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CASE REPORTS

Isolated Congenital Left Ventricular Diverticulum of the Fetal Heart in a Twin Pregnancy: Case Report and Literature Review

Maha Binfadel[1] · Abdah Hrfi[1,2] · Abdullah Al Sehly[1,3,4]

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Abstract We report a rare congenital heart disease in the form of isolated fetal left ventricular diverticulum in a spontaneous twin pregnancy, detected at 30 weeks of gestation. The diagnosis was confirmed postnatally with Echocardiography and Cardiac MRI. The case highlights the relevance of antenatal screening in detecting cardiac anomalies as early as possible to allow early postnatal work up and intervention if needed. The patient is currently 6 years old, and is followed regularly and is completely asymptomatic with no intervention required

Keywords Left ventricular diverticulum · LVD · LV outpouching · Antenatal diagnosis

Case Report

Our patient currently is a 6 years old boy who was part of a spontaneous twin pregnancy. He was diagnosed in utero with a congenital LV diverticulum. He is twin A for a 38-year old mother, gravida 3 para 2 (G3P2), living 0. The previous two pregnancies ended in preterm delivery due to preeclampsia at 29 weeks and 26 weeks respectively with early neonatal death due to prematurity complications. The mother is a known case of hypothyroidism on L-thyroxin. She was referred to our institute with spontaneous twin pregnancy, diamniotic, dichorionic for fetal cardiac evaluation at 30 weeks of gestational age with a suspicion of cardiac anomalies. With moderate to large LV outpouching of twin A. There was no family history of congenital cardiac disease. Fetal Echocardiogram in our hospital for our patient (Twin A) demonstrated atrio-ventricular and ventriculo-arterial concordance. The left and right ventricular outflow tracts and a dilated main pulmonary artery. Systemic and pulmonary venous connections were normal. Normal aortic arch and ductal arch were seen. There was a moderate size cavity connected to the left ventricle posteriorly with blood flow seen across: posterior wall LV diverticulum. There was a moderate circumferential pericardial effusion (Fig. 1). No clot was visualized inside the cavity. There was an echogenic LV myocardium and competent mitral and tricuspid valves. No ventricular septal defect was seen. Echocardiogram of twin B was normal. Pregnancy was smooth and the patient was delivered at 36 weeks gestation by elective cesarean section due to previous two cesarean sections. The baby was born with a birth weight of 2 kg and good APGAR score. There were no dysmorphic features. The baby maintaing his oxygen saturation with room air without any respiratory support. Systemic examinations were unremarkable. He was admitted to the neonatal care unit for evaluation. A chest X-Ray showed mild cardiomegaly with clear lung fields. Twelve lead ECG demonstrated sinus rhythm with regular heart rate no atrial or ventricular arrhythmia and no ischemia. Postnatal echocardiogram confirmed the presence of a large posterolateral LV diverticulum and mildly dilated right sided chambers. There was a moderate size

✉ Abdullah Al Sehly
aalsehly@kfshrc.edu.sa

[1] Department of Pediatric Cardiology, King Faisal Specialist Hospital and Research Center, Riyadh, Saudi Arabia

[2] Department of Pediatric Cardiology, Prince Sultan Cardiac Center, Qassim, Saudi Arabia

[3] Division of Pediatric Cardiology, King Faisal Heart Institute, Riyadh, Saudi Arabia

[4] Non-Invasive Cardiac Imaging Unit, King Faisal Specialist Hospital and Research Centre, MBC-61, PO Box 3354, Riyadh 11211, Saudi Arabia

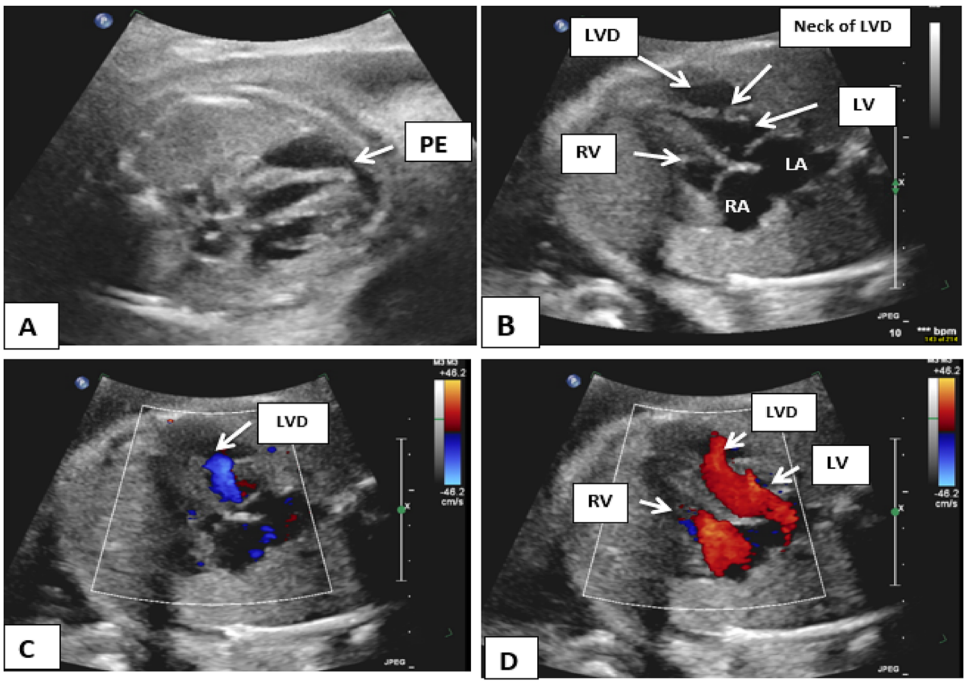

Fig. 1 2D and color Doppler echocardiographic images of four chamber views of twin A fetal heart at 30 weeks gestation demonstrating **A** moderate pericardial effusion (PE). **B** Large left ventricular diverticulum from the postero-lateral LV wall (LVD) with the neck of this diverticulum. **C**, **D** showing blood flow from left ventricle (LV) into the diverticulum through a narrow neck (red color) and from LVD to LV (blue color). *LV* left ventricle, *RV* right ventricle, *RA* right atrium, *LA* left atrium, *LVD* left ventricular diverticulum, *PE* pericardial effusion

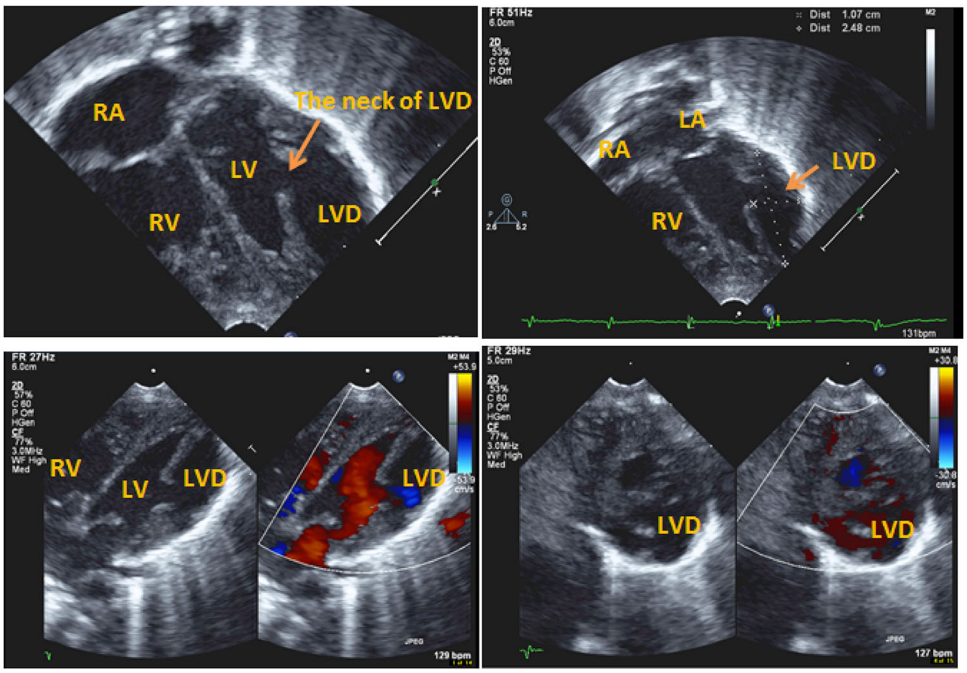

Fig. 2 Postnatal transthoracic 2D and color Doppler echocardiographic imaging of the four-chamber and short axis views demonstrating large posterolateral left ventricular diverticulum (LVD) and blood flow from left ventricle (LV) into and out the diverticulum. *RA* right atrium, *RV* right ventricle, *LA* left atrium, *LV* left ventricle, *LVD* left ventricular diverticulum

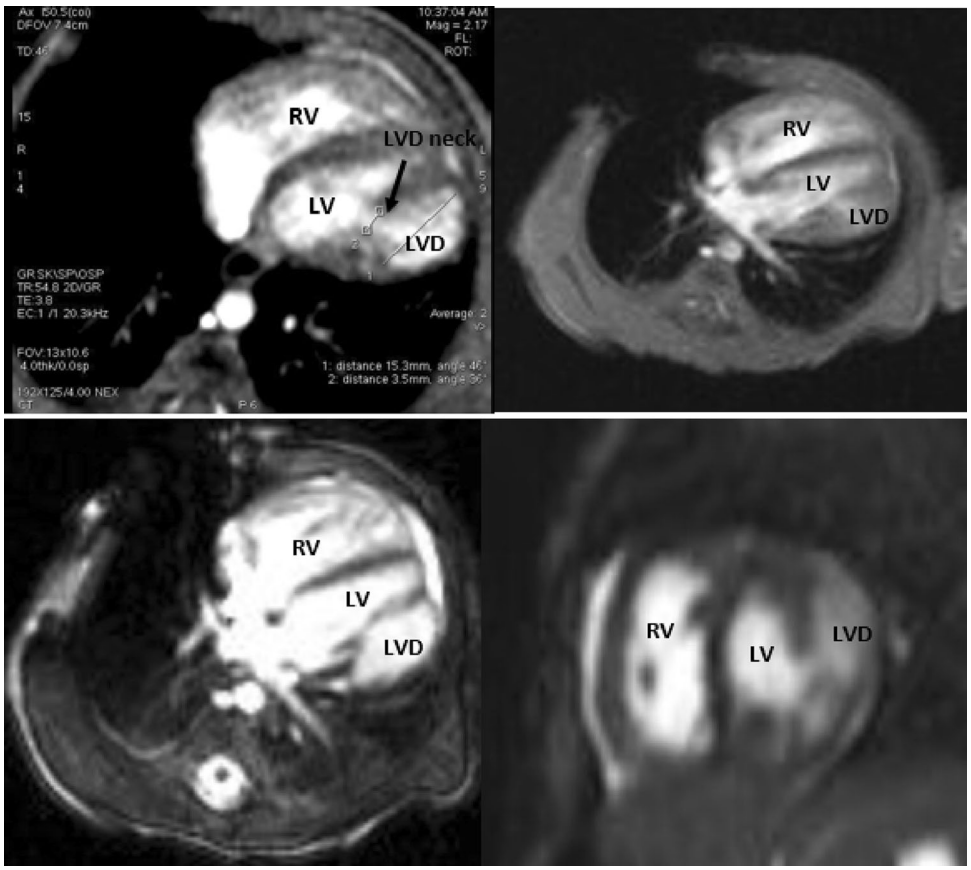

Fig. 3 Post-natal cardiac MRI and viability study showed: Large left ventricular diverticulum arises approximately from the mid lateral wall and measures around 5 mm in length, maximum length approximately 1.8 cm with no thrombus in diverticulum and no evidence of scar or fibrosis formation on viability study

atrial septal defect secundum type (which closed spontaneously at follow up echo studies). There was no pericardial effusion (Fig. 2). Cardiac MRI was done at the age of 6 days confirming the same finding of congenital diverticulum of left ventricular lateral wall arising approximately from the mid lateral wall measuring around 5 mm in length. Maximum diverticulum length was 1.8 cm. No thrombus was seen in the diverticulum cavity and no evidence of scar or fibrosis formation was demonstrated on viability study (Fig. 3). The condition of the baby was quite stable with no indications for surgical intervention. Currently, the child is 6 years old and is on regular follow up in our pediatric cardiology clinic. He remains asymptomatic, thriving well and developmentally normal. His last echocardiogram showed the congenital left ventricular diverticulum (LVD), located at the posterolateral aspect of left ventricle. The diverticulum is of the same size with no evidence of clots. There is a good sized opening seen between the LV cavity and diverticulum with bidirectional flow with normal biventricular systolic function. There is a normal AV valves inflow (Fig. 4). ECG showed sinus rhythm with no evidence of any arrhythmia or ischemic changes.

Discussion

Congenital LVD is a rare cardiac malformation. It is a localized outpouching from the cardiac left ventricular free wall [1]. It appears to be a developmental anomaly [4, 5]. The etiology of a ventricular diverticulum is uncertain and possibilities include developmental abnormalities in embryogenesis, viral infections, and myocardial ischemia [5, 6]. Histologically, this lesion contains all layers of the ventricular wall with preservation of the myocardial architecture [1–6]. Ventricular diverticulum can arise from the right or left ventricle or both together, however, the LV diverticulum is more frequent [7]. Apical LVD is the most common one, however, non-apical diverticula may occur [7]. The natural history of left ventricular diverticuli is uncertain as the prognosis is generally determined by associated abnormalities. Patients with isolated LVD

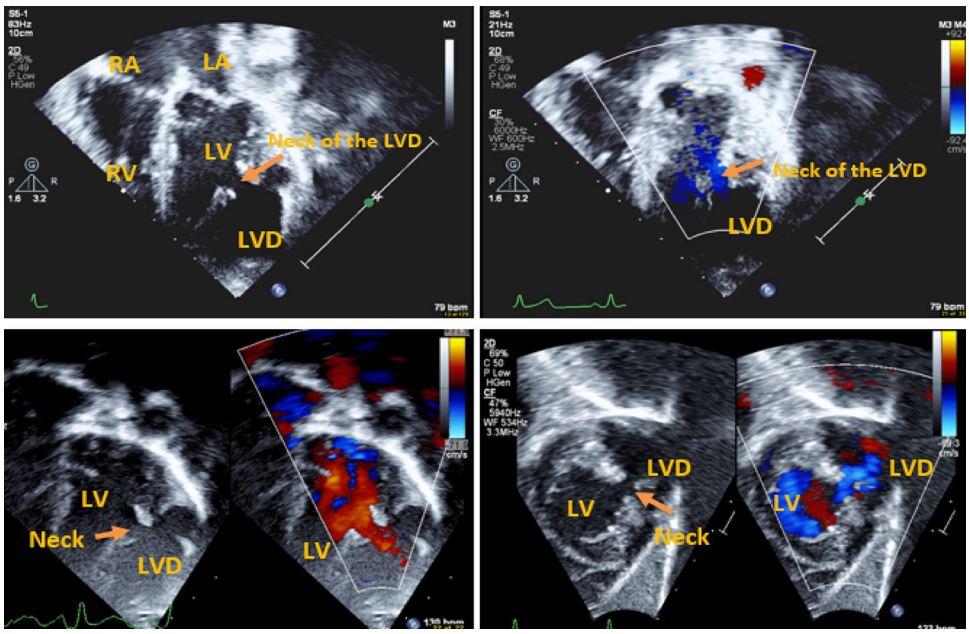

Fig. 4 Four chamber and short axis views of the last follow up transthoracic Echocardiogram showing large left ventricular diverticulum (LVD) with no thrombus, and blood going into and out of it

are often asymptomatic and diagnosed incidentally during diagnostic procedures or screening for other reasons [4, 5]. In general, there are multiple possible complications, like arrhythmia, infective endocarditis, embolism and thrombus formation. Rupture can rarely be the initial presentation [1–4]. Common in utero complications include cardiac dysrhythmias, pericardial effusion and heart failure [5]. The diagnosis is usually established postnatally by echocardiography, CT angiography and cardiac MRI [7]. Our review of the literature showed that the true incidence of congenital ventricular diverticula is difficult to ascertain, as these lesions are often grouped with congenital aneurysms when described in the literature [5] and only a few cases diagnosed during fetal life have been published [3]. However, diagnosis of congenital LVD can be established antenataly by fetal echocardiography and fetal cardiac MRI. Management of this rare congenital heart lesion depends on the clinical situation and associated abnormalities and complications during pregnancy. Most are asymptomatic and managed with a conservative approach and careful follow up during pregnancy. Treatment options for symptomatic high-risk cases may include planned delivery for postnatal management which might include surgical resection and/or anticoagulation, as well as heart failure and arrhythmias treatment if present after birth [6].

Conclusion

Isolated congenital LVD is a very rare form of congenital heart defect of the fetal heart and can be diagnosed prenatally. Fetal ultrasonography screening is an important and established technique for the prenatal diagnosis of congenital heart diseases in fetal life [2]. The four-chamber view of the heart is a standard view of the 18–20 week fetal anatomy scan, and 2D evaluation of the heart with color Doppler imaging has greatly enhanced the detection of rare and subtle cardiac anomalies in general, especially in non specialized centers for fetal echocardiogram which facilitate an early referral to a specialized center. In our case fetal echocardiogram allowed us to diagnose this case in utero with an early postnatal comprehensive evaluation plan and follow up.

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Declarations

Conflict of interest The authors declare no conflict of interest.

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