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CASE REPORT
Congenital heart disease

Ablation for idiopathic left ventricular tachycardia in a patient with double outlet right ventricle who underwent Fontan operation: a case report

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Background The incidence of ventricular tachycardia (VT) in patients following Fontan operation is reported as 3.5%. Furthermore, in patients with repaired double outlet right ventricle (DORV), scar-related VT and outflow tract VT have been reported; however, Purkinje-related VT has not previously been reported. In this report, we present the case of idiopathic left VT (ILVT) in a patient with DORV who underwent Fontan operation.

Case summary A 31-year-old man was diagnosed as having DORV with complete atrioventricular defect at birth. When he was 17 years old, he underwent surgical repair, including extracardiac Fontan operation and common atrioventricular valve replacement. Five years later, VT was detected. Since some medications were ineffective in suppressing VT, he was referred to our hospital for definitive treatment. Ventricular tachycardia was induced by atrial and ventricular programmed electrical stimulations. The mechanism of the VT was determined to be re-entry. The earliest activation site was located at the mid-inferior septum of the hypoplastic left ventricle, in which Purkinje potentials were observed before the local ventricular electrogram. Radiofrequency catheter ablation (RFCA) was performed at this site to eliminate VT.

Discussion Most VTs originate from surgical scars in patients with congenital heart disease. Catheter ablation was feasible in scar-related VT. To the best of our knowledge, this is the first report of ILVT treated successfully with RFCA in a DORV patient who had undergone Fontan operation.

Keywords Idiopathic left ventricular tachycardia • Double outlet right ventricle • Fontan operation • Purkinje potential • Case report

Learning points

- The incidence of ventricular tachycardia (VT) in patients following Fontan operation is reported as 3.5%.
- Some cases of scar-related VT and outflow tract VT have been reported in repaired double outlet right ventricle (DORV) patients.
- Idiopathic left VT in DORV patient following Fontan operation has not previously been reported and can be successfully treated with radiofrequency catheter ablation.

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Introduction

The population of patients with congenital heart disease (CHD) is continuously increasing as more patients are reaching adulthood. A significant proportion of these patients will suffer from various arrhythmias, including atrial tachycardias, ventricular tachycardias (VTs), and ventricular fibrillation owing to the underlying CHD itself or as a sequela of interventional or surgical treatment.[1,2] The Fontan operation is the primary surgical technique used to treat patients with single-ventricle physiology. Arrhythmias are frequently observed and associated with morbidity and mortality in patients who undergo the Fontan operation.[3] The frequency of arrhythmias increases after the Fontan operation over time, with an prevalence of up to 50% at 20 years post-operatively.[4] Atrial tachycardia and sinus bradycardia are most frequently observed in these patients. The incidence of VT in patients who have undergone Fontan operation is reported as 3.5%.[4] Anatomical boundaries and surgical scars are important substrates for VT arrhythmogenesis in a patient with repaired CHD.[2]Purkinje-related VT is known as idiopathic left VT (ILVT) without organic heart disease.[5,6] Although scar-related VT[7] or outflow tract VT[8] have been reported in patients with repaired double outlet right ventricle (DORV), Purkinje-related VT has not been reported. Here, we report the case of a patient who developed ILVT after repair of DORV following a Fontan operation and was treated with radiofrequency catheter ablation (RFCA).

Timeline

Age	Event
At birth	The patient was diagnosed with double outlet right ventricle with a complete atrioventricular defect, pulmonary stenosis, and polysplenia
1 year of age	He underwent left Blalock–Taussig surgery
17 years of age	He underwent surgical repair, including extracardiac Fontan operation and common atrioventricular valve replacement
24 years of age	Wide QRS tachycardia was detected, which was treated by adenosine infusion; temporary oral intake of verapamil was also effective in treating the wide QRS tachycardia
31 years of age	He was referred to our hospital because the wide QRS tachycardia was not controlled
After admission	Idiopathic left ventricular tachycardia (ILVT) was treated by radiofrequency catheter ablation (RFCA) at the mid-inferior septum of the hypoplastic left ventricle
After discharge	After RFCA, ILVT had not reoccurred at 12 months follow-up

Case presentation

A 31-year-old man was diagnosed as having DORV with complete atrioventricular septal defect, pulmonary stenosis, and polysplenia at birth. He underwent left Blalock–Taussig surgery when he was 1 year old. At the age of 17 years, he underwent surgical repair, including extracardiac Fontan operation and common atrioventricular mechanical valve replacement. Five years after the Fontan operation, he visited a hospital owing to palpitations and dizziness. A 12-lead electrocardiogram detected wide QRS tachycardia, which indicated right bundle branch block with superior axis morphology (*Figure 1A*). The wide QRS tachycardia was treated using adenosine infusion. Although the morphology of the wide QRS tachycardia was different from that of the sinus rhythm (*Figure 1B*), it was considered as supraventricular tachycardia with aberrant conduction. He was prescribed pilsicainide, and temporary oral intake of verapamil was also effective in terminating the wide QRS tachycardia. However, owing to persistent wide QRS tachycardia, emergency hospital visit was required, and he was referred to our hospital for definitive treatment.

Physical examination showed pulse rate of 66 b.p.m., blood pressure of 110/72 mmHg, no leg oedema, and oxygen saturation on room air of 82%. Pulmonary auscultation was normal but cardiac auscultation revealed systolic murmur of Levine II/VI at two left sternal border. The systemic ventricle was the anatomical right ventricle. Transcutaneous echocardiography showed that systemic ventricular contraction was diffusely reduced with end-diastolic area of 51.7 cm^2 (normal range: 10–24 cm^2), end-systolic area of 36.4 cm^2 (normal range: 3–15 cm^2), and fractional area change of 29% (normal range: 35–56%).[9] Cardiac magnetic resonance imaging (MRI) showed that end-diastolic volume was 115 mL (normal range: 42–100 mL), end-systolic volume was 73 mL (normal range: 16–52 mL), and ejection fraction was 36% (normal range: 42–68%).[9] Prior to electrophysiological study, cardiac MRI showed diffuse late gadolinium enhancement (LGE) observed in the right ventricle and left ventricle (LV) (*Figure 2A–C*). The VT was presumed to originate from the scar or low voltage area. During the electrophysiological study, an atrial catheter was advanced from the left jugular vein to the extracardiac conduit, which could record the atrial electrogram and stimulate the atrial myocardium. Ventricular catheters were advanced retrograde from the right femoral artery to the ventricle. Although diffuse LGE region was observed in the right ventricle, apparent low voltage area and fractionated potential were not obtained from endocardial voltage mapping (*Figure 2D*). Wide QRS tachycardia was induced by programmed atrial and ventricular electrical stimulations and was terminated by single ventricular extrastimuli. Wide QRS tachycardia was diagnosed as VT, because atrial programmed stimulation could not produce wide QRS morphology, and activation mapping showed that the earliest activation site was located at the mid-inferior septum of the hypoplastic LV (*Figure 3*). At this site, abnormal fractionated potentials or low voltage zone were not observed, however, Purkinje potentials were observed before the local ventricular electrogram obtained during VT and sinus rhythm (*Figure 4A–C*). Diastolic pre-Purkinje potential was not observed. Entrainment pacing from this site was not performed, because VT was not sufficiently sustained. The paced QRS morphology at this site was similar to that of

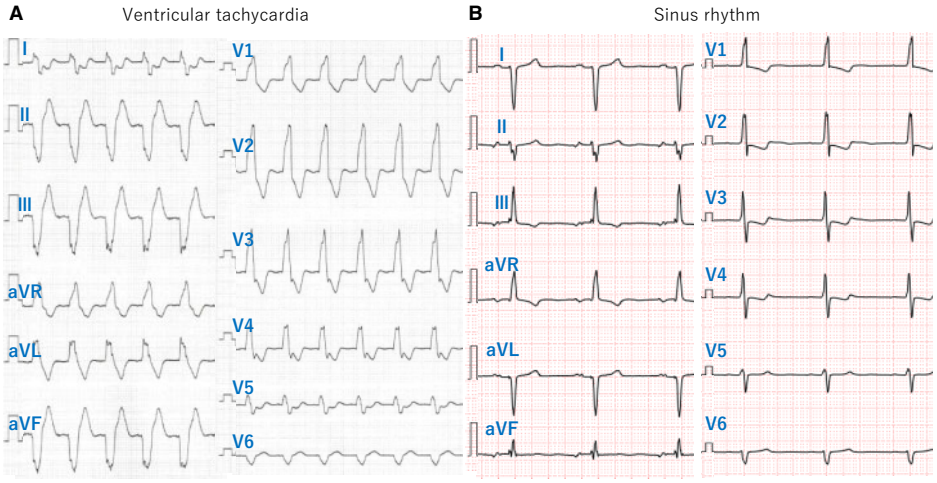

Figure 1 The electrocardiogram during ventricular tachycardia (*A*) show complete right bundle branch block, superior axis, and QRS duration of 156 ms. The electrocardiogram during sinus rhythm (*B*) show QRS duration of 115 ms.

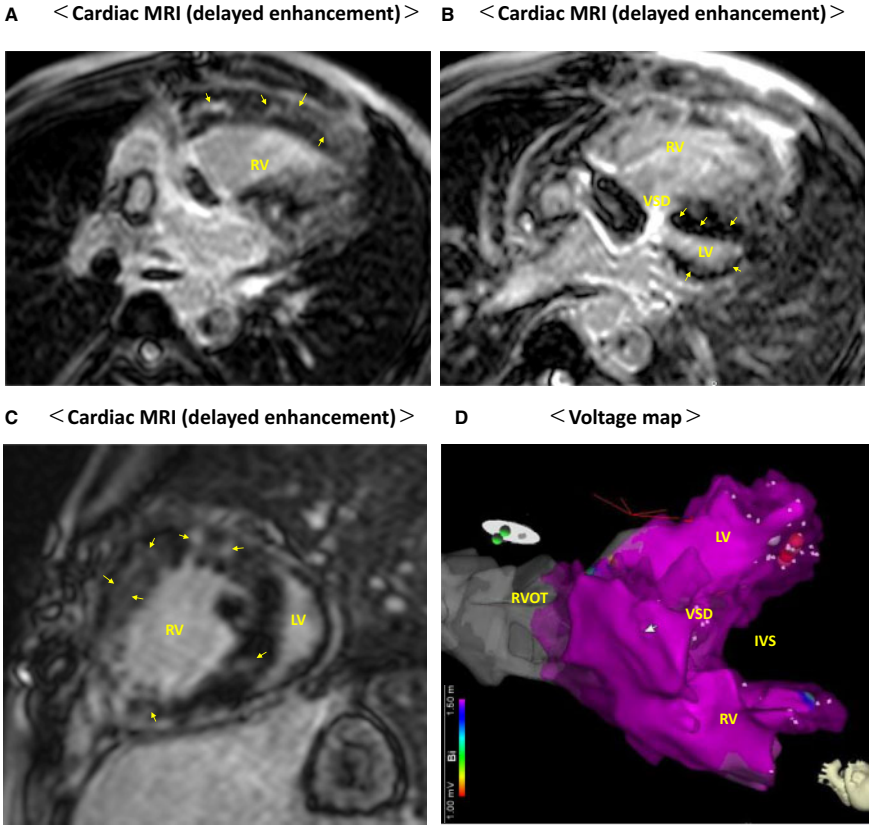

Figure 2 (*A–C*) Diffuse late gadolinium enhancement region observed in the right ventricle and the left ventricle on cardiac magnetic resonance imaging (yellow arrow). (*D*) No low voltage zone detected in either ventricle. LV, left ventricle; MRI, magnetic resonance imaging; RV, right ventricle; RVOT, right ventricular outflow tract; VSD, ventricular septal defect.

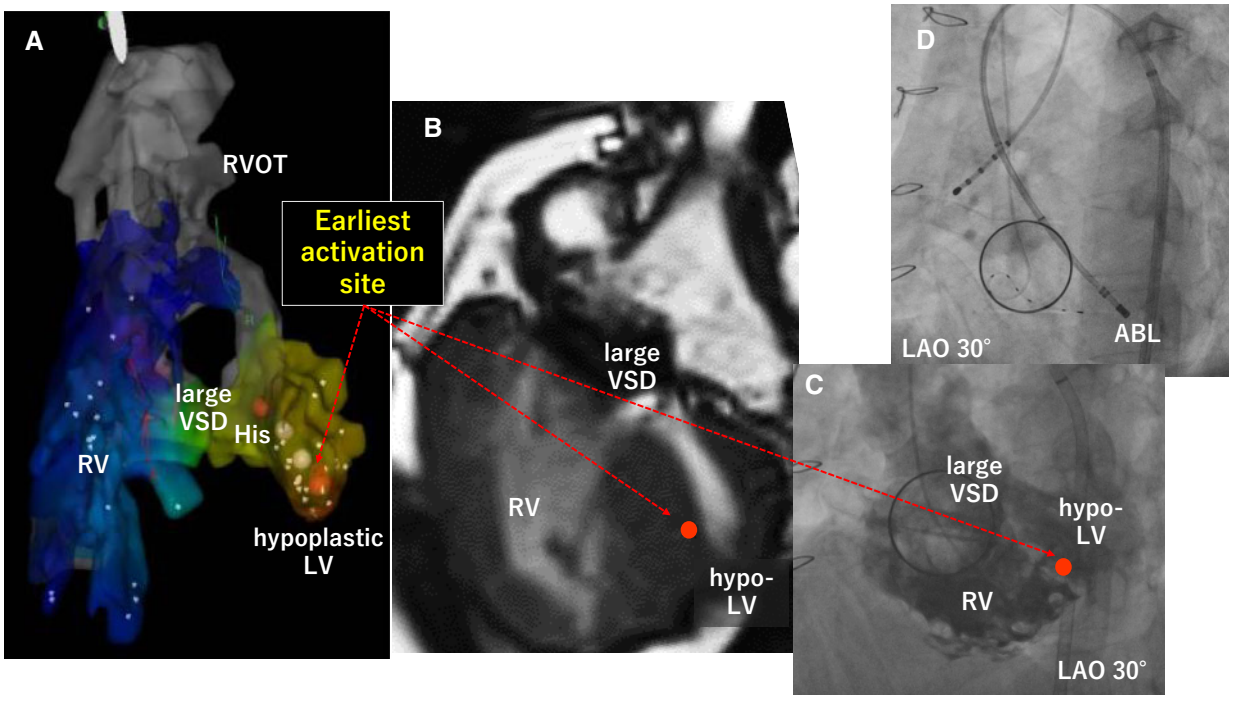

Figure 3 The earliest activation site during ventricular tachycardia is located at the inferior-septal wall of the hypoplastic left ventricle. [(*A*) 3D anatomical mapping, (*B*) magnetic resonance imaging, (*C*) ventriculography, (*D*) ablation catheter position]. ABL, ablation; LAO, left anterior oblique view; LV, left ventricle; RV, right ventricle; RVOT, right ventricular outflow tract; VSD, ventricular septal defect.

spontaneous VT. As the coupling interval of single ventricular extrastimuli became shorter, the return cycle of the VT became longer, which indicated that VT was caused by a re-entry mechanism and was not a triggered activity (*Figure 4D*). No further VT has been observed at 12 months following RFCA.

Discussion

To the best of our knowledge, this is the first report of successful treatment of ILVT using RFCA in a patient with DORV who had undergone Fontan operation. Patients with Fontan circulation are at a high risk of developing a variety of arrhythmias.[10] These arrhythmias are most often supraventricular tachyarrhythmias; however, ventricular tachyarrhythmias may occur as well.[11] The risk of sudden cardiac death in this population is as high as 9% during late follow-up.[12] Ventricular tachycardia has been reported in ~3.5–5% of patients who underwent Fontan surgery.[4,11] Most VTs originated from surgical scars in patients with CHD, and catheter ablation is feasible for scar-related VT.[13] Yang *et al.*[14] reported long term follow-up after VT ablation in patients with CHD. Ventricular tachycardia-free survival after multiple procedures was 85.4% (41 of 48) at a median follow-up of 52 months. Although 4 of 77 VTs were His-Purkinje-related VT (3 bundle branch re-entry and 1 interfascicular re-entry VT), the baseline heart disease was not DORV. Although the patient underwent extracardiac Fontan operation and common atrioventricular valve replacement, the origin of the VT was the mid-inferior septum of hypoplastic LV, which may not be associated with the scar of the previous Fontan operation. Furthermore, the substrate of the VT may not be influenced by the Fontan operation, but related to the baseline heart disease of DORV with atrioventricular septal defect. Double outlet right ventricle with atrioventricular septal defect is likely to have a conduction system abnormality or disturbance.[2,15,16]

On the other hand, verapamil-sensitive fascicular tachycardia is the most common form of ILVT. It was first recognized as an electrocardiographic entity by Zipes *et al.*[17] This VT is successfully suppressed by RFCA in the vicinity of the left posterior fascicle. Verapamil-sensitive fascicular tachycardia is characterized by the following: (i) during VT, retrograde activation of the His-bundle before QRS onset occurring with a significantly shorter HV interval during VT than that during sinus rhythm, (ii) the presence of common characteristics of re-entrant common ILVT, such as inducibility with ventricular and/or atrial stimulation, entrainment, and verapamil-sensitivity, and (iii) successful VT ablation in the left upper-middle ventricular septum, where the diastolic Purkinje potential was recorded during VT.[18]

Because entrainment pacing during VT was not performed, and diastolic pre-Purkinje potential was not observed, the precise diagnosis of VT was unknown. However, the mechanism of VT was re-entry, Purkinje potential preceded the VT, His bundle - Ventricular interval was normal, and verapamil was effective in suppressing the VT. This VT may therefore be the ILVT associated with DORV.

Other possible mechanisms of this VT are thought to be idiopathic Purkinje-related VT,[19] Purkinje-related VT associated with structure heart disease,[20] and bundle branch re-entry VT.

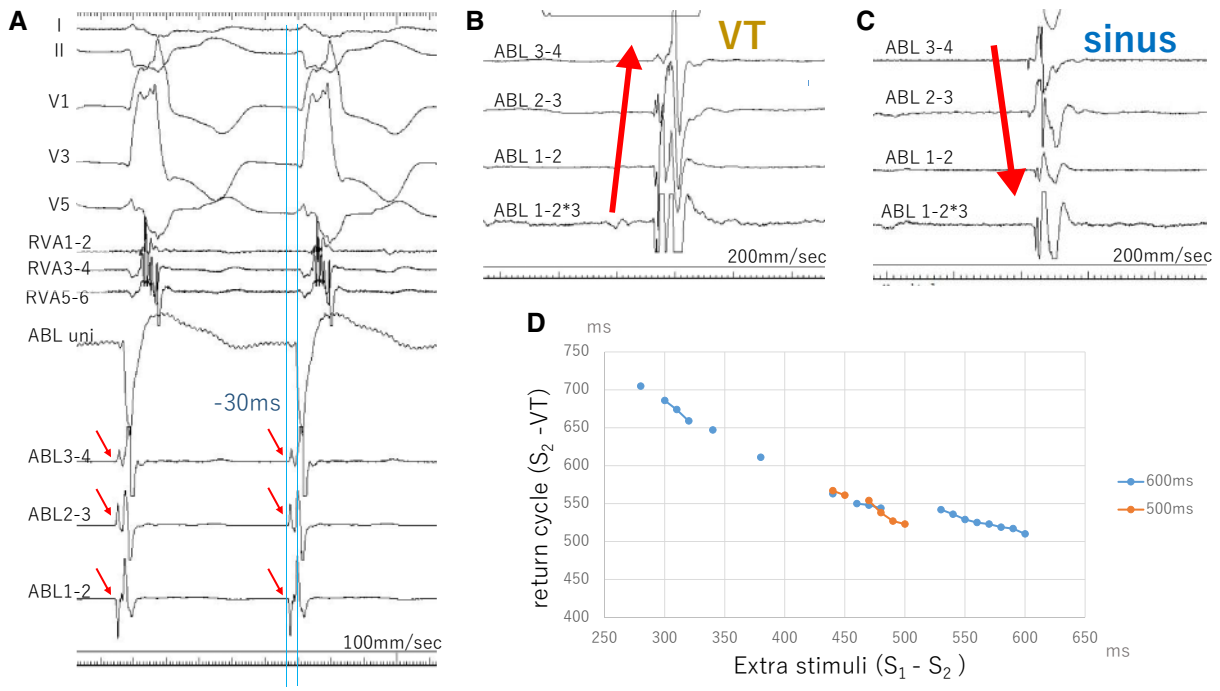

Figure 4 (*A*) The earliest activation potential during ventricular tachycardia is preceded by 30 ms from onset of QRS, in which Purkinje potential (red arrows) was recorded. (*B*) The activation sequence of Purkinje potential during ventricular tachycardia (red arrows) is from the distal to proximal. (*C*) The activation sequence of Purkinje potential during sinus rhythm (red arrows) was from the proximal to distal. (*D*) The interaction between the coupling interval of extrastimuli and return cycle. As the coupling interval of single ventricular extrastimuli becomes shorter, the return cycle of the ventricular tachycardia becomes longer. ABL, ablation; RVA, right ventricular apex; VT, ventricular tachycardia.

Conclusions

To the best of our knowledge, this is the first report of ILVT successfully treated using RFCA in a DORV patient who had undergone a Fontan operation.

Lead author biography

Masakazu Miyamoto, MD, graduated from Faculty of Medicine University of Miyazaki, Japan in 2004. He completed 2 years of Japanese post-graduate residency programme at Okayama University Hospital in Japan. Then he has worked as a fellow in cardiology at Kurashiki Central Hospital. Currently, he is working as a clinical fellow at Department of Cardiovascular Medicine, Okayama University Graduate School of Medicine, Dentistry, and Pharmaceutical Sciences, Japan. He is Fellow of the Japanese Society of Internal Medicine and Board Certified Member of the Japanese Circulation Society.

Supplementary material

Supplementary material is available at *European Heart Journal - Case Reports* online.

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Slide sets: A fully edited slide set detailing this case and suitable for local presentation is available online as Supplementary data.

Consent: The author/s confirm that written consent for submission and publication of this case report including image(s) and associated text has been obtained from the patient in line with COPE guidance.

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