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Early Repolarization Syndrome

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The electrocardiographic pattern of early repolarization (ER) is common, with a particularly high prevalence reported amongst athletes and adolescents. It has long been associated with benign outcome [1-3]. Recently, an association between inferolateral ER pattern and sudden cardiac death (SCD) has been established by different groups [4-7]. Population-based studies have also reported an increased mortality rate among patients with inferolateral ER pattern compared to controls [7-9]. To bring back together these differences, it is important to focus on the definition of ER pattern used in these different studies as well as the population included. The definition of ER pattern associated with sudden cardiac death was the presence of J point elevation more than or equal to 0.1mV in at least 2 contiguous inferior and/or lateral leads of a standard 12-lead ECG and not ST elevation as it was often the case in the studies with benign outcome. Any study dealing with ER should clearly indicate the definition used. Otherwise it cannot be interpreted. Talking about definition, ER syndrome is an ER pattern (as defined above) associated with symptoms (syncope or aborted SCD) and/or familial history of SCD as mentioned in the last HRS/EHRA/APHRS Expert Consensus Statement on the Diagnosis and Management of Patients with Inherited Primary Arrhythmia Syndromes [10]. It is important to recognise that having only an ER pattern is not a disease.

Another potential difference between the studies focusing on ER is the studied populations. Studies with a correlation between ER pattern and SCD included mainly Caucasian or Asian and very little African, African/American nor athletes. Therefore, their findings cannot be extrapolated to these populations.

Based on the different studies linking ER pattern and SCD, only a small minority of patients with this pattern will experience SCD, while the majority will remain asymptomatic. Therefore, the identification of this minority of patients represents a significant challenge. Currently, criteria for malignant ER pattern rely on 12 leads ECG parameters on top of arrhythmic syncope. The amplitude of J point more than 0.2mV and a descending ST segment are the 2 main pejorative parameters [5,7,11-13]. The presence of the ER pattern in inferior vs lateral leads, notch vs slur and QRS fragmentation have been reported as pejorative criteria but are still controversial.

Concerning risk stratification, it seems now clear that programmed ventricular stimulation has no interest in asymptomatic patient and should not be done [14]. Currently, asymptomatic persons with

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ER pattern should not be treated. However close follow-up is warranted in case of J point elevation more than 0.2mV with horizontal/descending ST segment. In patients with arrhythmic syncope, an implantable cardioverter defibrillator (ICD) may be considered (based on the expert consensus statement). In clinical practice, syncope may be of undetermined origin; ICD vs loop recorder implantation is then a case by case decision. Of note, patients with resuscitated SCD and ER pattern have a recurrence risk x2 compared to the same population without ER pattern. In case of recurrent ICD shocks, isoproterenol may halt arrhythmic storm and quinidine may prevent from recurrent ventricular fibrillation episodes [15,16].

Because 5% of a caucasian population may present this pattern with only 1/2500 of them at risk of ventricular fibrillation, better tools for risk stratification are necessary. However it is crucial to recognise this pattern in patients with a familial history of SCD, patients with syncope or patients with dramatic J point elevation and descending/horizontal ST segment.

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