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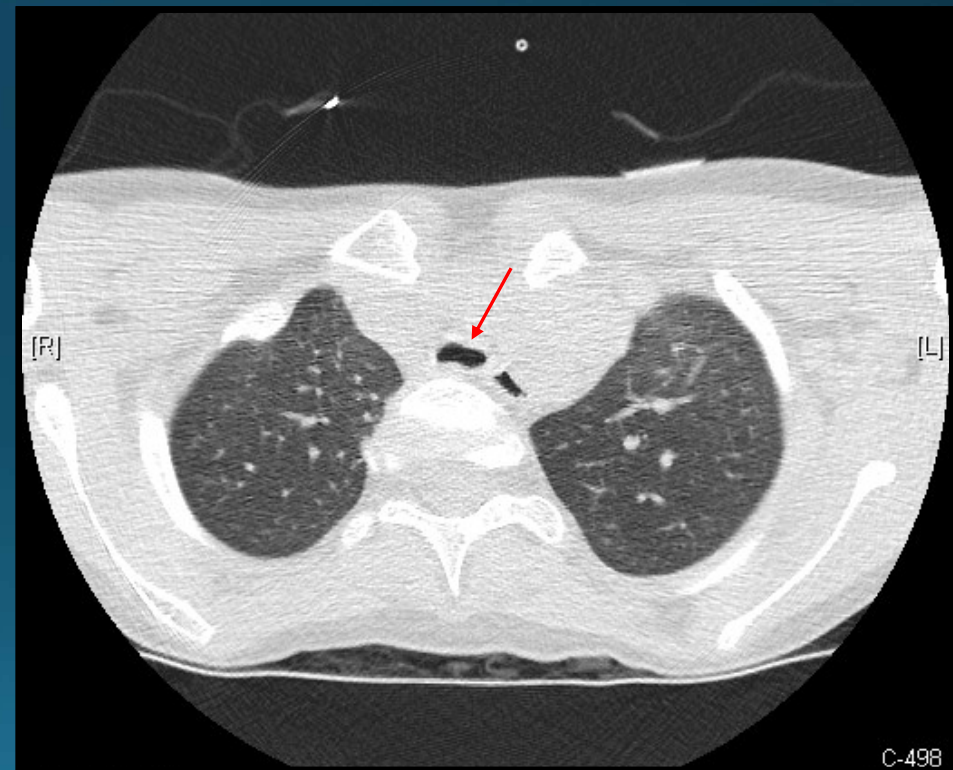
What do you do
without a
pulmonologist to
help manage
children with EOS

Disclosures

- Pediatric Pulmonary Section Editor, UpToDate

Pulmonary Issues related to EOS

- Restrictive lung disease
- Obstructive lung disease due to airway compression, torque and stretch
- Respiratory muscle weakness due to tethering, loss of intercostal muscle function
- Unilateral loss of lung function
- Hypoxemia during sleep and poor sleep quality
- Exercise intolerance/activity limitation
- Failure to thrive
- Pulmonary Hypertension and Early Death



Difficulties with Children Too Young or Disabled to Perform Lung Functions

- Children < 5 years old, inexperienced children of all ages
- Children with tracheostomies or developmental delay
- Accounts for 50% of all children in CSSG registry
- Spine interventions are required early in many children too young to do spirometry.
- So what to use as surrogates that reflect abnormal lung function?

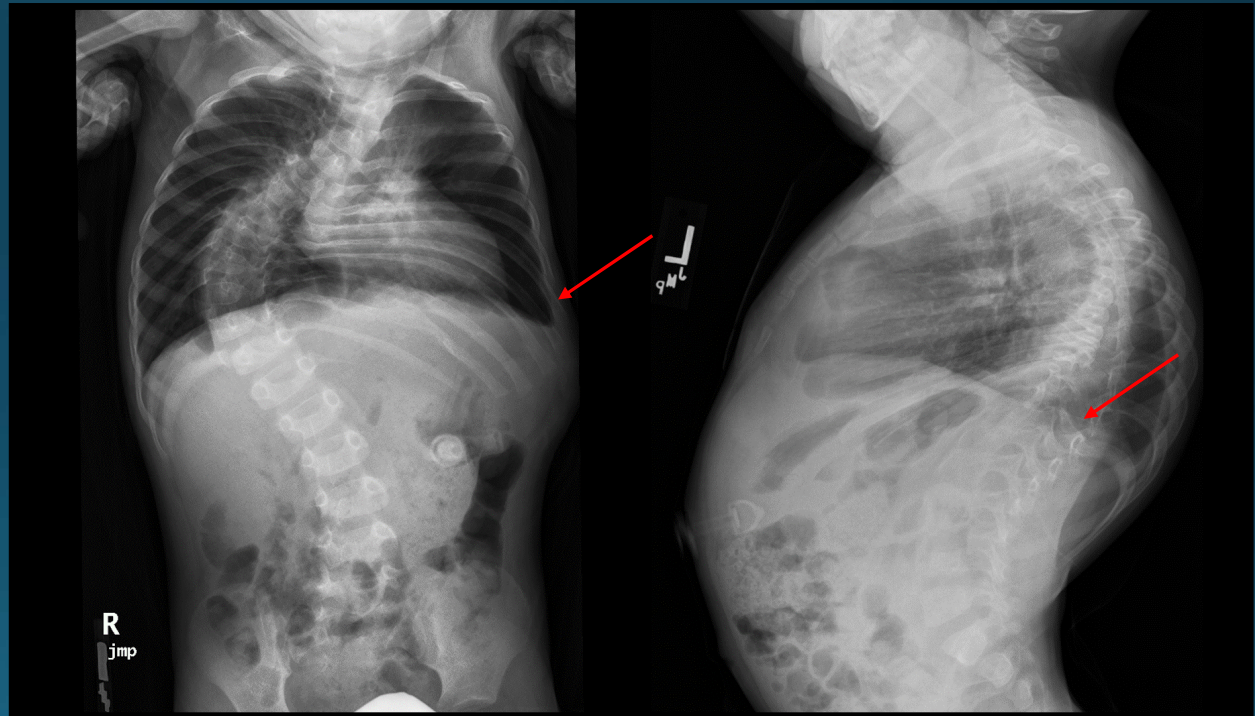
Pertinent Pulmonary Physical Findings

- Anterior Chest Wall shape (pectus excavatum, rib gaps, asymmetry in shape)
- Chest wall depth (pectus gracilis) often with thoracic hypokyphosis or lordosis
- Asymmetric breath sounds: may reflect reduced air entry or obstructed airways which make louder sounds
- Short expiratory abdominal push. “double pump”
- Tonsil size (maximal 3-5 years of age)
- Low weight for age (<2 years old) or BMI using arm span for height (<10% for age)



Radiographic Findings

- Evidence of parenchymal or airway disease complicating scoliosis
- Low lung volumes (often more apparent in the sagittal view)
- Pectus gracilis
- Flat diaphragm contour with no zone of apposition
- Heart vs Chest wall size (often due to small chest wall dimensions)



Diagnostic Tests that Don't Require Effort

- Lung perfusion scans to quantitate asymmetric lung function over time
- Overnight polysomnograms (sleep studies): Hypoxemia often due to EOS plus enlarged tonsils or adenoids in kids <5 years old.
 - 93% of children with EOS have abnormal sleep indices including hypoxemia*
 - 56% of sleep studies in children with EOS were abnormal enough to* prescribe BIPAP, CPAP, or night time oxygen use long-term.
 - Polycythemia is likely related to sleep-related hypoxemia at night.
- Serum electrolytes for elevated serum bicarbonate which may reflect nighttime hypercarbia
- Echocardiogram for pulmonary hypertension

Conclusions

- Abnormal physical and radiographic findings provide qualitative information about poor lung function in children too young to perform spirometry.
- The relative “badness” for each of these or combination of these features has not been determined.
- Serial assessments of these features should be correlated with progression of spine and chest wall deformities.
- New methods to quantitate regional lung function and thoraco-abdominal synchrony will improve this assessment.