CASE REPORT

The Presence of Epsilon Waves in All Precordial Leads (V₁–V₆) in a 13-Year-Old Boy with Arrhythmogenic Right Ventricular Dysplasia (ARVD)

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Electrocardiographic feature is included in the diagnostic criteria for arrhythmogenic right ventricular dysplasia (ARVD) based on the Revised Task Force criteria 2010. The epsilon wave, which reflects delayed conduction of the right ventricle, is considered to be one of the major diagnostic criteria. We reported a 13-year-old Thai boy with ARVD who presented with ventricular tachycardia. The presence of epsilon wave in all precordial leads (V₁–V₆) was observed in standard 12-lead EKG. Extensive scarring of the right and left ventricle was seen on cardiac MRI. The extensive Epsilon wave found in this patient may reflect the extensive ventricular wall involvement.

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Arrhythmogenic right ventricular dysplasia (ARVD) is a genetically inherited cardiomyopathy, which characterizes by replacement of right ventricular myocardium with fibrofatty tissue. Abnormal electrical conduction leads to generation of reentrant ventricular tachycardia (VT). ARVD is diagnosed by using the Revised Task Force criteria 2010, which were based on structural alteration, tissue characterization, repolarization-depolarization/conduction abnormalities, arrhythmias, and family history.¹ Definite diagnosis is made by completing two major criteria, or one major and two minor criteria, or four minor criteria from different categories. One of the major criteria in the diagnosis of ARVD is the presence of epsilon wave in right precordium leads (V₁–V₃). We reported a case of a 13-year-old boy with epsilon waves in all precordial leads (V₁–V₆)

associated with extensive scarring of the right and left ventricle.

A 13-year-old boy presented with palpitation, central chest tightness, and hypotension (BP 80/40 mmHg). Initial 12-lead EKG showed monomorphic VT (rate 160/min) with left-bundle branch block and left-axis morphology. A synchronized cardioversion (100 J) successfully converted the VT to normal sinus rhythm. The 12-lead EKG after VT termination showed sinus rhythm (rate 64/min) with generalized low voltage, right atrial enlargement, and right-bundle branch block. Epsilon waves and T-wave inversion were seen across all precordial leads (V₁–V₆; Fig. 1). Transthoracic echocardiography showed enlarged right atrium and right ventricle (RV) with moderate tricuspid regurgitation. Cardiac MRI documented severe RV dilatation (end-diastolic volume = 378.7 mL/m²) with impaired systolic function (RV ejection fraction = 13.1%) and diffused myocardial thinning

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Figure 1. Twelve-lead EKG demonstrated epsilon waves in V_1 – V_6 (marked by arrow).

Figure 2. Upper row: SSFP cine white blood cardiac MR imaging in end-systolic phase of short axis (A), axial (B), and right ventricular inflow tract views (C) showed severe dilatation of the right ventricle and small aneurysms at right ventricular wall (arrows). Lower row: delayed gadolinium enhancement imaging in axial (D) and short-axis views (E) showed diffuse delayed enhancement at the right ventricular wall and at apicolateral wall of the left ventricle, indicating myocardial fibrosis (arrowheads).

and delayed gadolinium enhancement of the RV compatible with fibrosis (Fig. 2). Mildly decreased left ventricular systolic function ($EF = 42.5\%$) with delayed enhancement was also seen. Both echocardiogram and cardiac MRI were compatible with the diagnosis of ARVD according to Revised Task Force Criteria for ARVD/C 2010.¹

Guy Fontaine first mentioned the epsilon wave in a patient with ARVD in 1977.² It is a reproducible low-amplitude signal between the end of QRS complex and the onset of T wave. Although it is one of the major criteria for diagnosis of ARVD, the sensitivity of epsilon wave for the diagnosis of ARVD is low, observed in only 1–29% of cases according to previous studies.³ Epsilon wave represents delayed activation of affected ventricular myocardial fibers after the excitation of main ventricle, also known as postexcitation wave. These localized prolongation of the end of QRS complexes (>110 ms) are best seen in right precordial leads (V_1 – V_3). The epsilon waves represent selective involvement of RV myocardium in ARVD patients; however, they can rarely be seen in posterior and right ventricular myocardial infarction, right ventricular hypertrophy, and infiltrative diseases.⁴

From the previous studies, the epsilon wave proved to be related with the extension of ventricular structural and functional abnormalities. It is also correlated with the progression of ARVD, but not correlated with high-risk ARVD (patient with VT and/or RV failure).⁵ In our patient, the initial EKG showed epsilon waves in all precordial leads (V_1 – V_6). It is best explained by the extensive involvement of both right and left ventricular walls in this patient as seen on the cardiac MRI.

The patient underwent an electrophysiologic study, which showed inducible two morphologies

of VT (rate 160/min). RF ablation could not be done due to hemodynamic instability during the VT. The patient later underwent implantable cardioverter/defibrillator (ICD) implantation and was given amiodarone. There was no recurrence of VT or ICD discharge during the 4-month follow-up.

REFERENCES

1. Marcus FI, McKenna WJ, Sherrill D, et al. Diagnosis of arrhythmogenic right ventricular cardiomyopathy/dysplasia. *Eur Heart J* 2010;31:806–814.
2. Wang J, Yang B, Chen H, et al. Epsilon waves detected by various electrocardiographic recording methods in patients with arrhythmogenic right ventricular cardiomyopathy. *Tex Heart Inst J* 2010;37:405–411.
3. Dalal D, Nasir K, Bomma C, et al. Arrhythmogenic right ventricular dysplasia: A United States experience. *Circulation* 2005;112:3823–3832.
4. Marcus FI, Zareba W. The electrocardiogram in right ventricular cardiomyopathy/dysplasia. How can the electrocardiogram assist in understanding the pathologic and functional changes of the heart in this disease? *J Electrocardiol* 2009;42:136.e1–136.e5.
5. Wu S, Wang P, Hou Y, Yang P, Xiao Y, Zhan X. Epsilon wave in arrhythmogenic right ventricular dysplasia/cardiomyopathy. *Pacing Clin Electrophysiol* 2009;32:59–63.