What Do We Know about Pulmonary Effects of Growing Rods and VEPTR Therapies for Children with TIS

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Pulmonary Features to Assess in Children with TIS

- Loss of rib mobility and chest wall distensibility; increased work of breathing and caloric expenditure; decreased exercise tolerance
- Loss of both regional and global lung volume
- Altered diaphragm position, movement, and function
- Poor sleep quality and failure to thrive
- Pulmonary hypertension and respiratory failure

The pulmonary consequences of surgical intervention depend on multiple factors:

1. Severity and age pre-operatively
2. Extra-pulmonary features of disease, e.g. neuromuscular weakness, developmental delay, cardiac disease
3. Type of TIS category (scoliosis, hypoplastic chest, flail chest)
4. Time of assessment post-operatively
5. Type of intervention (Growing Rods vs VEPTR vs other)

Respiratory Function Outcomes of Surgical Interventions

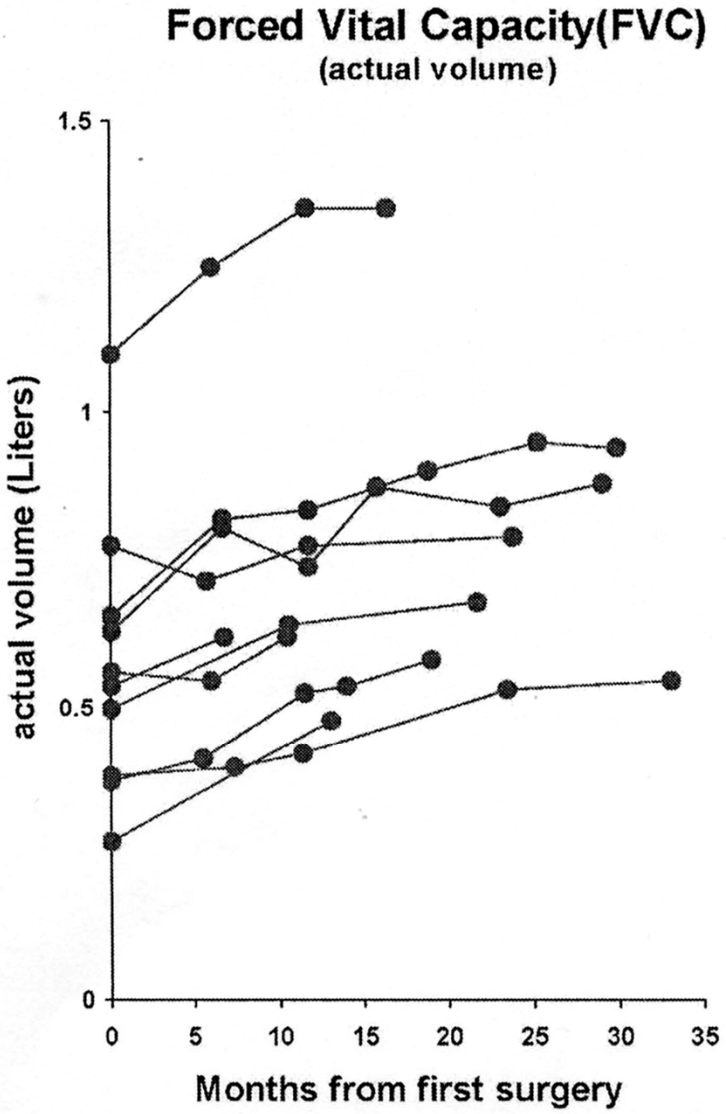
Influenced by:

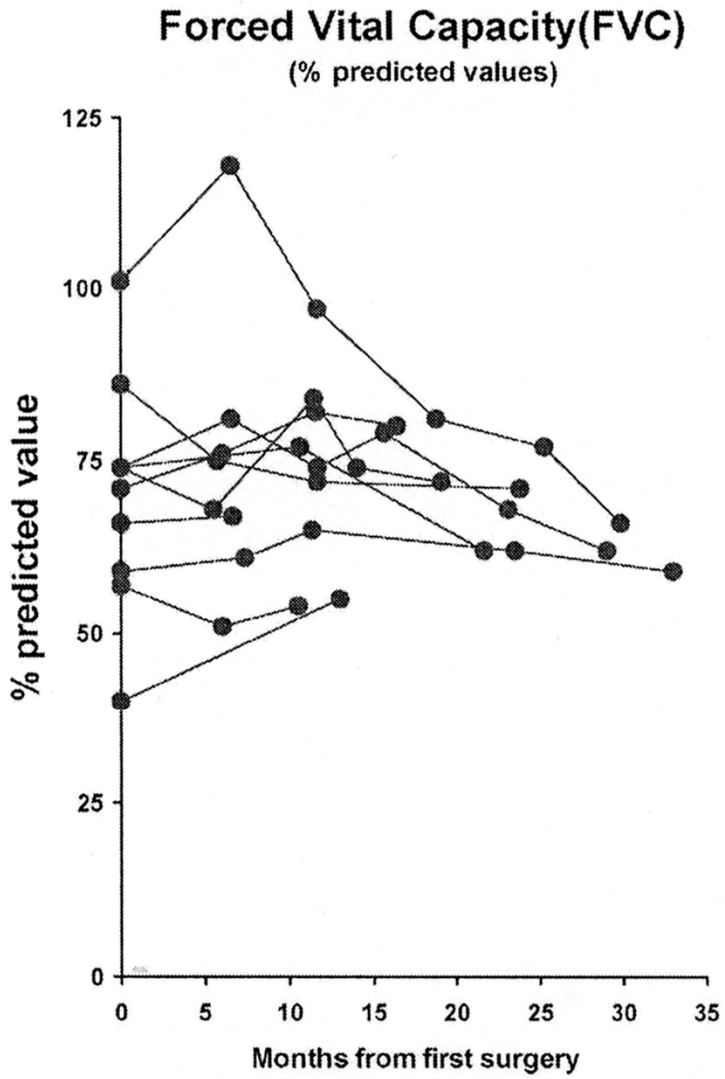
- Conditions of measurement, active vs passive efforts
- Position during testing, i.e. upright vs supine, awake or asleep
- Limited testing techniques in young children

- VEPTR and Expansion Thoracoplasty: Pulmonary Outcomes
- -Not the obvious outcome indices, e.g. spirometry
- -Heterogeneous responses
- -Time course of improvement after surgery

- Growing Rod Interventions
  - Same outcomes?
  - No data
  - Plans for direct comparisons?

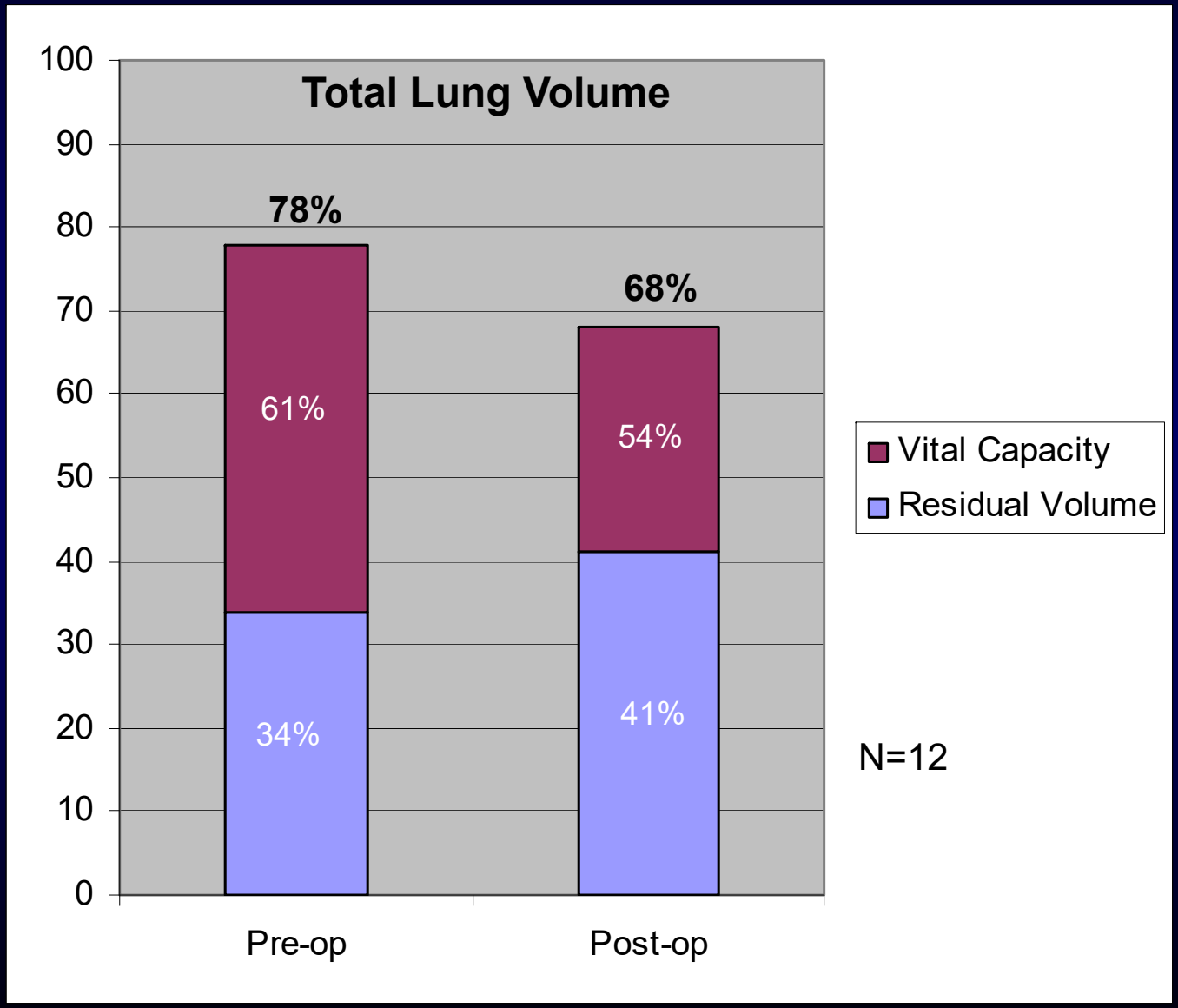
Serial Lung Functions Following Initial Expansion Thoracoplasty





Motoyama EK, et al. *Spine* 31:284-290

Lung Volumes Before and After VEPTR in Older Children

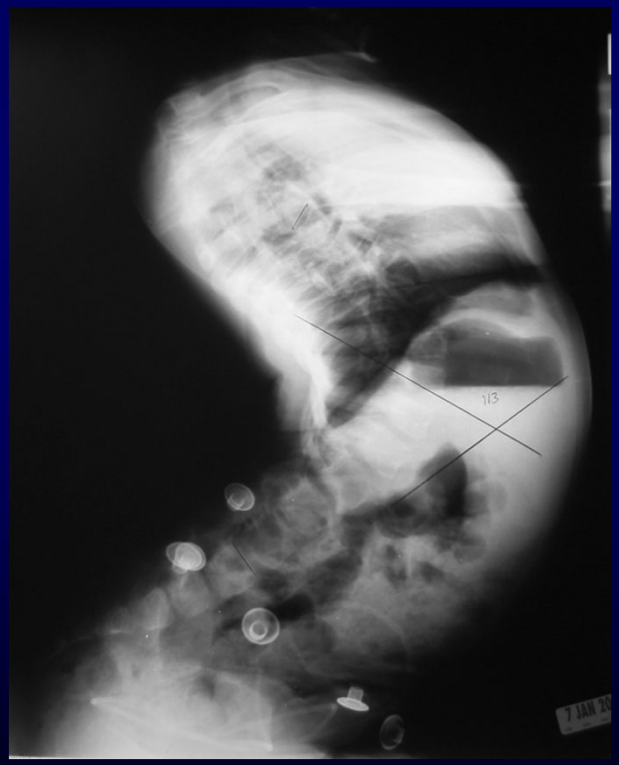


Mayer O, Redding G, et al. *J Pediatric Orthopedics,* 2008

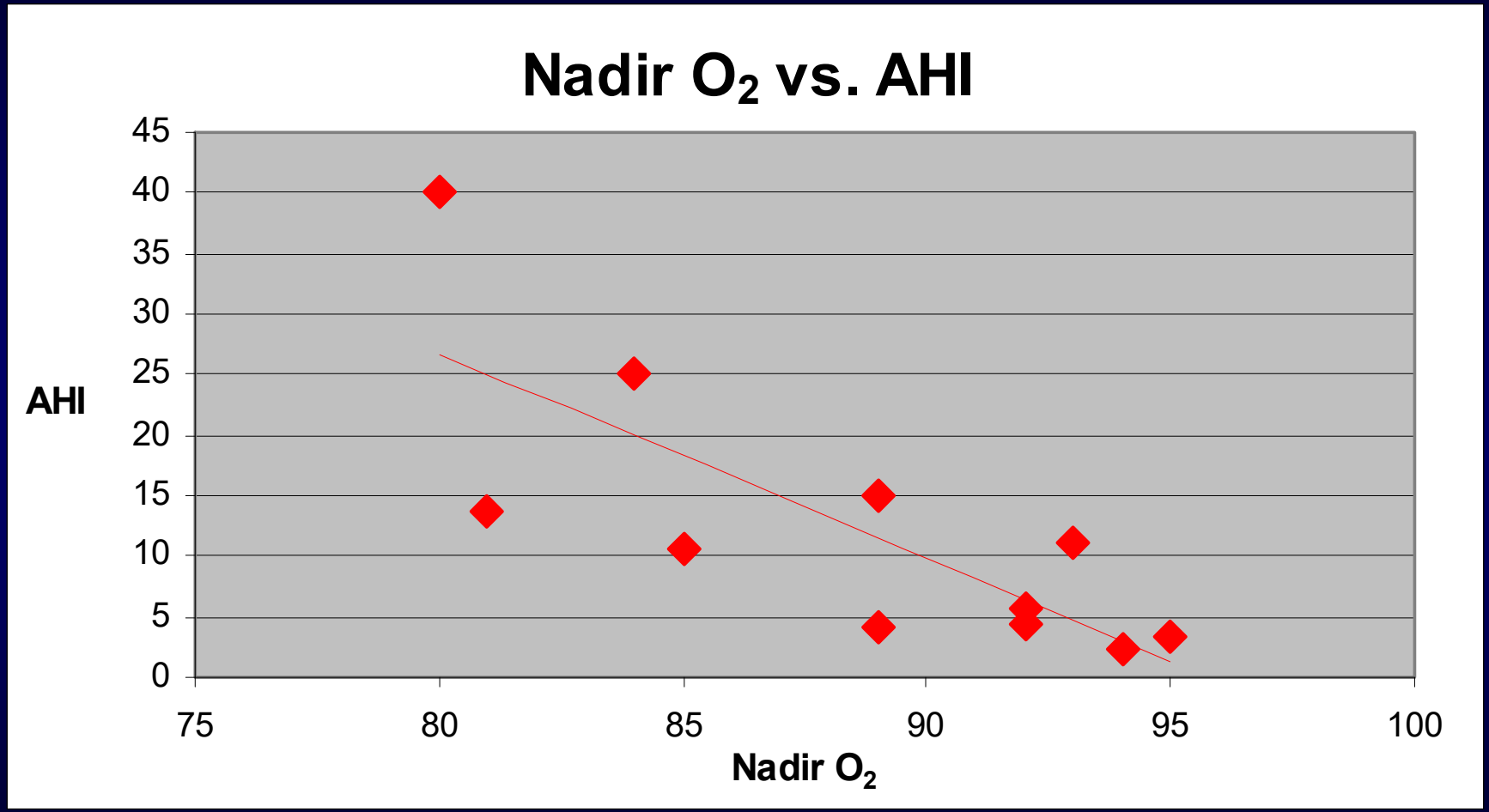
Secondary Thoracic Insufficiency Syndrome

Collapsing Neuromuscular Scoliosis/Myelodysplasia

- *With Chest deformity* and collapse into the abdomen



Apnea-Hypopnea in Children with TIS During Sleep

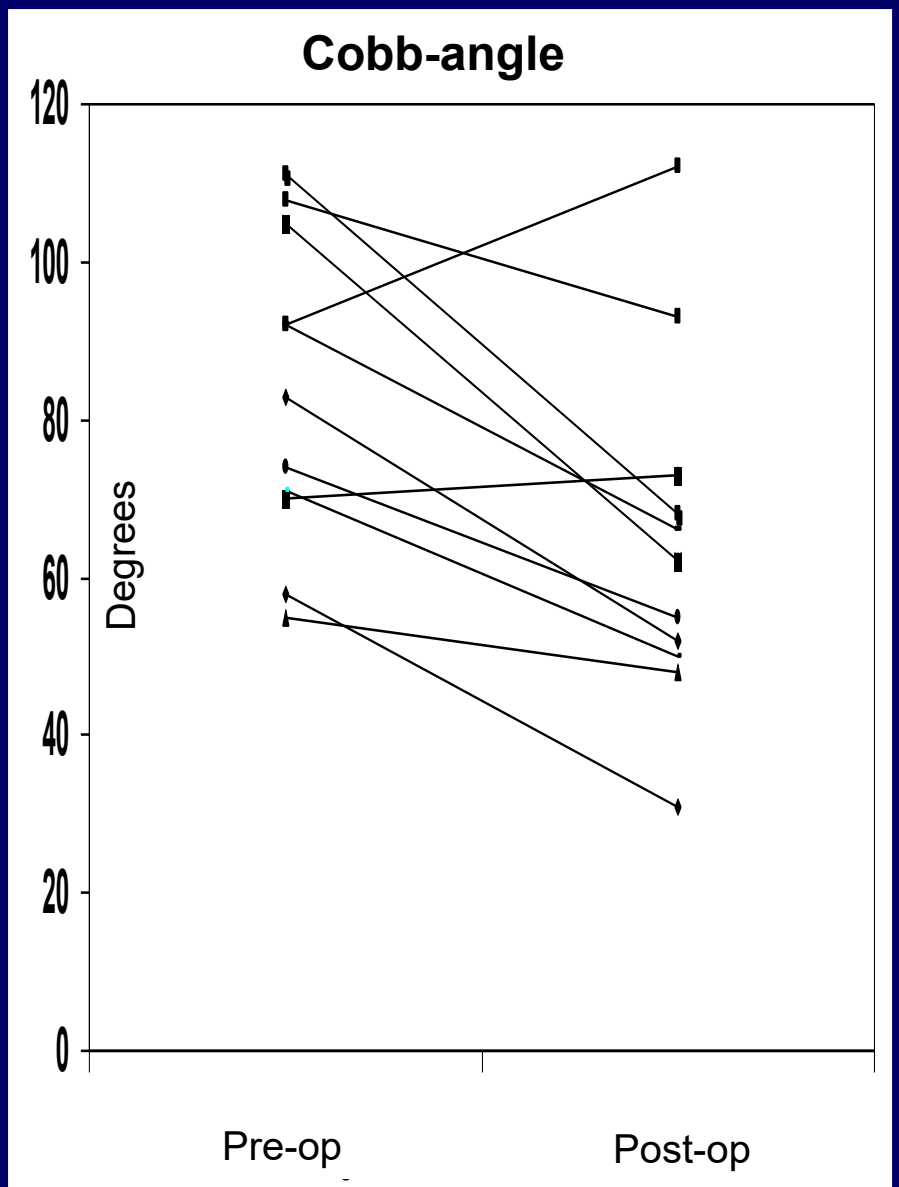


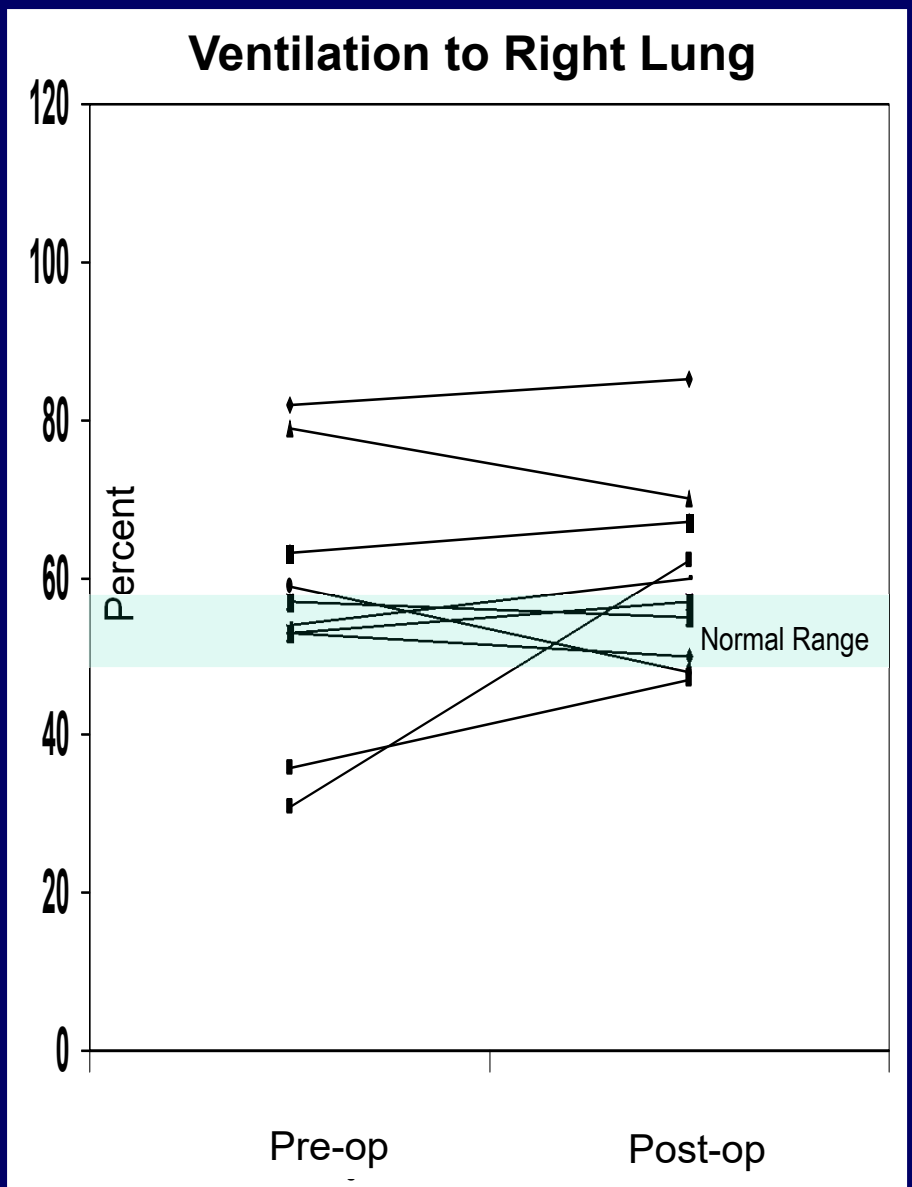
r=0.78, $p < 0.005$

Left Lung		Right Lung
114077.5	Total Counts	66041.8
63.3	Percent	36.7
63.3	1 Subregion % 2	36.7

Lung Distribution

Left Lung		Right Lung
137231.5	Total Counts	48125.0
74.0	Percent	26.0
74.0	1 Subregion % 2	26.0


Cobb-angle
120
100
80
60
40
20
0
Degrees
Pre-op
Post-op


Ventilation to Right Lung
120
100
80
60
40
20
0
Percent
Normal Range
Pre-op
Post-op

Changes Following Expansion Thoracoplasty by Diagnosis

	Discontinuation of Ventilation
Overall	10/35 (33%)
Hypoplastic Thorax	6/28 (21%)
Other Diagnosis	4/7 (57%)

**Time to initiation ventilator support post-operatively: mean = 29 months, range = 19-48 mo.*

Changes Following Expansion Thoracoplasty by Diagnosis

	Resolution of Hypercapnea	Appearance of Hypercapnea
Overall	14/21 (67%)	10/193 (5%)
Hypoplastic Thorax	9/14 (64%)	6/73 (8%)
Other Diagnosis	5/7 (71%)	4/120 (3%)

Summary of Pulmonary Changes with Expansion Thoracoplasty and VETPR Treatment

Documented Changes

- No change in vital capacity
- Increased residual volume
- Unpredictable change in distribution of right and left lung function
- Reduction in $PaCO_2$ (if elevated pre-op)
- Discontinuation of mechanical ventilation in a minority

Features to be Studied

- Resting energy expenditure and growth rate
- Sleep quality
- Respiratory muscle strength and endurance
- Regional chest wall excursion?

Summary

There is no comparative pulmonary function data on different surgical interventions that alter chest wall and spine configuration.

Vital capacity changes little after surgical therapy. Increased residual volume provides a better gas reservoir, especially during sleep, may reduce hypoxemia and frequency of arousals. This in turn might reduce caloric expenditure at night which will improve growth velocity after treatment.

A majority of patients with respiratory failure ventilate better with reduced CO2 levels but few will improve enough to become ventilator free if on this therapy pre-operatively.