MAX Healthcare

CONGENITAL THORACIC MALFORMATIONS

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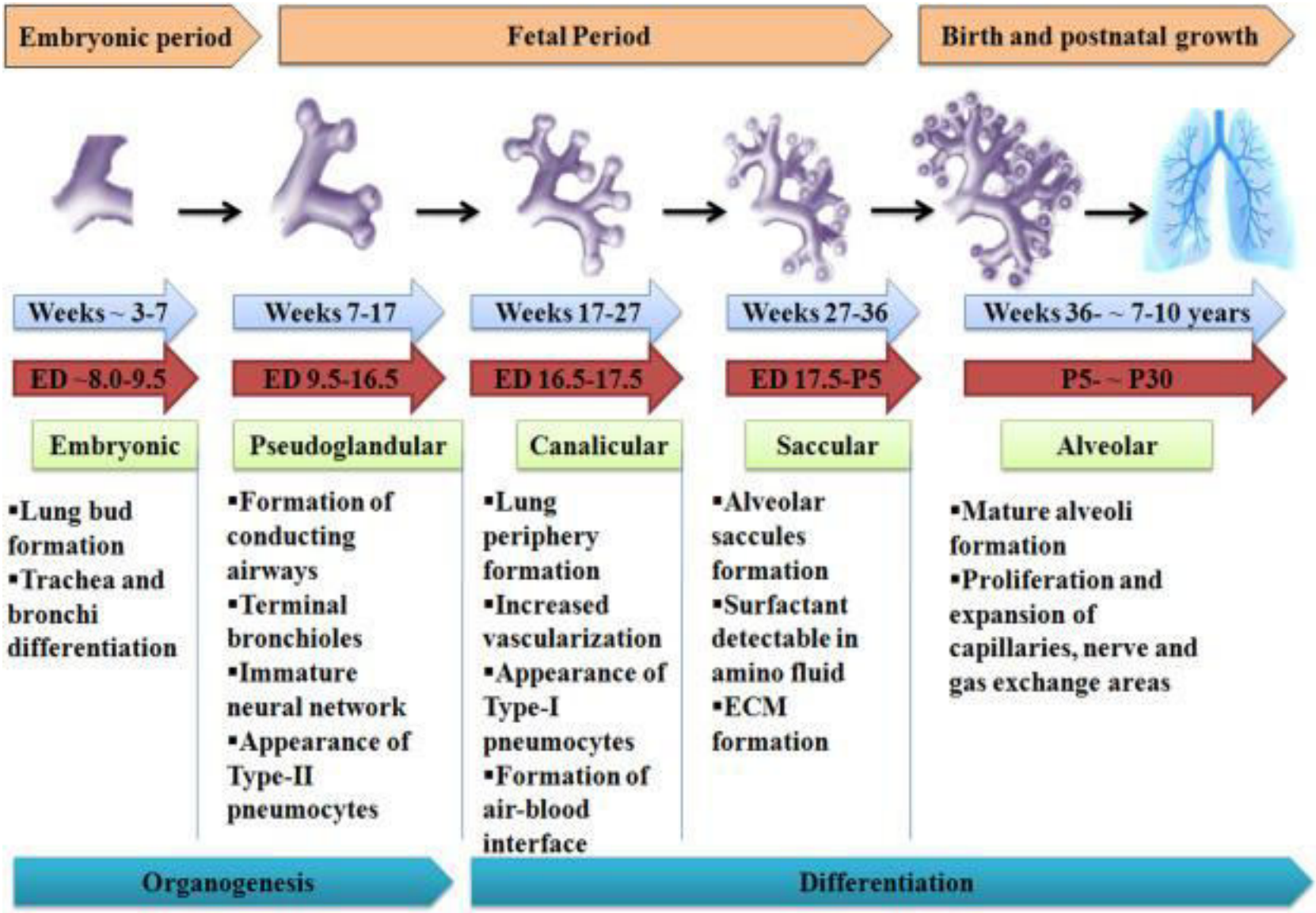
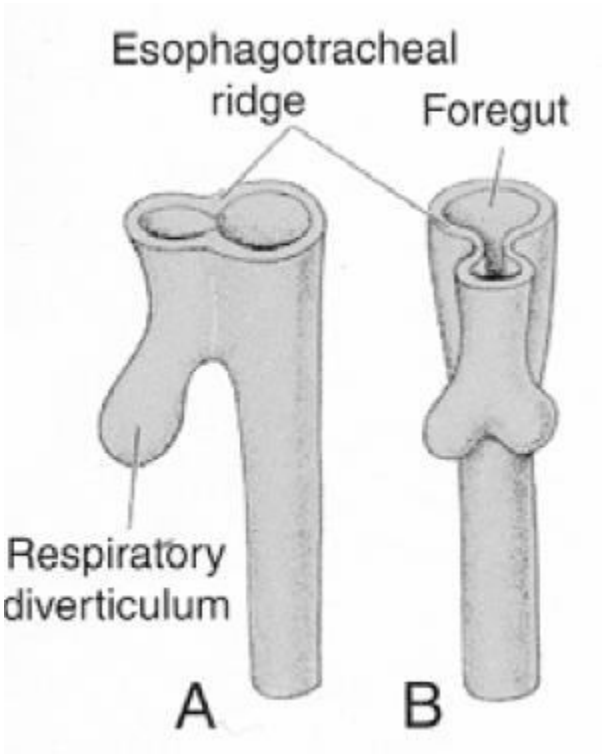
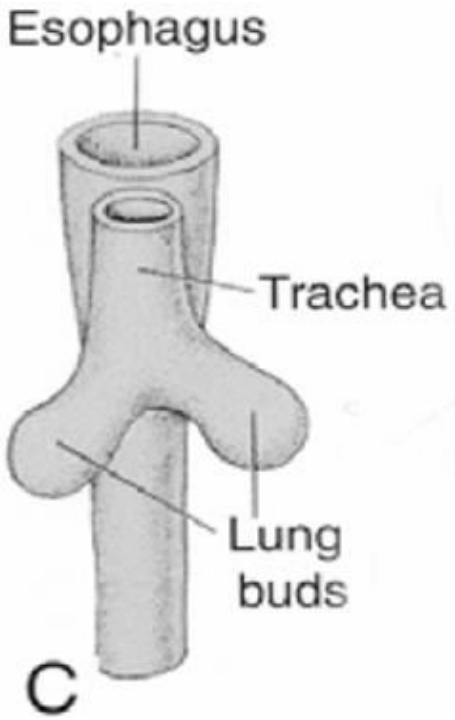
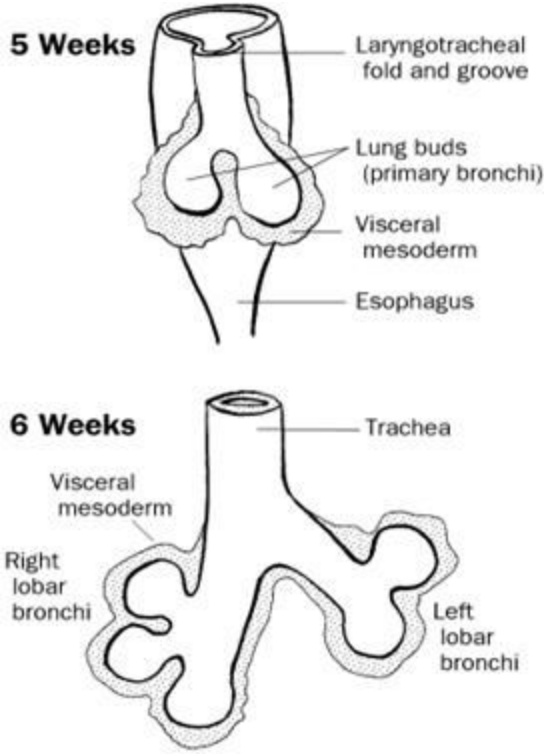

Embryonic period
Fetal Period
Birth and postnatal growth
Weeks ~ 3-7
Weeks 7-17
Weeks 17-27
Weeks 27-36
Weeks 36- ~ 7-10 years
ED ~8.0-9.5
ED 9.5-16.5
ED 16.5-17.5
ED 17.5-P5
P5- ~ P30
Embryonic
Pseudoglandular
Canalicular
Saccular
Alveolar
▪Lung bud formation
▪Trachea and bronchi differentiation
▪Formation of conducting airways
▪Terminal bronchioles
▪Immature neural network
▪Appearance of Type-II pneumocytes
▪Lung periphery formation
▪Increased vascularization
▪Appearance of Type-I pneumocytes
▪Formation of air-blood interface
▪Alveolar saccules formation
▪Surfactant detectable in amino fluid
▪ECM formation
▪Mature alveoli formation
▪Proliferation and expansion of capillaries, nerve and gas exchange areas
Organogenesis
Differentiation

Embryonic stage







Embryonic Stage - Anomalies

Error in separation of laryngo-tracheal groove

- TEF, tracheal atresia, tracheal stenosis

Failure of partitioning of lung bud

- Pulmonary agenesis (u/l, b/l)

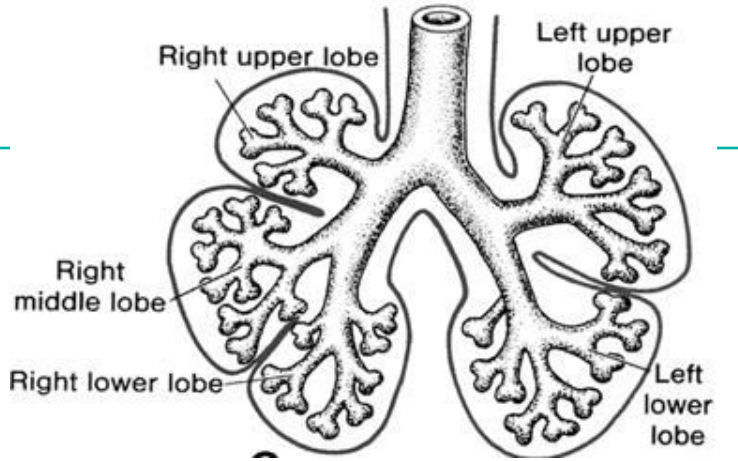
Formation of Accessory lung bud

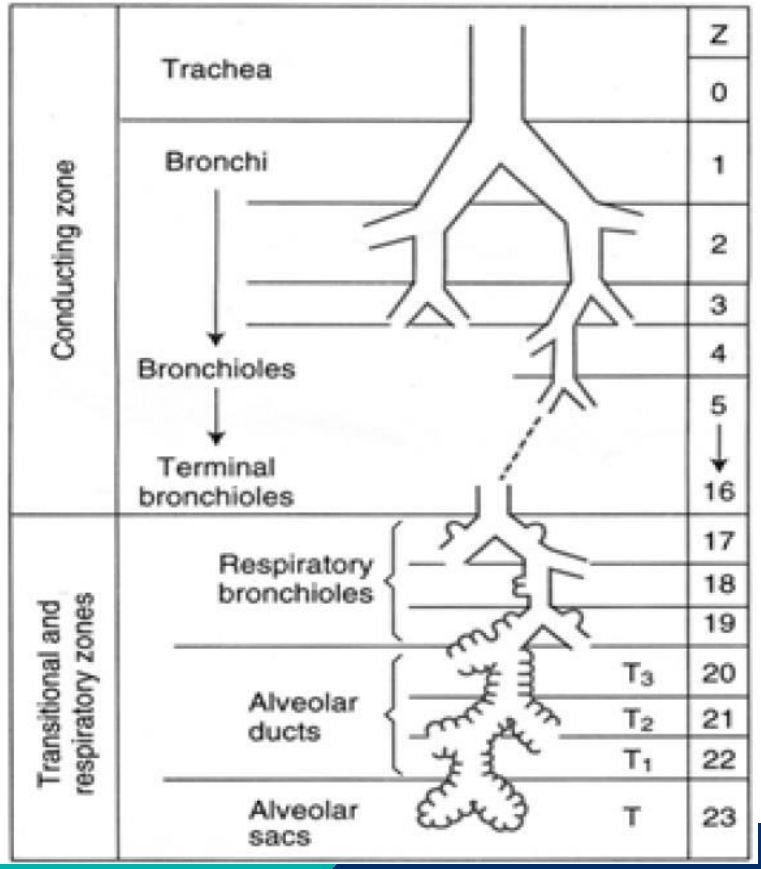
- **Pulmonary Sequestration**

***Foregut duplication cyst- abnormal budding from foregut**

Pseudoglandular stage

- **7-16 wk**
- Development of **conducting airways**
- By 7 wk - trachea, segmental & subsegmental bronchi evident
- **All bronchial divisions complete by 16 wk**
- Closure of pleuroperitoneal fold @ 7 wk





Pseudoglandular stage - Anomalies

No closure of pleuro-peritoneal fold

- CDH, Lung hypoplasia

Failure of partitioning

- Lobar agenesis (u/l, b/l)

Abnormal airway branching

- **CPAM**

Antenatal diagnosis

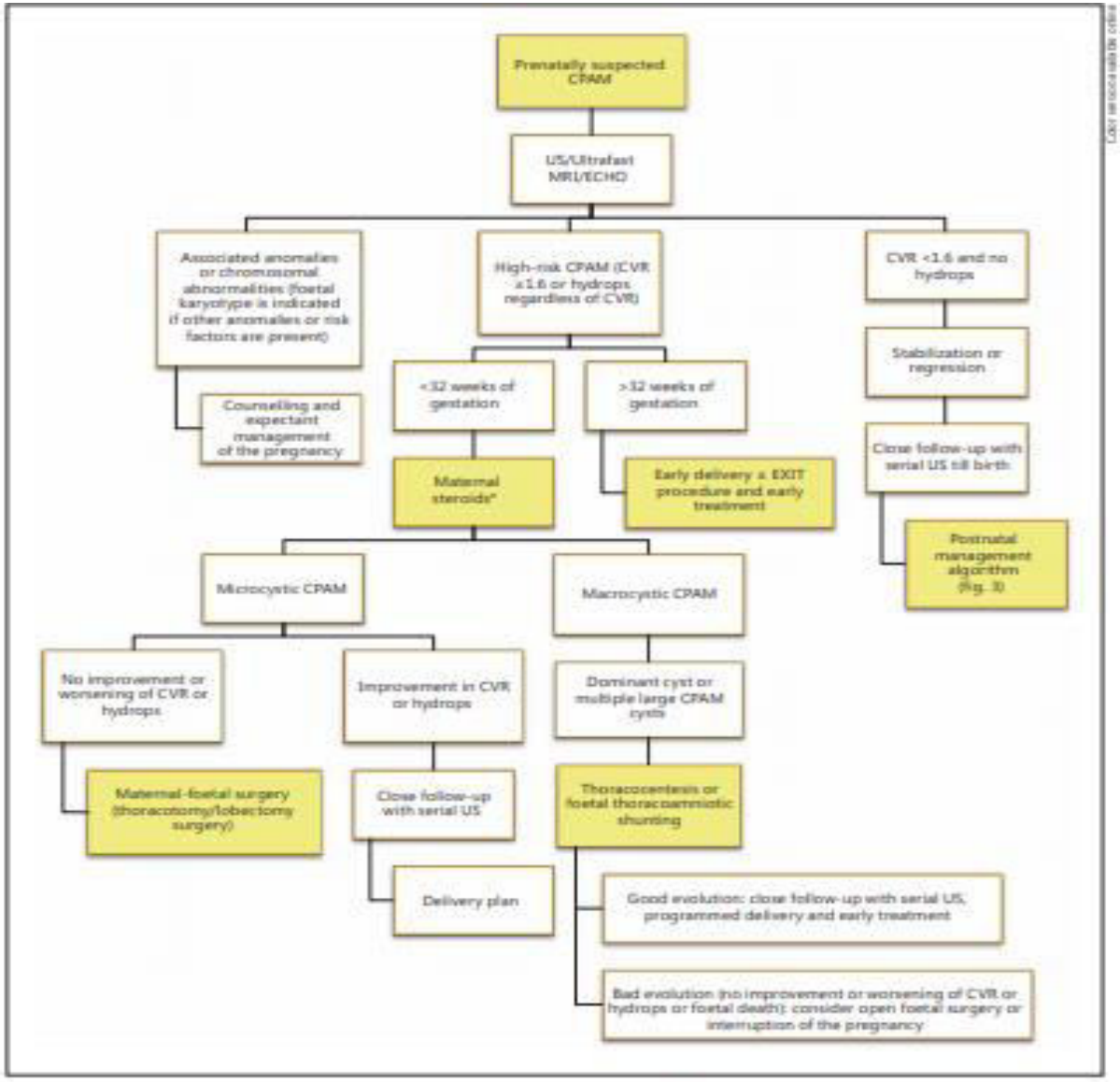
- USG- reliable and accurate in 2nd trimester
- Echogenic lung mass
- Difficult to differentiate various cystic entities
- Postnatal confirmation is required
- **Adzick** - classified antenatal cystic lesions as macrocystic (>5mm) and microcystic (<5mm) , of prognostic value
- Microcystic have poor prognosis

Clinical presentation

- Some with respiratory distress at birth
- Symptoms depend on size of lesion
- Most remain asymptomatic until complicated by secondary infection or compression of nearby structures

Antenatal Management

- Depends on symptoms- fetal hydrops
- Cystic lesion - Aspiration
- Large cyst with pleural effusion- Thoracoamniotic shunting
- Solid lesion – fetal lobectomy
- Ex utero intrapartum treatment (EXIT) procedure - resection of the abnormal lobe while on placental bypass
- Antenatal steroids- 1st line for high risk CPAMs

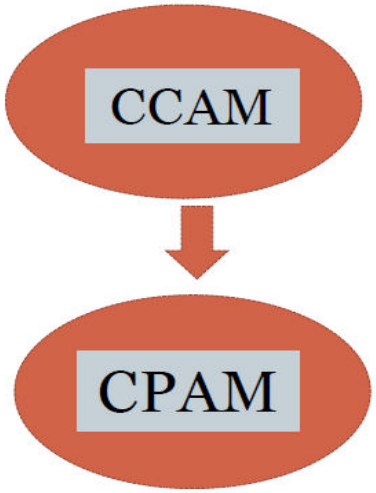

Prenatally suspected CPAM
US/Ultrafast MRI/ECHO
Associated anomalies or chromosomal abnormalities (foetal karyotype is indicated if other anomalies or risk factors are present)
Counselling and expectant management of the pregnancy
High-risk CPAM (CVR ≥1.6 or hydrops regardless of CVR)
<32 weeks of gestation
Maternal steroids*
>32 weeks of gestation
Early delivery ± EXIT procedure and early treatment
CVR <1.6 and no hydrops
Stabilization or regression
Close follow-up with serial US till birth
Postnatal management algorithm (fig. 3)
Microcystic CPAM
No improvement or worsening of CVR or hydrops
Maternal-foetal surgery (thoracotomy/lobectomy surgery)
Improvement in CVR or hydrops
Close follow-up with serial US
Delivery plan
Macrocystic CPAM
Dominant cyst or multiple large CPAM cysts
Thoracocentesis or foetal thoracoamniotic shunting
Good evolution: close follow-up with serial US, programmed delivery and early treatment
Bad evolution (no improvement or worsening of CVR or hydrops or foetal death): consider open foetal surgery or interruption of the pregnancy

Postnatal Management

- Symptomatic- immediate surgery
- Asymptomatic- controversial
- Surgical resection by 1 year of age is advised
  - To limit malignant potential (Sarcomatous and carcinomatous degeneration).
  - To rule out **pleuropulmonary blastoma** (radiographically similar to type 1 and 4 CPAM)
  - Recurrent infection, pneumothorax
- The mortality rate is <10%.

Congenital pulmonary airway malformation (CPAM)

- Multiple different hamartomatous lesions arising from abnormal branching of the immature bronchial tree, generally confined to 1 of lower lobes, Male preponderance.
- Incidence: 1 in 11000 to 35000 births



Because only 3 out of 5 are cystic

AND

Only 1 is adenomatoid

Congenital pulmonary airway malformation

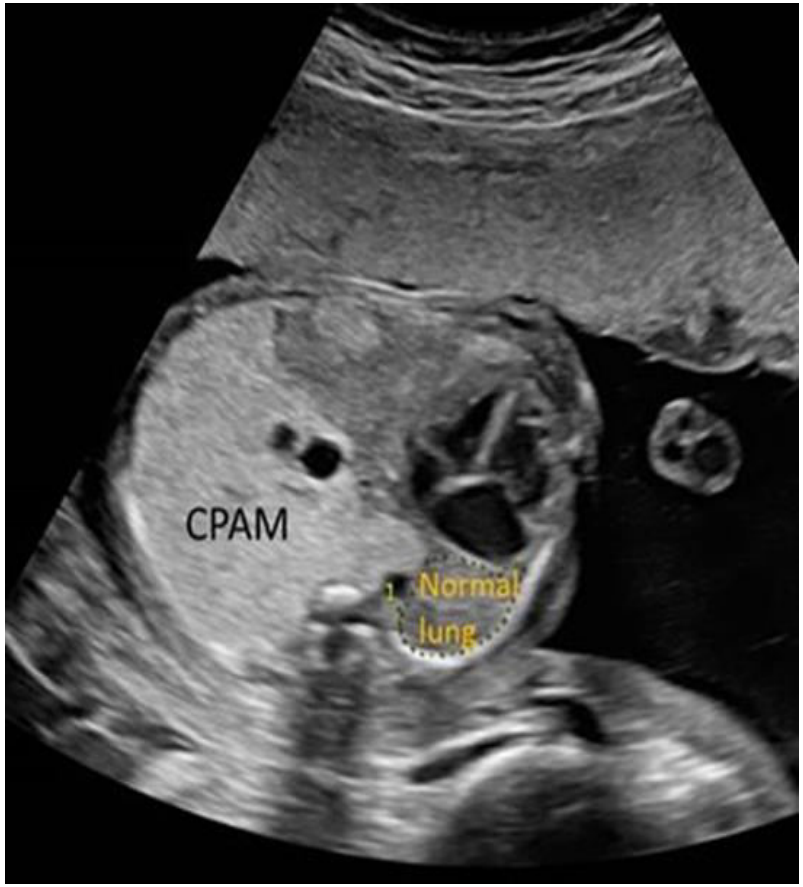
- Etiology: Embryologic injury during pseudoglandular stage.
- Connected to tracheobronchial tree and has pulmonary blood supply.
- Stocker's classification according to cyst size and histologic resemblance to segments of the developing bronchial tree and airspaces

CPAM Type	Type 0	Type I	Type 2	Type 3	Type 4
CCAM Type		Type I	Type II	Type III	
Developmental origin	Tracheal/ bronchial	Bronchial/ Bronchiolar	Bronchiolar	Bronchiolar/ alveolar	Acinar
Proportion of CPAM	<2%	60%–65%	15%–20%	5%–10%	10%
Timing of presentation	Birth	Prenatal (especially if large cysts)	Postnatally	Prenatal (if large)	Postnatal
Clinical presentation	Lethal pulmonary hypoplasia, absence or lack of alveoli	Asymptomatic, or immediate or delayed respiratory distress or infection	Respiratory presentation often less significant than associated anomalies	Prenatal (development of hydrops) Postnatal (depending on size) respiratory distress	Incidental finding, pneumonia or pneumothorax
Cyst size	None	0.5–10 cm	0.5–2 cm	Microcystic (solid)	Large multilocular
Lung involvement	All lobes involved (incompatible with life)	Lobar	Lobar	Lobar or entire lung	Lobar
Associated anomalies	CV anomalies, renal hypoplasia		50% associated anomalies (CV, CDH, BPS, renal dysplasia agenesis)	None	Pleuropulmonary blastoma
Malignancy risk	None	Bronchoalveolar carcinoma (rare)	None	None	Pleuropulmonary blastoma

Antenatal diagnosis

Ultrasonogram

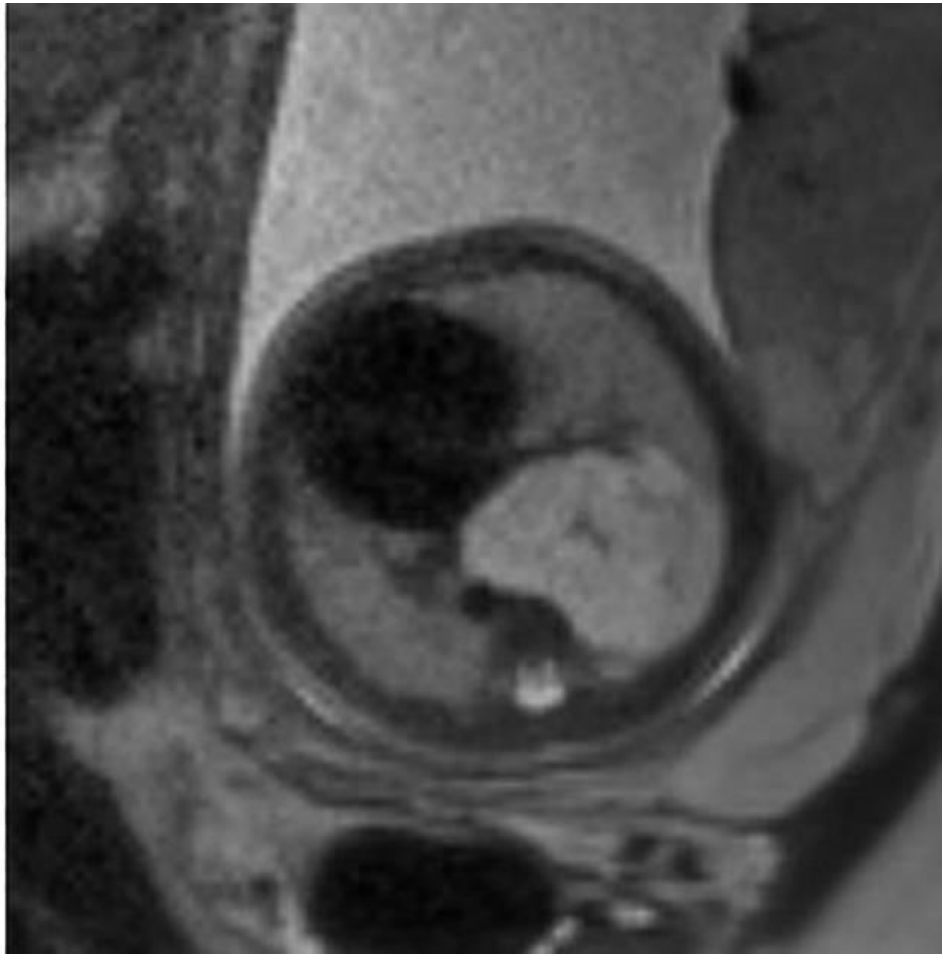
- Hyperechoic, heterogeneous tissue with multiple hypoechoic cysts
- CCAM volume ratio (CVR) - prognostication tool
- CVR >1.6 is predictor of hydrops and indication for intrauterine interventions



Predictors of hydrops

- CVR> 1.6
- Mass to thorax ratio >56%
- Cystic predominance
- Diaphragm eversion

Fetal MRI- CPAM



Scan shows an enlarged and T2 hyperintense right lung

Antenatal diagnosis

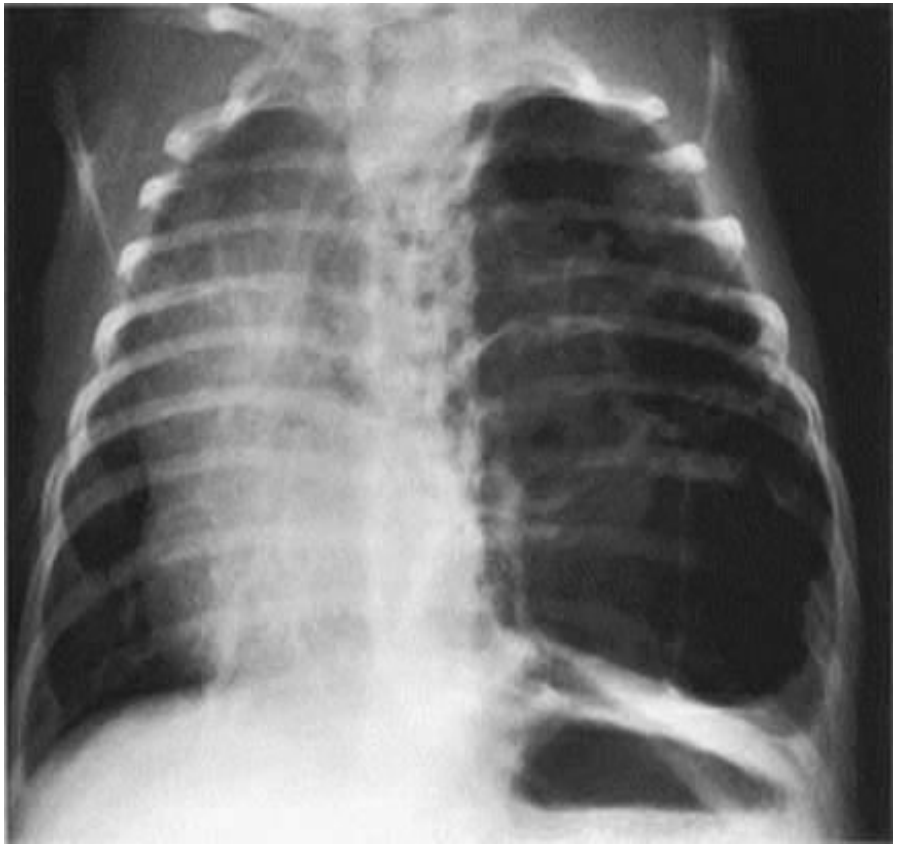
Colour Doppler US

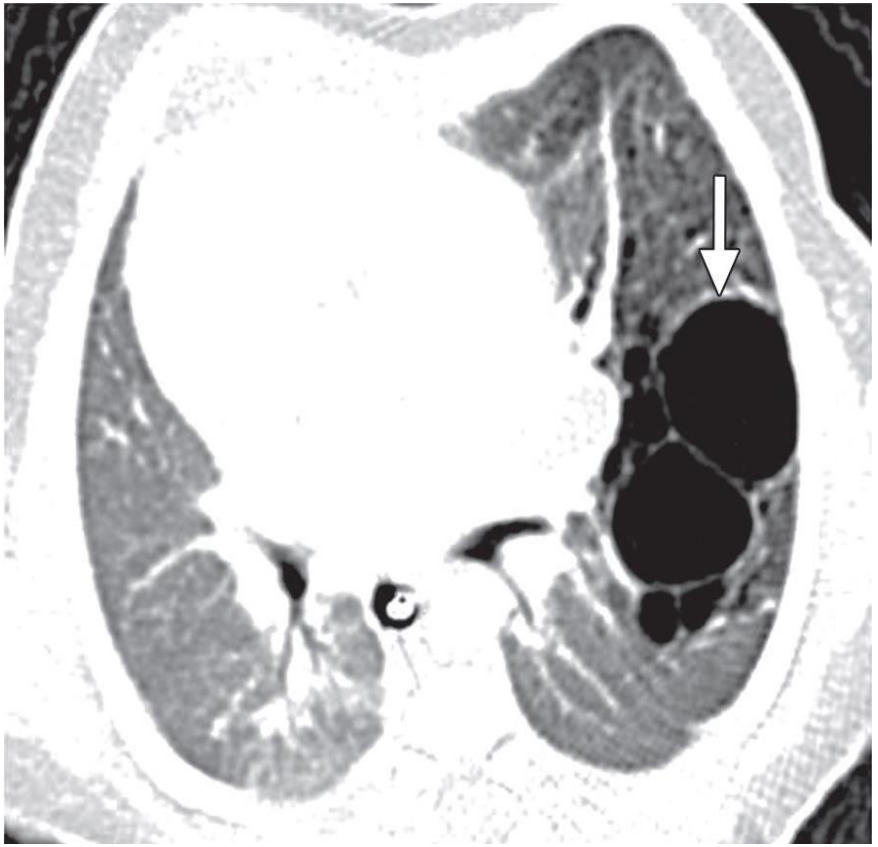
- Evaluates the arterial and venous blood flows, allowing prenatal differentiation between CPAM and BPS.
- While CPAMs derive their blood supply from the pulmonary circulation and drain via the pulmonary veins, BPS has a feeding systemic artery.

Poor prognostic factors - antenatally

- Microcystic lesions
- Fetal hydrops
- CVR >1.6
- Pulmonary hypoplasia
- Presence of other congenital anomalies

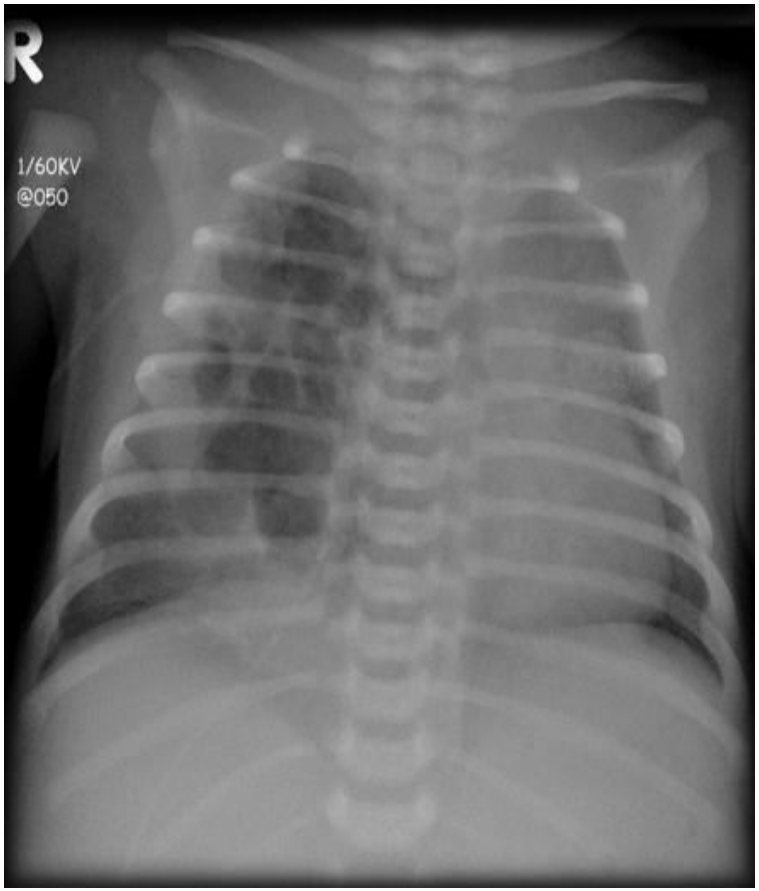
Postnatal diagnosis (Type I)

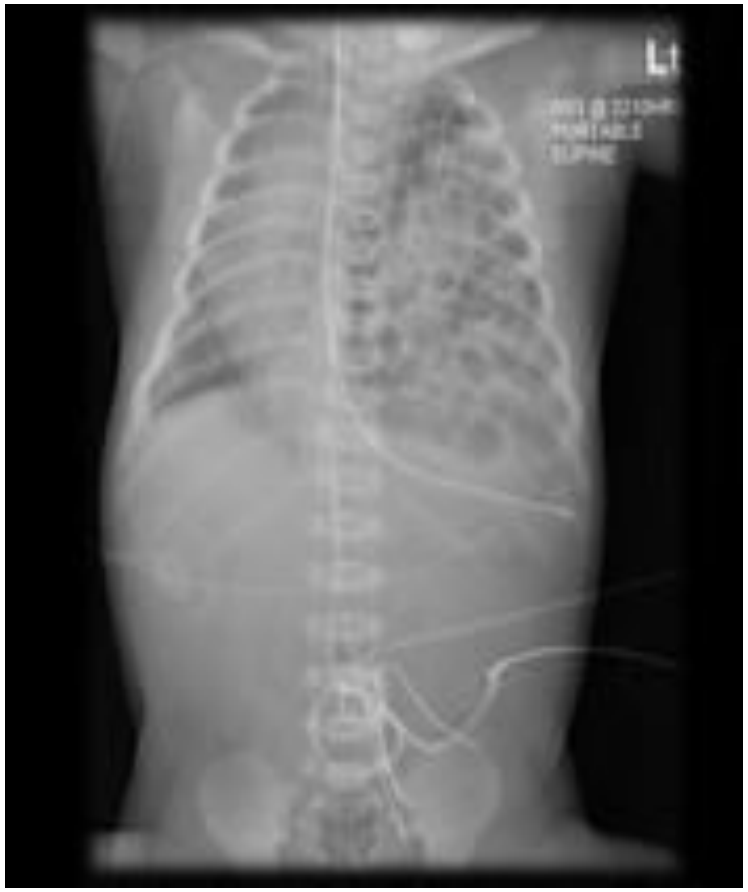




large multicystic mass in the left hemithorax with mediastinal shift

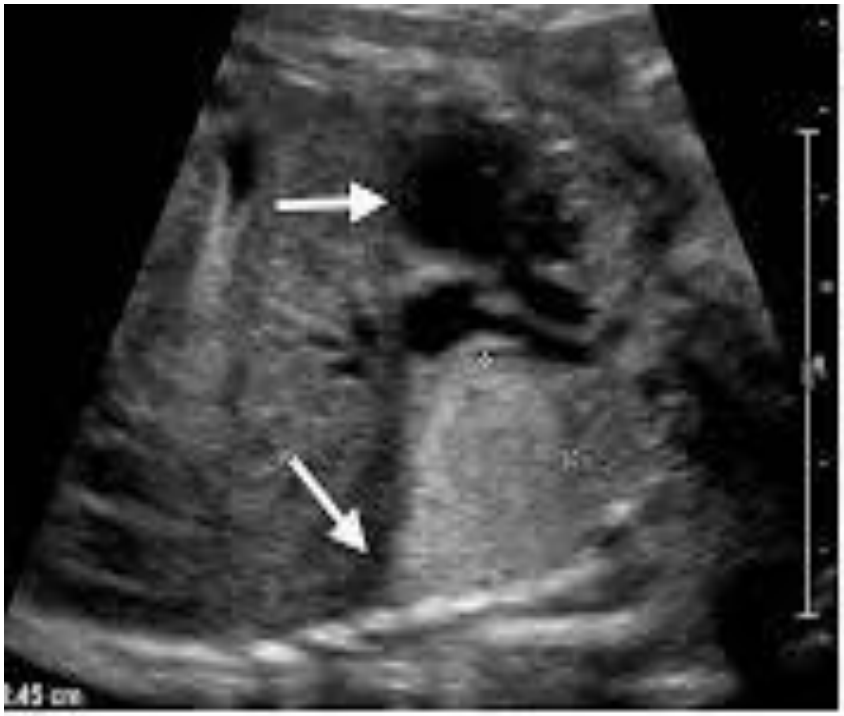
Postnatal diagnosis (Type II)



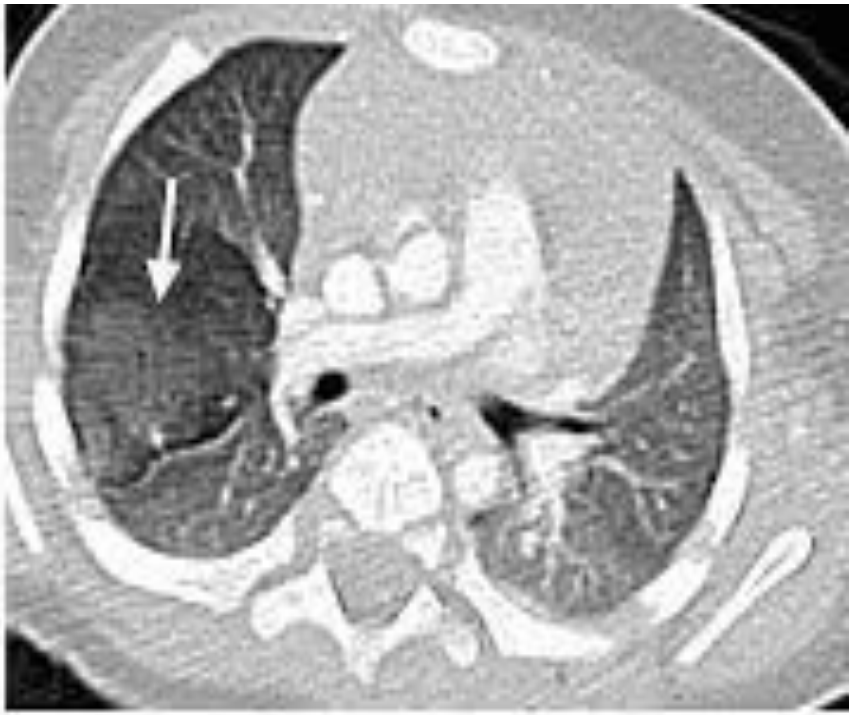


Multiple well defined small cysts (<2 cm in diameter)

Postnatal diagnosis (TYPE III)



a.



b.

Echogenic lung with small cysts less than 5mm.

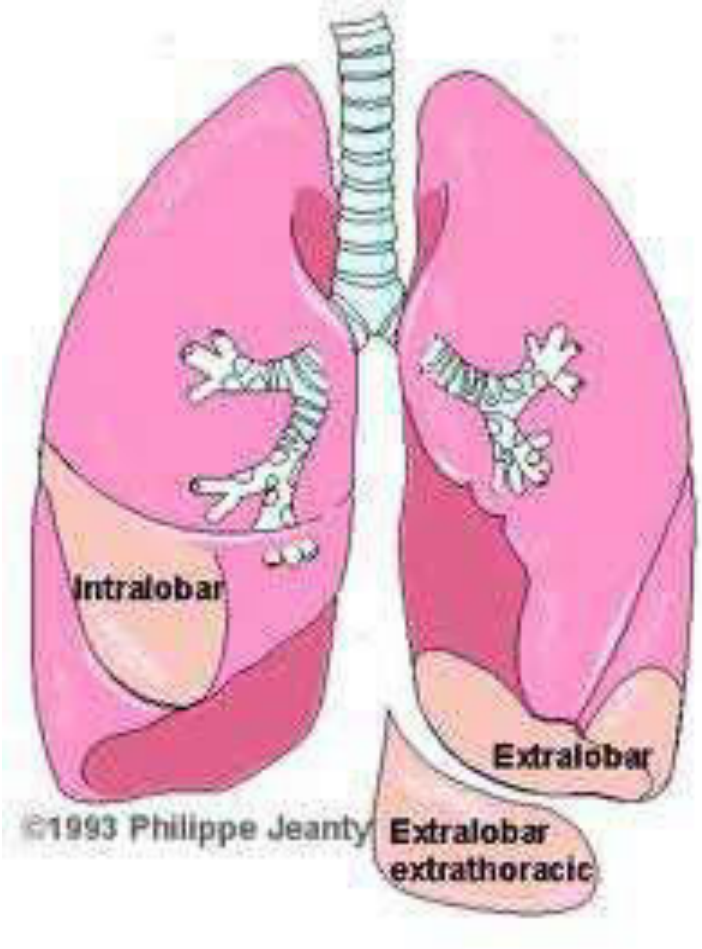
Well defined hyperdense area in right lung with multiple microcysts

Complications

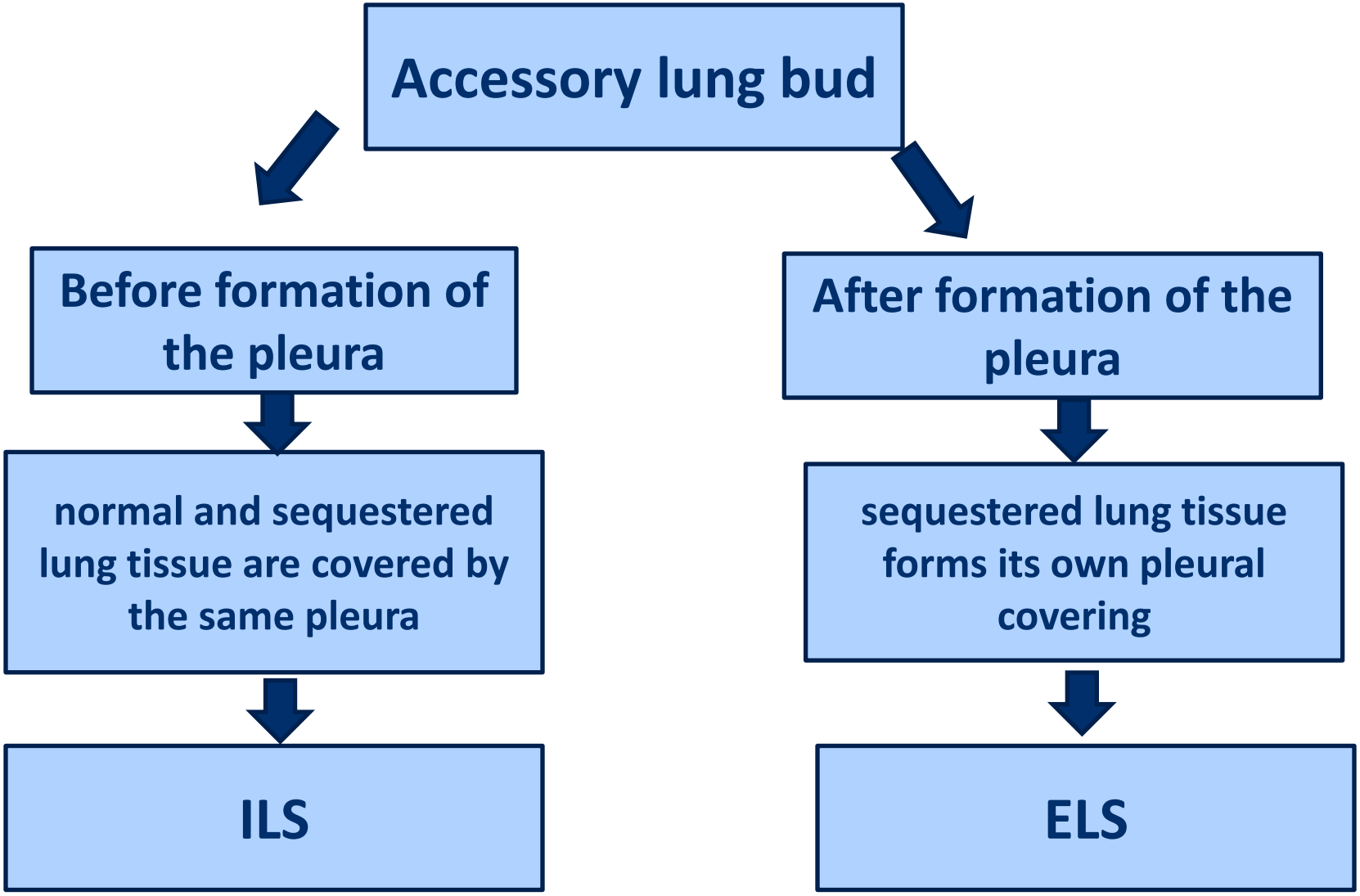
- Infection of the cyst , pneumonia , sepsis
- Rupture and cause pneumothorax
- Pulmonary hypoplasia and PAH
- Haemorrhage into the cyst
- Secondary malignancy

PULMONARY SEQUESTRATION

- Aberrant lung tissue mass that has no normal connection with the bronchial tree or with the pulmonary arteries.
- The arterial blood supply arises from the systemic arteries, usually the thoracic or abdominal aorta
- Types: Intralobar and extralobar



Pathogenesis



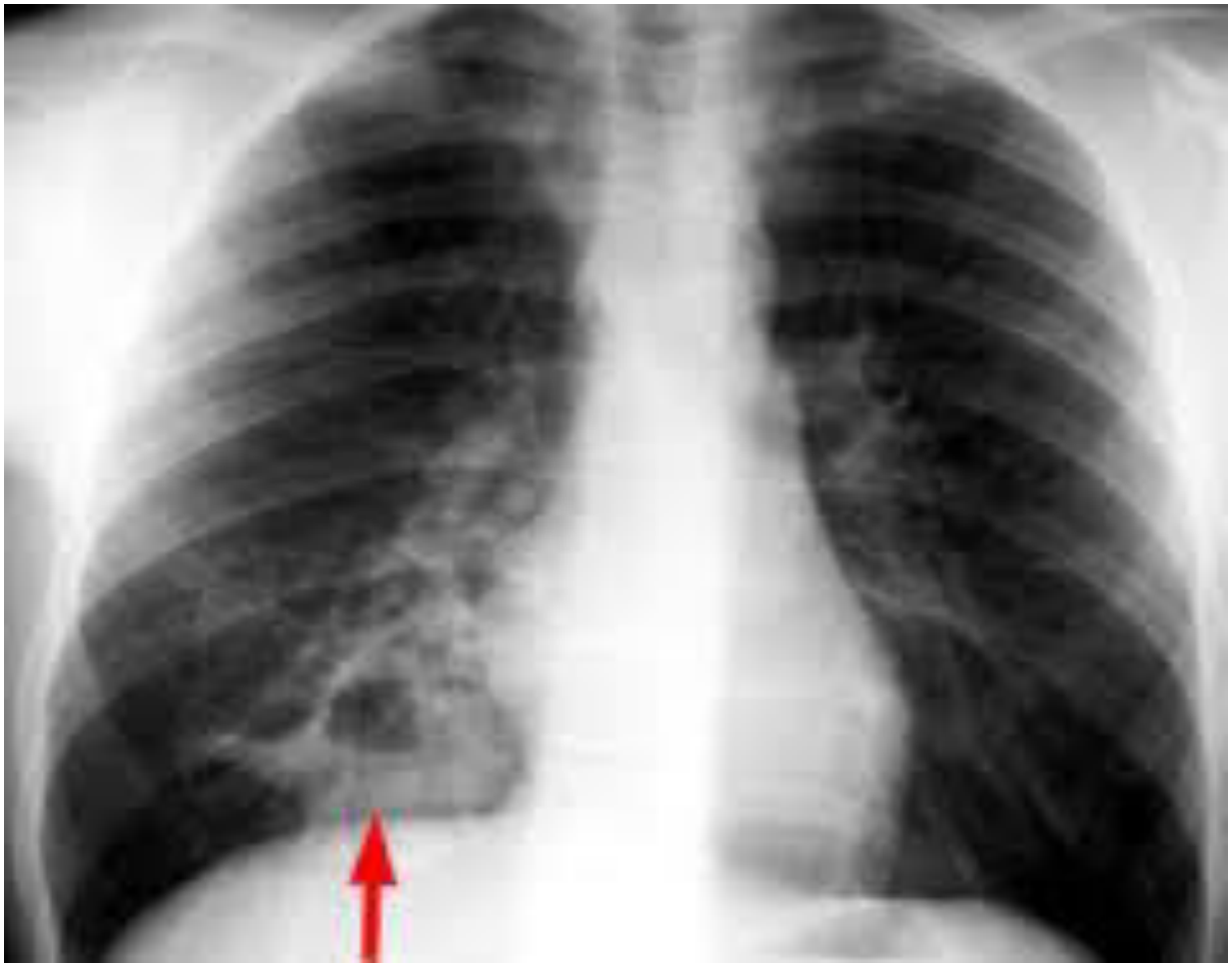
CHARACTERSTIC	INTRALOBAR	EXTRALOBAR
Incidence	More common (75 %)	Less common(25 %)
Gender predisposition	Equal	Men 4: 1
Pleural investment	Shares visceral pleura of parent lobe	Separate visceral pleura
Location	Posterior basal segments (Approx. 60% on left)	Above, below or within diaphragm (Approx. 90% on left)
Venous Drainage	Pulmonary venous	Systemic venous (azygos, IVC, portal)
Presentation	Early adulthood with a history of pulmonary infection, chronic cough, or asthma. Asymptomatic mass (15%)	Mostly present during first 6 months of life due to respiratory or feeding problems
Radiographic Features	Homogeneous consolidation with irregular margins or uniformly dense mass with smooth or lobulated contours.	Single well defined, homogeneous, triangular shaped opacity in the lower thorax. May present else where in the thoracic cavity.

Clinical features

- Common in males, left lower lobe (⅔ cases)
- Asymptomatic in new born and usually detected on chest radiographs taken for other reason
- Intra lobar may present with LRTI, recurrent pneumonic episode or massive haemoptysis
- High output cardiac failure
- Can cause fetal hydrops

Diagnosis

- **Antenatal USG-** well defined, solid, echogenic chest mass - systemic blood supply demonstrated by colour doppler.
- **MR imaging** - confirm the diagnosis.
- **Post natal X-ray** – triangular or oval-shaped basal lung mass on one side of the chest(left side)
- **USG** - confirms the presence of a thoracic mass
- **Chest CT** - identify structure and also the arterial and venous supply
- **MRI** - Identify the feeding arterial vessels & delineate the character of the mass



Right paracardiac triangular opacity

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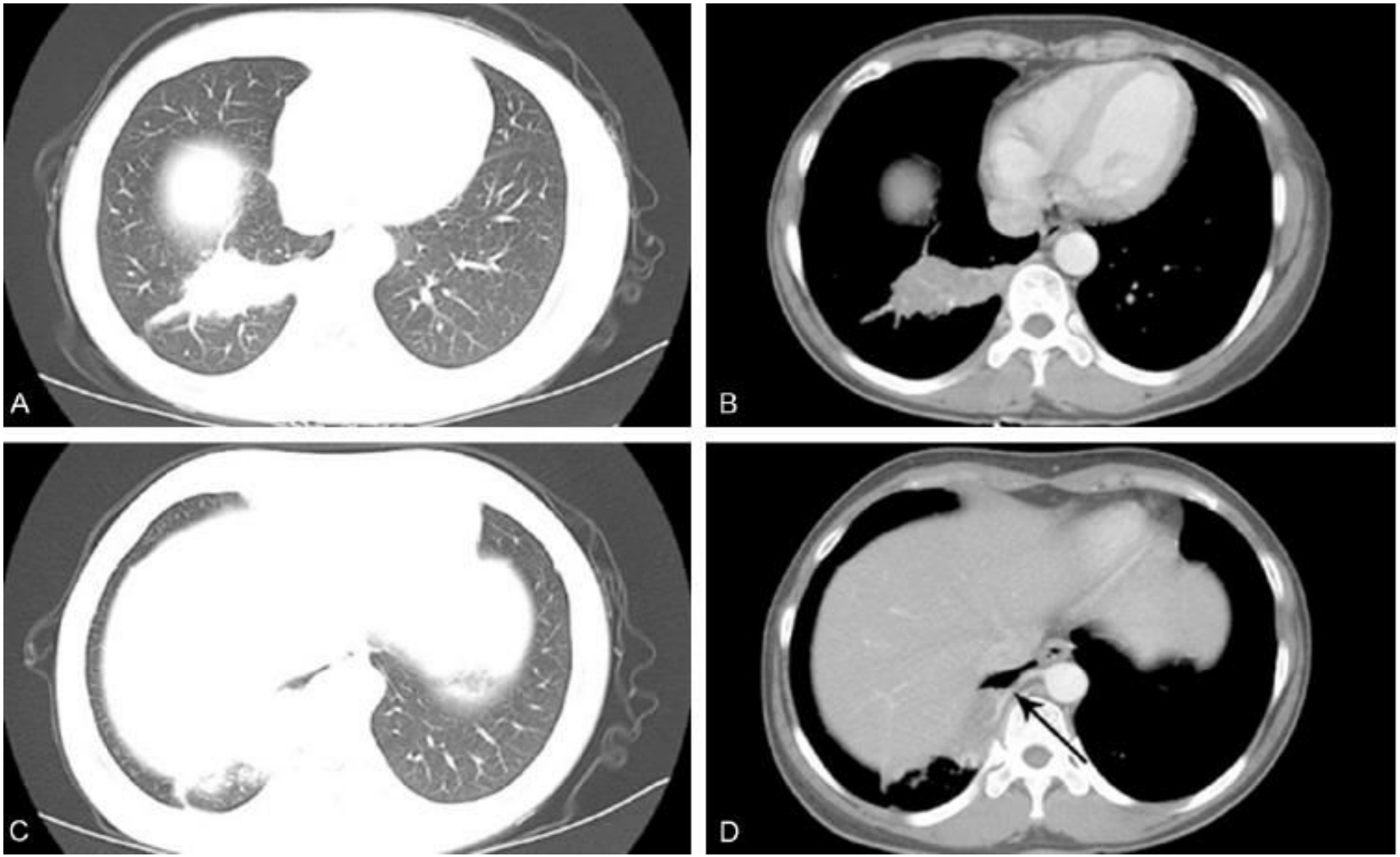
CECT Chest



Management

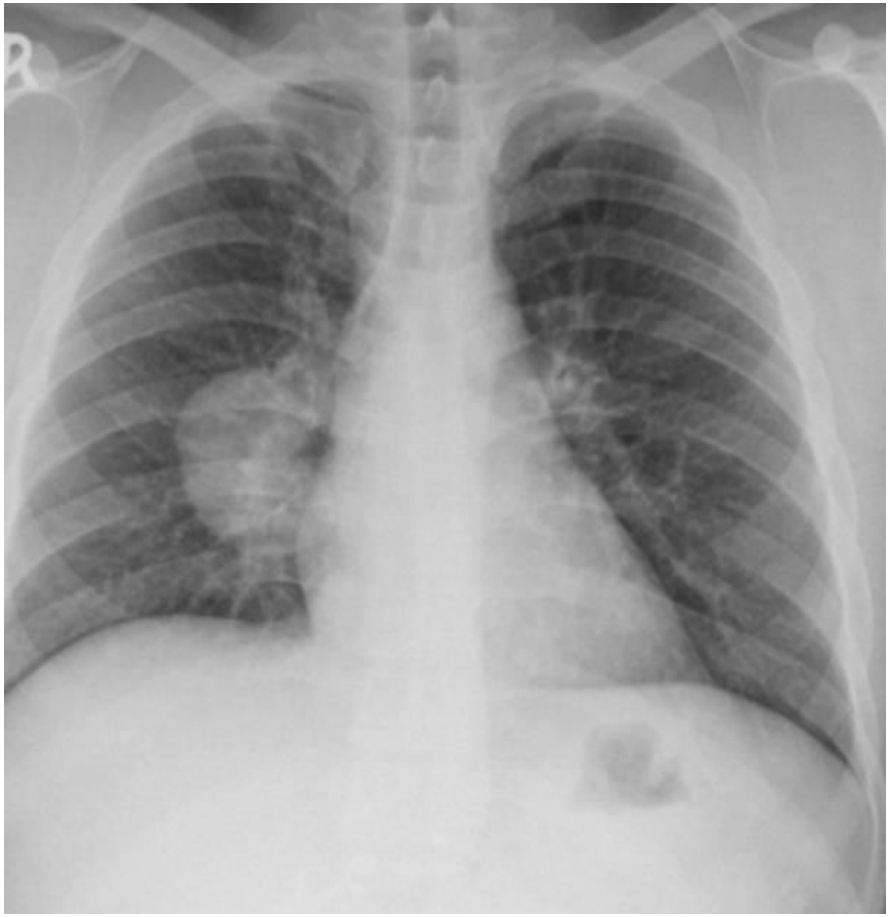
- Most managed surgically.
- Asymptomatic also resected to prevent risk of malignancy or infection
- ILS generally require a segmentectomy or lobectomy, particularly with history of recurrent infection.
- **Embolization** of the feeding arterial vessels has recently been described in the treatment

Bronchogenic cyst

- Single cyst lined by respiratory epithelium and covered with cartilage and smooth muscle
- Abnormal budding of foregut between the 26^{th} and 40^{th} day of fetal life-no further branching.
- Mostly found near the carina but can occur within the lung parenchyma, pleura or diaphragm.

Bronchogenic cyst

- most asymptomatic
- **Diagnosis-** fetal ultrasound and fetal MRI or incidentally on a chest radiograph after birth.
- A **CT scan** is the study of choice



Right mediastinal ovoid mass not obscuring hilum

Bronchogenic cyst

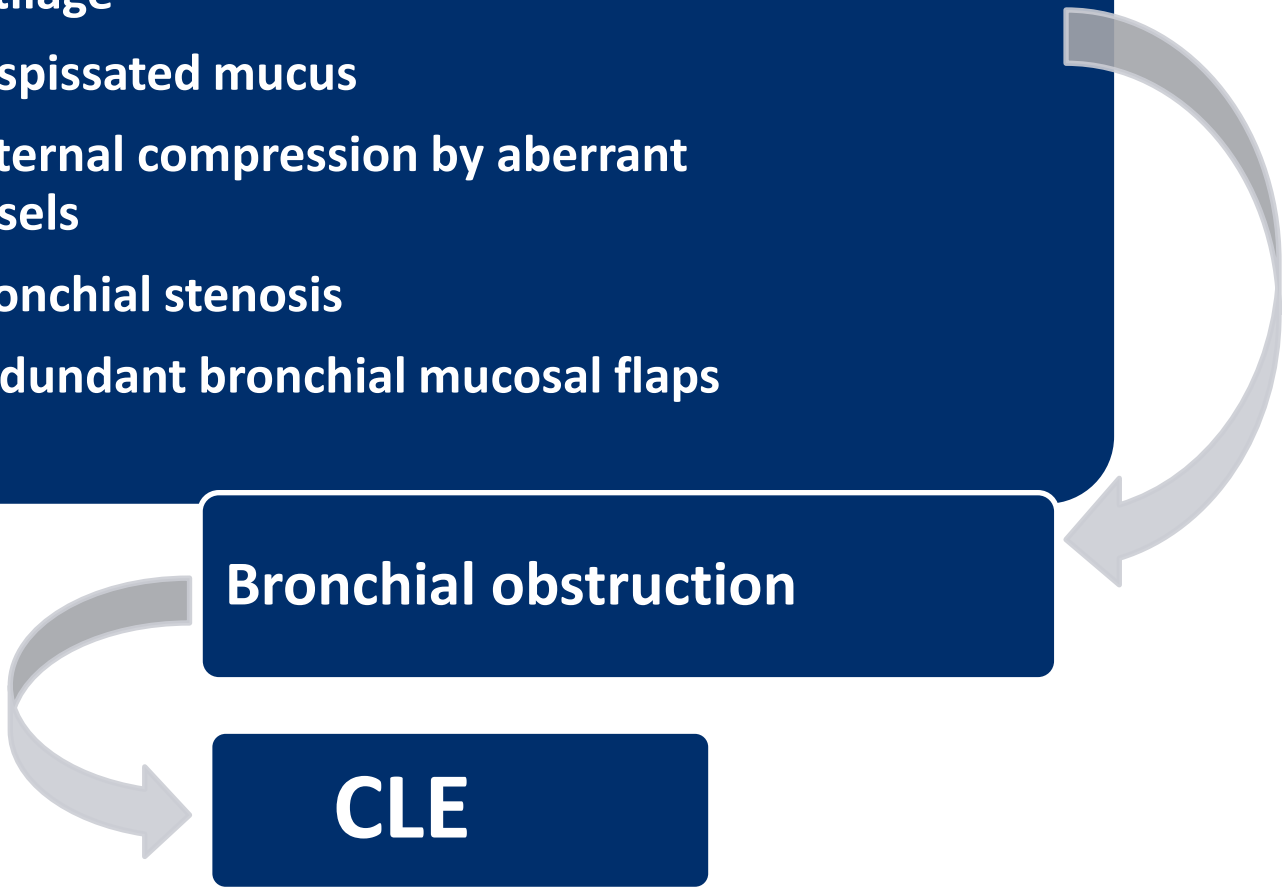
- Symptoms are due to mass effect
- Can cause airway obstruction and air trapping and picture resembling CLE
- Older children present with pneumonia, hemoptysis, pneumothorax, dysphagia or IVC obstruction
- Treatment is complete excision and has an excellent prognosis thereafter .

CONGENITAL LOBAR EMPHYSEMA

- Rare cause
- Males > females
- Left upper lobe > Rt middle lobe.
- Progressive overdistention of one or more segments or lobes - result from a ball-valve mechanism at the bronchial level.
- Cardiovascular anomalies in 12%–14% of cases

Pathology

- Congenital deficiency of the bronchial cartilage
- inspissated mucus
- External compression by aberrant vessels
- Bronchial stenosis
- Redundant bronchial mucosal flaps



Clinical Presentation

- Symptoms at birth (33%), during the 1st month(50%) and most before 1 year of age.
- Mild- severe Respiratory distress
- Examination like tension pneumothorax but are hemodynamically stable

Diagnosis

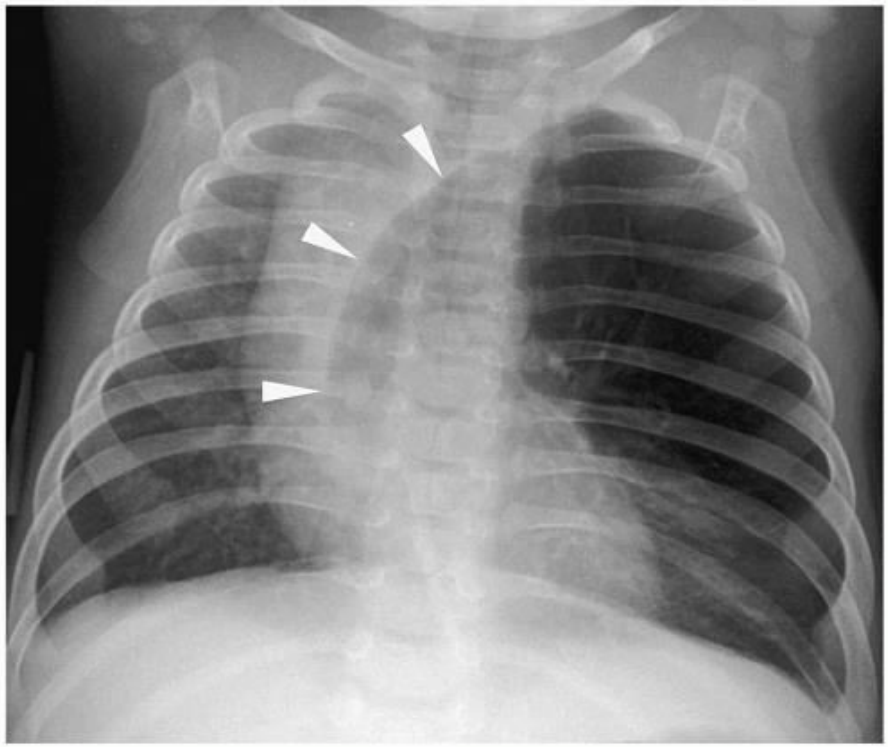
Antenatal

- Fetal USG - homogeneously hyperechoic mass
- Fetal MRI - mass with homogeneously high signal intensity

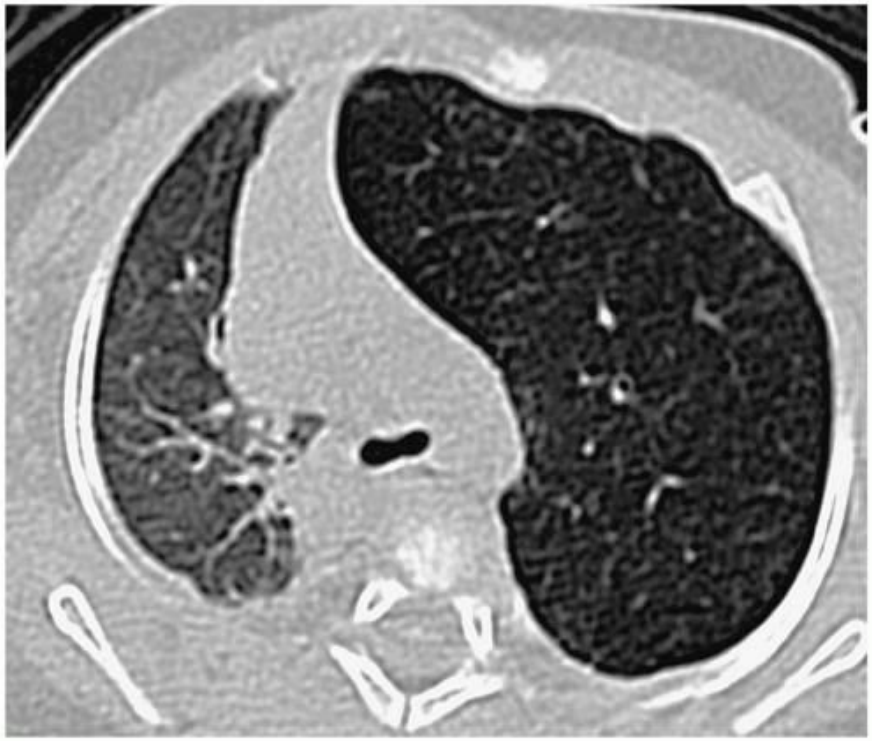
Postnatal

- Chest X-ray - lobar hyperinflation, mediastinal shift, contralateral atelectasis.
- CT scan - decreased vessels due to compression in the involved lung and increased vessels due to overperfusion on the opposite side

X-ray and CT Chest



Hyperinflation of the left upper lobe and herniation across the midline (*arrowheads*). Note the relative paucity of vascular markings and compressive atelectasis of the left lower lobe



Oligemic hyperinflated left upper lobe and mediastinal shift to the right.

Treatment

- Mild- supportive and conservative
- Severe- surgical resection
- Long term prognosis after resection is excellent

Take home message

- Congenital thoracic malformations are rare causes of respiratory distress in newborns
- A majority are self limited and donot require intervention in neonatal period
- Post surgery also there is >90% survival rate
- Outcomes have improved with antenatal diagnosis and management
- Multidisciplinary team approach is required for optimal antenatal and postnatal management

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THANK YOU