

# How can a 4-liter heart fit in the human chest? Ebstein anomaly diagnosed in adulthood

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A 38-year-old man presented to the outpatient cardiology clinic with palpitations and a slight limitation in physical activity (functional class II according to the New York Heart Association). Electrocardiography revealed atrial fibrillation. On transthoracic echocardiography, typical features of Ebstein anomaly were observed: apical displacement of the offset of the septal tricuspid leaflet  $>8$  mm/m<sup>2</sup> and significant tricuspid regurgitation. Apical displacement of the septal tricuspid leaflet divides the right heart into 3 chambers: the functional right ventricle, the atrialized right ventricle, which in combination with the anatomic right atrium constitutes the functional right atrium.<sup>1</sup> Echocardiography demonstrated a significant enlargement of the right ventricle (diameter of 104 mm in the parasternal long-axis view) with spontaneous echo contrast within the chambers (FIGURE 1A and 1B; Supplementary material, *Video S1*). Further imaging with cardiac magnetic resonance confirmed enormous volumes within the right heart chambers with a severe systolic dysfunction of the functional right ventricle (right atrium, 3300 ml; reference range, 37–169 ml; right ventricle, 426 ml; reference range, 118–250 ml; right ventricular ejection fraction, 27%; reference range, 52%–72%), whereas size and function of the left ventricle remained close to normal (left ventricle, 142 ml; reference range, 106–214 ml, left ventricular ejection fraction, 50%; reference range, 57%–67%).<sup>2</sup> No intraatrial shunt was demonstrated (Supplementary material, *Videos 2 and 3*).

Cardiopulmonary testing demonstrated moderately impaired exercise capacity (peak oxygen uptake [VO<sub>2</sub>], 17.9 ml/kg/min; 53% predicted

peak VO<sub>2</sub>; respiratory exchange ratio, 1.19) with blood oxygen saturation pre- and post-examination of oxygen saturation by pulse oximetry (SpO<sub>2</sub>) of 97% and 96%, respectively. Despite the cardiac size was extremely increased, basic pulmonary function was preserved with no features of restrictive lung disease (forced vital capacity, 4.32 l; 88% of predicted forced vital capacity; ratio of forced expiratory volume in 1 second to forced vital capacity, 70%). Brain natriuretic peptide was only mildly elevated (165.8 pg/ml; reference range, 0–100 pg/ml).

Due to paroxysmal atrial fibrillation and spontaneous echo contrast present in the severely enlarged cardiac chambers, oral anticoagulants along with bisoprolol were administered.

In Ebstein anomaly, a late diagnosis (in adult life) portends a favorable prognosis even without any previous cardiosurgical interventions.<sup>1</sup> Nevertheless, this degree of cardiomegaly, in combination with a clinically benign course, is highly unusual, even in patients with congenital heart disease. The most probable cause of such cardiac enlargement is the significant tricuspid regurgitation leading to volume overload.<sup>1</sup> The activation of the neurohormonal axis (renin–angiotensin–aldosterone) results in a dysregulated process of collagen turnover and, subsequently, increased myocardial fibrosis.<sup>3</sup> The described alterations favor the continuous cardiac enlargement, but also an occurrence of supraventricular arrhythmias, including atrial fibrillation.<sup>3,4</sup>

The preserved lung capacity and function, despite enormously enlarged heart, is also an unexpected finding. The adaptation in respiratory mechanics in this patient might be similar to that seen in pleural effusions. The fluid

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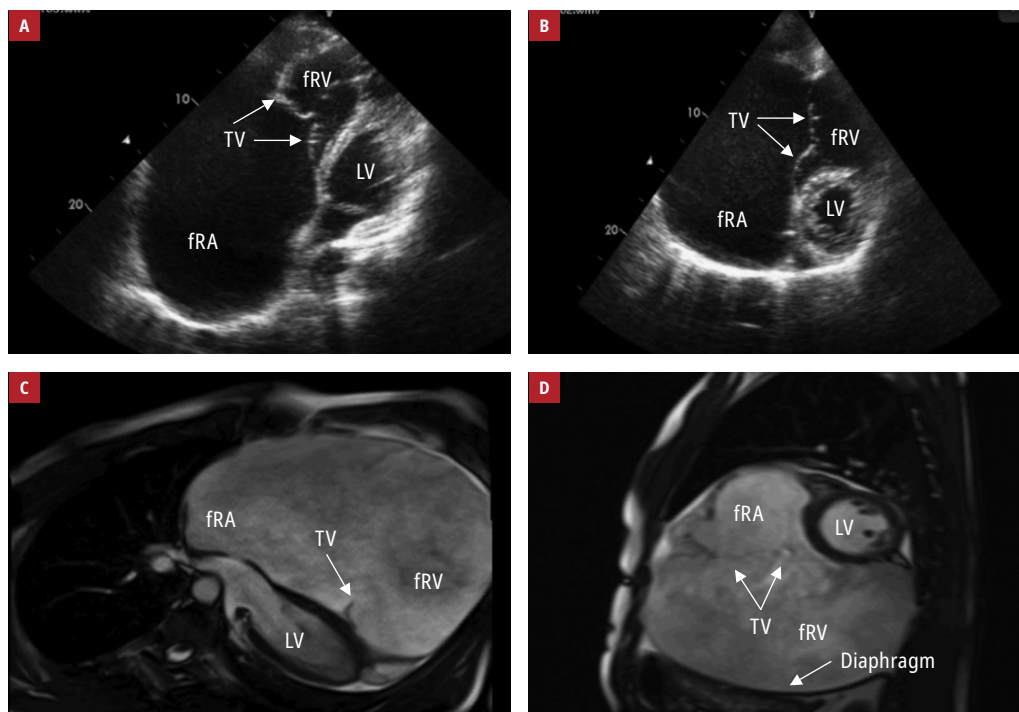
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**FIGURE 1** Different imaging modalities in Ebstein anomaly: **A** – transthoracic echocardiography, the apical 4-chamber view; **B** – transthoracic echocardiography, the parasternal short axis view; **C** – cardiac magnetic resonance, the transaxial plane; **D** – cardiac magnetic resonance, the sagittal plane. Notice the inverted diaphragm dome.

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

in the pleural cavity is usually accommodated mainly through the expansion of the thoracic cage (through the downward displacement of the diaphragm) rather than the lung compression and restrictive pattern.<sup>5</sup>

Ebstein anomaly might cause enormous cardiac enlargement without overt heart and lung failure symptoms. The congenital character of the disease probably allows for a clinically benign course. The adaptation mechanisms of the cardiac and respiratory system are long-lasting and multifactorial and they demonstrate an astonishing capacity of the human body to cope with pathology. This case also proves that despite the enormous remodeling in patients with complex congenital heart defects, even those unoperated, may survive until late adulthood. Therefore, indications for cardiac surgery should be carefully considered in every individual.

#### SUPPLEMENTARY MATERIAL

Supplementary material is available at [www.mp.pl/kardiologiapolska](http://www.mp.pl/kardiologiapolska).

#### ARTICLE INFORMATION

**CONFLICT OF INTEREST** None declared.

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