Perinatal Imaging in Congenital Thoracic Cystic Malformations.

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Learning objectives

1. To point out the clinical importance of the detection and proper diagnosis of congenital thoracic cystic malformations.
2. To learn when and why to use the different imaging techniques in these patients.
3. To recognise the impact of perinatal imaging in fetal and neonatal clinical management.

Background

Congenital thoracic malformations represent a wide spectrum of developmental anomalies whose origin is not well established.

Although they are rare, its clinical importance is remarkable.

We are going to focus on those malformations of cystic nature. The most common of them are those affecting the lungs:

- Congenital pulmonary airway malformation or CPAM.
- Bronchopulmonary sequestration and hybrid lesions.
- Bronchogenic cyst.
- Bronchial atresia.

There are even more rare congenital cystic thoracic anomalies that do not affect the lung but other chest structures:

- Mediastinal or pericardial teratomas.
- Mediastinal or axillary lymphangiomas.
- Chest wall tumors.

Diaphragmatic hernia is also another entity to be aware of because its prenatal appearance might be confused with a cystic pulmonary malformation.

Nowadays, congenital thoracic malformations are usually detected in prenatal studies. At present, ultrasound (US) and magnetic resonance imaging (MRI) are the diagnostic tools by which non-invasively examine fetal chest development.
Although US remains as the primary screening method for thoracic anomalies and for identifying specific lung lesions, mediastinal shift or the presence of pleural effusion, foetal MRI can obtain further relevant information about the nature or prognosis of these lesions, like the presence of lung hypoplasia. Therefore, advanced prenatal imaging techniques help to parental counseling and perinatal assessment.

**Postnatal** imaging should always start with a radiography because it gives the most valuable information in a fast and unexpensive fashion. If a cystic component or vascular supplies are suspected within a lesion, US and Doppler examination are very useful. Once the diagnosis of a congenital thoracic mass or lesion is made by radiography and/or Doppler-US, other studies like contrast enhancement **Computed Tomography** (CeCT) or MRI might be required to confirm prenatal diagnosis, rule out complications and decide the surgical planning.

Surgical removal is the most frequent treatment of these malformations and the time of surgery will depend on the clinical status of the patient. For instance, if a neonate presents with respiratory distress because of a big CPAM, surgery must be performed in first days of life. Nevertheless, if the neonate is asymptomatic and the diagnosis of a CPAM is made with a plain film and/or US, another study like a CeCT would be not necessary until surgery is considered to be performed (usually around 6 months of life).

**Imaging findings OR Procedure details**

**CONGENITAL CYSTIC MALFORMATIONS AFFECTING THE LUNGS**

**CPAM**

Formerly called congenital cystic adenomatoid malformation or CCAM, the congenital pulmonary airway malformation or CPAM is the result of an abnormal development of the bronchial tree. Most of these lesions appear in prenatal imaging as a multicystic mass (Fig.1) occupying one lobe. Macrocystic or microcystic types can be determined by imaging. Evolution of these lesions can widely vary in each patient: while some lesions may even disappear others may cause foetal hydrops and stillbirth.

Postnatal imaging must start with a chest plain film (Fig.2) to confirm the diagnosis and evaluate its extent and complications like mediastinal shift, pleural effusions, etc.

Symptomatic CPAM are normally managed with surgical resection in first week of life. On the other hand, management of asymptomatic CPAM is controversial although most
advocate elective resection because of risks of infection and malignancy. In our Hospital these patients are postnatally followed and at 6 months of age a CeCT is performed as a previous step to elective surgery.

**DIAPHRAGMATIC HERNIA (DH)**

We include congenital DH in this group as an important differential diagnosis of CPAM in prenatal imaging. Although currently DH are usually detected prenatally some may present late in pregnancy and suddenly present at birth or in the first days of life, being one of the most important surgical causes of neonatal respiratory distress to be recognised by radiologists.

DH may be prenatally diagnosed by US but lately MRI (Fig.3) has proven its capability to better determine DH important features like contralateral pulmonary hypoplasia, liver-up herniation, etc.

Postnatally, a neonatal chest radiography showing bubble-like lucencies within the chest with a dilated gastric bubble and paucity or absence of abdominal gas (Fig.4) is usually enough to make the right diagnosis. Surgery will be necessary to repair the diaphragmatic congenital defect.

**BRONCHOPULMONARY SEQUESTRATION (BPS)**

BPS is a congenital area of abnormal lung whose origin is similar to the other entities like CPAM. It is a dysplastic lung without connection to the bronchial tree but with systemic arterial supply (typically arising from descending aorta).

It usually presents as an echogenic lung mass in prenatal US with doppler demonstration of the arterial supply.

BPS seldom has cystic nature but it may occur in conjunction with cystic lesions as CPAM. This form of BPS+CPAM is called hybrid lesion and in our experience is not so rare as an association. Therefore, one should always try to detect arterial supply in a CPAM or in a congenital solid pulmonary lesion to rule out this sort of lesions.

**BRONCHOGENIC CYST (BC)**

BC are developmental lesions that are part of the family of foregut duplication cysts.

They most commonly appear as a solitary well-defined cystic mass in the paratracheal or subcarinal region of the mediastinum, although some of them can be located
intrapulmonary and can be indistinguishable from CPAM. BC are homogenously hypoechoic in US and hyperintense on a T2-weighted prenatal MRI sequence (Fig.5).

Postnatally, depending on the size and location of the BC the clinical impact will be different. When they are big, airway or esophageal compression can be present. If they are small they cannot even be identified in a chest radiograph. Usually CT or MRI are needed for further evaluation (Fig.6). Surgical resection is recommended.

**BRONCHIAL ATRESIA (BA)**

BA is a rare congenital atresia of a proximal segmental bronchus with normal distal architecture. It normally affects apicoposterior segment of Left upper lobe. It can be detected in utero or most commonly in neonatal period being similar to CPAM in appearance. CT usually allows for more definite diagnosis showing a tubular branching mass surrounded by hyperinflated lung (Fig.7)

**CONGENITAL CYSTIC MALFORMATIONS NOT AFFECTING THE LUNGS**

**LYMPHANGIOMAS (LYMPH.)**

Lymph. can be detected as early as in fetal period with prenatal US. They appear like big cystic masses normally arising in the lateral regions of the neck.

20% of them do not affect the neck but may affect the chest region. The most frequent here are axillary limph. growing as cystic masses between the arm and the chest wall (Fig.8). Sometimes, the patients can show multiple lymphangiomas and the **mediastinum** is commonly affected (Fig.9).

**MEDIASTINAL and PERICARDIAL TERATOMA**

Fetal tumors are very rare. The most common are teratomas but those growing in fetal chest are even rarer.

They may appear as a heterogeneous mass with calcifications in the superior **mediastinum** so the diagnosis is quite easy, but sometimes it can adopt a confusing "benign"aspect in prenatal US (Fig.10) and finally continue growing next to the heart and then develop as a mass within the pericardium. MRI can add valuable information. In **pericardial** teratomas the presence of pericardial effusion is the key point (Fig.11).

**CHEST WALL TUMOR (mesenchymal hamartoma, MH)**
It is very uncommon to find chest wall tumors in fetal period. There are some of them described in the literature. When they grow within the chest, and present cystic nature like the MH, they could mislead to a CPAM diagnosis (Fig.12). Fetal MRI here is very useful to identify and determine the origin of these confusing lesions (Fig.13). Anyway, most of these extremely rare tumors are normally diagnosed postnatally (Fig.14). Mesenchymal hamartomas are benign lesions arising from the ribs and can be detected in utero. Despite its benign origin they can produce fetal demise secondary to mediastinal compression.

Images for this section:

Fig. 1: CPAM. Axial plane of a T2-weighted MR image of a 21 week-old fetus. Multiple rounded cystic structures (white arrows) were seen in the left lung. No mediastinal shift or pleural effusion was demonstrated.
Fig. 2: Neonatal chest radiograph of a one-day old neonate(same patient showed in Fig.1). Over-inflated lucencies related to a CPAM in the left lung. Despite the mediastinal shift the patient was almost asymptomatic. Anyway, an early surgery was necessary due to the big size of the lesion.
Fig. 3: Diaphragmatic Hernia (DH). A coronal T2-weighted MR image of a 22 week-old fetus demonstrates the right hemithorax occupation by multiple hypointense structures (white arrows) corresponding to multiple bowel loops in a right-sided DH. It was a "Liver-down" type DH because liver was not included in the hernia. Dilated gastric bubble (red arrow).
Fig. 4: Right-sided DH in a neonatal chest radiograph (same patient that figure 3). Orotracheal tube. The absence of abdominal air, dilated gastric bubble and aieated rounded lucencies in the right hemithorax confirm the prenatal MRI diagnosis.
**Fig. 5:** Bronchogenic cyst. Sagital T2-weighted MR image of a 24 week-old fetus. Round-shaped middle-mediastinal located cystic mass corresponding to a BC (white arrow).
Fig. 6: BC. Postnatal non-enhanced CT of the same patient than Fig. 5. Region of interest marking the subcarinal liquid containing mass confirming the prenatal diagnosis.
**Fig. 7:** Bronchial atresia. Contrast-enhanced CT in a neonate. Overinflated anterior segment of the upper right lobe (white arrow) and central hilar mass confirming a BA.
Fig. 8: Giant axillary and chest wall lymphangioma. Transversal T2- weighted MR image showing a huge thin wall cystic mass in a 21 week-old fetus. White arrows indicate the limits of the mass. A hypointense tubular structure (red arrow) was seen arising from the fetal chest wall.

![Image of a T2-weighted MR image showing a giant axillary and chest wall lymphangioma.](image)

Fig. 9: Axial plane of a T2-weighted of a 31 week-old fetus. A cystic mass is located in anterior mediastinum (white arrow), close to the thymic gland. This fetus also presented abdominal wall and retroperitoneal cystic lesions (not shown). The patient was born and the presence of multiple lymphangiomas was confirmed. MRI is useful to determine the extent and number of lesions in cases of multiple fetal lymphangiomas.
Fig. 10: Axial plane of a prenatal Doppler-US of a 21 week-old fetus. A round avascular and hypoechoic cystic mass (white arrow) was identified and described as a possible bronchogenic or pericardial cyst.
**Fig. 11:** Pericardial teratoma. Axial plane of a FIESTA MRI sequence of the same fetus of picture 10 but 2 weeks later. The previous lesion had increased and was more complex than it was suspected (white arrow). Pericardial effusion (*) was also identified. Both findings changed our previous diagnosis and we reported it as a probable pericardial cystic teratoma, what was confirmed postnatally.
Fig. 12: A mid-axial plane of a 21 week-old fetus chest US. A complex cystic mass was identified in the right hemithorax (white arrow and between calipers). We thought of an atypical CPAM or even a tumor so MRI was performed (see next figure).
**Fig. 13:** Chest wall tumor. Axial SSFSE-T2 weighted image of the same fetus in Fig.12. The thoracic mass was identified as an heterogeneous complex lesion (red arrow) probably affecting fetal chest wall and ribs (white arrow). A thoracic tumor with chest wall affection was reported instead of a CPAM what completely changed the perinatal management.
**Fig. 14:** Mesenchymal hamartoma. Postnatal chest plain film of the same patient of Fig. 12 and 13. Right hemithorax was completely opacified by the presence of a complex mass with calcifications inside. Multiple right ribs were also affected. This rare congenital tumor of the chest wall has a typical appearance in plain film. Final diagnosis was made by CT and histopathologic analysis (not shown).
Conclusion

Congenital cystic thoracic malformations are rare but when they appear is very important to reach an accurate diagnosis for a precise parental counseling and perinatal assessment.

These malformations are currently detected in utero by prenatal imaging. US remains the main technique to study fetal chest but when big or atypical cystic lesions are suspected fetal MRI has a supplemental role and can add valuable information.

We encourage careful attention to prenatal and postnatal imaging findings because Perinatal Imaging plays a central role in the adequate diagnosis and management of these patients.

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