Case discussions

ICEOS 2009
Istanbul

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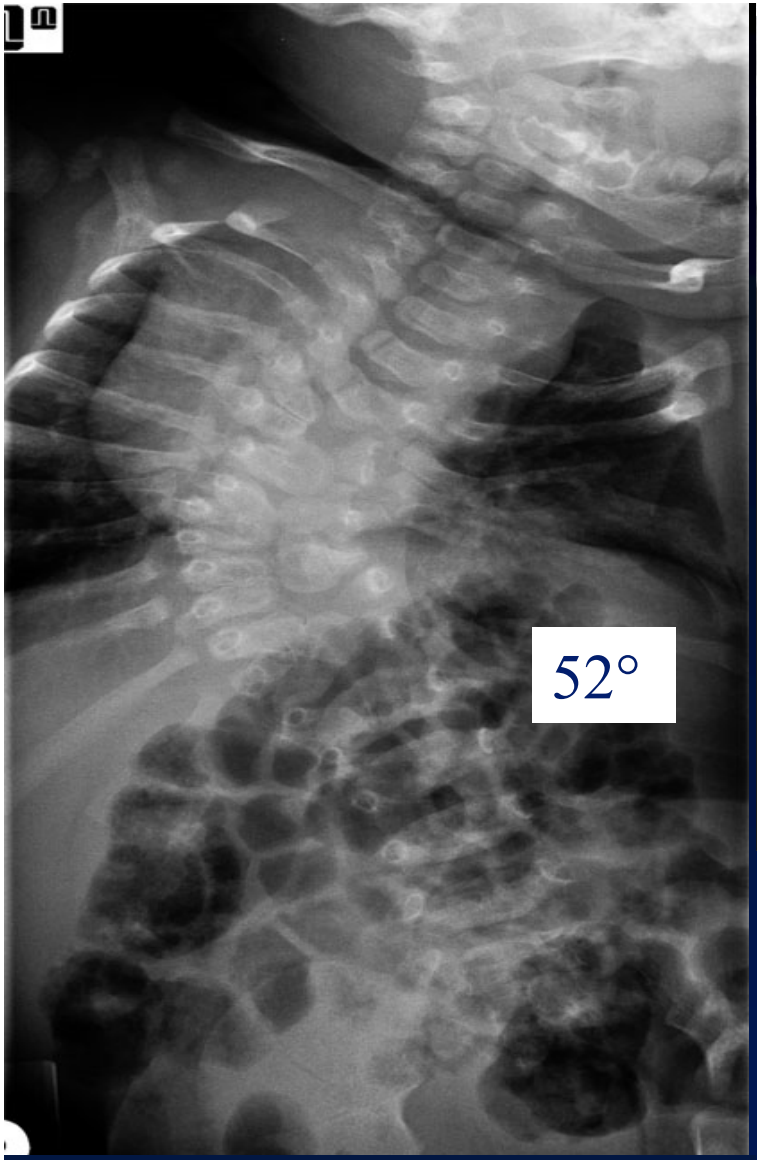


2 months old

December, 2005

- Twin boy
- Prenatal ultrasound noted polycystic kidney and congenital scoliosis
- Otherwise healthy at birth
- No respiratory support required

This cervico-thoracic spine film at age 4 mos most clearly shows the vertebral and rib anomalies

CT scan at 4 months of age

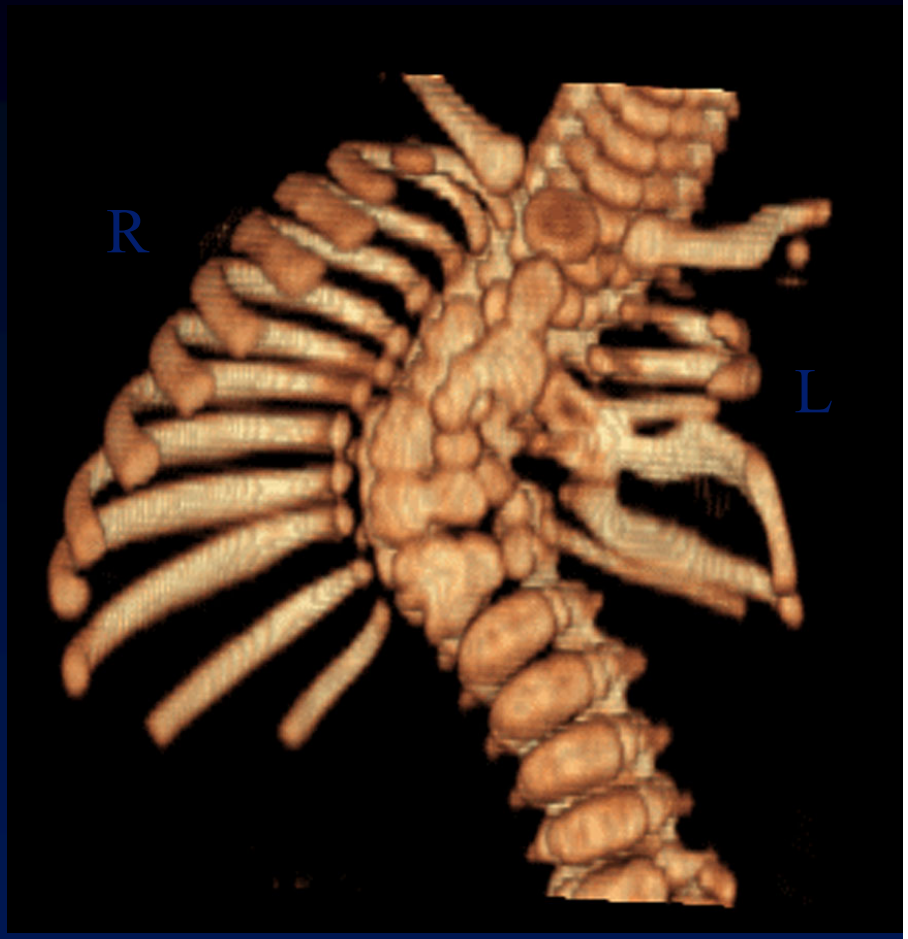
60° congenital scoliosis

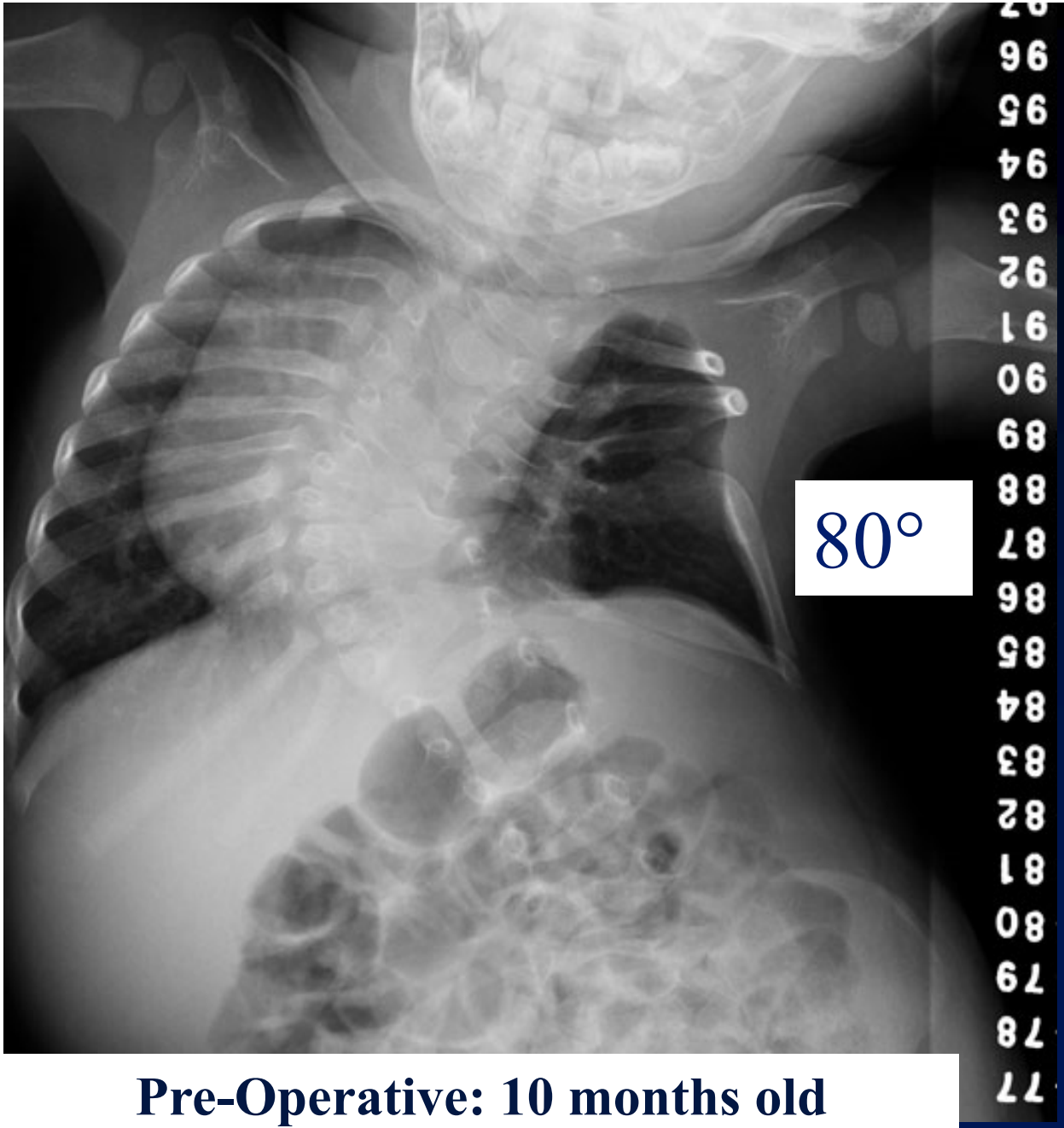
anomalies of multiple left-sided ribs and vertebra affecting the lower six thoracic vertebral bodies and multiple ribs

poor lung aeration on the left side

left lung volume 53 cc

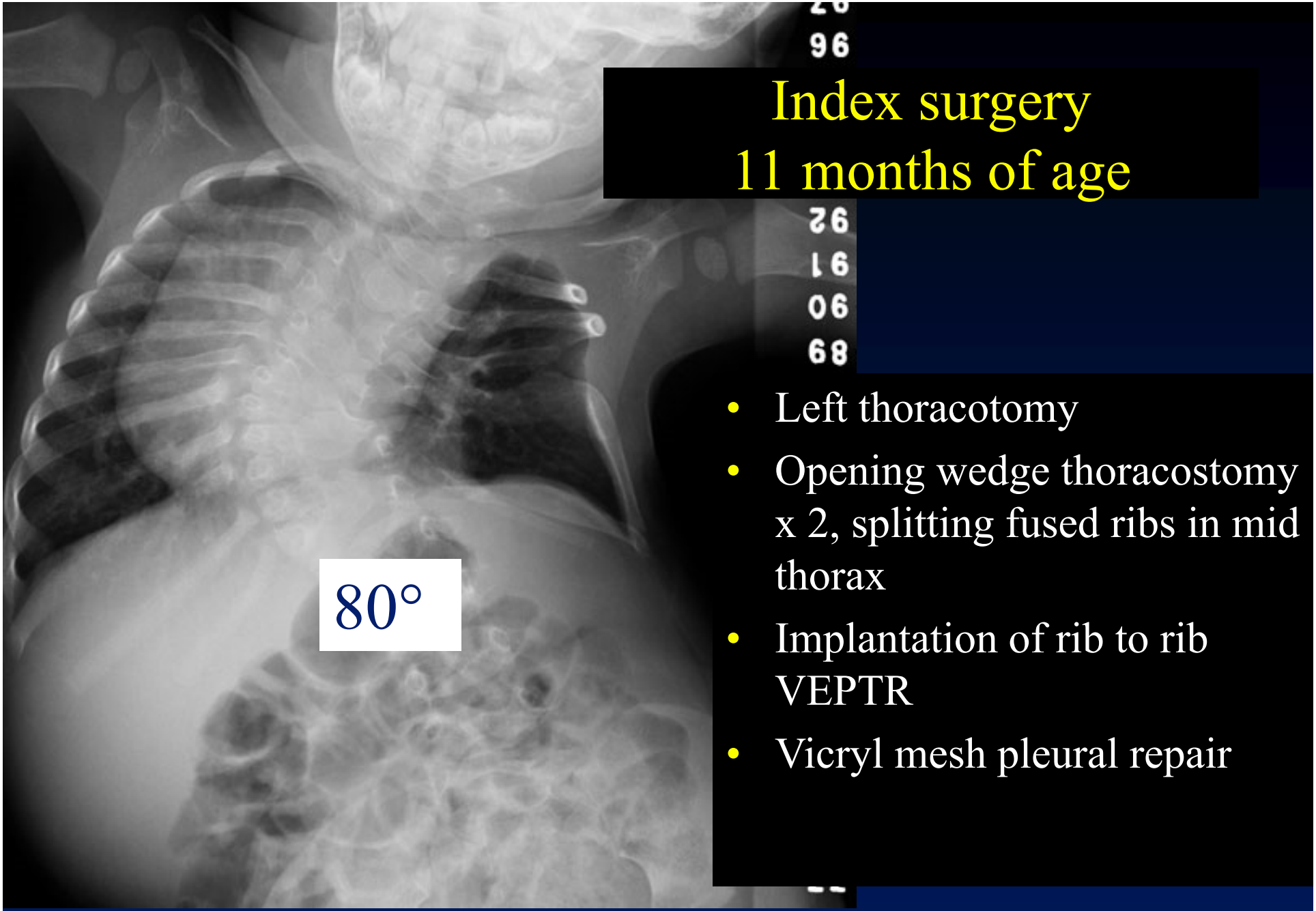
right lung volume 88 cc





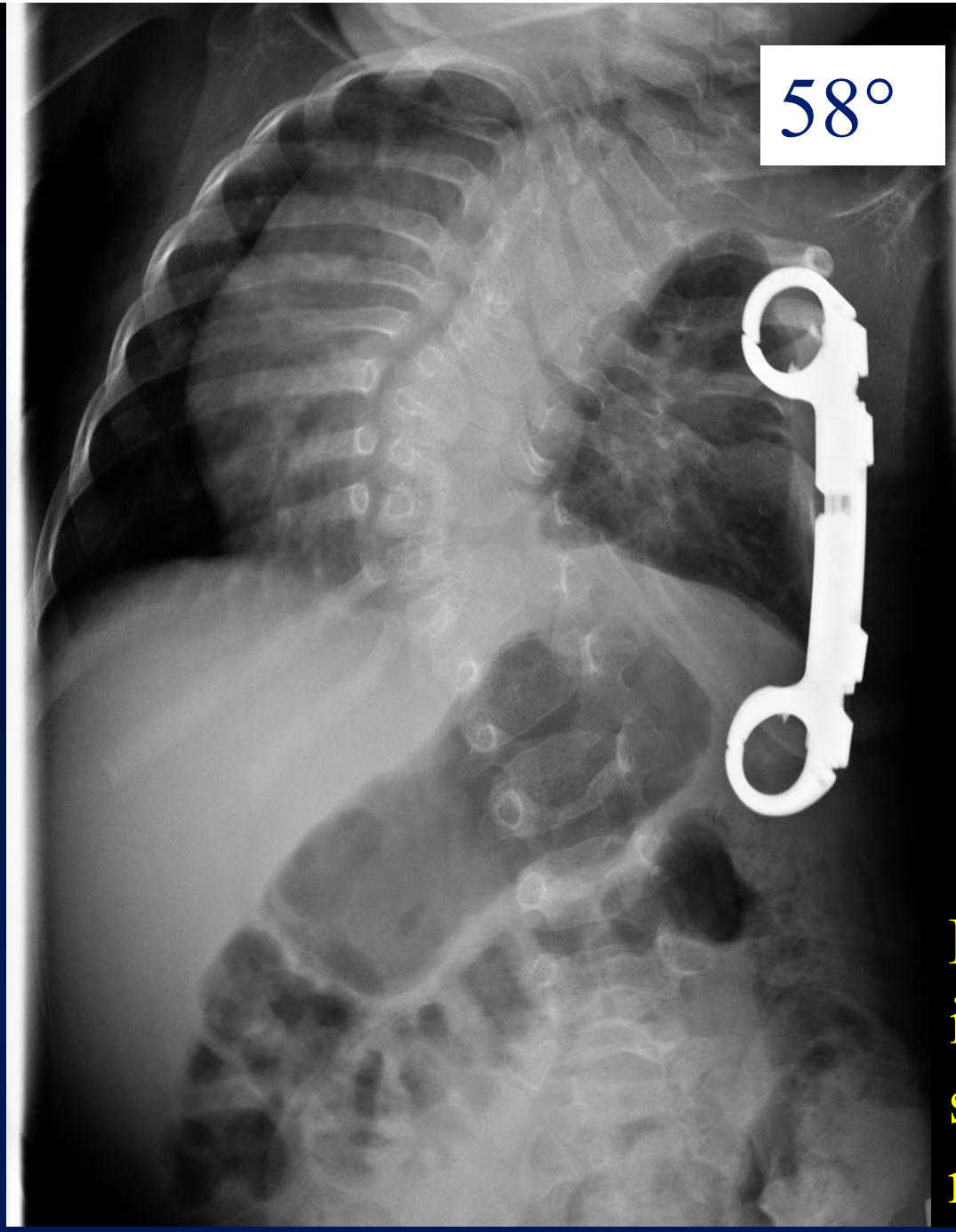
Pre-Operative: 10 months old

What would be your plan?



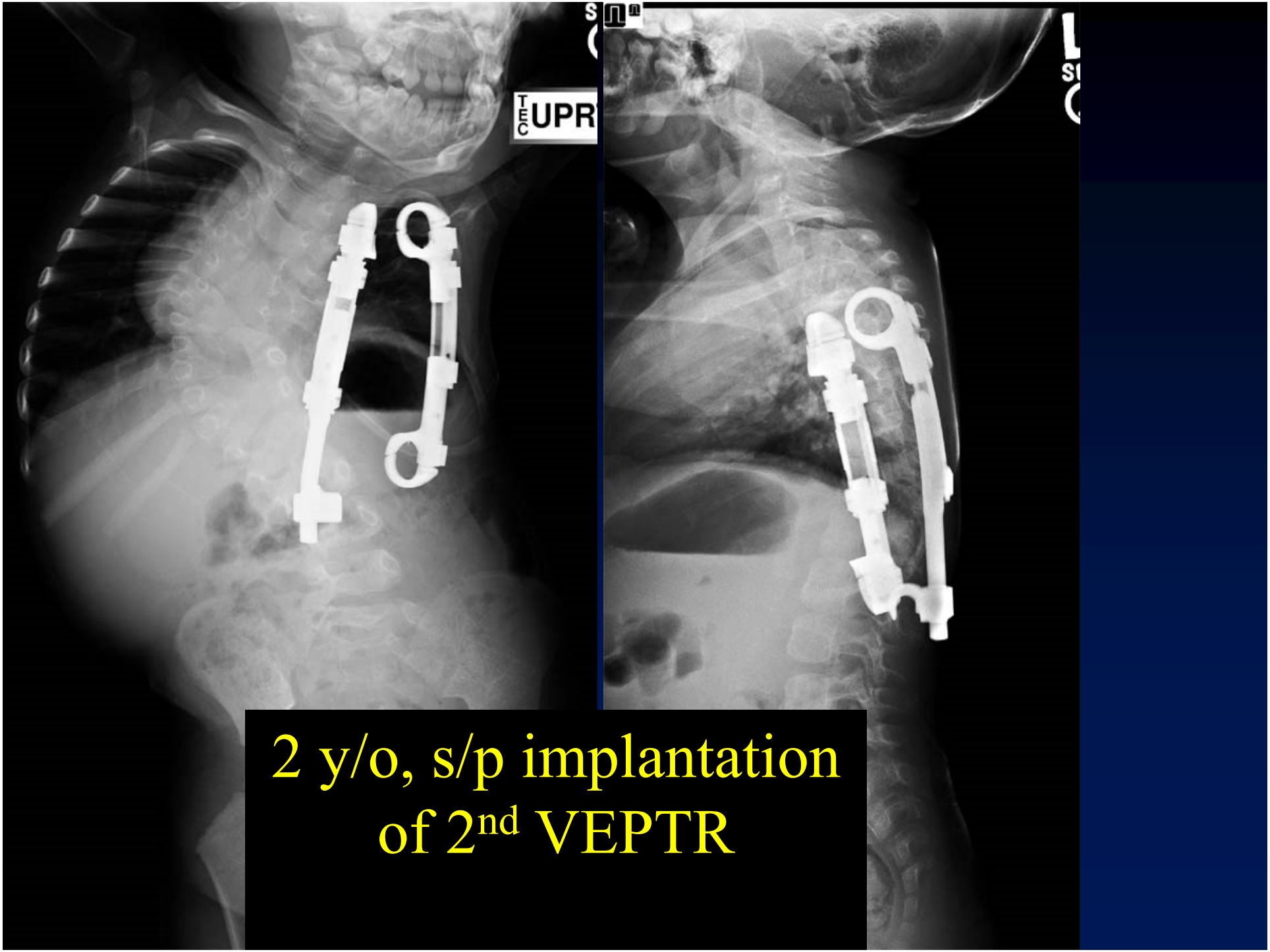
Index surgery
11 months of age

- Left thoracotomy
- Opening wedge thoracostomy x 2, splitting fused ribs in mid thorax
- Implantation of rib to rib VEPTR
- Vicryl mesh pleural repair

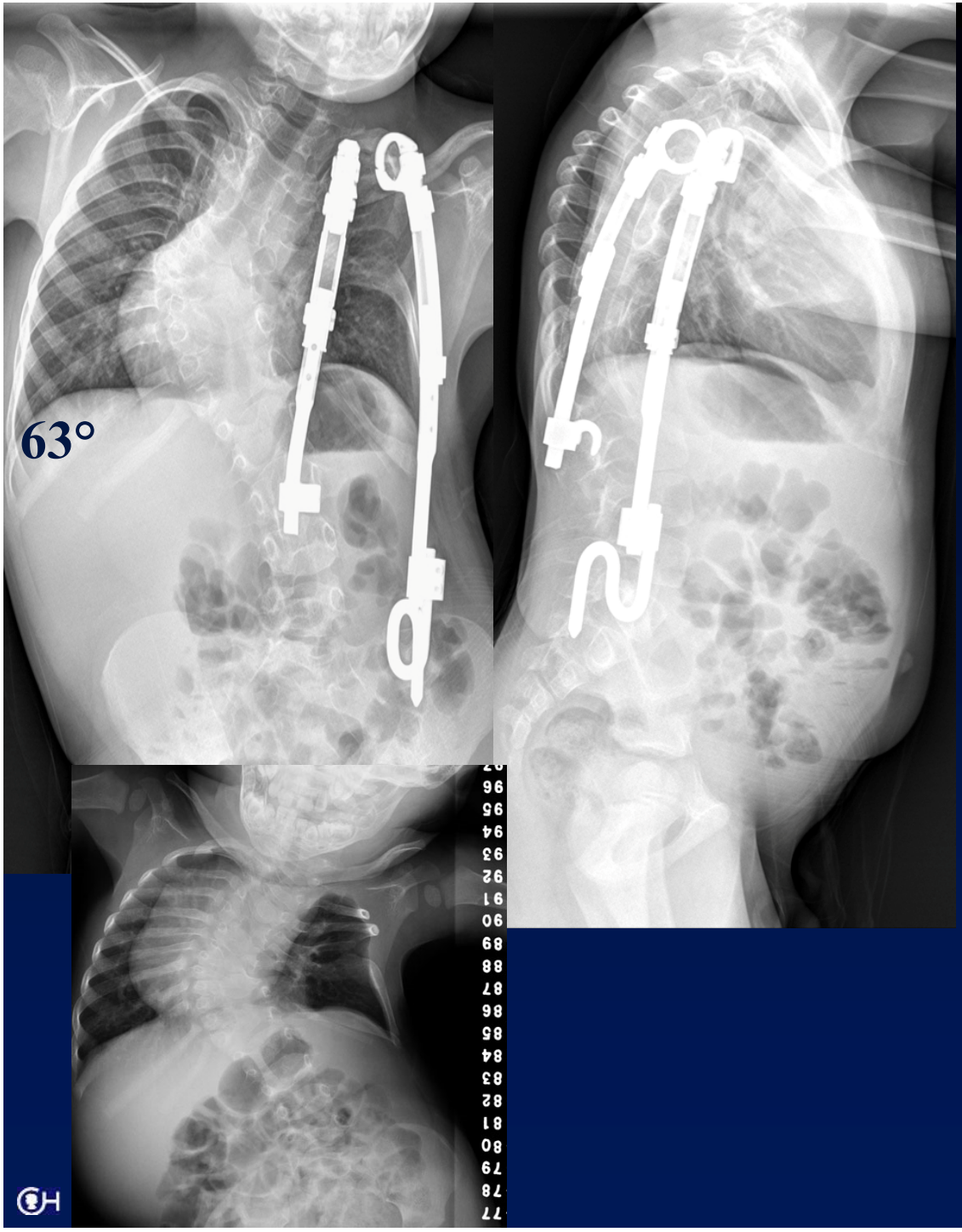


1 mo post op– 1 yr old

Plan: 6 months after index surgery, ad rib to spine device, and expand rib to rib device



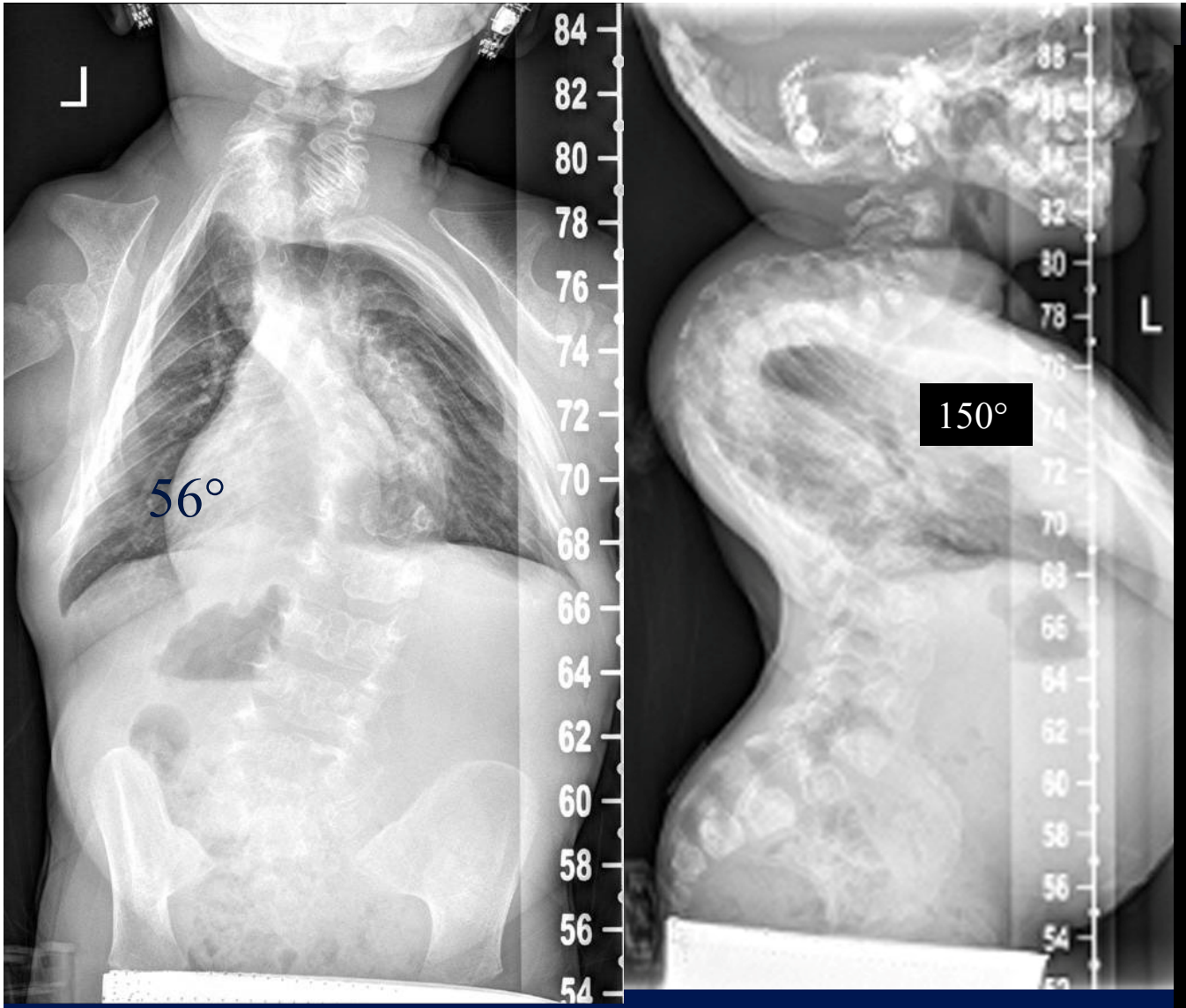
2 y/o, s/p implantation of 2nd VEPTR



5 y/o

4 yrs after index surgery

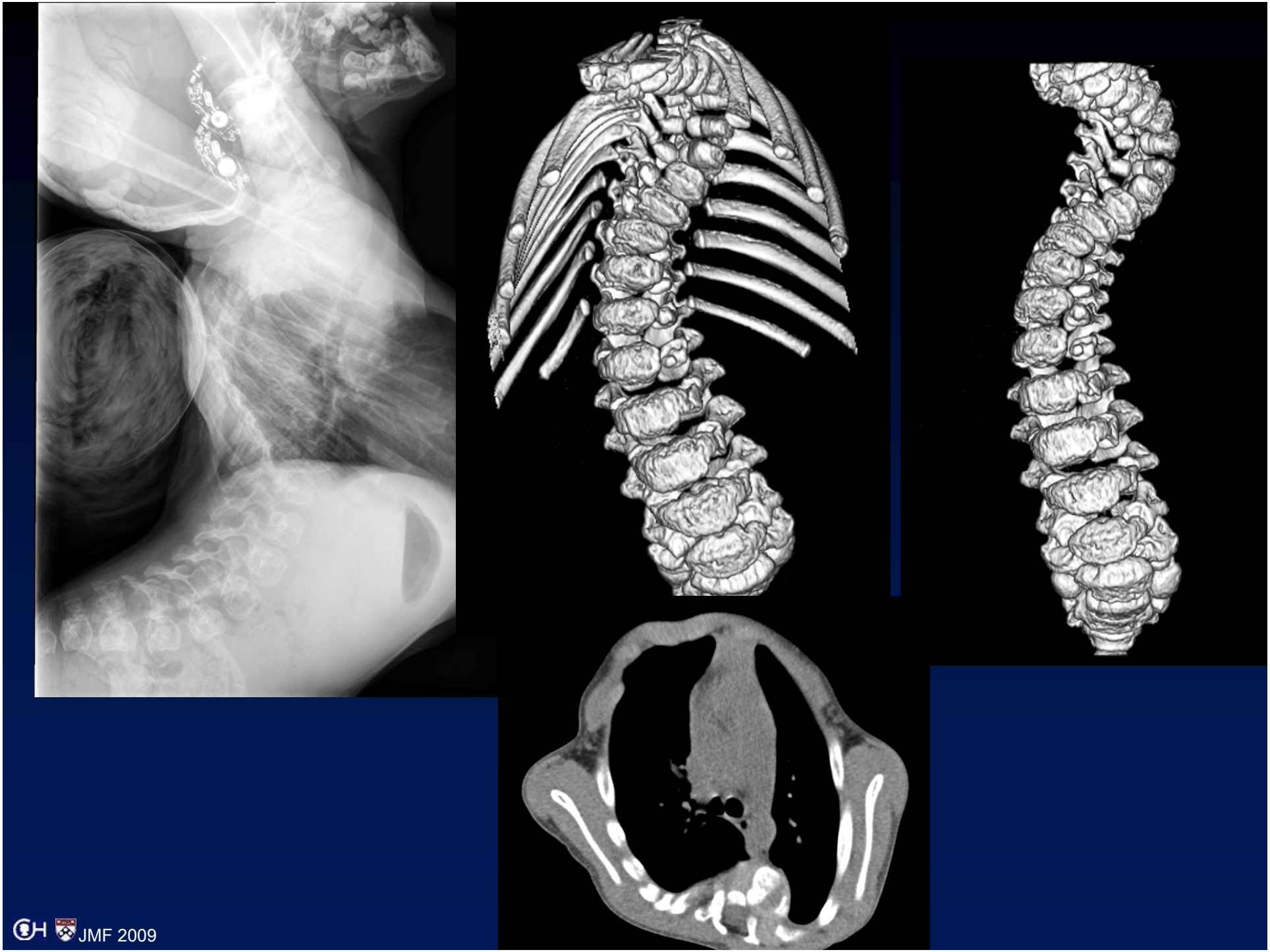
rib to rib has been exchanged for rib to pelvis



- 8 y.o. male from 700 miles away
- Cleidocranial Dysostosis Syndrome
- underwent brace treatment without effect on the deformity

Spirometry (BTPS)

- FVC 75%
- FEV1 57%

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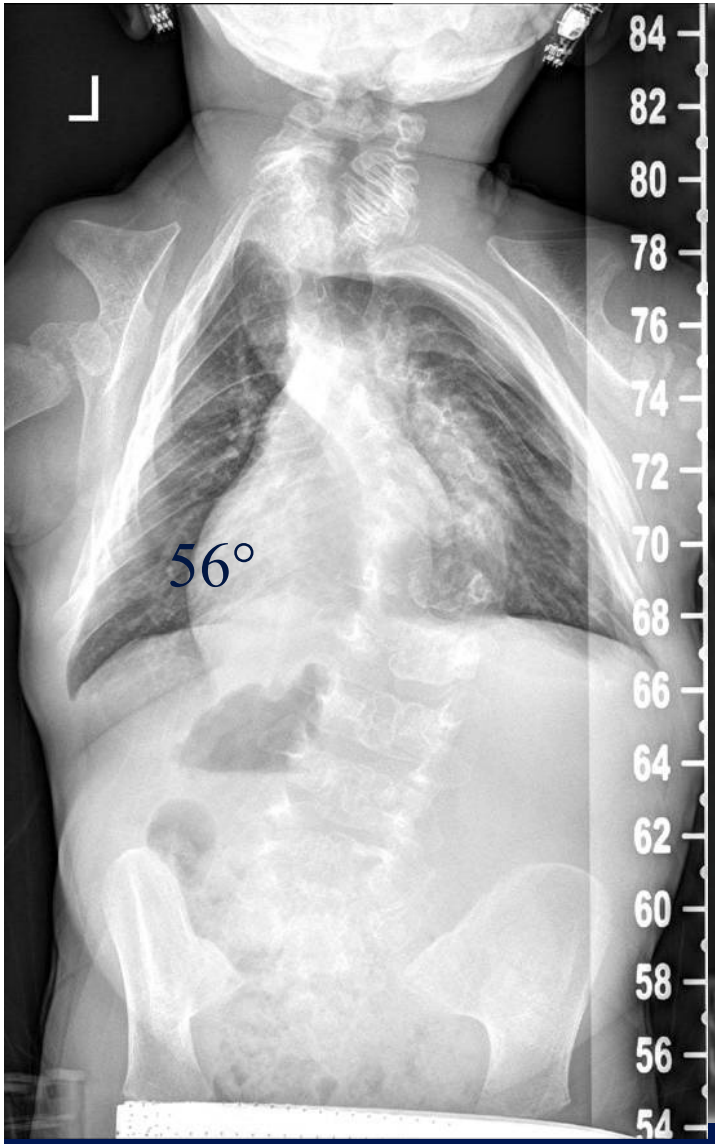
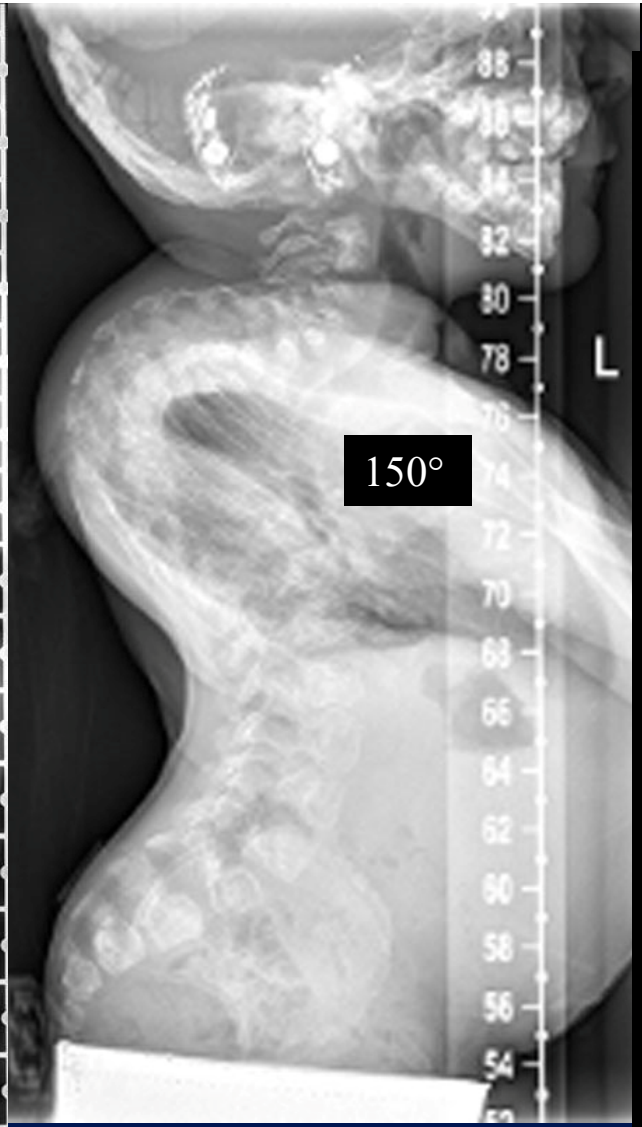
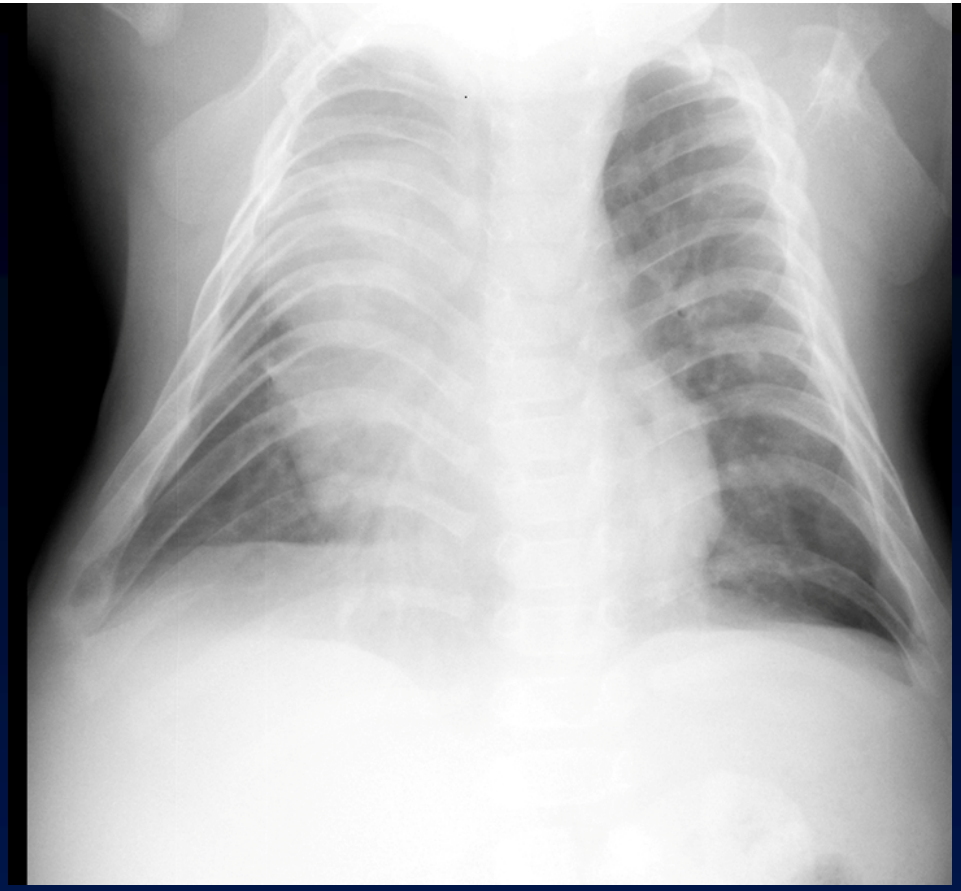
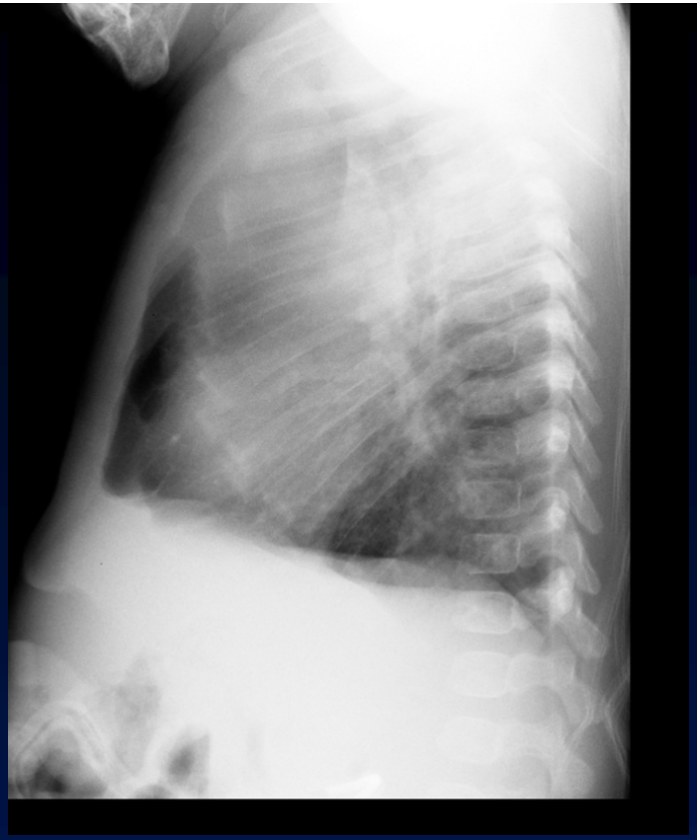


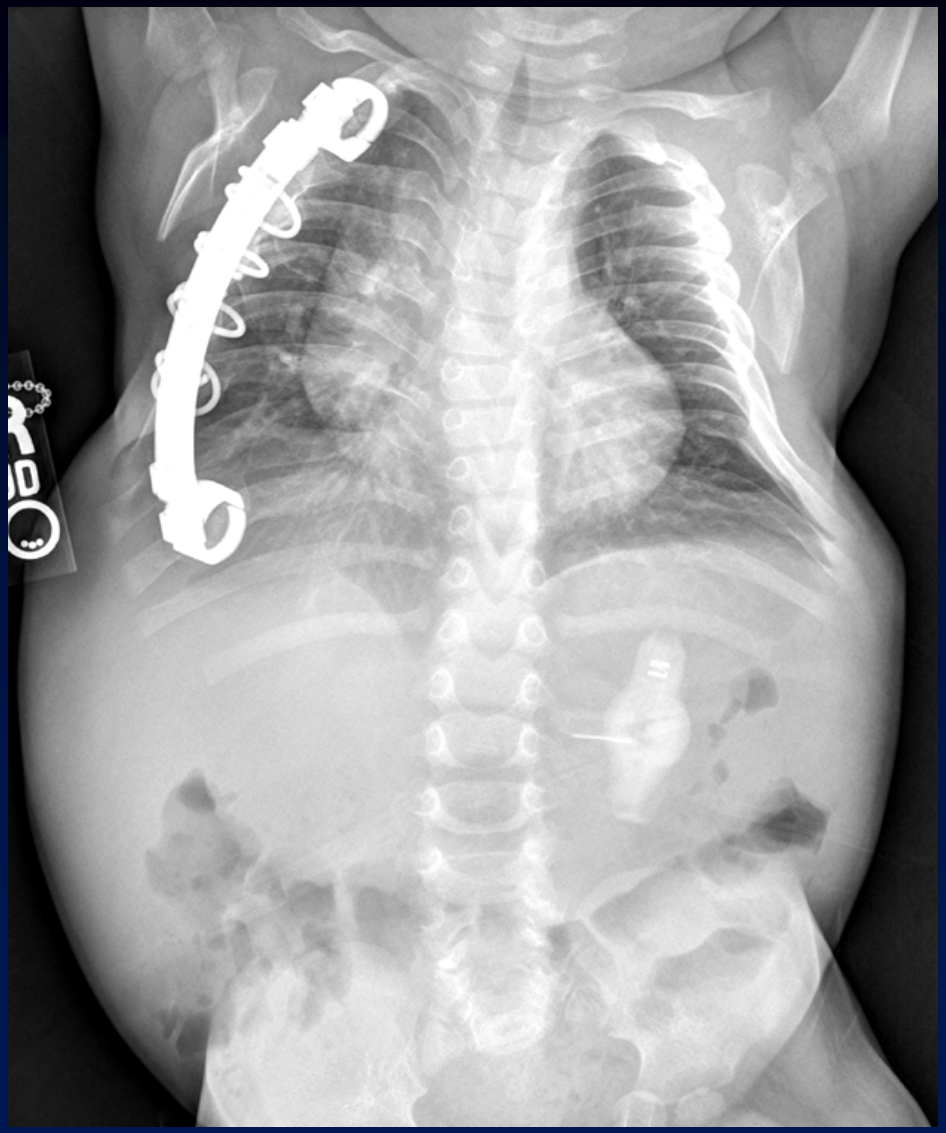


- 8 y.o. male
- Cleidocranial Dysostosis
- What is your plan?



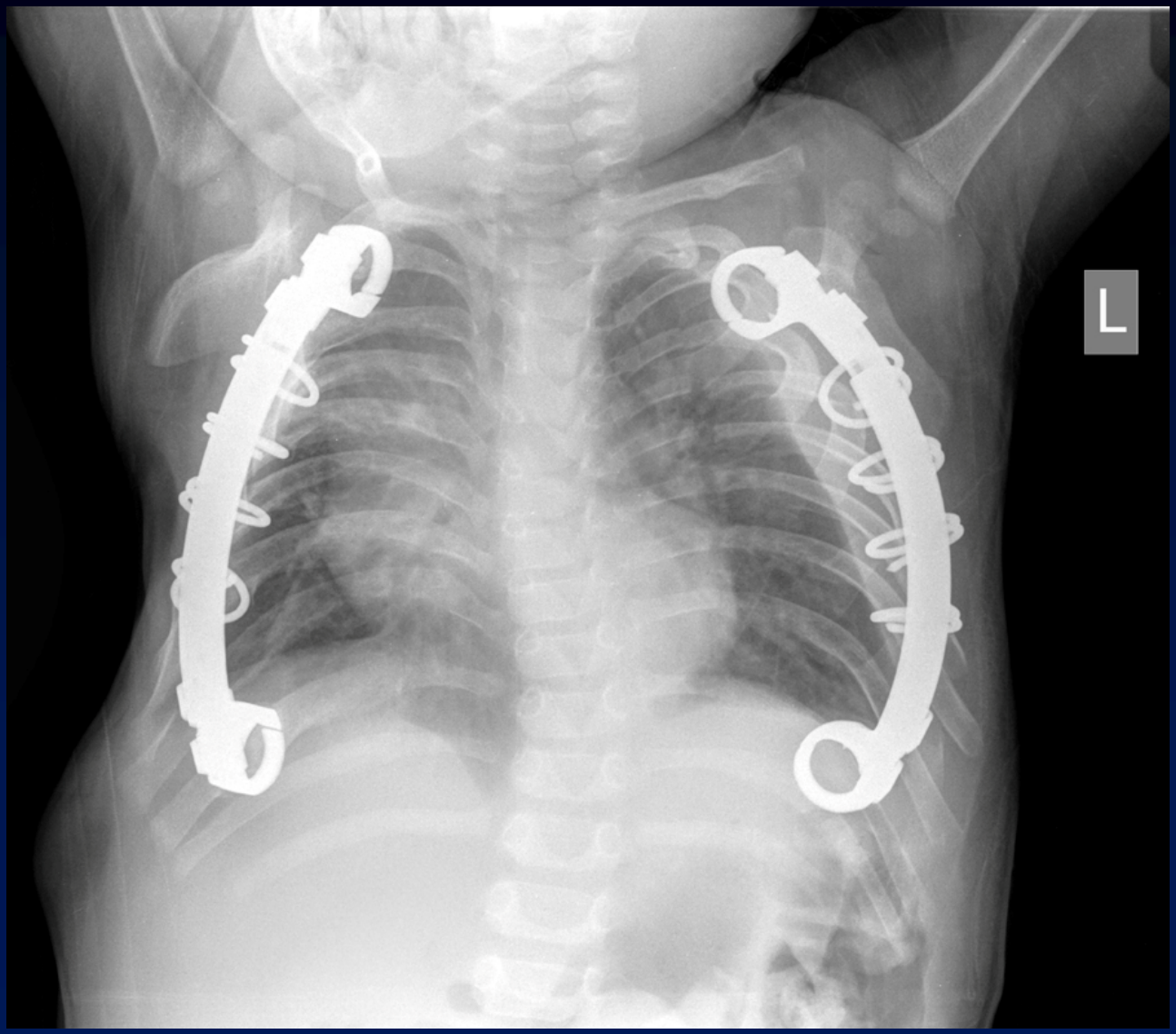


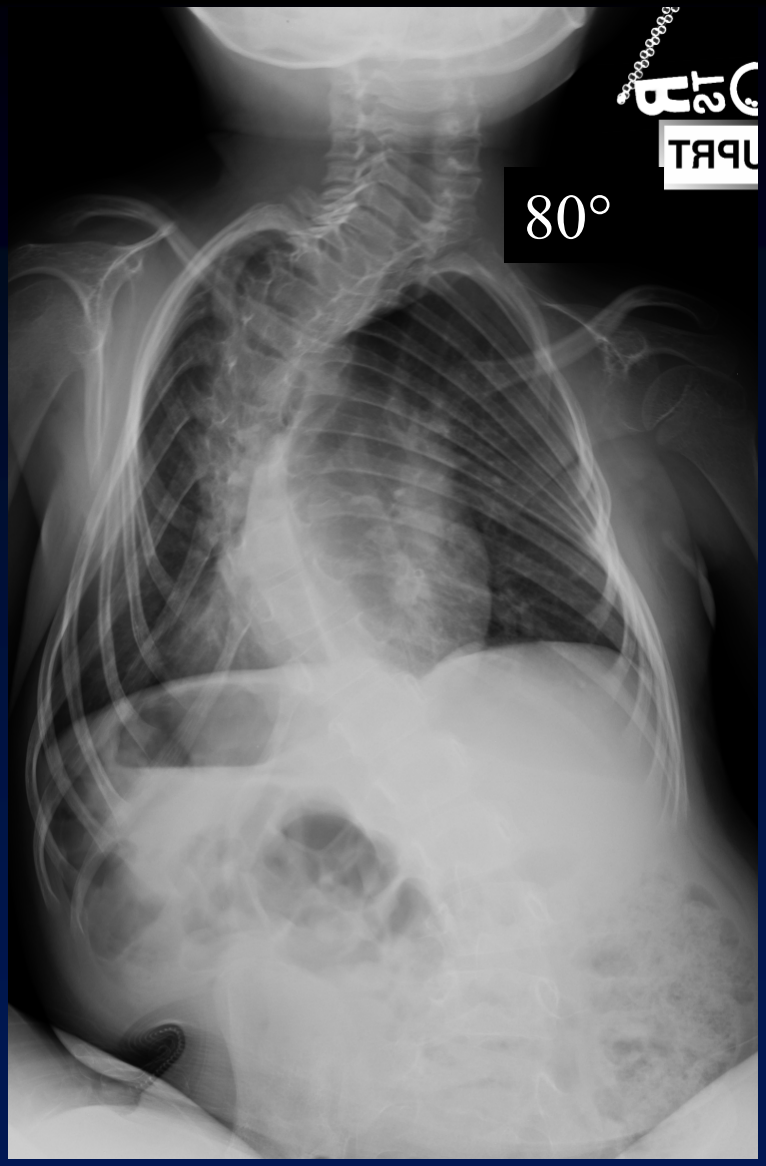
- 8 month old boy
- Chest wall deformity, similar to bell-shaped chest.
- Restrictive lung disease, with continual oxygen use.
- Status post genetics evaluation with no specific syndrome identified.



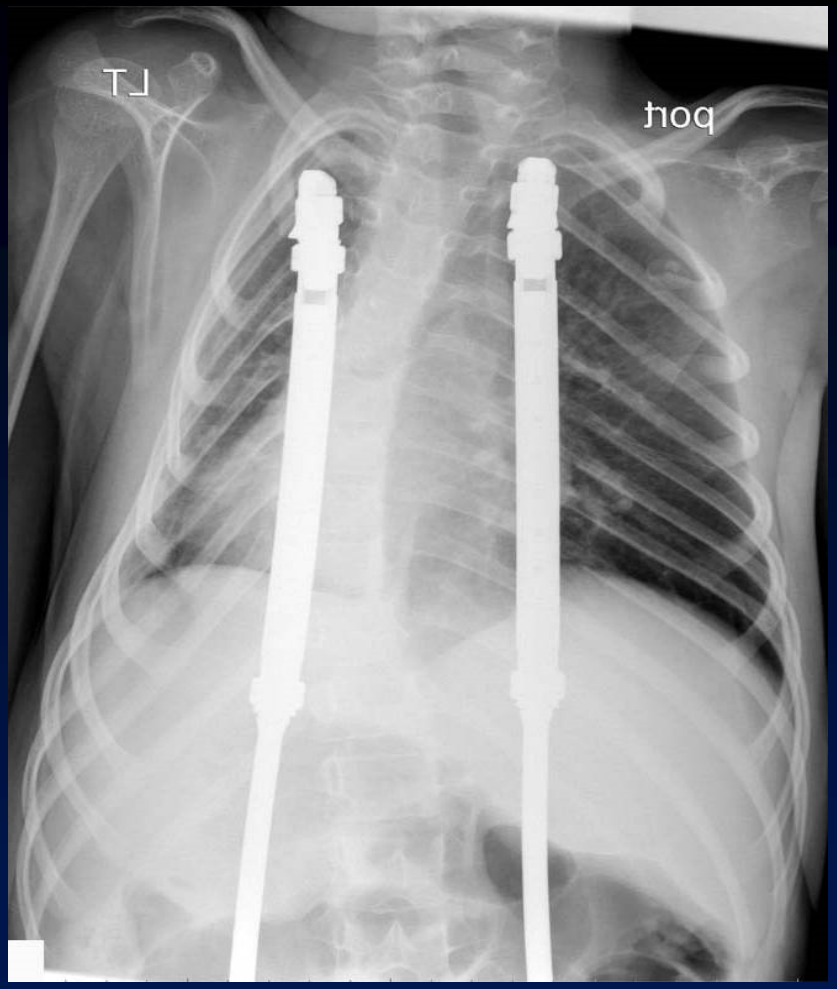
5 months post op from 1st surgery

Immediately after 2nd side

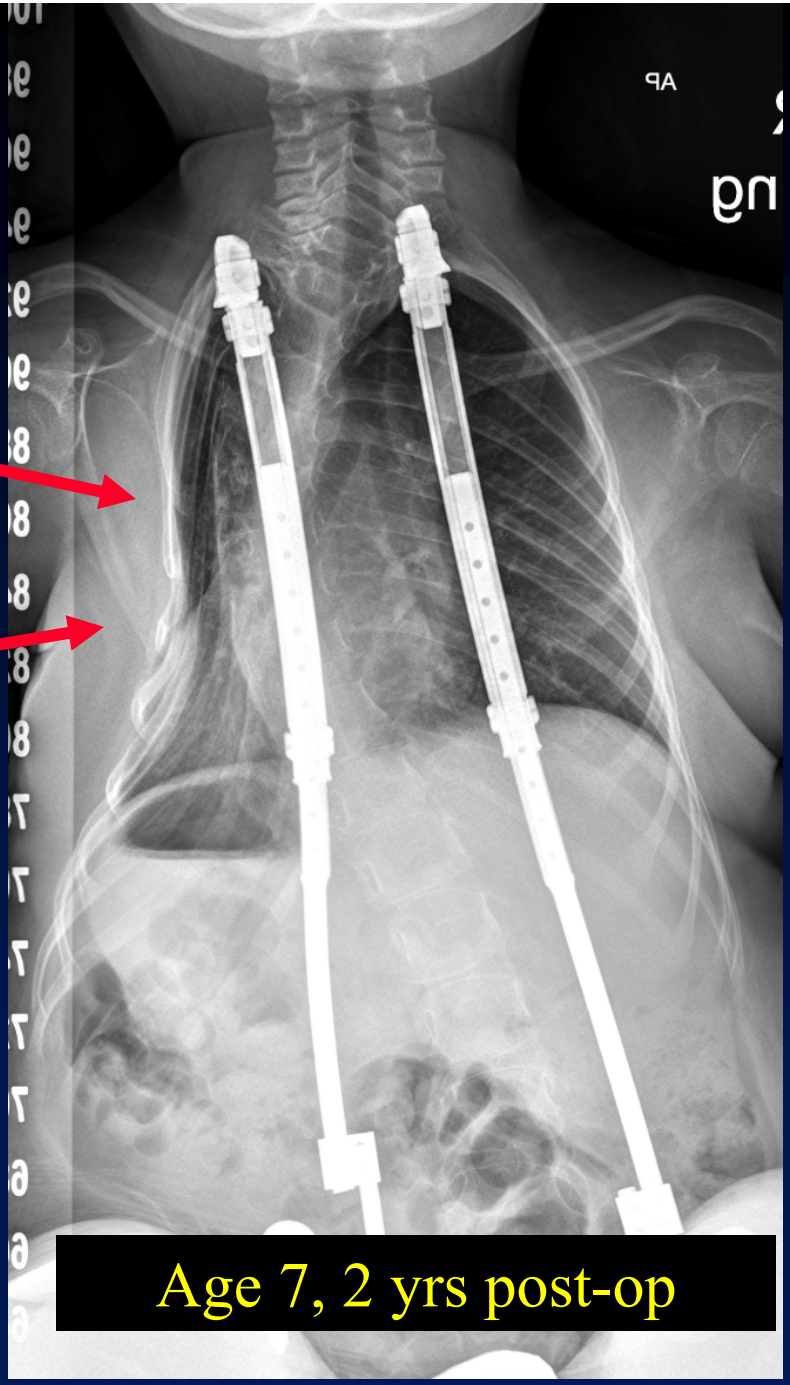




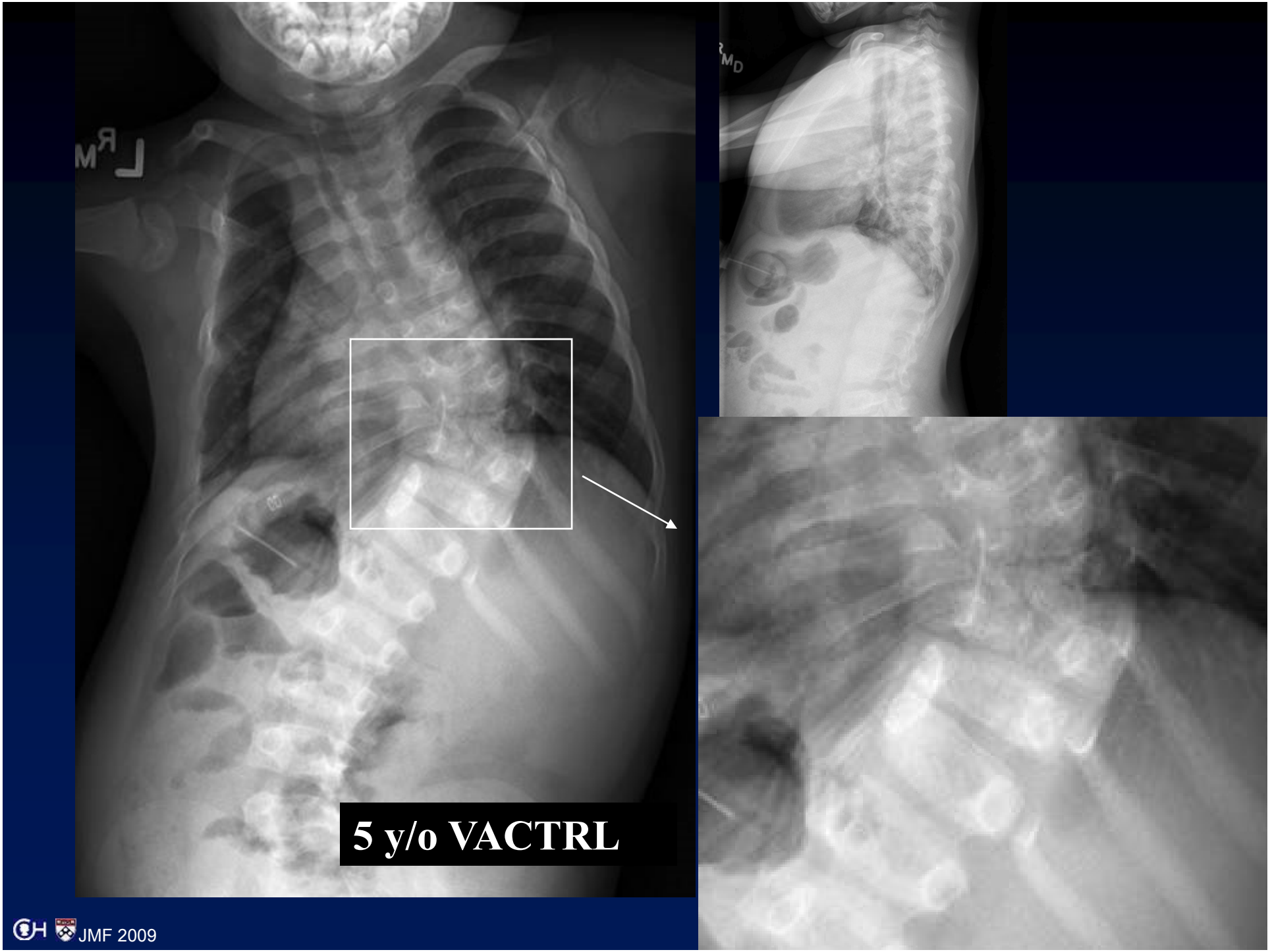
5 y/o with SMA type 2, declining respiratory function, progressive deformity

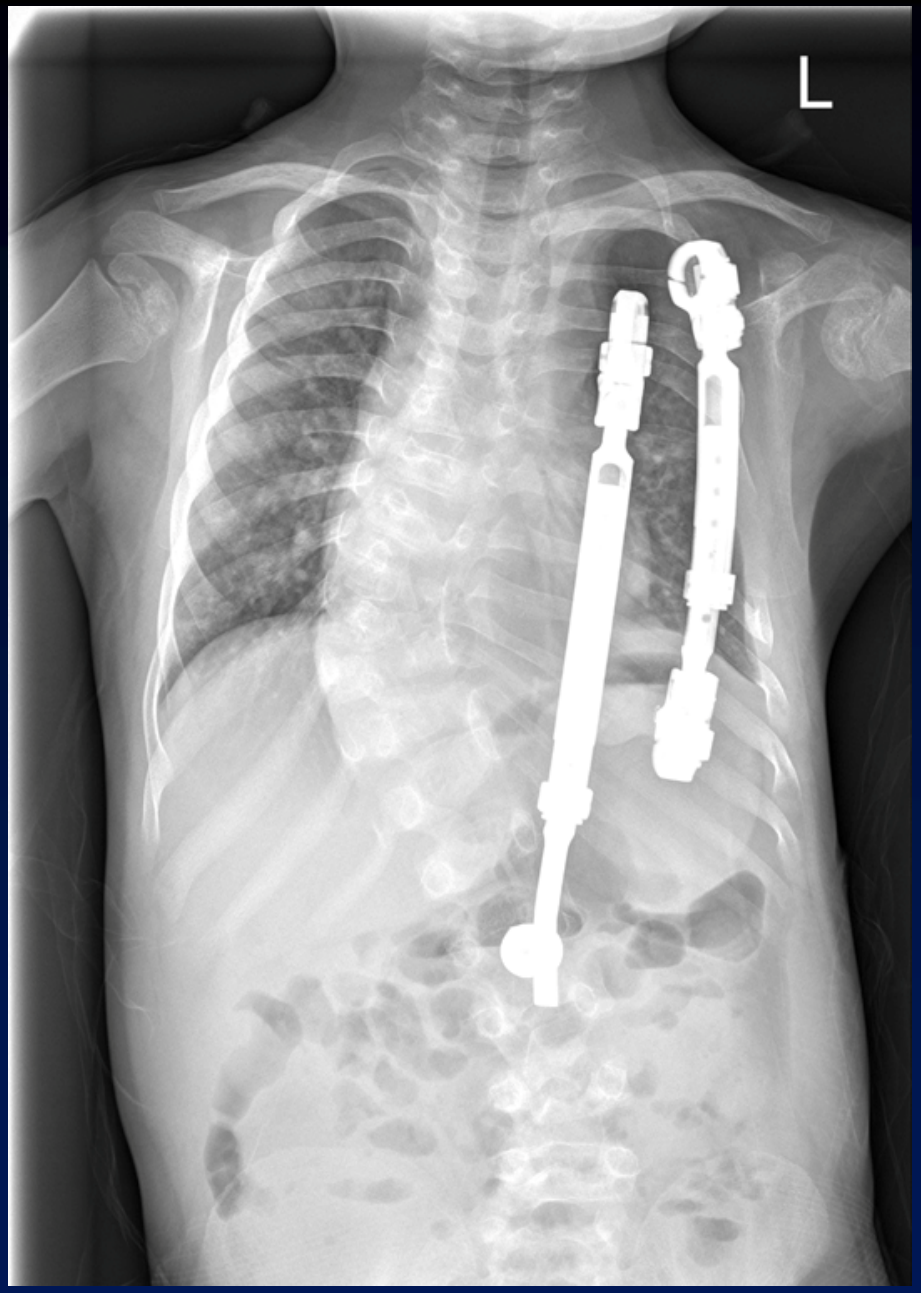


Immediately post-op



Age 7, 2 yrs post-op


5 y/o VACTRL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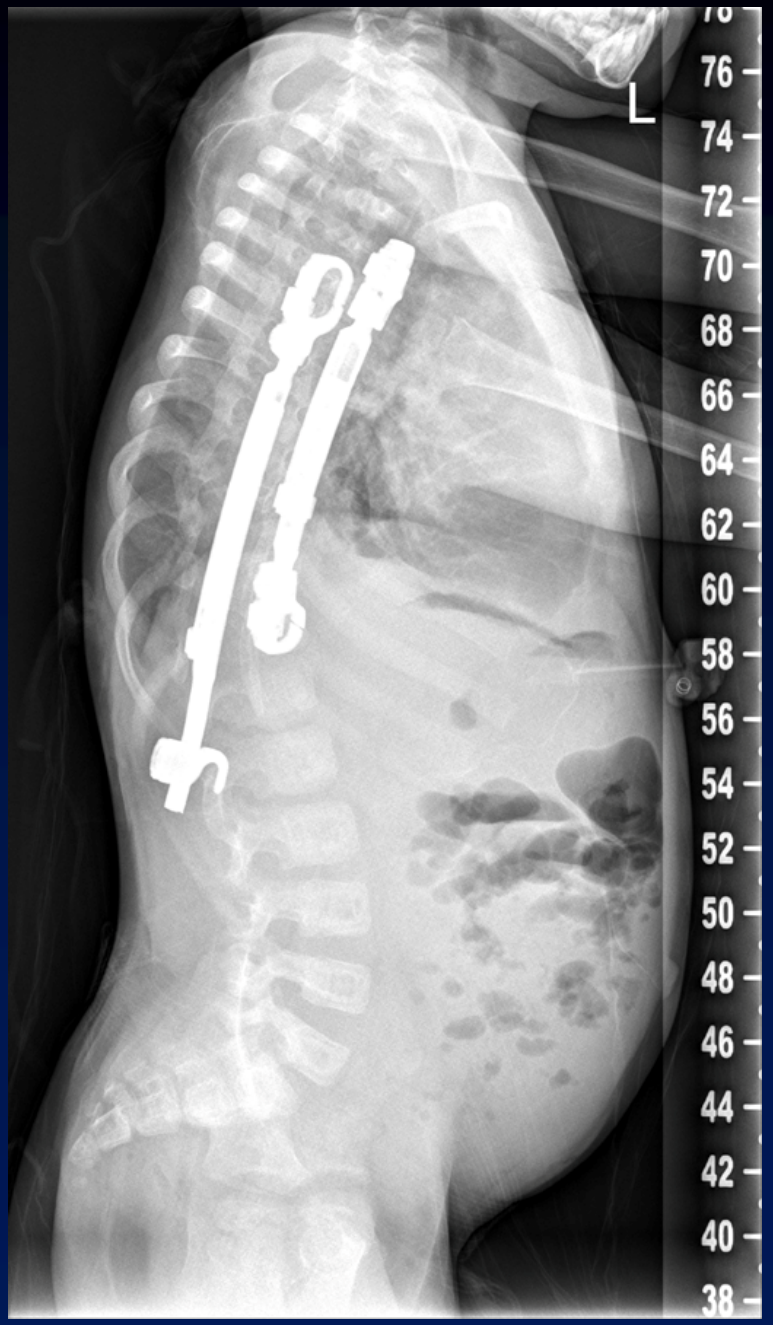
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Thank you