Assessment of Pulmonary Problems in EOS

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Disclosures

- Pediatric Pulmonary Section Editor for UpToDate
- NIH Data Safety Monitoring Board for AsthmaNet research collaborative

Lung Functions: Which Pertain to EOS?

- Lung mechanics and work of breathing
- Gas exchange ($PaCO_2$, PAO_2, SpO_2)
- Respiratory muscle function
- Pulmonary vascular function (cor pulmonale)
- Host defenses, e.g. cough effectiveness
- Distribution of lung function (lung scans)

Direct and Indirect Pulmonary Changes Due to EOS

Direct

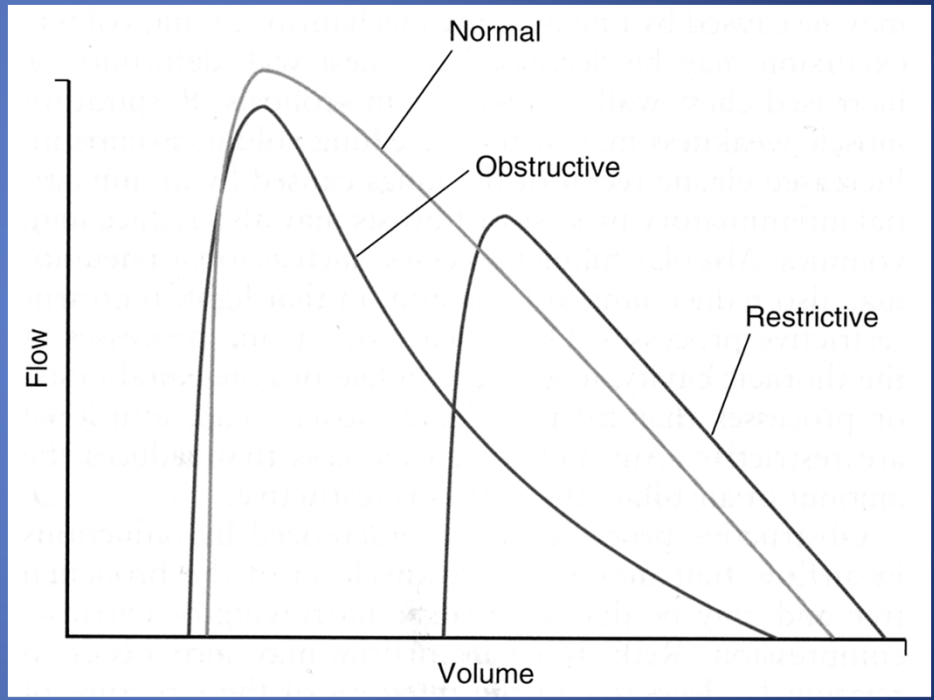
- Reduced thoracic cage and lung volumes
- Reduced thoracic excursion
- Reduced diaphragm excursion

Indirect

- Poor weight gain and reduced Body Mass Index
- Poor Sleep quality
- Poor Quality of Life
- Increased frequency of pulmonary infections
- Increased post-operative complications

Forced Vital Capacity

- FVC requires effort to breathe in maximally and out forcefully and completely. 5-6 year olds can do it.
- FVC is reduced due to all 3 RESTRIVE changes produced by EOS:
  - Small lung and thoracic volumes
  - Decreased chest wall mobility and flexibility
  - Decreased inspiratory and expiratory muscle strength and movement
  - Does not distinguish which mechanism is most important for a given child.



Castile RG, Davis SD. In: Disorders of the Respiratory Tract in Children (8th ed.) 2012.

How “severe” is the Pulmonary Impact of EOS?
Or
What does the FVC value signify?

- Vital Capacity <85% using arm span for height is abnormal. 2,4 and 6 standard deviations from the mean define mild, moderate, and severe disease.
- VC<40% predicts nighttime hypercapnia in children with neuromuscular disease. In EOS?
- VC<20% predicts risk of ventilatory support 24 hours/day in neuromuscular disease. In EOS?

Obstructive Lung Disease in EOS

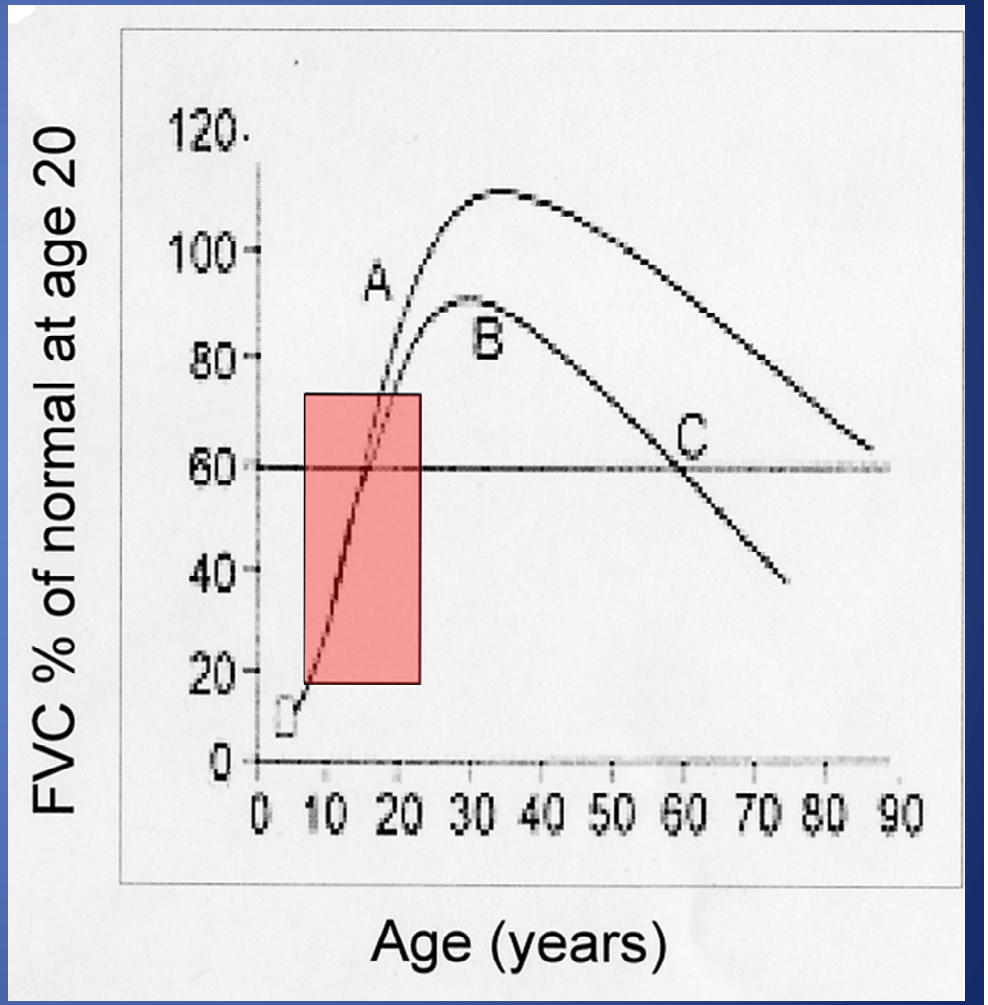
- Determined by FEV_1/FVC <80%
- Occurs in up to 1/3 of children with congenital scoliosis.*
- Due to mainstem bronchial compression by vertebral intrusion.
- May be a population of children with EOS that improves substantially after surgical intervention.
- Need to rule out asthma with a bronchodilator challenge.

Compression of Right Mainstem Bronchus with Spinal Lordosis

*McPhail G, Sturm P, Wall E, et al. JPO, 2012.

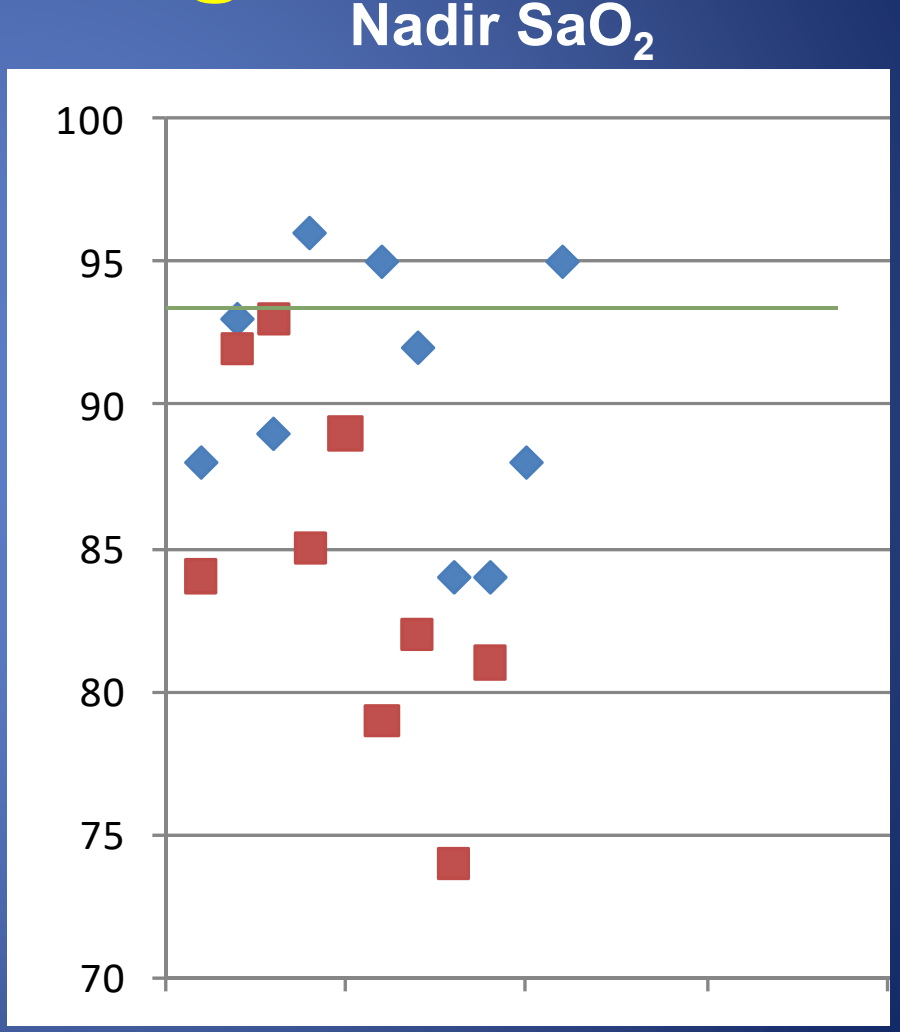
FVC and Growth

- A FVC value of 50% at 8 years does not portend the same FVC at 20 years.
- The respiratory goal of surgical treatment is normal lung function by the end of lung function development (17-23 years)
- VEPTR treatment provides almost enough thoracic growth to maintain VC as a % of predicted over 3 years of growth. Longer studies and studies with new devices are needed.

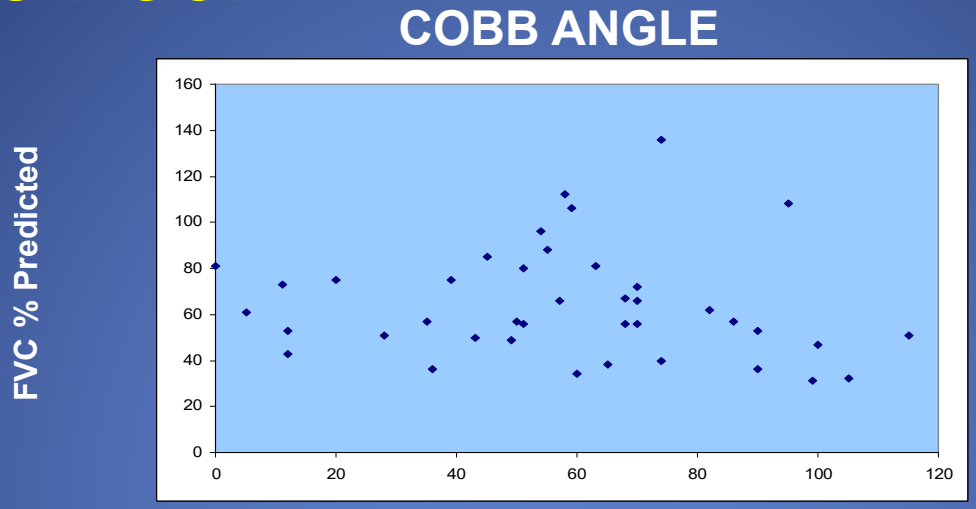


Pulmonary Functions for Screening

- Polysomnography (Overnight Sleep Study): Apnea Hypopnea Index (AHI), Nadir SaO2, and Maximum CO_2
- Echocardiogram or ECG: Screening for cor pulmonale and pulmonary hypertension.
- Serum CO_2 content on electrolytes and pre-operative PCO2
- Maximum inspiratory and expiratory pressures (MIP/MEP)



Structure-Function Correlations in EOS are Poor



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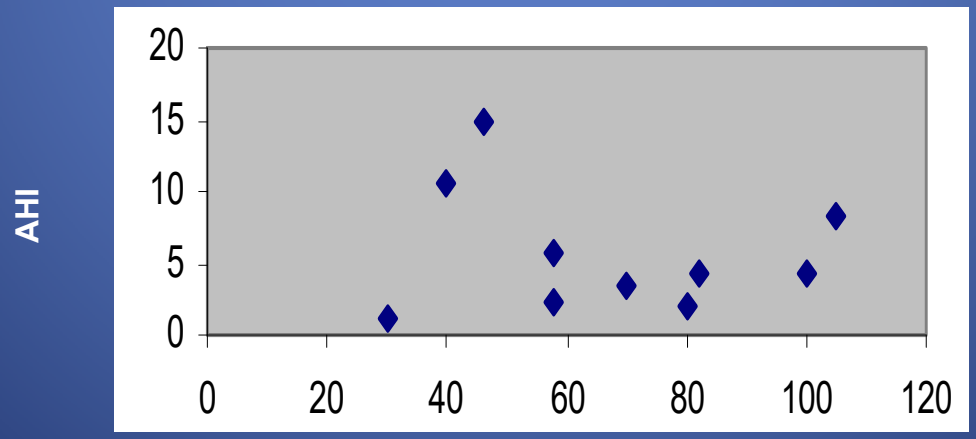

n=11
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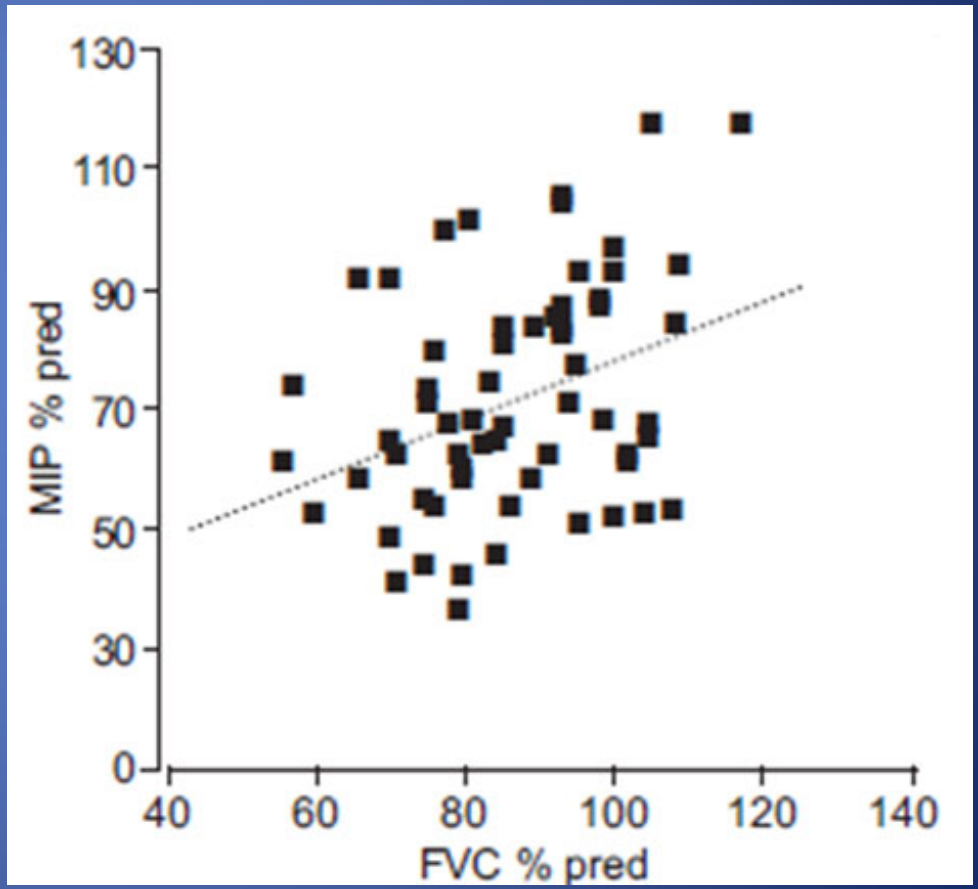
p=NS

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

Respiratory Function Correlations and Primary Mechanisms of Disease

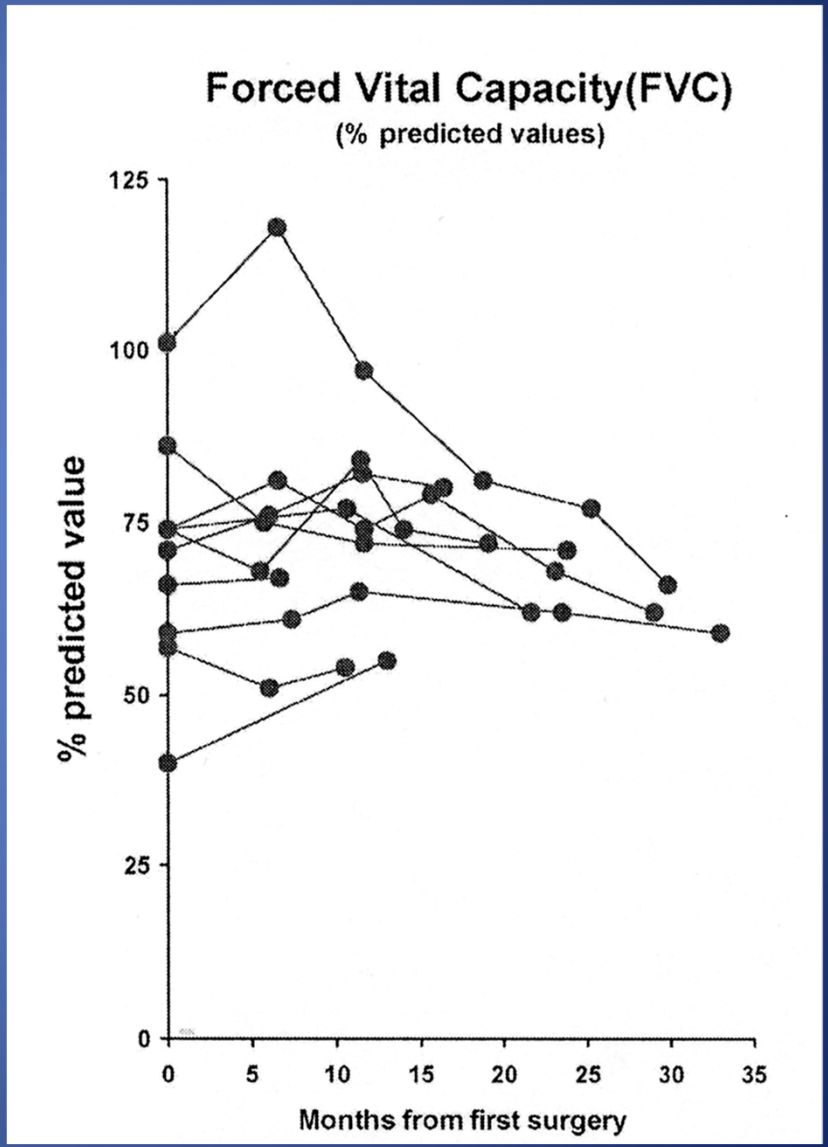
- Respiratory muscle strength and vital capacity
- Sleep–related hypoxemia and functional residual capacity.
- Polycythemia and night-time hypoxemia
- Hypoxemia and pulmonary hypertension
- Cough-generated peak flow and prolonged intubation post-operatively



Martinez-Llorens J, et al. *Eur Respir J* 36:393-400, 2010.

Pulmonary Function Uses

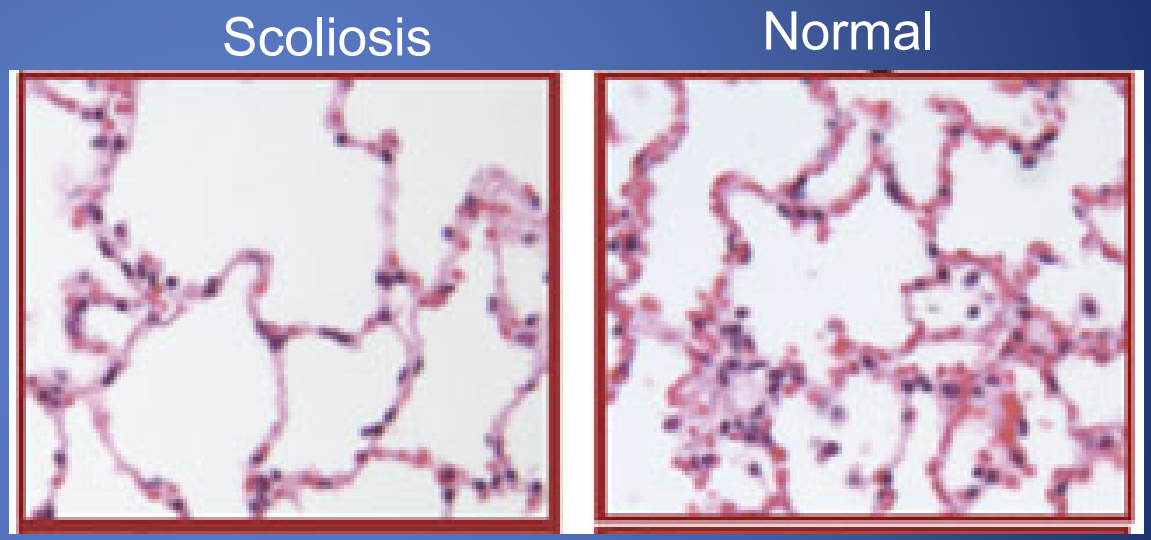
- Determine impact of disease over time
- Determine effects of interventions over time
- Determine short-term and long-term risk of intervening, e.g. casting, bracing, surgical instrumentation, fusion
- Risk of sudden death at home?
- Determine the mechanism of disease that needs attention, i.e. more strategic intervention



Motoyama EK, et al. *Spine* 31:284-290, 2006.

Future Directions in Pulmonary Assessment of EOS

- Measurement of Postnatal Pulmonary Hypoplasia: Lung Diffusion Capacity
- Methods to measure lung mechanics in 3-4 year olds
- Serial measurements of structure and function in EOS
- Effects of spine fusion after growing rod instrumentation
- Lung function effects of continuous spine distraction



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