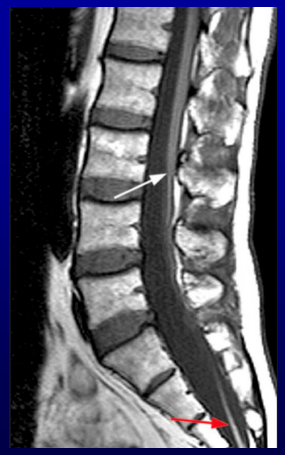
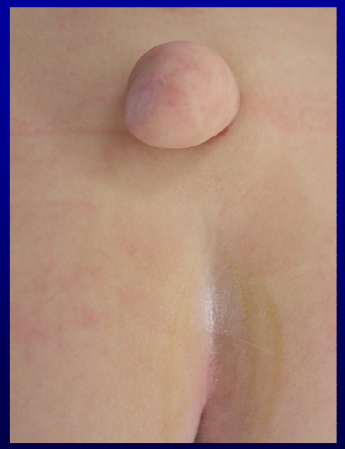
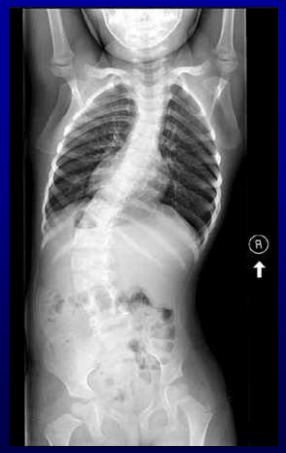
Tethered spinal cord and EOS: When to treat and when to watch?

Richard C. E. Anderson, M.D.

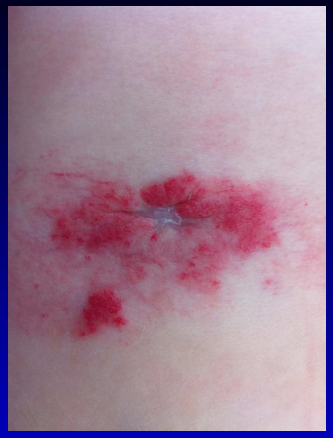


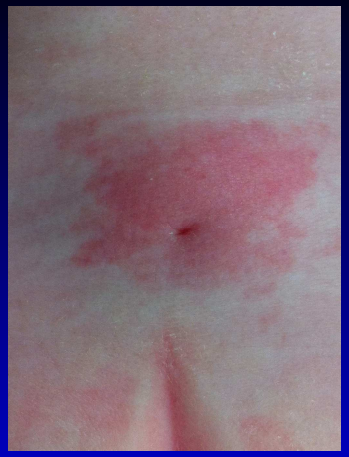


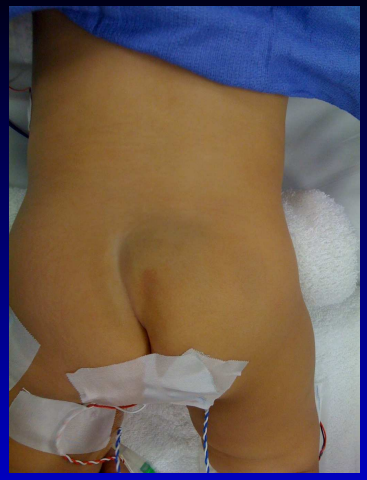


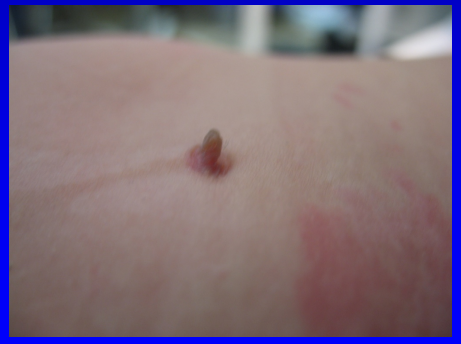
ICEOS
13TH INTERNATIONAL CONGRESS ON EARLY ONSET SCOLIOSIS
NOVEMBER 21-22, 2019

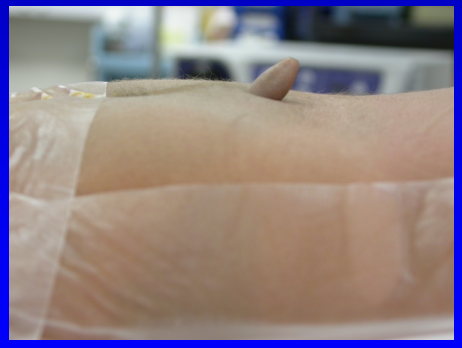
Disclosures up to date

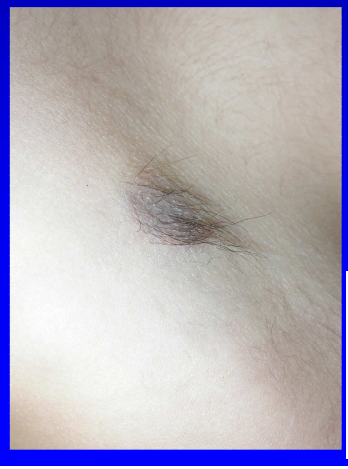








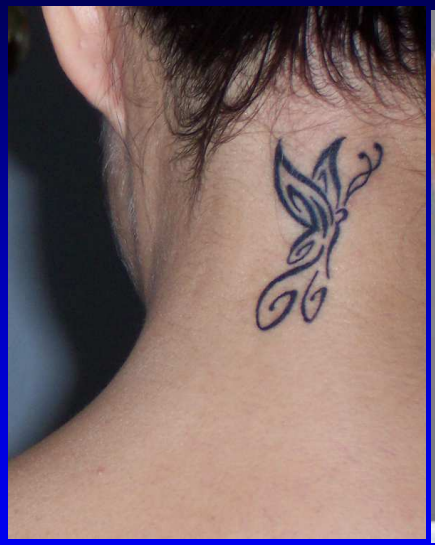




ICEOS

13TH INTERNATIONAL CONGRESS ON EARLY ONSET SCOLIOSIS

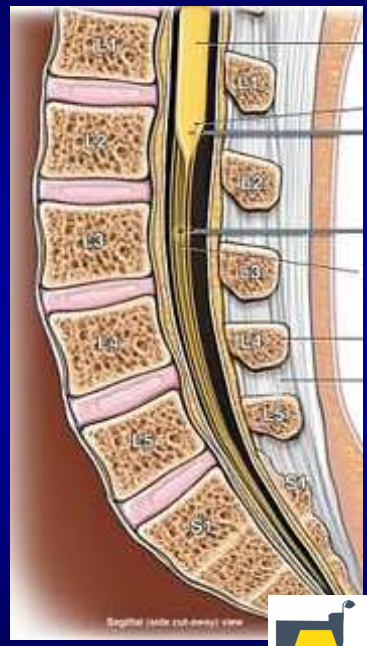
NOVEMBER 21-22, 2019



ICEOS
13TH INTERNATIONAL CONGRESS
ON EARLY ONSET SCOLIOSIS
NOVEMBER 21-22, 2019

Pathophysiology: Tethered Cord Syndrome

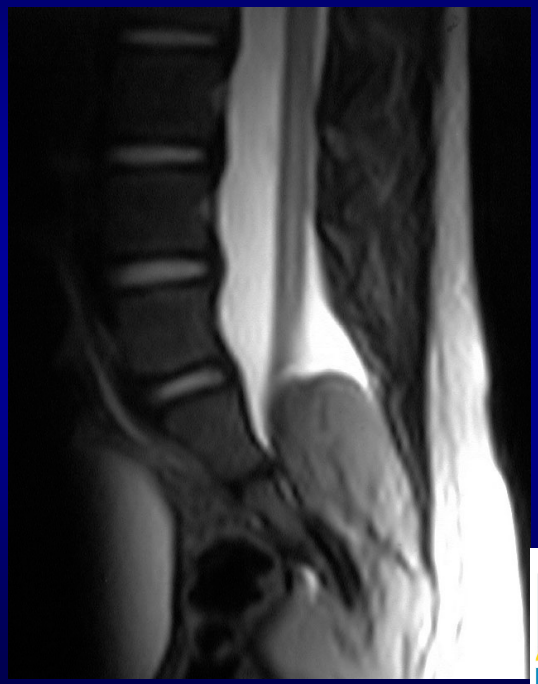
- Relationship between spinal cord and vertebral column changes during development
- Conus lies at approximately:
  - Coccyx 30mm embryo
  - L2-3 Birth
  - L1-2 1 years

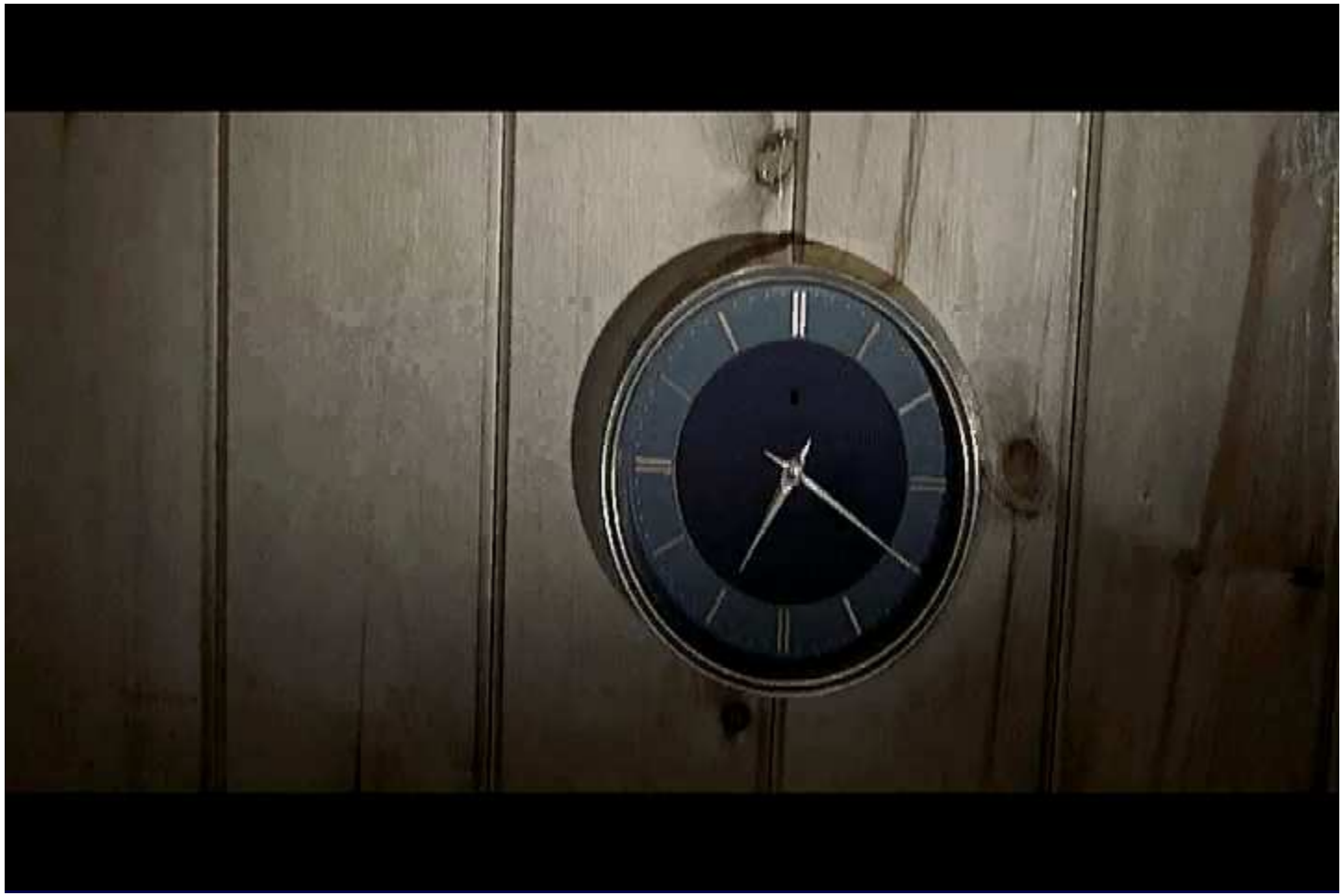


ICEOS
13TH INTERNATIONAL CONGRESS ON EARLY ONSET SCOLIOSIS
NOVEMBER 21-22, 2019

Pathophysiology: Tethered Cord Syndrome

- Causes neurological impairment by mechanically anchoring the spinal cord at the site of the cord's fusion with the lesion
- Leads to traction on the spinal cord with resultant relative ischemia
- If surgery is not performed, neurological deficits frequently are progressive (30-40%)
- Once a neurological deficit develops, it is often irreversible





Why be concerned about TSC with EOS?

- Intraspinal anomalies seen in 20-50% of children with congenital scoliosis
- TSC release can delay the progression of scoliosis
- Deformity correction in the setting of untreated TSC may be problematic
  - Neurological injury
  - Unreliability or loss of IOM signals limiting correction

Table 2. Intraspinal anomalies

Serial no.	Intraspinal anomaly	No. of patients (56/119)
1	Tethered cord and low conus	27 (48.2)
2	Diastematomyelia	18 (32.1)
3	Syringomyelia	17 (30.3)
4	Arnold Chiari malformation	11 (19.6)
5	Intradural cyst	1 (1.7)
6	Intradural lipoma	2 (3.5)

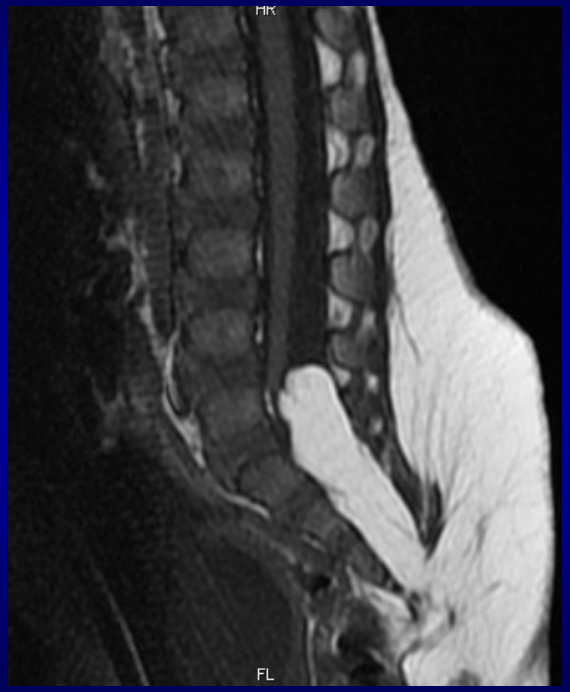
Values are presented as percent (number).
35.7% patients (20/56) had multiple intraspinal abnormalities.

Gupta et al (2016) Asian Spine J

ICEOS
13TH INTERNATIONAL CONGRESS ON EARLY ONSET SCOLIOSIS
NOVEMBER 21-22, 2019

When to act

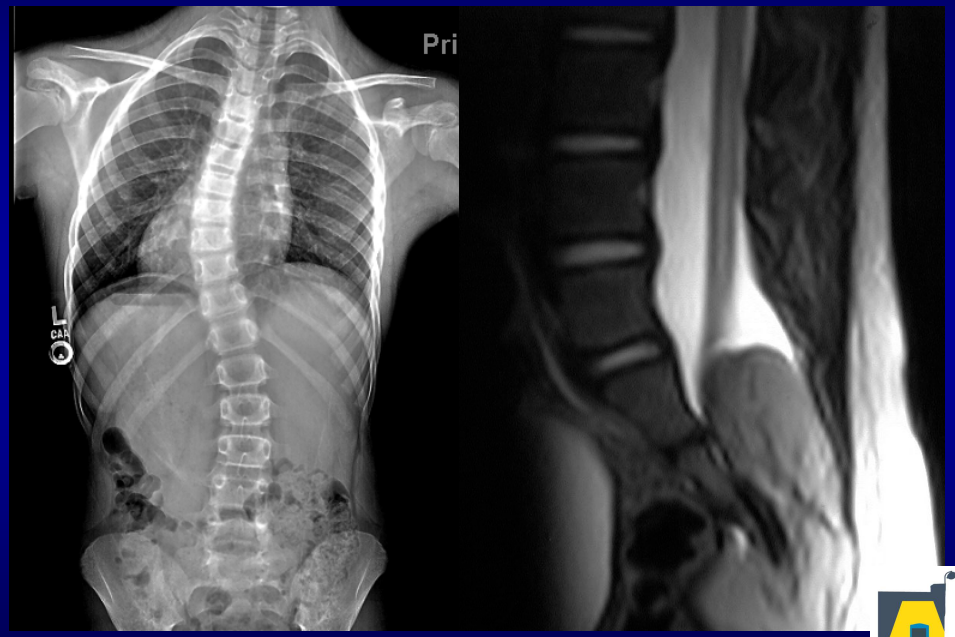
- Clear TSC with low lying conus:
  - Spinal cord lipomas
  - Split cord malformations
  - *Symptomatic* myelomeningoceles
  - Dermal sinus tracts
  - Others



ICEOS
13TH INTERNATIONAL CONGRESS ON EARLY ONSET SCOLIOSIS
NOVEMBER 21-22, 2019

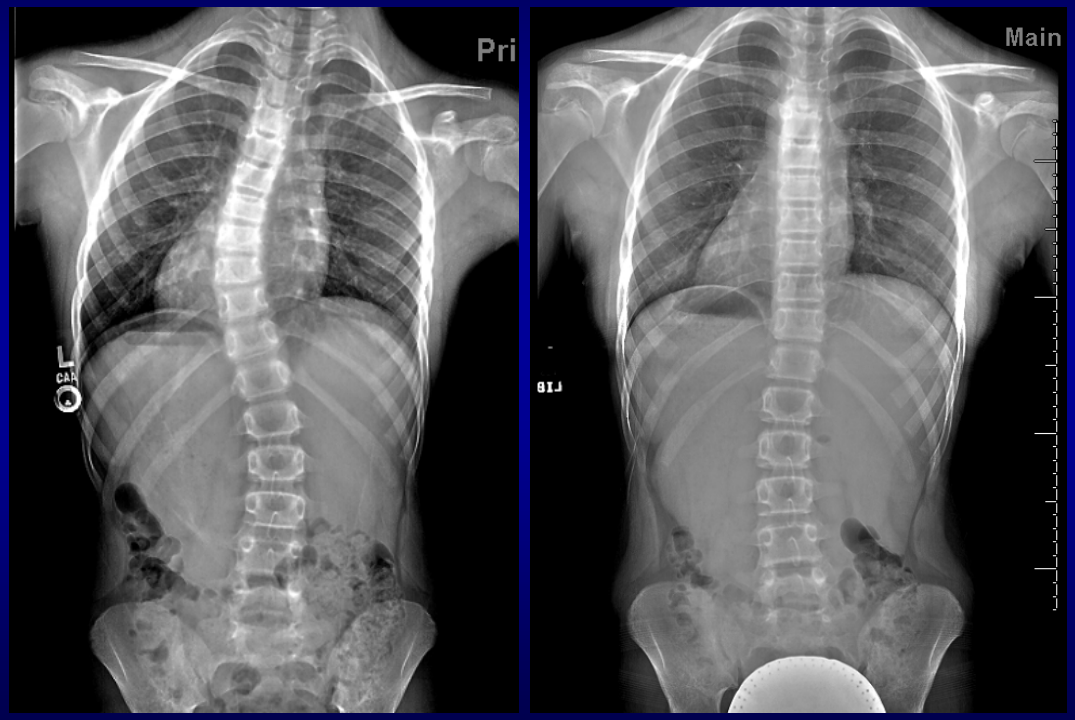
When to act

- 6 year old girl presented with scoliosis
- Back pain
- No skin lesions
- Neuro intact
- Underwent TSC release



ICEOS
13TH INTERNATIONAL CONGRESS ON EARLY ONSET SCOLIOSIS
NOVEMBER 21-22, 2019

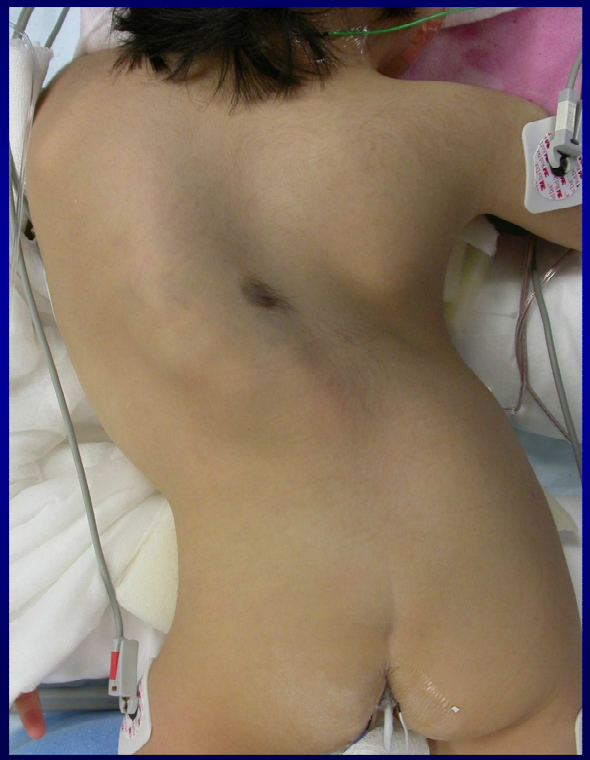
When to act



ICEOS
13TH INTERNATIONAL CONGRESS ON EARLY ONSET SCOLIOSIS
NOVEMBER 21-22, 2019

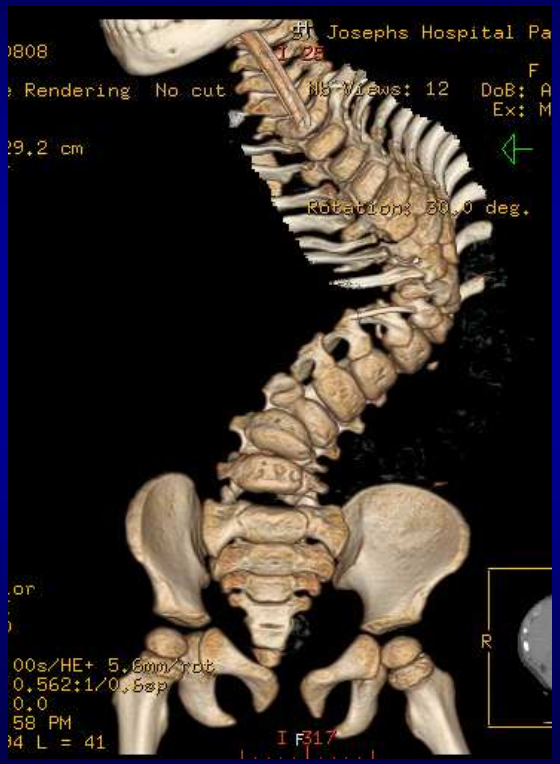
When to act

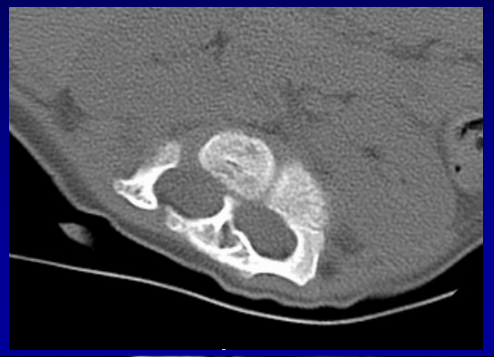
- 3 year old girl with progressive scoliosis and hairy patch in thoracic region

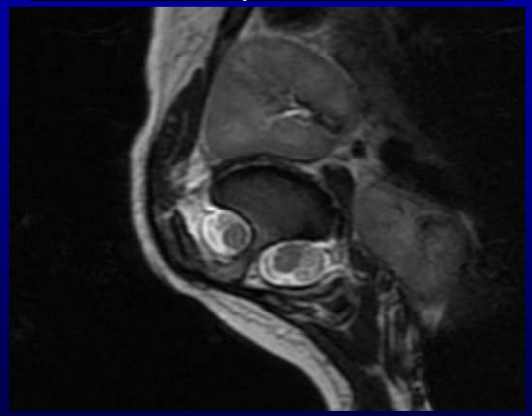


ICEOS
13TH INTERNATIONAL CONGRESS ON EARLY ONSET SCOLIOSIS
NOVEMBER 21-22, 2019

When to act







ICEOS
13TH INTERNATIONAL CONGRESS ON EARLY ONSET SCOLIOSIS
NOVEMBER 21-22, 2019

When to act

ICEOS
13TH INTERNATIONAL CONGRESS ON EARLY ONSET SCOLIOSIS
NOVEMBER 21-22, 2019

Effect of TSC release on scoliosis

- Bowman/McLone study:
  - 36pts, 7yr follow-up
  - Myelo only
  - 33% progression at 2 years
  - 66% progression at 7 years (36% fusion)
- Pierz study:
  - 21 pts, 5yr f/u
  - 57% progression
- Hassani study:
  - 20 pts, 4yr f/u
  - 60% progression (50% fusion)
- Newton study:
  - 15pts (10 TSC)
  - 47% progression
- Chern study:
  - 14pts
  - All tight filum
  - 64% progression (36% fusion)

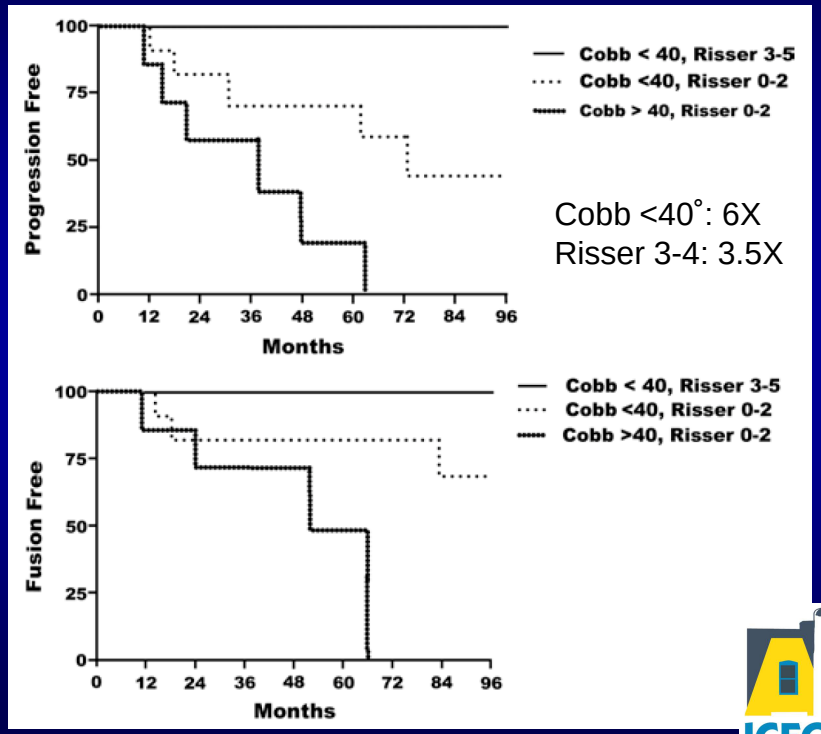
Effect of TSC release on scoliosis

TABLE 2: Summary of patient demographics, symptomatology, and scoliosis measurements*

Variable	Value
demographics	
mean age (yrs)	8.9 ± 3
male	16 (59)
skeletal maturity at TCS	
Risser Grades 0–2	17 (63)
Risser Grades 3–5	10 (37)
cause of TCS	
postmyelomeningocele repair	14 (52)
fatty filum	5 (18.5)
lipomeningocele	3 (11)
diastematomyelia	2 (7.4)
arthrogryposis	1 (3.7)
lipomyelomeningocele w/ occult dysraphism	1 (3.7)
imperforate anus w/ S-2 hemivertebrae	1 (3.7)

TABLE 3: Five-year incidence rate of radiological progression and subsequent fusion in 4 subgroups of patients with TCS-associated spinal deformity

Cobb Angle/ Risser Grades	No. of Patients	% w/ Radiological Progression	% w/ Subsequent Fusion
<40°/3–5	8	0	0
<40°/0–2	11	54	27
>40°/3–5	2	50	0
>40°/0–2	6	83	83



McGirt M et al (2009) JNS Peds

ICEOS
13TH INTERNATIONAL CONGRESS ON EARLY ONSET SCOLIOSIS
NOVEMBER 21-22, 2019

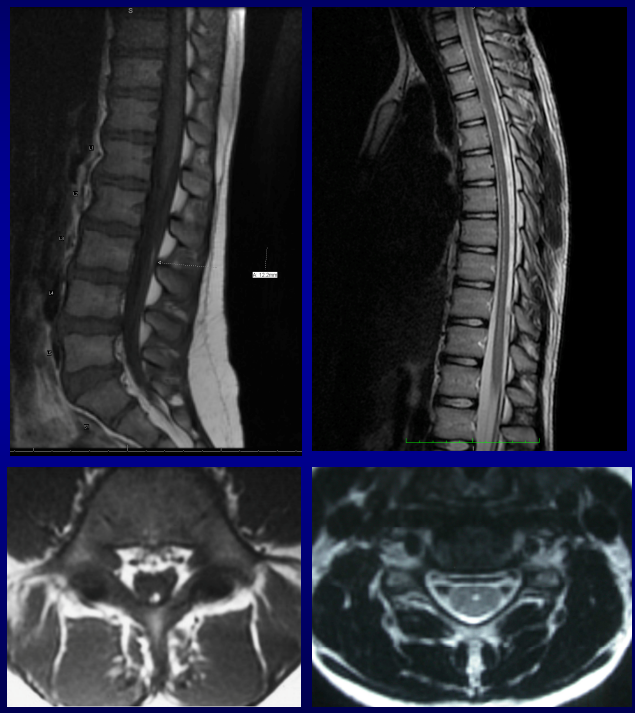
Effect of TSC release on scoliosis

- Higher risk of curve progression after TSC release:
  - *Bigger curve (Cobb angle >35-45˚)*
  - Younger age (<10 yrs)
  - Skeletal immaturity (Risser 0-2)
  - Thoracic level myelomeningocele
  - Greater vertebral rotation
- Delays the need for definitive scoliosis surgery by several years

McGirt M (2009) JNS Peds; Altiok H (2016)Top SCI Rehabil; Bowman RM (2009) JNS Peds; Jankowsky PP (2016) Spine

When NOT to act

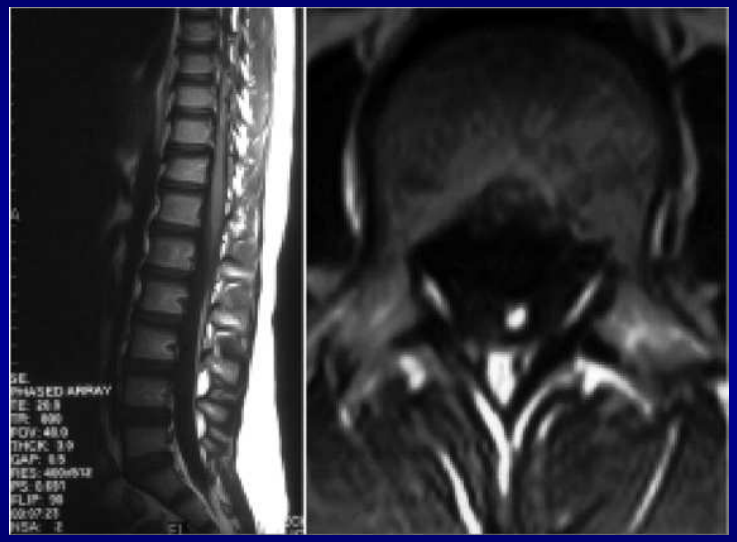
- Radiologists call “syrinx” when <3mm: dilation of the central canal
- Radiologists call “filum lipoma” when trace fat seen in filum (6% autopsy and MRI; 0.4% deterioration)
- *Asymptomatic* myelomeningocele
- Vast majority of cases with normal level conus



Cools MJ (2016) JNS Peds; Maher CO (2015) Pediatrics

When is there equipoise?

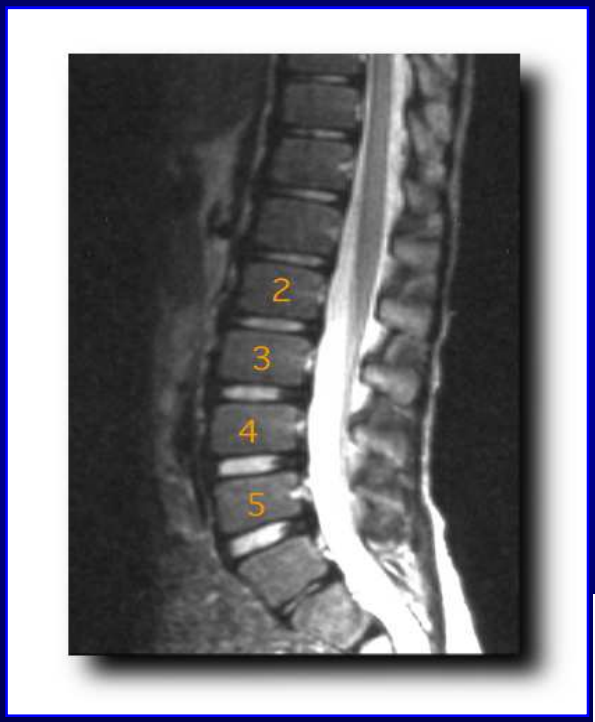
- Concept of an occult tethered cord syndrome (OCTS) first reported in 1990 by Khoury et al.
- Similar symptoms/signs of tethered cord syndrome but conus lies in a normal position at or above L2
- Can account for up to 20% of cases in large series



ICEOS
13TH INTERNATIONAL CONGRESS ON EARLY ONSET SCOLIOSIS
NOVEMBER 21-22, 2019

When is there equipoise?

- Key Question:
  - What percentage of patients with progressive scoliosis and are otherwise neurologically and urologically normal with a normal level conus have OTCS?
- Answer:
  - Unknown
  - *No definitive test exists to identify OTSC*



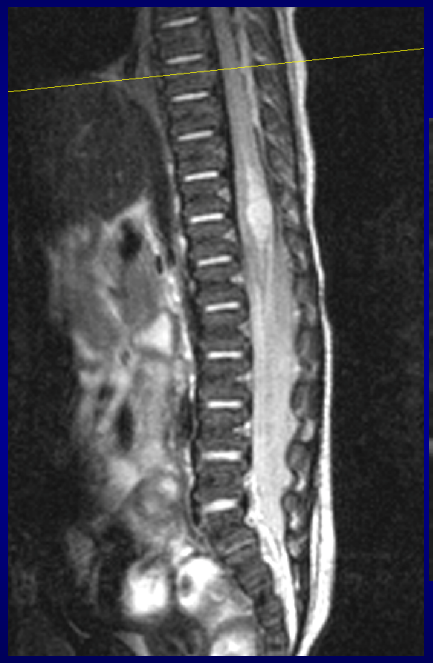
ICEOS
13TH INTERNATIONAL CONGRESS ON EARLY ONSET SCOLIOSIS
NOVEMBER 21-22, 2019

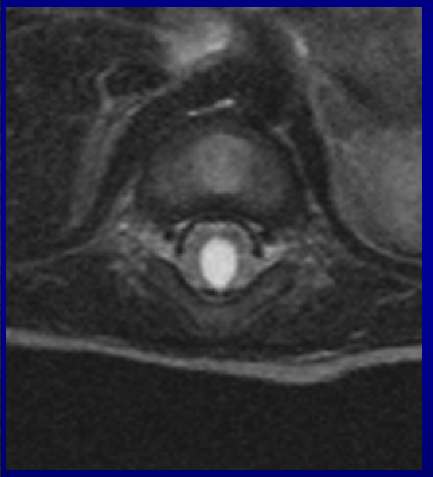
When is there equipoise?

- Consider TSC release if atypical or rapidly progressive curve under the following circumstances:
  - Any symptoms of TSC (progressive pain, weakness, sensory loss, urologic dysfunction)
  - Additional radiographic findings including fatty filum, syrinx, or vertebral body anomalies
- Why?:
  - Morbidity is low (1-2%)
  - Loss of potentials in the OR makes for a very bad day
  - EOS patients extremely difficult to manage
  - In some patients, it will buy some time

Occult TSC

- 3 month old girl born with sacral-coccygeal pit
- Gets MRI for evaluation
- Neuro exam normal

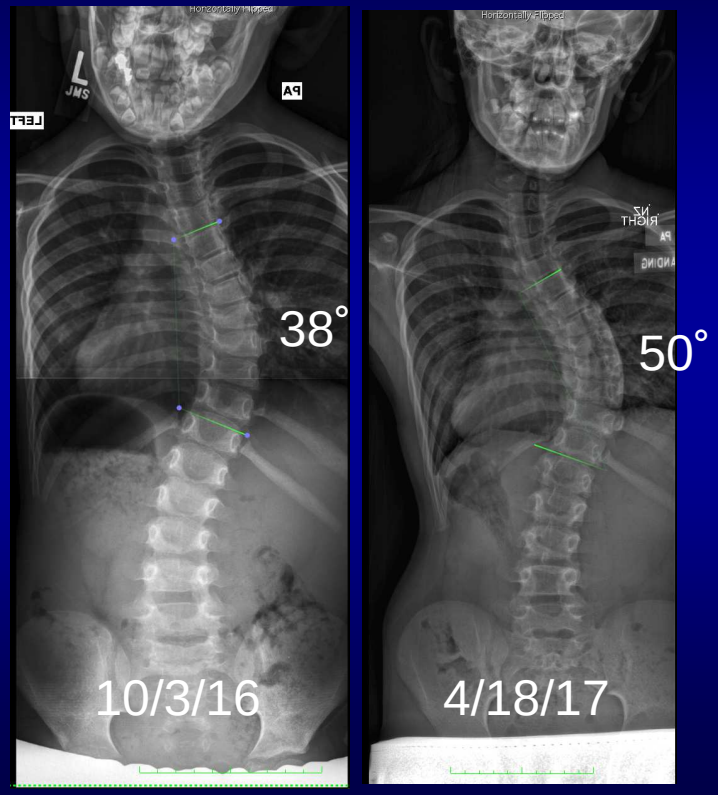




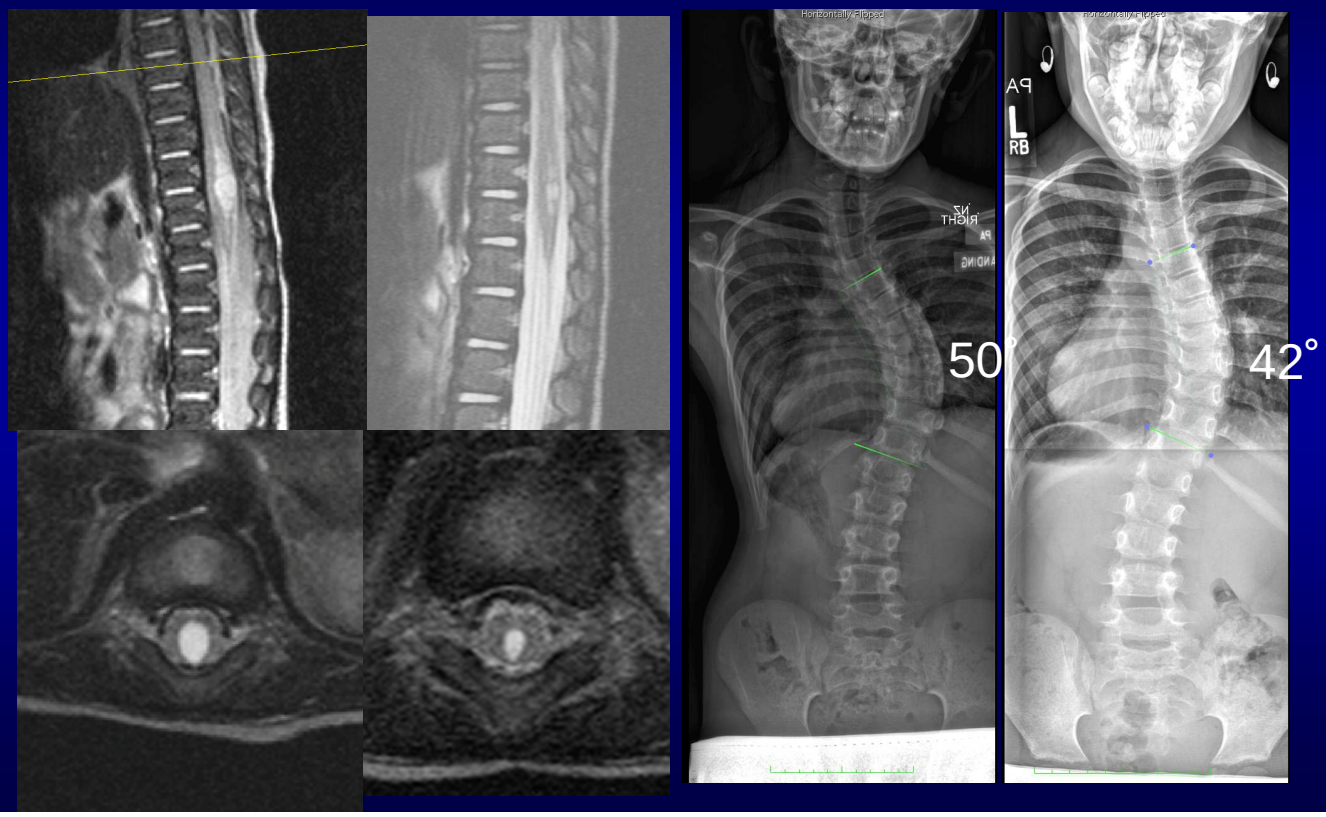
ICEOS
13TH INTERNATIONAL CONGRESS ON EARLY ONSET SCOLIOSIS
NOVEMBER 21-22, 2019

Occult TSC

- At 6 years of age, develops rapidly progressive scoliosis over 6 month period
- Repeat MRI shows no change
- Neuro exam remains normal



Occult TSC



Conclusions

- When to Treat:
  - Clinical symptoms of TSC
  - MRI showing low level conus
  - Consider occult TSC if rapidly progressive curve with additional symptoms or radiographic signs
- When to Watch:
  - Reports of “syrinx” < 3mm diameter
  - Reports of “fatty filum” with normal level conus and no other symptoms

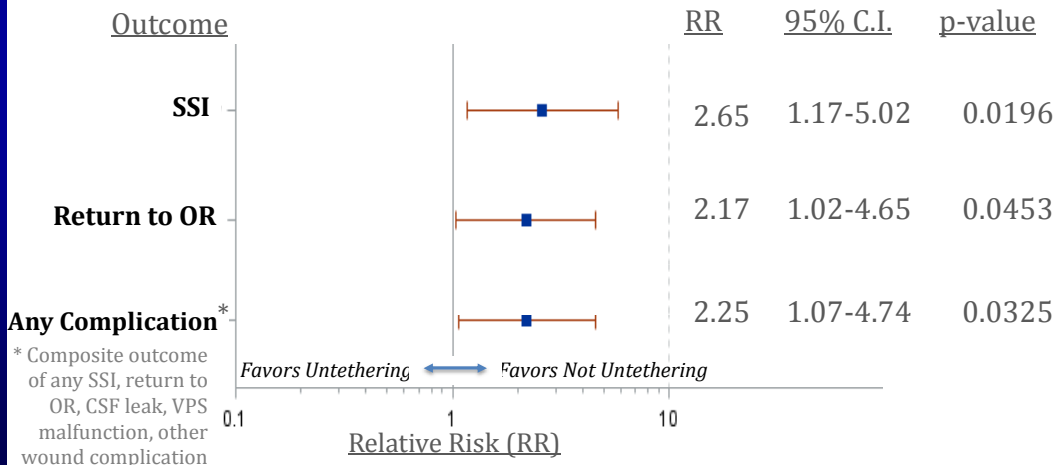
Thank you!

When NOT to act

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



Goldstein H (2019) Childs Nerv System

ICEOS
13TH INTERNATIONAL CONGRESS ON EARLY ONSET SCOLIOSIS
NOVEMBER 21-22, 2019