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## Augmentation of J Waves and Electrical Storms in Patients with Early Repolarization

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**To the Editor:** Early repolarization, consisting of an elevation of the QRS-ST junction (J point), QRS notching or slurring (J wave), and a tall, symmetric T wave, is generally considered to be benign.<sup>1</sup> On the basis of preclinical experimental evidence, it has been suggested that some forms of early repolarization seen in the clinic may not be benign, especially when associated with the occasional appearance of J waves or ST-segment elevation.<sup>2</sup> Sporadic case reports and basic electrophysiological research have suggested a critical role of the J wave in the pathogenesis of idiopathic ventricular fibrillation.<sup>3,4</sup> Clinical evidence in support of an association between early repolarization and idiopathic ventricular fibrillation was previously reported in preliminary form by Haïssaguerre et al. and is fully disclosed by these researchers in this issue of the *Journal*.<sup>5</sup> However, direct evidence of a relation between early repolarization and the appearance of accentuated J waves in idiopathic ventricular fibrillation has been scarce.

We evaluated the incidence of early repolarization among 1395 controls who were representative of the general population and 15 patients classified as having idiopathic ventricular fibrillation, excluding patients with long- and short-QT syndromes, the Brugada syndrome, or catecholaminergic polymorphic ventricular tachycardia. Among the 15 patients with idiopathic ventricular fibrillation, 4 presented with electrical storm (four or more episodes of ventricular fibrillation in 1 day). Continuous electrocardiographic monitoring of the patients with electrical storm was performed in the coronary care unit.

The incidence of early repolarization among controls was 3.3%. In contrast, the incidence of early repolarization among patients with idiopathic ventricular fibrillation was 60% (9 of 15 patients). All four patients with idiopathic ventricular fibrillation and electrical storm had early repolarization. These four patients, whose hearts were apparently structurally normal, had a unique electrocardiographic signature consisting of a baseline early repolarization pattern; dramatic but transient accentuation of J waves across the precordial and limb leads before the development of electrical storm, which was precipitated by relatively short-coupled premature ventricular contractions; and suppression of the accentuated J waves and ventricular fibrillation with the administration of quinidine and isoproterenol and with pacing at increasingly rapid rates. In these patients with idiopathic ventricular fibrillation, unlike patients with the Brugada syndrome, the electrocardiographic and arrhythmic abnormalities could not be provoked with intravenous flecainide, were not limited to the right precordial leads, and were not accompanied by abnormalities on the signalaveraged electrocardiogram.

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Our observations suggest that an early-repolarization pattern is not always benign, as was previously thought, and that the transient appearance of global J waves in this setting is indicative of a highly arrhythmogenic substrate, representing a unique clinical syndrome associated with a high risk of sudden death from cardiac causes. We propose that this unique phenotype represents a variant of a much broader syndrome, including the Brugada syndrome, in which the appearance of prominent J waves underlies the development of arrhythmogenicity.

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