Radiofrequency catheter ablation in a child with Wolff-Parkinson-White syndrome and congenitally corrected transposition of the great arteries

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Abstract
The case is presented of a five-year-old boy with Wolff-Parkinson-White syndrome, who had undergone three surgical palliations because of congenitally corrected transposition of the great arteries, ventricular septal defect and pulmonary trunk near-atriesia. Successful and uncomplicated radiofrequency catheter ablation of an accessory pathway located across the systemic atrioventricular valve ring was performed. The procedure was motivated by forthcoming corrective surgery, which would preclude venous access to the heart. This case emphasizes the need for precise timing of ablative therapy in such patients and shows that surgery itself may facilitate catheter ablation. (Cardiol J 2007; 14: 500–503)

Key words: congenital cardiac malformation, ventricular pre-excitation, orthodromic atrioventricular re-entrant tachycardia, catheter ablation

Introduction
It is more than a decade since radiofrequency catheter ablation became the most efficient treatment for many tachyarrhythmias in both adults and children, irrespective of the presence or absence of structural heart disease [1–6]. Owing to the natural course of most of the common tachycardias in children, it is not uncommon to wait until the child has reached the tenth year of age before undertaking ablative treatment [2]. The ablation may be performed at an earlier age when the tachycardia is highly symptomatic or when surgery for congenital heart disease is anticipated, which would make catheter ablation difficult or impossible [2].

Case report
The case reported is that of a boy aged 4 years and 11 months with a body weight of 22.5 kg and a height of 114 cm at the time of electrophysiological study. At birth he had been diagnosed with congenitally corrected transposition of the great arteries, large non-restrictive inlet-outlet ventricular septal defect, near-atriesia of the pulmonary artery and persistent arterial duct, with oxygen saturation of the capillary blood (SatO2) of 55% and no signs of heart failure. Treatment with prostaglandin E2 was started to prevent arterial duct closure. At the age of 5 days a central aortopulmonary anastomosis was created, with interposition of a vascular prosthesis between the ascending aorta and the right pulmonary artery, resulting in an immediate increase of SatO2 to 74%.

By the age of 20 months the effect of the surgery had become negligible. The SatO2 had decreased to 64% and the anastomosis was found to be non-functional. A second surgical palliation was performed at the age of 21.5 months. This consisted of modified Blalock anastomosis with interposition
of a vascular prosthesis between the left subclavian artery and the left pulmonary artery, resulting in an immediate increase of SatO2 to 88%.

The first stage of a third surgical operation was undertaken just before the boy reached 4 years of age. A bi-directional cavopulmonary Glenn anastomosis was created between the superior caval vein and the right pulmonary artery with the stump of the superior caval vein sewn. In addition, an atrioseptectomy was performed with creation of an interatrial communication measuring 12 mm in diameter. The Blalock anastomosis was clipped.

It was decided to proceed further to total cavopulmonary connection, with anastomosis of the inferior caval vein to the right pulmonary artery at a later stage.

Meanwhile, at the age of 3 months the boy had, for the first time, experienced narrow QRS tachycardia with a rate of 280 bpm. During the tachycardia the child had shown signs of heart failure and increase of the cyanosis. The tachycardia was terminated with adenosine, and then amiodarone treatment was started for one month. At the age of 9 months the tachycardia relapsed again and after its termination ventricular pre-excitation was registered for the first time. After this chronic amiodarone therapy had been reinstituted. Nevertheless, he had had a third paroxysm of the same tachycardia.

With a view to the forthcoming total cavopulmonary connection, which would preclude venous access to the heart and the performance of electrophysiologic study, it was decided to make the electrophysiologic study and radiofrequency catheter ablation before the second stage of the surgical operation.

The procedure was performed under general anesthesia. The catheters passed with some difficulty along the iliac veins, although during contrast injection both vessels appeared to be entirely normal, large enough and with a straight course. Through the femoral veins were inserted a 4-polar 5 F catheter with a Damato curve (Viking, CR Bard Inc., Lowell, MA, USA) in the venous ventricle (morphologically the left ventricle) and a 4-polar 5 F catheter with a Josephson curve (Viking, CR Bard, Lowell, MA, USA) in the anteroseptal area of the normally located right atrium. After several unsuccessful attempts at catheterizing the coronary sinus with a 4-polar 6 F steerable catheter (Polaris DX, Boston Scientific Corp., Natick, MA, USA), the catheter was inserted through the surgically created interatrial communication into the normally located left atrium close to the lateral part of the ring of the systemic atrioventricular valve (morphologically the tricuspid valve).

During positioning of the catheters narrow QRS tachycardia was repeatedly induced with a rate of 230 bpm. Sustained atrial flutter was also induced. The latter could not be terminated with pacing maneuvers, and transthoracic cardioversion was performed with restoration of sinus rhythm.

The narrow QRS tachycardia was found to be an orthodromic atrioventricular re-entrant tachycardia with the accessory pathway participating in the re-entrant circle. The earliest retrograde activation was recorded at the lateral part of the left atrium (Fig. 1A). The shortest delta-V interval in sinus rhythm was recorded in the same area (Fig. 1B).

Using the femoral venous approach a 5 F 4-mm tipped ablation catheter (Marin Sc, Medtronic Inc., Minneapolis, MN, USA) was inserted through the...
interatrial communication into the left atrium for mapping and ablation. The shortest local ventriculoatrial interval during tachycardia and the shortest local atrioventricular interval during sinus rhythm were found at the anterior part of the systemic atrioventricular valve annulus (Fig. 2A). Five radiofrequency applications were delivered in this area. Three of these led to immediate termination of the tachycardia or to immediate disappearance of the delta wave, but shortly after each application the pre-excitation pattern recurred. The atrioventricular valve ring was remapped with the ablation catheter. A second area was found, located approximately 1 cm more laterally, with a short ventriculoatrial interval during tachycardia (Fig. 2B). Two more radiofrequency applications delivered to this area during sinus rhythm led to permanent loss of pre-excitation (Fig. 3) without the ability to reinduce the tachycardia. The retrograde conduction became decremental and non-eccentric.

Figure 2. Fluoroscopic images at a left anterior oblique incidence of 45°, showing two different sites where the pre-excitation was eliminated. Abl — ablation catheter; HB — His bundle catheter; LA — left atrial catheter; LV — venous ventricle.

Figure 3. Twelve-lead surface ECG during the last radiofrequency application, showing the sudden loss of pre-excitation.
An adenosine bolus applied intravenously caused transient advanced atrioventricular block. The procedure and post-procedural course were uneventful and the boy was discharged from the hospital on the next day.

**Discussion**

Radiofrequency catheter ablation has been performed in children for more than a decade now. In the case of spontaneous or drug-induced remission of arrhythmia episodes ablation can be postponed until a later age, but in patients with congenital heart malformations supraventricular re-entrant tachycardias may lead to severe hemodynamic compromise and impose an early attempt at ablating the substrate [2]. Another reason for an early ablative approach is anticipated heart surgery that would make subsequent ablation difficult to perform [2]. In the case presented this was one of the reasons for proceeding with ablation, given the fact that the tachycardia was satisfactorily controlled by amiodarone.

In this case, after Glenn anastomosis was made, venous access to the heart could be effectuated only through the femoral veins. This could be the reason for the impossibility of catheterizing the coronary sinus. On the other hand, the surgically created interatrial communication made the procedure technically not very demanding, permitting access of diagnostic and ablation catheters to the left atrium and thus sparing the need for a trans-septal approach or femoral artery puncture and a retrograde approach to the systemic atrioventricular valve ring. Although in this type of congenital malformation the His bundle is not uncommonly displaced and susceptible to mechanical trauma, it was possible to locate it at quite a typical position and to avoid mechanically induced atrioventricular block [7]. Regardless of the presence of an interatrial communication, His bundle recordings were stable and reproducible throughout the study.

The pre-excitation pattern could be eliminated permanently and the tachycardia terminated by radiofrequency applications at two different sites separated by a distance of approximately 1 cm. One may speculate that two accessory pathways were present with similar conduction properties and located very close to each other. Such a coincidence might explain why only one pre-excitation pattern was observed and why the tachycardia characteristics were constant throughout the study. However, a discrete shift in pre-excitation and a subtle change in tachycardia characteristics at least would be expected to occur. This was not observed, which makes such an explanation highly improbable.

In conclusion, this case underscores the need for careful and precise timing of ablative treatment in children with congenital cardiac malformations, in whom totally corrective surgery is envisaged. It also shows that surgery itself, at least at some stage, may actually facilitate catheter ablation in children with congenitally corrected transposition of the great arteries.

**References**