Early Onset Spinal Deformity: Growing Rods or VEPTR – How to decide?

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Disclosures:

- *Conflicts of Interest*:
  - Helped design VEPTR II
    - Royalties from VEPTR II
  - Consultant:
    - Synthes spine
    - Medtronics spine
- *Off-Label devices discussed:*
  - All pedicle screws in children and growing rods (and staples and tethers) are off-label
  - VEPTR is FDA approved for Thoracic Insufficiency syndrome in growing patients. HDE approval is required

Growing Rods or VEPTR:

- Are they really very different?
  - Both distraction based:
    - Skaggs classification useful:
      - Rib-based distraction
      - Spine-based distraction
      - Combinations
  - Both have same major disadvantage:
    - Repetitive surgical lengthening required
    - Distraction-bases Rx not very good for kyphosis

Growing Rods or VEPTR

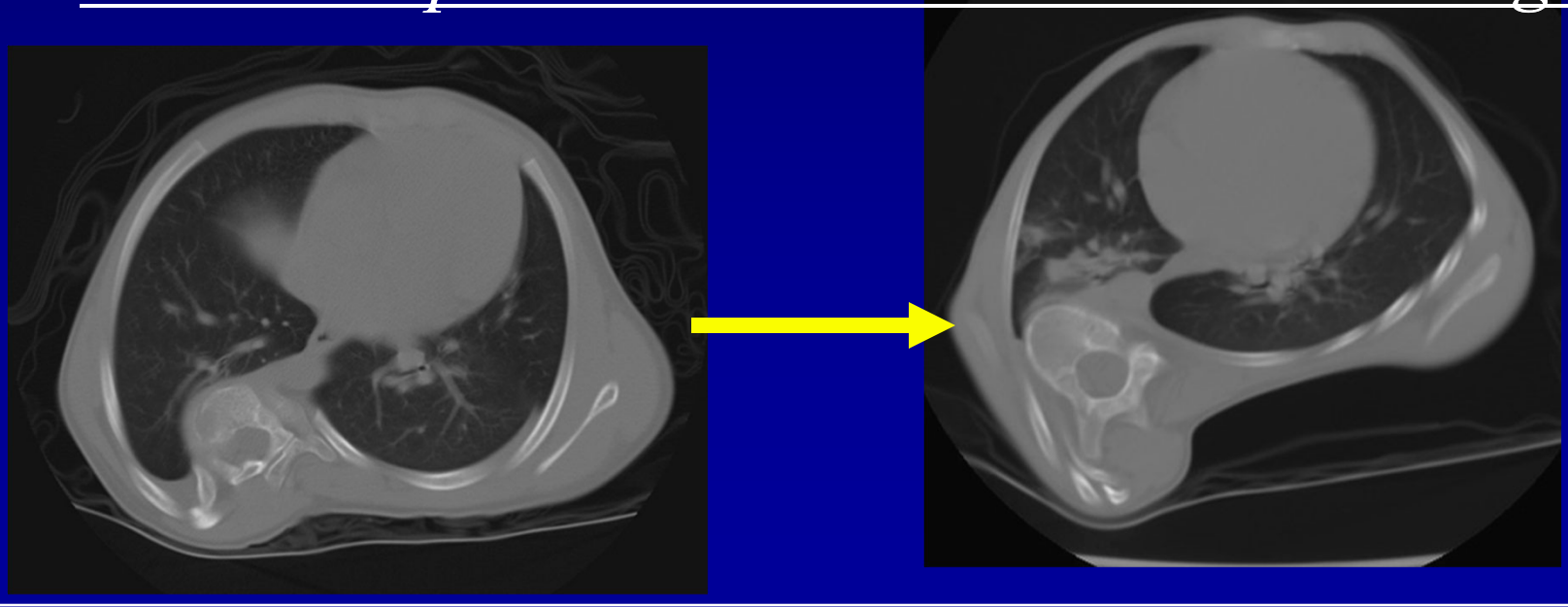
- *Same treatment goals in early onset deformity*:
  - At maturity try to achieve:
    - *Maximum*
      - Spine length, flexibility
      - Thoracic function (volume, movement)
      - Lung growth
    - *Minimum*
      - Surgery
      - Complications
      - Hospitalizations, disability

Growing Rods or VEPTR

- The difficult decision is *when* to intervene surgically?
  - Use evolution of chest deformity as a guide to timing of first surgery?
    - Rationale:
      - Surgical intervention can usually correct/control worsened spine deformity.
      - Surgical intervention less effective for established chest deformity
    - The dilemma:
      - Don't wait to intervene – the chest deformity may be too severe to reverse
      - Don't intervene too early – may get spontaneous fusion beneath growing rods after ? years causing early termination of growth-friendly treatment.

Things we think we know: (maybe)

- *Our EOS operations don't correct severe or established 3-D chest deformity*
  - Goal is therefore not to let severe thoracic deformity occur or progress
  - *When is this? How much is too much?*
  - *Thorax shape more relevant than the Cobb angle*



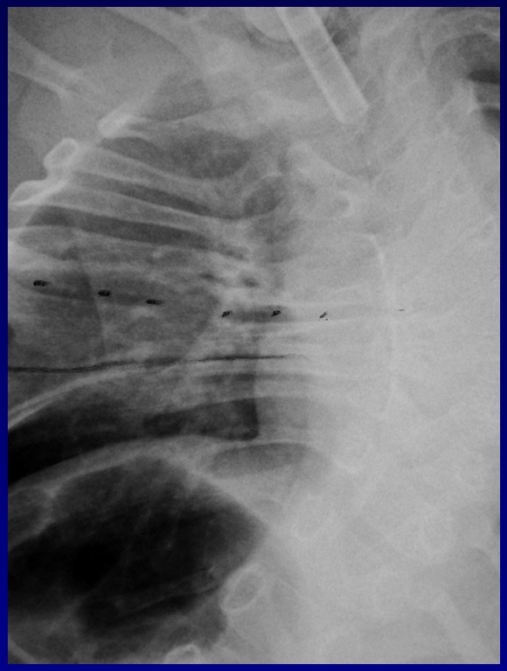
GR or VEPTR for Early onset deformity

- *Factors to consider*, ways to decide
  - *Etiology* of deformity
    - *Spine or chest* dominant or primary
  - Associated:
    - Bone quality
    - Kyphosis
    - Rigidity of deformity
    - Soft tissue coverage
    - Complications of treatment

Growing Rods or VEPTR

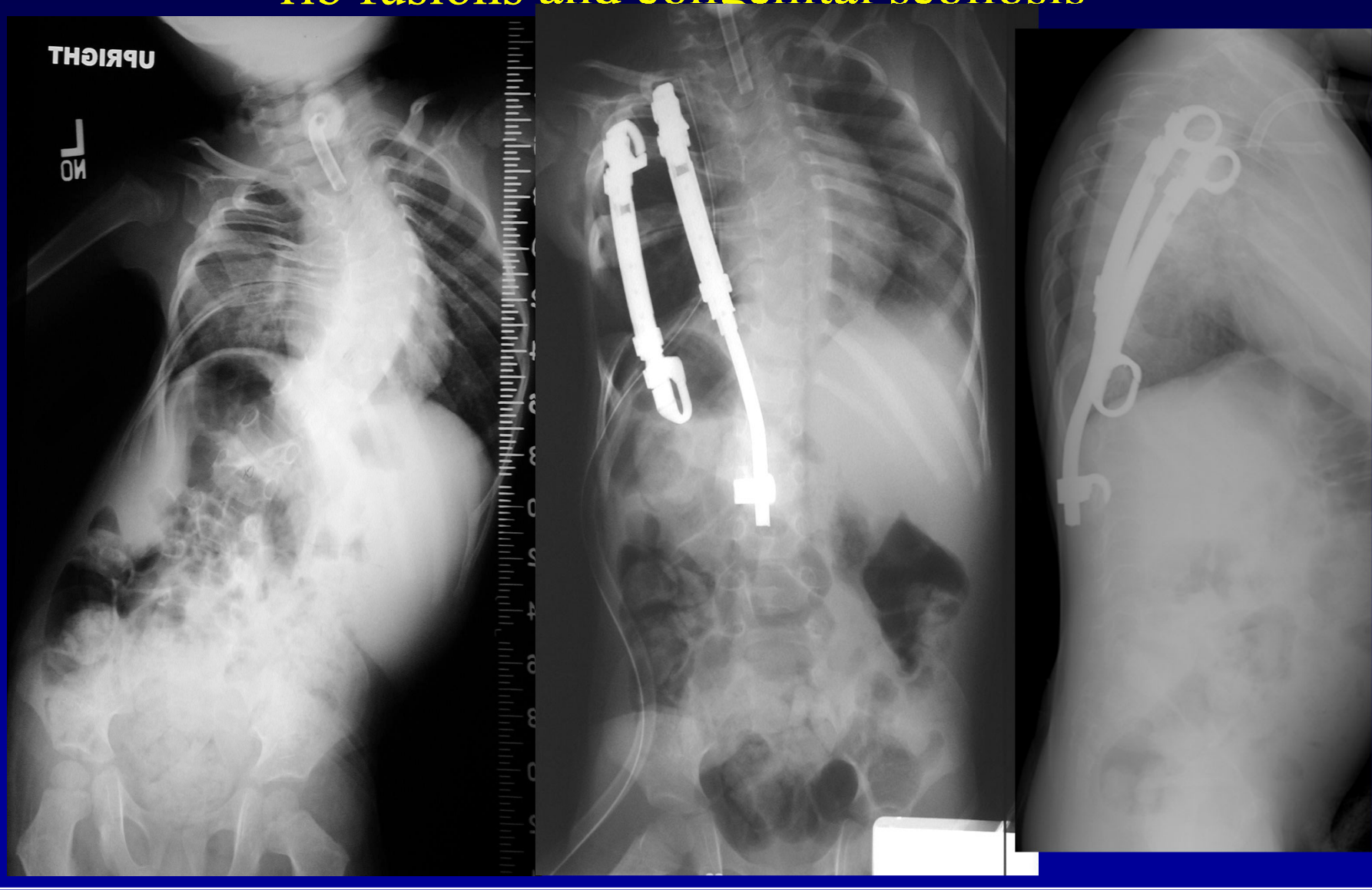
- Etiology of deformity
  - Is the *spine the primary deforming force*?
  - Is the *chest wall the primary deforming force?*
  - Are the *chest and spine* both etiologic factors?

Early Onset Deformity. – etiology as a factor:

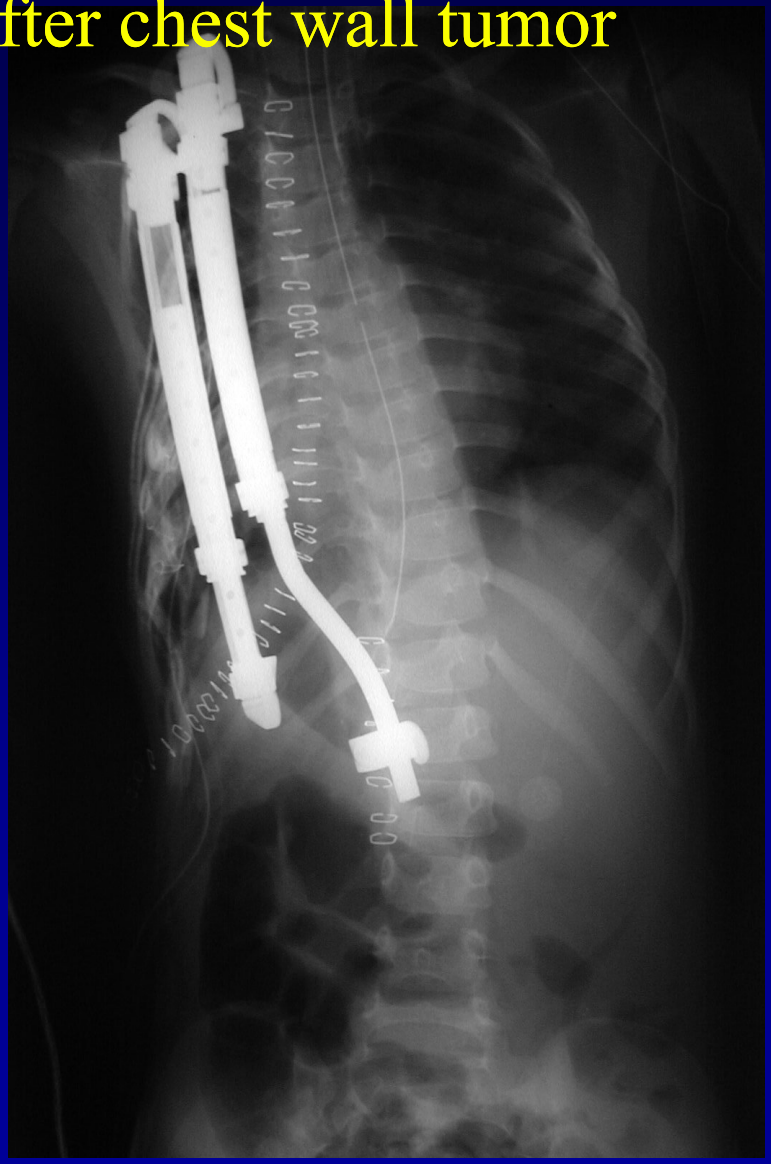


- *Chest wall is primary problem*
  - Massive rib fusions
  - Other *purely thoracogenic scoliosis*
    - Chest wall tumors
    - Esophageal atresia,
    - Multiple thoracotomies
    - Some congenital diaphragmatic hernias
  - *Expansion thoracostomy, VEPTR best choice*
  - 'Spine-only' treatment will be defeated by chest wall tether
- Thoracogenic scoliosis may need skin expanders, staged procedures, flaps,

GR or VEPTR - VEPTR and *expansion thoracostomy* for rib fusions and congenital scoliosis



GR or VEPTR?
Thoracogenic Scoliosis after chest wall tumor

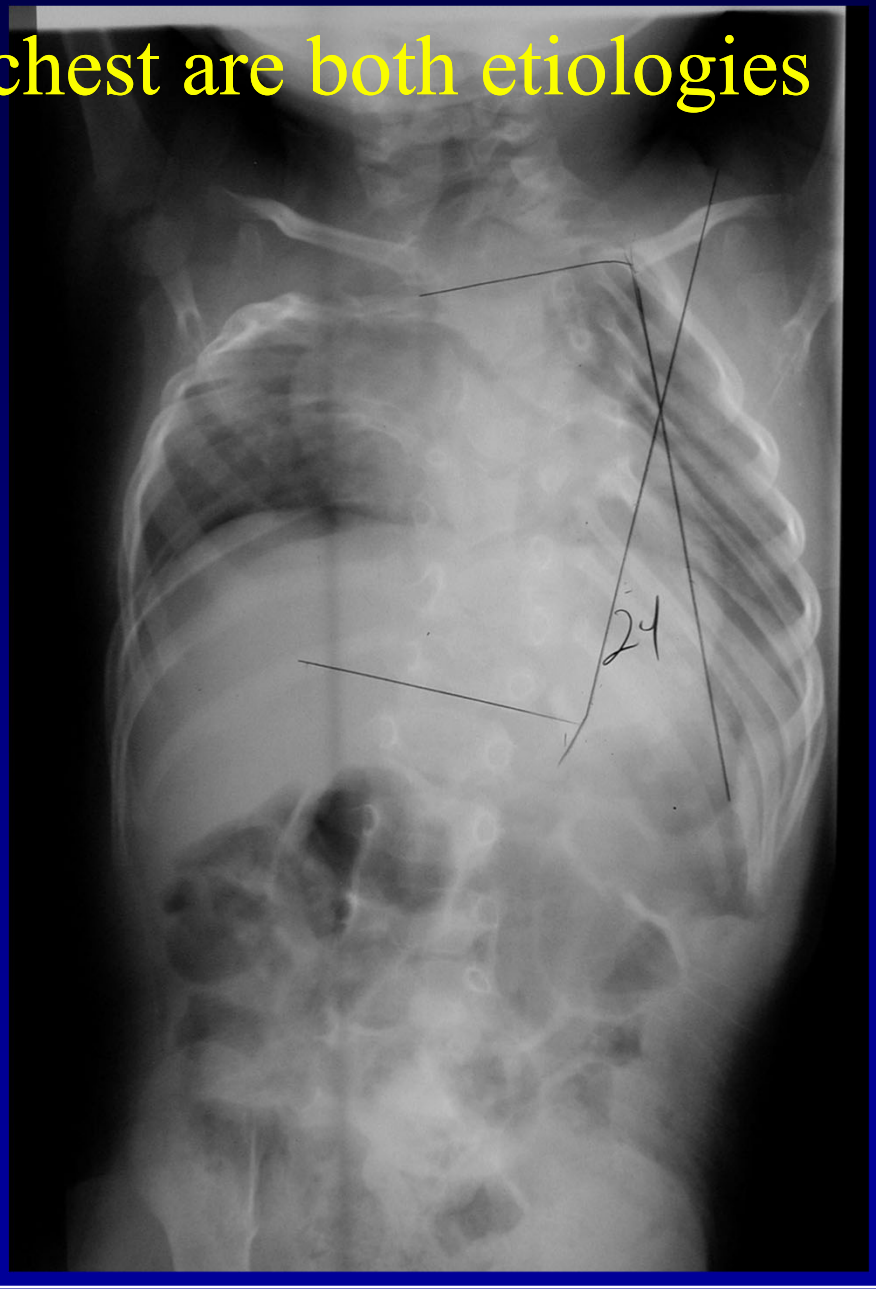


Spine and Chest are both etiologies

- Spine and chest wall *both etiologic factors*
  - *Rib fusions* with congenital scoliosis
    - *Expansion thoracostomy with VEPTR* or expansion thoracostomy with GR best choices
  - *Established, severe chest wall deformity plus severe spine deformity*
    - Severe windswept thorax
      - GR approaches primary spine problem
      - Is VEPTR any better?

GR or VEPTR Spine and chest are both etiologies

- 2 y.o with progressive curve, increasing nighttime O2 requirement
- Multiple unilateral rib fusions
- Vertebral bars
- *Note worsening 'normal' hemithorax*



GR or VEPTR Spine and chest are both etiologies

- Early intervention for the sake of chest shape
  - Three thoracostomies
  - VEPTR
  - ‘normal’ side of chest improving slowly?

Age 11, PFT's 45%

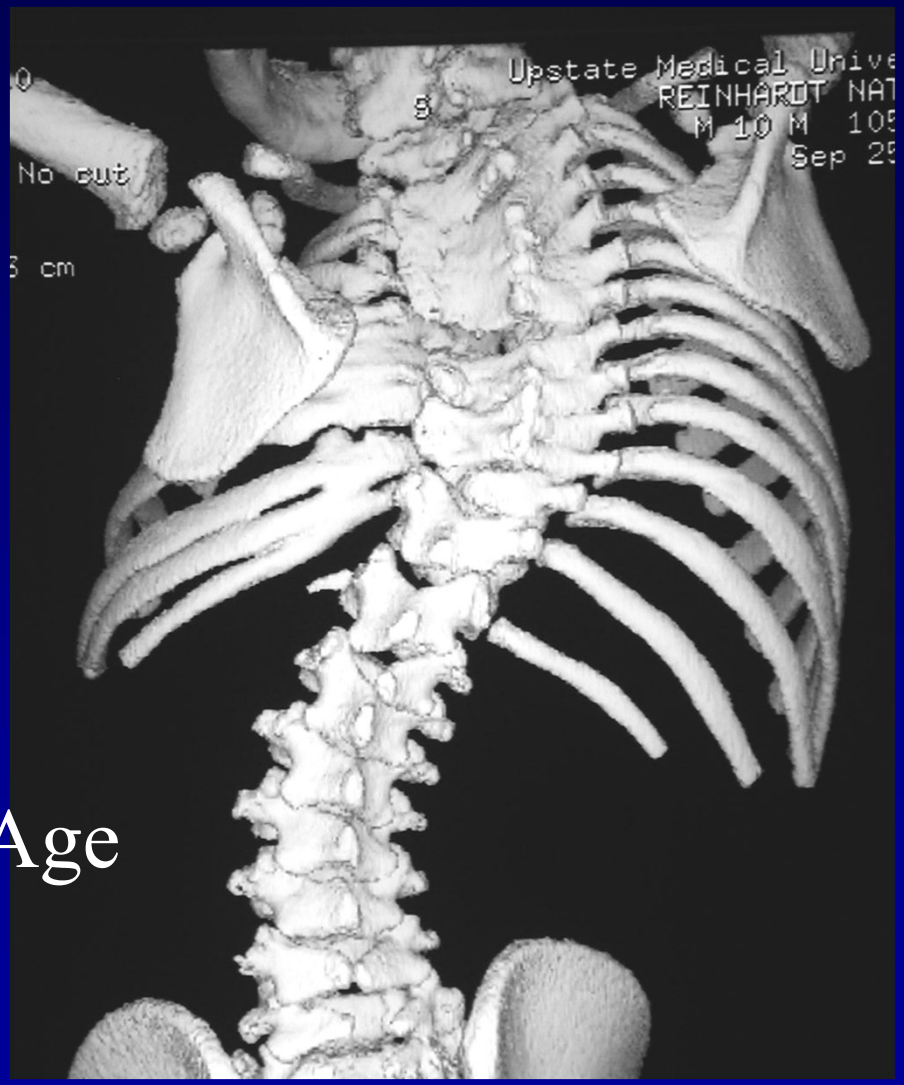
- *Partial* control of spine deformity, chest deformity

GR or VEPTR Spine and chest are both etiologies

Example: progression after in situ fusion

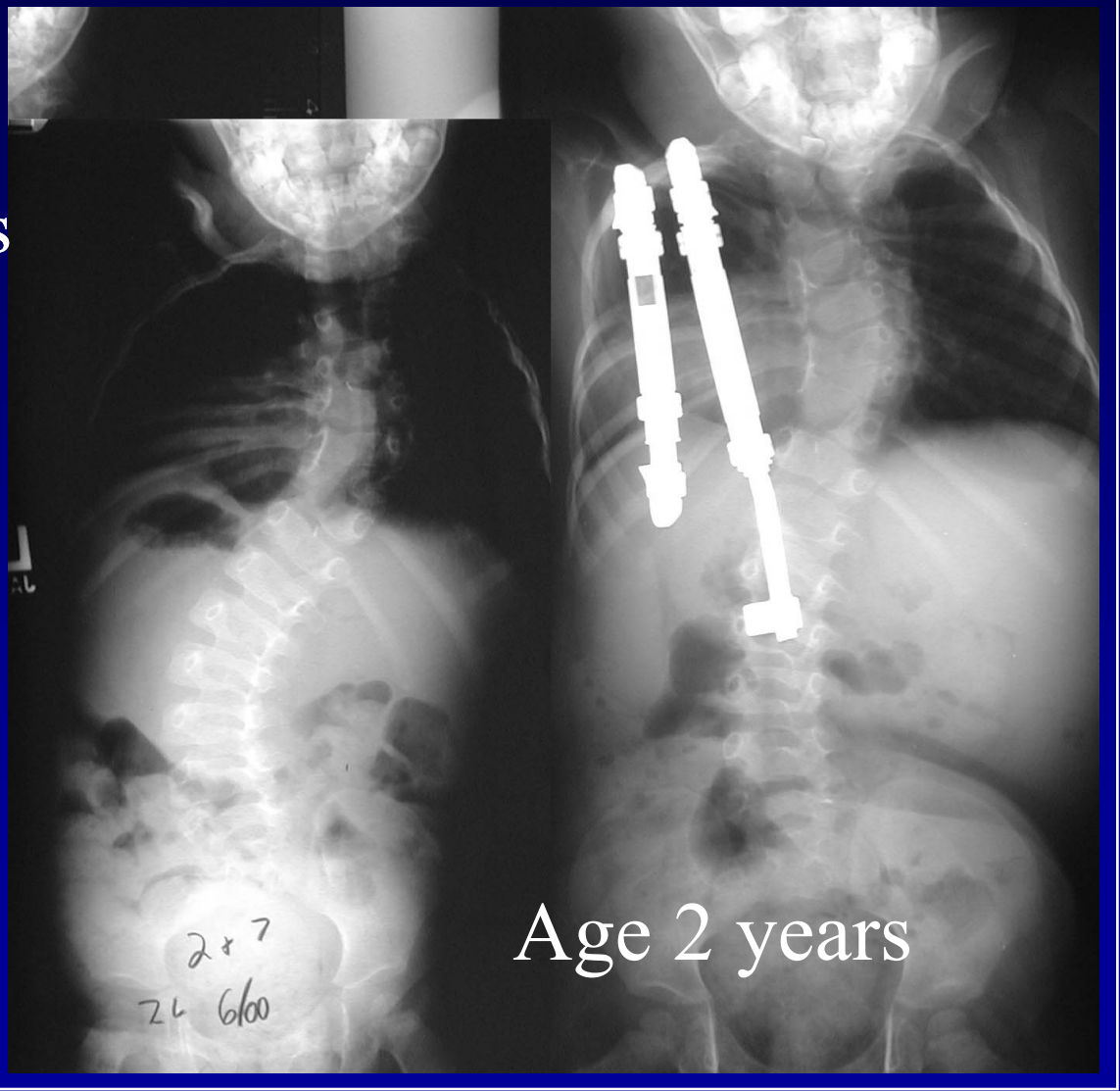
GR or VEPTR Spine and chest are both etiologies
Age 2 – s/p in situ fusion age 6 mos

Curve Progression at Age
2 years



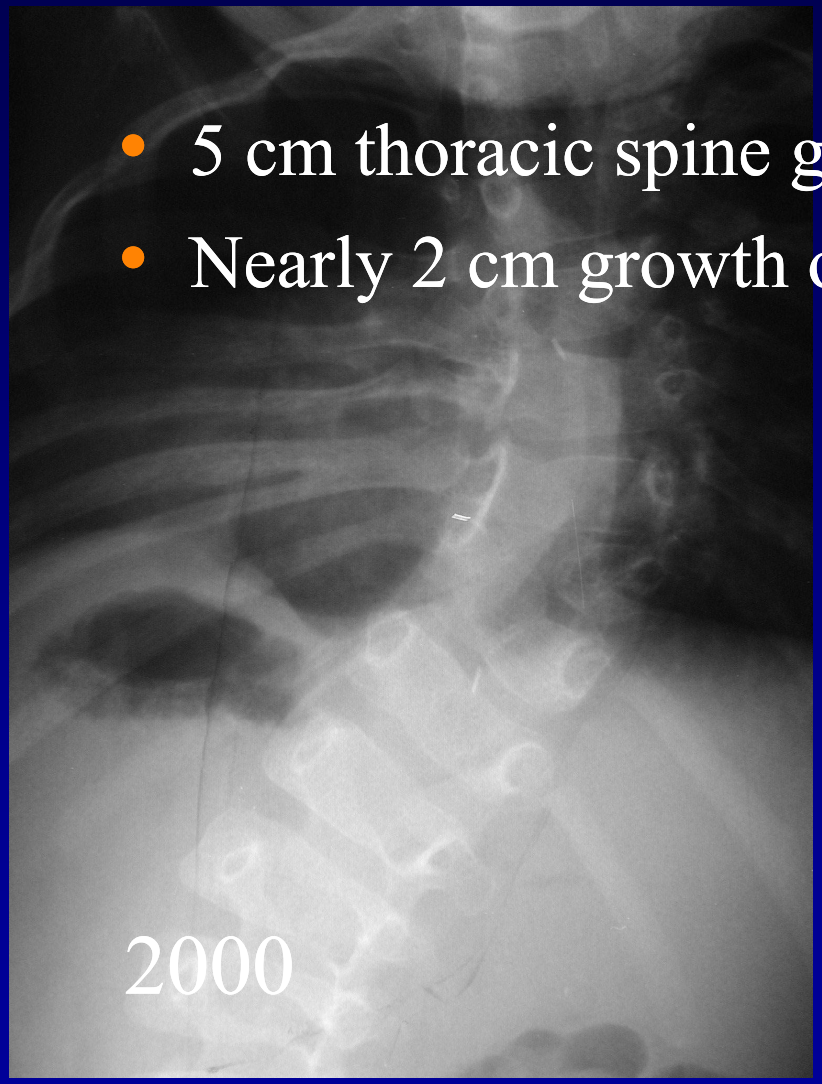
Curve Progression at Age 16 months
VEPTR, expansion thoracostomy, prior fusion

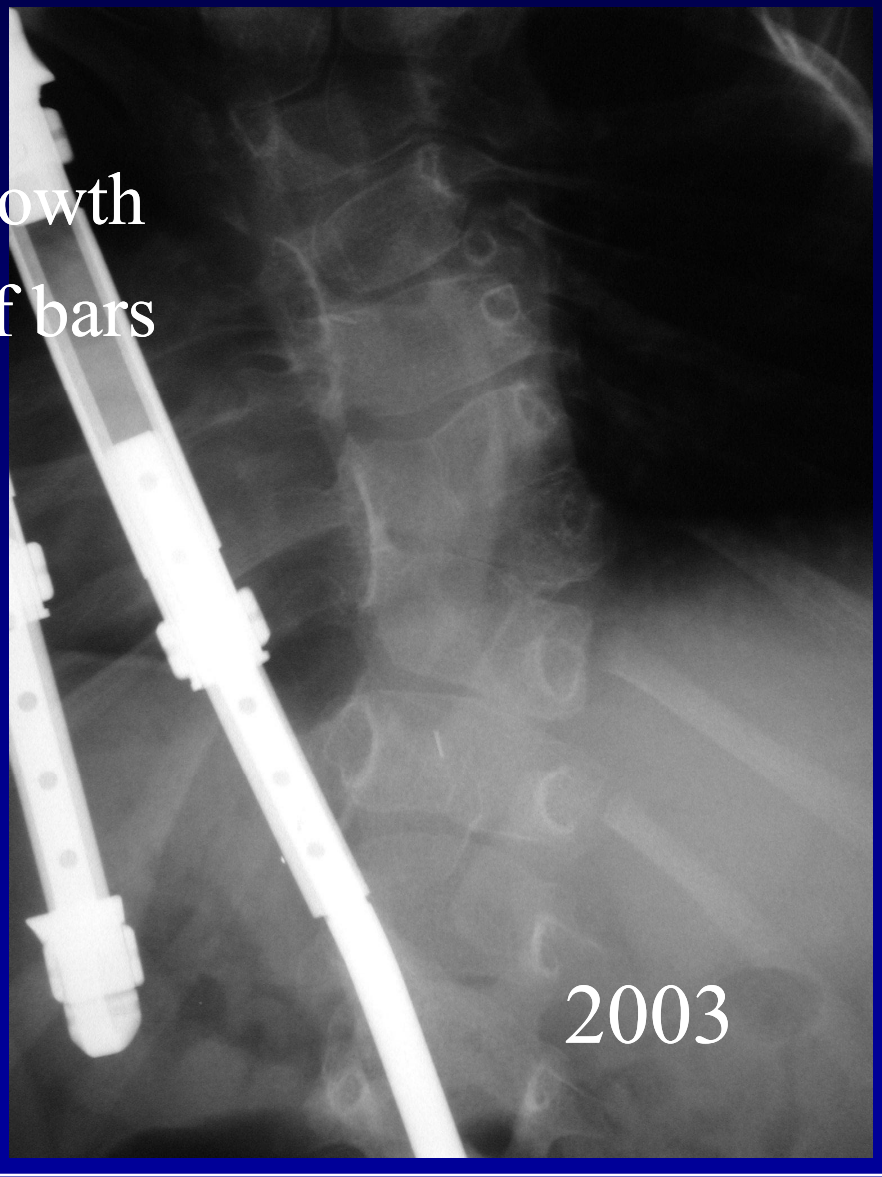
- Two rib osteotomies
- One rib-to-rib device
- One rib-to-spine device



Bar growth – 4 yrs after first VEPTR

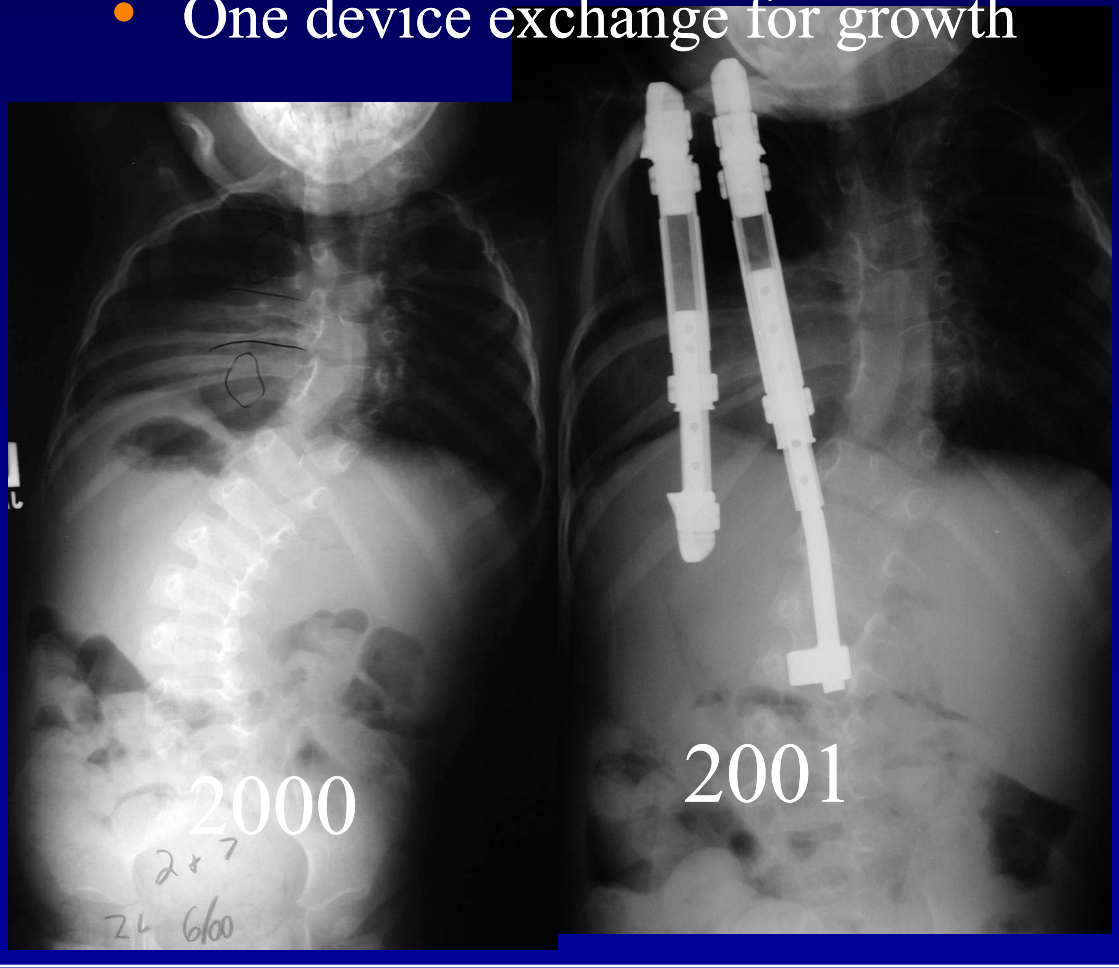
- 5 cm thoracic spine growth
- Nearly 2 cm growth of bars

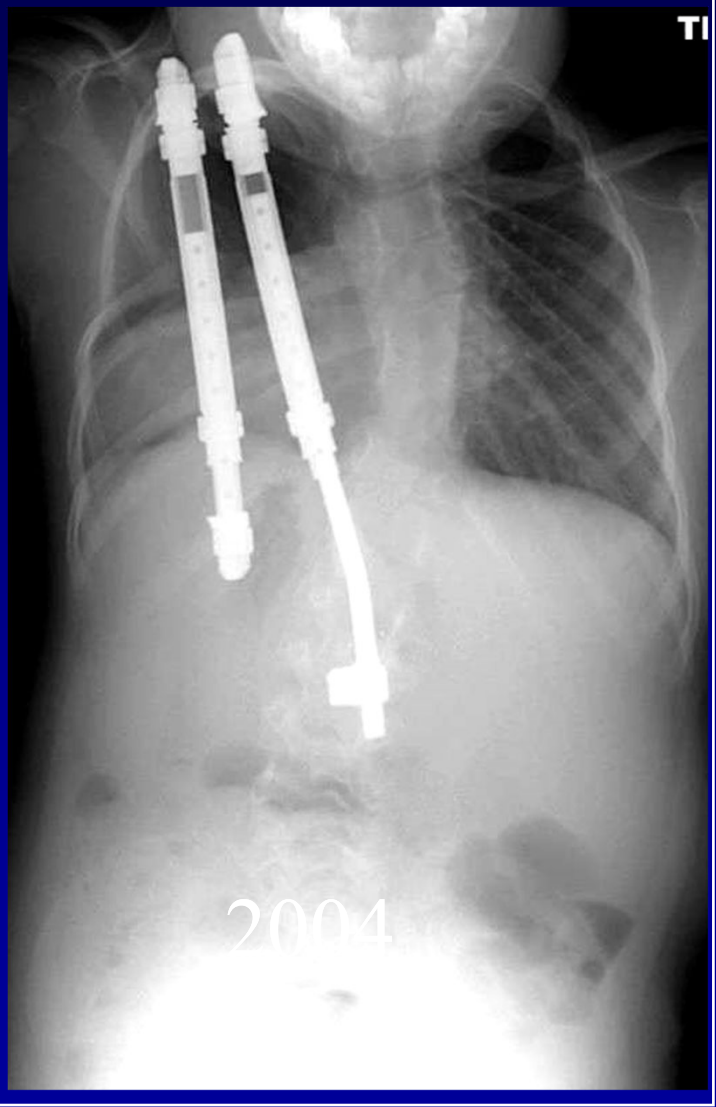




Spine growth – 4 yrs after first VEPTR

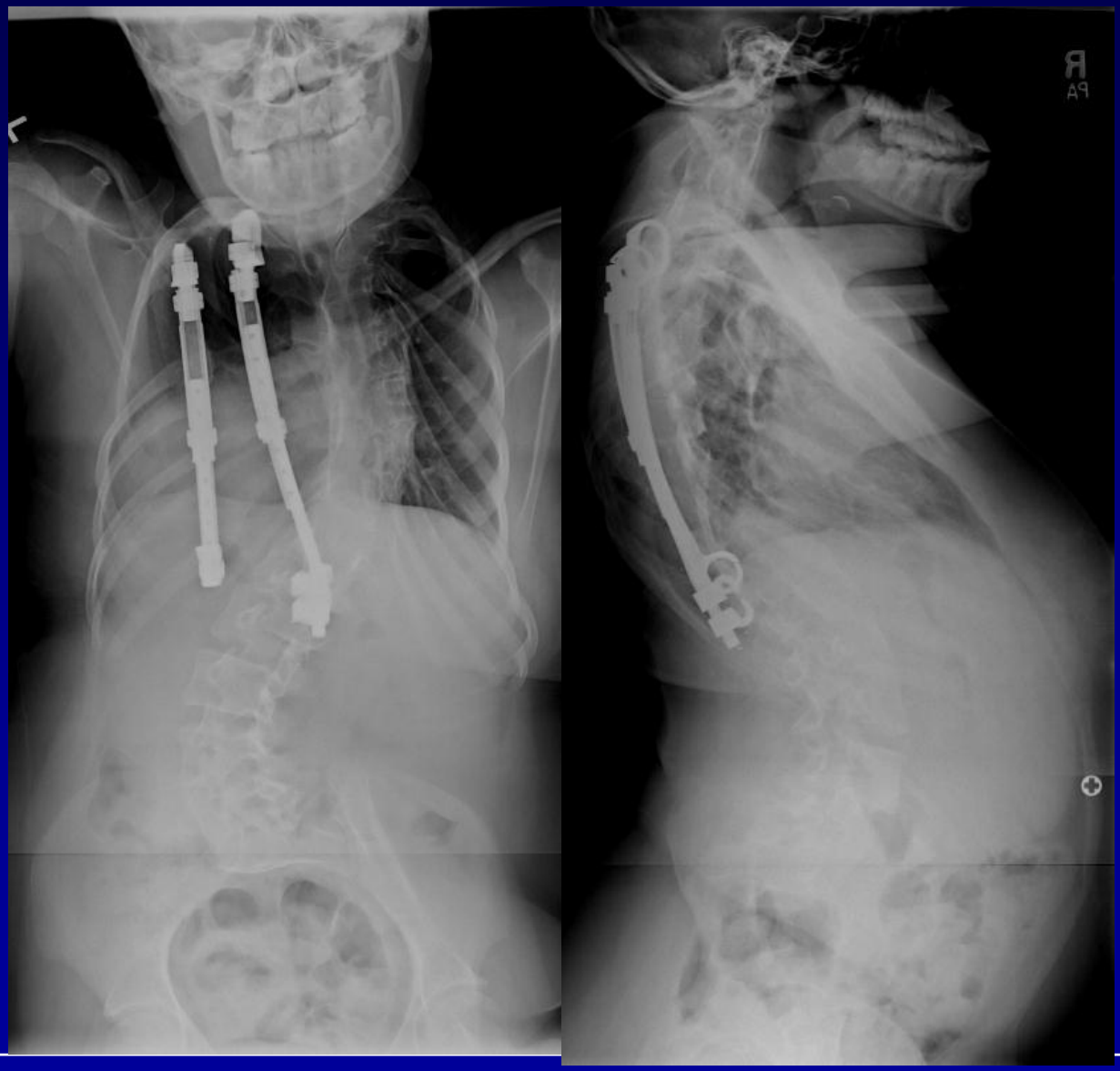
- Continued growth - 6 lengthenings
- One device exchange for growth





Age 13 – two years post menarchal- 11 years after first VEPTR

- PFT's 55%%
- Devices left in place.
- Thorax far from normal
- No fusion needed?



GR or VEPTR – Chest and Spine both a problem

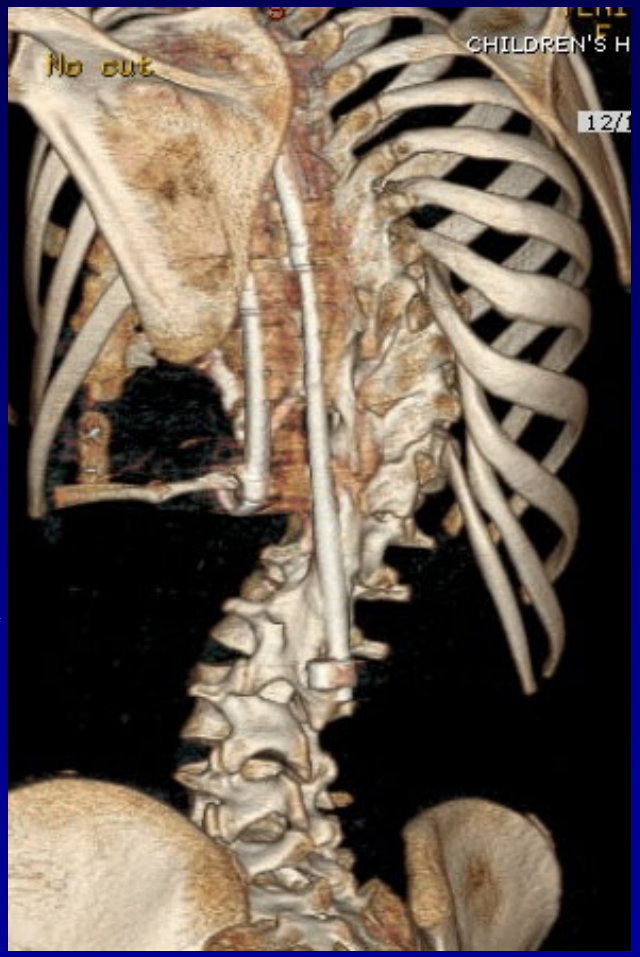
- 3yo with progressive thoracic deformity
  - Congenital rib fusions
  - Multiple vertebral anomalies.
- VEPTR and expansion thoracostomies age 3

GR or VEPTR – Chest and Spine both a problem

- Multiple lengthenings
- Exchange x 2
- Age 14, mature
- Unhappy with waist asymmetry
- Active as cheerleader
- VEPTR removal, osteotomies, final fusion

GR or VEPTR – Chest and Spine both a problem

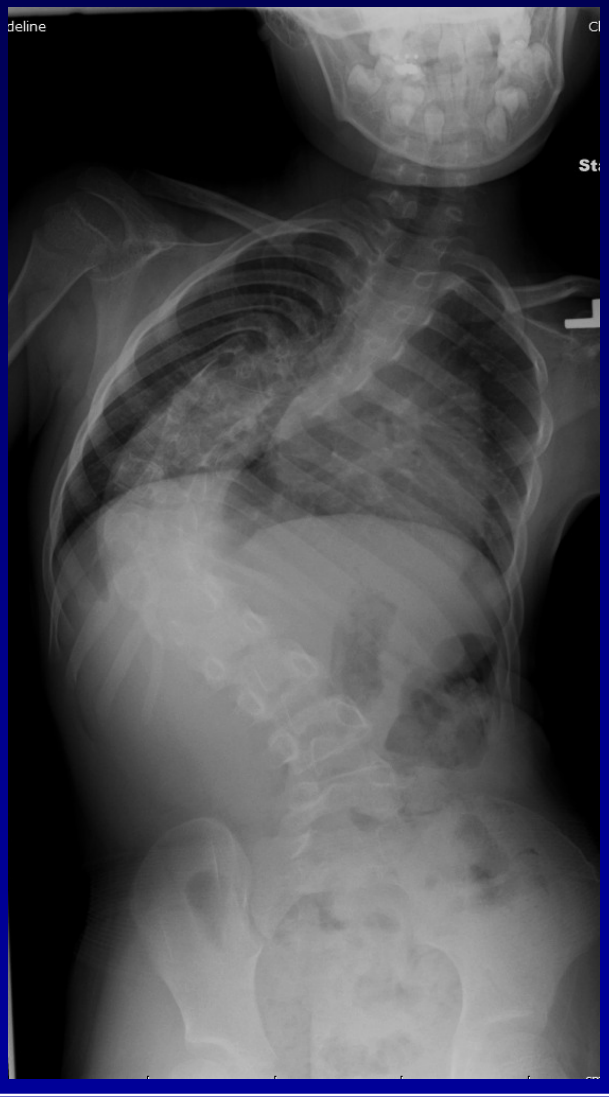
- *Final spine fusion easier with VEPTR*?
- Rib fusions, scarring expected
- Spontaneous spine fusions:
  - Below rod at lower end
  - In normally segmented part of curve
- *Far from easy after VEPTR*

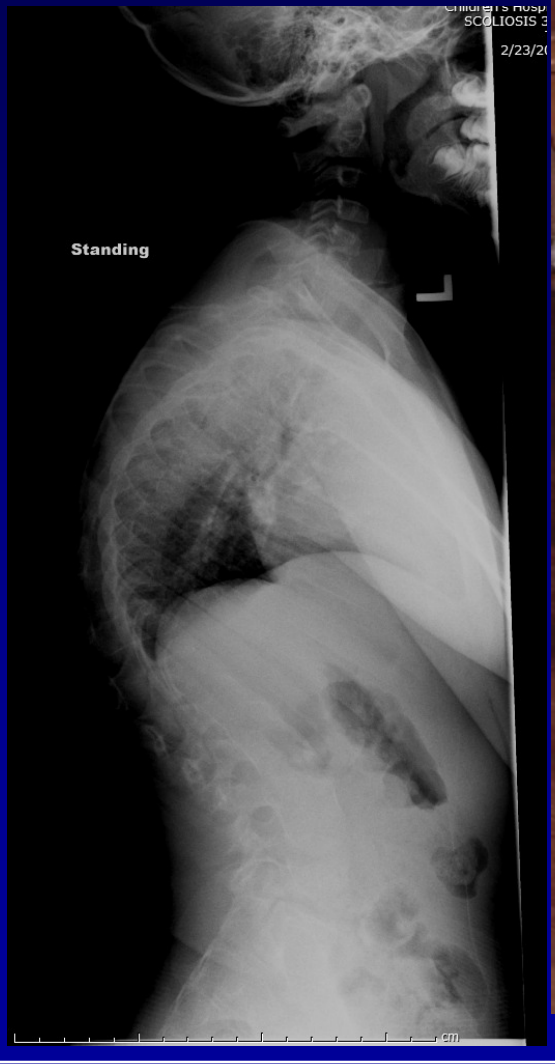


GR or VEPTR? – *Spine* is the problem

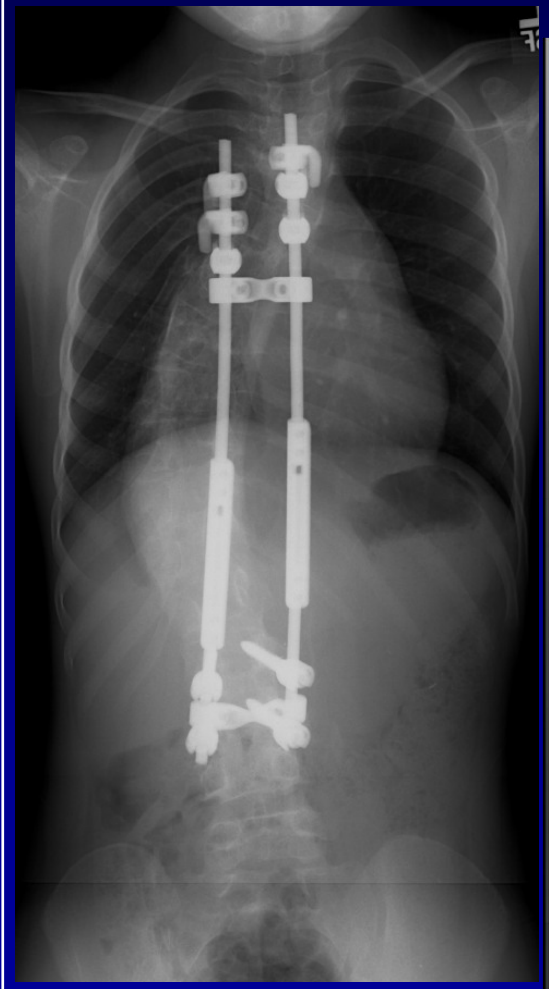
- *Infantile idiopathic* scoliosis (normal segmentation)
  - *Moderately severe deformity*
    - (Spine still worse than chest deformity)
  - *Dual Growing rods*
  - Rationale for choice:
    - Chest wall, although deformed, is mobile.
    - Expansion thoracostomy, VEPTR, may stiffen chest wall?

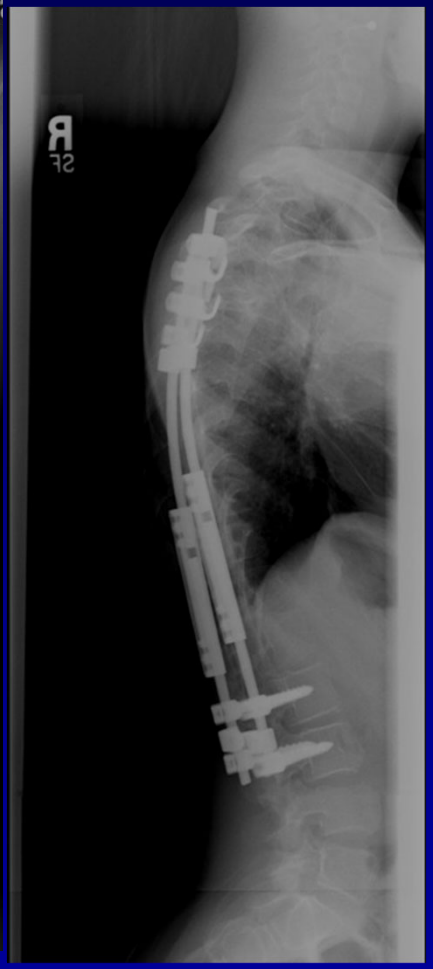
Infantile idiopathic scoliosis, moderately severe rotational chest deformity – Age 5



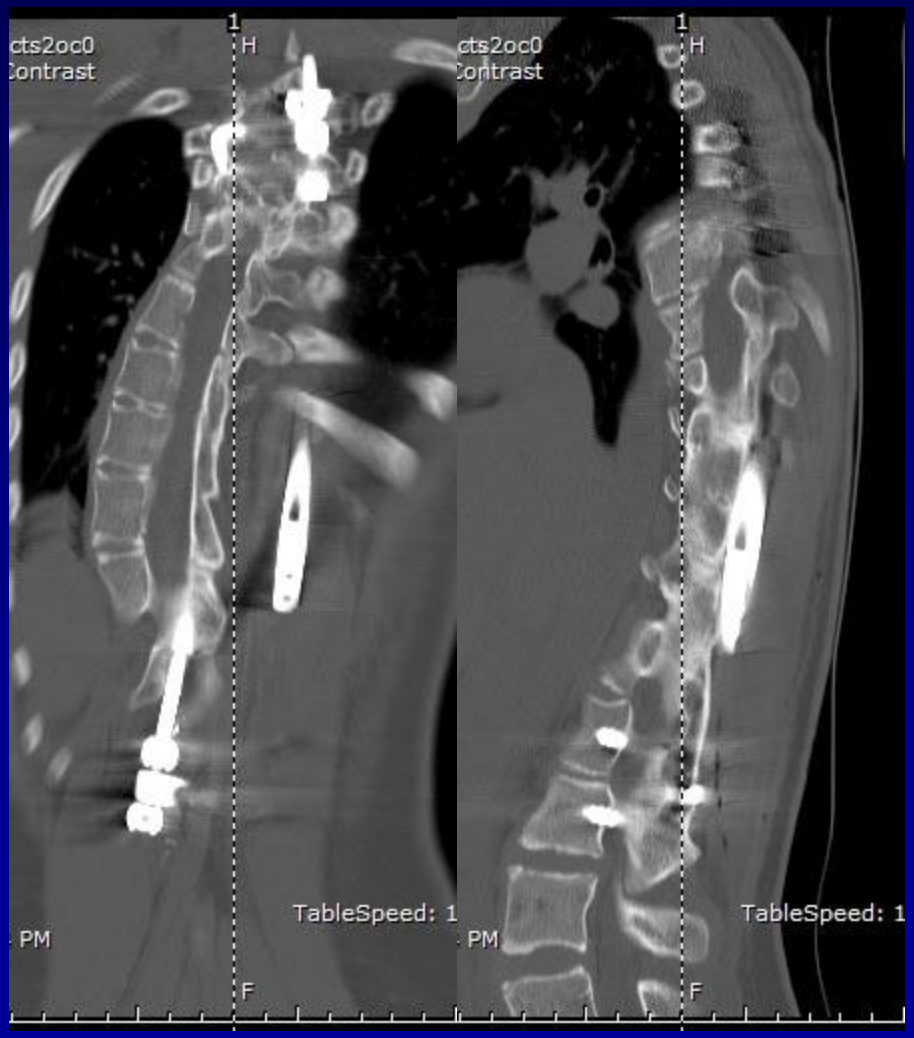


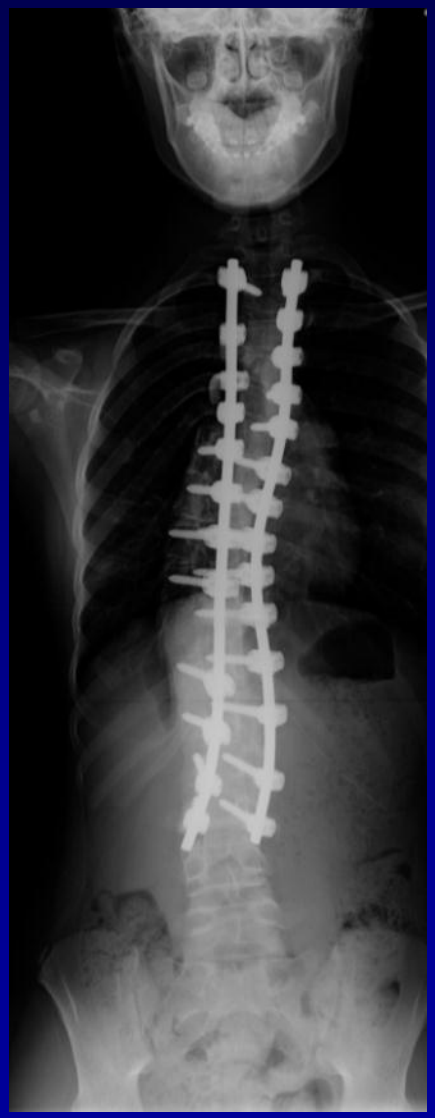
Infantile Idiopathic Scoliosis, dual growing rods – earlier would have been preferable? – Age 5





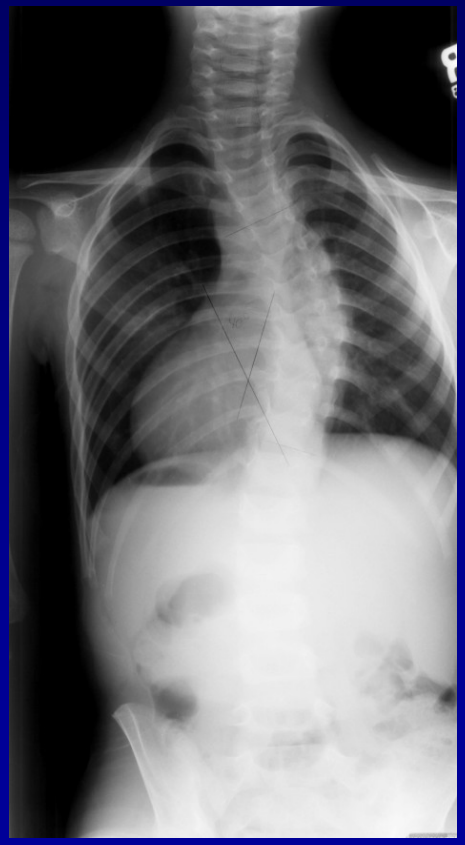
Age 12, after 7 years of lengthening, stopped by spontaneous fusion. Osteotomies, definitive fusion

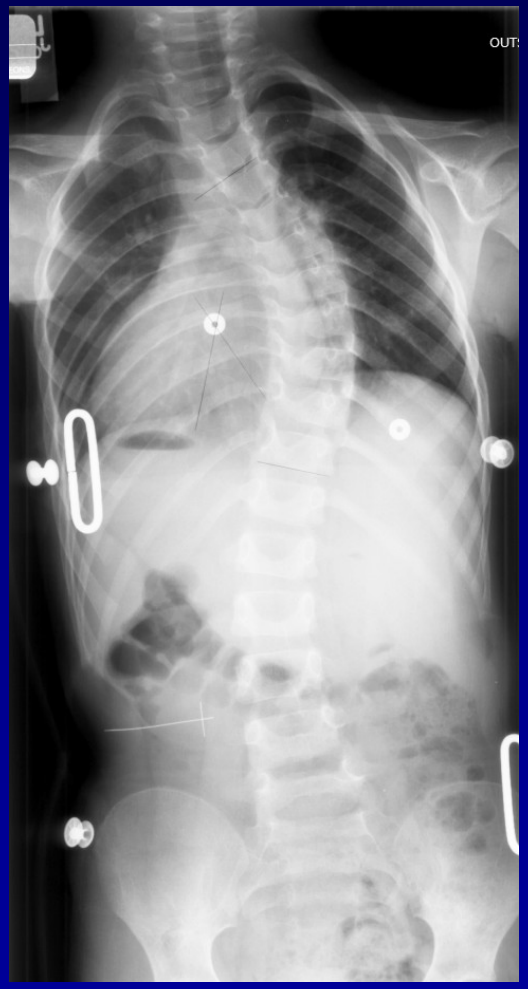




GR or VEPTR ?

Infantile idiopathic Age 3, age 4 in brace, age 5 after casting

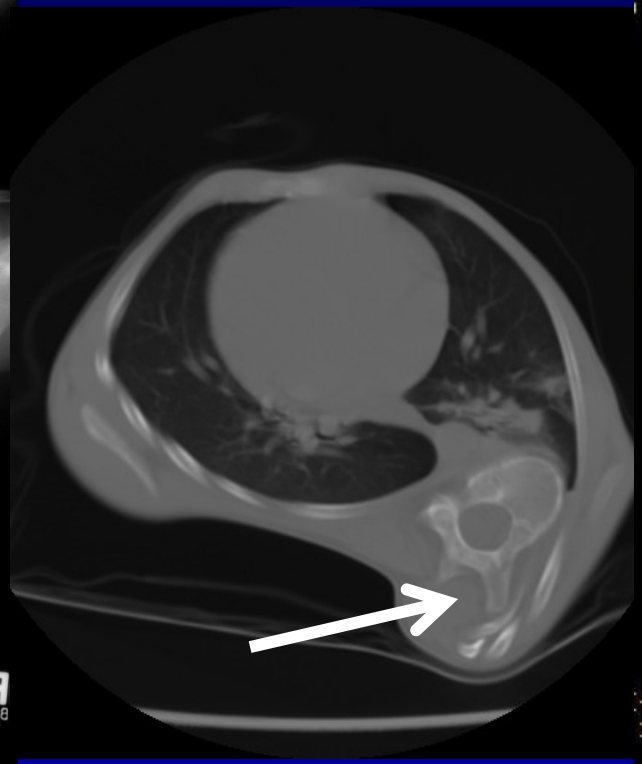




GR or VEPTR?
More casting, bracing -Age 7, Age 9

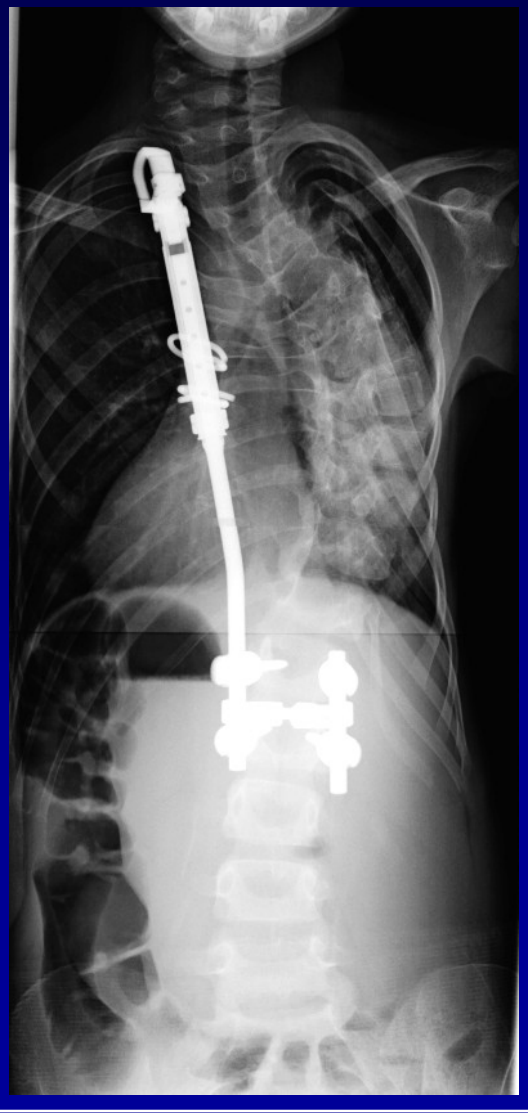
GR or VEPTR – IIS casted until age 9
Preop Windswept chest. Note severe thoracic lordosis

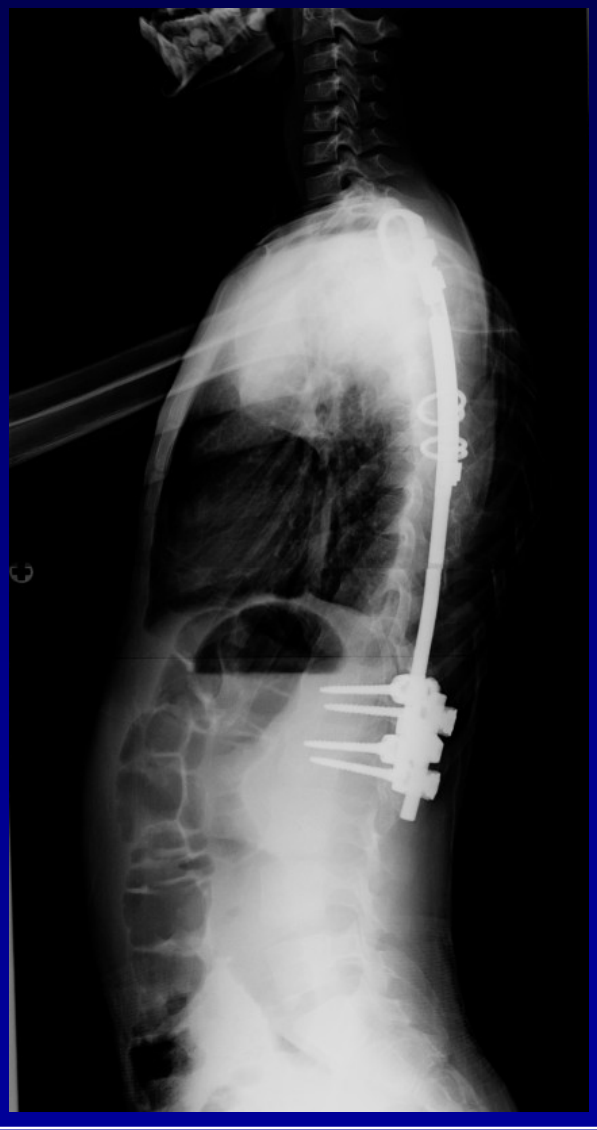
GR or VEPTR? - Infantile idiopathic, Age 9



Windswept Chest

IIS - VEPTR first stage

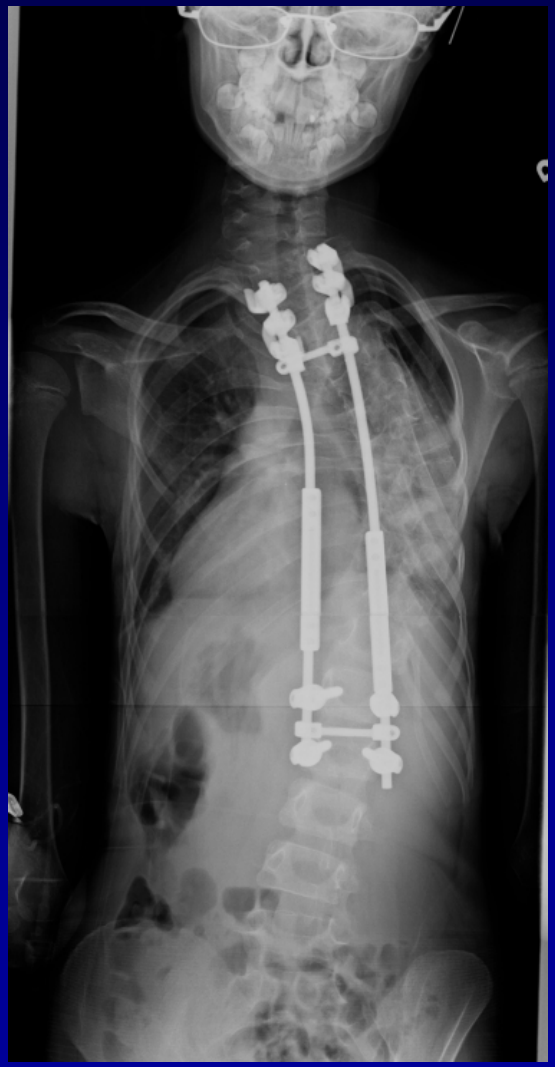


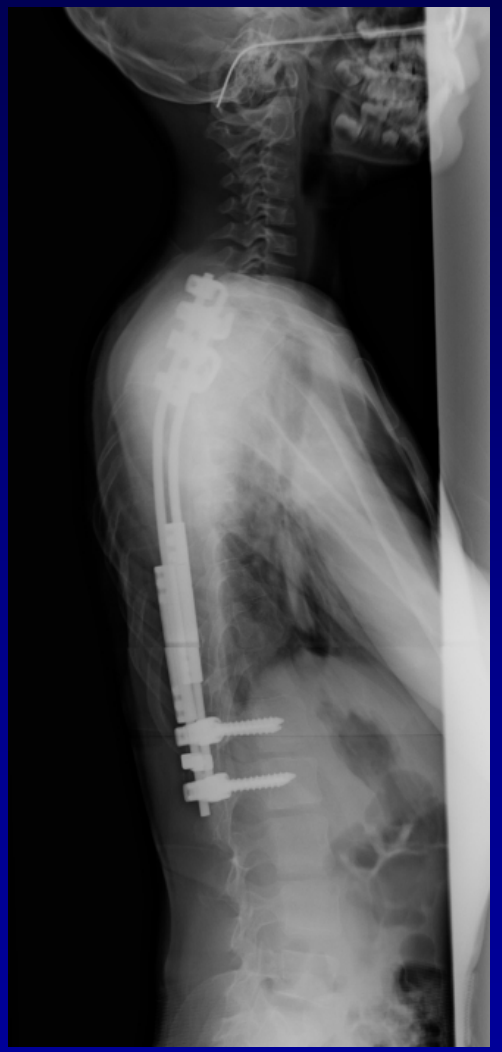


Chest shape after first VEPTR?

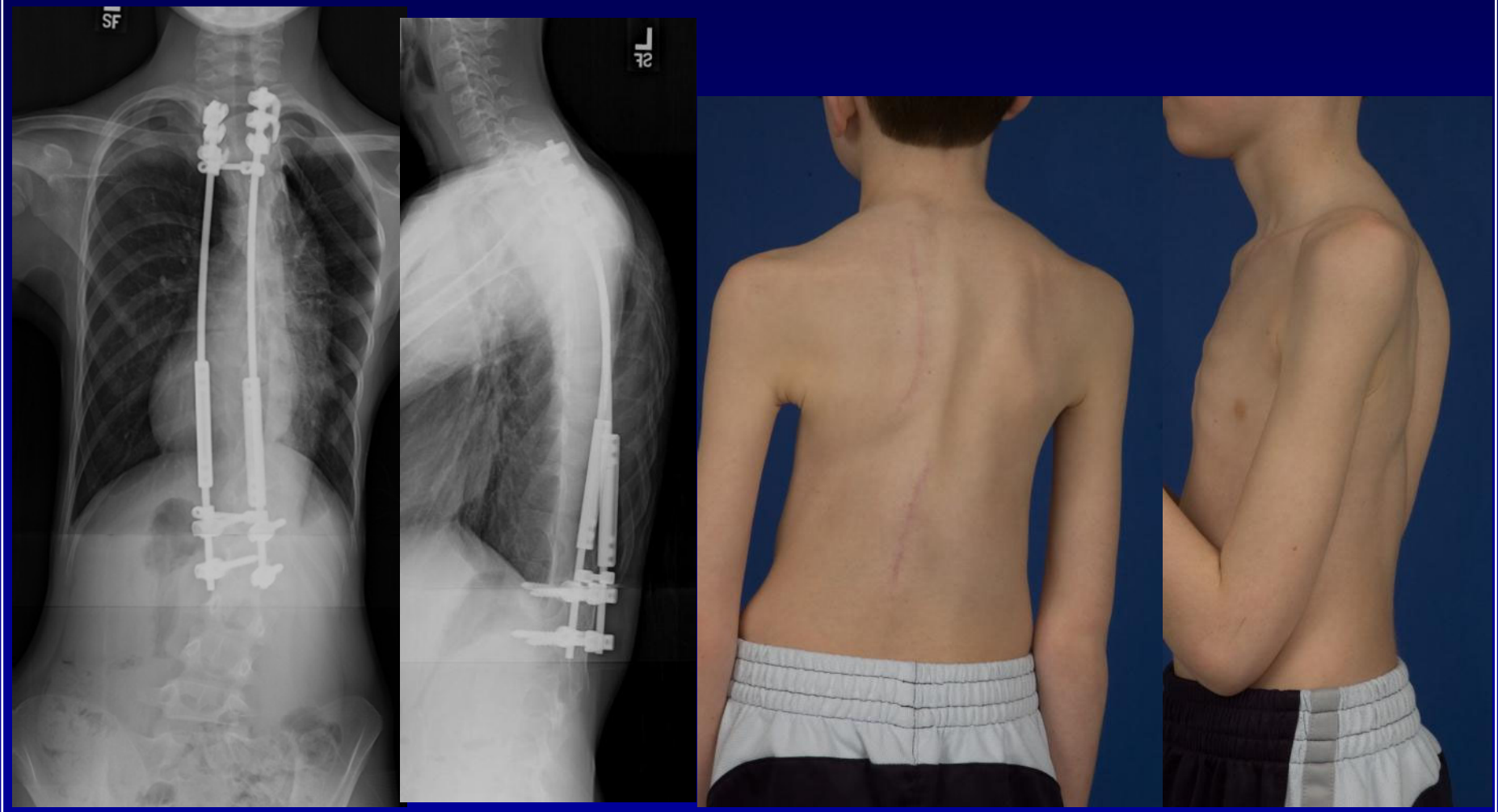
- No change in occluded convex thorax after concave expansion thoracoplasty
- "Parasol procedure" with VEPTR planned for convex, collapsed side
- VEPTR rib attachments cut through at 3 weeks
  - Better to have had an additional load-sharing device?
  - No other rib attachments available
- Converted to dual growing rods

VEPTR fixation lost. GR inserted.
Spine under control, *chest irrevocably altered*.





After 3 years lengthening, Age 12 .
FEV1 65% Chest still windswept

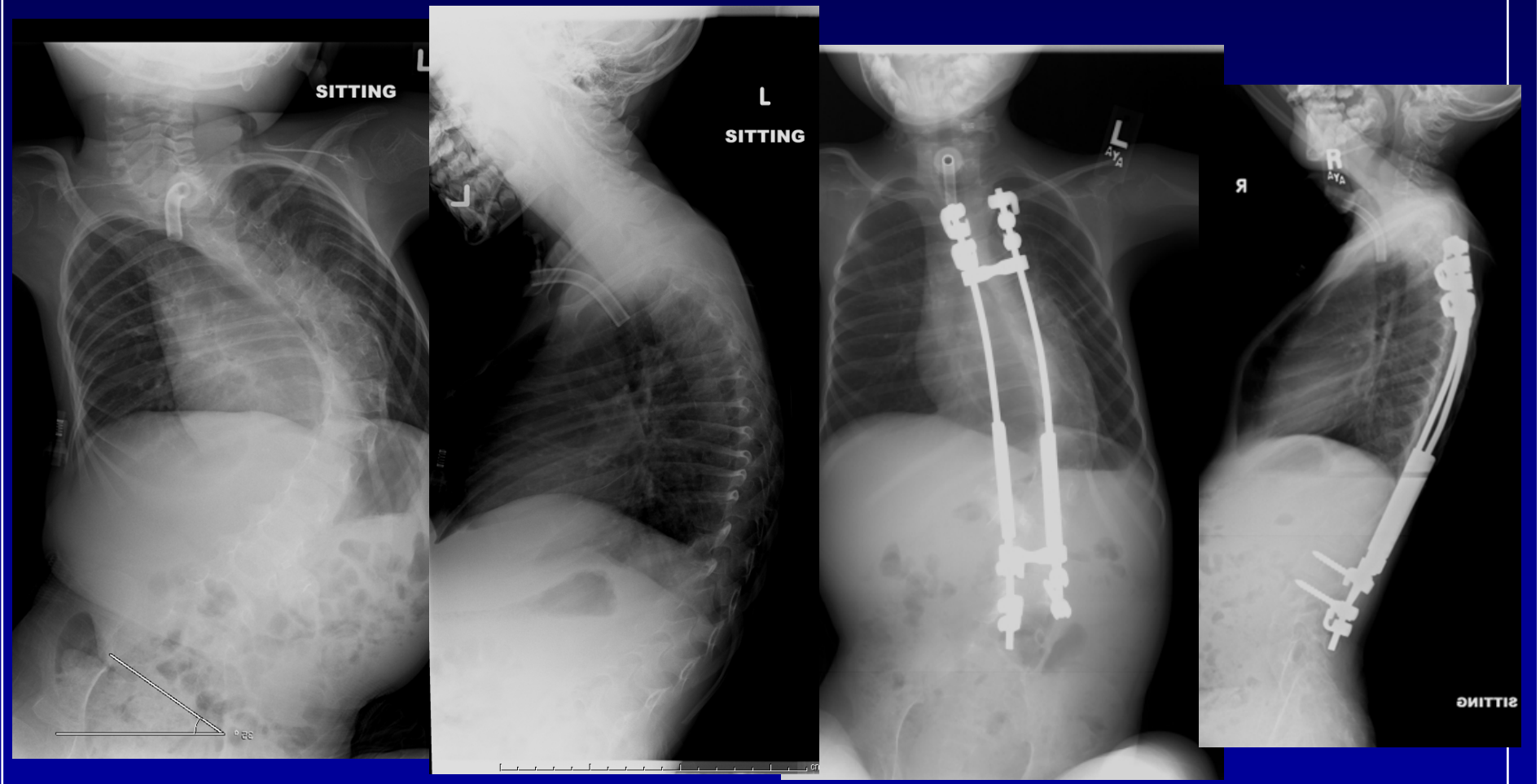


Growing Rods or VEPTR
Neuromuscular deformity – VEPTR or GR?

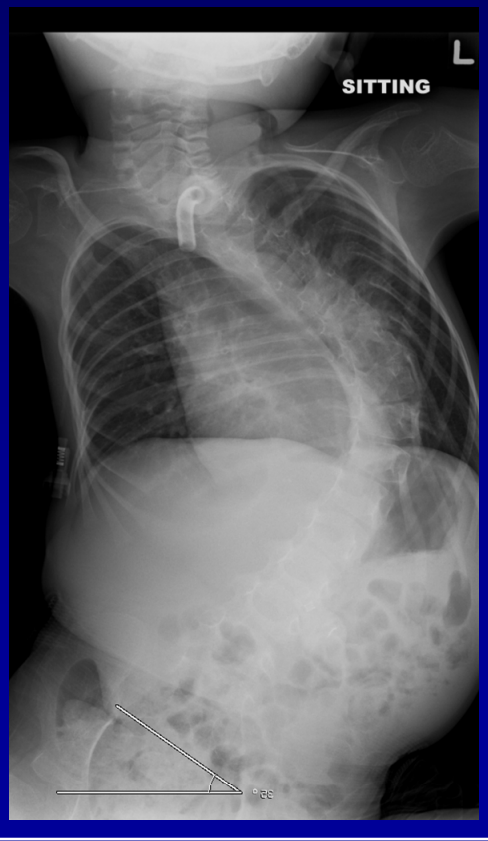
- High-tone ('CP')
  - Neither very good. Rarely indicated in 'CP' high tone
  - Personal opinion – more trouble with rib drifting in 'CP'

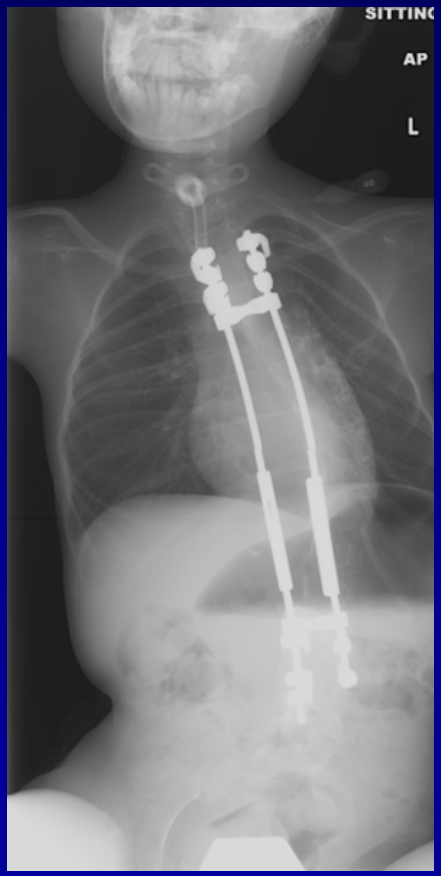
- *Low-tone: SMA*, arthogryposis, myopathy
  - VEPTR directly treats the 'parasol' deformity
  - Multiple fixation points – tandem connectors
    - *Xrays better with VEPTR, but are the patients any better*?
    - *Minimal chest wall movement* – breathing is all diaphragmatic, hence GR may be as good as VEPTR!

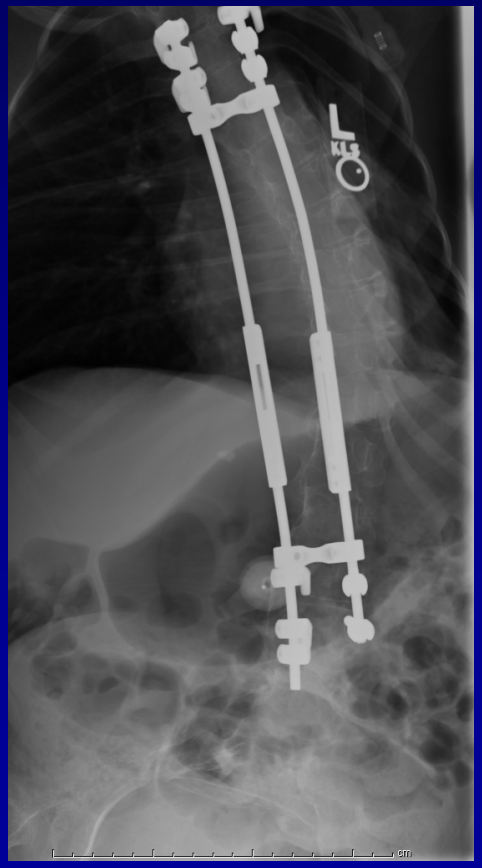
GR or VEPTR?
5 yo with SMA trach but not vent dependent



GR or VEPTR?
SMA ages 5, 10, 12



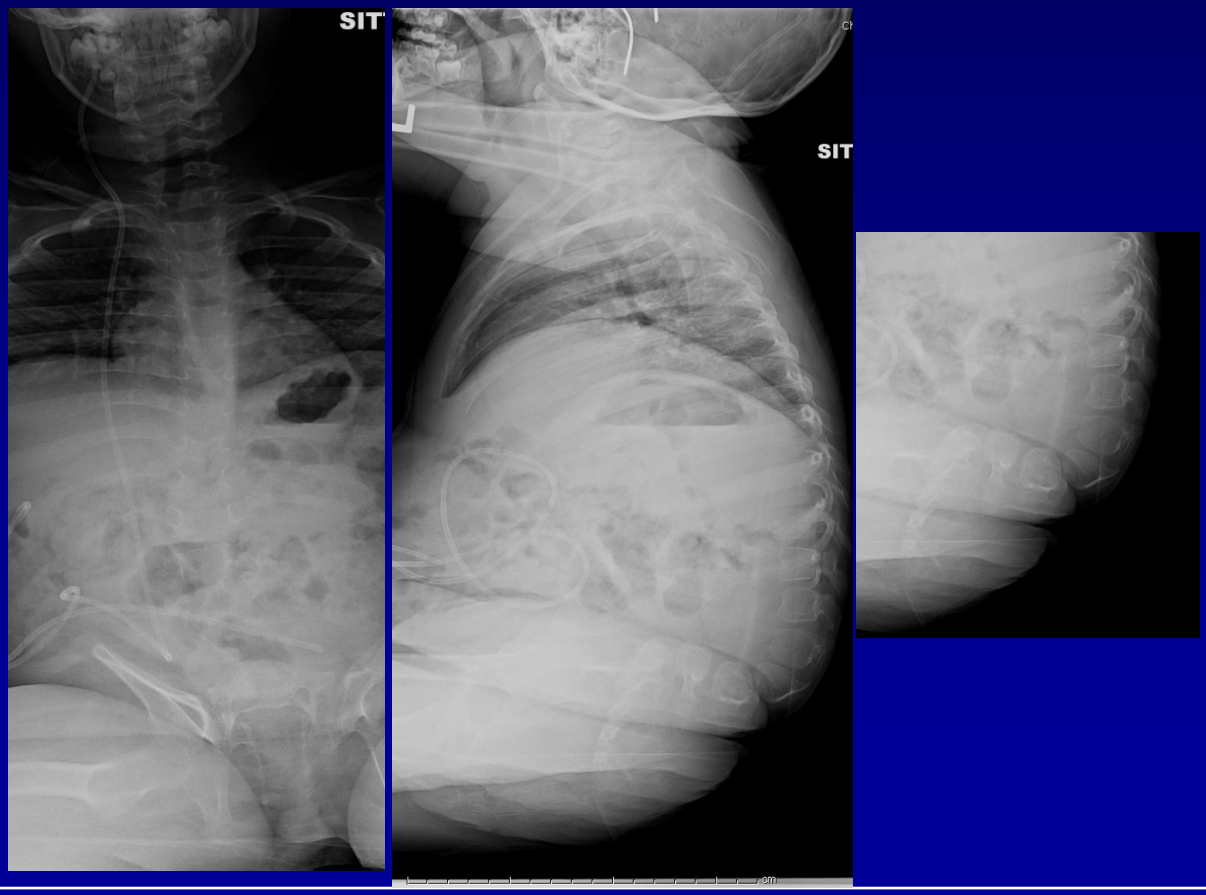




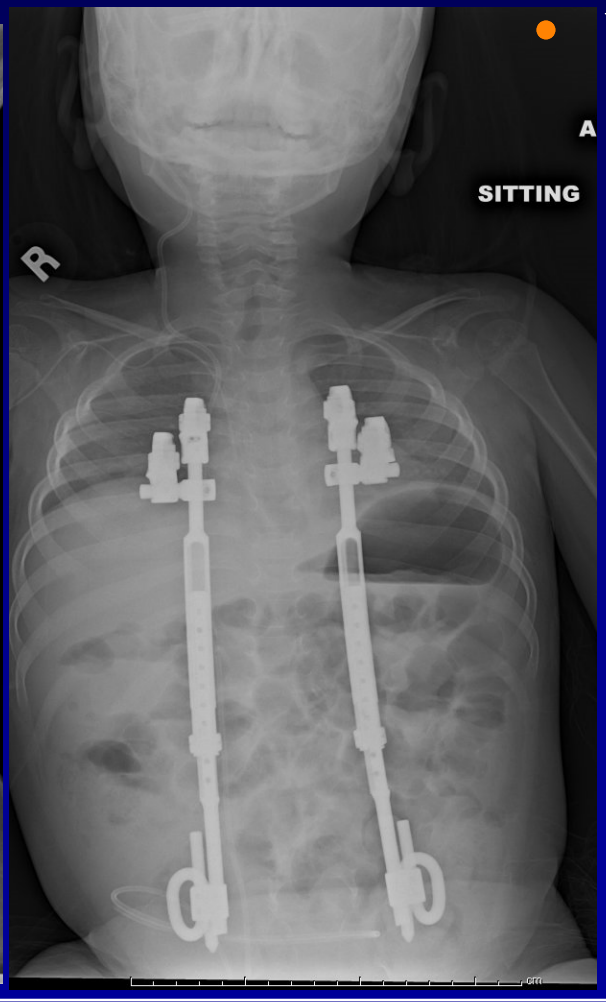
GR or VEPTR – usefulness of tandem VEPTR II anchors to horizontalize parasol deformity?

GR or VEPTR - *Myelodysplasia* with collapsing kyphosis or scoliosis

- Age 5, thoracic level.
- Worsening deformity, impending skin breakdown



GR or VEPTR - *Myelodysplasia* with collapsing kyphosis or scoliosis



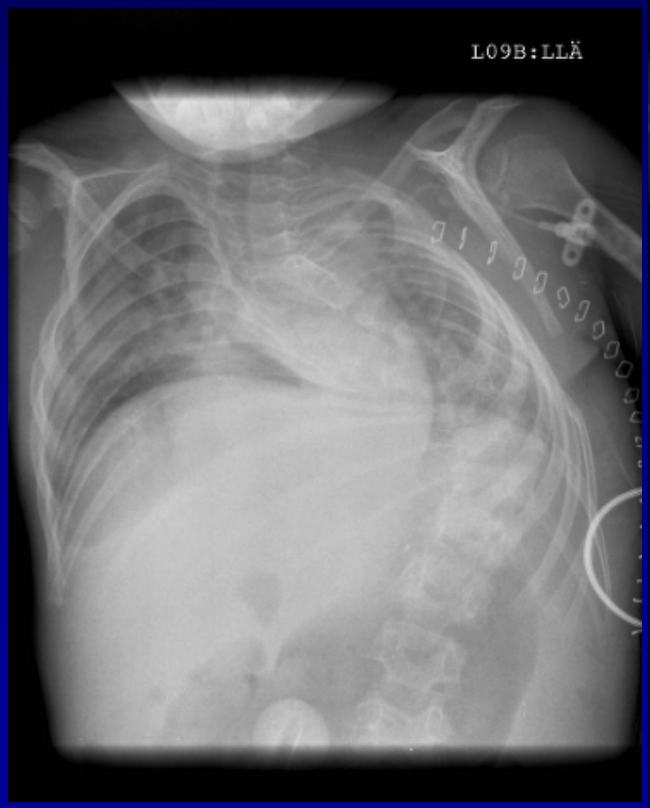
VEPTR II rib to pelvis.

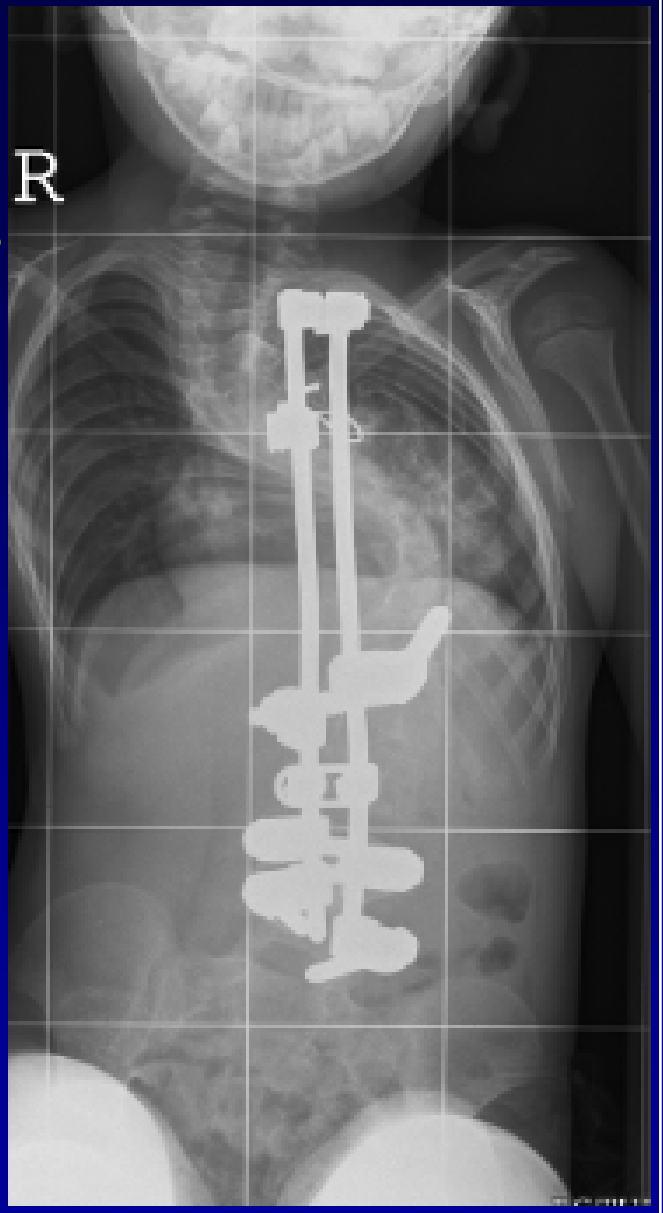
- Better able to maintain distraction, deformity control without involving dysplastic lower spine
- Control of spine without instrumentation of spine
- Tandem rib connectors useful.
  - Distribution of force
  - Resistant to pull out

GR or VEPTR in Infection

- Infection – VEPTR or GR?
  - Either is a good rescue for infection in the other!
  - Soft tissues a critical component
- Implant removal/retention with deep infection?
  - Depends upon
    - Extent
    - Duration after index procedure
    - Skin, soft tissues

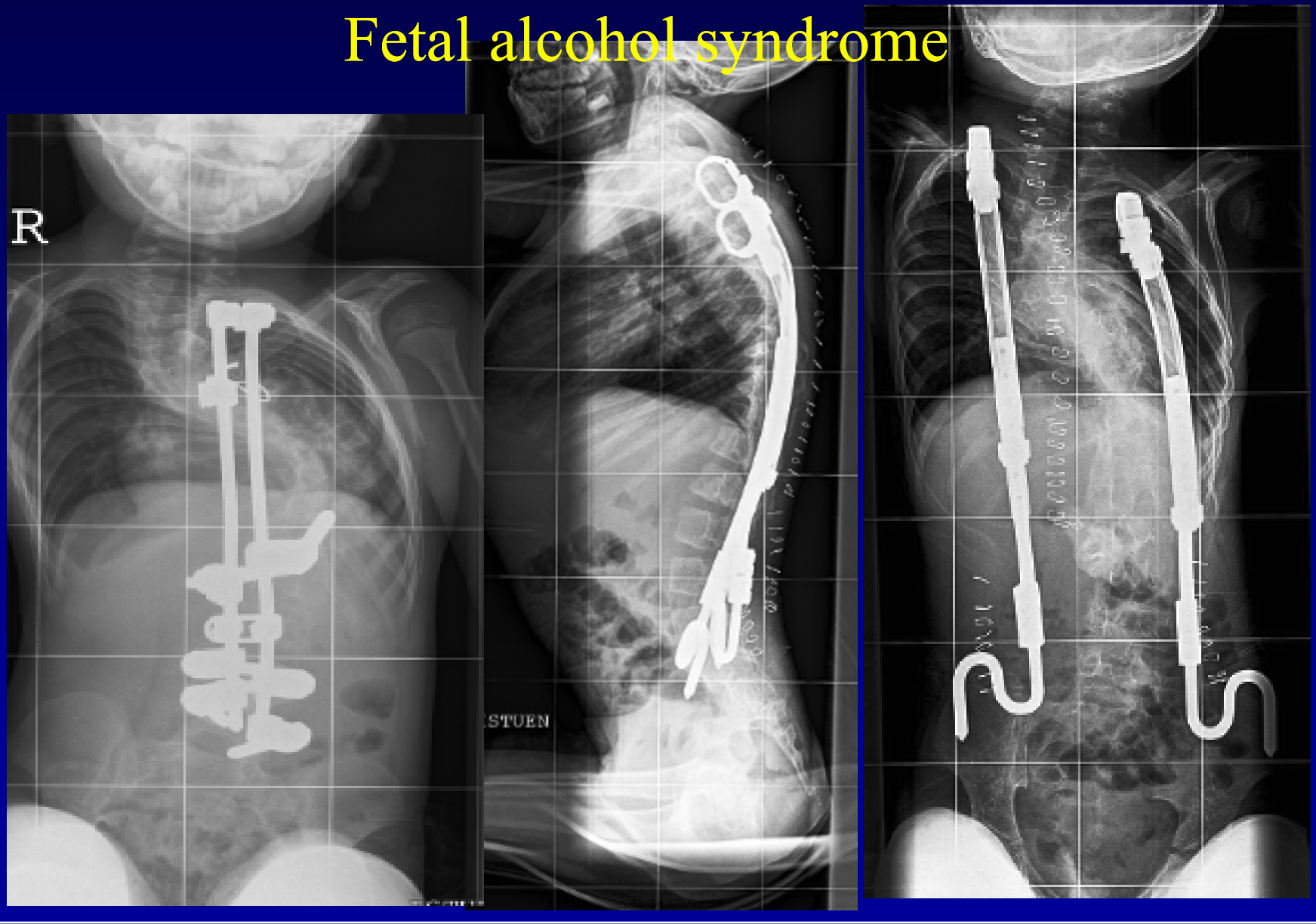
GR or VEPTR in Infection
Fetal alcohol syndrome Complex
congenital vertebral anatomy - age 6





GR or VEPTR in Infection
Fetal alcohol syndrome – long segment of spine

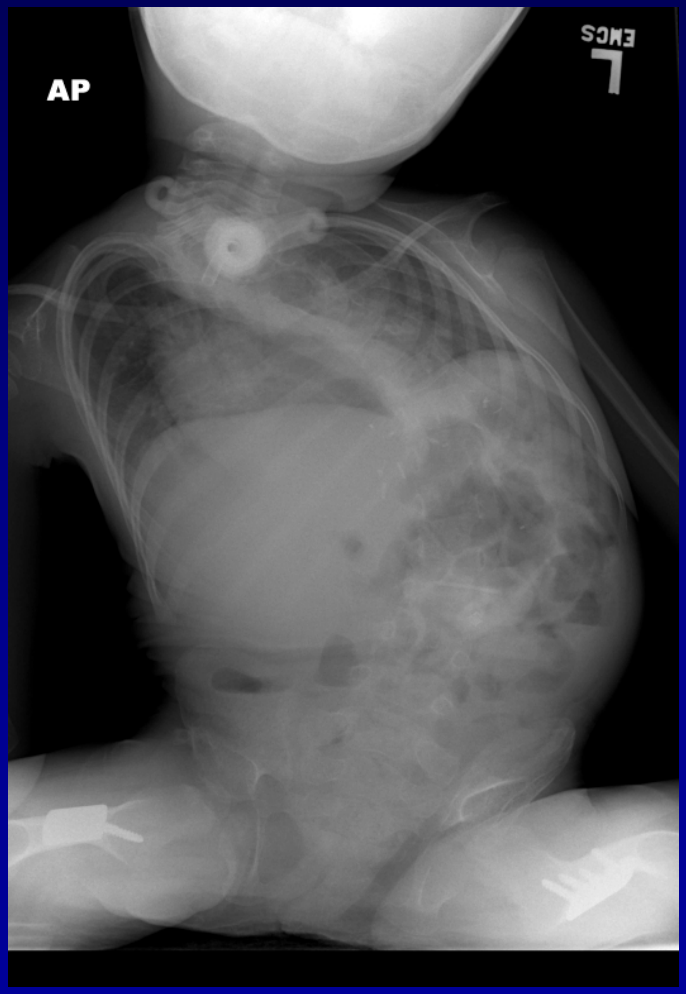
GR or VEPTR in Infection
Fetal alcohol syndrome

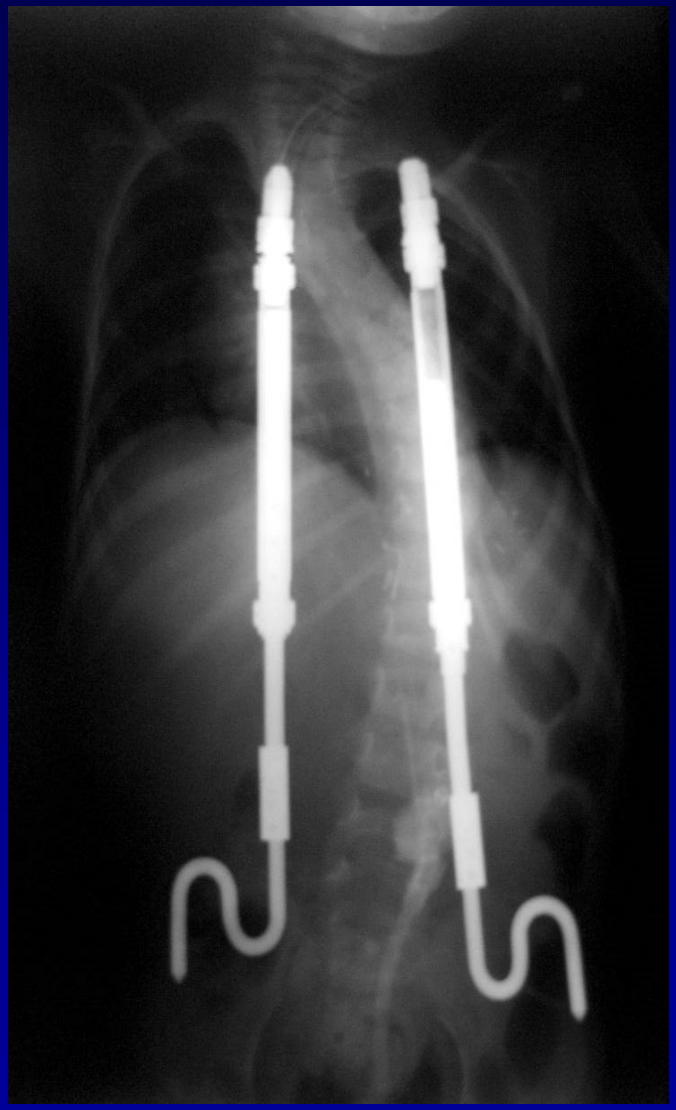


GR or VEPTR? – *Upper thoracic kyphosis*

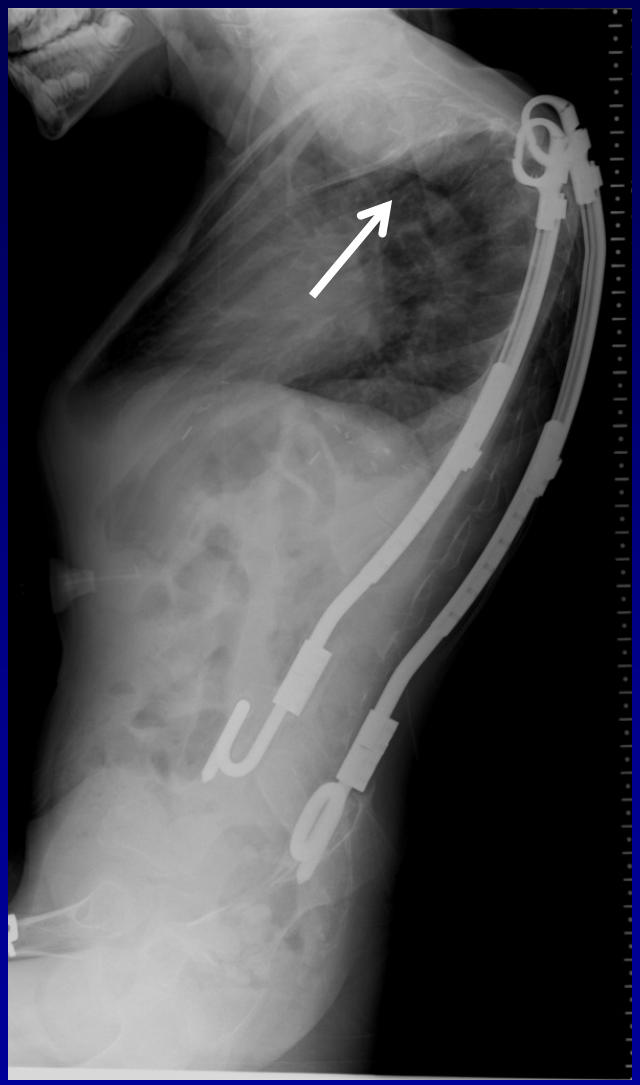
- *Kyphosis (*upper thoracic) problematic for both growing rods and VEPTR
  - Multiple factors:
    - Weak paraspinals
    - Junctional stresses above stiff segment
    - GR - Disruption of posterior elements.
    - VEPTR – lack of direct sagittal spine control
    - *Both GR and VEPTR – distraction-based*

GR or VEPTR> – collapsing deformity, arthrogryposis age 4. Coronal deformity controlled





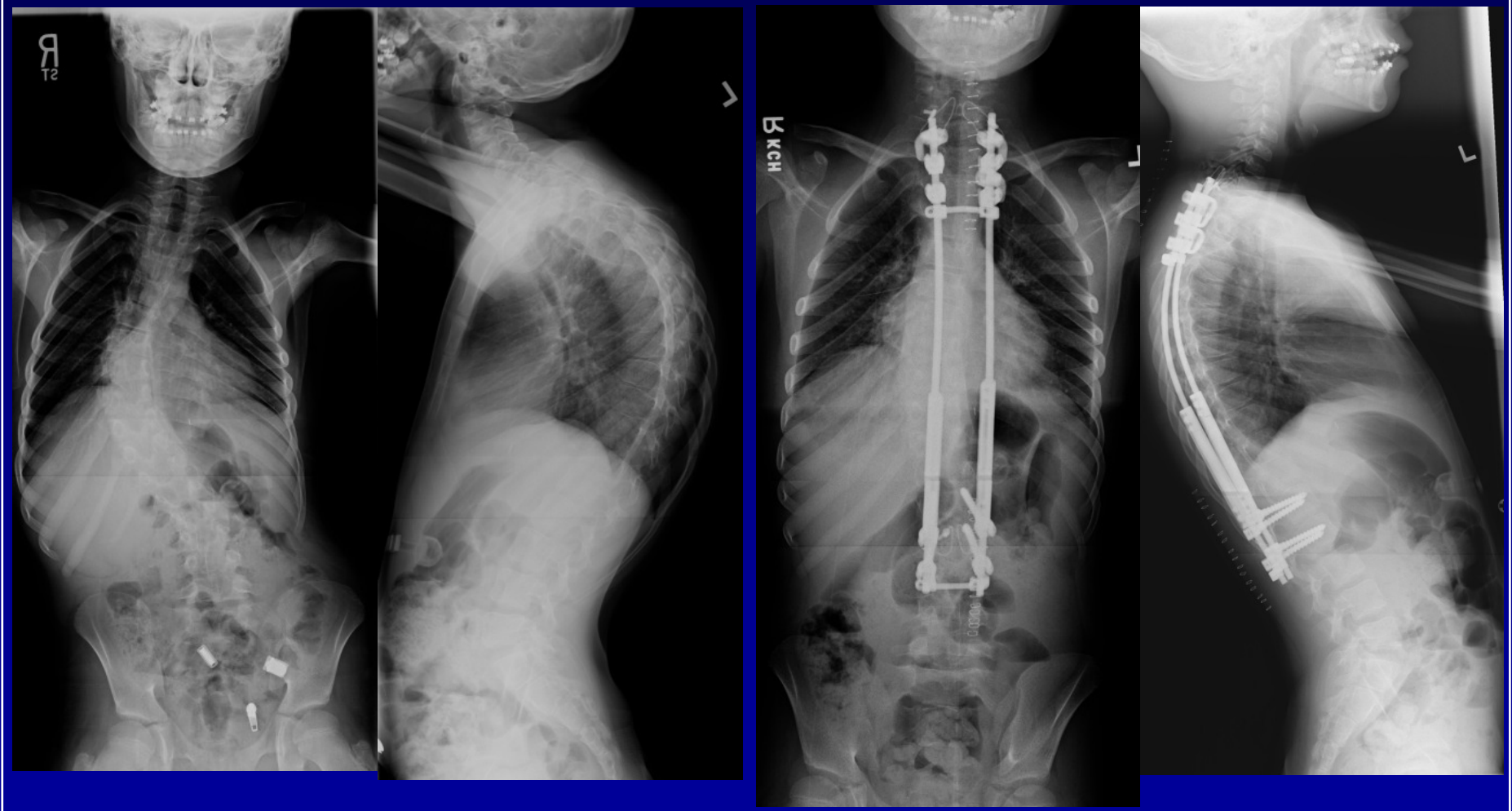
GR or VEPTR? arthrogryposis – age 4 to 8 –initial control then progressive PJK



VEPTR in arthrogryposis – poor control proximal kyphosis.

- Cervico thoracic junction collapsed further into kyphosis, rotating around VEPTR attachments
- Distraction based systems poor for upper thoracic kyphosis
- GR better than VEPTR?
  - Can extend more proximally with GR

GR or VEPTR for upper thoracic kyphosis?
7 yo with familial dysautonomia



GR or VEPTR for upper thoracic kyphosis? with familial dysautonomia age 7, 8, 16

GR or VEPTR for upper thoracic kyphosis - neither is perfect - GR has some advantages

- Strategies:
  - Leave some kyphosis
  - Pre-op halo gravity traction may facilitate device insertion by diminishing kyphosis
    - Tendency to reoccur
  - Growing rods can extend more cephalad than VEPTR - ? Past the kyphosis?
- GR preferable:
  - More cephalad extent possible
  - More contouring options
  - Direct control of spine

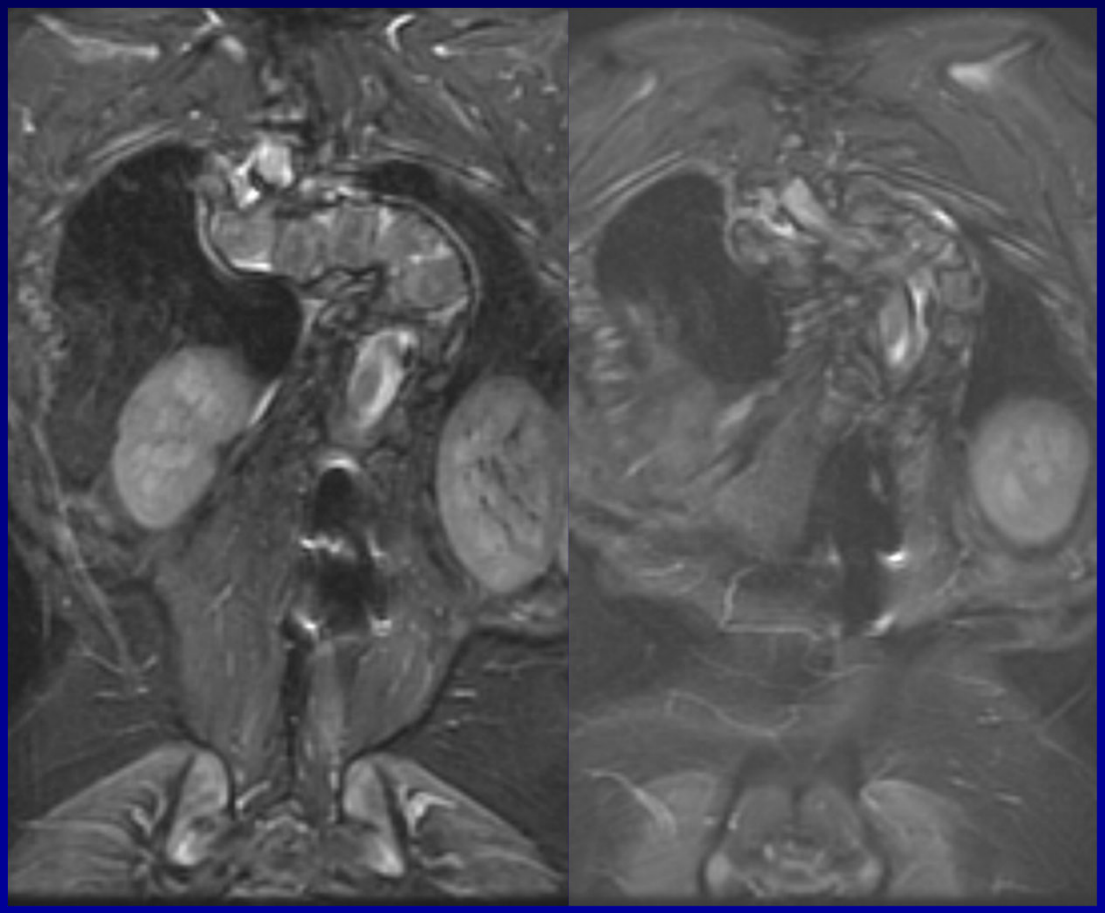
GR or VEPTR - Bone dysplasias

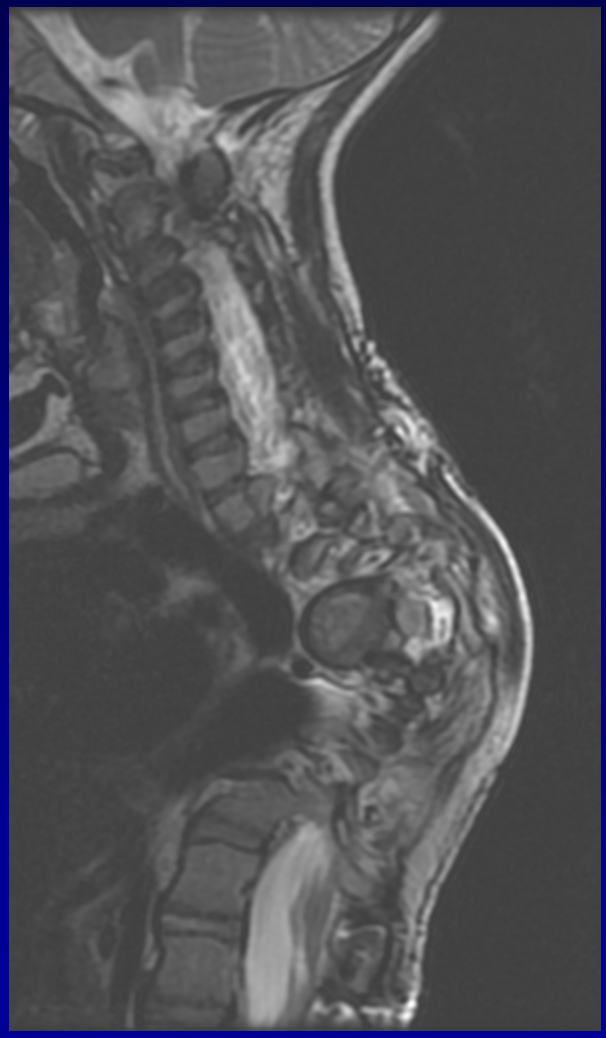
- Bone dysplasias
  - Spine or ribs better bone for anchor points?
    - Small soft vertebra? – VEPTR may be preferable
    - Spinal stenosis or hypoplastic pedicles? Prior laminectomy? VEPTR may have advantage
  - Beware waiting too long to establish contol over curves in spinal stenosis – neurologic risk with progression, correction.

Spondyloepiphyseal dysplasia

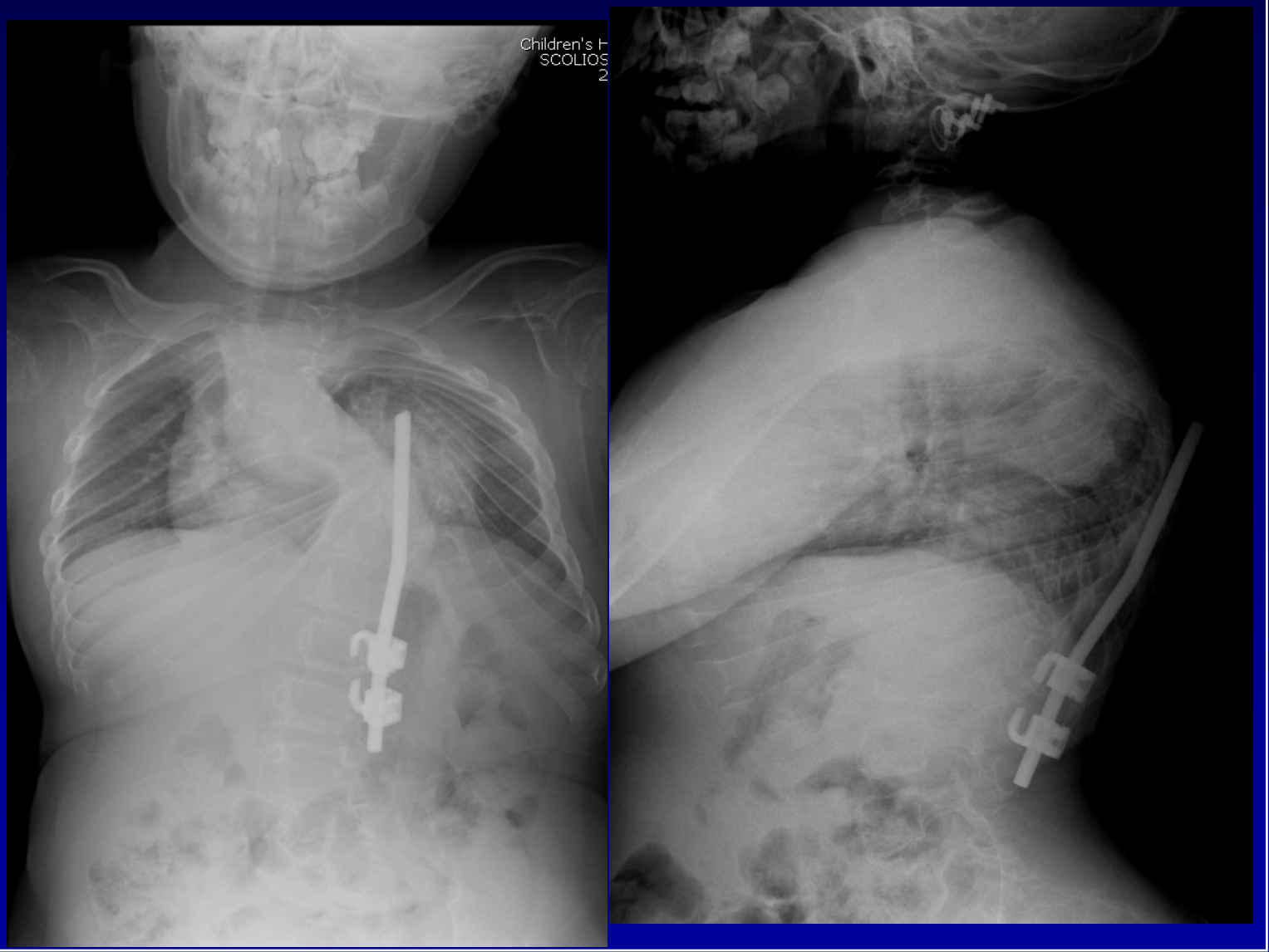
- Age 14 mos.
  - C1-C2 fusion
  - Growing rod for scoliosis, kyphosis
- Age 30 months – proximal disengagement
- Age 10 – paraplegia following hip osteotomies, epidural post-op. Slow resolution

SED

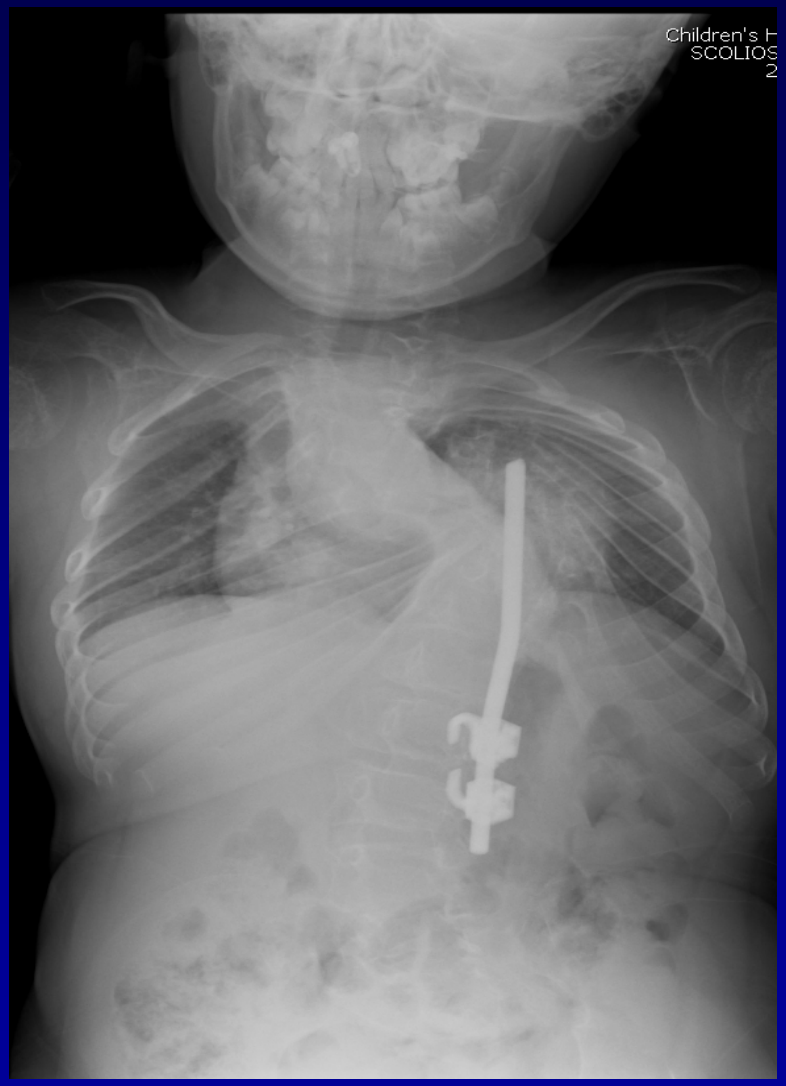


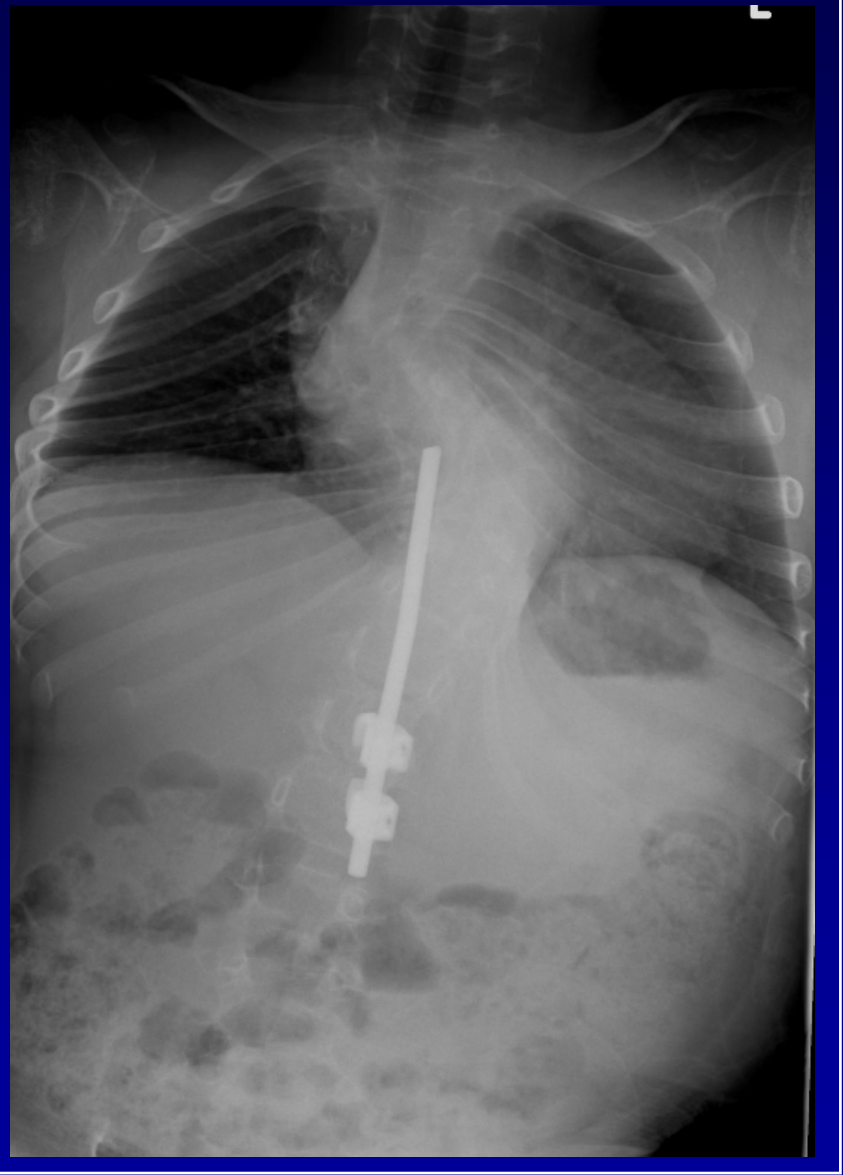


Pre - traction



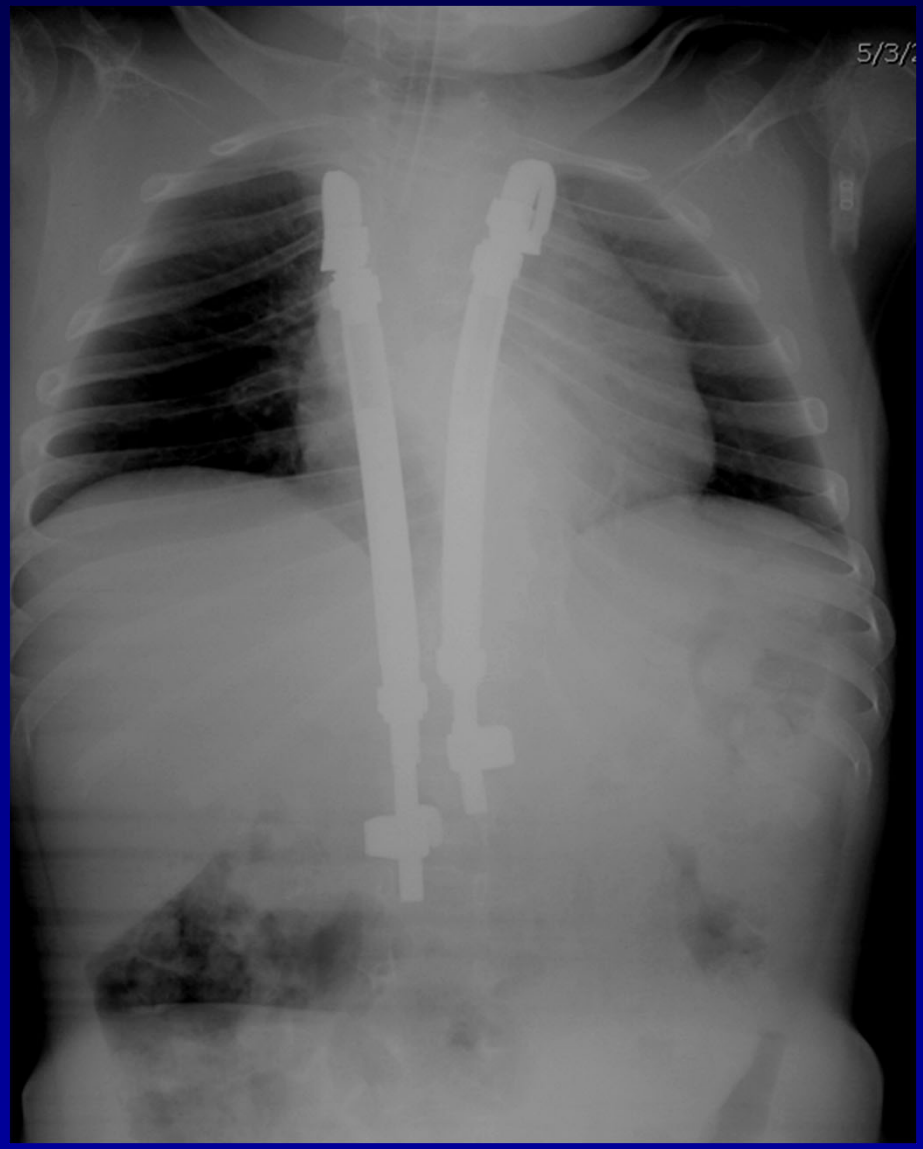
Halo gravity for 7 weeks





Post VEPTR

- Kyphosis well controlled
- One rib sleeve bent to accommodate kyphosis

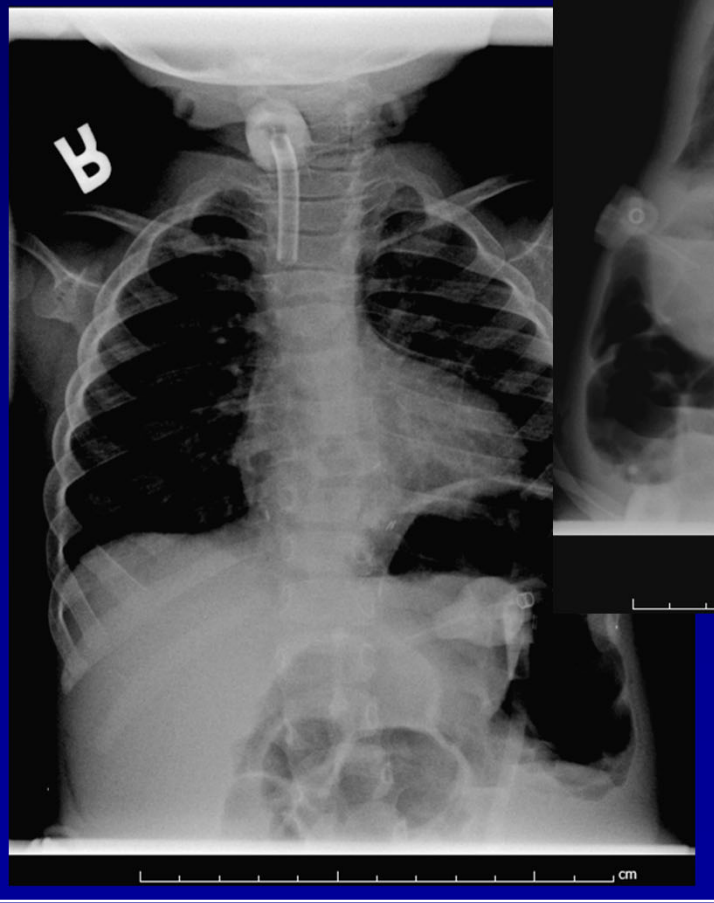


Early Onset Deformity. – etiology as a factor:

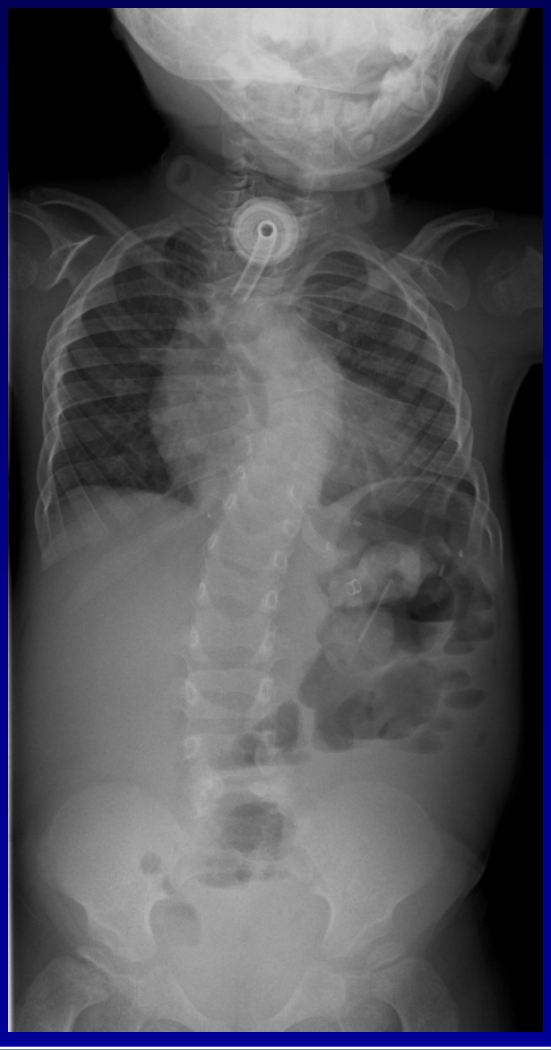
- Bone dysplasias:
  - If upper thoracic *kyphosis*, GR has an advantage over VEPTR
    - GR can be extended as far cephalad as needed

Campomelic Dysplasia

- Vent dependent
- Rapid progression after 6 months
- Age 18 months – minimal deformity



Age 30 months – severe kyphosis



Age 30 months – severe kyphosis

- Age 30 months – severe kyphosis – stiff
- ***GR or VEPTR?***

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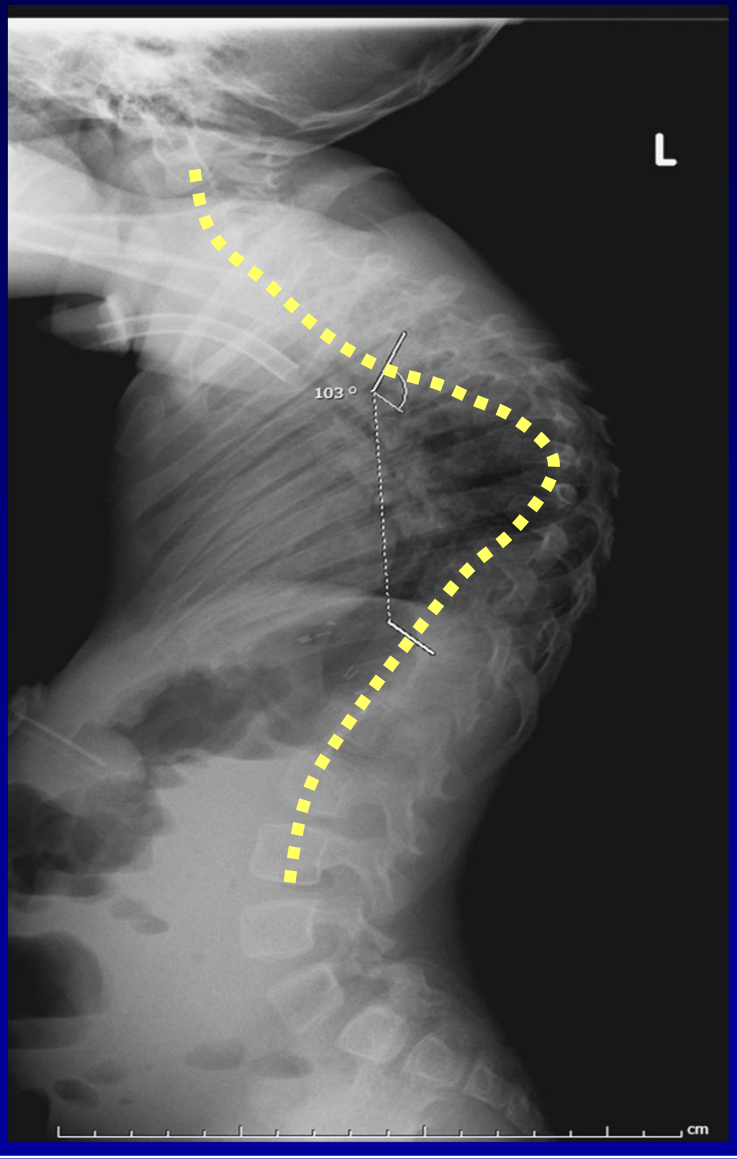
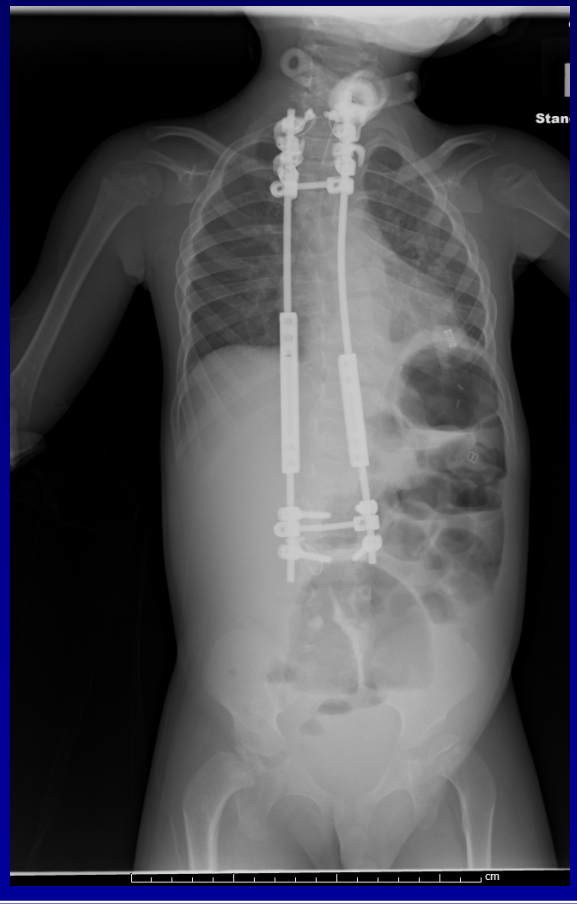
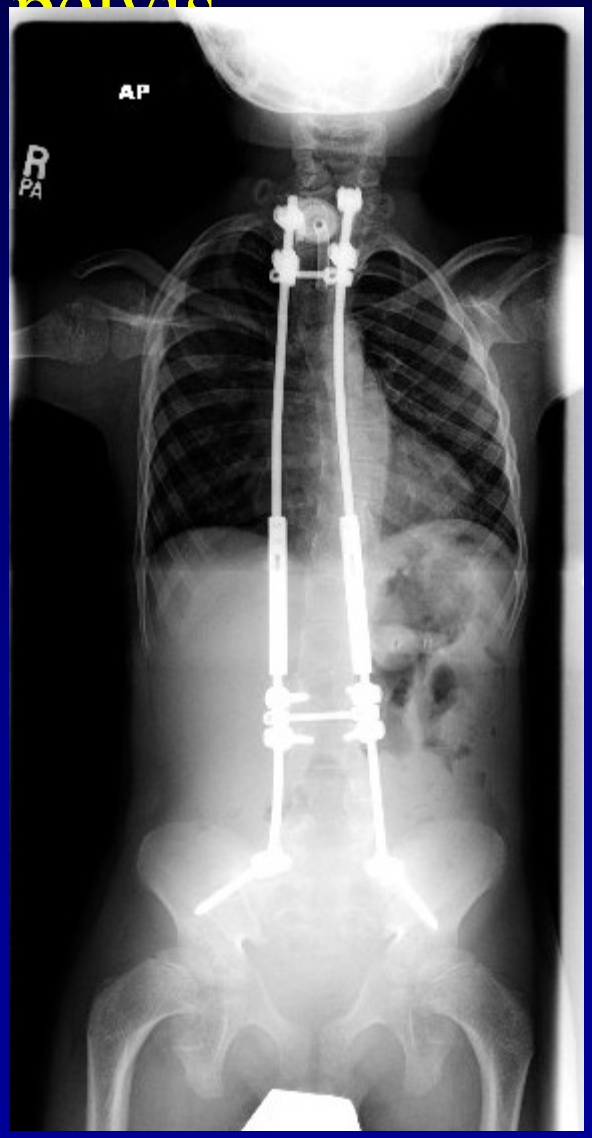
- ***GR enables more cephalad purchase***



Age 34 months – growing rods, extending to C7 with sublaminar cable at C7



Age 7 symptomatic spondylolisthesis required extension to pelvis

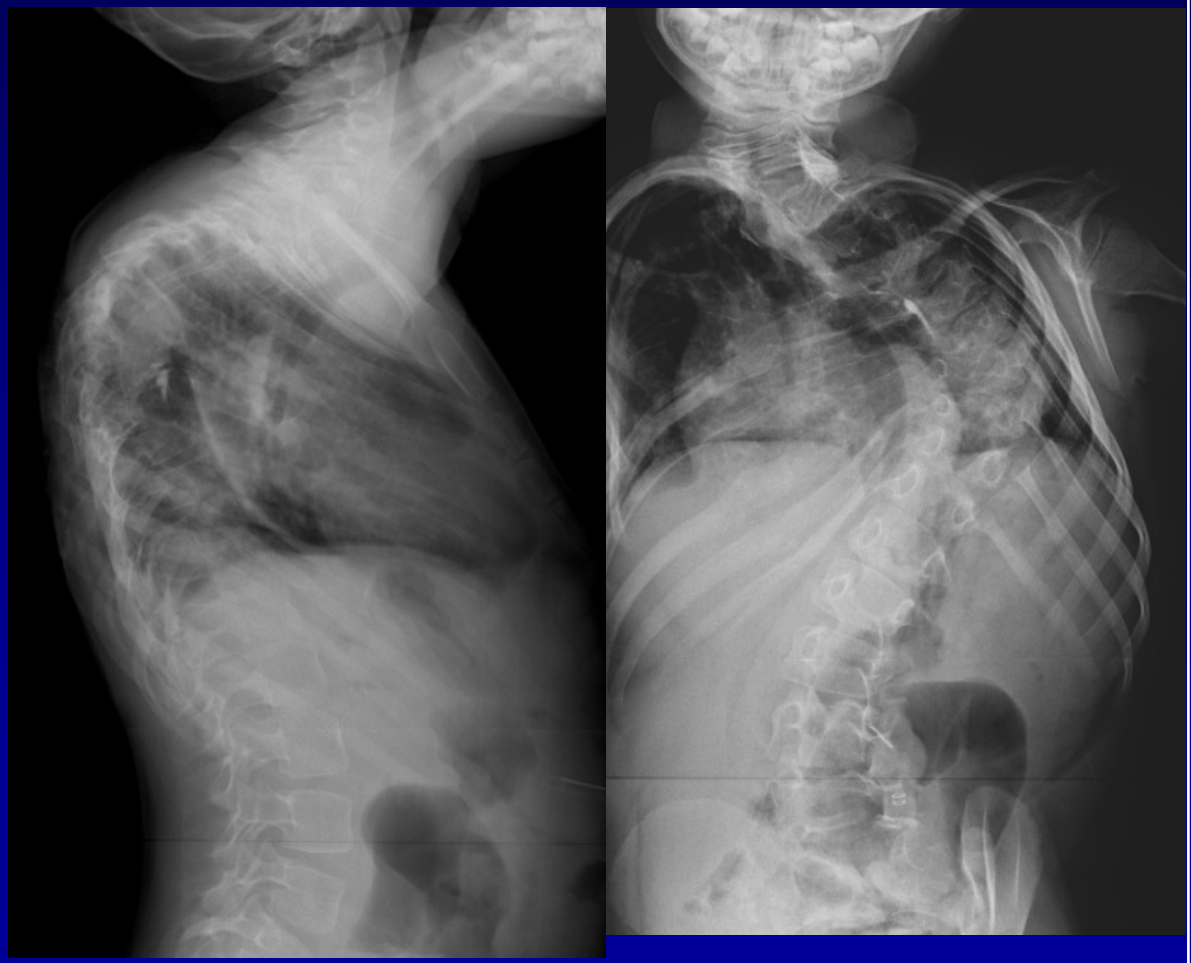


GR or VEPTR – Osteopenia in Early Onset Deformity

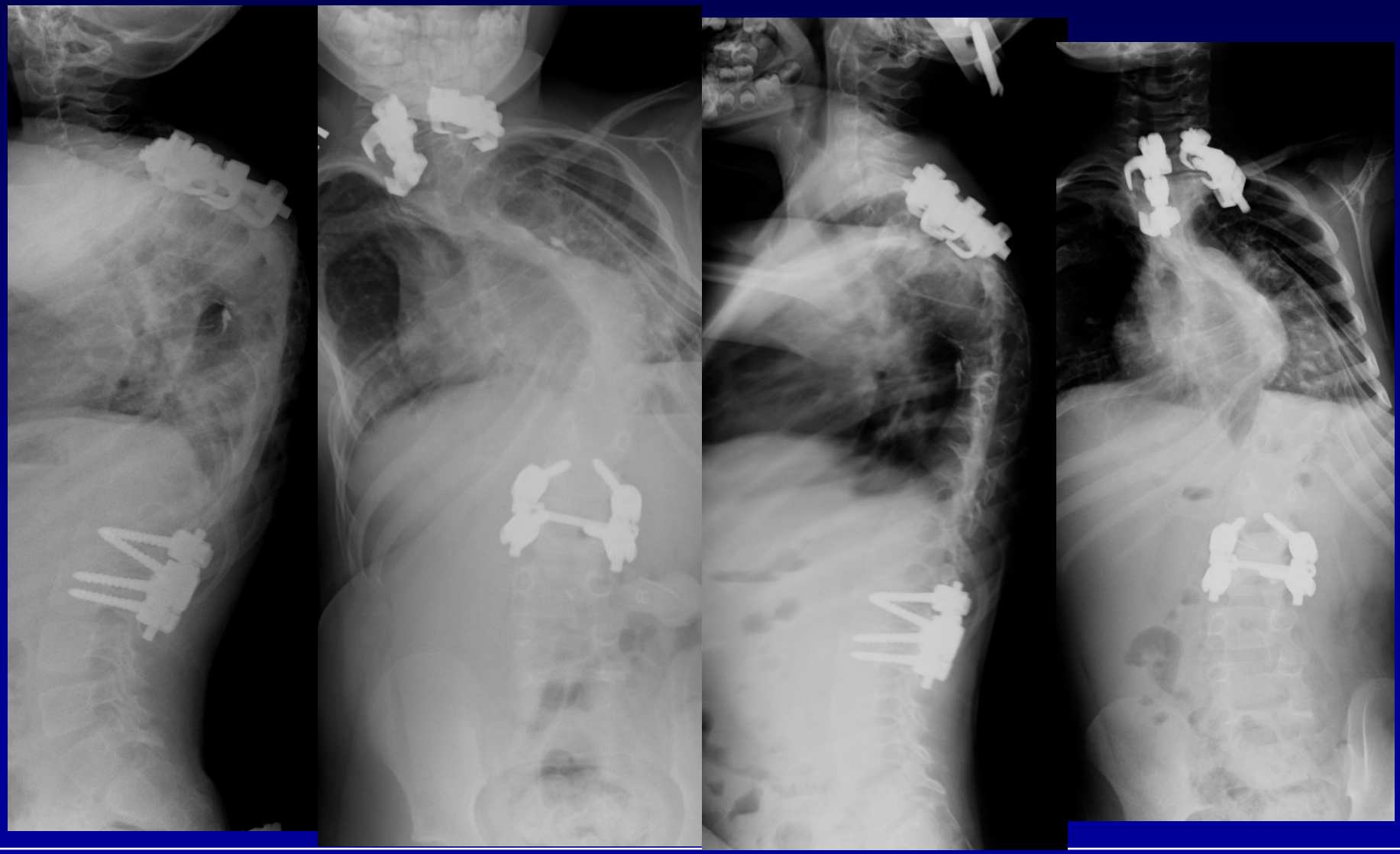
- Osteopenia, poor bone quality
  - VEPTR may have an advantage?
    - ?more 'give' in ribs than spine?
    - Experience with osteogenesis imperfecta?
  - *GR allows staged anchor placement*

VEPTR or GR? Osteopenia and kyphosis

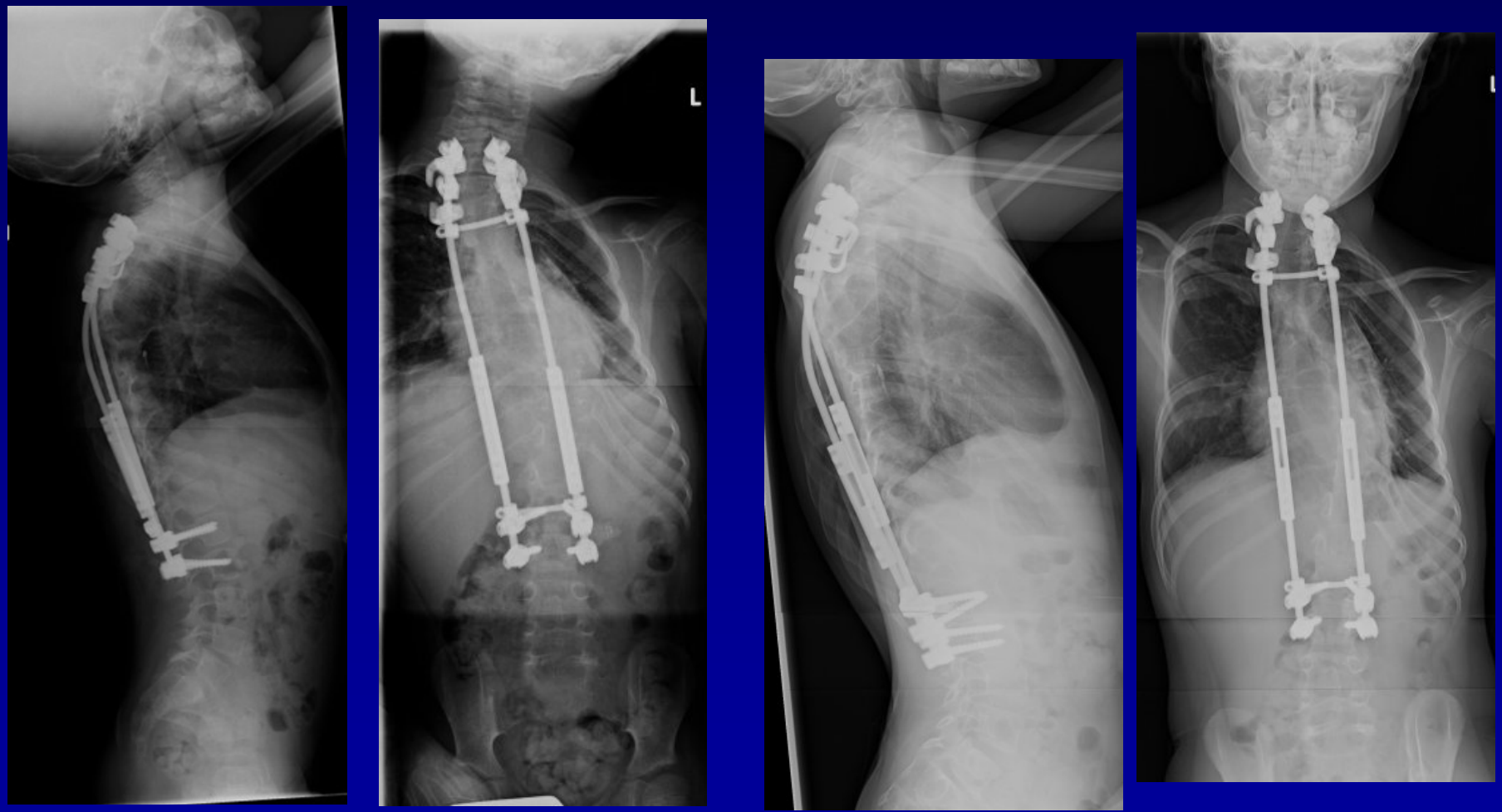
- 5 yo with recurrent TEF
- Failed VEPTR with severe osteopenia
- Progressive scoliosis
- Rigid upper thoracic kyphosis



VEPTR or GR? Osteopenia and kyphosis: Insert anchors, 2mos, apply H-G tx, 1 1/2 mos

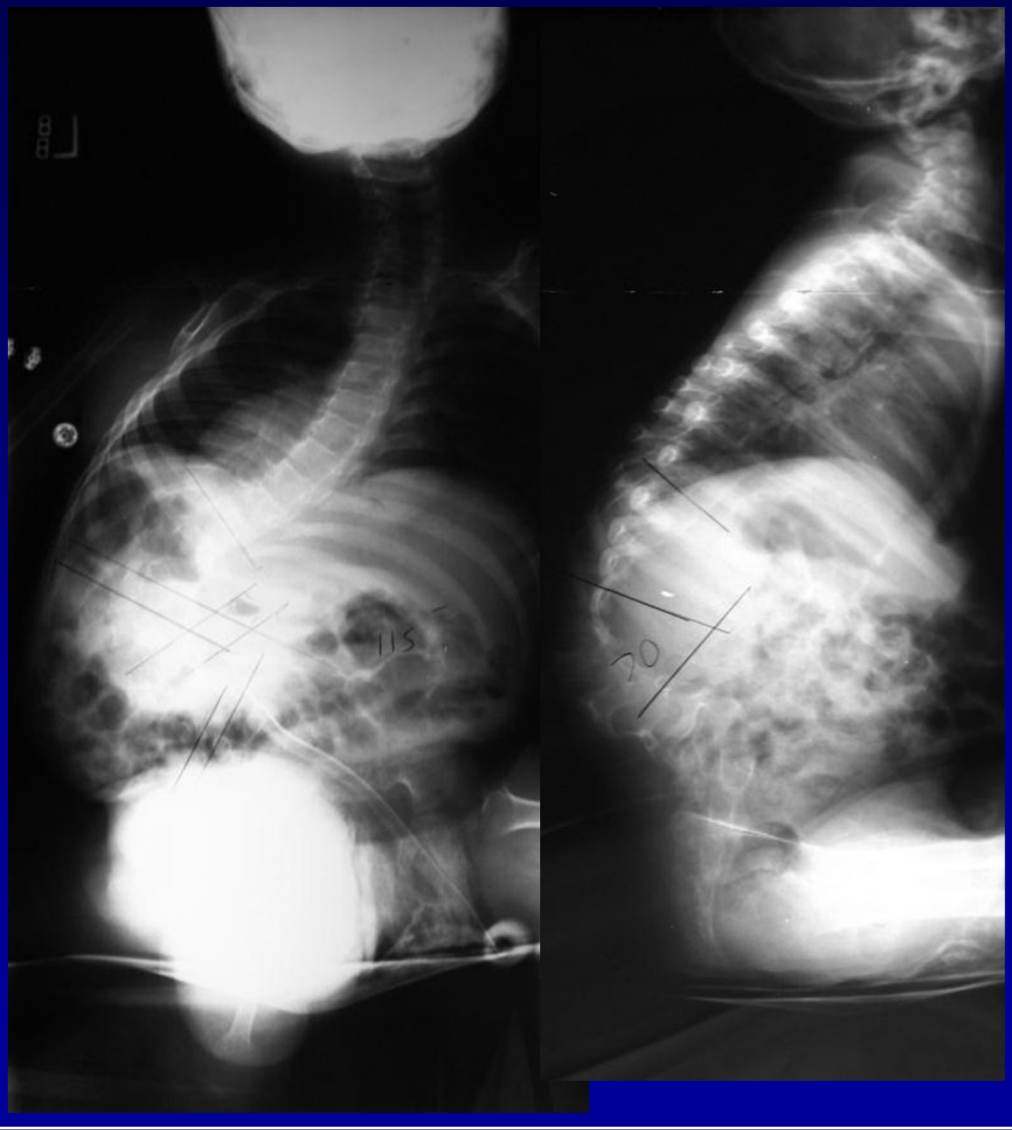


VEPTR or GR? Osteopenia and kyphosis Age 6, 8

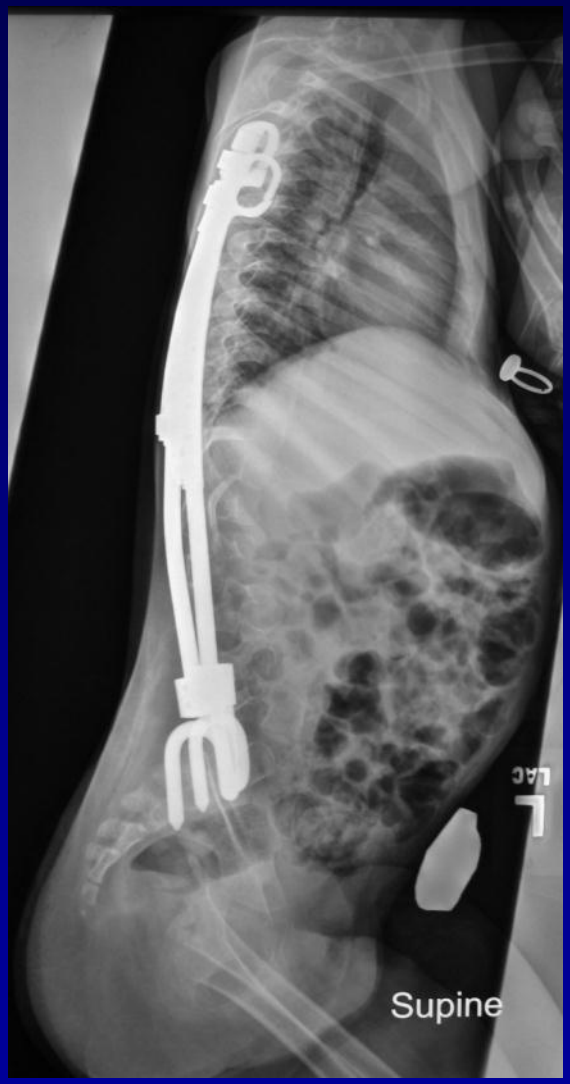


4 y.o. with Ehlers Danlos variant

- Worsening deformity
- Recurrent breakdown when attempting to sit or with a brace
- Severe osteopenia, recurrent fractures
- Increasing respiratory distress (secondary TIS)



Ehlers Danlos

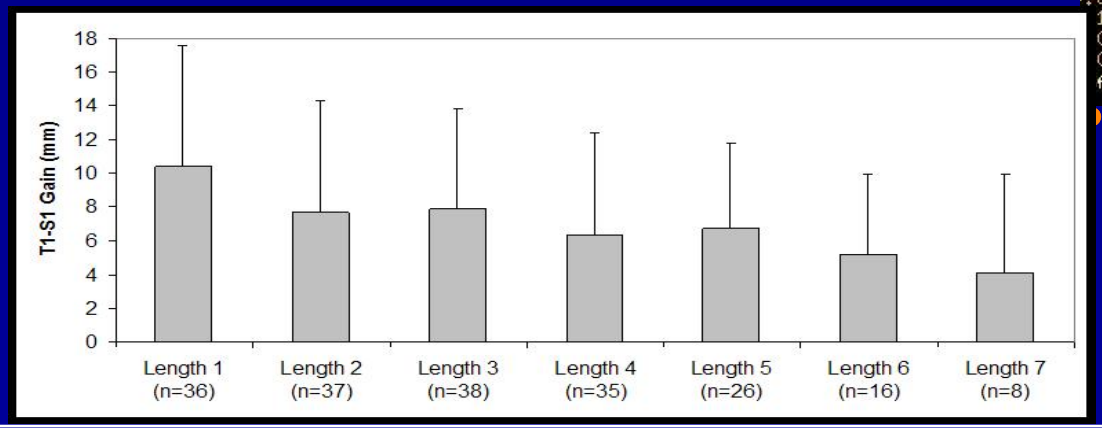


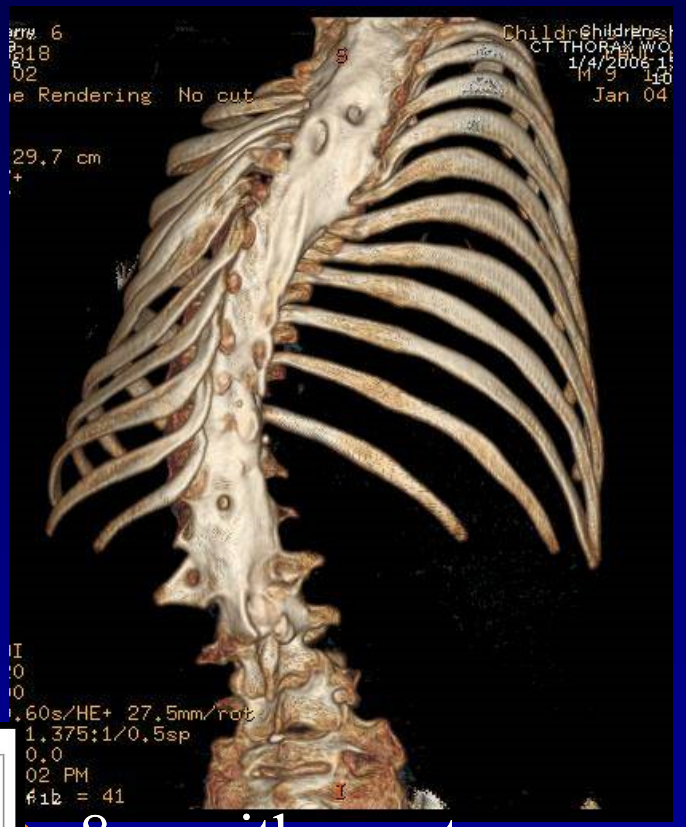
GR or VEPTR – Complications?

- Routine problems are different:
  - GR – rod fractures
  - VEPTR – rib anchor drift
- Both:
  - Infection
  - Scarring
  - Repetitive surgeries
    - Anesthetics
    - Psychological stress
- Serious or growth ending complications:
  - VEPTR – chest wall scarring, fusion
  - GR – spontaneous fusion
    - (VEPTR also?)

GR or VEPTR - complications

- *GR – growth stopping complication*
  - Lengthening may not be possible indefinitely
- Skaggs and GSSG data:

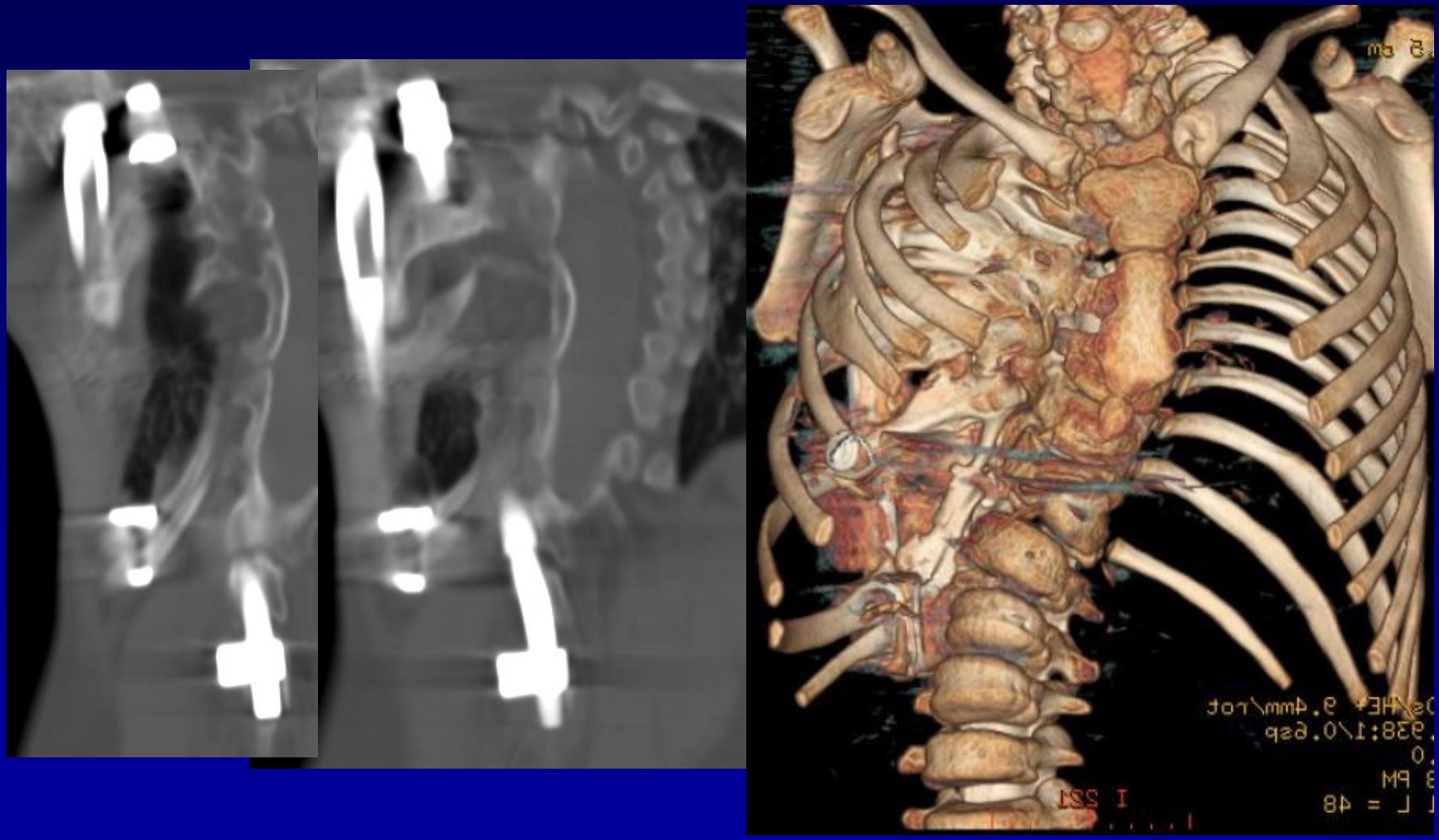




8yo with spontaneous posterior fusion after growing rods started at age 2

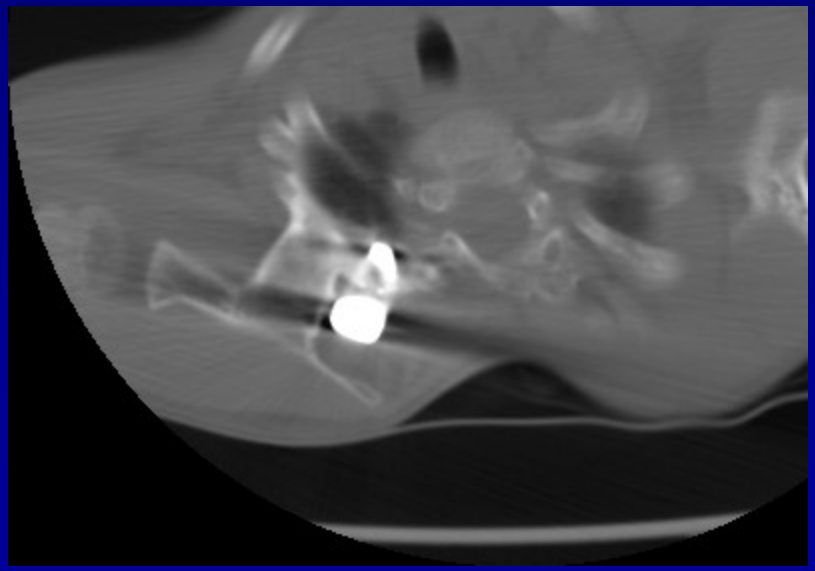
GR or VEPTR – complications

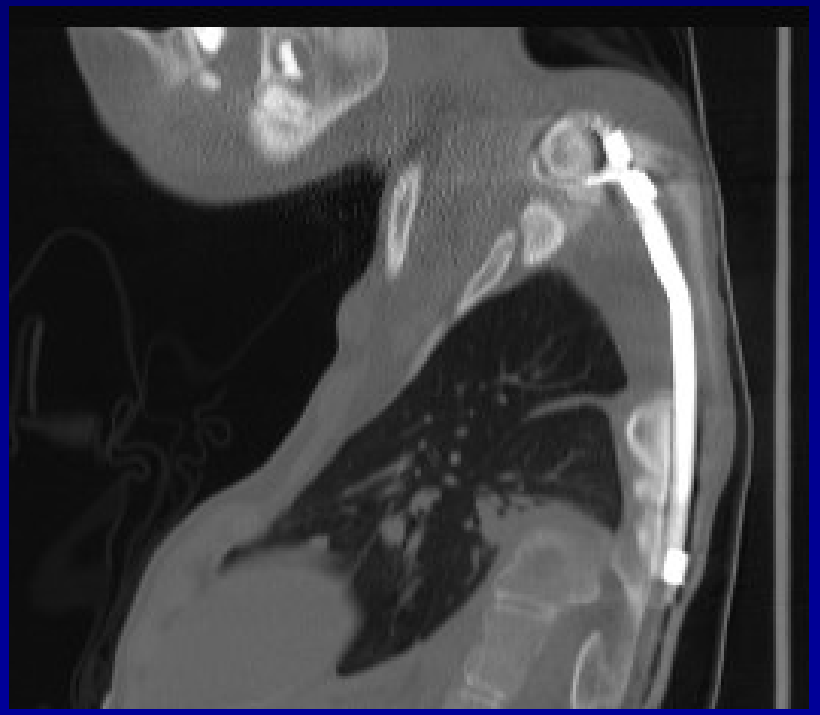
Recurrent rib fusion after thoracostomy



GR or VEPTR - complications
Inadvertent fusion of scapula to ribs:

- 2 patients with solid fusion of scapula to ribs
- Numerous with limited scapulothoracic function





GR or VEPTR - Final fusion easier?

- GR and VEPTR:
  - Spontaneous stabilization may have occurred by end of growth
    - *No fusion* may be needed if:
      - Modest deformity
      - Implants not troublesome
      - Likely underlying fusion
- Conversion to final instrumented fusion?
  - GR challenging conversion
    - Scar, spontaneous fusions, distorted anatomy
  - VEPTR easier conversion?
    - Spine "untouched"??
      - Spine spontaneously fused, esp lower spine anchor area
    - Rib fusions beneath device

VEPTR or Growing Rods?
How to choose between GR and VEPTR:

	Growing Rods	VEPTR
Spine growth preservation	+	+
Chest deformity correction	+/-	++
Ease of use, familiarity	+	-
Multiple ops	√	√
Final fusion needed	+/-	+/-
Complication which limits distraction	Fusion underneath rod	Chest wall stiffness, rib re-fusions
Solution to complication?	Early fusion	Repeat thoracostomy
Common device problems, failures	Rods break	Rib attachments drift
Upper thoracic kyphosis	Better	Poor
Osteopenia	Poor	Poor

Strong indications for VEPTR:

- Primary Chest wall problem
  - Massive rib fusions
  - Thoracogenic scoliosis
- Failed Growing rods
  - Infected spine anchors
- Poor spine anchors
  - Bone dysplasia with spinal stenosis
  - Spina bifida

Strong indications for growing rods

- Primary spine deformity with lesser, flexible chest deformity
- Normally segmented, unscarred chest wall
- High thoracic kyphosis
  - (GR/local fusion can extend into the cervical spine if needed)