Double orifice mitral valve; a coincidental finding

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Received 29 July 2005; accepted 30 August 2005
Available online 19 January 2006

Abstract A double orifice mitral valve (DOMV) represents a rare congenital malformation characterised by two valve orifices with two separate subvalvular apparatus. This case demonstrates the necessity of careful imaging of the mitral valve apparatus, not only in patients with atrioventricular septal defects, but also in patients with congenital left obstructive heart disease.

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Case description

A double orifice mitral valve (DOMV) represents a rare congenital malformation characterised by two valve orifices with two separate subvalvular apparatus (Figs. 1–3).1,2 It is most often associated with atrioventricular septal defects, 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 haemodynamic 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.

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Figure 1 Parasternal short axis view at the level of the mitral valve demonstrating the two separate orifices of the double orifice mitral valve.
The natural history of DOMV with intact atrioventricular septum is not known. We report a case of an asymptomatic 24-year-old man, referred to our department by his paediatric cardiologist. At the age of 1 he underwent correction for an aortic coarctation. Since then he has been reviewed regularly in our outpatient clinic for a bicuspid aortic valve with moderate to severe aortic stenosis and a moderately dilated ascending aorta, which will require future surgery. Routine echocardiography showed a previously non-recognized DOMV, with slight mitral regurgitation through both orifices. 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 operation.

This case demonstrates the necessity of careful imaging of the mitral valve apparatus, not only in patients with atrioventricular septal defects, but also in patients with congenital left obstructive heart disease.

References


Figure 2 Apical four chamber view demonstrating the two separate subvalvular apparatus of the double orifice mitral valve.

Figure 3 Apical four chamber view with color flow Doppler demonstrating two small regurgitant jets through the two orifices of the double orifice mitral valve.