Cardiology in the Young

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Double-orifice mitral valve causing left ventricular outflow tract obstruction: a hitherto unreported entity

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Case Report

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Abstract

We report the first documented case of isolated double-orifice mitral valve causing significant left ventricular outflow tract obstruction in an infant. Echocardiography revealed a complete bridge-type double-orifice mitral valve with aberrant chordal insertion into the interventricular septum, producing severe left ventricular outflow tract obstruction. This rare presentation underscores the importance of meticulous echocardiographic assessment for accurate diagnosis and surgical planning.

Introduction

Double-orifice mitral valve is a rare congenital malformation characterised by the presence of two distinct openings in the mitral valve, separated by a fibrous bridge.[1,2] First described by Greenfield in 1876, double-orifice mitral valve accounts for approximately 1% of CHDs.[2] It typically presents as an incidental finding during evaluation for associated anomalies, such as atrioventricular septal defects, ventricular septal defects, subaortic stenosis, coarctation, bicuspid aortic valve, or Shone's complex.[3,4] Most double-orifice mitral valves are haemodynamically insignificant; when symptomatic, they usually cause mitral regurgitation or mitral stenosis.[1,5]

While double-orifice mitral valve has been reported alongside left-sided obstructive lesions, [5] a direct causation of left ventricular outflow tract obstruction solely by a double-orifice mitral valve is hitherto unreported. We report a unique case where isolated double-orifice mitral valve was responsible for significant left ventricular outflow tract obstruction early in infancy.

Case report

An 11-month-old asymptomatic female infant presented to us with a diagnosis of left ventricular outflow tract obstruction which was detected incidentally on day 3 of life during evaluation of a murmur. Clinical examination revealed mild cardiomegaly, normal heart sounds, and a grade 3/6 harsh ejection systolic murmur at the left sternal border. Chest X-ray showed mild cardiomegaly and features of mild pulmonary venous hypertension. Electrocardiogram (ECG) showed left ventricular hypertrophy.

Transthoracic echocardiography revealed a double-orifice mitral valve with two distinct openings, separated by a fibrous bridge. Both the mitral orifices showed laminar flows with no gradient (Figure 1, Video 1). There was mild mitral regurgitation from both the orifices. The smaller, anterior orifice was directed towards the left ventricular outflow tract, and the chordopapillary apparatus of this orifice was partially inserted across the left ventricular outflow tract onto the interventricular septum. There was significant flow acceleration at the point of insertion of the chordopapillary apparatus into the left ventricular outflow tract (Video 1). Continuous wave doppler revealed a peak and mean gradient of 109 and 60 mm of Hg indicating severe left ventricular outflow tract obstruction (Figure 2). Concentric left ventricular hypertrophy was noted, and the left ventricular systolic function was normal with an ejection fraction of 65%. There were no other intracardiac defects, and aortic arch was normal.

A diagnosis of isolated double-orifice mitral valve causing left ventricular outflow tract obstruction was made, and parents were counselled for an elective surgical repair. However, the patient was subsequently lost to follow-up.

Discussion

Double-orifice mitral valve is a structural anomaly often associated with complex CHDs. Isolated double-orifice mitral valve is relatively rare.[1,2,5] Associated anomalies such as atrioventricular septal defects, ventricular septal defects, subaortic stenosis, coarctation, and Shone's complex have been well documented. While double-orifice mitral valve has been

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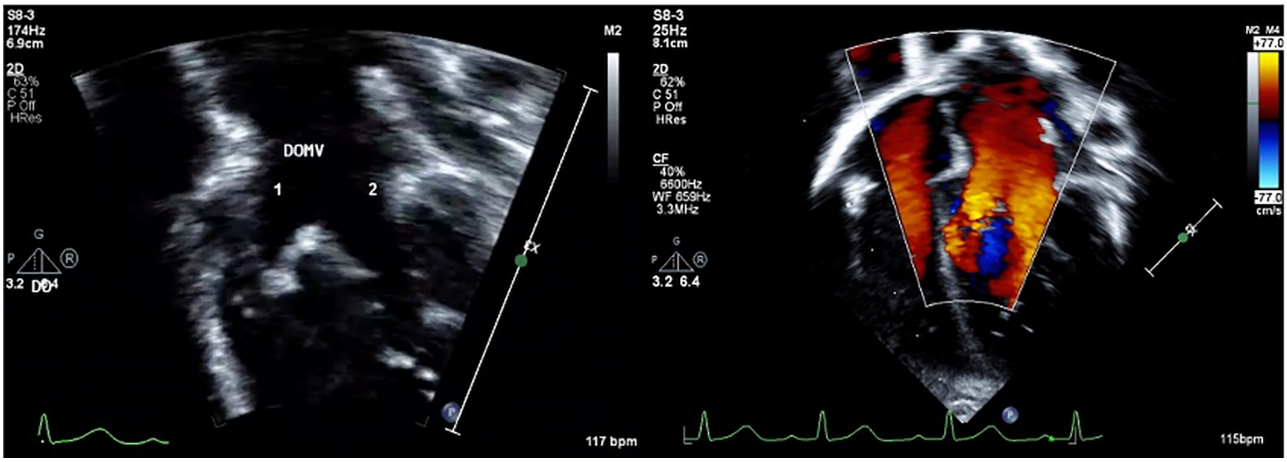


Figure 1. 2D image and colour interrogation of the mitral valve in apical four-chamber view showing double-orifice of the mitral valve with laminar inflows.

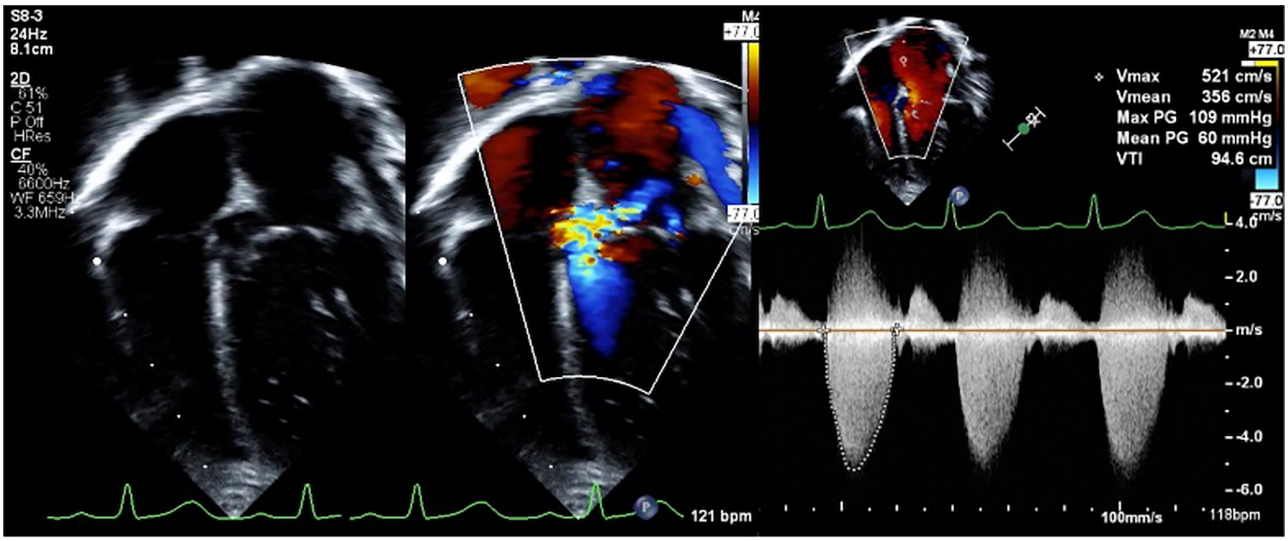


Figure 2. Apical four-chamber view showing severe subvalvar left ventricular outflow tract obstruction caused by chordal insertions of the smaller mitral orifice.

previously described with left-sided obstructive lesions,[5] the obstruction typically arose from discrete subaortic membranes or muscular narrowing rather than from the mitral valve complex itself.[3]

The embryologic basis of double-orifice mitral valve involves abnormal fusion of the endocardial cushions, resulting in duplication of the mitral valve orifice.[6] Anomalies in the tensor apparatus, including aberrant chordal insertion and accessory papillary muscles play a central role in the pathophysiology.[1] Trowitzsch E et al. classified double-orifice mitral valve morphologically as complete bridge type in which a fibrous bridge divided the atrioventricular orifice completely into equal or unequal parts, incomplete bridge type in which a small strand of tissue connected the anterior and posterior leaflets at the leaflet edge level and hole type in which an additional orifice with subvalvular apparatus occurred in the posterior commissure of the mitral valve. Zalzstein et al. classified double-orifice mitral valve into groups based on functional impact: normal, insufficiency, stenosis, or combined lesions.[5]

In the largest reported cases series of double-orifice mitral valve published by Zalzstein et al, they found that of the 46 children aged 2 days to 16 years, obstructive left-sided lesions were present in 19 patients in the form of aortic coarctation (12 patients), subaortic stenosis (6 patients), and Shone's complex in 1 patient.

In the present case, we report a complete bridge-type double-orifice mitral valve where the insertion of chordopapillary apparatus of the smaller orifice onto the IVS likely altered the outflow geometry, creating a fixed component of left ventricular outflow tract obstruction. Double-orifice mitral valve as the aetiology of left ventricular outflow tract obstruction has not yet been reported in literature.

This case uniquely highlights double-orifice mitral valve as the sole cause of left ventricular outflow tract obstruction, emphasising the variability and clinical significance of the subvalvular anatomy in such patients.

Modern echocardiographic techniques, especially three-dimensional imaging, improve the visualisation of complex mitral anatomy in greater detail.[6] However, we could not obtain optimal three-dimensional images in this young infant due to the lack of a dedicated paediatric 3D echocardiography probe. Recognition of double-orifice mitral valve as the aetiological basis of left ventricular outflow tract obstruction is critical, as it often involves

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complex mitral valve repair or mitral valve replacement which requires precise planning based on the age and clinical status of the patient.

Conclusion

We describe an extremely rare presentation of isolated double-orifice mitral valve causing significant left ventricular outflow tract obstruction in early infancy. This adds on to the various functional abnormalities described in association with double-orifice mitral valve. Awareness of this association can facilitate precise diagnosis and appropriate surgical repair involving the mitral valve, thereby improving patient outcomes.

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Competing interests. Nil.

Ethical standards. The authors certify that they have obtained all appropriate patient consent forms. The patient's guardians have consented to the reporting of clinical information with the understanding that anonymity will be maintained.

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