Pulmonary Function in EOS: Where Have We Succeeded

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Disclosures

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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

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.
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.