What Every Spine Surgeon Should Know About Neurosurgical Issues… When to Refer, When Not to Worry

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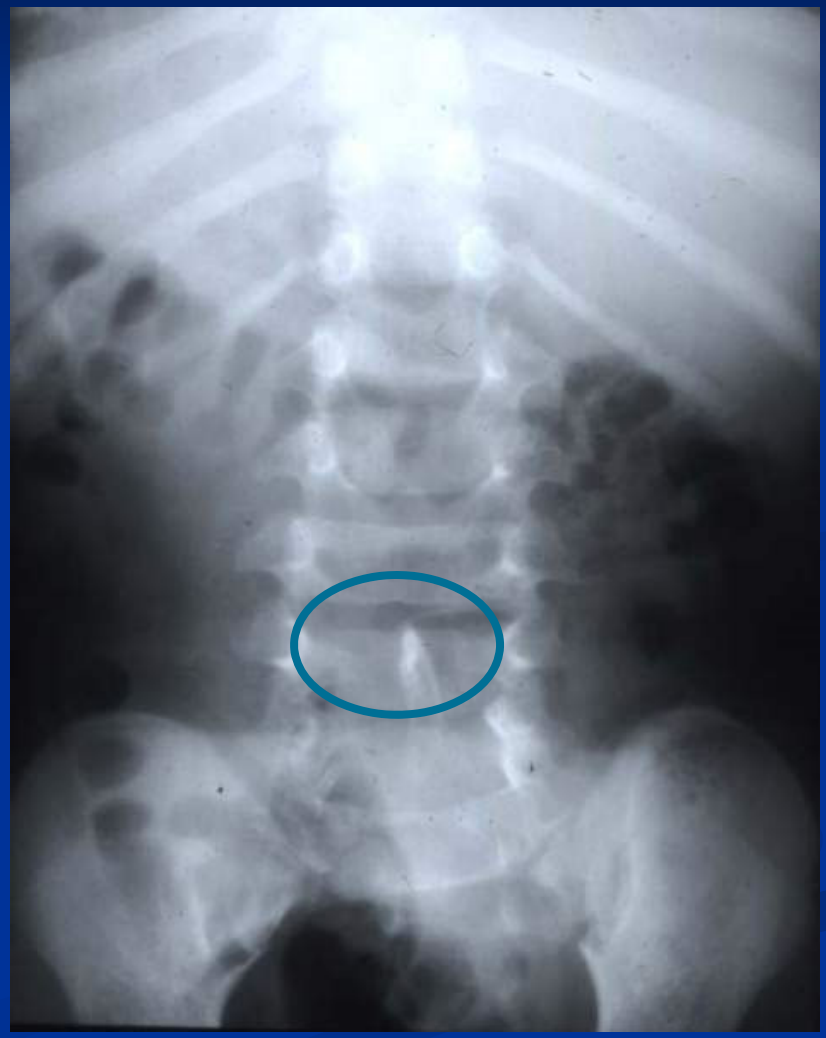
Objectives

- Incidence and most common anomalies seen
  - Chiari malformation
    - Syrinx
  - Tethered cord
  - Split cord malformation (diastematomyelia)
- Patient presentations

Incidence

- Congenital: 20-60% incidence
  - McMaster, JBJS 1984
    - 251 patients with congenital scoliosis
    - 46 (18.3%) with anomaly
    - Split cord malformation most common



Congenital Scoliosis: Incidence

- Shen et al, Spine 2013
  - 226 patients
    - 43% with intraspinal anomaly
      - Split cord malformation most common
- Basu et al, Spine 2002
  - 126 consecutive patients
    - Tethered cord most common
  - More common in:
    - Kyphosis, complex defects

Isolated Hemivertebra
Belmont et al, JBJS 2004

TABLE I Prevalence of Intraspinal Anomalies and Diagnostic Value of History and Physical Examination Findings

Group	No. of Patients	Intraspinal Anomaly *(No. of Patients)*	Accuracy *(%)*	Sensitivity *(%)*	Specificity *(%)*	Predictive Value *(%)*	
						Positive	Negative
Isolated hemivertebra	29	8 (28%)	62	63	62	38	81
Complex pattern	47	10 (21%)	77	50	84	45	86
Overall	76	18 (24%)	71	56	76	42	85

Isolated Hemivertebrae
Belmont et al, JBJS 2004

- 8 patients underwent neurosurgical intervention
- Only 4 had abnormal physical examination findings
- ".. *a magnetic resonance imaging evaluation of the entire spine should be considered for all patients with congenital scoliosis, including those with an isolated hemivertebra*."

Syrinx and Chiari Malformation

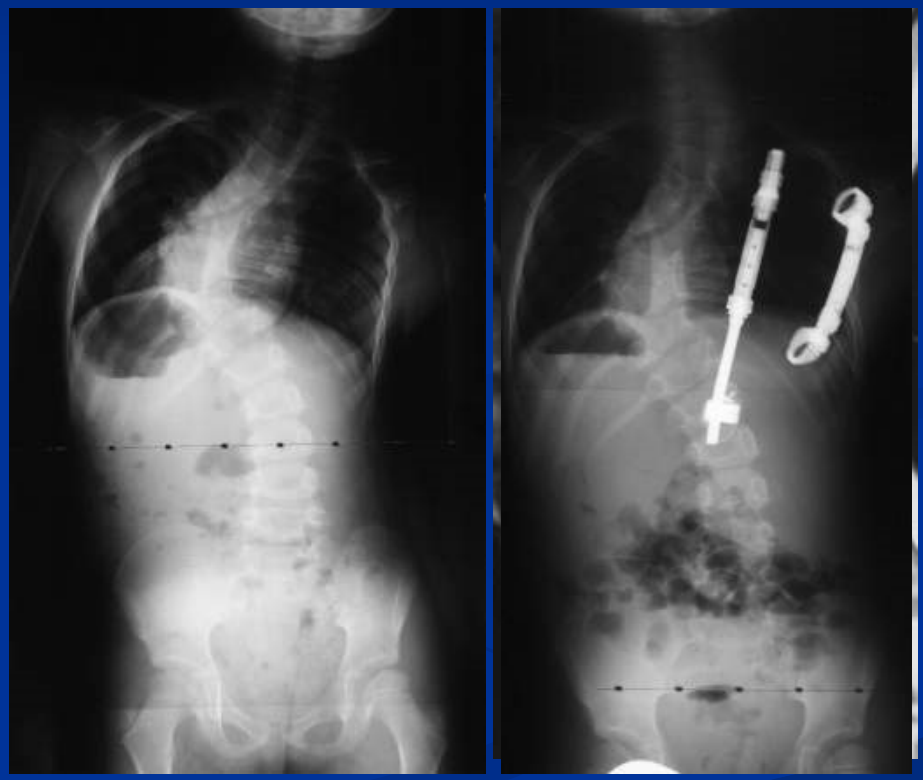
Outcome of Operative Treatment for Spinal Deformity in Patients with Syringomyelia: A Comparison Study to AIS Patients

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

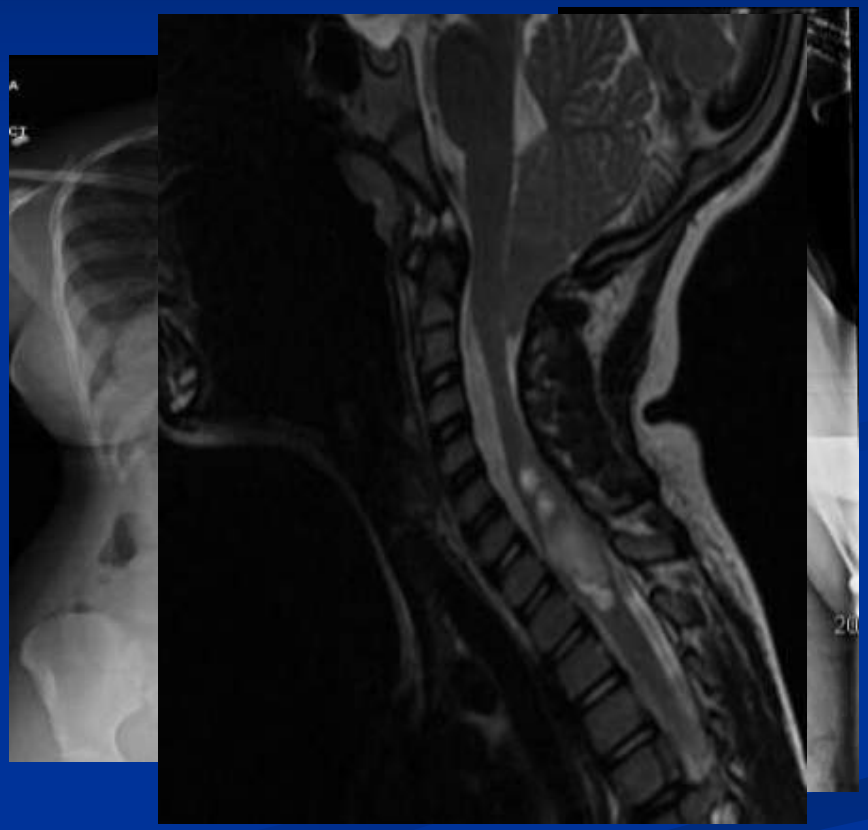
Small Syrinx: Patient CL

- 5 yo with progressive congenital scoliosis and fused ribs
  - Preoperative MRI shows small syrinx
    - No Chiari
  - Likely no further treatment: ? Up to 4mm a dilated central canal
  - Consider repeat MRI in 6 months prior to implants
  - Uneventful surgery



Patient LK: Large Syrinx with Chiari

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



Witten *et al,* J Neurosurg Pediatr 2008

Syrinx resolution after posterior fossa decompression in patients with scoliosis secondary to Chiari malformation type I

Tao Wu · Zezhang Zhu · Jian Jiang · Xin Zheng · Xu Sun · Bangping Qian · Feng Zhu · Yong Qiu

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Abstract

Introduction Description of syrinx resolution after posterior fossa decompression (PFD) in patients with scoliosis secondary to Chiari malformation type I (CMI) and syringomyelia (SM) has been rarely reported in the literature. This study was performed to investigate the outcome of PFD in patients with scoliosis secondary to CMI and to identify potential predictive factors for better outcome after PFD.

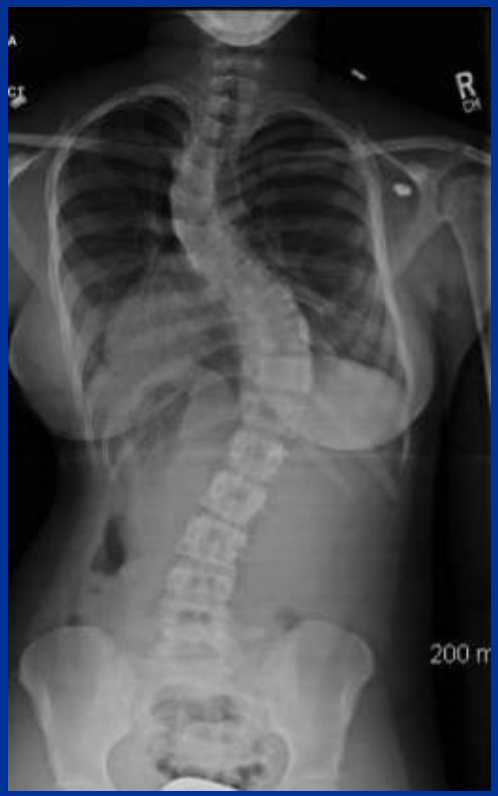
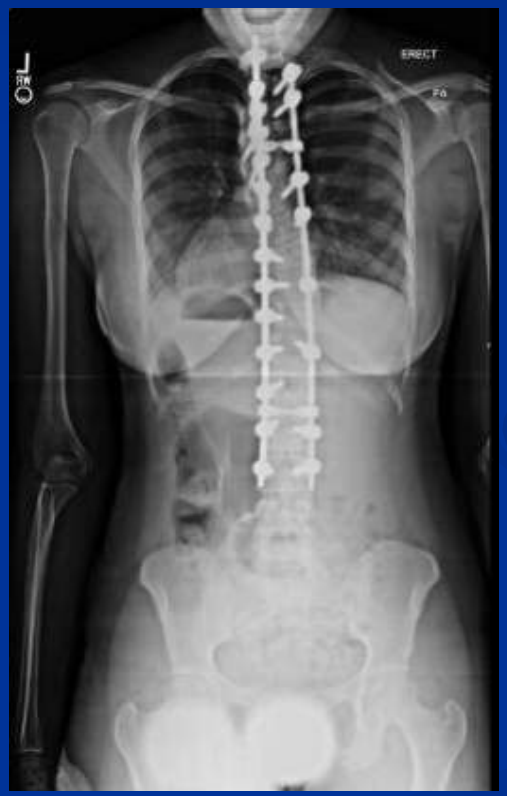
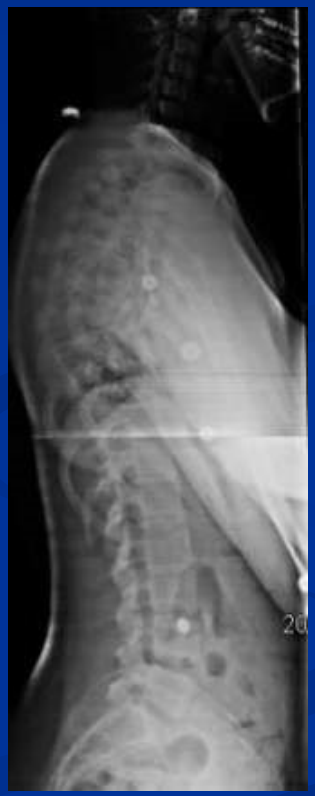
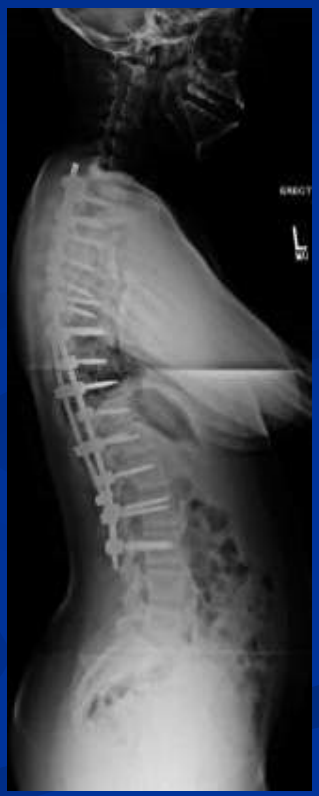
Material and methods Patients with scoliosis secondary to CMI and SM, who had undergone PFD during the period 2000 through 2009, were recruited. Inclusion criteria were (1) age $\leq$ 18 years, (2) diagnosis of SM associated with CMI, (3) scoliosis as the first complaint, (4) having undergone preoperative and follow-up magnetic resonance imaging (MRI). Patients with acquired CMI anomalies or who had received syringosubarachnoid shunting were excluded. The maximal S/C ratio and syrinx length were measured to evaluate syrinx resolution after PFD. A 20% decrease in S/C ratio or length at the latest follow-up was defined as a significant radiographic improvement and complete resolution was used to describe the syrinx disappearing after PFD.

Results 44 patients were recruited. Follow-up MRI was conducted for all 44 patients at 6 $\pm$ 3 months postoperatively, for 37 patients at 2 years $\pm$ 3 months, for 26 patients at 4 years $\pm$ 3 months, and for 15 patients at 6 years $\pm$ 3 months. 97.7% (43 of 44) of patients showed significant radiographic improvement by MRI. The distance of tonsillar descent (mm) was correlated significantly with the surgical outcome ($r = 0.116$, $P = 0.013$). Significant improvement was observed within 6 months postoperatively, with continued slow improvement after that.

Conclusion Syringes showed significant improvement after PFD in most patients with scoliosis secondary to CMI. Resolution generally occurred within 6 months follow-up and continued at a slow rate for several years. In addition, the severity of tonsillar descent is a potential predictor for better improvement after standard PFD.

Keywords Resolution · Syrinx · Chiari malformation type I · Syringomyelia · Scoliosis · Posterior fossa decompression

Postoperative Films

What if the syrinx does not resolve?

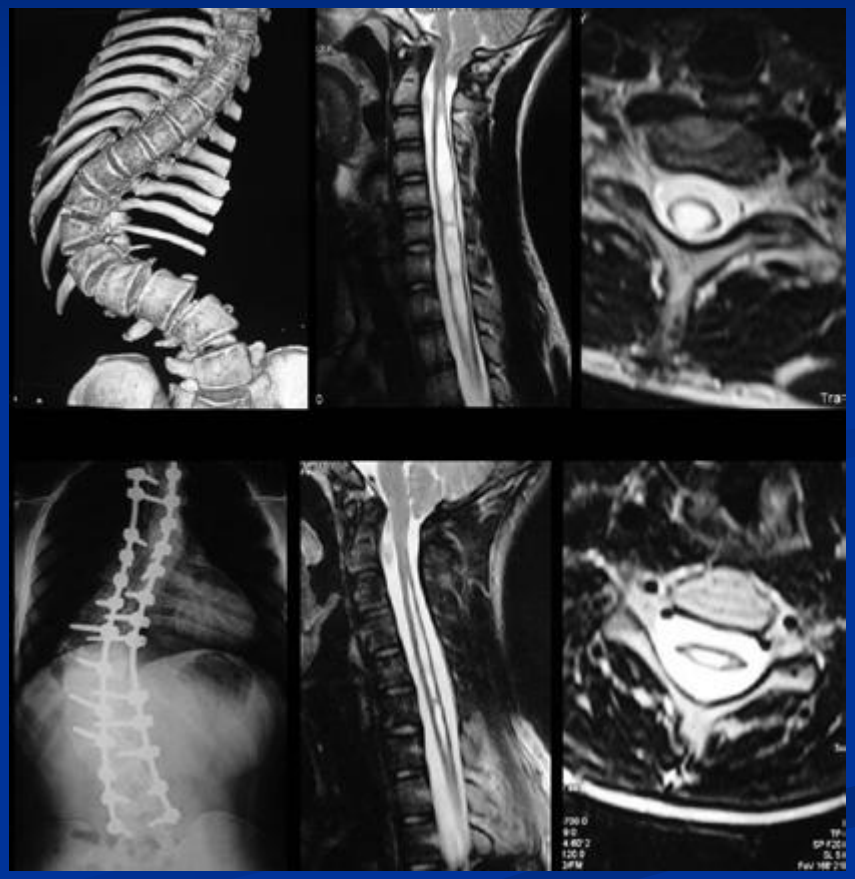
- Wait longer if deformity permits
  - Atenello *et al,* Neurosurgery 2008
    - Median time to resolution of syrinx = 10 months
- Drainage of syrinx
  - Morbidity
  - Aghakhani *et al,* Neurosurgery 2010

Syrinx and Neurologic Outcomes

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

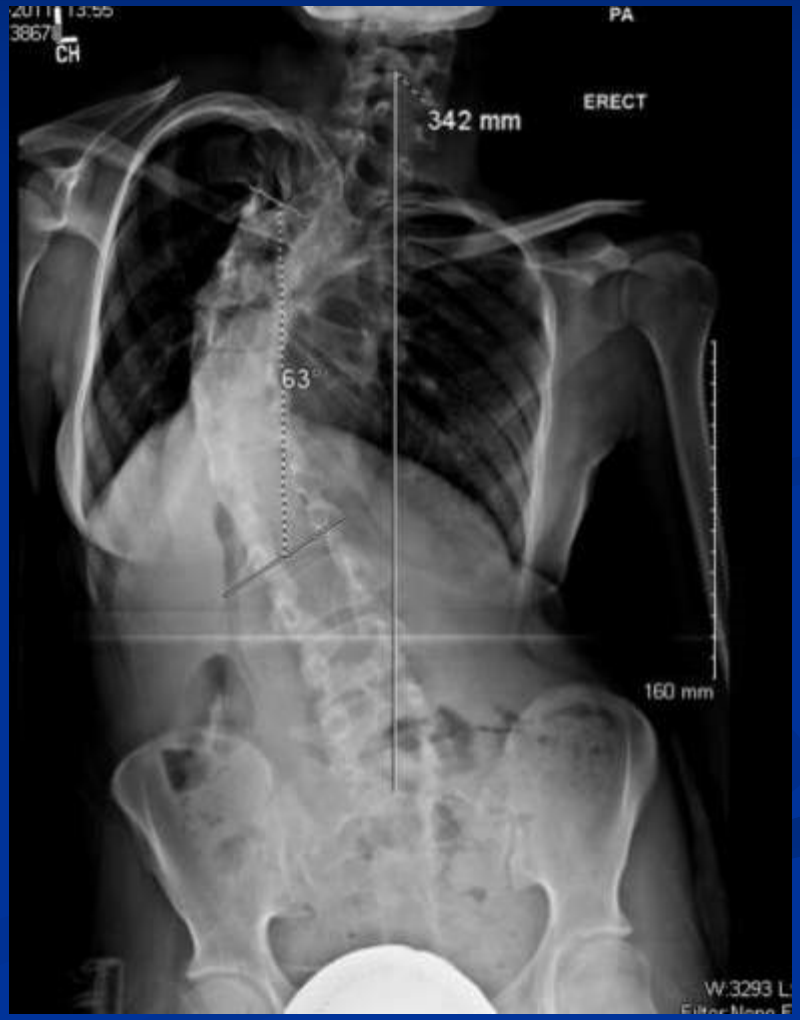
Large Syrinx Without Chiari

- Variable approach
  - Drain syrinx?
- SRS 2013
  - Xie et al
  - VCR shrinks syringomyelia
    - Chiari?



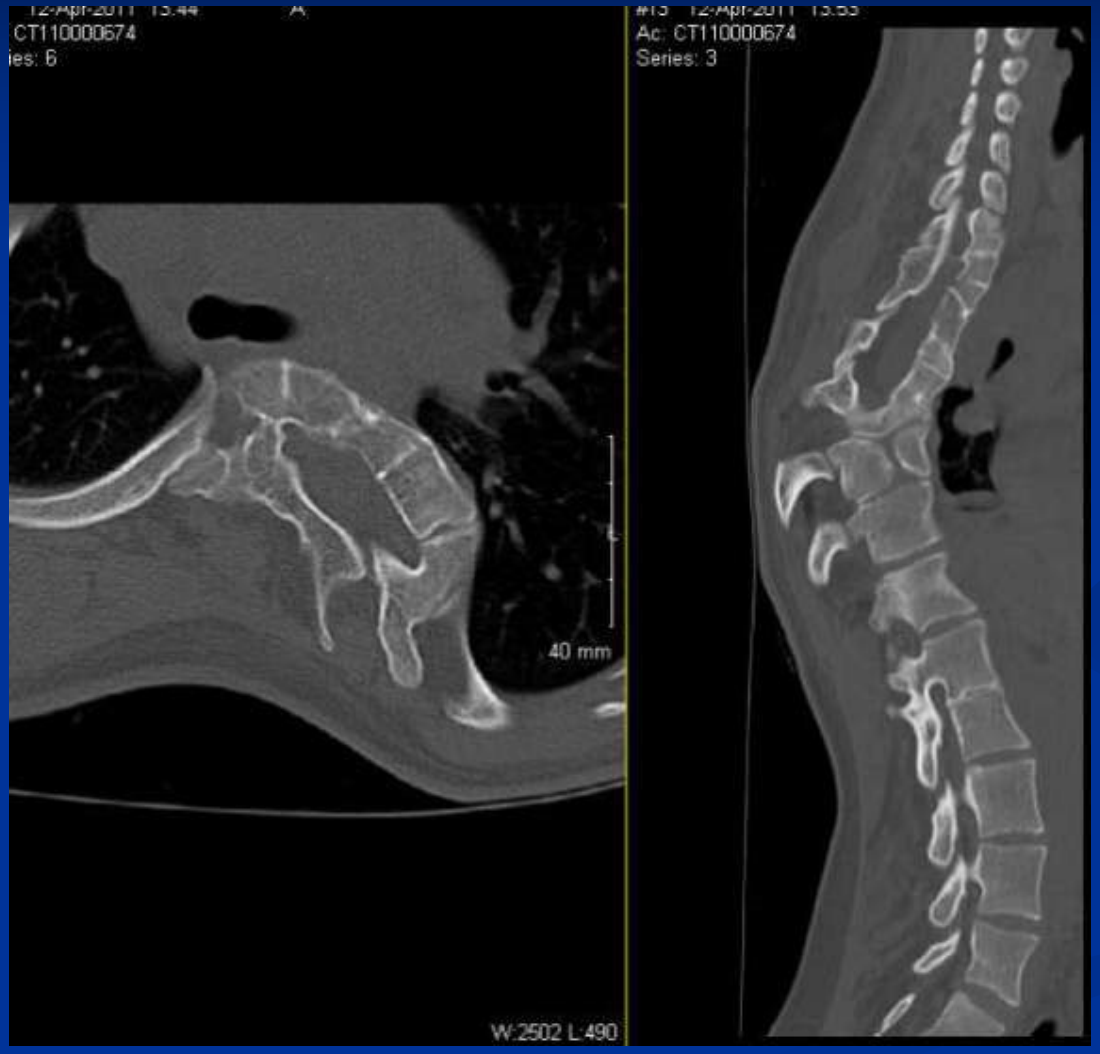
L.K.

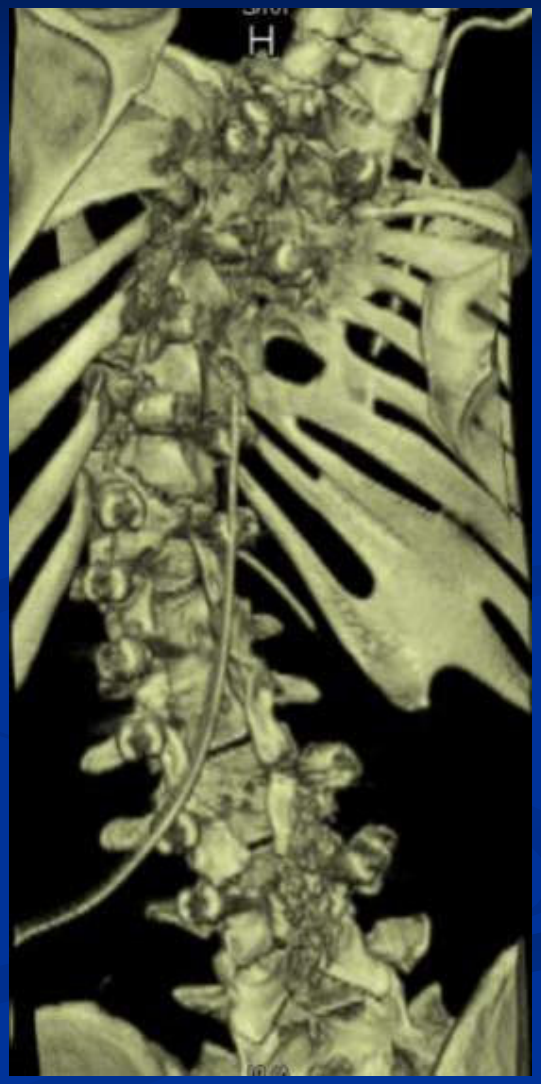
- 19 yo girl
  - Congenital scoliosis
  - Split cord malformation with bony spur
  - Syrinx
- Had resection of bony spur at age 10



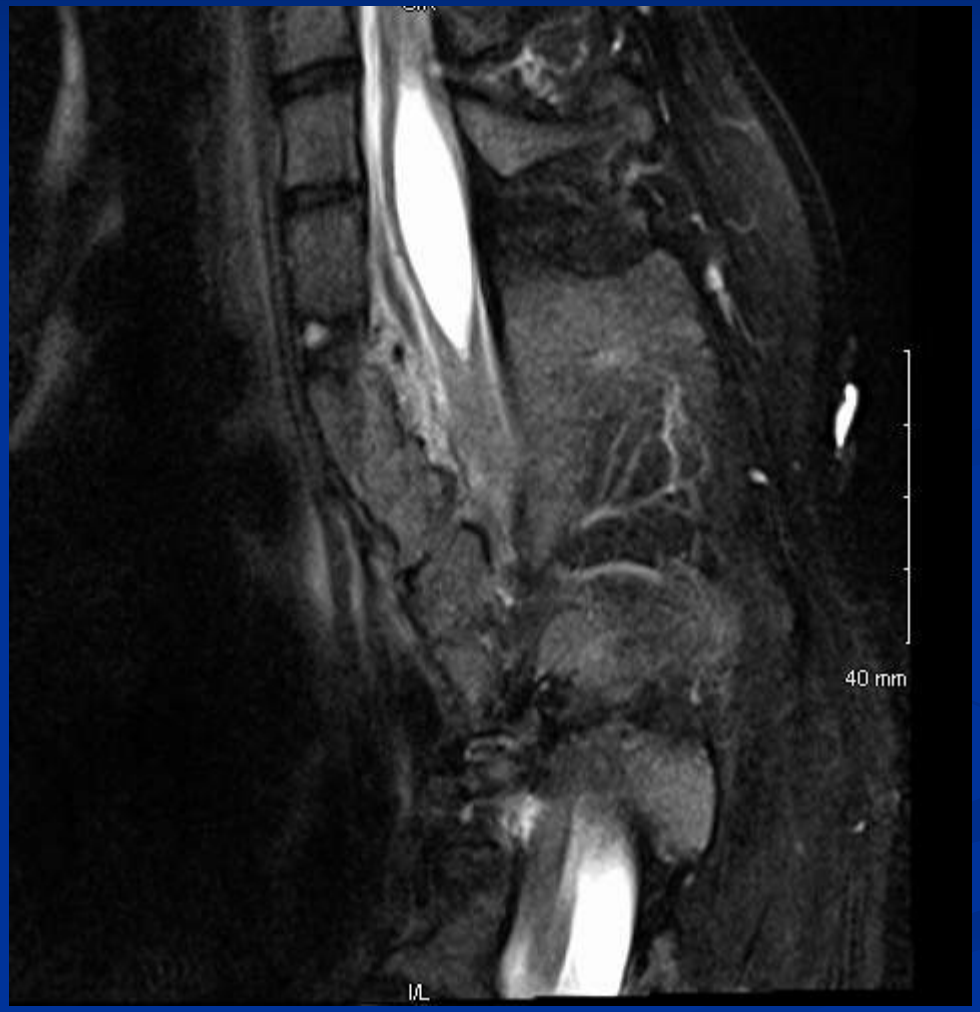
Clinical Photos

CT



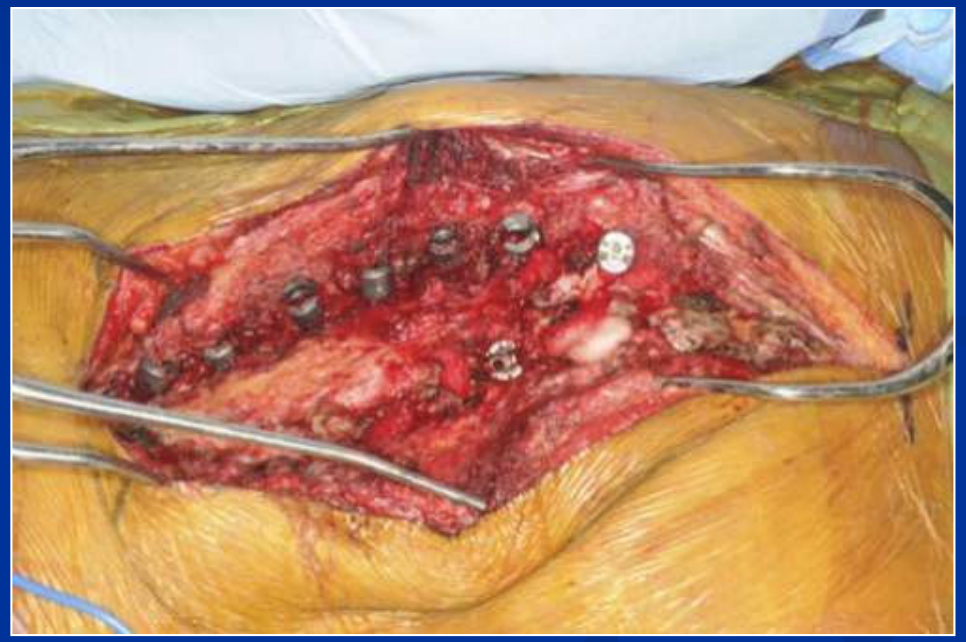


MRI

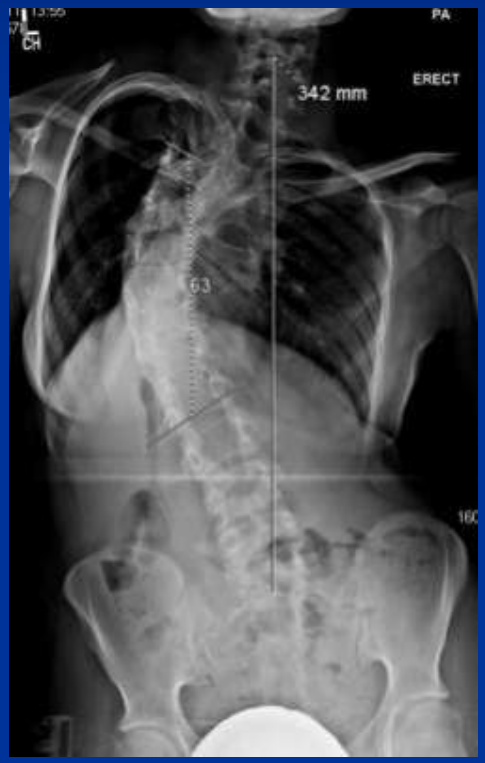


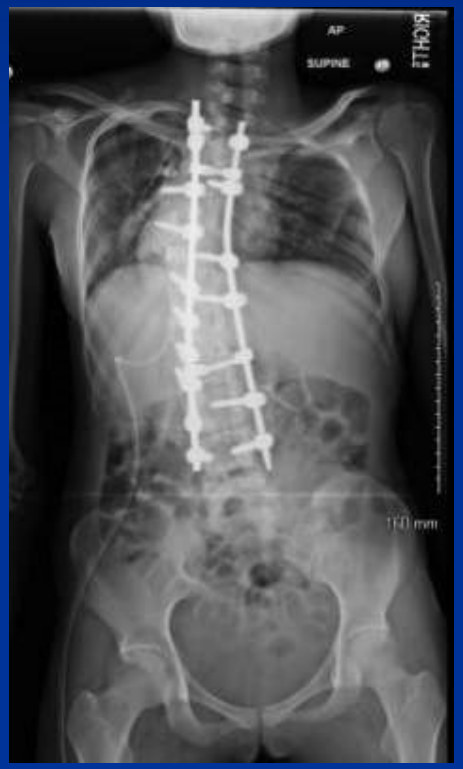
Intraoperative

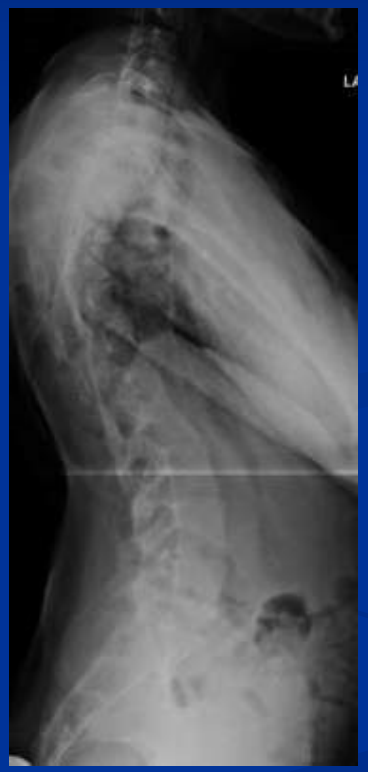
- T2 to L4 PSF
  - Osteotomies
  - Rib mass resection
- Intraoperative small MEPs, SSEPs
- T7 vertebrectomy with cage
- Prepared for
  - D- wave monitoring
  - Multiple wake-ups
    - After instrumentation
    - Correction

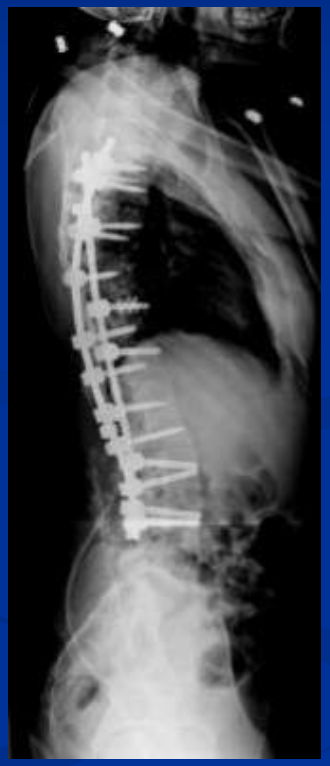


Postoperative



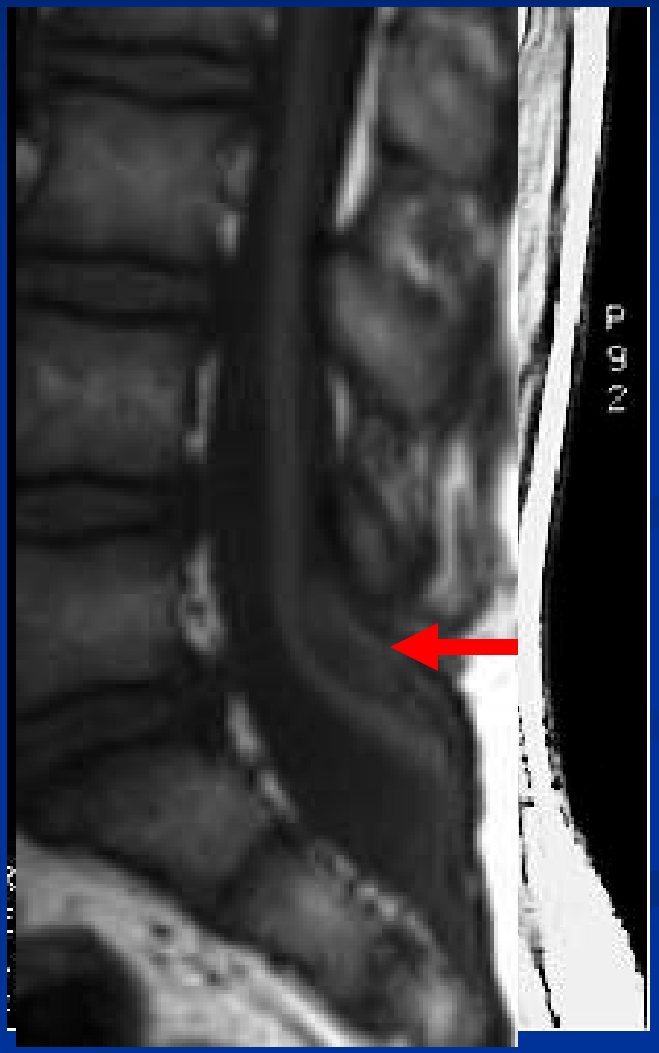






Tethered Cord

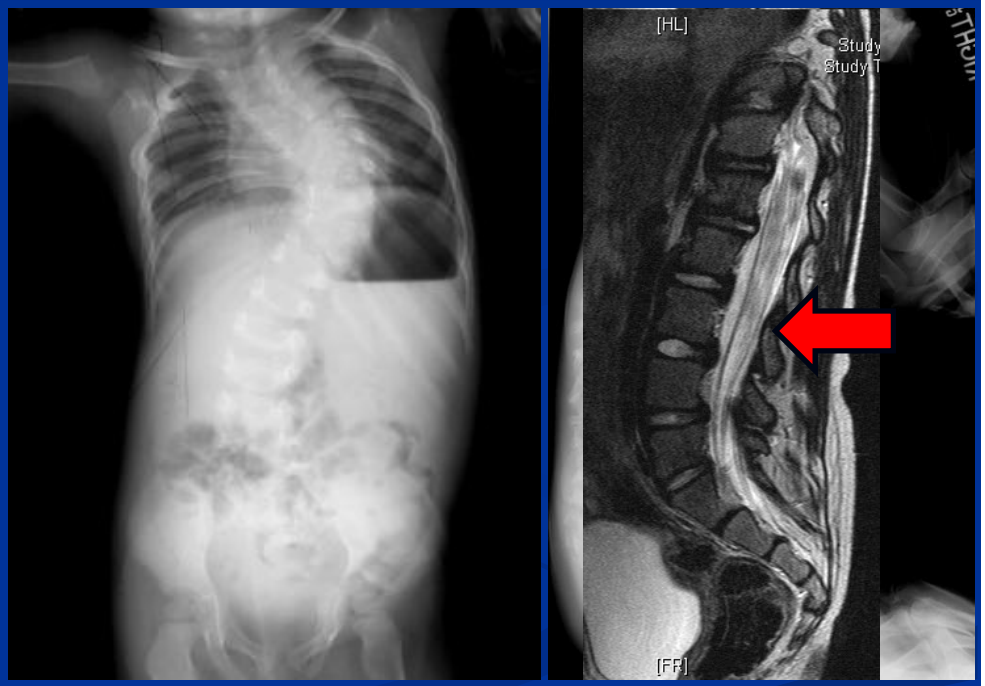
- Conus ends at L1-L2
- Variable etiologies
  - Fatty filum
  - Lipoma
  - Myelomeningocele
    - All radiographically tethered



McClone, Pediatr Neurosurg 1992

Patient CL: Tethered Cord

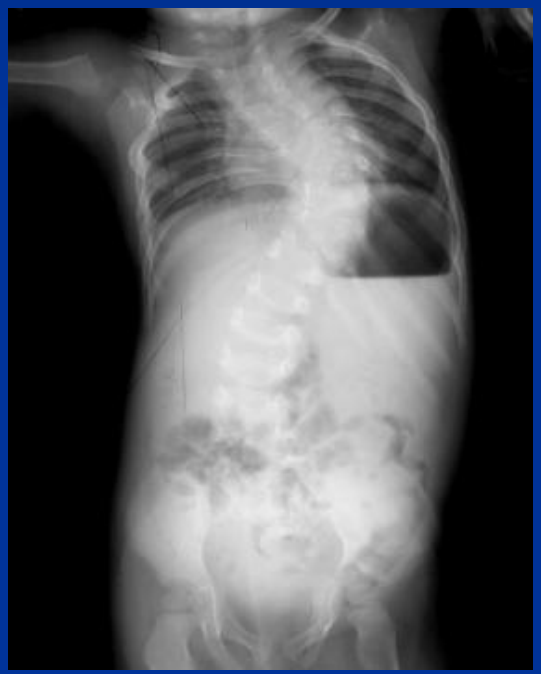
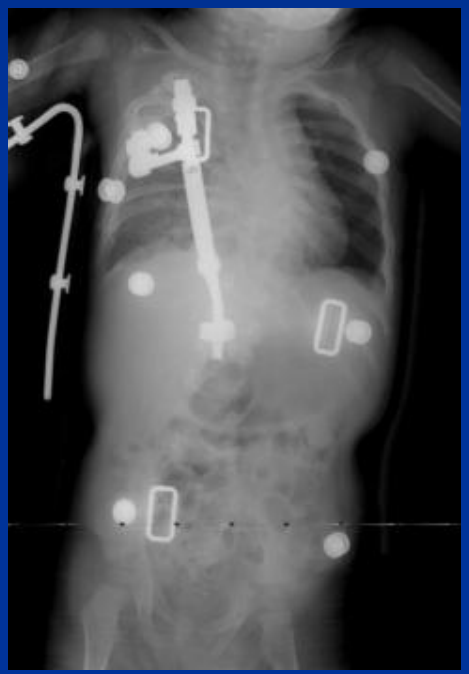
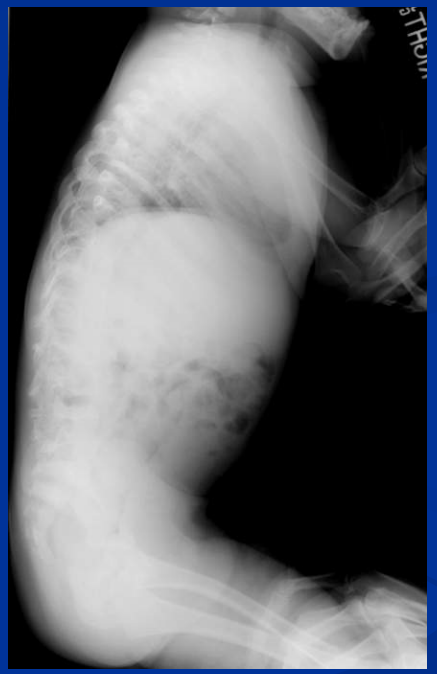
- 3 yo with progressive congenital scoliosis
- Neurologically non-focal exam
- MRI
  - ‘Low lying cord’ with fatty filum



Fatty Filum with Low Lying Conus

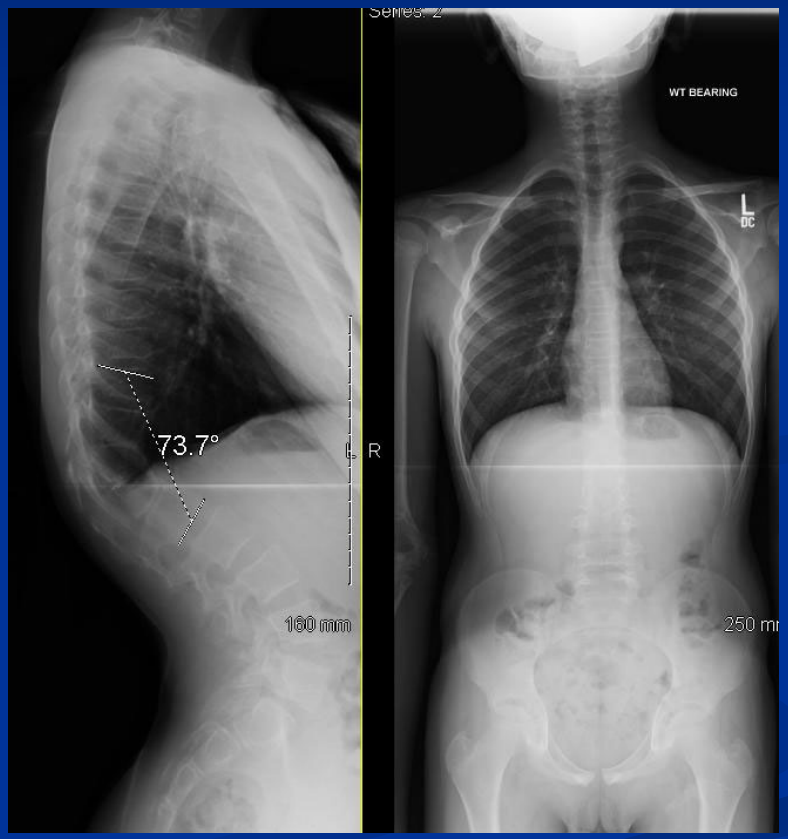
- Recommend untethering
  - Low morbidity
    - Bowman et al, J Neurosurg Pediatr 2009
- Family feels everything done

Uneventful Surgery

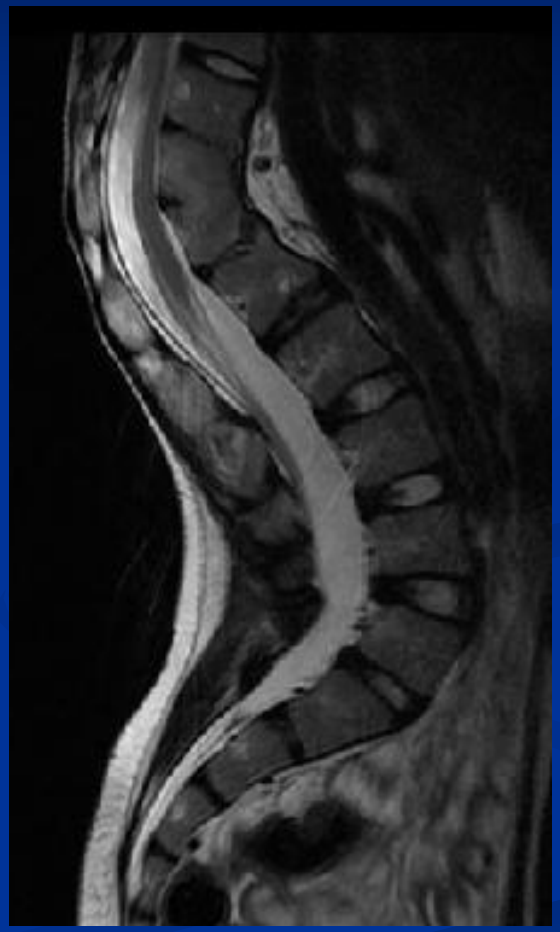

Patient NM

- 12 yo boy with congenital kyphosis
  - Plain films
    - 73º kyphosis T8 to L1
    - Anterior bar T10-12
  - MRI
    - No cord signal change
    - Low lying conus



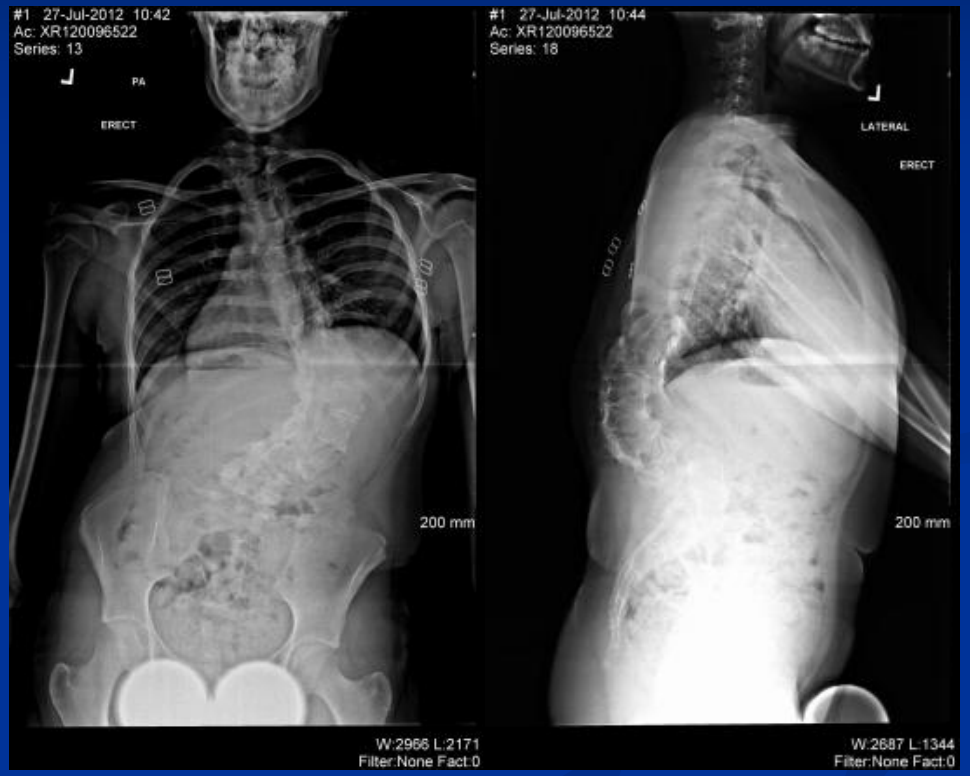
Patient NM

- Unclear if need to untether
- My preference is to leave alone as vertebral column shortening



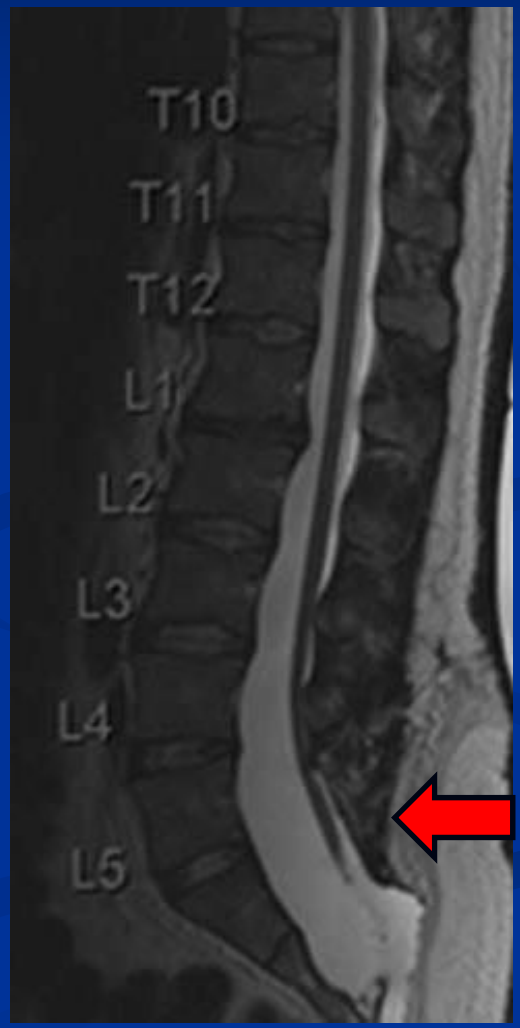
Patient EM

- 15 yo girl with a history of congenital scoliosis
- At 20 months of age she had A/P in situ fusion T10-L2 at outside hospital
- Several untetherings for lipoma
- Symptoms
  - Pain, progression

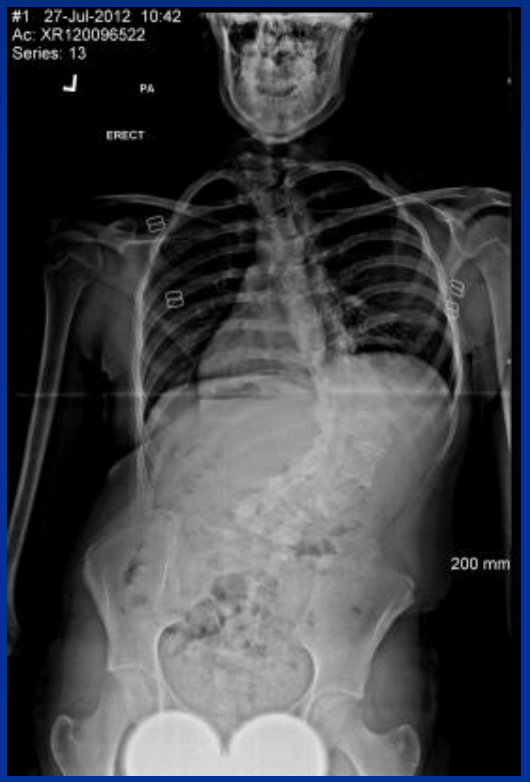


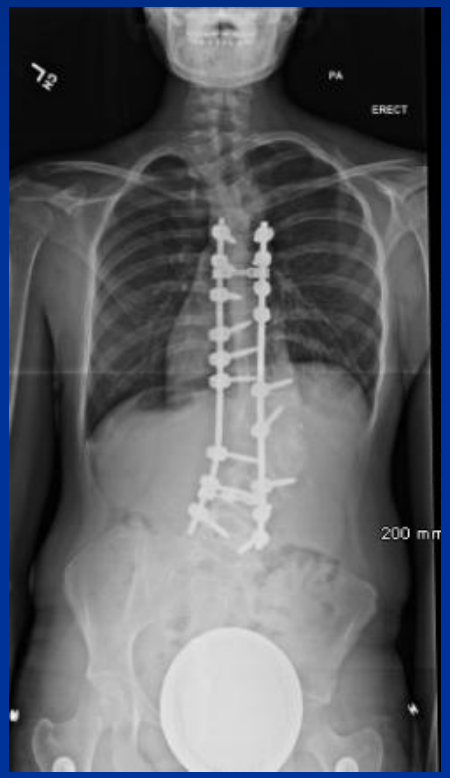
Options

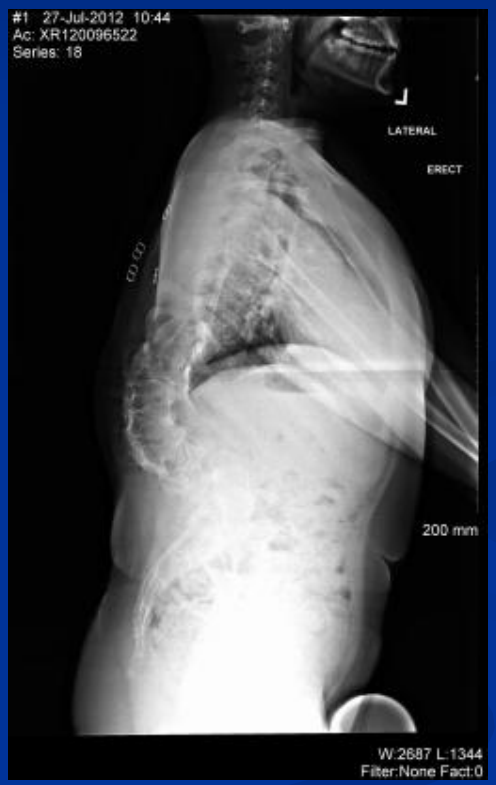
- Continued untethering
  - Neurologic risk
  - Risk of retethering
    - Surgery may be too long in same stage
- Vertebral column shortening
  - Deformity correction
  - Treatment for tethered cord
    - Hsieh et al, J Neurosurg 2009
    - Matsumoto et al, Spine 2009

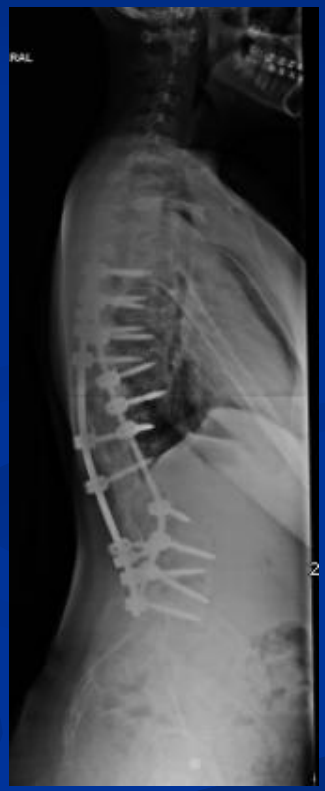


Postoperative





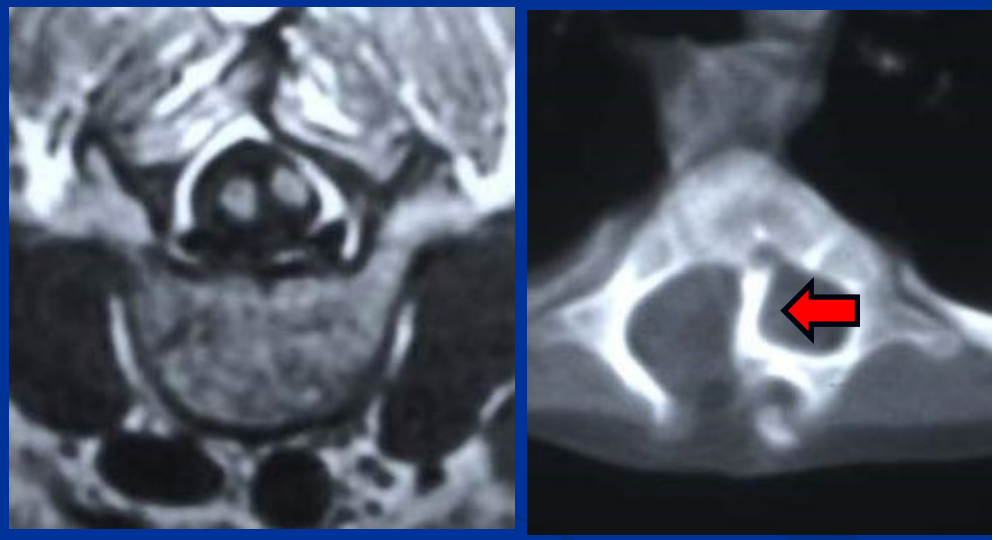




Patient EM

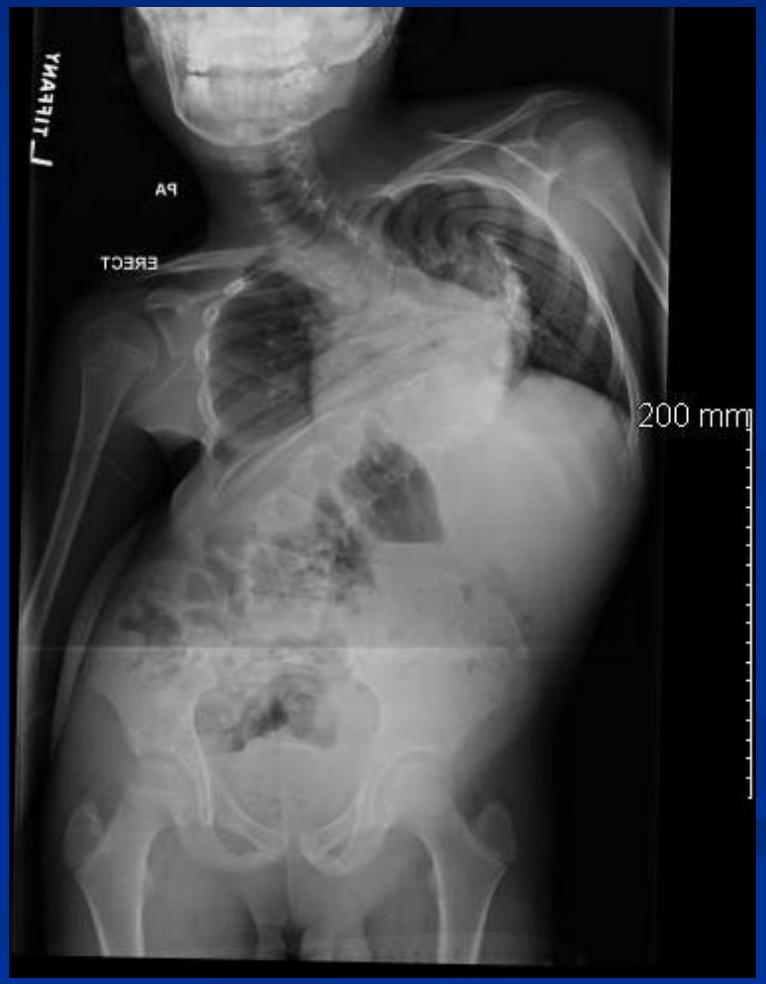
Split Cord Malformation (Diastematomyelia)

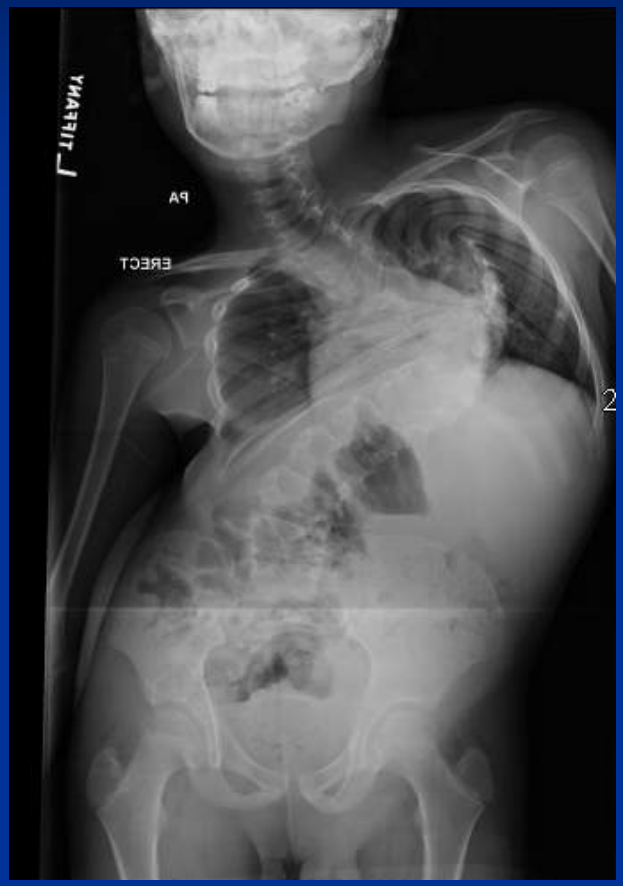
- Split cord syndrome
  - Associated with congenital scoliosis
  - Type 1: two separate dural sacs, with septum
  - Type 2: one dural sac with two cords
- Can cause tethering



Patient GF

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

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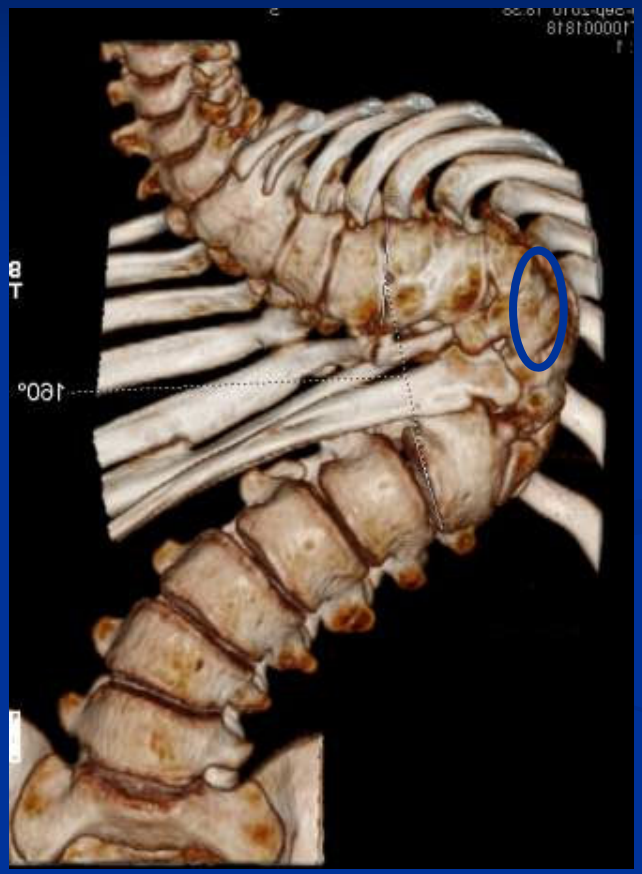
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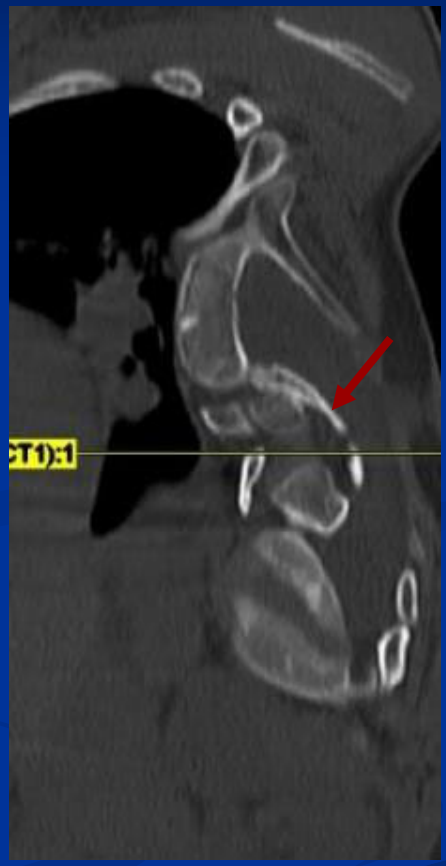
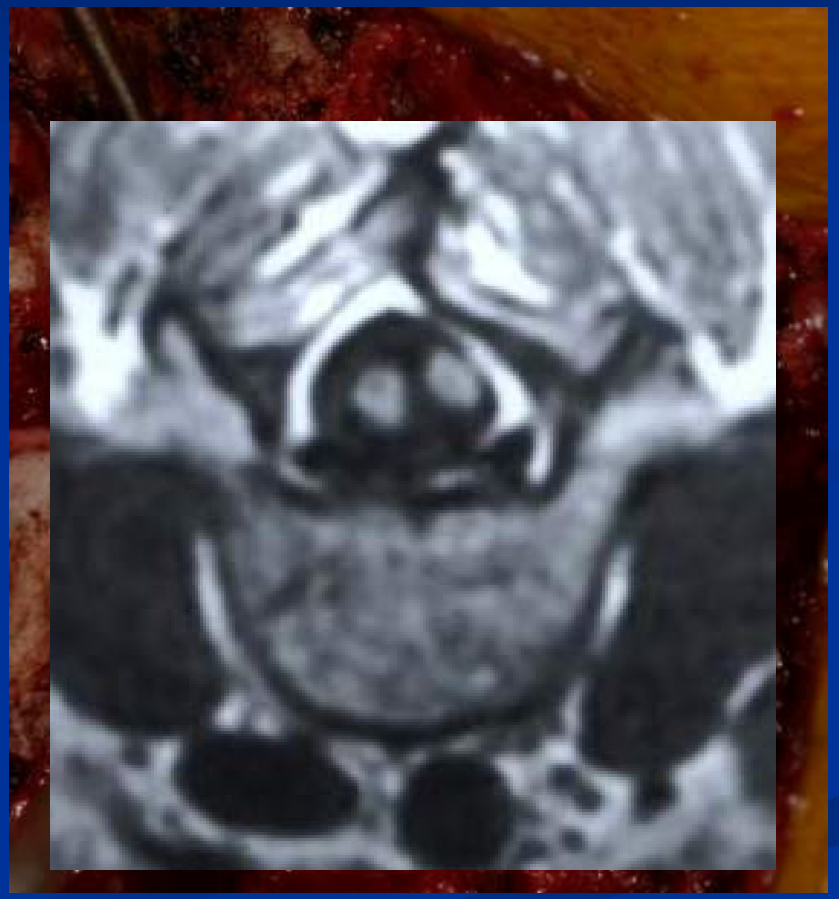


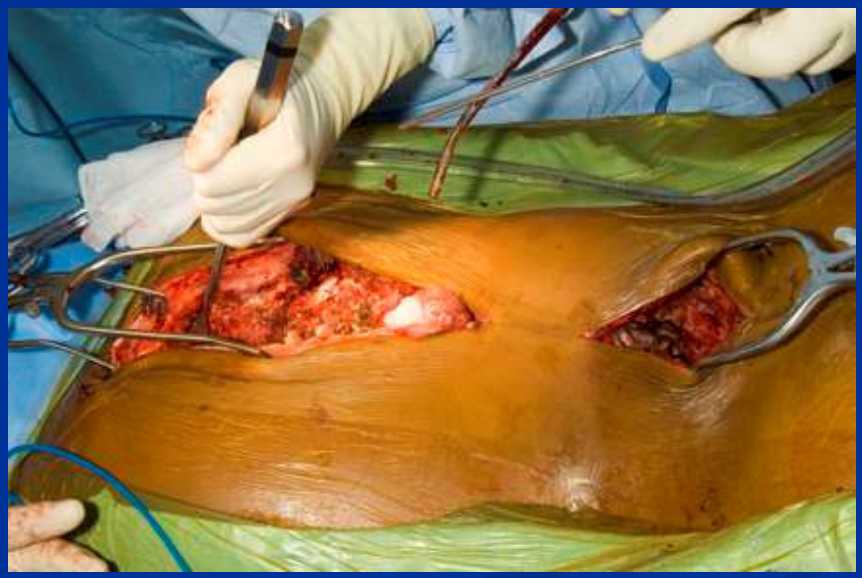
Options

- Considered reexploration of split cord malformation; however…
  - Davya et al, Spine 2009
    - 32 patients with SCM
  - Hui et al, Spine 2012
    - 45 patients with SCM
- Type 1: resect
- Type 2: leave alone

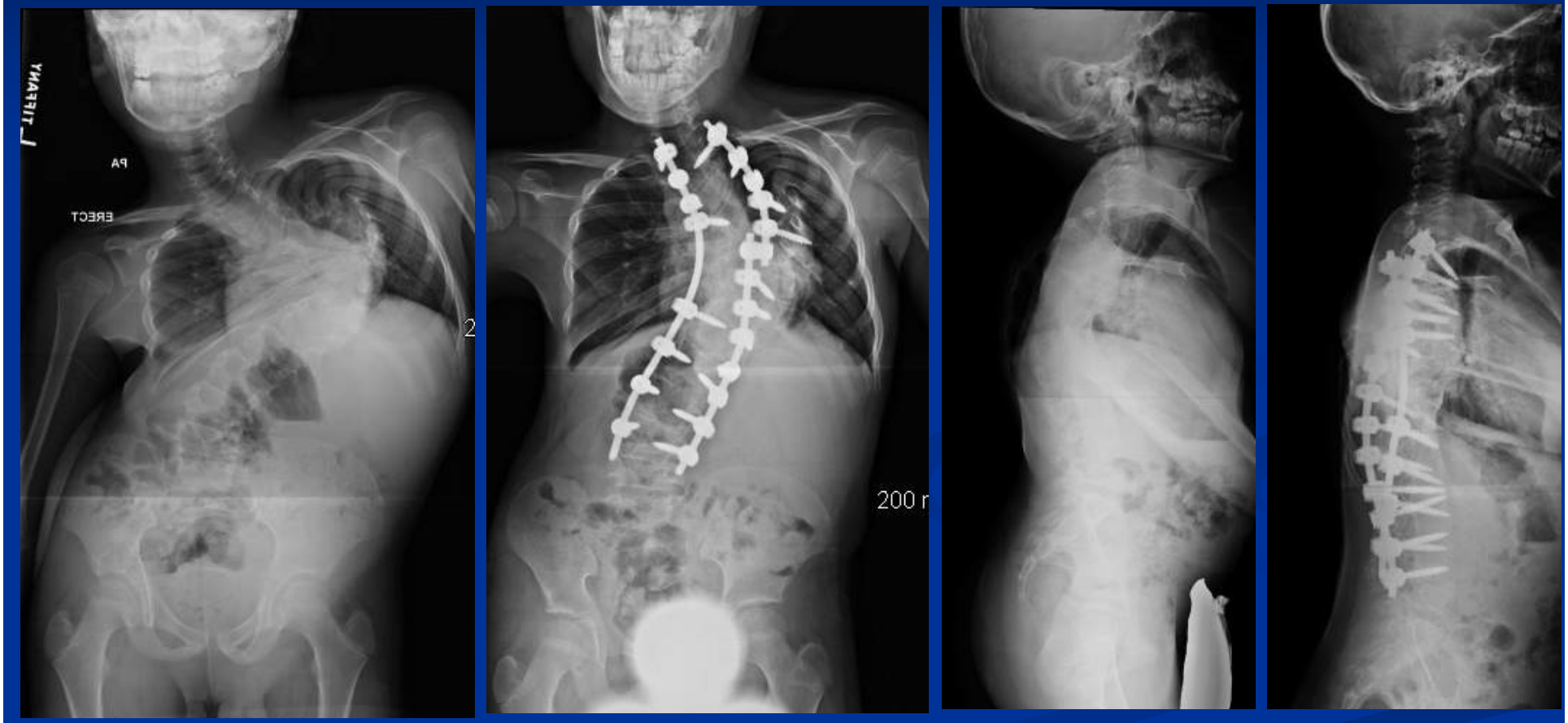


Intraspinal Anomaly and Scoliosis Correction

- 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



Post-op X-rays



Congenital Scoliosis and Neurologic Risk

- Hwang et al, JNS Pediatr 2013
- Possible reasons
  - ? Vascular anomalies to spinal cord
  - Vitale et al, JBJS 2010
    - Cardiopulmonary comorbidity risk factor
  - Mik et al, “Diminished spinal cord size associated with congenital scoliosis of the thoracic spine,” JBJS 2009

Summary

- Small syrinx without Chiari can likely be left alone, although a repeat MRI to demonstrate nonprogression should be considered
- Split cord malformation type 2 may not need to be treated prior to congenital spine deformity
- For complex anomalies spinal cord shortening may be safest