Epicardial Brugada syndrome ablation unmasking inferior J waves

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Abstract
Patients with Brugada syndrome are at risk of life-threatening ventricular arrhythmias. Epicardial substrate ablation for Brugada syndrome has been described as a means of controlling these arrhythmias and recent reports describe elimination of the Brugada phenotype with ablation. We describe a unique case in which a patient developed inferior J waves with an early repolarization-type electrocardiogram following successful epicardial infundibular substrate ablation (which eliminated the Brugada syndrome electrocardiogram on ajmaline challenge). We discuss the likely underlying pathophysiology responsible for this phenomenon, its relationship to the anatomic obstacles encountered during epicardial ablation, and the implications for long-term arrhythmic risk.

KEYWORDS
ajmaline, Brugada syndrome, early repolarization, epicardial ablation, J waves

1 | CASE PRESENTATION

A 39-year-old man presented following out-of-hospital cardiac arrest. He was found to be in ventricular fibrillation (VF) following 12 minutes of cardiopulmonary resuscitation and required three attempts at defibrillation before restoring sinus rhythm.

He had three previous episodes where he became "stiff and unresponsive," had an agonal breathing pattern, and suffered urinary incontinence. His sister died at the age of 8 months from sudden infant death syndrome. Another sister had an unexplained "seizure" in the shower at the age of 3. A maternal cousin suffered a VF arrest during sleep.

His electrocardiogram (ECG) on presentation demonstrated a Type I Brugada pattern but only when leads V1 and V2 were placed in the second intercostal space (Figure 1A), with 2 mm of coved ST elevation in V2. He also had J wave notching (>1 mm) in the inferior leads (II, III, aVF), though without any significant ST segment elevation. He was diagnosed with Brugada syndrome (BrS). Coronary angiography excluded obstructive coronary artery disease. A cardiac magnetic resonance imaging scan demonstrated normal biventricular function with no late gadolinium enhancement. On genetic testing, he was found to have an SCN5A mutation (substitution c.3837+1G>A) which results in haploinsufficiency due to protein degradation from altered mRNA splicing. This mutation has been classified as pathogenic in The Human Gene Mutation Database.

He underwent insertion of a single-chamber implantable cardioverter-defibrillator (ICD). He suffered mild cognitive deficits following the cardiac arrest but recovered well overall.

Three years later, he presented following a syncopal episode due to VF requiring high-voltage therapy in addition to four other episodes of ventricular arrhythmias (VAs). Quinidine was considered for suppression of VAs; however, this was declined due to concerns of compliance in light of his cognitive deficits and thus he was offered catheter ablation.

Under general anesthesia, multipolar catheters were advanced from the femoral vein. He had prolonged QRS width (132 ms) and mild His-Purkinje disease (HV 63 ms). Other baseline conduction intervals were within normal range. Percutaneous epicardial access was obtained without complication using the standard Tuohy needle technique and an open irrigated 3.5-mm catheter (Navistar®, Thermocoool®, Biosense Webster, Diamond Bar, CA, USA) was introduced through a deflectable sheath. Electroanatomic substrate mapping with computed tomography image integration revealed confluent areas of extreme fractionation with attenuated voltage over the epicardial aspect of the right ventricular outflow tract (RVOT) and basal right ventricular (RV) body extending to the inferior wall adjacent to the atrioventricular groove (Figure 2). Radiofrequency ablation titrated to 30–50-W power was performed targeting these areas of abnormal electrograms. Areas in proximity to the conus and acute
marginal branches of the right coronary artery (RCA) identified on coronary angiography were avoided (Figure 2), resulting in persistence of abnormal electrograms over the inferior aspects of the epicardial RV body. During ablation, there was accentuation of the ST segment elevation in the anterior precordial leads, resulting in an exaggerated Brugada pattern on the surface ECG.

Following ablation, there was not only complete abolition of the Brugada phenotype on the ECG but also accentuation of the early repolarization (ER) pattern with up to 2 mm of J point and ST segment elevation in the inferior leads (Figure 1B) seen immediately post-procedure. He underwent a delayed ajmaline challenge which resulted in accentuation of the inferior ER pattern on his ECG (Figure 3). No Brugada pattern was seen. He remains free of VAs off antiarrhythmic agents after 10 months.

2 | DISCUSSION

BrS has been considered a channelopathy affecting structurally normal hearts. However, recent series have shown regional epicardial fibrosis and electrogram abnormalities affecting the RVOT in patients with BrS. This corresponds to the anterior precordial ECG leads (V1-V2) where the Brugada phenotype is best seen.

In patients with BrS, published case series suggest that patients who undergo epicardial ablation with normalization of the Brugada ECG phenotype appear to have reduced recurrence of VAs during follow-up. Quinidine, a Class IA antiarrhythmic agent, can reduce recurrent VAs or ICD shocks by inhibiting the Ito current; however, quinidine has a significant side effect profile including risk of Torsades de Pointes,
thrombocytopenia, and cinchonism. Epicardial ablation, therefore, may be a reasonable option in some patients.4

During ablation we excluded areas of abnormal electrograms that were within 5 mm of the RV branches of the RCA. Injury to coronary vessels is a documented risk of epicardial ablation,5 though this has not been noted to occur in the published case series.

ER pattern is defined as the presence of a J wave of at least 1 mm in two contiguous leads excluding leads V1–V3. ER has recently been shown to be associated with increased risk of VAs, in both patients who have suffered idiopathic VF as well as in the general population.6–8 The more malignant pattern of ER affecting the inferior or inferolateral leads is associated with greater risk of VAs, compared to the relatively benign pattern seen in the anterior precordial leads.9 Some patients with BrS have concomitant inferolateral ER; these patients have a higher risk of recurrent VF.9 Patients may exhibit alternating patterns of ER and BrS at separate points in time.10 Both BrS and ER involve abnormalities of the J wave and can be considered two diseases on the spectrum of “J wave syndromes.”11 Common features include the role of the Ito channel in augmenting the J wave, propensity to Phase II reentry resulting in VF, and the therapeutic response to isoprenaline and quinidine. However, the primary region of J wave distortion differs in the two conditions; ER predominantly affects the inferolateral territory, while BrS affects the anterior RVOT.12

Sodium channel blocking agents such as ajmaline have been traditionally used to unmask a latent Type I Brugada pattern.13 However, a recent study suggests these same agents attenuate the inferolateral ER pattern in patients with concomitant BrS and ER.14 Ours is the first case in which an inferior pattern of ER has been accentuated by both epicardial ablation for BrS as well during a challenge with sodium channel blockade. Our patient meets criteria for both Brugada and ER syndromes, and the presenting ECG showed features

FIGURE 2 (A) Coronary angiogram in right anterior oblique view (RAO 30°) demonstrates proximity of the RV marginal branches to the ablation catheter. Abl = ablation catheter; RCA = right coronary artery. (B) Anteroposterior view of epicardial electroanatomic CT image integrated substrate maps. Areas in purple represent normal epicardial voltage (≥1 mV). Bipolar fractionated electrograms are annotated in black. Radiofrequency ablation lesions (red dots) concentrated in the superior aspect of the RV outflow tract due to mid-inferior RV body territory proximity to RCA branches (arrow). Representative bipolar electrograms from the epicardial RVOT and inferior RV body are illustrated (orange arrows). Ao = aorta; PA = pulmonary artery; RA = right atrium; RV = right ventricle; SVC = superior vena cava. (C) Highly fractionated local electrograms with attenuated voltage at sites over RV outflow tract (arrow = corresponding to black dots on B) [Color figure can be viewed at wileyonlinelibrary.com]
of both, raising the possibility of the coexistence of these two related entities in this patient. A recent case report describes successful ablation of coexistent inferior J waves in a patient with BrS. However, he has a pathogenic mutation in the SCN5A gene known to cause BrS. His ajmaline-induced inferior J wave accentuation is contrary to the expected response of J wave amplitude reduction in patients with true ER, although a recent case report suggests that sodium channel blockade may augment J waves over the lateral left ventricle in ER, though this is masked by the widened QRS on the surface ECG.

While BrS and ER may share a common pathophysiology distinguished by the region of the ventricles affected, the response in our patient of the inferior J waves and ST segments to ablation and sodium channel blockade suggests the inferior J wave augmentation is more likely to represent residual regional Brugada unmasked by our limited ablation in the inferior territory (due to coronary artery proximity) rather than true ER. This distinction may have prognostic implications as the presence of residual Brugada substrate following ablation raises the concern for recurrent arrhythmic events. On the other hand, patients with BrS and concomitant ER (in the absence of QRS fragmentation) do not appear to have a higher incidence of ICD shocks compared to those without ER.

### 3 | CONCLUSION

We present a case of successful epicardial ablation of BrS which unmasked an inferior ER pattern. This likely represents a residual regional Brugada phenotype masquerading as ER. While an ECG pattern of ER can exist in patients with BrS, our case is the first in which this was accentuated following successful epicardial outflow.

**FIGURE 3** 12-lead electrocardiograms demonstrating accentuation of the inferior “early repolarization” pattern following ajmaline challenge (arrows) [Color figure can be viewed at wileyonlinelibrary.com]
tract ablation of the BrS phenotype and during sodium channel blocker challenge.

REFERENCES


How to cite this article: Lee A, Kohler H, Wright D, Haqqani HM. Epicardial Brugada syndrome ablation unmasking inferior J waves. Pacing Clin Electrophysiol. 2017;00:1–5. https://doi.org/10.1111/pace.13095