New diagnosis of arrhythmogenic right ventricular cardiomyopathy in an octogenarian with the help of Fontaine electrocardiographic leads

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Received 31 December 2009; accepted after revision 5 March 2010

We report an 85-year-old man with arrhythmogenic right ventricular cardiomyopathy who presented with monomorphic ventricular tachycardia. This is the oldest patient recorded with this disease. The presence of epsilon waves by the Fontaine lead system provided a high degree of suspicion for the disease.

Introduction
Early and accurate diagnosis of arrhythmogenic right ventricular cardiomyopathy (ARVC) is challenging because of its variable clinical presentation, and often minor identifiable abnormalities.1 Only sparse reports exist documenting atypical features of antemortem ARVC in very elderly patients. Our case underscores the contribution of epsilon waves, which became readily identifiable only by utilizing Fontaine precordial lead positions, as an aid to the diagnosis of ARVC.

Case report
An 85-year-old man presented with sustained ventricular tachycardia of left bundle branch block configuration with left-axis deviation which was treated with electrical cardioversion. Following restoration of sinus rhythm, the conventional 12-lead electrocardiogram (ECG) demonstrated sinus rhythm with complete right bundle branch block (RBBB). Suspicious epsilon waves just after the QRS complex were occasionally visible in leads I, II, aVR, and V2, only when the ECG was taken at increased sensitivity of 20 mm/mV with filter setting 40 Hz (Figure 1). Repeated ventricular tachycardia episodes were depressed by intravenous and concomitant oral amiodarone. The patient’s medical history was unremarkable and he did not report any family history of syncope or premature/sudden death. Coronary angiography revealed non-significant coronary artery disease, and left ventriculography showed normal systolic function. Transthoracic echocardiography, right ventricular angiography, and magnetic resonance imaging were suggestive of ARVC showing moderately impaired right ventricular contraction, infundibular dysplasia and localized sacculation at the posterior wall. Both structural alterations, subpulmonary and posterior, showed systolic contraction without evidence of akinetic or dyskinetic areas typical for aneurysms. (Figure 1). With Fontaine ECG, readily identifiable epsilon waves were reproducibly documented in leads II and III (Figure 2). Our patient fulfilled the requirements of the proposed Task Force criteria for diagnosing ARVC,2 owing to the fact that he presented at least one major (epsilon waves) along with two minor criteria (ventricular tachycardia, mild global right ventricular dilatation with ejection fraction reduction). Programmed right ventricular stimulation induced two types of sustained ventricular

Figure 1 (Left panel) Conventional 12-lead ECG showing sinus rhythm with right bundle branch block. Epsilon waves (arrows) appear transiently in leads I, II, aVR, and are barely perceptible in V2. Recording 25 mm/s, sensitivity 20 mm/mV. (Right panel) Right ventricular angiography at 30° right anterior oblique projection showed infundibular dysplasia and sacculation at the posterior wall (arrows) with systolic contraction.
tachycardia of left bundle branch block morphology, with right- and left-axis deviation. An implantable cardioverter-defibrillator was placed before hospital discharge.

Discussion

The diagnosis of ARVC is still based on the presence of major and minor standardized Task Force Criteria which combine information from several diagnostic tests encompassing electrocardiographic, arrhythmic, right ventricular morphofunctional, histopathological, and clinical genetic factors. However, since clinical and cardiac imaging tools often are not sufficiently sensitive or specific to diagnose early ARVC, there is major difficulty in detecting minor forms of the disease in poorly symptomatic patients or atypical presentations.

The prevalence of the typical but hardly detectable epsilon wave by the standard ECG constitutes one of the two major diagnostic ECG criteria for ARVC. Epsilon waves are not specific for ARVC since a relationship to right ventricular abnormality has also been described in association with right ventricular infarction or sarcoidosis. Nevertheless, these discrete potentials are recognized as a hallmark feature of ARVC reflecting the delay excitation in islands of surviving myocytes which are interspersed with fibrous and fatty tissue. When examining the patient’s routine ECG, the recognition of epsilon waves is even more important in the presence of RBBB, where the other major ECG criterion, namely the QRS duration > 110 ms, cannot be appreciated. Typically, the epsilon waves may be most easily identified in the ECG leads V1 to V3 as well as in the right precordial leads by doubling the sensitivity of the record, and use of filter setting 40 Hz instead of 150 Hz to decrease the noise level. The bipolar Fontaine precordial electrocardiographic leads I–III may be also used to enhance the recording of epsilon waves by applying placement of the right arm electrode on the manubrium, of the left arm electrode on the xiphoid, and of the left leg lead in V4. Sensitivity of epsilon potentials waves by using the Fontaine lead positions can be accentuated when the ECG is taken at double paper speed and sensitivity and filter setting 40 Hz.

In the present patient, the classical epsilon waves were not identifiable by the normal routine ECG recording technique, even with high amplification of the recording, were hardly distinguishable within the QRS complex in lead V2, were not a consistent finding in leads I, II, and aVR, but were readily and reproducibly detectable in the Fontaine leads II and III. This suggest that epsilon waves may be more frequent than is thought and may be unmasked by the application of Fontaine leads. The Fontaine ECG may be particularly helpful to unmask epsilon waves in distinguishing uncertain cases of early AVRC presenting with RBBB.

Conflict of interest: none declared.

References