

Natural History of Adolescent Idiopathic Scoliosis in Skeletally Mature Patients: A Critical Review

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Abstract

The surgical treatment of adolescent idiopathic scoliosis is dependent on several factors, including curve type and magnitude, degree of curve progression, skeletal maturity, and other considerations, such as pain and cosmesis. The most common indication for surgery is curve progression. Most authors agree that surgical treatment should be considered in skeletally mature patients with curves $>50^\circ$ because of the risk of progression into adulthood. Furthermore, most authors would agree that curves measuring $<40^\circ$ to 45° in skeletally mature patients should be observed. When a skeletally mature patient with a curve measuring between 45° to 55° is presenting to an orthopaedic surgeon, it is not uncommon that the patient has no pain, no progression, and no imbalance. The generally accepted belief has been that curves that reach 50° are likely to progress into adulthood, progressing at a rate of 1° per year, based largely on the Iowa studies. However, the level of evidence for this is relatively weak, and the existing literature is equivocal in supporting the practice of performing surgery on these patients.

The treatment of adolescent idiopathic scoliosis (AIS) is dependent on several factors, including curve type and magnitude, curve progression, skeletal maturity, and other considerations, such as cosmetic deformity. The most common indication for surgery is curve progression. Skeletally immature patients are at a higher risk of progression, and curves that approach 50° in these patients are treated surgically. However, the decision to perform surgery on a skeletally mature patient with a curve in the range of 45° to 55° is not well-defined. Patients who have completed growth typically do not experience rapid curve progression, and they are often asymptomatic, without pain or imbalance. The task of the spine surgeon is to predict the effects of the deformity far into the future and

determine whether surgical treatment may be recommended now or sometime in the future if the curve becomes symptomatic or progresses.

Historical Perspective

The advent of Harrington instrumentation in the 1960s revolutionized the treatment of scoliosis. Prior to the Harrington era, surgery for AIS was not commonly performed, primarily because the deformity was more tolerable than the treatment, which often consisted of 6 to 12 months of casting. Before the 1960s, many scoliotic curves were not treated surgically, and in the ensuing decades (1960s through the 1980s) several studies were published on the natural history of these untreated

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curves. Our understanding of the natural history of untreated AIS is largely based on studies from this time period.

Surgical treatment of scoliosis became much more common after Harrington instrumentation was developed, and with further advances in spinal instrumentation in the 1980s and 1990s, the surgical correction of scoliosis became far less onerous for the patient and technically easier for the surgeon. As techniques evolved and curve correction became more feasible with a decrease in patient morbidity, many curves that were previously treated with observation were treated surgically.

Quality of Studies

The literature on the natural history of untreated AIS is fairly limited. The earliest studies revealed a grim prognosis,¹⁻³ mostly as a result of socioeconomic effects on work and marital status, the development of cor pulmonale, and increased disability secondary to back pain. Nachemson¹ reported on 130 patients with untreated scoliosis with 38-year follow-up; 30% claimed disability because of their deformity and there was a 100% increase in mortality compared with the normal population. However, only 45% of the patients had idiopathic scoliosis. Nilsson and Lundgren² reported on 113 patients, all with idiopathic scoliosis, with 50-year follow-up. Ninety percent of the patients had back pain and 30% were receiving disability benefits, with a mortality rate that was twice the rate of the general population. However, no radiographs were used to determine

the etiology of scoliosis, no Cobb angles were mentioned, and there was no differentiation between early-onset and adolescent curves. Similarly, Fowles et al³ reported on 55 patients with untreated scoliosis, but only 24 were idiopathic, and initial radiographs were not available to determine progression.

A major limitation of these earlier studies is the inclusion of patients with neuromuscular, congenital, and early-onset scoliosis. Therefore, their data cannot be extrapolated to an adolescent idiopathic population. Studies that focus more on AIS have shown a more benign natural history.⁴⁻¹⁰ However, such studies are few in number and are limited either by relatively short follow-up or a significant loss to follow-up that results in a small percentage of the original cohort being available for final analysis. A study that provides any meaningful information on the natural history of AIS must have a follow-up of several decades.

The Iowa Studies

For the last three decades, the Iowa studies have shaped our understanding of the natural history of AIS. In 1950, Ponseti and Friedman⁸ initially reported on 358 patients with untreated idiopathic scoliosis during their growing years, with a very short follow-up of 2 to 5 years. Four subsequent studies examined the natural history of these patients over five decades.^{5-7,9} As would be expected, many patients were lost to follow-up, some were determined not to have idiopathic curves, and others either refused to complete

questionnaires or did not have appropriate imaging studies. Therefore, the number of patients in each subsequent study gradually diminished. In the study by Weinstein and Ponseti,⁷ who published a 40-year follow-up on these patients, <30% (102 of 358) of the original cohort had completed follow-up. This study is somewhat limited by the small number of patients in each curve category, but it is still considered to be the best long-term follow-up study of untreated AIS as it relates to curve progression. The study by Weinstein et al⁹ is likely the final major report on the same cohort from 1932 to 1948. Of 314 eligible patients, 127 could not be located despite an extensive search, and after excluding patients who had died and those who had undergone fusion, 144 patients were located and eligible. Of these, 27 refused to participate, leaving 117 patients for this final study.

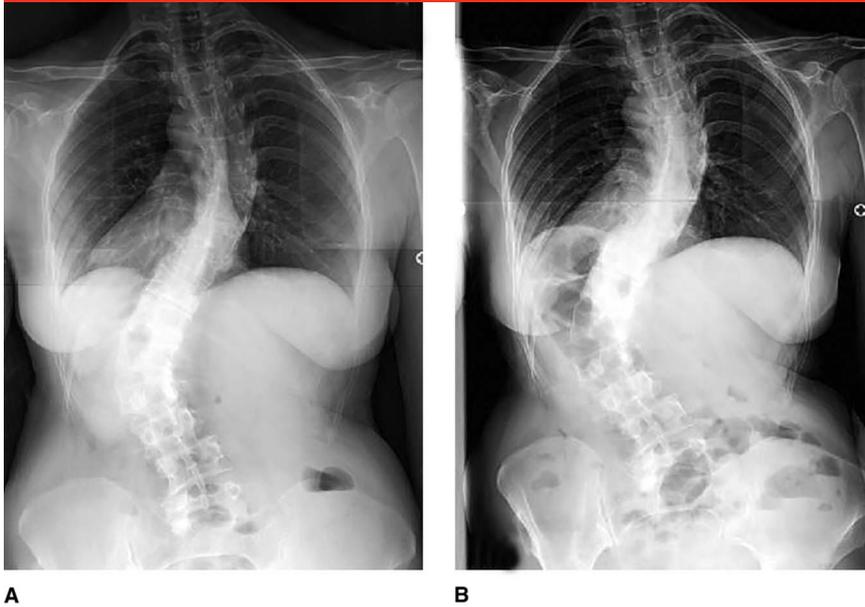
The Iowa studies, with their lengthy, detailed follow-up, have set the standard for natural history studies. Because surgical treatment of AIS has become more common, it is unlikely that any future studies of this magnitude on the natural history of untreated curves will be duplicated.

Curve Progression

The most compelling indication for surgery in AIS is curve progression. Predicting curve progression in skeletally mature patients can be challenging. With skeletal maturity, curve progression slows substantially compared with the progression that is observed during the adolescent growth spurt. Significant

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Figure 1



AP radiographs of a 53-year-old woman who has progression of her curve from (A) 53° to (B) 63° over a period of 12 years.

radiographic progression typically is noticed over a period of decades rather than months or years (Figure 1). A few long-term studies address curve progression in AIS, and these studies provide general guidelines on which clinical practice has been based for the last three decades.^{5-7,9,10}

When synthesizing the information available in these studies, most curves, especially those $>30^\circ$, progress at a rate of $<1^\circ$ per year.^{5-7,9,10} In general, as curve size increases, so does the likelihood of progression, possibly as a result of the influence of gravity on the concavity and the increasing imbalance that results from larger curves. However, several studies suggest that curves $>80^\circ$ to 90° either do not progress or do so at a much slower rate than smaller curves.^{5-7,9,10-12} Although the reasons for this observation are not clear, possible explanations include the spontaneous fusion that may occur at the apex of severe curves, the rigidity caused by degenerative changes, and

the “stability” that may be provided by the rib cage resting on the pelvis in these severe deformities.

Thoracic Curves

Several studies have shown that thoracic curves tend to be the most vulnerable to progression.^{6,7,9,10} In the Iowa study published in 1981,⁶ curves measuring between 50° and 80° in 1968 increased an average of 11.3° during the 10-year interval. Based on this information, it was recommended that thoracic curves reaching 50° at skeletal maturity should be fused. In the 1983 report,⁷ curves between 50° and 75° showed the most progression (ie, mean 29°), but it is important to note that there were only 11 patients with curves between 50° and 75° . Furthermore, closer analysis of the data reveals that only three patients had a curve in the 45° to 55° range, and all three progressed a mean of approximately 30° .

In a study by Ascani et al,¹¹ 17 thoracic curves between 40° and 49°

progressed a mean of $0.4^\circ/\text{yr}$. Eleven thoracic curves between 50° and 59° progressed a mean of $0.56^\circ/\text{yr}$. The study by Edgar¹⁰ included 28 patients with thoracic curves (ie, mean Cobb angle, 73° ; range, 36° to 118°) that progressed a mean of 8° over a 17-year follow-up ($0.47^\circ/\text{yr}$).

The recommendation to fuse curves that reach 50° at skeletal maturity is largely derived from the Iowa studies (from 1981 and 1983).^{6,7} The number of patients this recommendation was based on was small; 11 patients had curves between 50° and 75° , and 3 patients had curves between 45° and 55° .

Thoracolumbar Curves

In the Iowa series,⁷ only eight thoracolumbar curves measured $\geq 30^\circ$ at skeletal maturity (mean, 56°). At the 40-year follow-up, the mean curve size was 75.6° , for a mean progression of 19.6° , or $0.49^\circ/\text{yr}$. As in the thoracic group, thoracolumbar curves that measured between 50° and 75° at skeletal maturity progressed the most, increasing an average of 22.3° during the 40-year period. Each of these curves had developed a translatory shift at the lower end of the curve. However, there were only three thoracolumbar curves between 45° and 55° , two of which did not progress significantly, while one progressed approximately 40° (Figure 2).

In the study by Ascani et al,¹¹ which had a mean 34 years of follow-up, 14 patients had thoracolumbar curves $>40^\circ$, and the mean progression was 18.7° ($0.55^\circ/\text{yr}$). Of these, seven had curves between 50° and 59° and the mean progression for these curves was approximately 19° over 34 years. Fifteen patients had curves of $<40^\circ$, and the mean progression was 10° ($0.29^\circ/\text{yr}$). Edgar¹⁰ reported that 18 patients with a mean thoracolumbar curve of 54° had a mean progression of 10° over a 17.6-year follow-up; however, the curves were not separated based on the level of severity.

Thoracolumbar curves have the highest propensity for developing translatory shifts (ie, lateral listhesis) during adulthood, and back pain may be more prominent in more severe thoracolumbar curves. As degenerative changes develop and the curve becomes more rigid, surgical treatment may be more technically difficult in later life. Therefore, when thoracolumbar curves approach 50° to 60°, surgery should be considered.⁶

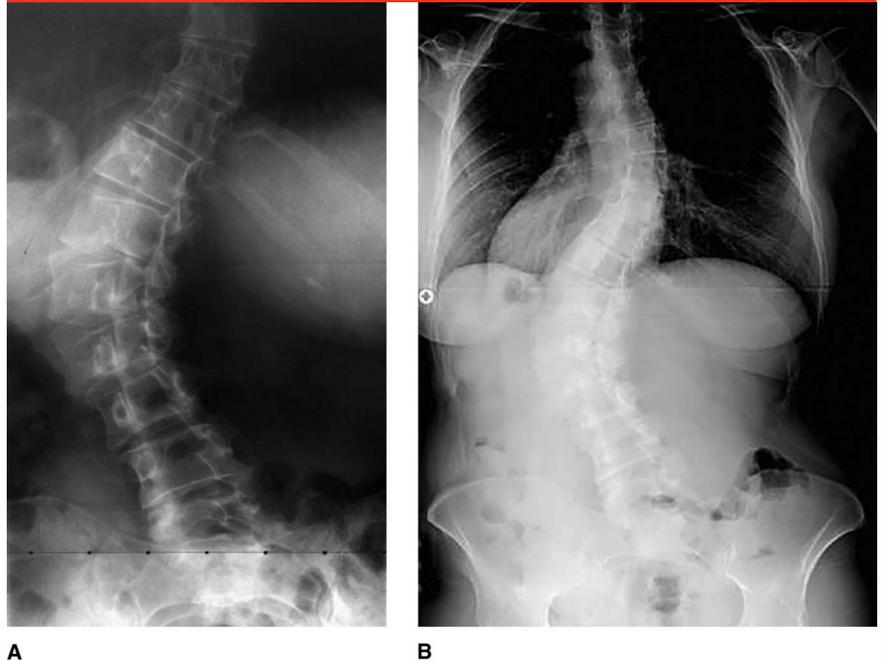
Lumbar Curves

In the Iowa series,⁷ there were 17 lumbar curves >30°, and the mean progression was 16.2° over 29 years. There were only five patients with lumbar curves >50°; two of the patients did not have significant progression and the other three progressed a mean of 20°. In 13 of 17 curves that were >30°, lateral listhesis developed at L3-4 and less commonly at L4-5. The study by Ascani et al¹¹ included 17 lumbar curves with a mean progression of 16° over 29 years, similar to the results by Weinstein and associates.^{6,7} The study by Edgar¹⁰ did not show any significant progression, with 11 lumbar curves progressing a mean of 3° over 19 years.

Double Curves

In the Iowa series,^{6,7} of all double curves, 11 had a thoracic component between 45° and 55° and progressed a mean of about 11°. Of double curves, only eight had a lumbar component between 45° and 55°, and the mean progression of these curves was about 20°. The study by Ascani et al¹¹ included 67 patients with double major curves. Of these, 25 had a thoracic component between 40° and 49° and progressed a mean of about 16° over 31 years. Twenty-five of the 67 double major curves also had a lumbar component between 40° and 59° and the mean progression was also similar to the thoracic curves.

Figure 2



AP radiographs of a 46-year-old woman with a thoracolumbar scoliosis that has not progressed over 5 years from (A) 2009 to (B) 2014.

Because there is considerable variation between the different studies, making any precise prediction for a particular curve in adult life is difficult. What is evident from the literature is that slow progression of curves does occur. As degenerative changes and osteoporosis develop in older age, an acceleration in curve deterioration may occur in some patients, although this may not necessarily be accompanied by clinical deterioration.

Most authors recommend surgery for the skeletally mature adolescent or young adult with a curve >55° to 60° because of the high risk of significant progression into adulthood. Furthermore, most authors would agree that curves of <40° to 45° in skeletally mature patients can be observed. Decision making is less well-defined in skeletally mature AIS patients with curves between 45° and 55°, and there is great variability in how these patients are treated. The best studies, with the longest

follow-up, have a small number of patients with curves in this range. Although it has become common for orthopaedic surgeons to recommend surgery when a curve reaches 50°, not all of these curves progress to a degree that leads to clinical sequelae in adult life. However, as patients age and other medical comorbidities develop, surgery becomes more technically challenging and surgical risks increase; therefore, surgery at a younger age may be justified in certain cases, even in the absence of documented progression. We feel that counseling the patient and family regarding the natural history data is important.

Cardiopulmonary Function and Mortality

Anatomists and clinicians have long postulated that severe spinal deformity leads to pulmonary hypertension with subsequent right ventricular

hypertrophy and cor pulmonale. Autopsies of scoliotic patients showed a high incidence of right ventricular hypertrophy and hypertensive pulmonary vascular changes.¹³ Proposed mechanisms responsible for pulmonary complications include reduced inspiratory muscle strength as a result of the abnormal shape and movement of the rib cage,¹⁴ limited range of movement and structural stiffness of the chest cage,¹⁵ and accelerated degenerative changes in scoliosis with accelerated stiffening of the rib cage with aging.¹⁶ The most common pulmonary function abnormality is a restrictive pattern of the lung volumes.

Several reports published in the mid to late 1900s correlated spirometric values with curve size.^{17,18} Several studies suggest limitations in cardiopulmonary function with curves $>60^\circ$.¹⁹⁻²² Both an airway obstructive component and a restrictive component have been implicated.¹⁹ It has been suggested that curves $>100^\circ$ may lead to cardiorespiratory failure²³ because of a significant reduction in lung volumes.¹⁴ An inverse relationship between curve size and vital capacity, percent predicted vital capacity, and total lung capacity has been observed.²⁴⁻²⁶ However, these studies had a heterogeneous group of deformities, with diagnoses other than idiopathic scoliosis, and no confirmatory data on the age of onset of deformity. Both the etiology and the age of onset play a critical role in the development of cardiorespiratory complications. The risk of cardiorespiratory complications is greater when scoliosis is apparent before the age of 5 years, with disabling or life-threatening respiratory complications occurring at or before late middle age in these patients.^{27,28} An increased mortality rate has been associated with early- and juvenile-onset idiopathic scoliosis but not with AIS.²⁹

Critical review of the literature does not support the hypothesis that idio-

pathic scoliosis that develops in adolescence leads to respiratory and ultimately cardiac failure in middle or old age. These patients have a good prognosis, and cardiorespiratory disability or disabling dyspnea is uncommon unless patients have independent cardiac or pulmonary disease.²⁷

However, these findings do not suggest that there are no cardiopulmonary consequences of moderate or severe scoliosis. Several studies have found reduced exercise tolerance, diffusion capacity, and lower VO_{2max} , even in adolescents with moderate to severe curves.^{20-22,30}

Kafer¹⁴ reported that lung volumes and the compliance of the total respiratory system and of the chest wall are inversely related to curve size. In the study by Collis and Ponseti,⁵ the authors reported that at least three patients in middle age with curves of $\geq 100^\circ$, who had no pulmonary symptoms, had reduced vital capacities ranging from 47% to 65% of normal. Although asymptomatic at that time, it is likely that these patients developed cardiopulmonary symptoms in older age that were at least partially explained by their spinal deformity.

In the study by Weinstein et al⁹ that completed the natural history studies stated earlier, scoliosis likely contributed to the death of three patients, all of whom had thoracic curves $>100^\circ$. It is unclear whether these are the same three patients discussed in the study by Collis and Ponseti.⁵ Twenty-two percent of patients reported shortness of breath during everyday activities compared with 15% of control subjects. Also, patients with thoracic curves $>80^\circ$ were more likely to experience shortness of breath than did patients with smaller thoracic curves.

The cardiopulmonary complications that are seen with early-onset scoliosis, such as significant loss of vital capacity, pulmonary hypertension, and cor pulmonale, are rarely

encountered in AIS. The larger the curve in the thoracic spine, the more likely that the patient will have gradual progression into adulthood, and with this progression will be a gradual reduction in vital capacity. An increased risk of death from cor pulmonale and right ventricular failure is rare and appears to occur only in patients with thoracic curves $>100^\circ$.⁸ Despite this finding, as patients age and other comorbidities develop, and as respiratory reserves and respiratory muscle strength diminish, this reduction in vital capacity may become clinically significant. This age-related impairment of respiratory function, when superimposed on a preexisting decrease in vital capacity, can lead to a variety of pulmonary consequences as patients age. If nonsurgical management is chosen for larger curves, it may be advisable to periodically monitor pulmonary function studies; if vital capacity drops to $<50\%$ of the value predicted, earlier treatment can be instituted. In general, AIS patients with thoracic curves generally do not face an increased mortality risk compared with people without scoliosis, and the recommendation for surgery for this indication is not supported in the literature.

Back Pain

Most adolescents with idiopathic scoliosis do not report significant pain. In counseling patients and families, a common question is whether untreated scoliosis causes back pain in adult life.

Adult scoliosis can be a late manifestation of untreated AIS; it may arise de novo as a result of asymmetric disk degeneration or facet degeneration, most commonly in the thoracolumbar or lumbar spine, or it may be secondary to other conditions, such as congenital deformity, limb-length discrepancy, or osteoporosis with

asymmetric fracture collapse. In the absence of long-term follow-up, the distinction between these three entities, particularly the first two, can be difficult. When an adult presents with a painful lumbar or thoracolumbar scoliosis, it is difficult to determine with certainty whether the curve is a de novo degenerative curve or an idiopathic curve that has undergone degenerative changes.

Adults with scoliosis often present with back pain and disability.^{31,32} The pain may result from spinal imbalance (ie, coronal or sagittal imbalance), facet arthropathy, muscle fatigue, or central or foraminal stenosis. Asymmetric loading of the disks and facet joints leads to incompetence of a spinal segment or segments and subsequent segmental instability, either in the sagittal plane (ie, spondylolisthesis) or more often in the coronal plane in the form of translation (ie, lateral listhesis) or three-dimensional rotatory subluxation. Osteophytes form at the vertebral end plates and facet joints, and hypertrophy of the ligamentum flavum and joint capsules leads to central and/or lateral recess stenosis and subsequent radiculopathy. On the concavity, foraminal stenosis is common and may also contribute to radicular pain. Increasing curve size has been associated with more severe pain, as has the degree of degenerative changes at the curve apex.³²

Thoracolumbar and lumbar curves may be more likely to cause back pain in adulthood.^{6,8,10} Cordover et al³³ evaluated 34 patients with thoracolumbar or lumbar curves measuring between 20° and 55° at skeletal maturity; at an average 22-year follow-up, 65% had back pain compared with 32% of age-matched control subjects. Although these patients were doing well and none of them underwent surgery for back pain, the mean age at final follow-up was only 36 years. Over a lifetime, as the curve gradually

increases in size and further degeneration occurs, symptomatic lateral listhesis or rotatory subluxation is not an uncommon occurrence in the mid to upper lumbar spine.

In the latest Iowa study⁹ with 50-year follow-up, it was reported that, on average, patients with scoliosis report more chronic back pain. However, those with pain have profiles similar to those of their peers in terms of duration and intensity and the ability to work and perform daily activities. Despite this generally benign long-term prognosis with regard to pain, many elderly patients with scoliosis and degenerative changes do present with back pain and/or radicular symptoms. As stated earlier, it is often difficult to determine whether these patients had scoliosis as adolescents or have developed de novo degenerative curves with attendant problems. As life expectancy increases, with expansion of the elderly population, adult scoliosis may take on increasing importance. But in counseling the asymptomatic adolescent or young adult with a large curve that has not shown progression, the possibility of back pain developing in the future should not, in and of itself, be an indication for surgical treatment. Table 1 summarizes the available literature on the natural history of idiopathic scoliosis.

Psychosocial Issues and Appearance

Studies that have addressed appearance and the psychosocial aspects of scoliosis are conflicting. Early studies reported that many patients were receiving disability benefits, and a large proportion of women with scoliosis were unmarried.^{1,2} The limitations of these studies have been discussed. Other studies suggest that patients with scoliosis perceive themselves to be less healthy than

their peers and experience restrictions in physical and social activities, such as lifting, walking long distances, and standing and sitting for long periods.^{34,35} Ascani et al¹¹ reported “real psychological disturbances” in 19% of their patients, 94% of whom had curves >40°.

In contrast, patients from Iowa⁹ were found to be gainfully employed except for four patients who were mentally handicapped; 90% had married and <20% had psychological effects from their deformity. Other studies note no significant difference between people with AIS and control subjects in their quality of life or the ability to undertake activities.³⁶

The clinical deformity caused by scoliosis is quite variable. Some patients with severe curves have little apparent deformity secondary to body habitus or musculature, whereas others with milder curves have more clinically apparent deformities. Right thoracic curves, particularly in the presence of a prominent rib hump, tend to be more deforming. The patient’s psychological makeup also influences the degree of dissatisfaction with appearance. Some patients with severe curves accept their deformity; others with mild curves have significant cosmetic concerns.

Compared with adolescents, cosmetic concerns are rarely the presenting symptom in adults with scoliosis, particularly in patients older than 40 years. These patients present with back or radicular leg pain. Occasionally, patients younger than 40 years with a thoracolumbar or lumbar curve may have cosmetic concerns. However, most patients are more self-conscious about their deformity in their adolescent and young adult years than in older age.^{5,6}

Patients with AIS who have more severe curves are less satisfied with their body image and appearance in swimsuits or tight clothing than are control subjects. It is common for these patients to have some degree of self-consciousness about their

Table 1

Natural History Studies for Adolescent Idiopathic Scoliosis

Study	No. of Patients (f/u period)	Back Pain/Disability	Curve Increase	Lung Function and Mortality	Comments
Collis and Ponseti ⁵	215 (20 to 36 yr)	No correlation between back pain and type/severity of curve. No relationship between severity of curve and extent of degenerative changes. Unemployment rate of <3%.	58 pts: Risser, 4; mean curve increase, 14.5° 61 pts: Risser, 5, mean curve increase, 10°	66% of curves >60° had diminished vital capacity. For thoracic curves, as curve size increased, vital capacity decreased. 7% of pts had died. Cor pulmonale or scoliosis not cited as cause of death in any patient.	Many pts lost to f/u; only 245 of original 348 pts were found. Of these, 215 formed basis of this study. Radiographs available in only 134 pts. Quality of f/u: 106 had history and physical and vital capacity measurements, 134 had radiographs, 89 returned questionnaires. Most patients (who were then middle-aged) were "leading active, productive lives" despite curves >50° in 71% of pts. Two patients committed suicide (thoracic curves 70° and 168°).
Scott and Piggott ⁴	30 (11 yr; range, 7 to 16 yr)	Not discussed	60% of curves progressed, always <10°. Curves >30° with rotation >25° were almost twice as likely to progress.	Not discussed	Only mild curves were included in this study (all curves <53°). Small sample size. 10 of 30 pts had curves <20° at skeletal maturity. Note: All pts had initial and follow-up radiographs.
Weinstein et al ⁶	194 (39.3yr; range, 31 to 51 yr)	Back pain more likely in thoracolumbar curves	Thoracic curves between 50° and 80° increased average of 11.3° during 10-yr period. Thoracolumbar curves increased by 3.3°.	29% had dyspnea. 2.5% had severe dyspnea (walking short distances). 83% with thoracic curves >60° had vital capacity of <75%. Pulmonary symptoms and diminished vital capacity correlated well with severity of thoracic curves. 15% mortality was no different from general population. One pt died of cor pulmonale attributed to scoliosis.	Located 206 of the 245 pts with recent information available for 194 pts. Current radiographs available for 120 pts, which represents 49% of 245 pts located by Collis and Ponseti ⁵ in 1969, and only 33% of the original 358 pts whose deformity began after age 8 yr. 21% not willing to wear tight clothes or a bathing suit; most pts were more self-conscious about deformity in teenage/young adult years than in older age.
Bjerkreim and Hassan ¹²	70 (8.5 yr)	Not discussed	Most curves increased 3°/yr before age 20 yr and 1°/yr after age 20 yr. Curves <40° increased significantly less than larger curves and curves from 60° to 80° increased the most. Single curves increased significantly more than double curves; thoracic curves increased significantly more than lumbar curves.	Not discussed	Mean follow-up of only 8.5 yr

(continued)

AIS = adolescent idiopathic scoliosis

Table 1 (continued)

Natural History Studies for Adolescent Idiopathic Scoliosis

Study	No. of Patients (f/u period)	Back Pain/Disability	Curve Increase	Lung Function and Mortality	Comments
Weinstein and Ponseti ⁷	102 (40.5 yr; range, 31 to 53 yr)	Not discussed	Curves <30° did not progress except lumbar curves in which L5 was not well-seated and apical rotation was >33%. Thoracic curves 50° to 75° increased the most (29°). Lumbar curves >30° increased by a mean of 16°. Curve progression related to degree of vertebral rotation.	Not discussed	Only 11 pts with thoracic curves between 50° and 75°. Only 17 lumbar curves >30°. Many patients lost to follow-up from original study.
Ascani et al ¹¹	187 (33 yr; range, 15 to 47 yr)	61% incidence of back pain. Thoracolumbar curves more often associated with back pain (79%).	All curves progressed after skeletal maturity (0.4°/yr). 17 thoracic curves between 40° and 49° progressed 0.4°/yr. 11 thoracic curves between 50° and 59° progressed 0.56°/yr.	Cardiopulmonary symptoms (not defined) present in 22%; more common with thoracic and thoracolumbar curves. Mortality rate 17% in first 50 yr, twice as high as mortality rate in general Italian population.	234 of 400 pts were found. 187 had initial radiographs. Only 47% of the original 400 pts were studied. "Real psychological disturbances" in 19%, most of whom had curves >40°.
Edgar ¹⁰	78 (17 yr; range, 10 to 27 yr)	Back pain in 79%, with 44% having frequent pain. Only definite factor correlating with extent of pain was presence of degenerative changes.	Significant deterioration occurred with curves >55° (>0.5°/yr). Curves >100° in thoracic region and >90° in thoracolumbar and lumbar regions do not progress as much. Lumbar curves are least predictable; increase in rotation out of proportion to the Cobb angle. Deterioration and lateral subluxation at L3-4 common.	Not discussed	5 of 78 pts had infantile scoliosis. Of 100 pts seen between 1950–1964, 78 had adequate radiographs and were skeletally mature at initial presentation. Original number of pts unknown; selected the first 100 pts who could be traced. Over 80% felt fulfilled in life (job satisfaction, marriage, acceptance of curve), with some self-consciousness but without significant depression.
Cordova et al ³³	34 (only thoracolumbar and lumbar curves studied; mean, 22 yr)	65% reported back pain compared with 32% of control subjects. Only 3 of 34 pts reported severe back pain. Only pts with thoracolumbar and lumbar curves were studied.	Follow-up radiographs not obtained. No data on progression reported.	Not discussed	Small sample of 34 pts; no follow-up radiographs. 29% of pts were bothered enough by their deformity to not wear a bathing suit.
Weinstein et al ⁹	117 (51 yr; range, 44 to 61 yr)	61% of pts had chronic pain (most mild or moderate) compared with 35% of control subjects. No evidence for a relationship between degree of back pain and osteoarthritis.	Not discussed	22% reported shortness of breath. More likely to be short of breath with thoracic curves >80°. No statistically significant difference in mortality compared with general population. However, 3 deaths potentially due to scoliosis (>100° thoracic curve).	Longest follow-up study ever published on natural history of AIS. Pts were slightly dissatisfied to slightly satisfied on body image (control subjects were slightly satisfied to moderately satisfied). No significant difference in rate of depression between pts and control subjects.

AIS = adolescent idiopathic scoliosis

deformity. However, during counseling, patients and their families should be made aware that cosmetic concerns tend to fade and become less prominent as patients age. Furthermore, patients are sometimes concerned about the appearance of the chest, and surgical correction of the curve, while successful at reducing the rib hump, is not as effective at restoring balance to the anterior chest cage. Whether cosmesis, in and of itself, should be an indication for surgery is debatable, and this decision is best left to the well-informed patient and family. However, when patients are presented with this evidence, most tend not to pursue surgery for this indication.

Summary

Natural history studies reveal a favorable natural history for AIS. With the exception of some increase in back pain and cosmetic concerns, patients tend to function fairly well into adulthood despite severe deformities. The mortality rate is not increased compared with that of normal control subjects. Although thoracic curves $>100^\circ$ may predispose patients to cardiopulmonary complications, progression to this degree is unusual. Curves $>50^\circ$ at skeletal maturity may be treated surgically to prevent severe deformity in adulthood, but this recommendation is based on a small number of patients in long-term follow-up studies. Each patient ultimately determines his or her own natural history, and long-term radiographic follow-up is important if nonsurgical management is chosen. Patients and families should be counseled on the generally benign natural history of AIS before surgery is recommended.

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Evidence-based Medicine: Levels of evidence are described in the table of

contents. In this article, references 1-36 are level IV studies.

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