Editorial

Concealed electrocardiographic features in arrhythmogenic cardiomyopathy

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Introduction

Arrhythmogenic cardiomyopathy is a life-threatening primary electrical disease with the potential of sudden cardiac death at the first manifestation of the disease. That is why it is very important to identify the disease as early as possible.

Standard ECG may describe arrhythmogenic cardiomyopathy with different markers:

- T-wave inversions as a marker of right ventricular dilatation [1].
- Epsilon waves as a marker of pronounced right ventricular outflow tract involvement [2].
- QRS fragmentation in inferior leads as a marker of inferior wall involvement [3].
- Localized right ventricular QRS prolongation as a marker of parietal block [4].
- Low voltage as a marker of advanced disease, in most cases caused by phospholamban mutations [1].

In the meantime, several ECG abnormalities have been described as features of concealed arrhythmogenic cardiomyopathy at the very early stage of the disease (prehistologic phase).

S-wave angle in lead V2

Daniel Cortez first described the S-wave angle in lead V2 as an early presentation of arrhythmogenic cardiomyopathy controlled by 66 probands without heart disease [5]. The first measurements were done by a protractor. After the initial phase, ECG’s were digitalized and analyzed. At a cut-off of 12.5°, most patients could be identified as having arrhythmogenic cardiomyopathy with a sensitivity of 47%, a specificity of 97%, and a negative predictive value of 65%.

Typical abnormalities in lead aVR

Stefan Peters described typical abnormalities in lead aVR in the ECG of patients with arrhythmogenic cardiomyopathy controlled by 1496 probands without heart disease [6]:

- Large (≥3 mm) Q wave as a marker of electroanatomic scar,
- Small (≤2 mm) R wave as a marker of myocardial atrophy,
- T wave inversion with an amplitude of ≤2 mm because of scar and myocardial atrophy [7].
The sensitivity of this parameter was 97%, specificity was 81%, and negative predictive value was 99%.

T wave inversion in lead V1 with an amplitude of ≥2 mm

The same author presented this marker as a typical finding in patients with arrhythmogenic cardiomyopathy also in cases with T wave inversion only in lead V1 [8]. Children below the age of 14 years were excluded, the mean age of this cohort was 46.8±11.8 years. In bipolar precordial leads, a T wave inversion of ≥3 mm in lead V1 has been described as a typical finding [9]. Controlled by the same number of normal probands, the sensitivity was 93%, specificity was 97%, and negative predictive value was 99%.

Combination of the above-mentioned features

If typical ECG abnormalities in lead aVR and T wave inversion in lead V1 were combined, the sensitivity was 94%, specificity was 99%, and negative predictive value was 99.9% [10,11].

QRS fragmentation in all leads

Fontaine described QRS fragmentation as „pre“, „top“, and „post“-silon in patients with typical arrhythmogenic cardiomyopathy in high values [12]. QRS fragmentation is an unspecific finding and can be described in a large number of other diseases like hypertrophic or dilated cardiomyopathy, long QT syndrome, and Brugada syndrome [13].

Conclusions

There are indeed ECG parameters of concealed arrhythmogenic cardiomyopathy which enable the diagnosis at a very early phase. Ajmaline challenge in an early stage is positive in some cases. These are patients with a very high risk of ventricular fibrillation described by Corrado [14] and Peters [15]. Perhaps these are those patients with sudden cardiac death as the first manifestation of the disease.

Conflict of interest

The authors declare that they have no conflict of interest.

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References