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OPEN ACCESS JOURNALS

Archives of Clinical and Experimental Cardiology

Review | Vol 1 Iss 1

Characterization of Early Stage Arrhythmogenic Cardiomyopathy: Better Understanding of Different Arrhythmias

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Received: March 01, 2020; **Accepted:** April 09, 2020; **Published:** April 17, 2020

1. Introduction

Arrhythmogenic cardiomyopathies include arrhythmogenic right ventricular cardiomyopathy, arrhythmogenic left ventricular cardiomyopathy, arrhythmogenic biventricular cardiomyopathy and arrhythmogenic dilated cardiomyopathy [1].

In terms of different arrhythmias (monomorphic ventricular tachycardia, polymorphic ventricular tachycardia or ventricular fibrillation) it would be useful to add early stage arrhythmogenic cardiomyopathy to the list of arrhythmogenic cardiomyopathies.

The problem is how to define early stage arrhythmogenic cardiomyopathy and to differentiate Brugada syndrome or right ventricular outflow tract tachycardia.

2. Dilated Right Ventricle

A non-dilated right ventricle without aneursyms characterizes early stage arrhythmogenic cardiomyopathy in terms of imaging techniques. Even in manifest arrhythmogenic cardiomyopathy a dilated right ventricle is no longer a parameter for diagnosis according to angiographic or other imaging techniques like echocardiomyopathy or cardiac MRI [2]. In the last Task Force criteria a dilated right ventricle was a matter of fact [3].

3. ECG Definition

Several attempts were made to characterize early stage arrhythmogenic cardiomyopathy by ECG. The S wave angle [4,5], but also the ECG appearance in lead aVR [4,6] and the amplitude of inverted S wave in lead V1 [4,7] seems to be markers of early stage arrhythmogenic cardiomyopathy. Nevertheless, typical appearance of lead aVR and T wave amplitude in lead V1 or more than 2 mm seems to be a sign of arrhythmogenic left ventricular cardiomyopathy or mutated ankyrin B [8].

4. The Role of Ajmaline Testing

In early stage arrhythmogenic cardiomyopathy the first manifestation of the disease can be sudden cardiac death. In about 15% of cases early stage arrhythmogenic cardiomyopathy is associated with provocable Brugada ECG [9]. In a recent paper the marker of high risk for dramatic arrhythmic events was localized right precordial QRS prolongation [10]. The so-called connexion - the association of gap junctions, sodium channel mutations and desmosome mutations - plays a major role and is described for plakophilin-2, but also for desmoglein-2 and desmoplakin, and in animal studies for plakoglobin [11].

There is a continuum between Brugada syndrome on the one side, and arrhythmogenic cardiomyopathy on the other side. It can be postulated that ventricular fibrillation of polymorphic ventricular tachycardia may be caused by Burgada syndrome later developing to arrhythmogenic cardiomyopathy.

5. Differentiation of RVOT Tachycardia and Arrhythmogenic Cardiomyopathy

It is well known that right ventricular outflow tract tachycardia can develop over years to arrhythmogenic cardiomyopathy. RVOT tachycardia do not normally lead to sustained tachycardia, otherwise arrhythmogenic cardiomyopathy do lead to sustained, long-standing ventricular tachycardia. In rare cases, non-sustained ventricular tachycardia in RVOT tachycardia can lead to sudden arrhythmic death [12].

6. QRS Prolongation in Ectopic Ventricular Beats

Early stage arrhythmogenic cardiomyopathy can be characterized in rare cases by a severely prolonged QRS complex in ectopic ventricular beats [13]. The ECG's of the patients were not systematically analyzed, whether S wave angle, typical features of lead aVR, or T wave amplitude in lead V1 were striking is not known.

7. QRS Fragmentation

QRS fragmentation in arrhythmogenic cardiomyopathy is a marker of severe arrhythmias in otherwise positive ECG findings. If QRS fragmentation without any other typical ECG findings in many leads is positive, this parameter seems to a sign of early stage arrhythmogenic cardiomyopathy [14]. If combined by ajmaline challenge this finding is likely to predict ventricular fibrillation.

8. Conclusions

Early stage arrhythmogenic cardiomyopathy is not easy to define. Imaging techniques, standard ECG and provocation tests are necessary to make a clear diagnosis. The diagnosis of early stage arrhythmogenic cardiomyopathy is important in order to make a clear prediction of ventricular arrhythmias and to define the risk of sudden arrhythmic death. Under the term of arrhythmogenic cardiomyopathies early stage arrhythmogenic cardiomyopathy should be included.

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