Storms of Ventricular Fibrillation Responsive to Isoproterenol in an Idiopathic Ventricular Fibrillation Patient Demonstrating Complete Right Bundle Branch Block

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Summary

A 45-year-old male was admitted to our hospital after successful resuscitation of cardiac arrest. Ventricular fibrillation (VF) had occurred during breakfast and was defibrillated by an automated external defibrillator operated by emergency medical service staff. On admission, his ECG demonstrated complete right bundle branch block as the sole abnormality. Intensive examination could not detect any structural disease leading to a diagnosis of idiopathic VF and implantation of an ICD. VF storm occurred one month after hospital discharge and beta-blocker, amiodarone, and sedative administration had no effect on VF. Likewise, catheter ablation for triggering premature ventricular beats failed to control the VF storm. The VF storm then subsided in the following weeks and the patient was discharged on amiodarone. A half month later VF storm recurred and the patient was admitted again. This time, isoproterenol infusion was effective in suppressing VF, and thereafter the patient was administered bepridil and followed up without recurrence of VF for 1.5 years. From these beneficial effects, the VF of the patient was suggested to share common arrhythmogenic characteristics to those of Brugada syndrome or J-wave associated VF. (Int Heart J 2013; 54: 240-242)

Key words: Sudden death, Implantable cardioverter defibrillator, Electrocardiogram, Catheter ablation, Electrical storm, Bepridil

Idiopathic ventricular fibrillation (IVF) is an occurrence of ventricular fibrillation (VF) in patients without structural heart disease. Among the types of IVF, Brugada syndrome is recognized as a distinct clinical entity. Some IVF might be associated with early repolarization, especially in the case of J waves. Both show some characteristic ECG phenotypes, but it has been reported that these ECG findings may be masked by complete right bundle branch block (CRBBB). Here, we report the case of an IVF patient who presented with CRBBB as the sole abnormality and developed VF storms. VF was well controlled by isoproterenol suggesting a close relation to the IVF caused by early repolarization.

Case Report

A 45-year-old male was found to have CRBBB at his annual health check in 2011. His past history was non-contributory, with no familial member believed to have died from sudden cardiac death. On July 13, 2011 the patient collapsed during breakfast and his wife immediately called an ambulance. On arrival of the emergency medical service, his conscious level was JCS300 and VF was confirmed using an automated external defibrillator. He was defibrillated by the external DC defibrillator 3 times and resuscitated. He was then admitted to hospital where emergency cardiac catheterization revealed neither coronary artery stenosis nor provocation of coronary spasm by acetylcholine performed on another day. After admission, he was intubated and put under mild hypothermia. During hypothermia, VF reoccurred and was terminated by external DC shock. He recovered in a few days without any neurological deficit. Cardiac echocardiography, RI scintigraphy, MRI, and exercise stress test results were all normal. He was transferred to our hospital on August 5, 2011 for implantation of an implantable cardioverter defibrillator (ICD).

On admission to our hospital, his ECG showed CRBBB (Figure 1A). After obtaining written informed consent, an ICD was implanted on August 6, 2011 (Figure 1B). During the positioning of the RV lead, premature ventricular contractions (PVCs) were induced which led to VF and required termination by external DC shocks. Following device implantation his clinical course was uneventful and he was discharged on August 11.

VF storm (2nd admission to our hospital): On the morning of September 17, 2011, the first ICD shock occurred while he was sitting down on his bed after returning from the bathroom.
The very next morning, he received another shock while attending a business meeting. During transport and upon arrival at our hospital, VF recurred and a total of 10 ICD shocks were discharged. ICD interrogation revealed VF episodes that were triggered by PVCs. Beta-blocker, amiodarone, and sedative administration failed to suppress the PVCs and VF storm, however, we were able to confirm VF-triggering PVC morphology by 12-lead ECG recording after admission (Figure 2A).

**EPS/ablation:** Emergency catheter ablation was performed on September 19, 2011 targeting the VF-triggering PVCs that were shown to be uniform via 12-lead ECG recordings. We performed 3-dimensional electro-anatomical mapping using the CARTO XP EP Navigation System (Biosense Webster, CA, USA). Voltage mapping did not indicate the existence of a pathological area, and the targeted PVCs were mapped and ab-
Figure 4. Clinical course of the patient. VF recurred in electrical storm (>4 times per day). A short acting beta-blocker (rudiolol) and intravenous amiodarone were not effective and VF still occurred after sedation by propofol. After catheter ablation was attempted, the patient was followed by oral amiodarone but VF recurred again. On the 3rd admission, isoproterenol (ISP) infusion was given immediately followed by oral bepridil, which was effective in preventing VF.

Although the catheter ablation was deemed a failure, VF disappeared after the procedure. The patient was discharged October 3 on oral amiodarone at a dose of 200 mg.

Recurrence of VF: On October 7, 2011, the patient experienced ICD shocks while sitting on his bed. Amiodarone was considered to be ineffective. We started infusion of isoproterenol (ISP) although ECG morphology did not show evidence of Brugada type ECG or J waves. While his heart rate increased, ISP infusion did not influence QRS morphology (Figure 3). ISP successfully suppressed VF, and it was given for 5 days followed by oral bepridil (200 mg/day). The patient was discharged from our hospital on October 16 on bepridil and has not had any recurrence of VF during 1.5 years of follow-up (Figure 4).

Summary: An ICD was implanted in a patient with IFV who did not show any typical Brugada-like or J wave ECG phenotypes with the exception of the CRBBB pattern. The patient developed VF storm, and amiodarone, beta-blocker, and sedative agent administration had no suppressive effect. Catheter ablation was considered to be a failure because VF was easily triggered by PVCs from multiple sites which were induced during catheter manipulation. However, the 2nd episode of VF storm was completely suppressed by ISP infusion and long-term oral bepridil therapy appeared to be effective for controlling VF.

**DISCUSSION**

Some IFV patients are known to have characteristic ECG patterns, and IFV has been re-categorized into Brugada syndrome, J wave syndrome, and others.

In the present patient, there was no underlying cardiac disease and the CRBBB pattern was the sole ECG anomaly. A diagnosis of Brugada syndrome was not supported from documented ECGs, including those recorded at higher intercostal spaces. However, the patient was found to share common pharmacological characteristics: responsiveness of VF to isoproterenol which is established as an effective drug for VF storm in Brugada syndrome and J-wave associated IFV patients. ISP acts as a beta-agonist and increases inward Ca current, restoring the loss of the action potential dome at the epicardial region of the ventricular wall. Cilostazol acts similarly to isoproterenol in Brugada syndrome and is often prescribed. Quinidine or bepridil were shown to block Ito currents and are also effective in Brugada syndrome. Bepridil seemed to be effective in controlling VF in the present patient.

More recently, several investigators have reported that CRBBB masks the Brugada type ECG. It is very reasonable to assume that J-wave associated IFV might also be masked by CRBBB. Therefore, an important step may be to verify whether resolution of CRBBB can reveal J waves or not.

**Conclusion:** We have described a patient with CRBBB who had recurrent VF storms. No ECG phenotypes for Brugada syndrome or J-wave associated IFV were evident because of CRBBB. However, the fact that isoproterenol infusion and oral bepridil therapy was so effective in controlling VF storms may suggest a common underlying arrhythmogenic substrate caused by abnormal repolarization.

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**REFERENCES**