Pacemaker Implantation in a Patient with Paroxysmal Atrial Fibrillation and Persistent Left Superior Vena Cava with Absent Right Superior Vena Cava

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To the Editor: Persistent left superior vena cava (PLSVC) with the absence of right superior vena cava (RSVC) is a very rare congenital anomaly that can complicate the implantation of a permanent pacemaker lead. If the atrial lead is placed beyond the right atrial appendage during atrial fibrillation (AF), assessment of adequate lead fixation might be challenging. We present a case of PLSVC with the absence of RSVC and sick sinus syndrome with paroxysmal AF, in which a permanent pacemaker was successfully implanted.

A 43-year-old man was admitted to our hospital with a complaint of recurrent palpitation for 1 week. A 12-lead electrocardiography revealed sinus rhythm with the left axis deviation of the P-wave and normal PR interval without PR segment. A Holter monitor revealed sinus arrest, atrioventricular junctional escape, and paroxysmal AF with a 4.3 s RR interval. Transthoracic echocardiography (TTE) showed a dilated coronary sinus (CS) and a PLSVC. Dual chamber permanent pacemaker was considered, and a right subclavian approach was used first. After puncture of the right subclavian vein, a guidewire was passed through the left-sided superior vena cava (SVC). Right subclavian vein venography demonstrated the absence of RSVC and drainage of right subclavian vein into the PLSVC via the innominate vein [Figure 1a]. A left subclavian approach was used due to ease of access. Due to the anomalous left-sided SVC, the leads took a circuitous course via the PLSVC. An active fixation pacemaker lead was successfully placed at the right ventricular apex with acceptable parameters (bipolar: threshold 0.5 V/0.4 ms, impedance 500 Ω, R-wave amplitude 10 mv) by method of Mora G[1] [Figure 1b-1d]. The other active fixation pacemaker lead was positioned in the posterior wall of right atrium (RA). During this process, AF persisted despite attempts to convert to sinus rhythm with propafenone. Based on our experience and an atrial fibrillatory wave amplitude of 1.6 mv with good atrial current of injury, the position of the atrial lead was considered to be acceptable. Sotalol 80 mg, taken orally, twice daily was prescribed after operation and sinus rhythm was restored 2 days later. Pacemaker analysis showed atrial parameters were acceptable (bipolar: threshold 0.5 V/0.4 ms, impedance 560 Ω, P-wave amplitude 4 mv). At 2-month follow-up, the patient was asymptomatic with no episodes of AF, acceptable lead parameters, and a fluoroscopically appropriate lead position.

PLSVC is usually a structural anomaly, present in 0.3–0.5% of the general population and in 4–10% of congenital heart disease.[2] It is always asymptomatic. During normal embryologic development, the left-sided anterior venous cardinal system regresses, leaving the CS and the ligament of Marshall. Failure of closure of the left anterior cardinal vein results in PLSVC. In most patients with left SVC, RSVC is present (bilateral SVC) and drains into the RA via a dilated CS. PLSVC with the absence of RSVC occurs in only 10–20% of cases of PLSVC.[3] In 90% of individuals, the PLSVC drains into the RA via the CS without hemodynamic consequence.[2,3] In the remaining cases, it might drain into the left atrium resulting in a right- to left-sided shunt or into the inferior vena cava or hepatic vein.[2,3] A PLSVC can cause problems during central venous catheterization (access to the CS can cause hypotension, angina, perforation of the heart, tamponade, and arrest), pacemaker implantation (due to the tortuous course of the electrode, it might be difficult to fix the electrode into position and obtain stable capture), or cardiopulmonary bypass (isolated PLSVC impairs the use of retrograde cardioplegia). In addition, a higher incidence of arrhythmias and conduction system abnormalities has been described in patients with PLSVC.[4] There are two proposed mechanisms for these: a dilated CS stretches the atrioventricular nodal tissue, which results in reentrant tachycardias; or the proximity of early conduction tissue to the cardinal venous tissue leads to sinus node dysfunction. Lenox *et al*.[4] found sinoatrial node abnormalities in some patients with absent RSVC that might predispose them to sick sinus syndrome. In this case, a Holter monitor revealed sinus arrest and paroxysmal AF with

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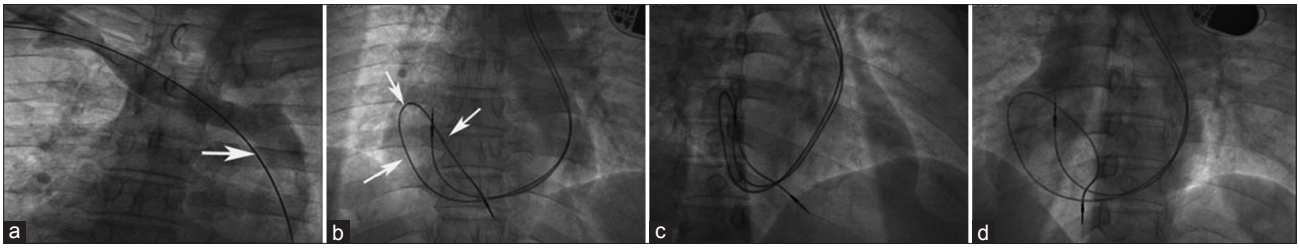

Figure 1: Right subclavian vein venography and dual chamber pacemaker implantation from the persistent left superior vena cava. (a) Right subclavian vein venography demonstrated the absent right superior vena cava and drainage of the right subclavian vein into the persistent left superior vena cava (white arrow) via the innominate vein; (b) the pacemaker leads successfully placed at the right ventricular apex and posterior wall of right atrium via the persistent left superior vena cava. The white arrows showed circuitous course (AP view); (c) RAO view of the pacemaker leads; (d) LAO view of the pacemaker leads. AP: Anteroposterior; RAO: Right anterior oblique; LAO: Left anterior oblique.

4.3 s RR interval and a pacemaker was successfully implanted. There is no specific problem in pacemaker implantation from the right subclavian vein in patients with bilateral SVC. However, when implanting pacemakers in PLSVC patients with absent RSVC, the left subclavian vein is preferred as lead manipulation is easier. Therefore, if TTE shows a dilated CS and a PLSVC is suspected, the diagnosis should be confirmed by additional contrast, transoesophageal echocardiography or computed tomography to help identify optimum vascular access before pacemaker implantation. There is an acute angle between the CS ostium and the tricuspid valve, which complicates pacemaker lead implantation; therefore, the ventricular lead should be looped in the RA to enter the right ventricle. Hand-shaped stylets and active fixation leads are also helpful in overcoming technical difficulties. Mora[1] introduced a method of ventricular placement with standard J stylet, which has a high success rate and is accompanied by less fluoroscope time and procedure than other series. The method described with four steps does not require a preformed stylet or use of special equipment, but rather uses a standard J stylet which is found in all pacemaker electrode package. In the case, it took 70 s for fluoroscope time and 3.5 min for the ventricular placement using this method. Other techniques, including using a steerable stylet and special sheath, are helpful. Generally, it is not difficult to implant the atrial active fixation lead through PLSVC. Sensing and pacing parameters can be used to determine whether the lead position is appropriate. Atrial fibrillatory electrogram measurement and lead motion when positioned in the right atrial appendage can help determine atrial lead position if AF occurs during pacemaker implantation. However, if the lead is placed beyond the right atrial appendage, lead motion is not helpful and atrial fibrillatory electrogram measurement is the only conventional way to predict adequate lead fixation. However, Saxonhouse *et al*.[5] revealed that atrial wave sensing alone was not reliable to predict adequate lead fixation. Instead, current of injury could guide adequate placement of the active fixation leads. In the case, the atrial active fixation lead was positioned with proper atrial wave sensing and current of injury during AF and sensing and pacing parameters of the lead was appropriate after converting to sinus rhythm.

PLSVC with absent RSVC is a rare congenital anomaly that can complicate the implantation of the permanent pacemaker lead. If the lead is placed beyond the right atrial appendage during AF, it is challenging to predict adequate lead fixation and current of injury is helpful.

Declaration of patient consent

The authors certify that they have obtained all appropriate patient consent forms. In the form, the patient(s)/patient's guardians has/have given his/her/their consent for his/her/their images and other clinical information to be reported in the journal. The patients/patient's guardians understand that their names and initials will not be published and due efforts will be made to conceal their identity, but anonymity cannot be guaranteed.

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Conflicts of interest

There are no conflicts of interest.

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