Learning Objectives

- To summarize the various imaging characteristics of congenital pulmonary malformations
- To list two reasons for the conceptual shift to approaching congenital lung diseases as a spectrum of abnormalities

The Conceptual Shift

- Isolated, distinct lesions to overlapping, hybrid lesions

Why?
- Information from prenatal ultrasound
- Prevalence of concurrent lesions
- Frequent occurrence of lesions not fitting into existing classification systems

Overlapping Spectrum of Congenital Lung Malformations
Characterization of Congenital Lung Malformations

- Tracheobronchial tree
- Parenchyma
- Arterial Supply
- Venous Drainage

Bronchogenic Cyst

- Abnormal budding from tracheobronchial tree
- No communication with airway
- Respiratory epithelium and cartilage
- Mostly paratracheal and subcarinal

- Symptoms result of mass effect or superinfection
- Imaging
  - Fluid or soft tissue density
  - Thin, well-defined wall
  - Lack internal enhancement
- Associations
  - Congenital Lobar Hyperinflation
  - Bronchial atresia

Case courtesy of John Engels MD

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  - Bronchial atresia
Bronchial Atresia

- Atresia of central bronchus, usually segmental
- Distal airways develop normally and become mucous impacted (mucocele or bronchocele)
- Etiology unclear
  - Secondary insult during development, possibly vascular
- Left upper lobe most commonly reported location
- Usually incidental finding in adult
  - Superinfection is possible

Imaging
- Branching tubular opacity (mucocele)
- Overinflation secondary to collateral air drift
- No systemic feeding artery
- No abnormal venous drainage
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**Intralobar Sequestration**

- Nonfunctioning pulmonary tissue isolated from the tracheobronchial tree
- Systemic arterial supply
  - Descending thoracic aorta
  - Celiac, intercostal, subclavian, etc.
- Lack separate pleural covering
- No longer considered acquired lesions
  - "Bronchial Atresia Sequence"
- Incidental or Superinfected

- Imaging
  - Lack communication with tracheobronchial tree
  - Can communicate with esophagus/GI tract
  - Usually normal pulmonary venous drainage, but can be systemic
  - Cystic or Solid

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Extralobar Sequestration

- Pulmonary tissue developing separate from normal lung
  - Separate pleural covering
  - Surgical importance
  - Systemic arterial supply
  - Slightly higher incidence of systemic venous return
  - May have parenchymal microcystic maldevelopment (CCAM)
  - Associated with diaphragmatic defects, pulmonary hypoplasia

Congenital Pulmonary Airway Malformation

- Disorganized lung tissue with bronchiolar proliferation and cystic change
  - Communicate with airways and usually have pulmonary arterial supply
Stocker classification (Expanded)

- Type 0 = Solid with small firm lungs
- Type I = large cysts (>2cm)
- Type II = multiple small cysts (<2cm)
- Type III = solid, microcystic lesion
- Type IV = large cysts, peripheral

Not true “cyst”, rather dilated bronchioles

Symptoms related to mass effect/midline shift or superinfection

Stocker Type I lesions
- Association with bronchoalveolar carcinoma

Stocker Type II lesions
- Associated with bronchial atresia and sequestration

Stocker Type III lesions
- Extreme of adenomatoid pulmonary hyperplasia

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Congenital Lobar Hyperinflation

Overinflation, not destruction

Intrinsic or extrinsic airway obstruction

Upper lobes (left) most common

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Swyer James Syndrome

- Unilateral acquired bronchiolitis obliterans, presumably due to prior infection
- Affected lung smaller
- Air trapping on exhalation
- Also may see bronchiectasis

Congenital Lung Malformations: Summary

- Cyst separated from tracheobronchial tree
- Fluid attenuation on CT = Stop
- Soft tissue density on CT = MR with contrast

- Focal airway atresia with distal air trapping
- Mucocele
- Minimum Intensity Projection to accentuate air trapping
Congenital Lung Malformations: Summary

- Isolated pulmonary tissue from the tracheobronchial tree with systemic feeding artery
  - Multiplanar reformats
  - Maximum intensity projection images

- Disorganized lung tissue with tracheobronchial communication
  - Multiloculated cystic lesions
  - Consider hybrid or associated lesions

- Post-obstructive overinflation
  - Hyperinflation with midline shift / mass effect
  - Minimum intensity projection images accentuate air trapping

- Be informative:
  - Tracheobronchial tree
  - Arterial supply
  - Venous drainage
  - Anatomic Localization