

## Intracardiac Correlate of the Epsilon Wave in a Patient With Arrhythmogenic Right Ventricular Dysplasia

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A 29-year-old man with no significant past medical history presented to the emergency department of a local hospital with complaints of dyspnea and palpitations. A 12-lead ECG was obtained, revealing a wide complex tachycardia (QRS duration 160 ms) at a rate of 196 beats per minute with a right bundle-branch block morphology and left axis deviation. In the emergency department, the ECG was thought to represent supraventricular tachycardia, and the patient was given adenosine and diltiazem intravenously. This did not terminate or change the rate of the tachycardia. He was then given metoprolol 5 mg intravenously, and the tachycardia slowed and terminated. A 12-lead ECG in normal sinus rhythm is shown (Figure 1).

A transthoracic echocardiogram revealed right atrial enlargement, right ventricular (RV) enlargement, and severe RV dysfunction and left ventricular regional wall motion abnormalities involving the anterior wall and septum. The ejection fraction was 25%. Because of the presence of Q waves on the ECG and regional wall motion abnormalities on the echocardiogram, a cardiac catheterization was performed. Coronary angiography revealed the absence of significant epicardial coronary artery disease. Right heart pressures were normal, and there was no increase in oxygen saturation to suggest an intracardiac shunt.

The patient was transferred to our institution for further management. A 3-catheter electrophysiological study was performed. A catheter positioned in the RV near the septal tricuspid valve leaflet demonstrated a His bundle recording, as well as atrial and ventricular electrograms. Also noted on this catheter recording was a postsystolic or late fractionated electrogram that temporally correlated with the epsilon wave on the surface lead V<sub>1</sub> (Figure 2).

Cardiac magnetic resonance imaging (Figure 3) was performed and demonstrated severe right atrial and RV enlargement and dysfunction with a RV ejection fraction of 9% (see Movie I in the online-only Data Supplement). The left ventricular ejection fraction was calculated to be 34%. The delayed enhanced images demonstrated complete transmural

enhancement of the RV myocardium from the apex to the base extending to the outflow tract. Enhancement was also noted in the basal and anteroseptal areas.

The magnetic resonance imaging findings in conjunction with the other diagnostic studies and clinical findings confirmed the diagnosis of arrhythmogenic RV dysplasia in this patient. The patient underwent implantation of a single-chamber defibrillator. During the implantation, the patient was noted to have poor sensing (R waves of <3 mV) and pacing thresholds despite extensive mapping of the RV, RV outflow tract, and coronary sinus.

Arrhythmogenic RV dysplasia is a rare autosomal dominant disorder that is characterized by structural and functional abnormalities of the right ventricle.<sup>1</sup> Fibrofatty replacement of myocardial tissue results in cardiomyopathy, in electric instability, and ultimately in an increased risk for ventricular arrhythmias and sudden cardiac death. The diagnosis is made by a combination of structural, histological, ECG, arrhythmic, and genetic factors. The presence of delayed enhancement correlates well with endomyocardial biopsy findings and predicts inducibility of sustained monomorphic ventricular tachycardia during electrophysiology testing.<sup>2</sup>

Abnormalities in ventricular depolarization are present on the surface ECG in nearly 90% of patients with arrhythmogenic RV dysplasia.<sup>3</sup> The epsilon wave, a terminal deflection within or at the end of the QRS complex in leads V<sub>1</sub> to V<sub>3</sub>, is present in 30% of patients, and T-wave inversion in the right precordial leads is present in 70%. In this case, the patient had both of these findings on his surface ECG. To our knowledge, correlation of these repolarization abnormalities on the surface ECG with intracardiac electrographic findings has not been described. In this case, we report the intracardiac manifestation of an epsilon wave.

### Disclosures

None.

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The online-only Data Supplement, consisting of movies, is available with this article at <http://circ.ahajournals.org/cgi/content/full/115/21/e538/DC1>.

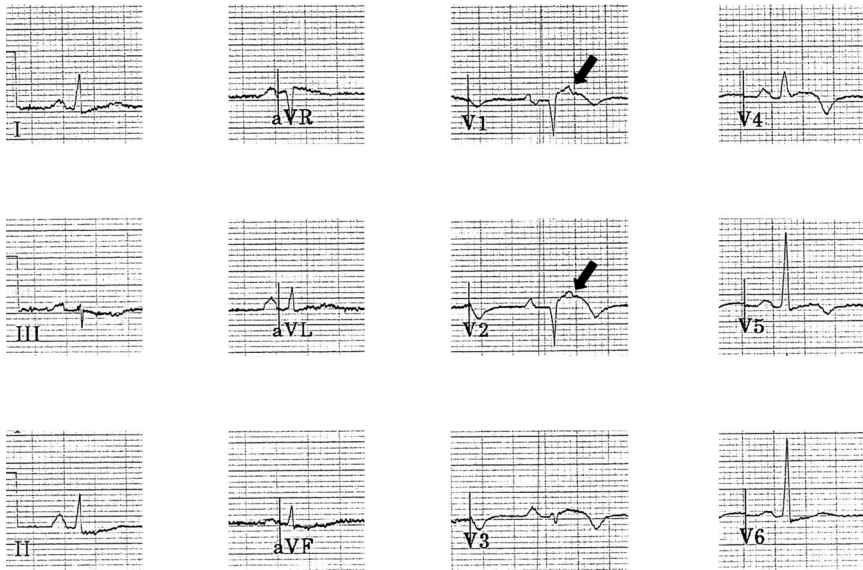
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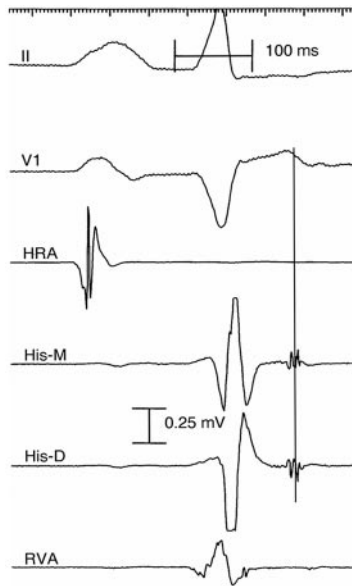
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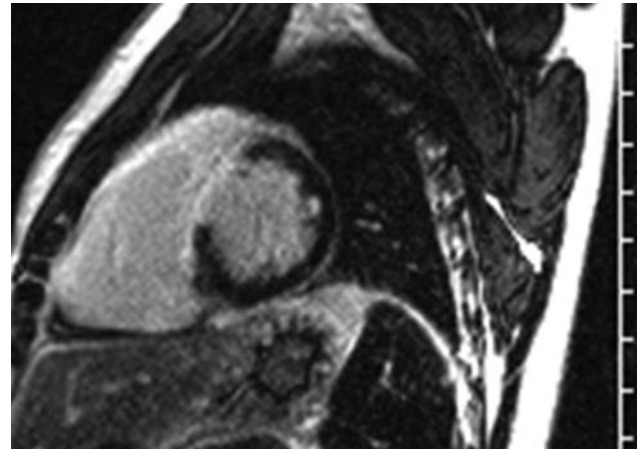
**Figure 1.** Twelve-lead ECG in normal sinus rhythm. The findings include right atrial abnormality, normal QRS axis, anteroseptal Q waves, and precordial T-wave inversions. The most striking finding is the presence of an epsilon wave (black arrow) in V<sub>1</sub> and V<sub>2</sub>.



**Figure 2.** In order from top to bottom, surface leads II and V<sub>1</sub>; intracardiac recordings from the high right atrium (HRA), mid (His-M) and distal His bundle (His-D), and RV apex (RVA). A line is drawn through the epsilon wave on the surface leads and the intracardiac electrographic representation of this late potential. For the intracardiac recordings shown, the notch filter was turned on, the gain was 5000, and the low-pass and high-pass filter limits were set to 500 and 30.0 Hz, respectively.

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**Figure 3.** High-resolution delayed enhanced magnetic resonance image. Notice the hyperenhancement (white) of the entire RV free wall extending into the RV outflow tract and the intraventricular septum. Also note severe RV enlargement. (For a moving image of this view, see Movie II in the online-only Data Supplement).

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