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CASE REPORT

Left Ventricular Accessory Chamber: A Case Report and Review of the Literature

Sawsan M. Awad · Anjali S. Patel · Anastasios Polimenakos · Ryan Braun · Ra-id Abdulla

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Abstract Congenital left ventricular out-pouching, consisting of left ventricular aneurysms, left ventricle diverticulae, and double-chambered left ventricle, is a rare cardiac malformation. Criteria to differentiate between left ventricular aneurysm and diverticulum are defined. Reports of left ventricular accessory chamber in the medical literature are rare. In this article, we present a case of left ventricular accessory chamber diagnosed during the late prenatal period. Review of the literature, imaging modalities used for differentiation of the nature of this lesion, and management plan are discussed.

Keywords Left ventricular aneurysm · Left ventricular diverticulum · Left ventricular accessory chamber

Left ventricular (LV) outpouching in the fetal/neonatal population is a rare condition. Differential diagnosis includes aneurysm, diverticulum (apical vs. nonapical), and double-chambered ventricle. Although distinguishment of these conditions is well defined in the literature, diagnosis based on echocardiography (fetal or postnatal) remains often challenging. LV aneurysm is defined as a saccular structure with a wide connection to the LV that does not contract synchronously with the LV but, rather, exhibits expansion with ventricular systole [6]. LV apical diverticulum contracts synchronously, as it normally contains all three layers of cardiac tissue and has a narrow connection to the ventricle [6]. Double-chambered left ventricle often exhibits mixed criteria for the previously described entities. The accessory chamber has a wide connection to the ventricular chamber as in an aneurysm but contains all three cardiac tissue layers as in a diverticulum. Early and accurate diagnosis is crucial to differentiate the above entities and thus to guide management [7].

Case Report

An African American boy was born at 39 weeks' gestation via vacuum-assisted vaginal delivery to a 17-year-old mother, G1P0, who had gonococcal infection in the second trimester that was treated successfully. Apgars were 8 and 9 at 1 and 5 min, respectively. Prenatal fetal echocardiogram at 37 weeks' gestation showed what was suspected to be an accessory LV chamber versus LV aneurysm, with normal ventricular function and no significant pericardial effusion. There was a wide communication between the accessory chamber and the LV, with bidirectional flow across the connection noted by color Doppler. Transthoracic echocardiogram (TTE) after delivery showed similar findings (Fig. 1) as identified previously during fetal life. Coronary artery origins and proximal courses appeared normal by both TTE and aortic root angiography.

Cardiac MRI without gadolinium (Fig. 2) showed thinning of the lateral wall of the LV, with focal outpouching of an abnormal myocardium at the LV free wall and near the apex. There was a global decrease in systolic

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S. M. Awad (✉) · A. S. Patel · A. Polimenakos · R. Braun · R. Abdulla
Rush Center for Congenital and Structural Heart Disease, Rush University Medical Center, 1653 West Congress Parkway, 770 Jones, Chicago, IL 60612, USA
e-mail: sawsan_m_awad@rush.edu

A. S. Patel
e-mail: anjali_patel@rush.edu

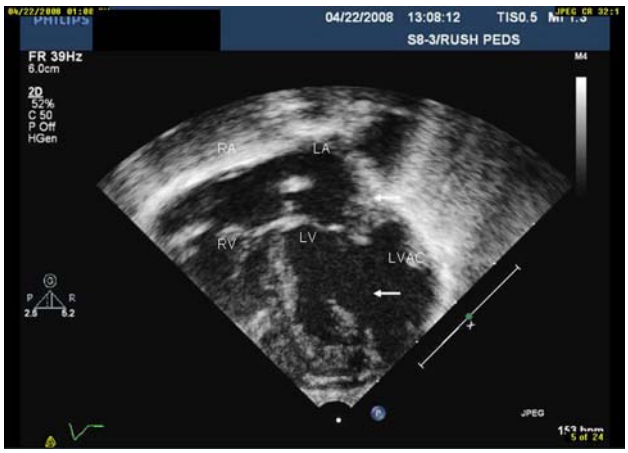

Fig. 1 Transthoracic echocardiography in the first few days of life. The apical four-chamber view shows the left ventricle (LV) and the LV accessory chamber (LVAC). White arrows point to the wide connection between the LV and the LVAC. *LA* left atrium; *RA* right atrium; *RV* right ventricle

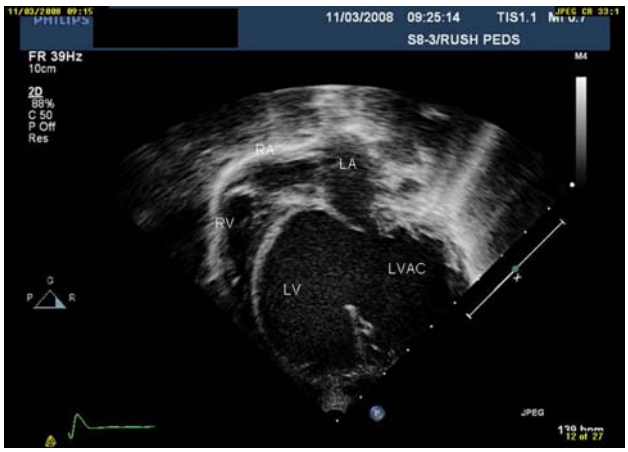

Fig. 3 Transthoracic echocardiography: apical four-chamber view at 6 months of age showing the dilated left ventricle (LV) with bowing ventricular septum to the right. *LA* left atrium; *LVAC* left ventricular accessory chamber; *RA* right atrium; *RV* right ventricle

contractility of the LV, with an LV ejection fraction of 15.8%. Antifailure medications (lasix, digoxin, and captopril) and salicylate were initiated due to poor LV function and to prevent thrombus formation.

Throughout the hospital stay, the infant remained stable, with no evidence of low cardiac output or arrhythmias. The patient was sent home, to be followed closely in an outpatient setting. During subsequent visits, progressively decreased LV systolic function, and dilation of LV were seen by echocardiography (Fig. 3). Poor weight gain was noted despite optimal caloric intake. Holter monitoring did not show significant arrhythmias. Repeat MRI (Fig. 4) with gadolinium at 6 months of age showed progressive dilatation of the LV, with progressively decreasing systolic function and an LV ejection fraction of 9%. It also identified the wall thickness of the accessory chamber to be of the same enhancement of the LV. Heart transplantation was recommended due to significant failure to thrive secondary to heart failure.

Discussion

Congenital LV outpouching is a rare anomaly. The exact nature of the outpouching is difficult to determine in most instances. In almost all cases, it is difficult to distinguish whether it is an aneurysm or an accessory chamber. Moreover, both terms are used interchangeably, with limited distinction between them.

Many reports describe the difference between ventricular aneurysm and diverticulum [1, 2, 6, 7, 10, 13]. Aneurysm has a wide communicating neck to the LV chamber. In addition, the wall of the outpouching is dyskinetic and there is a lack of full layers composing the ventricular wall. On the other hand, diverticulum has a narrow neck connection to the ventricle, similar layers as the ventricular wall, and absence of dyskinetic motion in systole and diastole. In our patient, the abnormal LV outpouching possessed mixed criteria, which made it difficult to categorize it into either group.

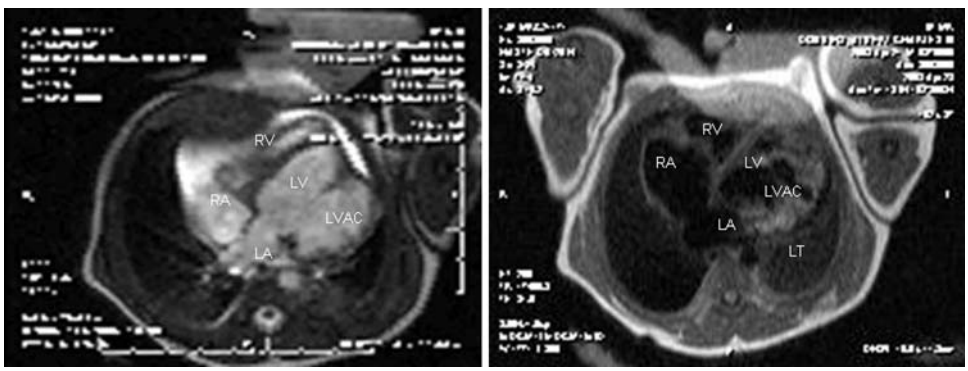

Fig. 2 Cardiac MRI in the first few days of life showing the left ventricle (LV) and the LV accessory chamber (LVAC). Note the cardiothoracic ratio compared to that on cardiac MRI 6 months later. *LT* lung tissue; *RA* right atrium; *RV* right ventricle

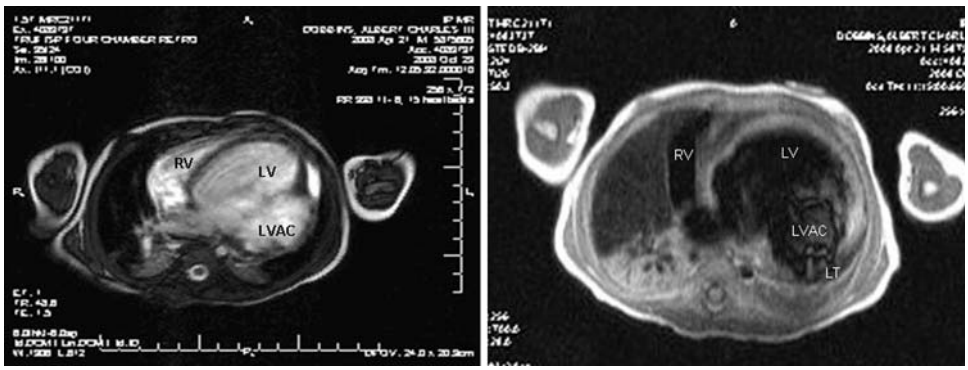

Fig. 4 Cardiac MRI at 6 months of age showing the dilated left ventricle (LV) compared to the initial cardiac MRI. Note the cardiothoracic ratio and the compressed right ventricle (RV). *LT* lung tissue; *LVAC* left ventricular accessory chamber

The etiology of this lesion is debated and not clearly understood. Several theories include ischemic insult [1], viral infection [2, 13], and, rarely, genetic abnormality. However, a more recent report of seven cases with congenital LV aneurysm showed no evidence of either ischemic insult or viral infection in all reported cases. Gross and histologic evaluation of coronary arteries in reported cases shows that normal coronary artery anatomy and polymerase chain reaction do not reveal evidence of viral myocarditis [10]. The etiology of the LV outpouching in our patient is not known, however, normal early obstetrical ultrasounds may point toward an ischemic insult in the late second or early third trimester.

In the case presented, it is clear that the lesion does not represent a diverticulum. It was difficult to differentiate whether this lesion is an aneurysm or an accessory chamber. Kay et al. reported a case in 1983 of a 4-month-old male with failure to thrive and a diagnosis of LV accessory chamber that was excised secondary to manifestation of heart failure. Histological analysis of the excised chamber showed muscular hypertrophy with irregularly arranged fibroelastic tissue [5].

Prenatal diagnosis of such an anomaly is increasing with the advances in prenatal care and fetal echocardiography [3, 11, 12]. Several cases were reported to have either LV aneurysm or diverticulum [1, 7, 8] with either intrauterine fetal demise or severe hydrops. The case in hand was diagnosed in the late prenatal period, with no evidence of heart failure or hydrops. Early prenatal ultrasound showed normal structures and function of the heart, which may point to a late prenatal ischemic insult as an etiology. The late onset of this event may be the reason for the absence of heart failure signs in utero.

MRI with gadolinium was helpful in verifying the nature of the accessory chamber. The presence of myocardium in the wall of the accessory chamber with the same degree of enhancement as the LV myocardium further favors the diagnosis of LV accessory chamber rather than aneurysm. However, no definite diagnosis may be obtained without pathological analysis.

The management plan for similar cases as reported in the literature consists of surgical resection of this chamber in asymptomatic patients and resection of the accessory chamber or cardiac transplant in symptomatic patients [4, 9]. In our patient, we elected medical management; however, this was abandoned in view of intractable heart failure and failure to thrive.

Surgical excision of the accessory chamber with interposition patch reconstruction and cardiac transplant were two valid options. The latter choice was favored due to the wide connection between the LV and the accessory chamber. This would have rendered the placed patch proportionally large compared to the LV free wall, with resultant poor LV function and failure to wean off bypass.

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