Are Magnetically Controlled Growing Rods a Good Option for Collapsing Spine Deformity in Spinal Muscular Athropy Type-II Associated Early Onset Scoliosis?

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• No disclosure!
SMA & Spine Deformity

• Collapsing spine deformity
  - C-shaped scoliosis / lack of compensation
  - Parasol rib deformity
  - Pelvic obliquity
  - Progression 5-15°/year
• Incidence is nearly 100%
  - Severe progression once patient becomes non-ambulatory
• Impairs functional status
  - Sitting imbalance
  - Unappealing cosmesis
  - Exacerbate pulmonary problems
• Conservative treatment fails
  - Bracing can not stop progression
  - Severe breathing impairment
# SMA & Spine Deformity

<table>
<thead>
<tr>
<th>SMA Type</th>
<th>Age of Onset</th>
<th>Motor Milestones</th>
<th>Age of Death</th>
</tr>
</thead>
<tbody>
<tr>
<td>I</td>
<td>&lt; 6 months</td>
<td>Unable to sit</td>
<td>&lt; 2 years</td>
</tr>
<tr>
<td>II</td>
<td>18 months</td>
<td>Sit independently, cannot stand</td>
<td>2nd – 3rd decade</td>
</tr>
<tr>
<td>III</td>
<td>&gt; 18 months</td>
<td>Stand and walk independently</td>
<td>Normal life expectancy</td>
</tr>
<tr>
<td>IV</td>
<td>Adolescent or adult onset</td>
<td>Retain walking, muscle pain</td>
<td>Normal life expectancy</td>
</tr>
</tbody>
</table>

**Early onset scoliosis**
GFT & SMA-related EOS

- **SMA&Traditional Growing Rods**
  - **McElroy et al, Spine 2011**
    Stabilizes the spine deformity, ↑spinal height
    Does not halt rib collapse
    Longer hospital stay but fewer major complications than idiopathic EOS
  - **Chandran et al, J Pediatr Orthop 2011**
    Pulmonary functions maintained early after surgery (↑FVC, 0.53 to 0.67 L)
  - **Lenhart et al, J Pediatr Orthop 2016**
    ↑Spinal height, ↑Thoracic cavity size
    ↑Absolute FVC
    ↓Predicted FVC% over time
GFT& SMA-related EOS

- SMA & Eiffel tower MCGR
  - Lorenz HM, JBJS Open Access 2017
    - Bilateral rib to pelvis construct with a magnetically controlled implant
    - Good control in coronal plane (main curve & pelvic obliquity) and spinal height
    - Poor control in sagittal plane
    - Effects on pulmonary functions are unclear
    - ↓ complication rates
    - ↓ the burden of repetitive lengthening surgeries
Aim

• A new treatment algorithm
  – SMA type 2 EOS
  – Spine based MCGR
• Does this algorithm work?
Methods

• Prospective study
• Single institution
• Inclusion criteria
  – SMA type-II
  – EOS (<10 years)
  – Collapsing spine deformity
  – Growing rods treatment with MCGR
  – >1 year follow-up
• Demographic, clinical, radiological information, surgical details and final status of the patients
Treatment algorithm

SMA - II Scoliosis

- Dual MCGR
- Instrumentation not extended to pelvis

- Early postop

- Dual MCGR
- Instrumentation extended to pelvis

- Preop TRUGA
- Apical vertebra inside the stable zone (n:6)
- Apical vertebra inside the stable zone

- Early postop

- MCGR (concave)
- Convex MCGR (convex)

- Preop TRUGA
Results

- Mean f/u
  - 29.9 months (12-50)
- # Outpatient lengthening
  - 6.7
- Deformity
  - Cobb angle
    - 81.8° (66-115) → 29° (11-57) → 23.6° (12-50)
  - Pelvic Obliquity
    - 21.9° (8-30) → 5.1° (2-8) → 4.4° (2-7)
Results

• T1-S1 height (mm)
  - 329 (280-376) → 356 (310-386)
  - 10.8 mm/year

• Thoracic kyphosis
  - 46.6 (28-66) (early postop) → 54.2 (35-71) (last f/u)

• Lumbar lordosis
  - 43.5 (25-62) (early postop) → 46.1 (27-65) (last f/u)
Results

- Distal adding-on in 2 pts. w/o initial pelvic fixation
  - Revised with extending the inst. to pelvis
Results
Conclusion

• MCGR is a good option in SMA-related EOS
  – Reduce the burden of repetitive lengthening surgeries
  – Good coronal/sagittal plane deformity correction/maintenance
  – Low complication rates

• Algorithm does work!
  – Except stopping @ lumbar spine
  – Natural course of underlying disease!