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

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?

Severe

- \( \uparrow \) PaCO\(_2\), Pulmonary Hypertension
- FVC < 30%

Mild

- FVC = 30-40%, MIP < 50%
- AHI > 5/hr

- FVC = 40-60%
- BMI < 50%

- FVC = 40-60%
- FVC > 80%
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:

<table>
<thead>
<tr>
<th>Condition</th>
<th>Mean week to week</th>
<th>2SD limits</th>
</tr>
</thead>
<tbody>
<tr>
<td>Normal children</td>
<td>5%</td>
<td>8%</td>
</tr>
<tr>
<td>Asthma</td>
<td>6%</td>
<td>12%</td>
</tr>
<tr>
<td>Cystic Fibrosis</td>
<td>6%</td>
<td>12%</td>
</tr>
<tr>
<td>Scoliosis</td>
<td>?</td>
<td>?</td>
</tr>
</tbody>
</table>

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

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*

Al*

SaO₂*

*Significant p<-05 by paired t-test

### Severe Restrictive Lung Disease

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

### Low-risk AIS

5.3-8.4+/-3.5 days

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
How do you Determine Long-term Pulmonary Outcomes?

Vital Capacity + Residual Volume = Total Lung Capacity

FVC % of normal at age 20

Total Lung Capacity

Vital Capacity + Residual Volume = Total Lung Capacity
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.