Ventricular arrhythmias induced by phase 2 reentry in a patient with J-wave syndrome

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Introduction

J-wave syndromes include early repolarization syndrome and Brugada syndrome.[1] Repolarization and depolarization abnormality hypotheses have been proposed to explain the mechanism underlying ventricular arrhythmias in patients with J-wave syndrome. Depolarization abnormalities can be detected as delayed potentials on the epicardial surface in patients with J-wave syndrome, and elimination of abnormal potentials by radiofrequency catheter ablation normalizes electrocardiography (ECG) findings and suppresses ventricular arrhythmias.[2,3] Repolarization abnormalities explain the mechanism underlying J-wave syndrome through epicardial action potential changes and phase 2 reentry.[4] However, demonstrating repolarization abnormalities is difficult because there are few methods capable of clinically detecting them.[5,6] Here we present the case of a patient with a J wave in which ventricular fibrillation (VF) occurred via a phase 2 reentry mechanism.

Case report

A 54-year-old male patient experienced repeated VF episodes and frequent appropriate shocks from an implantable cardioverter-defibrillator (ICD). He was initially referred to our hospital at 34 years of age with ECG findings suggestive of Brugada syndrome and a family history of sudden cardiac death. His older brother had died suddenly at night and his father had died suddenly while napping. The ECG showed saddleback-type ST elevation and J waves in the high lateral ECG leads. He had type 1 ECG by the sodium channel blocker test, and VF was induced by programmed electrical stimulation. *SCN5A* gene test results were negative. The patient was asymptomatic but at risk of VF; therefore, we implanted a dual-chamber ICD (DDD 50 ppm). He was free of arrhythmic symptoms until 51 years of age, when he experienced palpitations and atrial fibrillation (AF) was detected. Subsequently, pulmonary vein isolation was performed. At the age of 54 years, he experienced headaches and palpitations and consulted his family physician. AF recurrence was detected, and his family physician prescribed 2.5 mg bisoprolol once daily. He took a bisoprolol tablet at 10 AM and took a nap. His wife witnessed his shock from the ICD and called an ambulance. He experienced recurrent VF episodes, and 12 appropriate shocks were delivered within an hour. An ECG showed AF and prominent J waves in the inferior and lateral leads; however, a coved-type ECG did not appear in the right precordial leads. Premature ventricular contractions (PVCs) occurred frequently and induced VF. The PVC morphology was a right bundle branch block with a superior axis, suggesting an origination from the posterolateral area of the left ventricle (LV) (Figure 1). Isoproterenol infusion immediately relieved the VF storm. The patient was transferred to our hospital 5 days after the initial VF episode. VF and PVCs did not recur after cessation of the isoproterenol. Because the

KEY TEACHING POINTS

- A patient in whom transient J waves appeared experienced electrical storm of ventricular fibrillation.
- Significantly long pauses during atrial fibrillation induce prominent J waves in the inferolateral leads and premature ventricular contractions that trigger ventricular fibrillation.
- Phase 2 reentry can trigger premature ventricular contraction.

KEYWORDS Brugada syndrome; Early repolarization syndrome; J-wave syndrome; Phase 2 reentry; Ventricular fibrillation
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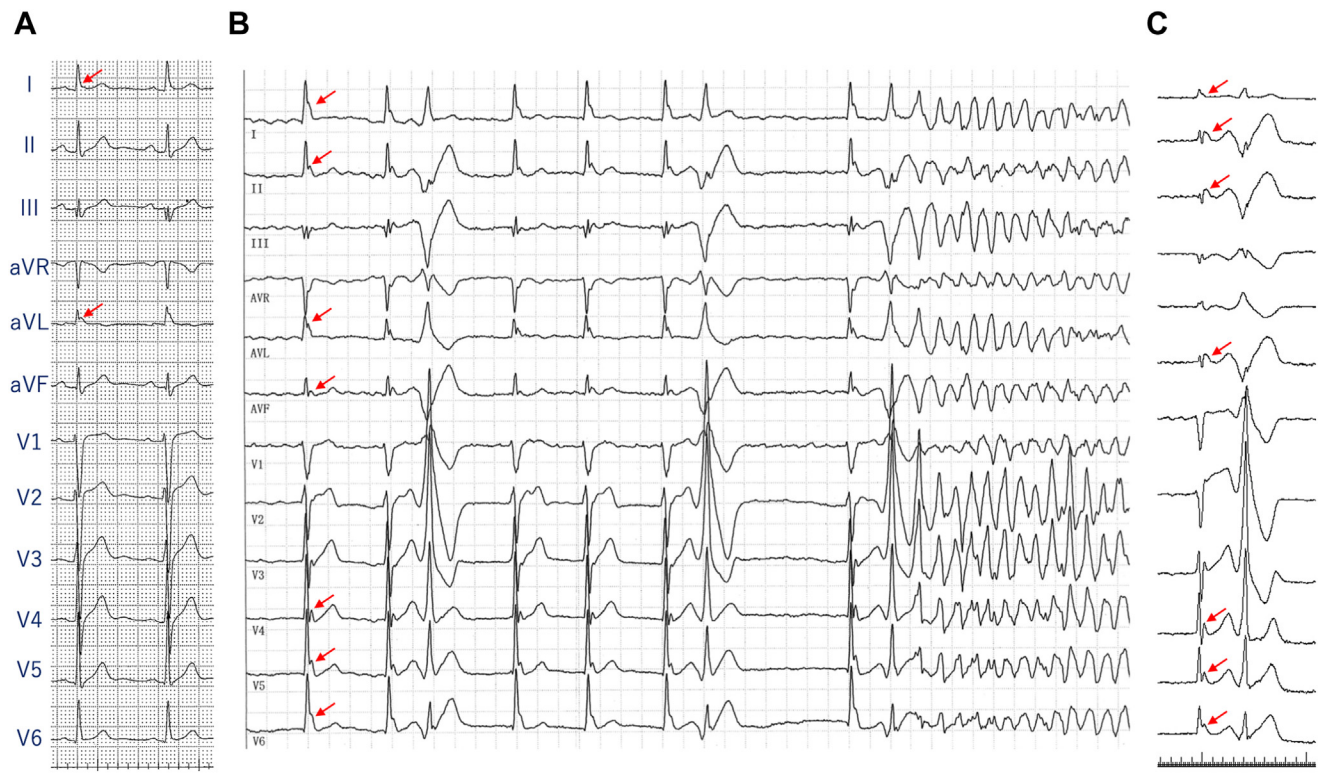

Figure 1 Twelve-lead electrocardiogram (ECG). **A:** The ECG was recorded at the patient's initial visit to the hospital. Slurred or notched J waves were observed in leads I and AVL (*arrows*). **B:** ECG taken at the onset of ventricular fibrillation (VF). Prominent J waves were observed in leads I, II, aVL, aVF, and V_4–V_6. The morphology of premature ventricular contractions (PVCs) was a right bundle branch block with a superior axis that arose from the posterolateral left ventricle. The same PVC initiated VF. **C:** A PVC was induced during an electrophysiological study. A long pause (7.4 seconds) after the injection of 20 mg adenosine triphosphate during atrial fibrillation induced trigger PVCs. A prominent J wave was visible in inferolateral leads (*arrows*).

PVC is a precursor of the VF, we planned catheter ablation of the PVC on day 10. Electrophysiological studies were performed under general anesthesia using a 3-dimensional mapping system (CARTO System; Biosense Webster, Inc, Diamond Bar, CA). A 6F 10-polar catheter was placed in the middle cardiac vein through the right jugular vein. A 2F 10-polar catheter was positioned in the left posterolateral vein through the right femoral vein. A ThermoCool catheter (3.5 mm tip, interelectrode spacing 2 mm, SmartTouch; Biosense Webster, Inc) with contact-force technology was used.

Large J waves were recorded at the catheter located in the posterolateral vein of the LV, whereas small J waves were observed at the catheter located in the middle cardiac vein (filter setting: 0.05–100 Hz). Low-frequency delayed potentials were recorded on unipolar and bipolar recordings (filter setting: 30–100 Hz) in the left posterolateral vein but not in the middle cardiac vein. The interval between the major ventricular (V) potential and low-frequency potential decreased during atrial pacing and was directly proportional to the cycle length of atrial pacing (Figure 2). To evaluate the delayed potential changes during bradycardia, we administered a bolus injection of 20 mg adenosine triphosphate (ATP). The pacing function was turned off during ablation to induce a long pause with ATP. The interval between the major V potential and the low-frequency potential increased as the V-V interval increased. Moreover, the low-frequency potential widened and became shallow at longer V-V intervals (Figure 3); however, the PVCs did not occur. As the patient experienced VF attacks during AF after bisoprolol intake, we induced rapid atrial pacing to induce AF after a 10 mg propranolol injection. We then administered 20 mg ATP during the AF rhythm. ATP-induced long pauses enhanced inferolateral J waves in 12-lead ECG and local J waves in the posterolateral vein and flattened delayed low-frequency potentials. The PVC induced after a long pause had the same morphology as the PVC that repeatedly triggered VF (Figure 1). Spiky prepotentials preceded PVC onset in the posterolateral vein. A small low-frequency delayed potential was recorded at the contralateral side of the endocardium; however, the PVC prepotential occurred earlier at the epicardial site in the posterolateral vein than in the endocardium (Supplemental Figures 1 and 2). Pace mapping was performed at the endocardial and epicardial sites of PVC origin (Supplemental Figure 3). The ablation catheter was placed on the contralateral side of the earliest activation site of the PVCs, which was the basal posterolateral left ventricle. Radiofrequency ablation was performed 12 times (output, 20 W; duration, 180 s/session) from the endocardial side. After ablation,

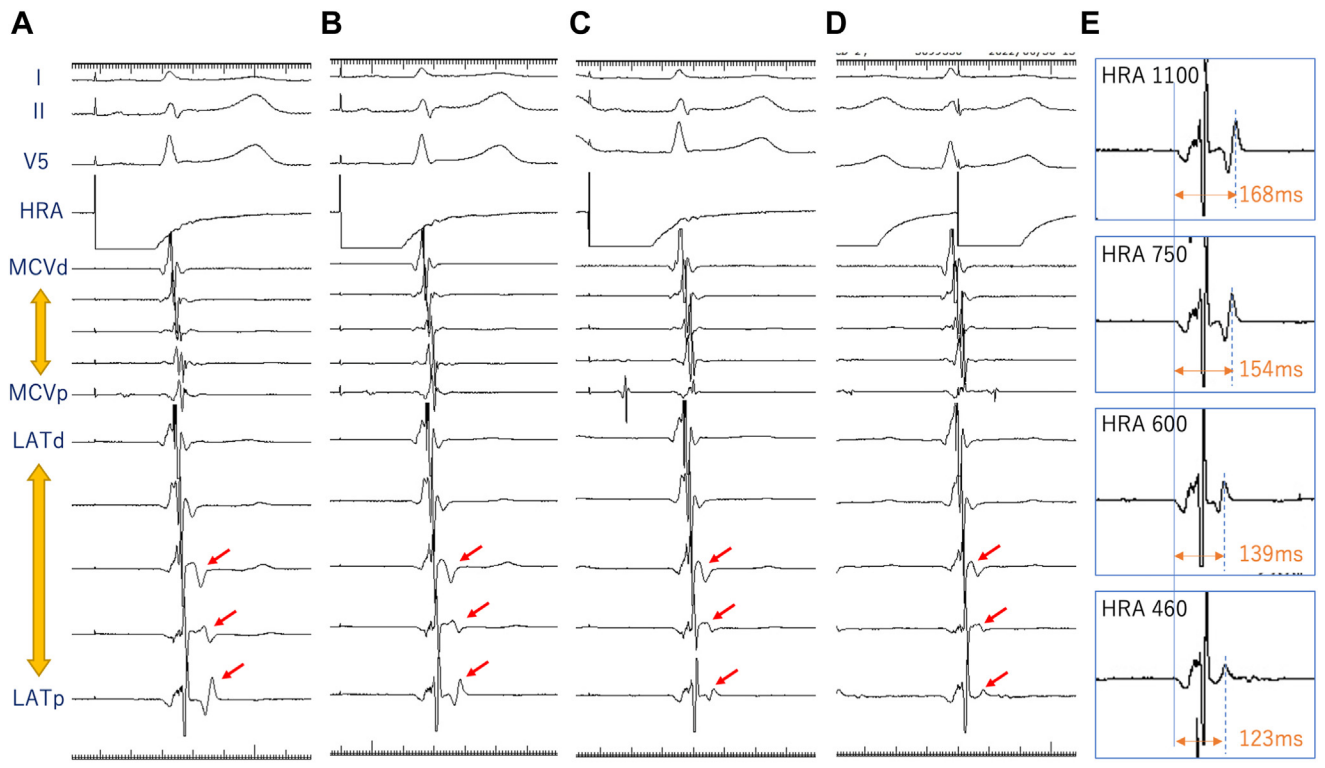

Figure 2 Dynamics of epicardial potential during tachycardia. Pacing in the high right atrium: **A:** cycle length (CL) = 1100 ms; **B:** CL = 750 ms; **C:** CL = 600 ms; **D:** CL = 460 ms; **E:** Magnification of the proximal portion of the posterolateral vein. Delayed low-frequency signals are visible at the electrodes in the posterolateral vein (*arrows*), but not at the electrodes in the middle cardiac vein. The delayed potentials approached the major ventricular potential according to the short-pacing CL. LATd = distal portion of the posterolateral vein; LATp = proximal portion of the posterolateral vein; MCVd = distal portion of the middle cardiac vein; MCVp = proximal portion of the middle cardiac vein.

ATP injection during AF did not trigger PVC. After eliminating the PVCs, re–pulmonary vein isolation of the left superior and right inferior PV was performed during the same procedure. The patient was discharged from our hospital without any medication and did not experience VF or AF recurrence for 1 year.

Discussion

The patient experienced VF storm associated with the transient appearance of inferolateral J waves. A long pause induced by ATP injection during AF triggered PVCs that arose from the epicardial side of the posterolateral LV. The low-frequency delayed potentials coincided with the focus of the PVCs. This delayed potential moved toward the major local V potential during tachycardia and grew distant from the major V potential owing to bradycardia. The delayed potential flattened during severe bradycardia and PVCs occurred at the peak of the flattened potential. Because epicardial puncture was not performed in this case, we could not precisely evaluate the epicardial potentials around the origin of the target PVCs. However, prepotentials preceding the onset of PVCs showed steep QS patterns at the Lat 8 and 9 electrodes on the unipolar lead, indicating that the catheters were positioned at the origin of PVCs (Figure 3 and Supplemental Figure 2). After ablation from the contralateral endocardial side of the focus, PVCs were not induced and VF did not recur after discharge. The relationship between the VF and autonomic nervous system activity in patients with J-wave syndrome has been reported previously. In this patient, VF occurred during a nap after taking beta-blocker, which was suspected to be owing to augmentation of parasympathetic tone.[7] As previously described, overdrive pacing at a slightly higher rate than the baseline rhythm could be useful in suppressing ventricular tachycardia/fibrillation if bradycardia or post-extrasystolic pauses facilitate the initiation of polymorphic ventricular tachycardia/fibrillation.[8]

J waves can arise from conduction abnormalities in the ventricle, whereas repolarization abnormalities can result in J waves in patients with early repolarization syndromes.[1–3] The J waves produced by conduction abnormalities become significant during a short cycle length.[9] In contrast, the J waves produced by repolarization abnormalities have the opposite characteristics.[10] According to the repolarization abnormality hypothesis, a deep phase 1 notch of the epicardial action potential results in a J wave in the ECG.[1,11,12] The upsloping limb of the deep phase 1 notch of the action potential can cause a low-frequency delayed potential.[13,14] A short pacing cycle

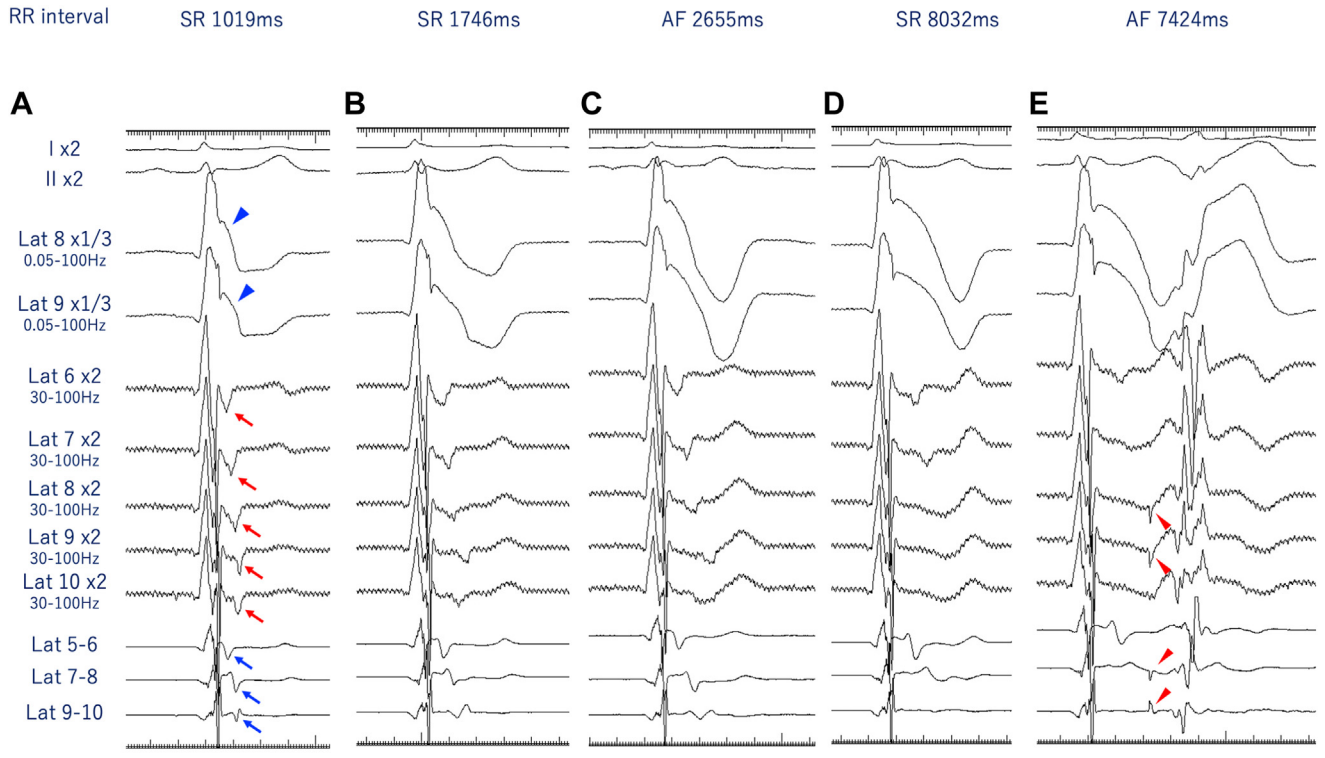

Figure 3 Dynamics of epicardial potential during bradycardia. **A:** Sinus rhythm (SR) with 1019 ms cycle length (CL). **B:** SR with 1746 ms CL. **C:** Atrial fibrillation (AF) with 2655 ms CL. **D:** SR with 8032 ms CL. **E:** AF with 7424 ms CL. Large J waves were observed in unipolar leads in the posterolateral vein with a filter of 0.05–100 Hz, while bradycardia increased the voltage and width (*blue arrowheads*). Delayed low-frequency potentials appeared in unipolar leads in the posterolateral vein with a filter of 30–100 Hz (*blue arrows*). Bradycardia delayed and flattened low-frequency potentials. Bipolar leads in the posterolateral vein also show delayed low-frequency potentials (*red arrows*). Potential changes after long pauses were more significant during the AF rhythm than the SR. Long pauses during AF induce premature ventricular complexes (PVCs), and a prepotential is observed at the electrodes in the posterolateral vein. The prepotential appeared at the bottom of the delayed potential (*red arrowheads*) and preceded PVC onset by 85 ms.

length shallows and abbreviates the phase 1 notch of the action potential, whereas a long pacing cycle length deepens and widens the phase 1 notch of the action potential.[14] The deepening of the phase 1 notch results in a loss-of-dome-type action potential and induces action potential heterogeneity. Action potential heterogeneity of the epicardium promotes phase 2 reentry.[1,14,15] The appearance of a loss-of-dome-type action potential can shallow the delayed potential. The low-frequency potential recorded in the epicardium in the present case had the same characteristic response to cycle length change as the abnormal potentials associated with repolarization abnormalities (Supplemental Figure 4). The observations in this case did not provide direct evidence of phase 2 reentry, but the dynamics of the epicardial delayed potential indicated that the PVC was promoted by phase 2 reentry.

Spontaneous PVCs appeared only during the VF storm when prominent inferolateral J waves appeared. Isoproterenol eliminates J waves and PVCs. Exercise testing, intracoronary acetylcholine injection, and intravenous propranolol did not induce PVCs. A bolus injection of 20 mg ATP administered during AF could induce PVCs. A longer R-R interval enhanced the inferolateral J waves associated with the focus of the PVCs. Because there are no definite induction methods for PVC-triggered VF in J-wave syndrome, the induction method used in the present case could be a clue for the detection of PVCs, which are precursors of VF. A long pause induced by ATP during sinus rhythm did not induce PVCs. Rapid atrial pacing for AF induction and R-R irregularities during AF may influence PVC inducibility.

Conclusion

A patient who underwent ICD implantation 20 years prior experienced a VF storm. The PVCs that triggered VF appeared only during the VF episodes and were associated with the transient appearance of inferolateral J waves. Triggering of PVCs could be induced by significantly long pauses by ATP during AF. Trigger PVCs are associated with an abnormally delayed potential in the epicardium, and phase 2 reentry is a possible mechanism of PVCs.

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Appendix

Supplementary Data

Supplementary data associated with this article can be found in the online version at https://doi.org/10.1016/j.hrcr.2023.06.008.

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