Atrial Tachycardia in a Patient with Extracardiac Conduit Fontan Circulation

ABSTRACT

Electrophysiology (EP) studies and radiofrequency catheter ablation (RFCA) are challenging in patients who have undergone extracardiac conduit Fontan procedures, because of the difficult vascular access. Here, we report on a 14-year-old male patient who underwent extracardiac conduit Fontan procedure for a double-inlet left ventricle, complete transposition of the great arteries, and large ventricular septal defect. The EP study was performed via a trans-conduit puncture. Focal atrial tachycardia originating from the mid portion of the interatrial septum was induced. RFCA of the origin of atrial tachycardia was successfully performed. EP studies and RFCA are feasible via a trans-conduit puncture in patients with extracardiac conduit Fontan circulation.

Key words: • atrial tachycardia • congenital heart disease • Fontan procedure

Introduction

The survival rate of patients with complex congenital heart disease has recently improved, most likely due to the development of new surgical techniques and improved perioperative medical management.1 As the number of adult patients with congenital heart disease has increased, arrhythmias and heart failure are becoming growing issues in these patients.1 Thus, it is not surprising that the demand for electrophysiologic (EP) studies and radiofrequency catheter ablation (RFCA) is increasing. EP studies and RFCA are challenging in patients who have undergone extracardiac conduit Fontan procedures for the palliative treatment of congenital heart disease, because the systemic venous blood is not drained into the heart in these techniques. Here, we report a case of focal atrial tachycardia, which was ablated via a trans-conduit puncture in a patient who had undergone an extracardiac conduit Fontan procedure.

Case

A 14-year-old male patient visited the emergency room complaining of palpitations for 3
Figure 1. (A) electrocardiogram during palpitation shows regular narrow QRS tachycardia with short RP interval. (B) electrocardiogram during sinus rhythm.
Figure 2. Cardiac CT shows findings compatible with complete transposition of the great arteries, double-inlet left ventricle, large ventricular septal defect and extracardiac Fontan conduit. 
Ao, aorta; C, Fontan conduit; LA, left atrium; PA, pulmonary artery; RA, right atrium; rV, rudimentary ventricle; V, ventricle.

Figure 3. (A) Fontan conduit angiography (B) Fluoroscopic image of performing trans-conduit puncture (C) Intracardiac echocardiography of the trans-conduit puncture (D) The Brockenbrough transseptal needle and the Swartz transseptal introducer sheath with holding the dilator tip of the Swartz sheath with the snare catheter to prevent it from sliding up along the conduit wall.
C, Fontan conduit; HV, hepatic vein; ICE, intracardiac echocardiography; IVC, inferior vena cava; LPA, left pulmonary artery; RA, right atrium.
hours. He had experienced several episodes of palpitations during the past year. His blood pressure was 82/49 mmHg. The electrocardiogram (ECG) showed regular, narrow QRS tachycardia with a rate of 160 beats/min and a short RP interval (Figure 1A). In the emergency room, the tachycardia spontaneously converted into sinus rhythm (Figure 1B). The QRS morphology of the tachycardia was similar to that in sinus rhythm. When he was 10 days old, he was diagnosed with double-inlet left ventricle (DILV), complete transposition of the great arteries (TGA), and large ventricular septal defect (VSD). When he was 5 months old, the bidirectional cavopulmonary shunt and interatrial septectomy were performed for palliation. At the age of 1 year, an extracardiac conduit Fontan procedure was performed with the autologous pericardium. We decided to perform the EP study for diagnosis and treatment of the tachycardia. Cardiac computed tomography (CT) was performed for assessment of the heart anatomy, showing findings compatible with TGA, DILV, large VSD, functional single ventricle, and extracardiac Fontan conduit (Figure 2).

Both femoral veins were punctured. Conduit angiography was performed with a Berman-type angiography catheter (Arrow International, Reading, PA, USA) (Figure 3A). Two SR-0 Swartz transseptal introducer sheath (St Jude Medical, St Paul, MN, USA), a snare catheter (PFM Medical, Nonnweiler, Germany), and an intracardiac echocardiography catheter (AcuNav, Siemens, Mountain View, CA, USA) were inserted into the Fontan conduit via the femoral veins. A BRK-1 Brockenbrough transseptal needle (St Jude Medical) was inserted into the Swartz sheath, and the dilator tip of the Swartz sheath was held with the snare catheter to prevent it from sliding up along the conduit wall (Figure 3B and D). We punctured the wall between the conduit and the right atrium with the Brockenbrough transseptal needle under intracardiac echocardiography guidance (Figure 3C). Right and left atrioography was performed with the pigtail catheter via the trans−conduit puncture. A deflectable decapolar catheter (St
Figure 5. Intracardiac electrogram of induction (A) and maintenance (B) of tachycardia. The tachycardia was maintained in spite of premature ventricular complex (red box in A) and atrioventricular block (red box in B). The atrioventricular or ventriculoatrial interval was varying.
Jude Medical) was placed in the high left atrium via the Swartz sheath and a decapolar catheter (St Jude Medical) was placed in the ventricle via the aorta (Figure 4A). The initial rhythm was normal sinus rhythm. During ventricular pacing and single ventricular extrastimuli, the atrial electrogram showed one-to-one ventriculoatrial conduction with decremental properties. During the atrial pacing of 240 ms and infusion of isoproterenol at a rate of 2 μg/min, tachycardia with a 290 ms cycle length was induced. Tachycardia was maintained despite the presence of a premature ventricular complex and atrioventricular block (Figure 5A and B). Therefore, atrioventricular reentrant tachycardia could be excluded. During tachycardia, the atrioventricular or ventriculoatrial interval varied (Figure 5B). It was not compatible with atrioventricular nodal reentrant tachycardia. The tachycardia was not entrainable by ventricular pacing. The decapolar catheter was moved to the right atrium (RA) side in the Fontan conduit, and an irrigated ablation catheter (Thermocool, Biosense Webster, Diamond Bar, CA, USA) was inserted into the atrium via the conduit puncture for tachycardia mapping (Figure 4B). During tachycardia, 3-dimensional electroanatomical mapping was performed with CARTO (Biosense Webster, Diamond Bar, CA, USA). The tachycardia was originating from the mid portion of the remnant interatrial septum (Figure 6). The tachycardia was compatible with focal atrial tachycardia originating from the interatrial septum. During sinus rhythm, we mapped the His bundle potentials. The His bundle area was located in the lower posterior potion of the interatrial septum. The origin of the tachycardia was away from the His bundle area by 13.6 mm (Figure 6B). We performed RFCA of the origin of the atrial tachycardia by delivering 30 watts of RF energy for 90 s with the irrigated ablation catheter during sinus rhythm. The procedure ended after we confirmed that the tachycardia was not induced.

**Figure 6.** LAO view of 3-dimensional electroanatomical mapping shows the origin (black arrow) of focal atrial tachycardia originating from the mid portion of the septum. His, His bundle area; LA, left atrium; LAA, left atrial appendage; LAO, left anterior oblique view; LSPV, left superior pulmonary vein; RA, right atrium; RSPV, right superior pulmonary vein.
by the programmed electric stimulation and isoproterenol infusion. The patient had no symptom and maintained sinus rhythm for 6 months after RFCA.

Discussion

The case was focal atrial tachycardia originating from the septum in a patient who had undergone an extracardiac conduit Fontan procedure. We performed an EP study and RFCA of the origin of the focal atrial tachycardia successfully via the trans-conduit puncture.

The lifelong prevalence of atrial tachycardia in patients with extracardiac Fontan circulation is approximately 50%, and it is considerably higher than in the normal population. In patients with extracardiac conduit Fontan circulation, it is difficult to perform an EP study, because the heart is completely excluded from the systemic venous system. There were previous case reports of EP studies and RFCA via various routes in patients with Fontan circulation, including via a trans-thoracic puncture, sternotomy approach, trans-apical access and trans-conduit puncture. The EP catheters can be transvascularly placed via 2 pathways: the trans-conduit puncture and retrograde transaortic approach. The approach via the trans-conduit puncture is suitable for gaining access to the atrium and the retrograde transaortic approach is suitable for gaining access to the ventricle. It is challenging to puncture the Fontan conduit because fibrosis forms around the conduit. In addition, the conduit wall is vertical—unlike the interatrial septum—and the transseptal needle tends to slide up along the conduit wall instead of puncturing it. The use of a Brockenbrough transseptal needle while holding the dilator tip of the Swartz sheath with a snare catheter is a useful method for puncturing the Fontan conduit. A large-curve BRK-1 transseptal needle is superior to a small-curve BRK needle. Moreover, the radiofrequency transseptal needle can be a good option for puncturing the fibrotic Fontan conduit.

In patients with congenital heart disease, the EP study and RFCA are very challenging because of the unusual anatomy of the heart. In addition, it is common for patients to have vascular anomalies including a persistent left superior vena cava and inferior vena cava interruption. It is important that the operator be completely aware of the anatomy of heart and vessels of each patient. Every patient has a unique heart structure, even though this patient group has the same diagnosis of congenital heart disease. The operator needs to review and understand the previous cardiac surgery and intervention. The operator should make a meticulous plan for the procedure, taking into consideration the types of EP catheters to be used for each chamber, pathways to be used for positioning of the EP catheters, and appropriate angles for the X-ray beam to improve visualization. The operator needs to discuss the current hemodynamics and long-term prognosis of the patient with the pediatric cardiologists. Given the complex heart anatomy, cardiac CT and a 3-dimensional electroanatomic mapping system are necessary for guiding the procedure. Intracardiac echocardiography can be helpful for real-time visualization of the anatomy and EP catheters. The activated coagulation time should be maintained at 350–400 ms by heparin infusion during the EP study in patients with a single ventricle, as the catheters are placed in the systemic chambers.

In the present case, the remnant interatrial septum might become arrhythmogenic after septectomy due to degenerative changes of the interatrial septum. This patient is likely to develop
atrial tachyarrhythmia originating from other parts of the atrium. In addition, an advanced atrioventricular block can occur in the future, although the peri-procedural ECG showed first degree atrioventricular block. Thus, the patient will require long-term follow-up.

**Conclusion**

EP studies and RFCA are feasible via a trans-conduit puncture in patients with extracardiac conduit Fontan circulation.

**References**