Early-Onset Scoliosis: A Review of History, Current Treatment, and Future Directions
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Early-onset scoliosis (EOS) is defined as curvature of the spine in children >10° with onset before age 10 years. Young children with EOS are at risk for impaired pulmonary function because of the high risk of progressive spinal deformity and thoracic constraints during a critical time of lung development. The treatment of EOS is very challenging because the population is inhomogeneous, often medically complex, and often needs multiple surgeries. In the past, early spinal fusion was performed in children with severe progressive EOS, which corrected scoliosis but limited spine and thoracic growth and resulted in poor pulmonary outcomes.

The current goal in treatment of EOS is to maximize growth of the spine and thorax by controlling the spinal deformity, with the aim of promoting normal lung development and pulmonary function. Bracing and casting may improve on the natural history of progression of spinal deformity and are often used to delay surgical intervention or in some cases obviate surgery. Recent advances in surgical implants and techniques have led to the development of growth-friendly implants, which have replaced early spine fusion as the surgical treatment of choice. Treatment with growth-friendly implants usually requires multiple surgeries and is associated with frequent complications. However, growth-friendly spine surgery has been shown to correct spinal deformity while allowing growth of the spine and subsequently lung growth.
occurs in children \( \leq 3 \) years of age and has a variable course over time. A unique feature of idiopathic EOS in infants is that it often improves spontaneously.\(^5\)\(^6\) Idiopathic EOS in juveniles occurs in children aged 4 to 10 years. Among children with neuromuscular disorders, scoliosis is common and compounds the restrictive lung disease produced by respiratory muscle weakness. Treatment strategies and duration differ significantly based on both etiology and the amount of anticipated growth remaining. The younger the child, the greater the risk that the spinal deformity will affect pulmonary development and function.

**GROWTH OF THE SPINE AND LUNG DEVELOPMENT**

The spine grows most rapidly in the first 5 years, with an average T1 to S1 segment length increase of 10 cm during this time (2 cm/year). After the first 5 years, there is a slower T1 to S1 growth from age 5 to 10 years of \( \sim 5 \) cm until adolescence (1 cm/year). From age 10 years to adulthood, T1 to S1 grows an additional 10 cm; this includes the adolescent growth spurt (2 cm/year).\(^7\)\(^8\)\(^9\) Because growth can promote the progression of the deformity, patients with EOS are at greatest risk for progression of spinal deformity in the first few years of life and during the adolescent growth spurt.

In EOS, the progressive spinal deformity occurs during a critical time of lung development. The number of alveoli and lung volume increase most rapidly in the first several years and continue to increase at a lower rate during adolescence and adulthood (Fig 2).\(^10\)\(^11\)\(^12\) Animal models of EOS produced early in life demonstrate alveolar simplification and reduced number, producing an example of postnatal hypoplasia.\(^13\) Autopsies of children dying of EOS report similar alveolar features and pulmonary vascular remodeling associated with pulmonary hypertension.\(^14\) Lung function studies of children with EOS demonstrate a variable severity of restrictive lung disease caused by small lung volumes, reduced chest wall compliance, and respiratory muscle dysfunction.\(^15\) The concept of thoracic insufficiency syndrome (TIS), popularized by Campbell et al,\(^16\) is defined as the inability of the thorax to support normal respiratory function and lung development in growing children. Because of scoliosis progression early in life, patients with severe EOS can potentially develop TIS. TIS has been associated with poorer quality of life scores than those of childhood epilepsy, heart disease, and cancer.\(^17\)

The natural history of untreated EOS is associated with significant morbidity and often profound cardiopulmonary compromise, including respiratory failure and cor pulmonale. A Swedish study comparing expected population death rates demonstrated more than twice the mortality rate by age 40 in patients with EOS compared with that of the general population.\(^18\) Consequently, the fundamental principle of treating EOS is to foster normal respiratory development and maximize spinal growth while preventing additional deformity that can lead to TIS.

**HISTORY OF TREATMENT STRATEGIES: WHAT WE HAVE LEARNED**

Before the introduction of spinal implants, the historical treatment of EOS consisted of casting or bracing the spine and thorax. Although casting for scoliosis has been performed for centuries, it fell out of favor as the primary treatment of scoliosis because of concerns that casting deformed the ribs. Paul Harrington introduced a spinal implant for scoliosis in the 1960s that provided a surgical alternative. In these cases, the rod was used to...
surgically correct scoliosis without fusion by applying distraction across the concavity of the curve. Harrington rods improved curves in a 2-dimensional plane, although this technique often led to a flat back deformity. Implant failure and dislodgment with this method were high, and its use was limited.

Early spinal fusion to halt deformity in EOS then became the preferred treatment, because a short and straight spine was thought to be superior to a progressively crooked spine. Subsequent studies demonstrated that early spinal fusion, which prevents continued spinal growth of the fused region, limits intrathoracic volume and hence lung volume. As a result, children developed severe restrictive lung disease with continued growth. Patients with idiopathic EOS who underwent spinal fusion at a mean age of 4.1 years demonstrated mean forced vital capacity (FVC) of 41% of normal when evaluated at skeletal maturity, whereas patients who underwent fusion at a mean age of 12.9 years demonstrated mean FVC of 68% of normal. In 1 study, the reduction in FVC ≥5 years after spine fusion directly correlated with the number of thoracic spinal segments fused. Early posterior spinal fusion techniques have often led to the crankshaft phenomenon, in which the anterior column of the immature spine continues to grow, leading to progressive deformity. Consequently, new treatment strategies have been developed that allow or promote spinal growth, usually referred to as growth-friendly techniques.

At worst these techniques may be thought of as delaying the need for a spine fusion to allow spinal growth, and at best they may cure the scoliosis or avoid the need for a spinal fusion. One exception to avoiding early spinal fusion is congenital scoliosis in which the spine deformity is limited to a small number of vertebrae. The classic example of congenital scoliosis that requires early fusion is the case of a hemivertebra causing progressive scoliosis (Fig 3). In such cases early fusion (with or without excision of the hemivertebra) can often correct the scoliosis in 1 surgery, with a fusion of only 2 vertebrae. This procedure is generally performed around the age of 3 to 6 years.

**CURRENT TREATMENT STRATEGIES: NONOPERATIVE**

Nonoperative treatment of EOS consists of bracing or casting. Bracing can be considered for mild progressive curves, although its efficacy remains unproven in EOS and ensuring compliance with brace wear can be difficult in young children. Commonly used braces are variants of a custom-molded thoraolumbosacral orthosis. Braces are often used to maintain correction obtained from serial casting or delay surgical intervention. Several case series have shown that 74% to 92% of idiopathic EOS in infants spontaneously resolves. For idiopathic EOS in infants diagnosed <3 years, many milder curves will resolve; however, some will require serial casting. For more severe curves, a brace may be necessary to maintain correction. For juvenile idiopathic scoliosis, surgery may be necessary despite prolonged casting or brace wear.

### TABLE 1 Summary of the Different Types of EOS and Their Unique Features

<table>
<thead>
<tr>
<th>Types of EOS</th>
<th>Characteristics</th>
<th>Associated Diagnoses</th>
<th>Treatment Considerations</th>
</tr>
</thead>
<tbody>
<tr>
<td>Congenital</td>
<td>Structural abnormality of the spine or thorax present at birth.</td>
<td>Cardiac, renal abnormalities</td>
<td>Hemivertebrae excision. Short-segment early spinal fusion in select cases in this group may be the exception to growth-friendly spine surgery.</td>
</tr>
<tr>
<td></td>
<td>Failure of formation (eg, hemivertebra).</td>
<td>Other musculoskeletal abnormalities (upper limb, club foot)</td>
<td></td>
</tr>
<tr>
<td></td>
<td>Failure of segmentation (eg, fused vertebra or ribs).</td>
<td>Associated with VATER/VACTERL syndromes</td>
<td></td>
</tr>
<tr>
<td></td>
<td></td>
<td>Intraspinal abnormalities (ie, diastematomyelia, tethered cord)</td>
<td></td>
</tr>
<tr>
<td>Neuromuscular</td>
<td>Abnormalities in muscular tone lead to scoliosis.</td>
<td>Examples: Cerebral palsy, muscular dystrophies, myopathies, spinal cord injuries</td>
<td>Generally higher-risk surgical patients with medical comorbidities (eg, respiratory, gastrointestinal).</td>
</tr>
<tr>
<td>Syndromic</td>
<td>Includes any other syndrome associated scoliosis (excluding neuromuscular or congenital scoliosis syndromes).</td>
<td>Examples: Connective tissue disorders, Marfan syndrome, neurofibromatosis, skeletal dysplasias, Prader–Willi syndrome</td>
<td>Each syndrome has unique considerations (eg, neurofibromatosis may have dural ectasia, making spinal implants more challenging to place. Generally healthier children.</td>
</tr>
<tr>
<td>Idiopathic</td>
<td>Scoliosis without a known attributable cause.</td>
<td>Higher incidence of Arnold–Chiari malformation and syringomyelia compared with adolescent idiopathic scoliosis</td>
<td>Casting shown to resolve some infantile curves.</td>
</tr>
<tr>
<td></td>
<td>Infantile (diagnosed &lt;3 y): Many milder curves will resolve but need observation.</td>
<td></td>
<td>Surgery often needed despite often long-term casting or brace wear.</td>
</tr>
</tbody>
</table>

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infants with unresolved progressive curves, Mehta demonstrated that casting may be effective in completely resolving some curves, especially those of lower magnitude. Because casting has proven to be a safe method to manage idiopathic EOS, there has been a resurgence of interest in expanding traditional cast methods to treat multiple subtypes of EOS to avoid the risks of surgery and early spine fusion (Fig 4). The cast is applied to the torso under anesthesia while the child is in traction, elongating the spine. The cast is molded while the child’s torso is derotated and flexed away from the concavity of the curve. Casts are changed every 8 to 12 weeks. For curves that resolved or stabilized in a cast, bracing is often used to help maintain the correction through skeletal maturity. If there is progression despite bracing, additional casting may be used to attempt to regain the correction. In other cases, growth-friendly implants or fusion may be the most appropriate step depending on the patient’s age and curve severity. Serial casting applied to young children with nonidiopathic EOS has been shown to be an effective way to delay surgical treatment. In 1 study, curve resolution was rare with serial casting in the nonidiopathic EOS, but progression of the curve was controlled sufficiently to delay spine surgery for at least 2 years. Normal longitudinal growth of the spine was observed while the patient was in the cast. Based on current evidence, a trial of casting in EOS regardless of curve etiology is considered a treatment option. The specific indications for the threshold to institute cast treatment continue to vary between institutions but are generally considered for EOS curves >25°, with >10° of documented progression.

**CURRENT TREATMENT STRATEGIES: SURGICAL**

The surgical treatment strategy for EOS has evolved significantly over the past decade with the use of modern growth-friendly implants. These implants attempt to maximize the growth of the spine and thorax while controlling curve progression to preserve normal lung volume. Growth-friendly implants can be classified into 3 distinct subtypes: distraction-based, guided growth, and compression-based strategies. Distraction-Based Implants

Distraction-based implants are the most common devices used in EOS. They apply traction to the spinal column between proximal and distal anchors joined by expandable rods. The rods are periodically lengthened as the child grows to maintain spine curve correction. Four types of implants have been used: the traditional growing rod (TGR), vertical expandable prosthetic titanium rib (VEPTR) device, hybrid systems, and magnetically controlled growing rod (MCGR).

**Growing Rod**

The TGR incorporates proximal and distal hook or screw anchors on the spine, joined by rods with connectors that allow serial distractions between the rods (Fig 5). Limited fusion is performed at the proximal and distal anchor sites on the spine to provide
solid sites for spine distraction. The area between the anchors is intentionally not fused, allowing motion and growth through this region. Lengthenings are typically performed at ~6-month intervals. Akbarnia et al\(^29\) reported on the use of TGRs for EOS in 24 patients, resulting in improvement of coronal plane major curves from 82° to 36°, with 1.2 cm growth in T1 to S1 length per year at mean 4-year follow-up. Akbarnia et al\(^30\) also demonstrated that patients whose spines were lengthened at \(\leq 6\)-month intervals had significantly higher annual T1 to S1 growth rate of 1.8 cm/year, compared with 1.0 cm/year in patients whose spines were lengthened less frequently. This finding has led many to believe that distraction may actually promote vertebral growth.

**VEPTR**

Developed by Bob Campbell, VEPTRs use ribs as anchors, and sometimes the spine and pelvis as well. VEPTRs are generally thought of as primarily providing thoracic expansion, compared with growing rods, which primarily provide control of scoliosis. In reality, thoracic and spinal deformities are closely linked. Similar to other distraction-based systems, these constructs undergo recurrent surgical expansion (Fig 6). Original descriptions of the VEPTR technique recommended incising between ribs to maximize thoracic expansion, but concerns of scarring and stiffening of the chest wall led most surgeons to cut between the ribs only in cases of multiple fused ribs. VEPTR treatment has demonstrated continued spinal growth with serial expansions (mean 71 mm over 4 lengthenings) while improving the coronal curve.\(^31,32\)

**Hybrid**

A hybrid distraction-based strategy incorporates the VEPTR concept of using ribs as anchor sites but also uses traditional spinal implants, as in the TGR system. Traditional spinal hooks are placed proximally along the ribs (Fig 7). As in the TGR strategy, the distal anchor site is incorporated by a fusion.
and lengthening is performed at a connector between the rods. The advantage of this technique is that it avoids fusion of the proximal anchor site of the thoracic spine, potentially allowing more total growth of the thorax. Furthermore, rib anchors without traditional rigid fusion at the proximal anchor site allow some motion of the spinal implant construct. This feature may reduce the stress and rigidity of the distraction system across an unfused mobile spine. Consequently, use of a hybrid system with proximal rib anchors has been associated with a decrease in the incidence of rod breakage.\textsuperscript{33}

**MCGR**

Recently the US Food and Drug Administration cleared the use of MCGRs, which can lengthen nonsurgically without anesthesia after the initial implantation.\textsuperscript{34,35} This implant is similar to other growing rod constructs with distal spine anchors and proximal rib or spine anchors that are connected by telescoping rods. This telescoping portion contains an internal magnet that can be lengthened from an external remote control (Fig 8). Because of the noninvasive nature of lengthening, it is also possible to distract at shorter intervals. Preliminary studies of MCGRs have been met with optimism with respect to achieving the same results of TGRs.\textsuperscript{34–36} Dannawi et al\textsuperscript{34} demonstrated that the mean coronal Cobb angle improved from \(69^\circ\) to \(41^\circ\) after MCGRs with a mean of 4.8 distractions per patient were used over 15 months. The T1 to S1 length increased a mean of 3.5 cm during this time period. A study comparing 12 MCGR- and TGR-treated patients demonstrated no significant difference in spine length gains, but there were 57 fewer surgical procedures in the MCGR group.\textsuperscript{37} Although MCGR avoids the need for repeat surgical intervention for routine lengthenings, long-term data are not yet available for this technique. Similar to growth-friendly implants, the complication rate is high: 33% of patients treated with MCGR within 2 years of follow-up.\textsuperscript{37}

**FIGURE 5**

Preoperative (A) and postoperative (B) radiographs of patient with traditional spine-to-spine growing rods. Radiographs obtained 5 years after the initial placement of growing rods (C) show that the scoliosis continues to be well controlled. Reproduced with permission of Children’s Orthopedic Center, Los Angeles, California.

**FIGURE 6**

Postoperative radiograph of an 8-year-old boy with VATER syndrome with congenital scoliosis, multiple rib fusions, and thoracic insufficiency syndrome that was treated with a VEPTR construct. Reproduced with permission of Children’s Orthopedic Center, Los Angeles, California.

**FIGURE 7**

A, Preoperative posteroanterior and lateral radiograph of a 4-year-old boy with severe progressive scoliosis and an 85° curve. He was not a casting candidate because of his restrictive lung disease. B, Postoperative radiograph showing a hybrid growing rod construct (rib to spine) with improvement to 37°. Reproduced with permission of Children’s Orthopedic Center, Los Angeles, California.
Complications of Distraction-Based Implants

Regardless of the implant used for distraction-based treatment of the growing spine, all strategies are associated with a high complication rate.\textsuperscript{29,30,38–43} Implant complications such as anchor malfunction (pullout from the spine, erosion through the rib) and rod breakage are common (Fig 9). Distraction-based posterior implants often produce kyphosis, which may result in an unfavorable overall sagittal plane balance. Wound complications are also common because of the prominence of implants under the skin and poor healing potential in small, thin, and often chronically malnourished patients with EOS (Fig 10). At least 1 complication of treatment has been reported to occur in 58\% to 86\% of patients undergoing distraction-based treatment, leading to multiple unplanned surgical procedures.\textsuperscript{38,43,44}

Among patients with complications, 1 study demonstrated a mean of 2.2 complications per patient.\textsuperscript{38} Application of stiff implants on an unfused spine that continues to have motion ultimately leads to fatigue failure of the implants. Risk factors for implant failures include severe thoracic kyphosis that produces proximal anchor pullout and increased number of lengthening procedures.\textsuperscript{40} Implantation strategies are being critically evaluated to decrease the incidence of implant-related complications. A comparison study of complications in TGRs, hybrid proximal rib anchor systems, and VEPTR treatments in EOS demonstrated a trend toward decreased implant-related complications in the hybrid system.\textsuperscript{43} Hooks are not as rigidly fixed to the spine compared with screws, theoretically allowing some motion and dispersion of stress, decreasing fatigue-related implant failures. Yamaguchi et al\textsuperscript{33} demonstrated 6\% rod breakage in proximal rib-anchored growing rods, compared with 29\% in proximal spine-anchored growing rods at a mean 56-month follow-up.

The multiple surgeries needed for treatment with distraction-based implants are associated with adverse outcomes. Each lengthening surgery has been shown to increase the risk of deep infection 3.3 times in EOS.\textsuperscript{41} The length gained from serial lengthening has also been shown to follow a law of diminishing returns, with decreased spinal length gained after each lengthening because of increased stiffness of the spine.\textsuperscript{45,46} Autofusion of the spine has been also described after repeated lengthenings.\textsuperscript{47} Ultimately, the utility of lengthening may be minimal after the sixth or seventh lengthening procedure, limiting the potential spinal growth to 4 to 5 years after initial surgery.

In addition to the physical effects on the spine, there are significant psychological effects from distraction-based treatment. Patients with repeated surgery in EOS demonstrate abnormal psychosocial scores, with a positive correlation between behavioral problems and the number of repetitive surgeries.\textsuperscript{42} Repeated general anesthesia in children may cause detrimental neurocognitive effects, based on animal and preclinical studies, although this remains an area of controversy.\textsuperscript{48–50} New technologies, such as MCGRs that obviate surgical lengthenings, will probably help minimize the total number of exposures to anesthesia in EOS.

Guided Growth Implants

In guided growth techniques, the spine is straightened with spinal implants that allow the vertebrae to grow along the path of the spinal implants. The original guided growth system used Luque wires wrapped around the lamina of each vertebrae, which were wrapped around straight rods that corrected scoliosis and then permitted guided growth as the wires slid along the rods. This technique was found to lead to spontaneous fusion and limited spinal growth. A more recent type of guided growth implant, called the Shilla technique, has been developed by Richard McCarthy. In this technique, screws are placed into vertebrae with minimal dissection in the hopes of avoiding spontaneous fusion and allowing 3-dimensional correction of

\section*{FIGURE 8}

A, Preoperative anteroposterior radiograph of a 7-year-old girl with spinal muscular atrophy and 100\textdegree{} thoracolumbar curve. B, Postoperative posteroanterior radiograph demonstrating a construct including the MCGR device. C, Model demonstrating an MCGR device. Reproduced with permission of Children’s Orthopedic Center, Los Angeles, California.
the spinal deformity and permitting growth along the rods (Fig 11). The major advantage of guided growth techniques over growing rods is that children avoid multiple surgical lengthenings. A recent study comparing the Shilla technique with growing rods demonstrated that patients treated with the Shilla technique had fewer surgeries (2.8) compared with growing rods (7.4) in >4-year mean follow-up. Shilla resulted in less spinal growth and less correction of scoliosis, with similar complication rates to growing rods.51

Compression-Based Implants

Compression-based implants involve correcting scoliosis by stopping the growth of the convex side of the scoliosis without fusion while allowing growth of the concave side of the curve. This correction is accomplished by placing staples, tethers, or other devices across the growth plates of the vertebrae from an anterior approach on the convex side of the scoliosis. Although there is 1 device approved by the US Food and Drug Administration, tethers and staples are most commonly used off label. Several case series on compression-based implants have demonstrated curve correction with growth in patients who underwent surgery at age <10 years.52,53 There have been cases of overcorrection with compression-based implants, in which the curve corrects and then develops in the opposite direction. Therefore, this technique is generally reserved for patients with limited growth remaining, such as 9- to 10-year-olds. Additional concerns include the potential pulmonary impact of ≥1 transthoracic surgeries. Serial measures of lung function in older children with scoliosis treated with transthoracic spine surgery have shown greater loss of function postoperatively when the thorax is opened.54

PULMONARY OUTCOMES OF RECENT EOS TREATMENT

The study of pulmonary function in this patient population is extremely challenging, complicated by the fact that many of these patients start treatment of EOS before they are old enough to undergo formal pulmonary function tests (PFTs). Much of the current literature evaluating the pulmonary outcomes after growth-friendly spine surgery has been based on VEPTR in children >6 years old. PFTs have been measured in both awake and anesthetized patients. Computed tomography has also been used to measure thoracic volumes as a surrogate for PFTs. Studies of lung function in EOS have not compared treatment strategies or different devices and are often small, descriptive case series. In 1 such series, 10 children with EOS (median age 4.3 years) demonstrated increased mean annual absolute FVC of 27% of predicted norms and maintenance of FVC as a percentage of normal after VEPTR treatment at a mean 22 months of follow-up.55 With longer periods of follow-up (mean 6 years) during and at the completion of growing construct surgery, FVC as a percentage of normal declined by an average of 28%.56,57 There are no untreated control groups to assess what loss of lung function might have occurred in the natural progression of the spine deformities. The implication of these studies is that lung function is not normalized or predictably improved after treatments for EOS but that progressive loss of lung function may be reduced with treatment.

Surrogate pulmonary outcomes that do not require voluntary effort by young children have also been reported. Several studies used weight gain as an indirect marker of improved pulmonary function and found that up to 50% of patients with EOS demonstrated a mean 24- to 26-percentage-point improvement after VEPTR or growing rod treatment.58,59 Overnight polysomnography in children with EOS demonstrates an increased Apnea–Hypopnea Index.

FIGURE 9
Radiograph demonstrating a broken rod with traditional spine-to-spine growing rods, identified by the red arrow. Reproduced with permission of Children’s Orthopedic Center, Los Angeles, California.

FIGURE 10
Clinical photo showing marked hardware prominence in a thin child with EOS. Reproduced with permission of Children’s Orthopedic Center, Los Angeles, California.
and hypoxemia associated with hypopneic events. Serial measures of breathing during sleep before and after treatment of EOS may also prove useful as an indirect measure of lung function.

Two-dimensional images of the spine, such as the Cobb angle, do not correlate with lung function measures and do not reflect changes in lung function after spine curvature has been reduced. However, an encouraging study recently demonstrated that radiographic T1 to T12 height and T1 to S1 height modestly correlate with improved pulmonary function in EOS. New imaging modalities, such as diaphragm and thoracic excursion, measured by dynamic MRI, hold some promise in improving assessment of spine structure–respiratory function relations. Persistent barriers to a high-quality literature on the topic include a lack of control groups, because untreated progressive EOS is known to have a poor outcome, and a lack of standardization and difficulty in evaluating respiratory function in young children.

CURRENT TREATMENT RECOMMENDATIONS

Management of EOS involves a diverse patient population, variable spinal and thoracic deformities, and multiple treatment options (Table 2). Optimizing the treatment of each child is a process in evolution. In many cases, a trial of serial casting can help control the scoliosis and allow growth while delaying surgery. In some idiopathic cases, these curves may resolve with casting alone. Many children may not be able to tolerate casting or demonstrate progression of the scoliosis despite casting necessitating the initiation of growth-friendly spinal surgery. There is considerable variation with regard to the optimal timing and indication of surgery. A recent survey of 14 pediatric spine surgeons found that the majority considered curves with recent progression and a magnitude of ≥60° an indication for distraction-based implants. The indications for any of the distraction-based implants are similar, although VEPTR has potential advantages in cases that require direct expansion of the thorax, such as cases of thoracic dystrophy. Compression-based therapies need more data in the treatment of EOS, although they appear to be an option in children.

![FIGURE 11](image)

A, Preoperative posteroanterior radiograph of a 7-year-old boy with a 61° curve. B, Initial postoperative radiograph shows correction with a Shilla construct with a fusion at the apex (red arrows) and screws at the upper and lower anchors that can slide along the rod (yellow arrows). C, Images 2.5 years later show how the child’s growth has been guided by the rods, as demonstrated by the shorter distance beyond the anchors that the rods extend (blue arrow). Reproduced with permission of Children’s Orthopedic Center, Los Angeles, California.

<table>
<thead>
<tr>
<th>Treatment</th>
<th>Pros</th>
<th>Cons</th>
</tr>
</thead>
<tbody>
<tr>
<td>Bracing</td>
<td>Helps delay need for surgery in very young patients.</td>
<td>Standard thoracolumbosacral orthosis brace cannot control curves with apex above midthoracic spine (around T7). Bracing wear compliance may be difficult.</td>
</tr>
<tr>
<td></td>
<td>Helps for idiopathic EOS curves in juvenile patients near adolescent age.</td>
<td>Not much literature about bracing in EOS. Some children may not tolerate full-time body cast well.</td>
</tr>
<tr>
<td>Casting</td>
<td>Maximizes spinal growth before surgery.</td>
<td>Is not a definitive treatment in most cases.</td>
</tr>
<tr>
<td></td>
<td>Some idiopathic curves may resolve.</td>
<td>Requires multiple periodic lengthening surgeries (exception: magnetically controlled rods).</td>
</tr>
<tr>
<td>Distraction</td>
<td>Effective method to correct curve and lengthen the spine before final spinal fusion.</td>
<td>High complication rates (e.g., implant failure, infection).</td>
</tr>
<tr>
<td></td>
<td>Holds the most clinical experience and literature in the surgical treatment of EOS.</td>
<td></td>
</tr>
<tr>
<td>Guided growth</td>
<td>Initial apical fusion procedure guides subsequent spinal growth.</td>
<td>Requires larger anatomy to allow instrumentation of apex (avoid in very small children).</td>
</tr>
<tr>
<td></td>
<td>No scheduled repeated lengthening procedures.</td>
<td></td>
</tr>
<tr>
<td>Compression</td>
<td>Fusionless procedure.</td>
<td>Limited data for use in EOS.</td>
</tr>
<tr>
<td></td>
<td>Requires a thoracic approach (potentially detrimental to pulmonary function).</td>
<td>Risk of overcorrection when used for young children.</td>
</tr>
</tbody>
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nearing adolescence, with less total growth remaining. After achieving maximal correction and growth with growth-friendly spinal implants, children can have their spine definitively fused at a later age if significant deformity remains.

**Future Directions in Early-onset Scoliosis**

The basic unanswered question is how much early and late treatment strategies for EOS maximize respiratory function when children reach maturity. This question remains difficult to study because it is unethical to have an untreated natural history comparison group in which the scoliosis is allowed to progress relentlessly. Collaboration between pediatric pulmonologists and orthopaedists is essential to standardize how pulmonary function evaluations are being performed for children who are not able to comply with traditional PFTs.

A better understanding of the 3-dimensional natural growth of the thorax and how it is affected by surgical treatment in EOS is crucial. Distraction-based implants help decrease the scoliosis, although how this result correlates with improved pulmonary function has not been established. Radiographic measurements in the 2-dimensional plane, such as Cobb angles, are not reliable predictors of severity of pulmonary disease. Three-dimensional understanding and functional imaging of the thorax warrant additional study to improve characterization of how the structure of the thorax relates to function to predict severity of pulmonary disease in EOS.

EOS incorporates a multitude of etiologies and associated diagnoses, and greater subclassification can help develop a framework for future study. Each etiology carries different implications, because congenital and idiopathic EOS may behave much differently with regard to the rate of curve progression. Williams et al developed a new classification scheme in EOS that may help establish the optimal treatment of each subtype of EOS. Multicenter groups such as the Growing Spine Study Group and the Chest Wall and Spine Deformity Study Group have been developed to collaborate in the study of this heterogeneous population of children.

**Abbreviations**

EOS: early-onset scoliosis

FVC: forced vital capacity

MCGR: magnetically controlled growing rod

PFT: pulmonary function test

TGR: traditional growing rod

TIS: thoracic insufficiency syndrome

VEPTR: vertical expandable prosthetic titanium rib

**References**


5. Lloyd-Roberts GC, Pilcher MF. Structural idiopathic scoliosis in infancy: a study of the natural history


32. Hasler CC, Mehrkens A, Hefti F. Efficacy and safety of VEPR instrumentation for progressive spine deformities in young children without rib fusions. Eur Spine J. 2010;19(3):400–408


37. Akbarnia BA, Pawelek JB, Cheung KMC, et al. Traditional growing rods versus magnetically controlled growing rods for the surgical treatment


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