

Double Orifice Mitral Valve: An Incidental Finding

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Abstract

A double orifice mitral valve (DOMV) represents a rare congenital malformation characterized by two valve orifices with two separate subvalvular apparatus. We present an interesting case of an asymptomatic 1 year and 8 months old male child with DOMV who successfully underwent surgery.

Keywords: Congenital heart disease, double orifice mitral valve, echocardiography, mitral valve

INTRODUCTION

A double orifice mitral valve (DOMV) is an uncommon anomaly that was first described by Greenfield in 1876. DOMV represents a rare congenital malformation characterized by two valve orifices with two separate subvalvular apparatus.

CASE REPORT

We report a case of an asymptomatic 1 year and 8 months old male child, referred to our department by a pediatric cardiologist. His echocardiography finding suggestive of partial atrioventricular (AV) canal defect with DOMV with the small mitral annulus [Figure 1]. Tricuspid annulus was 14 mm whereas mitral annulus was 9 mm.

During surgery, DOMV anatomy was assessed [Figure 2], the valve area was adequate for his body surface area without any regurgitation. At this moment, recognition of DOMV has no impact on the patient's management. It might, however, be of importance at the time of a further follow-up where he can develop regurgitation. The patient is on regular follow-up without any mitral valve regurgitation or stenosis for 6 months, and he is doing well.

DISCUSSION

A DOMV is an uncommon anomaly that was first described by Greenfield in 1876.^[1] DOMV represents a rare congenital malformation. Characterized by two valve orifices with two

separate subvalvular apparatus.^[2,3] The natural history of DOMV with intact AV septum is not known.

This case reveals the necessity of careful imaging of the mitral valve apparatus, not only in patients with AV septal

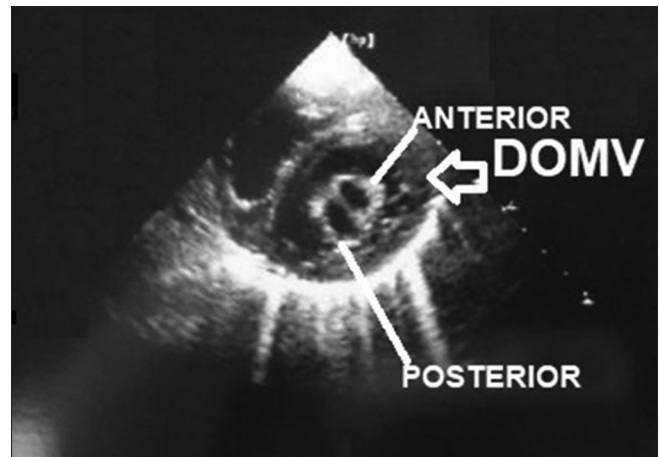


Figure 1: Parasternal short axis view at the level of the mitral valve demonstrating the two separate orifices of the double orifice mitral valve

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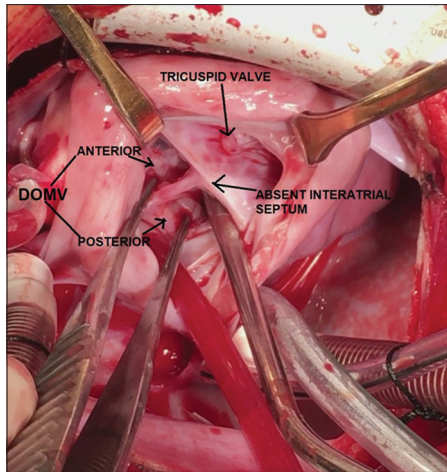


Figure 2: Operative finding

defects but also in patients with congenital left obstructive heart disease.

It is most often associated with AV septal defects (it has been found in as many as 4.9%–17.9% of cases in necropsy studies),^[4] but may also be present together with other congenital heart defects such as left-sided obstructive lesions, ventricular septal defects, or cyanotic lesions. Rarely, patients with isolated DOMV are reported. However, even in the modern era of echocardiography DOMV often remains unrecognized. The hemodynamic impact of DOMV varies from a normally functioning valve to (less frequent) significant mitral regurgitation (with or without cleft) or

stenosis. Surgical correction of DOMV is indicated in a minority of patients undergoing repair of associated lesions. However, these patients need regular follow-up to know any change in morphology and function of the left AV valve in the near future.

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.

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Conflicts of interest

There are no conflicts of interest.

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