitant increase in lumbar lordosis. In a study by Murray, patients with Scheuermann kyphosis between 65 and 85 degrees had the worst pain on validated questionnaires.

In addition to the standard history (age, gender, past medical history, and family history), one should determine maturity: age at menarche for girls, history of a growth spurt, current patient height, and the height of first-degree family members. Further assessment includes evaluation of pain or any neurologic symptoms.

**Physical Examination**  
The examination should start with an assessment of maturity based on secondary sexual characteristics and height compared to parents and older siblings. The skin should be examined for café au lait spots, axillary and inguinal freckling (which may indicate neurofibromatosis), and signs of dysraphism (skin markings, dimpling, hair on the back). The adolescent should be examined for asymmetry of shoulders, pelvis, and muscular development. Tightness of the hamstrings is common and can be evaluated with the straight-leg raise test or forward bending test. The patient should be examined for hip flexion contracture or fixed pelvic tilt with the Thomas test. A neurologic examination needs to be done on every patient with a spinal deformity. Neural compression has been reported from ruptured thoracic disk, intraspinal extradural cyst, and mechanical cord compression at the
apex of the kyphosis. These are incredibly rare findings, but necessitate a thorough neurologic examination.

Any spinal deformity should be examined and understood in the coronal, sagittal, and axial planes. Asymmetry of the shoulders and trunk can be a sign of coronal plane abnormality. The Adam's forward bend test can be used to assess coronal and axial deformities when viewing a patient from behind, and can assess sagittal deformity/kyphosis when viewed from the side. Tests of thoracic hyperextension, either standing or on an examination table, can give the examiner information on the rigidity of the curvature. True Scheuermann kyphosis will be rigid and not correct with hyperextension, but very immature patients may have more flexible, early forms of Scheuermann kyphosis.

**Radiographs**

The standard radiographic evaluation of kyphotic patients should include PA and lateral films shot on a full-length (36-in) cassette. Normal thoracic kyphosis is described as 20 to 45 degrees with normal lumbar lordosis being 40 to 60 degrees. The following features of Scheuermann kyphosis are seen on the lateral radiograph (Fig. 22.9):

- Anterior wedging (>5 degrees) of three or more consecutive vertebral bodies
- Increased kyphosis (>45 degrees), which can be measured by the Cobb method on the lateral x-ray
- Irregularity of vertebral end plates
- Schmorl nodes—depressions in the vertebral bodies that represent disk herniation into the end plate

Bradford suggests that anterior wedging of three or more consecutive bodies is not necessary for diagnosis if the patient has a rigid kyphosis on clinical examination. If flexibility is in question, or for preoperative planning, bending-type films with the back over a bolster (placed at the apex) can be obtained.

In 20% to 30% of patients, the PA x-ray shows an associated mild scoliosis in the area of the kyphosis. The scoliotic apex usually corresponds with the kyphotic apex. A lateral x-ray should be examined for spondylolisthesis in addition to kyphosis. In the later stages of Scheuermann kyphosis, x-rays often reveal changes consistent with degenerative arthritis, including decreased intervertebral disk spaces, marginal osteophytes, and ankylosis. MRI or CT can be performed if the patient has unusual symptoms, positive neurologic findings, or there is concern of disk protrusion/extrusion at the apex of the curvature.

**Treatment**

The reasons to initiate treatment in Scheuermann kyphosis are pain, cosmesis, progression of deformity, and neurologic or cardiopulmonary compromise. The most common of these are pain and cosmesis.

**Nonoperative**

There are several nonoperative options for the treatment of Scheuermann kyphosis and they are listed below:

- Serial observation
- Physical therapy with stretching and strengthening
- Bracing

Serial observation is an appropriate treatment method for immature patients with smaller, flexible curves. Many times serial observation and examination is combined with physical therapy and exercise regimens. Therapy regimens aim to strengthen back and core musculature while stretching the hamstring muscles. No study has clearly shown that this is an efficacious treatment, but, in theory, it can work to decrease lumbar lordosis. It is

![Figure 22.9](image)
less clear if stretching and therapy have any effect on a rigid kyphosis.

Bracing is the mainstay of nonoperative management in Scheuermann kyphosis. Because Scheuermann kyphosis is not a life-threatening problem until severe deformity is present, all patients with mild-to-moderate kyphosis (<75 degrees) should have a trial of conservative treatment to control symptoms and minimize deformity. A kyphosis brace applies three-point bending forces that decrease thoracic kyphosis. In skeletally immature patients with thoracic kyphosis greater than 45 degrees and less than 75 degrees, bracing can be considered. Attempts to brace curves greater than 75 degrees have led to a high failure rate, and this is not recommended. To influence a kyphotic deformity, a CTLSO or modified Milwaukee brace (Fig. 22.6) provides the best extension force, but wear compliance with this brace tends to be poor. More cosmetically acceptable underarm braces commonly are used now, but these require expert orthotic design and alterations to be effective. This type of brace is ideally used for curves with an apex at or below the T9 level.

There are mixed reports regarding brace efficacy in the literature. Acute application of a brace can influence the deformity and improve kyphosis by 40% to 50%; however, several series have shown at least partial loss of this correction when brace wear is stopped. It is recommended that a brace be worn full time (23 hours per day) for 12 to 18 months, then part time to full time (depending on the severity of the kyphosis) until the patient reaches skeletal maturity. If a teenager strongly refuses to wear a brace out of the house (a frequent reaction in a peer-conscious age group), a vigorous exercise program combined with nighttime brace wear can be considered. All kyphosis braces require careful orthotist attention to ensure fit and to recontour the posterior bars and pads every 2 months to gain further correction progressively. There is no indication to brace skeletally mature patients.

Operative
Operative intervention for Scheuermann kyphosis can be in the form of PSF, ASF, or a combination of the two. The principles of surgical correction are based on lengthening the anterior column, shortening the posterior column, or both. Changes in implant and instrumentation systems have made great strides in decreased rates of pseudarthrosis. Other than physiologic compromise, neurologic or cardiopulmonary, there are no gold standard indications for surgical intervention in Scheuermann kyphosis. Each patient and encounter must be treated individually based on the patient’s clinical examination, radiographic parameters, pain, and cosmesis. Tribus suggested the following five reasons to consider surgical treatment in thoracic Scheuermann kyphosis:

- Pain
- Progressive curve
- Neurologic compromise
- Cardiopulmonary compromise
- Trunk deformity

Rarely, thoracic disk herniation, epidural cysts, or a severe kyphosis (>100 degrees) can cause neurologic deficit in patients (usually adults) with Scheuermann kyphosis. Neurologic deficits are the only absolute indication for surgery. Relative indications include kyphosis greater than 75 degrees and kyphosis greater than 60 degrees associated with pain that is not alleviated by nonoperative measures. The goals of surgery include correction of the deformity and relief of pain. Surgical correction of the deformity always includes spinal fusion.

**Posterior Spinal Fusion.** PSF for kyphosis (Fig. 22.10) is a good option in curves that can be corrected to <50 degrees and held in place with instrumentation. Previous hook- and cable-based constructs could not provide the needed structural support for fusion without progression of curvature. These systems had an unacceptably high rate of pseudarthrosis and also required postoperative immobilization. With the introduction of thoracic pedicle screws by Suk we are able to control all three columns of the spine and provide the necessary support for sustained correction and fusion. In concert with instrumentation is the use of posterior spinal osteotomy techniques, such as the Ponte osteotomy. This osteotomy involves excision of the ligamentum flavum with osteotomy of the inferior and superior facets. Up to 10 degrees of correction at each level can be achieved with Ponte osteotomies (Fig. 22.11).

**Anterior Spinal Fusion.** Anterior release with instrumentation and fusion for kyphosis was originally described by Kostuku. In the procedure an anterior interbody fusion is performed at affected levels with Harrington-type instrumentation. Patients would then need supportive bracing postoperatively. The technique is not widely used despite good results reported by Kostuku.

**Combined Anterior and Posterior Spinal Fusion.** Anterior release and fusion at the apex of the deformity in addition to posterior fusion have provided significantly better results with regard to immediate and long-term correction of large kyphotic deformities and now is the standard of care for large, rigid deformities. The anterior procedure can be done open or endoscopically before the posterior instrumentation. It is worth mentioning, once again, that endoscopic/thoracoscopic techniques are technically challenging and have reported complication rates of up to 50% in some studies.
Fusion Levels
Debate remains regarding selection of fusion levels in Scheuermann kyphosis. It is clear from the literature that too short a fusion leads to junctional kyphosis developing at the proximal and distal ends of the rods. The criteria to determine the fusion levels in Scheuermann kyphosis are not as well established as they are for scoliosis. Current recommendations are to include the proximal end vertebra (determined by the modified Cobb method) and to extend the fusion past the transitional zone to the first lordotic disk distally. Lowe also recommended limiting correction of the kyphosis to 50% of the original deformity or less to prevent junctional kyphosis. Overcorrection should be avoided. Patients with lumbar Scheuermann kyphosis almost never require surgery.

Risks of Surgery
Reported complications of surgical correction of Scheuermann kyphosis include death, gastrointestinal obstruction, hardware failure, pseudarthrosis, progression of the deformity, hemothorax, pneumothorax, pulmonary emboli, neurologic injury, infection, and persistent postoperative back pain. The most feared complication is neurologic injury, including paralysis. Vascular insults to the cord and mechanical damage have led to paraplegia. Correction of kyphosis carries a higher than usual risk of neurologic injury, which is related directly to the amount of correction. Intraoperative neurologic monitoring is crucial during any surgery to correct kyphosis because the thoracic cord is at risk during correction and instrumentation. MEPs and SSEPs are used in most spine centers. The Stagnara wake-up test is the gold standard for motor monitoring if there are any concerns during surgery. If monitoring or the wake-up test indicates a neurologic deficit, any corrective maneuvers should be reversed.

There are significant risks to surgical correction of thoracic kyphosis that must be weighed carefully when considering surgery because the natural history of mild-to-moderate Scheuermann kyphosis still is not yet well defined. Corrective kyphosis surgery has benefited immensely from new developments in spine instrumentation (e.g., pedicle screws, in situ bending). Perhaps even more than idiopathic scoliosis, surgical management of Scheuermann kyphosis requires treatment by experienced spinal deformity surgeons.

Postoperative Management
Previously, most patients undergoing correction of Scheuermann kyphosis were braced for 3 to 6 months.
Figure 22.11  Ponte osteotomy to increase flexibility in the spine. A: First a facetectomy is performed removing the caudal facet from the superior vertebra exposing the articular surface. B: The spinous process including the intervening supraspinous and interspinous ligament is removed. C: The superior portion of the superior facet as well as the capsule is removed. D: The posterior elements are no longer connected across the released segments, thereby increasing the flexibility across the deformity. (Reprinted with permission from figure Lovell and Winter's pediatric orthopaedics seventh edition, Wolters Kluwer Health, Figure 17.29.)
With modern fixation techniques, like pedicle screws, the need for bracing is largely obviated. But, some patients with poor bone quality, rigid curves, or fewer points of fixation will require postoperative bracing. Patients typically are mobilized the day after surgery to help prevent atelectasis and gastrointestinal obstruction. They usually are hospitalized for 3 to 5 days and are encouraged to ambulate several times a day for the first 3 months. No bending, stooping, or lifting is allowed for at least 3 months. No sports are allowed for the first year after surgery. After 1 year, patients may return slowly to normal activities if x-rays show consolidation of the fusion and minimal progression of the kyphosis.

**SUGGESTED READING**


