Double orifice mitral valve with dysplastic tricuspid valve and intact interatrial septum: a three-dimensional echocardiographic study

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A 26-year-old male patient with a known diagnosis of congenital heart disease was referred to routine cardiological assessment. The patient was asymptomatic and no details were available about his cardiac condition.

In parasternal long axis view, echocardiography showed what seemed to be a rheumatic valve with mild mitral stenosis and no regurgitation. Parasternal short axis view, however, revealed a double orifice mitral valve (DOMV) with the larger orifice situated laterally (Figure 1). Tricuspid valve was dysplastic with tethered posterior leaflet (Figure 2), borderline apical displacement of the septal tricuspid leaflet of 1 cm, and severe tricuspid regurgitation. There was no other cardiac abnormality and a contrast study was negative for shunt. Real-time 3D scanning (Vivid 7, General Electric Medical Systems, Milwaukee, Wisc, USA) provided good visualization of the mitral valve when looked at from the apex in both parasternal (see Supplementary data online, Video 1) and apical views (see Supplementary data online, Video 2) showing a full bridge at leaflets edges level (see Supplementary data online, Video 1) but became partial only when the cutting plane was translated towards the annulus (see Supplementary data online, Video 3). Three-dimensional interrogation of the tricuspid valve confirmed the tethering of the posterior leaflet and allowed excellent morphological assessment of the regurgitant orifice with lack of leaflets coaptation at the posterior aspect of the valve (see Supplementary data online, Video 4).

Double orifice mitral valve is a rare congenital heart disease. We present a case in which 3D echocardiography helped clarify the underlying anatomy. The need exists for more extensive use of this technique to allow for surgical and pathological confirmation.

Double orifice mitral valve is a rare congenital malformation. It has been associated with various other congenital defects, mainly left heart obstructive lesions and AV canal, and the clinical picture varies from symptomatic to unexpected necropsy findings. The anatomical classification relies on identifying the extent of the fibrous ridge between the leaflets. In 2D echocardiography, this requires careful scanning from apex to base in parasternal short axis view. This provides a limited perspective and is not technically satisfactory in all patients. We describe an asymptomatic case of DOMV with associated dysplastic tricuspid valve. Combined tri-dimensional scanning in both parasternal and apical views identified a fibrous ridge at leaflets edge level only, suggesting an incomplete ridge-type DOMV. The added diagnostic value of 3D echo in this case was noteworthy.
and needs to be established in additional cases with surgical corroboration.

Supplementary data
Supplementary data are available at European Journal of Echocardiography online.

References