

Double-chambered Left Ventricle in Adult Patient with Asymptomatic Evolution

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Introduction

Double-chambered left ventricle is an expression used to refer to a rare congenital condition characterized by the subdivision of the ventricular cavity by a muscle bundle or by an anomalous septum.¹⁻³

Few cases are available in the literature with descriptions of varying presentations for this condition.² Diagnosis is usually established in the pediatric or neonatal age range and echocardiography and cardiac magnetic resonance imaging are the most used tests for detection and morphological and functional evaluation.³⁻⁵

Case Report

Female Caucasian 29-year-old patient was referred to a cardiologist to reevaluate the history of echocardiographic abnormalities from birth. The patient brought nine previous echocardiography scans with inconsistent conclusions. The first survey, conducted in 1988, reports a perimembranous ventricular septal defect; the other ones with descriptions ranging from normal test to the presence of left ventricular mass and findings suggesting uncompressed left ventricle.

The patient was asymptomatic on clinical evaluation, with no limitation for daily activities; on physical examination, holosystolic murmur in the lower left parasternal region with 2+/6+ intensity.

The following laboratory tests were performed: electrocardiogram with sinus rhythm with clockwise rotation and axis at about 100 degrees, with no other relevant findings. 24-hour Holter with sinus rhythm, absence of ventricular and supraventricular arrhythmias; no atrioventricular or intraventricular conduction disorders. Ventricular repolarization breaks or abnormalities were not evidenced.

Magnetic resonance imaging performed in 2010 identified muscle bundle in the left ventricle communicating the antero-septal wall to the anterolateral wall with a maximum thickness of 1.3 cm; prominent anterior papillary muscle,

associated with a potential non-compacted myocardium affecting the anterior and lateral walls in the middle and apical segments. No uptake areas were identified in the delayed enhancement phase.

Patient underwent another two-dimensional and three-dimensional echocardiography scan at our service, which identified an anomalous muscle bundle in the left ventricle (Figures 1, 3 and 4, Video 1), resulting in two large chambers aligned in parallel with the right ventricular inflow tract with non-obstructive filling (Video 2). Perforations were also observed in the muscle bundle with systolic flow from the anterolateral chamber to the medial chamber and minimal apical muscular ventricular septal defect (Figure 2), making up a double-chambered left ventricle. The left ventricular cavity dimensions were within normal limits.

Discussion

The first consistent reports of double-chambered left ventricle date from the late 1970s.^{6,7} The literature is scarce in terms of information about such anomaly and little is known about its prognostic evaluation and potential risks and complications. It may be the most fearsome thrombogenic capacity of the accessory chamber.^{2,3}

There is no evidence of electrocardiographic or chest X-ray findings with good accuracy for this diagnosis. The confirmed presence of double-chambered left ventricle in the previously described cases took place primarily through echocardiography and cardiac magnetic resonance imaging,¹⁻³ with a few reports mentioning computed tomography as an alternative.⁴

Despite the diversity of clinical presentations and morphological variations described, it is believed that the evolution of patients with double-chambered left ventricle is, as a rule, benign, whereas the chambers are often separated in parallel without determining pressure gradients or left ventricular outflow tract obstruction.³⁻⁵ Therefore, the treatment ends up being guided by the patient's functional impairment.²

Among the differential diagnoses, we must remember the left ventricular aneurysms characterized by structures that protrude from the ventricular cavity from a sort of neck, presenting an expanding trend rather than contraction during systole. In the double-chambered left ventricle, the muscle wall presents contraction and volume reduction simultaneous to the systole with a separation between the cavities by a sort of membrane or accessory muscle bundle.^{4,5}

Also as a differential diagnosis, the distinction for the double-chambered right ventricle (DCRV) should not be

Keywords

Heart Defects, Congenital; Ventricular Dysfunction, Left; Heart Septal Defects, Ventricular.

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forgotten, which is a condition with a completely different pathophysiology: the DCRV is a more common condition, usually presenting a clinical history of dyspnea on exertion and murmur on auscultation. There are reports of an association between DCRV and congenital heart defects, such as tetralogy of Fallot and transposition of the great vessels, an association not described in cases involving double-chambered left ventricle.^{5,8}

The genesis of DCRV is also distinguished, as there is progressive thickening of the right ventricular septum by the presence of anomalous muscle bundles. This phenomenon leads to higher gradients and the formation of double chambers in series,⁸ in contrast with the most frequently reported presentations of double-chambered left ventricle, in which the cavities are in parallel with lower pressure gradients and synchronous contraction.^{4,5}

Because of the rarity of the disease, lack of familiarity with this condition led to several misdiagnoses in echocardiograms

and even the magnetic resonance imaging findings did not lead to proper conclusion. The previous assumptions could lead to tests and interventions with potential unnecessary risks to the patient.

The innocuous behavior of this condition — found in most literary evidence — combined with the absence of signs and symptoms suggestive of left ventricular dysfunction led us to choose expectant management in this case, without the need for pharmacological therapies or further investigation.

Authors' Contributions

Data acquisition: Pretto J LCS, Oliveira RM; Data analysis and interpretation: Pretto J LCS, Oliveira RM; Manuscript drafting: Pretto J LCS, Oliveira RM; Critical revision of the manuscript as for important intellectual content: Pretto J LCS, Oliveira RM, Franciscatto T, Balestreri F, Roman RM.

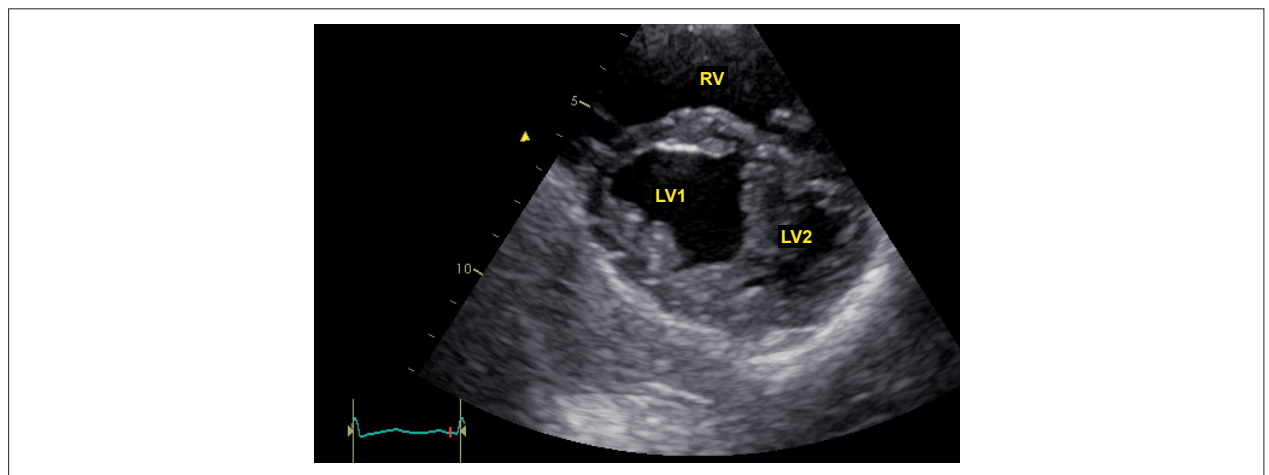


Figure 1 – Cross-section of the left (LV) and right ventricle (RV).

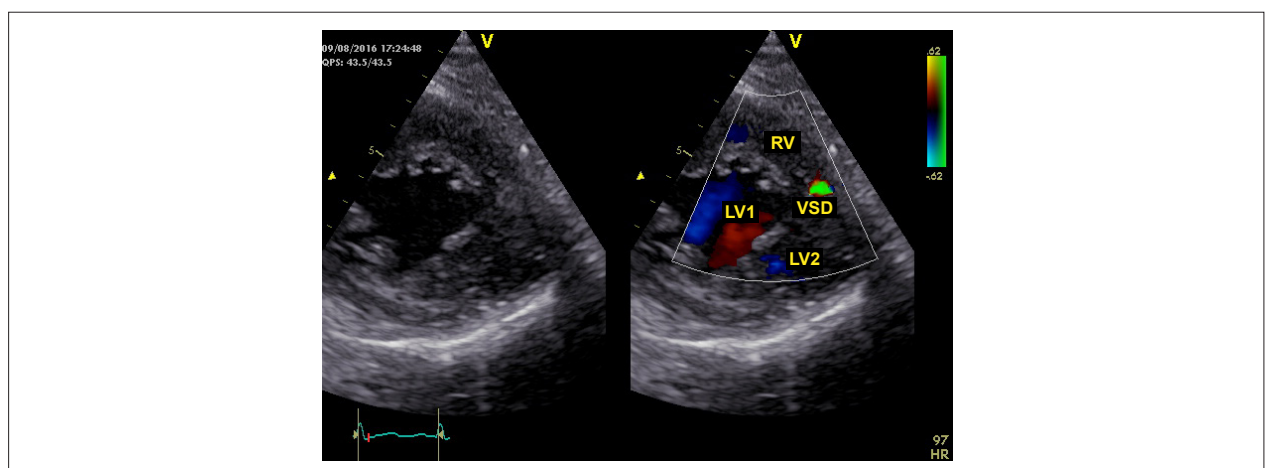


Figure 2 – Cross-section of the left and right ventricle showing a small ventricular septal defect (VSD).

Case Report

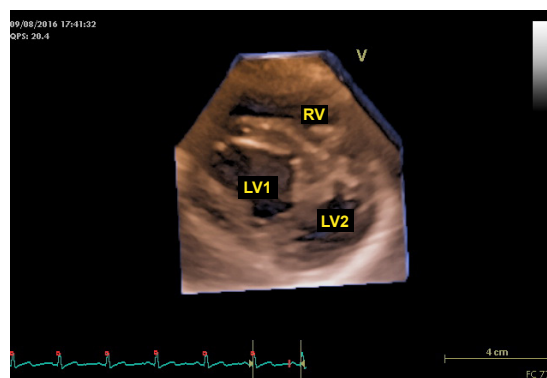


Figure 3 – Three-dimensional cross-section.

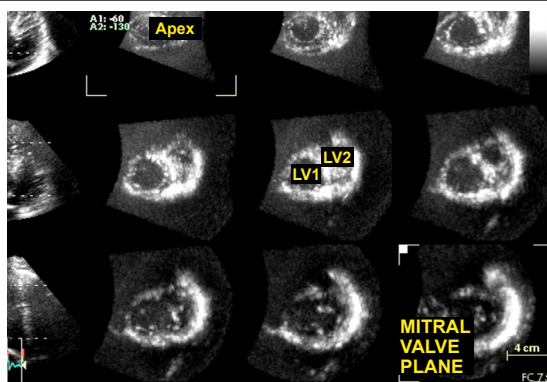


Figure 4 – Three-dimensional cross Multislice.

Video 1 – Click here. http://departamentos.cardiol.br/dic/publicacoes/revistadic/2016/v29_4/video_v29_4_166_ingles.asp

Video 2 – Click here. http://departamentos.cardiol.br/dic/publicacoes/revistadic/2016/v29_4/video_v29_4_166_ingles.asp

Potential Conflicts of Interest

There are no relevant conflicts of interest.

Academic Association

This study is not associated with any graduate program.

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