

# Derotational Casting for Progressive Infantile Scoliosis

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**Background:** Serial cast correction by using the Cotrel derotation technique is one of several potential treatments for progressive infantile scoliosis. This study reviews our early experience to identify which, if any, patients are likely to benefit from or fail this technique.

**Methods:** We followed all patients treated at our institutions for progressive infantile scoliosis since 2003 prospectively at 1 institution and retrospectively at the other 2. Data, including etiology, Cobb angles, rib vertebral angle difference, Moe-Nash rotation, and space available for the lung, were recorded over time.

**Results:** Fifty-five patients with progressive infantile scoliosis had more than 1 year of follow-up from the initiation of casting. The diagnosis of progressive scoliosis was made based upon either a progressive Cobb angle or a rib vertebral angle difference of more than 20 degrees at presentation. All but 6 patients responded to cast correction. Nine patients have undergone surgery to date, 6 because of worsening and 3 by parent choice. As shown in the table, initiation of cast correction at a younger age, moderate curve size (< 60 degrees), and an idiopathic diagnosis carry a better prognosis than an older age of initiation, curve > 60 degrees, and a nonidiopathic diagnosis. The space available for the lung improved from 0.89 to 0.93. No patient experienced worsening of rib deformities.

**Conclusions:** Serial cast correction for infantile scoliosis often results in full correction in infants with idiopathic curves less than 60 degrees if started before 20 months of age. Cast correction for older patients with larger curves or nonidiopathic diagnosis still frequently results in curve improvement along with improvement in chest and body shape.

**Significance:** Derotational cast correction seems to play a role in the treatment of progressive infantile scoliosis with cures in young patients and reductions in curve size with a delay in surgery in older and syndromic patients.

**Level of Evidence:** Level 4, therapeutic study.

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Progressive infantile scoliosis, also termed early-onset scoliosis, is one of the few potentially fatal noncancerous orthopaedic disorders if left untreated. Increased mortality seems to come from poor development of the chest's mechanics and volume, and from poor lung development. Campbell et al<sup>1</sup> have termed the pulmonary failure from this condition "thoracic insufficiency syndrome." Unfortunately, our basic treatments for scoliosis can also result in poor thoracic growth, particularly with definitive fusion before the age of 10 years.<sup>2,3</sup>

Various methods of growing instrumentation have been developed in an attempt to retain basic spinal alignment while continuing to allow further spine and subsequent chest growth. Current treatments, primarily the vertical expandable prosthetic titanium rib (VEPTR) and growing rods,<sup>4</sup> aim at delaying definitive fusion and almost never focus on a cure. Practitioners experienced in these methods are well versed in these difficulties. Growing rods often fracture, pull loose, develop infections, and create a variant of the crankshaft phenomenon with growth. VEPTR has similar complications with the additional problem of creating stiff, noncompliant chest walls and causing stretch on the brachial plexus.<sup>5</sup>

Cast correction represents another alternative for scoliosis, which was quite common until the development of effective spinal instrumentation. Casting itself can create pressure sores, significant rib or mandibular deformities, and constrict the chest. The historic term for superior mesenteric artery syndrome is even called "cast syndrome." Most of these problems seem to be the result of indiscriminate casting of all types of scoliosis with the use of improper techniques combined with a limited understanding of spinal, and particularly of chest wall deformities.

Scoliosis cast correction comes in several varieties. The most commonly used method in the United States is Risser casting,<sup>6</sup> which uses a 3-point bend for correction. Although it is possible to obtain significant curve correction with this technique, it does not sufficiently account for rotational abnormality, and, especially in younger children with flexible bones, can cause significant rib deformities and chest constriction. In France, the technique of Cotrel and Morel's<sup>7</sup> EDF (extension, derotation, flexion) cast correction has been

more common. Recently, Mehta<sup>4</sup> described her results of cast correction in 136 patients with infantile scoliosis by using the technique of Cotrel and Morel<sup>7</sup> with the philosophy that early rapid growth, if guided by the cast, would assist an initially curved spine to straighten. We have used serial cast correction in a select group of patients and are reporting our results.

The terminology of "infantile" scoliosis versus "early onset" scoliosis is controversial. Advocates of "early onset," who defined as scoliosis occurring before 5 years of age, believe that it separates those patients who are likely to develop pulmonary failure from "late onset" scoliosis patients who are unlikely to develop pulmonary compromise. Advocates of "infantile scoliosis", defined as scoliosis age 3 years and below at the time of detection, believe it is more appropriate since juvenile scoliosis, scoliosis identified between the age of 4 and 10 years, still has increased mortality rates though less than infantile scoliosis, making the terms "infantile" and "juvenile" useful. We have followed Mehta's precedent<sup>4</sup> of using the term "progressive infantile scoliosis" in this paper since it well describes this population.

## METHODS

We prospectively followed all patients treated at Shriners Hospitals for Children in Erie, Pennsylvania, and retrospectively reviewed those treated at the Shriners Hospitals for Children in Salt Lake City, Utah, and Chicago, Illinois with progressive infantile scoliosis since 2003. Each of the centers obtained their patient data independently. All of the patients with progressive early onset scoliosis more than 25 degrees in Erie (27 patients) and Salt Lake City (26 patients) during this period were treated with cast correction. Chicago (2 patients) instituted cast correction late in this series after having provided other options previously. We did not cast children with congenital scoliosis. Etiologies were delineated as idiopathic for children with no discernable syndrome or neurological disorder or as syndromic for those with known or apparent syndromes. Rib vertebral angles and Cobb angles at each of the centers and the initial precasting and last out of cast radiographs compared. The Erie facility also recorded the Moe-Nash rotation<sup>8</sup> and the space available for the lung using the method described by Campbell et al.<sup>9</sup> Any complications from treatment were also noted. Results were determined by the final Cobb angle. Only patients with more than 1 year of follow-up were included in the analysis with the vast majority more than 2 years.

### Technique

A proper casting table is crucial. Although a standard Risser frame will suffice, it is quite large for small children. Two of the centers (Erie and Chicago) use a table designed by Mehta,<sup>4</sup> which leaves the head, arms, and legs supported but the body free. The other center (Salt Lake City) uses a custom-designed table that performs a similar function of supporting the child in

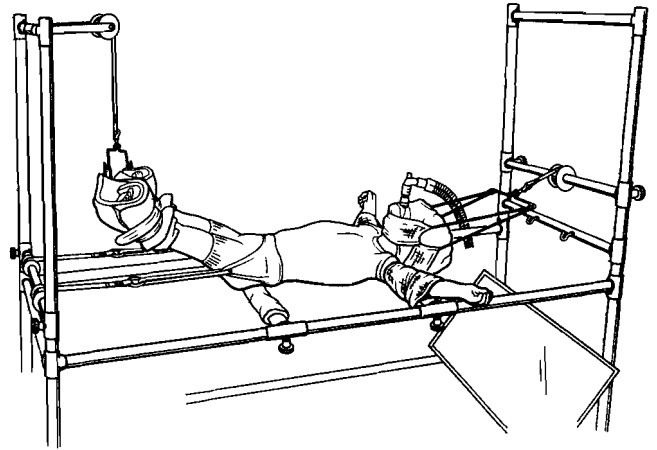


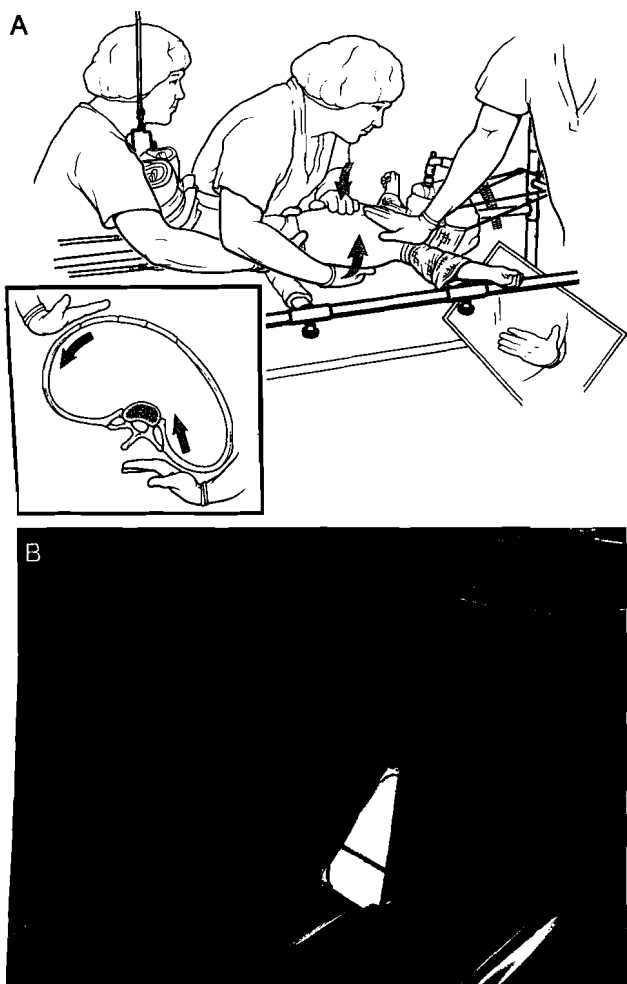
FIGURE 1. Positioning on the table with traction applied to head halter and pelvis.

traction while leaving the body free for cast application. Patients are intubated as thoracic pressure during cast molding can make ventilation temporarily difficult. A silver impregnated shirt is used as the innermost layer. Head halter and pelvic traction assists in stabilizing the patient and in narrowing the body (Fig. 1). Even though traction can correct the curve while applied, the position cannot be retained in the cast once traction is released and the body recoils. A mirror slanted under the table is useful for visualizing rib prominence, posterior cast, and molds. A thin layer of webril is applied with occasional felt on significant bony prominences. If there is a lumbar curve, the hips are slightly flexed to decrease lumbar lordosis and facilitate curve correction.

Plaster, or, in smaller curves, fiberglass, is applied. Plaster has the benefit of being more moldable than fiberglass and expands rather than constricts when set. The pelvic portion, as the foundation, is well molded. Cast molding is performed by using the EDF technique. We believe it important that the cast not push the ribs toward the spine with consequent narrowing of the space available for the lung. Rather, the posteriorly rotated ribs are rotated anteriorly to create a more normal chest configuration with counter rotation applied through the pelvic mold and upper torso (Fig. 2). Although the Cotrel-Morel technique<sup>7</sup> and Mehta's<sup>4</sup> modifications use an over-the-shoulder cast, one of the centers (Erie) has typically stayed below the shoulders as most infantile curves have low apices, typically at T9 to T11. An anterior window is made to relieve the chest and abdomen while preventing the lower ribs from rotating. A posterior window is made on the concave side allowing the depressed concave ribs and spine to move posteriorly. The 2 varieties of casts and the trim lines are shown in Figure 3A-C.

## RESULTS

Between the facilities, more than 100 patients with progressive infantile scoliosis have been treated with serial



**FIGURE 2.** A, The correction maneuver for derotation and translation of a typical left thoracic curve. Counter rotation is applied through the pelvis and pectoralis area. B, The rotational maneuver with the other individuals not in the photograph to show proper curve correction. The surgeon is using the mirror to identify proper location for the rotation.

cast correction. Fifty-five patients with progressive infantile scoliosis had more than 1 year of follow-up from the initiation of cast correction, 47 of these had 2 years or more. The diagnosis of progressive scoliosis was made based upon either a progressive Cobb angle 20 degrees or more or a rib vertebral angle difference (RVAD) of more than 20 degrees at presentation. Thirty-seven patients had no underlying syndrome, 2 had tethered cords, 1 had a minor brain abnormality, and 15 had syndromes. As the hospitals are referral centers, nearly all of the patients had been seen elsewhere, were often braced, and had documented progression. The average age at initial cast correction was 2.2 years (range: 7 to 64 mo) with an initial Cobb angle of 52 degrees (range: 25 to 100 degrees), an RVAD of 31 degrees (range: 5 to 82 degrees), and a Moe-Nash of 2.2 (range: 1 to 3). The vast majority of those whose curves have not resolved and who have not had surgery are still in casts

with periods of bracing, particularly during the summers expecting that their curves will require surgery in the future. This number is in continual flux. As the most detailed data are from the Erie site's prospective series, this more detailed information is presented initially with the entire series of all the sites then presented for comparison. As shown in the Table 1, initiation of cast correction at a younger age, moderate curve size (< 60 degrees), and an idiopathic diagnosis carry a better prognosis than an older age of initiation, curve of more than 60 degrees and a nonidiopathic diagnosis. Lesser rotation and a smaller RVAD are also indicative of success except in the double curves. The space available for the lung improved from 0.89 to 0.93 with no patient experiencing worsening rib deformities. The curve behavior of the Erie patients over time for the differing groups is shown in Figure 4. An example of a patient's radiographs is shown in Figure 5.

The results of the Salt Lake City and Chicago patients when added to those of the Erie patients have nearly identical results as shown in Table 2. All but 6 patients responded to cast correction by a decrease in curve magnitude manifest through the follow-up period. Nine patients have undergone surgery to date, 6 because of eventual worsening of the curves and 3 by parent choice.

The centers had very few complications with occasional cast irritation requiring short periods for recovery. The most important issue on a regular basis is temporary chest pressure making ventilation difficult while the cast is setting. Owing to this, we recommend intubating these patients for cast correction rather than relying on a mask or laryngeal mask airway.

## DISCUSSION

Infantile idiopathic scoliosis occurs in 2 basic types, resolving and progressive. Mehta was able to distinguish resolving from progressive scoliosis by using the RVAD of 20 degrees or more on an early supine radiograph highly prognostic of progression.<sup>10</sup> She also distinguished phase 1 from phase 2 rib rotation with phase 2, denoting the convex rib overlapping the vertebra body, always progressive. Double curves are problematic in that they may have a low RVAD except at the curve junction but are nearly always progressive. Mehta's findings have been confirmed by other investigators.<sup>11-14</sup>

Progressive infantile scoliosis is a problematic disease, which can be both disabling and fatal.<sup>15,16</sup> Definitive fusion before the age of 10 years can result in severe respiratory compromise. Owing to these difficulties, orthopaedists treating these patients have resorted to various methods of attempting to direct spine growth by using "growing" devices such as growth rods, the Shilla technique, or the VEPTR device.<sup>17,18</sup> Experience with these devices is fraught with difficulties and complications.

The Ponseti technique for clubfoot deformity has renewed the credibility of serial cast correction as superior to early surgery with its resulting scarring and stiffness.

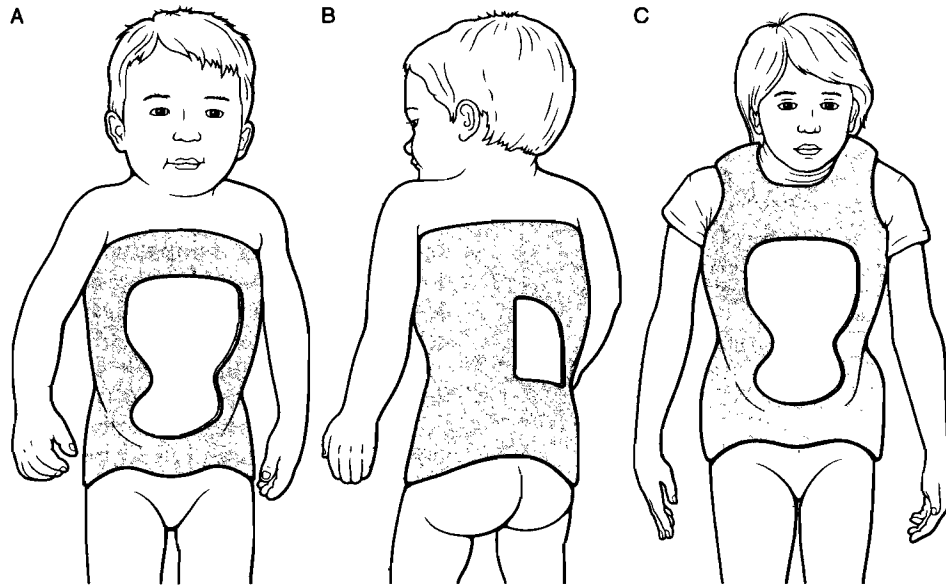


FIGURE 3. A, Under arm cast with timelines and windows anteriorly to allow abdominal distention while controlling ribs. B, Posterior window on concavity with posterior trim lines. C, Over the shoulders cast shown anteriorly. Posterior window is the same.

Mehta has reported on her experience in treating early onset scoliosis with serial cast correction to take advantage of the child's early flexibility and rapid growth to correct the deformity.<sup>4</sup> We began our cast correction for scoliosis with the goal of using the child's intrinsic growth capabilities and serial correction to the child's benefit while avoiding the complications and stiffness resulting from early growing rod surgery.

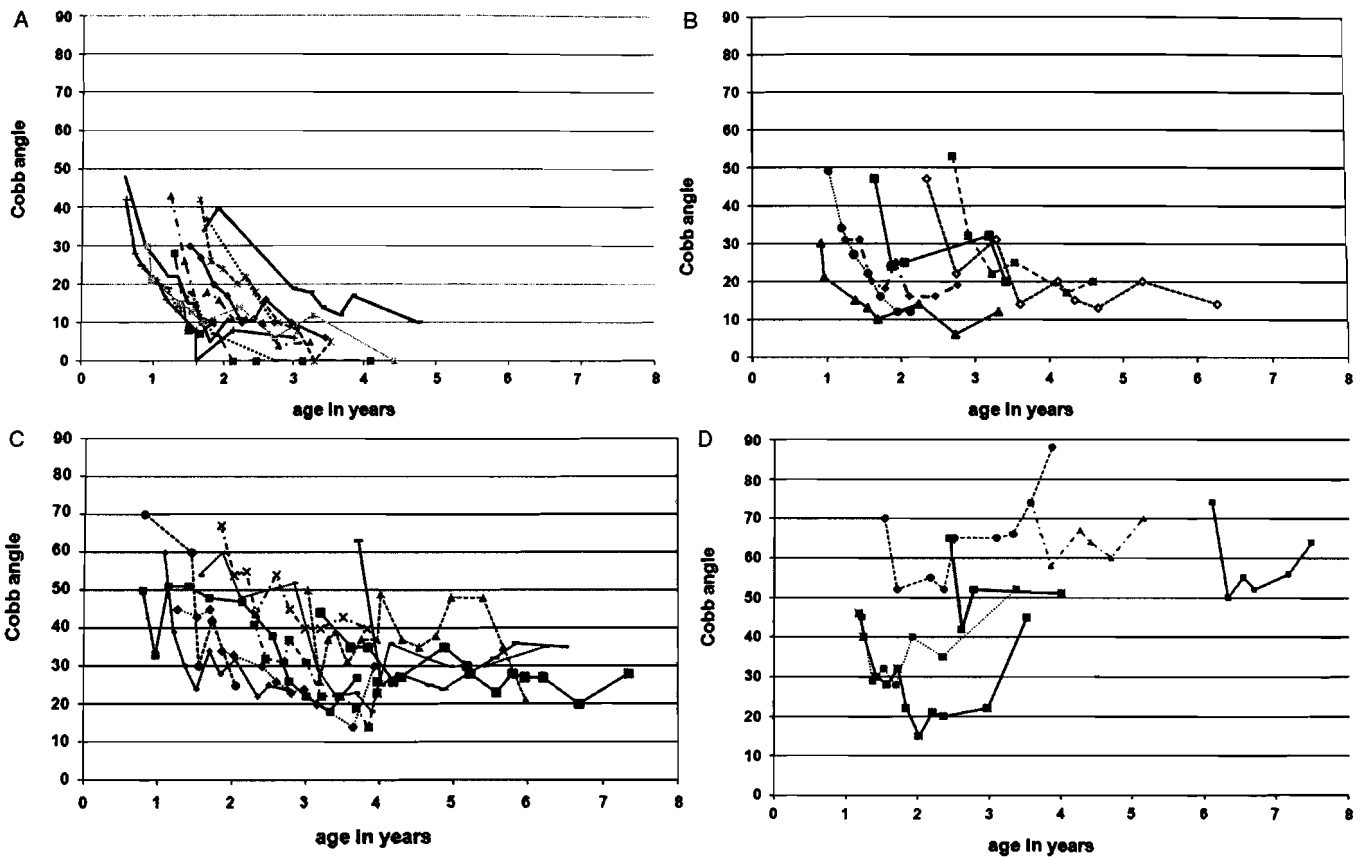
Our patients all exhibited progressive infantile scoliosis. Nearly all of the patients had some treatment before our cast correction. At the initiation of our casting programs, we were uncertain whether cast correction would prevent or delay the need for surgical intervention. The surgical indications were worsening curve magnitude or informed decision making by the parents. We do expect those who did not fully correct to ultimately have definitive deformity surgery though with most of them avoiding or delaying growing spine instrumentation. Once the diagnosis of progressive scoliosis is made, based on either a progressive Cobb angle or an RVAD of more than 20 degrees, rib phase 2, or a double curve, cast correction is recommended. We have typically required a magnetic resonance imaging

(MRI) of the spine before casting, but some centers initiate cast correction before the MRI. Mehta's program consists of cast changes under anesthesia in younger patients every 8 to 16 weeks until the curve is nearly resolved followed by an underarm brace that may be weaned if the patient's curve correction continues.<sup>4</sup> We have followed this protocol. We base our cast changes on the child's growth rate with changes every 2 months for those aged 2 years and below, 3 months for those age 3 years, and 4 months for those age 4 years and above. We aim for curves less than 10 degrees supine out of the cast and then use a brace molded just like the cast under anesthesia. Children are occasionally braced during the summer months with resumption of casting in the fall, particularly for children with larger curves in whom it is obvious that the casting will not be the definitive treatment. Whether these results will deteriorate or improve with further growth is unknown.

Mehta<sup>4</sup> identified 4 physiologic patterns: a "sturdy phenotype" with good muscle mass and tone, a "slender phenotype" with more delicate features, ligamentous laxity, and more rapidly progressive curves, those with known syndromes, and those with unknown syndromes.

TABLE 1. Relationship of the Final Curve to the Initial Curve for the Erie Patients

Follow-up Curve	Median Age in Years at Initial Casting	Average Curve at Cast Initiation	Average RVAD at Cast Initiation	Average Nash at Cast Initiation	No. Patients	Idiopathic Compared to Total Number	Average Follow-up in Years
10 or less	1.25	35	25	1.6	12	9/12—75%	2.1
11 to 20	1.78	45	21	2.5	5	3/5—60%	1.9
21 to 40	2.45	53	29	2.5	11	3/11—73%	2.3
> 40	2.69	59	40	2.3	6	3/6—50%	2.1
For the entire group of patients	1.97	46	28	2.2	34	23/34—68%	2.1



**FIGURE 4.** A, This graph shows the fairly rapid downward trend that occurred in these patients who had resolution of their curves to 10 degrees or less at follow-up. B, Similar to Figure 4A, this shows the downward trend of curves ending between 11 and 20 degrees during the follow-up period. These patients are slightly older and the drop in curve magnitude not as complete. C, This graph shows the course of those with curves 21 to 40 degrees at follow-up. The patients are again slightly older, have generally larger curves, and a slower and asymptotic response. D, This graph shows those whose curves were more than 40 degrees at follow-up. Although there was an initial response, these patient's curves tended to increase again after 1 to 2 years.

In her series, all groups responded if treatment was initiated early with smaller curves. In older patients with larger curves, the prognosis worsened from the sturdy to the slender to named and unnamed syndromes. Although we agree in principle with her classification, we have not found her classification of slender and sturdy reproducible in our hands and have divided the patients into the simple categories of idiopathic or syndromic. Those with abnormal MRIs were classified along with the syndromic.

Our results, similar to those of Mehta,<sup>4</sup> found the age at treatment onset a very important and significant factor in the success of the treatment. Mehta found cast correction much more likely to be successful if started under the age of 2 years. Our results are nearly identical with the patients achieving nearly full correction starting at average age of 1.1 years and full correction rare in those started over the age of 18 months. Cast correction to resolution typically takes 1 year or more. Curves with more rotation reflected in either the RVAD or Nash-Moe are less responsive than those with lesser rotation. We have also had less success in double than in single curves. Older and syndromic patients also had less correction

though some of the younger syndromic patients did respond as also noted by Mehta. Our goal in the older and syndromic patients is to delay the need for surgery until the spine has achieved sufficient growth for good pulmonary function as an adult. This endpoint is not well defined, but the hope is to delay fusion until the adolescent growth spurt. Both the VEPTR and growing rods are alternatives in this group of patients though we believe proper cast correction creates less spine or chest wall stiffness while still allowing spinal growth, though we lack comparison studies. The results of the centers in this study, with strong similarities in the principles, are nearly identical despite minor variations in the technique. The response of the smaller curves in younger children with less rotation emphasizes the importance of seeing and initiating treatment early rather than later after bracing has failed.

The argument can be made that bracing can afford the same corrective maneuvers as casting. Although this is theoretically true, the convenience of being able to remove a brace is also its difficulty. The lack of continuous corrective force and the difficulty in making sure the brace is donned in an ideal position each time do

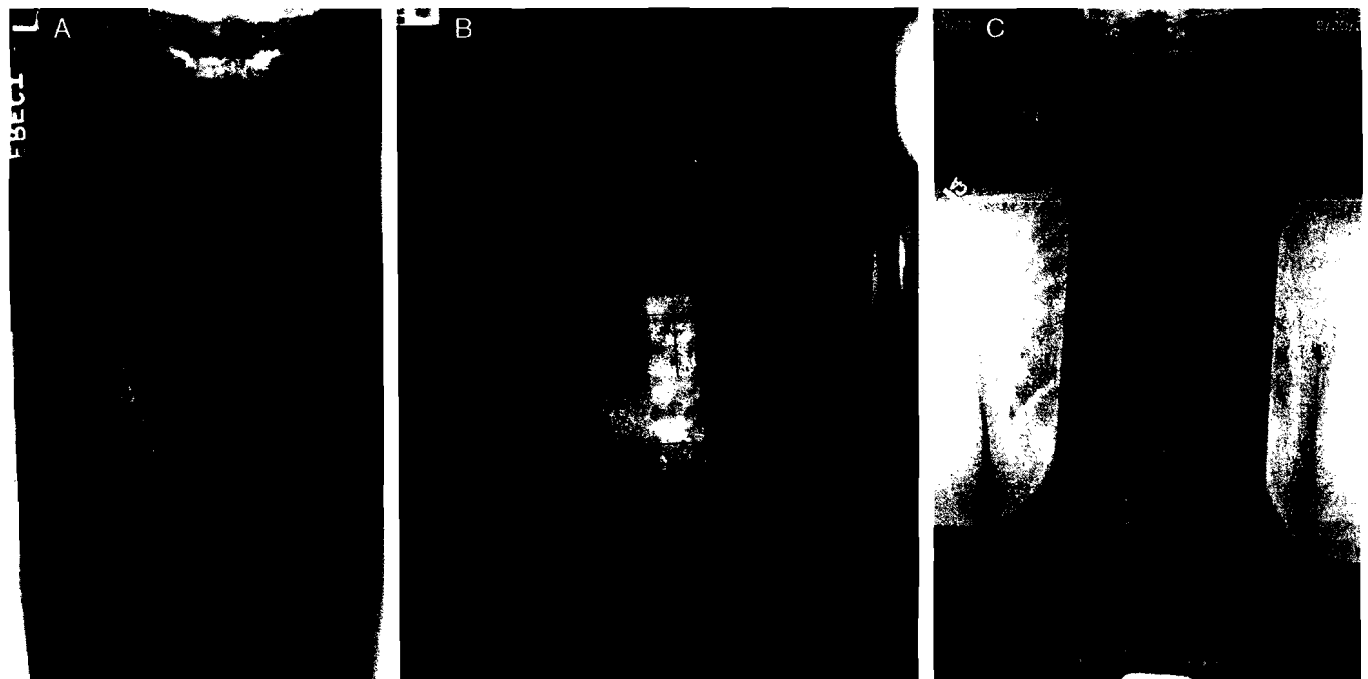
**TABLE 2.** Combined Data of the Centers

Follow-up Cobb Angle	No. Patients	Average Age at Start of Casting (y)	Average Cobb Angle at Start of Casting	Average RVAD at Start of Casting	Average Nash at Start of Casting	Etiology
10 or less	17	1.1	37	26	1.9	Fifteen idiopathic One possible tethered cord One minor brain MRI abnormality
11 to 21	10	2.2	41	20	2.2	Eight idiopathic One tethered cord One possible syndrome
21 to 40	13	2.6	53	33	2.4	Seven idiopathic Six with syndromes
> 40	15	3.1	71	37	2.6	Seven idiopathic Eight with syndromes
Cobb worsened	6	2.1	71	48	2.2	One idiopathic Five with syndromes
Total group	55	2.2	51	30	2.3	Thirty-eight idiopathic Fifteen with syndromes Two tethered cords One minor brain MRI abnormality

The relationship of the final curve to the initial curve and patient characteristics for the total combined series. The last row describes those patients whose curves actually worsened during the study period and are a subcategory of the row above it for final Cobb angles above 40 degrees and is not an additional group of patients. MRI indicates magnetic resonance imaging; RVAD, rib vertebral angle difference.

not afford the same ideal correction as the cast. In addition, with the pelvis and shoulders stabilized during the cast correction procedure, we believe that a true and effective derotational force can be applied to the primary spinal curvature and chest wall. Most children tolerate the casting far better than people expect and many parents indicate that their children are more comfortable in the cast compared with a brace.

The larger question is whether these patients are better served by serial cast correction, which requires a commitment and understanding on the family's part, or with growing rods or VEPTRs. The distinct advantages of the cast include the avoidance of the issues of spine and chest wall stiffness, rod fractures, infections, and pullout, and the rare neurological complication. Ultimately, determining this will require comparative studies.



**FIGURE 5.** A, Radiographs of an 8-month-old girl with a precasting curve of 52 degrees, rib vertebral angle difference of 39 degrees and phase 2 rib vertebral overlap. B, Radiographs showing correction in the cast at 11 months of age. C, Radiographs showing curve nearly complete resolution continuing at the age of 5 years, 3 years after completion of the treatment. Whether this will undergo later progression remains unknown.

## CONCLUSIONS

Serial cast correction for infantile scoliosis may result in complete correction in young patients (< 2 y) with curves of less than 50 to 60 degrees at the initiation of treatment. Cast correction for older patients with larger curves or nonidiopathic diagnosis still results in curve improvement. The Cotrel technique of derotation cast correction<sup>7</sup> seems to play a role in the treatment of progressive infantile scoliosis with cures in young patients and reductions in curve size with a delay in surgery in older and syndromic patients.

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## REFERENCES

- Campbell RM Jr, Smith MD, Mayes TC, et al. The characteristics of thoracic insufficiency syndrome associated with fused ribs and congenital scoliosis. *J Bone Joint Surg Am.* 2003;85-A:399-408.
- Goldberg CJ, Gillic I, Connaughton O, et al. Respiratory function and cosmesis at maturity in infantile-onset scoliosis. *Spine.* 2003;28:2397-2406.
- Karol LA, Johnston C, Mladenov K, et al. Pulmonary function following early thoracic fusion in non-neuromuscular scoliosis. *J Bone Joint Surg Am.* 2008;90:1272-1281.
- Mehta MH. Growth as a corrective force in the early treatment of progressive infantile scoliosis. *J Bone Joint Surg Br.* 2005;87:1237-1247.
- Emans JB, Caubet JF, Ordonez CL, et al. The treatment of spine and chest wall deformities with fused ribs by expansion thoracostomy and insertion of vertical expandable prosthetic titanium rib: growth of thoracic spine and improvement of lung volumes. *Spine.* 2005;30:S58-S68.
- Risser JC. The application of body casts for the correction of scoliosis. *Instr Course Lect.* 1955;12:255-259.
- Cotrel Y, Morel G. The elongation-derotation-flexion technique in the correction of scoliosis. *Rev Chir Orthop Reparatrice Appar Mot.* 1964;50:59-75.
- Nash CL Jr, MOE JH. A study of vertebral rotation. *J Bone Joint Surg Am.* 1969;51:223-229.
- Campbell RM Jr, Adcox BM, Smith MD, et al. The effect of mid-thoracic VEPTR opening wedge thoracostomy on cervical tilt associated with congenital thoracic scoliosis in patients with thoracic insufficiency syndrome. *Spine.* 2007;32:2171-2177.
- Mehta MH. The rib-vertebral angle in the early diagnosis between resolving and progressive infantile idiopathic scoliosis. *J Bone Joint Surg (Br).* 1972;54:230-242.
- Ceballos T, Ferrer-Torrelles M, Castillo F, et al. Prognosis in infantile idiopathic scoliosis. *J Bone Joint Surg Am.* 1980;62:863-875.
- Ferreira JH, Janeiro RD, James JI. Progressive and resolving infantile idiopathic scoliosis. The differential diagnosis. *J Bone Joint Surg Br.* 1972;54:648-655.
- Kristmundsdottir F, Burwell RG, James JI. The rib-vertebra angles on the convexity and concavity of the spinal curve in infantile idiopathic scoliosis. *Clin Orthop.* 1985;201:205-209.
- Thompson SK, Bentley G. Prognosis in infantile idiopathic scoliosis. *J Bone Joint Surg Br.* 1980;62-B:151-154.
- Pehrsson K, Larsson S, Oden A, et al. Long-term follow-up of patients with untreated scoliosis. A study of mortality, causes of death, and symptoms. *Spine.* 1992;17:1091-1096.
- Vitale MG, Matsumoto H, Roye DP Jr, et al. Health-related quality of life in children with thoracic insufficiency syndrome. *J Pediatr Orthop.* 2008;28:239-243.
- Thompson GH, Akbarnia BA, Campbell RM Jr. Growing rod techniques in early-onset scoliosis. *J Pediatr Orthop.* 2007;27:354-361.
- Thompson GH, Lenke LG, Akbarnia BA, et al. Early onset scoliosis: future directions. *J Bone Joint Surg Am.* 2007;89(suppl 1):163-166.