Persistent left superior vena cava: a blessing in disguise

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Persistent left superior vena cava (SVC) is an uncommon condition, usually encountered during cannulation of the left subclavian vein. We describe a patient who required a cardiac device upgrade to cardiac resynchronization implantable cardioverter defibrillator for biventricular failure. The presence of a persistent left SVC proved to be a blessing in disguise in this patient as he had since developed total occlusion of the left subclavian-innominate system.

Keywords Persistent left superior vena cava; Congenital cardiac anomalies; Cardiac resynchronization therapy

Introduction

Persistence of the left superior vena cava (SVC) is a rare congenital venous anomaly often detected during left subclavian vein cannulation. The technique of device implantation through the left persistent SVC, although challenging, has been described in the literature.

We present the case of a young man with a pre-existing dual-chamber pacemaker originally implanted from the left subclavian-innominate system, who required an upgrade to a biventricular defibrillator. An accidental discovery of a persistent left SVC proved to be a blessing in disguise as the left subclavian-innominate system was found to be occluded.

Case presentation

A 36-year-old Caucasian male with surgically corrected L-Transposition of the great vessels and a ventricular septal defect had a dual-chamber permanent pacemaker implanted in the past using a conventional left subclavian approach for postoperative complete heart block.

He presented with a monomorphic ventricular tachycardia in the presence of moderately reduced left ventricular function (ejection fraction 35%) and severely reduced right ventricular systolic function. Owing to symptoms of biventricular failure, a wide QRS (200 ms) with right ventricular pacing, and presentation with ventricular tachycardia, he was considered for an upgrade to a biventricular intracardiac defibrillator.

After obtaining access to the left subclavian vein, venography was performed which showed the innominate system to be completely occluded and an incidental discovery of persistent left SVC draining into a large coronary sinus was made (Figure 1). A coronary sinus angiogram revealed a large calibre lateral cardiac vein. After the failure of multiple attempts to cannulate the lateral cardiac vein with guiding sheaths of different sizes and curves, an Amplatz guiding sheath was used to successfully place the coronary sinus lead (St. Jude Medical, Quicksite, Model 1056) in the target vessel.

The implantation of any device via the persistent left SVC can be challenging. After advancing the right ventricular defibrillation lead via the persistent left SVC into the right atrium, a preformed curved stylet with a distal loop was introduced into the lead and a counter-clockwise rotation was given to the lead, thereby directing it towards the tricuspid valve. This manoeuvre facilitated the direction of the lead towards the right ventricular apex or the interventricular septum. Subsequently, with the introduction of a straight stylet, the lead was screwed into the right ventricular apex. The old right ventricular pacing lead was capped and the remaining leads were connected to the cardiac resynchronization device (St. Jude Medical, Atlas II DR, Model V-268). The old atrial lead was manufactured by Guidant (Model 4269).

A dual-coil defibrillation lead (St. Jude’s Medical, Riata, Model 7020) was used in this patient with the distal coil positioned in the right ventricle and the proximal coil in the main coronary sinus with the aim of achieving lower defibrillation thresholds (Figure 2). The defibrillation threshold achieved with this configuration was < 10 Joules.

On follow-up, the pacing and sensing parameters as well as the position of the leads were found to be stable.
Persistent left SVC is the most common thoracic venous congenital anomaly with a prevalence of 0.3–0.5% in the general population.\textsuperscript{1,2} This results from failure of the left anterior cardinal vein to degenerate during embryonic development.\textsuperscript{3} In more than 90% of cases, the left SVC drains into the right atrium via the coronary sinus\textsuperscript{4} and in the remaining 10% it drains into the left atrium resulting in partial anomalous systemic venous return.\textsuperscript{5} In 82% of reported cases, a right SVC is present.\textsuperscript{6} In the remainder, drainage of venous blood from the upper body occurs exclusively through a persistent left SVC.

Persistence of the left SVC is usually asymptomatic and is being discovered as an incidental finding with greater frequency because of increase in invasive cardiovascular and electrophysiology procedures.
Placement of dual-chamber and biventricular pacemakers and defibrillator leads via a left SVC has been reported. In our case, the presence of the left SVC proved to be very useful for upgrading the existing system instead of placing the whole system de novo via the right subclavian vein.

Device placement via persistent left SVC can be technically challenging, however, since the introduction of guiding sheaths with different sizes and curves, along with inner catheter systems with different distal angulations, this problem has by and large been circumvented. We found the technique of making a large distal loop in the lead with the help of a stylet and rotating the distal end towards the tricuspid valve to be very useful. In addition, we prefer to use a dual-coil system for defibrillation, with the proximal coil in the coronary sinus and the distal coil in the right ventricle. This allowed for most of the left ventricle to be included in the defibrillation pathway with the hope of achieving a lower defibrillation threshold. The time taken to place the lead in the right ventricle in our case although somewhat longer was not significantly different when compared with placing the lead through the normal route.

**Conclusion**

Placement of pacemaker and defibrillator leads via a left SVC although technically difficult is possible. Persistence of the left SVC is no longer considered a problem for implantation of devices and at times may prove to be a blessing in disguise as in our case.

**Conflict of interest:** none declared.

**References**