Atrial fibrillation ablation in a patient complicated by persistent left superior vena cava and absent right superior vena cava

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A 69-year-old man was referred to our hospital with a previously diagnosed drug-refractory paroxysmal atrial fibrillation (AF).

The delayed computed tomography angiography (CTA) prior to ablation revealed the presence of the huge persistent left superior vena cava (PLSVC) and the absence of right superior vena cava (SVC) (Figure 1). However, it was difficult to delineate the anatomy of right atrium (RA) and RA appendage (RAA) in detail.

ENSITE (Abbott) was used as the three-dimensional mapping system. Atrial septal puncture was difficult because it did not come with an anatomical orientation. Intracardiac echocardiography was helpful to achieve this puncture (Figure 2A). We performed the pulmonary vein isolation and the cavotricuspid isthmus ablation (Figure 2B). Then, we performed the three-dimensional mapping of RA, RAA, and PLSVC during sinus rhythm. Three-dimensional mapping during the sinus rhythm (Figure 3) depicted the location of sinus node, and the location of RA could be identified. The body of RA was looking like hypoplastic and RAA hyperplastic and extended cephalad, mimicking a remnant of SVC. During the procedure, we could not find any firing from PLSVC, we did not isolate PLSVC.

We reported on the characteristics of RA and RAA in a patient with AF complicated by PLSVC and absent right SVC. The main findings of this case are (1) the difficulty in delineating RAA on CTA, (2) the three-dimensional mapping depicted hyperplasia of RAA and hypoplasia of RA body, and (3) the difficulty of atrial septal puncture in a patient complicated by PLSVC and absent SVC.

PLSVC is the most frequent venous anomaly of the thorax, found 0.5%–2% of the general population. PLSVC with absent right SVC, which is also referred to as isolated PLSVC, is quite rare, and occurs in 0.09%–0.13% of patients who have other cardiac malformations.

The three-dimensional mapping system was able to depict the anatomy of RA and RAA that could not be depicted by CTA. The postcontrast-delayed CTA before AF ablation was performed to look at the morphology of the left atrium and pulmonary veins, and assess the presence or absence of thrombus in the left atrial appendage. However, the contrast-enhanced effect on RA and RAA is not sufficient at this timing of delayed LAA imaging. We believe that this is one reason for the poor delineation of RAA.

Three-dimensional mapping during sinus rhythm (Figure 2) allowed for easy identification of the location of the sinus node, and provided information regarding the anatomical location of RA and RAA. One reason for this hyperplasia of RAA might be that RAA had a pumping function, so that the blood flow returning from the whole body to RA could be easily delivered to the right ventricle. Figure 3 showed a comparison between the three-dimensional contact mapping created by the Ensite system and that created by contrast-enhanced CTA. RAA was not well delineated on CTA.

The standard transseptal procedure is following: advance the sheath and the dilator into SVC, position Brockenbrough needle inside assembly, adjust the needle pointer flange between 3 and 5 o’clock as viewed from foot end of the patient, drag assembly, and engage fossa ovalis. We advanced the sheath cephalad, but could not quite get it to point in the 3–5 o’clock direction. This was because the sheath was advanced into RAA, not in the direction of the remnant of SVC. This made it difficult to puncture the atrial septum. Because we performed the atrial septal puncture with intracardiac echocardiography, we were eventually able to perform the puncture without complications. We should mention the difficulty of atrial septal puncture in such cases.
FIGURE 1  The delayed computed tomography angiography prior to ablation. (A–D) were axial views of CT and (E, F) were three-dimensional images. Axial views revealed the presence of the huge persistent left supra vena cava and the absence of right supra vena cava. Open center asterisk showed right atrium (RA). Clean delineation of the body of RA and RA appendage from CT was difficult. A-Ao, ascending aorta; CA, coronary artery; CS, coronary sinus; D-Ao, descending aorta; LA, left atrium; LAA, left atrial appendage; LIPV, left interior pulmonary vein; LV, left ventricle; PA, pulmonary artery; RV, right ventricle

FIGURE 2  (A) Image of atrial transseptal puncture. The tenting image (red arrow) of the fossa ovalis was confirmed by intracardiac echocardiography, which facilitated the atrial septal puncture. (B) Image of catheters’ position during cavotricuspid isthmus linear ablation. Since there was no superior vena cava, a catheter of the coronary sinus was placed via the inferior vena cava. CS, coronary sinus; Eso, esophagus; LA, left atrium; LSPV, left superior pulmonary vein; needle, brockenbrough needle; RA, right atrium; RF, radiofrequency ablation catheter; TA, tricuspid annulus
To our knowledge, this is the first report about the three-dimensional mapping of RA, RAA, and coronary sinus in a patient with PLSVC and absent right SVC. In patients with these complications, other cardiac malformations are often present. Since the anatomy of cardiac malformations may not be clear on CT, the combined use of three-dimensional mapping might be important not only for understanding conduction in the heart but also for understanding detailed anatomy.

CONFLICT OF INTEREST
No conflict of interest.

FIGURE 3 A comparison between the image of three-dimensional contact mapping created by the Ensite system (left panels) and that created by contrast-enhanced computed tomography angiography (CTA) (right panels). Right atrial appendage was not well delineated on CTA. Three-dimensional mapping during sinus rhythm was easy to identify the location of the sinus node (left upper panel) and provide information regarding the anatomical location of right atrium (RA) body and RA appendage. PLSVC, persistent left supra vena cava; RA, right atrium; RAA, RA appendage; SN, sinus node; TV, tricuspid valve

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