Growing Rods for Scoliosis in Spinal Muscular Atrophy: What’s the role in 2016?

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Johns Hopkins Bloomberg Children’s Center
## Growing Rods for Scoliosis in Spinal Muscular Atrophy

**Disclosures**

<table>
<thead>
<tr>
<th>Paul Sponseller MD</th>
<th>DePuy Spine (a,e); Globus(e)</th>
</tr>
</thead>
<tbody>
<tr>
<td>Growing Spine Study Group</td>
<td>Growing Spine Foundation (a)</td>
</tr>
</tbody>
</table>

- a. Grants/Research Support
- b. Consultant
- c. Stock/Shareholder
- d. Speakers' Bureau
- e. Other Financial Support
History of Spine fusion in SMA

• 1974

The Orthopaedic Aspects of Spinal Muscular Atrophy

BY E. P. SCHWENTKER, M.D.‡, AND D. A. GIBSON, M.D., F.R.C.S.‡, TORONTO, ONTARIO, CANADA

ABSTRACT: To clarify the role of the orthopaedic surgeon in the management of spinal muscular atrophy, the records of 130 patients were reviewed. Seventy-three had died. Of the remaining fifty-seven, fifty were re-examined. The clinical manifestations of the disease, particularly those giving rise to orthopaedic problems, were studied.

At the time of review the average age of the patients was 11.5 years. Thirty-five could not walk and all had some degree of muscle weakness, more marked proximally and in the lower limbs. The most common medical problem was repeated respiratory infection and the major orthopaedic problem was scoliosis, often severe. Nine of the thirty-five patients with scoliosis had had spine fusion. Most of them suffered some functional loss in attaining spinal stability.

Review of Clinical Aspects

Childhood spinal muscular atrophy covers a spectrum of clinical manifestations, representing a variable expression of a single genetic entity, the composite of three or more entities.3,4,13

Infantile spinal muscular atrophy (Hoffmann's disease)10,11 starts in the first year of life with motor deterioration and it nearly always ends in death from respiratory causes by the age of four. Patients with this form of the disease are never able to sit without support, and their orthopaedic problems rarely require treatment because their respiratory status is so markedly involved.
Growing Rods

• Limited proximal & distal “foundations”
  – Allow the rest to grow
• Dual Rods
• Lengthen every 6-12 months
• ?Final fusion at maturity?
Rationale

• Improve sitting tolerance
  – ? Prevent pain
  – Applies to fusion as well as growing rods
• Improve Organ Function:
• “Function follows the Frame”
  – Minimize restrictive lung disease
  – Less abdominal crowding improves diaphragm?
  – Improve GI function, reflux
Our Patient Group

- 15 SMA patients treated with GRs
  - Age at insertion 8 years (range, 3.5-10.8)
  - Follow-up 54 months
  - Levels spanned ~16

- SMA patients compared to 80 early onset idiopathic scoliosis patients treated with GRs
• SMA patients have progressive rib collapse leading to increasingly triangular thoracic shape

• Quantified shape using
  – T6:T10 RVA ratio
  – T6:T12 thoracic width ratio

\[
\text{T6:T10 RVA} = \frac{22^\circ + 76^\circ}{58^\circ + 67^\circ} = 0.78
\]

\[
\text{T6:T12 Thoracic Width} = 0.69
\]
## Results

### Coronal Alignment

<table>
<thead>
<tr>
<th>Measurement</th>
<th>Mean</th>
</tr>
</thead>
<tbody>
<tr>
<td><strong>Cobb</strong></td>
<td></td>
</tr>
<tr>
<td>Preop</td>
<td>89°</td>
</tr>
<tr>
<td>Post-initial</td>
<td>45°</td>
</tr>
<tr>
<td>Latest f/u</td>
<td>55°</td>
</tr>
<tr>
<td><strong>Pelvic Obliquity</strong></td>
<td></td>
</tr>
<tr>
<td>Preop</td>
<td>31°</td>
</tr>
<tr>
<td>Post-initial</td>
<td>9°</td>
</tr>
<tr>
<td>Latest f/u</td>
<td>11°</td>
</tr>
</tbody>
</table>
# Length Measurements

<table>
<thead>
<tr>
<th>Measurement</th>
<th>Mean ± Range</th>
</tr>
</thead>
<tbody>
<tr>
<td>T1-S1 Length</td>
<td></td>
</tr>
<tr>
<td>Preop</td>
<td>26.7 cm</td>
</tr>
<tr>
<td>Latest f/u</td>
<td>34.5 ± 3.9 cm</td>
</tr>
<tr>
<td>Yearly growth during GR</td>
<td>1.2 ± 0.6 cm</td>
</tr>
</tbody>
</table>
Rib Collapse Over Time

<table>
<thead>
<tr>
<th>SMA</th>
<th>Idiopathic</th>
</tr>
</thead>
<tbody>
<tr>
<td>ΔT6:T12 Thoracic Width</td>
<td>ΔT6 Convex Angle</td>
</tr>
<tr>
<td>-0.04 ± 0.08</td>
<td>-15° ± 16°</td>
</tr>
<tr>
<td>0.00 ± 0.06</td>
<td>-2° ± 17°</td>
</tr>
</tbody>
</table>

- SMA group
  - Compared to idiopathic group more rib collapse and decrease in thoracic width ratio
  - GRs do not halt rib collapse in SMA, but may slow progression
    - No untreated controls
Use of Hospital Care

<table>
<thead>
<tr>
<th>Diagnosis</th>
<th>GR Insertion (days)</th>
<th>GR Lengthening (days)</th>
<th>Final Fusion (days)</th>
<th>% Outpatient Lengthenings</th>
</tr>
</thead>
<tbody>
<tr>
<td>SMA</td>
<td>7.8</td>
<td>2.1</td>
<td>10.6</td>
<td>39%</td>
</tr>
<tr>
<td>Idiopathic</td>
<td>5.3</td>
<td>0.8</td>
<td>5.6</td>
<td>47%</td>
</tr>
<tr>
<td>P Value (SMA &gt; Idiopathic)</td>
<td>0.08</td>
<td>0.01</td>
<td>0.11</td>
<td></td>
</tr>
</tbody>
</table>

- Hospital stay durations compared for 15 SMA patients and 80 idiopathic scoliosis patients

- SMA hospital stays longer for lengthening procedures
  - Trend towards longer stays in insertion and final fusion procedures
Complications (new intervention or prolonged stay)

- Fewer complications per patient in SMA
  - 0.5 per SMA Patient
  - 1.1 per Idiopathic Patient
    - $P = 0.05$

- More serious complications in SMA
  - 2 patients required tracheostomies
  - 1 intraoperative cardiac arrest (resuscitated)

- No
  - neurologic complications
  - No infections
  - No mortality
Saga of GR

- Pt #1: 7 yrs 10 cm no complications
  - No final fusion
SMA-2

- Poor head control
  - May develop neck deformity
    - Or just “unmask” it by correcting spine
    - Recognize and treat it!
• 16 children; 11 type 2
  – 5 yr follow up
  – 4 lengthenings; 5 cm T1-12 ht gained
    • 65% curve correction maintained
  – Respiratory support needs stabilized
  – Normalized PFTs declined
Feinberg 2016

- 64 patients with GR or VEPTR
- FVC maintained but % ile down
- HRQoL domain scores increased
Conclusions about Growing Rods

- GRs safely improve trunk/lung height, control curve, pelvic obliquity
- Do not halt rib collapse, but may slow progression
- Hospital stays longer in SMA
- Rate of significant complications lower in SMA
Game-Changers?

- Spine becomes stiff and “auto-fuses” over time
- Can “Final Fusion” Procedure be Avoided at Maturity After GR?
Results: Radiographic Outcomes

- **Correction of major curve**
  - NF group: 46% correction
  - FF group: 37% correction
  - No significant difference in curve correction (P=0.23)

- **Increase in trunk height (T1-S1 length)**
  - NF group: 30% (11.5cm)
  - FF group: 25% (9.5 cm)
  - Trunk height gain in NF significantly higher (P<0.01)
Idiopathic patient

6 yrs

14 yrs

17 yrs

No Final Fusion
The MAGEC Technology
MAGEC: next generation of distraction-based treatment for early onset scoliosis
MAGEC

- FDA approved 3/15
Shilla

- “Self-guided” spinal growth
- Fuse/control apex
- Rods grow off screws
- 5 year follow up
  - 35% less length gain
  - 35% less curve correction
  - Half as many procedures
4 yr f/u

3yo
89°

7yo
Screw pull-out
Rods Broke
Other Options:

“Trolley”
Wires slide with growth

[Images of X-rays showing spinal fusion with wires]
Other Options: Early Fusion

- Zebala, Sucato,
  - Recurrent Deformity mainly if fused <8 yrs

- Weinstein- Fusion at mean age 10
  - PFT declines slowed
  - 1 mortality at 20 yr follow up
Summary

- We can:
- Stabilize spine
- Grow the frame – but when?

- Evidence imperfect (level 3 &4) but evolving
- Newer options decrease the burden
Thank you
## Results

### Final Fusion

<table>
<thead>
<tr>
<th>Measurement</th>
<th>Mean ± Std Dev</th>
</tr>
</thead>
<tbody>
<tr>
<td>Number of patients</td>
<td>6</td>
</tr>
<tr>
<td>Age at fusion</td>
<td>13.1 ± 2.5 years</td>
</tr>
<tr>
<td>Preoperative Cobb angle</td>
<td>75 ± 17°</td>
</tr>
<tr>
<td>Pre-fusion Cobb Angle</td>
<td>61 ± 20°</td>
</tr>
<tr>
<td>Post-fusion Cobb angle</td>
<td>54 ± 12°</td>
</tr>
</tbody>
</table>

[Images of X-rays showing spinal fusion]
## 36 Case Matched Controls

### SHILLA Vs Growing Rods

<table>
<thead>
<tr>
<th></th>
<th>Growing Rod</th>
<th>Shilla</th>
<th>P-value</th>
</tr>
</thead>
<tbody>
<tr>
<td><strong>Total # of surgeries per patient</strong></td>
<td>7.0</td>
<td>2.8</td>
<td>&lt;0.001</td>
</tr>
<tr>
<td><strong>Average change in cobb angle</strong></td>
<td>-36 degrees</td>
<td>-23 degrees</td>
<td>0.019</td>
</tr>
<tr>
<td><strong>Average change in T1-S1</strong></td>
<td>8.5 cm</td>
<td>6.4 cm</td>
<td>0.031</td>
</tr>
</tbody>
</table>
Retrospective study

• Evaluate use of growing rods for scoliosis in SMA
  – Structural effectiveness
  – Perioperative hospital care
    • Rate of significant complications