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Electroanatomic Scar and Myocardial Atrophy in Arrhythmogenic Cardiomyopathy - Review of ECG Criteria

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Keywords: Arrhythmogenic cardiomyopathy; Electroanatomic scar; Myocardial atrophy; Terminal activation delay; T-wave inversions.

Abstract

Introduction: Electroanatomic scar and myocardial atrophy is the predominant finding of arrhythmogenic cardiomyopathy in either dominant right ventricular, biventricular of dominant left ventricular abnormalities. Lead aVR can describe electroanatomic scar by large Q waves and myocardial atrophy by small R waves. With these two conditions T wave is predominantly negative </= 2mm or in few cases biphasic.

Method: A large collective of patients 487 patients (309 males, mean age 42.2 ± 12.8 years) including own patients (436 cases), heart transplantation candidates with desmoglein-2 and plakophilin-2 mutations (41 cases), a large family (6 cases) with non compaction left ventricle and typical histologic arrhythmogenic cardiomyopathy of the right ventricle in FHL1 mutation, and 4 cases with filamin C mutations were analysed by right precordial negative T waves, epsilon waves, localised right precordial QRS prolongation, terminal activation delay, low voltage in limb leads, inferolateral T-inversions or flattening and typical findings in lead aVR.

Results: The most common electrocardiographic findings were large Q waves and small R waves in lead aVR defining electroanatomic scar and myocardial atrophy (100%). Localised right precordial QRS prolongation and terminal activation delay were found in 98%. Right precordial T-wave inversions were present of 69% of cases and epsilon waves in 30% of cases. In 7 cases low voltage in limb leads and inferolateral negative or flattened T waves were present. In the large family with FHL1 mutation and in two cases of the own patients only typical ECG signs in lead aVR were present.

Conclusion: The most common electrocardiographic finding in arrhythmogenic cardiomyopathy are large Q waves and small R waves in lead aVR closely followed by localised right precordial QRS prolongation and terminal activation delay.

These two criteria should belong to major diagnostic criteria for the electrocardiographic diagnosis of arrhythmogenic cardiomyopathy.

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Introduction

ECG criteria in arrhythmogenic cardiomyopathy-either right dominant or left dominant-are an essential diagnostic tool on the way to non invasive diagnosis. According to recommendations in 1994 [1] major diagnostic criteria were right precordial QRS prolongation and epsilon waves; minor criteria were right precordial T-wave inversions and typical findings of signal-averaged ECG.

In 2010 a modification of diagnostic criteria was published [2] defining right precordial T-wave inversions and epsilon waves as major diagnostic criteria, and terminal activation delay as a minor diagnostic criterium.

Last but not least, the so-called Padua criteria [3] excluded epsilon waves as major criteria and added low voltage in limb leads as minor criteria.

In 2014 typical findings in lead aVR with large Q waves as electroanatomic scar, small R waves as myocardial atrophy were added [4] and slighty inverted (< 2mm) or biphasic T-waves [5].

In a large collective of patients a variety of diagnostic ECG criteria were analysed again.

Matrial and Methods

In a total collective of 487 patients (309 males, mean age 42.2±12.8 years) including own patients (436 cases), transplantation candidates with desmoglein-2 and plakophilin-2 mutations (41 cases), six patients with FHL1 (Emory-Dreifuss) mutation, and four patients with filamin-C mutations several ECG findings were analysed including:

Right precordial T-waves inversions

right precordial epsilon waves

localised right precordial QRS prolongation

terminal activation delay

low voltage in limb leads

inferolateral T-inversions or flattening

and typical findings in lead aVR (large Q waves, small R-waves, and negative [< 2mm] or biphasic T-waves).

These ECG criteria were listed in percentage and correlated to current literature.

Results

Signs of electroanatomic scar and myocardial atrophy in lead aVR could be found in all patients (100%) independent of the aspect of arrhythmogenic cardiomyopathy-right dominant, left dominant or biventicular. Interestingly, the form of T wave was strictly negative (< 2mm) in right dominant or biphasic in left dominant cases.

Localised right precordial QRS prolongation and terminal activation delay combining right precordial QRS prolongation and epsilon waves were found in 479 cases (98%).

Right precordial T-wave inversions were found in 336 cases (69%).

Epsilon waves could be found in 146 cases (30%).

Low voltage in limb leads could be found in all patients with arrhythmogenic left ventricular cardiomyopathy (7 cases) and in 58 cases with biventricular disease (13%).

Inferolateral T-wave inversions of flattening were found in seven cases with arrhythmogenic left ventricular cardiomyopathy (1.4%).

Discussion

After extension of known ECG criteria beyond the so-called Pudua criteria it is a very interesting phemonenon that signs of electroanatomic scar and myocardial atrophy in lead aVR can be seen in all patients (100%) in this collective of patients with arrhythmogenic cardiomyopathy either right dominant, biventricular or (in seldom cases) left dominant. In particular, in left dominant cases ECG signs of large Q waves and small R waves in all cases is a very special finding, as lead aVR is completely directed to the right ventricle. The only different finding was that T-wave was in some cases biphasic. The special role of lead aVR was first described in 2014 [4] and added with regard to T waves [5].

The second most frequent finding was localized right precordial QRS prolongation and terminal activition delay combining QRS prolongation and epsilon waves. In the first paper published in 1994 [1] right precordial QRS prolongation was defined as major criterium regardless of ECG paper speed (25 mm/sec or 50 mm/sec). In the definition of localized right precordial QRS prolongation a ECG paper speed of 50 mm/sec is mandatory correlating right precordial and left precordial QRS complex width.

Terminal activation delay is mentioned in the review paper published in 2010 [2] and defined as minor diagnostic criterium.

Interestingly, these two criteria mentioned above are not included in the review of 2010 on diagnostic criteria and should belong to major diagnostic ECG criteria.

Right ventricular sarcoidosis is the only disease mimicking arrhythmogenic cardiomyopathy with all ECG features mentioned in this text. Fibrofatty infiltration are replaced by non-caseating necrosis and granuloma, the pathomechanism is completely identical.

Right precordial T-wave inversions and epsilon waves were defined as major diagnostic criteria in the review in 2010. In this large collective T-wave inversions and epsilon waves can be found in 69% and 30%, respectively, the definition as major diagnostic criteria must be questioned. Meanwhile, epsilon waves are devalueated as minor diagnostic criteria according to Padua criteria [3].

Low voltage in limb leads and T-wave inversions in inferolateral leads are effective, reliable diagnostic markers of arrhythmogenic left ventricular cardiomyopathy together with typical MRI findings.

In summary, special ECG criteria have a great impact on diagnosis of arrhythmogenic cardiomyopathy in right dominant forms and in the differentiation of right dominant and left dominant arrhythmogenic cardiomyopathy.

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