Three-dimensional echocardiographic evaluation of quadricuspid systemic atrioventricular valve

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A 56-year-old man became progressively dyspnoeic on exertion. He underwent an echocardiogram, which identified congenitally corrected transposition of the great arteries. Two-dimensional echocardiography showed that the systemic right ventricle was severely dilated with severely reduced systolic function. The left atrium was severely dilated (120 mL/m²). The systemic atrioventricular valve (tricuspid) was displaced apically by 18 mm (Panel A). Three-dimensional (3D) echocardiography demonstrated that the systemic atrioventricular valve (tricuspid) was quadricuspid with thickened leaflets (Panel B and see Supplementary data online, Video S1). Each papillary muscle was composed of two heads; each head connecting to the chordae of a single leaflet (Panel C and see Supplementary data online, Video S2). Three-dimensional colour Doppler identified severe tricuspid regurgitation into the dilated atrium (Panel D). Cardiac magnetic resonance imaging confirmed the findings of a quadricuspid systemic atrioventricular valve (Panel E). The patient underwent cardiopulmonary exercise testing, which showed reduced maximal oxygen consumption (peak VO₂ 17.4 mL/kg/min which is 41% of predicted). Morphological abnormalities of the systemic atrioventricular valve (tricuspid) are found in the majority of patients with congenitally corrected transposition of the great arteries. The most commonly abnormality is a displaced ‘Ebstein-like’ valve or less commonly a dysplastic valve. In our case, the valve was apically displaced but also composed of four leaflets. We believe that this is the first reported case of a quadricuspid systemic atrioventricular valve. Three-dimensional echocardiography demonstrated the precise morphological abnormalities of the quadricuspid systemic atrioventricular valve and its sub-valvular apparatus.

Supplementary data are available at European Heart Journal – Cardiovascular Imaging online.