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ORIGINAL ARTICLE

The prevalence of superior vena cava anomalies as detected in cardiac implantable electronic device recipients at a tertiary cardiology centre over a 12-year period.

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Abstract *Introduction:* The vast majority of cardiac implantable electronic device (CIED) recipients require transvenous lead insertion, which may be hindered by the presence of venous anomalies. The aim of this study was to determine the prevalence and variations of persistent left superior vena cava (PLSVC) and to conduct subsequent outpatient follow-up in terms of device function and the clinical condition of the recipients using data from CIED placement procedures conducted over a 12-year period.
Methods: The study population included patients undergoing first-time transvenous implantation of cardiac pacemakers and implantable cardioverter-defibrillators (ICDs). The presence of PLSVC was determined based on intra-procedure venography. Outpatient follow-up involved assessments of patient condition, radiological imaging, and CIED function.
Results: Of a total of 4708 CIED recipients, PLSVC was detected in eight patients (mean age 65.5 ± 13.9); five of them had double superior vena cava (DSVC), including three cases in which the vessels were bridged with a brachiocephalic vein (BCV). Three patients presented PLSVC associated with the absence of the right superior vena cava (RSVC), a very rare anomaly. Seven patients remain under observation, for a total of 78.4 ± 48.4 months of follow-up.
Conclusions: The rate of venous anomalies in the form of PLSVC detected in the evaluated population was 0.17%. These PLSVC cases were asymptomatic, which hindered their earlier

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detection. The presence of these anomalies made the procedures more challenging for the operator and increased the perioperative complication rates; however, neither patient condition nor CIED function was affected based on the long-term outpatient follow-up.

1. Introduction

Currently, the most common permanent cardiac pacing technique involves transvenous placement of cardiac pacemakers and implantable cardioverter-defibrillators (ICDs). Cardiac lead insertion is determined by both patency and favourable layout of the venous vascular system from the lead insertion site to final lead placement within the heart.[1,2] Intra-procedure detection of venous system variations that deviate from their typical anatomical layout may sometimes significantly affect the course of the procedure.[3–6] Unless such systemic vein anomalies co-occur with other clinically apparent congenital heart defects,[7] they typically remain undetected until certain cardiological and anaesthesiological procedures or diagnostic assessments are conducted for other reasons.[8–12]

The venous anomalies discussed here were diagnosed based on intra-procedure contrast-enhanced imaging revealing the presence of persistent left superior vena cava (PLSVC). Intra-procedure venography still remains the "golden standard" in venous diagnostics because it helps visualize vascular layout and morphology and helps determine the nature of the anomaly, which facilitates the selection of the optimal route of cardiac lead placement within the chambers of the right heart.[13] The goal of this study was to determine the prevalence and variants of PLSVC in cardiac electronic implantable device (CIED) recipients. The follow-up was conducted in an outpatient setting over a 12-year period and focused on the direct effects of the presence of PLSVC on the course of the procedure as well as on CIED function.

2. Methods

A total of 4708 CIED (i.e., cardiac pacemaker and ICD) recipients who underwent transvenous implantation between January 1, 2003, and December 31, 2014, were included in the study.

CIEDs were implanted in the infraclavicular region, and cardiac leads were inserted either via the cephalic vein (CV) cutdown and/or axillary vein (AV)/subclavian vein (SV) puncture approach. The cause of lead insertion difficulties and/or unusual lead position was investigated using contrast administration into the CV or directly into the AV or SV. Intra-procedure measurements of pacing parameters, such as action potential amplitude, stimulation threshold at a pulse duration of 0.5 msec and pacing impedance, were taken with device-specific readers. Post-procedure follow-up was continued in an outpatient setting and included periodic assessment of device function, location, and immediate surroundings, as well as the patient's clinical status.

3. Results

During the evaluated period, CIEDs were implanted transvenously in 4708 patients, 52% of which were female. In 97% of cases, the procedure was conducted via venous access from the left infraclavicular region. PLSVC was revealed in eight patients, including five women and three men, which constituted 0.17% of the study population (Table 1).

The PLSVC patient group included three different morpho-anatomical subtypes of this venous anomaly:

— three patients (cases number 1, 4, and 7) were diagnosed with double superior vena cava (DSVC) with a developed patent left brachiocephalic vein (LBCV) bridge connecting both vessels (Fig. 1)

— two patients (cases number 2 and 3) had DSVC without a brachiocephalic vein (BCV) bridge (Fig. 2)

!— three patients (see case reports 5, 6, and 8) had a single superior vena cava (SSVC), which is a PLSVC in the absence (agenesis) of the right superior vena cava (RSVC) (Fig. 3).

All of these PLSVC patients eventually received three single-chamber cardiac pacing systems, including one atrial (AAI type device — case 1) due to sinus node dysfunction and two ventricular (VVI type devices — cases 3, 6, and 7) due to third-degree (complete) atrioventricular block with sinus rhythm and chronic atrial fibrillation; in addition, four dual-chamber pacemakers (DDD type) were implanted due to tachycardia-bradycardia syndrome (cases 2 and 4) or complete atrioventricular block with sinus rhythm (cases 5). One patient (case 8) received an ICD-VR for the secondary prevention of ventricular tachycardia.

Venography conducted in three patients (cases 1, 4, and 7) because of an unusual course of the procedures (leads inserted through the PLSVC and the coronary sinus into the right atrium were unintentionally introduced into the RSVC) revealed the presence of DSVC with BCV, with contrast administration showing the precise layout of these vessels. In two patients (cases 2 and 3), the presence of DSVC without BCV was confirmed, independent of intra-procedure venography, via computed tomography angiography (CTA). A previous CTA performed in one patient (case 8) provided information about the presence and the type of LSVC prior to the procedure, which was confirmed via intra-procedure venography. In two patients (cases 5 and 6), lead position during lead introduction suggested the presence of PLSVC, and intra-procedure venography showed a lack of

Table 1 CIED implantation procedures in patients with PLSVC.

	Sex (F/M), age (years)	Venous anomaly	Indication for CIED implantation	Year of implantation, implanted device type and model	Lead model	Venous access
Case 1 (Fig. 1)	M, 41	DSVC with a BCV bridge	symptomatic sick sinus syndrome	2003, AAI, Biotronik Axios SR 2006, AAI, reimplantation due to lead damage - Vitatron C20SR	Biotronik SX-53-JBP /t/ Vitatron ICF09B /s/	left CV cutdown left AV puncture
Case 2 (Fig. 2)	F, 77	DSVC without BCV bridge	TBS (PAF / PSVT + sinus bradycardia)	2006, DDD, Medtronic Sigma SD303	Medtronic: A — 5592-53 /t/ V —5092-58 /t/	left AV puncture
Case 3	F, 80	DSVC without BCV bridge	complete AV block	2006, VVI, Medtronic Sigma SR 303	Medtronic 5092 -58 /t/	left AV puncture
Case 4	M, 61	DSVC with a BCV bridge	TBS (PAF + sinus arrest with escape nodal rhythm)	2007, DDD, St. Jude Medical Verity DR	St. Jude Medical: A-Tendril SDX-52 /s/ V-Tendril SDX-58 /s/	left AV puncture
Case 5	F, 52	PLSVC without RSVC	complete AV block in course of Wegener's granulomatosis	VI.2009, VVI, Biotronik Talos SR VII.2009, Biotronik Talos DR	Biotronik Selox ST /t/ Vitatron: A-Crystalline 52 /s/ V-Crystalline 58 /s/	right AV puncture
Case 6 (Fig. 3)	F, 67	PLSVC without RSVC	sustained AF with complete AV block	2013, VVI, Biotronik Ecuro SR-T	Biotronik Setrox S-60 /s/	left AV puncture
Case 7	F, 80	DSVC with a BCV bridge	sustained AF with AV block	2004, VVI, Medtronic Sigma SS203 2014, VVI, Medtronic Sensia SR	Medtronic 5092-58 /t/ St. Jude Medical Tendril STS 58 /s/	left AV puncture
Case 8	F, 66	PLSVC without RSVC	Turner syndrome with LQTS and *torsade des pointes*	2014, ICD-VR, Medtronic Cardia VR	Medtronic 6935-65 /s/	left AV puncture

F — female; M — male; CIED — cardiac implantable electronic device; DSVC — double superior vena cava; BCV — brachiocephalic vein; PLSVC — persistent left superior vena cava; RSVC — right superior vena cava; TBS — tachycardia-bradycardia syndrome; PAF — paroxysmal atrial fibrillation; PSVT — paroxysmal supraventricular tachycardia; AV — atrio-ventricular; AF — atrial fibrillation; LQTS — long-QT syndrome; lead model: A — atrial; V — ventricular; lead tip: s — screw-in; t — tined; CV — cephalic vein; AV — axillary vein

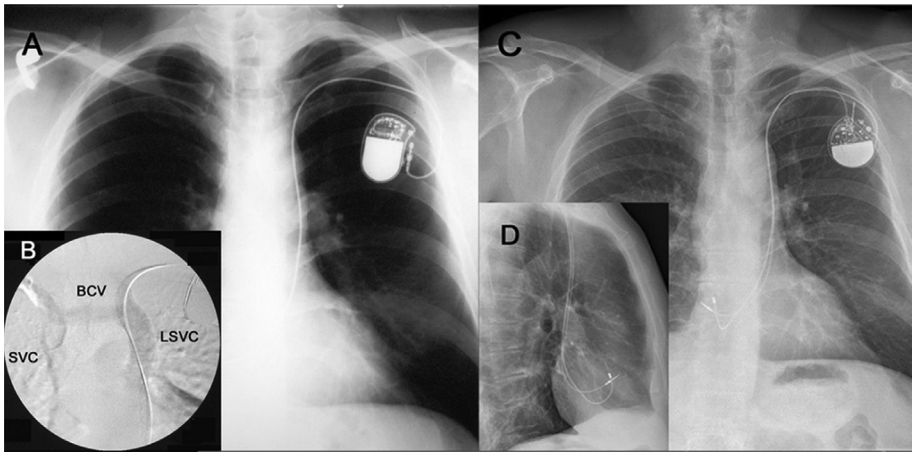

Figure 1 **(A, B, C, D). Case 1. (A)** Chest X-ray, posterior-anterior (PA) view, showing the atrial lead implanted in September 2003; **(B)** Venography showing contrast flow through both SVCs and the BCV connecting them (2003); **(C, D)** An X-ray film (January 2013) also showing the new lead implanted in May 2006, in **(C)** PA and **(D)** lateral views.

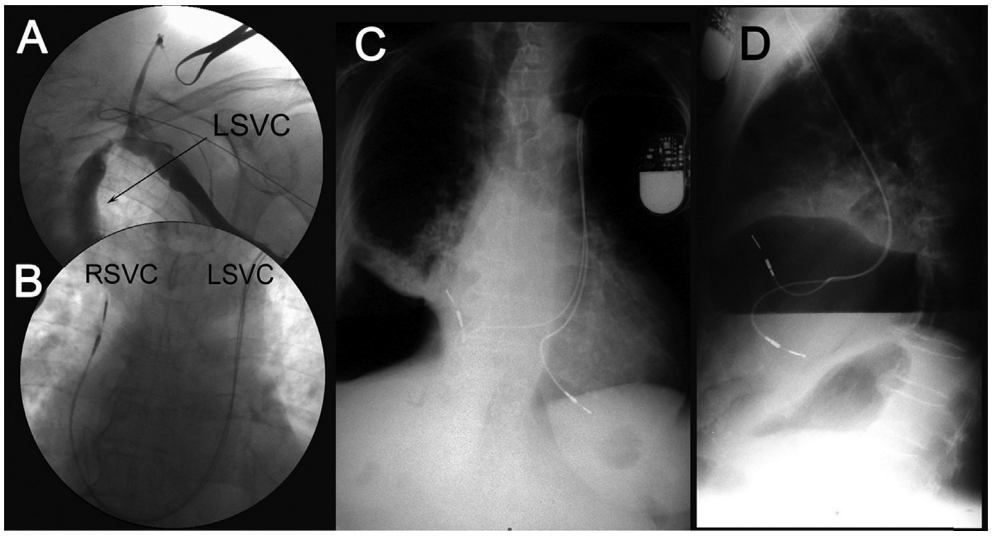

Figure 2 (A, B, C, D). Case 2. Intra-procedure fluoroscopy (Feb. 2006): contrast administration revealed LSVC without BCV. **(A)** Lead manipulation showed the presence of RSVC. **(B)** Chest X-ray from hospitalization for pneumonia (Jan. 2013). Massive opacities in the right middle lung field with parietal pleura thickening, pronounced adhesions, evidence of pneumothorax around the right lower lung field, and an air-fluid level in the right pleural cavity **(C, D)**.

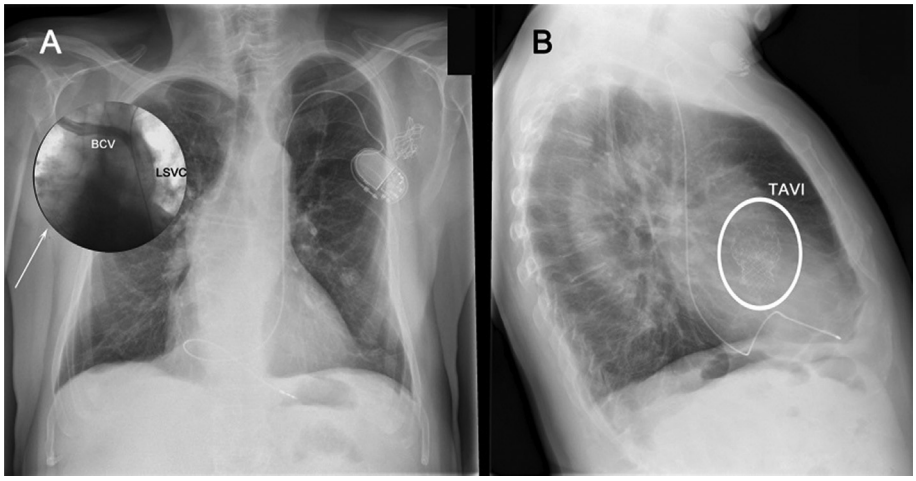

Figure 3 (A, B). Case 6. Radiographic image of lead position in PLSVC (arrow →, venography reveals a lack of RSVC) and the position of the implanted 29-mm aortic Medtronic Core Valve in **(A)** posterior-anterior and (**B**) lateral views.

RSVC. One of the patients described below (case 5) was diagnosed with PLSVC intra-procedurally at another centre. Ineffective stimulation by a device implanted from the left-side approach determined the need to repeat the procedure, this time via venous access through the right SV and BCV. Lead insertion into the cardiac chambers through the RSVC was impossible as the CTA showed evidence of the vessel's agenesis. In all of the PLSVC cases, venous blood collected by the coronary sinus (CS) drained into the right atrium. None of these patients showed any evidence of a congenital heart defect in post-procedure transthoracic echocardiography.

Three patients developed complications requiring reoperation. Two patients had cardiac lead displacement in the early postoperative period (in week 1 — case 2 and in week 2 — case 5); in both cases tined (passive-fixation) leads had been used. However, in the third patient (case 1), the lead was damaged as a result of trauma three years following pacemaker implantation, which necessitated another lead implantation. Following the required lead placement procedures, pacing parameters recorded during subsequent scheduled outpatient visits showed consistently normal stimulation thresholds, lead resistance, and impulse amplitude, as in the other patients.

The procedures described in this paper were characterized by a longer patient and staff X-ray exposure time, ranging from 1.75 min to 40.2 min [mean $\pm$ standard deviation (SD) 13.95 min $\pm$ 13.7 min], whereas X-ray exposure duration in 100 similar procedures in patients without venous anomalies varied from 0.12 min to 7.08 min (0.98 ± 1.1 minutes; $p < 0.005$).

Total outpatient follow-up duration from the time of the procedure until study follow-up completion (Dec. 31, 2014) ranged from 2 to 134 months (78.4 ± 48.4 months). None of the patients developed symptoms suggesting ipsilateral venous thrombosis.

4. Discussion

Venous vascular system development is characterized by a much greater variety than that of arterial vessels. In asymptomatic cases, a venous anomaly can be detected only in favourable circumstances. For example, PLSVC can be detected during transvenous implantation of a permanent cardiac pacing device.[1,8,14,15] The prevalence of persistent left superior vena cava in the population is approximately 0.3—0.5%, and the anomaly is usually found incidentally during post-mortem examinations, invasive cardiological procedures, cardio-thoracosurgical procedures or echocardiography.[9,11,12,16] The prevalence of this systemic vein anomaly in the evaluated patient population was 0.17%, which was slightly lower than that reported in the literature.

PLSVC draining into the left atrium, with the presence of a left-right shunt is found sporadically, usually in association with severe, symptomatic cardiovascular pathologies, such as unroofed coronary sinus defect type 1.[1,17,18]

The prevalence of PLSVC is higher when the anomaly coincides with congenital heart defects (3—10%), such as ventricular septal defect (VSD), atrial septal defect (ASD), pulmonary valve stenosis, common atrioventricular canal, dextrocardia, and triatrial heart.[5,11,19] In the cases described here, transthoracic echocardiography assessments showed no congenital heart defects. The observed individual vascular variations resulting from embryological development anomalies can be classified into three main anatomical forms of PLSVC: DSVC either with (Fig. 1) or without (Fig. 2) a connection via the innominate vein and a single PLSVC (SLSVC) with an underdeveloped RSVC (Fig. 3).[8,20—23]

A considerable majority of our patients underwent their procedures via a left-sided infraclavicular approach. This technique increases the likelihood of detecting a PLSVC-like systemic venous anomaly and leads to a more accurate estimate of its actual prevalence.

Patent DSVC can be found in approximately 85% of adults with PLSVC. Our data showed that PLSVC was accompanied by RSVC in 5 of 8 (62.5%) cases. However, the prevalence of DSVC with a bridging BCV (cases 1, 4, and 7) seems to be somewhat higher. This is most likely because this anomaly remains undetected in patients in whom the angle of BCV allows for lead insertion into RSVC, and it is only via venography that this systemic vein anomaly can be detected (case 7).

The absence of vascular bridge formation between both anterior cardinal veins (approximately 65%) leads to a lack of the innominate vein; this rate is higher than that observed in our study [two of five cases (33%)].[7,12]

!Developmental anomalies of the right common cardinal vein that result in the absence of RSVC occur in 10—20% of cases; the pertinent rate in our study being 37% (cases 5, 6, and 8). This venous configuration is hemodynamically significant, as the blood draining from the upper part of the body into the right atrium can flow only through PLSVC.[7,8,12]

The presence of PLSVC as well as its morphology (confluence angles, valves) hinders lead manipulation and placement in the right ventricle, as it requires negotiating the acute angle between the orifice of the coronary sinus and the tricuspid valve.[24] This manoeuvre may be facilitated by moulding the lead into a suitable shape, usually resembling the Greek letter alpha (α).[4,24]

The procedures presented here were characterized by a longer than usual patient and personnel exposure to X-rays. This was a result of both the non-routine cardiac lead insertion technique and the need for precise visualization and radiographic documentation of the detected PLSVCs.[8]

The increased challenge of non-routine cardiac lead insertion may also affect intra-procedure pacing parameters and contribute to postoperative problems (cases 2, 3, and 5).

The use of CTA either prior to or after the procedure helps accurately locate and assess the layout of systemic veins against the adjacent structures and sometimes helps detect significant concomitant pathologies.[25,26] CTA allowed for the accurate diagnosis of a left lung tumour detected during a CIED implantation procedure in one of our patients (case 5). This case was later elaborated in a separate report due to the rarity of atrioventricular conduction disturbances caused by Wegener's granulomatosis.[27]

The outpatient follow-up is ongoing in seven patients with PLSVC, and the conducted assessments confirm normal pacing parameters and lead placement. In January 2013, after 84 months of follow-up, one patient died as a result of cardiopulmonary failure associated with upper respiratory tract inflammation.

The literature mostly contains reports of CIED implantation in individual patients with PLSVC, only rarely describing larger patient groups and addressing variations .[13—15,28] Our study presents eight patients with PLSVC who received a CIED in a single centre during a 12-year period. Three of these patients had DSVC with a bridging BCV, two had DSVC without a venous bridge, and three had the rare variation of PLSVC with absent RSVC. This article presents morpho-anatomical variations of systemic vein anomalies and is one of only a handful of manuscripts that include postoperative follow-up.

5. Conclusions

The prevalence of PLSVC in our patients, 0.17%, was slightly lower than that reported in the literature. The presented PLSVC cases were asymptomatic, which hindered their earlier diagnosis. Although the presence of PLSVC made the implantation procedures more challenging, follow-up assessments showed that this had no effect on the long-term function of the implanted devices.

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