Section 2: Respiratory problems and management
Congenital cystic adenomatoid malformation of the lung (CCAM)
Date created: June 2006
Date revised: Sept 2014
Review date: Sept 2017

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CONGENITAL CYSTIC ADENOMATOID MALFORMATION OF THE LUNG (CCAM)

This 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. 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

There are 3 pathological types. Greater than one lobe may be involved but the defect is always restricted to one lung.

1. Type I is the most common and consists of large cyst(s) and some smaller ones that are localised to a single lobe of the lung.
2. Type II occurs in 40% of patients and are associated with other congenital anomalies which may be fatal.
3. Type III is localised to one area of the lung and contains multiple small cysts that form a firm mass. On histology there may be evidence of hamartomatous tissue.

CLINICAL PRESENTATION

The infant develops acute respiratory distress with tachypnoea and cyanosis immediately after birth. Symptoms can be absent or delayed for days/weeks in those with less extensive lesions. The cardiac apex may be shifted from the affected side with reduced breath sounds over the lesions. There is an increased risk of infection and adenoma of the lung if the CCAM is not removed. Hydrops is associated with a worse prognosis.

INVESTIGATIONS

- Often made antenatally, with the differential being CDH or sequestrated lung.
- Postnatally the CXR will show expansile cystic mass with mediastinal shift. 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

After discussion with the neonatal consultant refer to the Surgeons at PMH. Most infants have the CCAM excised by 1-2 years of age because of the increased risk of adenoma. If the infant is symptomatic at birth the CCAM will be excised earlier. Depending on the size of the lesion, there may be pulmonary hypertension, which should be treated as above.