Bundle branch re-entry ventricular tachycardia in a patient with myotonic dystrophy

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Abstract
Bundle branch re-entry (BBR)-ventricular tachycardia (VT) is relatively rare with an incidence of about 6% in sustained monomorphic VT series. However, physicians unexpectedly encounter it in clinical practice. BBR-VT is associated with serious hemodynamic decompensation, and the clinical presentation in approximately 75% of patients with inducible BBR-VT is syncope or cardiac sudden death. Thus, precise mapping of His—Purkinje and bundle branch potentials is necessary for an accurate diagnosis and treatment of re-entrant mechanisms especially for BBR-VT. However, simultaneous recording of both the left-bundle (LB) and right-bundle (RB), as well as His-bundle (HB), potentials is often difficult during tachycardia. Here we report the clear documentation of the activation sequence of the His-Purkinje system during BBR-VT, which could be readily and completely treated by radiofrequency catheter ablation in a myotonic dystrophy patient.

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Case presentation
A 43-year-old female was referred to the emergency room of our hospital with a chief complaint of fainting following the sudden onset of severe palpitations. The electrocardiogram (ECG) on admission revealed a wide QRS complex tachycardia with a heart rate of 235 bpm (Fig. 1A), and she was in shock (blood pressure, 64/32 mmHg). The ECG showed left bundle-branch block morphology with a superior axis. Her chest X-ray revealed pulmonary congestion. Although, in order to discriminate the supraventricular tachycardia with aberration and verapamil-sensitive ventricular tachycardia (VT), the doctors in the emergency room performed an
intravenous administration of 20 mg of adenosine and 5 mg of verapamil, neither of them affected the arrhythmia. Therefore, an external cardioversion was performed, and terminated the arrhythmia. The ECG after the cardioversion exhibited tall R waves in the right precordial leads with an R/S ratio greater than 1.0 and negative T waves in leads V1-3 (Fig. 1B). The echocardiogram revealed a normal cardiac function and normal images. Her physical examination revealed grip and percussion myotonia, and she had been developing more and more muscle weakness and a gait disturbance over the previous 10 years. Furthermore, myotonic discharges were observed on the electromyogram. Thus, she was diagnosed with myotonic dystrophy (MD). The patient had a family history of sudden death in her grandparents, and both had also suffered from MD.

Electrophysiological studies (EPS) were performed to elucidate the mechanism of this wide QRS tachycardia. Five multipolar electrode catheters were placed percutaneously in the coronary sinus...
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Figure 2  Fluoroscopic image showing the location of multipolar electrode catheters and a radiofrequency ablation catheter (ABL) placed in the coronary sinus (CS), His-bundle (HB), left bundle (LB), right bundle (RB), and right ventricle (RV).

The intracardiac electrogram revealed a prolonged HV interval (79 ms) (Fig. 1C). With hypotension and notable fainting, the clinical wide QRS tachycardia was reproducibly induced after the introduction of burst pacing from a distal site of the CS under isoproterenol administration (Fig. 1D). The direction of the impulse propagation resulted in an activation of the LB preceding the activation of the HB, which in turn, preceded that of the RB (Figs. 1E and 3). Although the phenomenon of entrainment could not be confirmed because of serious hemodynamic decompensation during wide QRS tachycardia, irregularities in the His—His and RB—RB cycle lengths followed by similar changes in the RV—RV intervals were found (Fig. 3). With a diagnosis of bundle branch re-entry (BBR)-VT,

Figure 3  Intracardiac recording during the wide QRS tachycardia. Tracings from top to bottom are the electrocardiogram lead I, intracardiac electrograms from the His bundle (HB), left bundle (LB), right bundle (RB), left ventricle (LV), and right ventricle (RV). The irregularities in the LB—LB, HB—HB, and RB—RB cycle lengths were followed by similar changes in the RV—RV intervals. The bar indicates 200 ms.
the radiofrequency (RF) catheter was positioned on the ventricular septum where an RB potential was recorded (Fig. 2). After delivering the RF energy to that site during VT, the BBR-VT was readily terminated (Fig. 1F). The wide QRS tachycardia could no longer be induced by programmed stimulation even under isoproterenol administration after the ablation. Moreover, since cardiac computed tomography confirmed a possible fat deposition in the apex of the ventricular wall and septum, which might later become a substrate for microre-entrant VT, an implantable cardioverter-defibrillator (ICD) was implanted. She has remained well without any symptoms as of her last follow up in August 2008.

Discussion

Cardiac involvement is common in many muscular dystrophies including MD. MD is characterized by a variety of ECG abnormalities (Fig. 1B), which appear to result from selective myocardial necrosis. Both disorders of the atrio-ventricular (AV) conduction and VT also often occur in MD patients. Thus, syncope and/or rapidly progressive congestive heart failure leading to cardiac sudden death may develop in these patients, despite extended periods of apparent circulatory stability during which the only detectable abnormalities are noted in the ECGs. In this present case, the EPS revealed a disorder of AV conduction with prolonged HV interval (79 ms), which was a prerequisite for BBR-VT (Fig. 1C). Several mechanisms for the development of VT in MD patients have been identified such as BBR [1], fascicular re-entry [2,3] and microre-entry in the ventricular wall [4]. BBR-VT is relatively rare with an incidence of about 6% in sustained monomorphic VT series [5], however, physicians unexpectedly encounter them in clinical practice. In BBR-VT, an impaired AV conduction is associated with the formation of macrore-entry between the LB and RB [1]. BBR-VT is associated with serious hemodynamic decompensation, and the clinical presentation in approximately 75% of patients with inducible BBR-VT is syncope or cardiac sudden death [6]. Precise mapping of His—Purkinje and bundle branch potentials is necessary for an accurate diagnosis and treatment of re-entrant mechanisms especially for BBR-VT. However, a simultaneous recording of both the LB and RB as well as HB potentials is often difficult during tachycardia. Here we report the clear documentation of the activation sequence of the His—Purkinje system during BBR-VT (Figs. 1E and 3), and could readily and completely treat by radiofrequency catheter ablation (Fig. 1F) in the MD patient. In view of these findings, a precise mapping of His—Purkinje and bundle branch potentials is important for an accurate diagnosis and treatment of re-entrant mechanisms especially for BBR-VT.

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References