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Double-Orifice Mitral Valve: Diagnosis and Characterization with Three-Dimensional Transesophageal Echocardiography

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Abstract

Double orifice mitral valve is a rare congenital anomaly that is often associated with other congenital cardiac abnormalities. Patients may present with valvular dysfunction or may be asymptomatic and have an incidental diagnosis of double outlet mitral valve. Whether symptoms due to this pathology are present or not, it is important to accurately characterize this lesion. 3-dimensional imaging is a powerful diagnostic modality for diagnosing and characterizing lesions such as this as highlighted in this case image series.

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Abstract

Double orifice mitral valve is a rare congenital anomaly that is often associated with other congenital cardiac abnormalities. Patients may present with valvular dysfunction or may be asymptomatic and have an incidental diagnosis of double outlet mitral valve. Whether symptoms due to this pathology are present or

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not, it is important to accurately characterize this lesion. 3-dimensional imaging is a powerful diagnostic modality for diagnosing and characterizing lesions such as this as highlighted in this case image series.

Case Image

Two patients with known bicuspid aortic valve stenosis presented for surgical aortic valve replacement. During the pre-cardiopulmonary bypass transesophageal echocardiogram (TEE) examination both patients were incidentally found to have a fibrous strand that could be intermittently identified traversing the anterior-posterior axis of the valve**(Figure 1A, Video 1A).** There was no significant regurgitation or stenosis appreciated on the Doppler examination of the valve. 3-dimensional (3D) TEE was subsequently used to characterize these lesions further in both patients. In our first patient 3D-TEE examination of the valve, showed a fibrous strand connecting the anterior and posterior leaflets at the leaflet edge level giving this patient a diagnosis of double orifice mitral valve (DOMV) 'incomplete bridge type **(Figure 1B&C, Video 1 B&C).'** In our second patient on 3D-TEE a fibrous bridge could be clearly seen dividing the atrioventricular orifice completely from the leaflet edge all the way through the valve annulus (Figure 1D) giving this patient a diagnosis of DOMV 'complete bridge type.'

DOMV is a rare congenital cardiac anomaly occurring in 1% of autopsied cases of congenital heart disease and is typically associated with other congenital cardiac pathologies.[1] DOMV is defined as a single fibrous annulus with two orifices opening into the left ventricle and can be classified into three types: the 'incomplete bridge type' the 'complete bridge type' as described for patients 1 & 2 in our case image series and the then the 'hole type' where a secondary orifice with subvalvular apparatus occurs in the lateral commissure of the MV.[2] 3D-TEE has been shown to be a valuable tool for characterizing and classifying congenital cardiac lesions such as DOMV.[3] In our first patient, we see how on 2-dimensional TEE the 'fibrous strand' could be mistaken for mobile vegetation or torn chordae tendineae **(Figure 1A, Video 1A)** , but when viewed with 3D-TEE and with multiplanar reconstruction we can clearly see the fibrous strand connecting the anterior and posterior leaflets at the leaflet edge level (**Figure 1B&C, Video 1B&C**). For our second patient, we see how 3D-TEE clearly demonstrates the fibrous bridge that divides the atrioventricular orifice completely from the leaflet edge all the way through the valve annulus **(Figure 1D)** .

The etiology of DOMV remains unclear with a defect in the endocardial cushion being one postulated cause. This etiology is supported by the fact that DOMV is rarely an isolated pathology as it is often associated with other cardiac congenital anomalies such as coarctation of the aorta, patent ductus or BAV as seen in both of our cases. Treatment of DOMV will depend on whether if it is associated with mitral valve regurgitation or stenosis with surgical repair or replacement being an option. Many patients remain asymptomatic with DOMV being picked up incidentally and no intervention is required as in both of our patients presented in this image series.

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Figure Legends:

Figure 1:

A) 2D-TEE image of the MV in patient - 1 that shows the fibrous strand that could be mistaken for a torn chordae tendineae or mobile vegetation,

B) 3D narrow sector view of the MV in the en-face view showing the fibrous strand connecting the anterior and posterior leaflets at the leaflet edge in patient -1,

C) Multiplanar reconstruction of the MV showing the fibrous strand connecting the anterior and posterior leaflets at the leaflet edge level in patient – 1,

D) 3D-TEE wide sector of the MV in the en-face view showing a fibrous bridge that traverses the anterior to posterior valve annulus and divides the atrioventricular orifice into two in patient – 2.

Abbreviations : 3D-TEE = 3-dimensional transesophageal echocardiography, MV = Mitral valve, 2D-TEE = 2-dimensional transesophageal echocardiography, AV = Aortic valve, TV = Tricuspid valve, PMC = Posteromedial commissure, Red arrow = Fibrous bridge.

Video 1:

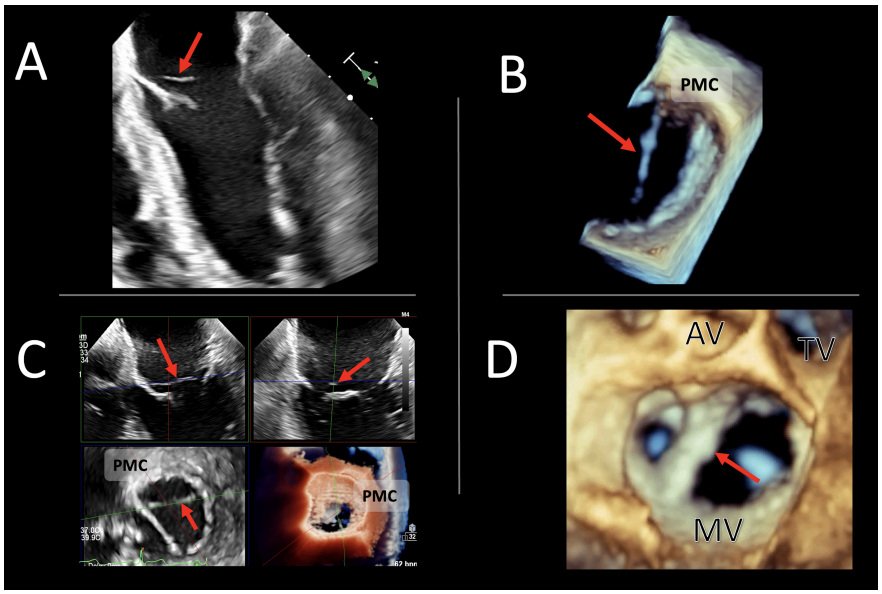
A) 2D-TEE image of the MV in patient - 1 that shows the fibrous strand that could be mistaken for a torn chordae tendineae or mobile vegetation,

B) 3D narrow sector view of the MV in the en-face view showing the fibrous strand connecting the anterior and posterior leaflets at the leaflet edge in patient -1,

C) Multiplanar reconstruction of the MV showing the fibrous strand connecting the anterior and posterior leaflets at the leaflet edge level in patient – 1,

D) 3D-TEE wide sector of the MV in the en-face view showing a fibrous bridge that traverses the anterior to posterior valve annulus and divides the atrioventricular orifice into two in patient – 2.

Abbreviations: 3D-TEE = 3-dimensional transesophageal echocardiography, MV = Mitral valve, 2D-TEE = 2-dimensional transesophageal echocardiography, AV = Aortic valve, TV = Tricuspid valve, LV = Left ventricle, PMC = Posteromedial commissure, Red arrow = Fibrous bridge.



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