Successful Radiofrequency Catheter Ablation for Electrical Storm of Ventricular Fibrillation in a Patient With Brugada Syndrome

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The case of a 41-year-old man with Brugada syndrome (BS) who suffered electrical storms (ES) of ventricular fibrillation (VF) is presented. Although intravenous infusion of isoproterenol (ISP) suppressed the VF occurrence, he consistently experienced recurrence of VF following discontinuation of ISP infusion. Quinidine and cilostazol were ineffective. An analysis of VF episodes on electrocardiogram monitoring revealed that the QRS morphology of the first beat of all VF episodes was identical to that of premature ventricular complexes (PVCs) with a left bundle branch-block morphology and inferior axis, which occurred repetitively before the episodes of VF and were recorded throughout the day. In addition, stored electrograms from the implantable cardioverter defibrillator showed that the first beat of all VF episodes had the same morphology. On electrophysiological study, the VF-triggering PVC was found to originate from the posterior portion of the right ventricular outflow tract area and their elimination, which was achieved with radiofrequency catheter ablation (RFCA), resulted in the suppression of ES. Although several other PVCs were still observed, the patient has been free of VF during the 29-month follow-up period. This case indicates that RFCA of VF-triggering PVCs may be useful in the treatment of drug-resistant ES in patients with BS. (Circ J 2008; 72: 1025–1029)

Key Words: Brugada syndrome; Electrical storm; Radiofrequency catheter ablation; Right ventricular outflow tract; Ventricular fibrillation

Brugada syndrome (BS) was reported for the first time in 1992 and is associated with a characteristic ST-segment elevation in the right precordial leads (V1–V3) in the absence of any demonstrable structural heart disease, as well as sudden cardiac death due to ventricular fibrillation (VF). Implantable cardioverter defibrillators (ICD) have been used to prevent sudden death in patients with BS. Recently, isoproterenol (ISP), oral quinidine bisulfate, and cilostazol have been reported to be effective in suppressing VF storms although there have been few reports on the efficacy of radiofrequency catheter ablation (RFCA) for the treatment of drug-resistant electrical storms (ES). In this report, we present a unique patient with BS who suffered drug-resistant electrical VF storms, in whom RFCA for the VF-triggering premature ventricular complexes (PVCs) successfully suppressed the recurrence of VF.

Case Report

A 41-year-old man with BS who suffered frequent episodes of VF was admitted to our hospital in June 2005. At 37 years of age, he experienced 2 syncopal episodes with generalized convulsions while he was watching television and driving a car. The diagnosis of BS was then established after documentation on a 12-lead electrocardiogram (ECG) showing ST-segment elevation (coved type) in lead V1, and excluding structural heart disease. He had no family history of BS or sudden cardiac death. He was implanted with an ICD (GEM-II DR 7273, Medtronic, Inc, Minneapolis, MN, USA) in our hospital on November 7, 2001. After discharge, the patient experienced 2 ESs of VF (defined as more than 3 episodes of VF within 24h) in June 2002 and March 2005, each of which was successfully treated with continuous infusion of ISP.

He subsequently experienced 4 ICD shocks due to recurrence of VF all in the same week, and was readmitted to hospital on June 4, 2005. An initial physical examination revealed normal findings and laboratory data were within normal limits. A 12-lead ECG revealed ST-segment elevation (coved type) in lead V1 with a J-point amplitude of 0.27 mV (Fig 1A). ISP was administered to prevent the recurrence of VF. Continuous intravenous infusion of ISP at 0.24 g/min normalized the ST-segment elevation (Fig 1B) and suppressed the occurrence of PVCs and VF. However, 10 min after infusion was discontinued, repetitive PVCs quickly recurred, followed by VF. Cilostazol, a phosphodiesterase type III inhibitor, which is reported to have suppressed ES in a patient with BS was given at a dose of 200 mg/day in combination with continuous infusion of ISP. Yet, it still could not prevent the recurrence of PVCs and VF following a reduction of ISP infusion. Quinidine bisulfate (600 mg/day), which is reported to have been useful in the treatment of ES in patients with BS also failed to prevent
Fig 1. (A) Standard 12-lead electrocardiogram (ECG) during sinus rhythm. Coved-type ST-segment elevation (arrow) is observed in lead V1 with a J-point amplitude of 0.27 mV. (B) A 12-lead ECG with normalized ST-segment in lead V1 after continuous infusion of isoproterenol.

Fig 2. (A) Electrocardiogram (ECG) recording of ventricular fibrillation (VF) initiation on the heart monitor. The QRS morphology of the first beat of VF (black arrow) is identical to those of the premature ventricular complexes (PVCs) (white arrow) that occurred shortly before the episode of VF. (B) ECG recording on the day before the episode of VF. Isolated PVCs with the same pattern of QRS morphology (white arrow) occurred repetitively with a late coupling interval of 390 ms. (C) Representative 12-lead ECG of the PVCs observed in (B). The PVCs exhibit left bundle-branch block morphology and inferior axis.
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Fig 3. Stored intracardiac electrograms in the implantable cardioverter defibrillators at the onset of ventricular fibrillation (VF). Three episodes are presented. The morphology of preceding premature ventricular complexes (white arrow) is identical to that of the first beat of VF (black arrow).

Fig 4. (A) Right (30°) and left (45°) anterior oblique (RAO and LAO, respectively) fluoroscopic images of the site of successful ablation. The ablation catheter (abl, white arrow) is located in the posterolateral portion of the right ventricular outflow tract area. (B) Intracardiac electrogram at the site of successful ablation. The local ventricular activation recorded by the distal electrode pair of the abl preceded all bipolar recordings from the basket (A,B,C,D) and coronary sinus (CS) catheters and the onset of the QRS complex by 44 ms. (C) The 12-lead electrocardiograms of ventricular fibrillation (VF)-triggering premature ventricular complexes (Left) and pace mapping (Right) at the site of successful ablation. Perfect pace mapping was obtained at this site. Basket, basket catheter; HIS, His-bundle catheter.
VF recurrence.

ECG recordings obtained from the heart monitor revealed that the QRS morphology of the first beat in all episodes of VF was identical to that of the PVCs that occurred repetitively before the episodes of VF (Fig 2A). The PVCs were recorded throughout the day with late coupling intervals ranging from 390 to 450 ms (Fig 2B), and 12-lead ECG revealed that the PVCs had a left bundle-branch block morphology and inferior axis, and appeared to originate from the right ventricular outflow tract (RVOT) area (Fig 2C). A total of 9,046 PVCs with the same morphology was observed on the 12-lead 24 h-Holter recording and accounted for 46% of all the PVCs during the whole day, and 285 episodes of non-sustained ventricular tachycardia (NSVT) of up to 6 beats were detected despite the continuous infusion of ISP. In addition, 215 out of 285 NSVT episodes had changing QRS morphologies and included at least 1 beat with the same morphology as the PVCs from the RVOT. Moreover, intracardiac electrograms stored in the ICD also suggested that the QRS morphology of the preceding PVCs was identical to that of the first beat in all VF episodes (Fig 3).

After obtaining written informed consent, we performed an electrophysiological study and catheter ablation for the VF-triggering PVCs. Under fluoroscopic guidance, a 5-Fr decapolar catheter (Irvine Biomedical Inc, Irvine, CA, USA), a multi-electrode basket catheter (FGM Inc, Tochigi, Japan) and a 2.5-Fr micro-sized mapping catheter with 16 electrodes (Cardima Inc, Benicia, Fremont, CA, USA) were introduced into the His bundle region, RVOT area and coronary sinus, respectively, via the femoral veins. Detailed activation mapping of the PVCs and pace mapping were performed in the right ventricular (RV) outflow and inflow tract area using a 7-Fr quadripolar ablation catheter with a 4-mm distal electrode, an embedded thermistor and a deflectable tip (EP Technologies, San Jose, CA, USA). A total of 8 types of PVCs from the RVOT area and 3 types from the RV inflow area were observed during the study. PVCs with morphology identical to the VF-triggering PVCs occurred most frequently. The coupling interval of the VF-triggering PVCs was 453±18 ms, whereas that of non-VF triggering PVCs from the RV inflow area was 464±29 ms. The earliest ventricular activation during the VF-triggering PVCs was found in the posterior portion of the RVOT lateral wall (Fig 4A). The intracardiac electrogram at this site during the PVCs preceded the onset of the QRS complex by 44 ms and revealed no abnormal morphology, such as fragmented potentials or double potentials (Fig 4B). At this site, perfect pace mapping was obtained (Fig 4C). Just after the application of radiofrequency current at this site, the VF-triggering PVCs were eliminated. After confirming the lack of recurrence of the VF-triggering PVCs and VF episodes even after discontinuation of ISP infusion for 1 h, we attempted to ablate the other PVCs from the RVOT and RV inflow tract. Finally, 3 types of PVCs from the RVOT and 2 types from the RV inflow tract remained after this treatment.

A 12-lead 24 h-Holter recording performed 6 days after the catheter ablation revealed that PVCs from the RV inflow tract area accounted for nearly all of 735 PVCs per day. One PVC from the RVOT, which was different from the VF-triggering PVC, and 1 episode of NSVT originating from the left ventricle were observed.

The patient has experienced no recurrence of VF during follow-up for 29 months. Several 24 h-Holter monitoring procedures have revealed that the number of PVCs has declined after catheter ablation, with 36 and 16 PVCs in 1 day at 2 and 6 months after catheter ablation, respectively. In addition, on ambulatory 12-lead 24 h-Holter recording at the 12-month visit, only PVCs from the RV inflow area were observed, without the occurrence of PVCs from the RVOT area.

Discussion

The major finding in this case is that RFCA for VF-triggering PVCs originating from the RVOT area successfully suppressed ES of VF in a patient with BS, despite the persistence of other PVCs originating from the RVOT and RV inflow tract areas.

BS is a clinical entity characterized by a coved- or saddle-back-type ST-segment elevation in the right precordial ECG leads in the absence of any structural heart disease, and episodes of VF are associated with sudden cardiac death.1,2-11 Studies of a recent experimental model developed by Yan and Antzelevitch, using arterially perfused canine RV wedge preparations, suggested that an increase in a transient outward current (Ito)-mediated phase 1 notch and a subsequent loss of the action potential dome in the RV epicardium, but not the endocardium, produces a transmural voltage gradient, resulting in a ST-segment elevation in the right precordial leads, and that heterogeneous loss of the action potential dome in the epicardium causes phase 2 re-entry, giving rise to VF.12,13 ISP has been shown to restore the epicardial action potential dome, eliminate the ST-segment elevation, and prevent the genesis of VF by increasing inward calcium current (Ica), in experimental models and patients with BS.8,13-16 Quinidine, a class I anti-arrhythmic agent with I0 blocking properties, has been shown to restore the epicardial action potential dome and normalize ST-segment elevation, and thus prevent phase 2 re-entry and VF in experimental models of BS.13 The effectiveness of quinidine in treating ES in patients with BS has been indicated in some reports.2-3 Cilostazol, an oral phosphodiesterase type III inhibitor that increases Ica, is also reported to have suppressed the frequently occurring VF episodes in a patient with BS.4 In the present case, ISP successfully normalized the ST-segment and suppressed the occurrence of VF. However, quinidine and cilostazol were ineffective in suppressing VF. The markedly increased Ica induced by ISP, followed by restoration of the epicardial action potential dome and elimination of ST-segment elevation, appeared to play an important role in the prevention of the VF-triggering PVCs and the recurrence of VF in our patient.

Some reports have described the mode of VF onset in patients with BS.6,13 and catheter ablation for triggering PVCs resulting in suppression of VF recurrence.5,6,13 These reports showed that the QRS morphology of the first beat of VF episodes was identical to that of the PVCs recorded before such episodes occurred.5,6,10 Haissaguerre et al reported successful catheter ablation for triggering PVCs for treatment of recurrence of VF in 3 symptomatic patients with BS.6 Two of the patients in their study had triggering PVCs with left bundle-branch block morphology and inferior axis originating from the RVOT area, and the other had triggering PVCs with left bundle-branch block morphology and superior axis originating from the anterior RV Purkinje network.5 The elimination of these VF-triggering PVCs, which was achieved with catheter ablation, resulted in suppres-
sion of VF recurrence for 7 months. Darmon et al reported one symptomatic patient in whom catheter ablation for triggering PVCs and 2 other types of PVCs originating from the RVOT area resulted in suppression of VF for 6 months.6

In the present case, we found that the VF-triggering PVCs originated from the posterior portion of the RVOT lateral wall. Despite the remaining PVCs originating from the RVOT and RV inflow tract areas, successful catheter ablation for the VF-triggering PVCs prevented the recurrence of VF for 29 months, the longest event-free period yet reported. Yan and Antzelevitch suggested that phase 2 re-entry in the RV free wall served as the trigger for VF.12,13 Morita et al demonstrated that ventricular arrhythmia was induced more frequently in the free-wall region of the RVOT area than in other ventricular regions in patients with Brugada-type ECG.16 These findings suggest that PVCs originating from the RVOT free wall may play an important role in initiating VF episodes in patients with BS.

Although we found no characteristic differences between VF-triggering and non-triggering PVCs, such as coupling intervals or number of successive beats, the detection of VF-triggering PVCs using ECG monitoring and 12-lead ECG played a very important role in the success of our catheter ablation. The Holter monitoring revealed that catheter ablation eliminated NSVT, as well as the VF-triggering PVCs from the RVOT, and that any remaining PVCs from the RV inflow tract area could not serve as the trigger for VF, suggesting that PVCs from the RVOT might have some effect on the VF-maintaining substrate and that catheter ablation modified it. Thus, catheter ablation for VF-triggering PVCs may be an alternative method of preventing VF in patients with BS. Identifying the characteristics of VF-triggering PVCs in more cases will improve the success of catheter ablation.

In conclusion, catheter ablation for VF-triggering PVCs was performed for treatment of drug-resistant ES and completely suppressed the recurrence of VF in a patient with BS.

References