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

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
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
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
Dynamic Lung MRI
2 preop only, 3 pre and post op
4-D Quantitative Dynamic Lung MRI (QdMRI)

Δ LRC
Δ LD
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

CTIS 📞
Preop QdMRI (5 pts)

- Concave side
  - $\Delta$ rib cage volume 46.5 cc
  - $\Delta$ diaphragm volume 25.7 cc

- Convex side
  - $\Delta$ rib cage volume 40.5 cc
  - $\Delta$ diaphragm volume 27.4 cc
Preop QdMRI (5 pts)

- **Concave side**
  - $\Delta$ rib cage volume 46.5 cc
  - $\Delta$ diaphragm volume 25.7 cc

- **Convex side**
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  - $\Delta$ diaphragm volume 27.4 cc
Preop QdMRI
Concave Hemi-thorax
Diaphragm volumes

• 2 pts
  – concave $\Delta$ diaphragm volume < convex

• 1 pt
  – concave > convex

• 2 pts
  – had equal volumes.
Concave Diaphragm 39.2 cc

Convex Diaphragm 20.1 cc
Pre-op Renal Excursion

Kidney excursion

- Concave was 2.58 mm
- Convex 3.02 mm.
What is normal superior/inferior renal motion?

1-2 mm

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

- Concave kidney excursion increased 6.3mm (320%)
- Convex 5.2mm (192%).
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%
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
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
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
The Kidney Pedicle and other structures probably play a complicated role in the pathologic biomechanics of TIS.
Thank You

Jaxx Lee
you were given
THIS LIFE
because you
are
STRONG ENOUGH
to live it