Imaging Evaluation of Congenital Lung Lesions in Children

Donald P. Frush, MD

No Disclosures
Current Perspectives

• **Etiologies:** alone or in combo
  – Obstruction, budding, vascular, genetic

• **Rename CCAM ..... CPAM**
  – congenital pulmonary airway malformation
  – not all are adenomatoid
  – types 0 - IV .... cystic and solid components
Current Perspectives

• Location, location, location
• Identify *all components*
  – location, extent, lung and vascular features
• “Elective” surgery
  – Follow asymptomatic, “small” lesions
• Hybrid (or overlap) lesions
Hybrid Lesions

- CCAM most frequent companion
  - e.g. sequestration, foregut cysts
- Sequestration probably second
  - e.g. CPAM, lung hypoplasia
Neonate
Hybrid lesion:

CPAM
Sequestration
Why MDCT?

- Fast: entire chest <1.0 second in neonate
  - Less sedation
  - Less movement artifact
- Better global assessment
  - than US
- Best lung evaluation
When do you get a CT?

- **Fetus:** suspected anomaly by US or MR
  - Even with "normal CXR"
- **Neonate:** persistent
  - Mass or persistent opacity
  - Abnormal aeration (including air-filled cyst)
  - Congestive failure (very rare)
- **Older child:**
  - Mediastinal mass (posterior…consider MRI)
  - “Fixed” lung mass/opacity/ x 3
  - Substantial…or fixed aeration abnormality x 3
  - Air-filled cyst without explanation
IV Contrast

- Non ionic, low osmolar
- (300) – 370** mgI/mL
- Bolus tracking in young child
Scan Parameters

- Use low kVp (80-100 under 10 yrs)
- routine (e.g. chest CT) mA
- Fast gantry cycle time
- Thin collimation
- Multiplanar/3D recons
The Big Four

Bronchogenic Cyst

CPAM (CCAM)

Congenital Lobar Emphysema (CLE)

Pulmonary Sequestration
The Big Four

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Imaging:

• **CXR:**
  – mediastinal mass (67%)
  – Parenchymal (i.e., hilar) mass (33%)

• **CT:** preferred modality
  – non-enhancing, thin-walled, CT
  – density depends on nature of fluid
  – no calcification
  – 2-10 cm in size
Bronchogenic Cyst
Differential Considerations

- Lung cyst
- Esophageal duplication cyst
- Teratoma (mature, usually benign)
- Pericardial cyst
- Lymphatic malformation
The Big Four

Bronchogenic Cyst

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Pulmonary Sequestration
CCAM

Imaging:

- Often detecting by fetal US or MRI
- Radiography: may be initially solid
- CT: variable sized cystic airspaces
- The 1 of 4 that can occur anywhere

In Garcia Pena Acta Radiol 2013

<table>
<thead>
<tr>
<th>Type</th>
<th>Percentage</th>
<th>Description</th>
</tr>
</thead>
<tbody>
<tr>
<td>Type 0</td>
<td>Rare</td>
<td>Acinar dysgenesis or dysplasia. Cardiovascular anomalies</td>
</tr>
<tr>
<td>Type 1</td>
<td>50–60%</td>
<td>Large cystic lesion (1–10 cm) with smaller cysts around</td>
</tr>
<tr>
<td>Type 2</td>
<td>10–40%</td>
<td>Multiple small cysts (0.5–2 cm). Cardiovascular and renal malformations may be associated</td>
</tr>
<tr>
<td>Type 3</td>
<td>5–10%</td>
<td>Adenomatoid type: small cystic lesions with solid aspect</td>
</tr>
<tr>
<td>Type 4</td>
<td>10–15%</td>
<td>Hamartomatous malformations with cysts at the lobe’s periphery</td>
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</tbody>
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Adapted from Stocker (11)
CCAM
Differential Considerations

- Congenital diaphragmatic hernia
- Diaphragm paralysis, paresis
- Pneumatocele
- CLE (by CXR; rare)
- Complicated pneumonia
- Other causes of nodules (usually adult)
The Big Four

Bronchogenic Cyst

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Pulmonary Sequestration
Congenital Lobar Emphysema

Imaging:

- **Location:**
  - Left upper lobe: 40-50%
  - Right middle lobe: 30-40%
  - Right upper lobe: 20%

- **CXR:** hyperinflation with contralateral shift; neonate: initially opaque

- **CT:** emphysema
  - Look for acquired causes: endo or exobronchial
Sublobar Emphysema/Bronchial Atresia
Sublobar Emphysema/Bronchial Atresia
Congenital Lobar Emphysema
Differential Considerations

• Acquired emphysema
• Bronchiolitis obliterans
• Contralateral volume loss
• CCAM (on CXR; rare)
The Big Four

Bronchogenic Cyst

CPAM (CCAM)

Congenital Lobar Emphysema (CLE)

Pulmonary Sequestration
## Comparison of Pulmonary Sequestration

<table>
<thead>
<tr>
<th>Feature</th>
<th>ELS</th>
<th>ILS</th>
</tr>
</thead>
<tbody>
<tr>
<td><strong>Occurrence</strong></td>
<td>Rarer</td>
<td>More common (4-6 times ELS)</td>
</tr>
<tr>
<td><strong>Sex ratio (M:F)</strong></td>
<td>Higher in males (4:1)</td>
<td>Approximately equal (1.5:1)</td>
</tr>
<tr>
<td><strong>Age at diagnosis</strong></td>
<td>60% &lt; 1 yr</td>
<td>50% &gt; 20 yr</td>
</tr>
<tr>
<td><strong>Location</strong></td>
<td></td>
<td></td>
</tr>
<tr>
<td><strong>Lung</strong></td>
<td>Left lung (90%)</td>
<td>Left lung (66%)</td>
</tr>
<tr>
<td><strong>Parenchyma</strong></td>
<td>Costodiaphragmatic sinus or below diaphragm</td>
<td>Posterior basal segments of the lower lobes; lingula</td>
</tr>
<tr>
<td><strong>Pleura</strong></td>
<td>Separate pleural coverage (pleural investment)</td>
<td>Enclosed within the visceral pleura of the lobe</td>
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* ELS = extralobar sequestration; ILS = intralobar sequestration. Bone: Pulmonary & Critical Care Medicine, 1998 ed; ML Rosado de Christenson STR Meeting San Francisco, 2002
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<tr>
<th>Feature</th>
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<tr>
<td>Pathology</td>
<td>Ectopia of lung tissue</td>
<td>Closed, nonfunctional segment with no connection to normal bronchial tree</td>
</tr>
<tr>
<td>Artery</td>
<td>Systemic: usually arteries from abdominal aorta (80%)</td>
<td>Systemic; longer vessels, from descending aorta (supra- or infradiaphragmatic); 5% from intercostal arteries</td>
</tr>
<tr>
<td></td>
<td>Pulmonary: rarely</td>
<td></td>
</tr>
<tr>
<td>Vein</td>
<td>Systemic 80% (usually azygous system)</td>
<td>Pulmonary veins; rarely systemic</td>
</tr>
<tr>
<td>Assc. defects of</td>
<td>60%</td>
<td>Rare</td>
</tr>
<tr>
<td>diaphragm</td>
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<td>Other congenital</td>
<td>Common (&gt; 50% cases) pericardial cysts; congenital heart disease</td>
<td>Rare; similar anomalies to those found in ELS</td>
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<tr>
<td>defects</td>
<td></td>
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<tr>
<td>Clinical manifestation</td>
<td>Respiratory distress; infection less common</td>
<td>Lower lobe pneumonia</td>
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<tr>
<td>Radiologic appearance</td>
<td>Supra- or infradiaphragmatic opaque lesion</td>
<td>Circumscribed mass (triangular or round) contiguous to the diaphragm</td>
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Pulmonary Sequestration Imaging:

- Radiology: usually solid mass, lower lobe, posterior
- Goal of imaging: identify systemic arterial supply
- MDCT preferred
11 yo male; 4 yrs recurrent pneumonia

Courtesy Murat Kocaoglu, MD
Congenital Lung Abnormalities

Conclusions

• Imaging plays critical role
• After CXR, MDCT is most useful
• Technique is critical
• Be descriptive: identify all components
References

