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Perinatal management  
of a large bronchogenic cyst

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This male infant was born to a 30-year-old G3/P3 by spontaneous vaginal delivery at 41 6/7 weeks of pregnancy. At 19 weeks, a right-sided cystic thoracic malformation had been detected on prenatal ultrasound examination (Fig. 1). Subsequently, the woman was regularly followed to detect any fetal compromise, such as evidence of congestive heart failure or hydrops. At 39 0/7 weeks of gestation, the lesion measured 3x3 cm on an axial view and displaced the mediastinal structures to the left (Fig. 2).

The infant adapted without difficulties with Apgar scores of 9, 9 and 10 at 1, 5 and 10 minutes, respectively. There was no evidence of any respiratory distress and the baby was initially left with his parents on continuous pulse oxymetry monitoring. At the age of eight hours, ap and lateral X-ray views of the chest were obtained and confirmed the presence of a cystic lesion in the right hemithorax. An air-fluid level was easily recognized suggesting that the lesion was connected either to the bronchial system or the esophagus (Fig. 3). On echocardiography, there was a structurally normal heart and mild pulmonary arterial hypertension which normalized one week later.

Preoperatively, a CT scan of the chest was obtained. It demonstrated a cystic lesion in the right upper lobe with a connection to the bronchial system (Fig. 4, 5). On day of life 9, the infant was operated. The lesion was found to be a large cyst within the right upper

lobe (Fig. 6, 7). Following aspiration of air, it was successfully separated from the intact lung tissue with the use of a hot knife. The postoperative course was uneventful.

On histology, the cyst was lined with respiratory epithelium and its wall contained small areas of cartilage (Fig. 8-10). These findings are consistent with the clinical diagnosis of a bronchogenic cyst.

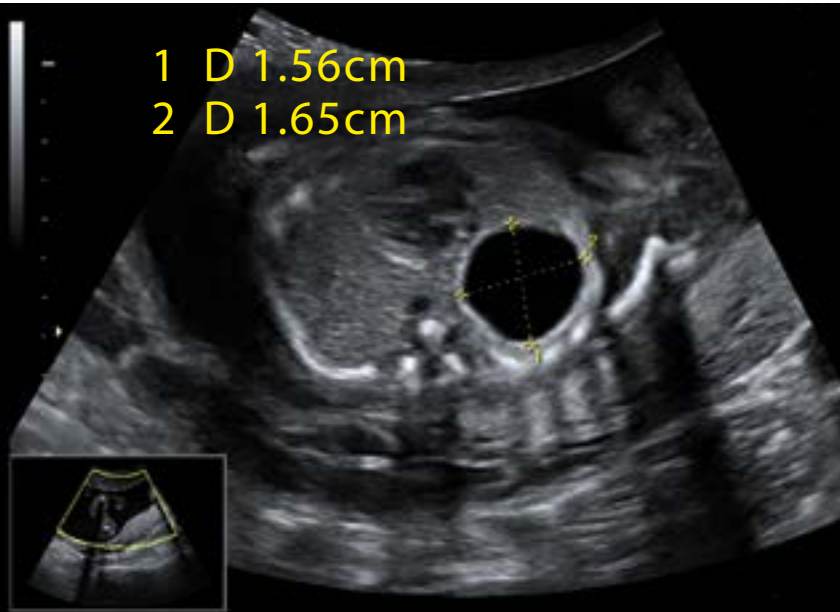


Fig. 1

Prenatal US at 19 0/7 weeks of pregnancy demonstrating a single right-sided thoracic cyst.

1 D 3.46cm  
2 D 3.26cm



Fig. 2

Prenatal US at 39 0/7 weeks of pregnancy demonstrating a single right-sided thoracic cyst.

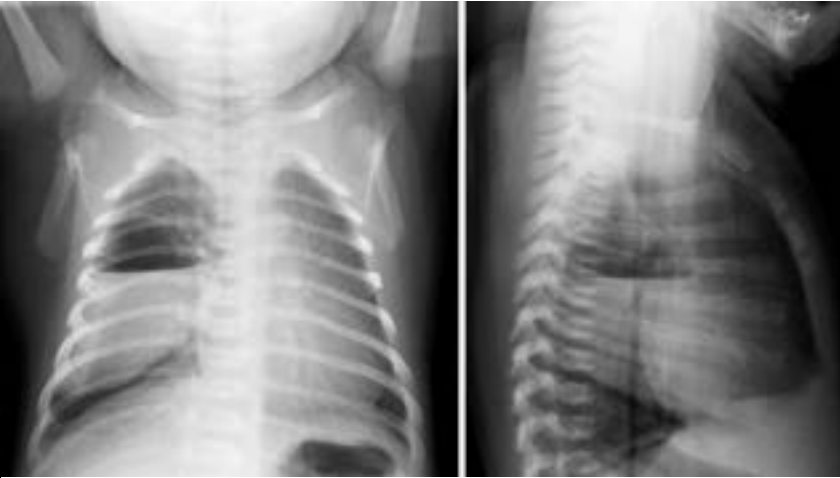
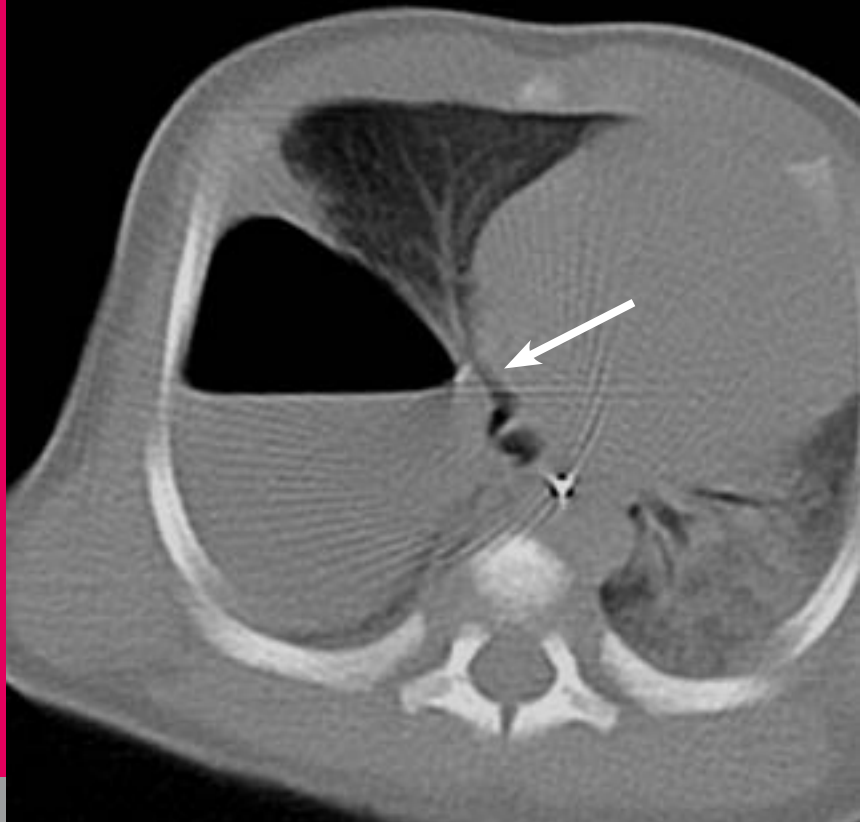


Fig. 3

CXR (ap and lateral views) on day 1 of life:  
right-sided dorsolateral cystic lesion with an air-fluid  
level.



**Fig. 4**

CT scan with contrast on day of life 4: large (4x5x4 cm) right-sided cyst with apparent connection to the bronchial system (arrow).



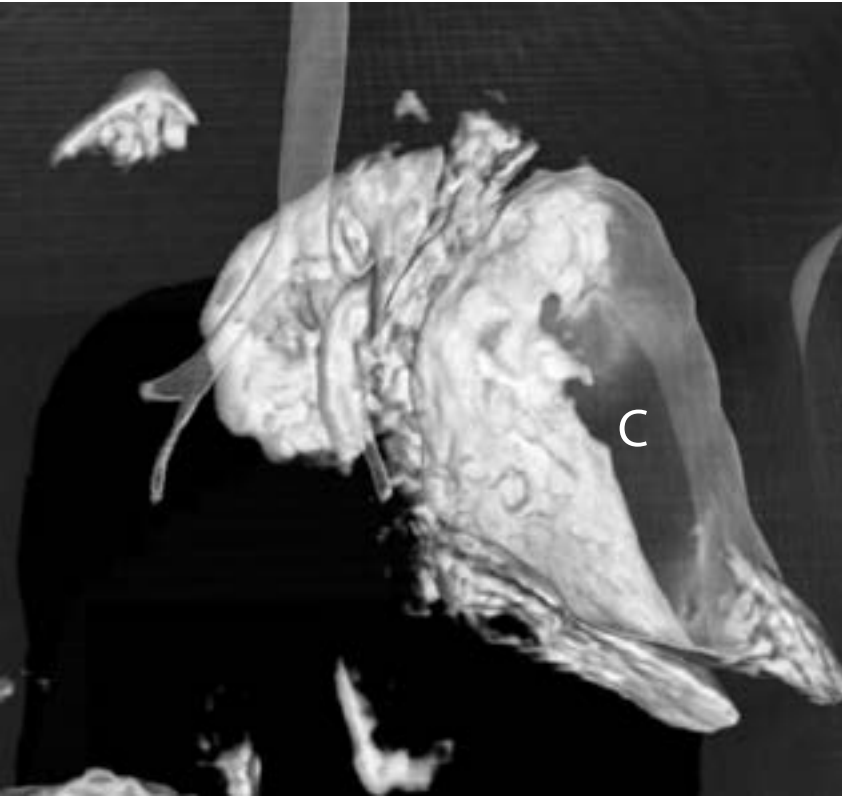


Fig. 5

CT scan with 3D reconstruction: the cyst appears hyperlucent (C) and is easily separated from the partially compressed white lung tissue.

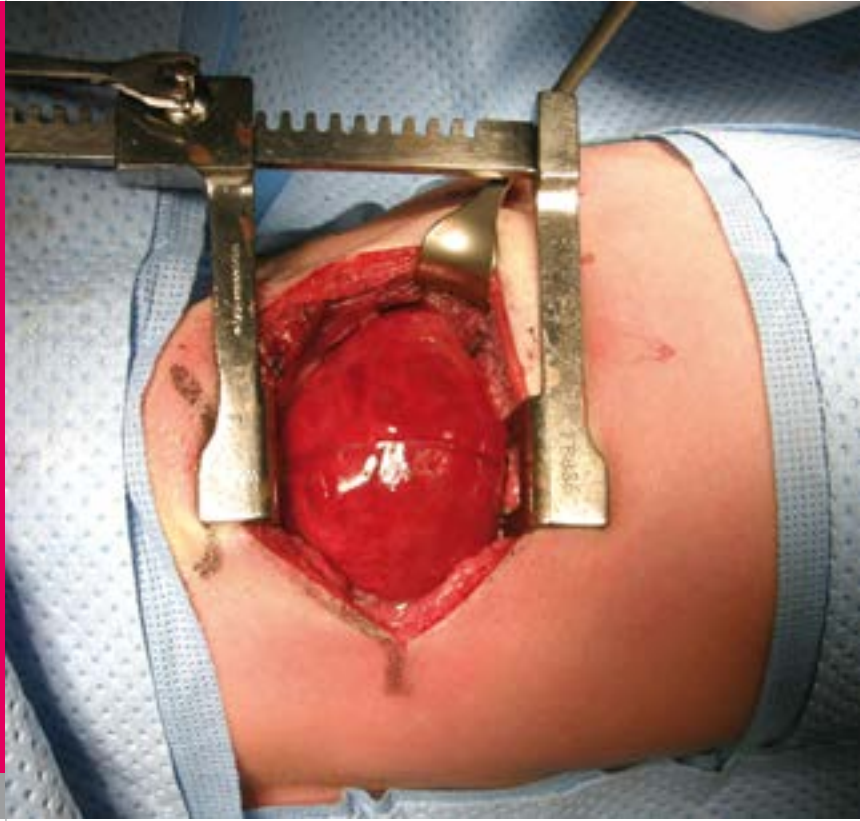


Fig. 6

Appearance of the cyst following opening of the chest through the 4th intercostal space.

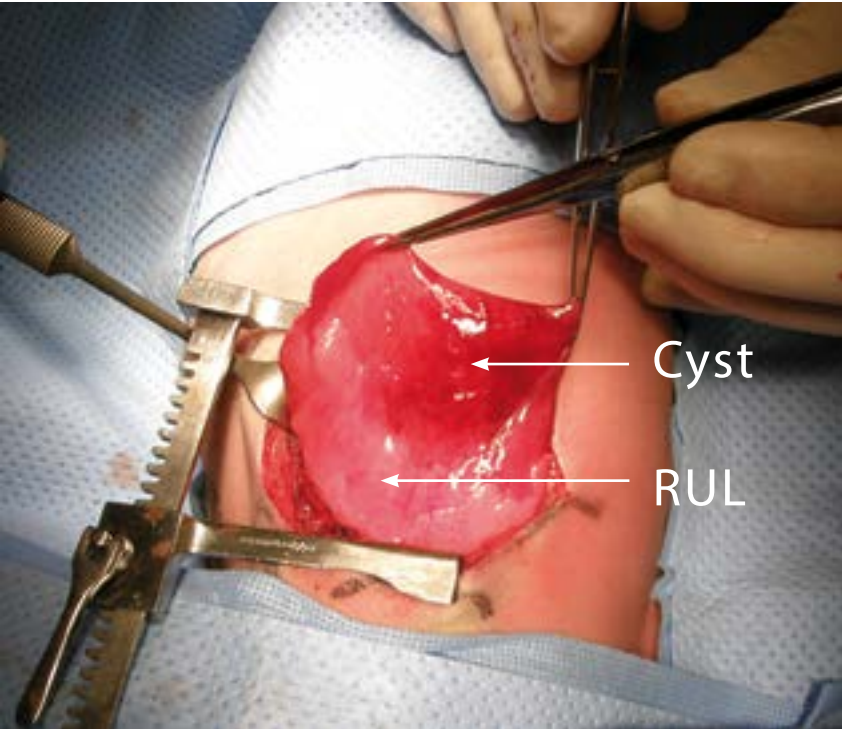
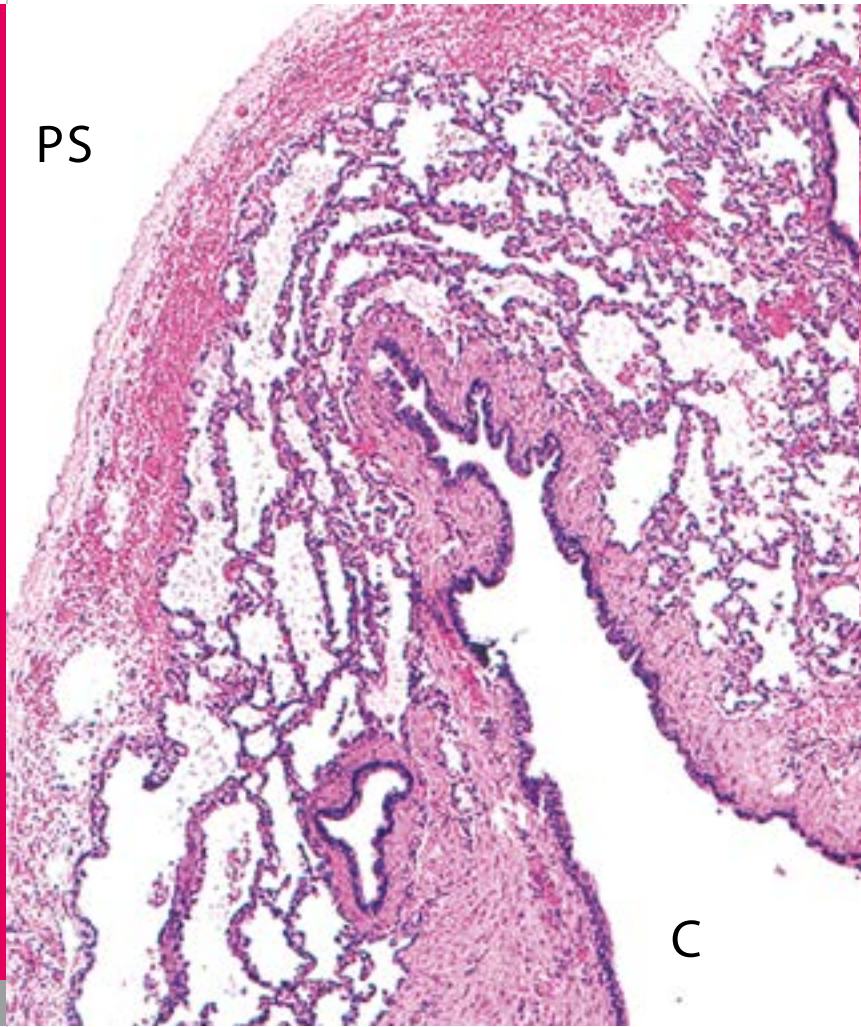


Fig. 7

Appearance of the cyst following aspiration of air: the cyst has collapsed and can now be delineated from the normally aerated part of the right upper lobe (RUL).

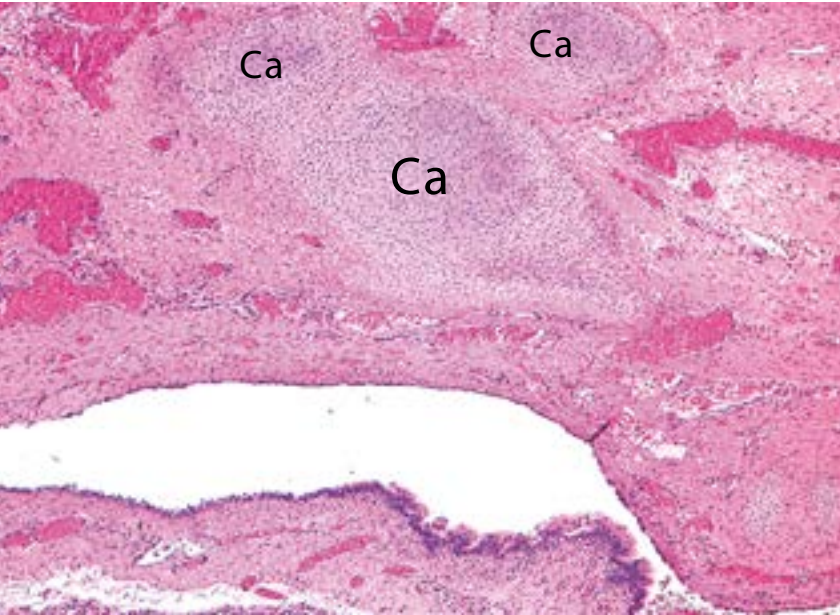
PS



C

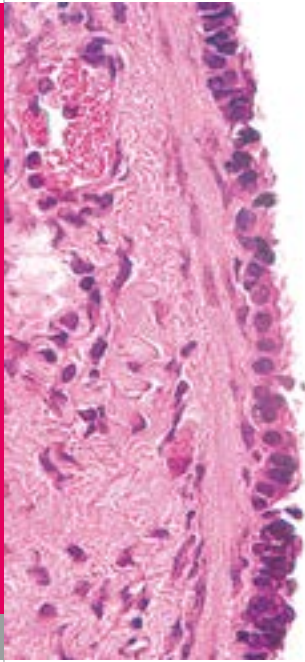
Fig. 8

Histology of the cyst (H&E stain): compressed lung tissue between pleural surface (PS) and cyst (C).



Histology of the cyst (H&E stain): islets of primitive cartilage (Ca) within the cyst wall.

Fig. 9



CL

**Fig. 10**

Histology of the cyst (H&E stain): higher magnification of the cyst wall with columnar respiratory epithelium (CL: lumen of cyst).

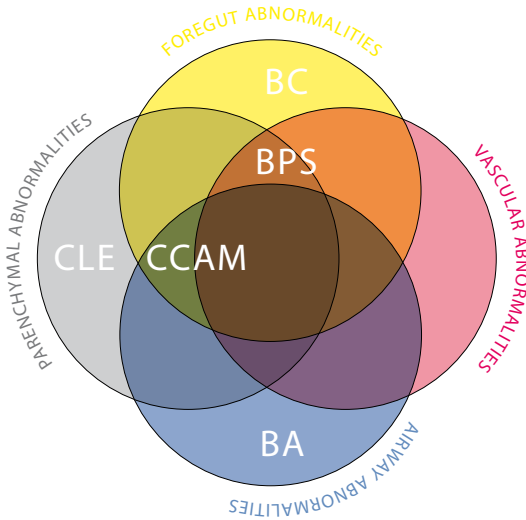


Fig. 11

Diagram of the spectrum of bronchopulmonary foregut malformations, including foregut, pulmonary, airway and vascular components (7) (BA: bronchial atresia; BC: bronchogenic cyst; BPS: bronchopulmonary sequestration; CCAM: congenital cystic adenomatoid malformation; CLE: congenital lobar emphysema).

New nomenclature	Old terms superseded
Congenital hyperlucent lobe (CHL)	Congenital lobar emphysema (CLE) Polyalveolar lobe
Congenital thoracic malformation (CTM)	Congenital cystic adenomatoid malformation (CCAM) Congenital pulmonary airway malformation Malinosculation Sequestration (intra- and extrapulmonary) Bronchogenic cyst Reduplication cyst Foregut cyst
Congenital small lung (CSL)	Pulmonary hypoplasia
Absent lung, absent trachea	Agenesis of lung, tracheal aplasia
Absent bronchus	Bronchial atresia

Table

Comparison of new nomenclature with old terms (1).



Congenital lung lesions are increasingly being recognized on prenatal ultrasound examinations. While they can be described as cystic, intermediate or solid, it is often not possible to make a more specific diagnosis. Some of these lesions disappear by the time the baby is born. Some infants are asymptomatic at birth, others have severe respiratory distress and may require emergency interventions. Postnatal imaging studies, including conventional X-ray, CT, MRI and ultrasound will help to better define the lesions and plan the appropriate surgical procedures. Finally, histological examination will allow a definitive description of the malformation.

Recently, several authors have argued that many of the terms used in the past to describe congenital lung malformations (e.g., congenital cystic adenomatoid malformation (CCAM), congenital lobar emphysema (CLE), bronchopulmonary sequestration (BPS), bronchogenic cyst) most likely describe extremes of a spectrum of malformations with the same underlying pathogenic mechanism (i.e., fetal airway obstruction) rather than separate disease entities (1, 2). In support of this concept, many authors have reported hybrid lesions with coexisting histological features of both CCAM and BPS (3, 4). Reviewing the final pathology reports of 25 patients who underwent surgical resection of prenatally diagnosed lung masses, Kunisaki et al. found that most congenital lung malformations were associated with an occult atretic bronchus (5).

Their observation suggests that fetal airway obstruction is likely to play a prominent role in the pathogenesis of congenital lung malformations (5).

Langston has proposed a malformation sequence to explain the spectrum of pulmonary anomalies, depending on the level, timing, and degree of bronchial obstruction. For example, an atretic bronchus early in gestation might favor the formation of a CCAM or a bronchogenic cyst, whereas obstruction later in gestation (e.g., 16-18 weeks of gestation) might predispose the lung to develop BPS or CLE (Fig. 11) (6, 7). Support for this theory also comes from experimental data that shows that cystic changes occur in fetal animal lungs after bronchial ligation. Newman has pointed out that this is also a common embryologic theme in other organ systems, such as the genitourinary system, with cystic renal dysplasia being a consequence of high-grade urinary obstruction (7).

Bush (1) has made a plea to replace the old terms by a simplified new nomenclature (Table). He recommends to use the following principles to classify congenital lung disease:

1. What is actually seen should be described, without indulgence in embryological speculation, which will almost certainly be proved wrong sooner or later.

2. The description should be in ordinary, everyday language.
3. The lung and associated organs should be approached in a systematic manner, because abnormalities are often multiple, and associated lesions will be missed unless carefully sought.
4. Clinical and pathological descriptions should be kept separate; the same clinical appearance (e.g., a multicystic mass) may have different pathological phenotypes.

The final pathological diagnosis in our patient was a bronchogenic cyst (Fig. 8-10). Bronchogenic cysts are thought to arise from abnormal buds from the primitive esophagus and tracheobronchial tree which do not extend to the site where alveolar differentiation occurs. Alternatively, they may be the consequence of bronchial obstruction early in gestation (5). In the majority of cases, they are located in the right paratracheal or carinal region, but intrapulmonary forms - as in our patient - have also been described (2). Depending on its location, a bronchogenic cyst may cause airway compression resulting in cough, wheeze, dyspnea, or even respiratory distress. Secondary infection of the cyst is a frequent complication. Malignant transformation of epithelial cells of bronchogenic cysts has been reported repeatedly (2).

Radiographic findings of bronchogenic cysts are variable and range from a rounded mass, with uniform

density similar to that of the cardiac shadow and projecting from the mediastinum, to hyperinflation or atelectasis of a lobe or an entire lung. As illustrated by our patient (Fig. 3, 4), when the lesion communicates with the esophagus or the tracheobronchial tree, air-fluid levels may be seen.

Because of the risk of complications, such as infection, hemorrhage, pneumothorax, sudden respiratory compromise and malignant transformation, most authors recommend elective resection of bronchogenic cysts. Histologically, the cyst wall has structural elements of the air way, including cartilage (as an obligatory finding to allow differentiation from an enteric duplication cyst), smooth muscle, mucous glands and respiratory epithelium (1).

1. Bush A. Congenital lung disease: a plea for clear thinking and clear nomenclature. *Pediatr Pulmonol* 2001;32:328-337
2. Eber E. Antenatal diagnosis of congenital thoracic malformations: early surgery, late surgery, or no surgery? *Sem Resp Crit Care Med* 2007;28:355-366
3. Samuel M, Burge DM. Management of antenatally diagnosed pulmonary sequestration associated with congenital cystic adenomatoid malformation. *Thorax* 1999;54:701-706
4. Roggin KK, Breuer CK, Carr SR, et al. The unpredictable character of congenital cystic lung lesions. *J Pediatr Surg* 2000;35:801-805
5. Kunisaki SM, Fauza DO, Nemes LP, et al. Bronchial atresia: the hidden pathology within a spectrum of prenatally diagnosed lung masses. *J Pediatr Surg* 2006;41:61-65
6. Langston C. New concepts in the pathology of congenital lung malformations. *Semin Pediatr Surg* 2003;12:17- 37
7. Newman B. Congenital bronchopulmonary foregut malformations: concepts and controversies. *Pediatr Radiol* 2006;36:773-791

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