Congenital lung anomalies - spectrum of imaging findings

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Learning objectives

- We aim to review the radiographic and CT imaging findings of some congenital lung diseases.

- We will divide this review in parenchymal anomalies (congenital pulmonary airway malformations; congenital lobar hyperinflation; congenital bronchial atresia and bronchogenic cyst) and combination of parenchymal and vascular anomalies (pulmonary sequestration and hypogenetic lung syndrome).
Background

Congenital lung anomalies are a heterogeneous group of developmental disorders with a wide distribution in imaging appearance and clinical manifestations.

At present, these anomalies have an annual incidence of 30 - 42 cases per 100,000.

In this review we divide them into those with pure parenchymal anomalies and those with both parenchymal and vascular manifestations.

Parenchymal anomalies

**Congenital pulmonary airway malformation (CPAM)**

- Congenital pulmonary airway malformations are a heterogeneous group of congenital cystic and noncystic lung lesions characterized by extensive overgrowth of the primary bronchioles, which are in communication with abnormal bronchial tree lacking cartilage.

- They are the most common congenital pulmonary anomaly accounting for 30 - 40% of cases.

- The most recent classification system divides CPAM into five types (0 - 4), with type classification based on cyst size and histologic similarities to the bronchial tree and airspace.

- The previous denomination (congenital cystic adenomatoid malformation) was abandoned because these lesions are cystic only in three of the five types (1, 2 and 4) and only one type has adenomatoid changes (type 3).

- Affected patients usually present with respiratory distress or infection between the neonatal period and 2 years age.

**Congenital lobar hyperinflation (CLH)**
- Congenital lobar hyperinflation, formerly known as congenital lobar emphysema, manifests as hyperinflation and distension of one or multiple pulmonary lobes. It results from air trapping caused by intrinsic or extrinsic bronchial luminal narrowing.

- There are two types of CLH based on alveolar number at histologic analysis - the hypoalveolar form has fewer than the normally expected number of alveoli, with associated hyperinflation of the individual alveoli; in the polyalveolar form there is an increase in the number of alveoli in the affected segment, with individual alveoli being normally inflated.

- Patient with CHL usually present during the neonatal period and infancy with respiratory distress, especially if there is marked hyperinflation in the involved lobe and mass effect on adjacent lung parenchyma or mediastinum.

**Congenital bronchial atresia**

- Congenital bronchial atresia results from focal obliteration of a proximal segmental or subsegmental bronchus that lacks communication with the central airways. The development of distal structures is normal.

- The bronchi distal to the stenosis become filled with mucus and form a bronchocele. The alveoli supplied by these bronchi are ventilated by collateral pathways and show features of air trapping. The upper lobe bronchi are more frequently involved.

- Although affected patients may present with recurrent infections or respiratory compromise, the majority of cases are incidentally detected on chest radiographs or CT obtained for other indications.

**Bronchogenic cyst**

- Bronchogenic cysts are developmental lesions resulting from abnormal ventral budding or branching of the tracheobronchial tree.

- They are a part of the spectrum of foregut duplication cysts that include bronchogenic, enteric and neurenteric cysts. In practice distinction between these entities is artificial.
- Although they are typically located within the mediastinum (70% of cases) near the subcarinal, hilar or right paratracheal regions, they can also occur in lung parenchyma, with the majority of them located in lower lobes.

- Mediastinal bronchogenic cysts usually do not communicate with the adjacent bronchial tree; intrapulmonary lesions often do.

- When they are small, affected patients are usually asymptomatic; when they are large enough to compress adjacent mediastinal structures, symptoms such as chest pain, respiratory distress or dysphagia may occur.

**Combination of parenchymal and vascular anomalies**

**Pulmonary sequestration**

- Pulmonary sequestration consists of a portion of nonfunctioning lung tissue which does not communicate with the adjacent tracheobronchial tree and receives systemic arterial blood supply.

- Based on the presence of pleural investment and venous drainage, it has been divided into two types: intralobar and extralobar.

- **Intralobar type** represents 75% of cases. It refers to a segment (or segments) of lung that has a systemic arterial supply and is invested by the same visceral pleura that covers the remainder of the lung. Most are located in the posterolateral segment of a lower lobe. The anomalous artery typically arises from the descending aorta while the anomalous vein drains into the left atrium via the inferior pulmonary vein. It often presents in older children or adults, with recurrent infections. It is seldom associated with other congenital anomalies.

- **Extralobar type** a portion of lung is invested by its own visceral pleura, separated from the rest of the lung. It has a systemic arterial supply and usually systemic venous drainage. The anomalous artery typically arises from the descending aorta or its branches such as celiac axis, while the anomalous vein drains into various systemic veins including the azygos, the portal, the subclavian or the internal mammary veins. Affected patients are usually infants who present during the neonatal period with lung masses, typically in the left lower thorax. Other less common locations include mediastinum, pericardium.
and within or below the diaphragm. Extralobar sequestration is frequently associated with other congenital anomalies such as diaphragmatic defect and congenital heart disease.

**Hypogenetic lung syndrome**

- Hypogenetic lung syndrome is a rare congenital anomaly consisting of hypoplasia of the right lung and pulmonary artery, associated dextroposition of the heart, ipsilateral partial anomalous pulmonary vein drainage and anomalous systemic arterial supply to the right lung. Most often the anomalous pulmonary vein drains into the inferior vena cava. Given the anomalous pulmonary venous drainage, there is a subsequent left-to-right shunt.

- Clinical presentation of hypogenetic lung syndrome is variable depending on the age of the patient. Young infants usually present with symptoms of congestive heart failure secondary to left-to-right shunt, while older children may present with recurrent right basilar pulmonary infections. However it may remain asymptomatic throughout life and be incidentally detected.

- Other congenital anomalies are seen in up to 25% of patients including atrial and ventricular defects, patent *ductus arteriosus*, tetralogy of Fallot, diaphragmatic abnormalities and pulmonary sequestration.
Findings and procedure details

Parenchymal anomalies

**Congenital pulmonary airway malformation (CPAM)**

- Congenital pulmonary airway malformations demonstrate a variety of imaging appearances based on the type and the presence or absence of associated superimposed infection.

- They usually appear as one or more air-filled "cystic" structures. Solid areas may be present.

- Type 0 is usually not imaged because it is incompatible with life.

- Type 1 is composed of variable-size cysts, with at least one dominant lesion (> 2 cm). It is the most common form (Fig. 1 on page 26 and Fig. 2 on page 26).

**Fig. 1:** Type 1 congenital pulmonary airway malformation. Axial lung window CT images show a macrocystic mass (*) in the left lower lobe. There are other smaller cysts adjacent to the dominant cyst (arrow). Given the cyst size, findings are consistent with a type 1 CPAM.

**References:** Serviço de Imagem Médica, Centro Hospitalar e Universitário de Coimbra - Hospital Pediátrico, Portugal
Fig. 2: Type 1 congenital pulmonary airway malformation. Axial lung window CT images demonstrate multiple cystic lesions in the left upper lobe, with a dominant cyst measuring more than 2 cm (*). Mediastinal shift to the right side is also seen.

References: Serviço de Imagem Médica, Centro Hospitalar e Universitário de Coimbra - Hospital Pediátrico, Portugal

- Type 2 has numerous small cysts of uniform size, each one measuring less than 2 cm in diameter (Fig. 3 on page 27 and Fig. 4 on page 27).
Fig. 3: Type 2 congenital pulmonary airway malformation. Axial lung window CT images demonstrate an air-filled multicystic mass in the right upper lobe. All cysts measure less than 2 cm which is consistent with a type 2 CPAM.

References: Serviço de Imagem Médica, Centro Hospitalar e Universitário de Coimbra - Hospital Pediátrico, Portugal
**Fig. 4:** Type 2 congenital pulmonary airway malformation. Axial lung window CT images show multiple cystic lesions in the right lower lobe. All cysts measure less than 2 cm which is consistent with a type 2 CPAM.

**References:** Serviço de Imagem Médica, Centro Hospitalar e Universitário de Coimbra - Hospital Pediátrico, Portugal

- Type 3 appears solid at imaging; however, at histologic evaluation, it is characterized by numerous microcysts.

- Type 4 usually presents as a large cystic lesion and is indistinguishable from type 1 in imaging studies.

- In patients with infected CPAMs, associated findings of internal air-fluid levels and enhancing thick wall are often seen (**Fig. 5** on page 28).

**Fig. 5:** Infected type 1 congenital pulmonary airway malformation. Axial lung window CT images demonstrate large cystic lesions in the left lower lobe, with air-fluid levels (arrows). Mediastinal shift to the right side is also seen.

**References:** Serviço de Imagem Médica, Centro Hospitalar e Universitário de Coimbra - Hospital Pediátrico, Portugal

**Congenital lobar hyperinflation (CLH)**

- Congenital lobar hyperinflation may initially appear as an opacity on radiographs owing to accentuated retention of fetal lung fluid just after birth. As fetal lung fluid is cleared and
replaced by air, the affected lobe generally shows hyperlucency with mass effect on the adjacent, non affected lobes (Fig. 6 on page 28 and Fig. 7 on page 29).

**Fig. 6**: Congenital lobar hyperinflation. Anteroposterior (a) and lateral (b) chest radiographs show a hyperlucent left upper lobe that herniates to the right (arrow), with displacement of mediastinal structures to the right.

**References**: Serviço de Imagem Médica, Centro Hospitalar e Universitário de Coimbra - Hospital Pediátrico, Portugal
**Fig. 7**: Congenital lobar hyperinflation. Frontal chest radiograph shows hyperinflation of the middle lobe (arrow).

**References**: Serviço de Imagem Médica, Centro Hospitalar e Universitário de Coimbra - Hospital Pediátrico, Portugal

- On CT images, a hyperinflated lobe with attenuated and displaced pulmonary vessels is typically seen. Adjacent lobes and structures may be compressed and sometimes ipsilateral or contralateral atelectasis may occur. Mediastinal shift to the contralateral side, and separated ribs and depressed diaphragm on the ipsilateral side can also be seen (Fig. 8 on page 30, Fig. 9 on page 31 and Fig. 10 on page 31).
**Fig. 8**: Congenital lobar hyperinflation (same case as in fig. 6). Axial lung window CT images show marked hyperinflation of the left upper which is herniated to the right (arrow). There is also mediastinal shift to the right.

**References**: Serviço de Imagem Médica, Centro Hospitalar e Universitário de Coimbra - Hospital Pediátrico, Portugal

**Fig. 9**: Congenital lobar hyperinflation (same case in figs. 6 and 8). Coronal lung window CT images show marked hyperinflation of the left upper lobe with attenuated lung markings and mediastinal shift to the right side.

**References**: Serviço de Imagem Médica, Centro Hospitalar e Universitário de Coimbra - Hospital Pediátrico, Portugal
Fig. 10: Congenital lobar hyperinflation (same case as in fig.7). Axial lung window CT images show hyperinflation of the middle lobe with attenuated lung markings (*).

References: Serviço de Imagem Médica, Centro Hospitalar e Universitário de Coimbra - Hospital Pediátrico, Portugal

- Although any lobe can be affected, there is a lobar predilection - the left upper lobe is the most frequently affected (42%), followed by middle lobe (35%) and right upper lobe (21%). Occasionally, only a portion of a lobe or more than one lobe may be involved.

**Congenital bronchial atresia**

- On chest radiography, impacted mucus distal to the atretic segment of bronchus appears as a round or oval opacity. Distal air-trapping appears as focal hyperlucency of the lung distal to the area of mucus plugging.

- CT accurately demonstrates the mucus-filled bronchus distal to the area of atresia as a tubular soft tissue opacity. Distal to the mucus-filled bronchus, there is lung hyperinflation which appears as a segmental area of hypoattenuation (Fig. 11 on page 32, Fig. 12 on page 32 and Fig. 13 on page 33).
Fig. 11: Congenital bronchial atresia. Lung window CT images show a dilated tubular-shaped opacity representing a bronchocele (arrow) in the left lower lobe with adjacent segmental hypoattenuation due to air trapping (*).

References: Serviço de Imagem Médica, Centro Hospitalar e Universitário de Coimbra, Portugal

Fig. 12: Congenital bronchial atresia (same case as in fig.11). Axial non-contrast (a) and contrast-enhanced (b) CT images demonstrate a tubular-shaped, soft tissue opacity representing a bronchocele (circle) in the left lower lobe. It shows no enhancement.

References: Serviço de Imagem Médica, Centro Hospitalar e Universitário de Coimbra, Portugal
Fig. 13: Congenital bronchial atresia (same case as in figs. 11 and 12). Coronal lung window (a) and soft tissue window (b) CT images show the dilated mucus-filled bronchus in the left lower lobe, surrounded by lung hypoattenuation due to air trapping (circle).

References: Serviço de Imagem Médica, Centro Hospitalar e Universitário de Coimbra, Portugal

**Bronchogenic cyst**

- On chest radiography bronchogenic cysts typically present as well-circumscribed, oval or round, soft tissue densities, most frequently in the subcarinal region (Fig. 14 on page 33a).

- On CT studies they are usually well-defined, smooth in contour, rounded lesions. About 50% of them show homogeneous water attenuation but the degree of attenuation depends on the amount of intracystic proteinaceous contents. After administration of intravenous contrast, bronchogenic cysts show no enhancement or a minimally enhancing wall (Fig. 14 on page 33 and Fig. 15 on page 34). Calcifications and septations are occasionally present.

- If infected, bronchogenic cysts may show thickening of their walls and air-fluid levels.
Fig. 14: Bronchogenic cyst. (a) Chest radiograph shows a large soft-tissue mass in the right hemithorax (arrow) which in coronal contrast-enhanced CT image (b) appears as a smooth, well-defined mass with water density (arrow).

References: Serviço de Imagem Médica, Centro Hospitalar e Universitário de Coimbra - Hospital Pediátrico, Portugal

Fig. 15: Bronchogenic cyst (same case as in fig. 14). Non-contrast (a) and contrast-enhanced (b) CT scan through the upper lobes show a well-defined water-density homogeneous mass with no contrast enhancement. The mass causes mediastinal shift to the left side and airway compression.

References: Serviço de Imagem Médica, Centro Hospitalar e Universitário de Coimbra - Hospital Pediátrico, Portugal

Combination of parenchymal and vascular anomalies
**Pulmonary sequestration**

- Imaging findings of pulmonary sequestration depend on the type, associated superimposed infection and the concomitant presence of other congenital pulmonary malformations.

- On chest radiography, pulmonary sequestrations appear as focal lung masses within the lower lobes, most commonly on the left. The aberrant systemic artery may also be evident on radiographs.

- MDCT angiography with 3D reconstructions can be particularly helpful in evaluating sequestrations. CT characteristics of the mass range from a heterogeneously enhancing solid mass to a complex cystic lesion, with air-fluid levels.

- The anomalous arterial supply may be readily detected and characterized. It usually arises from the descending aorta. In addition, the type of sequestration may be determined if the draining venous anatomy can be shown, which happens in 50% of cases.

- Intralobar sequestration venous drainage is typically into the inferior pulmonary vein (Fig. 16 on page 34, Fig. 17 on page 35 and Fig. 18 on page 36).

*Fig. 16: Intralobar sequestration. (a) Axial lung window CT image shows a focal opacity within the medial basal segment of the left lower lobe (white arrow). (b) Axial*
contrast-enhanced CT image demonstrates an anomalous artery arising from the descending aorta to feed the mass (black arrow).

**References:** Serviço de Imagem Médica, Centro Hospitalar e Universitário de Coimbra - Hospital Pediátrico, Portugal

**Fig. 17:** Intralobar sequestration (same case as in fig. 16). Axial contrast-enhanced CT images demonstrate the venous drainage of the mass into the left inferior pulmonary vein (arrows).

**References:** Serviço de Imagem Médica, Centro Hospitalar e Universitário de Coimbra - Hospital Pediátrico, Portugal
**Fig. 18:** Intralobar sequestration (same case as in figs. 16 and 17). Coronal maximum-intensity projection images show the feeding artery of the mass arising from the descending aorta (black arrow) and venous drainage into the left inferior pulmonary vein (white arrow).

**References:** Serviço de Imagem Médica, Centro Hospitalar e Universitário de Coimbra - Hospital Pediátrico, Portugal

- Typical drainage for extralobar sequestration is through the azygos vein, with drainage to the subclavian, internal mammary and portal veins being less common (Fig. 19 on page 37, Fig. 20 on page 38, Fig. 21 on page 38 and Fig. 22 on page 39).
**Fig. 19:** Extralobar sequestration. Lung window (a) and soft tissue window (b) CT images show a focal opacity in the left lower lobe.

*References:* Serviço de Imagem Médica, Centro Hospitalar e Universitário de Coimbra - Hospital Pediátrico, Portugal

![Fig. 19: Extralobar sequestration.](image)

**Fig. 20:** Extralobar sequestration (same case as in fig. 19). Axial and coronal CT angiography images demonstrate an anomalous artery arising from the descending aorta to feed the mass (arrows).

*References:* Serviço de Imagem Médica, Centro Hospitalar e Universitário de Coimbra - Hospital Pediátrico, Portugal

![Fig. 20: Extralobar sequestration.](image)
**Fig. 21**: Extrinsic sequestration (same case as in figs. 19 and 20). Axial contrast-enhanced CT images demonstrate the venous drainage of the mass into the portal vein (arrows).

**References**: Serviço de Imagem Médica, Centro Hospitalar e Universitário de Coimbra - Hospital Pediátrico, Portugal
Fig. 22: Extralobar sequestration (same case as in figs. 19, 20 and 21). Coronal maximum-intensity projection image demonstrates the feeding artery of the mass arising from the descending aorta (curved arrow) and the draining vein ending into the portal vein (arrows).

References: Serviço de Imagem Médica, Centro Hospitalar e Universitário de Coimbra - Hospital Pediátrico, Portugal

Hypogenetic lung syndrome (Scimitar syndrome)

- Hypogenetic lung syndrome can be definitively diagnosed on chest radiography in many cases. The anomalous pulmonary vein appears as a vertically oriented curvilinear opacity within the right hemithorax (the radiographic appearance of the anomalous vein has been
likened to a type of Turkish sword known as a scimitar, which gave the name to the syndrome. The right lung is small with resultant right hemithorax volume loss and shift of the mediastinum to the right. The left lung is hyperinflated (Fig. 23 on page 40).

![Radiograph showing partial hypogenetic lung syndrome (scimitar syndrome).](image)

**Fig. 23**: Partial hypogenetic lung syndrome (scimitar syndrome). Frontal chest radiograph shows a vertically oriented curvilinear opacity (arrows) projecting over the right lower hemithorax. Also noted is hypoplastic right lung.

**References**: Serviço de Radiologia, Centro Hospitalar Tondela - Viseu, Portugal

- CT imaging is useful in the evaluation of the course of the anomalous pulmonary vein and its drainage. Most commonly the scimitar vein drains into the inferior vena cava, with less common sites being the portal vein, hepatic vein, superior vena cava vein, right atrium and azygos vein (Fig. 24 on page 41). Using angiographic protocols, an anomalous systemic arterial supply can also be shown. Nonvascular findings include
hypoplasia of the right lung with parenchymal changes, abnormal lobation and anomalous bronchial branching.

Fig. 24: Partial hypogenetic lung syndrome (scimitar syndrome) (same case as in fig. 23). Axial contrast-enhanced CT images show the anomalous vein (arrows) draining into the inferior vena cava.

References: Serviço de Radiologia, Centro Hospitalar Tondela - Viseu, Portugal
**Fig. 1:** Type 1 congenital pulmonary airway malformation. Axial lung window CT images show a macrocystic mass (*) in the left lower lobe. There are other smaller cysts adjacent to the dominant cyst (arrow). Given the cyst size, findings are consistent with a type 1 CPAM.

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**Fig. 2:** Type 1 congenital pulmonary airway malformation. Axial lung window CT images demonstrate multiple cystic lesions in the left upper lobe, with a dominant cyst measuring more than 2 cm (*). Mediastinal shift to the right side is also seen.

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Fig. 3: Type 2 congenital pulmonary airway malformation. Axial lung window CT images demonstrate an air-filled multicystic mass in the right upper lobe. All cysts measure less than 2 cm which is consistent with a type 2 CPAM.

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**Fig. 4:** Type 2 congenital pulmonary airway malformation. Axial lung window CT images show multiple cystic lesions in the right lower lobe. All cysts measure less than 2 cm which is consistent with a type 2 CPAM.

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**Fig. 5:** Infected type 1 congenital pulmonary airway malformation. Axial lung window CT images demonstrate large cystic lesions in the left lower lobe, with air-fluid levels (arrows). Mediastinal shift to the right side is also seen.

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Fig. 17: Intralobar sequestration (same case as in fig.16). Axial contrast-enhanced CT images demonstrate the venous drainage of the mass into the left inferior pulmonary vein (arrows).

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**Fig. 18:** Intralobar sequestration (same case as in figs. 16 and 17). Coronal maximum-intensity projection images show the feeding artery of the mass arising from the descending aorta (black arrow) and venous drainage into the left inferior pulmonary vein (white arrow).

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**Fig. 19:** Extralobar sequestration. Lung window (a) and soft tissue window (b) CT images show a focal opacity in the left lower lobe.
Fig. 20: Extralobar sequestration (same case as in fig. 19). Axial and coronal CT angiography images demonstrate an anomalous artery arising from the descending aorta to feed the mass (arrows).
Fig. 21: Extralobar sequestration (same case as in figs. 19 and 20). Axial contrast-enhanced CT images demonstrate the venous drainage of the mass into the portal vein (arrows).

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**Fig. 22:** Extralobar sequestration (same case as in figs. 19, 20 and 21). Coronal maximum-intensity projection image demonstrates the feeding artery of the mass arising from the descending aorta (curved arrow) and the draining vein ending into the portal vein (arrows).

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Conclusion

- In this work we reviewed a group of congenital pulmonary malformations involving lung parenchyma and both lung parenchyma and vascular structures.

- These anomalies represent a diverse group of development disorders with a wide distribution in imaging appearance and clinical manifestations.

- They can be a source for important morbidity and mortality in infants and children. However, some of them may remain asymptomatic and be incidentally detected in adulthood.

- Thus it is imperative for both pediatric and adult radiologists to be familiar with the imaging characteristics of each lesion and proper imaging techniques in order to optimize diagnostic accuracy and disease management.
References


