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Risk of sudden death among young individuals with J waves and early repolarization: Putting the evidence into perspective

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The presence of J waves and ST-segment elevation on the electrocardiogram (ECG), jointly termed "the early repolarization pattern," has traditionally been considered a marker of "good health." However, recent case control series and long-term population studies have established a statistically significant association between this ECG pattern and an increased risk for arrhythmic death. This finding has raised concern among physicians, who now are asked to estimate the "arrhythmic risk" following the incidental discovery of J waves on routine ECG. Therefore, we review the literature linking early repolarization with arrhythmic risk to place this "fear of J waves" in the right perspective. We found five case control studies (involving 331 patients with idiopathic ventricular fibrillation [VF] and 8,649 controls). All of these studies showed that J waves, particularly of large amplitude and recorded in multiple leads, are more prevalent among patients with idiopathic VF. We also found three large population studies (involving >17,000 individuals) looking at the prognostic value of early repolarization. Two of these studies showed that the presence of J waves >2 mm in amplitude in asymptomatic adults is associated with a threefold increased of arrhythmic death during very long-term follow-up. Individuals with J waves do have some degree of increased dispersion of repolarization that places them at increased risk for arrhythmic death, but only in the presence of additional proarrhythmic factors or triggers. A sensible approach for the asymptomatic patient with J waves is proposed.

KEYWORDS Arrhythmic death; Early repolarization; Idiopathic ventricular fibrillation; J wave

ABBREVIATIONS **ECG** = electrocardiogram; **EP** = electrophysiologic; **ICD** = implantable cardioverter-defibrillator; **VF** = ventricular fibrillation; **VT** = ventricular tachycardia

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The presence of J waves and ST-segment elevation on the electrocardiogram (ECG), jointly termed "the early repolarization pattern," was harmoniously considered a benign finding devoid of clinical significance.[1–6] For decades, investigations of this phenomenon focused exclusively on how to distinguish this "normal variant" from clinically important causes of ST-segment elevation, most notably myocardial infarction. Given that early repolarization is more prevalent in the young and the fittest individuals[7–11] and predominates at slower heart rates, it was generally viewed as an ECG marker of "good health."[5] However, this view was recently challenged Table 1. First, several case reports highlighted the fact that patients with idiopathic ventricular fibrillation (VF), a highly lethal disease affecting young individuals with no organic heart disease, not only have obvious J waves but also demonstrate augmentation of J-wave amplitude immediately prior to the onset of malignant arrhythmias, the latter phenomenon suggesting cause and effect Figure 1C.[12–21] Support for the proarrhythmic significance of the early repolarization pattern also originated in experimental studies demonstrating that the J wave is a marker of increased dispersion of repolarization,[22] a substrate for reentrant arrhythmias.[23] Subsequently, several case control series Table 1 consistently showed that J waves and early repolarization were more prevalent among patients with idiopathic VF than among carefully matched healthy controls.[24–27] Finally, large population studies have shown that the presence of J waves is clearly associated with increased long-term cardiac and arrhythmic mortality Table 1.[28,29] Since then, the malignant attributes of J waves in the inferior and lateral leads were recognized in specific clinical conditions as diverse as the congenital short QT syndrome,[30] Brugada syndrome,[31,32] and coronary disease.[33] Moreover, because the tachyarrhythmias in these diverse entities share an arrhythmic mechanism (i.e., phase 2 reentry),[23] the concept of *J-wave syndromes* was put forward.[34,35] Even among athletes, the population group for whom early repolarization was long considered to be an innocent "occupational trademark,"[7–11] J waves have now been reported to portend an increased risk for arrhythmic death.[36]

The emergence of the concept of "malignant J waves"[35] as an accepted model has dramatically altered physicians' perceptions regarding the clinical significance of the early repolarization pattern. Consequently, physicians encountering asymptomatic young patients with "J waves" ought to

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Table 1 Evidence supporting the association between "early repolarization" and increased risk for arrhythmic death

Reference	No. of patients	Criteria for early repolarization	Main findings
Case reports	8	J wave ± ST- elevation	Early repolarization with marked J waves noted in patients with otherwise idiopathic VF J-wave amplitude increases during bradycardia and immediately prior to onset of VF.[14,15,17,20,47]
Case control series			
Haissaguerre et al[24]	Idiopathic VF = 206 Controls* = 412	QRS-ST junction elevation ≥0.1 mV (notching or slurring) in inferior or lateral leads	Increased prevalence of early repolarization in idiopathic VF (31% vs 5%, $P < .001$)
Rosso et al[25]	Idiopathic VF = 45 Controls* = 124 Athletes = 121	QRS-ST junction elevation (notching or slurring); QRS-ST junction elevation ≥0.1 mV also reported	Increased prevalence of early repolarization in idiopathic VF (42% vs 13%, $P = .001$) Intermediate prevalence among healthy athletes (21%)
Merchant et al[49]	Idiopathic VF = 39 Controls with benign early repolarization = 61	QRS-ST junction elevation ≥0.1 mV (notching or slurring) in 2 consecutive leads	Left precordial early repolarization more prevalent in malignant variants of early repolarization than in benign cases
Nam et al[27]	Idiopathic VF = 19 Controls = 1,395	QRS-ST junction elevation ≥0.1 mV (notching or slurring) in 2 consecutive leads	Early repolarization more common in idiopathic VF (58% vs 3%), especially early repolarization in multiple leads (73% vs 15%, $P < .05$)
Abe et al[26]	Idiopathic VF = 22 Control = 6,657	QRS-ST junction elevation ≥0.1 mV (J wave) in 2 consecutive leads	Early repolarization more common in idiopathic VF (32% vs 2%, $P < .001$)
Population-based studies			
Klatsky et al[5]	2,081 adults who voluntarily underwent ECG recording	Early repolarization defined mainly as ST elevation ≥1 mm (J waves noted in only 29%)	No increased risk of death (HR 0.8, 95% CI 0.6– 1.2) or hospitalization (HR 1.0, 95% CI 0.9–1.2) in individuals with early repolarization
Tikkanen et al[28]	10864 adults followed for 30 years	J-wave elevation ≥0.1 mV (notching or slurring) in inferior or lateral leads	Increased risk of cardiac mortality (adjusted RR 1.28, 95% CI 1.04–1.59) in individuals with early repolarization in inferior leads Increased arrhythmic mortality in those with J wave >2 mV
Sinner et al[29]	6,213 adults followed for 18 years	J-wave elevation ≥0.1 mV (notching or slurring) in all leads except V_1–V_3	2- to 4-fold increased risk of cardiac mortality in individuals 35–54 years old with early repolarization, particularly when present in inferior leads

CI = confidence interval; HR = hazard ratio; RR = relative risk; VF = ventricular fibrillation.
*Matched by age, gender, and race.

consider their potential arrhythmic risk.[37] Here we review the literature linking early repolarization with arrhythmic risk in general (Table 1) and with the syndrome of idiopathic VF in particular to place "the fear of J waves"[37] into perspective.

What is idiopathic VF?

A detailed description of this disease is beyond the scope of this essay and can be found elsewhere.[38,39] In brief, idiopathic VF is a disorder presenting as syncope or cardiac arrest caused by polymorphic ventricular tachycardia (VT) that invariably is triggered by ventricular extrasystoles with a very short coupling interval, falling on the peak or on the descending limb of the preceding T wave.[40] The triggering extrasystoles have been mapped mostly to the His–Purkinje system[41] of the left ventricle, and the arrhythmic substrate appears to correspond to a short QT syndrome.[42] Patients with idiopathic VF,[38] particularly those with prominent J waves,[43] tend to develop arrhythmic storms with numerous episodes of VF that fail to respond to conventional antiarrhythmic therapy but respond exquisitely to quinidine therapy.[38,43] Importantly, idiopathic VF affects young adults.[38] Extensive review of published cases suggests that the typical idiopathic VF patient is 20 to 40 years old.[39] In the largest multicenter study of idiopathic VF, the mean age was 35 ± 10 years.[24] This young age should be kept in mind when analyzing population studies suggesting an increased arrhythmic risk among individuals with J waves[28,29] (see following).

J waves and VF

The association of J waves with increased risk for VF was first noted for hypothermia.[44] Indeed, J waves were first termed "Osborn waves" subsequent to their initial description in hypothermia.[44] The proarrhythmic nature of this association was demonstrated by the high percentage of dogs that developed spontaneous VF during experimental hypothermia.[44] In fact, increased dispersion of ventricular repolarization and spontaneous phase 2 reentry leading to polymorphic VT have been elegantly demonstrated in experimental preparations by simply cooling the perfusion diluents.[45] Interestingly, quinidine is also effective in preventing hypothermia-induced VF.[46]