An asymptomatic 35-year-old male was referred for echocardiography due to an abnormal ECG. Two-dimensional transthoracic echocardiogram (2D TTE) showed a displacement of the septal leaflet of the tricuspid valve towards the apex (Panel A1). The diagnosis of Ebstein’s anomaly was established. The 2D TTE study suggested the existence of a double-orifice tricuspid valve (Panel A2; Supplementary data online, Movie S1) with mild tricuspid regurgitation across both orifices (Panel A3; Supplementary data online, Movie S2). Three-dimensional transthoracic echocardiogram (3D TTE) was performed to more accurately define the valve structures using GE Vivid 9. 3D TTE showed a tricuspid valve with double orifice, with normal and completely independent opening and closing movements. A larger main orifice appeared situated in the medial position with a minor orifice, close to the septum (Panels B and C; Supplementary data online, Movies S3 and S4).

Given the excellent functional class, the absence of complications, or significant tricuspid stenosis or regurgitation, the patient did not require any type of intervention.

A double-orifice valve is an uncommon anomaly. Most of the published cases are described in the mitral valve. This anomaly is characterized by a valve with a single fibrous annulus with two orifices that open into the ventricle. Ebstein’s anomaly is a congenital heart disease with a highly variable anatomical presentation. The existence of a double-orifice tricuspid valve is an extremely rare finding.

Transthoracic three-dimensional echocardiography can see the tricuspid valve from the right atrium and the right ventricle, providing a detailed information of the abnormal tricuspid valve anatomic structures in the Ebstein anomaly.

Panel A1. A four-chamber view. Apical displacement of the tricuspid septal leaflet with the consequent atrialization of part of the right ventricle. RA, right atrium; LA, left atrium; RV, right ventricle; LV, left ventricle; TV, tricuspid valve.

Panel A2. An apical coronary sinus view in diastole, which suggests a double-opening tricuspid with one larger most medial orifice (TVm) and other minor close to the ventricular septum (TVs) (Supplementary data online, Video S1).

Panel A3. The same view with colour Doppler in systole, appreciated a mild tricuspid insufficiency in both orifices (Supplementary data online, Video S2).

Panel B. Three-dimensional TTE showing the tricuspid valve from the atrium. Double-orifice opening of the tricuspid valve in diastole is displayed.

Panel C. Three-dimensional TTE showing the tricuspid valve from the ventricle. Double orifice of the tricuspid valve during the cardiac cycle is displayed. Closed in systole (C1), completely open in proto-diastole (C2), with partial closure in the diastasis (C3) and new aperture after the atrial contraction (Supplementary data online, Videos S3 and S4).

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

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