Pulmonary Consequences of TIS and EOS

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Thoracic Insufficiency Syndrome

Neuromuscular Disease Hypoplastic Thorax

Early Onset Scoliosis Flail Chest Syndromes





Spine and Thorax Deformities

Low lung volumes

Poor chest wall distensibility & excursion

Thoracic distortion & reduced regional lung function

<u>New</u> Idea...



Spine and Thoraco-Abdominal Deformities

Low lung Poor chest wall Thoracic distortion & Abnormal Abdominal volumes distensibility & reduced regional lung diaphragm position distensibility excursion function and movement and muscle tone?

Lung Volumes Before and 6 Months After VEPTR Insertion



Mayer OH, Redding GJ Pediatr Orthop 29(1):35-38, 2009

Sleep-related Breathing Problems in Children with TIS

<u>Apnea-Hypopnea Index</u> (NL<2/hr) TIS without weakness = 4/hr (2-11/hr) TIS with weakness = 11/hr (2-40/hr)

<u>Arousal Index</u> (NL<10/hr) TIS without weakness = 20/hr (3-52hr)

AHI worse if TIS occurs with weakness or obesity*



nadir O2 vs. AHI

Regional Lung Distortion, Volume, and Function









3-D CT Lung Volume vs Lung Perfusion and Ventilation Prior to Surgery



 $N=1\overline{1}$

Song K, et al. POSNA 2009

Altered Diaphragm Insertion, and Configuration Lead to Reduced Movement and Force Generation



Subpulmonic Contours



Dynamic MRI: Movements of the Diaphragm and Chest Wall





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

Inter-relationships of Adverse Pulmonary Outcomes





Correlations Between Lung Function Measures and Cobb Angle are Poor

n=39 r=0.14 p=NS

n=11 r=0.16 p=NS

Mayer OH, et al. J Pediatr Ortho 29:35-38, 2009 Redding GJ, et al. Spine J 8:639-644, 2007 Striegl A. American Thoracic Society (ATS), 2008 Children with TIS have severe, early onset, and progressive spine and chest wall deformities with years of somatic growth ahead



Chest Wall Constraints: Influence on Alveolar Development



Olson JC, Kurek KC, Mehta HP, et al. Evaluation of Pulmonary Cellular Response to Treatment of thoracic Insufficiency Syndrome Using Expansion Thoracoplasty in Scoliotic Rabbit Model

Consequences of Childhood Restrictive Lung/Chest Wall Disease



Age (years)

Concluding Remarks

- Early onset scoliosis results in loss of respiratory reserve which can worsen over time.
- How changes in lung function parallel progression in deformities is unclear.
- We do not have clear thresholds for intervention based on lung function values or trends.
- We have clearer ideas about how breathing is compromised and how this impacts growth and sleep, and to a lesser extent, daily activities. We know less about how poor lung functions influence longevity and quality of life in children with EOS.

Structural Features of Kyphoscoliosis: An Evolving 3-Dimensional Disorder

Scoliosis (Cobb) Kyphosis Rotation **Rib** alignment Progression Lordosis Distortion, e.g. rib hump

Lung Functions Before and After Serial VEPTR Expansions

N=24

Age at intervention = 4.6 years (1.8-11 years) Interval of Studies = 2.7 years (1-5.6 years) # Expansions = 2-12

	Pre-op	Post-op	% Change	P value
FVC (% of initial value in ml)	NR	NR	11 ± 10%	<.001
FVC as % predicted	$72\pm22\%$	66 ± 16%	-7 ± 2%	<.05
Crs (ml/cmH2O/kg)	1.2 ± 0.5	0.65 ± 0.3	$-44\pm22\%$	<.001
(NI Crs=6-20 ml/cmH2O)				
$ND = \dots + \dots + \dots$				

NR = not reported

Motoyama E, et al. Paediatric Respiratory Reviews 10:12-17, 2009