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IMAGING

CLINICAL CASE

Fetal Cardiac Magnetic Resonance Imaging in Left-Sided Diaphragmatic Hernia

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ABSTRACT

BACKGROUND Magnetic resonance imaging (MRI) has become an essential complementary imaging tool during pregnancy.

CASE SUMMARY In this case of left-sided congenital diaphragmatic hernia (CDH), fetal cardiac MRI at 29 gestational weeks showed subtle septal bounce and reduced biventricular ejection fraction. On follow-up MRI at 37 gestational week, the right ventricle was significantly dilated (right-to-left ventricle index: 1.9:1.0), indicating increasing right ventricular strain and raising the suspicion of pulmonary hypertension (PH). Post delivery, the newborn required ventilation and circulatory support. Diagnosis of PH was confirmed by echocardiography. After surgical repair, the clinical course was unremarkable.

DISCUSSION This case illustrates that fetal cardiac MRI can provide a prenatal assessment of cardiovascular sequelae of CDH, thereby informing perinatal care.

TAKE-HOME MESSAGES Fetal MRI in CDH is used to accurately quantify fetal lung volume and to detect associated malformations. Additional cardiac assessment can indicate right ventricular strain and should raise suspicion of PH and prompt information of perinatal management. (JACC Case Rep. 2025;30:105295)

HISTORY OF PRESENTATION

We present the case of a fetus with left-sided congenital diaphragmatic hernia (CDH) in a 34-year-old healthy woman (G1; P0). Ultrasound (US) screening at 12 + 5 gestational weeks (GW) revealed a left-sided CDH with partial dislocation of the small and large bowel into the thoracic cavity and the liver located underneath the diaphragm. Cardiac size, axes, and rhythm were regular, with only mild tricuspid regurgitation noted. US examinations revealed a favorable lung-to-head ratio without an

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ABBREVIATIONS AND ACRONYMS

CDH = congenital diaphragmatic hernia

CSF = cerebrospinal fluid

EF = ejection fraction

ECMO = extracorporeal membrane oxygenation

GW = gestational weeks

LV = left ventricular

MRI = magnetic resonance imaging

PH = pulmonary hypertension

RLVR = right-to-left ventricular ratio

RV = right ventricular

SF = shortening fraction

indication for fetoscopic endoluminal tracheal occlusion.[1]

PAST MEDICAL HISTORY

Pre-existing conditions are hypothyroidism and mild beta-thalassemia.

DIFFERENTIAL DIAGNOSIS

Not applicable.

INVESTIGATIONS

A fetal magnetic resonance imaging (MRI) was performed at 28 + 1 GW for lung volume quantification and the assessment of potential associated malformations. For this indication, fetal MRI is implemented as a routine examination in CDH cases at late second or early third trimester at our institution to inform prognosis and guide perinatal management, for example, the need for extracorporeal membrane oxygenation (ECMO).[2,3] No associated malformations were detected. Brain development was unremarkable (**Figure 1A**), but we observed rather large extra-axial cerebrospinal fluid spaces compared to published reference values[4] in both scans (**Figure 1B**).

Fetal MRI confirmed left-sided CDH. T1-weighted images displayed meconium-containing bowel loops (**Figure 2A**). T2-weighted images showed stomach, spleen, and small intestine, all displaced into the left thoracic cavity, with the liver remaining underneath the diaphragm. Mediastinal structures were displaced to the right (**Figure 2B**). No left lung parenchyma was detectable. Manual segmentation of the right lung revealed a volume of 14 mL, corresponding to ~28% of age-adequate lung volume of approximately 50 mL.[5]

Fetal cardiac balanced steady-state free precession cine imaging was conducted in long- and short-axis orientation. Cardiac postprocessing was performed using CVI42 (circle cardiovascular imaging). Left- (LV) and right-ventricular (RV) diameters were assessed between the lateral and septal endocardial borders at a mid-papillary level.[6] The right-to-left ventricular ratio (RLVR) was calculated as the ratio of RV diameter to LV diameter,[7] and the shortening fraction (SF) per ventricle was calculated as (end diastolic diameter − end systolic diameter)/end diastolic diameter.[6] LV ejection fraction (EF) and SF as well as ventricular ratio are protocolized measures

TAKE-HOME MESSAGES

- Fetal magnetic resonance imaging in congenital diaphragmatic hernia is used to accurately quantify fetal lung volume and to detect associated malformations.
- Additional cardiac assessment can indicate right ventricular strain, should raise suspicion of pulmonary hypertension, and prompt information of perinatal management.

VISUAL SUMMARY Timeline of Prenatal and Perinatal Management

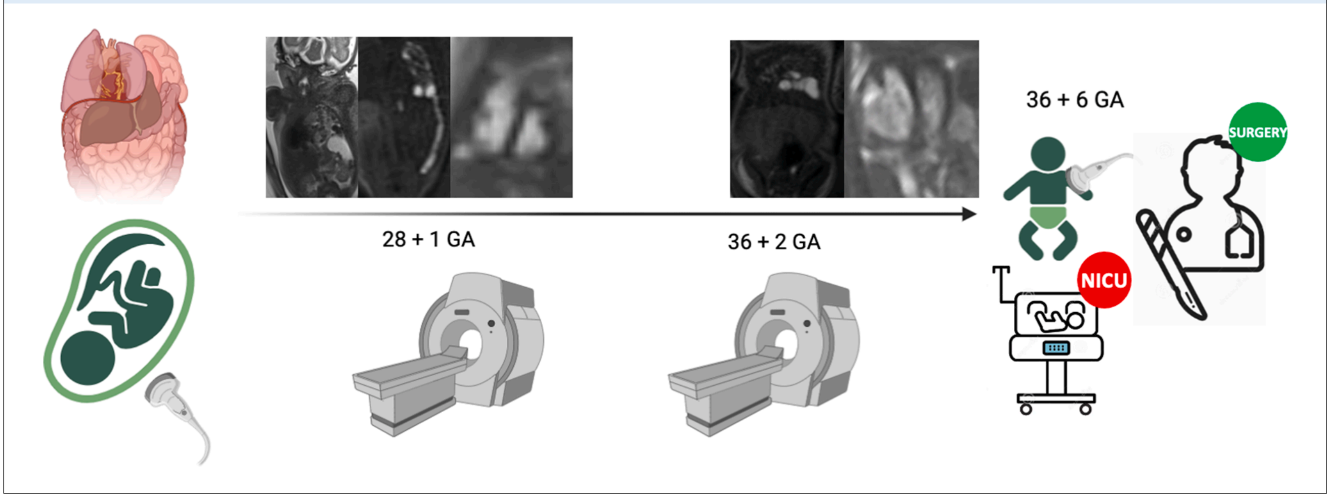
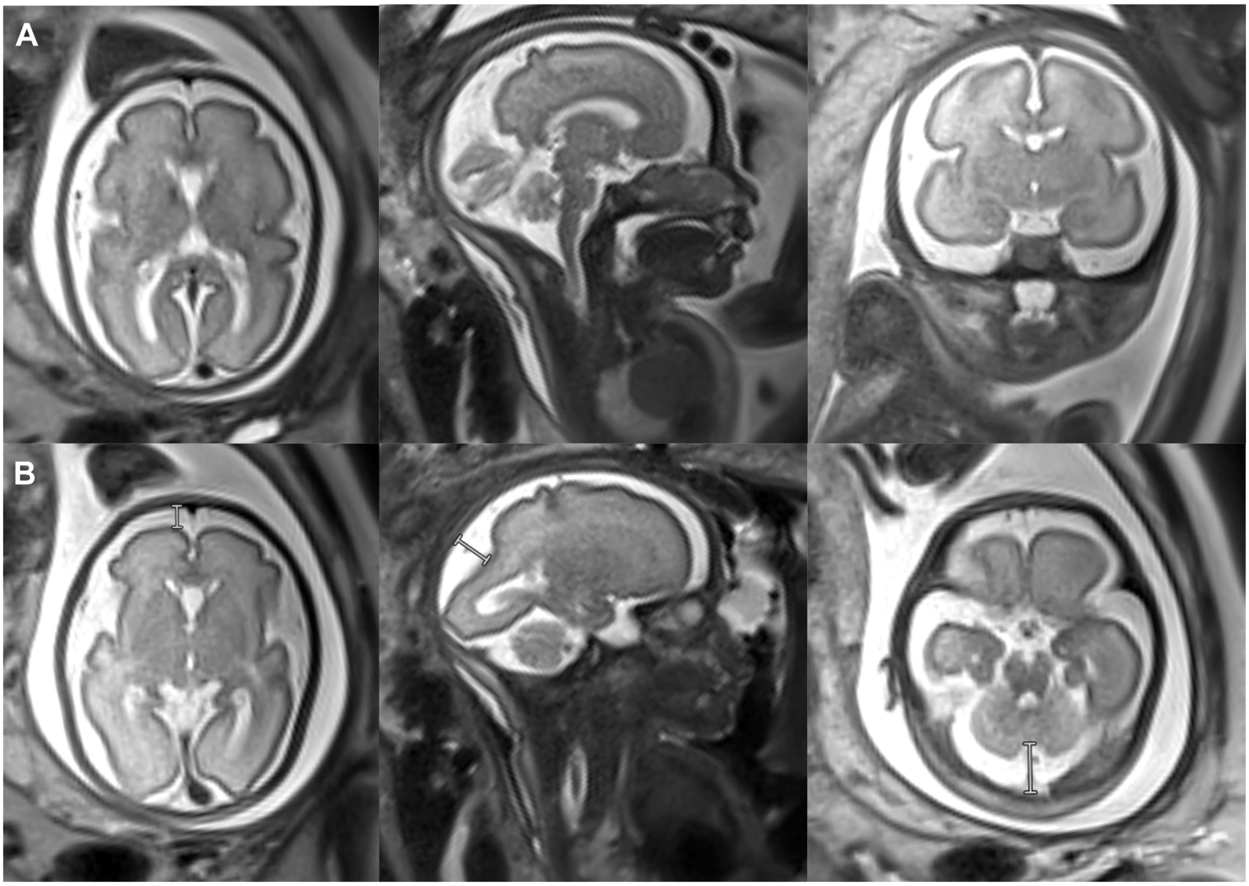


FIGURE 1 Images of the Fetal Brain at 29 GW



Representative T2-weighted images of the fetal brain at 29 GW in axial, sagittal, and coronal orientation demonstrating age-adequate fetal brain development (A) but enlarged extra-axial CSF spaces (B) (left frontal space = 6.5 mm, left posterior space = 11.7 mm, and cisterna magna = 14.4 mm as indicated in yellow). Values for follow-up scan (not shown in figure) at 37 GW: left frontal space 4.4 mm, left posterior space 10.9 mm, cisterna magna 14.2 mm; GW = gestational weeks; CSF = cerebrospinal fluid.

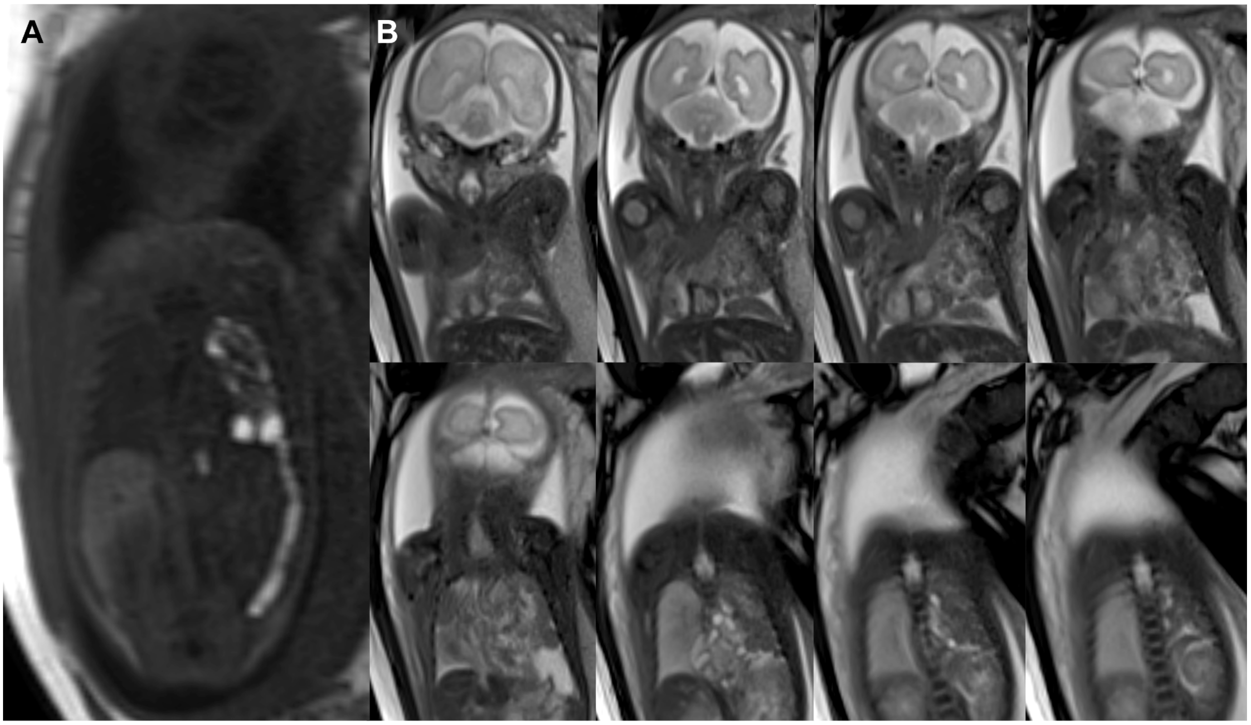
within the fetal cardiac MRI workflow. RV-EF and RV-SF were additionally evaluated given the RLVR and suspicion of RV strain.

Fetal cardiac MRI at 29 GW showed slightly reduced left- (49%) and RV (46%) EFs along with subtle septal movement disturbance and an elevated RLVR (1.2:1.0), raising suspicion of RV strain (Video 1, **Figure 3A**). LV-SF (23.53%) and RV-SF (17.35%) both ranged below reference values for the third trimester.[6] Following the first MRI as a routine examination at our institution, a second MRI in late third trimester was performed for follow-up of suspected RV strain at 36 + 2 GW. This showed a lung volume of 40 mL, corresponding to ~47% of age-adequate lung volume.[5] Ventricular EF were further reduced (44% LV, 45% RV), and RV enlargement had aggravated (RLVR 1.9:1.0) with clear septal bulging into the LV, indicating increasing RV strain and suggesting pulmonary hypertension (Video 2, **Figure 3B**). LV-SF (27.8%) and RV-SF (23.4%) both remained below reference ranges for the third trimester.[6]

MANAGEMENT

Following a cesarean section at 36 + 6 GW, the newborn was cyanotic and required immediate intubation. Blood gas analysis revealed acidosis with elevated pCO2. Blood pressure support with dopamine and noradrenaline was initiated, and the patient was admitted to the neonatal intensive care unit. Postnatal echocardiography confirmed persistent pulmonary hypertension, prompting therapy

FIGURE 2 Representative Images of the Fetal Body at 29 GW



(A) Representative T1w sequence in coronal orientation demonstrating meconium marking of intestinal loops in the left thoracic cavity and physiologically positioned meconium marking of the descending colon. (B) Representative balanced steady-state free precession (bSSFP) images in coronal orientation demonstrating the extent of herniated abdominal organs into the left thoracic cavity and displacement of the fetal heart to the right. GW = gestational weeks.

with inhaled nitric oxide. The diaphragmatic defect was closed by a standard Gore-Tex patch repair on the third day of life.

OUTCOME AND FOLLOW-UP

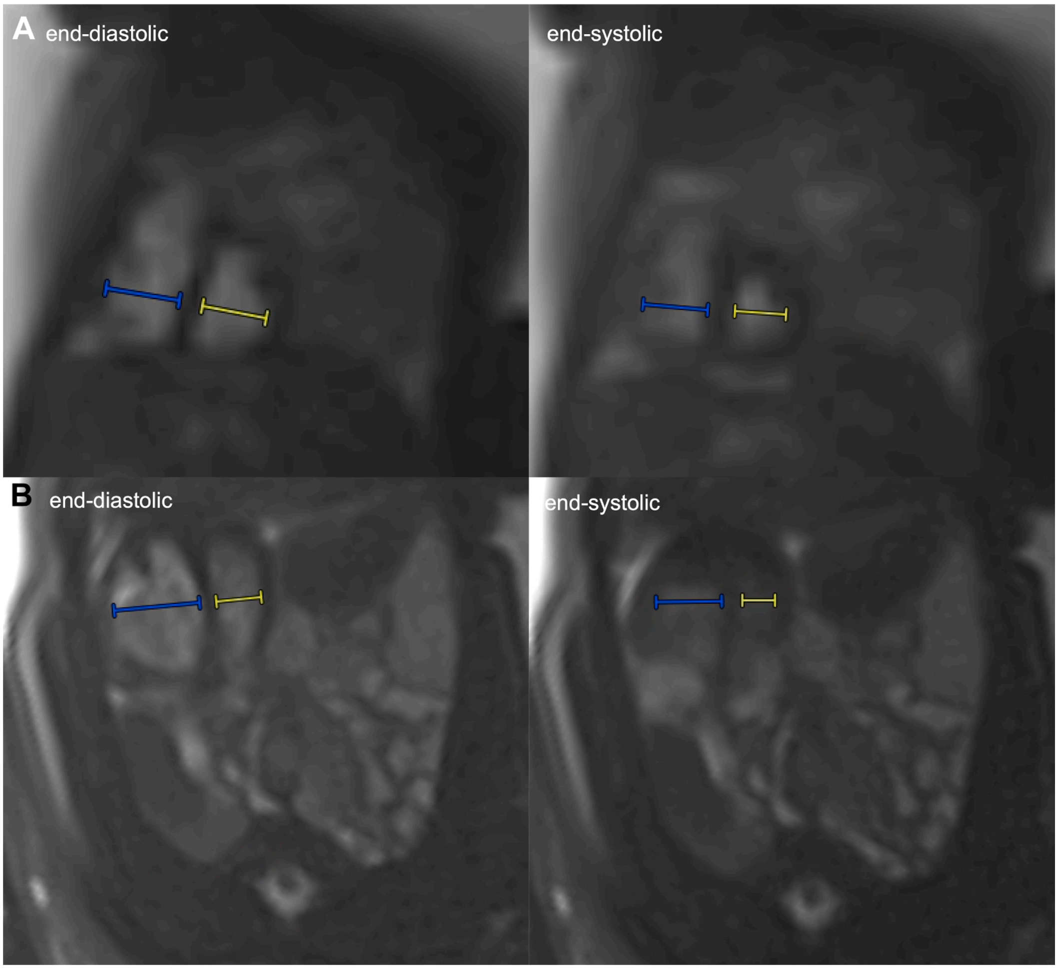
One month post operation, the newborn was discharged for outpatient follow-up, which proceeded in accordance with age-related expectations.

DISCUSSION

CDHs are common birth defects with a prevalence of 3/10,000 births[8] and are characterized by lung hypoplasia and abnormal pulmonary vascular development, often leading to significant morbidity and mortality.[9] Approximately 50% of CDH cases present additional anomalies, most frequently cardiovascular, which may imply a worse prognosis.[9] No associated extracardiac abnormalities were detected in this case; however, enlarged outer cerebrospinal fluid spaces may indicate impaired venous return due to increased intrathoracic pressure and link to delayed brain maturation.[10]

MRI has become an essential complementary imaging tool for the evaluation of fetal brain and body malformations, especially when US diagnostic capacity is limited.[11] In addition to US-based lung volume estimation via observed to expected lung-to-head ratio, MRI can accurately quantify fetal lung volume to provide prognostic information and guide perinatal management, for example, indicating the need for ventilation or ECMO.[2,3] Furthermore, it can depict the extent of organ dislocation into the thoracic cavity, with specific focus on the liver, where "liver-up" is prognostically unfavorable,[12] benefiting from its intrinsically high contrast between fluid-filled lungs and adjacent heart/liver parenchyma and the T1-weighted hyperintense display of the meconium-filled bowel.[13] With the recent development of Doppler-US gating based on the fetal cardiac cycle in the MRI scanner, fetal cardiac MRI has become feasible and has enabled the in-utero

FIGURE 3 Representative bSSFP Cine Images of the Fetal Heart Showing Manual Segmentation of the End-Diastolic and End-Systolic RV and LV Diameter at a Mid-Papillary Level



(A) Short axis at 29 GW: RV end-diastolic diameter = 9.8 mm, RV end-systolic diameter = 8.1 mm (indicated in blue), LV end-diastolic diameter = 8.5 mm, LV end-systolic diameter = 6.5 mm (indicated in yellow). (B) Four-chamber-view at 37 GW: RV end-diastolic diameter = 17.1 mm, RV end-systolic diameter = 13.1 mm (indicated in blue), LV end-diastolic diameter = 9.0 mm, LV end-systolic diameter = 6.5 mm (indicated in yellow). GW = gestational weeks. TE 1.66 ms, TR 33.93 ms, FoV read 360 mm, FoV phase 83%, flip angle 60°, slice thickness 4.0 mm, base resolution 224, phase resolution 104%, image acquisition under maternal breath-hold.

assessment of the fetal heart. Prior to that, the high fetal heart rate and a lack of fetal cardiac gating options constituted important limitations to fetal cardiac MRI.[14] Employing this technique could add valuable additional information to fetal MRI not only in congenital heart disease[15] but also in other syndromes involving the cardiovascular system and impacting cardiac function, for example, cardiovascular sequelae of lung malformations,[16] as in this case. Ventricular dysfunction is common in the early neonatal period of CDH cases and is a key contributor to disease severity and associated adverse outcomes, including the need for ECMO.[17] Two possible pathophysiological mechanisms may explain the emergence of pulmonary hypertension and ventricular strain observed in this case. First, increased intrathoracic pressure due to herniated abdominal organs may cause external compression of the lung parenchyma and thoracic vessels, increasing RV afterload, ultimately causing elevated RV pressure and dilatation. Second, while pulmonary resistance typically decreases as fetal vessels develop and expand in the third trimester,[18] CDH results in quantitatively and qualitatively impaired lung vascularization, potentially increasing pulmonary resistance and volume load of both ventricles.[9,18,19] Specifically, delayed growth of pulmonary vessels[18] and medial and adventitial thickening with premature differentiation of smooth muscle cells into a contractile phenotype[9,19] comprise pulmonary perfusion. Early in-utero assessment of cardiac function by fetal cardiac MRI may help to guide early individualized clinical decision-making, including intrauterine treatment strategies,[1] postnatal use of pulmonary vasodilators, cardiotropes, ECMO, and timing of surgical repair.[17] Fetal cardiac MRI is a new yet evolving method less affected by fetal position and maternal body habitus than US, offering diagnostic accuracy equivalent to echocardiography in congenital heart disease.[15] Despite good reproducibility,[20] fetal cardiac MRI is prone to motion artifacts, and interpretation may be limited by fetal motion and sparse MRI reference values.[6] To the best of our knowledge, we are the first to demonstrate this method's utility beyond congenital heart disease, such as in assessing cardiac sequelae of lung malformations, thereby potentially expanding the current practice of CDH workup.

CONCLUSIONS

In summary, fetal MRI in CDH can provide prognostic insights regarding exact lung volume estimation and extrapulmonary comorbidities. Adding Doppler-US gated sequences enables additional cardiac evaluation, supporting clinical decision-making and perinatal management in the expectation of not only respiratory but also cardiovascular challenges.

DATA SHARING STATEMENT

Data will be available upon reasonable request.

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KEY WORDS cardiac MRI, congenital diaphragmatic hernia, fetal MRI, pregnancy

APPENDIX For supplemental videos, please see the online version of this paper.