Evaluation and management of adolescent idiopathic scoliosis: a review

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Adolescent idiopathic scoliosis (AIS) is a 3D spinal deformity affecting children between the ages of 11 and 18, without an identifiable etiology. The authors here reviewed the available literature to provide spine surgeons with a summary and update on current management options.

Smaller thoracic and thoracolumbar curves can be managed conservatively with observation or bracing, but corrective surgery may be indicated for rapidly growing or larger curves. The authors summarize the atypical features to look for in patients who may warrant further investigation with MRI during diagnosis and review the fundamental principles of the surgical management of AIS.

Patients with AIS can be managed very well with a combination of conservative and surgical options. Outcomes for these children are excellent with sustained longer-term results.

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KEY WORDS adolescent idiopathic scoliosis; pediatric; spine; deformity

ADOLESCENT idiopathic scoliosis (AIS) is one of the most commonly treated types of scoliosis. We here summarize the classification, evaluation, and management of AIS and briefly discuss more subtle aspects of clinical care.

Etiology and Pathogenesis

Adolescent idiopathic scoliosis is a 3D spinal deformity that can involve one or more segments of the thoracolumbar vertebral column,^{19,61} affects children between the ages of 11 and 18,²⁷ and is of unknown etiology. In the simplest terms, scoliosis is described as lateral curvature of the spine > 10° with a rotational component of the vertebrae. Scoliosis is assigned to different subgroups based on patient age at onset, disease severity and etiology, and type of curve.²⁷ It is further subcategorized as idiopathic versus nonidiopathic. Idiopathic scoliosis is subcategorized as follows: infantile scoliosis (0–3 years of age),³³ juvenile

scoliosis (4–10 years of age),⁷ adolescent scoliosis (11–18 years of age),²⁷ and adult scoliosis (age > 18 years).²⁷

The exact prevalence of AIS is difficult to estimate based on the current literature. Varying definitions of scoliosis, the different age groups, and the inclusion of curves < 10° in studies all contribute to this issue. However, in looking at a meta-analysis of current data, one can estimate an annual prevalence of AIS to be 0.47%–5.2% with a female/male ratio of 1.5:1 to 3:1, increasing with age.^{25,27,42,54,64} In 2009 the total cost of treating AIS was estimated as \$514 million (\$137 million from Medicaid), ranking second only to appendicitis in children 10–17 years old.^{22,60}

The current literature cites several possibilities for causative mechanisms of AIS: genetics, biomechanical growth modulation, dorsal shear forces and axial rotational instability, uncoupled spinal neuro-osseous growth, postural abnormalities and hindbrain dysfunction, motor control problems, systemic melatonin-signaling pathway deficiency

ABBREVIATIONS AIS = adolescent idiopathic scoliosis; AP = anteroposterior; AVR = apical vertebral rotation; AVT = apical vertebral translation; CSVL = center sacral vertical line; PA = posteroanterior; TLSO = thoracolumbosacral orthosis; VATS = video-assisted thoracoscopic surgery.

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cies, and systemic platelet calmodulin dysfunction.^{1,4–6,10,14,20,21,24,44,53,58,65}

Various genetic factors have been implicated in the pathogenesis of AIS, but the inheritance of the disorder is unclear.¹¹ The literature is inconclusive as to whether the disorder is X-linked,²⁴ multifactorial,⁴⁴ autosomal dominant,⁶⁵ or a dominant major gene diallele model.¹

Clinical Presentation

The most common presentation of AIS is a disfiguring shape of the back and waistline in a teenage female. Common complaints include experiences with ill-fitting shirts, leaning to one side, or arm friction with the ipsilateral pelvis. A thorough neurological examination is necessary. Particular attention should be paid to reflexes, motor strength, and skin lesions. Clinical leg-length discrepancy should be ruled out as well. Evaluation of the back should focus on shoulder tilt, waistline asymmetry, and palpable masses or skin lesions. The Adams forward bend test is performed to identify rib prominence and is very useful in identifying axial and coronal plane curvatures with corresponding axial rotation. A scoliometer can also be used during this test to clinically quantify the rotation.

Patients with AIS do not usually present with pain or neurological deficits. Radicular symptoms, weakness, bowel or bladder incontinence, and sensory anomalies are exceptionally rare; when they are present, alternative diagnoses should be considered. However, a study by Ramirez et al.⁴¹ looked at 2442 patients with AIS and noted that 23% (560 patients) had back pain at the time of presentation and 9% had back pain during the period of observation. Furthermore, these authors found an association between back pain, an age > 15 years, skeletal maturity (Risser grade ≥ 2), postmenarchal stage, and a history of injury.⁴¹ Among the 560 patients with back pain in their study, 9% had an underlying pathological condition.

Pain, atypical curve patterns, or kyphosis in a patient is predictive of a potential underlying lesion.^{2,3,41} Other authors have suggested that the most common cause of pain in patients with AIS is osteoid osteoma and have recommended bone scans following the standard roentgenograms that are ordered to evaluate scoliosis.^{41,56} With regard to osteoid osteomas, patients will also report decreasing pain with the use of nonsteroidal antiinflammatory agents. If abnormalities are detected, further workup is necessary to elucidate the nature of the symptoms.

Radiographic Analysis

Historically, standing posteroanterior (PA), lateral, and side-bending 14 × 36-inch cassette radiographs have been the standard for scoliosis evaluation. Standing radiographs are optimal in ambulatory patients because spinal balance and curve magnitude change when a patient is supine. Standing radiographs should include the entire spine as well as the pelvis with clear views of the iliac crests. These views permit determination of the patient's skeletal age as well as the global balance of the spine. Advanced imaging such as CT or MRI is reserved for cases in which there are “red flags” (for example, left thoracic curve, kyphosis, pain, or neurological deficits). Lateral radiographs should

be reviewed with special attention to the degree of thoracic kyphosis (or in most cases hypokyphosis), lumbar lordosis, and sagittal balance. Cervicopelvic balance and sacropelvic parameters should also be evaluated. The PA films demonstrate the location and extent of scoliosis. The Cobb method is used to measure the degree of this lateral deformity. Computed tomography is not routinely used in the evaluation of AIS in order to limit radiation exposure in this young patient population. Magnetic resonance imaging should be performed in all patients younger than 11 years old with scoliosis > 20° or in patients with back pain, hyperkyphosis, unusual curves, or an abnormal neurological examination.^{23,40,51}

Assessment of Curves and Classifications

The most common method of measuring curve size is utilizing the Cobb angle on standing 14 × 36-inch cassette radiographs in the PA, lateral, and side-bending views.⁶³ To calculate the Cobb angle from PA radiographs, a line is drawn along the superior endplate of the upper-end vertebra (vertebra maximally tilted above the apex of the scoliotic curve) and the inferior endplate of the lower-end vertebra (vertebra maximally tilted below the apex of the scoliotic curve).⁶³

The pattern of a scoliotic curve is described by the apical vertebra (vertebra at the apex of the curve). It is important to assess the overall or global spinal balance in a patient with scoliosis. For example, if a patient has an isolated thoracic structural curve, he or she may have nonstructural compensatory curves (nonrotated curves) above and below the structural curves to bring the head into alignment over the pelvis. Hence, spinal balance is achieved when the sum of the angles of the compensatory curves equals that of the structural curves. Another important consideration is the pubertal stage of the patient. Puberty lasts 2 years and begins at a bone age of 11 in girls and 13 in boys. This stage is characterized by an “acceleration phase” of rapid growth lasting 2 years and is followed by a steady reduction of growth for 3 years, known as the “deceleration phase.” The skeletal maturation of the patient must be followed to evaluate the risk of AIS progression during the acceleration phase; the younger the child, the higher the risk for progression of a scoliotic curve.¹² Bone age, Tanner classification, stages of puberty, standing and sitting height, arm span, and weight should all be considered when evaluating patients and stages of puberty.^{49,50,52} An early indicator of puberty is ossification of the sesamoid bone of the thumb on anteroposterior (AP) radiographs.¹² As a general rule, during the acceleration phase of puberty, any spinal curve that increases by 1° per month is likely to be progressive, while one increasing by 0.5° can be monitored closely. Any curve increasing by < 0.5° per month is considered mild.¹³

A saliva-based genetic test (ScoliScore) analyzes 53 single nucleotide polymorphisms to predict the progression of AIS curves and showed promising initial results.⁵⁹ However, independent testing showed less reliable results with a positive predictive value of 0.27 and a negative predictive value of 0.87.⁴⁷ Ongoing research continues to attempt to better define predictive parameters associated

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)	$< 10^\circ$
N (Normal)	$10^\circ - 40^\circ$
+ (Hyper)	$> 40^\circ$

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

FIG. 1. Lenke classification system. CSVL = center sacral vertical line; SRS = Scoliosis Research Society. Reprinted from Lenke LG, Betz RR, Harms J, Bridwell KH, Clements DH, Lowe TG, et al: Adolescent idiopathic scoliosis: a new classification to determine extent of spinal arthrodesis. *J Bone Joint Surg Am* 83-A (8):1169-1181, 2001. <http://journals.lww.com/jbjsjournal/pages/default.aspx>. With permission from Wolters Kluwer Health Inc.

with curve progression, but currently no genetic testing is routinely used to predict curve progression.

Surgeons use the Lenke classification system³¹ to classify the types of curves noted in AIS and to determine the extent of arthrodesis necessary to correct scoliosis in this patient population. This system divides curves into 6 types with 3 types of lumbar modifiers (Fig. 1). The curves are described as structural or nonstructural and as main thoracic, double thoracic, double major, triple major, thoracolumbar/lumbar, or thoracolumbar/lumbar-main thoracic. Furthermore, the lumbar spine modifier is applied when a center sacral vertical line (CSVL) is between the pedicles in the lumbar spine curve (A), touches the apical body of the lumbar curve (B), or is medial to the lumbar spine curve (C).³¹

Nonsurgical Treatment

The primary nonsurgical treatment for AIS is bracing, whose goal is to obviate the need for surgery by limit-

ing curve progression. Many scoliotic curves in AIS that are $< 20^\circ$ can be observed and followed with serial radiographs or clinical examinations at 6-month intervals until skeletal maturity. If the curve is between 25° and 40° in a skeletally immature patient with a Risser Grade 0-1, most would agree that bracing is indicated.^{32,36,43,46,62} While the goal of bracing is to deter further progression of the curve, bracing will not result in curve regression. Two types of braces are used for treating AIS: a thoracolumbosacral orthosis (TLSO) and a cervicothoracolumbosacral orthosis (CTLTO). The former brace type includes the Wilmington, Boston, Lyon, Cheneau, Rigo-Cheneau, Malaga, and SPoRT orthoses. The latter type brace, such as the Milwaukee brace, is typically used for thoracic scoliosis with an apex above T-8, and a TLSO is employed for thoracic scoliosis with an apex at or below T-8.^{17,53} Though opinions in the literature vary, most authors recommend that braces be worn at least 16-20 hours per day with bracing treatment protocols lasting anywhere from 2 to 4 years or until skeletal maturity.^{26,28,45,55} Standing radiographs are

usually taken at 6-month intervals to assess bracing efficacy or curve progression.

The literature varies on the efficacy of bracing in AIS. Some authors report that Boston bracing is effective in girls with 25°–35° curves.³⁴ One Swedish study with a 16-year follow-up concluded that braced patients had no curve progression and that unbraced patients had 6° of curve progression.⁹ Conversely, a large meta-analysis by Dolan and Weinstein¹⁵ revealed no significant difference in surgical rates between braced and unbraced patients.

Ideal candidates for bracing are young, have scoliotic curves between 25° and 40°, are in acceleration growth phases, with a Risser Grade 0–1 and the goal of delaying surgery to maintain spinal and chest growth.⁵³

The most recent and definitive bracing trial was the Bracing in Adolescent Idiopathic Scoliosis Trial (BrAIST), the results of which were published in 2013.⁶⁰ The BrAIST was a multicenter prospective controlled trial comparing bracing to observation in patients with AIS. It included a randomized cohort and preference cohort. The population comprised patients 10–15 years old with Risser Grades 0, 1, or 2, a Cobb angle of 20°–40° for the largest curve, and no prior treatment for their scoliosis. The bracing group received a rigid TLSO, which was prescribed to be worn 18 hours/day. The trial was stopped early given the efficacy of bracing, which significantly decreased the progression of high-risk curves and was associated with a greater likelihood of reaching skeletal maturity with a major curve < 50°, as compared with observation alone. Furthermore, the likelihood of a successful outcome correlated with the average hours of daily brace wear.⁶⁰ There were important limitations to the study, however. It was not a true randomized controlled design. Treatment preferences limited enrollment in the randomized trial; as a result, the authors added observational patient preference groups, with 116 patients receiving randomly assigned care and 126 receiving patient-directed care, meaning that 71% of this group chose the bracing treatment. Additionally, not all of the patients wore braces for 18 hours/day, and 27% stopped using the brace altogether. Furthermore, the association between the duration of brace wear and better outcomes may have been skewed because patients with curves that were likely to progress may have been less inclined to wear the brace. In particular, these curves would be relatively stiff and would resist the corrective measures of bracing. Moreover, the end point of the trial was not curve progression but only curve magnitude and the need for surgery.

There is a need for further research on bracing, and newer studies are analyzing prognostic factors for bracing efficacy. Ogilvie et al. have looked at various genetic markers in AIS as a predictive measure of bracing efficacy.³⁹ Using a 30-marker genetic panel, the authors predicted which patients had curves that were likely to be brace resistant.^{39,53} Despite initial promising data, recent reviews have demonstrated that methods for predicting curve progression and bracing efficacy are not reliable and cannot be recommended as diagnostic criteria.³⁸

Surgical Treatment

The relative indications for surgery in patients with

AIS are curves > 45°–50° or rapidly progressing curves. The goals of surgery are to correct the deformity and stabilize the spinal curve, typically with instrumentation, while accounting for overall spinal balance. Posterior pedicle screw and rod fixation techniques represent the mainstay of approaches, with anterior approaches reserved for some thoracolumbar curves or anterior releases reserved for severe deformities. Anterior spinal fusion is typically indicated for skeletally immature patients to arrest vertebral growth and prevent a crankshaft deformity, reduce the number of vertebral bodies included in the fusion construct, and increase flexibility for the correction of rigid curves.⁵³

For either the anterior or posterior approach, all levels to be included in the fusion are typically exposed. For anterior approaches, discectomies are performed at the intended fusion levels. For posterior approaches, the ligamentum flavum can be resected to allow for greater correction of curves, with or without discectomies, depending on the extent and type of correction required.

For anterior approaches, a thoracic surgeon provides exposure to the anterior chest. From this approach, cortical screws and interbody constructs can be placed. Care must be taken to measure the length of the vertebral cortex in order not to penetrate the contralateral side with the screw. Furthermore, the interbody grafts or cages should be typically placed on the concave side of the scoliosis to facilitate curve reduction.³⁷

There is considerable debate in the literature regarding single versus dual rod placement, with dual rod constructs resulting in increased stiffness in torsion and flexion-extension loading.¹⁸ In a recent retrospective study, Nambiar et al.³⁵ analyzed patients undergoing an anterior approach for the correction of AIS. In that multicenter study, patients received either single or dual rod instrumentation via an anterior approach. The authors found no significant differences in postoperative radiographic measurements and functional outcomes between the groups.

With recent advances, video-assisted thoracoscopic surgery (VATS) has been employed for anterior approaches. Reviews of the procedure advocate its use based on superior cosmesis as compared to that obtained with standard approaches and less surgical trauma. However, in addition to increasing operative time, adequate anterior release and curve correction cannot be achieved through the limited VATS corridor.^{29,64}

Growth modulation via anterior surgery using internal implants has also been investigated as an alternative surgical option. Vertebral body stapling was developed using the Hueter-Volkman principle to anchor the convexity of a curve and allow for correction by differential growth along the concavity. However, the indications for vertebral body stapling were quite narrow, encompassing curves ideally between 25° and 34° in magnitude.⁸ Ultimately, research based on the staple led to further advances, and current investigations into a vertebral body tether are ongoing.⁴⁸

Currently, the most commonly adopted surgical approach is posterior spinal fusion with or without multiple Ponte osteotomies with derotation techniques to provide correction. Pedicle screw instrumentation is used to sta-

FIG. 2. Case 1. **A:** AP and lateral radiographs obtained in a 13-year-old girl who was 13 months postmenarche with progressive AIS despite bracing, demonstrating a right thoracic curve of 55° and left lumbar curve of 48°. **B:** AP and lateral radiographs demonstrating posterior spinal fusion from T-4 to L-3 with excellent correction of her deformity. **C:** AP and lateral radiographs obtained at her 6-month follow-up, demonstrating maintenance of correction and intact instrumentation. At that time, the patient had returned to full activities with no complaints.

bilize the correction, and arthrodesis is augmented with biologics per surgeon preference.

Surgical Intervention and the Lenke Classification System

Lenke et al.³¹ have defined the concept of selective fusion, which refers to the fusion of only the structurally major curve. The largest curve is considered structural along with any other curves that do not bend below 25° or have > 20° of kyphosis. The selective fusion addresses the structurally major curve while leaving the spinal segments along the minor curves unfused. Proximal thoracic curves range from T-2 to T-5, main thoracic curves from T-5 to T-9, and thoracolumbar curves from T-10 to L-2.

Furthermore, according to the Lenke classification system, there are 2 considerations for inclusion of the lumbar spine in a thoracic fusion construct: 1) flexibility of the

lumbar curve and 2) the degree of apical vertebral translation (AVT).^{31,37} Flexibility is assessed on a best-bend (to the opposite side) radiograph; if the coronal plane measurement is < 25°, the curve is classified as compensatory and should not be included in the thoracic fusion construct. Next, with respect to the AVT, the larger the translation of the apical vertebra from the CSVL, the more likely it is that a curve is structural and requires inclusion in the fusion levels.^{31,37} Curves assigned lumbar modifier A or B should not be included in the arthrodesis unless there is a kyphosis of at least +20° in the thoracolumbar region.³⁰

Selective fusion can be performed for Lenke Type 1C, 2C, and 5C curves. In order for Type 3 scoliosis to have a selective thoracic fusion, the thoracolumbar curve should be minimally rotated, small, and flexible, as denoted on bending lumbar radiographs. For Lenke Type 3C and 6C for which both thoracic and lumbar curves are structural, Cobb angles, AVT, and apical vertebral rotation (AVR) are

FIG. 3. Case 2. **A:** AP, lateral, and bending radiographs obtained in a 17-year-old boy with progressive AIS despite bracing, demonstrating a right thoracic curve of 57° that bent down to 47° and a flexible 40° left lumbar curve that bent out to 2°. **B:** AP and lateral radiographs demonstrating selective posterior spinal fusion from T-4 to T-12 with excellent curve correction of the thoracic and lumbar curves. **C:** AP and lateral radiographs obtained at his 6-week follow-up, demonstrating maintenance of correction and intact instrumentation.

considered. Selective thoracic fusion in Type 3C scoliosis may be considered if the ratios of Cobb angle, AVT, and AVR measurements for thoracic:thoracolumbar/lumbar curves are > 1.2 . In Type 6C scoliosis, selective thoracic fusion can be employed when the ratios of Cobb angle, AVT, and AVR measurements for thoracic:thoracolumbar/lumbar curves are > 1.25 . Ratios that approach 1 inher-

ently mean that the curves are similar in magnitude and structural in nature, and selective thoracic fusion will fail.³⁰

Postoperative Management

After scoliosis surgery, patients are transferred to a

surgical intensive care unit or surgical step-down unit. Pain management consists of patient-controlled anesthesia and long-acting oral pain medication. Surgical drains are placed and remain until the output is < 20 ml/day. Patients are expected to ambulate on postoperative Day 1 or 2. Most patients will not require postoperative bracing.^{16,57}

For revision surgery or for patients with prior pseudarthrosis or poor bone quality, postoperative bracing can be used.¹⁶ In this demographic, patients wear a brace for 6–8 weeks.¹⁶ At the senior author's institution, patients are able to return to school or work in 2–4 weeks and can resume preoperative activities, with a return to contact sports in 4–6 months.⁵⁷

Illustrative Cases

Case 1

Figure 2A features AP and lateral radiographs obtained in a 13-year-old girl who was 13 months postmenarche with progressive AIS despite bracing. She had a right thoracic curve of 55° and left lumbar curve of 48°. Bending radiographs had been obtained but were not available. The patient had a main thoracic structural curve and a rigid, structural lumbar curve, which is a Lenke C lumbar spine modifier. Thus, the patient had a Lenke Type 3C curve. Based on the magnitude of the lumbar curve, the AVT, and the Cobb angle ratio for thoracic:thoracolumbar/lumbar curve < 1.25 and since the lumbar curve was not flexible, a thoracolumbar fusion was performed. Figure 2B shows the final thoracolumbar construct: T4–L3 posterior fusion with instrumentation.

Case 2

Figure 3A demonstrates AP, lateral, and bending radiographs obtained in a 17-year-old boy with progressive AIS despite bracing. The patient had a right thoracic curve of 57° that bent down to 47° and a flexible 40° left lumbar curve that bent out to 2°. The patient had a main thoracic structural curve and a nonstructural lumbar curve with a Lenke B lumbar spine modifier. Thus, the curve was a Lenke Type 1B curve, and a selective thoracic fusion was employed. The patient underwent a selective T4–12 posterior spinal fusion with instrumentation as depicted in Fig. 3B.

Conclusions

Adolescent idiopathic scoliosis is a common disease entity facing spinal surgeons who treat pediatric patients. An understanding of the basic principles of evaluation and management is important. Pain symptoms or neurological deficits should alert the surgeon to other diagnoses. Mild (< 20°) or moderate (20°–40°) curves can be observed or treated with bracing, respectively. More extensive curves need surgical correction and stabilization. The predominant surgical approach is a posterior segmental pedicle screw and rod fixation construct.

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Disclosures

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Conception and design: Jada, Baaj. Acquisition of data: Jada. Analysis and interpretation of data: Jada. Drafting the article: Jada. Critically revising the article: all authors. Reviewed submitted version of manuscript: all authors. Study supervision: Hwang, Samdani, Baaj.

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