What Every Surgeon Wants to Know About Pulmonary Issues in EOS

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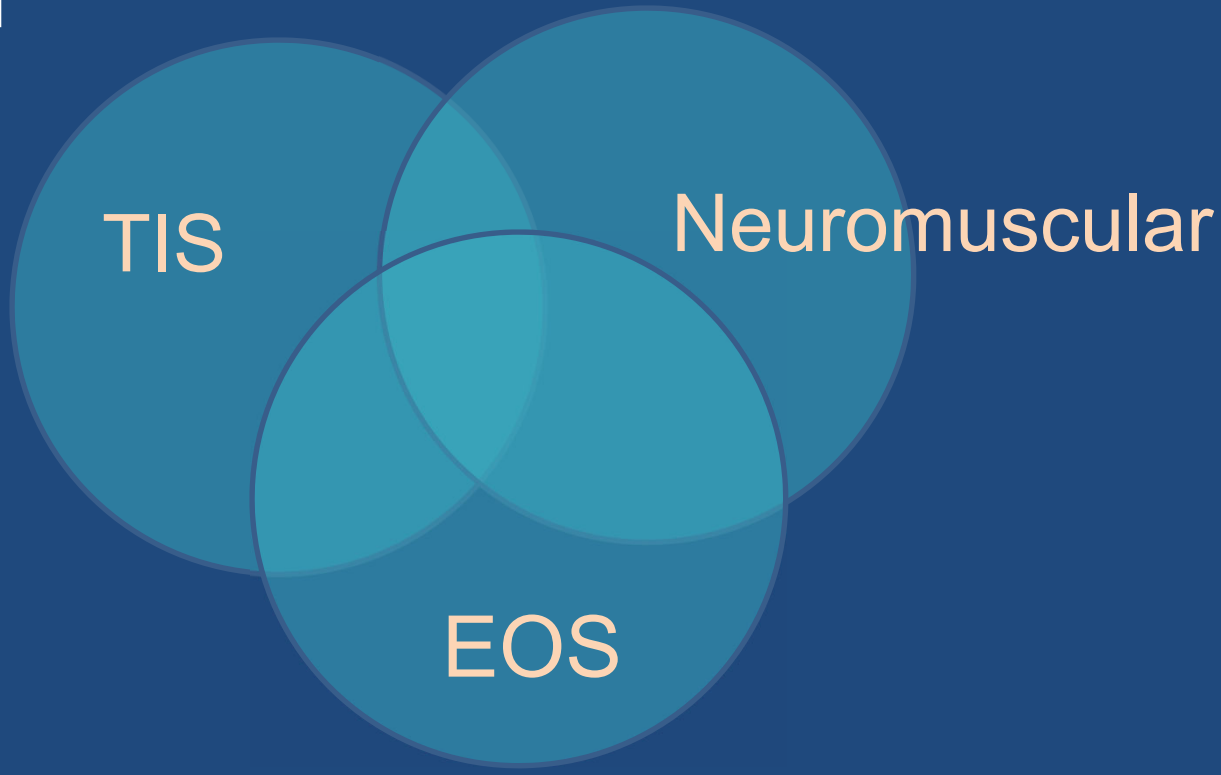
Thoracic Insufficiency and Early Onset Scoliosis

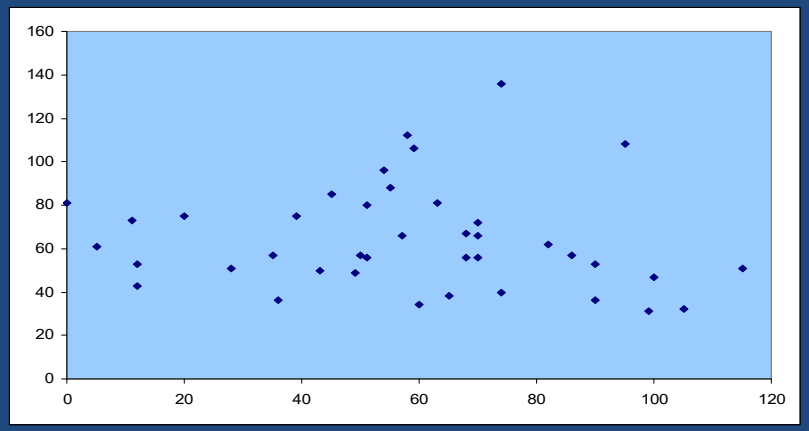
- In ability of the thorax to support normal respiratory function and postnatal lung growth



Correlations Between Lung Function Measures and Cobb Angle are Poor in EOS

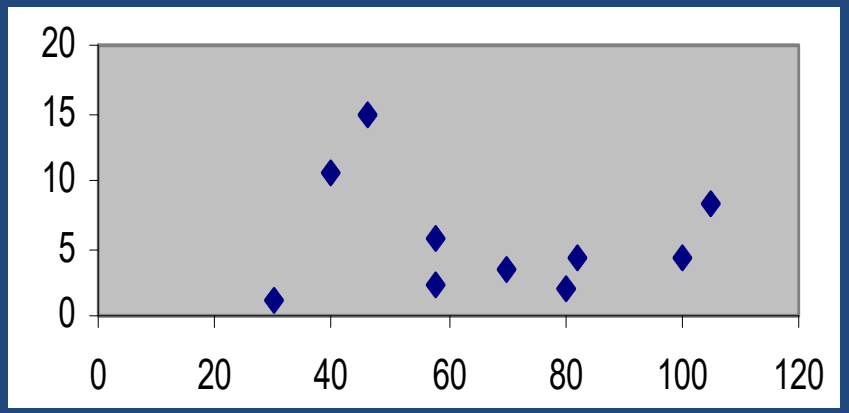
COBB ANGLE

FVC % Predicted



n=53
r=.11
p=NS

AHI



n=11
r=0.16
p=NS

Mayer OH, et al. J Pediatr Ortho 29:35-38, 2009.
Striegl A. *American Thoracic Society* (ATS), 2008.

Best Measures of Lung Function in EOS?

Direct Measures

- Spirometry
- Resp. Muscle Function
- Sleep Study
- Exercise testing
- Blood gas tensions
- Lung vent and perfusion scans
- Tidal volume and respiratory rate

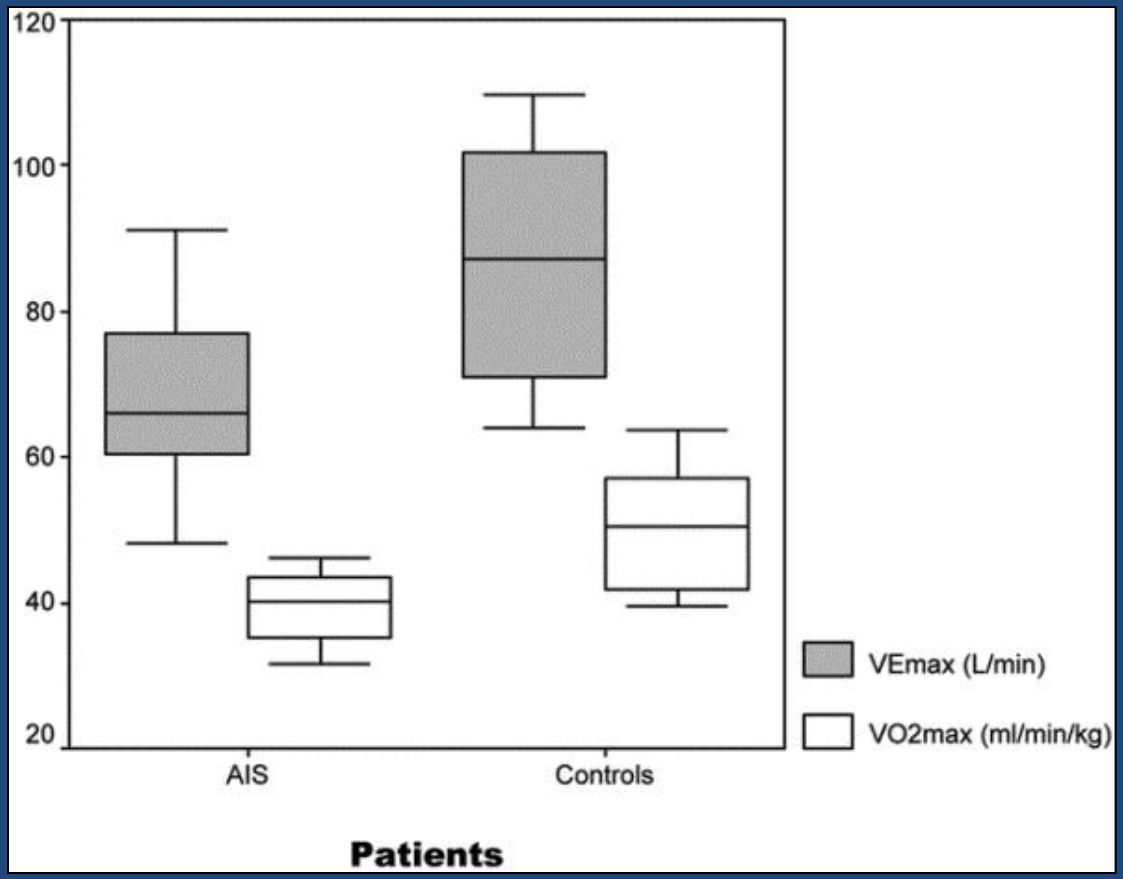
Indirect Measures

Body Mass Index
Echocardiogram

The choice depends on the question!

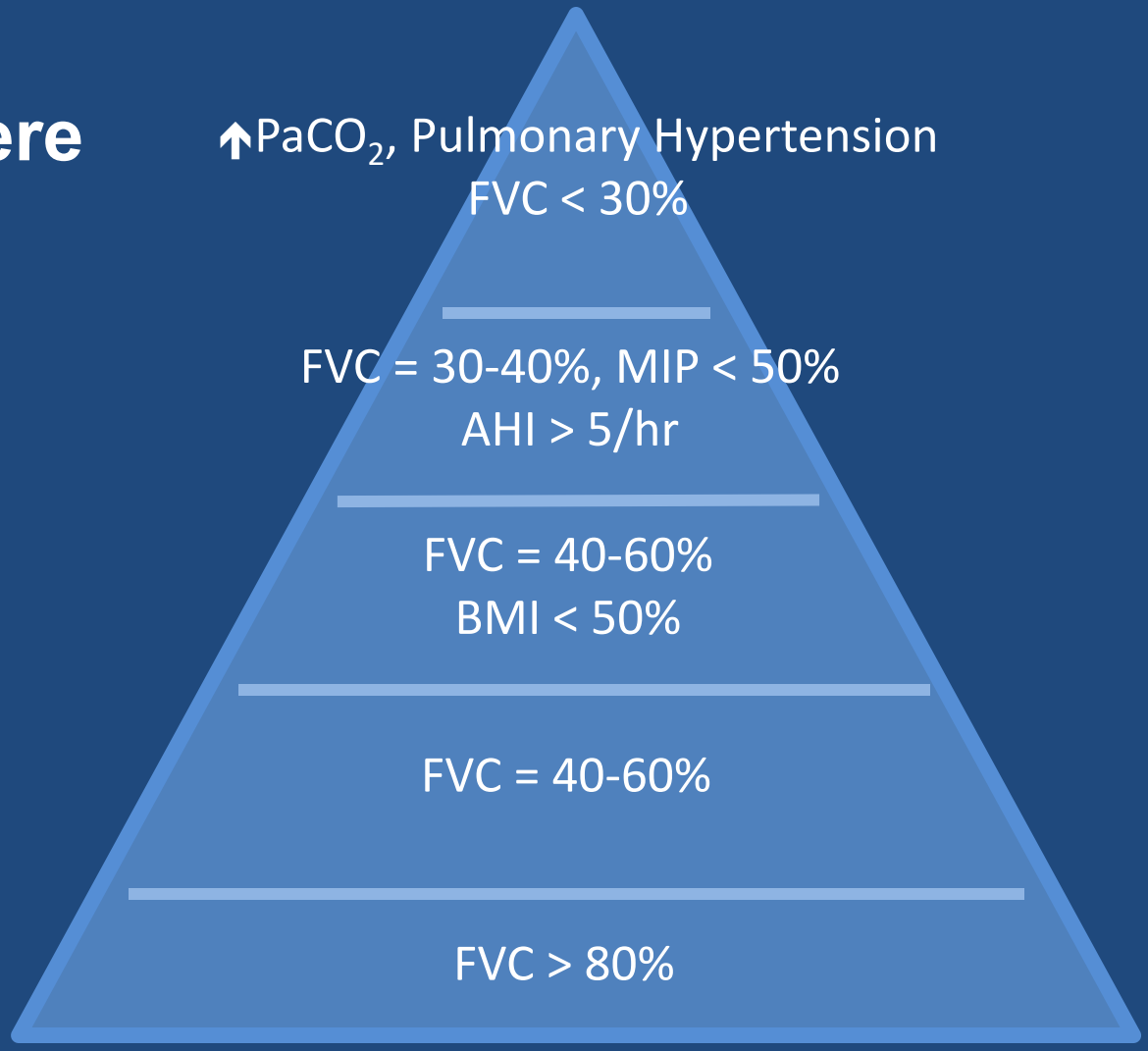
What is the most sensitive measure of respiratory changes in EOS?

VO2max (exercise)>AHI >FVC>Tidal Volume



AIS = 37
Controls = 10
Age 13 +/- 1.5 years
Cobb angle = 19-45°

What is the best measure of severe EOS?



Why measure Forced Vital Capacity?

- ↓ Low Lung Volumes
- ↓ Chest Wall Distensibility and Excursion
- ↓ Respiratory Muscle Force and Movement

Why not measure FVC?

- Age dependent, usually > 5 years old
- Variability in measurement of FVC:

	Mean week to week	2SD limits
Normal children	5%	8%
Asthma	6%	12%
Cystic Fibrosis	6%	12%
Scoliosis	?	?

Factors that contribute to variability:
Experience doing the test, age, disease, wellness

Pelkonen AS, et al. *Pediatr Pulmonol* 29:34-38, 2000.
Studnicka M, et al. *Pediatr Pulmonol* 25:238-243, 1998.
Sanders DB, et al. *Pediatr Pulmonol*, 43:1142-1146, 2008.

Active vs Passive FVC

ACTIVE

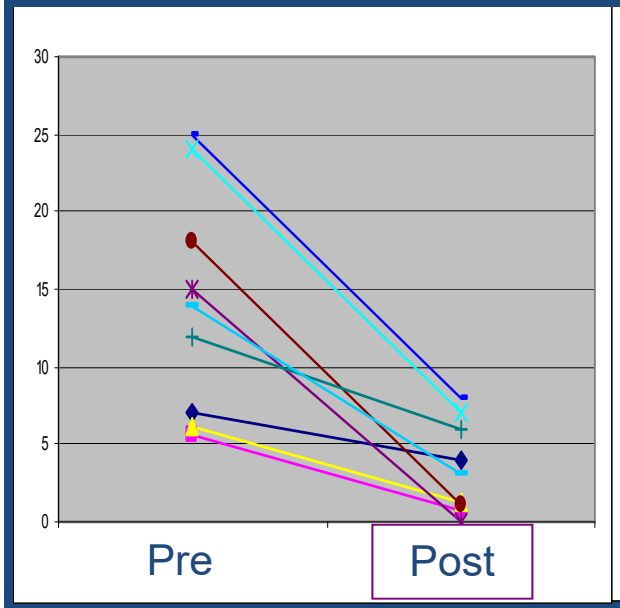
Awake, Effort Dependent

PASSIVE

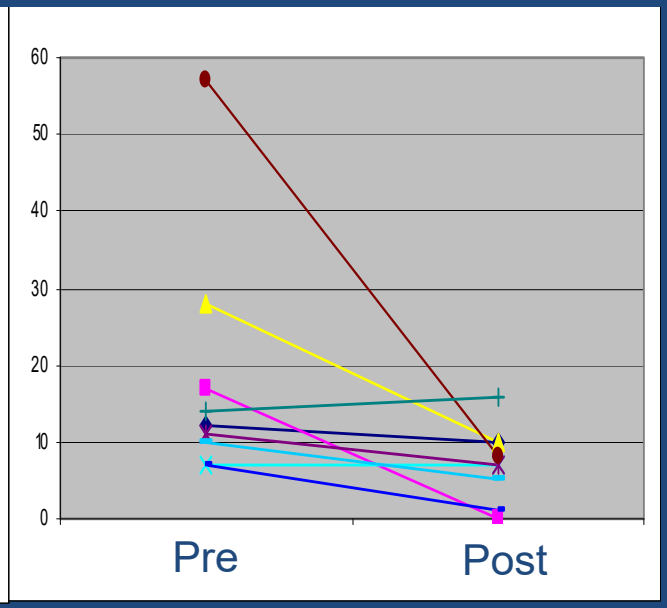
No active use of Respiratory Muscles,
Infant lung functions, OR Measurements

What respiratory measure of EOS will change supportive care?

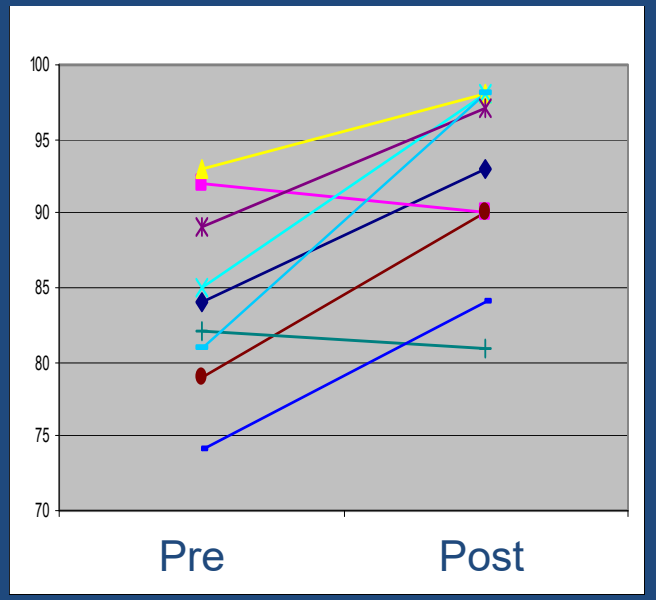
AHI*



AI*



SaO_2*



*Significant p<-05 by paired t-test

Redding G, Makris C, Song K. ICEOS, 2010.

What measure best predicts post-op pulmonary complications in EOS?

Severe Restrictive Lung Disease

N	Age (yr)	Cobb (d)	FVC (%)	LOS	% Pulm
21	12-19	82 (40-140)	18-43	18 d	10/15 (66%)
24	9-19	88 (40-129)	13-39	17 d	7/13 (54%)
32	7-17	87 (16-140)	16-39	27 d	6/32 (19%)
183	6-62	75 (45-141)	40-80	-	7/164 (4%)
			<40		6/19 (32%)

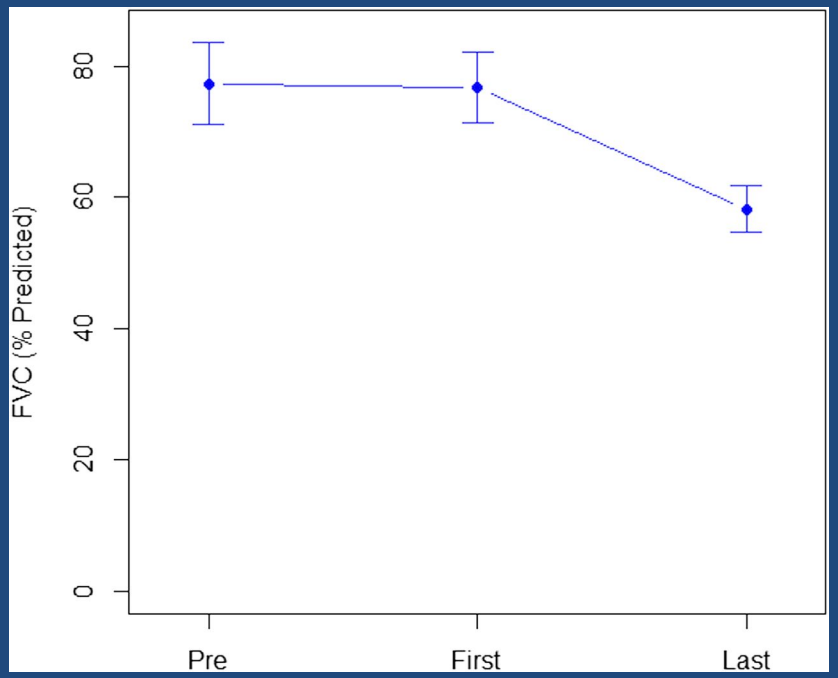
Low-risk AIS

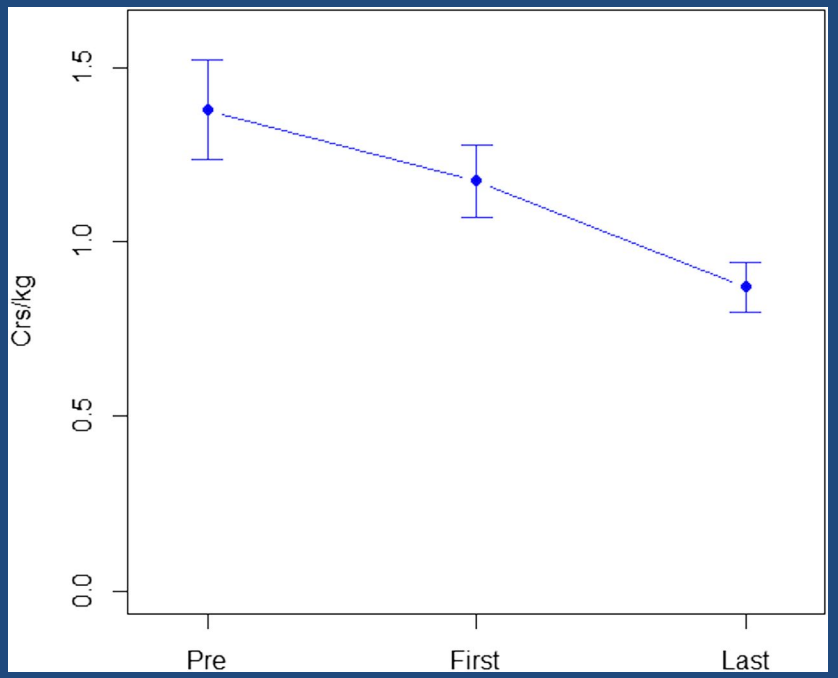
5.3- 8.4+/-3.5 days

Wazeka 2004, Payo 2009, Rawlins 1996, Gill 2006,* Zhang 2005.

What measure best quantified surgical impact?

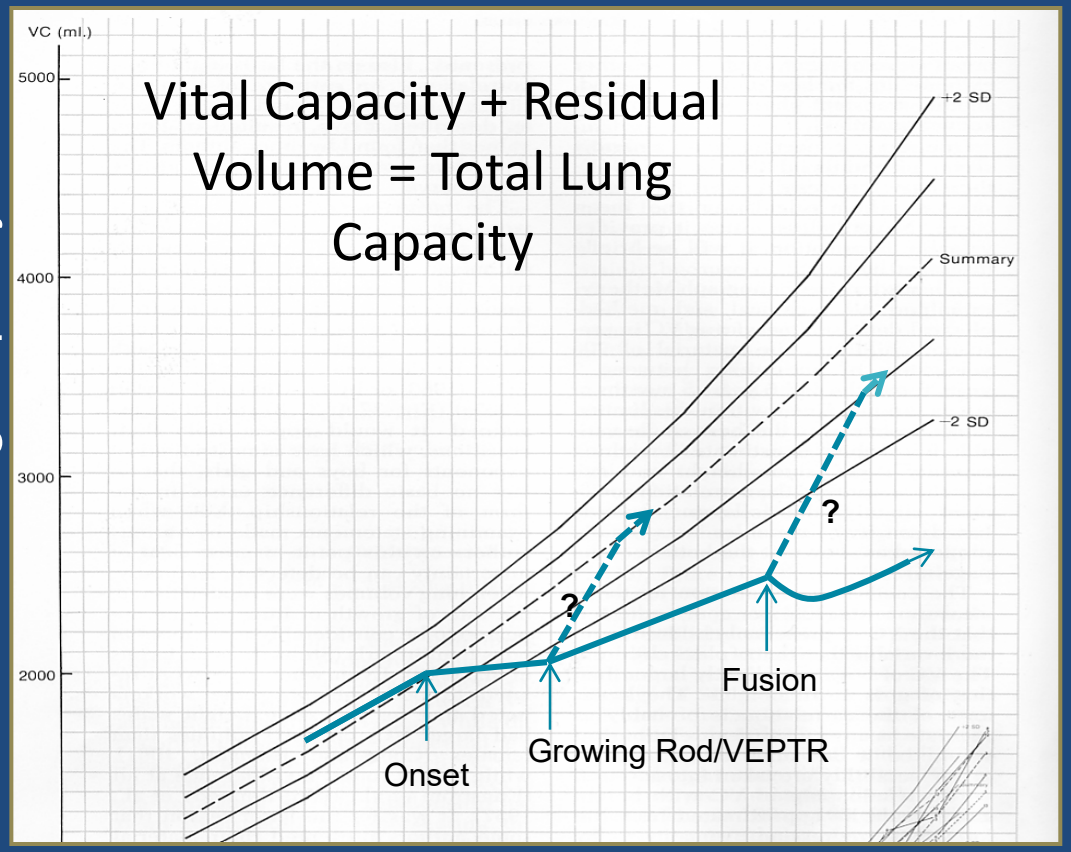
- FVC for respiratory reserve
- Chest Wall compliance for change in chest wall stiffness
- Maximum Inspiratory Muscle Strength
- Others? - Sleep quality - VO2max for exercise tolerance



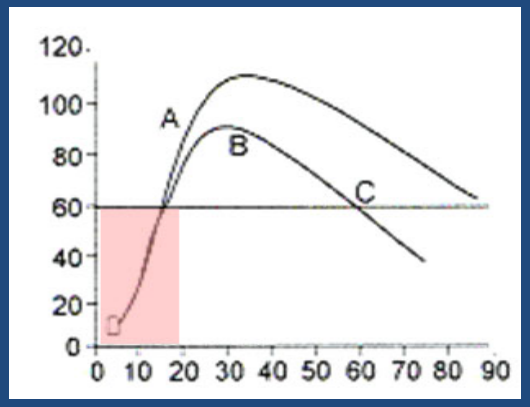


Dede O et al. *J Bone Joint Surg Am* 96:1295-1302, 2014.

How do you Determine Long-term Pulmonary Outcomes?



Total Lung Capacity



FVC % of normal at age 20

Age (years)

How Do We Improve Pulmonary Outcomes?

- Maximal three-dimensional correction including rotation
- Early onset intervention? Non-invasive approaches?
- Re-orientation of respiratory muscles? Sub-diaphragmatic release?
- Less force with first correction?
- Changes in distraction expansions non-invasively?
- Perhaps with late interventions, pulmonary hypoplasia precludes improvement?

The Role of the Pediatric Pulmonologist in the Management of EOS

- Find one that has an interest in this population.
- Find one that can interact directly with you and discuss the implications of test results.
- Find one who deals with uncertainty well.
- Find one who wants to improve current pulmonary outcomes in these children.
- Find one who is in this business for the long haul.

Summary

- Progressive EOS produces progressive pulmonary limitations and loss of reserve.
- Lung functions are useful to monitor changes over time and with treatment.
- Given the variation from patient to patient with EOS, lung function tests will help dictate care in some patients but not others.
- The lack of improvement in lung function with current surgical techniques calls for further treatment innovations for this group of children.