Objective: The persistence of a left superior vena cava (LSVC) has been observed in 0.3% of the general population as established by autopsy. In the adult population, it is an important anatomic finding if a left superior approach to the heart is considered. The aim of the study was to evaluate the prevalence of a LSVC in patients undergoing pacemaker (PM) and cardioverter-defibrillator (CD) implantation.

Design: We observed the prevalence of LSVC during a 10-year period; each patient undergoing PM or transvenous CD implantation received a left cephalic/left subclavian venous approach to the heart. With this technique, LSVC persistence is easily diagnosed during lead placement.

Results: A total of 1,139 patients consecutively underwent PM implantation during 10 years: 4 patients had persistent LSVC (0.34%). Among 115 patients undergoing CD implantation, 2 patients with LSVC (1.7%) were observed. Overall LSVC persistence was found in 6 of 1,254 patients (0.47%). Two patients, one of whom had no right superior vena cava (RSVC), received a left-sided PM, whereas two other patients received right-sided devices. Both CD patients received a left-sided active-can device: the first patient with a right-sided lead tunneled to the left pectoral pocket, as a result of poor catheter handling through the LSVC and coronary sinus, and the second patient with a screw-in lead from LSVC. Long-term follow-up of these patients (average ± SD, 41 ± 26 months) revealed absence of lead dislodgment and appropriate device function regardless of lead implantation site.

Conclusions: Persistence of LSVC in adults undergoing PM/CD implantation is similar to that of the general population (0.47% in our study). The left-sided implant can be achieved by stylet shaping and by use of active fixation leads in most patients, with a reliable outcome at short term in addition to appropriate device performance at follow-up. Assessment of the RSVC is advisable when planning a right-sided implantation, since a minority of patients lacks this vessel.

Key words: left superior vena cava persistence; pacemaker/defibrillator lead

Abbreviations: AV = atrioventricular; CD = cardioverter-defibrillator; LSVC = left superior vena cava; PM = pacemaker; RSVC = right superior vena cava; SSS = sick sinus syndrome; VT = ventricular tachycardia

The persistence of a left superior vena cava (LSVC) is due to an abnormal development of the sinus venosus in the early stages of fetal life. In the 4-mm embryo, this structure consists of three distinct parts, namely, the right horn, the transverse part, and the left horn, and collects three pairs of veins (the omphalomesenteric veins, the umbilical veins, and the common cardinal veins). Due to the rightward direction of blood flow, the right horn undergoes preferential growth and ultimately constitutes the intercaval (nonmuscular) part of the right atrium.1,2

The transverse part and the proximal left horn of the sinus venosus invaginate from the left atrium and form the coronary sinus. The distal left horn and the left cardinal vein obliterate, and form the so-called ligament of Marshall1,2 in the adult subject. In the case of patency of the left cardinal vein, its drainage reaches the right atrium through the coronary sinus, which becomes enlarged to variable degrees. In 92% of cases, drainage occurs in the right atrium; in the
Persistence of LSVC has been reported to occur in approximately 0.3% of the general population in a single, large report (> 4,000) of unselected autopsies. Its prevalence is in fact much higher in patients with congenital cardiac abnormalities compared to the general population, ranging from 2.8 to 4.3%. In addition, about 10% of these subjects with congenital cardiac abnormalities do not have a right superior vena cava (RSVC). When isolated, LSVC persistence is usually not recognized until a left superior approach to the heart is required, when it becomes a relevant anatomic finding. In fact, it can complicate the positioning of left-sided pacemaker (PM) and cardioverter-defibrillator (CD) leads, the placement of central venous lines for therapeutic purposes and hemodynamic monitoring, and cardiopulmonary bypass in patients undergoing cardiac/thoracic surgery procedures.

Isolated cases of LSVC persistence in patients undergoing PM or CD implantation have been reported in literature, the most common problems related to the unusual anatomic access to the heart were reaching a convenient pacing site and ensuring stable lead placement. This background activated our center to prospectively investigate the prevalence of LSVC persistence in adults undergoing either PM or CD implantation over a 10-year period, and to evaluate the relative benefits of the different approaches adopted by the distinct physicians to overcome implant-related difficulties.

### Materials and Methods

The study was prospectively conducted from October 1989 to September 1999 on all patients undergoing PM implantation; from January 1992 to September 1999, patients undergoing transvenous CD implantation were also taken into consideration. Patients with congenital thoracic-abdominal abnormalities were not considered due to the likelihood of associated LSVC persistence; this would have increased the prevalence in the patients with bradyarrhythmias and tachyarrhythmias. Patients requiring generator replacement and patients with known thrombosis of the left subclavian vein were not considered in the study.

From October 1989, all device implantations were consecutively carried out using the left-sided approach. In case of LSVC persistence, the guidewire (or the lead in case of cephalic access) enters the subclavian vein and does not cross the column, but lays parallel to it before entering the coronary sinus posteriorly to the left atrium. After entering the right atrium, the passage into the tricuspid orifice is accomplished from the posterior to the anterior and then leftwards, almost on the same horizontal plane, and requires skillful shaping of the stylet depending on right-heart size and geometry. The access to the right atrial appendage goes downwards, then upwards, and then anteriorly.

The physicians defined different steps to overcome implant-related difficulties: hand shaping the stylet in a predefined fashion, using active fixation leads, and moving to a right-sided implant after right peripheral venography by nonionic medium (iomeprol, 20 mL) or echocardiography. The practical outcome of the different policies was assessed by lead performance, total fluoroscopy time, and total procedure time.

### Statistical Analysis

Comparisons of total fluoroscopy times and total procedure times were performed using unpaired t tests.

### Results

During 10 years, 1,147 consecutive patients without congenital abnormalities underwent PM implantation for commonly accepted indications; 8 patients were occasionally discovered to have asymptomatic left subclavian thrombosis (angiographically documented during implantation) and were hence excluded from analysis due to the impossibility of ruling out LSVC persistence. Of the other 1,139 patients, 627 patients were male (55%) and 512 were female (45%); 4 of 1,139 patients had patency of LSVC (0.34%). In the period from 1992 to 1999, 115 consecutive patients (84 male [73%] and 31 female patients [27%]) underwent transvenous CD implantation; of these patients, LSVC persistence was found in 6 of 1,254 patients (0.47%) patients, all of whom were male.

The first patient (aged 57 years) had sick sinus syndrome (SSS) with episodic sinus arrest; LSVC persistence was recognized during lead advance by the left cephalic vein. An elective right-sided implant was performed after visualization of the RSVC by contrast venography (Table 1). Over 94 months of follow-up, this patient was asymptomatic with median pacing rate hysteresis.

The second patient (aged 60 years) had SSS. LSVC was recognized while advancing a standard bipolar ventricular lead through the left cephalic vein. Placement in the right ventricular apex was accomplished by a hand-shaped stylet consisting of a distal rounded “L” following a proximal leftward sharp bending (Fig 1, middle, B; Fig 2). Placement into the right appendage was successful with a rounded L-shaped stylet (Fig 1, top, A) and an active fixation lead (Fig 2). A normal RSVC entering the right atrium was visualized by contrast venography after implantation.

The third patient (aged 58 years) had SSS and mitral valve prolapse; LSVC was diagnosed after subclavian vein puncture by anomalous wire course. This patient was treated by shaping stylets in the same way as was the second patient (Fig 1) but received active fixation leads in both chambers. This patient had no RSVC at venography performed at the end of implantation.
The fourth patient (aged 65 years) had complete right bundle-branch block, second-degree atrioventricular (AV) block, and episodic complete heart block following an old inferior myocardial infarction. LSVC was diagnosed at subclavian vein puncture. The physician moved to a right-sided implantation after visualization of the RSVC by contrast venography. This patient was PM-dependent 6 months after implantation.

Both CD patients had dilated cardiomyopathy due to coronary artery disease and previous myocardial infarction.

The fifth patient (aged 58 years) had been rescued five times from monomorphic ventricular tachycardia (VT) while screening for heart transplantation. CD implantation was planned as a bridge to heart transplantation. LSVC was diagnosed following subclavian puncture; passage through the right cardiac chambers was difficult, and the lead neither reached the right ventricular apex nor demonstrated stable or reliable sensing. Then, the physician changed to another implantation strategy. The RSVC was visualized by echocardiography at the bedside in the operating room. The patient received a left pectoral active-can device (housing can takes active part into the defibrillation process) but with a right-sided lead tunneled to the left pectoral pocket. This patient eventually underwent heart transplantation 6 months later, having been rescued once more from fast VT. This patient is not considered in long-term follow-up.

The sixth patient (aged 64 years) had right bundle-branch block and sustained monomorphic VT unresponsive to drug therapy. He received a pectoral left-sided active-can device, with an active-fixation, single-coil lead passing through the LSVC and coronary sinus. Placement in the right ventricular apex (Fig 3) was accomplished by a hand-shaped stylet (Fig 1, middle, B). Right-sided venography performed during implantation demonstrated absence of the RSVC.

In this experience, total fluoroscopy time (mean ± SD) was 526 ± 83 s for left-sided implants and 766 ± 538 s for right-sided implants, respectively (p = not significant). Total procedure time was 160 ± 26 min for left-sided implants and 203 ± 15 min for right-sided implants, respectively (p = 0.07). Active fixation leads were used in two of two patients at the atrial site, and in two of three patients at the ventricular site in those with left-sided implantation. Absence of the RSVC was observed in two of six patients (33%).

Five patients were followed up for 41 ± 32 months on average; absence of lead dislodgment and appropriate function either for PM or CD patients regardless of lead implantation site were observed (Table 1). Follow-up times for leads placed through LSVC and coronary sinus were 46 months, 26

![Figure 1](http://journal.publications.chestnet.org/pdffacessashx?url=/data/journals/chest/21964/ on 04/15/2017)
months, and 20 months, respectively, whereas they were 94 months and 18 months for those placed through the RSVC. Lead integrity was 100%, as assessed by telemetry system check.

**DISCUSSION**

Persistence of LSVC in adult life is normal in rabbits and some other mammals, but it is a rare abnormality in man. Besides being associated with congenital diseases, its most relevant clinical implication is the association with disturbances of cardiac impulse formation and conduction. The ontogenetic development of the sinus node, the AV node, and the His bundle may be heavily influenced by the lack of regression of the left cardinal vein, since these structures are located at the junctions of the right and left cardinal veins with the sinus venosus. Persistence of LSVC and/or anomalies of RSVC have been described to alter the location and also the histologic organization of both the sinus

---

**Figure 2.** Radiographic views of lead placement into the right atrial appendage and right ventricle in a patient with SSS: anteroposterior view (top, A), right anterior oblique (middle, B), and left anterior oblique (bottom, C).

**Figure 3.** Radiographic views of lead placement into the right ventricle in a patient with VT, LSVC persistence, and absent RSVC: anteroposterior view (top, A), right anterior oblique (middle, B), and left anterior oblique (bottom, C).
node and the AV junction,\textsuperscript{24} causing small and poorly formed sinus node, fetal dispersion of the AV node and His bundle within the central fibrous body, small diameter of the His bundle, and poor arterial supply to either the AV node or the sinus node.\textsuperscript{25,26} This pathologic substrate may predispose patients to arrhythmias and also to sudden death.\textsuperscript{25,26}

\textbf{LSVC Prevalence}

The prevalence of LSVC in general population is about 0.3\%, as established by an autopsy study\textsuperscript{4}; however, to our knowledge, the prevalence in patients with symptomatic bradyarrhythmias requiring permanent cardiac pacing has never been investigated before. In our study, we observed a similar, although slightly higher, prevalence of LSVC persistence compared to general population as assessed by unselected autopsies\textsuperscript{4}; this may be viewed as lower than what expected \textit{a priori}, given the association with abnormalities of impulse formation and conduction.\textsuperscript{20–26} In our study, RSVC was absent in 33\% of patients, compared to an average 10\% in other reports.\textsuperscript{4,7,9} This difference may be due to the low prevalence of RSVC absence in each study. Indeed, this anatomic finding is of key importance from a surgical point of view (in this situation, the left vascular axis constitutes the only superior access to the heart) and implies a nontransvenous implantation in case of failure from the LSVC.

The finding of a LSVC complicating placement of left-sided PM or CD systems has been reported as a sporadic observation in literature, mainly in isolated patients. Of a total of 661 patients undergoing VVI-mode PM system implantation, Zerbe et al\textsuperscript{16} reported 4 patients with LSVC persistence, but the observation was retrospective and no systematic attempt to assess the prevalence of this abnormality by consecutive left-sided approach to each PM implantation was made. The customary left-sided approach adopted in our center allowed easy diagnosis of LSVC persistence and prevented missing “false-negatives,” thus giving a true estimate of its prevalence in this cardiology population.

\textbf{Practical Implications}

Different techniques have been used to obtain a reliable function of the implanted system in these patients,\textsuperscript{10–13,15–18} minimizing exposure to radiation and risk of lead dislodgment.\textsuperscript{10–12} Many authors found helpful ways to shape stylesets to enter the tricuspid valve or to reach the right atrial appendage\textsuperscript{15–17}; active fixation leads were used to ensure lead stability,\textsuperscript{17,19} although this was not mandatory in all the cases.

In our experience, lead placement by the left approach through the coronary sinus was feasible with reliable results. We suggest use of active fixation leads in the right appendage, guided by a large-curve L-shaped stylet (Fig 1, top, A). Careful shaping of the stylet (Fig 1, middle, B) allowed an easy access to the right ventricular apex in three of four patients in whom it was attempted. A standard ventricular lead was used in one patient with success; however, active fixation leads may be used when diagnosis is made before lead insertion.

In patients with poor handling through the coronary sinus, a right approach is recommended after visualization of a right vena cava entering the right atrium by echocardiography\textsuperscript{27} or contrast venography; the absence of RSVC would suggest an epicardial implantation. In our experience, successful implantation by the left side did not require significantly longer fluoroscopy time compared to a right-sided implantation except for the first CD patient. Crossing over to a right-sided approach requires a longer procedure time, echocardiography or contrast venography to ensure RSVC presence, and thus may cause greater discomfort to the patients. In fact, the two patients who crossed over to the right approach in “elective” manner (no attempt from the left side after diagnosis) had longer procedure times (190 min and 200 min, respectively) compared to average procedure time of the left-sided approach (160 ± 26 min). We therefore suggest that implantation be accomplished left sided when diagnosis is made intraoperatively, unless catheter handling appears difficult or fluoroscopy time exceeds a defined maximum.

In patients undergoing CD implantation, devices with outer shell taking part into the defibrillation process (termed active can or hot can, depending on different manufacturers) have lower defibrillation current requirements when placement is left sided compared to right-sided placement.\textsuperscript{28,29} Current CD generation is capable of achieving acceptable defibrillation thresholds also when placed right pectoral,\textsuperscript{30} so that tunneling the lead to a left subclavian pocket is unnecessary in several cases. A left-sided implantation may be mandatory in the minority of patients with high defibrillation thresholds from the right-sided configuration to improve the effectiveness of heart defibrillation. We suggest trying a transvenous left-sided approach first, and considering a right venous access in case of failure to reach a convenient site.

Relevant implications of our observation may apply to the fields of critical care, anesthesiology, general and thoracic surgery, oncology, and hematology when central lines for monitoring or therapeutic purposes are required\textsuperscript{12,14} or when permanent catheters for drug delivery are implanted. Awareness
of this venous anomaly may obviate unnecessary catheter removal and troublesome placement of a new one when arterial puncture is suspected by imaging techniques, but not obvious at bedside verification tests. In addition, assessment of the RSVC may be very important in these patients, since its absence may represent a major obstacle in providing care of critical patients.14

In conclusion, persistence of LSVC in patients undergoing PM/CD implantation is similar to that in the general population (0.47% in our study). Specifically, shaped stylets and active fixation leads are helpful tools to overcome technical difficulties in ordinary procedure times. Right-sided implantation is easily feasible once the presence of a RSVC has been confirmed either by echocardiography or by peripheral venography, since a substantial minority of these patients (two of six patients in our study) lacks a right-sided superior venous access to the right atrium.

REFERENCES

17. Rusk PA, Bexton JM, McComb JM. Persistent left sided and absent right sided superior vena cava complicating pacemaker insertion. Heart 1996; 75:413
23. Van Mierop LHS, Patterson PR, Reynolds RW. Two cases of congenital asplenia with isomerism of the cardiac atria and the sinoatrial nodes. Am J Cardiol 1964; 13:407