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Double orifice mitral valve associated with ventricular septal defect, patent ductus arteriosus and aortic coarctation

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Introduction: Double orifice mitral valve (DOMV), described by Greenfield in the 19th century is an uncommon anomaly. It is characterized by a mitral valve with a single fibrous annulus with two orifices that open into the left ventricle. Sub-valvular structures, like the tensor apparatus, invariably show various degrees of abnormality. Although it may allow normal blood flow between the left atrium and ventricle, it can substantially obstruct mitral valve inflow or produce mitral valve incompetence. It is generally associated with AV septal defects or left sided cardiac lesions. We report a symptomatic infant of DOMV with multiple cardiac lesions.

Case Report: Seven months old infant was referred in view of incidental cardiac murmur. On examination the child was found to be underweight, had tachypnea and tachycardia. SpO2: 94% (upper limb); 97% (lower limbs), BP of 85/55 mm Hg (upper limb); 92/60 mm Hg (lower limbs). The precordium was normal with the apex in the 5th intercostal space, at the midclavicular line. On auscultation, there was a mid diastolic rumble at the apex and a systolic murmur present at the lower left sternal border. ECG showed right axis deviation with right atrial enlargement. Chest radiograph revealed mild cardiomegaly with plethoric lung parenchyma. A 2D echocardiographic evaluation with Doppler was done which showed two mitral valve orifices with two sets of papillary muscles (double orifice mitral valve) with mild mitral stenosis, no mitral regurgitation, severe degree of aortic coarctation distal to the origin of left subclavian artery, restrictive perimembranous VSD, a small PDA, mildly dilated left heart without PAH. Repair of the aortic arch coarctation and closure of the PDA was done.

Discussion: Literature reveals four types of DOMV morphology: (1) eccentric or hole type: consisting of a small accessory orifice located in normal leaflets, at the posteromedial or anterolateral commissure; (2) complete bridge type: a central bridge of fibrous tissue connects the two leaflets of the mitral valve, creating two equal/ unequal openings; (3) incomplete bridge type: a small strand of tissue connects the anterior and posterior leaflets at the edge and (4) duplicate mitral valve type, with two separate mitral valve annuli, each with its own set of leaflets and sub-valvular apparatus. A few case reports of isolated asymptomatic DOMV are available, but mostly they are associated with other congenital anomalies. The combination of congenital anomalies with DORV in our case is unique in literature till date. Hence, thoroughly assessing all cardiac structures, ensured not only a comprehensive assessment of the primary indication for the scan, but also improved the detection of concomitant and otherwise unknown lesions.

Biography

Murtaza Kamal is currently working in the Department of Pediatric Cardiology at Star Hospitals, Hyderabad, India. His research interests are ventricular septal defect, pediatric cardiology, etc.

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