Trans–coronary sinus puncture for catheter ablation and left atrial appendage closure device implantation in a patient with dextrocardia and persistent right superior vena cava

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Adi Lador, MD, Apoor Patel, MD, Miguel Valderrábano, MD

From the Department of Cardiology, Houston Methodist DeBakey Heart and Vascular Center, Houston Methodist Research Institute, Houston Methodist Hospital, Houston, Texas.

Introduction

Dextrocardia is a rare congenital condition in which the apex of the heart is located on the right side of the body. Transseptal puncture and catheter ablation in the left atrium in patients with dextrocardia may be challenging. Dextrocardia with situs solitus, in which the viscera are in the normal position, is often associated with additional congenital anomalies. We describe a case of a patient with dextrocardia, situs solitus, persistent right superior vena cava (SVC) with atrial fibrillation, atrial tachycardia, and gastrointestinal bleeding who presented for catheter ablation and left atrial appendage closure. We performed transseptal access from a femoral approach via puncture of the coronary sinus (CS) directly into the left atrium.

Case report

A 77-year-old woman with a history of hypertension, dextrocardia, achalasia, nonischemic cardiomyopathy with an ejection fraction of 45%, and paroxysmal atrial fibrillation presented for ablation. She had atrial fibrillation diagnosed 2 years prior and had been initiated on amiodarone. Despite medical therapy, the frequency of her episodes had increased, prompting emergency room visits and hospitalizations. She was thus offered catheter ablation.

Given her history of dextrocardia, a chest and an abdominal computed tomography (CT) were performed to delineate her anatomy. A CT of her chest showed a duplicated SVC. Her left SVC drained into her right atrium. Her right SVC drained into her dilated CS. Notably, the inferior vena cava

> ## KEY TEACHING POINTS
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> - Trans–coronary sinus puncture may be of utility in patients in whom conventional transseptal puncture is difficult or not feasible.
> - Trans–coronary sinus puncture to the left atrium can be performed safely.
> - In patients with complex anatomy, preprocedural planning and gathering information of the anatomical details from multimodality imaging (computed tomography, intracardiac echography, and 3D mapping system) is vital.

(IVC) drained into the CS and not into the right atrium. The left atrium was 4.5 cm in diameter. She did not have situs inversus.

After careful review of her CT images, we considered performing transseptal access from a femoral approach via puncture of the CS directly into the left atrium. Other approaches were considered, such as a superior approach from her left internal jugular vein or a femoral approach from the fossa ovalis into her right atrium. Based on the CT images it was felt that the transseptal puncture directly from the fossa ovalis could be challenging, as the sheath would have to traverse the angle between the IVC and the CS, and then the CS into the right atrium.

During the procedure a fast anatomical map was made of her IVC, right atrium, CS, and both SVCs using a Pentaray catheter (Biosense Webster, Irvine, CA). This was integrated with ultrasound images from an intracardiac echocardiography (ICE) (SoundStar; Biosense Webster) catheter as well as the preprocedural CT (Figure 1A and B). The fossa ovalis was located on ICE, but as there was significant tortuosity reaching it from the IVC, we chose to perform the transseptal puncture from the dilated CS-persistent right SVC, more similar to a conventional puncture (right to left).
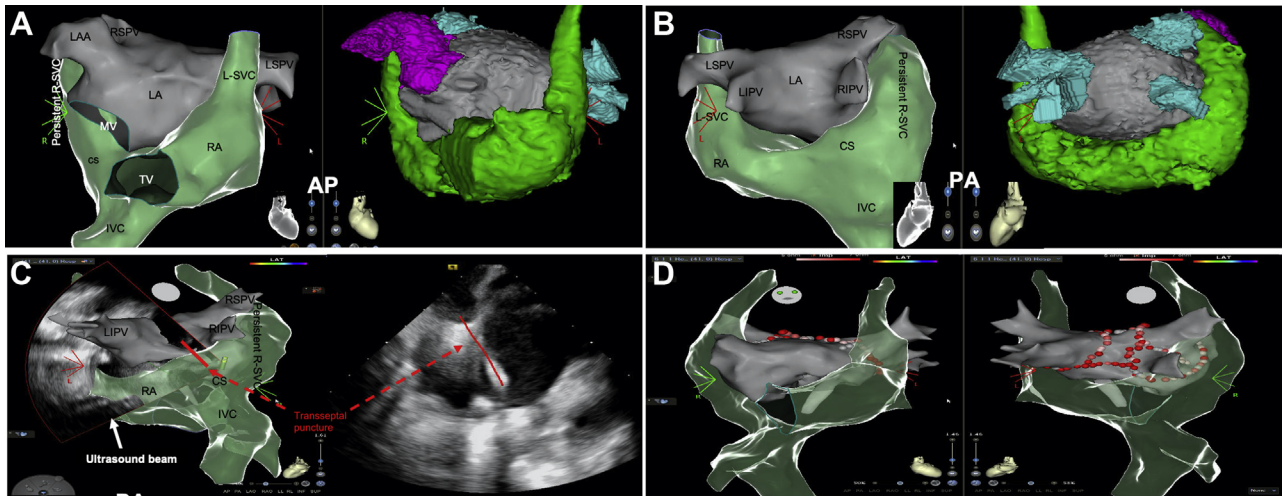
Figure 1   First ablation case: atrial fibrillation ablation. **A, B:** Patient's anatomy. CARTO (Biosense Webster, Irvine, CA) fast anatomical map (FAM) and computed tomography images in anterior posterior (AP) and posterior anterior (PA) projections. The patient has a duplicated superior vena cava (SVC). Her left SVC (L-SVC) drained into her right atrium (RA). Her right SVC drained into her dilated coronary sinus (CS). The inferior vena cava (IVC) drained into the CS and not into the RA. The left atrial appendage (LAA) is close to the right pulmonary veins. **C:** Transseptal puncture from the roof of the CS to the left atrium (LA). Carto FAM with ultrasound beam in PA projection and intracardiac echocardiography image. **D:** Ablation lesions: pulmonary vein isolation and posterior wall isolation. LIPV = Left inferior pulmonary vein; RIPV = right inferior pulmonary vein; RSPV = right superior pulmonary vein; R-SVC = right superior vena cava; TV = tricuspid valve.

The CS-to-left-atrium puncture was performed under ICE guidance without the use of fluoroscopy (Figure 1C). The ICE catheter was positioned on the floor of the CS with the imaging plane facing upward toward the left atrium. A Baylis needle (Baylis, Toronto, Canada) was then advanced into the Agilis sheath (Abbott, St Paul, MN), which was rotated up to the roof of the CS. The sheath was then advanced until tenting of the CS roof and inferior left atrium was seen. The needle was then advanced and a single transseptal puncture was performed. Catheter ablation was then performed using a Biosense Smart-Touch ablation catheter (Biosense Webster). The pulmonary veins (PVs) and posterior wall were isolated, and the patient was discharged the next day with no complications (Figure 1D).

On follow-up, the patient presented with severe gastrointestinal bleeding requiring blood transfusions. Endoscopy showed a cecal Dieulafoy lesion oozing and the patient underwent a hemoclip and embolization by angiography. She was felt to be at high risk of recurrent bleeding. In addition, the patient also had recurrent episodes of atrial tachycardia requiring recurrent cardioversions and admissions.

Therefore, the patient was brought to the electrophysiology laboratory to undergo catheter ablation and Watchman (Boston Scientific, St. Paul, MN) left atrial appendage (LAA) occlusion. Single trans-CS puncture via a femoral approach was again performed, guided by ICE, CT, and the CARTO fast anatomical map (Biosense Webster). The transseptal puncture had to be optimized to reach the LAA. After detailed ICE imaging from the right atrium and persistent right SVC, an imaging plane was found that showed the enlarged CS, left atrium, and LAA. This location was chosen for CS (transseptal) puncture, in order to align the Watchman delivery sheath with the LAA and facilitate Watchman deployment (Figure 2A and B). To maintain stability in the

chosen location for puncture, we avoided using a transseptal needle and instead used cautery applied to the back end of an 0.035" wire. Of note, the transseptal puncture during the second procedure was performed at a different location than the one in the first procedure (Figures 1 and 2).

The patient was found to have multiple focal atrial tachycardias. The sites of earliest activation were found to be in the left SVC–right atrium (RA) junction, in the RA anterior to the right atrial appendage and the anterior wall of the left atrium. We encircled the left SVC in the left SVC–RA junction, isolated the LAA, and did linear ablation from the LAA to the left PVs, from the left PVs to the mitral annulus, and a third line from the LAA to the mitral annulus. These lesion sets terminated the tachycardia and further tachycardia was not induced (Figure 2C).

Watchman implantation was then performed with the aid of fluoroscopy and ICE. The transseptal sheath performed for mapping and ablation was exchanged over the wire and a Watchman delivery sheath was inserted. A pigtail catheter was used to engage the LAA. A 24-mm device was then delivered and on the first delivery was found to have adequate compression, position, stability, and seal as measured by ICE with Doppler. Transesophageal echocardiography was not used (Figure 3).

On follow-up, the patient was in sinus rhythm and had a transesophageal echocardiogram with appropriate device placement and seal and no obvious residual communication between the CS and the left atrium; however, image quality was limited.

Discussion

Dextrocardia is a rare congenital condition in which the apex of the heart is located on the right side of the body. Situs
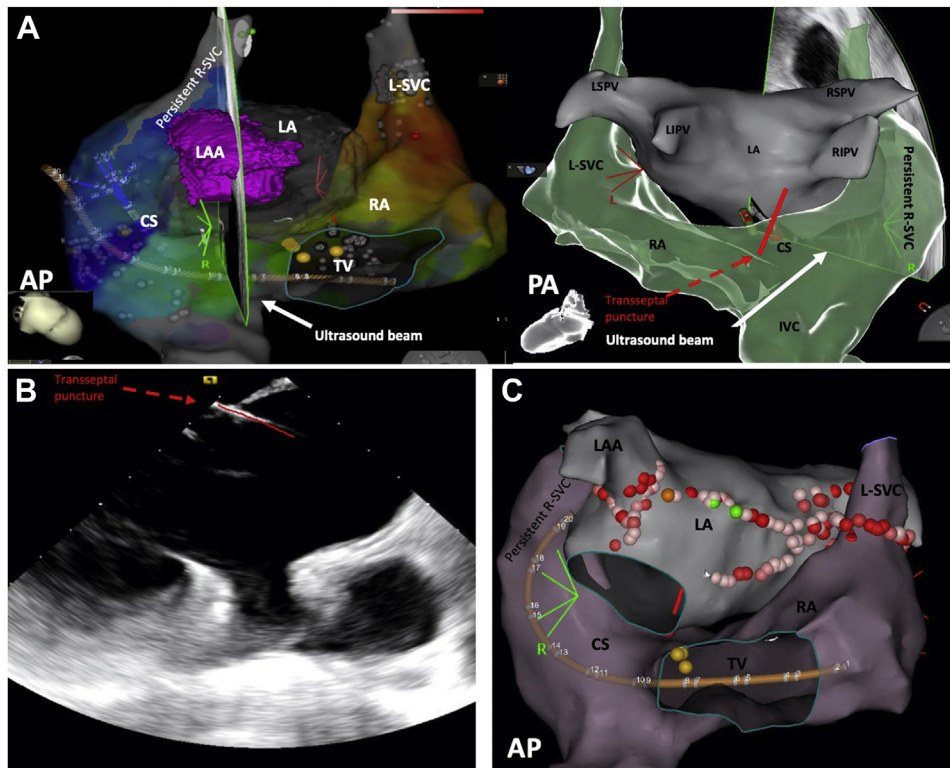
Figure 2   Second case: atrial tachycardia ablation. **A:** Transseptal puncture location on CARTO (Biosense Webster, Irvine, CA) fast anatomical map (FAM) with ultrasound beam. We performed trans–coronary sinus (CS) puncture. The puncture had to be optimized to reach the left atrial appendage (LAA) in order to align the Watchman delivery sheath (Boston Scientific, St. Paul, MN) with the LAA later in the procedure. Anterior posterior (AP) and posterior anterior (PA) projections. **B:** Intracardiac echocardiography image with transseptal location, opposite to the LAA location. **C:** We encircled the left superior vena cava (L-SVC) in the L-SVC–right atrium (RA) junction, isolated the LAA, and did linear ablation from the LAA to the left pulmonary veins (PVs), from the left PVs to the mitral annulus, and a third line from the LAA to the mitral annulus. These lesion sets terminated the tachycardia and further tachycardia was not induced. IVC = inferior vena cava; LA = left atrium; LIPV = left inferior pulmonary vein; LSPV = left superior pulmonary vein; RIPV = right inferior pulmonary vein; RSPV = right superior pulmonary vein; R-SVC = right superior vena cava; TV = tricuspid valve.

solitus describes viscera that are in the normal position, with the stomach on the left side; in situs inversus, the positions of the abdominal organs and viscera are reversed. Dextrocardia with situs solitus occurs in an estimated 7500–29,000 living people worldwide. Dextrocardia with situs solitus is often accompanied by additional cardiovascular abnormalities, notably atrioventricular discordance, single ventricle, atrial

or ventricular septal defect, anomalous pulmonary venous return, or transposition of the great arteries.[1]

Tripathi and colleagues[2] found in a retrospective study that situs solitus is present in about 43.1%, situs inversus in 38.1%, and situs ambiguous in 18.8% of patients with dextrocardia. Persistence of the second SVC occurs most often as an isolated persistent left SVC. This anomaly has an estimated
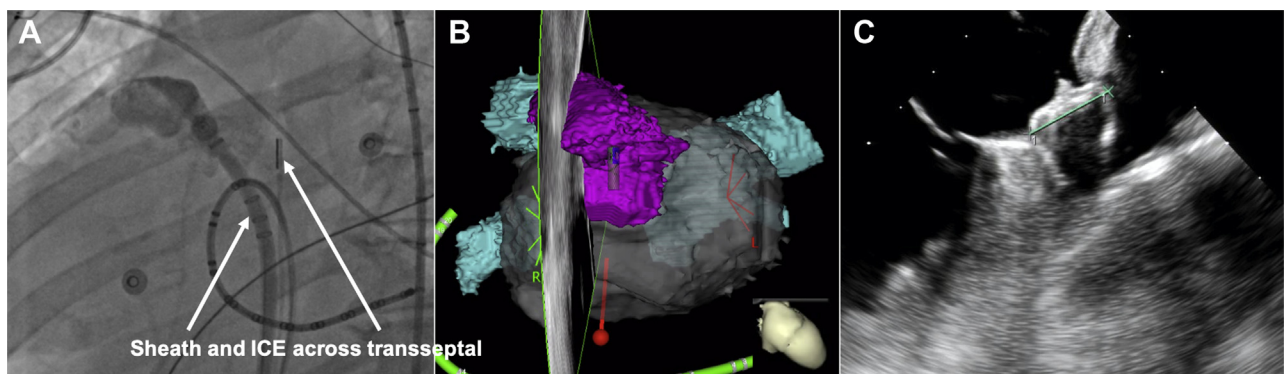

Figure 3   Second case: left atrial appendage closure with the Watchman device (Boston Scientific, St. Paul, MN). **A:** Fluoroscopy with final location of the Watchman device. **B:** CARTO (Biosense Webster, Irvine, CA) 3D map illustrates the transseptal location opposite to the left atrial appendage location. **C:** Intracardiac echocardiography (ICE) image with final Watchman location.

prevalence in the general population of 0.1%–0.4%.[3] The combination of dextrocardia with persistence of a second (right) SVC has been described.[4]

In patients with dextrocardia, knowledge of the anatomical details is vital because it allows the electrophysiologist to plan the technical approach. Thus, we ordered a chest and abdomen CT and integrated the images with ultrasound images from ICE and the 3D mapping system.

Transseptal puncture permits a direct route to the left atrium via the systemic venous system and intra-atrial septum. Access to the left atrium is required for numerous electrophysiology procedures, including ablation of atrial tachycardia, atrial fibrillation, accessory pathways, ventricular tachycardia, and LAA closure device.

Alternative approaches for the standard transseptal approach may be needed in case of anatomical variants. A superior approach through the SVC has been well described.[5–7] Transseptal access with a superior approach may be challenging because of the lack of sheath support when positioning and puncturing through the fossa ovalis. Additionally, the lack of support from the lower rim of the fossa ovalis may increase the difficulty of performing ablation adjacent to the left inferior PV,[8] and the tight curve required to ablate the right-side PVs also increases the difficulty of ablation in the antrum of the right-side PVs. Another alternative is the retrograde aortic approach.[9] Percutaneous transhepatic access was also described as a feasible alternative when the IVC is interrupted.[10]

To our knowledge, the puncture of the CS into the left atrium has not been described before. In our case we chose this approach after carefully reviewing the CT before the procedure. To perform a transseptal puncture from the fossa ovalis, the transseptal sheath and needle would have to traverse the angle between the IVC and the CS, and then the CS into the right atrium. Given this, it was decided in advance to directly puncture the roof of the CS, into the floor of the left atrium. We also altered the location of CS puncture based on whether we were performing PV isolation or appendage occlusion. When performing appendage occlusion, we ensured that the ICE imaging plane for CS puncture was aligned with the appendage.

The potential complications of trans-CS puncture are hemopericardium if the needle traverses the pericardial space in a region that is not directly opposed by the left atrium. In our case both punctures were uneventful and were closely guided by ICE. Although CS puncture may leave a residual communication between the CS and the left atrium, this would not be expected to be of any more hemodynamic significance than the traditional communication between the left and right atrium in the fossa ovalis. CS puncture may be of utility in patients in whom conventional transseptal puncture is difficult or not feasible—for instance, patients with a thick septum, severe lipomatous hypertrophy, or status post atrial septal defect closure device. It may also be of utility in other interventional cardiac procedures, and we have performed this in 2 patients who presented for mitral valve paravalvular leak closure where the inferior location of the leak was not

reachable via transseptal puncture through the fossa ovalis (unpublished data). In our case the CS was dilated, which facilitated trans-CS puncture. In a normal-size CS catheter manipulation may be more challenging, but from our experience CS-to–left atrium puncture is still possible.

Only 1 case has been described before of LAA closure in a patient with dextrocardia.[11] In that case Castriota and colleagues used transesophageal echocardiography and "reverse" fluoroscopic images to simplify anatomic guidance for the positioning of an Amulet device. Technical challenges in addition to left atrial access include that conventional sheaths, in our case the double curve, are not properly oriented to allow deployment in the anterior aspect of the appendage in a rotated heart. However, this was not a barrier to implantation in this particular case. Use of multimodal imaging including ICE, CT integration, and fluoroscopy was integral to successful deployment.

Conclusion

To our knowledge, this is the first reported demonstration of the safety and feasibility of trans-CS-to–left atrial puncture, catheter ablation, and Watchman delivery procedure in a patient with dextrocardia.

Acknowledgments

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