Double orifice mitral valve with normal function: an echocardiography and MRI study of a rare finding

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Double orifice mitral valve (DOMV) is a rare congenital lesion often associated with other abnormalities such as bicuspid aortic valve or aortic coarctation. Also abnormalities of the sub-valvular apparatus such as malformation of chordae tendineae (abnormal attachment, parachute type, etc.) and papillary muscles are found frequently. The atrio-ventricular connection consists of two anatomically distinct orifices separated by accessory fibrous tissue. In about 50% of DOMV cases, valvular function is normal, others present with stenosis or regurgitation. In most cases (~85%), a larger orifice is accompanied by a small eccentric accessory orifice, 15% (see our case) show duplicated mitral valves (MVs). Embryologically, the lesion results from abnormal leaflet fusion and persistence of the left part of the common atrio-ventricular canal.

We present a 59-year-old male patient with DOMV and coexisting stenotic bicuspid aortic valve, a low gradient aortic coarctation, and normal MV function. Parasternal 2D cross-sectional echocardiography (Panel A) demonstrates two MV orifices (1./2.), a modified apical four-chamber view shows four leaflets and two separated diastolic left ventricular inflow jets (Panel B). Three-dimensional echocardiography (Panel C) and MRI imaging in the two-chamber view (Panel D, Supplementary material online, Video S1) confirmed the central fibrous bridge dividing the atrio-ventricular orifice. The mild aortic coarctation is demonstrated in Supplementary material online, Video S2.

In cases of severely dysfunctional DOMV with stenosis or regurgitation, surgical intervention is recommended. In cases with normal MV function, surgical therapy is eventually necessary to repair the associated cardiac lesion. In our patient, aortic valve replacement and closure of a coexisting ASD were performed uneventfully.

RV, right ventricle; LV, left ventricle; LA, left atrium; Ao desc, aorta descendens.

Supplementary material
Supplementary material is available at European Heart Journal online.

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