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Bundle Branch Reentrant Ventricular Tachycardia Treated with Catheter Ablation in a Patient with Myotonic Dystrophy

Authors' Contribution:
Study Design A
Data Collection B
Statistical Analysis C
Data Interpretation D
Manuscript Preparation E
Literature Search F
Funds Collection G

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Conflict of interest: None declared

Patient: Male, 40-year-old
Final Diagnosis: Bundle branch reentrant ventricular tachycardia
Symptoms: Chest discomfort
Medication: —
Clinical Procedure: Radiofrequency catheter ablation
Specialty: Cardiology • Neurology

Objective: Rare disease

Background: Bundle branch reentrant ventricular tachycardia (BBRVT) is a rarely encountered ventricular tachycardia (VT) and is classically associated with advanced heart diseases. Importantly, the tachycardia is readily curable with catheter ablation. Without suspicion of BBRVT and recording of the His-Purkinje system, it is hard to diagnose accurately. Myotonic dystrophy (MD) is the most common neuromuscular disease in adults and is known to have a risk of development of BBRVT. Here, we report a case of BBRVT in an MD patient with normal cardiac configuration with typical clinical and electrophysiological features.

Case Report: A 40-year-old man presented with chest discomfort and weakness at the Emergency Department with unstable vital conditions. Electrocardiography showed wide QRS tachycardia with right bundle branch block pattern. The patient had been diagnosed with MD (type I) 3 years ago and had typical clinical features of MD. Transthoracic echocardiography showed normal left ventricular systolic function and no significant structural abnormalities. In the electrophysiologic study, VTs with left and right bundle branch block pattern were induced and diagnosed with BBRVT. Considering the risk of sudden death, implantation of an implantable cardioverter-defibrillator (ICD) was performed. One month later, VT had recurred and was successfully treated with ablation of the right bundle branch.

Conclusions: We present a case of 2 different morphologies of BBRVT in a patient with MD and normal ventricular function. Catheter ablation is a curative method for BBRVT and can be a tool for reducing ICD shock.

Keywords: Bundle-Branch Block • Cardiac Electrophysiology • Electrocardiography • Myotonic Dystrophy • Neuromuscular Diseases • Tachycardia, Ventricular

Full-text PDF: https://www.amjcaserep.com/abstract/index/idArt/929353

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Background

Sustained ventricular tachycardia (VT) can result in hemodynamic compromise and fatal consequences, such as syncope or even sudden cardiac death [1,2]. As myocardial scars play an important role for sustaining of VT, VT usually develops in structural heart disease [3]. Occasionally, VT can occur in patients without significant structural abnormality, with favorable prognosis.

Bundle branch reentrant VT (BBRVT) is a rarely encountered VT [4], and is often associated with advanced structural heart diseases including dilated cardiomyopathy, coronary artery disease, valvular heart disease, and congenital heart disease,

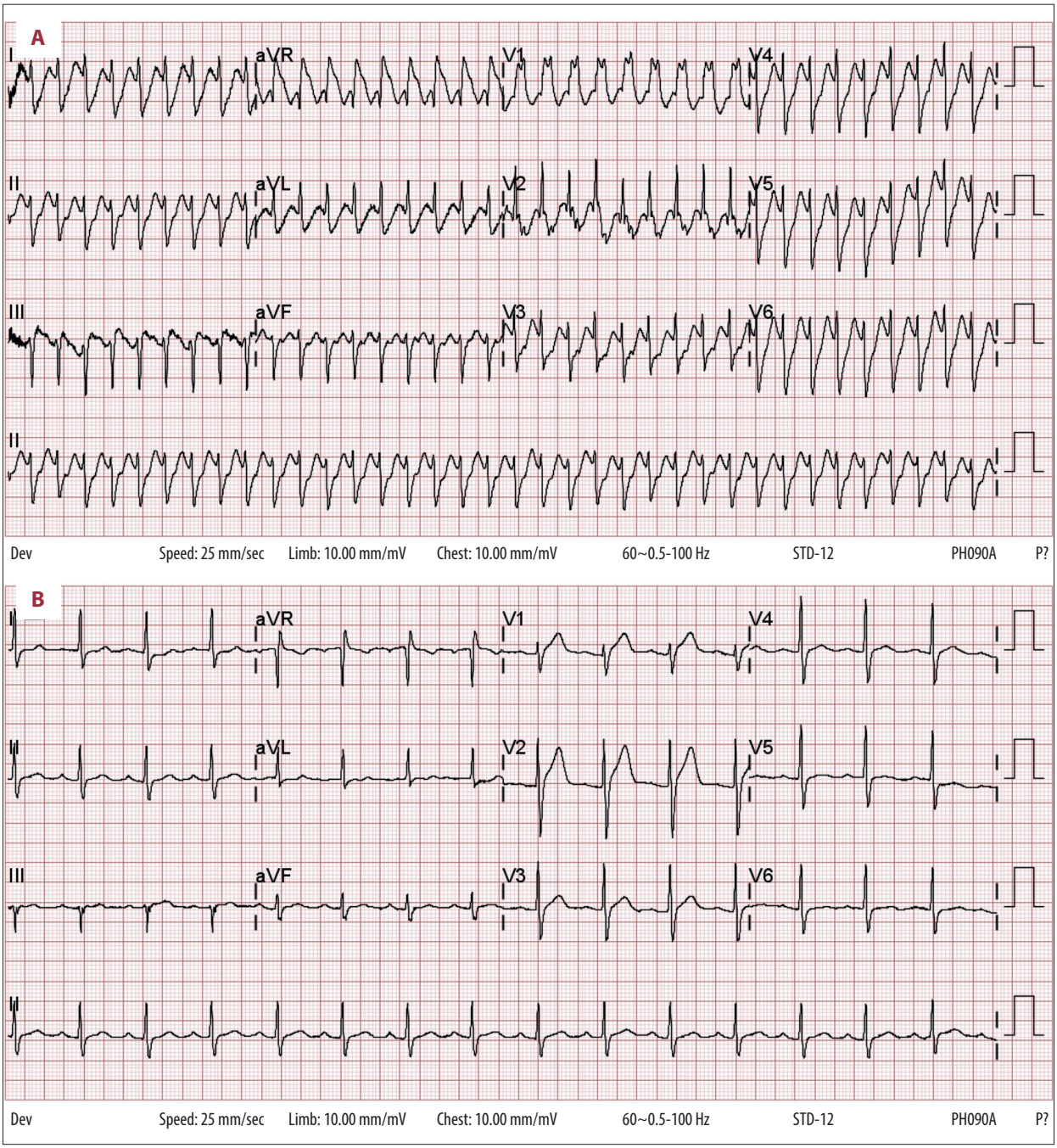

Figure 1. The electrocardiogram (ECG) during the wide QRS tachycardia (cycle length: 275 msec) with typical right bundle branch block pattern on admission (**A**). ECG during sinus rhythm showed delayed AV conduction (PR interval: 220 msec) and subtle intraventricular conduction delay (QRS duration: 104 msec) (**B**).

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but BBRVT can also occur in patients with normal ventricular size and function. Once diagnosed, BBRVT can be treated effectively by catheter ablation.

Myotonic dystrophy (MD) is a disease that involves the whole body, with characteristic clinical features suggesting neuromuscular disease [5]. Although it rarely progresses to a dilated cardiomyopathy in patients with MD, VT can occur as a consequence of reentry in the diseased distal conduction system, as characterized by bundle branch reentry and interfascicular reentry tachycardia [5]. BBRVT should be suspected when patients with MD present with wide QRS tachycardia, even without structural abnormality. Here, we report a case of BBRVT in a patient with MD and normal cardiac configurations.

Case Report

A 40-year-old man presented at the Emergency Department with chest discomfort and weakness. The initial survey revealed he had low blood pressure (70/59 mmHg) and high heart rate (218/min). Electrocardiography (ECG) showed wide QRS tachycardia with right bundle branch block (RBBB) pattern (**Figure 1A**).

Because the patient was hemodynamically unstable with tachycardia, DC cardioversion was performed. ECG during sinus rhythm showed first-degree AV delay and subtle intraventricular conduction delay with 104 msec of QRS duration (**Figure 1B**).

The patient had been diagnosed with MD (type I) 3 years ago. Physical examination showed baldness, thin face, and distal muscle atrophy suggesting MD (**Figure 2**). An elderly brother of the patient had similar physical finding but had not been diagnosed with MD. The heart examination was unremarkable. Although acute myocardial injury was evident with typical rise and fall pattern of cardiac enzymes, the epicardial coronary artery was normal in coronary angiography. Transthoracic echocardiography showed normal left ventricular systolic function and no significant structural abnormalities.

Electrophysiologic Study

To confirm the mechanism of the tachycardia, an electrophysiologic study (EPS) was performed with multipolar electrode catheters positioned at the high right atrium, coronary sinus, His bundle region, and right ventricular (RV) apex. The basic EP interval was remarkable for a prolonged HV interval (73 msec), but PA and AH interval were normal. No supraventricular tachycardia was induced with programmed electrical stimulation (PES). Wide QRS tachycardia with left bundle branch block (LBBB) pattern was induced by triple ventricular extrastimulation test (400/260/200) at a driving cycle length of 500 msec. Tachycardia cycle length

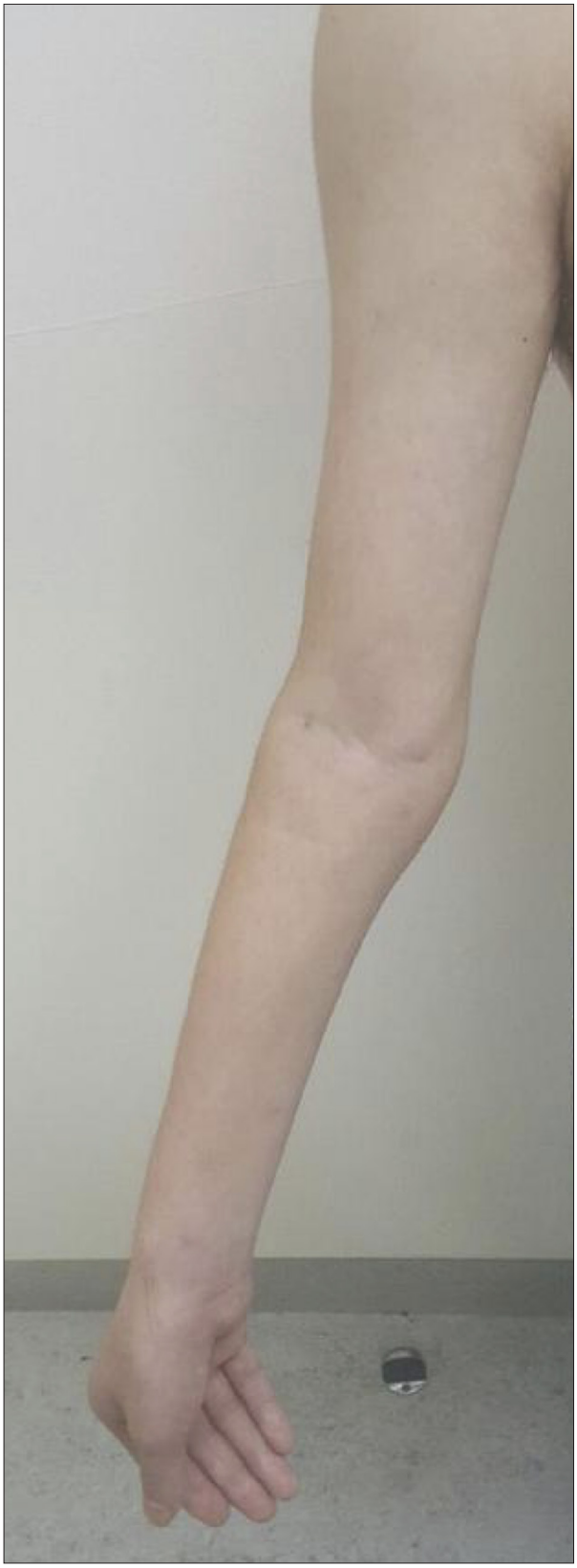

Figure 2. Clinical features of the patient. Distal muscle atrophy was noted in the upper extremity.

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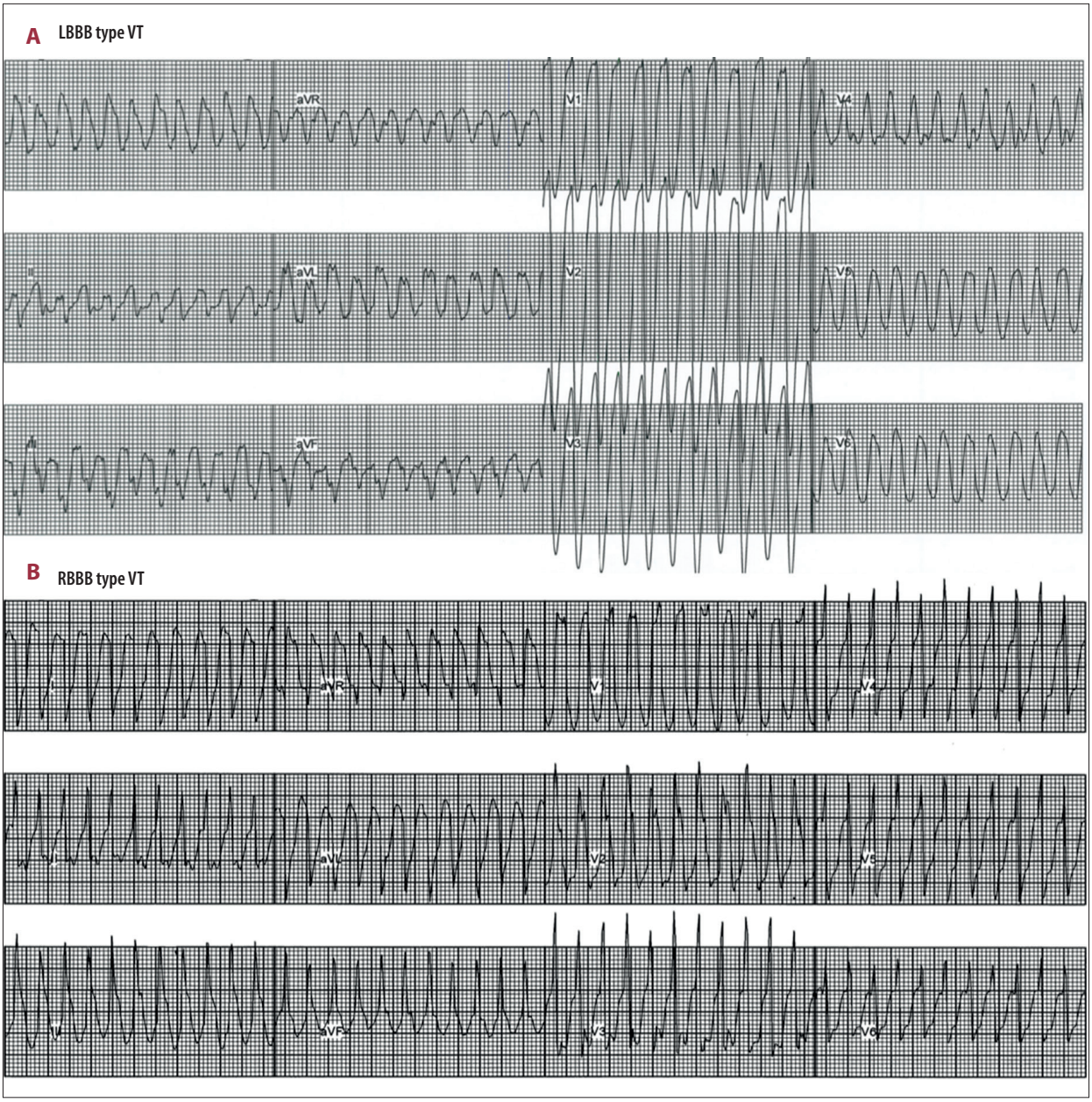

Figure 3. Induced tachycardia during initial (**A**) electrophysiologic study (EPS) and repeated EPS (**B**). Wide QRS tachycardia with left bundle branch block (BBB) pattern was induced with ventricular electrical stimulation. Tachycardia cycle length (TCL) was 216 msec (**A**). The other form of ventricular tachycardia with right BBB morphology with TCL of 228 msec (**B**).

(TCL) of the induced tachycardia was 216 msec and blood pressure dropped to 70/40 mmHg (**Figure 3A**). H-V interval during tachycardia was 112 msec, which was longer than H-V intervals during sinus rhythm. There was a 1: 1 H-V relationship and V-V intervals were dependent on H-H intervals (**Figure 4A**). Entrainment pacing at the RV showed short post-pacing interval – TCL, which was less than 30 msec (**Figure 4B**). These findings suggested BBRVT. We finished the EPS and implanted an implantable cardioverter-defibrillator (ICD). One month later, the patient presented with an appropriate ICD shock. As the patient had not been taking any anti-arrhythmic agents, immediate catheter ablation was planned, and repeated EPS was done with recording right bundle branch potentials. Activation sequences during VT with LBBB pattern were His bundle, right bundle branch, and RV (**Figure 5A**). The other VT with RBBB pattern, similar to the clinical tachycardia, was induced with PES (**Figure 3B**). Unfortunately, poor His bundle signals and easy termination of the VT hindered determining the exact mechanism of the tachycardia with RBBB pattern (**Figure 5B**). Radiofrequency catheter ablation was performed on the ventricular septum, where the right bundle branch

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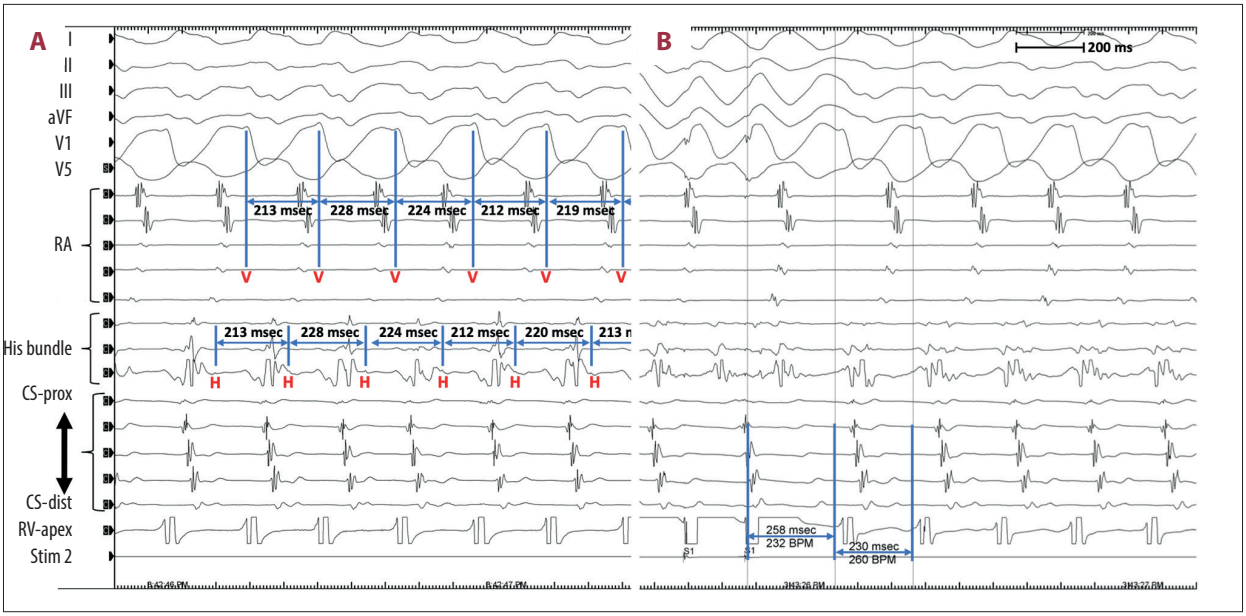

Figure 4. The intracardiac recording showing the activation sequence in the right atrium (RA), coronary sinus (CS-prox~CS-dist), His bundle, and right ventricle (RV apex). During wide QRS tachycardia with left bundle branch block pattern, the H-V relationship was 1: 1 with constant H-V intervals and V-V intervals were dependent on H-H intervals (**A**). Post-pacing interval – tachycardia cycle length was 28 msec at the RV (**B**).

potential was recorded (**Figure 6A, 6B**), and post-ablation ECG showed complete RBBB (**Figure 6C**). After successful ablation, no VT was recorded on ICD and the patient was doing well at a recent follow-up, without anti-arrhythmic agents for VT.

Discussion

MD is an autosomal dominant disorder and is the most common neuromuscular disease in adults. The characteristic features of MD are myotonia, which is a delayed muscle relaxation after contraction, weakness and atrophy of skeletal muscles, and systemic manifestations. BBRVT can occur as a cardiac manifestation of MD [5].

A delay in conduction through the His-Purkinje system is the mechanism of the arrhythmia [6]. The involvement of this system is manifested by intraventricular conduction delay or BBB in the ECG and by prolongation of the HV interval in the EPS. Precise mappings of His-Purkinje and bundle branch potentials are necessary for accurate diagnosis and treatment of BBRVT. In this patient, ECG showed subtle intraventricular conduction delay and prolongation of the HV interval was identified in the EPS, which indicated disease in the His-Purkinje system.

During EPS, VT with LBBB morphology was induced with PES and showed a 1: 1 relationship between His EGM and ventricular EGM. Spontaneous changes of V-V intervals were preceded by changes of H-H intervals. Because the postulated antegrade limb of the reentry circuit of LBBB type BBRVT is the His bundle to the right bundle branch and then to the RV, His bundle and right bundle branch potentials should precede the onset of ventricular activation, as demonstrated in this case (**Figure 5A**). RV entrainment pacing in the RV apex revealed short post-pacing interval – TCL, which indicated the RV-involved reentry circuit [7]. Unfortunately, atrial pacing maneuvers were not attempted.

Although VT with RBBB pattern, similar to clinical tachycardia, was induced with PES, we could not clearly demonstrate the mechanisms of the tachycardia due to the poor His bundle signals and easy termination of the VT. However, the findings suggesting BBRVT were typical RBBB pattern and apex to basal RV activation during tachycardia, longer HV interval than sinus rhythm, diagnosis of LBBB type BBRVT, and non-inducibility of the tachycardia after right bundle branch ablation.

In previous reports, the response to pharmacological treatment of BBRVT is poor, but ablation of one of the branches of the His system, usually the right branch, has been shown to be an effective treatment in these patients [6,7]. BBRVT is an easily ablatable cause of VT compared to complex substrate ablation for other VT. Hence, a diagnosis of BBRVT should not be overlooked.

Another importance of BBRVT is the high risk of sudden cardiac death in these patients [8]. Despite successful ablation, up to 6.5% of these patients died suddenly from arrhythmias [9]. In this particular case, ICD implantation is required due to future

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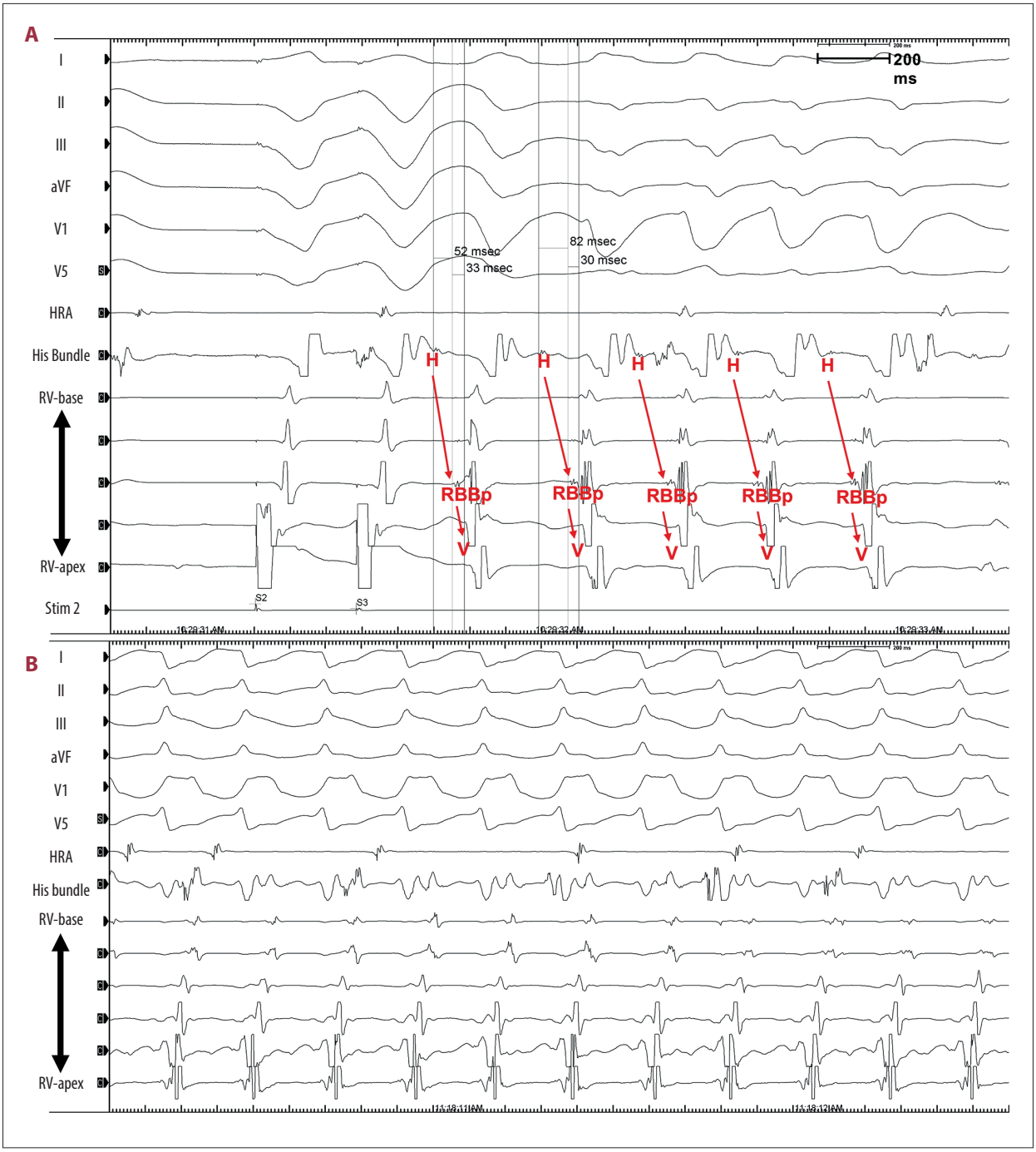

Figure 5. The intracardiac recording showing the activation sequences during ventricular tachycardia (VT) with left bundle branch block pattern. RV activation was preceded by His bundle and right bundle branch (**A**). Intracardiac tracing during VT with right bundle branch block (**B**). HRA – high right atrium; RBBp – right bundle branch potential; RV – right ventricle; V – ventricular activation; H – his bundle activation.

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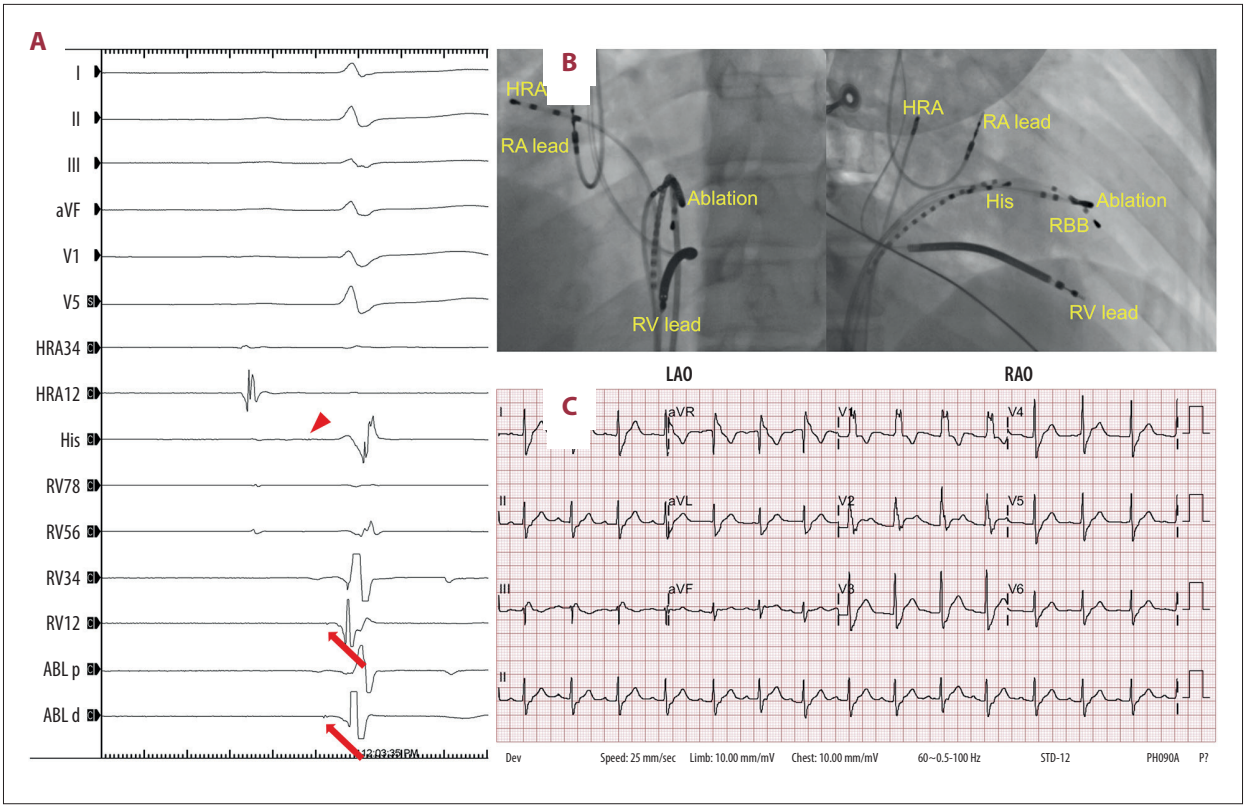

Figure 6. Right bundle branch potentials (arrows) were recorded in the intracardiac tracing of the ablation catheter (ABLp and ABLd). Tiny His bundle potential was recorded in the His catheter (arrowhead) (**A**). Fluoroscopic image showing the location of multipolar electrode catheters and ablation catheter (**B**). Post-ablation ECG showed complete right bundle branch block (**C**). HRA – high right atrium; RBB – right bundle branch; RV – right ventricle.

risk of sudden cardiac death, especially since the mechanism of the initial VT was unclear.

Conclusions

Catheter ablation can easily cure BBRVT, but there is also the potential risk of under-treatment, as BBRVT is mistakenly considered as VT that requires a complex ablation procedure. Hence, bundle branch reentry should be included in the differential diagnosis of patients presenting with sustained VT. This case report underscores that BBRVT can occur not only in patients with advanced structural heart disease, but also in those with normal heart configuration. In order to obtain an accurate diagnosis of BBRVT, the His and bundle branch potentials must be carefully recorded. With the high success rate of catheter ablation, BBRVT can be cured, thus avoiding unnecessary ICD shocks.

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