Lung Disease and Early Onset Scoliosis: What Surgeons Need to Know

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November 6, 2015
PRESENTER DISCLOSURE

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I am a Pediatric Pulmonologist!
I had spinal fusion surgery & now I’m all screwed up!

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Objectives

1. Review the effects early-onset scoliosis has on pulmonary physiology and lung function

2. Highlight the gaps in knowledge regarding assessment before and after surgical correction of scoliosis in infants and young children

3. Discuss the ways your local Pediatric Pulmonologist can assist with these challenging cases
Case

• 20 day old Native American newborn female infant born at 36 weeks via C-section transferred by helicopter to the University of Minnesota NICU from Minot, ND

• Known congenital abnormalities of spine and chest wall

• Arrived intubated and ventilated – soon extubated

• Orthopedic, Neurosurgery, General Surgery and Pulmonary consults obtained
What to do next?
Case – Next Steps

• Discussion of VEPTR on the right to help with lung volume – not done until 1-2 years of age

• Would need tethered cord released first

• Would need optimal nutrition (~10 kg)

• Would need to survive until surgery possible

• **At risk for recurrent pneumonia, thoracic insufficiency**
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The Normal Thorax

SPINE

STERNUM

RIBS
The ability to change lung volume depends on:

- Stable diaphragm inserting at T12
- Active rib motion (need separate ribs, intercostal muscle strength and symmetry of the thorax)
The Not Normal Thorax

**Inhibited Lung Growth**
- Low lung volumes
- Asymmetric lung function

**Restrictive Lung Disease**
- Chest wall distensibility and excursion
- Respiratory muscle force and movement

- Hypoxemia
- Sleep disorders
- Cor pulmonale
- Respiratory work
- Tachypnea
- Poor growth
- Exercise tolerance
- Respiratory failure
- Recurrent pneumonia
Thoracic Insufficiency Syndrome

- The inability of the thorax to support respiration:
  - Increased respiratory rate
  - Loss of intrathoracic volume
  - Loss of chest wall motion
  - Decreased exercise tolerance
- The inability of the thorax to support lung growth
- Treatment should increase thoracic volume and stabilize chest-wall defects without spine fusion

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Assessment Tools

- History of respiratory symptoms (decreased exercise tolerance, tachypnea, recurrent pneumonias, hypoxemia, hypercarbia, growth, etc.)

- CT/MRI imaging (Cobb angle, pelvic inlet width, T1-T12 height, T1-S1 height, coronal chest width, etc.)

- Pulmonary function tests (FVC and FEV1)

- Polysomnography (sleep disordered breathing, AHI)

Brief Review of Lung Volumes

Forced Vital Capacity (FVC): Amount of gas that can be exhaled from the lungs after a maximal inhalation

Residual Volume (RV): Amount of gas remaining in the lung after complete exhalation

Total Lung Capacity (TLC): The total amount of air in the lungs at maximal inflation

Functional Residual Capacity (FRC): Amount of air left in the lungs at the end of a passive exhalation

\[
FRC = ERV + RV
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Pulmonary Function Tests (PFTs)

- Used to assess for restrictive (decreased FVC) or obstructive lung disease (decreased FEV1 and decreased FEV1/FVC)

- Most children can’t perform until age 6

- Results compared to a “predicted” result which is based on standing height (can be inaccurate in spinal deformities)

Dreimann et al. Spine Deformity 2014.
Infant Pulmonary Function Testing

**Measurements:**
- FVC (Forced Vital Capacity)
- FEV 0.5 (Similar to FEV1)
- FRC (Functional Residual Capacity)

**Requirements:**
- Specialized equipment
- Expertise in the technique
- Sedated procedure
Case

- During first year of life:
  - Multiple hospitalizations for pneumonia
  - Respiratory insufficiency (oxygen, BiPAP dependent)
  - Poor growth and nutrition (g-tube placed)
- No infant pulmonary function tests performed
- Re-evaluation for VEPTR:
  - Needed to gain weight
  - Needed to improve pulmonary status
  - Needed to have tethered cord released
Case

• Decision made to try and optimize nutrition and pulmonary status before any surgery

• Socially complex situation – child was transferred back to Minot, SD to gain weight

• Suffered a respiratory arrest in the local hospital and died before any surgical intervention
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How we can help...

- These infants and children are often complex

- Need to partner to standardize preoperative and post-operative assessments using both structural and functional measures

- We can perform infant pulmonary function tests at the University of Minnesota

- We are happy to see your patient pre-operatively in clinic to help assess risk and plan the post-op period
Thank you

Questions?

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