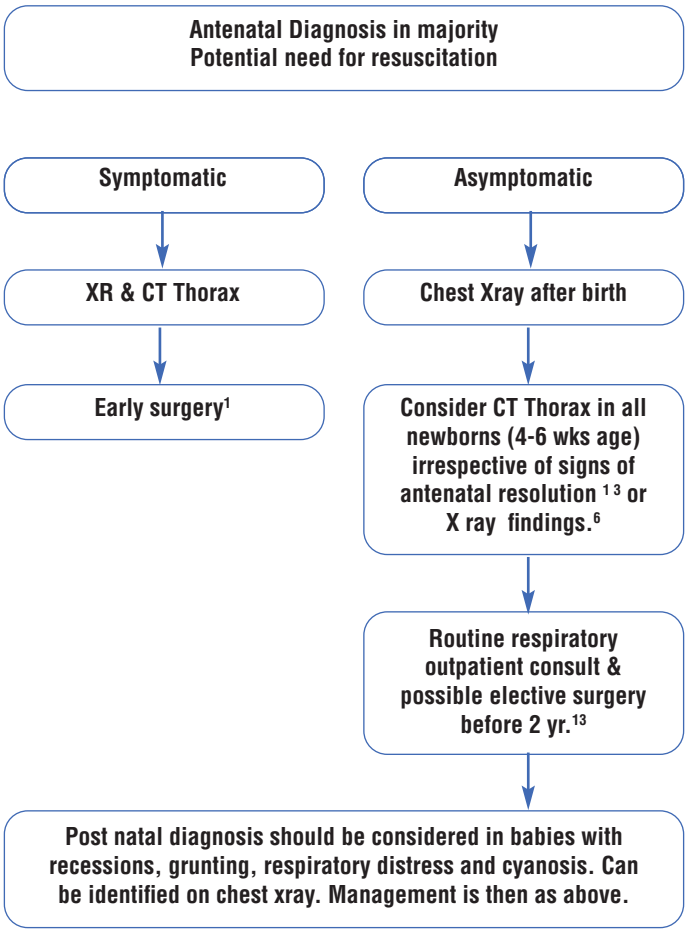
Congenital Cystic Adenomatoid Malformation

- CCAM is a congenital disorder which results in an entire lobe of lung being replaced by a non functioning cystic piece of abnormal lung tissue.
- In rare cases cystic portion can be so large that it affects growth of remaining lung causing pulmonary hypoplasia, cardiac compression & mediastinal shift.
- Fetus may develop hydrops which has significantly poorer outcomes.
- Should have antenatal echo.
- Classification:
  Macrocystic >5mm
  Microcystic <5mm
  Mixed
- Poorer outcome in microcystic, bilateral lesion.



- Majority (>75%) asymptomatic at birth [3].
- Signs include hydrops, bronchial atresia and hypoplastic lungs.
- Symptoms include resp distress, inc RR, decreased sats.
- Surgery recommended to decrease risk of infection, malignant transformation.
- Early surgery v serial CT surveillance in asymptomatic patients is controversial.
- Surgery shown to have better short term outcome.[4]
- Other studies have shown patients can be managed conservatively with 3-6 mth CT thorax until they become symptomatic (pneumonia) or changes in size are observed radiologically.[5]

References:

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This care pathway has been produced by the National Paediatric and Neonatology Clinical Programme. It is aimed at medical, nursing and allied health professionals working in Irish neonatal units.

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