Spinal Dysraphism 101 for Orthopedists: What Constitutes an “Actionable” MRI Finding?

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Intraspinal Anomalies and Spine Deformity

- Evolving understanding
- PubMed search: either ‘Chiari,’ ‘tethered cord,’ or ‘split cord malformation’ AND spine deformity
  - > 500 results
  - Over 100 in past 18 months!
- Regional differences
Patient C.L.: Tethered Cord

- 3 yo with progressive congenital scoliosis
- Neurologically non-focal exam
- MRI
  - ‘Low lying cord’ with fatty filum
Discussion

- When to get MRI?
- Indications for fatty filum release?
- Should it be done concurrently?
- What if tethered secondary to myelomeningocele?
- Brief overview of technique
  - Surgical video
Fatty Filum with Low Lying Conus

- Recommend untethering
  - Low morbidity
    - Bowman et al, J Neurosurg Pediatr 2009
- Family feels everything done
Uneventful Surgery
Case: 14 yo MM with rigid 90 degree curve and back pain

Shunt?
Results: Baseline

MM pts undergoing scoliosis correction surgery, 1994-2017 (n = 350)

Excluded due to rod lengthening procedure only (n = 140)

No Untethering (n = 155) VS

Prior (<3m) Untethering (n = 21)
Concomitant Untethering (n = 32)
**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>
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<tr>
<td>Any Complication *</td>
<td>2.25</td>
<td>1.07-4.74</td>
<td>0.0325</td>
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</tbody>
</table>

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

**Relative Risk (RR)**

**Favors Untethering**

**Favors Not Untethering**
Patient M.M.: Chiari Malformation

- 11-year-old girl presented with progressive scoliosis
  - Main thoracic curve 35 to 60°
  - Kyphosis T5-12: 75°
  - Neurologically intact
Scoliosis: When to Get an MRI?

Scoliosis >10°

- Typical curve
- Followed
  - Pre-op scan
  - Intra-op
  - Post-op
  - MRI

- Rapid progression
- MRI

Atypical curve:
- Left thoracic
- Age <11 y
- Rapid progression
- MRI


Imaging

Questions

• Does every Chiari need to be decompressed prior to curve correction?
• How long should we wait after Chiari decompression for curve correction?
• What if syrinx does not get better?
• What are the indications for another procedure, like a syrinx shunt?
• What are the risks of scoli surgery if the syrinx is still “big”? 

Questions

• Does every Chiari need to be decompressed prior to curve correction?
  • No. Most do (>95%), but not all
  • Minimal tonsillar herniation or a small syrinx could safely undergo curve correction without PFD
  • Syrinx is probably not driving the curve
  • Chiari is probably incidental
  • Consult with neurosurgeon; you probably don’t want to make this decision on your own!
This Patient’s Course

• Posterior fossa decompression with duraplasty
  • Randomized trial supports this approach
• MRI 3-6 months post-op to investigate syrinx size
  • If syrinx improved, then safe to undergo curve correction shortly thereafter
  • If not improved, then repeat imaging every 4-6 months up to a year
Six Months Later

- Repeat MRI and x-rays
  - No change in syrinx or scoli
- What now?
- Options:
  - Wait
  - Repeat PFD (with or without 4\textsuperscript{th} ventricle shunt)
  - Shunt the syrinx
- This patient: opted to wait longer

6 months
One Year Later
One Year MRI

- Cervical MRI
  - Decrease in syrinx size (whew!)
- Surgery
  - T2 - L3 PSF with Ponte osteotomies
  - Neuromonitoring
    - Motors inconsistent
    - Multiple wake-ups
- What are the risks of scoliosis surgery when syrinx is still “big”? 


Spinal cord monitoring in patients with spinal deformity and neural axis abnormalities: a comparison with adolescent idiopathic scoliosis patients.

E Horaev B1, Surato D5, Serrafana S, McClung A, Van Allen E, Harrow P


Murrow RD1, McClung A5, Harrow P, Van Allen E, Serrafana S, Surato D5.
Postoperative Films
L.K.

- 19 y.o. woman with severe congenital scoliosis
  - Diastematomyelia
  - Large syrinx
  - Age 10 partial removal of diastematomyelia
    - Loss of signals, operation aborted
    - Inability to walk for one month
    - Full recovery
Clinical Photos
CT
MRI April 2011
Surgical Plan

- Thoughts?
- Remove diastematomyelia?
  - Shen et al SRS 2010
    - Type 1 vs. Type 2
- Syringomyelia?
- Ketamine to enhance signals
- Consider spinal cord shortening procedure
Corrective Surgery for Congenital Scoliosis Associated with Split Cord Malformation: It May Be Safe to Leave Diastematomyelia Untreated in Patients with Intact or Stable Neurological Status


- 73 patients type-I SCM, 141 type-II SCM
- Mean follow-up 37 mos (range, 24-108 mos)
- Rate of scoliosis correction was lower in type-I than in type-II (p < 0.05)
- 11 patients (5.1%) experienced transient complications but no significant difference between the 2 groups
- No permanent neurologic deficits
SRS 2013: Large Syrinx With Chiari

- SRS 2013
- Xie et al
- VCR shrinks syringomyelia
- Chiari?
Changes in CSF Flow After One-Stage Posterior VCR in Scoliosis Patients with Syringomyelia and Chiari Malformation Type I


- 8 patients with Chiari malformation, syrinx and severe scoliosis
  - No Chiari decompression undertaken
  - Flow determined with phase contrast cine MRI
- PSF with VCR
  - Improved CSF to almost normal at one year
Intraoperative

- T2 to L4 PSF
  - Osteotomies
  - Rib mass resection
- Intraoperative small MEPs, SSEPs
- T7 vertebrectomy with cage
- Prepared for
  - D-wave monitoring
  - Multiple wake-ups
    - After instrumentation
    - Correction
Postoperative
Key Points

- Prophylactic release in growing patients with tethered cord and scoliosis is warranted in some patients
- Simultaneous treatment of intraspinal anomaly and scoliosis correction feasible and efficacious
- Scoliosis improvement in patients with Chiari malformation most likely to occur in those < 10 years of age with curves < 35°
- Not all patients with split cord malformation will need neurosurgical intervention prior to correction of their scoliosis