

CASE REPORT

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Bundle branch reentrant ventricular tachycardia in a patient with Fabry disease

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Abstract

Background Bundle branch reentrant ventricular tachycardia is a unique type of VT that employs both bundles and the ventricular septum as integral components of a re-entrant circuit. It is usually observed in dilated cardiomyopathy, and its circuit depends exclusively on the specialized conduction system.

Case presentation A 67-year-old man with a history of Fabry disease, and atrial fibrillation ablation, was admitted for a wide QRS tachycardia at 150 bpm, with an LBBB. Pharmacological therapy failed to stop the arrhythmia. Intra-cardiac recordings confirm the diagnosis. His Bundle recordings are essential, and Atrioventricular (AV) dissociation is typically present. The method used to induce left BBRVT is right ventricle apex stimulation. An electrophysiological study showed an AV dissociation a long HV sequence and a similar QRS morphology between VT and sinus rhythm. An overdrive stopped it. The same arrhythmia reappears and is restored to sinus rhythm by electrical cardioversion. We decided to implant an Implantable Cardiac defibrillator (ICD) with a backup pacing for secondary prevention and to ablate the right bundle branch.

Conclusion BBRVT is a rare entity that is underreported. Pharmacological therapy is usually inefficient. An ICD is recommended for secondary prevention and can provide backup pacing, frequently required after catheter ablation to prevent the development of AV block or an excessive prolonged HV interval. Catheter ablation of the bundle branch is the first-line therapy.

Keywords Bundle branch reentrant ventricular tachycardia, Ventricular tachycardia, Fabry disease, Electrophysiology

Background

Bundle branch reentrant ventricular tachycardia (BBRVT) is a unique type of ventricular tachycardia (VT) that employs both bundles and the ventricular septum as integral components of a re-entrant circuit. It is a life-threatening arrhythmia and accounts for 6–8% of all inducible sustained VTs [1].

Its incidence is still underreported in the literature, and its recognition is important because of the inefficiency of pharmacological therapy.

Case report

We report the case of a 67 years man with a history of Fabry disease with renal impairment under Migalastat, and atrial fibrillation ablation, without any coronary disease, admitted to our department for a wide QRS tachycardia at 150 bpm, with left bundle branch block (LBB) well tolerated (Fig. 1).

Transthoracic echocardiography showed hypertrophic left and right ventricles, a preserved left ventricular function, and an altered right ventricle function.

At the intensive care unit, pharmacological cardioversion and adenosine failed to stop and slow the

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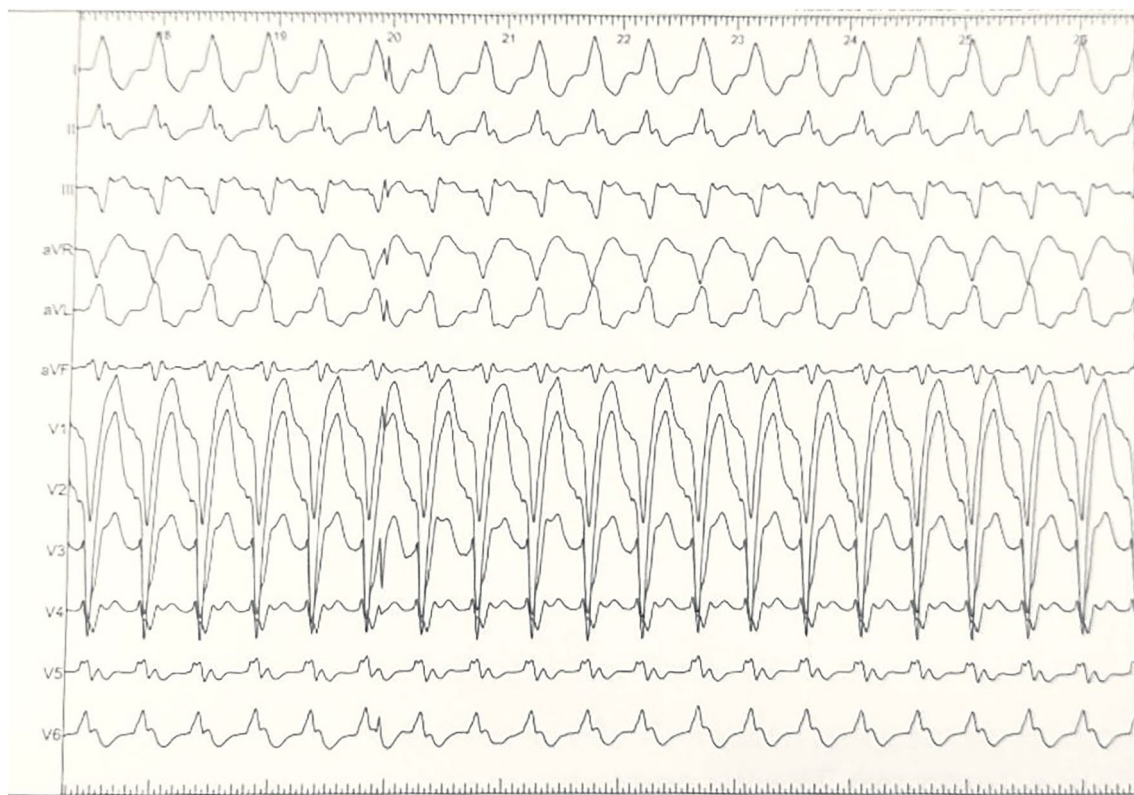


Fig. 1 EKG at admission showing wide QRS tachycardia

arrhythmia, atrioventricular reentrant tachycardia was excluded. An urgent electrophysiological study (EPS) showed an atrioventricular (VA) dissociation and a His-ventricular (HV) sequence at 50 ms, excluding supra-ventricular tachycardia (Figs. 2, 3, 4).

In addition to the similar morphology between VT and sinus rhythm, a left BBRVT (type A) diagnosis was done. An overdrive stopped the arrhythmia (Fig. 5).

A few hours later, the same arrhythmia reappeared, not well tolerated, with loss of consciousness. External electrical cardioversion restored it to a sinus rhythm.

We decided to implant a cardiac defibrillator for

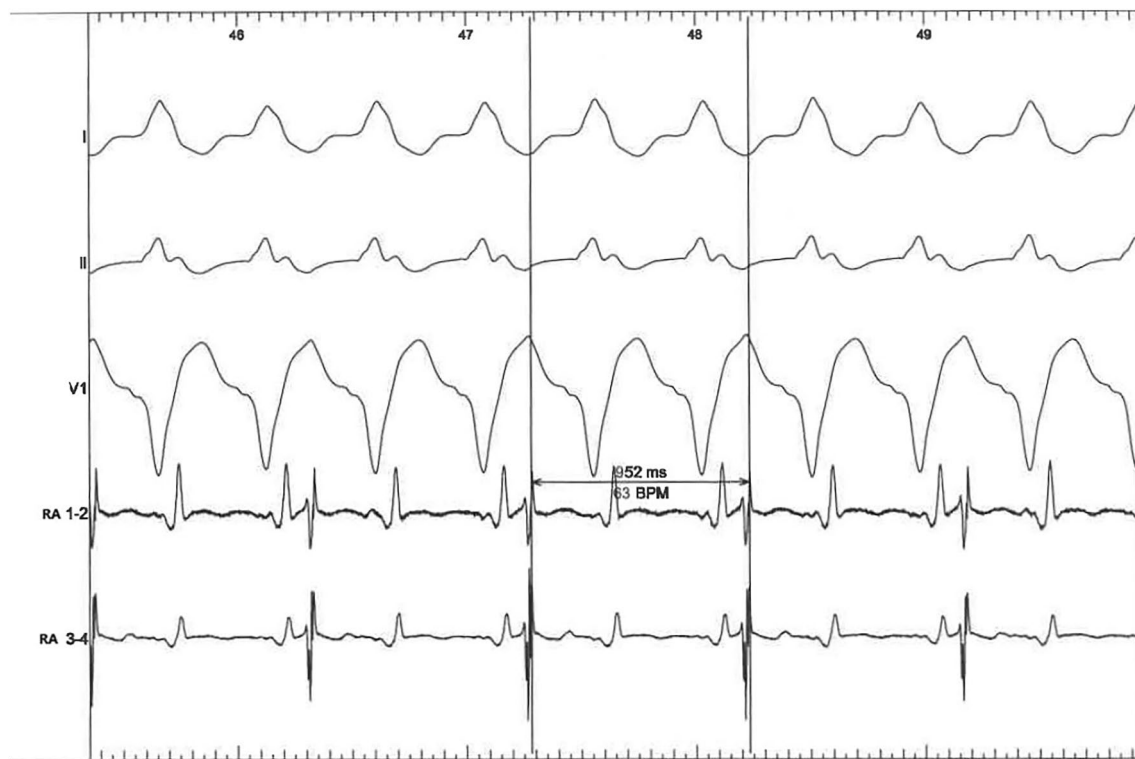


Fig. 2 EPS showing an AV dissociation

the right ventricle apex. In some patients, it can be inducible with atrial pacing, in others; procainamide and isoproterenol can favor its induction.

In the majority of patients with BBRVT, the HV interval is prolonged in sinus rhythm [6], and may be in normal limits, however in these patients functional His-Purkinje system impairment is manifesting as H-V interval prolongation or split His-bundle potentials and becomes commonly evident during atrial programmed stimulation or burst pacing.

The use of pharmacological therapy in BBRVT is usually inefficient. If sustained BBR VT is inducible during

programmed stimulation, catheter ablation of the bundle branch is the first-line therapy [7] but can result in an excessive prolonged HV interval or complete atrio-ventricular block in 30% of cases. An implantable cardioverter defibrillator is recommended for secondary prevention and can provide backup pacing [1].

Conclusion

BBRVT is a rare entity that is underreported.

The recognition of this tachycardia is important because of the inefficiency of pharmacological therapy.

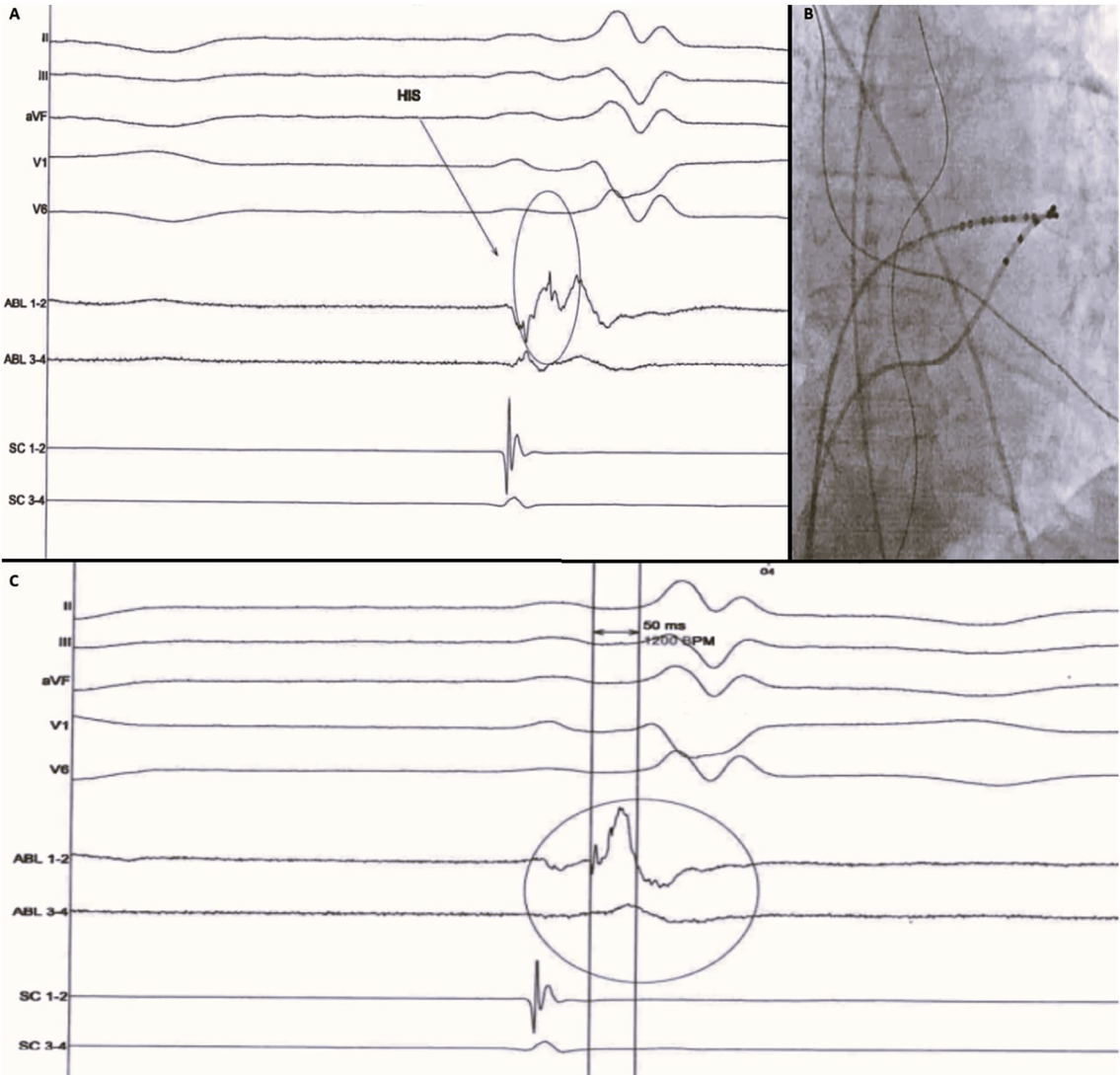


Fig. 3 EPS showing the HV sequence. **A** The His in sinus rhythm. **B** Fluoroscopic image of multipolar catheter position. **C** HV sequence measurement



Fig. 4 EPS showing HV sequence in BBRVT. **A** HV sequence measurement. **B** The His in BBRVT

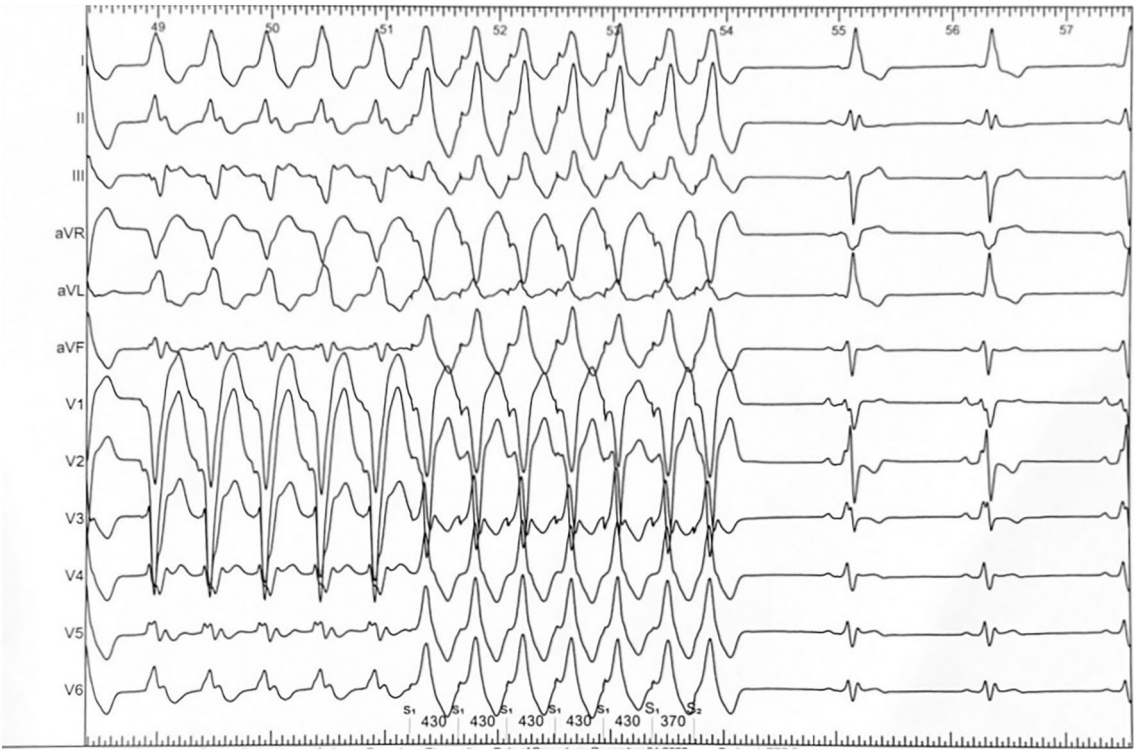


Fig. 5 An overdrive at the EPS that stopped the arrhythmia



Fig. 6 EKG after right bundle branch ablation

Abbreviations

AV	Atrio-ventricular
LBB	Left bundle branch block
BBRV	Bundle branch reentrant ventricular tachycardia
VT	Ventricular tachycardia
EPS	Electrophysiological study
EKG	Electrocardiogram
HV	His-ventricular
ICD	Implantable Cardiac defibrillator
RB	Right bundle
VES	Ventricular stimulation

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Declarations**Ethics approval and consent to participate**

Not applicable.

Consent for publication

The patient's consent for publication was obtained.

Competing interests

The authors declare that they have no competing interests.

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