Prošireni sažetak / Extended abstract

Three-dimensional echocardiography in classification of Ebstein's anomaly: a case report on agenesis of the tricuspid valve posterior leaflet

Ivo Planinc*, Željko Baričević, Vlatka Rešković Lukšić, Daniel Lovrić, Sandra Večerić, Jadranka Šeparović Haneževački

Clinical Hospital Center Zagreb, Zagreb, Croatia

We present an echocardiographic evaluation of a 27-year-old young male patient who was referred to our institution after a loud systolic murmur was found during his occupational health check-up.. Besides the murmur, his physical exam, and results of his laboratory tests were normal. He has no significant complaints.

His first 2D transthoracic echocardiogram (TTE) revealed severely dilated right ventricle (RV area greater than left ventricular area), volume overloaded and excentrically hypertrophied (wall thickness >6 mm), with severe tricuspid regurgitation (vena contracta 12 mm), large abundant sail-like anterior leaflet of the tricuspid valve (TV), apically displaced short septal leaflet (17 mm), and in the same time normal left ventricular systolic function and dimensions. These findings, especially apical displacement of septal leaflet indicate Ebstein's anomaly.

Ebstein's anomaly is a rare congenital heart defect that occurs in only about <1% of congenital heart disease. In normal human hearts the apical displacement of the septal and posterior TV leaflets is <8 mm/m^2 body surface area from the level of anterior mitral valve insertion. Differential diagnosis includes TV dysplasia, TV prolapse, TV endocarditis, and arrhythmogenic right ventricular cardiomyopathy, to name a few. With 2D TTE we were positive that this patient had some form of Ebstein's anomaly, however we could not generate important information about valvular apparatus, such as morphology of the leaflets, the true origin of septal and posterior leaflets as well as precise coaptation. Those information are necessary in order to complete classification of TV pathology.

Therefore we obtained 3D TTE and 3D TEE, and with multiplanar review mode and 3D reconstruction revealed complete agenesis of the posterior leaflet leaving almost unguarded tricuspid orifice with a large zone of lack of coaptation as the origin of severe tricuspid regurgitation. Subsequent 3D analysis also verified absence of complete posterior leaflet subvalvular apparatus, or in the context of embryological development, complete absence of posterior leaflet delamination.

We used 2D echocardiography to study morphology and function of right ventricle, but 3D echocardiography provided additional important information regarding the morphology and orientation of TV and its apparatus. Only 3D TEE obtained agenesis of the posterior leaflet. According to parameters derived by 2D TTE and 3D TEE this patient was classified as type I Ebstein's anomaly.

KEYWORDS: Ebstein's anomaly, three-dimensional echocardiography, congenital heart disease, multiplanar review mode, severe tricuspid regurgitation.

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*Address for correspondence: Klinički bolnički centar Zagreb, HR-10000 Zagreb, Croatia.

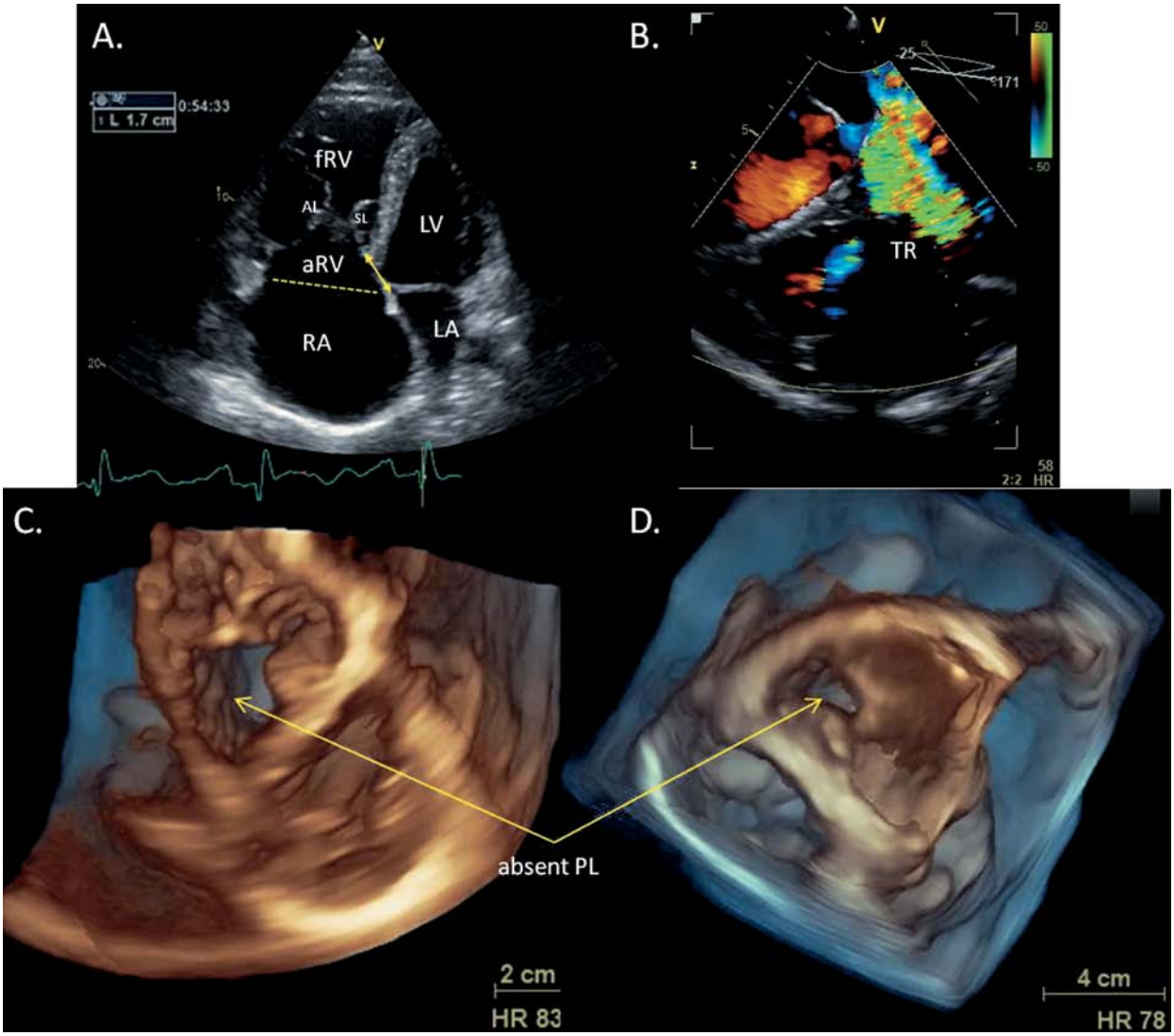
Phone: +385-1-2388-888

E-mail: ivo.planinc@gmail.com

Table 1. *Classification of Ebstein's anomaly based upon the anatomic findings at surgery (modified from Dearani JA, et al. Ann Thorac Surg. 2000;69:S106.).*

Type	Tricuspid valve leaflets			Size of atrialized RV
	Anterior	Posterior	Septal	
I	Large — Mobile	Apically displaced or dysplastic or absent		Varies
II	Small — Spirally displaced toward apex			Moderately large
III	Restricted motion Shortened and fused chordae	Displaced and dysplastic		Large
IV	Severely deformed Displaced in RVOT	Displaced or absent	Ridge or fibrous material	Extremely large

RV = right ventricle, RVOT = right ventricular outflow tract.



RA = right atrium; aRV = atrialized right ventricle; fRV = functional right ventricle; LA = left atrium; LV = left ventricle; AL = anterior leaflet; SL = septal leaflet; TR = tricuspid regurgitation; PL = posterior leaflet.

***Figure 1. A** The four chamber view showing displacement of septal leaflet, large anterior leaflet and size of atrialized right ventricle; **B** Transesophageal view of severe tricuspid regurgitation; **C** and **D** Three-dimensional reconstructed multiplanar view of tricuspid valve with large zone of no coaptation and absent posterior leaflet.*

Literature

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