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A review of the evaluation, diagnosis, and nonsurgical treatment of adolescent idiopathic scoliosis

Brandon Raudenbush, DO,^a Ashley Simela, DO,^a Hans Joseph, DO^b

From ^aOrthopedic Surgery Resident, University Hospital's Richmond Medical Center, Cleveland, OH; and ^bOrthopedic Surgery Resident, Millcreek Community Hospital, Erie, PA.

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Adolescent Idiopathic Scoliosis is a relatively common orthopedic condition, affecting approximately 0.5%-3.0% of the pediatric population. Although the condition is often self-limiting, the consequences of a late or delayed diagnosis, coupled with the potential for rapid progression of the deformity, make the detection and treatment of AIS critical for a satisfactory patient outcome. The scope of this article is to review the natural history, etiology, diagnostic workup, and nonoperative treatment options available for Adolescent Idiopathic Scoliosis.

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Introduction

Idiopathic Scoliosis (IS) is defined as a lateral curvature of the spine greater than 10⁰, measured via the Cobb method. To be defined as idiopathic, the vertebral rotation must occur in the absence of any congenital spinal anomaly or an associated neuromuscular condition.³⁻⁷ Although defined as a lateral or coronal curvature, scoliosis is in fact a 3-dimensional phenomena and deformity. Through spinal motion coupling, not only is scoliosis a lateral spinal curvature, but it is also associated with vertebral rotation as well as flexion and extension.⁸⁻¹⁰

A lordosis or hypokyphosis at the apical vertebra in the thoracic spine is seen as the initial insult. Adams, an English physician and surgeon, was the first to describe this in 1865 while performing autopsies on patients with scoliosis.¹⁰ He observed that the rotational prominence, or rib hump, was made worse by forward bending (Adams Forward Bend Test).

Corresponding author: Brandon Raudenbush, DO, Orthopedic Surgery Resident, University Hospital's Richmond Medical Center, Cleveland, OH 44143.

E-mail address: brandonraudenbush@yahoo.com.

Scoliosis: Classifications

Scoliosis is divided into 3 broad categories: *Idiopathic*, *Congenital*, and *Neuromuscular*.^{11,12} IS was classified according to the age of onset by James in England in 1954.¹³ These include *Infantile* (birth to 3 years), *Juvenile* (4-9 years), and *Adolescent Idiopathic Scoliosis* (AIS) (> 10 years old).^{5,13,14} IS is the most common form of scoliosis, comprising close to 80% of cases. It is distinguished from the other 2 major types of scoliosis, congenital and neuromuscular, by its unknown etiology.

Congenital scoliosis occurs as a result of abnormal growth and development of the vertebral column, likely due to intrauterine events at or about the sixth week of gestation.¹⁵ Neuromuscular scoliosis is often identified in pediatric patients with neurologic and myopathic diseases, such as cerebral palsy, among others.¹⁶ It is characterized by an early onset with rapid progression even after skeletal maturity. The curves are often long, extend into the sacrum, and have an associated pelvic obliquity^{11,16-18} (Figure 1).

A fourth category of scoliosis is occasionally referred to in some texts, called a *functional* scoliosis. This encompasses conditions causing a *relative* scoliosis including postural changes secondary to muscles spasms and short leg

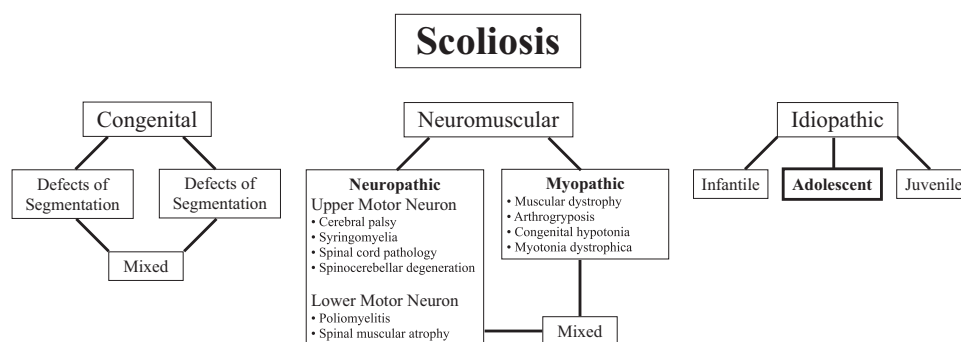


Figure 1 Scoliosis classification.

syndrome, among others, and is not truly due to an abnormality of the spinal column.^{1,19,20}

In recent years, the classification for IS has changed slightly. More emphasis has been placed on pulmonary development in relation to the onset of scoliosis, with the threshold of 5 years being critical for lung development.^{7,21-23} This has led to reclassifying IS as either *early onset* (before age 5) or *late onset* (after age 5), with AIS falling into the latter category.²³

Etiology, epidemiology, and natural history

Several theories exist as to the etiology of AIS, however none have been proven definitively. The role of genetic factors has been widely accepted, but the exact mode of inheritance and specific gene locations have not been determined.²⁴⁻²⁶ Due to an increased risk among first-degree relatives of patients with IS, as well as the high concordance rate among monozygous twins, it appears some form of autosomal inheritance pattern with incomplete penetrance is occurring.²⁷ A positive family history for scoliosis is observed in anywhere between 10%-30% of patients referred for medical evaluation.^{1,14,27}

A DNA-based test that incorporates 53 genetic markers has recently become available in late 2009 to help determine those patients who are at higher risk of curve progression.²⁸ The test is not fully incorporated into most orthopedic practices, however, but hopes are that it would help avoid costly follow-up visits and excessive radiation exposure to those at a lower risk of progression.²⁹ More research is being conducted in hopes to perfect future treatment algorithms using genetic testing such as this.

AIS comprises the vast majority of the 'idiopathic' cases of scoliosis. It is estimated that Infantile Idiopathic Scoliosis accounts for only 0.5% of all cases, and that for Juvenile Idiopathic Scoliosis, it ranges from 8%-16% of cases. Interestingly, the male-to-female ratio of mild curves in AIS is approximately 1:1. However, for larger and more significant curves, females predominate males by a ratio of 6:1, and according to some literature, upward to 8:1.^{30,31} Although there is no complete agreement, most clinicians view *significant curves* as those that are greater than 40° at skeletal maturity, or greater than 20° if the patient is skeletally immature.^{1,31}

The term *progression* in scoliosis refers to an increase in curve magnitude of greater than 5° from the original presenting curve. This is due to the measurement error of 3-4° that can occur with the Cobb method. With the prevalence of IS estimated to be only 0.5%-3%, the actual number of patients that would go on to develop a significant curve is even smaller, roughly 0.1% or 1 in every 1000 AIS patients.^{1,2} This is a substantially lower risk than that seen in Juvenile Idiopathic Scoliosis, which carries approximately a 70% chance of requiring some form of treatment.⁵

Risk factors that have been well associated with curve progression in IS deal primarily with indicators of skeletal immaturity. It has been documented that periods of rapid growth in adolescence, specifically the *peak height velocity* of the growth phase, can potentially lead to great increases in curve magnitude, thus requiring strict and periodic observation by the physician.^{1,30-34} Other risk factors for progression include the initial Cobb angle at presentation, curve pattern, vertebral rotation, and most importantly, female gender.^{1,30-33,35,43} (Figure 2).

School screening programs

In the last fifty years, screening programs for scoliosis have been implemented in the hopes of early diagnosis and treatment to decrease curve progression and severity. These programs were first instituted in the 1960s.³¹ The utility of these screenings has been called into question.^{44,45} Specifically, the United States Preventive Services Task Force issued an update in 2004, giving a grade D recommendation for routine screening (recommends against routine screening),

Risk Factors for Progression
Female Gender
Initial Curve Magnitude
Apical Vertebral Rotation
Curve Pattern
Bone Maturity
• Tanner Stage
• Age of Menarche
• Risser Grade
• Peak Height Velocity
• Open vs. Closed Triradiate Cartilage

Figure 2 Risk factors for curve progression.

feeling that there has been an overreferral to specialists.⁴⁶ This was a change from the United States Preventive Services Task Force 1996 recommendation that was inconclusive and did not favor for or against screening.⁴⁶

However many experts and expert panels disagree, including those from the American Academy of Orthopaedic Surgeons, the Scoliosis Research Society, the Pediatric Orthopaedic Society of North America, and the American Academy of Pediatrics.⁴⁷⁻⁴⁹ These societies admit that more standardized research is needed, but recommend continued screening, especially during periods of rapid growth.^{31,48} Screening allows for referral of high-risk patients, and early intervention that can potentially avoid operative treatment and spinal fusion. For this reason, focused or selective screening is recommended at least twice for females (at the age of 10 and 12 years, or in grades 5 and 7), and at least once for males (at the age of 13 or 14 years, or in grades 8 or 9).⁴⁸

Clinical assessment

The clinical assessment of patients with AIS begins with a thorough and detailed history. Close attention should be paid to the patient demographics, including current age, gender, menses status, and the original reason for seeking medical evaluation. The presence of significant back pain, which is not typical in IS, should raise concern.^{50,51} This must be distinguished from the routine aches and pains of musculoskeletal fatigue and mechanical low back pain, the presence of which is only slightly increased in AIS than in the general population of adolescent patients.^{50,52,53} Signs of pathologic back pain require further investigation to rule out potentially devastating conditions such as tumor, infection, or neurologic abnormalities.⁵¹ One must remember that adolescent IS is a diagnosis of exclusion.

Following a thorough history, the physical examination should commence with inspection of the patient, noting any dermatologic findings such as café au lait spots or axillary freckling seen in neurofibromatosis, abnormal hair growth or distribution as seen in spina bifida, or abnormal skin dimpling seen in other spinal dysraphisms (neural tube defects such as syringomyelia).⁵¹

A thorough neurologic examination should be performed and compared with the contralateral side, including gait evaluation, heel-and-toe walking, deep tendon reflex testing, and muscle strength testing. Additional neurologic testing includes the abdominal reflex, Babinski reflex, and other upper motor neuron tests looking for spinal cord pathology.¹⁴ Positive findings with any of these tests require further workup with an MRI of the spine to rule out spinal cord pathology.⁵¹

Inspection of the patient from the posterior to anterior view is performed next. The Adams forward bend test is a quick, easy, and sensitive method that has been used in school screening programs for many years.³¹ This test is used to assess both truncal rotation and sagittal contour. The patient bends forward with the knees fully extended, feet

together, and the palms opposed until the spine is horizontal.^{14,19,54} Truncal rotation can be visualized in this position as well as measured with an inclinometer or scoliometer. Approximately 7° of angle trunk rotation equilibrates to about 20° of coronal deviation (Cobb angle) and justifies referral to a spine specialist with relatively few false-positive results and a reduction in overreferring^{3,31,55} (Figure 3).

Other key areas of the patient that should be assessed during the physical examination include determining the level and position of the occiput, scapula, iliac crests, posterior superior iliac spines, anterior superior iliac spines, greater trochanters, and maleoli. These areas can alert a physician to somatic dysfunctions such as leg-length inequality and short leg syndrome, psoas syndrome and contracture, lumbosacral dysfunction, or sacral shear. All of these conditions can mimic or give the appearance of a lumbar scoliosis, but are not commonly found in AIS.^{20,46}

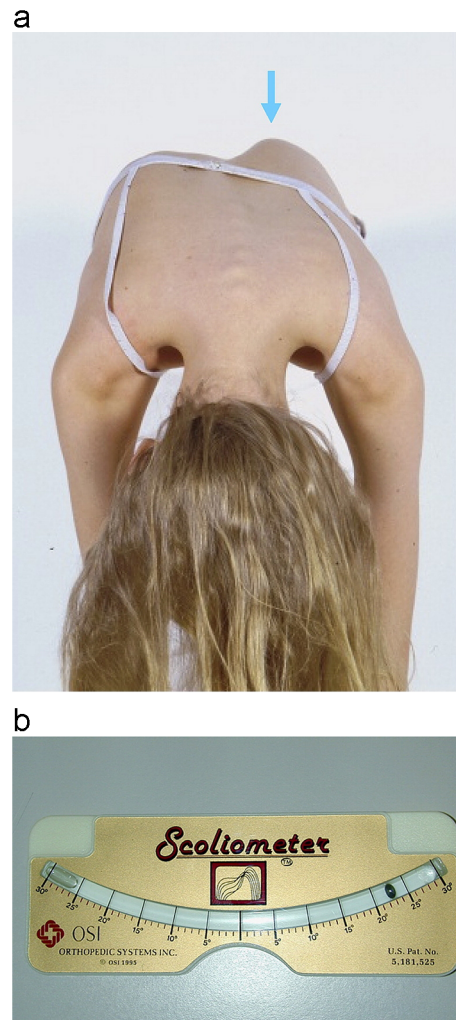


Figure 3 Adams Forward Bend Test (A) The patient bends forward at the waist with the arms relaxed and legs fully extended while the physician inspects from a posterior to anterior view. (B) Scoliometers can be used and are placed at the location of the rib hump; 7° = clinically significant curves that require follow-up.

Radiographic assessment

The definitive diagnosis of scoliosis lies within the imaging studies. Many radiographic measurements are used to assess scoliosis, with more than fifteen parameters having been described in the past.⁴² Most commonly, the radiographic assessment begins with full-length standing PA and lateral radiographic views of the spine on a 14 × 36-in film with the image intensifier positioned 72 in away from the patient and the patient standing erect with the elbows and knuckles rested into the supraclavicular fossa^{56,57} (Figure 4). More commonly, with digital imaging, two 14 × 17-in films are used and ‘stitched’ together electronically.⁵⁶

The Cobb method for measuring the extent of coronal plane deformity is similar to the earlier Ferguson method described in 1930.^{3,58,59} Despite their similarities, the Cobb angle is most widely used today.⁵⁸ This involves finding the angle formed by the intersection of perpendicular lines that are parallel to the superior end plate of the most superior vertebra, and to the inferior end plate of the most inferior vertebra of the curve.^{3,60} It is important to remember to be consistent with all future measurements, and to assess the same vertebral bodies each time (Figure 5 A and B).

The standard PA radiographic view not only allows for the Cobb angle to be determined, but also allows for the curve to be described based on the coronal plane deformity and the extent of the vertebral rotation. Curves are named in the direction of their respective convexity with multiple curve patterns existing, including thoracic, lumbar, thoracolumbar, cervicothoracic, cervical, and lumbosacral. Apical vertebral rotation has been described well in the

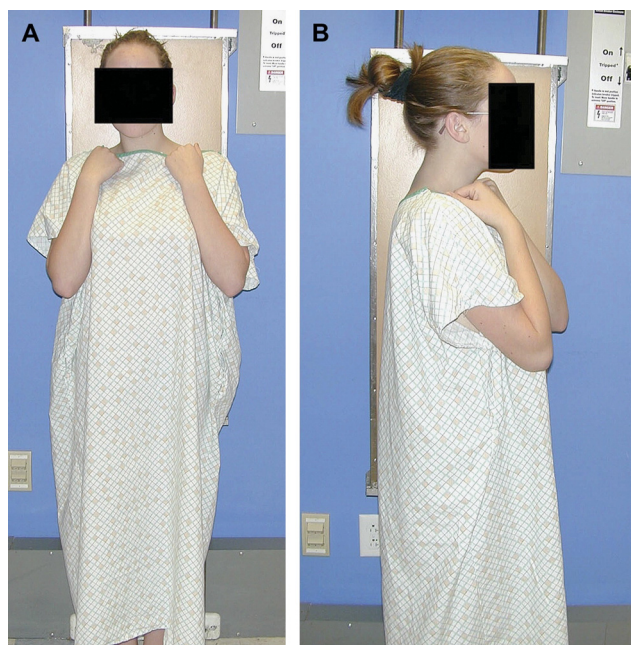


Figure 4 Full-length PA and lateral radiographs of the spine (A) The PA view was traditionally taken with 36-in film with the image intensifier at a distance of 72 in. (B) The lateral view is obtained in the same fashion, with the patient’s hands in the supraclavicular fossa.

past, by Cobb, Nash and Moe, as well as Perdirolle, and also signifies the aggressiveness of the curve and potential for progression^{3,32,43,61,62} (Figure 5C).

In the recent past, the classification system describing curve types and patterns has been changed. The curve characteristics as well as the amount of arthrodesis required for surgically treating AIS has been revolutionized by the introduction of the Lenke classification.⁶³ This newer classification is based on multiple curve characteristics and replaces the older and less reproducible King classification^{63,64} (Figure 6).

The full-length lateral radiograph is used to assess any sagittal plane deformity and for assessing spinal balance.^{14,54} The sagittal plane depicts the amount of kyphosis or lordosis that is in the vertebral column. In AIS, a lordosis or hypokyphosis of the thoracic spine is usually present, particularly at the apex of the curve or apical vertebrae.^{54,65} If there is increased thoracic kyphosis, further evaluation is warranted as this is atypical for AIS and is one of the most sensitive indicators for underlying neuronal disease.^{51,54}

The term *spinal balance* refers to the even distribution of weight over the sacrum and pelvis. This applies to both the sagittal and coronal planes, and is determined by drawing a plumb line in both vertical axes. To determine the coronal spinal balance, a plumb line is suspended from the inion or vertebra prominens to the sacrum. The line should lie within 1 cm of the middle of the sacrum. Plumb lines lying to the left of the center of the sacrum are said to have *negative coronal balance* and those to the right are said to have *positive coronal balance*.⁵⁶

To determine sagittal spinal balance, also called the sagittal vertical axis (SVA), a plumb line is suspended from the center of C7 inferiorly through the sacrum on the lateral radiograph. This line should lie within 2.5 cm of the posterior superior corner of S1. If the line is posterior to the L5-S1 disc space, it is considered *negative sagittal balance*; if it lies anterior to the L5-S1 disc space, it is considered *positive sagittal balance*. The sagittal vertical axis (SVA) should be neutral in the balanced spine^{56,66} (Figure 7).

Skeletal maturity

The iliac apophysis, or Risser sign, has been studied extensively in the past, and is used as a relatively good indicator of skeletal maturity.^{1,30,34,36-40,42,55,67-70} Skeletal maturity guides treatment and is used as an indicator for the risk of curve progression. The iliac apophysis is the secondary ossification center of the iliac crest. Ossification begins at the anterior superior iliac spine and continues along the iliac crest from an anterior to posterior direction. Ossification is completed most posteriorly, at the posterior superior iliac spine. Skeletal maturity can be assessed by reviewing the radiographs already obtained for the Cobb angle measurement and finding a Risser score. The average chronologic age when the apophysis is completed is 14 years in females and 16 years in males^{67,71} (Figure 8).

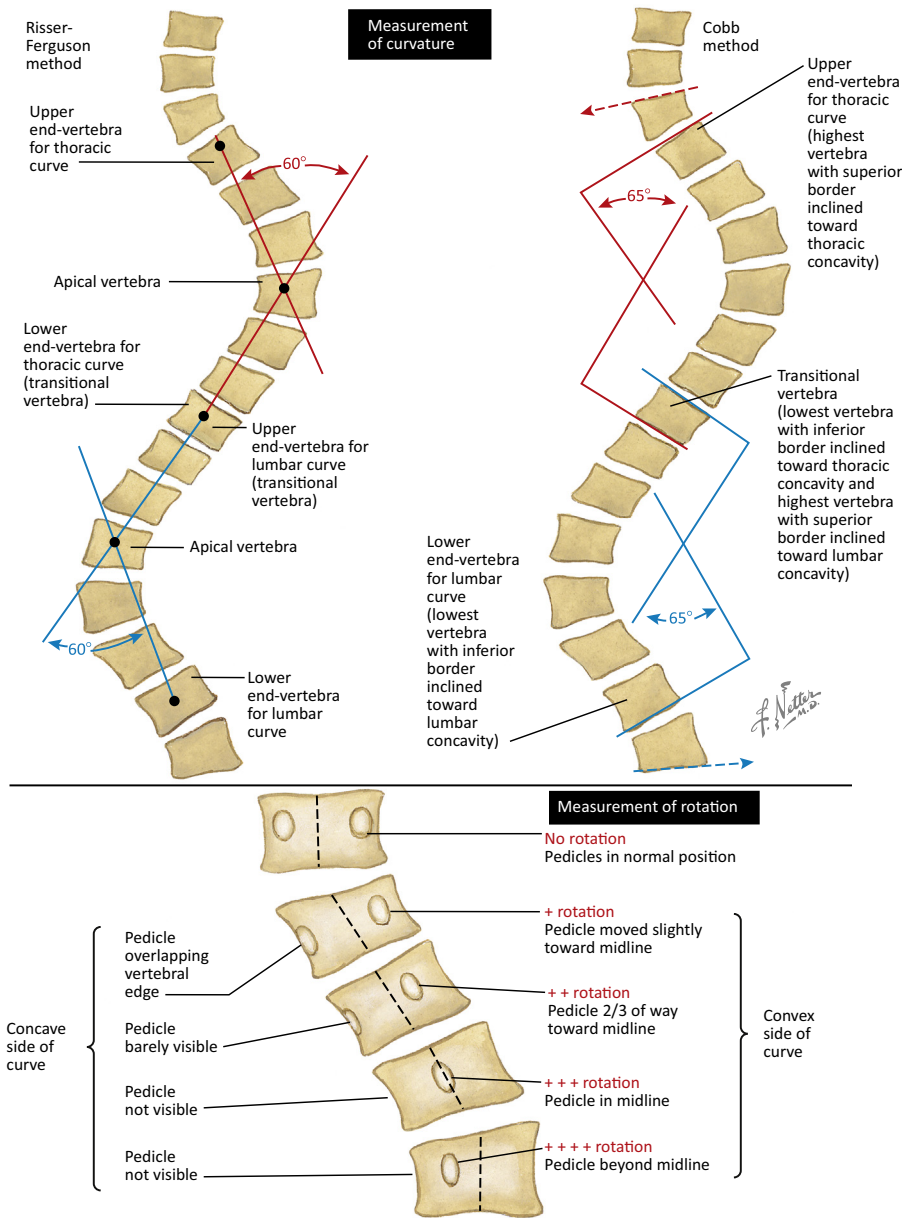


Figure 5 Measuring the curvature and the rotation of the spine (A) The Ferguson method that is no longer typically used. (B) The Cobb method, most popular among clinicians worldwide. (C) Vertebral rotation increases with increasing curve magnitude; it can be noted that as rotation increases, less of the pedicle is visible.

However, the utility of the Risser sign has been called into question as the majority of curve progressions occur at a Risser sign of 0-1, during the patient’s peak height velocity. During this time of rapid growth, the scoliosis can progress dramatically and the Risser sign has been faulted as being too late an indicator of skeletal maturity. Regardless, Risser sign is still considered by many to be a useful radiographic marker of skeletal maturity.⁷⁰

The radiographic appearance of the triradiate cartilage (TRC) is also an indicator of skeletal maturity.^{14,41,42,72} The triradiate cartilage is an epiphyseal growth plate cartilage complex of the 3 secondary ossification centers of the acetabulum; the ilium, ischium, and pubis.⁷³ On average, the TRC fuses between the ages of 11 and 13 years, and is

usually closed shortly before the Risser 1 is first apparent⁷² (Figure 9).

Other methods of determining skeletal maturity, or *bone age*, are used as well.⁷⁴ The Greulich and Pyle method was developed in the late 1950s and is a radiographic atlas of standardized hands and wrists of both sexes, but was limited to Caucasian subjects only.⁷⁵ A newer Tanner-Whitehouse-III RUS method has been published, which studies the radiographic features of the distal Radius, Ulna, and Small bones of the hands, similar to the Greulich and Pyle method.⁷⁶

Some researchers find this method too complex, with the need of an experienced pediatric radiologist to interpret the findings.^{76,77} Sanders et al. developed a

Curve Type				
Type	Proximal Thoracic	Main Thoracic	Thoracolumbar / Lumbar	Curve Type
1	Non-Structural	Structural (Major*)	Non-Structural	Main Thoracic (MT)
2	Structural	Structural (Major*)	Non-Structural	Double Thoracic (DT)
3	Non-Structural	Structural (Major*)	Structural	Double Major (DM)
4	Structural	Structural (Major*)	Structural	Triple Major (TM)
5	Non-Structural	Non-Structural	Structural (Major*)	Thoracolumbar / Lumbar (TL/L)
6	Non-Structural	Structural	Structural (Major*)	Thoracolumbar / Lumbar - Main Thoracic (TL/L - MT)

STRUCTURAL CRITERIA
(Minor Curves)

Proximal Thoracic: - Side Bending Cobb $\geq 25^\circ$
- T2 - T5 Kyphosis $\geq +20^\circ$

Main Thoracic: - Side Bending Cobb $\geq 25^\circ$
- T10 - L2 Kyphosis $\geq +20^\circ$

Thoracolumbar / Lumbar: - Side Bending Cobb $\geq 25^\circ$
- T10 - L2 Kyphosis $\geq +20^\circ$

*Major = Largest Cobb Measurement, always structural
Minor = all other curves with structural criteria applied

LOCATION OF APEX
(SRS definition)

CURVE	APEX
THORACIC	T2 - T11-12 DISC
THORACOLUMBAR	T12 - L1
LUMBAR	L1-2 DISC - L4

Modifiers		
Lumbar Spine Modifier	CSVL to Lumbar Apex	
A	CSVL Between Pedicles	A
B	CSVL Touches Apical Body(ies)	B
C	CSVL Completely Medial	C

Thoracic Sagittal Profile T5 - T12	
-	(Hypo) < 10°
N	(Normal) 10°- 40°
+	(Hyper) > 40°

Curve Type (1-6) + Lumbar Spine Modifier (A, B, or C) + Thoracic Sagittal Modifier (-, N, or +)
Classification (e.g. 1B+): _____

Figure 6 The Lenke classification. (The classification that has been adopted by spine surgeons for determining the amount of arthrodesis needed for adequate curve correction, stability, and balance.) Used with permission from Lenke LG, Betz RR, Harm J, et al. Adolescent Idiopathic Scoliosis: A new classification to determine extent of spinal arthrodesis. *J Bone Joint Surg Am.* 2001;83-A(8):1169-1181

simplified version of using the Tanner-Whitehouse method in 2008 that has been found to be both fast and reliable.⁷⁶ The search for a better radiographic marker of skeletal maturity continues, with research into using a variation of the radiographic appearances of the olecranon first described by Sauvegrain, as well as other methods⁴¹ (Figure 10).

Treatment options

Guidelines for treating AIS are based primarily on the initial curve severity and the amount of remaining skeletal growth. It is important to note that scoliosis, unlike spinal asymmetry, does not improve with time. Curves greater than 20° in those who are skeletally immature should be observed with radiographs on a routine basis, typically every 6 months until skeletal maturity. Alternatively, if no progression occurs and if other testing confirms a low likelihood of progression (DNA testing) or the patient is approaching skeletal maturity, x-rays can be discontinued.^{28,29} Although there still remains controversy, most experts agree that patients with curves more than 25° in the skeletally immature patient should be offered some form of an orthosis and brace treatment.^{5,14,78-80}

Orthotics

Scoliosis treatment has been performed for more than 2 millennia, with Hippocrates describing a distraction apparatus for deformity correction in the fifth century BC.^{10,81} Brace treatment and design has exploded during the last 30 years, with more orthotic devices on the market than ever before.^{80,81} The theory of brace treatment is to over-correct the coronal plane deformity in an attempt to stop further progression. This corresponds to a direct type of Osteopathic Balanced Ligamentous tension technique and is obtained with various brace devices depending on the location of the curve apex. The time required for theoretical correction varies depending on each device (Figure 11).

Some argue, however, that a 2-dimensional orthosis is not effective for correcting a 3-dimensional spine deformity.²⁶ The decision to brace can be quite troubling for both families and physicians. This is due to the psychological effect of wearing a brace, as well as the conflicting evidence of brace efficacy and the possible need for later surgery despite brace compliance.^{79,82-88}

Brace-effectiveness studies are quite hard to control in terms of randomization, brace wear time (night time vs 23-hour bracing), and patient compliance.⁸⁹ Because of a universal lack of consistency, a new criteria for AIS bracing was

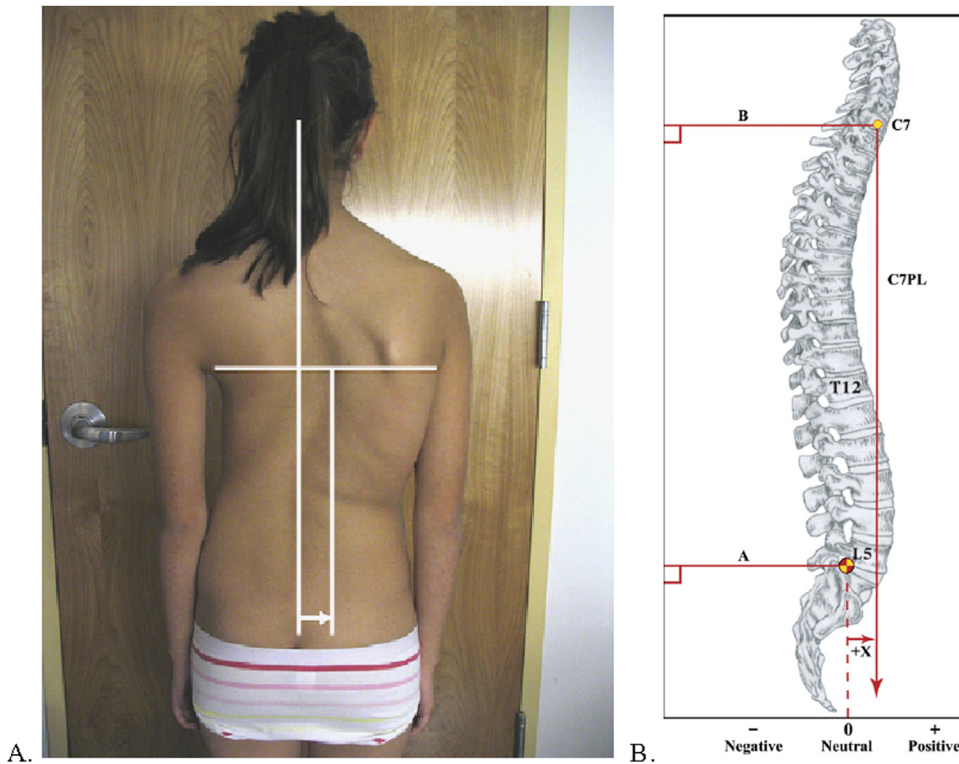


Figure 7 Spinal balance (A) Coronal balance is maintained in this patient because of a compensatory thoracolumbar curve, allowing the patient’s occiput to rest over the sacrum and in line with the natal cleft. (B) Sagittal balance refers to the position of the C7 vertebrae in relation to the sacrum, and can be negative, neutral, or positive.

introduced in 2005 in a hope to help standardize future studies.⁹⁰ A multicenter partially randomized prospective study that is finishing data collection in August 2012 hopes to finally determine if bracing is effective in AIS.⁹¹ The *Bracing for Adolescent Idiopathic Scoliosis Trial*, or BrAIST, will attempt to clarify the efficacy of orthotic treatment for AIS.⁹¹

Manipulation and modalities

The concept of manipulative medicine and other physical therapy or modalities to correct scoliosis has been promoted by some groups and nonoperative practitioners, especially in the European literature.⁹²⁻⁹⁶ Similar to bracing treatment,

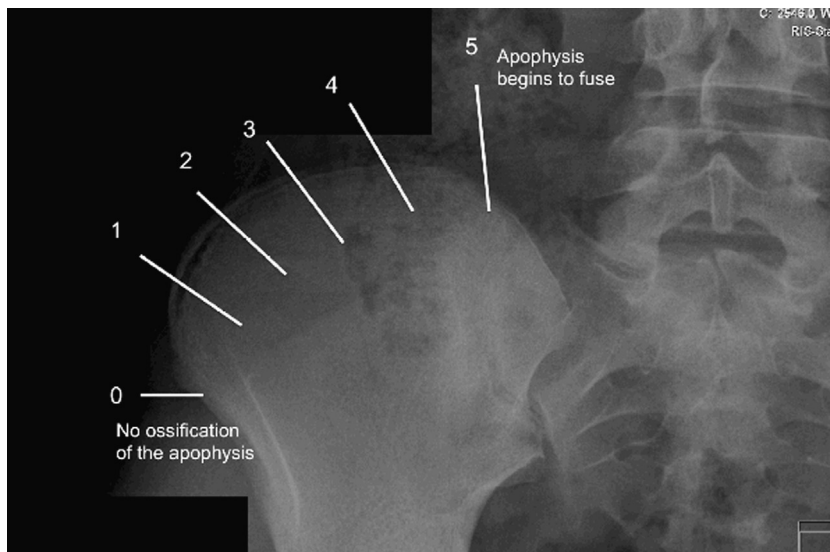


Figure 8 The Risser grading system. (The appearance of the iliac apophysis is referred to by a grade of 1 and signifies skeletal immaturity; fusion of the apophysis is a grade of 5 and signifies skeletal maturity.)



Figure 9 The triradiate cartilage (A) An immature TRC. (B) A mature and fused TRC.

there are limited high-quality studies to substantiate therapy or modality use to prevent further progression of scoliosis. Several studies have found little benefit with these treatments; however the use of such therapies may be of benefit for overall health, fitness, core strengthening, or symptom relief.^{19,78,97-99}

Surgery indications

It is seen that the early onset subgroup of IS has the greatest risk of progression and likely need for surgical intervention. This is done to prevent associated cardiopulmonary complications that occur with curves greater than 70°. In AIS patients, curves that are >50° are

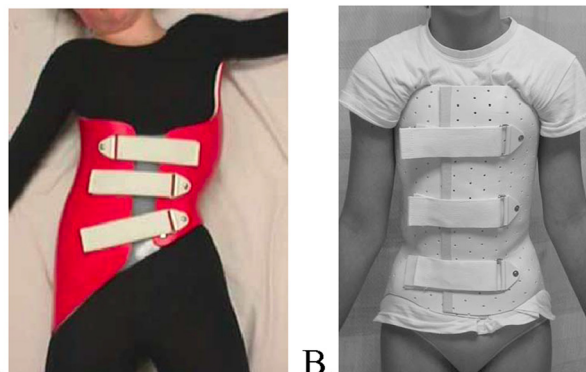


Figure 11 Spinal orthoses (A) The providence night time brace; by overcorrecting the curve it is believed that orthotic wear can be limited to the night. (B) The Wilmington Brace.



Figure 10 Tanner-Whitehouse-III stage. (Examples of digital skeletal maturation is seen here in the middle phalanx. Please refer to the original article for a complete description.)

usually recommended for surgery because of the likely progression throughout adulthood.¹⁴ Surgical indications cannot be taken lightly, as there can be approximately a 15% nonneurologic complication rate (bleeding, wound infection, and ileus) as well as the potential for new neurologic deficits after surgery¹⁰¹⁻¹⁰³ (Figure 12).

Curve Magnitude	Skeletal Maturity	Treatment
0-24°	Immature	Observation
25-40°	Immature	Bracing
>45°	Immature	Surgery
>50°	Mature	Surgery

Figure 12 Treatment guidelines.

Conclusions

AIS is relatively common in the pediatric population. The guiding factors for the diagnosis, initial nonsurgical treatment, and possible surgical treatment of AIS are based on many factors, primarily those concerning skeletal maturity and the potential for the risk of rapid and future progression. Although there is limited level I evidence for bracing or other nonsurgical treatment options, current and future studies hope to elicit treatment algorithms to separate those patients at highest risk of progression.

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