



ADOLESCENT IDIOPATHIC SCOLIOSIS: GENERAL REVIEW

Radwan Nouby Mahmoud^a, Yasser Mahmud El-Banna^b, Ashraf A. Abdellatif^c Eslam Elsayed Elkhatib^{c*}

^aNeurosurgery Department, Faculty of Medicine, Assiut University.

^bNeurosurgery Department, Faculty of Medicine, Alexandria University.

^cNeurosurgery Department, Faculty of Medicine, South Valley University.

Abstract

Adolescent Idiopathic Scoliosis (AIS) is an abnormal curvature of the spine exceeding 10 degrees, diagnosed in adolescence and in which the etiology is unknown. The gold standard for describing scoliosis is the Cobb angle. 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. Patients can be managed very well with a combination of conservative and surgical options. Outcomes for these children are excellent with sustained longer-term results.

Keywords: adolescent, scoliosis.

Introduction:

Adolescent Idiopathic Scoliosis (AIS) is an abnormal curvature of the spine exceeding 10 degrees, diagnosed in adolescence and in which the etiology is unknown [1].

Scoliosis is a deformity affecting the spine in all three planes. In the frontal plane, there are lateral curves with vertebrae deviating from the midline. A relative anterior overgrowth of the vertebral bodies changes the sagittal alignment of the spine, resulting in a hypokyphosis and/or hyper-lordosis. Each vertebra within the deformity is not just laterally deviated and tilted, it is also rotated in the axial plane. This means that the scoliotic spine is deformed in all three planes [2].

Even though the spine is deformed in all three dimensions, the evaluation of scoliosis has, historically, been based on two-dimensional (2D) frontal and lateral radiographic examinations. The gold standard for describing scoliosis is the Cobb angle [3]. Right lateral deviation and axial vertebral rotation (AVR) are to some degree physiologic in the mid and lower part of the thoracic spine in adolescence and beyond. It has been hypothesized that this is due to the location of the heart in the chest [4].

According to curve magnitude and maturity of the patients, the main treatment options for scoliosis include observation, bracing, and operation. For curvature less than 25 degrees, patients can be observed on a 6-to-12-monthly basis with clinical and radiological follow-up [5-6].

For unmatured patients with curves between 25 and 45, bracing should be considered. Correction surgery is indicated to patients with curves beyond 45°. However, curves in some



patients continued to progress even after skeletal maturity, especially for those who had curves larger than 50° [7].

There are two main approaches to carry out the correction surgery, posterior and anterior, while posterior approach becomes the trend due to the safety and correction outcome. Although anterior surgery can save surgical segments, there are many vessels and organs anteriorly, which affects the safety of the surgery. In addition, back pain was more often seen in adult patients than in adolescent patients, which could be a reason for pursuing corrective surgery [8].

Epidemiology:

The prevalence of AIS with a Cobb angle $>10^\circ$ is approximately 3%, but only 10% of adolescents with AIS require treatment (0.3% of the population). Males and females are affected equally. However, the risk of curve progression (and therefore the need for treatment) is 10 times higher in females than in males [9].

The severity of AIS on initial presentation appears to be increased in patients who are overweight or obese. In a retrospective cohort of 150 patients (≥ 10 years of age) referred for spinal asymmetry, body mass index ≥ 85 th percentile was associated with greater average curve (approximately 24 versus 18°) at presentation, increased risk of having a curve $\geq 40^\circ$ at presentation, and more advanced skeletal maturity at presentation. Increased severity of AIS patients who are overweight/obese at presentation may be related to delayed detection for a variety of reasons, primarily the limited utility of scoliometer readings in such patients [10].

The thoracolumbar region is an area of transition with specific mechanical conditions attributing to its vulnerability under traumatic loading. The thoracic spine is stiff owing to its attachment to the ribs and the intervertebral disc being less deformable. While the lumbar spine is more dynamic as it is not attached to the rib cage and it is responsible for spine flexion and extension. The thoracolumbar junction is a connection between these two regions creating an area of high stress concentration rendering it weak against high speed dynamic loading [11].

Classification:

The King-Moe classification was the first to divide the different features of the curves in order to define the area of fusion. It derives from the experience of the surgical treatment of scoliosis with Harrington's instruments, but some controversies about sufficient interobserver and interobserver reliability [12].

As segmental instrumentation systems began to gain favor over the Harrington rods this classification system failed to give accurate and reliable guidelines for choosing the proper levels for fusion. This led to the development of a new classification system which was presented by Lawrence Lenke [13].

According to Lenke classification A few new definitions were made: **(1)** Major curve: the curve of greatest magnitude and is always structural. **(2)** Minor curve: a smaller curve which may be structural or nonstructural. **(3)** Nonstructural curve: a curve which bends to less than 25° on side bending radiographs [14-15].

Type	Proximal Thoracic	Main Thoracic	Thoracolumbar/Lumbar	Curve Type
1	Non-structural	Structural (majora)	Non-structural	Main thoracic (MT)
2	Structural	Structural (majora)	Non-structural	Double thoracic (DT)
3	Non-structural	Structural (majora)	Structural	Double major (DM)
4	Structural	Structural (majora)	Structural (majora)	Triple major (TM) ^b
5	Non-structural	Non-structural	Structural (majora)	Thoracolumbar/lumbar (TL/L)
6	Non-structural	Structural	Structural (majora)	Thoracolumbar/lumbar-main thoracic (TL/L – MT)
Minor curve structural criteria	Side bending cobb $\geq 25^\circ$ T2-T5 kyphosis $\geq +20^\circ$	Side bending cobb $\geq 25^\circ$ T10-L2 kyphosis $\geq +20^\circ$	Side bending cobb $\geq 25^\circ$ T10-L2 kyphosis $\geq +20^\circ$	

a

Table (1): Lenke classification

In this classification the lumbar spine modifier is very important because permits identifying the curves in A, B, C, based on the ratio between central sacral vertical line (CSVL) and the lumbar curve. The thoracic sagittal modifier permits according to thoracic kyphosis (T5- T12) to identify the profile as positive if $>40^\circ$ neutral in case of values included from 10° to 40° and negative if kyphosis $<10^\circ$ [16].

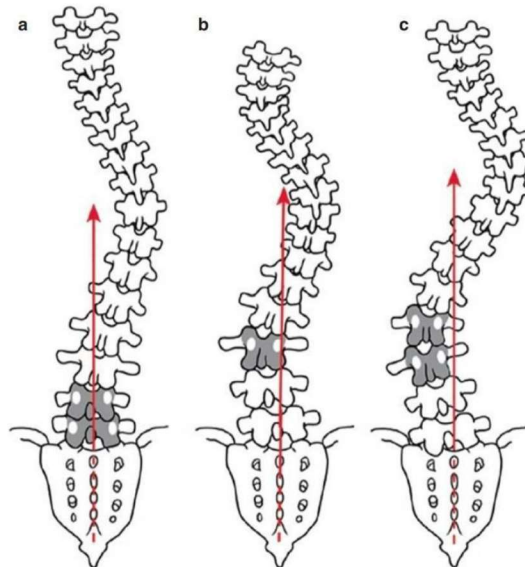


Figure (1): Lumbar Modifier. (a) CSVL between Pedicles (Apical Disc). (b) CSVL Touches Apical Pedicle (Apical Body). (c) The apicalvertebral bodies are completely lateral to the CSVL (Apical Disc)

Diagnosis:

- Clinical presentation:

AIS patients typically present with a deformity of the back, unequal shoulder levels, waistline asymmetry and a rib prominence. Occasionally back pain, not a typical finding in AIS, may be reported. Rightward thoracic curves predominate in the majority of AIS cases; thus, atypical scoliosis curve patterns, combined with rapidly progressing curves or neurological symptoms, should warrant an investigation into a possible underlying lesion [17]. The physical examination includes assessment of curve patterns, shoulder levels and waist asymmetry. Gait and posture are assessed, especially for a short-leg gait due to leg length discrepancy and listing to one side seen in severe curves. The Adams forward bending test may reveal a rib rotational deformity (rib hump) on the convex side of the curve whilst the patient is bending forward as shown in figure (3) [18].

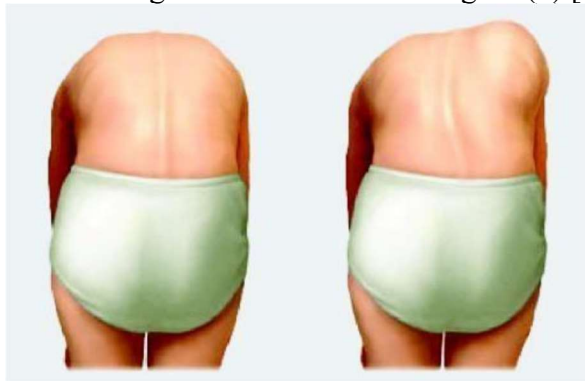


Figure (2): The Adam's forward bend test performed by (left) a patient without scoliosis, and (right) a patient with scoliosis showing a rib prominence.

A scoliometer is used to measure the angle of vertebral rotation. An angle of 7° rotatory asymmetry suggests referral for evaluation of scoliosis. As remaining spinal growth is associated with a risk of curve progression in AIS, monitoring growth velocity in every clinical examination is imperative and one of the most reliable methods for this is simple height. A scoliometer is an instrument that is placed on the back and can be used to provide an objective measure of curve rotation [19].

- Radiological evaluation:

1. X-ray:

Standard radiological images include upright standing posteroanterior (PA) and lateral views. The location of the apex vertebrae should be determined and corresponds to the curves name: cervical, thoracic, thoracolumbar or lumbar curves. The main Cobb angle is measured by identifying the largest curve and its two end vertebrae (EV), defined as the maximally tilted vertebrae cephalad and caudal to the curve's apex [20]. The Cobb method is then utilized by drawing lines along the superior border of the upper EV and the inferior border of the lower EV to form the Cobb angle. Additional imaging, such as magnetic resonance imaging, is reserved for patients with an atypical presentation of AIS suggestive of other underlying etiologies [21].

2. Computed tomography (CT):

The main roles of CT are to assess for an underlying occult pathology causing a scoliosis, and for pre-operative planning. In AIS, there are currently no clear guidelines regarding which

children with scoliosis would benefit from this more expensive study with much higher dose of ionizing radiation compared to radiographs [22].

3. Magnetic resonance imaging (MRI):

MRI study of the spine is routinely used for AIS patients. An MRI is usually ordered if your physician finds some subtle neurologic abnormalities on physical examination, or if you have significant pain or an "atypical" curve pattern. The likelihood of having something abnormal on the MRI is very small [23].

4. Dual Energy X-ray Absorptiometry (DEXA):

Low bone mineral density for age is associated with a higher risk of curve progression in AIS. The Cobb angle was found inversely related to bone mineral density in peripubertal females [24].

Management:

Current treatment options include observation, bracing, and surgery, depending on the severity of the scoliotic curve. Since some degree of scoliosis is commonly seen in the general population [25].

a) Observation:

Observation is the mainstay of therapy in patients who are still growing with a Cobb angle $<25^\circ$. Observation is often the initial approach for mild scoliosis cases. The healthcare provider will monitor the curvature through regular check-ups and X-rays. The frequency of monitoring depends on the degree of curvature and the age of the patient. This approach is used because many mild cases of AIS do not progress significantly and may not require active treatment [26].

b) Bracing:

In skeletally immature patients with AIS, bracing reduces the risk of curve progression to $\geq 50^\circ$ (the usual threshold for surgery) at skeletal maturity. The efficacy of bracing is directly related to the number of hours per day that the brace is worn [27]. Bracing is indicated for skeletally immature patients (Risser sign 0 to 2 or Sanders stage 1 to 3) whose Cobb angle is 25 to 39° at the time of presentation and skeletally immature patients whose Cobb angle is between 20 and 24° with documented progression $\geq 5^\circ$ over any six- to nine-month period during observation [28]. Bracing also is an option for skeletally immature patients with Cobb angle between 20 to 24° or 40 to 49° who choose bracing over observation or surgery, respectively [29]. The rate of brace failure appears to be increased in patients at Risser stage 0 (particularly those with open triradiate cartilage) and in patients with thoracic curves [30].

Bracing is contraindicated for skeletally mature patients or those with little remaining growth (Risser grade 3 to 5 or Sanders stage 5 to 8). It is also contraindicated for patients with Cobb angle $> 50^\circ$ or $< 20^\circ$. Thoracic lordosis is considered a relative contraindication.

Most curves can be managed with an underarm brace (athoracolumbar-sacral orthosis [TLSO], also known as the Boston brace). The TLSO is relatively easy to hide under clothing and fairly well accepted by most patients. Other types of underarm braces include the Charleston brace and the Providence brace, which are designed to be worn only at night. These nighttime bending orthoses may be effective for isolated thoracolumbar or lumbar scoliosis curves with Cobb angles less than 35° . Anon rigid brace (Spine Cor), which consists of a series of straps, is also available. Data regarding the efficacy of the different types of braces are mixed; differing inclusion criteria preclude direct comparisons [31].

Small percentage of curves require a brace with an under-chin extension (a cervico thoracolumbar-sacral orthosis [CTLSO], also known as the Milwaukee brace). The CTLSO is more difficult to hide under clothes and less well-tolerated by patients. Indications for CTLSO bracing

include thoracic curve with apex at or above T8, and double thoracic curves (high-left and lower-right thoracic curve pattern and right thoracic/left thoracolumbar pattern) [32].

Adverse effects of bracing may include psychosocial effects (e.g., diminished self-esteem, disturbed peer relationships), skin irritation, disturbed sleep, restriction of physical and recreational activities, and difficulty finding clothes that fit properly [33].

Surgery:

The primary goal of surgical treatment of AIS is prevention of curve progression through spinal fusion, but there is limited evidence regarding this outcome. Successful fusion occurs in approximately >95% of cases. Secondary goals include curve correction and improved quality of life. In a multicenter study of the outcomes of surgical treatment, patients reported improved self-image, function, and level of activity on validated measures two years after surgery for AIS [34].

Surgical correction is indicated for skeletally immature patients with curves with Cobb angle $\geq 50^\circ$ and some skeletally immature patients with Cobb angle between 40 and 50°. Surgical correction also is an option for skeletally mature patients with Cobb angle $\geq 50^\circ$. Surgery also may be warranted for patients with lumbar curves with marked trunk shift [35].

Procedures for correction of scoliosis involve spinal fusion, which may be performed posteriorly or anteriorly. Achieving bony fusion is the most important aspect of surgery, and either autograft or allograft may be used [36]. The selection of fusion levels should be individualized to the patient and is based on the curve pattern, curve size, flexibility, and sagittal alignment (presence or absence of kyphosis) [37-38].

a) Anterior spinal fusion (ASF) with instrumentation and bone grafting:

This procedure may be performed for thoracolumbar and lumbar scoliosis. The convex side of the spine is exposed anteriorly (often with the assistance of a general surgeon) by a thoracotomy and/or retroperitoneal approach. The curve is corrected by shortening the convex side of the deformity [38].

The purported advantages of the anterior approach include less blood loss, lower risk of neurologic injury (because of correction by shortening instead of distraction), and no disturbance of the paraspinal muscles. Disadvantages include increased complexity and decreased pulmonary function if the thoracic cavity is entered and/or the diaphragm is opened [39].

With improvements in PSF instrumentation and techniques, there are relatively few indications for isolated ASF and it is rarely performed. In a retrospective comparison, patients treated with posterior instrumentation and fusion for lumbar curves had better outcomes and shorter hospital stays than those treated with anterior instrumentation and fusion [40].

b) Thoracoscopic anterior spinal fusion:

Anterior instrumentation of the thoracic spine may be performed thoracoscopically. The potential advantages of video-assisted thoracoscopic ASF over PSF with thoracic pedicle screws include reduced blood loss, fewer total levels fused, and the preservation of nearly one caudal fusion level. The disadvantages include increased operative time and slightly less improvement in pulmonary function. Thoracoscopic anterior instrumentation is technically demanding and applicable to a limited number of curves. Its use seems to be decreasing [41].

c) Posterior spinal fusion (PSF) with instrumentation and bone grafting:

This is currently the most common surgical procedure for AIS. Contemporary implants for PSF are "segmental": a variety of hooks, screws, and wires are used to attach contoured rods to the spine at multiple vertebrae, or "segments" [42].

The original implants for PSF were straight stainless steel rods connected to the cephalad and caudal regions of the spine with simple hooks (Harrington rods). Segmental instrumentation

gives the surgeon greater control over the position and rotation of the spine. The increased stability of segmental instrumentation permits early mobilization with ambulation the day after surgery without external support such as a body cast or brace. Newer generation implants that use titanium with or without cobalt chrome are more compatible with magnetic resonance imaging (MRI) than stainless steel implants [43].

In patients with open triradiate cartilages, it is rarely necessary to perform an ASF, as well as a PSF, to prevent the "crankshaft phenomenon," in which the anterior spine continues to grow after the PSF, causing a severe rotational and sagittal alignment deformity. However, there is some evidence to suggest that modern segmental instrumentation systems make anterior fusion unnecessary [44].

d) Growth modulation techniques:

Growth modulation techniques include vertebral body tethering and unilateral periapical distraction. These techniques are minimally invasive, preserve motion, and do not preclude spinal fusion if they are unsuccessful [45].

Vertebral body tethering – Vertebral body tethering devices are intended to gradually correct scoliosis by slowing growth on the convex side of the curve. A single tethering device has been approved by the US Food and Drug Administration (FDA) for the correction of idiopathic scoliosis that has not responded to conservative treatment options [46]. Vertebral body tethering has been associated with progressive correction and appear to be safe, though overcorrection is a concern, and few long-term results are available. Additional adverse effects include tether breakage and pneumothorax [47].

Unilateral periapical distraction implant – A unilateral periapical distraction implant (minimally invasive deformity correction system, posterior dynamic deformity correction device) has been approved by the FDA as a humanitarian use device for the treatment of AIS. This device is implanted on the concave side of the curve it stabilizes and corrects the deformity through incremental ratchet lengthening that is activated by exercise [48].

Complications:

Complications of surgery include blood loss, infection, implant failure, neurologic injury including paralysis and blindness, progression of deformity around the fusion, and pseudoarthrosis (failure of fusion). Intraoperative blood salvage ("cell saver") and antifibrinolytics (epsilon aminocaproic acid and tranexamic acid) may be used to reduce blood transfusion requirements [49].

Postoperative care:

Monitoring neurologic function – Neurologic function must be monitored closely for 48 hours after surgery because delayed neurologic injury may occur. However, modifications in devices, techniques, and intraoperative monitoring have reduced the risk of catastrophic complications [50].

Recommendations for resuming sports participation vary by surgeon, from as little as 4 months to 12 months postoperatively. We suggest approximately six months. Once the spine has fused, all sports are permitted, with the possible exception of collision sports (eg, football, hockey, rugby). The reduced motion of the fused spine may make it more difficult to perform activities such as gymnastics and dancing at a high level [51].

MRI studies can be safely performed following spinal instrumentation for scoliosis, although some artifacts are produced. The amount of artifact depends on the composition of the implants; greater artifact has been noted with stainless steel than with titanium alloys. Routine removal of spinal implants used in correction of scoliosis is not usually performed [52].

Physical therapy is commonly recommended for AIS to help improve posture, muscle balance, and flexibility. Scoliosis-specific exercises may be prescribed, such as the Schroth Method. These exercises aim to strengthen the muscles around the spine and promote better alignment. Physical therapists may also provide education on proper body mechanics and posture to help patients manage their condition [53].

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