The Characterization of the Thoracic Biomechanics of Respiration in Thoracic Insufficiency Syndrome by Dynamic Lung MRI: A Preliminary Report

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Disclosures

- RM Campbell
  - Grant Support
    - NORD, FDA Office of Orphan Product Development
  - Royalites
    - Synthes Spine Co
- JW Simmons
  - Honorarium
    - Synthes Spine Co

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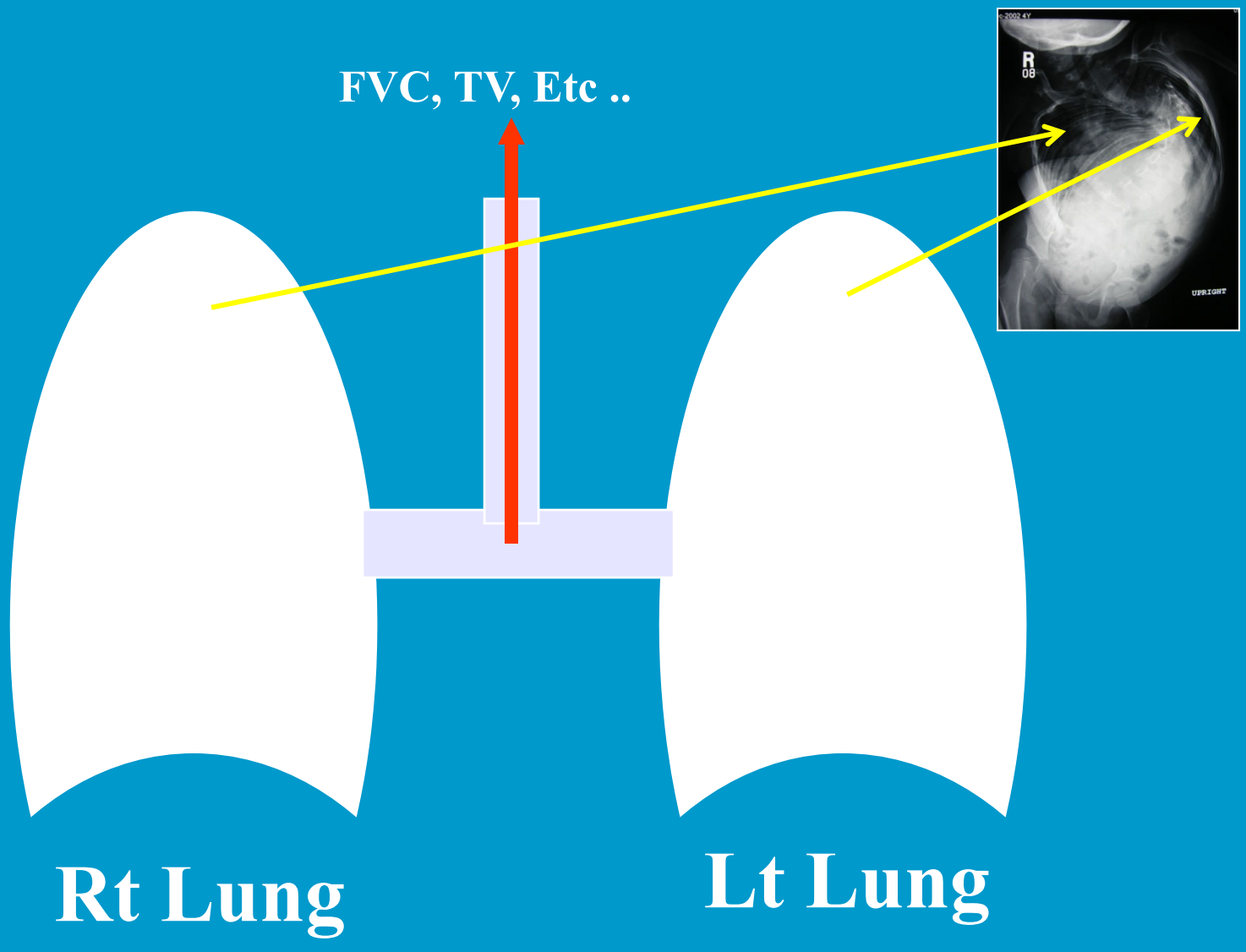
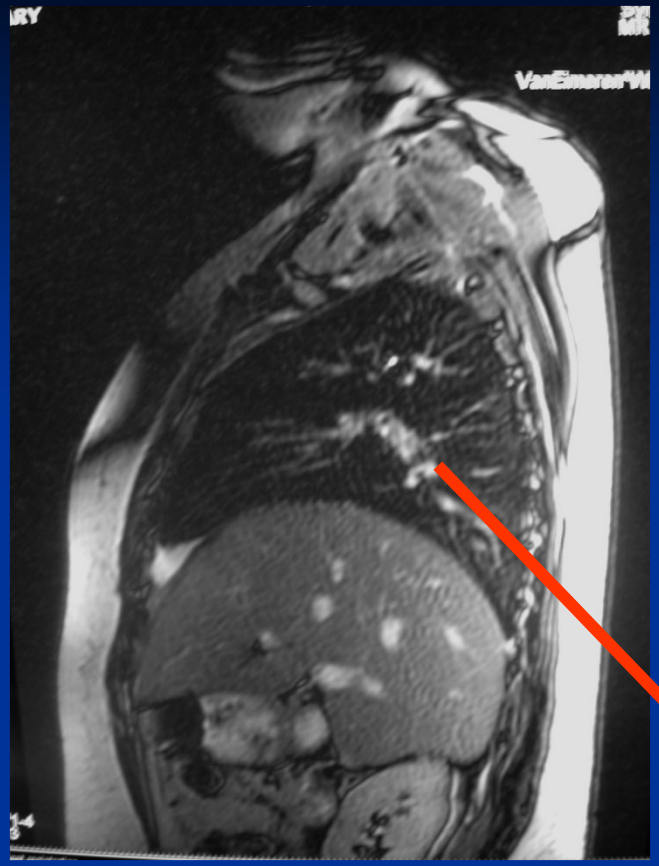


PFT's are Lacking Biomechanically

Dynamic Lung MRI and Spine Deformity



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Methods

- 20 pts
  - evaluated for TIS, studied with radiographs, CT scans, PFT's (when practical), and dynamic lung MRIs performed with spontaneous breathing (tidal volume)
  - 6 had EOS
  - 7 had congenital scoliosis
  - 4 had kyphoscoliosis
  - 3 hypoplastic thorax.

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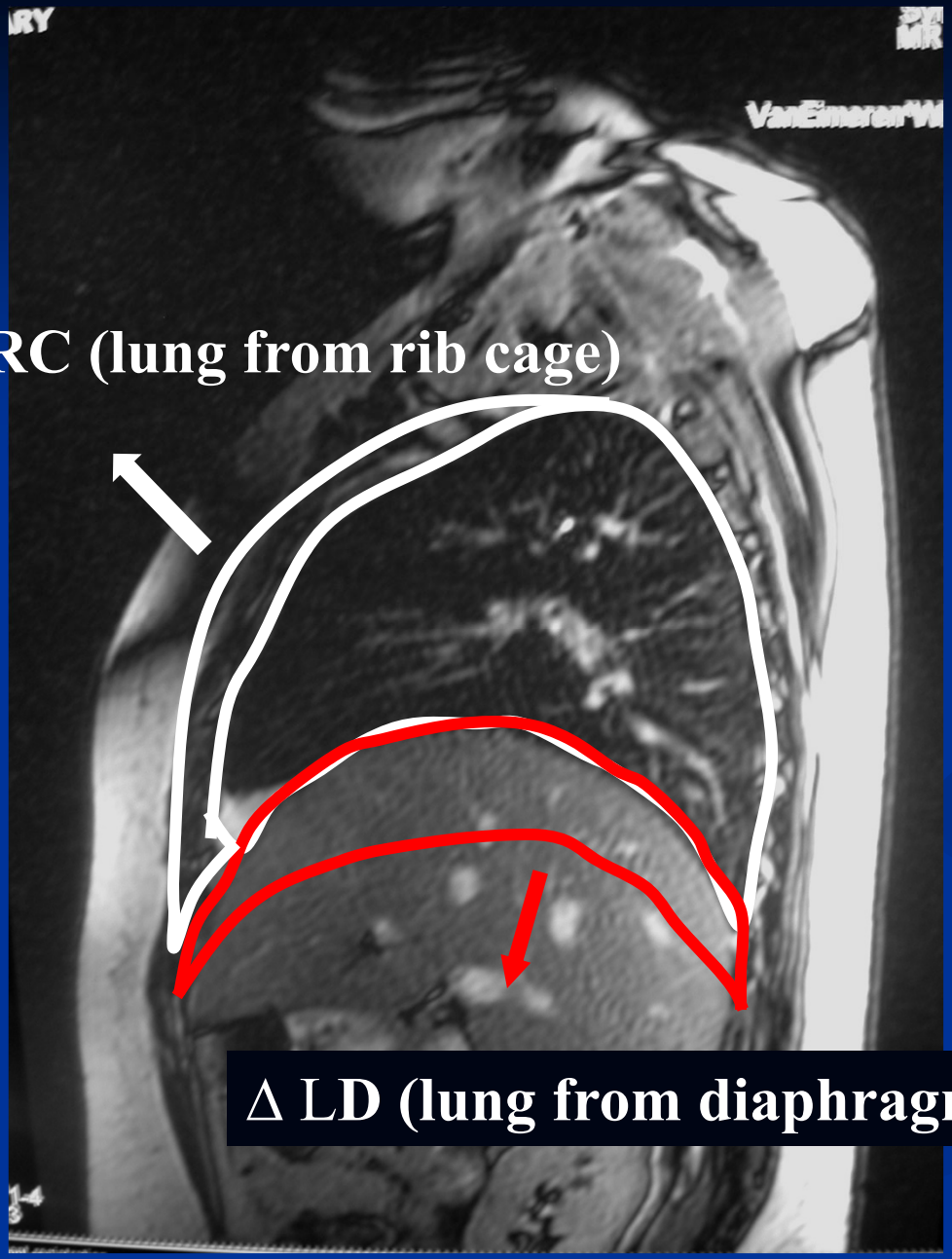
Δ LRC (lung from rib cage)



Δ LD (lung from diaphragm)

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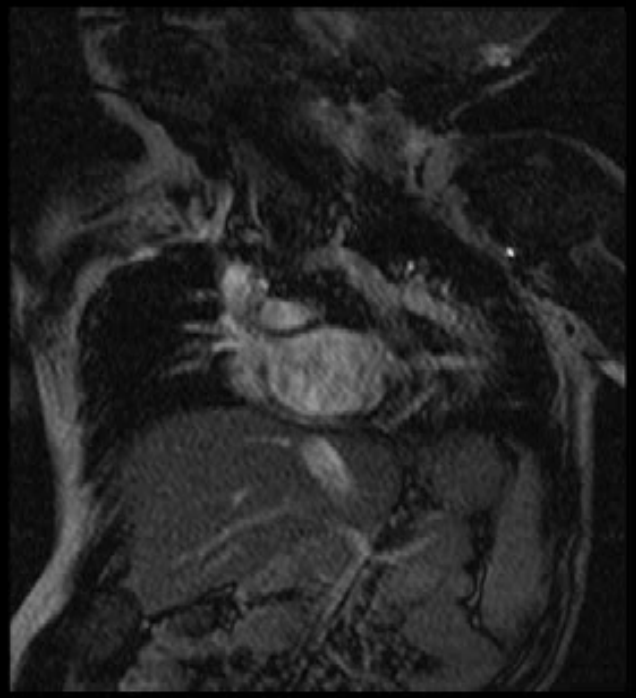
Positive Marionette sign

Patient

Normal 4 y/o



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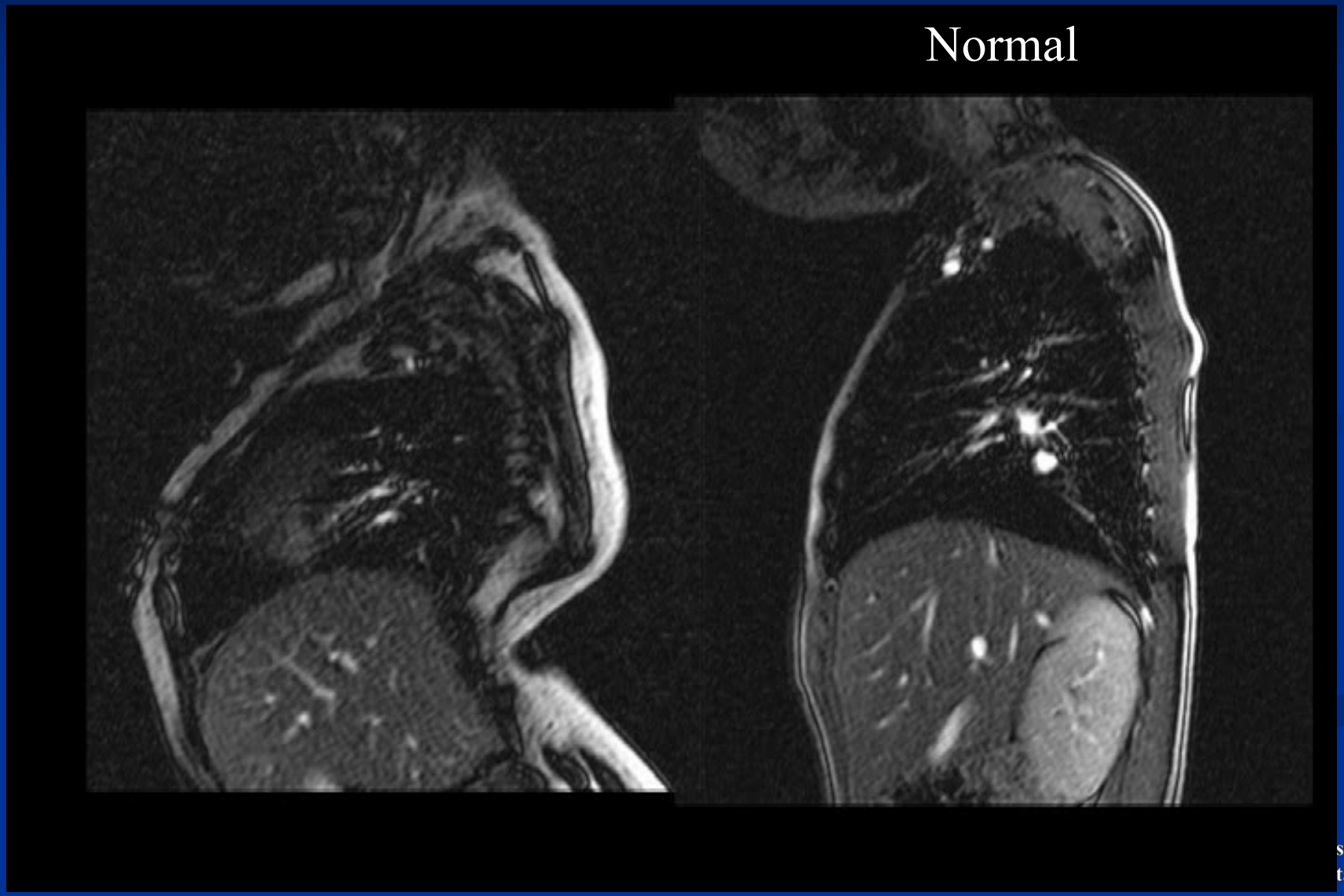
TL 58 s.c. **Δ L RC** 6.3 (11%)
Δ L D 5.2 (9%)

TL 89 s.c. **Δ L RC** 5.3 (6%)
Δ L D 6.9 (8%)



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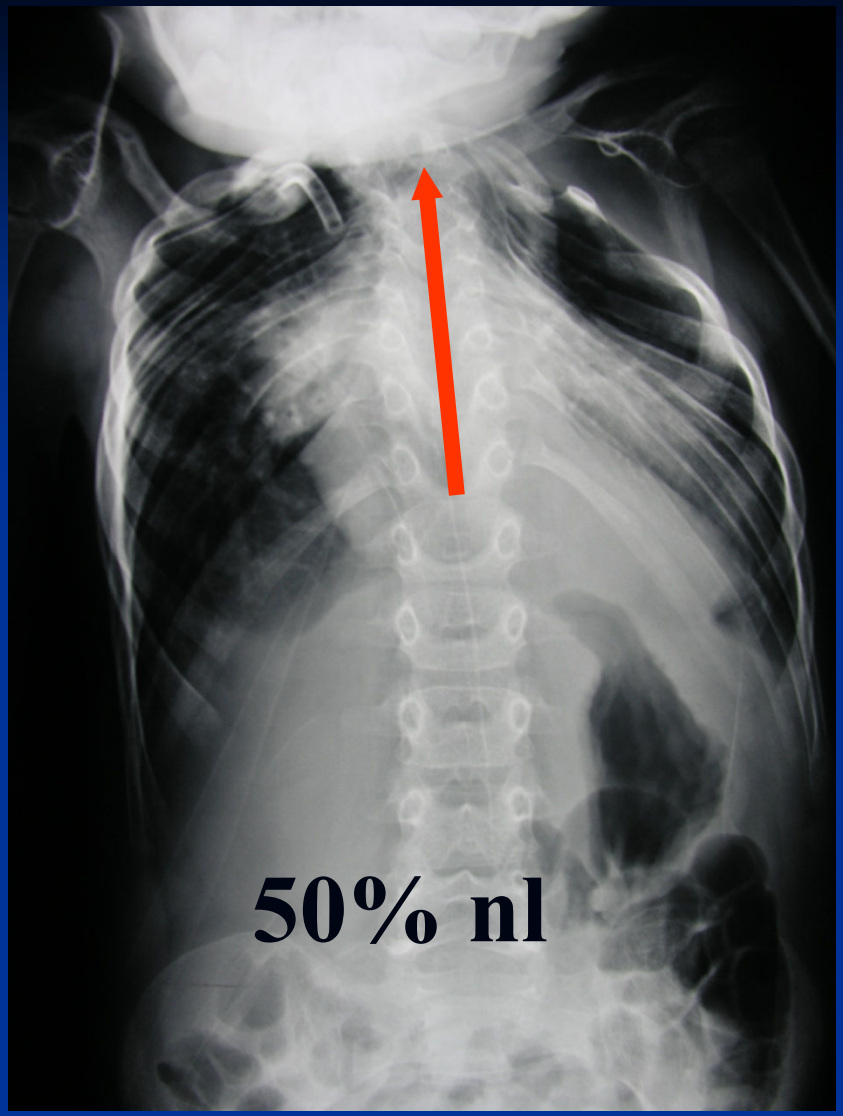

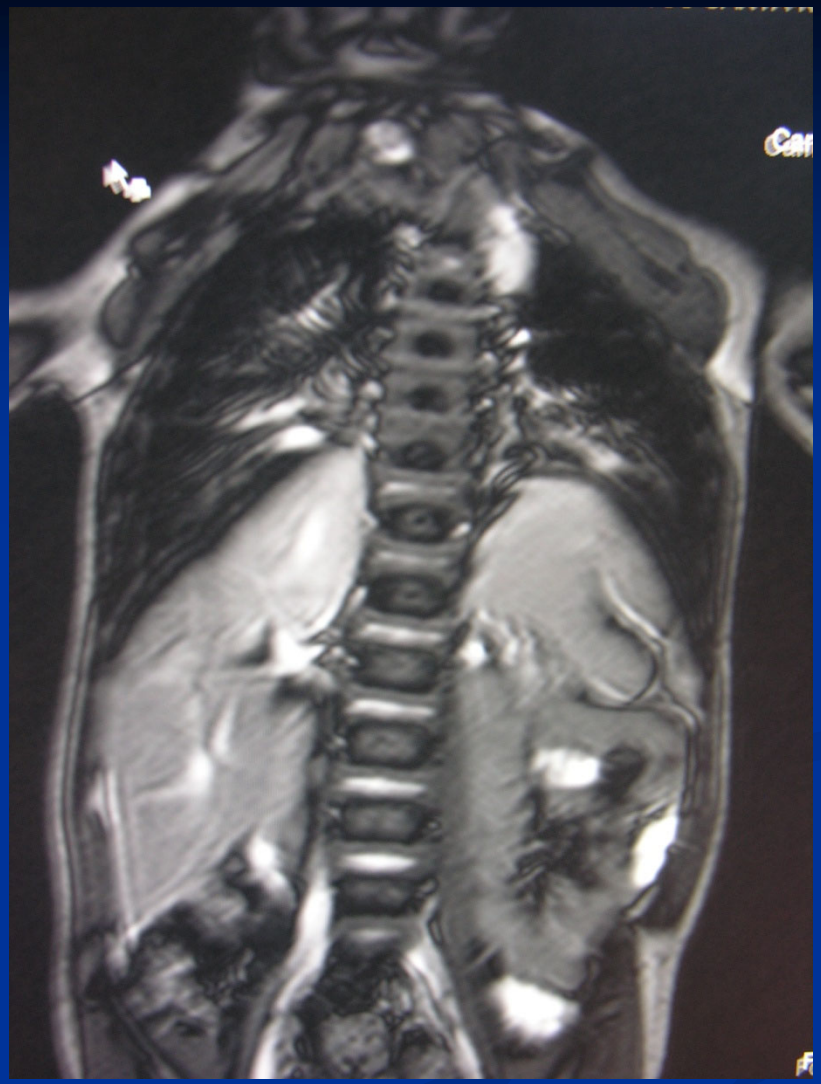

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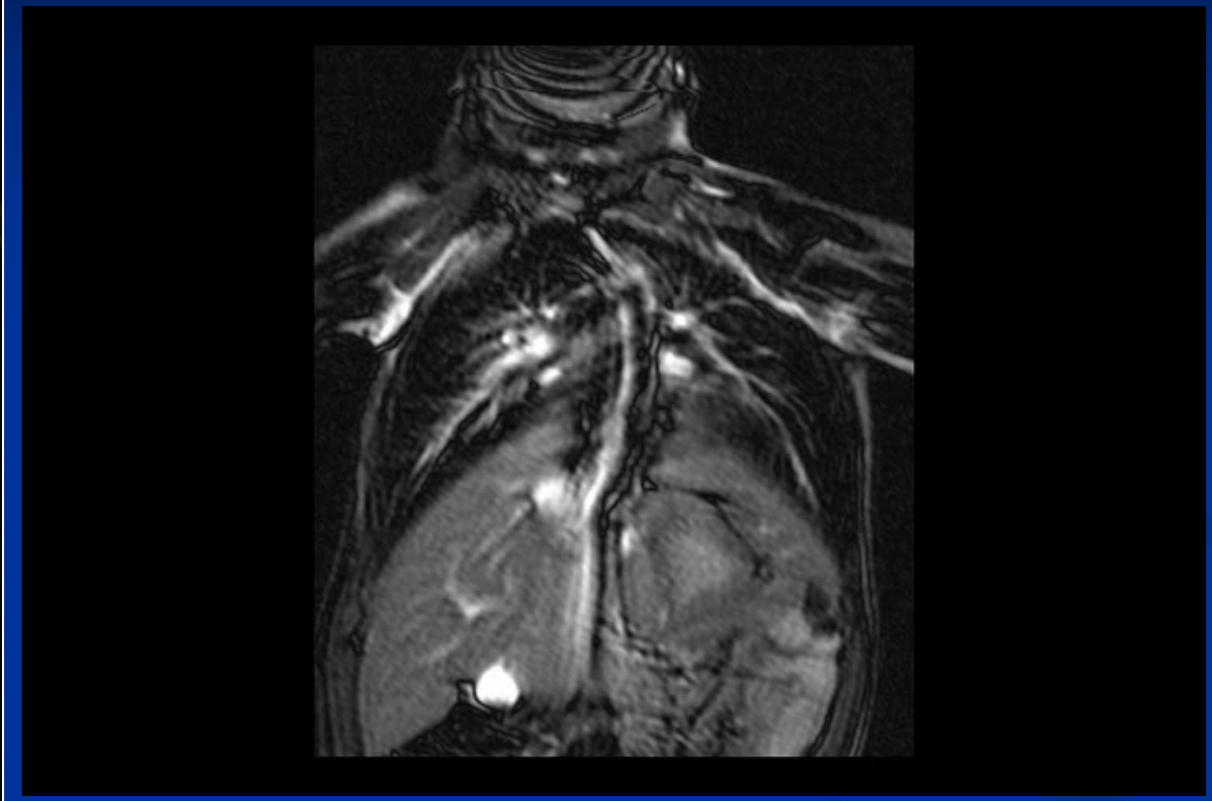

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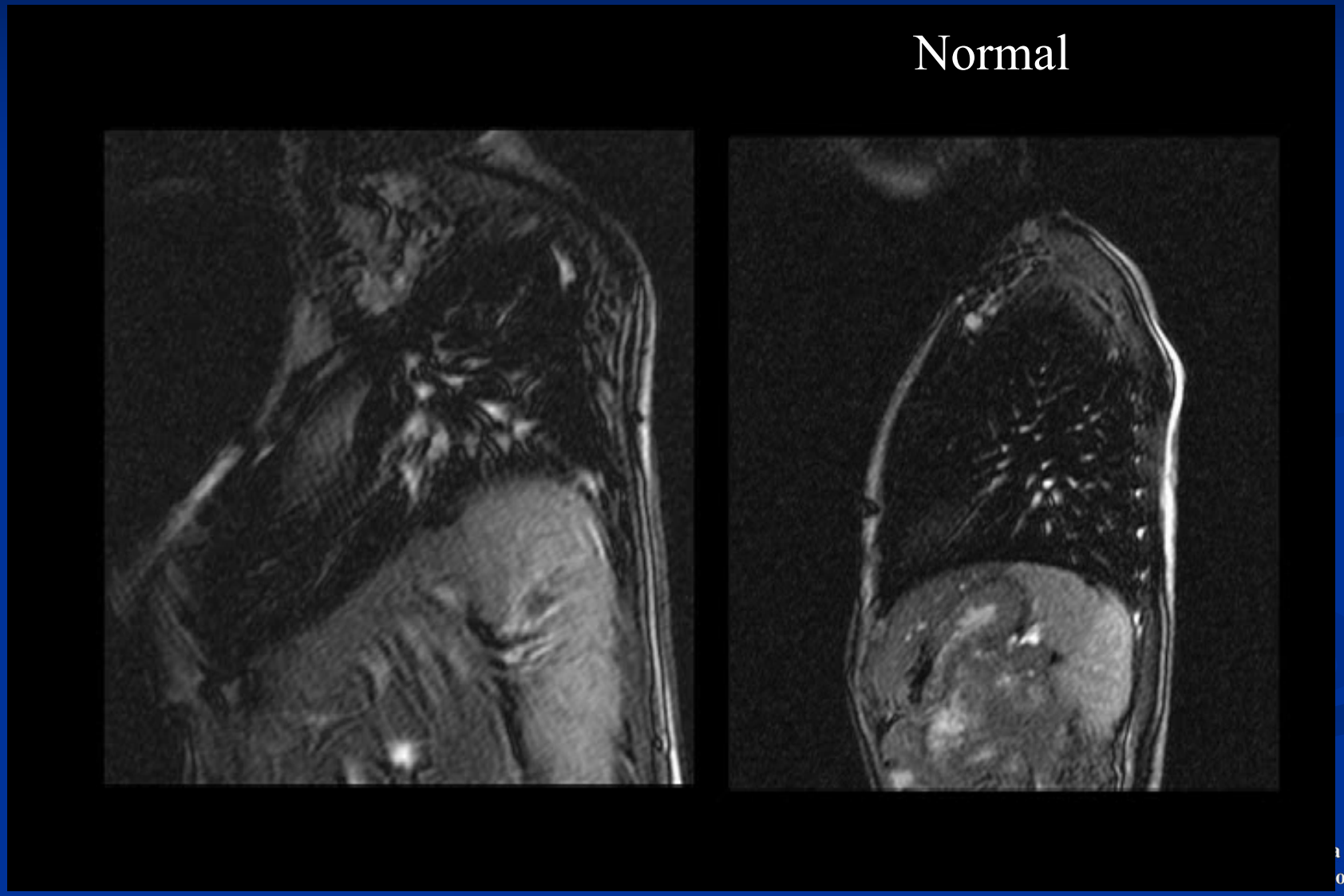
TL 74 s.c. Δ **L RC** 0.37 (0.4%)
(7 y/o) Δ **L D** 8.4 (11%)

TL 89 s.c. Δ **L RC** 5.3 (6%)
(4 y/o) Δ **L D** 6.9 (8%)



Results

	Δ Lung from Diaphragm % total lung area	Δ L Rib Cage % total lung area
EOS		
Convex hemithorax	8.2 %	1.1 %
Concave hemithorax	8.3 %	2.5 %
CONGENITAL SCOLIOSIS		
Convex hemithorax	12.1 %	2.6 %
Concave hemithorax	12.8%	3.4 %
KYPHOSCOLIOSIS		
Convex hemithorax	6.1 %	1.1 %
Concave hemithorax	7.1%	1 %
HYPOPLASTIC THORAX (hemi-thorax symmetric)	8.9 %	5.4 %

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Conclusions

- The convex hemi-thorax in EOS and congenital scoliosis is less efficient than the concave hemi-thorax in the rib cage expansion contribution to respiration
- Kyphoscoliosis degrades respiratory hemi-thorax expansion symmetrically
- Hypoplastic thorax hemi-thorax expansion seems least affected

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Thank You!

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