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Echocardiography in a young adult with congenital heart disease presenting with isolated right-sided heart failure – Ebstein's anomaly

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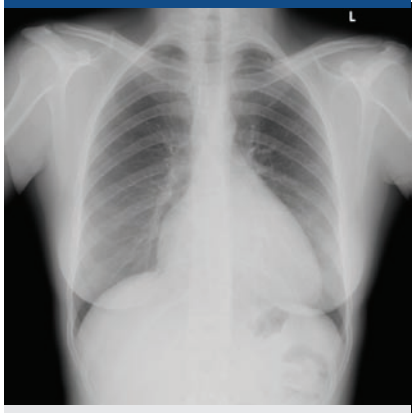
Ebstein's anomaly as a rare congenital disorder serves as a model of right ventricle dysfunction and altered atrial and ventricular coupling. It is characterized by failure of delamination of tricuspid valve leaflets and downward-apical displacement of the tricuspid valve attachments, apical displacement of the tricuspid valve due to adherence of the septal and posterior leaflets to the interventricular septum, redundancy, fenestration and tethering of the anterior tricuspid valve leaflet, dilatation of the anatomic (true) valve annulus, resulting in valve insufficiency and partial atrialization of the right ventricle.[1-4]

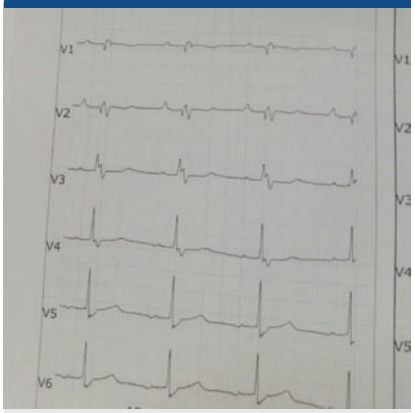
We report 36-year-old female. She presented with exertional dyspnoa. Enlarged right atrium and ventricle, a hump-shaped infundibulum was evident on chest radiograph (**Figure 1**). ECG showed atrial intraventicular conduction delay (**Figure 2**). The 2D echocardiogram (**Figure 3**) revealed the presence of poor right ventricular function and atrialization of the right ventricle, malformation of the tricuspid valve (TV) and the right ventricle (RV). The most prominent morphological feature of EA was degree of apical displacement of the TV into the RV, dividing the RV into a proximal chamber of atrialized RV (aRV) and distal portion of functional RV. Massive tricuspid regurgitation (TR), extensive dilatation, and dysfunction of the right atrium (RA) and RV were found.

FIGURE 1.



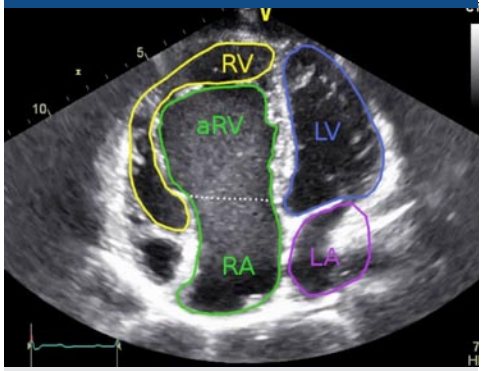
Chest radiograph showing enlarged right atrium and right ventricle.

FIGURE 2.



Fragmented QRS complexes observed in adult patients with Ebstein anomaly.

FIGURE 3.



Measurement of the severity of Ebstein anomaly. Planimetry was performed in the apical 4-chamber view at end diastole.

Poor right ventricular function was shown by the 2D echocardiogram including atrialization of the right ventricle, malformation of the tricuspid valve (TV) and the right ventricle (RV). The most prominent morphological feature of EA was degree of apical displacement of the TV into the RV, dividing the RV into a proximal chamber of atrialized RV (aRV) and distal portion of functional RV.

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LITERATURE

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