Congenital Scoliosis: Selection and Timing of Surgery

Sorting through the options

John B. Emans, MD

Children's Hospital
Harvard Medical School
Boston, MA 02115

Disclosures:

- *Conflicts of Interest* :
  - Consultant:
    - Medtronics spine
    - Synthes spine
      - Royalties from VEPTR II
- *Off-Label devices discussed:*
  - Pedicle screws in children and *growing rods are off-label.*
  - VEPTR is on-label but requires HDE

Congenital Spinal Deformities:
A spectrum of deformity, natural history and *diverse clinical significance:*

Deformity

Respiratory function

Neurologic compromise

Shift in available treatments for congenital deformity:

- *Old: early in situ fusion* for progressive (or predicted progressive) deformity
  - 'short and straight' better than severe deformity
- *Contemporary: multiple newer surgical options*:
  - *Growth awareness, TIS*
  - *Growth-sparing treatments*
    - Growing rods, VEPTR
  - *Safer radical deformity correction*
    - Posterior only Hemi excision, VCR

How to sort through the options for congenital spine deformity?

- *Analyze* the deformity
  - Understand the *natural history*
  - Understand the *consequences* of treatment
- Keep *treatment goals* in mind
- Decide:
  - *What* treatment
  - *When* to initiate treatment (for me this is the hardest)

How to sort through the options for congenital spine deformity?

- Understand the *growth remaining*:
  - What are the effects on *spine and chest growth* of
    - *Untreated* deformity
    - *Treatment* (and its side effects)
      - Definitive fusion
      - Local fusion

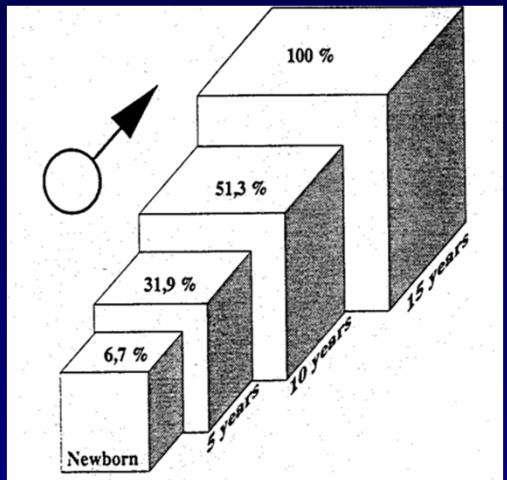


FIG. 15. Boys: Diagram of thoracic volume and its evolution expressed as a percentage.

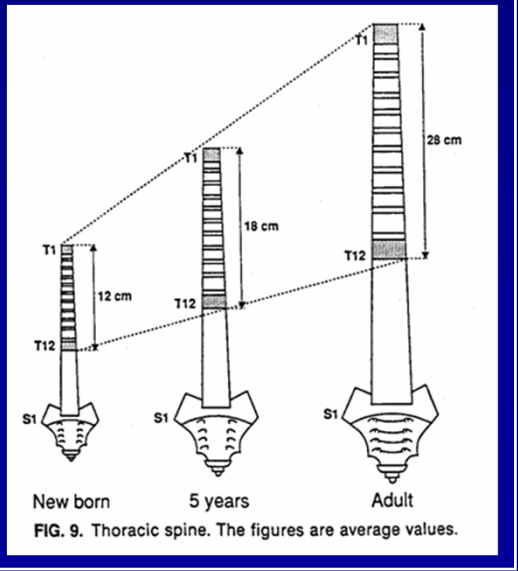
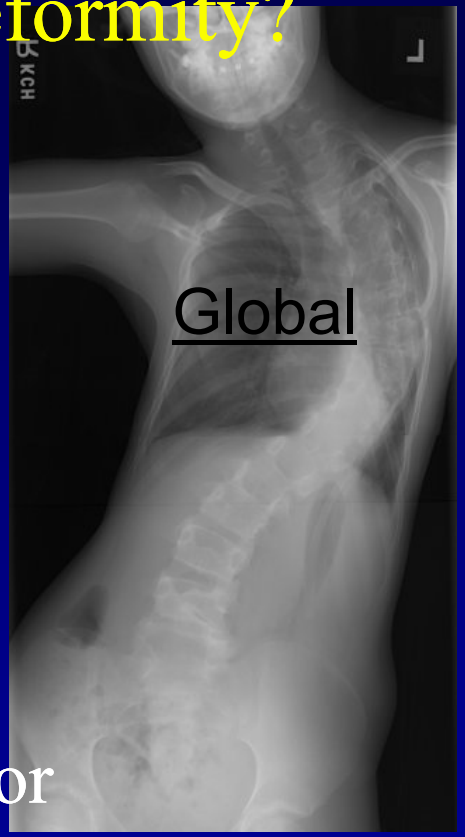
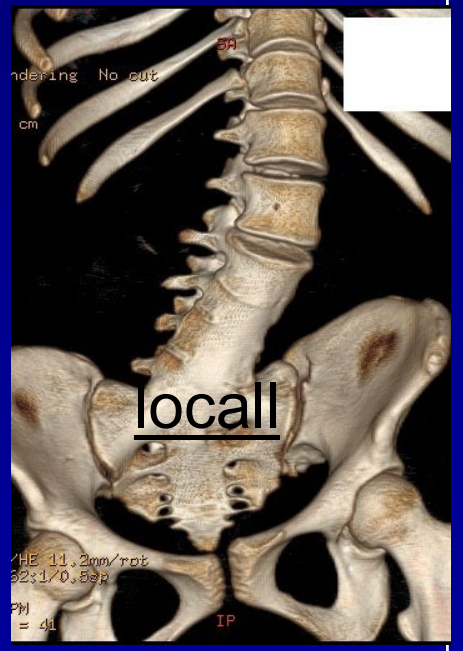


FIG. 9. Thoracic spine. The figures are average values.

How to sort through the options for congenital spine deformity?

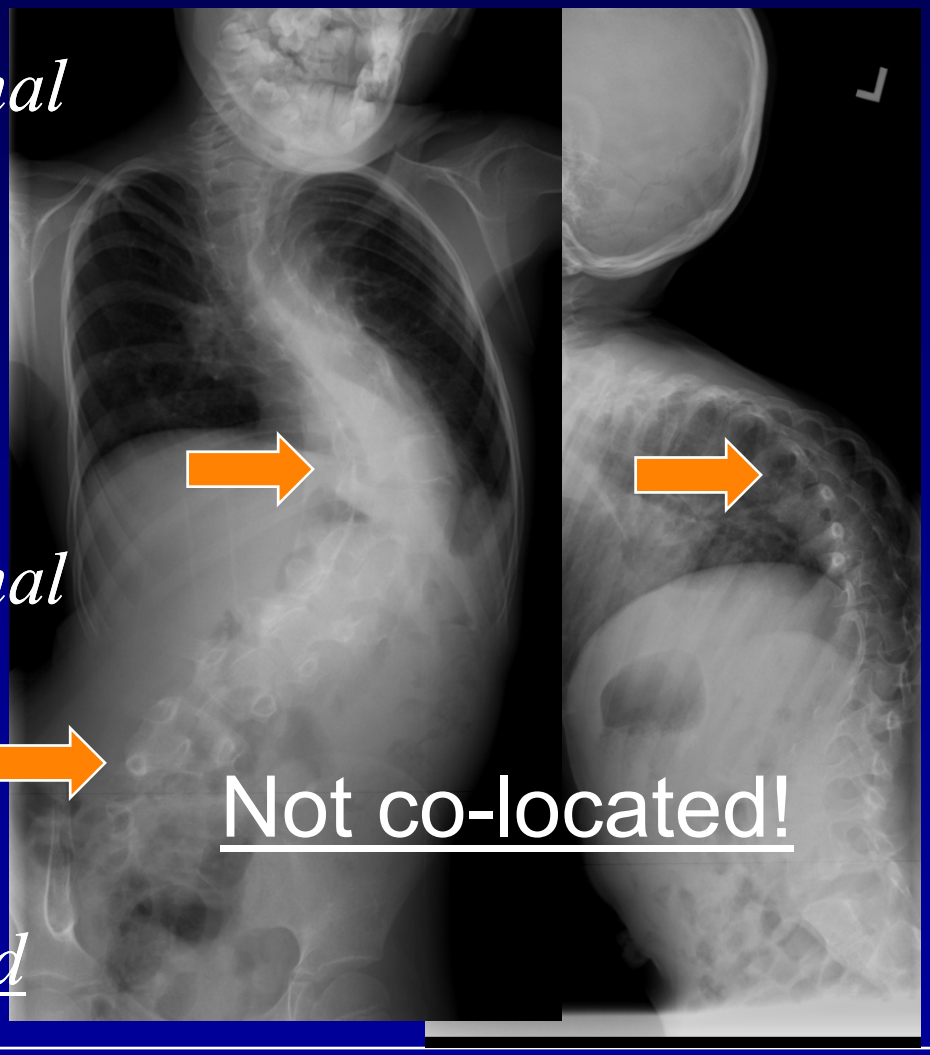
- *Analyze* the deformity
  - Progressive?
  - Secondary curves?
  - Effect on thorax?
- Attempt to *classify*:
  - *Global* deformity (involves large section or all of spine)
  - *Local* deformity (involves short section of spine)





How to sort through the options for congenital spine deformity?

- Are the *sagittal* and *coronal* deformities located in the *same region* of the spine?
  - Kyphosis/Lordosis/ Scoliosis *co-located?*
- Are the *sagittal* and *coronal* deformities in *different locations* on the spine?
  - Kyphosis/Lordosis / Scoliosis *not co-located*



Older Children – growth less of a concern

Congenital Spine Deformity in *older children approaching maturity:*

- A *pure deformity decision* (no growth sparing needed!)
- Contemporary options:
  - In situ (instrumented) fusion
  - Correction, fusion, instrumentation
    - Traction? Anterior release?
  - *Osteotomies*
    - Particularly helpful for congenital deformities
      - Wedge resection
      - Vertebral Column Resection
  - *Combinations- think out of the box*
- *How to decide*?

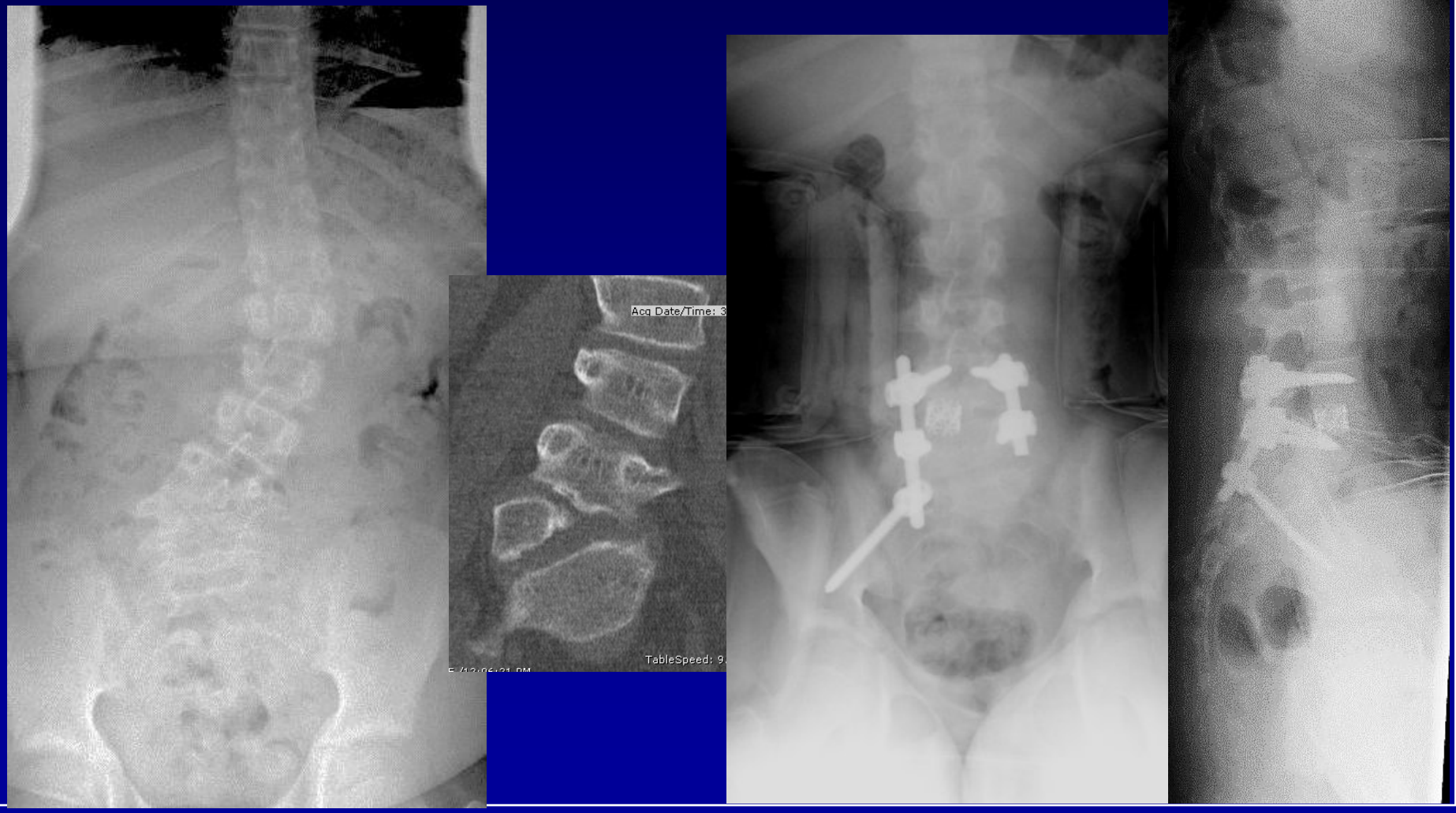
How to sort through the options:

Osteotomy needed for correction

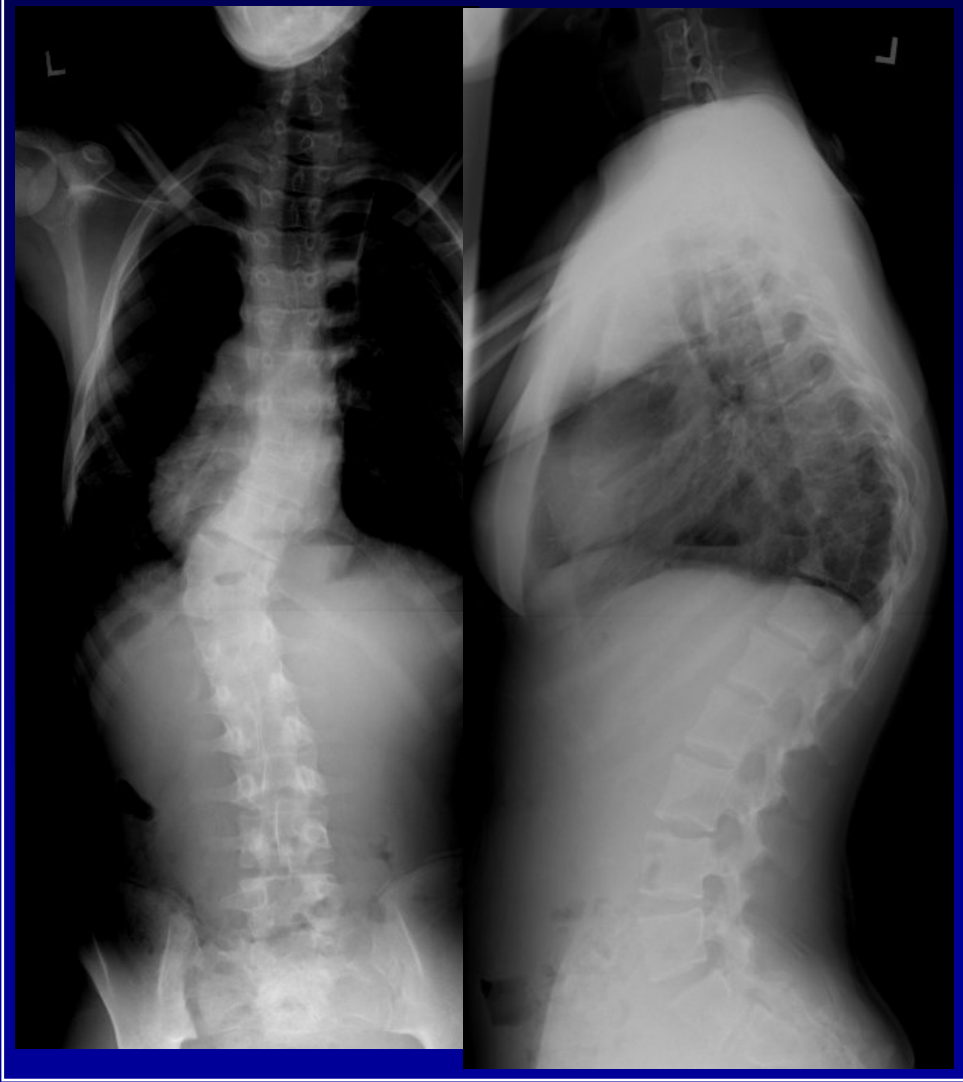
Which osteotomy is the correct choice?

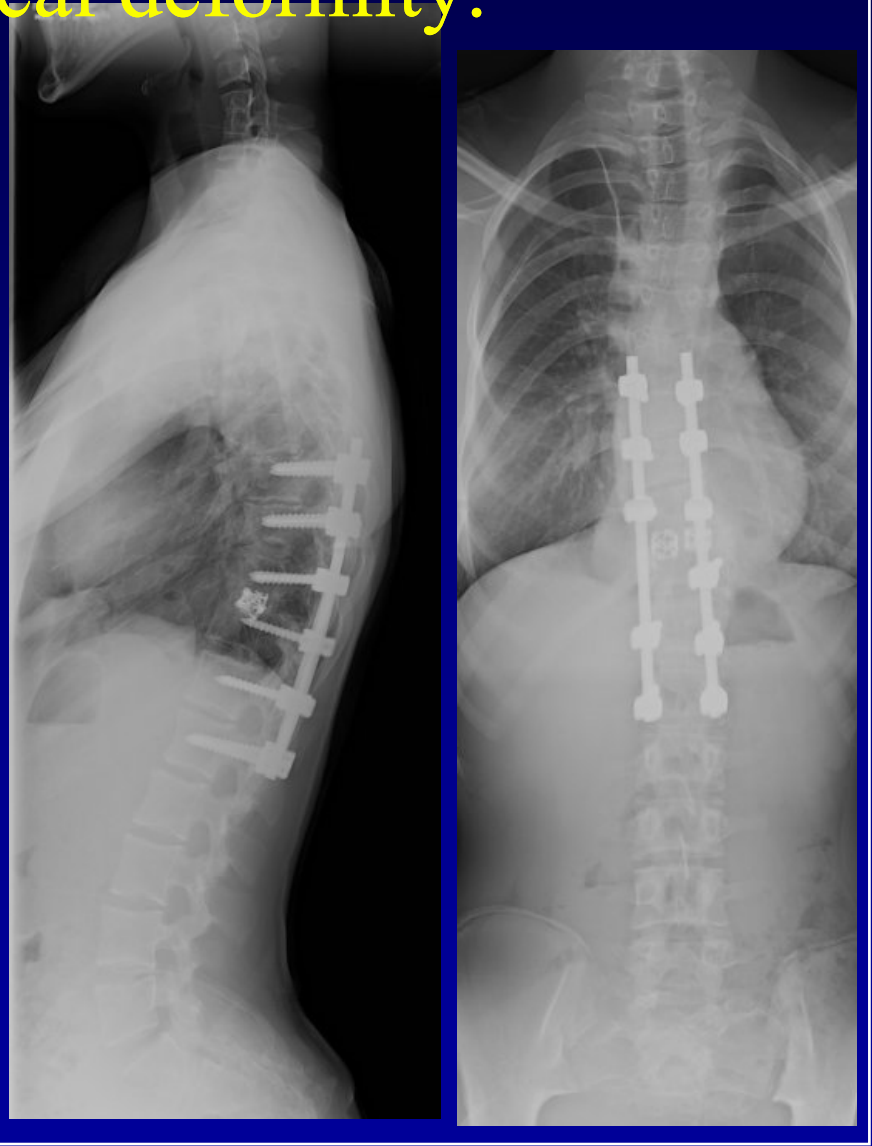
Deformity:	Multiple releases (include SPO's)	VCR, Wedge Resection
Global	+	
Local		+
Sagittal and coronal: *co-located*	+	+
Sagittal and coronal: *separate*	+	

Postero lateral L/S anomaly with secondary curve. Age 8. *Local deformity. Co-located scoli and kypho. Rx:* Wedge resection/ VCR

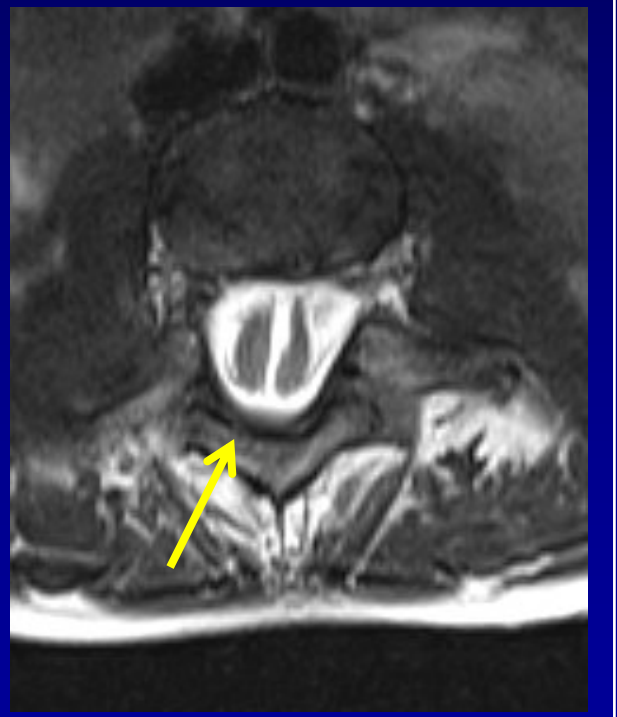


VACTERL age 14. *Kyphosis and Scoliosis co-located* *Rx:* -VCR for local deformity.



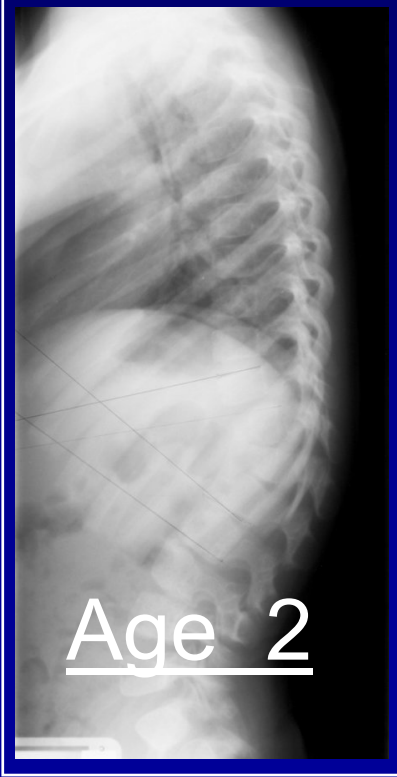


Congenital Kyphosis and Diastematomyelia. - age 14 -*local deformity*



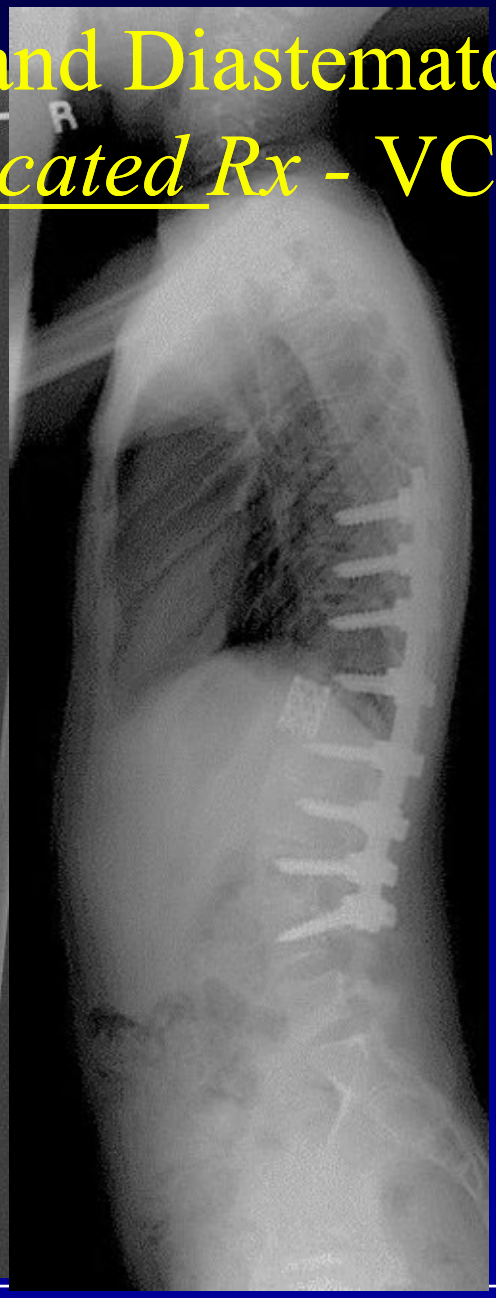
Congenital Kyphosis and Diastematomyelia.

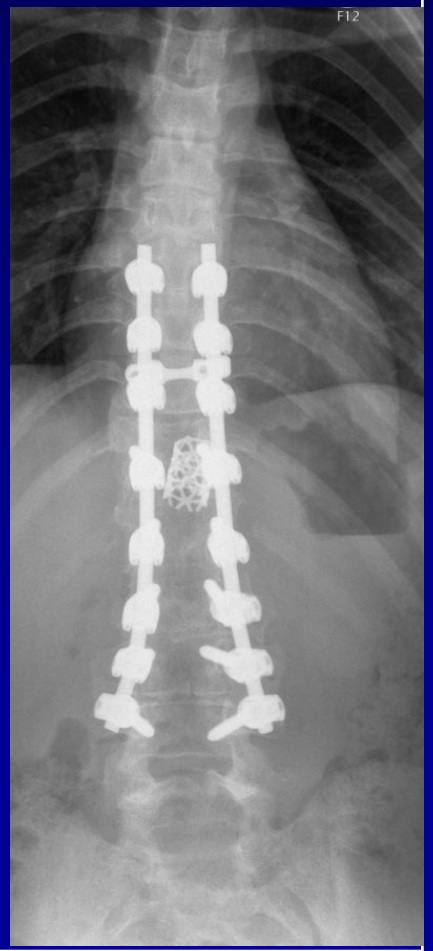
Local deformity, co-located Rx - VCR,



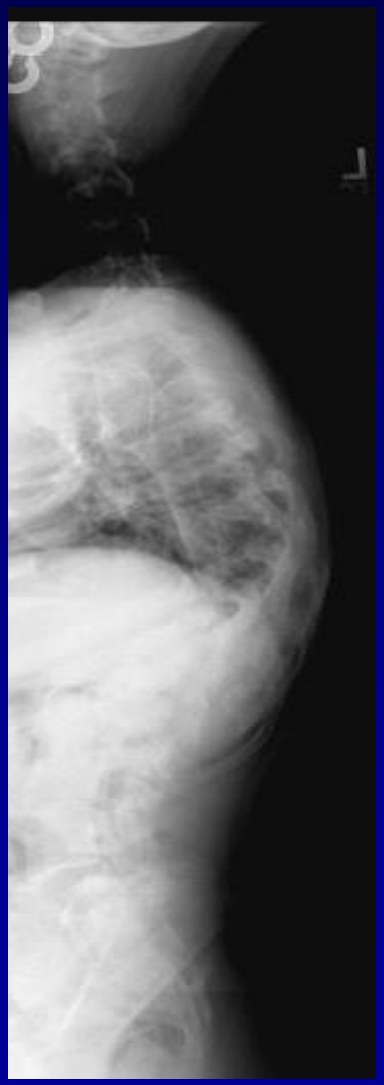
Age 2

e 13

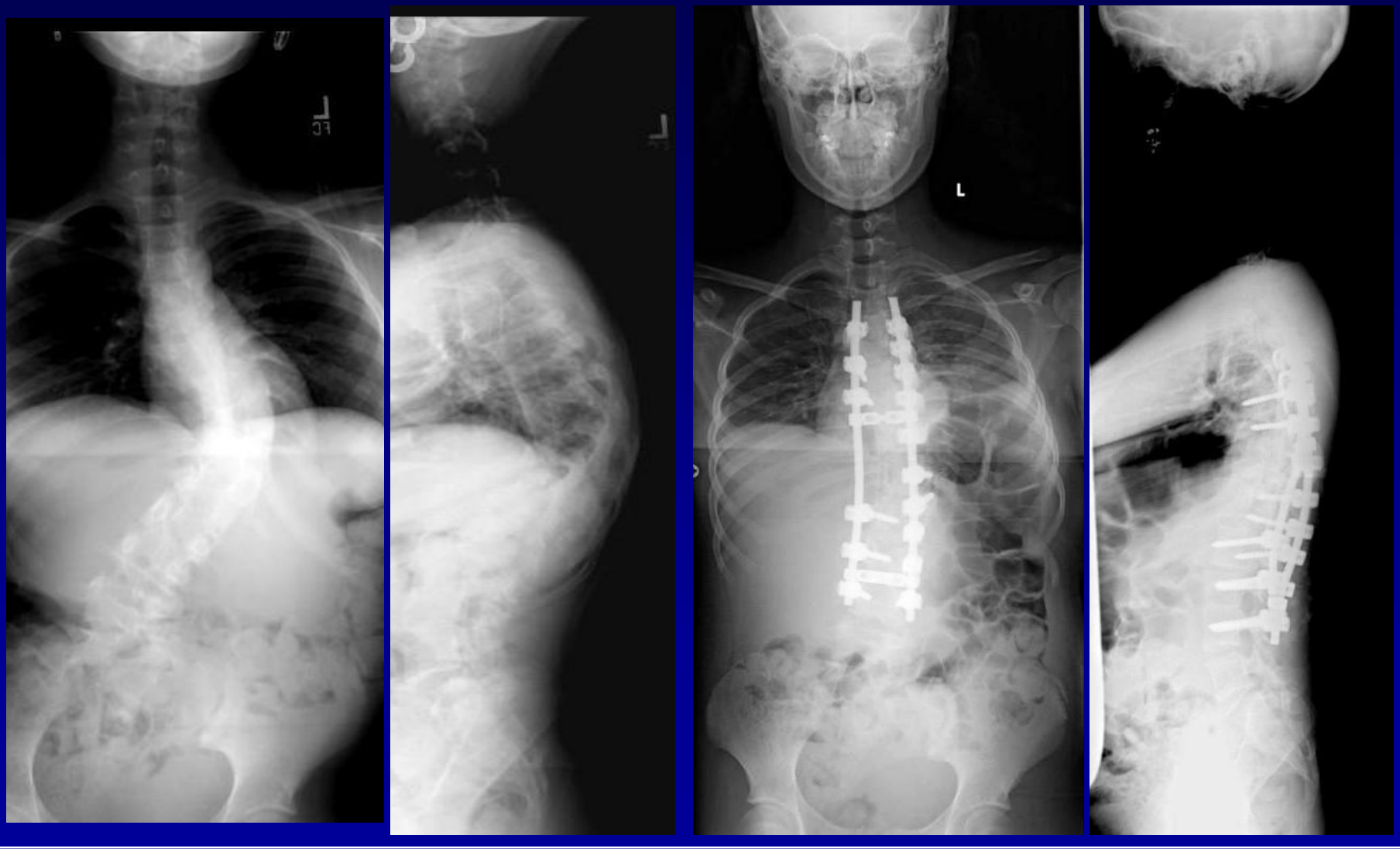




Congenital scoliosis, failed in-situ fusion – *local deformity, coronal and sagittal colocated*

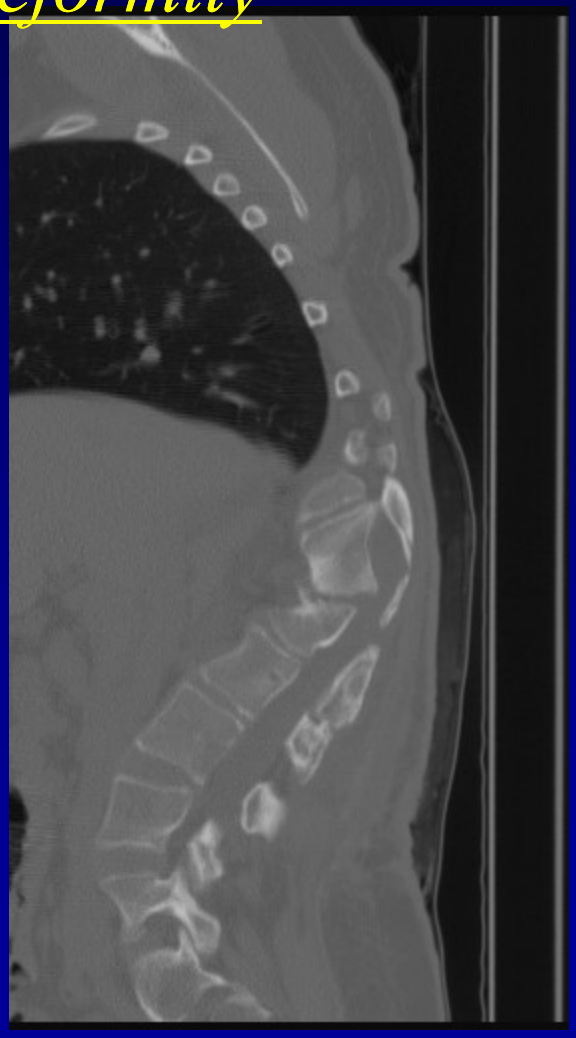


VCR for congenital scoliosis, failed in-situ fusion – *local deformity, coronal and sagittal colocated - VCR*

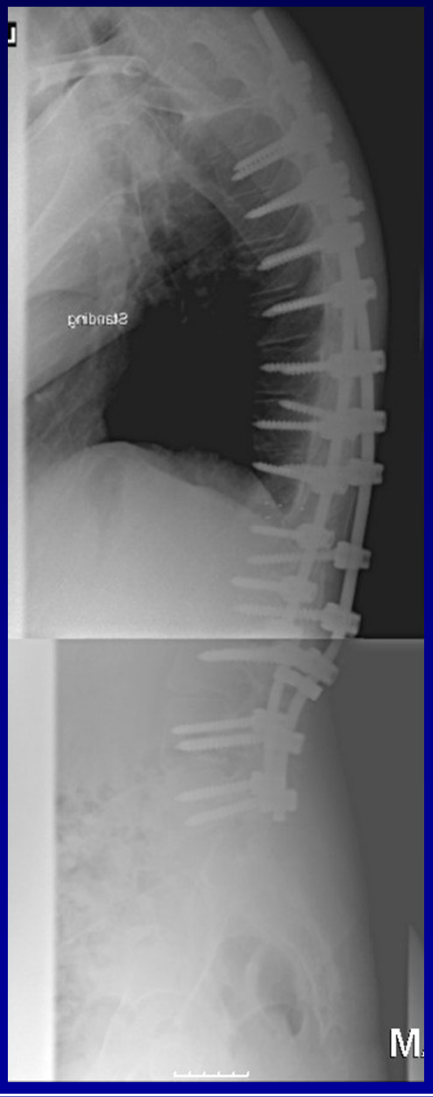


19 yo failed congenital kyphosis A and P resection. Prior infection. Progressive spasticity

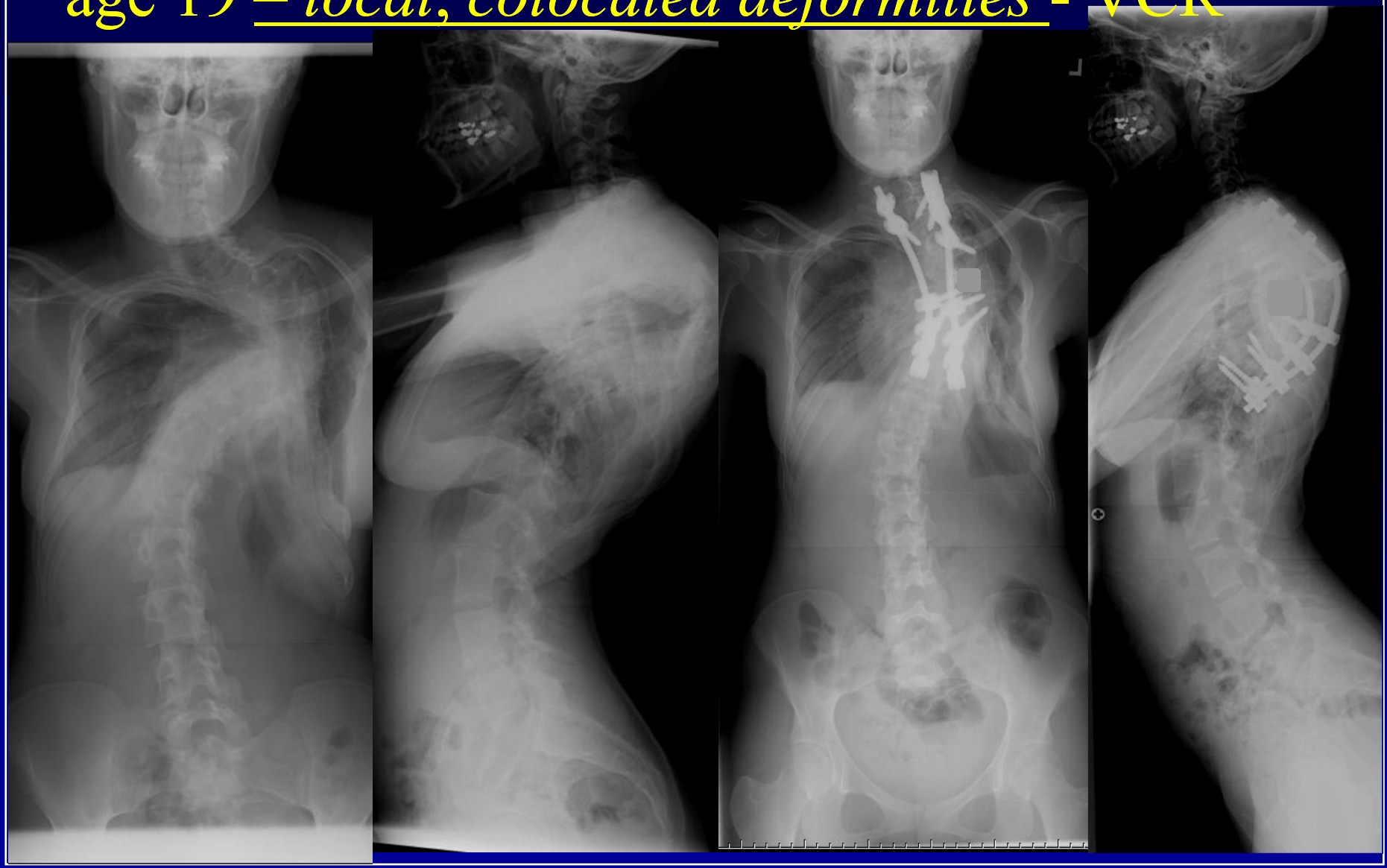
Local and global deformity



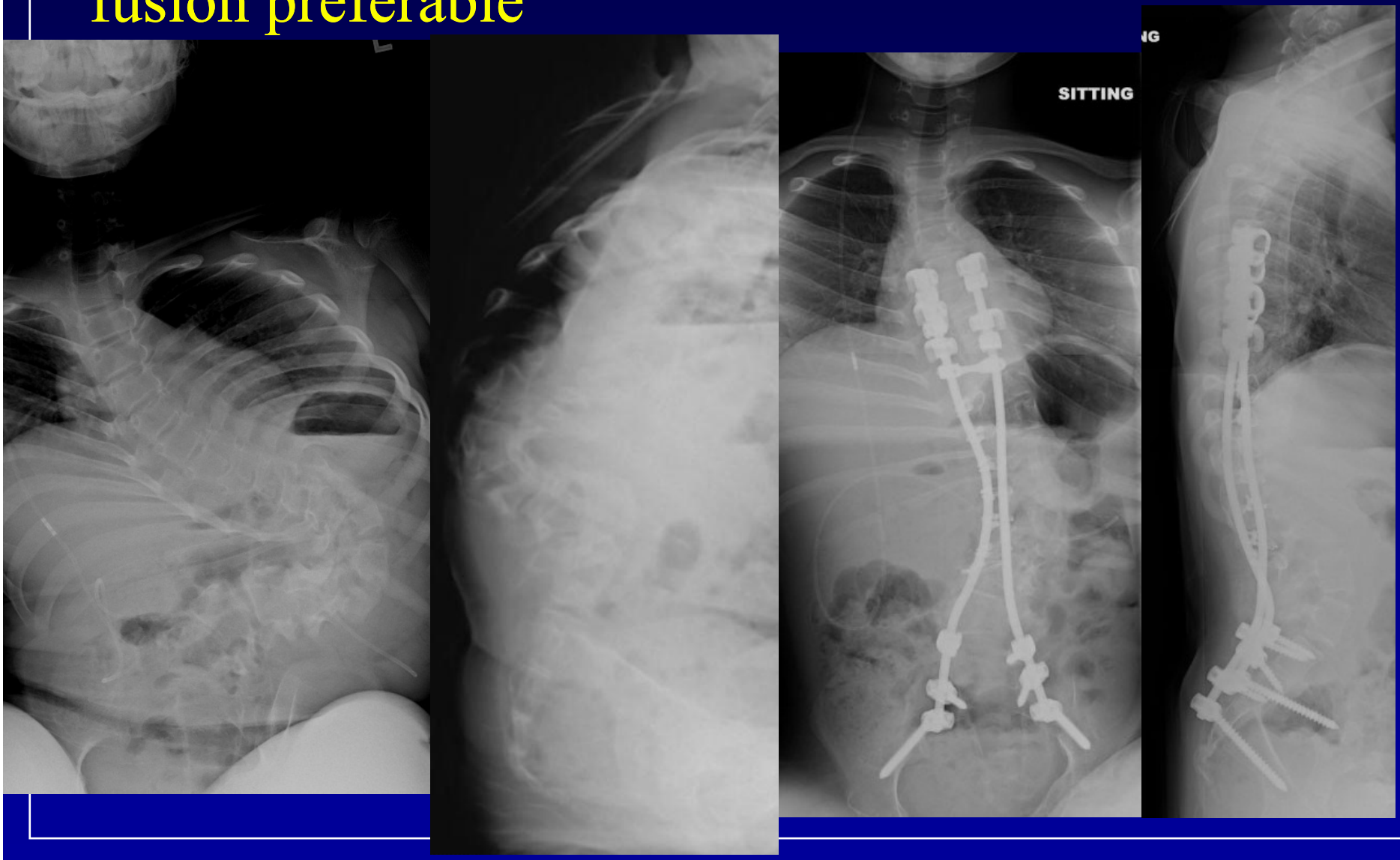
19 yo failed congenital kyphosis Rx. *Local and global Deformity colocated.* VCR *<u>and</u>* long fusion



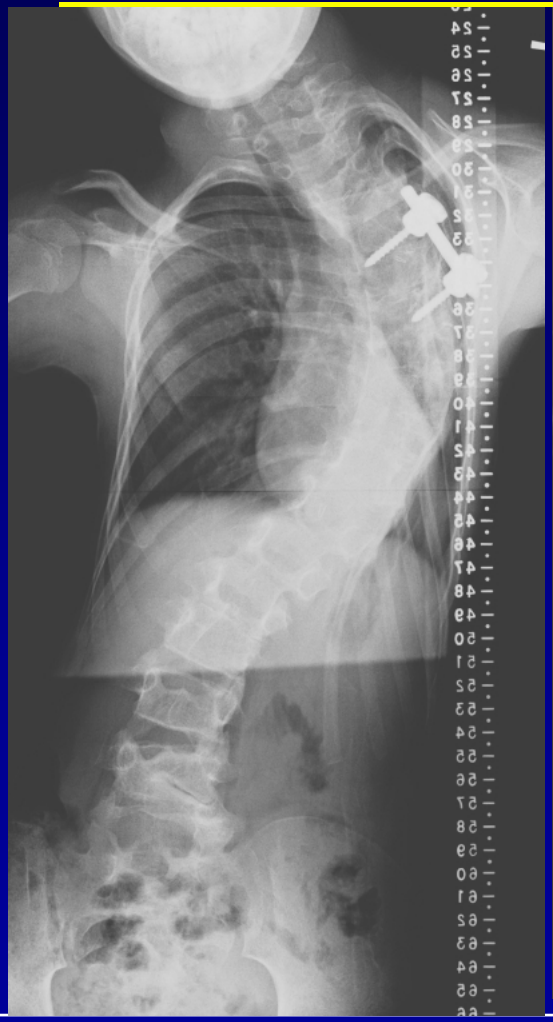
Thoracogenic and congenital kyphoscoliosis age 19 – *local, colocated deformities* - VCR

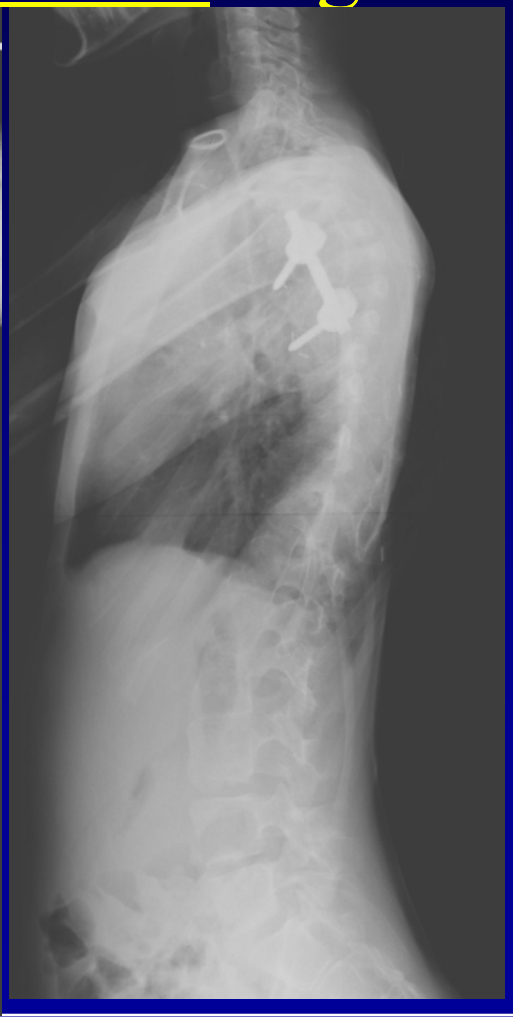


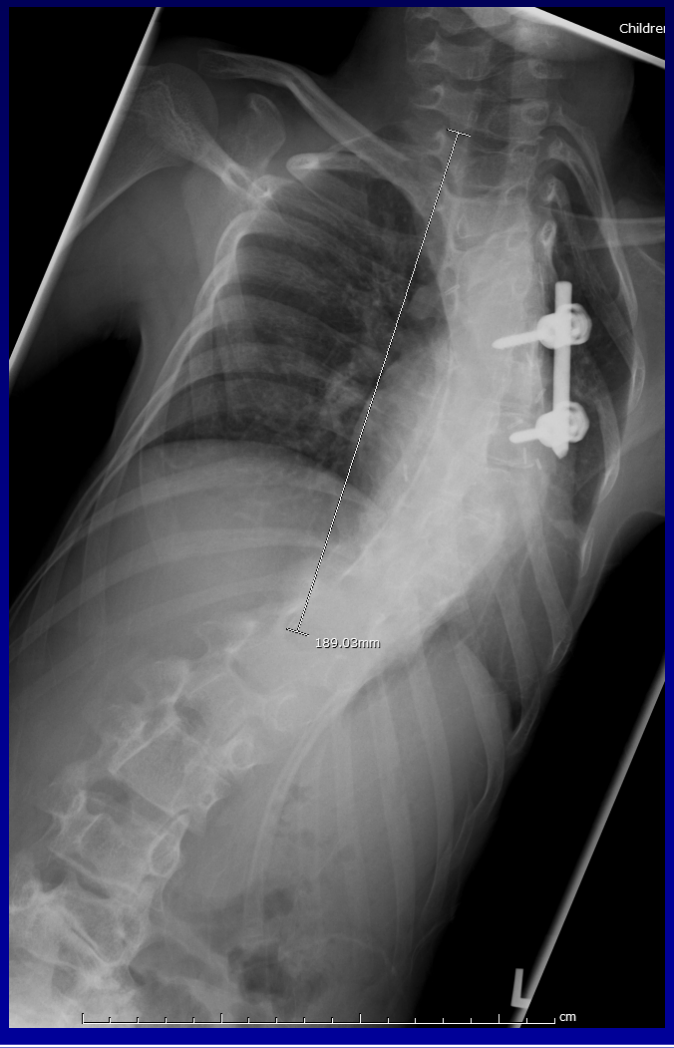
Congenital scoli, failed in situ fusion. *Local and global deformity not co-located* – A/P release, fusion preferable



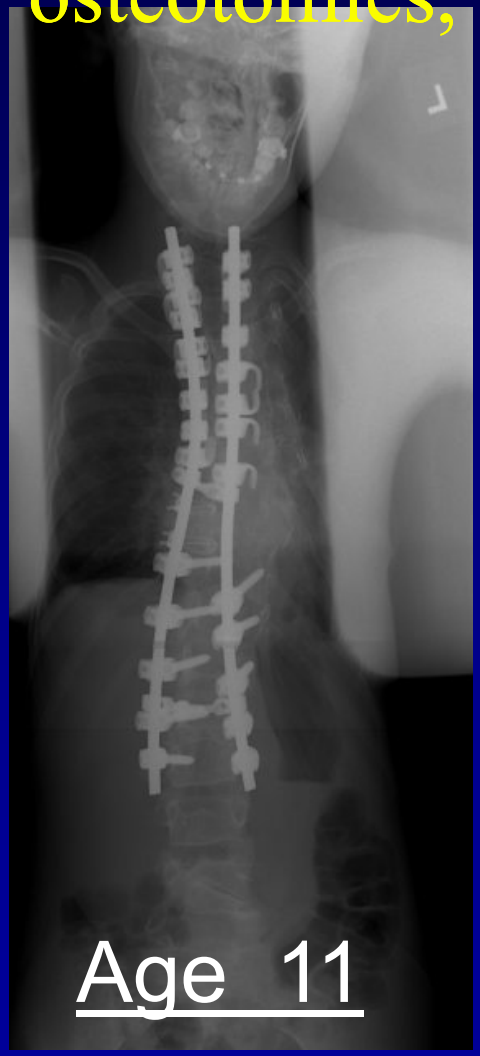
Congenital Scoliosis. Failed early fusion attempt. *Global (not local) deformity. kyph and scoli not co-located*. Age 11

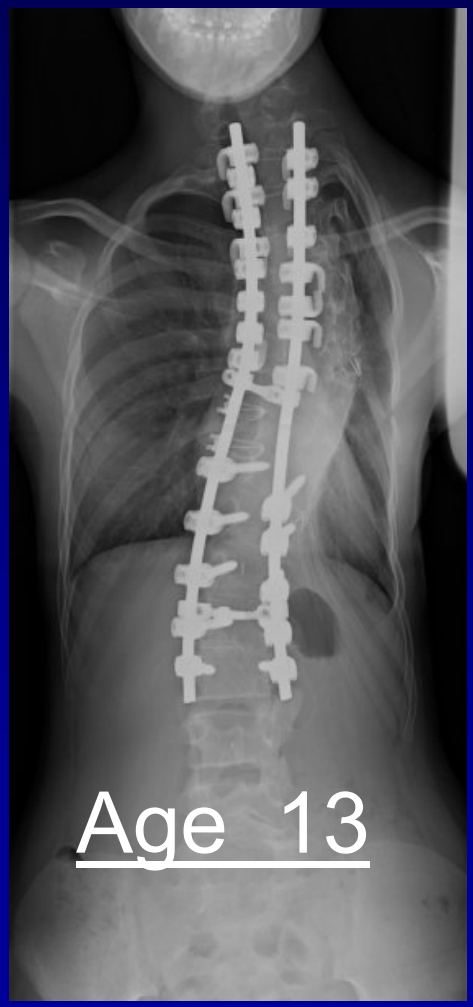






Congenital Scoliosis, *global deformities not colocated*. Anterior release, posterior osteotomies, fusion



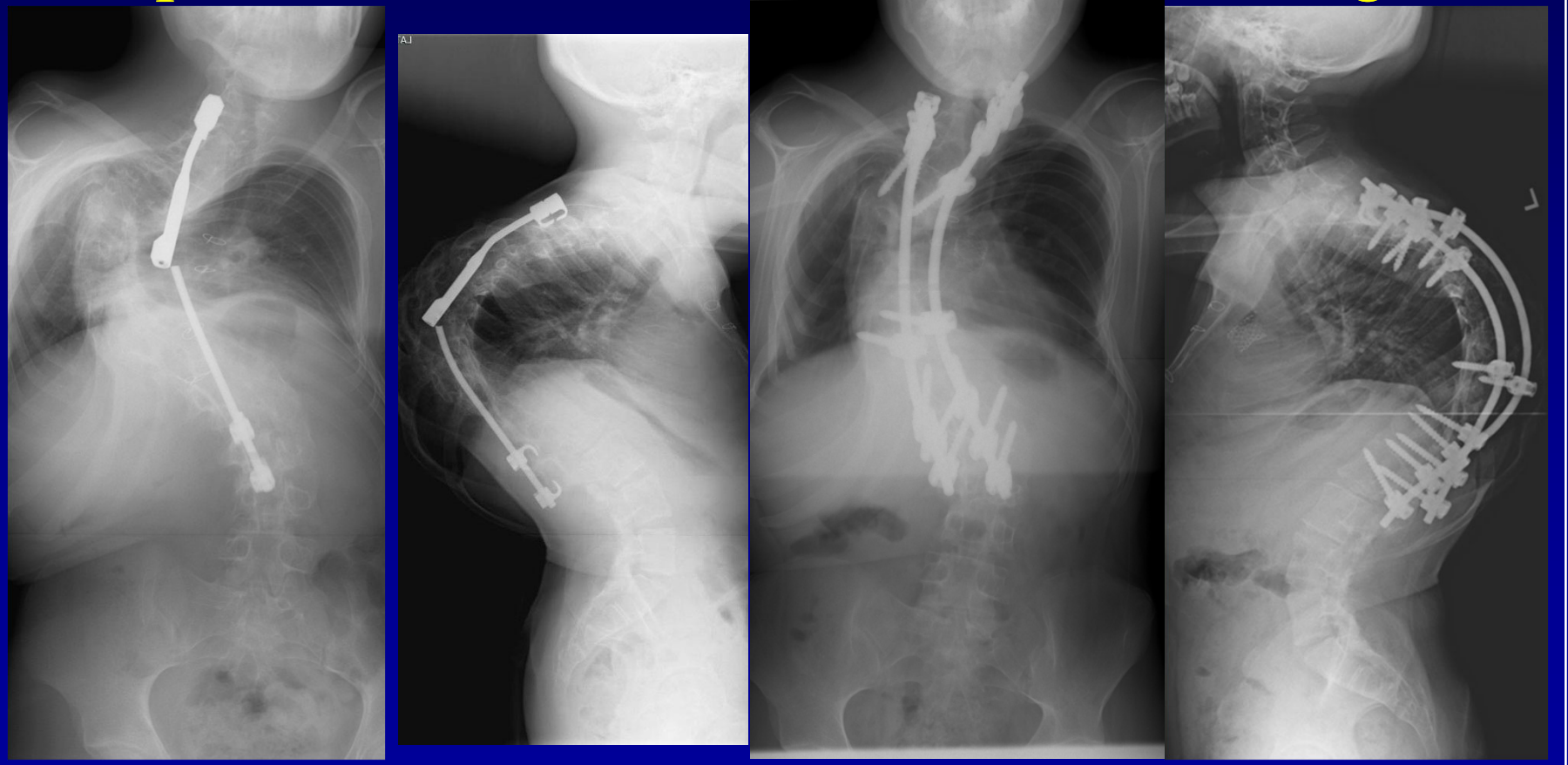


Special challenges for complex osteotomies in congenital deformity:

- Neuraxis anomalies
- Anomalous vertebrae with +/- *pedicles*
  - Prior fusion, scarring from GR or VEPTR
- *Anchor points critical to VCR*
- *What if anchor points inadequate*?
  - *Staged VCR:*
    - *1 – Fusion and instrumentation - healing*
    - *2 – VCR with pre-placed anchor points.*

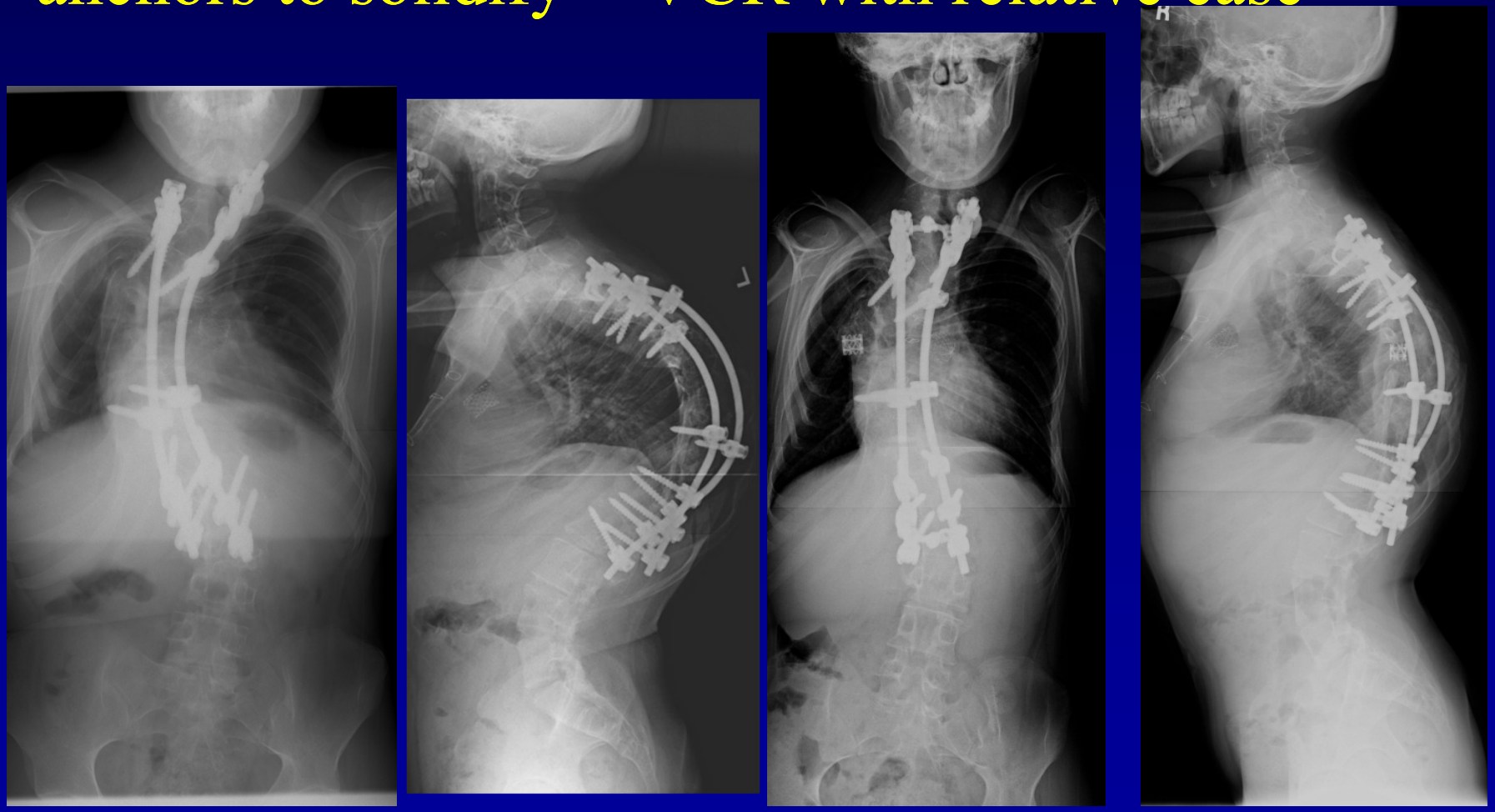
Staged VCR

Age 16 Severe CHD, failed GR–Anchor points complex, unsatisfactory quality for VCR – accept instrumentation and fusion as first stage



Staged VCR

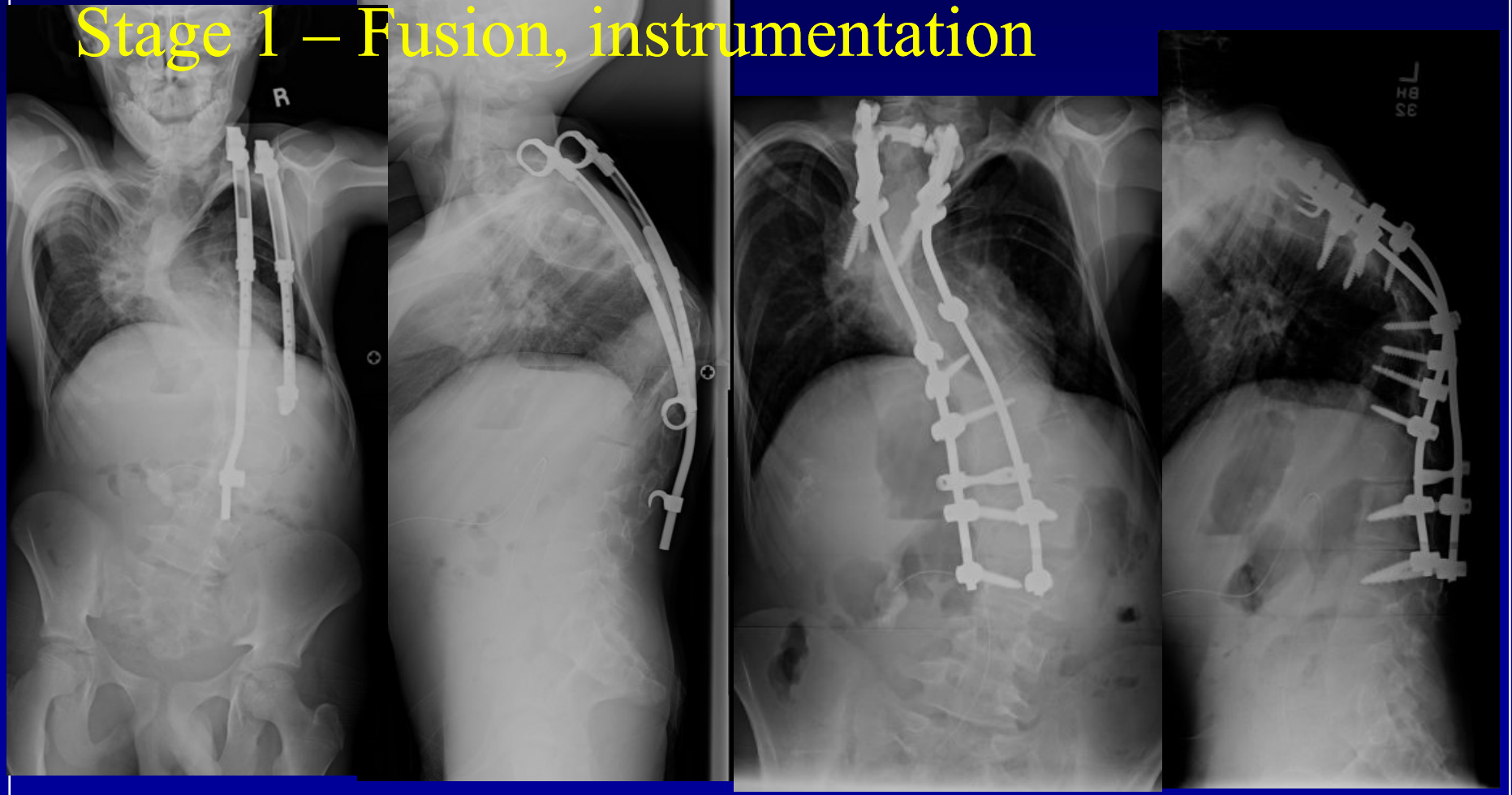
Age 17 – One year for fusion to occur and anchors to solidify - VCR with relative ease



Staged VCR

Age 13, VACTERL, prior GR, in situ fusion, VEPTR, . Distorted anatomy. *Osteopenia*.

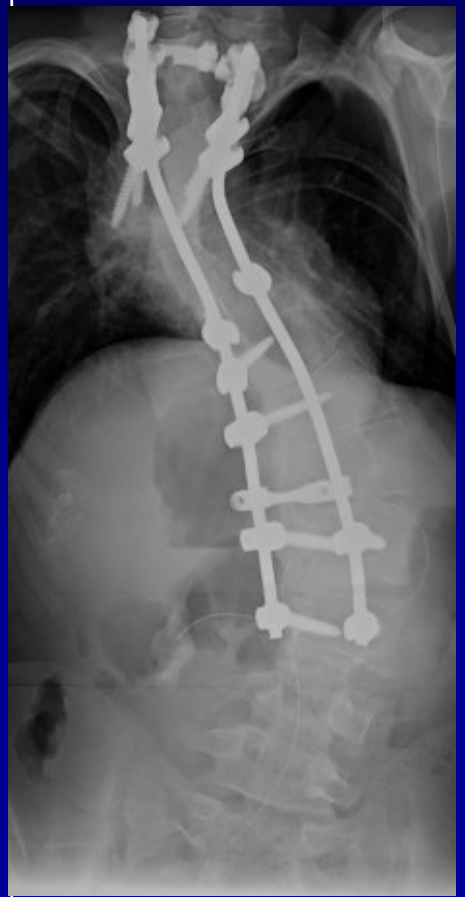
Stage 1 – Fusion, instrumentation

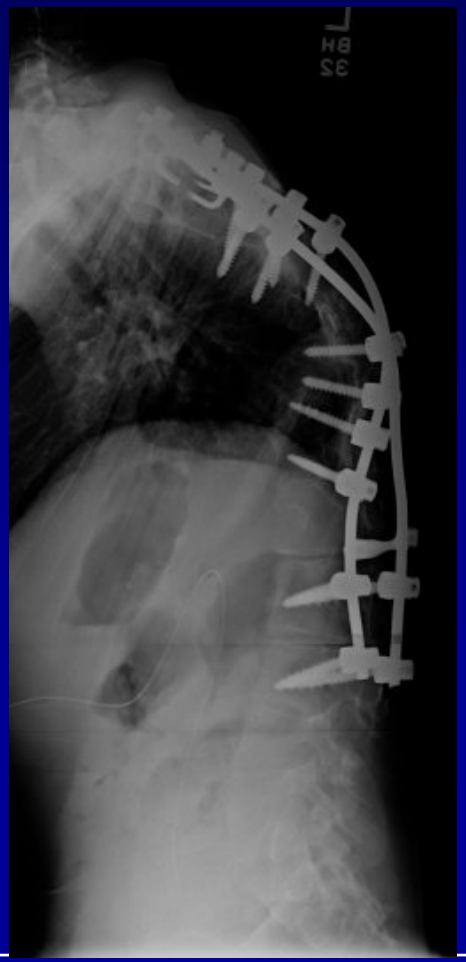


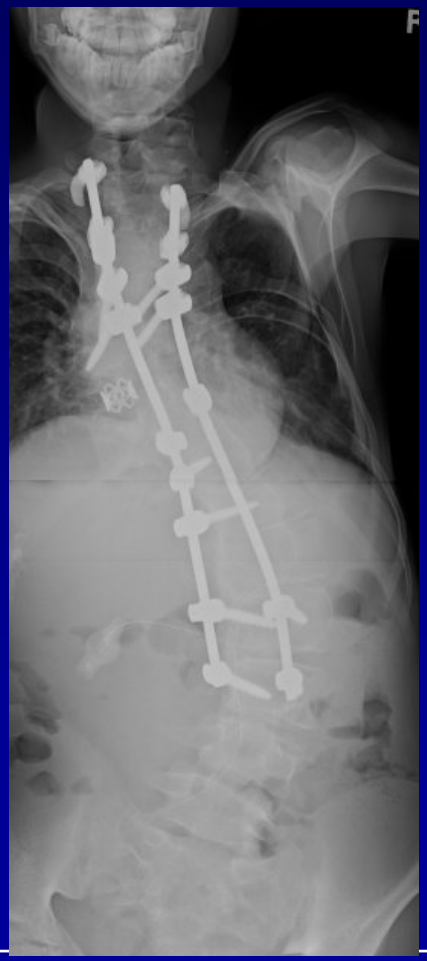
Staged VCR

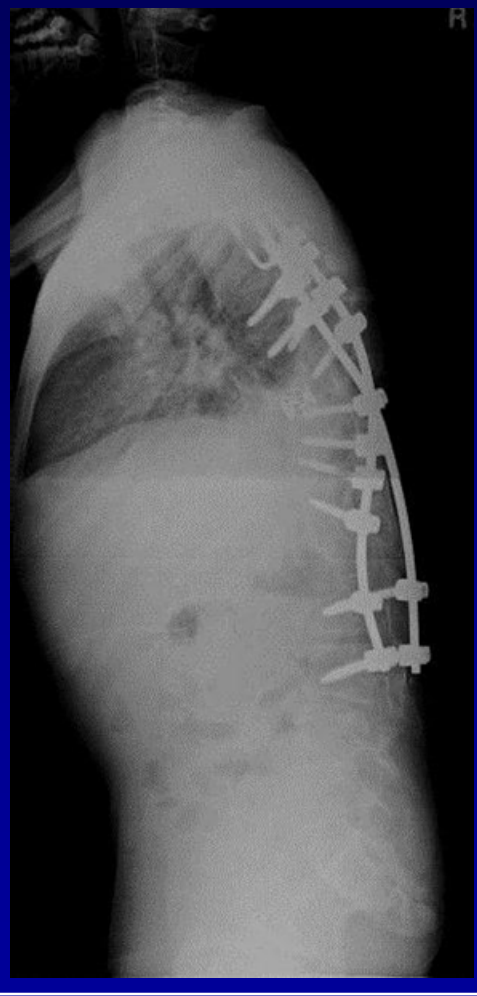
VACTERL:

Stage 2 - 1 year later - VCR





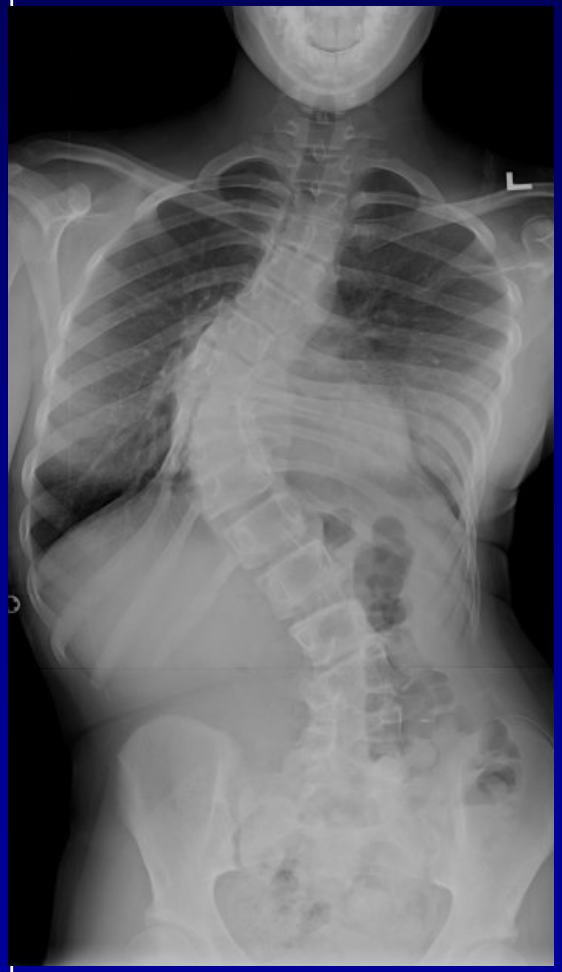




Congenital scoliosis, late progression, dancer, *one local, one global deformity deformities not co-located*.
Rigid congenital segments in opposite directions

bends

Solution: Anterior release, posterior osteotomies for thoracic curve, posterior VCR for L/S curve




Young Children – growth a major concern

Congenital Spine Deformity *with substantial growth remaining*

- Treatment decisions – how to sort through?
- Keep goals in mind
  - Do you know the natural history?
- *Local* deformity or *Global* deformity or both
  - *Local definitive* treatment?
  - *Global growth-friendly* treatment
- *Consequences of early intervention on spine growth*?

Treatment *goals* in congenital scoliosis and early onset deformity:

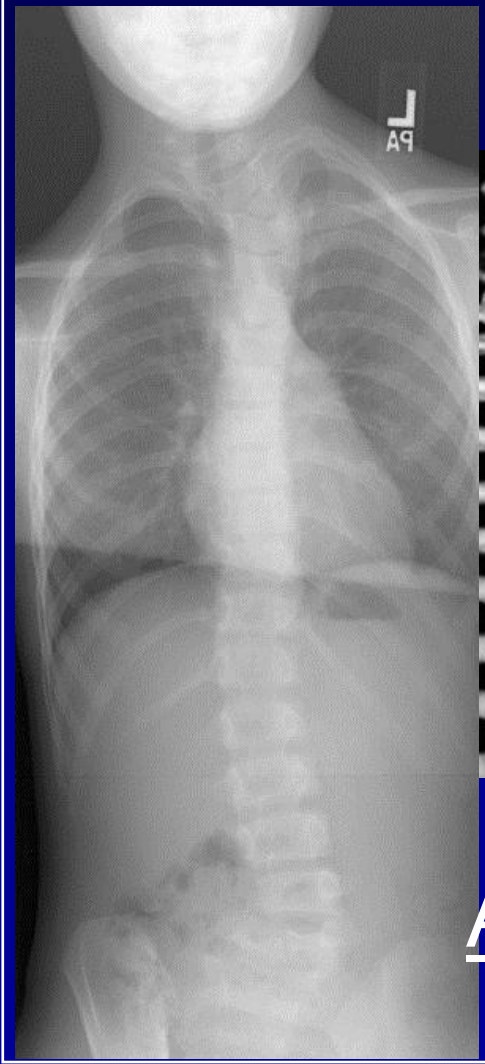
- At maturity try to achieve:
  - Maximum
    - Spine length, flexibility
    - Thoracic function (volume, movement)
    - Lung growth
  - Minimum
    - Surgery
    - Complications
    - Hospitalizations, disability

Early onset deformity decisions: which technique?

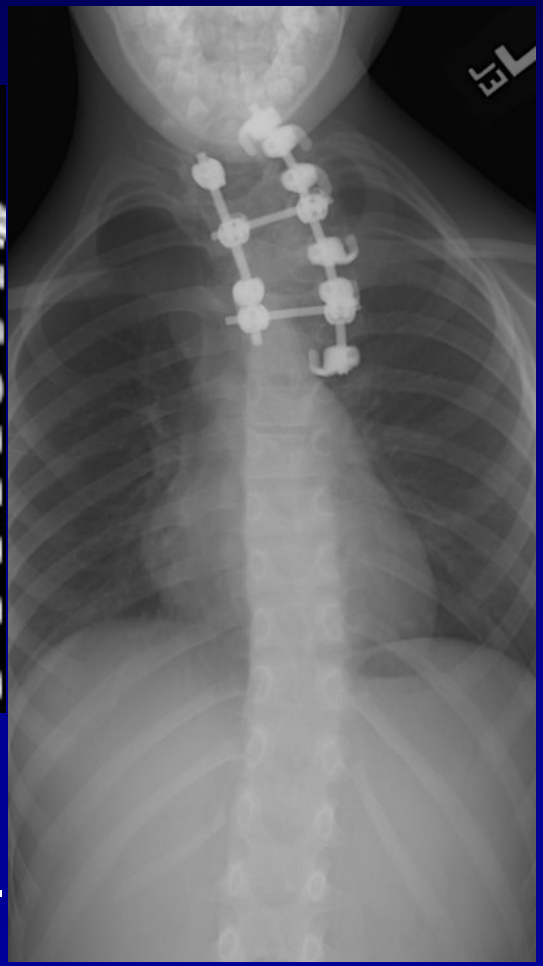
- *Local Deformity - Short section* of spine involved:
  - Wedge resection or local fusion
  - (loss of growth tolerated)
- *Global Deformity -Long section* of spine involved
  - Growth friendly
    - growing rods
    - VEPTR
    - Shilla

Both - use combinations

Local deformity progressive In Situ Fusion, some correction with growth – osteotomy more than needed



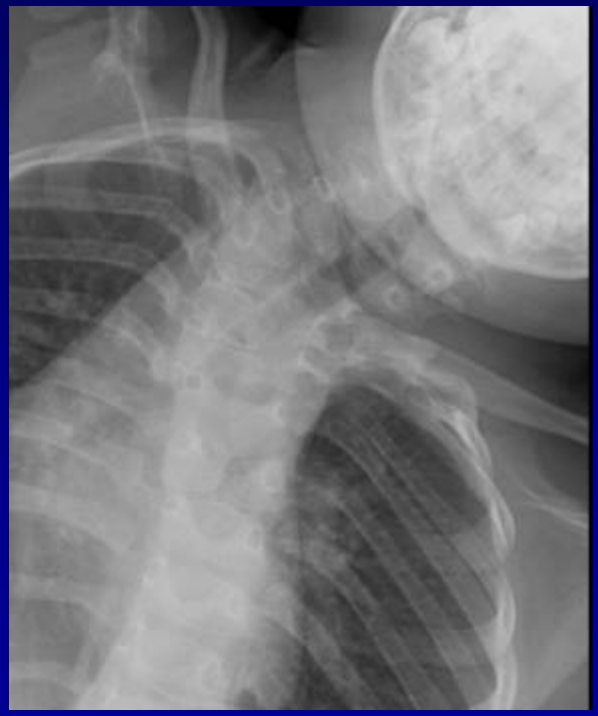
Age 7

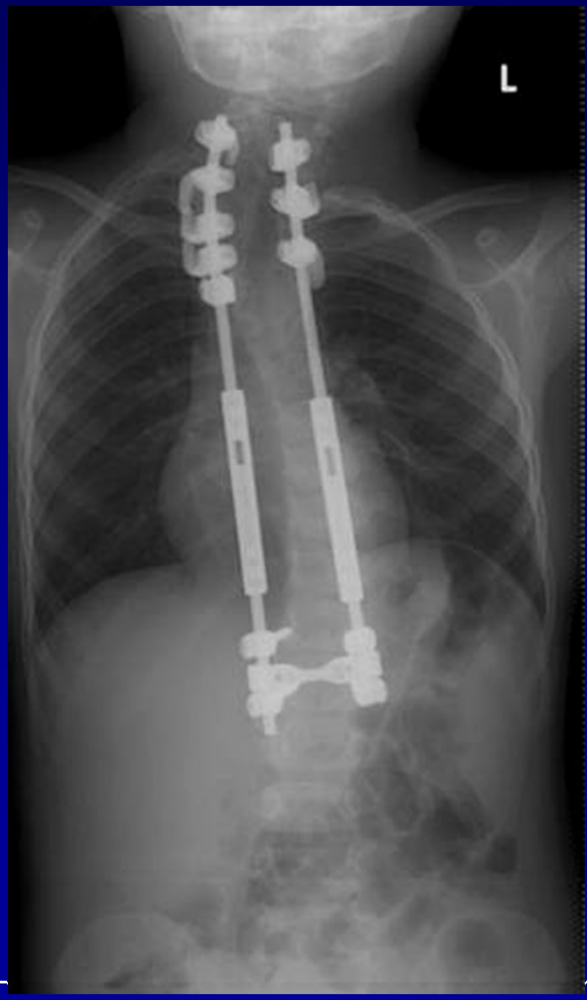


Age 15

Congenital Scoliosis, short and long section of spine involved

Definitive local treatment, Growth-oriented global treatment: Growing Rods *and* In Situ Fusion

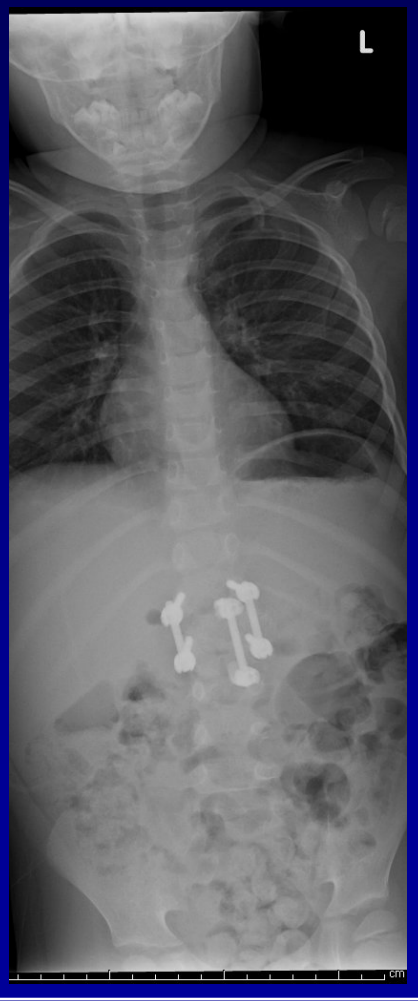


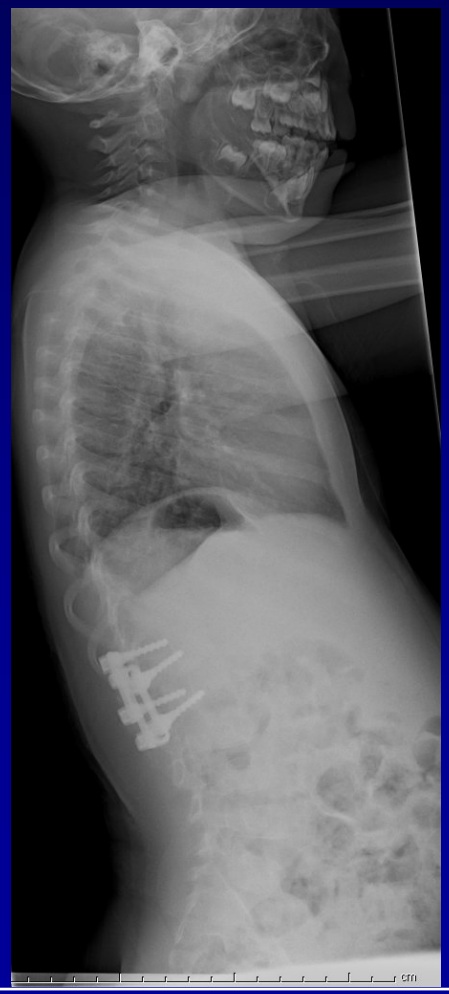


Congenital Scoliosis: *Short segment of spine involved – definitive treatment – growth loss acceptable*

- Short segment of spine involved
- Hemi excision

Local deformity only – Growth not a problem Posterior only hemivertebra excision age 3
Hedequist 3- rod technique –

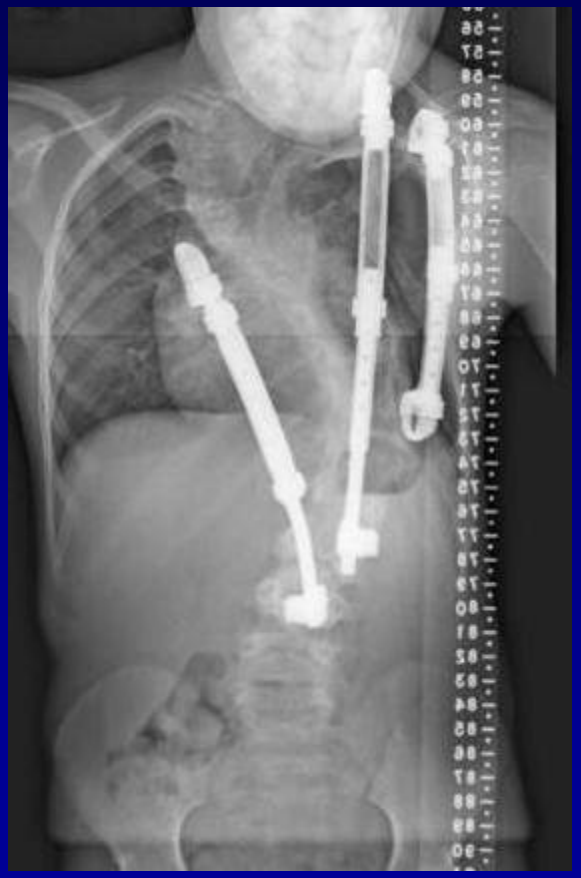




Growth-oriented global treatment: cong scoli, rib fusions

VEPTR – *with* expansion thoracostomy

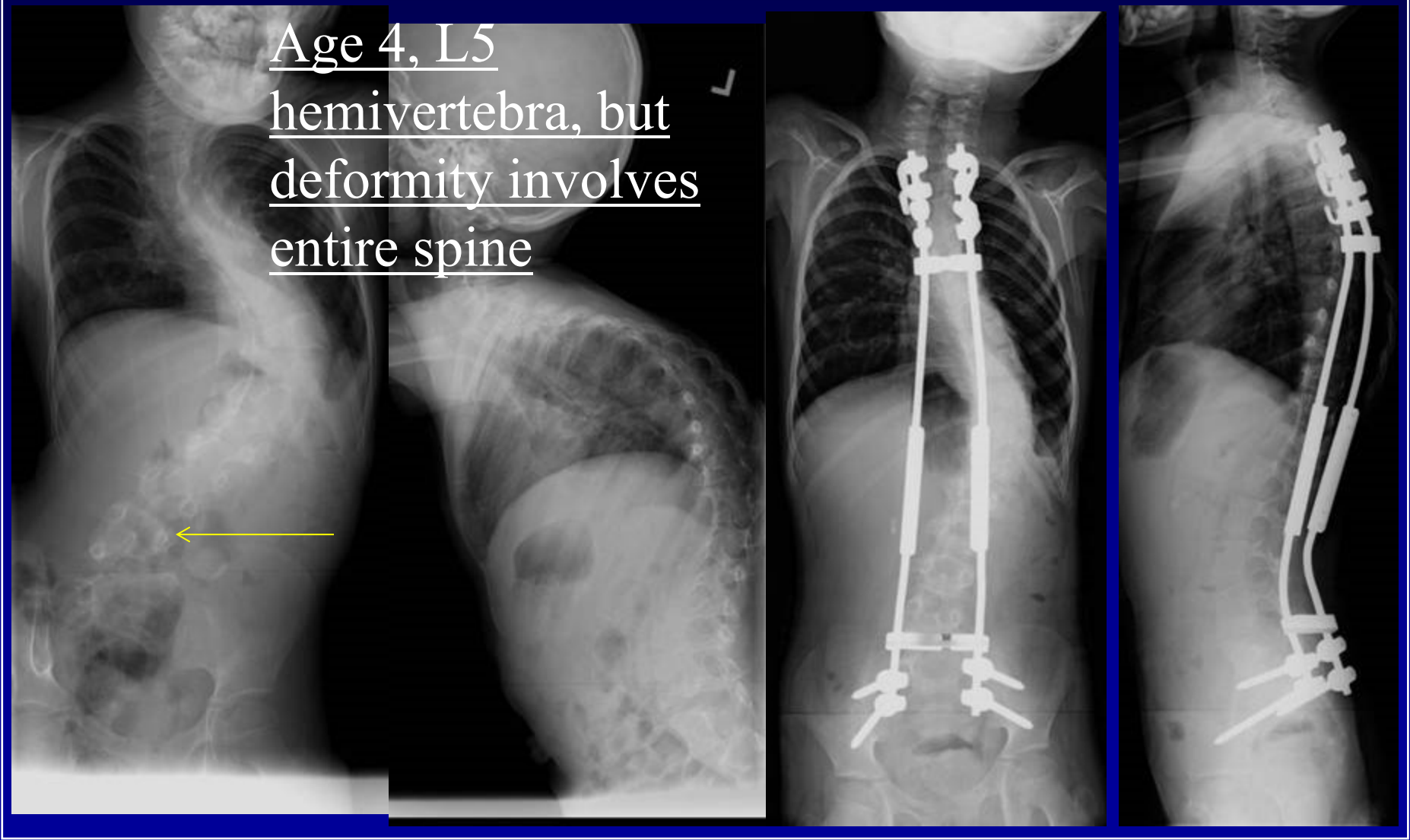
Age 3



Age 8

Age 11

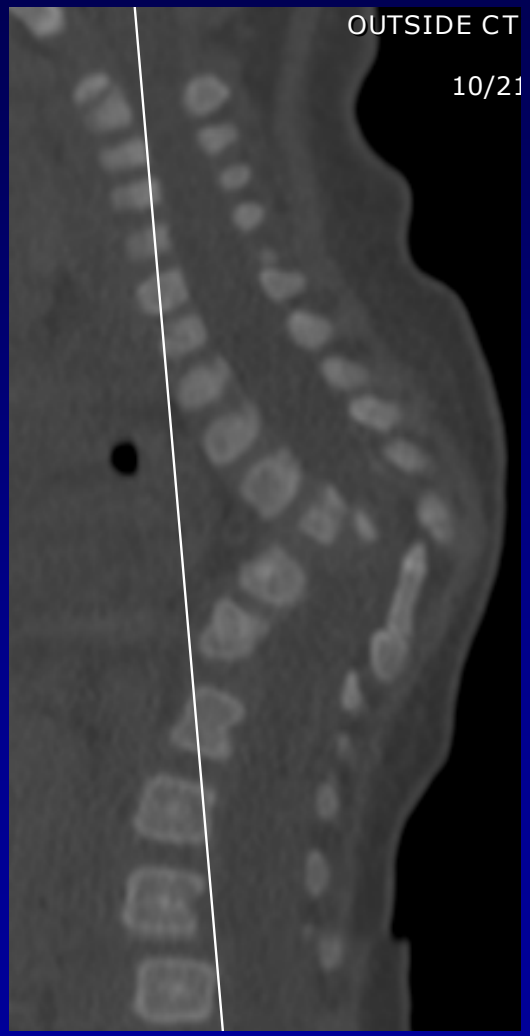
Congenital Scoli, long section of spine involved
Growth-oriented global treatment : Growing Rods

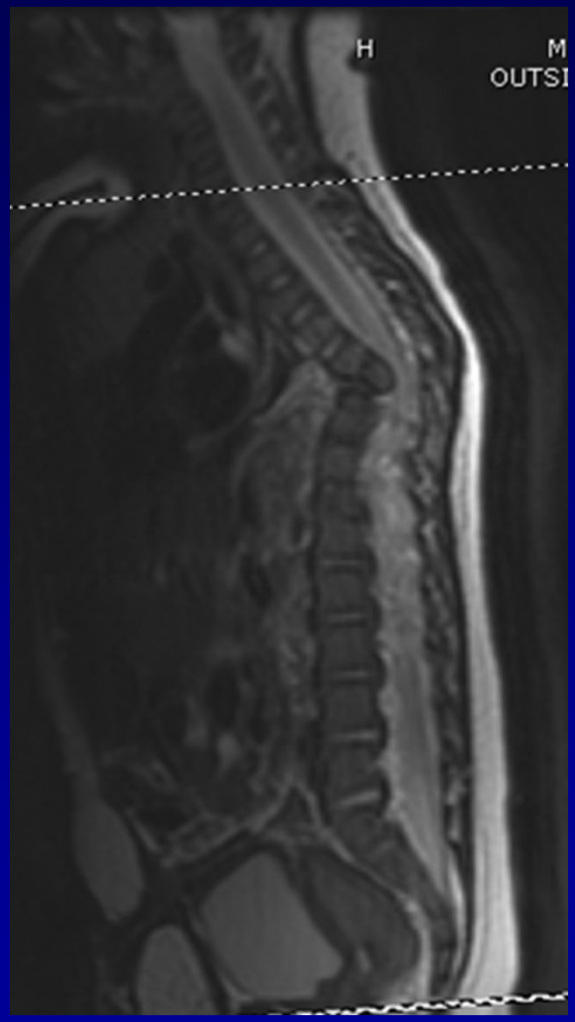


Age 4, L5 hemivertebra, but deformity involves entire spine

Congenital Spinal Dysgenesis/ Dislocation

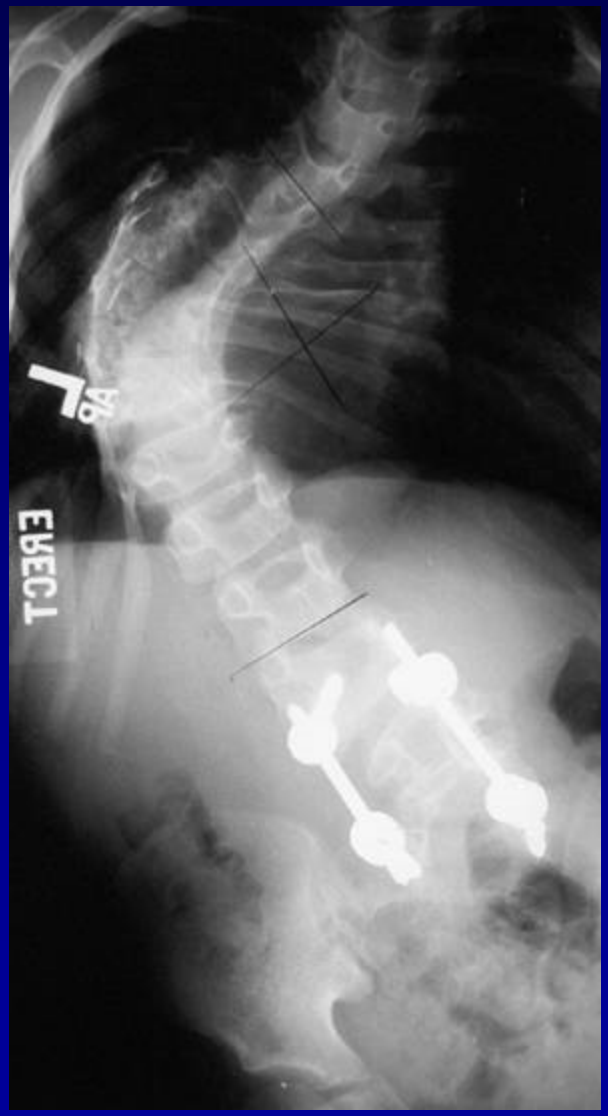
Local (for dysgenesis) *<u>and Global</u>* (for deformity) treatment needed





Congenital Spinal Dysgenesis/ Dislocation:
VCR for dysgenesis/dislocation
Growing Rods for fixation, then growth

Hemi excision and thoracic in situ fusion– not enough global control



When to intervene in congenital deformity?

Options for congenital deformity- *When to intervene surgically? Is earlier always better?*

Earlier intervention:

- Early chest expansion = max opportunity for lung growth
- Deformity more flexible, milder but
  - Max # surgeries, max # complications?
  - Spontaneous spinal fusion beneath rod earlier?
- Current thoughts? – we may only have 5+ years of lengthening before growth ending complications likely
  - *Where would you choose those 5+ years*?

Options for congenital deformity- When to intervene surgically? *Is earlier always better*?

Later intervention:

- Bones bigger, stronger
  - Instrumentation easier
  - Fewer surgeries, complications?
  - Spine deformity worse?
- *Lung growth stunted*
- *Chest deformity irrevocably worse*?
  - (little we can do for severe chest wall deformity once it has occurred)
- Campbell data – earlier intervention = better PFT's

Options for congenital deformity- *When to intervene surgically? Don't wait if -*

- *Lungs significantly constricted*
- *Neuro staus threatened*
- *More of spine involved – secondary curves*
- *Otherwise:*
  - *Use evolution of chest deformity as a guide to timing of first surgery*?
    - *Less urgency about controlling spine deformity than chest deformity ?*

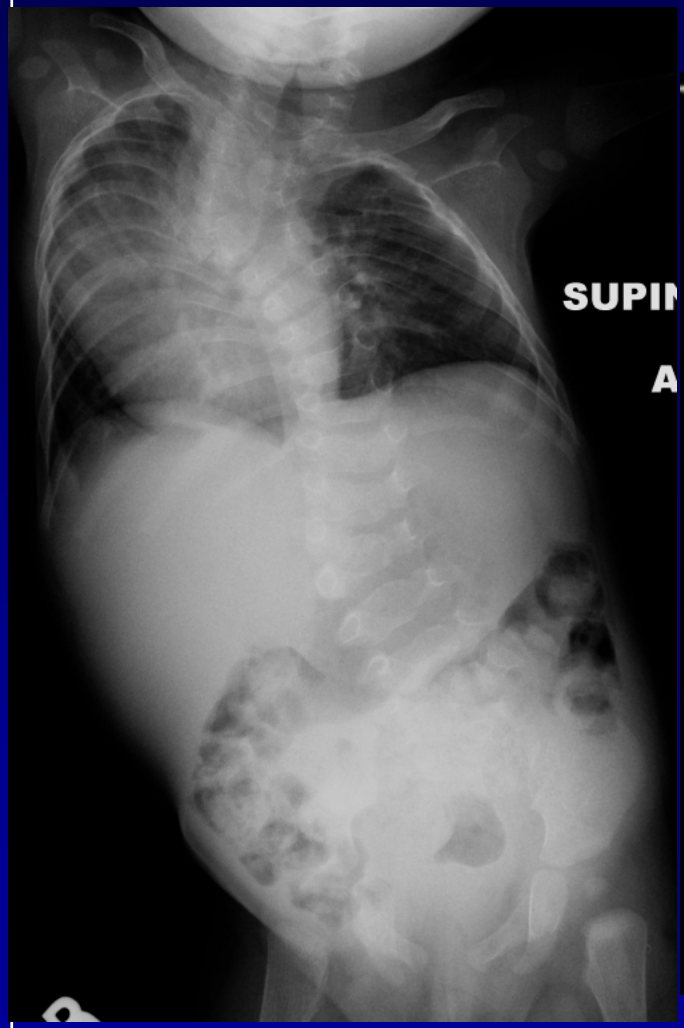
Use evolution of chest deformity as a guide to timing of first surgery?

- Rationale?
  - Surgical intervention can usually correct/control worsened spine deformity.
  - Surgical intervention less effective for established chest deformity
- Therefore less urgency about controlling spine deformity than chest deformity ?

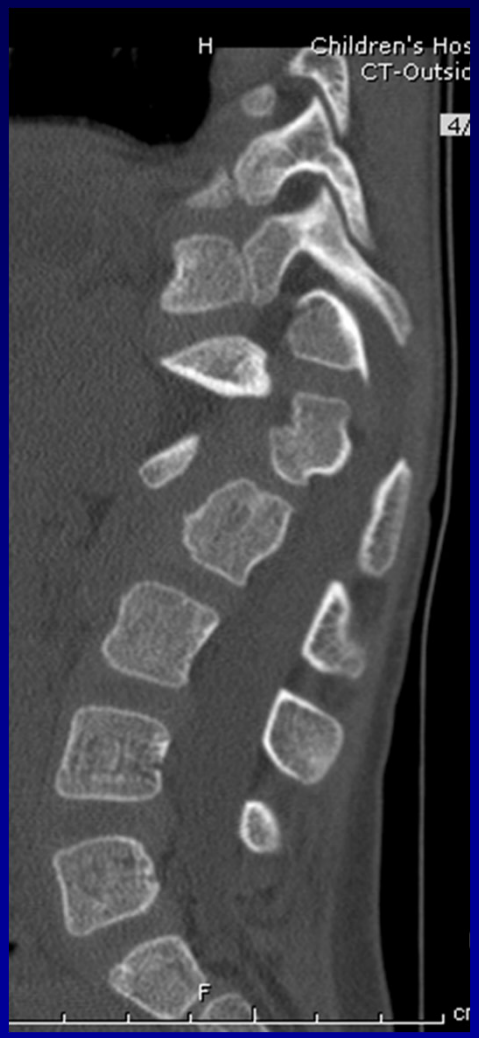
The dilemma:

- *Don't wait* to intervene – the chest deformity may be too severe to reverse
- *Don't intervene too early* – may get spontaneous fusion or other complication with premature cessation of growth friendly treatment.

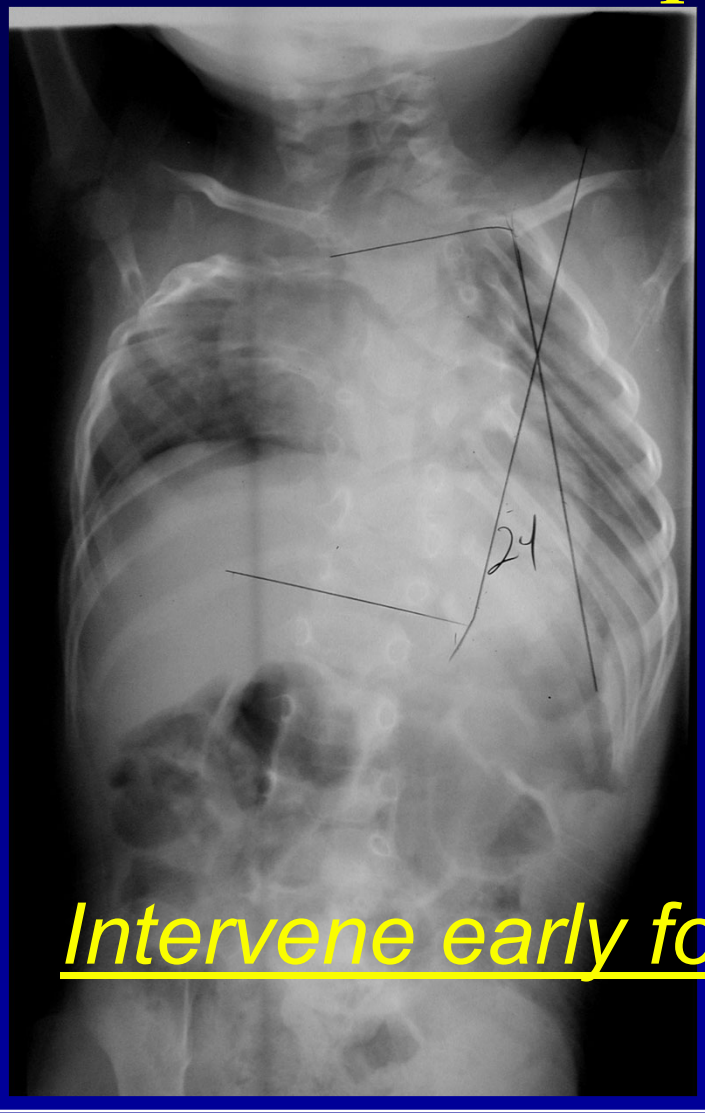
Multiple vertebral, chest wall anomalies
Lung growth - *Don't wait!*

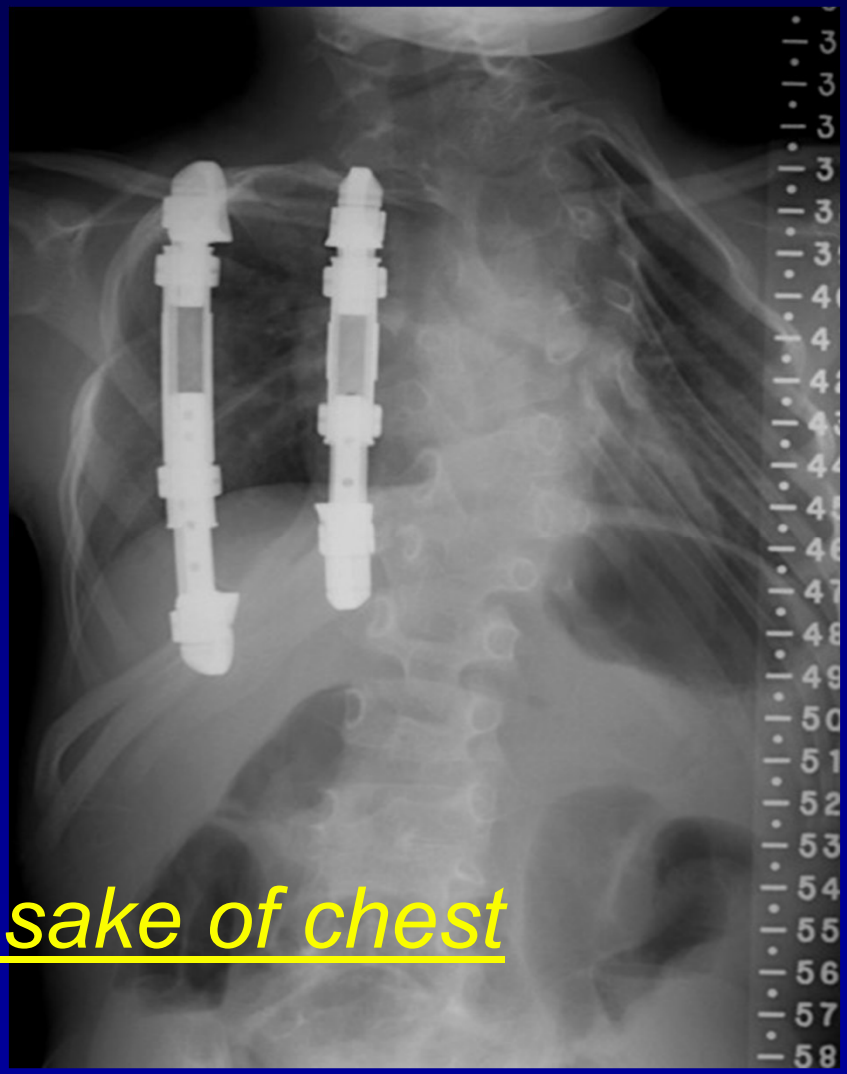


Congenital spinal dislocation, dysgenesis. Worsened spasticity *Don't wait!*



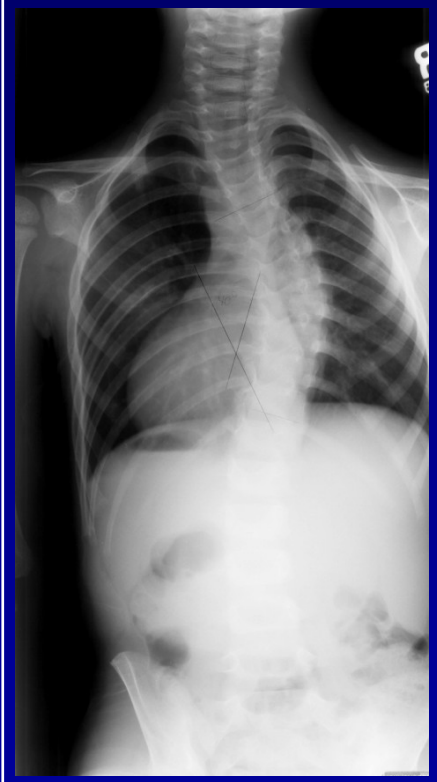
Congenital deformities age 2 – ‘stable curve’ – When to operate? *Don’t wait!*



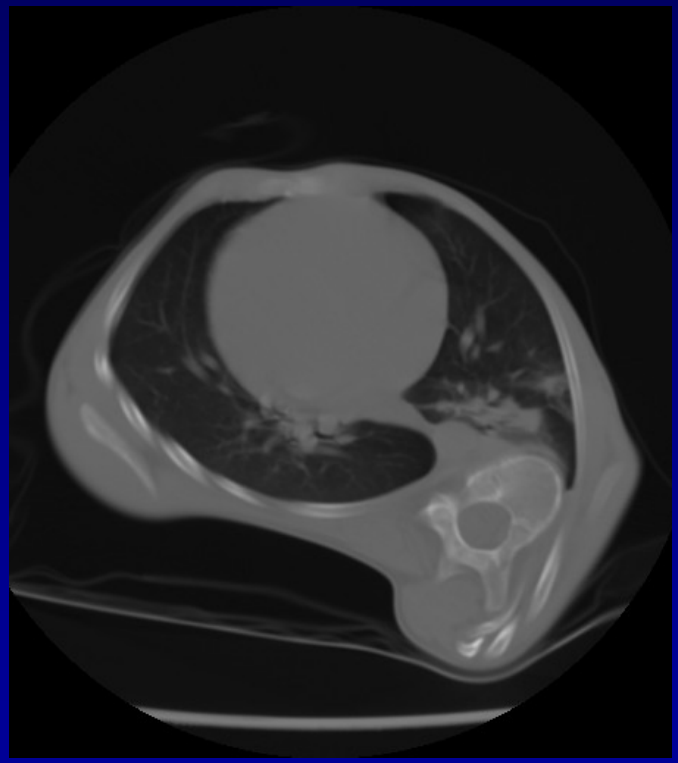


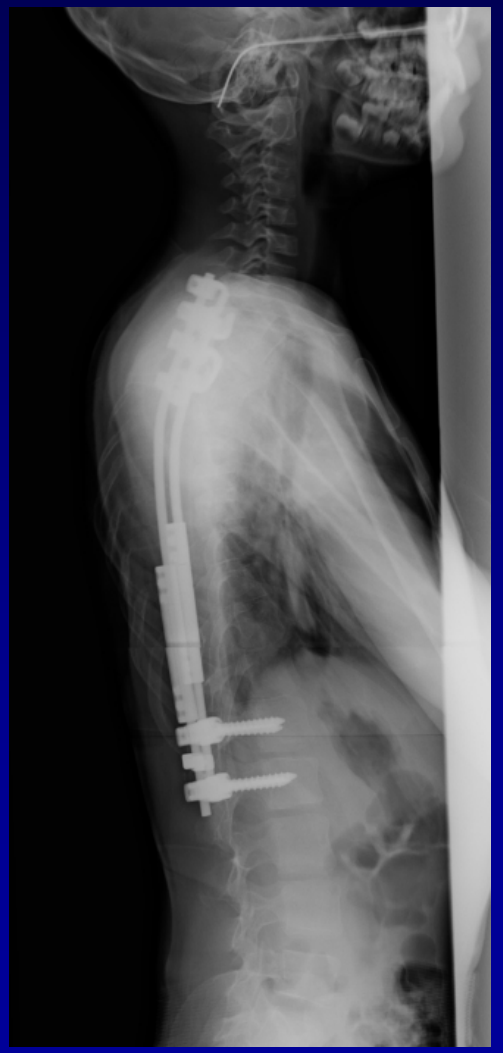
Intervene early for sake of chest

IIS Age 3, age 9 after casting, brace – much too long to wait – severe, irrevocable windswept chest deformity

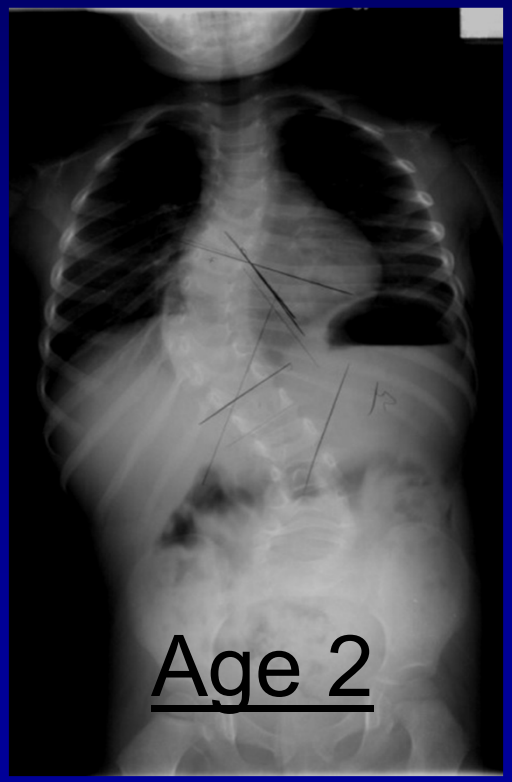


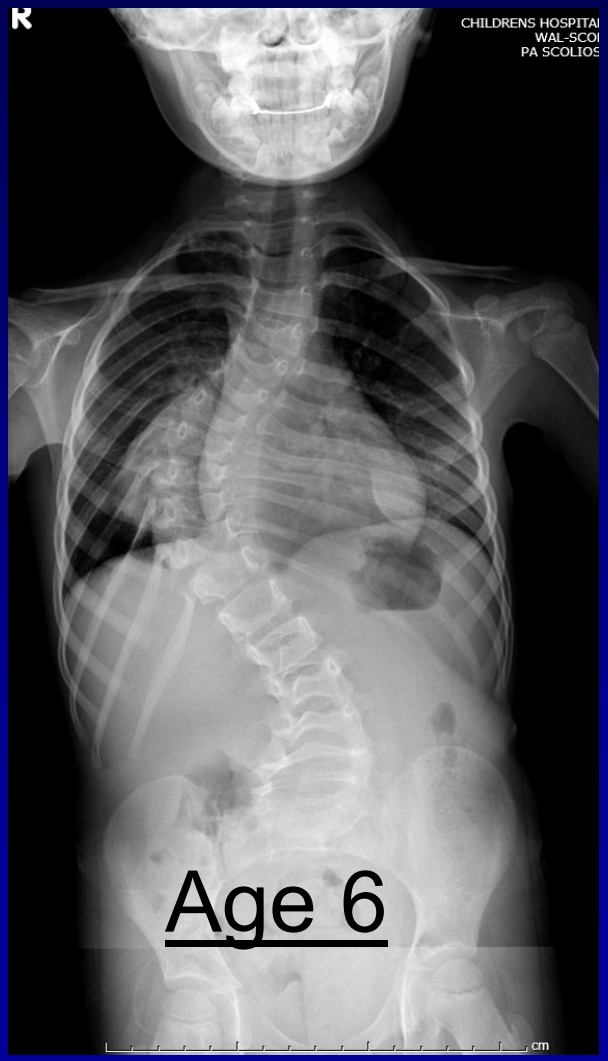
IIS – Age 9 – GR Spine under control, chest irrevocably damaged, PFT's 50% predicted.



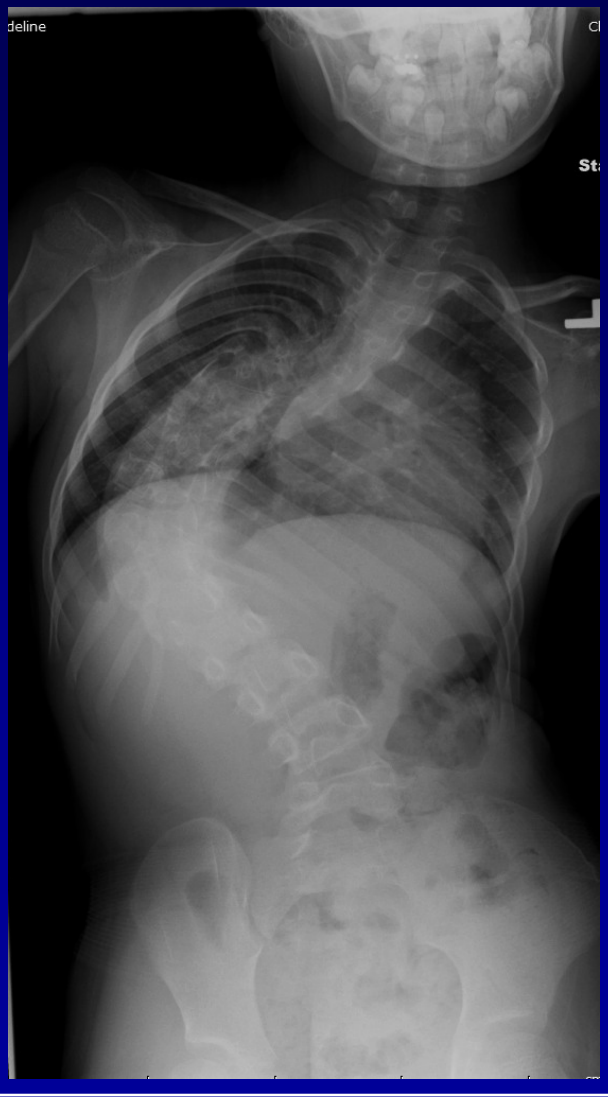


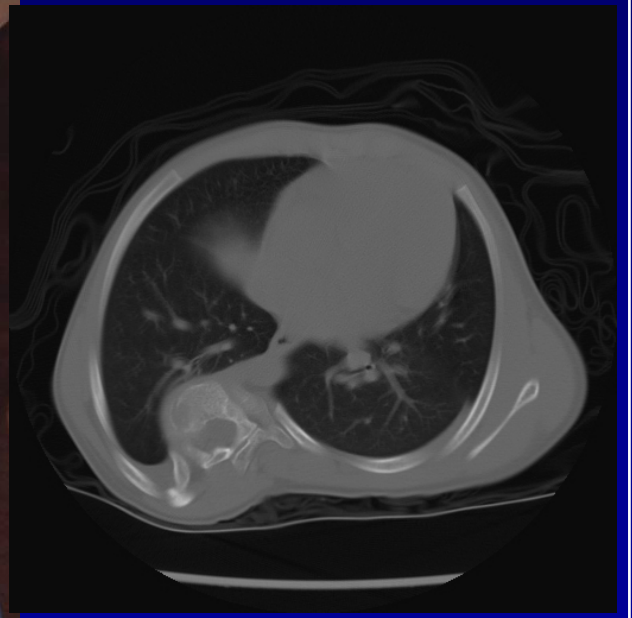
Infantile idiopathic, worsening spine deformity, modest but worsening chest deformity – wait? operate?



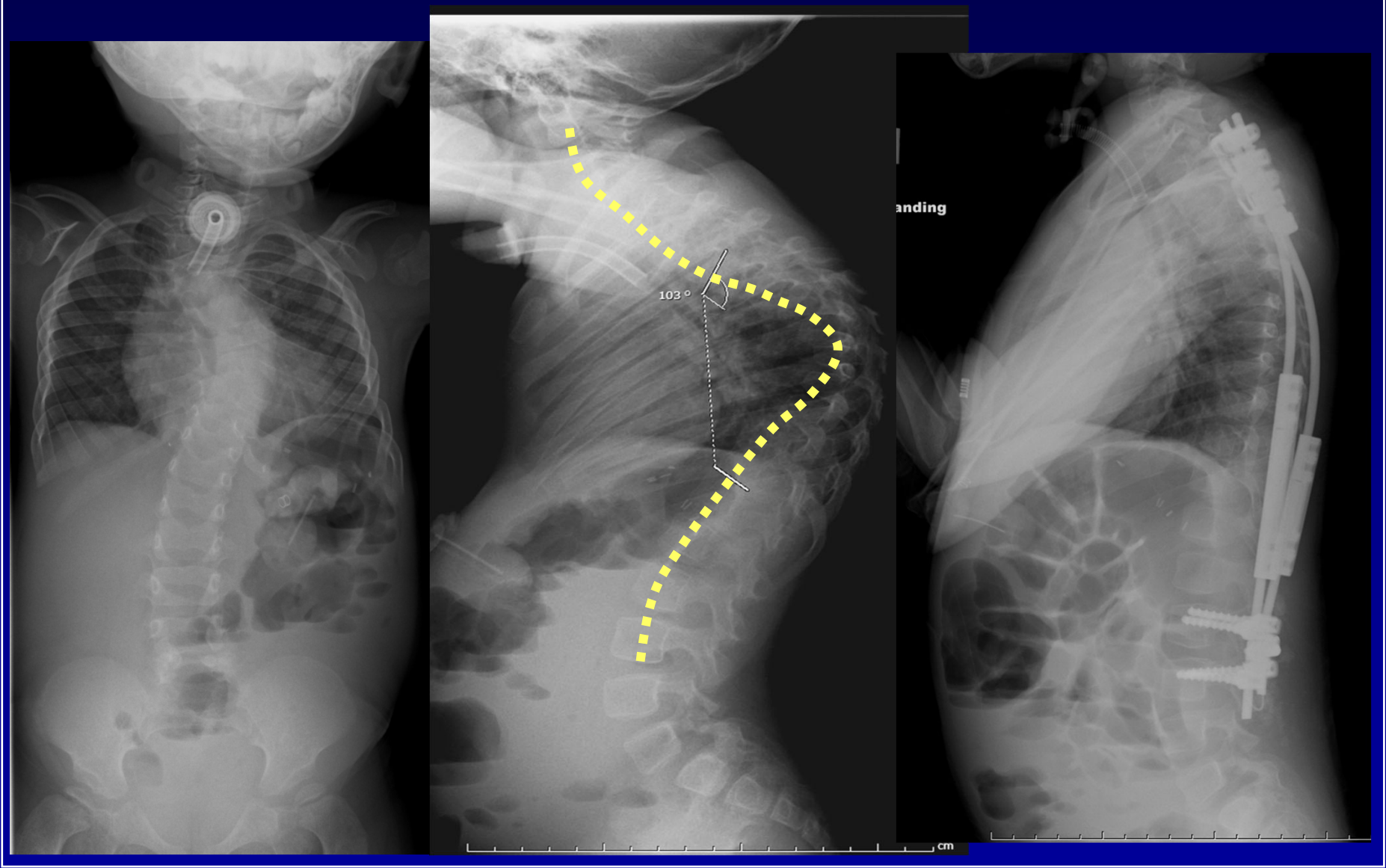


Infantile idiopathic scoliosis, moderately severe rotational chest deformity – braced until now Age 5 – too long?





Age 30 months – progressive kyphosis – don’t wait!



When to begin operation for EOS?

- *Chest-based decision-making?*
- *Use evolution of chest deformity as a guide to timing?*

- *Goals for timing operative decision*
  - Don't miss the opportunity to help lung growth and development → ***Be aggressive with chest deformity –operate early***
  - Don't do more operations than needed! → Otherwise wait
  - Don't make the operation impossibly hard → ***Control thoracic kyphosis early***

- *Actions:*