Growing Spine ICL: Options, Tips, Techniques and Results

ICEOS Toronto Canada

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The Recognition of Thoracic Insufficiency Syndrome

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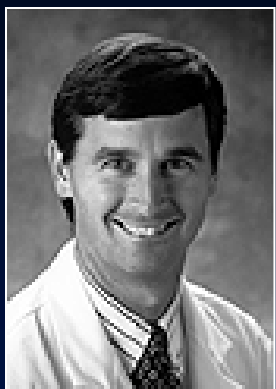

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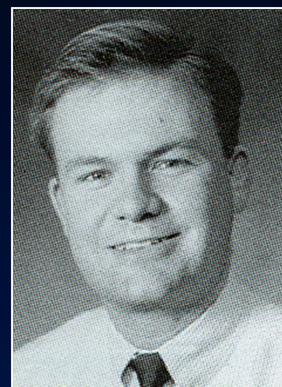
Pulmonary



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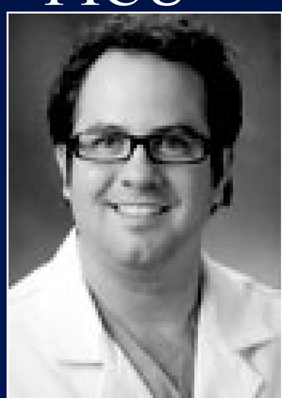

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Disclosures

- Grant Support
  – NORD
  – FDA Office of Orphan Product Development
- Royalties
  - Synthes Spine Co.
- Volunteer Advisor Spine-Form Co.
- Medical Advisory Committee member National Organization of Rare Disorders (NORD)
- Advocate for inventors/companies trying to develop safe and effective devices for children

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Thoracic Insufficiency Syndrome

The Inability of the Th ort

No

UNPOPULAR

Lung

Growth

-Campbell, Smith, et al.
J Bone Joint Surg, Mar , 2003
J Bone Joint Surg, Aug, 2004

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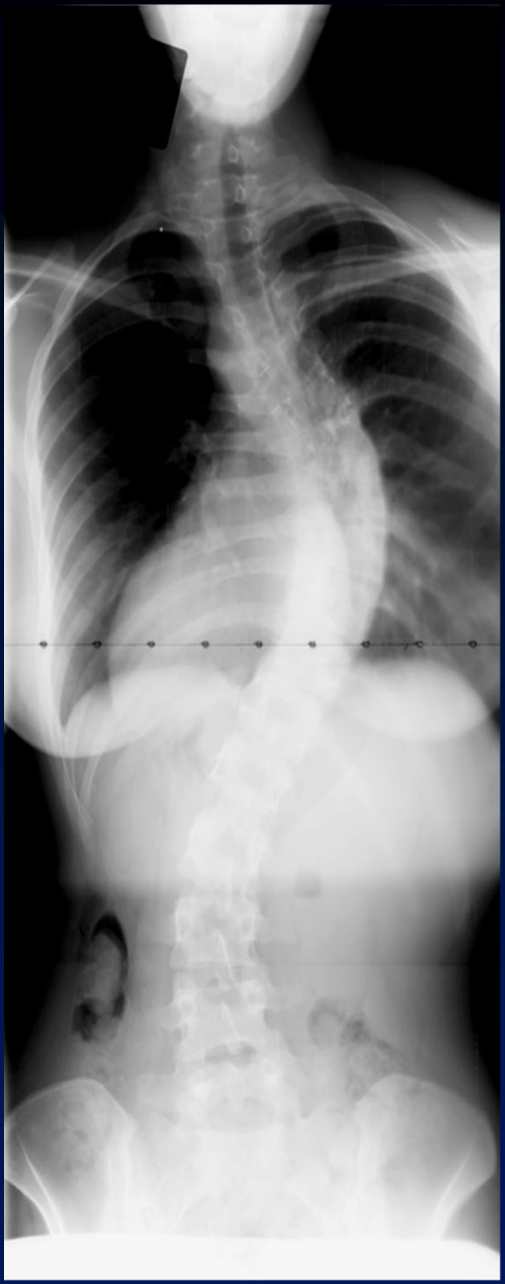

99%

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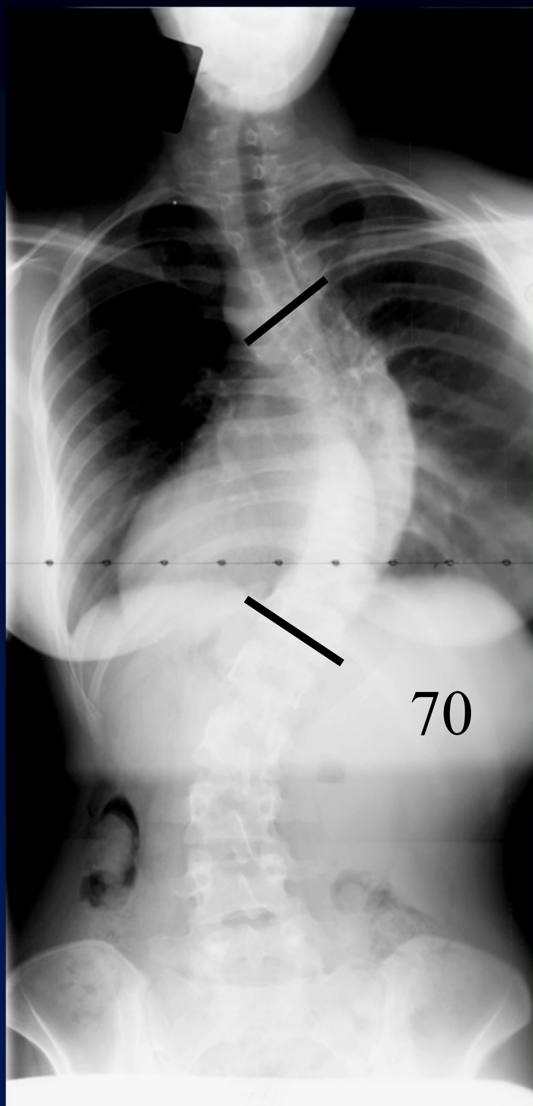

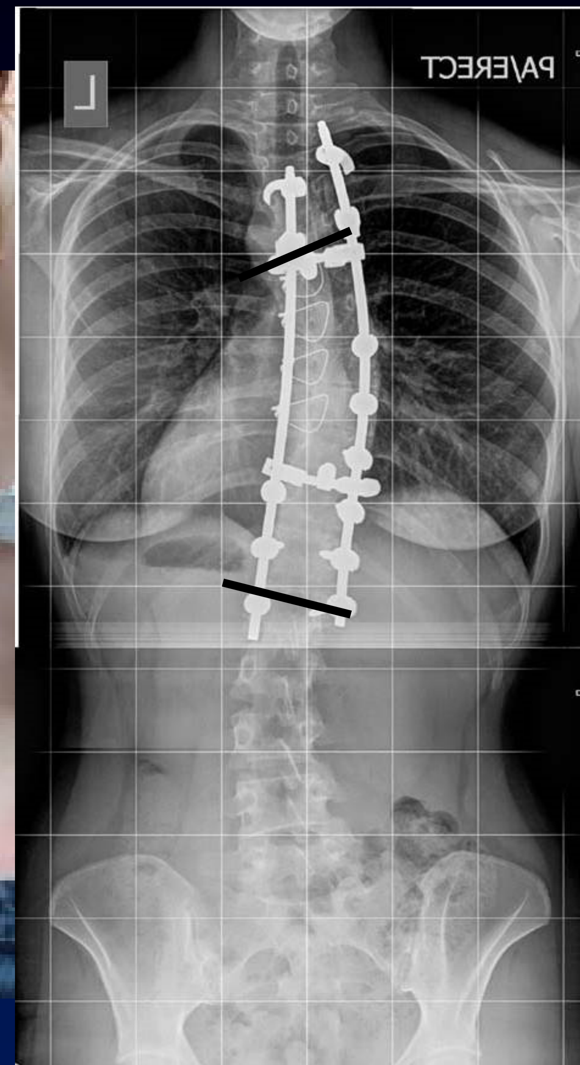

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Thoracic Insufficiency Syndrome

The Inability of the Thorax to Support

Normal Respiration

or

Lung Growth

Thoracic
Performance
And
Growth
Potential

-Campbell, Smith, et al.
J Bone Joint Surg, Mar , 2003
J Bone Joint Surg, Aug, 2004

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Normal Respiration

- Effortless at-rest breathing, able to respond to challenge activity without stress, ie, running
- Normal diaphragm excursion and rib cage expansion

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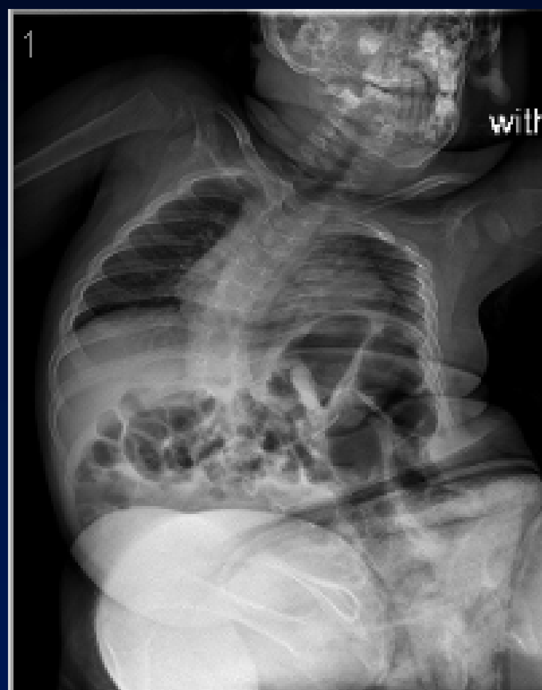
Normal Respiration?



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Thoracic Insufficiency Syndrome

- Mild, moderate or severe?
- Progressive? Getting worse without growth?
- So bad that respiratory insufficiency develops
  - Occult- clinically “seems” fine, but elevated respiratory rate, decreased play, other compensation mechanisms
  - Frank- cannot compensate, needs O2, CPAP, ventilator

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Normal Thoracic Growth Potential

- Thoracic spine is normal height
- Thorax has normal volume
- Thorax has normal symmetry
- And all of the above continues to be appropriate for age

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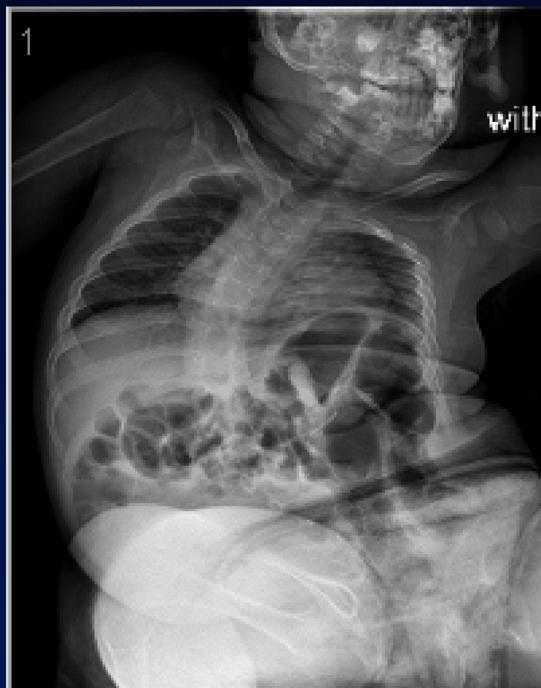
Normal Growth Potential?



Growth Potential of the Thorax

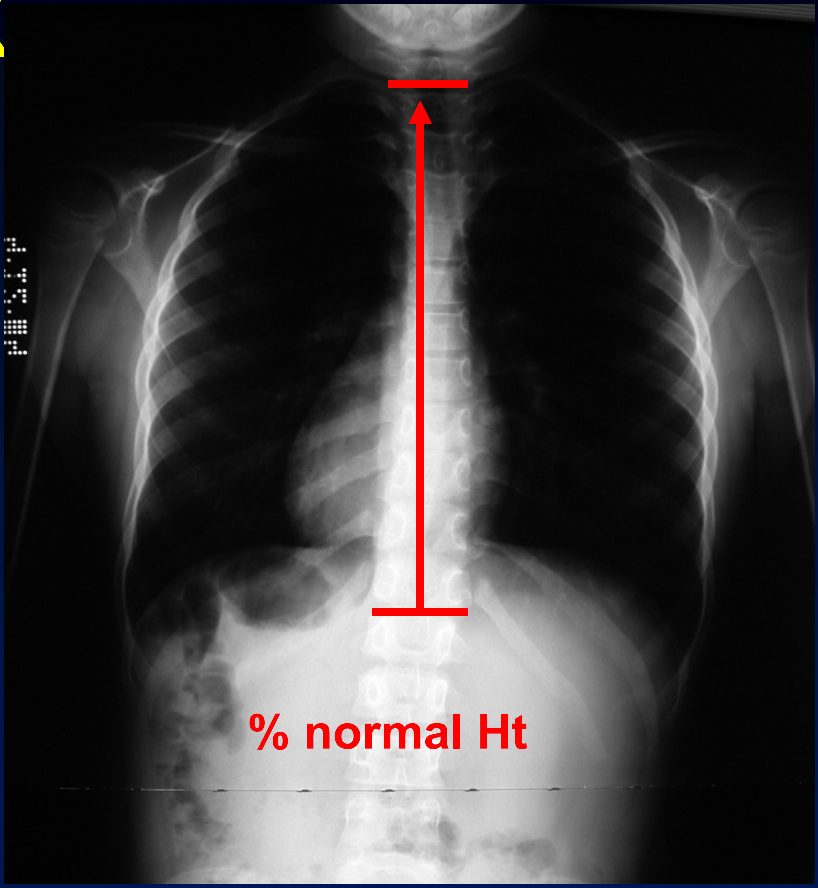
– **Height:**

Thoracic spinal growth

- **0-5 yrs: 1.4cm/yr,**
- **5-10 yrs: 0.6 cm/yr**
- **10-15 yrs 1.2 cm/yr**

-Dimeglio

– **Clinical Chest Circumference**



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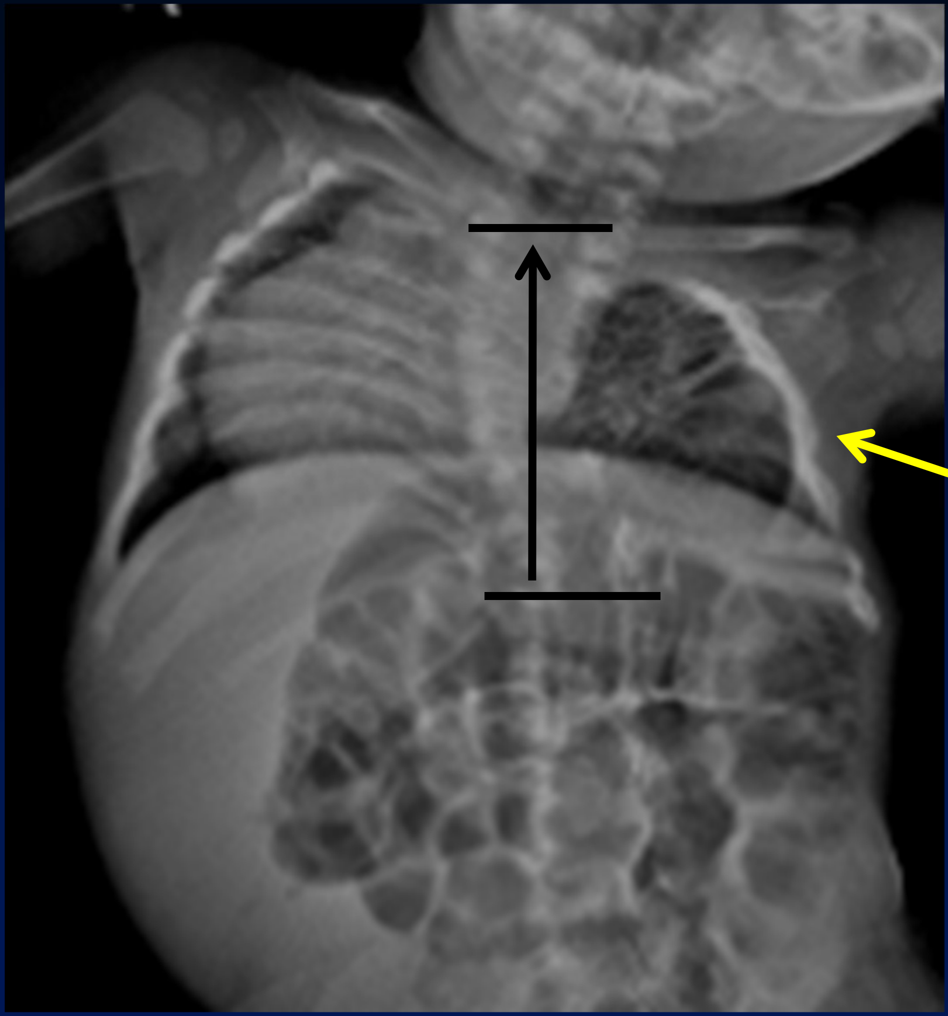
Growth Potential Problems



Thoracic Spinal height
60% normal

Fused rib cage
will grow poorly

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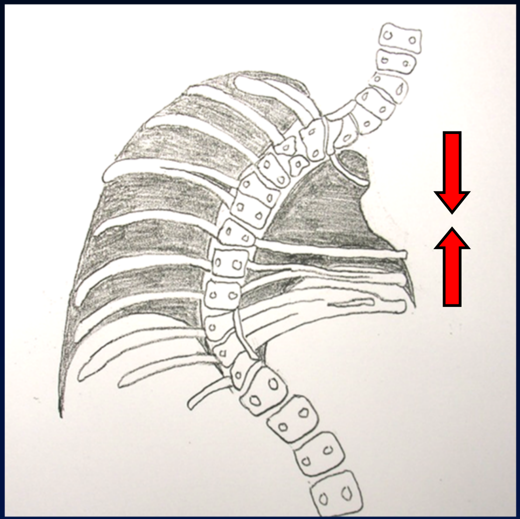
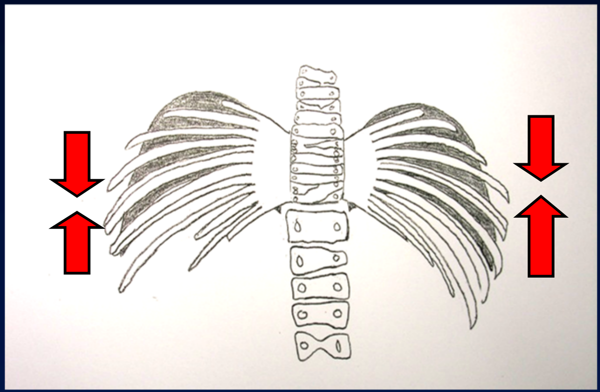
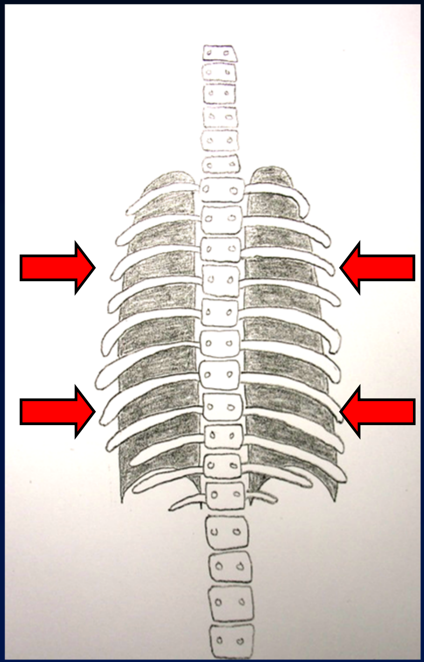
Volume Depletion Deformities of the Thorax

I







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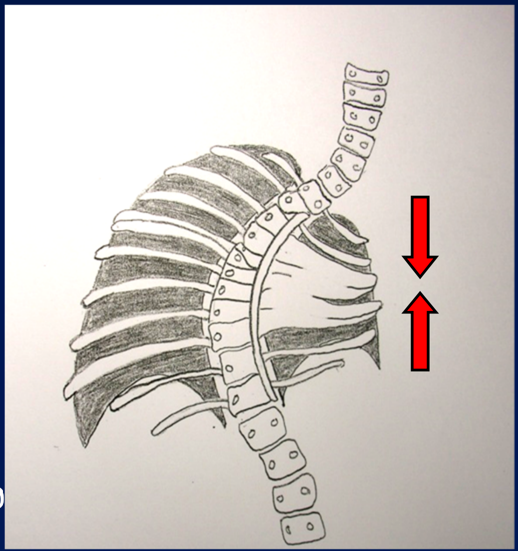


III a **III b**

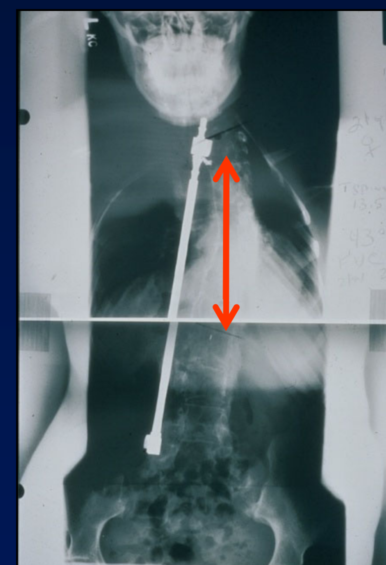
-Campbell Smith,
JBJS, supp, 2007

R Camp

Fusing scoliosis early may contribute to shortening of the thoracic spine and decreased thoracic/lung volume

- 28 pts
- Low FVC more prevalent in proximal spine fusions

Thoracic spinal height	FVC < 50%
< 18 cm.	63% pts
18 to 22 cm	25% pts
22 cm to normal	0% pts



- Karol et al., JBJS, 2008

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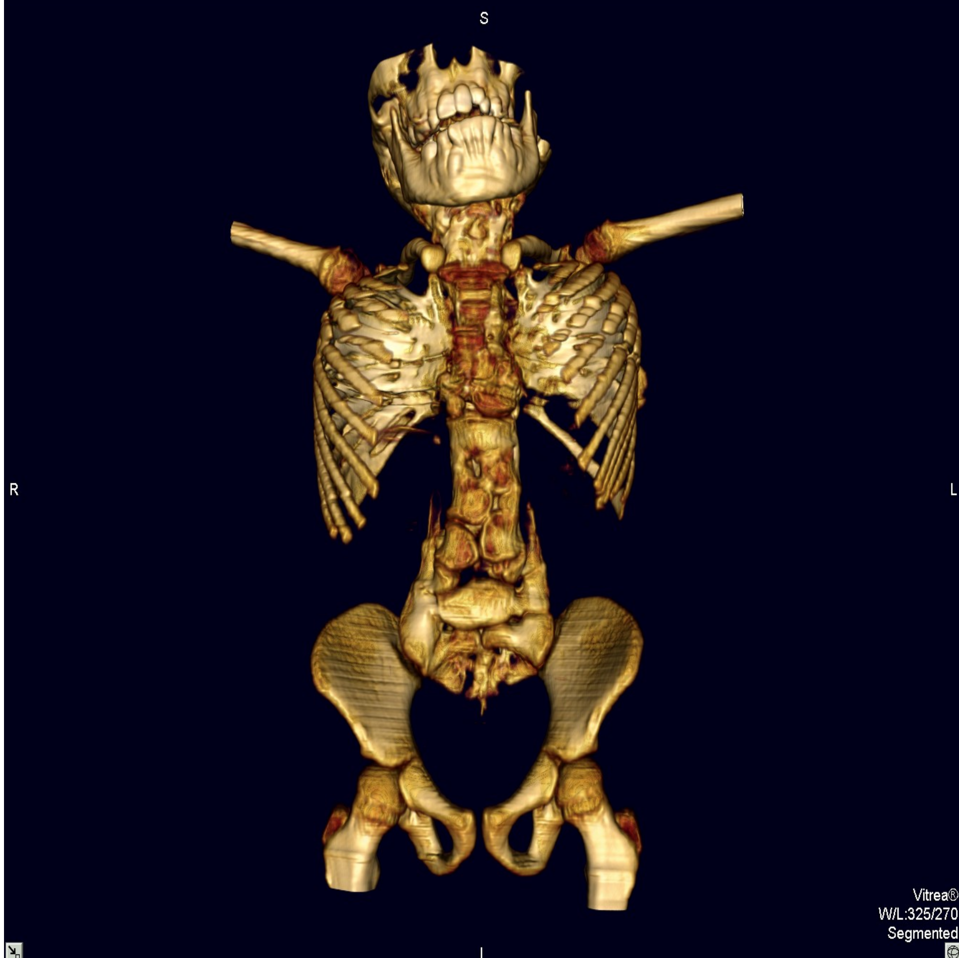
Spondylo-Thoracic Dysplasia



thoracic spinal height 24.2% nl
VC 27% nl

Ramirez, et al. JBJS, 2008

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Karol / Johnston 22 cm Rule ?

- **Pros:**
  - **Could be used to calculate tolerable growth deficiency of thoracic spinal height from viewpoint of VC**
- **Cons:**
  - **Does not consider rib cage deficiencies, ie VATER, hypoplastic thorax/ Jeune's type chest**

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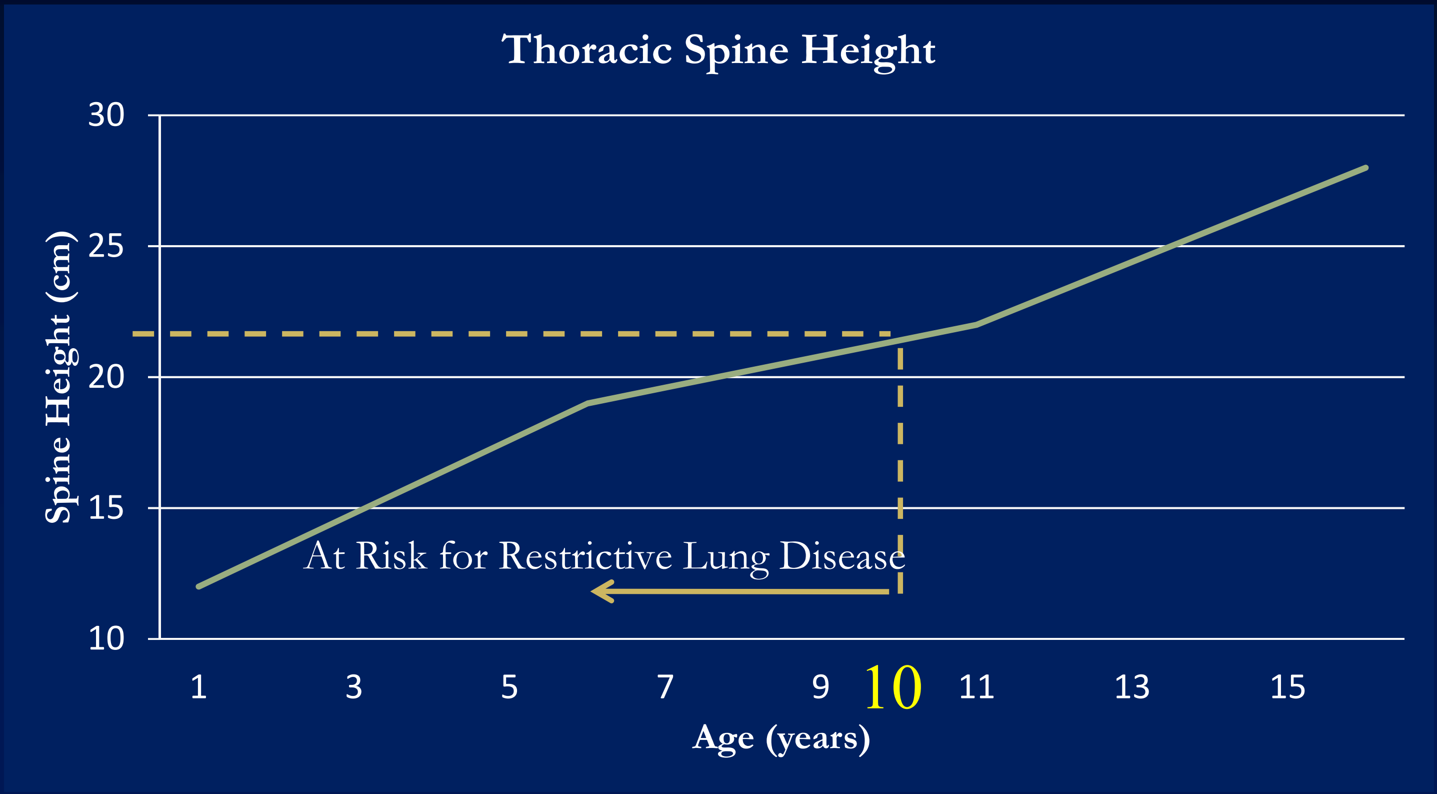
Early Fusion: A Critical Pulmonary Threshold?

-Fusing a spine before age 10 years may shorten a thoracic spine below the 22 cm threshold height
-Fusing a spine already shortened by congenital deformity may make matters even worse



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(Courtesy, Behrooz Akbarnia)

The original VEPTR concept

- Acutely reconstruct the malformed thorax , ie , opening wedge thoracostomy
- Add VEPTRs, as needed, to hold the thoracic reconstruction in place
- Expand VEPTRs to accommodate patient growth and maintain thoracic volume and symmetry

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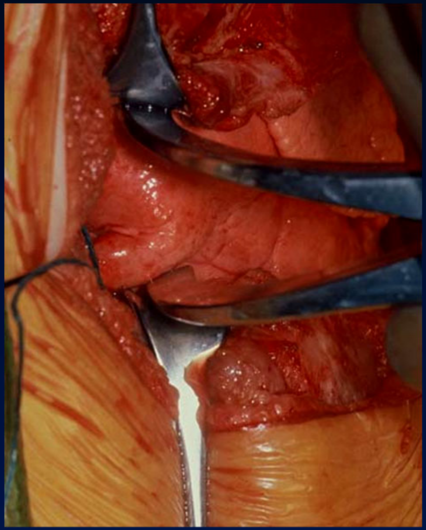
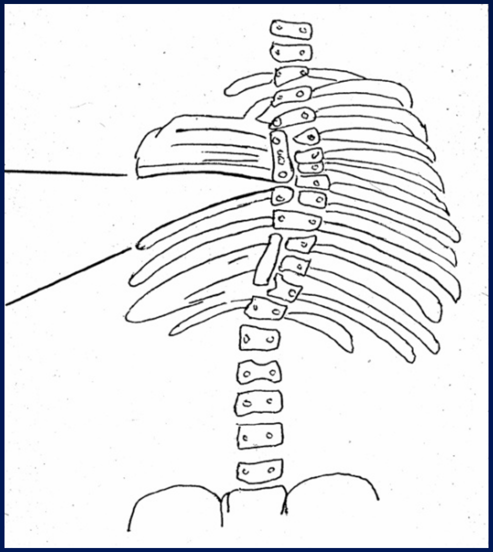
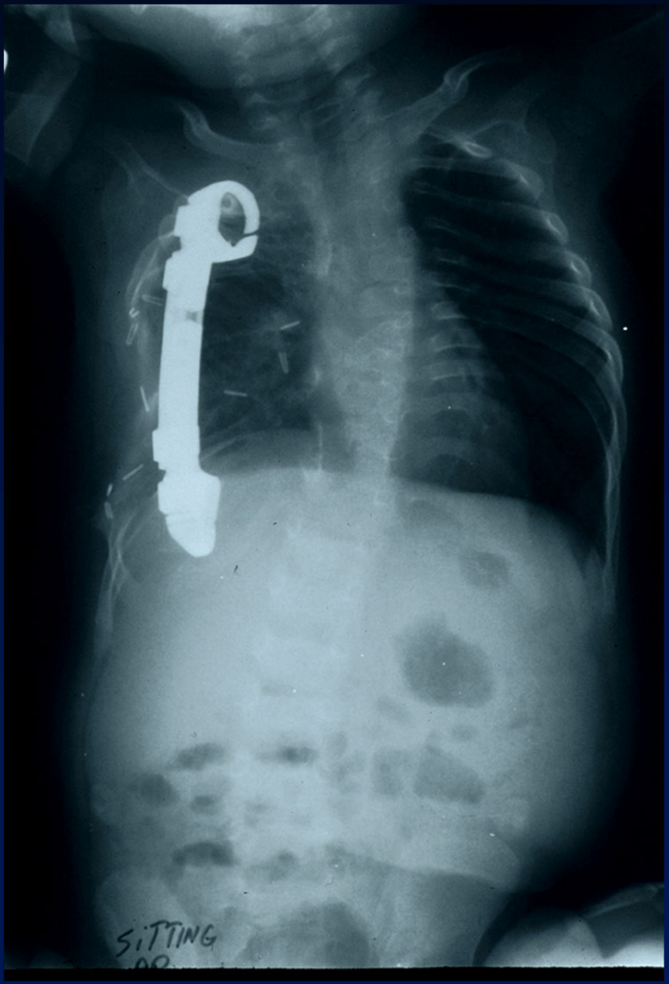
VEPTR Opening Wedge Thoracostomy







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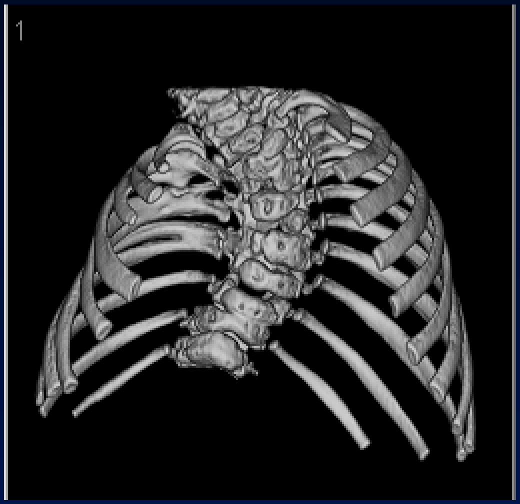
Congenital Scoliosis and rib fusion



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Percutaneous VEPTR without opening wedge thoracostomy
-John Smith, MD , Utah

Instrumentation drives correction,
rather than stabilizing the reconstruction

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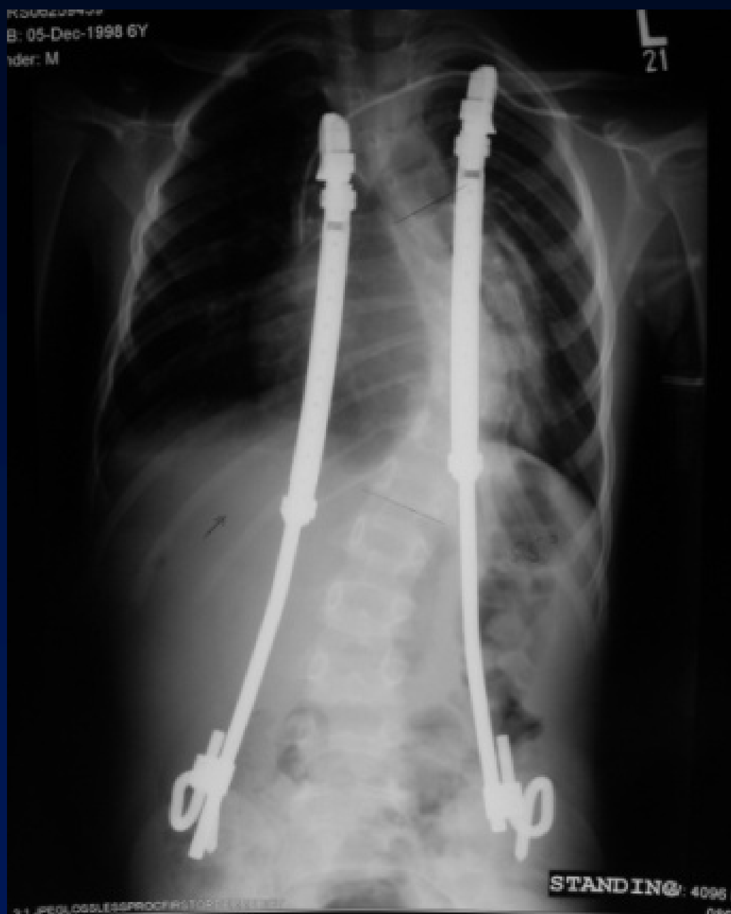
RIB BASED
spine distraction
instrumentation



SPINE BASED
spine distraction
instrumentation

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The choice?

Depends on your goals

The original goal of VEPTR surgery

- To obtain the largest, most symmetrical , most functional thorax possible by skeletal maturity

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Goals of most VEPTR/GR Users

- **A. Correct the Cobb angle, with the spine continuing to grow**
- **B. Minimize complications**

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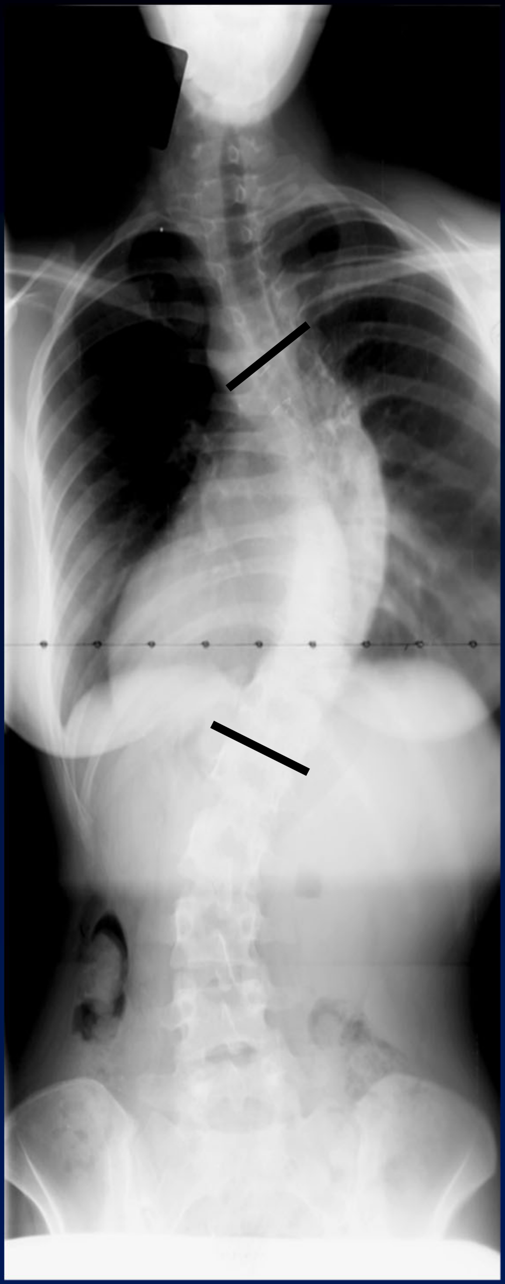

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Clinical Evidence

- **Growing rods: Small clinical series, 1982-present, heterogeneous populations**
- **VEPTR: Small clinical series, 2004-present, heterogeneous populations**
- **Both “appear” to function about the same for goals a and b**

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Expanding the Growth Sparing Instrumentation Goals

a. Correct the Cobb angle with the spine continuing to grow

b. Minimize complications

c. Minimize cost and morbidity

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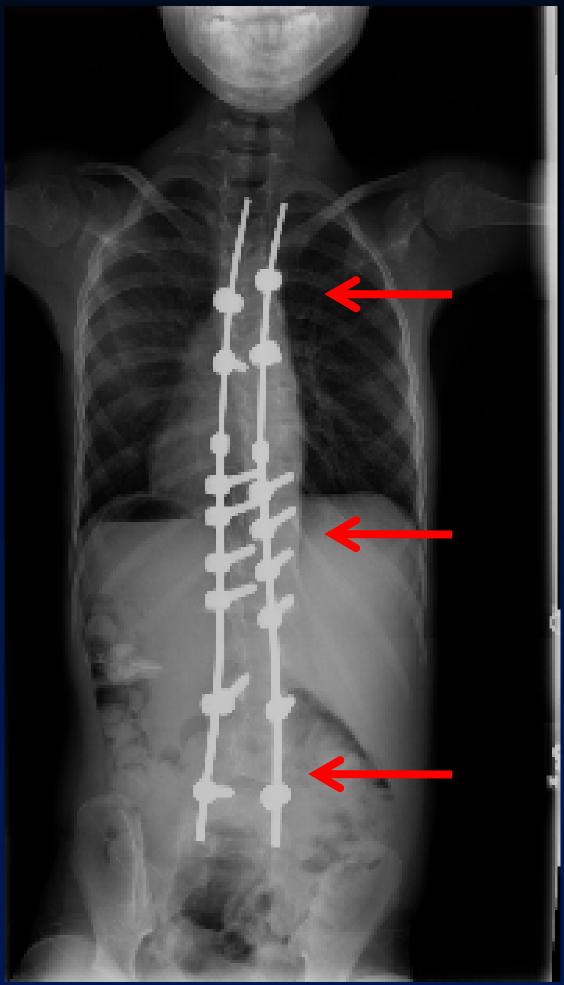
SHILLA



Pros:

- One Operation corrects deformity
- Then instrumentation “guides growth”
- Three (8?) point fixation

Cons:

- Additional operations for complications
- Unknown loss of thoracic spine growth because of central fusion/anchor points
- Long term complication rate unknown

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Growing Rods and VEPTR

Self expansion capability?

Pros:

- Distraction by physician/parents without surgery
  - Less cost
  - Less morbidity
- **Cons:**
- Long term cost?
- "Point of diminishing returns" ?
- Will it work long term?

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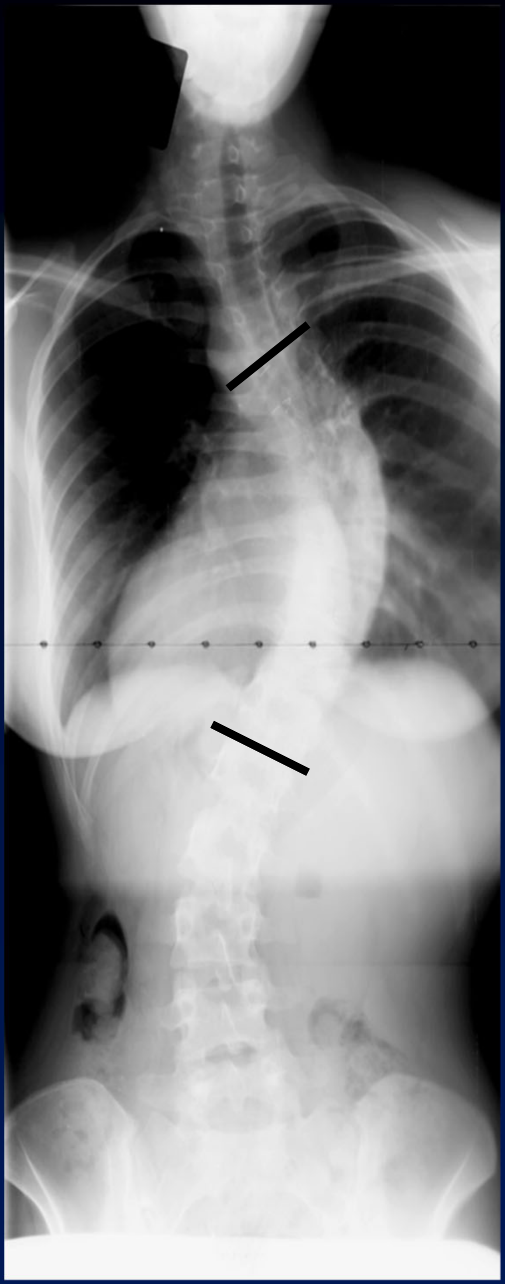
Adding Health Centered Goals

a. Correct the Cobb angle with the spine continuing to grow

b. Minimize complications

c. Minimize cost and morbidity

d. Improved Operative Long Term Health over Natural History



Natural History:
Mortality
Untreated Scoliosis

Thoracic Insufficiency Syndrome?

•Infantile: 0 to 3 years

•...e: 4 to 9 years

•Adolescent: 10 to 16 years

-Pehrsson, Larssson, Oden & Nachemson, Spine, 1992

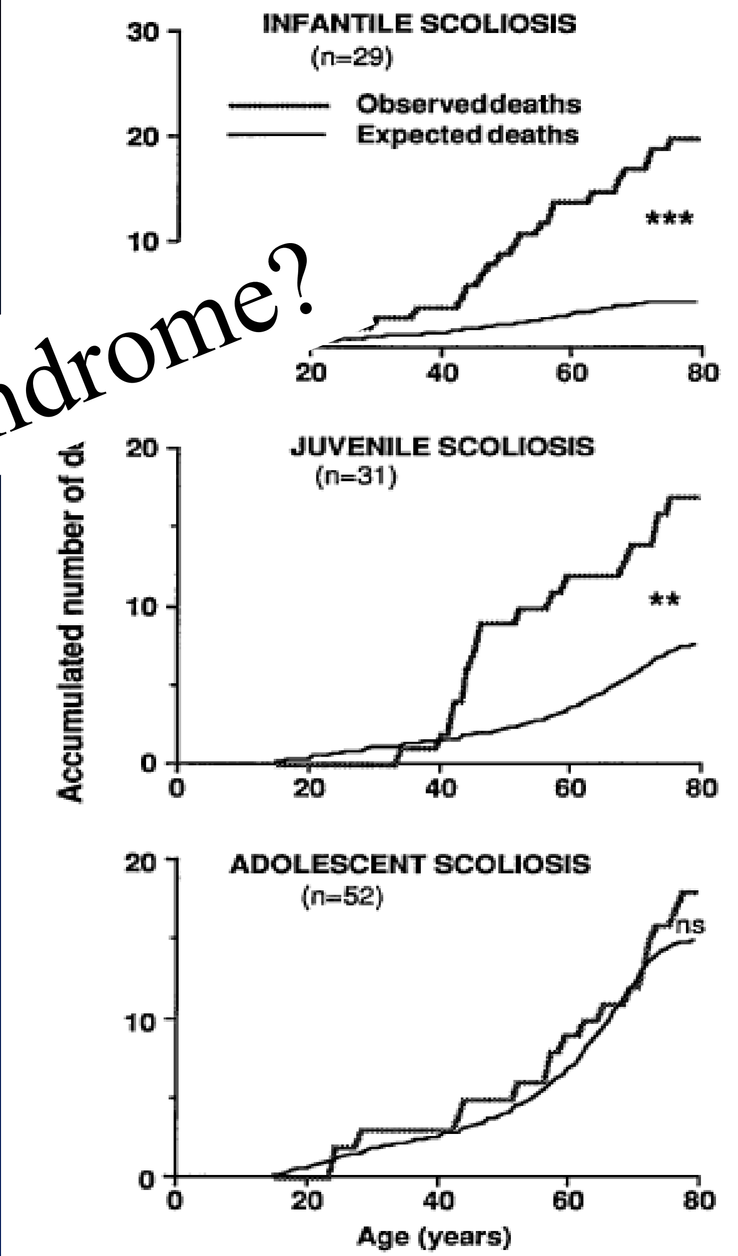


Figure 2. Cumulated number of observed deaths in patients with infantile, juvenile and adolescent scoliosis, and expected deaths.

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Morbidity

- Pulmonary
  - Higher risk of respiratory failure curves >110º
- Cardiac
  - Higher risk cor pulmonale, severe curves
- Appearance/Self Image
  - Probably worse
- “QOL” questionnaire data
  - Worse scores with Thoracic Insufficiency Syndrome
    - Vitale,et al.

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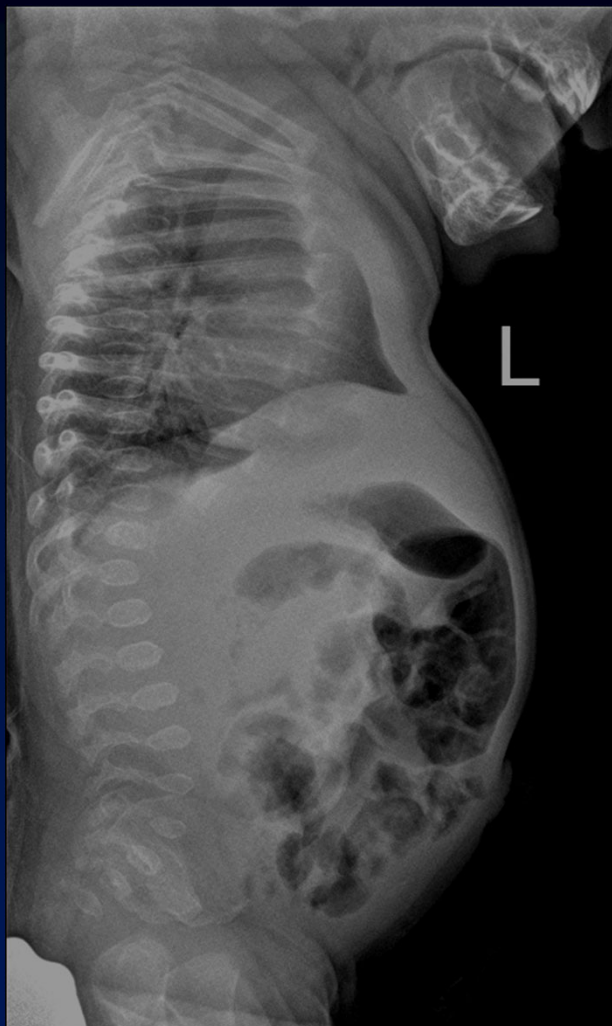
Long Term Health: Operative History of EOS



Mortality

- **Growing Rods:** Unknown
- **VEPTR:** Higher mortality hypoplastic cohort
- **SHILLA:** Unknown

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Morbidity

Pulmonary

- **Growing Rods:**
  - Unknown
- **VEPTR:**
  - Decreased % nl VC, Fused ribs scoliosis patients operated > 2 yrs age
  - Stable% nl VC post op?
- **SHILLA:**
  - Unknown

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Morbidity

Cardiac

- **Growing Rods:**
  - **Unknown**
- **VEPTR:**
  - **Unknown**
- **SHILLA:**
  - **Unknown**

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Morbidity

Appearance/Self Image

- **Growing Rods:**
  - **Unknown**
- **VEPTR:**
  - **FDA data**
- **SHILLA:**
  - **Unknown**

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Morbidity

"QOL" questionnaire data

- **Growing Rods:**
  - **Unknown**
- **VEPTR:**
  - **FDA data**
- **SHILLA:**
  - **Unknown**

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Quality of Life Instruments for Scoliosis

- SRS 22
- SRS 22r
- SRS 23
- Walter Reed Visual Assessment Scale
- SRS 24
- SRS 36
- Spinal Appearance Questionnaire
- Turkish , Spanish, Italian Version SRS 22
- Child Health Questionnaire-CF87

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Would it make sense to go back to the original goals of VEPTR surgery?

- To obtain the largest, most symmetrical , most functional thorax possible by skeletal maturity

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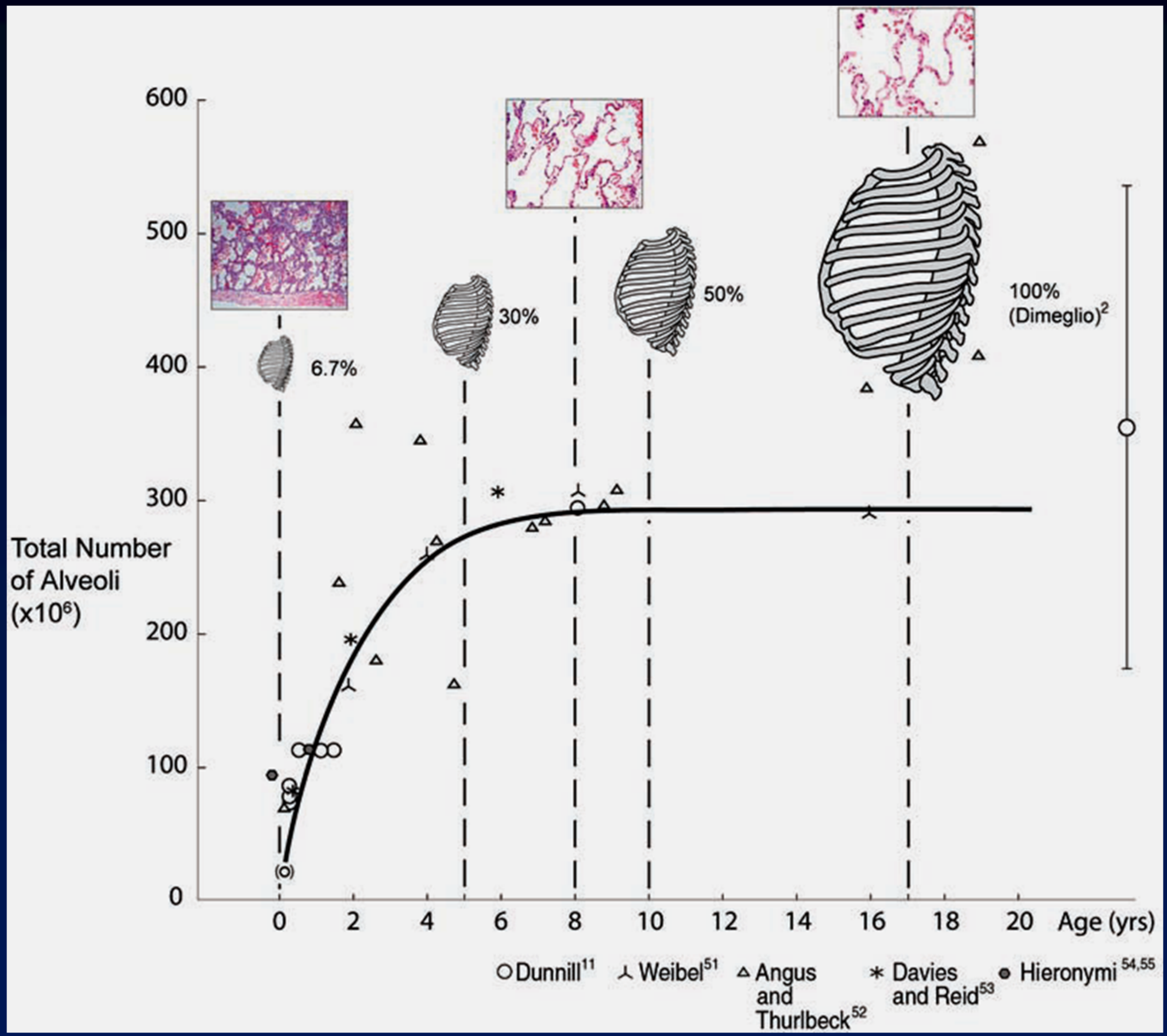
Use biology



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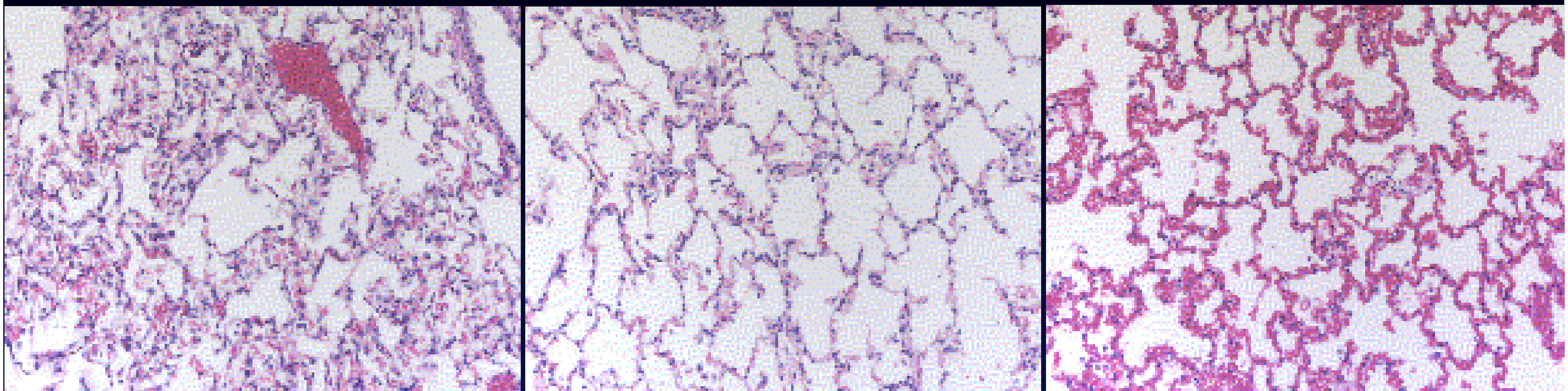
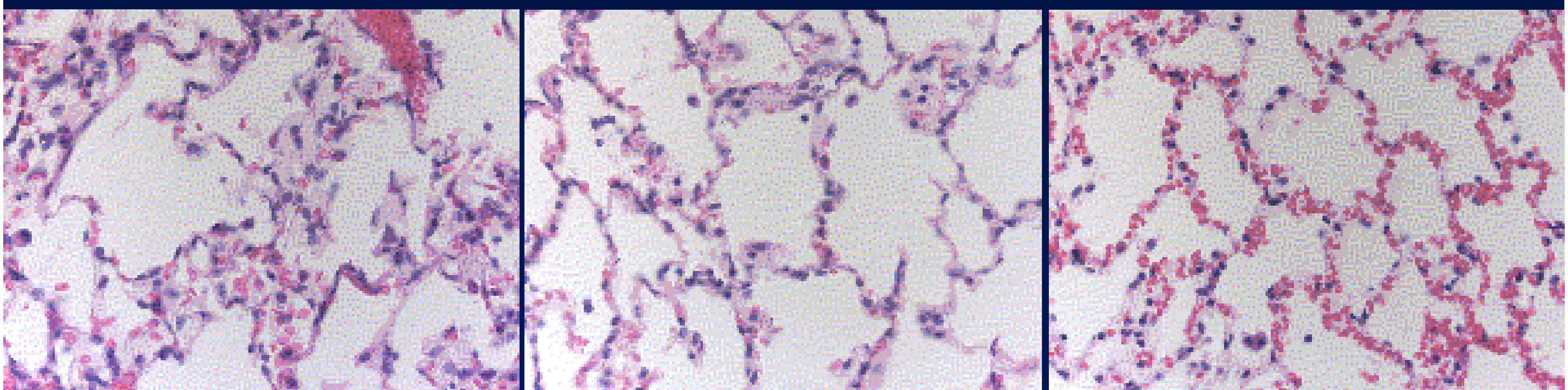
Lung Histology: Animal model (Mehta, Olson,Snyder)

Disease Control | **Normal Control** | **VEPTR Treated**



200x **200x** **200x**



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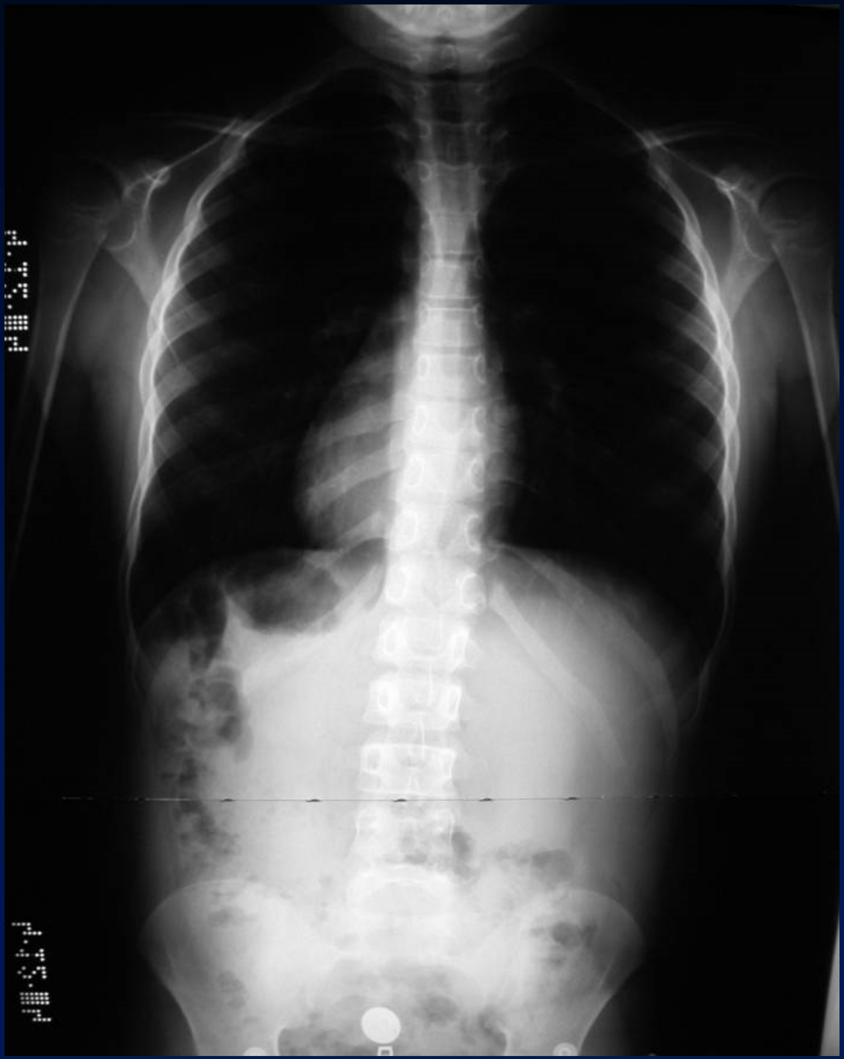
400x **400x** **400x**

Measuring Thoracic Performance



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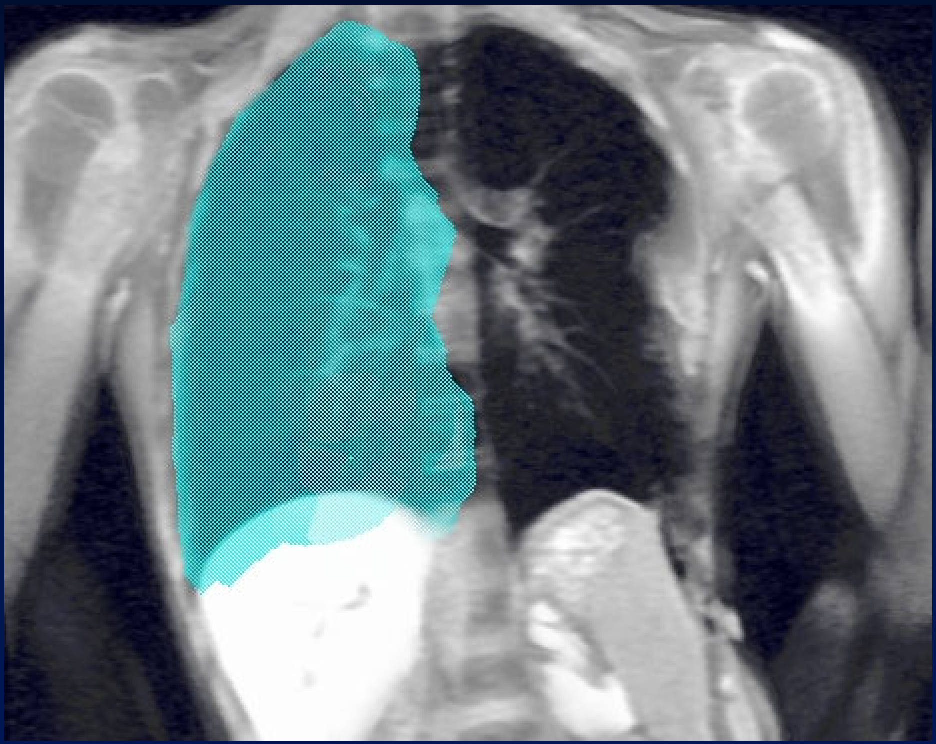
CT scan Lung Volumes

- (Johnston, et al.)



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Emans, et al. Spine, 2005

Lung volume by CT:

- Pre-op: 369 ± 279 cm3
- First Post-op 394 ± 289 cm3
- Last Follow-up: 736 ± 462 cm3

Lung on side of VEPTR increased:
219% ± 306% (range, 13%–1,160%)

Lung not on side of the VEPTR increased:
147% ± 176% (range, 24%–731%)

The ratio of right to left lung volume compared with a normal value of 0.85 improved by 13%

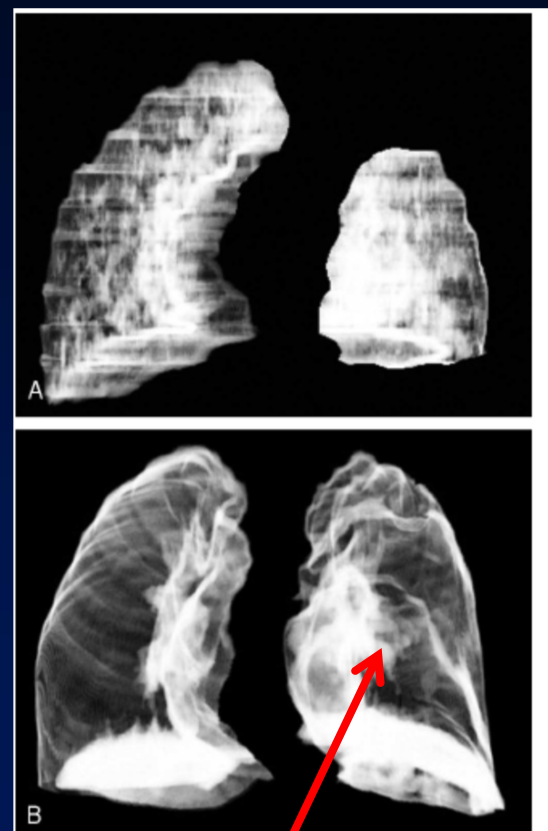
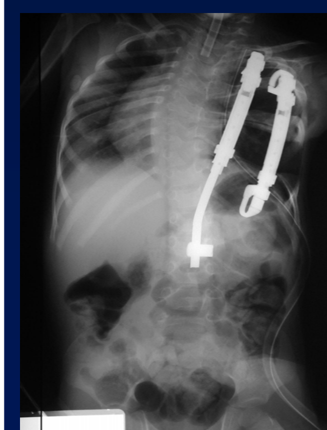


Figure 5. Preoperative (A) and postoperative (B) individual three-dimensional lung reconstructions for measurement of lung vol-



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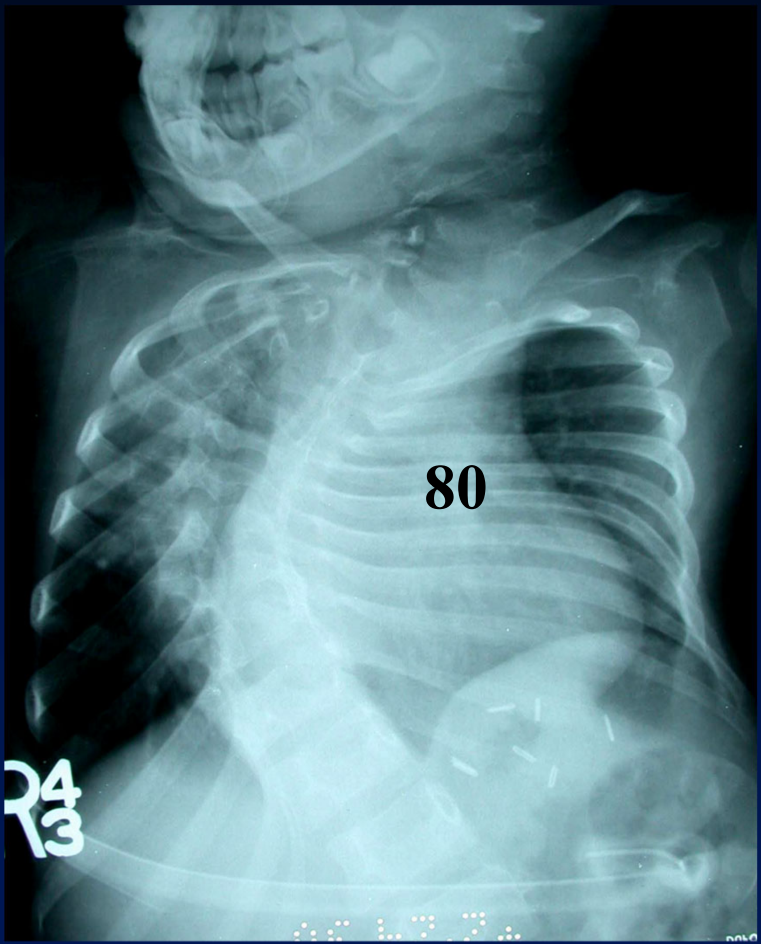
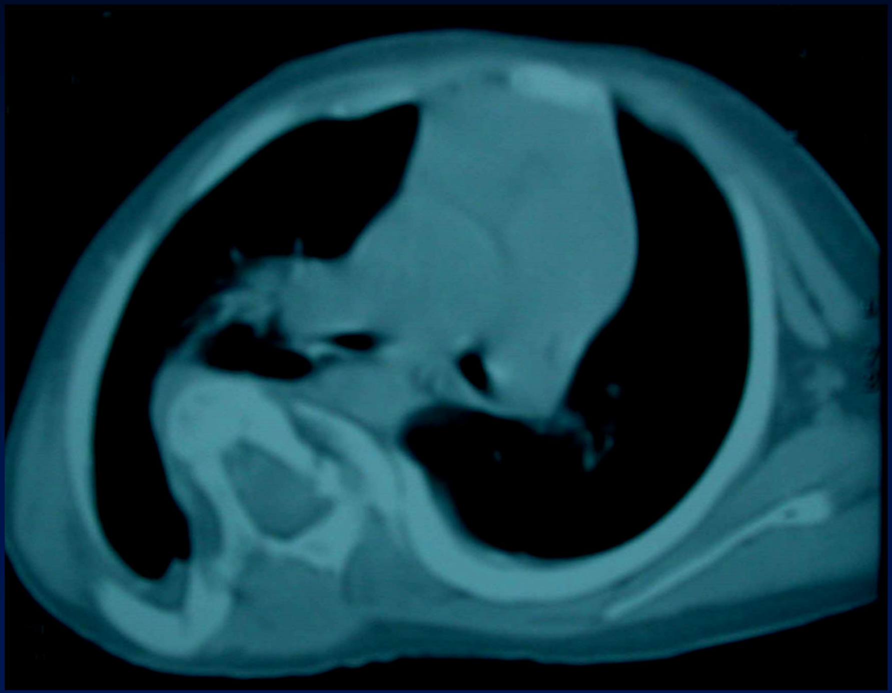
Helps us understand the 3-D deformity





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Pulmonary Function Testing

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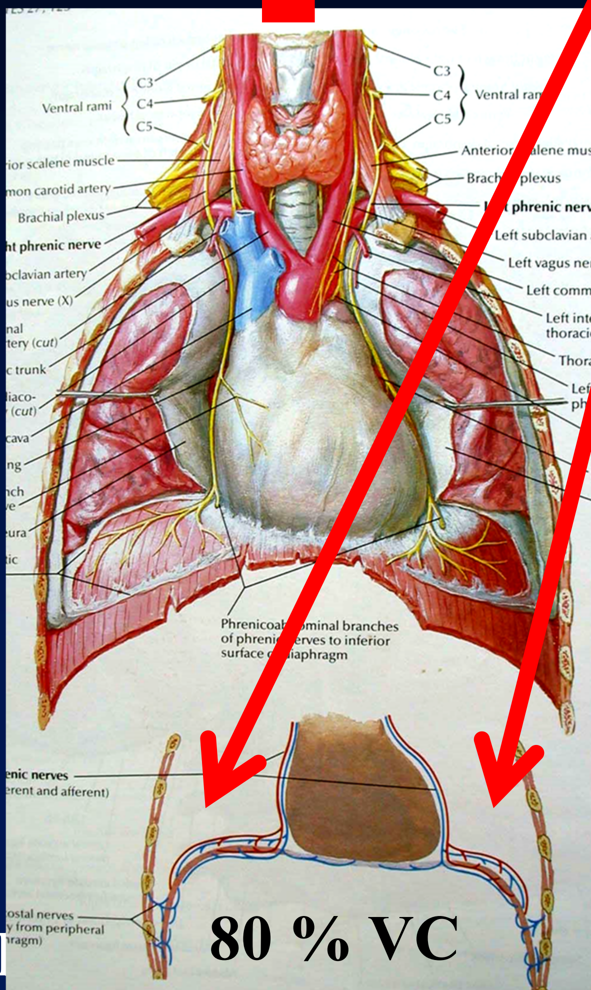
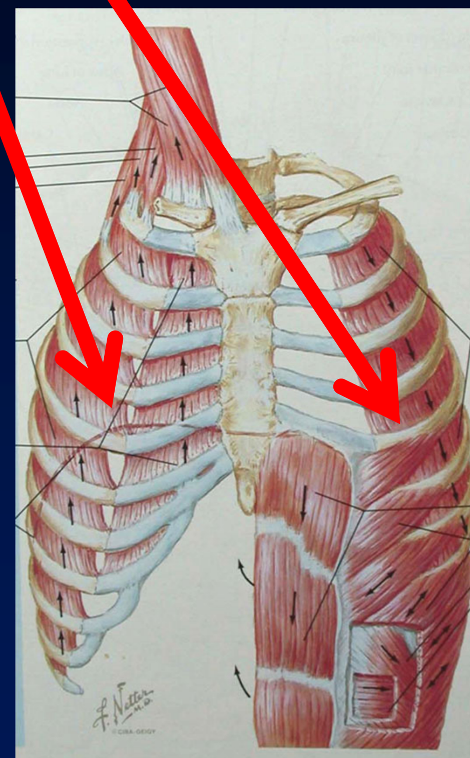
Thoracic Performance drives PFTs

VC



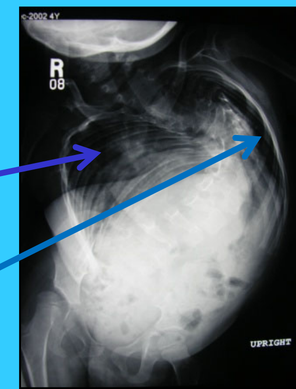
CT

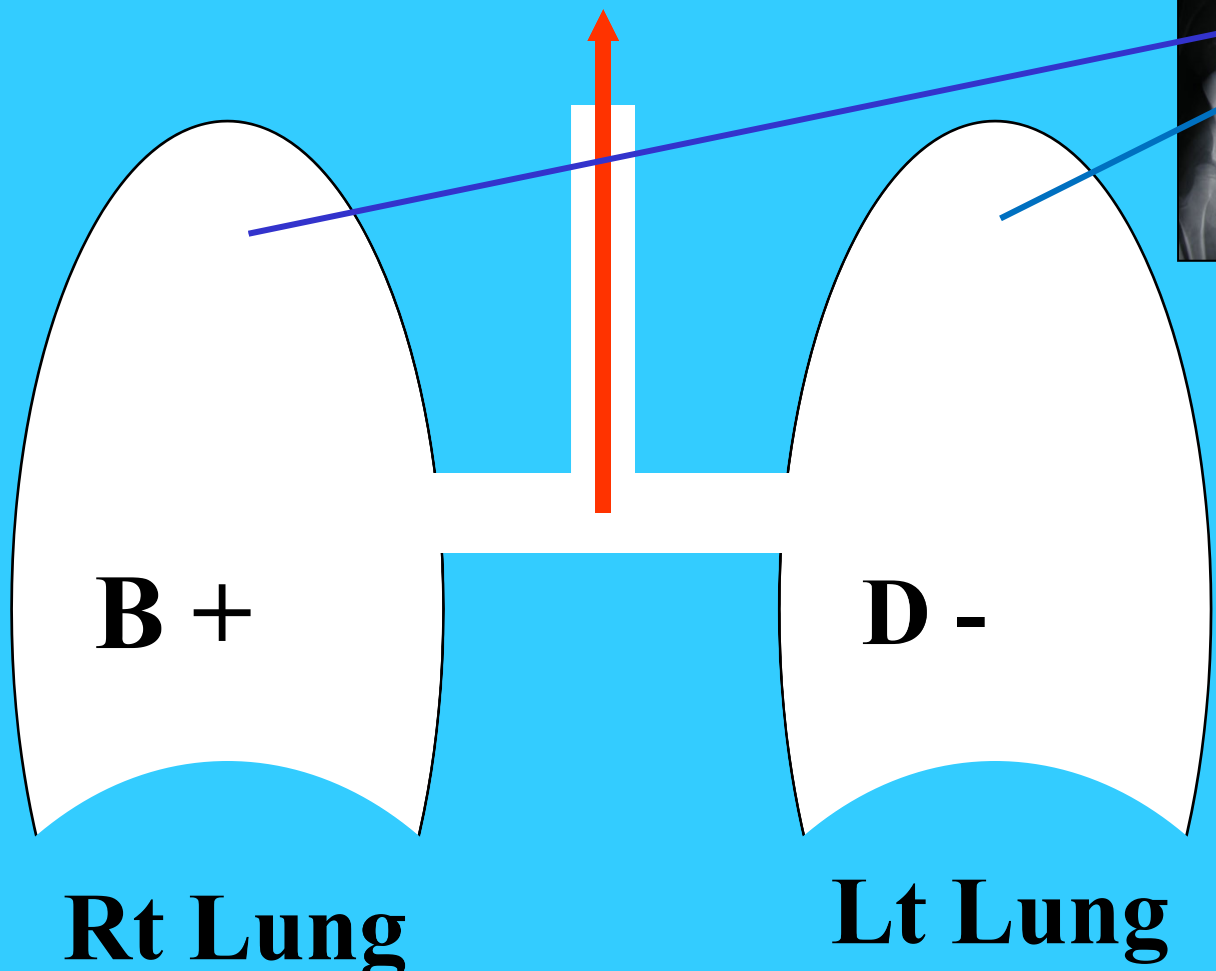
80 % VC



20 % VC

PFT <u>averages</u> these : FVC, TV, Etc .. = C + ?





PFT's are summative


QANTAS AUSTRALIA'S OVERSEAS
V-JET
Individual Engine Oil Pressure

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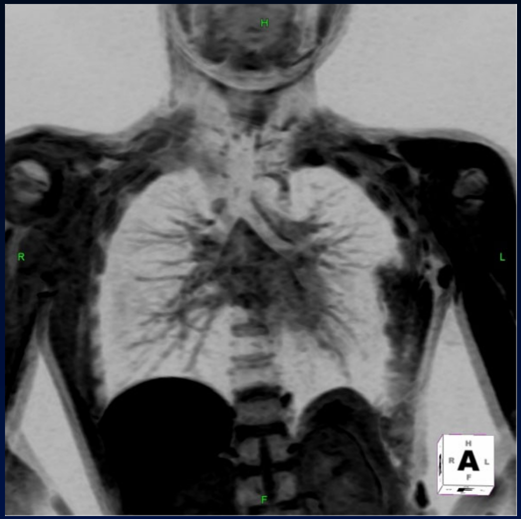
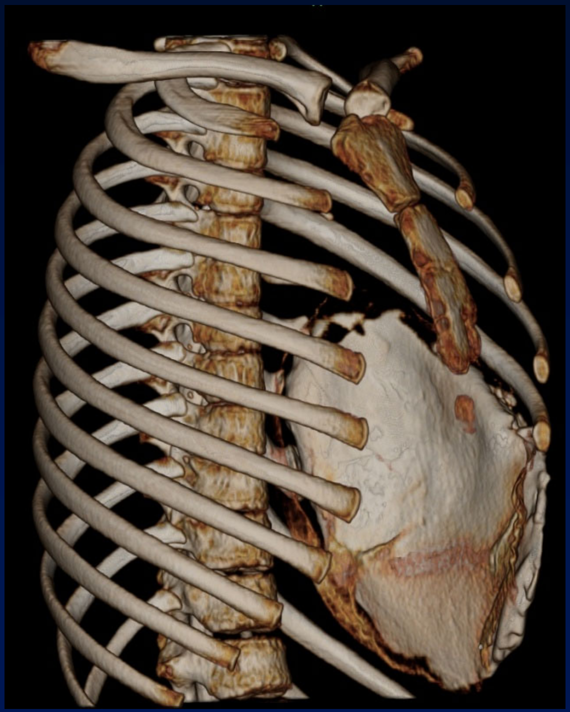
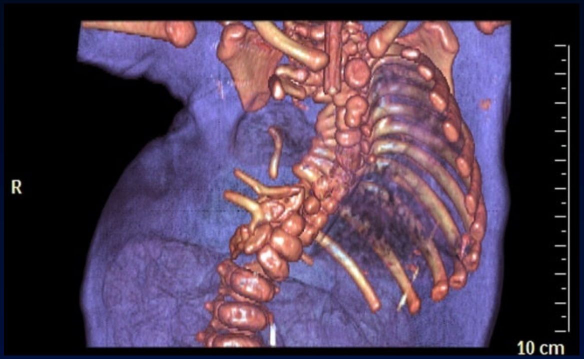
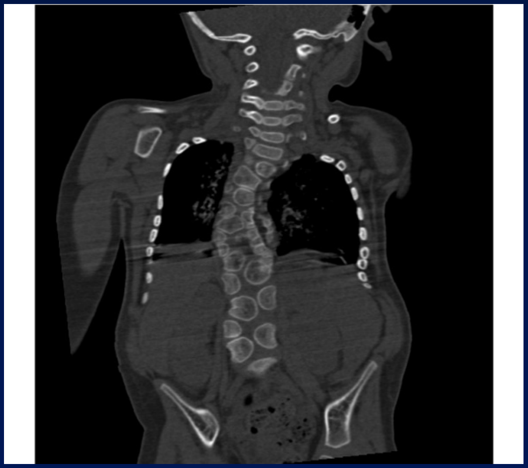
The Future









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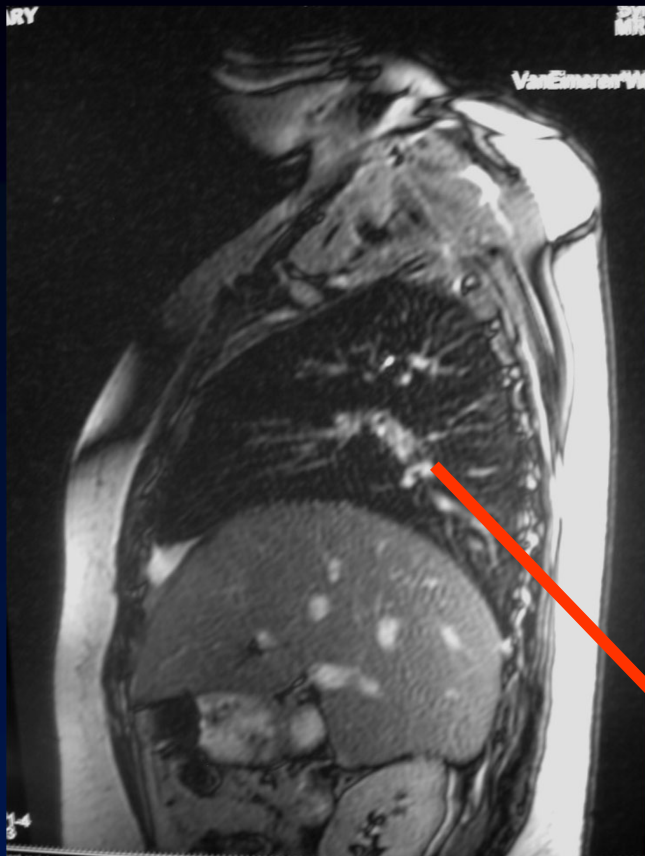
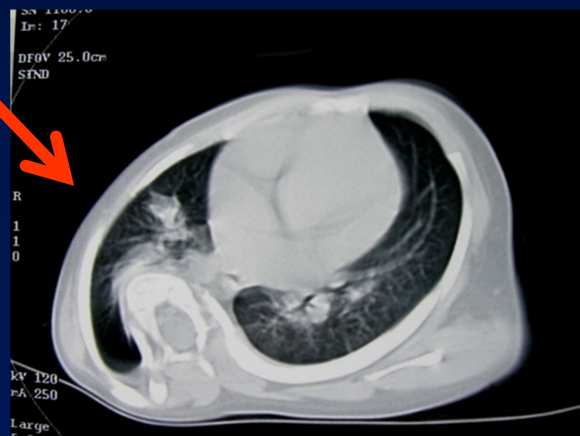
Dynamic Lung MRI and Spine Deformity



Chu, WCW, et al.
-SRS 2005
Kotani, T. et al.
Spine, 2004.



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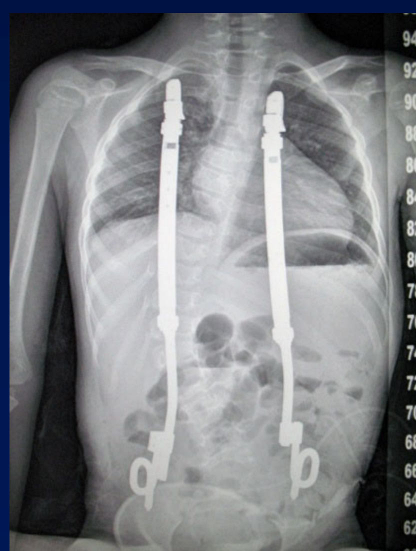
Dynamic Lung MRI (Campbell, et al.)

Pre-op



Post op

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My Goal: Recognize, measure, and effectively treat Thoracic Insufficiency Syndrome

- Do what it takes to have the largest, most symmetrical, most functional thorax by skeletal maturity

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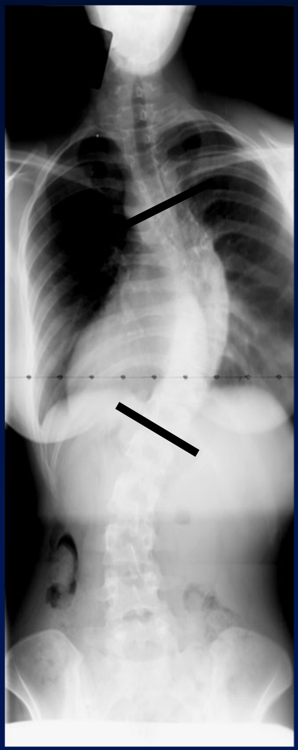
We need Courage



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Thank You

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