A 23-year-old man was admitted to the hospital with severe heart failure and cachexia. Ventricular arrhythmias and progressive heart failure (predominantly right heart) had been observed in the previous 3 years. Physical examination was unremarkable except for a widely split second heart sound, a systolic left precordial lift, third and fourth heart sounds, and signs of increased venous pressure. Chest radiography showed significant cardiomegaly. The ECG was characterized by right atrial enlargement, low QRS voltages, wide complexes in the right precordial leads (epsilon

Figure 1. Top, Two-dimensional echocardiography. Left, Parasternal short-axis view at basal level. Right, Modified apical 4-chamber view. Both frames in systole. Severe right ventricular (RV) enlargement with aneurysmal dilatation of outflow tract (RVOT) and multiple wall bulges (right) are present. PA indicates pulmonary artery; RA, right atrium; LV, left ventricle; and LA, left atrium. Bottom, M-mode echocardiographic tracing at ventricular level. RV is severely enlarged; LV is not enlarged, but severe hypokinesis of both septum (IVS) and posterior wall (PW) is evident.
waves?), and negative T waves. Nonsustained ventricular tachycardia with polymorphic configuration was observed at ambulatory ECG monitoring.

The echocardiogram (Figure 1) disclosed severe right ventricular enlargement, with aneurysm at the level of the outflow, severe depression of systolic pump function, and multiple wall bulges. The left ventricle was not dilated but showed severe diffuse hypokinesis and depression of the ejection fraction (20%).

Considering the severity of clinical symptoms refractory to medical treatment and the echocardiographic findings, a cardiac transplantation was scheduled; unfortunately, the young patient died a few weeks later in low output during an acute febrile illness.

Postmortem examination was compatible with a severe form of biventricular dysplasia, with severe widespread myocardial atrophy and fibrofatty substitution (Figure 2). Postmortem MRI examination of the heart (T1-weighted spin-echo technique) showed severe right ventricular enlargement, with wall thinning and increased brightness, compatible with fatty infiltration (Figure 3); focal hyperintensity was also present at the level of the left ventricular apical aneurysm as a result of involvement by the pathological process.

Arrhythmogenic right ventricular dysplasia is a recently described cardiomyopathy characterized by predominant right ventricular involvement with myocardial atrophy and fatty or fibrofatty substitution, clinical signs of ventricular arrhythmias, and rarely, heart failure. Biventricular involvement was observed in several cases. MRI has the unique capability to identify noninvasively the structural abnormalities that characterize this disease, particularly the fatty infiltration.5

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Figure 2. Top left, Postmortem pathological section of heart (4-chamber): severe widespread fatty infiltration of right ventricular (RV) wall is present; an apical aneurysm is present at left ventricular level (arrow). Top right, Histological section at level of RV inflow (hematoxylin-eosin, ×2.5): severe transmural fibrofatty infiltration of RV wall is present, compatible with RV dysplasia. Bottom, Histological section at level of left ventricle (outflow) (hematoxylin-eosin, ×2.5) shows focal severe fibrofatty infiltration with myocardial atrophy, compatible with left ventricular involvement by pathological process.
References


Figure 3. Postmortem MRI (4-chamber view, similar to pathological section, Figure 2, top left) shows extensive hyperintense signal at level of right ventricular (RV) wall, compatible with fat; hyperintense areas are also evident at left ventricular level (LV). Note close relationship between presence of fat at pathology and hyperintense areas at MRI.
Right Ventricular Dysplasia With Biventricular Involvement
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