Pulmonary Outcomes after VEPTR Intervention

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Disclosures

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Spine and Thoracic Cage Deformities

Low Lung Volumes

Hypoxemia
Poor sleep
Cor pulmonale

Reduced Chest Wall Distensibility and Excursion

Increased Work

Poor Growth

Reduced Respiratory Muscle Force and Movement

Reduced Exercise Tolerance

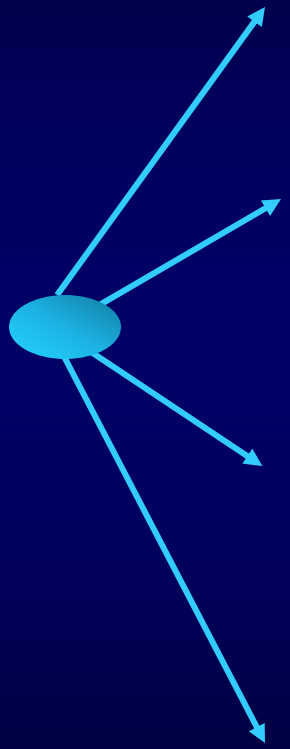
Respiratory Failure

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

Outcomes of Surgical Interventions for TIS

PRESENTING STATUS



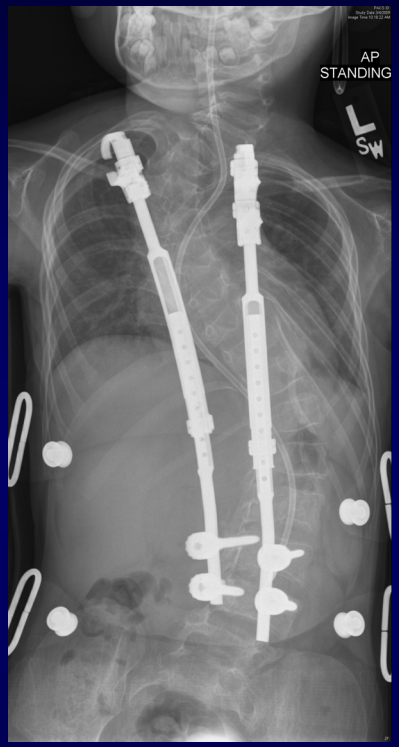
Ongoing Progression

Halt in Progression but no further growth

Improvement Proportional to Growth

Catch-up Improvement Over Time

Structural Outcomes of Surgical Spine/Thoracic Treatments

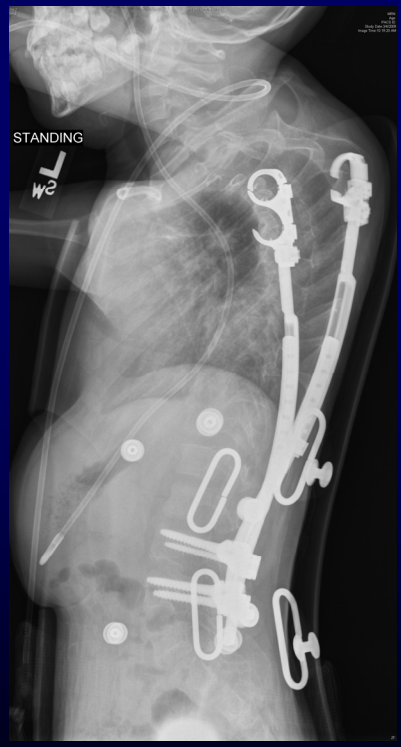


Gradual Changes

- Increased Annual Vertebral Body Growth Velocity
- Increased Thoracic Height

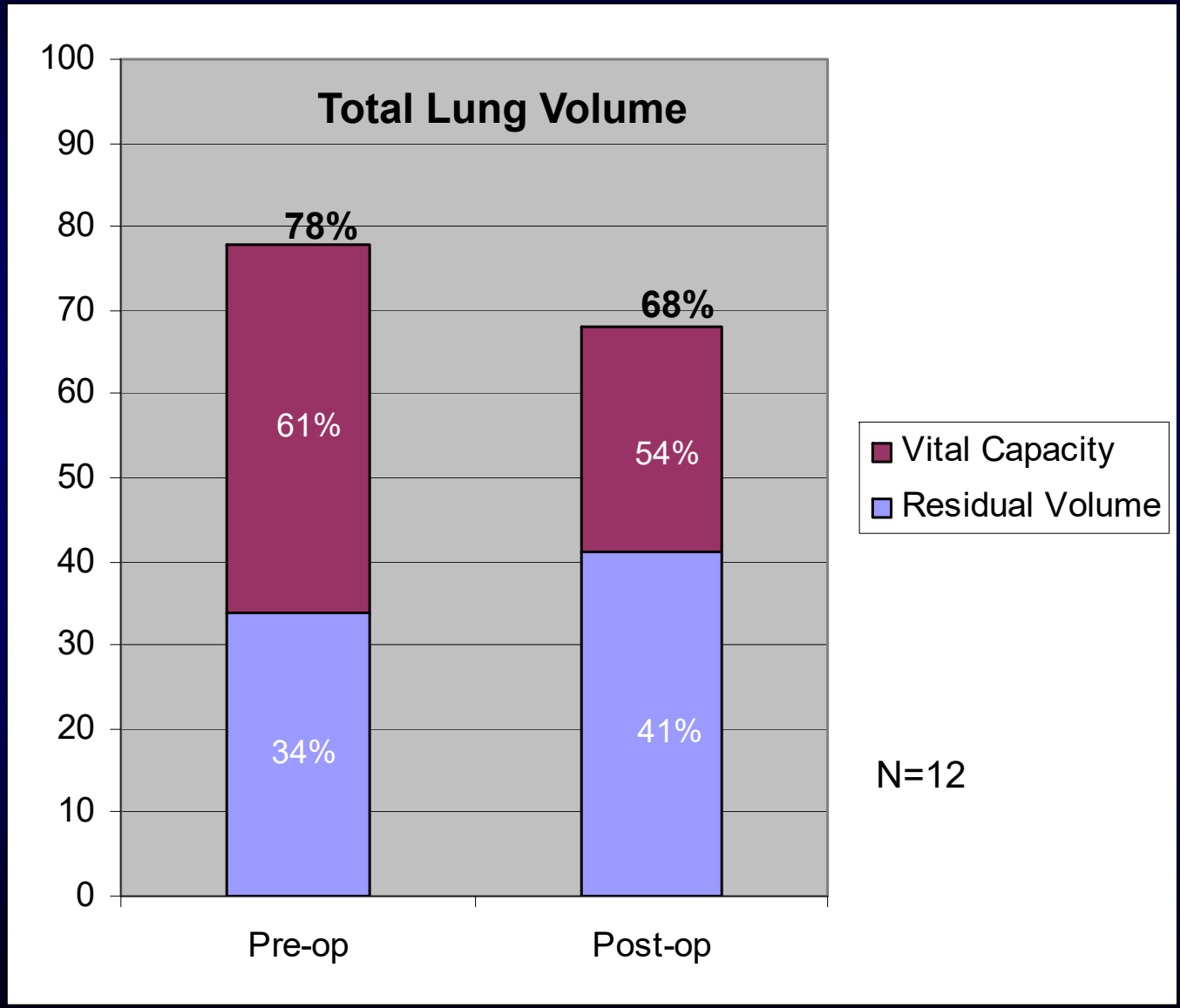
Immediate Changes

- Reduced Cobb Angle
- Reduced Kyphosis



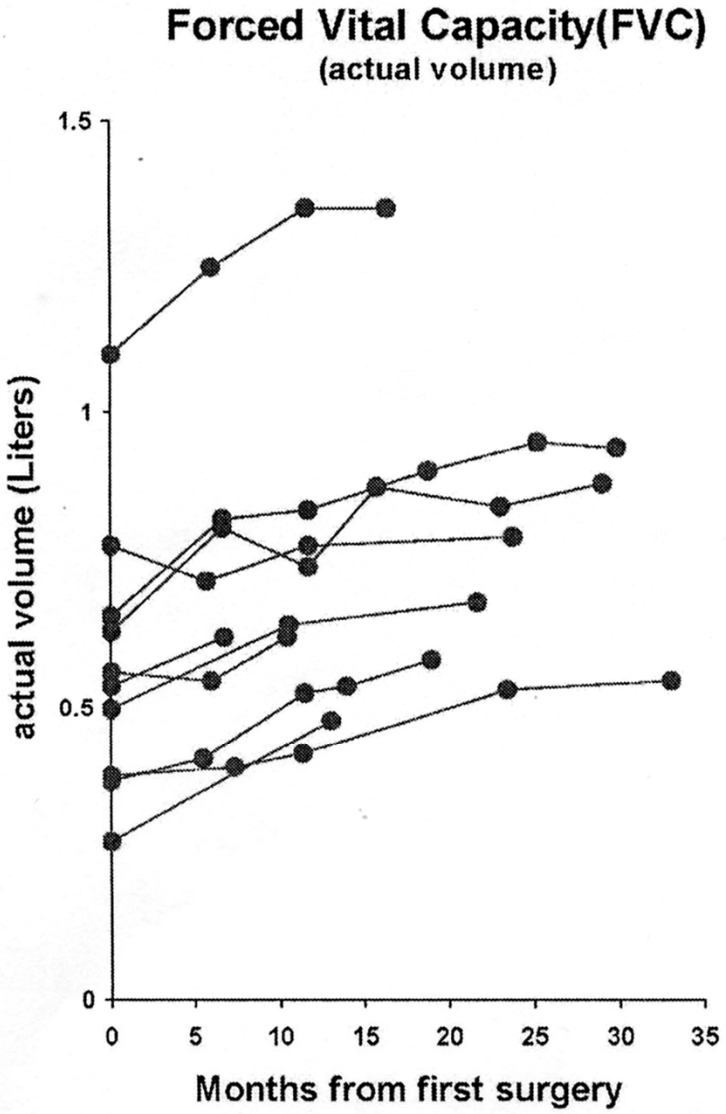
Campbell RM, et al. *J Bone Joint Surg Am* 85:409-420, 2003

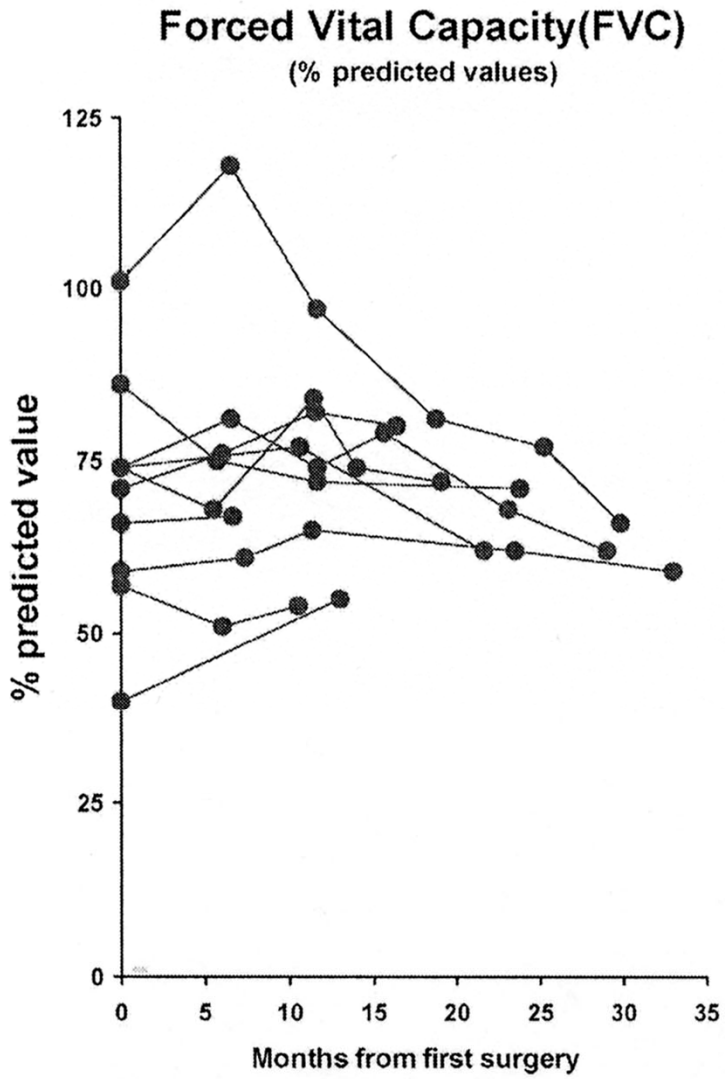
Lung Volumes Before and After VEPTR in Older Children



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

Serial Lung Functions Following Initial Expansion Thoracoplasty





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

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 ± 22%	66 ± 16%	-7 ± 2%	<.05
Crs (ml/cmH2O/kg)	1.2 ± 0.5	0.65 ± 0.3	-44 ± 22%	<.001

NR = not reported

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

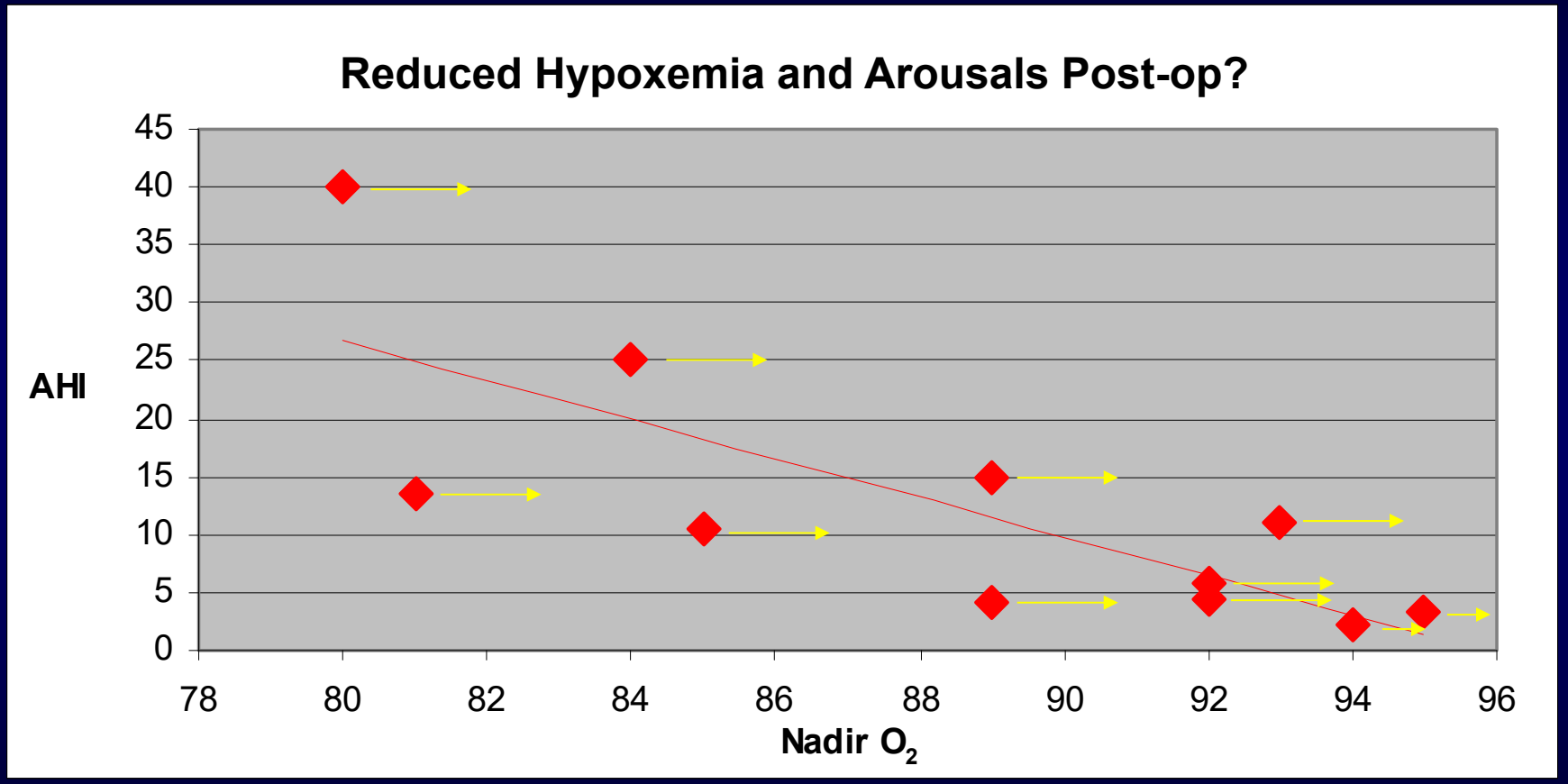
Increase in FVC After VEPTR Use: Effect of Age

Age at Surgery	*N*	Increase in FVC per year*
<6 years	16	14.7+/-8.5%
>6.5 years	7	6.5+/-5/.5%

*in absolute cm of lung volume

Motoyama et al. *Paed Resp Rev* 10:12-17, 2009.

Apnea-Hypopnea in Children with TIS During Sleep



r=0.78, $p < 0.005$

Elevated Hemoglobin Levels and TIS

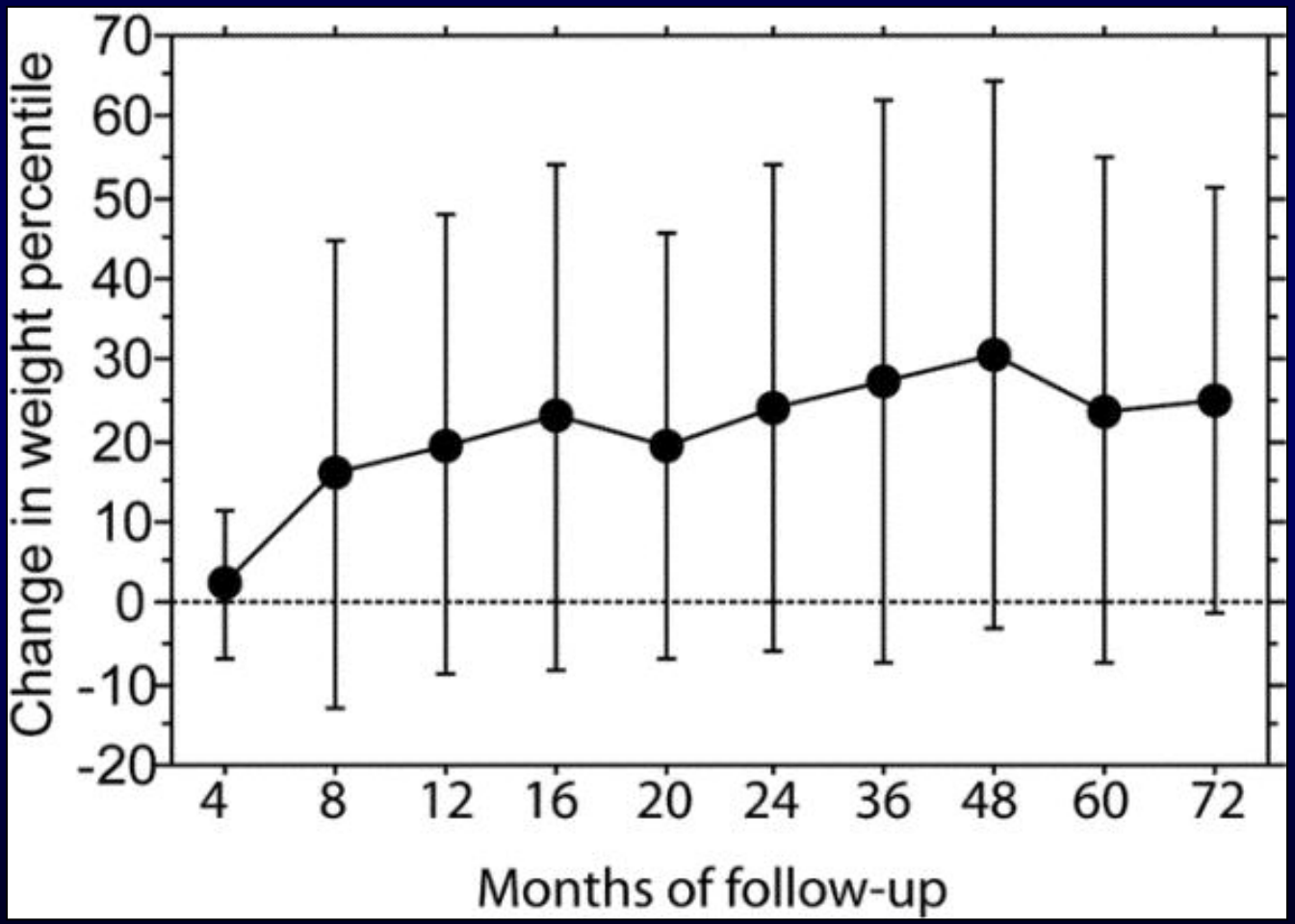
23% of patients* with EOS with Hgb levels with Z score>2

	Pre-op Z score	Post-op Z score	p value
Serum Hgb level	1.26+/-1.85	0.92+/-1.90	0.03
Serum Hematocrit	0.9_/-1.82	0.88+/-1.91	0.90

*n=138; ages 1-10 years old

Caubert J, et al. *Spine* 34(23):2534-2536, 2009.

Body Weight Before and After Spine Surgery



Skaggs DL, et al. *Spine* 34(23):2530-2533

Changes in Ventilator Needs Following Expansion Thoracoplasty

	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.*

Summary of Pulmonary Changes after VETPR Treatment

Documented Changes

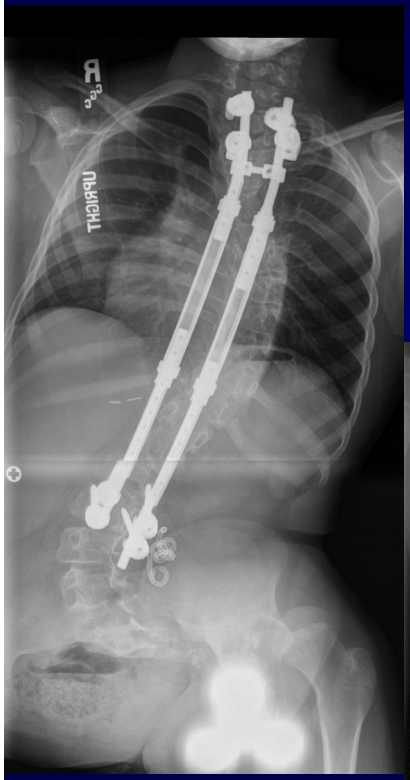
- No change in vital capacity
- Increased residual volume
- Weight gain (more than continued growth?)
- Reduced Hemoglobin levels (from elevated to more normal)
- Discontinuation of mechanical ventilation in a minority

Features to be Studied

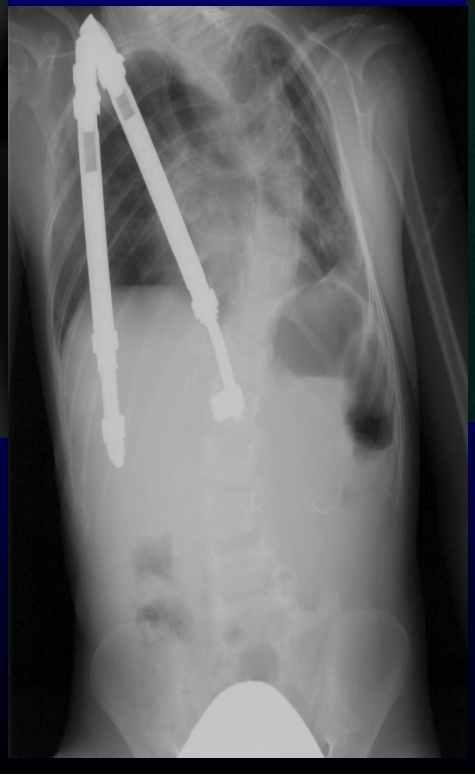
- Respiratory muscle strength and endurance
- Chest wall excursion
- Sleep quality
- Exercise tolerance and daily activity
- Pulmonary vascular recovery
- Nutritional requirements after correction

Fusionless Growing Spine Devices: Comparative Outcomes?

Growing Rods

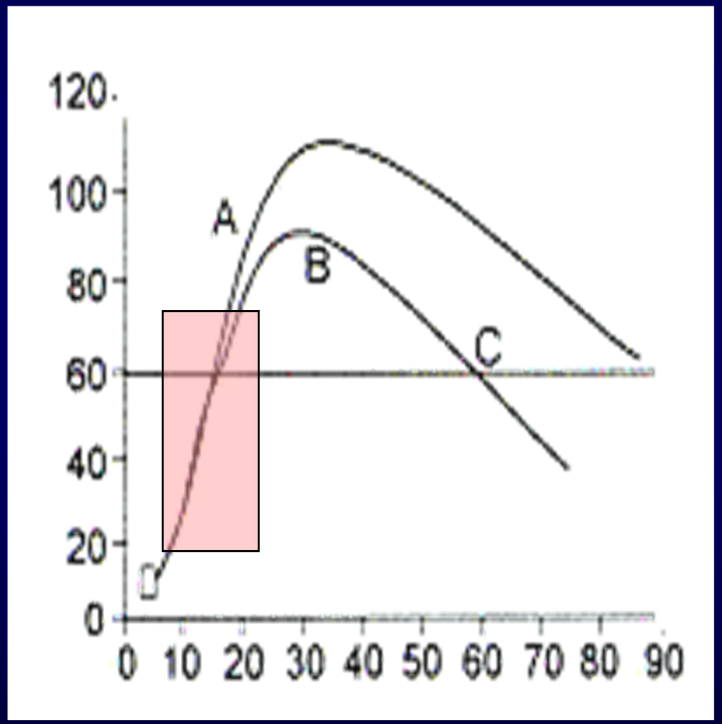


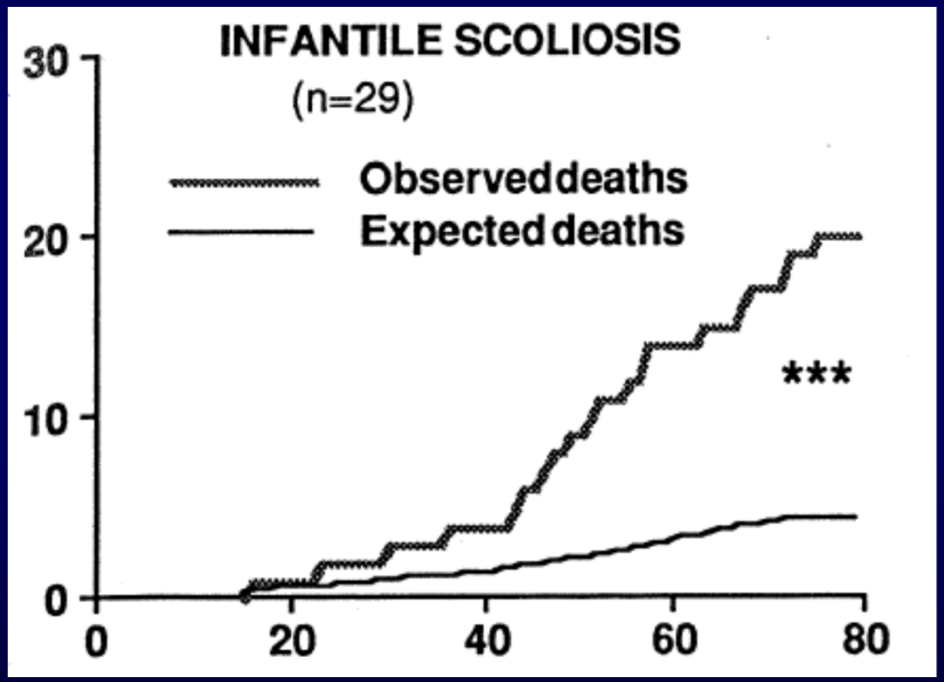
Vertical Expandable Prosthetic Titanium Ribs (VEPTRs)



Spine Staples (Vertebral Growth Modulation)

Adult Consequences of Childhood Chest Wall Disease: Long-term Prognosis of Restrictive Chest Wall Disease?





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

Summary

The respiratory goals of therapy in EOS are to increase lung/thoracic volume, chest wall mobility, and respiratory muscle movement. Of these, only lung volumes have been increased (but not vital capacity).

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

Outcome data after combined surgical and medical interventions are needed to better describe the new natural histories of children with EOS with and without cormorbid conditions.