Bronchopulmonary Malformations

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Introduction

- Developmental anomalies of the bronchopulmonary unit account for 90% of lung lesions in children
- Common origin and overlapping features (spectrum). Differences may be due to variations in level of obstruction, severity, timing
  - Congenital pulmonary airway malformation (CPAM)
  - Bronchopulmonary sequestration (BPS)
  - Congenital lobar emphysema (CLE)
  - Bronchogenic cyst
  - Bronchial atresia (BA)
- Define anatomy clearly first before giving the lesion a name

Part - 1

CPAM

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- formerly called CCAM (*congenital cystic adenomatoid malformation*)
- The most common bronchopulmonary malformation (1.5 per 10,000 LB)
- There are 5 types but practically can be divided into two
  - macrocystic = ≥5mm
  - microcystic = solid mass
- All lesions require surgical resection

Part - 1

CPAM

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Pathogenesis & Classification

Presentation

Diagnosis

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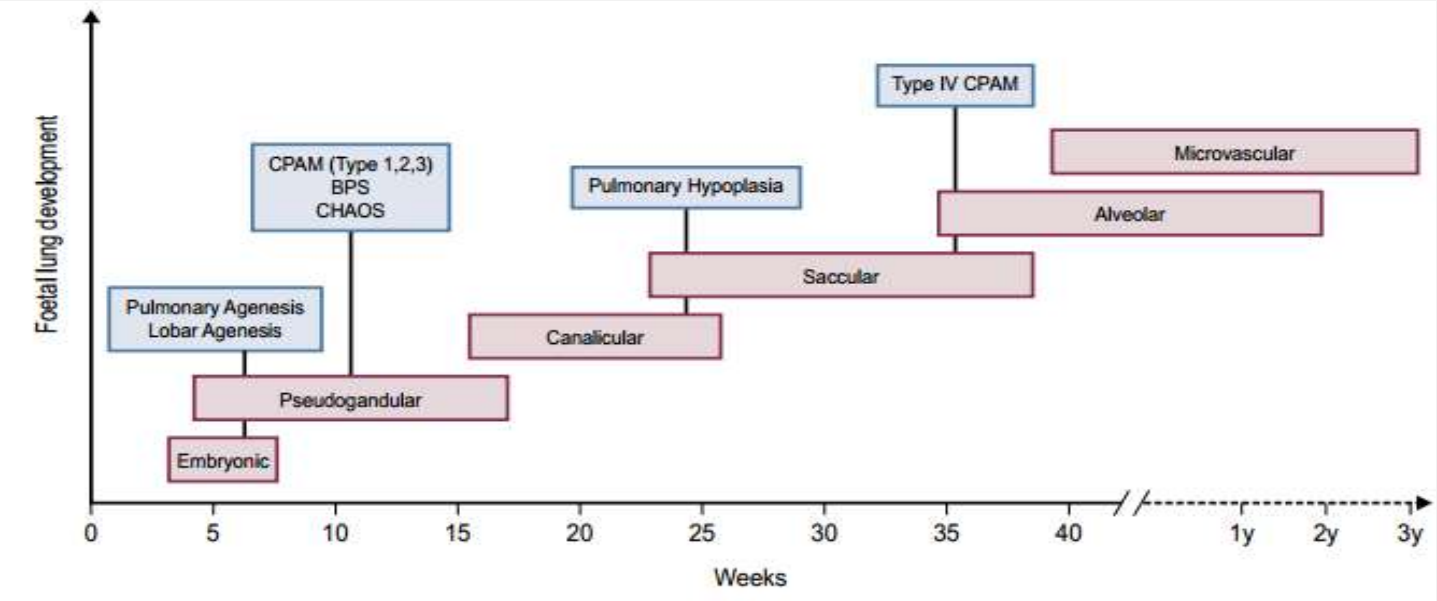
Pathology

- *Group of cystic and non-cystic lung masses*
  - Usually unilateral and limited to one lobe.
  - More common in lower lobes. Rt = Lt.
- cysts communicate with tracheobronchial tree but doesn't participate in gas exchange
  - May lead to air trapping
- Vascular supply & drainage from pulmonary vessels
- Cyst Lined with resp epithelium (polypoid proj), wall contain fibromuscular/elastic tissue
  - Absence of cartilage and inflamation
  - 1/3 of type 1 contain mucus producing cells (gastric mucins >>bronchoalveolar ca)

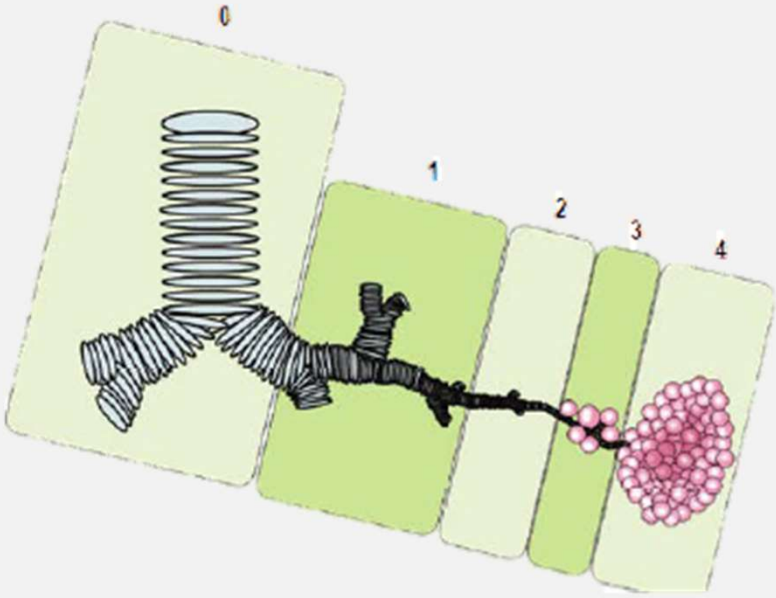
Etiology



- **Maturation defect** – Abnormal proliferation of immature segment of bronchial tree. Exact mechanism not known but thought to be imbalance b/n proliferation & apoptosis. Arise at different stages of lung development. Sporadic except type 4 (pleuropulmonary blastoma syndrome)

Classification

- **Type 0** (2%) – very small, diffuse (involve whole lung)
  - Incompatible with life (fatal at birth)
- **Type 1** (65%) – few, large cysts (>2cm)
  - ass. with bronchoalveolar ca (rare)
- **Type 2** (15-20%) – multiple, small cysts (0.5-2cm)
  - ass. anomalies (60%)- BPS (hybrid), CDH, TEF, CHD, renal agenesis
- **Type 3** (5-10%) – solid (large microcysts-<5mm cysts)
  - Poor prognosis
- **Type 4** (10%) – appear similar to type 1 (large cyst)
  - ass.. with pneumothorax and pleuropulmonary blastoma



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Part - 1

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Presentation

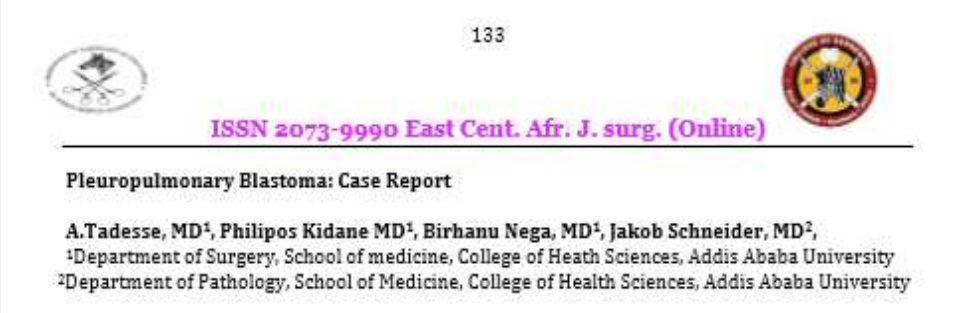
- Prenatal diagnosis
- resp distress (at birth in 2/3)
- chronic cough, dyspnea on exertion, or pleuritic chest pain
- Complications
  - recurrent infection
  - Pneumothorax
  - Malignant transformation (pleuropulmonary blastoma)

Recurrent infection

- infected parenchyma (pneumonia) or infected cyst
- Presentation in 1/3 of non-antenatally diagnosed infants
- difficult to clear with antibiotics

Pleuropulmonary blastoma

<500 cases worldwide (one report from Ethiopia)



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ISSN 2073-9990 East Cent. Afr. J. surg. (Online)

Pleuropulmonary Blastoma: Case Report

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- Embryonic pulmonary neoplasm (analogue to WT, hepatoblastoma…)
  - Been described as rhabdomyosarcoma but may contain other tissue elements
- Resembles non-malignant CPAM, can be solid/cystic (difficult to diagnose preop)
- Highly aggressive tumor

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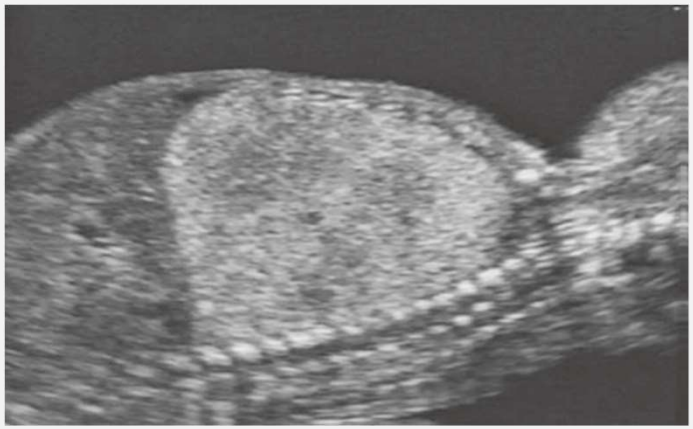
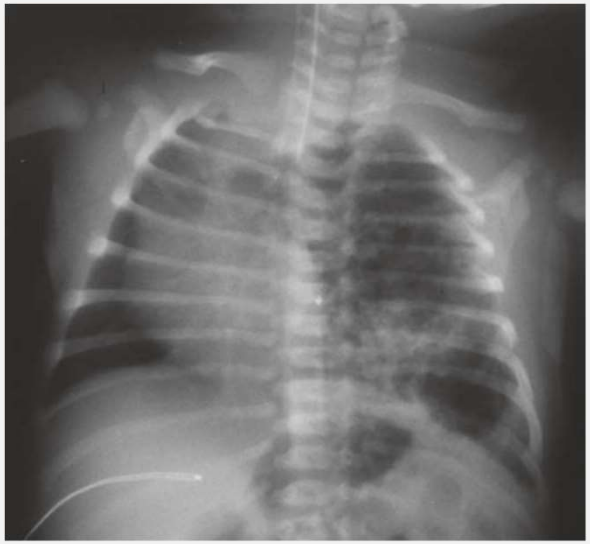
Prenatal ultrasound

Also predict prognosis

- Cyst type, size
- Mediastinal shift, Diaphragmatic eversion
- Polyhydramnios, Hydrops fetalis (40%)
- CVR (CPAM volume ratio)
    - CPAM/HC >1.6 indicate risk of hydrops
- Associated anomalies

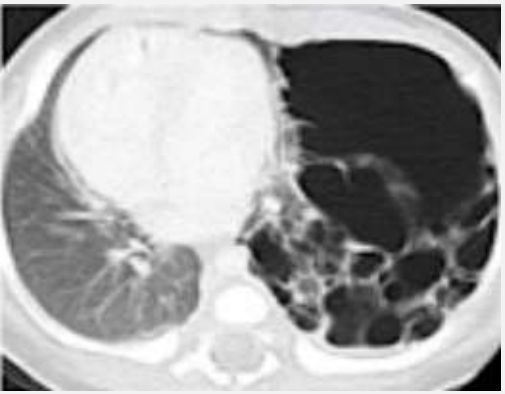
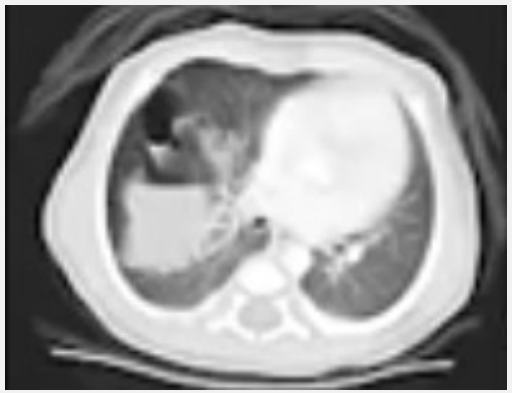
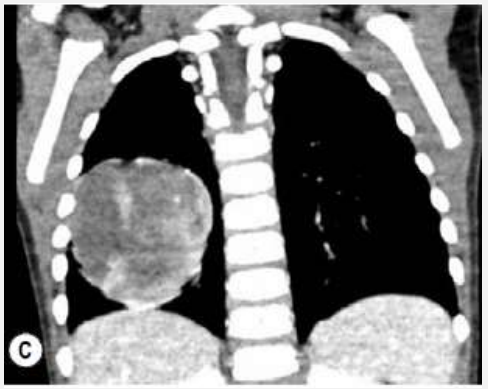


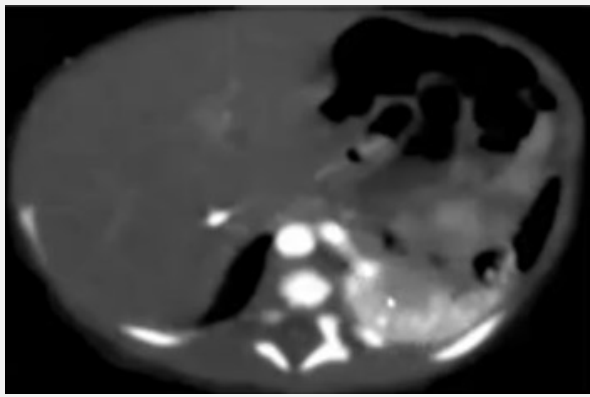
Chest X-ray



- Chest x-ray may be normal. CT scan is almost 100% sensitive in picking up the diagnosis. CXR ordered for other pathology may also detect incidental CPAM. CT is required nonetheless.

CT scan



- **Air filled cysts** (single large or multiple) **embedded in parenchyma.**
- **Sometimes filled with mucus**
- **Solid mass**
- **Hybrid lesion** (type II CPAM with systemic blood supply)

Differential diagnosis

- CDH - bowel peristalisis
  - *difficult to d/t large microcystic CPAM from in liver Rt side CDH
- BPS – no connection to bronchial tree, systemic blood supply
  - *some CPAMs might have systemic vessels (hybrid CPAM-BPS lesions)
- Foregut duplications (bronchogenic cyst)
- CLE

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CPAM

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Pathogenesis & Classification

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Part - 1

CPAM

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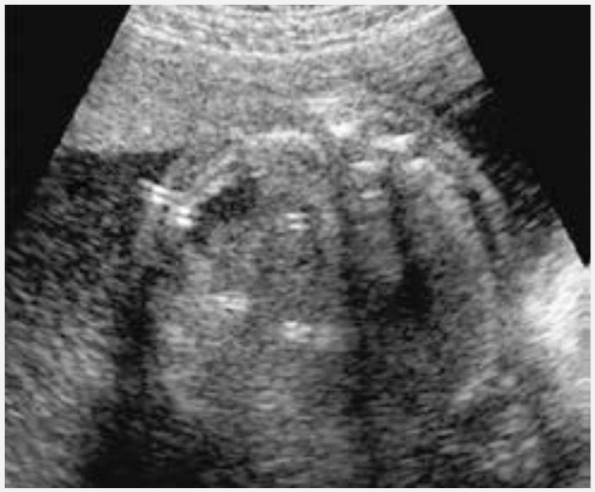
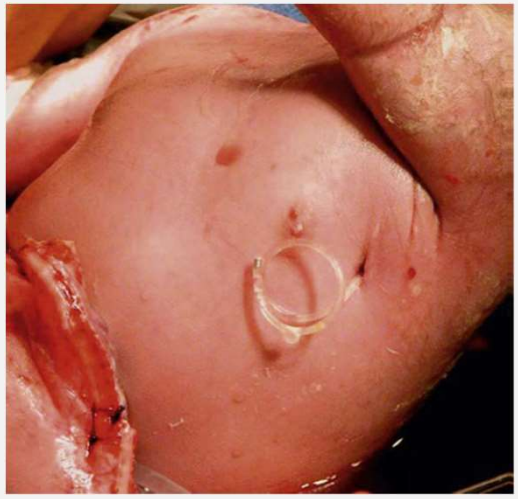
Natural history

- Grow with fetus until 28th wk after which size plateaus
  - No change in 50%
  - Enlarge in 10%
  - "disappearing CPAM" in 20% (*due to inc lung echongenicity in 3rd TM--always persist on postnatal imaging*)

Fetal intervention





Indicated if CVR>1.6 / hydrops before 32wk GA

- **Maternal IV betamethasone** (inhibit cyst growth, regress hydrops)
- **fetal surgery (lobectomy)** – microcystic, use declined after bethamethasonne
- **Thoracoamniotic shunting** (*pictures above) – macrocystic CPAM (less response to steroid)

Timing of surgery

- **Severe mediastinal shift/hydrops after 32 wk:** delivery by EXIT and CPAM resection under placental support
- **Mild-moderate mediastinal shift, symptomatic:** immediate surgery (within few days of delivery/diagnosis)
  - *emergency thoracotomy-lung decompression in extreme cases (followed by lobectomy)
- **Regressing CPAM:** postnatal CT at 4-6wk, elective surgery at 2-3mo
  - *asymptomatic lesions need resection – can't predict malignancy or d/t by imaging
  - *Controversy if don't completely regress despite being undetectable on X-ray

Type of surgery

- **Lobectomy :** preferred (ascertain margin, less bleeding and air leak)
- **Segmentectomy:** consider if multiple lobes involved (1-2%)
  - *if bilateral most involved side should be resected first
- **Non anatomic (wedge resection):** if no clear plane (segments of both upper and lower involved)

Part - 2

BPS

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- Mass of abnormal pulmonary tissue without bronchial communication
- Commonly left side (Intralobar- LLL, Extralobar – b/n LLL & diaphragm)
- Arterial supply from systemic vessels (thoracic/abdominal aorta)
- Venous drainage variable (pulmonary- iBPS, systemic- eBPS)

Part - 2

BPS

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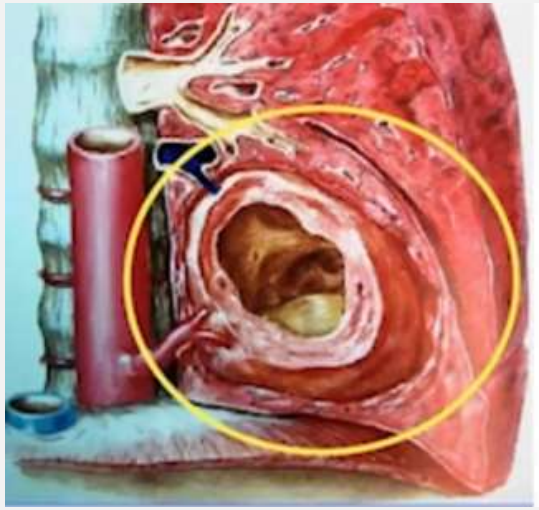
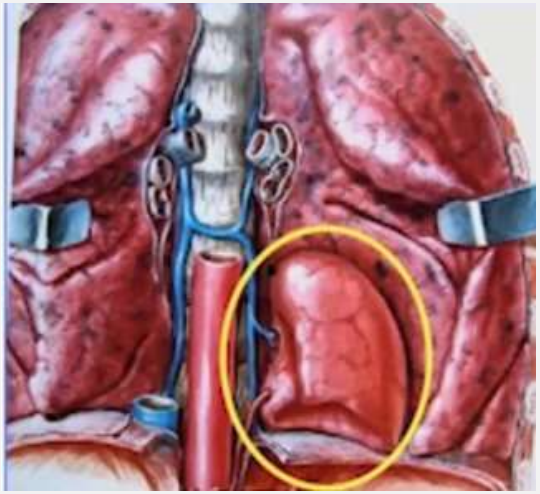
Pathology

- Abnormal budding in embryogenesis
  - iBPS - before pleura develops
  - eBPS - After pleura
- Arterial supply from systemic vessels
  - thoracic/abdominal aorta (20% infra diaphragmatic)
  - single/multiple (30-40% are multiple)
- Venous drainage variable
  - iBPS – pulmonar
  - eBPS – systemic (SVC, azygus)

Types





- **Intralobar (75%) IBPS** within visceral pleura. May establish 2ry connection to airway.
- **Extralobar (25%) eBPS** "Accessory lung" in its own pleura. May have associated anomalies
- **Hybrid lesions:** 50% of extralobar and 10% of intrlobar have associated CPAM

Part - 2

BPS

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Part - 2

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Presentation

- iBPS – present later with recurrent infection (communication with airway)
  - Hemoptysis
- eBPS – present earlier with resp distress.
  - Older infant may have heart failure (excessive flow through feeding vessel)
  - Associated anomalies (40%)
    - CDH (5% have eBPS)
    - CCAM (50%)
    - Vertebral, cardiac
    - foregut malformations, colonic duplication

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Part - 2

BPS

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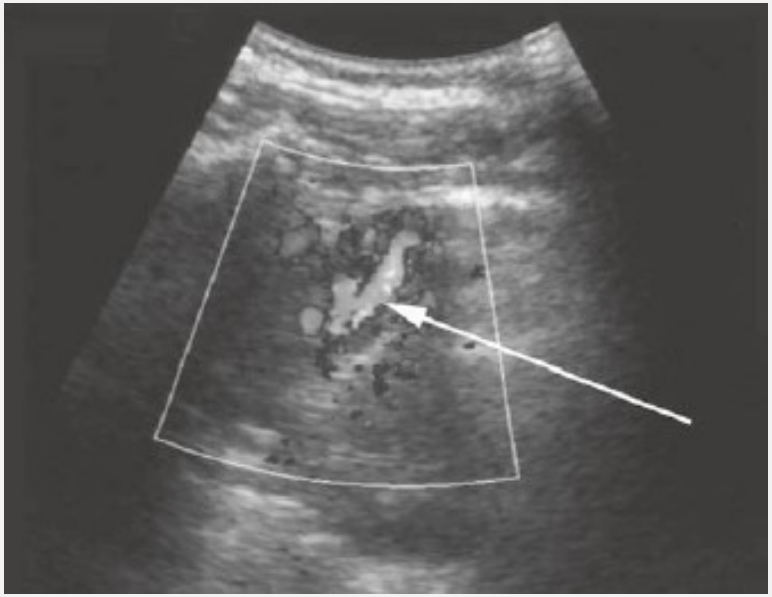
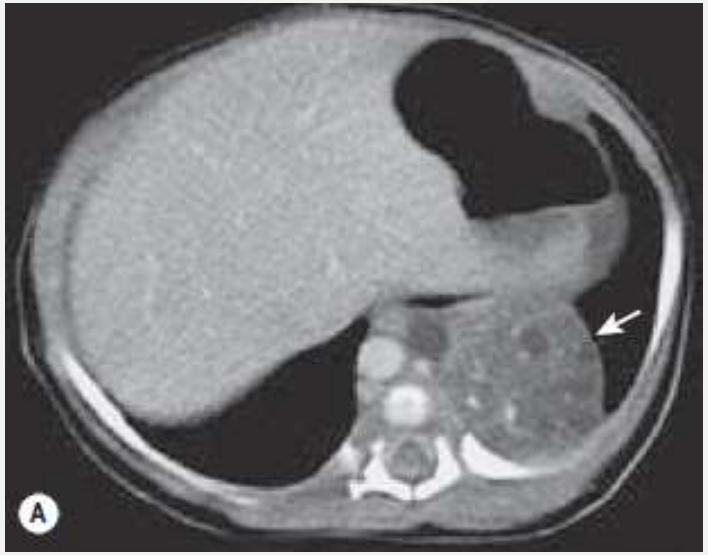
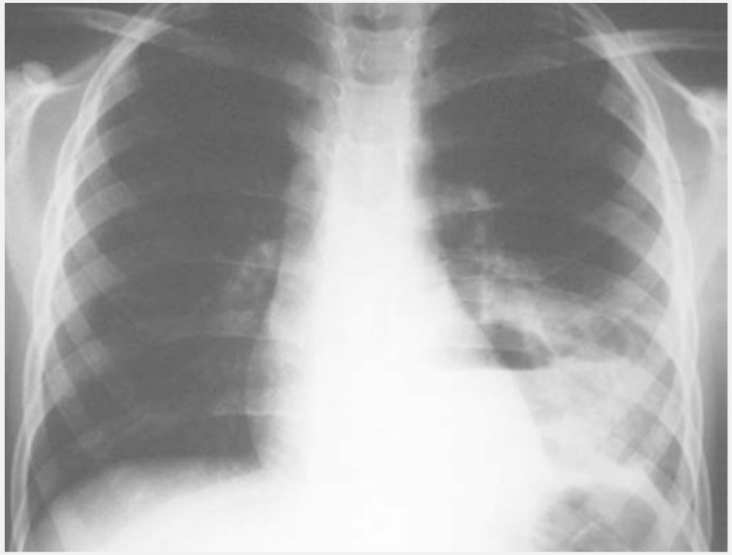
Prenatal US



- echodense homogenous mass separate from lung. Systemic artery identified with doppler.

Postnatal Imaging





- **eBPS:** seen as extralobar homogenous hyperechoic mass in paraspinal location
- **iBPS:** may show air fluid level on CXR due to bronchial communication. CT will show abnormal parencyma

Part - 2

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Part - 2

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Prenatal management

- iBPS – don't usually manifest

- eBPS – large mass, can have mediastinal shift, effusion, hydrops

  (may need TA shunting or laser ablation of feeding vessels)

Postnatal management

- Surgery:
  - All iBPS – risk of infection, high output physiology
  - Cystic areas on imaging – hybrid CPAM elements
  - Large lesion - lung compression, mediastinal shift
  - Large feeding vessel – less likely to regress
  - Location within or under diaphragm
- Controversy: small intrathoracic or subdiaphragmatic eBPS with small feeding vessel

Type of surgery

- **eBPS** - require ligation of systemic vessel and removal of mass
  - Thoracotomy (intrathoracic)
  - Laparatomy (retroperitoneal eBPS- commonly in lesser sac)
- **iBPS** - ligation of vessel and lobectomy

Part - 3

CLE

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- AKA congenital alveolar hyperinflation (no emphysema on histology)
- BPM characterized by air trapping with preserved alveolar anatomy
- prevalence of 1 in 20,000 -30,000 (Rare)
- Male predominance (3x) for unknown reason
- Has option of conservative treatment unlike other BPMs (not associated with risk of infection or malignancy)

Part - 3

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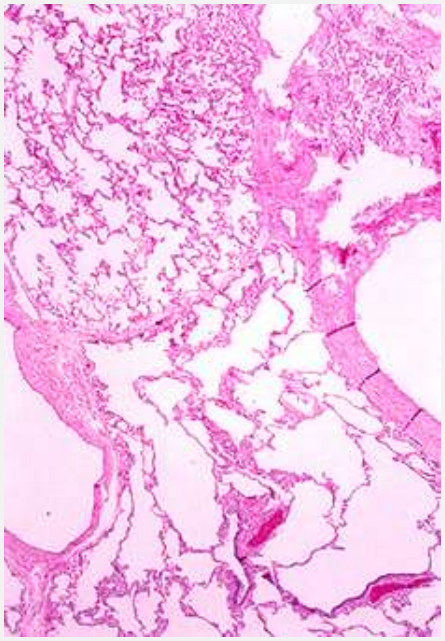
Pathogenesis

- amniotic fluid trapping lead to lobar expansion
- Left upper lobe (40-50%) >> right middle (25%) >> right upper (20%) >> lower lobes (2-10%)
- rare bilateral or multifocal involvement
- variable degrees of compression of the adjacent lung
- The lesion does not change during breathing but has a reduced blood supply

Pathology



- **gross inspection** of CLE of the LUL demonstrates hyperexpansion and pallor. Lingula is unaffected and looks dark without expansion. **Histology** of CLE demonstrates uniformly enlarged distal airways and alveoli. Their walls are normal

Etiology

? abnormal interactions between embryonic endodermal and mesodermal components of the lung

- unknown in 50%
- Bronchial Obstruction with "ball valve" mechanism (25%)
  - **intramural** - dysplastic bronchial cartilage/bronchial atresia, infection
  - **Endobronchial** - meconium or mucus plugs, mucosal fold
  - **extrinsic** – PA sling (anomalus origin of LPA), intrathoracic masses

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Clinical features

- generally not associated with significant prenatal pathophysiology
  - Moderate mediastinal shift
  - *very limited may require fetal intervention (??resection +/- EXIT)
- Almost all present in first 6 months of life
  - 25% at birth, 50% in neonatal age.
- severity depends on size of lobe, compression of lung & mediastinum
  - respiratory distress (rapid or insidious)
  - Recurrent pneumonia, poor feeding with failure to thrive
  - wheezing, dec breath sounds, hyperresonant

Associated anomalies

- cardiac in 14%
- Other possible anomalies
  - Renal
  - pectus excavatum
  - hiatal hernia

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Part - 3

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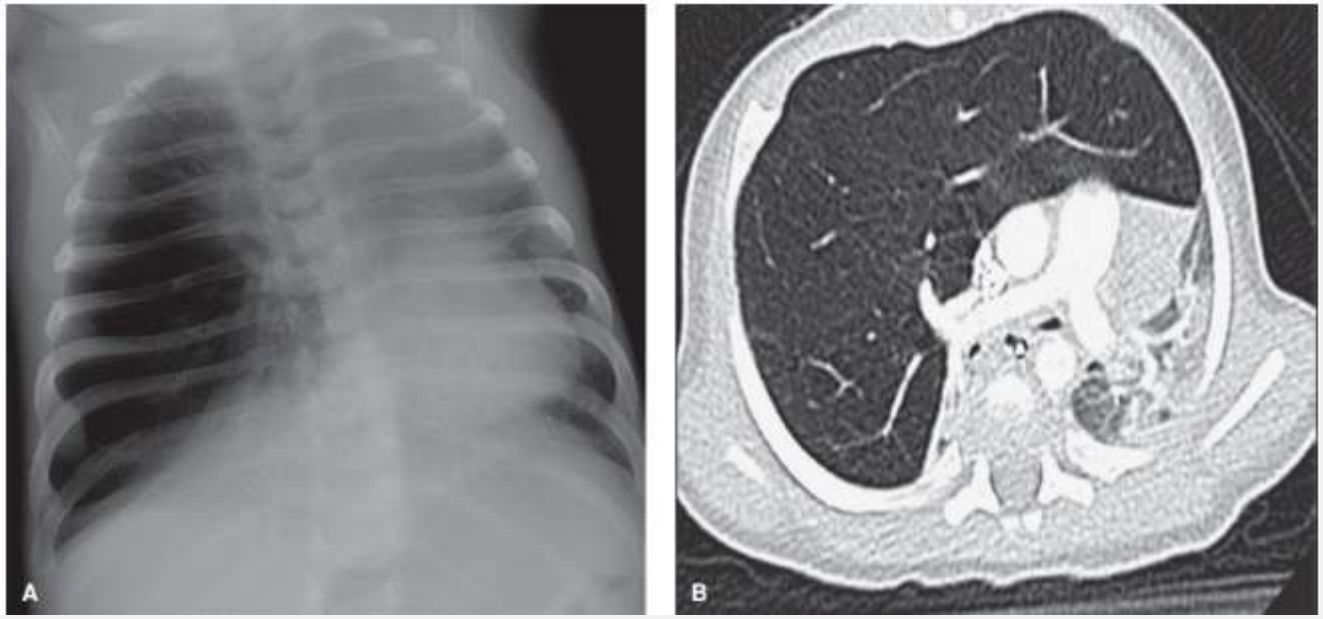
Imaging

- **Prenatal ultrasound** - increased echogenicity (cystic)
  - Decrease in size during pregnancy and become apparent again postnatal
- **X-ray** - hyper-lucent lobe, flattened diaphragm, mediastinal shift, contralateral atelectasis
  - immediately after birth, the affected lobe may appear opacified due to retained fetal lung fluid
- **CT scan** – in non diagnostic chest radiograph
  - may also demonstrate an intrinsic or extrinsic source of airway obstruction
- **Echocardiography** - vascular structures causing airway compression

Imaging



- X-ray showing marked hyperinflation of RML with expanded intercostal space, depression of diaphragm, mediastinal shift and contralateral atelectasis

Differential diagnosis

- Localized pulmonary interstitial emphysema/pneumothorax (MV)
- Foreign body
- Other bronchopulmonary malformations
- CDH
- Pulmonary agenesis (contralateral hyperinflation)
- Unilateral hyperlucent lung syndrome (post bronchiolitis)
- Poland syndrome

Part - 3

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Part - 3

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Decision and timing

- **respiratory distress** - urgent/emergent open lobectomy (MV worsen condition)
- **minimal symptoms–** elective lobectomy (don't require O2, good growth)
  - *controversial, but recommend <1 yr based on postop PFT (< 1yr = alveolar multiplication of remaining lung, > 1yr = overinflation of contralateral lung)*
  - **? If a distended aberrant pulmonary artery is the cause, repair of the vascular anomaly is the treatment and lobectomy could be sometime avoided
- **Asymptomatic/mild symptoms** - may not need surgery (stabilize / regress)

Part - 4

Bronchogenic cyst

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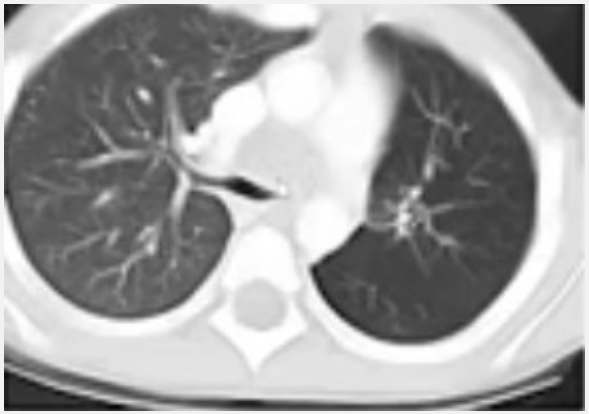
- Most common mediastinal cyst
- Duplication- can occur anywhere but usually found in close to carina
- Thin, well defined wall with homogenous fluid inside (mucus)



- All cysts should be excised. Plus lobectomy if bronchial obstruction-- irreversible damage

Part - 5

BA

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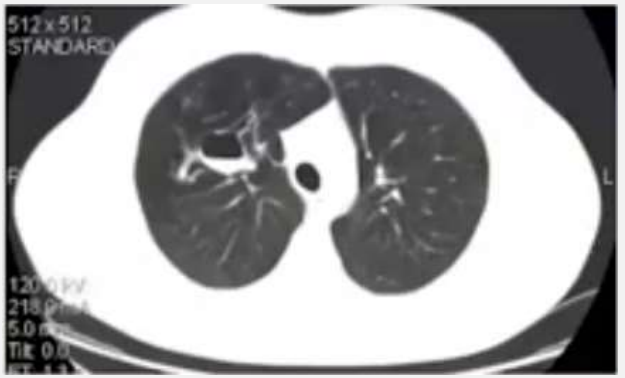
- Segmental bronchus commonly affected
  - Lobar enlargement (hyperplasia, fluid)
  - Distal bronchus dilated (mucocele)
- May be small, not affecting surrounding (only small hypoattenuation of lung)



- Can be large microcystic lesion (similar to CPAM, but don't respond to steroids)
- Can be isolated, ass. with intralobar sequestration or communicate with GIT

Thank You