Getting to the right left atrium: Catheter ablation of atrial fibrillation and mitral annular flutter in cor triatriatum

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Introduction

Cor triatriatum is a rare congenital cardiac anomaly that is characterized by a thin membrane separating the left atrium both functionally and anatomically. Patients with cor triatriatum are at risk for atrial arrhythmias, which can be especially symptomatic owing to a functional mitral stenosis physiology. We present a case of a patient with cor triatriatum and drug-refractory atrial fibrillation (AF) and atrial flutter (AFl) requiring imaging-guided ablation in both partitions of the left atrium.

Case report

A 51-year-old man with no significant medical history presented with palpitations and AF. An echocardiogram demonstrated cor triatriatum with a membrane separating the superior-posterior (including the pulmonary vein inflow) and inferior-anterior (including the mitral valve) left atria. There was a mild membrane gradient, which was not an indication for surgery. Despite medical therapy, he continued to have symptomatic AF and AFl and was referred for catheter ablation. Preprocedural imaging was performed with computed tomography and transesophageal echocardiography, delineating the partitions of the left atrium and the membrane fenestrations (Figure 1).

The patient was brought to the electrophysiology laboratory in sinus rhythm. Using intracardiac echocardiography (ICE) guidance, a double transseptal puncture was performed to gain access into the superior-posterior portion of the left atrium. Wide antral circumferential ablation using a force-sensing, open-irrigated catheter (SmartTouch; Biosense Webster, Diamond Bar, CA) resulted in pulmonary vein isolation (PVI) with entrance and exit block, as demonstrated by a circular mapping catheter (Figure 2). The use of adenosine revealed no dormant conduction.

Electrophysiological testing was performed once PVI was achieved. Rapid atrial pacing induced a narrow complex tachycardia with a cycle length of 250 ms. Entrainment maneuvers confirmed left atrium roof flutter, which was successfully ablated with a roof line connecting the wide antral PVI of the left and right pulmonary veins in the superior-posterior partition, resulting in termination of tachycardia and bidirectional block.

Repeat pacing was performed, which induced a second tachycardia with a cycle length of 264 ms and eccentric coronary sinus activation (distal to proximal). Entrainment pacing confirmed mitral annular flutter. Under ICE guidance, access to the inferior-anterior compartment was obtained by traversing a posterior fenestration. Ablation was performed from the anterior mitral annulus to the circumferential ablation lesions around the left superior pulmonary vein, requiring delivery of lesions on both sides of the atrial membrane. AFl terminated and bidirectional block was attained with completion of the line on the superior-posterior partition of the left atrium (Figures 2 and 3). Repeat atrial pacing induced a third AFl with cycle length 285 ms and concentric coronary sinus activation. Entrainment pacing confirmed cavotricuspid isthmus (CTI)-dependent flutter. A series of ablation lesions were delivered along the CTI with slowing and termination of flutter. No other arrhythmias were induced, and the patient has remained free of recurrence for over 1 year off all medications.

Discussion

Cor triatriatum is a rare form of congenital heart disease (CHD) in which AF can be commonly associated (up to 30%).[1] This report describes the successful use of ICE to help guide ablation in both partitions of the left atrium in a patient with cor triatriatum, AF, mitral annular-, roof-, and CTI-dependent flutter. Though few reports have described successful ablation of AF and atypical AFl in patients with

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KEY TEACHING POINTS

- Preprocedural and intraprocedural imaging is invaluable in guiding ablation in complex congenital heart disease.
- Pulmonary vein isolation and ablation of left atrial flutters can be standardly performed in patients with cor triatriatum as long as the appropriate left atrial chamber is accessed. Access to both partitions of the left atrium can be challenging but successfully accomplished using intracardiac echocardiography.
- Assessing endpoints, including entrance and exit block in the pulmonary veins and bidirectional block across lines, remains essential for a successful procedure.

cor triatriatum, they have not done so in both partitions of the left atrium using ICE guidance for traversing the membrane fenestration.[2–4] This case highlights both the challenges of catheter ablation among patients with CHD and the invaluable resource of preoperative and intraoperative imaging (computed tomography, transesophageal echocardiography, ICE). In this case, ICE imaging allowed for successful ablation along either side of the atrial membrane.

Advances in surgical techniques and treatment of associated comorbidities have led to significant improvement in quality and quantity of life among patients with CHD. Traditionally, the most prevalent arrhythmia in CHD patients has been intra-atrial reentrant tachycardia (IART). However, as a result of improvements in longevity, AF is increasing in prevalence among these patients and varies between 3.7% and 15%, a much larger percentage than that of the general population (0.95%).[5,6] The initiation and maintenance of AF among patients with CHD is multifactorial and includes left atrial dilation (related to volume overload, elevated systemic blood pressure, or left ventricular dysfunction), incisional scarring related to cardiac surgery, sinus node dysfunction (contributing to atrial triggers), and chronic cyanosis.[5]

As AF becomes more common among these patients, advanced therapies will be needed. For example, among patients with CHD referred for cardioversion, 31% had AF and 20% had AF as their sole presentation (without prior IART).[7] Among a cohort of patients with tetralogy of Fallot, AF was the most prevalent atrial arrhythmia (surpassing IART) among patients over the age of 55 years.[8]

Specific recommendations regarding the management of adults with CHD and AF are largely extrapolated from adult practice and literature on surgical Maze procedures in CHD. However, it is reasonable to infer that strategies such as PVI could be considered after failure of antiarrhythmic medical therapy. Small case series have demonstrated subtle differences in outcomes of catheter ablation in patients with CHD.

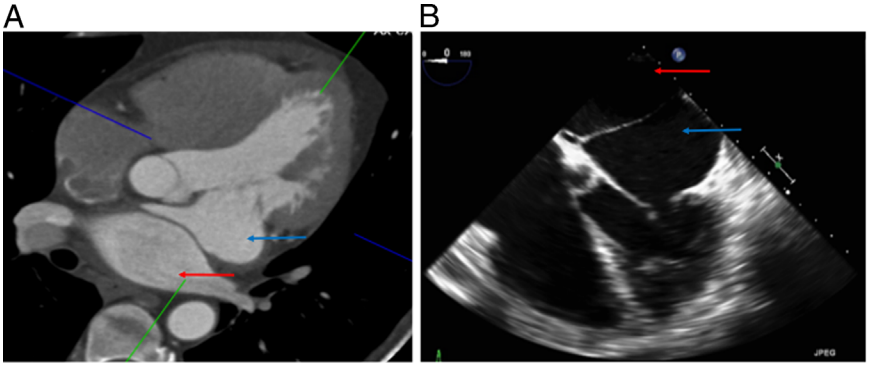

Figure 1 Axial cardiac computed tomography **(A)** and transesophageal echocardiogram **(B)** demonstrating the delineation of the superior-posterior (*red arrow*) and inferior-anterior (*blue arrow*) partitions of the left atrium.

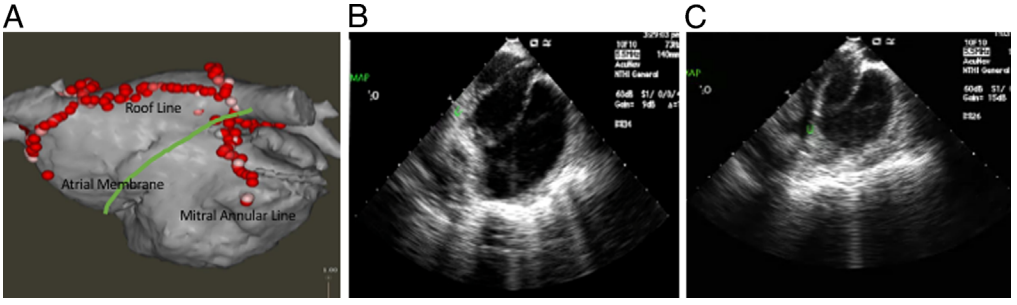

Figure 2 **A:** Carto electroanatomic map. Red tags are ablation lesions resulting in wide antral pulmonary vein isolation (PVI), left atrial roof line, and mitral annular ablation lines. Green line represents the atrial membrane. **B, C:** Intracardiac echocardiography showing the ablation catheter (*green tip*) in the superior-posterior **(B)** and inferior-anterior **(C)** partition of the left atrium while performing PVI of the left pulmonary veins.

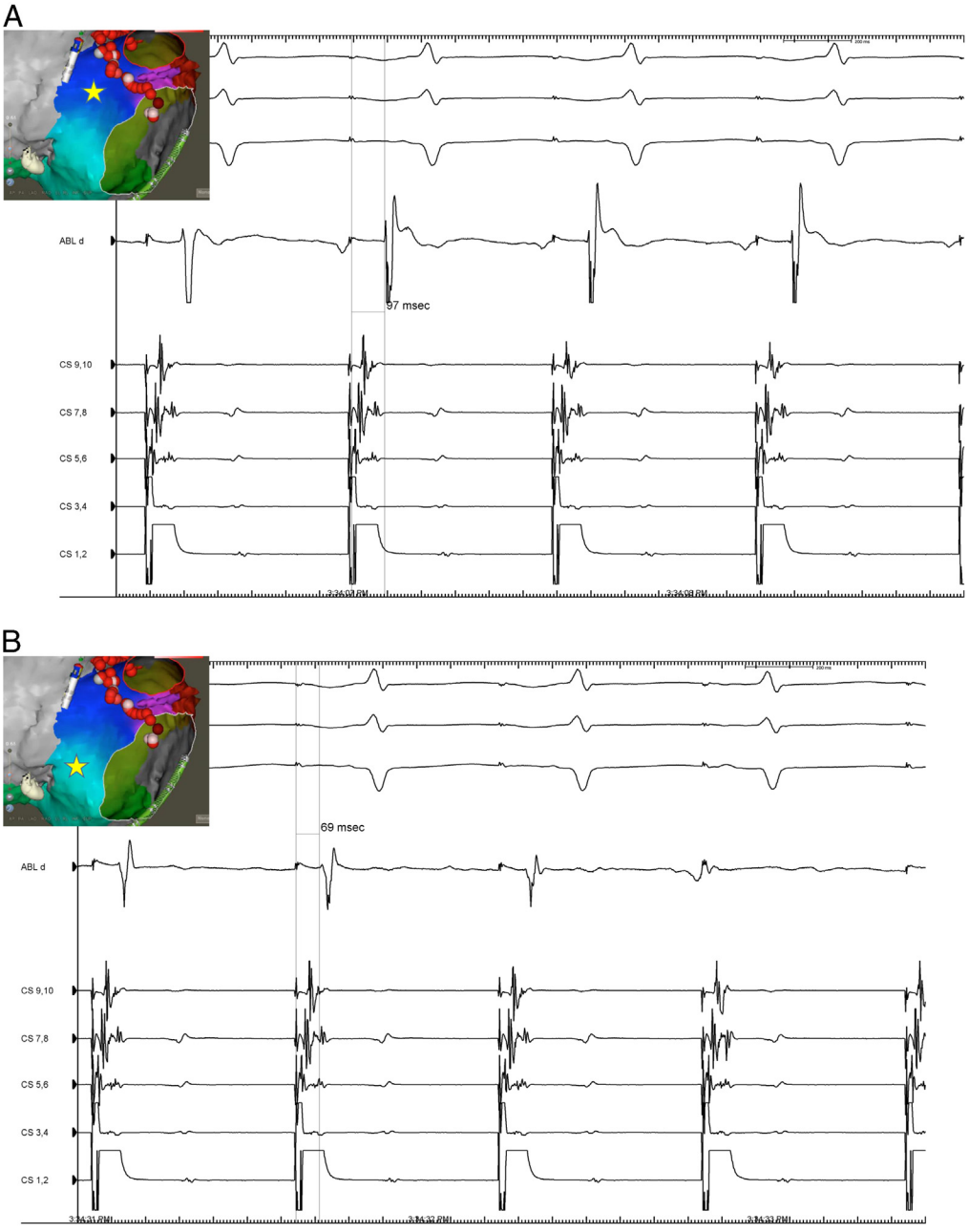

Figure 3 Demonstrating bidirectional block across the mitral annular line. Pacing distal coronary sinus catheter shows differential activation—the area of latest activation (*deep blue isochrone*) was directly medial to the mitral annular line. Pacing from the ablator (*yellow stars*) at 2 different medial sites demonstrated differential pacing (pacing from a site closer to the mitral annular line resulted in a longer conduction time). Similar pacing maneuvers were performed on the lateral side of the line to confirm bidirectional block.

Among 36 patients with simple forms of CHD (mostly atrial septal defects) undergoing PVI, success at 300 days was 42%, compared with 53% of control patients without CHD.[9] Current guidelines provide a IIa indication for PVI in adults with CHD with symptomatic drug-refractory AF (level of evidence C).[6]

There are aspects to AF ablation in this population that have unique challenges. For example, the transseptal puncture is oftentimes more difficult given modified anatomy and/or closure of prior atrial septal defects. ICE is invaluable when performing catheter-based techniques in CHD patients. As in the above example, among patients with surgically or percutaneously closed atrial septal defects, ICE can accurately demonstrate a safe and unobstructed area to perform transseptal puncture to gain access to the left atrium. As newer and better technologies and imaging are

introduced, safety and efficacy of catheter ablation will improve among patients with CHD.

Conclusion

By utilizing preprocedural and intraprocedural imaging to define the anatomy, ablation of AF and AFl can be safely and successfully treated in patients with cor triatriatum, especially among patients with challenging anatomy such as an accessory atrial membrane.

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