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Best practice guidelines

Congenital cystic lesions of the lung

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ABSTRACT

Congenital cystic lesions of the lung are present in 1 in 10,000–35,000 births and present as a spectrum of anomalies. Majority of these cystic lesions comprise congenital cystic adenomatoid malformations, pulmonary sequestrations, congenital lobar emphysema, and bronchogenic cysts. Most of these lesions are nowadays detected antenatally, however some will present either in the newborn or during later childhood. A review of the aetiology, classification, natural history, investigations, and treatment of congenital cystic lung lesions is discussed.

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1. Congenital cystic adenomatoid malformations

Congenital cystic adenomatoid malformations (CCAM) are characterised by multicystic areas of over-proliferation and dilatation of terminal respiratory bronchioles with lack of normal alveoli. CCAM are intrapulmonary lesions that contain various types of epithelial linings and maintain communication with the normal trachea–bronchial tree and retain a normal blood supply. They are usually unilobar and unilateral. The reported incidence is between 1 in 10,000 to 1 in 35,000 with no predilection for side of lung, sex, or race. Close to 100% of CCAMs are detected on antenatal ultrasound by 20 week gestation [1].

There are two classifications in practice — Stocker's classification which subtypes the resected cysts into 5 categories dependent upon the size and epithelial lining of the cyst. [Table 1] This classification system has been criticised for failing to recognise hybrid lesions that have features of CCAM and pulmonary sequestrations and not being applicable to antenatally detected lesions. There has been a correlation between Stocker type 1 with bronchoalveolar carcinoma and Stocker type 4 and pleuropulmonary blastomas. A further classification was developed by Adzick based on antenatal scanning which sub-divides antenatally detected lesions into two categories — macrocystic (type 1) and microcystic (type 2) [1,2] [Table 2].

Close monitoring of the antenatally detected lesion with serial ultrasounds to detect the size of the lesion, location, volume, blood supply and compromise to the foetus is performed. The cystic adenomatous malformation volume ratio [3] predicts an 80% increased risk of hydrops fetalis if more than 1.6 and a ratio less than 1.6 is associated with a survival rate of 94% and less than 3% risk of hydrops fetalis. In 10% of cases there will be need for foetal intervention due to enlarging mass or development of hydrops fetalis in association with an isolated lung lesion. Pre-requisites for intervention include normal chromosomal analysis and absence of other obvious anomalies. These interventions include thoracocentesis, thoracoamniotic shunt placement, percutaneous laser ablation, and rarely open foetal surgery. In a review by Adzick and Kitano [4] of children with CCAM requiring foetal intervention, it was found that thoracocentesis was not beneficial as it rapidly reaccumulated and did not seem to alter the long-term outcome and that open foetal surgery carried a 50% mortality. The risk of open foetal surgery in Adzick and Kitano's review found a 23% risk of pulmonary

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Table 1
Stocker classification.

Type	Histological features	Prevalence
0	Involvement of all lung lobes, stillborn	<2%
1	Single or multiple cysts >2 cm, pseudostratified columnar epithelium	60–70%
2	Single or multiple cysts <2 cm, cuboidal or columnar epithelium	15–20%
3	Predominately solid lesions, <0.5 cm cysts, cuboidal epithelium	5–10%
4	Large air-filled cysts, flattened epithelial cells	<10%

interstitial oedema in mothers, 34% undergo chorioamniotic separation, and amniotic fluid leak from hysterotomy site or from rupture of membranes [4].

The sensitivity of chest X-ray in the postnatal period for detecting CCAM is only 61% [5], thus necessitating the need for other modalities of postnatal imaging to determine the persistent presence of the CCAM. This same study also showed that CT was 100% sensitive in detecting postnatal CCAMs. Our local policy is that any child with an antenatally diagnosed CCAM will have a CT chest performed at 1 month of age without a general anaesthetic (feed and wrap technique) with clinic assessment at 2 months old and surgery, if necessary, at 3–6 months old. [Fig. 1] Although it can be argued that MRI can be substituted as the postnatal imaging of choice due to the radiation dose in children and the lifelong risk of malignancy, neonates currently require a general anaesthetic for MRIs to be performed and, therefore, until the technology improves to allow an MRI scan to be performed without a general anaesthetic we recommend the CT scan as the imaging modality of choice.

A further area of controversy is the timing of surgery in the asymptomatic group. A recent review has shown that early surgery is technically easier; risk of infection reduced and need for respiratory support is decreased. Thus the recommended timing of surgery is between 3 and 6 months of age as the procedure is well tolerated, there is a lower risk of infection, and allows more time for compensatory lung growth [5].

Management of symptomatic CCAM is well established. Most babies will require respiratory support at birth and the diagnosis is confirmed using CT scan or MRI. The open thoracotomy surgical technique is recommended as the thoracoscopic approach has a tendency to destabilise the symptomatic patient.

The treatment of the asymptomatic child with CCAM is controversial. Arguments for excising a CCAM include the risk of recurrent pneumonia, lung abscess, empyema, or malignancy as bronchoalveolar carcinoma, pleuropulmonary blastoma, and rhabdomyosarcoma are known to have an association with CCAMs. [6] Also, delaying the excision until the child becomes symptomatic carries a greater morbidity as evidenced in the study by Sueyoshi et al. [6] which demonstrated that in all patients that presented as postnatal diagnoses following pneumonia there was a significantly longer duration of surgery and a significantly greater intra-operative blood loss. Performing follow up X-rays or CT scans carries the risk of associated malignancy with radiation exposure.

With the technological advancements in minimally invasive surgery, congenital lung lesions are now commonly removed by video-assisted thoracoscopy (VATS). In our institution, VATS is the procedure of choice for CCAM excision as it has been demonstrated that VATS is a safe and feasible alternative to performing open thoracotomies in children with congenital lung lesions [7].

Those that argue against surgical intervention in asymptomatic children reason that the natural history of CCAM is not well defined thus the true risk of developing future symptoms may be exaggerated. With reports of CCAMs spontaneously resolving on CXR, although it has been shown that the sensitivity of detecting CCAM on CXR is only 61% [5], during postnatal follow up there is the argument of watch and wait and that the risk of radiation exposure can be reduced with the use of MRI, although MRIs in children require a general anaesthetic which carries its own risks. With these arguments, the group against surgical intervention in the asymptomatic child believe that surgical intervention may subject the child to more risks than conservative management [8].

Table 2
Adzick classification.

Type	Ultrasound features	Prevalence
Macrocystic (type 1)	Single or multiple cysts >5 mm	58%
Microcystic (type 2)	Single or multiple cysts <5 mm	42%

2. Pulmonary sequestration

Pulmonary sequestrations are portions of the lung that are in isolation from its neighbouring lung tissues — it has no communication with the bronchial tree and receives its blood supply from a systemic artery, and venous drainage may occur through either a pulmonary or systemic vein. [Fig. 2] It is estimated to represent up to 6% of congenital lung anomalies. Pulmonary sequestrations can be divided into two categories: intralobar (15%) and extralobar sequestrations (85%).

The aetiology of these lesions is under much debate. The predominate theory was proposed by Langston [9] that pulmonary sequestrations are embryologic developmental anomalies and are derived from an accessory lung bud. Other theories include vascular abnormalities, recruited vascular supply secondary to infection, and as an additional lung bud that has developed its own blood supply.

Extralobar sequestrations are contained within a pleural covering, thereby maintaining an anatomical boundary between itself and surrounding lung tissue. Most are found within the thoracic cavity, however, in a minority of cases, they can be found below the diaphragm. Most patients present within the first 6 months of life, of which about 25% will experience respiratory distress shortly after birth. Antenatal presentation may consist of foetal hydrops, polyhydramnios, or pleural effusion. In the postnatal period, children may present with a chronic cough, recurrent chest infections, difficulty feeding, or abdominal pain. Sixty percent will have another developmental anomaly. Congenital diaphragmatic hernia is the most common associated anomaly, with other anomalies including lung hypoplasia, congenital cystic adenomatoid malformation, congenital lobar emphysema, and bronchogenic cysts. There are also numerous associated cardiac malformations which include pericardial defects, dextrocardia, truncus arteriosus, and total anomalous pulmonary venous drainage. In 75% of cases, the arterial blood supply is derived from either the thoracic or abdominal aorta, however the arterial supply may originate from the subclavian, intercostal, phrenic, internal thoracic, coeliac trunk, or left gastric arteries [9].

Intralobar sequestrations are found within normal lung tissue. If not detected on antenatal scans, presentation is usually after the age of two years with chronic pneumonia. They predominately affect the medial and posterior basal segments of the lower lung lobes [10]. The arterial supply is typically derived from the lower thoracic or upper abdominal aorta.

Pulmonary sequestrations usually appear as defined homogenous echo-dense mass on ultrasound and on antenatal scans may present as a small lesion or may occupy a large portion of a hemithorax, causing mediastinal shift and occasionally hydrops fetalis. Although lesions may appear to regress on antenatal scans, postnatal imaging should be performed to confirm complete regression. Importance during investigation of pulmonary sequestrations is placed on the mapping of the blood supply. Doppler ultrasound, CT, and MRI are the primary modalities used; however each has its advantages and pitfalls. Doppler ultrasound can demonstrate the typical appearance of pulmonary sequestration and can be used to map the venous and arterial blood supply but will

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