Pulmonary Consequences of Thoracic Insufficiency Syndrome

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#### **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 disorderSecondary TIS:a chest wall disorder, such as scoliosis, due to<br/>neuromuscular conditions (weakness or spasticity)<br/>affecting the respiratory functionAcquired TIS:post-operative chest surgery (rib resection, etc.)

## **Classification of Primary TIS**

Type I:Absent Ribs and Scoliosis- Absence of ribs, congenital scoliosisType II:Fused Ribs and Scoliosis- Congenital scoliosis with fused ribs

Type IIIa: Foreshortened thorax- Jarcho-Levin syndromeType 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 together.



## Characteristic Changes in Spirometry Due to TIS



## **Pre-operative FVC Values by Diagnosis**

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

predicted by height: \*\* FVC > 80% predicted

**Conditions Requiring Respiratory Reserve** 

- Aerobic Exercise (>6 years of age)
- Sleep (all ages)
- Illness

# Sleep-Related Changes in Breathing\*

- Reduced ventilatory responses to hypoxemia and hypercapnia
- Reduced muscle tone in upper airways and intercostals
- Reduced sigh frequency
- Increased cough threshold
- Awakening is a defense mechanism in response to hypoxemia, hypercapnia, and increased work of breathing
- \*Worse in Rapid-Eye Movement (REM) Sleep

## **Apnea-Hypopnea in Children with TIS During Sleep**



#### Nadir O<sub>2</sub> vs. AHI

r=0.78, *p* < 0.005

#### **Regional Lung Volumes Dictate Regional Ventilation: Role of Rotation?**



## Lung Perfusion Scan in Kyphoscoliosis



# Unpredictable Distribution of Lung Function



Redding G, et al. Spine J 8:639-644, 2007

#### **Dynamic MRI: Change in Chest Wall and Diaphragm Configuration**



Cluzel, P. et al. *Radiology* 2000;215:574-583

### Changes in Diaphragm Strength and Movement relate to Contour and Orientation of Fibers



Normal sagital costophrenic depth ratio



Abnormal sagital costophrenic depth ratio in Spondylothoracic dysplasia.



Loss of Diaphragm Force Correlates with Respiratory Failure in Adults

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

#### Frequency of Respiratory Abnormalities Associated with TIS and Useful Testing Methods



## Consequences of Childhood Restrictive Lung/Chest Wall Disease

FVC % of normal at age 20



A: Failure to Match Maximum Best Value

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

Age (years)