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

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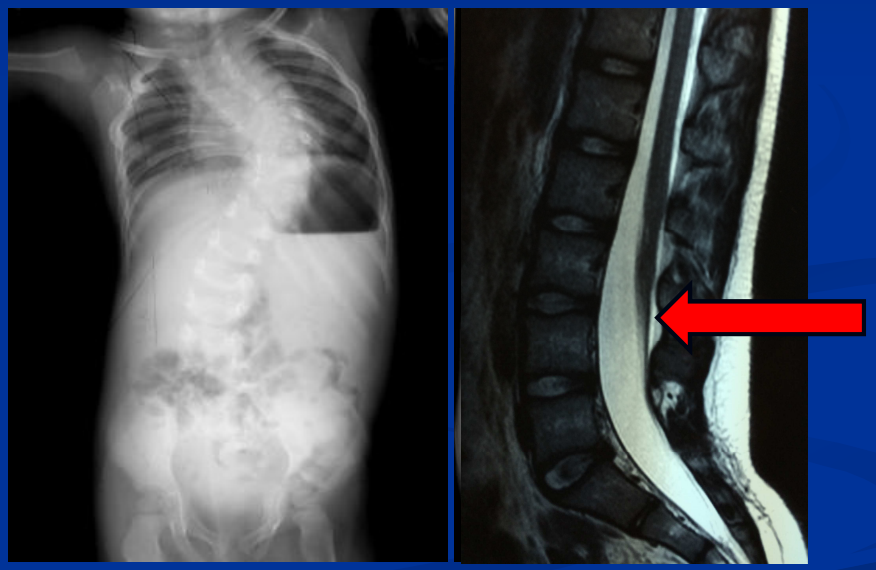
Intraspinal Anomalies and Spine Deformity

- 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

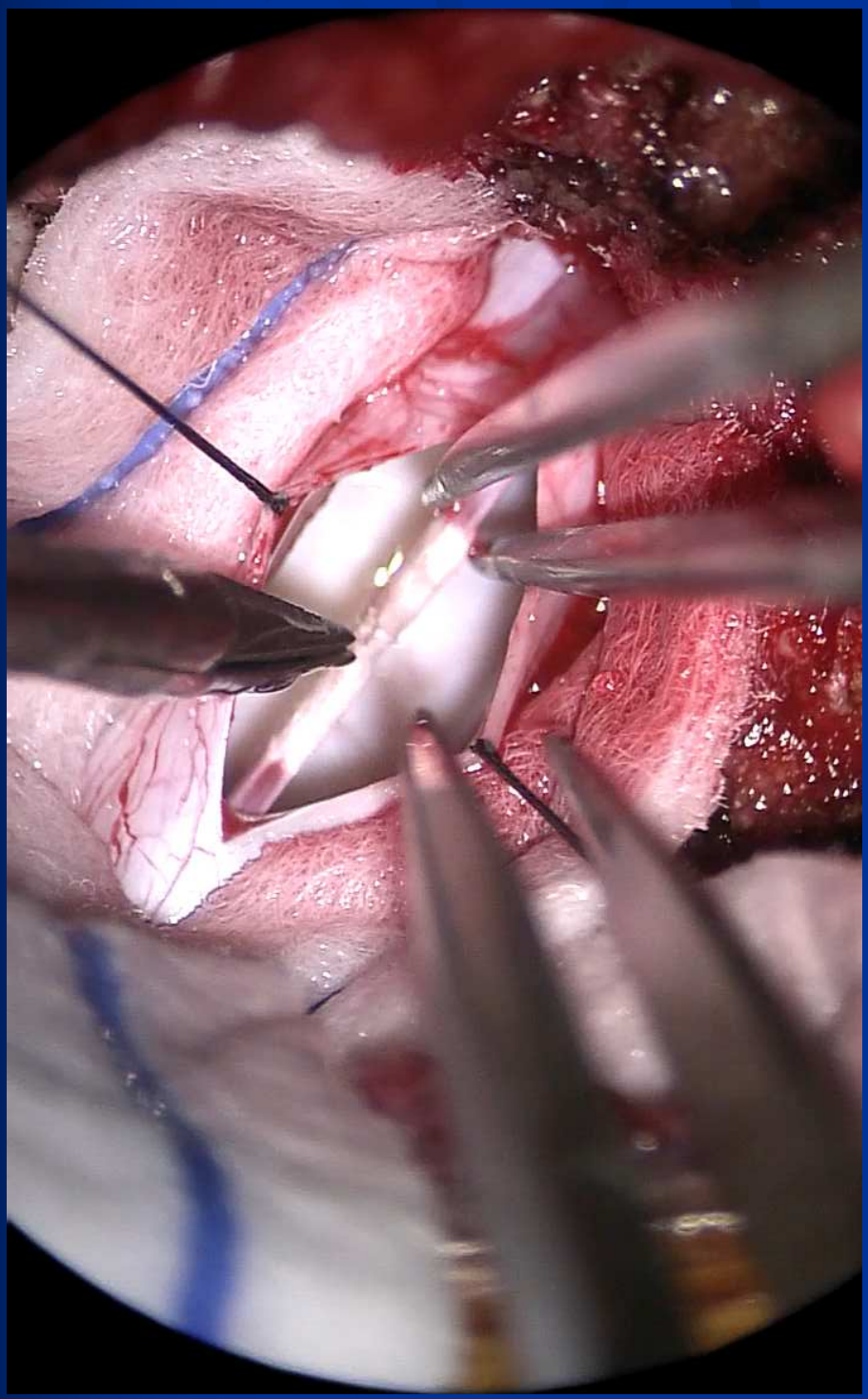
Patient C.L.: Tethered Cord

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



Discussion

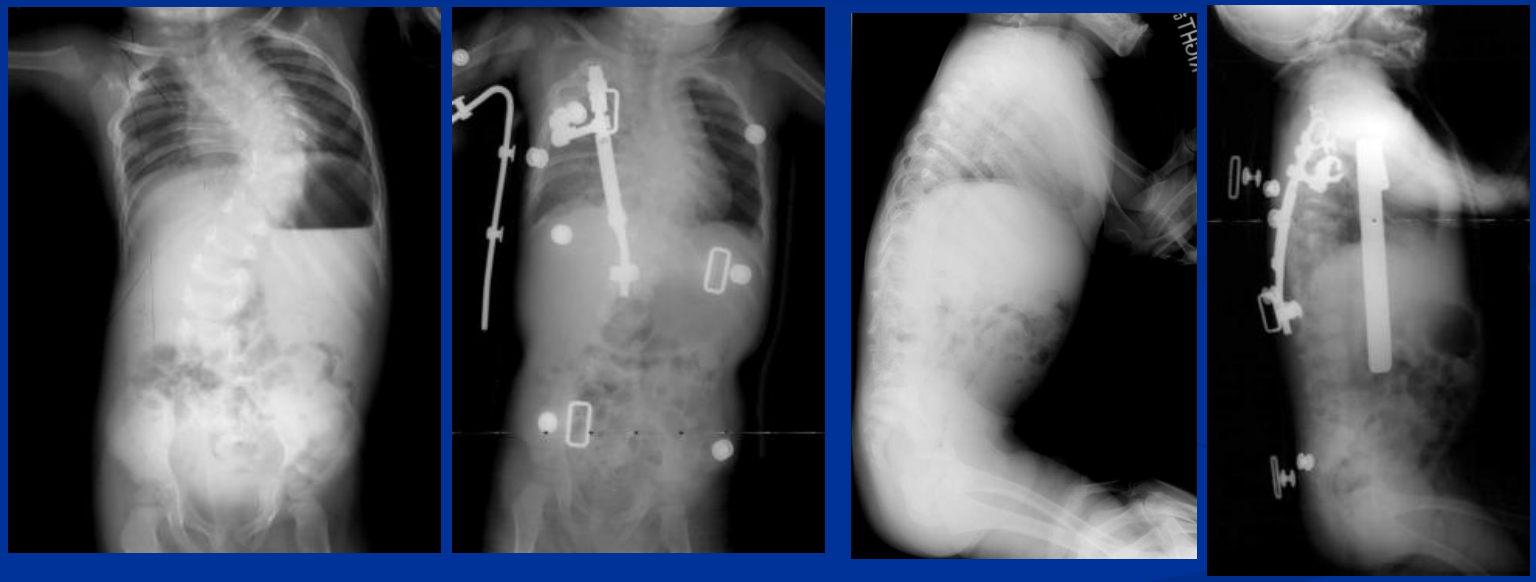
- When to get MRI?
- Indications for fatty filum release?
- Should it be done concurrently?
- What if tethered secondary to myelomeningocele?
- Brief overview of technique
  - Surgical video



Fatty Filum with Low Lying Conus

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

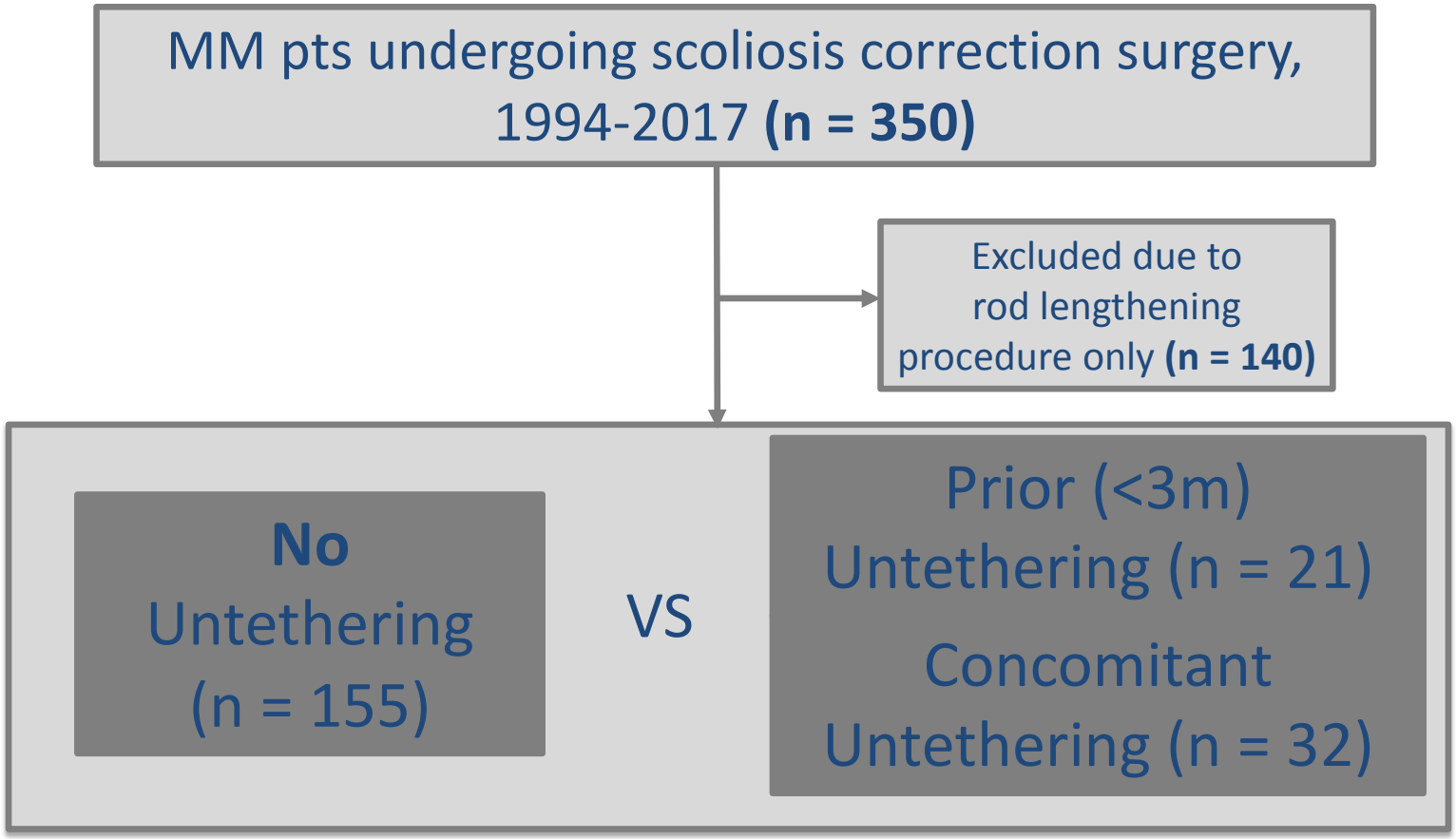
Uneventful Surgery



Case: 14 yo MM with rigid 90 degree curve and back pain

Shunt?

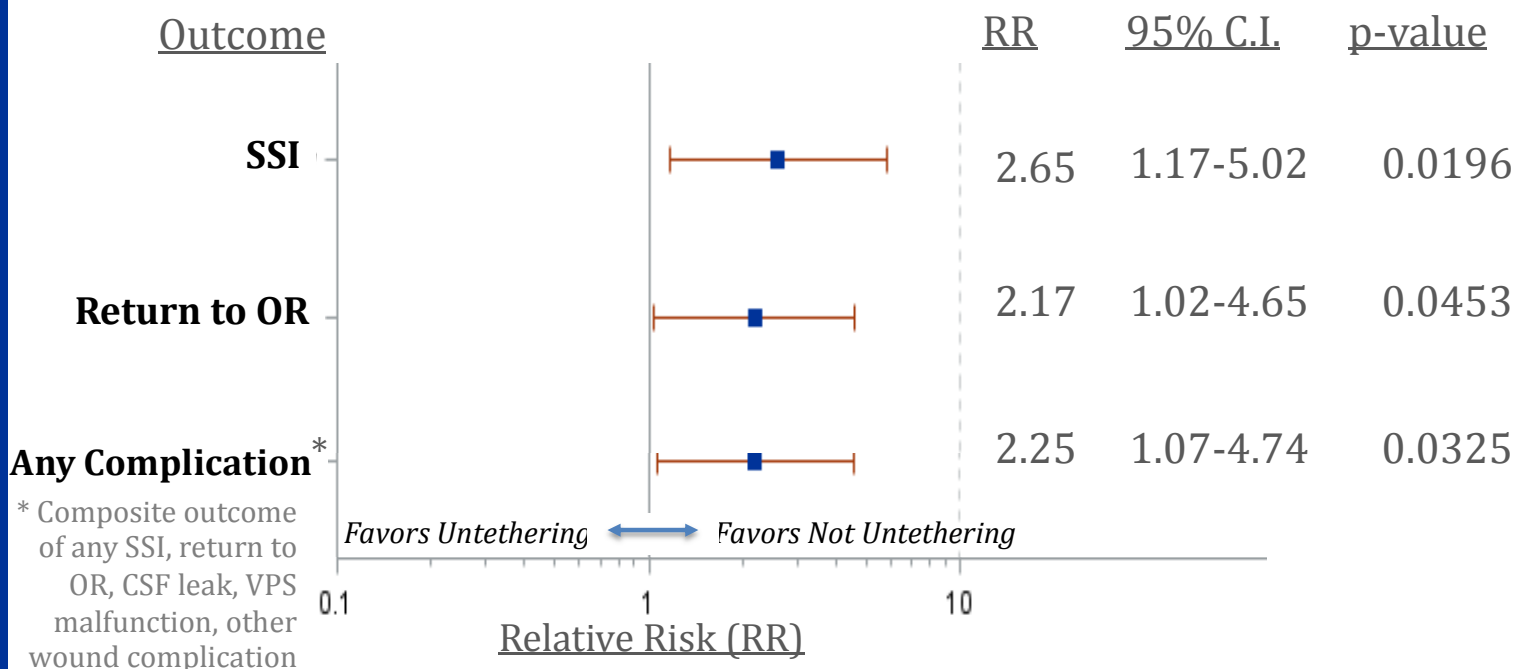
Results: Baseline



Results: Multivariate Analysis

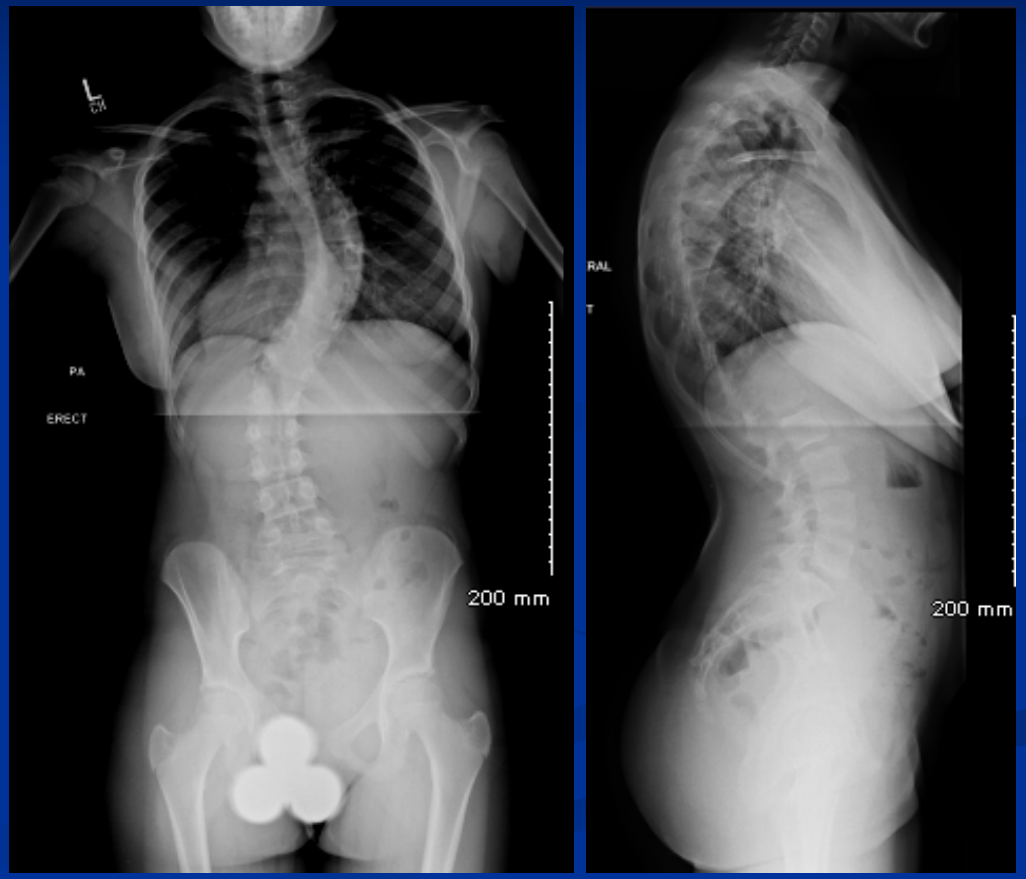
Relative Risk of **Post-Operative Complications** Associated with Prophylactic Untethering

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

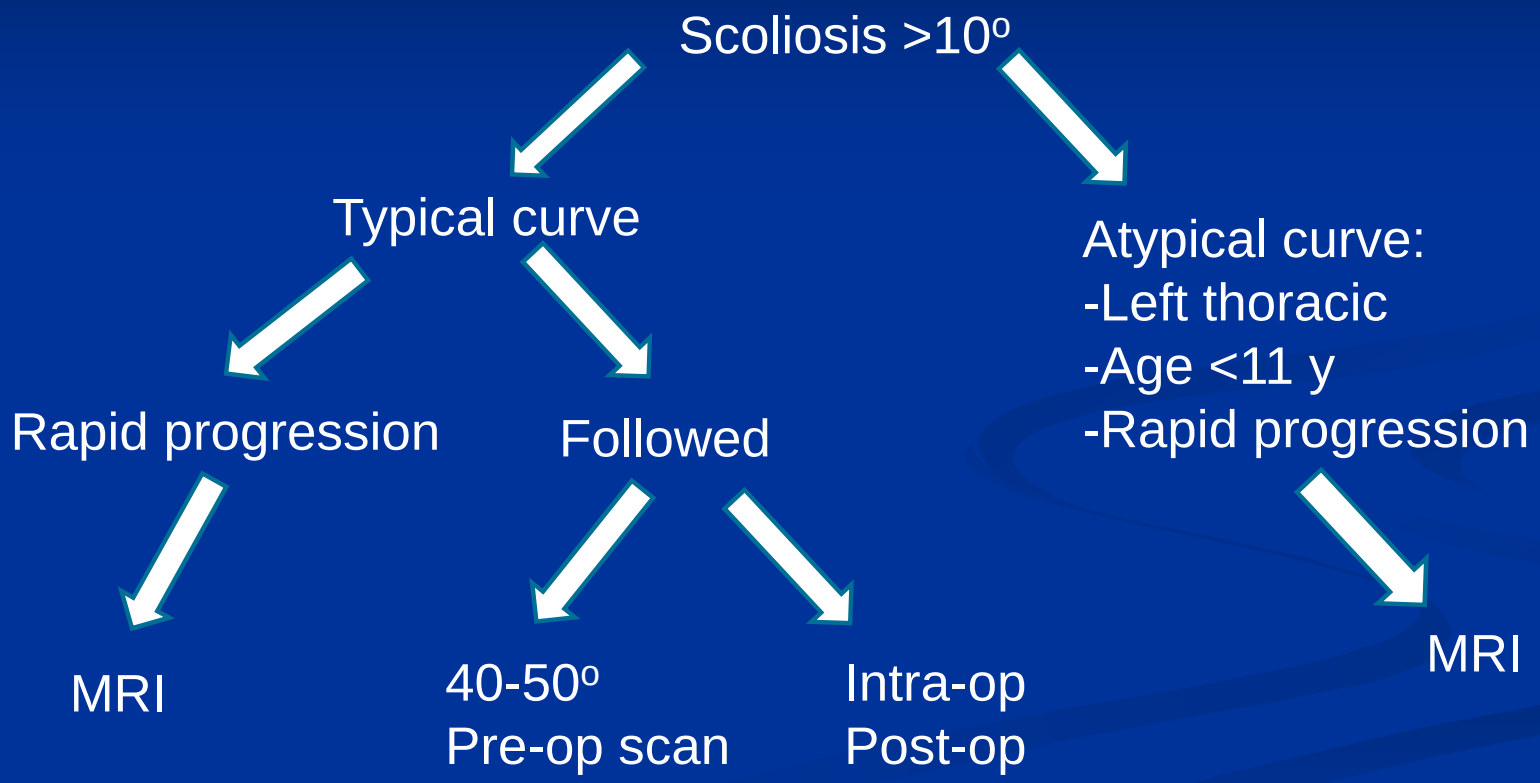


Patient M.M.: Chiari Malformation

- 11-year-old girl presented with progressive scoliosis
  - Main thoracic curve 35 to 60º
  - Kyphosis T5-12: 75º
- Neurologically intact



Scoliosis: When to Get an MRI?



Imaging

Questions

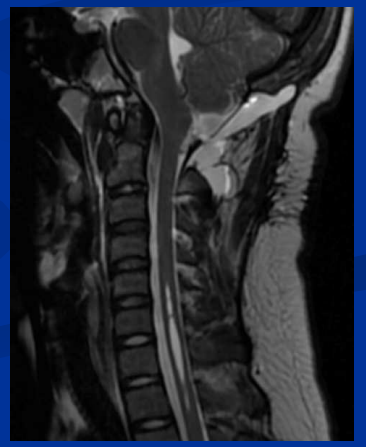
- Does every Chiari need to be decompressed prior to curve correction?
- How long should we wait after Chiari decompression for curve correction?
- What if syrinx does not get better?
- What are the indications for another procedure, like a syrinx shunt?
- What are the risks of scoli surgery if the syrinx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

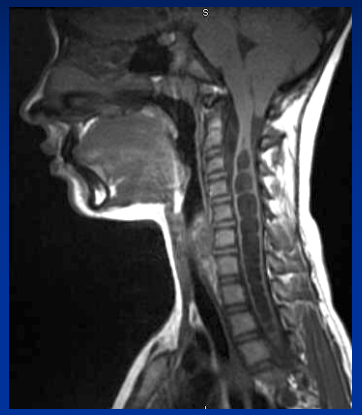
This Patient's Course

- 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

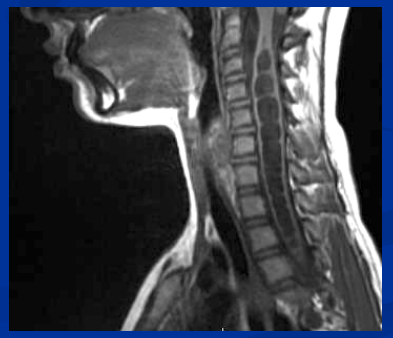


Six Months Later

- Repeat MRI and x-rays
  - No change in syrinx or scoli
- What now?
- Options:
  - Wait
  - Repeat PFD (with or without 4th 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” ?

Spine (Phila Pa 1976). 2006 Sep 1;31(19):E698-706.

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

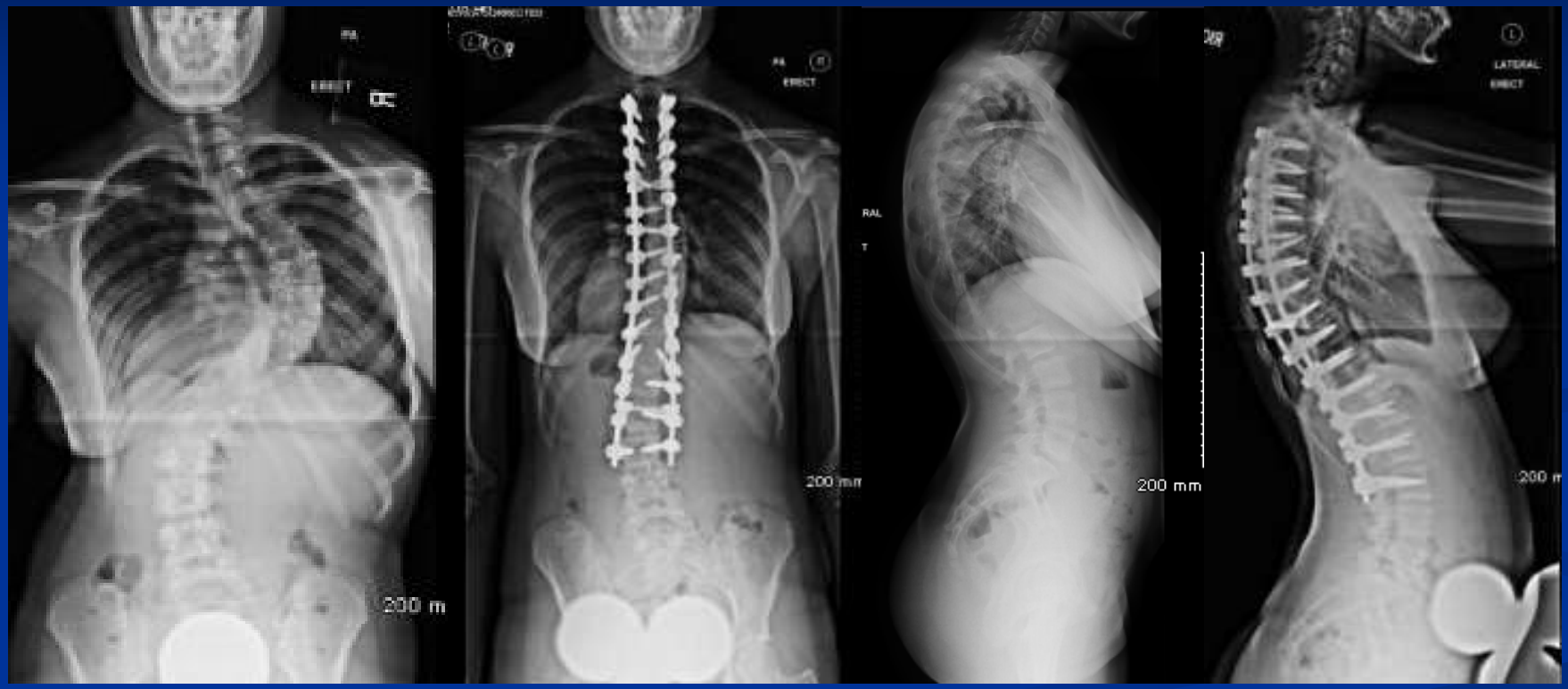
El-Hawary R[1], Sucato DJ, Sparagana S, McClung A, Van Allen E, Rampy P.

Spine Deform. 2013 May;1(3):205-210. doi: 10.1016/j.jspd.2013.02.002. Epub 2013 Jun 6.

Spinal Cord Monitoring With Transcranial Motor Evoked Potentials in Patients With Neural Axis Abnormalities Undergoing Spinal Deformity Surgery.

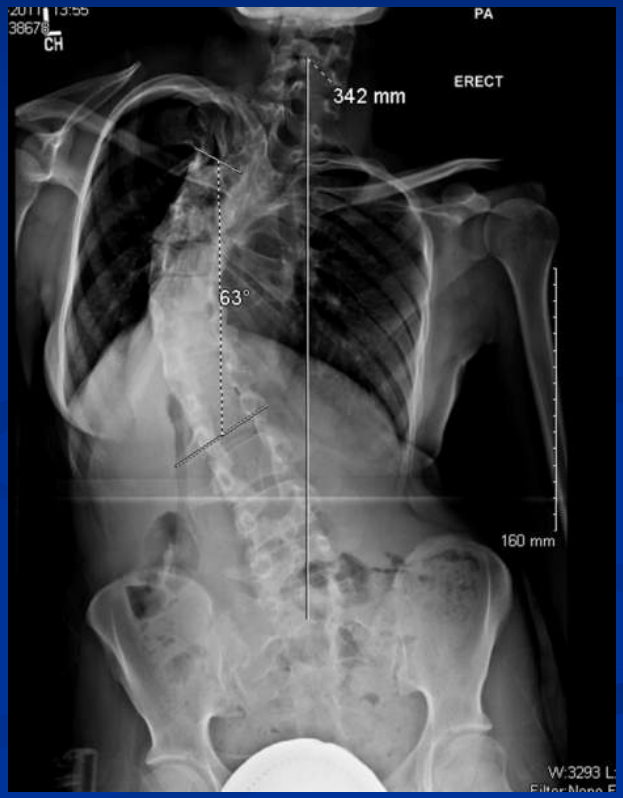
Muchow RD[1], McClung A[2], Rampy P[2], Van Allen E[2], Sparagana S[2], Sucato DJ[2].

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

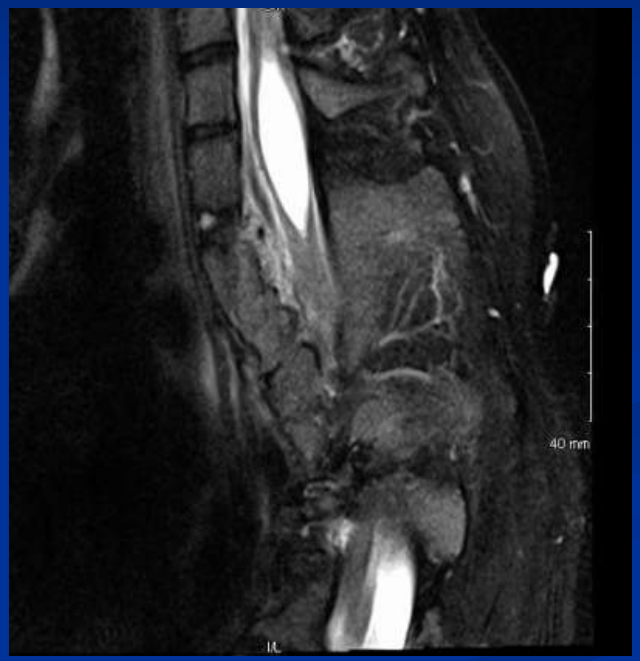


Clinical Photos

October 2011

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

Shen J et al, JBJS-A 2016;98:926-36

- 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

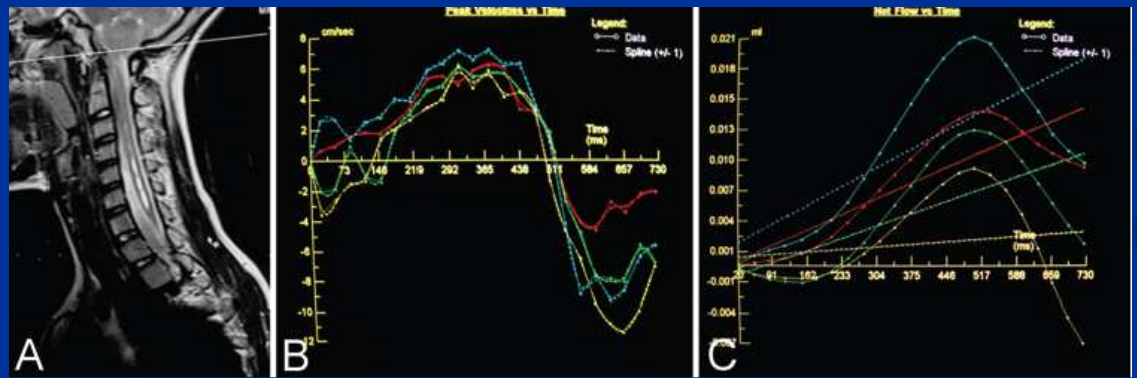
SRS 2013: Large Syrinx With Chiari

- 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

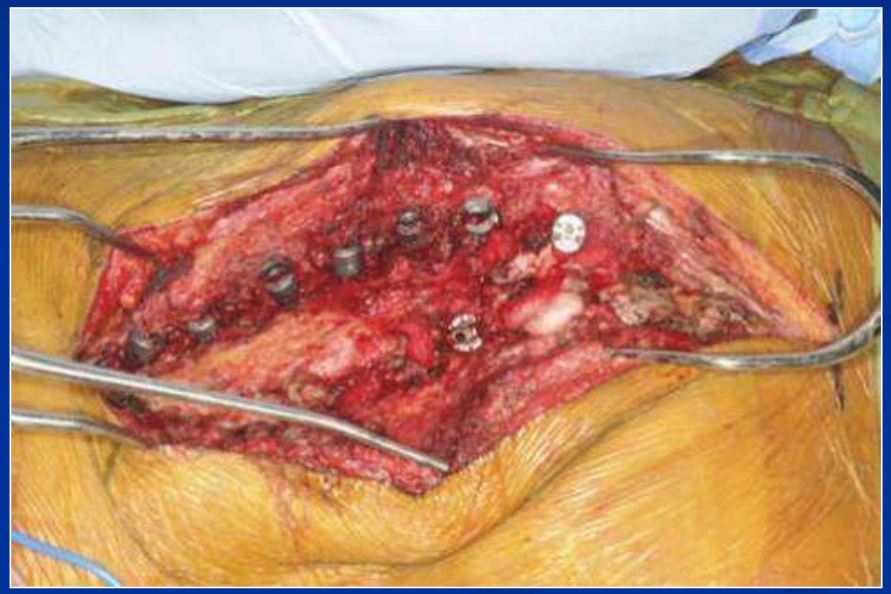
Wang Y *et al*, JNS Spine 18:456, 2013

- 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

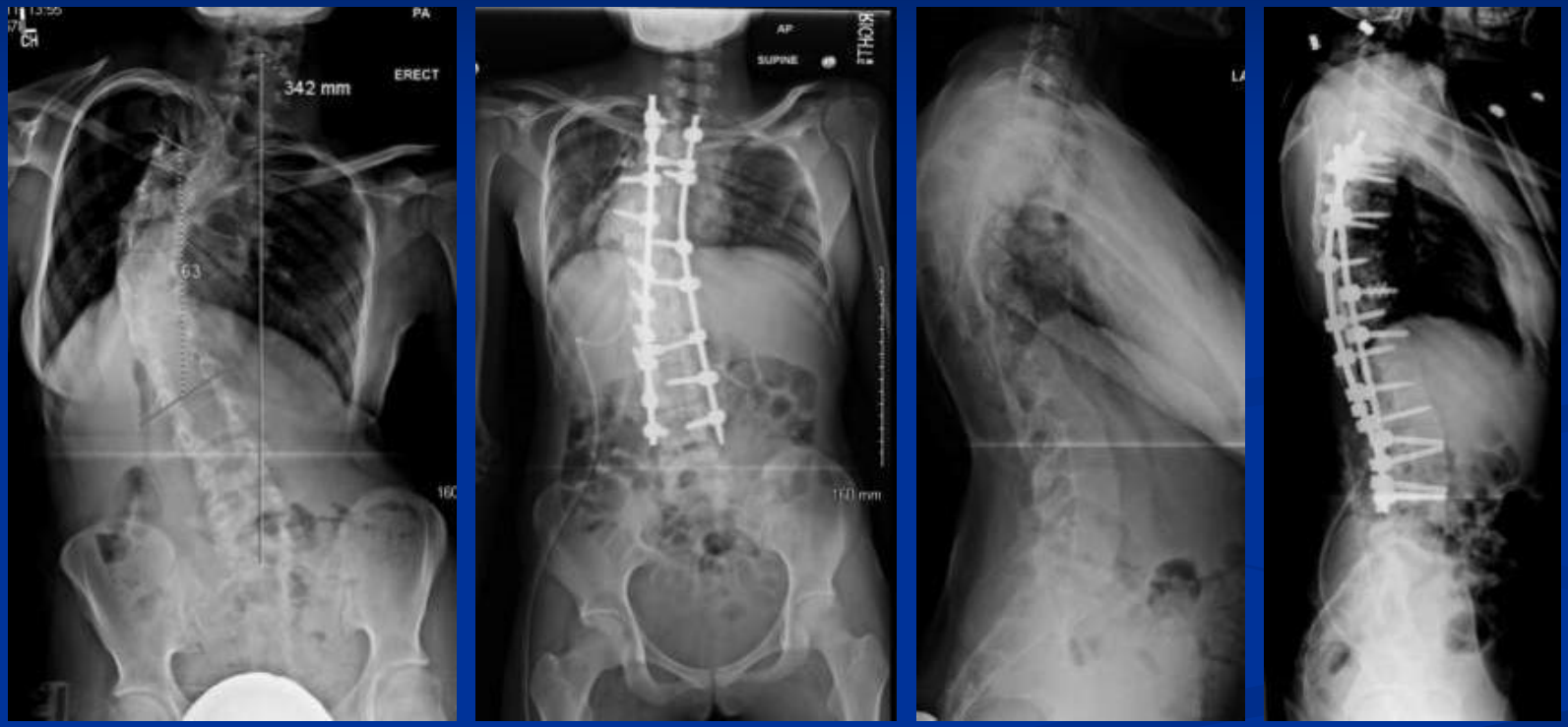


Intraoperative

- 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 < 35º
- Not all patients with split cord malformation will need neurosurgical intervention prior to correction of their scoliosis