

Three-Dimensional Echocardiographic Reconstruction of Double-Orifice Mitral Valve and Mitral Leaflet Prolapse

In-Cheol Kim, MD, PhD; Yun-Kyeong Cho, MD, PhD; Hyungseop Kim, MD, PhD;
Nam-Hee Park, MD, PhD; Kwon-Bae Kim, MD, PhD

A 51-year-old man was referred to our cardiology department for examination of a systolic murmur. Physical examination indicated a grade 3 systolic murmur at the fourth left intercostal space. Transthoracic echocardiography (TTE; Figure 1A) and heart dynamic computed tomography (Figure 1B) demonstrated a double-orifice mitral valve (DOMV) with 4 papillary muscles (triangles). Two valve orifices were separated by a complete bridge (arrow). The posteromedial mitral valve was larger, and the anterolateral mitral valve was smaller. The anterior leaflet of the anterolateral mitral valve prolapsed owing to chordae rupture, while color Doppler showed severe mitral regurgitation (Figure 2, Movie I in the online-only Data Supplement). No other combined congenital abnormalities were detected. Three-dimensional TTE clearly showed two orifices of the mitral valve (Movie II in the online-only Data Supplement). Transesophageal echocardiography (TEE) confirmed the diagnosis of DOMV and mitral valve prolapse (Figure 3). Three-dimensional TEE provided a better understanding of the mitral valve and the prolapsed leaflet (Figure 4, Movie III in the online-only Data Supplement).

Discussion

Double orifice mitral valve is a rare congenital defect characterized by 2 orifices in the left atrioventricular valve area that usually possess independent chordal attachment to papillary muscles. After the first report by Greenfield in 1876, various types of DOMV have been reported according to the morphology of DOMV and combined abnormalities. Three-dimensional echocardiography has recently become available and has extended the morphological understanding of the disease to over conventional 2-dimensional echocardiography.¹

Combined heart defects are common, but DOMV can also occur as an isolated congenital anomaly. The morphology

of DOMV in our case was also an isolated congenital defect with complete bridge type (type 1), of the other 3 types described previously.² Each mitral valve has 2 separate mitral leaflets, 2 papillary muscles, and attached chordae tendinae that could be clearly visualized using both TTE and heart dynamic computed tomography scan. Previously reported cases of DOMV had single or multiple papillary muscles.^{3,4} However, previous echocardiography techniques had lower resolution; therefore, an accurate papillary muscle count could be determined only by open heart surgery or autopsy. In the present case, we could identify 2 complete sets of mitral valves including 4 papillary muscles and related structures by using a multimodal approach of TTE, heart dynamic computed tomography scan, and TEE. Three-dimensional TEE also provided valuable information about the location of the prolapsed site from the complex mitral valve anatomy.

Disclosures

None.

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From the Division of Cardiology, Department of Internal Medicine (I.-C.K., Y.-K.C., H.K., K.-B.K.), and the Department of Thoracic Cardiovascular Surgery (N.-H.P.), Keimyung University Dongsan Medical Center, Daegu, Republic of Korea.

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Correspondence to Yun-Kyeong Cho, MD, PhD, 56 Dalseong-no, Jung-gu, Daegu, 700-712, Republic of Korea. E-mail ds010042@gmail.com (*Circulation*. 2014;130:e87–e88.)

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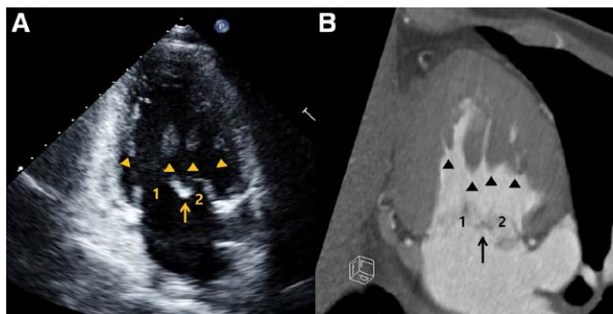


Figure 1. Two-chamber view on transthoracic echocardiography (A) and heart dynamic computed tomography (B) showing a double-orifice mitral valve with 4 papillary muscles (triangles) and a bridge between the 2 mitral valve orifices (arrow). A large posteromedial mitral valve (1) and small anterolateral mitral valve (2) are indicated.



Figure 2. Apical 2-chamber view on transthoracic echocardiography showing mitral regurgitation. Note the flow convergence at the anterolateral portion of the mitral valve (triangle) and the regurgitant flow in the opposite direction (arrow).

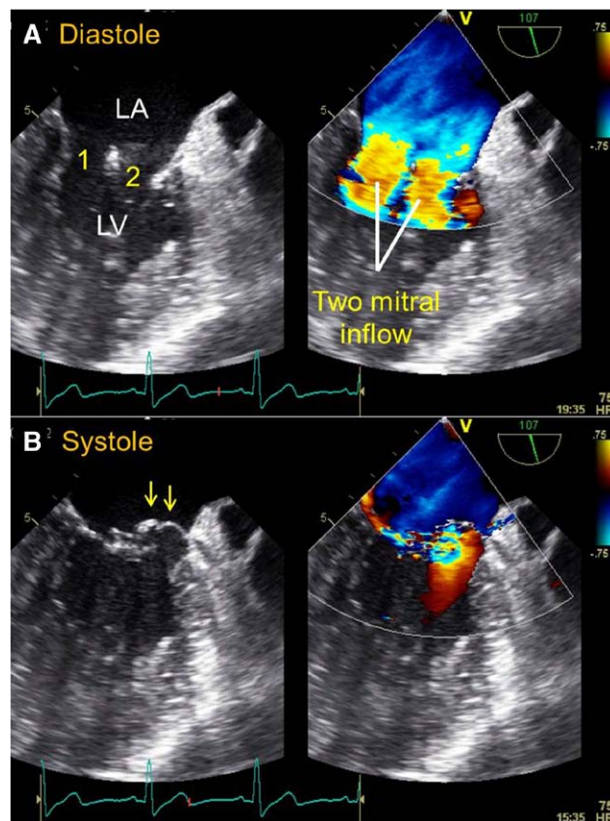


Figure 3. Two-dimensional and color Doppler transesophageal echocardiography image. A, Diastolic phase showing the double orifice of the mitral valve with dual mitral inflow. B, Systolic phase showing prolapse of the anterior leaflet of the anterolateral mitral valve (arrow) and mitral regurgitation color flow. LA indicates left atrium; and LV, left ventricle.

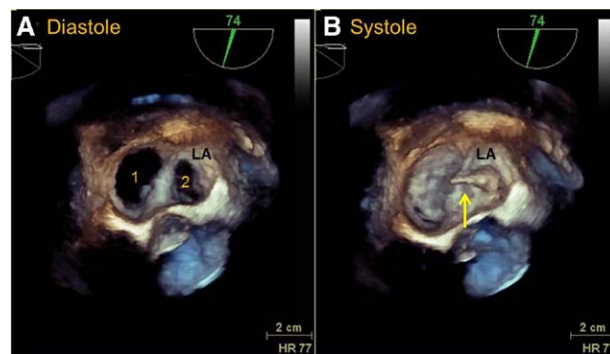


Figure 4. Three-dimensional transesophageal echocardiography images of the diastolic (A) and systolic (B) phase showing 2 mitral valves and mitral valve prolapse (arrow) from the aspect of the left atrium. LA indicates left atrium.

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