



## British Paediatric Orphan Lung Diseases (BPOLD)

### **Congenital Cystic Adenomatoid Malformation (CCAM) and other congenital cystic lung lesions - [Dr Andrew Bush](#)**

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##### **Definition**

A group of congenital malformations of the developing lung, which may be supplied by either or both of the pulmonary or systemic arterial system, and drain to pulmonary or systemic veins. The histology of the excised specimen may show many different cell and tissue types.

##### **Causes**

No known causes identified.

##### **Clinical Presentations**

Hitherto, CCAM and other congenital cystic malformations of the lung (which I group together as congenital thoracic malformations, or CTMs [1]) usually presented with symptoms such as recurrent infection, and it was obvious that treatment was needed. Now they present in the pre-symptomatic phase, and it is difficult to know how best to advise the parents. In most cases, the baby is well after delivery.

##### **Investigations**

Serial monitoring antenatally with ultrasound scans. Postnatally, the chest radiograph (CXR) is either normal, or reveals minor changes only. The CT scan often shows a more extensive lesion than the CXR. The blood supply to the CTM should be defined by contrast CT or magnetic resonance imaging angiography.

##### **Treatment**

In the short term, the prognosis of antenatally diagnosed lesions is very good; although at 20 weeks gestation the lesions may be large, in many babies they regress considerably, and most survive to be born at term. The Kings group have reported on 67 fetuses with an antenatally diagnosed congenital lung [2]. Sixty four were born alive, and 42 underwent postnatal surgery. Surgery was required in 45% of lesions showing late gestation 'resolution'. Although there was some correlation between the antenatal appearances and the need for surgery, this was not usefully predictive for an individual, and the need for operation was judged on postnatal features. Specifically, hydrops and mediastinal shift are not such bad prognostic features as was once thought.

Babies with a very large CTM, or one that is compressing the airway, may present with respiratory distress in the newborn period, in which case immediate surgery is likely to be required. If the baby is well, are no data on which to base treatment decisions. It is not possible to quote a risk of complications, because the denominator of the risk equation (how many undiagnosed CTMs in the past have caused no problems) is unknown.

Possible complications include infection, pneumothorax, bleeding from or into the CTM, malignant change, and high output heart failure if the CTM is fed by a large systemic artery. Complications are rare in early life, and many of the protagonists of surgery would usually recommend resection of the lesion at about 2 years of age. As an alternative to



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surgery, a CTM supplied by a large systemic vessel may be considered for coil embolization, which may be the only therapy needed. Others would advise an expectant approach, particularly with small CTMs. If a CTM does become infected, then surgery is inevitable; antibiotics can lead to temporary improvement, but recurrent infection is inevitable. The risk of malignancy is less clear. Primary intrathoracic malignancy, irrespective of the presence of a CTM, is rare. There are isolated case reports of malignant tumours in children with a CTM. The risk attributable to the malformation is not known. Metaplasia or pre-neoplastic change is not a feature of excised CTMs [3]. Furthermore, malignant disease may develop in sites distant from the original malformation [4], implying that the malformation is merely a marker of increased malignant potential throughout the lungs, in which case removing the malformation would not deal with the underlying problem. It is suggested that a higher incidence of suspicion is justified in those with bilateral disease, a family history of pleuropulmonary blastoma, pulmonary cysts or renal anomalies, or a close relative with a childhood malignancy, especially Wilm's tumour or medulloblastoma

### **Useful references:**

<b>1. Congenital lung disease: a plea for clear thinking and clear nomenclature.</b>	<b>Bush A.</b>	<b>Pediatr Pulmonol 2001; 32: 328-337</b>
<b>2. Current outcome of antenatally diagnosed cystic lung disease.</b>	<b>Davenport M, Warne SA, Cacciaguerra S, Patel S, Greenough A, Nicolaides K.</b>	<b>J Pediatr Surg 2004; 39: 549-56</b>
<b>3. Pulmonary sequestration: a review of 26 cases.</b>	<b>Halkic N, Cuenoud PF, Corthesy ME, Ksontini R, Boumghar M.</b>	<b>Eur J Cardiothorac Surg 1998; 14: 127-133</b>
<b>4. Pleuropulmonary blastoma: is prophylactic resection of congenital lung cysts effective?</b>	<b>Papagiannopoulos KA, Sheppard M, Bush A, Goldstraw P.</b>	<b>Ann Thorac Surg 2001; 72: 604-605</b>

### **Web links:**

Further information on the relationship between CCAM and pleuropulmonary blastoma can be found at [www.ppbregistry.org](http://www.ppbregistry.org).

Clearly, more evidence about the natural history of untreated CTM, and the risk of complications, is needed. Whether or not the baby has surgery, it is essential that normal child care, including immunizations, is carried out. There is an ongoing natural history study which is attempting to answer some of these questions prospectively, recruiting babies at the time of antenatal ultrasound diagnosis. If you are interested in being part of



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this study (LOTOS), further details can be obtained from the principal investigator, Dr Lyn Chitty ([l.chitty@ich.ucl.ac.uk](mailto:l.chitty@ich.ucl.ac.uk)).