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Double-orifice mitral valve (DOMV) is a very rare congenital malformation characterized by the presence of two anatomically independent orifices in the left atrioventricular (AV) valve area. The DOMV results from the abnormal fusion of the endocardial cushions with the persistence of the left part of the common AV canal. Usually, DOMV is associated with other congenital malformations (such as aortic coartation and bicuspid aortic valve) and subvalvular apparatus abnormalities.

In more than 50% of the cases, valvular function is normal. Otherwise, valve dysfunction may be present (usually mitral regurgitation and rarely mitral stenosis).

We present the case of a 17-year-old female with a past medical history of uneventful surgical correction of aortic coartation presenting with a DOMV determining severe mitral stenosis (Panel A—mean gradient was 16 mmHg on each valve orifice; total valve area was 0.8 cm²) and severe pulmonary artery hypertension (70 mmHg at rest). Pre-operative blood pressure was 110/80 mmHg on both the left and right arms. No clinical or instrumental signs of recoartation were present. The patient was symptomatic for recently worsened exertion dyspnoea (NYHA functional class III).

Pre-operative bi- and three-dimensional echocardiography demonstrated two separated diastolic left ventricular inflow jets (Panel B) with two symmetrical mitral orifices (1 and 2—Panel C). The patient underwent uneventful mitral replacement. Intraoperative aspect was evocative of a duplicated mitral valve, with two small orifices separated by a central fibrous bridge (1 and 2—Panel D). Post-operative echocardiography showed an acute reduction in the pulmonary artery pressure (45 mmHg).

LV, left ventricle; LA, left atrium; AV, aortic valve; TV, tricuspid valve.