Early Onset Scoliosis: To operate or to brace?

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Introduction

Fourth dimension with regards to scoliosis is potential for progression with growth.

Scoliosis: a review
Early-onset: below age of 3 years. Eighty to 90% resolve before age of 2 years. Ten to 20% develop extremely severe curves with associated affects on cardiac and respiratory function and death in early adult life.
Early-onset scoliosis is frequently associated with \textit{genetic disorders} but many with \textit{nonspecific} clinical features and without an associated diagnosis.
Cast treatment is a valuable delaying tactic for younger children with early-onset scoliosis.

**Comparison of Growing Rod Instrumentation Versus Serial Cast Treatment for Early-Onset Scoliosis**

*Spine Deformity, Volume 1, Issue 5, September 2013, Pages 339-342*

Charles E. Johnston, Anna M. McClung, George H. Thompson, Connie Poe-Kochert, James O. Sanders, Growing Spine Study Group
Cotrel technique of derotation casting seems to play a role in treatment of progressive infantile scoliosis.
Casting in Early-Onset Scoliosis


James O. Sanders, Charles E. Johnston, Jacques D'Astous
Asymmetry in lung function influenced by deformity of chest wall in multiple dimensions, and cannot be ascertained by chest radiographs alone.

Lung function asymmetry in children with congenital and infantile scoliosis
The Spine Journal, Volume 8, Issue 4, July–August 2008, Pages 639-644
Gregory Redding, Kit Song, Steve Inscore, Eric Effmann, Robert Campbell
Physical Significance of the Rib Vertebra Angle Difference and Its 3-Dimensional Counterpart in Early-Onset Scoliosis

*Spine Deformity, Volume 1, Issue 4, July 2013, Pages 259-265*

Genevieve Foley, Carl-Eric Aubin, Stefan Parent, Hubert Labelle, Jacques d' Astous, Charles Johnston, James Sanders

Rib vertebra angle difference not only indicates *asymmetry of ribs in relation to spine*, it is a compound of physical factors including *vertebral axial rotation.*
Evaluation of a Three-Dimensional Reconstruction Method of the Rib Cage of Mild Scoliotic Patients

Spine Deformity, Volume 1, Issue 5, September 2013, Pages 321-327

Aurélien Courvoisier, Brice Ilharreborde, Barbara Constantinou, Benjamin Aubert, Raphael Vialle, Wafa Skalli

Obtaining low-dose, weight-bearing, reliable, 3D morphological analyses of scoliosis of spine and chest simultaneously, with reasonable acquisition and processing times, is of particular interest, especially in children and adolescents.
Operative Strategies for Thoracic Insufficiency Syndrome by Vertical Expandable Prosthetic Titanium Rib Expansion Thoracoplasty

Operative Techniques in Orthopaedics, Volume 15, Issue 4, October 2005, Pages 315-325

Robert M. Campbell Jr

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### Operative Strategies

<table>
<thead>
<tr>
<th>Type of Volume Depletion Deformity</th>
<th>Thoracic Deficit</th>
<th>Mechanism of Lung Volume Loss</th>
<th>Examples</th>
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<tbody>
<tr>
<td>I. Absent ribs and exostotic scoliosis</td>
<td>Unilateral thoracic hypoplasia</td>
<td>Lung prolapses into the chest with volume loss.</td>
<td>VATER, absent ribs and congenital scoliosis</td>
</tr>
<tr>
<td>II. Fused ribs and exostotic scoliosis</td>
<td>Unilateral thoracic hypoplasia</td>
<td>Constriction of lung due to fused ribs shortening hemithorax</td>
<td>VATER, fused ribs and congenital scoliosis, thoracogenic scoliosis from prior thoracotomy</td>
</tr>
<tr>
<td>III. Foreshortened thorax</td>
<td>Global thoracic hypoplasia</td>
<td>Bilateral longitudinal constriction of lungs from loss of thoracic height</td>
<td>Jarcho-Levin syndrome</td>
</tr>
<tr>
<td>IV. Transversus constricted thorax</td>
<td>Global thoracic hypoplasia</td>
<td>Lateral constriction of lungs from rib deformity</td>
<td>Jeune’s asphyxiating thoracic dystrophy, wind-swept deformity of the thorax in scoliosis</td>
</tr>
</tbody>
</table>

### FDA Indications for VEPTR Expansion Thoracoplasty

1. Thoracic insufficiency in skeletally immature patients associated with:
   1. Fail chest syndrome
   2. Constrictive chest wall syndrome, including fused ribs and scoliosis
   3. Hypoplastic thorax
   4. Progressive scoliosis of congenital or neurogenic origin without rib anomaly

### Contraindications

1. Soft thoracic kyphosis greater than 50 degrees
2. Spinal cord syrinx, tethered spinal cord or other spinal cord abnormality
3. Inadequate soft tissue coverage for the VEPTR (usually body weight less than 25 percentile)
4. Absence of proximal ribs for attachment of the VEPTR
5. Inability to tolerate repetitive episodes of general anesthesia (usually due to pulmonary, cardiac disease, or other medical conditions)
6. Absence of diaphragm function
7. Active pulmonary infection

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[Diagrams and illustrations showing the procedures and tools used in the surgical techniques.]
Thoracic malformation with early-onset scoliosis: Effect of serial VEPTR expansion thoracoplasty on lung growth and function in children

*Paediatric Respiratory Reviews, Volume 10, Issue 1, March 2009, Pages 12-17* Etsuro K. Motoyama, Charles I. Yang, Vincent F. Deeney

Low surgical risks, clinical improvements and significant increases in lung volume. Better before 6 years of age.
Carbon Coated Implants as a New Solution for Metal Allergy in Early-Onset Scoliosis: A Case Report and Review of the Literature

Spine Deformity, Volume 2, Issue 1, January 2014, Pages 76-80

Julie Zielinski, Thomas A. Lacy, Jonathan Huw Phillips

An innovative carbon coating applied by plasma spray to VEPTR system, with good results.
An unusual case of congenital scoliosis associated with rib agenesis in the upper part of the concavity treated by VEPTR vertebra to vertebra


**Vertebra-to-vertebra construct**, a solution in treatment of early-onset scoliosis requiring surgery and associating **rib agenesis in** area of **proximal** module insertion.
Kyphosis: difficult to manage with repetitive distraction strategies, and some considered severe rigid kyphosis a contraindication to VEPTR.
Growing Rods

Early fusion results in negative long-term consequences.

Measuring Outcomes in Children with Early-Onset Scoliosis
Seminars in Spine Surgery,
Volume 24, Issue 3, September 2012, Pages 140-143
Jaime A. Gomez, Michael G. Vitale
Pulmonary function correlates with height of thoracic spine (T1–T12). **Growth of thoracic spine** to 18 Cm at age of 5 and 22 Cm at age of 10 minimizes risk of pulmonary complications.

10 years=10 cm again
Assessment of skeletal maturity in scoliosis patients to determine clinical management: a new classification scheme using distal radius and ulna radiographs

The Spine Journal, Volume 14, Issue 2, 1 February 2014, Pages 315-325
Keith D.K. Luk, Lim Beng Saw, Samuel Grozman, Kenneth M.C. Cheung, Dino Samartzis

New DRU classification: a practical and easy-to-use scheme that provide skeletal maturation status.
Growing Rods in the Treatment of Congenital Spinal Deformity

Seminars in Spine Surgery, Volume 22, Issue 3, September 2010, Pages 121-125

Pooria Salari, Behrooz A. Akbarnia, Hazem El-Sebaie, Muharrem Yazici

Best indicated in patients in whom primary problem is at vertebral column.
Growing rods have evolved into an effective and safe treatment option for children with early-onset scoliosis.
Growing rods in early-onset scoliosis. Do they really help to control the deformity and spinal and thoracic growth?


J.M. Sánchez Márquez, F.J. Sánchez Pérez-Grueso, N. Fernández-Baíllo, A. García Fernández

Moment at which spines of patients with early-onset scoliosis submitted to expandable techniques are in best condition is precisely after primary surgery.
Growth rods used to limit progression of scoliosis without restraining opportunity for spine to grow. Growth sustained by consecutive distraction at intervals of 6 months.
Treatment of juvenile scoliosis: Increasing the lengthening interval with the growing rod technique should not necessarily compromise thoracic growth


For less severe juvenile scoliosis: spacing out lengthenings over more than a year can decrease number of surgeries, while still controlling deformity and allowing longitudinal thoracic growth.

<table>
<thead>
<tr>
<th></th>
<th>Preoperative</th>
<th>Postoperative</th>
<th>End of follow-up</th>
</tr>
</thead>
<tbody>
<tr>
<td>Cobb angle</td>
<td>56.5 ± 8.1</td>
<td>24.7 ± 12.7</td>
<td>23.7 ± 11.8</td>
</tr>
<tr>
<td>Vertebral apical translation</td>
<td>4.8 ± 2.8 cm</td>
<td>2.1 ± 1.2 cm</td>
<td>1.9 ± 0.5 cm</td>
</tr>
<tr>
<td>Coronal balance</td>
<td>1.8 ± 2.2 cm</td>
<td>0.8 ± 0.92 cm</td>
<td>1 ± 1.2 cm</td>
</tr>
<tr>
<td>T1-T12 length</td>
<td>20.8 ± 4.6 cm</td>
<td>24.4 ± 4.0 cm</td>
<td>26 ± 4 cm</td>
</tr>
<tr>
<td>T5-T12 kyphosis</td>
<td>33.5 ± 15</td>
<td>23.4 ± 11.8</td>
<td>32.1 ± 14.6</td>
</tr>
<tr>
<td>Proximal junctional kyphosis</td>
<td>7.7 ± 8.8</td>
<td>9.1 ± 8.5</td>
<td>10.1 ± 7.9</td>
</tr>
<tr>
<td>Lumbar lordosis</td>
<td>42.5 ± 7.4</td>
<td>40.4 ± 15.4</td>
<td>44.7 ± 10.2</td>
</tr>
</tbody>
</table>
Three-dimensional stereoradiographic modeling of rib cage before and after spinal growing rod procedures in early-onset scoliosis

Marc Sabourin, Erwan Jolivet, Lotfi Miladi, Philippe Wicart, Virginie Rampal, Wafa Skalli

Growing rods techniques: a corrective effect on chest wall geometry.
Magnetically controlled growing rods for severe spinal curvature in young children: a prospective case series

Kenneth Man-Chee Cheung, Jason Pui-Yin Cheung, Dino Samartzis, Kin-Cheung Mak, Yat-Wa Wong, Wai-Yuen Cheung, Behrooz A Akbarnia, Keith Dip-Kei Luk

MCGR procedure safely and effectively used in outpatient settings, minimises surgical scarring and psychological distress, improves quality of life.
A novel approach to gradual correction of severe spinal deformity in a pediatric patient using the magnetically-controlled growing rod

The Spine Journal, Volume 14, Issue 7, 1 July 2014, Pages e7-e13

Jason Pui-Yin Cheung, Dino Samartzis, Kenneth Man-Chee Cheung

Correction of deformity while patient awake and repeated surgery avoided.
Reducing radiation exposure in early-onset scoliosis surgery patients: novel use of ultrasonography to measure lengthening in magnetically-controlled growing rods

The Spine Journal, Volume 14, Issue 10, 1 October 2014, Pages 2397-2404

Oliver M. Stokes, Elizabeth J. O'Donovan, Dino Samartzis, Cora H. Bow, Keith D.K. Luk, Kenneth M.C. Cheung

Use of ultrasonography: fewer whole spine radiographs when MCGRs.
Direct costs associated with the management of progressive early onset scoliosis: Estimations based on gold standard technique or with magnetically controlled growing rods

Orthopaedics & Traumatology: Surgery & Research, Volume 100, Issue 5, September 2014, Pages 469-474


Management of severe EOS using conventional growth rods: a substantial cost due to iterative distraction surgeries and associated hospital stays.
Precautions

Non fusion techniques, pedicle screws excellent anchor foundations in lumbar or sacral spine. Caution in upper thoracic spine because of risk of catastrophic pullout or placement in narrow concave pedicles closely approximated to spinal cord.

Current State of Pedicle Screw Constructs in Children with Spinal Deformity
Seminars in Spine Surgery, Volume 24, Issue 3, September 2012, Pages 192-201
Richard M. Schwend, Sreeharsha V. Nandyala
Somatosensory-evoked potential monitoring during instrumented scoliosis corrective procedures: validity revisited

The Spine Journal, Volume 14, Issue 8, 1 August 2014, Pages 1572-1580
Parthasarathy D. Thirumala, Lance Bodily, Derrick Tint, W. Timothy Ward, Vincent F. Deeney, Donald J. Crammond, Miguel E. Habeych, Jeffrey R. Balzer

SSEP monitoring alone during idiopathic scoliosis: a highly reliable method for detection and prevention of iatrogenic injury.
Minimally invasive thoracoscopic techniques, and endoscopic techniques of spinal instrumentation and scoliosis correction are now well established.

**Recent developments in scoliosis surgery**
*Current Orthopaedics, Volume 22, Issue 1, February 2008, Pages 42-47*
Patrick J. Kiely, Michael P. Grevitt
Early onset scoliosis. What are the options?


D.M. Farrington, Á. Tatay-Díaz

<table>
<thead>
<tr>
<th>Based on distraction</th>
<th>Guided growth</th>
<th>Inhibition of growth by compression of the convexity</th>
<th>Others</th>
</tr>
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<tbody>
<tr>
<td>Single growing rods</td>
<td>Luque Trolley</td>
<td>Staples with shape memory</td>
<td>Costal lengthening/shortening</td>
</tr>
<tr>
<td>Double growing rods</td>
<td>Shilla</td>
<td>Tether</td>
<td>Hemiepiphyseodesis of the neurocentral synchondrosis</td>
</tr>
</tbody>
</table>

Source: Skaggs et al.  

Hybrid growing rods  
Pfenix® / MAGEC®

Images of X-rays showing different stages of scoliosis treatment.
Improvement of Kyphoscoliosis in a 9-Year-Old Using Growth Modulation With a Posterior Tether: A Case Report

Spine Deformity, Volume 1, Issue 1, January 2013, Pages 79-83

Kira F. Skaggs, Karen S. Myung, Aimee E. Brasher, David L. Skaggs

A posterior unilateral tether successful at progressively improving kyphosis and preventing worsening of scoliosis in a 9-year-old boy, but led to fusion of spine within 31 months.
Study Group

12 patients since 2010 to date.

- 7 girls and 5 boys
- Cobb 40°
- 5 years
- 3 rods distraction and 6 braces
Comprehensive safety protocols include participation of all team members, make surgery safer over time, and use of postoperative management protocols and multidisciplinary teams to manage complex patients improve outcomes and shorten hospital stays.
Conclusion

Significant improvement in thoracic height percentile normalized for expected values based on pelvic width after initial surgery, and maintained over time.

Distraction-Based Treatment Maintains Predicted Thoracic Dimensions in Early-Onset Scoliosis

Michael P. Glotzbecker, Meryl Gold, Patrica Miller, Behrooz A. Akbarnia, Charles Johnston, Francisco Sanchez Perez Grueso, Suken A. Shah, John Emans