Adolescent Idiopathic Scoliosis

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INTRODUCTION

Traditionally idiopathic scoliosis is divided into three categories according to the age of onset: Infantile (0–3 years), juvenile (4–9 years), and adolescent (10 years to skeletal maturity). Adolescent idiopathic scoliosis (AIS) is a structural lateral curvature of the spine measuring at least 10 of Cobb’s angle and occurring at or near the onset of puberty. The prevalence of adolescent scoliosis in the adolescent population is 2 to 3%.1-4 However, as the curve magnitude increases, the prevalence decreases. The larger curves have a female preponderance in a ratio of 4:1. In the smaller curves, males and females have almost an equal prevalence.3,4 The prevalence of curves greater than 30° is about 0.2%, and it is less than 0.1% for curves greater than 40°.3,4

ETIOLOGY

As the term idiopathic indicates, the etiology of adolescent scoliosis is unknown. However, hereditary, environmental, collagen and connective tissue defects, neuromuscular and biomechanical factors have all been indicated in the etiology of adolescent scoliosis.

Hereditary Factors

It has long been known that hereditary factors play a role in the etiology of idiopathic scoliosis (IS).5 Inheritance of scoliosis in five generations was described by Garland.6 In a cohort of 1,463 individuals with IS, Grauers et al found that among those treated with a brace or surgery for scoliosis, 53% reported one or more relatives with scoliosis, compared to 46% of the untreated, pointing towards a slightly higher risk of treatment in the presence of a family history of scoliosis.7

Endocrine Factors

Thillard discovered that pinealectomized chickens developed scoliosis.8,9 This was repeated in bipedalized rats and a deficiency of melatonin was suggested to be causative of IS.10,11 Further studies, however, showed that adolescent IS patients had normal melatonin levels and pinealectomized monkeys did not develop scoliosis.12

Calmodulin is a calcium-binding receptor protein which regulates contractile properties in platelets and muscles, and also interacts with melatonin. Increased levels of calmodulin in platelets and an asymmetrical distribution of calmodulin in paraspinal muscles compared to healthy controls have been described in IS patients.13

Biomechanical Factors

Dickson et al showed that vertebral bodies were wedged in the sagittal plane in IS patients, causing an apical lordosis in thoracic curvatures. They suggested that lordosis, in a region, i.e., normally kyphotic, creates a rotational moment in the spine thus causing scoliosis.14

Neuromuscular Factors

Neuromuscular conditions producing an asymmetry of the transverse-spinalis muscle, abnormality in visual, vestibular, proprioceptive and postural control have also been implicated in AIS.15 In addition, tonsillar ectopia with abnormal somato-sensory evoked potentials, larger foramen magnum, and left-right brain asymmetries point to neural origin of AIS.16,17 These findings explain a poor performance in combined visual and proprioception, as well as spatial orientation tests and impaired postural balance in AIS patients.

The higher prevalence of AIS observed in females may be due to the fact that girls attain adolescent skeletal growth spurt in relative postural immaturity, compared to boys who go through their pubertal rapid growth at later age when their posture is more mature. Spine slenderness and ectomorphy are other risk factors in girls.18
NATURAL HISTORY OF ADOLESCENT IDIOPATHIC SCOLIOSIS

The main concerns are that patients with IS relate mainly to curve progression, occurrence of low back pain, and pulmonary function.

Curve Progression

In the immature patient, factors affecting curve progression relate mainly to curve factors and growth potential. The majority of information on curve progression relates mainly to scoliosis in girls. Six factors influencing curve progression have been identified. These include:

1. The younger the patient at the time of diagnosis, the more risk of progression.4,19
2. The greatest risk of progression happens just prior to the onset of menarche.
3. Risser grades 0 and 1 are more likely to progress than the higher Risser grades.19

Joseph C Risser first described what now is called the Risser sign in 1958. Risser observed that the state of ossification of the iliac apophysis was associated with the state of a patient’s spinal skeletal maturity.20

In the US system, the ossification of the iliac apophysis is divided into quarters, whereas in the European system the iliac apophysis ossification is divided into thirds with the fourth stage representing the commencement of fusion of the apophysis to the crest posteriorly21 (Fig. 1).

Lonstein et al collated the risk of progression based on Risser grade and curve magnitude19 (Table 1).

4. Males with comparable curves have one-tenth the risk of progression compared to females.4,22
5. Double curves have a greater chance of progression. Lumbar curves more than 30°, and where the fifth lumbar vertebra is high riding are more likely to progress.23
6. Finally, curves with higher magnitude are more likely to progress after skeletal maturity. Curves between 50 to 75° are likely to progress at 0.5 to 1.0° per year.23

Sanders et al modified the Tanner-Whitehouse staging system to assess risk of progression in adolescent scoliosis. They emphasized the use of the physes

![French risser staging system](image1)

![US risser staging system](image2)

Fig. 1: Depiction of Risser sign
Table 1: Risk of progression

<table>
<thead>
<tr>
<th>Risser</th>
<th>Curve magnitude 5–19</th>
<th>Curve magnitude 20–25</th>
</tr>
</thead>
<tbody>
<tr>
<td>0–1</td>
<td>22%</td>
<td>68%</td>
</tr>
<tr>
<td>2–4</td>
<td>1.6%</td>
<td>23%</td>
</tr>
</tbody>
</table>

Table 2: Skeletal maturity based on hand physes

<table>
<thead>
<tr>
<th>Stage</th>
<th>Key features</th>
</tr>
</thead>
<tbody>
<tr>
<td>1 Juvenile slow</td>
<td>Digital epiphyses are not covered.</td>
</tr>
<tr>
<td>2 Preadolescent slow</td>
<td>All digital epiphyses are covered.</td>
</tr>
<tr>
<td>3 Adolescent rapid—early</td>
<td>The preponderance of digits are capped. The second through fifth metacarpal epiphyses are wider than their metaphyses.</td>
</tr>
<tr>
<td>4 Adolescent rapid—late</td>
<td>Any of distal phalangeal physes are clearly beginning to close.</td>
</tr>
<tr>
<td>5 Adolescent steady—early</td>
<td>All distal phalangeal physes are closed and others are open.</td>
</tr>
<tr>
<td>6 Adolescent steady—late</td>
<td>Middle or proximal phalangeal physes are closing.</td>
</tr>
<tr>
<td>7 Early mature</td>
<td>Only distal radial physis is open. Metacarpal physisal scars may be present.</td>
</tr>
<tr>
<td>8 Mature</td>
<td>Distal radial physis is completely closed.</td>
</tr>
</tbody>
</table>

Pulmonary Function

Findings in pulmonary function studies in patients with untreated AIS demonstrate that only in patients with thoracic curves is there a direct correlation between decreased pulmonary function and increasing curve severity. Vital capacity and forced expiratory volume in 1 second decreases as the severity of thoracic curves increases. The pattern of pulmonary disease in affected patients is, uniformly, restrictive lung disease. Smokers are generally affected much more severely than nonsmokers. Results in most studies show that significant limitations of forced vital capacity in nonsmokers do not occur until the curve approaches 100 to 120°. However, the presence of thoracic hypo-kyphosis increases the loss of pulmonary function associated with curve severity.

Kim et al, in a study of pulmonary function in adolescent scoliosis relative to the surgical procedure demonstrated a significant negative correlation between the preoperative Cobb’s angle and percent predicted pulmonary function test values and a significant negative correlation between the number of involved vertebrae in the major curve and percent-predicted pulmonary function test values. The smaller number of involved vertebrae in the major curve had a strong correlation with higher pulmonary function test values preoperatively and hence, a better pulmonary function in Lenke 5 curves.

CLASSIFICATION OF ADOLESCENT SCOLIOSIS

In 1983, Howard King presented his classification system for AIS. This system was based on the experience of John Moe in the surgical treatment of AIS patients with Harrington rod instrumentation. Curves were divided into five types and guidelines and recommendations for which levels should be instrumented were given according to those different curve types to preserve motion as much as possible.

King and Moe defined five curve types:
- **Type I:** An “S” shape deformity, in which both curves are structural and cross the CSVL, with the lumbar curve being larger than the thoracic one.
- **Type II:** An “S” shape deformity, in which both curves are structural and cross the CSVL, with the thoracic curve being larger or equal to the lumbar one.
- **Type III:** Major thoracic curve in which only the thoracic curve is structural and crosses the CSVL.
- **Type IV:** Long “C” shape thoracic curve in which the fifth lumbar vertebra is centered over the sacrum and the fourth lumbar vertebra is tilted into the thoracic curve.
- **Type V:** Double thoracic curve.

of the hand to grade the skeletal maturity into eight stages (Table 2).

Back Pain

The incidence of back pain in the general population is reported at between 60 and 80%. Some studies have indicated that the prevalence of back pain in adolescents with IS are similar to the general adolescent population while other studies concluded that adolescents with IS experience more back pain and more severe back pain than their peers. Ramirez et al reported a retrospective study of 2,442 patients (6–20 years) with IS showing that the lifetime prevalence of back pain was around 23% and the point prevalence of back pain was 9%. He concluded that these results were similar to those reported by other authors in the general pediatric and adolescent population. Dickson et al also reported that back pain was significantly more prevalent among the 165-subjects with scoliosis compared to the 100 age- and sex-matched control subjects (73 vs 52%) using a questionnaire survey (average age at survey was 17 years). Sato et al, in an epidemiological study of over 43,000 adolescent school children in Japan the point and lifetime prevalence in Scoliosis group were 27.5 and 58.8% respectively, which both were significantly higher than in no scoliosis group. The pupils in scoliosis group also experienced significantly more back pain, a longer duration of back pain, and more recurrences of back pain in comparison to those without scoliosis.
The drawbacks of the King classification was the failure to address the sagittal profile of the deformity and the low inter and intra-observer reliability.\textsuperscript{37,38}

In an attempt to address these drawbacks, Lenke et al\textsuperscript{37} proposed a classification, which is now widely used. This classification uses the following terminologies:

- Major curve: The curve of greatest magnitude and is always structural.
- Minor curve: A smaller curve which may be structural or nonstructural.
  - Nonstructural curve: A curve which bends to less than 25° on side bending radiographs.

According to these definitions, there are six different curve types (Fig. 2).

**Type I**: Main thoracic (MT) is the only structural curve while the others (proximal thoracic and lumbar or thoracolumbar) are nonstructural.

**Type II**: Double thoracic in which the MT is the major curve, the proximal thoracic (PT) is the minor curve but is structural, and the thoracolumbar (TL) or lumbar curves are minor and nonstructural.

**Type III**: Double major curve pattern in which the MT is the major curve and the lumbar is the minor one but is structural, whereas the PT is nonstructural.

**Type IV**: Triple major curve pattern when the MT is the major curve but all three curves are structural.

**Type V**: The TL or lumbar curve is the major and only structural curve, with the PT and/or MT curves being minor and nonstructural.

**Type VI**: The TL or lumbar curve is the major curve measuring at least 5° more than the MT which is the minor but structural curve.

To these basic six curve types, the lumbar spine modifier is added. This modifier is defined by the location of

\begin{table}[h]
\centering
\begin{tabular}{|c|c|c|c|c|c|}
\hline
\textbf{Lumbar spine modifier} & \textbf{Type 1} & \textbf{Type 2} & \textbf{Type 3} & \textbf{Type 4} & \textbf{Type 5} & \textbf{Type 6} \\
\textbf{(main Thoracic)} & \textbf{(double Thoracic)} & \textbf{(double major)} & \textbf{(triple major)} & \textbf{(TL/L)} & \textbf{(TL/L MT)} \\
\hline
\textbf{(No to minimal curve)} & 1A* & 2A* & 3A* & 4A* & & \\
\hline
\textbf{(Moderate Curve)} & 1B* & 2B* & 3B* & 4B* & & \\
\hline
\textbf{(Large curve)} & 1C* & 2C* & 3C* & 4C* & 5C* & 6C* \\
\hline
\textbf{Possible sagittal structural criteria (to determine Specific curve type)} & Normal & PT kyphosis & TL syphosis & PT+TL kyphosis & & \\
\hline
\end{tabular}
\caption{Pictorial depiction of Lenke classification\textsuperscript{37}}
\end{table}
the CVSL on the apical vertebra of the lumbar curve. Lenke defined three lumbar modifiers (Fig. 3).

In addition, weightage is given to the sagittal profile of the curve by introducing the concept of a thoracic kyphosis modifier as follows:

- + (plus): When thoracic kyphosis measures >40°.
- N (normal): When thoracic kyphosis measures between 10 and 40°.
- − (minus): When thoracic kyphosis measures <10°.

**ASSESSMENT OF SCOLIOSIS**

A detailed history is essential with respect to age of onset of deformity, menarche, and family history as well as the presence and absence of back pain. In addition the history should cover symptoms related to other systems including the central nervous system (developmental milestones, visual/hearing problems, gait abnormalities), cardiovascular system (cardiac murmurs), and the genitourinary system.

The physical examination should include an observation of presence or absence of secondary sexual characteristics to determine the status of skeletal maturity, and therefore, the risk of progression of the deformity. Gait should be examined for any evidence of neurological deficit or limb length discrepancy. The examination of the spine should include any skin discoloration (café-au-lait spots: Fig. 4), shoulder levels, contour of the spine, scapular, rib prominence, and symmetry of the waist creases (Figs 5A and B). A plumb line observation is done to see whether the head is aligned over the sacrum (a balanced spine, Fig. 6). The Adam’s forward bend test is performed to assess for the rib prominence (Fig. 5B) and side bending in this position is used to assess for the flexibility of the curve. Alternately, the patient is placed prone and flexibility of the curve assessed manually by pushing against the rib prominence. A neurological examination is then undertaken.

Erect radiographs of the spine in PA and lateral projections are undertaken to measure the magnitude of the curve using the Cobb’s angle and determine the Risser status and the status of the triradiate cartilages on the radiographs. Side bending PA radiographs of the spine will give an indication of the flexibility of the curve and help determine the extent of fusion at surgery.

**NONOPERATIVE TREATMENT OF ADOLESCENT SCOLIOSIS**

**School Screening**

There have been several publications on screening for scoliosis since 2007, including a systematic review of
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...the literature and large retrospective studies.39-41 Labelle et al,39 published a consensus statement developed by an international task force of the SRS regarding screening for AIS. The task force performed a systematic review of the literature through 2012 and used a modified Delphi process following the framework of the World Health Organization to reach consensus on the validity of a screening program. The panel reached consensus on the five domains studied, with four of the domains: Technical efficacy and clinical, program, and treatment effectiveness, supportive of screening, but there was insufficient evidence to make a statement with respect to cost-effectiveness.

The Adam’s forward bend test with the use of a scoliometer (a specialized inclinometer) was agreed upon by the SRS task force as an effective quantitative measure, with 5 to 7° of deformity as a threshold for positive screening. The clinical effectiveness of screening for the detection of curves greater than 20° was supported in a large retrospective study by Luk et al.40 Out of 115,190 adolescents followed until the age of 19 years. In their study, 2.8% of adolescents were referred for a radiograph. At final follow-up, the positive predictive value for spinal curvature greater than 20° was 43.8 and 9.8% for treatment. Sensitivity was near 90% for both diagnosis and treatment. Conversely, Yawn et al41 reported on a population-based school screening program in Rochester, Minnesota. In this retrospective cohort study, 4.1% (92 out of 2,242) of the children screened positively and were referred for evaluation. The positive predictive value was low (0.05), and they concluded that roughly 450 children would need to be screened for every child who subsequently received treatment as a result of screening. The discrepancy in these studies points out the need for effective screening systems, as inappropriate false-positive screening may lead to unnecessary referrals and radiographs with higher population cost. Although well-done population screening may be an effective means to capture all children at risk, many communities may not have sufficient resources to carry out these programs.

**Bracing**

Treatment with rigid bracing (thoracolumbosacral and lumbosacral orthoses) is the most common nonoperative strategy to prevent curve progression.42-44 Many different designs exist, but all attempt to restore the normal contours and alignment of the spine through the use of external forces and, with some designs, the stimulation of active correction as the patient moves the spine away from pressures within the brace. Permanent correction of the curve is typically not expected. Instead the brace functions as a holding device during the high-risk growth phase. Bracing is generally indicated for curves between 20 and 40° in adolescents who still have significant skeletal growth remaining (Risser status 0-2). The recommended wear time varies across clinicians, ranging from 12 to 23 hours per day until skeletal maturity is reached (2–4 years of treatment).45

In a hallmark study, Weinstein et al46 reported on the efficacy of brace treatment bracing in adolescent idiopathic scoliosis trial (BrAIST). The study reported 75% success rate in patients randomly assigned to the bracing group as opposed to 42% in the group randomly assigned to observation alone. Success was defined as failure of progression of curve to 50° or more at skeletal maturity (Risser 4 for girls and Risser 5 for boys).

Katz et al,45 demonstrated the efficacy of bracing in a non-controlled population, where 82% of patients who wore a brace for greater than 12 hours per day had less than 5° of curve progression compared to only 31% of those who wore the brace for less than 7 hours per day. An important feature of this study was that a temperature-sensitive data recorder imbedded in the spinal orthosis monitored brace wear compliance.

Bracing, however, has many disadvantages for patients including the need for radiographs to monitor brace fit and curve response, interference with sports and other activities, limited clothing choices, and self-consciousness about the brace. Brace wear for many patients is also a reminder of their medical condition.

**Surgical Treatment of Scoliosis**

The accepted indications for surgery in adolescent scoliosis are curves greater than 50° with associated cosmetic implications. The goals of surgery include correction of deformity, prevent deterioration, and improve cosmesis.
Hibbs reported early results from 59 patients, most with paralytic scoliosis, who had undergone posterior fusion surgery. His fusion technique involved elevating the bone flaps from the laminae and turning these flaps upward or downward to bring them into contact with the adjacent decorticated laminae.

Modern instrumentation surgery for scoliosis was pioneered in the early 1950s by Harrington, who used a stainless steel rod-and-hook system to correct spinal deformities. Luque reported a segmental spinal instrumentation method that used rods and sublaminar wiring. Unlike Harrington’s system, which uses a distractive force distractive force applied to the spine through hooks, Luque’s system used transverse forces applied segmentally through sublaminar wires.

Cotrel and Doubusset described a multisegmental system, called CD instrumentation, in which multiple laminal and pedicle hooks and later, pedicle screws (PSs) were placed on the concave and convex sides of the curve. This allowed multiple fixation points using hooks, translation of the spine, and the creation of kyphosis by rod rotation. Cotrel and Doubusset believed that the CD system allowed for a shorter fusion area, derotation of the spine and the creation of kyphosis and lordosis in the thoracic spine and lumbar spine respectively.

Suk et al, reported the first in a case series using PSs for fixation of the thoracic curve, and compared the results of three different surgical constructs: Hooks only, screws only, or a combination of screws and hooks. They found that PS constructs provided better correction of coronal, sagittal, and rotational deformity with less loss of correction, a shorter fusion area, and less risk of neurological complications. In Japan in 1992, Abe et al also reported using PS constructs to correct thoracic curves in three patients, with a 78% correction rate. Several researchers have since compared surgical results between patients treated with PS-only constructs and those with hybrid constructs of hooks, sublaminar wires, and PSs. Kim et al compared the outcomes for patients treated with PS or hybrid constructs (29 each) and found that PS constructs offered better correction of the major curve and more improved pulmonary function than hybrid constructs, whereas the junctional change, lowest instrumented vertebra, time in surgery, and postoperative SRS-24 outcome scores were similar in both groups. Other authors echoed Kim’s findings in retrospective comparative studies and systemic reviews, reporting that PS constructs provided better, or at least similar, correction and maintenance of the main curve, and required fewer revisions due to their biomechanical stability.

The PS constructs have an important advantage in that vertebrae can be derotated directly with the PSs, thus reducing vertebral rotation by 42 to 60 % and reducing thoracic and lumbar hump. The PS constructs also allow for osteotomies, including Ponte and pedicle-subtraction osteotomies and posterior vertebral column resection, so that even rigid and severe curves can be corrected efficiently without anterior procedures (Figs 7A to E).

**Anterior Approach Surgery**

Dwyer developed an anterior instrumentation system using a titanium cable and screws to correct scoliosis.

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**Figs 7A to E:** (A and B) Preoperative radiographs; (C) side bending radiograph; and (D and E) postoperative radiographs after Ponte osteotomies and pedicle screw instrumentation
After thorough discectomy and a morselized rib graft, the cable was threaded through the screw heads, and a tensioning device was applied to approximate the adjacent vertebral bodies. Fusion was achieved in 91% of 51 patients treated with this device; however, there was a loss of correction in 19 patients, and others experienced loss of lumbar lordosis and instrumentation failure. Zielke et al. developed ventral derotation spondylodesis (VDS), an anterior instrumentation system that was claimed to allow derotation and restoration of lordosis of the thoracolumbar spine, and to yield better correction than either the Harrington or Dwyer systems. After following 53 patients for at least 10 years after undergoing treatment with Dwyer or Zielke instrumentation, Otani reported a 62% correction rate, a 6% rate of instrumentation failure, and patient satisfaction in most cases. However, other researchers have reported implant failure, loss of correction, progressive kyphosis, and pseudarthrosis in association with the VDS system. Kaneda et al. treated 25 patients with thoracolumbar or lumbar curves using an anterior dual-rod system, and obtained a correction rate of 83% for scoliosis and 86% for rotation with restoration of lumbar lordosis. This 2-rod system is biomechanically robust enough to prevent loss of correction after surgery.

The main advantage of an anterior approach to the thoracic spine is preservation of mobile segments through a shorter fusion: In some scoliosis’ curve patterns both proximal and distal levels can be saved, which would have been instrumented in a posterior spinal fusion. Other advantages of an anterior approach are resection of the growth plate and thereby prevention of the crankshaft phenomenon, and correction of the deformity by compression rather than distraction, which theoretically should have a lower rate of neurological complications.

FUTURE ADVANCES

Fusionless Surgery

Whereas conventional instrumented fusion for scoliosis has stood the test of time, it has the disadvantage of fusing mobile segments of the spine. Vertebral body stapling (VBS) utilizes C-shaped nitinol staples to compress across convex vertebral physes in growing children with IS with the goal of halting or reversing progressive curvature. The concept is to correct deformity without fusing the spine. The recommended indications are patients with Risser sign of 0 or 1 and coronal curve measuring between 20 and 45° deemed to be at risk of progression. Betz et al. using VBS in 41 patients followed for 2 years postoperatively reported success rate of 87% in all lumbar curves and in 79% of thoracic curves less than 35°.

GENETIC TESTING

Genetic markers associated with AIS progression have the potential to be used as a prognostic test. Linkage analyses have identified candidate regions on chromosomes. A DNA-based prognostic test was developed by Ward et al. to predict spinal curve progression in white patients with AIS. The test algorithm incorporates genotypes for 53 DNA markers and the patient’s presenting Cobb’s angle. The test reports a score between 1 and 200; this prognostic score correlates with the patient’s risk of progression.

The results have not been replicated in other studies and there may be a variation based on ethnicity.

REFERENCES


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