What Every Spine Surgeon Should Know About Neurosurgical Issues

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Objectives

- Main intraspinal lesions
  - Chiari malformation
  - Tethered cord
  - Diastematomyelia
- Timing and effect of neurosurgical intervention
Chiari Malformation

- Chiari 1
  - Herniation of cerebellar tonsils below foramen magnum
  - Usually asymptomatic
  - Associated syrinx
- Chiari 2
  - Almost exclusively seen in children with myelomeningocele
  - Herniation of tonsils, vermis, 4th ventricle
- Chiari 3
- Chiari 4


Signs/Symptoms

- Anything that can be ascribed to the brain stem or craniocervical junction
  - Occipital headaches (80%)
  - Neck pain (70%)
  - Upper extremity paresthesias (50%)
  - Scoliosis

Tubbs et al, Childs Nerv Syst 2007
How common is scoliosis with Chiari 1?

<table>
<thead>
<tr>
<th>Study</th>
<th>No. of Patients</th>
<th>Rate of Scoliosis</th>
</tr>
</thead>
<tbody>
<tr>
<td>Navarro et al, 2004</td>
<td>94</td>
<td>15%</td>
</tr>
<tr>
<td>Tubbs et al, 2003</td>
<td>130</td>
<td>18%</td>
</tr>
<tr>
<td>Alzate et al, 2001</td>
<td>66</td>
<td>17%</td>
</tr>
<tr>
<td>Park et al, 1997</td>
<td>68</td>
<td>28%</td>
</tr>
</tbody>
</table>

How Common is a Chiari Malformation in Children with Scoliosis?

- Age dependent
- Infantile (0-3)
  - Dobbs, Lenke et al, JBJS 2002
    - 21.7% intraspinal anomaly
    - 10% Chiari
  - Pahys, Samdani et al, Spine 2010
    - 13% intraspinal anomalies
    - 4% Chiari
- Majority required surgical intervention
Incidence

- Adolescent idiopathic scoliosis
  - Do et al, JBJS 2002
    - 2% (7/327)
      - 1.2% Chiari
    - None required neurosurgical intervention
    - MRI reserved for high risk

Result of Chiari Decompression on Spine Deformity

- Eula et al, Spine 2002
- BrockMeyer et al, Spine 2003
- Flynn et al, Spine 2004
- Bangor et al, Childs Nerv Syst 2006
- Krieger et al, J Neurosurg Pediatr 2011
SRS 2009: The Natural History of Scoliosis Secondary to Chiari I Malformation and Syringomyelia after Suboccipital Decompression in Young Patients

Li Wei-guo, MD; Prof. Quiz Yong

- Largest series to date of 121 patients with scoliosis and Chiari decompression
- Progressors older than nonprogressors (14.2 vs. 9.2 years)
- Mean Cobb angle of 43° for progressors versus 31° for nonprogressors

Chiari with No Syrinx

- 12 yo girl with AIS
  - MRI probably not necessary
  - Do et al, JBJS 2002
  - Richards et al, Spine 2011
    - 529 patients
    - 6.8% with intraspinal anomalies
    - Risk factors: increased rotation and kyphosis
**MRI**

- ‘Low lying tonsils’
- No syrinx
- Underwent uneventful surgery

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**Chiari/No Syrinx And Scoliosis: Are They Related?**

- Very rare
- Tubbs *et al*, Childs Nerv Syst 2006
  - Case report
  - 13 degree right thoracic curve
- Krieger *et al*, JNS Pediatr, 2011
  - 79 children with scoliosis
  - All had Chiari with syrinx
Chiari with Syrinx

- 14 yo girl with a Lenke 2 curve
  - MRI revealed a Chiari with large syrinx
- Underwent Chiari decompression
- How long should one wait prior to deformity correction?
  - Repeat MRI 4-6 months to document decrease in syrinx

Witten et al, J Neurosurg Pediatr 2008
What if the syrinx does not resolve?

- Wait longer if deformity permits
  - Atenello et al, Neurosurgery 2008
    - Median time to resolution of syrinx = 10 months
- Drainage of syrinx
  - Morbidity
    - Aghakhani et al, Neurosurgery 2010
- Increased risk of not obtaining reliable neuromonitoring
Syrinx and Neurologic Outcomes

- Increased neurologic risk
  - Noorden et al, Spine 1994
  - Charry et al, J Pediatr Orthop 1994
  - Ozerdemoglu et al, Spine 2003

Outcome of Operative Treatment for Spinal Deformity in Patients with Syringomyelia: A Comparison Study to AIS Patients
Sucato et al, SRS 2011

- 38 patients with syringomyelia and scoliosis compared with 82 patients with AIS
- No differences in rate of obtainable neuromonitoring and similar correction
- However, no quantification of syrinx size
L.K.

• 19 yo female
  • Congenital scoliosis
  • Diastematomyelia
  • Syrinx
• Age 10 had resection of diastematomyelia

Clinical Photos
CT

MRI
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
Tethered Cord

- Conus ends at L1-L2
- Variable etiologies
  - Fatty filum
  - Lipoma
  - Myelomeningocele
    - All radiographically tethered

McClone Pediatr Neurosurg 1992
Tethered Cord

- 3 year old with presumed idiopathic scoliosis
- Neurologically nonfocal exam

MRI reveals a low-lying conus with a fatty filum

Recommend untethering

- Low morbidity
  - Bowman *et al*
  - *J Neurosurg Pediatr* 2009
Myelomeningocele and Tethered Cord
Hudgins & Gilreath, 2004

- All patients with MM radiographically tethered BUT 10-30% symptomatic
  - Weakness
  - Gait
  - Pain
  - Scoliosis
  - Worsening of foot and hip deformities
  - Urologic

Aim

- Is untethering necessary in the asymptomatic patient with MM prior to deformity correction?
Methods

- We retrospectively identified 19 patients with MM who had:
  - no evidence of a clinically symptomatic tethered cord
  - a spinal fusion for deformity correction
  - no untethering for at least one year prior to surgery
- Minimum follow-up after fusion was 2 years
- Charts and radiographs were reviewed for neurologic or shunt complications intra-op and within 3 months of surgery

Patient Demographics

<table>
<thead>
<tr>
<th>Total Patients</th>
<th>19</th>
</tr>
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<tbody>
<tr>
<td>Males</td>
<td>8</td>
</tr>
<tr>
<td>Females</td>
<td>11</td>
</tr>
<tr>
<td>Avg. age at surgery</td>
<td>12.2 years (10-17)</td>
</tr>
<tr>
<td>Follow-up</td>
<td>3.9 years (2-8)</td>
</tr>
<tr>
<td>Shunt present (%)</td>
<td>14/19 (74%)</td>
</tr>
<tr>
<td>Motor level</td>
<td></td>
</tr>
<tr>
<td>Thoracic</td>
<td>8</td>
</tr>
<tr>
<td>L1 or L2</td>
<td>7</td>
</tr>
<tr>
<td>L3</td>
<td>2</td>
</tr>
<tr>
<td>L4</td>
<td>2</td>
</tr>
</tbody>
</table>
Results

- Untethering 1 year prior to spine correction: 0%
- New neuro deficit: 1 patient *
- Pre-op major Cobb: 81°
- Post-op major Cobb: 33°

* Transient lower extremity weakness which returned to baseline within one month of surgery

Samdani et al Neurosurg Focus 2010

Conclusion

- Our results suggest that spinal cord untethering may not be necessary in all patients with myelomeningocele undergoing spinal deformity surgery
- Future studies with larger numbers are needed
Myelomeningocele: Always Consider the Shunt

- Majority of patients with MM will have a VP shunt
  - Talamonti et al
    J Neurosurg 2007
- Early postoperative death following deformity surgery from shunt malfunction
  - Geger et al Eur Spine J 2007

Patient KP

- 15 yo boy with MM and VPS
  - Severe curvature and pelvic obliquity
  - VPS placed at birth and not revised
- Multiple stage procedure planned
  - Stage 1: Halo, instrumentation, osteotomies
  - Stage 2: VCR with completion
Stage 1

- One hour after being positioned prone
  - Bradycardia with hypertension
- Emergent shunt tap performed
  - Elevated intracranial pressure
  - Surgery aborted
  - Head CT no change
  - Neurologically intact

Possible Reasons for Increased Intracranial Pressure

- Coincidental shunt malfunction
  - Unlikely
- Increased abdominal pressure resulting in shunt malfunction
  - Miele et al 2004 Neurosurgery
Options

- Cancel surgery
- Externalize the shunt
  - Would require multiple neurosurgical procedures
- Intracranial monitoring and CSF drainage
- Procedure completed
  - Required CSF drainage to maintain normal ICP

Recommendations

- Consider baseline HCT
- Aggressive bowel prep pre-op
- Abdomen free during surgery
- Prep patient in a manner to allow easy access to the shunt
Diastematomyelia

- Split cord syndrome
  - Associated with congenital scoliosis
  - Type 1: two separate dural sacs
  - Type 2: one dural sac
- Can cause tethering

Patient GF

- 12 yo boy with congenital scoliosis, diastematomyelia
  - Laminectomy and partial resection of diastematomyelia at age 4
  - Progressive, severe scoliosis and increased pain with ambulation
Options

- Considered resection of diastematomyelia; however…
  - Shen *et al* SRS 2010
    - 95 patients with diastematomyelia underwent deformity surgery
    - None prophylactically removed
    - No neurological injuries
Chiari malformation with syrinx should be decompressed. A repeat MRI 4-6 months obtained to document a decrease in syrinx size.

Not all MM patients need to be dethered prior to scoliosis correction.

Not all patients with a diastat need removal prior deformity correction.