

# Pulmonary Function in EOS: Where Have We Succeeded

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# Disclosures

- ▶ Section Editor, Pediatric Pulmonary Medicine, UpToDate

# Defining “Success” and Lung Function

- ▶ Recovering impaired lung function
- ▶ Preventing progressive decline in lung function
- ▶ Keeping up with somatic growth (FVC in liters or in % predicted based on arm span or ulnar length).
  
- ▶ Lung Function: Gas Exchange, Lung Mechanics (FVC, FEV1, MVV, Compliance measures), Respiratory Muscle function, Pulmonary Hypertension, Pulmonary host defenses (e.g.cough), Exercise tolerance, Sleep quality.

# Recovery of Lung Function in EOS

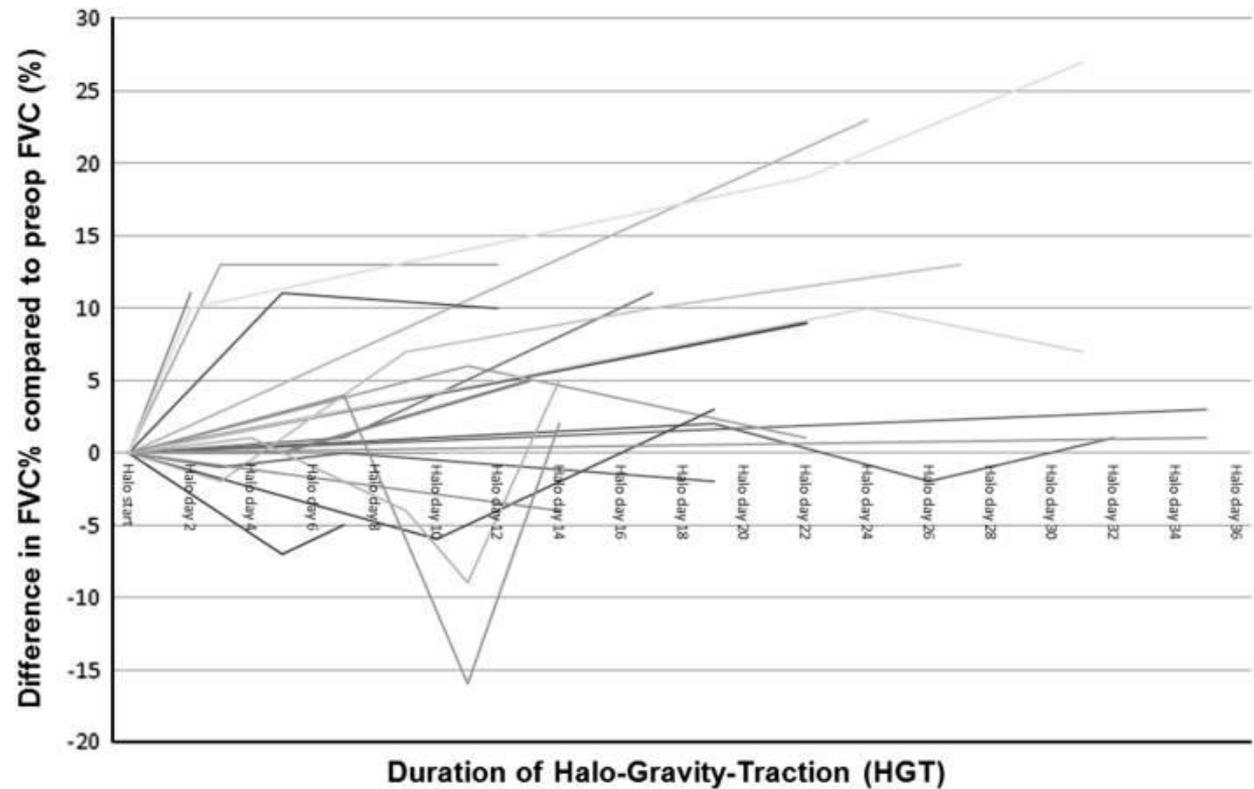
- ▶ Short term FVC changes after VEPTR use: 92 patients(3 studies)of EOS/TIS 7–30 months after surgery.
  - No changes in mean values (pre-vs post-op) in each study
  - 15 (28%) of 53 patients had improved FVC (how much?)
- ▶ Short term Lung volume by CT imaging 2–3 years after VEPTR use.
  - Mean % increase in lung volume 121% (range 24–326%)
  - 11 /17 (65%) increased lung volume by >50% of baseline

# Improvement in Children with EOS on home respiratory support

- ▶ 77 patients from CSSG registry on day/night respiratory support.
- ▶ Within 6 years following VEPTR surgery:
  - 24% required less support
  - 12% required more support
  - 64% required the same level of support as before surgery.
- ▶ Diagnoses most likely to improve were:
  - Congenital scoliosis
  - Neuromuscular weakness conditions

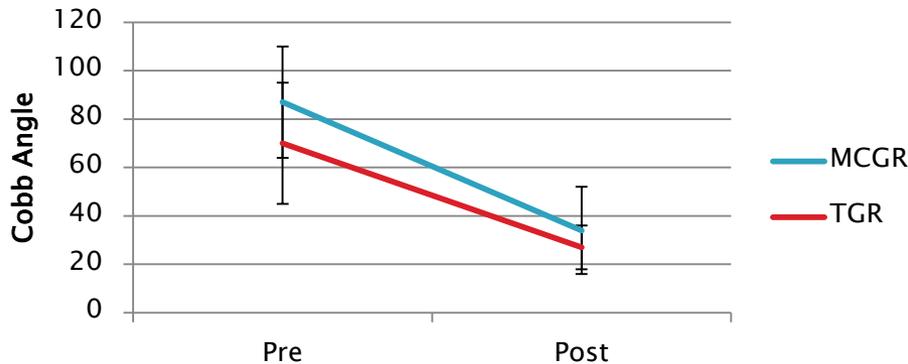
# Improvement in FVC (% predicted) with Halo Traction

9 of 24 patients with increased FVC% by >10% with 2–8 weeks of Halo treatment.

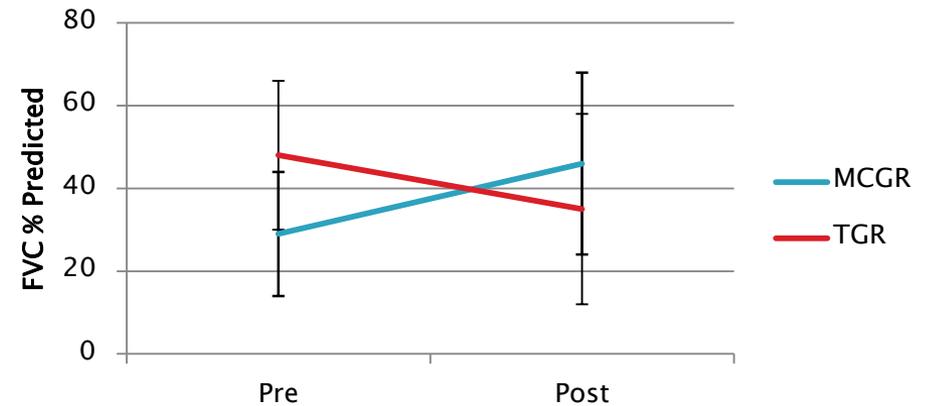


# Improvement in FVC among Children with SMA II and EOS

## Pre- and Post-Operative Cobb Angles

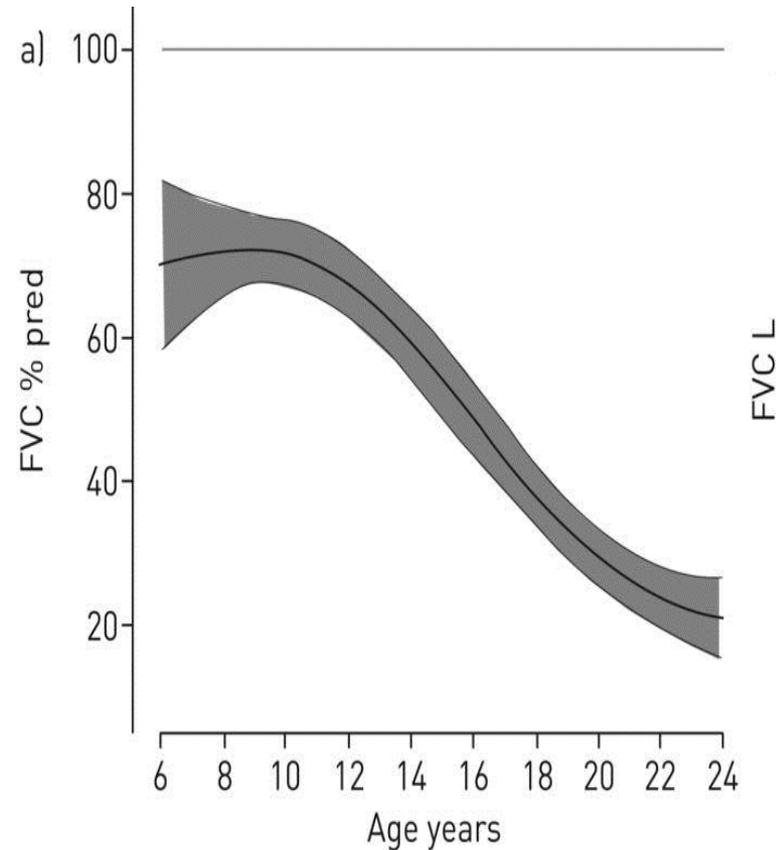
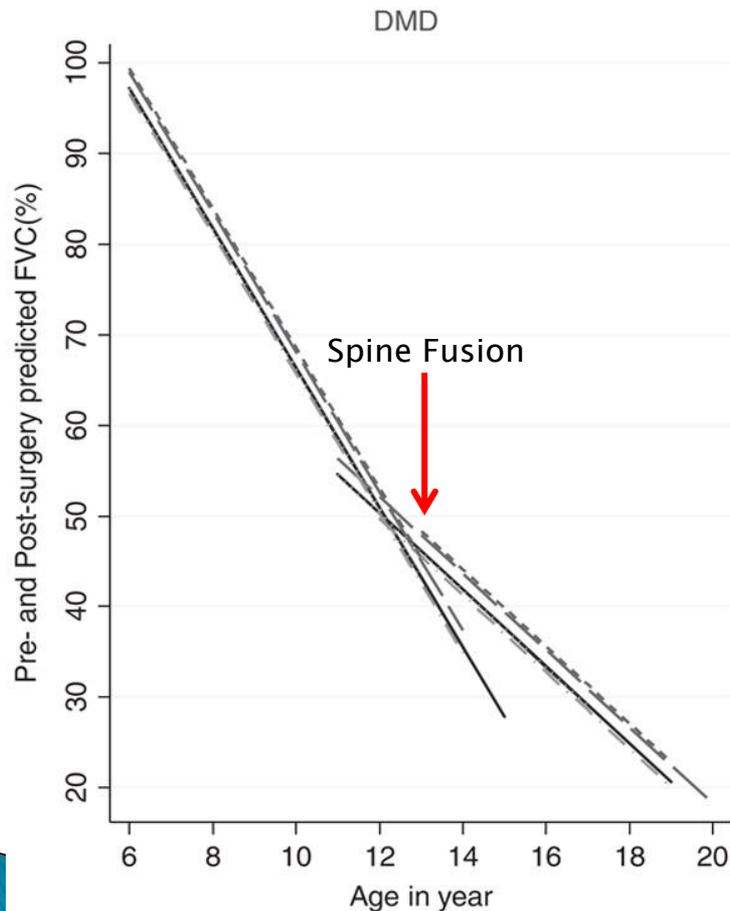


## Pre- and Post-Operative FVC



N=12

# Effects of Spine fusion on Boys with Duchenne's Muscular Dystrophy



# Current Gaps

- ▶ Few long term studies.
  - ▶ Using absolute values, e.g. Liters instead of % predicted does not separate growth from therapeutic effects. Can't use height for % predicted values.
  - ▶ No serial PFT studies on most surgical devices or strategies (Traditional rods, MCG rods, Shilla, etc.)
  - ▶ Heterogeneous patient population.
  - ▶ Inability to assess lung function <5 years of age.
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# Summary

- ▶ Studies that describe mean group differences in pulmonary outcomes are unlikely to find large improvements.
  - ▶ Proportions of children whose lung functions improve with spine treatments need to be reported as do features of those who dramatically improve (or worsen).
  - ▶ Prevention of progression over years should be considered a success for many patients with EOS.
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