Letter to the Editor:

J wave syndromes: where's the scar?

Refers to "Structurally abnormal myocardium underlies ventricular fibrillation storms in a patient diagnosed with the early repolarization pattern" by Boukens et al. DOI: 10.1016/j.jacep.2020.06.027

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We read with great interest the paper by Boukens and colleagues (1) detailing the pathophysiology underlying inferior Early Repolarisation (ER) pattern in a patient with VF storms and Early Repolarisation Syndrome (ERS) (2). Open chest epicardial mapping demonstrated delayed activation and heterogeneous repolarization of the inferior right ventricular free wall and fractionated local potentials coinciding with the ECG ER pattern, which were successfully treated by radiofrequency ablation. Most interestingly, morphometric analysis on biopsied cardiac tissue from this region showed areas of extensive fibrosis. This study provides the first *in vivo* evidence that the ER pattern can be the ECG manifestation of temporal heterogeneity in cardiac activation due to regional replacement fibrosis.

ERS and the Brugada Syndrome (BrS) are characterised by J-point elevation (J Wave Syndromes), both predisposing to sudden cardiac death (SCD) in young adults with no apparent structural heart disease. This paradigm has, however, been challenged: it has been postulated that pathological remodelling of the epicardium of the right ventricular outflow tract of patients with BrS leads to decreased cellular excitability and inhomogeneous slowing of the cardiac conduction, manifesting in delayed, fragmented electrograms and a type 1 Brugada pattern on the ECG: the *depolarization theory*. We have shown that increased fibrosis and dysregulation of gap junctions are present at these sites, and that the application of radio-frequency energy can eliminate the abnormal electrical potentials, reduce the J point and ST elevation and suppress

arrhythmias (2). Epicardial mapping and ablation have demonstrated similar findings and success in ERS patients, although abnormal electrograms were present predominantly in the right ventricular inferior wall (3). Thus, Boukens and colleagues have given further support to regionalised epicardial fibrosis as a unifying pathophysiology underlying much of the J Wave Syndromes.

This unlocks new potential strategies for the investigation and prevention of unexplained SCD. For example, whilst the ER pattern is common in the general population, it is overrepresented in families of autopsy negative SCD victims (4). Currently, robust risk stratification strategies are lacking in asymptomatic subjects with ER pattern, making the assessment of ERS in autopsy negative SCD families particularly challenging. BrS is frequently diagnosed in these families, but the true incidence of ERS in autopsy negative SCD is unknown (5). Now that a depolarisation mechanism has been confirmed as relevant by Boukens et al, research into diagnostic and prognostic evaluation can focus on new non-invasive and invasive markers of localised fibrosis and conduction delay.

References:

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