Case Report

Implantation of a Dual-Chamber Cardioverter Defibrillator System in a Patient with Dilated Cardiomyopathy, Pulmonary Hypertension and Persistent Left Superior *Vena Cava*

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We report the case of a dual-chamber cardioverter defibrillator (ICD) implantation in a young patient with a persistent left superior *vena cava*. The patient had biventricular dilatation, pulmonary hypertension and severely depressed left ventricular ejection fraction due to non-ischemic cardiomyopathy. Appropriate use of currently available, low profile active fixation leads allowed safe implantation of both leads through the left subclavian vein.

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Persistence of the left superior *vena cava* is a venous system malformation that may complicate procedures associated with left superior thoracic venous cannulation, such as placement of a pacemaker or ICD leads. In the present case we report the implantation of a dual chamber ICD system in a patient with a persistent left superior *vena cava* (PLSVC), biventricular dilatation, pulmonary hypertension and a severely depressed left ventricular ejection fraction due to non-ischemic cardiomyopathy.

Case presentation

A 38-year-old male patient was admitted to our hospital for an elective implantation of an ICD system. The patient had a history of non-ischemic cardiomyopathy with an ejection fraction of 25%, a left ventricular end-diastolic diameter of 85 mm and an estimated pulmonary artery systolic pressure of 60 mmHg. The patient was in New York Heart Association stage II under maximal tolerated medical treatment and had experienced two syncopal episodes. The patient had also manifested non-sustained ventricular tachycardias on 24-hour Holter monitoring and inducible ventricular tachycardia during programmed ventricular stimulation. The duration of the QRS complex was <120 ms; thus the patient was not considered a suitable candidate for a biventricular ICD.

The presence of a PLSVC was an incidental finding during the implantation and was recognized due to the unusual course of the J-tipped guidewire when advanced under fluoroscopy (Figure 1). The diagnosis was established with contrast venography. This venous malformation raised difficulties in gaining access to the right chambers. The guidance of the ventricular lead was achieved after manually forming the shape of a stylet with a proximal bend and a distal rounded curve (Figure 2). The ventricular lead (Durata® Defibrillation Lead, St. Jude, USA) was finally positioned in the low septum—it is our routine

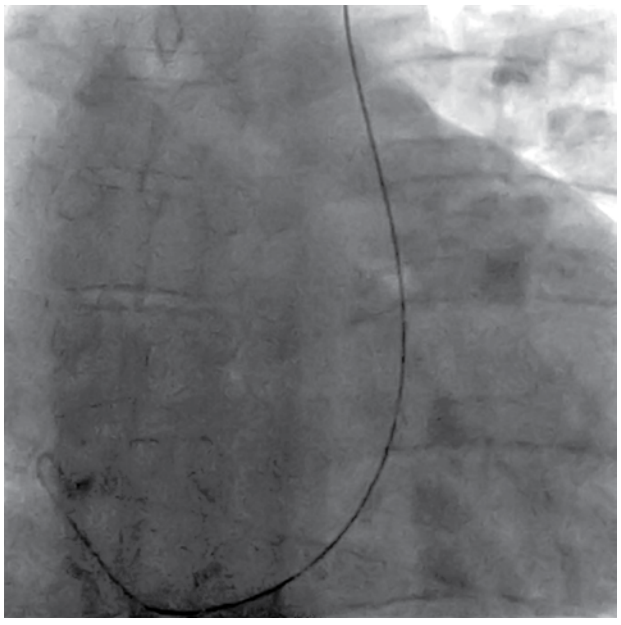

Figure 1. Fluoroscopic view of the J-tipped guidewire through the persistent left superior *vena cava* and the coronary sinus to the right atrium.

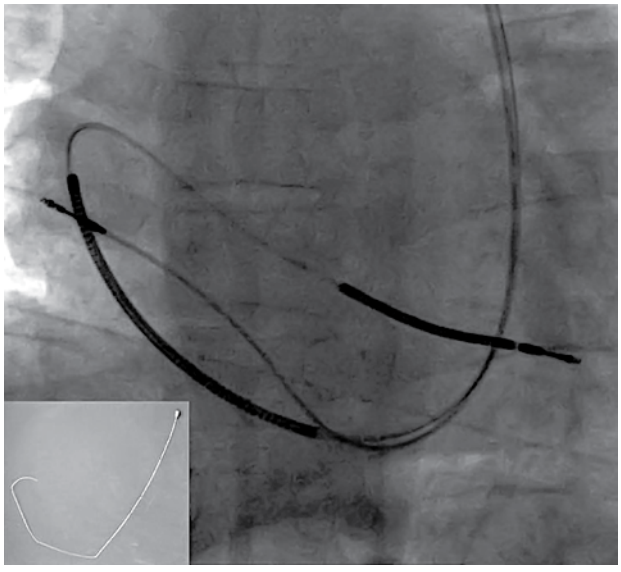

Figure 2. Fluoroscopic view of atrial and ventricular lead placement in our patient with persistent left superior *vena cava*. Please note that the ventricular lead is positioned in an alternate pacing site (low septum). In the lower left corner the manually formulated shape of the stylet used for placement of the ventricular lead is shown.

practice to seek alternate sites for ventricular lead implantation in patients with left ventricular dysfunction who are likely to receive ventricular pacing in the future. The atrial lead was positioned in the lateral atrial wall with the use of a large-curved stylet. Active fixation leads were used in order to minimize the risk of subsequent lead dislodgment. Both leads presented optimal measured parameters intraoperatively (ventricular lead: R wave >12 mV, slew rate 3 V/s, impedance 490 Ω, pacing threshold 0.75 V / 0.5 ms, shock impedance 63 Ω; atrial lead: P wave >2.5 mV, slew rate 0.6 V/s, impedance 450 Ω, pacing threshold 1.2 V / 0.5 ms). These values were maintained in the system interrogation which was performed during the one-month follow-up visit.

Discussion

The persistence of the left superior *vena cava* is a variation of the venous system that has been reported to occur in 0.3% of the general population and at higher frequencies, ranging from 2-9%, among patients with different types of congenital heart disease.[1,2] In 90% of cases, the PLSVC drains to the right atrium via a dilated coronary sinus, while in 10% of cases it connects to the left atrium, thus resulting in a right-to-left shunt. In a percentage of patients with PLSVC that has been reported to range from 10% to 36%, the right superior *vena cava* is also absent.[3-5] In a prospective assessment of the prevalence of PLSVC among consecutive patients undergoing ICD or pacemaker implantation over a 10-year period, Biffi et al reported a PLSVC occurrence rate of 0.34% among pacemaker recipients and a rate of 1.7% among ICD recipients.[4]

The presence of PLSVC complicates the implantation of pacemaker or ICD systems when a left-sided approach is attempted, while in a considerable percentage of cases a right approach is not feasible due to the concomitant absence of the right SVC. In the majority of cases the diagnosis is documented intraoperatively, although the recognition of a markedly dilated coronary sinus (CS) without right atrial dilatation in a standard parasternal long-axis view during preoperative transthoracic echocardiography may raise the suspicion of PLSVC. Radiographic features or clinical examination findings that may also suggest the presence of a PLSVC include a left paramediastinal venous crescent on the chest X-ray and a greater distension combined with a double A wave in the left jugular vein in comparison to that of the right vein.[6]

Technical difficulties associated with PLSVC-related anatomic peculiarities need to be addressed during pacemaker/ICD implantation. The major barrier that has to be overcome is the steering of the ventricular lead through the PLSVC, the coronary sinus

and then through the tricuspid annulus in the right ventricle. When the lead is advanced out of the ostium of the CS it points away from the tricuspid orifice and towards the opposite right atrial wall. Therefore, the use of a preshaped stylet is needed in order to redirect its course around the tricuspid annulus and through the valve. In our case, this manipulation was further impaired by the presence of increased systolic and diastolic pressure in the dilated right ventricle and by the jet of regurgitant blood flow due to coexistent tricuspid insufficiency. Apical lead positioning has been reported to be facilitated by manually shaping the stylet used so that a proximal leftward sharp bend followed by a distal rounded L are formed.[3] Zerbe et al have reported that reshaping the stylet end into a pigtail configuration 3-4 cm wide without complete "closing" of the loop enables lead placement in the right ventricle without difficulty.[7] However, the shape of the stylet usually has to be formed on an individualized basis in order to account for the anatomic peculiarities of each patient. Placement of the ventricular lead can be further complicated when the implanter seeks an alternate site in order to avoid the detrimental effect of conventional right ventricular apical pacing, as was the case in our patient. Furthermore, handling of a dual-coil ventricular lead may be impeded by the presence of the "SVC" coil in the coronary sinus. Indeed, if the authors had been aware of the anatomy before choosing the type of the lead, they would have preferred an ICD electrode without an "SVC" coil. However, the function of the defibrillator system is not affected by presence of the "SVC" coil, because the configuration of the defibrillation shock can be programmed accordingly, omitting the involvement of the "SVC" coil.

Another obstacle to device implantation in patients with PLSVC is the length of the leads used. Taking into consideration the circuitous, elongated course of the ventricular lead along the PLSVC, the CS and the occasionally dilated right ventricle, the length of conventional leads may be inadequate and therefore longer leads may be necessary. In similar demanding implantations, it is common practice to favor the use of active fixation leads in order to minimize the risk of subsequent lead dislodgement.[3,4,8]

In conclusion, PLSVC may complicate left-sided implantation of pacemaker or ICD systems. Technical difficulties can be overcome with the use of specially shaped stylets and thin, high performance handling, active-fixation leads, thus ensuring reliable and proper system performance even in patients with the aforementioned characteristics.

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