Successful atrioventricular junction ablation in a patient with situs inversus with dextrocardia and complex venous anatomy

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Introduction

Situs inversus with dextrocardia is an uncommon condition and is present in approximately 0.02% of the general population. Infrahepatic interruption of the inferior vena cava (IVC) is a congenital anomaly found in 1.3%–3.0% of patients with congenital heart disease, and it occurs in 8–18% of patients with dextrocardia. In this condition, the IVC is interrupted above the renal branches, and systemic venous flow from below the interruption usually drains into the superior vena cava (SVC) via an enlarged azygos vein. Radiofrequency ablation of the atrioventricular node (AVN) is the most definitive rate-control strategy for patients with refractory atrial fibrillation. Although there are prior reports of radiofrequency ablation of cardiac arrhythmias via the anomalous IVC and azygos continuation, to the best of our knowledge this is the first case report of successful catheter ablation of the AVN in patients with dextrocardia, situs inversus, persistent bilateral SVC, and abnormal IVC.

Case report

The patient is an 82-year-old male with situs inversus with dextrocardia, persistent atrial fibrillation (AF), bioprosthetic aortic-valve replacement for severe aortic stenosis, prior MAZE procedure, and a dual-chamber pacemaker implanted via the persistent right SVC for sinus-node dysfunction. He presented with highly symptomatic AF with rapid ventricular response refractory to medical therapy and was referred for atrioventricular node junction (AVJ) ablation.

Standard electrophysiological intervals were measured at baseline. Given the dextrocardia, surface electrocardiographic leads, using inverted I (i. I), inverted II (i. II), RV1, and RV3 were displayed and recorded by the electrophysiological laboratory recording system (Prucka CardioLab IT system, GE Healthcare, Little Chalfont, UK). A true left anterior oblique view was obtained by positioning the fluoroscopy arm in a right anterior oblique position, and vice versa. Fluoroscopic images of the pacemaker lead and the bioprosthetic valve were obtained to define the anatomic location of the septum (Figure 1).

Bilateral femoral venous access was obtained, and a hydrophilic guidewire was advanced from both groins. The wires were initially inserted into the dilated azygos vein, which terminated in the distal hepatic vein below the level of the diaphragm, suggestive of an interrupted IVC. An 81-cm long SL-1 venous sheath was advanced to the azygous vein, and results of the venography performed demonstrated a direct azygous connection to the right persistent SVC (Figure 1A). Further venography via a pigtail catheter inserted through the SL-1 sheath confirmed the presence of a persistent left SVC with a patent communication into the right atrium (Figure 1B). The hydrophilic guidewire was navigated into the right ventricle to facilitate positioning of the SL-1 sheath into the right ventricle. A large curve ablation catheter (Chilli II, Boston Scientific, Marlborough, MA) was advanced through the SL-1 sheath toward the septum, which was identified by the fluoroscopic landmarks outlining the bioprosthetic aortic-valve replacement. An electroanatomic map of the right side of the heart and its great venous vessels was rendered using the EnSite NavX mapping system (St. Jude Medical, St. Paul, MN). The His bundle potential was recorded on the distal tip of the ablation catheter, as shown in Figure 2. Radiofrequency ablation at this location achieved transient junctional rhythm and
subsequent complete heart block heralded by the onset of right ventricular pacing from his preexisting dual-chamber pacemaker (Figure 2A–C).

Because no preprocedure imaging was performed, a computed tomography angiogram was performed postablation, and its results confirmed the presence of a right persistent SVC draining directly into the dilated coronary sinus, a left persistent SVC communicating with the right atrium, and an azygous vein entering the right SVC prior to connecting with the coronary sinus (Figure 3A and B). There were no procedural complications, and the patient was rendered pacer dependent, with amelioration of his AF symptoms.

Discussion

There have been reports of successful catheter ablation for supraventricular tachycardia in patients with dextrocardia and situs inversus.3,4 Ablations in patients with dextrocardia without sinus inversus or anomalous IVC have also been reported.5–7 Among those, only 3 cases had anomalous IVC.4,8,9 To our knowledge, this is the first clinical case report of successful AVJ ablation in a patient with dextrocardia, sinus inversus, anomalous IVC, and bilateral persistent SVC.

Venous access via the jugular or subclavian vein may be considered in patients with an interrupted IVC. However, in our case, there were chronic pacing wires through a persistent right SVC, and therefore a right internal jugular approach was not feasible. Taniguchi et al8 reported that in a patient with dextrocardia with complete situs inversus, an accessory connection between the coronary sinus and left ventricle was achieved with a right cubital venous approach. Although access via the left subclavian vein has been previously described10 and could have been considered, the advantage of a femoral venous approach is the ability to

| KEY TEACHING POINTS |
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| - This article is the first case report of successful catheter ablation of the AV node in patients with dextrocardia, situs inversus, persistent bilateral superior vena cava (SVC), and abnormal inferior vena cava (IVC) |
| - In the presence of these anomalies, access to the right cardiac chambers from the femoral approach via the azygos vein and persistent SVC is challenging, as a result of the longer course and the numerous angulations of the azygos venous system communicating with the SVC, but feasible. |
| - Preprocedural imaging to define anatomic variations may help with ablation success. |

Figure 1.  A: Results of the venography performed via a long sheath inserted from the femoral venous access. The venogram suggested an azygous connection to a persistent right superior vena cava (SVC) with communication to the persistent left SVC. B: A second venogram performed through a pigtail catheter inserted into the persistent left SVC confirming a connection to right atrium (RA).
Figure 2. Fluoroscopic position of the ablator and signal at the site of successful atrioventricular node junction ablation. A: Left anterior oblique (LAO) view, 40°: Note that in this patient with dextrocardia, the LAO view represents a true right anterior oblique (RAO) view and highlights the anterior location of the RA lead inserted through the persistent right superior vena cava (SVC) system. The right ventricular (RV) lead tip is directed septally toward the bioprosthetic AV (septum located near RV curve). The ablation catheter tip is oriented parallel to the RV lead tip, toward the septum. B: RAO view, 40°: Similarly, this image represents the true LAO view. Note the RA lead is directed toward the lateral RA wall and the RV lead points toward the atrioventricular node junction, which is located septally. C: Distal His potential intracardiac electrogram on the ablation catheter (ABLd) captured adjacent to the AV junction after deploying a large curve ablation catheter via a long femoral venous sheath. Radiofrequency ablation delivered at this location resulted in junctional beats.
Figure 3. These results from the postprocedure computed tomography angiography confirm the right-sided superior vena cava (SVC) with leads entering a grossly dilated coronary sinus (CS) due to the persistent left sided SVC with direct drainage into the RA. The azygous vein inserts directly to the right SVC prior to connecting with the CS.)
navigate the ablation catheter through a long sheath that can provide additional stability and manipulation to the desired location in the setting of complex anatomy. A retrograde approach to ablate the AVJ through the left side could have been considered and used. However, in the presence of a bioprosthetic aortic valve, we thought that the femoral approach would be more optimal. Our case was uniquely challenging given that the azygos continuation of the IVC occurred in a patient with dextrocardia, complete situs inversus, and bilateral persistent SVC. In the presence of these anomalies, access to the right cardiac chambers from the femoral approach via the azygos vein and persistent SVC is feasible. However, positioning and manipulation of the catheters were unusually challenging as a result of the longer course and the numerous angulations of the azygos venous system communicating with the SVC.

In conclusion, this case highlights the uniquely complex venous anatomy associated with situs inversus, dextrocardia, complete left SVC, dilated coronary sinus, and an azygos system connected with the right SVC but disconnected from the IVC in a patient requiring precise localization of the AVN for therapeutic radiofrequency ablation. Preprocedural imaging to define anatomic variations may potentially help with ablation success.

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