

A very rare case: Ebstein anomaly, biventricular noncompaction and pre-excitation in the same patient

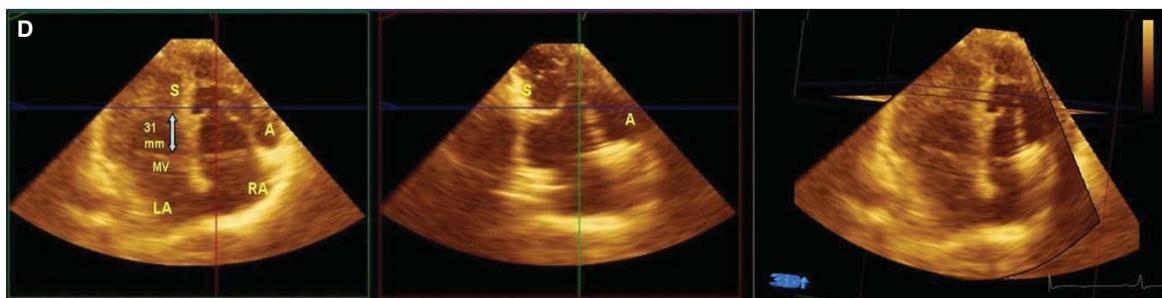
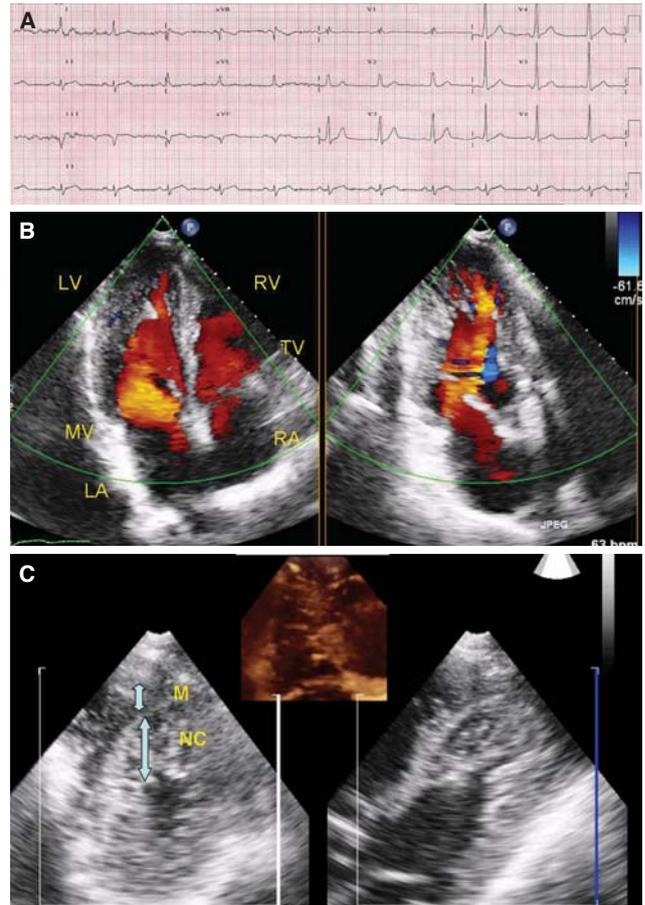
Çok nadir bir olgu: Aynı hastada Ebstein anomalisi, biventrikül süngerimsi miyokart (noncompaction) ve pre-eksitasyon

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A 20-year-old man was referred to our hospital for evaluation of a cardiac murmur. He was asymptomatic except for shortness of breath with strenuous exercise. On examination, a grade 1/6 systolic ejection murmur was audible at the apical region. An ECG showed a pre-excitation pattern (Fig. A). There were multiple intertrabecular recesses in two dimensional echocardiographic views (Fig. B, Video 1^{*}). The ratio of thicknesses of noncompacted and compacted layers of the myocardium in end-systole was greater than 2 (Fig. C). There was also moderate tricuspid regurgitation and multiple prominent trabeculations of both ventricles. Three-dimensional (3D) transesophageal echocardiography (TEE) was also carried out and revealed apical displacement of the septal leaflet of the tricuspid valve, tethering of the anterior leaflet and atrial dilatation with atrialized right ventricle (Video 2^{*}). 3D TEE was also showing prominent trabeculae and deep intertrabecular recesses of both ventricles. The three-plane analysis of reconstructed 3D echocardiographic images revealed that the septal leaflet of the tricuspid valve was apically displaced in both apical images (Fig. D). The distance from the mitral septal annulus to the septal leaflet attachment was

measured as 31 mm. Due to concerns about potential thromboembolic events, we started warfarin therapy. The patient is on regular follow up.



Figures– (A) Surface ECG showing the pre-excitation pattern. **(B)** Blood flow is shown in the recesses in both ventricles with color Doppler. **(C)** Ratio of noncompacted subendocardial layer to compacted myocardium exceeds two in systole. **(D)** The post-processing analysis of the reconstructed three dimensional images. The image with a green frame corresponds to apical four chamber view, showing septal leaflet attachment 31 mm apically to the mitral annulus. The image with the red frame is orthogonal to the image with the green frame, showing similar findings. **Supplementary video files associated with this case can be found in the online version.* LV: Left ventricle; RV: Right ventricle; LA: Left atrium; RA: Right atrium; MV: Mitral valve; TV: Tricuspid valve; M: Myocardium; NC: Noncompaction myocardium; S: Septal leaflet; A: Anterior leaflet.