5yo with multiple anomalies and rib fusions
Early in Brace (not by me) film clarifying deformity

- Two 3-level unilateral bars with contralateral hemivertebrae
Age 8, VEPTRs with thoracostomy
Age 10 – curve worsening

Added a growing rod between the two unilateral bars
Age 12

- VEPTRs could not generate sufficient force to prevent trunk tilting
Age 13 – “definitive fusion”
Age 17 – on O2 and methadone
Pulmonary Function in EOS: Where We Have Failed

Confessions of a Greg Redding Disciple

Jim Sanders
Professor of Orthopedics and Pediatrics
University of Rochester
Orthopedists Think Structurally

- EOS makes the chest too small.
- Expand a small chest volume
- Lengthen the spine
- Pull or push the ribs into a better position
Karol, et al: T1-T12 – Based on 28 patients with congenital scoliosis
Restrictive Respiratory Disease

- Loss of lung volume and lung distensibility
- Loss of rib mobility and normal chest wall expansion with inspiration
- Increased reliance on diaphragm function as the primary muscle of inspiration
Additional Consequences of TIS

- Diminished cough effectiveness, with prolonged and severe respiratory infections
- Diminished exercise tolerance
- Airway torque and kinking with localized airway obstruction
- Pulmonary hypertension and cor pulmonale
- Hypercapnia (respiratory failure)
- Postnatal lung growth arrest
Lung Volumes Before and 6 Months After Growing Construct with Rib Based Device Insertion

- TLC Normal
- FRC
- RV

TIS-pre-op
- FVC 62 ± 4%
- RV 77 ± 12%

TIS-post-op
- FVC 54 ± 3%
- RV* 96 ± 16%

n=53

*n=12

Serial FVC Over a 6 Year Interval

28% Reduction from Pre to LAST

16% Reduction Over 6 Years


The Johnston Concept:
What do we really do?

Goal:
- Lengthen spine to make a bigger box.
- Push up the ribs to increase chest diameter.
- Correct lordosis/rotation to improve airway geometry

But:
- Create spontaneous spine fusion
- Create rib fusions minimizing expansion/contraction
- Do we really correct the airway issues?
What about our outcomes? EOSQ-24

### Pulmonary Function: During the past 4 weeks

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<thead>
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<tbody>
<tr>
<td>5.</td>
<td><strong>How difficult has it been for your child to cry/babble/speak (appropriate for age) without experiencing shortness of breath?</strong></td>
<td></td>
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<tr>
<td></td>
<td>Difficult</td>
<td>Somewhat Difficult</td>
<td>Neutral</td>
<td>Somewhat easy</td>
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<tr>
<td>6.</td>
<td><strong>How often has your child experienced shortness of breath during activities?</strong></td>
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<tr>
<td></td>
<td>All of the time</td>
<td>Most of the time</td>
<td>Some of the time</td>
<td>A small amount of the time</td>
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### Physical Function: During the past 4 weeks

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<td>8.</td>
<td><strong>How difficult has it been for your child to move his/her upper body?</strong></td>
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<td></td>
<td>Difficult</td>
<td>Somewhat difficult</td>
<td>Neutral</td>
<td>Somewhat easy</td>
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<tr>
<td>9.</td>
<td><strong>How difficult has it been for your child to sit up on his/her own?</strong></td>
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<tr>
<td></td>
<td>Difficult</td>
<td>Somewhat difficult</td>
<td>Neutral</td>
<td>Somewhat easy</td>
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<tr>
<td>10.</td>
<td><strong>How difficult has it been for your child to keep his/her balance while crawling, walking, or running?</strong></td>
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</tr>
<tr>
<td></td>
<td>Difficult</td>
<td>Somewhat difficult</td>
<td>Neutral</td>
<td>Somewhat easy</td>
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Where have we failed?

- Focus on Cobb angles or spine length
- Both poorly related to what children’s needed pulmonary function
- Outcome measures not adequately addressing the pulmonary domain
What’s needed?

- Effective chest volume expansion and contraction.
  - Intercostal Muscles
  - Diaphragm
  - Costovertebral joints
  - Rib alignment
  - CNS control
  - Adequate baseline volume

- Lung parenchyma that permits effective gas exchange

- Unobstructed airway geometry
Step 1: Adequately determine pulmonary status

- PFTs are hard to get in children.
- Nearly impossible in young children.
- PFTs only provide limited data and not function.
The Redding Construct:

- If children are breathing well, then three things should happen.
- If not, they will be negatively effected:
  - Do they sleep?
  - Do they play(work)?
  - Do they grow?
Pulmonary Function Domains

• Do they sleep?
  – Sleep studies

• Do they play (work)?
  – Need to develop criteria

• Do they grow?
  – Need to define uniform growth parameters for children with differing disorders
Step 2: The orthopedic mental change - start considering chest volume in EOS as dynamic not static

• Our current devices work to make a static box bigger.

• Can we design implants or supports that facilitate rather than block motion?
We need a better paradigm

- It’s not just a bigger box.
- It’s dynamic expansion and contraction with a bigger box—a larger, more effect bellows.
- Listen to Redding—pulmonary function has three main domains:
  - Can they sleep
  - Can they play (work)
  - Do they grow?
- We need to measure them well.