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## Double orifice mitral valve associated with Ventricular Septal Defect in an infant: Case Report

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### Abstract

Double orifice mitral valve (DOMV) is an uncommon anomaly characterized by a mitral valve with a single fibrous annulus with two orifices opening into the left ventricle. Here we present a rare case of DOMV with ventricular septal defect (VSD) which was detected by 2-D echocardiography.

**MeSH:** Heart defects, congenital, Double orifice mitral valve, Ventricular septal defect

### Introduction

DOMV is a rare congenital malformation which was first reported by Greenfield in 1876.<sup>1</sup> It is often associated with atrioventricular canal defects, although it can be seen either as an isolated malformation or in association with other cardiac anomalies. It's usually does not lead to any significant hemodynamic effects except regurgitant and rarely stenotic.<sup>2</sup> We present here a rare case of DOMV with VSD which was detected by 2-D echocardiography.

### Case Report

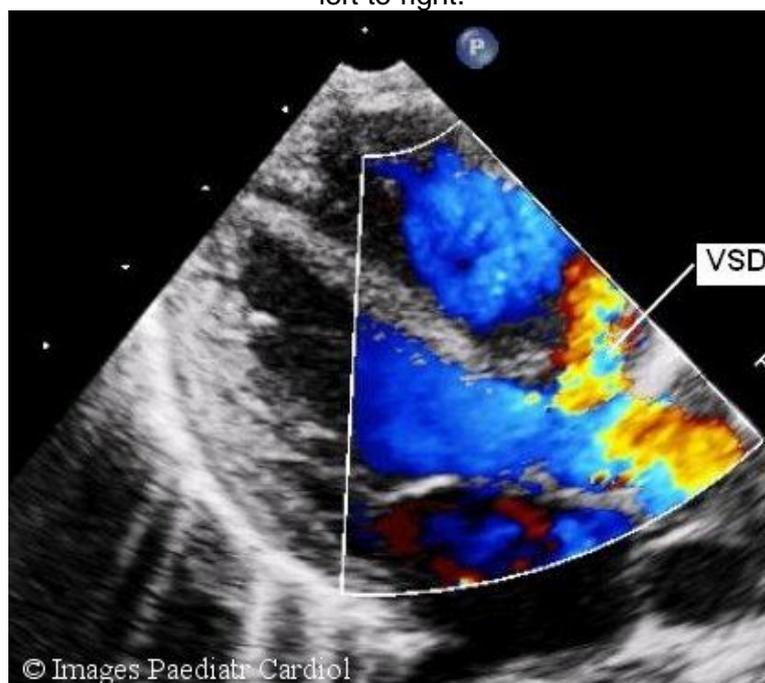
A 2 month old female child, product of non-consanguineous marriage, presented with history of rapid breathing. On physical examination, a pansystolic murmur [Grade 3/6] was heard in 3<sup>rd</sup>, 4<sup>th</sup> and 5<sup>th</sup> intercostal spaces in left parasternal area. On electrocardiography, no abnormality was detected. Chest x-ray showed normal cardiac silhouette and lung vasculature. Transthoracic echocardiography revealed dilated left atrium and left ventricle. Mitral valve was divided into 2 separate valve orifices by a fibrous bridge (Figure 1).

Figure 1 Short axis parasternal view shows double-orifice mitral valve



During diastole in four chamber view, the anterior mitral leaflet appeared like a 'V' due to an abnormal bridge of fibrous tissue connecting the two leaflets. Color Doppler flow imaging of the mitral valve showed 2 separate envelopes of antegrade flow into the left ventricle through the double orifices during diastole. No mitral or aortic regurgitation was documented by color Doppler flow imaging. The left ventricular ejection fraction was within normal limits. There was a small perimembranous VSD with left to right shunt (Figure 2). A small PFO was present shunting from left to right. In view of patient age and the cardiac lesion i.e. DOMV associated with small size VSD without mitral valve obstruction, patient was advised conservative management.

Figure 2 Long axis parasternal view shows the ventricular septal defect shunting from left to right.



## Discussion

The normal mitral valve consists of a central orifice located between a sail-like anterior leaflet and a C-shaped posterior leaflet. The anterior mitral leaflet occupies roughly one-third of the annular circumference and the posterior leaflet occupies roughly the remaining two-thirds of the annular circumference. In a DOMV, however, abnormal tissue divides the mitral orifice into 2 parts.<sup>3</sup> Mitral orifice are unequal in size, with the smaller orifice directed towards the anterolateral commissure (41%) or the posteromedial commissure (44%). The mitral valve can function reasonably well in about 50% of patients with DOMV. In the other 50%, it can cause clinically significant mitral stenosis or mitral regurgitation. The commonly associated lesion is atrioventricular septal defect, VSD, coarctation of aorta, interrupted aortic arch, patent ductus arteriosus, primum atrial septal defect, tetralogy of Fallot, and ebstein anomaly.<sup>4–9</sup> Rosenberg et al reported,<sup>8</sup> 25% of patients with DOMV have partial persistent AV canal and about 5% of patients with partial persistent AV canal have DOMV. Bibhuti Das et al found that various abnormalities along with DOMV were left-sided obstructive lesions, VSD; anomalies of the tricuspid valve.<sup>3</sup> However, in our case, there was only a ventricular septal defect as an associated anomaly.

The classification based on echocardiographic imaging was proposed by Trowitsch et al,<sup>5</sup> which divided DOMV into 3 different types: 1) hole type (accessory orifice surrounded by leaflet tissue that may have a chordal ring), 2) complete bridging (fibrous bridge in the plane of the mitral valve sails, dividing the mitral valve opening into 2 parts that may be equal or unequal), and 3) incomplete bridging (small strand of fibrous tissue connects only the tips of the anterior and posterior leaflets. Our patient falls into the complete bridge type of DOMV as per the Trowitsch classification. Rosenberg et al<sup>7</sup> reported that the true developmental basis of duplication of the mitral valve is unknown. DOMV might be the result of foetal endocarditis or may be a purely developmental anomaly.<sup>8</sup> Another study proposed that the double mitral valve was the result of an early arrest of development and that the accessory orifice represents retention of the left portion of the common AV canal with subsequent reduction of the mitral ostium and alignment with it.<sup>4</sup>

Management depends on the type and severity of mitral valve dysfunction. Isolated DOMV causing neither obstruction nor regurgitation needs no active intervention. Surgical intervention is necessary when stenosis or incompetence is severe or if repair of an associated cardiac lesion is needed. Long-term follow-up is necessary to detect subsequent abnormal hemodynamics or complications. More recently, two dimensional echocardiography has allowed a non-invasive and more frequent detection of this abnormality. In conclusion, this case demonstrates the necessity of careful imaging of the mitral valve apparatus in patients with ventricular septal defects.

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