Congenital Lung Lesions
Neonates to Adults

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Conflict of Interests

• None
Learning Objectives

• Describe imaging appearances of the common congenital anomalies
• Correlate with pathologic features
• Emphasize “clues” to diagnosis
Congenital Lung Anomalies

- **Normal vascularity**
  - Lobar emphysema
  - Cystic adenomatoid malformation
  - Bronchogenic cyst
  - Bronchial atresia
  - Pulmonary agenesis, hypoplasia

- **Abnormal vascularity**
  - Scimitar syndrome
  - Sequestration
  - Arteriovenous malformation
Congenital Lobar Emphysema

• Misnomer; true emphysema not present
• Lobar “emphysema” = “overinflation” from bronchial obstruction
  – absence of bronchial cartilage
  – intraluminal web
• Onset: 50% 1st week
  – 90% < 6 months
  – dyspnea, cyanosis, cough
Lobar Emphysema: LUL

- Lobar hyperinflation
- Atelectatic adjacent lung
- Mediastinal shift
- Lobar predilection:
  - LUL > RML > RUL
DDX: Swyer-James Syndrome

- Bronchiolitis obliterans
  - viral infection in childhood **
- Imaging findings
  - unilateral hyperlucent (low attenuation) lung
  - small or normal size **
  - bronchiectasis **
  - air-trapping on expiration

** Helps to differentiate from CLE
Cystic Adenomatoid Malformation

- 90% symptomatic as neonates
  - Cyanosis, grunting, tachypnea
- 10% older children & adults
  - Presents as pneumonia or recurrent infection
- May be antenatal diagnosis
CCAM: Histologic Types

- Type I: (50%) Large cyst(s) (> 2 cm)
- Type II: (40%) Multiple cysts (< 2 cm)
- Type III: (10%) Microcysts on cut-section

Stocker JT. Hum Pathol 1977; 8:155-171
CCAM: Imaging

- Air-filled mass, mediastinal shift
- Type I
  - At least one dominant cyst
- Type II
  - Small cysts
- Type III
  - Solid mass, usually not imaged
  - High perinatal mortality
Cystic Adenomatoid Malformation
CCAM: Adults

- 10% diagnosed in adolescents & adults
- Infection common
- Imaging studies
  - Thick walled cavities
  - Fluid-fluid levels
  - Pneumonia
Bronchogenic Cyst

- Lung (30%)
  - mediastinum (70%)
- Incidental finding or symptoms from mass effect
- Path
  - Lined by respiratory mucosa
  - Clear or mucoid contents
Bronchogenic Cyst: Imaging

- Round, *unilocular* mass
- Air-filled +/- fluid
Bronchial Atresia
(Congenital Mucocele)

- Failure of bronchial bud to maintain communication with airway
- Wheezing, cough or incidental detection
- Imaging:
  - nodular opacity
    - fluid-filled dilated bronchi
  - surrounding emphysema (distal air drift)
Bronchial Atresia

Nodular/tubular opacity & overaeration
Arrested Pulmonary Development

- **Agenesis**: Complete absence of lung tissue, artery, & small or absent bronchus
- **Hypoplasia**: Small lung & bronchus (artery may or may not develop)
Adult: Lung Agenesis

Absent lung & artery--small bronchus
Pulmonary Hypoplasia Adult

Absent pulmonary artery - small lung & bronchus
Congenital Anomalies with Abnormal Vasculature

- Hypoplasia with anomalous venous return
  - Scimitar syndrome
- Pulmonary sequestration
- Arteriovenous malformation
Hypogenetic Lung Syndrome

- AKA scimitar syndrome
- Lung hypoplasia with PAPVR
  - RLL vein returns to IVC, portal or hepatic vein or RA
- May be symptomatic or incidental finding
Hypogenetic Lung Syndrome
Pulmonary Sequestration

- No normal connection with bronchial tree or pulmonary arteries
- **Systemic** blood supply
- 2 types
  - Intralobar (acquired)
  - Extralobar (congenital)
<table>
<thead>
<tr>
<th><strong>Extralobar</strong></th>
<th><strong>Intralobar</strong></th>
</tr>
</thead>
<tbody>
<tr>
<td>Own pleura</td>
<td>Shared pleura</td>
</tr>
<tr>
<td>Neonate</td>
<td>Children &amp; adults</td>
</tr>
<tr>
<td>90% left</td>
<td>90% left</td>
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<tr>
<td>Syst. arterial supply</td>
<td>Syst. arterial supply</td>
</tr>
<tr>
<td>Syst. venous drainage</td>
<td>Pulm. venous drainage</td>
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</tbody>
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Sequestration

- Intralobar (infected)
  - Symptomatic
  - Cough, recurrent pneumonia
- Extralobar (not infected)
  - Asymptomatic, incidental finding
Sequestration: CT Features

- Anomalous feeding artery
- Drains to pulmonary or systemic veins
- Parenchymal findings
  - Intralobar:
    - Infiltrate &/or abscess
  - Extralobar
    - Triangular/round solid mass
Which Sequestration?

ILS

ELS
Arteriovenous Malformation

- 80% Hereditary telangiectasia (OWR)
  - 15% sporadic
  - 5% cardiac surgeries (Glenn or Fontan)
- Symptomatic in older patients (cyanosis, polycythemia, dyspnea)
- 80-90% are simple AVMs
  - single feeding and draining vessel
Pulmonary AVM

Simple architecture
Congenital Lung Anomalies

ABNORMAL LUNG
NORMAL VASCULATURE

CLE  Cyst  CCAM  Hypoplasia  Sequestration  Scimitar  AVM

NORMAL LUNG
ABNORMAL VASCULATURE