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# An unexpected cause of syncope

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**An unexpected cause of syncope**

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A 60-year-old female patient with hypertension, and no history of tachy- or bradyarrythiams, admitted to the Emergency Department due to fatigue and shortness of breath during mild exercise. She referred 4 episodes of lost of consciousness the previous months, the last one 2 days ago. On arrival, the patient was stable with normal vital signs.

The electrocardiogram (ECG) was in sinus rhythm, with right bundle branch block (heart rate 58 bpm). Cardiovascular examination revealed a holosystolic murmur in the fourth intercostal space in the parasternal region. In transthoracic echocardiogram the left ventricle had normal function with normal wall thickness, increased dimensions of the left and right atrium, normal left ventricle filling pressures, mild regurgitation of the aortic valve, mild to moderate regurgitation of the mitral valve, moderate regurgitation of the tricuspid valve (Figure 1D) and normal dimensions of the inferior vena cava. The most important finding was the apical displacement of the septal leaflet of the tricuspid valve and the variable tethering of the leaflet tissue to the myocardium of the right ventricle (Figure 1B–C). Due to high suspicious of atrial septal defect, the patient underwent a transesophageal echocardiogram, that confirmed the findings of the bedside transthoracic echocardiogram and especially the apical displacement of the septal leaflet of the tricuspid valve and the variable tethering of the leaflet tissue to the right ventricle (RV) myocardium (atrialized ventricle) and moderate regurgitation of the valve. It also revealed an atrial septal defect, with no hemodynamic significance (Figure 1E–F). During her

hospitalization the patient had an episode of fading. A new ECG revealed a 2nd degree heart block (2:1). (Figure 1A). With the combination of clinical, echocardiogram and electrocardiogram findings we were able to confirm the diagnosis. It was a 2nd degree heart block due to the first diagnosed Ebstein anomaly disease and the cause of the symptoms was chronotropic incompetence. The patient was transferred to the electrophysiology department and a permanent pacemaker was implanted. Because of the technical difficulties caused by the enormous and without anatomical borders right atrium, a VVI pacemaker was implanted to the patient. European Society of Cardiology guidelines recommend surgical repair in patients with severe tricuspid regurgitation and symptoms or objective deterioration of exercise capacity (class I, level C). They also recommend surgical repair regardless of symptoms in patients with progressive right heart dilatation or reduction of RV systolic function (class IIa, level C) [1]. Our patient had no episodes of dyspnea or other symptoms in the past. She was admitted to a Congenital Diseases Center for evaluation and a conservative therapy with regular follow up was decided for the patient.

Ebstein anomaly is a rare congenital disorder that involves dysplasia of the triscuspid valve. Specifically, it is characterized by right atrial dilatation, apical displacement of the septal or anterior leaflet, tethering of the leaflet tissue, atrialized RV and reduced RV systolic function [2]. Symptoms and sings can occur at any age , but rare at the 6th decade of life [3]. ECG usually includes PR interval prolongation, tall P waves, and the presence of right bundle branch block [4]. Rarely, Ebstein anomaly is combined with conduction disorders [5]. We report the rare case of a first — diagnosed Ebstein anomaly in a 60-year-old, who presented with a 2nd degree heart block that was successfully managed with the implantation of a VVI pacemaker.

**Article information**

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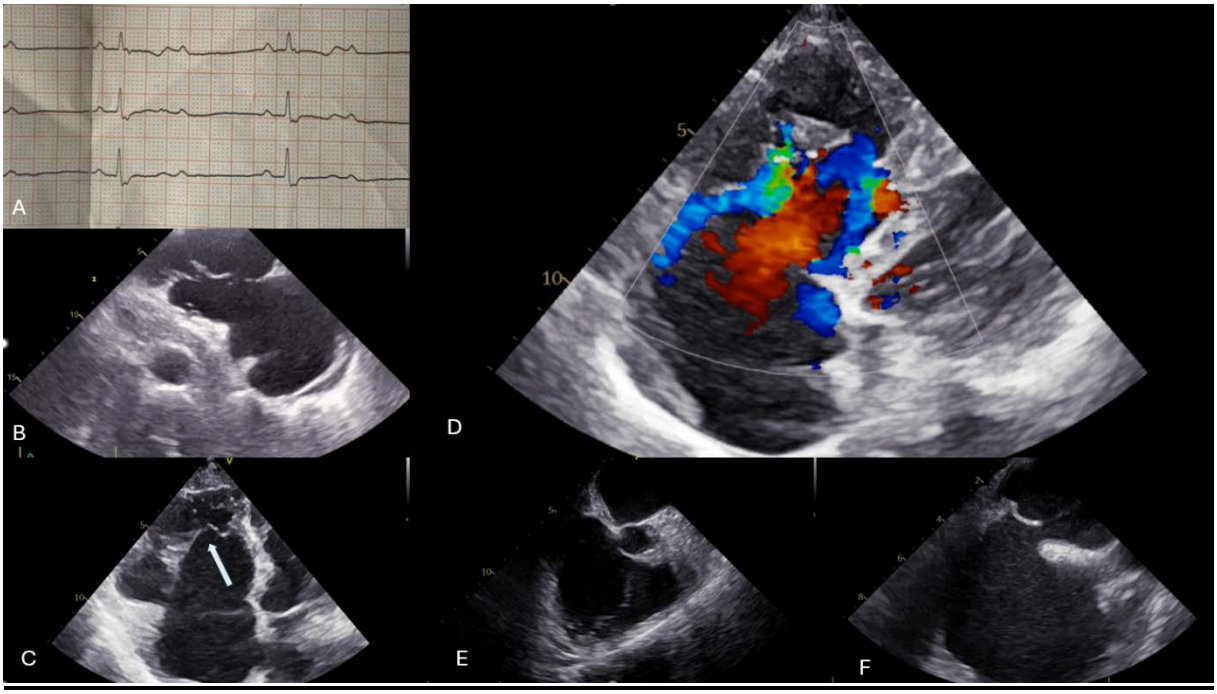


**Figure 1.** ECG, TTE and TOE in a patient with Ebstein Anomaly. **A.** ECG revealed a 2nd degree heart block (2:1). **B.**, **C.** TTE revealed apical displacement of the septal leaflet of the tricuspid valve and the variable tethering of the leaflet tissue to the myocardium of the right ventricle

(white arrow). **D.** Moderate regurgitation of the tricuspid valve. **E.**, **F.** TOE confirmed the TTE findings. It also revealed an atrial septal defect, which no hemodynamic significance