Unique Applications of VEPTR For the Management of TIS

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Thoracic Insufficiency Syndrome

- Inability of the thorax to support normal respiration and/or lung growth
- Infantile and early onset spinal deformity
- Primary thoracic dysplasias – constricted hemithorax

Atypical Application

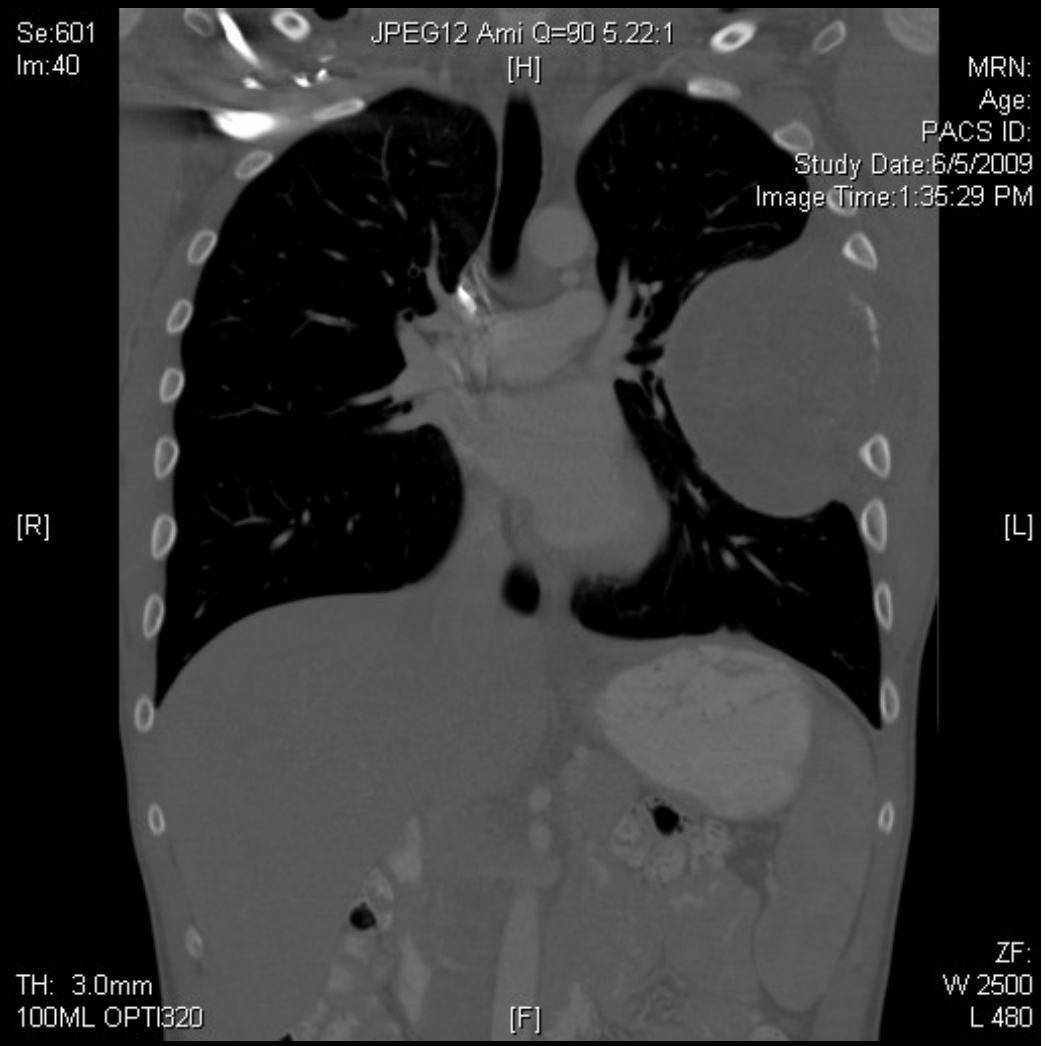
- Primary chest wall insufficiency 5 Pts.
- Primary spinal deformity with unique limitations for implantation 2 Pts.
- From 2001-2009:
  - 88 Rib based VEPTR implantations 51 Pts.
  - 26 Spine based VEPTR implantations 25 Pts.

Primary Chest Wall Insufficiency

- All IRB/HDE consent for off label use
- Chest wall resection due to tumor (4)
  - Ages 8-19 years
  - Dx;
    - Ewing's Sarcoma (2)
    - Osteosaroma (1)
    - Desmoid tumor (1)
- Primary absence of chest wall – (1)
  - Age 5
  - Poland's syndrome

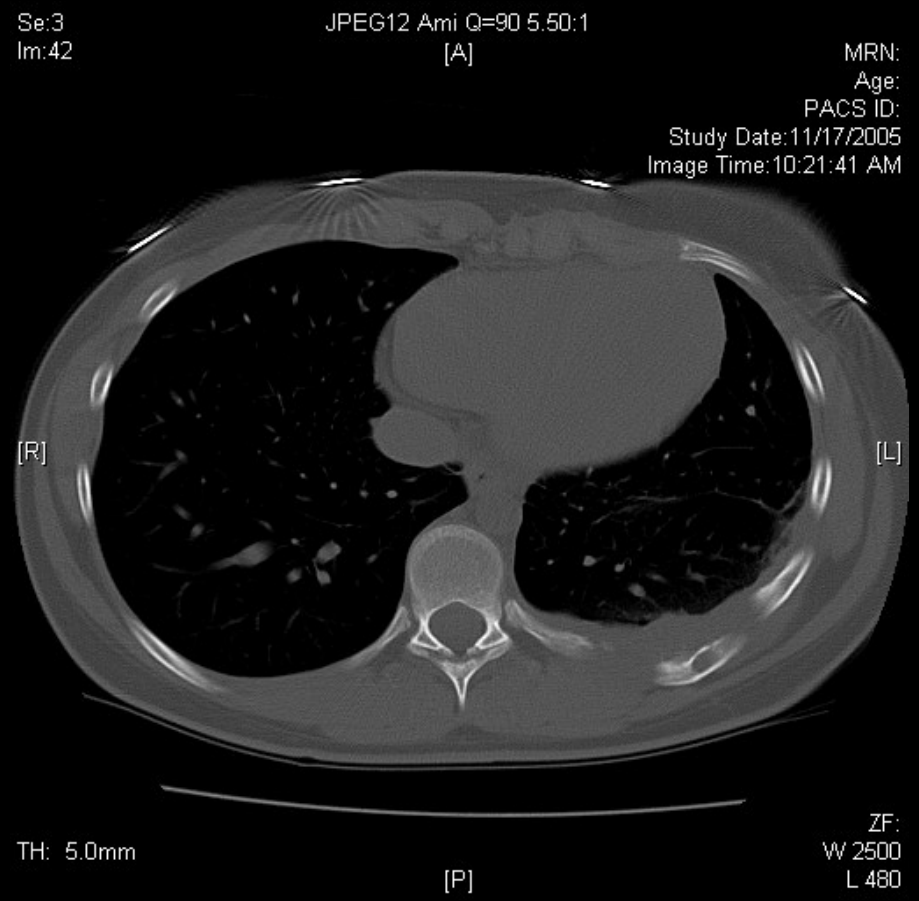
Chest Wall Resection

- Implantation at time of tumor resection
- Resectable
- Survivable
- 3 rib resection



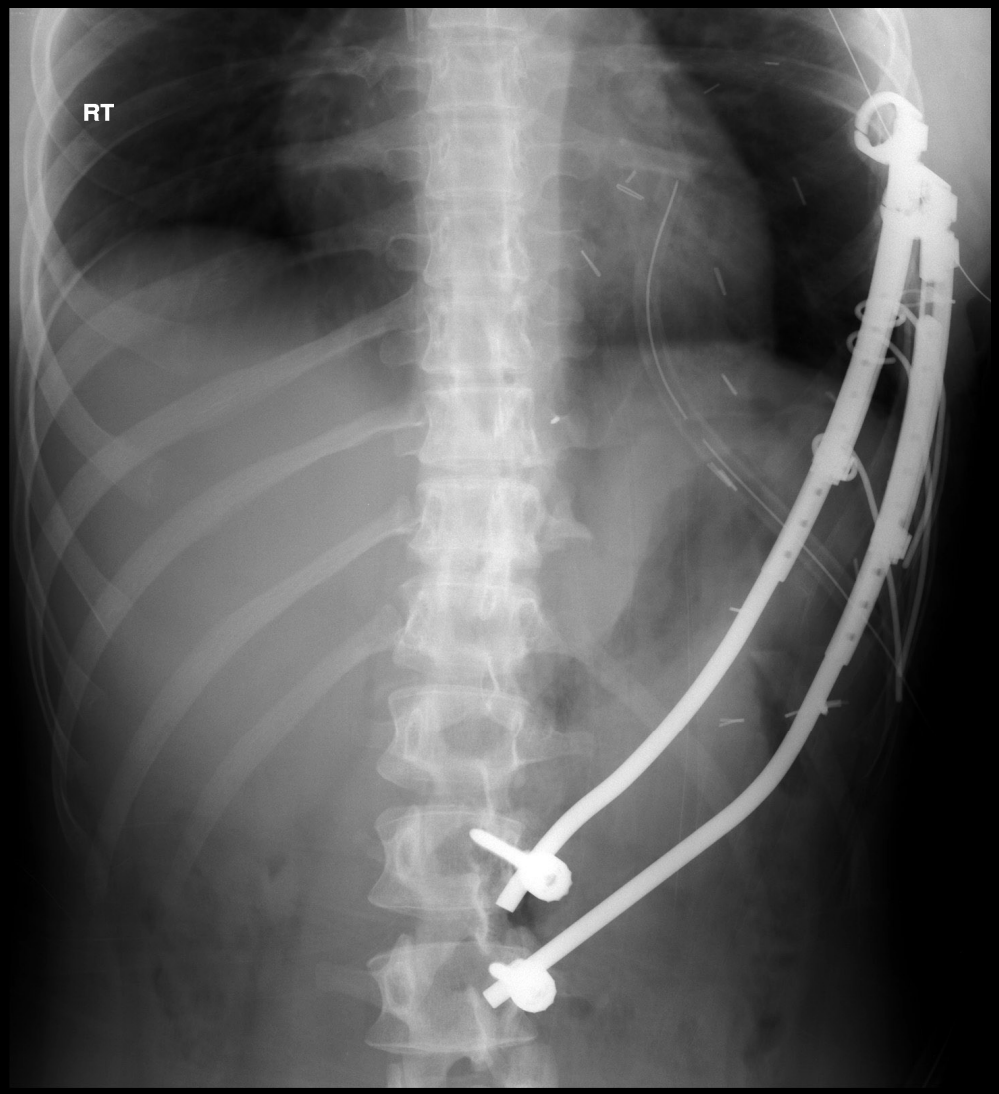
Technique

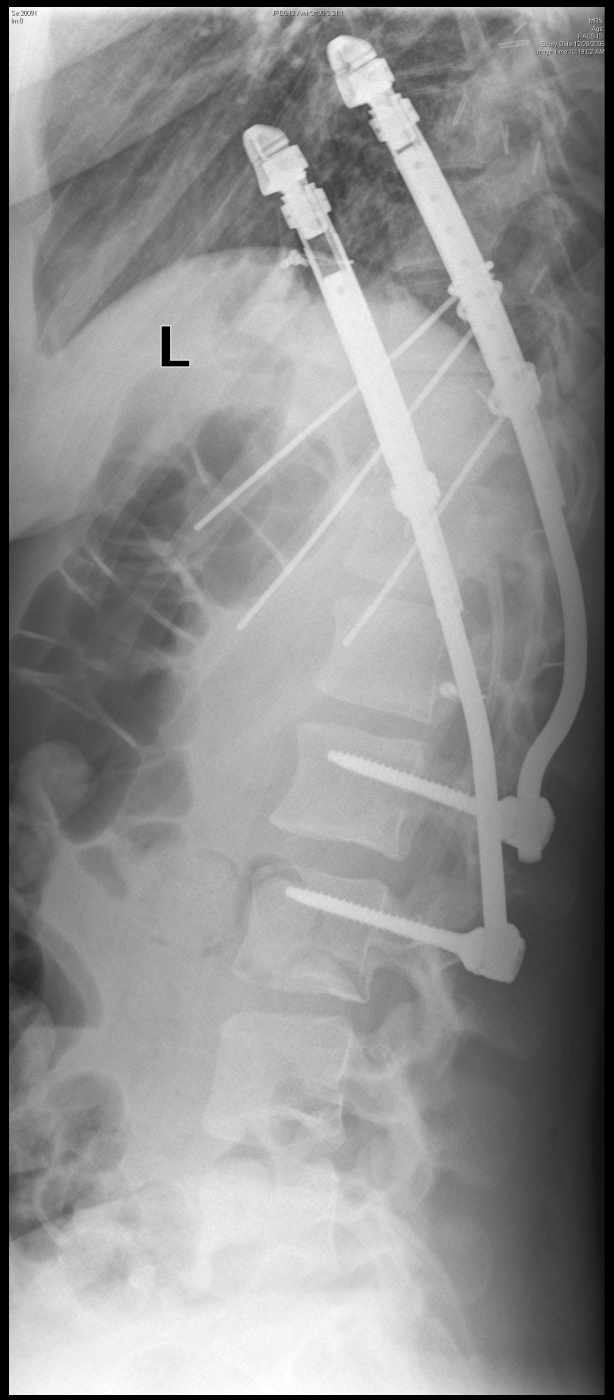
- Rib to Spine
- Rib to Rib
- Transverse rods or plates
- Limited number of expansions



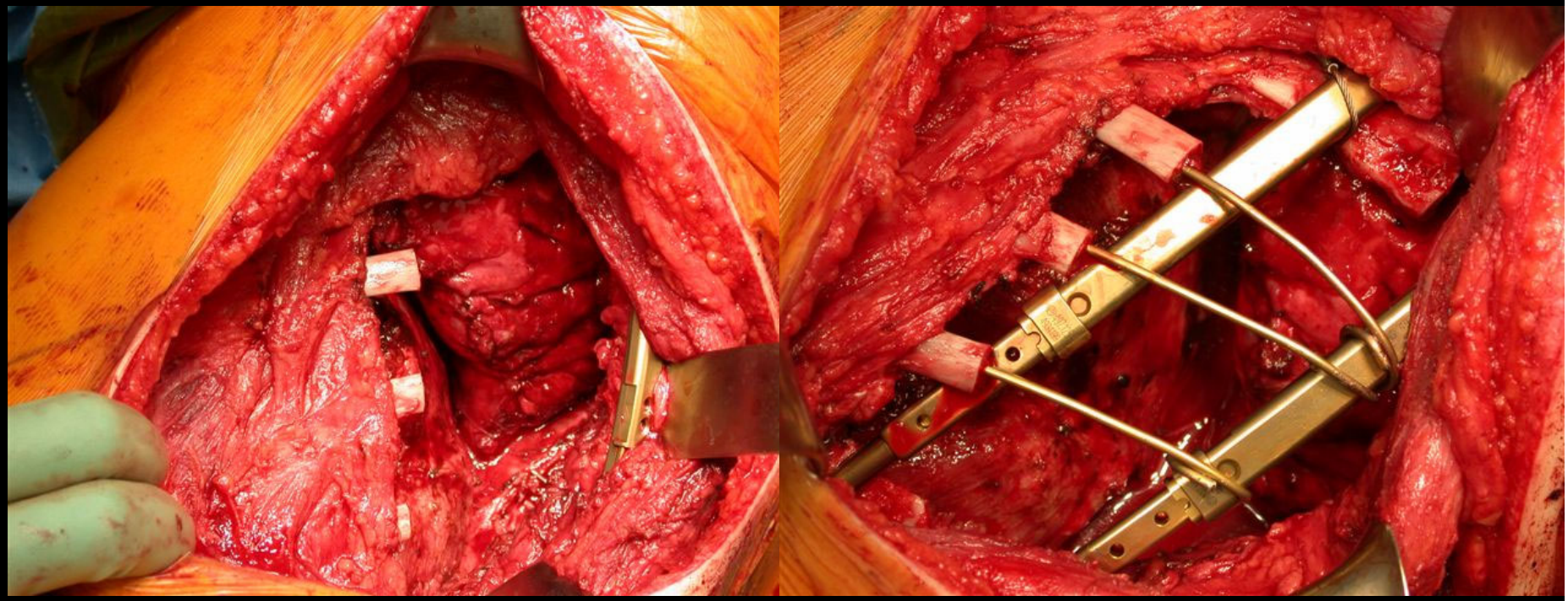
Desmoid tumor – planned resection post ribs 7-10

Rib to Spine

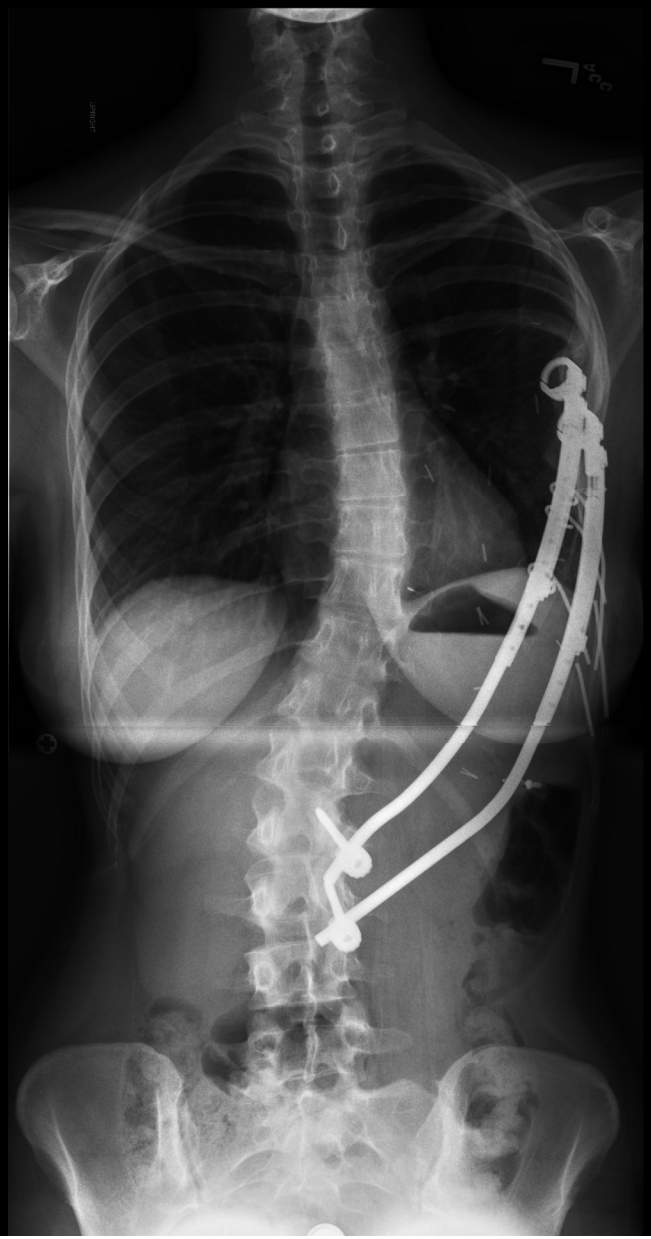


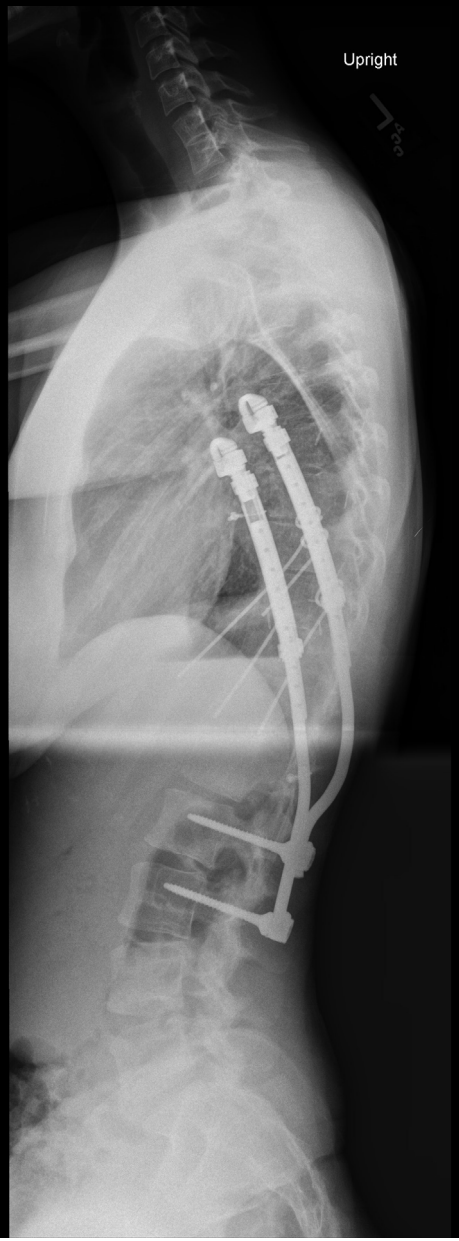


Rib to Spine



Rib to Spine – 5 yr F/U





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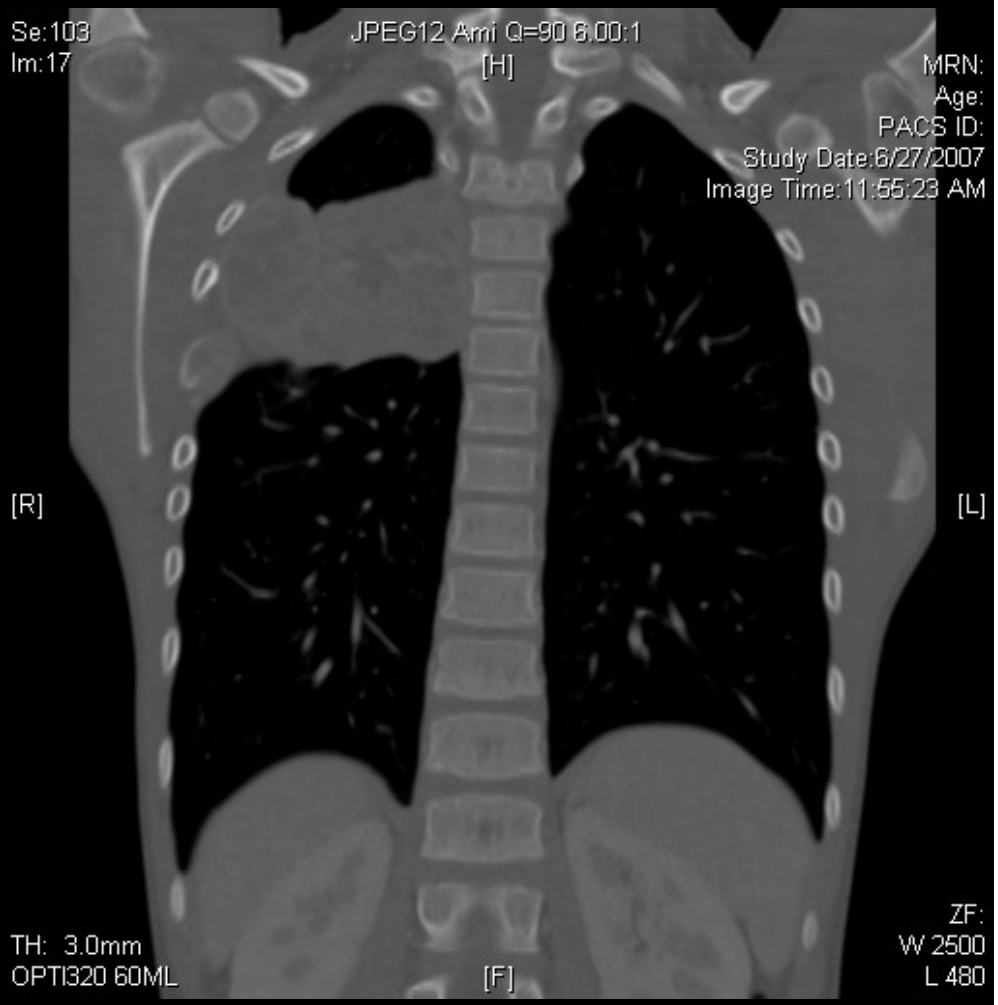
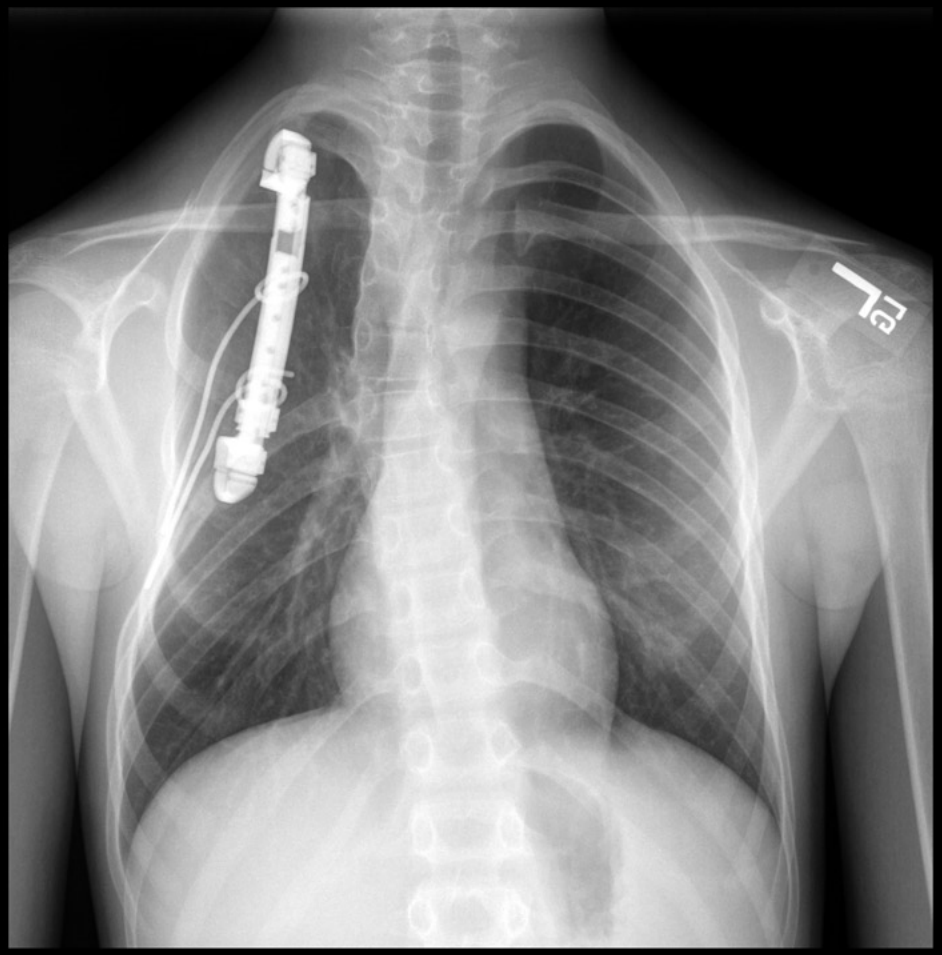
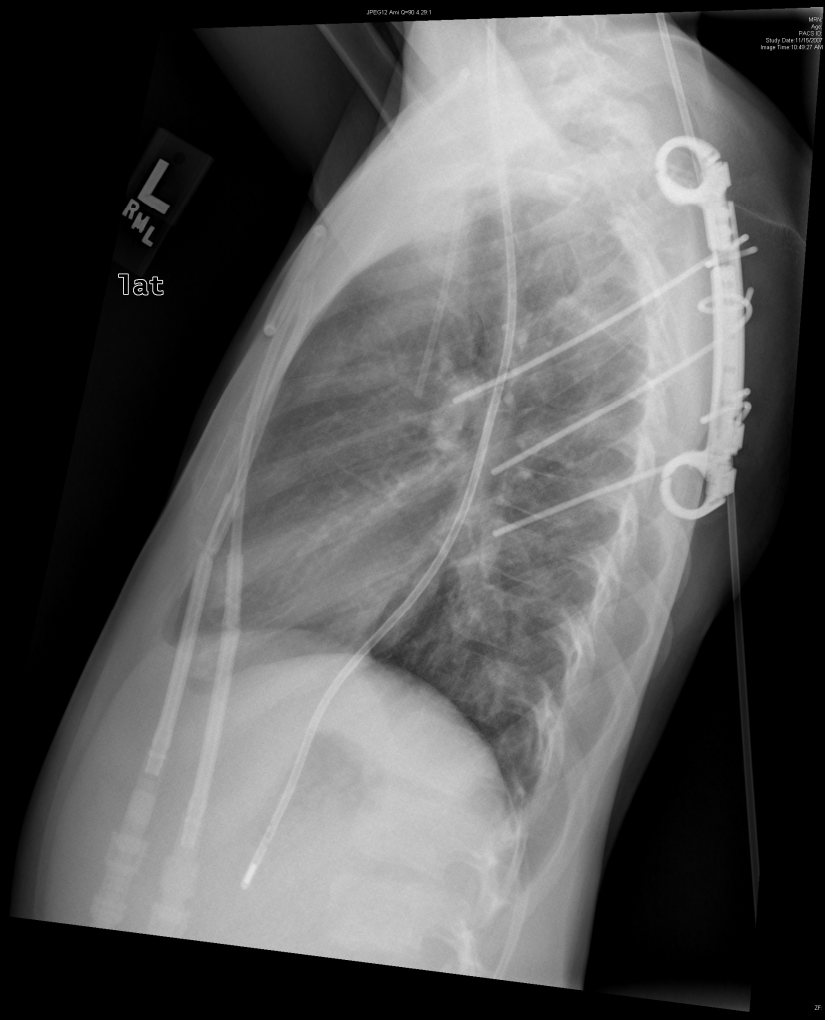


10 y.o Ewing's Sarcoma – planned resection ribs 4-6

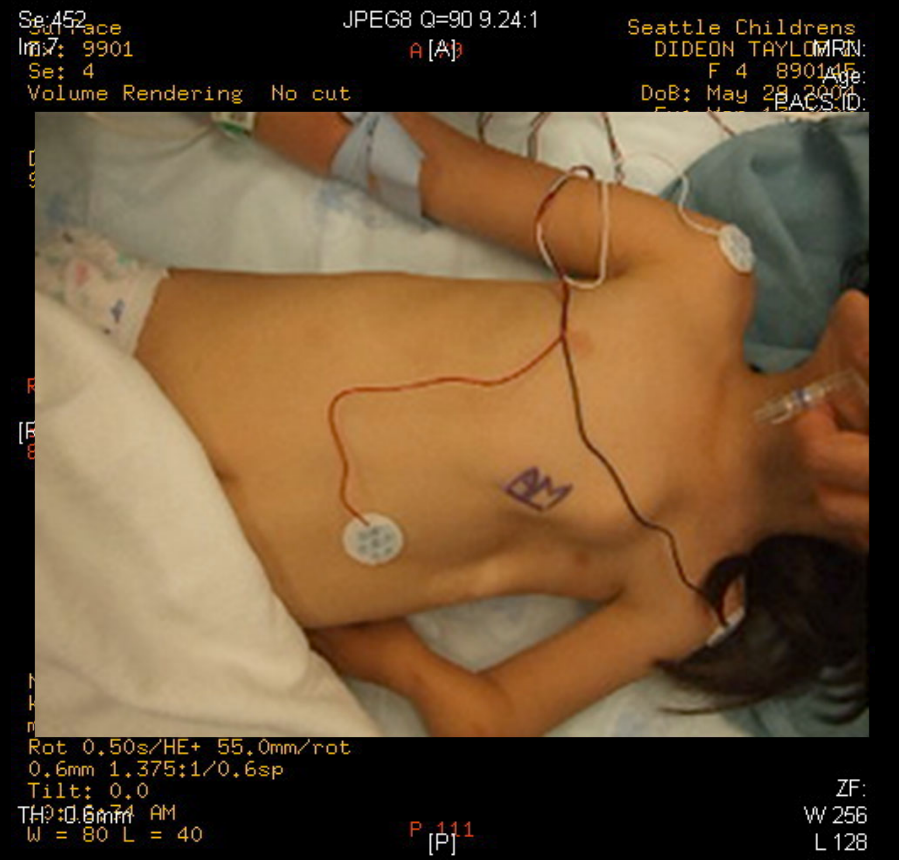
Rib to Rib





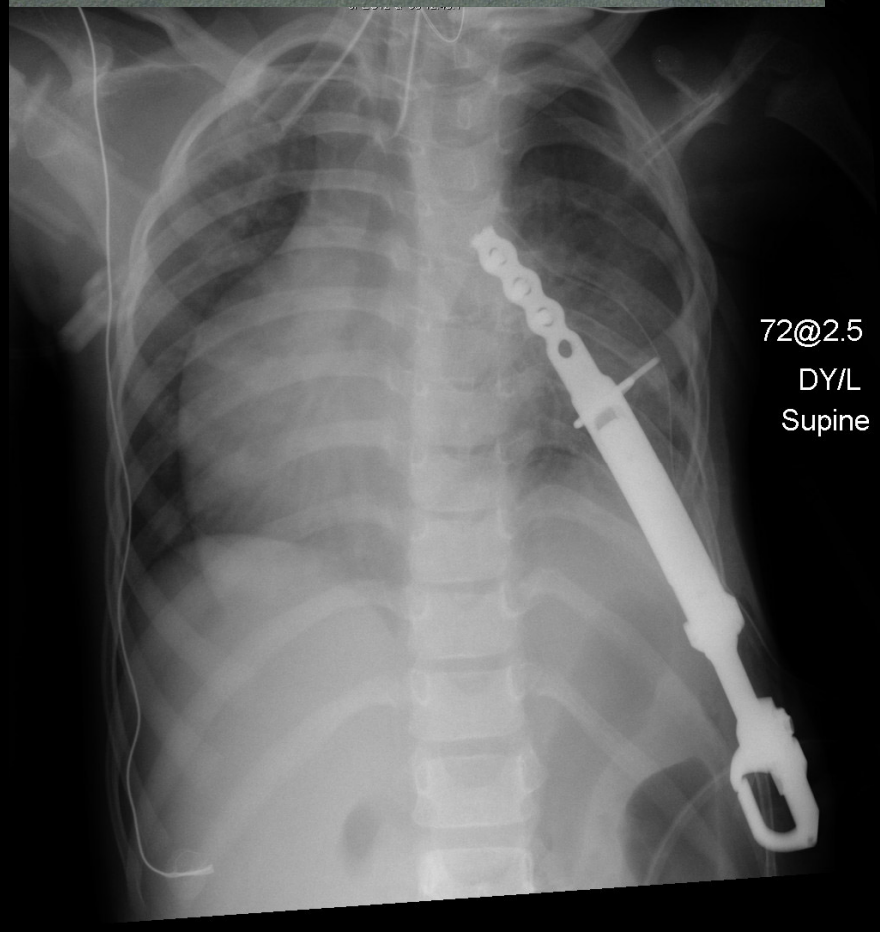
Primary Chest Wall Deficiency

- Poland's Syndrome
- Flail chest with worsening asymmetry of function
- Protection of pericardium



Technique

- Modification to custom VEPTR with sternal plate
- Sternum to rib construct
- Transverse fixation – limited due to size



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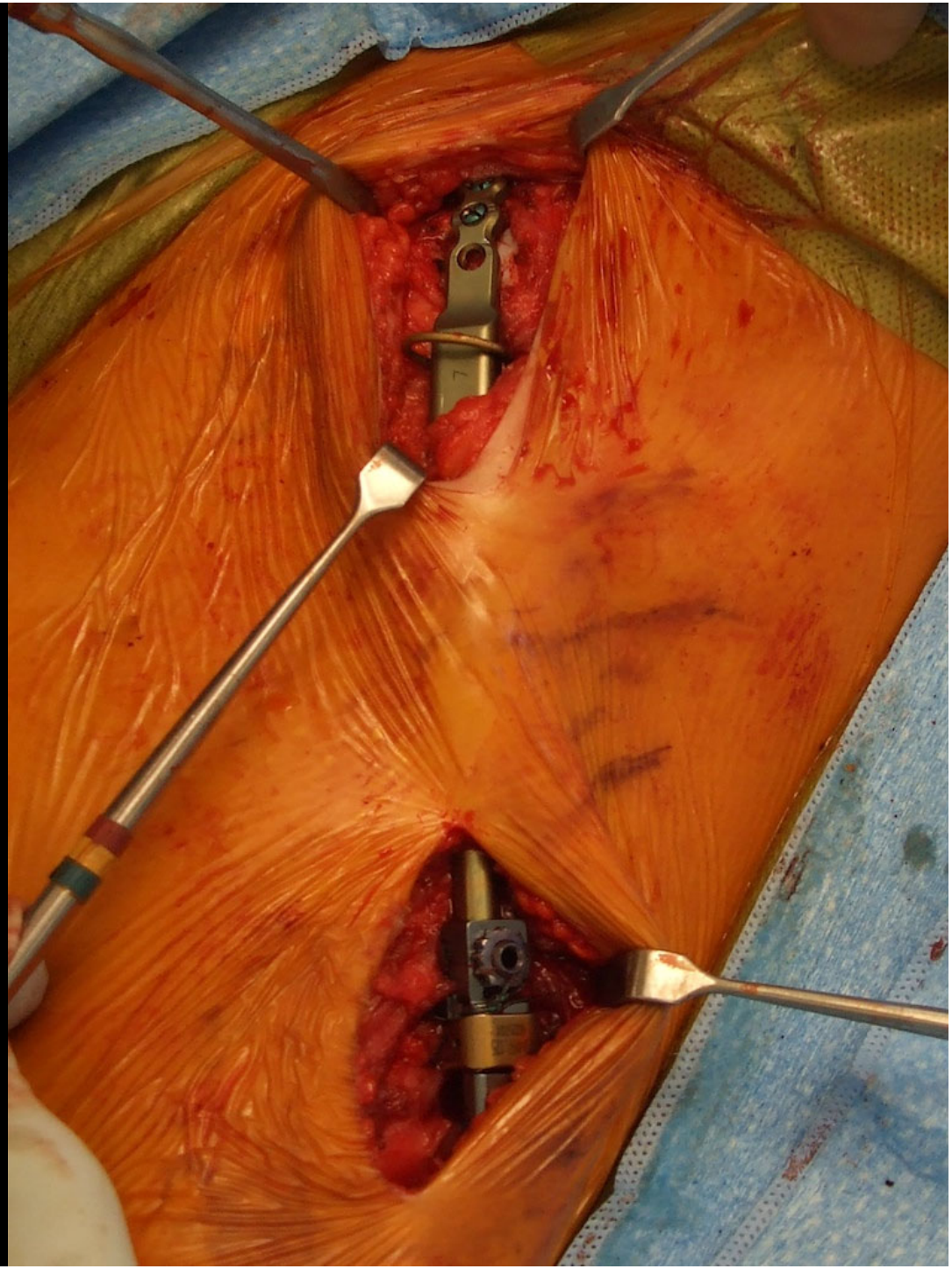
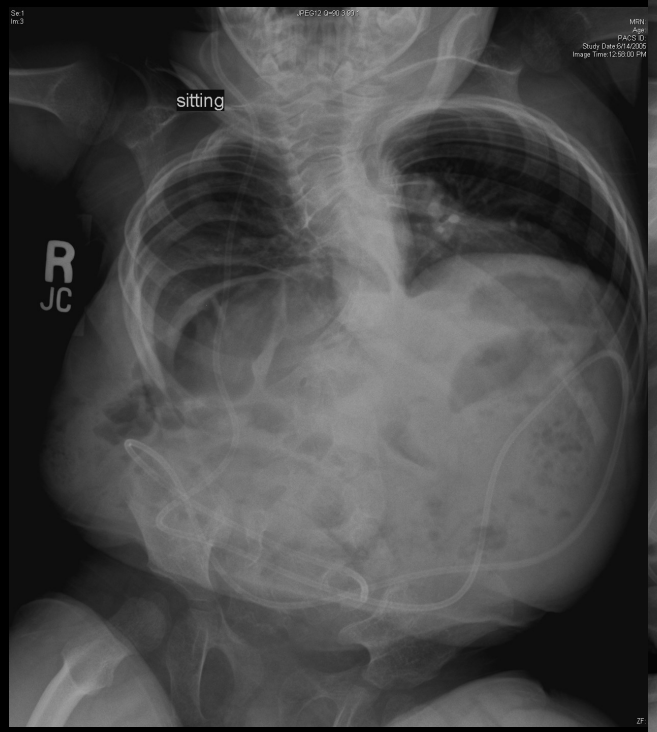
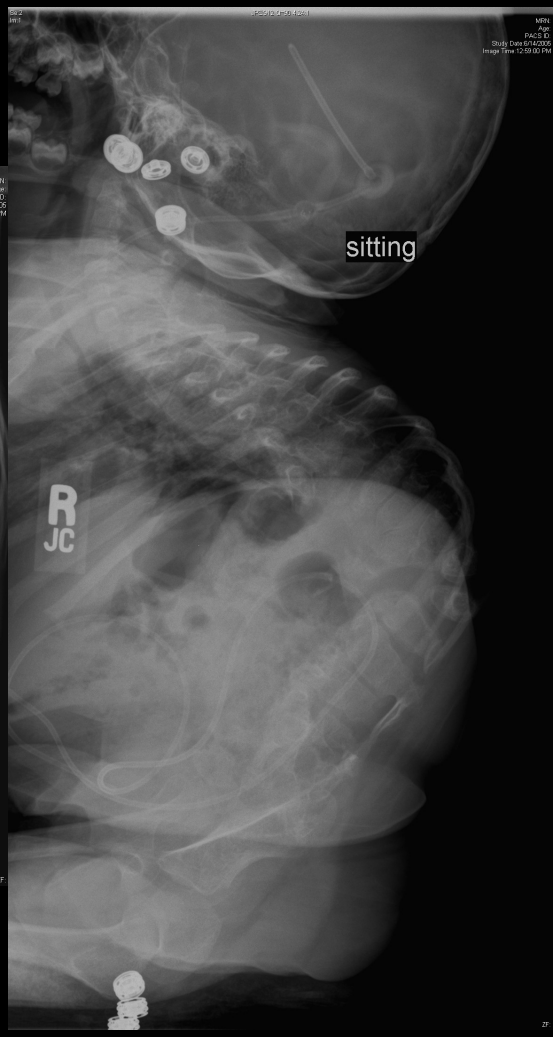


Unique Limitations for Implants Severe Anatomical Constraints

- 2 y.o. large lumbar meningocoele
- Progressive congenital curve
- Failed prior attempt at fusion in situ with cast immobilization





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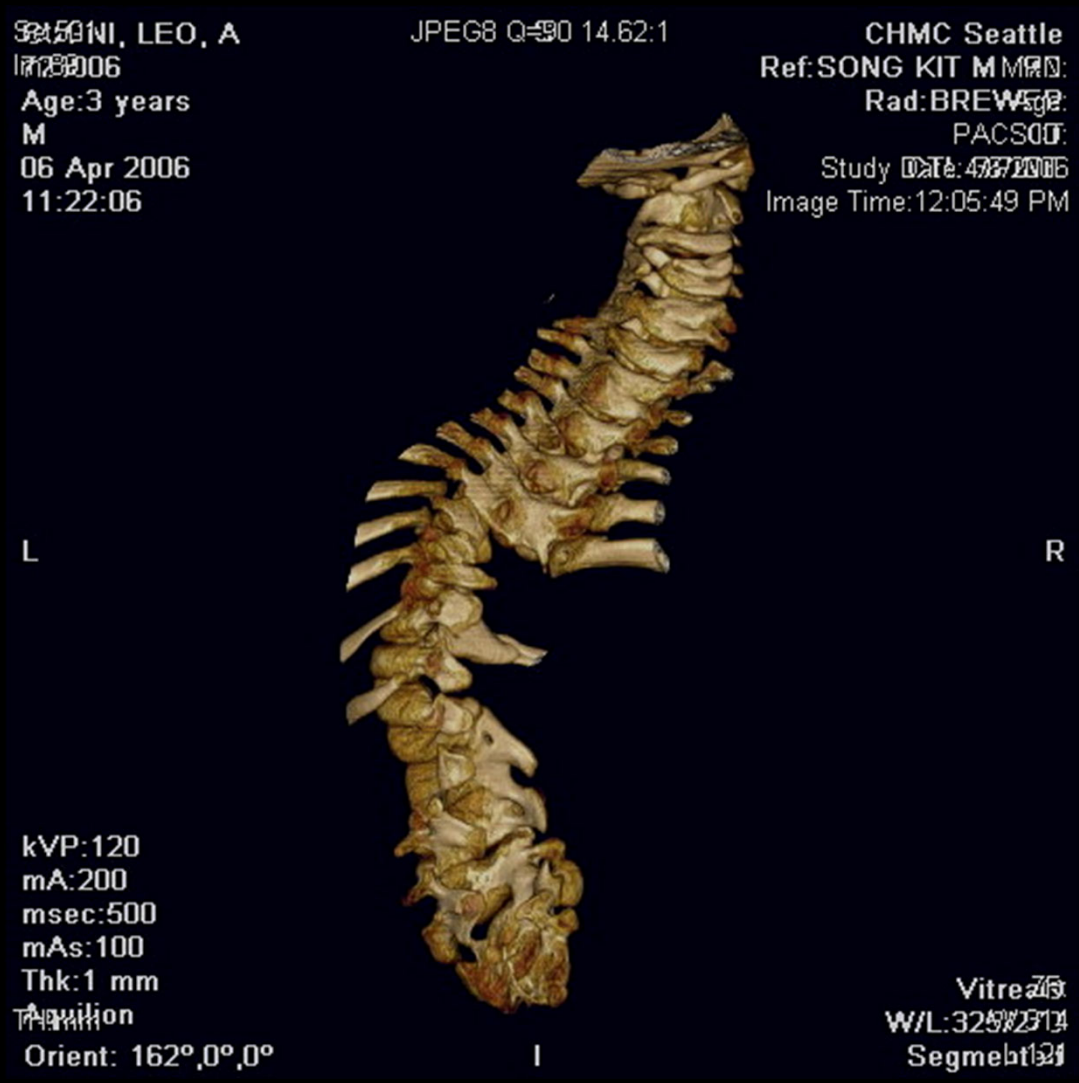

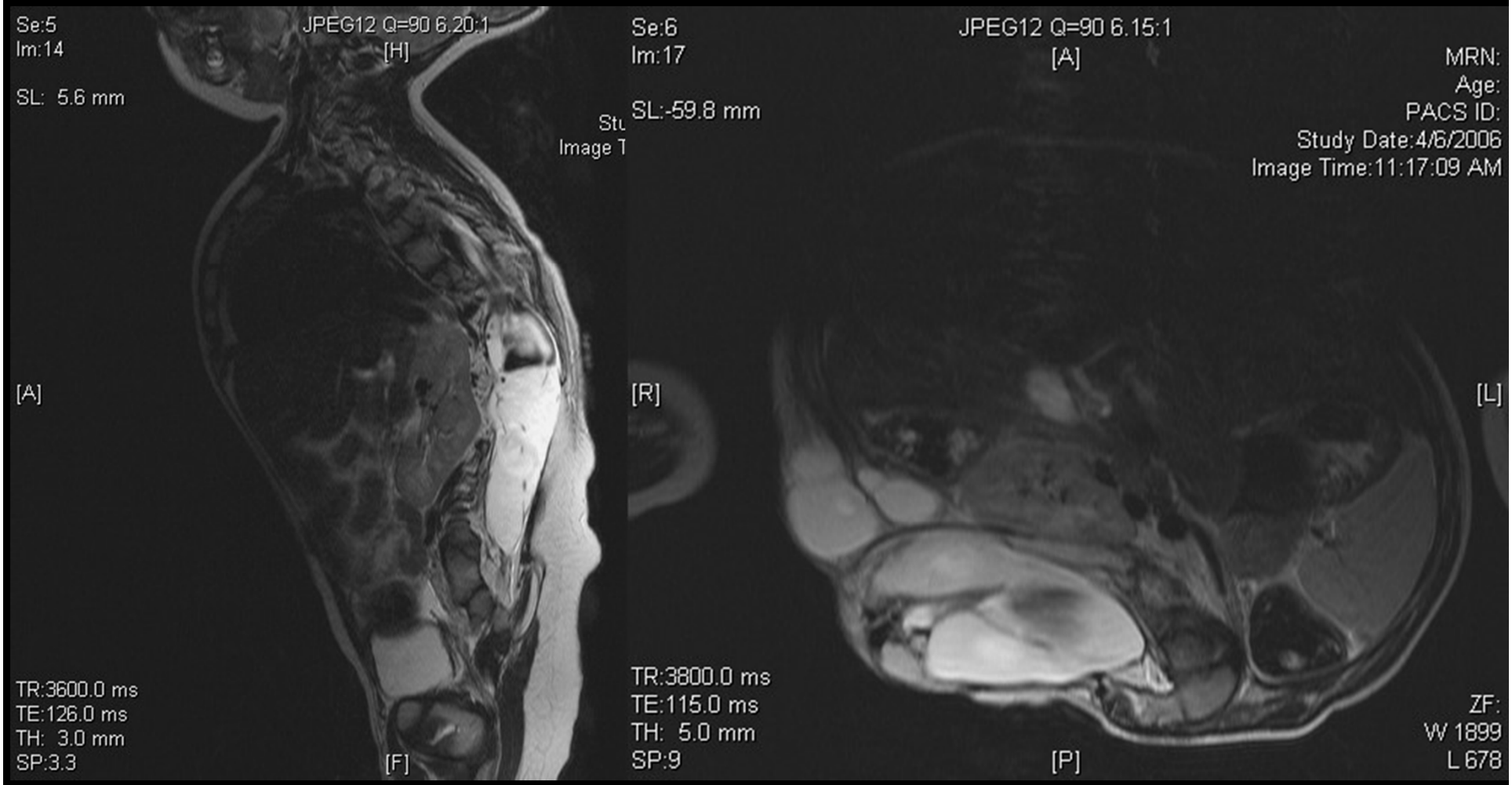
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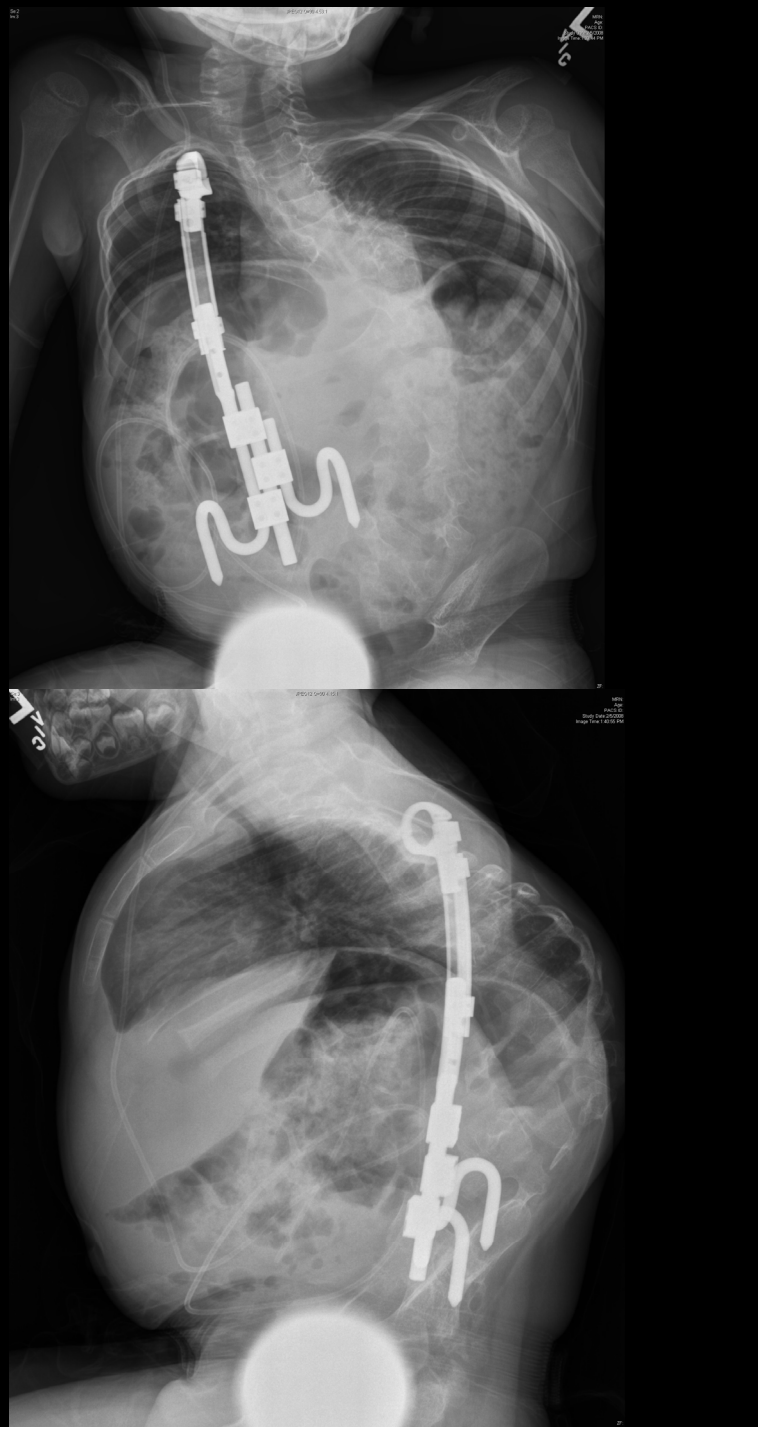
Technique

- Retroperitoneal placement
- Reversal of male/female component
- Expansions proximal
- Gortex liner
- 3 expansions – 1 revision to longer construct



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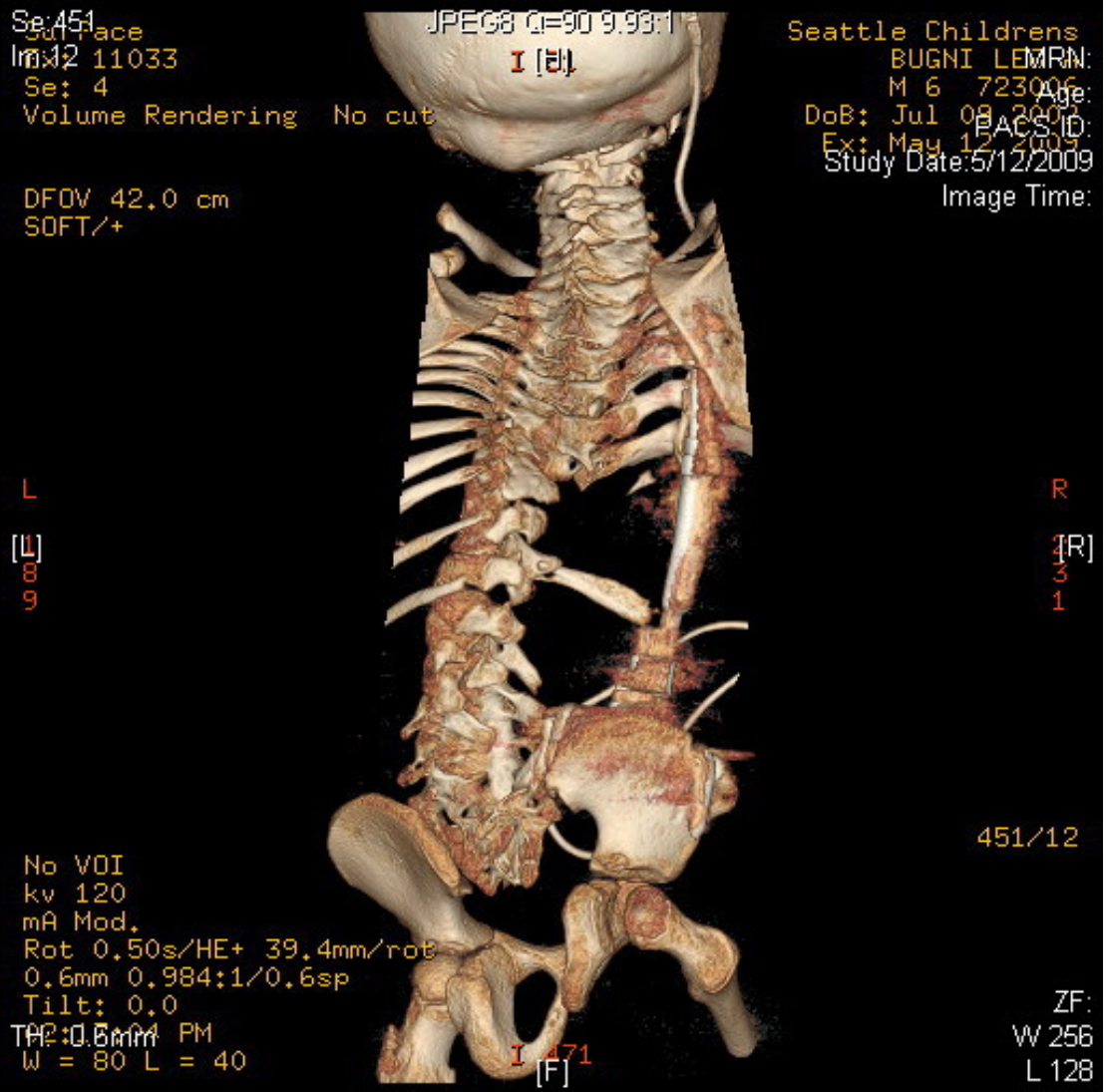
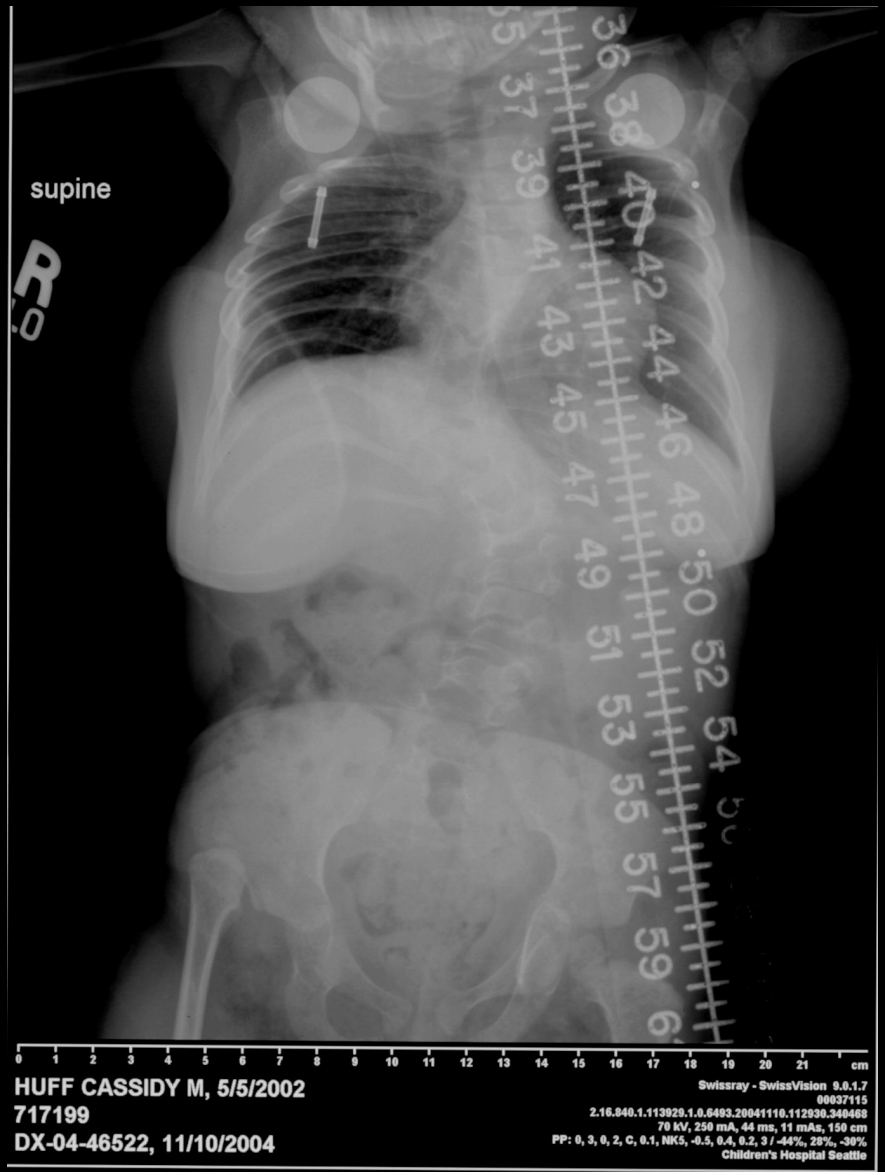


Unique Limitations for Implants Small Size / Hardware Prominence

- 3 year old
- Conradi Hunnerman Syndrome
- 16 kg weight
- Progressive kyphoscoliosis



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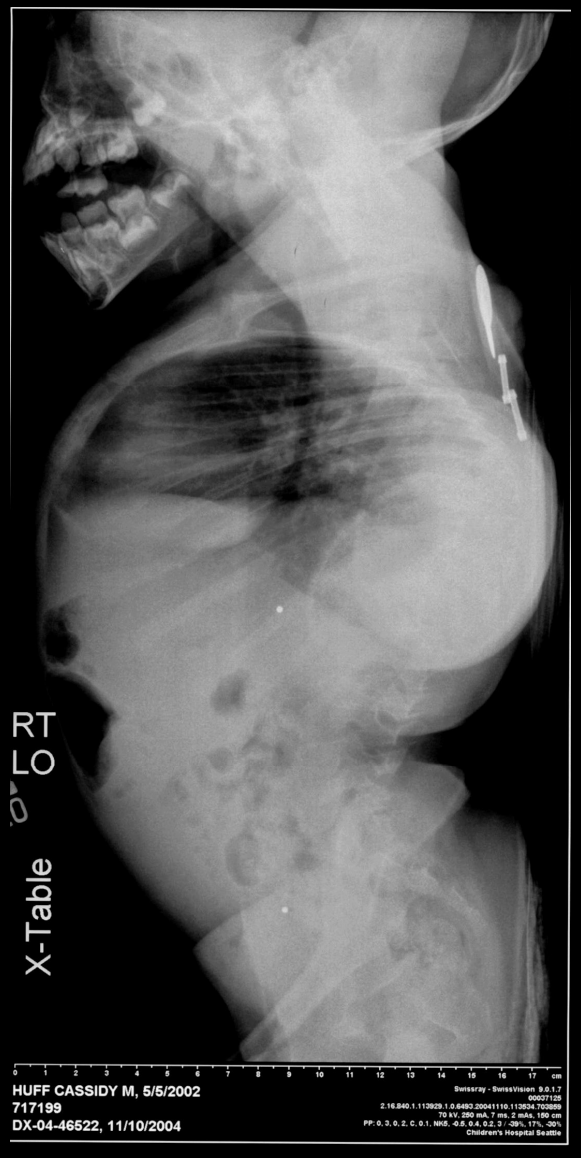

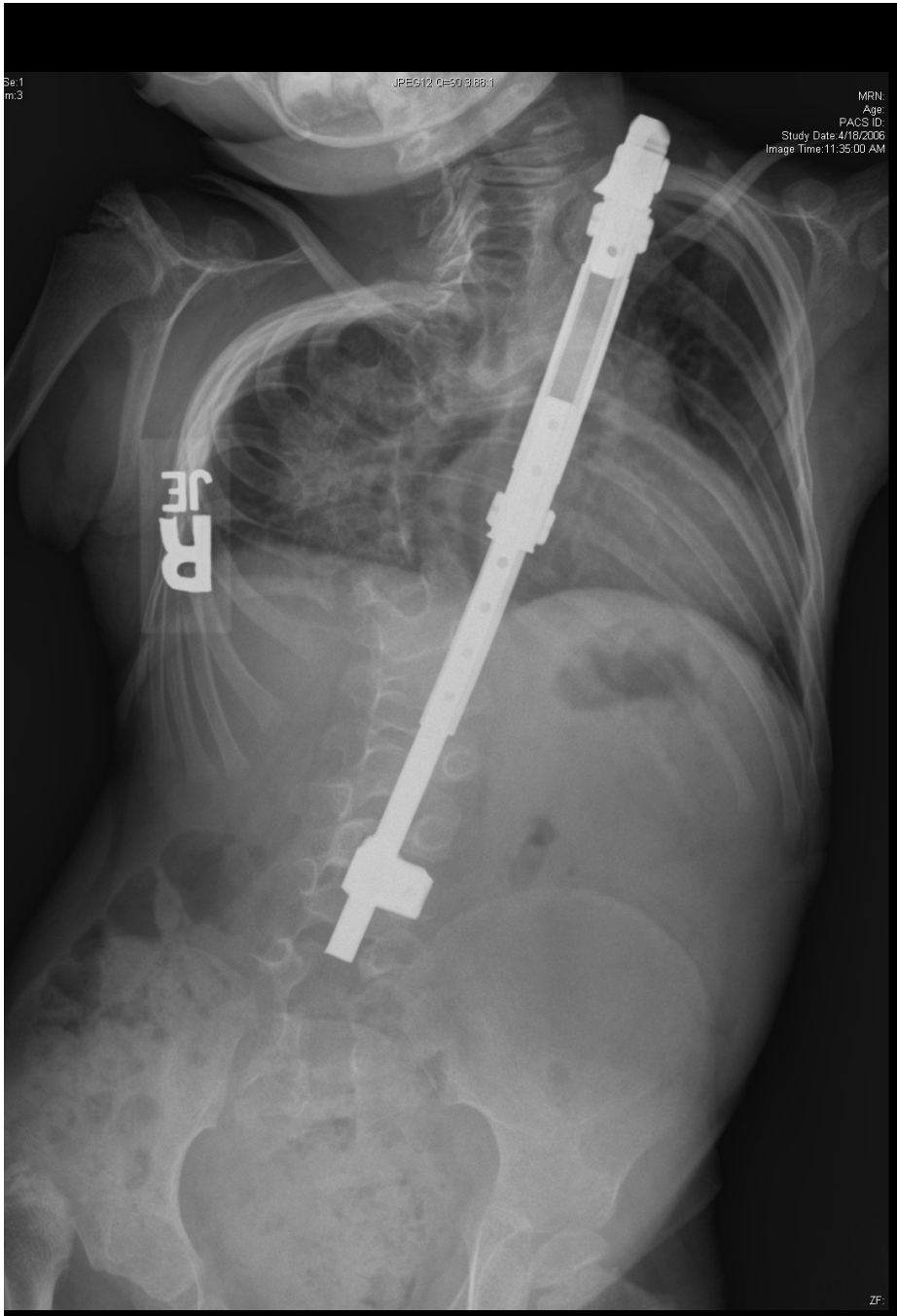
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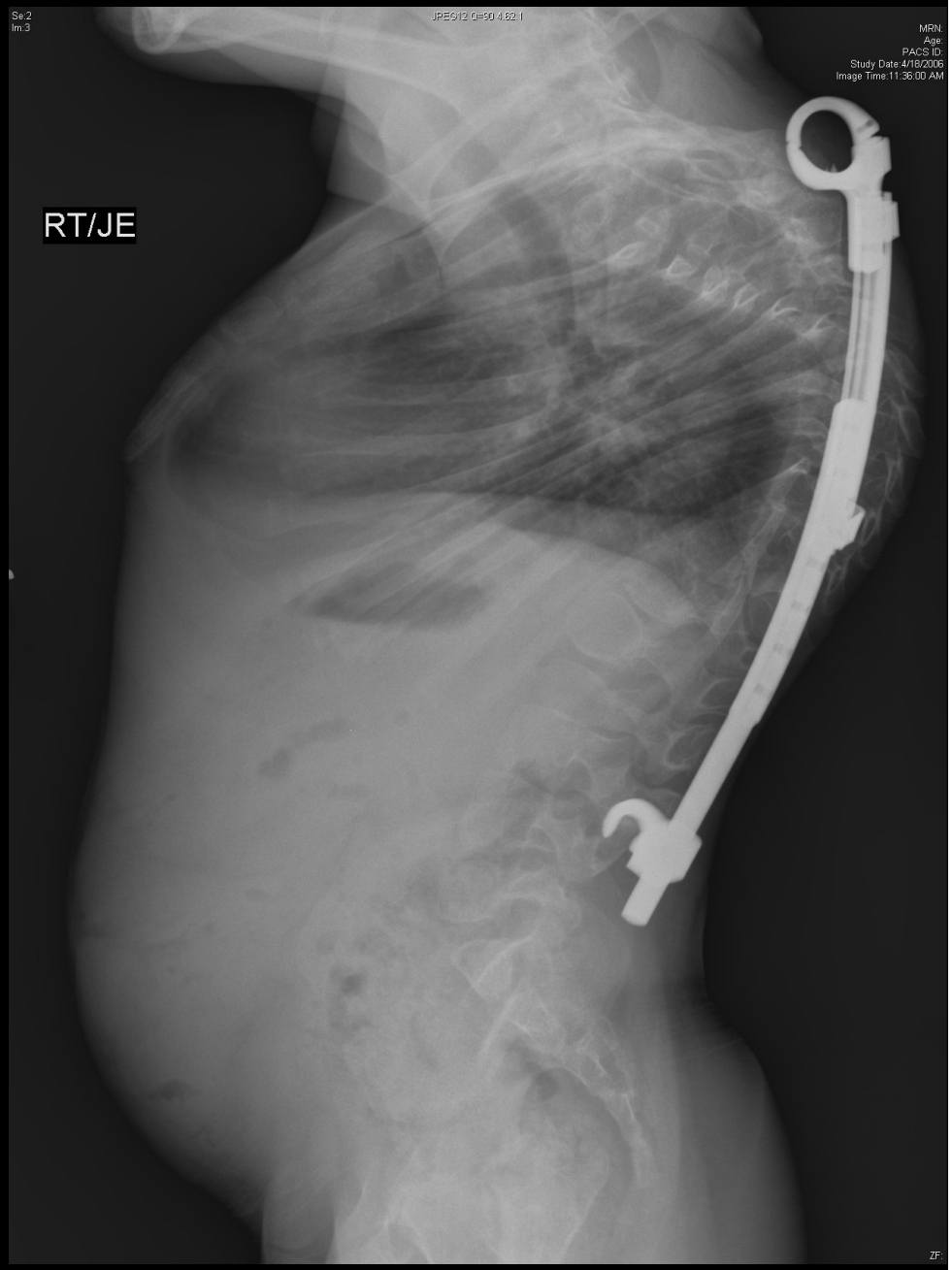
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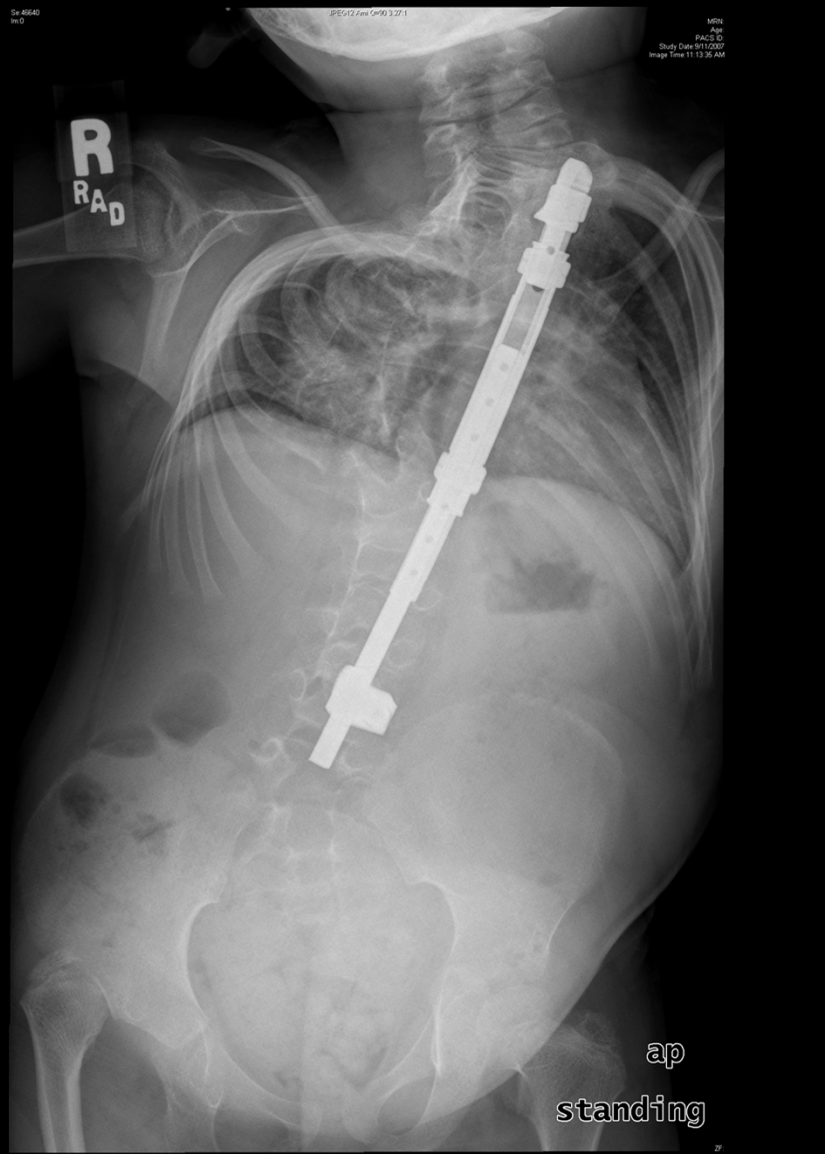
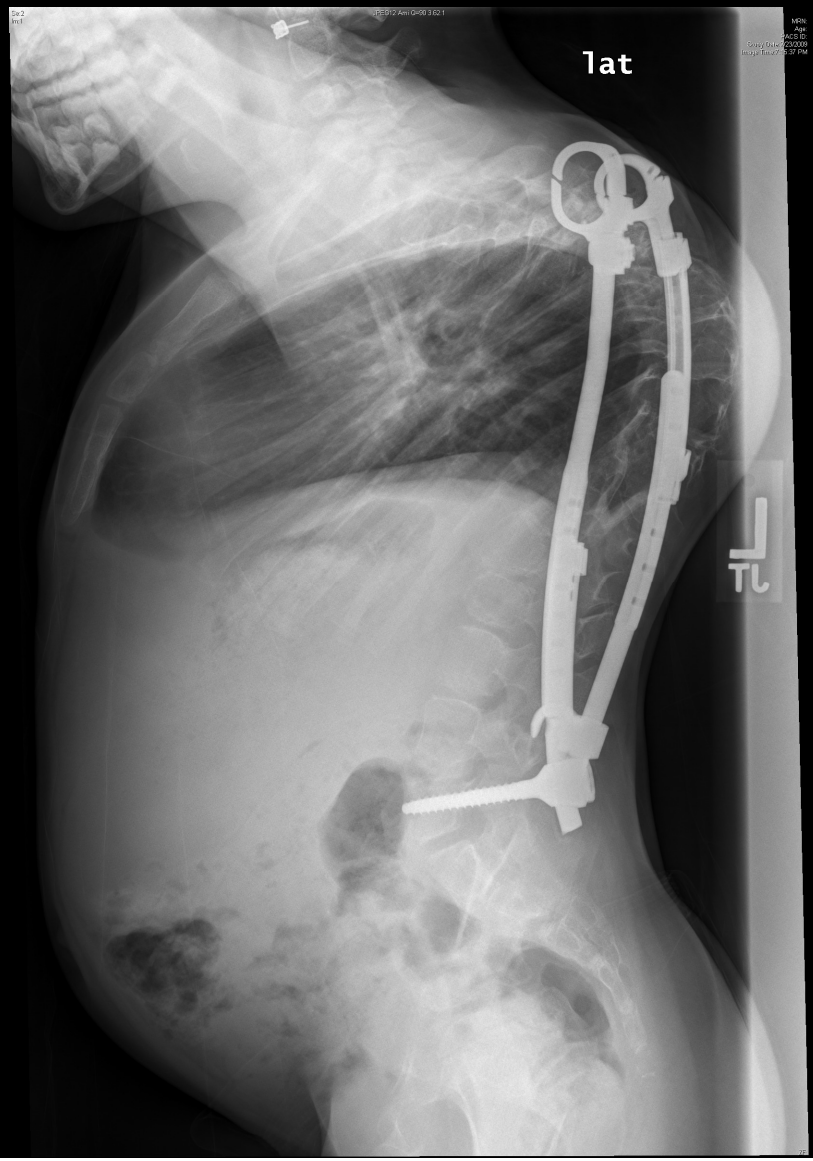
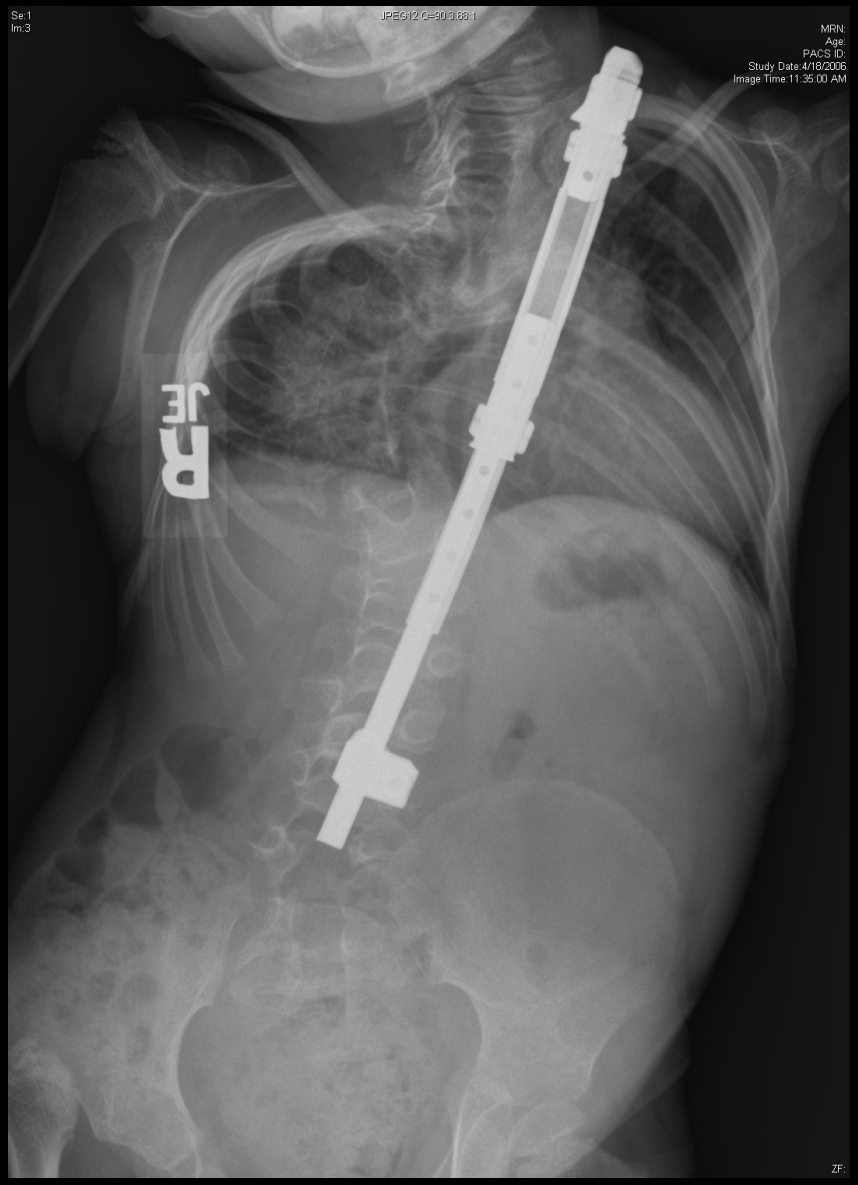
- V/Q 9-05 45:55 R/L

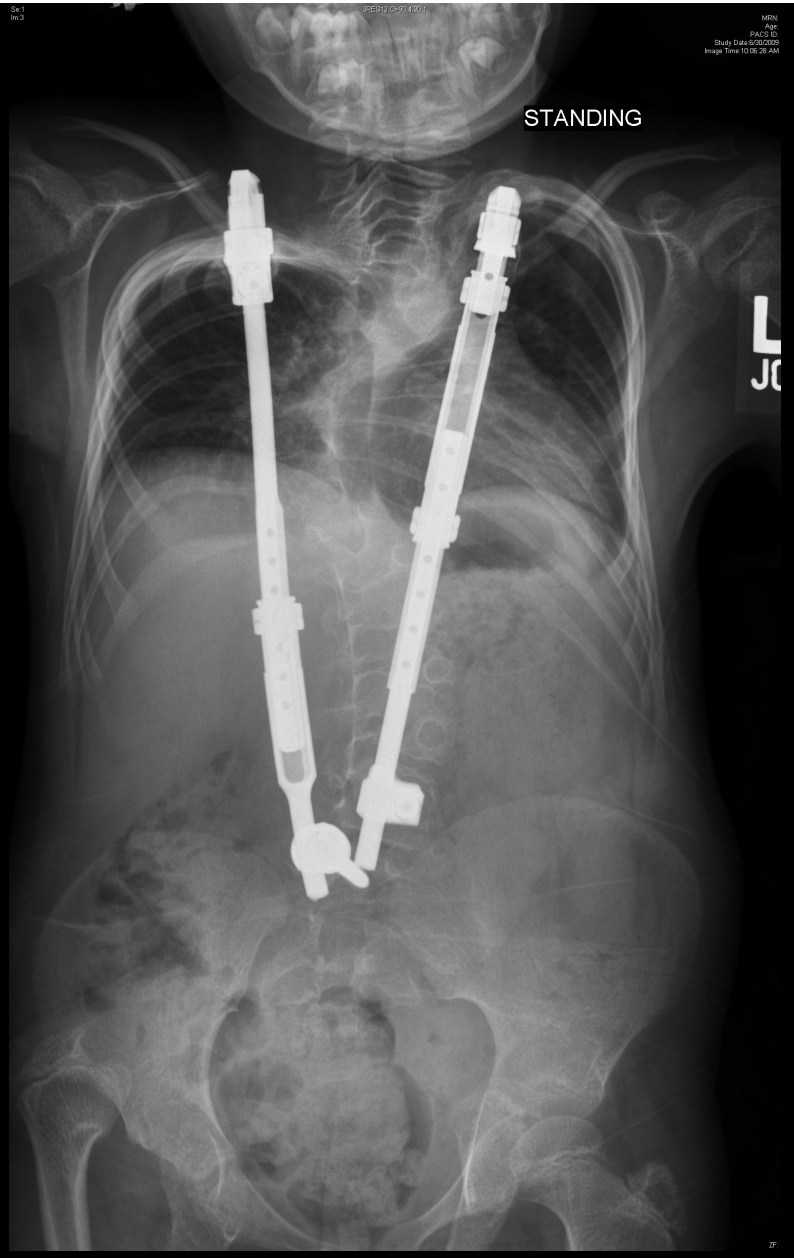


Technique

- Intra-thoracic VEPTR
- Custom lumbar component in lordosis
- 2 expansions - extrathoracic







V/Q 3-09 60:40 R/L

Summary

- TIS stabilization
  - Chest wall reconstruction
  - Extracavitary location
  - Modified or custom implants
- Desired – facile expansion
- Short term revisions/expansions possible
- Long term linkage to definitive solution