Fetal Magnetic Resonance Imaging of Congenital Chest Malformations: A Pictorial Review

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ABSTRACT

Congenital chest malformations may affect the foregut, pulmonary airway, vasculature, chest wall, and mediastinum. An understanding of fetal chest masses and their complications is essential for patient counseling, appropriate management of pregnancy, delivery, and neonatal care planning. Ultrasonography is the primary imaging modality used for the prenatal evaluation of chest malformations. Magnetic resonance imaging (MRI) is a well-established and complementary diagnostic tool in cases of inconclusive and inadequate findings of ultrasonography, particularly for the assessment of complex fetal abnormalities. In this study, we have reviewed the fetal MRI technique as well as discussed and illustrated the common congenital chest abnormalities observed by fetal MRI.

Congenital chest malformations may involve the lung parenchyma, bronchi, vasculature, and rarely, the chest wall and mediastinum [1, 2]. The most common congenital chest anomalies include congenital pulmonary airway malformation (CPAM), bronchopulmonary sequestration (BPS), congenital lobar fluid overload (CLFO), congenital diaphragmatic hernia (CDH), and congenital pleural effusions. Less common diseases include chest wall lymphangioma, congenital bronchogenic cyst, esophageal duplication cyst, pulmonary hypoplasia or aplasia, bronchial atresia, congenital high airway obstruction syndrome (CHAOS), pulmonary arteriovenous malformation (PAVM), and congenital pulmonary lymphangieectasia [2-6]. In this study, we have reviewed the fetal MRI technique and discussed and illustrated the common congenital chest abnormalities observed by fetal MRI.

MRI Technique

MRI is performed using a 1.5 T MR scanner (Twin-speed, GE Medical Systems, Milwaukee, WI), torso phased-array coil, and single-shot fast-spin echo sequence. The ultrafast T2-weighted images are the most useful for evaluating the lung anatomy and abnormalities. The imaging protocol consists of a single-shot fast-spin echo sequence following the axial, coronal, and sagittal planes of the fetal chest (TE: 90 ms, TR: minimum, field of view: 270 × 270 mm, matrix: 256 × 224, slice thickness: 4 mm, scan time: 34 s/19 slices). All image sequences are performed without maternal breath holding. The entire examination does not exceed 30 min.

Normal Fetal Lungs on T2-weighted Images

The trachea, bronchi, and lungs reveal homogeneous hyperintensity on T2-weighted images in comparison to chest wall muscles, because they contain a significant amount of amniotic and alveolar fluid. An increase in the signal intensity of the lungs can be observed as the lungs mature, and a relative hypointensity is found in cases of pulmonary underdevelopment owing to the decreased amount of alveolar fluid.
Types of Congenital Chest Malformations

**Congenital Pulmonary Airway Malformation**

CPAM is the most common congenital lung malformation and accounts for 30-40% of all congenital chest diseases [2, 3, 6, 7]. It is characterized by adenomatoid proliferation of bronchiole-like cysts in the lung and a lack of normal alveolar development [7]. The term CPAM has been renamed as being preferable to the term congenital cystic adenomatoid malformation, since the lesions are cystic in only three of five types of these lesions and adenomatoid in only one type [4, 7]. These malformations may consist of cystic and solid components, resulting in various MRI manifestations.

The findings of MRI depend on the size of the cysts. Typically, the signal intensity of the cysts is higher than that of the surrounding normal lung parenchyma [8-10]. Macro-cystic CPAM appears as a lobulated mass with inhomogeneous hyperintensity (Fig. 1) [2, 9, 10], and microcystic CPAM as a lobulated mass of homogeneous hyperintensity with arterial vascular architectural distortion without visible cysts (Fig. 2) [6, 10]. Regression (either partial or nearly complete) of the masses in utero appears to be the rule. Regressed CPAM usually appears as an ill-defined inhomogeneous mildly hyperintense mass (as compared to the normal lung), which shows decreased signal intensity on follow-up MRI (Fig. 3) [9, 10]. The differential diagnosis for CPAM includes congenital bronchogenic cyst (single macrocystic CPAM), BPS, and CLFO (microcystic CPAM).

**Bronchopulmonary Sequestration**

BPS consists of nonfunctioning pulmonary tissue, which fails to connect to the normal tracheobronchial tree and receives systemic blood supply [9-13]. It is enveloped by the visceral lung pleura (intralobar subtype) or has its own visceral lung pleura (extralobar subtype). The majority of cases frequently occur in the lower lobes, especially in the left lower lobe [2-4].

MRI findings of pure BPS include a well-defined triangular homogeneous hyperintense mass with a higher signal intensity than that of a normal lung, but with lower signal intensity than that of the amniotic fluid, as well as visualization of the systemic feeding artery (Fig. 4) [2, 4, 8, 10, 11]. In the hybrid lesions of BPS and CPAM, the margin may be lobulated, and the signal is inhomogeneous and hyperintense (Fig. 5) [2, 6, 10]. BPS may also regress either partially or completely in utero. Regressed BPS tends to have a lobulated margin, with decreased and inhomogeneous signal intensity (Fig. 6) [10, 12]. The differential diagnosis includes microcystic CPAM and CLFO, and systemic arterial supply is the key finding for BPS [2, 3, 10, 11].

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**Figure 1.** Macro-cystic CPAM in a 25-week-old fetus. Axial MRI shows a lobulated, multicystic mass with inhomogeneous hyperintensity (arrows) in the right lower lobe causing mild compression of the heart toward the left (arrowhead).

**Figure 2.** Micro-cystic CPAM in a 23-week-old fetus. Sagittal MRI shows a lobulated mass of homogeneous hyperintensity without visible cysts (arrow).
Figure 3. Regressed CPAM in a 35-week-old fetus. Follow-up MRI shows a small, inhomogeneous, mildly hyperintense mass (arrow) in the right lung base, with decreased signal intensity, compared to the initial MRI results depicted in figure 1.

Figure 4. Pure BPS in a 24-week-old fetus. Coronal MRI shows a triangular, homogeneous, hyperintense mass in the left lower lobe (arrows) and visualization of the systemic feeding artery (arrowhead).

Figure 5. Hybrid lesion of BPS and CPAM in a 25-week-old fetus. Coronal MRI shows a well-defined, homogeneous, hyperintense mass in the left thorax (arrows), visualization of the systemic feeding artery (arrowhead), and a hyperintense cystic lesion in the left upper lobe (asterisk).

Figure 6. Regression BPS in a 35-week-old fetus. Follow-up MRI shows a regressed mass with a lobulated margin, and a decrease and inhomogeneity in signal intensity (arrows) compared to the initial MRI results shown in figure 4.
Congenital Lobar Fluid Overload

CLFO is a phenomenon that occurs because of alveolar fluid trapped in the affected lung segment or lobe due to a ball-valve mechanism, which is synonymous with congenital lobar emphysema seen postnatally. CLFO occurs when transient mucus plug impaction or extrinsic compression by a mass lesion or cartilage abnormalities causes bronchial narrowing; it usually involves the left upper and right middle lobes [6, 14, 15]. However, CLFO may progress, improve, or disappear completely in utero, depending on the etiology of the blockage [10, 15-18].

CLFO appears as a fluid-overloaded, expanded, but structurally intact lung segment or lobe causing mediastinal compression (Fig. 7). On MRI, CLFO can be differentiated from microcystic CPAM, BPS, and bronchial atresia due to its homogeneity and intact lung segment or lobe with stretched hilar vessels without pulmonary architectural distortion [6, 14, 15].

Congenital Diaphragmatic Hernia

In CDH, a diaphragmatic defect causes herniation of the abdominal viscera (such as the liver, stomach, and bowel) into the thorax (Fig. 8), leading to lung developmental deficiencies, and it predominantly occurs on the left side [2-4]. Lung hypoplasia is found most frequently in the ipsilateral lung, and the contralateral lung can be involved.
by mediastinal shift and compression [2-4]. Kilian et al. observed that fetuses with an MRI relative fetal lung volume less than 14.3% died, whereas all neonates with an MRI relative fetal lung volume greater than 32.8% survived, and no neonate with a relative volume greater than 44% needed extracorporeal membrane oxygenation [19].

**Congenital Pleural Effusions**

Congenital pleural effusions are abnormal accumulations of fluid in the pleural cavity. Massive pleural effusions can cause poor development of the lungs or heart failure. The primary cause of congenital pleural effusion is chylothorax (abnormal lymphatic drainage) and the secondary causes include infection, heart conditions, genetic or chromosome problems, and other lung problems [2, 3]. MRI of fetal pleural effusion reveals fluid collection surrounding the lungs (Fig. 9). The T1- and T2-weighted images can differentiate the pleural effusion content: transudate, rich in lipid, and protein or blood products, and may help to make the etiological diagnosis. MRI can also provide more information that may be required to investigate the associated abnormalities [2, 3, 20], which are present in approximately 40% of fetuses.

**Chest Wall Lymphangiomas**

Chest wall lymphangiomas develop because of inefficient connections between the lymphatic and venous pathways and carry a low incidence of chromosomal and structural anomalies, as compared to nuchal lymphangiomas [21, 22]. On MRI, chest wall lymphangiomas appear as well-defined unilocular or multilocular homogeneous hyperintense cystic masses, with or without septation (Fig. 10). MRI can provide more information regarding the tumor extent and tissue characteristics as well as allow differential diagnosis of this type of malformation [22-25].

**Congenital Bronchogenic Cysts**

Congenital bronchogenic cysts are caused by abnormal budding of the embryonic foregut and subsequent differentiation into a fluid-filled, blind-ended cyst typically located in the mediastinum and less commonly within the lung parenchyma, pleura, or diaphragm [2]. Fetal MRI helps in determining the location of the lesion and usually shows marked hyperintensity (Fig. 11) [2, 4].

**Esophageal Duplication Cyst**

Esophageal duplication cyst is the second most common duplication in the gastrointestinal tract and can rarely be diagnosed in the fetus [26]. A large esophageal duplication cyst in a fetus may manifest with polyhydramnios and pleural effusion due to compression of the fetal esophagus and mediastinum, and consequently, it may
obstruct the physiological swallowing of amniotic fluid by the fetus [27]. On MRI, an esophageal duplication cyst appears as a well-defined, round or tubular, homogeneous hyperintense cystic lesion at the posterior mediastinum (Fig. 12) [27, 28].

**Pulmonary Hypoplasia**

The majority of cases of pulmonary hypoplasia are secondary to a mass effect, which may be of intrathoracic or extrathoracic origin, limiting lung development in the thoracic cavity. The most common cause of pulmonary hypoplasia is congenital diaphragmatic hernia (Fig. 13) [2]; other causes include abnormal fetal breathing movements, abnormal amniotic fluid volume (Fig. 14), and abnormal fetal lung fluid pressure [4]. Diffuse homogeneous hypointensity of the hypoplastic lung, in comparison to the surrounding amniotic fluid, can be observed on MRI [29].

**Bronchial Atresia**

Bronchial atresia results from interruption of focal developmental of a lobar, segmental, or subsegmental bronchus and associates with the collateral alveolar fluid drift (not “fluid trapping” in CLFO) over the surrounding lung parenchyma through the intra-alveolar pores of the Kohn, bronchoalveolar channels of Lambert, and interbronchial channels [2, 4]. The apicoposterior segment of the left upper lobe is most often involved. Bronchial atresia and CLFO

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**Figure 10.** Chest wall lymphangioma in a 26-week-old fetus. Coronal MRI shows a well-defined unilocular, hyperintense cystic mass over the left chest wall (arrows).

**Figure 11.** Congenital bronchogenic cyst in a 25-week-old fetus. Sagittal MRI shows a marked hyperintense cyst (asterisk) within the lung parenchyma near the mediastinum.

**Figure 12.** Esophageal duplication cyst in a 28-week-old fetus. Sagittal MRI shows a well-defined, homogeneous, hyperintense cystic lesion at the posterior mediastinum (arrows), just above the normal lung parenchyma and anterior to the thoracic aorta (asterisk).
usually have the same fetal MRI finding of fluid-overloaded and expanded lung parenchyma due to difficulty in visualization of the atretic bronchi [2, 10]. Postnatal computed tomography is recommended to determine the tubular structure of the atretic bronchi near the hilum.

**Congenital High Airway Obstructive Syndrome**
CHAOS is caused by complete or near-complete airway obstruction; it is associated with a poor prognosis, causing outflow obstruction of the fetal lung fluid, pulmonary hyperplasia, and tracheal dilatation [30]. Fetuses with CHAOS may develop hydrops due to cardiac compression and obstruction of venous return. MRI in CHAOS shows enlarged, hyperintense lungs with flattened or inverted hemidiaphragms and massive ascites. Visualization of the dilated airway helps to localize the obstruction [2, 30, 31].

**Pulmonary Arteriovenous Malformation**
PAVM is caused by abnormal communication between the pulmonary arteries and veins [32]. Fetal MRI is helpful in identifying multiple lesions and shows a hypointense irregular lesion, usually located near the hilum [2, 32].

**Congenital Pulmonary Lymphangiectasia**
Congenital pulmonary lymphangiectasia probably results from pulmonary interstitial connective tissue failure, leading to dilatation of the pulmonary lymphatic vessels [33]. Fetal MRI shows bilateral inhomogeneity of the lung parenchyma and hyperintense linear structures, suggesting engorged lymphatic vessels [2, 34].

**CONCLUSIONS**
MRI is a helpful and complementary diagnostic tool adjunct to ultrasonography for the detection, characterization, and evaluation of small to extensive fetal chest malformations. This tool is essential for both prenatal counseling of the parents and to guide perinatal and postnatal management in selective patients when ultrasonographic findings are inconclusive or inadequate.
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