Cryoballoon pulmonary vein isolation after extracardiac Fontan operation

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Introduction

Atrial fibrillation (AF) poses risk to patients with single ventricle physiology and Fontan palliation, but the pulmonary vein isolation (PVI) procedure has not been reported after extracardiac Fontan operation. We present a case in which a transcaval puncture technique was used to access the pulmonary venous atrium for cryoablation in a patient with tricuspid atresia and an extracardiac Fontan conduit.

Case report

A 30-year-old man with a history of tricuspid atresia with normally related great arteries and extracardiac Fontan conduit presented for ablation of AF. The patient had undergone Blalock-Thomas-Taussig shunt in the initial days of life, followed by a Bjork-type Fontan operation (right atrium to right ventricular outflow tract conduit) at 3 years of age. Because of the subsequent development of AF, his Fontan conduit was converted to an extracardiac conduit at 19 years of age. In the subsequent year, he developed atrial flutter. Amiodarone was initiated and later discontinued after the onset of thyroiditis.

The patient received intermittent medical care for the next 10 years, after which time he presented to his primary care doctor with abdominal pain. An initial evaluation for gastrointestinal abnormality was performed, followed by electrocardiography, which demonstrated coarse AF at a rate of 165 beats/min. The patient was prescribed warfarin and diltiazem and referred to an adult congenital heart disease specialist. At that appointment, nearly a month later, his rhythm was still noted to be AF. Elective cardioversion was performed. Since arrhythmia recurrence was expected to be poorly tolerated and he had been intolerant of amiodarone previously, he was referred for catheter ablation.

The decision was made to proceed with PVI for AF. On review of the contrast-enhanced computed tomography scan of the heart, it was appreciated that the inferior vena cava abutted the morphologic right atrium inferior to the anastomosis with the Gore-Tex conduit (W. L. Gore and Associates, Flagstaff, AZ) (Figure 1).

Catheterization was performed under general anesthesia with both transesophageal echocardiography and fluoroscopy guidance. Access was obtained in bilateral femoral veins, left femoral artery, and right internal jugular vein. Before PVI, hemodynamic catheterization revealed a mean Fontan pressure of 15–16 mm Hg. Using a 4-F pigtail catheter, angiography was performed and a region of cavoatrial overlap was confirmed just below the extracardiac conduit.[1]

An 8.5-F Mullins sheath (Medtronic, Minneapolis, MN) was advanced over a wire into the inferior vena cava below the extracardiac conduit. The wire was removed, and a BRK transseptal needle (St. Jude Medical, St. Paul, MN) was placed against the portion of inferior vena cava tissue adjacent to the right atrium. Contrast was injected in order to stain the tissue. Then, the needle was advanced into the morphologic right atrium, through the large atrial septal defect, and into the morphologic left atrium.[1] Positioning was confirmed with both pressure transduction and contrast injection. Heparin was administered and activated clotting time determined to be therapeutic. The Mullins sheath was exchanged over a wire for a FlexCath steerable sheath (Medtronic) after serial dilations. The EnSite NavX electroanatomic system (St. Jude Medical) was used for catheter localization and guidance. A deflectable decapolar catheter (St. Jude Medical) was advanced into the left pulmonary artery in order to record atrial electrograms, but later pulled back into the superior vena cava in order to document phrenic nerve capture during PVI.

Using a 20-mm Achieve circular mapping catheter (Medtronic), a voltage map of pulmonary venous atrium

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KEY TEACHING POINTS

- In patients with single ventricle physiology and Fontan palliation, atrial fibrillation is associated with an increased risk of thromboembolism and worsening hemodynamics.
- The prevalence of atrial fibrillation in this population increases with age and with the complexity of congenital heart disease.
- A transcaval puncture technique can be used to access the pulmonary venous atrium and perform cryoablation for pulmonary vein isolation.

was created, demonstrating electrical activity in all 4 pulmonary veins. With a 28-mm Arctic Front Advance cryoballoon catheter (Medtronic), each of the pulmonary veins was targeted (Figure 2). Cryoballoon ablation completely eliminated conduction in the right superior pulmonary vein, but only partially eliminated conduction in the remaining veins, likely because of inadequate balloon-tissue contact related to the distorted anatomy. Therefore, isolation of these pulmonary veins was completed with a contact-force irrigated radiofrequency ablation catheter (TactiCath, St. Jude Medical). After delivery of these lesions, there were no recorded electrograms in any of the 4 pulmonary veins, demonstrating entrance block (Figure 3). Pacing was performed at each of the pulmonary vein ostia to ensure exit block, confirming bidirectional electrical isolation of all 4 pulmonary veins.

Six months after the procedure, the patient remains free of recurrent AF without antiarrhythmic drugs, with only a brief episode of atrial tachycardia (<1 minute) demonstrated on a rhythm monitor.

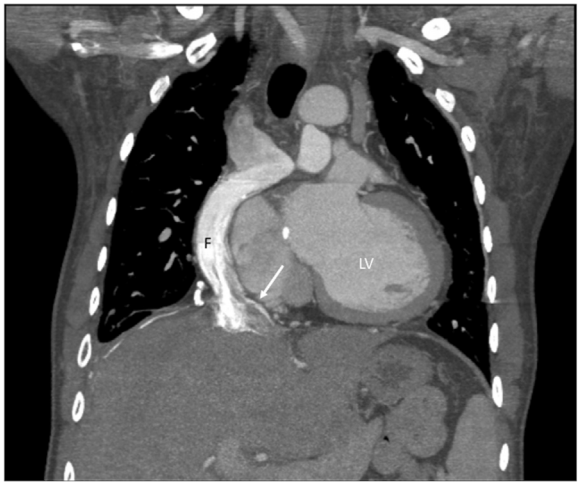

Figure 1 Contrast-enhanced computed tomography angiogram demonstrating that the inferior vena cava is in direct contact with the native atrium (*arrow*). F = Fontan; LV = left ventricle.

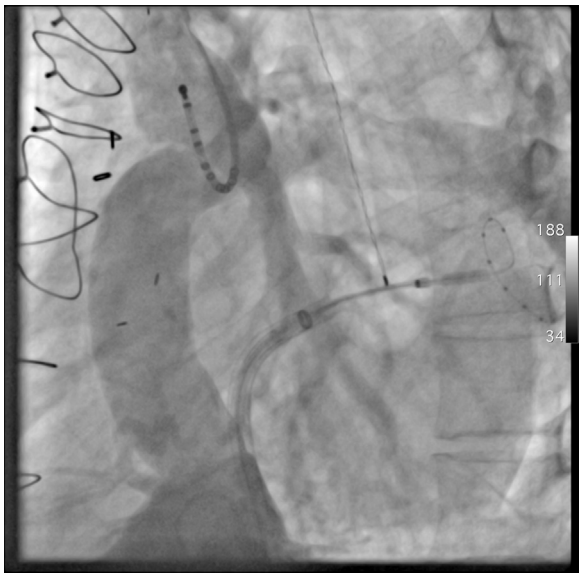

Figure 2 Left anterior oblique projection of the cryoballoon in the left superior pulmonary vein. A pigtail catheter has been placed in the inferior vena cava for simultaneous venography of the extracardiac Fontan conduit.

Discussion

As many as 50% of patients with Fontan palliation will develop atrial tachyarrhythmias, which are associated with an increased risk of thrombosis and worsening hemodynamics.[2] In a large multicenter cohort of patients with Fontan palliation, 32% underwent arrhythmia treatment at 12 years after operation.[3] The prevalence of AF in particular seems to increase with age and with the complexity of congenital heart disease.[4,5]

Ablation of AF poses numerous challenges to this population, including the presence of multiple tachycardia circuits, limited catheter access, distorted anatomy, difficulty achieving adequate lesion depth, and hemodynamic instability.[2] In a heterogeneous population with congenital heart disease, the success rate of PVI has been reported to be comparable to the rate in patients with acquired heart disease,[6,7] but PVI experience in the population with Fontan palliation is limited and has not previously been reported after the extracardiac modification.

Completion of an extracardiac Fontan includes connection of the inferior vena cava and the pulmonary artery by way of a Gore-Tex tube, so percutaneous access to the native atria may prove challenging. A transconduit puncture has been reported,[8] but this technique requires serial balloon dilation to advance the guiding sheath, a process that is difficult and time-consuming. When there is adequate contact between the inferior vena cava and the native atrium, transcaval puncture can be performed for more facile access. Such apposition is believed to develop in most patients because of the fixed size of the rigid extracardiac conduit in a heart that enlarges after Fontan operation as a result of either somatic or pressure-related growth.[1] Direct access to the morphologic left atrium using the technique described here facilitates the use of cryoballoon ablation, a newer method for PVI.[9]

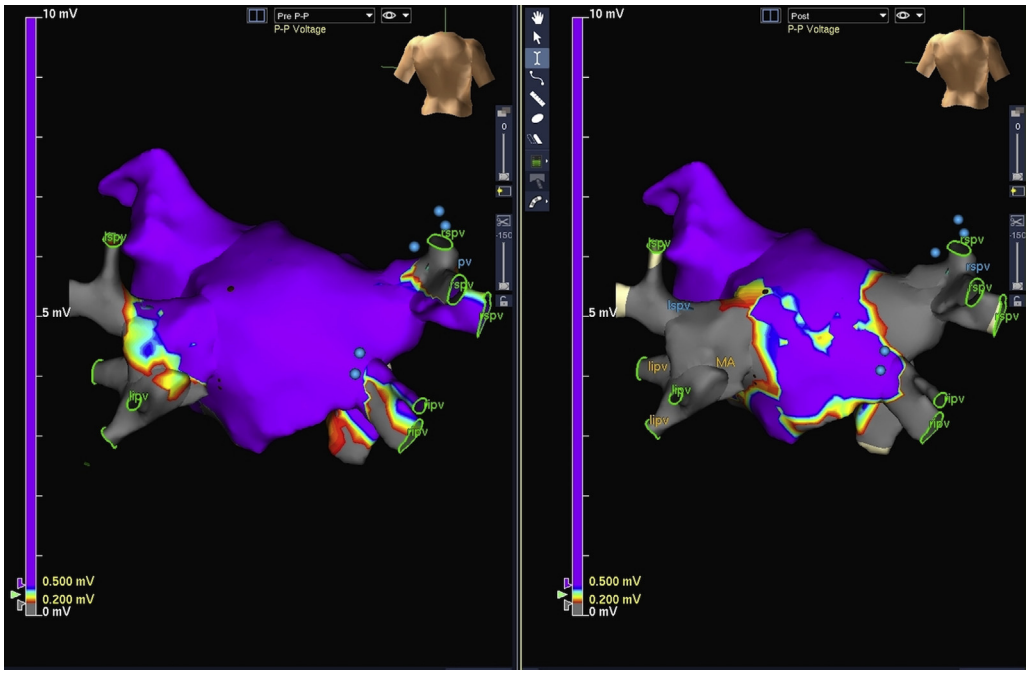

Figure 3 Electroanatomic maps before (left) and after (right) the procedure, demonstrating electrical isolation of all 4 pulmonary veins.

Further experience with this approach will likely grow as this population continues to age, and AF becomes more prevalent in patients with surgically treated congenital heart disease.

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