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Long-term follow-up after pacemaker implantation via persistent left superior vena cava

Paweł Dąbrowski[1], Bogdan Obszański[1], Andrzej Kleinrok[1], Andrzej Kutarski[2]

[1]Department of Cardiology, John Paul II Hospital, Zamosc, Poland
[2]Department of Cardiology, Medical University of Lublin, Poland

Abstract

Background: *The persistent left superior vena cava (PLSVC) is a relatively common congenital venous return anomaly. It may have serious clinical implications especially in case of pacemaker/cardioverter-defibrillator implantation, venous catheter insertion, radio-frequency ablation and cardiac surgery. There is also some evidence that PLSVC may be accompanied by arrhythmias. The aim of this report is to present the effectiveness and safety of cardiac pacing via PLSVC, clinical outcome and appearance of arrhythmias in a long-term follow-up.*

Methods and results: *Four cases of pacing electrodes implanted via PLSVC in patients without any other cardiac congenital heart anomalies were observed for at least 6 years. There was 1 patient with AAI, 2 patients with VVI, and 1 with DDD pacemaker. Atrial electrodes were implanted on the free right atrial wall, 2 ventricular electrodes were implanted in right ventricular outflow tract, 1 in postero-lateral cardiac vein. During the mean 110 months of observation, pacing was efficient. One patient underwent an upgrade from AAI to biatrial pacing due to progressive interatrial conduction delay during the follow-up. No other intervention or pacemaker related events were noticed except for battery replacement.*

Conclusions: *The presence of PSVC may complicate implantation, but it does not influence the long-term follow-up of pacing parameters.* (Cardiol J 2014; 21, 4: 413–418)

Key words: persistent left superior vena cava, congenital venous anomaly, pacemaker implantation, arrhythmia, atrial fibrillation

Introduction

The pacemaker (PM) implantation may cause some difficulties in case of persistent left superior vena cava (PLSVC) reported in approximately 0.3–0.5% of the general population [1–3]. The abnormal early embryonic development of great thoracic veins may also result in abnormalities of the conduction tissue because of the proximity of these areas. In most cases (80%), there are double superior veins. Usually, PLSVC drains away to the markedly dilated coronary sinus (CS), and only sporadically to the left atrium (LA). Very rarely, there is a CS ostial atresia and PLSVC drains backward from CS to the right superior vena cava [4]. PLSVC occurs in 3–10% of patients with other congenital cardiac malformations, most often with the atrial septal defect. An important clinical implication of PLSVC is its influence on different types of arrhythmias [5–7]. There are some case reports concerning patients with PLSVC [8–12], but not a long-term observations after PM implantation.

Methods

There were 3,452 patients, who underwent PM implantation from October 1995 to December 2006 in our center. Four cases of PLSVC patients

Address for correspondence: Paweł Dąbrowski, MD, PhD, Department of Cardiology, The John Paul II Hospital, Al. Jana Pawła II 10, 22–400 Zamość, Poland, e-mail: pawel.dabrowski.md@gmail.com
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Table 1. Baseline characteristics of patients (Pt.).

Pt.	Sex	Age [years]	Diagnoses	Arrhythmia		Type of venous anomaly
				Tachy-	Brady-	
1	M	54		Paroxysmal AF	Sinus bradycardia Interatrial block	Single left SVC
2	M	73	AH, CAD	Paroxysmal AFL Paroxysmal AF	Sinus bradycardia Sinoatrial block Impaired AV conduction (measured during pacemaker implant)	Single left SVC
3	F	75	AH, CHF	Permanent AF, ventricular arrhythmia	Paroxysmal AV block	Double SVC
4	M	75	AH, DM, CHF	Permanent AF	Paroxysmal AV block	Single left SVC

AF — atrial fibrillation; AFL — atrial flutter; AH — arterial hypertension; AV — atrio-ventricular; CAD — coronary artery disease; CHF — congestive heart failure; DM — diabetes mellitus; F — female; M — male; SVC — superior vena cava

Table 2. Pacemaker (PM) follow-up.

Pt.	Pacing mode	Follow-up period [month]	Threshold [V] (0.5 ms impulse width)		Sensing [mV]		Resistance [Ω]	
			At PM implant	After follow-up	At PM implant	After follow-up	At PM implant	After follow-up
1	AAI/AAT (BiA)	89	RA 0.7 LA* 0.5	RA 1.8 LA 4.2	RA 2.4 LA* 2.4	2.0**	RA 620 LA* 530	1200**
2	DDDR (RA + RVOT)	84	A 0.4 V 0.1	A 0.8 V 0.7	A 2.2 V 17.4	A 1.8 V 23.6	A 640 V 780	A 580 V 690
3	VVI (LV)	48	1.1	1.5	20	15.7	1100	980
4	VVI (RVOT)	14	0.9	0.8	12.5	13.5	720	810

*Electrode for LA stimulation was placed 48 months after RA (pacing mode was changed); **measurement between RA and LA (interatrial); A — atrium; LA — left atrium; LV — left ventricle; RA — right atrium; RVOT — right ventricular outflow tract; Pt. — patient; V — ventricle

were enrolled for the observation. All the patients had fully documented clinical and electrocardiographic (ECG) indications for PM implantation, and the procedures were performed using standard leads and pulse generators. After the discharge, regular visits took place in the out-patient clinic according to the scheme: 1 month, 3 months after implantation, and every 6 months during follow-up. An ambulatory ECG monitoring was performed to assess the presence of arrhythmias and efficacy of pacing. Clinical characteristics of 4 patients are presented in Table 1, and PM parameters in Table 2.

Patient 1 (Case 1). A 54-year-old man with tachycardia-bradycardia syndrome and a history of ischemic stroke was referred for PM implantation in 1997. The patient complained of palpitations, dizziness and syncopal episodes. At the time of implantation in a standard 12-lead ECG there was a normal sinus rhythm with PQ duration of 120 ms (P-wave duration 80 ms) but the 24-h ECG monitoring showed many episodes of atrial fibrillation (AF) which ended in pauses and severe sinus bradycardia. Trans-thoracic echocardiography did not show any significant structural changes. During the implantation procedure the left subclavian vein approach was not successful, therefore the J-shape, unipolar lead with a passive fixation was inserted via right subclavian vein. The route of the lead was unusual: it passed through the mediastinum from the right to the left side and then went down along the left border of the heart and turned to the right atrium (RA). The tip of the electrode was placed on the lateral wall of the RA because a RA appendage (RAA) could not be achieved. After the PM implantation stimulus to Q wave interval (SQ) duration was 220 ms and P-wave duration was 160 ms. A contrast echocardiography was performed. The micro-bubbles appeared first in the dilated CS, than in the RA after the injection of contrast (cooled 0.9% NaCl) to the left and right antecubital veins.

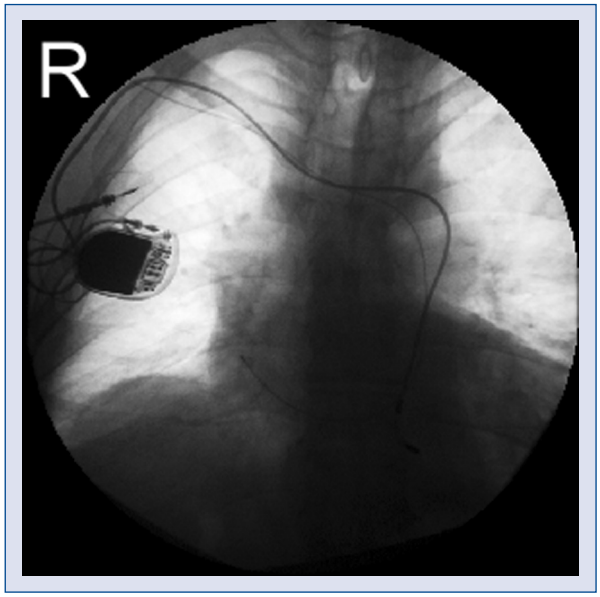

Figure 1. Postero-anterior chest radiograph of AAT BiA pacemaker. Leads are passing through persistent left superior vena cava and are positioned in right atrium free wall and coronary sinus (Case 1).

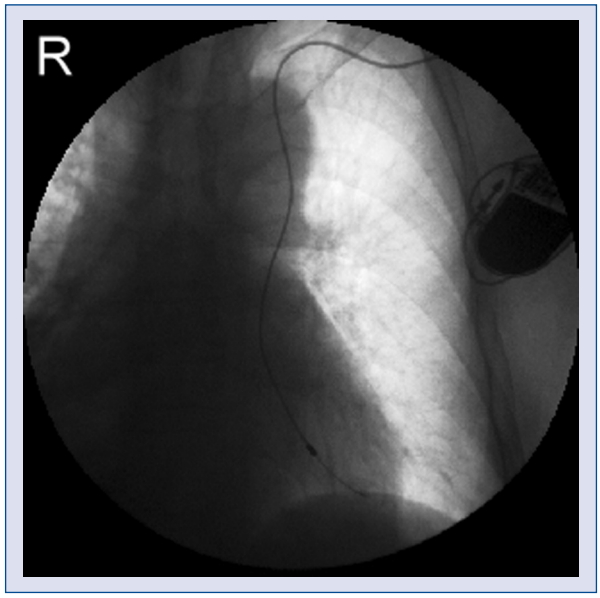

Figure 2. Postero-anterior chest radiograph demonstrates the ventricular lead in a postero-lateral cardiac vein (Case 3).

Angiography was performed to confirm a single, left sided superior vena cava.

Despite antiarrhythmic treatment with 3 different drugs, the patient was hospitalized and cardioverted several times for paroxysmal AF. Biatrial stimulation (BiA) was considered an alternative. In April 2001, a bipolar electrode with an active fixation was inserted into the CS to pace the LA. Both leads were connected by a Y-connector to the SSI pacemaker (Fig. 1). In the described case, the RA electrode was a cathode and the CS electrode was an anode. BiA pacing reduced P wave duration and the SQ from 180 to 120 ms and 220 to 160 ms, respectively. After the BiA system implantation, the patient was not hospitalized for paroxysmal AF and had only self-terminating palpitations. However, in 2005 his rhythm degenerated to a chronic AF.

Patient 2 (Case 2). A 73-year-old man with paroxysmal atrial flutter and AF followed by symptomatic sinus bradycardia and paroxysmal sinoatrial block was presented for PM implantation. His echocardiogram revealed left ventricular (LV) hypertrophy, mild mitral and aortic regurgitation. The dilated CS was noticed in a modified parasternal long-axis view with the diameter of 38 × 15 mm. The left sided approach was chosen for a PM implantation. The course of advanced lead indicated the presence of PLSVC. The attempt to reach RAA was not successful and a bipolar active fixation lead was screwed in RA free wall. Next, bipolar passive fixation lead was placed in the right ventricular outflow tract (RVOT). The presence of PLSVC was confirmed by angiography and the trans-esophageal echocardiography. No other malformations were found. Amiodarone (200 mg/day) was prescribed to prevent existing supra ventricular tachyarrhythmias with a good clinical effect.

Patient 3 (Case 3). A 75-year-old woman with a congestive heart failure and permanent AF was admitted due to syncope, palpitations, weakness and exercise intolerance. Holter monitoring documented many episodes of severe bradycardia < 35/min. Trans-thoracic echocardiography revealed mildly impaired LV ejection fraction (45% Simpson's method), mild mitral, aortic and tricuspid regurgitation. In addition, an enlarged CS was found. During the PM implantation, the lead was advanced along the left border of the sternum (Fig. 2). The electrode was intentionally placed into one of the cardiac veins on the posterior wall of the LV, without CS visualization, but only a stylet preforming, with good electrical parameters. Pacing at 10 V did not cause any diaphragmatic stimulation. Finally, the LV pacing with the right bundle branch block morphology in a standard 12-lead ECG was obtained. The diagnosis of PLSVC was confirmed by angiography. Clinical improvement and ventricular arrhythmia reduction were observed during the follow-up. In January 2013, after almost 10 years, the PM was changed due to a battery timeout.

Table 3. Estimation of arrhythmias and related symptoms during follow-up based on Holter monitoring.

	Case 1			Case 2			Case 3			Case 4		
	Before PMI	3 months after PMI	Follow up (95 months)	Before PMI	3 months after PMI	Follow up (152 months)	Before PMI	3 months after PMI	Follow up (115 months)	Before PMI	3 months after PMI	Follow up (81 months)
Symptoms of arrhythmia	Yes	Yes	No	Yes	No	No	Yes	No	No	Yes	No	No
NYHA class	I	I	I	II	I	I	III	II	II	III	II	II
SPB (per hour)	20	34	18	60	56	47	NA	NA	NA	NA	NA	NA
PVB	38	82	41	38	76	22	2273	898	746	61	75	125
Pairs of PVB	0	0	0	0	0	0	76	0	1	0	0	0
NSVT	0	0	0	0	0	0	4	0	0	0	0	0

NA — not applicable (patients with permanent atrial fibrillation); NSVT — non-sustained ventricular tachycardia, NYHA — New York Heart Association; PMI — pacemaker insertion; PVB — premature ventricular beats; SPB — supraventricular premature beats

Patient 4 (Case 4). A 75-year-old male was admitted to the hospital due to numerous presyncopal attacks and symptoms of congestive heart failure. His medical history included hypertension, permanent AF, diabetes mellitus, dyslipidemia and obesity (BMI = 35). Ambulatory ECG monitoring revealed many episodes of bradycardia < 40/min and pauses. The transthoracic echocardiography revealed dilated LV (end-diastolic diameter = 63 mm) with a normal ejection fraction, enlarged LA (52 mm) and a moderate mitral regurgitation. A passive fixation ventricular lead was inserted via the left subclavian vein, again the electrode advanced along the left edge of the sternum. Therefore, venography was performed intra-operatively confirming the presence of PLSVC draining to the large CS. The J-shaped stylet was used to introduce the lead via the tricuspid annulus to the RVOT. The patient was discharged and remained free of symptoms ever since. An anticoagulation therapy was continued to prevent thromboembolic complications.

Results

During the mean follow-up period of 110 months, pacing was effective. One patient (Case 1) required another operation and change of the pacing mode due to progressive interatrial conduction prolongation during the follow-up. No serious complications related to the PM were observed. The basic data concerning parameters of pacing are presented in Table 2. Selected information obtained from 24-h ECG monitoring, symptoms related to arrhythmias like syncope, palpitations, and functional class (NYHA) during the follow-up are presented in Table 3.

Discussion

Historically, the first diagnosis of PLSVC could be made only by means of autopsy but at present, this anomaly may be diagnosed by several vital diagnostic examinations. A trans-thoracic echocardiography is the most relevant method for PLSVC recognition, especially with a contrast injection. The enlargement of the CS present in echocardiography should lead to PLSVC suspicion. However, visualization of the CS by trans-thoracic echocardiography is not possible in about 20% of patients. A contrast trans-esophageal echocardiography is necessary to exclude any other congenital cardiac malformations concomitant with this anomaly [13]. Another simple and cheap method is venography [14], which may be performed intra-operatively during a PM implantation procedure. However, it should be avoided in patients with

severe renal dysfunction due to the risk of contrast nephropathy. In addition, magnetic resonance imaging, computerized tomography and first pass radionuclide angiocardiography are useful but rarely performed methods in evaluation of PLSVC [15]. Despite all this advanced diagnostic methods, PLSVC is most often recognized during medical procedures such as central venous catheter insertion, electrophysiological study, cardiac surgery and PM or cardioverter-defibrillator implantation. The presence of PLSVC may be suspected during the PM implantation on the basis of a left sided route of the electrode observed during fluoroscopy. In case of PLSVC, the proper positioning of a lead is difficult, the total time of procedure and the time of fluoroscopy are longer than usual.

Some authors suggest that PLSVC is coexistent with arrhythmias. Rhythm abnormalities were observed in 36% in a series of 121 published cases with PLSVC and absence of the right superior vena cava [16]. A few mechanisms leading to arrhythmias are distinguished:

— Histological changes in sinus and atrio-ventricular nodes. A hypoplastic sinus node was reported in cases of absent right superior vena cava. An atrio-ventricular node also may have hypoplastic, fragmented and disorganized histological structure. Therefore, a DDD pacemaker should be considered in all cases of PLSVC and a sick sinus syndrome. The proximal part of the great thoracic veins contains cardiac muscles connected to the atrium. Some of the cardiomyocytes have their own PM activity and may be responsible for the arrhythmic activity [17]. Hsu et al. [6] studied 5 patients with PLSVC and symptomatic drug refractory AF after a pulmonary vein isolation. Many electrical connections between PLSVC and CS and the LA were documented during an electrophysiological examination. Additionally, repetitive ectopic beats originating from PLSVC were observed. In each case, the ectopic beats initiated AF. Electrical activity of PLSVC and many electrical connections between PLSVC and the LA, and between PLSVC and CS were reported by Maruyama et al. [7] in a 25-year-old man with a Wolff-Parkinson-White syndrome, who underwent radiofrequency ablation. Thus, the electrical properties of PLSVC may play a role in the genesis of atrial tachyarrhythmias.
The prevalence of accessory pathways in the group of patients with PLSVC is higher than in general population [18]. Most often, these are left free wall and posteroseptal pathways, which are remnants of incomplete separation of myocardium between atria and ventricles by the annulus fibrosis during cardiogenesis.

— Prolonged interatrial conduction may constitute another problem in patients with PLSVC. It is known that delayed LA depolarization deteriorates hemodynamic function of the heart leading to concurrent (instead of sequential) LA and LV contraction and may induce AF. The interatrial conduction time may be prolonged before the PM implantation. Chan et al. [19] described a 63-year-old man with paroxysmal AF and widened P-wave (130 ms) in ECG. A DDD BiA pacemaker was implanted via PLSVC and the patient was free of AF recurrence without antiarrhythmic medications during the 6-month follow-up. Similar result was obtained in our patient no. 1, after upgrading to BiA pacing. A prolongation of interatrial conduction time may also appear after a typical RAA and especially RA free wall electrode placement. BiA pacing may prevent hemodynamic and electrical consequences. This should be considered in patients with PLSVC because it is difficult to place the atrial lead into the RAA, or septal position and CS ostium offers insufficient sensing. In our Case 1, the interatrial conduction time was long after PM implantation (P-wave duration 160 ms) and became even longer during the follow-up (200 ms). After changing the mode of pacing from AAI to AAT BiA the P-wave duration shortened to 120 ms. A clinically good result in prevention of paroxysmal AF was confirmed.
PLSVC as a sole anomaly has no influence on hemodynamics and does not take part in the development of heart failure directly. However, concomitant arrhythmias may precipitate a heart failure. Atrial tachyarrhythmias with a rapid ventricular rate reduce ventricular filing and cardiac output and may even lead to tachycardiomyopathy. On the other hand, marked bradycardia also depresses cardiac output. Depressed hemodynamic function and irregular rhythm may induce ventricular arrhythmias. The non-sustained ventricular tachycardia observed in Case 3 was probably induced by severe bradycardia and did not occur after PM implantation (Table 3).

One of the main purposes of our report was to demonstrate safety of the cardiac pacing via PLSVC in a long-term follow-up. Almost all cases presented in the literature have short follow-up periods,

which do not allow assessing the consequences of electrodes presence in PLSVC on the PM and heart function. Biffi et al. [20] implanted 2 PM and 2 cardioverter-defibrillators via PLSVC. After 41 months no device malfunction or lead dislodgement were observed. No complications occurred during the 4-year follow-up in another patient with DDD pacemaker implanted via PLSVC [21].

In Cases 2 and 4, ventricular leads were implanted into the right ventricle septal aspect of the outflow tract. This position of the lead seems hemodynamically more profitable than the classic right apical (RVA) pacing and is easier to achieve via PLSVC. In Case 3, the passive fixation bipolar lead was advanced into the left postero-lateral vein via PLSVC. This route was also used by Gaba et al. [22] during a DDD pacemaker implantation but an over-the-wire electrode was used after the CS venography. In our case, leads dedicated for the LV pacing were not available at that time.

Conclusions

1. The presence of PLSVC may complicate pacemaker's implantation.
2. Yet, it does not influence the long-term follow--up of pacing parameters.

Conflict of interest: none declared

References

1. Nsah EN, Moore GW, Hutchins GH. Pathogenesis of persistent left superior vena cava with a coronary sinus connection. Pediatric Pathology, 1991; 11: 261–269.
2. Niszczota CS, Koleśnik A, Clinical morphology of the abnormal pulmonary and systemic venous return. Pediatria Pol, 2012; 87: 293–304.
3. Tak T, Crouch E, Drake GB. Persistent left superior vena cava: Incidence, significance and clinical correlates. Int J Cardiol, 2002; 82: 91–93.
4. Giebel J, Fanghanel J, Hauser S et al. A case of a persistent left vena cava superior with atresia of the right atrial ostium of the coronary sinus. Anat Anz, 2000; 182: 191–194.
5. Huang SK. Persistent left superior vena cava in a man with ventricular fibrillation. Chest, 1986; 89: 155–157.
6. Hsu LF, Jais P, Keane D et al. Atrial fibrillation originating from persistent left superior vena cava. Circulation, 2004; 109: 828–832.
7. Maruyama M, Ino T, Miyamoto S et al. Characteristics of the electrical activity within the persistent left superior vena cava: comparative view with reference to the ligament of Marshall. J Electrocardiol, 2003; 36: 53–57.
8. Sarodia BD, Stoller JK. Persistent left superior vena cava: Case report and literature review. Respir Care, 2000; 45: 411–416.
9. Ashida Y, Mori T, Kuroda H et al. Transvenous dual chamber pacing via a unilateral left superior vena cava. PACE, 1998; 21: 137–139.
10. Waśniewski M, Mitkowski P, Ochotny R et al. Pacemaker or ICD electrode implantation in patients with persistent left superior vena cava. Cardiol J, 2006; 13: 154–158.
11. Corbisiero R, DeVita M, Dennis C. Pacemaker implantation in a patient with persistent left superior vena cava and absent right superior vena cava. J Interv Card Electrophysiol, 2003; 9: 35–37.
12. Kempa M, Lubiński A, Wilczek R et al. Cardioverter-defibrillator implantation in a patient with persistent left superior vena cava: A case report. Kardiol Pol, 2006; 64: 1281–1283.
13. Broka SM, Ducart AR, Collard EL et al. Persistent left superior vena cava: transoesophageal echocardiographic findings and surgical strategies. Acta Chir Belg, 1997; 97: 86–89.
14. Hsiao SH, Lee D, Hsu TL et al. Diagnosis of an isolated persistent left side superior vena cava by contrast echocardiography compared with invasive angiographic study. Zhonghua Yi Xue Za Zhi (Taipei) China, 2002; 65: 320–325.
15. Michałowska I, Kowalski M, Kwiatek P, Hoffman P. The evaluation of congenital heart diseases in adults: Can computed tomography supplement echocardiography? Pol Przegl Kardiol, 2010; 12: 291–296.
16. Bartram U, Praagh S, Levine JC et al. Absent right superior vena cava in visceroatrial situs solitus. Am J Cardiol, 1997; 80: 175–183.
17. Chen YJ, Chen YC, Yeh HI et al. Electrophysiology and arrhythmogenic activity of single cardiomyocytes from canine superior vena cava. Circulation, 2002; 105: 2679–2685.
18. Morgan DR, Hanratty CG, Dixon LJ et al. Anomalies of cardiac venous drainage associated with abnormalities of cardiac conduction system. Europace, 2002; 4: 281–287.
19. Chan NY, Lau CL. Implantation of a biatrial pacemaker in a patient with sinus venosus atrial septal defect and persistent left superior vena cava. J Cardiovasc Electrophysiol, 2006; 17: 104.
20. Biffi M, Boriani G, Frabetti L et al. Left superior vena cava persistence in patients undergoing pacemaker or cardioverter-defibrillator implantation: A 10-year experience. Chest, 2001; 120: 139–144.
21. Kilickap M, Altin T, Akyurek O et al. DDD pacemaker implantation in patient with persistent left superior vena cava and absent right superior vena cava: A four-year follow-up report. Can J Cardiol, 2005; 21: 1221–1223.
22. Gaba D, Kittusamy P Ho RT et al. Permanent pacing from a left ventricular vein in a patient with persistent left superior vena cava and absent right superior vena cava: use of an over-the-wire system. J Interv Card Electrophysiol, 2003; 9: 357–360.