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Risk stratification in early stage arrhythmogenic cardiomyopathy and provocable brugada ECG

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Abstract

Risk factors of malignant tachyarrythmias should be assessed in cases with early stage arrhythmogenic cardiomyopathy and provocable Brugada ECG. Spontaneous right precordial coved-type ST segment elevation, tall R wave in lead a VR, QRS fragmentation, 1° AV block, inducibility during electrophysiological examination, and localised right precordial QRS prolongation should be tested in 20 out of 128 patients.

Only QRS fragmentation and localised right precordial QRS prolongation were positive in these patients. QRS fragmentation was positive in one patient, however without any arrhythmic events. Localised right precordial QRS prolongation was positive in five patients with serious arrhythmic events – four cases with previous ventricular fibrillation and one case with subsequent slow ventricular tachycardia. All other risk factors could be ruled out.

In summary, localised right precordial QRS prolongation seems to be the strongest risk marker in cases of developing arrhythmogenic cardiomyopathy and provocable Brugada ECG.

Introduction

The risk of aborted sudden cardiac death is very high in cases with developing arrhythmogenic cardiomyopathy and provocable Brugada ECG [1]. In about 10 - 15% of cases there seems to be an association between arrhythmogenic cardiomyopathy and Brugada syndrome by the so-called connexome – a linkage of desmosomal proteins, sodium channel complexes and gap junctions including desmosomal genes like plakophilin-2 [2], desmoglein-2 [3], desmoplakin [4], and plakoglobin (in animal models).

Risk assessment in these cases is of utmost importance by spontaneous coved-type ST elevation in right precordial leads as the only independent risk marker [5], QRS fragmentation [6], positive R wave in lead a VR [7], 1° AV block [8], inducibility during electrophysiological examination [9], and as a new finding localized right precordial QRS prolongation demonstrated in the last ESC meeting in Barcelona 2017 [10] with a sensitivity of 78% and a specificity of 87%.

Above mentioned risk factors were analysed in cases with possible or typical developing arrhythmogenic cardiomyopathy and provocable Brugada ECG.

Method

In a cohort of 128 patients the diagnosis of typical (n=7, two major diagnostic criteria) and possible (n=13, one major and one minor diagnostic criteria) arrhythmogenic cardiomyopathy

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(72 males, meanage 46.1 +/- 13.6 years) was made. Diagnostic criteria were evaluated according to the paper published by Frank Marcus in 2010 [11]. Ajmaline challenge was used to provoke Brugada ECG that was positive in 20 cases (15%). In provoked Brugada ECG cases localised right precordial QRS prolongation, spontaneous Brugada ECG, tall R wave in lead a VR, 1° AV block, and inducibility during electrophysiological examination was analysed.

Results

Localised right precordial QRS prolongation could be detected in 5 cases – four cases with aborted ventricular fibrillation before ICD implantation with the diagnosis of an early stage of arrhythmogenic cardiomyopathy and Brugada syndrome and one case of slow ventricular tachycardia 2 years after ICD implantation with the same diagnosis.

In a single patient QRS fragmentation could be detected after ajmaline administration with the result of provocable Brugada ECG. This patient had no arrhythmice vents in a 72 months follow-up.

Spontaneous coved-type ST elevation in right precordial leads, tall R wave in lead a VR, 1° AV block, and inducibility during electrophysiological examination could be excluded.

Discussion

Since 2014 it is known that arrhythmogenic cardiomyopathy and Brugada syndrome have a common relationship in 10 – 15% of patients. The so-called connexome - an association between gap junctions, desmosomal proteins and sodium channel complexes – is the cause of this phenomenon and was first described in 2014 [1].

In isolated Brugada syndrome without structural abnormalities some risk factors for the development of arrhythmic events have been described: A positive R wave in lead a VR [7], spontaneous coved-type ST elevation [5], QRS fragmentation [6], inducibility of tachyarrythmias during electrophysiological examination [9], and I° AV-block [8] .

In early stages of arrhythmogenic cardiomyopathy with provocable Brugada ECG the situation is different. Generally, the risk of ventricular fibrillation is extremely high, supporting the theory that Brugada syndrome is the main disease with developing arrhythmogenic cardiomyopathy [12].

In the majority of patients (n=13) there was the diagnosis of possible arrhythmogenic cardiomyopathy without dilation and aneurysms of the right ventricle. In our group of patients there are no spontaneous cove-type ST-elevation in right precordial leads, no 1° AV block and during electrophysiological examination there were no inducibility of ventricular tachyarrhythmias in the cohort of 20 patients. In this group of patients a positive R wave of 3mm or more could be excluded; typical electrocardiographic appearance in lead a VR with significant Q wave, small R wave (2 mm or less), and negative T wave (2 mm or less) characterizes arrhythmogenic cardiomyopathy [13,14].

QRS fragmentation after ajmaline administration was positive in a single case, however, without arrhythmic events in a 72 months' follow-up.

During a last ESC meeting in Barcelona 2017 another risk factor gained further importance: Localized right precordial QRS prolongation [10]. This risk factor was tested in more than 200 patients with a sensitivity of 78% and a specificity of 87%.

In four patients (3 females) with aborted ventricular fibrillation before the diagnosis of arrhythmogenic cardiomyopathy was made and in one female patient with slow ventricular tachycardia after the diagnosis of arrhythmogenic cardiomyopathy with provocable Brugada ECG localized right precordial QRS prolongation was positive.

In this small study sensitivity and specificity were both 100%, suggesting that localised right precordial QRS prolongation is a strongest predictor of previous and future arrhythmic events.

Localized right precordial QRS prolongation is a strong diagnostic marker of arrhythmogenic cardiomyopathy [15] and - as it appears – the strongest risk factor in cases with developing arrhythmogenic cardiomyopathy and provocable Brugada ECG.

The value of drug-induced type 1 Brugada ECG and cardiac arrest is controversely discussed: Newest register data reveal an increasing risk for cardiac arrest of about 50% [16].

These results should be tested a larger collective of patients to gain statistically significant results. This aim is difficult to reach: Arrhythmogenic cardiomyopathy is a rare diagnosis and ajmaline testing in these patients is positive in only 15%.

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