Tethered spinal cord and EOS: When to treat and when to watch?

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Disclosures up to date
Pathophysiology: Tethered Cord Syndrome

• Relationship between spinal cord and vertebral column changes during development
• Conus lies at approximately:
  – Coccyx 30mm embryo
  – L2-3 Birth
  – L1-2 1 years
Pathophysiology: Tethered Cord Syndrome

- Causes neurological impairment by mechanically anchoring the spinal cord at the site of the cord’s fusion with the lesion
- Leads to traction on the spinal cord with resultant relative ischemia
- If surgery is not performed, neurological deficits frequently are progressive (30-40%)
- Once a neurological deficit develops, it is often irreversible
Why be concerned about TSC with EOS?

- Intraspinal anomalies seen in 20-50% of children with congenital scoliosis
- TSC release can delay the progression of scoliosis
- Deformity correction in the setting of untreated TSC may be problematic
  - Neurological injury
  - Unreliability or loss of IOM signals limiting correction

<table>
<thead>
<tr>
<th>Serial no.</th>
<th>Intraspinal anomaly</th>
<th>No. of patients (55/119)</th>
</tr>
</thead>
<tbody>
<tr>
<td>1</td>
<td>Tethered cord and low conus</td>
<td>27 (48.2)</td>
</tr>
<tr>
<td>2</td>
<td>Diastematomyelia</td>
<td>18 (32.1)</td>
</tr>
<tr>
<td>3</td>
<td>Syringomyelia</td>
<td>17 (30.3)</td>
</tr>
<tr>
<td>4</td>
<td>Arnold Chiari malformation</td>
<td>11 (19.6)</td>
</tr>
<tr>
<td>5</td>
<td>Intradural cyst</td>
<td>1 (1.7)</td>
</tr>
<tr>
<td>6</td>
<td>Intradural lipoma</td>
<td>2 (3.5)</td>
</tr>
</tbody>
</table>

Values are presented as percent (number). 35.7% patients (20/56) had multiple intraspinal abnormalities.
When to act

- Clear TSC with low lying conus:
  - Spinal cord lipomas
  - Split cord malformations
  - *Symptomatic* myelomeningoceles
  - Dermal sinus tracts
  - Others
When to act

- 6 year old girl presented with scoliosis
- Back pain
- No skin lesions
- Neuro intact
- Underwent TSC release
When to act
When to act

• 3 year old girl with progressive scoliosis and hairy patch in thoracic region
When to act
Effect of TSC release on scoliosis

- Bowman/McLone study:
  - 36pts, 7yr follow-up
  - Myelo only
  - 33% progression at 2 years
  - 66% progression at 7 years (36% fusion)
- Pierz study:
  - 21 pts, 5yr f/u
  - 57% progression
- Hassani study:
  - 20 pts, 4yr f/u
  - 60% progression (50% fusion)

- Newton study:
  - 15pts (10 TSC)
  - 47% progression
- Chern study:
  - 14pts
  - All tight filum
  - 64% progression (36% fusion)
Effect of TSC release on scoliosis

**TABLE 2: Summary of patient demographics, symptomatology, and scoliosis measurements**

<table>
<thead>
<tr>
<th>Variable</th>
<th>Value</th>
</tr>
</thead>
<tbody>
<tr>
<td>mean age (yrs)</td>
<td>8.9 ± 3</td>
</tr>
<tr>
<td>male</td>
<td>16 (59)</td>
</tr>
<tr>
<td>skeletal maturity at TCS</td>
<td></td>
</tr>
<tr>
<td>Risser Grades 0–2</td>
<td>17 (63)</td>
</tr>
<tr>
<td>Risser Grades 3–5</td>
<td>10 (37)</td>
</tr>
<tr>
<td>cause of TCS</td>
<td></td>
</tr>
<tr>
<td>postmyelomeningocele repair</td>
<td>14 (50)</td>
</tr>
<tr>
<td>fatty tumour</td>
<td>5 (18.5)</td>
</tr>
<tr>
<td>lipomeningocele</td>
<td>3 (11)</td>
</tr>
<tr>
<td>diastematomyelia</td>
<td>2 (7.4)</td>
</tr>
<tr>
<td>arthrogryposis</td>
<td>1 (3.7)</td>
</tr>
<tr>
<td>lipomyelomingocele w/ occult dysraphism</td>
<td>1 (3.7)</td>
</tr>
<tr>
<td>imperforata anus w/ S-2 hamisvertebra</td>
<td>1 (3.7)</td>
</tr>
</tbody>
</table>

**TABLE 3: Five-year incidence rate of radiological progression and subsequent fusion in 4 subgroups of patients with TCS-associated spinal deformity**

<table>
<thead>
<tr>
<th>Cobb Angle/ Risker Grades</th>
<th>No. of Patients</th>
<th>% w/ Radiological Progression</th>
<th>% w/ Subsequent Fusion</th>
</tr>
</thead>
<tbody>
<tr>
<td>&lt;40°/3−5</td>
<td>8</td>
<td>0</td>
<td>0</td>
</tr>
<tr>
<td>&lt;40°/0−2</td>
<td>11</td>
<td>54</td>
<td>27</td>
</tr>
<tr>
<td>&gt;40°/3−5</td>
<td>2</td>
<td>50</td>
<td>0</td>
</tr>
<tr>
<td>≥40°/0−2</td>
<td>6</td>
<td>83</td>
<td>83</td>
</tr>
</tbody>
</table>

McGirt M et al (2009) JNS Peds
Effect of TSC release on scoliosis

- Higher risk of curve progression after TSC release:
  - Bigger curve (Cobb angle >35-45°)
  - Younger age (<10 yrs)
  - Skeletal immaturity (Risser 0-2)
  - Thoracic level myelomeningocele
  - Greater vertebral rotation
- Delays the need for definitive scoliosis surgery by several years

When NOT to act

- Radiologists call “syrinx” when <3mm: dilation of the central canal
- Radiologists call “filum lipoma” when trace fat seen in filum (6% autopsy and MRI; 0.4% deterioration)
- *Asymptomatic* myelomeningocele
- Vast majority of cases with normal level conus

When is there equipoise?

- Concept of an occult tethered cord syndrome (OCTS) first reported in 1990 by Khoury et al.
- Similar symptoms/signs of tethered cord syndrome but conus lies in a normal position at or above L2
- Can account for up to 20% of cases in large series
When is there equipoise?

Key Question:

- What percentage of patients with progressive scoliosis and are otherwise neurologically and urologically normal with a normal level conus have OTCS?

Answer:

- Unknown
- *No definitive test exists to identify OTSC*
When is there equipoise?

- Consider TSC release if atypical or rapidly progressive curve under the following circumstances:
  - Any symptoms of TSC (progressive pain, weakness, sensory loss, urologic dysfunction)
  - Additional radiographic findings including fatty filum, syrinx, or vertebral body anomalies
- Why?:
  - Morbidity is low (1-2%)
  - Loss of potentials in the OR makes for a very bad day
  - EOS patients extremely difficult to manage
  - In some patients, it will buy some time
Occult TSC

- 3 month old girl born with sacral-coccygeal pit
- Gets MRI for evaluation
- Neuro exam normal
Occult TSC

- At 6 years of age, develops rapidly progressive scoliosis over 6 month period
- Repeat MRI shows no change
- Neuro exam remains normal
Occult TSC
Conclusions

• When to Treat:
  – Clinical symptoms of TSC
  – MRI showing low level conus
  – Consider occult TSC if rapidly progressive curve with additional symptoms or radiographic signs

• When to Watch:
  – Reports of “syrinx” < 3mm diameter
  – Reports of “fatty filum” with normal level conus and no other symptoms
Thank you!
# Results: Multivariate Analysis

## Relative Risk of Post-Operative Complications Associated with Prophylactic Untethering

with multivariable logistic regression adjustment for age, gender, VPS, and level of myelomeningocele

<table>
<thead>
<tr>
<th>Outcome</th>
<th>RR</th>
<th>95% C.I.</th>
<th>p-value</th>
</tr>
</thead>
<tbody>
<tr>
<td>SSI</td>
<td>2.65</td>
<td>1.17-5.02</td>
<td>0.0196</td>
</tr>
<tr>
<td>Return to OR</td>
<td>2.17</td>
<td>1.02-4.65</td>
<td>0.0453</td>
</tr>
<tr>
<td>Any Complication*</td>
<td>2.25</td>
<td>1.07-4.74</td>
<td>0.0325</td>
</tr>
</tbody>
</table>

* Composite outcome of any SSI, return to OR, CSF leak, VPS malfunction, other wound complication

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**When NOT to act**

Goldstein H (2019) Childs Nerv System