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IMAGING VIGNETTE

ADVANCED

CLINICAL VIGNETTE: EDITOR'S HIGHLIGHTS

A Heart Elsewhere

The Unusual Diagnosis of a Congenital Ventricular Diverticulum in the Abdomen

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ABSTRACT

Congenital left ventricular (LV) diverticulum is a rare condition characterized by the presence of a contractile appendix originating usually from the cardiac apex, but with high variability in location, dimension, and clinical presentation. We describe the diagnostic process and clinical management of an isolated apical diverticulum discovered during fetal life. (**Level of Difficulty: Advanced.**) (J Am Coll Cardiol Case Rep 2021;3:1453–1455) © 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 fetal echocardiogram in a 27-year-old woman identified an anomaly of the left ventricular (LV) apex with a contractile pouch aspect, measuring 11 × 6 mm (**Figure 1A**), in the context of mesocardia and persistence of left superior vena cava. The prenatal genetic evaluation and parents' echocardiographic assessment were negative. The pregnancy course was unremarkable.

After birth, a contractile subcutaneous mass was identified in the neonate's epigastric region, extending up to the umbilicus (Video 1). Transthoracic echocardiogram showed a contractile appendix originating from the LV apex and extending toward the abdomen, anterior to the liver (**Figure 1B,** Video 2), suspicious for LV congenital diverticulum herniating through an anterior diaphragmatic hernia.

Thoraco-abdominal computed tomography (CT) was performed, confirming the presence of a huge diverticulum measuring approximately 40 × 6 × 3 mm (**Figure 1C**), without evidence of thrombus inside. The lower part of the sternum and the xyphoid process were absent, suggesting an incomplete Cantrell's syndrome.

Considering the risks of thrombus formation/rupture and diverticulum's dimension, the patient was put on aspirin therapy and then underwent elective cardiac surgery 1 month after birth. The diverticulum was removed and the apex was closed with a patch of heterologous pericardium (**Figure 1D,** Video 1). The histological assessment showed normal myocardial layers. The neonate had an uncomplicated post-operative course.

Congenital LV diverticulum is a rare defect, accounting for 0.05% to 0,7% of all cardiac malformations, according to the different imaging techniques. It is either isolated from or associated with midline thoraco-abdominal abnormalities as part of Cantrell's syndrome, a rare midline disease with several variants, due to the developmental failure of mesoderm at early embryonic life (1).

The clinical presentation is highly variable, ranging from asymptomatic adult patients to cases of prenatal rupture, and syncope and arrhythmias are the most common symptoms. The prognosis is related to the underlying cardiac and systemic defects (2).

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ABBREVIATIONS AND ACRONYMS

CT = computed tomography

LV = left ventricular

The treatment should take into consideration the timing of diagnosis, symptoms, underlying comorbidities and size of the diverticulum. Both anticoagulant and aspirin have been proposed to reduce the risk of thrombosis, but there is no consensus regarding the best option.

Surgery is usually the treatment of choice in neonates/children with associated cardiac defects or big diverticula, due to higher risk of complications such as rupture, thrombosis, or arrhythmias (3). In adults, the best strategy is still unknown.

This case highlights the importance of a multimodality imaging approach in the diagnostic process of such rare and complex cases.

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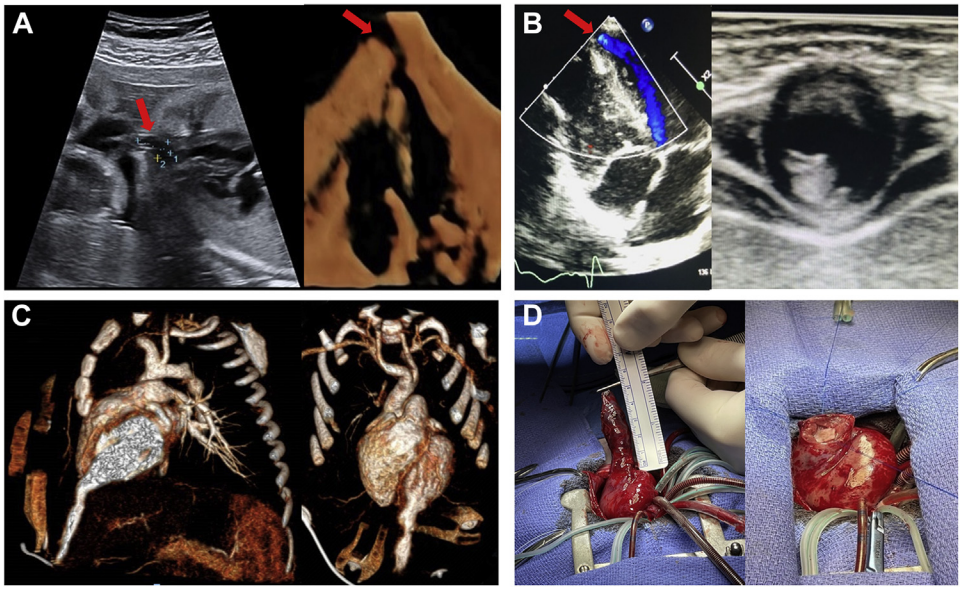
FIGURE 1 Left Ventricular Diverticulum



(A) Left ventricular diverticulum **(red arrow)** seen during fetal scan on 2-dimensional (2D) **(left)** and 3D **(right)**. **(B)** Transthoracic echocardiography: short axis from the abdomen **(left)** and apical 4-chamber view with color Doppler. **(C)** Computed tomography scan: 3D lateral **(left)** and frontal **(right)** reconstruction of the heart, showing the position and extension of the diverticulum. **(D)** Surgical view before **(left)** and after **(right)** the diverticulum was removed.

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