Editorial

Double orifice mitral valve: rare disease with a wide range of presentation

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Double orifice mitral valve (DOMV) is a rare congenital anomaly, as implied by the name, that results in two distinct orifices within the mitral valve, rather than usual single orifice. Due to its rarity, there is no established guidelines on evaluation of DOMV.

Theerasuwipakorn *et al.* (1) have highlighted two distinct cases of DOMV and their different outcomes. This condition is rare with an incidence of approximately 1 in 100,000. The exact embryogenesis of DOMV is unknown, but it is thought to be due to abnormal development of the endocardial cushions during embryonic life (2). Baño-Rodrigo *et al.* reported that an anomaly of tensor apparatus was always found in 27 post-mortem cases that included chordal ring, accessory papillary muscle, subdividing muscle ridge, fused papillary muscle, crossing chordae tendineae and central fibrous division (2).

The pathophysiology of DOMV is complex, and it may involve a combination of hemodynamic abnormalities, such as regurgitation, stenosis, and turbulence, as well as structural abnormalities, such as prolapse, redundancy, and fibrosis, of the valve leaflets and the chordae tendineae. It is commonly associated with other congenital anomalies, most commonly atrioventricular septal defects, patent ductus arteriosus or coarctation of aorta (3,4). Thus, a thorough evaluation is needed if DOMV is detected.

The diagnosis of DOMV can be challenging, as it may be mistaken for other conditions, such as mitral stenosis, mitral regurgitation, or mitral valve prolapse. Echocardiography is the gold standard for the diagnosis of DOMV, and it can provide detailed information on the anatomy, function, and severity of the valve abnormalities (5). In the current era of advanced imaging, we believe that multimodality imaging approach is essential and can be help in achieving accurate diagnosis and, more importantly, it allows for better assessment of its hemodynamic consequences and in evaluating concomitant congenital anomalies. 3D transthoracic echocardiogram (TTE) provides a comprehensive assessment of the morphology of DOMV, over and above the information obtained by 2D imaging (6,7). Cardiac magnetic resonance imaging (MRI) can also help assess DOMV especially in the setting of poor echocardiographic windows, and in addition it can better assess potential other congenital anomalies (8). The entity is not well established with cardiac MRI; however, once we use it to assist in diagnosis, it will gain traction to be used as a diagnostic tool in the evaluation of DOMV.

The management of DOMV depends on the severity of the valve dysfunction, the presence of associated congenital heart defects, and the clinical symptoms of the patient. Asymptomatic patients with mild or no valve dysfunction

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may be managed conservatively with regular follow-up and surveillance (9), while symptomatic patients with severe valve dysfunction or associated heart defects may require surgical intervention. The surgical options for DOMV include mitral valve repair or replacement, with the choice of procedure depending on the extent of the valve abnormalities, the age and comorbidities of the patient, and the surgeon's experience and preference.

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