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Cystic Lung Lesions in Newborns and Young Children: Differential Considerations and Imaging

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Numerous diverse entities produce cystic lung changes in neonates and young children. This review provides an evidence-based, age-appropriate, differential diagnostic framework to use when confronted with pulmonary cystic changes. The categories of diseases that have been discussed include congenital cystic bronchopulmonary malformations, neoplastic conditions, infections, collagen or soft tissue abnormalities, and mimics of cystic lung disease. An understanding of the pathophysiology, imaging appearance, and demographics of these entities is essential in guiding optimal care. Important educational points include differentiating bronchopulmonary malformations from neoplasms and the management and surveillance of lung cysts in young children.
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Introduction

The purpose of this review is to help the reader establish a framework for use when confronted with a cystic lung lesion(s) in an infant or young child (Tables 1 and 2). This discussion includes relevant, age-appropriate differential considerations using an evidence-based approach. The differentiation of bronchopulmonary malformations (BPMs) from neoplasms has been specifically addressed with discussion of the imaging, management, and surveillance of cystic lung lesions in young children.

Cystic lung lesions may contain either air or fluid or a combination of both, often with an air-fluid level(s). In this review, we attempt to distinguish between focal air trapping or emphysematous lung and actual cystic lung changes in which no internal lung markings are present, although the differentiation can be difficult.

Causes of cystic lung lesions in the immediate newborn period primarily consist of congenital cystic BPMs and occasional cystic neoplasms, whereas the differential diagnosis is broader in infants and young children, with the addition of infectious lesions, cystic changes associated with collagen or connective tissue abnormalities, and vasculitic lesions and pulmonary fibrosis occasionally (Table 1). In both age groups, there are a number of potential pitfalls—entities that can mimic or be mistaken for cystic lung changes (Tables 1 and 2).

Congenital Cystic BPMs

The cystic congenital lung lesions encountered include bronchogenic cyst, congenital pulmonary airway malformation (CPAM) and pulmonary sequestration (Figs. 1-4). Congenital lobar overinflation and bronchial atresia are additional congenital BPMs that are encountered; they are not truly cystic lesions but have regional or focal overinflated lung that may mimic cystic lung changes. Often, overlap between the various congenital lung lesions is observed on both imaging and pathology, with frequent hybrid or overlap lesions containing elements of more than 1 entity. This is particularly seen with a combination of CPAM and sequestration occurring in approximately 50% of lesions (Fig. 4).[1] Most congenital lung lesions are thought to be caused by in utero airway obstruction, often with an aberrant, stenotic, or atretic supplying airway.[2] This unifying theory of an obstructive malformation complex with varying dysplastic lung manifestations related to the degree and timing of obstruction in utero[3] helps to explain the considerable overlapping features of the various lesions. Many of

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Table 1 Causes of Pulmonary Cystic Changes in the Neonate and Young Child

Bronchopulmonary malformations
Bronchogenic cyst
Congenital pulmonary airway malformation
Pulmonary sequestration
Pulmonary neoplasms
Lymphangioma
Langerhans cell histiocytosis
Respiratory papillomatosis
Pleuropulmonary blastoma
Peribronchial myofibroblastic tumor
Fetal lung interstitial tumor
Lymphoma and cavitating metastases
Infections/inflammatory
Pneumatocele
Cavitary necrosis
Abscess
Septic emboli
Specific: staphylococcus, streptococcus, TB, cocidiodomycosis, aspergillus, hydatid, and pneumocystis
Vasculitic
Granulomatosis with polyangiitis and collagen vascular disease (very uncommon in young children)
Collagenopathies, connective tissue disorders, and syndromes
Down syndrome
Pulmonary fibrosis or hypoplasia
Mimics of cystic lung changes
Air leak including pneumomediastinum or pneumothorax and PIE
Bronchopleural fistula
Focal emphysema: BPD, CLO, bronchial atresia, bronchiolitis obliterans, and other vascular and lung growth abnormalities
Bronchiectasis (rare in age group)
Loculated pleural fluid
Diaphragmatic and paraesophageal hernia
Trauma—air leak, cyst, hemorrhage, and infarct
Artifact

BPD, bronchopulmonary dysplasia; CLO, congenital lobar overinflation.

these lesions are initially discovered by ultrasound (US) screening in utero, whereas in the past they may have presented with respiratory distress at birth or due to secondary infection or mass effect on adjacent structures or as an incidental, often asymptomatic finding. Prenatally, these lesions can be further defined and evaluated by magnetic resonance imaging (MRI), which usually allows postnatal computed tomographic (CT) imaging to be deferred to a later age if the child is asymptomatic at birth.

Typically, the congenital lung lesions are largest in size in the second trimester and tend to decrease in size or remain stable in the third trimester. They may be more difficult to appreciate on imaging later in pregnancy because of decreased relative or absolute size and mass effect and more similar echogenicity (US) or intensity (MR) compared to normal lung (Fig. 2). Large or rapidly expanding lesions may cause hydrops fetalis,[1] related to central venous compression by the lesion, thereby producing congestive failure; these cases typically require emergency intervention and are associated with a high morbidity and mortality. When radiography is performed at or soon after birth, BPM lesions are usually fluid filled, dense opacities that mimic a solid mass, and there is delayed clearance of fetal lung fluid and replacement with air, which is likely related to an obstructed or abnormal airway connection[3] (Fig. 2). Many lesions discovered in utero are either not visible or produce only subtle radiographic abnormalities at birth, although they are usually readily identified by CT imaging[4,5] (Fig. 4). We strongly advocate CT angiography rather than routine chest CT for investigation of suspected congenital lung malformations postnatally, and the imaging should extend inferiorly to include the celiac axis to identify any infradiaphragmatic feeding arteries. Because of considerable overlap among the lesions, aberrant systemic vessels may be present in a variety of pathologic entities and can readily be missed if small. Additionally, thin-section angiographic spiral imaging allows for high-quality multiplanar and 3-dimensional reformations, which may be very useful in defining the arterial supply and venous drainage of the lesion as well as providing anatomical details of specific features including cysts, airway narrowing, bronchial mucoid impaction, air trapping, and parenchymal abnormality.[5] Postnatal US may also be useful, especially soon after birth when the lesion is likely to be fluid filled (Fig. 4). MRI is often employed prenatally, but it is generally used less often than other modalities postnatally because of relative inability to see lung parenchyma well, especially air-filled structures. US and MRI are the modalities of choice for evaluating suspected abdominal BPMs and even mediastinal lesions. MRI has been used to follow up some cases