Complex cardiac anatomy and catheter access: the role of imaging in patients referred for catheter ablation

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A 66-year-old man with Kartagener’s syndrome, situs inversus totalis, and recurrent supraventricular tachycardia and a 49-year-old man with atrial fibrillation and drug-refractory rapid ventricular rate response were referred for catheter ablation. In the first case, the mirrored anatomy of the right atrium was reconstructed using three-dimensional electroanatomical mapping, which guided successful ablation of a typical atrioventricular nodal reentrant tachycardia. In the second case, computerized tomography showed the presence of left atrial isomerism with interruption of the inferior vena cava and azygous continuation into the superior vena cava, guiding advancement of the ablation catheter for access into the positionally right atrial ablation site. These cases illustrate the role of imaging in patients with unusual anatomy of the cardiac chambers and major blood vessels guiding optimal catheter access for catheter ablation.

Case 1: atrioventricular nodal reentrant tachycardia ablation in a patient with Kartagener’s syndrome

A 66-year-old man with Kartagener’s syndrome, situs inversus totalis, and recurrent atrioventricular nodal reentrant tachycardia was referred for electrophysiologic study. After having received informed consent, an electrophysiologic study was performed. Two quadripolar catheters were positioned on the His position and in the right ventricular apex, and a decapolar catheter was positioned within the coronary sinus. The electrophysiologic study confirmed that a typical atrioventricular nodal reentrant tachycardia was responsible for the clinical arrhythmia. Fluoroscopic view of the catheters during the ablation is shown in Figure 1A. The CARTO mapping system (Biosense Webster Inc., Diamond Bar, CA, USA) was used to map the anatomy of the mirrored right atrium and to guide the ablation performed using a 7 French D-curve 4 mm tip ablation catheter (Navistar®, Biosense Webster Inc.) as shown in Figure 1B. After the ablation, there was no residual slow pathway conduction and the clinical arrhythmia was not inducible without and with infusion of isoproterenol.

**Figure 1** (A) Fluoroscopic image with an antero-posterior projection showing the ablation catheter at the region of the slow pathway (Abl), a decapolar steerable catheter positioned within the coronary sinus (CS), and a quadripolar catheter on the His position. (B) Right anterior oblique (30° projection) electroanatomical map of the mirrored right atrium (grey) in the patient with Kartagener’s syndrome and situs inversus. The atrioventricular node is pointed out with an orange point and the successful ablation sites in the slow pathway region with red points. The coronary sinus is shown in blue and both superior and inferior vena cava in green.

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Case 2: atrioventricular node ablation in a patient with atrial fibrillation and left atrial isomerism

A 49-year-old man with atrial fibrillation and drug-refractory rapid ventricular rate response was referred for atrioventricular nodal ablation and cardiac resynchronization therapy. He had a history of congenital aortic stenosis due to a dysplastic tricuspid aortic valve, as well as mitral valve prolapse. The intracardiac anatomy was otherwise normal. Prior to his referral, he had implantation of a dual-chamber pacemaker for ‘sick sinus’ syndrome and uncomplicated prosthetic mechanical aortic valve replacement, mitral valve repair, and intraoperative left atrial cryoablation combined with clipping of the appendage of the positionally left atrial appendage.

Computerized tomography of the thorax and the abdomen showed the presence of left atrial isomerism with interruption of the inferior vena cava, ayzygous continuation into the superior vena cava, and polysplenia syndrome (Figure 2A and B). The anatomy of the coronary sinus was normal and suitable for implantation of a left ventricular lead. After having informed consent, the patient was brought to the electrophysiology laboratory. An ablation catheter was inserted through the sheath in the right femoral vein. The ablation catheter was advanced through the ayzygous vein into the superior vena cava and the positionally right atrium. The position of the 7 French D-curve 4 mm tip ablation catheter (Celsius®, Biosense Webster Inc.) on the atrioventricular nodal ablation site is shown in Figure 3.

Discussion

Complex cardiac anatomy in the presence of congenital anomalies of the heart may be challenging to electrophysiologists during catheter ablation procedures. For example, instead of the normal atrial arrangement (situs solitus), one may encounter mirror imaging (situs inversus) or independent of the atrial arrangement the apex of the heart may point to the right (dextrocardia). While all forms of abnormal atrial arrangement can occur in isolation, associated intracardiac defects are common, particularly in patients with heterotaxy syndromes. These conditions may complicate the interpretation of intracardiac signals and manipulation of the electrophysiology catheters must be performed in an unfamiliar environment by the operator who is accustomed to anatomical landmarks.
The incidence of dextrocardia is estimated to be 1 in ~12,000 births, while one-third of these have situs inversus. Kartagener’s syndrome is a rare condition with autosomal recessive transmission comprising situs inversus, primary ciliary dyskinesia with nasal polyposis, and bronchiectasis. Its prevalence is estimated as 1 in 130,000 of the general population. There have been only few case reports of catheter ablation of supraventricular tachyarrhythmias in patients with dextrocardia and situs inversus published. This report is the first to provide a three-dimensional electroanatomical map of a patient with the Kartagener’s syndrome and atrioventricular nodal reentrant tachycardia. Tagging the atrioventricular node and the slow pathway on the three-dimensional electroanatomical map was useful to avoid atrioventricular block in the presence of unusual anatomy. Likewise, novel technologies for remote robotic catheter navigation, such as magnetic navigation, may be helpful in such cases.

Left atrial isomerism with azygous continuation and interruption of the inferior vena cava may be observed unexpectedly during insertion of electrophysiology catheters. Isomerism is present in 1–4% of patients with congenital heart defects with a large heterogeneity at clinical presentation depending on associated cardiac malformations. Intracardiac involvement in left atrial isomerism is often less severe than in right atrial isomerism. The presence of interrupted suprarenal portion of the inferior vena cava with azygous continuation strongly suggests left atrial isomerism, which is frequently associated with two or more spleens (polyspenia syndrome) as it was the case in our patient.

During a standard catheter ablation procedure, the catheter is targeted to the ablation region based on typical local electrogram characteristics and anatomical landmarks. As indications for catheter ablation expand, the complexity of the anatomical substrates in patients with congenital anomalies may pose a challenge to the electrophysiologist. Our cases illustrate the role of imaging in patients with unusual presentation of the anatomy of the cardiac chambers and major blood vessels. The identification of the accurate anatomy using imaging modalities such as computerized tomography and magnetic resonance imaging and three-dimensional image reconstruction using mapping systems may be useful not only to help the electrophysiologist enabling a safe and successful catheter ablation procedure, but also to understand the complex anatomical structures and to guide for optimal catheter access.

References