

Electrocardiographic Features in the Diagnosis and Risk Assessment of Arrhythmogenic Right Ventricular Cardiomyopathy/Dysplasia

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ABSTRACT

There are numerous ECG features making the diagnosis of arrhythmogenic right ventricular cardiomyopathy/dysplasia possible such as localised right precordial QRS prolongation, right precordial QRS prolongation, terminal activation delay, S wave upstroke, epsilon waves, QRS fragmentation, reduced amplitude in precordial leads, reduced amplitude ratio in right precordial leads and more than complete right bundle branch block. What is new are large Q waves, small R waves and negative T waves in lead aVR and epsilon waves in lead aVR as a risk marker of heart failure. Early repolarisation phenomenon appears in 24% of cases, but it is not clear to decide, whether it is a hint of inferior aneurysm or a sign of recurrent ventricular tachycardia. Atrial fibrillation appears either early in the disease progression or very late in intensive form of the disease.

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Key words: Localised right precordial QRS prolongation; Epsilon waves; QRS fragmentation; Eight precordial T-wave

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REVIEW

Arrhythmogenic (right ventricular) cardiomyopathy is defined as electroanatomical scar due to fibrofatty degeneration and myocardial atrophy. This can be best reproduced by voltage mapping^[1]. Myocardial atrophy is defined as loss of myocardial cells of 40% or more in the latest paper of diagnostic criteria published of Marcus and coworkers^[2].

The conduction of the right ventricle is slowed due to fibrofatty abnormalities leading to cell-to-cell conduction delay. Even if the dominant finding is arrhythmogenic left ventricular cardiomyopathy with leading abnormalities mainly of the left ventricle right ventricular conduction delay is still evident.

The main characteristics of arrhythmogenic cardiomyopathy are: (1) electroanatomical scar; (2) myocardial atrophy; (3) conduction delay of the right ventricle.

All these characteristics can be reproduced by standard ECG making ECG the best diagnostic option for the diagnosis of arrhythmogenic cardiomyopathy.

Conduction delay of the right ventricle is indeed best diagnosed by standard ECG. There are many electrocardiographic characteristics which describe conduction delay. If the speed of ECG writing is 50 mm/s - which is unusual the most countries except in Germany and Austria - a difference between QRS duration in right precordial and left precordial leads of more than 1.2 can be defined. If QRS duration in right precordial leads is 100 ms or more the most accurate formula is $QRS \text{ duration in } (V1+V2+V3) / (V4-V5-V6)$ is 1.2 or more^[3].

Other options are right precordial QRS duration $\geq 110 \text{ ms}^{[4]}$, terminal activation delay $\geq 55 \text{ ms}^{[5]}$ and S-wave upstroke duration in $V1 \geq 55 \text{ ms}^{[6]}$.

A very good option is describe arrhythmogenic cardiomyopathy by ECG is the appearance of so-called epsilon waves^[2,7]. Epsilon waves are major criteria in the definition of arrhythmogenic cardiomyopathy very well characterising conduction delay of the right ventricle. Another clue for ECG definition is QRS fragmentation in all leads including “pre-silons” (QRS fragmentation in the Q wave), “top-silons” (QRS fragmentation in the R wave) and “post-silons” (QRS fragmentation in the S wave) reported by G.Fontaine and published in diverse literature^[8]. QRS fragmentation of right precordial leads is probably the same ECG phenomenon as epsilon waves. QRS fragmentation is of diagnostic value in arrhythmogenic cardiomyopathy^[9] and has prognostic effects for risk stratification^[10]. QRS fragmentation is an unspecific finding in arrhythmogenic cardiomyopathy and can also be found in diverse entities like coronary artery disease, Ebstein anomaly, Brugada syndrome, dilated cardiomyopathy and hypertrophic cardiomyopathy^[11].

In some ECG's in arrhythmogenic cardiomyopathy the amplitude of precordial leads are reduced^[12] and by the formula of QRS amplitude ratio in $(V1+V2+V3) / (V1+V2+V3+V4+V5+V6) \leq 0.48$ risk assessment is possible^[13].

In the presence of complete right bundle branch block with QRS prolongation in right precordial leads – defined as “more than” complete right bundle branch block^[14] – arrhythmogenic cardiomyopathy can be diagnosed by conduction delay. This ECG criterion can be seen in arrhythmogenic cardiomyopathy, cardiac sarcoidosis superimposed on arrhythmogenic cardiomyopathy and arrhythmogenic biventricular cardiomyopathy with progressive severe left ventricular impairment sometimes leading to heart transplantation. The value of electrocardiogram as a predictor of right ventricular dysfunction in patients with chronic right ventricular volume overload can be best characterised by a QRS width of 140 ms or more^[15].

To sum up ECG characteristics of conduction delay there are diverse ECG features such as: (1) localised right precordial QRS prolongation; (2) right precordial QRS prolongation; (3) terminal

activation delay; (4) S wave upstroke; (5) epsilon waves; (6) QRS fragmentation; (7) reduced amplitude in precordial leads; (8) reduced amplitude ratio in right ventricular leads; (9) more than complete right bundle branch block; (10) QRS width of 140 ms or more.

Electroanatomic scar and myocardial atrophy are other characteristics of arrhythmogenic cardiomyopathy. These features can be best interpreted in lead aVR. Lead aVR is the only lead which is completely directed to the right ventricle. As it is an unipolar lead the findings in arrhythmogenic cardiomyopathy can also be found in some cases (18.9%) in healthy volunteers. The statistical significance of large Q wave of 3mm or more, small R wave of 2mm or less and inverted T waves are very high; negative predictive value is nearly 100% thus excluding arrhythmogenic cardiomyopathy if other ECG criteria are positive^[16]. Large Q wave characterises electroanatomic scar; small R wave characterises myocardial atrophy.

An epsilon wave in lead aVR is rarely seen (mostly in patients with complete right bundle branch block). The mortality is high due to heart failure^[17] or - to a lesser extent – arrhythmic events due to for example electrical storm^[18].

T wave inversions in right precordial leads are characteristic, but completely unspecific findings which are in the first report of diagnostic criteria published by William McKenna and co-workers^[4] a minor criterion but in the second report a major finding^[2]. In combination with other diagnostic ECG criteria the significance is high. The appearance of recurrent ventricular tachycardia or ventricular fibrillation can be best predicted by the extent of T-wave inversions including inferior or lateral leads according to the extent of electroanatomic scar in voltage mapping^[19,20].

From a not yet published paper the amplitude of T wave in lead V1 of 3mm or more appears to have a specificity of 97%.

The association between arrhythmogenic cardiomyopathy and Brugada syndrome is ongoing debate. In some patients with definite arrhythmogenic cardiomyopathy ajmaline challenge leads to coved-type ST-segment elevation in right precordial leads. Recently ECG

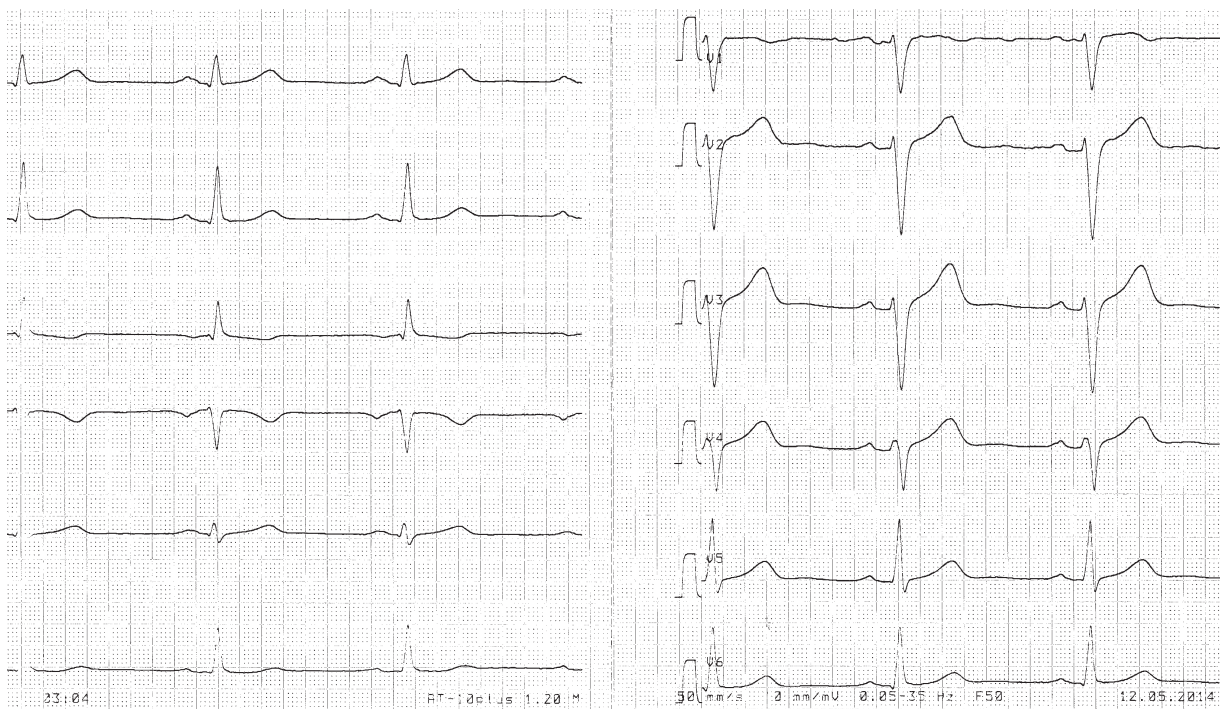


Figure 1 ECG of a patient with dominant arrhythmogenic left ventricular cardiomyopathy. Left side: lead I, II, III, aVR, aVL, aVF (from above); right side: lead V1, V2, V3, V4, V5, V6 (from above).

criteria to differentiate “true” and “false” brugada syndrome were published^[21,22] including: (1) QRS-ST at least 2mm high in lead V1; (2) the duration of the QRS in leads V1 and V2 is greater than in the middle and left precordial leads; (3) QRS morphology shows progressive decline; (4) low QRS amplitude after ajmaline testing.

If these criteria are positive, “true” Brugada syndrome is likely. In a cohort of 19 patients with typical arrhythmogenic cardiomyopathy and provokable coved-type ST elevation in right precordial leads in 16 patients (84%) “true” Brugada syndrome could be suggested^[23]. In 3 patients “false” brugada syndrome probably resulted from maldevelopment of the right ventricular outflow tract^[24].

In a few cases with arrhythmogenic left ventricular cardiomyopathy criteria of right ventricular conduction delay dominants ECG appearance. We describe a 32-year old male patient with syncope. Although the right ventricle showed no echocardiographic abnormalities the ECG revealed signs of conduction delay of the right ventricle (Figure 1).

It is well known that additional left ventricular involvement increases the risk of arrhythmic events^[25]. An electrocardiographic marker of additional left ventricular involvement is left precordial JT prolongation of 30ms or more. With the help of this simple ECG criterion it was possible to predict arrhythmic risk^[26].

In about 24% of cases typical ECG criteria of arrhythmogenic cardiomyopathy were associated with early repolarization phenomenon

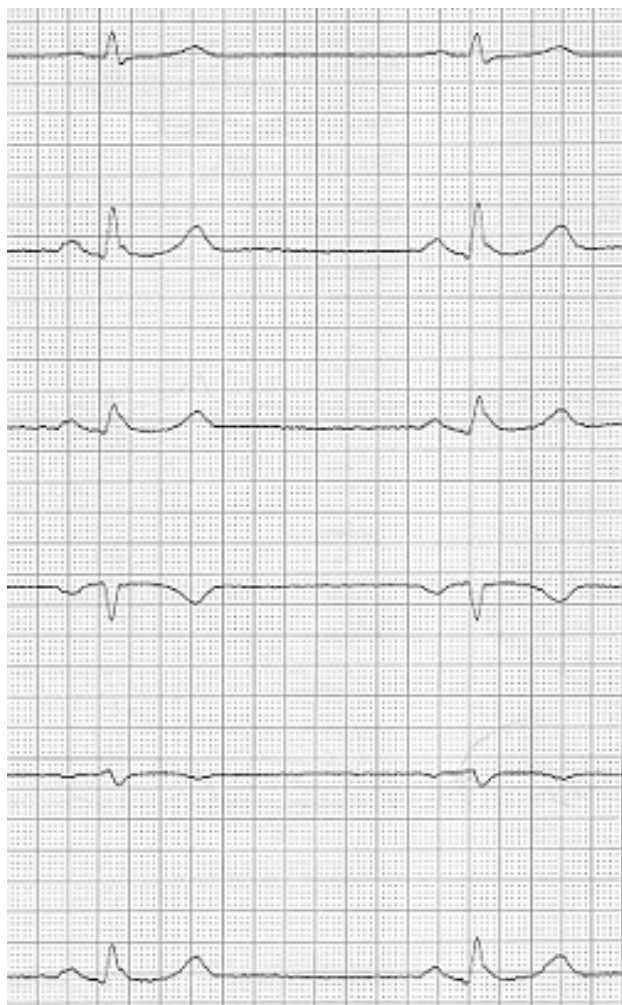


Figure 2 Early repolarization phenomenon in a patient with typical arrhythmogenic cardiomyopathy. Lead I, II, III, aVR, aVL, aVF (from above).

(Figure 2). This is so far not a marker of arrhythmic risk, but a hint of inferior aneurysm in the development of arrhythmogenic cardiomyopathy^[27]. Recently, Chen and coworkers described early repolarization as a marker of sustained ventricular tachycardia best treated with epicardial ablation^[28].

Finally, atrial fibrillation before ventricular arrhythmias^[29] appear and in the long-term follow-up^[30] can be documented in nearly 40% of cases. Atrioventricular block and the necessary of pacing appear in up to 6% of cases^[31] mostly in association with fibrofatty infiltration of the AV node and His bundle.

CONFLICT OF INTERESTS

There are no conflicts of interest with regard to the present study.

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