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Thoracic Function: A new Thoracic Performance Classification based on Dynamic Lung MRI with Identification of a new Mechanism for Restrictive Lung Disease in EOS, Posterior Obstructive Blockade of the Diaphragm

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CTIS

Disclosures: RMC

- Royalties from Synthes Spine for the VEPTR device
- Non-compensated volunteer member Medical Advisory Board SpineForm Co.
- Medical Advisory Committee member National Organization of Rare Disorders (NORD)
- Grants
  - NORD and FDA Office Orphan Product development
- Provide advocacy for companies or inventors trying to develop safe and effective devices for children

What causes significant restrictive lung disease in EOS?

- Curve > 110˚

-Pehrsson, Bake, Larsson, Nachemson, Thorax ,1991

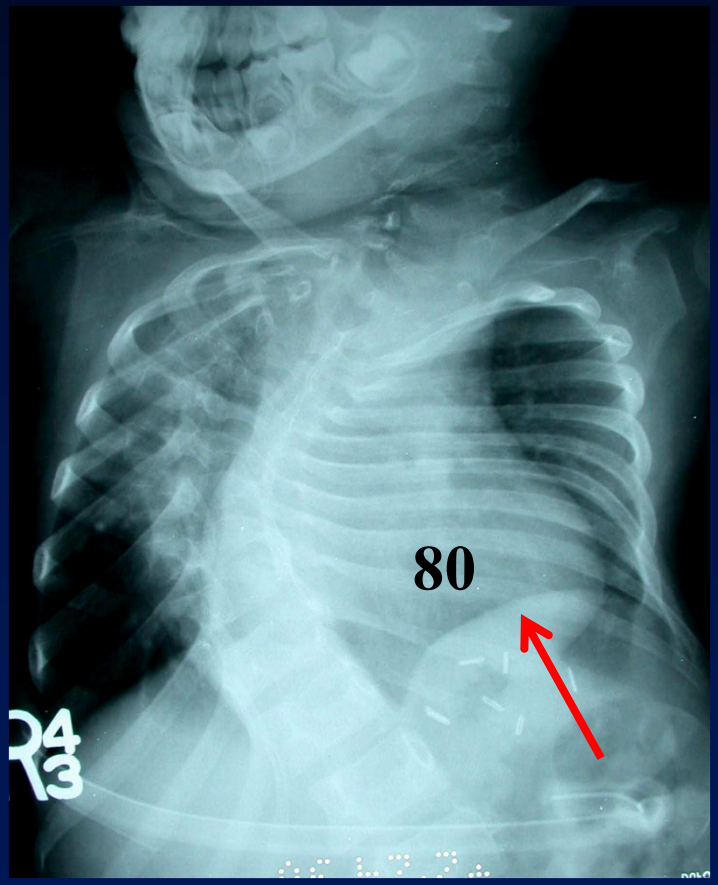
WHY ?

Primary Thoracic Insufficiency Syndrome

- Campbell, Smith JBJS 2004

Spine Deformity

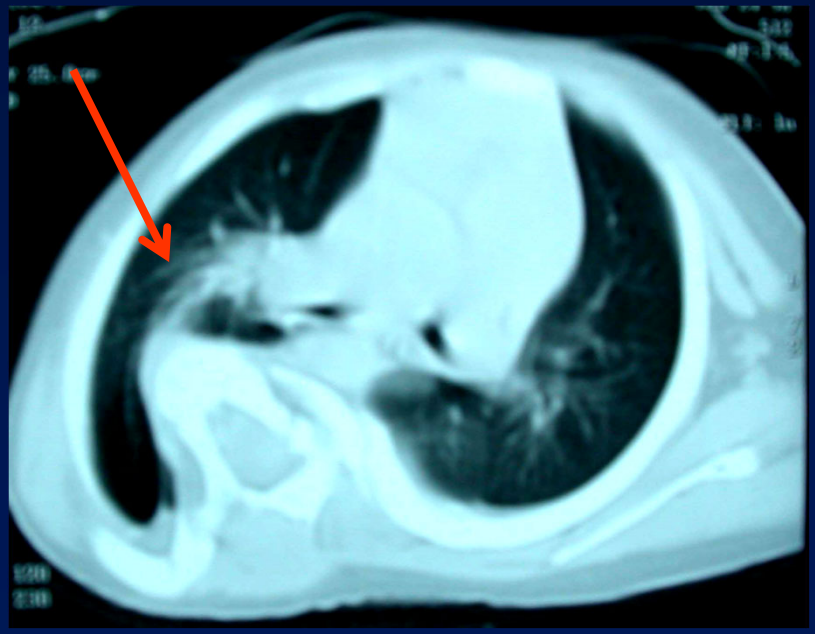
- Scoliosis
- Rotation



Effect on Rib Cage

" Wind Swept Thorax"

- Volume Reduction of Lung
- Motion Restriction of Ribs
  - Campbell, Smith et al JBJB 2003



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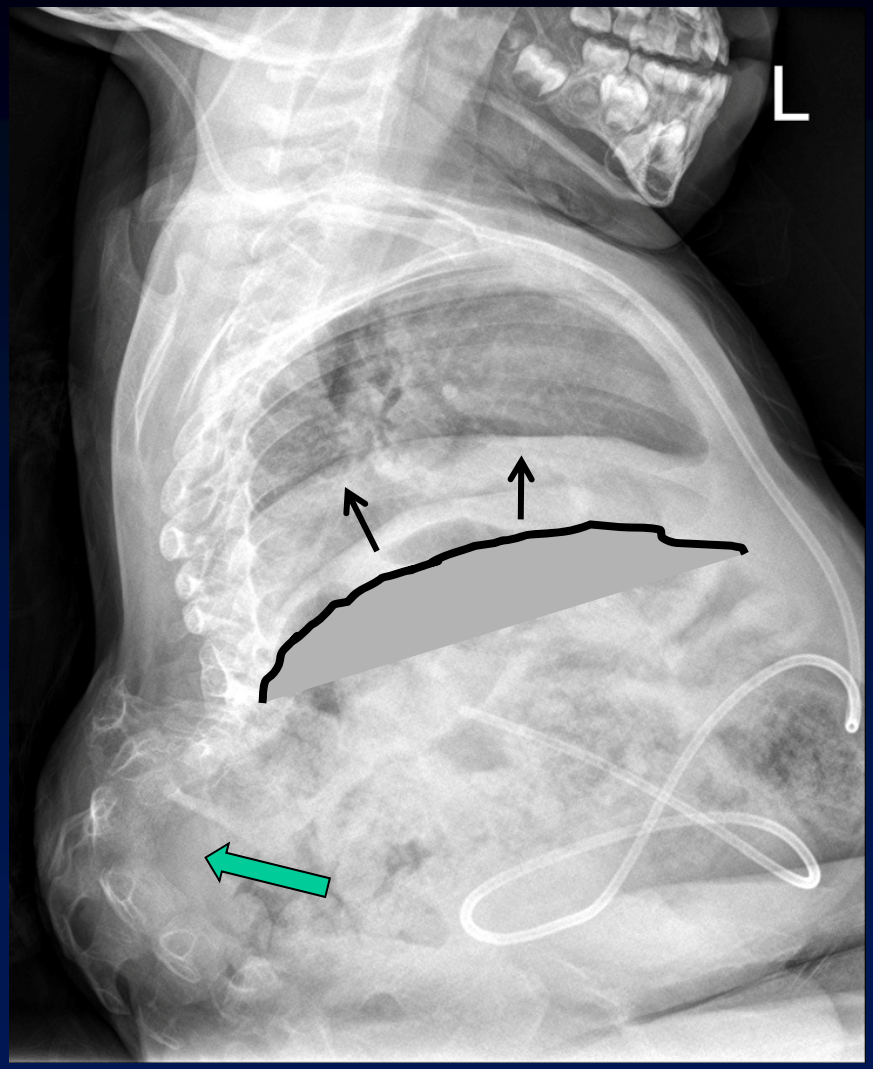
Secondary Thoracic Insufficiency Syndrome

- Campbell, Smith JBJS 2004



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Methods

- 6 Males, 4 Females
- Age 5.8 yrs (9 mo-10.75 yrs)
- All had TIS
- Supine Dynamic Lung MRI , most under sedation/gen anesthesia, spontaneous breathing

Each hemi-thorax graded

- 1- Intact motion of both chest wall and diaphrag
- 2- primarily loss of chest wall motion with minimal diaphragm abnormality
- 3- substantial loss of diaphragm excursion with minimal loss or compensatory hyper-kinesis of chest wall,
- 4- substantial loss of both diaphragm and chest wall motion.

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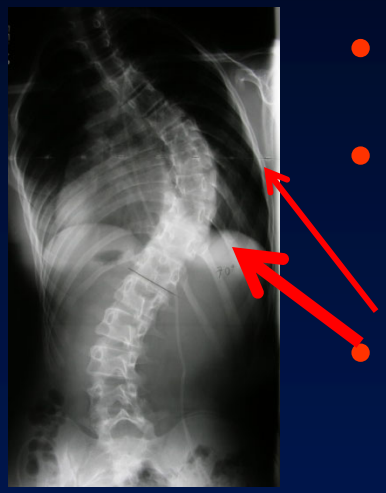
Each hemi-thorax graded



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Thoracic Function Score

- Average of

  Rt Hemithorax grade and Lt Hemithorax Grade

Levels of Clinical Thoracic Performance

- Level I thorax : 1 -1.5
- Level II thorax : > 1.5 – 2.5
- Level III thorax : >2.5 -3.5
- Level IV thorax : > 3.5 - 4.0

9 EOS pts

- 2 were level I
- 3 were level II
- 4 were Level III

- 1 pt with hypoplastic thorax without scoliosis was Level II .

PFT's and Thoracic Score

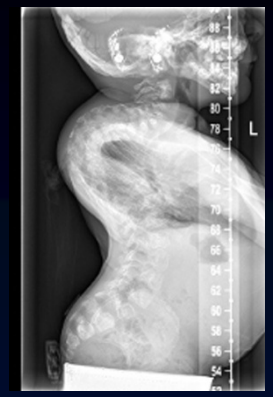
- Level I FVC 98% nl (I) 98% nl
- Level II FVC 63% nl
- Level II FVC 91% nl (II) 71% nl
- Level II FVC 58% nl
- Level III FVC 36% nl
- Level III FVC 71% nl (III) 61% nl
- Level III FVC 75% nl

Posterior Obstructive Blockade of Diaphragm

- In 4 patients there was a surprisingly marked posterior obstruction of diaphragmatic excursion by soft tissue organs.

POBD

- Level III FVC 75% nl



- Level III

- Level III FVC 36% nl

- Level II FVC 58% nl

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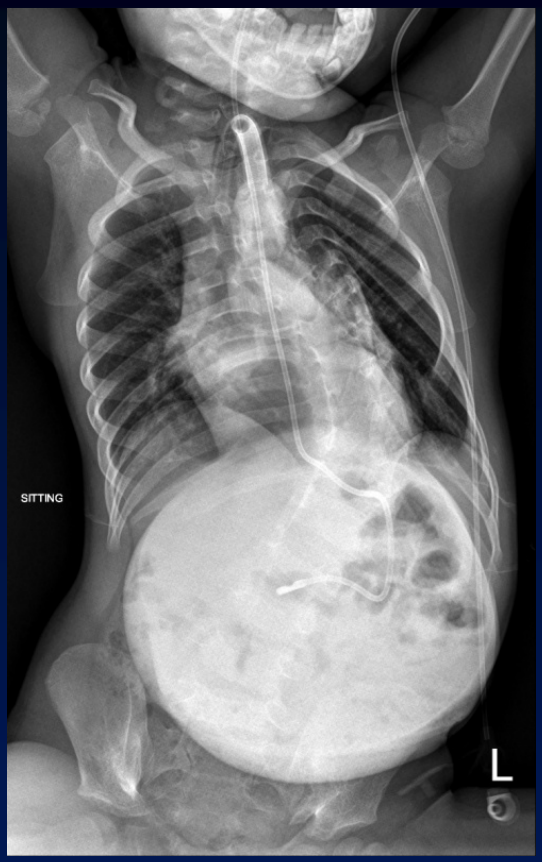
Giant omphalocele



- Level III thorax
- FVC 36% nl

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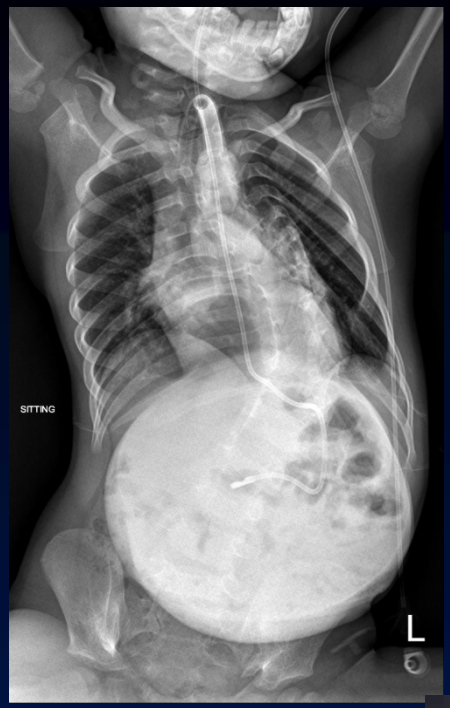

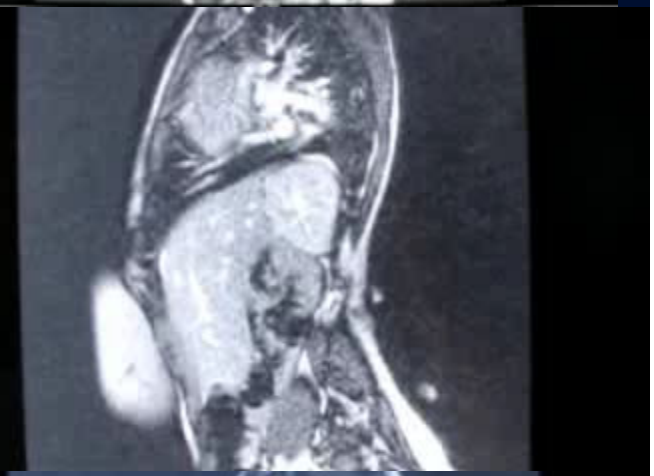

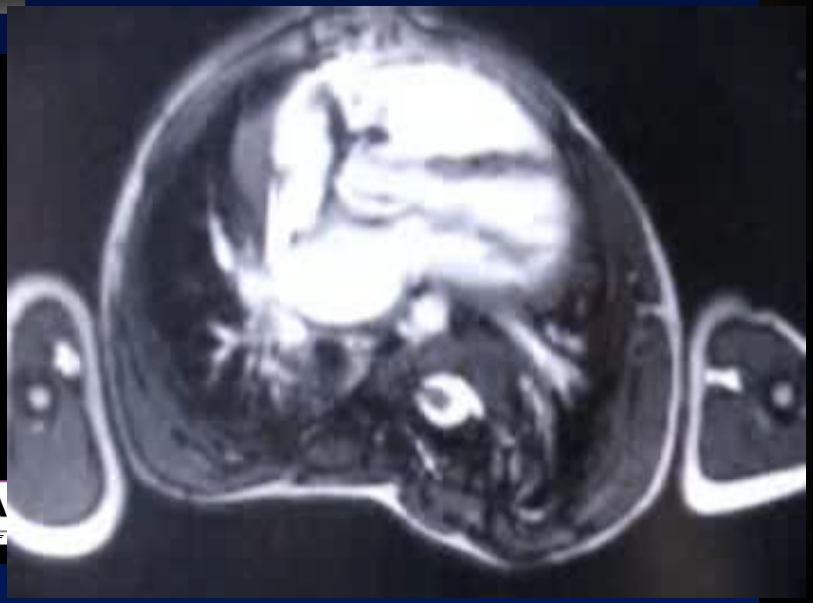

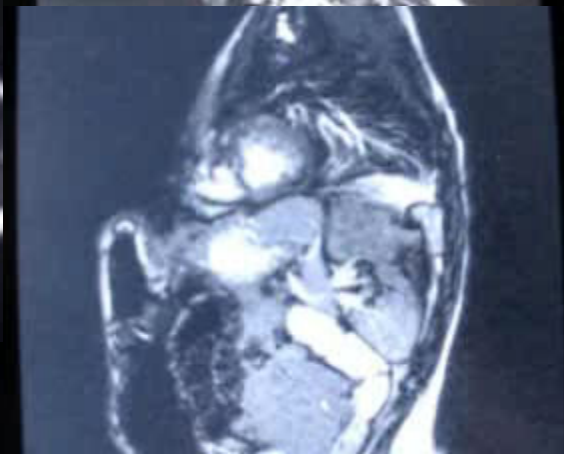

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Neuromuscular scoliosis
Level II thorax, FVC 58% nl

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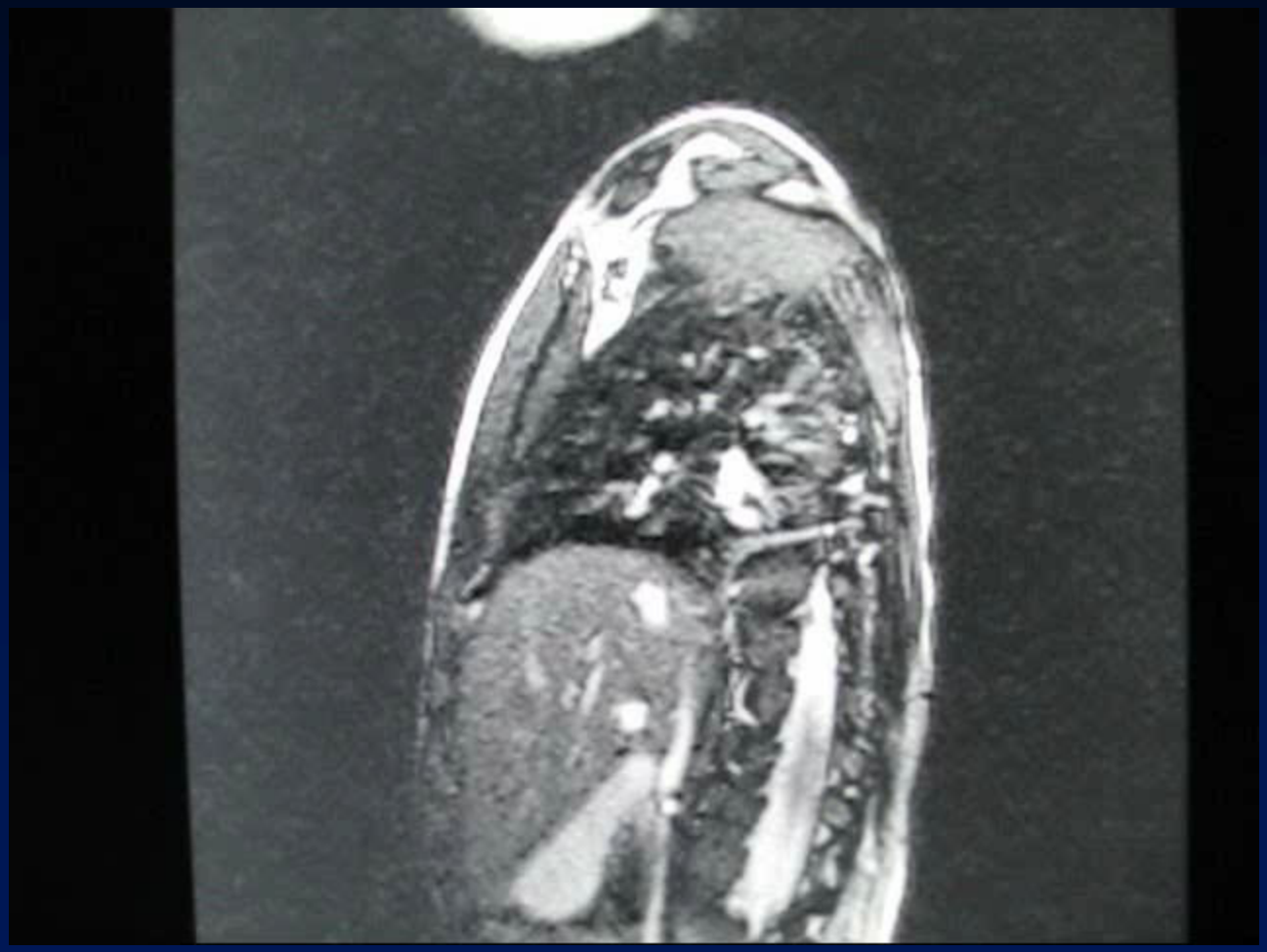
Concave side



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Convex side

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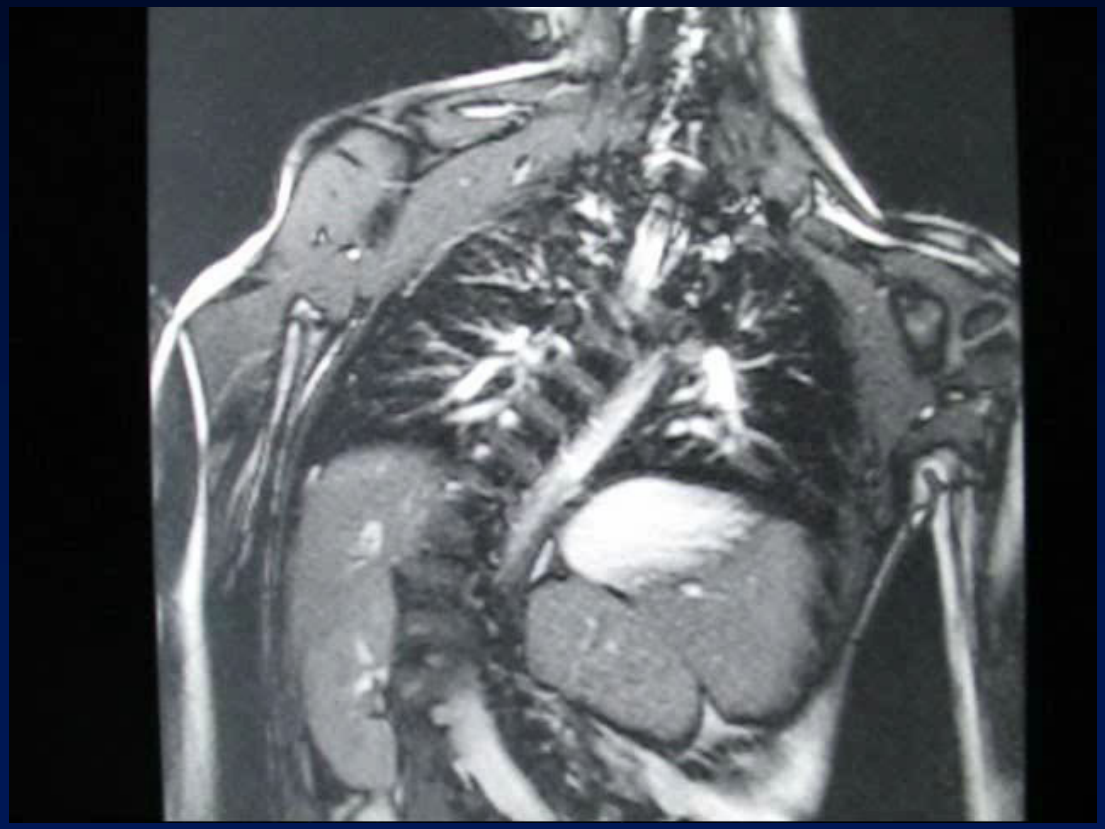
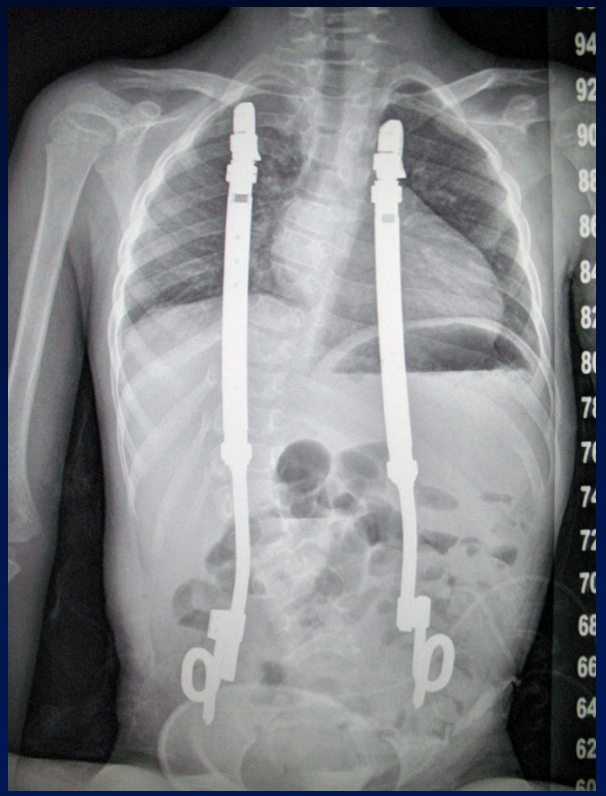
Will this relieve POBD?





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Conclusions

- Posterior Obstructive Blockade of the Diaphragm may help explain severity of restrictive lung disease in EOS
- More research will help refine our understanding of this new finding
- Thoracic Function Score is a way to look at the biomechanical basis of restrictive lung disease in EOS

Thank You!

R Campbell