Irregular wide QRS complex tachycardia in a patient with pulmonary hypertension: What is the mechanism?

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Introduction

Pulmonary hypertension (PH) results in right ventricular (RV) hypertrophy and dilation that could lead to the development of arrhythmogenic substrates for cardiac arrhythmias, which are important predictors and contributors to morbidity and mortality in these patients.[1] To date, the mechanism of ventricular arrhythmia in patients with PH are less well understood.

Here, we reported a case presenting sustained and nonsustained ventricular arrhythmia owing to the increased automaticity of the right Purkinje arborization in a patient with severe chronic PH and the promising effectiveness of radiofrequency catheter ablation in elimination of the ventricular arrhythmia.

Case presentation

An 83-year-old woman had history of atrial fibrillation (AF) with controlled ventricular response and chronic PH. She was clinically stable on medical therapy but developed 1 month of dyspnea, dizziness, and near syncope. The echocardiography revealed severely dilated left and right atria, severe PH (RV systolic pressure of 74.9 mm Hg) with moderate tricuspid regurgitation, mild RV hypertrophy, and mild RV systolic dysfunction. Her 24-hour Holter monitor demonstrated incessant wide QRS complex tachycardia with a mean ventricular rate of 124 bpm. Figure 1 demonstrated the 12-lead electrocardiogram (ECG) before and during wide QRS complex tachycardia. Antiarrhythmic drugs with amiodarone and verapamil were used after admission to coronary care unit, with unsatisfactory effects. The recorded ECG monitoring showed frequent nonsustained wide QRS complex tachycardia. What is the mechanism of the wide QRS complex tachycardia?

Electrophysiological study

After providing informed consent, the patient underwent an electrophysiological study in fasting state without sedation. A multielectrode catheter was placed in the RV apex in standard fashion. The initial rhythm presented with AF with both narrow and wide QRS complexes. The wide QRS complex morphology was left bundle branch block, superior axis, V5 transition with QRS duration of 142 msec. His potential was recorded occurring after the wide QRS complex during tachycardia, supporting the diagnosis of ventricular tachycardia/ventricular premature complexes (VT/VPC). Drug test with adenosine did not suppress the VT/VPC. The 3D geometry was created by the CARTO 3.2 UDM system (Biosense-Webster, Baldwin Park, CA) using a 3.5-mm-tip open-irrigated catheter (Thermocool; Biosense-Webster) and voltage mapping depicted no remarkable low voltage zone or abnormal fractionated electrograms (Figure 2A). Activation mapping of the VT/VPC demonstrated the earliest activation site localized at the anterior mid free wall of the right ventricle, and a Purkinje potential preceded the onset of wide QRS tachycardia by 30 msec. Pace mapping at the earliest activation site yielded 12/12 leads matched QRS morphology (Figure 2B). Radiofrequency energy was delivered in a temperature-controlled mode at 30-35 watts targeting for an impedance drop of 10 ohms. The morphology of VT/VPC changed in leads I, II, III, AVL, AVF, V3, and V4 after initial attempts of ablation. Remapping of the VT/VPC demonstrated the earliest activation localized near the previous ablation site at a distance of 11.2 mm. Similarly, a Purkinje potential preceding VT/VPC by 32 msec was noted, while pace mapping also yielded

KEYWORDS Ventricular tachycardia; Ventricular premature complex; Pulmonary hypertension; Ablation; Purkinje potential; Automaticity

ABBREVIATIONS **AF** = atrial fibrillation; **ECG** = electrocardiogram; **MB** = moderator band; **PH** = pulmonary hypertension; **RV** = right ventricular; **VPC** = ventricular premature complex; **VT** = ventricular tachycardia (Heart Rhythm Case Reports 2016;2:63–66)

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KEY TEACHING POINTS

- Currently, human studies on the mechanisms of ventricular arrhythmias in patients with pulmonary hypertension are still lacking.
- The Purkinje system may play a role in the arrhythmogenesis of ventricular tachycardia in patients with pulmonary hypertension.
- The recording of discrete potentials from the Purkinje system provides help in the effectiveness in elimination of right Purkinje system–related ventricular tachycardia by radiofrequency catheter ablation.

12/12 leads matched QRS morphology (Figure 2C). Radiofrequency ablation successfully terminated VT/VPC. After ablation, VPCs and nonsustained VT were not inducible by the infusion of isoprenaline and programmed stimulation. The patient was uneventful during clinical follow-up 3 months later.

Discussion

During wide QRS complex tachycardia, it is important to differentiate between supraventricular and ventricular origin of the arrhythmia in order to guide us in clinical management. The irregular cycle length of wide QRS complex tachycardia might easily be misinterpreted as aberrant conduction during AF. Also, using the Brugada algorithm[2] in this case did not differentiate the wide QRS tachycardia as supraventricular tachycardia with bundle branch block aberrancy from VT. However, the recording of His potential after the wide QRS complex during electrophysiologic study and the earliest activation sites localized at anterior mid RV free wall supported the diagnosis of VT/VPCs.

The activation map of the nonsustained VT/VPCs showed the focal earliest activation from the right Purkinje system. Purkinje potentials were recorded 30 msec and 32 msec earlier than the VT/VPCs from the 2 successful ablation sites (shown in the ablation catheter bipolar electrogram in Figure 2B and C). The Purkinje potentials at the successful ablation sites preceding the ventricular activation were also noted during AF with narrow QRS complexes. The recording of discrete potentials from the Purkinje system provides help in localization and facilitating ablation of these arrhythmias.[3]

In this case, it is also possible that VT/VPCs originated from the moderator band (MB), which usually extends from the septum to the RV free wall and encompasses the RV Purkinje fibers.[4] VPC arising from the MB may variably arise from the septal insertion site, from the body, and from the RV free wall insertion site. The Purkinje system is a complex network of fibers that spreads through the subendocardium of the right and left ventricles and VT/VPCs may arise from these fibers surrounding the MB.

In contrast to idiopathic left fascicular VTs, which mostly could be suppressed by verapamil,[5,6] the VT/VPCs demonstrated poor response to either verapamil or amiodarone, implying different electrophysiologic characteristics in this case. Moreover, a focal activation pattern with radial

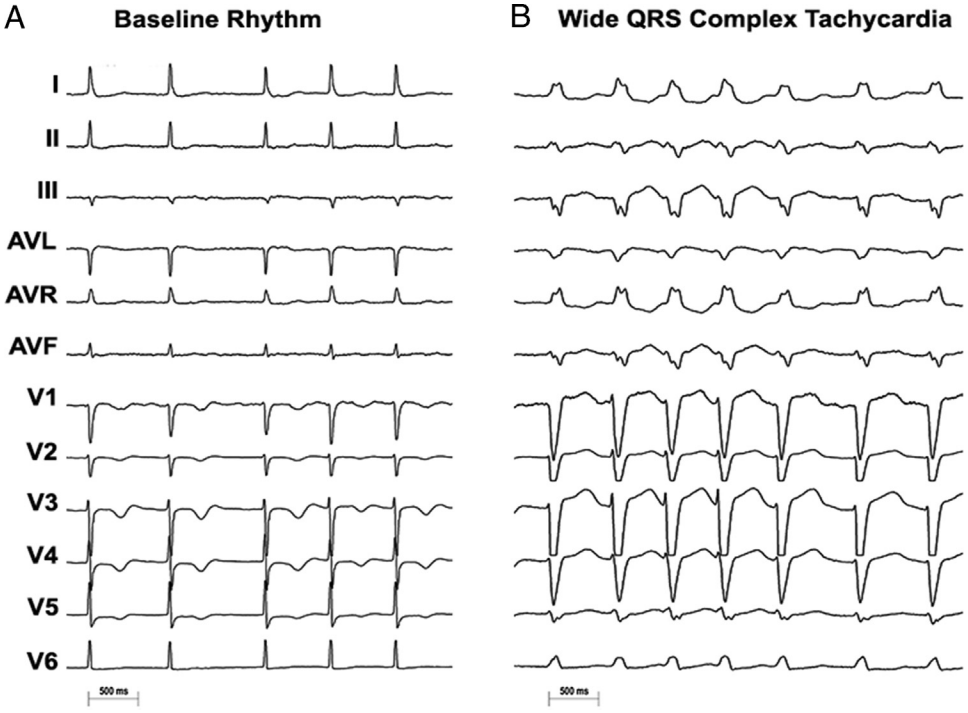

Figure 1 The 12-lead electrocardiogram (ECG) at baseline and during tachycardia. **A:** The baseline ECG showed atrial fibrillation and narrow QRS complex. **B:** ECG during wide QRS complex tachycardia with left bundle branch block morphology, a superior axis, V5 transition, and QRS duration of 142 msec.

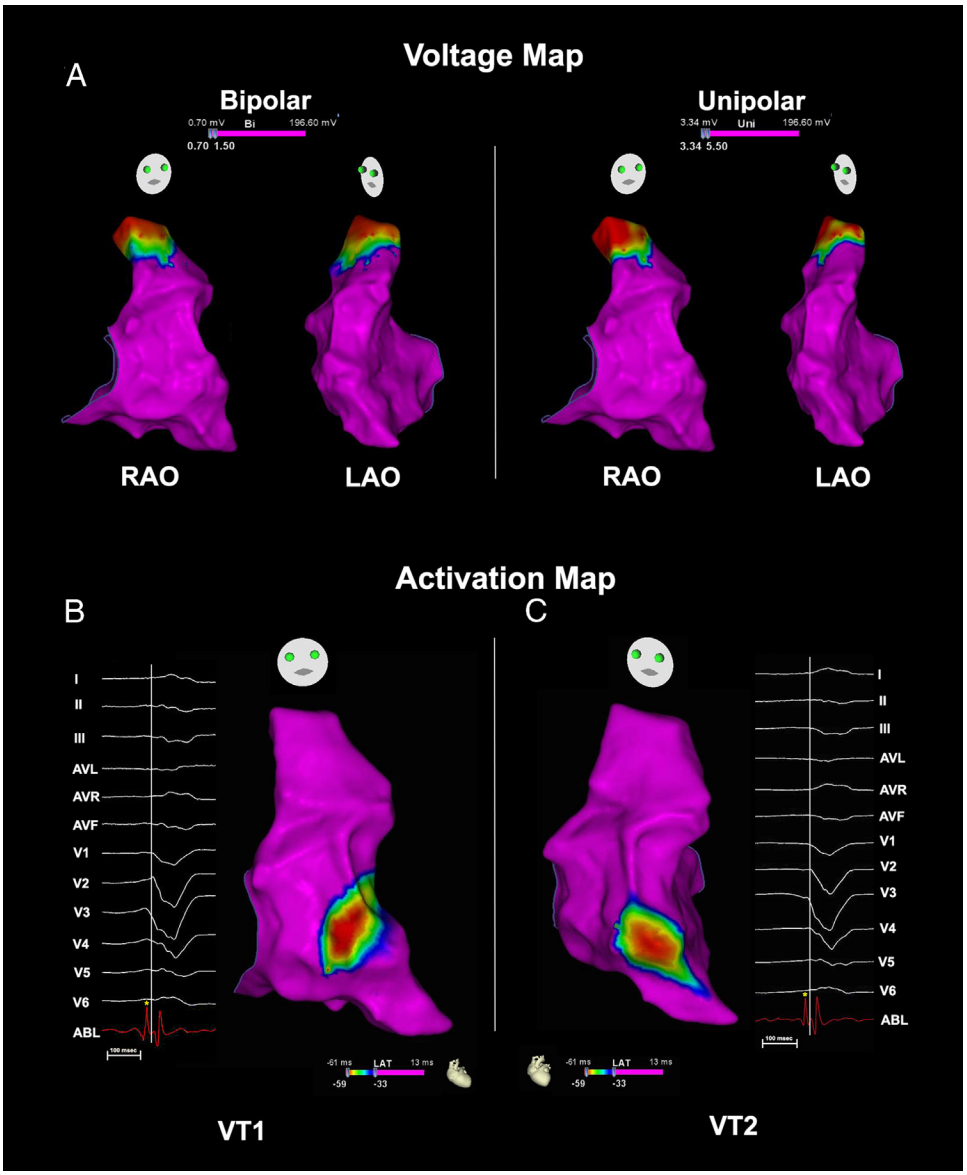

Figure 2 Voltage and substrate mapping. **A:** Voltage mapping depicted no remarkable low voltage zone (defined by voltage <1.5 mV). **B:** The activation map of the ventricular tachycardia (VT) / ventricular premature complex (VPC) demonstrated the earliest activation site localized at the anterior mid free wall of the right ventricle, and a Purkinje potential (*) preceded the onset of wide QRS tachycardia by 30 msec. Pace mapping at the earliest activation site yielded 12/12 leads matched QRS morphology. **C:** Remapping after initial ablation showed another VT/VPC originating from the foci at a distance of 11.2 mm from the previous ablation site with a Purkinje potential (*) preceding VT/VPC by 32 msec. Pace mapping at the foci also yielded 12/12 leads matched QRS morphology comparing with the documented VT/VPC. Radiofrequency ablation successfully terminated the VT/VPC.

spreading of VT/VPCs activation map and irregular VT cycle lengths proved that the mechanism of ventricular arrhythmia was less likely to be re-entrant, while triggered activity was excluded by the fact that VT/VPCs were also unresponsive to adenosine. To the best of our knowledge, sustained VTs owing to automaticity of the right Purkinje system and manifesting as electrical storm in a patient with chronic PH have not been previously reported.

Purkinje fibers have been implicated in both the initiation and the maintenance of ventricular tachyarrhythmias in animal models.[7,8] Haïssaguerre et al[9] also demonstrated the role of the Purkinje system in triggering of idiopathic ventricular fibrillation. Several previous studies[10–12] demonstrated that the spontaneous re-entrant ventricular arrhythmia originating from conduction system might become frequent in patients who have developed conduction abnormalities in the His-Purkinje system. Additionally, early afterdepolarization–mediated triggered activity in failing RV cardiomyocytes as the mechanism for initiation of nonsustained VT has been demonstrated in rats with monocrotaline-induced PH and RV failure.[13]

Previous study demonstrated that supraventricular arrhythmias such as AF and atrial flutter contribute to the worsened outcomes in patients with PH.[1] In addition, prospective and retrospective studies by Bandorski et al[14,15] showed increased incidence of nonsustained VT in patients with chronic PH undergoing electrophysiological study or a 72-h Holter ECG. However, these studies were limited by moderate sample size

and the lack of underlying pathophysiological mechanism of ventricular arrhythmias. To date, studies on recognition of the origin of ventricular arrhythmias in patients with PH are still lacking and are warranted for further prospective investigation.

In conclusion, we reported a case with nonsustained and irregular wide QRS complex tachycardia originating from right Purkinje arborization owing to the increased automaticity in the patient with chronic PH. Mapping the earliest Purkinje potential could provide help and promising effectiveness on elimination of VT/VPCs by radiofrequency catheter ablation.

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