Hyperechogenic solid masses of lungs: Prenatal diagnosis and outcome

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Learning objectives

The objectives of this work is to describe the accuracy of prenatal diagnosis by ultrasound Doppler color imaging and MRI and to review outcome of foetal hyperechogenic lung lesions.
Background

With current advances in both fetal ultrasonography (US) and magnetic resonance (MR) imaging, abnormalities of the thorax are increasingly being recognized antenatally. These chest lesions may be detected and diagnosed on ultrasound when there is an abnormal echogenicity of the lung compared with normal lung or the liver or when there is a really mass effect on the heart and mediastinum.

The most common fetal hyperechogenic lung lesions are congenital cystic adenomatoid malformation (CCAM), pulmonary sequestration (PS) congenital lobar emphysema (CLE) and congenital high airway obstruction syndrome (CHAOS).
Findings and procedure details

1/ Normal imaging features of fetal lung:

Normal fetal lung is visible at the end of the first trimester, but this study is easier from the second trimester. At US, the parenchyma is homogeneous echogenic. The echogenicity compared to the liver increases with gestational age (Figure 1). The trachea and bronchi are visualized more easily as hypoechoic linear images limited by two hyperechoic lines (Figure 2).

Normal lung, at MRI, is T2-weighted hyperintense compared to the muscle wall, but the signal is lower than fluid structures. Trachea and bronchi axis are T2-weighted hyperintense.

Compressed normal lung is hypointense compared to uncompressed normal lung; this could be related to the decrease of intra alveolar liquid in the compressed lung.

2/ Congenital cystic adenomatous malformation of the lung (CCAM):

CCAM is the most common developmental lung malformation diagnosed prenatally. It accounts for 50% to 75% of detected fetal lung abnormalities.

Prenatal diagnosis has dramatically increased as a result of improvement of US equipment in recent years.

It’s a heterogeneous group of cystic and noncystic lung lesions that largely result from early airway maldevelopment. At pathologic analysis, CCAM have been classified by Stocker according to cyst size and histologic resemblance to segments of the developing bronchial tree and airspaces.

The newer classification scheme includes five types and is an extension of the original scheme (three types):

Type 0 has a tracheal or bronchial origin and is really acinar dysgenesis or dysplasia. It is a very rare type which is incompatible with life.

Type 1 has a bronchial or bronchiolar origin. It’s characterized by single or multiple cysts greater than 2cm in diameter.

Type 2 has a bronchiolar origin formed by small cysts less than 2cm in diameter.

Type 3 has a bronchiolar-alveolar duct origin (adenomatoid type with cysts smaller than 5mm).
Type 4 has a distal acinar origin. It is represented by large, thin walled peripheral cysts caused by hamartomatous malformation of the distal acinus.

At imaging, there are only three types of CCAM which are distinguished: large cyst CCAM (type 1), macrocytic CCAM (type 2), and microcystic or solid type (type 3).

- **Ultrasound:** (Figure 3)

  Systematic evaluation of the lung parenchyma shows an abnormal echogenic lung mass. Once is identified, the location, size, existence of lung cyst, and blood supply must be evaluated using conventional spectral or power Doppler US.

  A CCAM is usually unilateral and unilobar with a slight predilection of lower lobes of the lung. The mass is usually detected in the second trimester.

  The study should include fetal echocardiography to screen for potential congenital heart malformations and comprehensive US evaluation to rule out associated anomalies and evaluate signs of fetal hydrops and early manifestations of cardiac failure.

  Classic signs include echogenic unilateral, well-defined, solid or cystic lung lesion which involves part of the lung with predilection to the lower lobe. Doppler study shows no feeding arterial vessels arising from the systemic circulation.

- **MRI:**

  MRI has been used to evaluate fetal lung masses but has not shown a substantial advantage over US. It shows a homogeneously hyperintense solid mass with normal adjacent lung parenchyma.

  A fast-growing CCAM may cause mediastinal shift with displacement of the heart to the contralateral part of the thorax, flattening of the diaphragm and subsequent development of polyhydramnios and hydrops.

  In microcystic CCAM with no hydrops, the survival rate is more than 95% without the need for antenatal intervention. In 50% of cases there is apparent antenatal resolution of the hyperechogenic lesion, usually at around 32 weeks of gestation.

  Prognosis is generally good as long as there is adequate residual normal lung tissue.

3/ Pulmonary sequestration (PS):

PS is the second most common lung lesion detected antenatally. It is characterized by a portion of lung that does not connect to the tracheobronchial tree and has a systemic arterial supply, usually from the thoracic or abdominal aorta.
Two types of sequestration have been described: intralobar and extralobar. The extralobar form, which is the most commonly diagnosed in the prenatal, has its own pleural investment and systemic venous drainage, whereas the intralobar form shares the pleural investment with the normal lung and usually drains into the pulmonary venous system.

Extralobar PS can be positioned in the chest cavity, above the diaphragm, with slight predominance in the left hemithorax (up to 50%); located in anterior and posterior mediastinum (8% and 6%); or located outside the chest cavity, below the diaphragm (up to 18% of cases).

- **Ultrasound:** (Figure 4)

At prenatal US, extralobar pulmonary sequestration is seen as a homogeneous hyperechoic mass in a paraspinal location, most often the left lower thorax. The feeding artery originating from the descending aorta may be seen at color Doppler US.

In some cases of extralobar PS can be situated below the diaphragm, making it difficult to distinguish PS from suprarenal masses, such as mesonephric blastoma or neuroblastoma.

In cases of an intrathoracic mass, the distinction between PS and the microcystic form of (CCAM) is very difficult especially when aberrant vessels may not be identified at Doppler US.

- **MRI:**

Prenatal MR imaging shows a solid, well-defined, uniformly hyperintense mass on T2-weighted images and the feeding artery may be identified.

The natural history of PS depends on the size and location of the mass. Generally it has a high rate of spontaneous resolution (75%).

Neonatal outcome depends on several factors, as follows:

- Mass size and position: Large intrathoracic masses may lead to lung hypoplasia, mediastinal shift, and fetal hydrops owing to blood vessel compression. Intraabdominal ELS generally has a better prognosis because it very rarely leads to fetal hydrops, and it usually has no mass effect on developing lung and so does not cause lung hypoplasia.

- Development of fetal hydrops.

- Presence of associated anomalies.

**4/ Congenital lobar overinflation or emphysema (CLE):**
CLE is characterized by progressive lobar overexpansion, usually with compression of the remaining (ipsilateral) lung. The underlying cause can be secondary to an intrinsic cartilaginous abnormality with resultant weak or absent bronchial cartilage or extrinsic compression of an airway. Congenital lobar emphysema is usually diagnosed in the neonatal period with respiratory distress.

The left upper lobe is involved in 42.2% of cases, the right middle lobe in 35.3%, the right upper lobe in 20.7%, and either lower lobe in less than 1%.

**Ultrasound:**

The main fetal sonographic features of CLE include a bright echogenic lung with or without cystic or mixed cystic lesions without abnormal blood flow. A mediastinal shift, polyhydramnios, and fetal hydrops can also be seen and are predictors of severe respiratory distress or mortality.

**MRI:**

Fetal MR imaging may demonstrate a mass with homogeneously high signal intensity on T2-weighted images.

CLE may be associated with cardiovascular anomalies in 14% of cases and uncommonly, renal, gastrointestinal, musculoskeletal, and cutaneous malformations.

Often CLE, CPAM, and PS are described as a single clinical group representing a spectrum of the same disease which can decrease in size during pregnancy.

### 5/ Congenital high airway obstruction syndrome (CHAOS):

CHAOS is a rare congenital anomaly caused by an obstruction of the upper airway tract. The causes of obstruction include laryngeal atresia, stenosis or laryngeal cysts, and tracheal atresia or stenosis. Congenital high airway obstruction syndrome should be distinguished from extrinsic causes of tracheolaryngeal obstruction such as lymphatic malformation, cervical teratoma, and vascular rings like double aortic arch [8]

**Physiopathology:**

The obstructed airway results in decreased clearance of the fluid produced by foetal lungs and increased intratracheal pressure which causes the lungs to expand and develop abnormally.

This causes thinning of the alveolar walls, reduction of Type II pneumocytes, and reduced surfactant. This further leads to hyperexpanded lungs which cause compression of the heart and inferior vena cava. Ultimately, these events culminate in decreased venous return and lead to non-immune hydrops [10].
• Ultrasound: (Figure 5)

Prenatal US shows enlarged symmetrical echogenic lungs and their enlargement relative to the chest wall may cause flattening or inversion of the diaphragm. The proximal airway appears dilated up till the level of the obstruction. The heart appears small compared with the enlarged lungs and appears somewhat anteriorly positioned in the chest due to compression by the lungs. Hydrops may be present. Fetal MR imaging in demonstrates enlarged, hyperintense lungs with flattened or inverted hemidiaphragms. Visualization of a dilated airway helps localize the level of obstruction.

• MRI:

The MRI findings include increased lung volumes with abnormally increased signal. There may be flattening or inversion of the diaphragm, small anteriorly displaced heart with centrally positioned axis, ascites, and other features of non-immune hydrops.

The MRI also shows a dilated airway up to the level of the obstruction and is better at identifying the level of obstruction due to higher intrinsic soft tissue contrast.
**Fig. 1:** Ultrasound Imaging in fetuses with normal lungs. Echogenicity compared to the liver increases with gestational age.

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**Fig. 2:** Transverse section of the fetal thorax demonstrating the fluid-fluid trachea.

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Fig. 3: Transverse section of a 24 weeks gestation: Type III CPAM (asterisk), displacing the heart, right and the left lung in the left side. h: heart, RL: right lung, LL: left lung.

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Fig. 4: sagittal US view in a fetus in 21 weeks gestation: Left-sided hyperechoic triangular basi pulmonary mass.

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Fig. 5: Bilateral pulmonary hypertrophy (asterisk) responsible for a reversal of the diaphragm (arrows) associated with a high abundance of intra peritoneal effusion.

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Conclusion

The evolution of better antenatal imaging has been crucial in diagnosis of foetal hyperechogenic lung lesions. Recognizing the US and MR imaging features of these abnormalities is necessary for prenatal counseling and appropriate peri- and postnatal management.
References


