Atrioventricular reentrant tachycardia in a child with tricuspid atresia
A case report of catheter ablation

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Abstract

Rationale: Atrioventricular reentrant tachycardia (AVRT) is the most common supraventricular tachycardia occurring in children. However, in complex congenital heart disease patients with a different heart anatomy and conduction system morphology, accessory pathway modification may be particularly challenging because of distortion of typical anatomic landmarks.

Patient concerns: A 10-year-old boy with tricuspid atresia and history of bidirectional Glenn operation had recurrent chest distress and palpitation for 3 months. He had multiple hospitalizations for narrow-QRS tachycardia with poor hemodynamic tolerance, despite the use of adenosine and amiodarone.

Diagnoses: AVRT. Tricuspid atresia with secundum atrial septal defect, large ventricular septal defect, and right ventricular outflow tract stenosis.

Interventions: Cardiac catheterization, electrophysiological examination, and ablation.

Outcomes: The child has not had a recurrent AVRT during 6 months of follow-up and is waiting for Fontan operation.

Lessons: Since there is an increased risk of accessory pathways in patients with tricuspid atresia, all these patients should be checked before the Fontan operation to exclude congenital accessory pathways.

Abbreviations: AV = atrial-ventricular, AVRT = atrioventricular reentrant tachycardia, LV = left ventricular.

Keywords: Atrioventricular reentrant tachycardia, catheter ablation, children, tricuspid atresia

1. Introduction

Atrioventricular reentrant tachycardia (AVRT) is the most common supraventricular tachycardia occurring in children. Catheter ablation is considered the definitive therapy of choice in the majority of patients.[1] However, in complex congenital heart disease patients with a different heart anatomy and conduction system morphology, accessory pathway modification may be particularly challenging because of distortion of typical anatomic landmarks.[2,3] Here, we report a case of AVRT in a child with tricuspid atresia and history of bidirectional Glenn operation, and describe the procedure of catheterization and ablation.

The ethics committee of Hunan Children's Hospital approved this study as a case report for retrospective analysis. The ethical approval number was HCHLL-2018-68. Informed written consent was obtained from the patient for publication of this case report and accompanying images. We anonymized all information before analysis.

2. Case report

A 10-year-old boy with complex congenital heart disease presented with hemodynamically poorly tolerated refractory paroxysmal supraventricular tachycardia. The heart presented a single, leftsided ventricle due to severe tricuspid hypoplasia, associated with a large secundum atrial septal defect and ventricular septal defect with right ventricular outflow tract stenosis. The patient had already undergone a bidirectional Glenn procedure at age 3.

Over the last 3 months, he had multiple hospitalizations for narrow-QRS tachycardia with poor hemodynamic tolerance, despite the use of adenosine and amiodarone. When arrhythmia-free, he remained functionally stable in New York Heart Association functional class II–III/IV. Physical examination revealed cyanosis and clubbing, with a baseline oxygen saturation of 85%. Electrocardiogram showed normal sinus rhythm at 90 beats/min, no preexcitation, biatrial enlargement. The patient was pending the Fontan operation, and a preliminary electrophysiological study was requested.

The patient was brought to the cardiac electrophysiology laboratory in the fasting and unsedated state. Routine monitoring equipment including electrocardiogram and defibrillation pads was placed. With the child under intubation and general anesthesia, access was obtained in both femoral veins and the right femoral artery. Angiocardiography showed a secundum
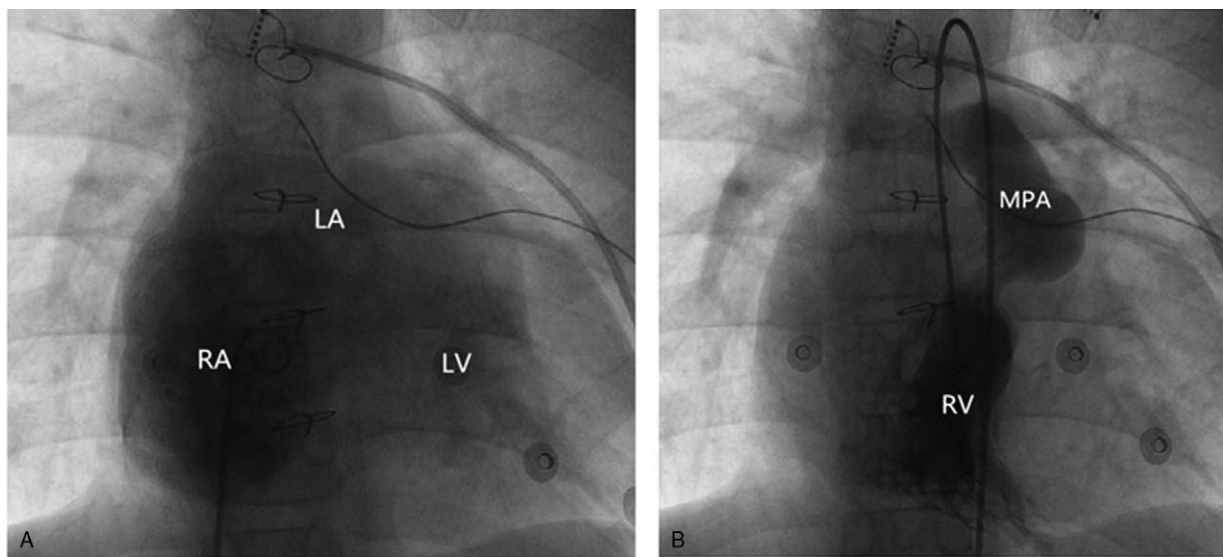
Figure 1. Angiocardiography showed anatomy of the heart in anteroposterior views. A, Power contrast injection via a 5-French pigtail catheter in the right atrium (RA) showed the absence of a right-sided AV valve, a secundum atrial septal defect, and a patent left-sided AV valve with left ventricular (LV) filling. B, A 5-French pigtail catheter was located in right ventricular (RV) through the ventricular septal defect. Angiography showed severe stenosis in right ventricular outflow tract. LA = left atrium, MPA = main pulmonary artery.

atrial septal defect and large ventricular septal defect with right ventricular outflow tract stenosis (Fig. 1). The connection between superior vena cava and right pulmonary artery was unobstructed. The pressure and oxygen saturation values of different chambers were taken during cardiac catheterization.

A 6-French 4-polar electrode catheter and a 4-mm ablation catheter were introduced through the right femoral vein into the left ventricle and right interatrial septum. Ventricular extrastimulus testing from the left ventricular (LV) demonstrated nondecremental ventricular-atrial conduction with retrograde refractory period of 280 ms. Atrial extrastimulus testing showed decremental atrial-ventricular (AV) conduction and induced a supraventricular tachycardia with a cycle length of 390 ms (Fig. 2A). A His-bundle electrogram was recorded from right
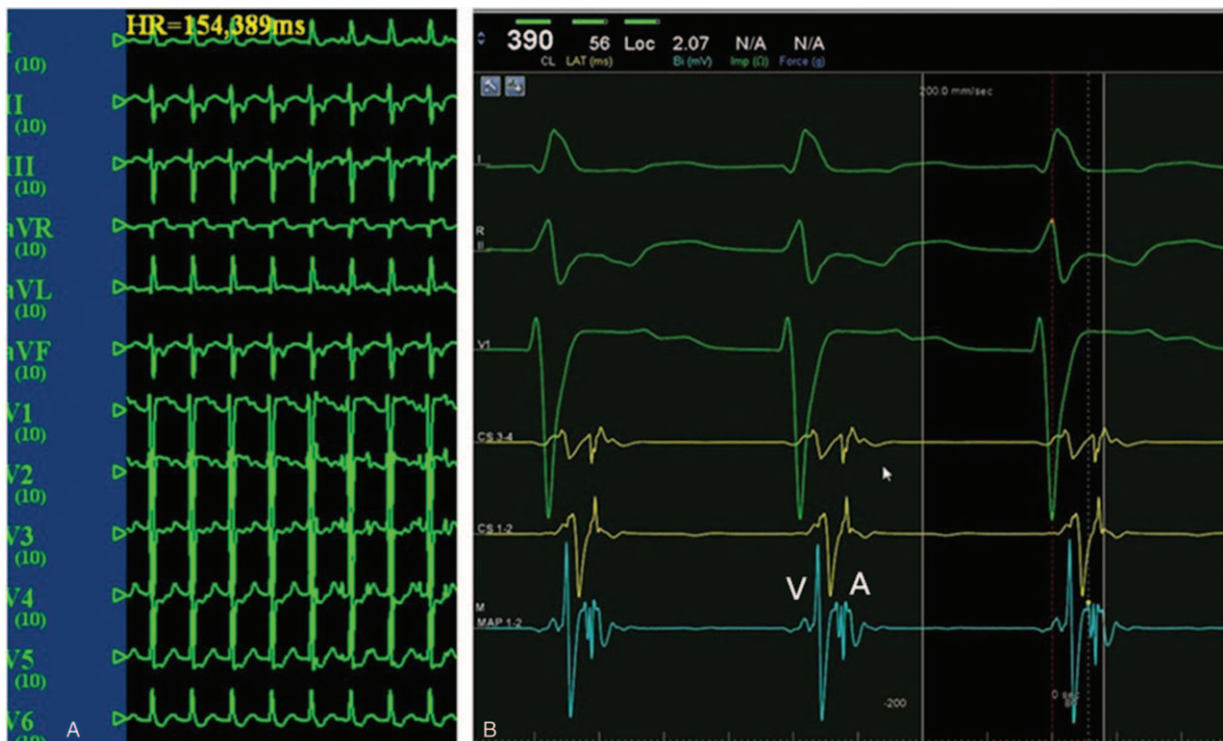

Figure 2. A, Electrocardiogram of clinical tachycardia with a cycle length of 389 ms. B, The site of earliest atrial activation was localized in the coronary sinus, which was recorded by the ablation catheter, between electrodes 1-2 and 3-4.
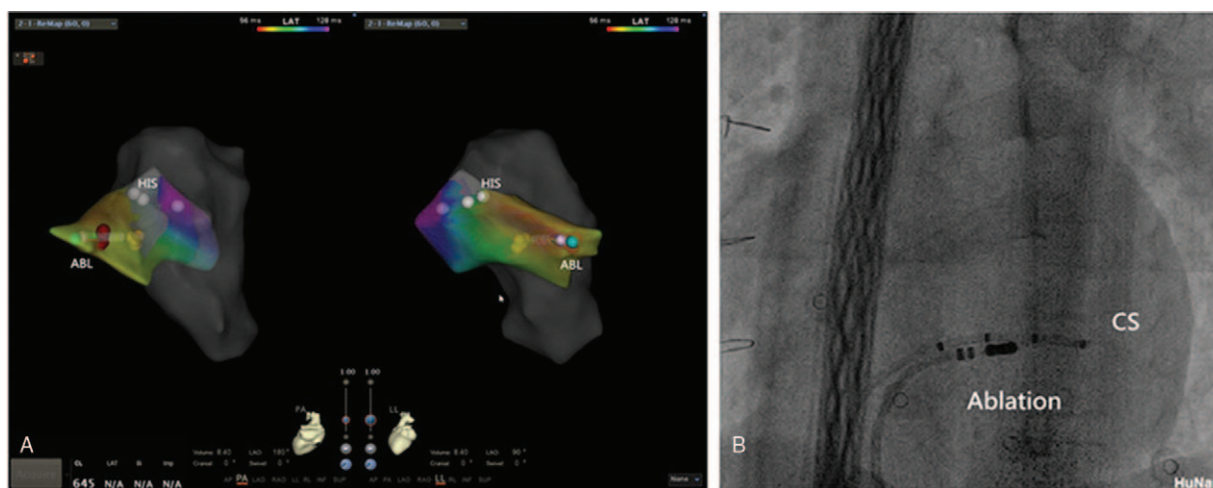
Figure 3. Location of the His-bundle electrogram and site of ablation. A, Electroanatomic maps of right atrial during tachycardia. Red circles and the catheter tip indicate the location of ablation lesions (ABL). White circles represent sites where His-bundle electrograms were recorded (HIS). B, Site of ablation, in the coronary sinus (CS) between CS1-2 and CS3-4 in left anterior oblique view.

interatrial septum, superior to the coronary sinus ostium with the ablation catheter. The tachycardia was pace-terminable, with anterograde atrioventricular block.

Electroanatomic mapping of right atria was performed when the patient was in tachycardia. The site of earliest atrial activation was localized in the coronary sinus, between electrode 1-2 and 3-4 (Figs. 2B, 3). In sinus rhythm, radiofrequency ablation was attempted at this point and a temperature controlled ablation system with a maximum temperature of 60°C and power output of 30 W for 120 seconds. In the process of ablation, junctional acceleration did not occur. Postablation, the tachycardia cannot be induced by atrial extrastimulus testing and overdrive pacing. Ventricular extrastimulus testing from the LV demonstrated decremental ventricular-atrial conduction with retrograde refractory period of 400 ms. These findings suggested that the most likely diagnosis was posteroseptal accessory pathway-mediated tachycardia. The child has not had a recurrent event during 6 months of follow-up and is waiting for Fontan operation.

3. Discussion

Children with complex congenital heart disease are at risk of developing arrhythmias as a result of cardiac defects, the hemodynamic stress imposed by such malformations, or postoperative sequelae following cardiac surgery.[3] Some congenital heart malformations are associated with an abnormal architecture of the conduction system. Apart from arrhythmias caused by anatomic malformations and/or scar-related arrhythmias due to surgical corrections, all known forms of supraventricular tachycardia can occur in patients with tricuspid atresia.[4] Recurrent tachycardia can cause cardiac insufficiency and increase the risk of sudden death.[5] Therefore, a detailed electrophysiological examination is necessary to determine the mechanism of arrhythmia and risk of ablation. We completed cardiac catheterization, pulmonary artery pressure assessment, and electrophysiological examination at the same time, which can provide an effective support for Fontan operation.

As in previous studies, congenital accessory pathways in patients with tricuspid atresia are mostly on the right side,[6,7] which is the same in our case. This confirms the theory of Misaki et al[7] that the accessory pathway is on the side of the congenitally malformed atrioventricular valve, although this theory is based mainly on patients with Ebstein malformation and only on a very few number of other congenital atrioventricular valve defects. Children with tricuspid atresia usually need staged surgical correction. Arrhythmia may occur before and after surgical treatment, especially after Fontan operation. Due to the presence of atrial baffle and changes of atrioventricular connection, it is very difficult to treat the arrhythmia and even lose opportunities for ablation.[8]

Since there is an increased risk of accessory pathways in patients with tricuspid atresia,[2,9] all these patients should be checked before the Fontan operation to exclude congenital accessory pathways. If an accessory pathway is detected, it should be localized and ablated either in the same session or during surgical intervention, even if the patient is asymptomatic.[6] The application of three-dimensional ablation system has greatly improved the success rate and safety of ablation in children with complex congenital heart disease.[10] Radiofrequency ablation is possible, even in infants with a similar success rate compared with that seen in older children.

Author contributions

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Supervision: Qiming Liu, Zhi Chen.
Writing – original draft: Yefeng Wang.
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