Successful atrial fibrillation ablation without pulmonary vein isolation utilizing focal impulse and rotor mapping in an atriopulmonary Fontan

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Introduction

Fontan palliation is designed to separate systemic and pulmonary circulations to ameliorate systemic hypoxemia and relieve ventricular volume overload in patients with a functional univentricular heart. Since its introduction in 1971, the surgical techniques have evolved considerably.[1] Early atriopulmonary (AP) versions of the Fontan incorporated the right atrium (RA) into the systemic venous circuit and are associated with considerable burden of early and late atrial dysrhythmias.[1–5] Although the majority of atrial arrhythmias are macroreentrant RA circuits, such as intra-atrial reentrant tachycardia (IART), the prevalence of late atrial fibrillation (AF) in Fontan patients is increasing.[2,4–7] The mechanisms responsible for AF initiation and maintenance in this population and the role and techniques of catheter ablation for treatment of AF are not well defined.[6,8–12] We present a case of successful AF ablation without pulmonary vein (PV) isolation by utilizing focal impulse and rotor mapping to identify and target mechanisms of AF maintenance in a patient with atrial arrhythmias and medication intolerance in the setting of an AP Fontan.

Case report

A 25-year-old woman with complex congenital heart disease presented with recurrent palpitations and exertional syncope. She had a history of tricuspid atresia, initially palliated with a left modified Blalock-Taussig shunt and a subsequent shunt ligation and AP Fontan. Her childhood and adolescence were relatively unremarkable with the exception of an embolic stroke at age 20 while taking oral contraception, from which she had a complete neurologic recovery and for which she is maintained on anticoagulation. Because of progressive cyanosis and arrhythmias, her adult cardiologist referred her to a children's hospital, where catheterization revealed significant collateralization without focal hemodynamic derangement. A Fontan conversion was recommended, but she declined and shortly afterward relocated and was referred to our program.

On the initial visit, the patient reported significant functional impairment due to fatigue and daily symptomatic palpitations, lasting minutes to hours. Examination was notable for cyanosis and clubbing, with resting O_2 saturation of 83%. Echocardiography demonstrated tricuspid atresia with an unobstructed AP Fontan, severe RA dilation, and normal systemic ventricular function. Baseline electrocardiography demonstrated sinus rhythm with a prolonged PR interval of 216 ms, evidence of RA enlargement, and left-axis deviation. Exercise testing was notable for desaturation to 72%, poor exercise capacity (peak VO_2 17.2 mL/kg/min), and salvos of a regular tachyarrhythmia with ventricular rate of 166 bpm (Figure 1A). Event monitoring revealed symptomatic paroxysmal AF (Figure 1B). Catheterization revealed unobstructed Fontan flow with mean resting Fontan pressure of 5–6 mm Hg. A large venoatrial collateral from the superior vena cava to the left atrium was identified and the coil embolized, with improvement in systemic saturation to 89%. Medical management with beta-blocker was not tolerated. Given her overall normal hemodynamics and her aversion to Fontan conversion or antiarrhythmics, the decision was made to attempt catheter ablation. Preprocedural imaging was performed with cardiac magnetic resonance imaging to evaluate the atrial anatomy and identify pulmonary venous structures.

Description of the procedure

The patient was brought to the electrophysiology laboratory in sinus rhythm. Intracardiac echocardiography was used to

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KEY TEACHING POINTS

- Atrial tachyarrhythmias are a common source or morbidity in patients who undergo classic atriopulmonary (AP) Fontan. The prevalence of atrial fibrillation (AF) is increasing in this population.
- Catheter ablation of AF in AP Fontan patients can be feasible, safe, and efficacious.
- Factors driving AF in Fontan patients are ill defined, and nonpulmonary vein triggers should be considered. Pulmonary vein isolation may not be necessary to maintain sinus rhythm in AP Fontan patients with nonpulmonary vein triggers for AF.
- Ablative strategies targeted toward mechanisms of AF maintenance, such as rotors and focal impulses, should be considered in AP Fontan patients with AF.
- Meticulous identification of triggers and judicious, focused ablative approaches applied by electrophysiologists skilled in catheter ablation and familiar with complex congenital heart disease should be considered an option to treat drug refractory atrial arrhythmias in Fontan patients. A sustained reduction in arrhythmia burden and improvement in quality of life can be achieved in select patients.

guide catheter placement, to create an anatomic geometry of the Fontan pathway to merge with her preprocedural cardiac magnetic resonance imaging, and to monitor hemodynamics and ablation lesions. Therapeutic anticoagulation was maintained during the case. Various catheters, including a decapolar catheter, PentaRay multielectrode catheter (Biosense Webster, Diamond Bar, CA), and a ThermoCool SmartTouch ablation catheter (Biosense Webster), were positioned throughout the Fontan pathway (Figure 1D). A dense voltage map of the Fontan showed mostly normal voltage, except for a dense strip of scar within the inferolateral limb of the Fontan, contiguous with the inferior vena cava (Figure 1E).

Electrophysiological testing was performed once the voltage map was acquired. Triggers initiating an IART with a tachycardia cycle length of 280–310 ms were noted during isoproterenol infusion (Figure 1C). Activation mapping demonstrated a microreentrant circuit with early activation within the inferolateral Fontan, at the border zone between normal tissue and scar. A series of ablation lesions delivered within the border zone resulted in slowing of tachycardia before termination and transition into AF. Given the persistence of AF, substrate modification was then performed, extending the ablation lesions from the region of IART termination superiorly and inferiorly through the border zone to transect channels within scar.

Furthermore, in order to identify possible drivers of AF maintenance, the decision was made to map potential rotors and focal impulses. Using a focal impulse and rotor mapping multielectrode basket catheter and RhythmView mapping system (Topera Inc, Abbott EP, Abbott Park, IL), rotors and focal impulses were identified. A focal impulse and an adjacent fixed rotor were identified in the lateral aspect of the Fontan pathway near the previously identified border zone of the Fontan scar (Figure 2). The region containing the rotor and focal impulse was ablated to noncapture, and further ablation was performed to anchor this region inferiorly to the previously identified area of scar. AF self-terminated. Bidirectional block was then confirmed across ablation lines. Several other rotors noted to be within areas of normal voltage or along the atrioventricular node/His region were not targeted for ablation given the potential risks of complete heart block and proarrhythmia.

An additional macroreentrant cavo-tricuspid isthmus (CTI)–dependent atrial flutter (AFL) circuit with a tachycardia cycle length of 230 ms was induced with rapid atrial pacing and isoproterenol infusion. CTI ablation was performed with AFL termination, and bidirectional block was confirmed with differential pacing. Repeat pacing failed to induce sustained atrial arrhythmias. The patient has remained free of significant arrhythmias without medications for over 18 months, with marked improvement in her functional capacity and quality of life.

Discussion

Atrial arrhythmias are highly prevalent in patients with Fontan palliation for functional univentricular heart and are associated with significant morbidity and mortality.[2,5] Arrhythmias may arise from obstructive hemodynamic perturbations, progressive atrial dilation and remodeling, or scar. There is a high failure rate of antiarrhythmic drugs used to treat atrial arrhythmias in these patients.[13] Catheter ablation of atrial arrhythmias in patients with Fontan palliation, the majority of which are right-sided, has historically been associated with lower success than in other forms of congenital heart disease, with recurrence rates of 30%–60% reported within 1 year.[5] Ablation of atrial arrhythmias is challenging in Fontan patients for myriad reasons, including distorted anatomy resulting in anatomic and electrical barriers to native atrial tissue, restricted catheter access, inability to deliver lesions of sufficient penetrance, and hemodynamic instability during protracted cases.

AF ablation in Fontan patients is uncommon. Whether the optimal ablative strategy is to target all possible triggers utilizing empiric lines for IART prophylaxis and PV isolation or to selectively identify and target clinically relevant triggers is unclear. We and others have demonstrated durable AF ablation in Fontan patients by solely targeting right-sided triggers.[7,11,14] AF recurrence rates are higher in patients with non-PV triggers for AF,[12] and non-PV triggers for AF, including right-sided triggers, are more common in patients with a normal-sized left atrium,[11] such as many

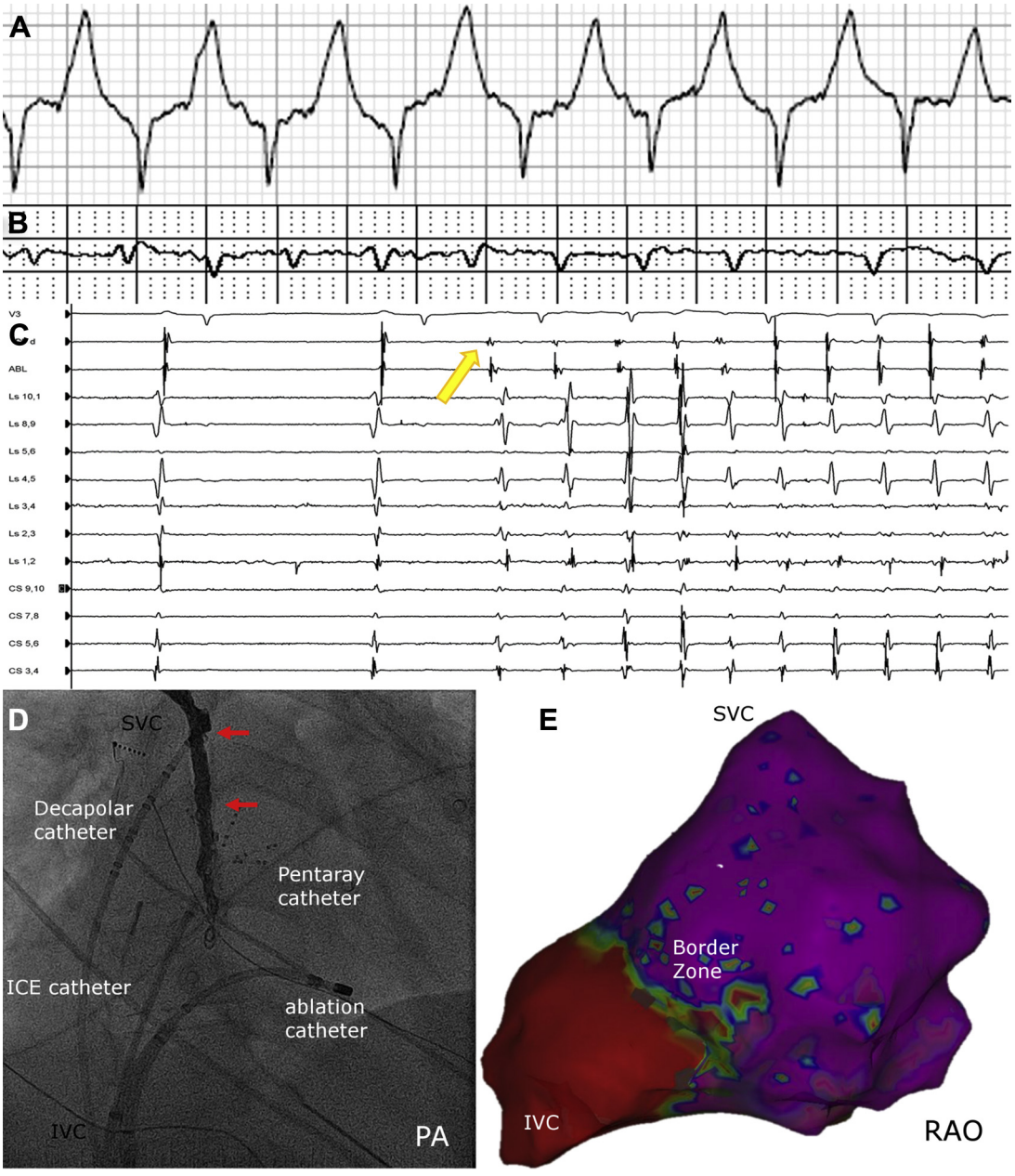

Figure 1 **A:** Regular narrow-complex long RP tachycardia elicited during exercise study. **B:** Symptomatic irregularly irregular tachycardia without P waves consistent with atrial fibrillation was noted on event monitor. **C:** During isoproterenol infusion, triggers were noted with runs of intra-atrial reentrant tachycardia with earliest signals at the ablation catheter (*yellow arrow*). **D:** An array of catheters was utilized to appropriately investigate and ablate tachycardias. The ablation catheter is positioned at the border zone. Embolization coils are seen within the collateral vessel (*red arrows*). **E:** Voltage map of the right atrium/Fontan pathway revealed a dense strip of scar (*red*) within the inferolateral limb of the Fontan, continuous with the inferior vena cava, with a border zone between normal tissue and scar. ICE = intracardiac echocardiography; IVC = inferior vena cava; PA = posteroanterior; RAO = right anterior oblique; SVC = superior vena cava.

patients with AP Fontan. In this case, the patient did not have significant hemodynamic derangements and had not previously undergone ablative attempts. Multiple arrhythmias were demonstrated, as is often the case in Fontan patients.[15] We targeted all relevant mechanisms of initiation and maintenance of AF that we uncovered. A microreentrant IART circuit in the inferolateral Fontan, a macroreentrant CTI-dependent AFL, and focal impulses and rotors within zones of abnormal voltage were targeted (Figure 3). Empiric PV isolation was not performed. In this case, the ablation was successful and durable.

We propose that catheter ablation is a viable treatment strategy to address arrhythmia burden in AP Fontan patients who have drug intolerance or drug-refractory atrial arrhythmias without apparent hemodynamic abnormalities, especially those who are at high surgical risk.[2] Surgical conversion of AP Fontan to an extracardiac total cavopulmonary connection with biatrial surgical maze procedure is

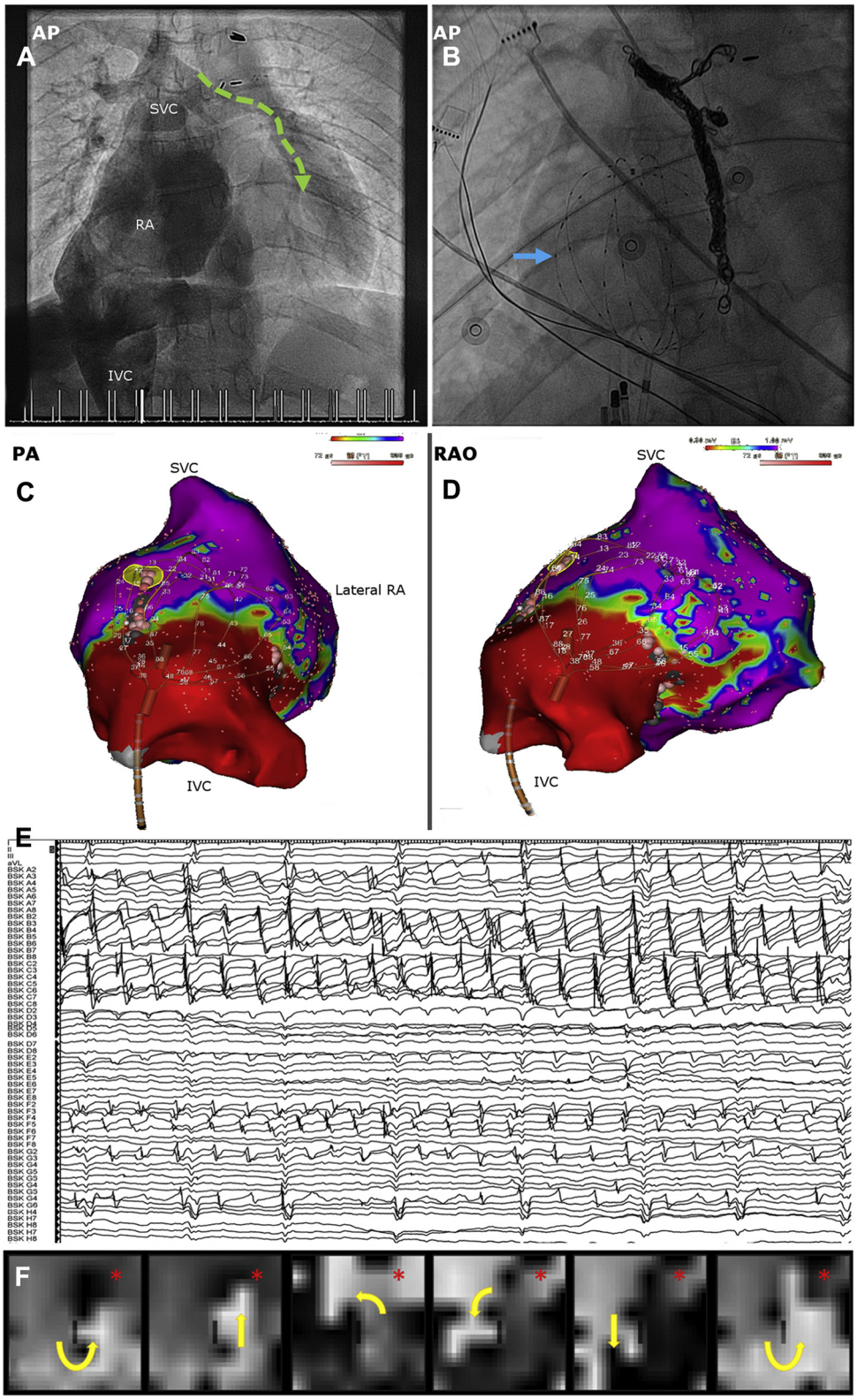

Figure 2 **A:** Fontan angiography revealed a markedly dilated IVC and RA, with unobstructed Fontan pathway to branch pulmonary arteries. A large venoatrial collateral from the SVC to left atrium was seen (*green arrow*) and subsequently embolized. **B:** A 64-pole FIRM multipolar basket (*blue arrow*) was introduced into the Fontan. **C, D:** Electroanatomic mapping was performed to define any underlying electrophysiological abnormalities contributing to atrial fibrillation maintenance. The FIRM basket catheter is visualized within 3-dimensional high-density electroanatomic maps. A rotor (*yellow area*) was identified within the Fontan near the border zone. **E:** Raw unipolar electrograms collected from the FIRM basket catheter, which are then exported to a proprietary mapping system that determines physiologically plausible activation paths. **F:** FIRM-derived computational phase map depicting electrical propagation during atrial fibrillation, revealing a rotor (*yellow arrows*) and adjacent focal impulse (*asterisks*). AP = anteroposterior; FIRM = focal impulse and rotor mapping; IVC = inferior vena cava; PA = posteroanterior; RA = right atrium; RAO = right anterior oblique; SVC = superior vena cava.

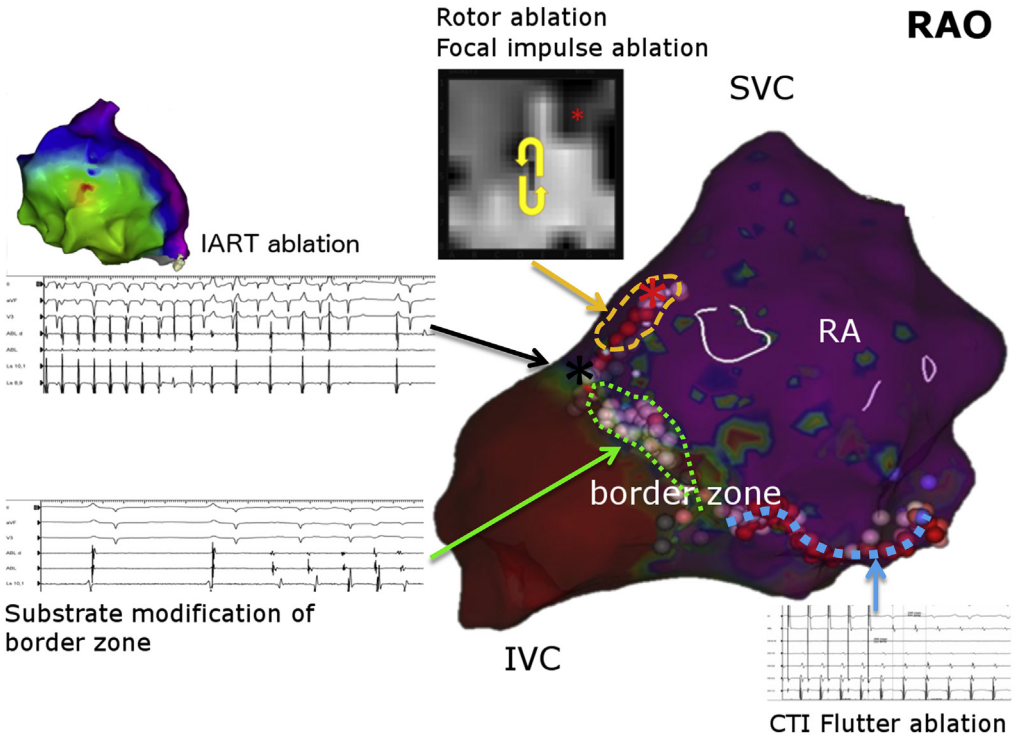

Figure 3 Representative electroanatomic map of the right atrium/Fontan pathway demonstrating the various identified pathologic triggers and mechanisms of maintenance of arrhythmia and subsequent sites of ablation. CTI = cavo-tricuspid isthmus; IART = intra-atrial reentrant tachycardia; IVC = inferior vena cava; RA = right atrium; RAO = right anterior oblique; SVC = superior vena cava.

associated with decreased burden of recurrent atrial arrhythmias, including AF, and has been increasingly proposed as the definitive therapy for significant atrial arrhythmias in AP Fontan patients.[2,3,5] However, conversion is associated with an early surgical mortality of 1%–6% and a prolonged recovery, and it may not be the optimal choice in patients without significant hemodynamic derangements who have not undergone attempts at catheter ablation.

Conclusion

Radiofrequency catheter ablation is an alternative for reduction of arrhythmia burden and improvement in quality of life in carefully selected Fontan patients with drug-refractory or drug-intolerant arrhythmias. Ablation of AF is challenging in Fontan patients. Novel mapping techniques, remote magnetic navigation catheters, and ablative strategies targeting potential triggers and drivers of AF are powerful tools to aid in the identification of electrophysiological triggers for AF in patients with complex univentricular anatomy. We demonstrate successful and durable AF ablation using methodical identification of potential triggers without targeting PV triggers. Such approaches may be widely applicable. Meticulous individualized ablative strategies applied by electrophysiologists skilled in catheter ablation and familiar with complex congenital heart disease are of paramount importance in this patient population.

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