Congenital Lung Malformations, what the radiologist should know

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Learning objectives

- To know the wide spectrum of congenital lung malformations of the foregut
- To understand the current nomenclature
- To show the imaging findings of each malformation with different techniques
Background

Pulmonary malformations, also called congenital anomalies of the primitive foregut, represent a broad spectrum of relatively frequent abnormalities with a common origin at some point of the embryological development of the foregut/lung.

These anomalies are detected in most of cases in infants and children, although asymptomatic cases can be diagnosed in adults. The current spread of prenatal ultrasound has meant a large increase in the diagnosis of these anomalies.

Pulmonary malformations form a continuous spectrum. At one end there are cases with abnormal lung with normal vasculature (congenital lobar hyperinflation, bronchial atresia, bronchial cyst…) and in the other end, cases with abnormal vasculature and normal lung (arteriovenous malformation). In between, there are lesions with both, parenchyma and vascular abnormalities (pulmonary sequestration, hypogenetic lung syndrome…) Fig. 1 on page 13.

To understand their development, the lung embryological development is going to be remembered. Five phases are described:

- **Embryonic phase** (3-7 weeks): begins with the formation of the respiratory diverticulum from the ventral wall of the primitive foregut. The tracheoesophageal septum, which separates respiratory and digestive systems, derived from an invagination of the mesoderm. Around 33 days of gestation, the distal end of the respiratory diverticulum bifurcates into two buds (left and right primary bronchi) Fig. 2 on page 13.

- **Pseudoglandular phase** (7-17 weeks): segmental and subsegmental bronchi are formed through a series of divisions. Initially three segmental bronchi on the right lung and two on the left side. Later in time, segmental bronchi are divided dichotomously to form 10 subsegmental bronchi in the right lung and 8 in the left lung. In subsequent divisions, the bronchi give rise to a number of alveolar ducts and alveoli Fig. 3 on page 14.

In the central air-conducting portions of the lungs, the epithelium begins to differentiate into cilia-carrying and goblet cells. After the 10th week, cartilage and smooth muscle cells as well as bronchial glands can be found into the bronchi wall.

The new pulmonary circulation system is formed out of the 6th pharyngeal arch artery, and it is divided according to the airways Fig. 4 on page 14.
- **Canalicular phase** (16-26 weeks): the distal acinar unit develops and further airspaces are being canalized and aproximated by a network of capillaries. Into the lumen of the alveolar sacculi are formed the pneumocytes I and II.

- **Saccular phase** (22-36 weeks): the alveoli and terminal sacs continue to develop, with compression of intervening interstitium and the beginning of alveolar septation.

- **Alveolar phase** (> 36 weeks): It is the lung maduration period, which is maintained until postnatal 2-3 years. Pneumocytes I and II increase in number and also increase the surfactant production. New vessels are formed and the primary septa decrease its thickness and contain more elastic fibres.

It's difficult and in most of cases confusing to understand the pathogenesis of lung malformations. According with some theories, these lesions are to defective foregut budding and differentiation. Others authors, believe that the cause is an airway obstruction with secondary pulmonary dysplastic changes. Variability in the embryologic timing and severity of the obstruction can explain hybrid and overlapping lesions.

Diagnosis and follow up of the bronchopulmonary malformations are performed with different imaging techniques (ultrasound, plain radiograph, MR, CT). The radiologist should select the appropriate imaging technique to provide the information necessary for the proper therapeutic management of the patient Fig. 5 on page 15 .

- **Ultrasound**: prenatal Ultrasound is an excellent initial screening test and the Doppler mode allows to know the lesion vasculature.
- **Plain Radiography**: It's the initial test for the diagnosis of these malformations. It should be performed in any infant with respiratory distress and in those with suspected bronchopulmonary malformation on prenatal ultrasound. This imaging technique provides information about location, size and mass effect. It is also useful for monitoring.
- **CT**: It is the technique of choice for surgical planning, allowing a better evaluation of the lung parenchyma, airway and vascular structures and concomitant malformations. It should be done with iv contrast to adequate assessment of vascularization.
- **MR**: fetal-MR is a very useful technique in the characterization of the lesion and measurement of the healthy lung volume. MR angiography is also useful for the assessment of vascularity although less than CT in the assessment of lung parenchyma

The most frequent broncopulmonary malformations in children will be explain in detail.
a) Bronchogenic Cysts

Bronchogenic cysts are thought to arise secondary to abnormal budding of the primitive ventral foregut, probably between 4-7 weeks of fetal life. They are mostly situated in the mediastinum (70%), typically subcarinal, hiliar or right paratracheal locations, but they can also be intraparenchymal (predominantly in lower lobes). Rarely it may appear in the neck, pericardium or abdominal cavity.

Most bronchogenic cysts are asymptomatic and are found incidentally. The symptoms are generally caused by compression of the trachea, bronchi or esophagus, leading wheezing, stridor, dyspnea, and dysphagia. Spontaneous pneumothorax is rare. Intraparenchymal cysts may complicated with recurrent infections or hemorrhage, and more rarely, malignant tumors (rhabdomyosarcoma, pulmonary blastoma and adenocarcinoma).

The bronchogenic cyst has the following characteristics:

- **Prenatal ultrasound**: anechoic lesion with posterior acoustic enhancement in fetal thorax Fig. 6 on page 15.

- **Chest radiograph**: the findings are non specific, with a water-density mass, well demarcated, in the mediastinum or central lung. Airway compression may lead to overinflation of an adjacent lobe or lobar atelectasis. Infection of the cyst may lead to surrounding acinar shadowing. An air-fluid level may be present if tracheobronchial connection develops. In some cases peripheral calcifications can be seen Fig. 7 on page 16.

- **CT**: helps in characterizing the precise anatomic location, intrinsic density (0-20 UH) and the relationship with adjacent structures. Generally appearing as rounded, well demarcated, thin wall and non enhancing masses. In some cases it can have a higher attenuation (>30 UH) due to the presence of proteinaceous-mucoid material or by intracyst hemorrhage Fig. 8 on page 17 Fig. 9 on page 18.

- **MR**: typically bronchogenic cysts have high signal on T2-weighted images. The T1-weighted signal intensity is variable depending on the protein content or intracystic hemorrhage Fig. 10 on page 19.

Differential diagnoses include congenital pulmonary airway malformation, neumatocele, lung abscess, round pneumonia Fig. 11 on page 19, hydatic cyst Fig. 12 on page 20 and neurenteric cyst Fig. 13 on page 21 Fig. 14 on page 22.
b) Pulmonary sequestration

Pulmonary sequestration is defined as a dysplastic pulmonary tissue that does not communicate with the tracheobronchial tree, and that received its blood supply from the systemic circulation, usually from the thoracic or abdominal aorta.

Two forms of pulmonary sequestration have been described: extralobar and intralobar Fig. 15 on page 22 Fig. 16 on page 23.

- **Intralobar pulmonary sequestration (ILPS):**
  - The most frequent (70%).
  - It’s a segment of pulmonary tissue that shares the visceral pleura covering the normal adjacent lung tissue.
  - It's usually located in the posterobasal portion of the left lower lobe.
  - The arterial supply is systemic, typically from the thoracic or upper abdominal aorta.
  - The venous drainage is into the pulmonary venous system.
  - The origin is usually congenital but some authors argue that some of these lesions could be acquired, due to recurrent infected lung segment.

- **Extralobar pulmonary sequestration (ELPS):**
  - This type has its own pleura investment.
  - The artery supply is typically from the thoracic or abdominal aorta.
  - The venous drainage is systemic, into azygos, hemiazygos or portal veins.
  - It may have communication with the gastrointestinal tract.
  - Usually it has paraspinal location in the lower chest, even below the diaphragm.
  - Usually it is associated with other congenital malformations (congenital diaphragmatic hernia, congenital heart disease, congenital pulmonary airway malformation …) Fig. 17 on page 23.

ELPS is usually diagnosed in the neonatal period or in the first year of life. Respiratory distress and feeding difficulties are the most frequent symptoms, but it may be an
incidental prenatal or postnatal imaging finding, in some cases during the study of other congenital anomalies.

ILPS is most commonly diagnosed later in life (childhood or even adulthood) and is associated with recurrent pneumonias.

The radiological findings of these lesions are:

- **Prenatal ultrasound**: homogeneous, hyperechoic, well defined mass in a paraspinal location, most often the left lower thorax. At color Doppler US we can see the feeding artery originating from the descending aorta Fig. 18 on page 24.

- **Chest X-ray**: it is difficult to distinguish between ELPS and ILPS on plain radiographs alone. ELPS Fig. 19 on page 24 are usually found as well defined, retrocardiac masses, in the cardiophrenic angle. Air bronchogram is absent. It may be found as a subdiaphragmatic or mediastinal mass lesion. It can be associated with opaque ipsilateral hemithorax and pleural effusion. ILPS tends to be more heterogeneous and less well defined. Focal bronchiectasis, areas of atelectasis, cavitation, and cyst formation may also be recognized.

Radiography may be normal at birth Fig. 20 on page 25, even if there was a suspicion in the prenatal US, posterior studies should be performed.

- **CT**: postnatal CT is the technique of choice. CT-angiography techniques and 3-dimensional images can help to show aberrant arterial supply and anomaly drainage to the surgical planning.

ELPS shows an homogeneous enhancing, well circumscribed mass of soft tissue attenuation. ILPS may show more irregular edge Fig. 21 on page 26 Fig. 22 on page 27 and heterogeneous in appearance (cystic containing air or fluid, focal emphysema). Hypervascularity have also been showed.

- **MR**: MR imaging may also be used to identify the feeding arterial vessels. Pulmonary sequestration mass is usually of high signal on both T1 and T2-weighted images. Hemorrhage or cystic components are easy to demonstrate Fig. 23 on page 27.

Most pulmonary sequestration are surgically resected electively, even in asymptomatic patients, due to the risk of infection, hemorrhage, and questionable malignancy. The artery embolization may precede the surgery.
c) Congenital pulmonary airway malformation

Congenital pulmonary airway malformation (CPAM) is the current name of the congenital cystic adenomatoid malformation (CCAM). It is the most commonly diagnosed lung malformation in prenatal period and accounts the 25% of all congenital bronchopulmonary malformations.

These malformations are characterized by an abnormal branching of immature bronchioles, with a lack of normal alveolar development. CPAM can develop in any lung lobe, except in the middle lobe where is very rare, and usually communicates with the normal tracheobronchial tree.

The Stocker classification Fig. 24 on page 28 have been widely used and divided this malformation in 3 groups, according to the cysts size:

- **Type I** (the most common type 75%): usually diagnosed in neonatal period and characterized by containing one or more cysts measuring over 2 cm. The cysts are lined by ciliate columnar epithelium and their walls contain abundant elastic fibres.

- **Type II** (10-15%): contains cysts less than 2 cm in diameter, and are lined with cuboidal or columnar epithelium. This type is associated with other congenital malformations (congenital heart disease, pulmonary sequestration, renal agenesis ...). Tumoral degeneration (rhabdomyosarcoma and pulmonary blastoma) may occur.

- **Type III** (10%): have a poor prognosis. Usually contain cysts less than 0’5 cm in a compact and homogeneous mass. Cysts are delimited by cuboidal epithelium.

The new classification is an extension of the Stocker classification and include five types:

- **Type 0**: tracheal or bronchial origin and it is really acinar dysgenesis or dysplasia.

- **Type I**: bronchial or bronchiolar origin (cysts with 2-10cm diameter)

- **Type II**: bronchiolar origin ( 0’5-2 cm cyst lesions)

- **Type III**: bronchiolar-alveolar duct origin (adenomatoid type)

- **Type IV**: distal acinar origin. This type at imaging is indistinguishable from a predominantly cystic pleuropulmonary blastoma.
Most CPAM derive their blood supply from the pulmonary artery and drain via the pulmonary veins.

The symptoms are highly variable, from asymptomatic cases that are late diagnosed by recurrent lung infections, until early fetal or neonatal death due to hypoplasia or hydrops fetalis.

In the past, CPAM was diagnosed in the early neonatal period, in infants who presented acute respiratory distress. Nowadays, is increasingly being diagnosed on prenatal US (echogenic soft tissue with or not cysts images). This lesion can increase in size during the pregnancy and be associated with polyhydramnios or hydrops fetalis.

The chest radiograph findings are variable, and correlate with the type of CPAM:

- Type I: multicystic lesion formed by large cysts with or without a dominant cyst Fig. 25 on page 28. Early radiographs may demonstrate a water density mass if the cysts are dilled with retained fetal lung fluid. Mass effect can cause contralateral mediastinal shift, inversion of the ipsilateral hemidiaphragm and atelectasis of the ipsilateral and contralateral lung.

- Type II: multicystic mass with cysts of a smaller size (# 2 cm) filled with air or a focal area of consolidation Fig. 26 on page 29.

- Type III: homogeneous, soft tissue density mass.

In cases of prenatal suspicion of CPAM and normal neonatal chest radiograph, the study must complete with a CT.

CT helps in characterizing the lesion, document the involved pulmonary lobe or segment and also confirm the diagnosis in asymptomatic infants. Intravenous contrast media must be used to define the presence of any systemic arterial vessels supplying the involve lung (a hybrid lesion), and it is important to the surgery planning. CPAM appears as a multicystic mass with varying sizes cysts depending on the type of malformations Fig. 27 on page 30. Fluid-filled cystic and air-fluid levels area appreciated. In the case of type III malformation, a homogeneous solid mass is seen (microcysts diagnosis is a pathologic feature). CT also helps to diagnose other associated congenital malformations.

Differential diagnoses should be made with pulmonary sequestration, bronchogenic cyst and diaphragmatic hernia.
Type II and type III are often diagnosed in childhood and associated with other congenital anomalies (specially pulmonary sequestration and bronchial atresia) Fig. 28 on page 31.

The treatment of these malformations is the surgery due to the risk of complications and malignancy, although asymptomatic CPAMs surgical resection remains controversial.

d) Bronchial atresia

Bronchial atresia is a rare anomaly resulting from focal obliteration of a lobar, segmental or subsegmental bronchus, with secondary dysplastic changes in the distal lung parenchyma. The bronchi distal to the stenosis are dilated and filled with mucus, resulting in the formation of a bronchocele. The alveoli supplied by these bronchi are ventilated by collateral pathways and show features of air-trapping, resulting in a region of hyperinflation around the dilated bronchi.

In bronchial atresia the airway is occluded rather than narrowed, consequently there is no ball-valve effect hence the lobe or segment does not become hyperinflated as with in other congenital malformations like congenital lobar overinflation, explained later.

The lesion may be solitary or multiple. The upper-lobe bronchi are more frequently affected (left upper lobe more than right one), middle and lower lobes are rarely involved.

In neonatal period, bronchial atresia is seen as water density mass by the trapped lung fluid distal to the atresia on chest-X ray. Later, the fetal lung liquid escapes and bronchial atresia is found because of focal air trapping with a bronchocele Fig. 29 on page 31, although not always it can be seen Fig. 30 on page 32.

CT is an excellent modality for demonstrating the typical features of bronchial atresia. It shows the mucus-filled bronchus near the hilium (bronchocele), as well as the surrounding areas of air trapping Fig. 31 on page 33. Recall that there may be more malformations in the same patient Fig. 32 on page 34 Fig. 33 on page 35.

MR usually shows the bronchocele as a branching structure radiating from the helium, whit high signal intensity in T1 ant T2- weighted images, however MR cannot depict regional air trapping.

e) Congenital lobar hyperinflation
Congenital lobar hyperinflation, formerly called congenital lobar emphysema, is characterized by progressive overdistension of a lobe or lobes to result from a check-valve mechanism at the bronchial level that causes progressive hyperinflation of the lung.

It can either be acquired or congenital. The underlying cause can be secondary to an intrinsic cartilaginous abnormality or extrinsic compression of an airway. In either case, the collapsed airway can act as a one-way valve, resulting in air trapping. There is no destruction of the alveolar walls.

The most commonly affected lobe is the left upper lobe, followed by the middle lobe and the right upper lobe.

Most patients become symptomatic during the neonatal period with respiratory distress and in these cases, lobectomy is indicated.

Diagnosis is obtained by means of chest X-ray and CT, which show hyperinflation of the segment or lobe affected.

- **Chest X-ray**: radiograph shows a hyperlucent area in the lung which size depends on the overinflation degree. Adjacent lobes and structures may be compressed by the emphysematous lobe, and sometimes ipsilateral and contralateral atelectasis may occur. Rib space widening, hemidiaphragm depression and mediastinal shift Fig. 34 on page 36 can also be seen.

- **CT**: during the neonatal period may show a high attenuation area in the lung, since the involved section is still filled with fluid. Over time, the fluid will resolve, subsequently the affected lung will progressive overinflate and become lower attenuation with the septa and vascular structures in the periphery of the distended alveoli Fig. 35 on page 37. The same findings described above in radiography (mediastinal shift, hemidiaphragm depression, atelectasis...) can also be seen on CT.

**RADIOLOGY REPORT.**

The classification of bronchopulmonary malformations is complex, it is under constant review and the radiological findings can be common to several malformations, some authors advocate a simple classification that include all under one name: THORACIC CONGENITAL MALFORMATIONS.
Moreover, it is important include in our radiology report the following points:

- Solid / cystic or mixed malformation. If the lesion is cystic, specify whether microcystic or macrocystic.
- Presence or absence of systemic arterial vascularization.
- Lobar location (uni or bilateral, single or multiple lobes).
- Description of the adjacent airway and parenchyma.
- Communication with the gastrointestinal tract.
- Associated malformations vascular, chest wall, diaphragm, superior abdomen.)
Fig. 1: Graphic representation of the bronchopulmonary malformations spectrum (CLH: congenital lobar hyperinflation, BC: bronchogenic cyst; CPAM: congenital pulmonary airway malformation; BA: bronchial atresia; PS: pulmonary sequestration; AVM: arteriovenous malformations)

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Fig. 2: Graphic representation of the embryological development of the trachea and main bronchi

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Fig. 3: Graphic representation of the dichotomous bronchial division

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Fig. 4: Graphic representation of the pulmonary vasculature development
Fig. 5: Scheme of the different diagnostic techniques available for the diagnosis of bronchopulmonary malformations and more frequent use

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Fig. 6: Prenatal ultrasound: anechoic lesion with posterior acoustic enhancement, in the fetal chest cavity. The lesion corresponded to a bronchogenic cyst

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**Fig. 7:** Chest Radiograph: Well-defined mass of water density, superimposed on right upper lobe (arrow)

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Fig. 8: Coronal CT image with intravenous contrast. Rounded, thin-walled, well-defined lesion with heterogeneous content, located in the right lower lobe

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**Fig. 9:** A) Coronal CT image with intravenous contrast: rounded and well defined left paraspinal lesion. B) Axial section of the same lesion: rounded, hypodense and well defined lesion, which corresponds to a bronchogenic cyst

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**Fig. 10:** A) Coronal T2-weighted MR image: rounded and well-defined hyperintense lesion, localized in the upper mediastinum, which corresponds to a bronchogenic cyst. B) Axial T1-weighted image, of the same lesion: well circumscribed hipointense lesion

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**Fig. 11**: Chest radiograph: rounded water density lesion in right lower lobe, corresponding to a round pneumonia

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**Fig. 12:** Axial CT image with intravenous contrast: well-defined and thin edges cystic lesion, in right lower lobe, which corresponds to a pulmonary hydatid cyst.

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Fig. 13: Sagittal A) and axial B) CT images with intravenous contrast: low attenuation and ill-defined lesion in the posterior medistinum displacing the posterior side of the trachea and the esophagus (arrows).

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Fig. 14: Surgical specimen of the lesion described in Figure 13

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Fig. 15: Graphical representation of the types of pulmonary sequestration

© radiographics

Fig. 16: Summary table of intra and extralobar sequestration characteristics

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**Fig. 17:** Surgery images of a newborn with diaphragmatic hernia and a left extralobar sequestration: A) Left thoracotomy exposing the diaphragmatic defect and pulmonary infradiaphragmatic sequestration. B) Surgical specimen. C) Diaphragmatic hernia repaired and partially left lung reexpanded.

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**Fig. 18:** Prenatal ultrasound of a sequestration: A) Well defined hyperechoic lesion in the fetal thorax. B) Sagittal color Doppler image which shows the arterial supply of the lesion.

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Fig. 19: Chest radiograph: well-defined mass in the left lower lobe which determines loss of volume of the left lung and mediastinal shift to the right. The lesion corresponded to an extralobar sequestration

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Fig. 20: Normal chest radiograph of a newborn with pulmonary sequestration

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**Fig. 21:** CT scan with intravenous contrast of an intralobal sequestration in the left lower lobe: A) Coronal image in lung window: complex attenuation lesions, with central high attenuation area surrounded by low attenuation parenquima. B) Coronal MIP image: the arterial supply is from the abdominal aorta and the venous drainage is into the pulmonary venous system. C) Volume rendering image depicting clearly the vessels of the sequestration.

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**Fig. 22:** CT scan with intravenous contrast of the intralobar sequestration in right lower lobe: A) Axial image in mediastinal window: high attenuation lesion in the right pulmonary base. B and C) Oblique MIP images demonstrating the arterial supply from abdominal aorta and drainage into pulmonary veins.

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**Fig. 23:** T2-weighted A) and T1-weighted B) MR images of a pulmonary sequestration. High intensity on T2 and low intensity on T1 lesion. Note the arterial supply from thoracic aorta.
Fig. 24: Graphic representation of the congenital pulmonary airway malformation (CPAM) and their corresponding histology

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Fig. 25: Chest radiograph: large multicystic lesion in the right lung that associate significant mass effect and contralateral mediastinal shift. The lesion corresponds to a type I CPAM

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**Fig. 26:** Chest radiograph: ill-defined water density mass on right upper lobe with small radiolucencies inside, findings that are compatible with a type II CPAM.

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**Fig. 27:** Coronal (A) and sagittal (B) CT images of a type II CPAM: right upper lobe mass, formed by multiple cysts.

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**Fig. 28:** Axial (A) and coronal (B) CT images in lung window and coronal (C) in mediastinal window of the same patient of Figure 27. This patient besides the CPAM also had a cystic lesions adjacent to the right main bronchus, corresponding to a bronchogenic cyst.

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Fig. 29: Bronchial atresia. Chest radiograph: water density opacity leading from the right hilum to right lower lobe (bronchocele), hyperinflation of the right lung, increased intercostal space and mediastinal shift to the left.

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**Fig. 30:** Chest radiograph: hyperinflation of the left lung, increased intercostal space and mediastinal shift to the right

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**Fig. 31**: Axial CT image: dilated bronchi filled with mucus in the right lower lobe, corresponding to a bronchocele (arrow), surrounded by an area of air trapping

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**Fig. 32:** Same patient as Figure 34, axial CT image in mediastinal window: right paraspinal, well defined, soft tissue attenuation lesion, which corresponds to an associated extralobar sequestration

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**Fig. 33:** CT scan axial (A) and saggital (B) images: nodular lesion with soft tissue attenuation (bronchocele, arrow) in left upper lobe, surrounded by a hyperlucent area of air trapping, corresponding to a segmental bronchial atresia. B) Sagittal image: hypodense lesions in lingula and left lower lobe, with a cystic component, corresponding to CPAM (stars).

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**Fig. 34:** Chest radiograph: low density and expanded left lung with important mass effect (mediastinal shift to the right, contralateral atelectasis and increased of the left intercostal space) compatible with congenital lobar hyperinflation

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Fig. 35: A) Axial CT image with intravenous contrast, in lung window: marked hiperlucency of the left upper lobe, compatible with congenital lobar hyperinflation. B) Surgical specimen

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Imaging findings OR Procedure details

a) Bronchogenic Cysts:

- **Prenatal ultrasound**: anechoic lesion with posterior acoustic enhancement in fetal thorax Fig. 6 on page 42.

- **Chest radiograph**: the findings are non specific, with a water-density mass, well demarcated, in the mediastinum or central lung. Airway compression may lead to overinflation of an adjacent lobe or lobar atelectasis. Infection of the cyst may lead to surrounding acinar shadowing. An air-fluid level may be present if tracheobronchial connection develops. In some cases peripheral calcifications can be seen Fig. 7 on page 42.

- **CT**: helps in characterizing the precise anatomic location, intrinsic density (0-20 UH) and the relationship with adjacent structures. Generally appearing as rounded, well demarcated, thin wall and non enhancing masses. In some cases it can have a higher attenuation (>30 UH) due to the presence of proteinaceous-mucoid material or by intracyst hemorrhage Fig. 8 on page 43 Fig. 9 on page 44.

- **MR**: typically bronchogenic cysts have high signal on T2-weighted images. The T1-weighted signal intensity is variable depending on the protein content or intracystic hemorrhage Fig. 10 on page 45.

b) Pulmonary sequestration

- **Prenatal ultrasound**: homogeneous, hyperechoic, well defined mass in a paraspinal location, most often the left lower thorax. At color Doppler US we can see the feeding artery originating from the descending aorta Fig. 18 on page 45.

- **Chest X-ray**: it is difficult to distinguish between ELPS and ILPS on plain radiographs alone. **ELPS** Fig. 19 on page 46 are usually found as well defined, retrocardiac masses, in the cardiophrenic angle. Air bronchogram is absent. It may be found as a subdiaphragmatic or mediastinal mass lesion. It can be associated with opaque ipsilateral hemithorax and pleural effusion. **ILPS** tends to be more heterogeneous and less well defined. Focal bronchiectasis, areas of atelectasis, cavitacion, and cyst formation may also be recognized.

Radiography may be normal at birth Fig. 20 on page 52, even if there was a suspicion in the prenatal US, posterior studies should be performed.
- **CT**: postnatal CT is the technique of choice. CT-angiography techniques and 3-dimensional images can help to show aberrant arterial supply and anomaly drainage to the surgical planning.

ELPS shows an homogeneous enhancing, well circumscribed mass of soft tissue attenuation. ILPS may show more irregular edge Fig. 21 on page 47 Fig. 22 on page 48 and heterogeneous in appearance (cystic containing air or fluid, focal emphysema). Hypervascularity have also been showed.

- **MR**: MR imaging may also be used to identify the feeding arterial vessels. Pulmonary sequestration mass is usually of high signal on both T1 and T2-weighted images. Hemorrhage or cystic components are easy to demonstrate Fig. 23 on page 48.

c) **Congenital pulmonary airway malformation**

The chest radiograph findings are variable, and correlate with the type of CPAM:

- Type I: multicystic lesion formed by large cysts with or without a dominant cyst. Early radiographs may demonstrate a water density mass if the cysts are dilled with retained fetal lung fluid. Mass effect can cause contralateral mediastinal shift, inversion of the ipsilateral hemidiaphragm and atelectasis of the ipsilateral and contralateral lung.

- Type II: multicystic mass with cysts of a smaller size (# 2 cm) filled with air or a focal area of consolidation Fig. 26 on page 50.

- Type III: homogeneous, soft tissue density mass.

In cases of prenatal suspicion of CPAM and normal neonatal chest radiograph, the study must complete with a CT.

CT helps in characterizing the lesion, document the involved pulmonary lobe or segment and also confirm the diagnosis in asymptomatic infants. Intravenous contrast media must be used to define the presence of any systemic arterial vessels supplying the involve lung (a hybrid lesion), and it is important to the surgery planning. CPAM appears as a multicystic mass with varying sizes cysts depending on the type of malformations Fig. 27 on page 51. Fluid-filled cystic and air-fluid levels area appreciated. In the case of type III malformation, a homogeneous solid mass is seen (microcysts diagnosis is a pathologic feature). CT also helps to diagnose other associated congenital malformations.

d) **Bronchial atresia**
- CT is an excellent modality for demonstrating the typical features of bronchial atresia. It shows the mucus-filled bronchus near the hilium (bronchocele), as well as the surrounding areas of air trapping Fig. 31 on page 53.

- MR usually shows the bronchocele as a branching structure radiating from the hilium, with high signal intensity in T1 and T2-weighted images, however MR cannot depict regional air trapping.

e) Congenital lobar hyperinflation

- **Chest X-ray**: radiograph shows a hyperlucent area in the lung which size depends on the overinflation degree. Adjacent lobes and structures may be compressed by the emphysematous lobe, and sometimes ipsilateral and contralateral atelectasis may occur. Rib space widening, hemidiaphragm depression and mediastinal shift Fig. 34 on page 54 can also be seen.

- **CT**: during the neonatal period may show a high attenuation area in the lung, since the involved section is still filled with fluid. Over time, the fluid will resolve, subsequently the affected lung will progressive overinflate and become lower attenuation with the septa and vascular structures in the periphery of the distended alveoli Fig. 35 on page 55. The same findings described above in radiography (mediastinal shift, hemidiaphragm depression, atelectasis...) can also be seen on CT.
Fig. 6: Prenatal ultrasound: anechoic lesion with posterior acoustic enhancement, in the fetal chest cavity. The lesion corresponded to a bronchogenic cyst

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Fig. 7: Chest Radiograph: Well-defined mass of water density, superimposed on right upper lobe (arrow)

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**Fig. 8:** Coronal CT image with intravenous contrast. Rounded, thin-walled, well-defined lesion with heterogeneous content, located in the right lower lobe

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Fig. 9: A) Coronal CT image with intravenous contrast: rounded and well defined left paraspinal lesion. B) Axial section of the same lesion: rounded, hypodense and well defined lesion, which corresponds to a bronchogenic cyst

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Fig. 10: A) Coronal T2-weighted MR image: rounded and well-defined hyperintense lesion, localized in the upper mediastinum, which corresponds to a bronchogenic cyst. B) Axial T1-weighted image, of the same lesion: well circumscribed hipointense lesion

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Fig. 18: Prenatal ultrasound of a sequestration: A) Well defined hyperechoic lesion in the fetal thorax. B) Sagittal color Doppler image which shows the arterial supply of the lesion.

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**Fig. 19:** Chest radiograph: well-defined mass in the left lower lobe which determines loss of volume of the left lung and mediastinal shift to the right. The lesion corresponded to an extralobar sequestration

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**Fig. 21:** CT scan with intravenous contrast of an intralobar sequestration in the left lower lobe: A) Coronal image in lung window: complex attenuation lesions, with central high attenuation area surrounded by low attenuation parenquima. B) Coronal MIP image: the arterial supply is from the abdominal aorta and the venous drainage is into the pulmonary venous system. C) Volume rendering image depicting clearly the vessels of the sequestration.

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**Fig. 22:** CT scan with intravenous contrast of the intralobar sequestration in right lower lobe: A) Axial image in mediastinal window: high attenuation lesion in the right pulmonary base. B and C) Oblique MIP images demonstrating the arterial supply from abdominal aorta and drainage into pulmonary veins.

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**Fig. 23:** T2-weighted A) and T1-weighted B) MR images of a pulmonary sequestration. High intensity on T2 and low intensity on T1 lesion. Note the arterial supply from thoracic aorta.

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**Fig. 25:** Chest radiograph: large multicystic lesion in the right lung that associate significant mass effect and contralateral mediastinal shift. The lesion corresponds to a type I CPAM

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Fig. 26: Chest radiograph: ill-defined water density mass on right upper lobe with small radiolucencies inside, findings that are compatible with a type II CPAM.

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**Fig. 27:** Coronal (A) and sagittal (B) CT images of a type II CPAM: right upper lobe mass, formed by multiple cysts

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Fig. 20: Normal chest radiograph of a newborn with pulmonary sequestration

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Fig. 31: Axial CT image: dilated bronchi filled with mucus in the right lower lobe, corresponding to a bronchocele (arrow), surrounded by an area of air trapping

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**Fig. 34:** Chest radiograph: low density and expanded left lung with important mass effect (mediastinal shift to the right, contralateral atelectasis and increased of the left intercostal space) compatible with congenital lobar hyperinflation

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Fig. 35: A) Axial CT image with intravenous contrast, in lung window: marked hiperlucency of the left upper lobe, compatible with congenital lobar hyperinflation. B) Surgical specimen

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Conclusion

- There is a constant review of the nomenclature and classification, due to overlap radiological and histological findings, which suggest that thoracic congenital malformations present a common pathogenesis.

- CT is the diagnostic imaging method of choice, and always should be performed with intravenous contrast because it is essential to know the vascular supply malformation.

- There is a frequent association of these malformations so several of them can be find in the same patient.
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