

# Double-chambered left ventricle in a young previously healthy man presenting for a routine echocardiographic study (RCD code: IV-1B.2o)

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## Abstract

A double-chambered left ventricle (DCLV) constitutes a rare congenital malformation. It is usually diagnosed at a neonatal or pediatric age, and often exhibits mixed criteria for diverticulae and aneurysms. This anomaly is characterized by the subdivision of the left ventricle into two chambers by an abnormal septum or muscle band. In this article, we present the case of 30-year-old asymptomatic patient, in whom DCLV was diagnosed by routine echocardiographic study. JRC D 2013; 1 (4): 155–157

**Key words:** a double-chambered left ventricle, diverticulum, aneurysm

## Background

A double-chambered left ventricle constitutes a rare congenital malformation. It is usually diagnosed at a neonatal or pediatric age, and often shows mixed criteria for diverticula and aneurysms. This anomaly is characterized by the subdivision of the left ventricle (LV) into 2 chambers by an abnormal septum or muscle band.

## Case presentation

We present a case involving a 30-year-old patient who was admitted to our outpatient clinic for a routine echocardiographic study that was required for his employment health check. He was asymptomatic and had no history of cardiac diseases or any other chronic diseases. He reported no symptoms of heart failure, dyspnea, or chest pain. A physical examination did not reveal any abnormalities. Sinus rhythm was observed on an electrocardiogram, and no rhythm disturbances were seen.

Transthoracic echocardiography revealed normal LV function (Figure 1). No valvular dysfunction or hypertrophy was observed. However, an additional cavity lying within the LV was visualized in two-chamber (Figure 2) and modified two-chamber (Figure 3) apical long-axis views. The additional cavity of the LV was located

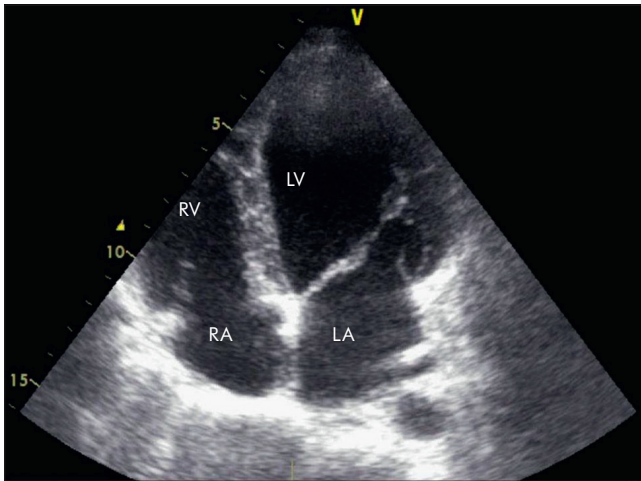
in the posteroinferior LV wall. It was not clearly visible in the traditional parasternal long-axis 2D view, and no pathology could be definitely determined on the images in the apical four-chamber view (Figure 1). The global LV systolic function was normal. The muscle of the additional LV cavity thickened during systole; however, the myocardium was nonhomogenous with small lacunae. The ratio of spongy / compacted layers was about 1. No thrombus was observed. There was a septum dividing both cavities, which was rich in fenestrations and lacunae that enabled free flow between the two of them (Figure 3). Turbulent systolic inflow and diastolic outflow with high velocities were present in the pulse and color Doppler imaging. Cardiac magnetic resonance imaging showed the dimensions of the additional LV cavity to be 77 × 60 × 39 mm; moreover, it revealed reduced ejection fraction (20%). Narrow neck of the cavity was observed (17 × 20 mm). The cavity consisted of fibrous membrane (thickness of 3–4 mm) with some fenestrations (max. 4 mm). At the same time, the main LV cavity had preserved ejection fraction of 58% as well as end-diastolic volume of 168 mL and end-systolic volume of 71 mL (Figure 4, 5). The final diagnosis was a double-chambered LV.

The patient was evaluated by a cardiac team and the decision was made to administer conservative treatment. He was advised to take anticoagulants to prevent thrombotic complications of the pathology (which are possible but not frequent). He was also advised to undergo regular echocardiographic check-ups.

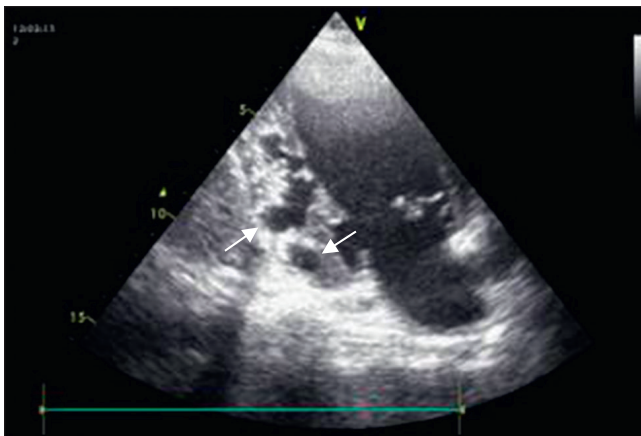
Conflict of interest: none declared.

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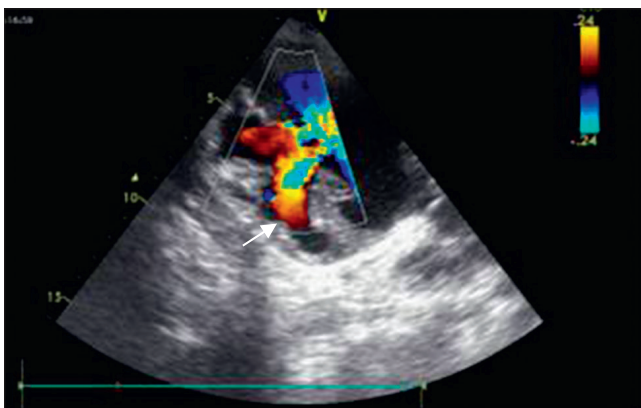
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**Figure 1.** Echocardiography. Apical four-chambered view. LV – left ventricle, RV – right ventricle, LA – left atrium, RA – right atrium



**Figure 2.** Echocardiography. Apical two-chamber view. Additional cavity within the left ventricle – subdivision of the left ventricular cavity by an abnormal septum into 2 chambers; fenestrations and lacunae within the pathology (arrows)



**Figure 3.** Echocardiography. Apical two-chamber view, modified. Color Doppler imaging. Turbulent flow between the two cavities is observed (arrow)

## Discussion

In the literature, only a few case reports of a double-chambered LV have been described to date. When the PubMed database was searched for “Double-chambered left ventricle” (DCLV), only 19 citations were found. The majority of the papers focused on single cases. Thus, this pathology may be rightly named an “orphan disease”, according to the classifications of the Centre for Rare Cardiovascular Diseases.

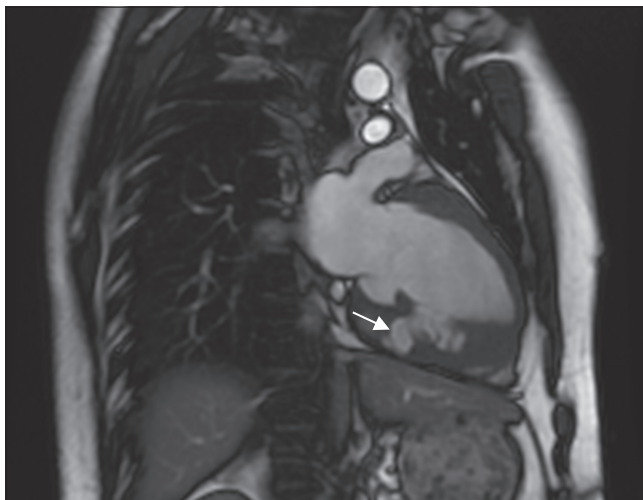
A double right ventricle (RV) is a much more common pathology. This is probably because the RV is normally partially divided into the inlet and outlet portions by the muscular ridge of the supraventricular crest and the septomarginal trabecula. Abnormalities of the various muscle bands in this region are the most frequent cause of a two-chambered RV.[1]

A rare congenital disorder diagnosed in our patient is best classified as double-chambered LV, a term which has been used to describe the subdivision of the LV cavity into 2 chambers by an abnormal septum or muscle bundle. The etiology of this disorder is not well-known, but the anomaly is generally thought to be congenital if it is diagnosed late and is nonprogressive. Only a few cases with variable morphologies have been reported in the literature, mostly with either a diverticular appearance or small contracting chambers attached to the LV lateral wall or within the apex.[2] The exact mechanism of a double ventricle is unknown; however, cardiomyopathy is often an underlying pathology.

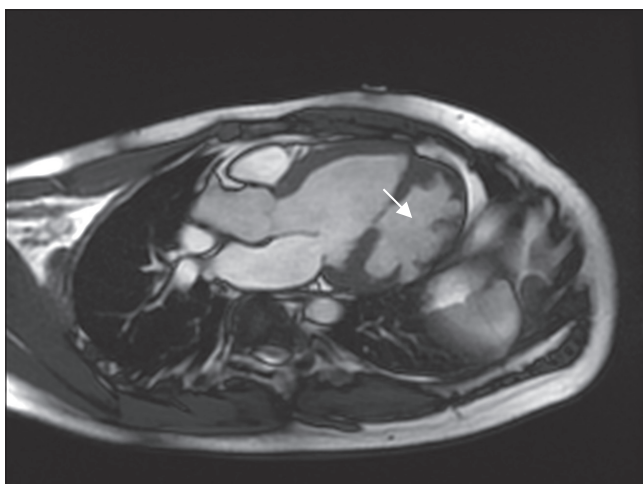
Gerlis et al.[1] reported 3 cases with endomyocardial fibroelastosis and cardiomyopathy. All 3 patients had double-chambered ventricles. All of those patients died in the first year of life from cardiomyopathy. A double-chambered ventricle coexisting with tetralogy of Fallot has also been described.[3] Our patient, in contrast to some other cases shown in the literature, had no echocardiographic evidence of cardiomyopathy and was diagnosed relatively late in life, as no symptoms had been previously reported.

The differential diagnosis is extremely important in trying to define the pathology. Echocardiography and computed tomography can aid in the detection of double-chambered ventricles. Magnetic resonance imaging will characterize the condition better because of its higher spatial resolution and the ability for tissue characterization, especially with the differentiation between fibrosis and normal myocardium.[4]

Detection of an accessory chamber in communication with the LV cavity needs special consideration, because an LV aneurysm or diverticulum can sometimes mimic a double-chambered LV. Therefore, a precise evaluation of their wall motion during systole and diastole as well as the width of their communication with the main LV cavity would help to differentiate between these entities [5]. Even more importantly, the treatment modalities are different for each of these pathologies. The diagnosis with the worst prognosis is an LV pseudoaneurysm, which is a contained rupture of the LV free wall. However, this disease follows chest trauma, myocardial infarction, or endocarditis. An aneurysm of the LV is described as a wall that is thinner than the adjacent myocardial segments, while a diverticulum is an outpouching containing the endocardium, myocardium, and pericardium. Both an aneurysm and a diverticulum need to be resected when symptomatic [6].



**Figure 4.** Cardiac magnetic resonance. The additional cavity is seen (arrow)



**Figure 5.** Cardiac magnetic resonance. A large additional cavity (arrow) consisting of fibrous membrane with some fenestrations and a narrow neck

Another diagnosis that should be taken into consideration is LV noncompaction. In our patient, the ratio between the spongy and compact layers (about 1 in the systole phase) did not fulfill the criteria for this cardiomyopathy.

Accessory cavities sequestered by aberrant muscular ridges are not well recognized. Surgically, they may be resolved by a resection of the aberrant tissue [4]. Indications for surgery must be made for each individual case as there have only been few reports on this pathology. We decided on a conservative treatment for the patient. Our decision was based on the fact that the patient was totally asymptomatic, had no history of thromboembolism, and there seemed to be a low probability of developing thrombus in the additional LV cavity. Another point that we considered important was that the condition did not affect the overall function of the LV.

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