Diagnosis of double-chambered left ventricle by contrast echocardiography: a case report

Lin Jin1, Feifei Yuan2, Ping Li1 and Yingchun Wang1

Abstract
Double-chambered left ventricle (DCLV) is a rare congenital cardiac abnormality. We retrospectively analyzed the diagnosis and treatment of a patient with DCLV who was admitted to Jiading Central Hospital of Shanghai Medical College in August 2019. The initial symptom of the 55-year-old male patient was stroke. He was preliminarily diagnosed with DCLV by echocardiography and this diagnosis was confirmed by contrast-enhanced echocardiography. Our findings indicate that contrast-enhanced echocardiography of the left heart is useful for clinical application in the clinical diagnosis, treatment, and prognosis of DCLV. We also review the relevant literature for our case.

Keywords
Contrast-enhanced echocardiography, contrast agent, congenital heart disease, double-chambered left ventricle, stroke, thick muscle bundle

Introduction
Double-chambered left ventricle (DCLV) is a rare congenital heart disease. In DCLV, the left ventricle is separated into a primary chamber and a secondary chamber by abnormally thick muscle bundles or a fibrous septum.1 These patients are often asymptomatic. Therefore, imaging diagnosis is important for the treatment strategy of...
DCLV, especially in surgical decisions. We report a case of DCLV with stroke as the initial symptom in our hospital. The related literature was also reviewed to examine the value of contrast-enhanced echocardiography in the left heart for diagnosing DCLV.

**Case presentation**

A male adult aged 55 years had symptoms of dizziness, nausea and vomiting, weakness of the right limb and vague speech without obvious inducement. There was no history of hypertension, diabetes, or coronary heart disease. His body temperature was 37°C, pulse rate was 80 beats/minute, respiratory rate was 20 breaths/minute, and blood pressure was 110/70 mmHg. An electrocardiogram showed sinus rhythm and occasional ventricular premature beats. Magnetic resonance imaging showed new and multiple cerebral infarctions in the left cerebellar hemisphere and occipital cortex, and there was no major abnormality in brain magnetic resonance angiography.

Routine echocardiography showed that the patient had normal atrioventricular internal diameters, and normal thickness and motion amplitude of the ventricular septum and left ventricular posterior wall. Additionally, forward blood flow velocity in the main pulmonary artery and aorta, as well as the left ventricular ejection fraction, were normal. A thickened muscle bundle was discovered in the left ventricular cavity, of which one end was connected to the left ventricular lateral wall, while the other end was connected to the apex, dividing the left ventricle into two oblique chambers (Figure 1). The mitral valve and aortic valve were located in the primary chamber. In the resting state, the left ventricular apical wall and apical lateral wall became thinner and bulged slightly outward, and their contraction movement was weakened. In color Doppler flow imaging, blood flowed from the primary chamber into the secondary chamber in diastole, but flowed back in systole.

In contrast-enhanced echocardiography of the left heart (including left ventricular opacification [LVO]), an ultrasound contrast agent (SonoVue, Bracco, Italy) filled the left ventricular cavity and the endocardium was clearly observed. The primary and secondary chambers were connected by multiple transportation channels. Filling of blood was good in the primary chamber, while blood flow was slow in the secondary chamber and appeared as a swirl-like shape. These findings suggested the diagnosis of DCLV. Furthermore, myocardial perfusion in the whole left ventricle was good.

Coronary computed tomography (CT) showed no major stenosis in the left anterior descending artery and the papillary muscles were greatly thickened. A thick muscle bundle separated the left ventricle into two chambers, which was consistent with the echocardiogram (Figure 2).

**Discussion**

Based on the position of the chambers, there are two types of DCLV, including the upper and lower chambers type (A type) and the left and right chambers type (B type). The difference between these types is that the secondary chamber in the A type is located at the apex, and this chamber is located at the lateral wall of the primary chamber in the B type. In this case, an abnormally thick muscle bundle connects the left ventricular side wall with the apex of the heart. Although the primary chamber is located at the base, in which the mitral valve and the aortic valve are located, the volume of the secondary chamber is larger, including the apex and part of the lateral wall. The secondary chamber does not directly connect with the left atrium and aorta. Our patient had no history of cardiac trauma or myocardial infarction. Additionally coronary CT showed that there was no significant
coronary artery disease. Therefore, the double-chamber should not have been caused by cardiac trauma or myocardial infarction.

LVO can improve display of the left ventricular membrane, especially of the endometrium at the left ventricular apex. This improves the diagnostic accuracy of echocardiography for abnormal cardiac structures. In our case, LVO showed that the contrast agent in the primary chamber flowed into the secondary chamber in diastole and the transportation channel was porous. There was no obvious obstruction in the left ventricular inflow and outflow tracts. Therefore, the patient was asymptomatic for many years. The time of appearance and degree of severity of clinical symptoms of DCLV are closely related to the size and function of the transportation channel between the two chambers. In severe cases, symptoms of cardiac insufficiency can occur, such as palpitations, shortness of breath, lower limb edema, and arrhythmia. However, LVO in our case showed that blood velocity in the secondary chamber was slow, in a swirl-like shape. Slow hemodynamics in the

Figure 1. Echocardiogram of a double-chambered left ventricle. (a) In the apical four-chamber view, a large muscle bundle (arrows) divides the left ventricle into two chambers, with the upper chamber as the primary chamber and the lower chamber as the secondary chamber. The mitral valve and the aortic valve are located in the primary chamber. (b) Short-axis view of the left ventricle shows thickened anterolateral papillary muscles (arrow). (c) Apical four-chamber view of contrast-enhanced echocardiography of the left ventricle shows that the primary and secondary chambers are connected by multiple transportation channels (arrows). (d) Apical four-chamber view of contrast-enhanced echocardiography of the left ventricle shows that the left ventricular apical myocardium is thin, but myocardial perfusion is good.
secondary chamber is a risk factor for thrombosis, which is consistent with a study by Gopal et al.\textsuperscript{6} Therefore, administration of anticoagulation is necessary, such as oral warfarin. In our patient, although a history of stroke led to the indications for surgical treatment, there was no systolic dysfunction or left ventricular obstruction. Therefore, we chose to perform medical treatment.

Previous studies have shown that the pathogenesis of DCLV may be due to hyperplasia or incomplete degeneration of trabecular muscle in the middle or apex of the ventricle in the embryonic stage.\textsuperscript{7} Endomyocardial fibroelastosis in the secondary chamber can lead to coronary microcirculation disorders. The coronary microcirculation plays an important role in myocardial perfusion, and therefore, its disorder can cause microvascular angina and increase the risk of adverse cardiovascular events. Consequently, early identification of the cause of DCLV to guide clinical treatment strategies is extremely important to improve the prognosis of patients. Myocardial contrast echocardiography can be used to observe left ventricular myocardial blood perfusion for evaluating the coronary microcirculation.\textsuperscript{4} In our case, myocardial contrast echocardiography showed that the left ventricular myocardium was well perfused. This is helpful for distinguishing DCLV from a ventricular aneurysm and also providing a meaningful reference for the patient’s treatment choice and evaluation of prognosis. Zhang et al.\textsuperscript{8} also found that echocardiography has great clinical value in diagnosis, treatment and evaluation of prognosis of DCLV. However, a 5-year follow up study showed that an asymptomatic 15-year-old boy had electrocardiographic abnormalities.\textsuperscript{9} Transthoracic echocardiography also showed that he had DCLV with mild left ventricular systolic dysfunction.

Figure 2. Computed tomographic image. Cardiac computed tomography shows a thick muscle bundle separating the left ventricle into two chambers.
After 5 years of follow up, he was still asymptomatic, but two-dimensional speckle tracking echocardiography showed subtle progressive deterioration of left ventricular systolic function. Therefore, patients with DCLV require regular visits to hospital to control development of this disease.

In our patient, the findings of CT were consistent with those of echocardiography, which showed anatomical abnormalities of DCLV. However, LVO is able to dynamically observe blood flow between the primary and secondary chambers, and indicate obstruction or an eddy current in real time. Additionally, myocardial contrast echocardiography has more advantages than CT in displaying left ventricular myocardial perfusion.

Because the morphology of DCLV is similar to that of left ventricular diverticulum, left ventricular aneurysm, and left ventricular myocardial noncompaction, DCLV needs to be differentiated from these conditions during an ultrasonic examination. Left ventricular diverticulum is a type of local myocardial tissue with weak development or a defect, and it bulges to the outside of the heart. Ultrasonic appearance of a diverticulum shows a cystic structure outside of the heart cavity. The diverticulum is connected with the ventricle by a relatively narrow channel. The difference between left ventricular diverticulum and DCLV is that there is no abnormal tissue separation in left ventricular diverticulum. A ventricular aneurysm is due to local myocardial ischemia or infarction caused by local bulging. In this condition, necrosis of left ventricular wall heart muscle is replaced by fibrous scar tissue, the wall of local left ventricular is thin, and systolic movement amplitude disappears or is even reversed, with outward expansion. Noncompaction of the myocardium refers to failure of normal myocardial thickness in the embryonic stage. This leads to the persistent presence of intra-trabecular myocardial sinusoids, abnormal development of trabeculae, and a reduction in dense myocardium in corresponding regions.

In conclusion, contrast-enhanced echocardiography of the left heart can clearly show the size and function of the primary and secondary chambers of DCLV and effectively determine the degree of obstruction between these two chambers. This visualization is important in the clinical diagnosis, treatment, and assessment of prognosis for DCLV.

Declaration of conflicting interest

The author(s) declared no potential conflicts of interest with respect to the research, authorship, and/or publication of this article.

Ethics statement

The study protocol was thoroughly reviewed and approved by the Medical Ethics Committee of Jiading District Central Hospital. The patient provided written informed consent for publication of this report.

Funding

The author(s) disclosed receipt of the following financial support for the research, authorship, and/or publication of this article: This work was supported by Jiading District Agricultural and Social Enterprise Research Project (No. JDKW-2017-W14) and Jiading District Health and Family Planning Commission Scientific Research Project (No: 2018-QN-06).

ORCID iDs

Lin Jin https://orcid.org/0000-0002-7912-4836
Feifei Yuan https://orcid.org/0000-0002-5582-0765
Ping Li https://orcid.org/0000-0001-7370-308X
Yingchun Wang https://orcid.org/0000-0003-4820-1672
References

1. Maschi PG, Pucci A, Fontanive P, et al. Double-chambered left ventricle in an asymptomatic adult patient. *Eur Heart J Cardiovasc Imaging* 2012; 13: E1–E3.

2. Wang XF. *Echocardiography*. Beijing: People’s Medical Publishing House, 2015, p.11.

3. Bilici M, Demir F, Akın A, et al. Echocardiographic diagnosis of double-chambered left ventricle. *J Echocardiogr* 2016; 14: 176–178.

4. Porter TR, Mulvagh SL, Abdelmoneim SS, et al. Clinical applications of ultrasonic enhancing agents in echocardiography: 2018 American Society of Echocardiography guidelines update. *J Am Soc Echocardiogr* 2018; 31: 241–274.

5. Sharma S, Dinwoodey DI, Chaudhry GM, et al. Congenital double-chambered left ventricle presenting as monomorphic ventricular tachycardia. *CASE (Phila)* 2019; 3: 51–55.

6. Ghosh GC, Alex AG, George PV, et al. ‘Chamber within a chamber’: a rare cardiac anomaly. *Cardiol Young* 2017; 27: 1423–1425.

7. Demirel M, Toprak C, Tabakçı MM, et al. Case images: cardiac hydatid cyst presenting as double-chambered left ventricle. *Turk Kardiyol Dern Ars* 2016; 44: 350.

8. Xin Z, Xiaolin Z, Yongli C, et al. The value of echocardiography on the diagnosis of the double-chambered left ventricle. *Chinese Journal of Ultrasound in Medicine* 2017; 33: 643–646.

9. Harada K, Mori K, Ichimiya C, et al. Five-year follow-up two-dimensional speckle tracking echocardiography in a juvenile with a double-chambered left ventricle. *Echocardiography* 2017; 34: 791–793.

10. Makkuni P, Kotler MN and Figueredo VM. Diverticular and aneurysmal structures of the left ventricle in adults. *Tex Heart Inst J* 2010; 37: 699–705.

11. Novo G, Dendramis G, Marrone G, et al. Left ventricular noncompaction presenting like a double-chambered left ventricle. *J Cardiovasc Med* 2015; 16: 522–524.