



When Is It Time To Do Surgery from a Pulmonary Perspective

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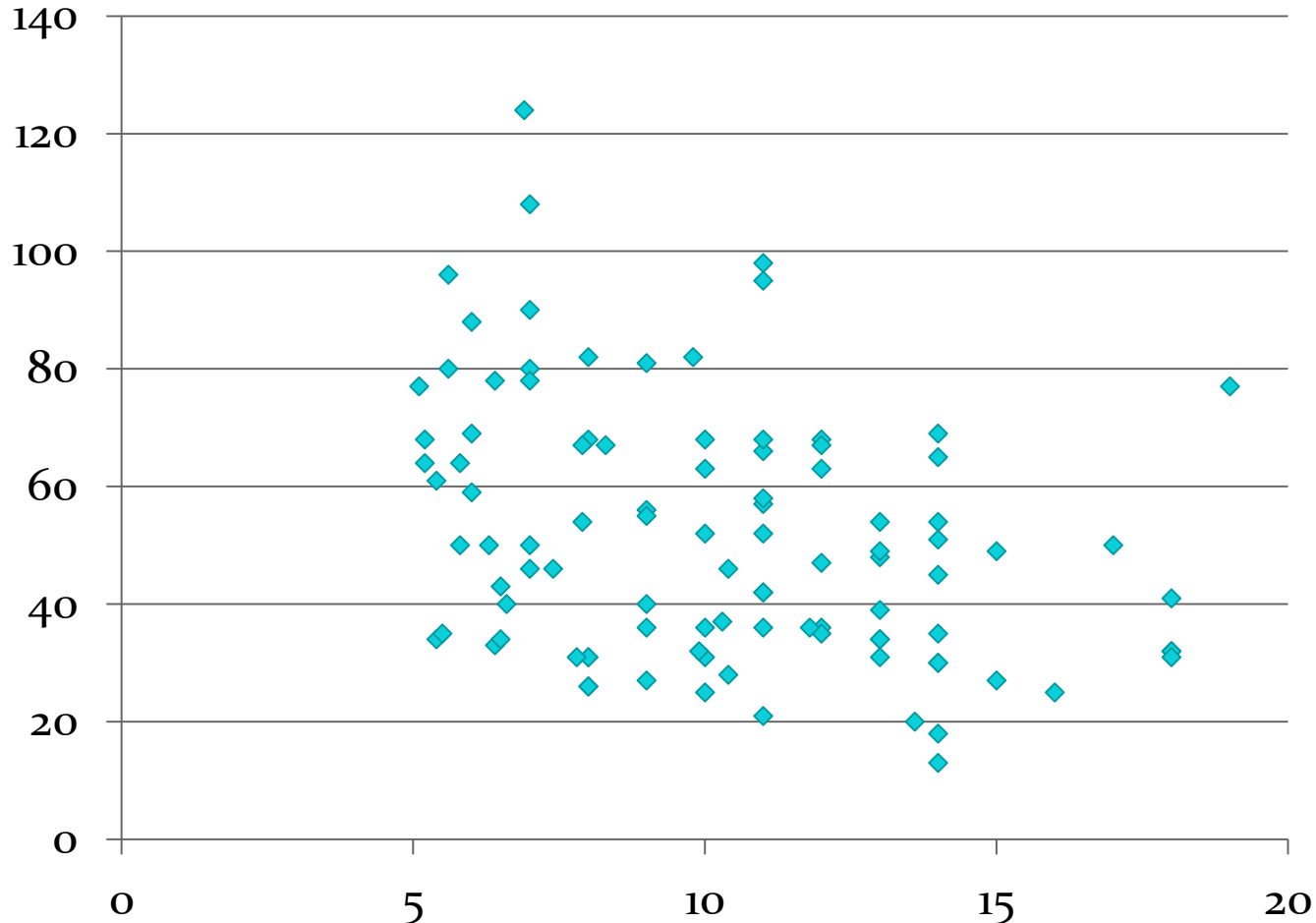
Disclosures

- Pediatric Pulmonary Section Editor of UpToDate

What do we (think we) Know?

- The 3-dimensional spine/thoracic deformity leads to lung impairment that worsens as the deformity worsens.
- Pre-operative lung function is a major determinant of post-operative lung function (long and short term).
- Lung function after insertion of growth friendly distraction devices does not improve in most children but allows further lung function growth (in liters).
- Co-morbid states, such as neuromuscular weakness often determine the pulmonary outcome of children despite the scoliosis repair.

Seattle-Philadelphia-San Antonio: First FVC (% predicted) *



CSSG Registry: 54% of 3,968 patients > 5 years old at presentation

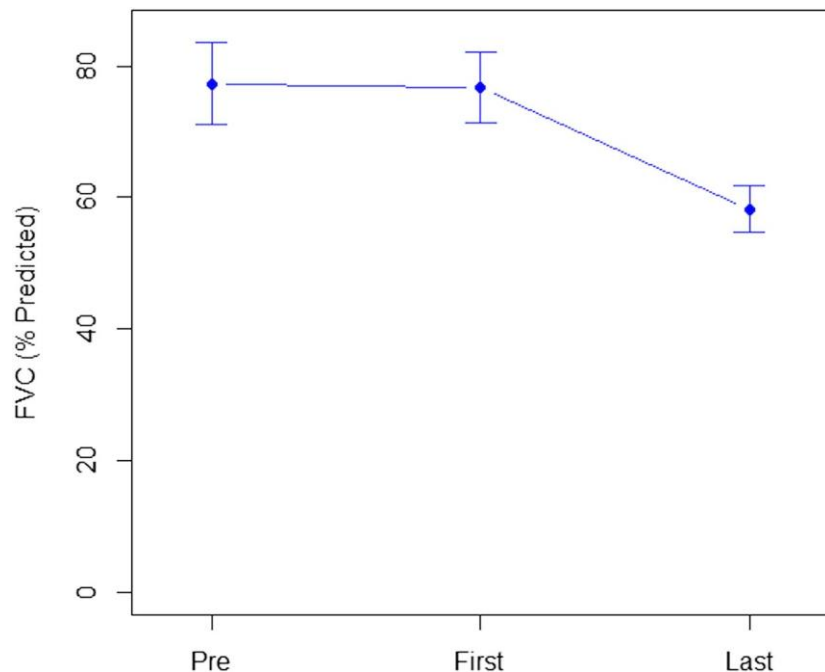
Classification for Early Onset Scoliosis + Pulmonary Data

Age	Etiology	Major Curve Angle	Kyphosis	APR Modifier	FVC	PFT Modifier
Continuous Prefix	<u>C</u> ongenital/ Structural	<u>1</u> : <20°	(-): <20°	<u>P</u> ⁰ : <10°/yr	>70%	<5%/yr
	neuro <u>M</u> uscular	<u>2</u> : 20-50°	<u>N</u> : 20-50°	<u>P</u> ¹ : 10-20°/yr	50-70%	5-10%/yr
	<u>S</u> yndromic	<u>3</u> : 51-90°			30-50%	
	<u>I</u> diopathic	<u>4</u> : >90°	(+): >50°	<u>P</u> ² : >20°/yr	<3%	>10%/yr

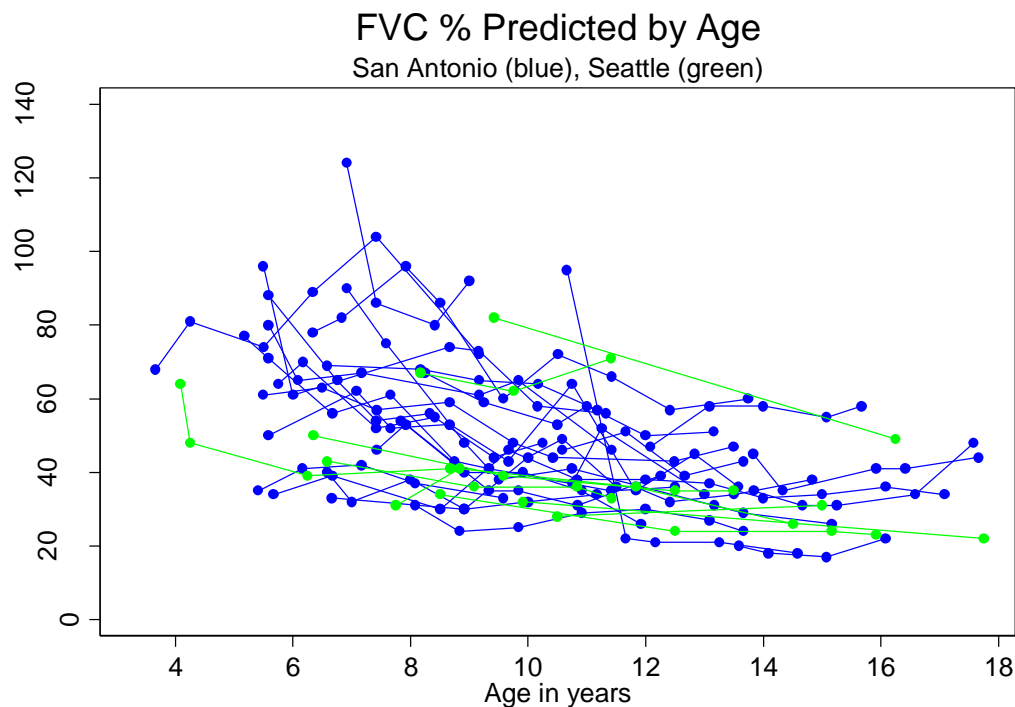
Spine/Pulmonary Joint Decision Points

- Is there a threshold lung function value where I have to intervene immediately?
- What degree of lung function loss can I tolerate as a spine deformity progresses?
- How often do I follow lung function to assess trends?
- How do I juxtapose this decision with age of surgical preferences, e.g. fusion?

Serial FVC Over a 6 Year Interval



28% Reduction from Pre to LAST

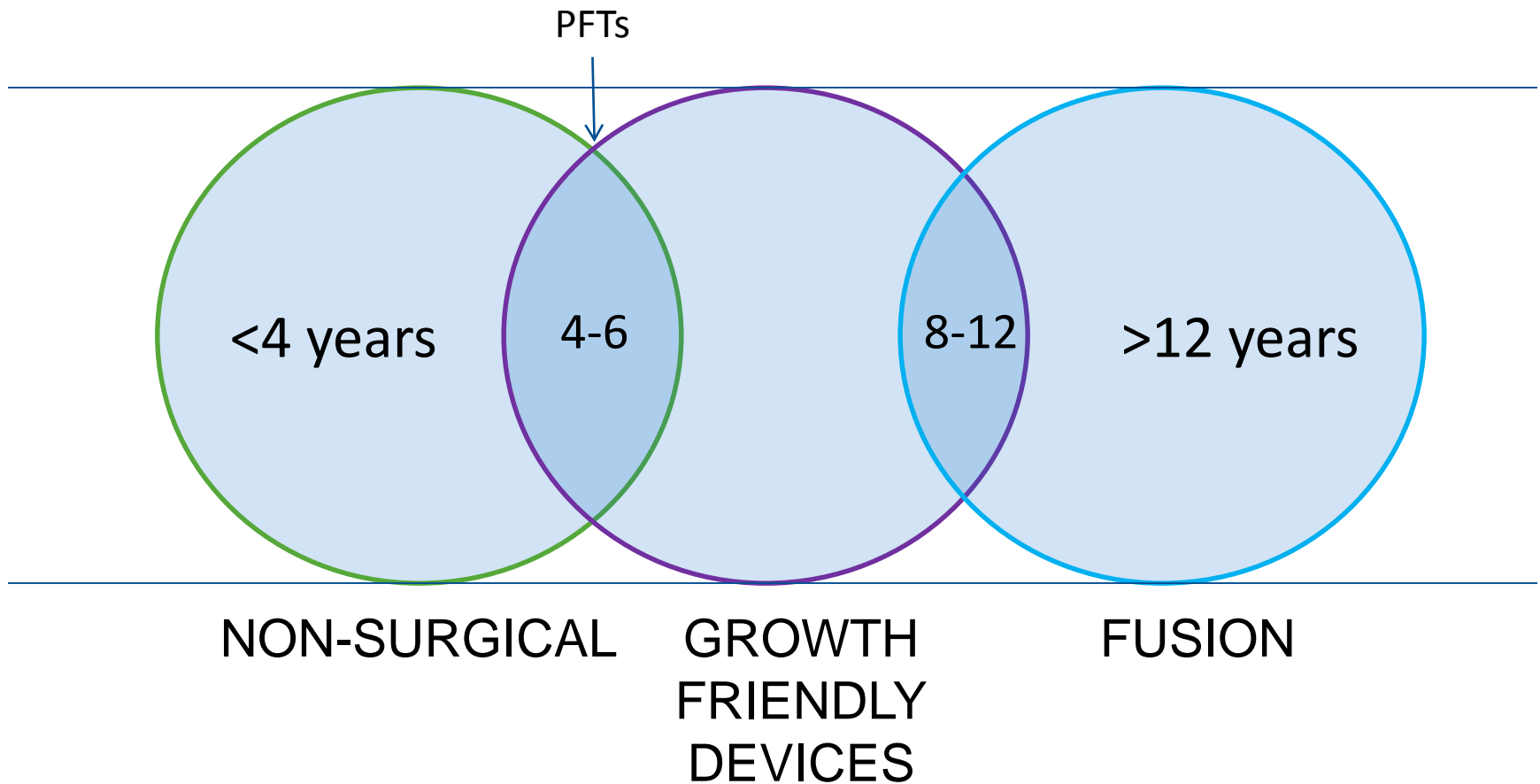


16% Reduction Over 6 Years

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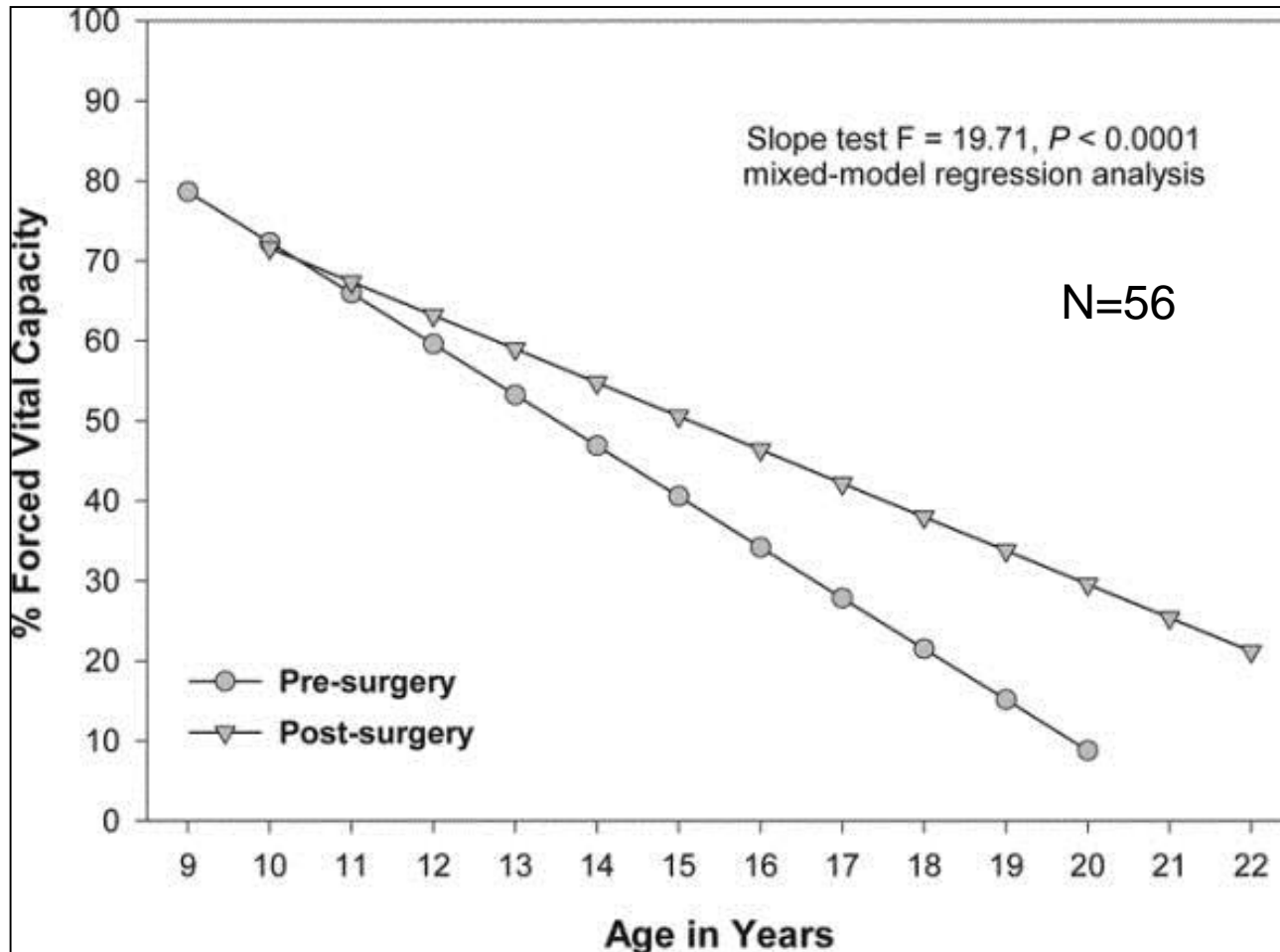
Ages and Treatment Options for Children with EOS



Evidence for Earlier Intervention

- VEPTR insertion and expansion (6 years F/U)
 - N=16 <6 y/o; N= 7 >6 y/o
 - FVC change per year: <6 y/o: 14.7+/-8%
>6 y/o: 6.5+/-5.5%
- Vertebral Column Resection (2 year F/U)
 - N=27 children, ages 8-18 years
 - Correlation between age of surgery and change in FVC (not % predicted) $r=-0.44$; $p=.02$

Decline in FVC after surgery spine fusion for DMD based on age



Speculation

- Earlier *intervention* to prevent deformity progression is better for lung function preservation than later. Intervention does not mean surgery or type of surgery.
- *Ideal timing* of surgical interventions may not coincide with ideal timing to stop lung function decline. How do we decide next steps?
- New methods are needed to salvage lung function which is severely impaired at presentation.