

# **Pulmonary Evaluation of Infants and Children with Congenital Chest Wall and Spine Deformities**

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## The Pulmonary Evaluation Assesses:

1. Severity at the time of initial encounter
2. Progression of disease
3. Effect of intervention (VEPTR implantation and expansion)

## Serial Pulmonary Assessments May Help Determine:

1. When to intervene
2. Short and long-term benefits of interventions

# Thoracic Structure and Function

Scoliosis

Kyphosis

Rotation

Distortion



Work of  
Breathing

Chest Wall  
Movement

Asymmetric  
Ventilation

Respiratory  
Muscle  
Function

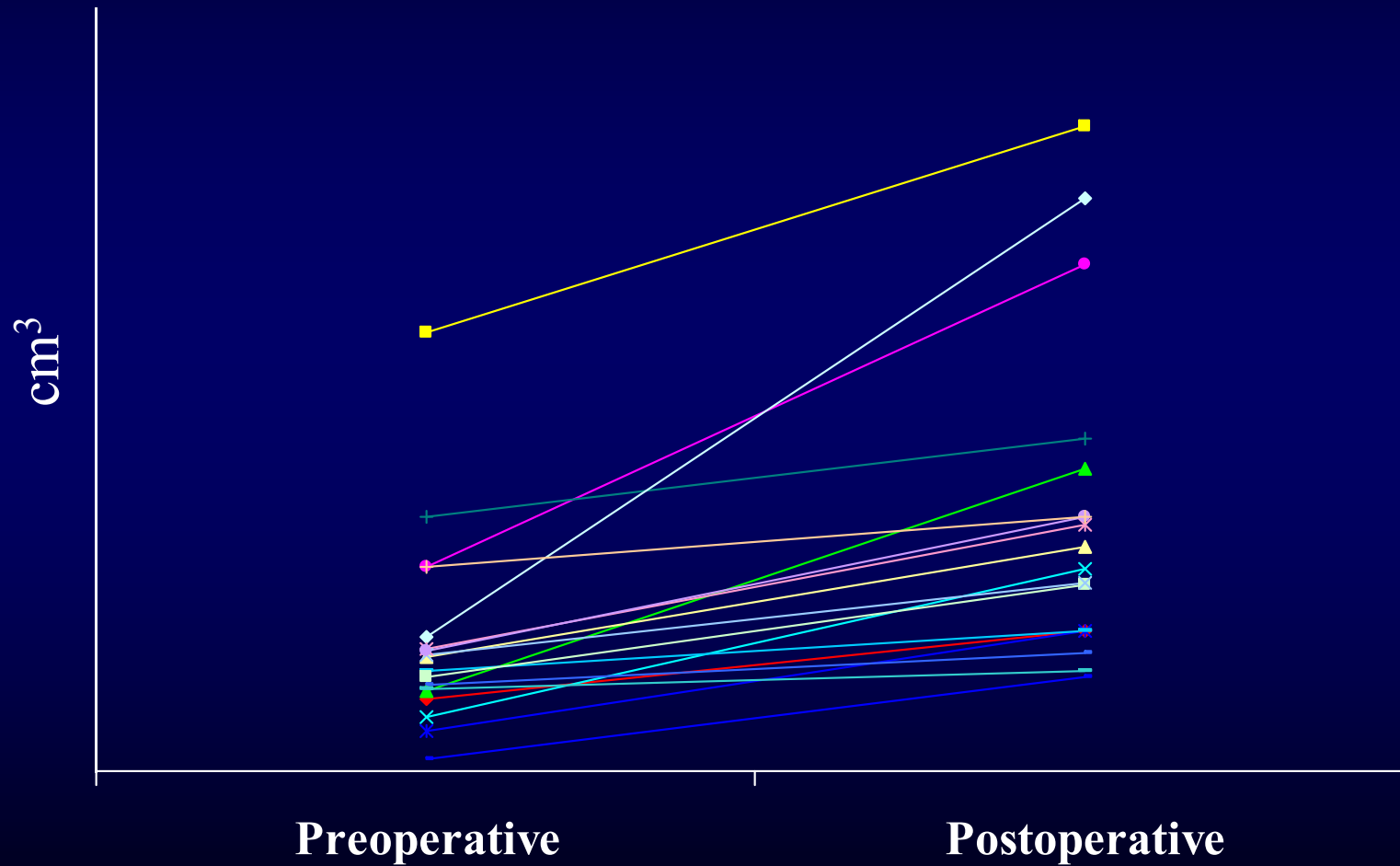
# Conceptual Pulmonary Outcomes of Kyphoscoliosis

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

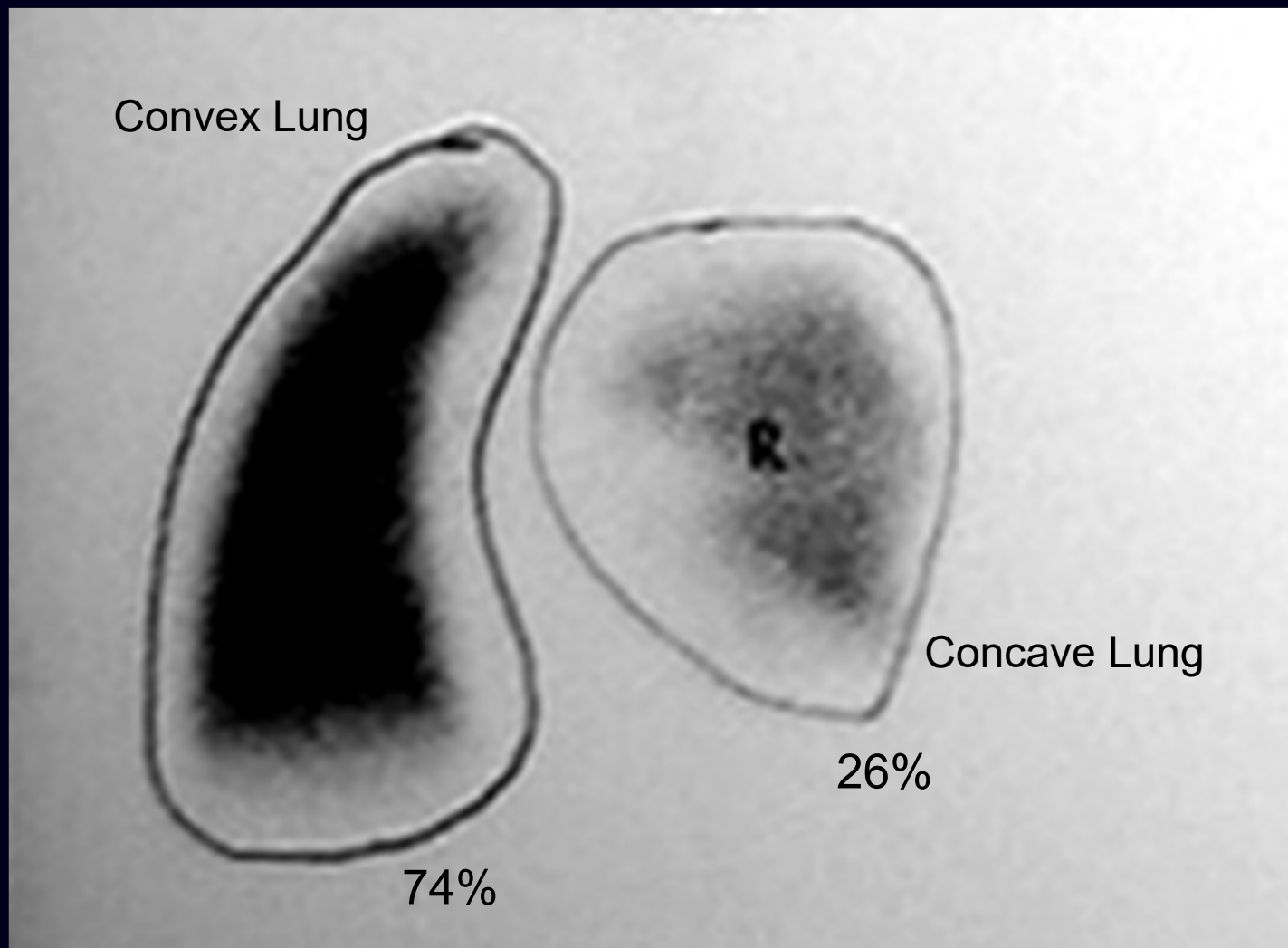
# Pulmonary Evaluation Techniques in Children

Test	Index	Age of Useful Measurement
<u>Respiratory rate</u>	Respiratory rate	All ages (age-specific norms)
<u>Oxygenation</u>	SaO <sub>2</sub> in room air	All ages
<u>Ventilation</u>	CO <sub>2</sub> (capillary) or HCO <sub>3</sub>	All ages
<u>Spirometry</u>	Forced vital capacity (FVC)	5-6 years (using arm span)
Respiratory muscle strength	Maximal inspiratory and expiratory pressure	6-8 years
Exercise tolerance	Maximal oxygen consumption, external work, ventilation	8 years
	6 minute walk?	5 years
<u>Echocardiogram</u>	Estimated pulmonary artery pressure Right ventricular wall thickness	All ages (may need sedation)
Ventilation scan	Right vs. left ventilation Homogeneity of ventilation	5-6 years
<u>Perfusion scan</u>	Right vs. left lung blood flow	Any age (may need sedation)

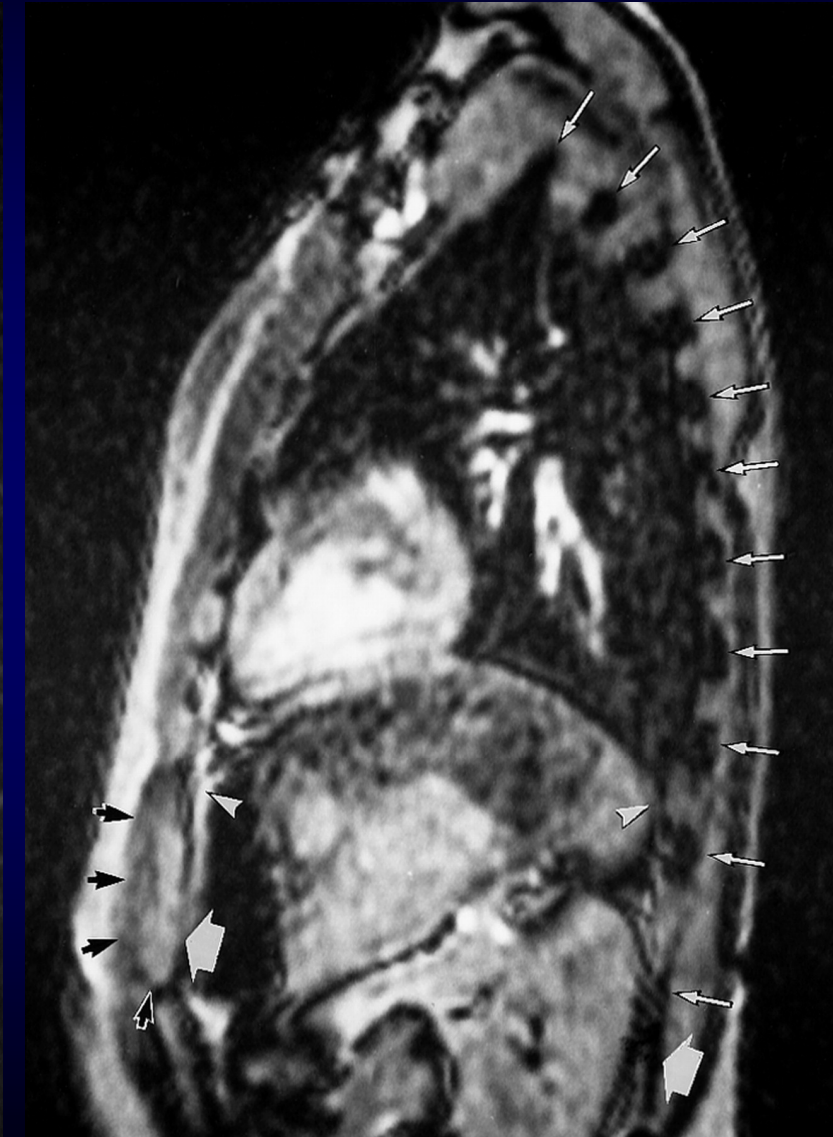
# Total Lung Volume by CT Scan 2 Years After Expansion Thoracoplastasty



# Lung Perfusion Scan in Kyphoscoliosis

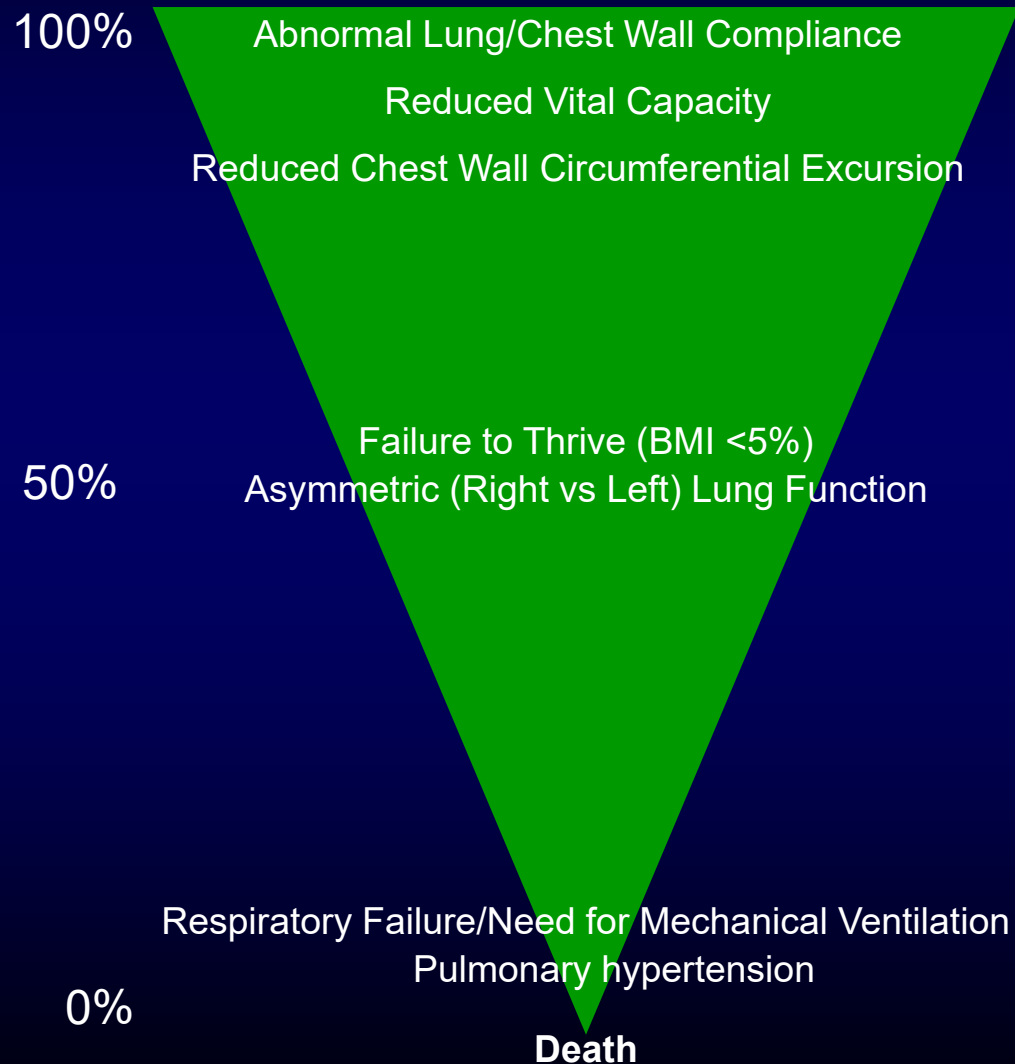


# Dynamic MRI: Change in Chest Wall and Diaphragm Configuration





# Frequency of Respiratory Abnormalities Associated with Congenital and Infantile Scoliosis



# TIS Severity Score: Evolving Functional Considerations\*

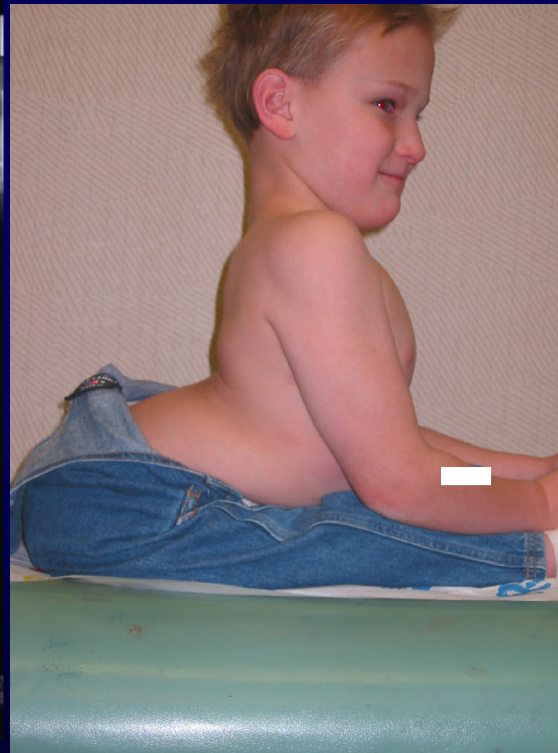
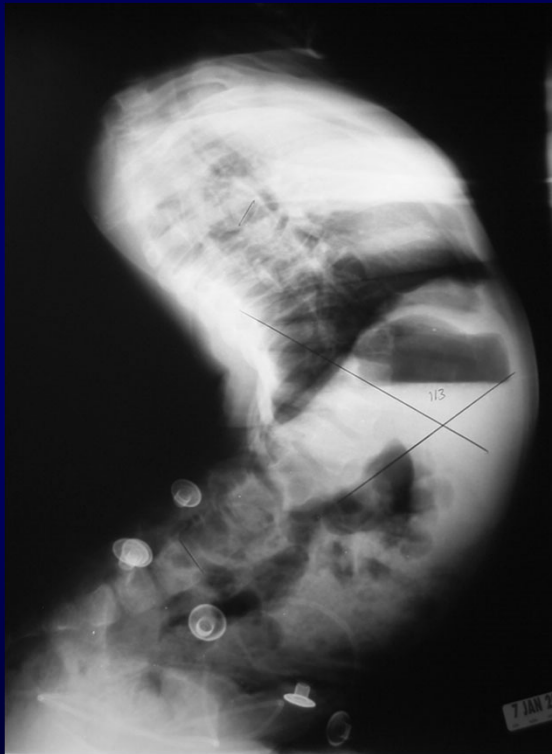
Severe: Ventilator Dependent  
CO<sub>2</sub> Retention  
Cor Pulmonale  
Vital Capacity <50%

Moderate: Failure to Thrive  
Asymmetric Lung Function  
Vital Capacity 50-70%  
Tachypnea at Rest

Mild: Vital Capacity >70%  
Exercise Limitation

*\*Excluding Neuromuscular Weakness Conditions*

# Neuromuscular Weakness and Secondary Thoracic Insufficiency Syndrome

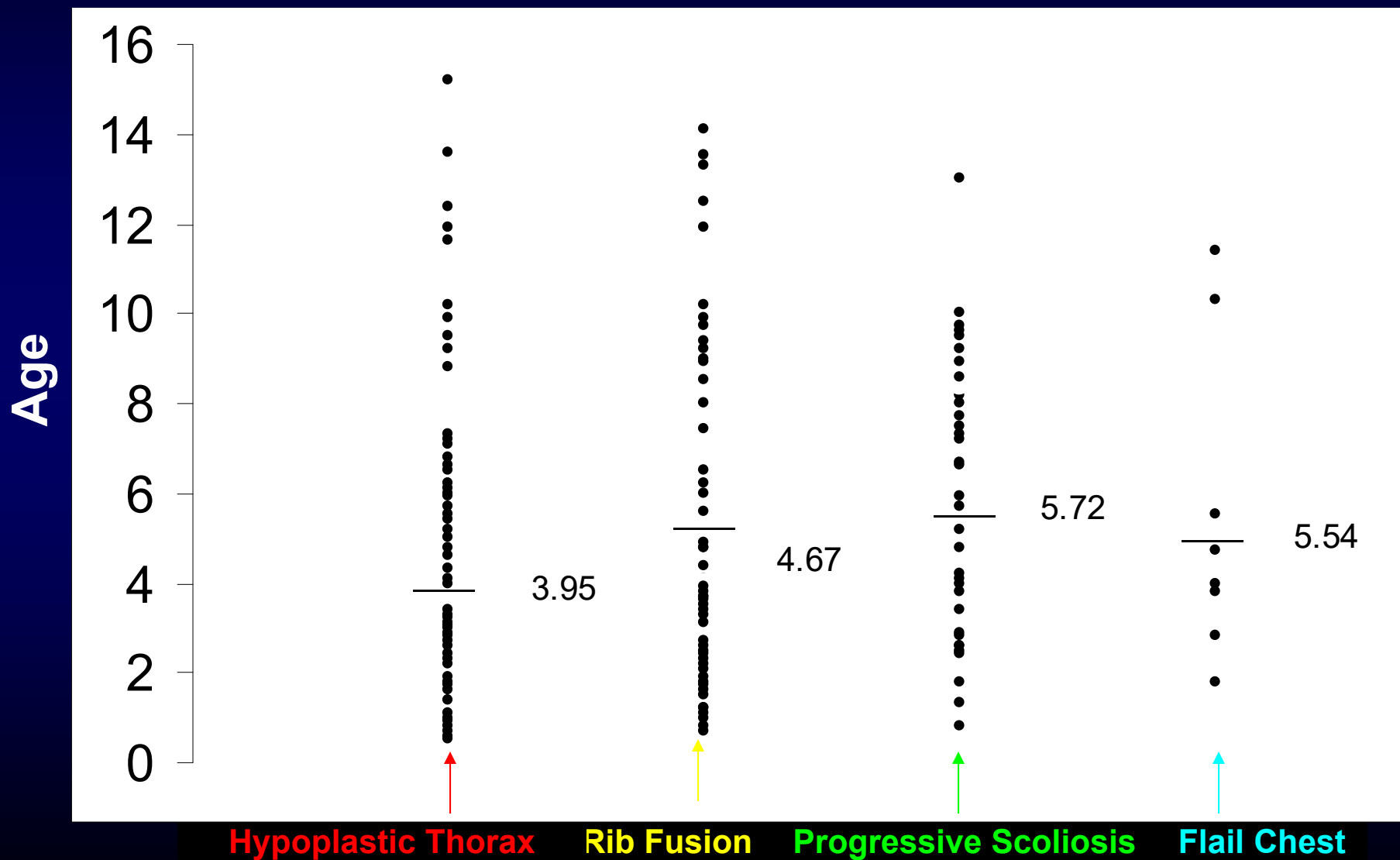


↓FVC, ↓FRC

Hypoxemia and CO<sub>2</sub> retention during sleep

Decreased muscle strength and cough

# Age Distribution for TIS Subjects at ET by Diagnosis



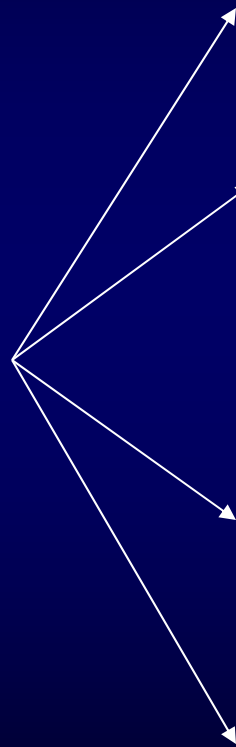
# Pulmonary Function Testing in Infants and Children

	Invasive	Non-Invasive
Effort-- Dependent*	Trans-Diaphragm Pressures	Spirometry Respiratory Muscle Strength and Endurance
Effort-- Independent	Chest Wall & Lung Compliance Lung volumes	Oximetry, Blood Gas Tensions, Perfusion Scan, Sleep Study, Echocardiogram

\*Voluntary Effort at 5-6 Years of Age

# Pulmonary Outcomes of Surgical Interventions for TIS

**PRESENTING  
STATUS**



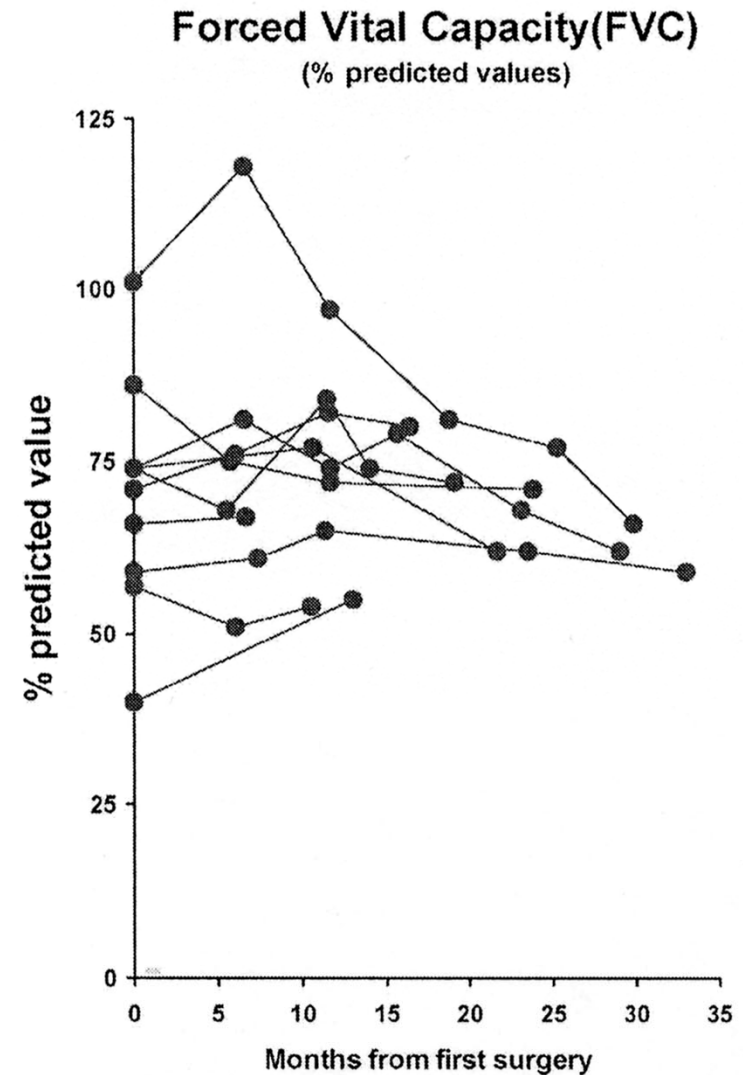
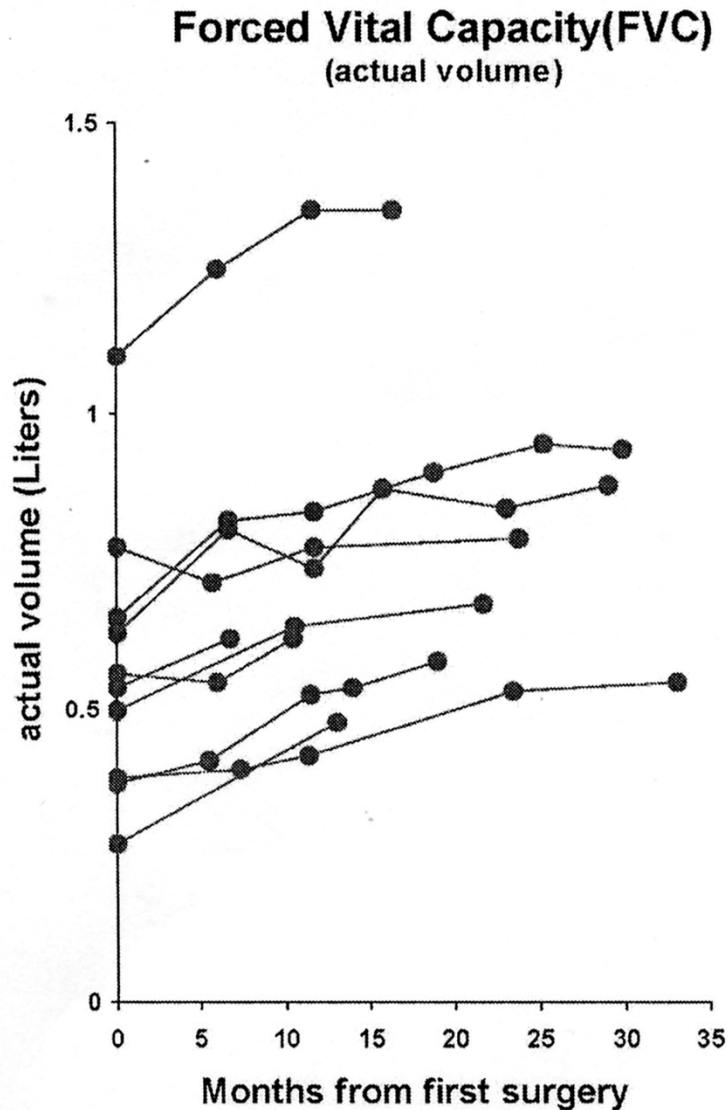
Progression

Halt in Progression but  
no further growth

Improvement  
Proportional to Growth

Catch-up Improvement  
Over Time

# Serial Lung Functions Following Initial Expansion Thoracoplasty



# Summary

- Specific and standardized criteria to intervene surgically in children with Thoracic Insufficiency Syndrome do not exist.
- Functional criteria tend to identify and/or confirm the loss of pulmonary reserve suggested by structural abnormalities of the spine and thoracic cage.
- Serial correlations of structure and functional indices among one another will help to determine when surgical intervention is ideally timed.