A Rare Case of Double Orifice Mitral Valve: A Case Report

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INTRODUCTION

Double-orifice mitral valve (DOMV) is a rare congenital anomaly characterized by a mitral valve (MV) with two orifices that are anatomically separated by an accessory bridge of fibrous tissue. As first described in 1876,[1] the incidence of DOMV has been reported to be 0.05%,[2] although one autopsy study found it to be present in as many as 1% of cases with congenital heart defects.[3] We present an unusual case of DOMV with mitral regurgitation (MR) in a middle-aged man associated with atrial septal defect (ASD), highlighting the role of three-dimensional transesophageal echocardiography (TEE).

CASE HISTORY

The patient was a 50-year-old male presenting with New York Heart Association functional class II dyspnea, a grade 3 systolic murmur best heard over the apex and in normal sinus rhythm. Preoperative transthoracic echo showed an ASD of 8 mm and flow acceleration across the MV. TEE was then done to assess the morphology of the MV and severity of MR.

TEE showed a DOMV with prolapse of the anterior leaflet of anterolateral MV orifice with an eccentric anteriorly directed jet of MR.

The patient was admitted, stabilized, decongested and further evaluated. Routine blood and urine examinations were within normal limits. The carotid and peripheral Doppler were normal and angiography revealed normal coronaries. A detailed cath study was done to assess the operability. Our patient's basal pulmonary vascular resistance (PVR) index was 5.9 WU/m² and post-oxygen therapy for 10 minutes it came down to 2.261 WU/m², thus a decrease of more than 20%, and therefore, the patient was operable.

In the operating room, standard monitoring was done. The TEE revealed a DOMV with two distinct orifices.
Kandpal, et al.: A rare case of double orifice mitral valve

The smaller orifice placed anterolaterally, measuring 1.8 cm showed anterior leaflet prolapse with an eccentric jet of MR; whereas the postero-medially placed larger orifice, measuring 2.56 cm, had normally functioning leaflets with no MR. The gradient across the valve was 2 mmHg. The left atrium was 5.3 cm with no clot in the left atrial appendage or cavity. Other valves were normal. A small ostium secundum ASD of 8 mm with the left-to-right shunt was seen. Left ventricular function was normal [Video 1].

Real-time 3D (RT3D) imaging was done and the en face view of the MV showed valve being divided into two unequal orifices by a complete bridge of tissue and the prolapse of the anterior MV leaflet in the smaller anterolateral orifice [Video 2].

The MV repair was done by mid sternotomy. The anterolateral orifice was repaired with neochord at A2, commissuroplasty and ring annuloplasty with 25 mm Tailor ring. The postero-medial orifice was repaired with a 29-mm Tailor ring annuloplasty as the annulus was dilated and there was only minimal length of coaptation. Leaving it unrepaired would pose a risk of developing regurgitation in the future. The ASD was closed with a pericardial patch.

Weaning off from cardiopulmonary bypass (CPB) was uneventful. Post-CPB TEE confirmed adequate repair of ASD with no flow across the atrial septum and no residual mitral stenosis or regurgitation. Gradients across both the small and large orifices were 2 and 1 mmHg, respectively, thus no mitral stenosis. The pressure half-time revealed adequate valve area of both the orifices, 1.67 and 1.69 cm², respectively [Figure 1 and Video 3].

The post-op 3D en face view of the MV revealed adequately repaired both orifices with the two rings in situ and on color flow there was no residual MR. The postoperative period was uneventful and the patient was discharged on the sixth postoperative day [Video 2].

DISCUSSION

DOMV can be of three types as follow. (A) Eccentric or Hole type, the most common type, which is characterized by a small accessory orifice situated at one of the commissures. It is visible at the mid-leaflet level and disappears when the probe is redirected towards the base or apex. (B) Incomplete bridge type, where the connection is seen between the anterior and posterior leaflets at the leaflet edge, with the annulus and leaflet base unaffected. (C) Complete bridge type as exemplified by our case, where the complete ridge of tissue extends from the anterior to the posterior annulus with the separate orifices visible throughout the valvular ring.[4]

Rarely reported in isolation, DOMV usually presents in early childhood or adolescence due to its association with other congenital heart defects such as coarctation of the aorta, patent ductus arteriosus and atrioventricular septal defects.[4] As evidenced in our patient, who had an associated ASD. However, he was asymptomatic in the early years of his life and presented at 50 years of age.

DOMV is suspected by the finding of a pivot point at the level of the bridging tissue in the midesophageal mitral commissural view. Transgastric basal short-axis or en face views of the MV, obtained with 2D and 3D imaging, respectively, may be used to identify the separate orifices.[9]

Approximately 50% do have significant regurgitation or stenosis that needs to be surgically repaired. It is much more common to have regurgitant lesions rather than stenotic ones with DOMVs. When examining these patients for mitral stenosis, the pulsed-wave Doppler sampling gate must be placed at the tips of the MV leaflets of both orifices. If there is only one orifice with stenosis, the echocardiographer should calculate the total MV area (MVA) by combining the area of both orifices. If this total MVA is <1.0 to 1.5 cm², it necessitates surgical repair. The MVA of both orifices can be calculated by the pressure half-time method or deceleration time and added together. Likewise, it can be calculated by the continuity equation but two combined velocity time integrals (VTIs) of the MV orifices should be in the denominator.[9]

The 3D imaging in multiplanar reconstruction (MPR) improves the accuracy of measurement of the narrowest

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**Figure 1:** Postoperative mean pressure gradients across both mitral valve orifices: (a) Anterolateral orifice and (b) Posteromedial orifice. Post repair valve area by pressure half-time of (c) Posteromedial orifice and (d) Anterolateral orifice.
orifice (albeit at a lower temporal resolution). Highly gained 3D acquisitions can underestimate the orifice area due to the MV appearing falsely smaller than it actually is.\(^\text{[6]}\)

A potential 3D application would be to measure the functional orifices of the valve using 3D MPR with CFD; however, this method has not been validated for MVA post edge-to-edge repair.\(^\text{[7]}\)

Mitral regurgitation should be evaluated as if it is a single-orifice valve. Severe mitral regurgitation through only one orifice would constitute the need for surgical repair. This could be evaluated by measurement of the vena contracta width, regurgitant volume, or effective regurgitant orifice area. As corroborated by our case where the patient was taken for mitral valve repair due to severe MR.\(^\text{[5]}\)

Owing to the en face atrial view of the MV, RT3D echo allows the echocardiographer to visualize the tissue bridge more clearly.\(^\text{[5]}\)

Recognition of the different types of DOMV with the use of echocardiography has been accomplished by carefully examining the different levels of the mitral valve apparatus. However, this process can be significantly improved with the use of a 3D echo. The presence of arrhythmias should not hinder the use of 3D imaging, as long as the frame rate is adequately optimised.\(^\text{[3]}\)

The management strategy depends on the type and severity of MV dysfunction and associated congenital defects. An isolated DOMV with normal hemodynamics needs no active intervention. Division of the bridging tissue results in severe regurgitation that is not amenable to repair. Thus no attempt is made to convert the double orifice into a single one. Standard repair techniques can be applied as needed as in our case where both the orifices were repaired with ring annuloplasty and neo chord at the anterolateral orifice. Valve replacement is considered as the last resort.\(^\text{[8]}\)

**CONCLUSION**

DOMV is a rare congenital cardiac anomaly and we presented a successful mitral valve repair in the case of DOMV. RT3D TEE helped delineate valve morphology and the mechanism of valve dysfunction thereby assisting the surgeon in planning a surgical repair and also helpful to evaluate valve post repair.

**Declaration of patient consent**

The authors certify that they have obtained all appropriate patient consent forms. In the form, the patient(s) has/have given his/her/their consent for his/her/their images and other clinical information to be reported in the journal. The patients understand that their names and initials will not be published and due efforts will be made to conceal their identity, but anonymity cannot be guaranteed.

**Financial support and sponsorship**

Nil.

**Conflicts of interest**

There are no conflicts of interest.

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