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CASE REPORT

Transvenous Dual-Chamber Pacemaker Implantation in a Patient with Persistent Left Superior Vena Cava Undergoing Maintenance Hemodialysis

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Background: Persistent left superior vena cava (PLSVC) is a rare congenital venous anomaly. Permanent pacemaker implantation (PPI) in patients with PLSVC presents challenges in placing both the right ventricular and atrial leads. The article describes a technique for dual-chamber PPI using standard leads in a PLSVC patient with chronic kidney disease on maintenance hemodialysis.

Case Presentation: A 69-year-old male patient with sick sinus syndrome (SSS), hypertension, moderate mitral regurgitation, dilated left ventricle, diabetes, chronic kidney disease (on hemodialysis), and NYHA III heart failure underwent dual-chamber pacemaker implantation via the left axillary vein. Venography revealed a persistent left superior vena cava, a challenge for the placement of leads. The ventricular lead was positioned in the right ventricular outflow tract using a Biotronik active fixation lead with a "C" shaped stylet, and the atrial lead was placed in the right atrial lateral wall. The procedure took 115 minutes with 17.5 minutes of fluoroscopy. After 4 months, the patient showed symptom improvement and stable pacing parameters.

Conclusion: Dual chamber pacemaker implantation through PLSVC in a patient undergoing maintenance hemodialysis using a "C" shaped stylet technique is feasible, safe, and effective.

Keywords: persistent left superior vena cava, permanent pacemaker, maintenance hemodialysis

Background

Persistent left superior vena cava (PLSVC) is a rare congenital anomaly of the major venous system, where the superior vena cava (SVC) is positioned on the left side of the body instead of its usual location on the right. The prevalence of PLSVC in the general population is approximately 0.3% - 0.5%,[1] but in patients with congenital heart disease, it increases to 1.3% - 11%. In about 90% of cases, PLSVC drains into the right atrium via the coronary sinus (CS), while in 10–20% of cases, it drains into the left atrium.[2]

Implanting a pacemaker through the left axillary vein in patients with PLSVC draining into the right atrium via CS is complex and presents numerous challenges for placing both the ventricular and atrial leads. Right ventricular lead placement is particularly complex due to navigation difficulties and risks of lead dislodgment,[3] often requiring modifications to the lead stylet in a "C" or "J" configuration and careful navigation through the tricuspid valve into the right ventricle.[4,5]

These challenges are further amplified in long-term hemodialysis patients, particularly those with left ventricular dilation and tricuspid regurgitation caused by arteriovenous fistula (AVF). Such patients face higher risks of infection and

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venous occlusion.[6–8] We present a technique and successful outcomes of permanent dual-chamber pacemaker implantation in patients with PLSVC and end-stage renal disease undergoing periodic hemodialysis.

Case Presentation

A 69-year-old male patient suffered from sick sinus syndrome (SSS) with a heart rate ranging from 40 to 50 beats per minute, hypertension, moderate mitral regurgitation, dilated left ventricle, diabetes, chronic kidney disease (on hemodialysis), and NYHA III heart failure. On a follow-up day, he reported fatigue, mild dyspnea, and inability to walk independently. Examination showed a pulse of 40 bpm, blood pressure of 132/51 mmHg (on antihypertensives), pale skin, no edema or fever, normal lung sounds, and non-palpable liver/spleen. Muscle strength was 4/5 bilaterally.

Laboratory tests revealed mild anemia (Hb 98 g/L), glucose 7.7 mmol/L, HbA1c 7.3%, eGFR <5 mL/min, potassium 4.1 mmol/L, and normal thyroid function. Brain MRI was normal. ECG in Figure 1 showed sinus bradycardia (49 bpm), and 24-hour Holter monitoring confirmed persistent bradycardia (HR: avg 45 bpm, min 36 bpm, max 77 bpm) with bradycardia present 86% of the time, along with occasional premature ventricular contractions classified as Lown Class IVA.

Echocardiogram showed left ventricular dilation (Dd: 60mm; Ds: 36mm), concentric hypertrophy, moderate mitral regurgitation, mild tricuspid regurgitation, moderate pulmonary hypertension (PAPs 46 mmHg), normal left ventricular ejection fraction (EF 70%) and dilated coronary sinus measuring 20.9×24.8 mm. A coronary angiogram revealed 20% stenosis in the mid-left anterior descending artery. The patient consented to permanent dual-chamber pacemaker implantation after a thorough explanation, with all documentation completed.

Preoperative Preparations

The pacemaker was implanted a day after the patient's last hemodialysis session. Due to a right forearm AVF, the pacemaker was placed on the left side. The patient fasted for 6 hours, and 2 g of cefotaxime was administered prophylactically. A Biotronik Evity 6 DR-T dual-chamber pacemaker with active fixation leads Solia S 60, and S 53 leads were used.

Implantation Procedure

A 7 cm incision was made below the left deltopectoral groove to create the pacemaker pocket. Venography revealed a dilated PLSVC draining into the right atrium via the coronary sinus (Video S1). Two access points were established in the left axillary vein under fluoroscopic guidance. The right ventricular lead was implanted first, followed by the right atrial lead.

Right Ventricular Lead Implantation

Firstly, using a straight stylet, advance the lead into the right atrium as far as possible, visualized in an antero-posterior (AP) view. Secondly, replace the straight stylet with a "C"-shaped stylet (Figure 2A). Gently push the entire system (lead with "C" shaped stylet – Figure 2B) into the lateral right atrial wall and rotate the stylet counterclockwise so the lead tip points towards the tricuspid valve annulus. Thirdly, the crucial step of guiding the lead through the tricuspid valve is performed. At this position, gently withdraw the stylet 2–3 cm while simultaneously pushing the lead forward. This

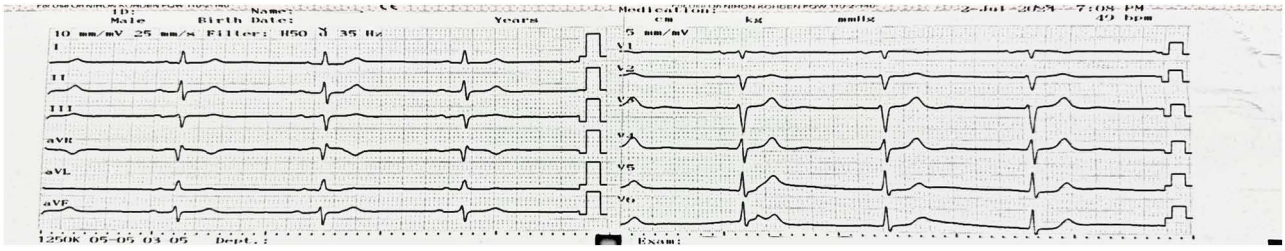

Figure 1 Patient's electrocardiogram (ECG) before pacemaker implantation.

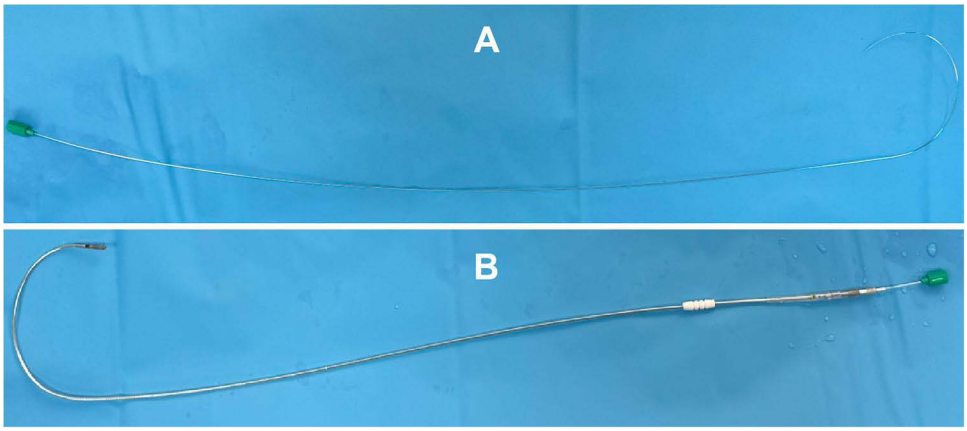

Figure 2 (**A**) "C" shaped stylet; (**B**) Lead with "C" shaped stylet.

should allow the lead to pass smoothly through the tricuspid valve into the right ventricle (Video S2). This step may need to be repeated 2–3 times for success. Fourthly, navigate and fix the lead to an appropriate position in the right ventricle. Continue gently pushing the lead and fixing the stylet to allow contact with the right ventricular outflow tract or interventricular septum. For apical positioning, gently rotate the stylet clockwise and advance the lead towards the apex. Finally, the lead secured with 7–10 turns. Pacing parameters were checked, and the "C" shaped stylet was gently withdrawn and the lead slightly pushed to test stability. Replace the "C" shaped stylet with a straight one, advancing it to the coronary sinus ostium or partially into the right atrium to recheck lead stability. If stable with good pacing parameters, suture the lead in place. Fluoroscopic views (RAO 30°, LAO 30°, and AP) were used for guidance (Video S3).

Right atrial lead implantation: The lead was advanced into the right atrium using a straight stylet, then replaced with a "C" stylet. Due to the curved path and angle, placing the atrial lead in the right atrial appendage was challenging, so we navigated and fixed the lead to the high lateral right atrial wall. Fixation and good pacing threshold. Secure the atrial lead.

Pacing parameters: Right ventricular lead: 0.5V at 0.4ms; impedance 700 Ohm; sensing 10.7 mV. Right atrial lead: 0.8V at 0.5ms; impedance 450 Ohm; sensing 1.8 mV. Total procedure time: 115 minutes, fluoroscopy time: 17.5 minutes.

Results and Follow-Up

Post-implantation, the patient continued antibiotic therapy and inpatient treatment for 7 days before discharge. Follow-up at 1, 3, and 4 months showed clinical improvement, including reduced fatigue, better mobility, and improved self-care. The device pocket healed well, and pacemaker parameters remained stable with 100% atrial pacing, <1.0% ventricular pacing, and minimal pacing thresholds, sensing, and impedance changes (Figure 3). The chest X-ray film confirmed

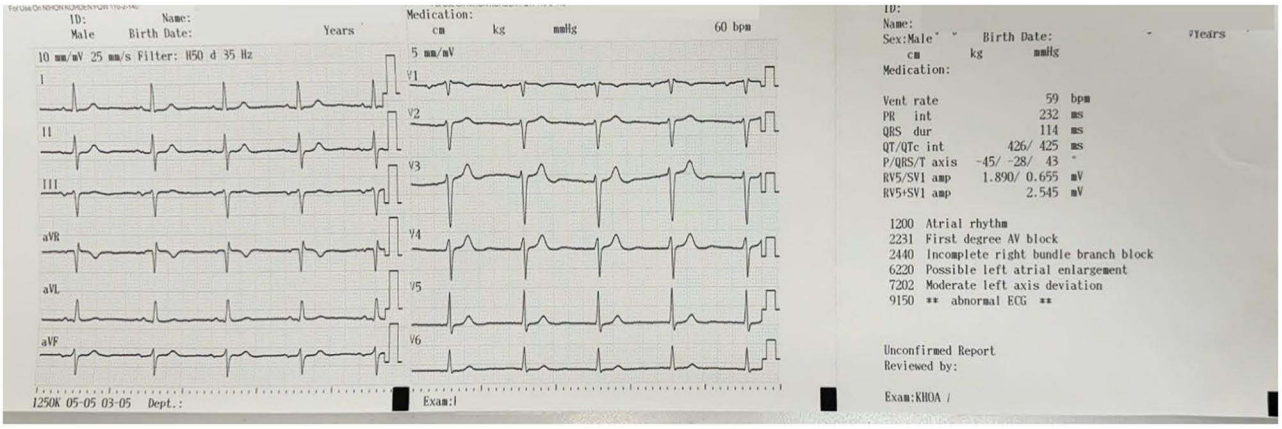

Figure 3 ECG post dual chamber pacemaker implantation, A pacing, and V sensing.

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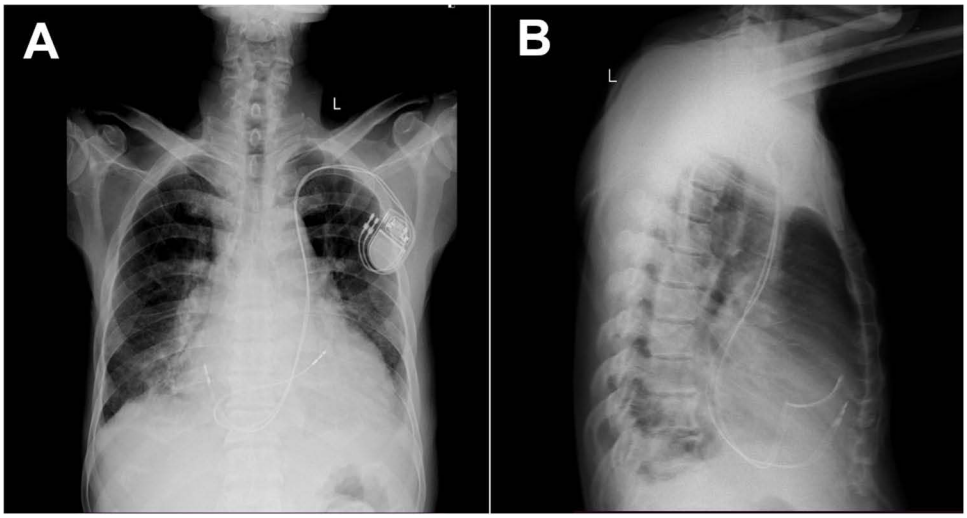

Figure 4 Chest X-ray after 3-month follow-up ((**A**): PA view; (**B**): Left lateral view).

proper device and lead positioning (Figure 4), and echocardiography parameters showed no significant changes in valves, left ventricular size, or LVEF.

Discussion

Persistent left superior vena cava (PLSVC) is a congenital anomaly caused by the failure of the left anterior cardinal vein to regress during embryonic development, affecting 0.3–0.5% of the population.[1] The left anterior cardinal vein regresses typically, leaving the right vein to form the superior vena cava (SVC). In PLSVC, the left vein persists, leading to either double SVCs or, less commonly, a single left-sided SVC if the right vein regresses.[9] PLSVC typically drains into the right atrium via the coronary sinus, but in rare cases, it drains into the left atrium or pulmonary veins, causing left-to-right shunting, cyanosis, or paradoxical embolism. The anomaly is often incidentally detected during cardiovascular procedures or imaging, including echocardiography, CT, MRI, or venography.

The prevalence of PLSVC in hemodialysis patients is rare, and comprehensive statistics are lacking. However, there are clinical reports of hemodialysis cases involving PLSVC. Catheter placement through the subclavian vein or internal jugular vein in these patients can be challenging and carries risks such as arrhythmia, vascular wall injury, and embolism. Despite these challenges, complications and adverse events are very low, comparable to procedures performed in normal veins.[10,11]

Most PLSVC cases are asymptomatic but may complicate procedures like pacemakers, defibrillator implantation, or other cardiac interventions,[3,12,13] with an incidence of 0.37% during PPI (9/2420 cases).[3] Left axillary vein access is standard, but in PLSVC patients, the right SVC approach is preferred if available. In cases of absent right SVC, right-hand dominance, or right-sided AVF in hemodialysis patients, the PLSVC route may be optimal.

Pacemaker implantation through PLSVC presents challenges in navigating the lead through the tricuspid valve into the right ventricle due to the posterior-inferior position of the coronary sinus ostium relative to the interatrial septum and a high risk of lead dislodgement.[3]

The technique for permanent pacemaker implantation in patients with persistent left superior vena cava (PLSVC) depends on the type of PLSVC (Type I with a normal superior vena cava, type II with only PLSVC, Type IIIA with Right and left superior vena cava connection, and Type IIIB with right and left superior vena cava without connection),[14] and the characteristics of the lesion or previous procedures performed in the superior vena cava system. For Type II, left venous access is recommended, while for Type III, the right axillary or subclavian vein access is more convenient for implanting the right ventricular lead. In our case, the patient had Type IIIB PLSVC and was undergoing hemodialysis with the right AVF so that we opted for left venous access to minimize the risks of thrombosis, venous obstruction, and infection.

In most cases without PLSVC, using a J-shaped, C-shaped stylet or Mond's curve allows for successful implantation of the right ventricular lead. However, in patients with PLSVC, inserting the right ventricular lead through the PLSVC is challenging due to the acute angle formed by the CS orifice and the tricuspid valve. In such cases, the C-shaped technique facilitates lead implantation into the septum or right ventricular outflow tract more effectively than the J-shaped stylet.[3]

We used a "C"-shaped stylet technique in our hemodialysis patient with dilated cardiac chambers. The implantation steps were as described above, allowing us to easily navigate through the tricuspid valve and fix the lead in the right ventricular outflow tract position. However, after securing the lead to the right ventricular wall and achieving good pacing parameters, removing the "C"-shaped stylet from the lead proved challenging due to its curved shape and multiple bends, risking lead dislodgement and necessitating repetition of the procedure. In some cases, due to the tortuous and twisted course of PLSVC, there is an increased risk of lead kinking or inability to reach optimal pacing sites, as well as potential vein injury or perforation causing pericardial effusion. Some authors have used long sheaths to facilitate and optimize pacemaker implantation in PLSVC patients.[15]

Additionally, PLSVC can alter the electrophysiological characteristics of the heart, making it difficult to achieve stable lead signals and effective pacing.[16] These factors contribute to longer procedures and fluoroscopy times. Due to the increased risk of lead dislodgement in PLSVC patients, Santosh Kumar Sinha introduced the Alpha loop configuration for ventricular lead to minimize this risk.[17] However, the Alpha loop configuration is suitable for lead implanted in the right ventricular apex or the lower region of the interventricular septum.

For the right atrial lead, due to PLSVC, we attempted to approach and fix it in the right atrial appendage but experienced multiple dislodgements. We successfully fixed it on the high lateral right atrial wall using a "C"-shaped stylet. Data on dual-chamber permanent pacemaker implantation in PLSVC patients is limited. However, implanting the right ventricular lead typically involves using a "J" or "C"-shaped stylet, with potential implantation sites including the right ventricular apex, interventricular septum, or right ventricular outflow tract. The right atrial lead is usually placed on the left lateral atrial wall. The general consensus is that the technique is challenging, with longer procedures, fluoroscopy times, and potential complications along the lead path, but it can be successful.[3,15,18]

End-stage renal disease patients on hemodialysis have a fivefold higher incidence of bradyarrhythmia requiring pacemakers compared to the general population.[19] While transvenous pacemakers offer significant benefits, these patients face higher risks of subclavian vein stenosis (up to 70%),[7] venous occlusion affecting AVF function,[20] and severe infections like pacemaker pocket infection and endocarditis, especially when AVF is ipsilateral to the cardiac implantable electronic devices (CIEDs).[7,8,21] Most cases of central venous stenosis or occlusion are asymptomatic. Patients who become symptomatic can present with edema of the face, neck, breast, shoulder, and arm. Most of these stenoses or occlusions occur within 6 months of CIEDs implantations.[7,22]

CS dissection during PPI or CRT device implant is not common (0.6%).[23] It is usually well tolerated, related tamponade and other life-threatening conditions are rare.[23,24] Coronary sinus thrombosis is a rare phenomenon and potentially fatal entity. There are some cases report occurred CS thrombosis such as pacemaker with lead, catheter ablation for arrhythmias in CS, Swan-Ganz catheters, and central venous lines.[11,25] The risk of CS thrombosis increases in cases of CS dissection during procedures and in patients with advanced age, diabetes, or malignancies.[26]

Leadless pacemakers can help reduce risks of venous stenosis, occlusion, and infections in hemodialysis patients, who often face challenges like dilated cardiac chambers and higher rates of cardiovascular events and procedural complications. [6]

In our patient with a right-sided AVF, we used the left axillary vein approach to minimize complications. The pacing position in the right ventricular outflow tract is our choice to reduce heart failure compared to the lead position in the apex. After four months, no pacemaker-related issues were observed.

Conclusion

Persistent left superior vena cava is a rare congenital anomaly, making dual-chamber pacemaker implantation challenging and high-risk. Using a "C"-shaped stylet, we successfully implanted a dual-chamber pacemaker in a patient with symptomatic bradycardia, PLSVC, and multiple comorbidities, including mitral regurgitation, left ventricular dilation, diabetes, and chronic kidney disease on hemodialysis. After four months, the patient's condition remained stable, with improved symptoms, stable pacemaker parameters, and no complications.

Ethics and Consent

Institutional approval was not required for publication; therefore, informed written consent was obtained from the patient for using their information and case report publication, ensuring that all identifying data remained anonymous.

Disclosure

All authors declare no conflicts of interest in this work.

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