Double-orifice mitral valve: a rare congenital anomaly

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A 39-year old male presented with dyspnoea, NYHA class II. The diagnosis of an ostium primum atrial septal defect (ASD) with a double-orifice mitral valve was established by transthoracic echocardiography (TTE) (Fig. 1A) as well as two-dimensional (2D) and 3D transoesophageal echocardiography (TEE) (Figs 1B and 2A). Intraoperative findings are revealed in Fig. 2B. Successful closure of the primum ASD was performed.

Figure 1: Two-dimensional TTE and TEE. (A) An apical two-chamber view showing the double-orifice mitral valve. LA: left atrium; LV: left ventricle. (B) Two-dimensional TEE showing the double-orifice mitral valve (arrows). LA: left atrium; LV: left ventricle; TEE: transoesophageal echocardiography.

Figure 2: Three-dimensional TEE and intraoperative picture. (A) A real-time 3D TEE atria en face view (surgical view) showing the double-orifice mitral valve with two equal-sized orifices and an abnormal bridging tissue in between. ANT: anterior leaflet; POST: posterior leaflet; MED: medial orifice; LAT: lateral orifice; TEE: transoesophageal echocardiography. (B) Intraoperative image showing the ostium primum ASD and double-orifice mitral valve with equal-sized openings and a bridging tissue in between with separate chordal attachments for each orifice. The chordae from the bridging tissue is attached to the anterolateral and posteromedial papillary muscle. The mitral valve was non-stenotic and trivial regurgitation was present, which did not require any intervention. Closure of the ASD was performed using McGoon’s technique using autologous untreated pericardial patch keeping coronary sinus on the right atrial side.

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