Syrinx, Chiari, Tether – Need for Treatment

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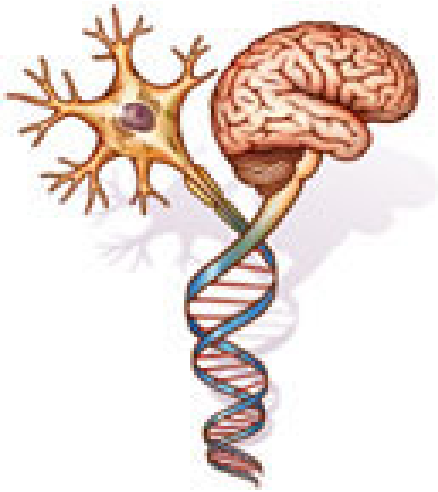
ICEOS Toronto November 2010

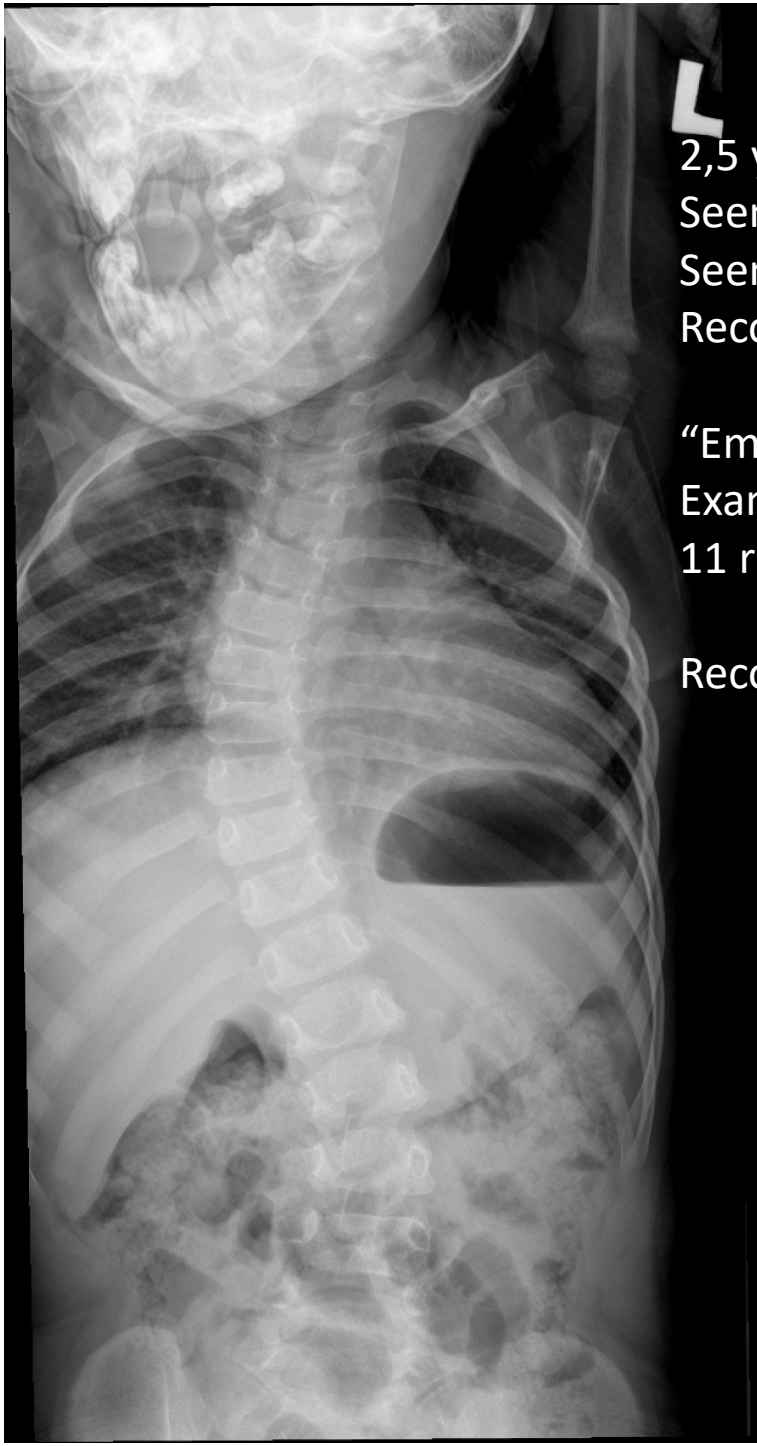
The Hospital for Sick Children
University of Toronto, Canada

SickKids



Objectives

- Learn about the nervous system embryology, and common anomalies associated with early onset scoliosis, their diagnosis and management.
- Disclosures - none

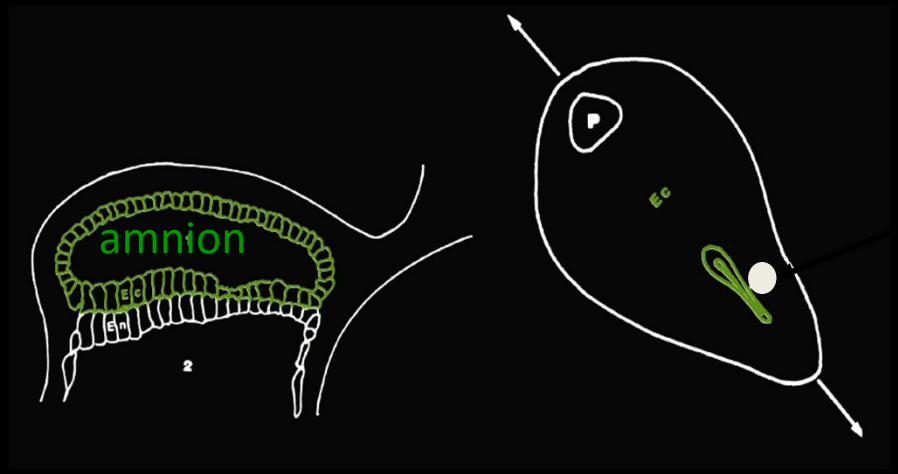


2,5 yr old female infantile scoliosis
Seen in orthopedic clinic HSC July . MRI 3-6 months ordered
Seen at Shriner's USA 2 weeks, MRI conus L2-L3
Recommended de-tethering operation by Shriner neurosurgeon

"Emergency" second opinion neurosurgery HSC
Exam normal (? Absent lt ankle jerk)
11 ribs, lumbarized S1, filum normal on MRI

Recommended observation

embryology

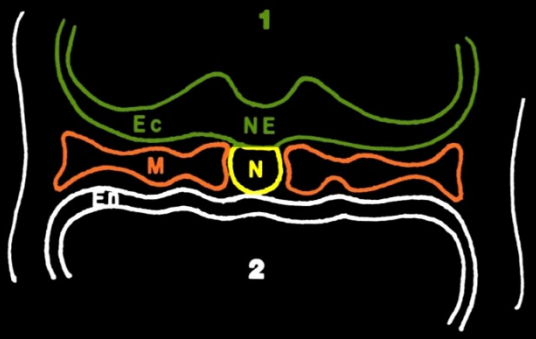


Primitive streak forms
in the midline of bilaminar disc

midline cells form notochord

mesenchyme

embryology: neurulation



neural plate thickens

neural folds

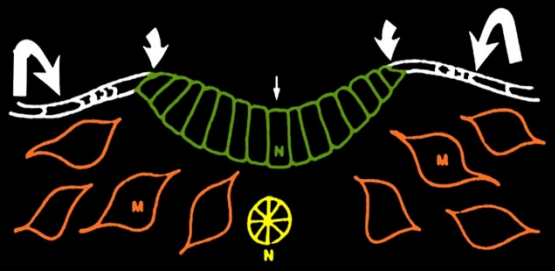
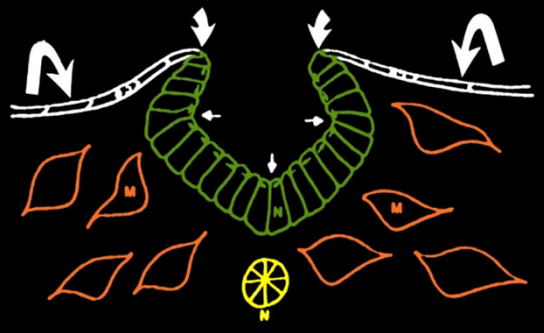


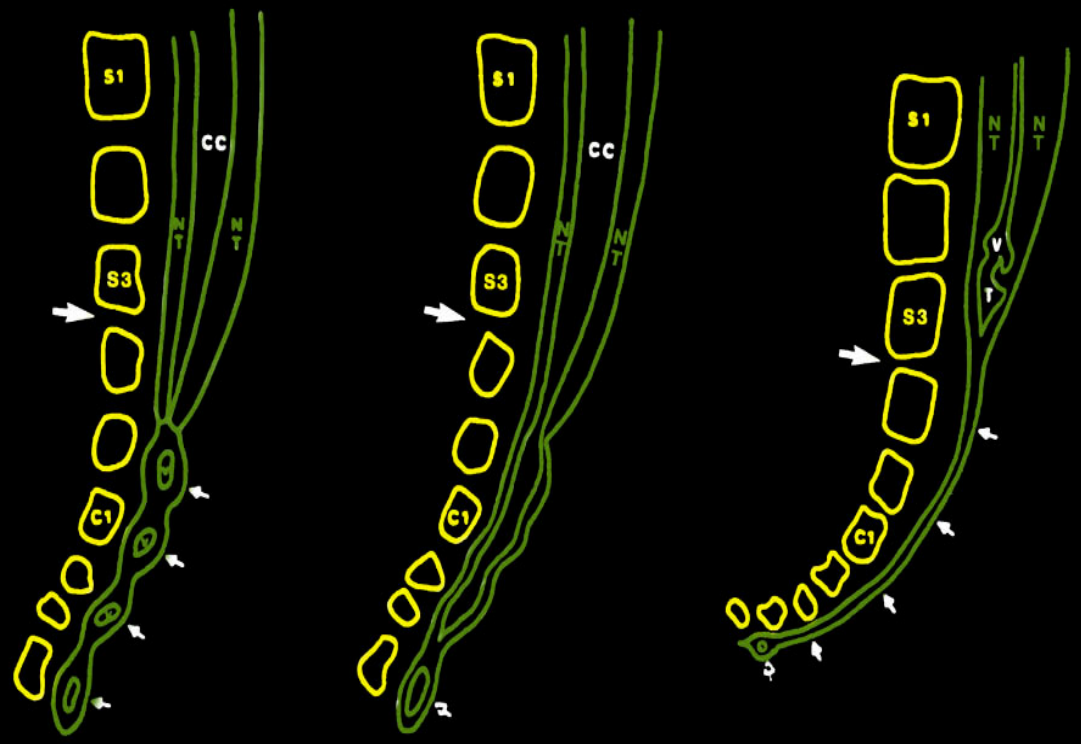
plate flexes



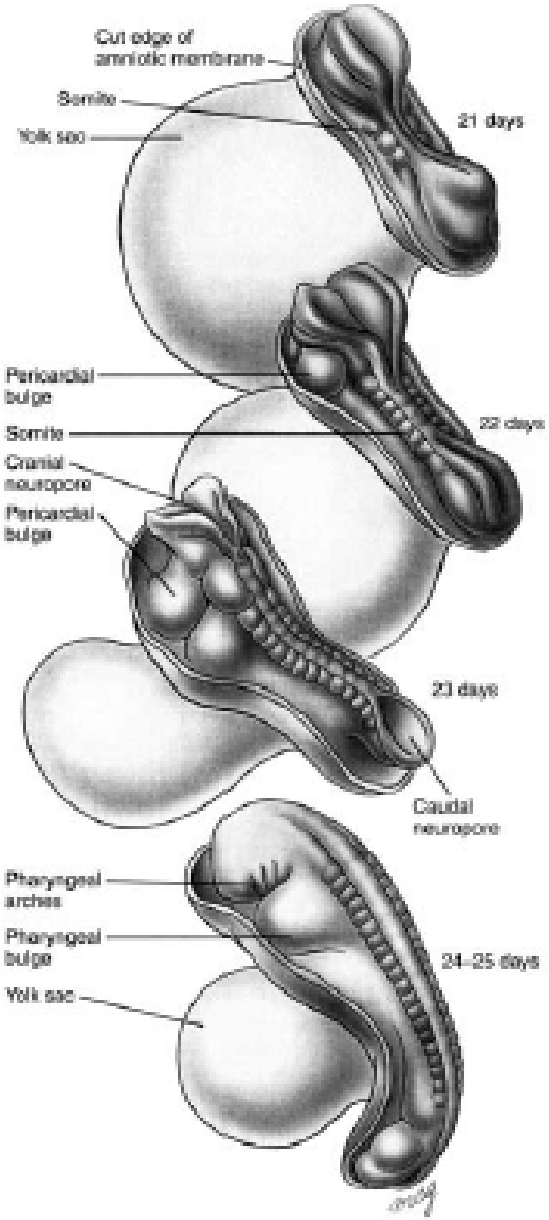
midline sinks into
neural groove

canalization / retrogressive differentiation

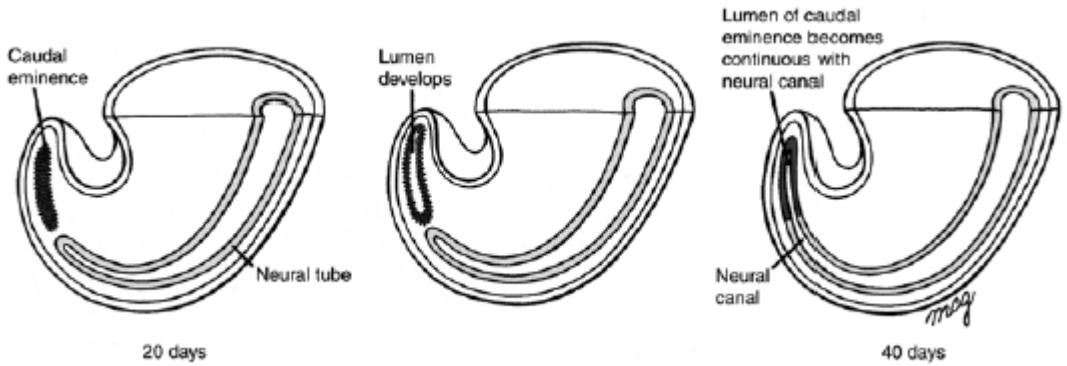
- cell agglomeration, vacuolation and involution



Primary Neurulation



Secondary Neurulation

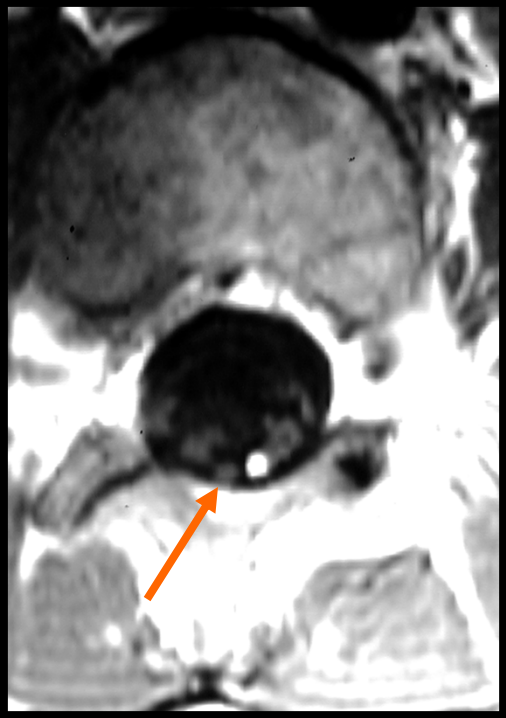


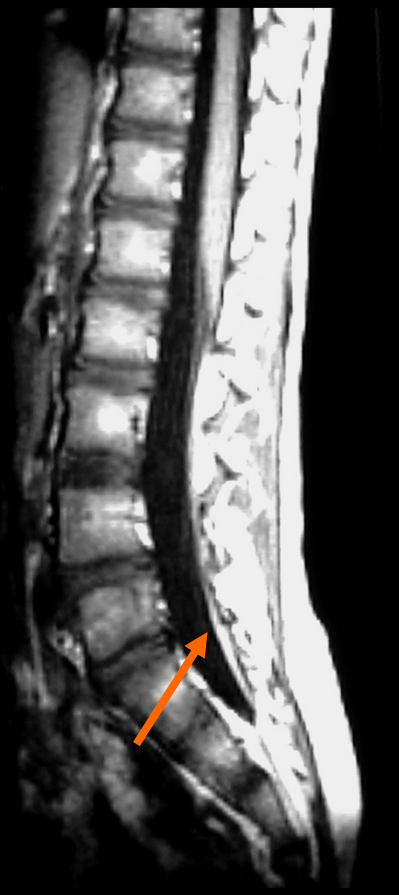
Between 13 and 18 weeks' gestation, the Conus Medullaris was situated at the level of the L4 vertebra, or more caudally, in 100% of the fetuses. At term, all fetuses showed the CM above L2. A distinct ascent of the CM was detected between 13 and 40 weeks' gestation.

Zalel Y, Lehavi O, Aizenstein O, Achiron R. Development of the fetal spinal cord: time of ascendance of the normal conus medullaris as detected by sonography. *J Ultrasound Med. 2006 Nov;25(11):1397-401; quiz 1402-3.*

filum lipoma: failure of involution of distal band

- distal lipomas are caused by failure of canalization and retrogressive differentiation





fetal MR: lipomyelomeningocele

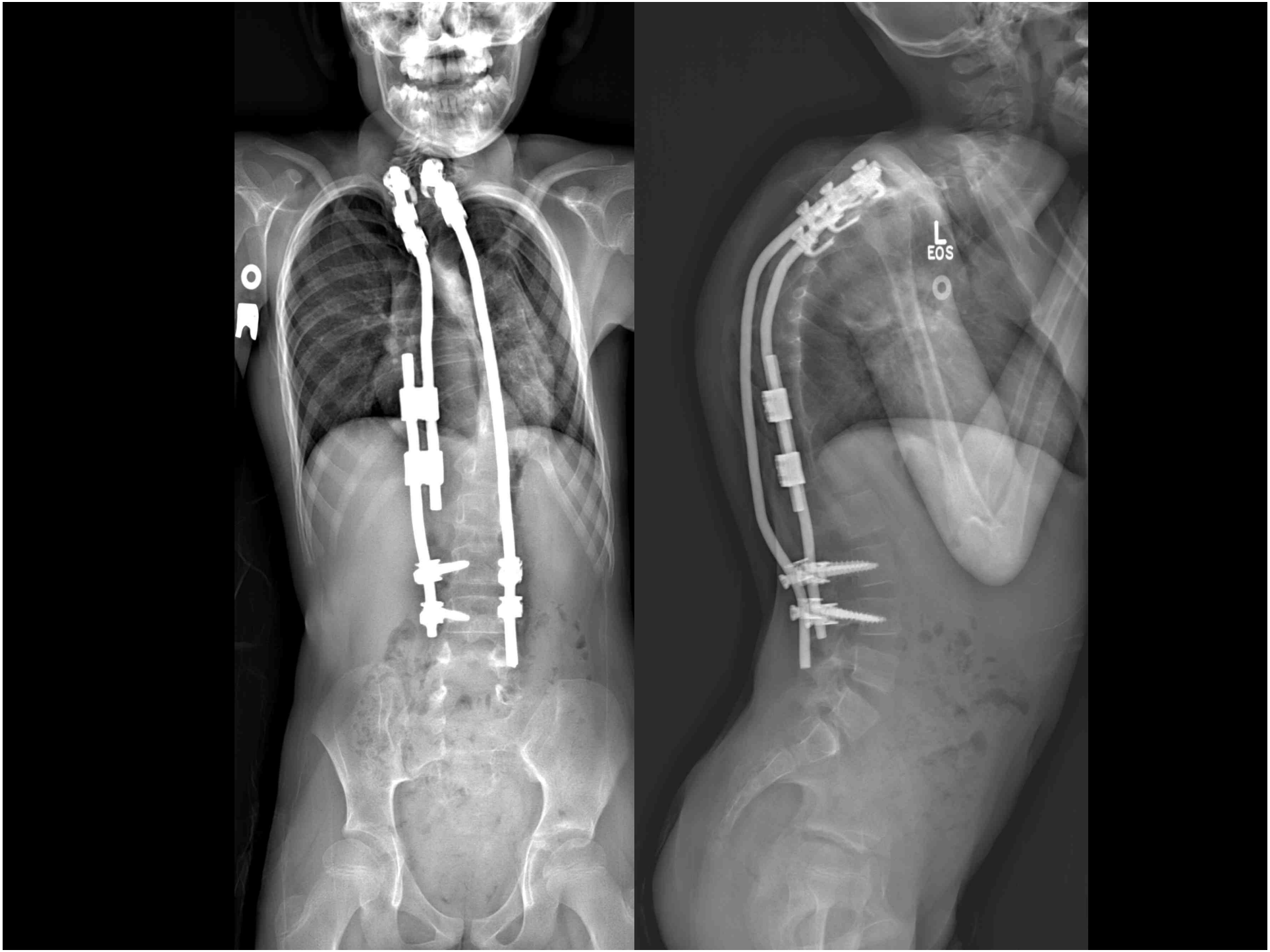
Tethered Spinal Cord

- A concept – no definitive test
- Low lying spinal cord under tension
- Causes low level spinal cord ischemia & dysfunction (rarely if ever measured)
- Leg muscle atrophy, weakness, foot deformity, pain, bladder dysfunction

Tethered Spinal Cord

10 yr old boy, hairy patch on back
Neurologically intact. Still same 5 yrs later
Plays soccer.


R
(9 yr old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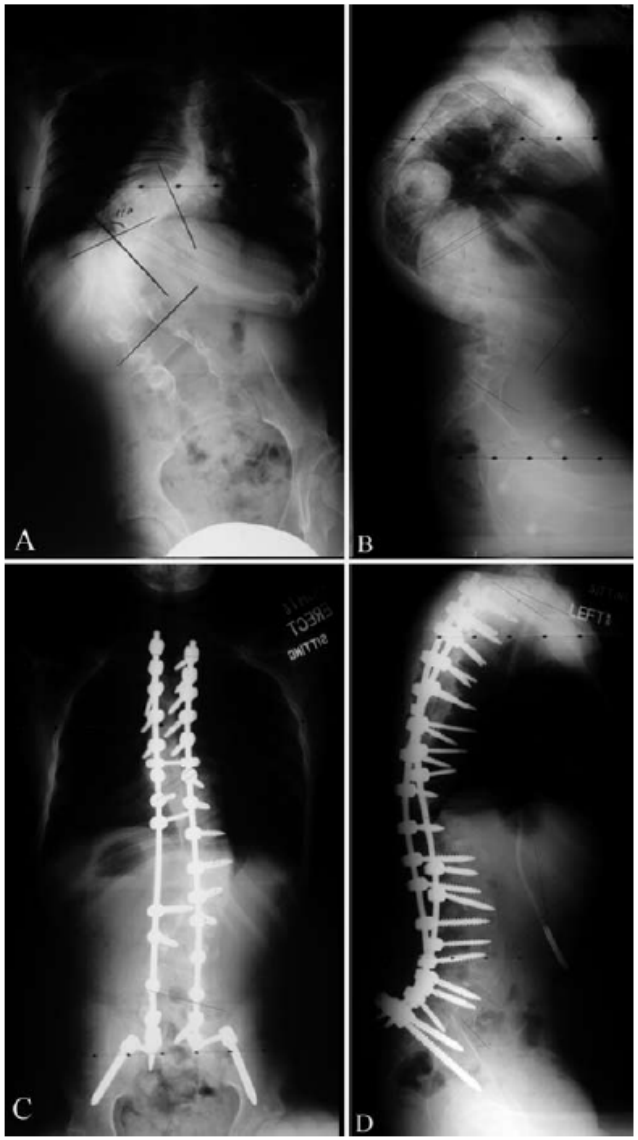
Neurosurg Focus 29 (1):E8, 2010

A patient with myelomeningocele: is untethering necessary prior to scoliosis correction?

Amer F. Samdani, M.D., Anthony L. Fine, B.S., Sukhdeep S. Sagoo, D.O., Shailja C. Shah, B.S., Patrick J. Cahill, M.D., David H. Clements, M.D., and Randal R. Betz, M.D.

Shriners Hospital for Children, Philadelphia, Pennsylvania



Conclusions. The study results suggested that spinal cord untethering may be unnecessary in patients with MM who are undergoing scoliosis corrective surgery and do not present with clinical symptoms of a tethered cord, even though tethering is radiographically demonstrated.

J Neurosurg (Pediatrics 1) 103:24–35, 2005

Neurosurgical causes of scoliosis in patients with myelomeningocele: an evidence-based literature review

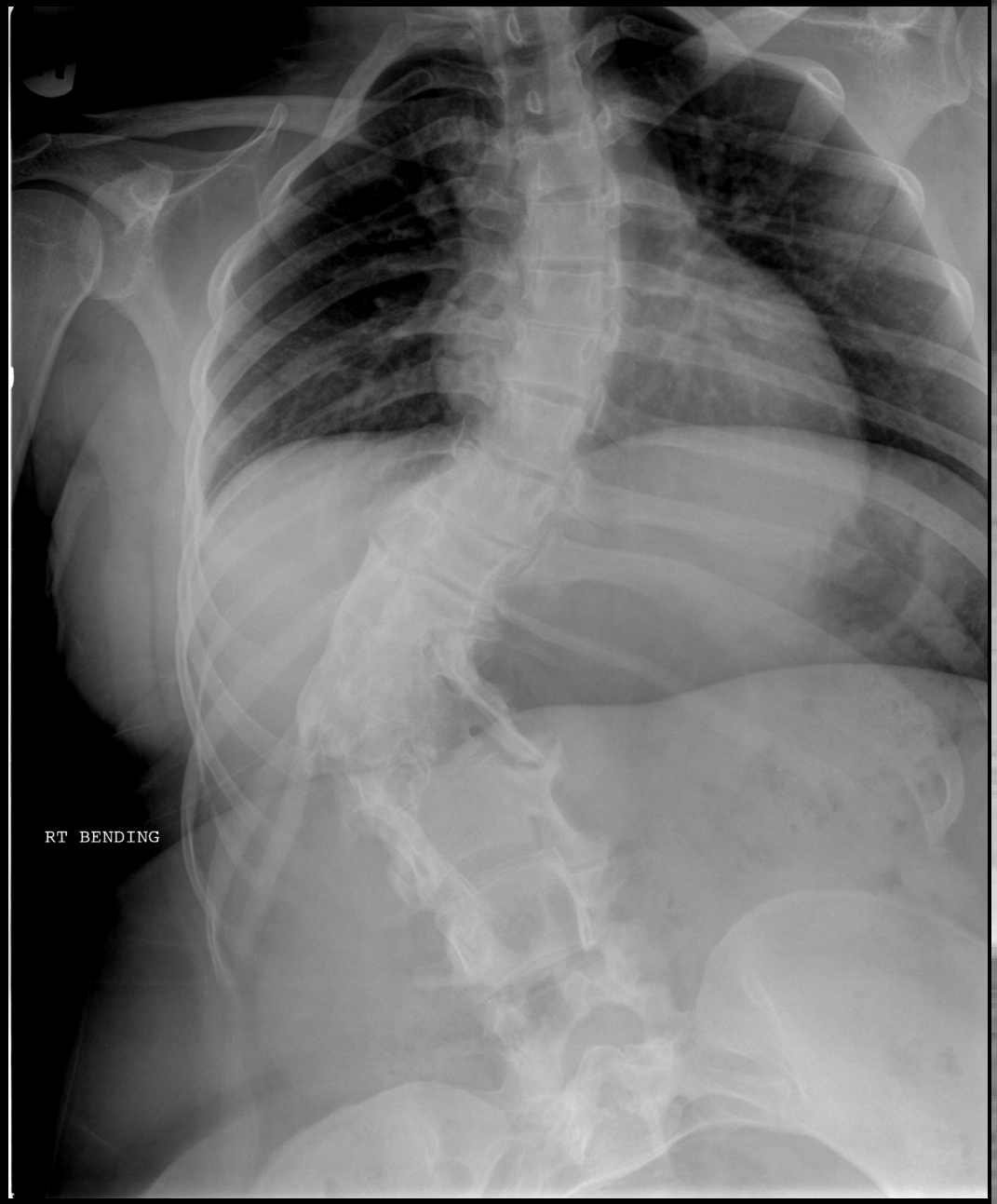
MARK S. DIAS, M.D.

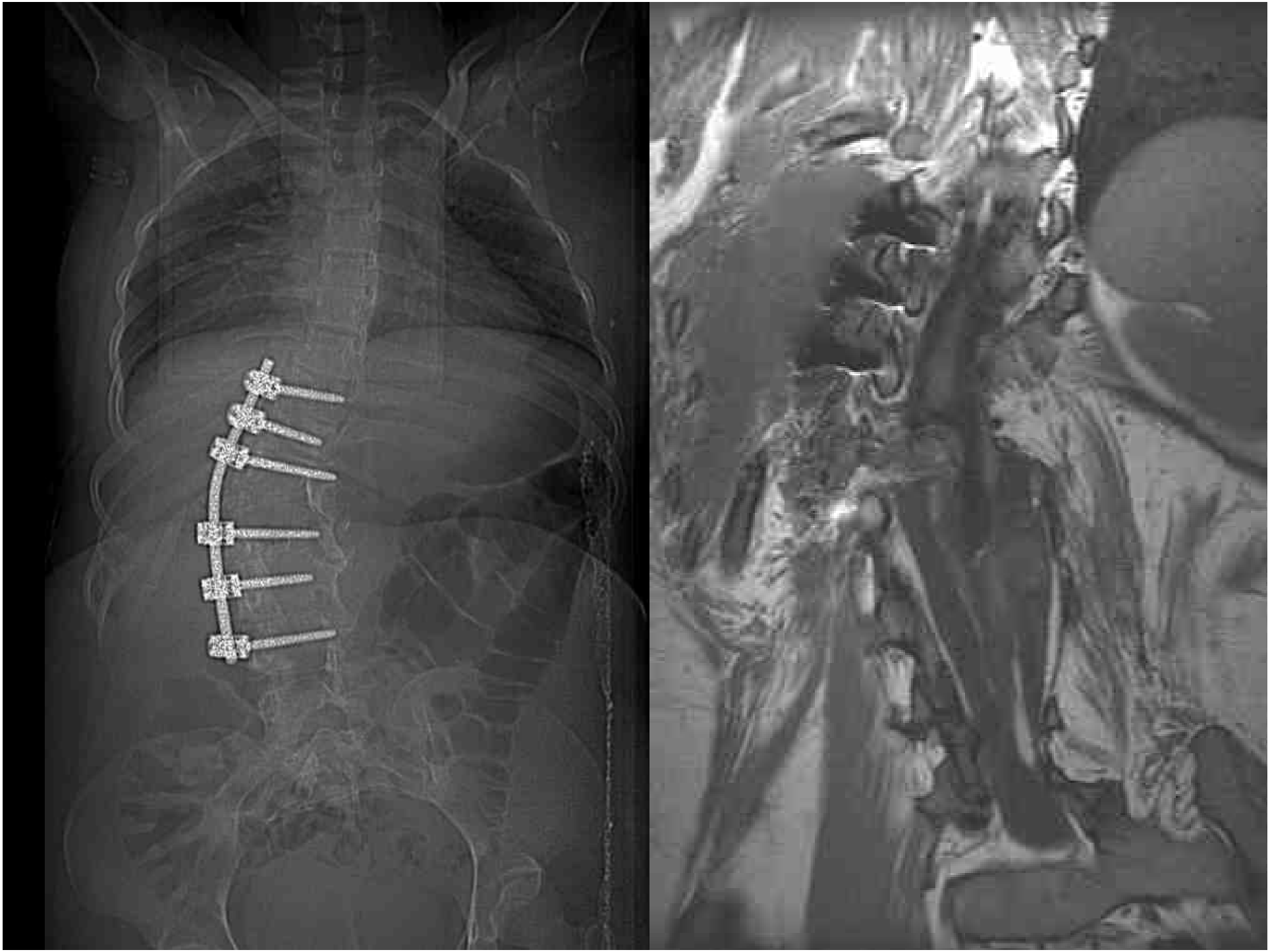
Department of Neurosurgery, Penn State Milton S. Hershey Medical Center, Hershey, Pennsylvania

Conclusions. All available studies were classified as Level 4 studies (case series and flawed cohort and case-control studies). Based on the strength of the available data, there may be an association between spinal cord tethering and scoliosis, particularly in those patients with upper lumbar lesions and spinal curves less than 45°. There is little evidence to support a causal relationship between scoliosis and Chiari malformation or syringomyelia in this population.

MYELOCYSTOCELE

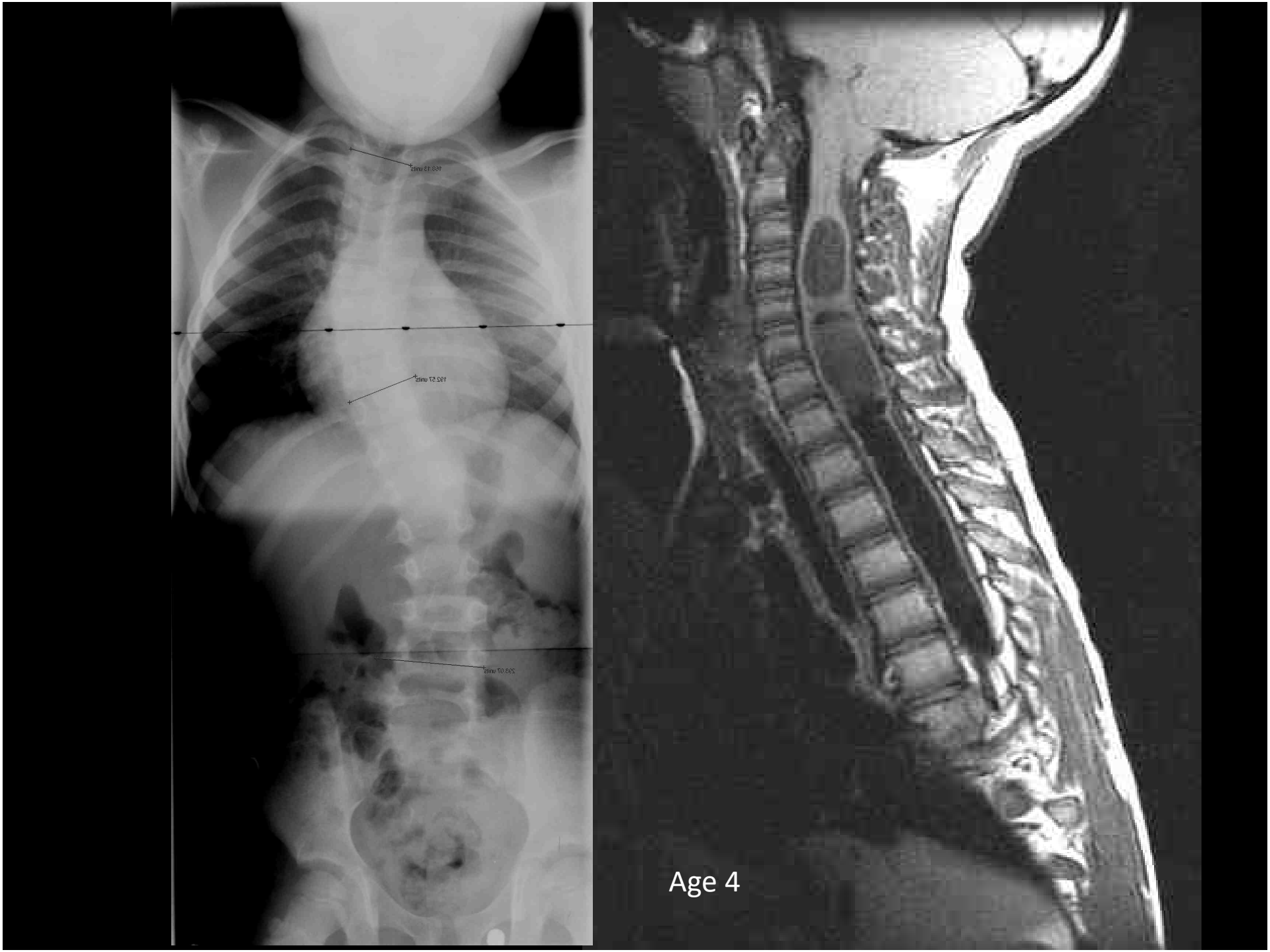
1 Yr of age, rapidly progressive scoliosis
Repeat surgery for un-tethering


RT BENDING

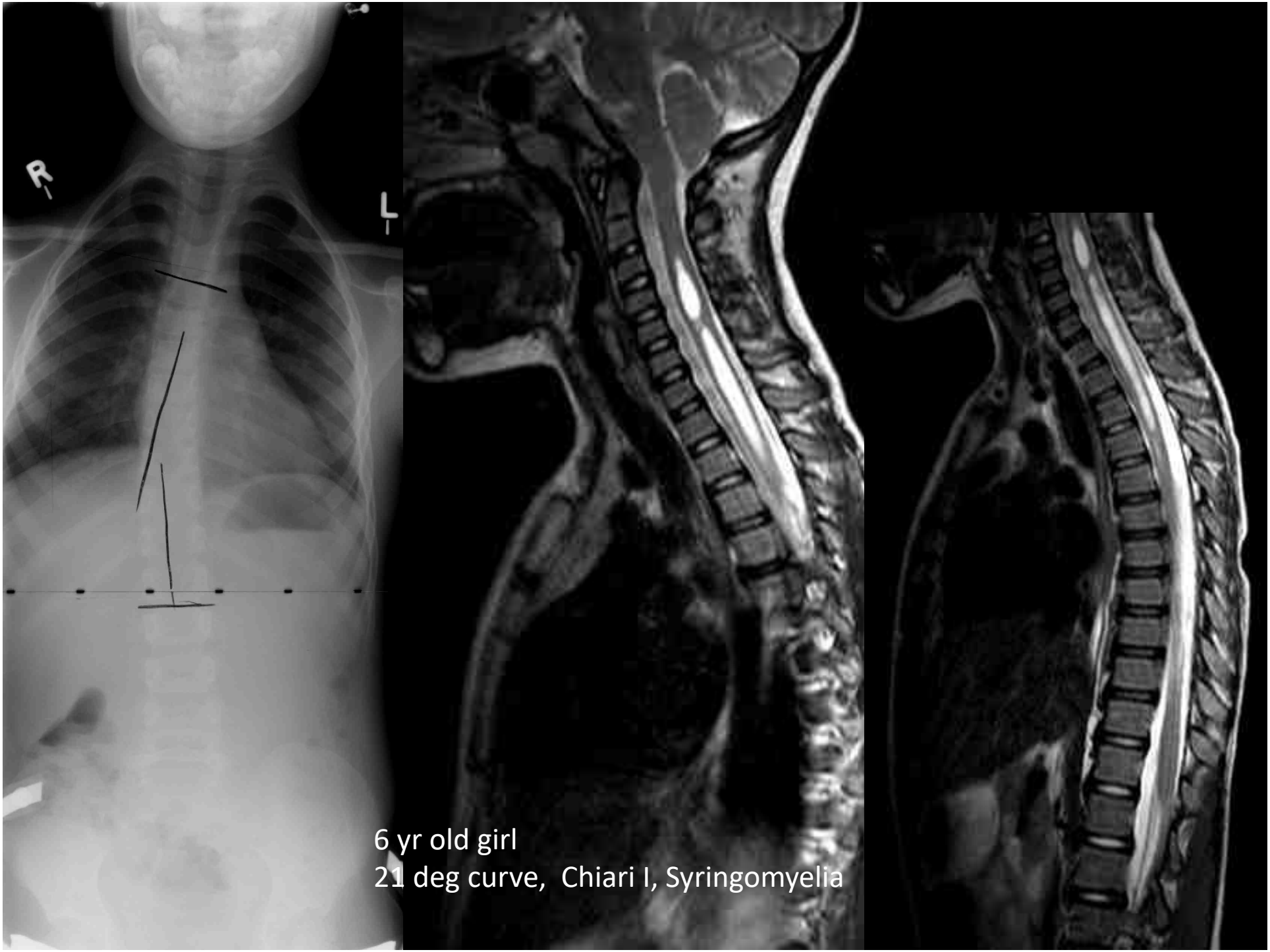


Tethered Cord and Scoliosis

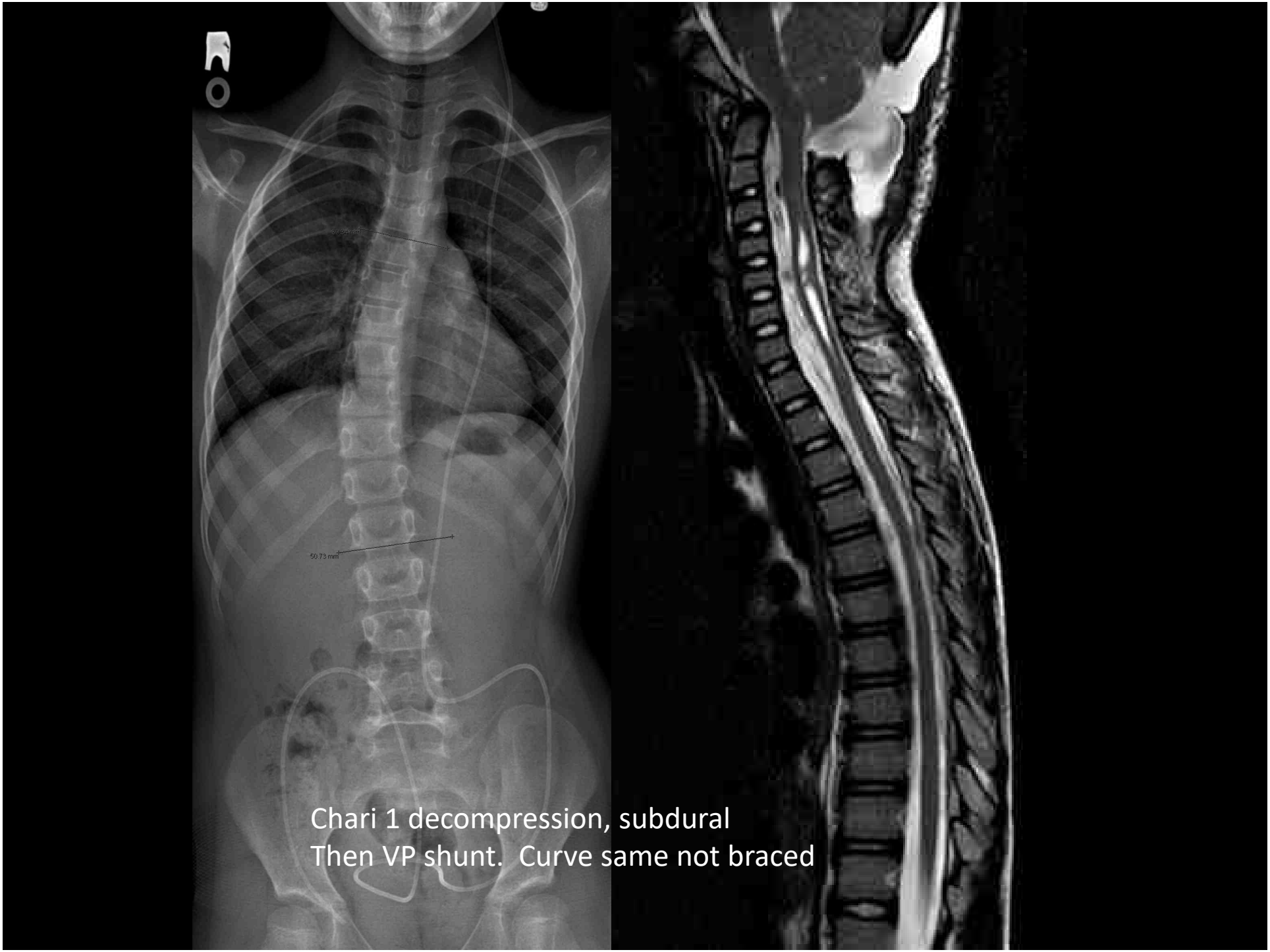
- Diverse spectrums of abnormalities
- Causal relationships tenuous and risks of tethering/de-tethering not well understood
- Case by case decision making taking into account
  - Degree of congential abnormality
  - Degree of pre-existing neurlogical impairment
  - Magnitude of correction


Age 4


Age 12



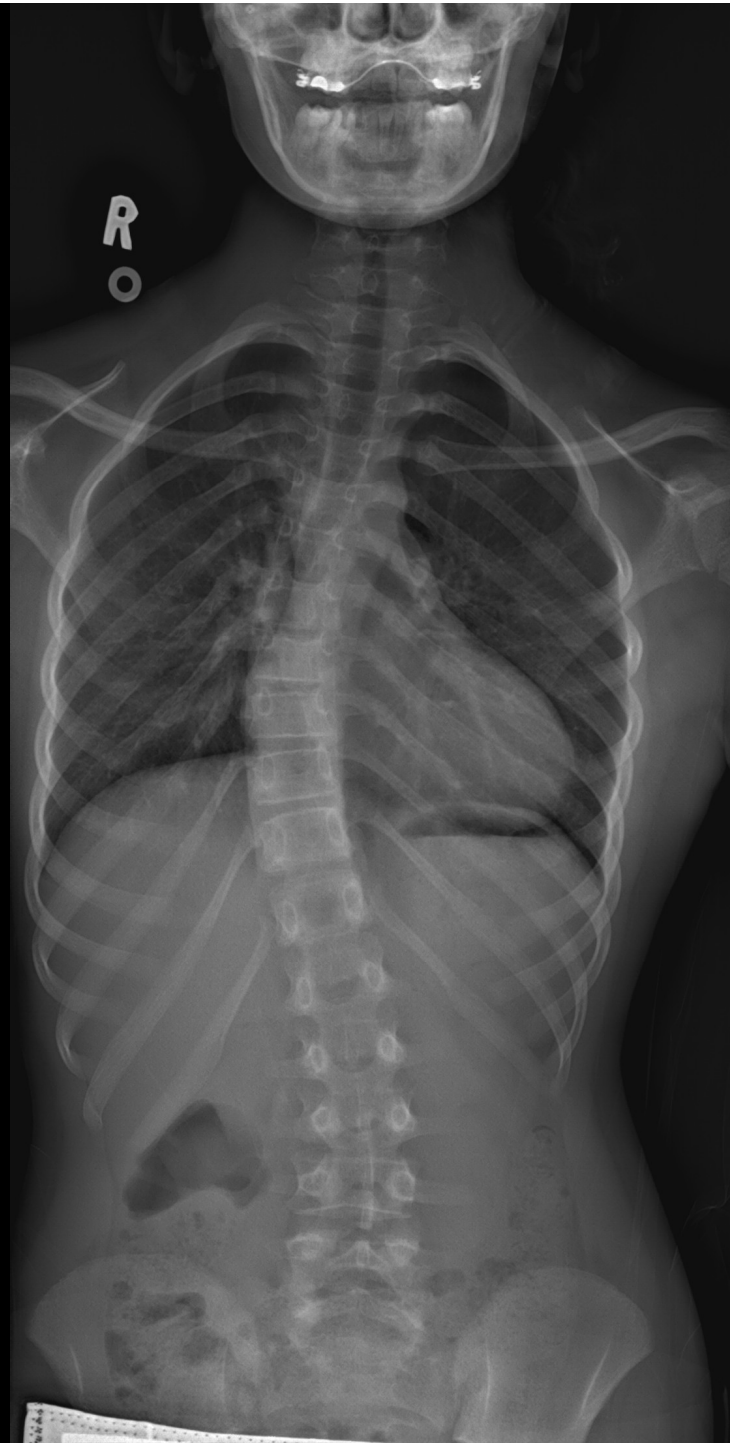
6 yr old girl
21 deg curve, Chiari I, Syringomyelia

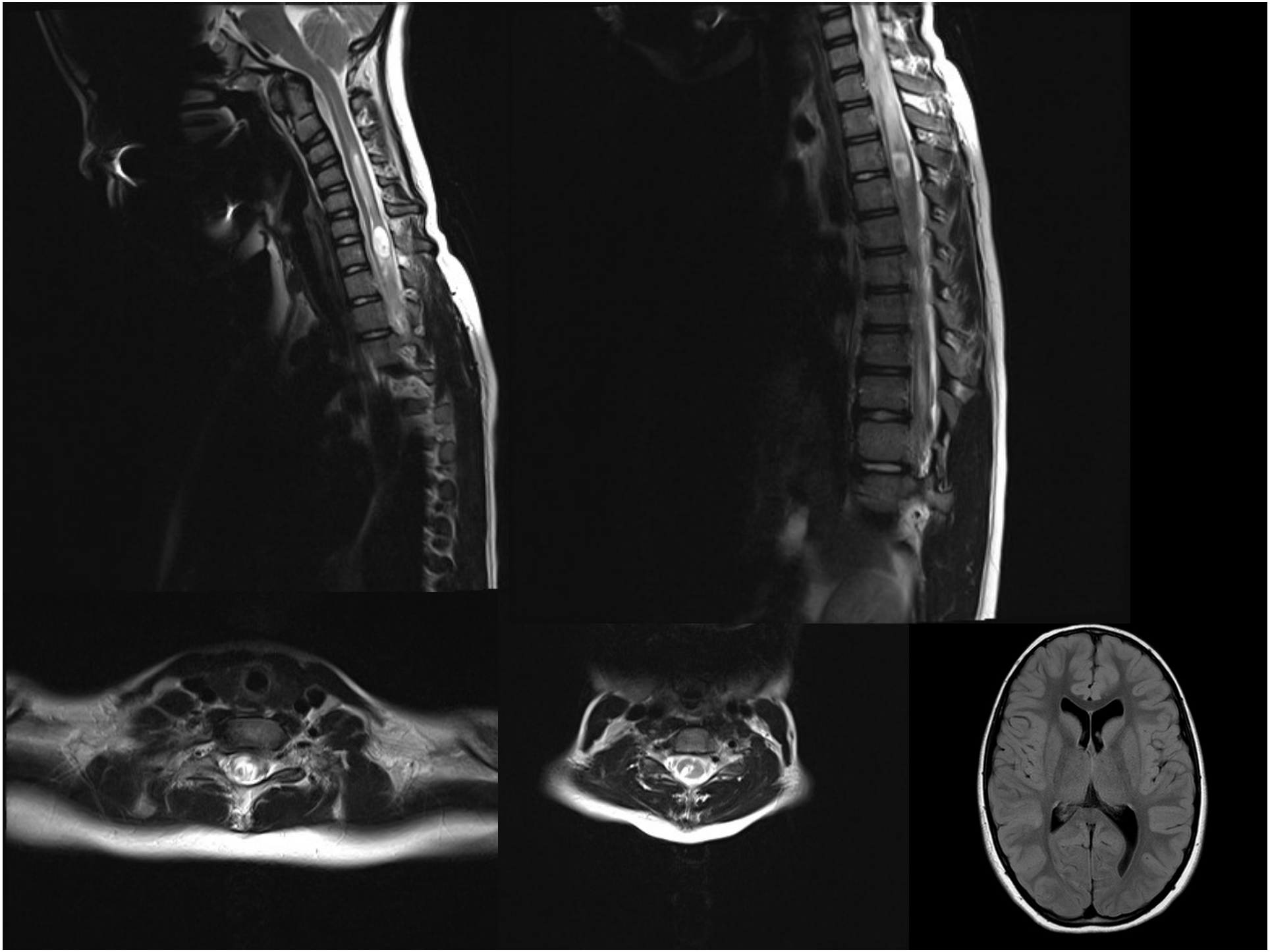


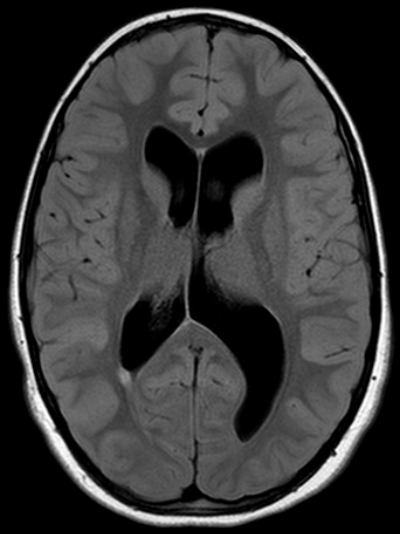
Chari 1 decompression, subdural
Then VP shunt. Curve same not braced

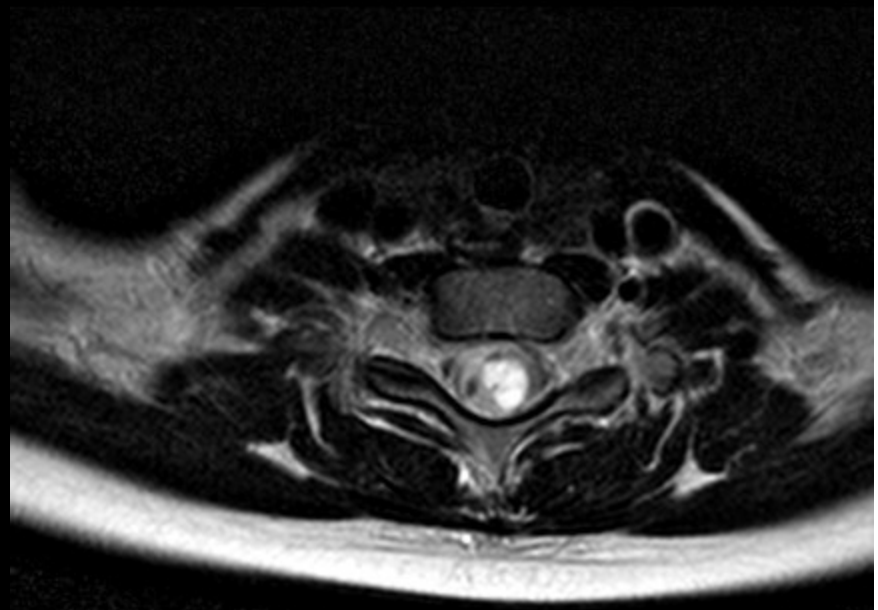

4 yr old boy, foot drop, loss of pain/temp
Absent abdominal reflexes

13 yr old female, arthrogryposis, scoliosis. Previous history of palatal insufficiency, mechanical.

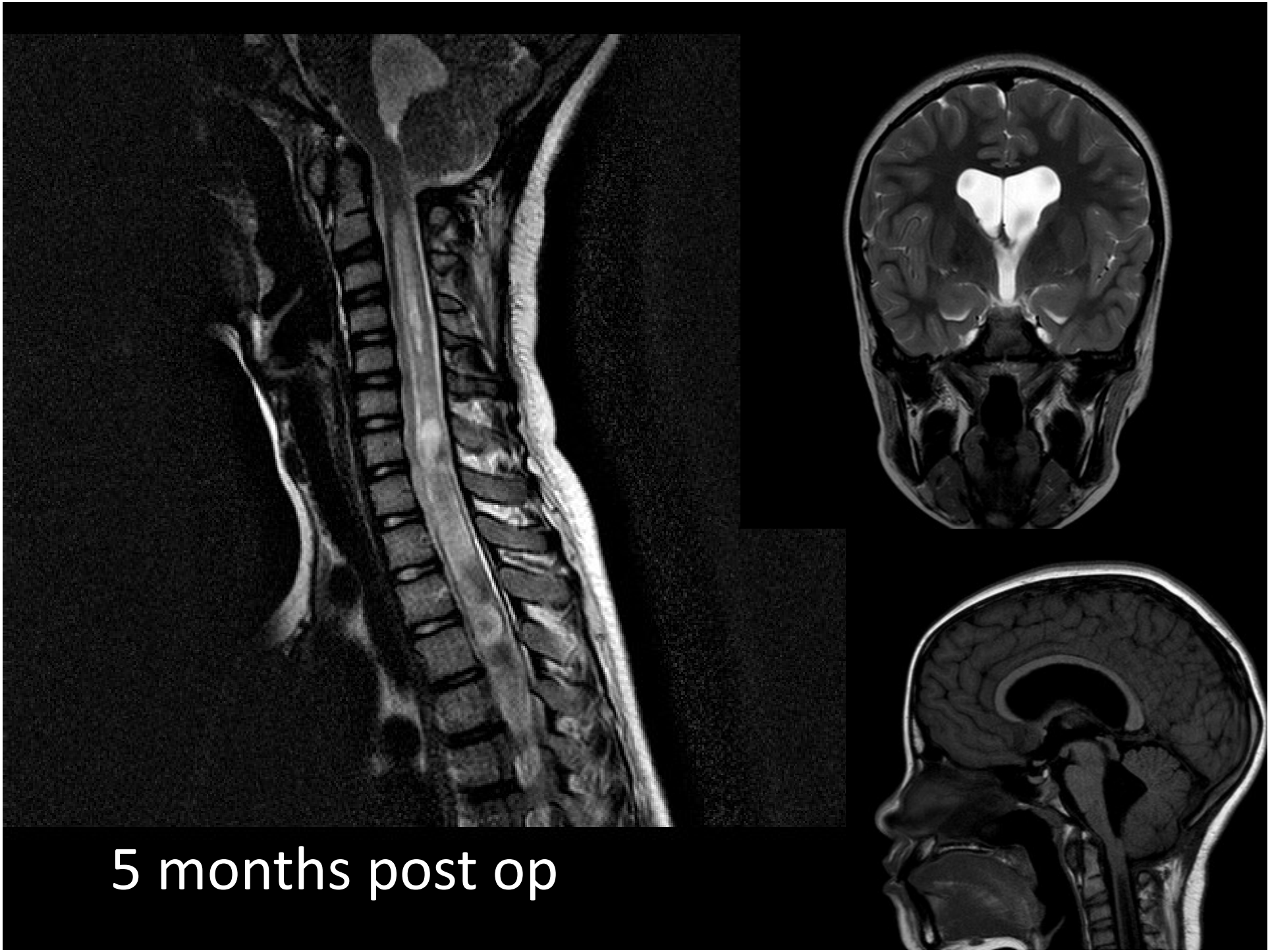








2 months post op


5 months post op

Eule JM, Erickson MA, O'Brien MF, Handler M.
Chiari I malformation associated with syringomyelia and scoliosis: a twenty-year review of surgical and nonsurgical treatment in a pediatric population. Spine (Phila Pa 1976). 2002 Jul 1;27(13):1451-5.

CONCLUSION: Early decompression of Chiari I malformation with syringomyelia and scoliosis resulted in improvement or stabilization of the spinal deformity in 5 cases. Each of these patients underwent decompression before 8 years of age and before the curve was severe. However, this series represents a few patients demonstrating this trend, and further follow-up and investigation are warranted.

Childs Nerv Syst. 2006 Oct;22(10):1351-4.

Scoliosis in a child with Chiari I malformation and the absence of syringomyelia: case report and a review of the literature.

Tubbs RS, Doyle S, Conklin M, Oakes WJ.

CASE REPORT: We report a child with Chiari I malformation and scoliosis who presented with Valsalva-induced headache/neck pain. MRI revealed no syringomyelia or hydrocephalus. Radiographs revealed that her scoliotic curvature was approximately 13 degrees and was a single levoscoliotic curve. This patient underwent a posterior fossa decompression with duraplasty for her Valsalva-induced pain. Postoperatively, she had resolution of her pain and there has been no progression of her scoliosis at 3 years follow-up.

PROGNOSIS: Intriguingly, and scattered throughout the medical literature, many have noted cases of scoliosis in patients with only a Chiari I malformation and no syringomyelia. Moreover, experimental studies have induced scoliosis in animals after compression of the dorsal columns. After a review of the medical literature regarding a potential cause and effect of herniated hindbrain-induced scoliosis in the absence of syringomyelia, this association although rare, does seem plausible.

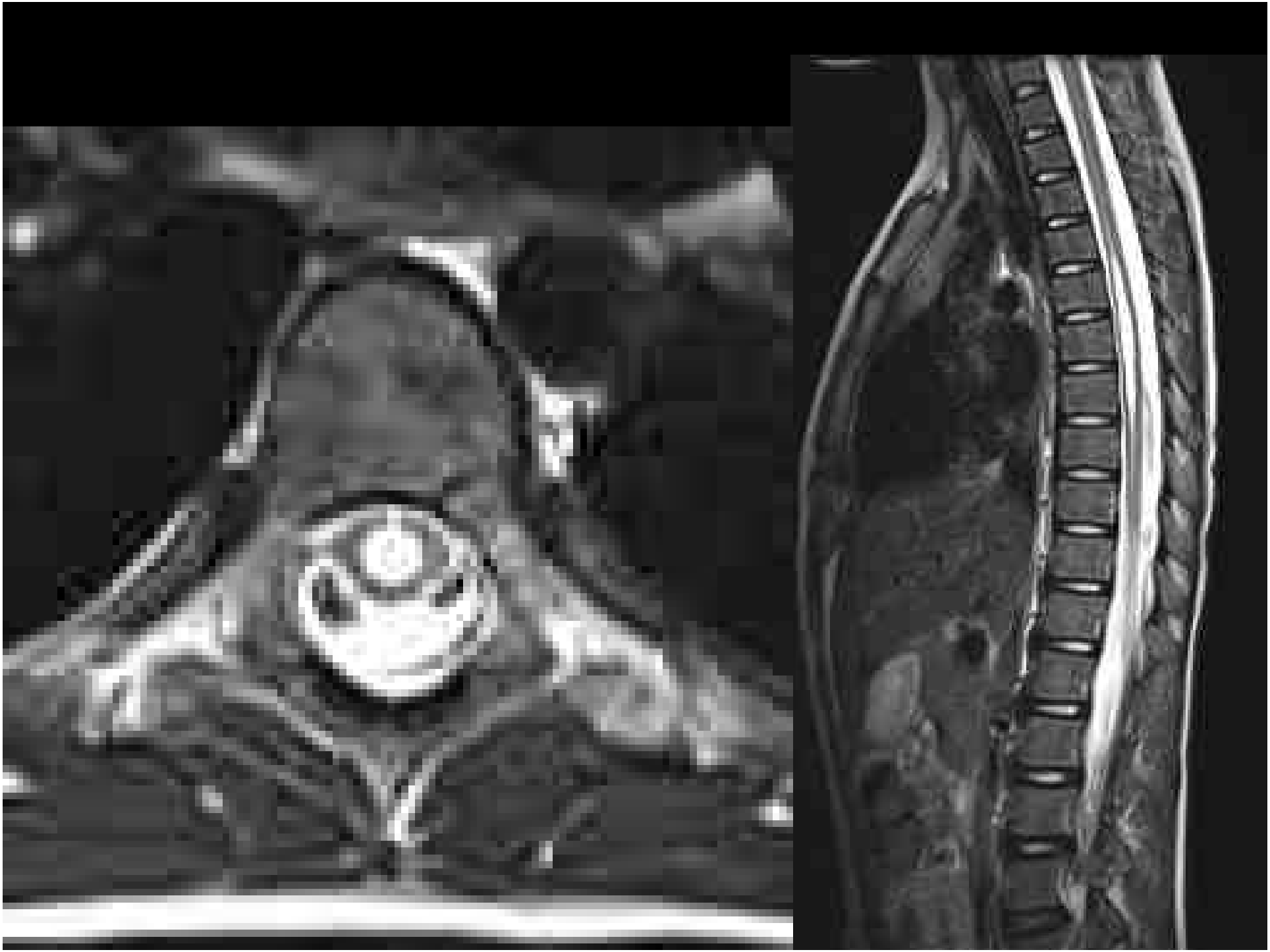
Childs Nerv Syst. 2004 May;20(5):341-8.

International survey on the management of Chiari I malformation and syringomyelia.

Schijman E, Steinbok P.

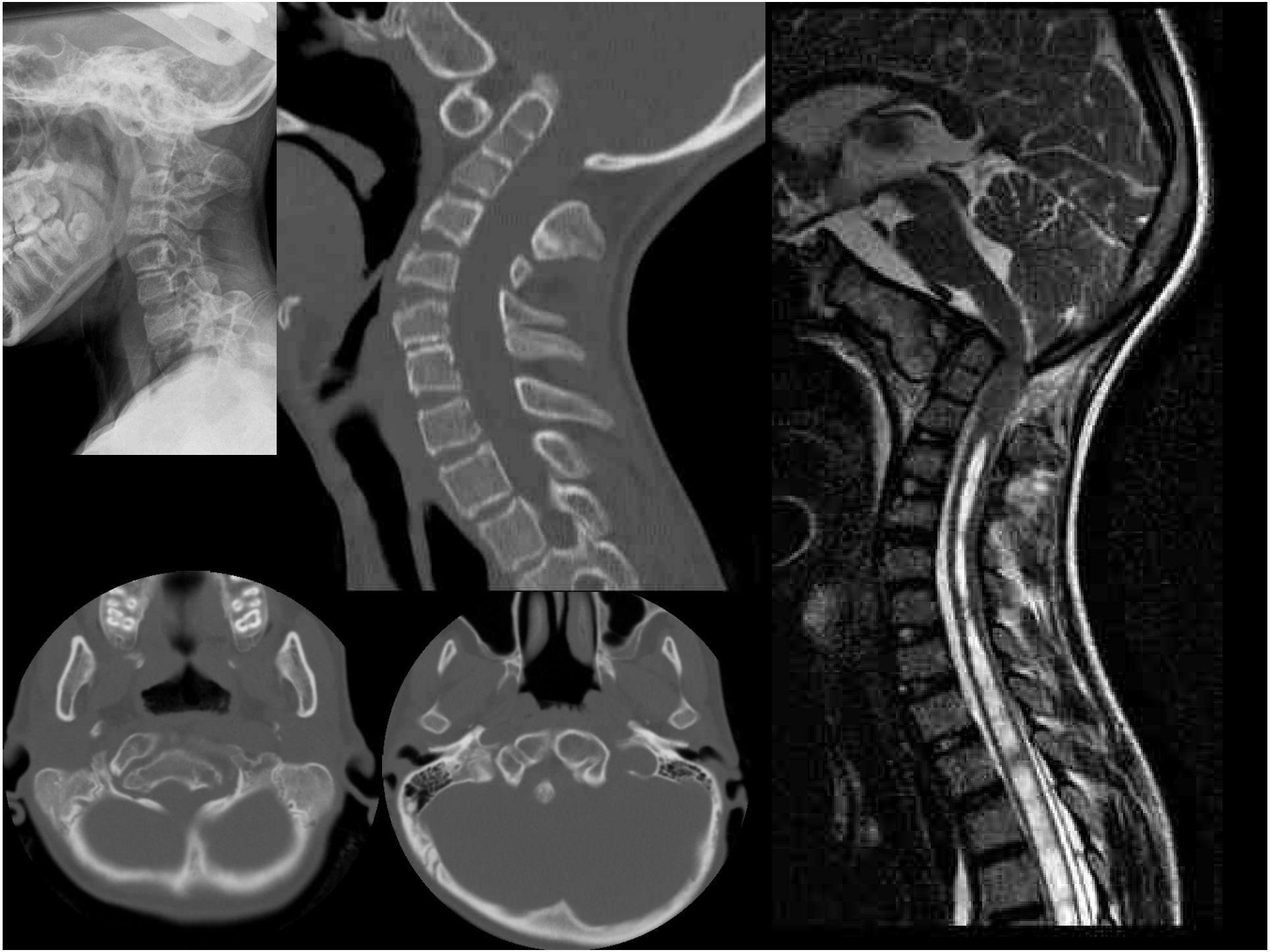
RESULTS: Of 246 questionnaires distributed, 76 (30.8%) were completed and returned. There was a consensus that no operation should be carried out in asymptomatic patients with a Chiari I malformation, unless there is associated syringomyelia. There was a consensus that decompression of the Chiari malformation should be performed in patients with scoliosis when syringomyelia is present, and the majority decompressed the Chiari malformation in scoliotic patients even in the absence of syringomyelia. Suboccipital decompression was the standard surgical procedure for Chiari I malformations. The majority of respondents favored routine dural opening at surgery and closure with a pericranial or synthetic patch graft. In the case of a persistent or progressive syrinx after suboccipital decompression, the majority recommended shunting of the syrinx to the subarachnoid space or to the pleural cavity.

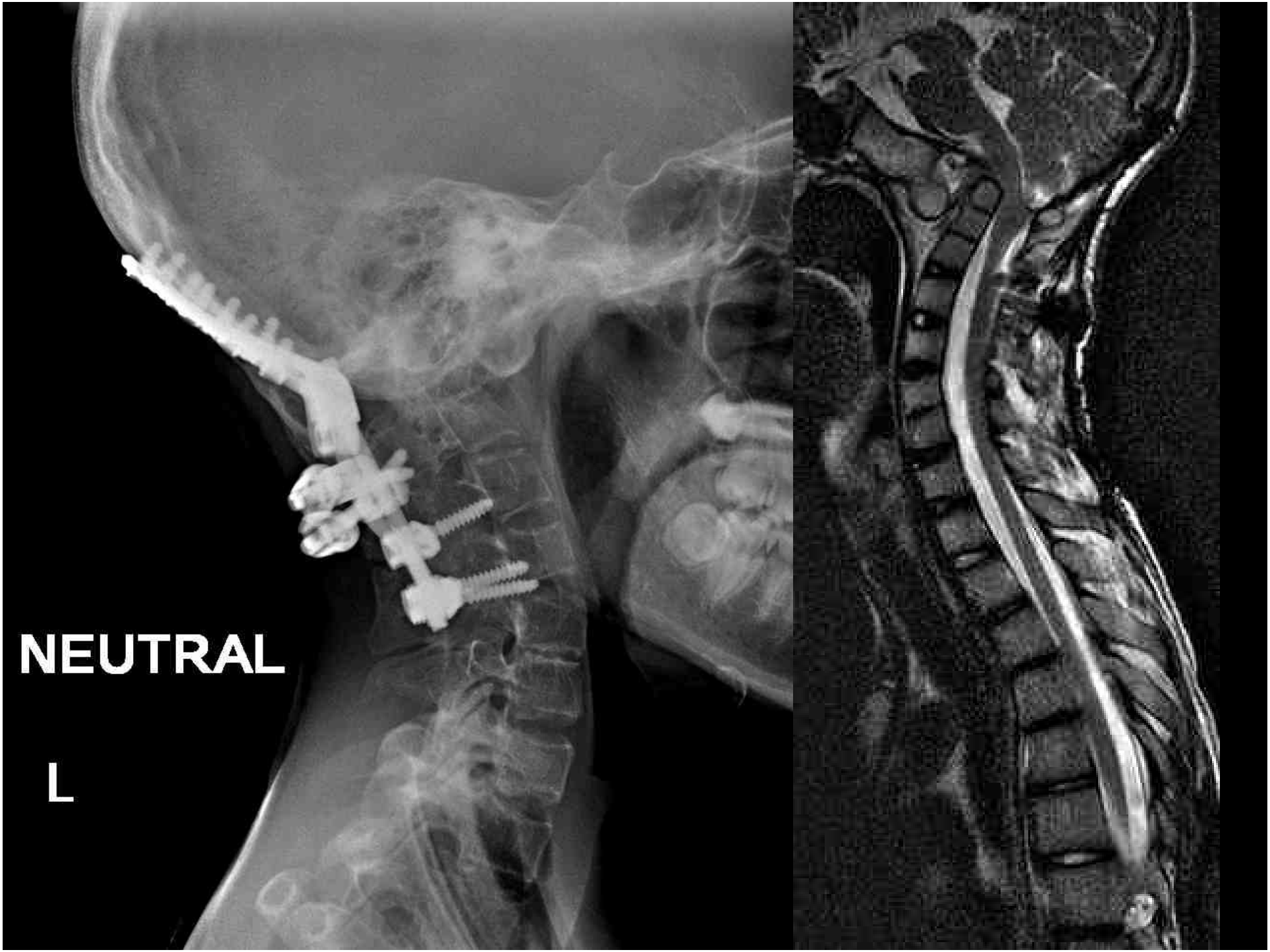
CONCLUSION: There continues to be much variation in the management of the Chiari I malformation.




L
R
3 monthg female
Multiple congenital anomoalies
Chromosome 4 deletion, Wolf Hirschorn Syn


MRI 2 yrs
Clinically well

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Syringomyelia and Scoliosis

- Chiari I, syringomyelia, and scoliosis commonly associated, often good response to surgery, especially if early
- Chairi I and scoliosis without syringomyelia more controversial
- Smaller syrinxes (dilatation of central canal) in absence of any clear pathogenetic mechanism are very common, usually benign, rarely seem to require interention_

J Neurosurg Pediatrics 5:000–000, 2010

Do baclofen pumps influence the development of scoliosis in children?

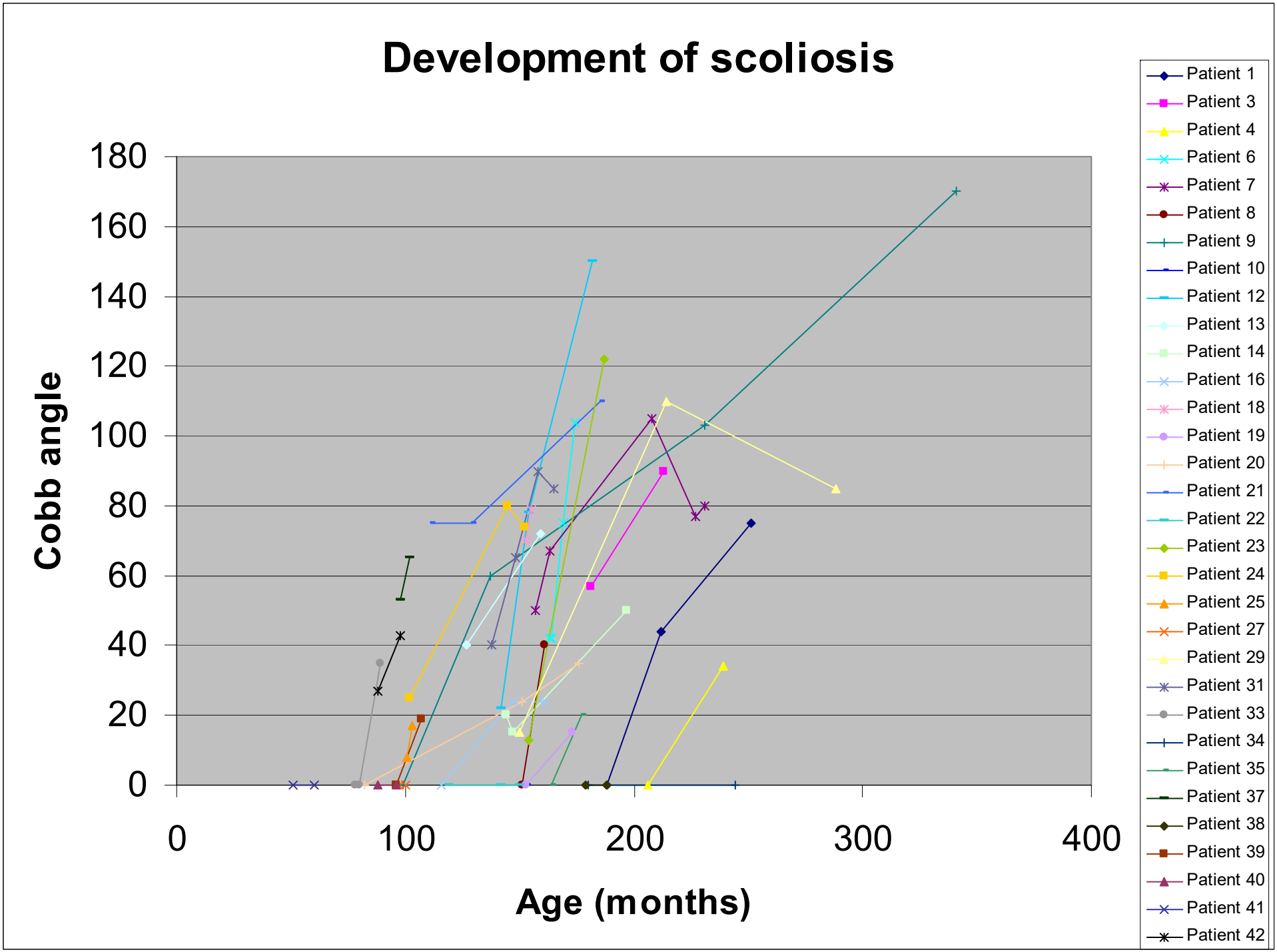
Clinical article

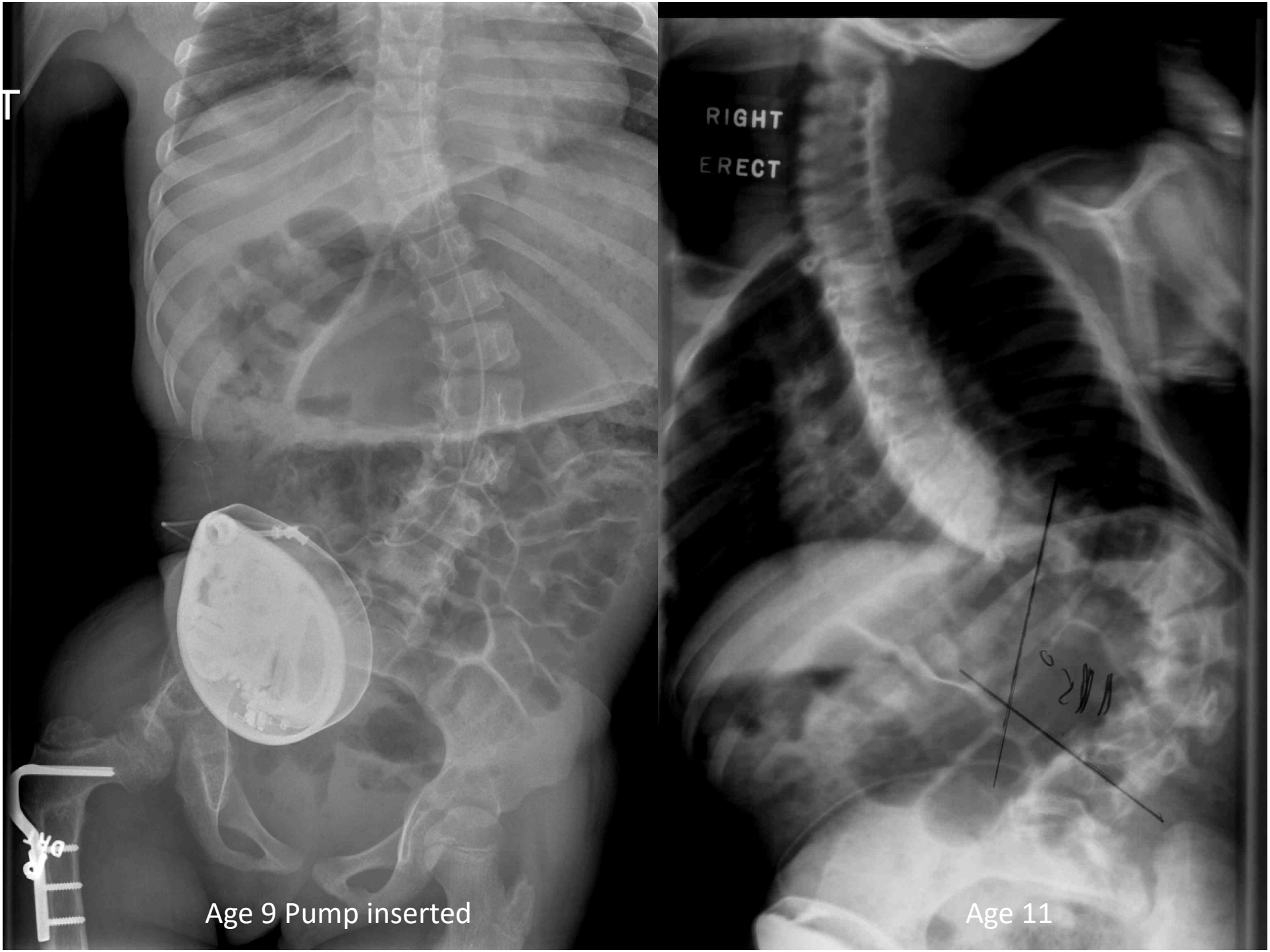
Sasha C. Burn, F.R.C.S.(SN),[1] Reinhard Zeller, F.R.C.S.C.,[2] and James M. Drake, F.R.C.S.C.[1]

Divisions of [1]Neurosurgery and [2]Orthopaedic Surgery, The Hospital for Sick Children, Toronto, Ontario, Canada

	Whole group	CP	CP/dystonia	Head injury	other
Mean	18.40	20.27	15.54	19.77	18.38
Median	11.88	12.26	11.76	5.7	18.48

Table 2 Mean and median annual progression rate of Cobb angle (degrees per year)


Development of scoliosis
Cobb angle
180
160
140
120
100
80
60
40
20
0
0
100
200
300
400
Age (months)
Patient 1
Patient 3
Patient 4
Patient 6
Patient 7
Patient 8
Patient 9
Patient 10
Patient 12
Patient 13
Patient 14
Patient 16
Patient 18
Patient 19
Patient 20
Patient 21
Patient 22
Patient 23
Patient 24
Patient 25
Patient 27
Patient 29
Patient 31
Patient 33
Patient 34
Patient 35
Patient 37
Patient 38
Patient 39
Patient 40
Patient 41
Patient 42



Age 9 Pump inserted

Age 11

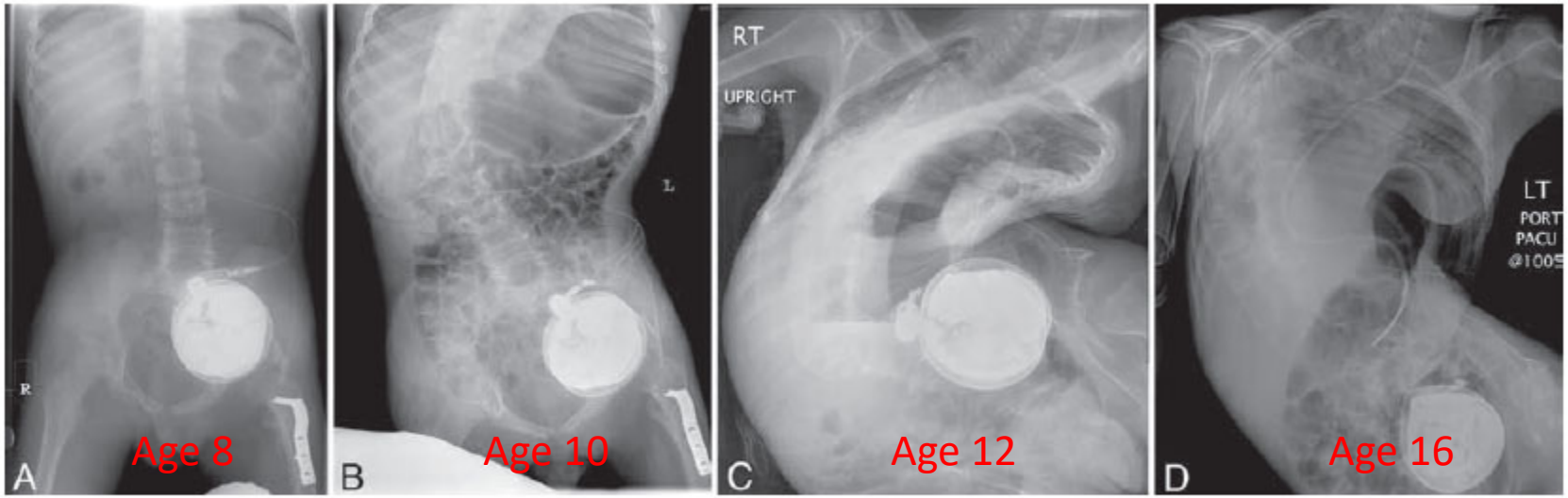


FIG. 2. Case 9. Radiographs demonstrating development of scoliosis following baclofen pump insertion at 8 years of age: at 17 **(A)**, 51 **(B)**, 97 **(C)**, and 118 **(D)** months after insertion of a baclofen pump.

- Above patient died – respiratory failure
- Scoliosis etiology ?
- Intrathecal baclofen human model of scoliosis development ??

Do Baclofen Pumps Cause Scoliosis?

Yes

Spine 2007 Nov 15;32(24):2745-50.

Progression of scoliosis in patients with spastic quadriplegia after the insertion of an intrathecal baclofen pump.

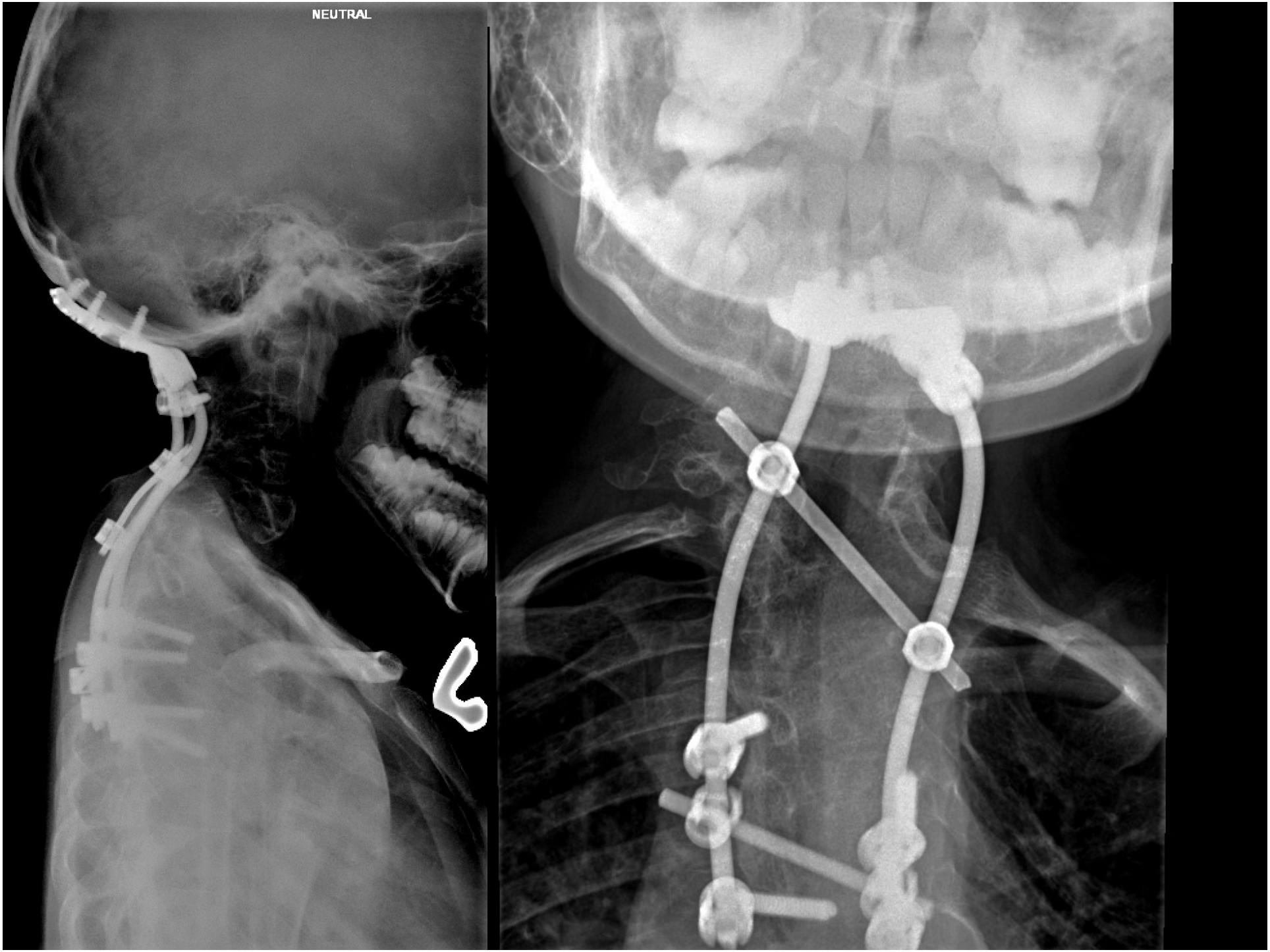
No

J Pediatr Orthop. 2008 Sep;28(6):684-7.

The impact of intrathecal baclofen on the natural history of scoliosis in cerebral palsy.


7 year old neurenteric cyst excised at birth, progressive myelopathy


cut
Ex. Feb
AIL/+
i/10
VOI
120
200
0.50s/HE+ 27.5mm/rot
mm 1.375:1/0.6sp
t: 0.0
38:31 AM
541 L = 160
IPR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Conclusions

- Early onset scoliosis often associated with neuro- anatomic abnormalities or pathologies
- Clear pathogenetic mechanisms are not well understood making decision making difficult
- Careful measured approach, often in multidisciplinary setting, provides reasonable and very satisfying approach