# Pulmonary Consequences of Spine and Chest Wall Abnormalities in Young Childhood

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# Chest Wall/Spine/Lung Interactions

The spine dictates rib function:
Kyphoscoliosis alters rib alignment and mobility

The ribs dictate spine function:
Fused ribs lead to scoliosis

The lungs dictate rib and spine function:
Corrected congenital diaphragmatic hernia produces scoliosis (18% of patients).

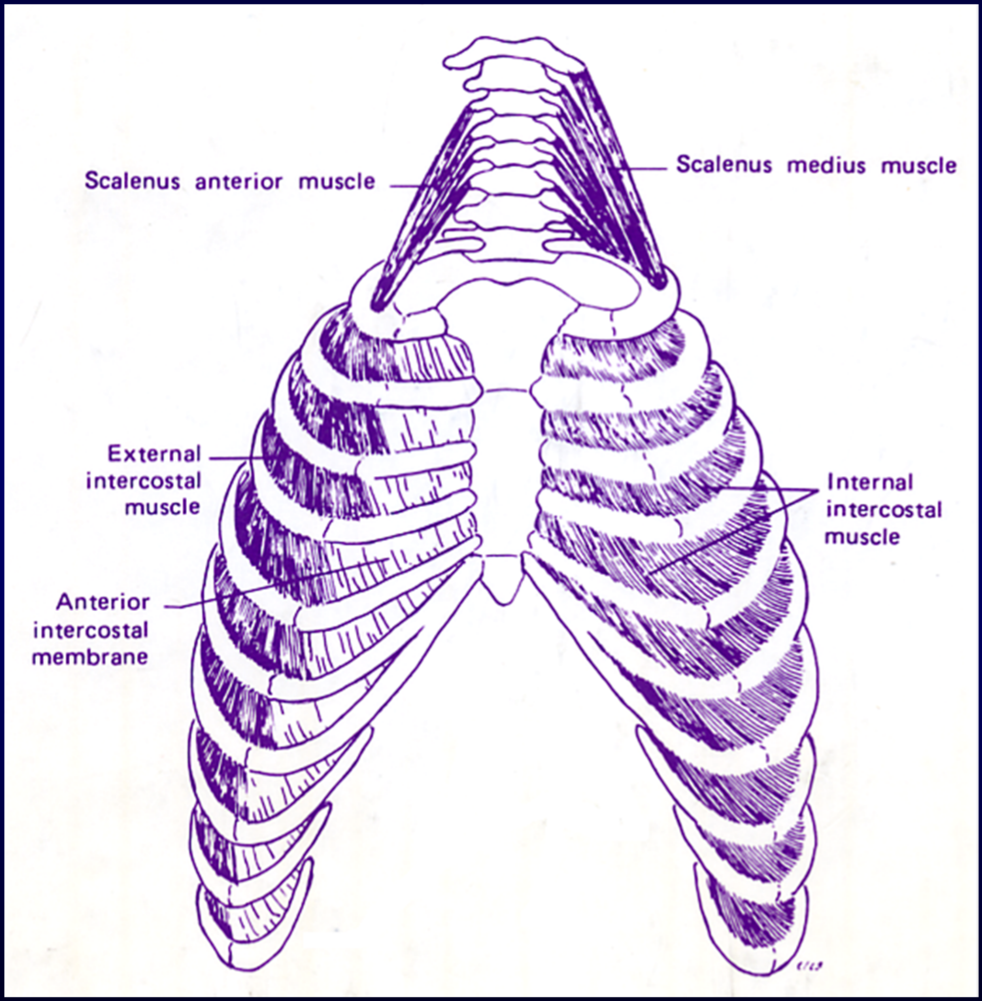
THE RIBS AND SPINE DICTATE LUNG FUNCTION

# Thoracic Anatomy and Composition

Bones: Spine
Ribs

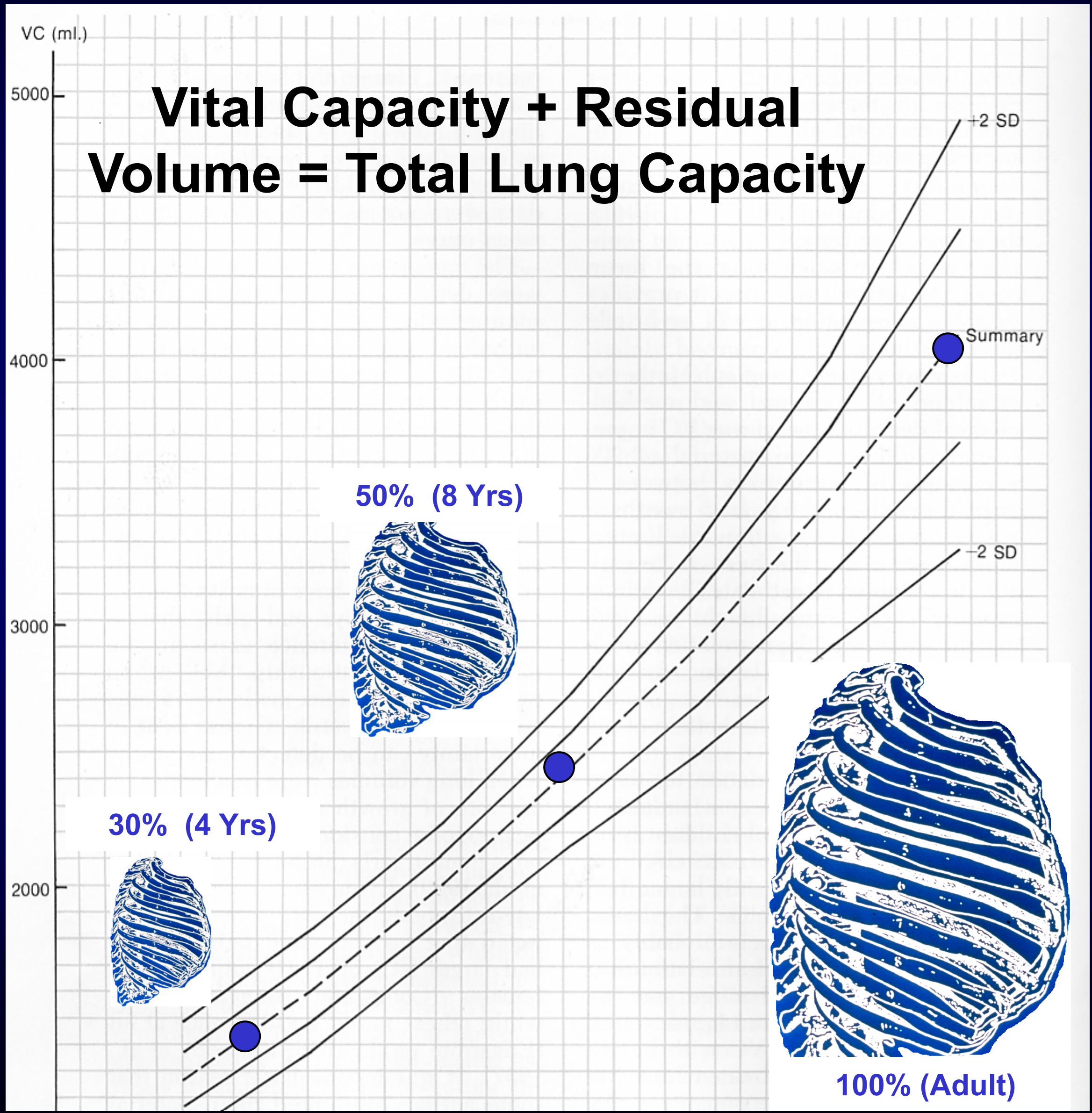
Muscles: Chest, Neck,
Abdomen, and
Diaphragm

Alterations in size
Alterations in configuration
Alterations during growth




Roussos C, Macklem PT. In: The Thorax (vol 29):Marcel Dekker, Inc., 1984

# Lung Volumes Change with Age



## Thoracic Insufficiency Syndrome (TIS)

**FDA DEFINITION: TIS is the inability of the thorax to support normal respiration or lung growth.**

*Assumption:*

*There is no inherent lung disease (e.g. unilateral hypoplasia) that is affecting spine/chest wall growth.*

| | |
|---|---|
| *Primary TIS*: | primary chest wall disorder |
| *Secondary TIS*: | a chest wall disorder, such as scoliosis, due to neuromuscular conditions (weakness or spasticity) affecting the respiratory function |
| *Acquired TIS*: | post-operative chest surgery (rib resection for tumors, Siamese twin separation, etc) |

# Classification of Primary TIS

Type I: Absent Ribs and scoliosis

- Absence of ribs, congenital scoliosis

Type II: Fused Ribs and scoliosis

- Congenital scoliosis with fused ribs

Type IIIa: Foreshortened thorax

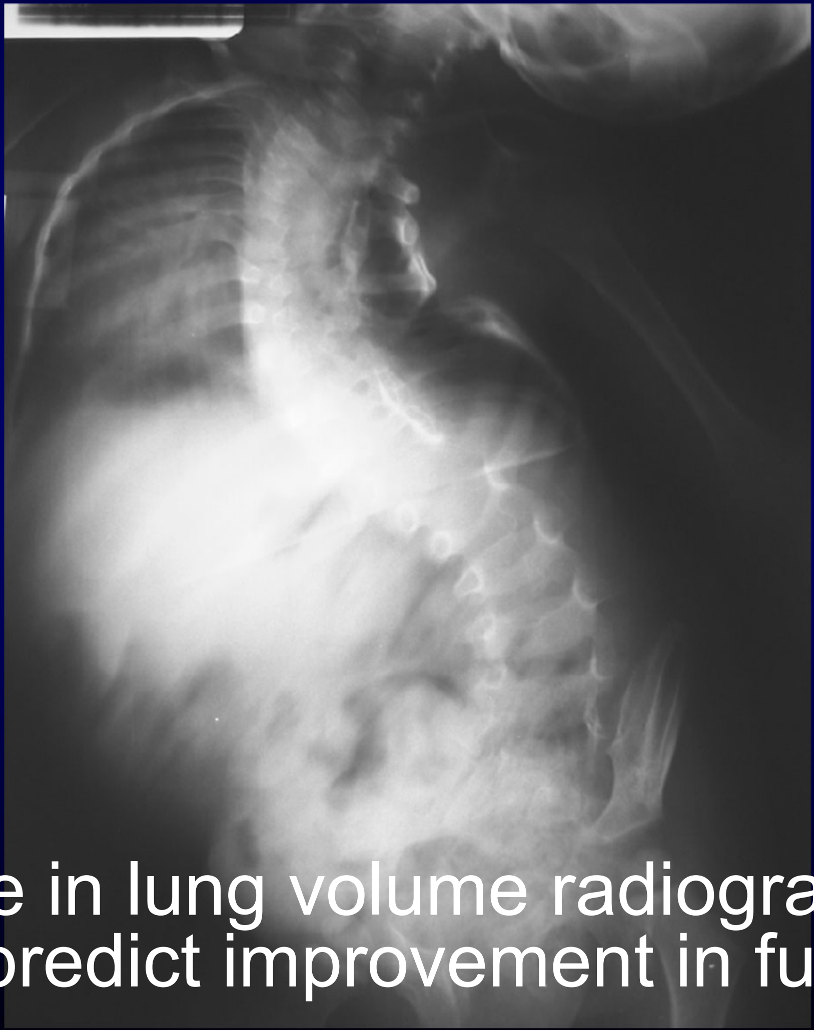
- Jarcho-Levin syndrome

Type IIIb: Transverse constricted thorax

- Jeune’s aphyxiating thoracic dystropy

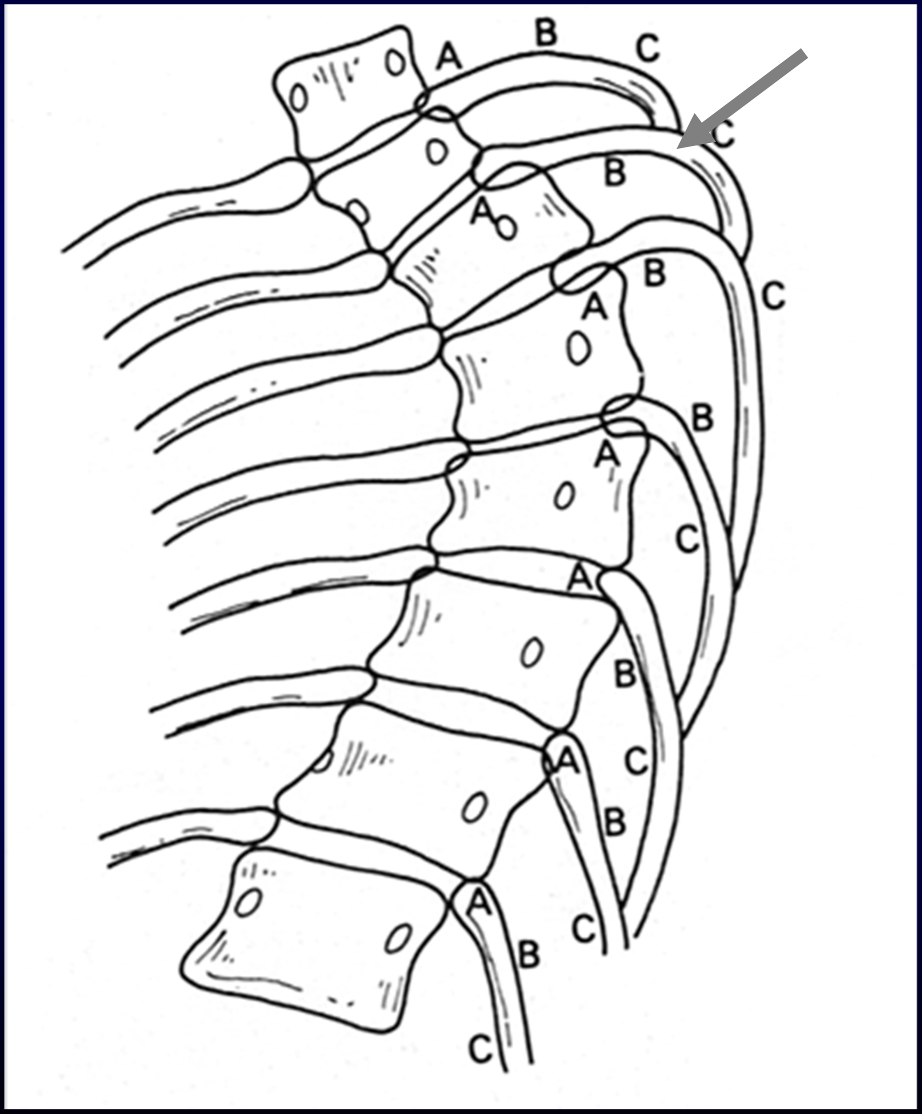
Campbell RM, Jr., Smith MD. *J Bone Joint Surg Am* 89:Suppl 1:108-22, 2007

**Kyphoscoliosis**: two lungs surrounded by chest walls with different shapes, sizes, and respiratory muscle configuration which interact.



An increase in lung volume radiographically does not predict improvement in function.

# Abnormal Rib-vertebral Alignment and Motion +/- Abnormal Intercostal Tissue in Scoliosis




Roaf, 1977

# Effects of Spine Rotation in Lung Size and Shape

# Pulmonary Outcomes of Kyphoscoliosis

- Restrictive Respiratory Mechanics
- Loss of Chest Wall Excursion
- Asymmetric Loss of Lung Function
- Inefficient Diaphragm Function

# Restrictive Respiratory Disease

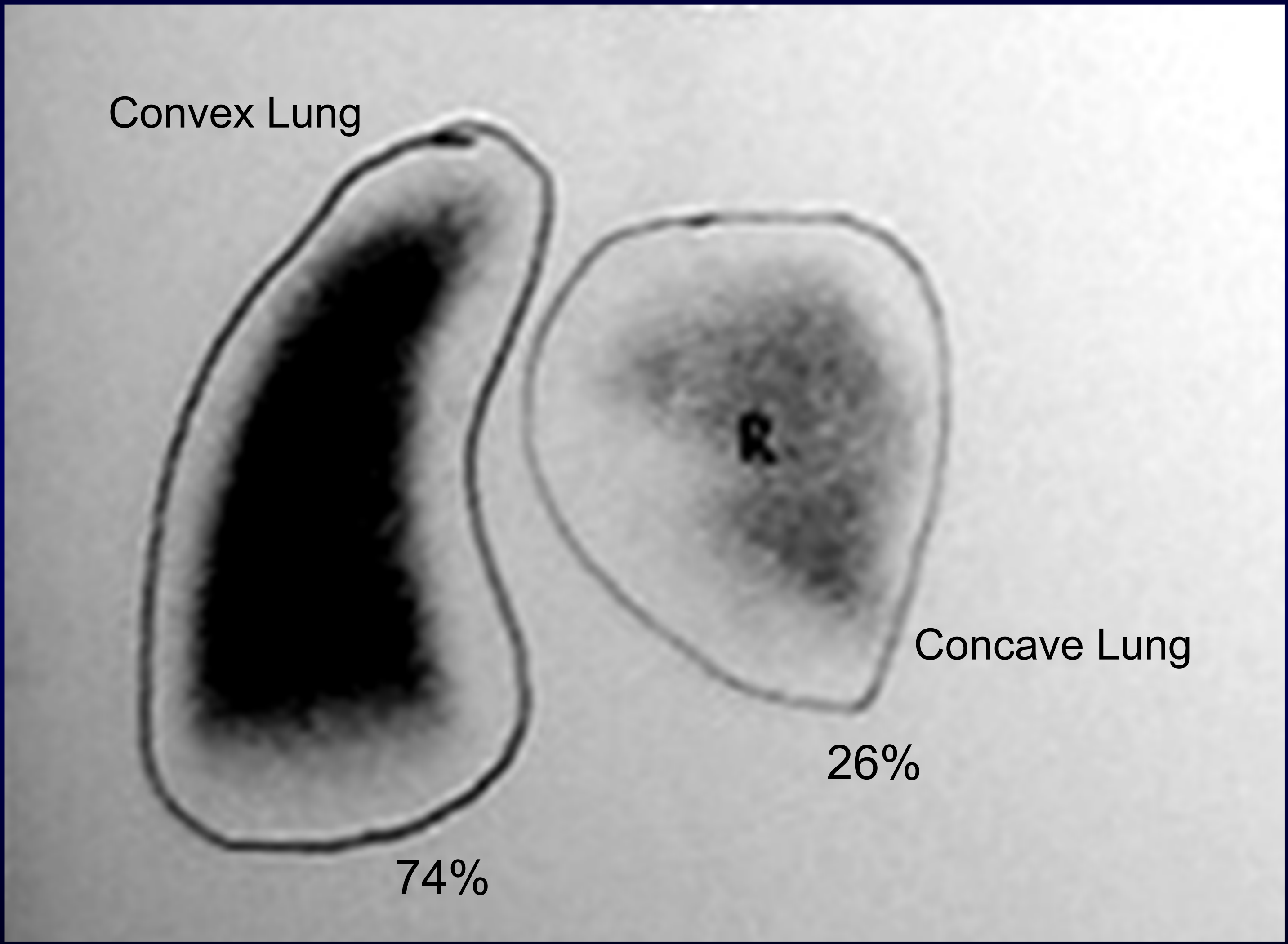
- Loss of lung volume and lung distensibility
- Loss of rib mobility and normal chest wall expansion with inspiration
- Increased reliance on diaphragm function as the primary muscle of inspiration

# Pre-operative FVC Values by Diagnosis

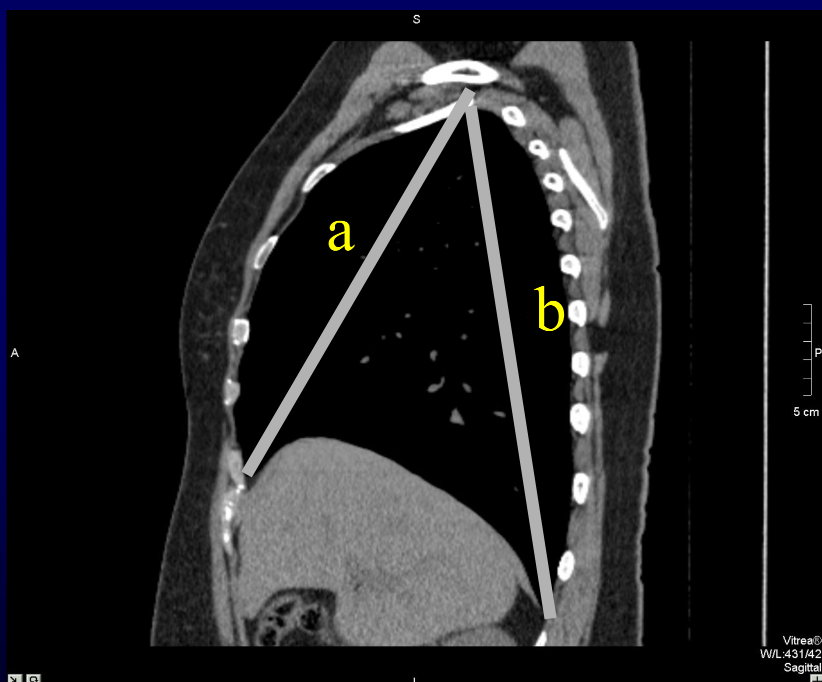
| | HT | RF | PS | FC |
|---|---|---|---|---|
| *n* | 8 | 14 | 17 | 2 |
| *Age* | 11.5 | 10.1 | 7.9 | 10.9 |
| Median | 55% | 56% | 67% | 47% |
| Range | 26-85% | 36-115% | 38-136% | 33-61% |
| # of with Normal FVC** | 1 (12%) | 1 (8%) | 5/17 (29%) | 0/2 |

* % of predicted by height; ** FVC > 80% predicted

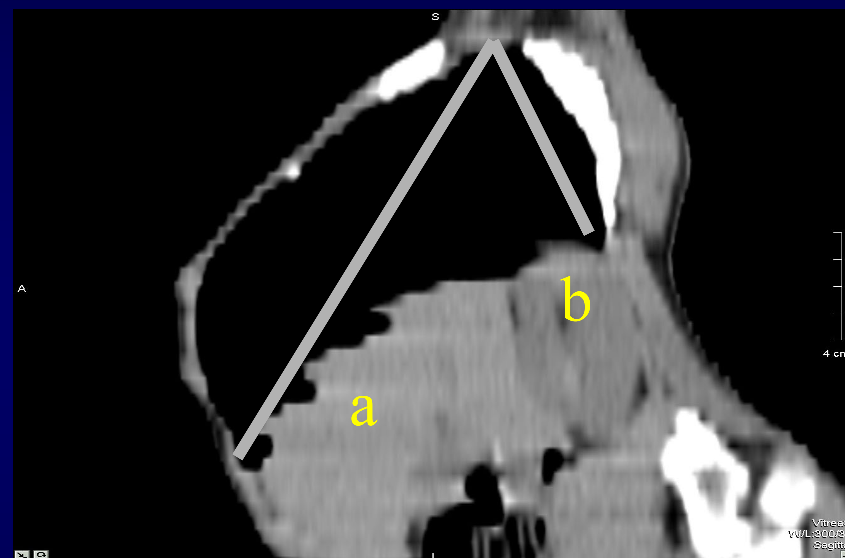
# Lung Perfusion Scan in Kyphoscoliosis



# Diaphragm contour in Jarcho-Levin Syndrome



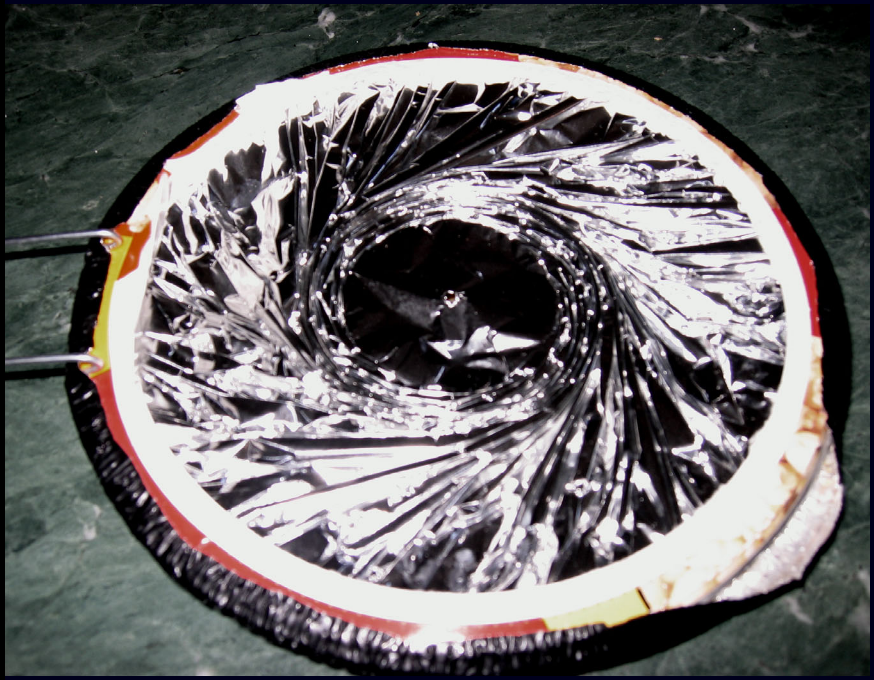

Normal sagital costophrenic depth ratio

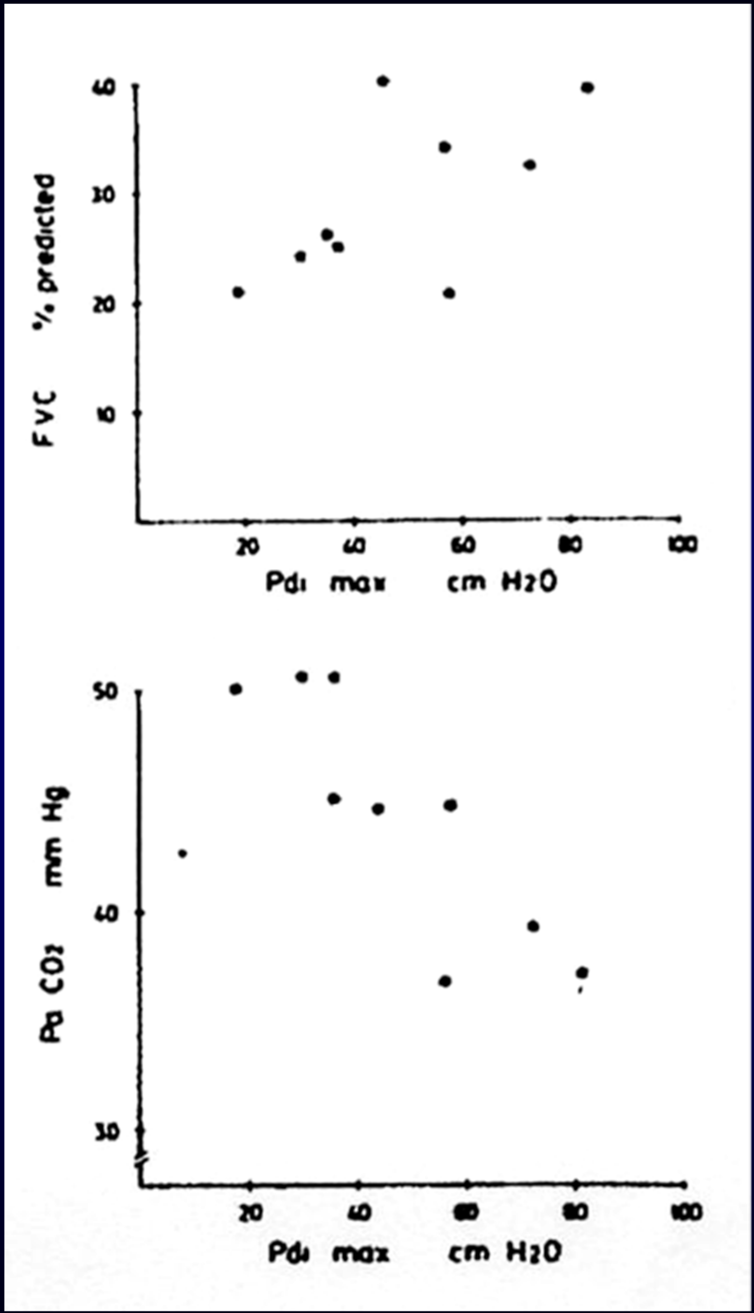



Abnormal sagital costophrenic depth ratio in Spondylothoracic dysplasia.

Ramirez, et al. JBJS, in review

# Spine Rotation → Effects on Diaphragm Function?






# Loss of Diaphragm Force Correlates with Respiratory Failure in Adults

Moreno LC, et al. *Am Rev Respir Dis* 1985; 132(1):48-52

# Inter-relationships of Adverse Pulmonary Outcomes

Restrictive Lung Mechanics

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Asymmetric Lung Function → Progression of Chest Wall Disease → Progressive Loss of Functional Reserve

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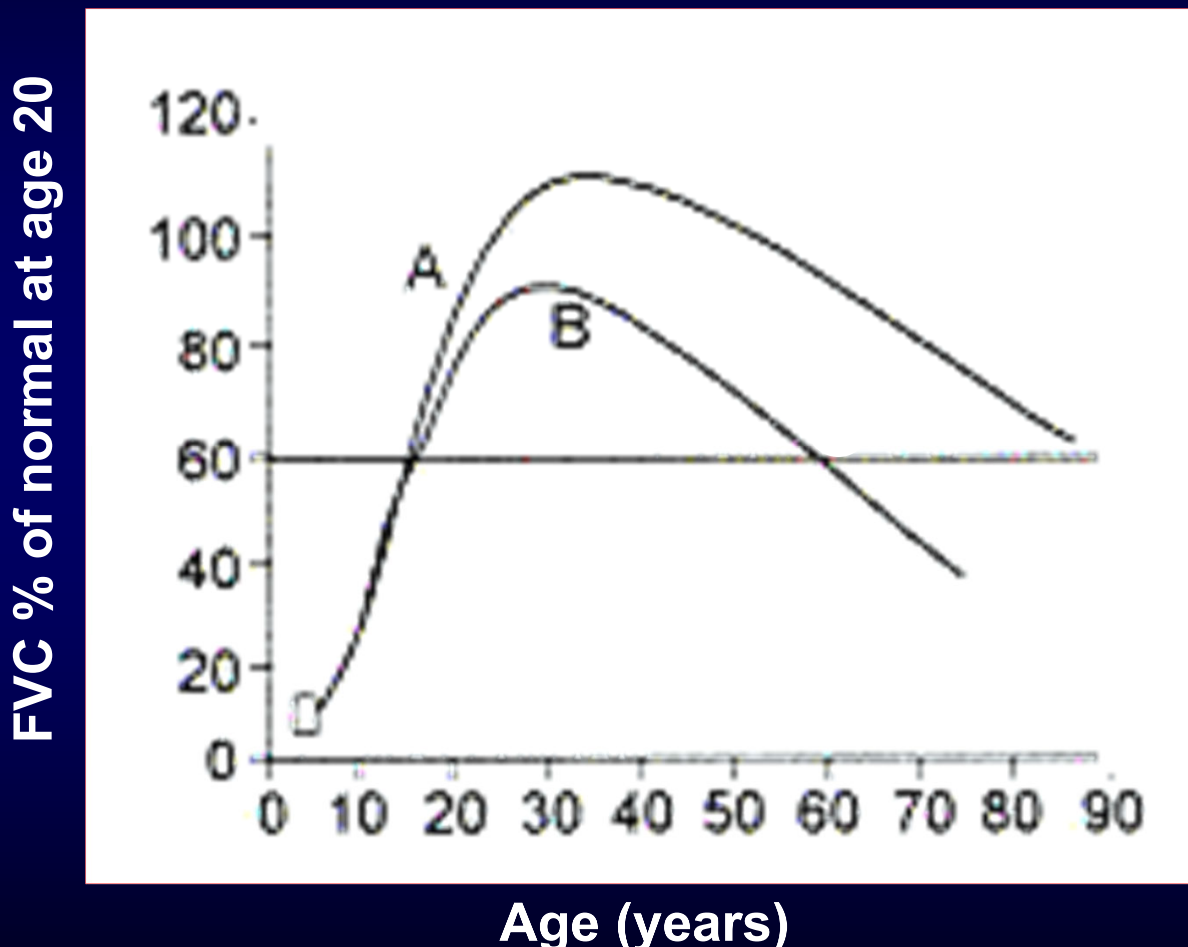
Diaphragm Inefficiency

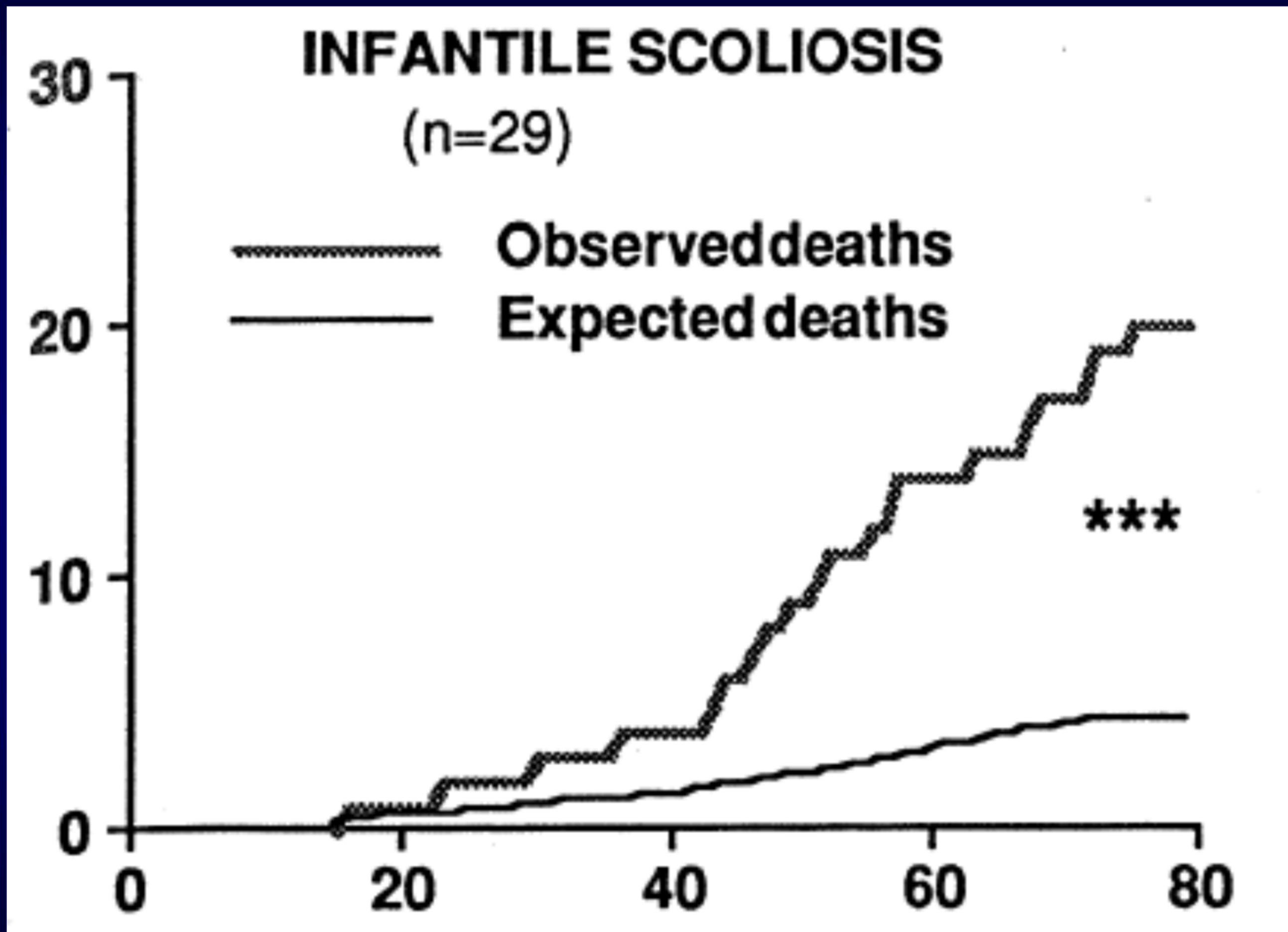
Increased Mortality

# Consequences of Childhood Restrictive Lung/Chest Wall Disease




**A:** Failure to Match Maximum Best Value

**B.** Rapid Decline to At Risk Values with Age




Pehrsson K. et al. *Spine* 1992; 17:1091-96


McMaster MJ. *Spine* 1998; 23:998-1005