Catheter ablation of multiple, surgically created, atrioventricular connections following Fontan–Björk procedure

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It is well known that after a Fontan procedure, patients are prone to supraventricular arrhythmias.1–3 In addition, the association between tricuspid atresia and Wolff–Parkinson–White (WPW) syndrome has previously been described.4–6 Furthermore, several cases have been documented of orthodromic re-entrant tachycardia due to surgically acquired accessory pathways (APs), following right atrial (RA) to right ventricular (RV) anastomosis, after a Fontan–Björk-type connection.6–11 However, to the best of our knowledge, the development of multiple acquired AP has only been reported once. We describe the case of a patient who developed WPW syndrome due to multiple atrioventricular (AV) connections following Fontan operation.

Case report

A 24-year-old woman with WPW syndrome was admitted to our unit for electrophysiological study and radiofrequency (RF) ablation. At birth, she was diagnosed of tricuspid atresia and Wolff–Parkinson–White (WPW) syndrome has previously been described.4–6 However, to the best of our knowledge, the development of multiple acquired AP has only been reported once. We describe the case of a patient who developed WPW syndrome due to multiple atrioventricular (AV) connections following Fontan operation.

After informed consent was obtained, an electrophysiological study was performed. An ablation catheter and three quadripolar diagnostic catheters were inserted through the right femoral vein and artery and placed, under fluoroscopic guidance, at the RA, His bundle area, and RVOT. A His-bundle electrogram was recorded at the left side of the septum. Baseline HV interval was 8 ms. From time to time, we observed a second pattern of pre-excitation (Figure 2A). The patient remained asymptomatic for many years, but at the age of 18, she began experiencing brief episodes of palpitations. Holter monitoring documented persistent pre-excitation and several runs of non-sustained narrow-QRS complex tachycardia. An echocardiogram showed marked dilatation of the RA and the RV outflow tract (RVOT) with a wide, non-obstructive connection between the two chambers. The left ventricle (LV) showed normal dimensions with hypokinesia, atrophic movement of the interventricular septum, and mild LV dysfunction (LV ejection fraction: 43%).

An increased incidence of Wolff–Parkinson–White (WPW) syndrome with tricuspid atresia has been reported. Although atrioventricular accessory pathways may develop across suture lines after the Fontan–Björk procedure, the presence of multiple acquired accessory pathways has only been described rarely. We report on a case of a female with tricuspid atresia who underwent the Fontan operation at 5 years of age. One year later, she developed a WPW pattern. Narrow complex tachycardias started at the age of 18. An electrophysiological study revealed the presence of three accessory pathway connections at the surgical anastomosis level. All of them were successfully ablated and there were no recurrences.
reproducibly induced by atrial stimulation. The VA interval during tachycardia was 95 ms. Activation mapping during tachycardia was performed with a 4 mm deflectable-tip electrode ablation catheter, first around the atrial aspect of the atretic tricuspid annulus with late activation times. Mapping of the RA to RV anastomosis showed the shortest ventriculoatrial conduction times on the mid-portion of the anastomosis (Figures 3A and 4). No other tachycardias were induced at the electrophysiological (EP) study.

Subsequently, we proceeded to map during sinus rhythm the RA–RVOT anastomosis and found the earliest activation times (20 ms before the onset of the delta wave) at its inferior portion. RF application at this site resulted in a transient change in the pattern of pre-excitation, but AP conduction recovered shortly afterwards. We mapped the atrial insertion of the AP during ventricular pacing, and retrograde continuous activity was found 1 cm above the previous application (Figures 3A and 4). A single 60 s RF application blocked the VA conduction, confirmed by the presence of VA dissociation (Figure 3A), but with persistent anterograde pre-excitation, suggestive of a concealed AP. Another RF application was delivered during sinus rhythm close to the first application and the predominant pre-excitation pattern disappeared (Figures 2A and 4). Then, we determined an effective refractory period of the second manifest AP of 310 ms. Finally, activation mapping of this AP was performed during sinus rhythm, and a point with early activation time was found in the most superior portion of the RA–RVOT anastomosis (Figure 4) where an AP potential was recorded (Figure 3B). RF application at this site permanently abolished AP conduction and the pre-excitation pattern (Figure 2B). The patient is now asymptomatic and without recurrences of palpitations or pre-excitation after a 2 year follow-up. An echocardiogram performed 6 months after ablation showed normalization of the mitral valve function.
of the asynchronic movement of the interventricular septum and the LV ejection fraction.

Discussion

Since the technique for the treatment of tricuspid atresia was first described by Fontan and Baudet in 1971, there have been several modifications of this procedure. Despite these, patients are prone to supraventricular arrhythmias after a Fontan procedure.1–3 In addition, the WPW syndrome and congenital heart diseases, such as Ebstein’s anomaly and tricuspid atresia, are well-known associations.4–6 In many cases, they had been treated in the same surgical procedure.5 Sometimes APs are detected on the basis of surface ECG criteria only after the Fontan operation. This could be due to the presence of previously unapparent congenital AP. Changes in the conduction properties in the atrial myocardium after surgical intervention and surgical injury to the AV node could slow conduction, allowing a previously unapparent AP to become manifest. However, surgically created accessory connections at the atrioinfundibular anastomosis are another cause of WPW syndrome after the Fontan–Björk procedure. This possibility must be considered in these patients, and the surgical anastomosis must be carefully mapped when searching for an AP during ablation procedures.

In 1992, Razzouk et al.7 described the finding of an AP through the RA–RV anastomosis in a patient with supraventricular tachycardias following a Fontan operation. Since then, there have been several case reports of AP surgically created after the Fontan procedure with a Björk-type connection and orthodromic AV tachycardia.5–11 However, in previously reported cases, only a connection through the RA–RV anastomosis has been found and the presence of multiple AV connections has been rarely reported. Hager et al.6 recently reported on the results of EP studies in five patients with WPW syndrome tricuspid atresia treated by Fontan–Björk procedure and haemodynamically symptomatic AV re-entrant tachycardia. Two of these five patients presented acquired WPW syndrome after the Fontan procedure. In one patient, three different APs were located at the atrioinfundibular Fontan anastomosis in superior, left lateral, and inferior positions and were successfully treated with RF ablation.

It can be argued that our findings could be related to a broad connection rather than to separate connections. However, the distance between the RF applications in our case and in that of Hager et al. favour the presence of different APs. The growth of myocardial cells across the suture line, or the presence of electrotonic transmission through this line, is the main explanation for this finding. The existence of multiple AP connections suggests that the growth of ‘de novo’ excitable tissue is the most probable cause of WPW syndrome development in this type of procedure. There is further evidence that supports the growth of tissue across suture lines. Atrioatrial conduction has been described after orthotopic heart transplantation.12,13 Furthermore, the description of WPW syndrome years after heart transplantation is another example of ‘acquired bypass tract’.14

From a clinical point of view, early treatment of these APs is mandatory because AV re-entrant tachycardia can deteriorate into atrial flutter or fibrillation that can be conducted 1:1 to the ventricle. Furthermore, anti-arrhythmic drug treatment often fails in these patients. Another interesting observation in our case was the normalization of the LV ejection fraction in the echocardiogram performed several months after the ablation procedure. The presence of asynchronic contraction of the interventricular septum due to the presence of the AP, resolved after RF ablation, could explain this finding.

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References