

Double-chambered left ventricle

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We describe a rare congenital anomaly in a 39-year-old woman who was referred for a preoperative echocardiographic assessment in February 2012. A double-chambered left ventricle was suspected on transthoracic echocardiography and was confirmed by transesophageal echocardiography.

Keywords:

double chamber, echocardiography, left ventricle

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Introduction

Double chambered is a term that has been used to describe the subdivision of a ventricle as a result of an anomalous septum or muscle bundle. Subdivision of the left ventricular cavity is a rare cardiac anomaly compared with subdivision of the right ventricle (RV).

We describe a case of double-chambered left ventricle (DCLV), highlighting the importance of echocardiography, both transthoracic and transesophageal, in the detection of double-chambered ventricles.

The case

A 39-year-old woman was referred for a preoperative cardiac assessment because of an accidental discovery of cardiac murmur. The patient was completely asymptomatic. Physical examination was inconspicuous, with a faint cardiac murmur and a regular heart rhythm. The ECG showed a regular sinus rhythm with normal QRS axis and duration, and delayed R-wave progression on the precordial leads. Transthoracic echocardiography indicated atrial situs solitus with atrioventricular and ventriculoarterial concordance; there was moderate left ventricle (LV) systolic dysfunction and an estimated ejection fraction of 40%. The LV was abnormally divided into two distinct contracting LV chambers, separated by a thick-walled muscular septum extending from the apex up to the LV outflow tract, inserted into the base of a noncoronary cusp and causing a mild degree of aortic incompetence. The mitral valve was overriding the major LV chamber, LV inflow was unrestricted, and the minor LV chamber was connected to the major chamber through large fenestrations. The RV was normal in size and function. Color-Doppler echocardiography indicated intact IAS and IVS. These findings were confirmed by transesophageal echocardiography. No significant ventricular arrhythmia was recorded during holter monitoring or during the exercise ECG test. Coronary angiography indicated a normal coronary artery anatomy.

Discussion

This rare congenital disorder is best classified as a 'double-chambered left ventricle', a term that has been

used to describe the subdivision of the LV cavity by an abnormal septum or muscle bundle into two chambers. Only a few cases with variable morphologies have been reported in the literature, most with either a diverticular appearance or small contracting chambers attached to the LV lateral wall or within the apex [1,2].

The differential diagnosis of DCLV includes left ventricular aneurysm or pseudoaneurysm; hence, it is referred to in some studies as an aneurysm-like structure 'ALS'. However, in a DCLV, there is contractile motion of the wall of the ALS that is separated from the main lumen by a membrane or an accessory muscle bundle, whereas LV aneurysm and pseudoaneurysm are out-pouching of the LV wall, connected to the cavity through a neck, and as it lacks the muscular contractile part of the LV free wall, it tends to expand rather than contract during systole [3,4].

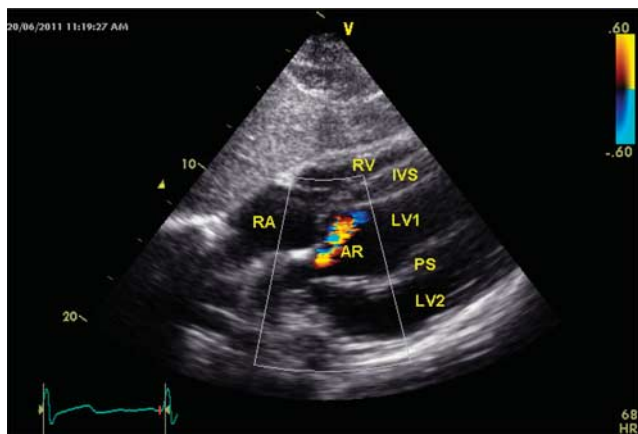
Another pathology related to DCLV, at least literally, is double-chambered right ventricle (DCRV); however, they are clearly distinct pathologies.

DCRV is a more common congenital heart disease presenting early in life with exertional shortness of breath and usually a loud murmur of right ventricular outflow tract (RVOT) obstruction; it is frequently associated with other congenital heart disease, for example fallot's tetralogy, ventricular septal defect (VSD), and transposition of great arteries (TGA), whereas DCLV is usually an asymptomatic disorder discovered accidentally as in our case [5,6].

The pathology in both conditions is also different. In DCRV, there is progressive thickening of the right ventricular septum because of a hypertrophied muscle bundle. This muscle bundle divides the RV into two sequentially contracting RV chambers with a significant gradient in between; this explains the early symptoms, the murmur, and the frequently associated RVH pattern on surface ECG. In DCLV, the two chambers are contracting in parallel and present less of a pressure gradient, as both contract synchronously; thus, it is usually nonobstructive and asymptomatic.

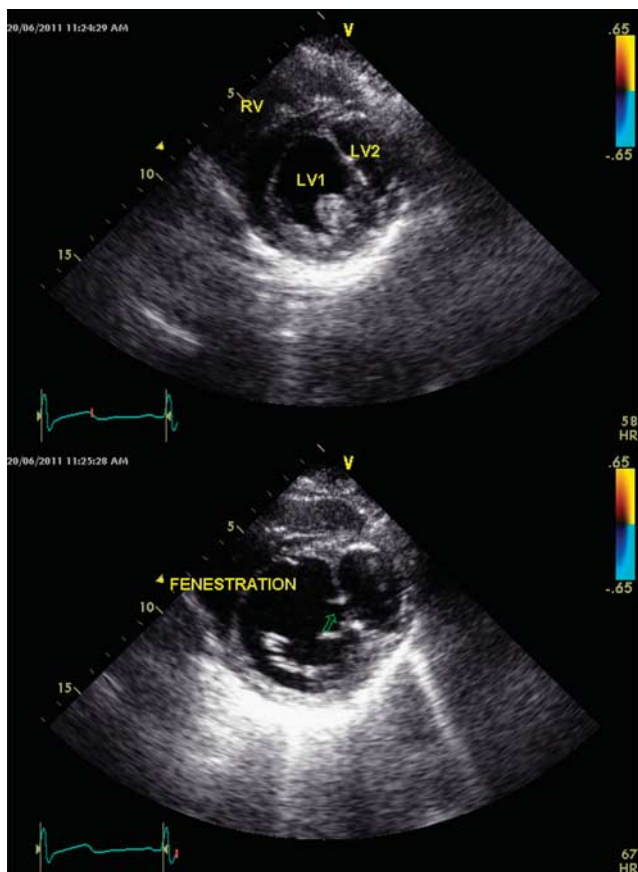
As this is an extremely rare finding, no definite data on the prognosis, outcomes, and potential complications, such as risk of embolism, of DCLV are available. It is generally

Figure 1



Subcostal long axis view showing the full length of the LV pseudoseptum from the apex up to its insertion point into the left ventricular outflow tract (LVOT), at the base of NCC, resulting in mild aortic regurg (Grade 1) (AR). LV, left ventricle, NCC, noncoronary cusp.

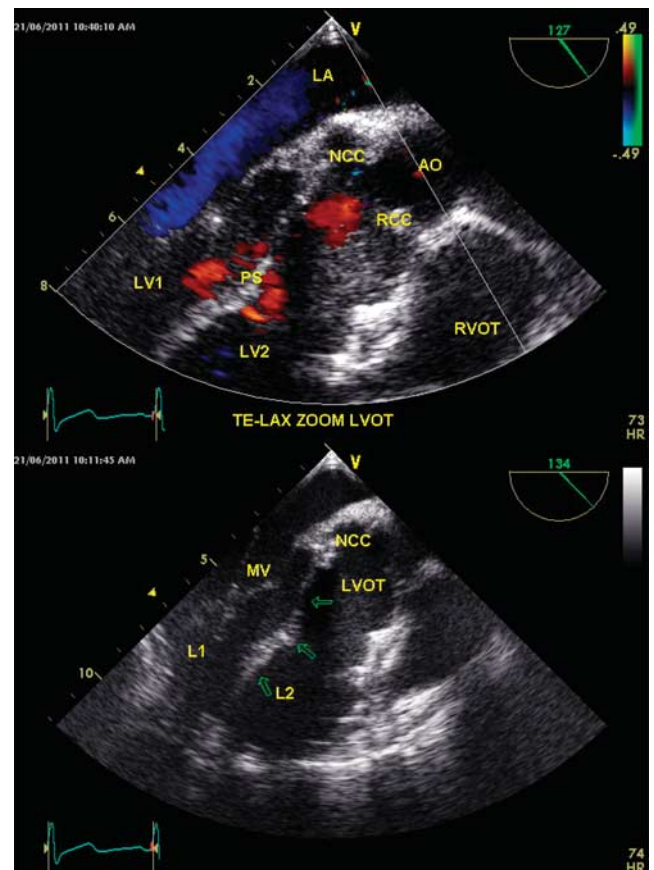
Figure 2



Parasternal short-axis view of multiple LV segments starting from the apex to the base, with a 'double-barreled' heart appearance. The septum-like muscle bundle (middle) divides the left ventricle into a round, larger chamber on the left side (LV₁) and a small, more oval-shaped chamber on the right (LV₂). LV, left ventricle.

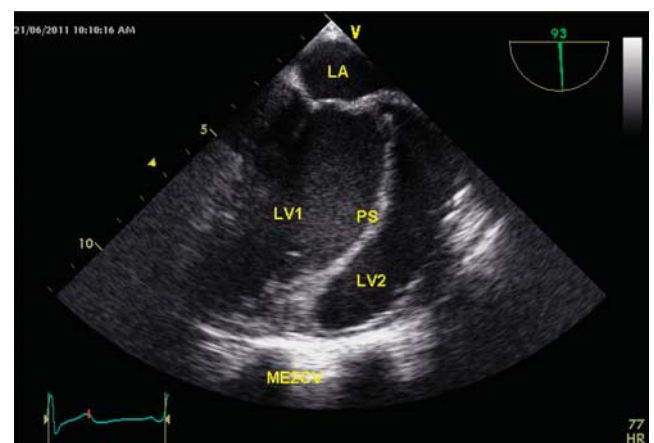
believed that DCLV poses little risk to the patient. Treatment, if any, is usually guided by the presence of other associated abnormalities.

Figure 3



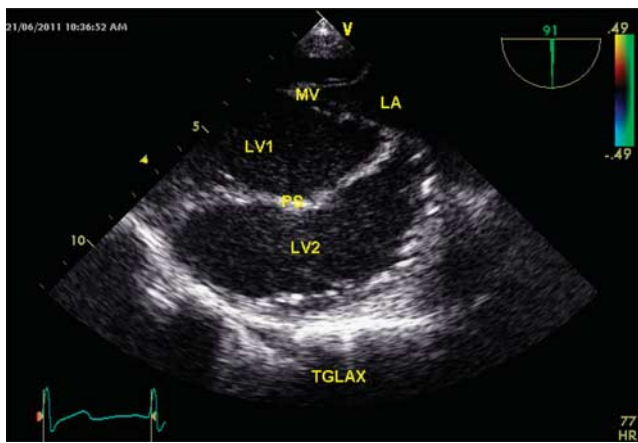
Transesophageal echo, midesophageal long axis (LAX) images showing the site of insertion of pseudoseptum (PS) into the base of NCC and the resultant jet of AR. NCC, noncoronary cusp.

Figure 4



Transesophageal echo, midesophageal, showing the extent of PS down to the apex.

In the case presented, the completely asymptomatic course, suggesting that the patient's condition is not immediately life threatening, eliminated the need for surgical intervention. We had two concerns: first, the possibility that a clot will form in the minor ventricle, although in the patient's case, good blood flowthrough

Figure 5

Transesophageal echo, TGLAX, showing the extent of PS down to the apex.

the communication gap lessened this risk. The other concern was the asymptomatic LV systolic dysfunction; hence, the patient was discharged on carvedilol + telmi-

sartan combination and instructed to follow-up in the cardiac outpatient clinic (Figs 1–5).

Acknowledgements

Conflicts of interest

There are no conflicts of interest.

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