Congenital deformity Dangerous, Benign

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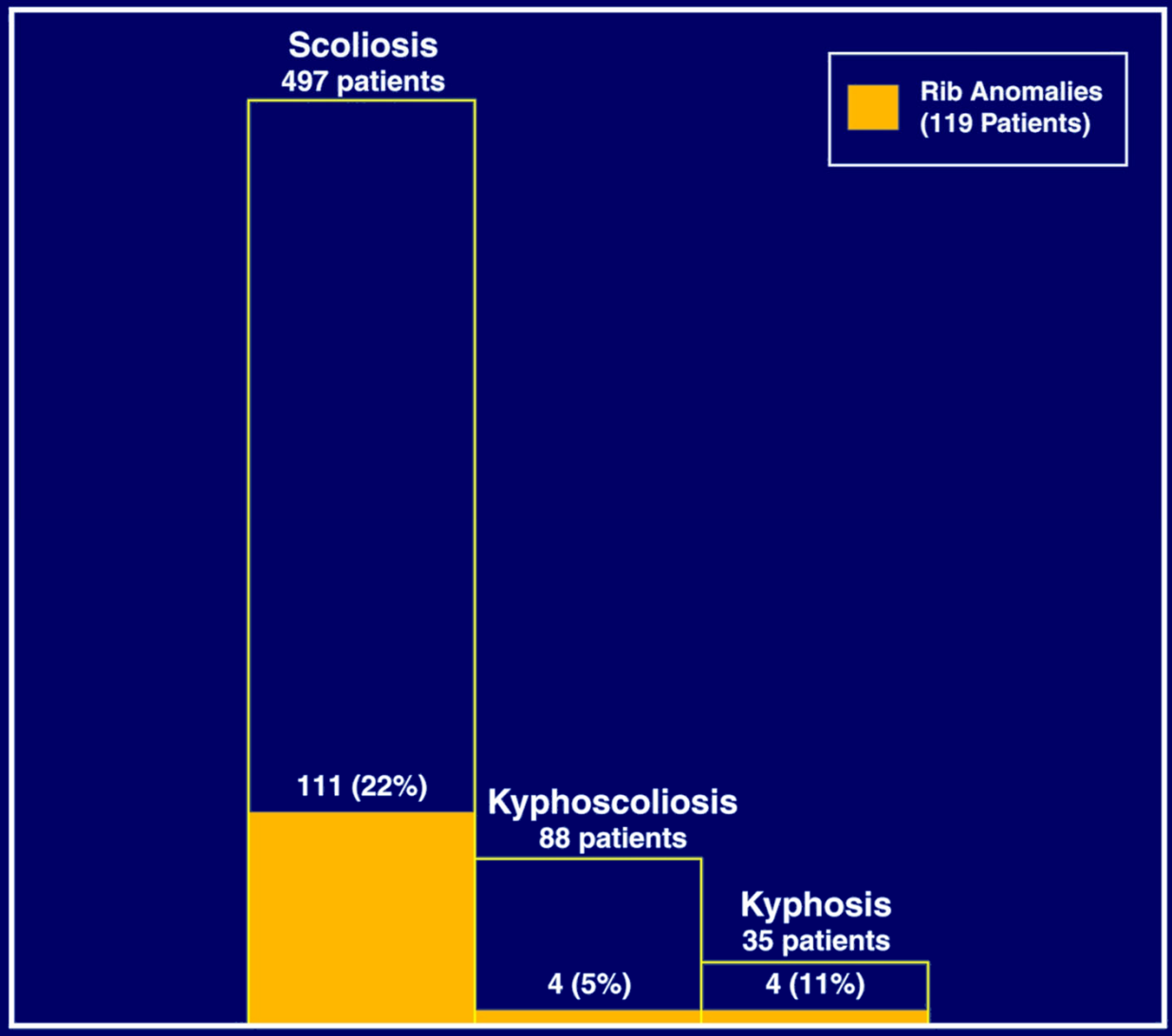
Pathogenesis

- represent a gradually blending spectrum of spinal deformities
- developmental anomalies producing a localized imbalance in the longitudinal growth of the spine
- congenital scoliosis: during the first 6 weeks of intrauterine life (mesenchymal stage)

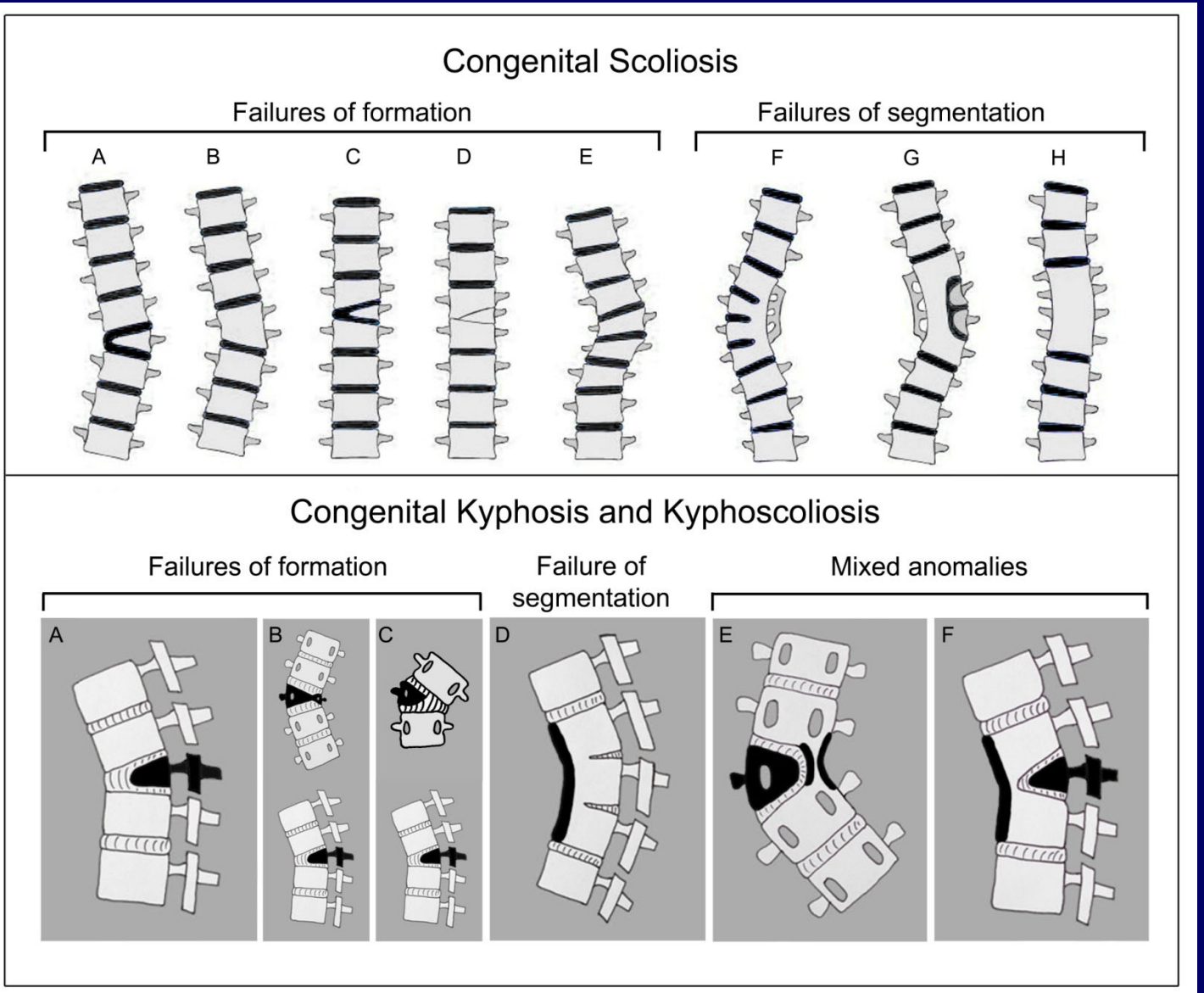
Pathogenesis

- congenital kyphosis: during the later stages of chondrification and ossification
- failure of formation: due to localized failure of vascularization of the developing cartilaginous centrum
- failure of segmentation: due to bony metaplasia occurring in the anterior part of the annulus fibrosus and ring apophysis

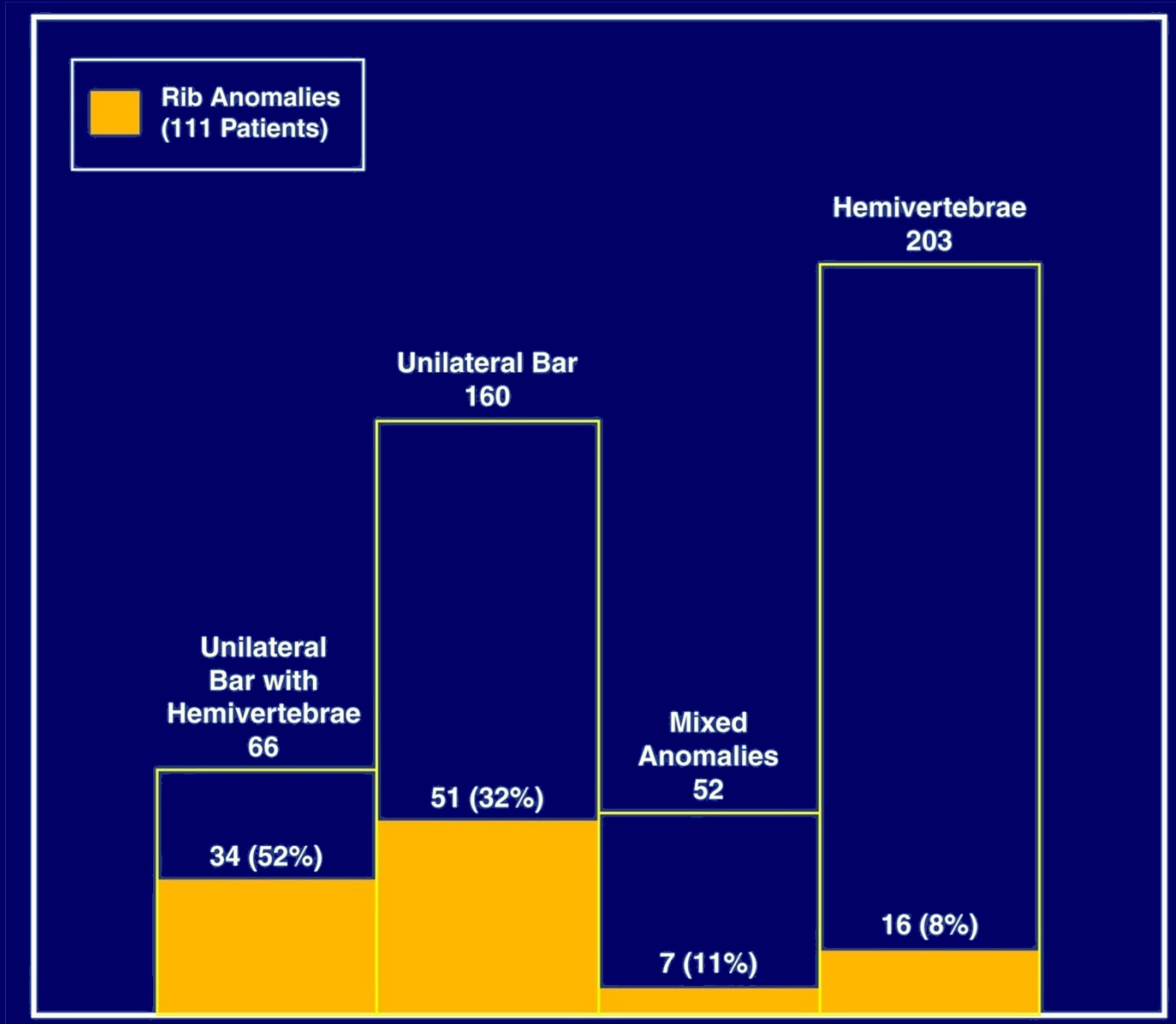
Type of spinal deformity



Classification

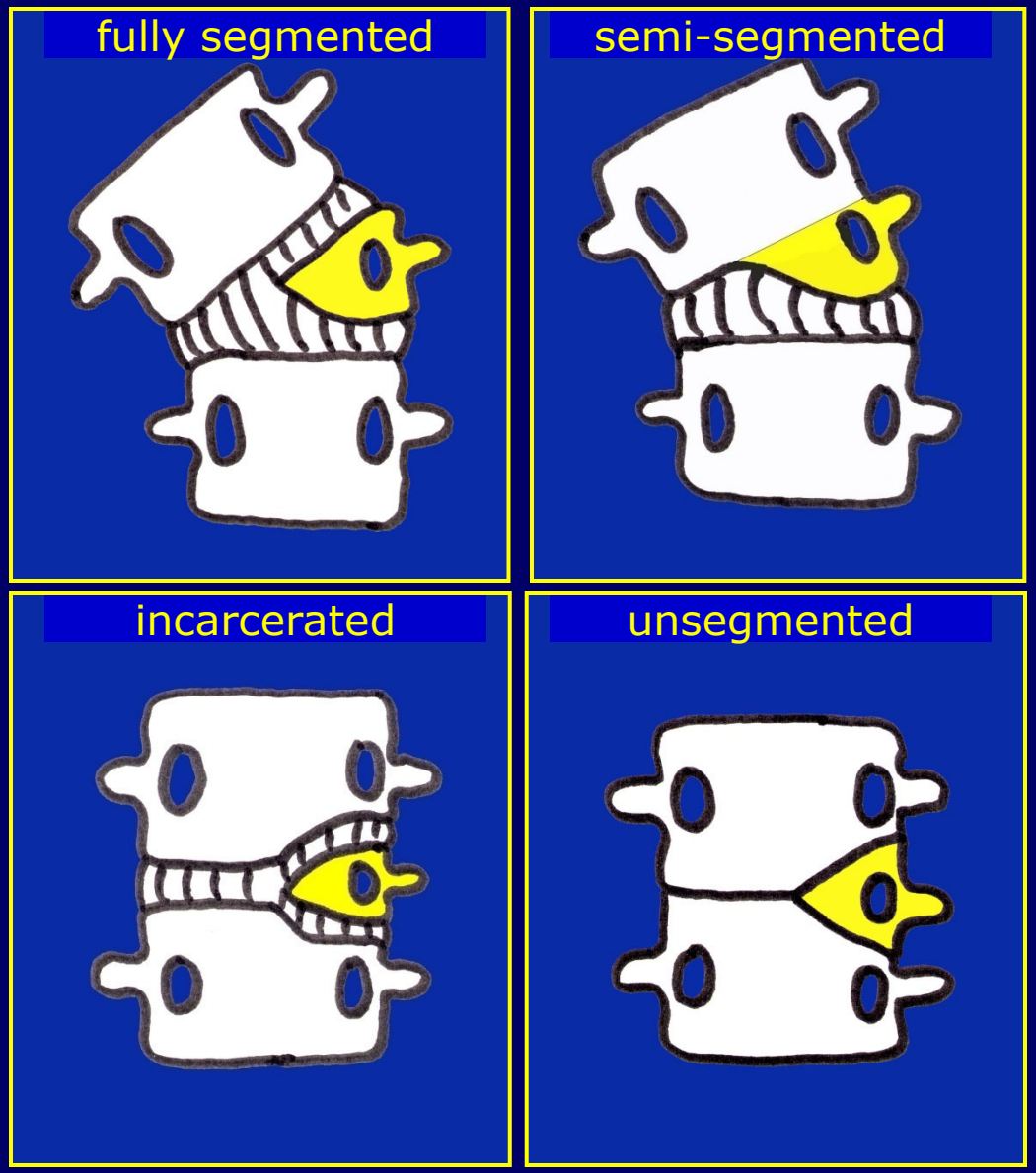


CONGENITAL SCOLIOSIS (481 patients)

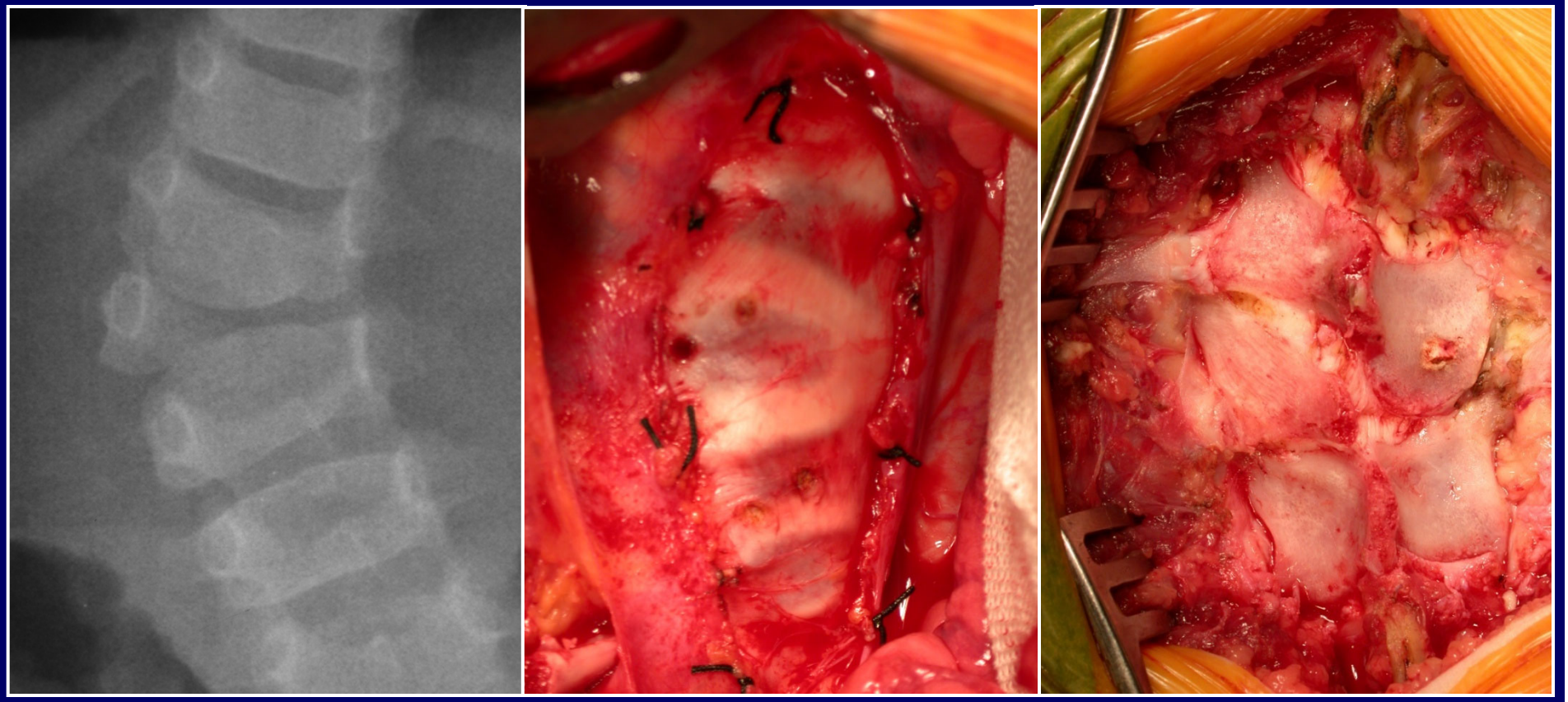


Hemivertebra-prognosis

- pathologic anatomy
- number and relationship to the adjacent vertebrae
- site

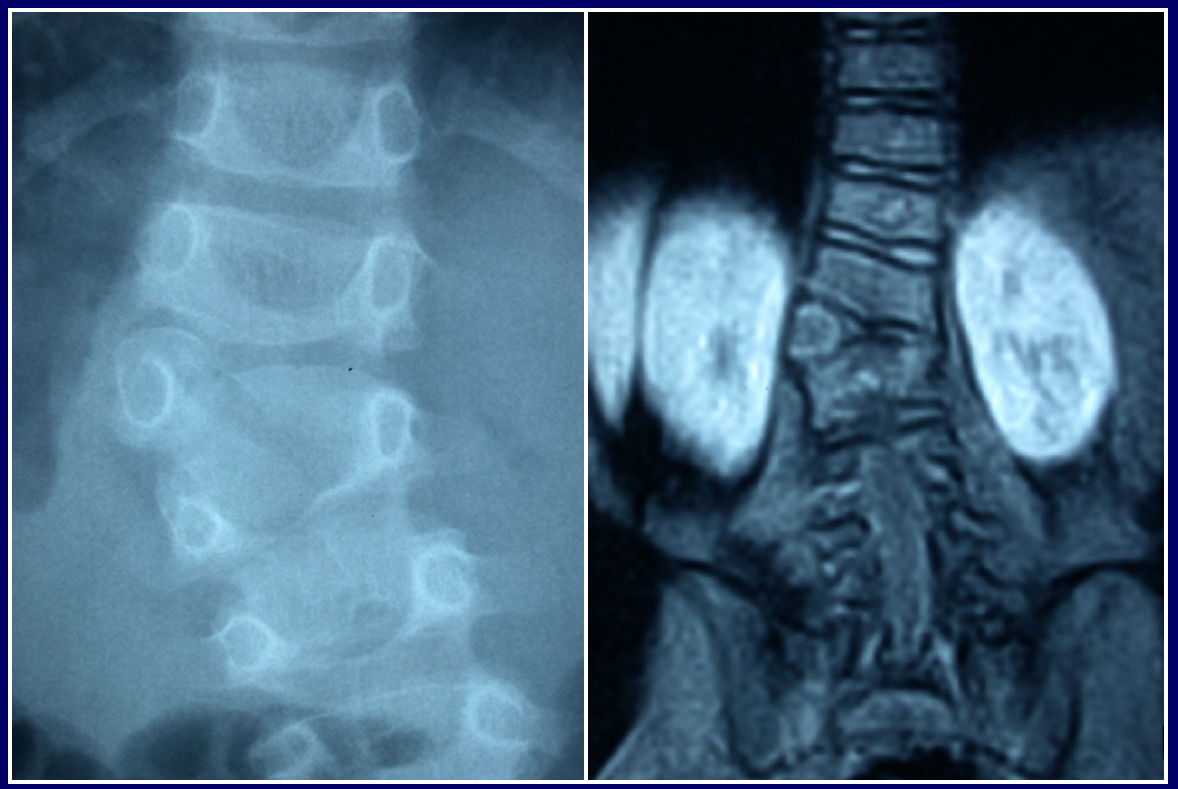


Fully segmented hemivertebra (65%)

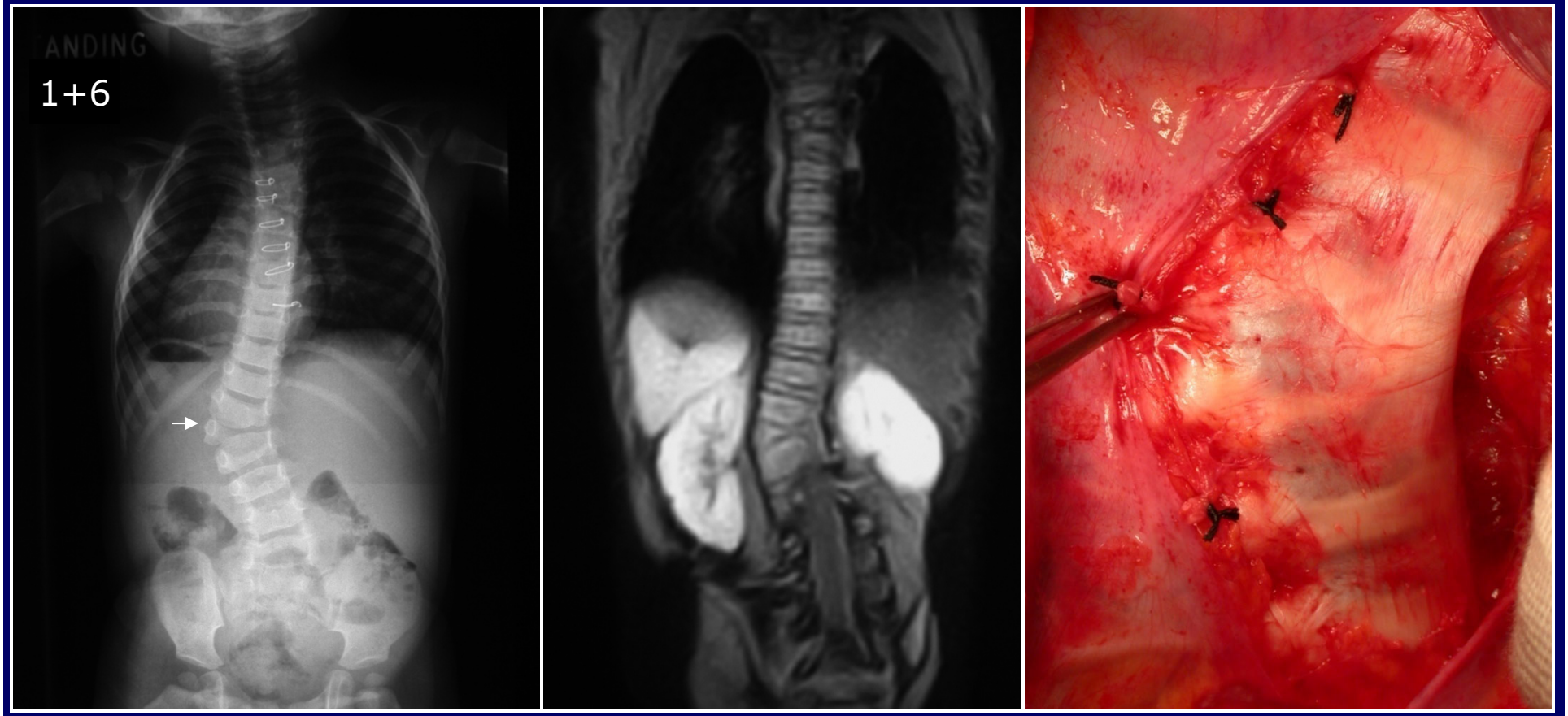


Single fully segmented hemivertebra

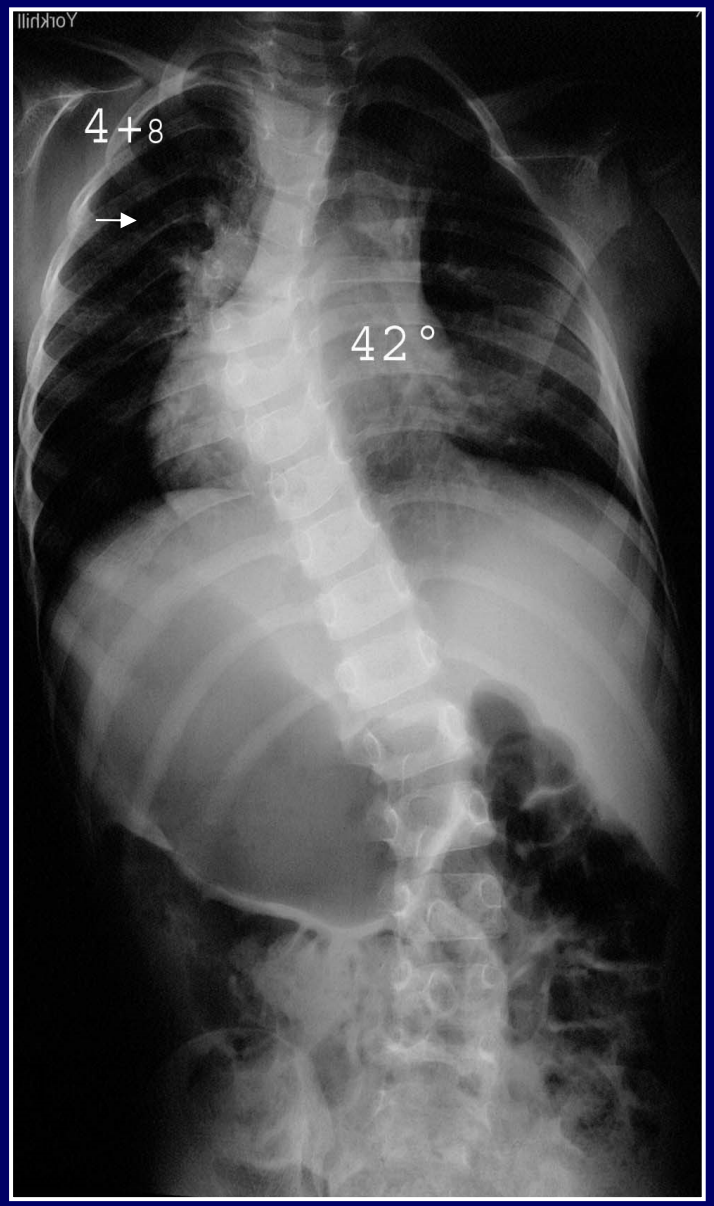
- prognosis is difficult to predict
- scoliosis progresses at a mean rate of 1-2°/year

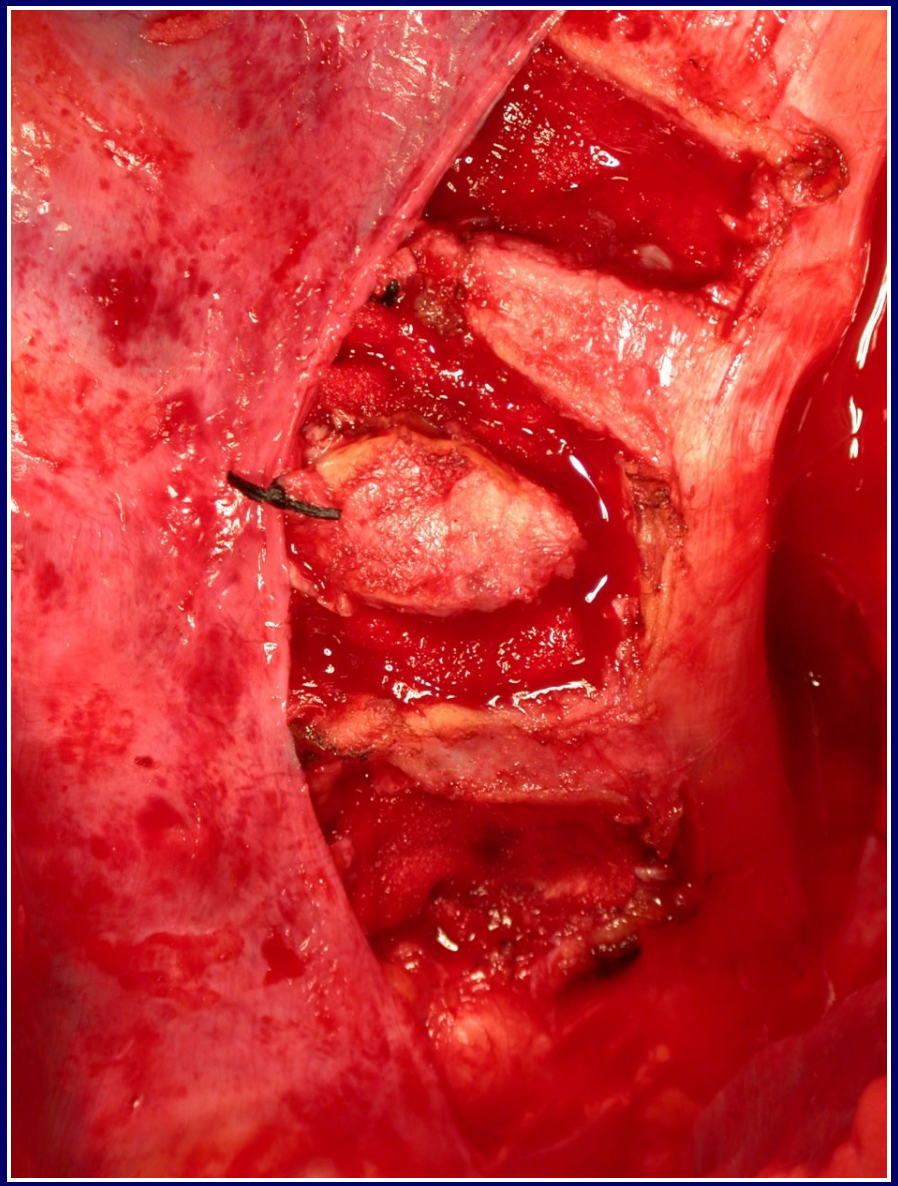


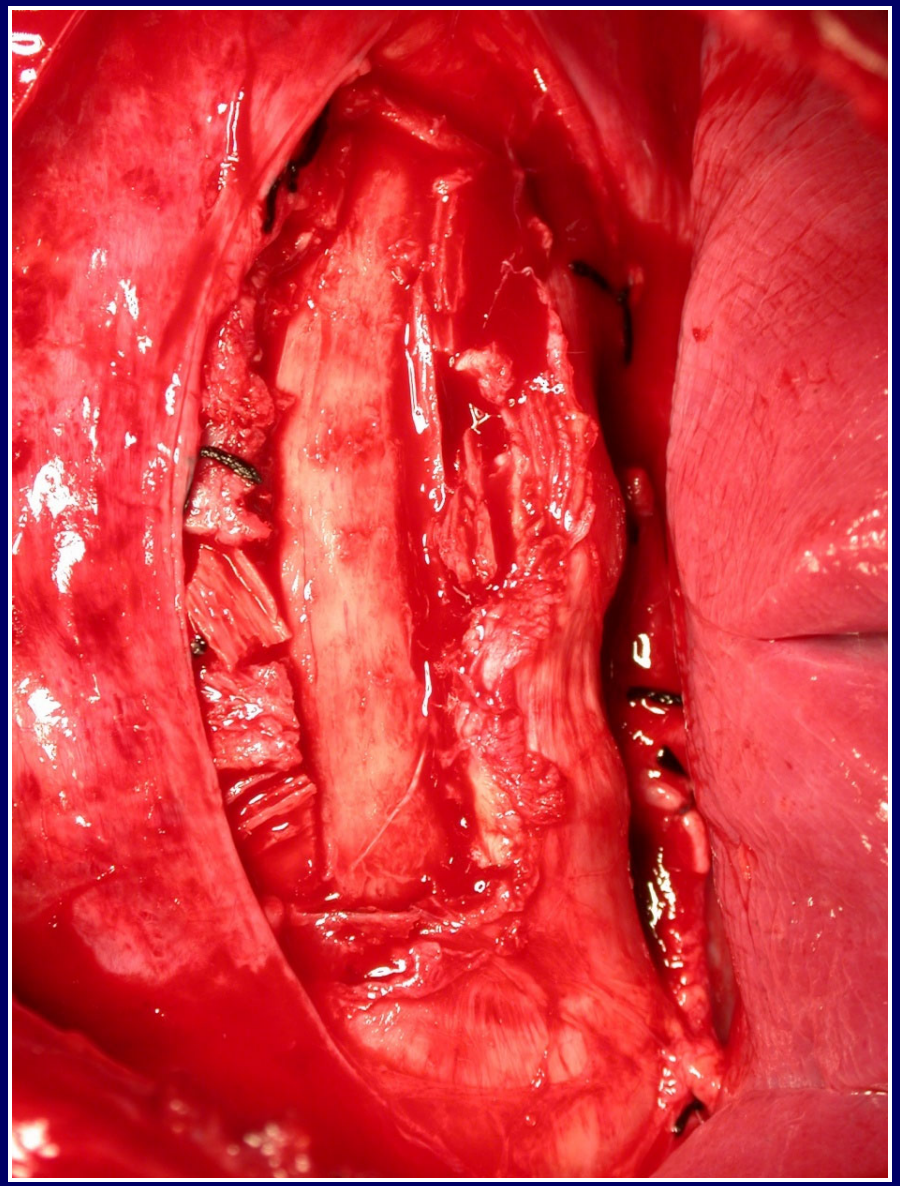
Fully segmented hemivertebra

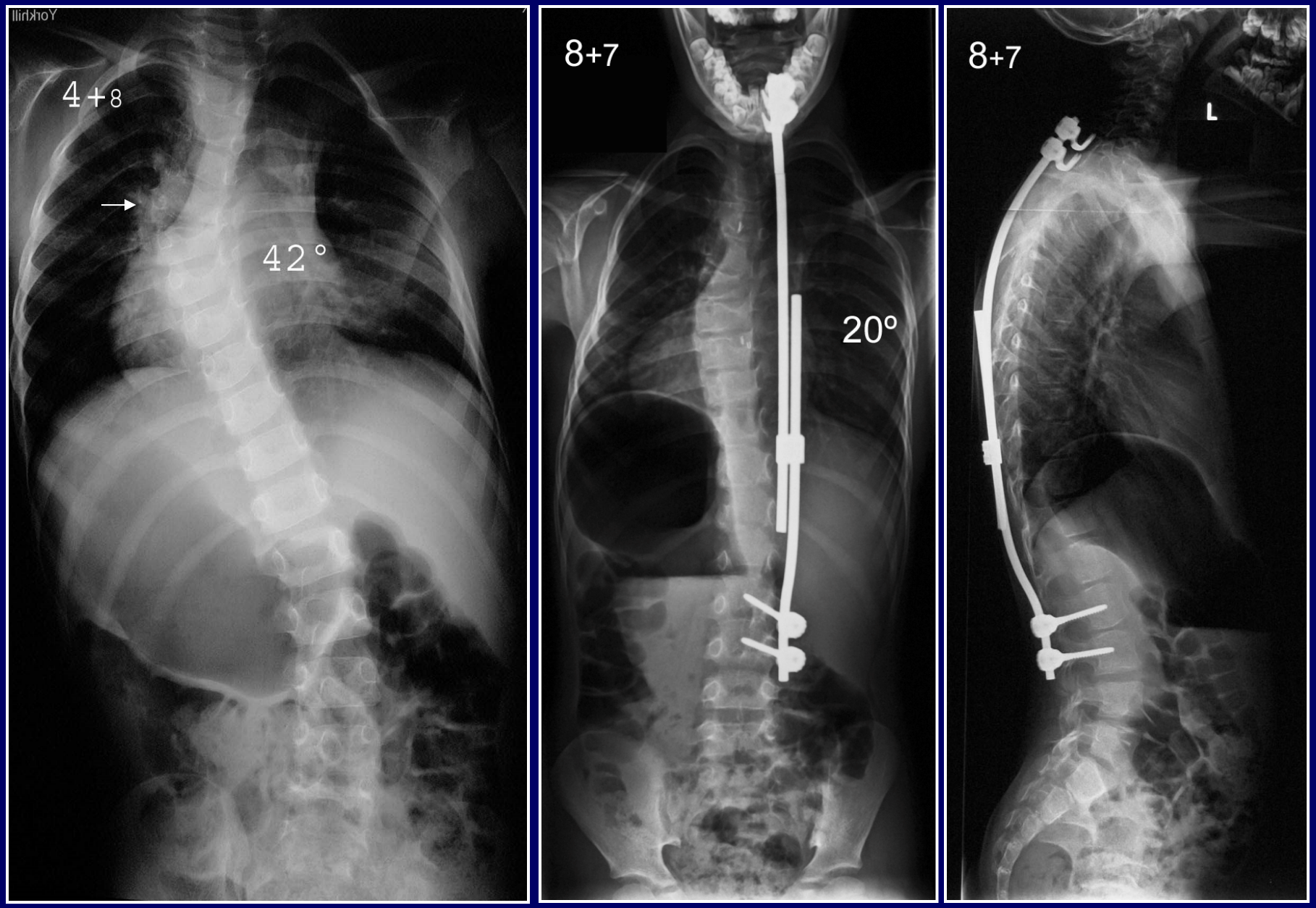


Convex growth arrest procedure

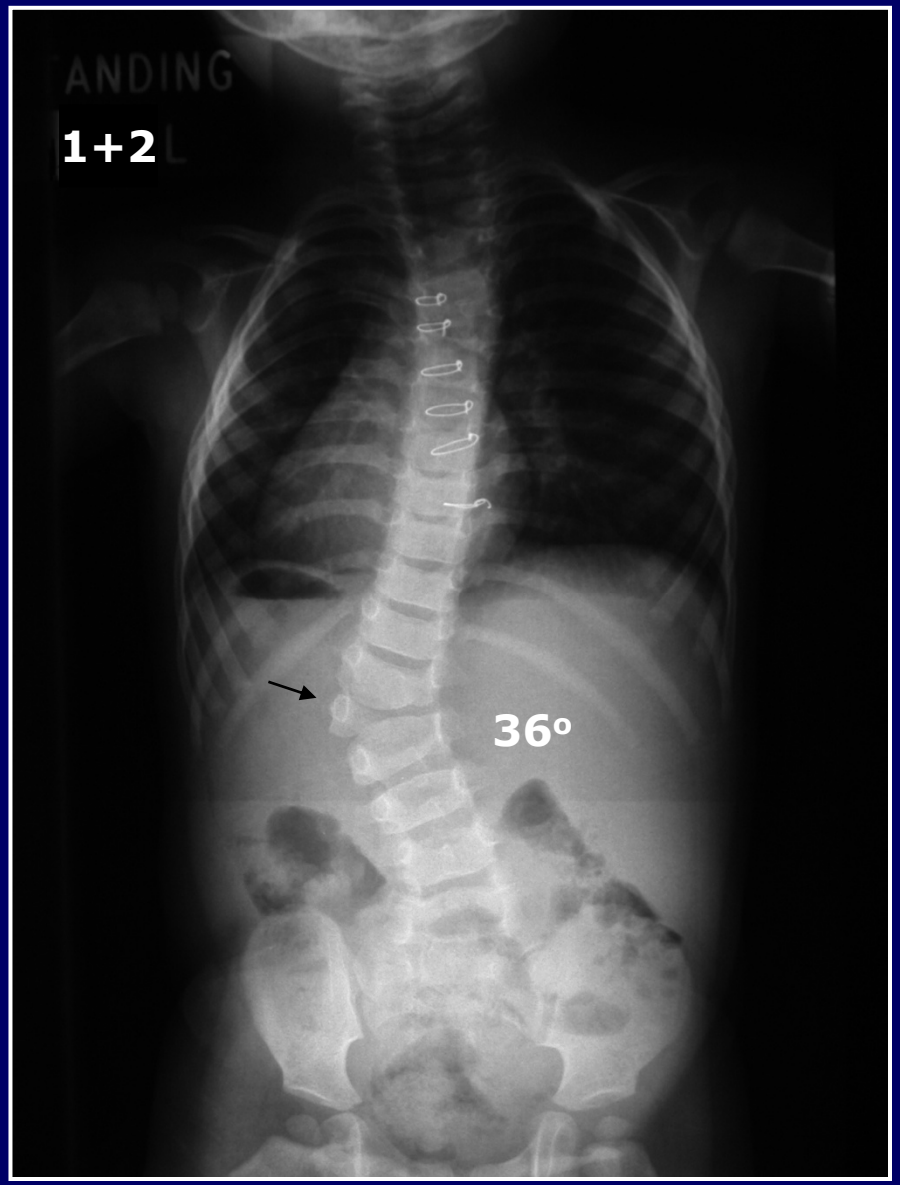


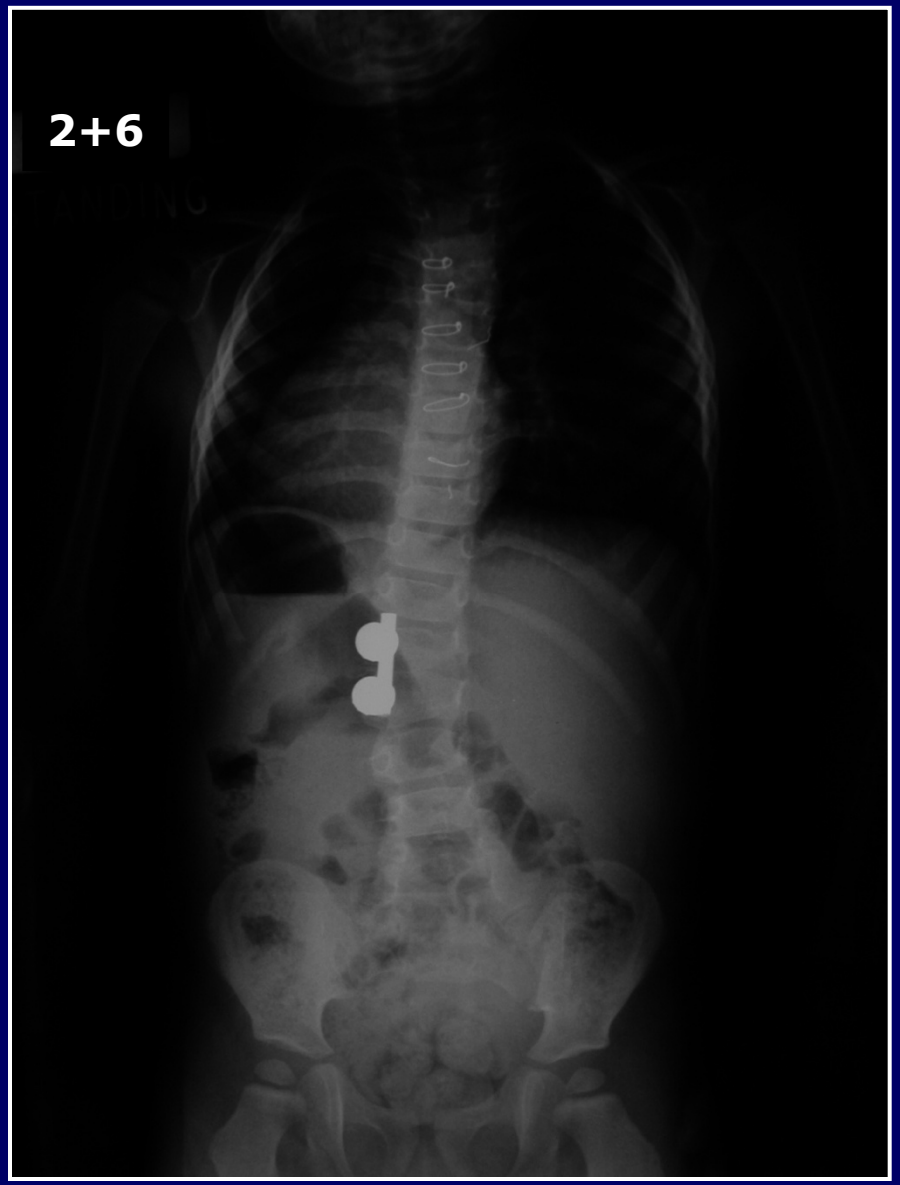





4+8
42°
8+7
20°
8+7
L

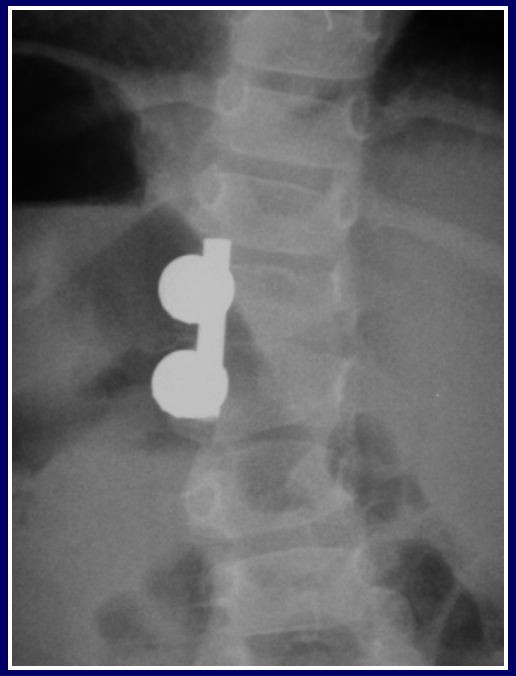
Hemivertebra resection



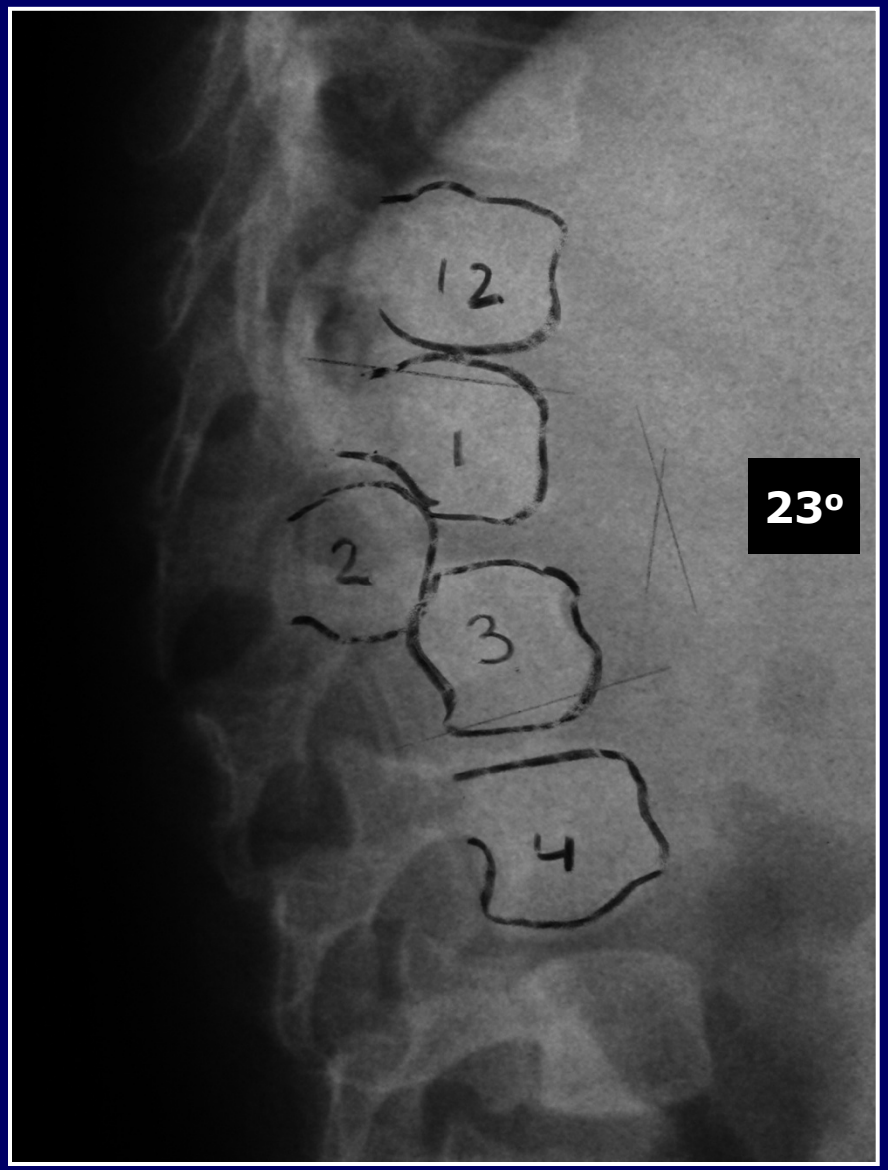


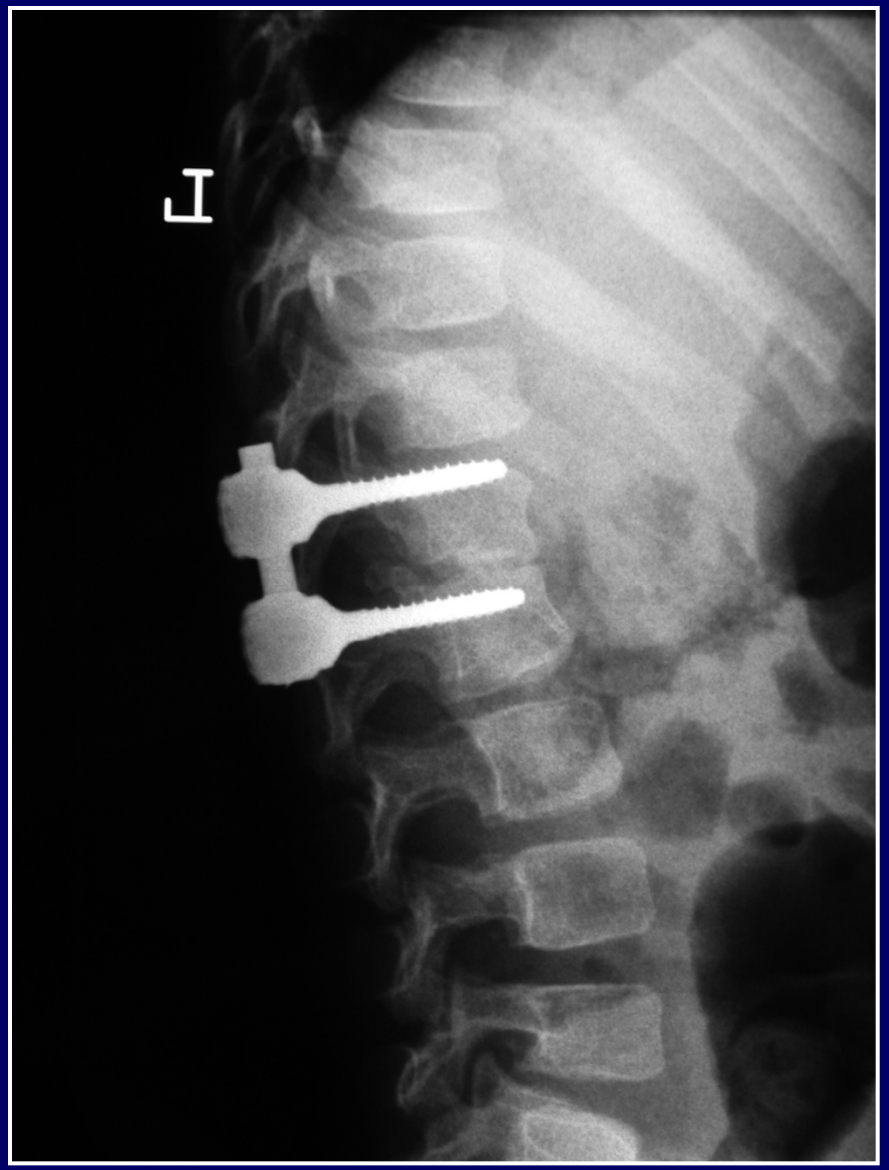




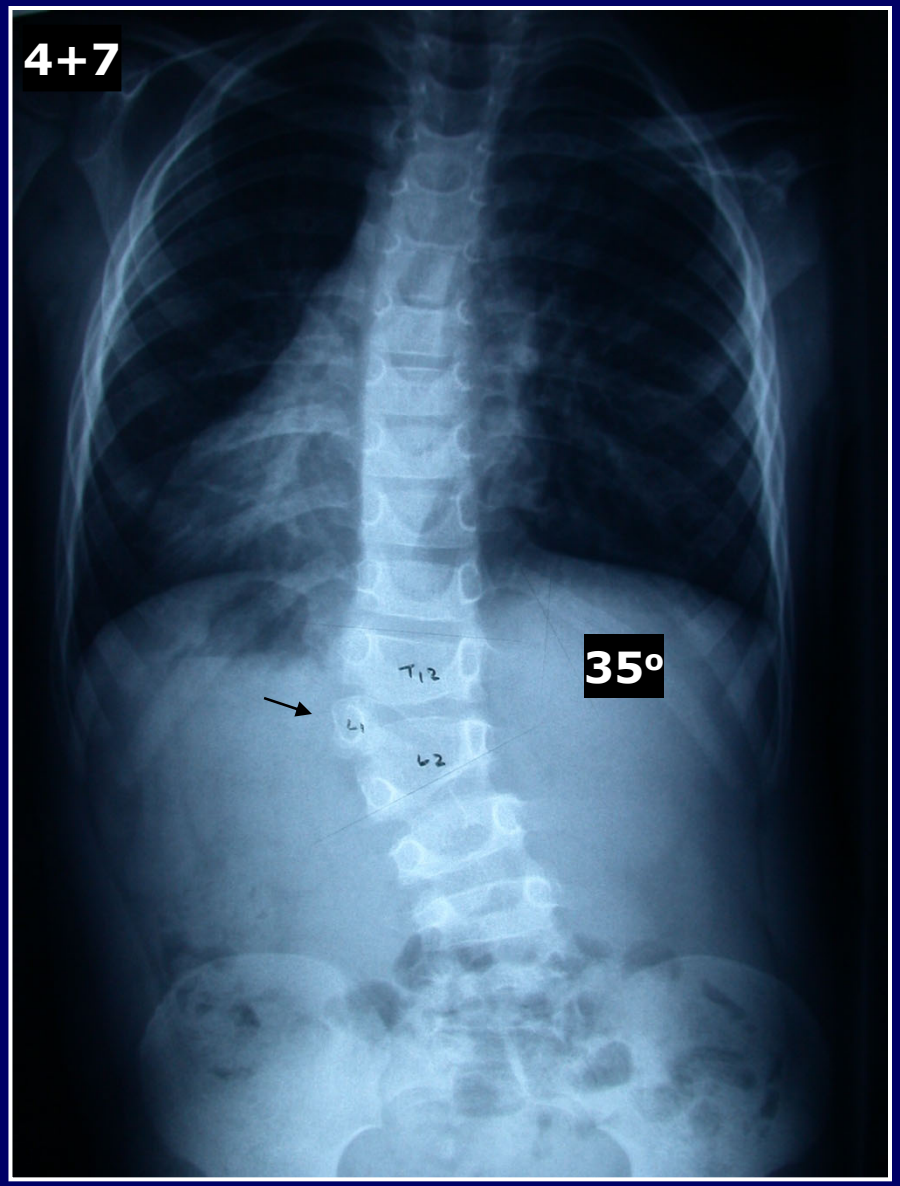


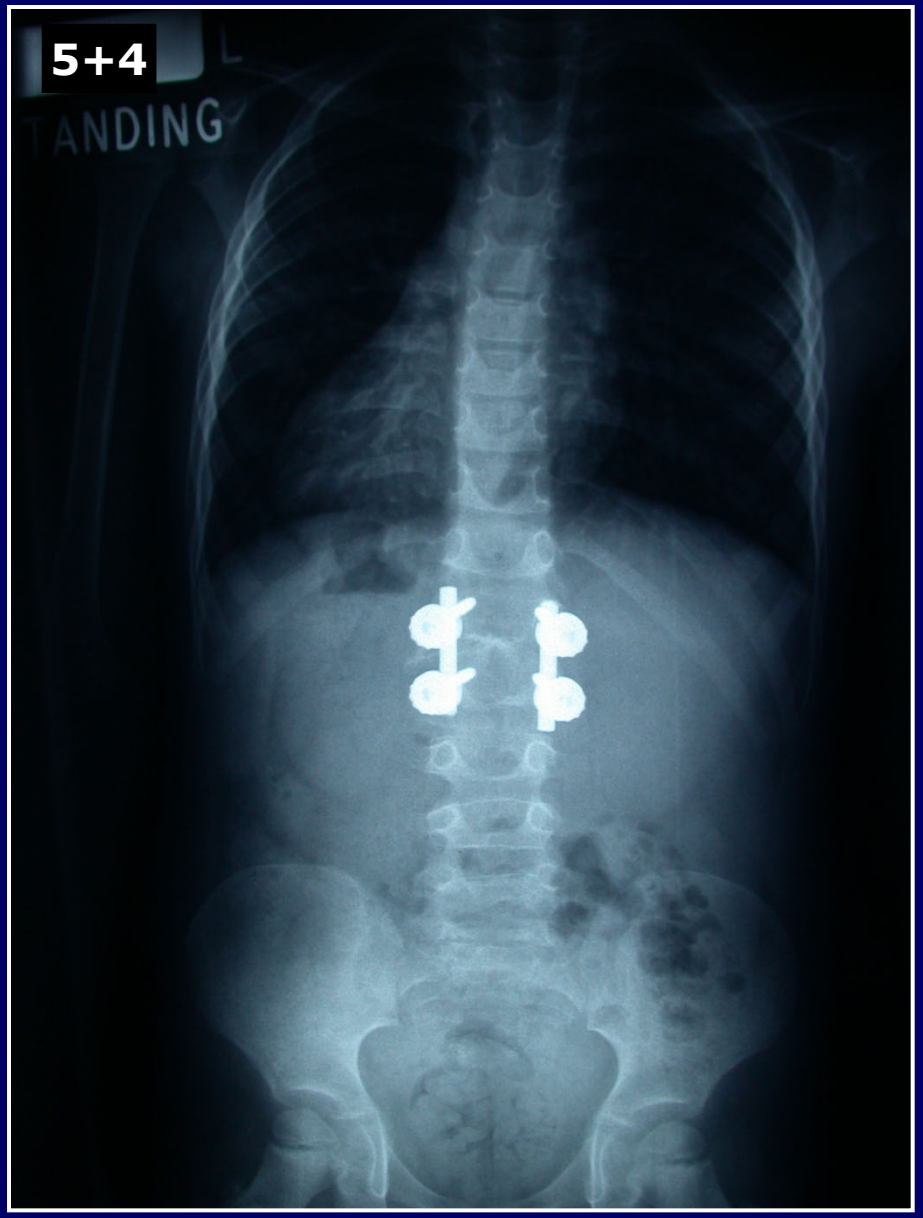
Hemivertebra resection

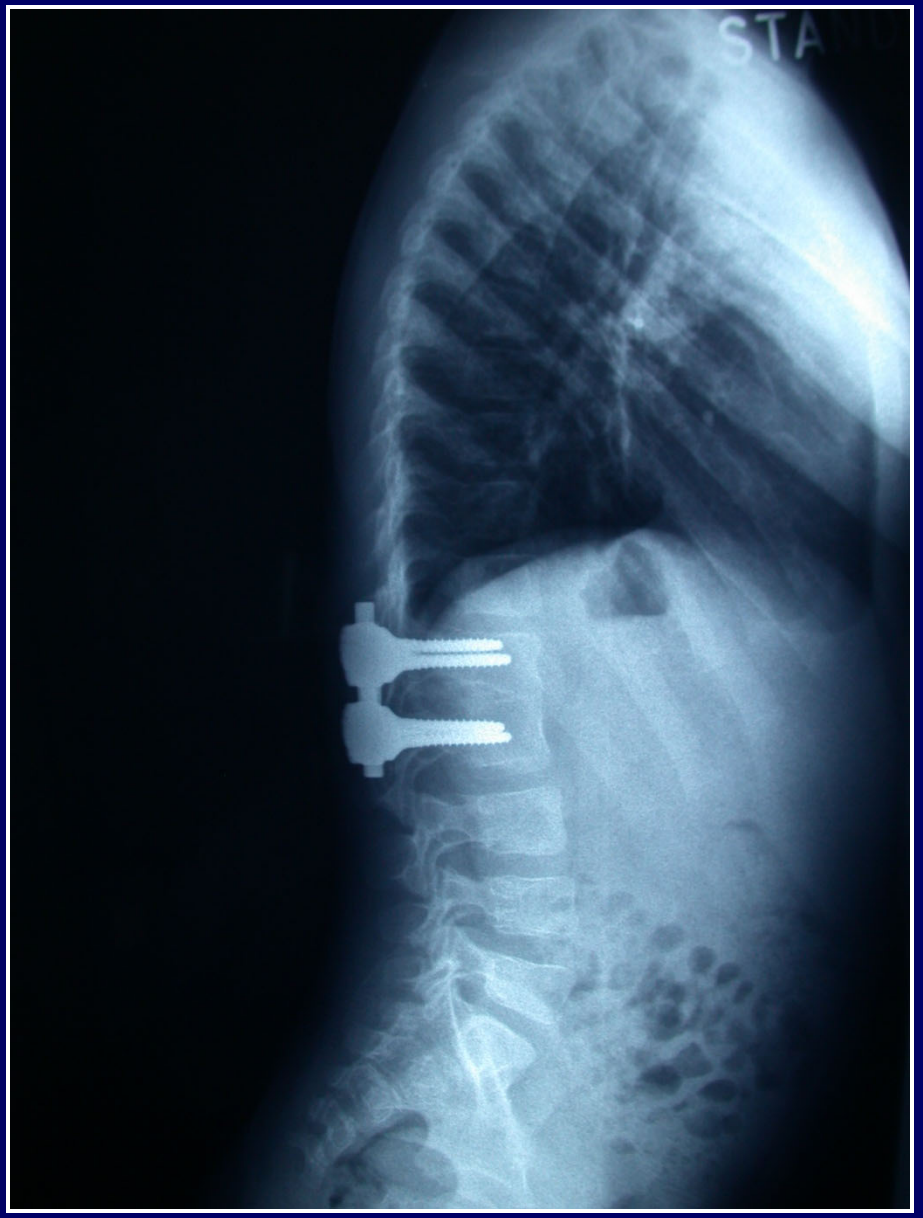




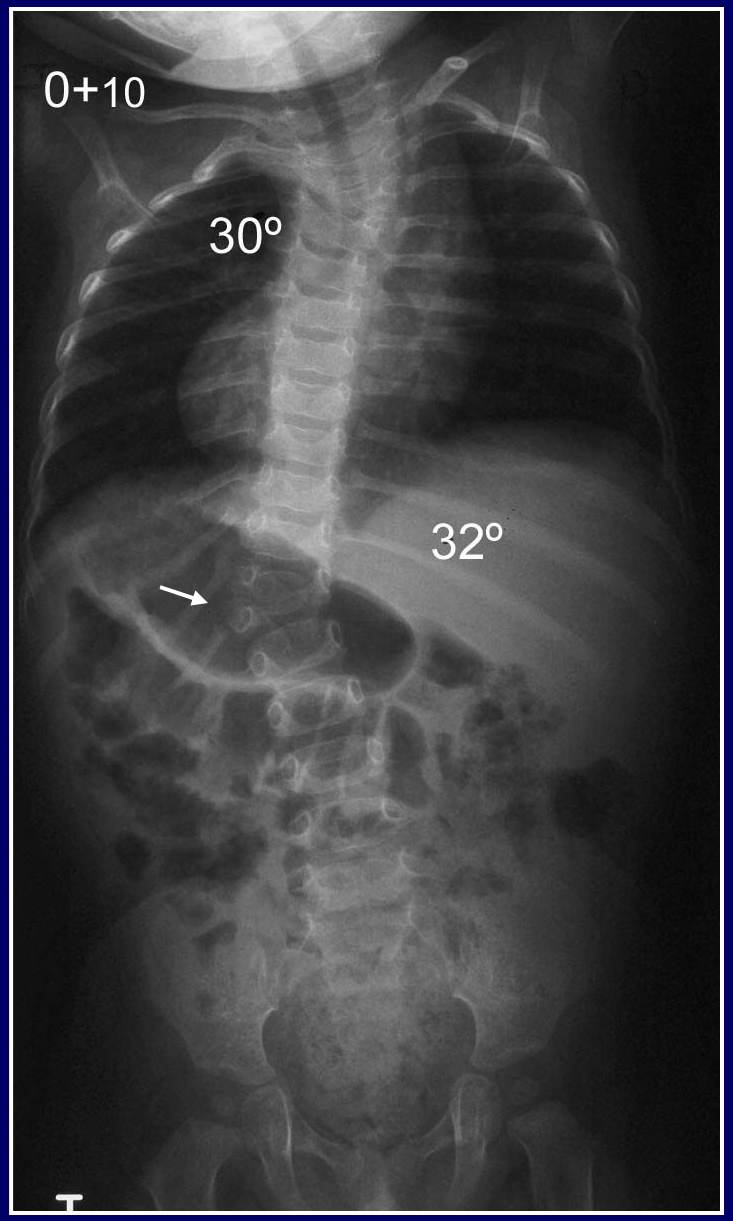
Hemivertebra resection

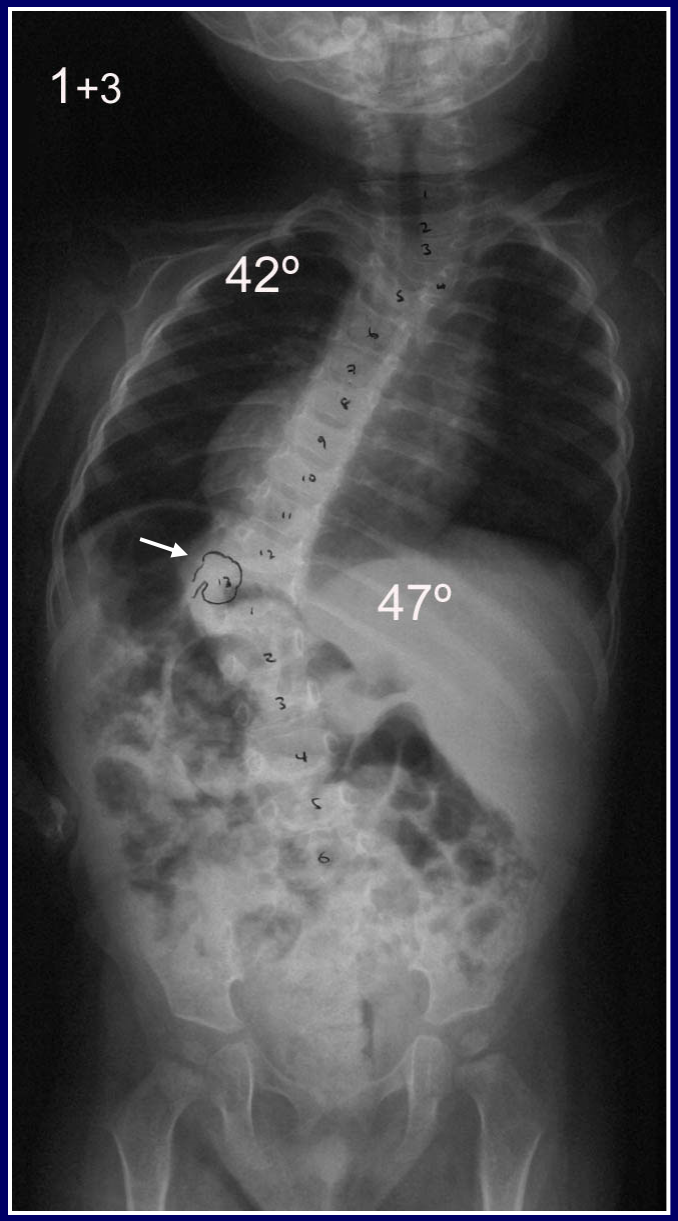


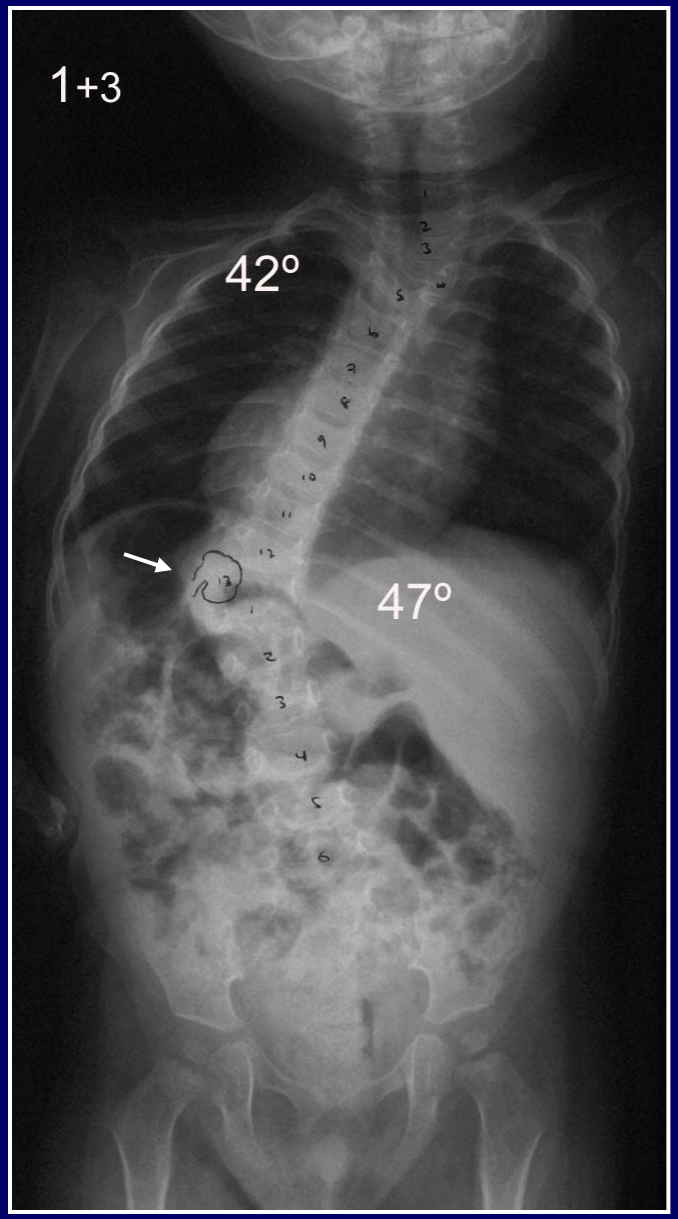

5+4
L
TANDING

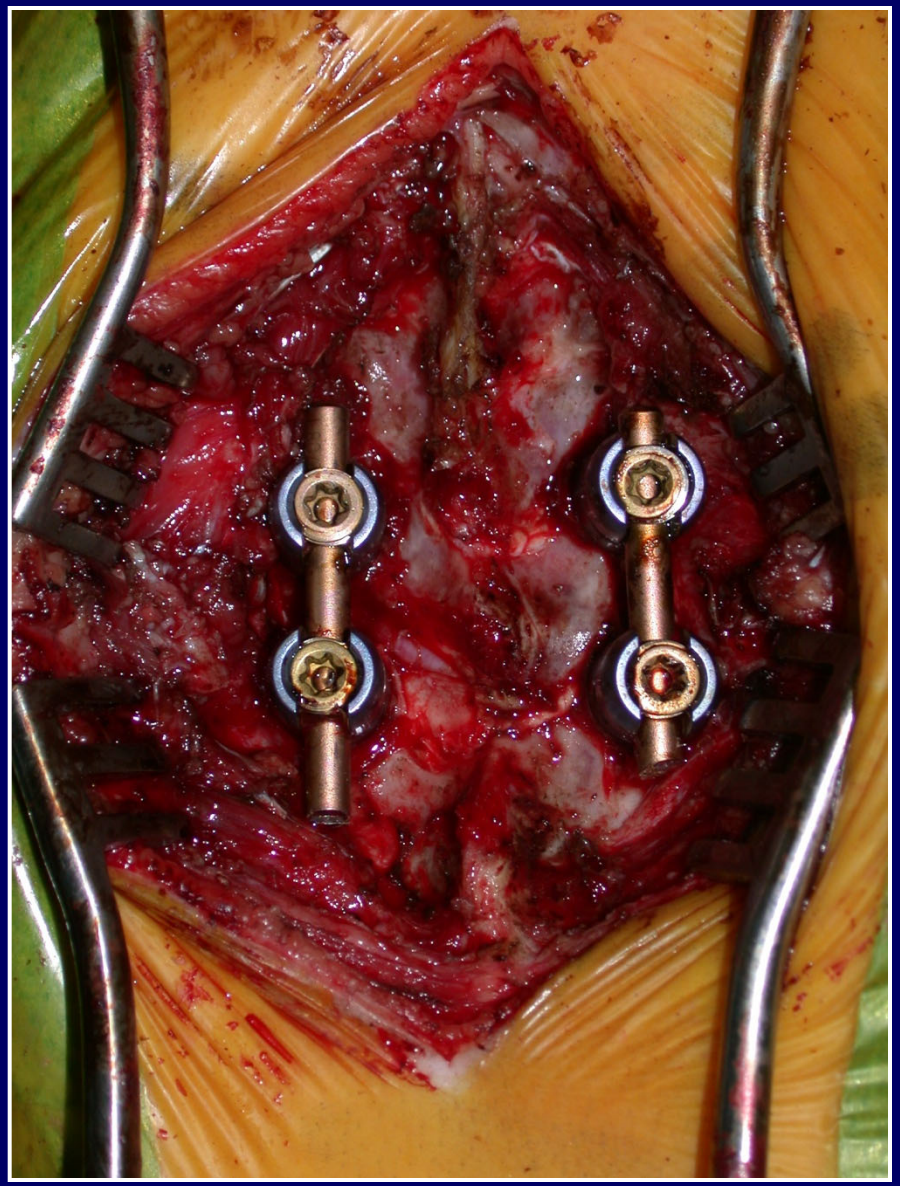

STA

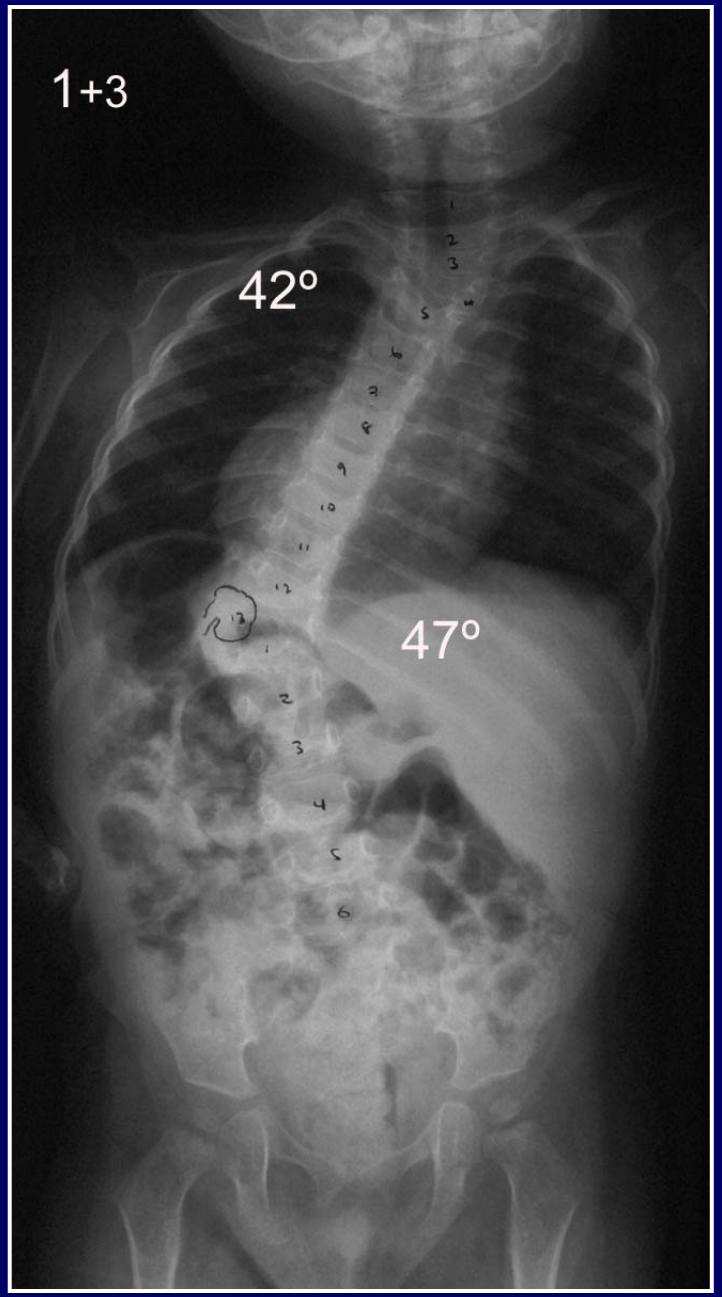
Complex vertebral anomalies

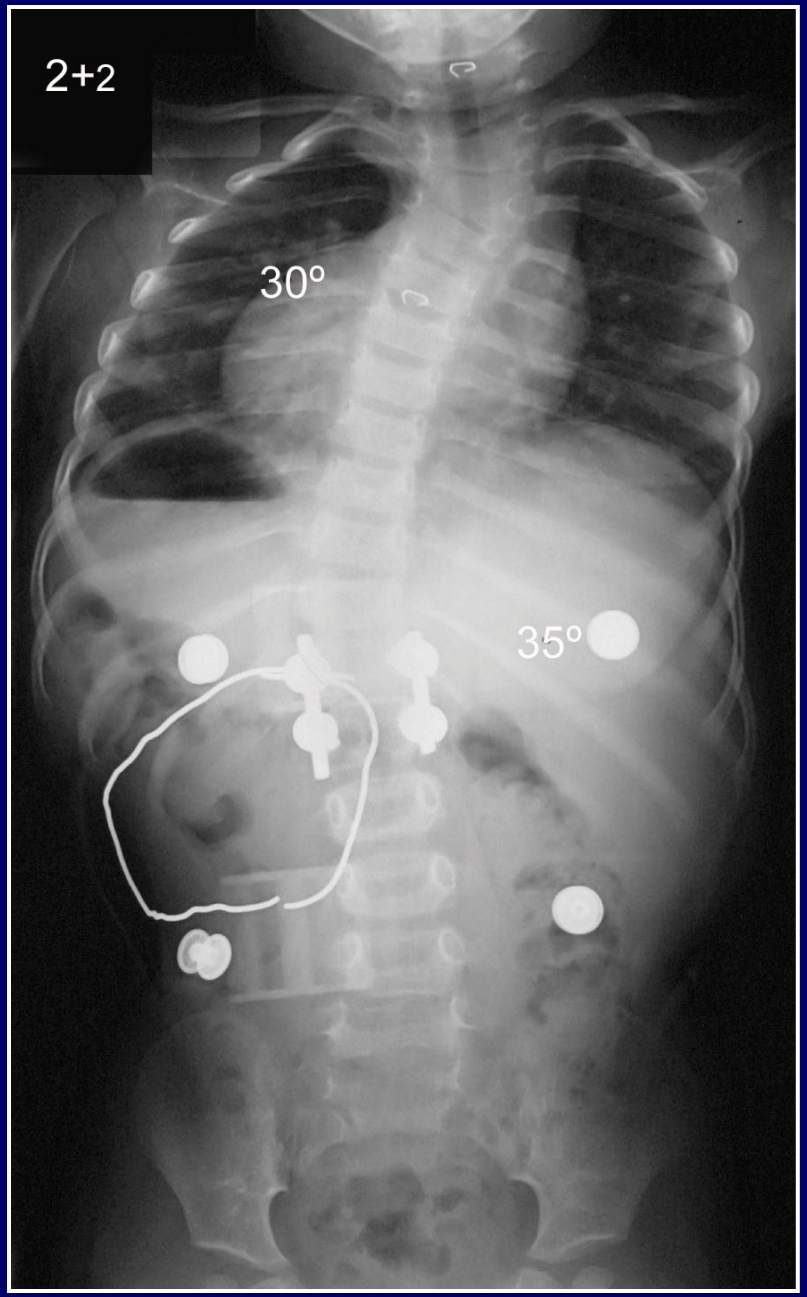




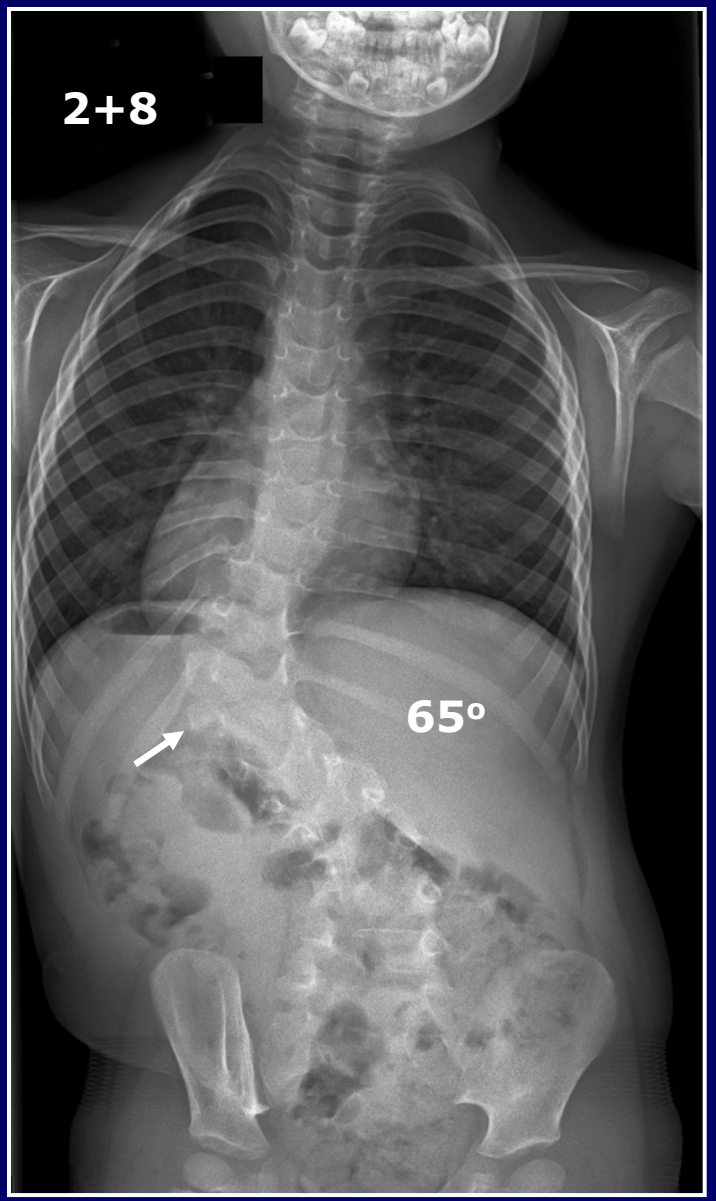

1+3
42°
47°

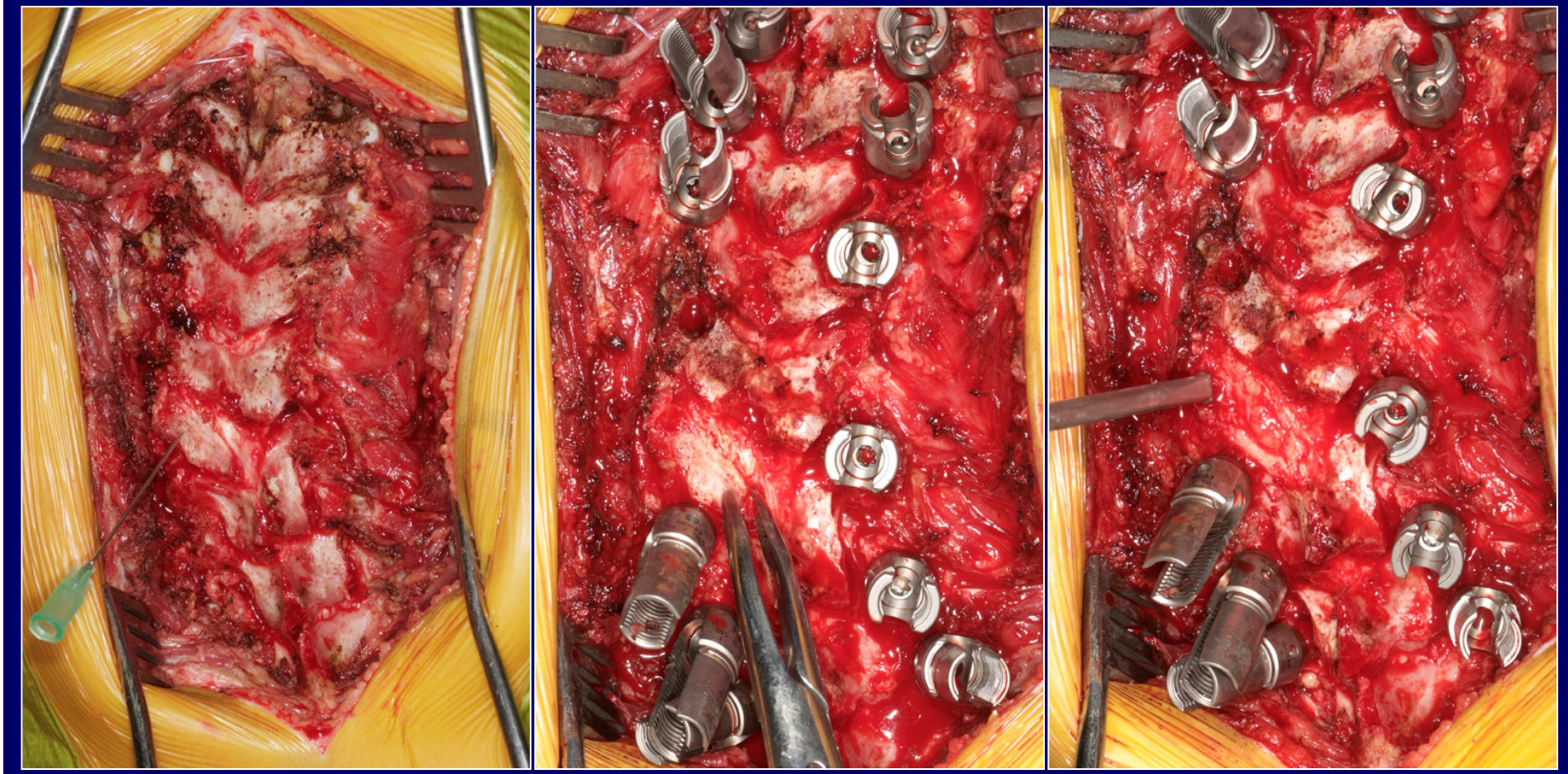


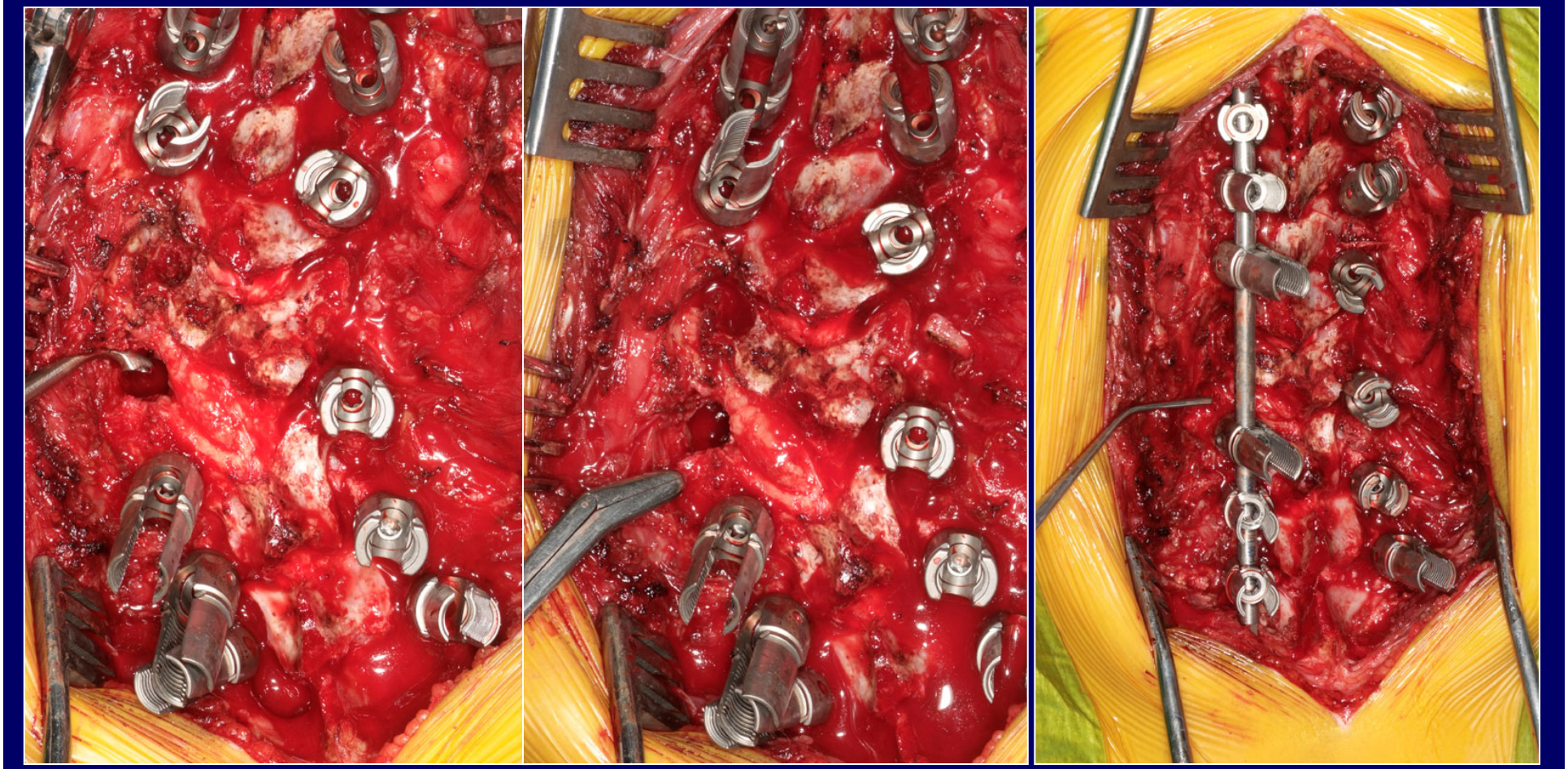

1+3
42º
47º

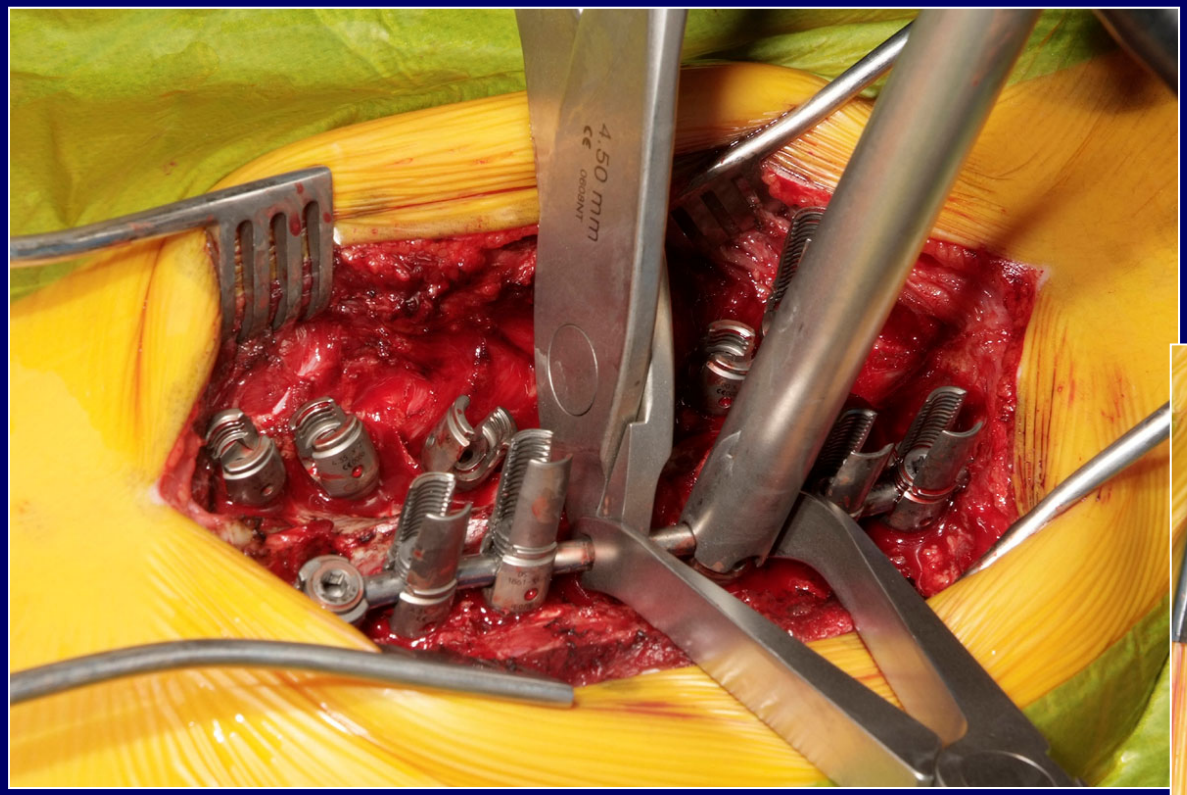

2+2
30º
35º

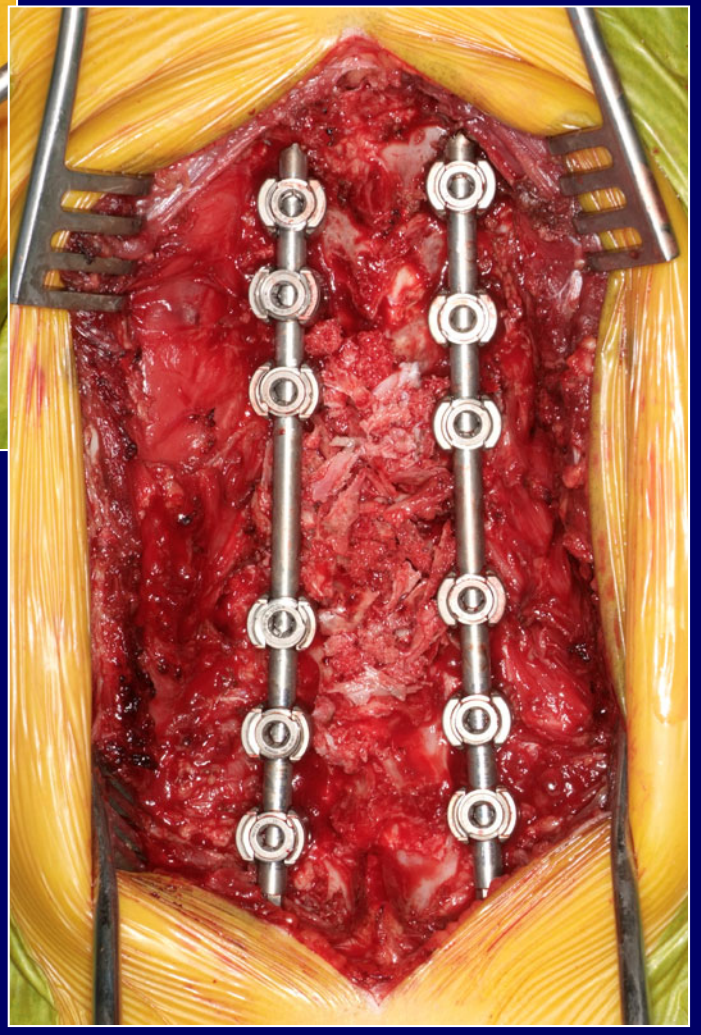
Hemivertebra resection

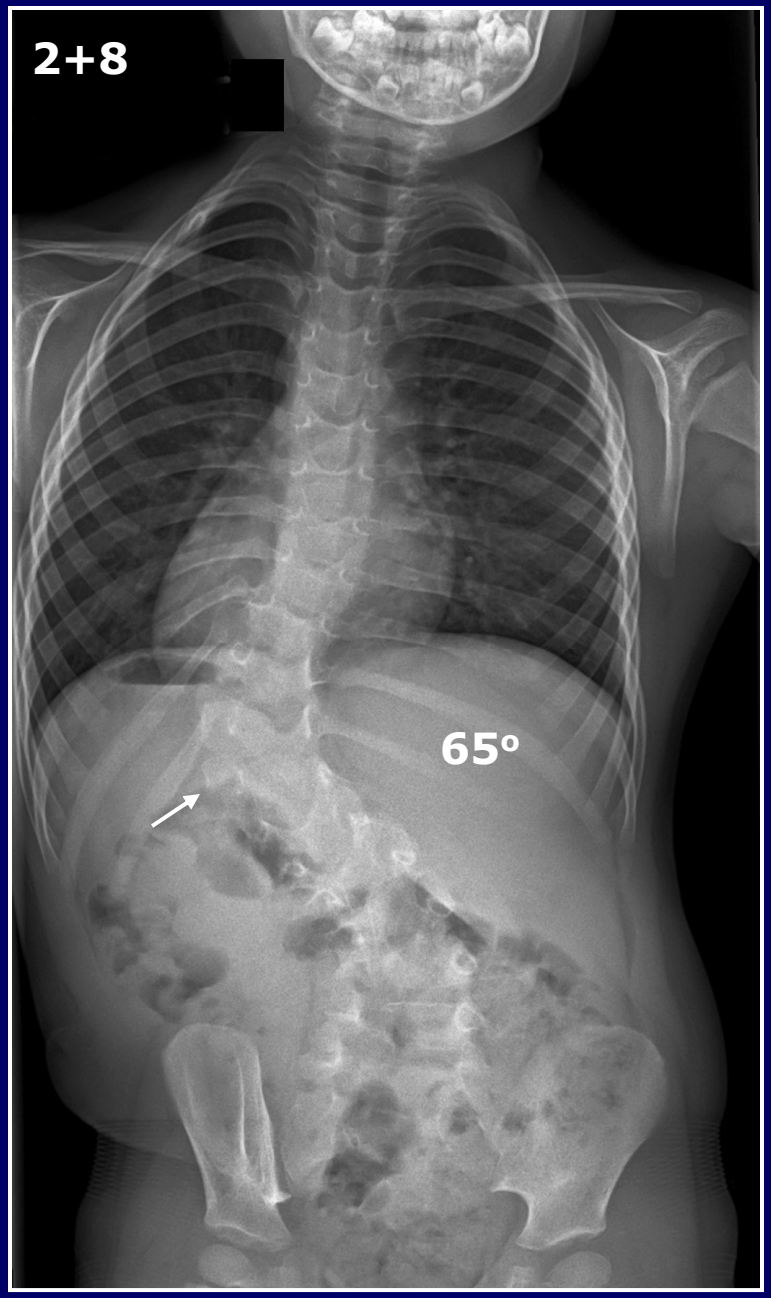


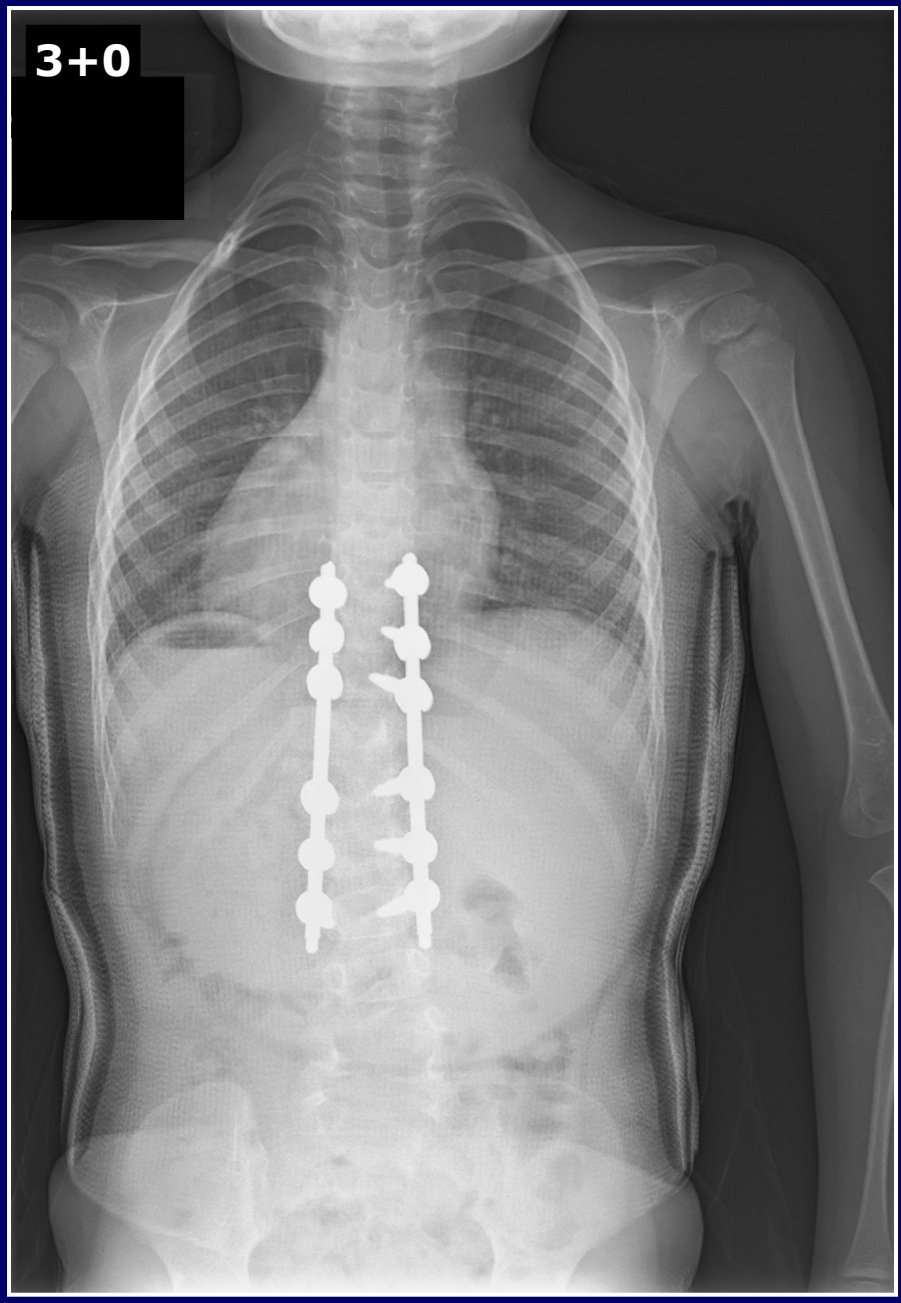


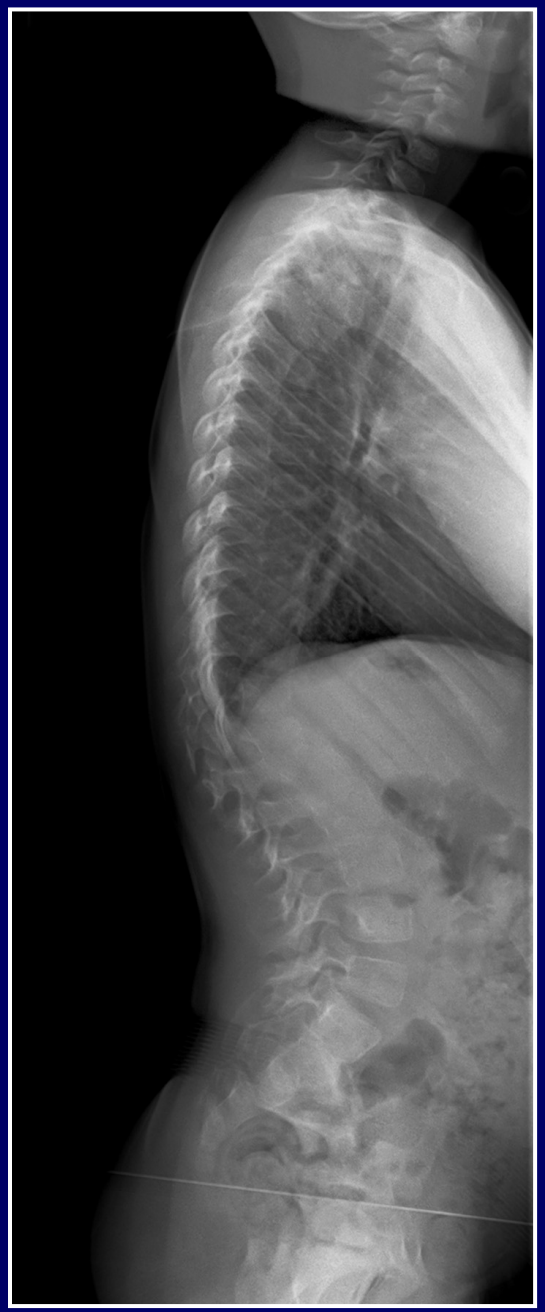


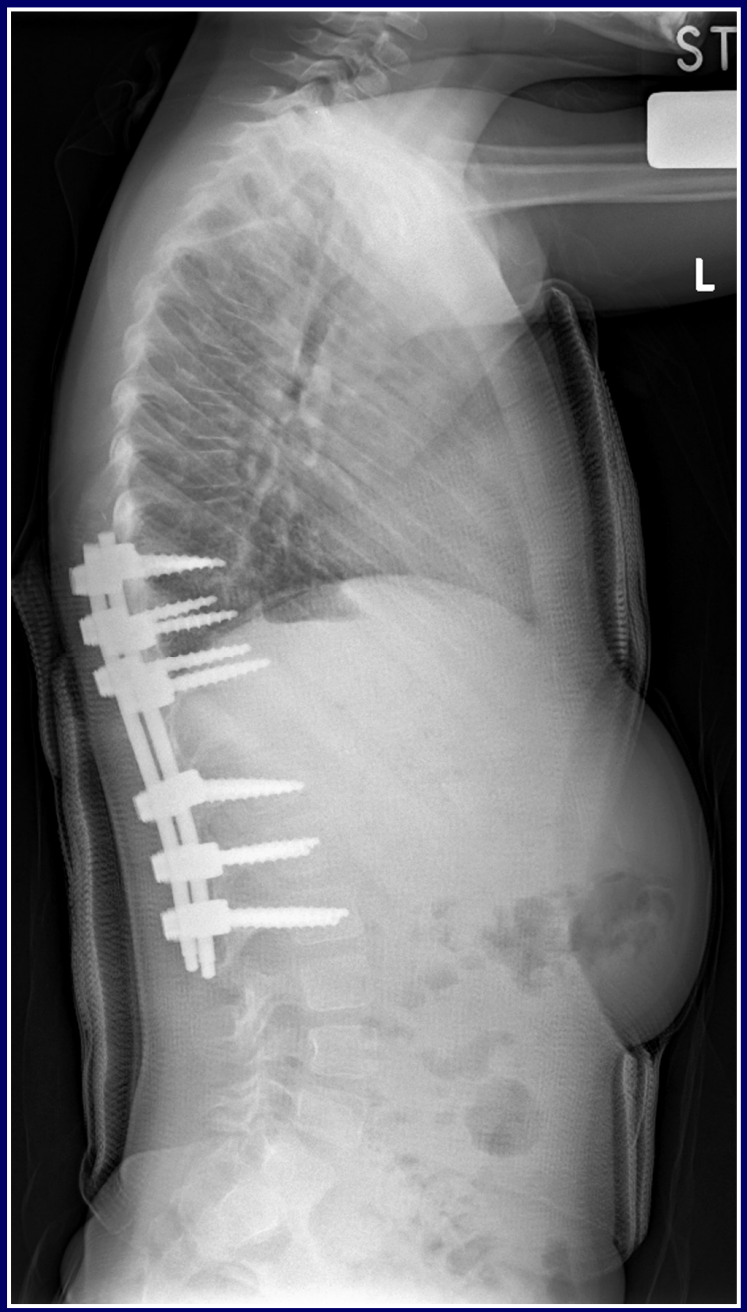

4.50 mm



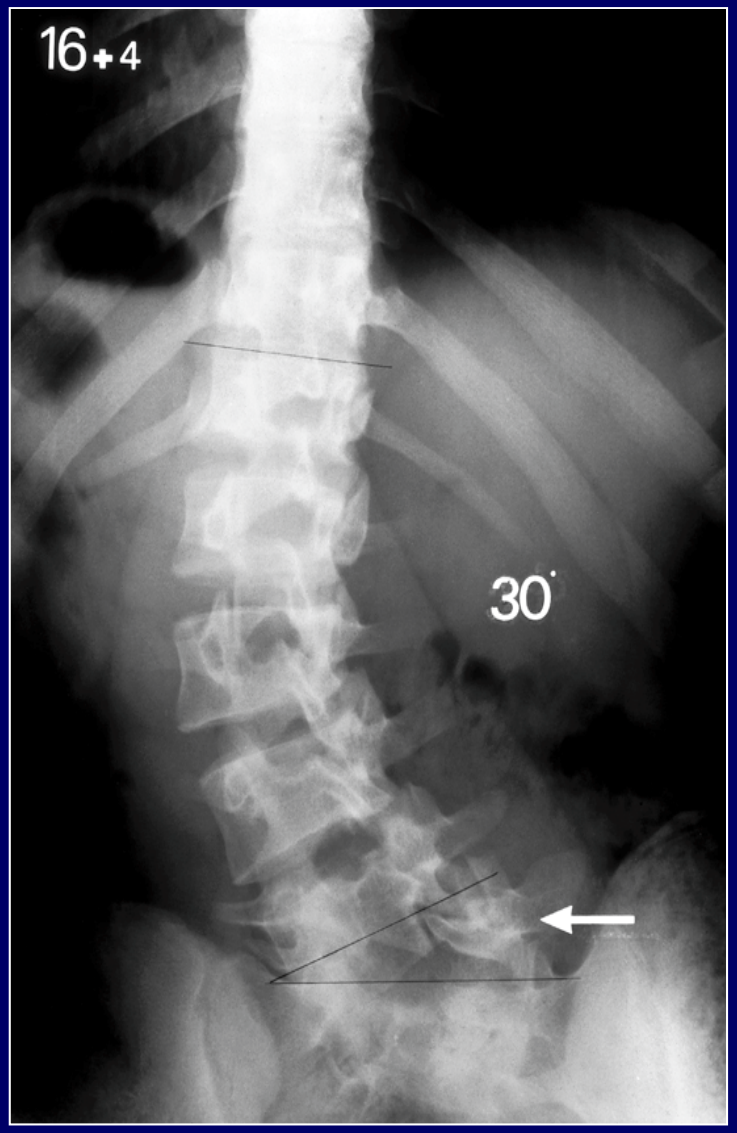

2+8
65º


3+0



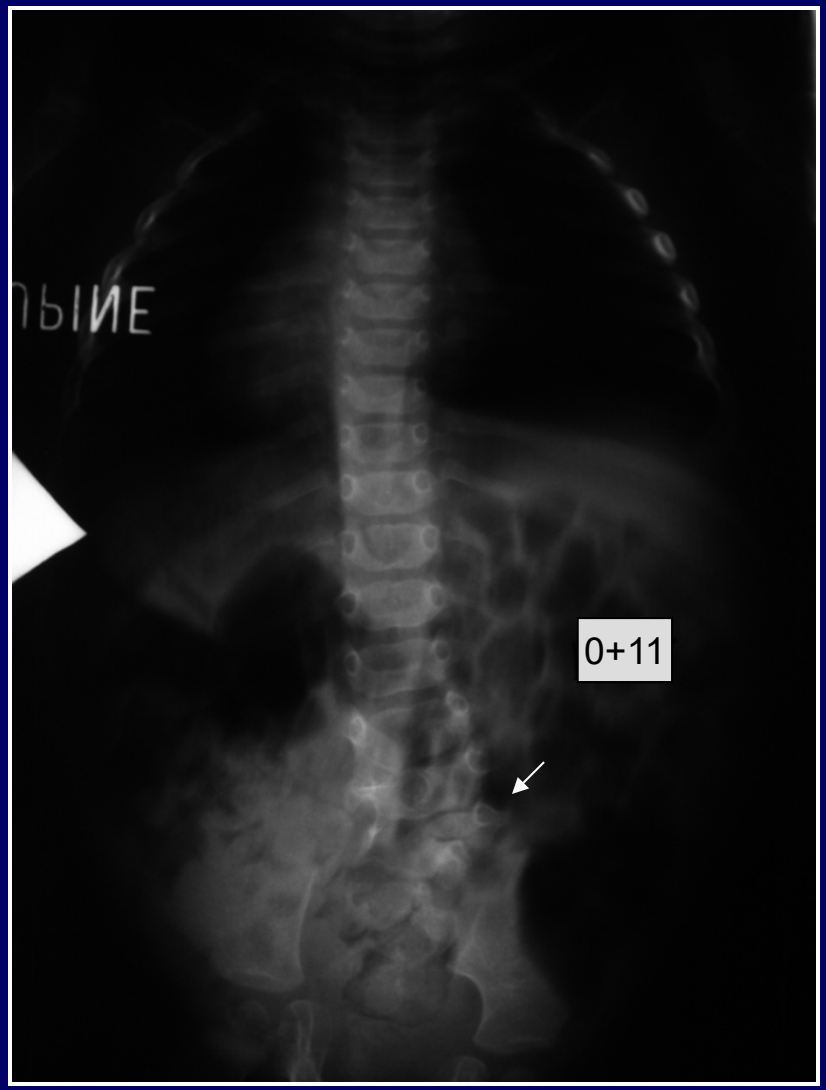

ST
L

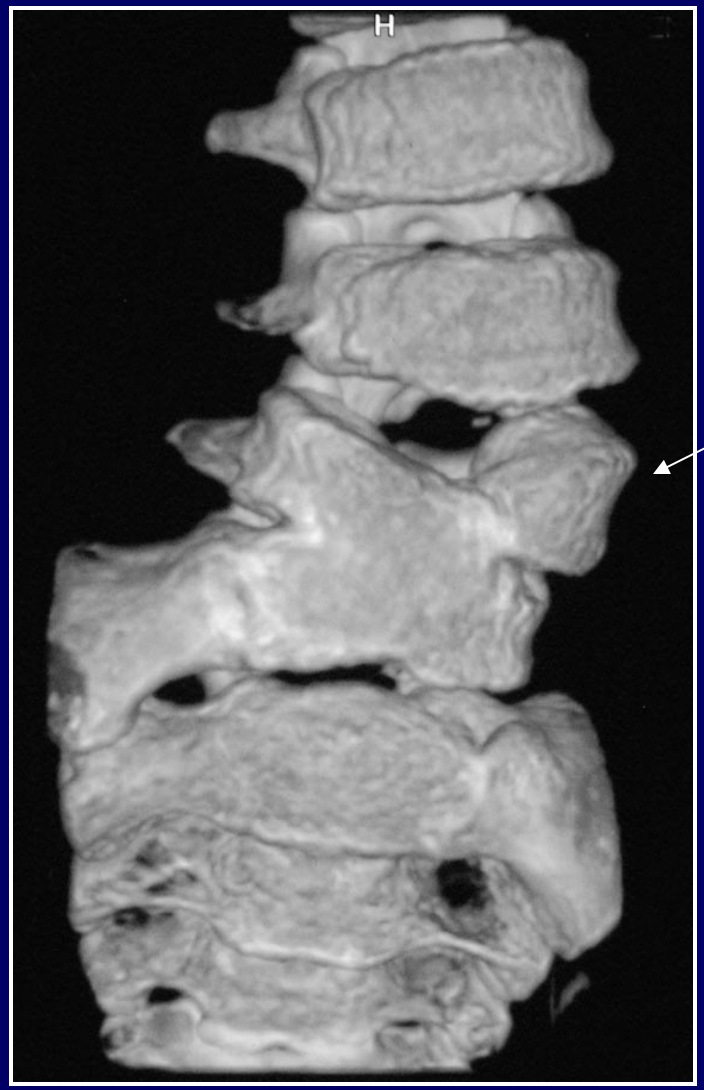
Lumbosacral hemivertebra



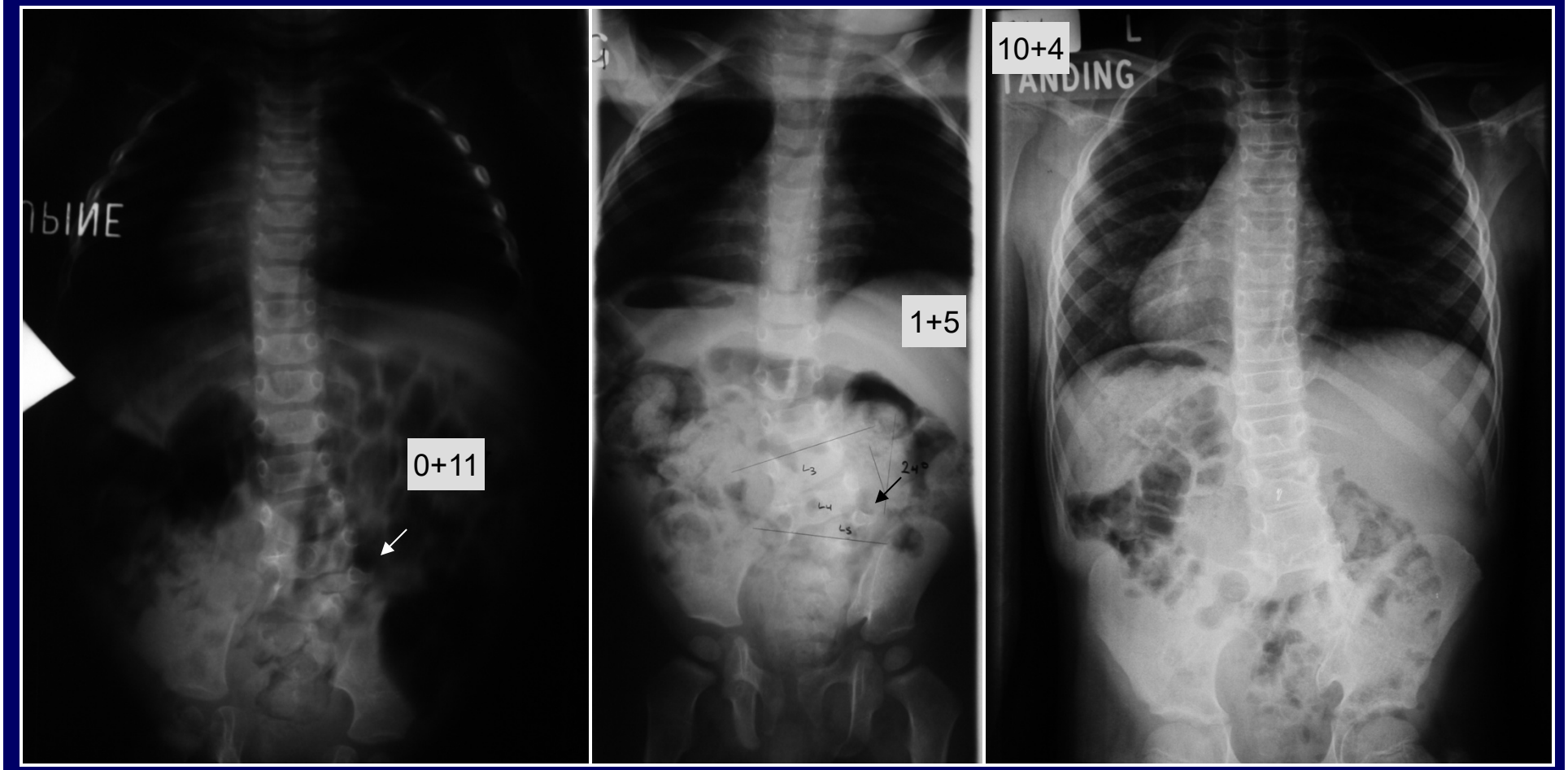
McMaster

Lumbosacral hemivertebra

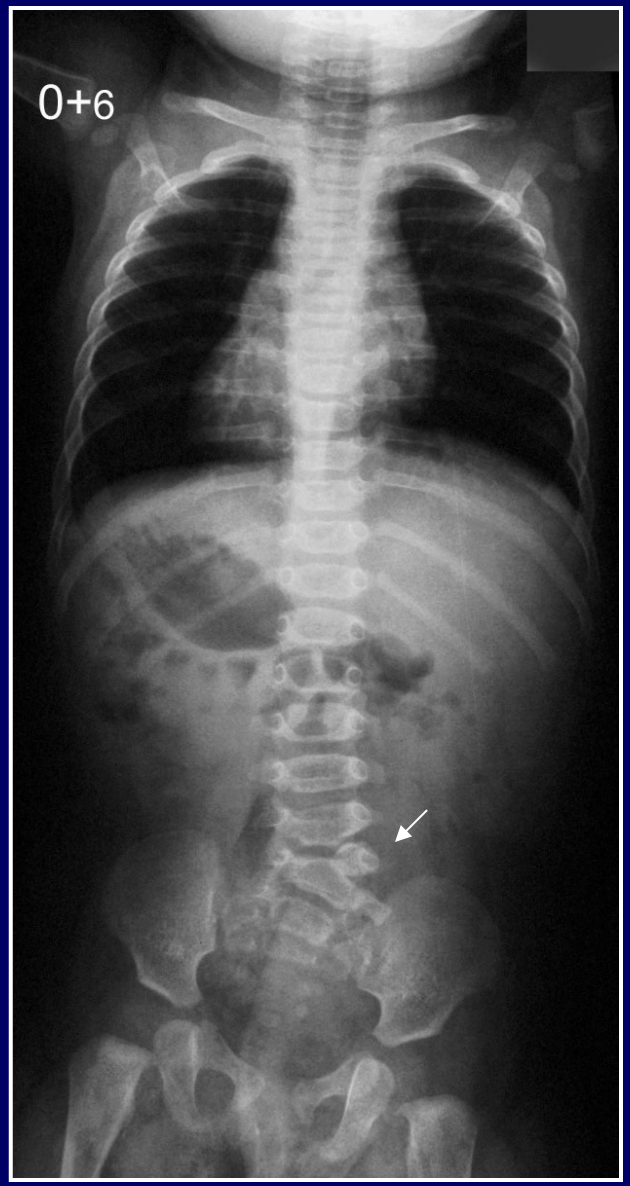


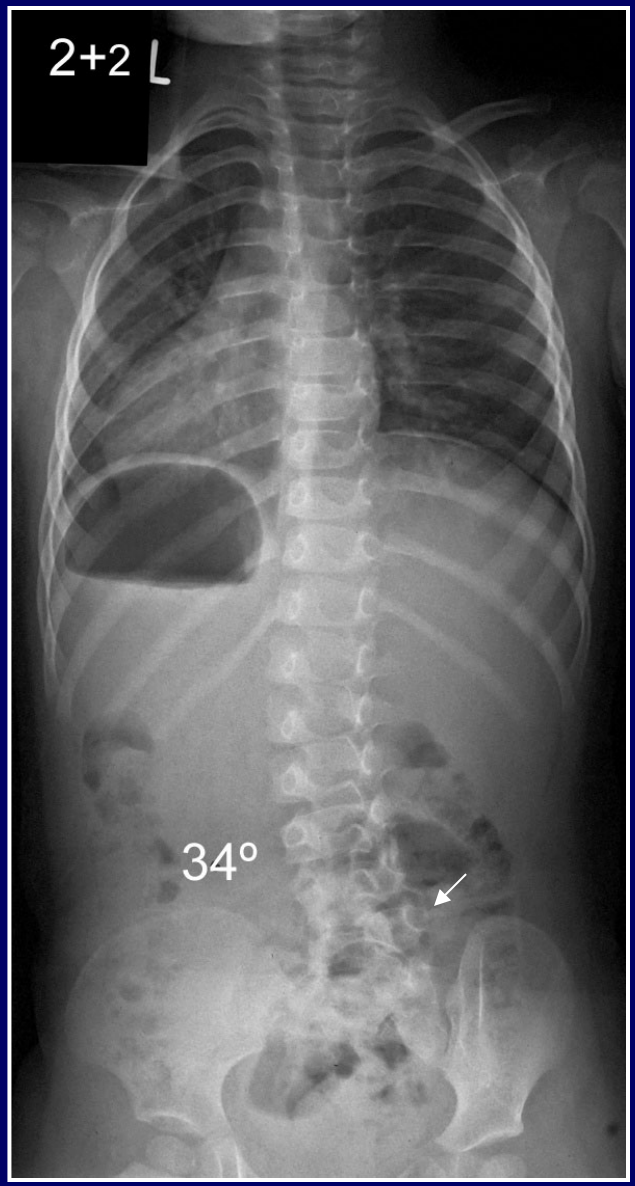


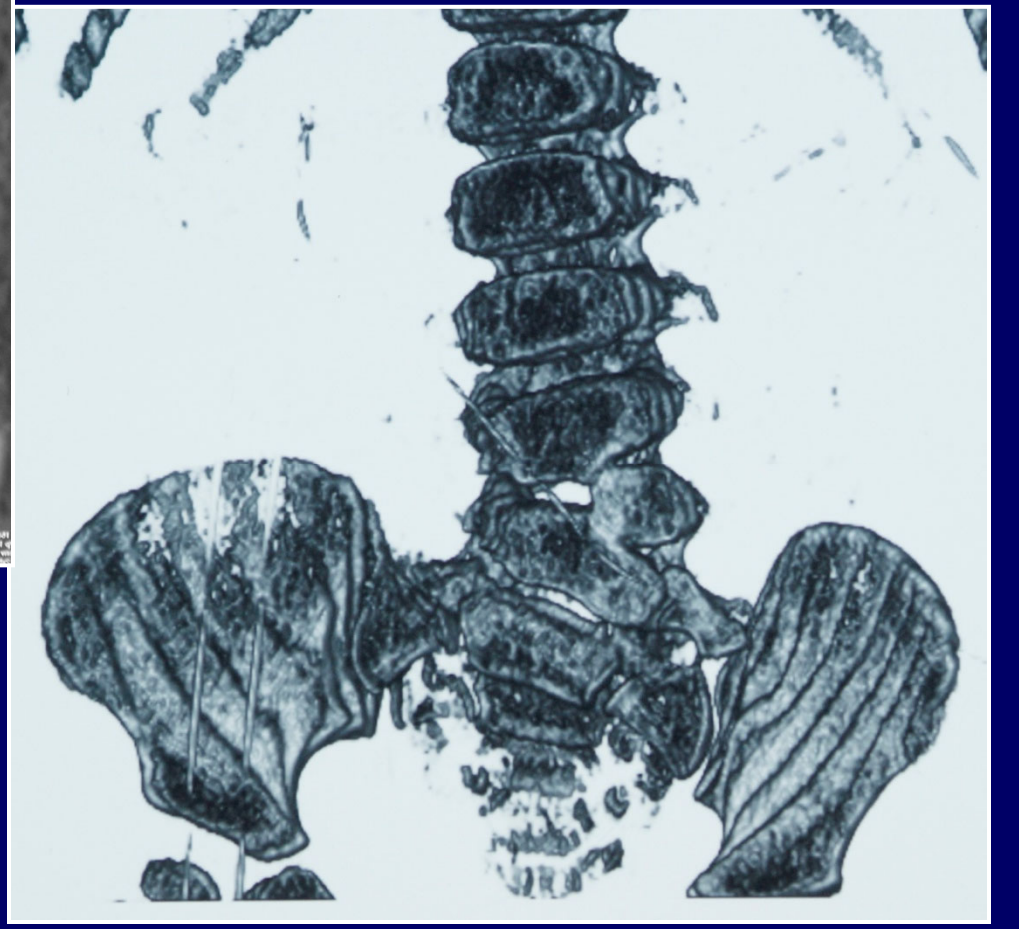
Posterior in situ fusion

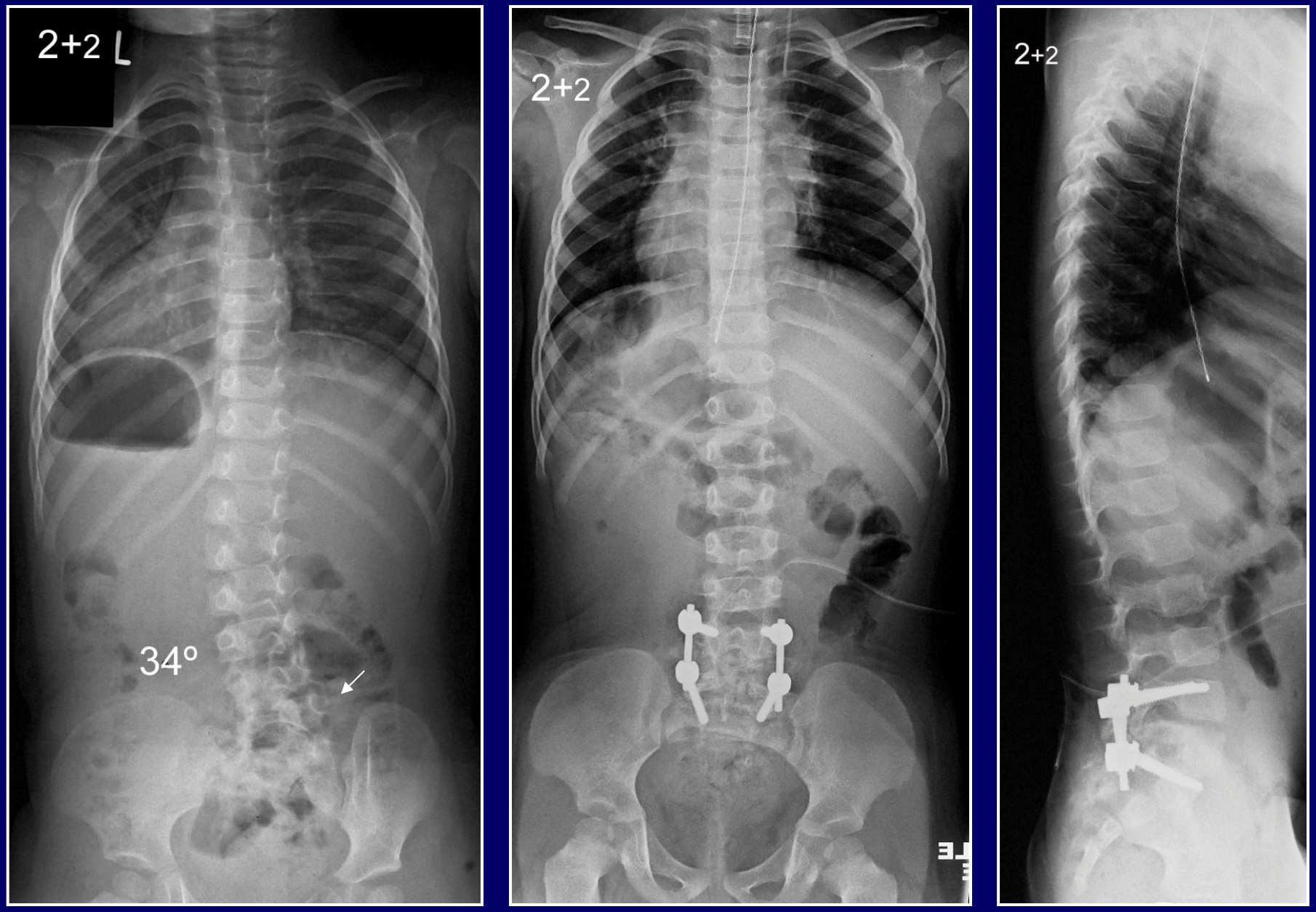


LS hemivertebra resection



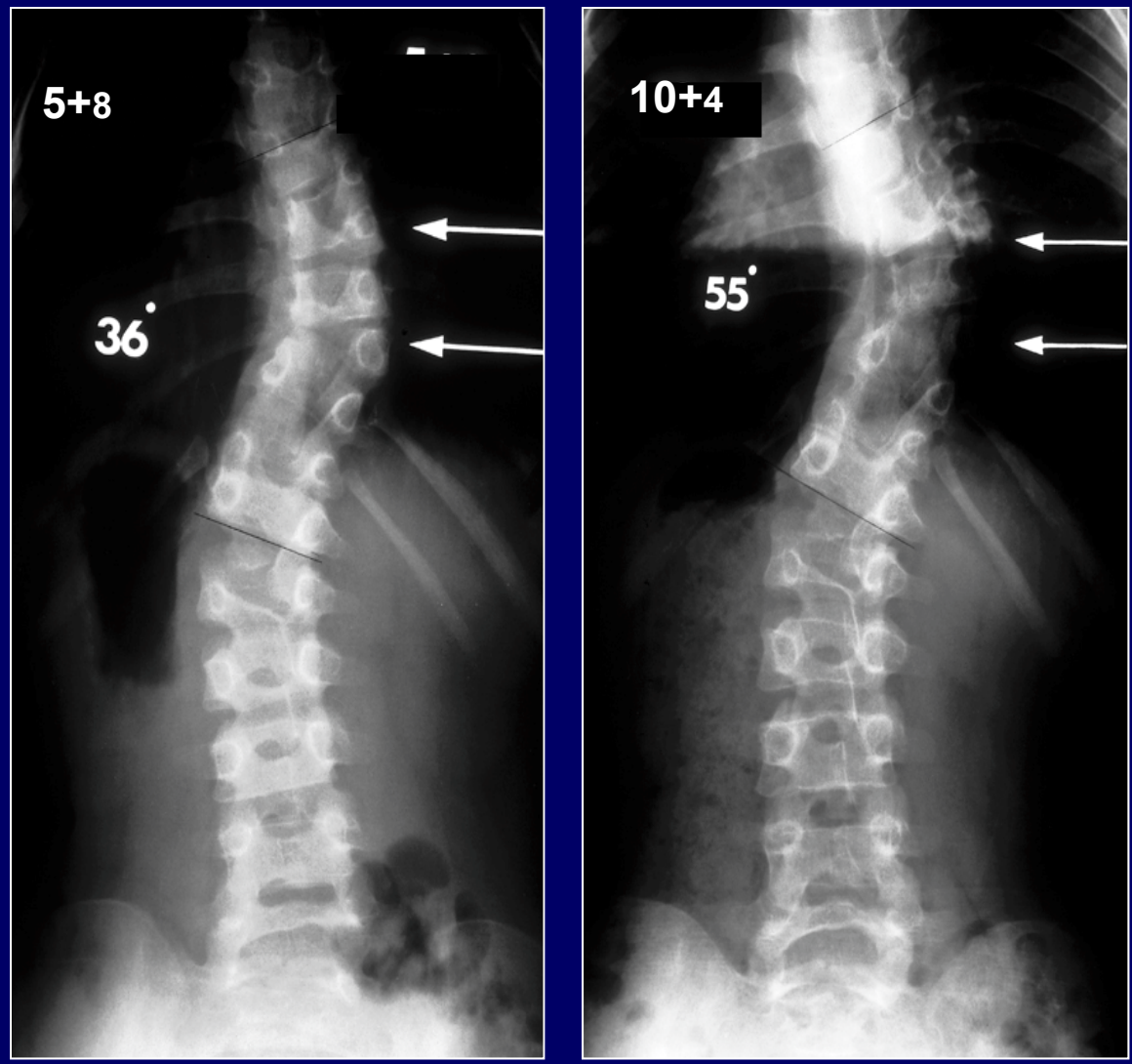




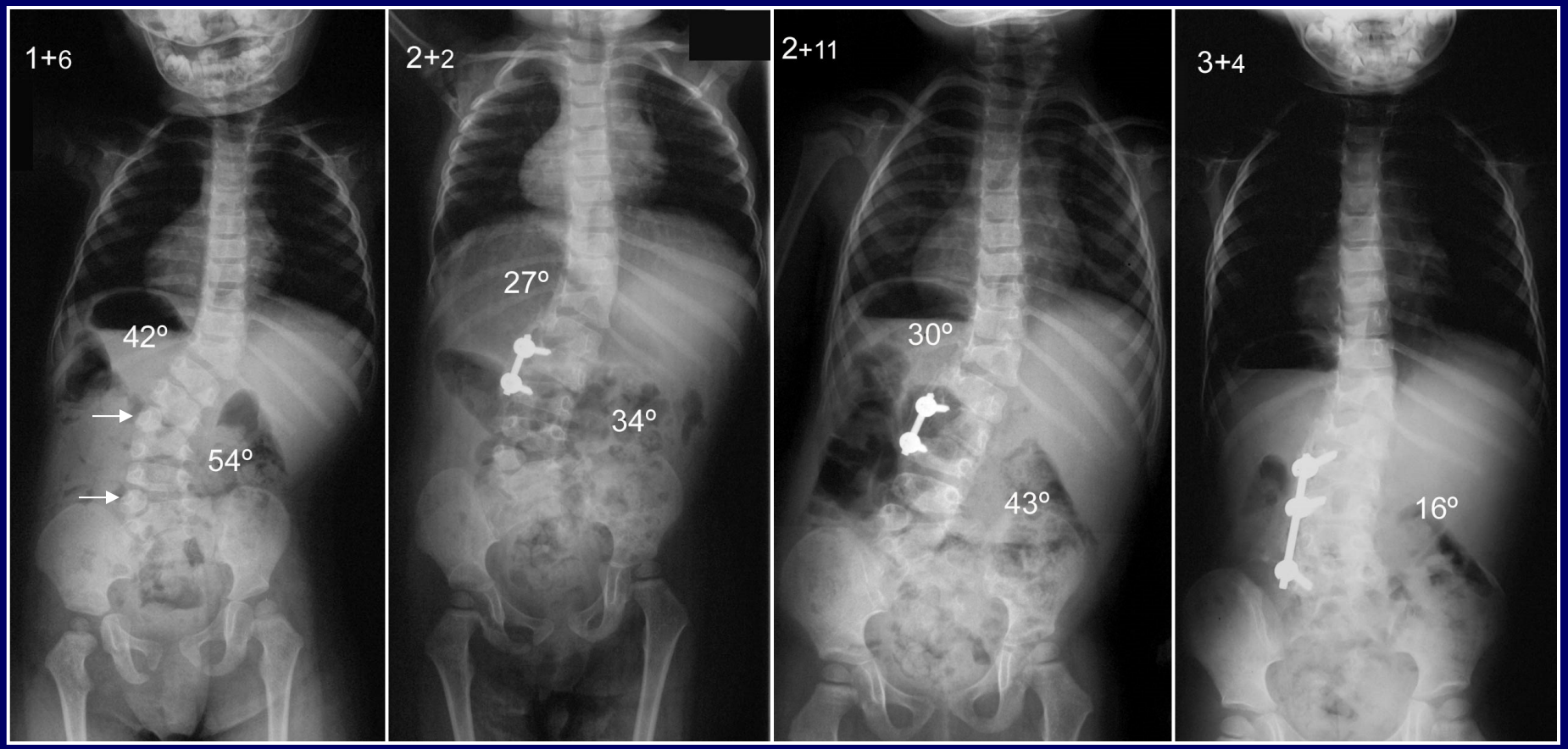

2+2 L
34°
2+2
2+2

Two ipsilateral hemivertebrae

- scoliosis progresses at a mean rate of 3-4^{o}/year
- exceeds 50^{o} by 10 yrs
- reaches 70^{o} by skeletal maturity
- early prophylactic surgical treatment is required

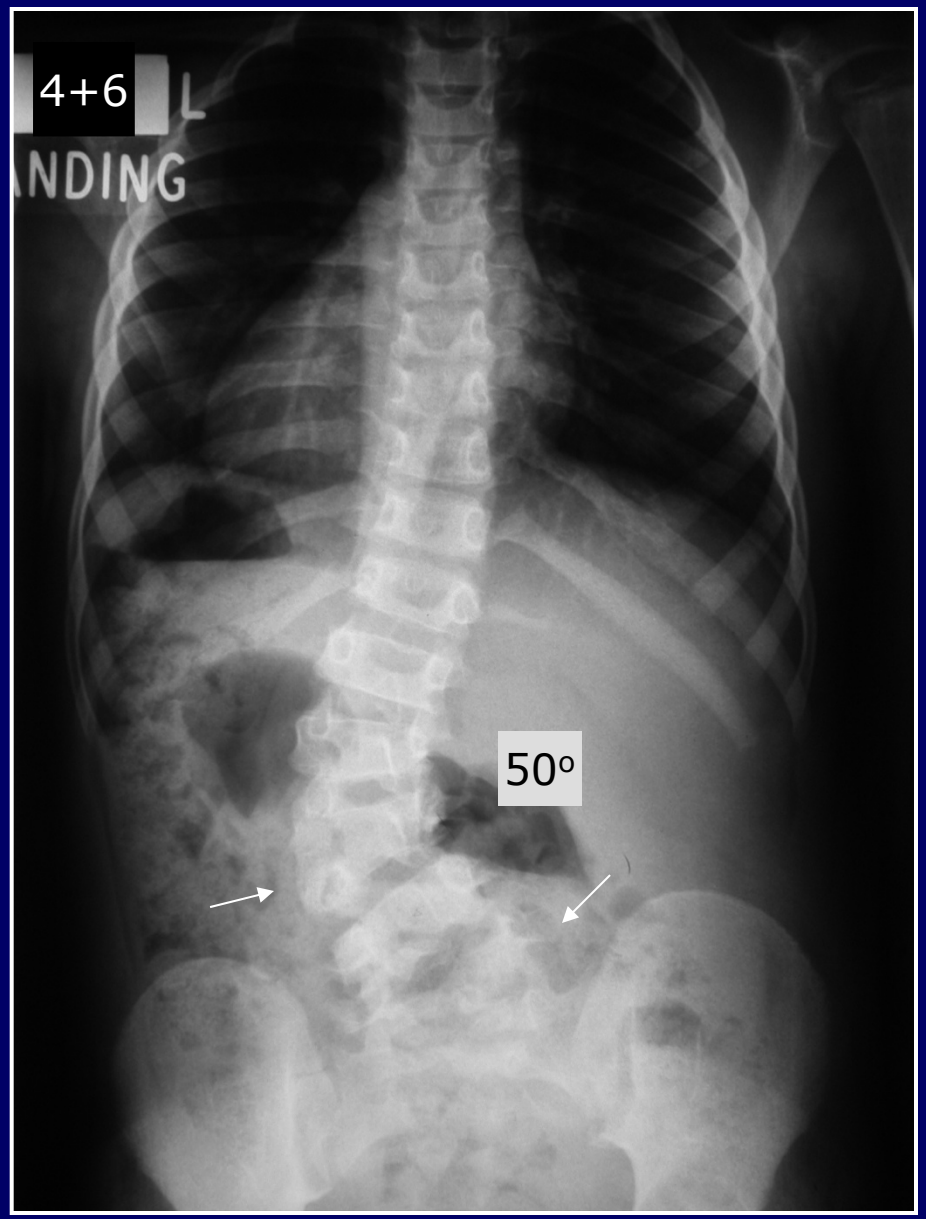


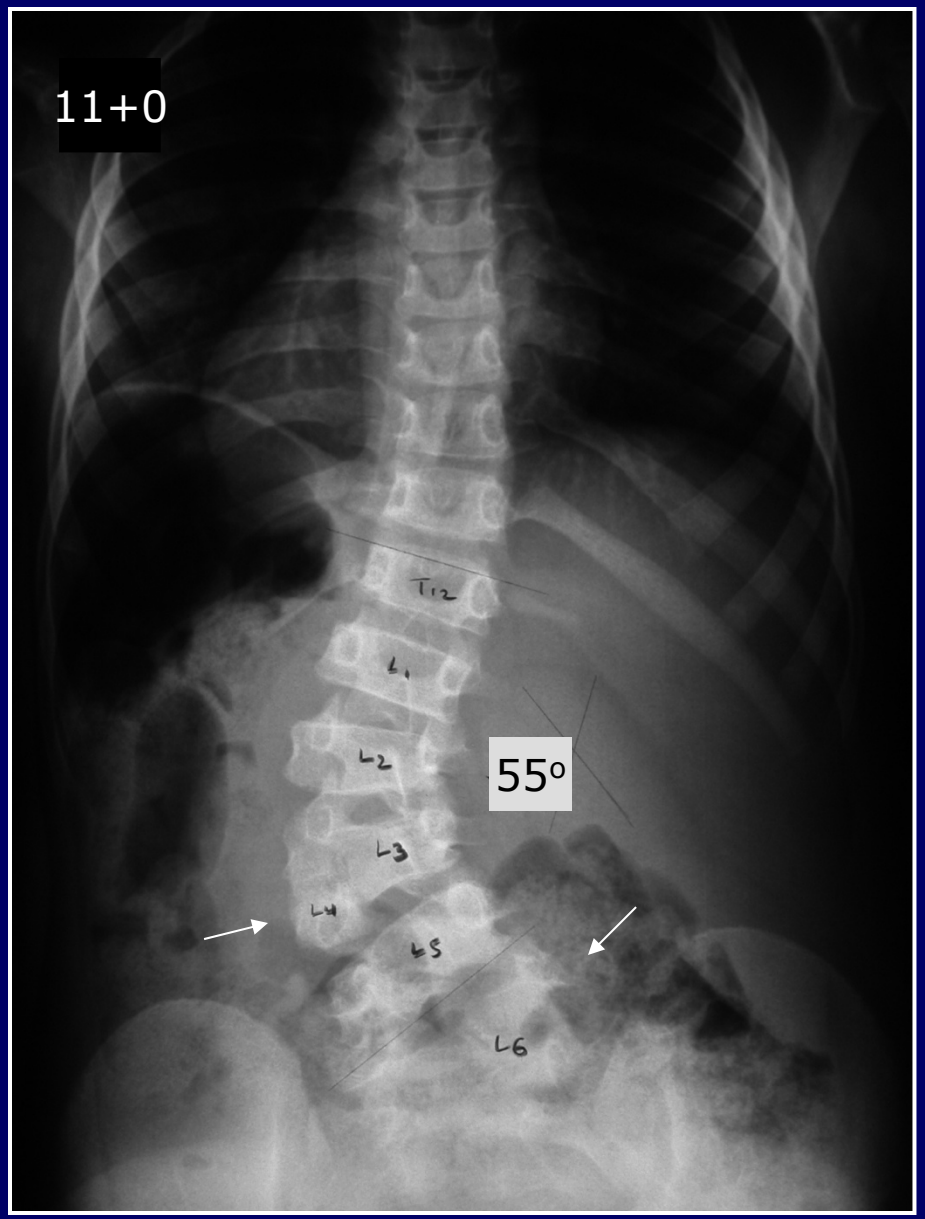
Two ipsilateral hemivertebrae



Two opposing hemivertebrae (hemimetameric shift)

- prognosis depends on site and relationship of the hemivertebrae
- if located in different regions of the spine they produce a significant deformity and usually require treatment


4+6
L
NDING
50°

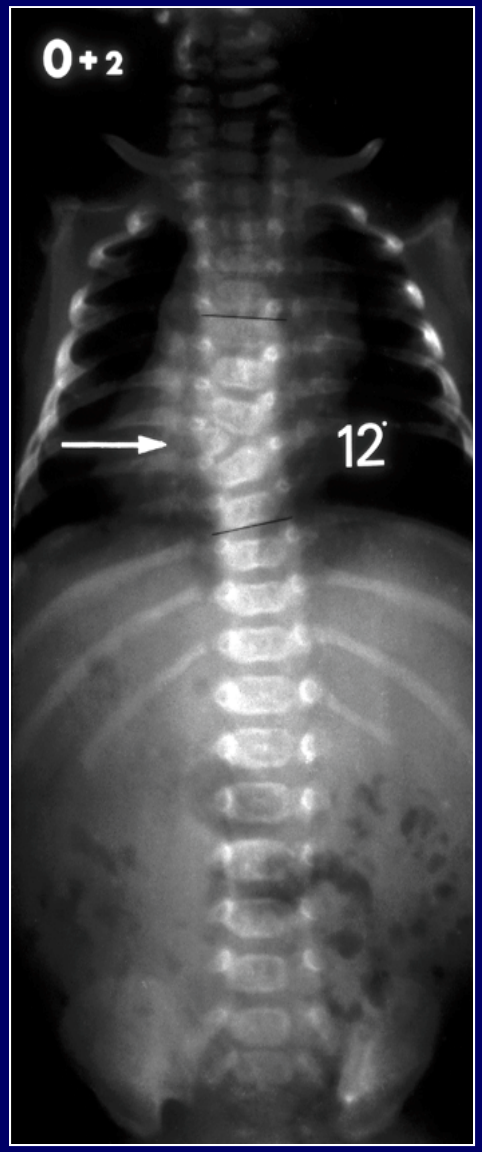

11+0
T12
L1
L2
L3
L4
L5
L6
55°

Semi-segmented hemivertebra (22%)

- slowly progressive scoliosis
- usually does not exceed 40^{o} at skeletal maturity
- treatment is often not required, except LS hemivertebra

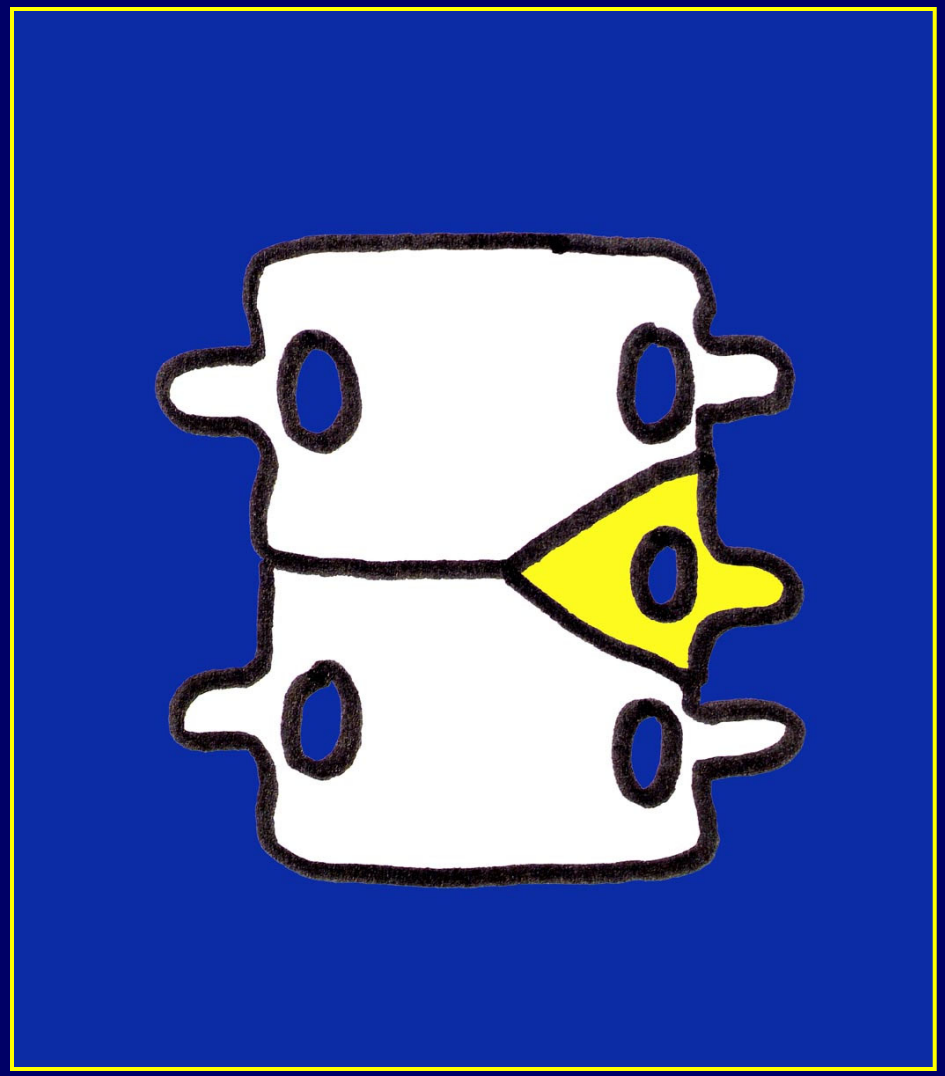
Incarcerated hemivertebra (13%)

- hemivertebra sets into a niche scalloped out of the adjacent vertebrae
- produces minimal deformity due to poor growth potential
- treatment is not required

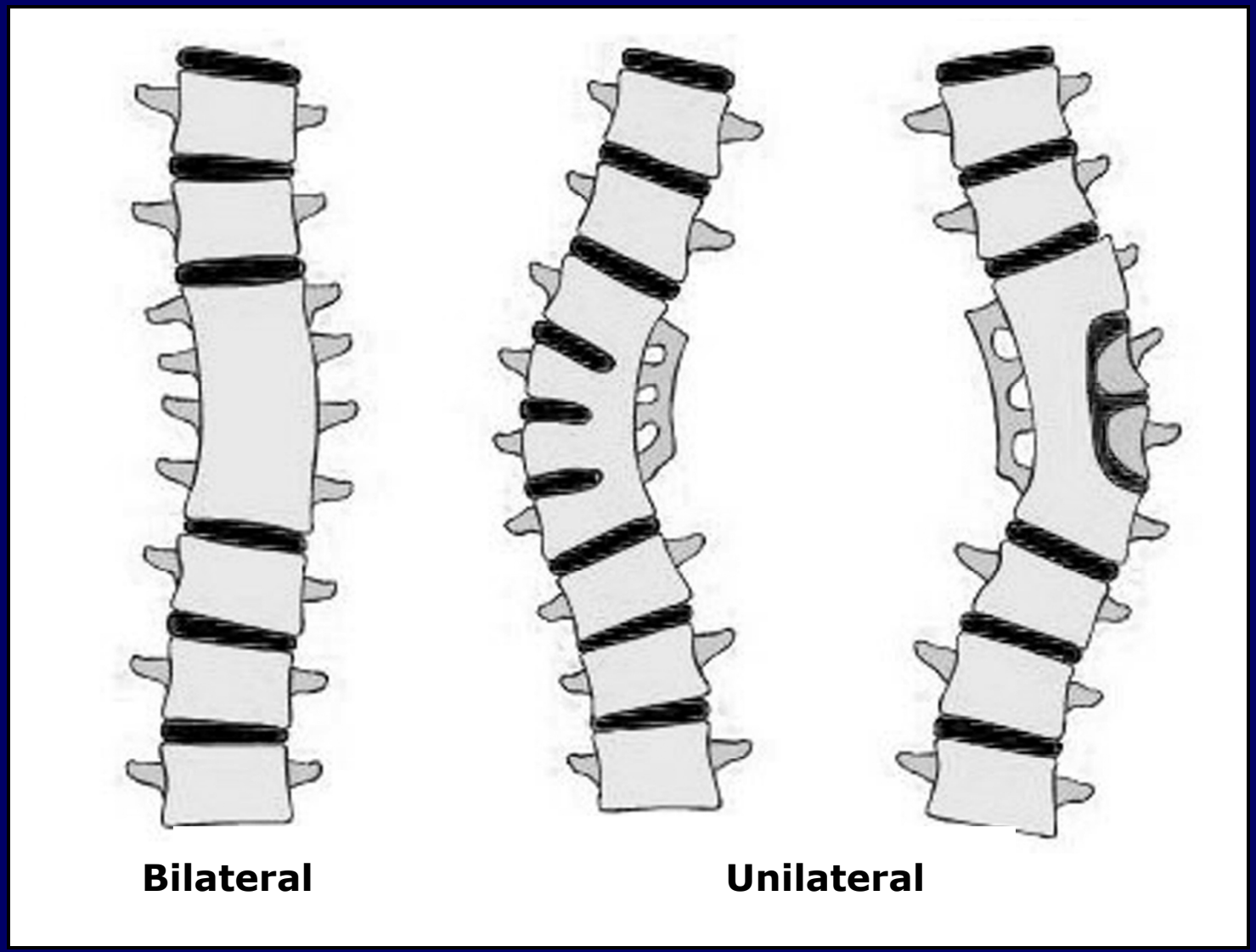


Unsegmented hemivertebra

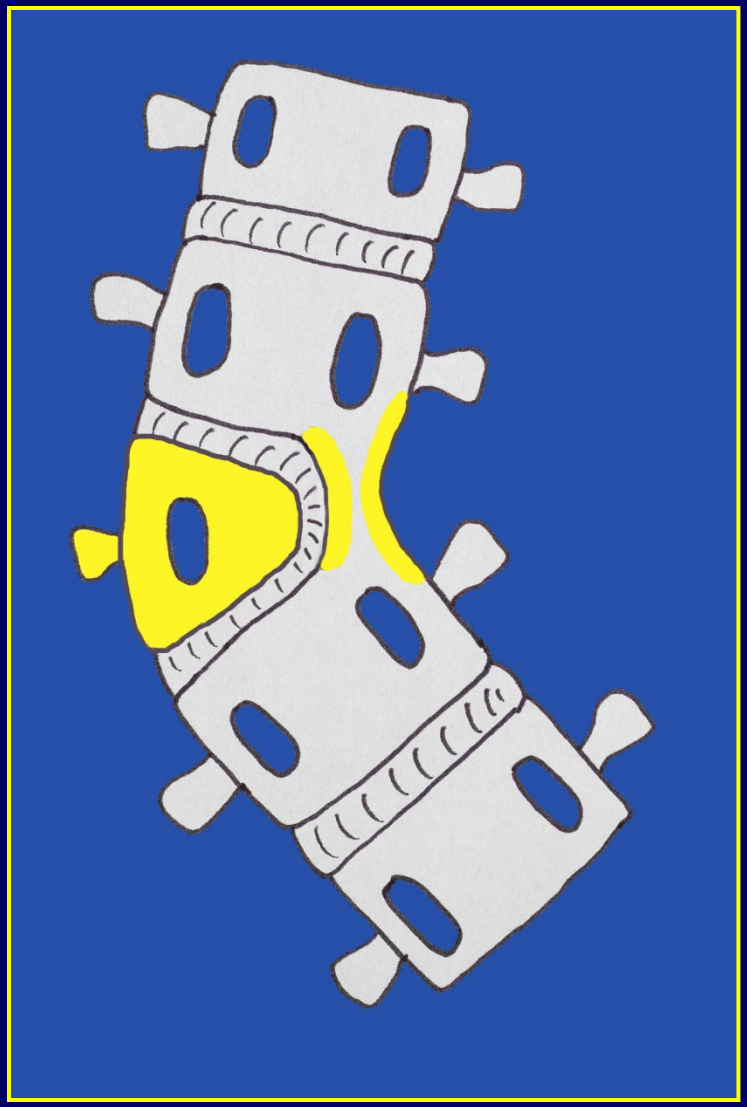
- usually occurs in the thoracic spine
- has no growth potential
- does not cause progressive scoliosis
- no treatment is required



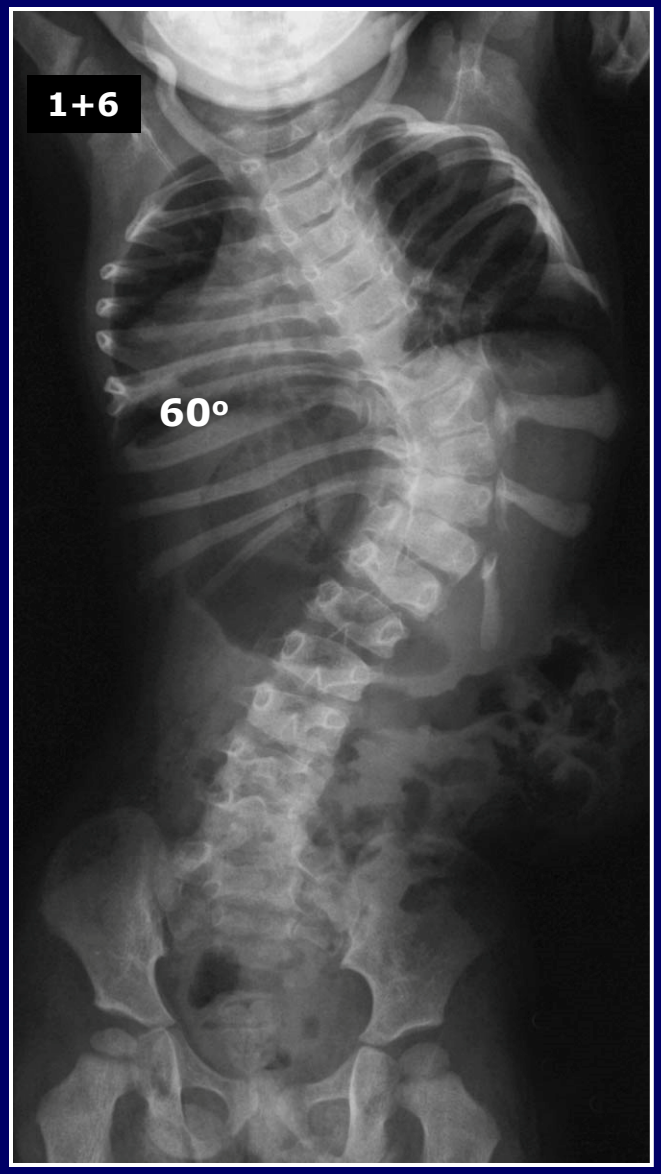
Failures of Vertebral Segmentation



Unilateral Failure of Segmentation

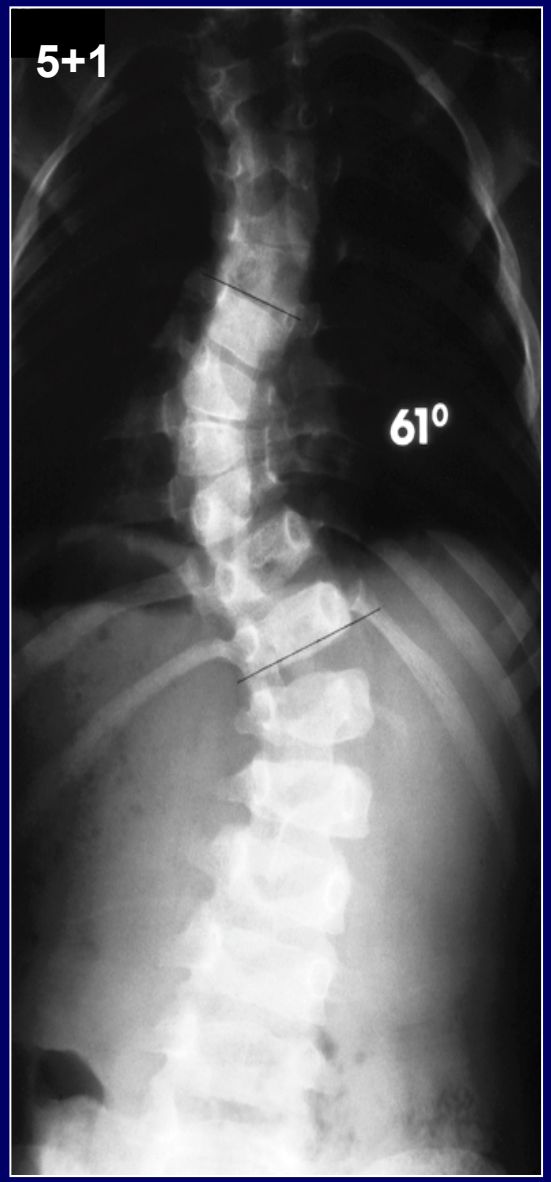


Extent of Bar/Convex growth



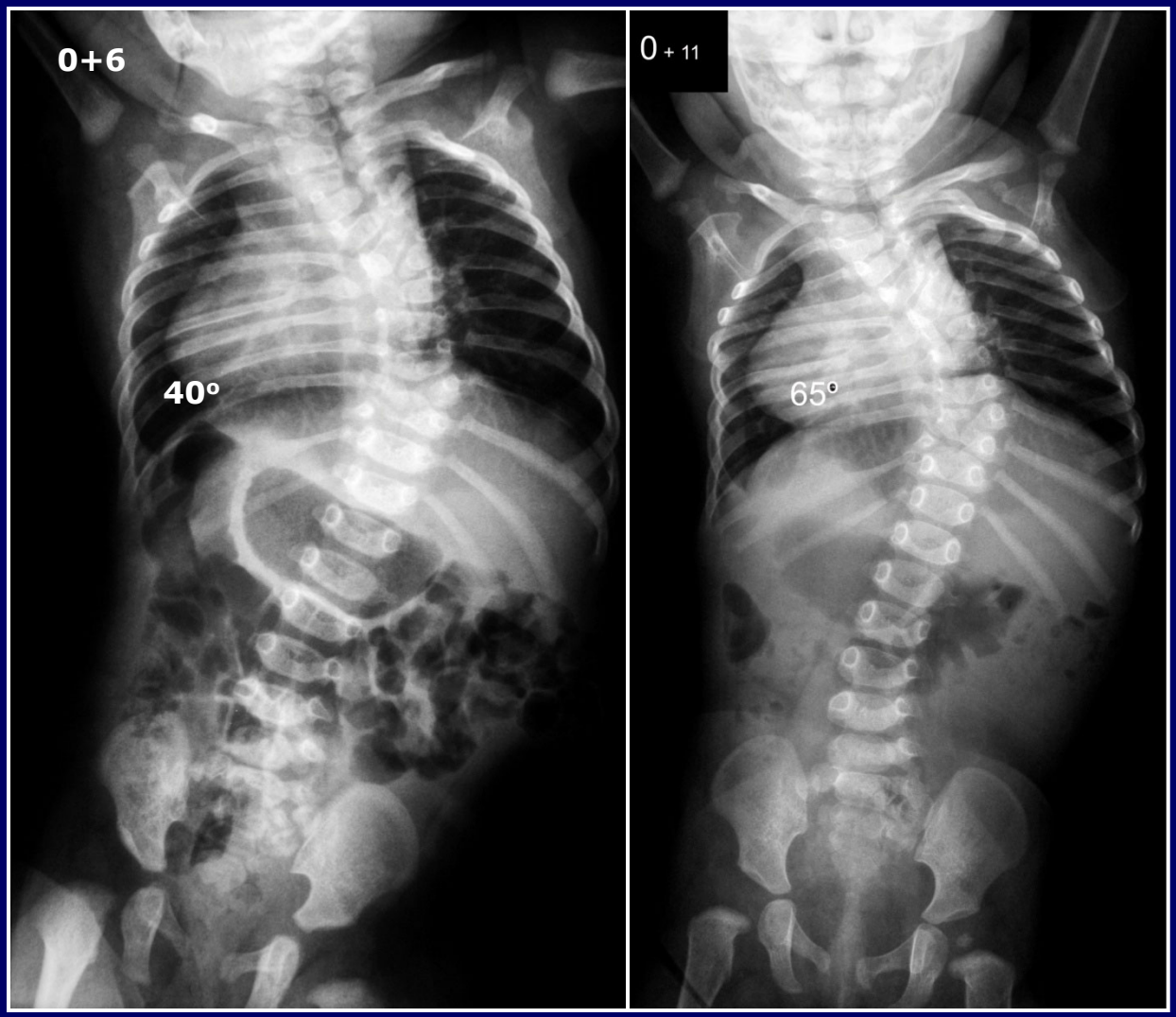
Unilateral Unsegmented Bar

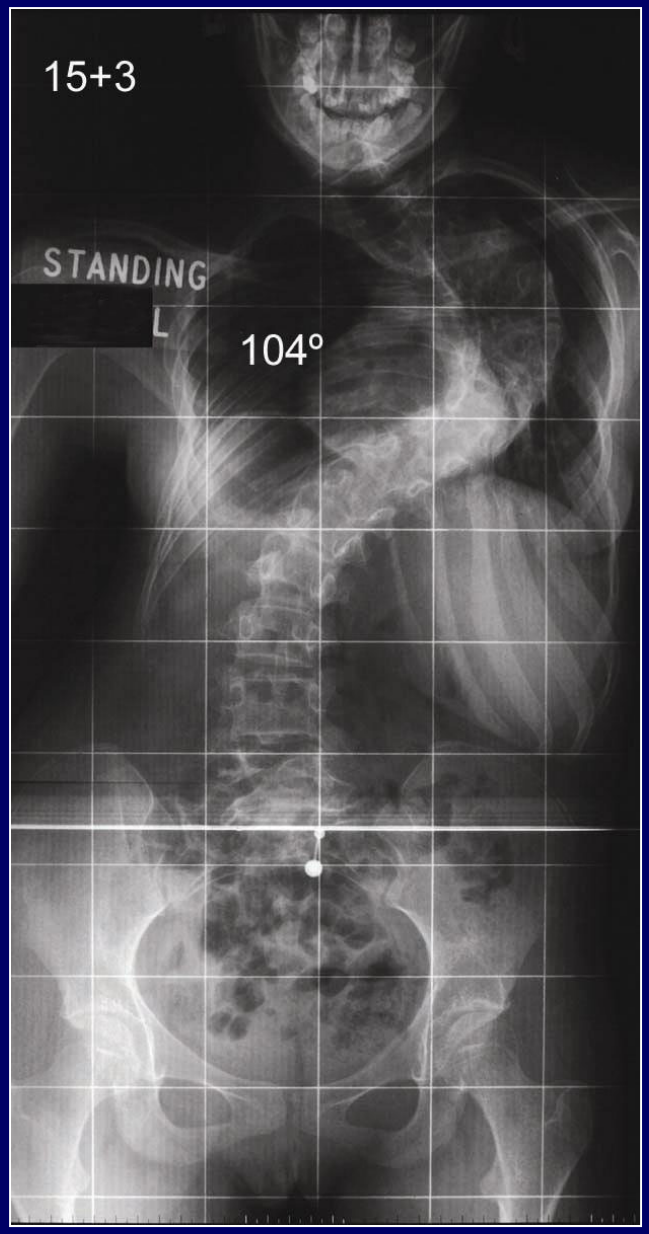
- mean rate of progression 5°/year
- majority exceed 50° by 10 yrs
- all require early prophylactic treatment



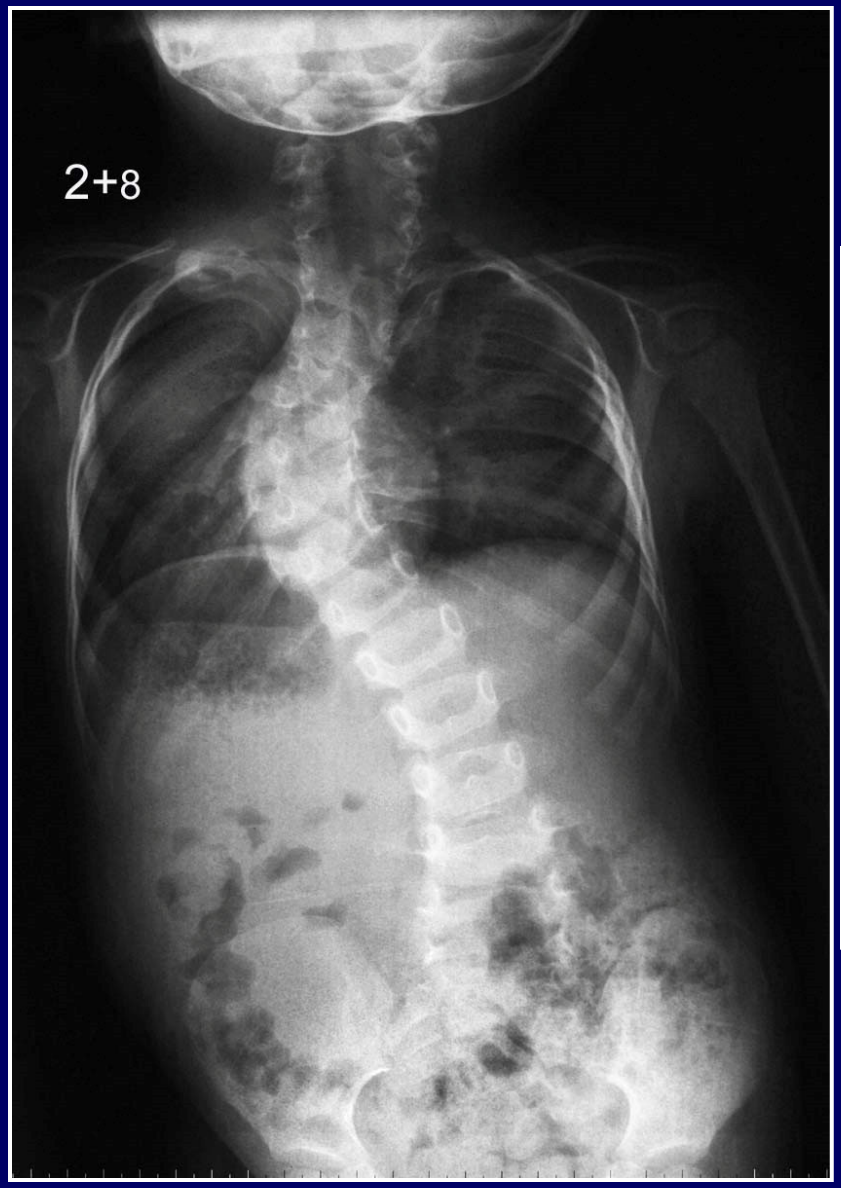
Unsegmented Bar and Hemivertebrae

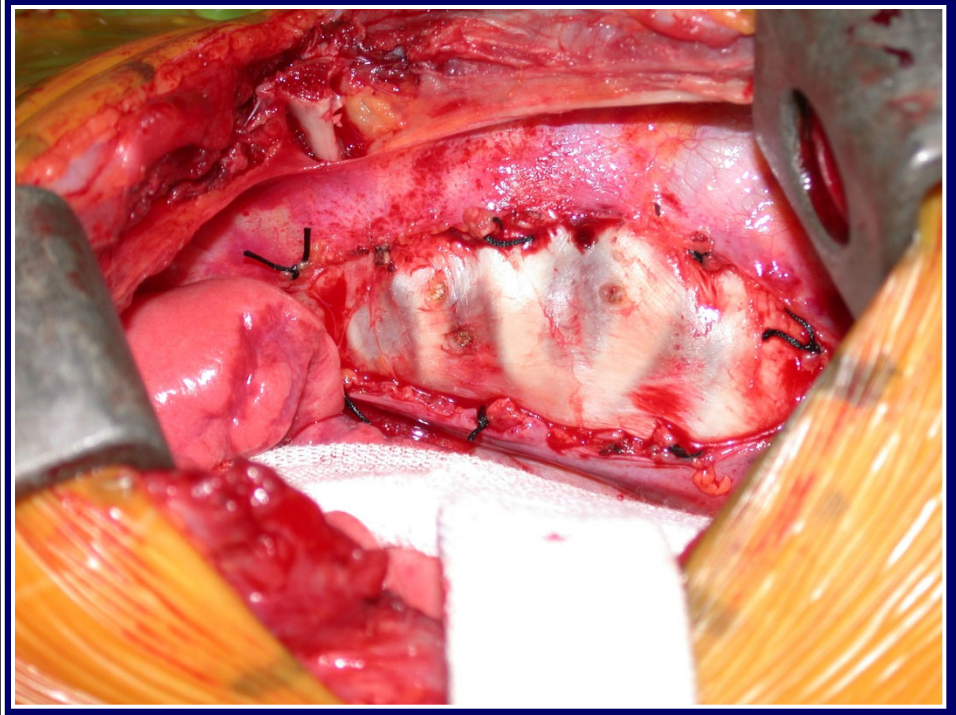
- mean rate of progression 6°/year
- all exceed 50° by age 2 yrs



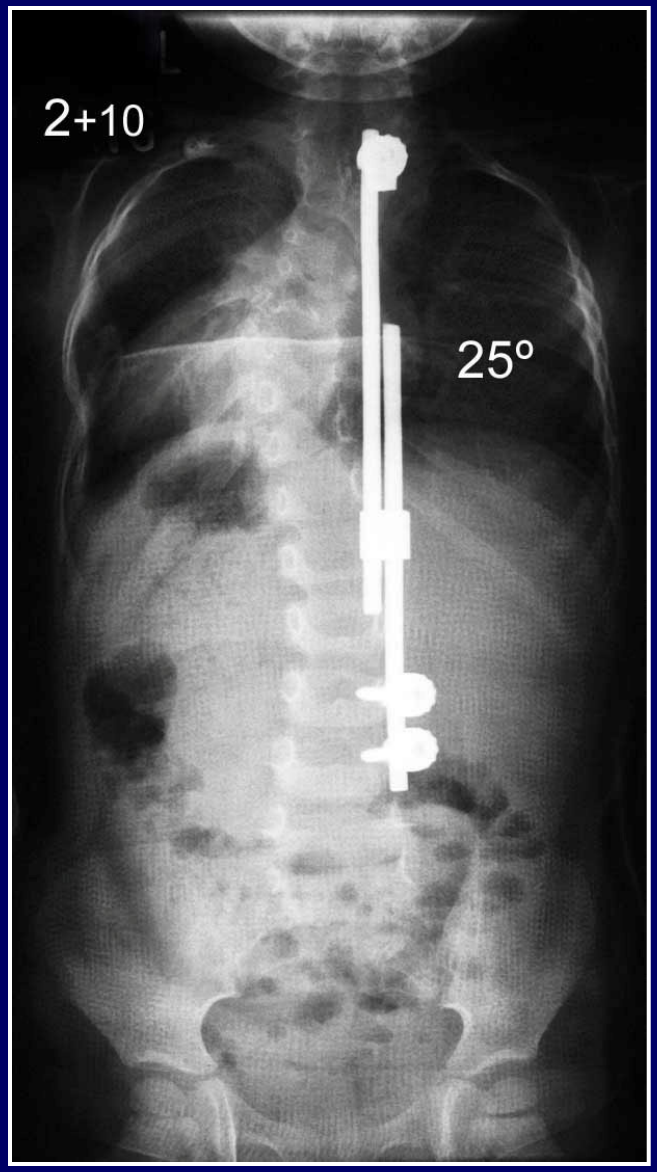

15+3
STANDING
L
104°

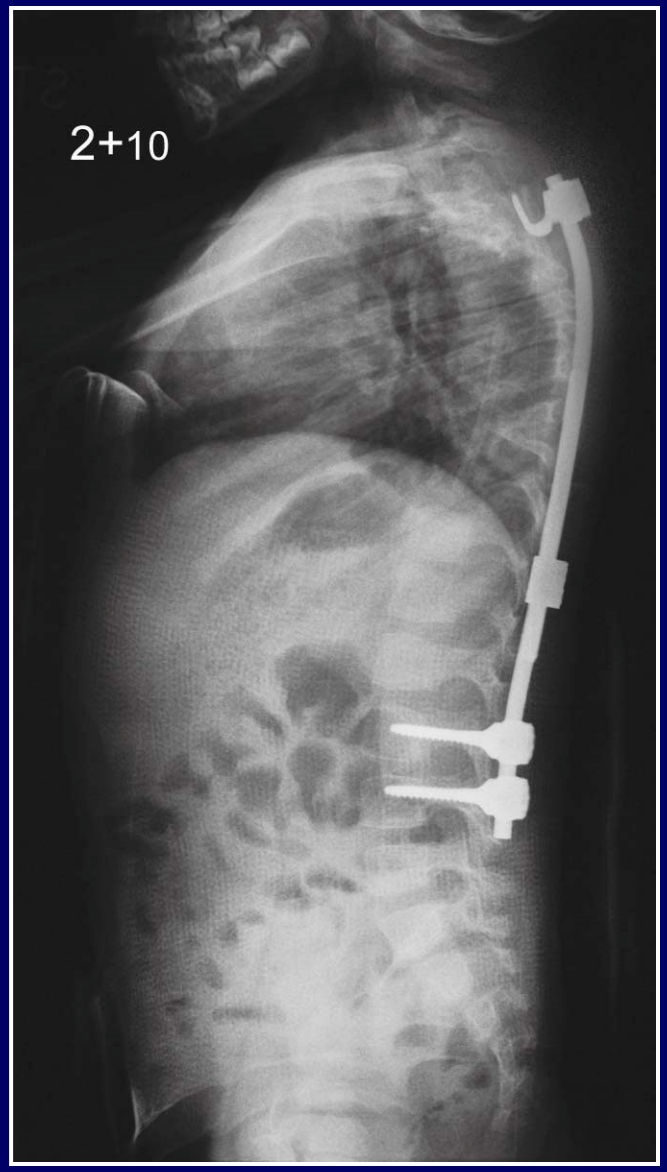
Convex fusion-GR



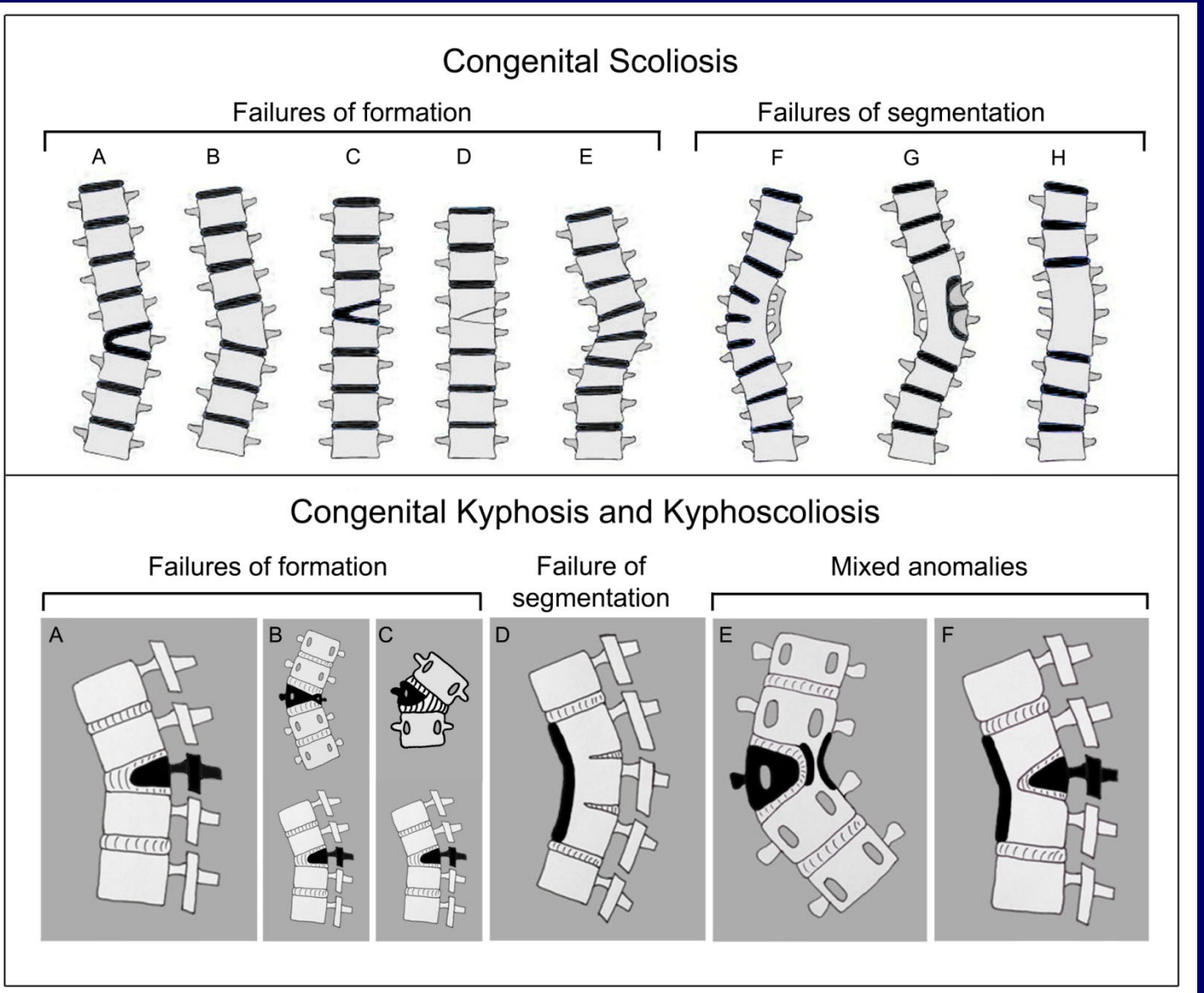


Convex fusion-GR





Classification



Alignment of the spinal canal

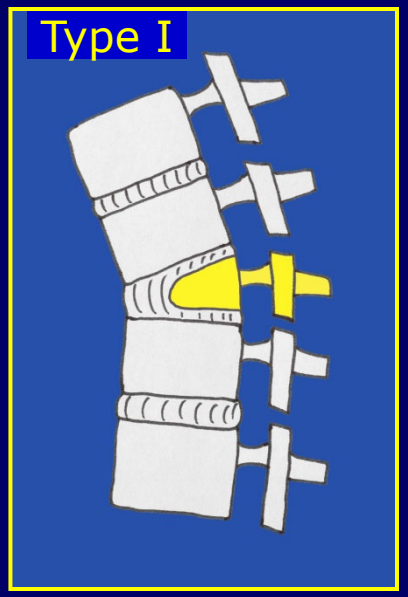
Type II-aligned

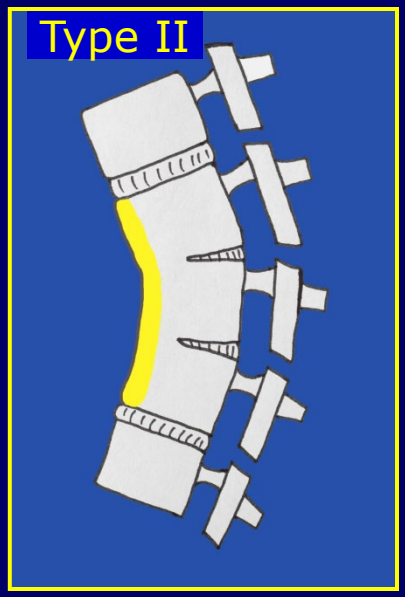
Type I-aligned

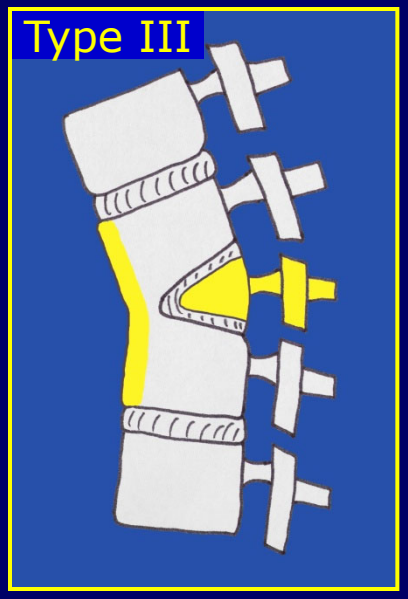
Type I-displaced

Prognosis

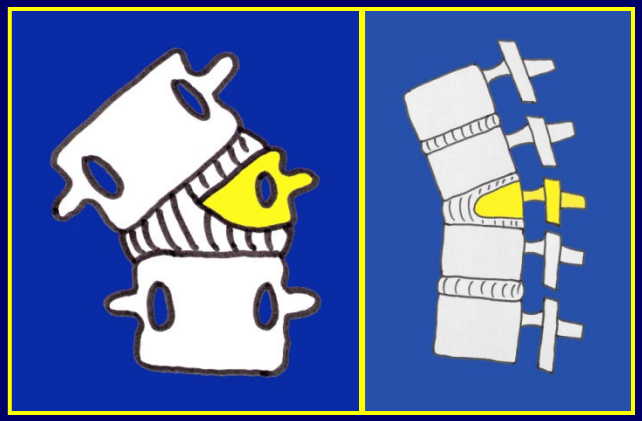
- pathologic anatomy and type of anomaly
  -Types I and III: worst prognosis
- site
  -usually TL
  -if TH: higher risk for neurological complications
- alignment of the spinal canal







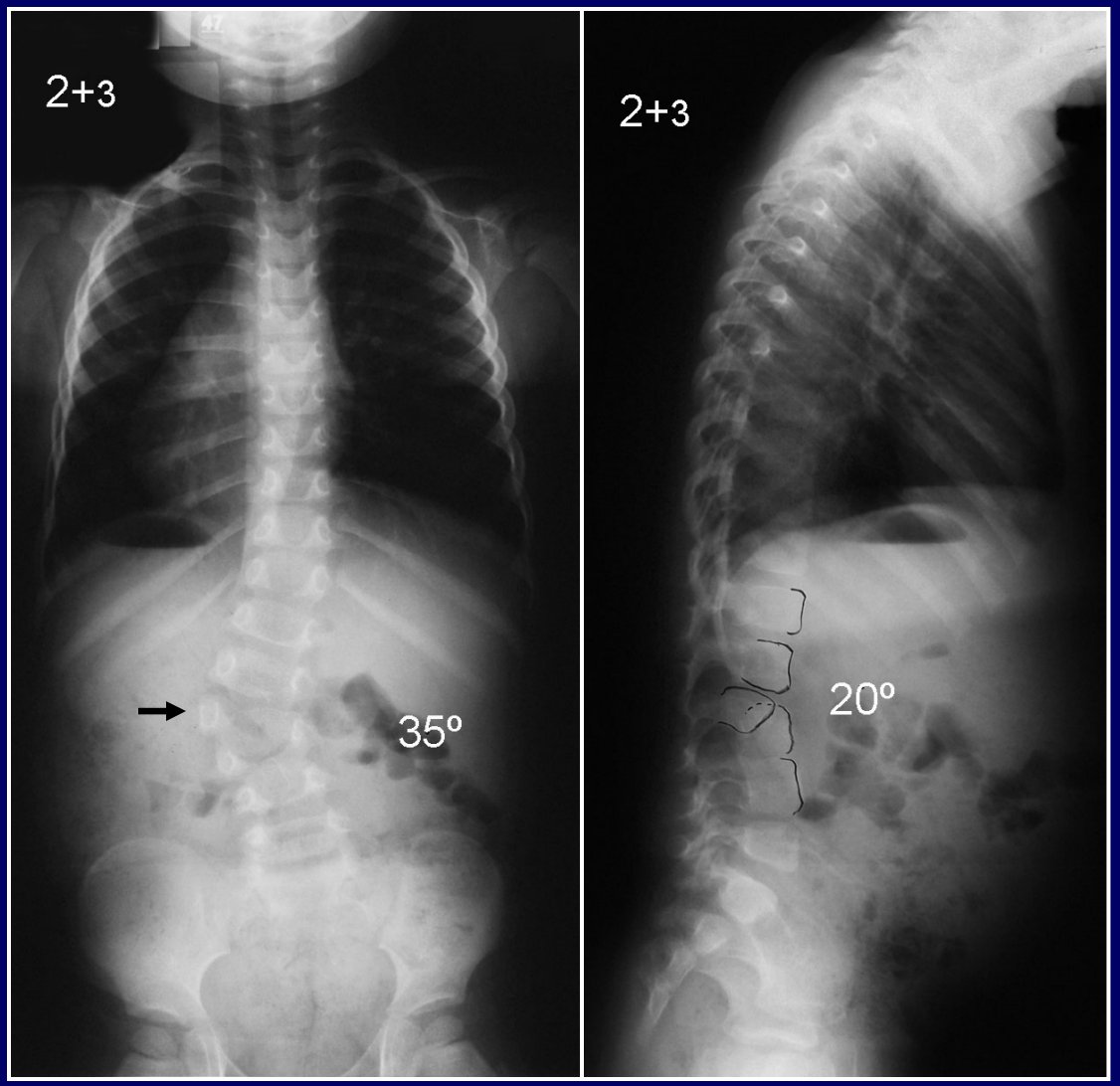
Posterolateral Quadrant Vertebra (35%)



- kyphosis progresses at a mean rate of:

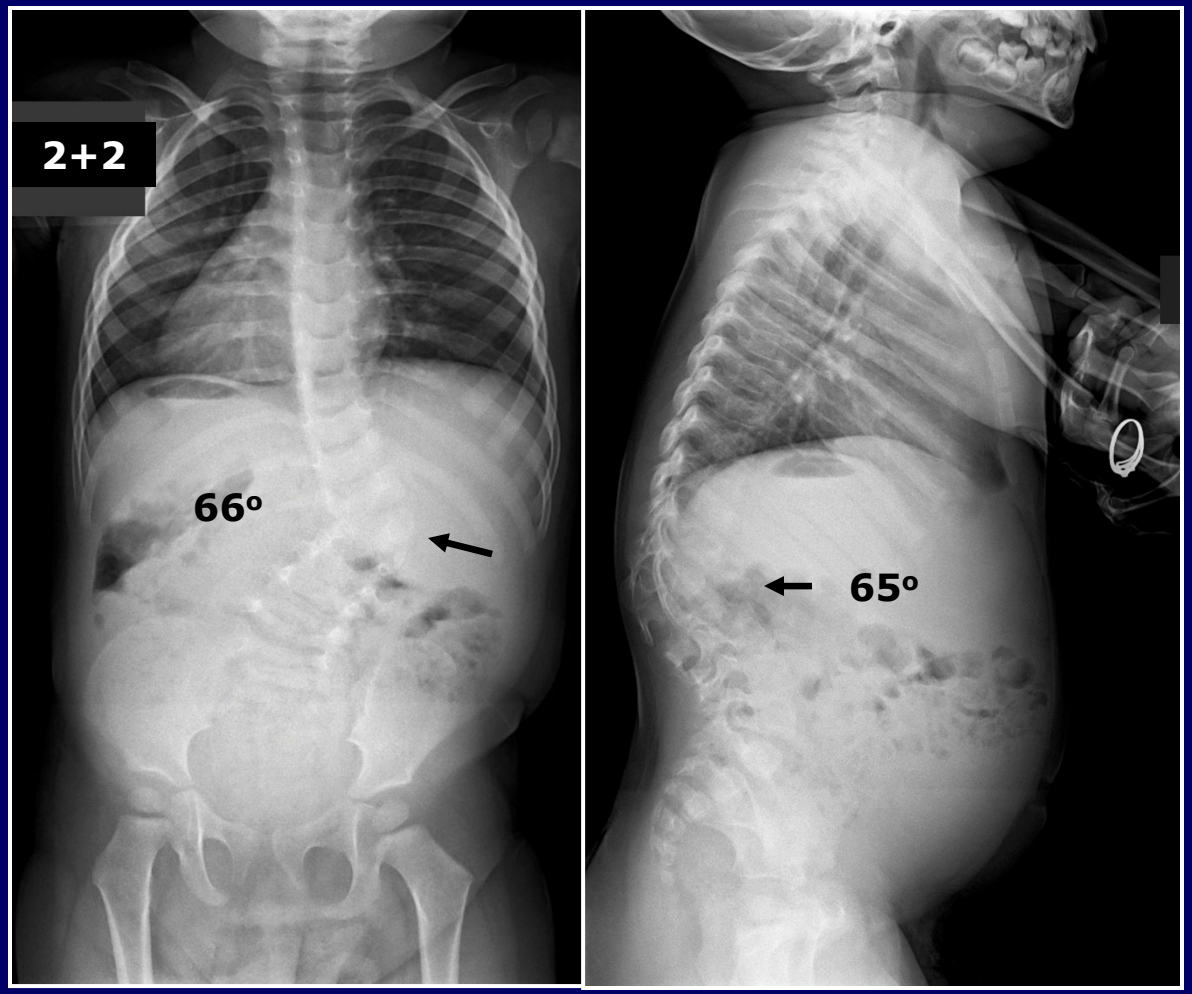
  2.5°/yr (before 10 yrs)

  5°/yr (after 10 yrs)

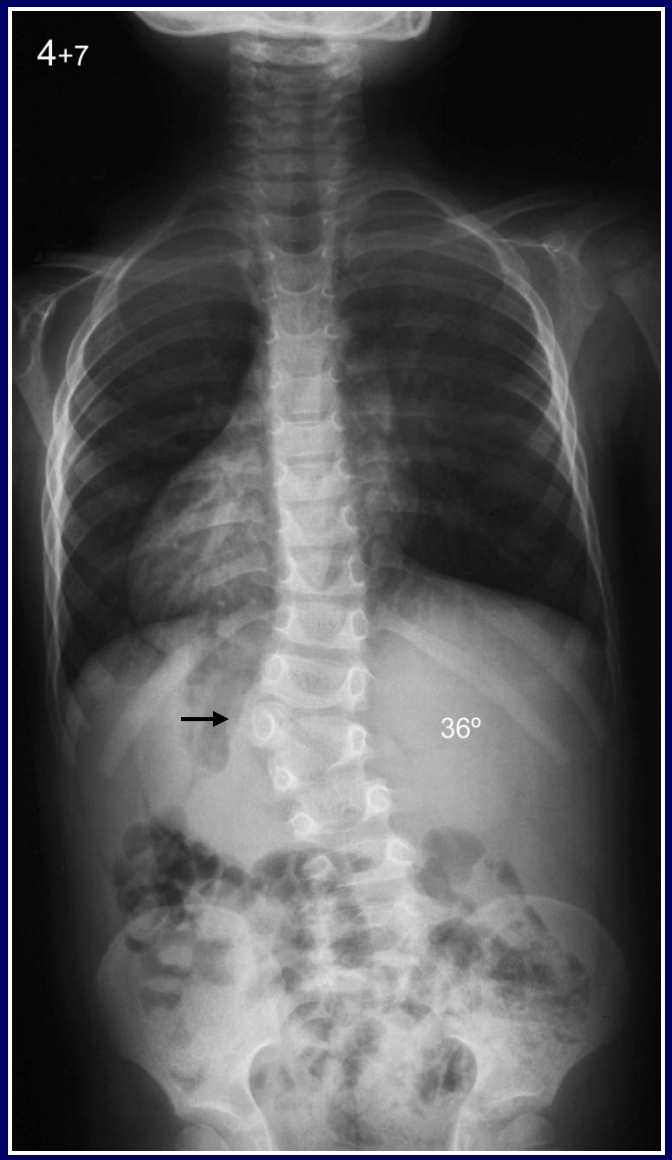


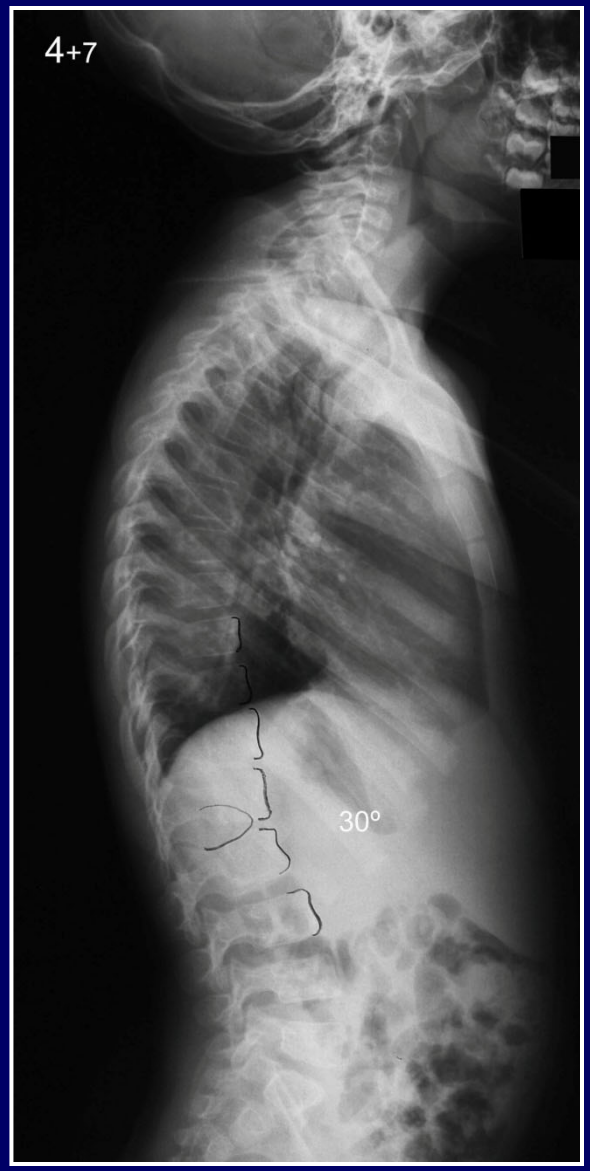
Posterolateral Quadrant Vertebra (35%)

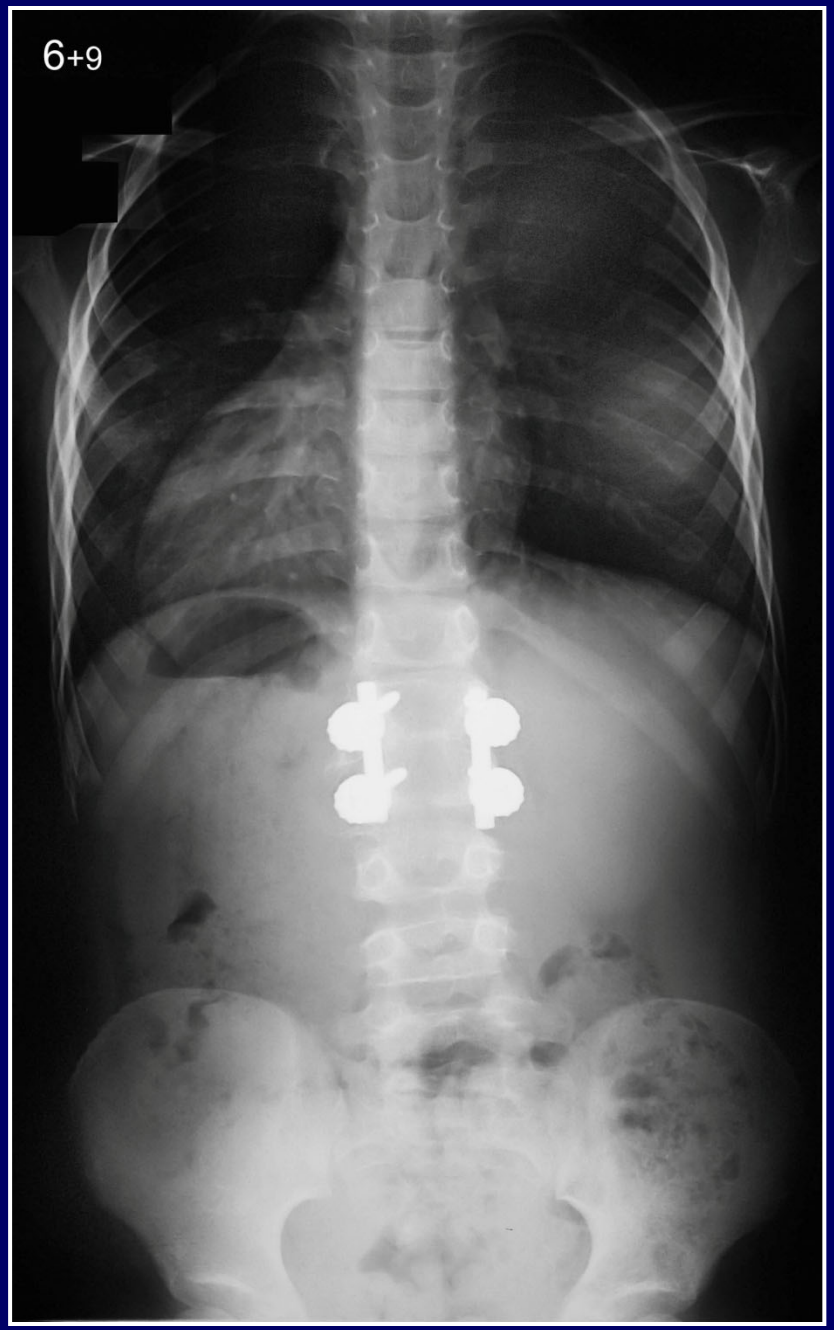
- majority exceed 80° by 11 yrs of age
- highest risk of spinal cord compression



PL Hemivertebra resection

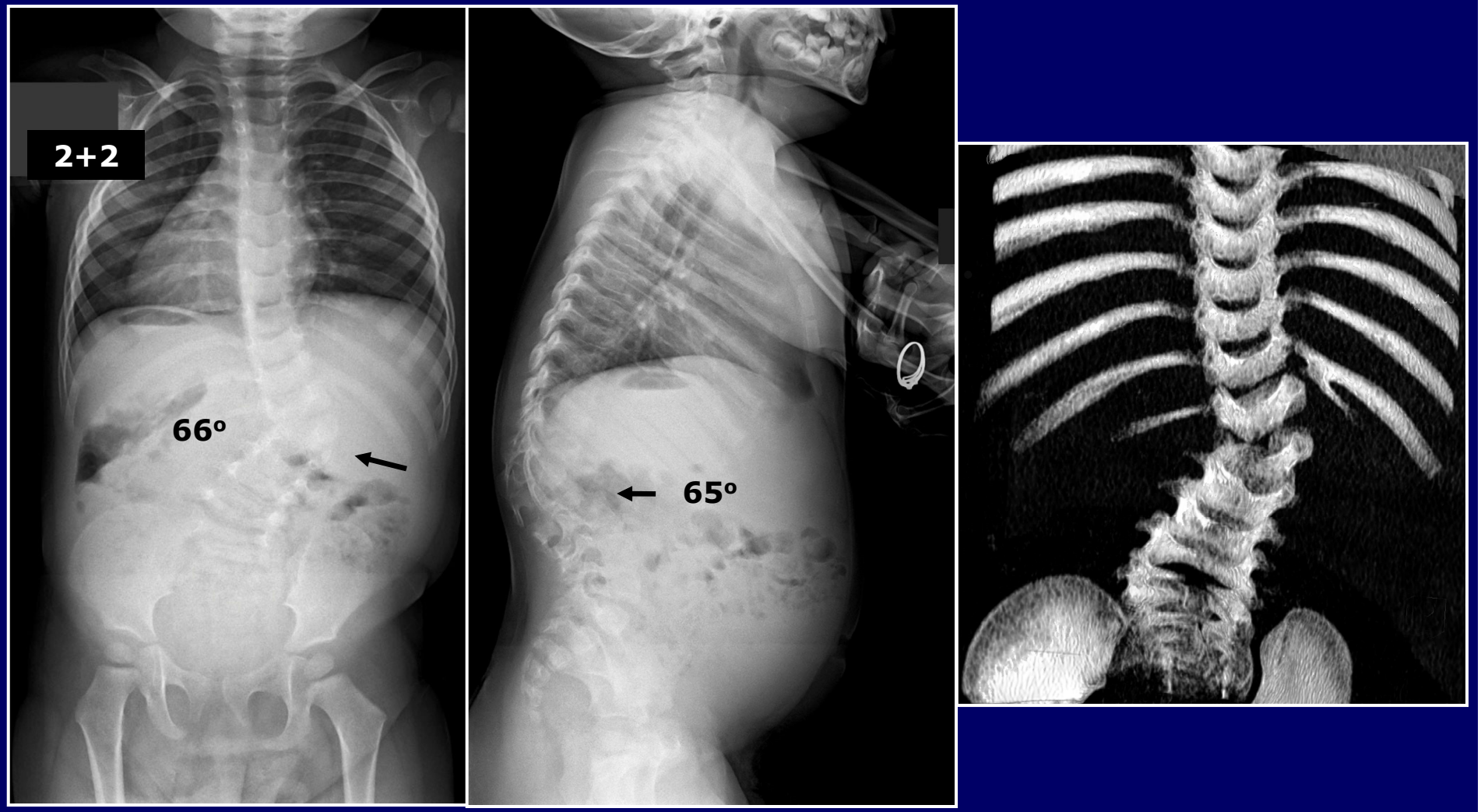


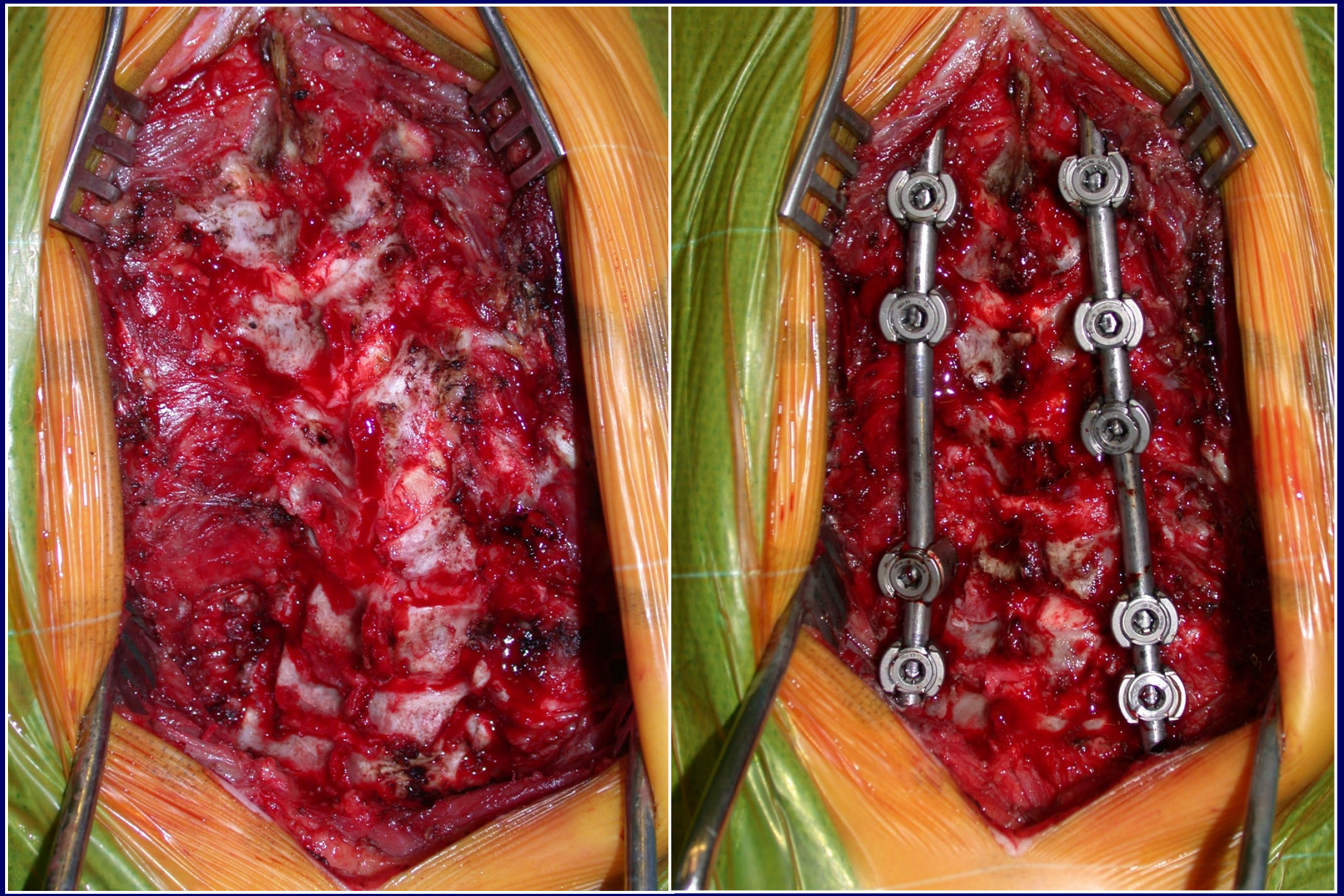


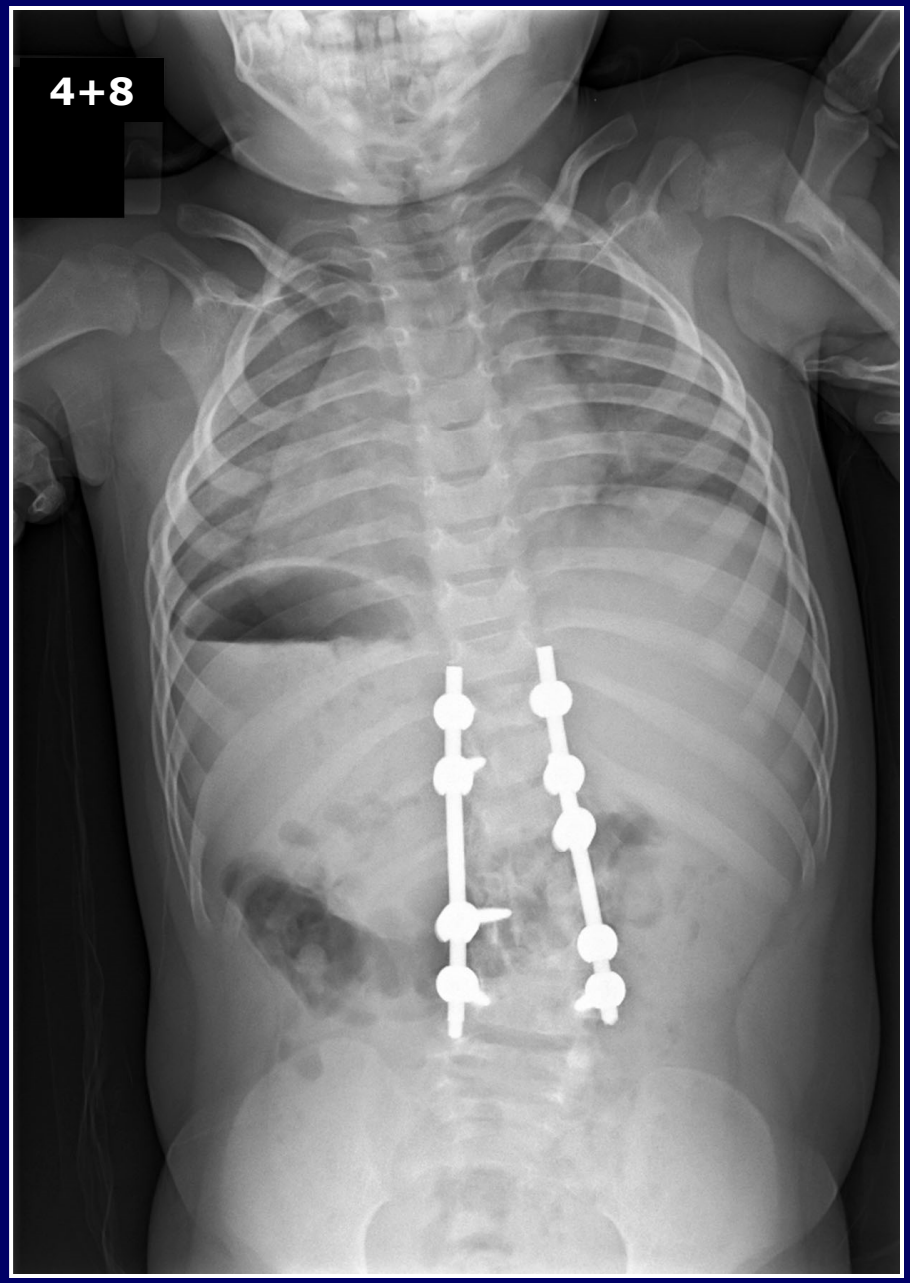

6+9

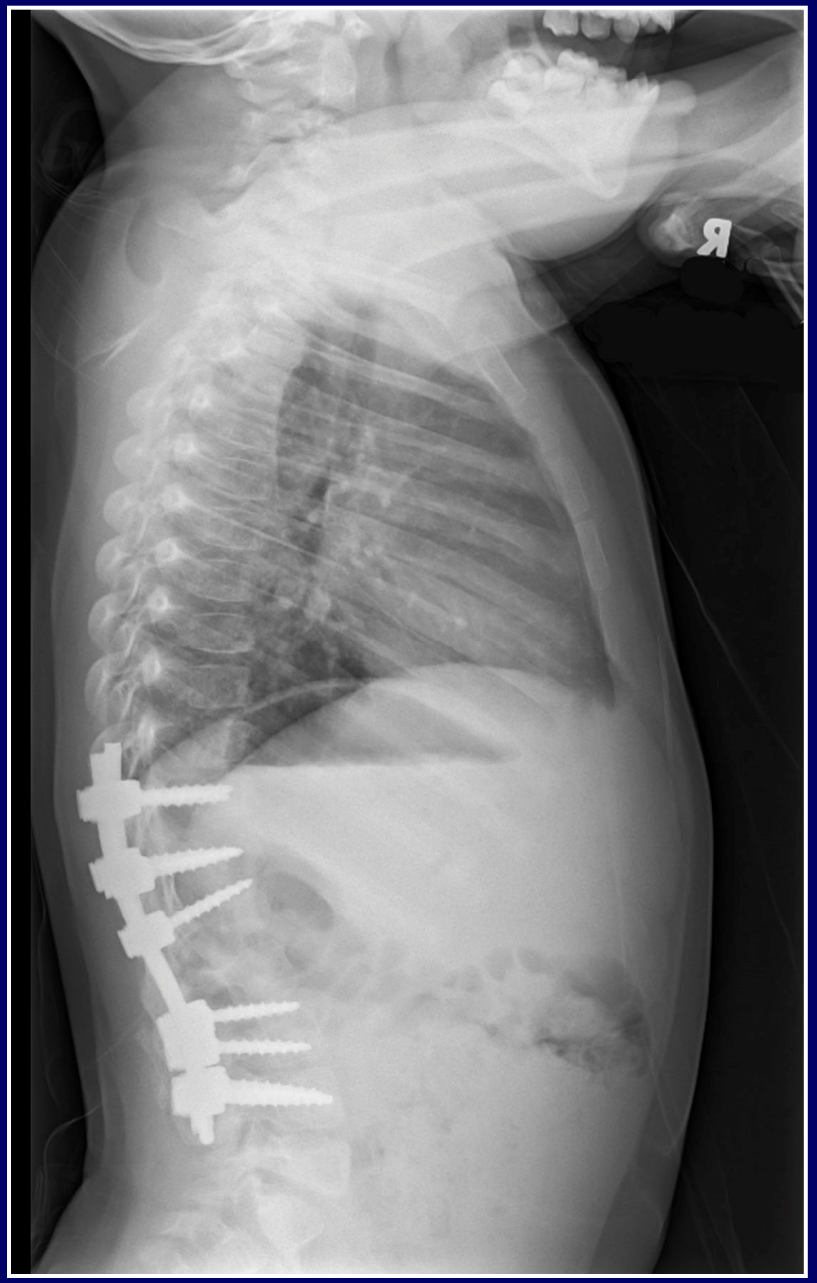

6+9

PL Hemivertebra resection

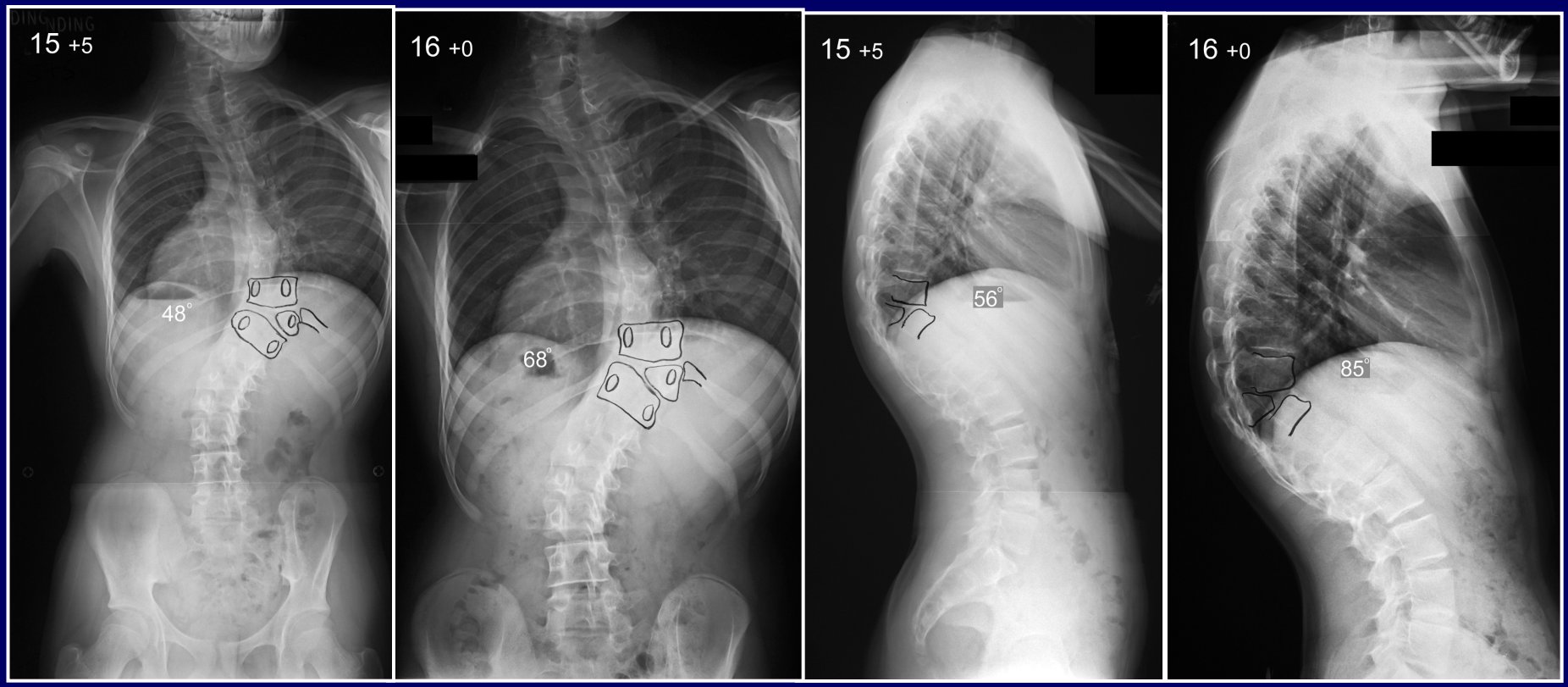


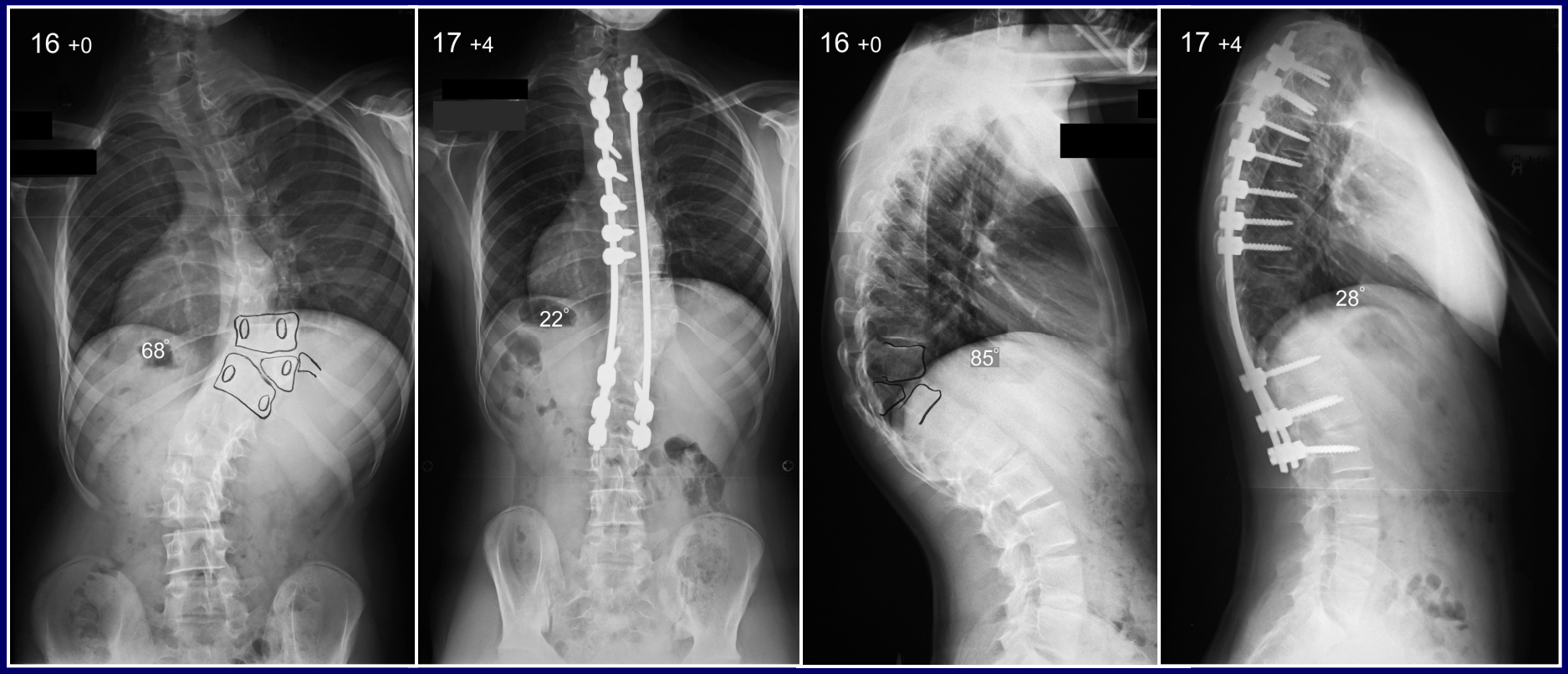



4+8


Я

PL Hemivertebra-late presentation




16 +0
68°
17 +4
22°
16 +0
85°
17 +4
28°

Posterior Hemivertebra

Prophylactic posterior fusion
(Type I and III congenital kyphosis and kyphoscoliosis)

- Indications:
  -children <5 yrs
  -kyphosis up to 50^{o}
- Objective:
  -produce a posterior spinal tether
  -gradual correction with remaining anterior growth
- Considerations:
  -should extend to at least one level above and below the most sagittally tilted vertebra

Prophylactic posterior fusion

- Pseudarthrosis

  -difficult to detect

  -kyphosis progression >5°

  6 months following posterior arthrodesis

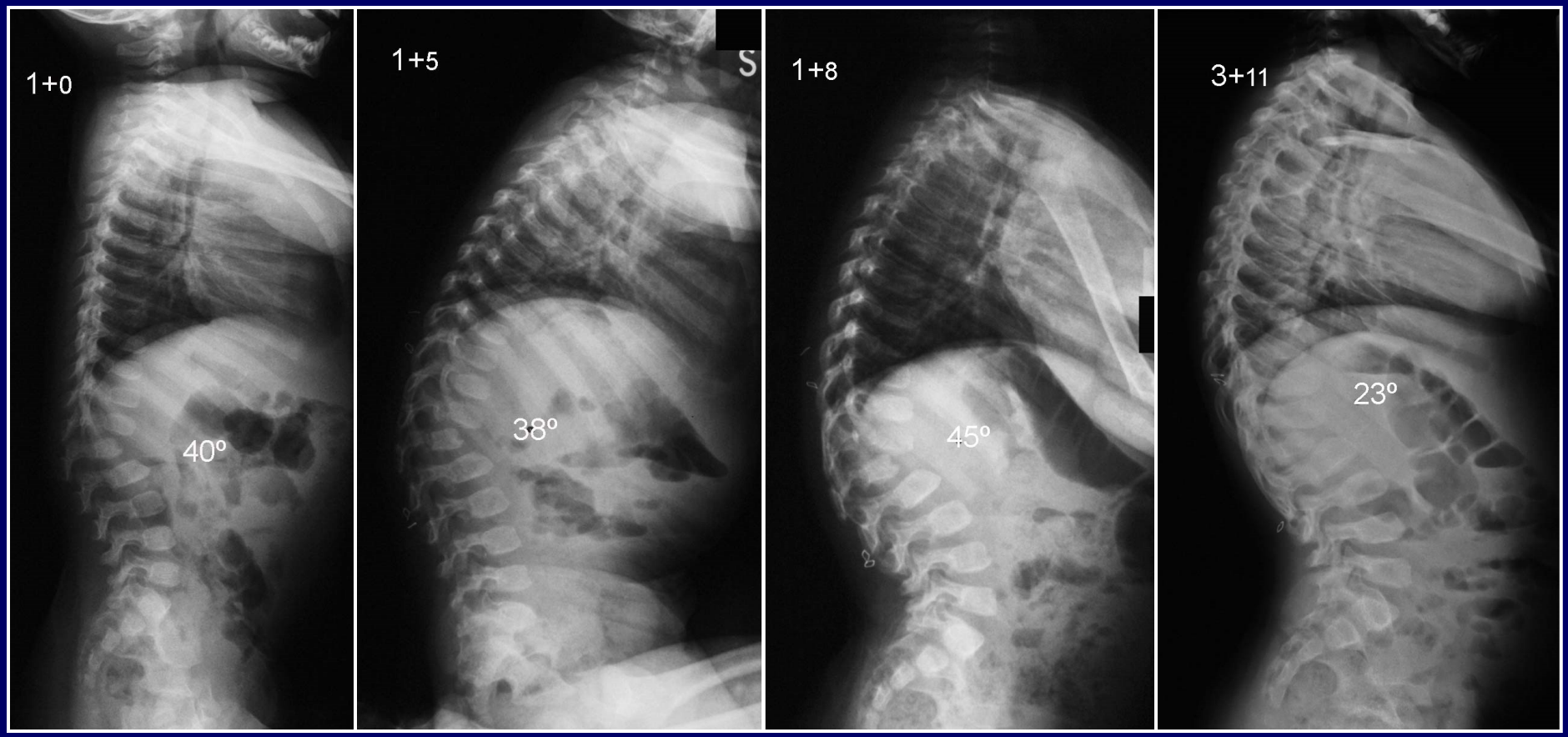
- exploration to repair the nonunion or augment a weak fusion mass

preoperative

postoperative

pseudarthrosis

follow-up

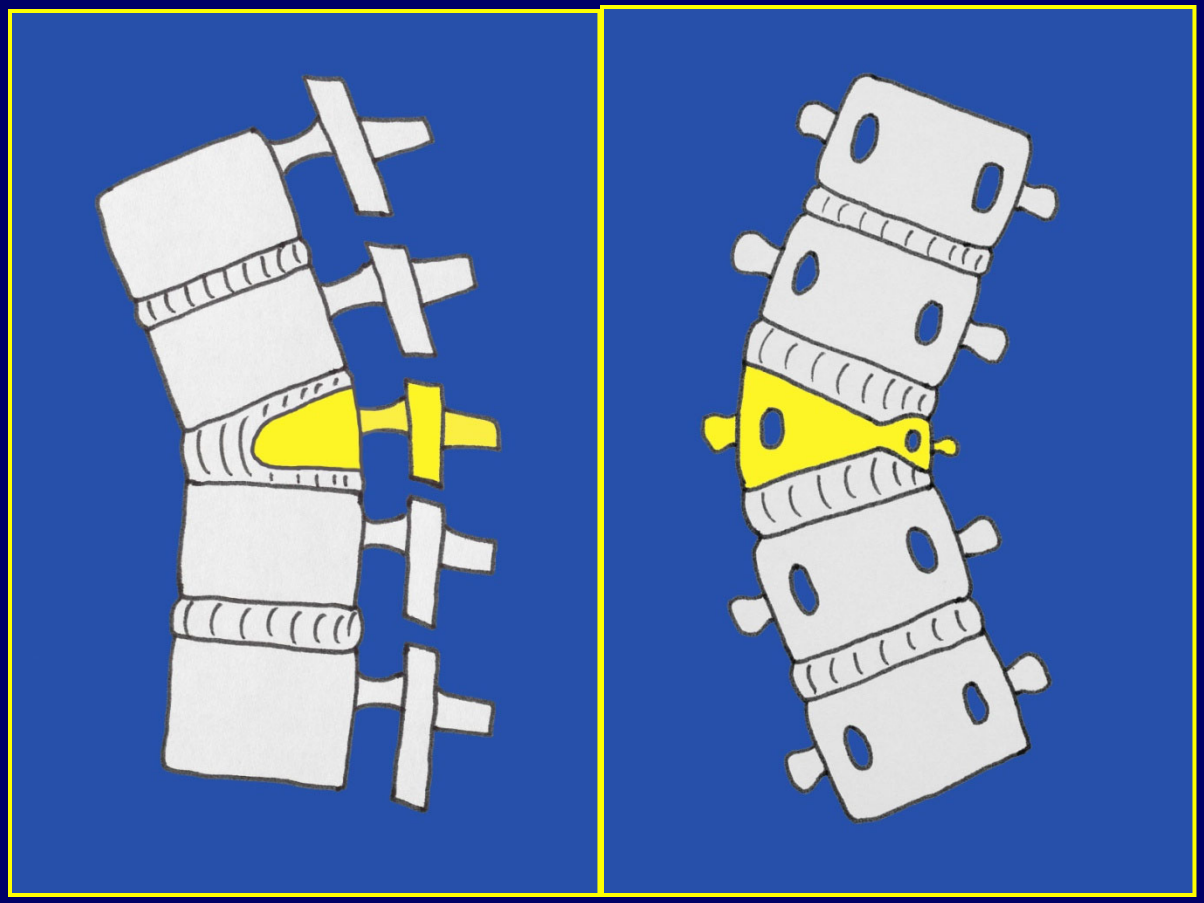


Butterfly (Sagittal Cleft) Vertebra (13%)

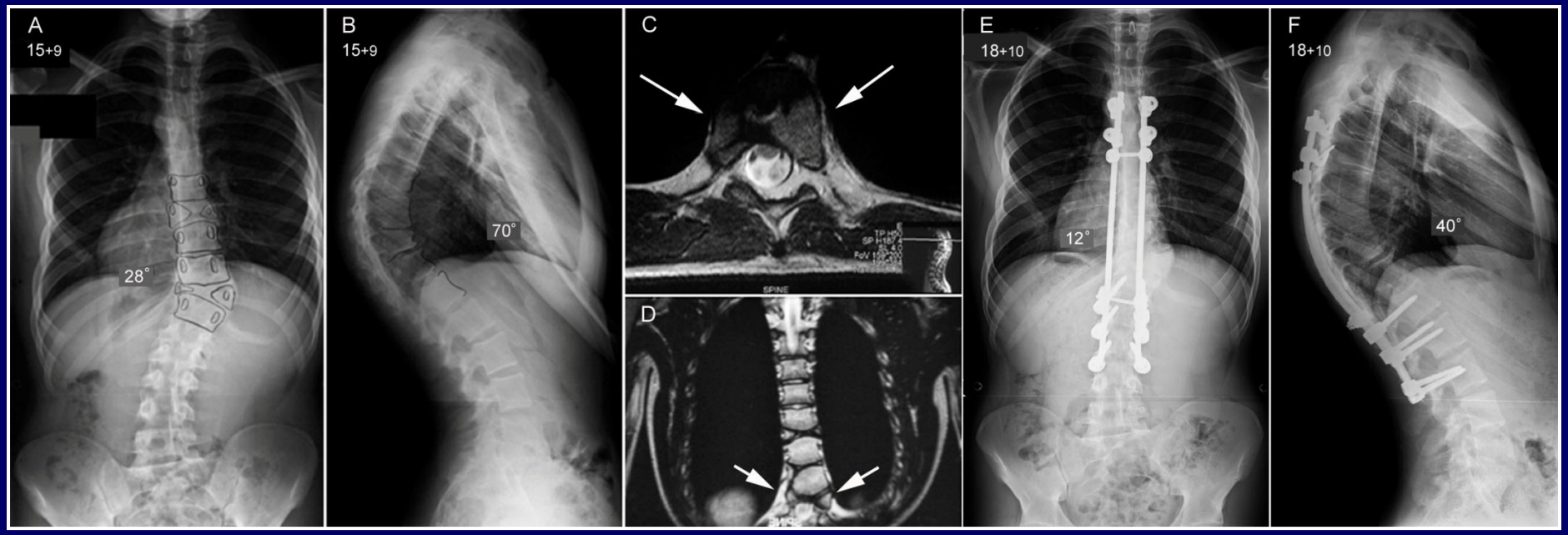
- kyphosis progresses at a mean rate of:

  1.5°/yr (before 10 yrs)

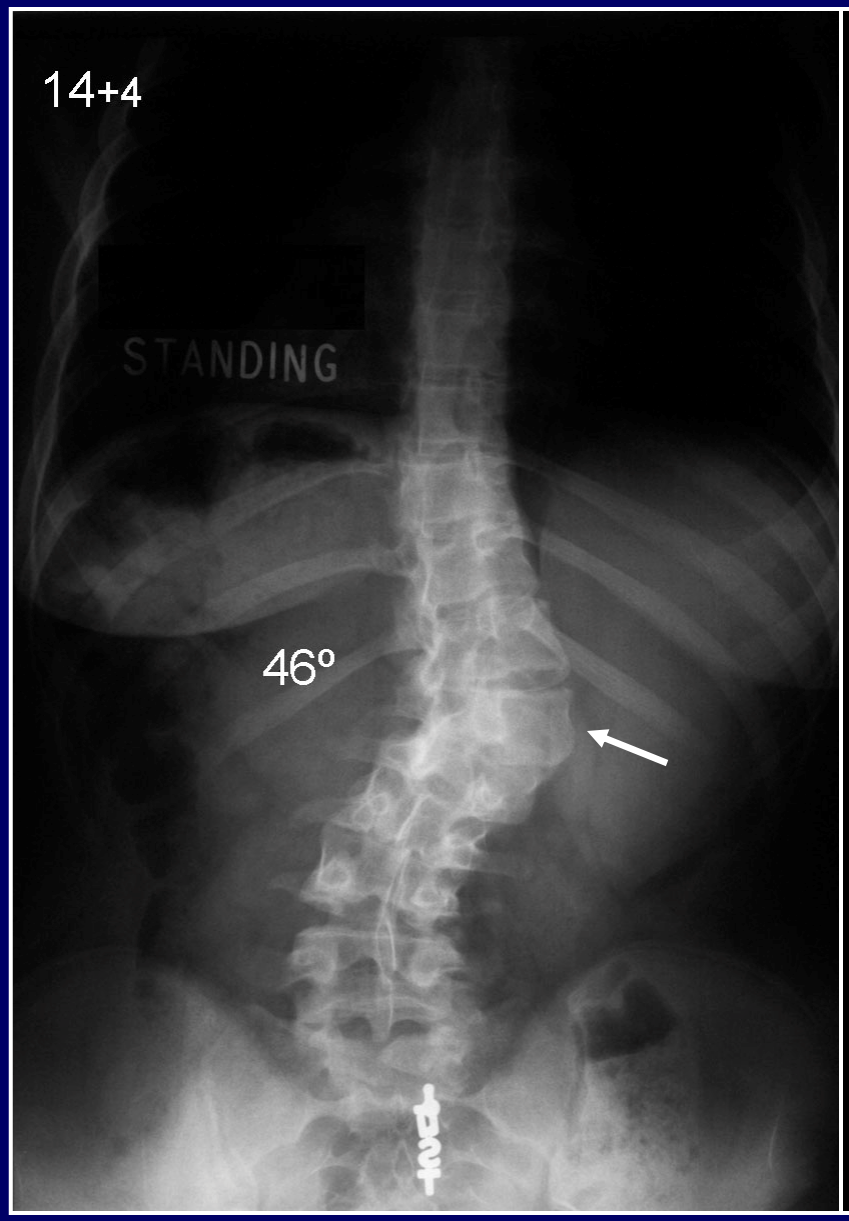
  4°/yr (after 10 yrs)

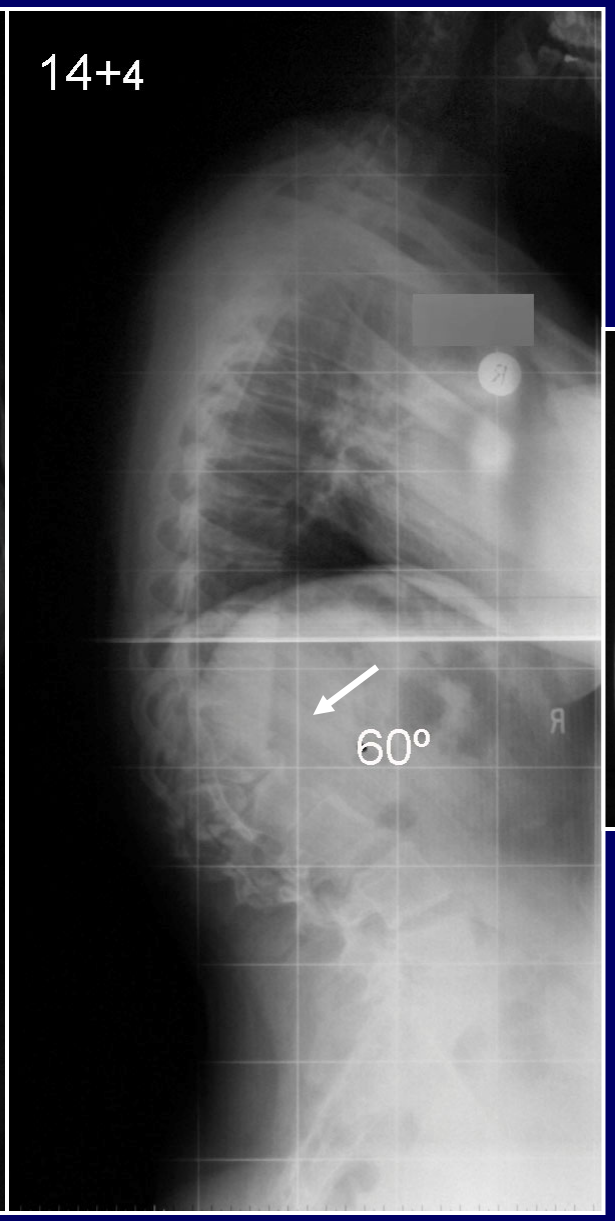


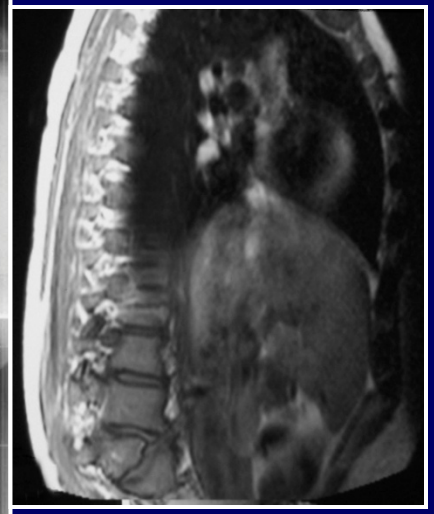
Butterfly vertebra



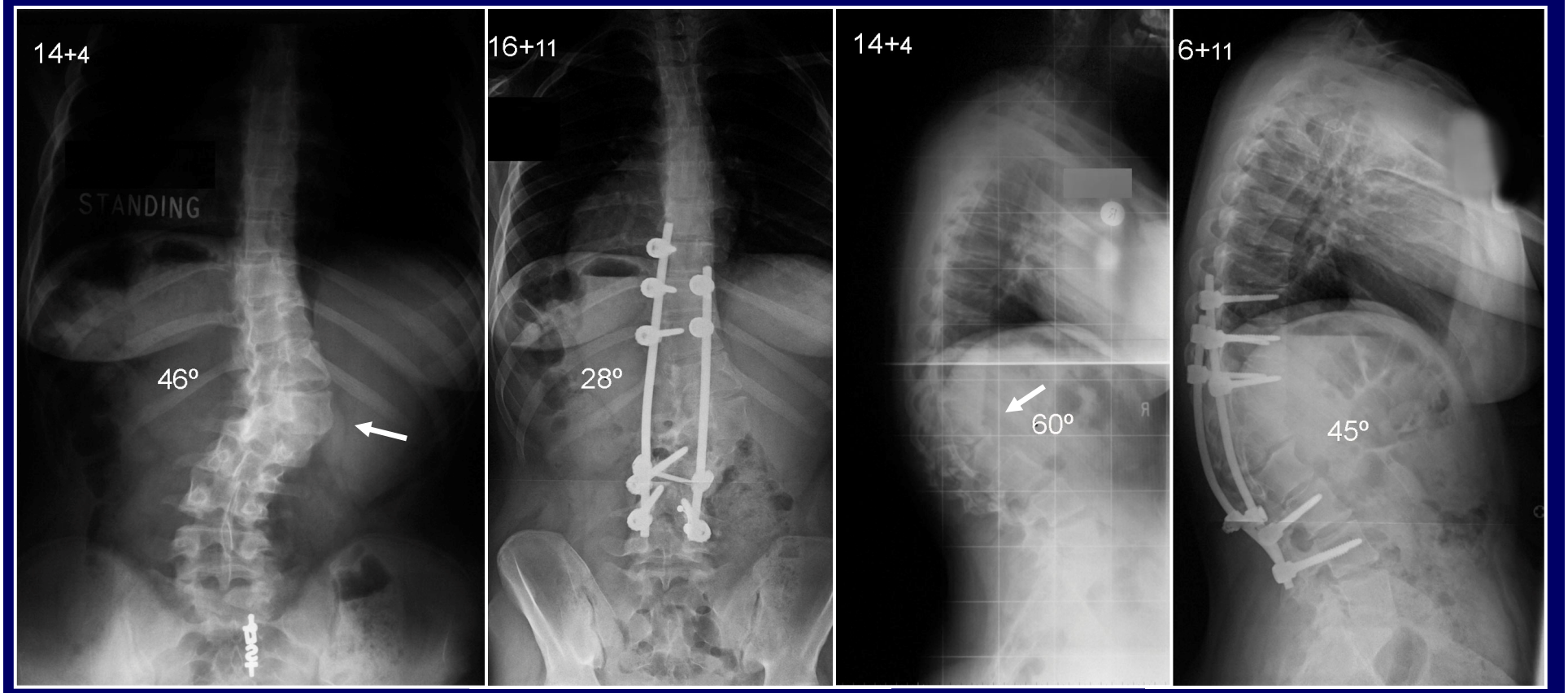
Wedged Vertebra







Posterior Instrumented Fusion

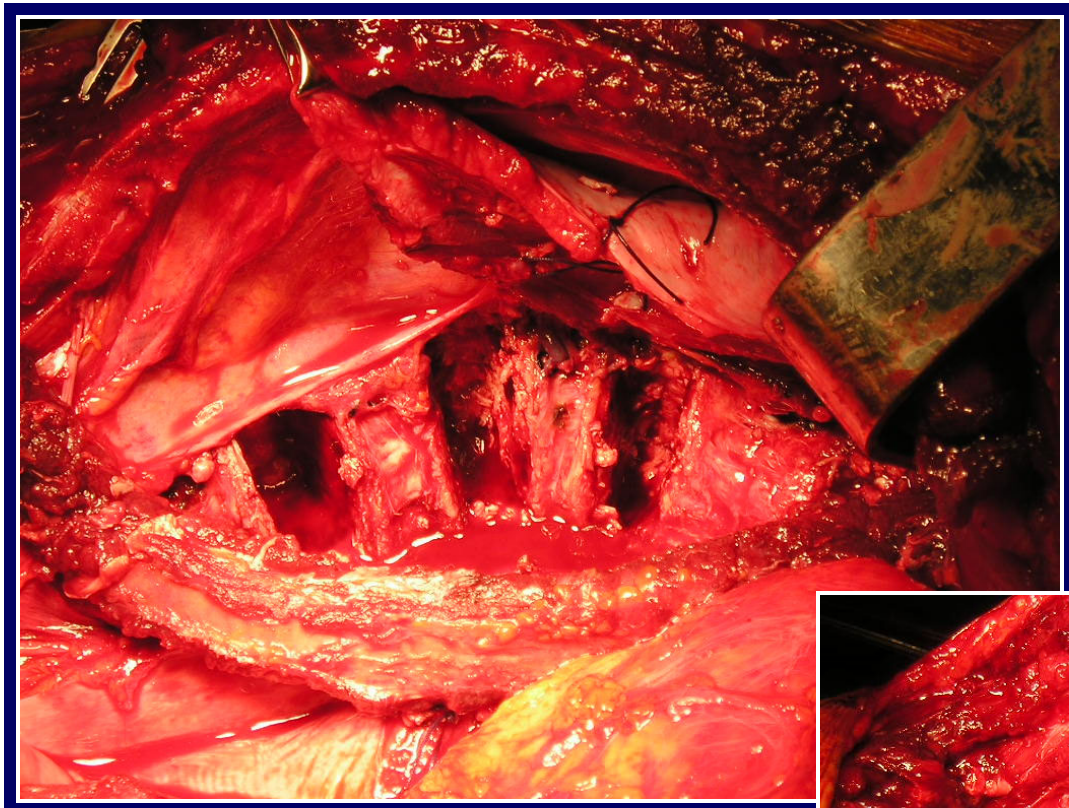


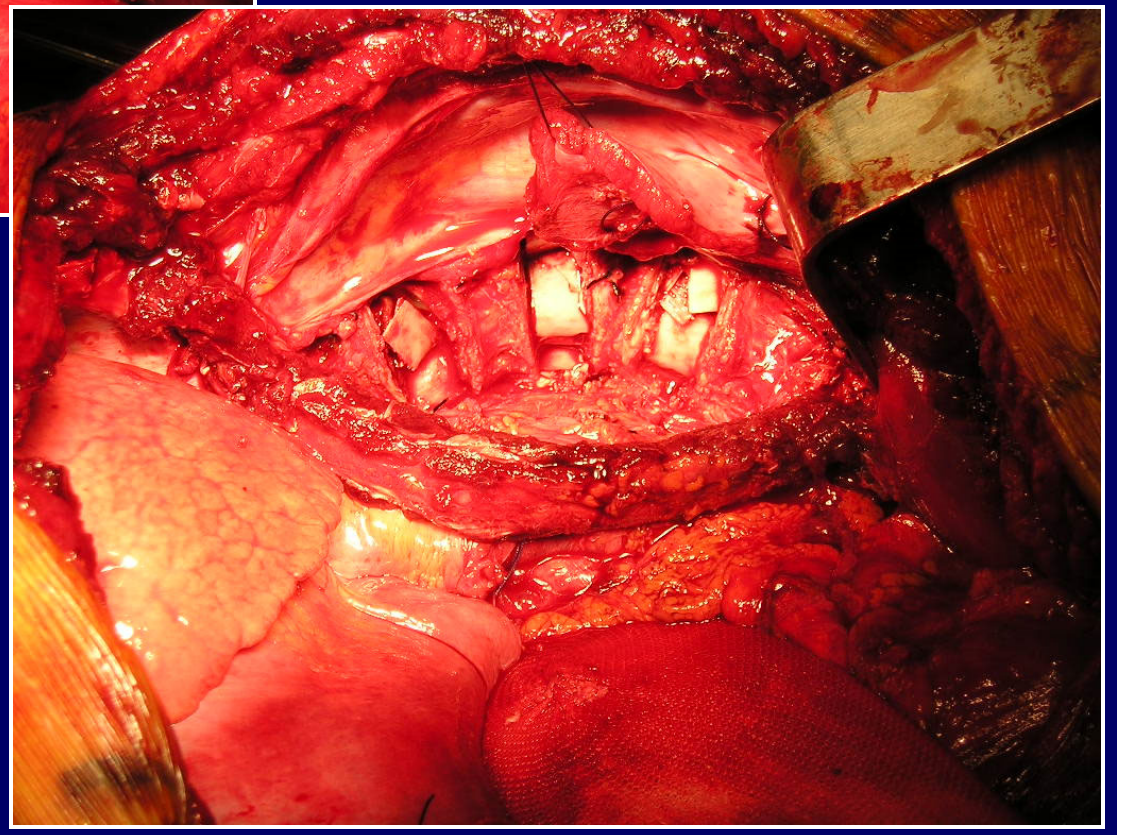
Type I-wedged vertebra

Combined A/P fusion

- Indications:
  -children >5 yrs
  -kyphosis >50-60^{o}
- Objective:
  -partly correct a severe and rigid kyphosis
  -stabilize the spine and prevent neurological complications
- Considerations:
  -anterior strut graft
  -posterior instrumentation

Vascularized rib strut graft (stage 1)

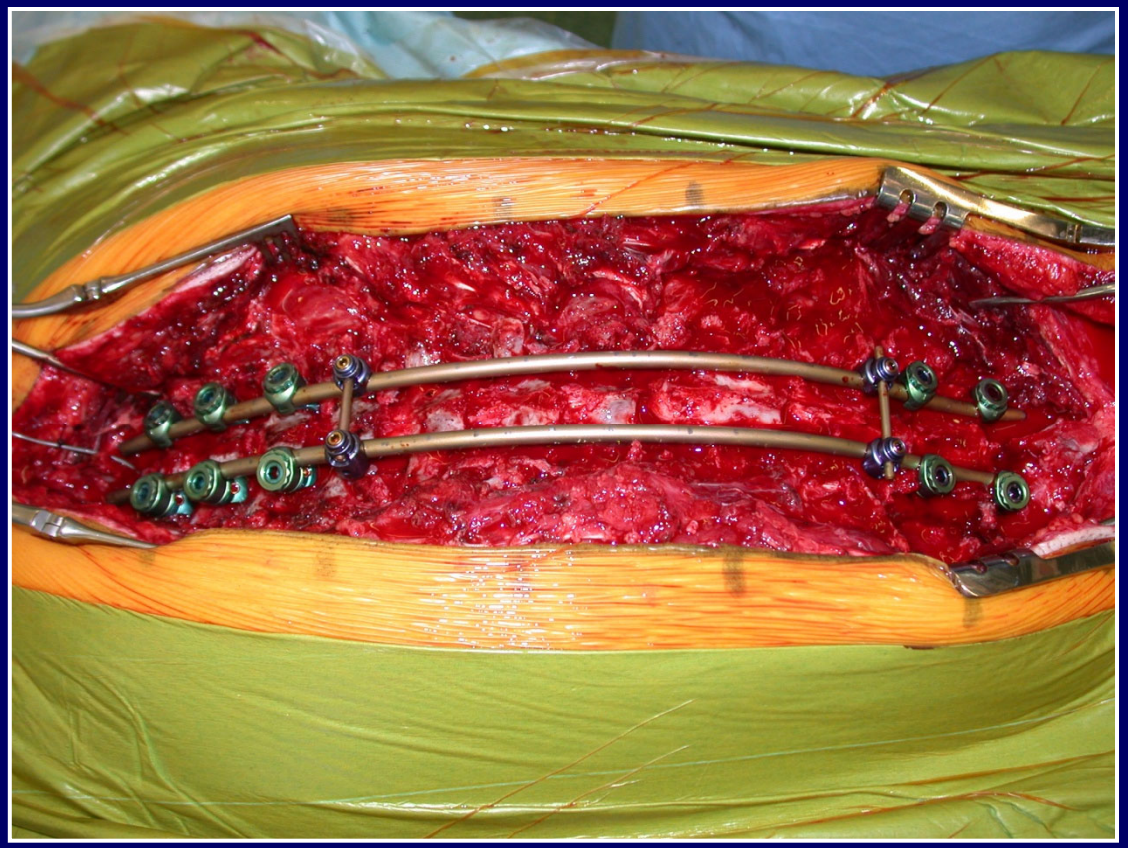




Posterior spinal instrumentation (stage 2)

Problems

- young children
  - -difficult to apply
  - -covers area available for fusion
- large/rigid, upper thoracic kyphosis
  - -less effective
  - -prominent under the skin



VCR and Instrumented Fusion

- Indications:

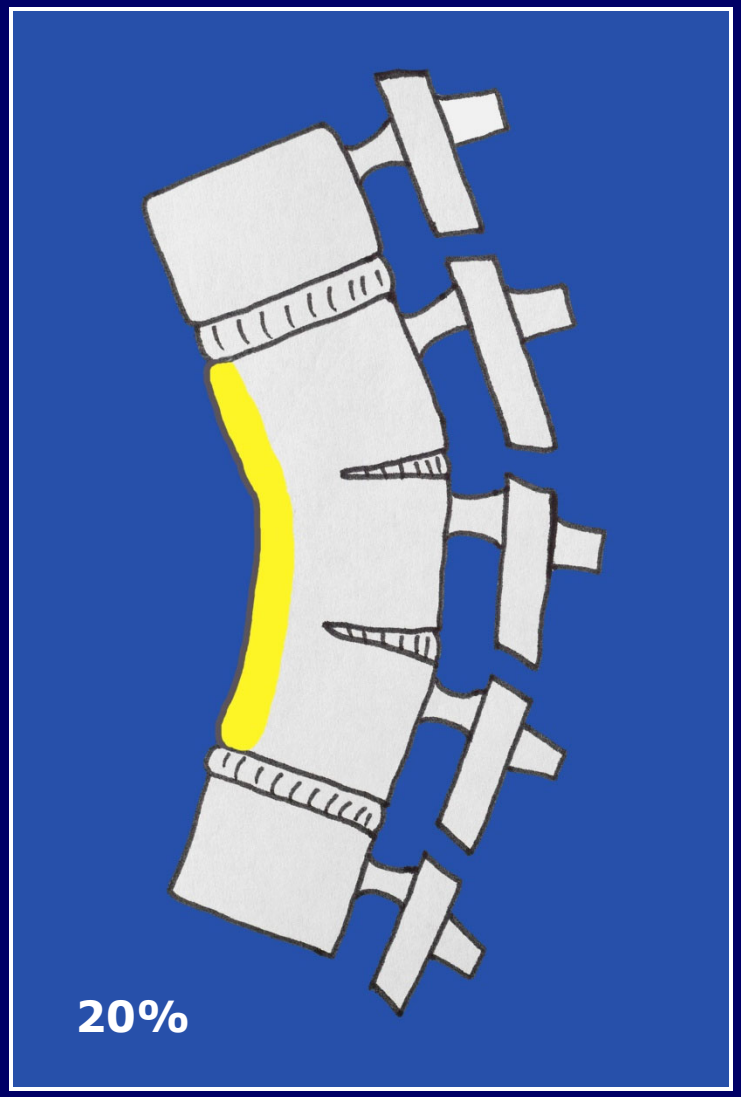
  -alternative to combined A/PSF for very severe and rigid deformities

  -neurological compromise due to spinal cord compression

- Considerations:

  -complex and dangerous procedure with increased neurological risks

Anterior Failure of Vertebral Segmentation (Type II)

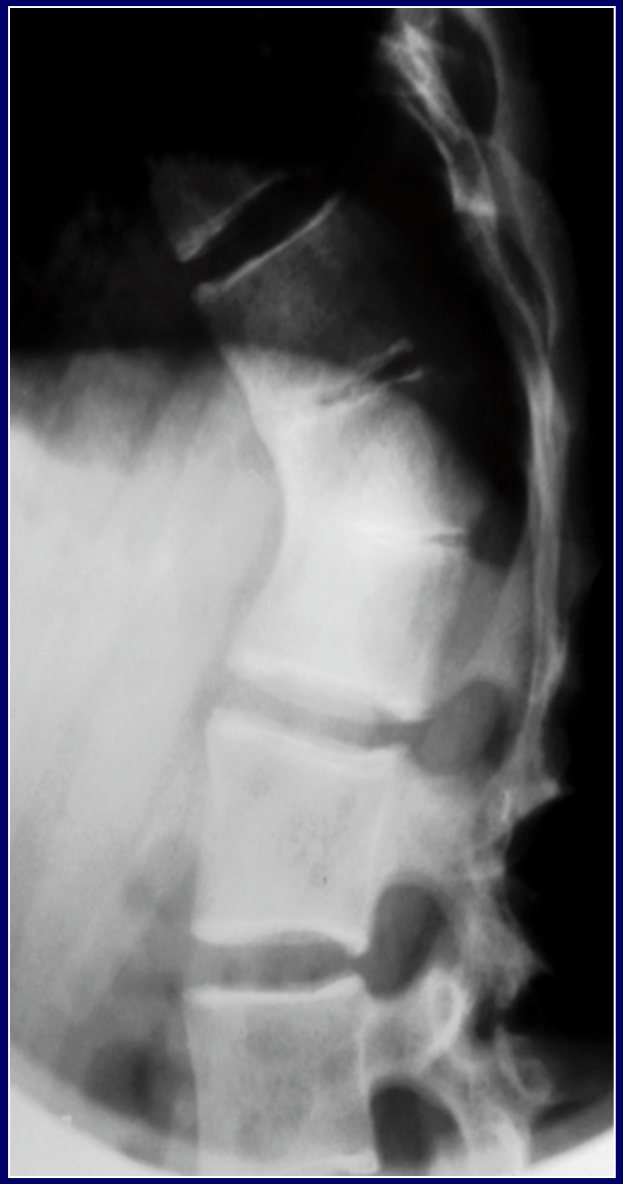


Anterior Unsegmented Bar

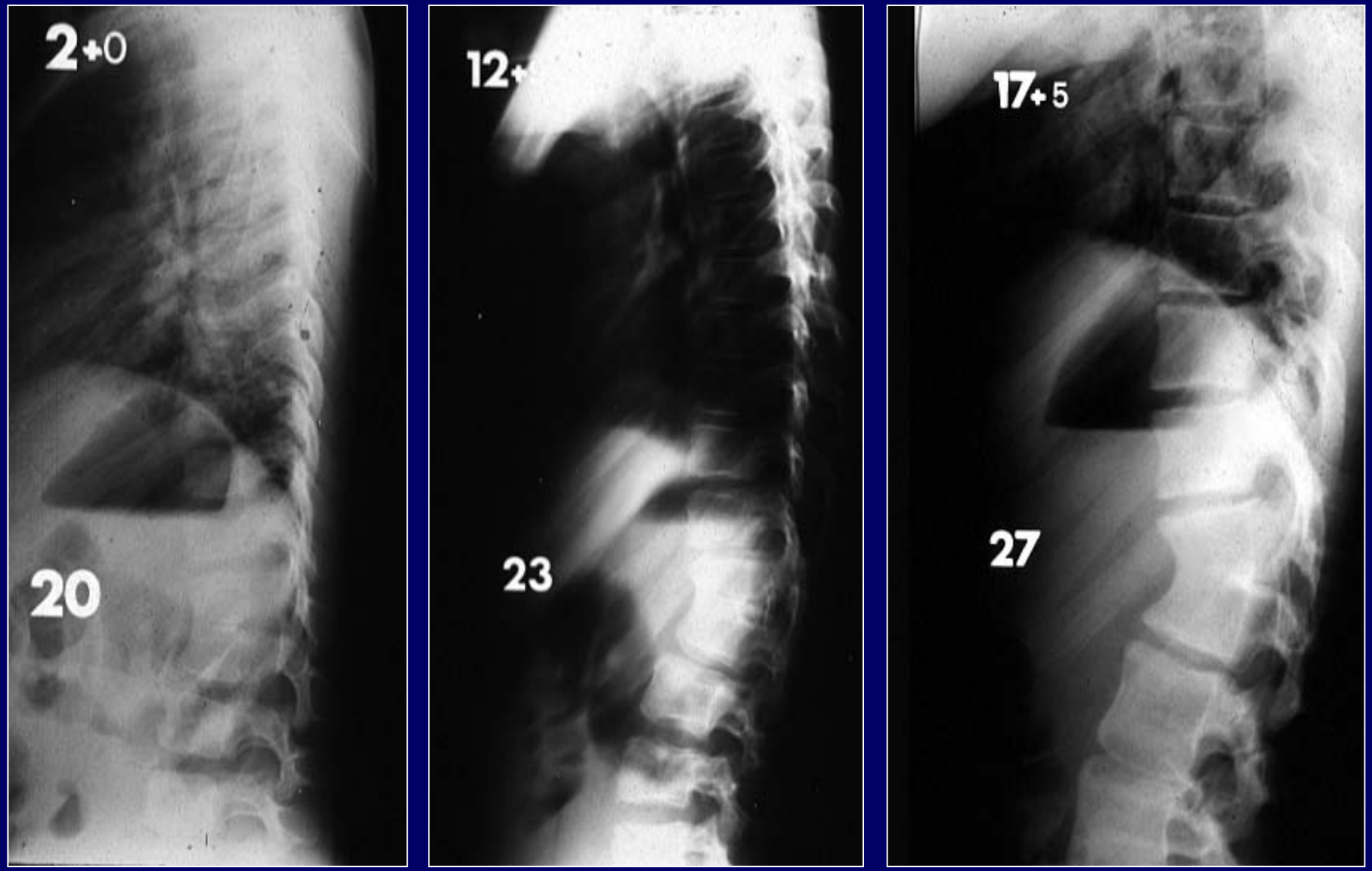
- produces a pure kyphosis
- kyphosis progresses slowly at a mean rate of:

  1^{o}/yr (before 10 yrs)

  $<2^{o}$/yr (after 10 yrs)
- does not cause spinal cord compression



Anterior Unsegmented Bar



McMaster

Anterolateral Unsegmented Bar

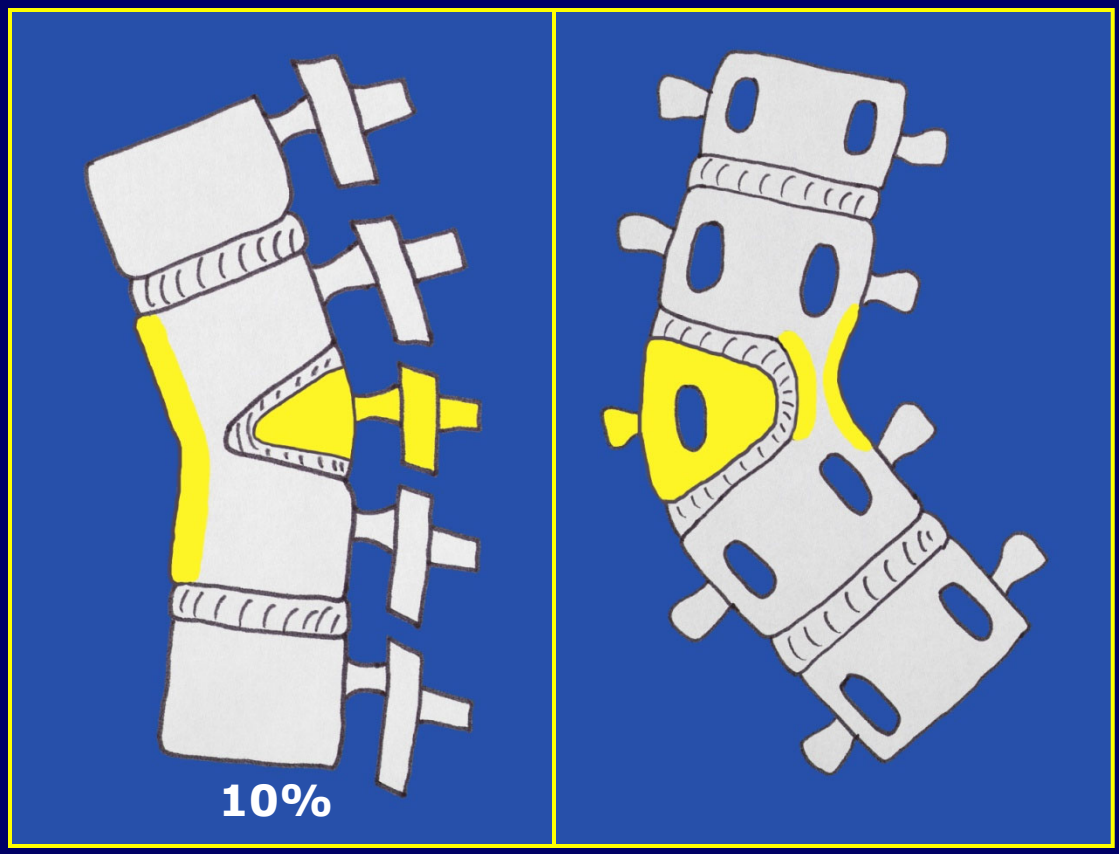
- produces a kyphoscoliosis
- worse prognosis compared to an anterior unsegmented bar
- kyphosis exceeds 90^{o} at skeletal maturity

Anterolateral Bar and Contralateral Quadrant Vertebra (Type III)

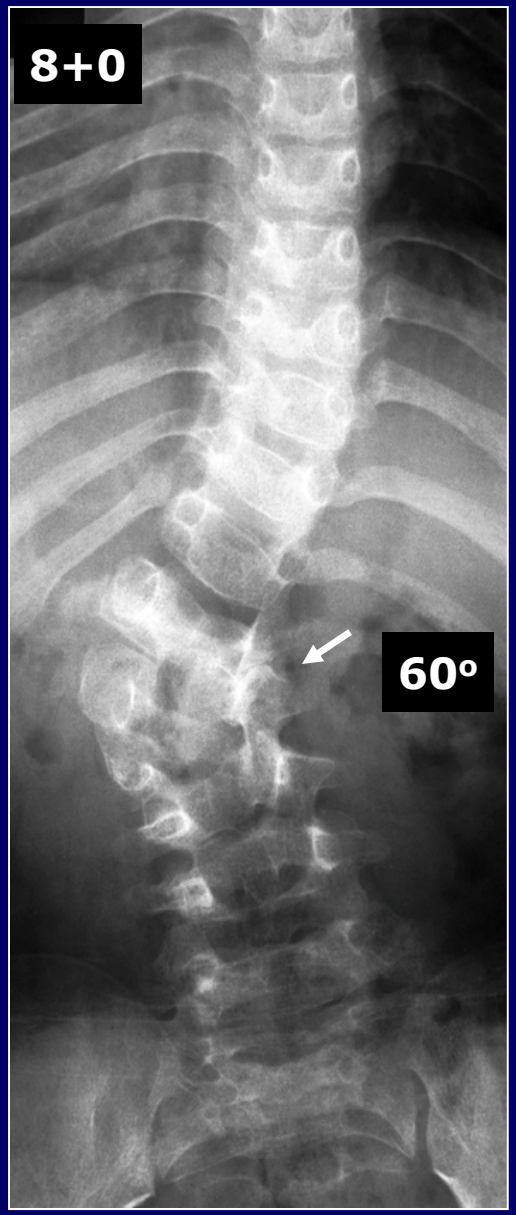
- kyphosis progresses at a mean rate of:

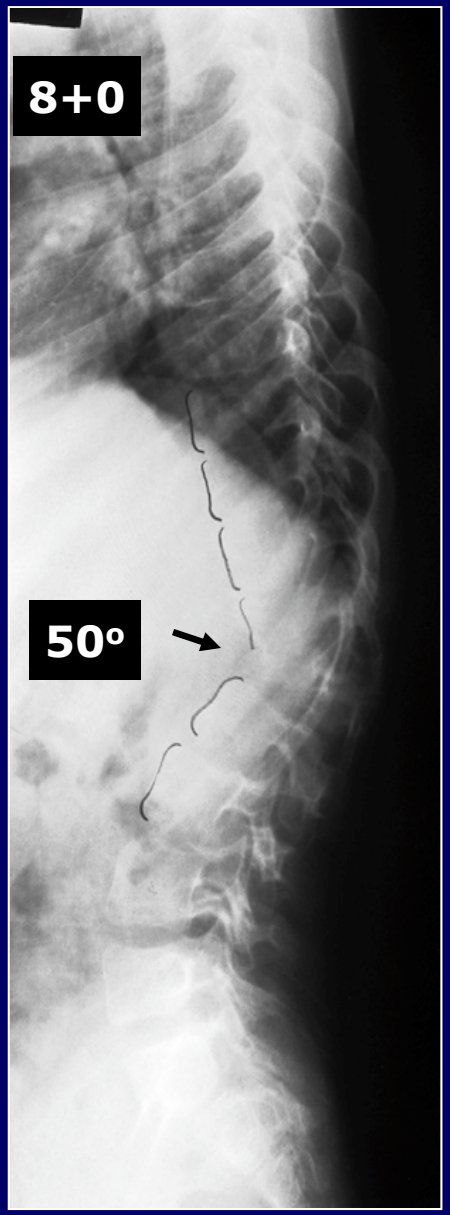
5°/yr (before 10 yrs)

8°/yr (after 10 yrs)



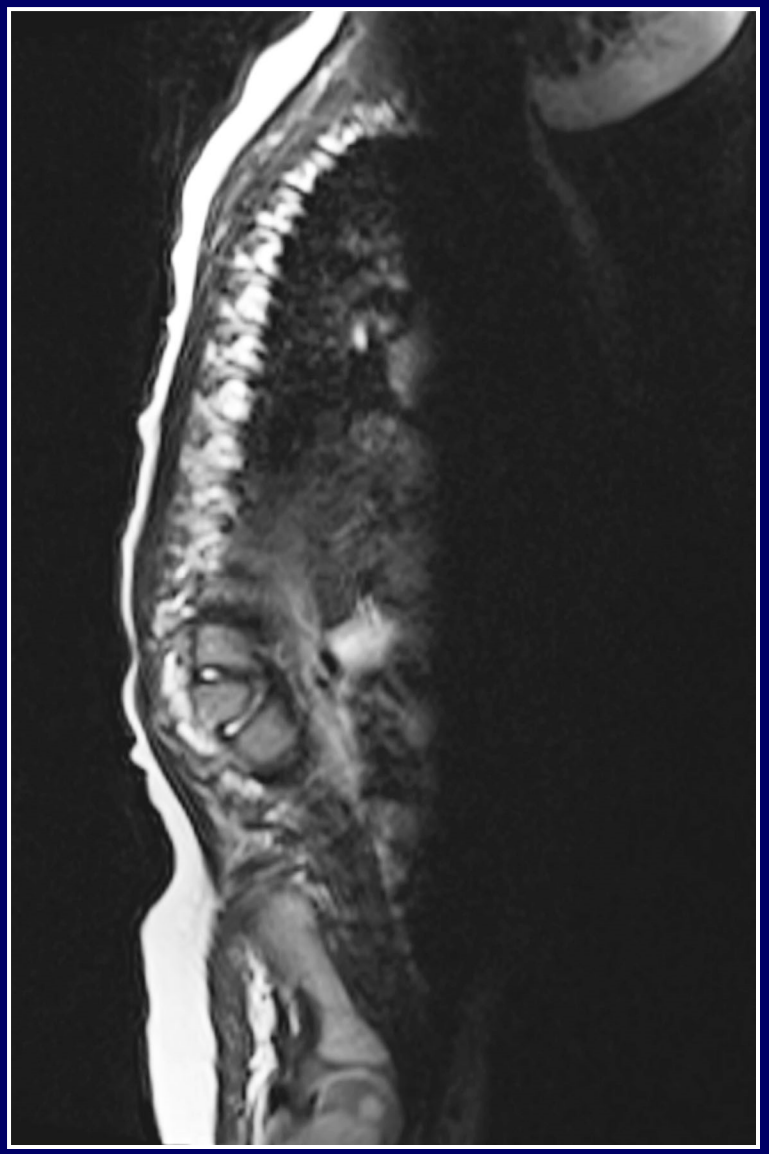
Mixed Anomalies (Type III)





Spinal Cord Compression

- the most dreaded complication
- only occurs with an anterior failure of vertebral body formation (Type I)

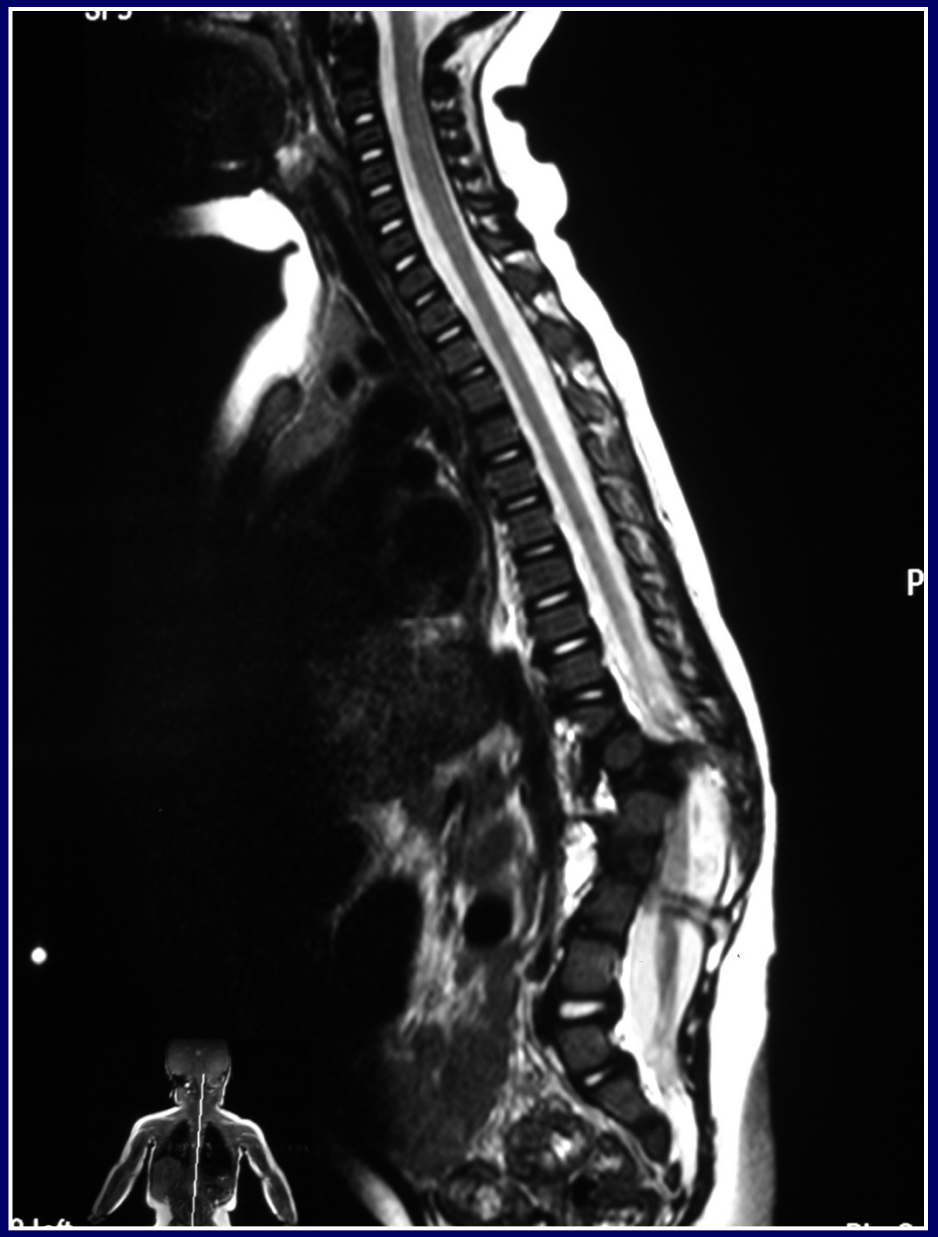


- prevalence of spinal cord compression

  -McMaster and Singh 10%
  -Winter et al. 12%

- onset of symptoms usually during adolescence with variable size of kyphosis

- untreated always results in paraplegia




12+4

Dangerous

- Unilateral unsegmented bar with or without contralateral hemivertebra
- Anterolateral unsegmented bar with or without contralateral hemivertebra
- Fully-segmented hemivertebra (LS)
- Posterior or posterolateral hemivertebra ⟶ neurological complications

Benign

- Block vertebra
- Anterior unsegmented bar
- Unsegmented hemivertebra
- Incarcerated hemivertebra
- Semi-segmented hemivertebra (unless LS)

Surgical treatment

- age of the patient
- type of vertebral anomaly
- type and size of deformity
- presence of spinal cord compression

Select the correct procedure and apply at the appropriate time

Treatment Principles

- not possible to create growth of the spine on the deficient side
- early surgical treatment is required to balance spinal growth and prevent a severe deformity and spinal cord compression
- anticipate deformity progression based on patient age and type of anomaly


Thank you for your attention