

MINI-FOCUS ISSUE: CONGENITAL HEART DISEASE

ADVANCED

IMAGING VIGNETTE: CLINICAL VIGNETTE

# Foramen Ovale Restriction in a Late Gestation Fetus Resembling Hypoplastic Left Heart Syndrome



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## ABSTRACT

A pregnant woman presented at term with a fetus with foramen ovale restriction. We illustrate the contrast between fetal and postnatal echocardiography of this rare entity. (**Level of Difficulty: Advanced.**) (J Am Coll Cardiol Case Rep 2021;3:721-3) © 2021 The Authors. Published by Elsevier on behalf of the American College of Cardiology Foundation. This is an open access article under the CC BY-NC-ND license (<http://creativecommons.org/licenses/by-nc-nd/4.0/>).

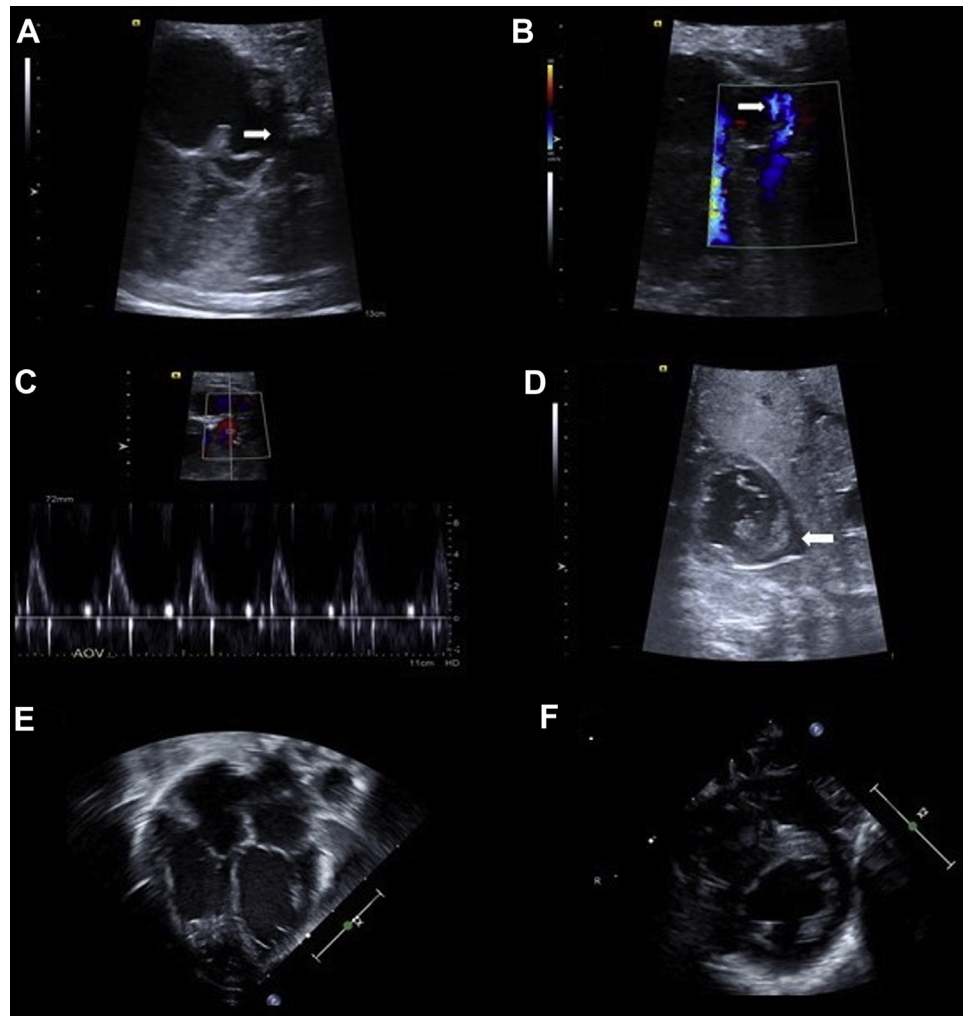
A 19-year-old woman was referred from a tertiary care center at  $37 \pm 6$  weeks due to suspected fetal hypoplastic left heart syndrome. A fetal echocardiogram showed restriction of the foramen ovale with a bulging atrial septal aneurysm nearly touching the left atrial wall (**Figures 1A and 1B, Video 1**). The right atrium and ventricle were moderately dilated (**Video 1**). There was normal right ventricular systolic function and trivial tricuspid regurgitation. The left ventricle appeared underfilled, with little antegrade mitral and aortic valve flow (**Figure 1C**). There was reversal of flow in the transverse arch. The ductus venosus and hepatic venous Doppler patterns were normal. A male infant was delivered via vaginal delivery. Prostaglandin was initiated at birth. The postnatal echocardiogram showed a tiny patent foramen ovale, near the apex-forming left ventricle with mild right atrial and right ventricular dilation (**Figures 1E and 1F**), low normal arch dimensions, and a large, predominantly right to left patent ductus arteriosus. Prostaglandin was discontinued. Oxygen via nasal cannula was continued until day of life 3, and he was discharged home.

Fetal restriction of the foramen ovale in a normal heart is a rare entity with an incidence of 0.2% to 1.4% (1). The pathogenesis is unclear, but it may occur at any gestation (1). Premature foramen ovale closure has been associated with left heart hypoplasia, right heart failure, hydrops, and death (2). Associated atrial septal aneurysm can also obstruct mitral inflow, contributing to an appearance of left ventricular hypoplasia (3). Postnatally, infants may develop pulmonary hypertension (1). This case illustrates that the disease may grossly mimic hypoplastic left heart syndrome in utero and the contrast between fetal and postnatal life. The preserved fetal right ventricular systolic function is unusual and suggests that the foramen ovale restriction happened late in gestation.

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The authors attest they are in compliance with human studies committees and animal welfare regulations of the authors' institutions and Food and Drug Administration guidelines, including patient consent where appropriate. For more information, visit the [Author Center](#).

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**FIGURE 1** Foramen Ovale Restriction With Atrial Septal Aneurysm Pre- and Post-Natally

(A) Fetal echocardiogram showing the dilated right atrium and right ventricle and the underfilled left heart. The **arrow** points to the atrial septum, which is nearly touching the left atrial wall. (B) Color flow over the atrial septum and mitral inflow. The **arrow** points to flow across the foramen ovale. (C) Doppler of the aortic valve showing low flow in utero. (D) Off-axis sagittal view showing the underfilled left ventricle (**arrow**) and dilated right ventricle. (E) Postnatal transthoracic echocardiogram showing the near apex-forming left ventricle and mild right atrial and right ventricular dilation. (F) Parasternal short-axis view demonstrating the left ventricle postnatally, which had a normal left ventricular end-diastolic dimension (1.9 cm, z-score:  $-0.4$ ).

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## REFERENCES

1. Gu X, Zhang Y, Han J, Liu X, Ge S, He Y. Isolated premature restriction or closure of foramen ovale in fetuses: echocardiographic characteristics and outcome. *Echocardiography* 2018; 35:1189-95.
2. Li YD, Li ZA, He YH. Premature closure or restriction of the foramen ovale: prenatal diagnosis by directional enhanced flow imaging. *J Ultrasound Med* 2013;32:1291-4.
3. Channing A, Szwast A, Natarajan S, Degenhardt K, Tian Z, Rychik J. Maternal hyperoxygenation improves left heart filling in fetuses with atrial septal aneurysm causing impediment to left ventricular inflow. *Ultrasound Obstet Gynecol* 2015;45:664-9.

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**KEY WORDS** echocardiography, fetus, foramen ovale restriction

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 **APPENDIX** For a supplemental video, please see the online version of this paper.