# Idiopathic Ventricular Fibrillation in a 10-Year-Old Boy: Technical Aspects of Radiofrequency Ablation and Utility of Antiarrhythmic Therapy

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*Idiopathic ventricular fibrillation (VF) is defined as spontaneous VF in the absence of structural heart disease. No prior reports exist addressing the technical aspects of idiopathic VF ablation in a child. We present the case of a 10-year-old boy with idiopathic VF, who presented a unique management challenge, particularly as regards the technical aspects of the ablation procedure. Ablation of idiopathic VF is feasible in a 10-year-old boy and oral quinidine seems more effective than other antiarrhythmic drugs in this condition. (PACE 2011; 34:e85–e89)*

***mapping, ablation, electrophysiology—clinical, pediatrics, VT***

## Introduction

Idiopathic ventricular fibrillation (VF) is defined as spontaneous VF in the absence of structural heart disease and identifiable electrophysiologic abnormalities,[1] likely first described by Dock[2] in 1929. Characterized by a high recurrence rate of VF,[3] the implantable cardioverter-defibrillator (ICD) has been shown to prevent sudden cardiac death in this condition.[4] An important trigger for idiopathic VF has been demonstrated to be a premature ventricular complex (PVC) originating predominantly in the distal Purkinje system of the right or left ventricle (LV), the ablation of which can significantly reduce or abolish recurrence of VF.[5] We present the case of a 10-year-old boy suffering from idiopathic VF, who, because of age and small body size, presented a unique challenge as regard the technical aspects of the ablation procedure.

## Case History

A 10-year-old boy presented for evaluation after an episode of syncope. Past medical history was only significant for mild asthma, and family history was negative for sudden cardiac death. Physical examination was normal. A 12-lead electrocardiogram demonstrated a normal QRS and QT/QTc durations. Serum electrolytes, an echocardiogram, and coronary angiogram were normal. A week later, he had a second episode requiring cardiopulmonary resuscitation and was found to be in AF with rapid ventricular response, requiring cardioversion. Cardiac and brain magnetic resonance imaging were normal. An electrophysiologic study showed normal baseline function and intervals. Polymorphic ventricular tachycardia (VT) was induced with programmed stimulation and an ICD was implanted. Genetic testing did not identify any long-QT syndrome variants. Due to frequent ICD shocks for recurrent VF, Sotalol was started, with no effect. Sotalol was switched to amiodarone and a $\beta$-blocker without success. Holter monitoring revealed frequent PVCs that triggered VF and polymorphic VT (Fig. 1). The patient had over 30 ICD shocks in the ensuing 2 months, with resultant negative psychological effects. He was taken to the electrophysiology laboratory for an ablation procedure. Written, informed consent was obtained for publication of this case report and accompanying images.

## Ablation Procedure

Under general anesthesia, a quadripolar catheter was advanced to the right ventricular cavity; a 9-Fr, 9-MHz intracardiac echocardiography (Ultra ICE™, Boston Scientific Corp., Natick, MA, USA) catheter was advanced and positioned in the right atrium. Transseptal access was gained using a 71-cm Brockenbrough needle and an 8.5-Fr Agilis™ NxT deflectable sheath (St. Jude Medical, Inc., St. Paul, MN, USA). Full anticoagulation with intravenous heparin was started, maintaining

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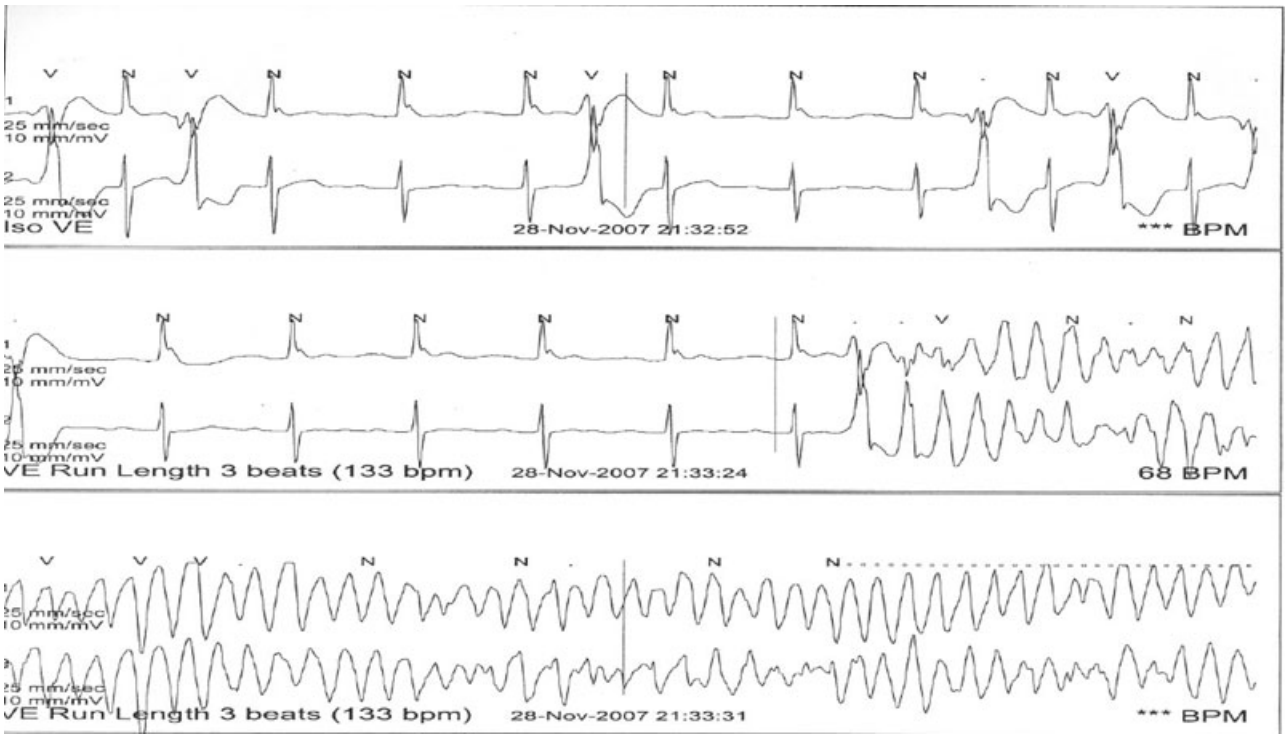


**Figure 1.** *Continuous 2-channel Holter monitor strips showing sinus rhythm and dimorphic PVCs, with the predominant PVC initiating a run of polymorphic ventricular tachycardia.*

an activated clotting time of 300–350 seconds. The transseptal sheath was positioned across the mitral valve and a 38-mm 64-electrode Constellation® basket catheter (Boston Scientific Corp.) was advanced through the sheath under fluoroscopic guidance into the LV. A 3.5-mm tip, B-curve, saline-irrigated Thermocool® (Biosense Webster, Inc., Diamond Bar, CA, USA) ablation catheter was advanced via the retrograde aortic approach into the LV. The small-sized catheters and curves were selected based on the patient's size (50 kg, 1.35 $m^2$ body surface area). Using the EnSite NavX™ system (St. Jude Medical, Inc.) a three-dimensional (3D) electroanatomic map of the LV was created using the eight splines of the basket catheter. Four distinct PVC morphologies were identified, all originating from the septal aspect of the LV. The localization of each PVC was facilitated by the simultaneous recording from all eight splines of the basket catheter. PVC-1 had a right bundle, right inferior axis morphology, and originated from the mid-anteroseptal LV. PVC-2 had a right bundle, right axis morphology, and originated from a slightly more inferior location to PVC-1 (Fig. 2A). PVC-3 had a right bundle, right inferior axis, and originated from a location more apical to PVC-1 and was preceded by a Purkinje potential (PP) 35 ms pre-QRS (Fig. 2C). PVC-4 had a right bundle, superior axis, and originated from a low mid-septal and more apical location, preceded by a PP 29 ms pre-QRS (Fig. 3). There were two distinct populations of PVC: PVCs 1 and 2 were relatively short coupled (mean coupling interval 267.5 ± 9.6 ms), not preceded by PP and had a longer QRS duration (mean 161.1 ± 7.0 ms), whereas PVCs 3 and 4 were relatively late coupled (mean coupling interval 421.8 ± 24.7 ms), with a preceding PP and had a shorter QRS duration (mean 117.9 ± 9.4 ms). These differences were highly statistically significant; $P < 0.0001$ for both comparisons. The earliest PP site for PVC-3 (H 3,4) and subsequent activation sequence of the Purkinje system were different to that for PVC-4 (earliest at G 3,4). These differing activation sequences strongly suggest two distinct Purkinje fiber foci. Right ventricular activation was consistently later than the LV. An activation map was created for each PVC. The sites of earliest activation (and ≥11/12 pace-map match when used) were targeted for ablation (45 W, 60–120 seconds). Four applications of radiofrequency energy were delivered, for a total of 425 seconds. At the conclusion of the procedure, no further spontaneous or inducible PVCs were noted and both QRS duration and morphology and His-ventricular interval (39 ms pre/postablation) remained unchanged. Fluoroscopy time was 44.17 minutes. No complications occurred.

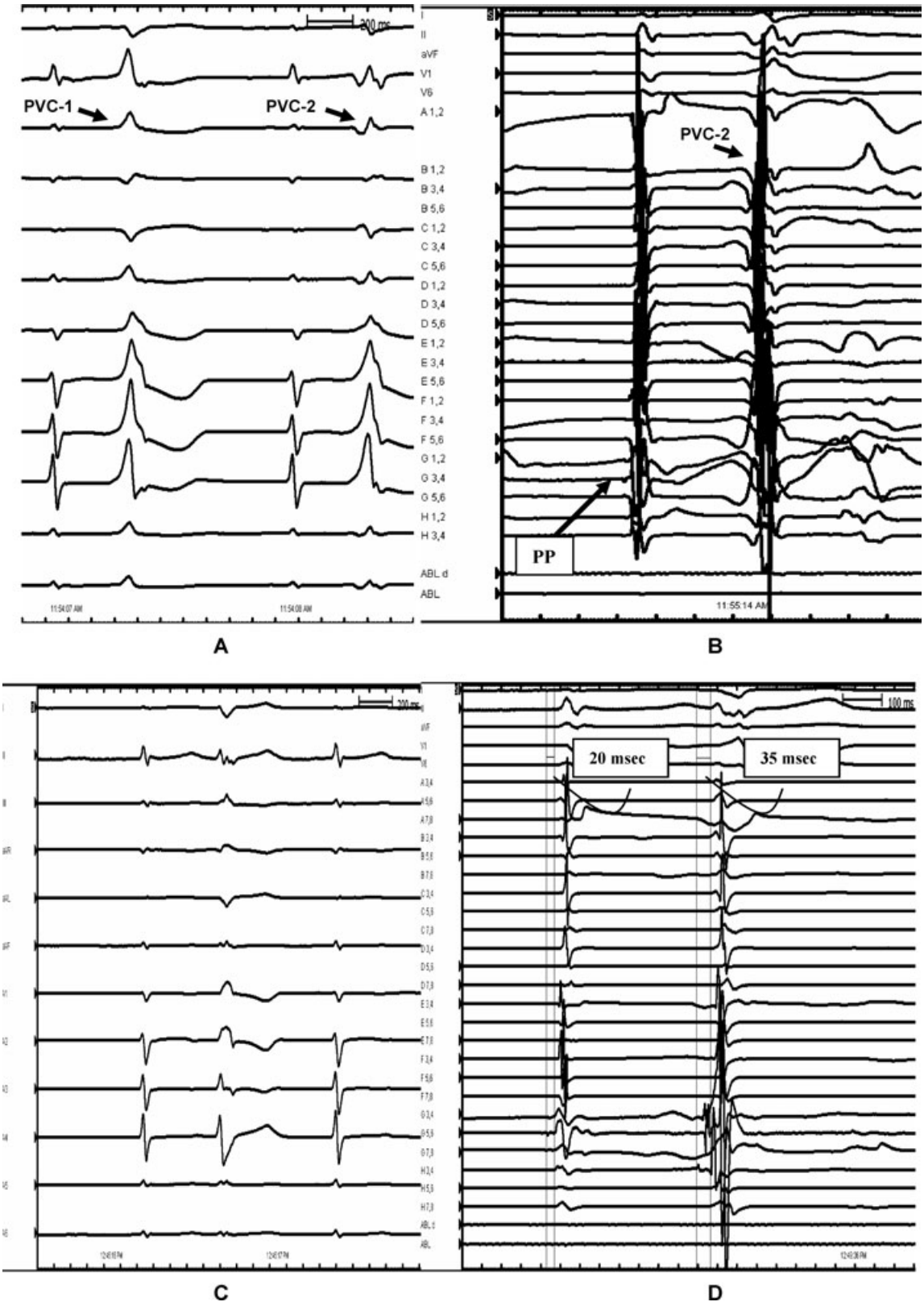


**Figure 2.** *(A) Surface ECG tracing showing polymorphic premature beats in a bigeminy fashion, labeled PVC-1 and PVC-2. From top to bottom, channels are leads I, II, III, aVR, aVL, aVF, V1–V6. (B) Surface ECG and intracardiac*

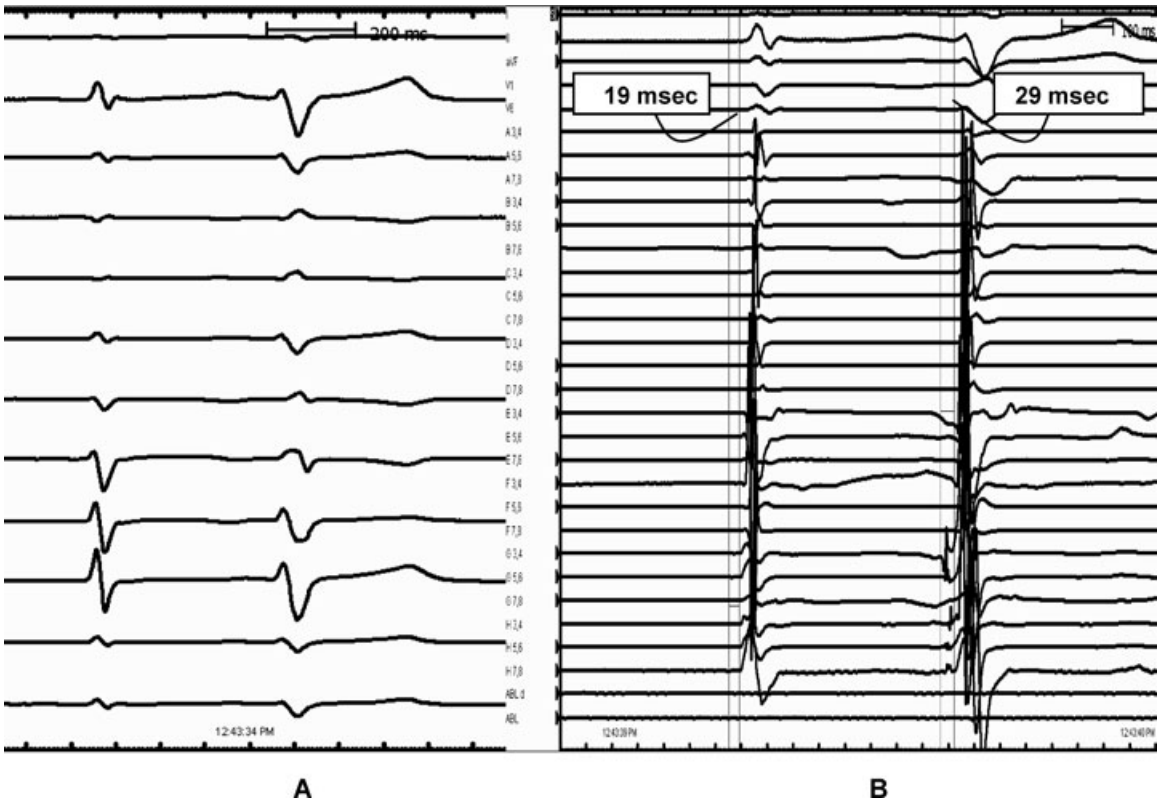


**Figure 3.** *Channels are arranged as in Figure 2. (A) Surface electrocardiographic tracing showing PVC-4. (B) Surface electrocardiographic and intracardiac tracings from the basket catheter showing a sinus beat followed by PVC-4 with a long coupling interval (400 ms, calipers not shown). A Purkinje potential precedes both beats, but it precedes the premature beat by a greater duration (29 ms) than the sinus beat (19 ms) and has a different activation pattern (earliest on G 3,4) to the sinus beat and PVC-3. ABL = ablation catheter channel.*

### Postablation Follow-Up

Over the ensuing 3 months, there was an 80% reduction in ICD shocks compared to preablation. Quinidine gluconate was started at a dose of 300 mg bid. At follow-up of 21 months, he has no VT/VF and no ICD shocks. He has returned to school.

### Discussion

This is the first report of ablation of idiopathic VF in a child. Although all four PVC morphologies originated from the left ventricular septal surface, it is likely that there were two different origins; the short-coupled PVCs (1 and 2) were wider and not preceded by PPs and likely originated in the ventricular muscle, whereas the late-coupled and narrower PVCs (3 and 4) were preceded by PPs and likely arose from the distal Purkinje network. These findings differ from those reported by Haissaguerre et al.[5] Although the hallmark of a PVC of muscular origin was a longer QRS duration in both reports, we report a shorter coupling interval for such PVCs compared to PVCs originating in the Purkinje system, whereas they reported a longer coupling interval. This may be related to the site of origin of the PVC, right ventricular outflow tract in their report and left ventricular septum in ours, or to a difference in mechanism. In

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*tracings from the basket catheter (labeled A–H) showing a sinus beat followed by PVC-2. Note the Purkinje potential (18 ms pre-QRS) during the sinus beat (left) and its absence on the premature beat, suggesting an origin from ventricular muscle. The same was true for PVC-1. (C) Surface ECG tracing showing PVC-3. (D) Surface ECG and intracardiac tracings from the basket catheter showing a sinus beat followed by a PVC-3. A Purkinje potential precedes both beats, but it precedes the premature beat by a greater duration (35 ms) than the sinus beat (20 ms) and has a different activation pattern (earliest on H 3,4) to the sinus beat. ABL = ablation catheter channel; PP = Purkinje potential.*

the same report, left ventricular Purkinje sources displayed markedly different morphologies compared with right ventricular Purkinje sources but the issue of multiplicity of Purkinje foci versus different activation routes from the same focus could not be resolved due to paucity of endocardial mapping coverage. In our report, however, we were able to confirm the presence of more than one Purkinje source due to the extensive LV mapping data simultaneously obtained for each PVC. Interestingly, while both short- and late-coupled PVCs have been described to initiate VF[5] in different patients, we describe both forms operative in a single patient. The patient had more than one morphology of PVC, up to four clinically. Using a point-by-point mapping strategy would have been extremely tedious and made it nearly impossible to map all four different PVC morphologies. The use of a 3D mapping system allowed us to quickly spot the site of earliest activation on the 64-pole catheter to quickly target that area for closer mapping and subsequent ablation. The use of the saline-irrigated catheter allowed accurate mapping with the small 3.5-mm tip while allowing the delivery of deep lesions as needed. In cases with a single PVC morphology occurring relatively frequently, one may forgo the use of 3D mapping and the use of a multipolar mapping catheter.

Class Ia antiarrhythmic drugs have been used with success in this condition. Quinidine sulphate was initially reported by Dock[2] to control idiopathic VF and, subsequently, Belhassen et al.[6] used quinidine in four of a total of five patients with idiopathic VF to suppress VF both acutely and long term with success. This may be related to unique antiarrhythmic properties of quinidine as well as to its vagolytic effects. While our findings agree with, and extend the findings of, these prior reports on the utility of quinidine, we cannot arrive at solid conclusions based on a single case. It is also possible that lesion maturation would have led to the same reduction of PVC, and thus VF, frequency. But this is less likely given an arrhythmia-free 3 weeks postablation period, which was followed by a resurgence of arrhythmia, albeit to a lesser extent, and the subsequent complete absence of all events upon quinidine initiation.

This case demonstrates the feasibility of ablation of idiopathic VF in a child with multiple PVC origins and highlights the value of extensive endocardial mapping data simultaneously acquired from multiple sites.

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