Cardiology in the Young

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Congenital giant left ventricular aneurysm

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Brief Report

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Abstract

We report a case of congenital giant left ventricular aneurysm with severely depressed systolic cardiac function who underwent early surgical resection with subsequent recovery of left ventricular systolic function.

Case presentation

A gravida 3, para 2 mother at 32 weeks' gestation was referred for fetal cardiac evaluation. Family history was significant for consanguinity, CHD and a previous neonatal death due to anencephaly. Fetal echocardiography demonstrated a large poorly contractile left ventricular outpouching and moderate pericardial effusion. The case was planned for delivery at our centre, and several surgical options were entertained including a single-ventricle route based on postnatal findings.

The patient was born at term via normal vaginal delivery with birth weight of 3.3 kg and Apgar scores of 4 and 7 at 1 and 5 minutes, respectively. She was transferred to neonatal ICU. Clinically, the patient had mild tachypnea with respiratory rate 65/min, oxygen saturation 95 %, mean arterial blood pressure 40 mmHg with acceptable perfusion and arterial blood gases, and no murmurs or dysmorphic features. She was connected to nasal continuous positive airway pressure, positive end-expiratory pressure 6 mmHg. Chest X-ray showed significant cardiomegaly. Abdominal and cranial ultrasound were normal. Full laboratory work-up was unremarkable.

Electrocardiogram revealed sinus rhythm with heart rate 160/min, left axis deviation, left ventricular hypertrophy, and T wave inversion in the inferior and lateral leads. Transthoracic echocardiography confirmed the diagnosis of a large left ventricular outpouching arising from the apical lateral left ventricular wall (Fig 1), with a wide neck and depressed left ventricular systolic function. There was conventional coronary artery origins. Cardiac MRI, performed under general anesthesia at 18 hours of life, demonstrated a large left ventricular outpouching (36 × 27 mm) from the apical and mid-lateral free wall with a wide connection to the native left ventricle (13 × 12 mm) and a volume comparable to the native left ventricle (Fig. 2). The medial walls of the outpouching had myocardium which exhibited focal fibrosis, while its lateral wall was very thin with akinesia and dyskinesia. Findings were consistent with a giant left ventricular aneurysm. The left ventricle was dilated with severely depressed global systolic function (EF 25%).

The patient developed mild respiratory distress and hypotension and was started on dobutamine infusion. On the 5th day of life, the patient underwent surgical aneurysm resection and left ventricular edges were approximated via a linear suture line followed by a double layer of continuous sutures without a patch (supplemental file 1). The patient came off cardiopulmonary bypass easily, and chest was closed and transferred to ICU in stable condition. Post-operative course was uneventful. Left ventricular systolic function gradually improved, and ejection fraction became 50% by day 5 post-op. EKG continued to demonstrate T wave inversions in the lateral leads, and no ventricular ectopy were noted in the post-operative period or on Holter monitor a couple of months later. Histopathological examination of the resected aneurysm revealed degenerative changes of the cardiac muscle with dense surrounding fibrosis (Supplemental file 2).

Discussion

Congenital left ventricular aneurysm and left ventricular diverticulum are rare cardiac congenital malformations, first described in 1816 in Germany.[1] They appear to be developmental anomalies that arise during the 4th embryonic week of gestation.[2] Most cases of left ventricular aneurysm/left ventricular diverticulum are asymptomatic but carry a risk of systemic embolisation, congestive heart failure, ventricular wall rupture, ventricular tachycardia or sudden cardiac death.[3] Left ventricular aneurysm is a ventricular akinetic or dyskinetic protuberance of fibrous tissue with no organised myocardium. On the other hand, congenital left ventricular diverticulum is an outpouching from a ventricle which contracts synchronously with that

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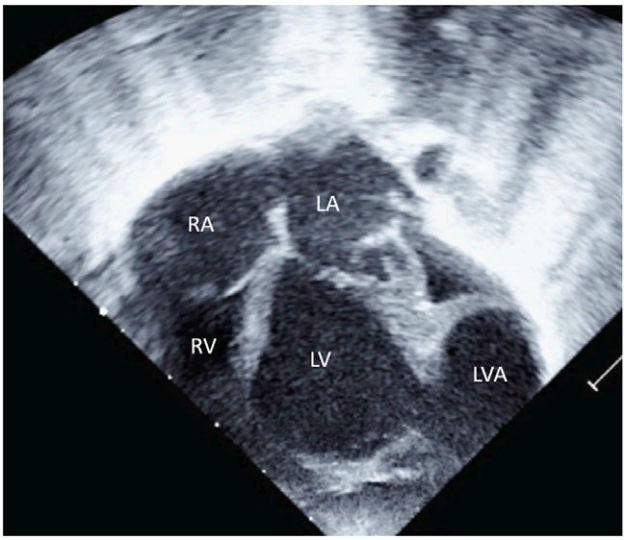


Figure 1. Transthoracic echocardiogram: apical four-chamber view demonstrating a large aneurysm arising from the left ventricle lateral wall.

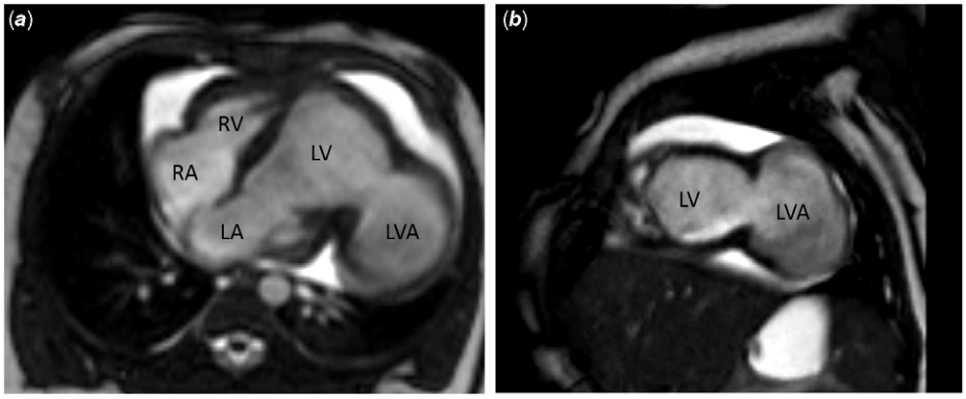


Figure 2. Cardiac MRI: four-chamber (***a***) and short-axis (***b***) projection balanced steady-state-free precession images demonstrating a large LV aneurysm arising from the lateral LV wall with a wide neck and small to moderate pericardial effusion. LA = left atrium; LV = left ventricle, LVA = left ventricular aneurysm; RA = right atrium; RV = right ventricle.

chamber and histologically contains all three layers of the ventricular wall (endocardium, myocardium, and pericardium).[4]

A retrospective study of 12,924 consecutive autopsies in children found 750 (5.8%) cases with congenital cardiac defects and 3 cases (0.02 %) with left ventricular diverticulum.[5] Congenital left ventricular aneurysm and left ventricular diverticulum are associated with other congenital anomalies including cardiac, vascular, or extracardiac anomalies. The prevalence of associated anomalies is markedly higher in patients with left ventricular diverticulum (34%) compared to left ventricular aneurysm (11%) suggesting a different aetiology and pathogenesis during embryonic development.[6]

A universal classification of left ventricular outpouching anomalies is not yet established. In 1961, Loogen et al. proposed a two-group classification where one group included pulsating palpable masses below the diaphragm and the other included masses above the diaphragm.[7] A more recent approach classified cases of congenital left ventricular outpouchings according to left ventricular geometry, wall thickness, and motion.[8] Accordingly, our case would be classified as type IIc, which carries a poor prognosis if left untreated.

In the perinatal period, left ventricular diverticulum and left ventricular aneurysm are mostly discovered on echocardiography. Asymptomatic smaller lesions are diagnosed incidentally later in life.[6] Our case was diagnosed by routine fetal echocardiography. Shuplock et al. described 86 cases of prenatally diagnosed congenital left ventricular outpouchings.[9] Pericardial effusions (44%) and ventricular dysfunction (17%) were the most common cardiac findings. Mortality rate was 17%, with the majority occurring prenatally. Factors associated with mortality included an outpouching located on the left ventricle, hydrops fetalis, pericardial effusion, and an earlier gestational age at diagnosis. Of those who survived to delivery, 57% remained asymptomatic without intervention, 15% regressed, and an additional 15% resolved.[9] Our patient did not have any arrhythmias but developed respiratory distress and low systolic blood pressure that required respiratory and inotropic support that could be related to the severely depressed left ventricular systolic function with giant dyskinetic aneurysm.

Treatment modalities of left ventricular aneurysm and left ventricular diverticulum are determined by the clinical presentation and associated findings of the patient. Conservative management may be sufficient in asymptomatic cases. Surgical resection is needed in symptomatic cases with associated cardiac anomalies. The first successful resection of an apical left ventricular diverticulum in a newborn was documented in 1944.[10] Surgical technique depends on the type and extension of the outpouching and usually requires cardiopulmonary bypass, although up to 30% can be resected without extracorporal circulation.[6]

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In our case, we proceeded with early surgical resection taking into consideration the high risk of rupture, thromboembolic complications, and congestive heart failure.

In conclusion, congenital left ventricular aneurysm and left ventricular diverticulum are rare congenital cardiac defects that can be diagnosed prenatally. Early surgical resection can be associated with favourable outcomes in large symptomatic cases.

Supplementary material. To view supplementary material for this article, please visit https://doi.org/10.1017/S1047951122003651.

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Conflicts of interest. None.

Ethical standards. This work has been approved by the institutional review board at King Faisal Specialist Hospital & Research Center – Jeddah IRB: 2022-CR-16.

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