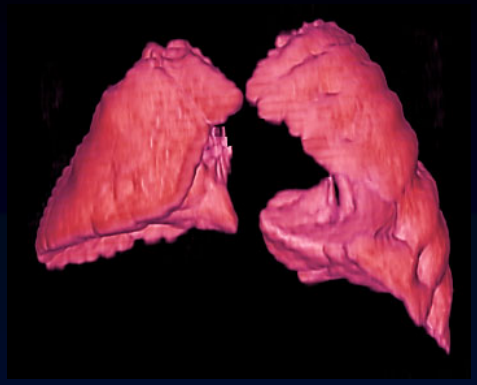

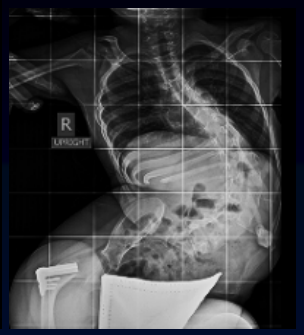

The Children's Hospital *of* Philadelphia®

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Center for Thoracic Insufficiency Syndrome

The Etiology of Thoracic Insufficiency Syndrome in Neuromuscular Scoliosis based on Quantitative Dynamic Lung MRI (QdMRI)

Campbell, RM, Udupa, J*, Flynn, J, Mayer, H, Nance, M, Panitch, H, Wang, Wei-Hsun, Tong, Y*, Wu, I*, Baldwin, K, McDonough, J, Mong, A.

Division of Orthopaedics, Pulmonary, Surgery, Radiology CHOP, *Medical Image Processing Group, Dept. Radiology, University of Pennsylvania

Disclosures RMC

- Grant Support
  - National Organization of Rare Disorders (NORD)
- NORD Medical Advisory Committee member
- Spinal Consultant to the FDA
- Advocate for inventors/companies trying to develop safe and effective devices for children

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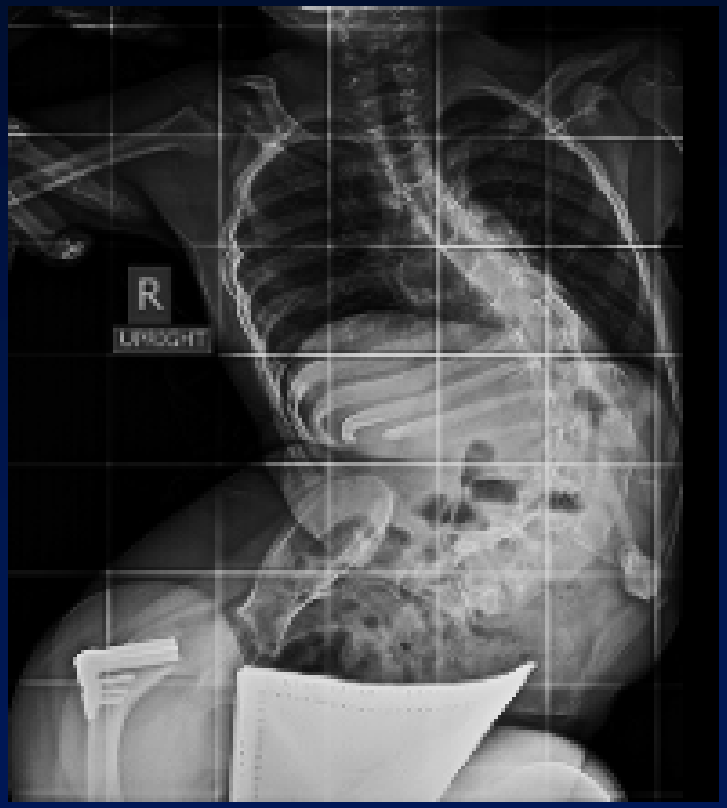
Thoracic Insufficiency Syndrome in neuromuscular scoliosis

- The thorax fails to support normal respiration or lung growth
  - Because of muscle weakness
  - Because of increased thoracic disability from deformity



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Retrospective review
5 NM scoliosis pts

- Avg age 7.9 yrs,
- Avg f/u 2.13 yrs
- Scoliosis 75˚ preop,
  45˚ f/u.
- SAL .84 preop,
  .96 at f/u.
- Pelvic obliquity 19.6˚
  f/u 13.6˚
- Complications: 2 rib cradle migrations

- Eifel Tower VEPTR constructs

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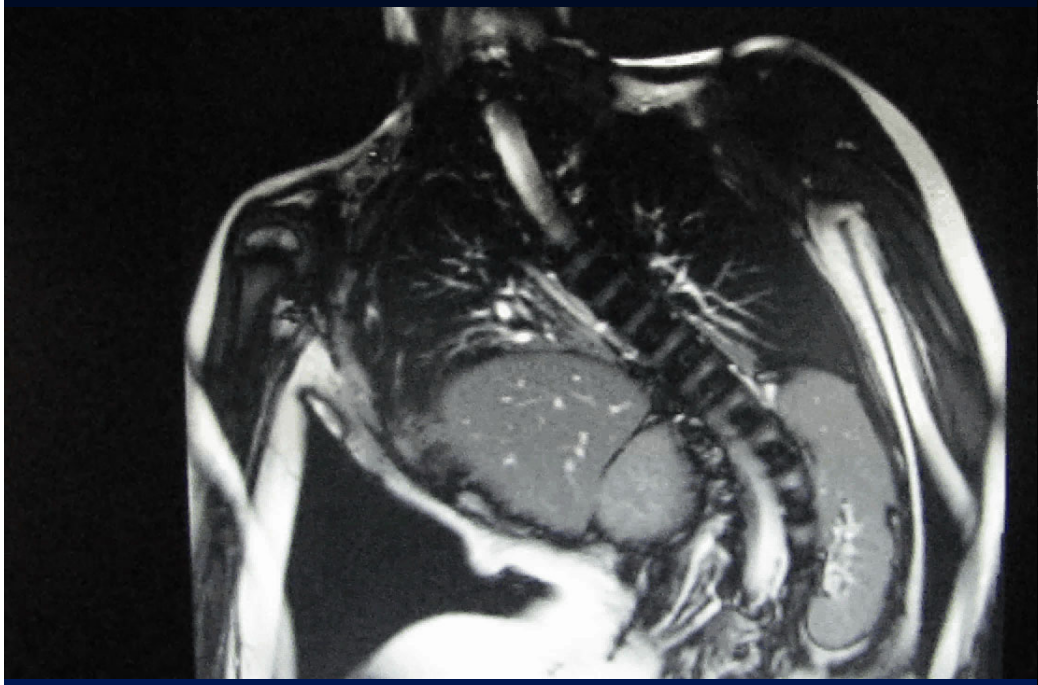
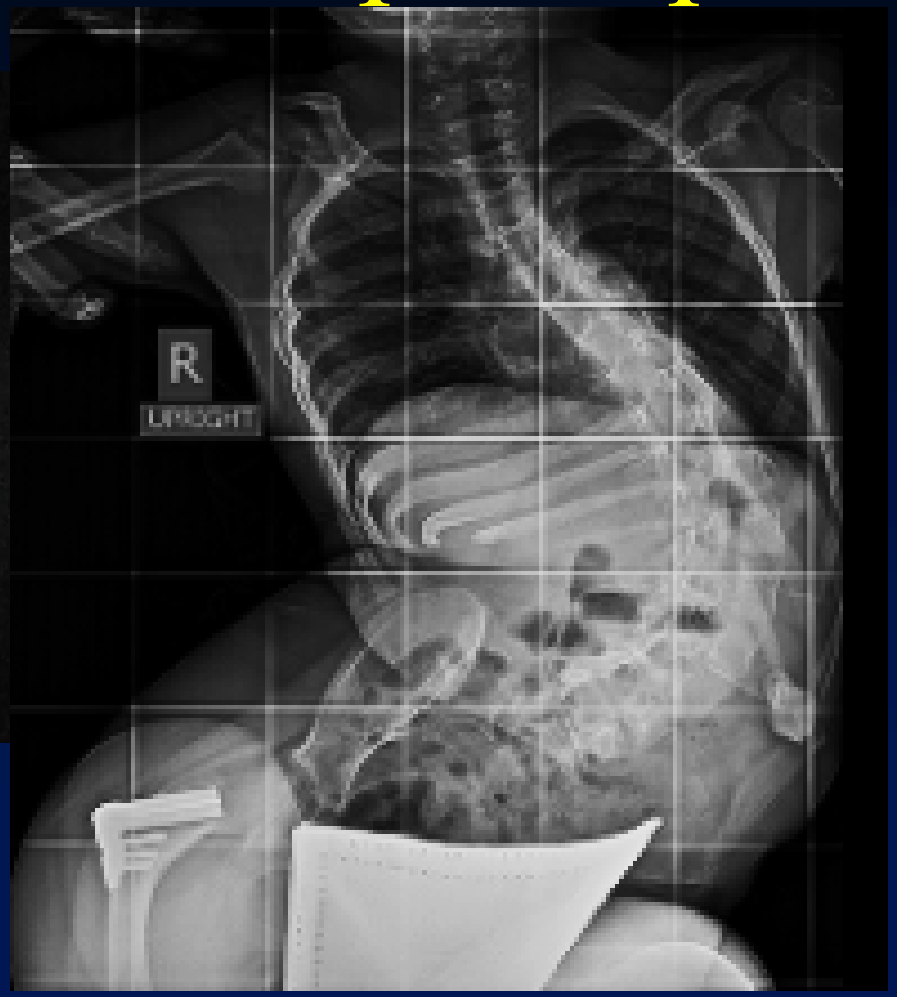
Dynamic Lung MRI
2 preop only, 3 pre and post op





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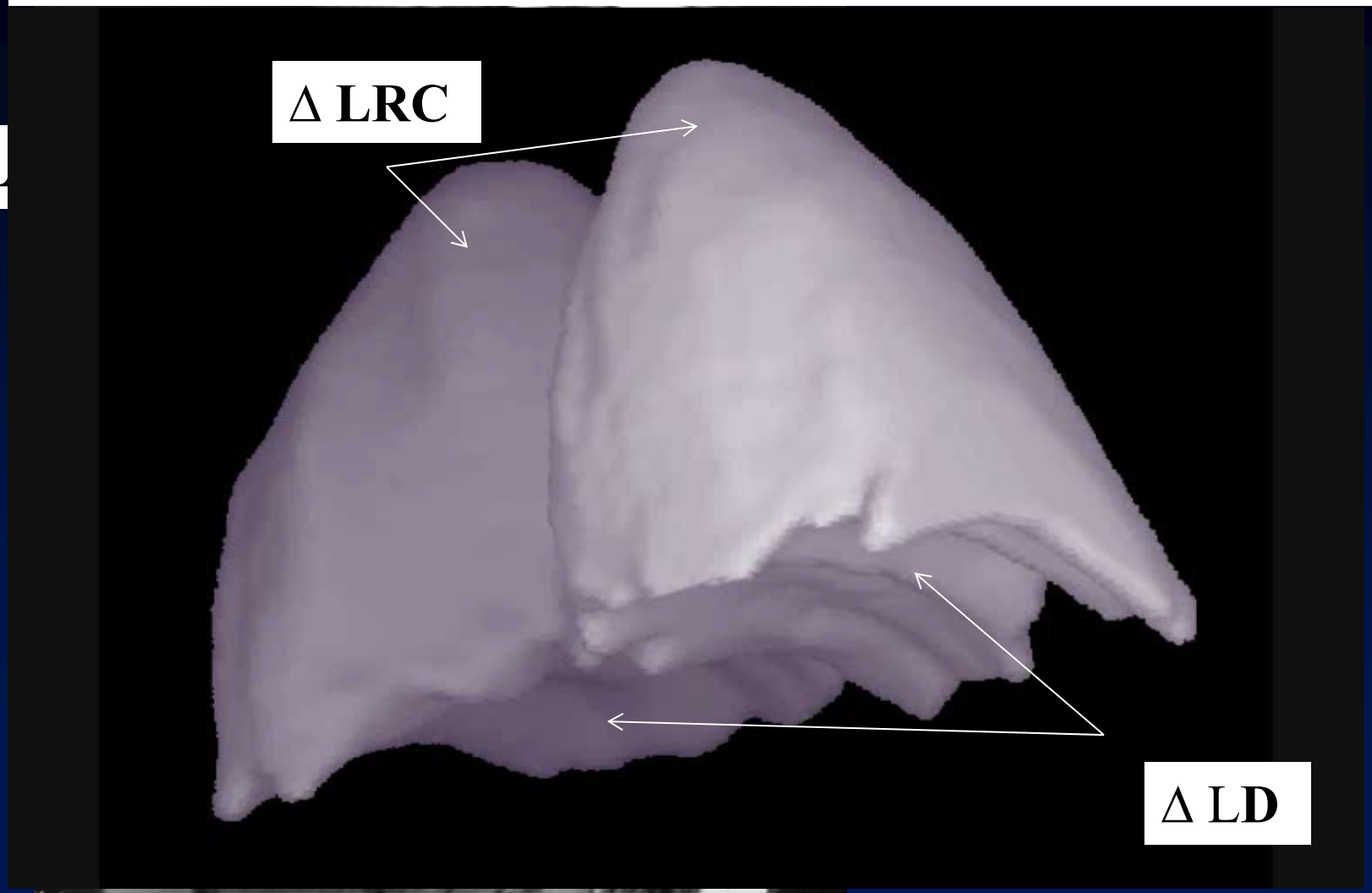
4-D Quantitative Dynamic Lung MRI (QdMRI)



Δ L

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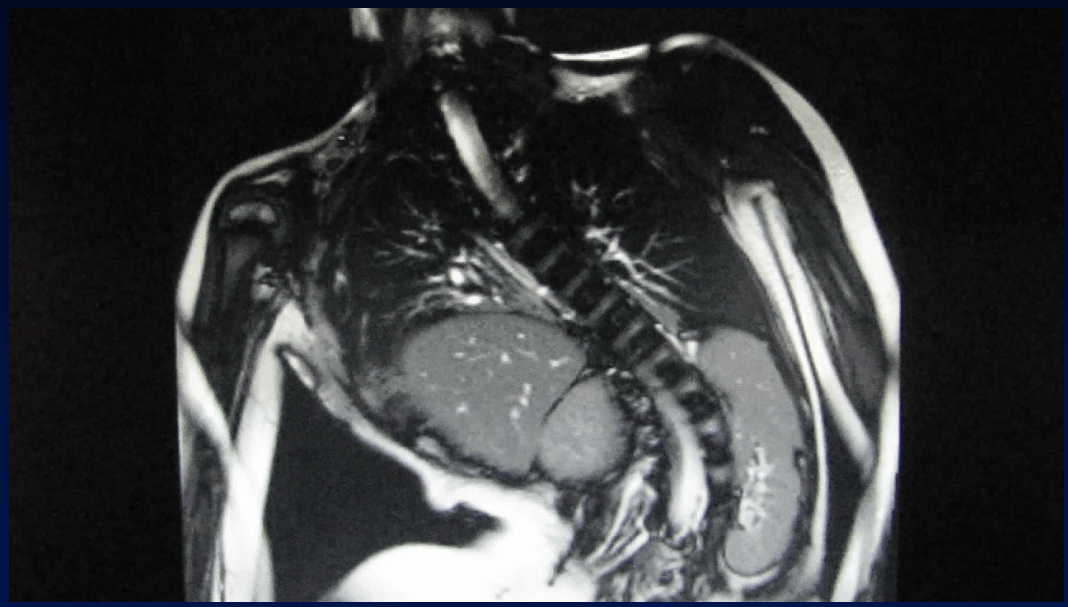
Hypothesis

- The concave side would be more obstructed with less change in volume with respiration
  - Renal motion more obstructed
- The convex side (rib hump)would have less rib cage volume changes with respiration



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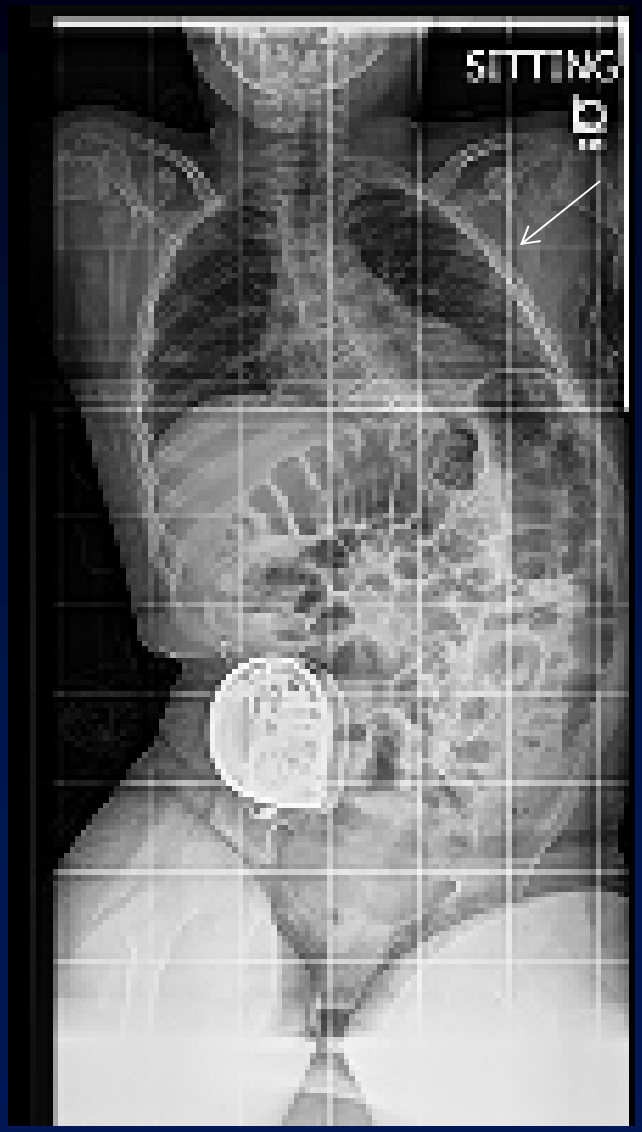
Preop QdMRI (5 pts)

- Concave side
  - Δ rib cage volume 46.5 cc
  - Δ diaphragm volume 25.7 cc
- Convex side
  - Δ rib cage volume 40.5 cc
  - Δ diaphragm volume 27.4



CTIS

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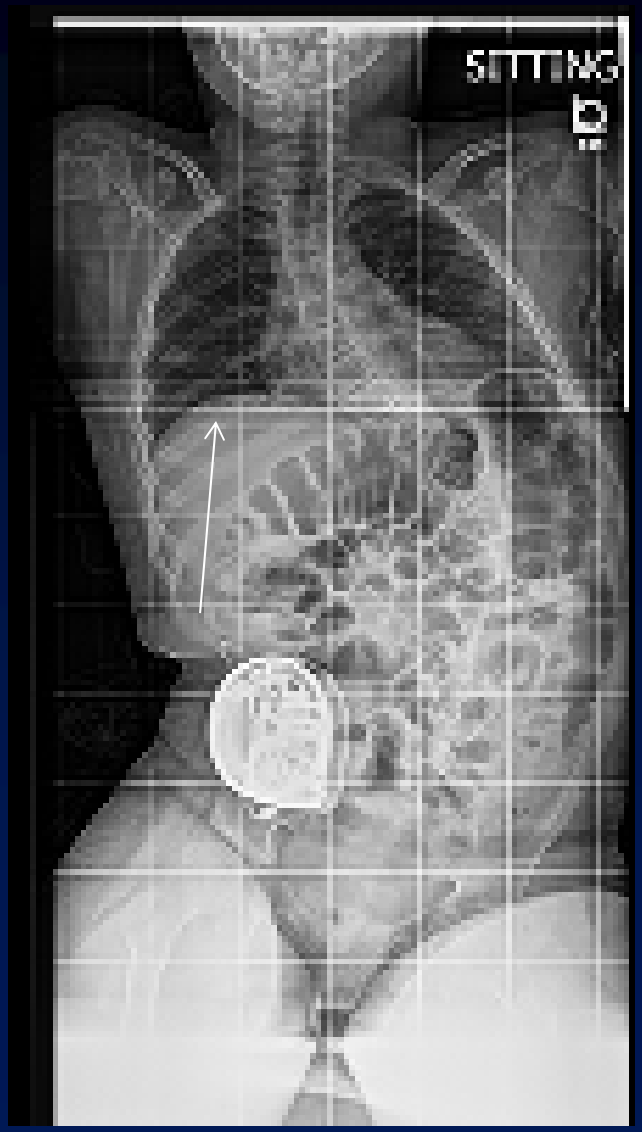

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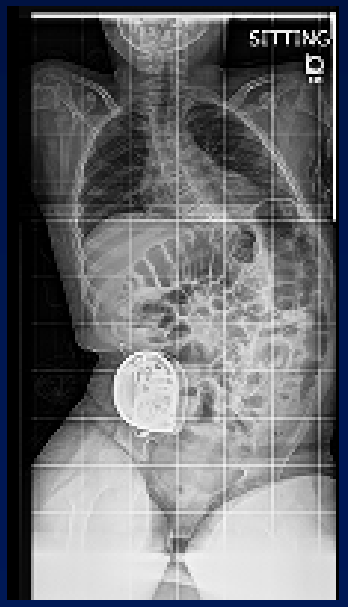
Preop QdMRI
Concave Hemi-thorax
Diaphragm volumes

- 2 pts
  - concave Δ diaphragm volume < convex
- 1 pt
  - concave > convex
- 2 pts
  - had equal volumes.



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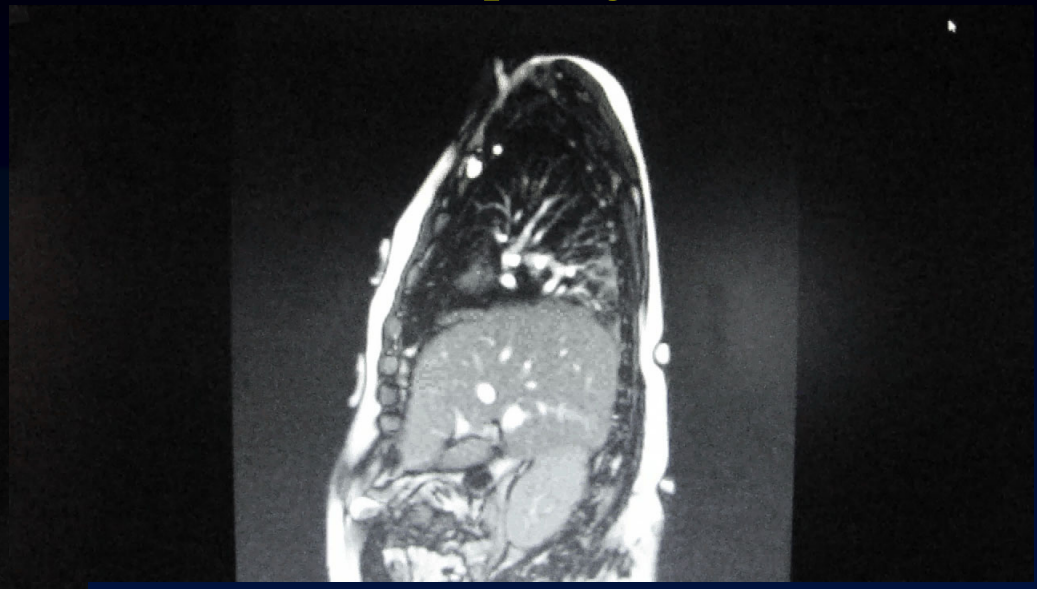
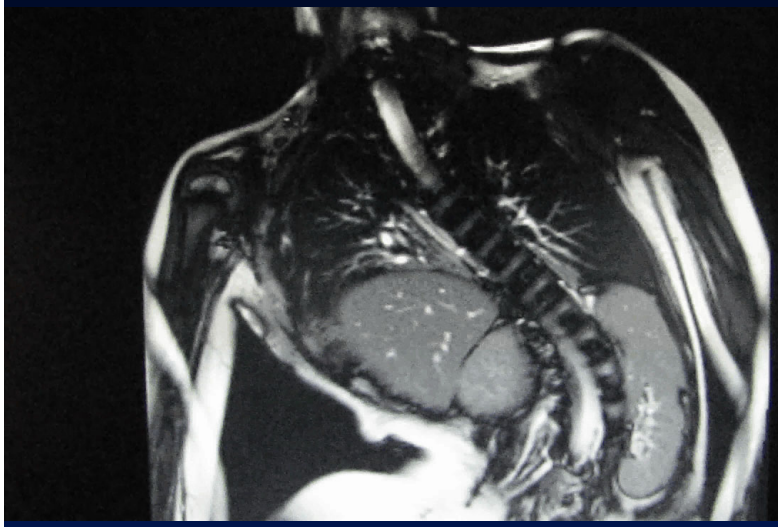
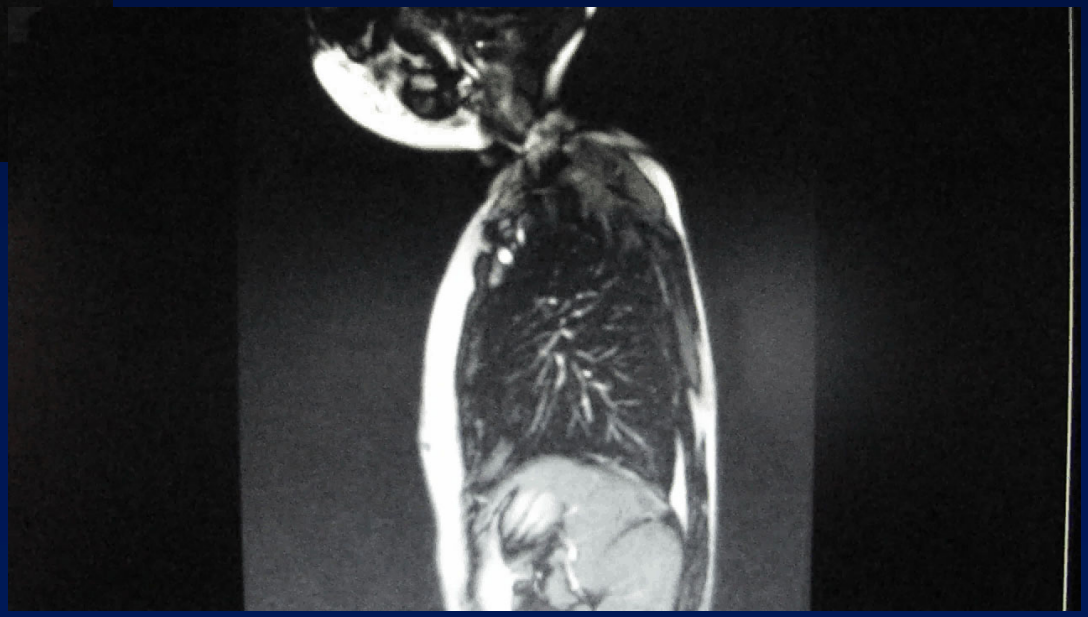
Concave Diaphragm 39.2 cc





Convex Diaphragm 20.1 cc



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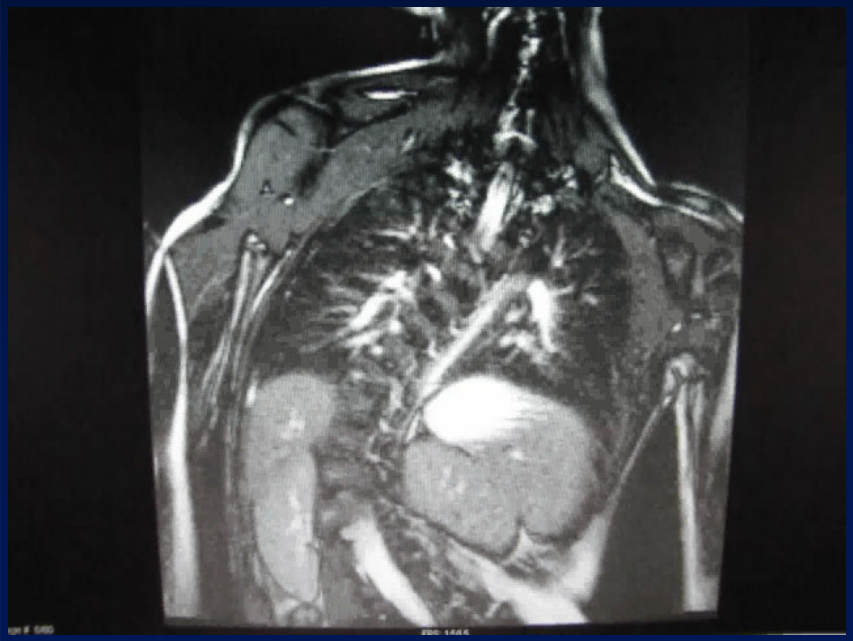
Pre-op Renal Excursion

Kidney excursion

- Concave was 2.58 mm
- Convex 3.02 mm.



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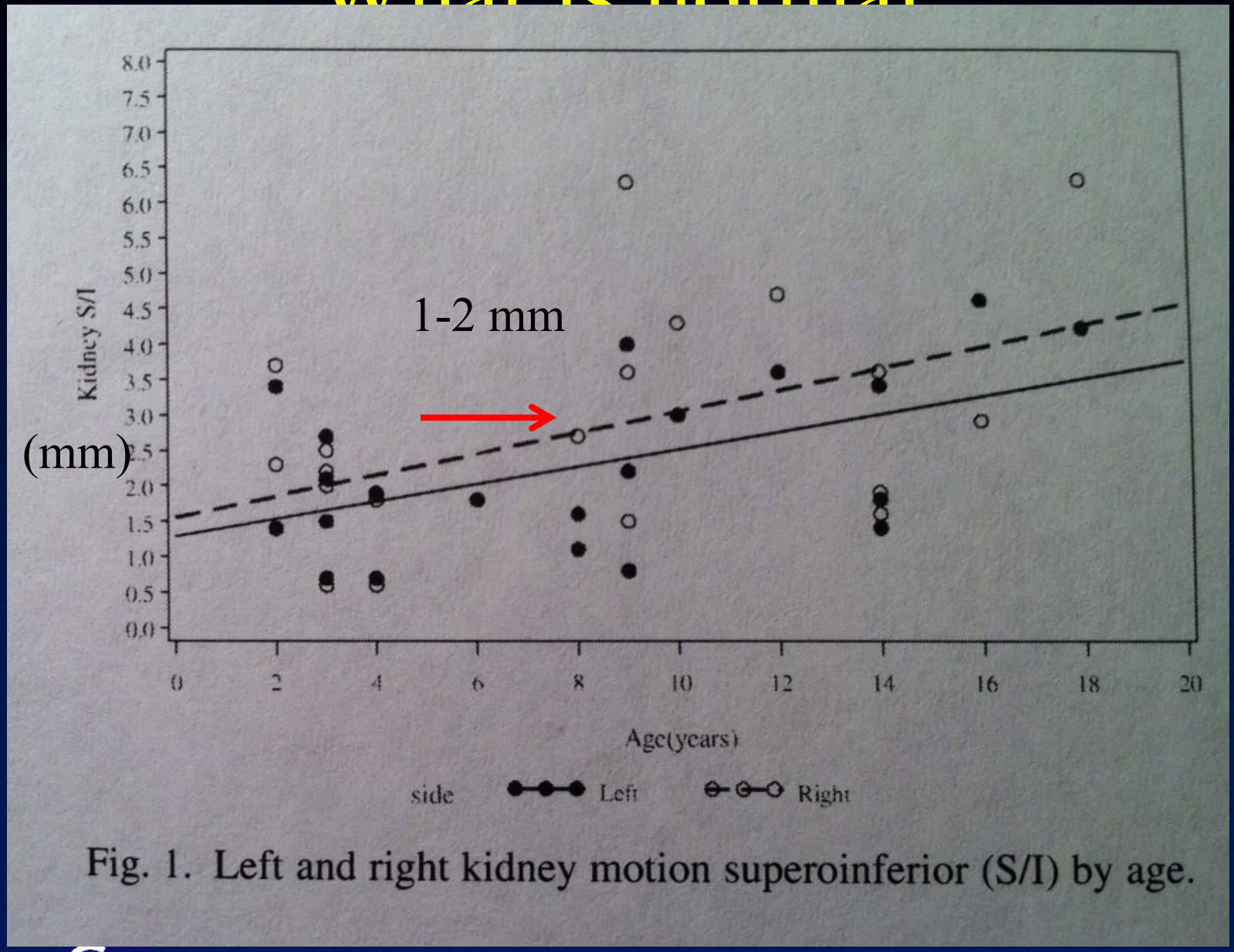
What is normal



1-2 mm

(mm)

Fig. 1. Left and right kidney motion superoinferior (S/I) by age.

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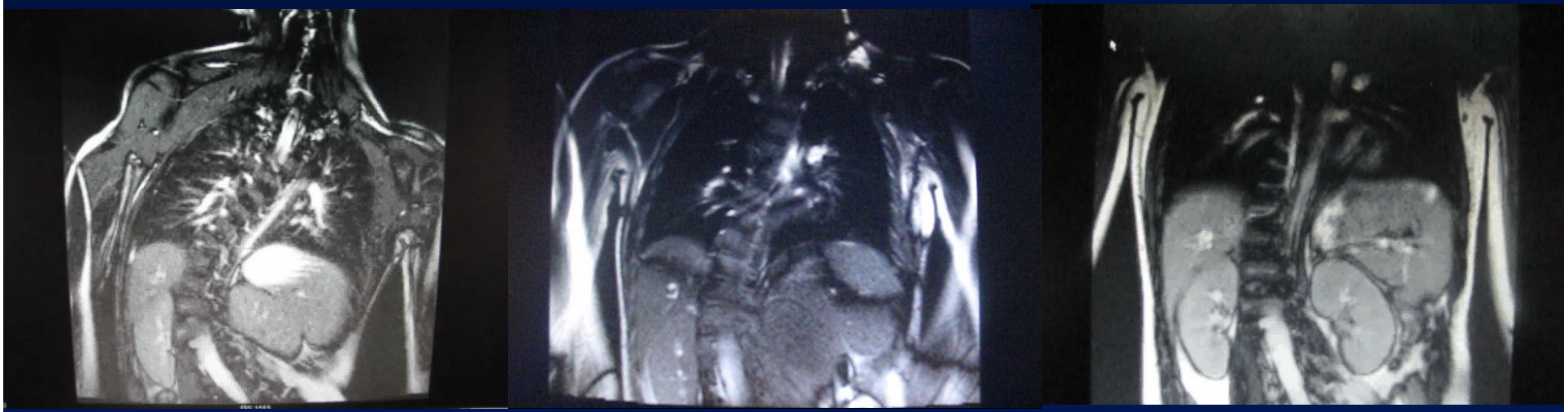
Post op Renal Excursion

- Concave kidney excursion increased 6.3mm (320%)
- Convex 5.2mm (192%).



1.2 mm

13.1mm

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3 pts postop QdMRI scans

Concave side

- Δ rib cage volume increased 57%
- Δ diaphragm volume increased 128%

Convex side

- Δ rib cage volume increased 72%
- Δ diaphragm volume increased 109 %

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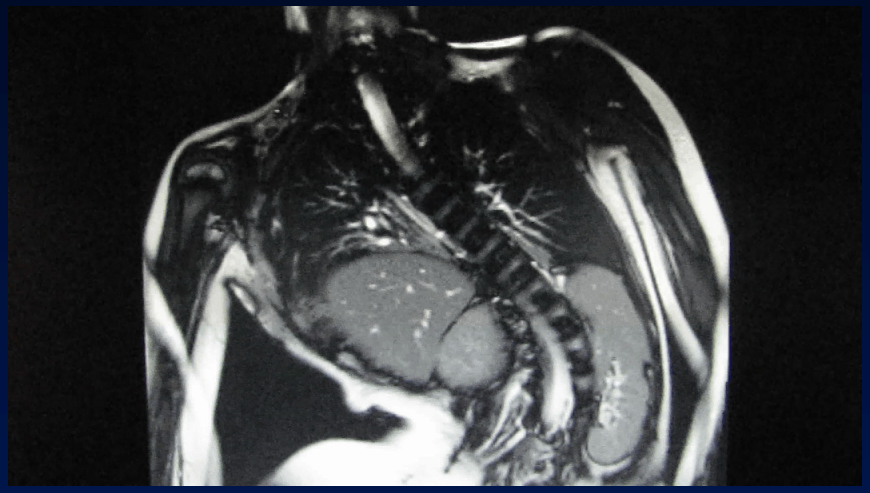
Summary

- QdMRI can accurately assess thoracic performance parameters such as unilateral diaphragm/rib cage lung volume changes
- It has great potential to increase our understanding of these diseases



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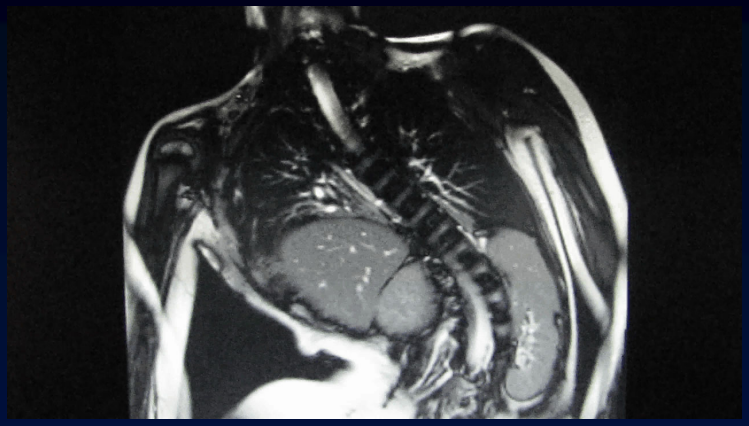
Summary

- VEPTR treatment appears to increase concave hemi-diaphragm and convex rib hump hemi-thorax performance in TIS due to neuromuscular scoliosis
  - But, this needs much more study



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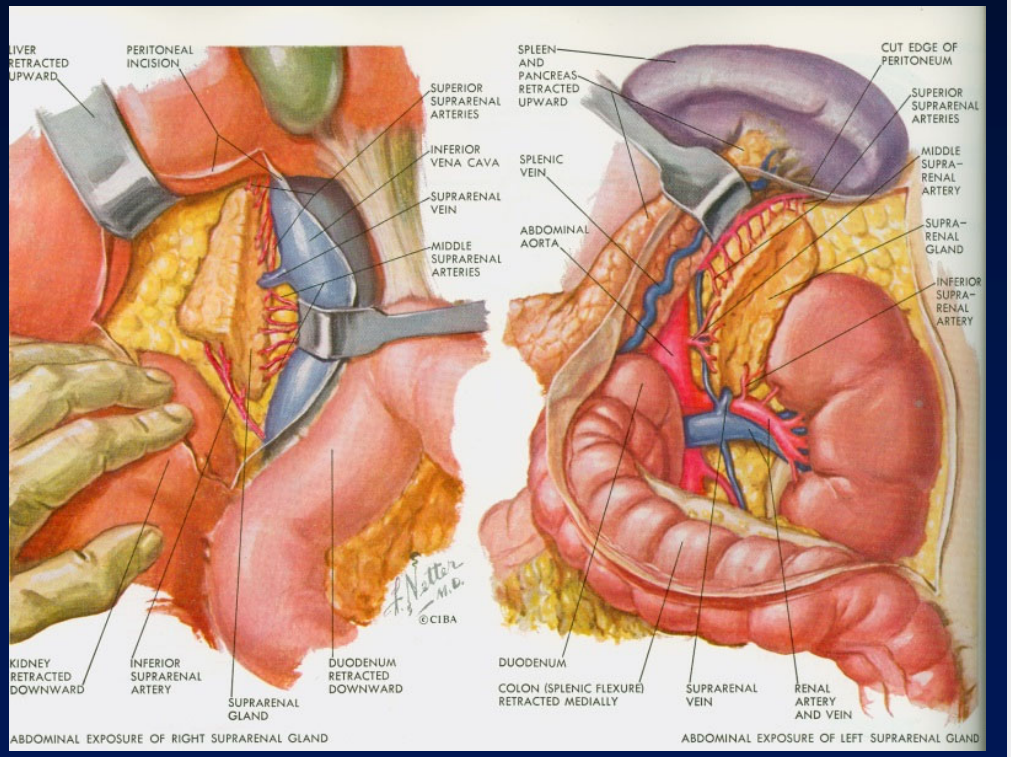
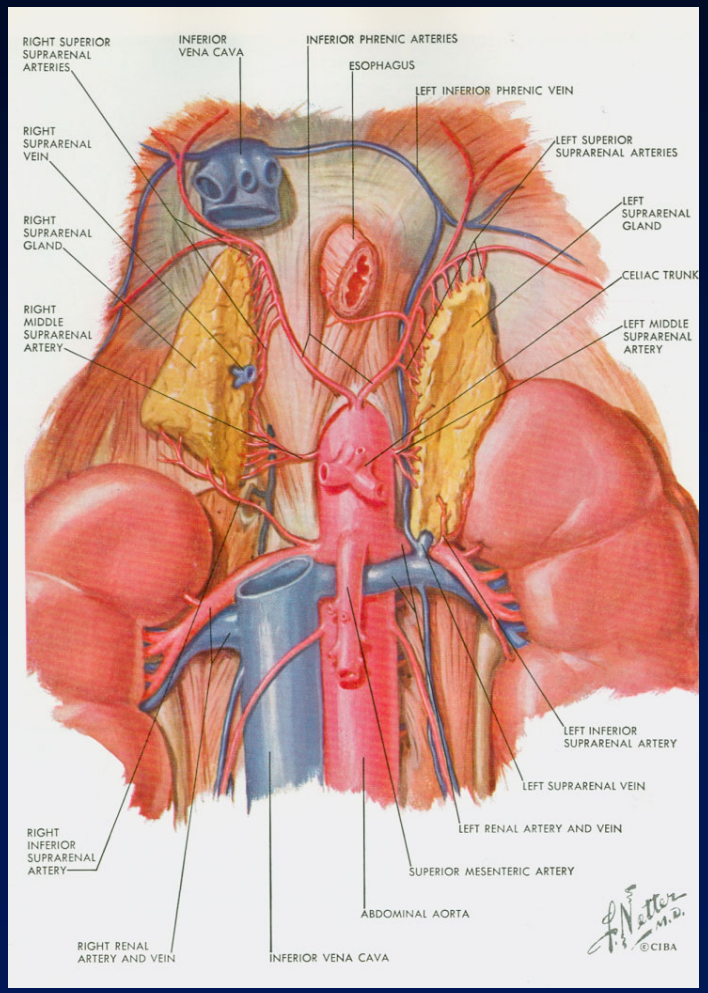
The Future Breakthroughs in Treatment

- To really advance surgical treatment of spine deformity in children, we have to better understand the dynamic biomechanical component of these diseases
  - X rays can't help us much
- We have the technology to do this
    - But we have a long way to go

CTIS

The Kidney Pedicle and other structures probably play a complicated role in the pathologic biomechanics of TIS





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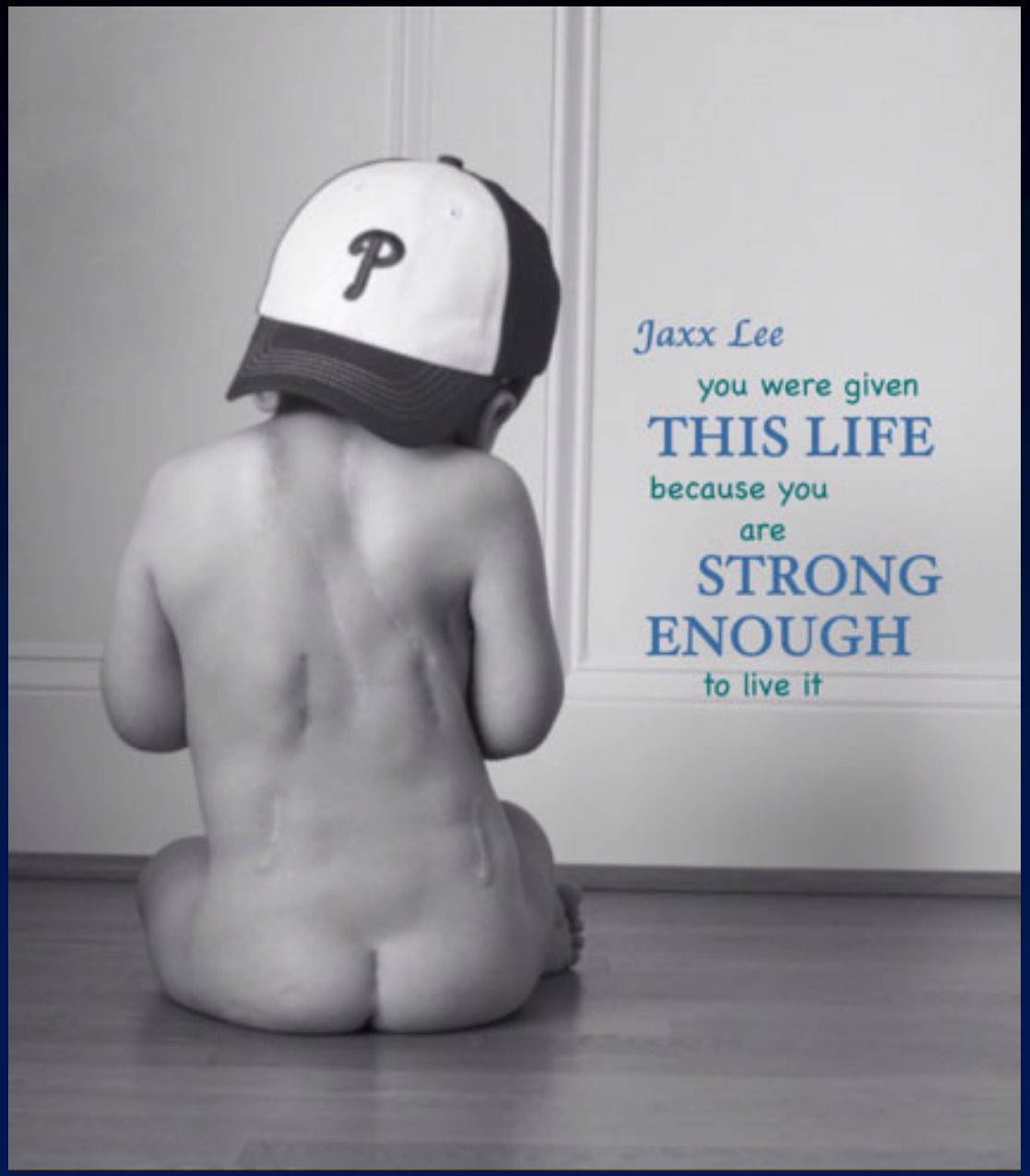
Thank You



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