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2-1-2007

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Management of Juvenile Idiopathic Scoliosis

By Lawrence G. Lenke, MD, and Matthew B. Dobbs, MD

Idiopathic scoliosis is a structural, lateral curvature of the spine for which no etiology has been established. Chronologically, idiopathic scoliosis can be categorized on the basis of the age of the patient at first identification of the deformity: infantile (birth to two years and eleven months), juvenile (three years to nine years and eleven months), and adolescent (ten years to seventeen years and eleven months). Thus, this article will describe the scoliotic deformities of patients who are at least three years of age but younger than ten years of age when the deformity is first identified. It has been demonstrated that spinal growth is fairly steady during this juvenile period[1]. For this reason, Dickson and Archer believed that true juvenile-onset scoliosis was rare enough not to warrant a separate category. They proposed a two-group classification that included early onset (five years of age or less) and late onset (six years of age and older) scoliosis[2]. In addition, patients who receive a diagnosis of scoliosis at five years of age or younger have a much higher chance of having a large curve develop, which may lead to pulmonary complications and cor pulmonale[1,3]. In this paper, we adhere to the classic age-at-onset definition as described by Dickson and Archer but do not describe adolescent idiopathic scoliosis.

Natural History and Epidemiology

Between 12% and 21% of patients with idiopathic scoliosis can be included in the category of juvenile idiopathic scoliosis[4], the gradual transition period between infantile idiopathic scoliosis and adolescent idiopathic scoliosis. This is certainly true with regard to sex predilection: the female-to-male ratio is 1:1 in children between three and six years of age, 2:1 to 4:1 overall in children who are at least three but less than ten years of age[5], and 8:1 by the time the children are ten years of age, which is a ratio close to that reported for patients with adolescent idiopathic scoliosis[1]. Hefti and McMaster reported that males are normally diagnosed with juvenile idiopathic scoliosis at approximately five years of age, whereas females are diagnosed at approximately seven years of age[6]. This difference, coupled with the fact that most males become skeletally mature at a later age than females, means that the risk of curve progression is higher for males with early onset juvenile idiopathic scoliosis than for females.

The natural history of juvenile idiopathic scoliosis is usually slow-to-moderate progression[4-8]. Since these curves occur at such a young age, there is a higher risk of severe deformity for these patients than there is for patients with adolescent idiopathic scoliosis. Approximately 70% of curves in patients with juvenile idiopathic scoliosis progress and ultimately require some type of treatment. Tolo and Gillespie reported progression in forty-two of fifty-nine curves (71%), sixteen of which required surgery[9]. Similarly, Figueiredo and James reported that fifty-five of ninety-eight patients (56%) with juvenile idiopathic scoliosis required spinal surgery[8]. Mannherz et al., in a retrospective review of forty-three patients, found that thirty-one curves required bracing, twenty-six continued to progress, and thirteen ultimately required surgery[10]. However, there are curves that do not progress or that may actually regress with time. Mannherz et al. found that, in six of twelve patients, the curves that did not require treatment ultimately resolved[10]. All patients presented with curves of <25°. Thus, most juvenile curves do progress while some spontaneously regress, but regression does not occur as often in juvenile curves as it does in infantile curves.

The curve patterns of juvenile idiopathic scoliosis very closely resemble those of adolescent idiopathic scoliosis, with right thoracic and double major curves predominating. Dobbs, Lenke, and Bridwell modified the adolescent idiopathic scoliosis classification system of Lenke et al. for the purpose of classifying juvenile idiopathic scoliosis[11,12]. Similar to the adolescent idiopathic scoliosis classification system, six curve types were designated: type 1, main thoracic; type 2, double thoracic; type 3, double major; type 4, triple major; type 5, thoracolumbar/lumbar; and type 6, thoracolumbar/lumbar-main thoracic. Instead of using side-bending radiographs to distinguish structural from nonstructural minor curves, the authors utilized the deviation from the midline of the apex of the curve from the main thoracic region (from the C7 plumb line) and the thoracolumbar/lumbar region (from the center sacral vertical line). In a structural minor curve, the apex of the curve is completely off the line, whereas the

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apex in a nonstructural minor curve is not. The structural characteristics of the proximal portion of the thoracic region are determined from evaluation of the first rib in comparison with the main thoracic curvature. For a right thoracic curve, if the left first rib is elevated, then the proximal portion in the thoracic region is structural. If the right first rib is higher than or level with the left first rib, then the proximal thoracic region is nonstructural. In addition, a lumbar modifier and a sagittal thoracic modifier are also assigned, similar to the adolescent idiopathic scoliosis classification system of Lenke et al.[12] (Figs. 1-A and 1-B).

Evaluation of a patient with juvenile idiopathic scoliosis is very similar to that of a patient with adolescent idiopathic scoliosis. However, obvious growth differences and skeletal maturation patterns for these young patients must be evaluated. In addition, there is a much higher risk of an intraspinal abnormality, and it is recommended that any juvenile patient with a scoliosis of >20° undergo magnetic resonance imaging of the total spine to rule out an intraspinal abnormality such as a syringomyelia or an Arnold-Chiari malformation[13,14]. Approximately 20% of juvenile patients will have an intraspinal abnormality on magnetic resonance imaging[13,14]. In addition to a careful evaluation of the trunk and extremities, a thorough neurologic examination must be performed as well.

Baseline radiographic evaluation includes an upright or standing long-cassette radiograph of the entire spine and pelvis in frontal (posteroanterior) and lateral views, and Cobb angles of all curves are then measured. Although the Mehta angle (the rib-vertebra angle difference [RVAD]) can also be measured, it has not been as useful in predicting progression of juvenile idiopathic scoliosis as it has been for infantile idiopathic scoliosis[15].

Treatment

Observation

Observation is still the main treatment for the majority of small curves in patients with juvenile idiopathic scoliosis, especially for curves that are <20° to 25° at first recognition.

TABLE I Juvenile Idiopathic Scoliosis: Traditional Treatment Options
1. Anterior convex epiphysiodesis combined with anterior hemiarthrodesis
2. Anterior and posterior spinal fusion with or without posterior instrumentation
3. Anterior spinal fusion with instrumentation (older juveniles)
4. Posterior growing single-rod systems
5. Posterior Luque trolley growing-rod system

Follow-up every four, six, nine, or twelve months is indicated, based on the age of the patient, the magnitude of the curve, and the characteristics of the clinical deformity[14,16]. For curves of 25° to 30° or higher, some treatment should be considered because of the high probability of progression.

Orthotic Management

Curves between 25° and 50° and even up to 60° are initially treated with an orthotic or bracing program[16,17]. Although an underarm thoracolumbosacral orthosis (TLSO) or a Boston-type brace is usually effective for patients in this age range (depending on the location of the apex of the major curve), traditionally the cervicothoracolumbosacral orthosis (CTLSO) or Milwaukee brace has been preferred for these young patients[18]. The thoracolumbosacral brace should be used with caution because of the amount of rib-cage compression that can be attained with this brace and because of the length of time that is usually required for brace treatment. In contrast, the Milwaukee brace provides correction more by way of axial lengthening than by rib-cage compression. Thus, the Milwaukee brace is generally preferred if the curve is fairly flexible. If the curve is rigid, then preliminary serial casting via a Risser cast, similar to what is recommended for patients with infantile idiopathic scoliosis, is commenced to obtain some correction prior to fitting the child with a Milwaukee brace[16,17].

Lumbar Spine Modifier	CSVL to Lumbar Apex	
A	CSVL between pedicles	
B	CSVL touches apical body(ies)	
C	CSVL completely medial	

Fig. 1-A
Lumbar spine modifiers A, B, and C are described. CSVL = center sacral vertical line.

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TABLE II Juvenile Idiopathic Scoliosis: Newer Treatment Options
1. Intervertebral stapling
2. Intervertebral tethering
3. Rib-cage distraction
4. Dual posterior growing-rod constructs (with hook/screw implants)
5. Shilla technique of posterior apical growth arrest construct with bilateral screw fixation

Bracing is performed on a part or full-time basis, depending on the size of the curve and the age of the child. Reported success with bracing in the management of juvenile idiopathic scoliosis has been variable. Kahanovitz et al. reported an excellent prognosis with part-time bracing for curves ≤35° and RVADs ≤20°[18]. However, patients with curves of ≥45° and RVADs of ≥20° had a poor prognosis for successful brace treatment. If curves progress despite brace treatment, the goals of treatment obviously change. Usually orthotic treatment will have to be abandoned once the curve is 60°, depending on curve flexibility and the size of the patient. Thus, surgical management may need to be considered, especially with progression despite good compliance with brace-wearing. Certainly, surgery should be avoided if at all possible in very young juveniles (three to six years of age).

Surgical Management

Surgical treatment is not as clearly indicated for juvenile idiopathic scoliosis as it is for adolescent idiopathic scoliosis. Because of the differing characteristics of a patient who presents at three years of age as compared with one who presents at nine years of age, the decision to proceed with surgery can be extremely variable and difficult.

There are many important considerations in the surgical treatment of patients who have juvenile idiopathic scoliosis. One of the foremost issues is the expected loss of spinal height, and thus limited chest-wall growth and lung growth, due to a spinal fusion procedure. Winter devised a formula for determining the amount of potential shortening of the spinal column following spinal fusion: 0.07 cm multiplied by the number of vertebral segments fused and then multiplying the product by the number of years of remaining spinal growth[19]. This formula assumes complete cessation of longitudinal spinal growth after posterior fusion and thus allows the surgeon to inform the family of the estimated postoperative spinal shortening. However, the family must understand that, without spinal fusion, more truncal height will be lost because of the untreated progressive scoliotic deformity.

Another important consideration is the crankshaft phenomenon, which describes continued anterior growth and spinal curvature with increased rib prominence despite a solid posterior fusion in a skeletally immature patient[20]. Most patients with juvenile idiopathic scoliosis and open triradiate cartilages will be at risk for the crankshaft phenomenon, as documented by Sanders et al.[21]. To prevent it, anterior release and fusion has been recommended in addition to a posterior fusion (Figs. 2-A, 2-B, and 2-C). Another possible option is the use of segmental bilateral pedicle screws to maintain a posterior-only fusion without an anterior fusion, although this method has not yet been proven efficacious in the juvenile idiopathic scoliosis patient population. Even patients with posterior growing-rod instrumentation can be affected by the crankshaft phenomenon.

Growing-Rod Systems

Many standard and nonstandard surgical treatments may be applicable to patients with progressive juvenile idiopathic scoliosis (Tables I and II). The most common traditional treatment is a posterior instrumentation and fusion. Because of concern about the crankshaft phenomenon, often a preliminary anterior release and fusion is also performed[22]. Obviously, one of the main concerns with posterior instrumentation and fusion, especially in the thoracic spine, is diminished chest-wall height and volume and, thus, limited lung development and subsequent growth. For this reason, a growing-type of posterior instrumentation has been in use for several decades in an attempt to sequentially lengthen the

Thoracic Sagittal Profile T5-T12		
-	(Hypokyphosis)	< 10°
N	(Normal)	10° - 40°
+	(Hyperkyphosis)	> 40°

Fig. 1-B
Sagittal thoracic modifiers are described for hypokyphosis (–), a normal curve (N), and hyperkyphosis (+).

Fig. 2-A

(Left to right). Anteroposterior, left-side-bending, right-side-bending, and lateral radiographs showing a progressive left thoracic curve to 64° with appropriate flexibility in a nine-year and five-month-old female with an associated syringomyelia and Arnold-Chiari decompression.
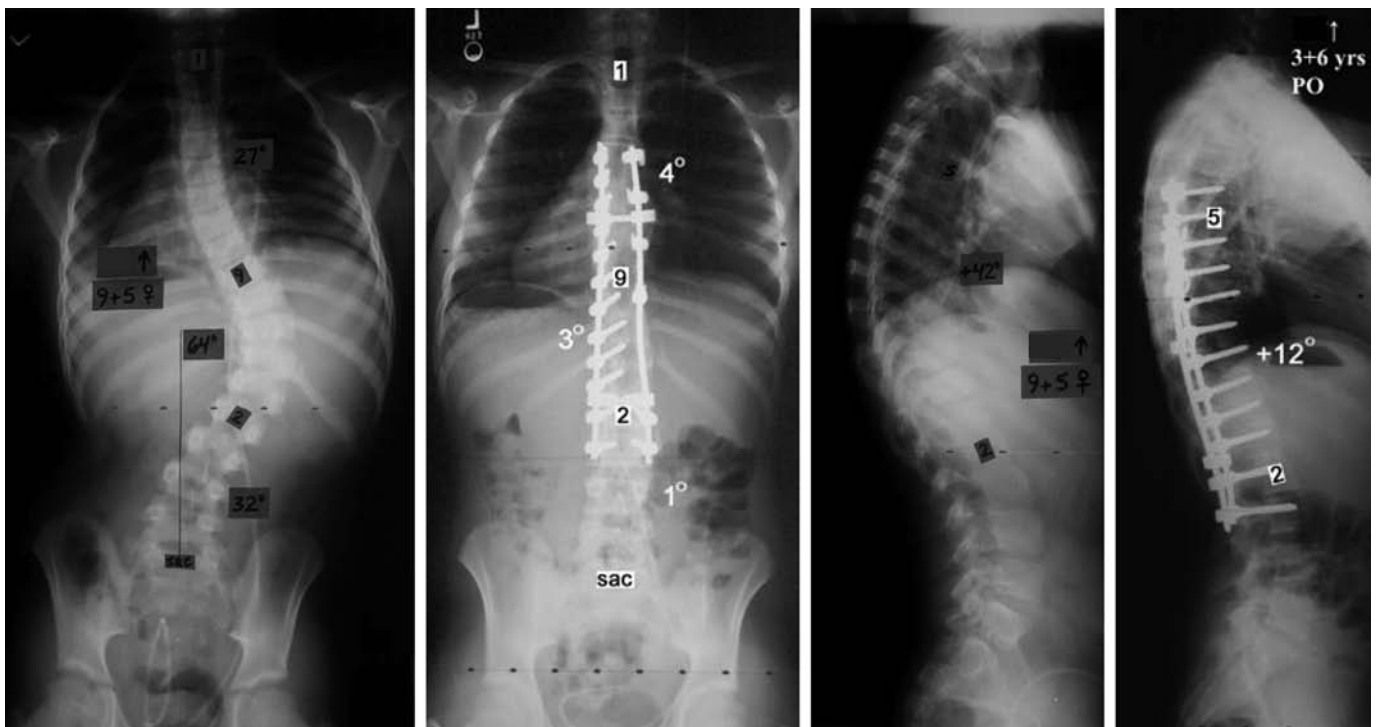

Fig. 2-B

(Left to right). Preoperative and postoperative anteroposterior and lateral radiographs of the patient, who underwent a same-day anteroposterior spinal fusion from T4 to L3 with placement of a 4.5-mm screw-rod system. As of the time of the 3.5-year postoperative follow-up, the patient had maintained excellent coronal and sagittal balance.
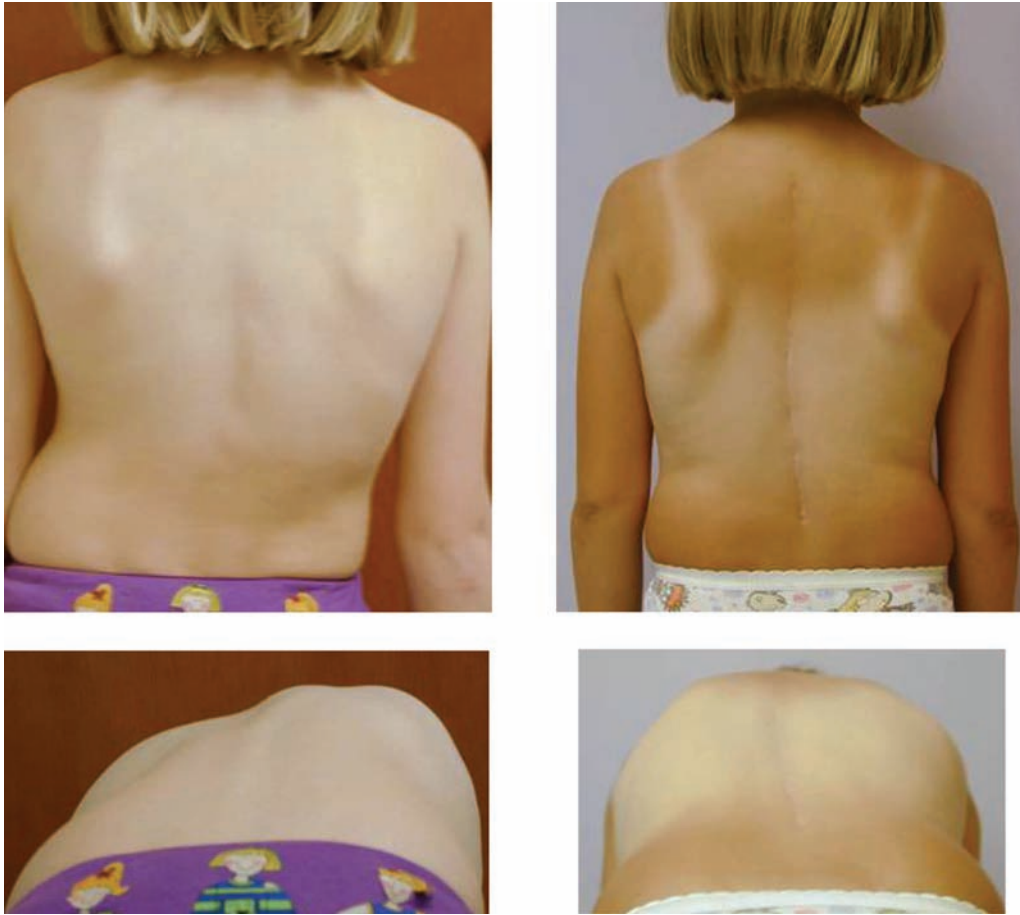
Fig. 2-C
Preoperative (left) and postoperative (right) clinical photographs of the deformity.

spine and allow longitudinal growth while still attempting to control progressive spinal deformities[23-25]. This approach, however, has been met with less-than-ideal results. The standard growing-rod construct, consisting of a claw-hook proximal as well as distal to the curve, with both hooks connected to a single rod, led to many instrumentation failures, including implant pull-out and rod breakage, requiring multiple revision surgeries[23-25]. In addition, unintended spontaneous spinal fusion or autofusion may occur over the instrumented levels[26]. In addition to internal fixation, long-term brace-wear may also be required[27].

Recently, the use of dual rod implants, as is standard practice in conventional posterior instrumentation and fusion, and the addition of pedicle screws in the cephalad and/or caudad aspect of the implant construct, have also become quite popular in an attempt to lessen the instrumentation problems of the past[28] (Figs. 3-A through 3-D). In addition, the performance of an anterior and posterior growth arrest with fusion has been found to provide suboptimal results compared with the use of dual growing-rod constructs without apical fusion[28]. Other growing-rod construct options include the traditional Luque trolley[29], in which sublaminar wires are utilized, and a new type of nonlocking pedicle screw implant called the Shilla technique, which was recently devised by McCarthy[30]. In this system, the apex of the deformity is fixed and fused with pedicle screws, while the ends of the construct are instrumented with screws that are not locked to the rod. Thus, this technique theoretically allows for apical control of the deformity and continued axial lengthening of the spine with growth. This system can be considered a type of "pedicle screw-trolley" procedure.

Another recent growing-rod system, developed by Campbell et al., utilizes implants that extend from rib-to-rib or ribs-to-spine in an attempt to control progressive juvenile spinal deformity and expand the chest[31]. In these techniques, claw-hooks on bilateral ribs are applied laterally, with caudal implants resting on ribs that are more distal or more often on the upper to midlumbar spine than the ribs chosen for use with hook or pedicle screw implants. These types of hybrid rib-spine constructs are as yet unproven for the treatment of juvenile idiopathic scoliosis; however, they represent another option in order to avoid exposure and instrumentation of proximal thoracic vertebrae, which are often quite small and provide a more tenuous fixation in very young patients.

Halo-Gravity Traction

One common technique often utilized, especially in juvenile patients with severe scoliotic deformities (>80°, with or without a severe thoracic or thoracolumbar kyphosis >70°), is the use of halo-gravity traction[32]. In this technique, with the patient under general anesthesia, a six up to eight-pin halo is placed; then, the patient is placed in upright halo-gravity traction for two to six weeks or longer. Daily, the patient sits, stands, and walks while undergoing axial traction applied to the spine through the halo. Progressive weight is added until 32% to 50% of body weight is applied, which usually allows the entire trunk to be suspended in the sitting position by the traction weight through the skull. We have found this technique to be very useful for slow correction of severe deformities, especially in young patients and in those with fairly severe preoperative pulmonary compromise, as it provides a safer environment for definitive anterior and/or posterior procedures. In addition, the axial traction performed while the patients are awake, in our experience, has decreased the risk of the development of neurologic deficits during the traction and subsequent surgical correction[32]. As there is a risk of pin-site problems and cranial nerve palsy from the traction, the patients must be monitored carefully. Acute cranial nerve palsies usually respond to a lessening of traction weight. Traction can be applied preoperatively or perioperatively following anterior release in very serious cases.

Anterior and Posterior Spinal Fusion

Definitive procedures for older juvenile patients (eight to ten years of age) usually consist of an anterior and posterior spinal fusion. This is usually performed on the same day with an open or endoscopic anterior release followed by a definitive posterior spinal fusion and segmental spinal instrumentation with use of an appropriate-size instrumentation system for these often small-size patients[33-37]. It is very important not to attempt to cheat proximal or distal levels as continued growth will often lead to an "adding-on phenomenon" with deformity progression cephalad to and/or caudad to the spinal instrumentation and fusion levels. Standard segmental spinal instrumentation techniques are performed with hooks, wires, and/or pedicle screws, depending on the preference of the surgeon. In these young patients it is important to achieve a solid posterior fusion mass, which can be accomplished with autograft bone supplemented with allograft bone as required.

Occasionally, if the patient is large enough, isolated anterior instrumentation and fusion may be applied in an attempt to correct a single major curve. However, one must always be aware that continued spinal growth may cause other minor curves to progress even if those curves initially seem innocuous. Anterior instrumentation and fusion often can be done in the thoracolumbar and/or lumbar region, but the limiting factors are often the size of the vertebral bodies to be instrumented and the bone, which is often quite soft. Single-screw, single-rod constructs may be applied along with anterior structural devices, such as structural allograft bone or titanium cages in the thoracolumbar/lumbar spine, if required, to maintain optimal sagittal alignment[38,39].

Growth Modulation Techniques

Alternative devices that modulate spinal growth are becoming available, including intervertebral stapling to produce a tethering effect. Convex disc stapling represents an attempt to apply principles similar to those used in physeal stapling in the lower extremities for correction of angular malalignment. Thus with convex stapling, continued concave growth will hopefully maintain or even correct scoliosis deformities over time[40]. Indications for the use of this technique in patients with juvenile idiopathic scoliosis are not precisely known at this time. Posterior growing-rod instrumentation and definitive spinal fusion may be performed at any time in patients with juvenile idiopathic scoliosis who have previously undergone stapling anteriorly.

Another treatment option that may be available in the future for progressive juvenile idiopathic scoliosis is anterior tethering. In this technique, single screws are placed in the vertebral bodies anteriorly, and then a polypropylene tether
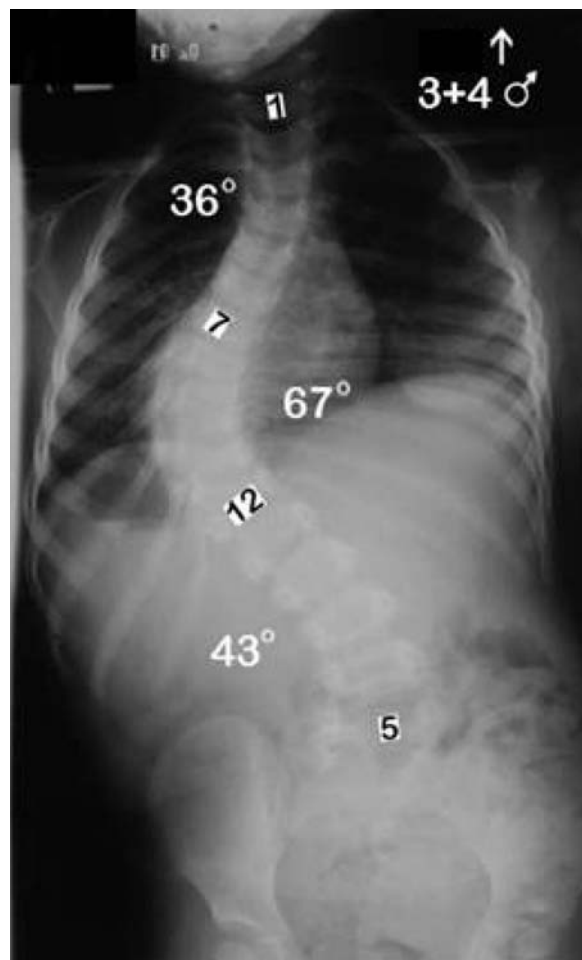

Fig. 3-A

Radiograph showing a three-year and four-month-old male with a left thoracic scoliosis that has progressed to 67°.
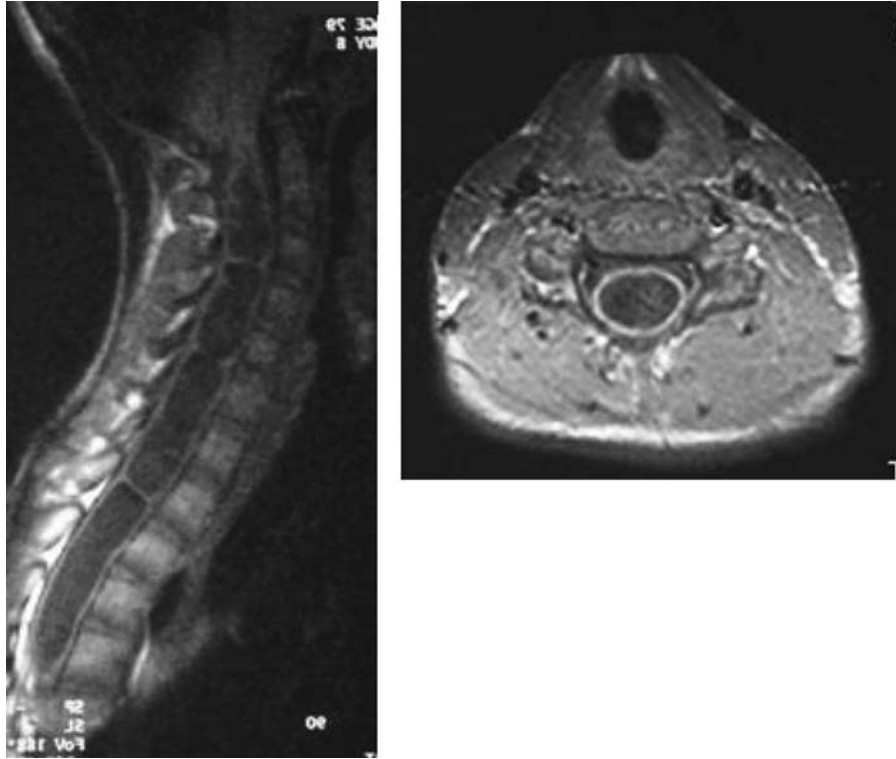
Fig. 3-B

T1-weighted magnetic resonance image of the cervicothoracic junction showing an associated Arnold-Chiari malformation and syringomyelia.
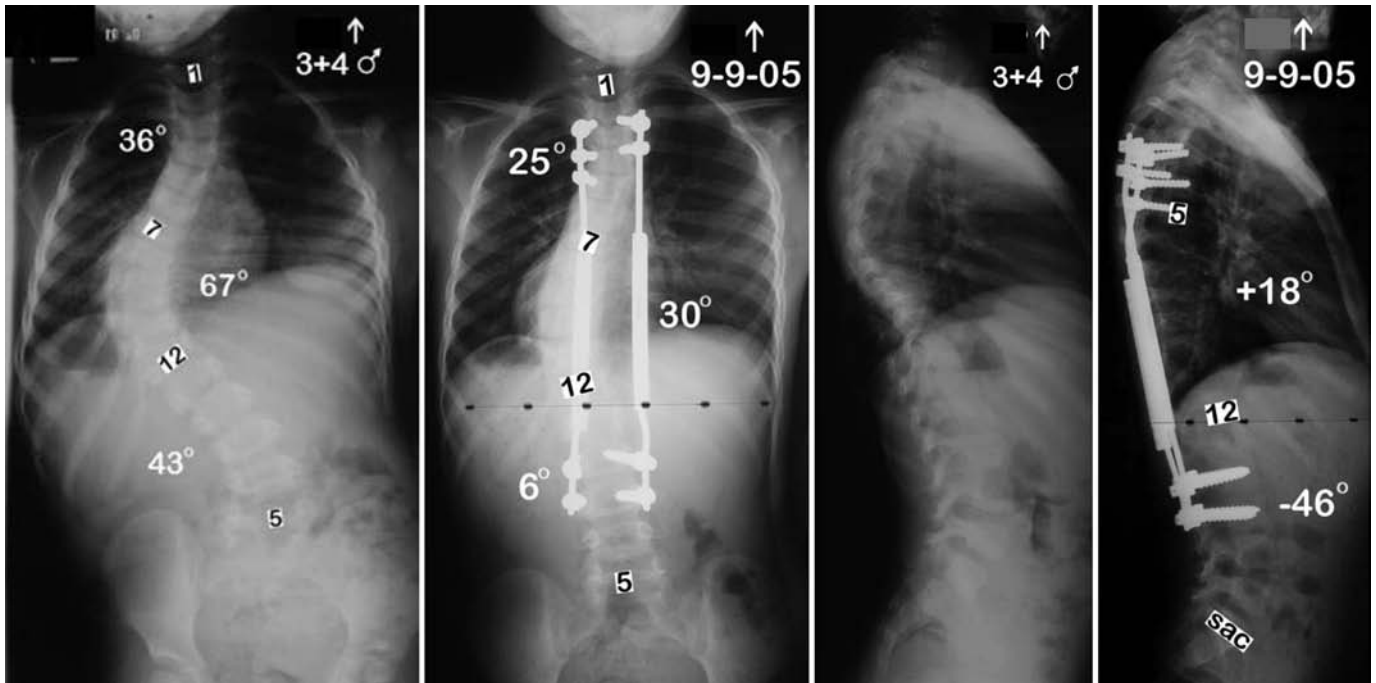

Fig. 3-C

Preoperative and postoperative anteroposterior and lateral radiographs showing the curve before and after placement of a posterior growing-rod construct that was comprised of a 3.5-mm screw-rod system with growth connectors-lengtheners. The scoliosis was nicely controlled through lengthening of the construct every six months.
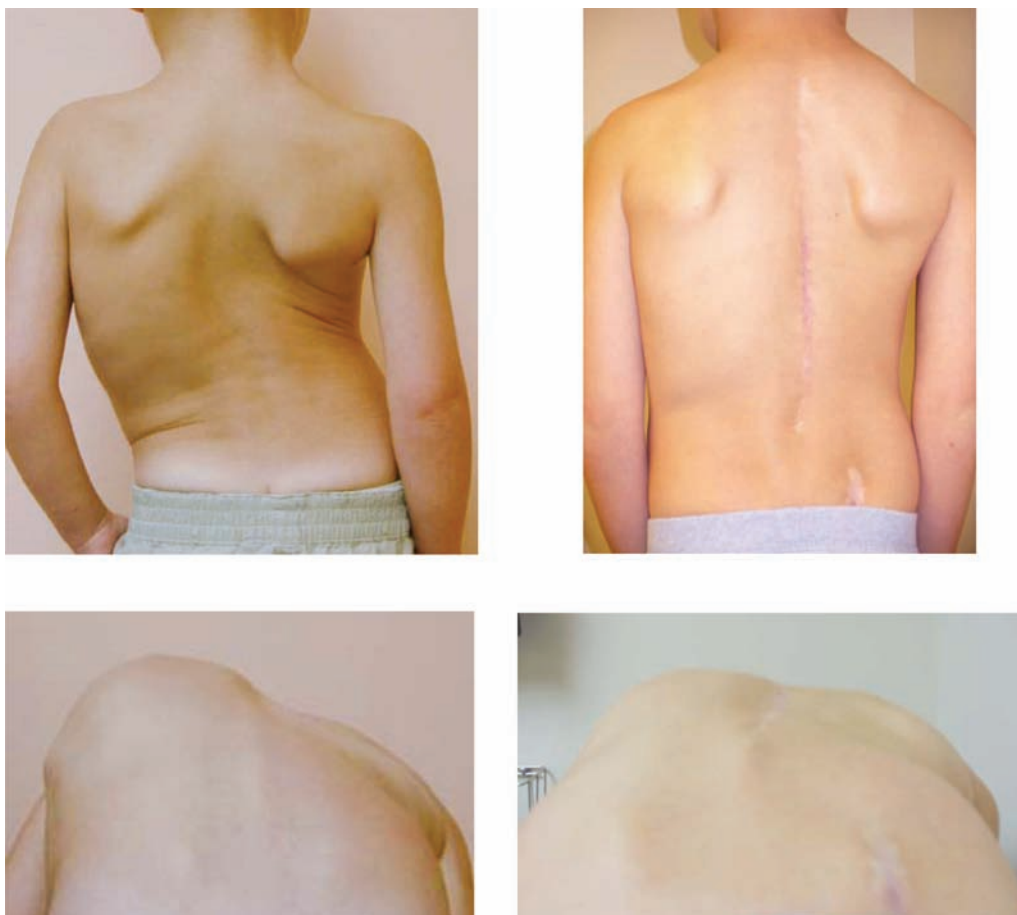
Fig. 3-D
Preoperative (left) and postoperative (right) clinical photographs of the deformity.

is placed between the screw heads, which may be shortened and compressed to correct the deformity during this surgical procedure. In this technique, the discs are left unaffected, and motion of the spine, although not normal, theoretically allows 5° of freedom, with a loss of motion only when bending away from the tether (to the left for a right-sided scoliosis tethering). Stapling and/or tethering procedures are growth-modulation procedures, which ultimately may be utilized for smaller deformities in an attempt to avoid secondary manifestations of scoliosis that are much more difficult to correct with definitive spinal instrumentation and fusions.

Discussion

All surgical procedures to correct scoliosis involve a struggle to control spinal deformity in a growing spine while guarding against the complete immobilization of the vertebrae. It is hoped that the next decade will usher in major advances in the treatment of juvenile idiopathic scoliosis, as these deformities are among the more challenging of progressive spinal deformities. There will come a day when progressive scoliosis will be treated with correction of the deformity, either internally or externally, without a spinal fusion. Until then, we should analyze each deformity on an individual basis, keeping in mind that the optimal treatment for the control or correction of deformity will be the one that entails the least amount of short and long-term morbidity. ■

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