A rare case of superior approach for fluoroless atrial flutter ablation in a patient with dextrocardia, situs inversus, and interruption of the inferior vena cava

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Introduction
Dextrocardia with situs inversus is a congenital anomaly seen in approximately 0.02% of the general population. Congenital interrupted inferior vena cava (IVC) is seen in about 0.2%–0.6% of the population and it occurs in about 5%–15% of patients with dextrocardia.1–3 In these patients with complex anatomy and limited femoral access, catheter ablation is difficult. Several past reports of radiofrequency ablation of cardiac arrhythmias have been published using different approaches: the inferior approach via IVC and azygos continuation and the superior approach via the left subclavian vein.4–8 We report the first case of successful isthmus-dependent atrial flutter ablation using 3-dimensional anatomic mapping system and intracardiac echocardiography without the use of fluoroscopy in a patient with dextrocardia, situs inversus, and an incomplete IVC via a superior approach through persistent right superior vena cava (SVC) into the coronary sinus.

Case report
A 56-year-old woman with known situs inversus was sent to the emergency department from the outpatient setting after being found to have symptomatic atrial flutter with a rapid ventricular response. Initially, cardioversion was performed and sinus rhythm was restored. On subsequent outpatient follow-up, she was noted to have recurrent atrial flutter and a reduced left ventricular ejection fraction of 20%, which was thought to be arrhythmia-induced. Further management was discussed and the patient elected to undergo electrophysiology study and ablation.

KEY TEACHING POINTS
- This is the first reported case of successful fluoroless catheter ablation of isthmus-dependent atrial flutter in patients with dextrocardia, situs inversus, and interrupted inferior vena cava (IVC) via superior approach through the persistent right superior vena cava (SVC).
- In patients with unusual anatomical variations, access to the right cardiac chambers via the persistent right SVC is challenging but feasible using 3-dimensional anatomic mapping and intracardiac echocardiography to identify landmarks and a coherent timing map in the absence of fluoroscopic exposure.
- When considering catheter ablation for atrial flutter in patients with dextrocardia, situs inversus, and interrupted IVC, options are IVC approach through azygos continuation, right persistent SVC approach, and left jugular SVC approach.
- Preprocedural cardiac imaging is of huge significance, particularly in patients with known congenital anomalies, to correctly plan the procedure.

The patient had known dextrocardia with situs inversus totality. Preprocedure imaging with computed tomography angiography showed a persistent right-sided SVC emptying into the coronary sinus. No other venous anomalies were noted. The patient had no history of previous cardiac surgery or prior intracardiac ablation. Electrocardiography done upon presentation to the emergency department showed findings of dextrocardia with limb reversal in I and aVR as well as underlying atrial arrhythmia (Figure 1).

Right femoral vein vascular access was obtained with ultrasound guidance and 2 8F and 1 11F sheaths were placed within the vein. Initially, a 10F SoundStar catheter (Soundstar catheter-Biosense webster, Irvine, CA), an F-J decapolar...
mapping catheter, and a ThermoCool SmartTouch SF ablation catheter (ThermoCool SmartTouch SF ablation catheter-biosense webster, Irvine, CA) were passed through the sheaths into the IVC to perform mapping of the right atrium (RA) anatomy. On SoundStar catheter placement, there was no direct connection between the IVC and inferior portion of the RA. The structures of the RA, including the tricuspid valve and the cavotricuspid isthmus (CTI), were visualized, but tissue was detected between the IVC and the RA. Further mapping with the ablation catheter showed that the IVC connected with the superior venous system through the azygos vein and left SVC draining into the RA. The tricuspid valve and CTI were visualized and there appeared to be a pouch of tissue where the IVC would normally be present. Detailed 3-dimensional (3D) anatomic mapping was performed from the femoral access. An initial activation map was created with the ablation catheter wrapping around the azygos and left SVC into the RA (Figure 2).

Based on known anatomy from preprocedural imaging, vascular access was obtained into the right internal jugular vein with ultrasound guidance. An 8F sheath was placed and the ablation catheter was advanced. This immediately went into the large coronary sinus. The coronary sinus and the RA were mapped, adding to the activation map created previously. The baseline cycle length of the atrial flutter was 220 ms. The map showed an arrhythmia that appeared

Figure 1  Electrocardiogram showed findings of dextrocardia with limb reversal in I and aVR as well as underlying atrial arrhythmia.

Figure 2  A: Right anterior oblique position showing the ablation catheter through the coronary sinus (CS) and on the cavotricuspid isthmus (CTI). Activation map in this projection shows a clockwise circuit around the tricuspid valve going through the CTI. B: Right-sided cranial view showing both the CS and the azygos vein emptying into the left-sided superior vena cava, which enters the right atrium from above. The remnant pouch of the inferior vena cava (IVC) can be seen in both these views. C: Intracardiac echocardiography image that can be seen in panels A and B shown in detail. Ablation catheter can be seen on the CTI between the tricuspid valve and the IVC pouch.
to progress clockwise around the tricuspid valve using the CTI, suggestive of an isthmus-dependent arrhythmia. The pouch of the remnant IVC that was seen on intracardiac echocardiography (ICE) was mapped and no atrial signals were found, indicating that this could be used as a source of connection for ablation with the tricuspid valve. Ablation was performed along the isthmus through the persistent right-sided SVC and the arrhythmia terminated during ablation. Throughout, the decapolar F-J coronary sinus catheter was positioned through the femoral vein access through the left superior vena cava and around the left SVC into the RA adjacent to the isthmus and opposite the coronary sinus (Figure 3). Following completion of ablation, differential pacing was performed on the opposite sides of the isthmus and measurements were taken across the line. Bidirectional transisthmus block was demonstrated with conduction times of greater than 160–170 ms across the isthmus, split signals seen on the line, and differential atrial pacing. In total, 17 applications of ablation were used for 10.5 minutes of radiofrequency delivered. Postprocedure testing showed normal sinus node function and atrioventricular conduction. There were no procedural complications, and the patient was discharged later that day and prescribed 1 month of anticoagulation.

The patient was seen at 1-month and 6-month follow-ups with electrocardiograms showing sinus bradycardia and premature atrial contractions. No further atrial arrhythmias were identified. Repeat echocardiogram demonstrated a left ventricular ejection fraction of 50%–55%.

Discussion

There have been several reports published in the past on successful catheter ablation for arrhythmias in patients with dextrocardia and situs inversus.4–8 Successful ablations in patients with dextrocardia without situs inversus or interrupted IVC have also been reported.9,10 There have only been 4 reported cases so far with anomalous IVC.5–8 All of them have used fluoroscopy in

Figure 3  Right anterior oblique caudal projection showing ablation catheter extending from the coronary sinus and onto the cavitricuspid isthmus (CTI). The CTI is well defined between the tricuspid valve and remnant pouch. The pouch of the inferior vena cava (IVC) left in the right atrium is visualized. The decapolar catheter that was brought around from the femoral vein access through the left superior vena cava can also been seen later in the CTI. Pacing from this catheter confirmed the block along the CTI.
addition to 3D anatomic mapping system and ICE. The common approaches mentioned are inferior approach via IVC and azygos continuation and the superior approach via the left subclavian vein. This is the first case of successful isthmus-dependent atrial flutter ablation using 3D anatomic mapping system and intracardiac echocardiography without the use of fluoroscopy in a patient with dextrocardia, situs inversus, and an incomplete IVC via a superior approach through persistent right SVC into the coronary sinus.

Procedural feasibility in patients with dextrocardia and situs inversus is often challenged by the degree of anatomic variability. An accessory connection between the coronary sinus and left ventricle has been described before in a patient with dextrocardia with complete situs inversus via a right cubital venous approach. Venous access using the subclavian vein has been reported before and could have been considered in our case, but the femoral venous approach was preferred owing to ease of manipulation and improved stability that can be achieved to navigate the ablation catheter through a longer sheath, especially in a patient with complex anatomy. Our case was challenging in that the azygos continuation of the IVC occurred in the patient with dextrocardia, complete situs inversus, and persistent right IVC. Although preprocedural cardiac imaging in our patient did not identify the anomaly of the IVC, it still provided great benefit when needing to adjust the intraprocedural plan. In the presence of such complex anatomy, access to the RA through a stable femoral approach via the azygos vein and persistent right SVC can be challenging but feasible. This led to the pursuit of a different approach in our patient through the right internal jugular vein and subsequent mapping around the coronary sinus and RA, which allowed for further arrhythmia characterization and successful ablation. Furthermore, these complex procedures in patients with anomalous cardiovascular anatomy often led to difficulty in positioning and manipulation of the catheters and, thus, longer fluoroscopy time. In this case, we were able to forgo the use of fluoroscopy and accomplish therapeutic atrial flutter ablation by using a 3D anatomic mapping system and ICE.

Conclusion
Successful ablation of atrial flutter in patients with dextrocardia and situs inversus is often challenging owing to underlying anatomical variants that often limit vascular access and procedure feasibility. The use of 3D anatomic mapping along with ICE may aid and lead to successful cardiac ablations with minimal to no fluoroscopy time in patients with rare anatomical variants of dextrocardia with situs inversus, such as an incomplete IVC with a persistent right-sided SVC.

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