Early Onset Scoliosis

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dayton children's

Defining Early Onset Scoliosis (EOS)

- EOS generally refers to pathologies of the growing spine that manifest and progress in different forms
- Ponseti and Friedman 1950 (1)
  - First used to describe idiopathic scoliosis in patients < 10 years old
- James 1954 (2)
  - Subdivided idiopathic scoliosis based on age of onset: infantile (<3 years), juvenile (4-9 years), adolescent (10 years – maturity)
- Growing spine study group (GSSG) (3)
  - Any spinal deformity that is present before the age of 10 regardless of etiology

Etiology

Idiopathic

- No causal agents or association with other diseases

Neuromuscular

- Underlying neuropathic or myopathic disease resulting in muscle tone imbalance

Congenital (Structural)

- Defects during vertebral development that lead to asymmetric growth

Syndromic

- Clinically defined patterns associated with scoliosis not directly attributable to congenital or NM causes (Marfans, EDS, NF, Noonan)

Natural History

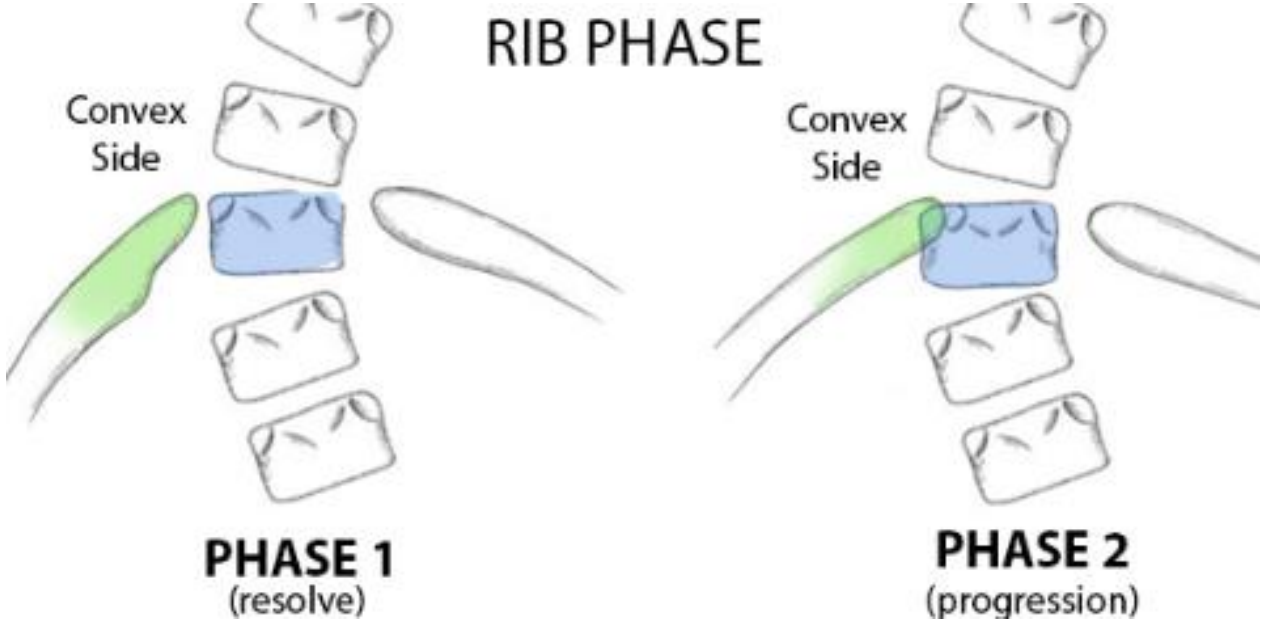
- EOS can either be progressive type or resolving type
- Progressive EOS
  - Earlier the onset, worse final curvature and prognosis
  - Cardiopulmonary deterioration and increased mortality (4,5)
- Increased mortality is explained by:

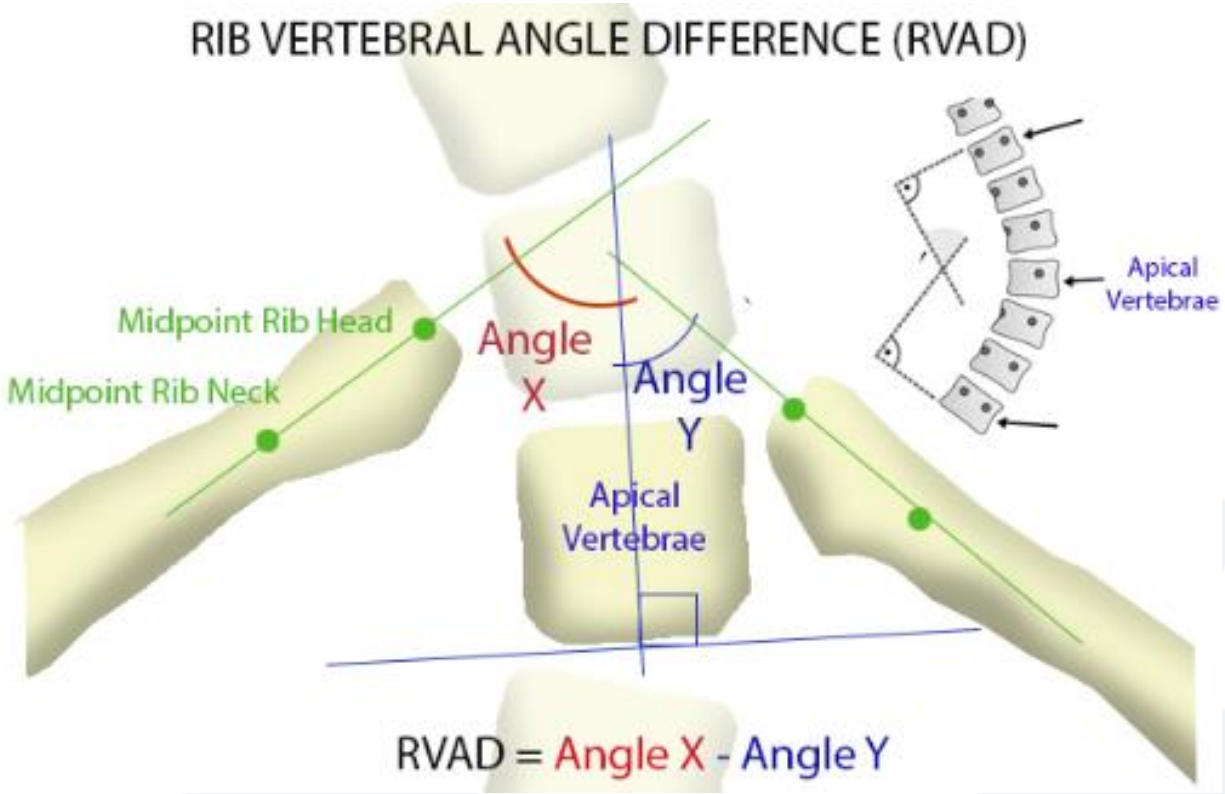
Constraints on the thoracic cavity → Restrictive Lung Disease → Cardiovascular complications → Respiratory failure

Progressive vs. Resolving EOS

- Cobb angle
  - >20 degrees associated with progression
- Rib phase
  - Phase 2 associated with progression
- Rib vertebral angle difference (RVAD)
  - >20 degrees associated with progression

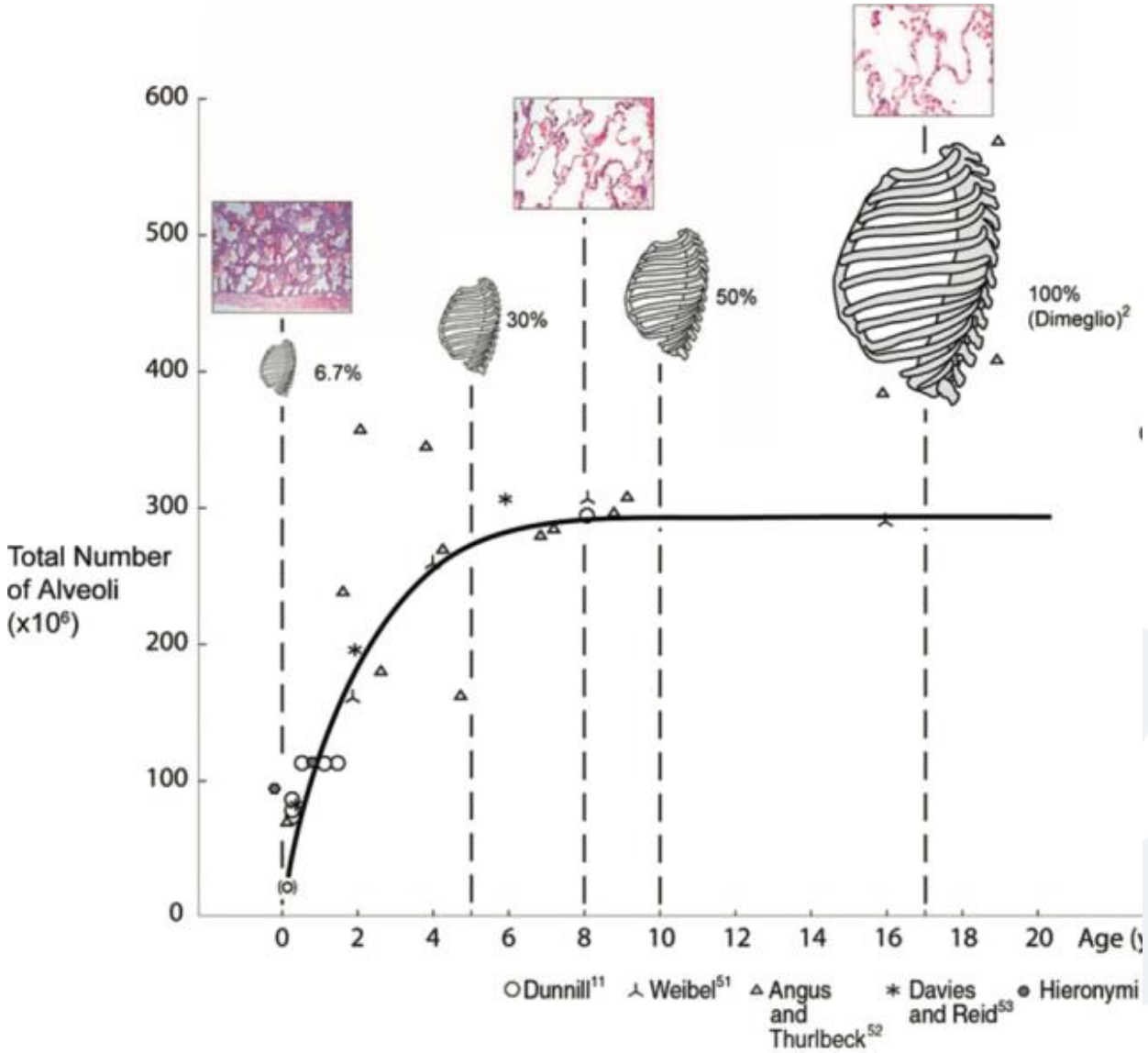
Progressive vs. Resolving EOS





Lung Development

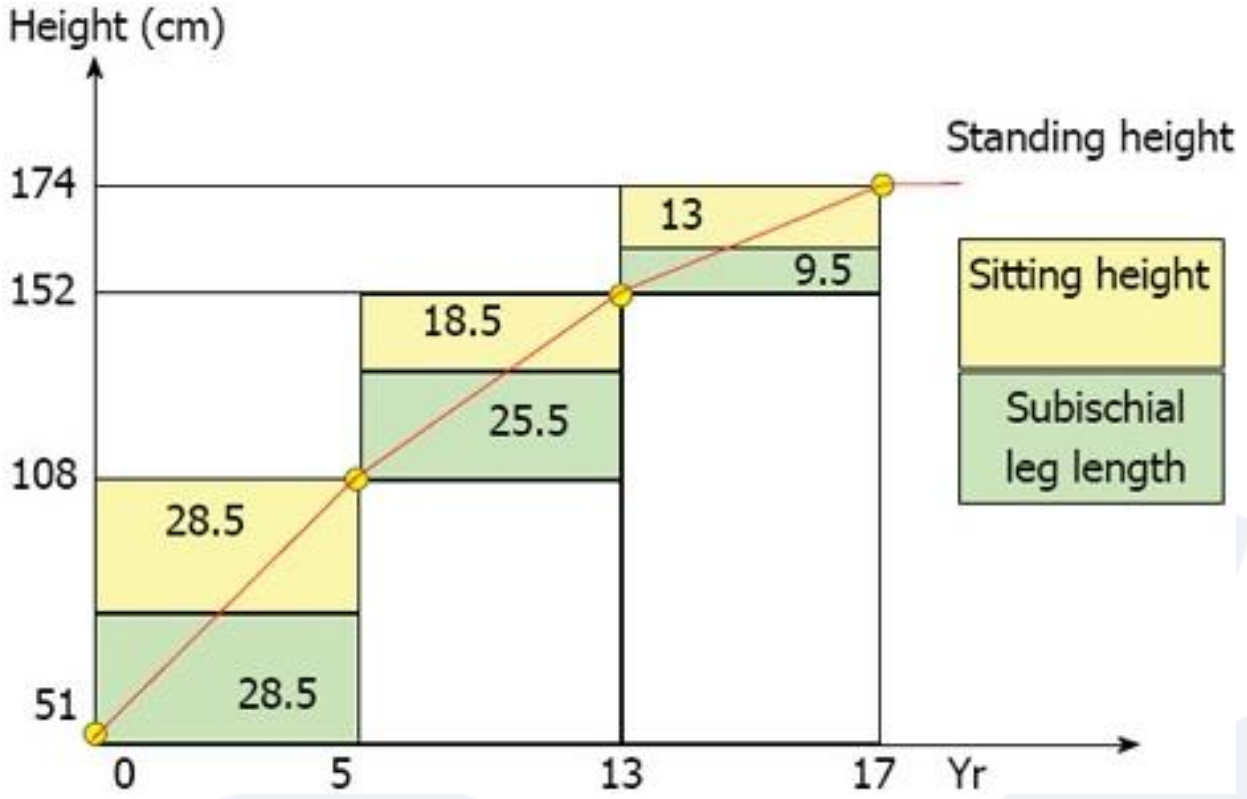
- Most rapid period of development occurs during first 8 years of life, exponential growth (6)
- By age 10, typically see 50% of normal adult volume
- Early development of scoliosis constrains thoracic cavity leading to lung hypoplasia and emphysematous changes (7)



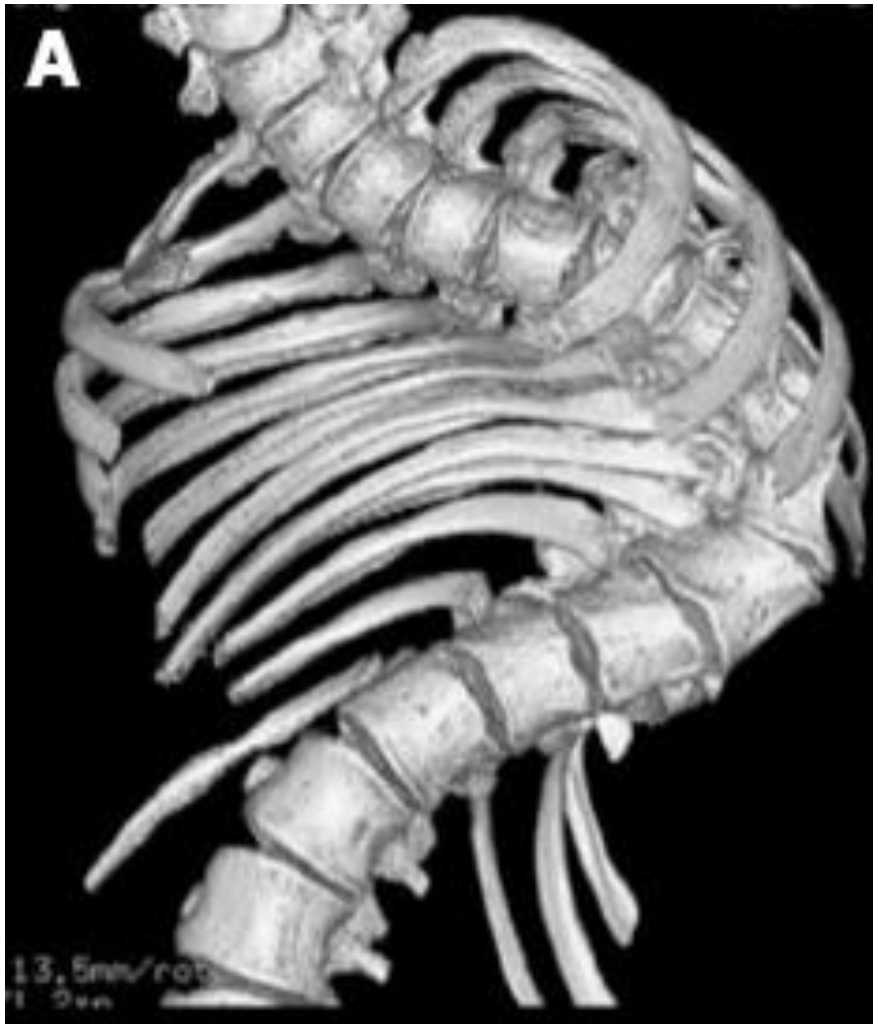
Spine Development

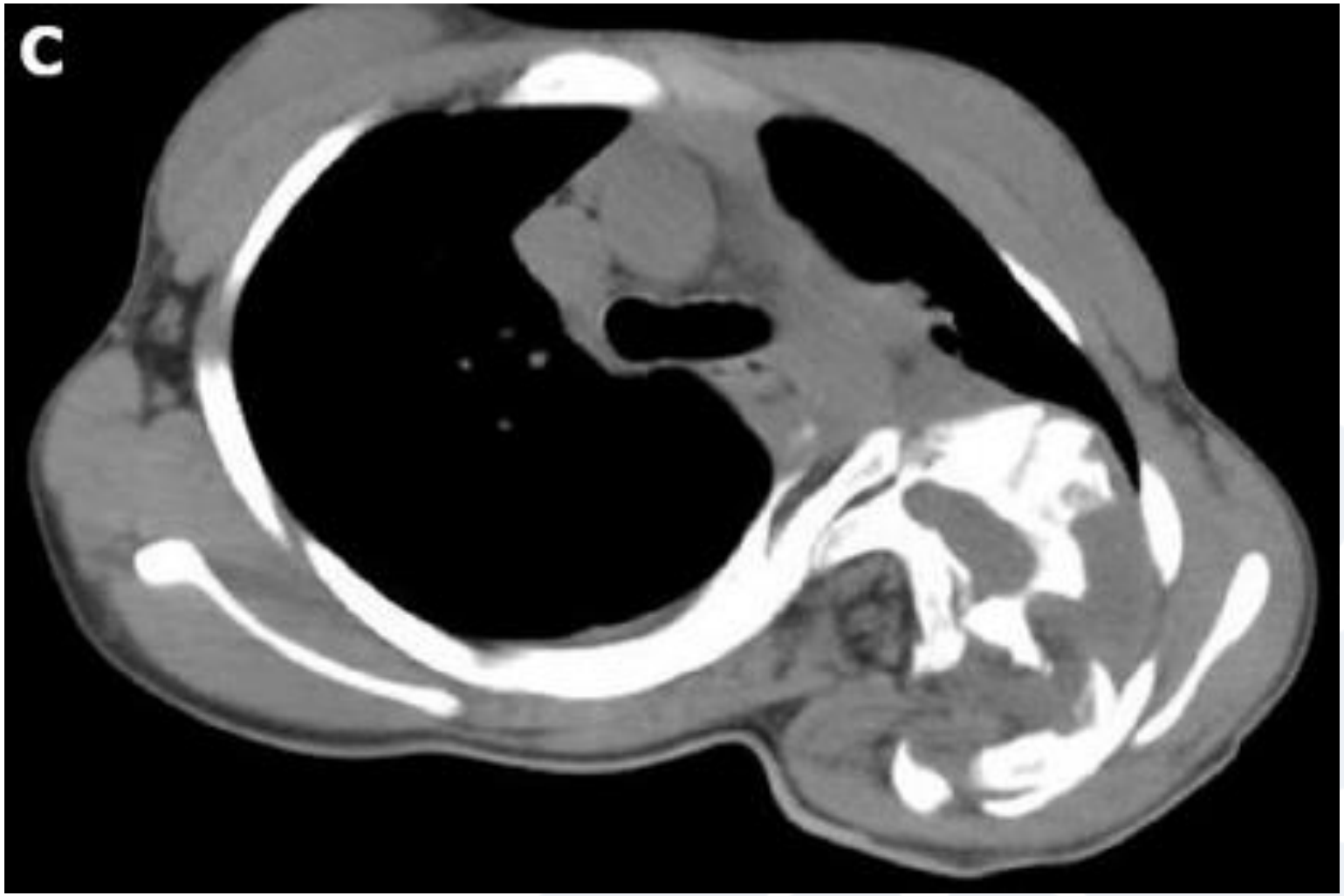
- Spinal growth is the product of more than 130 growth plates working at different paces (8)

- Spine growth is a succession of acceleration and deceleration phases comprising 3 periods (8)
  - Birth – age 5: rapid spinal growth (2.2 cm/year, 12cm in first year of life)
  - 5 – 10 years: quiescent phase (2.2 -> 1.1 cm/year, more lower extremity growth)
  - Puberty: Remaining standing height 20cm, 2/3 is at the level of the trunk



Spine Development





Psychological Burden

- Patients with EOS often subject to multiple medical visits, hospitalizations, comorbidities, repetitive surgeries
- EOS patients
  - Depression and anxiety more prevalent, and dysfunctional areas of daily living (9)
- 58% of individuals with repeated surgical intervention show
  - Conduct problems, aggression, and anger management issues (10)
- This is not just an orthopedic problem
  - Multidisciplinary approach including consideration for psychological impact needs to be considered
  - Access to mental health services should be prioritized as part of a holistic approach

Treatment Options

- Historical context

- Early spinal fusion in the past was the treatment of choice (11, 12)
  - A short straight spine was better than a progressing curve

Pulmonary function following early thoracic fusion in non-neuromuscular scoliosis

Lori A Karol [1], Charles Johnston, Kiril Mladenov, Peter Schochet, Patricia Walters, Richard H Browne

- Early spinal fusion may halt progressive deformity, but it does not facilitate lung growth (13)
- Early fusion patients had average 58% of normal FVC
- Extent of fusion had negative correlation with pulmonary function
- More proximal the fusion, the worse the pulmonary outcome

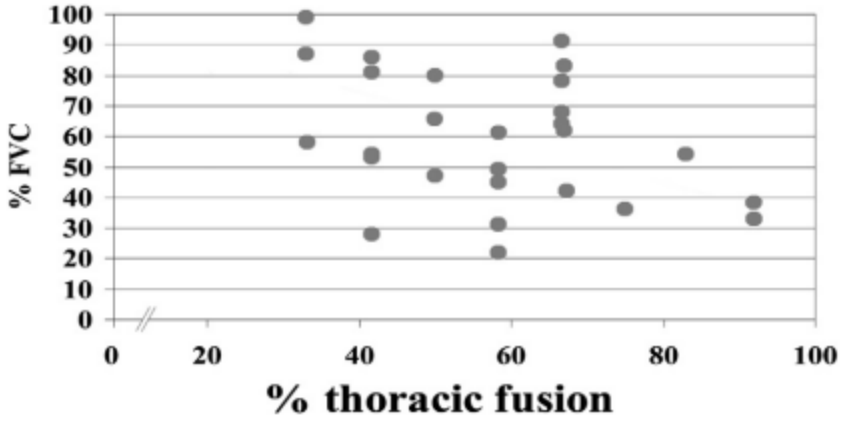
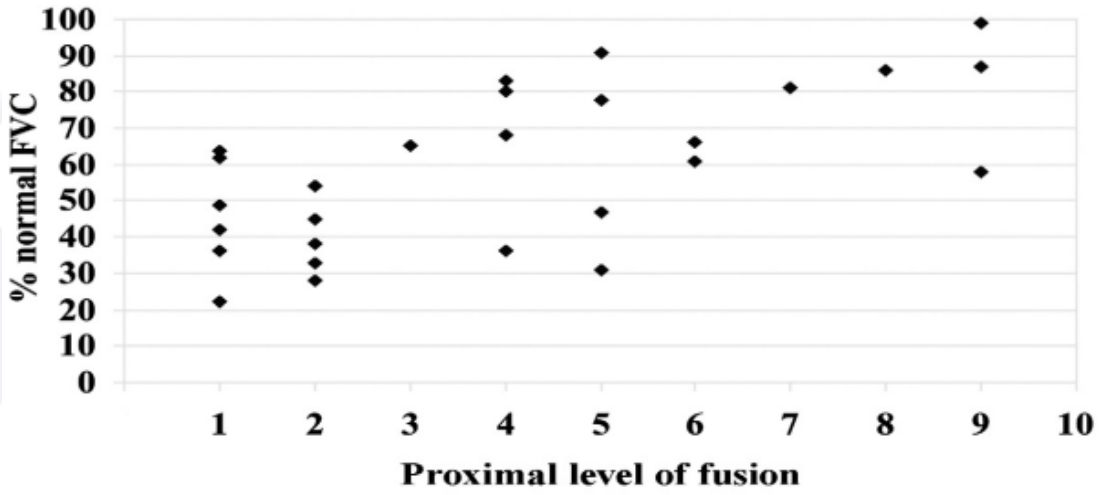


Fig. 1
The percentage of the thoracic spine fused at the index procedure plotted against the percentage of predicted forced vital capacity (FVC) for twenty-eight patients. More extensive thoracic fusions were associated with diminished forced vital capacity at the time of follow-up ($r = -0.46$, $p = 0.01$).



Treatment Options

- Push for growth-friendly treatment options
  - These comprise both surgical and non-surgical techniques
- Non-surgical
  - Casting/bracing
- Surgical
  - Distraction based (TGR, VEPTR, MCGR)
  - Compression based (VBT)
  - Guided growth (Shilla)

“Mehta” Casting

- Casting for spinal curve correction described in 1863 by Bradford (14)
- Technique involves using traction combined with elongation-derotation-flexion casting to provide 3D correctional forces
- Casts then changed every 2-3 months
- Has experienced a resurgence in popularity thanks in part to Dr. Mehta

Growth as a corrective force in the early treatment of progressive infantile scoliosis

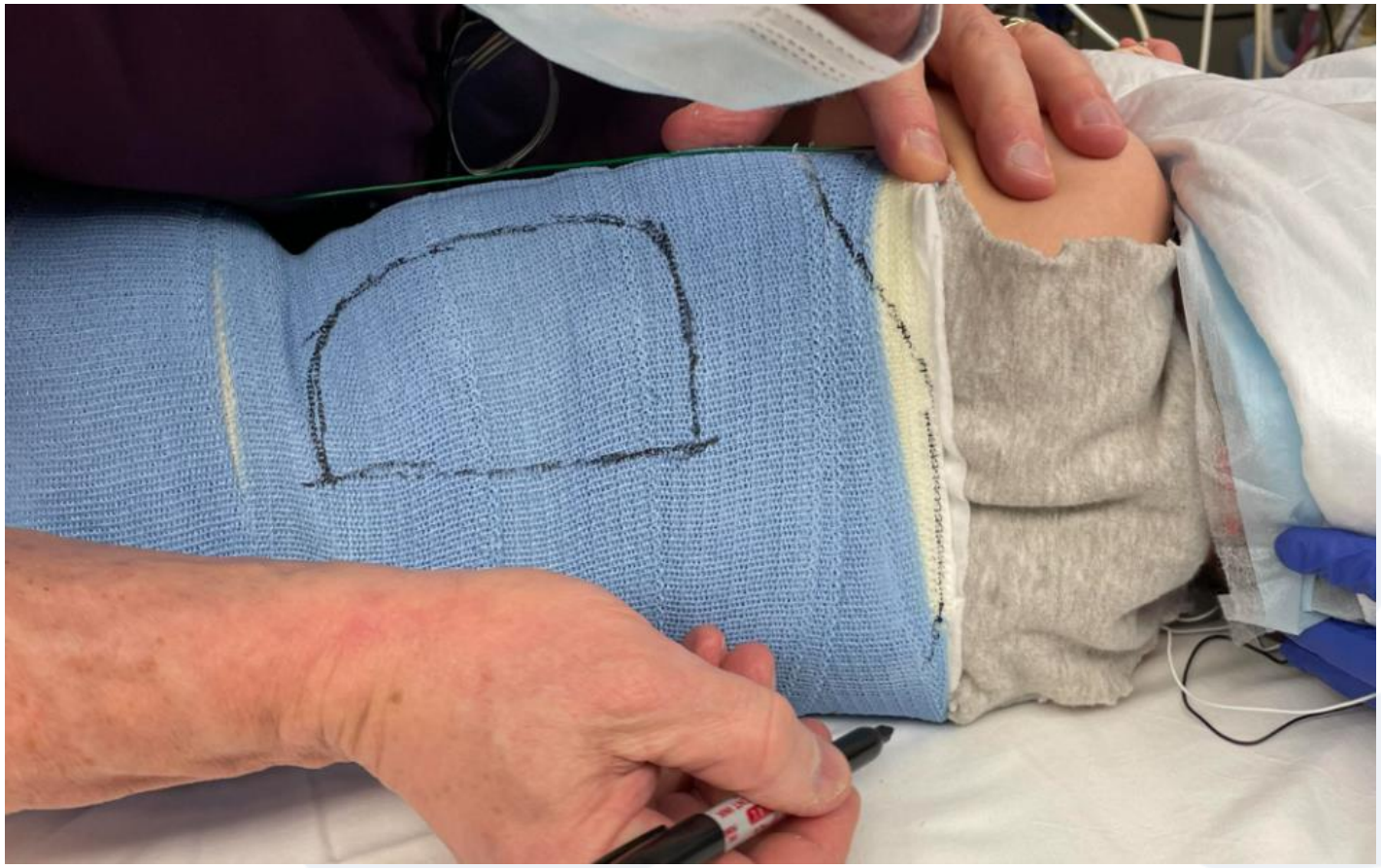
M H Mehta [1]

- Prospective study of 136 children with progressive infantile scoliosis with 9-year follow-up (15)
- Children <2 years, Cobb angle ~30 degrees, casting can provide long standing correction of scoliotic curves
- Children >2 years, Cobb angle ~50 degrees, casting can reduce but not reverse the deformity
  - Majority will go on to spinal fusion, but can delay until more skeletally mature

“Mehta” Casting

“Mehta” Casting

“Mehta” Casting

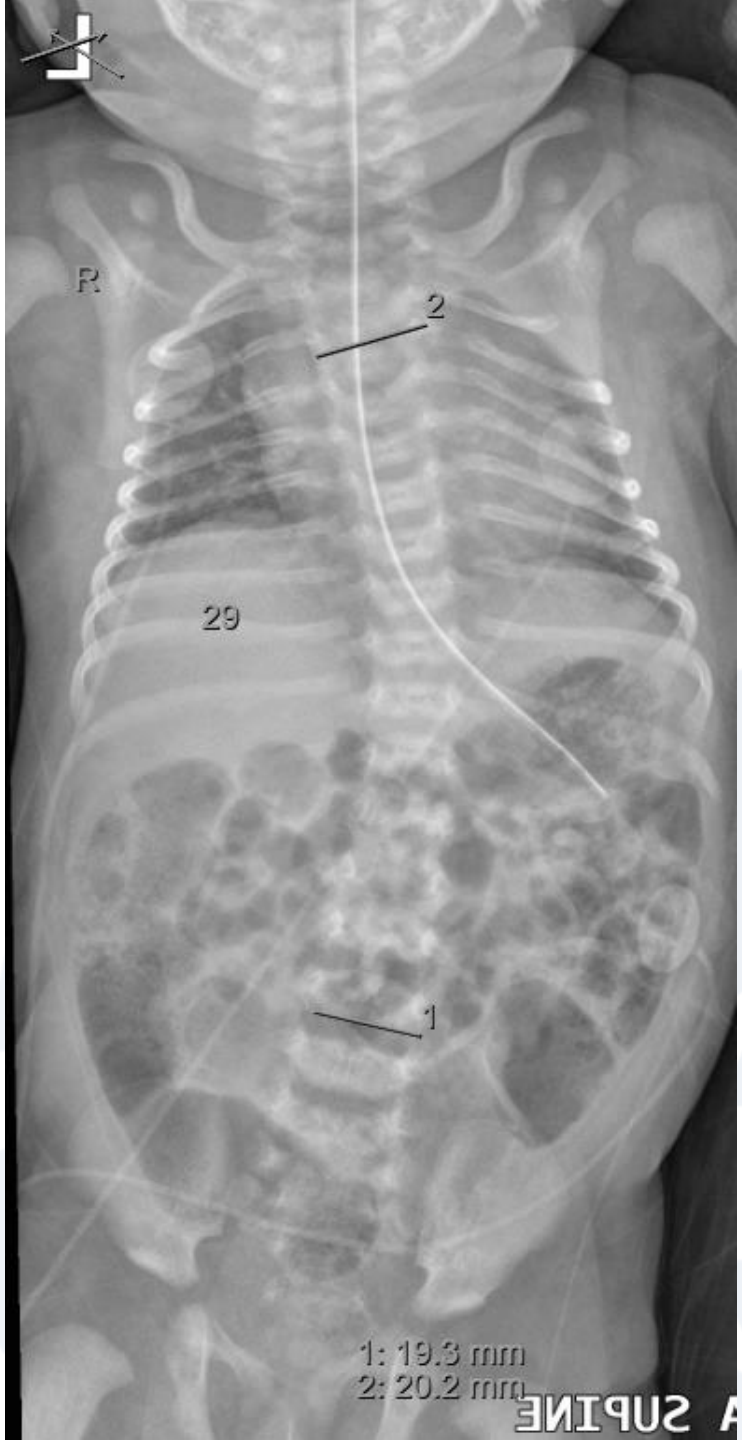


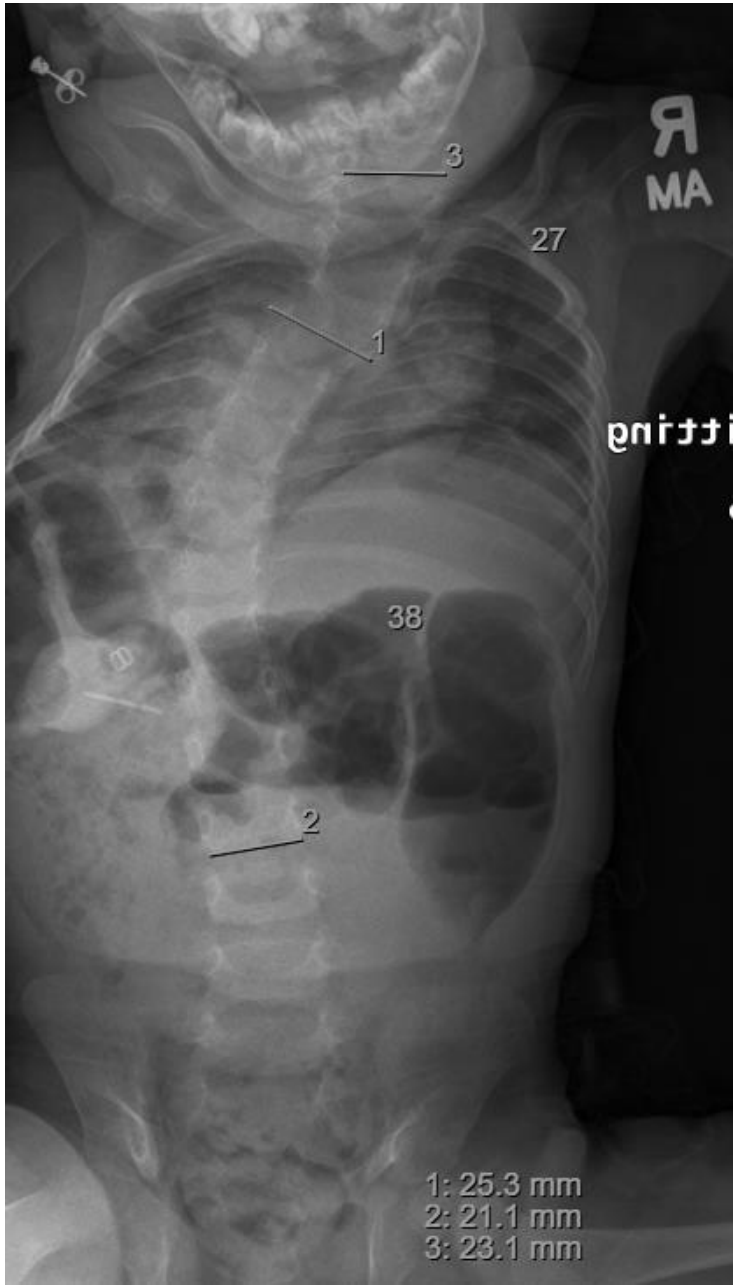
“Mehta” Casting

Case 1

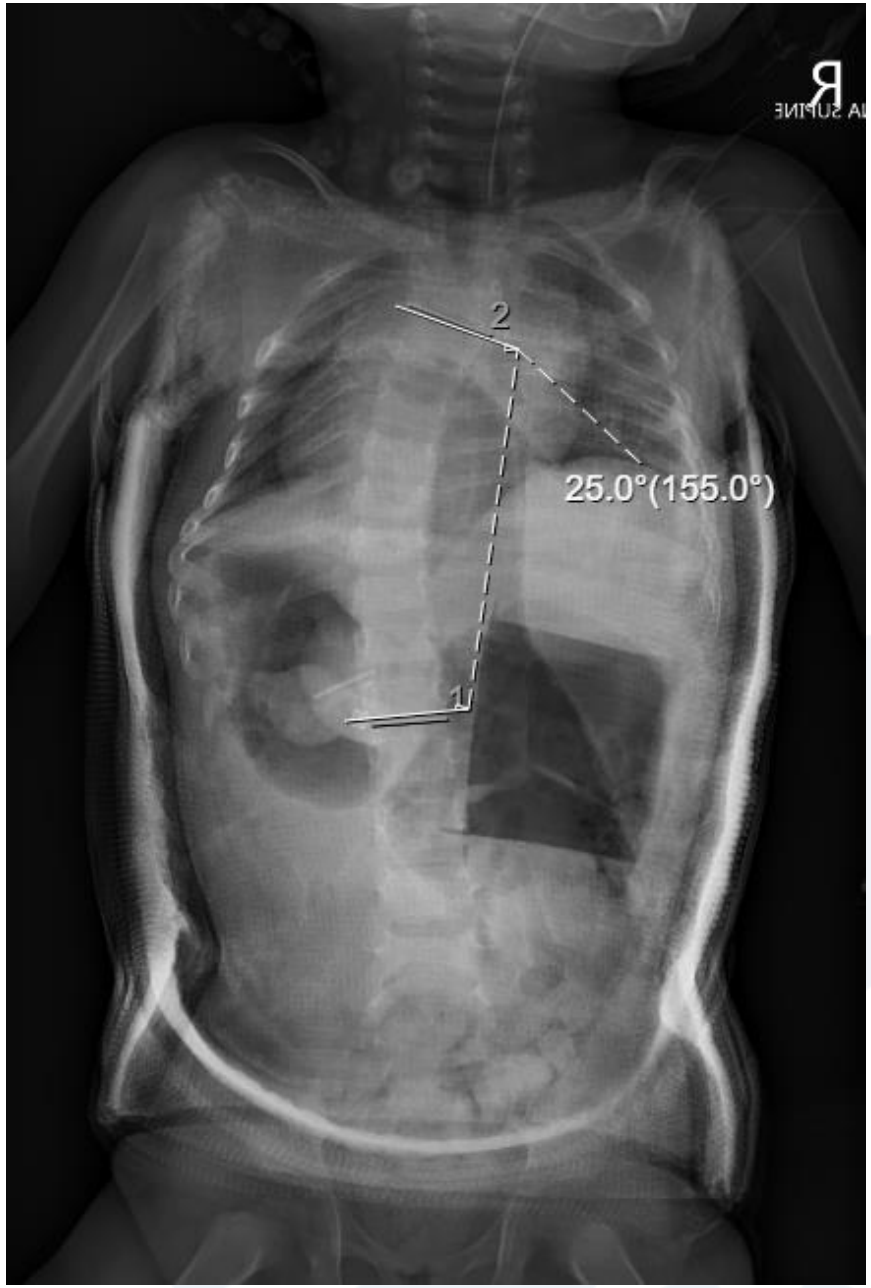
- DH presented to our clinic at 9mo’s of age for spinal asymmetry concern

- Mild developmental delay, g-tube and left renal agenesis

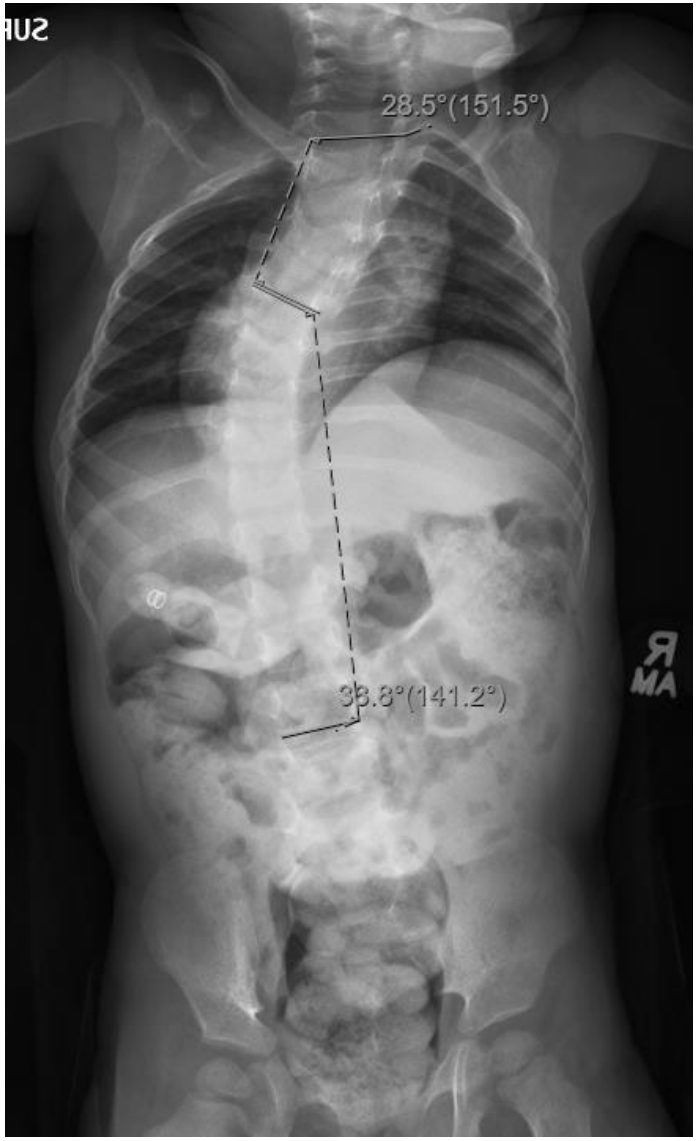




1 year of age



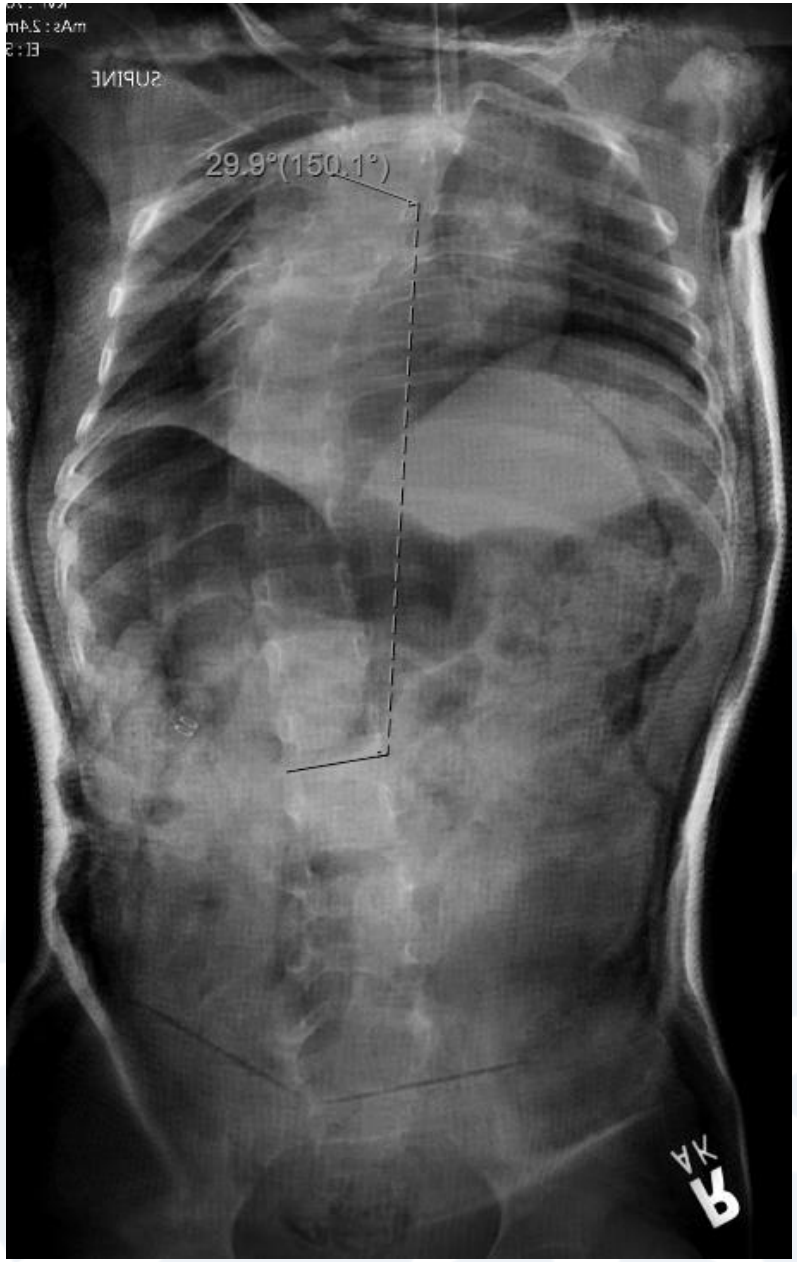
1st Cast



2 years of age, having undergone 4 casts

Cast Holiday, returns at 3 years of age


28.4°(151.6°)
121.9°(58.1°)
47.1°(132.9°)
68.4°(111.6°)


SUPINE
29.9°(150.1°)

Bracing for EOS

- Bracing effectiveness in EOS is under debate (16, 17)
- Provides the benefit of being removed as desired, but lacks continuous corrective force of casting
- Thometz et al 2018 (18)
  - Can provide curve correction in up to 44% of patients
  - 67% of patients had 5 degrees or less or progression
  - Median cobb angle reduction of 15 degrees

Surgical Management

- Distraction-based systems
- Traditional growing rods (TGR) are the most applied technique for treating EOS
- Technique uses anchors as a mechanical distractive force to correct spinal deformity
- Dual-rod technique described by Akbarnia (19) preferred
- Requires repetitive surgical lengthening in the operating room

Complications of growing-rod treatment for early-onset scoliosis: analysis of one hundred and forty patients

- Up to 58% of patients have at least one complication (20)
- Implant failure, surgical site infection, wound problems most common
- Exposure to repeated anesthesia
- Psychological impact of surgical intervention every 6 months

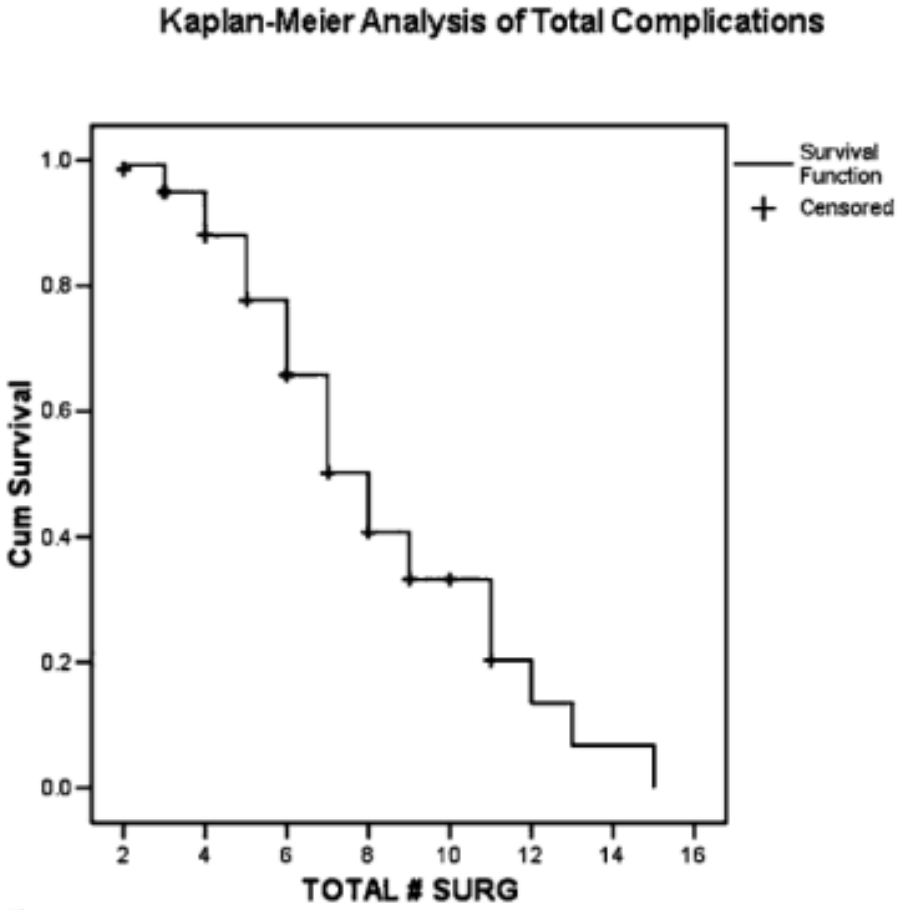
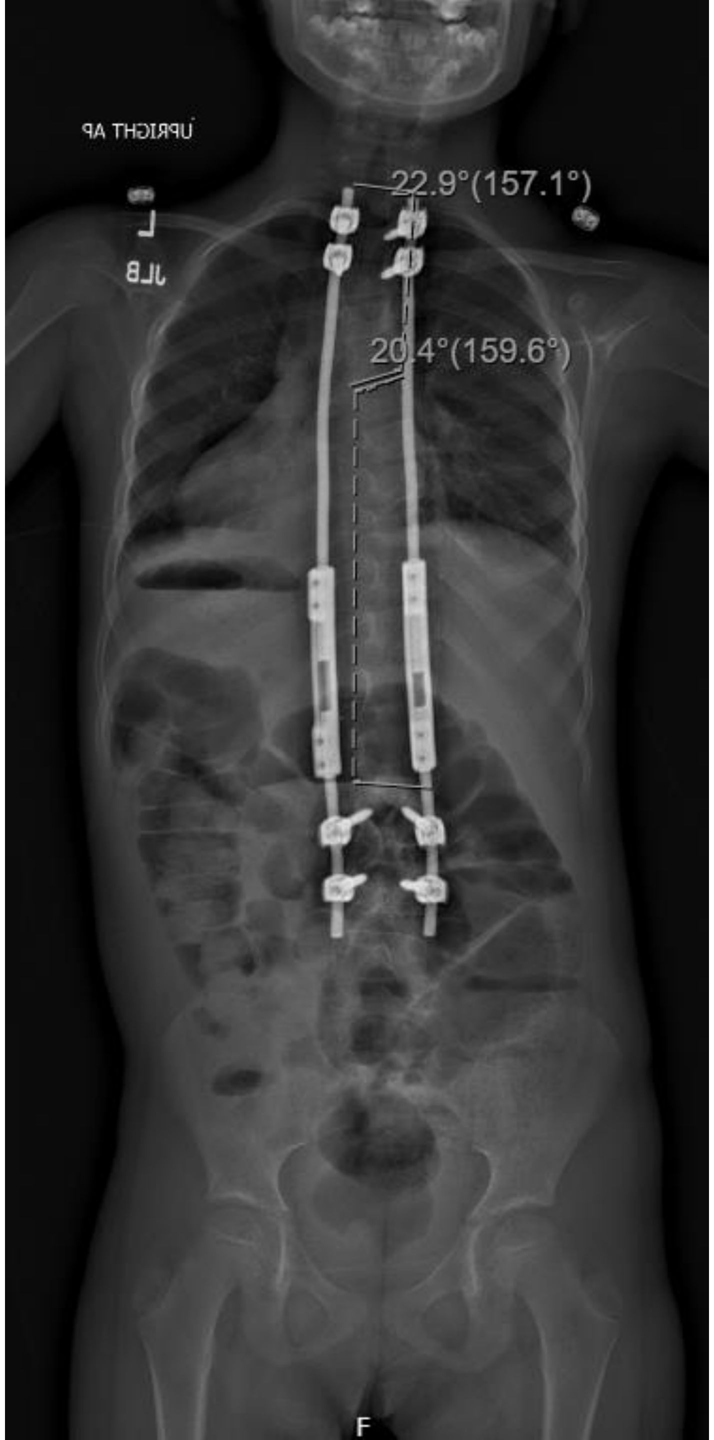


Fig. 2

Kaplan-Meier analysis of total complications showed a linear decrease in complication-free rates for each surgical procedure performed. At six surgical procedures, the complication-free rate was approximately 70%, indicating that 70% of the patients did not have a complication after six surgical procedures. At approximately fifteen surgical procedures, the complication-free rate was zero, indicating that 100% of the patients treated with fifteen surgical procedures had a complication.

Case 2

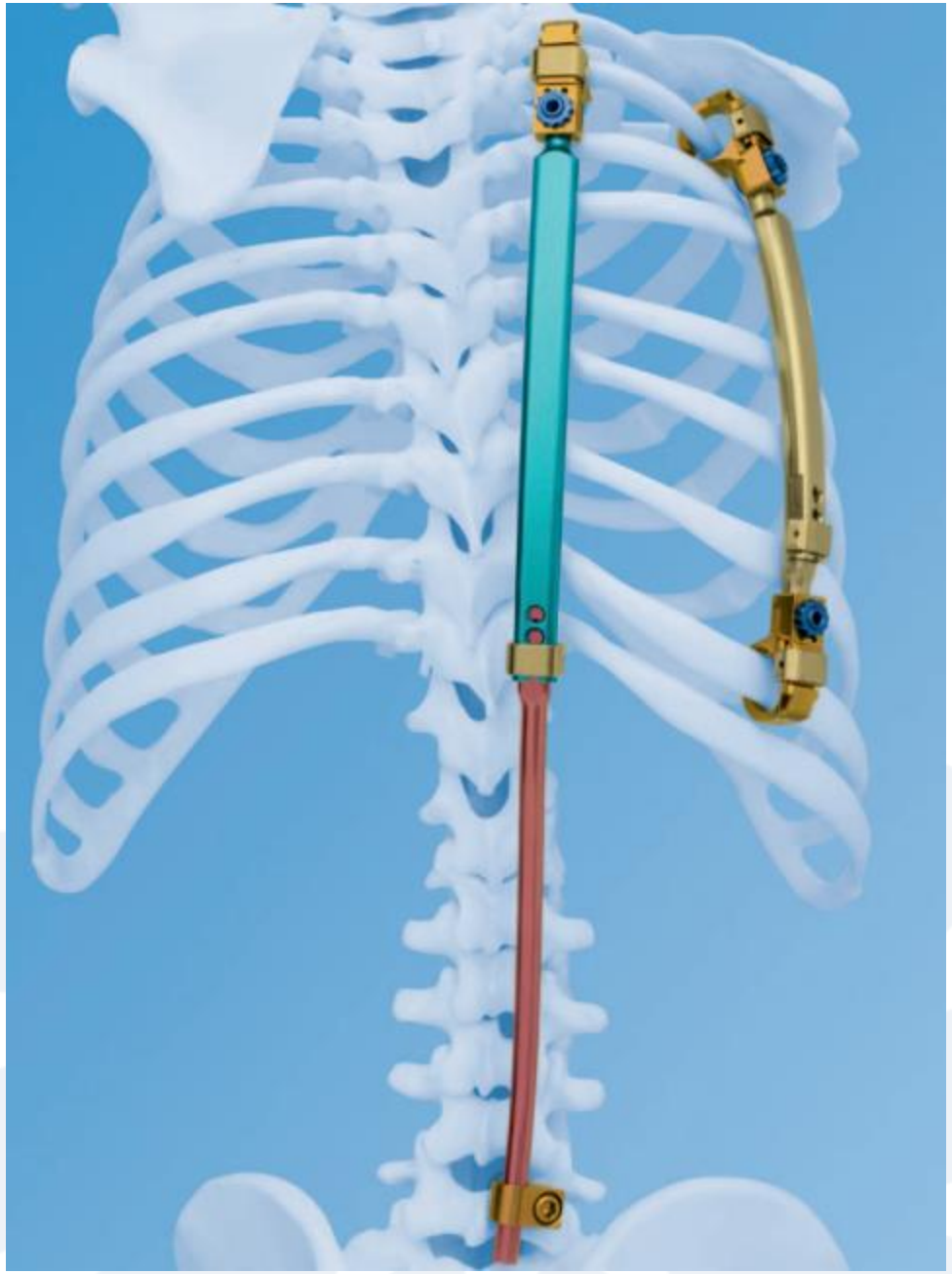
- TG is a 6-year-old Female, hx of psoriasis but otherwise healthy
- Dad noticed shoulder asymmetry which prompted referral
- Normal neurologic exam and normal MRI


22.9°(157.1°)
20.4°(159.6°)
F


F

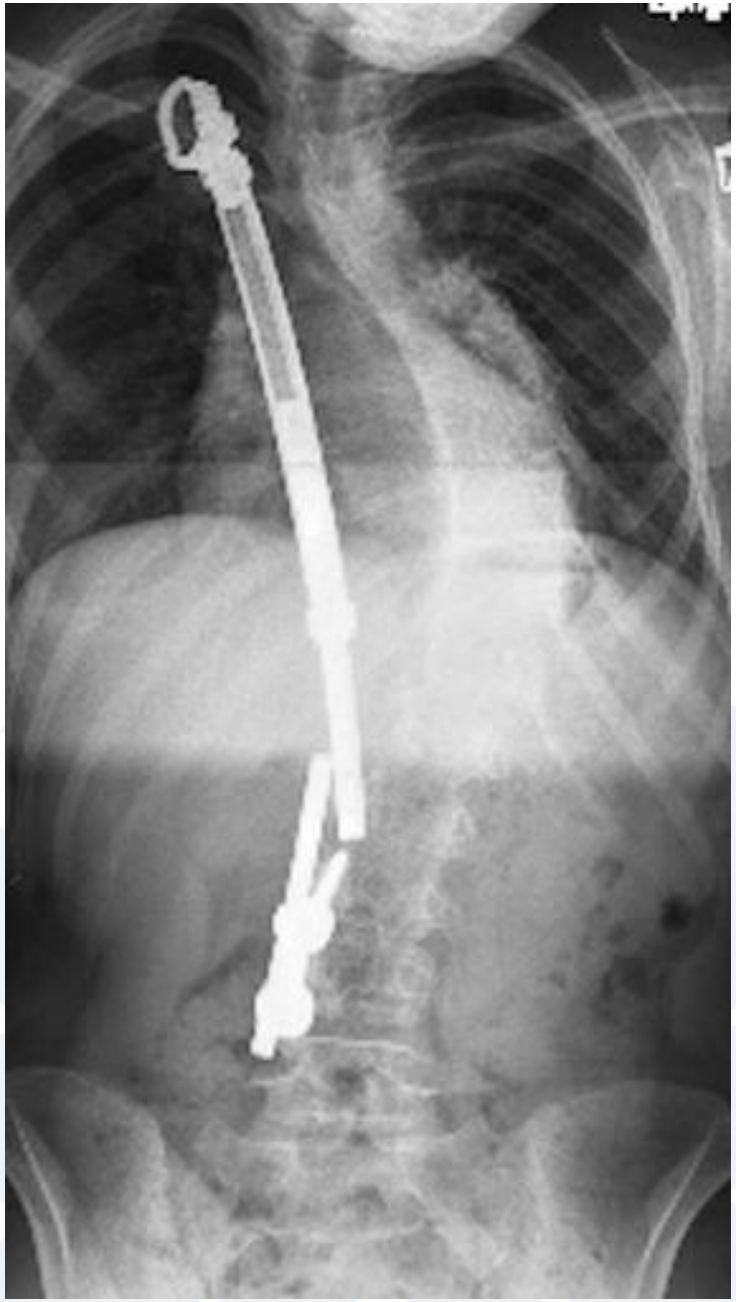
Surgical Management

- Distraction-based systems
- Vertical expandable prosthetic titanium rib (VEPTR)
- First described by Campbell for treatment of thoracic insufficiency syndrome (20)
- Can use rib-rib, rib-spine, rib-pelvis anchor points
- Early promise, but disappointing late results



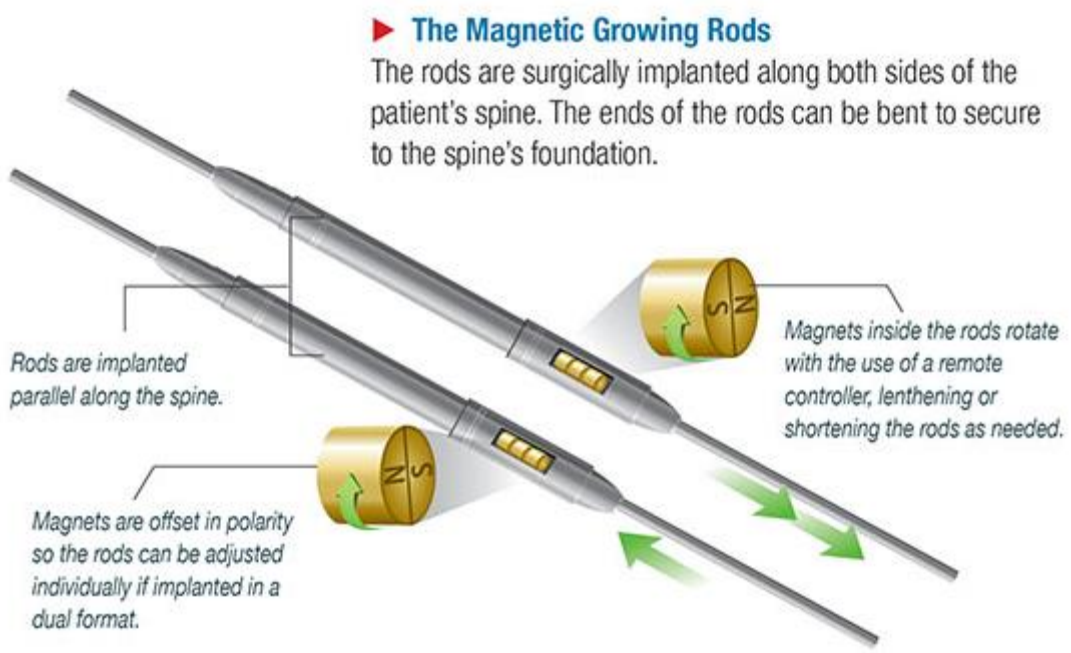
Surgical Management

- Articles by Campbell (22), Hasler (23) and Ramirez (24)
  - Inadequate correction of Cobb angle
  - Up to 100% complication rate
  - Spine growth was moderate
  - Respiratory function did not improve
  - Failure of proximal anchors and auto-fusion most common complications



Surgical Management

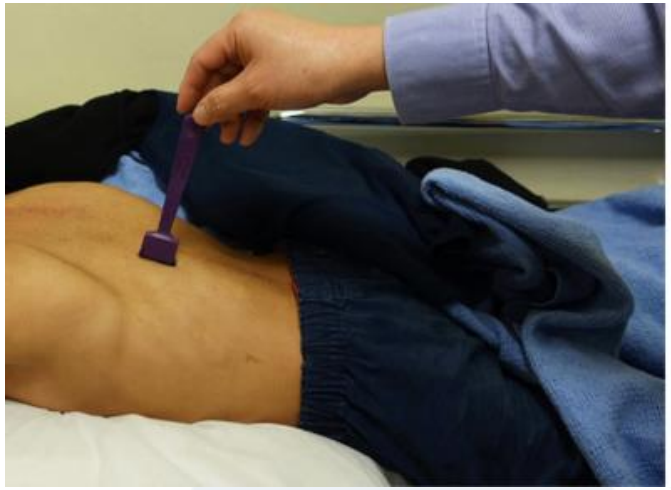
- Distraction-based systems
- Magnetically controlled growing rods (MCGR)
- Developed to offset the need for repeated surgical lengthening as seen with TGRs



Systematic review of the complications associated with magnetically controlled growing rods for the treatment of early onset scoliosis

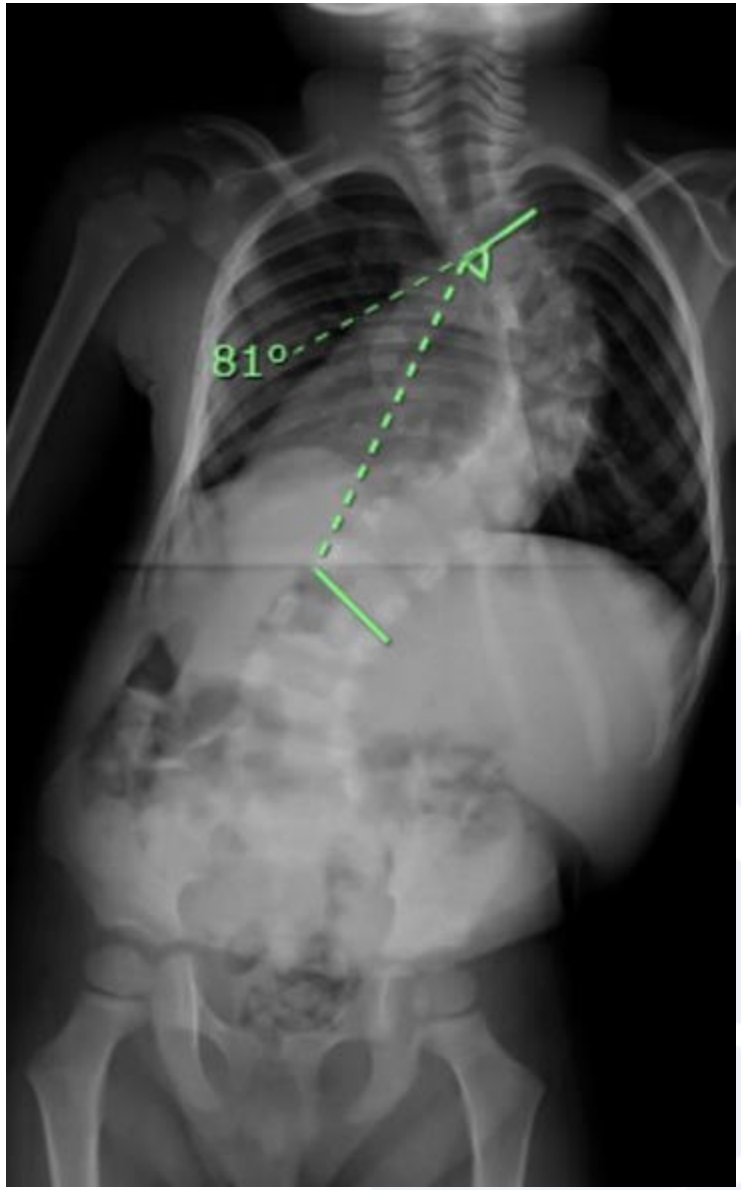
Chrishan Thakar [1], David Christopher Kieser [2 3], Mihai Mardare [1], Shahnawaz Haleem [1], Jeremy Fairbank [1], Colin Nnadi [1]

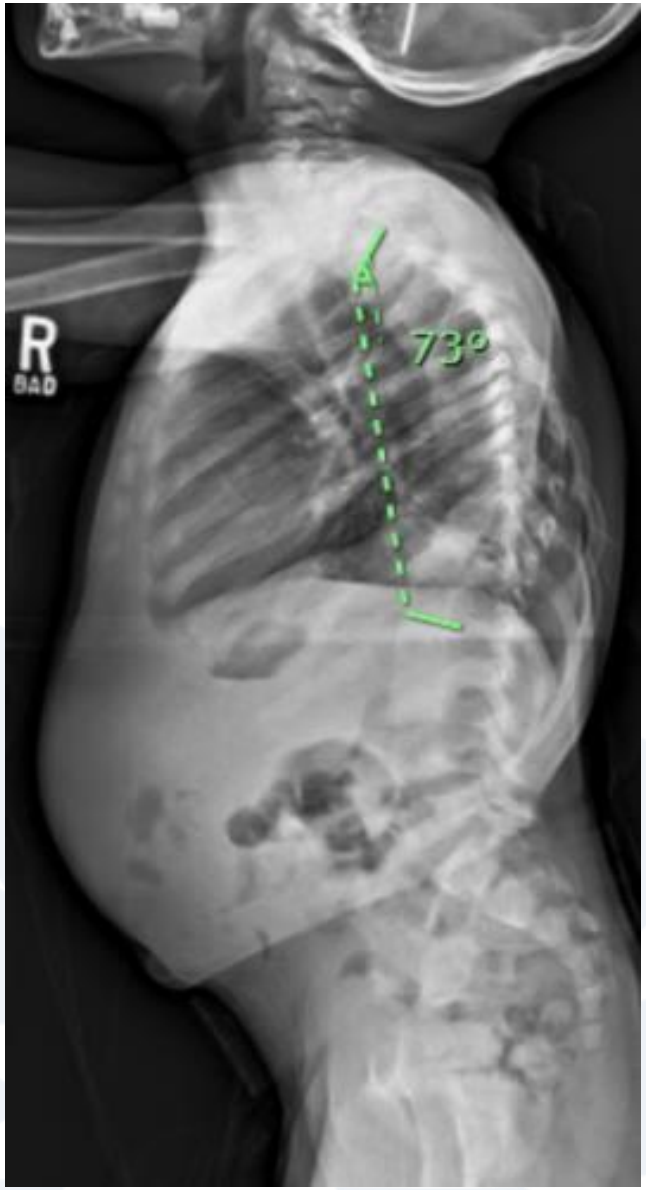
- MCGRs are able to obtain good curve correction
- 45% complication rate with 33% unplanned revision rate (25)
- 4.67 times the risk of problems with metallosis compared to TGR
- No improvements in mental health compared to TGR (26)



Case 3

- EM is a 10-year-old male with chromosome 8 abnormality and syndromic scoliosis
- Underwent serial mehta casting (7 casts) but failure to control his curvature



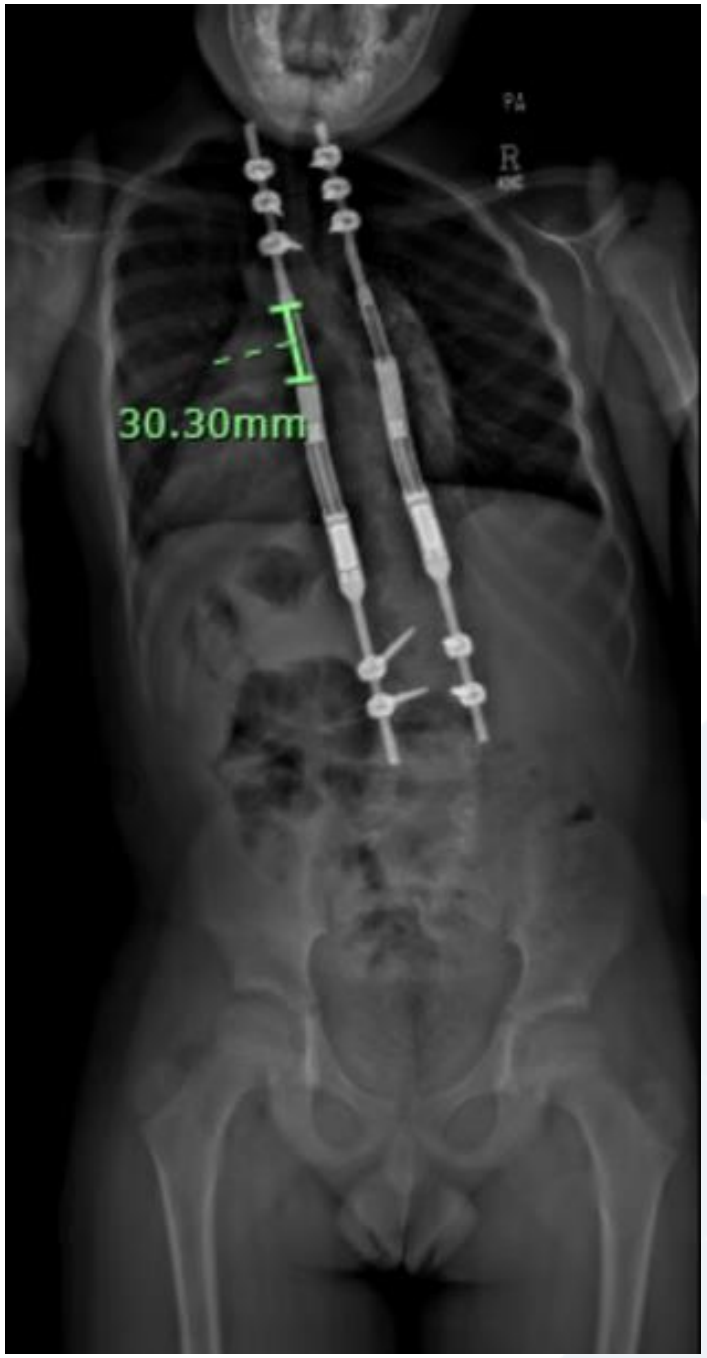


Case 3

- MCGRs placed
- Initially good correction of his curve
- Lengthened 3cm over 2 years

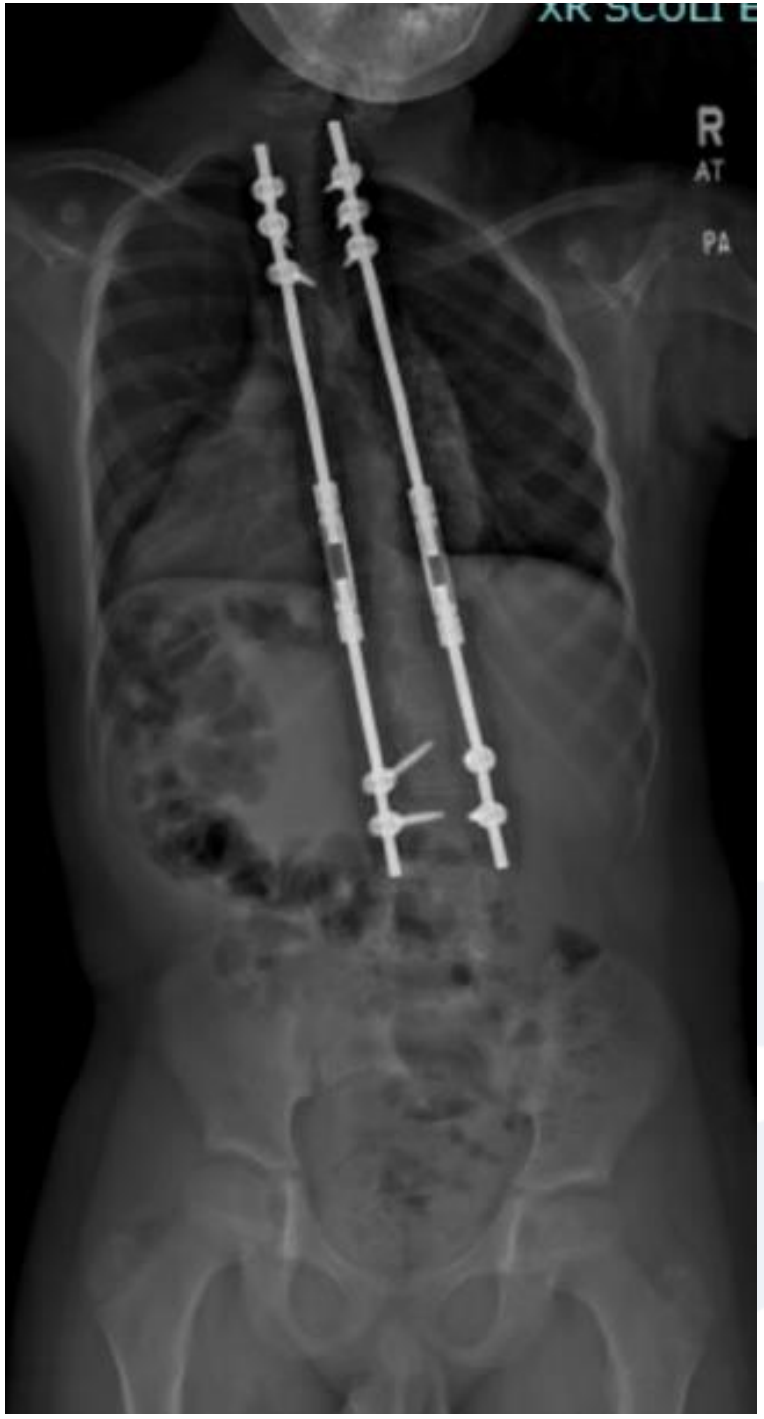
Case 3

- Rods stopped lengthening
- Developing PJK

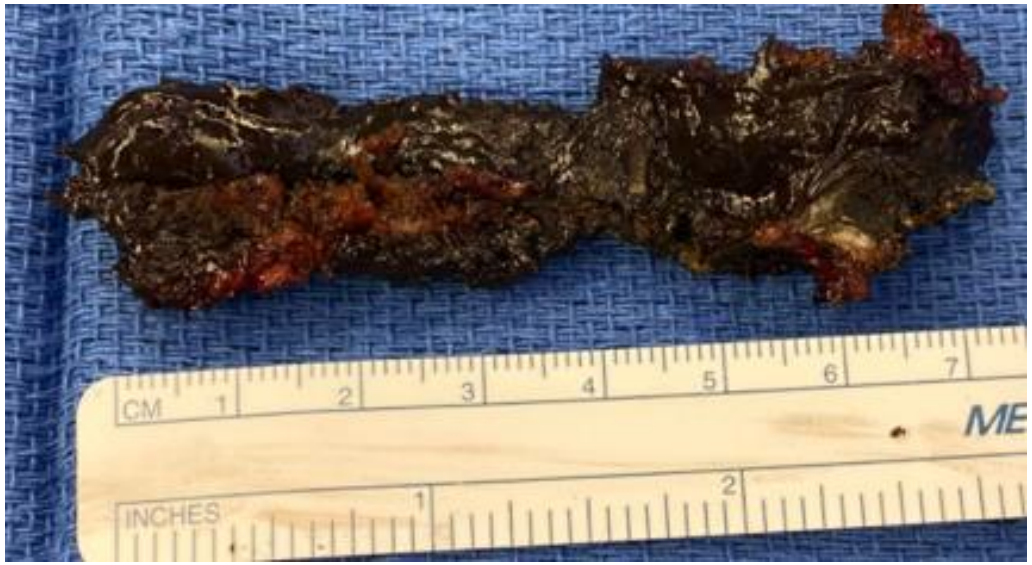


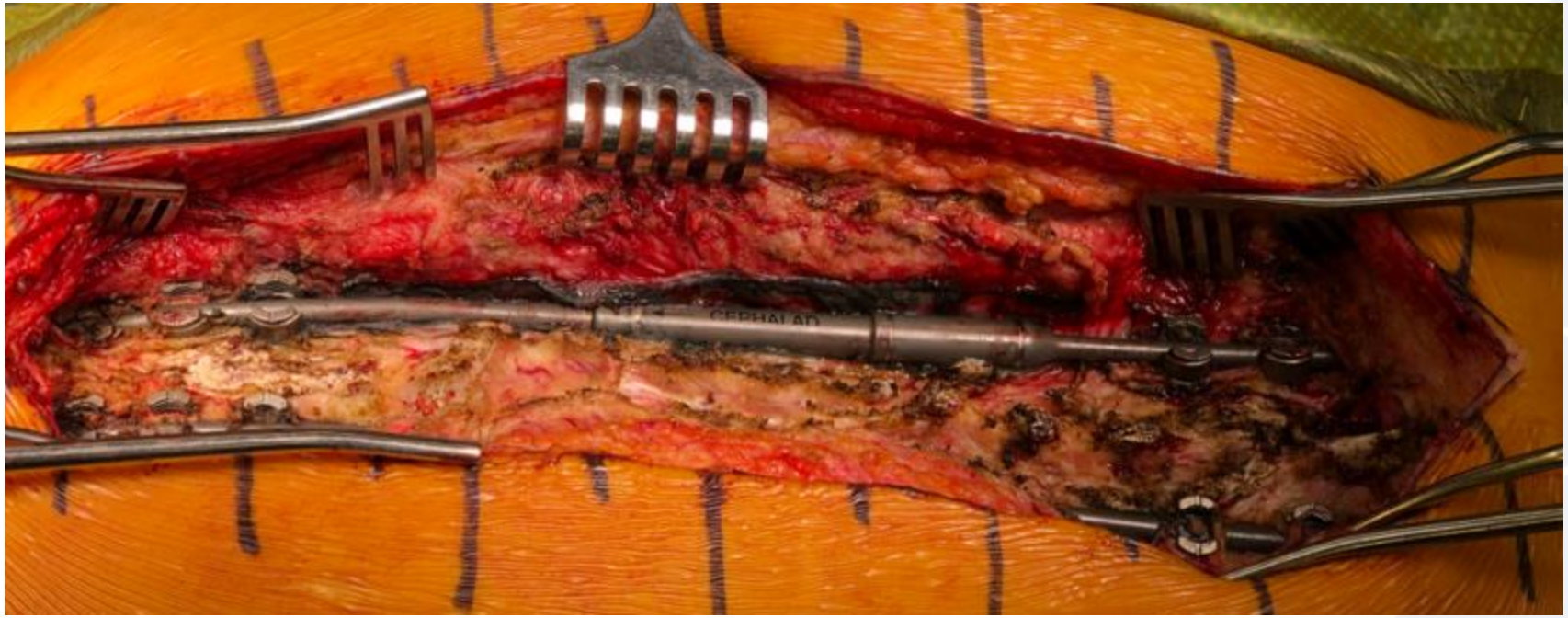
Case 3

Case 3



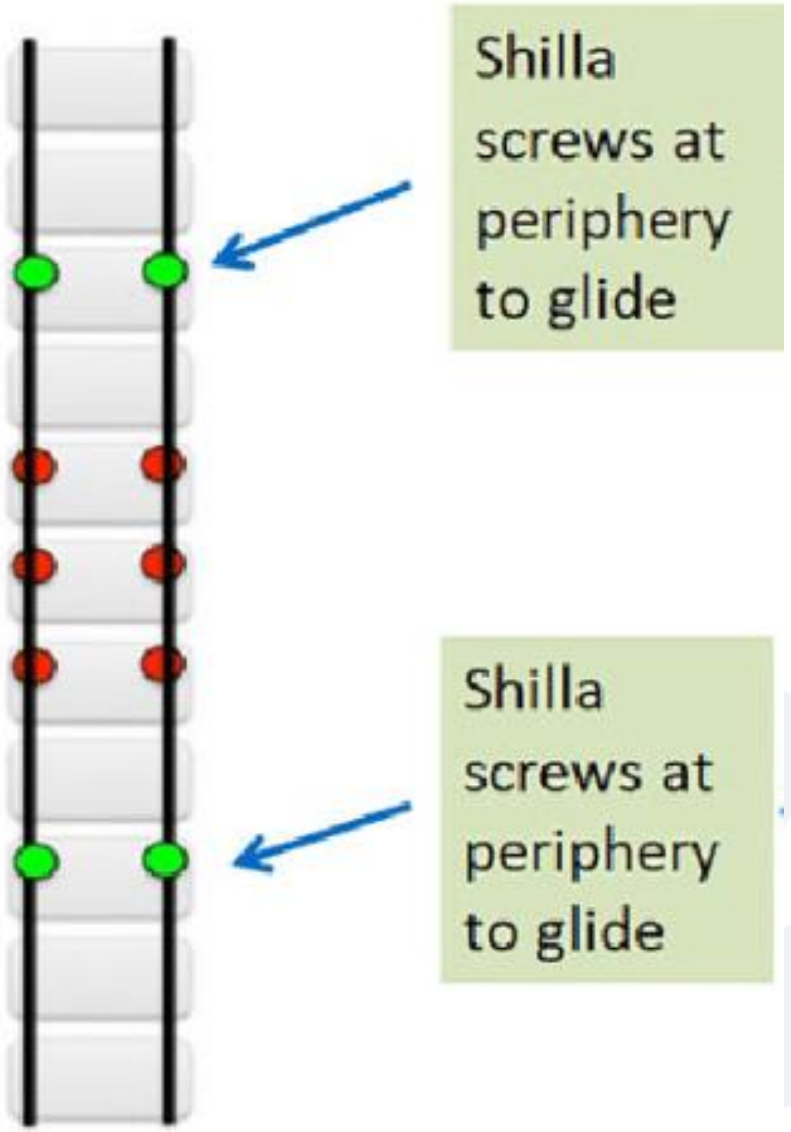
Case 3





Surgical Management

- Guided-Growth systems
- Shilla technique described by McCarthy in 2014 (27)
  - Apical fusion
  - Percutaneous fixation top and bottom
  - Allows spine to grow along the rods
- Has theoretical benefit of avoiding repeated surgical intervention



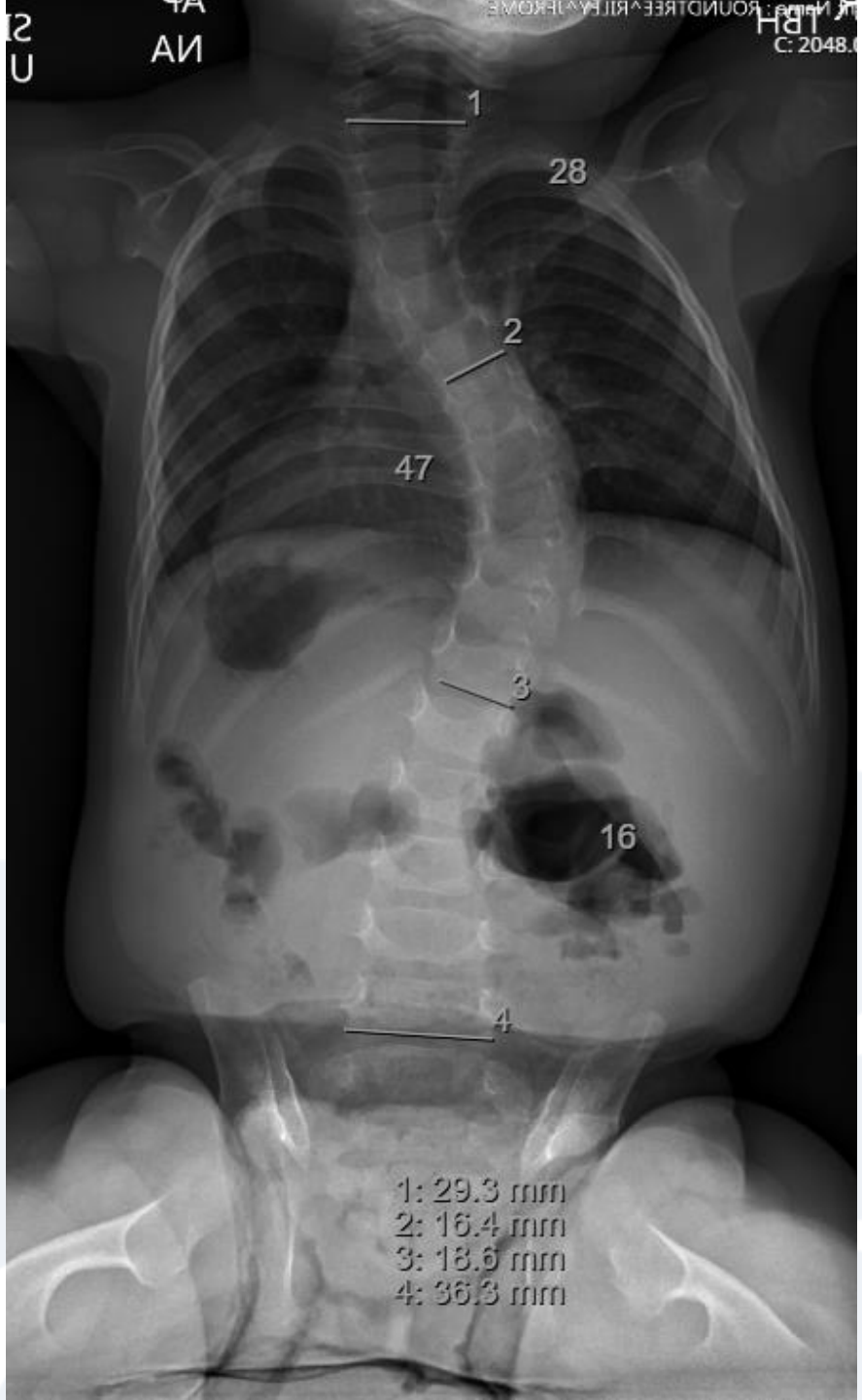
Radiographic Outcomes of Shilla Growth Guidance System and Traditional Growing Rods Through Definitive Treatment

Scott J Luhmann [1], June C Smith [2], Ann McClung [3], Frances L McCullough [4], Richard E McCarthy [4], George H Thompson [5]; Growing Spine Study Group

- No difference in amount of correction obtained (28)
- 4.6cm trunk growth for Shilla compared to 5.2cm for TGR
- Complication rates equivalent
- Threefold decrease in number of surgeries with Shilla compared to TGR
- But….with more time complications increase
  - McCarthy et al. 2015: 5 years follow-up
  - 73% complications: wound infections, alignment concerns, implant problems

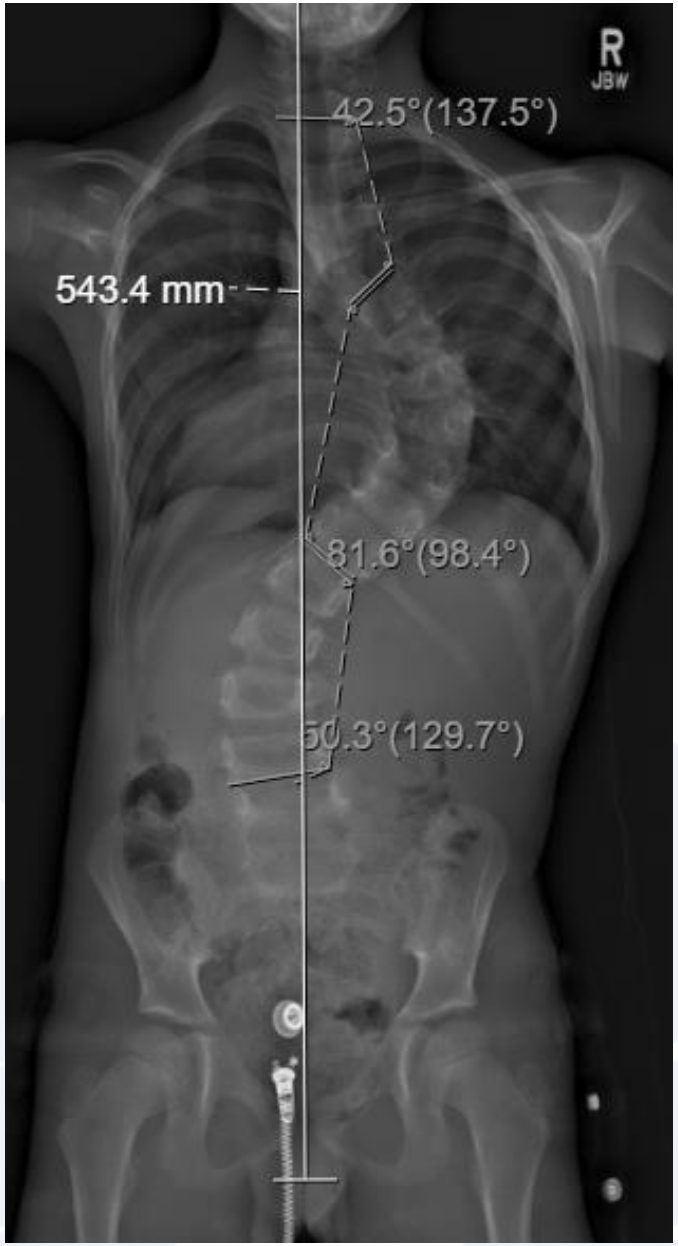
Case 4

- RR is an 8-year-old male with EDS and hx of tethered cord
- Presented to us at 1 year of age
- Underwent serial mehta casting and bracing until age 5



Case 4

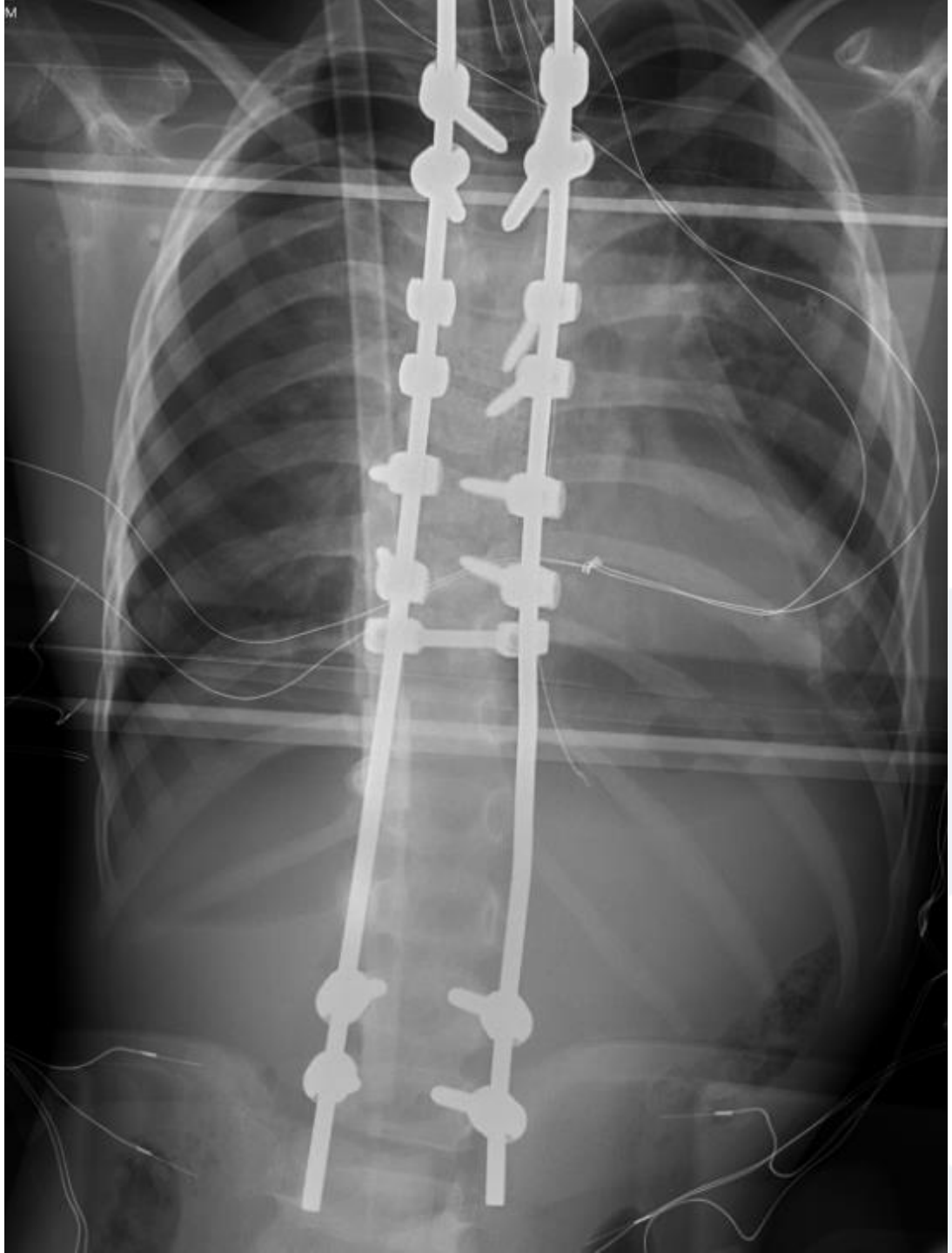
- At age 5



- Lost to follow-up for > 2 years

Case 4

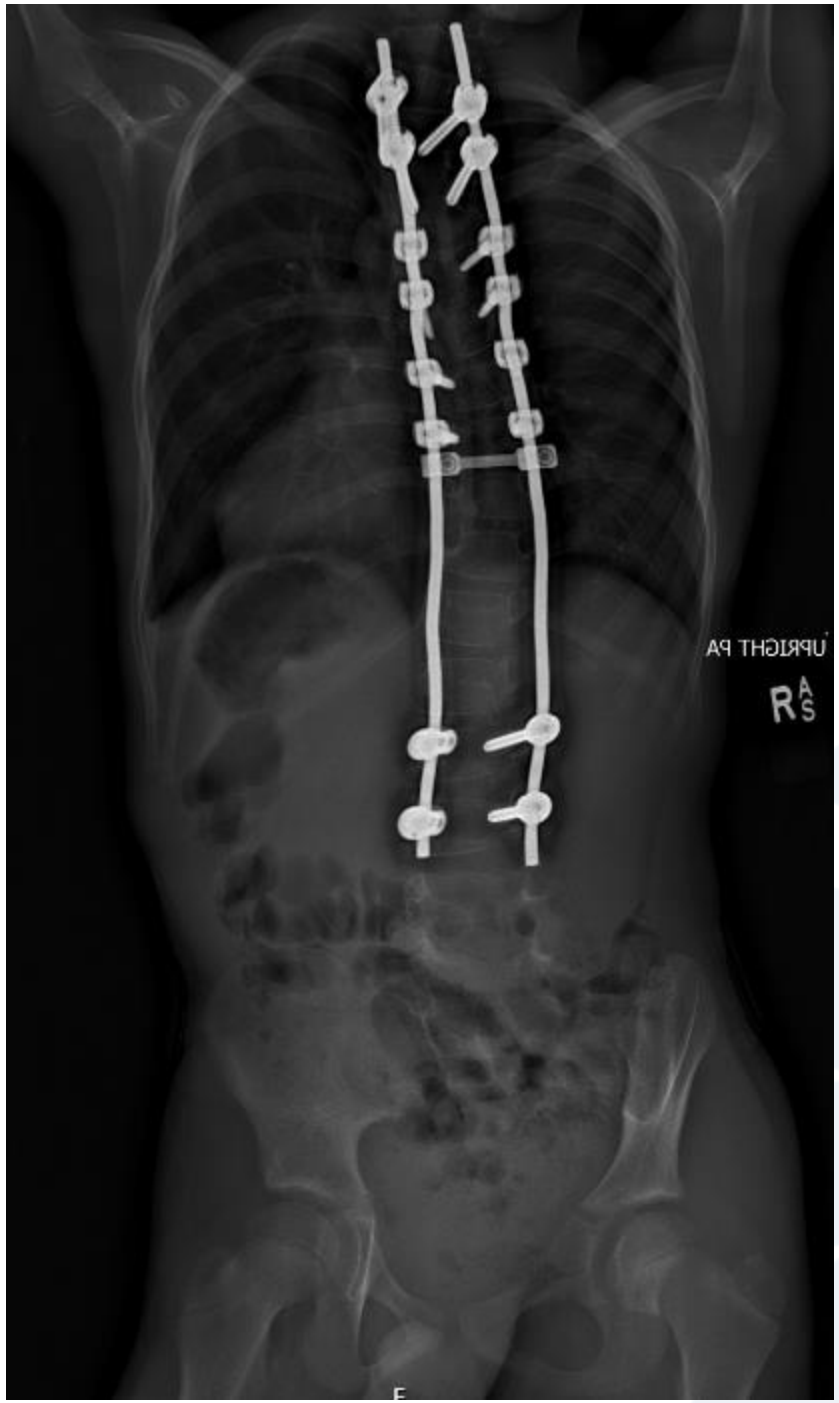
Case 4



Case 4

Case 4

- 6 months postop

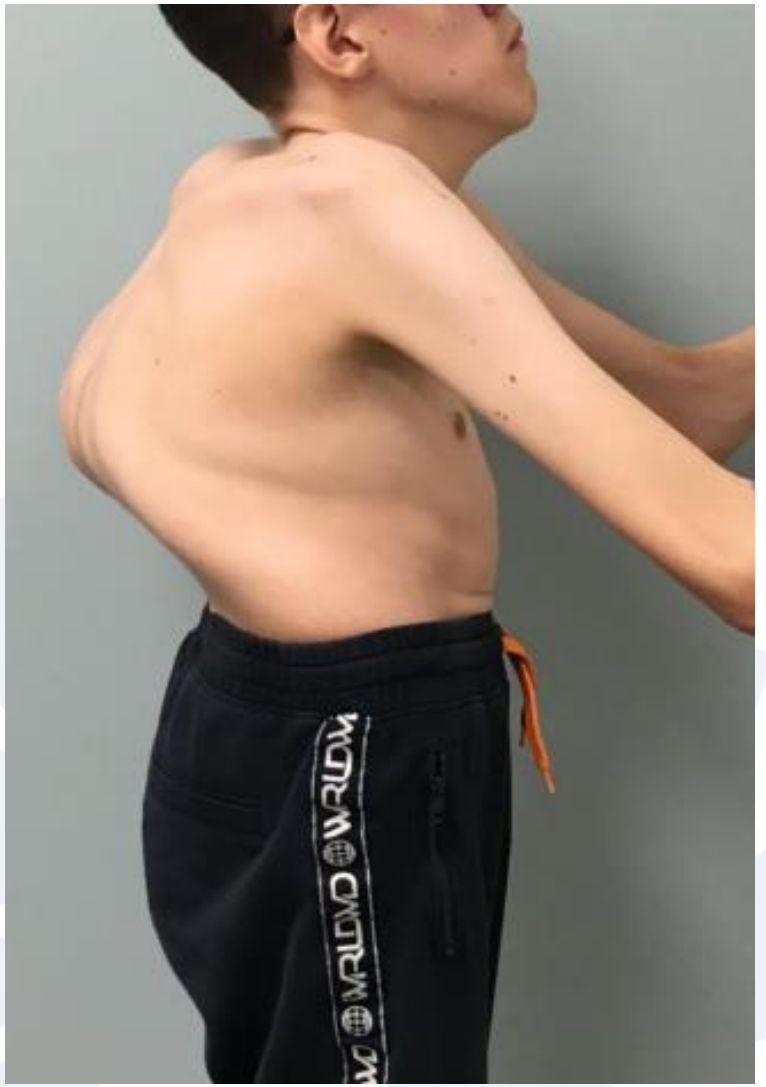


Summary

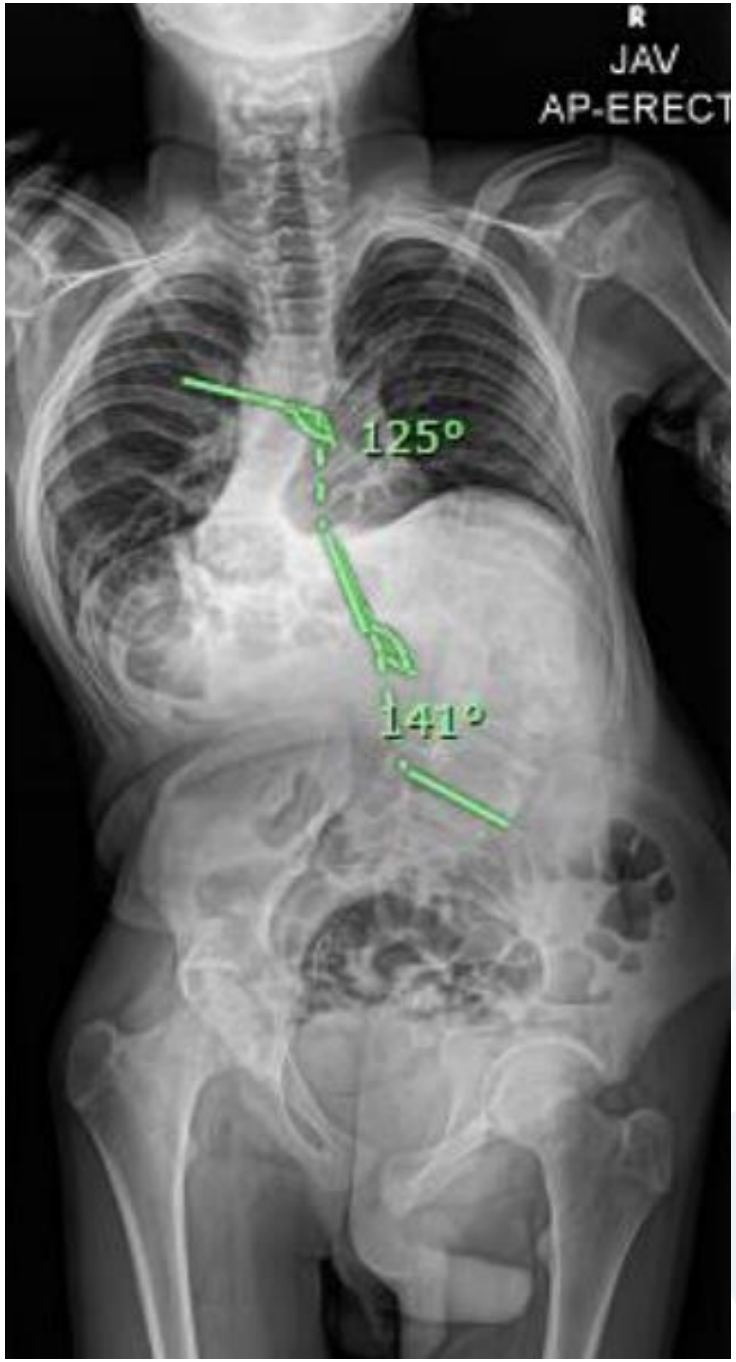
- EOS is a set of complex pathologies related to the growing spine
- It is a very distinct and more severe entity compared to AIS with increased mortality and psychological burden
- Numerous treatment modalities exist, all are fraught with high complication rates
- No gold standard treatment exists
  - Likely worst thing you can do is allow to progress unchecked

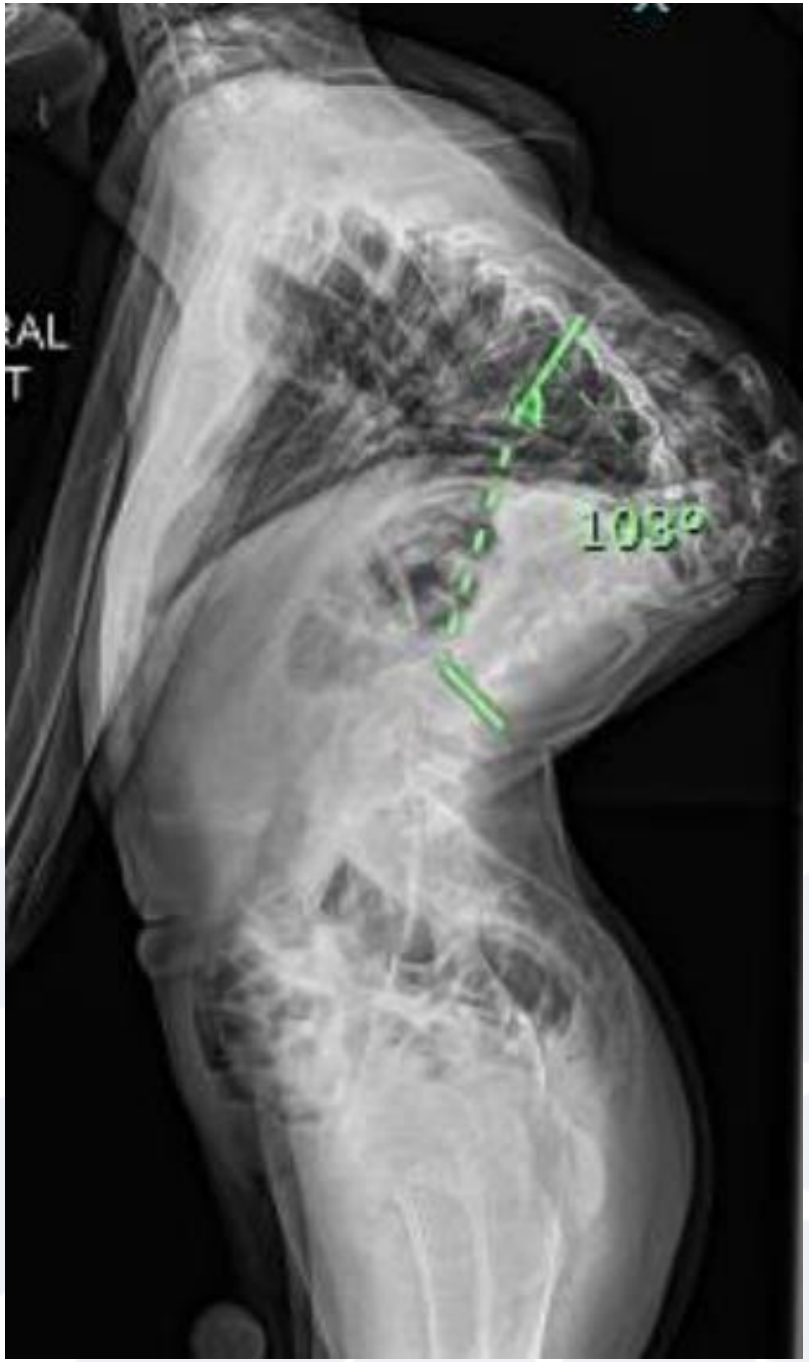
Case 5

- 17m from Poland with undiagnosed syndrome
- Dx with kyphoscoliosis at early age
- Underwent casting. Prior surgery “aborted” due to spine being too stiff.



Case 5

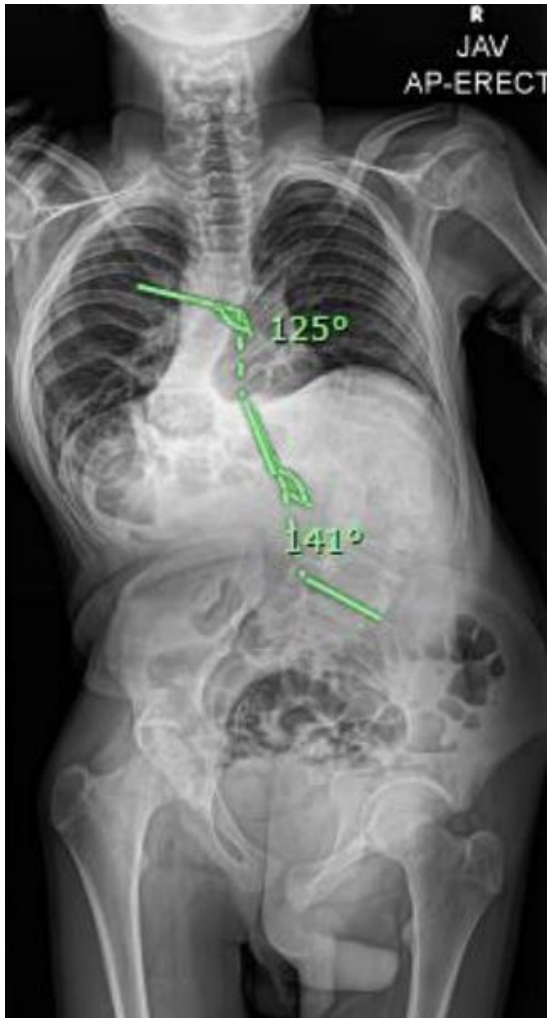


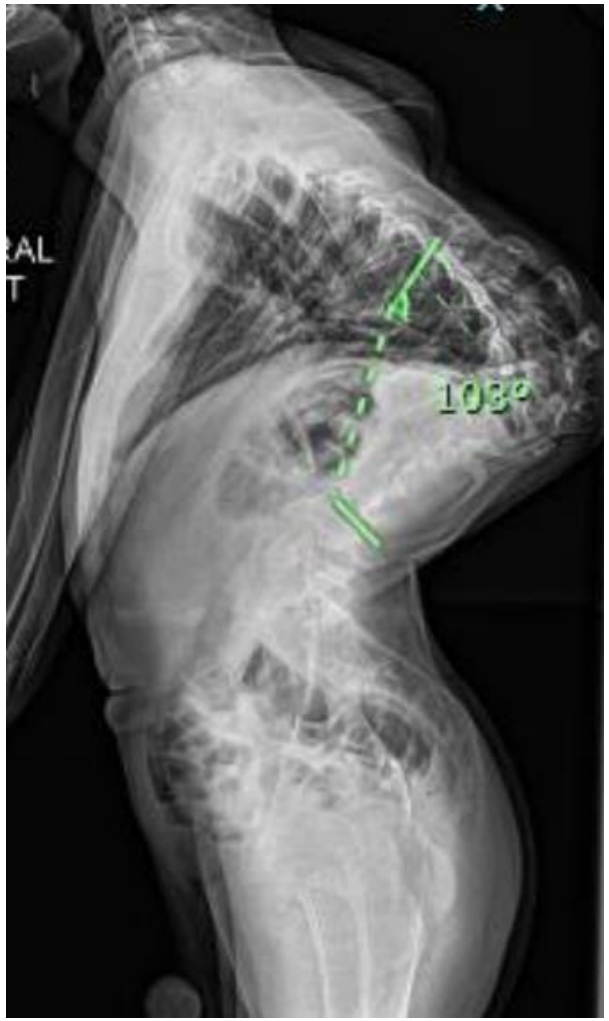


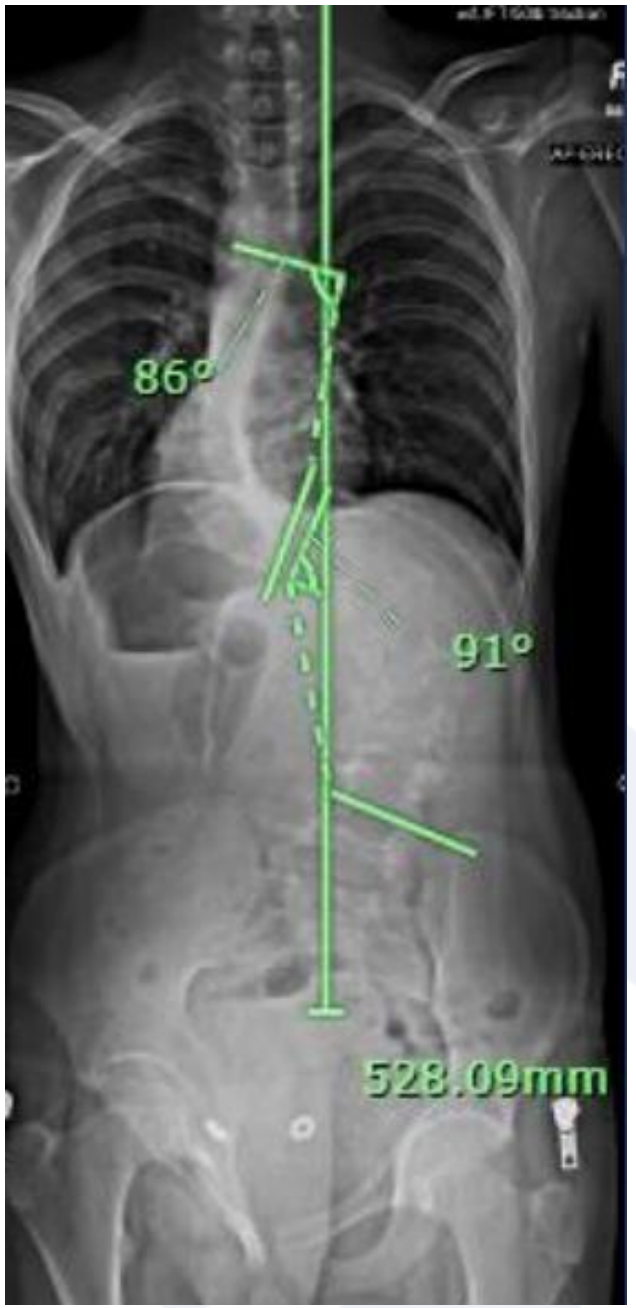
Case 5

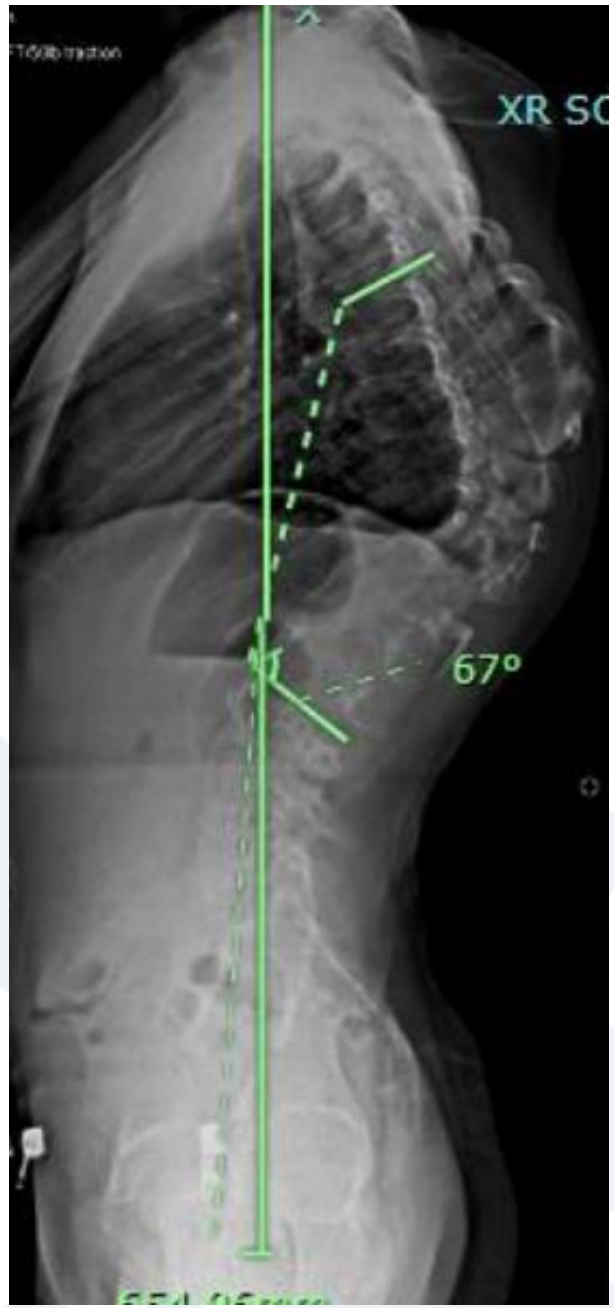
- Underwent 3 stage procedure
- Halo application and HGT
- Posterior column osteotomies, convex rib resections, anterior discectomies through costotransversectomy approach
- PSF T3-L4, concave rib osteotomies

Case 5

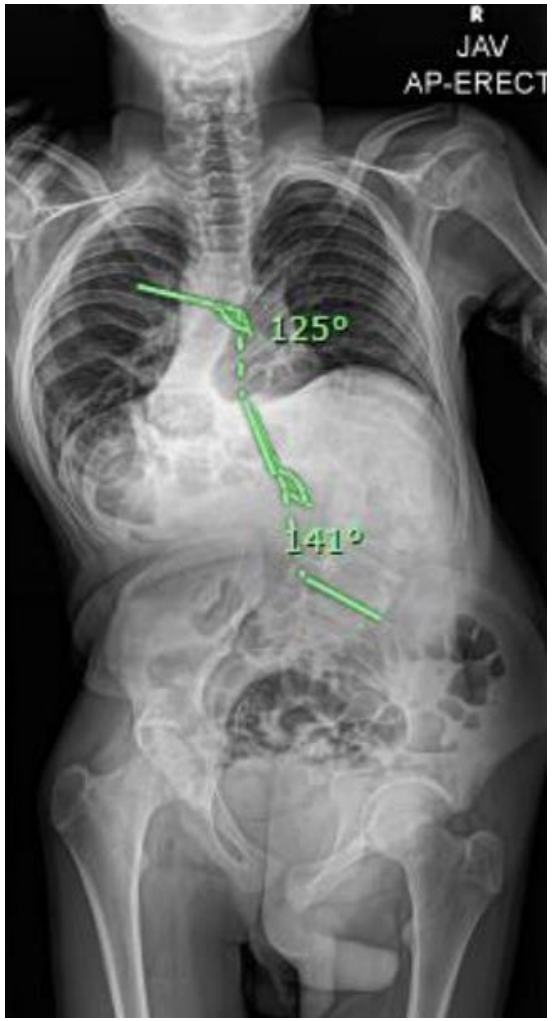


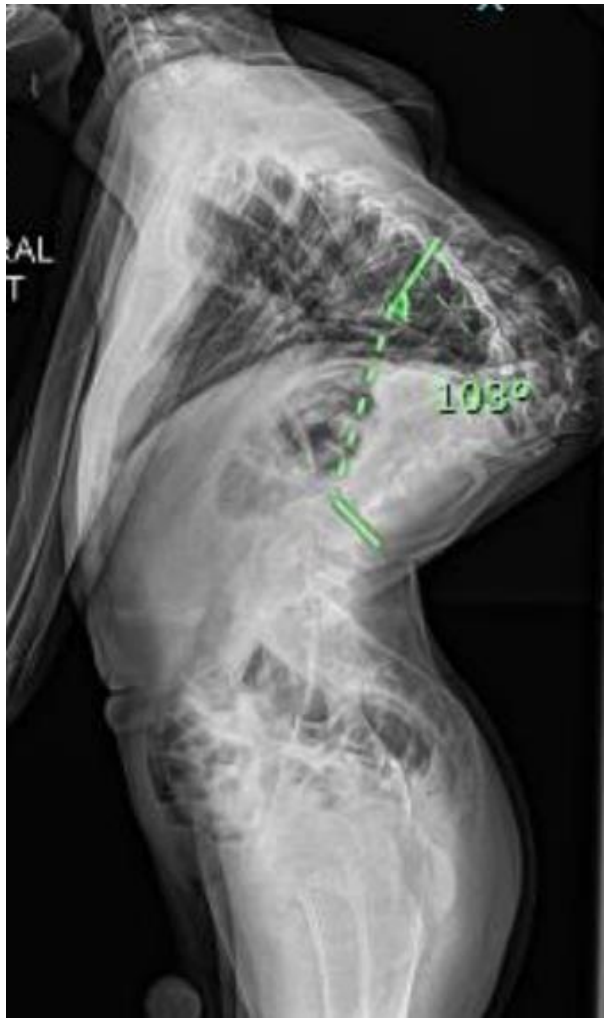






Case 5





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Thank You!