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CASE REPORT
Arrhythmias / electrophysiology

Radiofrequency ablation for paroxysmal atrial fibrillation in a patient with dextrocardia and interruption of the inferior vena cava: a case report

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Background Dextrocardia with interruption of the inferior vena cava (I-IVC) is a very rare anatomical variant. Catheter ablation of atrial fibrillation (AF) in patients with this anatomical variant is challenging for electrophysiologists. This case report presents a safe, effective, and radiation-free approach for high-power ablation of AF via a superior transseptal approach in patients with dextrocardia and I-IVC.

Case summary A 57-year-old man with paroxysmal AF with dextrocardia and I-IVC with azygos continuation was referred to our hospital for radiofrequency (RF) ablation. It was evident that transseptal puncture and pulmonary vein isolation (PVI) would be impossible using an IVC approach via the femoral vein. Therefore, we decided to perform left atrium (LA) ablation via the superior vena cava approach. A phased array intracardiac echocardiography (ICE) catheter was inserted in the right femoral vein. Three-dimensional (3D) anatomical reconstruction of LA, right atrium (RA), and coronary sinus (CS) ostium were performed using ICE with azygos vein and RA imaging. Navigation-enabled electrodes were inserted into annotated CS on cardiac 3D ICE image. The left internal jugular vein was accessed using an SL1 transseptal sheath and Brockenbrough needle. Transseptal puncture was performed under ICE with an RF-assisted approach. We accomplished ablation index guided high-power pulmonary vein isolation using a bi-directional guiding sheath with visualization capabilities and a surround flow contact force-sensing catheter. No complications occurred during or after the procedure.

Discussion With the application of multitude of newer technologies, we can accomplish safe, effective, and fluoroscopy-free RF ablation of AF using the superior approach in patients with complex anomaly.

Keywords Dextrocardia • Interruption of the inferior vena cava • Transseptal puncture • Atrial fibrillation • High-power ablation • Case report

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Learning points

- Catheter ablation of atrial fibrillation (AF) in patients with dextrocardia and interruption of the inferior vena cava (I-IVC) is challenging for electrophysiologists.
- A safe and fluoroscopy-free transseptal puncture can be successfully performed in patient with dextrocardia and I-IVC via radiofrequency (RF)-assisted approach from the jugular vein.
- With the application of multitude of newer technologies, we can accomplish safe, effective, and fluoroscopy-free RF ablation of AF from superior approach in patients with rare congenital anomaly.

Introduction

Dextrocardia is detected in ~1 in 20 000 in the general population and interruption of the inferior vena cava (I-IVC) is seen in 0.6% of patients with congenital heart disease.[1,2] Dextrocardia and I-IVC are both rare congenital malformations of the heart and their presence together can make catheter ablation for atrial fibrillation (AF) extremely challenging. Here we present a case of catheter ablation of AF in a patient with both dextrocardia and I-IVC under guidance of intracardiac echo without fluoroscopy. In this report, we describe in detail our safe, effective, and fluoroscopy-free approach to ablation of AF in a patient with both dextrocardia and I-IVC.

Timeline

Time	Events
Two years before	Patient has been diagnosed with atrial fibrillation (AF)
In the past 12 months	He had chest discomfort and palpitation with 2–3 times a month on average during tachycardia due to AF
One month before	Patient was found to have a rare congenital anomaly, consisting of dextrocardia and interrupted inferior vena cava
Hospitalization	Fluoroscopy-free radiofrequency ablation of AF was performed from superior approach in this patient. No complications occurred during or after the procedure
Six months post-ablation	At 6-month post-ablation, the patient was asymptomatic without recurrence of arrhythmia

Case presentation

A 57-year-old man with symptomatic paroxysmal AF was referred to our hospital for catheter ablation. The patient has no previous history of heart disease or other health problems. For the last 12 months, patient has been experiencing chest discomfort and palpitations (European Heart Rhythm Association Class 3) correlating well with episodes of AF with 5–10 min duration and 2–3 times a month on average. Coronary computed tomography (CT) angiography showed dextrocardia, but no coronary artery problems were found. He has previously been tried on metoprolol, propafenone, and amiodarone without significant improvement in his symptoms even with a high oral maintenance dose (metoprolol 100 mg twice daily, propafenone 200 mg q8h, and amiodarone 600 mg per day). Patient was referred to a local electrophysiology centre but was eventually referred to us due to the anticipated technical difficulties. Physical examination revealed apical impulse located on the right side of his chest. Transthoracic echocardiography revealed a slightly dilated left atrium (LA) with an anteroposterior diameter of 43 mm. Contrast-enhanced CT at our hospital demonstrated that his IVC was completely obstructed at the infrarenal level. Abdominal venous return occurred via the azygos vein draining into the superior vena cava (SVC) (*Figure 1*). Therefore, SVC approach was chosen for AF ablation.

After written informed consent was obtained, the procedure was performed under general anaesthesia with midazolam, fentanyl, and propofol. Ablation was performed using anticoagulation with a novel oral anticoagulant.

A phased array intracardiac echocardiography (ICE) catheter (10 Fr) was placed at the level of the LA in the azygos vein and mid-right atrium to create the left and right atrial anatomic map (*Figure 2*). The steerable, navigation-enabled multi-electrode mapping catheter (DECANAV, Biosense Webster) were successfully inserted into coronary sinus via right femoral vein cannulation without fluoroscopy guidance (*Figure 3*).

We manually curved a Brockenbrough needle (BRK, St. Jude Medical, St Paul, MN, USA) with a 120° angle and an 8 cm curve to manipulate the tip towards the fossa ovalis horizontally (*Figure 4A*). An 8.5 Fr transseptal long sheath (SL1, St. Jude Medical, Minneapolis, MN, USA) with a Brockenbrough needle (BRK, St. Jude Medical) via the right jugular vein was introduced at the posterior portion of the tricuspid annulus, maintaining the relative position of the sheath with the tip oriented in the 1 o'clock position from the operator's view. Next, we gently manipulated the SL1 sheath and needle with 60° clockwise rotation and a 2 cm pull-back (*Figure 4B*). Once correct positioning in the fossa ovalis of the sheath/needle assembly was confirmed using ICE (*Figure 4C*), the needle was advanced almost to the tip of the dilator without exposing the needle beyond the dilator. A SmartTouch Surround Flow (STSF) ablation catheter (ThermoCool, Biosense Webster) was brought into contact with the proximal extremity of the needle as its tip was advanced out of the dilator. At this point, unipolar radiofrequency (RF) energy of 30 W was transmitted from the ablation catheter to the needle, the tip of which being still in contact with the fossa ovalis (*Figure 4D*). The puncture of the septum occurred without further tenting of the septum. After successful transseptal puncture, we added heparin intravenously to an activated

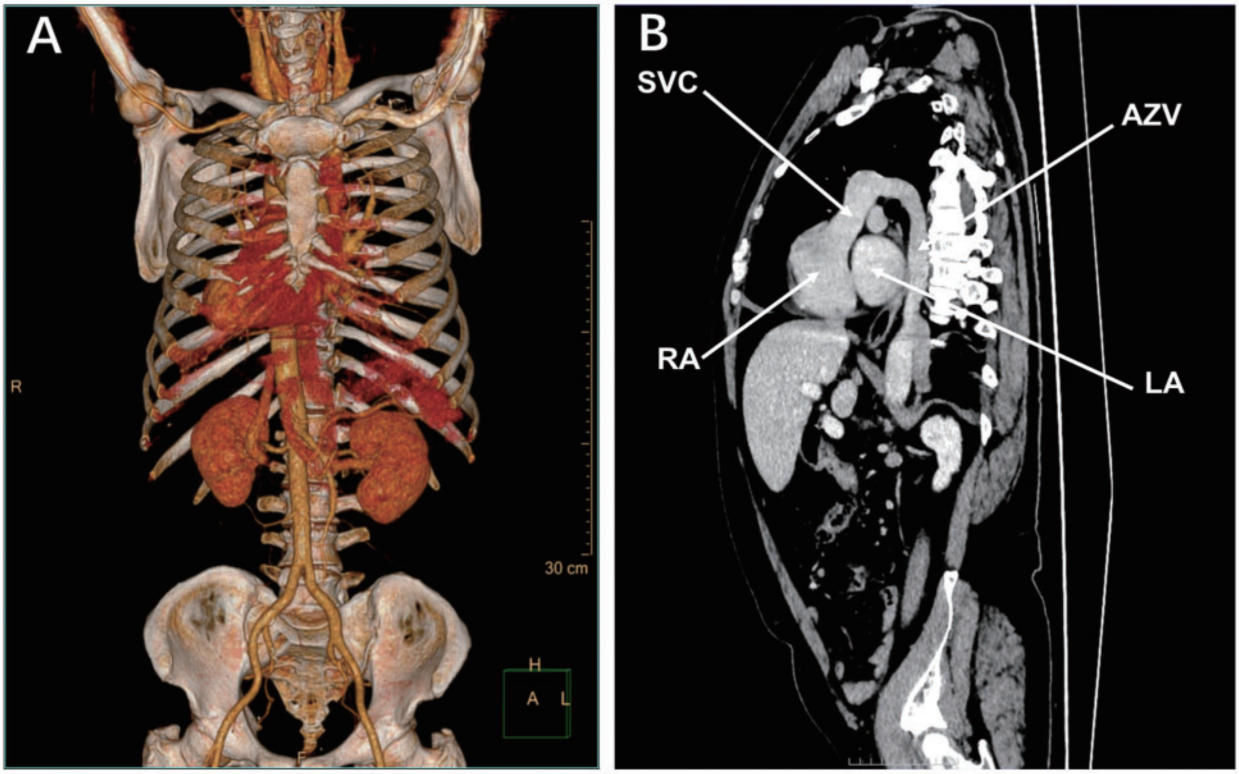

Figure 1 Three-dimensional reconstruction image of dextrocardia (*A*) and sagittal view of contrast-enhanced computed tomography presenting the atrium and veins (*B*). The inferior vena cava drains into the azygos vein and then to the superior vena cava. There was no additional connection between the inferior vena cava and right atrium. AZV, azygos vein; LA, left atrium; RA, right atrium; SVC, superior vena cava.

clotting time of 300–350 s throughout the procedure. Next, we replaced the first 8.5 Fr SL1 sheath via the right jugular vein using a VIZIGO bi-directional guiding sheath (Destino; Japan Lifeline Co., Tokyo, Japan) in the LA.

RF energy was delivered with STSF catheters in power-controlled mode with 35 W for the posterior wall and 40 W elsewhere. Ablation index (AI) targets were 350 for the posterior wall and 450 elsewhere. Left pulmonary veins (PVs) (common trunk) were accomplished via a direct approach by maintaining the sheath in the body of the LA and advancing the catheter to map and ablate different aspects of the PVs (*Figure 5A*). We accomplished right PVs by creating a large loop with the sheath and catheter (*Figure 5B*). Afterwards, to check the PV isolation, the STSF catheter was exchanged for a Lasso catheter (Biosense Webster). As there were good contact force achieved and high power guided by AI during procedure, all PVs electrical isolation were achieved. The procedure time was 89 min and RF delivery time was 15 min.

At 6-month post-ablation, the patient was asymptomatic without recurrence of arrhythmia.

Discussion

To the best of our knowledge, there has been no published case of PVI performed manually via superior transseptal access in patients with dextrocardia and I-IVC. The presence of both dextrocardia and I-IVC is rare and pose significant technical challenges to AF ablation.

In patients such as those with congenital or acquired I-IVC, LA transseptal catheterization is not feasible via the femoral veins. Additional options for patients with I-IVC include surgical AF ablation, ablation via a retrograde aortic approach, transhepatic access, or transseptal access using a superior approach. Surgical epicardial ablation approach is effective but remains a more invasive technique than percutaneous catheter ablation and may be associated with increased complication rates and hospitalization time. Transhepatic access allows for an inferior approach, which is more familiar to electrophysiologists and allows for greater degrees of manoeuverability and catheter manipulation.[3] However, after consulting radiologists regarding the CT for our patient, this approach was deemed risky with regard to risk of hepatic haemorrhage and the narrow calibre of the hepatic vein. Additionally, transhepatic access requires more effective haemostasis measures and should only be performed in a centre familiar with the approach.[4] Hence, our interventional radiologists discouraged this approach. Another alternative is the retrograde aortic approach. Okajima *et al.*[5] reported the technique of trans-aortic pulmonary vein isolation using the magnetic navigation system in a patient with dextrocardia, situs inversus, and IVC continuity with the azygos vein. Considering our case, with the lack of a magnetic navigation system in our hospital and the technical difficulty of

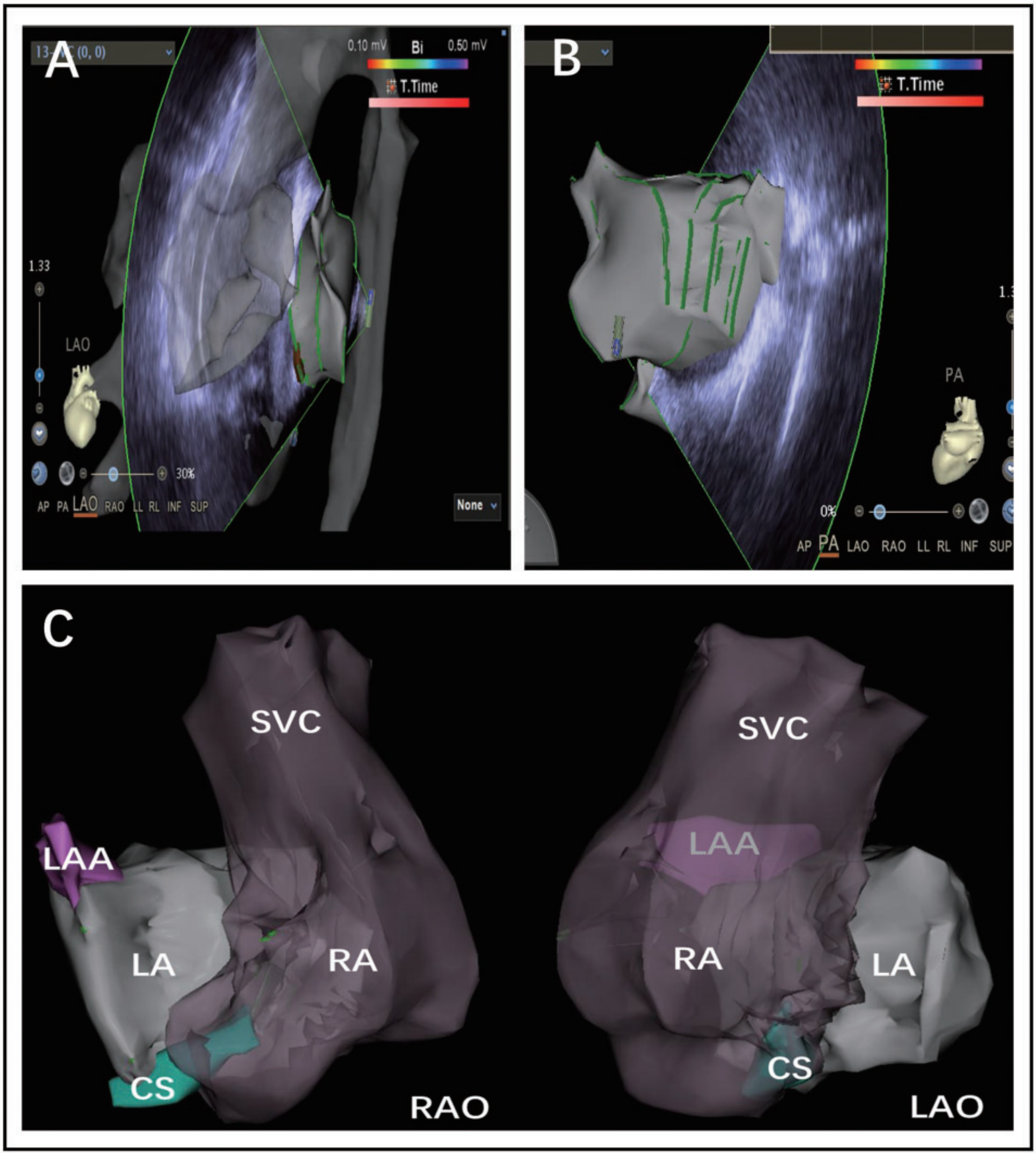

Figure 2 Reconstruction of the left and right atrial anatomic map. Three-dimensional anatomical reconstruction of the left atrium with azygos vein (*A*) and right atrium (*B*) imaging using intracardiac echocardiography. (*C*) Three-dimensional anatomical reconstruction of left atrium, right atrium, left atrial appendage, superior vena cava, and coronary sinus ostium. CS, coronary sinus; LA, left atrium; LAA, left atrial appendage; RA, right atrium; SVC, superior vena cava.

placing continuous linear lesions to permanently isolate the PVs this approach was ruled out much earlier. Taking into consideration these factors, AF ablation via the superior approach was considered optimal in our case.

An RF-assisted transseptal approach has been shown to perform transseptal puncture safely and successfully without the need for significant mechanical force.[6,7] The main advantage of RF-assisted transseptal approach is negation of mechanical pressure potentially

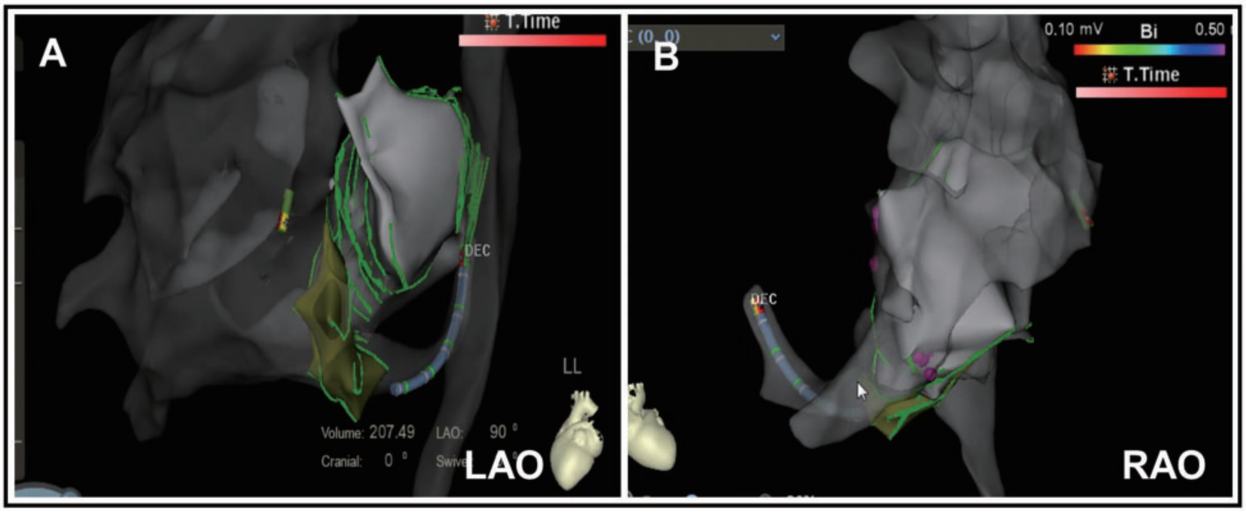

Figure 3 Deflectable DECANAV electrodes were successfully inserted into coronary sinus via right femoral vein cannulation without fluoroscopy. (*A*) Left anterior oblique view. (*B*) Right anterior oblique. LAO, left anterior oblique; RAO, right anterior oblique.

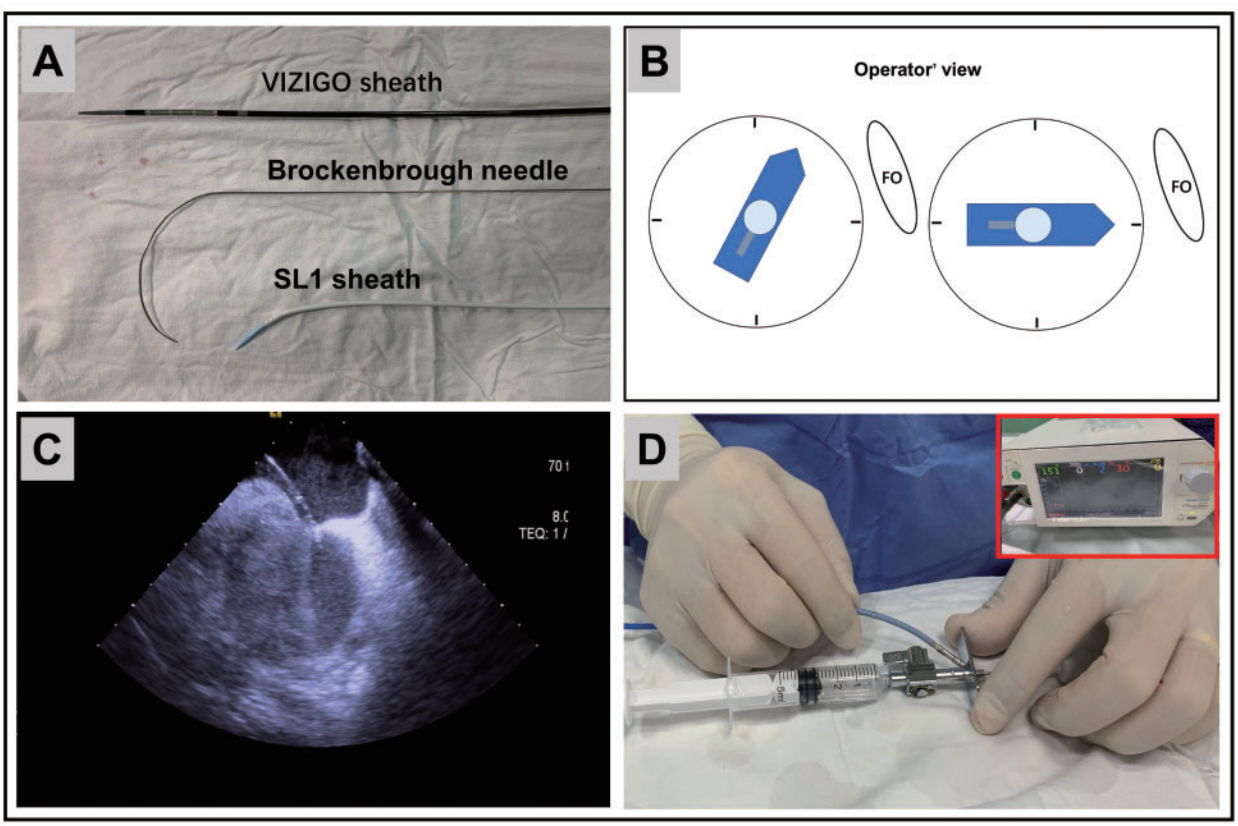

Figure 4 Transseptal puncture procedures from the superior approach in dextrocardia. (*A*) The Brockenbrough needle was manually curved with a 120° angle and 8 cm curve. (*B*) The SL1 sheath and needle were located at 1 o'clock from the operator's view. We manipulated the SL1 sheath and needle with 60° clockwise rotation and 2 cm pull-back, gradually changing the relative position of the sheath with the tip oriented to the 3 o'clock position. (*C*) Tenting of the interatrial septum by the sheath-dilator-needle was confirmed by intracardiac echocardiography. (*D*) The SmartTouch Surround Flow ablation catheter was brought into contact with the proximal extremity of the needle, the tip of which was still in contact with the fossa ovalis. Unipolar radiofrequency energy of 30 W was then transmitted from the ablation catheter to the needle by simple contact. FO, fossa ovalis.

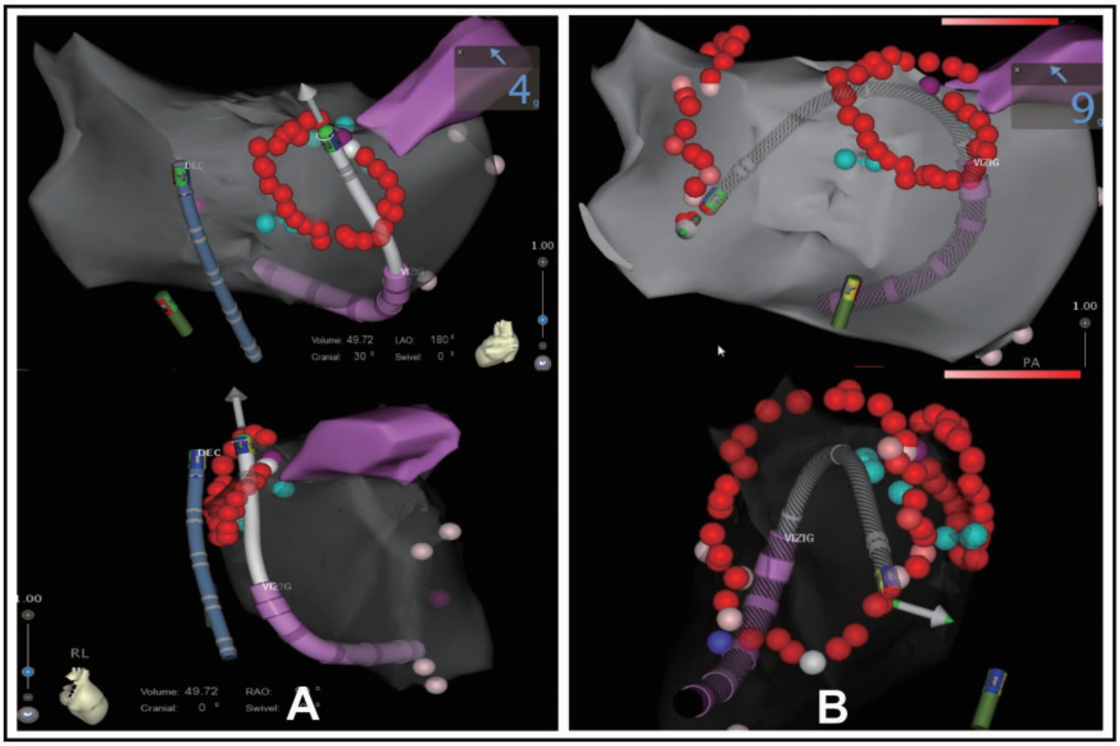

Figure 5 The approach used to map and ablate the left pulmonary veins (*A*) and the right pulmonary veins (*B*).

preventing excessive needle movement and inadvertent injuries to surrounding structures. Use of RF-assisted transseptal approach for various procedures has been consistently shown to result in shorter time to transseptal LA access and shorter fluoroscopy duration.[8,9] Because the abrupt leftward movement (snap-in), which indicates passage over the limbus into the fossa ovalis, cannot be observed from the SVC approach, real-time echocardiographic imaging for tenting of the fossa ovalis enhanced the safety of the transseptal access into the LA. We believe that the use of RF-assisted transseptal approach and ICE make transseptal puncture safer and more effective when using the SVC approach in complex case.

The introduction of contact force-sensing catheters and use of force-time integral targeted ablation has improved results, but not to the levels hoped for, with still more than one-third of patients exhibiting one or more gaps.[10] AI is a novel marker incorporating contact force, time, and power in a weighted formula.[11] High-powered ablation can be safely delivered with STSF catheters when guided by AI. This ablation strategy was associated with lower complication rates, more localized and durable RF ablation lesions, and reduced procedural and fluoroscopy time.[12] In addition, VIZIGO sheath enables real-time steerable sheath visualization on the electroanatomical map during a procedure without depending solely on fluoroscopy. This allows for quicker isolation of the pulmonary veins because of the decreased catheter manipulation time and better contact force.

Catheter manipulation via the superior approach is associated with higher radiation exposure. Even with a lack of experience, the fluoroscopy and procedure time in our case was much shorter than reported previously, and may be further shortened with increased procedural familiarity. Potentially, with the application of multitude of newer technologies, we can accomplish safe, effective, and fluoroscopy-free RF ablation of AF from the superior approach in patients with rare congenital anomalies.

Conclusion

A multitude of newer technologies can be readily implemented in the modern electrophysiology laboratory to accomplish safe, effective, and fluoroscopy-free ablation of AF in a complex case despite significant technical difficulties.

Lead author biography

Xiaofeng Hu was born in Zhejiang, China, in 1986. He received the MD degree from Shanghai Jiao Tong University School of Medicine in 2014. Since 2014, he began to work and receive interventional cardiology training at the Shanghai Chest Hospital. His clinical and research interests focus on catheter ablation of cardiac arrhythmias including atrial fibrillation, supraventricular tachycardia, and ventricular tachycardia.

Supplementary material

Supplementary material is available at *European Heart Journal - Case Reports* online.

Slide sets: A fully edited slide set detailing this case and suitable for local presentation is available online as Supplementary data.

Consent: The authors confirm that written consent for submission and publication of this case report including images and associated text has been obtained from the patient in line with COPE guidance.

Conflict of interest: None declared.

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