Successful ablation of an outlet septum ventricular tachycardia in a double-outlet right ventricle patient who underwent an extracardiac Fontan operation

Masayoshi Mori, MD, Hisaaki Aoki, MD, PhD, Yoshihide Nakamura, MD, Yoichiro Ishii, MD, PhD, Kunihiko Takahashi, MD, PhD, Futoshi Kayatani, MD

From the Department of Pediatric Cardiology, Osaka Women's and Children's Hospital, Osaka, Japan.

Introduction

Arrhythmias are one of the most common causes of death in the late period post Fontan operation[1] and are associated with a 3.5% incidence of ventricular tachycardia (VT).[2] The extracardiac Fontan (EC-Fontan) has recently become the most commonly used approach in the Fontan operation. In such patients, catheter ablation (CA) is difficult to perform because the venous access to the heart is limited. A transcaval cardiac puncture (TCP) technique for gaining access to the heart chamber has previously been suggested for EC-Fontan patients.[3] To our best knowledge, there have only been a few reports describing VT ablation in Fontan patients.[4,5] In normal hearts, ablating preferential potentials leads to elimination of the multiple exit sites of outflow tract premature ventricular contractions (PVCs).[6] We report a case of VT from the outlet septum in a double-outlet right ventricle (DORV) after an EC-Fontan, wherein CA was performed via a TCP. Multiple PVCs were completely eliminated by ablating the site of distinct prepotentials on the aortic side of the outlet septum.

KEY TEACHING POINTS

- The outlet septum could be one of the origins of the ventricular arrhythmias in Fontan patient.
- Look for the early discrete potentials in outflow ventricular arrhythmias in tough cases.
- A transconduit approach is effective in extracardiac Fontan patients.

Case report

The patient had a left atrial isomerism, DORV, subaortic ventricular septal defect, common atrioventricular valve, and aorta originating from the right ventricle with subvalvular stenosis. She underwent pulmonary artery banding at 2 months, a Damus–Kaye–Stansel operation (DKS), bidirectional Glenn operation, and central shunt surgery at 1 year of age and an EC-Fontan operation at 3 years of age (Figure 1, Supplemental Figure 1). She had recurrent syncope and headaches since she was 13 years old. A 12-lead electrocardiogram (ECG) showed 2 types of PVCs and 24-hour Holter monitoring documented an episode of nonsustained VT (NSVT), which was not related to the syncope. Her neurological evaluation was normal. She underwent an electrophysiological study and no arrhythmias were induced by the electrophysiological study. An implantable loop recorder was implanted; however, she did not have any further episodes of syncope, and the device was removed owing to battery depletion 3 years postimplantation. After the removal, she had further syncopal episodes, and 24-hour Holter monitoring showed a significant burden of PVCs (12%) with NSVT. She underwent a CA at 18 years of age owing to recurrent syncope and high PVC burden.

The resting ECG (Figure 2A) exhibited a right upper axis in the frontal plane, qR in V_1, and R/S ratio <1 in V_5 and V_6 during sinus rhythm, suggesting that the atrioventricular node could be in a posterior position of the common atrioventricular canal. There were 2 types of PVCs with a left bundle branch block pattern and inferior axis. Each PVC morphology slightly differed in leads I and V1: R in I and rS in V_1 (PVC1) and RS in I and small rS in V_1 (PVC2). Those PVCs were thought to originate from the outflow tract with slightly different exit sites. Cardiac magnetic resonance imaging did not show any significant late gadolinium enhancement. A computed tomography scan (Figure 1) was performed to plan for the transcaval puncture and ablation strategy. We prepared for both a retrograde approach via

KEYWORDS Extracardiac Fontan operation; Catheter ablation; Premature ventricular contraction; Double-outlet right ventricle; Outlet septum; Transcaval puncture

(Heart Rhythm Case Reports 2022;8:543–547)

Funding Sources: This research did not receive any specific grant from funding agencies in the public, commercial, or not-for-profit sectors. Disclosures: All of authors have no conflicts to disclose. **Address reprint requests and correspondence:** Dr Hisaaki Aoki, Department of Pediatric Cardiology, Osaka Women's and Children's Hospital, 840 Murodo-cho, Izumi, Osaka 594-1101, Japan. E-mail address: aokihisaaki@hotmail.com.

https://doi.org/10.1016/j.hrcr.2022.04.015

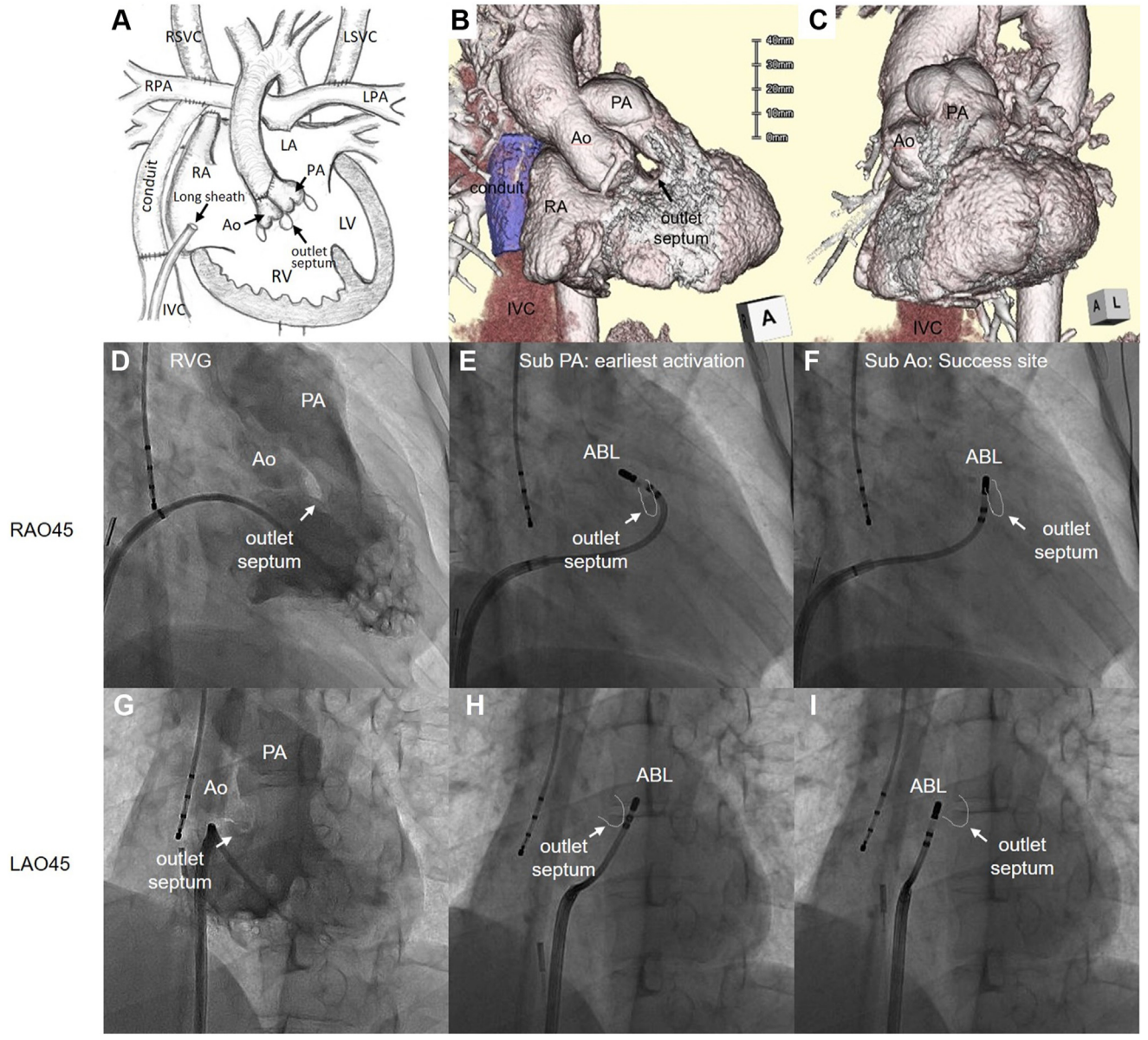

Figure 1 **A:** The scheme of the heart structure. The aorta (Ao) and pulmonary artery (PA) were anastomosed (Damus–Kaye–Stansel operation). The Agilis NxT sheath (Abbott Medical) was advanced into the atrium by a transcaval cardiac puncture, which involved a direct needle puncture through a segment of the inferior vena cava (IVC) adjacent to the wall of the pulmonary venous atrium. **B, C:** Three-dimensional contrast cardiac computed tomography. **D, G:** Right ventriculography images show the location of the subpulmonary (sub-PA) and subaortic (sub-Ao) sides of the outlet septum. **E, H:** The cine-angiography images indicate the location of the earliest activation potential during a premature ventricular contraction (PVC) on the sub-PA side of the outlet septum. **F, I:** The cine-angiography images indicate the successful site with a confirmed prepotential on the sub-Ao side of the outlet septum. ABL = ablation catheter; LA = left atrium; LPA = left pulmonary artery; LV = left ventricle; RA = right atrium; RPA = right pulmonary artery; RV = right ventricle; RVG = right ventricle angiography; SVC = superior vena cava.

the DKS anastomosis and antegrade approach via a transcaval puncture because of the possibilities of failure to access the target.

CA was performed under general anesthesia with propofol, remifentanil, and rocuronium. After an inferior vena cava angiography, we performed a TCP under intracardiac echocardiography guidance using a radiofrequency (RF) needle (NRG Transseptal Needle; Baylis Medical, Montreal, Canada) and Swartz introducer (SL-0, Fast-cath; St. Jude Medical, St. Paul, MN). A deflectable long sheath (Agilis NxT sheath; Abbott Medical) was inserted into the atrium after balloon dilatation of the puncture hole (Sterling 4-mm-diameter balloon; Boston Scientific, Tokyo, Japan). Diagnostic catheters were placed in the conduit via the internal jugular vein and right ventricle from the femoral artery in a retrograde manner. At baseline, there were frequent PVCs with 3 different morphologies, all with a left bundle branch block and inferior axis QRS morphology (Figure 2B). An NSVT with the PVC1 morphology was induced with an isoproterenol infusion. The PVCs were reproducibly elicited

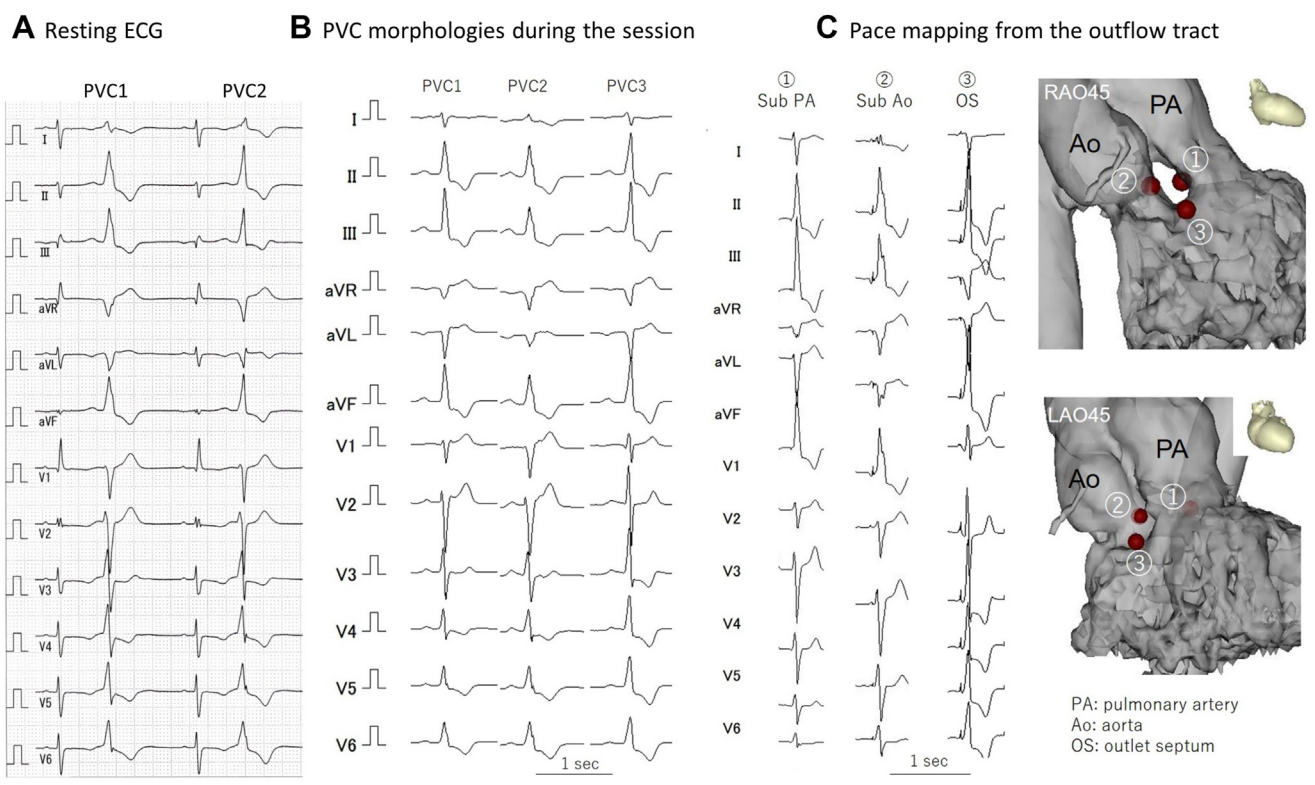

Figure 2 **A:** The resting 12-lead electrocardiogram (ECG) before the ablation. The 12-lead ECG shows a right upper axis in the frontal plane, qR in V_1, and R/S ratio <1 in V_5 and V_6. The premature ventricular contractions (PVCs) have a left bundle branch block pattern and inferior axis. Each PVC morphology slightly differs in lead I and V_1: R in I and rS in V_1 (PVC1) and RS in I and small rS in V_1 (PVC2). These PVCs could have originated from the outflow tract and it was assumed that their exit sites slightly differed. **B:** Twelve-lead ECGs exhibiting multiple PVCs during the session (PVC1, PVC2, and PVC3). **C:** Pace mapping obtained by pacing from the subpulmonary artery (sub-PA), subaortic (sub-Ao) region, and inferior part of the outlet septum. Good pace maps were obtained from the sub-PA side (PVC1) and sub-Ao side (PVC2) of the outlet septum.

by programmed stimuli. The His bundle electrogram was confirmed inferior to the common atrioventricular valve and far from the outflow septum. Catheter manipulation was performed with an Agilis sheath via the TCP, because the catheter manipulation was challenging via the retrograde approach owing to the aortic stenosis and DKS anastomosis.

A PVC activation map using CARTO III mapping and a NAVISTAR catheter (Biosense Webster, Irvine, CA) revealed the earliest activation in the outlet septum. The PVC1 activation map on the subpulmonary side of the outlet septum revealed the earliest site of the ventricular activation, which was 32 ms earlier than the QRS complex on the surface ECG (Figure 1E and 1H and Figure 3B). The prepotential was recorded at that site, which preceded the QRS complex by 61 ms. Pace mapping nearly matched the QRS morphology of PVC1 (Figure 2C). The PVC transiently disappeared after delivering RF energy for 60 seconds at a temperature below 55°C with a power limit of 40 W. The earliest ventricular activation during PVC2 was on the subaortic side of the outlet septum, which was 28 ms earlier than the QRS complex on the surface ECG. However, its discrete prepotential, which was 106 ms earlier than the QRS complex, was recorded (Figure 1F and 1I and Figure 3A), and its pace mapping coincided with the PVC2 morphology (Figure 2C).

All PVCs were eliminated by delivering RF energy on the subaortic side of the outlet septum (Figure 3A). At 3 months postablation, she has not had any syncopal episodes and Holter monitoring revealed a PVC burden of 2% with no VT.

Discussion

To the best of our knowledge, this is the first report of a successful CA of NSVT originating from the outlet septum in a patient with a DORV after an EC-Fontan operation via a transcaval puncture. Three PVC morphologies were simultaneously eliminated by an RF application at the site of the distinct prepotential electrograms during the PVC on the subaortic side of the outlet septum (not at the earliest ventricular activation site during the PVC). Three studies have discussed percutaneous VT ablation in patients that underwent a Fontan operation, which were ablated via a retrograde approach.[4,7,8]

The approach to the heart is restricted after an EC-Fontan operation: retrograde approach and transbaffle or transcaval approaches. Catheter manipulation can be quite challenging via a retrograde approach in the setting of subaortic stenosis. However, a transbaffle and transcaval approach could be useful for CA or the creation of a fenestration. In this case, the subaortic stenosis made the retrograde approach difficult.

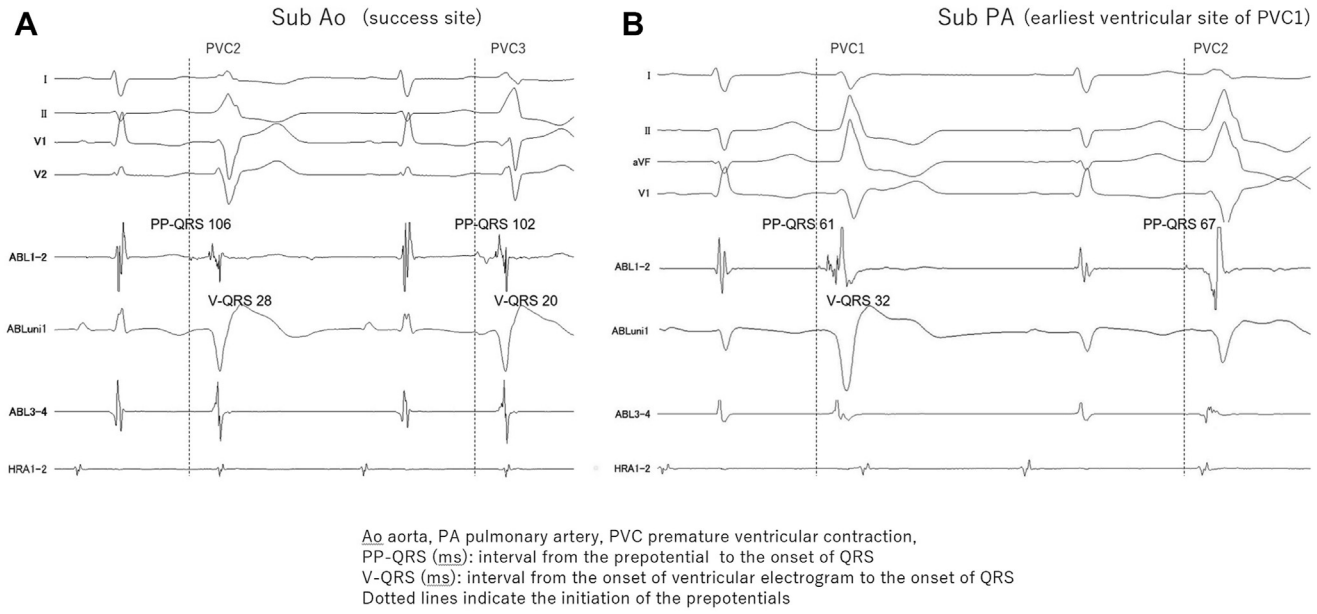

Figure 3 The intracardiac electrograms at the ablation sites. **A:** The earliest ventricular activation during premature ventricular contraction (PVC) 2 was on the subaortic side of the outlet septum, which was 28 ms earlier than the QRS complex on the surface electrocardiogram (ECG). However, a discrete prepotential, which was 106 ms earlier than the QRS complex, was recorded. All PVCs were eliminated by delivering radiofrequency energy on the subaortic side of the outlet septum. **B:** PVC1 activation map on the subpulmonary side of the outlet septum revealed the earliest site of ventricular activation, which was 32 ms earlier than the QRS complex on the surface ECG. A prepotential was recorded on that side, which preceded the QRS complex by 61 ms. The pace map nearly matched the QRS morphology of PVC1.

The transcaval approach using a TCP and deflectable sheath made it easier to manipulate the ablation catheter. Thus, when a PVC/VT originating from the outlet septum in Fontan patients is suspected, the operators should be prepared to attempt multiple approaches to accomplish a successful ablation.

Idiopathic ventricular arrhythmias commonly originate from the right (RVOT) and left ventricular outflow tracts. The adult RVOT forms from the embryonic outflow tract, which is primarily composed of myocardium and exhibits slow conduction and spontaneous activity. During development, the embryonic outflow tract acquires a working myocardial phenotype (eg, fast conduction) and transforms into the RVOT. A small ring of primary myocardium, however, remains just below the pulmonary valve, and this may give rise to automaticity, as seen in patients with idiopathic RVOT tachycardia.[9] On the other hand, the "dead-end tract" has been linked to the occurrence of ventricular arrhythmias. This is a remnant of the developing conduction system and is seen in addition to the right and left bundle branch, fading out on the crest of the muscular ventricular septum.[10] Hachiya and colleagues[11] speculated that prepotentials represent preferential conduction (ie, the activation of a tract connecting the focus of the arrhythmia to the ventricular myocardium).[11] In this case, the multiple PVCs were completely eliminated by delivering RF on the aortic side of the outlet septum, rather than on the pulmonary artery side, which was the earliest ventricular site during the PVCs. This suggested the existence of preferential conduction (Supplemental Figure 2). Although there is a secondary interventricular foramen (ie, a defect between the muscular ventricular septum and conus septum [outlet septum]), the outlet septum inserts into the ventriculo-infundibular fold and continues to the muscular ventricular septum, wherein a preferential pathway can exist. When it is difficult to ablate outflow PVCs/VTs in complex congenital heart diseases, the existence of preferential conduction should be considered.

Conclusion

To the best of our knowledge, this is the first reported case of the successful ablation of NSVT originating from the outlet septum via the transcaval approach in a patient with a DORV after an EC-Fontan operation. Multiple PVCs were completely eliminated by ablating the site of distinct prepotentials on the aortic side of the outlet septum, suggesting the existence of preferential conduction. Thus, in such cases, it is important to study the patient's anatomy, conduction system, mechanism of the arrhythmia, and strategic approach, as well as prepare the required equipment.

Acknowledgments

We are grateful to Hiroko Asakai for carefully proofreading the manuscript. We would like to thank enago (www.enago.jp) for the English-language editing.

Appendix

Supplementary data

Supplementary data associated with this article can be found in the online version at https://doi.org/10.1016/j.hrcr.2022.04.015.

References

1. Alsaied T, Bokma JP, Engel ME, et al. Factors associated with long-term mortality after Fontan procedures: a systematic review. Heart 2017; 103:104–110.
2. Stephenson EA, Lu M, Berul CI, et al. Arrhythmias in a contemporary Fontan cohort: prevalence and clinical associations in a multicenter cross-sectional study. J Am Coll Cardiol 2010;56:890–896.
3. Moore JP, Gallotti RG, Tran E, et al. Ten-year outcomes of transcaval cardiac puncture for catheter ablation after extracardiac Fontan surgery. Heart Rhythm 2020;17:1752–1758.
4. Miyamoto M, Nishii N, Morita H, et al. Ablation for idiopathic left ventricular tachycardia in a patient with double outlet right ventricle who underwent Fontan operation: a case report. Eur Heart J Case Rep 2020;4:1–6.
5. Izumi G, Yokoshiki H, Tachibana T, et al. A hybrid therapy for arrhythmogenic congestive cavity in a single ventricle. Eur J Cardiothorac Surg 2021; 59:911–913.
6. Shirai Y, Goya M, Isobe M, et al. Preferential pathway pacing within the aortic sinus of valsalva: strong evidence for the existence of preferential conduction with different exit sites traversing the ventricular septum. J Cardiovasc Electrophysiol 2015;26:805–808.
7. Reiter T, Ritter O, Nordbeck P, et al. MRI-guided ablation of wide complex tachycardia in a univentricular heart. World J Cardiol 2012;4:260–263.
8. Correa R, Sherwin ED, Kovach J, et al. Mechanism and ablation of arrhythmia following total cavopulmonary connection. Circ Arrhythm Electrophysiol 2015;8:318–325.
9. Rivaud MR, Blok M, Jongbloed MRM, et al. How cardiac embryology translates into clinical arrhythmias. J Cardiovasc Dev Dis 2021;8:70.
10. de Vries L, Hendriks A, Szili-Torok T. The “dead-end tract” and its role in arrhythmogenesis. J Cardiovasc Dev Dis Apr 5 2016;3:11.
11. Hachiya H, Yamauchi Y, Iesaka Y, et al. Discrete prepotential as an indicator of successful ablation in patients with coronary cusp ventricular arrhythmia. Circ Arrhythm Electrophysiol 2013;6:898–904.