

SPECIALTY AREA: Fetal Anomalies in the Body

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Highlights

-Fetal MR evaluation of the thorax is performed for evaluation of congenital diaphragmatic hernia, and congenital lung lesions, offering certain prognostic predictors of survival.

-MR of the Abdomen is performed less frequently to evaluate abnormalities of the hepatobiliary system, gastrointestinal tract, genitourinary tract and complex dysmorphology of the abdominal wall; certain MR findings offer important diagnostic information.

This information is intended as a review of indications of fetal MR imaging of the body as pertains to dysmorphology.

The purpose of this of presentation is to review the normal and abnormal findings on the MR fetal body examination and discuss the common indications and prognostic value.

Thorax

Congenital diaphragmatic hernia (CDH) is the principal indication for fetal thoracic imaging. Numerous articles have compared ultrasound with MR in predicting outcomes and survival in fetuses with congenital diaphragmatic hernia. They calculated lung volumes and degree of liver herniation, major prognostic indicators for CDH, as postnatal morbidity and mortality is primarily due to pulmonary hypoplasia and pulmonary hypertension¹⁻⁸. Specifically, MR can depict herniated contents in a CDH. One recent study compared ultrasound(US) and MR measurements, evaluating US parameters of lung to head ratio (LHR) and observed to expected LHR (O/E LHR) to the MR observed to expected total lung volume (O/E-TLV) and the percent herniated liver(%HL)⁷. Receiver operator curves were constructed based on univariable logistic regression to predict survival. Both MR variables had significantly better areas under the curve than the US parameters (p=0.02). The liver is well demarcated on T1 weighted images, and discrimination of large versus small bowel can be performed owing to T1 bright appearance of meconium in large bowel⁹.

Congenital lung lesions encompass a spectrum of abnormalities, including congenital pulmonary airway malformation (CPAM), bronchopulmonary sequestration (BPS), and congenital lobar overinflation (CLO). CPAMs account for approximately 50% of all lesions, and contain either macrocystic or microcystic components. Blood supply is via the pulmonary artery and vein. BPS account for approximately 1/3 of lesions and consist of nonfunctioning bronchopulmonary tissue with systemic blood supply, the majority are supra-diaphragmatic (85-90%), with 10-15% in an infra-diaphragmatic location. Of all BPS masses, ¾ will be hybrid lesions, with components of both CPAM and sequestration. CLO results from bronchial obstruction of a main or lobar bronchus, either from mucus plug or extrinsic compression due to vessel or mass. Macrocystic CPAMs tend to be more heterogeneous, multilocular, and cystic in appearance with architectural distortion. Microcystic CPAM is homogeneous but also demonstrates architectural distortion. BPS lesions appear homogeneous, without architectural distortion, and if a feeding vessel is visualized can be diagnosed with confidence as a BPS. Hybrid lesions demonstrate features common to both CPAM and BPS. CLO appears uniformly hyperintense on T2 weighted imaging secondary to fluid overdistension of segments and lobes¹⁰⁻¹². MR plays a complementary role to ultrasound, confirming or providing alternative diagnoses.

Abdomen

Abnormalities in the fetal abdomen are an infrequent indication for MR evaluation. Fetal abdominal pathology encompasses abnormalities of the hepatobiliary system, GI tract, GU tract, and abdominal-pelvic masses.

Hyperintense T1 signal within bowel loops represents meconium usually seen in distal ileum or colon¹³. T2 hyperintense bowel dilatation usually indicates proximal small bowel dilatation, and T1 hyperintense bowel dilatation has been thought to represent distal ileal or colonic occlusion¹⁴. However a recent study demonstrated proximal jejunal atresia in 9 cases of suspected ileal atresia, signifying the imprecision of determining the exact site of occlusion¹⁵.

Genitourinary anomalies are well demonstrated on MR, and imaging is not hindered by oligohydramnios. Renal cystic disease, duplications, hydroureteronephrosis, megacystis-microcolon-intestinal hypoperistalsis syndrome, and cloacal exstrophy with 2 characteristic hemibladders can be visualized¹⁶. Furthermore, identification of signal void compared to bright signal in the bladder on T-2 weighted imaging has been found to be very discriminating ($p < 0.001$) in the prediction of severe and lethal renal abnormalities from those that result in a non-lethal outcome¹⁷.

Cystic masses within the abdominal cavity may be categorized according to location, upper abdominal, lower abdominal, and retroperitoneal. The majorities of masses are lower abdominal and are urogenital in origin, and gynecologic in origin, including ovarian cysts and hydrometrocolpos. Upper abdominal cysts include choledochal, mesenteric, hepatic, and splenic cysts. Retroperitoneal masses include lymphangioma, adrenal cysts, and adrenal neuroblastoma. MR has shown benefit in evaluating cystic abdominal masses with respect to not only improved tissue characterization but also anatomical localization¹⁸.

Abdominal wall defects include gastroschisis, omphalocele, cloacal exstrophy, limb body wall complex (LBWC), and pentalogy of Cantrell. Recognition of common features of each entity and potential pitfalls are essential for proper diagnosis. Ruptured omphaloceles may mimic gastroschisis, congenital hernia of the umbilical cord may mimic omphalocele, and a dilated vagina may be mistaken for a normal urinary bladder in cases of cloacal abnormality¹⁹. MR is beneficial in visualizing omphalocele sac content, size of anterior abdominal wall defect, quantity of extruded bowel, suggestion for bowel atresia, and presence of additional anomalies²⁰. Omphalocele may be isolated, or part of a severe anomaly, including cloacal exstrophy, LBWC, and pentalogy of Cantrell. Fetal MR imaging is particularly helpful in defining the more complex lesions and differentiation of treatable abdominal wall defect from lethal entities can help guide appropriate counseling and management²¹.

1. Lazar DA, Ruano R, Cass DL, Moise KJ, Jr., Johnson A, Lee TC, Cassady CI, Olutoye OO. Defining "liver-up": does the volume of liver herniation predict outcome for fetuses with isolated left-sided congenital diaphragmatic hernia? *Journal of pediatric surgery*. Jun 2012;47(6):1058-1062.
2. Debus A, Hagelstein C, Kilian AK, Weiss C, Schonberg SO, Schaible T, Neff KW, Busing KA. Fetal lung volume in congenital diaphragmatic hernia: association of prenatal MR imaging findings with postnatal chronic lung disease. *Radiology*. Mar 2013;266(3):887-895.
3. Walleyo A, Debus A, Kehl S, Weiss C, Schonberg SO, Schaible T, Busing KA, Neff KW. Periodic MRI lung volume assessment in fetuses with congenital diaphragmatic hernia: prediction of survival, need for ECMO, and development of chronic lung disease. *AJR. American journal of roentgenology*. Aug 2013;201(2):419-426.
4. Nawapun K, Eastwood M, Sandaite I, DeKoninck P, Claus F, Richter J, Rayyan M, Deprest J. The correlation between the observed-to-expected total fetal lung volume and intra-thoracic organ herniation on magnetic resonance images in fetuses with isolated left-sided congenital diaphragmatic hernia. *Ultrasound in obstetrics & gynecology : the official journal of the International Society of Ultrasound in Obstetrics and Gynecology*. Oct 21 2014.

5. Ruano R, Lazar DA, Cass DL, Zamora IJ, Lee TC, Cassady CI, Mehollin-Ray A, Welty S, Fernandes CJ, Haeri S, Belfort MA, Olutoye OO. Fetal lung volume and quantification of liver herniation by magnetic resonance imaging in isolated congenital diaphragmatic hernia. *Ultrasound in obstetrics & gynecology : the official journal of the International Society of Ultrasound in Obstetrics and Gynecology*. Jun 2014;43(6):662-669.
6. Bebbington M, Victoria T, Danzer E, Moldenhauer J, Khalek N, Johnson M, Hedrick H, Adzick NS. Comparison of ultrasound and magnetic resonance imaging parameters in predicting survival in isolated left-sided congenital diaphragmatic hernia. *Ultrasound in obstetrics & gynecology : the official journal of the International Society of Ultrasound in Obstetrics and Gynecology*. Jun 2014;43(6):670-674.
7. Zamora IJ, Olutoye OO, Cass DL, Fallon SC, Lazar DA, Cassady CI, Mehollin-Ray AR, Welty SE, Ruano R, Belfort MA, Lee TC. Prenatal MRI fetal lung volumes and percent liver herniation predict pulmonary morbidity in congenital diaphragmatic hernia (CDH). *Journal of pediatric surgery*. May 2014;49(5):688-693.
8. Worley KC, Dashe JS, Oliver Q, Megison S, Barber R, McIntire DD, Twickler DM. Magnetic resonance imaging as a predictor of outcome in fetuses with isolated congenital diaphragmatic hernia. *American Journal of Obstetrics and Gynecology*. Dec 2006;195(6):S78-S78.
9. Zizka J, Elias P, Hodik K, Tintera J, Juttnerova V, Belobradek Z, Klzo L. Liver, meconium, haemorrhage: the value of T1-weighted images in fetal MRI. *Pediatric radiology*. Aug 2006;36(8):792-801.
10. Recio Rodriguez M, Martinez de Vega V, Cano Alonso R, Carrascoso Arranz J, Martinez Ten P, Perez Pedregosa J. MR imaging of thoracic abnormalities in the fetus. *Radiographics : a review publication of the Radiological Society of North America, Inc*. Nov-Dec 2012;32(7):E305-321.
11. Pacharn P, Kline-Fath B, Calvo-Garcia M, Linam LE, Rubio EI, Salisbury S, Brody AS. Congenital lung lesions: prenatal MRI and postnatal findings. *Pediatric radiology*. Sep 2013;43(9):1136-1143.
12. Barth RA. Imaging of fetal chest masses. *Pediatric radiology*. Jan 2012;42 Suppl 1:S62-73.
13. Farhataziz N, Engels JE, Ramus RM, Zaretsky M, Twickler DM. Fetal MRI of urine and meconium by gestational age for the diagnosis of genitourinary and gastrointestinal abnormalities. *AJR. American journal of roentgenology*. Jun 2005;184(6):1891-1897.
14. Saguintaah M, Couture A, Veyrac C, Baud C, Quere MP. MRI of the fetal gastrointestinal tract. *Pediatric radiology*. Jun 2002;32(6):395-404.
15. Colombani M, Ferry M, Garel C, Cassart M, Couture A, Guibaud L, Avni F, Gorincour G. Fetal gastrointestinal MRI: all that glitters in T1 is not necessarily colon. *Pediatric radiology*. Jul 2010;40(7):1215-1221.
16. Caire JT, Ramus RM, Magee KP, Fullington BK, Ewalt DH, Twickler DM. MRI of fetal genitourinary anomalies. *AJR. American journal of roentgenology*. Nov 2003;181(5):1381-1385.
17. Hawkins JS, Dashe JS, Twickler DM. Magnetic resonance imaging diagnosis of severe fetal renal anomalies. *American Journal of Obstetrics and Gynecology*. Mar 2008;198(3).
18. Gupta P, Sharma R, Kumar S, Gadodia A, Roy KK, Malhotra N, Sharma JB. Role of MRI in fetal abdominal cystic masses detected on prenatal sonography. *Archives of gynecology and obstetrics*. Mar 2010;281(3):519-526.
19. Nakagawa M, Hara M, Shibamoto Y. MRI findings in fetuses with an abdominal wall defect: gastroschisis, omphalocele, and cloacal exstrophy. *Japanese journal of radiology*. Mar 2013;31(3):153-159.
20. Sugai Y, Hosoya T, Kurachi H. MR imaging of fetal omphalocele: a case report. *Magnetic resonance in medical sciences : MRMS : an official journal of Japan Society of Magnetic Resonance in Medicine*. 2008;7(4):211-213.
21. Aguirre-Pascual E, Epelman M, Johnson AM, Chauvin NA, Coleman BG, Victoria T. Prenatal MRI evaluation of limb-body wall complex. *Pediatric radiology*. Nov 2014;44(11):1412-1420.