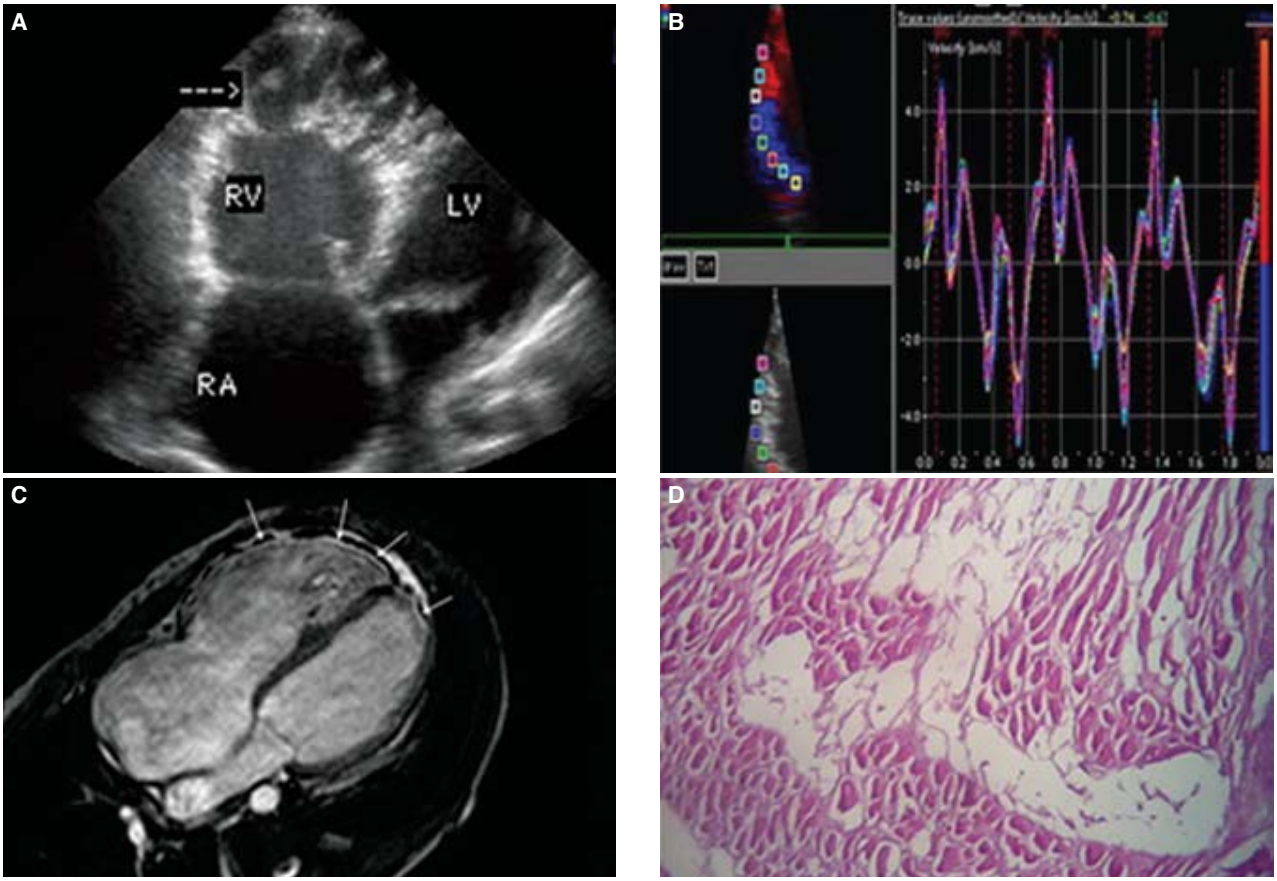
# Arrhythmogenic right ventricular cardiomyopathy with severe biventricular heart failure

# *Aritmojenik sağ ventrikül kardiyomiyopatisi ve ileri biventriküler kalp yetersizliği*

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Arrhythmogenic right ventricular cardiomyopathy (ARVC) is characterized by replacement of the myocardium with fatty and fibrous tissue. A 17-year-old male presented with a six-month history of progressive dyspnea, fatigue and abdominal pain. On evaluation, severe heart failure symptoms (predominantly right heart) were observed. Transthoracic echocardiography showed severely hypokinetic right ventricle, dilated right chambers, and bulging of apical right ventricular (RV) aneurysms with multiple septations (Fig. A). The left ventricle was not dilated, but showed moderately reduced systolic function (ejection fraction 40%). Tissue Doppler velocity curve analysis was used to assess regional RV function. When the sample volume was placed in the free wall, from the tricuspid annulus to the apex, it was observed that there was no base-mid-apex gradient with decreased peak systolic velocity and early diastolic velocity (Fig. B). Cardiac magnetic resonance imaging showed fatty infiltration of the right and left ventricular myocardium (Fig. C) with thinning of the RV free wall. Dilation of the right chambers was also noted. The patient underwent orthotopic heart transplantation with the diagnosis of ARVC. Fibrofatty replacement of myocytes in the right and left ventricles (lateral and apical wall) was observed on microscopic examination of the explanted heart (Fig. D).

**Figures. (A)** Transthoracic four-chamber view shows dilated right chambers and apical right ventricular bulging aneurysms with multiple septations (arrow). **(B)** Tissue Doppler velocity curve showing decreased systolic and early diastolic velocity with absence of base-mid-apex gradient. **(C)** Long-axis MRI scan shows extensively bright signals at the level of the right ventricular wall and left ventricular apex consistent with fat (arrows). **(D)** Histological section of the right ventricular wall showing myocytes replaced by fibrous and fatty tissue (H-E, x100).