Developmental Anomalies of the Lungs

Catherine Kier, M.D.
Professor of Clinical Pediatrics
Stony Brook Medicine, Stony Brook, New York
Division Chief, Pediatric Pulmonary
Director, Cystic Fibrosis Center
Director, Pediatric Sleep Disorders Center
Objectives

• Objective #1
  review developmental, clinical and radiographic features of lung anomalies

• Objective #2
  review medical and surgical approaches of therapy
Embryonic period (4th week)

Lung bud from ventral foregut
Stages of Lung Development

- Pseudoglandular
- Canalicular
- Saccular
- Alveolar
Pulmonary vasculature

Airway epithelium and vascular epithelium

Pulmonary arteries form after and follow the arteries
Congenital lung anomalies

• Vascular versus non-vascular
• Location of the anomaly
• Predicts when mishap happens in development
• Associated conditions
Congenital lobar hyperinflation
Adult male wheezing
Congenital lobal hyperinflation

• Misnomer: lobar emphysema
• Etiology: partial bronchial obstruction
  • Deficient bronchial cartilage
  • Intraluminal web, stenosis, malacia
• Onset: 90% <6 months
  • neonates: dyspnea, cyanosis, cough
  • older patient: wheezing or incidental
Bronchogenic Cyst

Well-defined, spherical cystic lesion
Thin wall
Bronchogenic Cyst

- Majority in middle/posterior mediastinum (80%)
- Asymptomatic and incidental finding
- Symptomatic from mass effect
Bronchogenic Cyst

- Abnormal foregut budding
- Lined with respiratory epithelium
- Mural cartilage, smooth muscle, mucous glands
Bronchogenic Cyst (adult)

Unilocular; air-filled
Differential diagnosis: CPAM
congenital pulmonary airway malformation

Multiple cysts; septations
CPAM =
congenital pulmonary airway malformation

• Formerly, congenital cystic airway malformation (CCAM)
• 25% of congenital lung lesions
• Abnormal mass of pulmonary tissue with cystic change
  • Result of abnormal airway proliferation
  • Normal arterial supply and venous drainage
CPAM

Type I (3-10 cm cysts) 70%

Type II (1-3 cm cysts) 15-20%

Type IV (up to 10 cm) 15%

Type II – associated with other abnormalities (renal agenesis, pulmonary sequestration, cardiac anomalies)
CPAM = congenital pulmonary airway malformation

- Type III – 5-10%, microcysts <5mm, can appear solid (high mortality)
  - Cysts not imaged
  - Fetal hydrops
    - Ascites, anasarca, placental edema
Bronchial Atresia

- Congenital mucocoele
- Failure of bronchial bud to maintain communication with airway (atresia of central segmental bronchus)
- Wheezing, cough or incidental finding

Fluid-filled (mucus) dilated bronchi; overinflated surrounding lung (air drift)
Pulmonary underdevelopment

• Agenesis
  - Complete absence of lung tissue, artery, and small or absent bronchus

• Hypoplasia
  - Small lung and bronchus (artery may or may not develop)
Lung Agenesis

Small bronchus
No lung or pulmonary artery
Lung Agenesis - in an adult
Pulmonary hypoplasia

- Arrested pulmonary development
- Small lung (hypoplasia)
- Small bronchus
- Absent or small pulmonary artery
- Mediastinal shift to side of hypoplasia
Pulmonary hypoplasia

2 month old male presenting with mild dyspnea
Pulmonary hypoplasia
Pulmonary hypoplasia
Pulmonary hypoplasia

Small lung and bronchus
No pulmonary artery
Potter syndrome

- Oligohydramnios
- Cause: renal agenesis
- Small lungs due to compression of the fetal thorax
- Pneumothorax and no other lung disease
- Flattened nose, small chin, low-set ears
Potter syndrome

Small bell-shaped chest, clear lungs, pneumothorax
Congenital Anomalies
with Abnormal Vasculature

- Hypoplasia with anomalous venous return
  - Scimitar syndrome
- Pulmonary sequestration
Scimitar syndrome

• Hypogenetic lung syndrome
• Lung hypoplasia with vascular anomaly
  • PAPVR (partial anomalous pulmonary venous return)
  • Right lower lobe (RLL vein) returns to the inferior vena cava (IVC), portal or hepatic vein or right atrium (RA)
  • Normally, should return to pulmonary vein

• May be symptomatic or incidental finding
  • Infants may present congenital heart disease or pulmonary sequestration
  • Adults are generally asymptomatic
Scimitar syndrome

- abnormal vein on the chest x-ray creates a gentle curve bulging into the right chest from the mediastinum
- resembles the Turkish sword scimitar
Scimitar syndrome
Pulmonary sequestration

• Systemic blood supply instead of bronchial or pulmonary arteries
• Arises from supernumerary lung bud
• If the lung bud arises before the pleura develops, it shares pleura of the adjacent lung (INTRALOBAR)
• If the lung bud arises after the pleura develops, it acquires its own pleural covering (EXTRALOBAR)
Pulmonary sequestration (systemic arterial supply)

Intralobar
- majority (75-85%)
- present as recurrent infections (infiltrate or abscess)
- pulmonary venous drainage

Extralobar
- less common (15-25%)
- infra-diaphragmatic (10%)
- could present in the neonatal period with respiratory distress, cyanosis, or infection (solid mass)
- systemic venous drainage
Pulmonary sequestration

Intralobar

Extralobar
Bronchopulmonary sequestration
Pulmonary arteriovenous malformation

• abnormal communications between the pulmonary arterial and venous systems without interposed capillaries
• can lead to highoutput cardiac failure
• symptoms
  • unusual in childhood
  • adulthood, exertional dyspnea; hemoptysis in patients with cutaneous telangiectasis
  • continuous bruit is often heard over the lesion
  • Brain/liver abcess (associated AVM in other organs)
Pulmonary arteriovenous malformation
Summary
Congenital lung anomalies

Abnormal lung
Normal vasculature

CLH = congenital lobar hyperinflation
Cyst = bronchogenic cyst
CPAM = congenital pulmonary airway malformation
BA = bronchial atresia
Parenchymal agenesis, hypoplasia

Normal lung
Abnormal vasculature

CLH Cyst CPAM BA Hypoplasia Sequestration Scimitar AVM

Pulmonary sequestration
Scimitar syndrome
AVM = arteriovenous malformation
Think outside the box if does not fit
Infected CPAM
congenital pulmonary airway malformation
CPAM with systemic arterial supply