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INTERMEDIATE

CASE REPORT: CLINICAL CASE

Prominent Epsilon Waves in a Patient With Cardiac Sarcoidosis

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ABSTRACT

Epsilon waves are the surface manifestation of myocardial regions with delayed activation and are considered the hallmark of arrhythmogenic right ventricular cardiomyopathy. However, other conditions can also result in epsilon waves and simulate arrhythmogenic right ventricular cardiomyopathy. In this case, a patient presents with recurrent ventricular tachycardia and large epsilon waves due to cardiac sarcoidosis. (**Level of Difficulty: Intermediate.**) (J Am Coll Cardiol Case Rep 2020;2:577-82) © 2020 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/>).

A 40-year-old African American man without significant past medical history except for a recent long flight developed sudden onset of substernal chest pain associated with shortness of breath and dizziness. An electrocardiogram obtained by the emergency medical services documented a ventricular tachycardia at a rate of 188 beats/min with left bundle branch morphology and inferior and rightward axis (**Figure 1**), suggesting origin in the right ventricular (RV) outflow tract. This tachycardia terminated after a bolus of amiodarone, and

the patient was transported to our medical center. On recovering sinus rhythm (**Figure 2**), the electrocardiogram showed right atrial enlargement, low-voltage QRS complexes, and large epsilon waves in all leads, most prominently in right precordial and inferior leads. The echocardiogram showed normal left ventricular (LV) size and systolic function; a dilated RV with reduced systolic function; septal flattening in diastole, suggesting RV volume overload; and moderate tricuspid regurgitation. Echocardiographic findings and the presence of epsilon waves in the electrocardiogram were initially suggestive of arrhythmogenic right ventricular cardiomyopathy (ARVC). A computed tomography scan searching for pulmonary embolism revealed multiple enlarged mediastinal and axillary lymph nodes.

LEARNING OBJECTIVES

- To understand the origin of epsilon waves and their association with regions of delayed ventricular activation.
- To recognize that epsilon waves can occur in other disorders besides ARVC, including cardiac sarcoidosis.

DIFFERENTIAL DIAGNOSIS

Differential diagnosis included ARVC, pulmonary embolism, lymphoma, acute coronary syndrome, and cardiac sarcoidosis.

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The authors attest they are in compliance with human studies committees and animal welfare regulations of the authors' institutions and Food and Drug Administration guidelines, or patient consent where appropriate. For more information, visit the *JACC: Case Reports* [author instructions page](#).

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**ABBREVIATIONS
AND ACRONYMS****ARVC** = arrhythmogenic right
ventricular cardiomyopathy**LV** = left ventricle**RV** = right ventricle**INVESTIGATIONS**

Searching for the etiology of mediastinal and axillary lymphadenopathy, a core biopsy of a left axillary lymph node revealed non-caseating granulomas with negative stain for acid fast bacilli and fungal elements typical of sarcoidosis (Figure 3). The patient had no family history of heart failure, syncope, or sudden death, and genetic testing did not identify any mutations known to be implicated in ARVC.

Cardiac magnetic resonance imaging revealed a decreased LV ejection fraction of 49% and a severely dilated RV with an ejection fraction of 21%. Late gadolinium enhancement disclosed hyperenhancement in the basal portion of interventricular septum and LV free wall near the mitral annulus (Figure 4).

MANAGEMENT

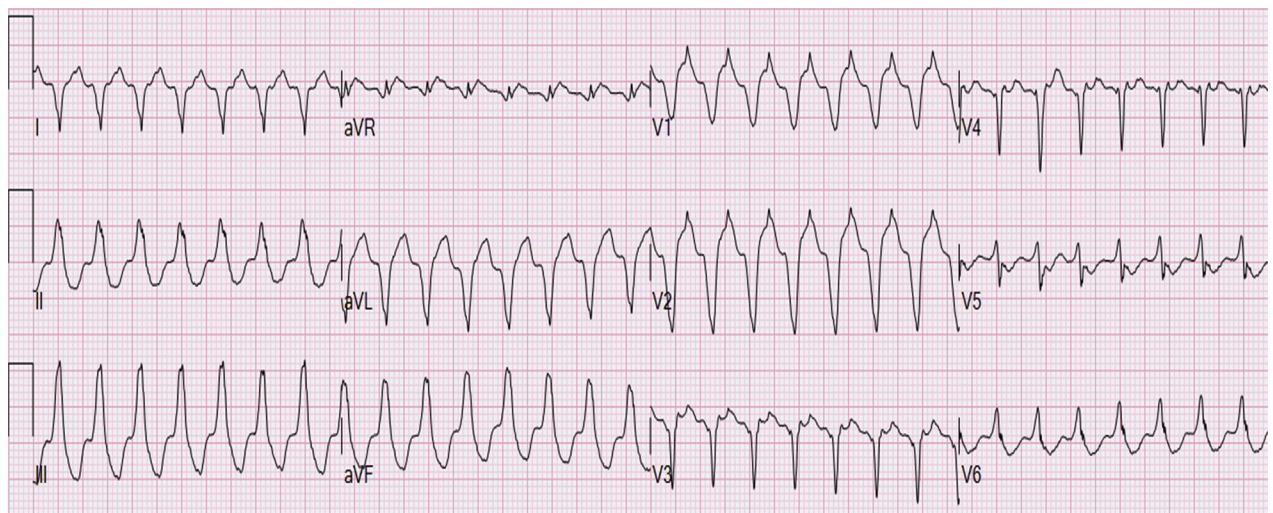
After the initial presentation, the patient had further episodes of ventricular tachycardia resulting in hypotension. During electrophysiology study, the RV was mapped in sinus rhythm using a contact force electrode, and it was found that there was delayed activation of the right free wall and the RV outflow tract (Figure 5). Endocardial late potentials in the right free wall occurred simultaneously with the

epsilon wave recorded in the electrocardiogram (Figure 5).

Ventricular stimulation induced 4 different unstable ventricular tachycardias. Radiofrequency ablation targeting the late potentials during sinus rhythm eliminated the induction of ventricular tachycardia. Following ablation, an implantable cardioverter-defibrillator was implanted, and treatment with prednisone and methotrexate was initiated. A year later, the patient presented with a ventricular tachycardia of different morphology (left bundle, superior axis) than the original tachycardia. An ablation was performed targeting late potentials, rendering the patient noninducible. Amiodarone 100 mg daily was initiated in the setting of an evolving substrate. He remains asymptomatic without recurrence of ventricular tachycardia over the past 3 years.

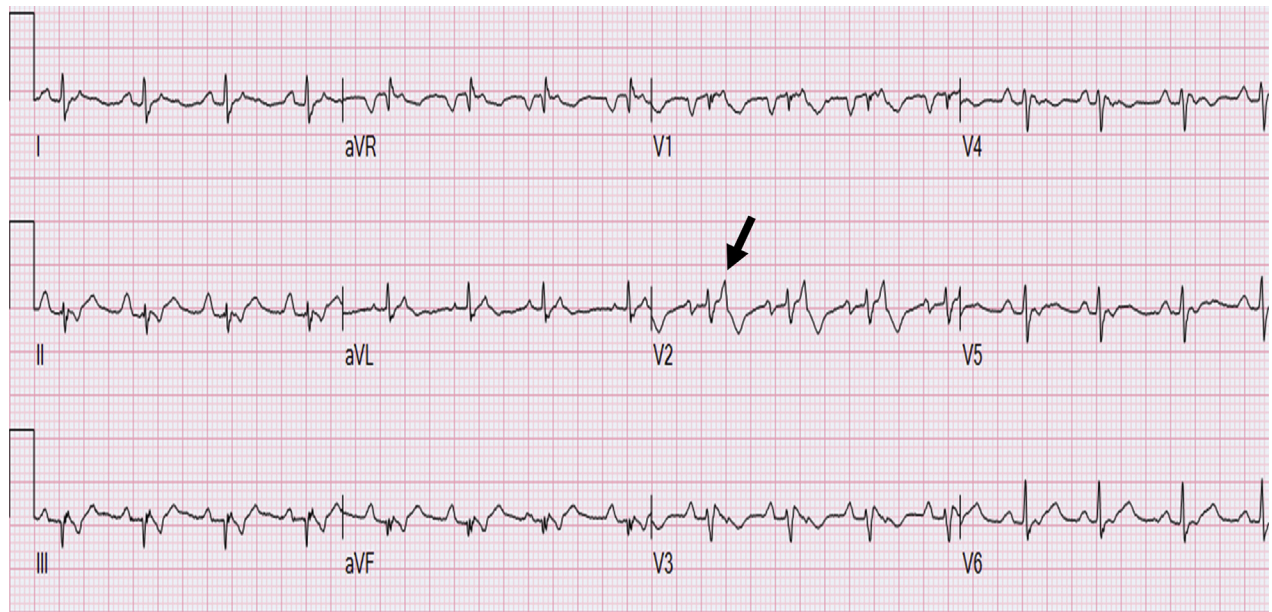
DISCUSSION

Epsilon waves are considered the hallmark of ARVC (1). The term epsilon wave was coined by Dr. Guy Fontaine in 1977 to describe a post-excitation phenomenon in patients with recurrent ventricular tachycardia (2). The epsilon wave is the electrocardiographic manifestation of localized delayed ventricular myocardial activation secondary to myocardial infarction, fibrosis, or infiltrative

FIGURE 1 ECG Obtained at Presentation

A 12-lead electrocardiogram (ECG) obtained at presentation shows ventricular tachycardia with a left bundle branch morphology at a rate of 188 beats/min. The axis in the frontal plane is directed inferiorly and to the right at approximately $+120^\circ$.

FIGURE 2 ECG Obtained in Sinus Rhythm



This 12-lead electrocardiogram (ECG) was obtained in sinus rhythm after termination of the ventricular tachycardia with intravenous amiodarone. Right atrial enlargement and low-voltage QRS complexes are observed in the frontal plane. **Arrow** indicates a prominent epsilon wave in V₂ but can be observed following the QRS complex in all leads.

processes. This wave is inscribed after the end of the QRS complex despite being part of the depolarization process (3). In the present case, endocardial late potentials were recorded in the right free wall occurred simultaneously with the epsilon wave (Figure 5). Epsilon waves can be recorded in patients with other conditions besides ARVC, including ventricular infarction and infiltrative processes. Epsilon waves are a major criteria for the diagnosis of ARVC (4), although they are not pathognomonic and are subject to individual interpretation (5). Sarcoidosis is a systemic disorder that mainly affects the lungs, although necropsy studies find up to 70% of individuals with cardiac involvement (6). Furthermore, in a large registry of 1,140 patients diagnosed as ARVC, 15 patients were found in whom cardiac sarcoidosis and not ARVC was the correct diagnosis (7). Patients with cardiac sarcoidosis tend to be older and more frequently have heart failure, conduction system disease, multiple inducible ventricular tachycardias, involvement of the interventricular septum and LV free wall, and mediastinal lymphadenopathy (6).

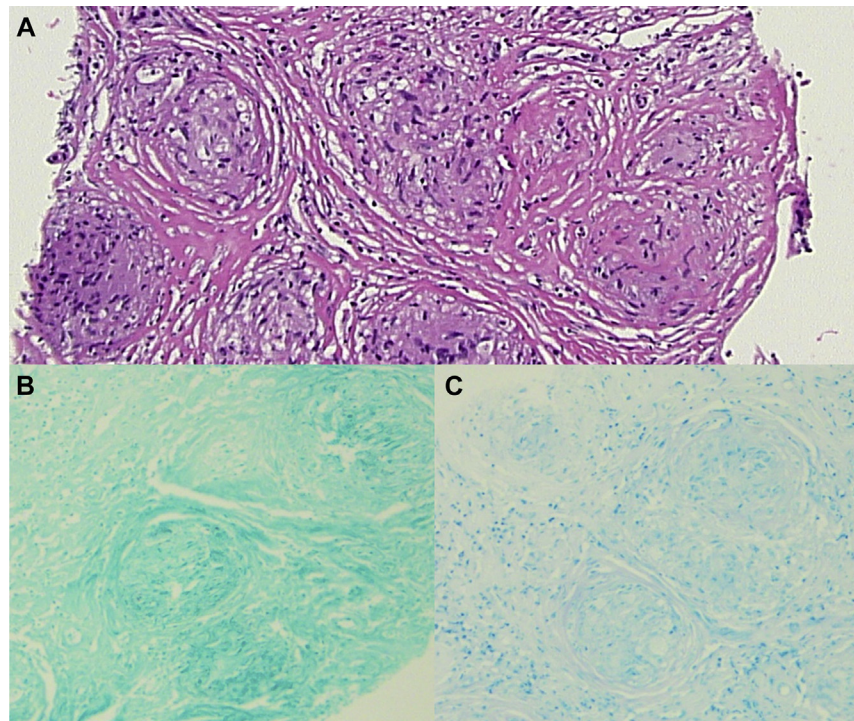
Epsilon waves can be observed both in patients with cardiac sarcoidosis and ARVC (6). Therefore, epsilon waves should not be interpreted as pathognomonic of ARVC. In the present case, the presence of lymph node biopsy-proven sarcoidosis along with the findings in the electrocardiogram, echocardiogram, and cardiac magnetic resonance imaging were used to establish the correct diagnosis (7,8).

FOLLOW-UP

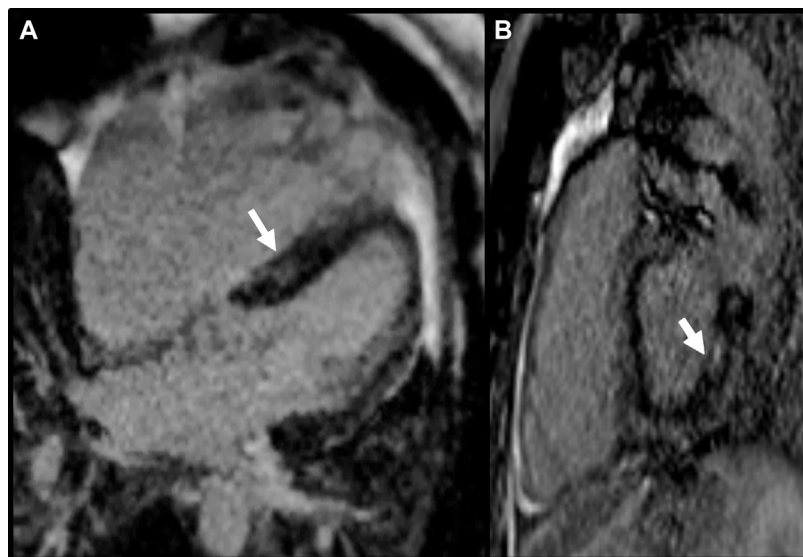
The patient remains asymptomatic 3 years after his second ablation procedure and is maintained on prednisone 2.5 mg daily and amiodarone 100 mg daily without recurrence of ventricular tachycardia.

CONCLUSIONS

Epsilon waves can be seen in advanced forms of ARVC, but they can also be observed in other conditions, as in the present case with cardiac sarcoidosis. In the present case, the combination of

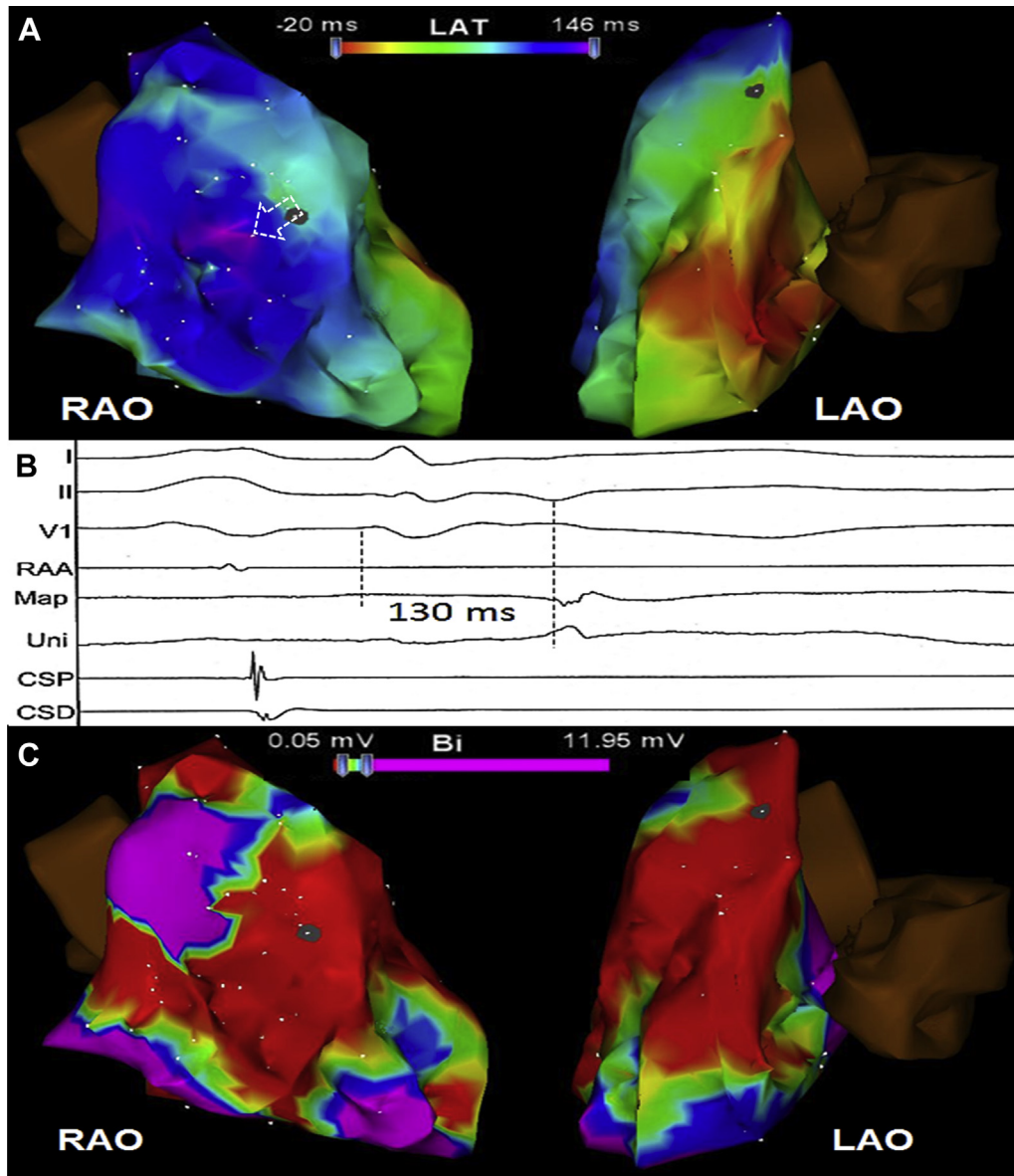
FIGURE 3 Core Biopsy From Axillary Lymph Node

(A) Gram stain demonstrates noncaseating granulomas. (B) Grocott methenamine silver stain shows absence of fungal elements. (C) Ziehl-Neelsen stain was negative for acid fast bacilli.

FIGURE 4 Cardiac Magnetic Resonance

Cardiac magnetic resonance imaging shows (A) 4-chamber spin echo with fat suppression and (B) short-axis spin echo with fat suppression. This study demonstrated decreased wall thickness in the left ventricular free wall and delayed gadolinium enhancement in the interventricular septum (arrow in A) and left ventricle free wall (arrow in B).

FIGURE 5 Voltage Map of Right Ventricle in Sinus Rhythm



(A) Sequence of endocardial activation of the right ventricle obtained in sinus rhythm using a contact force mapping catheter and a non-fluoroscopic mapping system (Carto 3, Biosense Webster, Irvine, California). Late activation (**white dotted arrow**) occurs in the right ventricular free wall. At this site, late potentials occurring simultaneously with the epsilon wave were recorded (**B**). Bipolar mapping showed extensive areas with low voltage in the right free wall and right ventricular outflow tract (**C**). Bi = biventricular; CSD = coronary sinus distal; CSP = coronary sinus proximal; LAO = left anterolateral projection; LAT = lateral; RAA = right atrial appendage; RAO = right anterolateral projection.

immunosuppressive drugs, catheter ablation, and antiarrhythmic drugs may be required to halt or delay disease progression. Despite apparently successful ablation of ventricular tachycardias, an implantable cardioverter-defibrillator is needed in most patients due to a high risk of recurrence of these arrhythmias.

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KEY WORDS arrhythmogenic right ventricular cardiomyopathy, cardiac sarcoidosis, epsilon waves