Real Congenital Scoliosis
(Rib Fusions/Jumbled Spine)

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Disclosures

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- Honorarium/consulting funds directed to research from Medtronic, Zimmer
Congenital Scoliosis

- Associated Conditions
  - Renal
  - Spinal cord
  - Cardiac
• 3 yo M, evaluation for congenital scoliosis

• Family complains of gait abnormality
  • Left thumb hypoplasia
  • Imperforate anus
  • Dextrocardia
  • Fused kidneys
The Effect of Opening Wedge Thoracostomy on Thoracic Insufficiency Syndrome Associated with Fused Ribs and Congenital Scoliosis

Management of thoracic insufficiency syndrome in patients with Jarcho-Levin syndrome using VEPTRs (vertical expandable prosthetic titanium ribs).

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2. Spinal & Thoracic Treatment and Research (STTAR) Center, Children’s Hospital of San Antonio, 333 North Santa Rosa Street, San Antonio, TX 78207. E-mail address for M.K. Roth: megan.roth@christushealth.org.
Does expansion thoracostomy with fused ribs result in improved outcomes?

The Effect of Expansion Thoracostomy on Spine Growth in Patients with Spinal Deformity and Fused Ribs Treated with Rib-Based Growing Constructs

Fady J. Baky, MD, A. Noelle Larson, MD, Tricia St. Hilaire, MPH, Jeff Pawelek, MD, David L. Skaggs, MD, John B. Emans, MD, Joshua M. Pahys, MD, Children’s Spine Study Group, Growing Spine Study Group

Rib Fusion Severity

- Rated rib fusions as: mild/moderate/severe

Mild
1-2 Rib Fusions

Moderate
> 2 Rib Fusions

Severe
Altered Chest Architecture
<table>
<thead>
<tr>
<th>Measure</th>
<th>Thoracostomy (N=103)</th>
<th>No Thoracostomy (N=48)</th>
<th>P-value</th>
</tr>
</thead>
<tbody>
<tr>
<td>Follow-Up (Years)</td>
<td>6.8</td>
<td>5.8</td>
<td>0.08</td>
</tr>
<tr>
<td># of Lengthenings</td>
<td>8.3</td>
<td>6.6</td>
<td>0.017</td>
</tr>
<tr>
<td>Total # of Surgeries</td>
<td>11.5</td>
<td>9.6</td>
<td>0.031</td>
</tr>
<tr>
<td># of Revision Surgeries</td>
<td>2.3</td>
<td>2</td>
<td>0.48</td>
</tr>
<tr>
<td>Underwent Final Fusion</td>
<td>19/103</td>
<td>10/48</td>
<td>0.83</td>
</tr>
<tr>
<td>Change in T1-S1 height at initial procedure (cm)</td>
<td>2.01</td>
<td>1.24</td>
<td>0.20</td>
</tr>
<tr>
<td>Change in T1-S1 height with subsequent distractions (cm)</td>
<td>6.7</td>
<td>4.7</td>
<td>0.05</td>
</tr>
<tr>
<td>Change in T1-S1 height at Final Fusion (cm)</td>
<td>2.2</td>
<td>1.6</td>
<td>0.57</td>
</tr>
<tr>
<td>Total Change in T1-S1 (cm)</td>
<td>7.2</td>
<td>4.8</td>
<td>0.0043</td>
</tr>
</tbody>
</table>
What Proximal Anchors are Best?

• To determine outcomes (thoracic height and Cobb angle) in patients with congenital fused ribs treated with proximal spine anchors (spine-based growing devices) compared to constructs with proximal rib anchors (rib-based devices).

Spine Deformity With Fused Ribs Treated With Proximal Rib- Versus Spine-Based Growing Constructs.

Larson AN\textsuperscript{1}, Baky FJ\textsuperscript{2}, St Hilaire T\textsuperscript{3}, Pawelok J\textsuperscript{4}, Skaggs DL\textsuperscript{5}, Emans JB\textsuperscript{6}, Pahys JM\textsuperscript{7}; Children's Spine Study Group\textsuperscript{3}; Growing Spine Study Group\textsuperscript{4}.

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\textsuperscript{4} Growing Spine Foundation, 555 East Wells Street, Suite 1100, Milwaukee, WI 53202, USA.
\textsuperscript{5} Children's Hospital Los Angeles, 4650 Sunset Blvd, Los Angeles, CA 90027, USA.
\textsuperscript{6} Children's Hospital, Harvard Medical School, 300 Longwood Ave, Boston, MA 02115, USA.
\textsuperscript{7} Shriners Hospital for Children, 3551 N Broad St, Philadelphia, PA 19140, USA.
Proximal fixation assessed as spine-based or rib-based (excluded those with both types of anchors)
Study Design

- Retrospective review of prospectively collected data from multicenter databases (GSSG & CSSG).
- Minimum 2 year follow-up
- Early onset scoliosis and rib fusions
- 176 patients identified
  - 16 proximal spine anchors
  - 160 proximal rib anchors
    - 154 VEPTR, 6 other
    - 106 had thoracoplasty at implantation
    - 90 had rib-to-rib construct
## Results: Severity of Rib Fusions

<table>
<thead>
<tr>
<th>Rib Deformity</th>
<th>Spine-Based Devices (N=16)</th>
<th>Rib-Based Devices (N=160)</th>
</tr>
</thead>
<tbody>
<tr>
<td>Mild (%)</td>
<td>6 (38%)</td>
<td>60 (38%)</td>
</tr>
<tr>
<td>Moderate (%)</td>
<td>6 (38%)</td>
<td>66 (41%)</td>
</tr>
<tr>
<td>Severe (%)</td>
<td>2 (13%)</td>
<td>26 (16%)</td>
</tr>
<tr>
<td>Indeterminate</td>
<td>2 (13%)</td>
<td>8 (5%)</td>
</tr>
</tbody>
</table>
## Results: # of Surgeries

<table>
<thead>
<tr>
<th></th>
<th>Spine-Based Devices (N=16)</th>
<th>Rib-Based Devices (N=160)</th>
<th>P-value</th>
</tr>
</thead>
<tbody>
<tr>
<td># Lengthening Surgeries</td>
<td>6.3 (1-14)</td>
<td>7.9 (1-21)</td>
<td>0.12</td>
</tr>
<tr>
<td># All Surgeries</td>
<td>8.0 (2-18)</td>
<td>11.2 (2-30)</td>
<td>0.007</td>
</tr>
<tr>
<td># Revision Surgeries</td>
<td>1.6 (0-7)</td>
<td>2.3 (0-12)</td>
<td>0.17</td>
</tr>
</tbody>
</table>
## Results: Spinal Height

<table>
<thead>
<tr>
<th></th>
<th>Spine-Based Devices (N=16)</th>
<th>Rib-Based Devices (N=160)</th>
<th>P-value</th>
</tr>
</thead>
<tbody>
<tr>
<td>Total Change in T1-T12</td>
<td>6.0 (-4.5-22.4)</td>
<td>3.4 (-3.01-11.7)</td>
<td>0.26</td>
</tr>
<tr>
<td>Total Change in T1-S1</td>
<td>9.1 (3.0-13.1)</td>
<td>6.3 (-4.1-18.2)</td>
<td>0.06</td>
</tr>
<tr>
<td>Distraction Change T1-T12</td>
<td>5.7 (-4.5-22.4)</td>
<td>3.3 (-3.3-11.7)</td>
<td>0.35</td>
</tr>
<tr>
<td>Distraction Change T1-S1</td>
<td>8.1 (-1.3-7)</td>
<td>5.9 (-5-10.3)</td>
<td>0.04</td>
</tr>
<tr>
<td>Length/Distraction</td>
<td>0.29</td>
<td>0.3</td>
<td>&gt; 0.05</td>
</tr>
<tr>
<td>Final Fusion Change* T1T12</td>
<td>0.75 (-0.7-2.4)</td>
<td>1.4 (-2.1-6.2)</td>
<td>0.36</td>
</tr>
<tr>
<td>Final Fusion Change* T1S1*</td>
<td>0.58 (-2.2-2.7)</td>
<td>2.1 (-5.5-8.2)</td>
<td>0.16</td>
</tr>
</tbody>
</table>

**Only 46 patients (40 rib-construct and 6 spine-construct) had final fusion.**
Results: Coronal and Sagittal Plane

<table>
<thead>
<tr>
<th></th>
<th>Spine-Based Devices (N=16)</th>
<th>Rib-Based Devices (N=160)</th>
<th>P-value</th>
</tr>
</thead>
<tbody>
<tr>
<td>Post-Treatment Cobb</td>
<td>36.7 (0-62)</td>
<td>57.8 (11-117)</td>
<td>&lt;0.001</td>
</tr>
<tr>
<td>Post-Treatment Kyphosis</td>
<td>34.7 (0-72)</td>
<td>52.4 (0-108)</td>
<td>0.003</td>
</tr>
<tr>
<td>Δ in Cobb Angle</td>
<td>24.4 (-18-66)</td>
<td>11.3 (-31-88)</td>
<td>0.049</td>
</tr>
<tr>
<td>Δ in Kyphosis</td>
<td>20.3 (-10-62)</td>
<td>-7.3 (-63 - 74)</td>
<td>0.002</td>
</tr>
</tbody>
</table>
Discussion

- No difference in T1-T12/T1-S1 growth achieved
- More surgeries in the rib-based group
- Increased thoracic Cobb angle and kyphosis at latest follow-up in rib-based group
  - Previously rib-based devices have been found to be associated with increasing kyphosis
Case: 6 month old

- Thoracic meningocele
- Solitary right kidney
- Rectal urethral fistula status post colon diversion takedown
- Left upper extremity abnormalities
Early Onset Scoliosis – Congenital 3 yo

- 10.6 kg
- 83.7 cm
- T1-T12 8 cm
Congenital Deformities/Rib Fusions

- Three rib blocks
  - Planned two rib cradles
  - Two screws distally
- No thoracostomy (ribs somewhat mobile)
- Lack of soft tissue coverage
• Lower lesion
• Poor intrinsic function
Neuromonitoring During Index Procedure

- 3 lower extremity
- 1 upper lead (not ulnar n.)

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Congenital Deformities/Rib Fusions

- Can sustain brachial plexus injuries from rib-based devices:
  - Compression from 1st rib
  - Retracted Superior pole of scapula
  - Secondary to Sprengel deformity reconstruction by pulling scapula down (Joiner et al., JBJS 2013)
Revised Construct POD2
2 Years Follow-Up → Upsized Rod and Screws

6 yo M

10.6 → 14 kg

83.7 → 98.7 cm

T1-T12

8 → 13.7 cm
• 1 yo M
4 yo

Left Lung Volume

Volume = 174.3 cm³

Volume = 161.3 cm³
12 yo M

- 16 cm thoracic ht
Summary: Congenital Scoliosis with Rib Fusions

- Always look for associated conditions (MRI, etc)
- Monitor arms and legs during surgical procedures
- PSSG database
  - Better deformity control with spine-based proximal rib anchors?
  - Improved overall spine length with thoracostomy
    - No data on PFTs, chest wall flexibility/function
- Area for further research
  - Use of MCGR
  - PFTs, more functional outcome measures
Thank you!

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