Successful pregnancy course and outcome in a patient with unusual coincidence of two structural heart defects: Ebstein anomaly and biventricular non-compaction cardiomyopathy — extremely rare, but there!

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We present the case of a 26-year-old pregnant woman (GIPI, 30 hbd) with an Ebstein anomaly (EA) confirmed by two-dimensional transthoracic echocardiography (2D TTE). The patient was referred to the Cardiology Department because of dry cough, dyspnea at rest, and cardiac arrhythmias that had persisted since 27 hbd. On admission, the patient's general condition was average, and laboratory tests (high sensitivity troponin I, B-type natriuretic peptide) were normal. An electrocardiogram (ECG) showed sinus tachycardia, dextrogram, incomplete right bundle branch block, ST-segment elevation in lead V1, tall and peaked T waves in leads I, II, V2–V5; biphasic P waves in lead V1 (Figure 1A). A Holter ECG was performed, which recorded 4 non-sustained ventricular tachycardias (nsVTs) consisting of polymorphic evolutions (Figure 1B). 2D TTE showed displacement of the tricuspid valve (TV) leaflets by 21 mm with visible signs of the right ventricle (RV) atrialization and severe tricuspid regurgitation. Additionally, impaired left ventricular (LV) contractility (LV ejection fraction [LVEF] ~43%) and increased LV trabeculation were noted in the apical region, which was not visualized during previous TTEs (Figure 1C–D). Following the tests, extended-release metoprolol was added to therapy. During hospitalization, the fetus's well-being was also monitored — the correct fetal position and optimal flows in the umbilical artery.

Subsequently, in 36/37 hbd the patient delivered a live daughter (birth weight — 2280 g, Apgar 10 at 1 min) during a planned C-section. The early postpartum period was uneventful; no arrhythmias were observed. After postpartum discharge, a control Holter ECG was performed twice at home; in neither examination did nsVTs recur. Due to the inability to perform magnetic resonance, a cardiac tomography multiplanar reconstruction was then performed, confirming EA with an intraventricular displacement of the TV plane by approximately 50 mm; significant deepening of the LV free and inferior wall trabeculae and periapically in the RV were clearly visible. Global and regional contractility disorders of both ventricles were noted — LVEF ~29%, RVEF ~38%. (Figure 1E–F; Supplementary material, *Figure S1*). Due to significantly decreased LVEF, implantation of a cardioverter-defibrillator for primary prevention of sudden cardiac death was considered. The patient started treatment for heart failure with reduced LVEF according to the European Society of Cardiology guidelines consisting of a beta-blocker, angiotensin-receptor neprilysin inhibitor, and loop diuretic.

EA constitutes a rare congenital heart disease with varying degrees of downward displacement of the septal and posterior TV leaflets, triggering RV atrialization and dysfunction; the prevalence is 1:200 000 live births. [1] During pregnancy, the probability of right ventricular failure rises due to an increase in blood volume, cardiac output, and afterload. [2] The clinical picture of non-compaction cardiomyopathy varies, ranging

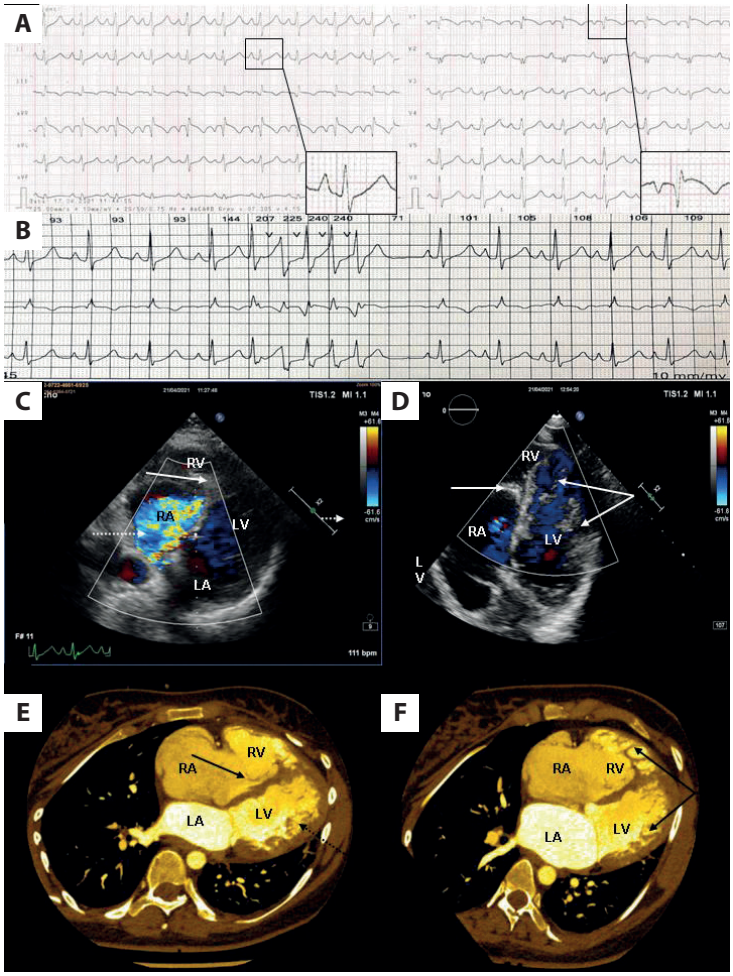

Figure 1. A. ECG on admission: sinus tachycardia 105/min, right axis deviation, IRBBB. ST-segment elevation in lead V1, tall and peaked P waves in leads I, II, and V2–V5 I; biphasic P waves in lead V1. **B.** Holter ECG: nsVT consisting of polymorphic evolutions. **C.** Echocardiography — 2D TTE SC: displacement of the septal leaflet of the tricuspid valve towards the RV (solid arrow) with visible signs of RV atrialization. **D.** Echocardiography — 2D TTE modified 4C: displacement of the septal leaflet of the tricuspid valve towards the RV (solid arrow) with visible signs of RV atrialization and increased LV trabeculation with blood turbulence in the intertrabecular lacunas on color Doppler in the LV apical region (double solid arrows). **E.** CT MPR 4C: displacement of the septal leaflet of the tricuspid valve towards the right ventricle (solid arrow) with visible signs of RV atrialization and increased left ventricular trabeculation in the LV apical area (dashed arrow). **F.** CT MPR 4C: visible increased ventricular trabeculation in both the RV and LV apical areas (double solid arrows)

Abbreviations: 2D TTE 4C, two-dimensional transthoracic echocardiography 4-chamber apical view; 2D TTE SC, two-dimensional transthoracic echocardiography subcostal view; CT MPR 4C, cardiac tomography multiplanar reconstruction 4-chamber view; CT MPR TV, cardiac tomography multiplanar reconstruction transverse view; ECG, electrocardiogram; IRBB, incomplete right bundle branch block; LA, left atrium; LV, left ventricle; nsVT, non-sustained ventricular tachycardia; RA, right atrium; RV, right ventricle

from asymptomaticity to dangerous manifestations, such as chronic heart failure, arrhythmias, thromboembolic episodes, and sudden cardiac death [3]. Non-compaction cardiomyopathy, a rare myocardial abnormality, sometimes coexists with Ebstein disease in approximately 5% of patients, significantly worsening the prognosis [4]. The described coexistence of these two very uncommon heart diseases can definitely be included in the casuistry. Despite medical advances, cardiovascular diseases are still the leading cause of non-obstetric maternal deaths during pregnancy (0.5%–4% in the West) [5].

Supplementary material

Supplementary material is available at https://journals.viamedica.pl/kardiologia_polska.

Article information

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