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Case Report

Right-Sided Permanent Pacemaker Implantation in Patients with Persistent Left Superior Vena Cava with Absent Right Superior Vena Cava: 3-Case Series, Technique and Discussion

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ABSTRACT

Persistent left superior vena cava (PLSVC) is the most common variant of abnormal venous return to the heart. While usually asymptomatic, it is known to complicate transvenous cardiac procedures, such as implantation of cardiac electronic devices and ablations. PLSVC can present with or without the concomitant absence of right superior vena cava (RSVC). Depending on the operator's preference, implantation of permanent cardiac pacemakers (PPMs) may be performed from the left or right side. As most often the PLSVC is only identified at the time of intervention, it follows that the variant with the absence of RSVC can be diagnosed in practice only when implanting from the right side. For this reason, the true prevalence of this variant is largely unknown because most published cases of cardiac device implantations in patients with PLSVC have been performed from the left side. We present a short 3-case series of PPM implantations in a tertiary center from the right side in patients with PLSVC and absent RSVC. We found that the use of a standard curve for ventricular lead septal placement and a wide C-curve for right atrial lead placement in these patients was a feasible technique with good outcomes.

Introduction

Persistent left superior vena cava (PLSVC) is the most frequent venous return anatomical variant encountered in 0.3-0.5% of the general population and is characterized by the presence of an anomalous left superior vena cava which usually drains into the coronary sinus [1-3]. Around 20-30% of PLSVC cases are thought to be associated with the absence of the right superior vena cava (RSVC), and in these patients, all thoracic venous drainage takes place through the PLSVC [2, 4, 5]. PLSVC may complicate cardiac device implantation, as transvenous lead placement is more challenging due to distorted anatomy. Most cases of devices implanted via a PLSVC published in the literature are left-sided implants, some employing special techniques [6-9]. The number of right-sided implants in patients with PLSVC and absence of RSVC is obviously lower and far fewer cases have been published, using a range of techniques [10-12]. We present a short 3-case series of patients with PLSVC and absent right SVC who received right-sided permanent pacemakers (PPMs), and briefly describe our technique and outcomes.

Case Reports

Case 1

A 76-year-old woman with tachycardia-bradycardia syndrome, characterized by rare episodes of atrial fibrillation (AF) with fast atrioventricular conduction and sinus pauses of 5 seconds at spontaneous conversion into sinus rhythm, documented on an ambulance ECG strip. Her symptoms were palpitations and recurrent syncope. Her past medical history was remarkable only for mild hypertension; her chronic treatment included an anti-vitamin K oral anticoagulant with good INR control, an angiotensin-receptor blocker, and a diuretic. Her physical examination, resting ECG in sinus rhythm and echocardiographic functional parameters were unremarkable. After PPM implantation (the

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operator opted for a single-chamber VVI pacemaker due to the venous anomaly), she was started on beta-blockers and propafenone, and the rest of her chronic medication was continued. The PPM was programmed VVI 55/min with hysteresis at 40/min to avoid unnecessary ventricular pacing. Her in-hospital course was uneventful. Pacemaker checks at 6 and 12 months showed good electrical parameters, no sustained high-rate episodes, a median heart rate of 60-70/min, and 1% ventricular pacing.

Case 2

A 57-year-old man, also with tachycardia-bradycardia syndrome, characterized by frequent episodes of AF with fast atrioventricular conduction and symptomatic sinus bradycardia after conversion into sinus rhythm (documented on ECG in the emergency room and later also on a 24-hour Holter recording) was presented. His symptoms were palpitations, presyncope and fatigue. His past medical history was otherwise remarkable for hypertension, dyslipidemia and type 2 diabetes; his chronic treatment included a direct oral anticoagulant, a beta-blocker, an angiotensin converting enzyme inhibitor, dihydropyridine calcium channel blocker, statin and insulin. His physical examination and resting ECG in sinus rhythm were within normal range except for sinus bradycardia and a mid (grade 2/6) systolic murmur at the second left intercostal space. Echocardiography showed mild left ventricular (LV) hypertrophy, preserved LV ejection fraction, mild aortic stenosis, mild left atrial enlargement and normal right-sided chambers. The patient refused the option of catheter ablation as treatment for AF but agreed to PPM implantation and upscaling of medication for rhythm control. After dual-chamber PPM implantation, he was started on oral amiodarone, and beta-blockers were re-introduced. The PPM was programmed DDD 60/min with atrioventricular interval hysteresis to avoid unnecessary ventricular pacing. His in-hospital course was uneventful. PPM check at 12 months showed good electrical parameters, rare short episodes of atrial fibrillation, 50% atrial pacing and <1% ventricular pacing.

Case 3

A 69-year-old man presented with recurrent syncope. He had no angina or heart failure symptoms on usual exertion. His past medical history included an inferior myocardial infarction ten years prior, percutaneous revascularization with 2 drug-eluting stents in the right coronary artery (ten and three years prior), right bundle branch block, hypertension, dyslipidemia and type 2 diabetes. Chronic treatment included aspirin, beta-blocker, angiotensin converting enzyme inhibitor, statin and oral antidiabetic medication. His physical examination showed no signs of heart failure. Resting ECG showed sinus rhythm, grade I atrioventricular (AV) block, right bundle branch block and left axis deviation (-70º), with pathological Q waves in the inferior leads. 24-hour Holter recording while in-hospital documented paroxysmal high-grade (AV) block, symptomatic by presyncope. Echocardiography showed mildly dilated left ventricle (LV) with mild systolic dysfunction (LV ejection fraction 45%) and grade II mitral regurgitation. After dual-chamber PPM implantation, his in-hospital course was uneventful. The PPM was programmed DDD 60/min with atrioventricular interval hysteresis to avoid unnecessary ventricular pacing. His in-hospital course was uneventful. PPM check at 48 hours post-implantation showed good electrical parameters, <5% atrial pacing and 11% ventricular pacing. Long-term follow-up data is not yet available, as the patient was recently implanted.

Implant Technique

The patients were informed about the benefits and risks of PPM implantation, agreed to the procedure and signed informed consent forms. All 3 patients received the usual preparation for implantation: pre-operative antibiotic (Vancomycin 1g IV infusion); the right anterior thoracic area was aseptically cleaned and draped. After the right subclavian puncture, the guidewires were noted to pass beyond the midline and then curve caudally into the PLSVC. Contrast venography from both left and right antecubital veins was performed in all cases, confirming thoracic venous drainage via PLSVC and absence of right SVC (Figure 1 shows an example of venography). Right ventricular (RV) active-fixation leads were implanted in all 3 patients via 7F sheaths. We used a wide-curve stylet to advance the leads into the pulmonary artery with a modified classical technique, the lead entering the right atrium (RA) from the coronary sinus and hinging on the lateral right atrial wall to create an arching loop [13]. Then, we used another stylet, C-shaped with a proximal wide curve and a distal short "neck" (Figure 2B) for septal placement by slowly retracting the lead from the outflow tract onto the septum, maintaining counter-clockwise torque and screwing it in when a stable position was reached. Electrical parameter check showed positive lesion current, good sensing (>5mV) and good pacing threshold (<1V @0.4ms) in all cases.

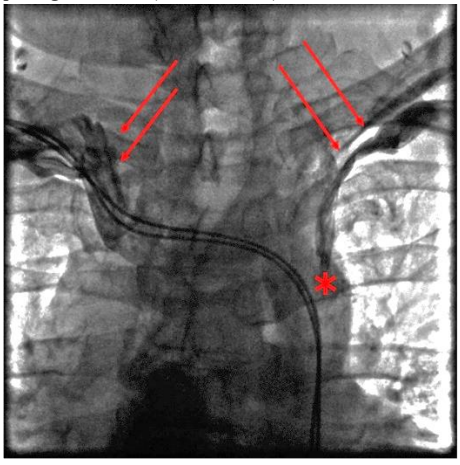

Figure 1: Contrast venography performed post-implant in patient 3, showing that both left and right subclavian veins (arrows) drain into a persistent left superior vena cava (*).

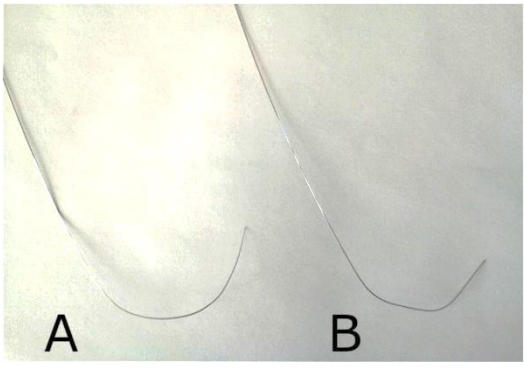

Figure 2: Stylet shapes for **A)** atrial (a wide-radius C curve) and **B)** ventricular (C-shaped with a proximal wide curve and a distal short "neck") lead placement in our patients.

RA active-fixation leads were implanted in 2 of the 3 patients via 7F sheaths. We used a wide-curve C-stylet (Figure 2A) to advance the lead from the PLSVC into the RA and screw it into the high lateral RA wall. Electrical parameter check showed positive lesion current, good sensing (>1.5mV) and good pacing thresholds (<1V @0.4ms). Lead positioning was confirmed fluoroscopically in the anteroposterior, right anterior oblique and left anterior oblique views for all patients (Figure 3, for patient 2 all views were checked, but only the anteroposterior image was recorded). Procedure and fluoroscopy times are detailed in the (Table 1).

Table 1: Procedure and fluoroscopy times for each of the three patients.

Case no	Procedure time (min)	Fluoroscopy time (min), including contrast venography
1	100	9.5
2	80	3.7
3	115	6.6

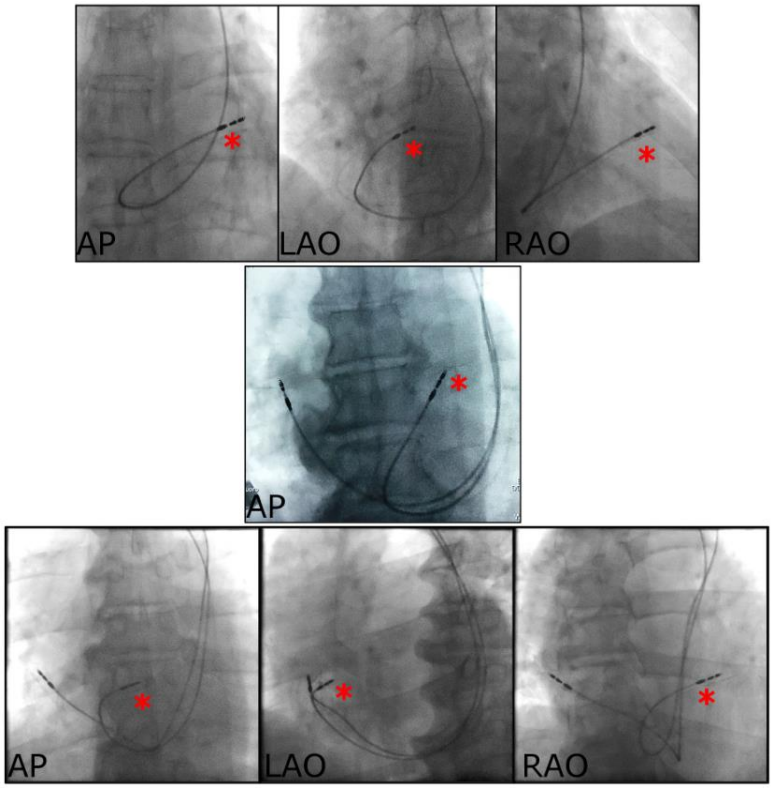

Figure 3: Fluoroscopy views of final lead positions in all 3 patients (upper row, patient 1; middle row, patient 2; lower row, patient 3). (*) marks the right ventricular lead. AP=antero-posterior; LAO=left anterior oblique 30º; RAO=right anterior oblique 30º.

Discussion

PLSVC, resulting from persistent patency of the left anterior cardinal vein (which usually degenerates in its caudal portion during embryonic development), is rare in the general population and consequently among patients with indications of permanent pacing (0.3-0.5%) [3]. This venous anomaly has several anatomical variants, one of which is the concomitant absence of the right SVC, when an important part of the right cardinal vein that should remain functional, atrophies [14]. The prevalence of the latter is difficult to assess since the available information comes mainly from limited series derived from patients with other congenital abnormalities [14, 15]. However, from the available data, its occurrence can be estimated to be at most one-fifth to one-third of LSVC cases [3, 5, 16].

Generally, PLSVC has aroused interest for implanting physicians, especially because of the difficulty of implantation of the ventricular lead, since at the exit from the coronary sinus, it has a reverse direction from the entrance to the RV, towards the lateral wall of the RA. Moreover, another aspect that has been taken into account is the risk of damage to vascular structures by unusual and prolonged manipulation of the leads [10]. Regarding the preferred implanting approach (from the right or left side), when the diagnosis of PLSVC is established before the intervention, some authors advocate the right side for the variant in which there is also a right SVC present [17]. However, when the right SVC is absent, the approach can be right or left-sided because, in both situations, the leads reach the right atrium through the LSVC via the coronary sinus in the same position. The implantation of PPM leads from the right side in such patients is cited in the literature, but essentially in the form of case reports and a short series totaling only a few dozen cases [10-12]. These articles describe the placement of both passive and active fixation leads. To place active fixation leads, several methods have been described to specifically shape the stylets [10, 16, 17]. In our center, the preferred side for “de novo” PPM implantation is the right one. Out of a total of 2359 consecutive PPM implantations performed in our center over a 15-year period (2005-2020), 1950 were right-sided. We described

above the only three cases of PLSVC with absent right SVC we have encountered in this period and the main characteristics of the procedures. In our experience, similar to Li *et al.*, there was no need for a special form of a stylet for the implantation of the ventricular lead [18]. Using a classic shape suitable for implants at the median interventricular septum (Figure 2) after looping the lead in the right atrium and few manipulations needed for orientation of the tip towards the interventricular septum, the advancement of the stylet allowed the fixation of the lead in a stable position without special difficulties. For the atrial lead, we found that the most suitable placement was on the lateral wall of the right atrium (where the orientation was natural) with a wide-curve stylet (Figure 2). The standard length of the ventricular leads (58-60 cm) was sufficient to reach the median interventricular septum. On the contrary, the standard length of the atrial leads (52-53 cm) may be insufficient, especially in patients of higher size or/and when a decision is made to place the lead in the right appendage. A ventricular length lead may be more useful in these cases. In our experience, right-sided implantation of single- or dual-chamber PPMs in patients with PLSVC and absent RSVC did not require significantly longer fluoroscopy or procedure times compared to standard procedures. The exposure doses were higher because intraprocedural venography was performed, but we found this necessary in order to confirm the absence of the RSVC.

Conclusion

PLSVC with absent right SVC is a rare anatomical variant that can, however, pose difficulties during cardiac device implantation. Knowledge of anatomy and adequate implantation techniques can improve outcomes in patients with PLSVC and absent RSVC. In our experience, performing right-sided implants using a conventionally shaped stylet for RV septal placement and a wide-curve C-shape stylet for lateral wall RA placement is a feasible technique with good outcomes.

Author Contributions

All authors have made substantive contributions to the article, and all authors endorse the data and conclusions.

Conflicts of Interest

None.

Funding

None.

References

1. Buirski G, Jordan SC, Joffe HS, Wilde P (1986) Superior vena caval abnormalities: their occurrence rate, associated cardiac abnormalities and angiographic classification in a paediatric population with congenital heart disease. *Clin Radiol* 37: 131-138. [Crossref]
2. Bartram U, Van Praagh S, Levine JC, Hines M, Bensky AS et al. (1997) Absent right superior vena cava in visceroatrial situs solitus. *Am J Cardiol* 80: 175-183. [Crossref]
3. Biffi M, Bertini M, Ziacchi M, Martignani C, Valzania C et al. (2009) Clinical implications of left superior vena cava persistence in candidates for pacemaker or cardioverter-defibrillator implantation. *Heart Vessels* 24: 142-146. [Crossref]
4. Pálinkás A, Nagy E, Forster T, Morvai Z, Nagy E et al. (2006) A case of absent right and persistent left superior vena cava. *Cardiovasc Ultrasound* 4: 6. [Crossref]
5. Peltier J, Destrieux C, Desme J, Renard C, Remond A et al. (2006) The persistent left superior vena cava: anatomical study, pathogenesis and clinical considerations. *Surg Radiol Anat* 28: 206-210. [Crossref]
6. Beig JR, Dar MI, Tramboo NA, Hafeez I, Lone AA et al. (2017) An Innovation in Pacemaker Lead Implantation via Persistent Left Superior Vena Cava: The "3D Alpha Curve" Stylet. *Pacing Clin Electrophysiol* 40: 1042-1044. [Crossref]
7. Naqvi SHR, Ahmed I, Ali PS, Alam M, Zab J et al. (2020) Two Cases of Cardiac Implantable Electronic Device Placement via Persistent Left Superior Vena Cava. *Eur J Case Rep Intern Med* 7: 001484. [Crossref]
8. Sundhu M, Syed M, Gul S, Saqi B, Mosteller R (2017) Pacemaker Placement in Persistent Left Superior Vena Cava. *Cureus* 9: e1311. [Crossref]
9. Chen X, Yu Z, Bai J, Wang W, Qin S et al. (2018) Transvenous cardiac implantable electronic device implantation in patients with persistent left superior vena cava in a tertiary center. *J Interv Card Electrophysiol* 53: 255-262. [Crossref]
10. Sinha SK, Goel A, Razi M, Jha MJ, Mishra V et al. (2019) Permanent Pacemaker Implantation in Patients With Isolated Persistent Left Superior Vena Cava From a Right-Sided Approach: Technical Considerations and Follow-Up Outcome. *Cardiol Res* 10: 18-23. [Crossref]
11. Dilaveris P, Sideris S, Stefanadis C (2011) Pacing difficulties due to persistent left superior vena cava. *Europace* 13: 2. [Crossref]
12. Rizkallah J, Burgess J, Kuriachan V (2014) Absent right and persistent left superior vena cava: troubleshooting during a challenging pacemaker implant: a case report. *BMC Res Notes* 7: 462. [Crossref]
13. Mond HG, Hillock RJ, Stevenson IH, McGavigan AD (2007) The right ventricular outflow tract: the road to septal pacing. *Pacing Clin Electrophysiol* 30: 482-491. [Crossref]
14. Karnegis JN, Wang Y, Winchell P, Edwards JE (1964) Persistent Left Superior Vena Cava, Fibrous Remnant Of The Right Superior Vena Cava And Ventricular Septal Defect. *Am J Cardiol* 14: 573-577. [Crossref]
15. Lenox CC, Zuberbuhler JR, Park SC, Neches WH, Mathews RA et al. (1980) Absent right superior vena cava with persistent left superior vena cava: implications and management. *Am J Cardiol* 45: 117-122. [Crossref]
16. Kanjwal K, Soos M, Gonzalez Morales D, Shah I, Madala M et al. (2019) Persistent Left Superior Vena Cava and Absent Right Superior Vena Cava with Left Subclavian Vein Stenosis: Technical Challenges with Pacemaker Implantation. *Case Rep Cardiol* 2019: 7271591. [Crossref]
17. Biffi M, Boriani G, Frabetti L, Bronzetti G, Branzi A (2001) Left superior vena cava persistence in patients undergoing pacemaker or cardioverter-defibrillator implantation: a 10-year experience. *Chest* 120: 139-144. [Crossref]
18. Li T, Xu Q, Liao HT, Asvestas D, Letsas KP et al. (2019) Transvenous dual-chamber pacemaker implantation in patients with persistent left superior vena cava. *BMC Cardiovasc Disord* 19: 100. [Crossref]