# Comprehensive Guide to Adult Spinal Deformity

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Edited by

Evalina Burger and Christopher Kleck

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# INTRODUCTION


## PAUL C. CELESTRE, M.D.[1], AND STEVEN D. GLASSMAN, M.D.[2]

[1] OCHSNER SPINE CLINIC, NEW ORLEANS, LOUISIANA, U.S.A

[2] NORTON LEATHERMAN SPINE CENTER, LOUISVILLE, KENTUCKY, U.S.A.


### Understanding Adult Spinal Deformity

Spinal deformities are a part of the human condition. The terms *scoliosis*, *kyphosis*, and *lordosis* were introduced by Hippocrates and further delineated by Galen more than 2,000 years ago (Vasiliadis, Grivas, and Kaspiris 2009). Although spinal deformities have been recognized since antiquity, until recently the majority of historical treatments were focused on attempts to prevent and correct spinal deformity in pediatric patients. These endeavors are the genesis of the term *orthopaedics* (Kohler 2010). In addition to untreated adolescent idiopathic scoliosis, Pott disease and de novo degenerative deformities were the most common etiologies of adult spinal deformity (ASD). Iatrogenic deformity, as a consequence of posterior distraction instrumentation, is a relatively recent phenomenon in the arc of human history (Potter, Lenke, and Kuklo 2004). Until recently, although the physiologic and psychologic effects of untreated adolescent idiopathic scoliosis in adults were well described (Ascani et al. 1986; Fowles et al. 1978), operations to treat adults with spinal deformity were generally considered too dangerous to perform (McDonnell et al. 1996), and these patients were left to pursue ineffective nonoperative treatments (Sciubba et al. 2016).

In the absence of reliable outcome measures, radiographic results were used as a proxy for surgical success; however, correlating radiographic findings of spinal deformity with patient function was challenging and unreliable (Schwab et al. 2002; D'Andrea et al. 2000). Before the development of validated patient outcomes measures, the results of treatment, both operative and nonoperative, were subjectively reported by the surgeon and commonly graded on an arbitrary scale from excellent to poor. The quality of data, and thus the ability to understand the effects of treatment, improved as researchers developed more powerful outcome measures.

Our contemporary understanding of ASD is based on patient-reported outcome measures. Generic health outcomes measures, such as the SF-12 (12-item Short Form survey), lead to spine-specific questionnaires like the Oswestry Disability Index, and finally to spinal deformity–specific tools including the Scoliosis Research Society's SRS-22r questionnaire (Fairbank and Pynsent 2000; Ware, Kosinski, and Keller 1996; Bridwell et al. 2005). These tools allowed surgeons to understand treatment effects from a patient's perspective and to reliably compare results between studies. Finally, normative population data provided surgeons with the ability to understand the consequences of scoliosis compared with an unaffected population (Diarbakerli, Grauers, and Gerdhem 2017).

In 2005, a fundamental shift in thinking about the management of ASD occurred when it was recognized that decompensation in the *sagittal* plane was the most reliable predictor of disability in adults with spinal deformity (Glassman et al. 2005). Before this, the focus on the *coronal* deformity confounded our desire to better understand the consequences of scoliosis. The paradigm shift that occurred when physicians began to focus on sagittal plane imbalance, rather than coronal curvature, continues to be the focal point of adult spinal research nearly two decades later. Critically, this fundamental change in our understanding of ASD was enabled by the aforementioned patient-related outcome measures.

As surgeons began to focus on sagittal plane deformities, the foundational role of the pelvis, particularly the role of pelvic incidence, pelvic tilt, and the relationship between pelvic incidence and lumbar lordosis became another major focus of research (Roussouly and Nnadi 2010). Like sagittal plane imbalance, abnormal pelvic tilt and mismatch between pelvic incidence and lumbar lordosis (PI-LL) closely correlated with worse patient-related outcomes, and these parameters form the basis of our contemporary classification systems for ASD (Roussouly and Nnadi 2010; Schwab et al. 2013; Schwab et al. 2012).

Building upon the refinement of data collection centered on patient-reported outcomes and a new focus on the sagittal plane deformity and the spinopelvic relationship, multicenter research consortia generated the next evolution in our understanding of ASD. Collaborative entities such as the Spinal Deformity Study Group, International Spine Study Group, and the Scoliosis Research Society allowed for the pooling of data, creating the statistical power needed to understand the results of treatment, particularly surgical treatment, for patients with spinal deformities. This breadth of experience has been further augmented by an increased interest in spinal deformity treatment within the neurosurgical community.

## Technological Advances

As our collective understanding of the consequences of ASD—and particularly sagittal plane imbalance—increased, operative techniques were developed and refined to correct these deformities. The transition to pedicle screw-based anchors afforded surgeons the ability to create constructs capable of withstanding the large biomechanical forces inherently resulting from spinal realignment operations. This transition was controversial and initially took place outside the United States (Boos and Webb 1997; Suk et al. 1995). In the United States, class action lawsuits against both surgeons and medical device companies created significant barriers to advances in the care of patients with ASD. Indeed, the International Meeting on Advanced Spine Techniques (IMAST) is a legacy of this hostile climate. Its genesis was in allowing surgeons to learn about implants and techniques not yet approved in the United States. It was not until 1998 that the US Food and Drug Administration (FDA) reclassified pedicle screws, allowing for their quotidian use.

Leveraging the more robust fixation provided by posterior pedicle screw-based constructs, ASD surgeons began to more aggressively correct spinal deformities via spinal osteotomy techniques. In particular, the pedicle subtraction osteotomy has become a powerful tool in the treatment of sagittal plane imbalance and spinopelvic mismatch (Kim et al. 2007). The enthusiastic adoption of pedicle subtraction osteotomies (Gum et al. 2016) was not without consequences, particularly the high rate of complications and nonunions associated with the procedure itself (Cho et al. 2012). Additionally, pelvic fixation techniques have provided a more robust foundation for long fusion constructions, helping to decrease lumbosacral pseudoarthrosis (Shen et al. 2013).

While achieving fusion continues to be a challenge, particularly when compared with pediatric deformity surgery, the commercialization and wide adoption of recombinant human bone morphogenetic protein-2 (rhBMP-2) in ASD surgery, outside of the original FDA indications, allows surgeons to realistically achieve osseous union in long fusion constructs. Utilization of rhBMP-2 demonstrates both superior fusion rates and improved long-term cost-utility compared with conventional bone grafting techniques (Jain et al. 2020; Kim et al. 2013).

Advances in spinal navigation technology have allowed surgeons to place pedicle screws more quickly and accurately. ASD operations are commonly revision procedures, and navigation techniques are particularly beneficial here, given the absence of normal anatomic landmarks (Flynn and Sakai 2013). Navigation technology is also helpful in the placement of pelvic fixation. Furthermore, developments in imaging technology, specifically the EOS biplanar imaging device (EOS Imaging, Paris, France), allow providers to better visualize and evaluate not only spinal deformities but also the relationship of the spine to the pelvis and lower extremities (Melhem et al. 2016; Wybier and Bossard 2013).

Although intraoperative neurophysiologic monitoring techniques are well established, their role in allowing surgeons to safely complete ASD operations cannot be understated. The combination of somatosensory-evoked potentials, transcranial motor-evoked potentials, and electromyographic screw stimulation allows surgeons to make intraoperative decisions in response to real-time neurophysiologic data, identifying and potentially reversing neurological injuries. These techniques represent a dramatic improvement upon the historical standard of care—the wake-up test (Glassman et al. 1995; Nuwer et al. 1995; Hilibrand et al. 2004).

Collaborative relationships with anesthesiology have improved anesthesia protocols for ASD procedures. Cardiac anesthesia techniques, such as the use of antifibrinolytics, have helped decrease not only perioperative blood loss but also transfusion rates (Peters et al. 2015). Blood and fluid management protocols have also helped to minimize coagulopathy and diminish the dreaded complication of postoperative blindness (Kla and Lee 2016).

## Future Directions

In less than 20 years, our ability to understand and treat ASD has expanded logarithmically. Operations once deemed too dangerous to consider are now performed on a routine basis worldwide. The continued emphasis on data collection, particularly by research consortia and society-driven databases, will continue to advance our understanding of the treatment of ASD. The assimilation of information technology methodologies represents the next evolution in ASD surgery. Artificial intelligence, predictive modeling, and machine learning techniques are rapidly being deployed to assist in decision-making algorithms for patients with ASD (Chang et al. 2020; Joshi et al. 2019). These modalities can evaluate data sets beyond human comprehension, searching for patterns that are otherwise occult. A critical human component will nonetheless remain: without the continued collection of high-quality data and intelligent interpretation of results, these powerful data analysis techniques are impotent.

The shift in a surgeon's perspective on patients with ASD, from perioperative episodic–based care to understanding the longitudinal nature of ASD care, will continue. Preoperative optimization of patients with modifiable risk factors is the foundation of this new approach; however, understanding the associated psychosocial component of ASD and its effects on patient outcomes is equally important (Sethi et al. 2014; Sikora et al. 2020). In an increasingly cost-conscious healthcare environment, demonstrating the patient-centered value of expensive ASD operations will play an outsized role in the future of spinal deformity care (Glassman et al. 2020). The incorporation of new operative techniques, such as percutaneous instrumentation, lateral interbody cages, and robotically assisted operations, must be studied and validated from the standpoint of outcomes *and* cost-effectiveness.

The editors of this textbook have assembled an outstanding group of thought leaders to provide the most up-to-date information on ASD. Without question, this will become an indispensable textbook for those caring for patients with ASD.

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# CHAPTER ONE

# TRANSITIONAL SPINE CARE: CARE OF YOUNG ADULTS 18–25 YEARS OLD

DENNIS P. DEVITO, M.D.[1,2,3]

[1] MEDICAL DIRECTOR, SPINE PROGRAM, CHILDREN'S HEALTHCARE OF ATLANTA, ATLANTA, GEORGIA, U.S.A.
[2] CONSULTING ASSOCIATE, DUKE UNIVERSITY ORTHOPEDICS, DURHAM, NORTH CAROLINA, U.S.A.
[3] CLINICAL ASSISTANT PROFESSOR, EMORY UNIVERSITY ORTHOPEDICS, TUCKER, GEORGIA, U.S.A.

## Abstract

Young adults 18–29 years of age with scoliosis, with or without prior spinal surgery, are a poorly characterized patient group, because they are transitioning from traditional pediatric to adult care, and may be unaware of the need for close monitoring. The expected surgical results are influenced by spinal curve progression and related back pain, particularly that associated with degenerative spine disease, along with changes in spinal flexibility and perioperative complication rates. Innovative spinal surgery techniques previously used to treat adults may be combined with typical deformity correction to achieve optimal outcomes in young adults. Considerations for early surgical intervention are highlighted.

## Introduction

Transitional spine care lacks a set definition but may be considered to apply to individuals aging out of the typical pediatric age range and entering adulthood. From a practical standpoint, this age ranges from 18 to 29 years. Scoliosis in this age group is referred to as young adult scoliosis (YAS). This designation is important because this subgroup of patients with spine deformity is often neglected: patients might cease to be followed by specialists in pediatric deformity and subsequently experience delays in starting care with physicians specializing in the adult spine. In addition, most patients with YAS are much younger than the typical patients treated by surgeons specializing in adult deformities. Many young adults might be unaware of the need for continued or long-term follow-up for spine care, regardless of whether they have previously been observed, have worn braces, or have undergone surgery. Patients may lack clear or well-facilitated access to surgeons with knowledge and familiarity regarding issues of youth and the aging spine. Thus, clear identification and characterization of this group of young adults provides benefits to patients.

Limited data specific to YAS are available, according to a literature review; consequently, nuances in treatment have not been well defined. Subtle changes in the spine in young adults, such as flexibility and early degenerative disease, must be elucidated to enable optimal surgical management. However, great opportunity exists for innovation, such as the application of certain adult procedures, in combination with standard traditional adolescent techniques, in the surgical management of young adult patients, when indicated. Some crossover certainly exists between the goals of adolescent deformity correction and the techniques used for adult deformity correction. Specifically, the emphasis on ideal sagittal balance and preservation of spinopelvic relationships is particularly relevant but may be overlooked by pediatric surgeons treating deformity of the immature spine. The margin for neglecting these anatomic insights may be narrower in young adults than in children, particularly regarding the long-term preservation of favorable outcomes.

The purpose of this chapter is to better define this unique category of patients with spine deformity and to provide insights into which adolescents with scoliosis are at risk of future problems and should be followed closely as young adults.

## What is Young Adult Spinal Deformity?

Adult spinal deformity (ASD) is generally divided into two main categories: (1) scoliosis present before skeletal maturity, which is usually idiopathic, and (2) scoliosis secondary to superimposed degenerative changes in the aging spine. YAS is classified in the former group, and spinal curve progression is considered nondegenerative in nature, in contrast to the etiology of typical ASD, although mild to moderate degenerative disc disease may be present in both subgroups. This discussion focuses primarily on idiopathic scoliosis, although the principles are likely to also apply to congenital and neuromuscular scoliosis.

Patients with YAS may present with several different etiologies, depending on the initial clinical conditions, but can generally be divided into two main categories. The first category includes patients with mild to moderate curvature that is observed during adolescence and is predicted to not progress, and patients who were braced and released with a similar expectation. This category may also include patients with substantial spinal curvature who deferred surgical intervention because of school or extracurricular activities, a family preference for avoiding surgery in the absence of major symptoms, or a general fear of surgery. Another less common subset of this category is patients presenting with de novo scoliosis in young adulthood, with problematic curvature not diagnosed during adolescence but presumably having an etiology of adolescent idiopathic scoliosis (AIS). This group is large, given that all patients with AIS eventually become young adults with scoliosis and consequently should receive medical attention.

The second category of patients with YAS comprises those with prior surgery during adolescence for deformity, typically posterior spinal instrumentation and fusion. These patients present as young adults because of adverse developments associated with prior surgery, such as the adding-on phenomenon, progression of the unfused portion of the spine, truncal imbalance, junctional deformity (proximal or distal), hardware failure or nonunion, degeneration distal to the last instrumented level, and/or associated worsening back pain. Although all patients do not require close follow-up indefinitely after surgery, identifying patients with risk factors for future problems is critical, so that periodic observation during young adulthood can be recommended, and early referral to a qualified spine provider can be achieved.

## Nonoperative Spinal Curve Behavior and Surgical Indications

The decision to undergo spinal reconstructive surgery in young adults follows similar guidelines to those for skeletally immature patients but carries less urgency. Because rapid spinal curve progression is generally not a concern, attention can be focused on curve magnitude, curvature-related pain, or other exacerbating symptoms (such as curve location and cosmetics) as surgical indications. Functional limitations and pronounced sagittal deformity, frequent components of ASD, are generally not factors in YAS. To appropriately advise patients regarding surgical intervention, a basic understanding of spinal curve behavior, with or without prior treatment, is essential. In addition, the rate of perioperative complications associated with the proposed procedure, as they relate to advancing age, should be delineated to assess surgical urgency.

Long-term studies of patients with untreated AIS have reliably demonstrated curve progression of 0.5°–1° per year, after skeletal maturity, for curves with a magnitude of 50° or greater (Weinstein et al. 2008; Ascani et al. 1986). Thus, this number has become a standard threshold for recommending prophylactic spinal fusion, although curve progression may not follow a linear pattern over a patient's lifetime. However, recent evidence indicates that female patients with curvature below rather than above 50° at skeletal maturity may progress at faster rates, as demonstrated by Grothaus et al. (2020) in a cohort of 89 patients with an average curve of 33° at skeletal maturity, which progressed 2.3° per year during a 2-year follow-up. Approximately half the patients progressed 5° or greater, 19% progressed 10°, and 12.4% crossed the 50° threshold and subsequently underwent surgery. The authors concluded that curves 40° or greater at relative skeletal maturity were 150 times more likely than curves less than 40° to progress to 50°, and even curves less than 40° had the potential to increase (figure 1-1).

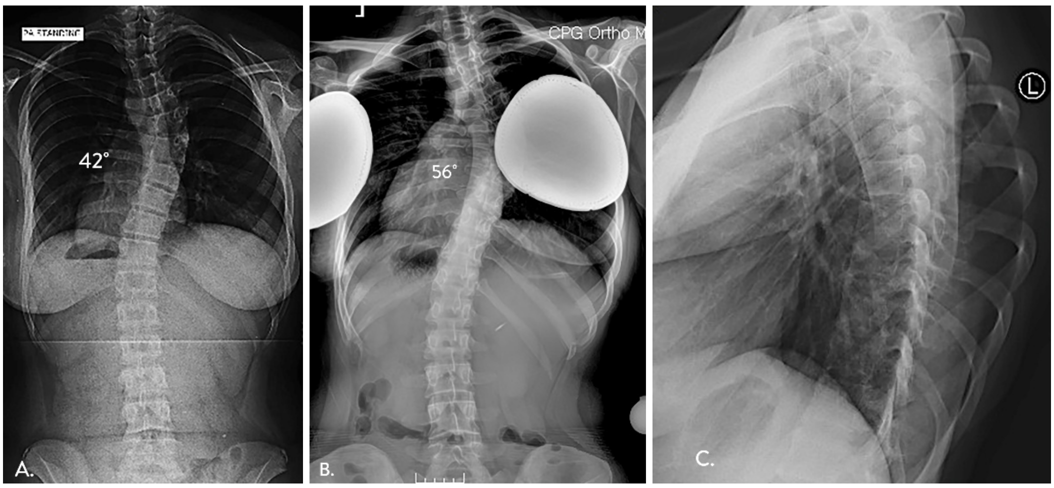


Figure 1-1. (A) A 15-year-old girl with an asymptomatic 42° thoracic curve. (B) At the age of 20 years, 15° progression (57°), greater shoulder tilt, and back pain. (C) Significant rib rotation, producing pain and prominence.

Although moderate single thoracic curves may be fairly well tolerated in patients with YAS, growing evidence indicates that compensatory lumbar curves and thoracolumbar/lumbar (TL/L) curves can be problematic well below the 50° threshold. Concerns apply to both curve progression and the development of clinically significant low back pain (LBP).

Ohashi et al. (2018a) evaluated 56 women with TL/L scoliosis with an average age of 39 years and a mean Cobb angle of 37° at adolescent maturity, which progressed to 47.8° after only 12½ years (0.4° per year). The predictors of curve progression were significant apical vertebral translation and L3 tilt; however, L3 tilt exceeding 16.5° was the only independent predictor of TL/L curve progression and amounted to ≥ 0.5° per year (figure 1-3). In addition, L4 tilt exceeding 16° was positively correlated with LBP (greater Visual Analogue Scale and Oswestry Disability Index [ODI] scores than observed in controls) and early lumbar disc degeneration, as shown on MRI. Similar outcomes regarding lumbar curve behavior can be extrapolated for double major (DM) curves (Lenke 3) and thoracic curves where the apex of the compensatory lumbar curve crosses the central sagittal vertical line (Lenke 1C). In a similar study, Ohashi et al. (2018b) investigated the long-term behavior of primary right thoracic curves (Lenke 1AL, 1AR, and 1B) in a group of young adults with curves initially measuring at least 30° at skeletal maturity, focusing on changes in the lumbar component. The main thoracic curve significantly progressed in all three curve types, but the lumbar curve significantly progressed in only the type 1B group, at approximately 0.3° per year. Moreover, the L4 tilt significantly shifted to the right in both type 1AR and 1AL curves, along with a rightward C7 translation, but was of greater significance in the 1AR group (which initially had a rightward tilt). These patients also had a much greater incidence of Modic changes at the L4-L5 level on MRI examination, as well as higher Visual Analogue Scale scores for LBP (figure 1-2). Patients with AIS with primary thoracic scoliosis with a rightward L4 tilt should also be followed periodically into adulthood because of the curve changes and asymmetric loading of the lower lumbar spine; consequently, relatively early selective thoracic fusion may be indicated.

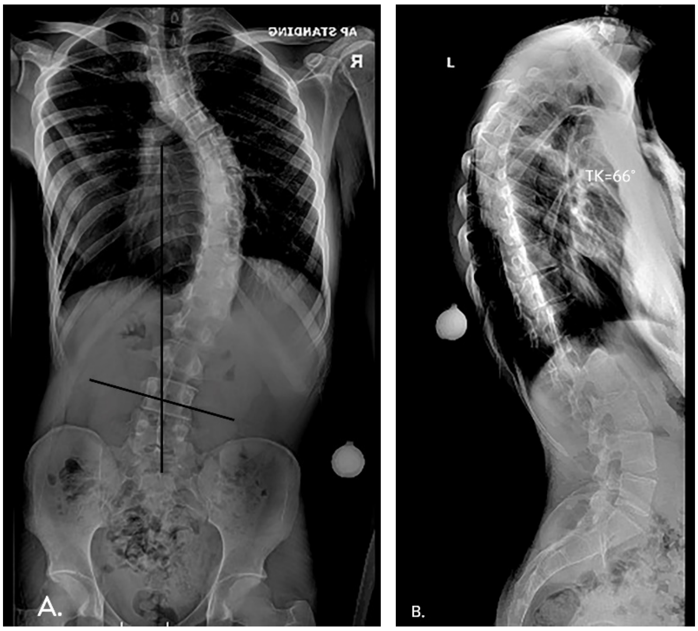


Figure 1-2. (A, B) A 21-year-old man with a progressive curve and increasing LBP associated with a 60° Lenke 1AR pattern with a right tilt of L4 and a 7.4 cm apical trunk shift.

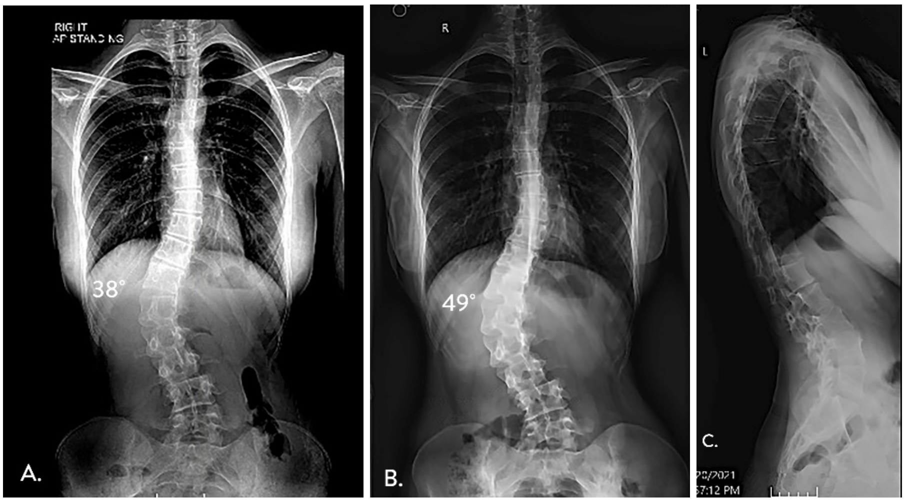


Figure 1-3. (A) A 16-year-old girl with an asymptomatic 38° thoracolumbar curve at skeletal maturity, with last touched vertebrae L3 and 18° tilt, and 3.3 cm apex translation. (B) Six years later.

Patients with YAS often present with a complaint of LBP, although the causal relationship between scoliosis and nonspecific back pain is somewhat nebulous. Yuan et al. (2019) in a comparison of nonspecific LBP between young adult women 18–30 years of age with or without lumbar scoliosis, have observed that the pain was unilateral and on the convexity in most patients with scoliosis. Lumbar prominence was also suggested to contribute to the discomfort complaint. In contrast, the nonscoliotic group usually showed midline or symmetric pain over a greater area. However, in contrast to findings from other studies, nonspecific LBP was less severe in the group with scoliosis, presumably because of the absence of any superimposed degenerative disease. In a large study of health-related quality of life (HRQOL) in 1,187 adults 38 years old, on average, with scoliosis, Diarbakerli et al. (2018) determined that brace-treated and untreated individuals with Cobb angles 30° or greater had lower Scoliosis Research Society (SRS)-22R subscores in all domains than individuals with Cobb angles below 30°. However, the HRQOL scores did not appear to decrease with age in untreated patients. In another review of young adults with moderate scoliosis at skeletal maturity, Watanabe et al. (2019) demonstrated curve progression into adulthood for all curve types at 0.5° per year, with significantly worse SRS-22 scores in the self-image domain, and significantly worse HRQOL scores for LBP and walking ability in the TL/L and DM scoliosis groups than in age- and sex-matched controls. In a comparison of back pain in adolescents (with a mean age of 15 years) and young adults (with a mean age of 22½ years), Fekete et al. (2019) have shown that, despite matched curve severity, back pain scores above 4/10 were observed in 52% of the YAS group but only 38% of adolescents ($p = .004$).

The decision to delay spinal surgery until young adulthood in patients with AIS involves delineating whether significant differences in outcomes exist among age groups. A primary consideration relates to the loss of flexibility with age and how outcomes might be affected, particularly regarding fusion levels and perhaps the surgical approach. Chen et al. (2011) in a comparison between skeletally immature and mature patients with AIS, have shown that curve flexibility decreases with age, and this decrease occurs very slowly in adolescents but dramatically and rapidly in adults. In addition, an increasing Cobb angle adversely affects flexibility; therefore, delaying surgery may allow for curve progression and confound the issue of aging. Zhu et al. (2017) in a matched cohort of patients with AIS and patients with YAS undergoing surgery, observed a difference of 8% in curve flexibility despite an average age interval of only 8 years between groups. Further analysis by Deviren et al. (2002) has demonstrated that the flexibility of structural TL/L curves is inversely correlated with curve magnitude and age: every 10° increase in curve magnitude above 40° was associated with a 10% decrease in flexibility, and every 10-year increase in age was associated with a 5% decrease in flexibility. In addition, for every 10-year increase in age, the flexibility of the fractional lumbosacral curve decreased by 10%. Additionally, axial pain correlated with advancing age but was not predicted by curve magnitude, and radicular pain tended to increase with age and the magnitude of lateral listhesis. Therefore, progressive loss of flexibility with age, particularly in conjunction with an increase in curve size, has implications for deformity correction and the maintenance of caudal mobile segments.

Another concern relates to the relationship between age and complications in scoliosis surgery. In an analysis of the SRS database, Shaw et al. (2016) reviewed the cases of 5,470 adult patients and observed an overall complication rate of 13.5%, among which dural tears, deep infections, and implant failures were the most common problems. The data revealed a statistically significant increase in complication rates with each decade of age, from 7% in the 19-year-old group to 10% in the 30-year-old group, to 24% in the group older than 80 years. An independent risk factor for increased perioperative complications is the addition of spinal osteotomy to the surgical procedure, as often occurs in the correction of relatively large and stiff curves (Wu et al. 2021). Other perioperative measures that should be considered between patients with AIS or YAS are the estimated blood loss (EBL), number of spinal levels fused (including the lowest level), duration of surgery, and length of hospital stay. In direct comparison, both Lonner et al. (2019) and Lavelle et al. (2020) have reported more levels that were fused, more distal fusion levels at or below L4, and greater EBL in patients with YAS rather than AIS, whereas the length of stay varied but was longer in patients with YAS. Additional surgeries were required only in the YAS cohort. Chan et al. (2021) have also observed stiffer curves and a trend toward less correction in a study of propensity-matched women with AIS versus YAS; however, they did not observe significant differences in the perioperative outcomes of operation time, EBL, or length of stay. This finding was attributed to the operative strategy of two experienced spine surgeons performing the surgery together.

In summary, curve magnitude may not be the only consideration in spinal reconstructive surgery, although this single indication may be appropriate for young adults. Other issues include pain resolution, loss of flexibility with aging, effects on curve correction and the extent of spinal fusion, and elevated incidence of perioperative complications.

## Curve Behavior after Adolescent Spinal Surgery

The indications for spinal instrumentation and fusion for AIS are well delineated in the literature. Most patients are healthy and asymptomatic, although approximately one-third have minor curve-related back discomfort. The surgical goals, beyond arresting curve progression, are to create balanced spinal alignment in three dimensions, achieve favorable cosmesis, avoid subsequent operative interventions, and do no harm. Spinal instrumentation and fusion always carry some risk in disrupting an otherwise balanced spine and creating an iatrogenic malalignment in the coronal and/or sagittal plane, thereby resulting in a need for revision surgery in the near term. Furthermore, the incidence of creating pain in a previously asymptomatic patient, regardless of etiology, is undesirable and defies the aim of achieving good long-term outcomes. In a multicenter review of patients with AIS, Hariharan et al. (2020) documented a 10-year complication rate of 12% and a reoperation rate of 6.7%, in agreement with other reports (Weiss and Goodall 2008). The most common structural complications include adding-on phenomena, junctional kyphosis, caudal disc degeneration, nonunion with or without implant failure, and associated increased back pain.

## Coronal Imbalance

The challenge of selecting which spinal levels to instrument is based on the desire to preserve the greatest lumbar range of motion possible, thus presumably maintaining function. This goal is the impetus for selective thoracic fusions and sometimes fusing short of the end vertebra, usually of the lumbar component of a curve. Coronal decompensation is frequently associated with selective fusion of Lenke 1B and particularly Lenke 1C, 2C, and 3C curves, thus potentially explaining why surgeons chose selective thoracic fusions for Lenke 1C curves only half the time. In a review of selective thoracic fusions for Lenke 1C and 2C curves, Kwan et al. (2018) observed a 20% incidence of coronal decompensation, and a 25% incidence of adding-on phenomena in their cohort, although no patients required revision surgery during the 2-year follow-up period. However, others have reported a 6%–8% surgical revision rate for this problem (Chang et al. 2010; Ahmed et al. 2017). Edwards et al. (2004) have identified preoperative decompensation as a risk factor for postoperative coronal imbalance, and inferior outcome scores in patients with decompensation greater than 2–5 cm postoperatively (figure 1-4). Substantial decompensation occurred when the lower instrumented vertebrae (LIV) were located distal to the stable vertebra; however, improvements in imbalance have been observed by Ishikawa et al. (2017), and the stability of the lumbar curve is usually maintained, as verified in a 20-year follow-up by Larson et al. (2012). Coronal imbalance after posterior spinal instrumented fusion (PSIF) can also occur in the treatment of primary thoracic scoliosis, particularly with large and stiff curves, and is associated with significantly diminished HRQOL scores. Severe residual imbalance exceeding 4 cm is 23.8 times more likely to occur when the preoperative curve flexibility is below 20% (Anari et al. 2020).

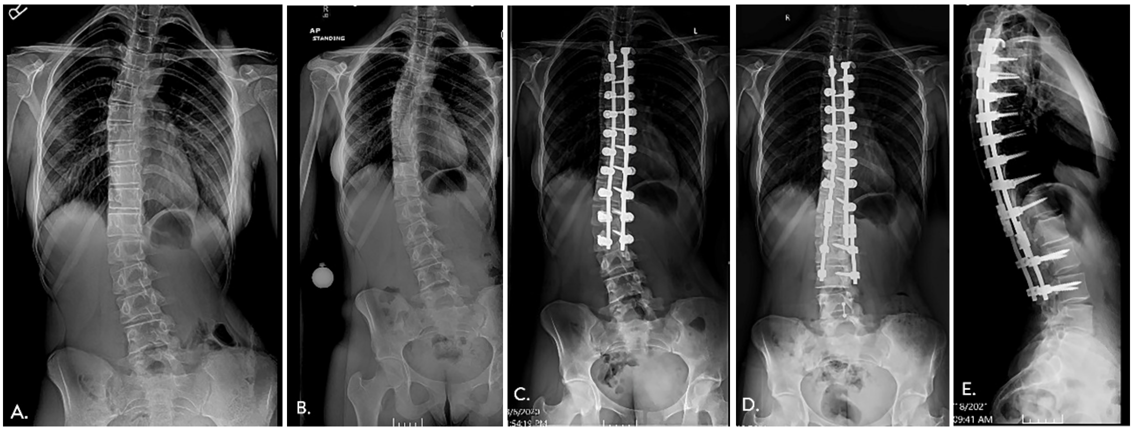


Figure 1-4. (A) A 14½-year-old skeletally mature girl with a prior history of bracing terminated 1 year prior, now with a 30° curve and mild trunk decompensation to the right. (B) At the age of 18 years, curve progression to 41°, increased truncal decompensation to 6.8 cm, and back pain. (C) At 2 years after PSIF, with LIV L2 (cephalad to last touched vertebrae) and worsening LBP, L2-L3-disc wedging of 11°, and L3 tilt of 18°. (D) At the age of 21 years, revision surgery with caudal extension to L3 E and (E) preservation of sagittal balance.

The adding-on phenomenon is defined as a postoperative deterioration of the curve below the LIV, thus increasing the number of vertebrae within the curve, the translation of the first vertebra below the LIV (greater than 10 mm from the central sacral vertical line [CSVL]), and the angulation of more than 5° of the first disc space below the LIV. In a meta-analysis of the risk factors for adding-on phenomena after surgery for idiopathic scoliosis, Yang et al. (2018) reported an overall prevalence of adding-on phenomena of 14%, with a range of 9%–16% according to scoliosis type. The authors concluded that LIV selection is an independent risk factor for adding-on phenomena and is more likely to occur when the LIV is cranial to the end vertebra or stable vertebra. Suk et al. (2003) have observed an elevated risk of adding-on phenomena when the LIV is proximal to the neutral vertebrae by more than two vertebrae, and have recommended extending fusion to the neutral vertebrae if it is located below two levels from the end vertebra. Lakhal et al. (2014) have suggested that the LIV should be the vertebra above the first flexible intervertebral disc, as demonstrated on coronal and sagittal benders, to avoid this complication. Finally, in rigid TL/L curves, selecting the lowest instrumented vertebra is often based on balancing the goal of preserving as many mobile lumbar segments as possible with the ability to achieve better curve correction and to avoid possible disc wedging and vertebral tilt below an excessively cephalad LIV. A disc wedge angle exceeding 5° and an L4 tilt angle exceeding 10° are associated with poor outcomes when the LIV is L3 (figure 1-5). Nohara et al. (2015), in a 10-year follow-up study, have observed that greater L4 tilt angle and L3-L4-disc wedging after lumbar fusion are associated with disc degeneration. However, in a study by Cho et al. (2019), the most increased tilt and disc angulation were observed at 6 months postoperatively; subsequently, spontaneous improvements in L4 tilt and trunk shift occurred and were maintained during the 5-year follow-up (although the disc wedge angle did not change). For primary TL/L curves, Zhuang et al. (2021) have recommended the following criteria for selecting the LIV: the most cephalad vertebra touched by the CSVL, 25% rotation or less on a standing anteroposterior radiograph, and the CSVL crossing between pedicles on the lateral bend and not at the apex of kyphosis, although adding-on phenomena still occurred in 8.7% of cases. LaValva et al. (2021) have also added the parameters of body mass index above 28 kg/m$^2$, apical vertebral translation above 6 cm, L3 angulation greater than 25°, and greater than 4 cm L3 translation from the CSVL; if these parameters are not met, selection of a more caudal LIV is required.

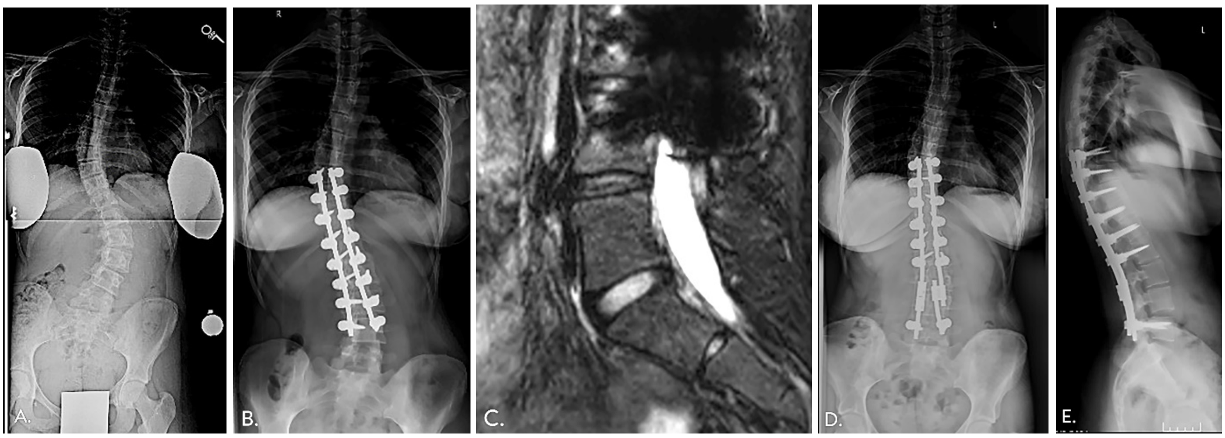


Figure 1-5. (A) A 16-year-old girl with a 57° Lenke 5 curve from T11 to L4, and mild pelvic obliquity treated with PSIF from T9 to L4. (B) At 4 years postoperatively (age 20 years), 8° disc wedge and 12° tilt of L5, residual translation of L4-L5, and LBP unresponsive to conservative measures. (C) MRI showed a healthy L5-S1 disc (CT scan demonstrated solid fusion). (D, E) Revision PSIF with extension to L5 with improved coronal balance, maintenance of sagittal alignment, and resolution of LBP.

## Sagittal Imbalance

Except for several typical early complications of scoliosis surgery such as wound issues or infection, the primary problems are due to sagittal misalignments resulting in a junctional deformity created by the surgery. Most adolescents are balanced preoperatively, although Abelin-Genevois et al. (2018) have brought attention to patients with AIS with abnormal sagittal regional deformities that affect the surgical plan regarding rod contouring and instrumentation levels. Although the exact cause of junctional deformity remains obscure, disrupting sagittal balance may be the central issue. Fortunately, very few patients with AIS or YAS require instrumentation across the lumbosacral region, where respecting spinopelvic parameters is critical for good long-term results, according to studies of ASD (Schwab et al. 2010). Most thoracic fusions extending no more caudally than L2 have little effect on lumbopelvic alignment postoperatively, and any changes in corresponding lumbar lordosis (LL) typically return to normal after 6–12 months (Clément et al. 2019; Yeung et al. 2021). Problems with flattening of the already hypokyphotic thoracic spine can result in proximal junctional kyphosis (PJK), and attention must be paid to preexisting kyphosis from T10 to L2, to avoid creating distal junctional kyphosis by failure to include the junctional level in the instrumentation (figure 1-6). An improper rod contour with a "lordotic tail" at the LIV can also produce distal junctional kyphosis, which has an overall incidence of approximately 5%–7%.

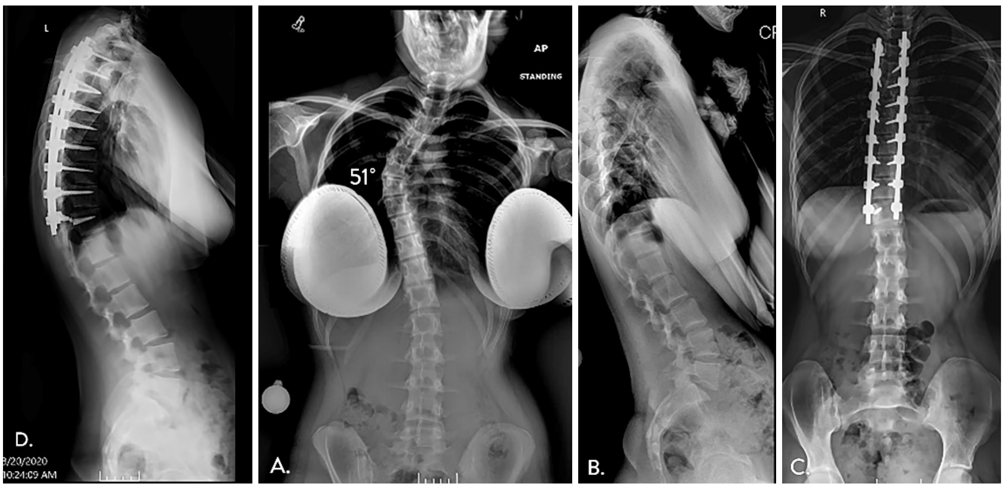


Figure 1-6. (A) A 15-year-old girl with a 51° thoracic curve. (B) Abnormal sagittal profile with thoracolumbar kyphosis of 21° (T10-L1), TK of 33°, and LL of 62°. (C) A 5-year follow-up at the age of 20 years, with maintained coronal alignment but broken screws at T11. (D) Moderate distal junctional kyphosis due to excessive cephalad LIV selection (at apex kyphosis). Increases in thoracolumbar kyphosis to 24°, TK to 43°, and LL to 65°, with a sagittal vertical axis of 0. However, the patient remained asymptomatic.

PJK is a more common problem, occurring in 8%–25% of AIS cases. However, radiologically measured PJK does not frequently translate to major clinical problems or necessitate surgical revision (Cho et al. 2015) (figure 1-7). In one causal mechanism, the hyperkyphotic thoracic spine is excessively overcorrected or flattened. Lonner et al. (2017) have noted that the risk of developing PJK increases by 7% with every degree of kyphosis lost during correction. A pelvic incidence (PI) above 55° generally requires a matching higher degree of thoracic kyphosis (TK). An additional risk factor for PJK is failing to recognize hyperkyphosis from T1 to T4, and selecting the uppermost instrumented vertebra caudal to this level. Finally, Burton et al. (2020) have suggested overcorrection of LL as an etiology of PJK, emphasizing the need to maintain the relationship between preoperative PI and LL. Low PI and LL warrant careful attention to the rod contour, even when the LIV is at L3 (and particularly at L4), to avoid overlordosis of this segment and a posterior shift of the uppermost instrumented vertebra. In summary, proper postoperative sagittal balance requires creating TK within the ideal physiologic range of 20°–45°; maintaining lumbopelvic congruence (PI – LL = ~9°) and avoiding overcorrection of LL; and keeping the inflection zone between T10 and L1, with a sagittal Cobb angle of approximately 0° in this zone, to decrease the risk of compensatory malalignments and later back pain (Ilharreborde 2018).

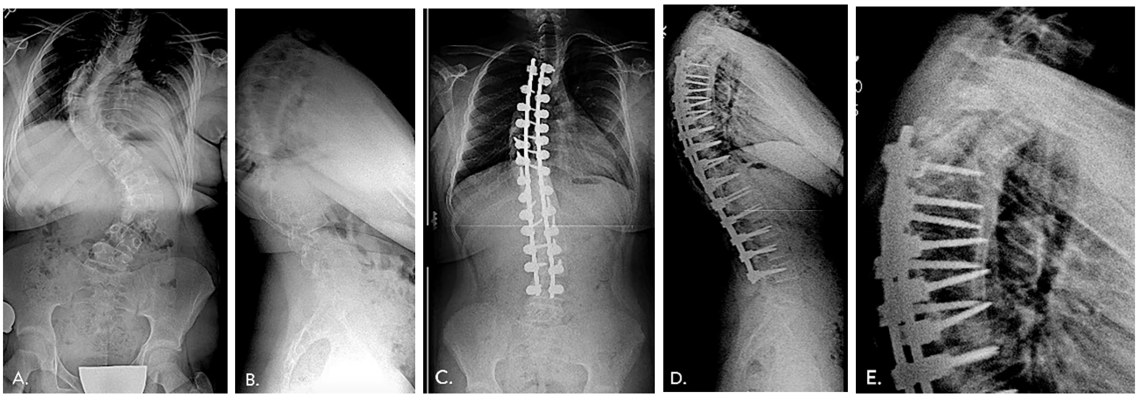


Figure 1-7. (A) A 12-year-old girl with a large DM curve of 87°/74° and a high body mass index. (B) Preoperative sagittal parameters: low PI of 30°, LL of 47° (PI − LL= −17°), and TK of 50°. (C) Balanced coronal correction. (D) At 2-year follow-up, LL of 37° but instrumented TK of 34° with asymptomatic PJK.

## Disc Degeneration and Low Back Pain

Given the association between LBP and scoliosis, approximately 50% of patients with YAS present with relevant back pain—an incidence nearly twice that in adolescents (Fekete et al. 2019). Additionally, Watanabe et al. (2019), in a 25-year follow-up study of untreated patients with AIS (average age of 30 years), have identified that TL/L curves greater than 30° are a factor associated with greater LBP and lower HRQOL scores in middle age. Fortunately, the patient-reported outcomes in all domains improved after scoliosis surgery in patients with AIS or YAS, and the long-term fusion results were comparable to those in individuals with untreated scoliosis. However, the pain scores were slightly lower (negative effect) for fusions extended below L3 (Diarbakerli et al. 2018). Whereas Upasani et al. (2008) have shown that, despite stable satisfaction scores, pain levels increased 5 years after surgery in patients with AIS, no correlation was observed with the level of the lowest instrumented vertebra. In a 10-year follow-up after fusion for AIS, Lonner et al (2018) found that 7% of patients had significant degenerative disc disease (DDD), and a LIV at L4 was associated with the highest risk. In fact, disc wedging exceeding 5° and translation exceeding 2 cm subjacent to the LIV produced a six-fold increase in DDD development. Ohashi et al. (2018a) have also identified that a residual L4 tilt greater than 16.5° correlates with more back pain and DDD. In a group of patients with AIS with no preoperative DDD and caudal fusion no lower than T12, Akazawa et al. (2020) noted that the residual lumbar curve developed at least one segment with DDD after 5 or more years of follow-up (47% incidence). The DDD group had greater residual lumbar curvature, but the percentage of DDD involvement was similar for each intervertebral disc level from L3 to S1. Ghandhari et al. (2018) have identified DDD in 15% of patients with AIS preoperatively, and in another 16% of patients after only 5 years postoperatively, noting that most of their cohort had fusions to the L4 level. Although corrective surgery for YAS can result in significantly better patient-reported outcomes and stopping of curve progression (Zuckerman et al. 2021), the potential for further improvement may be enhanced by identifying preoperative disc disease, appropriate LIV selection, and optimal residual spinal alignment and balance.

## Nonunion and Implant Failure

Nonunion is an uncommon feature of AIS surgery, as is implant failure, such as rod breakage or screw fracture; the true incidence is obfuscated by the stability provided by the use of two rods and segmental vertebral fixation. Frequently, the cause of late pain, particularly at the caudal instrumented level, is due to nonunion, as often suggested by screw loosening or fracture (Klineberg et al. 2016). Clinical suspicion may be confirmed with thin-section CT scans of the involved area (Patel and Spiker 2008). The incidence is higher among non-idiopathic than idiopathic patients;

nonetheless, the need for revision surgery during young adulthood is likely. The approach may entail the replacement of the screw with a larger diameter and augmentation of the fusion mass; an anterior discectomy and interbody fusion at that level; or both (figure 1-8). Using both techniques may be more appropriate for patients with YAS to ensure solid fusion.

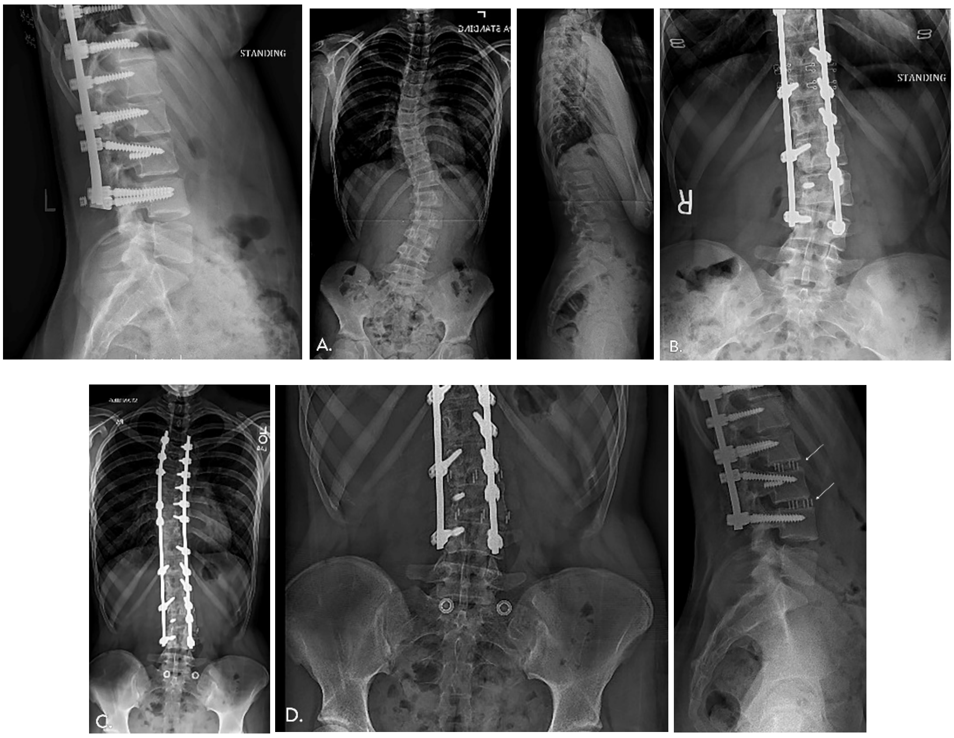


Figure 1-8. (A) Anteroposterior/lateral radiograph of a 13-year-old skeletally immature girl with 35°/49° curve treated with PSIF to L3, with subsequent adding-on phenomenon and screw fracture at LIV 2 years postoperatively, revised to L4. (B) At the age of 19 years, increased LBP, screw cap dislodgement, and L3-L4 nonunion, are associated with an 18° tilt of the LIV. (C, D) Reinsertion of caudal screws and then two-level X-LIF to restore coronal and sagittal balance, and obtain anterior fusion.

## Surgery for Young Adult Scoliosis

Although the reasons for performing scoliosis surgery in young adults who have experienced problems from prior adolescent surgery are relatively straightforward, the recommendation for intervention for curves of similar magnitude to those in older adolescents is multifactorial and requires detailed communication regarding goals and expectations, particularly because treatment decisions are no longer brokered by parents or guardians. Indications may include significant curve progression, worsening curve-associated back pain not responsive to standard conservative measures, or simply the absolute magnitude of scoliosis. Questions that must be answered include whether delaying the surgery beyond the age of 30 years is associated with greater morbidity, whether similar correction results are achieved and similar regions of the spine are fused, and whether waiting might be associated with further complications. These considerations are the same as those discussed in decision-making for mature adolescents with significant scoliosis.

A comparison of surgical outcomes of idiopathic scoliosis in adolescents and young adults by Lavelle et al. (2020) has revealed that, in general, more spinal levels are fused, particularly to L4 or below, and a lower major Cobb correction is obtained, in patients with YAS rather than AIS. This finding has been corroborated by another matched-pair analysis of 160 patients by Zhu et al. (2017) in which the results were attributed to significantly less flexibility of the spine in the young adult deformity group. Spinal alignment was successfully maintained in both groups, and HRQOL analysis demonstrated no major differences at the 2-year follow-up (figure 1-9). In the first study, greater EBL was observed in patients with YAS rather than AIS, but the complication rates were similar. However, in the second study, the adding-on phenomena were significantly greater in patients with AIS rather than YAS; this finding was attributed to further spinal growth in the younger group and a lower LIV in the older group. Avoiding this problem may be a potential advantage of waiting until maturity before PSIF. In a comparison between patients undergoing AIS surgery and a slightly older adult cohort of patients with ASD (average age 44 years), Riouallon et al. (2016) have reported a higher complication rate, with more revisions, infections, and pseudarthroses, in the latter age group. In another matched study comparing adolescent patients and adults close to the typical age range of patients with YAS, Lonner et al. (2019) documented more spinal levels fused, with one-third extended to the pelvis. In addition, longer operative times and greater EBL were observed in the YAS group than in the AIS group, in which no spinal levels were fused to the pelvis. Again, the rate of major complications was significantly greater for YAS than AIS (25% versus 5.4%, respectively). Furthermore, stiffer curves in older patients may necessitate spinal osteotomy to mobilize the spine, and this has been identified as an additional independent risk factor for perioperative complications (Wu et al. 2021).

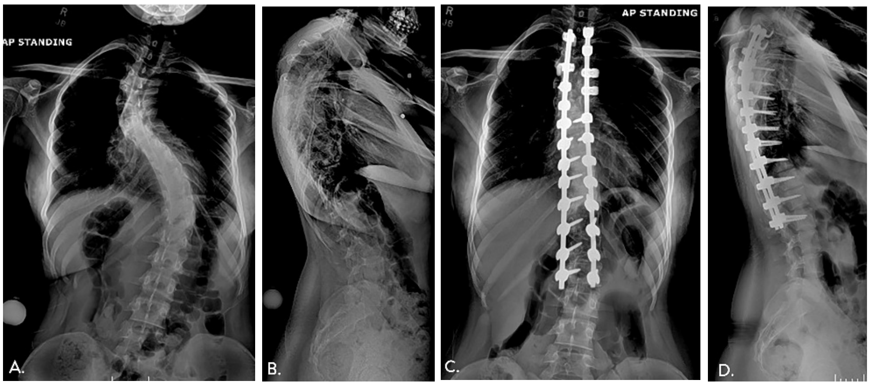


Figure 1-9. (A) A 22-year-old woman with a progressive Lenke 2C idiopathic curve (65°/62°) with 43° TK, PI = 48°, and LL = 63° (PI − LL = −15°). (B) Two years after PSIF from T2 to L2, with improved global spinal balance; 35° TK, LL = 48° now with matched PI − LL. A risk of adding-on phenomenon was identified based on the LIV one-level cephalad to last touched vertebrae; however, excellent curve correction was possible, even in this patient with YAS.

Thoracic-based scoliosis curve progression into adult life is expected with curves greater than 50° and is ultimately linked to a decline in cardiopulmonary function, thus potentially simplifying decision-making. However, TL/L curves (either primary or compensatory) have been shown to progress from curves as low as 30° in adolescents, and progression is compounded by adult lumbar degenerative disease with the potential development of sagittal malalignment. Avoiding the challenges of correcting a larger and stiffer curve, having more levels fused, and subjecting patients to a higher risk of major perioperative complications might be prudent, even without considering all the comorbidities associated with aging. In contrast, patients with YAS can expect very similar results to those in patients with AIS, with only small adjustments based on spinal flexibility and the presence of any degenerative disease (Zhu et al. 2017; Lavelle et al. 2020).

Opportunities exist to improve the results of YAS surgery through collaboration between adult and pediatric spine surgeons to combine selected surgical techniques with typical maneuvers for deformity correction. For moderately severe thoracolumbar ASD, Strom et al. (2016) have combined lateral interbody fusion with open posterior surgery and demonstrated greater coronal correction, greater improvements in Visual Analogue Scale and ODI scores, and fewer nonunions than observed with open posterior procedures alone. In another study, Bae et al. (2018) compared three surgical strategies for correcting mild to moderate thoracolumbar deformities (lateral interbody plus posterior spinal fusion; anterior interbody plus posterior spinal fusion; or posterior spinal fusion alone) and observed satisfactory radiographic outcomes in all groups. However, the patients treated with lateral lumbar interbody fusion plus posterior spinal fusion had lower rates of PJK, less back pain, less disability, and better SRS-22 scores. In another innovation for treating the rigid components of TL/L curves, Mikhail et al. (2020) have proposed a multilevel posterolateral convex disc release and removal through a transforaminal lumbar interbody fusion approach combined with PSIF, to enable better deformity correction while avoiding the morbidity associated with a formal anterior approach. The authors have demonstrated 72% major curve correction and were able to save a distal fusion level in some patients, thus decreasing the LIV tilt from 26.8° to 8.3° on average. Although most patients with YAS do not require fusion across the lumbosacral junction, enhanced alignment parameters are achieved when the apical segment of the TL/L curve is also treated with multilevel lateral interbody fusion in combination with PSIF, as compared with PSIF alone, with transforaminal lumbar interbody fusion at only L5-S1 (Theologis et al. 2017).

Ultimately, after surgery, HRQOL measurements show minimal differences between patients with AIS or YAS, and improvements are observed in both groups. Although pulmonary function generally remains unchanged after adolescent PSIF (Kato et al. 2019), a significant measurable decline in the absolute and predicted percentage pulmonary function has been observed in patients with ASD (mean age of 45 years), approximately one-quarter of whom may experience clinical deterioration (Lehman et al. 2015).

## Conclusions

Many of the considerations for recommending spinal fusion surgery in young adults coincide with those for mature adolescents with idiopathic scoliosis, particularly regarding curve magnitude and expected progression; however, curve location may also be considered. Although main thoracic curves are relatively better tolerated than TL/L and DM curves in terms of pain in young adults, curve progression to 80°, associated with thoracic lordosis, and apical rotation greater than 25° are associated with a risk of restrictive cardiopulmonary disease. Moderate curves involving the TL/L regions in adolescents have been shown to progress in adulthood, contrary to the traditional viewpoint, and are subject to further adverse changes in alignment (both coronal and sagittal) with superimposed degenerative joint disease experienced with aging. Given the commensurate loss of flexibility seen in the aging spine, late surgical intervention typically entails fusion of more caudal levels and a greater likelihood of extension across the lumbosacral junction, thus resulting in less spinal motion and greater risk of degenerative disc disease. Furthermore, the incidence of complications increases linearly with each decade of aging, thus underscoring the advantages of earlier surgery. Finally, a subset of patients with AIS who have undergone spinal surgery are at risk of future clinical deterioration due to truncal imbalance, adding-on phenomena, junctional deformity, or worsening back pain; these patients must be closely monitored for possible revision surgery during the transition into young adulthood.

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# CHAPTER TWO

# CONSIDERATIONS FOR PEDIATRIC DEFORMITY TREATED NONOPERATIVELY

STUART L. WEINSTEIN, M.D., AND LORI A. DOLAN, PH.D.
UNIVERSITY OF IOWA, IOWA CITY, IOWA, U.S.A.

## Abstract

Adolescent idiopathic scoliosis (AIS), defined as a Cobb angle >10° on coronal plane upright radiography, is a structural deformity of the spine of unknown etiology. Although most adolescents are asymptomatic, those with more severe disease (i.e., a Cobb angle >20°) are at risk for progression and thus require treatment. Nonsurgical treatments indicated for this condition include scoliosis-specific exercises (SSE) and bracing. While there is only limited evidence for the effectiveness of SSEs, the results of the Bracing in Adolescent Idiopathic Scoliosis Trial (BrAIST) provided definitive support for bracing as a means to reduce the likelihood of curve progression. Risk calculators are now available to clinicians and families and can be used to help with the decisions involved in the management of AIS.

## Introduction

Adolescent idiopathic scoliosis (AIS) is a three-dimensional structural deformity of the spine for which no known etiologic cause has been established. The diagnosis is made when other causes of scoliosis have been ruled out. Between 2% and 3% of children between 10 and 16 years of age will have this condition, defined as having a Cobb angle >10° on a coronal plane upright radiograph. Of these patients, only a few (0.3%-0.5%) will have scoliosis with a Cobb angle >20°, meeting traditional indications for active treatment. Boys and girls are affected equally but, for yet unknown reasons, girls more often require active treatment.

The vast majority of patients with AIS are asymptomatic and often present due to truncal asymmetry noted during screening or incidentally during a well-child examination. As AIS cannot be prevented, active treatment attempts to prevent curve progression into adulthood. Nonoperative treatments such as scoliosis-specific exercises (SSE) or bracing are first-line treatments if the curve meets appropriate criteria; curves measuring 50° or greater require surgical intervention to correct the deformity.

Evidence-based decisions require knowledge of the natural history of AIS and treatment goals, as well as risk factors for curve progression. Finally, the treating physician must understand the current and ever-evolving literature on the effectiveness of current nonsurgical treatments.

## Natural History

The evidence base for AIS treatment relies heavily on a small body of natural history literature, including the University of Iowa longitudinal studies. The overwhelming majority of patients with AIS have small, nonprogressive curves <20° that will not interfere with their lives and require no treatment. Curves >20° in skeletally immature patients are at risk for progression and require active treatment.

The early natural history literature of scoliosis presented a grim prognosis, suggesting that patients become disabled by back pain and die young due to pulmonary compromise. Unfortunately, these early studies included not only patients with AIS but also those with early-onset idiopathic scoliosis and those with congenital, neuromuscular, and syndromic etiologies. The unfortunate consequence of these reviews was the prevalent misperception that all types of scoliosis inevitably led to high mortality rates, disability from back pain, and cardiopulmonary compromise.

The majority of clinical treatment decisions in AIS are made based on spinal curve magnitude and progression, assuming that, if the curve worsens, the patient will develop problems such as pain, increased risk of early mortality, increasing deformity, and negative psychosocial effects. Over the lifetime of patients, the magnitude of the spinal curve generally increases. However, how much the curve actually progresses and over what time frame varies for each patient. Factors that can predict spinal curve progression include curve magnitude and location, age at diagnosis, and maturity factors. These factors include age at menarche and the amount of growth remaining, as judged by either the Risser grade of ossification of the iliac apophysis or the more reliable simplified skeletal maturity system (SSMS) (Sanders et al. 2007; Sanders et al. 2008). Curve progression is more probable in skeletally immature patients and those with larger curve sizes (even after maturity) than in patients who are skeletally mature and have smaller curves. Curves with a thoracic apex and a Cobb angle of >50° have the highest prevalence of progression.

Pulmonary function is the only symptom that is consistently associated with curve size in AIS. Pulmonary function can be affected by other factors in addition to curve magnitude, including decreased respiratory muscle strength and the degree of thoracic lordosis and vertebral rotation. Unlike in early-onset idiopathic scoliosis, pulmonary hypertension, and right heart failure are rare occurrences and then only in patients with severe thoracic curves. In AIS, thoracic curves with a Cobb angle of >50° are associated with reduced vital capacity and more frequent shortness of breath than those experienced by patients with smaller spinal curves. Large thoracic curves are only rarely associated with severe cardiopulmonary compromise (Weinstein et al. 1981).

With respect to back pain, Deyo et al. (2006) report that approximately 50% of adults who do not have scoliosis have an episode of low back pain in any particular year, and 15% report frequent back pain or pain that lasts for >2 weeks in a given year. Most long-term follow-up investigations of patients with AIS report a frequency of back pain in these individuals similar to that in the general population. In contrast, the Iowa 50-year AIS natural history follow-up reported that patients with AIS had a greater frequency, intensity, and duration of chronic back pain than the general population (Weinstein et al. 2003). These patients, however, were *not* disabled. They were able to work and undertake everyday activities at a level similar to that of their unaffected peers. Although most patients with AIS will develop radiographic osteoarthritis changes, the presence or absence of radiographic osteoarthritis or curve severity has not been strongly correlated with the history of backache. This lack of association is also true for back tenderness to palpation, except for areas of lateral listhesis in lumbar and thoracolumbar curves. Lumbar and thoracolumbar curves have the highest frequency of back pain compared with other spinal curve patterns (Weinstein et al. 1981; Weinstein et al. 2003).

With respect to overall function and self-esteem in patients with AIS, the literature is sparse and conflicting. Patients with scoliosis compare favorably to age- and sex-matched controls in terms of the psychological aspects of the condition, including the presence or absence of clinical depression. Unfortunately, older patients with AIS who have not been treated are much less satisfied with their body image and appearance compared with individuals without the condition. One-third of these patients feel that their spinal curvature has restricted their life in some way. They express difficulty in purchasing clothing, reduced physical ability, and increased self-consciousness (Weinstein et al. 2003).

Over their lifetime, untreated patients with AIS generally function well. As young adults they become employed, form relationships, have children, and become active older adults. With increasing age, most patients with untreated AIS can have back pain and feel they have important cosmetic concerns. Accordingly, treatment recommendations such as watchful waiting, physiotherapy, bracing, and surgery must be decided on an individual basis. The patient and their family should be well educated about the natural history of the disease to help them make informed decisions.

## Nonsurgical Management for "At-Risk" Patients

"At-risk" patients are those who have a high risk of curve progression to surgical indications (curve >45°-50°) if left untreated. This group includes patients with a curve between 20° and 40° who are skeletally immature. Skeletal maturity is generally assessed by various maturity markers, including menstrual status in girls, status of closure of the triradiate cartilage, ossification stages of the iliac crest (Risser grade), and digital skeletal age as determined by the SSMS. Although other markers exist; for example, thumb ossification composite index (TOCI) (Hung et al. 2017; Hung et al. 2018), olecranon ossification (Greene et al. 2021), and proximal humeral ossification (Li et al. 2018), these are the most commonly used in current practice. The high-risk group is generally those with an open triradiate cartilage, Risser <2, and SSMS <4. All of these patients require active treatment. Treatment options include SSE, bracing, or a combination of both.

### *Exercises*

Although SSE programs continue to gain popularity around the world, the literature is somewhat limited. Since 2003, over 10 systematic reviews/meta-analyses have been conducted to estimate the effect of SSE on various clinical and radiographic outcomes of AIS, the most recent being published in 2020 by Fan et al. (2020). These studies suggest that SSE may have a positive effect, but their conclusions are limited by the heterogeneity of patient characteristics, SSE programs, and outcomes assessed across the various studies.

### *Bracing*

Modern brace treatment for AIS began in the late 1940s, with the development of the Milwaukee brace. This orthotic, originally developed as a postsurgical alternative to casting, was soon widely adopted around the world as the standard of nonsurgical care for AIS, despite the lack of any high-quality evidence supporting its effectiveness. Most modern braces use a system of intermittently applied forces to apply distraction and lateral pressure to reduce the deformity. The goal of treatment is to prevent further curve progression, but the ultimate goal of patients and parents is to avoid having the curve reach the surgical threshold of >45°. Early reviews of brace treatment at our own institution led to equivocal results. A summary of systematic reviews in 2009 demonstrated considerable variability, and hence inconclusive and inconsistent evidence, concerning the effect of bracing on the risk of surgery (Dolan and Weinstein, 2009).

This lack of high-level evidence led to the Bracing in Adolescent Idiopathic Scoliosis Trial (BrAIST). The primary aim of BrAIST was to compare the risk of curve progression to 50° or greater before skeletal maturity (a common indication for surgery) in subjects treated using a brace with those treated by observation. Secondary aims included a comparison between health and functioning, quality of life, and self-image over time in the two treatment groups, as well as a determination of the relationship between bracing dose (wear time) and curve response. An additional aim was to develop predictive models based on individual patient characteristics at initial presentation (e.g., sex, skeletal maturity, chronological age) and curve characteristics (e.g., curve magnitude, location) to establish baseline risk. Models including these variables plus treatment characteristics, such as hours of brace wear, were then used to establish risk reduction associated with treatment.

BrAIST was planned and funded as a randomized study, but a preference arm was added during the course of the study to address slow enrollment. This allowed patients to participate by choosing their own treatment. Therefore, the final design included both a randomized cohort and a preference cohort, with identical inclusion criteria, protocols, and outcomes assessments (Weinstein et al. 2013a). Enrollment began in March 2007 and was completed in February 2011. The trial was terminated when an interim analysis provided definitive evidence that bracing decreased the risk of curve progression relative to observation (Weinstein et al. 2013b). The primary analysis (combining the randomized and preference groups) yielded an adjusted odds ratio of 2.03 (95% CI, 1.12 to 3.68; $p < .0197$), indicating a treatment benefit in favor of bracing. The rate of treatment success was 72% in the bracing group and 48% in the observation group. A similar positive effect was also found in the randomized analysis: 75% success after bracing compared with 42% after observation. The study also demonstrated a strong positive association between the average hours of brace wear and treatment success. Of the patients averaging between 12.9 and 17.6 hours of brace wear per day, 90% reached skeletal maturity with a Cobb angle of <50°; this percentage increased to 93% with 17.7 or more hours of wear. Hence, this study demonstrated for the first time, with Level I evidence, that bracing was effective in preventing curve progression to a surgical threshold in high-risk patients with AIS and that there was a dose-response effect (Table 2-1). The models of thoracic-lumbar-sacral orthosis (TLSO) used in the BrAIST trial varied according to the choice of individual center. Although many TLSO designs are used in routine practice (e.g., Boston, Rigo-Chêneau type), no specific type can currently be considered more appropriate for any given patient or group of patients.

Table 2-1. Probabilities of Curve Progression in Skeletally Immature Patients

| **Curve Magnitude** | **Age at Detection (years)** | | |
|---|---|---|---|
| **<19°** | **10–12** | **13–15** | **16** |
| **<19°** | 25% | 10% | 0% |
| **20–29°** | 60% | 40% | 10% |
| **30–39°** | 90% | 70% | 30% |

Reprinted from Nachemson et al. (1982) with permission.

## *Brace Prescription and Risk Calculator*

Trying to predict the ideal brace candidate has been a goal of treating physicians since the advent of modern bracing. In 1982, Nachemson, Lonstein, and Weinstein developed a "probability of progression" based on age as a surrogate for maturity, using patient data from Gothenburg (Sweden), Minneapolis, and Iowa City (table 2-1) (Nachemson et al. 1982). It was clear that most bracing studies included many patients who were unnecessarily braced because of their low probability of progression to a surgical threshold. The rigidly controlled BrAIST study, using traditional brace indications as outlined above, estimated the number needed to treat as 3, suggesting that (1) low-risk patients were unnecessarily braced, and (2) high-risk patients were undertreated in terms of hours of bracing per day. An important additional aim of BrAIST was to develop predictive models based on individual patient characteristics at initial presentation (e.g., sex, skeletal maturity, chronological age) and curve characteristics (e.g., curve magnitude, location) to establish baseline risk. Models including these variables plus treatment characteristics, such as hours of brace wear, could then be used to establish risk reduction associated with treatment.

An extensive systematic review published in 2015 identified no high-quality validated prognostic models to guide the treatment of AIS (Noshchenko et al. 2015). In 2019, we published the development and validation of a prognostic model in untreated AIS using the SSMS (Dolan et al. 2019). The development sample included 115 untreated BrAIST participants, and the external validation sample was combined from three institutions not involved in the study. The study provided a validated model and classification system to predict the risk of curve progression to surgical indications in untreated AIS patients using the SSMS. The model (https://uichildrens.org/ais-prognosis-calculator-simplified) was designed for easy use in all clinical settings where coronal spine views and hand films are obtained at the initial visit. The risk classification system produced reasonably accurate predictions in both the low- and high-risk groups.

There is now strong evidence supporting bracing as the standard of care for high-risk AIS patients to reduce the risk of the curve reaching a surgical threshold. The risk calculators provide clinicians and families with an additional tool to be used in a shared decision-making process (figures 2-1 and 2-2).


Figure 2-1. (A–C) A female patient, aged 12 years, 11 months, with a 24° low thoracic curve. She is menses 0, Risser 0, a standard traditional indication for bracing. Her digital skeletal age is four. Putting this information into the risk calculator shows only a 3% to 4% chance of the curve progressing to the surgical threshold. The family can make an informed choice as to whether they wish to brace or observe. The family chose observation, with no change in the curve at the final follow-up.

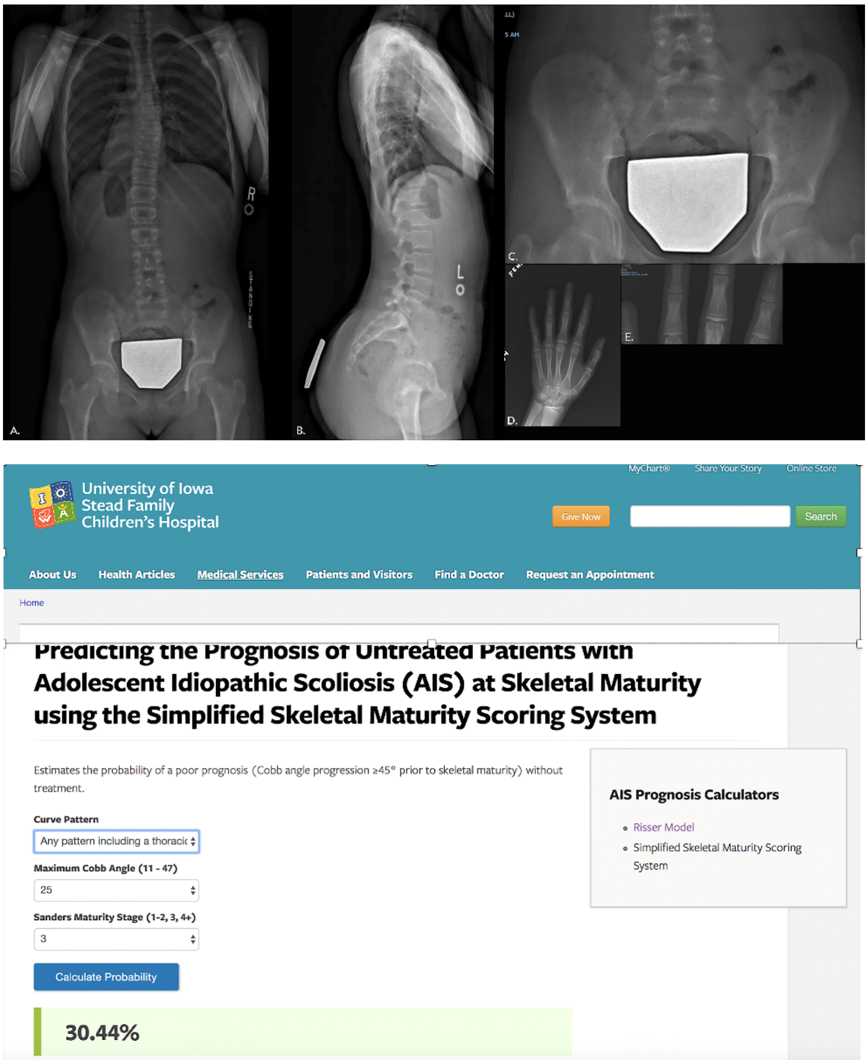


Figure 2-2. (A–E) A female patient, aged 12 years, 7 months, with a 25° right thoracic adolescent idiopathic scoliosis (AIS). She is menses 0, Risser 0, a traditional indication for bracing. Her digital skeletal age is three. The risk calculator shows her having a 30% risk of progressing to a surgical threshold without treatment. An informed shared decision can be made with the patient and family. As the family was risk averse, they chose bracing.

## Conclusions

Over their lifetime, untreated patients with AIS generally function well. As young adults they become employed, form relationships, have children, and become active older adults. With increasing age, most patients with untreated AIS can have back pain and feel they have important cosmetic concerns. Thoracic curves with a Cobb angle of >50° are associated with reduced vital capacity and more frequent shortness of breath compared with symptoms experienced by patients with smaller spinal curves. At-risk patients are those who have a high risk of curve progression to surgical indications (curve >45°-50°) if left untreated. This group includes patients with a curve between 20° and 40° who are skeletally immature. There is now Level I evidence supporting bracing as the standard of care for high-risk AIS patients to reduce the risk of the curve reaching a surgical threshold. Risk calculators provide clinicians and families with an additional tool to be used in a shared decision-making process in the nonsurgical management of AIS.

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