Catheter ablation of ventricular ectopy originating from the left fascicular conduction system triggering polymorphic ventricular tachycardia in Brugada syndrome

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Introduction

In patients with no evidence for structural heart disease, idiopathic ventricular fibrillation and polymorphic ventricular tachycardia (PVT) are frequently triggered by ventricular premature beats (VPBs) arising from the Purkinje system or the right ventricular outflow tract that can be successfully eliminated by radiofrequency (RF) catheter ablation.[1,2] Such triggers of malignant arrhythmias have also been demonstrated and successfully eliminated in patients with Brugada, long QT, and early repolarization syndromes.[3–5] We report a case involving a patient with Brugada syndrome in whom malignant PVT was eliminated by RF ablation of VPBs as a trigger originating from the left fascicular conduction system.

Case report

Work on this case was done at Department of Cardiology and Angiology, Augusta-Kranken-Anstalt, Bochum, Germany.

A 17-year-old female patient with a history of recurrent syncope since the age of 9 years had no evidence for structural heart disease. Holter monitoring showed massive repetitive ventricular salvoes as well as rapid PVT episodes at a maximum duration of 12 seconds (Figure 1A), the latter associated with syncope. Repeated recordings of the 12-lead electrocardiogram (ECG) revealed sinus beats with a normal QRS complex and short-coupling VPBs of right bundle branch block pattern with a right superior or inferior axis as a trigger for complex ventricular arrhythmia. The challenge test revealed a diagnostic Brugada ECG after intravenous administration of 40 mg ajmaline (Figure 1B) associated with a marked aggravation of ventricular arrhythmia.

KEYWORDS Brugada syndrome; Monomorphic ventricular ectopy; Polymorphic ventricular tachycardia; Radiofrequency ablation; Syncope
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Mapping and catheter ablation procedures

An electrophysiological study was performed using 3 multipolar catheters with 10 mm interelectrode spacing (Daig, St. Jude Medical, St. Paul, MN). A quadripolar catheter was used for stimulation and recording from the right ventricular apex. Two hexapolar catheters were positioned within the coronary sinus for left atrial activity recording and across the tricuspid valve for His bundle (distal pair) and low right atrial (proximal pair) activity recording. A steerable quadripolar 7F catheter with a 4 mm-tip electrode (Marinr, Medtronic, Minneapolis, MN) was used for retrograde transaortic mapping and temperature-guided RF ablation.

During electrophysiologic study, sinus beats showed an atrial-His interval of 100 ms and a prolonged His-ventricular interval of 70 ms. Sharp presystolic fascicular potentials (P) were reproducibly recorded in the distal and proximal pairs of the ablation catheter (Abl) in both sinus beats and VPBs (Figure 2A–C) during mapping the left ventricular mid septum in an area of the presumed distal left anterior fascicle (LAF) (Figure 2D). During sinus beats, these P potentials as a result of their anterograde activation from the His bundle (H) showed an HP interval of 30–35 ms and were closely followed by local ventricular electrogram (V) with a PV interval of 10–15 ms. During VPBs at prematurities (P-Pe) of 315–265 ms, these potentials (Pe) activated the His bundle retrogradely with a Pe-He delay of 20–40 ms and preceded the QRS complex by 30 ms (Figure 2A). As shown, these VPBs were narrow (115–120 ms). At local prematurities $\leq$255 ms, no retrograde His bundle activation could be observed (Figure 2B and C). During VPBs, the more marked the increase in prematurity of local triggering Pe potentials, the more a prolonged delay of both subsequent local ventricular activation (Pe-V) and the QRS complex (Pe-QRS) could be observed. On the surface ECG, this conduction delay resulted paradoxically in longer coupling intervals of VPBs, masking the true ectopic prematurity (Figure 2C). The short local VPB prematurity ($\leq$255 ms), regardless of local conduction delay, was usually associated

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KEY TEACHING POINTS

- Syncope or cardiac arrest associated with malignant ventricular arrhythmias may occur with a normal QRS complex during sinus rhythm and no evidence for structural heart disease.
- In idiopathic malignant ventricular arrhythmias, a careful search for primary channelopathies and ectopic triggers is essential.
- The diagnosis of unmasked Brugada syndrome points out the need for performing the challenge test and ventricular premature beats identifiable as triggers can be successfully ablated.

with both the QRS widening and variable QRS morphology with right superior (Figure 2B) or right inferior (Figure 2C) axis, as well as an increase in complexity of ventricular arrhythmia, the latter triggering self-terminating PVT (Figure 2C). It is of note that a subsequently initiated ventricular echo beat in Figure 2B demonstrated a recordable His bundle potential activated retrogradely. Pace mapping performed at this site demonstrated a nearly perfect match for the narrow VPBs (Figure 2E). During ablation, 4 RF applications for 20–30 seconds resulted in transient rapid or accelerated ventricular runs, progressive arrhythmia suppression, and transient prolongation of local PV time. Ultimately, after the fifth RF application, complete arrhythmia elimination could be achieved. Postablation, a persistent prolongation of the local PV time (35 ms) and no change in both the HV interval and the QRS morphology during sinus rhythm were observed (Figure 2F). A subcutaneous implantable cardioverter-defibrillator (ICD) was implanted in view of Brugada syndrome and risk of sudden death. There was no recurrence of any ventricular arrhythmia (Figure 3) during a follow-up period of 43 months.

Discussion

The major findings in this case with no evidence for structural heart disease and sinus beats with a normal QRST complex is that unifocal narrow VPBs demonstrating polymorphic QRS pattern triggering malignant PVT were successfully ablated from the distal left-sided fascicular conduction system in a patient in whom the challenge test unmasked a diagnostic Brugada ECG associated with aggravation of ventricular arrhythmia preablation. Generally, ventricular fibrillation is considered to be idiopathic after exclusion of structural heart disease and genetic disorders, including channelopathies.[1,2,6] Several cases with sinus beats showing a normal QRST complex on a 12-lead ECG and idiopathic PVT and/or ventricular fibrillation have been reported in which VPB triggers were successfully ablated.[7–10] In all but 1 of them, no challenge test was performed,[7–9] even though a novel *SCN5A* gene mutation was found in 1 patient.[9] In the remaining case, intravenous procainamide 1000 mg was administered for suppressing VPBs, but possible ECG changes suggestive of Brugada syndrome were not evaluated.[10] Otherwise, one may not exclude that malignant ventricular arrhythmias could be incorrectly classified as idiopathic.

This case demonstrates that a normal sinus rhythm may not necessarily exclude the diagnosis of Brugada syndrome and points out the need for performing the challenge test.

Nevertheless, for ventricular fibrillation, either idiopathic or occurring in Brugada, long QT, and early repolarization

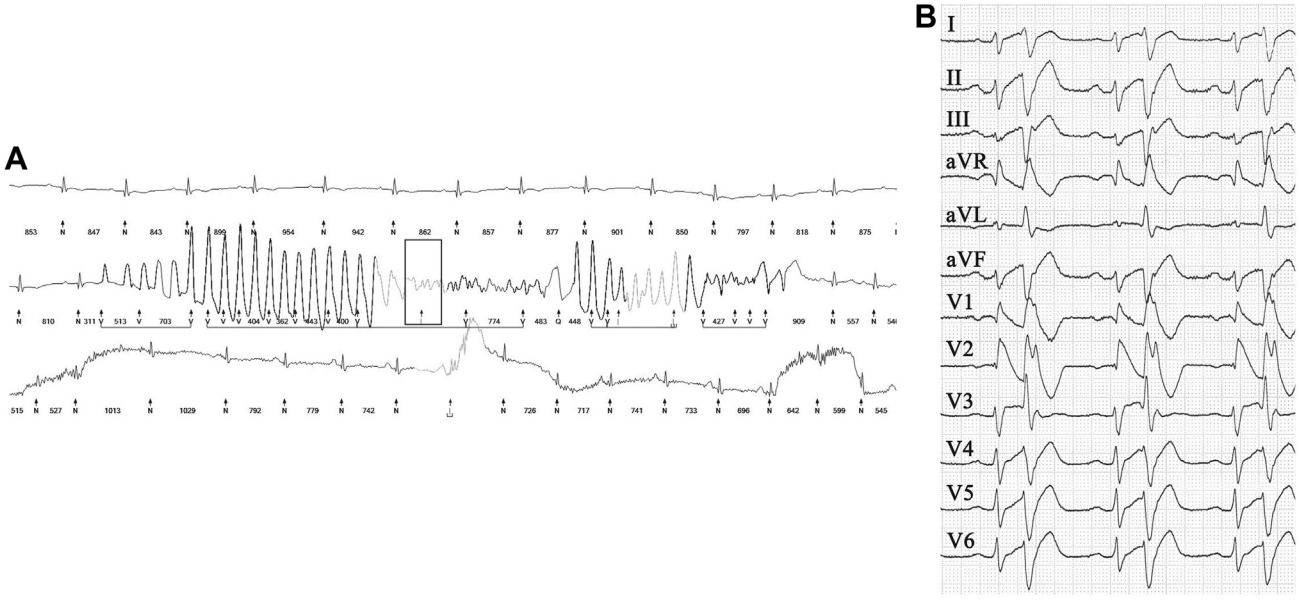

Figure 1 **A:** Polymorphic ventricular tachycardia associated with syncope. **B:** The challenge test (intravenous ajmaline 40 mg) revealing a diagnostic Brugada electrocardiogram with ventricular bigeminy.

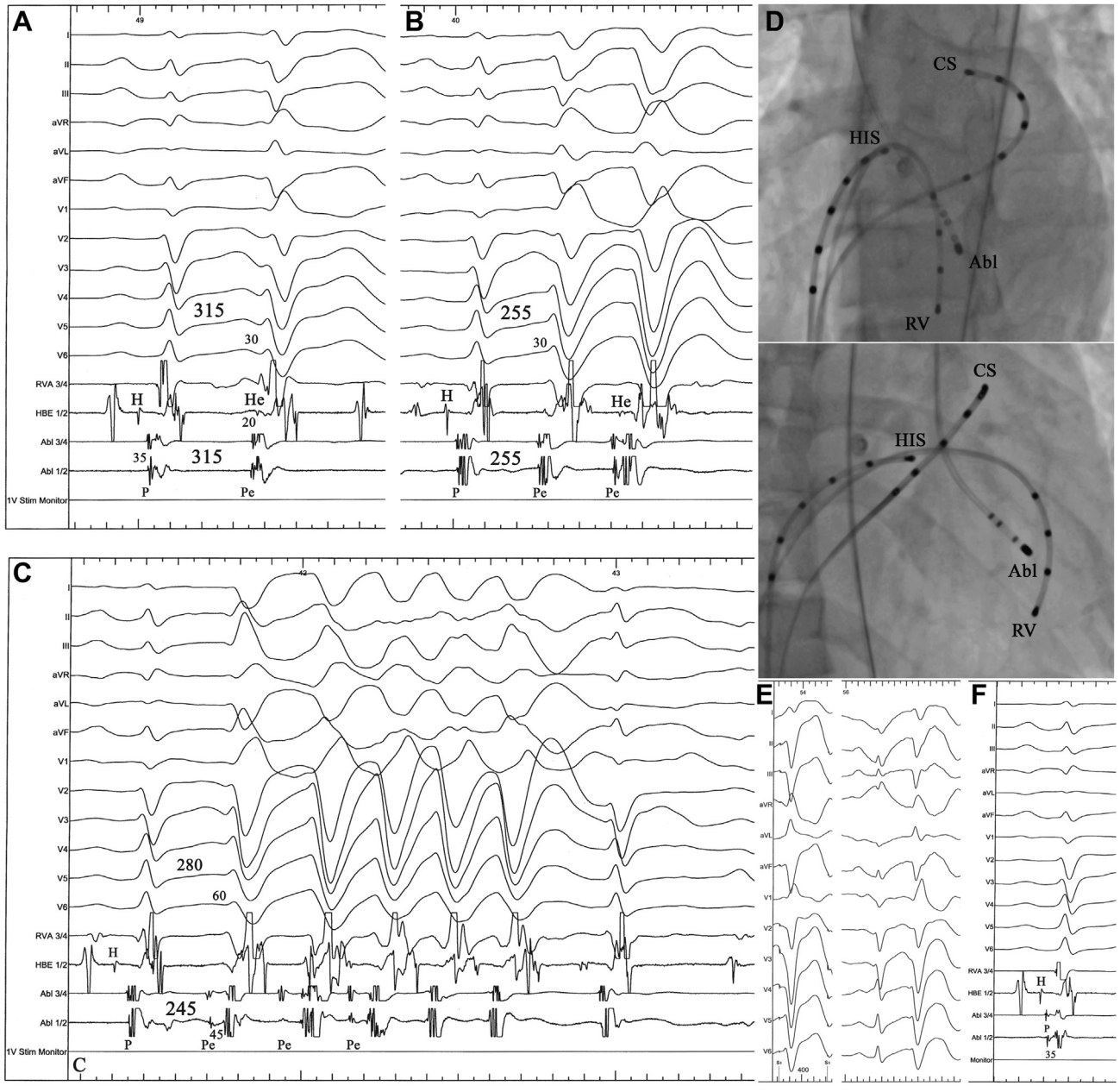

Figure 2 **A–C:** Intracardiac recordings showing sinus and ventricular premature beats. **D:** Left anterior oblique (*upper panel*) and right anterior oblique (*lower panel*) radiographs of the ablation catheter (Abl) positioned at the successful ablation site. **E:** Pace mapping showing a nearly perfect pace map obtained from the successful ablation site. **F:** Intracardiac recording showing a prolonged fascicular potential-ventricular interval (PV) of 35 ms postablation. CS = coronary sinus; H/He = His bundle potential; HBE = His bundle electrogram; HIS = His bundle catheter; P/Pe = fascicular potential; RV = right ventricle; RVA = right ventricular apex.

syndromes, VPBs identifiable as triggers can be successfully ablated from the right ventricular outflow tract and/or the right or left Purkinje conduction system.[1–5]

In 1 report, 2 cases were described in which idiopathic narrow VPBs in 1 case and narrow ventricular tachycardia in the other, originating from the proximal LAF, were eliminated by ablation, followed by development of LAF block.[11] VPBs of right bundle branch block pattern shown in Figure 2, narrow or widened with right superior axis (Figure 2A and B, respectively) and widened with right inferior axis (Figure 2C), were those documented clinically on the 12-lead ECG, as mentioned in the Case report section. During intracardiac mapping, these 3 distinct VPB forms were recorded with sharp presystolic fascicular potentials and eliminated at the same ablation site. This ablation site resembles that at the left anterior fascicle reported by Suzuki and colleagues[12] for idiopathic left ventricular fascicular tachycardia that initially presented with right bundle branch block morphology and left superior axis. In their report, the ablation catheter was located more proximal and ablation was successful after previous ablation sessions at the left posterior fascicle, inducing permanent left posterior fascicular block

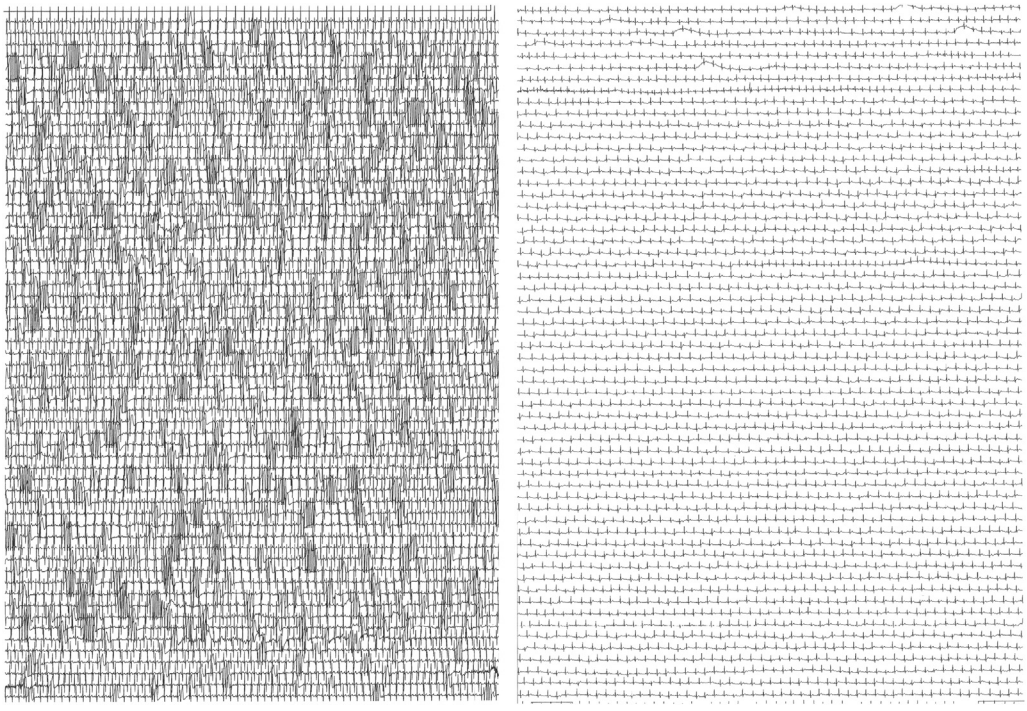

Figure 3 Electrocardiogram monitoring showing frequent ventricular salvoes (*left*) before and arrhythmia-free sinus rhythm (*right*) after ablation.

and the change in tachycardia morphology at the same cycle length. Regarding a trifascicular left-sided conduction system, it is likely in our patient that this ablation site is located at the left septal fascicle.[13] In this case with 3 distinct VPBs, the narrow VPB type with retrograde His bundle activation, morphologically resembling that described for left posterior fascicle ventricular tachycardia, may be a result of retrograde propagation up the septal fascicle and anterograde conduction down the left posterior fascicle. One may suggest that 2 other widened VPBs originating from the same focus are due to local myocardial conduction delay with propagation to the left posterior (Figure 2B) and left anterior (Figure 2C) fascicle regions. Though no retrograde His bundle activation was recorded, retrograde conduction up the septal fascicle reaching the upper turnaround in the vicinity of the left bundle branch followed by anterograde conduction over the left posterior and left anterior fascicles may be an alternative explanation for right bundle branch block pattern with right superior and right inferior axes in these VPBs, respectively. In support of this, an initiated ventricular echo beat was recorded in Figure 2B, showing a His bundle potential activated retrogradely.

In general, recurrent syncope associated with the diagnosis of Brugada syndrome would conventionally justify implantation of an ICD. In this case, elimination of focal ectopics triggering malignant ventricular arrhythmias associated with recurrent syncope previously mistaken for epilepsy resulted in a symptom-free state during a follow-up period of 43 months. In absence of Brugada syndrome, such cure would justify not implanting an ICD, not only in the short term but also in the long term. However, in 1 study after ablation of VPB triggers for idiopathic ventricular fibrillation, 18% experienced recurrence of ventricular fibrillation at a median of 4 months.[14] Our patient unequivocally presents the diagnosis of Brugada syndrome, with its unchanged arrhythmogenic substrate possessing marked ventricular vulnerability, as demonstrated preablation. This, along with a prolonged HV interval during sinus rhythm and positive late potentials, indicates that the young patient remains at risk of sudden cardiac death and justifies implantation of an ICD. ICD implantation is the most widely accepted first-line treatment in patients with channelopathies who have recurrent syncope and sustained ventricular tachyarrhythmias, including ventricular fibrillation.[6] Catheter ablation may reduce the risk of sudden death and decrease the frequency of cardiac events.

Conclusion

We report a case involving a patient with Brugada syndrome, in which malignant PVT was most likely eliminated by RF ablation at the distal left septal fascicle of VPBs as a trigger. Though the local fasciculoventricular interval prolonged by 20–25 ms postablation, no septal fascicular block developed. Preablation, our case demonstrated prematurity-dependent multiple QRS morphologies of triggering unifocal VPBs. Fortunately, pace maping produced a nearly perfect 12-lead ECG match for a narrow VPB form, which was additionally helpful for successful ablation. Otherwise, the remaining VPB morphologies would be important challenges for this mapping approach.

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