CONGENITAL CYSTIC ADENOMATOID MALFORMATION [CCAM]

A series of 8 cases operated in our department.

Dr. Rashmi A Patil.
Department of Paediatric Surgery,
Dr. D. Y. Patil Hospital, Pimpri.

Summary of operated cases

Sr. no	Characteristic	Feature	Number of cases
1	Age at presentation -	Neonates Infants 2-3 years age	3 3 2
2	Gender -	Male Female	5 3
3	Laterality -	Unilateral	8
4	Side -	Right sided left sided	5 3
5	Mode of surgery -	Open thoracotomy	8
6	Type of resection -	Lobectomy of the involved lobe	8

INTRODUCTION

- CCAM is the most common congenital cystic lung malformation, with an incidence of 1:25,000 to 1:35,000 live births and represents 30-47% of fetal thoracic lung lesions.

- It may be diagnosed antenatally and/or in early infancy if symptomatic.

- Surgical resection of the mass is the treatment of choice.

Definition - Now called as CPAM i.e. Congenital Pulmonary Airway Malformation: CCAM/CPAM is a term applied to a cystic, solid or mixed intrapulmonary mass characterized by proliferation of distal airway-like structures and suppression of normal alveolar formation.

- The lesion may communicate with the normal tracheobronchial tree, although this communication is abnormal.
- Mostly unilateral, affects single lobe(95% cases) and mainly lower lobes involved.
- No side/gender predilection.

Pathology

- **Histologically** characterised by –
  - excessive proliferation of terminal respiratory bronchioles
  - polypoid mucosa
  - the cysts lined by respiratory epithelium
  - their walls containing increased elastic tissue and smooth muscle and absence of cartilage.

Types

Stocker classification-

5 types based on the size of cysts and its histologic resemblance to segments of the developing bronchio-alveolar tree.

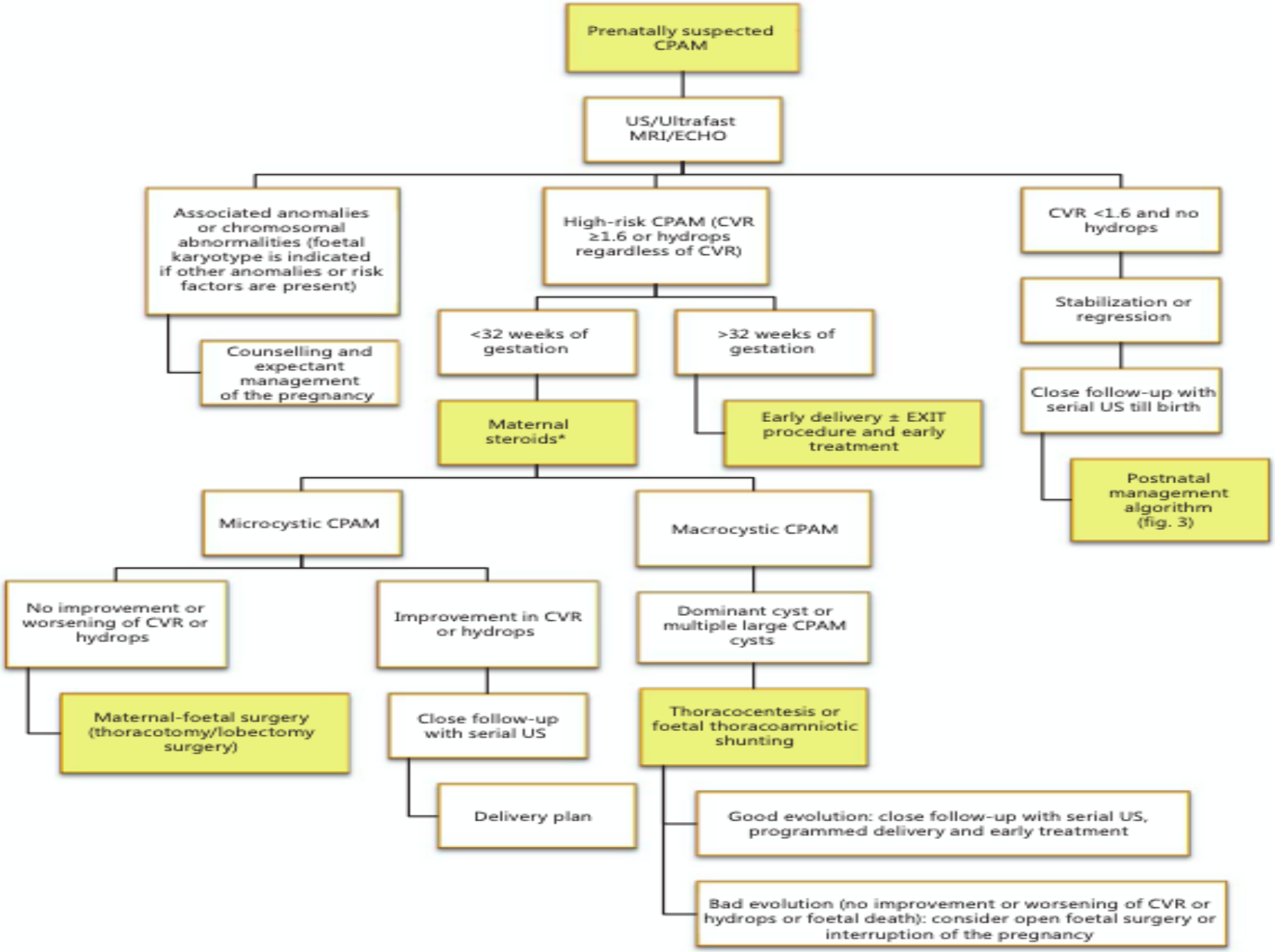
- **Type 0-** Multilobar acinar dysgenesis of major tracheobronchial airways.
- **Type 1-** The most common type(70%); Macrocysts of >2cm size and of bronchial or bronchiolar origin.
- **Type 2-** Same as type 1 but with cysts of 0.5-2 cm size.
- **Type 3-** predominantly solid lesions with microcysts <0.5 cm and of broncho-alveolar duct origin.
- **Type 4-** Large air or fluid-filled cysts of distal acinar origin.

Prenatal diagnosis and treatment

- CCAM can be detected on anomaly scans at 18-20 weeks and can be either microcystic, macrocystic or mixed.

- When diagnosed before 28 weeks of gestation, and
  - If CVR <1.6 --→ weekly USG surveillance
  - If CVR >1.6 --→ twice weekly USG surveillance along with maternal corticosteroid administration.

- When diagnosed after 28 weeks gestation, or massive cysts or with hydrops-(severe Type II and III) -
  - In-utero thoraco-amniotic shunting
  - USG-guided fetal sclerotherapy
  - Laser therapy of lesions
  - Fetal lobectomy using EXIT approach.


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
>32 weeks of gestation
Maternal steroids*
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
Macrocystic 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
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 diagnosis and treatment

- Only 25% cases of CCAM present in neonatal and early infancy.

- The symptoms of varying degrees respiratory distress and infection may be seen.

- Asymptomatic cases may be diagnosed incidentally later in life or may present with late symptoms or with features of suspected malignancy.

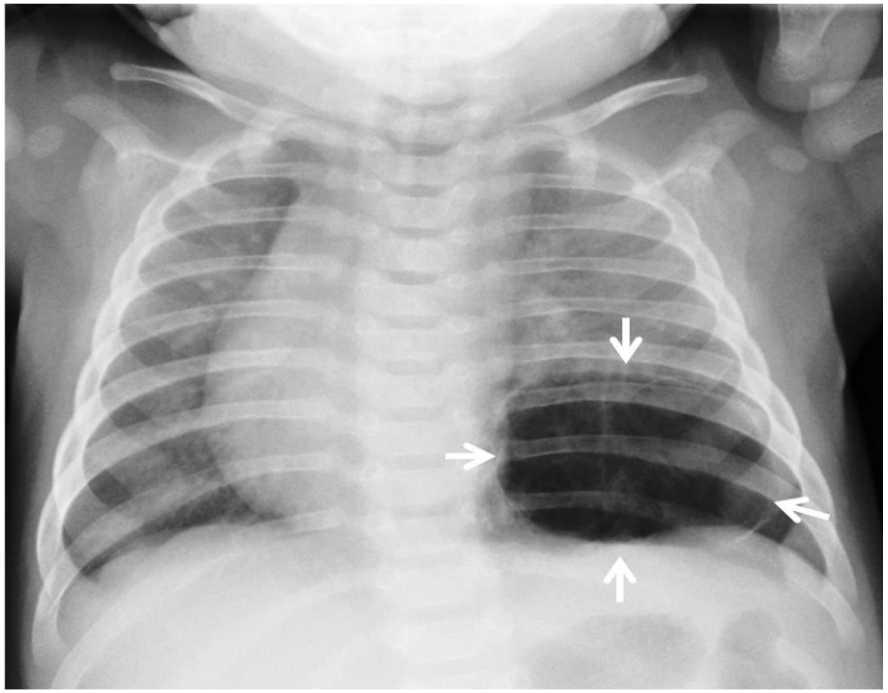
- **Prognosis** - depends on
  - Size of the lesion and
  - The physiological derangements it causes like - mediastinal shift, lung hypoplasia, cardiovascular compromise, polyhydramnios and fetal hydrops.

Diagnosis by Imaging

The radiological features of CPAM differ depending on its type, the content, size, and the number of cysts present.

1) Plain Radiograph of the Chest -

Unilocular Type 1 CCAM →

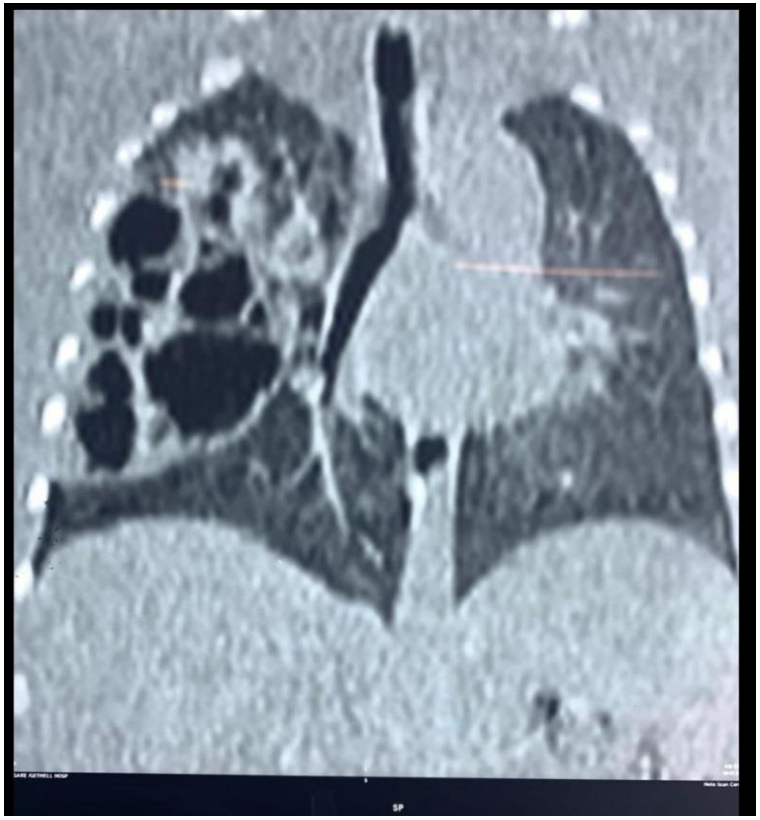


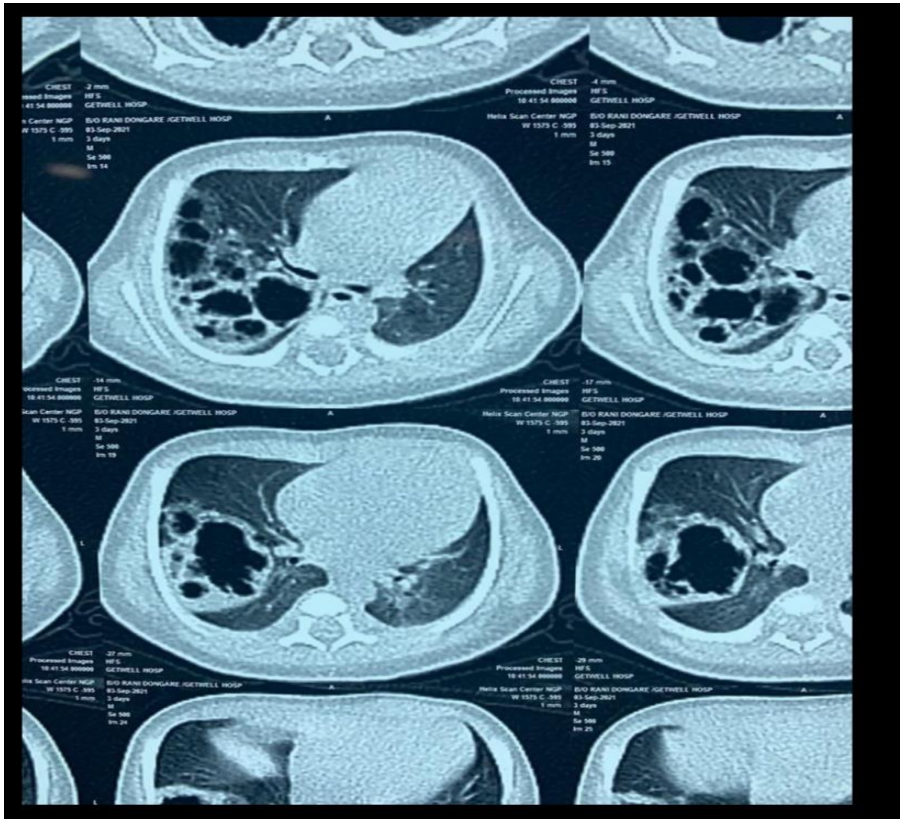
2) Ultrasound -

3) Computed Tomography -

Contrast-enhanced chest CT is the gold standard.

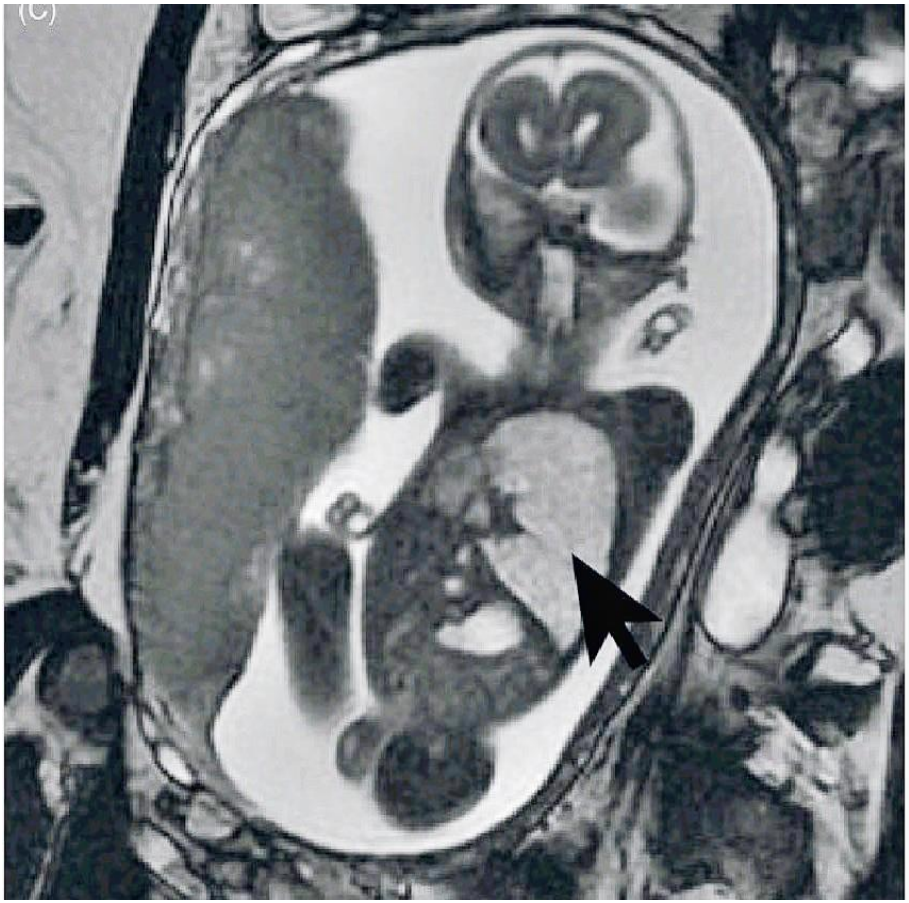
Very useful for the pre-surgical treatment planning.





4) Magnetic Resonance Imaging –

Fetal MRI- showing large left sided CPAM(arrow) with mediastinal shift



Differential Diagnosis

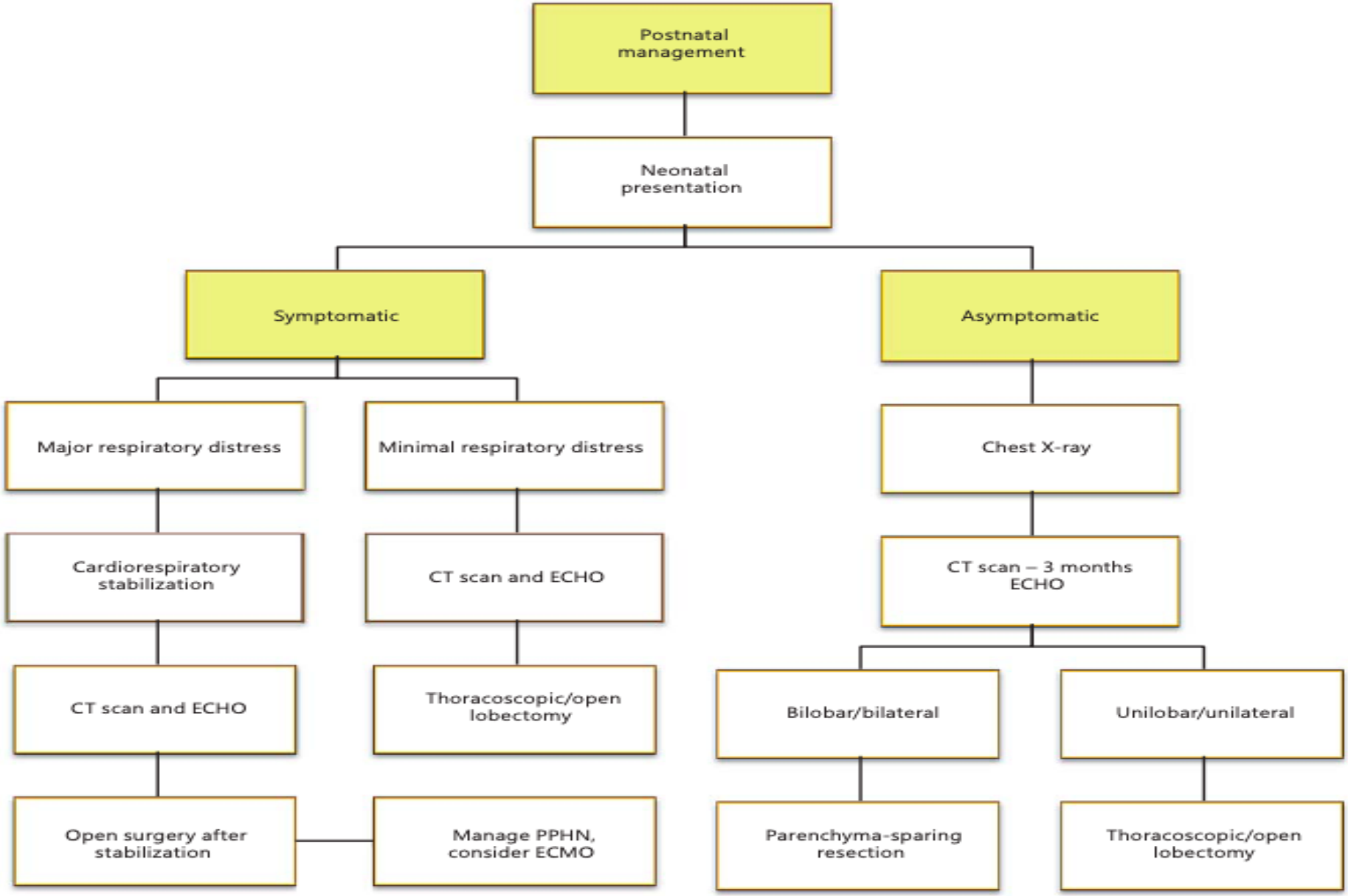
- Congenital lesions-

- Pulmonary sequestrations
- Bronchial atresia
- Congenital lobar emphysema
- Foregut cysts
- Bronchogenic cysts.
- Lung tumours.

- Acquired lesions-

- Pulmonary interstitial emphysema
- Post-infectious pulmonary cysts
- Cystic changes in tumours.

Postnatal management strategy



Preoperative stabilization

- Optimum oxygen delivery with gentle assisted ventilation-
  - Conventional ventilation
  - HFO - High frequency oscillatory ventilation
  - ECMO.

- Cardiovascular and Inotropic support.

- Treatment of PPHN-
  - Inhaled nitric oxide and
  - ECMO

The rationale for operative intervention in asymptomatic CCAM cases

- To avoid occurrence of
  - lung infections,
  - air-leak, and
  - malignancy.
- To have less surgical and post-operative complications.
- To allow for lung maturity and increase in lung volume.
- To relieve parental anxiety about presence the lung lesion in their child.
- To avoid frequent monitoring with CT Scans.

Surgical Treatment

Timing of Surgery -

- In symptomatic cases can be done early or after respiratory stabilization – depending on the severity of the case

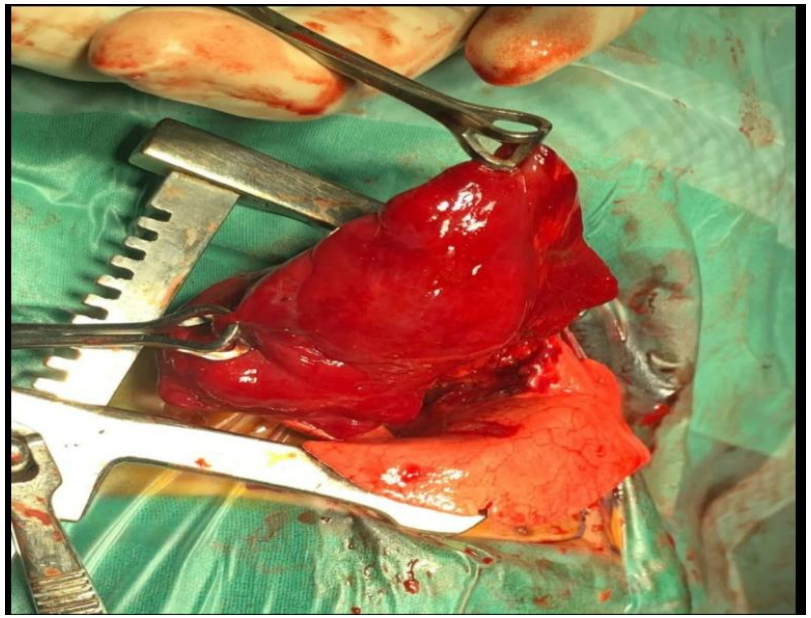
- In asymptomatic cases surgery can be carried out at 6 to 12 months of age.

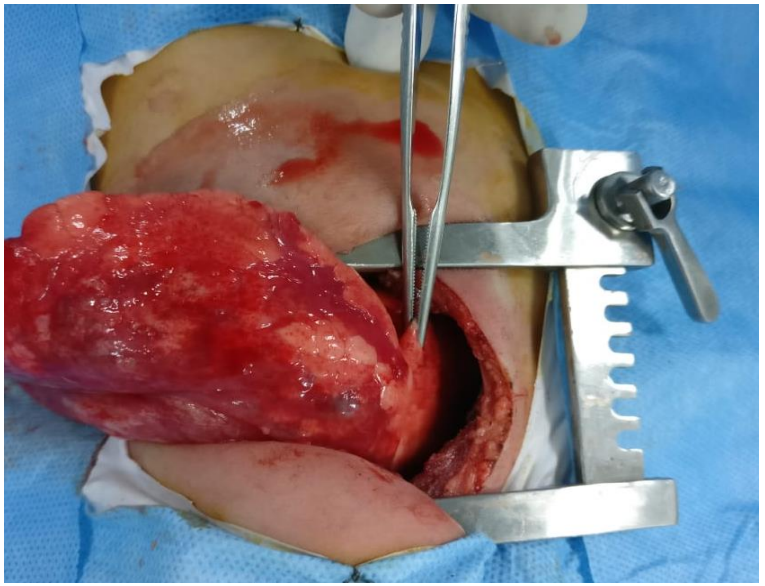
Surgical options -

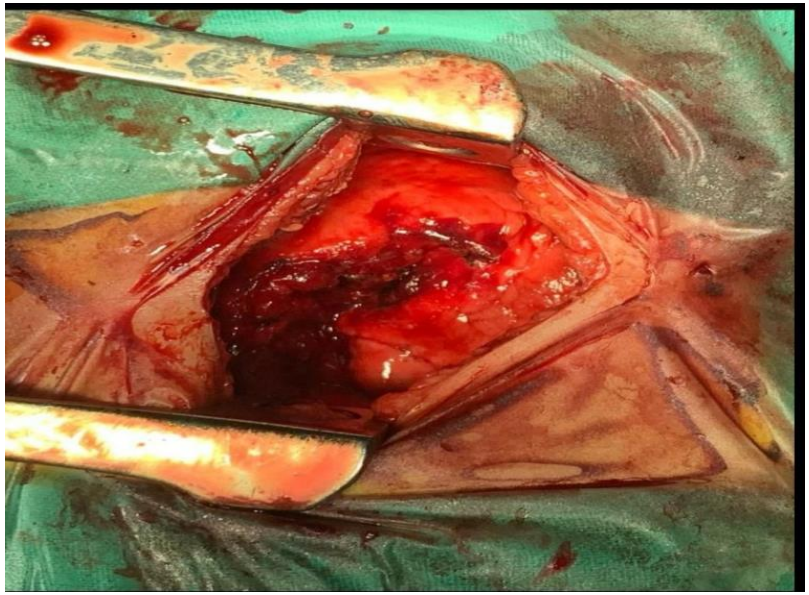
- Complete resection of CCAM is achieved by
  - -> Lobectomy - remains the treatment of choice
  - -> Segmental lung resection.

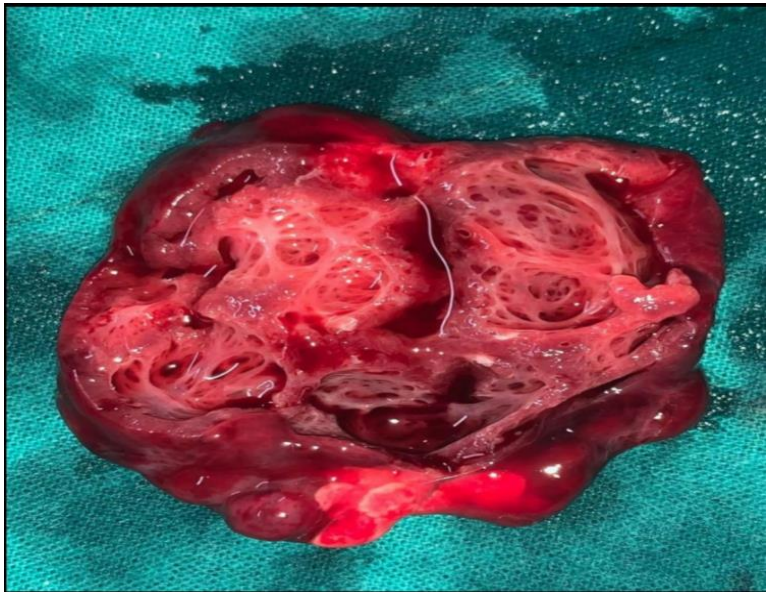
- Can be done either by
  - -> Thoracoscopy
  - -> Conventional open thoracotomy

INTRA - OPERATIVE IMAGES









CONCLUSION

- An index of suspicion for CCAM should be kept especially in recurrent pneumonia cases.
- CECT chest provides detailed anatomical diagnosis.
- Symptomatic cases require surgery irrespective of age.
- Prophylactic excision of asymptomatic lesions should be done before 1 year of age.

THANK YOU