Ebstein anomaly with severe tricuspid valve regurgitation: An unusual case of an 84-year natural course

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Ebstein anomaly (EA) is one of the rarest congenital heart defects (1%) [1]. This disorder is characterized by the abnormal formation of the leaflets of the tricuspid valve (TV) with their displacement towards the apex of the right ventricle (RV) [2]. The consequence of the described changes is the creation of a large, "functional" right atrium (RA), which morphologically consists of the RA and the atrialized RV (aRV), by which the right ventricle (fRV) is anatomically reduced and accompanied by TV regurgitation [2].

A female patient with accidentally diagnosed EA, who lived to be 84, was under the care of the Adult Congenital Heart Defects Clinic for 12 years. The patient was referred to our center at the age of 70 (2007) by a cardiologist who performed a follow-up echocardiographic examination due to moderate hypertension and hyperlipidemia. She had two uncomplicated natural births. During the first visit, the patient's condition was good — she did not report cardiac arrhythmias, and was in the New York Heart Association class I. She was permanently on a statin, an angiotensin-converting enzyme inhibitor, and a calcium channel blocker. Physical examination — regular heart rate of 70/min, systolic murmur in the 4th and 5th intercostal spaces on the right side, and a typical symmetrical vesicular murmur. Electrocardiogram showed regular sinus rhythm and a right bundle branch block. Transthoracic echocardiography showed typical features of EA: apical displacement of the septal TV leaflet by 22 mm and significant TV regurgitation. The examination also showed enlargement of the right heart (56 mm in the parasternal long-axis view), good RV function, and the Celermajer echocardiographic index ([Cel-ind] = [RA + aRV]/[fRV + left atrium + left ventricle]) was 1.1, which indicates a significant severity of the defect (grades: 1 = Cel-ind <0.5; 2 = 0.5–0.99; 3 = 1.0–1.49; 4 >1.5) [3]. The Cel-ind assessed by MRI was 0.72 (grade 2) (Figure 1) [3]. Significant TV regurgitation was confirmed (Supplementary material, *Video S1* and *S2*). During the next follow-up visit (2019), the patient reported moderate fatigue and periodic swelling around the ankles, with the presence of venous insufficiency (B-type natriuretic peptide — 89.2 pg/ml). In 2021, the patient died of an ischemic stroke.

The presented case report is (to the best of our knowledge) the only published description of a patient who lived to such an advanced age, despite a severe, unoperated form of EA, without presenting clinical symptoms associated with this congenital defect. It is known that diagnosis in adulthood is a favorable prognostic factor in this population [1]. The observation of 51 patients with EA performed by Celermajer et al., among whom only 18% were operated on, showed good long-term survival in the majority of the conservatively treated group — as many as 81% of the patients reached the age of 60 [4] in our adult congenital heart disease (ACHD). Although surgical treatment is recommended and more widely used in symptomatic patients, as much as 34%–38% require reoperation, and the survival rate is estimated at 71.3%–84% 20 years after surgery [2, 5]. The case we described proves that the wait-and-see attitude proposed by Celermajer may be an acceptable approach even in patients with a significant intensification of the defect because, as it can be seen, they can live to old age in good general condition. Therefore, it is

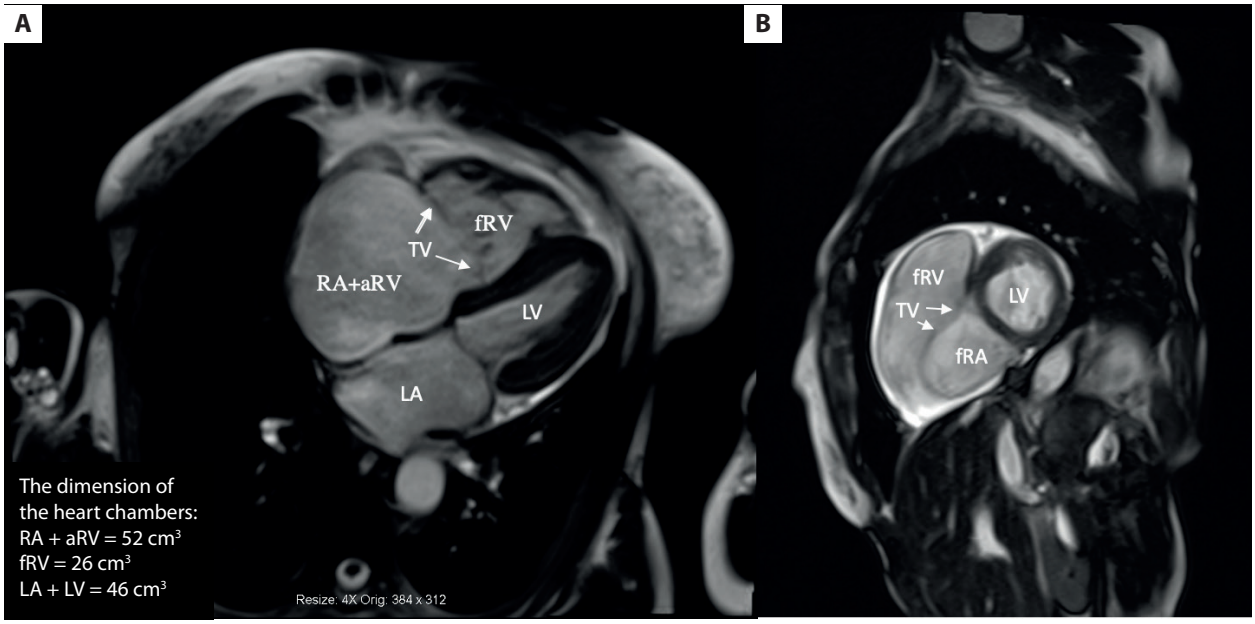

Figure 1. Cardiac magnetic resonance. **A.** Four-chamber view. **B.** Short axis view

Abbreviations: aRV, atrialized right ventricle; fRA, functional right atrium; fRV, functional right ventricle; LA, left atrium; LV, left ventricle; TV, tricuspid valve leaflets

crucial to carefully consider indications for cardiac surgery in each patient with congenital heart disease.

Supplementary material

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

Article information

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