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IMAGING VIGNETTE

INTERMEDIATE

CLINICAL VIGNETTE

Combining 3-Dimensional Electroanatomic Mapping and Cardiac Magnetic Resonance

A New Tool for Arrhythmogenic Cardiomyopathy

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ABSTRACT

We present the case of a 17-year-old asymptomatic boy with a diagnosis of arrhythmogenic cardiomyopathy. Merging of cardiac magnetic resonance imaging and three-dimensional electroanatomic mapping provided striking visualization of the association between structural and electrical alterations and guided the decision to implant an implantable cardioverter defibrillator. (**Level of Difficulty: Intermediate.**) (J Am Coll Cardiol Case Rep 2022;4:1200–1202) © 2022 The Authors. Published by Elsevier on behalf of the American College of Cardiology Foundation. This is an open access article under the CC BY-NC-ND license (http://creativecommons.org/licenses/by-nc-nd/4.0/).

The diagnosis of arrhythmogenic cardiomyopathy (ACM) remains a clinical challenge. An upgrade of previous criteria for diagnosis of the entire spectrum of the phenotypic variants of ACM was recently published.[1] We present the case of a 17-year-old asymptomatic boy who was referred to our center for an abnormal electrocardiogram: incomplete right bundle branch block, T-wave inversion in all precordial leads, epsilon waves in lead V_1-V_3, and low voltages in inferior leads (**Figure 1A**). Written informed consent has been obtained by the patient to publish this case. Exercise stress test was negative for inducible arrhythmias and 24-h Holter monitoring revealed frequent premature polymorphic ventricular contractions (15,000/24 h). Transthoracic echocardiogram revealed mild left ventricle (LV) dysfunction with a LV ejection fraction of 49%, severe right ventricle (RV) dysfunction with RV fractional area change of 24%, and RV free wall akinesia.

cMRI (cardiac magnetic resonance imaging) was suggestive of biventricular ACM with severe RV impairment and diffuse transmural late gadolinium enhancement in both ventricles, particularly in the RV free wall (**Figure 1B**). ACM was diagnosed with the presence of 4 major criteria.[1] To further characterize the arrhythmic risk, we opted for an electrophysiological study with biventricular 3-dimensional (3D)-electroanatomic mapping (EAM). Unipolar and bipolar Rhythmia (Boston Scientific) 3D-EAM with an Orion mapping catheter demonstrated diffuse scar areas in the RV anterolateral wall and a scar area limited in the LV apex (**Figures 1C and 1D**). Local abnormal ventricular activity signals were recorded in the RV basal free wall and in the LV apex (**Figures 1E and 1F**). No arrhythmias were inducible. After image merging, unipolar and bipolar scar areas at 3D-EAM matched with fibro-fatty replacement areas evidenced by cMRI with good approximation but unipolar

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RV scar areas extended beyond the areas of infiltration at cMRI, indicating wider RV transmural electroanatomic scar than fibrofatty area (Figures 1C and 1D).[2]

The proband was found to be the carrier of a heterozygous mutation in the desmoglein-2 gene. Considering the extent of biventricular ACM detected by cMRI and 3D-EAM, the degree of electrical instability, and the results of genetic screening, a subcutaneous implantable cardioverter defibrillator was implanted. At 1-year follow-up no arrhythmic events were recorded. The combination of MRI and EAM represents a relatively new tool in this setting, which revealed the real structural and electrical extent of the disease combining electroanatomic transmural scar extension with fibro-fatty distribution and guiding therapeutic decisions.

ABBREVIATIONS AND ACRONYMS

3D = 3-dimensional

ACM = arrhythmogenic cardiomyopathy

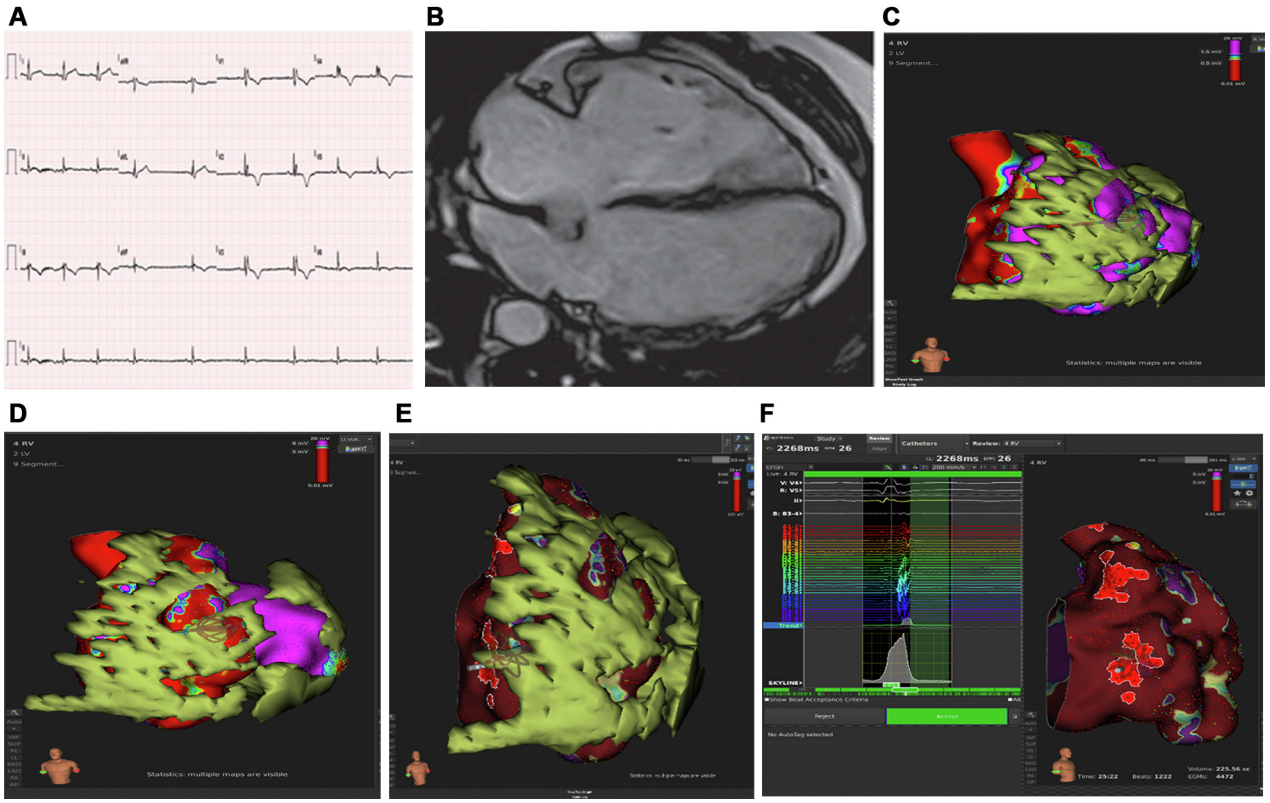
cMRI = cardiac magnetic resonance imaging

EAM = electroanatomic mapping

LV = left ventricle

RV = right ventricle

FIGURE 1 **Electrocardiogram, Cardiac Magnetic Resonance, and Electroanatomic Mapping Features**



(A) Resting electrocardiogram showing diffuse negative T waves in precordial leads and epsilon waves **(red arrows)** in V_1-V_3. Inferior leads with low QRS voltages. **(B)** Cardiac magnetic resonance imaging: severe right ventricle dilation and dysfunction with areas of diffuse transmural late gadolinium enhancement in both ventricles **(black arrowheads)**. **(C)** Merging of biventricular bipolar electroanatomic mapping and cardiac magnetic resonance imaging: low voltages areas in **red** with fibrofatty replacement superimposed in **yellow**, showing diffuse biventricular involvement (scar <1.5 mV). **(D)** Merging of biventricular unipolar electroanatomic mapping and cardiac magnetic resonance: low voltage areas in **red** with fibrofatty replacement superimposed in **yellow**. Compared to bipolar images, unipolar mapping reveals larger areas of diseased myocardium, reflecting the prevalent epicardial involvement in arrhythmogenic cardiomyopathy (ACM). Cardiac magnetic resonance and electroanatomic mapping images are not redundant: encircled in **yellow** areas of electrically diseased myocardium that extend beyond fibro-fatty replacement (scar <5 mV). **(E and F)** Relationship between right ventricular local abnormal ventricular activity **(bright red)**, unipolar electroanatomic mapping and areas of fibro-fatty replacement **(yellow)**.

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KEY WORDS arrhythmogenic ventricular cardiomyopathy, cardiac magnetic resonance, electroanatomic mapping