Imaging of congenital lung malformations in infants and children

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Learning objectives

- To describe and illustrate the principal congenital lung malformations.
- To provide a concise radiologic approach to congenital lung malformations.
Background

- Congenital lung malformations are uncommon: an incidence of around 1 in 5000 to 10000 births have been suggested; only 10% of cases are identified at birth, whereas 14% are diagnosed later by 15 years of age.
- May lead to considerable morbidity and mortality.
- Radiologic appearance will change depending on the type. Plain radiograph and CT scan are the primary tool for imaging.
- Congenital lung malformations include:

  Congenital cystic adenomatoid malformation (CCAM)
  - Broncho pulmonary sequestration
  - Congenital lobar emphysema (CLE)
  - Bronchogenic cysts.
  - Pulmonary agenesis and hypoplasia
  - Polyalveolar lobe
  - Alveolocapillary dysplasia
  - Arteriovenous malformation
  - Pulmonary lymphangiectasis

- Congenital lung lesions result from perturbations in lung and airway embryogenesis.
- The diagnosis of these conditions may sometimes be made by prenatal ultrasonography but usually the lung malformation is discovered after birth.
- Clinical symptoms: Respiratory symptoms in newborn or infant; Pneumonia that may respond poorly to medical treatment (most common complication); Development of malignancies (carcinomas and pleuro-pulmonary blastomas); Pneumothorax; Hemoptysis or hemothorax. Most likely asymptomatic and discovered incidentally on thoracic imaging.
Findings and procedure details

Procedures details:

We retrospectively reviewed the imaging findings of congenital lung abnormalities from our departmental archives. Different imaging modalities were used including plain radiograph, CT scan and prenatal ultrasound.

**Congenital lobar emphysema (CLE)** is a developmental anomaly of the lower respiratory tract that is characterized by hyperinflation of one or more of the pulmonary lobes. Other term for CLE include congenital lobar over inflation and infantile lobar emphysema. The most frequently identified cause of congenital lobar emphysema (CLE) is obstruction of the developing airway. This leads to the creation of a "ball-valve" mechanism in which a greater volume of air enters the affected lobe during inspiration than leaves during expiration, producing air trapping. Most common location: left upper lobe (40-45%); occasionally, more than one lobe. The emphysema can compress the adjacent healthy lobe. Herniation of the emphysematous lobe through the mediastinum and bronchial cartilaginous dysplasia are often associated.

**Imaging finding:**

**Chest radiograph:** Immediate post-partum period: The affected lobe tends to appear opaque and homogeneous because of fetal lung fluid or it may show a diffuse reticular pattern that represents distended lymphatic channels filled with fetal lung fluid.

Later findings (fig.1) : appears as an area of hyperlucency in the lung with a paucity of vessels, mass effect with mediastinal shift and hemidiaphragmatic depression, decubitus films lying on the affected side will show little or no change in lung volume, lateral film may demonstrate posterior displacement of the heart.

**CT scan** (fig.2): shows more features in greater detail; attenuation of vascular structures in affected lobe; May also show compressive atelectasis of neighbouring lobes.

**Treatment:** Left upper lobectomy (fig.3)

**Congenital cystic adenomatoid malformation (CCAM):**

Also known as congenital pulmonary airway malformation (CPAM), it is a ccongenital anomaly arising from the abnormal proliferation of the bronchial elements. There is 3 histologic types (Stoker et al):
• Type I (most common): variable-size cysts, with sometimes at least one dominant cyst (>2 cm in diameter).
• Type II: smaller, more uniform cysts less than 1 cm in diameter.
• Type III: solid mass composed of Broncho alveolar micro cysts.

Imaging finding:

Antenatal: solid mass within fetal chest with maternal hydramnios and fetal anasarca.

Postnatal: Chest radiograph (fig.4): First investigation in a neonate with respiratory distress, it showed multiple air filled cystic spaces usually in a single lobe with normal lung interposed between the cysts. CT scan (fig.5): is conclusive by demonstration of air filled cystic spaces with and between normal lung parenchyma. The cyst may contain air fluid level (fig.6 and 7) and more other smaller cysts (fig8).

Differential diagnosis of classical CCAM: Congenital diaphragmatic hernia, staphylococcal pneumonia with pneumatocele formation, congenital lobar emphysema in case of a large unilocular cyst, bronchogenic cyst, sequestration, pneumothorax.

Sometimes in the early neonatal period the lesion is not air filled and may mimic airspace disease. However, subsequently fluid is replaced by air producing classical cystic.

**Intra pulmonary bronchogenic cyst:**

Commonly located in the mediastinum (70-90%) or in lung parenchyma. It arise from abnormal budding of the primitive tracheobronchial tube. The location depends on the embryological stage of development at which the anomaly occurs: early development (the cyst tends to be located along the tracheobronchial tree); later (cysts are more peripheral and may be located in the lung parenchyma). Bronchogenic cyst is usually asymptomatic and presents as an incidental finding on the chest roentgenogram. Diagnostic studies, include chest CT scan, magnetic resonance imaging, and definitive tissue diagnosis can only be established by means of surgical excision.

Imaging finding:

Standard chest radiographs (fig.9): most presented as homogeneous water density shadows, air-fluid levels can be also seen. Computed tomography (fig.10): round, well circumscribed, unilocular or multilocular mass with density ranging from water density. Occasionally computed tomography showed an air-filled cyst (fig.11).

Lung cysts often include a cartlaginous wall (fig.12).

**Pulmonary sequestration:**
It is a dysplastic, non-functioning lung parenchyma, there is no communication with the tracheobronchial tree. PS has an anomalous systemic arterial supply, usually from the thoracic or abdominal aorta (from the celiac, splenic, intercostal, subclavian, or even coronary arteries is less common). Most common location is within the left lower lobe.

Sequestrations can be (fig.13) : Intralobar in 75% (within visceral pleura and venous drainage via the inferior pulmonary vein). Extra lobar in 25% (separate pleural covering with venous drainage usually via systemic veins, typically the azygous vein and less commonly via the portal, left subclavian, or internal mammary veins). Many variations and combinations of these classical patterns have been described.

Intralobar PS: diagnosis usually made in adolescence or adulthood, presenting with recurrent pneumonia.

Extralobar PS: often asymptomatic but discovered during evaluation of other anomalies in neonantes or infants.

Imaging findings: appear as a persistent opacity or mass; may be associated with congenital pulmonary airway malformation (CPAM), in which case air may be present within the lesion. It can be infected.

Plain radiographs (fig.14): Single homogeneous opacity in the base of one lung; less commonly, a cystic mass.

**CT angiogram**: Establish the diagnosis by showing both abnormal lung parenchyma (fig.15) and the systemic arterial supply (fig.20).

Surgical treatment: ligature of the systemic artery + lobectomy for the intralobar PS; resection of the PS for extralobar form (fig.16).

**Pulmonary arterio venous malformation:**

Anomalous communications between arteries and veins that bypass capillary system. 70 % are located in the lower lobes. Lesions less than 2 cm are usually asymptomatic. MDCT technique with high quality 3D images (VR) provides angiographic like images of the pulmonary vasculature, complete picture of the malformation, including information on the feeding and draining vessels (fig.17 and 18).

**Pulmonary agenesis and hypoplasia:**

Incomplete development of lung tissue; very rare; Severity of the lesion depends on the appearance time of the malformation during the timeline of lung development. (fig.19)
Images for this section:

![Chest x-ray image](image_url)

**Fig. 1:** Chest x-ray show marked overdistention of the upper left lobe with mediastinal shift and partial collapse of the contralateral remaining lung field.

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Fig. 2: CT scan shows a hyperlucent, hyperextended lobe with midline substantial herniation and compression of the remaining lung. The mediastinum is shift away from the side of abnormal lobe.

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Fig. 3: Left upper lobectomy. Photograph of the resected specimen

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**Fig. 4:** Chest radiograph shows air filled, multi-cystic lesion of the right lower lobe

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Fig. 5: Axial CT section, lung window reveals multiple air filled cysts in the right lower lobe with normal lung parenchyma in between
Fig. 6: Chest radiograph: Upper right lobe cyst with air fluid level
Fig. 7: CT scan show cystic lesion with air-fluid level and lower lobe alveolar condensation.

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Fig. 8: Photograph of the resected specimen showing a large cyst wall lined by necrotic debris (arrow), along with multiple small cysts.

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**Fig. 9:** Chest radiograph: upper right lobe opacity with air fluid level (arrow).

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**Fig. 10:** CT scan: right Bronchogenic cyst.

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Fig. 11: CT scan: unilocular air-filled bronchogenic cyst of the right lower lobe.
Fig. 12: Lung parenchymal bronchogenic cyst; histological view: Respiratory epithelium (arrows).

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Fig. 13: schematic view of the two types of PS.

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Fig. 14: Chest radiograph: retrocardiac excaved opacity.

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Fig. 15: CT scan of the chest: Extralobar PS.

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Fig. 16: Extralobar PS: ligature of the systemic artery (arrow) before resection.

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**Fig. 17:** CT scan of the chest: right laterobasal opacity.

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Fig. 18: CT scan angiogram: Pulmonary Arteriovenous Malformation.

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Fig. 19: Unilateral left pulmonary hypoplasia.

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Fig. 20: Systemic artery from aorta (arrow) with cystic pulmonary sequestration (large arrow).
Conclusion

Although congenital lung malformations are rare, they are important disorders because they may lead to considerable morbidity and mortality. The radiologic examination should give the clinical information required, be noninvasive and use the lowest radiation dose possible. Early diagnosis and prompt surgical treatment offer the possibility of absolutely normal lung development.
References