



A young male with progressive dyspnea: Ebstein's anomaly

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A 32-year-old man presented to the out-patient clinic with complaints of progressive dyspnea. The symptoms began 2 years before the patient's presentation to our clinic, he also reported occasional palpitations in the prior month. On initial examination, the pulse was regular, heart rate was 107 bpm, and blood pressure was 110/70 mmHg in the right arm and 110/60 mmHg in the left arm in both standing and supine positions. There was no jugular venous distention, no kussmaul sign, and no hepatojugular reflux. On chest auscultation, there was a systolic, III/VI murmur located on the left parasternal border, fifth intercostal space that increased during inspiration. Also, there was a fixed splitting of the second heart sound, there was no evident lower extremity edema. A 12-lead electrocardiogram showed a complete right bundle branch block, and non-specific abnormalities of the QRS complex. A transthoracic echocardiogram was performed, findings are shown in Fig. 1a, b. A diagnosis of Ebstein's anomaly (EA) was established based on the echocardiographic findings, the patient was started on diuretics and inotropes, and referred to the cardiothoracic surgery clinic, but was lost to follow-up.

Ebstein's anomaly is a congenital malformation of the tricuspid valve (TV) and right ventricle (RV) that accounts for < 1% of all congenital heart diseases [1]. It is secondary to a failure of the primitive TV to delaminate from the developing right ventricular myocardium. It is characterized by adherence of the septal and posterior leaflets of the TV to the right ventricular myocardium with apical displacement of the TV functional annulus and atrialization of the

RV [1, 2]. Echocardiography is the diagnostic method of choice, showing an apical displacement of the septal leaflet (compared with the position of the anterior leaflet of the mitral valve) $\geq 8 \text{ mm/m}^2$ of the body surface area being a key diagnostic finding [3]. Abnormal coaptation of the TV leaflets during systole with consequent TV regurgitation and atrial septal defect (ASD) is also a common feature [1, 3]. Surgical repair typically consists of TV repair, plication of the atrialized RV, right reduction atrioplasty, and partial or total closure of any ASD [2]. The "cone reconstruction," which includes complete surgical delamination and recruitment of the undelaminated leaflet tissue for re-anchoring at the anatomic right atrio-ventricular junction, with the creation of a 360° leaflet cone, can achieve nearly anatomic restoration of the TV and has shown improvement in the grade of tricuspid regurgitation, functional class, and mortality [2, 4]. Risk of infective endocarditis in patients with EA is low; however, antibiotic prophylaxis is recommended before high-risk procedures (i.e., dental procedures with manipulation of the gingival region or perforation of the oral mucosa), particularly in cyanotic patients [1, 5]. EA is an uncommon cause of dyspnea. Symptoms depend on the severity of anatomic alterations, RV function, size of ASD, and coexisting dysrhythmias [1]. Although symptoms can be non-specific, an echocardiogram can show typical features that can lead us to the diagnosis.

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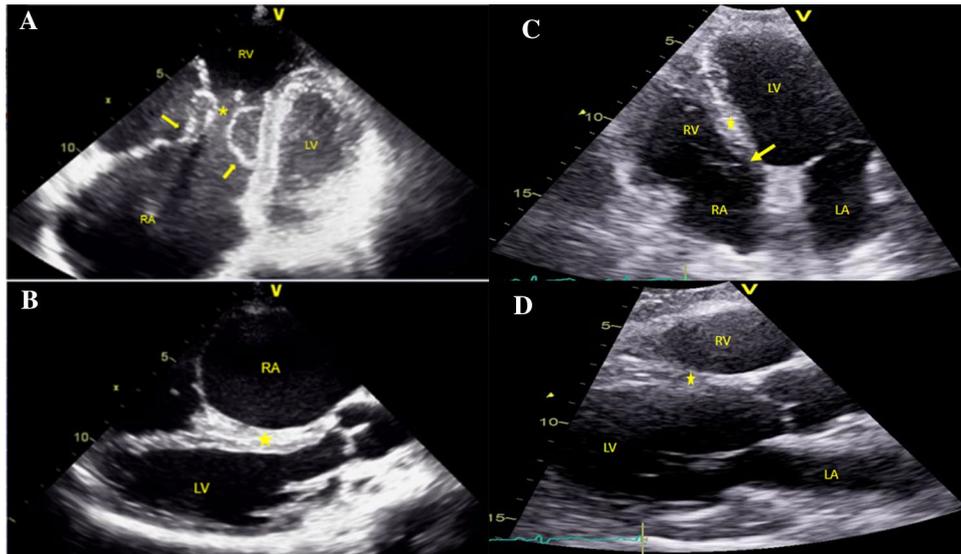


Fig. 1 **a** Transthoracic echocardiogram in the apical four-chamber view, right atrium (RA) that shows severe dilatation. Arrows point to the anterior and posterior leaflets of the tricuspid valve that are attached to the right ventricular myocardium. Asterisk—absence of coaptation of the tricuspid valve leaflets during systole that caused severe tricuspid regurgitation. Small functional right ventricle (RV). **b** A long-axis parasternal view on the echocardiogram. Severely dilated right atrium (RA). Filled star—Interventricular septum, note the leftward bowling of the septum as a consequence of the severe dilatation of the right cavities. Left ventricle (LV) that is being com-

pressed by the right cavities. **c** A four-chamber view of a normal heart. See how right ventricle (RV) is normal in morphology. Arrow points at the tricuspid valve, which is normally placed, and there is no atrialization of the right ventricle. Right atrium (RA), with normal diameter and morphology. **d** A long-axis parasternal view of the same normal heart. Right ventricle (RV), note how RA is not visible in this view because it is not dilated. Filled star—interventricular septum, which is normally placed, with no leftward bowling. LV shows the left ventricle with which the diameter is preserved. Left atrium (LA)

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Compliance with ethical standards

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