A 35-year-old woman was referred for further cardiovascular assessment after an incidental finding of a systolic murmur and symptoms of exertional chest tightness. On examination, she had an anacrotic pulse with an aortic ejection systolic murmur. Two-dimensional (2D) transthoracic echocardiogram (TTE) demonstrated normal left ventricular (LV) size and systolic function with a thickened and calcified bicuspid aortic valve. The mean gradient was 47 mm Hg, and valve area was 0.8 cm², consistent with severe aortic stenosis. There was mild coarctation of the aorta with a peak velocity of 2.5 m/s. The mitral valve appeared structurally abnormal (Figures 1 and 2 and online-only Data Supplement Movies I and II). However, 2D TTE images were suboptimal. Therefore, 2D transesophageal echocardiography (TEE) was performed. This confirmed a bicuspid aortic valve with severe aortic stenosis and mild aortic regurgitation with no associated aortopathy. The images were suggestive of 2 separate mitral valve orifices (Figure 3 and online-only Data Supplement Movie III) without stenosis (mean gradient, 2 mm Hg) and only trivial mitral regurgitation. Real-time 3-dimensional (3D) TEE was performed with the use of the Philips X72T probe for further clarification. This clearly demonstrated division of the mitral valve into 2 separate orifices, each with its own leaflets and subvalvular apparatus, consistent with a double-orifice mitral valve (DOMV) (Figures 4 and 5 and online-only Data Supplement Movies IV and V) and coexisting bicuspid aortic valve (Figure 6 and online-only Data Supplement Movie VI). The images demonstrated

Figure 1. Two-dimensional TTE short-axis view of the mitral valve. RV indicates right ventricle.

Figure 2. Two-dimensional TTE 2-chamber apical view showing a structurally abnormal mitral valve. LA indicates left atrium.

Figure 3. Two-dimensional TTE 4-chamber view. LV indicates left ventricle.

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a bridge-subtype DOMV with a central bridge of abnormal leaflet tissue connecting the 2 leaflets that divided the orifice into unequal medial and lateral parts. The planimetry area of the 2 orifices with the use of offline advanced quantification software was 1.6 cm² and 2.0 cm² respectively.

DOMV is a rare congenital malformation that may be associated with other anomalies of the heart.1 It is characterized by a mitral valve annulus with accessory bridging fibrous tissue that separates it into 2 anatomically distinct orifices of usually unequal size.

To our knowledge, this is the first report of real-time 3D TEE evaluation of a DOMV. Three-dimensional echocardiography is a novel and rapidly evolving imaging modality that is complementary to 2D imaging in the assessment of cardiovascular function and anatomy. There is evidence that it provides improved accuracy and reproducibility over 2D methods in the assessment of LV volumes, LV function, and the assessment of mitral valves.2

Three-dimensional TTE has been found previously to provide a more reliable assessment of DOMV and incrementally more information than that obtained by 2D echocardiography alone.3,4 However, 3D TTE often is limited by suboptimal image quality, and, in comparison, 3D TEE offers superior resolution of the mitral valve. Three-dimensional TEE clearly demonstrated all components of the mitral valve apparatus and allowed identifi-

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**Figure 3.** Two-dimensional TEE view suggesting the presence of 2 distinct orifices within the mitral valve (arrows). LA indicates left atrium.

**Figure 4.** Real-time 3D TEE atrial en face “surgical” view of the mitral valve confirming a double-orifice mitral valve as the “central” type with a bridge of abnormal tissue (arrow). ANT indicates anterior leaflet; POST, posterior leaflet; MED, medial orifice; and LAT, lateral orifice.

**Figure 5.** Real-time 3D TEE LV en face view of the mitral valve. MED indicates medial orifice; LAT, lateral orifice; ANT, anterior leaflet; and POST, posterior leaflet.

**Figure 6.** Real-time 3D TEE view demonstrating bicuspid aortic valve with thickened, calcified aortic cusps causing moderate to severe aortic stenosis. ANT indicates anterior cusp; POST, posterior cusp.
cation of the specific subtype of DOMV and accurate quantification of the mitral valve area by planimetry.

Disclosures

None.

References


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