

## J waves in arrhythmogenic cardiomyopathy versus primary electrical disease

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### Abstract

In case reports early repolarization (ER) is associated with arrhythmogenic cardiomyopathy. In smaller case analyses ER is a discussable risk marker. Furthermore, the association with inversions of right precordial T-waves should be analysed.

Method: In a cohort of 360 patients with typical ESC/ISFC criteria of arrhythmogenic cardiomyopathy (176 males, mean age 47.3 +/- 13.7 years) the presentation with ER and the risk assessment for sudden cardiac arrest was analyzed.

Results: A total of 113 cases (31%) with inferior (22%), inferolateral (3%), and lateral (6%) notching or slurring was presented as typical signs of early repolarization syndrome. Together with the results of ajmaline challenge it is in close relation with the number of right precordial T-wave inversions. Early repolarization does not identify patients at risk for sudden cardiac arrest. In single cases the risk was increased in cases with ST elevation in early repolarization pattern as primary electrical disease.

Conclusions: The number of early repolarization pattern in typical arrhythmogenic cardiomyopathy is increased and relates to the number of right precordial T-wave inversions of 55% in this cohort. Early repolarization is generally not a risk marker, but in single cases overlapping primary electrical disease can be documented.

**Keywords:** arrhythmogenic cardiomyopathy; early repolarization; right precordial ER; Brugada ECG

### Introduction

Early repolarization pattern is present in a normal, healthy population in a total prevalence of 7.3%, in males 10.5%, in females 1.9% [1]. In idiopathic ventricular fibrillation early repolarization pattern is present in 31% with higher recurrence rate [2]. The correlation between arrhythmogenic cardiomyopathy and early repolarization syndrome was been discussed for a long period of time. Case reports of single patients with typical diagnosis of arrhythmogenic cardiomyopathy, missing right precordial T wave inversions and signs of positive early repolarization criteria were published [3].

In a paper of Boukens et al. an diagramm of the severity of structural abnormalities leads to a relation between early repolarization, Brugada syndrome, arrhythmogenic cardiomyopathy, and finally Uhl's anomaly [4]. In the same paper a basis for overlap between fibrosis in early repolarization syndrome and brugada syndrome is presented [4]. In another paper the same author presented a case with fibrofatty abnormalities in early repolarization syndrome in inferior leads [5].

For this purpose J waves were looked for in patients with typical signs of arrhythmogenic cardiomyopathy. A correlation to right precordial T-wave inversions was analyzed.

### Method

In a cohort of 360 cases with AHA/ESC criteria of arrhythmogenic cardiomyopathy (176 males, mean age 47.3 +/- 13.7 years) typical electrocardiographic signs of early repolarization syndrome was searched for. Early repolarization syndrome was characterised of notching or slurring of the QRS complex in inferior, lateral, and inferolateral leads. In a certain number of patients (n= 102) Brugada challenge was done in order to search for

provocable Brugada ECG.

### Results

In a whole cohort of patients analyzed there were 22 cases with cardiac arrest (6%), 65 cases with syncope (16%), 98 cases with sustained ventricular tachycardia (27%), 67 cases with non-sustained ventricular tachycardia (19%), and 34 cases with palpitations [9]. No arrhythmias were documented in 83 cases (23%).

A total of 113 cases (31%) with inferior (22%), inferolateral (3%), and lateral (6%) notching or slurring was presented as typical signs of early repolarization syndrome.

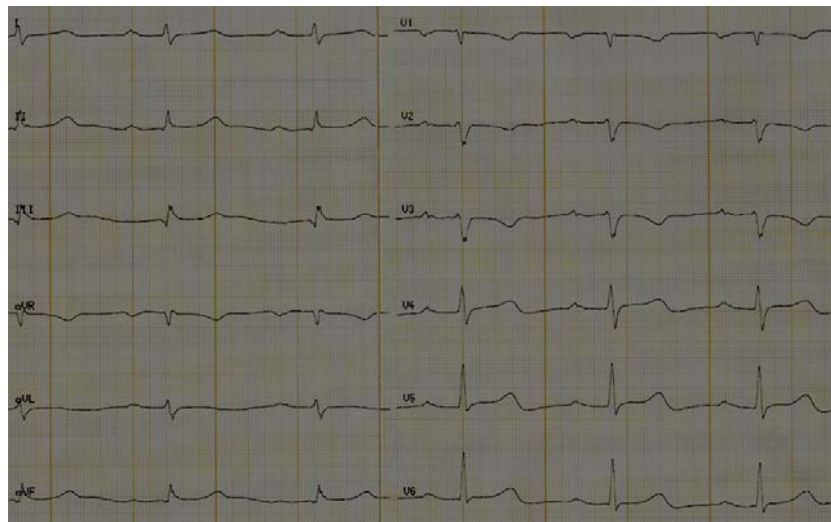
A comparison of clinical findings between cases with and without early repolarization syndrome was analyzed in this context. With regard to cardiac arrest (n=22), syncope (n=56), non-sustained ventricular tachycardia (n=67) and no arrhythmias (n=83) there were no differences. Merely for sustained ventricular tachycardia (n=98) and palpitations (n=34) there were slight differences. No signs of early repolarization favored sustained ventricular tachycardia ( $p < 0.05$ ), but existance of early repolarization favored palpitations ( $p > 0.0025$ ).

In additional, 15% of 102 cases right precordial J waves could be provoked by ajmaline challenge presenting as typical brugada ECG.

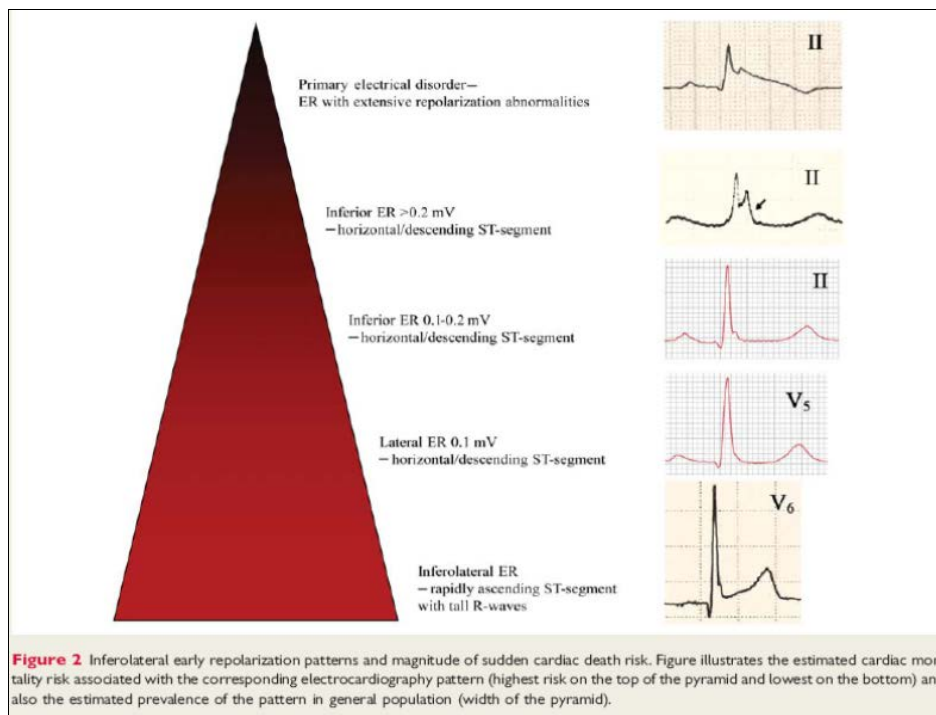
Right precordial T-wave inversions were present in a total of 55%. An example of right precordial T-wave inversion or early repolarization syndrome in the same patient is presented in table 1 and 2. The first table presents typical early repolarization syndrome in inferolateral leads with T-wave inversion only in lead V1 just after resuscitation due to ventricular fibrillation. In table 2 ECG was recorded days after resuscitation. Right precordial T-wave inversion is present in lead V1, V2 and V3. QRS fragmentation is present in lead II, III, aVF and in lead V2 and V3.



**Fig 1:** The standard ECG of the patient just resuscitated for ventricular fibrillation



**Fig 2:** The ECG of the same patient some days ago



**Fig 3:** Early repolarization diagram and risk of sudden cardiac arrest

## Discussion

After careful analysis of T-wave inversions and early repolarization syndrome in the analyzed cohort of patients with definite arrhythmogenic cardiomyopathy it seems that either typical right precordial T-wave inversions or inferolateral early repolarization are typical changes of repolarization in patients with arrhythmic cardiomyopathy. Together with abnormalities of depolarization (epsilon waves, right precordial QRS prolongation, terminal activation delay, and QRS fragmentation) these findings describe well electrocardiographic components in the definition of arrhythmogenic cardiomyopathy. Right precordial T-wave inversions in this cohort accounts for 55% of cases <sup>[6]</sup>, characterising histologic abnormalities in the right ventricular outflow tract <sup>[7]</sup>. Early repolarization in inferior leads characterise histologic abnormalities in the inferior, tricuspid valve – near region <sup>[5]</sup>. Inferior QRS prolongation, mentioned in a paper <sup>[8]</sup>, is caused by inferior early repolarization.

For all we know early repolarization is not a marker of risk, but an essential diagnostic tool in defining arrhythmogenic cardiomyopathy electrocardiographically. The opposite is true for Brugada syndrome, where early repolarization in inferolateral leads seems to be a risk marker for sudden cardiac death <sup>[9]</sup>.

The presented case documents a dynamic process with early repolarization pattern just after resuscitation and right precordial T-wave inversions and QRS fragmentation days ago. This process presents an interaction between right precordial T-wave inversions and early repolarization pattern demonstrating in this special case a certain risk factor of early repolarization for ventricular fibrillation with ST elevation in lead III and aVF as primary electrical disease (see figure 3). In this respect it must be discussed whether early repolarization is actually a repolarization criterion or indeed a depolarization criterion. In other cases early repolarization is definitely not a risk factor for sudden cardiac death. The number of patients with survived sudden cardiac death was rather low, the analysis of early repolarization as a risk marker inconclusive.

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