CASE REPORT

Congenital Double-Orifice Mitral Valve in Asymptomatic Patients
Comprehensive Assessment by Three-Dimensional Echocardiography
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Summary
Congenital double-orifice mitral valve (DOMV) is a rare malformation that may affect both mitral valve and apparatus, and it is usually associated with other cardiac disorders. Asymptomatic DOMV is extremely rare. We present two similar cases of asymptomatic congenital double-orifice mitral valve. Both cases presented functionally normal mitral valve apparatus with two equally sized orifices, which were formed by a central fibrosis. In addition, final confirmatory diagnosis was made using three-dimensional (3D) echocardiography in both cases.

Key words: Valvular disease, Fibrous bridge, Transthoracic

Double-orifice mitral valve (DOMV) is an uncommon congenital malformation characterized by the presence of fibrous bridge and two separate valve orifices. First recognized by Greenfield in 1876. It was thought to be rare since it only accounts for 1% of congenital heart diseases. It is generally associated with other congenital abnormalities, including aortic coarctation, atrial septal defect, patent ductus arteriosus, or complex congenital heart disease. About half of patients had reasonable function, whereas others presented with significant mitral stenosis or regurgitation. Most cases can be diagnosed by transthoracic two-dimensional echocardiography (2D TTE). However, it is difficult to recognize it in asymptomatic patients. Transthoracic three-dimensional echocardiography (3D TTE) provides a reliable and comprehensive assessment of mitral valve and associated malformations. We describe two asymptomatic cases of DOMV fully assessed by 3D TTE.

Case Reports
Case 1: A 57-year-old man was referred to our hospital for cardiac assessment. His blood pressure was 120/80 mmHg and heart rate 76 bpm on physical examination. The heart sound was normal with auscultation. Electrocardiography (ECG) and chest X-ray were inconclusive. The parasternal long axis view showed a restricted movement of the mitral valve (Figure 1A); however, the parasternal short axis view revealed double-orifice mitral valve with equal size in mitral position (Figure 1B). There was trivial mitral regurgitation in mitral position. Color Doppler imaging showed that the blood flow in the mitral valve was divided into two parts (Figure 1C), and pulsed wave Doppler confirmed no mitral stenosis in this regard (Figure 1D). 3D TTE showed that the mitral valve was divided into two adequate orifices by a central bridge of fibrous tissue, and the papillary muscles were normal with chordate inserting into two papillary muscles (Figure 2). The patient was scheduled for follow-up.

Case 2: A 20-year-old man presented for further cardiac examination. He was in good physical condition with blood pressure 120/80 mmHg and heart rate 93 bpm. His ECG and chest X-ray were without remarks. 2D TTE showed normal cardiac dimension and function. The aortic valve was bicuspid without stenosis or regurgitation. The parasternal long axis view demonstrated restricted movement of the mitral valve, but adequate opening was visualized when slightly rotating the transducer. The parasternal short axis view revealed double-orifice mitral valve of approximately equal size in mitral position. The mitral flow was normal without regurgitation. 3D TTE confirmed central fibrous bridge in the middle of mitral valve without other associated abnormalities except bicuspid aortic valve (Figure 3). Since there was no significant mitral regurgitation or stenosis, the patient underwent periodic follow-up.

Discussion
DOMV is a relatively rare congenital heart disease, which may affect both mitral valve and subvalvular apparatus. Isolated DOMV is extremely rare. The embryologic mechanisms of DOMV include abnormal fusion of dorsal endocardial and left lateral endocardial and persistence of left part of the common atrioventricular canals.

Trowitzsch et al. divided the anomaly into three

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Figure 1. Conventional transthoracic echocardiography views. A: The parasternal long axis view showed restricted movement of the mitral valve (white arrow). LA indicates left atrium; LV, left ventricle; RV, right ventricle; and AO, aorta. B: The parasternal short axis view revealed double-orifice mitral valve in mitral position (asterisk sign). C: Color Doppler echocardiography demonstrated the presence of two separate inflow jets. D: Pulsed wave Doppler imaging showed a normal blood flow velocity in mitral valve.

Figure 2. Three-dimensional echocardiography (3D Flexi Slice mode) showed the orifices (asterisk sign) and the papillary muscles (arrows). A (apical four-chamber view) and B (apical long-axis view) show the slices we chose; C is the image at the level of central bridge; and D represents the images of slices at the four levels (papillary muscle, chordal attachment, central bridge, and mitral valve).
Figure 3. Three-dimensional echocardiography confirmed the presence of central fibrous bridge (arrows) in the middle of mitral valve. A: The central fibrous bridge observed from the left atrium. B: The central fibrous bridge observed from the left ventricle.

types according to echocardiographic manifestation: complete bridge type, incomplete bridge type, and hole type. The clinical manifestation and management not only depend on the severity of mitral valve dysfunction, but also on the associated malformations. Both of our cases were complete bridge type, with a fibrous bridge dividing the left atrioventricular orifice into approximately equal parts, as demonstrated by 3D TTE.

Although the DOMV diagnosis mainly depends on two-dimensional and Doppler echocardiography, it may be difficult to recognize it in patients with no obvious morphological and hemodynamic characteristics. In addition, it may be even more difficult to recognize and completely assess the mitral valve function and associated malformation in patients with poor image quality. In our cases, the parasternal long axis view showed stenosis of mitral valve at first, but when we adjusted the transducer, the mitral valve showed normal opening. Therefore, sonographers should be trained to obtain the delicate images to make an appropriate diagnosis.

Compared with 2D TTE, 3D TTE in addition to providing a more vivid mitral valve anatomic structures and function, also demonstrates a more accurate size and spatial relationships among the intracardiac structures to help define the associated anomalies of subvalvular apparatus, including redundant chordal attachment, chordal ring. Thus, 3D TTE is a diagnostic imaging method complementary to 2D TTE. Combining 2D and 3D TTE is extremely beneficial in the complete cardiac assessment and management of DOMV. Additionally, surgery is required when there are other complex cardiac malformations present, complicated with severe stenosis, insufficiency, or both. Thus, a complete assessment of mitral valve abnormality including apparatus is necessary during clinical practice. In our patients, 3D TTE not only helped to confirm the presence of central fibrous bridge in the absence of any significant mitral stenosis or regurgitation, but it also excluded severe subvalvular malformation. Finally, both the patients were discharged without any medical interventions.

Disclosures

Conflicts of interest: None.

References