Successful catheter ablation of persistent atrial fibrillation and common atrial flutter in a patient with dextrocardia, situs inversus, and interrupted inferior vena cava with azygos continuation

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Introduction

Congenital interruption of inferior vena cava (IVC) is rare condition seen in 0.1%–0.6% of the population.[1] In patients with limitation in femoral venous access, catheter ablation has been met with difficulty, with several case reports of approach via superior vena cava (SVC), hepatic vein, or aorta using magnetic navigation system. Here we report a case of dextrocardia, situs inversus, and interrupted IVC with azygos continuation, in which successful manual catheter ablation of persistent atrial fibrillation (AF) and common atrial flutter (AFL) was conducted concomitantly with SVC-only approach.

Case report

A 78-year-old man with dextrocardia and situs inversus was scheduled for catheter ablation for symptomatic persistent AF. At age 71, he was diagnosed with paroxysmal AF and had a dual-chamber pacemaker implanted from the left subclavian vein after experiencing syncope caused by bradycardia-tachycardia syndrome. At age 78, his underlying rhythm had transitioned to persistent AF. As he developed worsening dyspnea on exertion, pilsicainide and then propafenone were administered sequentially. As neither of them was effective, the patient chose to undergo catheter ablation for persistent AF. He had a history of type 2 diabetes mellitus.

On electrocardiogram at admission, the baseline rhythm was AF (Figure 1A and 1C). Chest radiograph showed dextrocardia and enlarged cardiothoracic ratio at 52% (Figure 1B). Transthoracic echocardiogram revealed dilation of the morphologic left atrium (LA) with LA volume index of 47 mL/m^2 and mildly decreased left ventricular (LV) ejection fraction at 47%. Transesophageal echocardiography showed no intracardiac thrombi.

Three-dimensional computed tomography (3D CT) revealed dextrocardia, situs inversus totalis, and interrupted IVC (Figure 2A–2E). Instead of directly draining into the right atrium (RA), IVC continued to the azygos vein, which then drained into the morphologic RA via the SVC. The hepatic veins drained directly into the low RA. At this point, because there was limitation in femoral venous approach and difficulty was expected in accessing the morphologic LA, we offered to the patient a referral to another facility

KEY TEACHING POINTS

- Congenital interruption of inferior vena cava (IVC) is seen in 0.1%–0.6% of the population. When considering catheter ablation for atrial fibrillation in such cases with limited femoral access, options are via superior vena cava (SVC)-only approach, IVC approach through azygos continuation, and transaortic retrograde approach.
- The keys to successful transseptal puncture via SVC-only approach are larger curve of the transseptal needle and real-time images acquired with intracardiac echography.
- In patients with unusual anatomical variations such as interruption of IVC and situs inversus, catheter ablation under 3D navigation system is especially useful in obtaining accurate location information and effectively achieving pulmonary vein isolation with electrically continuous lesions with less fluoroscopy time.

KEYWORDS Atrial fibrillation; Atrial flutter; Catheter ablation; Dextrocardia; Interrupted inferior vena cava; Situs inversus; 3D navigation system

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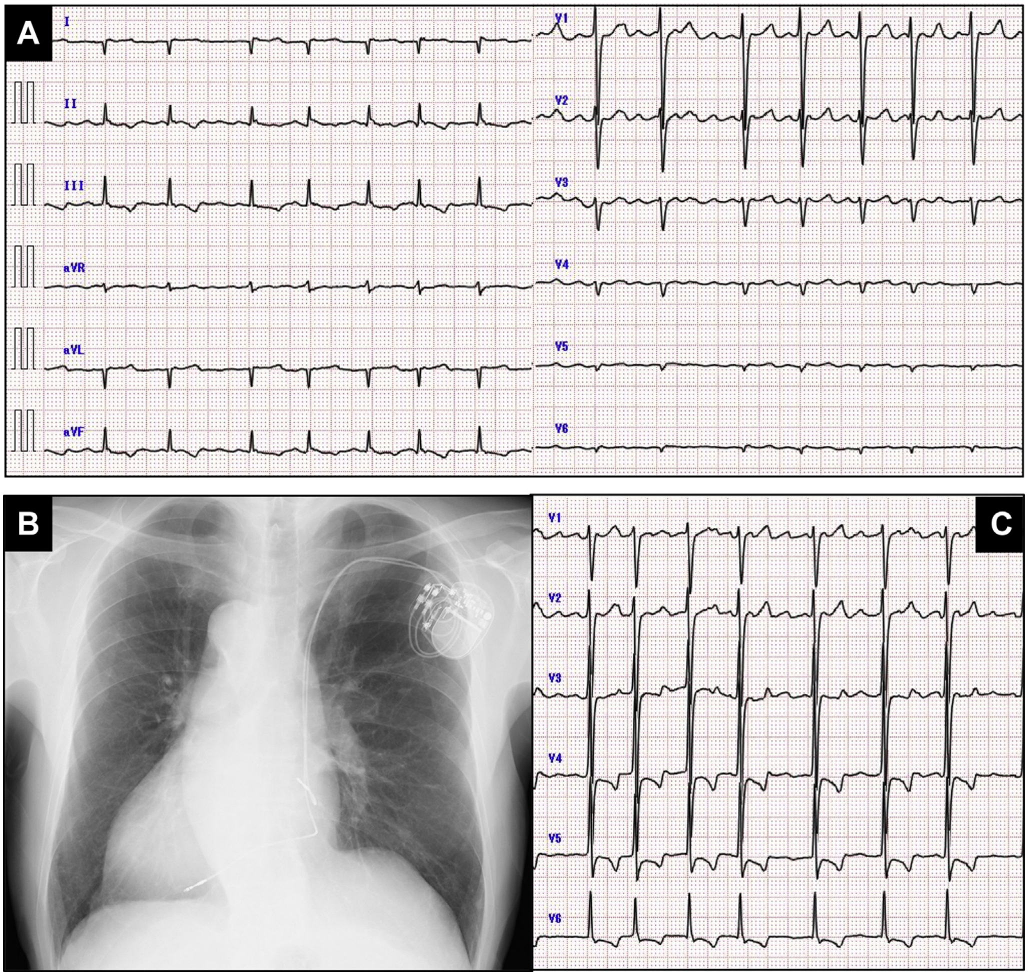

Figure 1 Electrocardiogram (ECG) and chest radiograph at admission. **A:** ECG showing the baseline rhythm as atrial fibrillation. **B:** Chest radiograph showing dextrocardia and enlarged cardiothoracic ratio at 52%. **C:** Right precordial leads of the ECG.

where a transaortic approach was feasible. However, the patient preferred to undergo SVC-only approach at our hospital. To obtain accurate location information of the catheters, we decided to utilize a 3D navigation system (CARTO; Biosense Webster, Irvine, CA) to integrate images acquired with 3D CT and 3D mapping.

The procedure was conducted under local anesthesia and sedation with dexmedetomidine and fentanyl. Ablation was

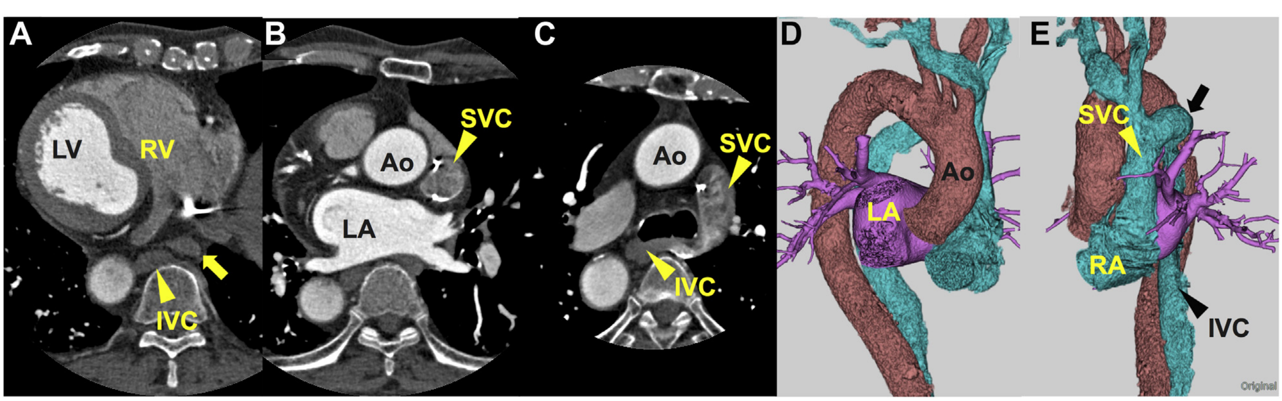

Figure 2 Computed tomography (CT) images. **A:** CT showed situs inversus totalis and dextrocardia. The inferior vena cava (IVC) did not drain into the morphologic right atrium (RA). The hepatic veins (*arrow*) entered the low RA. **B:** The superior vena cava (SVC) entered the high RA. The aorta (Ao) and the left atrium (LA) are also shown. **C:** The IVC continued to the azygos vein and then drained into the SVC. **D:** Three-dimensional CT showing the right anterior oblique (RAO) image of the aorta (Ao) and the LA. **E:** Three-dimensional CT showing the left anterior oblique (LAO) image of the azygos continuation (*arrow*) of the IVC that entered the RA via SVC.

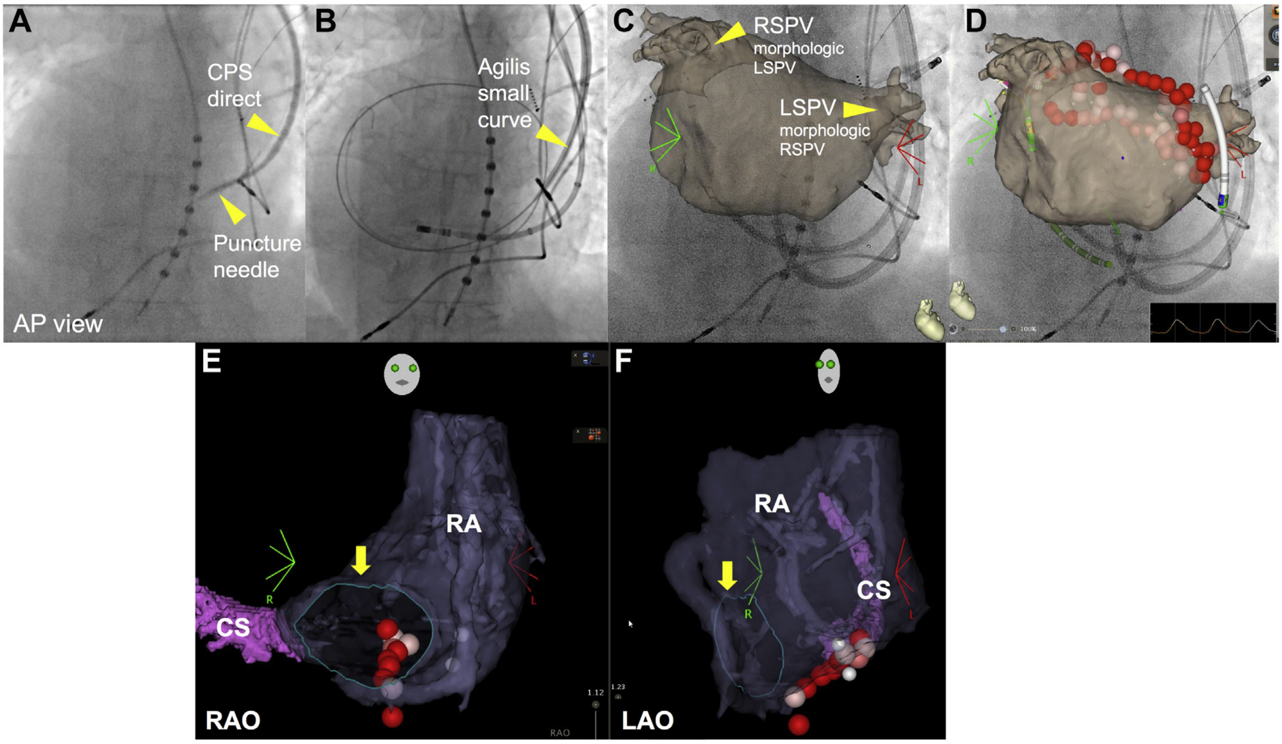

Figure 3 Fluoroscopy and 3D navigation images during catheter ablation. **A:** When the transseptal needle was pushed, it kept slipping below the fossa ovalis. **B:** The septum was punctured and a guidewire was placed into the morphologic left atrium (LA). **C:** The integrated images of computed tomography and fluoroscopy on 3D navigation system showing the LA. An electrode catheter was placed in the morphologic right pulmonary vein (PV), and an ablation catheter was tightly curved and placed in the left PV. **D:** The integrated images after bilateral extensive PV isolation and linear ablation across the LA roof and posterior wall. **E:** After the rhythm transitioned to atrial flutter, the block line was made connecting the confluence of the hepatic vein at low right atrium (RA) and the tricuspid annulus (*arrow*). The right anterior oblique (RAO) image with coronary sinus (CS) is shown. **F:** The left anterior oblique (LAO) image.

performed on anticoagulation with heparin, with activated clotting time maintained above 300 seconds.

Radiofrequency catheter ablation was conducted with an approach from the SVC via right internal jugular and subclavian veins under 3D navigation system. First, a deflectable sheath was inserted to the RA through right internal jugular vein, and ultrasound geometry acquired with intracardiac echography (ICE) (SoundStar; Biosense Webster, Irvine, CA) was merged with reconstructions on 3D navigation system. Then, a catheter introducer sheath (CPS Direct; Abbott, St Paul, MN) and a transseptal puncture needle (Baylis Medical, Montreal, Canada) were advanced through the right subclavian vein. When the transseptal needle was pushed on the right atrial septum, it slipped below the fossa ovalis, and there was difficulty in effectively applying the pushing force toward the septum (Figure 3A). By manually bending the needle and making the curving angle larger, the septum was successfully punctured and a guidewire was placed into the morphologic LA. Next, we attempted to replace the introducer sheath with a deflectable long sheath (Agilis small curve; Abbott, St Paul, MN) via the guidewire, but the sheath again slipped below the fossa ovalis when it was pushed (Figure 3B). The same method was tried through the long sheath inserted via right internal jugular vein, but the manipulation was still difficult. With replacement of the long sheath to one with a larger curve (Agilis large curve; Abbott, St Paul, MN), it was finally advanced to the LA via right subclavian vein.

Next, a multipolar electrode catheter (PentaRay; Johnson & Johnson, Minneapolis, MN) was inserted through the long sheath, and voltages of the bilateral pulmonary veins (PVs) were obtained. As its manipulation was not felt to be smooth, we decided to simplify the procedure by omitting electrophysiological mapping of the LA. An ablation catheter (SmartTouch Surround Flow; Biosense Webster, Irvine, CA) was inserted into the LA through the long sheath, and bilateral extensive PV isolation was completed with radiofrequency energy (power 50 W, contact force 10–20*g*, and ablation index 400–500) (Figure 3C and 3D). Then, while the linear ablation across LA roof and posterior wall was added (power 50 W, contact force 10–20*g*, and ablation index 400–500) and bidirectional block of the PVs and the LA posterior wall were confirmed by pacing the each site with the ablation catheter, the rhythm transitioned to AFL. A multipolar electrode catheter was inserted to coronary sinus by the femoral approach via IVC and then to azygos continuation and SVC, and when the left low RA was paced, postpacing interval matched the interval of AFL. Because the voltage of the area thought to be the confluence of the hepatic veins at low RA indicated a scar region, the block line was made connecting the scar at low RA and tricuspid annulus. The isthmus-dependent AFL was terminated and sinus rhythm was restored (Figure 3E and 3F).

At 10 months after the ablation, the patient remained asymptomatic without dyspnea on exertion, and the rhythm was maintained at sinus. In transthoracic echocardiography, both decrease in LA volume and improvement in LV systolic function were observed, with LA volume index of 31 mL/m^2 and LV ejection fraction of 63%.

Discussion

Situs inversus is a rare condition found in 0.01%–0.02% of the population,[2] and congenital interruption of IVC in 0.1%–0.6%.[1] There have been reports on catheter ablation for patients with limited femoral access, including ones with SVC-only approach, IVC approach through azygos continuation, transhepatic venous approach, and transaortic retrograde approach using magnetic navigation system.[3]

Successful pulmonary vein isolation (PVI) via SVC-only approach has been previously conducted in patients with interrupted IVC, but difficulty was reported in maneuvering transseptal needles.[4–8] Therefore, we first offered a referral to another facility where transaortic approach to the LA with magnetic navigation system was feasible, but the patient preferred to undergo SVC-only approach at our hospital. On the other hand, ablation through azygos continuation was not our option, because difficult catheter manipulation was anticipated through the tortuous course.

As reported previously in SVC-only approach, there was difficulty in effectively propagating the pushing force toward the septum in transseptal puncture. Instead of moving toward the septum, the needle kept slipping below the fossa ovalis. In addition, because this patient concomitantly had situs inversus, the push from the right side of the body only resulted in the needle shifting to the left side of the morphologic right atrium, away from the septum. As this patient had had a dual-chamber pacemaker implanted from left subclavian vein, the only remaining approach was via the right jugular and the right subclavian veins. In addition, the specially designed radiofrequency transseptal needle for SVC approach was not available in Japan.

By manually bending the needle and making the curve extra-large to provide sufficient back-up against the wall of the right atrium as reported previously,[5-8] and by utilizing images acquired with the ICE, the needle was finally pushed toward the fossa ovalis.

Likewise, there was difficulty in advancing the long sheath to the LA until the sheath was replaced with one with a larger curve and counter-traction maneuver was utilized. Additionally, the lack of support from the lower rim of the fossa ovalis might have increased the trouble in controlling the electrode and the ablation catheters, as very tight curves were required to ablate the left-side PVs.

Catheter ablation under the 3D navigation system has been shown to reduce fluoroscopy time, lead to comparable safety end point, and improve ablation outcomes.[9,10] In this case of situs inversus and dextrocardia, the 3D mapping system was especially useful in accurately locating the mirror-image pulmonary veins by providing integrated images of CT scan and 3D navigation. In addition, because manipulation of the catheters through the SVC was not smooth, the markers on 3D images and the visualization of the contact force also had aided in achieving continuous and transmural lesions. Because there was initial difficulty in accessing the LA, the total procedure time was relatively long at 400 minutes. However, under the guidance of the 3D navigation system, PVI time was acceptable at 63 minutes, with fluoroscopic time of only 8 minutes.

In patients with interrupted IVC with azygos continuation and AFL, the circuit of AFL was shown to traverse the isthmus of the atrioventricular valve and the confluence of the hepatic veins as they entered the right atrium.[11,12] In the current case, because the voltage of the area thought to be the entrance of hepatic veins at the morphologic low RA indicated a scar region, the block line was made connecting the confluence and the tricuspid annulus, successfully terminating the isthmus-dependent AFL.

In summary, the keys to accessing the LA were large curves of the transseptal needle and sheaths and real-time images acquired with ICE in catheter ablation with SVC-only approach. As the patient concomitantly had situs inversus totalis, the 3D navigation system was useful in effectively achieving PVI with continuous and transmural lesions.

Conclusion

In a patient with dextrocardia, situs inversus, and interrupted IVC with azygos continuation, our case is the first successful manual catheter ablation of persistent AF and common AFL conducted with SVC-only approach.

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