



CLINICAL PRACTICE GUIDELINE

Guideline coverage includes NICU KEMH, NICU PCH and NETS WA

Congenital Cystic Adenomatoid Malformation (CCAM) of the Lung

This document should be read in conjunction with the [Disclaimer](#)

Definition

Congenital cystic adenomatoid malformation is a mass of cysts lined by proliferating bronchial or cuboidal epithelium with intervening normal portions of lung.

Epidemiology

About 25% of affected infants are stillbirths. 20% may have associated congenital abnormalities. CCAM is mainly associated with other pulmonary conditions such as sequestration. Of the heart lesions, CCAM is most commonly associated with Tetralogy of Fallot. Of CCAMs diagnosed antenatally, up to 40% may show regression in size of the cysts as the pregnancy progresses.

Pathophysiology

The hypothesis is that CCAM occurs when there is failure of interaction between endoderm and mesoderm tissue leading to an imbalance with increased cell proliferation and reduced cell death. This leads to abnormal development of the lung. New hypotheses suggest that CCAM may be part of a unifying pathogenesis that would also include sequestration, bronchial atresia and lobar emphysema.

Classification

At present we are still using the modified Stoker classification:

1. Type 0 – acinar atresia
2. Type I – cysts up to 10cm. The cysts are lined by pseudostratified ciliated cells that are often interspersed with rows of mucous cells. Most common form.
3. Type II – Sponge-like multime small cysts (<2cm) and solid pale tumour-like tissue. Occurs in 40% of patients. Most anomalies are associated with type II.
4. Type III – Solid. Excess of bronchiolar structures with cuboidal lining. It is localised to one area. On histology there may be evidence of hamartomatous tissue.
5. Type IV – cysts up to 10cm. The cysts are lined by flattened epithelium resting on loose mesenchymal tissue.

Clinical Presentation

CCAMs present in 3 main ways:

1. Antenatal diagnosis (symptomatic and asymptomatic)
 - Many may have “resolved” on ultrasound and
2. Symptomatic post-natal diagnosis

3. Asymptomatic post-natal diagnosis (incidental on CXR).

Common symptoms and signs are early tachypnoea and poor feeding. The majority of infants are asymptomatic and present later with tachypnoea, repeated chest infections, bronchiectasis and rarely lung abscess.

Approximately 20% have associated malformations of which the majority are other pulmonary associations. Other associations include renal and cardiac.

Long term prognosis depends on whether the infant becomes symptomatic with recurrent infections prior to surgical removal. Emergency surgery carries the highest risk. The majority that are asymptomatic at birth will remain asymptomatic until surgery.

Investigations

- Often made antenatally, with the differential being CDH or sequestered lung.
- Postnatally the CXR should be done on day 1 of life
 - If the infant is asymptomatic delay the CXR until the infant is over 12 hours old. This allows for fluid resorption to occur and improves interpretation of the CXR.

Management

For antenatal diagnosis, refer to antenatal management plan for early resuscitation and post-natal management.

Symptomatic


If the infant is symptomatic at birth then admit to the neonatal unit and perform an early CXR. Discuss acute management with the neonatal consultant and the surgical team at PCH.

If the symptoms are related to the CCAM rather than normal newborn respiratory disease then the baby should be transferred to PCH for evaluation. Infants will likely require a CT chest +/- lobectomy if the baby remains symptomatic.

Asymptomatic

If the infant is asymptomatic then the baby can be managed on the postnatal ward. Perform a CXR on day 1-2 of life and contact the surgical team for management. Most infants can be discharged home with outpatient follow-up and CT scan.

Most infants have the CCAM excised by 1-2 years of age because of the increased risk of adenoma.

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