Seminars in Pediatric Surgery (2005) 14, 16-33

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SEMINARS IN PEDIATRIC SURGERY

# Asymptomatic congenital lung malformations

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**KEYWORDS**
Congenital cystic adenomatoid malformation;
Pulmonary sequestration;
Bronchogenic cyst;
Congenital lobar emphysema;
Primary pulmonary neoplasm

Congenital lung malformations are often discovered incidentally on routine prenatal sonography or postnatal imaging. Lesions such as congenital cystic adenomatoid malformations (CCAM), sequestrations, bronchogenic cysts and congenital lobar emphysema may be asymptomatic at birth or at the time of discovery later in life. Some authors advocate simple observation because of the lack of data on the incidence of long-term complications. However, there are very few described cases where CCAM and intralobar sequestration have remained asymptomatic throughout life; complications eventually develop in virtually all patients. The most common complication is pneumonia, which may respond poorly to medical treatment. Other complications include the development of malignancies (carcinomas and pleuropulmonary blastomas), pneumothorax and hemoptysis or hemothorax. Since lung resection will be required sooner or later for CCAM, intralobar sequestration and intrapulmonary bronchogenic cysts it is best not to wait for complications to occur. For patients diagnosed prenatally, we recommend surgery at 3 to 6 months of life at the latest, so that compensatory lung growth can occur. At this age the postoperative course is usually smooth and long-term follow-up has shown normal respiratory function. Mediastinal bronchogenic cysts also tend to become symptomatic and elective resection is recommended. On the other hand, asymptomatic congenital lobar emphysema may regress spontaneously and observation is warranted. The management of small noncommunicating extralobar sequestrations is more controversial; it is known that these lesions can remain asymptomatic throughout life but complications may develop and they are sometimes difficult to differentiate from neuroblastoma.
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Congenital lung malformations include a wide spectrum of developmental abnormalities, some of which are incompatible with life or cause severe symptoms in the prenatal or neonatal period. This review focuses on malformations that may be discovered incidentally on routine prenatal sonograms or postnatal imaging. These include congenital cystic adenomatoid malformations, pulmonary sequestrations, bronchogenic cysts and congenital lobar emphysema (polyalveolar lobe and lobar overinflation).[1-4]

*Congenital cystic adenomatoid malformation* (CCAM) is considered a hamartomatous lesion of the bronchial tree by some, while others favor a localized arrest in the development of the fetal bronchial tree as the etiology.[1-5] Because some types are not cystic and only one type has the adenomatoid appearance, the term "congenital pulmonary airway malformation" (CPAM) has recently been proposed.[4] These lesions were initially classified into three types by Stocker,[6] who more recently added two more variants (types 0 and 4).[7] The pathogenesis of CCAM has been the subject of controversy.[5,8-10] Langston believes that the five types may represent different malformations with varying etiologies.[11]

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doi:10.1053/j.sempedsurg.2004.10.022

Other studies support the notion that dysregulation in the branching morphogenesis of the lung is associated with the development of abnormal lung tissue, both in CCAM and sequestration.[12] A dysregulation of lung maturation is also suggested by the increased cell proliferation seen in CCAM.[13,14] The incidence of CCAM has been estimated at 1:25,000 to 1:35,000 pregnancies in one study,[15] while another reports a population prevalence of 9 per 100,000 total births.[16]

*Pulmonary sequestration* is generally thought to result from an abnormal accessory tracheobronchial bud arising from the foregut.[3,4] Intralobar (ILS) and extralobar (ELS) types are recognized, based on whether the visceral pleura is shared with the adjacent normal lobe or not. Typically, the lung tissue in sequestrations does not have a connection to the normal tracheobronchial tree and is supplied by an anomalous systemic artery, but many variants exist.[3,4,17] Most ILS are located in the lower lobes; most ELS are found postero-medially in the left lower chest but can occur within the diaphragm, below it or rarely in other locations.[18-20]

*Bronchogenic cysts* result from abnormal budding of the foregut. As foregut duplication cysts, they share common features with esophageal duplication cysts but are characterized by the presence of cartilage, smooth muscle and glands in their wall. The majority are located in the mediastinum, usually adjacent to the distal trachea or proximal mainstem bronchi, but they can also be found within the parenchyma of the lung.[1-4,11,21] They are usually unilocular, filled with fluid or mucus and generally do not communicate with the airway.

*Congenital lobar emphysema* (CLE) is a term used to describe a distended, hyperlucent lobe on plain radiographs, usually the left upper or the right middle lobe.[22] Pathologically, a distinction is made between a *polyalveolar lobe*, in which the number of alveoli is greatly increased, and *congenital lobar overinflation* (CLO), in which the alveoli are markedly distended. CLO is thought to be caused by a partial bronchial obstruction creating a ball-valve effect. This obstruction may be intrinsic (bronchomalacia) or less commonly extrinsic (vascular, bronchogenic cyst), but in many instances an exact cause cannot be determined.[1-4,11] The pathogenesis of polyalveolar lobe remains uncertain but transient bronchial obstruction in utero has been suggested.[23]

Although the terms CCAM, sequestration, bronchogenic cyst and CLE are entrenched in clinical usage and comfortably correspond to rigid pathological definitions, there is a considerable overlap in the findings. Numerous reports describe single lesions fulfilling the criteria for CCAM and ILS or ELS, coexisting lesions in different lobes, and lesions typical of one entity on imaging but corresponding to another entity pathologically (Figures 1 and 2).[11,17,20,24-31] Furthermore, the nomenclature and definitions have changed over time[6,7] and prominent pathologists disagree. For example, Stocker[4] believes that intrapulmonary bronchogenic cysts and other congenital unilocular lung cysts are type 1 CCAM while others disagree.[11] There have been attempts at revising the classification and pathogenesis of congenital lung malformations,[2,11,32] but the new nomenclatures proposed have either led to oversimplification[33] or have not gained wide acceptance. The emerging consensus is that imaging findings should simply be described, without attempting to make a pathological diagnosis. Adzick in 1985 first made the distinction between macrocystic and microcystic (hyperechoic) lung lesions on prenatal sonograms.[34] This is frequently interpreted as CCAM, but can be found in sequestration and other lung malformations or may represent a transient finding in normal babies.[15,16,29,31,35,36]

With the increasing number of obstetrical ultrasounds performed and the improving quality of these examinations, congenital lung malformations are diagnosed more frequently before birth. These appear as hyperechoic or cystic or mixed lesions within the chest (or abdomen for some sequestrations) that may displace the heart and mediastinum and occupy most of the thoracic cavity. Doppler interrogation may reveal a systemic arterial supply from the thoracic or abdominal aorta. This generally confirms a diagnosis of sequestration, although mixed lesions occur, as discussed earlier. While some congenital lung malformations may give rise to serious complications in utero, such as polyhydramnios with premature labor or hydrops with fetal demise, the majority remain stable or show evidence of regression.[15,16,26,29,37,38] Some newborns will develop symptoms related to pulmonary hypoplasia, left to right shunting (in a sequestration), overinflation (within a CCAM or a CLE) or pneumothorax.[39] However, most remain asymptomatic after birth and in many cases the chest radiograph (CXR) appears completely normal. Controversy thus exists as to the management of these newborns with asymptomatic congenital lung lesions discovered during routine obstetrical ultrasound examinations. The same is true for malformations discovered incidentally in infants or children, either by plain radiograph done for minor cold symptoms or by sonography, computed tomography (CT) or magnetic resonance imaging (MRI) done for unrelated reasons (Figures 3 and 4).

Based on our own experience[15,17,24,40] and an extensive review of the literature, we believe that elective resection is indicated for most congenital lung malformations. We will discuss the evidence on which this recommendation is based and explain which lesions may not require intervention.

## The arguments

Some authors recommend simple observation of patients with asymptomatic CCAM,[41-45] sequestration[46-48] or prenatally diagnosed "lung masses."[49] However, most authors favor surgical resection, at least for CCAM, intralobar sequestation and bronchogenic cysts.[2,3,7,15,24,29,31,39,50-69] Finally, others recommend surgery for "significant" lesions

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