Neurosurgical Management of Intraspinal Lesions: Considerations for the Deformity Surgeon

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Objectives

- Main intraspinal lesions
  - Chiari malformation
  - Tethered cord
  - Diastematomyelia
- Patient with myelomeningocele
- 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 MM
  - Herniation of tonsils, vermis, 4th ventricle
- Chiari 3
- Chiari 4

Hankinson *et al* Neurosurg Clin North Am 2007

Case Presentation

- 8 yo girl presented with 30° right thoracic curve
  - MRI scan demonstrated Chiari malformation with holocord syrinx
- Underwent Chiari decompression
- What are chances of curve improvement or stabilization?

Result of Chiari Decompression on Spine Deformity

Eula *et al* Spine 2002

Brock Meyer *et al* Spine 2003

Flynn *et al* Spine 2004

Bangor *et al* Childs Nervous System 2006

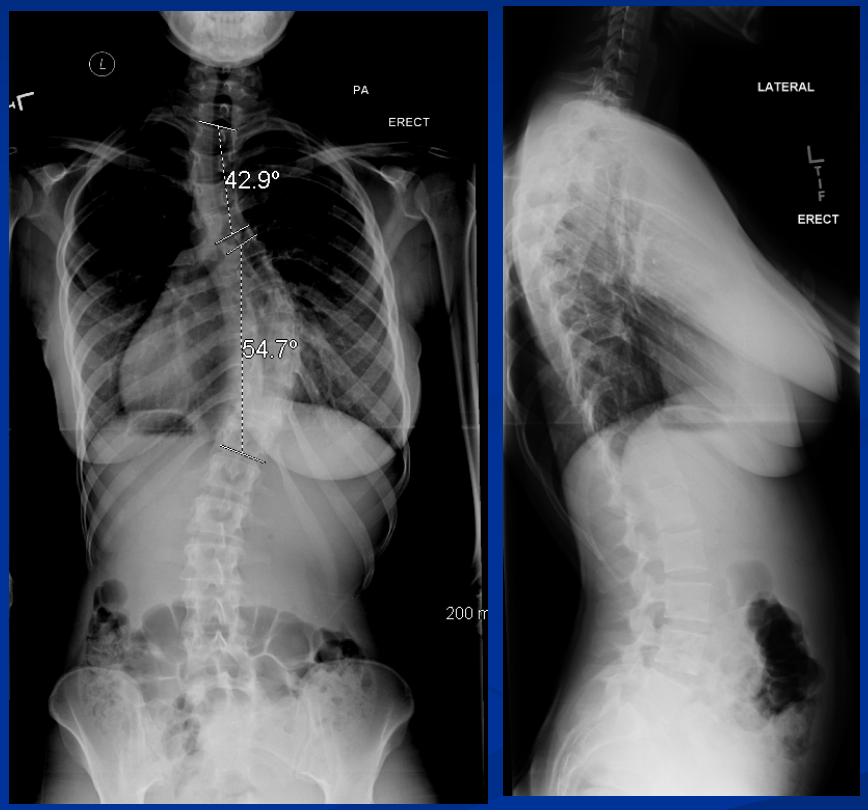
SRS 2009: The Natural History of Scoliosis Secondary to Chiari I Malformation and Syringomyelia after Suboccipital Decompression in Young Patients

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)
- Progressors mean Cobb angle of 43° versus 31° for nonprogressors

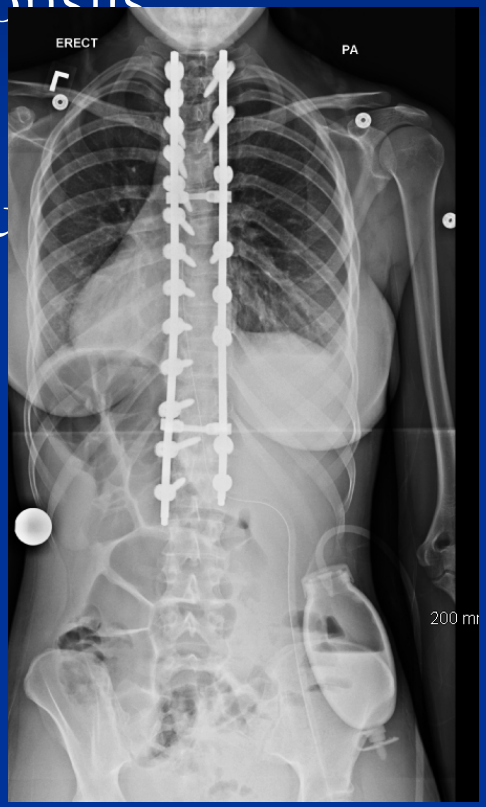
Chiari with No Syrinx

- 12 yo girl with AIS
  - MRI probably not necessary
  - Do *et al* JBJS 2002
    - 2% (7/327)
    - None required neurosurgical intervention
    - MRI reserved for high risk
  - Richards *et al* Spine 2011
    - 529 patients
    - 6.8% with intraspinal anomalies
    - Risk factors: increased rotation and kyphosis



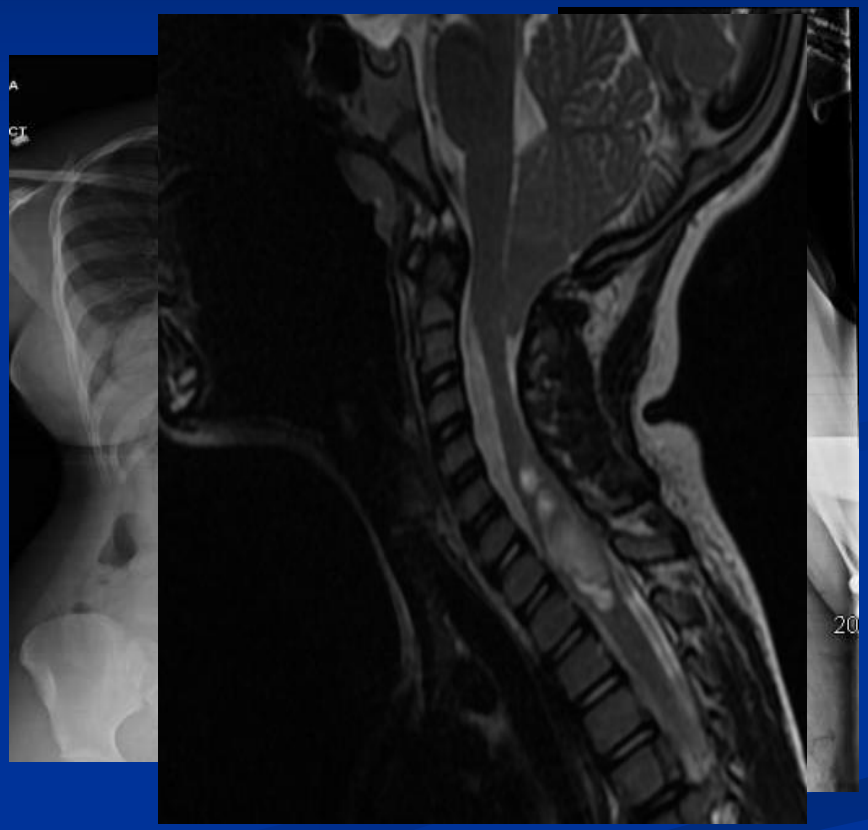
MRI

- ‘Low lying tonsils’



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

Postoperative Films

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

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
- Very high incidence of intraspinal anomalies
  - Dobbs *et al* JBJS 2002
    - 21.7% (10/46) neural axis abnormalities
  - Philadelphia Shriners
    - Pahys, Samdani, Betz Spine 2009
      - 13% (7/54)

Tethered Cord

- MRI reveals a low-lying conus with a fatty filum
- Recommend untethering
  - Low morbidity
    - Bowman *et al*

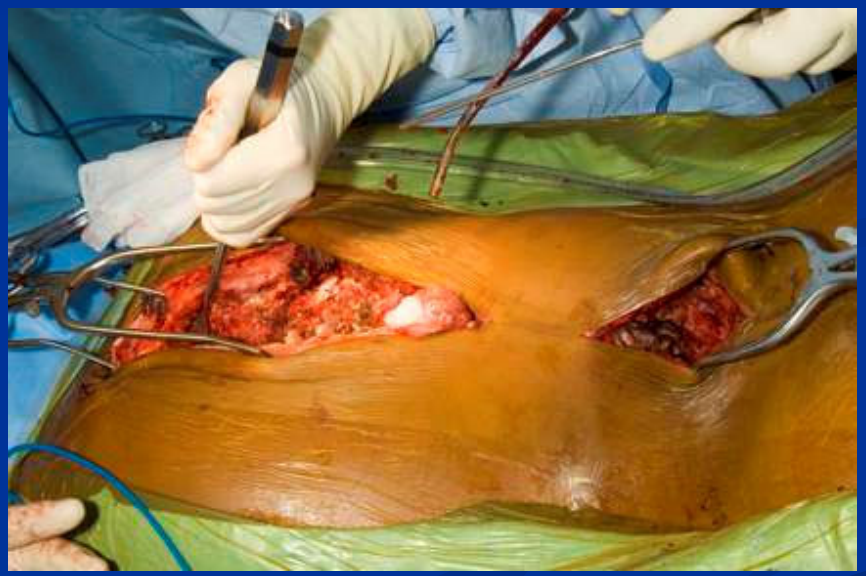
      J Neurosurg Pediatr 2009

Intraspinal Anomaly and Scoliosis Correction

- Timing of surgery
  - Traditionally 2 stages operated on three months apart
    - CSF leak
    - Two hospitalizations
    - Risk of retethering

Intraspinal Anomaly and Scoliosis Correction

- Improved neuromonitoring allows for one stage procedures
  - Samdani *et al* Spine 2007
  - Hamzaoglu *et al* Spine 2007
    - 21 patients with congenital scoliosis/kyphosis
    - No infections, neurologic deficits



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

Outcomes Following Detethering

- Bowman *et al* J Pediatr Neurosurg 2009
  - 114 patients
- Pierz *et al* J Pediatr Orthop 2000
  - 21 patients
- Herman *et al* Pediatr Neurosurg 1993
  - 153 patients
- Fone *et al* J Urol
  - 39 patients
- Reigel *et al* Pediatr Neurosurg 1994
  - 262 patients
- Palmer *et al* J Urol 1998

Untethering in Patients with MM

- Risks
  - Neurologic worsening
  - CSF leak
  - Wound problems

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

Total Patients	19
Males	8
Females	11
Avg. age at surgery	12.2 years (10-17)
Follow-up	3.9 years (2-8)
Shunt present (%)	14/19 (74%)
Motor level *Thoracic* *L1 or L2* *L3* *L4*	 8 7 2 2

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 you 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

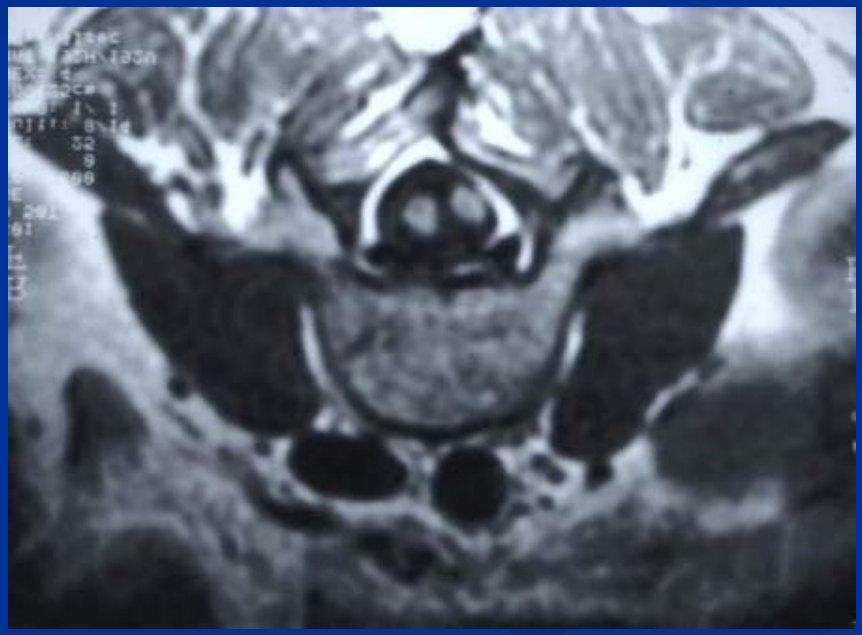
Recommendations

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

Samdani et al *JBJS in press*

Diastematomyelia

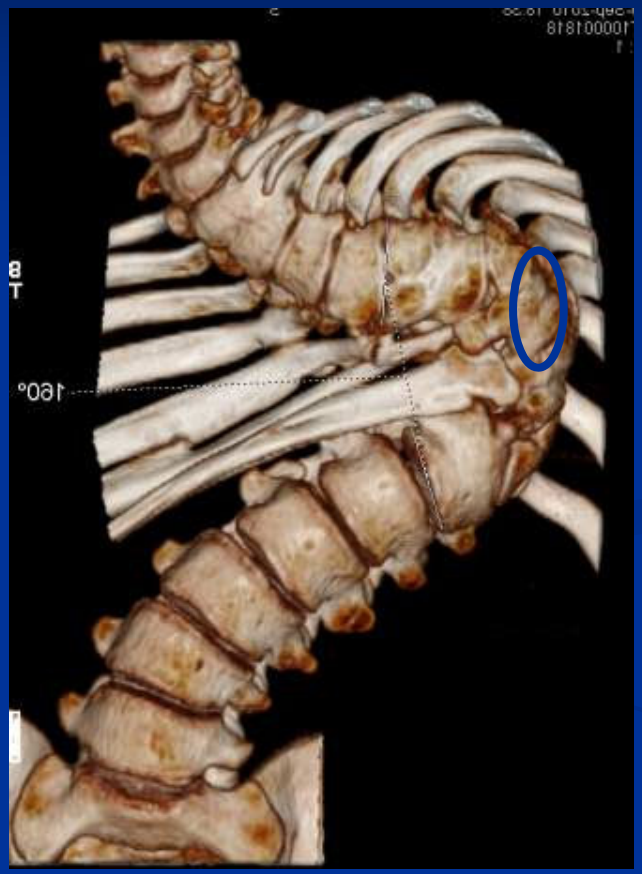
- Split cord syndrome
  - Associated with congenital scoliosis
  - Type 1: two separate dural sacs
  - Type 2: one dural sac
- Can cause tethering



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

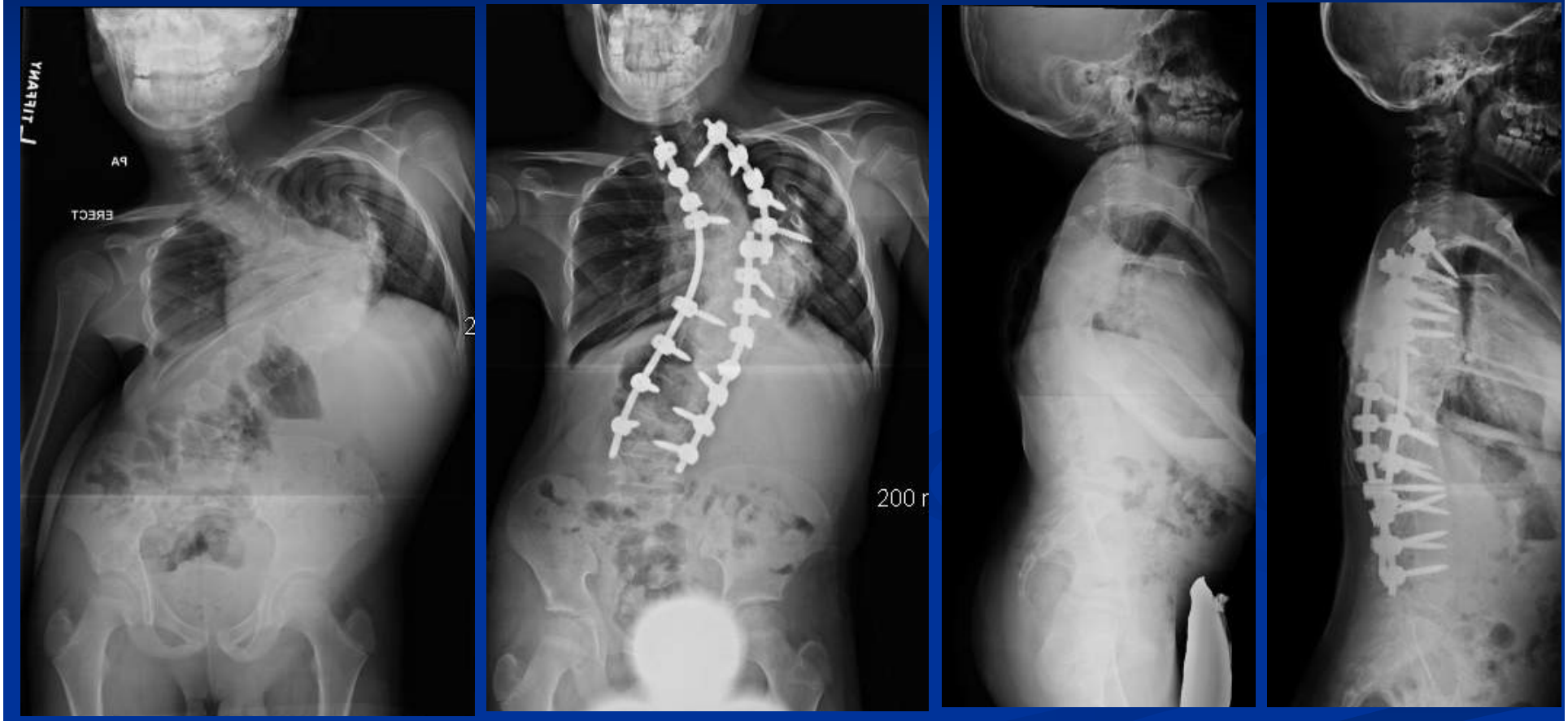

PA
ERECT


180°

Options

- Considered resection of diastematomyelia; however…
  - Shen *et al* SRS 2010
    - 95 patients with diastematomyelia underwent deformity surgery
    - None prophylactically removed
    - No neurological injuries

Post-op X-rays



Summary

- 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.