Double Orifice Mitral valve with Muscular Ventricular Septal defect: A case report

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Citation

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Abstract

Double orifice mitral valve is a rare congenital heart anomaly. We present here a case in which this anomaly was detected during routine echocardiography done for evaluation of a systolic murmur.

INTRODUCTION

The first description of double orifice mitral valve was made by Greenfield in 1876 A.D and about 220 cases have been reported so far [1] . This abnormality may be present as an isolated lesion, but more often, it is associated with other cardiac diseases such as atrioventricular septal defects, bicuspid aortic valve, and coarctation of the aorta. [234]

Usually this anomaly does not lead to any significant hemodynamic effects, sometimes it is regurgitant and rarely stenotic. Hence most descriptions of this anomaly were casual observations at autopsy or during surgery for other associated cardiac defects. More recently, two dimensional echocardiography has allowed a non-invasive and more frequent detection of this abnormality. [456] In this particular case we have performed two dimensional and three dimensional echocardiography for complete elucidation of the findings.

CASE REPORT

A 15 year old boy, product of non-consanguineous marriage, presented with history of breathlessness on exertion NYHA class II for 6 months. There was no history of recurrent lower respiratory tract infections during childhood. On physical examination a pan systolic murmur [Grade III] was heard in 3 rd and 4 th intercostal spaces in left parasternal area.

On ECG no abnormality was detected. Chest x-ray showed normal cardiac silhouette and lung vasculature. Two and three dimensional echocardiography with colour Doppler [Phillips iE 33] showed that one papillary muscle was attached to the septum (Fig 1) and this finding immediately attracted attention. On detailed interrogation from craniocaudal direction, double mitral inflow became obvious. This finding was confirmed in short axis view which showed two adjacent nearly equal sized circular orifices appearing like a spectacle (Fig 2).

Figure 1

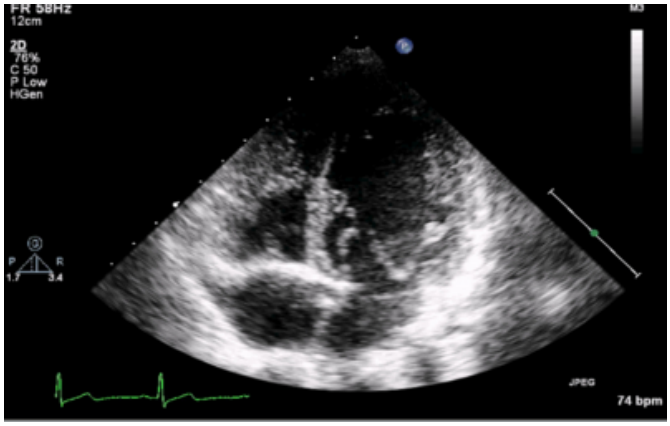
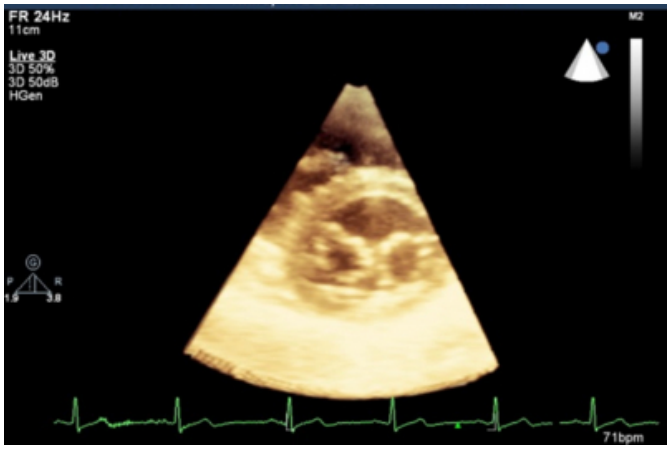
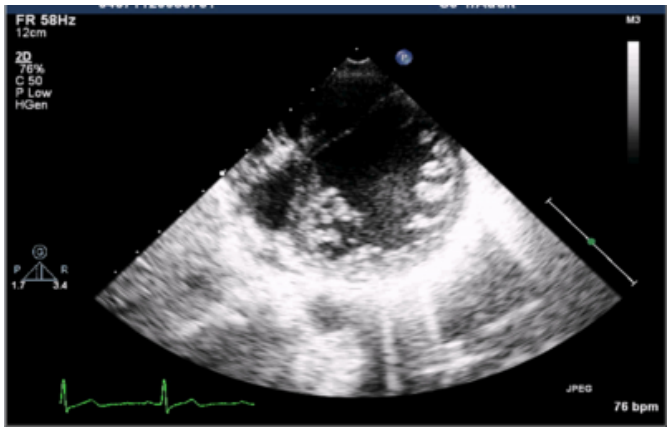


Figure 2



Short-axis view at the level of the papillary muscles demonstrated four papillary muscles (Fig 3).

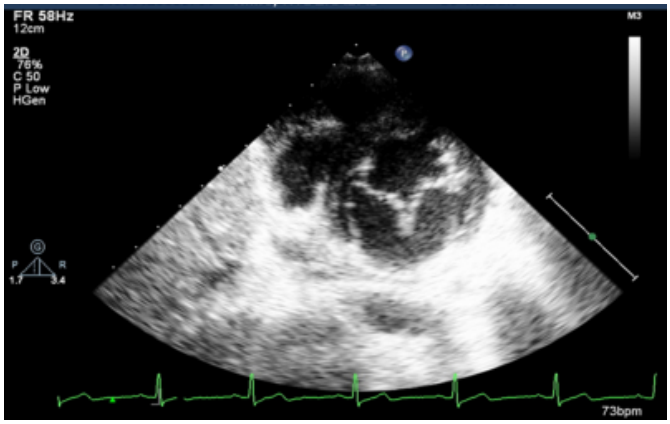
Figure 3



Unlike the rare anomaly ‘duplication of mitral valve’ which has two annuli, this condition has a single mitral annulus.

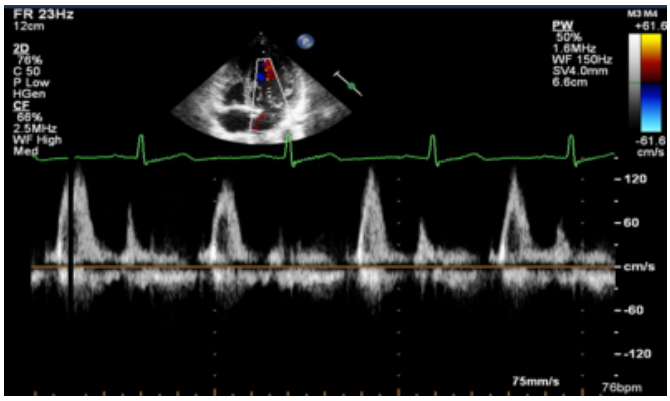
During diastole in four chamber view, the anterior mitral leaflet appeared like a ‘V’ due to an abnormal bridge of fibrous tissue connecting the two leaflets (Fig 4).

Figure 4



The four-chamber view with colour Doppler showed the left ventricular filling through two separate jets without any stenosis (Fig 5).

Figure 5



A trivial mitral insufficiency was seen from the medial orifice. The atrioventricular septum was intact. There was a small restrictive mid muscular VSD with left to right shunt. In view of minimal symptoms and a small VSD coupled with the fact that there is no obstruction at the level of mitral valve, patient was advised conservative management with prophylaxis for infective endocarditis.

EMBRYOLOGY

The embryology of double-orifice mitral valve is incompletely understood and controversial. However, as most of valve leaflets, chordae tendineae, and papillary muscles are the result of invagination of the atrioventricular sulcus and undermining of ventricular myocardium, it is conceivable that an abnormality of these processes can lead to such a defect. [7]

DISCUSSION

Double-orifice mitral valve (DOMV) is a rare malformation

characterized by 2 valve orifices with a separate subvalvular apparatus for each orifice. The presence of tendinous attachments to both openings distinguishes DOMV from acquired lesions, such as perforation or partial fusion of valve leaflets caused by inflammatory lesions, perforated aneurysm of a leaflet, traumatic ruptures, and complications of interventional procedures (eg, balloon valvotomy. [89] It is most commonly reported in association with AV septal defect, where it has been found in

as many as 4.9%-17.9% of cases in necropsy studies. [1011] In the largest published series of 27 postmortem cases of DOMV, 44% had an intact AV septum and 56% had an AV septal defect. [3]

Based on the echocardiographic studies, DOMV has been classified into three types by Trowitzsch et al [5] These are as follows:

1. Complete bridge type: Two separate complete orifices are visible from the leaflet edges all the way through the body of the leaflet. Both the orifices are circular, almost equal in size, and appear like a pair of spectacles.

2. Incomplete bridge type: A connection is seen between the anterior and posterior leaflets only at the leaflet edges. At the mid-basal level, the mitral valve appears normal.

3. Hole type: A single orifice is present at the leaflet level. An additional smaller orifice is visible in one of the two commissures oriented roughly at a right angle to the main orifice.

The two orifices are balanced in size with a central subdivision in only 15% of cases. More commonly, they are unequal in size, with the smaller orifice directed towards the anterolateral commissure (41%) or the posteromedial commissure (44%). In the later case, atrioventricular septal defects are a common associated condition (90%) and mitral regurgitation is often present. The incidence of functional mitral stenosis is more common if the atrioventricular septum is intact.

Our patient falls into the complete bridge type of DOMV as per the Trowitzsch classification.

Bano-Rodrigo et al concluded, based on necropsy data, that DOMV was always associated with an anomaly of the subvalvular apparatus [3] . Bibhuti Das et al agree with these findings and they could demonstrate this association in all their cases by echo [4] . They found various abnormalities: parachute arrangement in 1 case, redundant chordae in 2 cases, chordal ring in 4 cases, and septal attachments of chordae in 5 cases. Papillary muscle anatomy was abnormal in 10 patients. Two patients had 3 papillary muscles, and 2 other patients had fused papillary muscles. In 3 cases the papillary muscles were displaced from their normal position, and in another 3 cases the papillary muscles were unequal in size. The most common associated anomalies in this series were left-sided obstructive lesions, found in 8 of the 18 patients. Ventricular septal defects (VSDs) were seen in 5 patients; anomalies of the tricuspid valve in 2 patients. In 4 patients the DOMV was an isolated finding in an otherwise structurally normal heart. In all of their cases, the accessory (ie, smaller) opening in DOMV with an intact AV septum was located in the anterior leaflet of the mitral valve rather than in the posterior leaflet, as is usual in DOMV associated with AV septal defect.

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References

1. Greenfield WS. Double mitral valve. Trans Pathol Soc London 1876;27:128-9.
2. Sharma S, Loya YS, Daxini BV. Coarctation of aorta with unusual association of diverticulum of the left ventricle and double-orifice mitral valve. Int J Cardiol 1991;30:113-5.
3. Bano-Rodrigo A, Van Praagh S, Trowitzsch E, Van Praagh R. Double-orifice mitral valve: a study of 27 postmortem cases with developmental, diagnostic and surgical considerations. Am J Cardiol 1988;61:152-60.
4. Bibhuti B. Das, Linda B. Pauliks, Ole A. Knudson, Jr, Scott Kirby, Kak-Chen Chan, Lilliam Valdes-Cruz, Raul O. Cavre et al. Double-Orifice Mitral Valve with Intact Atrioventricular Septum: An Echocardiographic Study With Anatomic and Functional Considerations. J Am Soc Echocardiogr 2005;18:231-6.)
5. Trowitzsch E, Bano-Rodrigo A, Burger BM, Colan SD, Sanders SP. Two-dimensional echocardiographic findings in double-orifice mitral valve. J Am Coll Cardiol 1985;6:383-7.
6. Hoffman P, Stumper O, Groundstroem K, Sutherland G. The transesophageal echocardiographic features of double-orifice left atrioventricular valve. J Am Soc Echocardiogr 1993;6:94-100.
7. Wenink ACG, Gittenberger-de Groot AC. Left and right ventricular trabecular patterns. Consequence of ventricular septation and valve development. Br Heart J 1982;48:462-8.
8. Linka AZ, Fatio R, Jost CA. Images in cardiology: double orifice mitral
valve. Heart 2000; 84: 244
9. O'Shea JP, Abascal VM, Wilkings GT, Marshall JE, Brandi S, Acquatella H, et al. Unusual sequelae after percutaneous mitral valvuloplasty: a doppler echocardiographic study. J Am Coll Cardiol 1992;19:186-91.
10. Wakai CS, Edwards JE. Pathologic study of persistent common
atrioventricular canal. Am Heart J 1958;56:779-94.

11. Sherman FE. An atlas of congenital heart disease. Philadelphia: Lea & Febiger; 1963. p. 123, 146, 152-4.

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