Case Report

Iatrogenic atrioventricular reentrant tachycardia following Bjork/Fontan palliation of tricuspid atresia: Electro-anatomic mapping, ablation, review and possible mechanism

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S U M M A R Y

This case report describes the successful ablation of an iatrogenic accessory pathway in a Fontan patient. A 15-year-old girl with tricuspid atresia was palliated with a Bjork modification Fontan procedure; six years later she developed supraventricular tachycardia. Electro-anatomic mapping during electrophysiology study localized a concealed iatrogenic atrioventricular accessory pathway and facilitated successful ablation using radiofrequency energy. This report reviews the reported cases of anomalous atrioventricular conduction (antegrade only, retrograde only [Hager, et al., J Thorac Cardiovasc Surg 2005;130:48–53], both [Liberman, et al., Pacing Clin Electrophysiol 2000;23:914–6]) after the Fontan/Bjork repair, outlines a potential pathophysiologic mechanism for the post-operative tachyarrhythmia and highlights the usefulness of electro-anatomic mapping in identifying unusual arrhythmias in post-operative patients with complex congenital heart disease.

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Introduction

Recent advances in management of cardiac arrhythmias include the development of non-radiologic mapping techniques to identify and localize the mechanism of tachyarrhythmias, particularly in the young person with repaired complex congenital heart disease. In this report we describe a 15-year-old girl with tricuspid atresia 6 years after the Bjork modification Fontan operation who developed a paroxysmal tachyarrhythmia and review the reported 7 cases [1–6]. Electro-anatomic mapping played a key role in identifying the mechanism and facilitating its successful elimination by transcatheter radiofrequency ablation [1]. Finally, we update previously noted possible pathophysiologic mechanisms for the tachycardia [2,7–9].

Case report

The patient underwent a Bjork modification of the Fontan procedure [10] at 3 years of age consisting of suturing the right atrial appendage to the anterior right ventricular outflow tract via a ventriculotomy. There was no ventricular preexcitation and no tachycardia prior to her Fontan palliation. At 9 years of age she presented with a paroxysmal narrow QRS complex tachycardia at 280 beats/min; the tachycardia was successfully cardioverted to sinus rhythm on several occasions with adenosine; she was treated with digoxin and verapamil. At 15 years of age, because of recurrent tachycardia, she underwent electrophysiologic (EP) study using a multi-channel electrophysiologic recording and pacing system (EP Med System, West Berlin, NJ, USA). A 4F electrode catheter was placed in the left ventricle and a 2F active fixation temporary pacing lead (Medtronic Inc, Minneapolis, MN, USA) in the high right atrium. No His potentials were recorded. Adenosine (10 mg), given by bolus injection during left ventricular pacing, demonstrated, constant retrograde conduction consistent with a concealed accessory pathway. Ventricular burst and extrastimulus pacing revealed a constant ventricular-atrial (VA) conduction interval. Atrial extra stimulus induced supraventricular tachycardia (SVT; cycle length = 250 ms) with a 1:1 atrioventricular relationship. Premature ventricular extra stimuli during the SVT advanced atrial activation during probable His refractoriness (based on estimated HV interval). Using the CARTO electro-anatomic mapping system (Biosense, Webster, Diamond Bar, CA, USA), the location of the atretic tricuspid valve was identified in the area of tricuspid valve. VA conduction intervals mapped and measured on the atretic tricuspid valve annulus (onset of the QRS complex to the first rapid deflection of the atrial electrogram) during SVT were 137 ms, 156 ms, 118 ms, and 140 ms at 3:00, 6:00, 9:00, and 12:00 respectively. The shortest VA

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interval during SVT was 71 ms and was measured at the anterior right atrium, near the mouth of the right atrial appendage (RAA) where it was anastomosed to the right ventricular outflow tract (RVOT, Fig. 1). Radiofrequency energy (maximum temperature 51 °C) at 50 Watts for 30 s delivered at that site eliminated VA conduction (Fig. 2), two additional lesions were applied due to the initial low temperature. Post ablation EP testing with and without isoproterenol demonstrated decremental retrograde conduction and failed to induce SVT. Intravenous adenosine given during left ventricular pacing resulted in retrograde block. The patient was discharged 6 h after the procedure; she has been arrhythmia free for 5 years.

**Discussion**

Atrial arrhythmias, especially intra-atrial reentrant tachycardia (IART), are common following the Fontan operation, occurring in 7–35% of patients [11–13]. In contrast to the expected IART in the tricuspid atresia patient following a Fontan operation, an atrioventricular reentrant tachycardia (AVRT) supported by an accessory pathway (either manifest or concealed) is unusual [1,14,15], but not infrequently suggested by the electrocardiogram. On the other hand, the atrioventricular anastomosis Fontan (Bjork modification [10]) has been reported to produce an inadvertent, surgically created, electrically active connection supporting atrioventricular reentry tachycardia in 7 previous cases [1–6]. In addition 4 of these 7 patients had both antegrade (preexcitation, Wolff-Parkinson White electrocardiogram pattern) and retrograde conduction (AVRT) through the atrioventricular anastomosis [1,4,5]. One additional patient developed only antegrade conduction after surgery [16]. In our patient, electro-anatomic mapping was used in the anticipation of an inducible IART. Six previous reports [1–6] described ablation (6 by radiofrequency ablation, 1 by surgery) of a concealed only [1] or manifest accessory pathway [2] (two multiple) in these 7 patients. The authors
interpreted the findings differently. Based on the angiographic images of the RA-RV anastomosis and the catheter positions superimposed on those images, one postulated a pre-existing congenital concealed accessory pathway in the region of the tricuspid valve [3] as apparently occurred in 3 of Hager’s 5 patients [1], while the others hypothesized the development of an iatrogenic accessory pathway mediating AVRT following the Fontan palliation [1,2,5,6].

The surgical creation of an electrically conducting atrioventricular connection is of considerable interest and not dissimilar to the observed atrio-atrial electrical connectivity that has been reported in patients following cardiac transplantation [17–19]. As suggested by others [2,17,19] and based on more recent experimental observations [7–9,19,20], we propose that electrotonic transmission may account for impulse propagation through a narrow area of impaired conduction, which may exist in an anastomotic “scar,” with just enough ionic channels and electrotonic current spread across myocyte-myofibroblast connections in the absence of an action potential generation – to support downstream transmission [7,9,19,20]. Four patients with tachycardia supported by the new anastomosis [1] demonstrated antegrade anomalous conduction across the atrioventricular anastomosis. Four patients (including the present case) exhibited only retrograde conduction across the anastomosis, perhaps related to asymmetry in the geometry of the conduction pathway [8] or the greater amplitude of the ventricular signal (compared to the atrial signal) sufficient to recruit more ionic channels and produce more electrotonic current to excite the atria retrogradely. Other authors have suggested that some fibers from the upstream tissue may become “trapped” within and along the border of the downstream insertion tissue or grow into it and establish conduction [18,19].

Several factors support the interpretation that the post-surgical AVRT in our patient also arose from an iatrogenic, post-surgical formation of a ventricular-atrial conducting pathway between atrial and ventricular tissue. First, careful electro-anatomic mapping disclosed a long VA conduction interval during the tachycardia at the tricuspid valve ring and coronary sinus compared to the short VA conduction interval recorded at the site of successful ablation – the RAA–RVOT anastomosis. Second, the tachycardia in all 8 patients did not occur until after the Fontan operation, allowing for the high probability that a pathway did not exist prior to

Figure 2. Electrocardiogram and intracardiac electrogams obtained during ventricular pacing and radiofrequency ablation. A loss of retrograde conduction is seen 2.2 s into the radiofrequency application.

the Fontan operation. Third, the interruption of VA conduction and tachycardia after ablation supports the observation that there was no residual accessory conducting pathway near the tricuspid valve site and that pre-ablation retrograde conduction occurred only at a site, identified by electro-anatomic mapping, anatomically distant from the tricuspid valve but anatomically immediate to the RAA–RVOT anastomosis.

Conclusion

Electro-anatomic mapping is a highly effective adjunct in the diagnosis and treatment of IART in the post-operative congenital heart disease patient. In this case, as in a previous case [1] electro-anatomic mapping was very useful in confirming and then precisely localizing the concealed iatrogenic retrograde limb of the reentrant circuit. Based on recent experimental observations, we propose that sufficient electrotonic current through myocyte-myofibroblast connection established by the atrio-ventricular anastomosis is the likely mechanism that supports the acquired anomalous conduction and the AVRT. This experience, along with the review of the previous cases, underscores the importance of establishing the mechanism of the tachycardia, as well as the value of high definition electro-anatomic mapping, when evaluating a patient with arrhythmias following cardiac surgery for complex congenital heart disease.

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References


