Catheter Ablation of Atrial Fibrillation in Transposition of the Great Arteries Treated With Mustard Atrial Baffle

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The survival of patients with D-transposition of the great arteries (D-TGA) has dramatically improved with corrective surgical procedures, including the atrial and arterial switch operations. In the current era, the majority of the adult patients typically have undergone atrial switch operations, either a Senning or a Mustard procedure. Late complications include baffle obstruction, systemic ventricular failure, systemic atrioventricular valve regurgitation, and rhythm disturbances. The most common atrial arrhythmia in this population is intra-atrial reentrant tachycardia (IART), which has been associated with development of heart failure and death. Proarrhythmic factors include atrial enlargement from tricuspid regurgitation and systemic right ventricular failure, as well as suture lines. As this population ages, chronic hemodynamic stress may result in atrial fibrillation as well. We present the first report of catheter ablation for treatment of atrial fibrillation in a patient with D-TGA treated with prior Mustard repair.

Case Report

A 41-year-old man with D-TGA status post Mustard repair presented with longstanding, recurrent atrial arrhythmias. He originally underwent balloon atrial septostomy 2 days after birth and a subsequent atrial switch procedure using the Mustard technique at 2 years of age. He did well until age 32, at which time he had recurrent symptomatic IART. EP study was performed. Cavotricuspid isthmus (CTI)-dependent flutter was induced and confirmed by entrainment mapping. Lines of ablation were drawn across the systemic venous baffle portion of the “medial” CTI and the pulmonary venous “lateral” CTI using a retrograde, aortic approach. Flutter terminated with ablation and could not be reinduced. Bidirectional block was demonstrated across the CTI. He remained arrhythmia-free for 2 years, then presented again at age 34 with exertional intolerance and fatigue due to partial obstruction of the pulmonary venous baffle. He underwent pulmonary venous pathway between the pulmonary veins to the base of the left atrial appendage, extending through the narrowest portion of the pathway. A large Gore-Tex patch was secured to the pulmonary venous pathway. His systemic right ventricle was moderately impaired at that time, with moderate tricuspid regurgitation. Over the following 4 years, he had recurrent, highly symptomatic atrial fibrillation requiring several DC cardioversions despite treatment with dofetilide and then amiodarone. After much discussion, catheter ablation was recommended.

A CT scan (Figure 1D) was obtained before the ablation both to facilitate procedural planning and to integrate with the electroanatomic mapping system (NavX, St Jude Medical, St Paul, MN). Intracardiac echocardiography (AcuNav, Siemens, Mountain View, CA) was used to guide catheter positioning and monitor pulmonary venous flows (Figure 1C). The patient was placed under general anesthesia, using a propofol infusion, and was intubated for the procedure. A transbaffle puncture was performed, using fluoroscopy and contrast angiography to access the pulmonary venous atrium (Figure 1A and 1B). The circular mapping catheter (Lasso, Biosense Webster, Diamond Bar, CA) was positioned in the left common pulmonary vein and the ablation catheter in the right inferior pulmonary vein (Figure 2B and 2C). Atrial fibrillation triggers were elicited with isoproterenol (3–30 μg/min) and with atrial burst pacing. During infusion of isoproterenol, isolated premature atrial depolarizations were recorded from the right inferior pulmonary vein (Figure 2A, asterisk) and atrial tachycardia was recorded from the left common pulmonary vein, which did not initiate atrial fibrillation (arrows). Circumferential antral pulmonary vein isolation was performed; the right veins were targeted independently (Figure 3A). Ablation was performed with an open irrigated catheter (EZ Steer Thermocool, Biosense Webster, Diamond Bar, CA). Both entrance block (Figure 3B) and exit block (Figure 3C) were demonstrated in all 3 pulmonary vein ostia. After ablation, isoproterenol was administered again, this time without any triggers identified; dissociated firing within the
The pulmonary vein ostia was seen. Bidirectional block was confirmed across the CTI, from the prior ablation. Aggressive atrial burst pacing and programmed stimulation failed to induce sustained arrhythmias.

There were no complications from the ablation procedure. Dofetilide was restarted, in accordance with our practice of treating patients with an antiarrhythmic drug for 6 weeks following atrial fibrillation ablation. The patient was discharged on the second postoperative day. Over the following 6 months, he had 2 episodes of IART, with cycle length 290 ms, superiorly directed p wave, and 2:1 AV conduction. Both episodes were symptomatic and required DC cardioversion. He has had no recurrences of atrial fibrillation.

**Discussion**

To our knowledge, this is the first report of catheter ablation for treatment of atrial fibrillation in a patient with D-TGA status post Mustard atrial switch. Although survival of patients with D-TGA has dramatically improved with corrective surgical procedures, atrial arrhythmias are a common late sequela, approaching 25% and increasing with time. Although the most common incident arrhythmia is IART, atrial fibrillation may occur as well. Electrophysiology studies have confirmed abnormalities of sinus node function as well as atrial conduction and refractoriness.

Our patient had several risk factors for atrial arrhythmias, including complex atrial reoperation, impaired systemic right ventricular hemodynamics, and tricuspid regurgitation. Atrial arrhythmias in patients with systemic right ventricular dysfunction are often highly symptomatic and poorly tolerated. Although antiarrhythmic drugs are a reasonable first line of treatment for atrial fibrillation, efficacy is often inadequate and proarrhythmia may occur. Therefore, in patients with D-TGA treated with Mustard atrial baffle, with recurrent

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**Figure 1.** D-transposition of the great arteries with Mustard atrial switch anatomy. A, Contrast angiography opacifies the inferior vena cava baffle, the systemic venous atrium, and the superior vena cava baffle. B, Transbaffle puncture is performed with the second sheath now in the pulmonary venous atrium. Contrast opacifies the right superior pulmonary vein as well as the pulmonary venous atrium. C, The intracardiac echocardiography catheter, placed in the systemic venous atrium, is used to guide positioning of the circular mapping catheter (L) at the ostium of the left common pulmonary vein. D, Contrast enhanced CT scan demonstrates the pulmonary venous atrium and pulmonary veins in gray, and the systemic venous atrium, inferior vena cava baffle, and superior vena cava baffle in purple. SVC indicates superior vena cava; IVC, inferior vena cava; LCPV, left common pulmonary vein; RIPV, right inferior pulmonary vein; RSPV, right superior pulmonary vein; and SVA, systemic venous atrium.

**Figure 2.** Pulmonary vein triggers. A, During isoproterenol infusion (30 μg/min), a premature depolarization is recorded from the right inferior pulmonary vein (3rd beat, asterisk) followed by atrial tachycardia from the left common pulmonary vein (4th through 9th beats, arrows). B, Right anterior oblique projection with duodecapolar catheter from the right internal jugular vein, extending through the superior vena cava (SVC) baffle and the systemic venous atrium into the inferior vena cava (IVC) baffle; decapolar catheter positioned within the systemic venous atrial appendage; circular mapping catheter positioned in left common pulmonary vein; and ablation catheter positioned in right inferior pulmonary vein. C, Left anterior oblique projection of same catheter positions, LCPV indicates left common pulmonary vein; RIPV, right inferior pulmonary vein.
atrial fibrillation despite antiarrhythmic drugs, catheter ablation is a viable treatment option. We were able to demonstrate pulmonary vein triggers for atrial fibrillation, as is nearly uniformly the case in patients without congenital heart disease.9 We also showed that pulmonary vein isolation is technically feasible using a transbaffle puncture approach to access the pulmonary venous atrium. Further, by isolating the pulmonary veins, atrial tachycardia could not be induced with the same dose of isoproterenol that induced atrial tachycardia before ablation. Last, our case highlights the limitations of arrhythmia inducibility in the electrophysiology laboratory, perhaps exacerbated by the use of general anesthesia. Best catheter ablation results are likely to be achieved with collaboration between adult and pediatric electrophysiologists and interventional cardiologists in this population with unique challenges and risks.10

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Disclosures

None.

References


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Figure 3. Catheter ablation. A, Circumferential antral ablation was performed around the left and right pulmonary veins. The left common pulmonary vein was ablated as a single ostium. The right superior and inferior pulmonary veins were ablated as separate ostia. B, During ablation around the left common pulmonary vein, entrance block is achieved, with disappearance of pulmonary vein potentials between the 2nd and 3rd beats (arrows). C, Exit block is demonstrated by failure to capture the atrium with pacing (10 mA, 2 ms) from contiguous lasso poles around the circumference of each vein ostium. SVC indicates superior vena cava; IVC, inferior vena cava; LCPV, left common pulmonary vein; RIPV, right inferior pulmonary vein; and RSPV, right superior pulmonary vein.
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