Windows, Wings and Wonder: Advanced Cardiac Imaging Via Transesophageal Two-Dimensional and Transthoracic Three-Dimensional Echocardiography for the Accurate Diagnosis of Double-Orifice Mitral Valve in the Background of Primum Atrial Septal Defect: A Case Report

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Abstract

The assessment of mitral valve anatomy can be difficult or even misleading on standard cardiac imaging procedures. Such is the scenario in a 40-year-old woman with an incidental finding of atrial septal defect (ASD) and consideration of a trileaflet versus double-orifice mitral valve (DOMV) on transthoracic two-dimensional echocardiography. Further interrogation using transesophageal two-dimensional and transthoracic three-dimensional (3D) echocardiography (to allow more accurate assessment and simultaneous viewing of multiple imaging planes) revealed the typical “seagull wing” configuration of the mitral valve on long-axis view and two distinct but nearly equal-sized orifices on short-axis view, one oriented anteriorly and the other posteriorly. The patient’s DOMV was classified as an incomplete bridge type as confirmed on 3D imaging, with each orifice having its own set of papillary muscles and chordal attachments. The mitral regurgitation was graded as severe, resulting from leaflet prolapse of the anterior orifice. No left ventricular outflow tract obstruction was demonstrated in this case, and the overall left ventricular systolic function was preserved. The ASD was of the primum type, with the configuration and exact location verified through 3D imaging. The accurate detection of congenital anomalies via noninvasive techniques and complementary advanced modalities is vital for preoperative planning, as well as anticipation of potential complications related to the structural anomalies. To our knowledge, this is the first local report of DOMV with primum ASD in an adult.

KEYWORDS: double-orifice mitral valve, primum atrial septal defect, transesophageal and three-dimensional echocardiography

INTRODUCTION

The double-orifice mitral valve (DOMV) is a very rare congenital anomaly first discovered postmortem in 1876 by Greenfield,[1] with only several hundred cases being reported worldwide since through case reports and necropsy-based series.[2,3] The rarity may be a result of underdetection, as most cases of DOMV are incidentally discovered only upon echocardiography for other more common congenital malformations, such as primum atrial septal defect (ASD), atrioventricular canal defect, ventricular septal defect, coarctation of the aorta, patent arterial duct, interrupted aortic arch, or subaortic stenosis.[4] Moreover, no clinical presentation is considered specific for DOMV.[5] In our patient's case, both the primum ASD and the DOMV were incidental findings, previously undetected in past consultations. The use of transthoracic three-dimensional (3D) echocardiography to complement and augment the findings in transthoracic and transesophageal two-dimensional (2D) echocardiography in a more comprehensive assessment of mitral valve anatomy is demonstrated in this case.

REPORT OF CASE

Clinical Summary

A 40-year-old woman sought specialist consultation for an incidental finding of suspected ASD on transthoracic 2D echocardiogram as part of her cardiovascular workup prior to planned hysterectomy. Her medical history was quite unremarkable, with neither report of perinatal or childhood diseases nor comorbid medical conditions such as hypertension or diabetes. Her obstetric history was likewise noncontributory, with four successful pregnancies, three of which were via spontaneous vaginal delivery. She only reported a history of mild exertional dyspnea, which she initially attributed to her obesity, something that she had recently addressed with dieting. Past physical examinations failed to disclose any significant murmurs, and there was no hint of any congenital heart disease by history or physical examination.

Echocardiographic Findings

The unusual M-mode pattern gives the first clue as to the anatomic variation in this patient's mitral valve apparatus (Figure 1). The multiple linear wave patterns suggest the presence of superfluous leaflets and chordae. The parasternal long-axis view (Figure 2) showed a "seagull's wing" configuration typical of a DOMV, with two mitral valve openings, one located in the usual position (orifice 1) and a more anteriorly situated orifice (orifice 2) located proximal to the aortic valve (AV). This dual-orifice configuration was not evident during systole (Figure 2A) but was more visible during diastole (Figure 2B). The two orifices had two distinct leaflets each (a longer anterior leaflet and shorter posterior leaflet), with simultaneous excursion throughout the cardiac cycle.

The posterior orifice (orifice 1) seemed to have a smaller "annulus" with an apically tethered posterior leaflet. The anterior orifice (orifice 2) opened directly toward the anterior interventricular septum, as opposed to orifice 1, which was directed toward the left ventricular (LV) apex. The anterior leaflet of orifice 2 appeared to obstruct the LV outflow tract only during maximal diastolic excursion (Figure 2B).

Transesophageal 2D echocardiography allowed a better short-axis view of the mitral valve using a deep transgastric approach (Figure 3). This view revealed an anterior-posterior orientation of the two orifices with respect to each other. Angulation of the probe to 120 degrees gave a long-axis view of the mitral valve and its subvalvar apparatus, showing the presence of multiple small papillary muscles with their respective chordal attachments (Figure 4). At the midesophageal level, color Doppler showed a dual LV inflow pattern across the two orifices during diastole (Figure 5). During systole, two regurgitant jets were demonstrated, with a predominant mitral regurgitation at the area of the more anterior orifice (Figure 6). This was likely due to systolic prolapse of the redundant leaflets of this particular orifice.

A transthoracic 3D echocardiography was performed to reconstruct the mitral valve appearance from a 3D perspective. Through multiplanar reconstruction, a 3D image of the mitral valve was created, allowing better visualization of the spatial relationships of various structures in the neighborhood of the mitral valve. Figure 7 shows how the full volume image was cropped to allow simultaneous viewing of the long and short axes of the mitral valve apparatus. Meanwhile, Figure 8 shows how the full volume image was cropped to unveil an "en-face" view of the anterior orifice from the LV outflow tract perspective, something that was technically impossible via transthoracic and transesophageal 2D echocardiography. Such a maneuver was crucial because the two orifices were in fact oriented nearly perpendicular to each other. The multiplanar advantage of 3D is once again highlighted in Figure 9, as it illustrates how structures are visualized from unconventional viewing planes not possible with 2D. The "figure-of-8" configuration of the mitral valve orifices is shown in this image.

Serial cropping of the mitral valve apparatus from the LV aspect allowed comparison of the en-face views of the mitral valve at different levels. Figure 10 demonstrates how 3D imaging verified diagnosis of an incomplete bridge type of DOMV, with absence of intervening tissue at the basal portion of the mitral valve.

The ASD noted on transthoracic 2D echocardiography was confirmed to be of the primum type, as transesophageal views clearly showed the defect location at the most inferior portion of the interatrial septum (Figure 11). Note that the midportion of the septum (areas of the fossa ovalis) was thinned out but still intact and deviated to the right atrium, indicating increased left atrial pressure. Color Doppler showed mosaic color flow across the defect, with a left-to-right shunt and a pulmonic (Qp) to systemic (Qs) flow ratio of 1.5:1. Additional 3D echocardiography revealed an ellipsoid ASD (Figure 12), with maximum dimensions at end-systole (anteroposterior diameter = 2.8 cm, superoinferior diameter = 0.88, area = 2.30 cm^2) by 3D echocardiography.

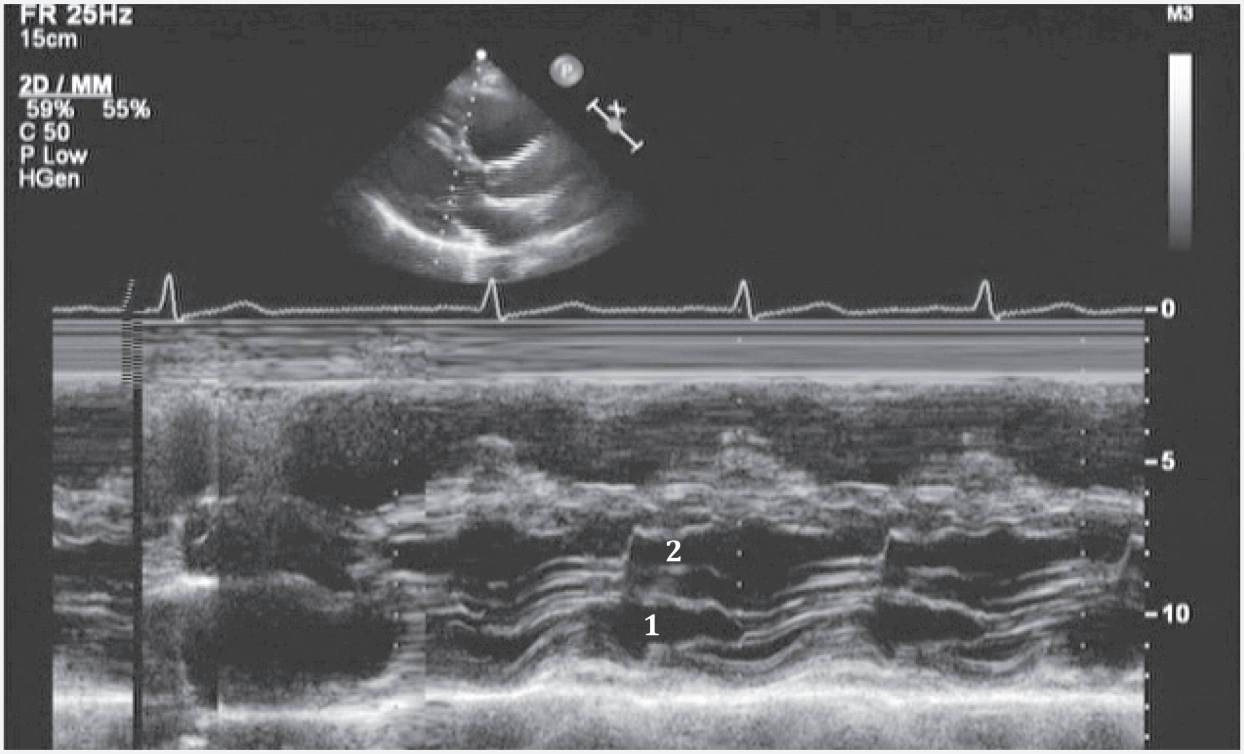

Figure 1. Unusual M-mode pattern of the mitral valve demonstrating multiple linear echodensities, suggesting the suspected abnormal mitral waveform patterns created by orifices 1 and 2.

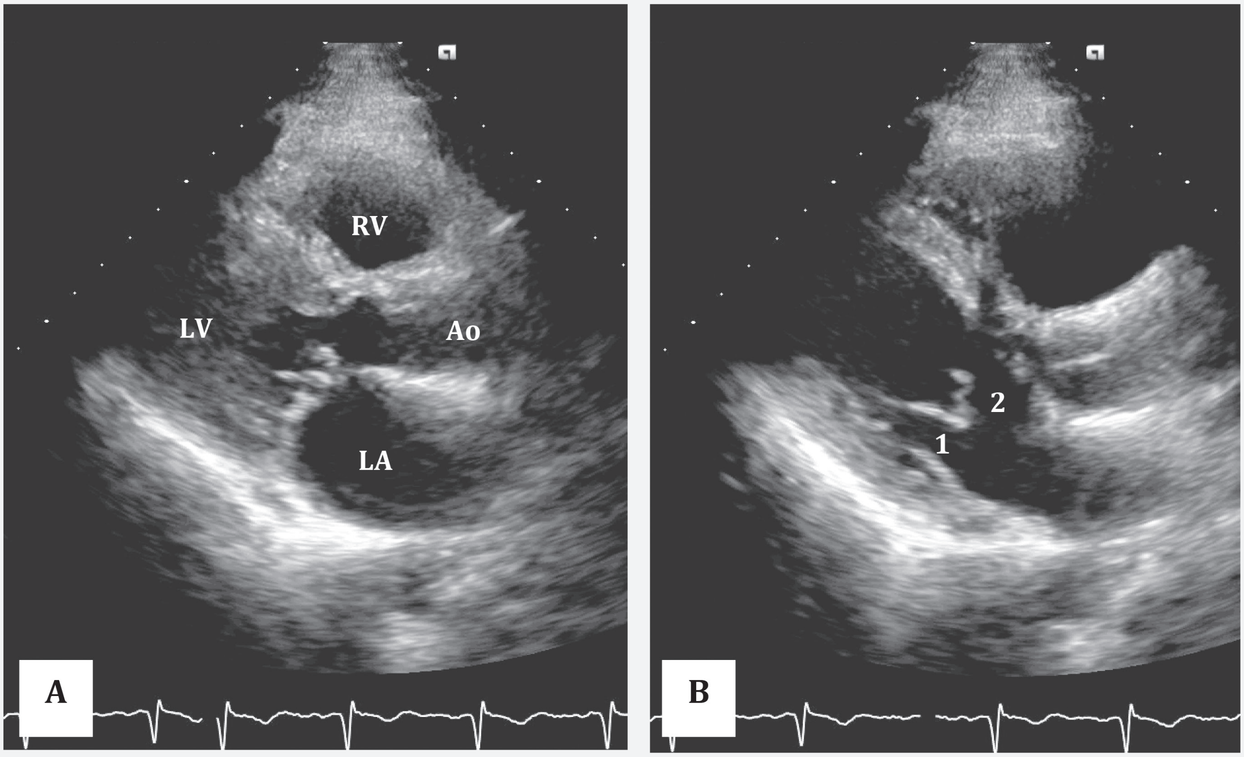

Figure 2. Transthoracic 2D echocardiography parasternal long-axis view of the mitral valve during systole (A) and diastole (B). Note the "seagull's wing" configuration of the central portion of the mitral valve during diastole, separating orifices 1 and 2. Abbreviations: 2D, two-dimensional; Ao, aorta; LA, left atrium; LV, left ventricle; RV, right ventricle.

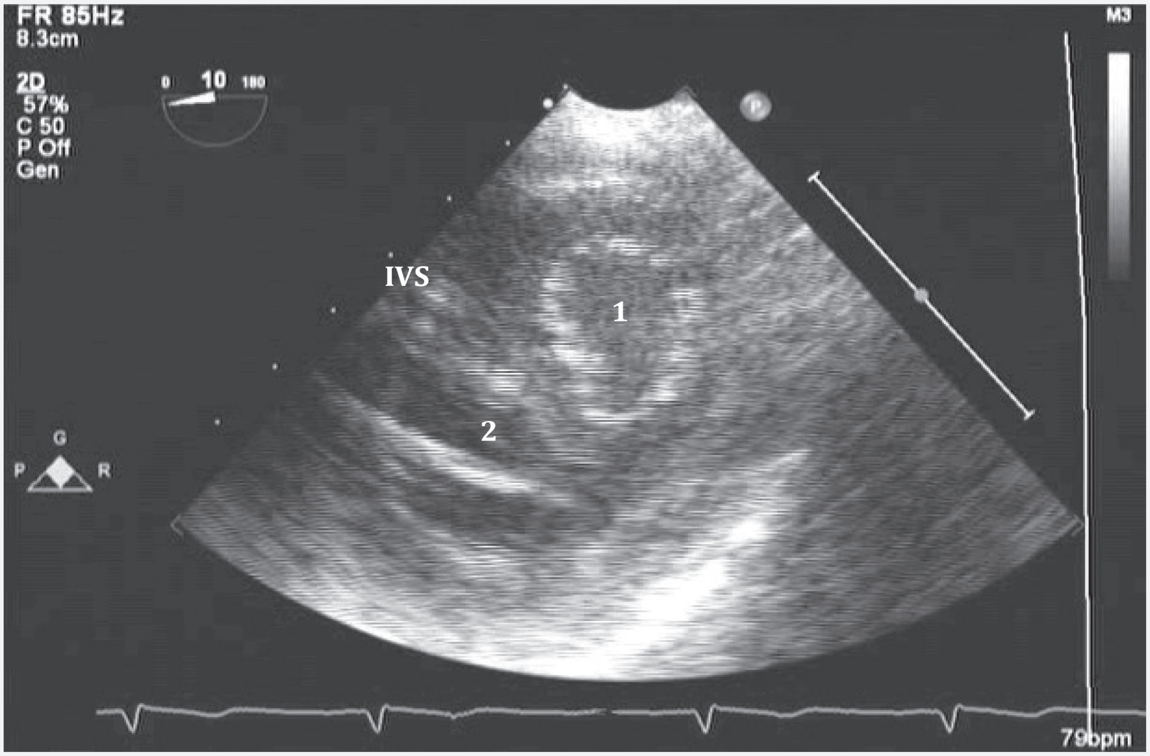

Figure 3. Transesophageal 2D echocardiography deep transgastric short-axis view of the mitral valve demonstrating two mitral valve orifices—one located posteriorly (1) and the other more anteriorly (2). Abbreviations: 2D, two-dimensional; IVS, interventricular septum.

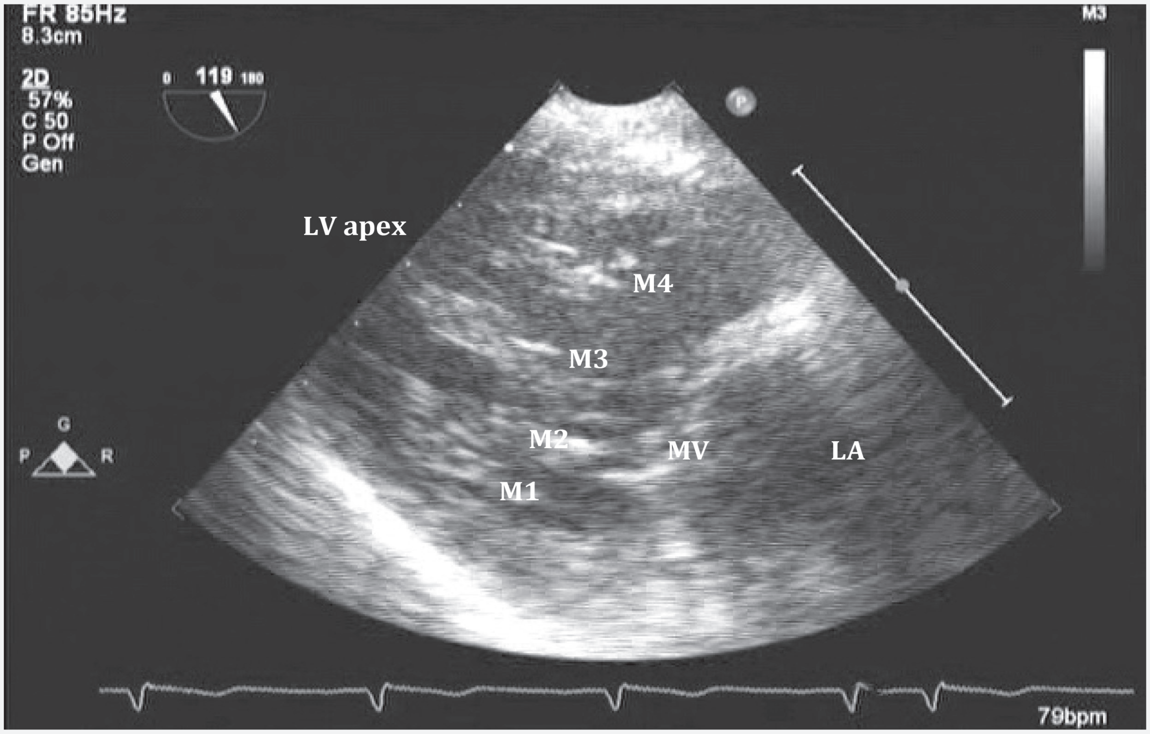

Figure 4. Transesophageal 2D echocardiography deep transgastric long-axis view of the mitral valve demonstrating chordal attachments to multiple papillary muscles (M1–M4). Abbreviations: 2D, two-dimensional; LA, left atrium; LV, left ventricle; MV, mitral valve.

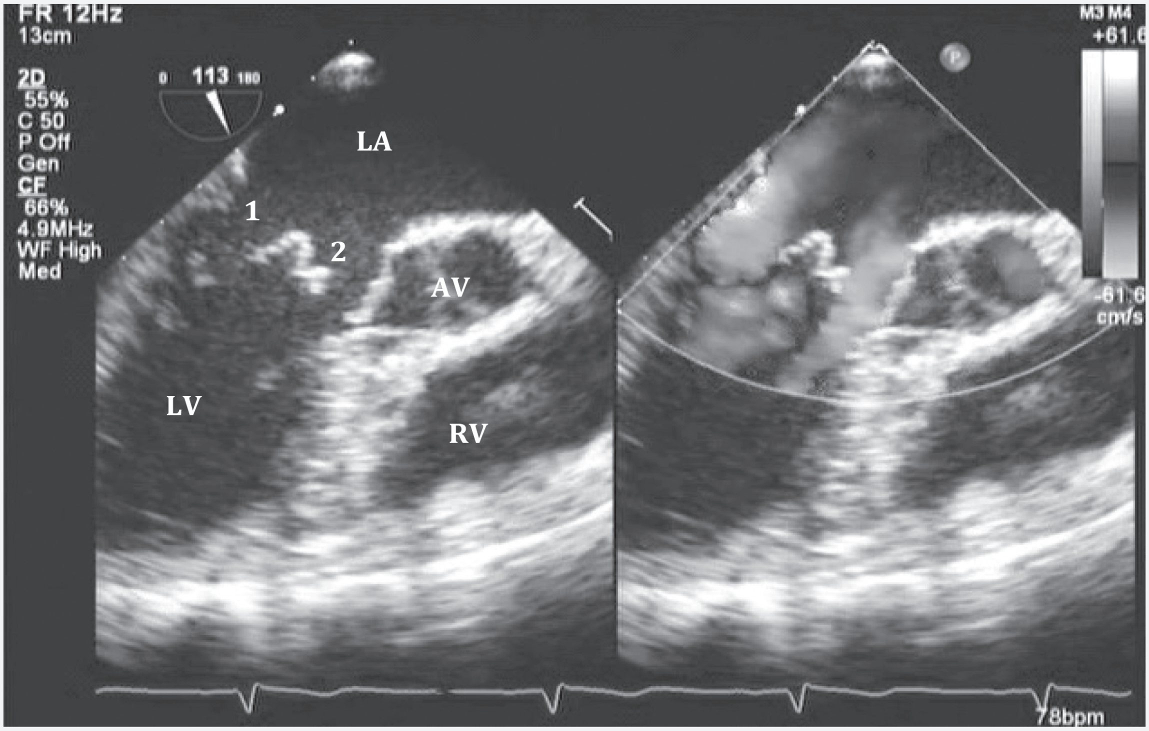

Figure 5. Transesophageal 2D echocardiography midesophageal long-axis view of the mitral valve demonstrating the dual left ventricular inflow pattern across the two orifices (1 and 2) during diastole. Abbreviations: 2D, two-dimensional; AV, aortic valve; LA, left atrium; LV, left ventricle; RV, right ventricle.

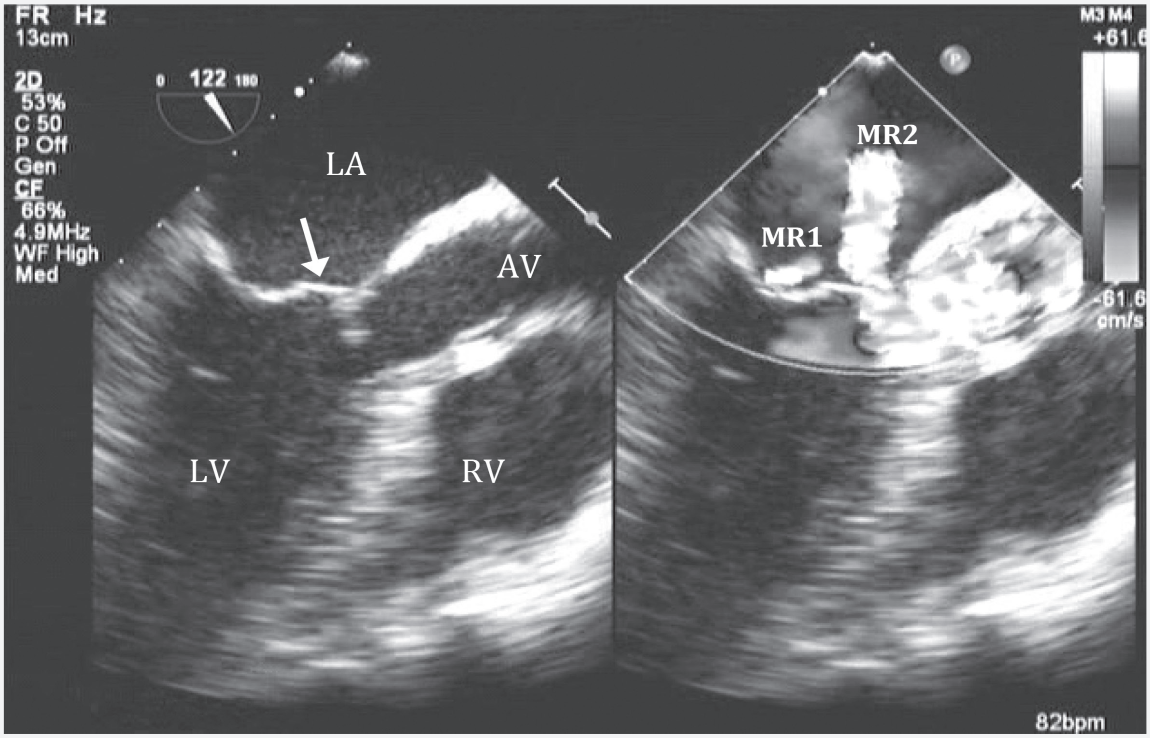

Figure 6. Transesophageal 2D echocardiography midesophageal long-axis view of the mitral valve demonstrating two regurgitant jets (MR1 and MR2) during systole, with predominant mitral regurgitation at the more anterior orifice due to prolapsing leaflets (arrow). Abbreviations: 2D, two-dimensional; AV, aortic valve; LA, left atrium; LV, left ventricle; RV, right ventricle.

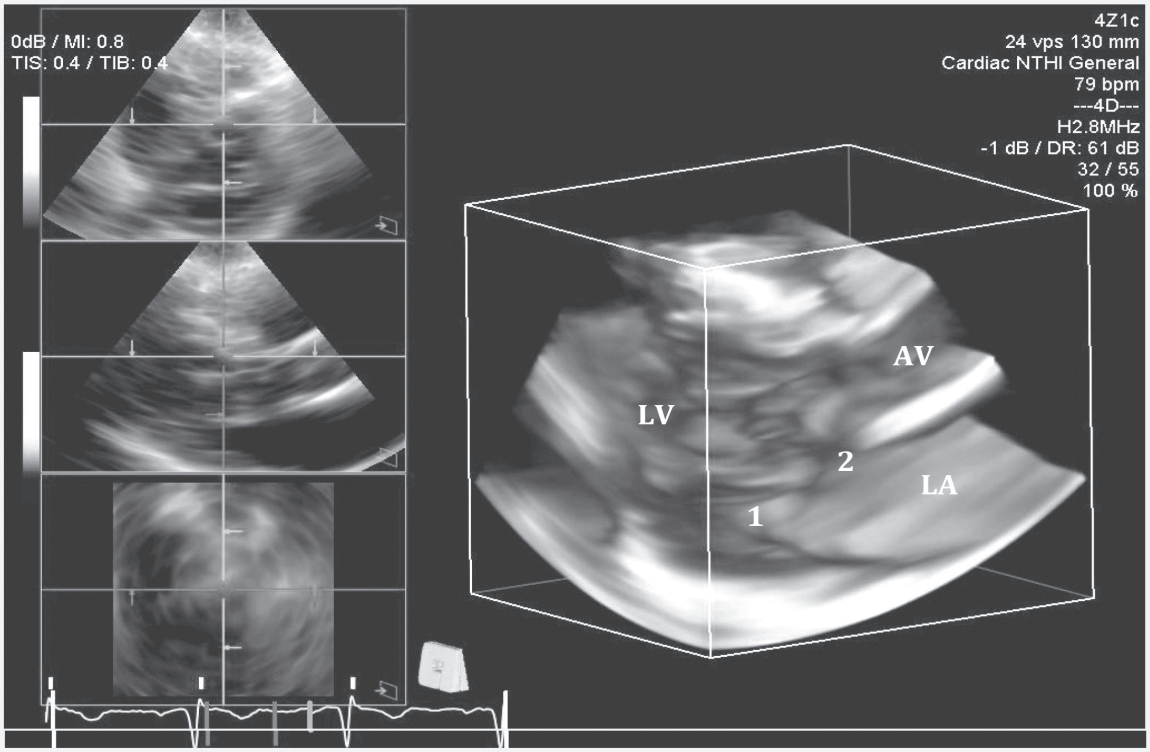

Figure 7. Transthoracic 3D echocardiography simultaneous imaging of mitral valve from multiple planes. Freezing the 3D image during diastole reveals the presence of two distinct mitral valve orifices (1 and 2). Abbreviations: 3D, three-dimensional, AV, aortic valve; LA, left atrium, LV, left ventricle.

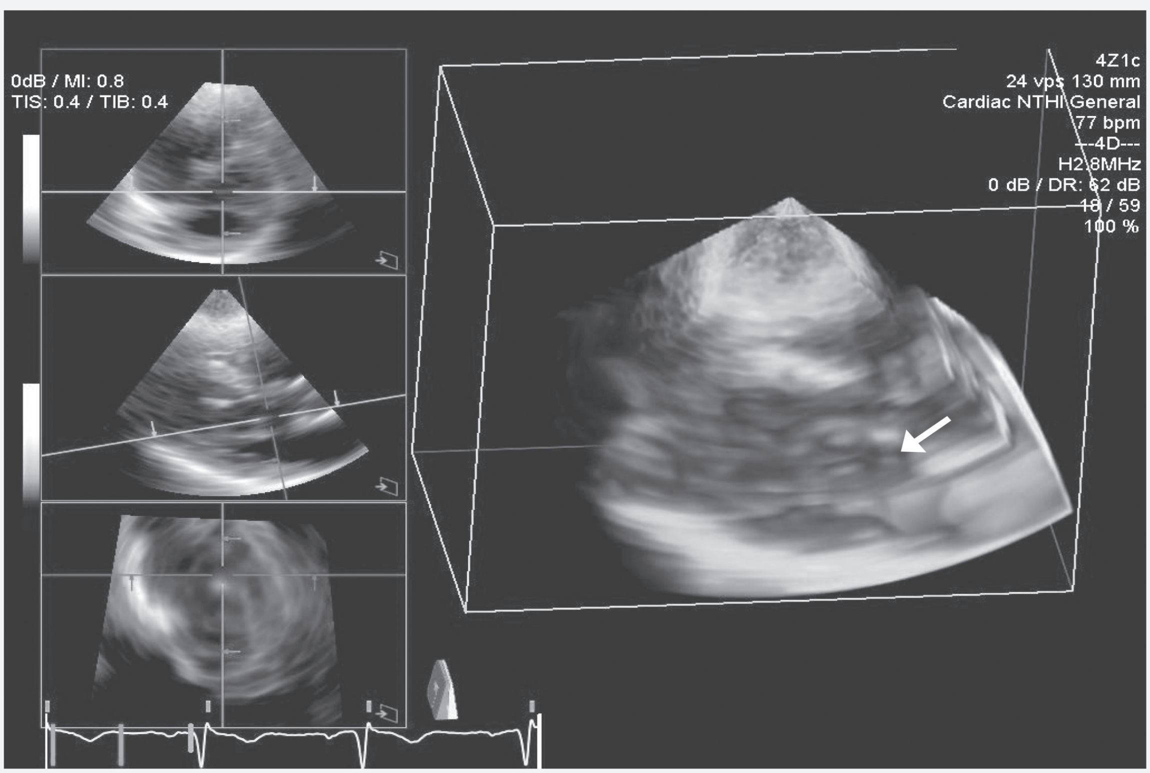

Figure 8. Transthoracic 3D echocardiography en-face view of the anteriorly located mitral valve orifice (1) from an LV outflow tract perspective. This image demonstrates how the said orifice opens directly into the LV outflow tract, as opposed to the more posterior orifice, which opens into the LV cavity. Abbreviations: 3D, three-dimensional; LV, left ventricular.

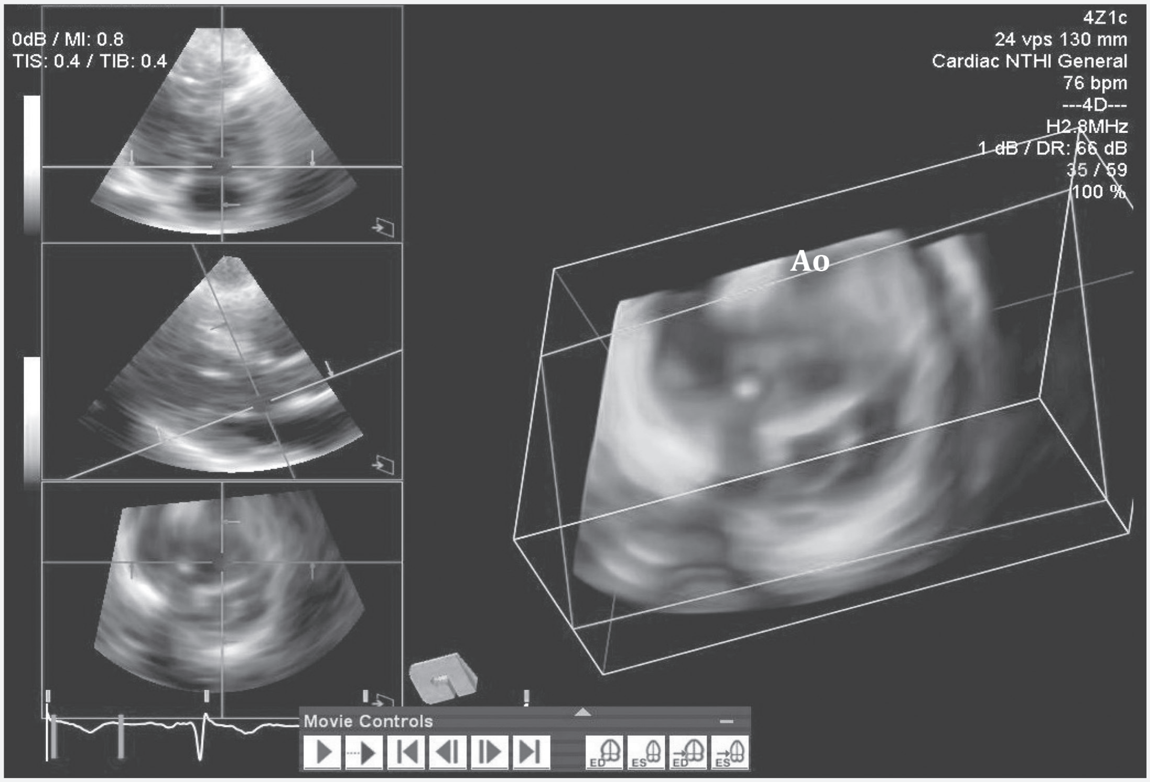

Figure 9. Transthoracic 3D echocardiography en-face view of the two mitral valve orifices from left ventricular perspective showing a “figure-of-8” configuration. This image demonstrates how the multiplanar capability of 3D allows visualization of structures from unconventional viewing planes not possible with 2D. Abbreviations: 2D, two-dimensional; 3D, three-dimensional; Ao, aorta.

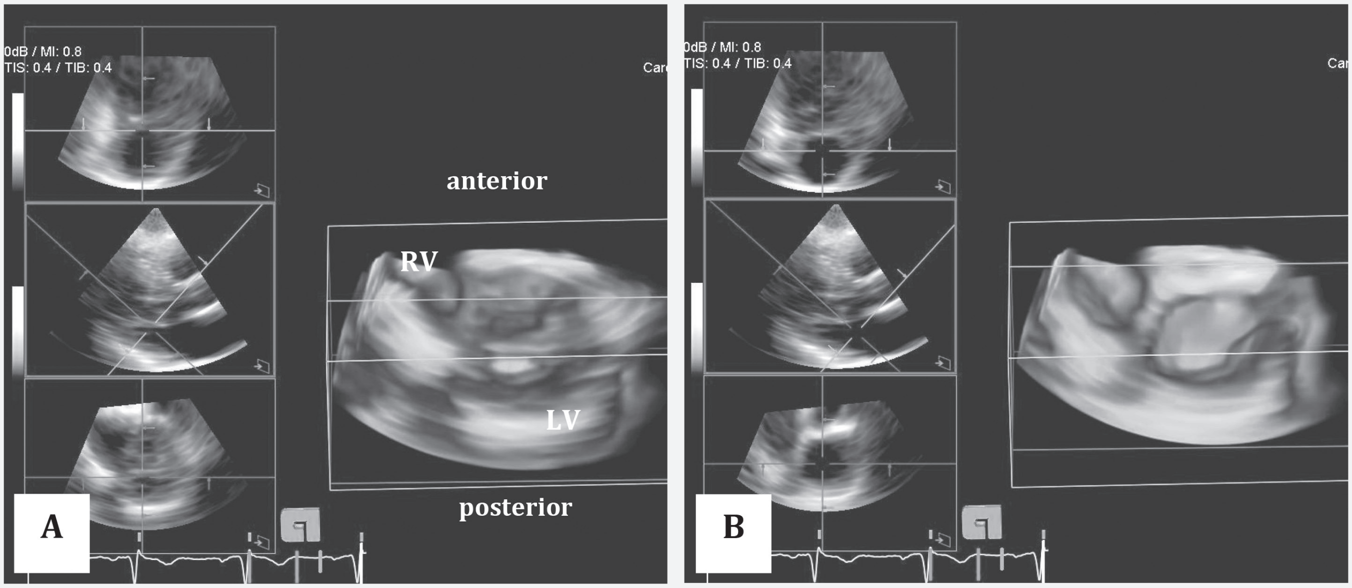

Figure 10. Transthoracic 3D echocardiography en-face view of the mitral valve apparatus from left ventricular perspective demonstrating presence of intervening tissue at the mid- to apical portions (A) and its absence at the basal portion (B) of the mitral valve, suggesting an “incomplete bridge type” of double-orifice mitral valve. Abbreviations: 3D, three-dimensional; LV, left ventricle; RV, right ventricle.

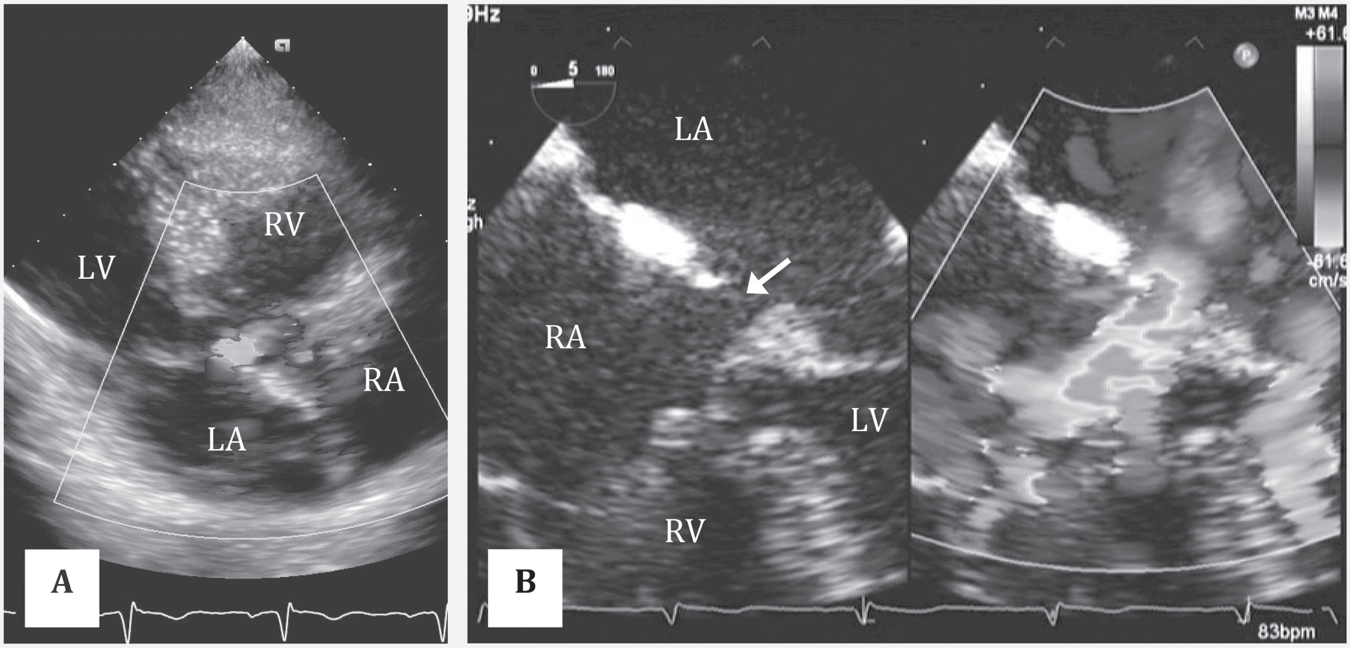

Figure 11. Images of the primum atrial septal defect (arrow) on transthoracic 2D using the apical modified four-chamber view (A) and on transesophageal 2D using the midesophageal four-chamber view (B). Color Doppler clearly demonstrates a left-to-right shunt. Abbreviations: 2D, two-dimensional; LA, left atrium; LV, left ventricle; RA, right atrium; RV, right ventricle.

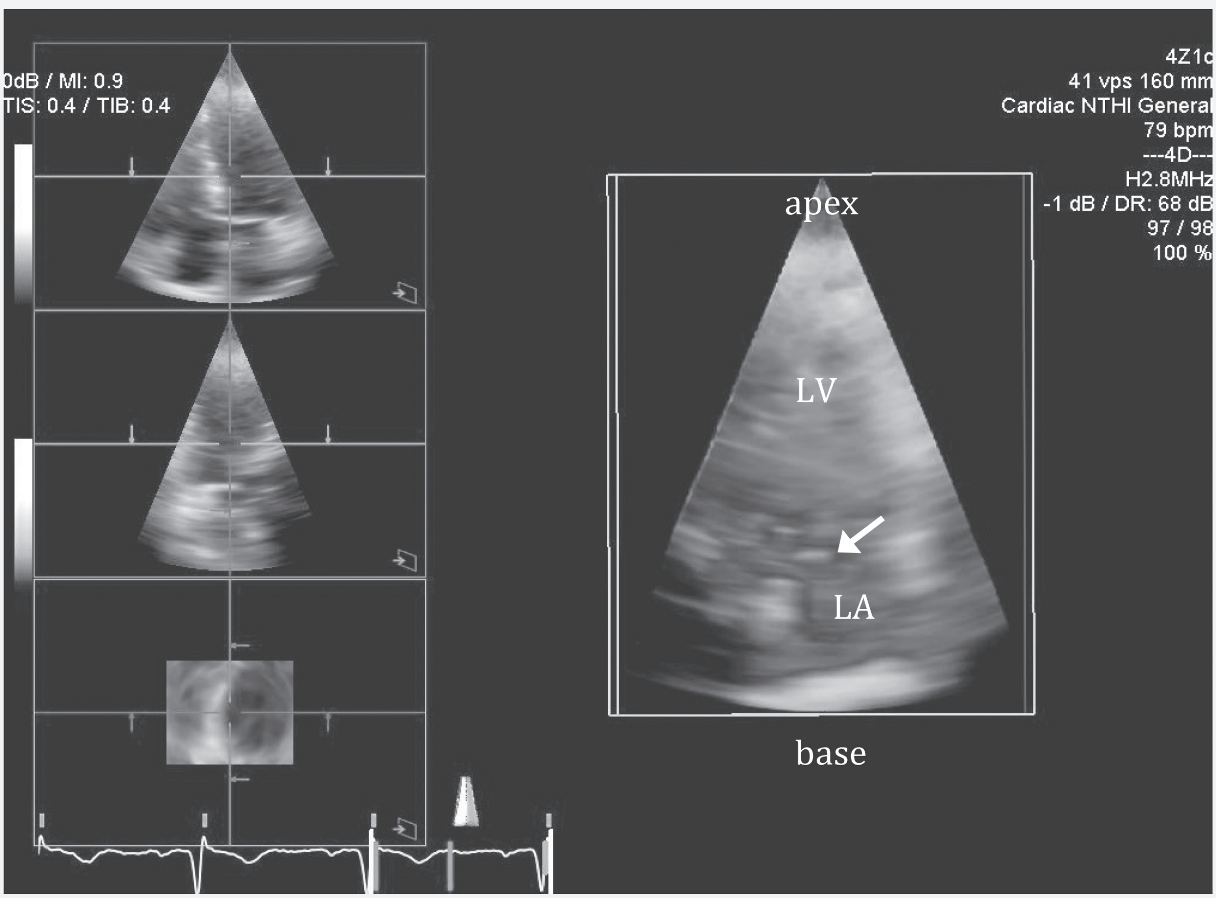

Figure 12. Transthoracic 3D echocardiography showing an en-face view of the primum ASD (arrow) from a left atrial perspective. Abbreviations: 3D, three-dimensional; ASD, atrial septal defect; LA, left atrium; LV, left ventricle.

DISCUSSION

Anomalies in Mitral Valve Anatomy

Several structural anomalies of the mitral valve apparatus have already been described and published, with most of the congenital variants demonstrating either an excess or lack of certain mitral components, such as the leaflets, chordae, or papillary muscles. In the echocardiographic detection of a DOMV, it is necessary to clarify certain terminologies and be able to distinguish this rare anomaly from other masqueraders, such as the trileaflet mitral valve, the duplicate mitral valve, the true parachute mitral valve, and the parachute-like asymmetric mitral valve. The true parachute mitral valve is characterized by a unifocal attachment of mitral valve chordae tendinae independent of the number of papillary muscles. It has a single papillary muscle that receives all chordae, unlike the parachute-like asymmetrical mitral valve, which has two papillary muscles, one being elongated and located higher in the left ventricle.[6] In the case of a duplicate mitral valve, two independent mitral valve apparatuses (leaflets and annulus) and subvalvular apparatuses (chordae and papillary muscles) function well by themselves. Such an anomaly is usually isolated.[7] The trileaflet mitral valve is defined as a left atrioventricular valve with three distinct leaflets and commissures, resulting in the classic "Mercedes Benz" configuration on short-axis view more typical of the AV.[8]

The Double-Orifice Mitral Valve

The DOMV is characterized by the presence of a single fibrous annulus with two distinct orifices in the left atrioventricular valve area, each with its own set of leaflets and chordal attachments.[9] From an embryologic standpoint, DOMV arises as a result abnormal fusion of the endocardial cushions of the primitive AV canal, resulting in two orifices of the mitral valve.[4] This endocardial cushion abnormality in DOMV probably also explains the coexistence of other anomalies such as atrial and ventricular septal defects. The echocardiographic detection of this structural anomaly is important because of its potential physiologic or hemodynamic impact, with mitral regurgitation being the most common functional abnormality arising from valve incompetence.[4,5]

Different variants of DOMV have been recognized, and one classification scheme segregates these variants into three groups: (1) the complete bridge type, wherein both circular orifices are visible from the leaflet edge to the valve ring, with each orifice having separate chordal attachments to structurally normal papillary muscles; (2) the incomplete bridge type, wherein the orifices are distinct only at the level of leaflet edges, creating pseudonormal short-axis views of the mitral valve at more basal levels; and (3) the hole type, wherein a small accessory orifice can be found at the anterolateral or posteromedial commissure of the main orifice, which is seen only at the midleaflet level.[10] The hole type is believed to be the most prevalent variant (80% of patients), commonly associated with a common AV canal.[9] The present case falls within the second category, the incomplete bridge type, which is believed to be the rarest of all DOMVs.[10]

Role of Advanced Cardiac Imaging

Several studies have already demonstrated how 3D reconstruction transoesophageal echocardiography provides better accuracy in patients with complex mitral valve pathology when compared with 2D transoesophageal echocardiography.[11,12] One distinct advantage of 3D echocardiography over its 2D counterpart is its ability to demonstrate mitral valve morphology from virtually any perspective. The nonplanarity of the mitral valve (being inherently saddle-shaped) makes this capability for multiplanar viewing particularly useful.[13] The present case demonstrated how real-time 3D allowed simultaneous imaging of multiple 2D planes. Moreover, the 3D matrix array ultrasound transducer provides flexibility for the following maneuvers: (1) multiplane imaging during the same cardiac cycle or over multiple cardiac cycles, (2) leveraging of 2D image quality, and (3) quantitation, on axis imaging, and rapid acquisition.[14]

The unusual configuration of the mitral valve in the present case possibly resulted in the exaggeration of the saddle-shaped geometry of the mitral annulus. We speculate that, because of the rigid nature of the aortomitral fibrous skeleton, the mitral annulus had to become distorted to accommodate itself within the limited confines of the LV cavity. Such an anatomic distortion has implications from a surgical standpoint. First, the accurate measurement of mitral annular dimensions cannot be made solely on 2D images, but rather by using the multiplanar capability of 3D echocardiography. Second, the risk for arrhythmogenesis can, in theory, be affected by disruption of the aortomitral fibrous geometry. A study of 404 consecutive patients who presented for catheter ablation of idiopathic premature ventricular complex/ventricular tachycardia over a period of 9 years revealed that 5% of the population had an ablation site at the mitral annulus.[15] Based on this observation, it may be logical to surmise that congenital anomalies involving structural deformation of the mitral annulus (such as a DOMV) may, in theory, increase the risk of such arrhythmias. Third, the type of surgical intervention has to be analyzed carefully, weighing the advantages and disadvantages of valve repair as opposed to replacement, with special consideration for the coexisting primum ASD. Fourth, the distortion of the normal spatial relationships of the mitral valve to its chordal and papillary muscle attachments has to be elucidated in the event that repair is to be strongly considered. Finally, despite the seemingly equal contribution of each orifice to LV inflow, the present case revealed a nearly exclusive site for mitral regurgitation, at the more anteriorly situated orifice. This may have important implications in terms of selective repair or total replacement of the mitral valve.

Recommended Management

Patients diagnosed as having DOMV need to be well-evaluated for the hemodynamic consequences of mitral regurgitation. The presence of heart failure signs and symptoms warrants close outpatient follow-up or even hospital admission if there is urgent need for decongestion and stabilization of hemodynamics.

Mechanical intervention for the dysfunctional valve may include either balloon dilatation via transcatheter approach,[17] especially in instances where the bridge is deemed to be incomplete.[18] If such a percutaneous strategy is not feasible, or if the valve is grossly stenotic or malformed, surgical intervention is recommended, either through valve repair[17] if feasible or outright replacement.[19]

CONCLUSION

The rare combination of congenital anomalies in this patient deserves utmost care in terms of noninvasive diagnosis in anticipation of surgical intervention. The inherent advantages of transesophageal 2D as well as transthoracic 3D echocardiography must be recognized and maximized to arrive at the most accurate anatomic and mechanistic description of the cardiac lesions present in the patient. The insights provided by advanced cardiac imaging techniques cannot be undermined, as the latter's configuration can give potentially reliable clues to the true anatomy of the mitral valve and its amenability for repair or replacement.

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