

The Etiology and Natural History of Idiopathic Scoliosis

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Idiopathic scoliosis (IS) can be defined as a lateral curvature of the spine, greater than 10° as measured by the Cobb method,¹ in the absence of any relevant congenital spinal anomalies or associated musculoskeletal conditions. The true etiology of IS remains unknown, but its cause is thought to be multifactorial. The intent of this article is primarily threefold: 1) To summarize the etiological factors thought to be associated with IS, consistent with an outline recently published by the Scoliosis Research Society's (SRS) Etiology Committee²; 2) To summarize what is currently understood about the natural history of IS, identifying various "risk factors" for possible curve progression, and 3) To identify the general thinking about when orthotic management is indicated, and summarize the evidence of its ability to alter the natural history of the disease.

GENETIC FACTORS

The role of genetic or hereditary factors in the etiology of IS is widely accepted. Harrington³ studied women with a scoliotic curve that exceeded 15° and found a 27 percent prevalence of scoliosis among their daughters. The mode of inheritance is debated, but studies collectively characterize IS as a single-gene disorder. However, a so-called simple gene disorder is susceptible to variability in penetrance, meaning a certain percentage of individuals carrying the gene will not express the observable trait. It is extremely difficult to isolate family or small sample populations to yield a positive result for genetic linkage.

PRIMARY MESSAGE

Genetics plays a role in the etiology of IS as a single-gene disorder, but because of the variability of penetrance and the heterogeneity of the population, it is extremely difficult to isolate this as a singular etiological factor.

THE ROLE OF MELATONIN

Melatonin is a hormone made by the pineal gland in the brain that helps regulate sleep. Doubousset and Machida⁴ reported patients with progressive scoliosis had a 35 percent decrease in melatonin levels throughout the night compared with those with stable scoliosis or control subjects. However, vari-

ation in melatonin levels found in other diseases has not demonstrated an effect on the development of scoliosis, and patients with IS do not have documented difficulties with sleep or immune function.

PRIMARY MESSAGE

Melatonin may play a secondary role in the development of IS, but it is unlikely that scoliosis results from a simple absence of melatonin.

EFFECTS OF CONNECTIVE TISSUE

Collagen and elastic fibers are principle elements in the supporting structures of the spine. Changes in the distribution of collagen in patients with IS differ from those of seen in subjects without IS, but these changes are not consistent among those with IS. Echenne et al.⁵ reported that skin in patients with IS has significant differences within the middle and deep dermis compared with that of subjects without IS. Elastic fiber abnormalities in the spinal ligaments have also been reported in a substantial number of patients with IS compared with those of individuals without IS.⁶

PRIMARY MESSAGE

Most researchers concede that abnormalities within the connective tissue elements of the majority of those with IS probably are secondary to the structural forces of the scoliotic deformity itself.

SKELETAL MUSCLE ABNORMALITIES

The differences between type I (slow-twitch) and type II (fast-twitch) muscle fibers in those with IS have been studied. Decreased type II fibers in the paraspinous and gluteus medius muscles have been reported.^{7,8} One report showed a normal distribution of type I and type II fibers on the convexity of the curve, but a lower frequency of type I fibers on the concavity.⁹ However, another report showed a decrease in the number and size of type II fibers, with no preference for the convex or concave side.¹⁰ Another study found similar results from distant muscle sites (deltoid, trapezius, gluteus, and quadriceps), so the authors suggested a myopathic process¹¹ may play a significant role in the etiology of IS.

PRIMARY MESSAGE

There are no definitive conclusions. Most abnormalities are likely secondary to the deformity itself, but histochemical changes may indicate a defect of the cell membranes.

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THROMBOCYTE ABNORMALITIES

Thrombocytes are the same as platelets, which are described as nonhemoglobin carrying, irregularly shaped discs found in the bloodstream. Many investigators have noted abnormalities in the structure and the function of thrombocytes in patients with IS. Those with larger scoliotic curves have a higher concentration of a more dense type of platelet, compared with those with small curves or control subjects.¹² Calmodulin is a calcium-binding receptor protein, and it regulates the contractile properties of muscles and platelets. Increased calmodulin levels in platelets have been shown to be associated with the worsening of adolescent IS.¹³ These data are particularly important when compared with the reports on melatonin, in which decreased levels of melatonin were found in those with adolescent IS and curve progression greater than 10°. Melatonin binds to calmodulin and has been shown to act as a calmodulin antagonist.

PRIMARY MESSAGE

Changes in platelet morphology and physiology suggest a cell-membrane defect in patients with IS.

NEUROLOGICAL MECHANISMS

Numerous, sophisticated neurological investigations have been conducted during the past 20 years. Although studied extensively, peripheral proprioception has not yielded consistent results. Similarly, abnormal "sway" patterns of the trunk were initially reported, but these patterns are thought to be secondary to the curve itself. Any such patterns ultimately resolve at the end of growth. Abnormalities in the cerebral cortex, cervicothoracic syrinx associated with Chiari type-I malformations, and such have been studied.

PRIMARY MESSAGE

Lowe summarizes these investigations nicely²: "Any hypothesis that proposes a neurological defect must account for the impression that many patients with IS have above-average ability in sports. These observations have been largely anecdotal, but a study of girls attending ballet school showed an increased prevalence of IS as high as 20%. It's difficult, therefore, to account for a neurological defect that allows the patient to excel in activities demanding high proprioception and coordination."

THE ROLE OF GROWTH AND DEVELOPMENT

There is an association between hypokyphosis and IS. A prospective review of Finnish children showed that those with scoliosis were taller and had less kyphosis compared with children without scoliosis.¹⁴ An imbalance of growth that appears to exist between the anterior and posterior structures of the spine has been hypothesized as a contributing factor to the etiology of IS. This hypothesis argues that IS is essentially the opposite of Scheuermann's kyphosis. It

suggests the anterior structures grow more rapidly than the posterior ones, in effect forcing the spine to rotate to the side upon forward bending. In an article suggesting the pathogenesis of IS, Dickson et al.¹⁵ stated: "Idiopathic *kyphoscoliosis* cannot and does not exist, from the mildest cases in the community to the most severe case in pathology museums." In that review of 70 patients with thoracic curves, the average Cobb angle was 39° with an average kyphosis of 20°, as measured by a simple standing lateral radiograph. However, by using a technique with which a "true" lateral of the apical vertebrae of the thoracic curve could be viewed by taking into account the vertebral rotation associated with the curve, Dickson et al. reported this portion of the spine to be lordotic an average of 3°. They noted the apical one or two vertebrae were distinctly wedged, with the anterior height greater than the posterior height. Thus, Dickson et al. argued the coronal plane curvature as seen in IS is entirely secondary to the sagittal plane imbalance of growth.

The period of accelerated growth in puberty starts about 1 year earlier in girls with scoliosis, but there are no differences in maximum growth rates.^{16,17} The overall height, and the height-to-width ratio of the sixth thoracic vertebrae were notably greater in patients with IS. Girls have a greater height of the vertebral bodies than do boys, and this difference increases with age.

PRIMARY MESSAGE

In general, girls with IS have a tendency to be taller and more slender than their peers. There are also indications that the scoliotic spine is more slender and longer than the nonscoliotic spine. This condition is thought to predispose the spine to "column-buckling," as described below.

BIOMECHANICAL FACTORS

The manner in which a structure is supported (the boundary conditions) is an important determinant of its mechanical behavior. For instance, pelvic obliquity or motor imbalance can increase the likelihood of causing scoliosis in neuromuscular or myopathic disorders. For this reason, some have investigated the potential role of bone quality in those with IS. Authors have reached different conclusions on mechanical property variances of bone found in patients with IS, but there is no firm evidence that this may be an important factor in the etiology of the disease.

Soft-tissue extensibility and joint laxity may be an important risk factor for the progression of IS, but like bone quality, there is little evidence as an etiological factor. Others have considered the asymmetrical loading of the spine that forces the collapse of the overloaded side.¹⁸ An example of this is known as the "thoracospinal concept." This theory is based on the finding that in convex right thoracic scoliosis, the ribs on the left are consistently longer than those on the right.¹⁹ These findings are supported by those of similar studies suggesting the left side of the thorax is more vascular than the right and thus likely to grow more. The hypoky-

photic spine may be more sensitive to axial loads, and thus have greater risk for buckling. The direction of the collapse depends on local conditions but is often determined by the lateral pressure exerted by the aorta. A “rotational lordosis” can result from the growth of the posterior vertebral elements lagging behind that of the anterior bodies.²⁰ The amount of growth difference (anterior to posterior) need not be great, but the greater the difference, the higher the risk of collapse. Aortic loading has been reported to cause mild asymmetry of the thorax, resulting in a mild right-sided rib hump upon forward bending in the otherwise healthy child. It is hypothesized that this particular predisposition to right-sided asymmetry can account for the more common convex-right thoracic curve pattern found in IS.

A growth imbalance between the anterior and posterior portions of the vertebrae can be exacerbated by the Heuter-Volkman principle. This principle can be described simply as decreased concave growth. Thus, this principle can serve only as a “vicious circle” of asymmetric loading of the spine coupled with true plastic deformation.

PRIMARY MESSAGE

The thoracic vertebrae in IS are known to demonstrate a decreased posterior element growth and increased anterior body wedging, resulting in hypokyphosis or even lordosis (in the sagittal plane), predisposing the spine to scoliosis (in the coronal and transverse planes). Thus, in the absence of external (orthotic) support, such a spine is more susceptible to collapse into a scoliotic posture.

THE ROLE OF SKELETAL MATURITY

There are four forms of IS, defined by the age of onset or discovery:

- Infantile onset: scoliosis that develops before 3 years of age.
- Juvenile onset: scoliosis developing between the ages of 3 and 10 years.
- Adolescent onset: scoliosis that appears (generally) after age 10 years, but before the onset of puberty and before skeletal maturity.
- Adult onset: Scoliosis that appears after skeletal maturity.

Infantile onset is unique in that it is the only form of IS known to spontaneously resolve. That is, it can improve or disappear without treatment (beyond observation). As early as 1930, Harrenstein²¹ reported, “Spontaneous correction does occur without treatment but at the moment it is not possible to distinguish between the two at the time of diagnosis.” In 1972, Mehta²² described use of the rib-vertebra angle in the early diagnosis between resolving and progressive infantile scoliosis (see Figure 1 for how this angle is constructed). The primary finding of that study was that in patients in whom the initial rib-vertebra-angle-difference (RVAD) was less than 20°, approximately 80 percent experienced spontaneous resolution of their scoliosis, whereas in those with an RVAD

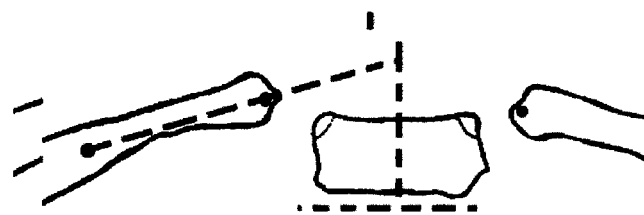


Figure 1. The construction of the rib-vertebra angle.²² This is drawn referencing the apical vertebrae in a thoracic scoliosis, where the RVAD value is the difference in the size of the angles between the convex and concave sides of the curve. (Reprinted from Mehta MH. The rib-vertebra angle in the early diagnosis between resolving and progressive infantile scoliosis. *J Bone Joint Surg [Br]* 1972;54:230–243. Used with permission.)

greater than 20°, 80 percent experienced progression of their scoliosis.

Numerous articles have reported on factors that influence the likelihood for curve progression in juvenile and adolescent onset IS. The following are the primary factors reported in the literature:

- Gender
- The degree of maturity at the time of curve discovery (eg, chronological age, skeletal age as described by the Risser sign, and the timing of the primary adolescent growth spurt)
- The degree of curvature at the time of discovery
- The location and pattern of the curve

GENDER

In general, the incidence of IS is similar between boys and girls in smaller curves. However, girls are more likely to experience curve progression (Table 1).

MATURITY

In their landmark article on the prediction of curve progression in untreated IS, Lonstein and Carlson²⁴ described the relationship between maturity and curve progression. In that review of 727 school-age children (575 girls, 152 boys), with initial curves of 5° to 29°, the authors described three primary prognostic factors in predicting curve progression in the growing child: curve size, chronological age, and Risser sign. They reported the younger the child, the lower the

Table 1. Prevalence of gender for idiopathic scoliosis according to the severity of the curve in 1,222 subjects*

Curve (°)	Girls	Boys	Girls: Boys
6–10	316	322	1:1
11–20	299	208	1.4:1
21 or more	65	12	5.4:1
Under treatment	36	5	7.2:1

*Data reflect those reported in Rogala et al.²³

Risser sign, and the larger the curve at discovery, the higher the likelihood for curve progression (Table 2).

Based on the data from patients with curves ranging from 20° to 29°, a nomogram devised from a simple formula to calculate a progression factor was developed (Figure 2):

Progression factor

$$= [(Cobb\ angle - 3) \times Risser\ sign] \div\ chronological\ age$$

The authors stressed this formula could be used primarily as a tool to develop a sense of risk for progression in communicating with a concerned patient and family, but not as a tool for clinical decision making. This is because the formula does not take into account other important factors, such as the location and pattern of the curve, the patient's gender, or in girls, menarchal status.

As for the long-term prognosis of scoliosis, Weinstein and Ponseti²⁵ reported on a series of 102 untreated scoliosis patients representing 133 curves, with radiographs having an average follow-up of 40.5 years. Sixty-eight percent of the curves progressed after skeletal maturity. Curves that were less than 30° at skeletal maturity tended not to progress, regardless of curve pattern. Progression of curves greater than 30° appeared to be related to the amount of vertebral rotation. Curves that measured between 50° and 75° at skeletal maturity, particularly thoracic curves, progressed the most.

However, in a more recent publication, Weinstein et al.²⁶ studied outcomes related to health and general function in untreated patients with adolescent IS. Citing previous long-term studies as having a more heterogeneous population of patients with true IS and those with non-IS, Weinstein et al. argued late-onset IS (LIS) is a distinct entity with a natural history that differs from other forms of "idiopathic" scoliosis. The authors defined LIS as "a structural lateral curvature of the spine arising in otherwise normal children usually during puberty," and compared 117 prospectively evaluated, untreated patients with 62 age- and gender-matched volunteers. This review, which had a 50-year follow-up, found that those with LIS were significantly more likely to experience shortness of breath in the presence of a thoracic curve >80° than were control subjects. Patients with LIS also were significantly more likely to report chronic back pain than were

Table 2. Progression incidence relating to the magnitude of the curve and the Risser sign*

Risser Sign	Percentage of Curves that Progressed	
	5-19° Curves	20-29° Curves
Grade 0 or 1	22	68
Grade 2, 3, or 4	1.6	23

*Data reflect those reported in Lonstein and Carlson.²⁴

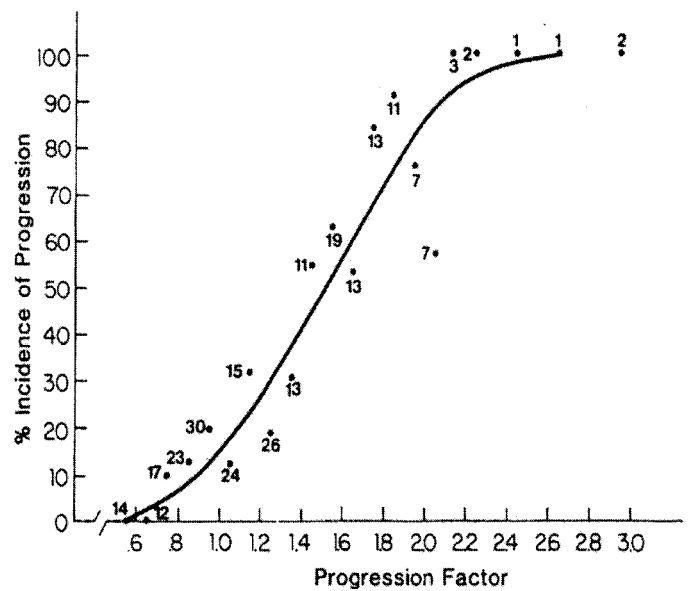


Figure 2. Percent incidence of progression according to the progression factor. (Reprinted from Lonstein JE, Carlson JM. The prediction of curve progression in untreated IS during growth. *J Bone Joint Surg [Am]* 1978;60:173-176. Used with permission.)

control subjects. However, there was no significant difference in the estimated probability of survival between the two groups. Thus, the authors concluded that adults with untreated LIS experienced little physical impairment other than back pain and cosmetic concerns compared with age- and gender-matched control subjects.

THE RELATIONSHIP BETWEEN PEAK GROWTH VELOCITY AND CURVE PROGRESSION

Little et al.²⁷ reported on the relationship of growth rates, as expressed in overall height velocities, in girls with IS. Peak height velocity was defined as a growth rate of at least nine centimeters per year, as calculated by standing height measurements at 6-month intervals for a minimum of 2.5 years. The timing of peak height velocity, also known as the peak growth age (PGA) was plotted against other maturity indicators, such as chronological age, Risser sign, and menarchal status, to determine its usefulness as a prognostic tool for curve progression.

Of special interest to the orthotist, 88 of 120 girls reviewed demonstrated progression of their primary curve despite being braced (Boston and Charleston braces were used). Sixty of these patients had a curve of more than 30° at PGA, and in 50 (83 percent) of the 60 patients, the curve progressed to 45° or more. The remaining 28 patients had a curve of 30° or less at PGA, with only one curve (4 percent) progressing to 45° or more. Thus, there appeared to be a significant relationship between the size of a primary curve size, with 30° as an important threshold, and the timing of PGA.

Citing the aim of all maturity scales as being an aid for the clinician in decision making regarding whether to treat or observe a patient with scoliosis, Little et al. reported PGA was

found to have a stronger correlation with curve progression than any other maturity indicator (age, Risser sign, or menarchal status). Little et al.²⁷ state: "Peak height velocity also grouped patients for maximal progression of the curve more accurately than did the other maturity scales, as most of the curve progressed maximally at peak height velocity. There was a wider spread of timing of maximal progression when chronological age, menarchal age, and Risser sign were used to predict progression." Of particular interest from a clinical standpoint is the fact that the median age at which PGA was achieved was less than the median age at menarche and Risser 1 (Figure 3).

THE SIZE OF THE CURVE

In general, the larger the curve at presentation for all children and adolescents, the higher the likelihood for curve progression (Figure 4).

THE LOCATION AND PATTERN OF THE CURVE

There tends to be agreement in the literature that single lumbar and single thoracolumbar curves are the least likely to progress and are the most receptive to brace treatment. According to some natural history studies,²⁸ single thoracic and double patterns are more likely to progress, by as much as a factor of two, than are single lumbar or single thoracolumbar curve patterns (Table 3).

CAN AN ORTHOSIS ALTER THE NATURAL HISTORY OF IDIOPATHIC SCOLIOSIS?

Numerous manuscripts on the efficacy of bracing scoliosis have been published during the past half-century. However, most of the natural history studies were not published until the 1980s. These reports exposed the fact that a number of earlier reports on the efficacy of orthotic treatment, at no fault to the original authors, included patients who were

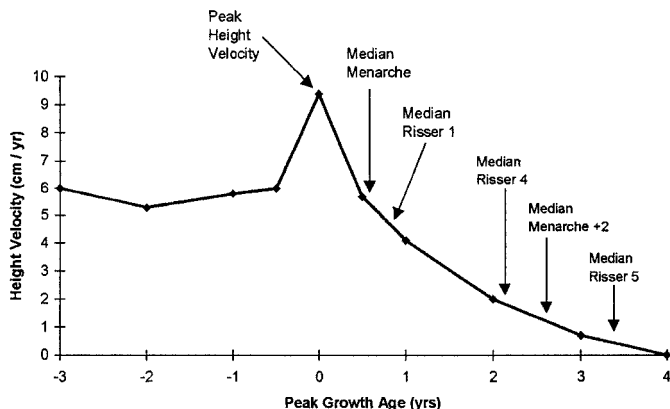
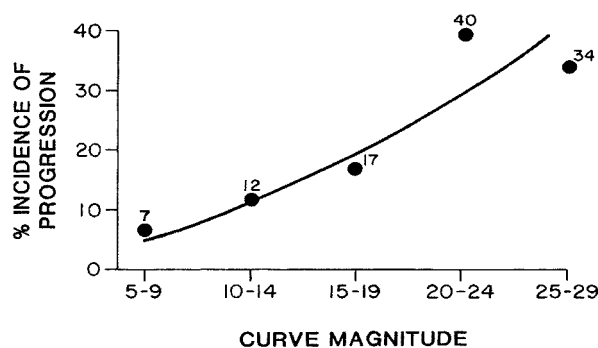


Figure 3. Plot of height velocity curve with median occurrence of menarche, age at menarche plus 2 years, and Risser grades 1, 4, and 5 in the 120 female patients with IS. (Reprinted from Little DG, Song KM, Katz DE, et al. Relationship of peak height velocity to other maturity indicators in IS in girls. *J Bone Joint Surg [Am]* 2000;82:685–693. Used with permission.)



This graph shows the probability of curve progression as a function of curve magnitude. This graph combines all ages that have any given curve magnitude.

Figure 4. Probability of curve progression as a function of curve magnitude. Data reflect those reported in Lonstein and Carlson.²⁴ (Reprinted from Bunch WH, Patwardhan AG, eds. *Scoliosis: Making Clinical Decisions*. St. Louis: CV Mosby Co.; 1989. Used with permission.)

Table 3. Percentage of curves that progressed*

	Double	T/L	Thoracic	Lumbar
Study 1	67%	47%	42%	12%
Study 2	75%	69%	54%	25%
Study 3	27%	27%	44%	18%
Study 4	27%	12%	26%	9%

*Data reflect those of studies cited in Bunch and Patwardhan.²⁸

unlikely to experience curve progression if not treated (Risser 3–5, or curves less than 20°). Thus, many of the reported "success rates" on the ability of an orthosis to prevent curve progression immediately became suspect.

Reports that followed the publication of natural history reports are also difficult to compare because many used differing definitions of treatment success, with equally varying criteria for inclusion. Studies by the SRS attempted to address these concerns in the mid-1990s. The SRS Natural History Committee published "A Meta-Analysis of the Efficacy of Nonoperative Treatments for Idiopathic Scoliosis."²⁹ This reported on 37 peer-reviewed articles on scoliosis that were published between the years 1975 and 1993, which reflected the outcomes of 1,459 braced patients, 322 patients treated with lateral electrical stimulation, and 129 patients treated only with observation. Although the methods of meta-analysis can be controversial, this group concluded that bracing alters the natural history of progressive scoliosis, whereas electrical stimulation did not yield a significantly different outcome than that seen with observation. The meta-analysis also suggested the Milwaukee brace or thoracolumbosacral orthosis that is worn 23 hours a day is the most successful treatment regimen; bracing 8 or 16 hours per day is less effective than bracing 23 hours per day ($p < .0001$).

Then in 1995, on behalf of the Brace Study Group of the SRS, Nachemson and Peterson³⁰ wrote a much-needed pro-

spective review of the efficacy of bracing girls with thoracic and thoracolumbar curves, ranging in size from 25° to 35°. The authors defined a treatment success as no curve progression beyond 5°; 111 patients were braced, with a 74 percent treatment success rate; 129 were observed, with a 34 percent success rate; and 46 were treated with electrical stimulation, with a 33 percent success rate ($p < .0001$). This study showed that even in a worst case scenario, in which cases lost to follow-up were counted as treatment failures, bracing significantly alters the natural history of curve progression in this patient population. It also showed, as previous reports suggested,³¹ that electrical stimulation of select trunk musculature did not change the natural history of IS in these patients.

From the mid-1990s to the time of this writing, reports have become more consistent with respect to study design, bolstering the argument of the ability of orthoses to alter the natural history of adolescent IS.³¹⁻³⁵ The most common criteria in these most recent reviews involve adolescents who are Risser 0, 1, or 2, with curves ranging from 25° to 45°, for whom treatment success was defined as curve progression of no more than 5° at the end of treatment. Surgical rates in those treated with a brace also are reported as criteria for brace efficacy. In general, some reports suggest orthoses designed for nighttime wear are selectively effective in single lumbar and single thoracolumbar curves of less than 35°,^{33,36} whereas other reports suggest single thoracic curves may be effectively controlled by this approach to part-time bracing.^{32,35,37,38} Some reports suggest double curves can be successfully treated with nocturnal brace designs,^{32,35} but treatment success rates for this population generally are lower than those for treating single curves using this approach. Other researchers suggest double curves should be treated only with an orthosis designed to be worn in the upright position (thoracolumbosacral orthosis or Milwaukee brace design), with many placing a universal emphasis on this approach in the treatment of larger curves (between 36° and 45°).^{29,33,34,36,39,40} However, boys appear to experience less response to brace treatment than do girls.^{33,41}

INDICATIONS FOR ORTHOTIC TREATMENT FOR IDIOPATHIC SCOLIOSIS

Orthotic treatment for IS typically should be considered in skeletally immature individuals (typically defined as having 2 or more years of growth remaining) with a curve that is large enough to fall within a natural history category that suggests additional worsening of the deformity is likely.

For infantile onset IS, the amount of apical vertebral rotation and the RVAD, as described by Mehta,²² should be considered. This can help differentiate between a curve that may otherwise be expected to spontaneously resolve versus one that would otherwise be expected to worsen with time.

For juvenile or adolescent onset IS, the SRS suggests the following: "Brace (orthotic) management of adolescent IS is used in children with spinal deformity and curve magnitudes of 25–40° who are skeletally immature and with significant

growth remaining."⁴² More specifically, most current reports in the literature suggest orthotic management is indicated in adolescent IS for those who are Risser 0, 1, or 2, with a curve ranging from 25° to 45°.^{31,33-35,43-46} No natural history studies support the idea of bracing a curve less than 20°. Lonstein and Winter³⁴ suggest any adolescent presenting with a curve of at least 25° and Risser 0 maturity should begin orthotic treatment immediately. However, there is still some question in the literature regarding the possible value of observing for curve progression in those with curves between 20° and 29° before initiating orthotic treatment. Most researchers agree that curves of 30° to 39° in subjects with significant growth remaining should be braced upon presentation, and curves ranging from 40° to 45° are considered in the upper end of orthotic treatment candidacy. Curves larger than 45° typically are thought to exceed the limit at which bracing may be effective.

TREATMENT OBJECTIVES

The goal for all orthotic treatment is to stop curve progression, thus preventing the need for surgical correction or stabilization. It is not uncommon for a curve that progressed to a point at which orthotic treatment is needed to demonstrate some straightening during treatment. However, in time the curve is expected to return to its original size at prescription if treated successfully with an orthosis. Any permanent correction is considered a "bonus."

CONTRAINDICATIONS TO ORTHOTIC TREATMENT AND RATIONALE FOR DISCONTINUATION OF BRACE USE

Most studies concur that large curves (exceeding 45°) in the growing adolescent are both a contraindication to treatment and a reason to abandon orthotic treatment for the pursuit of surgical stabilization. In some patients with larger curves, even if progression can be controlled by an orthosis, the curve itself may be considered cosmetically unacceptable, thus indicating surgical correction.⁴⁷ According to the SRS: "Surgery for IS is suggested when curve magnitude is 50° or more in either the previously untreated patient or in one who fails brace treatment. Surgery is undertaken with two goals in mind. The primary one is to prevent spine deformity progression and the secondary one is to diminish spinal deformity. The natural history of IS during adulthood is one of continued progression if the curves tend to be more than 50° at the end of growth."⁴²

Another contraindication to orthotic treatment may be for patients who find the wearing of an orthosis to be emotionally intolerable.⁴⁷ The entire clinic team and parents or caregivers of the patient need to be cognizant of this possibility. However, psychological counseling may prove helpful in enabling an otherwise reluctant adolescent to accept orthotic treatment. Other contraindications may include:

- Extreme thoracic hypokyphosis,⁴⁸ for which corrective

forces from an orthosis may be impossible to apply to the scoliotic curve without significantly worsening the hypokyphosis that exists.

- Patients in whom bracing would have no benefit because of skeletal maturity.
- A deformity located in the high thoracic (apex cephalad to T7) or cervicothoracic spine, where orthotic treatment has not been shown to be effective in controlling curve progression. A superstructure may be used in such cases, but applying forces at or around the shoulder girdle makes in-brace curve correction and patient tolerance a formidable challenge.

Brace use discontinuation generally is indicated in those who are Risser 4 or 5 and, if the patient is female, 2 years beyond menarche.^{40,47}

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