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TRANS-CONDUIT PUNCTURE FOR CATHETER ABLATION OF ATRIAL TACHYCARDIA IN A PATIENT WITH EXTRA-CARDIAC FONTAN PALLIATION

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catheter ablation; Fontan surgery; conduit puncture; electrocautery

INTRODUCTION

The Fontan palliation and its variants are the most widely used and accepted approach to regulate pulmonary blood flow and relieve cyanosis in patients with single ventricle physiology¹. First generation Fontan procedures involved connection of the right atrium (RA) to the pulmonary artery (PA). However, this frequently caused RA dilation, sluggish RA flow, atrial fibrosis, and was associated with a high incidence of atrial arrhythmias^{2,3}. Exclusion of most or all of the RA decreases the incidence of atrial arrhythmias; therefore, the Fontan procedure evolved by establishing a total cavopulmonary connection (TCPC) by way of either the lateral tunnel (an intra-atrial tunnel) or the extra-cardiac Fontan^{4,5}. Patients who underwent first generation RA-PA Fontan surgery with subsequent development of atrial arrhythmias may benefit from surgical conversion to TCPC with concomitant arrhythmia surgery^{6,7}. Despite significant reduction in arrhythmia burden, a sub-group of patients still develop recurrent arrhythmias⁸. The extra-cardiac Fontan consists of an IVC to PA conduit and direct connection of the SVC to the PA. Being completely outside the heart, it is thought to have less arrhythmic sequelae⁹; however, the conduit being made of stiff Gore-Tex, it also precludes access into the heart. This report outlines an approach to catheter ablation in a patient with extra-cardiac Fontan palliation where the heart was accessed via the conduit.

CASE REPORT

Our patient is a 34 year old male with functionally single (left) ventricle, with tricuspid atresia but not pulmonary stenosis/atresia, and with atrial septal defect (ASD), ventricular septal defect

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(VSD), and D-transposition of the great arteries in situs solitus (S,D,D). He underwent PA banding at age 8 months. At age 7, this band was loosened and his VSD was enlarged. At age 15, he underwent non-valved modified Fontan anastomosis of RA to PA with pericardial augmentation, ASD closure, and further enlargement of the VSD. A few years later, he began having palpitations: these were about once weekly, lasted up to 30 seconds, were associated with dyspnea, but without syncope or presyncope. At age 30, in 2005, for reduced exercise tolerance he underwent revision to an extra-cardiac non-fenestrated Fontan, a bidirectional Glenn, and resection of subaortic stenosis (Figure 1). This surgery involved resection of the atrial septum, resulting in a single pulmonary venous atrium with 'right' and 'left' atrial portions. Concomitant arrhythmia surgery was not performed. Subsequently, his exercise tolerance improved significantly. His atrial arrhythmias were treated successfully with amiodarone following surgery. In 2008, this was stopped due to elevated liver enzymes, and he was switched to dofetilide which unfortunately was ineffective. His palpitations progressively increased to several times a week, lasting minutes. They were sudden in onset and offset and were associated with significant dyspnea. He continued to be free of syncope. Holter monitoring showed predominantly atrial tachycardia and rare episodes of atrial fibrillation. In April, 2009 he underwent cardiac catheterization, plug occlusion of a large venous collateral, and electrophysiologic testing that showed an atrial tachycardia with 472ms cycle length, easily inducible with burst atrial pacing at the IVC/conduit junction at 400ms cycle length. Cardiac MRI showed close apposition of the extracardiac conduit and the free wall of the right atrial portion of the pulmonary venous atrium (RA).

He was referred for catheter ablation. Conduit angiography was performed (Figure 2A). Intravenous heparin was administered to achieve an ACT over 300s and was given prior to trans-conduit puncture to minimize risk of clot formation (standard for all transseptal access procedures in our lab). Protamine was kept ready in case of perforation or significant bleeding. The tip of an SL-0 sheath with a 71cm Brockenbrough (BRK-1) needle (St. Jude Medical, Minnetonka, MN) inside of it was positioned along the left border of the conduit adjacent to the RA. Correct positioning was confirmed with biplane fluoroscopy; transesophageal echocardiography and intracardiac echocardiography were less useful for this step. The needle was advanced but would not cross readily through the Gore-tex conduit material. Multiple advances were made with radiofrequency energy (Valley Lab, Boulder, CO) at 45W and 90W of energy using the "cut" setting. A second needle was required when the first needle was found to have been bent. Using a combination of constant forward pressure and radiofrequency energy, the BRK-1 needle was able to cross the conduit, and contrast injection showed staining of the conduit and atrial myocardium (Figure 2B). A 0.032" Toray valvuloplasty wire (Toray Corporation, Japan) was advanced through the SL-0 sheath into the atrium. The dilator crossed into the atrium over the Toray wire, but the sheath would not readily cross (Figure 2C). The sheath and dilator were exchanged for an Agilis 8.5 Fr sheath (St Jude Medical, Minnetonka, MN). Again the dilator crossed but the sheath would not track over the dilator. With the Toray wire and no dilator in place, the hemodynamics were stable and no effusion / hemothorax was noted on transesophageal echocardiography. A 4 mm × 20mm peripheral percutaneous transluminal angioplasty balloon (OptaPro, Cordis Endovascular, USA) was advanced over the Toray wire and positioned to straddle the border between the extracardiac conduit and the atrium (Figure 2D). Two inflations were performed at 6 atmospheres and 8 atmospheres for 10 seconds each. Following balloon dilation, the SL-0 sheath was able to cross readily. Atrial position was confirmed with contrast injection. Transesophageal echocardiography and intracardiac echocardiography showed no evidence of bleeding. The Berman angiographic balloon-tipped catheter (Arrow International, Reading, PA) was advanced through the SL-0 sheath into the atrium, and cine-angiography was performed (Figure 2E).

A deflectable tip duodecapolar catheter was positioned across the aortic valve into the (left) ventricle. A 3.5mm tip externally irrigated radiofrequency ablation catheter (Celsius

Thermocool, Biosense Webster, Diamond Bar, CA) was positioned in the atrium and electroanatomic mapping was performed (Figure 3) using the Ensite system (St Jude Medical, Minnetonka, MN). Scar was noted in the anterior peri-mitral valvular region. The clinical atrial tachycardia was induced and entrainment mapping confirmed the scar border zone to be part of the circuit. Ablation was performed along this scar using 30 to 50W of radiofrequency energy with 30cc/minute of irrigation, keeping tip temperatures limited to below 42°C. Following ablation, the targeted (clinical) tachycardia was non-inducible with pacing maneuvers and also with isoproterenol infusion. Two different rapid narrow complex tachycardias (cycle lengths 377ms and 300ms) were induced with isoproterenol infusion but were poorly tolerated hemodynamically and required pace-termination and external electrical cardioversion. These were not further targeted as they were felt to be non-clinical. Further, there was no inducible atrial fibrillation.

Following the procedure, the patient was observed overnight and discharged home the next morning. He had an uneventful post-procedure course. On follow-up, he has not had any symptomatic arrhythmias (4 months).

DISCUSSION

This is the first report to describe an endovascular method to access the pulmonary venous atrium for catheter ablation in patients with an extra-cardiac Fontan palliation. The use of electrocautery and balloon dilatation of the puncture allows an endovascular approach to solving an extremely challenging access problem for catheter ablation in this patient population.

Most patients born with single ventricular physiology eventually undergo the Fontan operation. With the older types of Fontan and modified Fontan procedures, the incidence of late supraventricular tachyarrhythmias has been reported to be 29%¹⁰, 17%¹¹ and 18.5%¹². With the extracardiac Fontan procedure, it is reported to be as low as 5% in a series of 200 patients⁹. However, as in this case, many patients will have received palliative surgical procedures prior to extracardiac anastomosis. These palliative surgeries can include procedures such as PA banding to reduce pulmonary blood flow, atrial septectomy, cavopulmonary (e.g. Glenn) shunts, aortopulmonary (e.g. Blalock-Taussig) shunts, and pacemakers, among others. Arrhythmogenic sequelae can be related to atriotomy scars, alterations in atrial pressure or volume load, or alterations in atrial refractoriness secondary to sinus node dysfunction.

The clinical need for catheter access to the pulmonary venous atrium following Fontan surgery led Nehgme et al (2006)¹³ to develop a transthoracic puncture technique allowing direct access to the pulmonary venous atrium. They reported a series of six procedures in five patients, each having supraventricular arrhythmias in the setting of a lateral tunnel Fontan. Access was achieved by puncture in a selected intercostal space adjacent to the right margin of the sternum. They achieved a 100% success rate in mapping and RF ablation. The arrhythmias treated were one ectopic atrial tachycardia and five atrial reentrant tachycardias. There was one pneumothorax and two hemothoraces. One patient (who did not develop pneumothorax or hemothorax) remained in the intensive care unit and underwent Fontan palliation but ultimately died with multiorgan failure and sepsis.

A patient with an extracardiac Fontan conduit with significant leak of a patch-closed right atrioventricular valve was included in a case series¹⁴ describing the use of direct transapical left ventricular (LV) puncture as a method to obtain access to the left heart for interventions including ablation. In this series of 5 patients, there was one puncture of the left anterior descending coronary artery and one hemothorax following sheath removal, neither of these occurring in the patient with the extracardiac Fontan. Their conclusion was that “blind”

percutaneous LV puncture was effective and could be considered, but that puncture under direct visualization through a purse-string suture following mini-thoracotomy was preferable due to fewer complications.

Khairy and colleagues¹⁵ described a case in which catheter access was obtained via atrial puncture under direct visualization through a purse-string suture. Their patient was a 6 year old girl who had only hours earlier undergone Fontan palliation with an extracardiac conduit for hypoplastic left heart and Ebstein's malformation of the right-sided atrioventricular valve. She had received concomitant surgical WPW ablation, but unfortunately pathway-mediated hemodynamically unstable SVT recurred almost immediately after surgery. They were able to perform successful ablation of the accessory pathway without cardiopulmonary bypass, but commented on the challenge of hand and catheter stabilization for delicate positioning while working directly on a beating and compressible heart.

Our patient with a non-fenestrated extracardiac conduit developed highly symptomatic supraventricular arrhythmias from atrial tachycardia, likely intraatrial reentry. These arrhythmias began prior to and progressed despite extracardiac Fontan conversion surgery. Catheter based ablation was performed by way of trans-conduit puncture using a transseptal needle and radiofrequency energy to cross the conduit. Although multi-modality imaging was utilized to minimize the risk of peri-cardiac bleeding, fluoroscopy was the mainstay for the trans-conduit puncture. Balloon angioplasty of the conduit puncture was necessary to place a sheath across the puncture site. Following trans-conduit sheath placement, electroanatomic mapping and radiofrequency ablation was performed successfully and with relatively unrestricted access to the atrium.

With improved survival of patients like these, the incidence of atrial arrhythmias requiring intervention can be expected to increase. Even if the atrial arrhythmia incidence is decreased in patients with extracardiac as compared to intracardiac conduits, many will have had previous surgeries including atriectomy conferring a high risk of atrial arrhythmias. Further, these atrial arrhythmias tend to be poorly tolerated from the hemodynamic standpoint in these patients. Trans-conduit puncture, though challenging, provides a minimally invasive option for definitive treatment of these highly symptomatic and often life-threatening arrhythmias. In the appropriate setting, this technique may prove to be a viable alternative to on- or off-pump arrhythmia surgery, hybrid atrial or ventricular puncture through a purse string, or direct transthoracic puncture. Although no complications were encountered in this patient, further studies are needed to determine the safety of this procedure and a careful assessment of risks versus benefit needs to be made on a case by case basis.

Supplementary Material

Refer to Web version on PubMed Central for supplementary material.

Acknowledgments

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Figure 1. Rendering of post-surgical anatomy

There is tricuspid valve atresia with hypoplastic right ventricle and well-developed systemic left ventricle (LV). Great arteries are D-transposed with the aorta (Ao) anteriorly dextraposed and the banded pulmonary artery (PA) posteriorly levoposed. Superior vena cava (SVC) is connected to PA via bidirectional Glenn shunt. Extracardiac conduit connects inferior vena cava to PA. Note close proximity of extracardiac conduit to "right atrial" portion (RA) of pulmonary venous atrium. The "left atrium" (LA) and mitral valve (MV) are labeled.

Figure 2.

Fluoroscopic images showing (A) still-frame from conduit angiogram, (B) transseptal needle with contrast staining of conduit prior to crossing, (C) dilator of transseptal sheath crossing conduit into pulmonary venous atrium, (D) Toray valvuloplasty wire coiled in atrium with angioplasty balloon inflated across conduit crossing, and (E) still-frame from angiography of pulmonary venous atrium.

Figure 3.

(A) ECG tracings of the atrial tachycardia. (B) Catheter positions in LAO at ablation site (Abl) along anterior border of atrio-ventricular (mitral) valve. A duodecapolar catheter (Ao) crosses aortic valve into LV. (C) Electroanatomic voltage map of pulmonary venous atrium is shown on right with healthy atrial tissue in purple and a scar visible on this anterolateral view. Extracardiac conduit tube is in beige. Solid black line marks approximate location of mitral valve. Voltages below 0.05V were mapped to gray, between 0.05V and 0.5V to colors between red and purple, and above 0.5V to purple.