Spinal Deformity Treatment in Children with Myelomeningocele-the BCH Experience

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Disclaimers

Detethering of the spinal cord to treat scoliosis: the role of serial urodynamic data for patient selection

The use of computer generated 3-D models of the spine in the surgical treatment of spinal deformity

Spinal Cord Detethering in Children with Myelomeningocele for the Treatment of Scoliosis

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McLone DG, et al.: Tethered Cord as a Cause of Scoliosis in Children with a Myelomeningocele. Pediatr Neurosurgery 1990

- **30 myelomeningocele with scoliosis as one of presenting signs – lumbar level**

- **At one year**
  - <50˚ :23/24 stable or improved
  - >50˚ :5/6 progressed
- **Data suggest scoliosis due to tethered cord and support (1) early untethering even when scoliosis is the only finding AND (2)repeat untethering with progression after 1 year**

Pierz,K: The Effect of Tethered Cord Release on Scoliosis in Myelomeningocele.JPO. 2000

21 patients (18 myelomeningocele)

- **Follow-up 5 yrs (2+5-9+9)**
- **10/21 required spinal fusion**
- **6 stable, but only one had scoliosis>30˚ - and that patient had thickened filum only**
- **Thoracic level- all had spinal fusion**
- **Lumbar level- 58% progressed**
- **Sacral level- 33% progressed**
- **8 surgical complications in 6 patients**
- **"….scoliosis alone must be analyzed critically before recommending such surgery."**

Bowman RM:Tethered cord release: a long-term study in 114 patients.J Neurosurg Ped; 2009

- **36 patients**
  - Improved – 8
  - Stable-4
  - 11 progressed (mean 26 °) but no spinal susion as yet
  - 13 progressed (mean 34°) and had spinal fusion
- **<u>Untethering does not prevent need for spinal fusion in the majority of the patients</u>**
- **<u>Insufficient follow-up to determine if untethering slows progression to allow fusion at maturity</u>**

? Methods

- Defined scoliosis? (10˚ not appropriate)
- Inadequate follow-up
- Varied population
- Importantly- ? Patient selection: Since all children with myelomeningocele have an anatomically tethered spinal cord, how do you know when the tether is responsible for the curvature.

Spinal Cord Detethering for the Treatment of Scoliosis in Children with Myelomeningocele

- Does it stabilize or improve the deformity?
- Does it slow the progression to allow delay in stabilization surgery?

METHODS

- Myelomeningocele – open neural tube defect
- Skeletal Immaturity
- Minimum 2 y follow-up
- Scoliosis defined as 20˚or more Cobb
- Eliminate other causes (shunt, Chiari..)
- <u>Additional findings (other than scoliosis) to suggest symptomatic tethering</u>
- <u>Serial urodynamic studies</u>
- Define progression and improvement as 10˚ change

RESULTS

Demographics

- **20 Children (9 boys, 11 girls)**
- **Age: mean 5.6 years (1.0- 13.6) at time of detether**
- **Follow-up mean 4.3 years (2.0- 8.5)**

Confirmatory Symptoms

Pain	5
Motor Change	7
Spasticity	2
Foot Deformity	1
UDS	17
hypertonicity	12
fibrillations	5
Synergy/Dyssynergy	1

Curvature Response

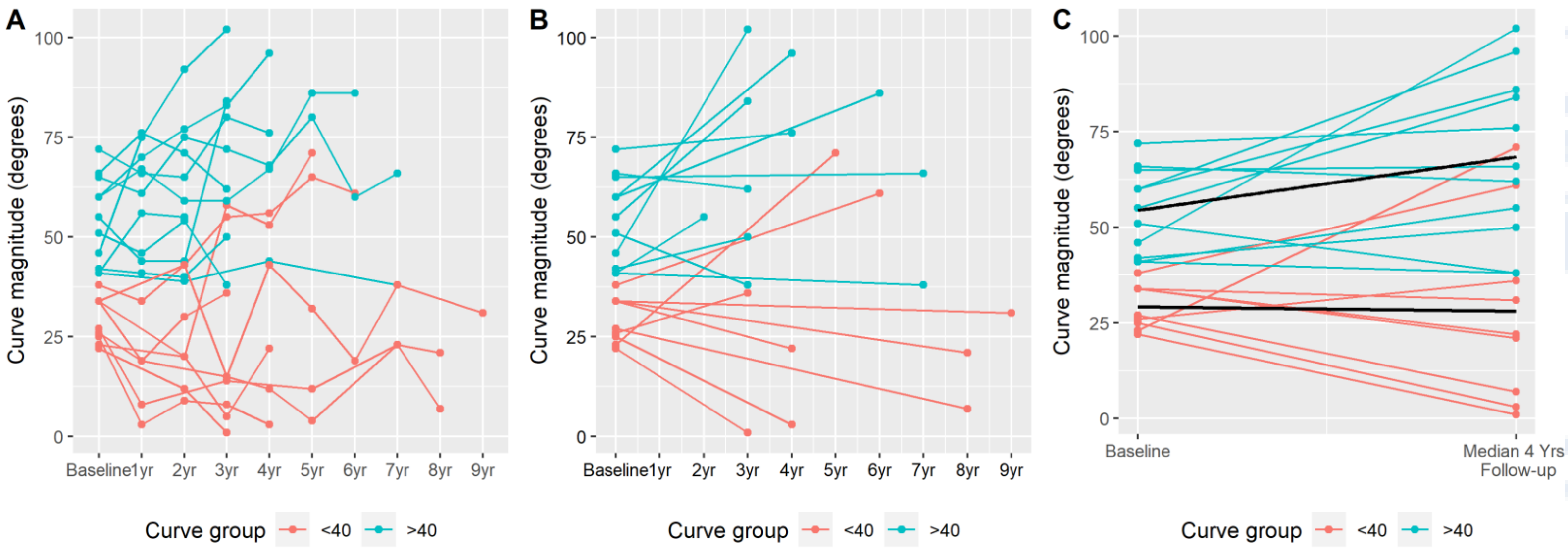
- **Improved: 6 at mean 5.2 years**
- **Stable: 7 at mean 3.4 years**
- **Progressed 7 at mean 4.4 years**

Relationship to Scoliosis Magnitude

- **< 40˚:**
  - Stable or improved 7
  - Progressed 2

- **=/> 40:**
  - Stable or Improved 6
  - Progressed 5

Detethering and Curvature Response



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Stable or Improved

Z.P.

?

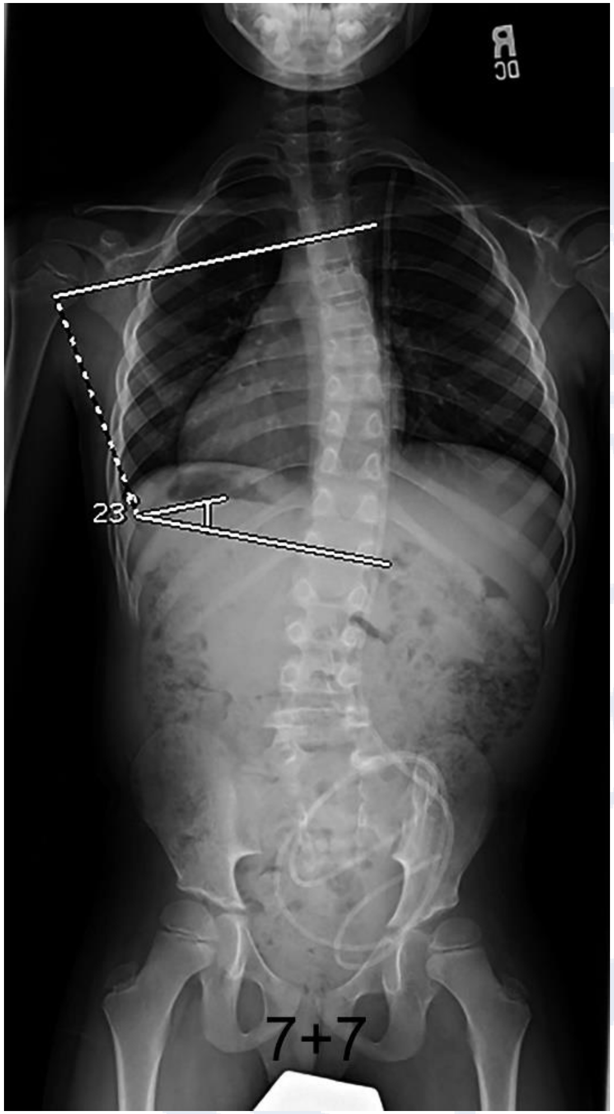
7+11

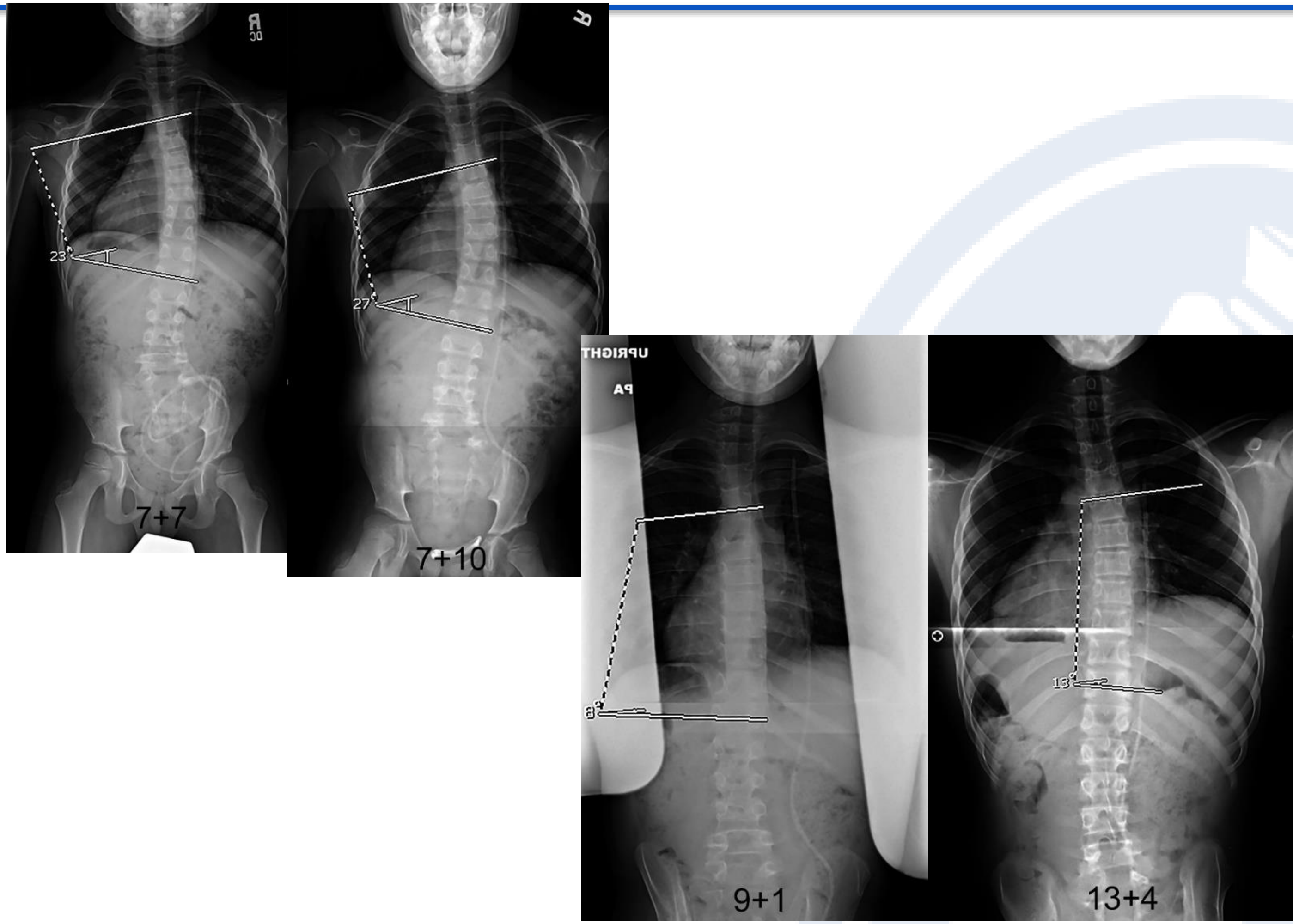
27°

Falling

Scoliosis

UDS – FIBRILLATIONS




23°
7+7
27°
7+10
8°
9+1
13°
13+4


Acq Date/Time: 11/0


XR-Spine Entire 2 or 3
Acq Date/Time: 01/29

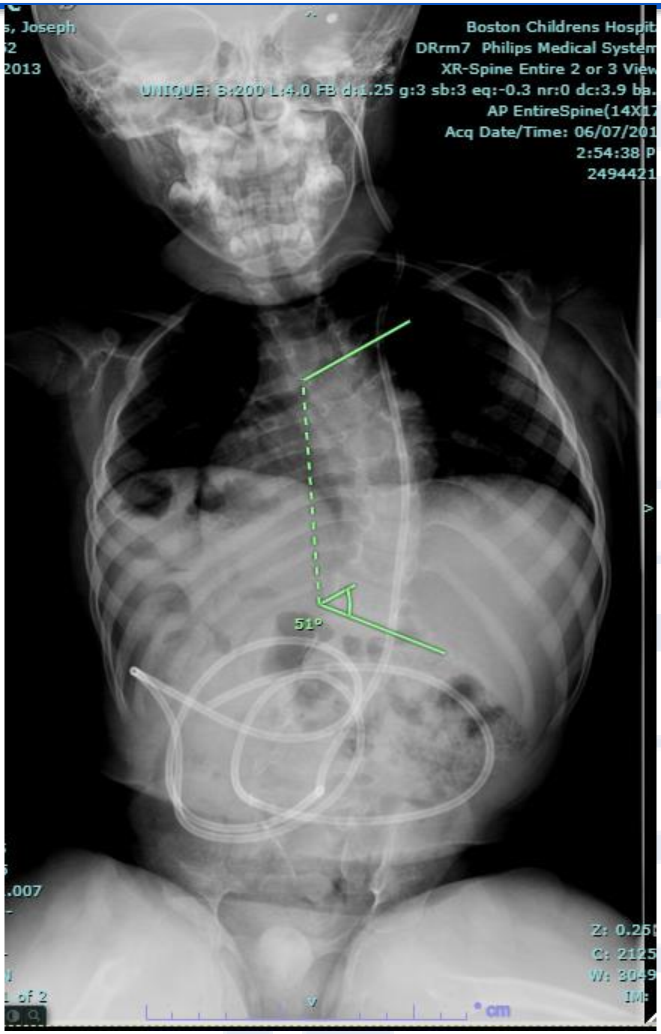
JC

4+2 Y

51˚

Back Pain

L.E. Spasticity



JC

6+2 Y

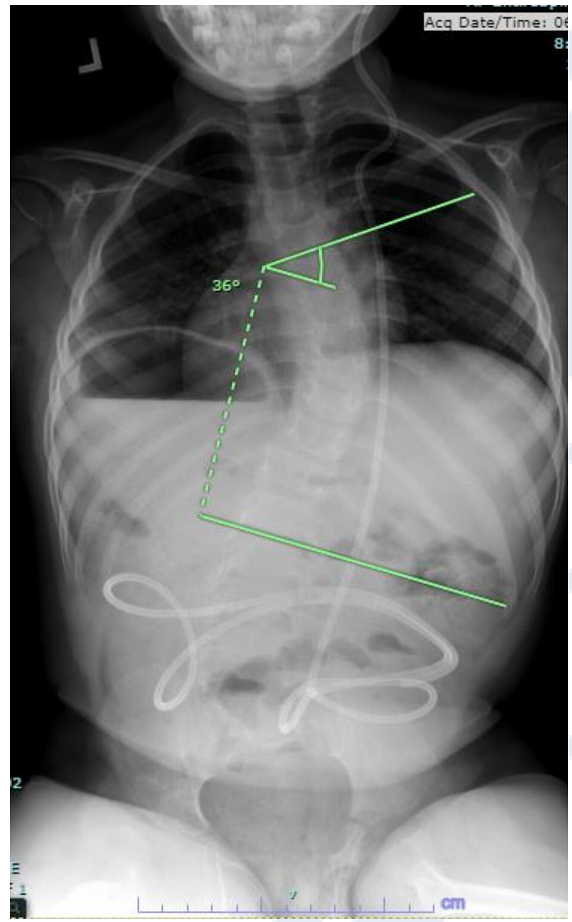
54˚


5/2013
XR-Spine Entire 2
SCOLIOSIS PA
Acq Date/Time: 07
54°
425\424

JC

7+2

36˚



Progressed

J.L.

1+2 y- untethered

scoliosis

bladder contractions

5+5y- untethered

gait change

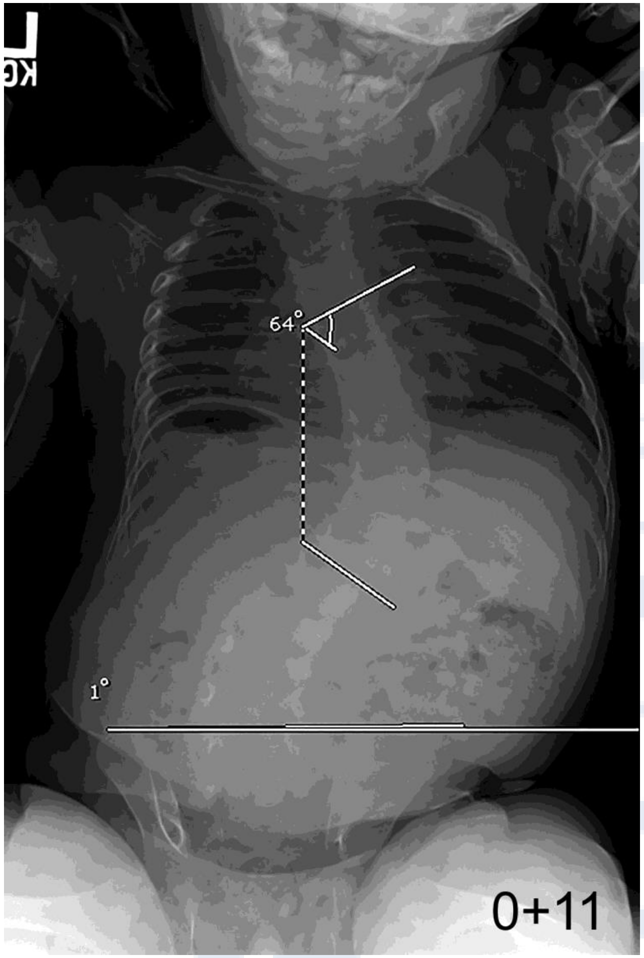
dyssynergy-

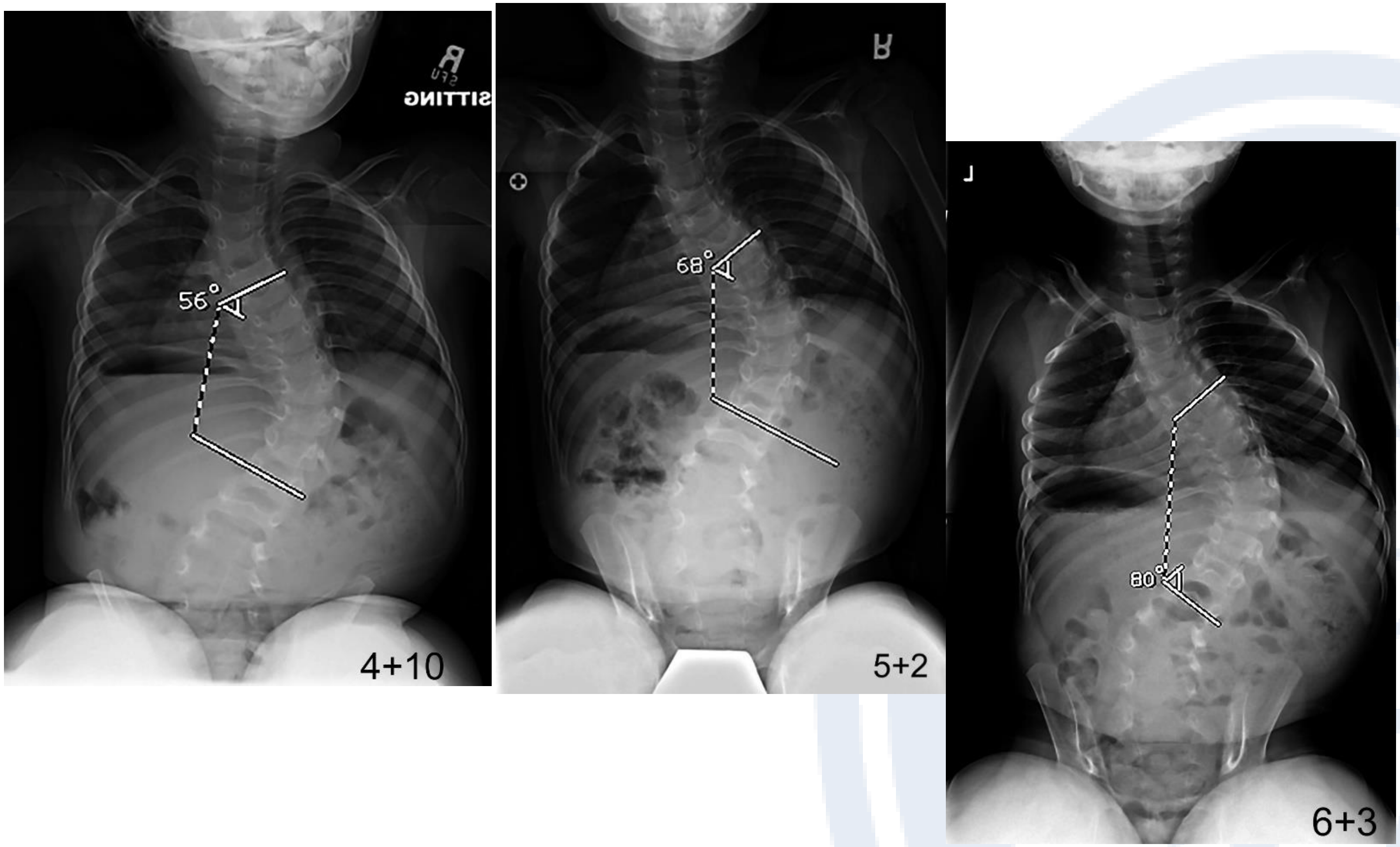
overactive bladder contract.

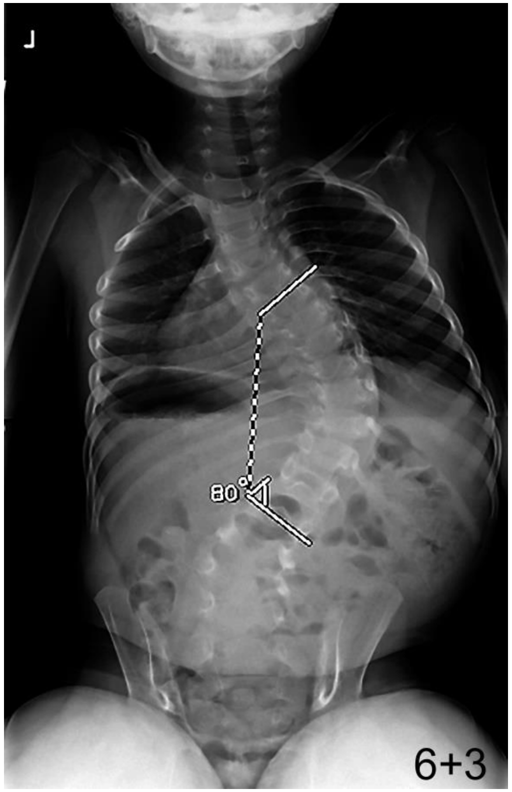
6+5- untethered

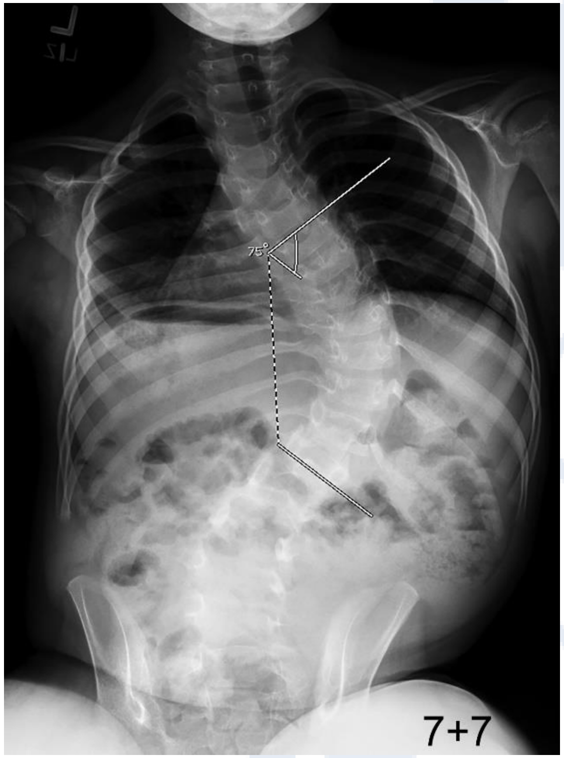
dyssynergy

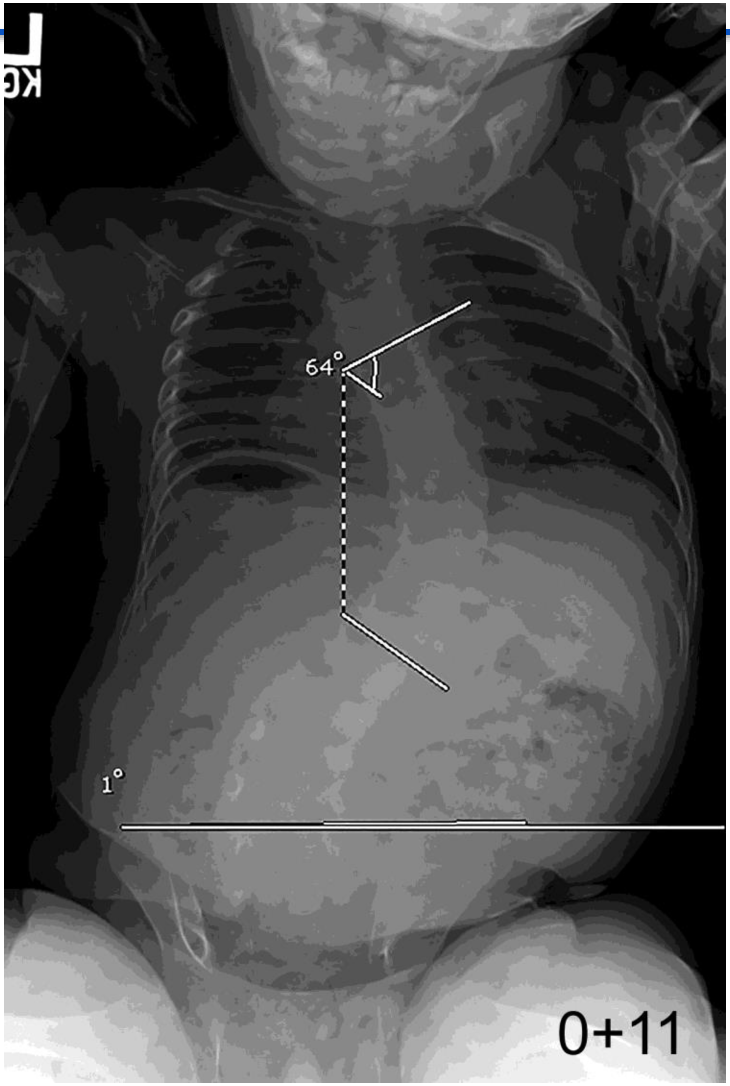
foot deformity

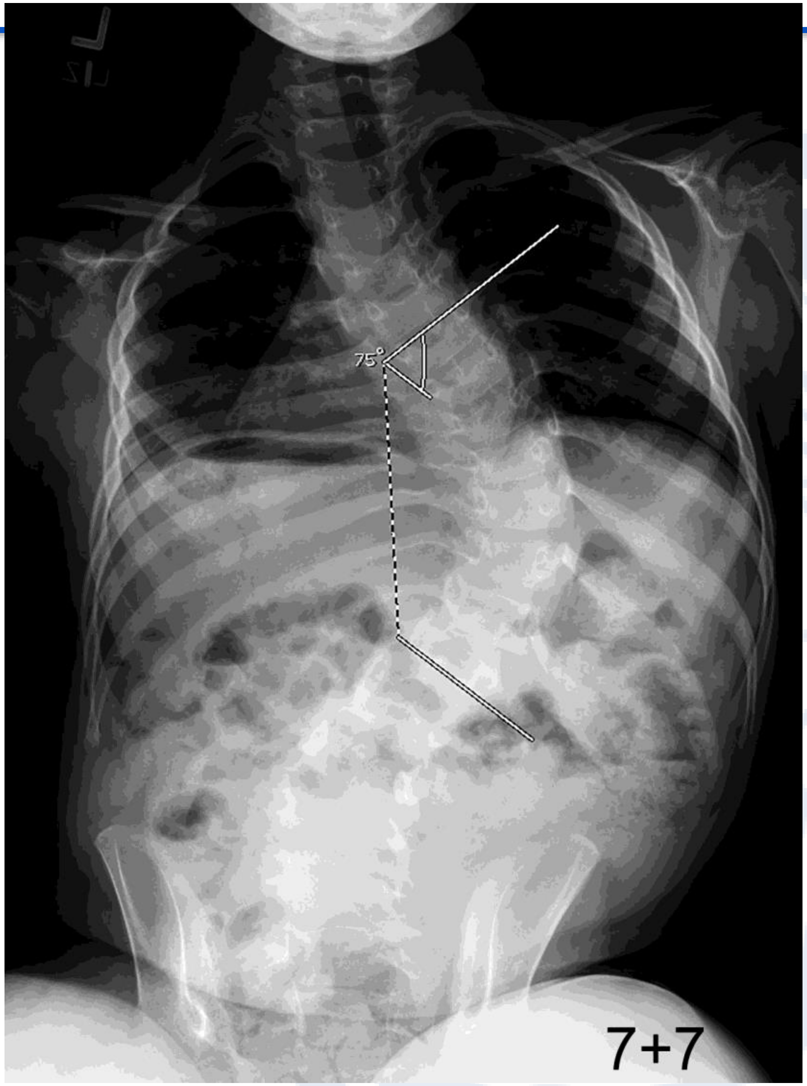



56°
4+10
68°
5+2
80°
6+3


80°
6+3


75°
7+7





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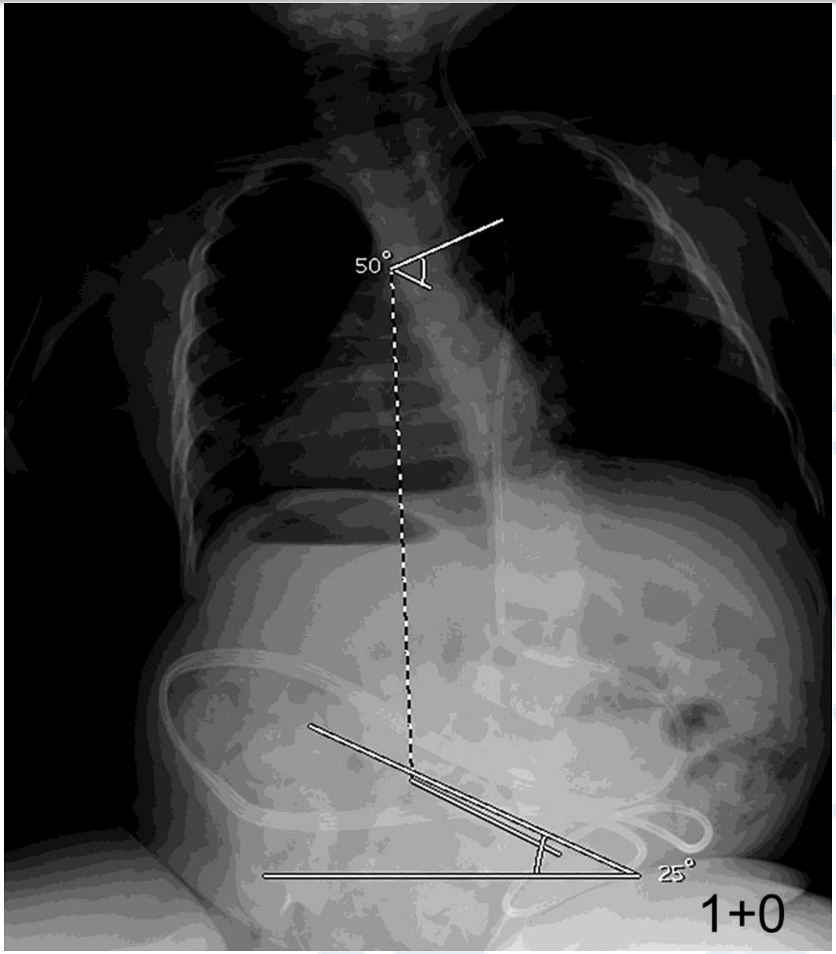
L.S.

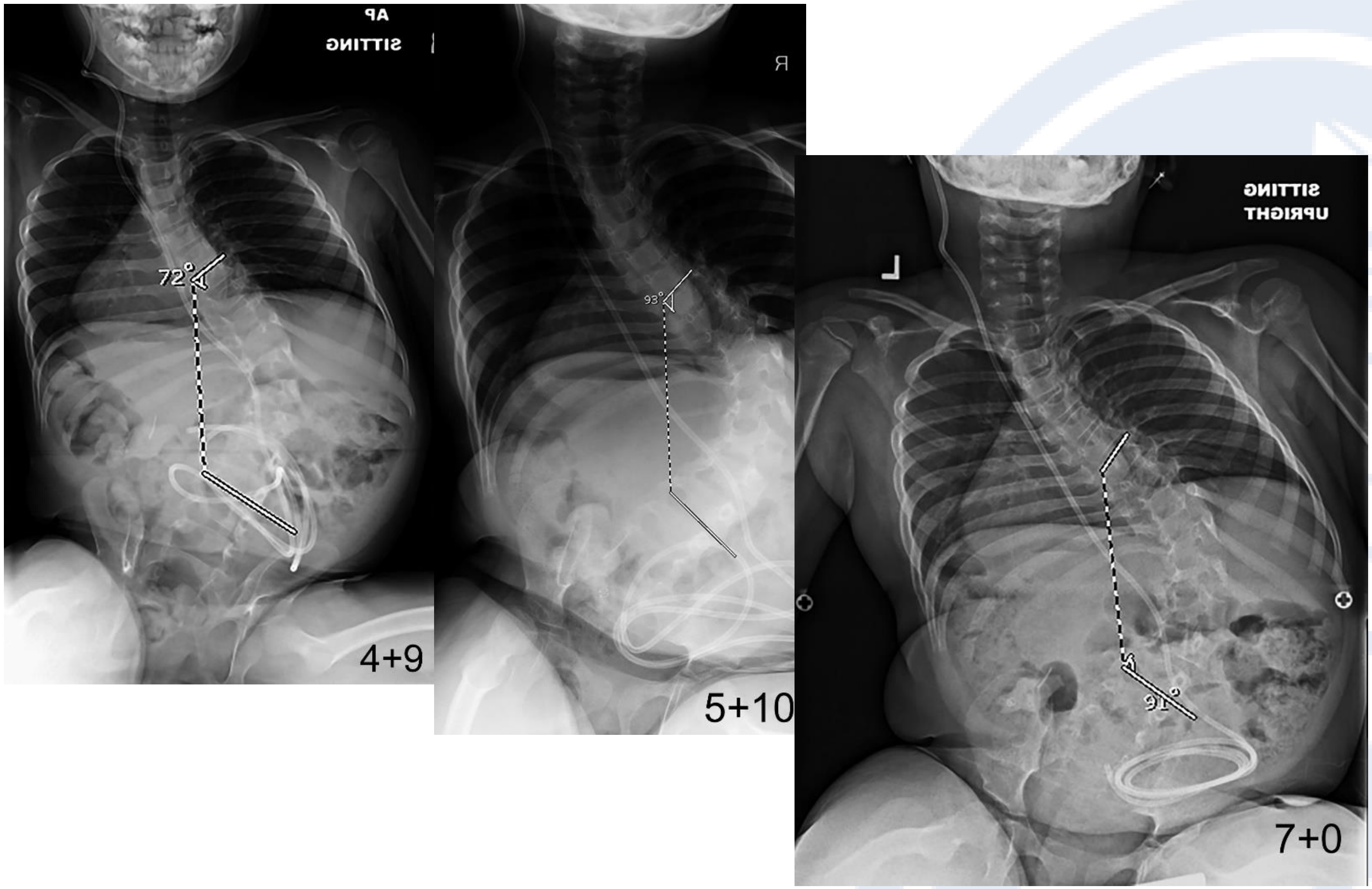
1+1y- untethered for
scoliosis
UDS- fibrillations

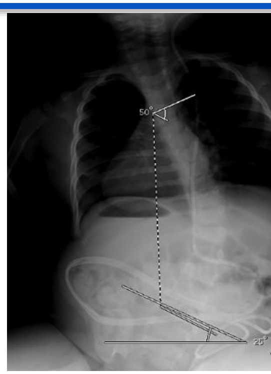
1+8y- cerebellar infarction-2° shunt failure- vent dependent

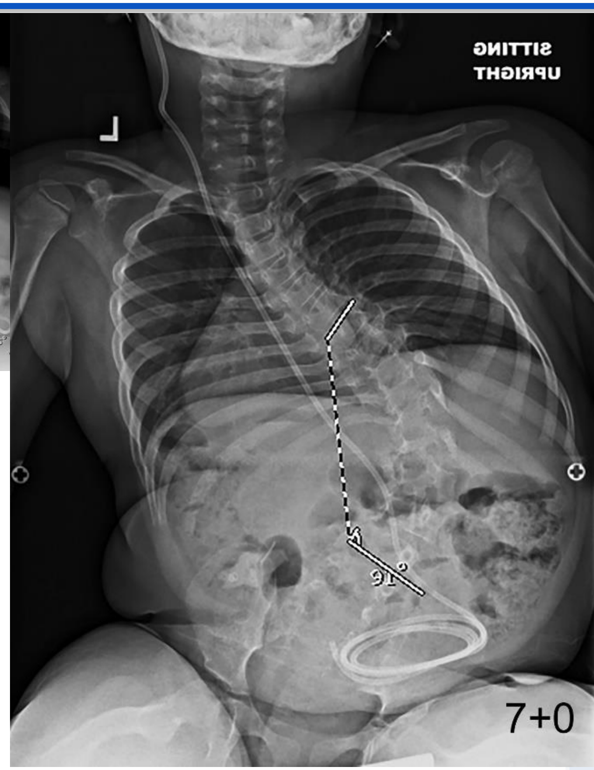
Multiple shunt revisions

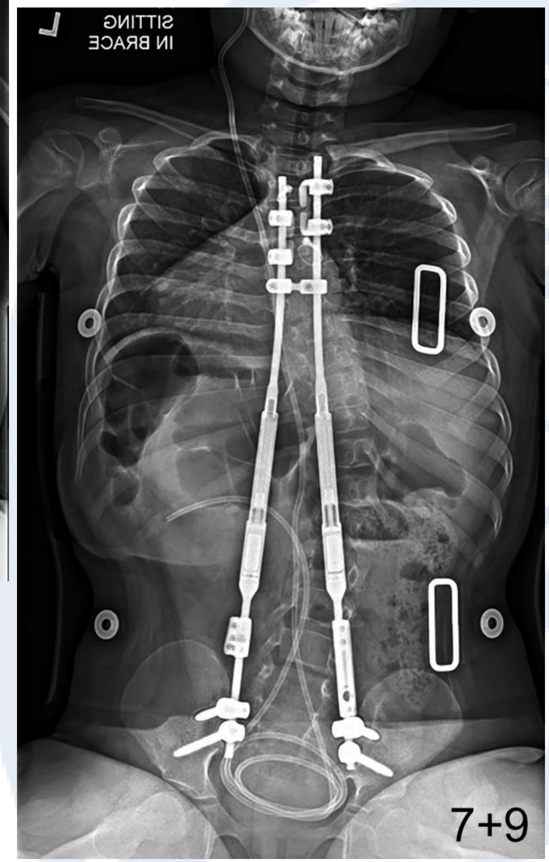
6+1y- untethered for
UDS- dyssynergy
scoliosis




72°
4+9
5+10
7+0




SITTING
UPRIGHT
91°
7+0


SITTING
IN BRACE
7+9

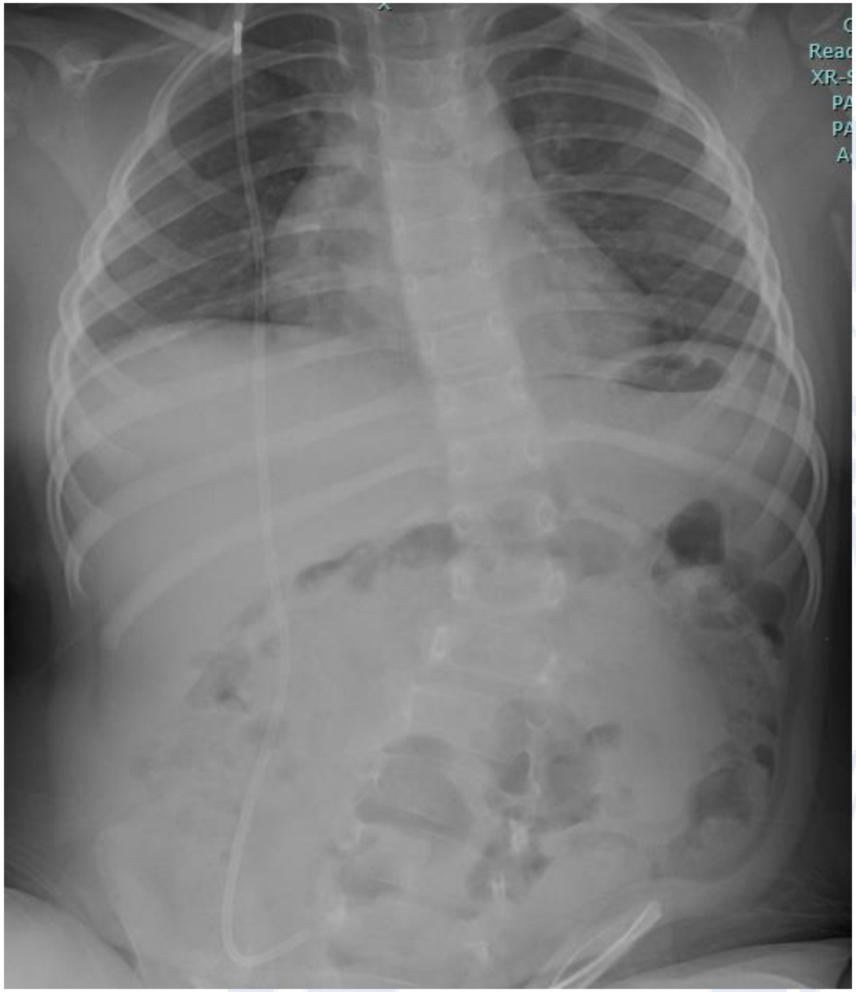
GL

4+8

32˚

Back Pain

L.E. Spaticity



GL

7+10

??0˚

GL

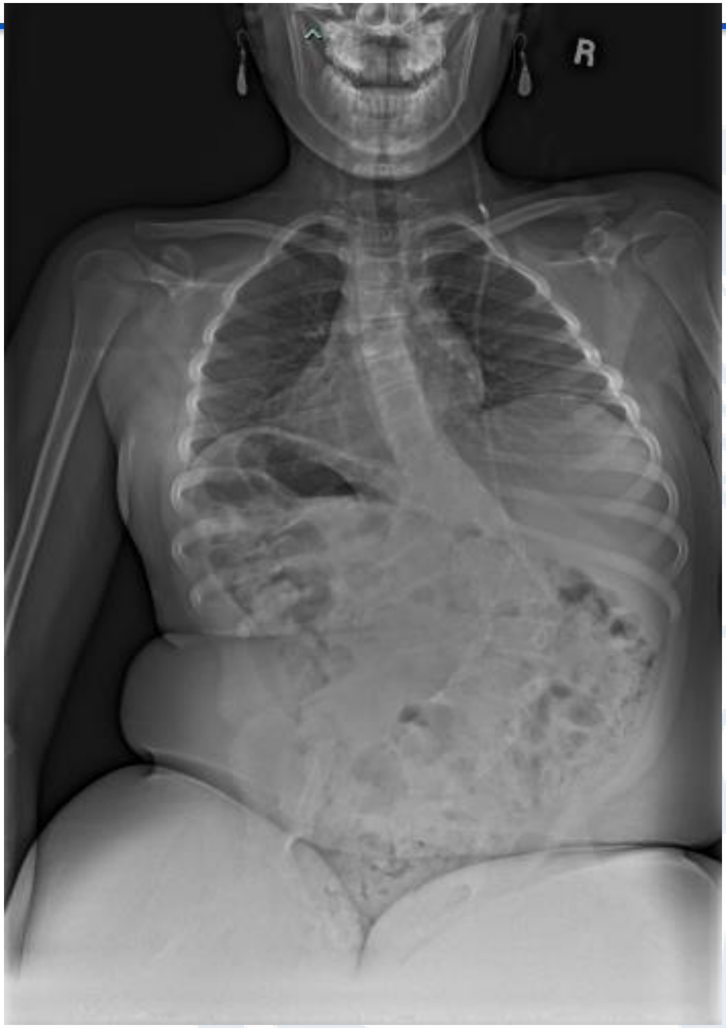
9+10

22˚

GL

11+5

72˚



Scoliosis Progression

	Year progressed	Cobb increase that year(°)	Total progression/total years(°)
40° &>			
TB	1	29	56/3
LS	5	19	26/5
AB	1	7	14/2
MP	2	7	36/4
VB	3	40	29/3
<40°°			
AM	3	39	35/3
GR	4	12	23/6

Surgical Stabilization

Age Detether (y)	Age Surgery (y)	Delay (y)	Cobb(°) vs (baseline)
11.2	14.8 (D)	3.6	110 (46)
1	7.2 (GR)	6.2	90 (50)
6.4	10.5 (D)	4.1	83 (72)
8.0	11.4 (AVT)	3.4	55 (41)
6.8	11.4 (D)	4.6	95 (60)
1.1	10.5 (AVT)	9.4	75 (64)
9.5	14.7 (D)	5.2	67 (23)
3.6	11.3 (D)	7.7	77 (34)
6	11.5	5.5	81 (49)

Conclusion

- **In selected children detethering surgery often- but not consistently- alters the natural history of scoliosis:**

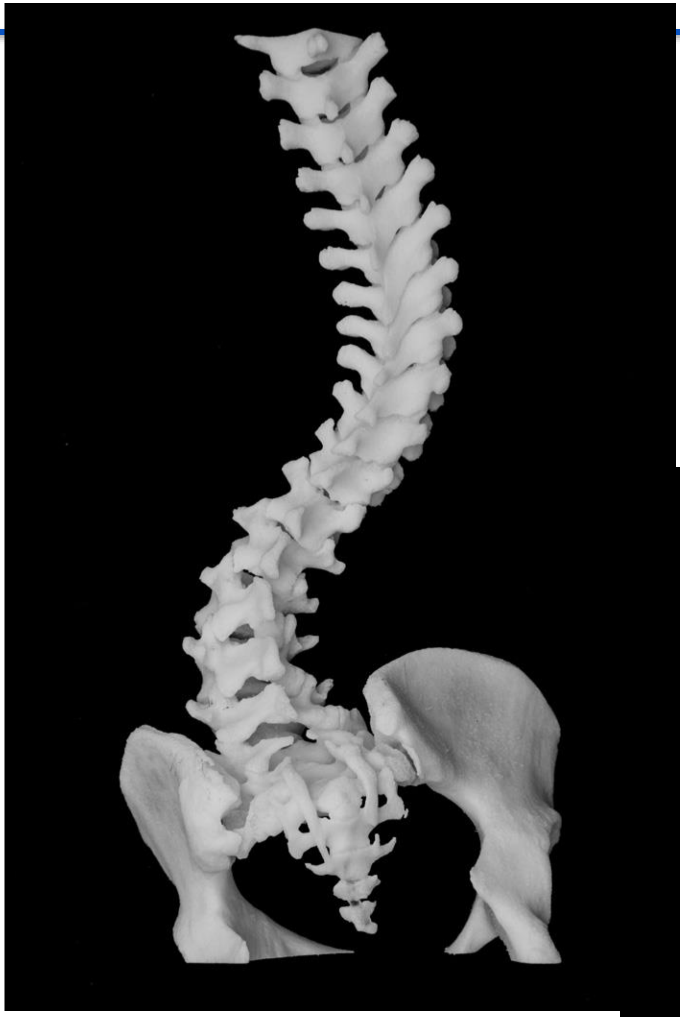
  stabilizes or improves the deformity

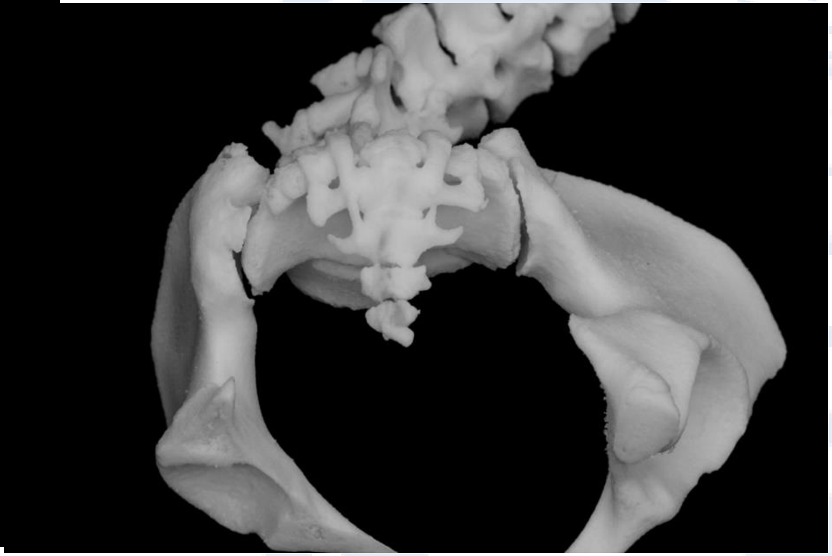
  slows progression of the deformity and delays a definitive surgery allowing further growth

Challenges

- Successful treatment requires compulsive (burdensome) attention – frequent visits throughout childhood
- ?Success of repeat detethering when progressive deformity is only indication
- ? Extend indication to include suspicious scoliosis (early onset, rapid progression)without other indicators of tethered cord

The use of computer generated 3-D models of the spine as an aid to planning and performing spinal deformity surgery in children with myelomeningocele





Challenges of Spinal Deformity Surgery in Children with Myelomeningocele

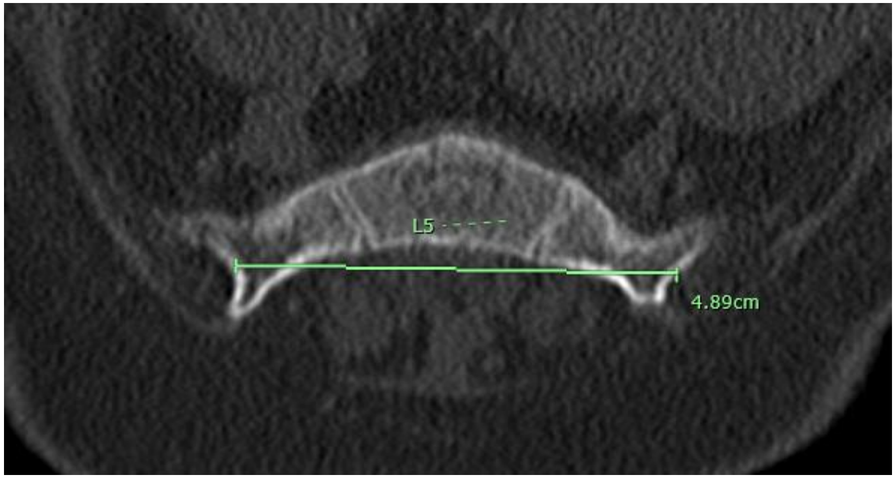
- **Absent posterior elements: addressed by modern segmental fixation- pedicle screws**
- **The dysplastic vertebral bodies have highly variable anatomy- anchor placement must be individualized – addressed by CT analysis**
- **The intersegmental relationships vary – it is difficult connect the pedicle screws to one another without putting stress on the anchors and without using bulky instrumentation – <u>3-D models</u>**

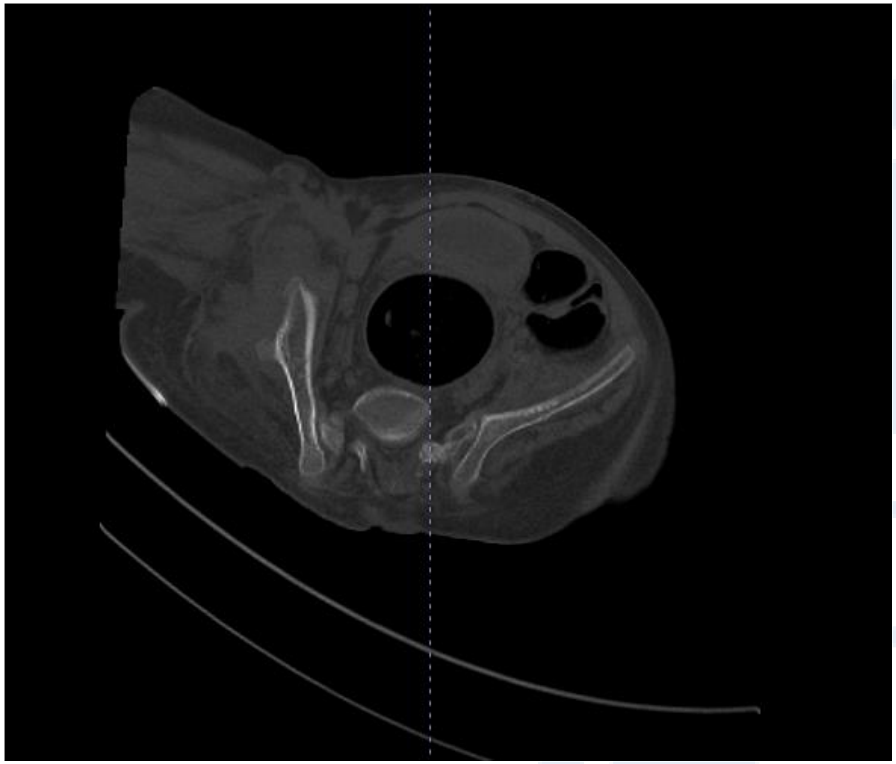
Preoperative Planning

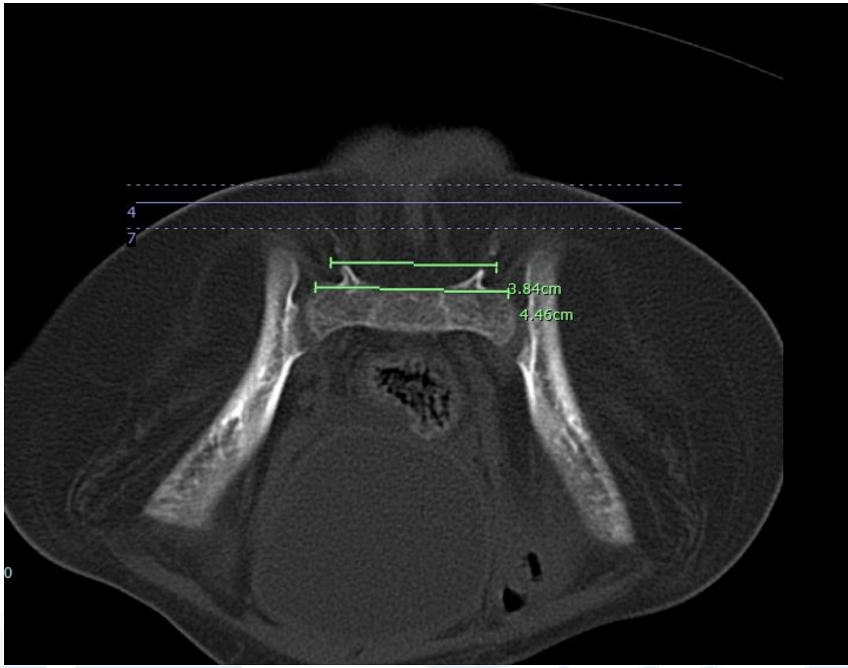
- **3-D segmental and global anatomy**
  - Design of osteotomies
  - Entry point and orientation of anchors for:
    - **Maximum segmental stability**
    - **<u>Harmonious inter-segmental alignment for ease of multi-anchor capture</u>**
    - **Ideal force vector**
    - **Low-profile**
- **Construct customization- rod contouring/pedicle screw sizing**

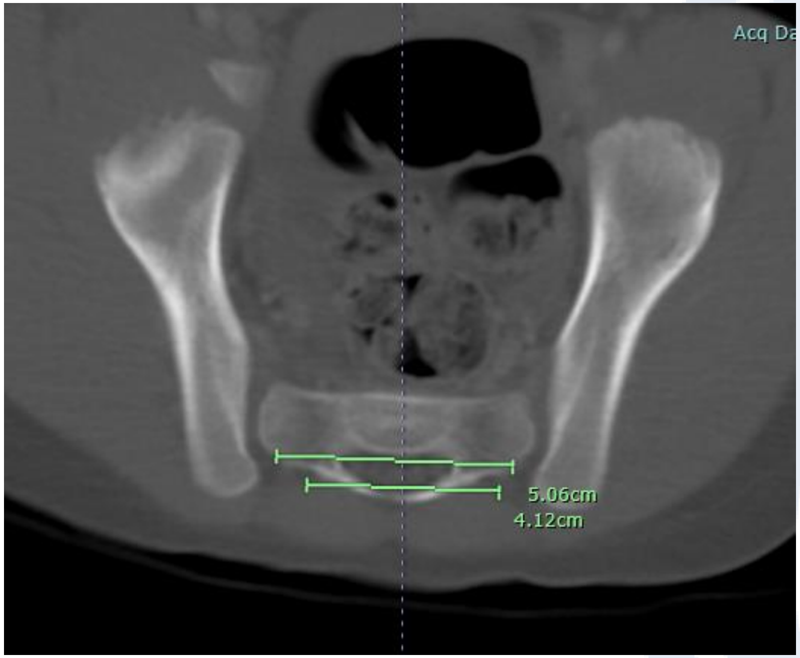
Intra-operative

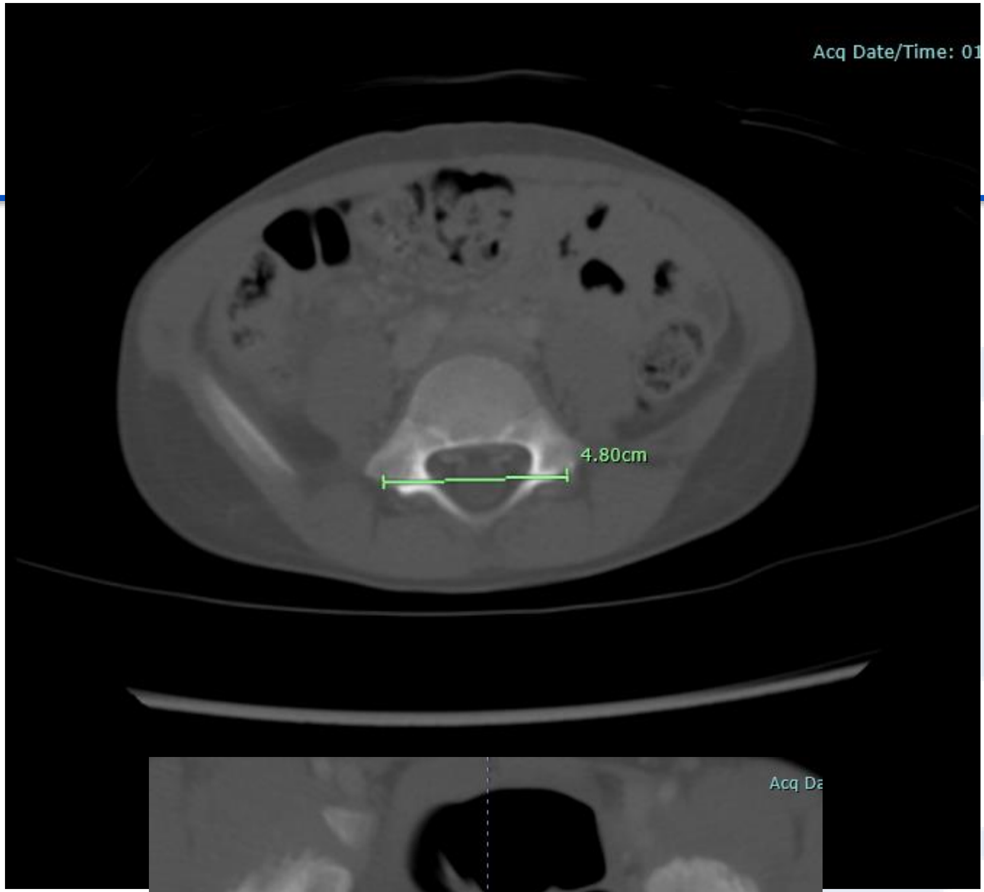
- **Reference to determine anchor entry points and orientation**
  - Avoid fluoroscopy- radiation exposure, inaccurate
  - Additional dissection- additional blood loss, possible dural injury


L5
4.89cm




4
7
3.84cm
4.46cm


Acq Da
5.06cm
4.12cm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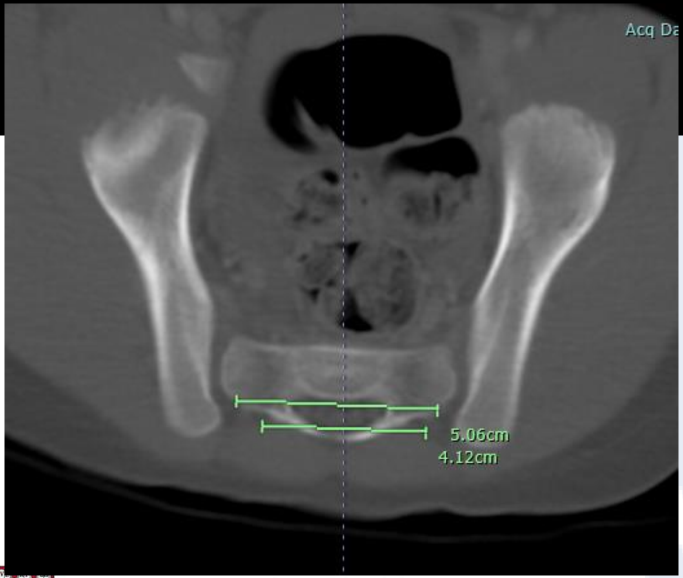
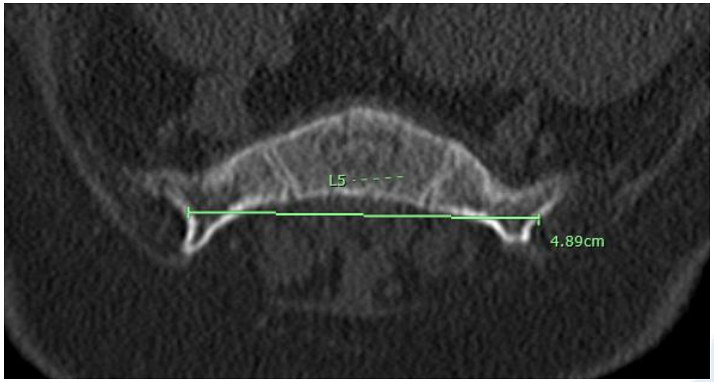
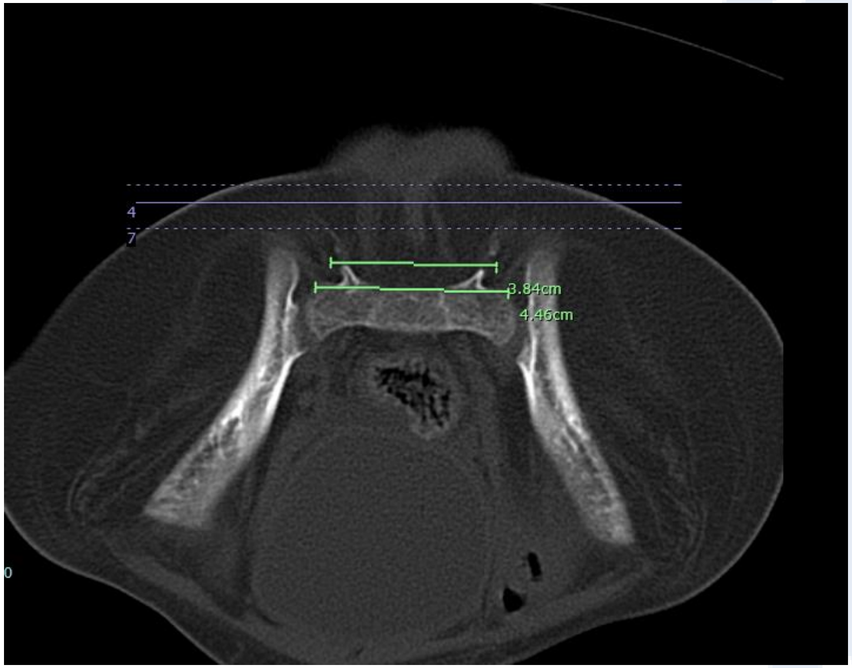
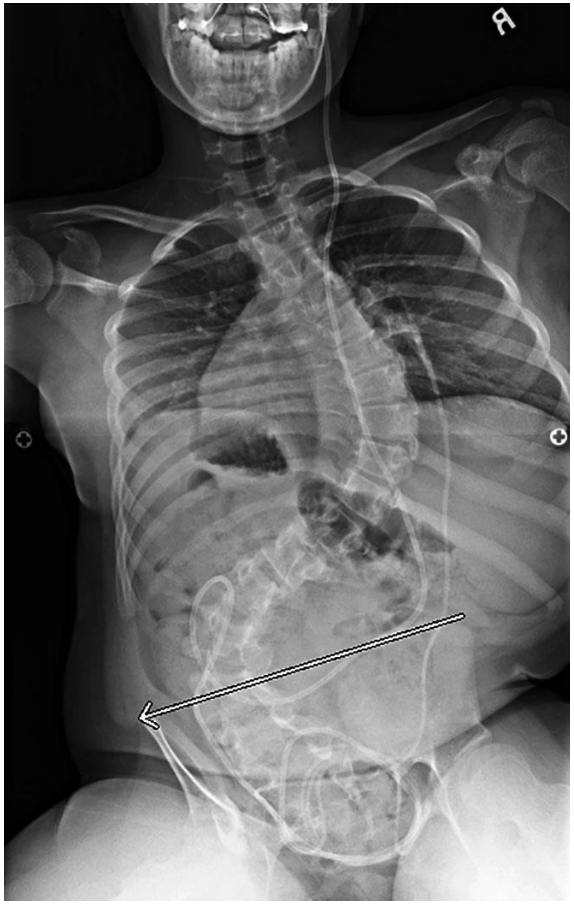
4.80cm


Acq Da
5.06cm
4.12cm


L5
4.89cm


4
7
3.84cm
4.46cm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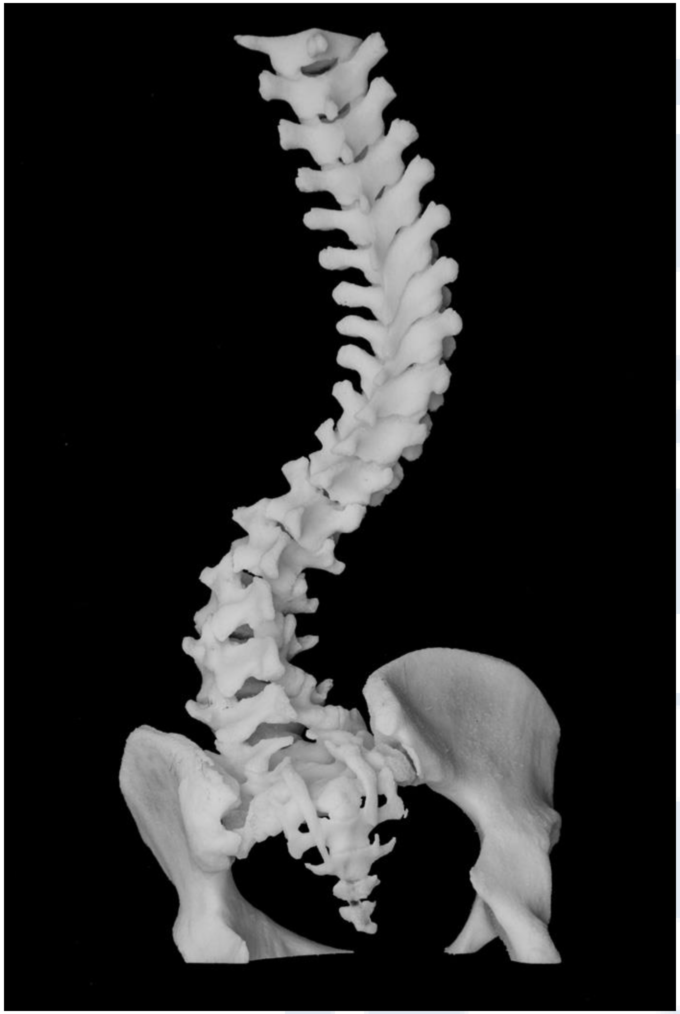

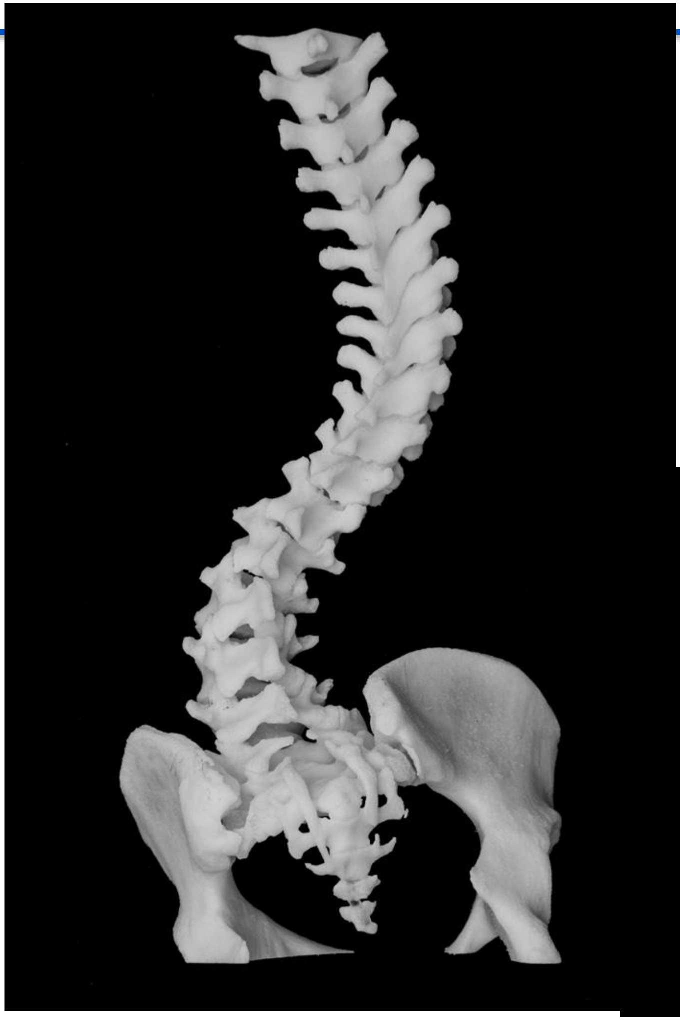

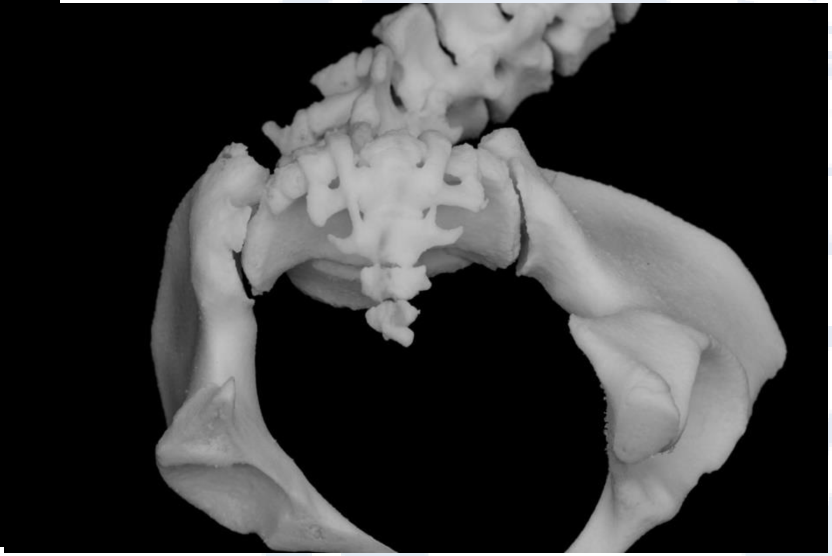

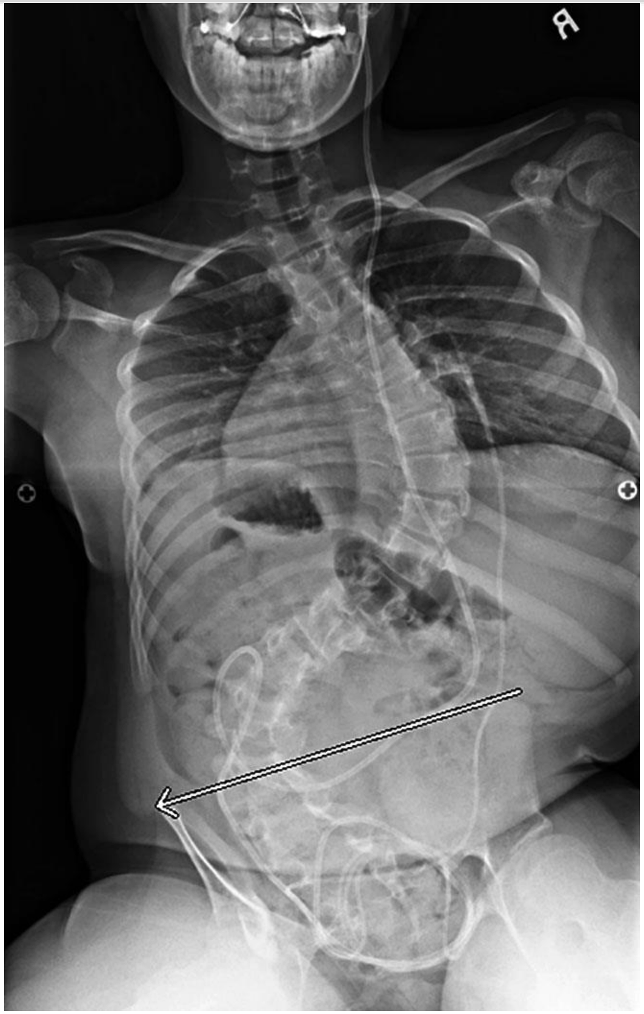

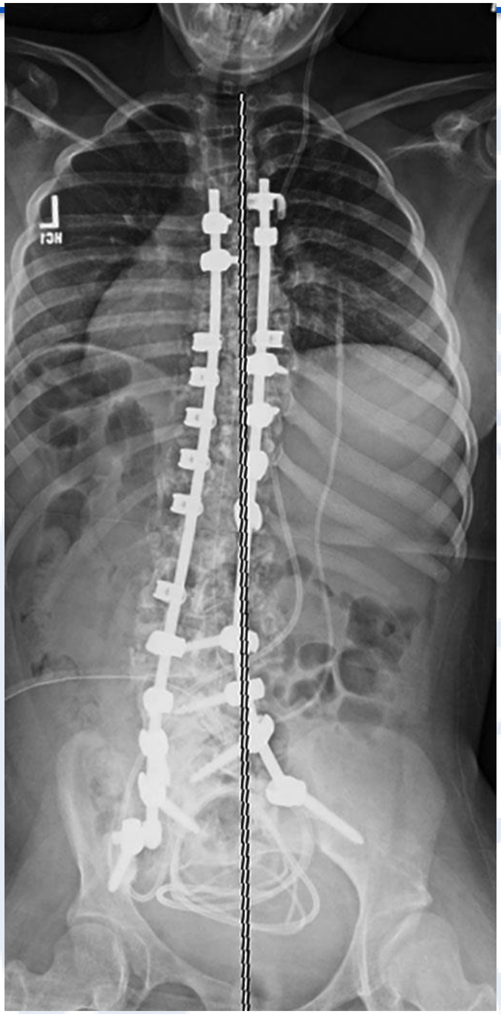

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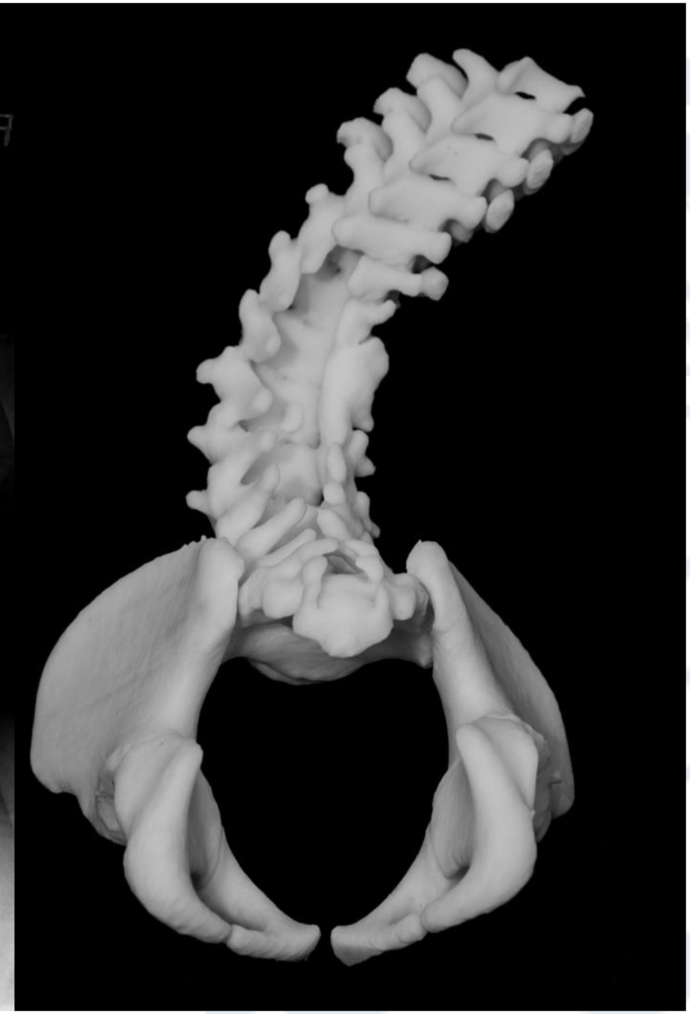

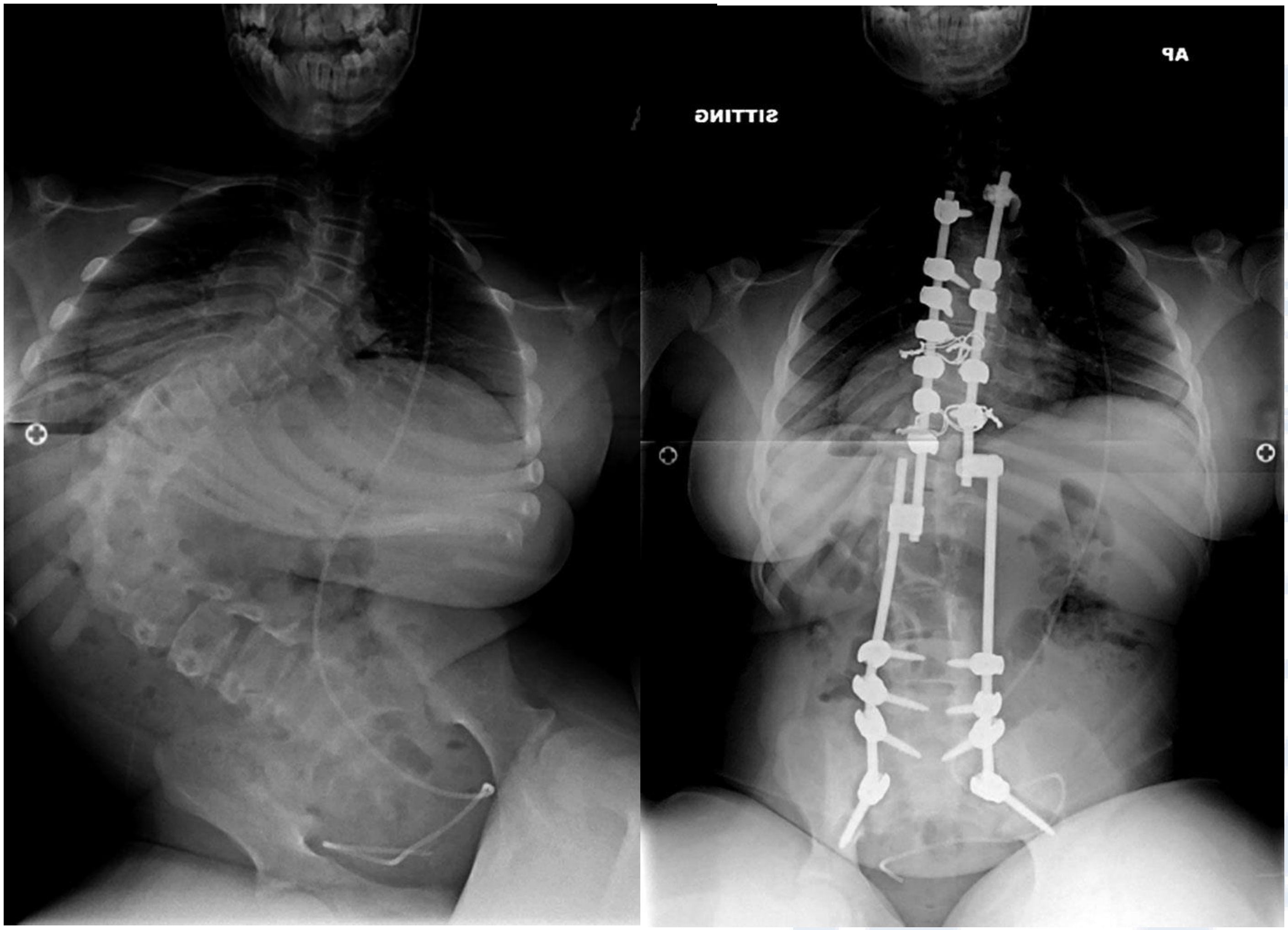

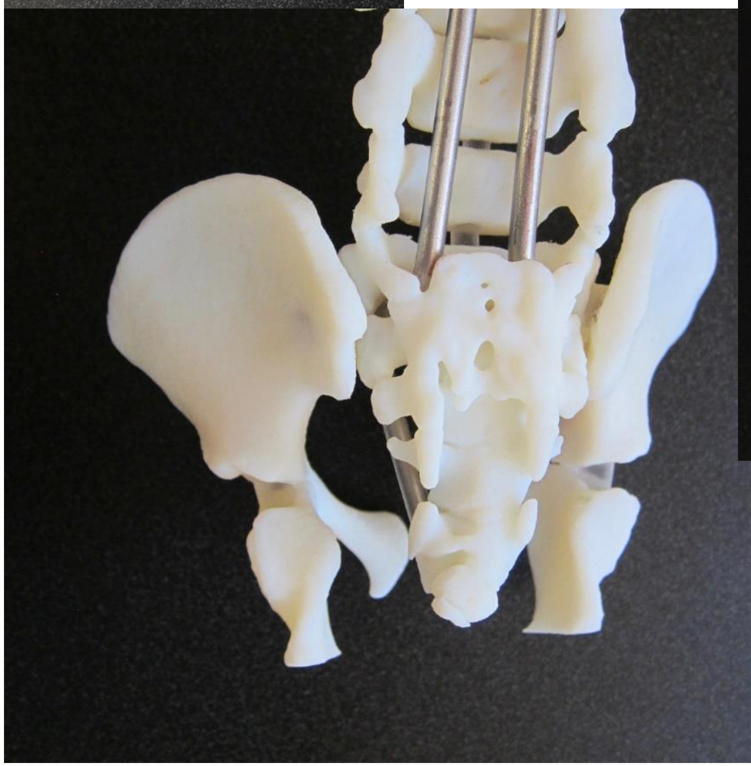

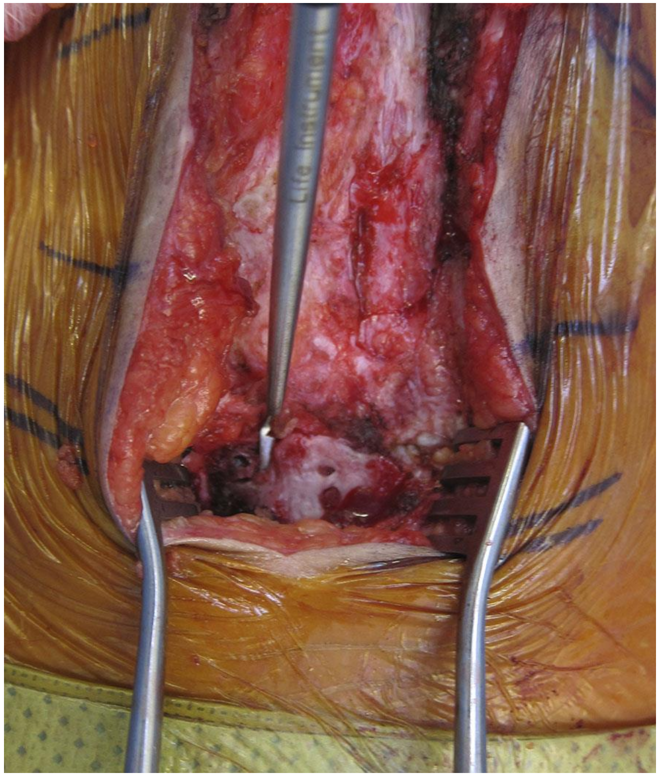


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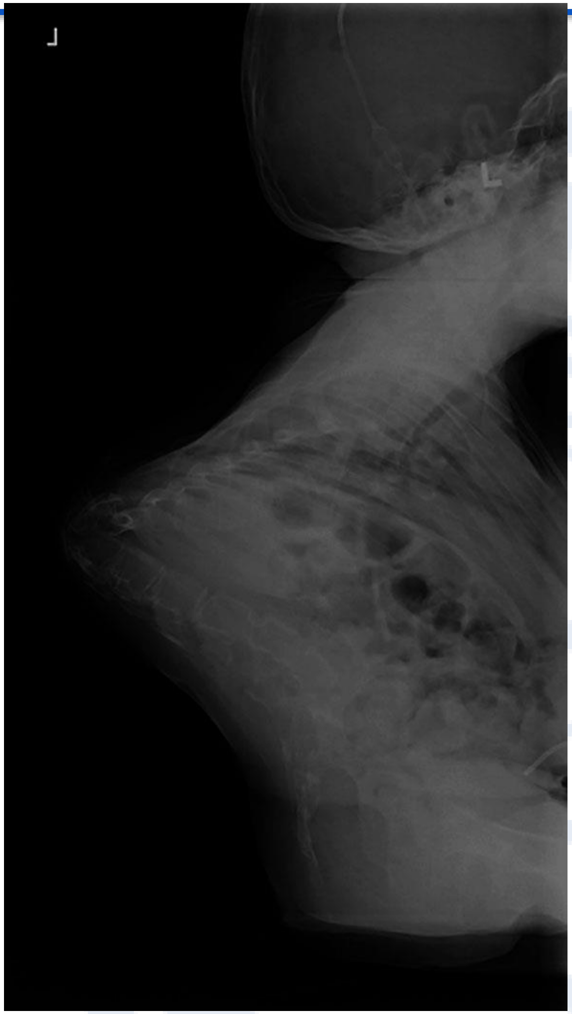

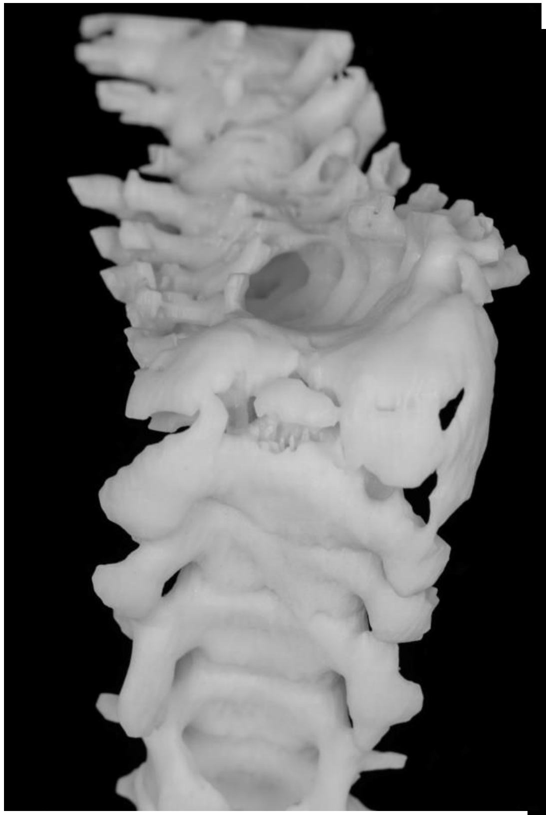

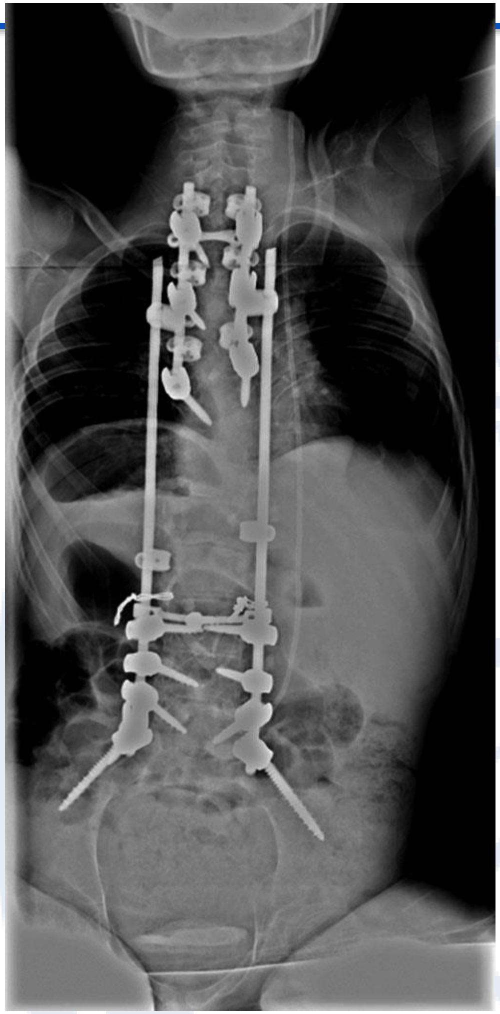

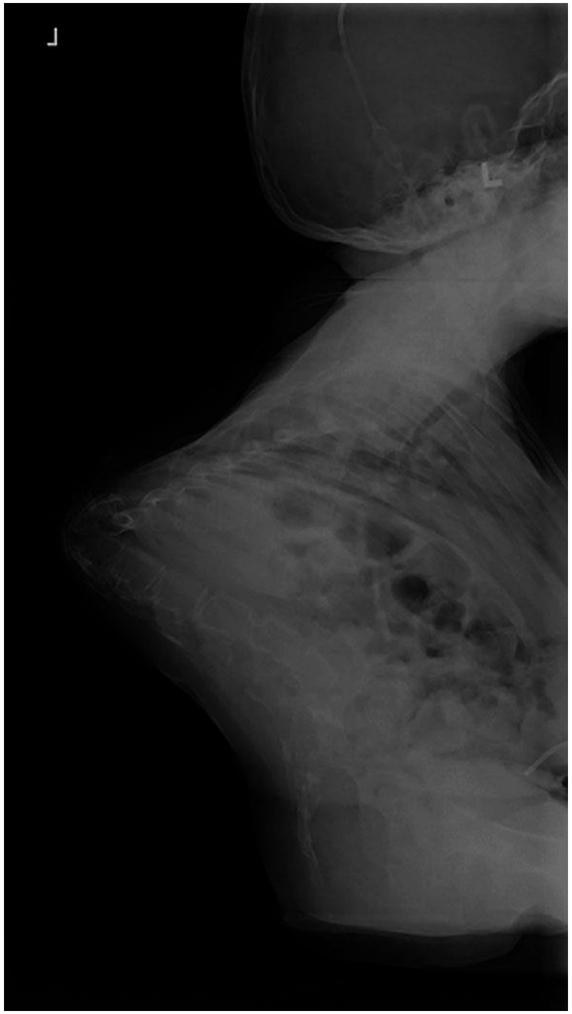


AP
SITTING
R

Efficiency

Group	Fluoroscopy Time	Blood Loss (% blood volume)
A	0.2 min.	24
B	0.42 min.	26

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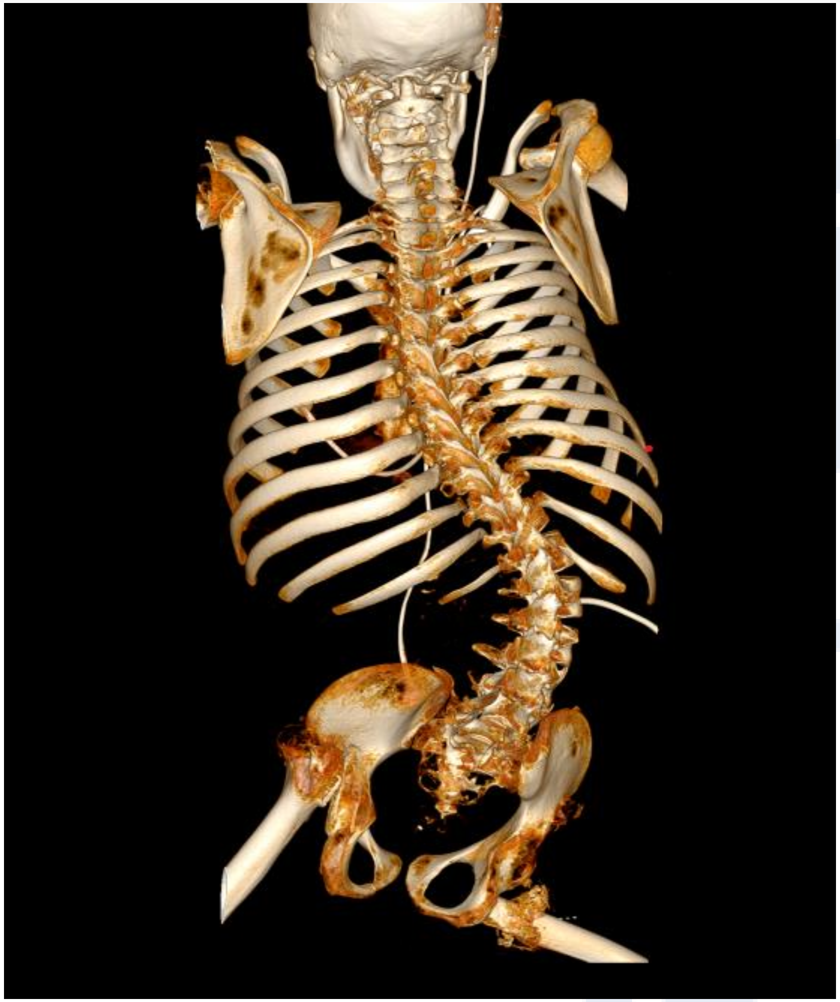
A (7 models) vs B (10 no models)

Group	Deformity	Pre-op Cobb (°)	Post-op Cobb(°)	% Correction
A	Scoliosis	123 (102-145)	21 (7-32)	83
	Kyphosis	112 (60-176)	13 (-2-37)	88
B	Scoliosis	74 (53-98)	23 (14-34)	70
	Kyphosis	152 (107-178)	42 (14-90)	76

Conclusion

- **3-D models – They help**

Thank you



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