American Pediatric Surgical Association

Standardized Toolbox of Education for Pediatric Surgery

Congenital Pulmonary Airway Malformations

APSA Education Committee 2019-2020

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Congenital Pulmonary Airway Malformations

- **Laura Hollinger, MD**
  - **Medical University of South Carolina**

- **Edited by:**
  - Grace Mak, MD (University of Chicago)
  - Steven Lee, MD (UCLA Mattel Children's Hospital)

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History

- **Term newborn born earlier today with prenatal diagnosis of fetal lung malformation**
  - Baby initially required CPAP, but then was weaned to room air
  - Prenatal ultrasound: right sided cystic lung lesion with a CCAM Volume Ratio (CVR) of 1.1
  - No mediastinal shift
  - No diaphragm flattening
  - No hydrops
  - No feeding vessel identified

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History

Echogenic CPAM on prenatal ultrasound

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History Discussion Slide

What history do you want to know?

- **Age:** Term baby, day of birth
- **Mass:** Prenatal u/s @21 weeks: right sided cystic lung lesion, CVR 1.1, no diaphragm or mediastinal shift. No hydrops
- **Birth hx:** Spontaneous vaginal delivery, CPAP required initially, then weaned to room air
- **Medical hx:** uneventful pregnancy; maternal steroids given
- **Family hx:** none
- **ROS:** baby is now breathing normally on room air

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Differential Diagnosis

- **Lung lesions**
  - Congenital Pulmonary Airway Malformation (CPAM)
  - Bronchopulmonary sequestration (BPS)
  - Congenital Lobar Emphysema
  - Bronchogenic Cyst
- **Other lesions**
  - Enteric duplication cyst
  - Fetal lung interstitial tumor
  - Cystic Pleuropulmonary Blastoma (PPB)
  - Congenital diaphragmatic hernia (CDH)
  - Vascular malformation

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Lung Lesion Definitions

Congenital Pulmonary Airway Malformation (CPAM)

- Abnormal development of lung tissue with blood supply from pulmonary system
- Classically communicate with lung tissue via bronchial airways
- Can be solid or cystic
- Lower lobes more common

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Lung Lesion Definitions

Bronchopulmonary sequestration (BPS)

- Abnormal development of lung tissue with systemic blood supply
- Lack of communication with bronchial airways
- Usually solid
- Can be intra- or extra-lobar
- Lower lobes more common

Bronchogenic Cyst

- Abnormal development from tracheo-bronchial airway
- Occurs centrally in mediastinum

Congenital Lobar Emphysema

- Congenital Lobar Overinflation
- Left upper lobe most common

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Physical Exam

- **What specifically would you look for?**
  - Vitals: P 120, RR 30, BP 70/50, T 99F
  - Appearance: Healthy term male, comfortable and swaddled in warmer
  - PE exam findings:
    - No retractions
    - No increased work of breathing
    - No palpable masses
    - No wheezing

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Studies (Labs, Imaging)

- **What labs are needed?**
  - None

- **What imaging is needed?**
  - Chest X ray initially
  - CT Chest with IV contrast
    - Closer to time of surgery

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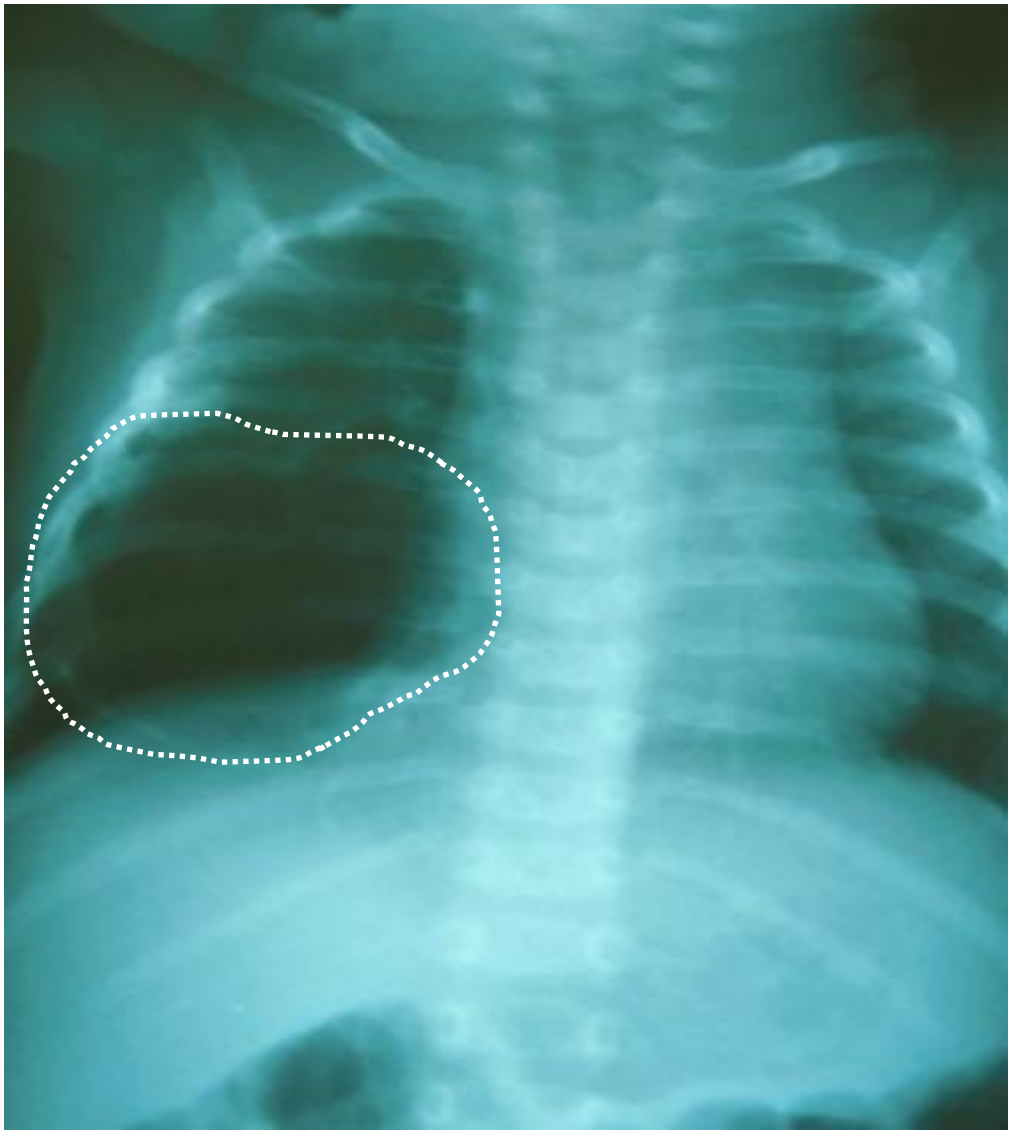
Study Results



Large cystic lung lesion in right chest

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Case Discussion

- **Diagnosis**
  - Congenital Pulmonary Airway Malformation (CPAM)
- **Management:**
  - Pre-op: Imaging (Chest XR, CT scan with angiography)
  - Operative: Lobar resection

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Interval Steps Before Surgery

- **Chest X-ray**
  - Rule out potentially life-threatening issues (mediastinal shift, pneumothorax, larger space occupying lesion)
  - Not sensitive enough to see all lesions
- **CT Chest with angiography or IV contrast**
  - Typically performed after neonatal period (age 3-6 months)
  - Allows for anatomic operative planning (i.e. vascular supply)

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Interval Steps Before Surgery

- **Operative planning/Timing of surgery**
  - Symptomatic patient: quickly after birth
  - Asymptomatic patient: sometime in the first year of life to allow for post-resection compensatory lung growth
  - If active infection, treat with antibiotics initially then schedule interval surgery 6-8 weeks later once infection adequately treated

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Interval Steps Before Surgery

- **Indications for surgery**
  - Symptoms
  - Future complications of infection (pneumonia), pneumothorax, bleeding, heart failure, malignant degeneration

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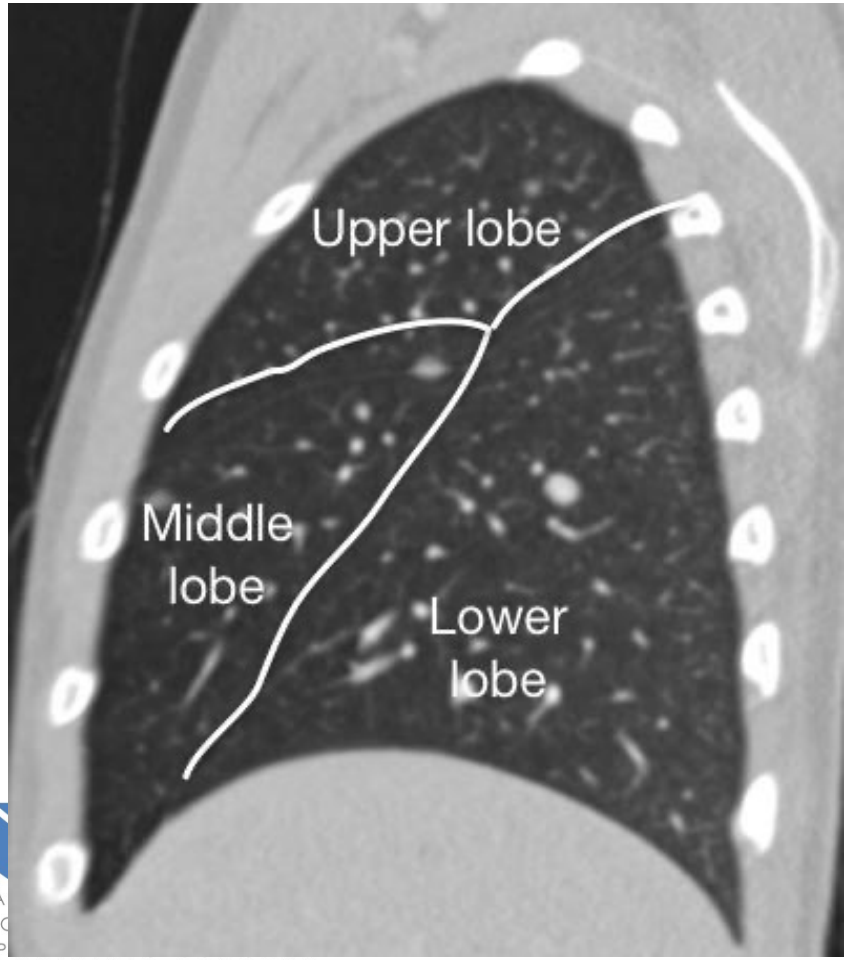
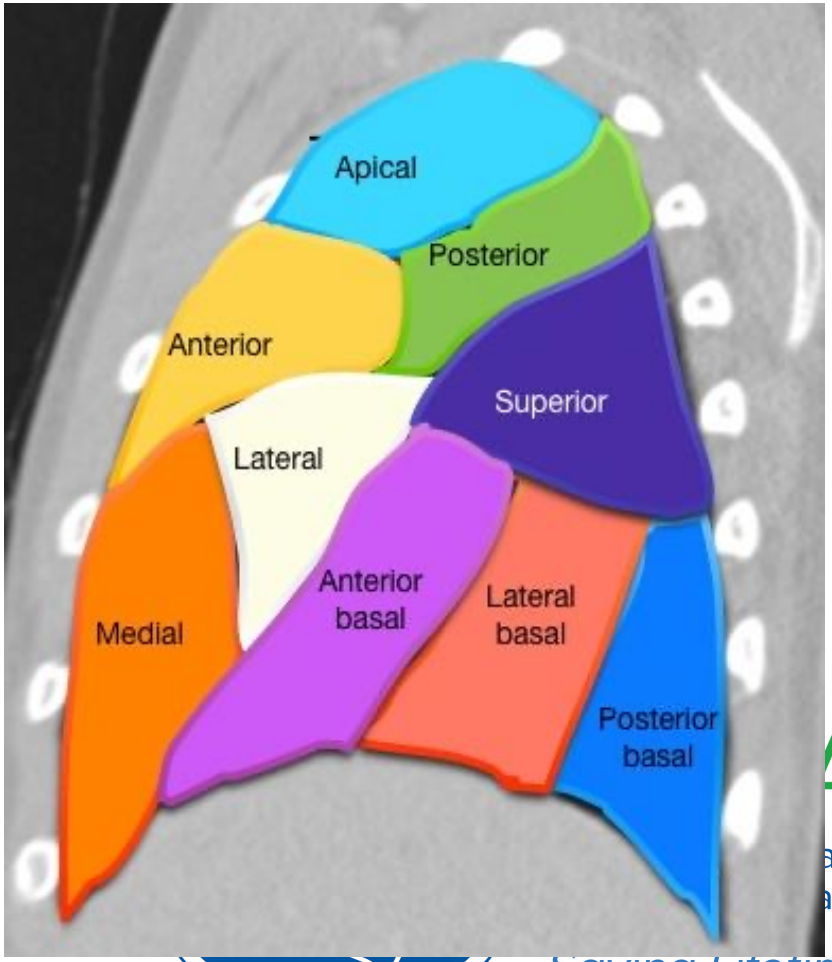
Operation

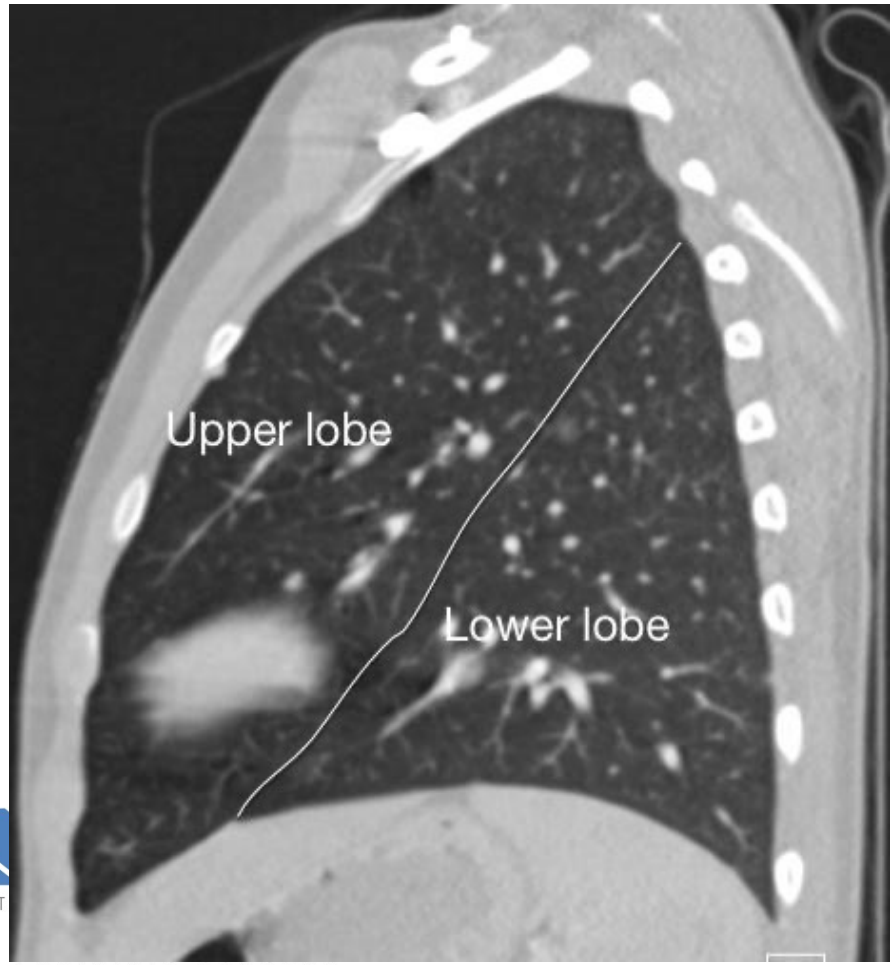
- **Anatomic Lobar Resection: Right Lung**

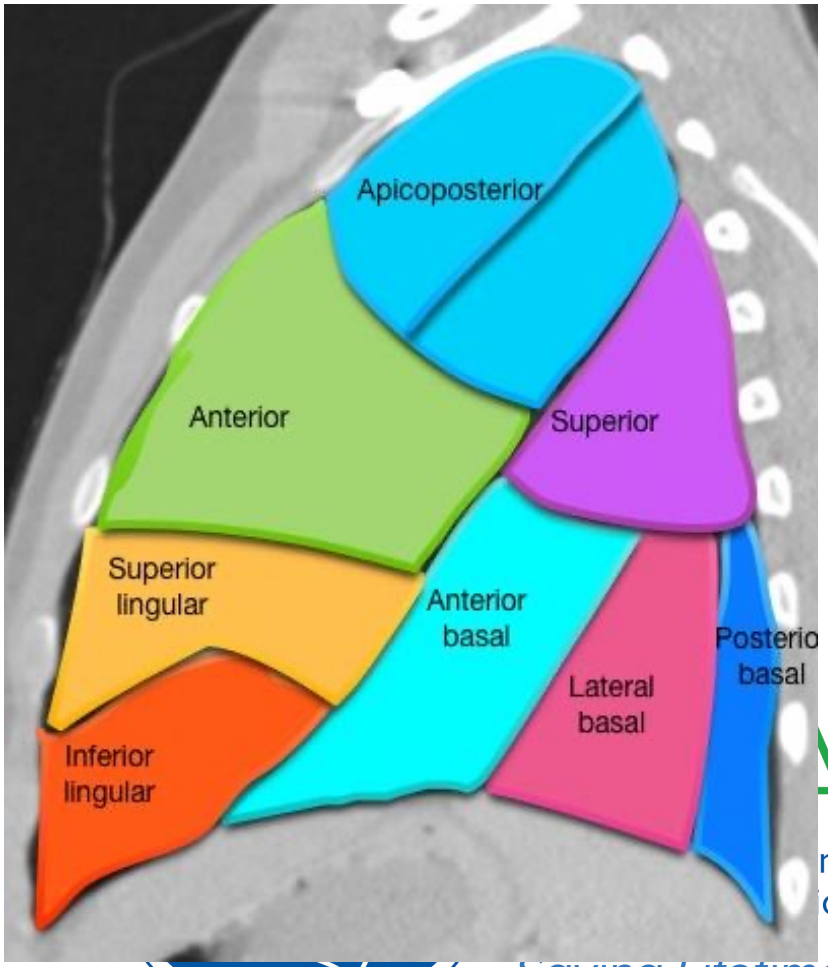




Operation

- **Anatomic Lobar Resection: Left Lung**





Operation

- **Lobar Resection:**
  - Open or Thoracoscopic
  - Inferior pulmonary ligament divided
    - Look for a systemic feeding vessel and ligate if present
  - Division of the fissure
  - Ligation of lobar artery, vein, and bronchus
  - Air leak test
  - Chest tube placed

Post-Operative Management

- **Chest tube**
  - Remove once output is low and air leak resolved

- **Pain management**
  - Regional (epidural, nerve block) more common for thoracotomy
  - NSAIDS, Tylenol, opioids as needed

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Complications

- Peri-operative
  - Bleeding
  - Need for postoperative respiratory support
  - Bronchial plug
  - Bronchial stump leak
- Long Term
  - Typically, no long term sequelae
  - If resection performed early, patients have compensatory lung growth

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Questions

- **Newborn with respiratory distress and cystic appearance to left lower lobe on chest X ray. Most likely diagnosis?**

  A. Bronchopulmonary sequestration
  B. Congenital pulmonary airway malformation (CPAM)
  C. Congenital lobar emphysema
  D. Pneumothorax

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Questions

- **3 month old with known CPAM presents to your office, asymptomatic. What is the best study to order to evaluate the lesion prior to discussing surgery?**

  A. Chest X-ray
  B. Ventilation/perfusion scan (V/Q scan)
  C. MRI of the chest
  D. CT of the chest with IV contrast

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Final Discussion/Review

- **Types of congenital lung lesions**
  - Congenital pulmonary airway malformation (CPAM)
  - Bronchopulmonary sequestration (BPS)
  - Congenital lobar emphysema
  - Bronchogenic cyst
- **Diagnosis**
  - Often, prenatally by ultrasound
  - Later in life, can present as frequent pneumonias or as an incidental finding

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Final Discussion/Review

- **Imaging modalities**
  - Chest X-ray
  - CT Chest with IV contrast (even if lesion not visible on CXR)
- **Management of CPAM**
  - Elective resection in first year of life if asymptomatic
    - Risk of recurrent pneumonia, pneumothorax, heart failure, or malignant degeneration
  - Watchful waiting

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Final Discussion/Review

- **Operative approach**
  - Open or thoracoscopic
  - Complete resection of the affected lobe
  - Typically performed within the first year of life to allow for compensatory lung growth

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Acknowledgement Slide

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