Successful Ablation of Atrial Tachycardia Originating from Inside the Single Atrium and Conduit After a Fontan Operation
Using an Ultra-High-Density 3-Dimensional Mapping System

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Summary
An 18-year-old male who had a past medical history of an intracardiac total cavopulmonary connection (TCPC) operation was referred to our hospital for radiofrequency catheter ablation (RFCA) of supraventricular tachycardia (SVT). Two types of SVTs were induced, and 3-dimensional (3D) maps were created using an ultra-high-density 3-dimensional mapping system (Rhythmia). The earliest atrial activation site (EAAS) of SVT1 was at the superior part of the conduit, and the EAAS of SVT2 was at the inferior part of the single atrium (SA). The SVTs were terminated by energy deliveries to the EAAS from the conduit in SVT1 and from inside the single atrium in SVT2. Detailed maps of the SVTs were important to understand the mechanisms of the SVTs. The Rhythmia system was useful for the detailed mapping of complex arrhythmias. The use of Rhythmia in patients after a TCPC is difficult, because puncturing the TCPC conduit and proceeding and manipulating the Orion catheter via a narrow puncture hole are difficult. We were the first to succeed in ablating two atrial tachycardias (ATs) originating from the inside and outside of the conduit after a TCPC operation by using an ultra-high-density 3-dimensional mapping system.

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It is widely known that a supraventricular tachycardia (SVT) may become life-threatening in patients after a Fontan operation.1 It is rarely reported to be about 17-42% within several years after the operation.2,3 SVTs after a Fontan operation could be a major cause of morbidity and mortality and usually are very difficult to control.4 Catheter ablation of SVTs in these patients is sometimes technically difficult but has been reported to be safe and effective.5,6 Intra-atrial reentrant tachycardia (IART) is the most common SVT, and the incidence of focal atrial tachycardia (AT) is relatively rare in these patients.2,5 The type of Fontan was associated with the presence of IART. Patients with an atriopeumonial connection were more likely to have IART (19%) than those with intracardiac lateral tunnel (7%), extracardiac lateral tunnel (5%), or extracardiac conduit (2%) type.7

We report a case of focal AT originating from the superior part of the conduit and inferior part of a single atrium (SA) in a patient after a Fontan operation. An ultra-high-density 3-dimensional mapping system was useful for detecting the origin of this focal AT.

Case Report
An 18-year-old male was referred to our hospital for radiofrequency catheter ablation (RFCA) of a supraventricular tachycardia (SVT). At birth, he was diagnosed with pulmonary atresia, double outlet right ventricle, hypoplastic left ventricle, and ventricular septal defect. An intracardiac total cavopulmonary connection (TCPC) with a fenestration using a Gore-Tex conduit was performed at 17 years old. SVT was first noted one day after the operation. Amiodarone was administered, and the SVT was partially controlled; however, 3 months after starting amiodarone, it was discontinued due to an elevation of KL-6. The tachycardia recurred when he was 18 years old, and many cardioversions were required to terminate that tachycardia, and therefore, he was referred to our hospital to undergo RFCA.

Cardiac computed tomography (CT) was performed before the RFCA to analyze the 3-dimensional (3D) anatomy, and the RFCA was performed with the use of a high-density 3D mapping system (Rhythmia; Boston Scientific, Marlborough, MA) and an Orion catheter (Boston). These procedures were performed under general an-
esthesia by an anesthesiologist.

A 5 Fr decapolar electrode catheter (Snake®; Japan Lifeline, Tokyo, Japan) and 4 Fr 4-pole electrode catheter (Inquiry®; Abbot, St. Paul, MN) were positioned in the left pulmonary artery (PA) from the internal jugular vein and the single ventricle (SV) from the femoral artery. Due to right femoral vein stenosis, an 8.5 Fr steerable sheath (Agilis®; Abbot, St. Paul, MN) was introduced into the TCPC conduit from the left femoral vein. Intravenous heparin was used to maintain the activated clotting time at 300-350 seconds. Angiography from the conduit was performed to confirm the location of the fenestration along the TCPC, and then, the Agilis sheath was introduced from the fenestration into the SA. Pulmonary venous arteriography was performed from the Agilis sheath.

An atrial substrate map during sinus rhythm using an Orion catheter revealed a wide low-voltage area (< 0.5 mV) in the contact region between the SA and conduit. There was no ventriculoatrial conduction during ventricular burst pacing. An SVT was induced during atrial burst pacing under an isoproterenol (ISP) infusion. Two types of SVTs were induced during this study; the cycle length (CL) of SVT1 was 341 ms and the earliest atrial activation site (EAAS) in PA catheter was at PA 7-8 (Figure 1A); the CL of SVT2 was 253 ms and the EAAS in PA catheter was at PA 5-6 (Figure 1B). Each SVT spontaneously converted to the other SVT without any stimulation. Ultra-high-density 3D mapping was performed during both SVT1 and SVT2. The Rhythmia system could automatically distinguish each SVT without manual reannotation by a change in the earliest activation site of the reference catheter in the PA. The EAAS of SVT 1 was at a superior part of the conduit (Figure 2A), and the EAAS of SVT 2 was at an inferior part of the SA (Figure 3A). Although we tried to perform entrainment pacing, the tachycardias spontaneously changed from one to the other or terminated, and we could not measure the post-pacing interval and tachycardia CL. We considered the mechanism of the tachycardia was a micro-reentrant AT or focal AT from the distribution pattern of the activation maps. RFCA was attempted at the EAAS of each tachycardia. The local potential at the EAAS of a SVT1 was 37 ms earlier than the reference potential; however, the voltage amplitude was 0.0565 mV (Figure 1A and Figure 2A). The local potential of the EAAS of SVT2 was 35 ms earlier than the reference potential; however, the voltage amplitude was 0.393 mV (Figure 1B and Figure 3A). Ablation energy (30 W) was delivered to the EAAS of SVT1 from inside the baffle and SVT2 from inside the SA. Although the tachycardia was terminated by the energy delivery, the tachycardia could still be induced by atrial stimulation. Several energy deliveries were required before the tachycardias could no longer be induced. During a follow-up after the ablation, the patient was aware of palpitation due to atrial premature constrictions 3 months after ablation. Administration of flecainide was started, and the symptoms were improved. However, no further recurrences of SVT have been observed.

Discussion

To the best of our knowledge, this is the first case report to describe the detailed mapping of SVTs after a TCPC operation using an ultra-high-density 3D mapping system. Most SVTs after a Fontan operation are scar-related macroreentrant ATs, atrial flutter, or atrial fibrillation, and the incidence of focal ATs has been relatively rare. Our patient had two different focal ATs. In this patient, there was no space outside the heart to place the TCPC conduit in mediastinal space, and the surgeon sutured the conduit inside the SA (intracardiac TCPC). Because the surgeon worried about bleeding between the
conduit and inferior vena cava or PA, he sutured the atrial muscle around the conduit to prevent bleeding. We could confirm this muscle by cardiac CT (Figures 2B, 3B). This muscle could be considered as an arrhythmogenic substrate in this patient. As a result of this operation, the SVT originated from the contact region between the conduit and SA. The arrhythmogenicity of the TCPC is frequently related to the index surgical approach. Surgeons should be aware of the different mechanisms of postoperative arrhythmias and perform perioperative ablation if possible.

The local amplitude of SVT1 in the conduit was very small; however, the Rhythmia system could accurately describe the activation map. The voltage amplitude of SVT1 and SVT2 was 0.0565 mV and 0.393 mV, respectively, which was generally considered as low voltage or scar tissue (bipolar voltage ≤ 0.5 mV). The electrodes of the Orion catheter have a 0.4-mm² area with a 2.5-mm inter-electrode spacing and are useful to record tiny potentials. This characteristic of the catheter was useful for the accurate mapping of SVT1.

Our patient had an adequate size of the fenestration...
confirmed by angiography from the conduit. Without a transbaffle puncture, the steerable sheath was easily placed into the SA. The steerable sheath via the fenestration provided the greatest stability of the mapping catheter and was useful for creating the detail 3D map. Postprocedural oxygen desaturation, which is caused by a right-to-left shunt via a fenestration, is observed in some patients after ablation in Fontan patients.7) Our patient also had oxygen desaturation; however, it improved one day after the procedure.

Placement of a stable atrial sensing electrode as a locational reference and potential reference is a crucial step for accurate 3D maps. Several doctors chose to place the reference catheter in the esophagus, but a catheter placement in the PA could be more useful than in the esophagus.5) In our patient, the reference catheter was positioned in the PA, and it was more stable for acquiring the 3D map. The limitations of the reference catheter positioned outside the pulmonary venous atrium have also been reported due to a reduced ability to detect abrupt changes in the activation pattern during the tachycardia by shifting from one tachycardia circuit to another. In the present patient, SVT1 and SVT2 spontaneously shifted between each other. However, the Rhythmia system could distinguish between each SVT by the change in the EAAS on the reference catheter in the PA.

Conclusion

The border region between the conduit and SA could be an arrhythmogenic substrate. The ultra-high-density 3D mapping system was useful for recording the activation map of the AT after the Fontan operation.

Disclosure

Conflicts of interest: All authors declared no conflict of interest associated with this report.

References