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Short Original Article

Superior Vena Cava Malformations: Diagnosis and Technique for Endocavitary Lead Implantation in Permanent Cardiac Pacing

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The authors declare no competing interests.

Abbreviations
CAVB: complete atrioventricular block
CT: computed tomography
PLSVC: persistent left superior vena cava
PSVT: paroxysmal supraventricular tachycardias
RA: right atrium
RSVC: right superior vena cava
RV: right ventricle/ventricular

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ABSTRACT

Introduction: *The implantation of endocavitary leads for permanent cardiac pacing requires specialized skills and knowledge regarding the anatomy of the superior venous vessels that drain into the right atrium. The superior vena cava typically courses to the right of the vertebral column before draining into the right atrium. However, it may present anatomical variants —such as a Persistent Left Superior Vena Cava (PLSVC)— that pose a challenge for the specialist.*

Objective: *To describe the clinical and imaging characteristics of patients with superior vena cava system malformations due to the technical difficulties these present for endocavitary lead implantation.*

Method: *Five patients, aged 36 to 72 years, were studied. These patients were referred for pacemaker implantation, but the initial procedure was unsuccessful due to anatomical and technical challenges. An imaging study protocol was established and applied on a case-by-case basis.*

Results: *Three patients were diagnosed with PLSVC plus absence of the right superior vena cava, and two others with a dual superior vena cava. Novel electrode implantation techniques, not previously described in the literature, were established and successfully applied to achieve endocavitary implant success.*

Conclusions: *Knowledge of the anatomical variants of this malformation, the diagnostic modalities, and the techniques for endocavitary lead implantation are of vital importance for achieving procedural success. Specialists who identify that the electrode descent is via the left paravertebral region should establish similar diagnostic protocols and master these implantation techniques.*

Keywords: *Congenital malformations, Superior vena cava, Persistent left superior vena cava, Pacemaker implantation*

Malformaciones de vena cava superior: diagnóstico y técnica para el implante de electrodos endocavitarios en la estimulación cardíaca permanente

Authors' Contribution

ECG and LES: Conception, conceptualization, and study design; literature review; data acquisition, curation, and formal analysis; and manuscript drafting.

LFVF, NRLP, and RIA: Data acquisition and analysis; assistance with image acquisition and reconstruction; and support in manuscript drafting.

FRG and JMCE: Study conception and design; data analysis; and critical revision of the manuscript.

AEHC: Data analysis and manuscript review.

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RESUMEN

Introducción: La implantación de electrodos endocavitarios para la estimulación cardíaca permanente requiere habilidades y conocimientos sobre la anatomía de los vasos venosos superiores que drenan en la aurícula derecha. La vena cava superior normalmente discurre por la derecha de la columna vertebral hasta desembocar en la aurícula derecha. Sin embargo, puede presentar variantes anatómicas —como la vena cava superior izquierda persistente (VCSIP)— que representan un reto para el especialista.

Objetivo: Describir las características clínicas e imagenológicas de los pacientes con malformaciones del sistema de las venas cava, debido a la dificultad que esto implica en la implantación de electrodos endocavitarios.

Método: Se estudiaron cinco pacientes entre 36 y 72 años de edad, quienes fueron referidos para el implante de marcapasos, pero las dificultades anatómicas y técnicas impidieron el procedimiento inicial. Se estableció un protocolo de estudio de imágenes según cada caso.

Resultados: Se diagnosticaron tres pacientes con VCSIP más ausencia de vena cava superior derecha y otros dos con doble vena cava superior. Se establecieron y aplicaron técnicas de implantación de los electrodos, no descritas previamente en la literatura, para lograr el éxito del implante endocavitario.

Conclusiones: El conocimiento de las variantes anatómicas de esta malformación, las formas de diagnóstico y las técnicas para el implante de los electrodos endocavitarios son de vital importancia para lograr el éxito del procedimiento. Los especialistas que identifiquen que el descenso de los electrodos es por la región paravertebral izquierda deben establecer protocolos diagnósticos similares y dominar las técnicas de implante.

Palabras clave: Malformaciones congénitas, Vena cava superior, Vena cava superior izquierda persistente, Implantación de marcapasos

INTRODUCTION

Since the late 1950s, various types of cardiac pacing devices have been rapidly developed, along with the indications for their implantation. Currently, access to the superior venous system (vena cava) is sometimes required to place up to three electrodes through it (e.g., in cardiac resynchronization therapy), which usually drain into the right atrium (RA). In most countries, electrophysiologists are the specialists who daily face the challenges of implanting endocavitary leads.

During embryonic development, the superior venous system of the embryo and fetus undergoes transformations. Normally, at birth, each individual has a single right superior vena cava (RSVC) that receives venous drainage from the upper body. The failure of these embryonic changes leads to malformations of the superior venous system, which include: 1) the presence of a dual superior vena cava, 2) a persistent left superior vena cava (PLSVC) with the absence of the RSVC, or 3) the absence of both vena cava.

The presence of these malformations can pose a challenge for specialists implanting electrodes, catheters, or other prostheses that require access to the superior venous system. Occasionally, when endocardial access via the superior venous system is impossible for pacemaker implantation, the patient undergoes surgery for an epicardial lead implant or, as described, the femoral route is used for endocavitary access[1].

PLSVC with the absence of the RSVC is an extremely rare anatomical variant that is generally associated with other congenital diseases or alterations in heart position. It is essential to have a thorough knowledge of this anatomical variant to manage patients undergoing invasive procedures of the superior venous system[2-5].

Suspicion of these superior venous system malformations almost always occurs accidentally during the intraoperative procedure. In the case of pacemaker lead implantation, immediate suspicion is warranted when the lead, via either access route (cephalic or subclavian vein, right or left), "capriciously" follows a trajectory to the left of the vertebral column. However, this observation often stops the specialist, and the implant is considered a failure.

The use of imaging modalities, such as echocardiography, computed tomography (CT), magnetic resonance imaging, and angiography, is useful in diagnosing anatomical variants. This allows the specialist to decide on the most appropriate technique for advancing the intracavitary leads during pacemaker implantation[4-6].

We aim to describe the clinical and imaging characteristics of patients referred to our department due to difficulties in endocavitary lead implantation, with a final diagnosis of vena cava system malformation.

METHOD

A descriptive, observational, cross-sectional study was conducted from January 2019 to October 2023. The study included seven patients referred for difficulty in implanting endocavitary leads for permanent cardiac pacing to the territorial electrophysiology department of the Ernesto Guevara Cardiocenter in Santa Clara, Cuba. This department covers the needs of the provinces of Cienfuegos, Villa Clara, Sancti Spíritus, Ciego de Ávila, and Camagüey.

The sample consisted of five patients with a definitive diagnosis of vena cava system malformation. CT scanning or hemodynamic study (venous angiography) was used for diagnostic confirmation.

Information Collection

Data for the present investigation were obtained through interviews and individual clinical records, which were used to create a collection form.

The variables studied were: age, sex, the diagnosis that motivated permanent pacemaker implantation, anatomical characteristics of the anomalous venous system, and imaging tests used to corroborate the diagnosis.

Statistical Processing

Data were stored in the Microsoft Office Excel program.

Additionally, representative "witness" images of the diagnoses are included for inclusion in the study.

Ethics

The present investigation was carried out in accordance with the five basic ethical principles of all medical research: respect for persons, beneficence, nonmaleficence, justice, and informed consent.

The agreements of the World Medical Association's Declaration of Helsinki[7] on ethical principles for medical research involving human subjects were taken into account, complying with the basic principles of all research. The study patients were explained the objectives of the research and the importance of their participation, guaranteeing the confidentiality of the information, as well as the possibility of withdrawing from the study if they wished.

RESULTS

Five patients were studied: four women and one man (**Table**), with an age range between 36 and 72 years, referred to the reference center due to the impossibility of implanting endocavitary leads for

Table. Distribution of the data studied for each patient.

Patient	Age (years)	Sex	Diagnosis that motivated pacemaker implantation	Diagnosis of venous system malformations	Diagnostic imaging technique
1	36	Female	CAVB	PLSVC + No RSVC	CT
2	41	Female	CAVB	PLSVC + No RSVC	Venous angiography
3	56	Female	CAVB	PLSVC + No RSVC	CT
4	62	Male	CAVB	DSVC	CT
5	72	Female	CAVB	DSVC	Venous angiography

CAVB, complete atrioventricular block; DSVC, dual superior vena cava; CT, computed tomography; RSVC, right superior vena cava; PLSVC, persistent left superior vena cava permanent cardiac pacing.

Four of the patients required a primary pacemaker implant. The remaining patient, the oldest, had been treated at her original center for a pacemaker pocket infection (primary implant) implanted twelve years earlier (in the right region) and required a new implant via the left region. The diagnosis motivating the need for permanent pacing in all five patients was complete atrioventricular block (CAVB). However, the youngest patient also presented with paroxysmal supraventricular tachycardia (PSVT). In the latter patient, both diagnoses (CAVB and PSVT) were confirmed by a twenty-four-hour Holter study and in follow-up consultations.

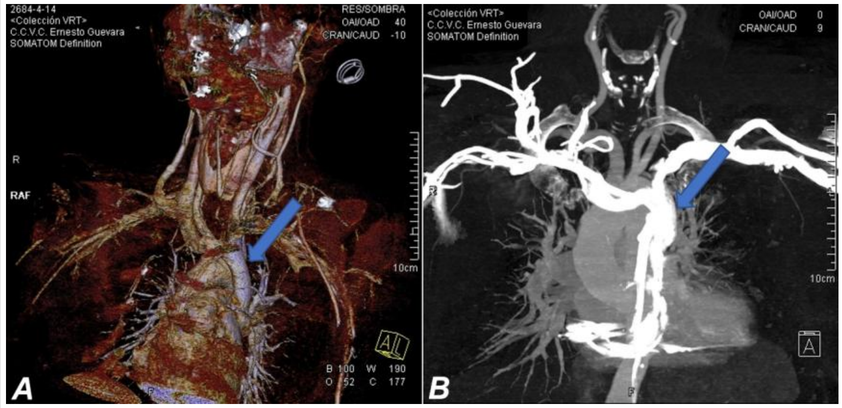

Figure 1. Images of chest angiotomography (VRT Reconstruction [Volume Rendering Technique]) of patients 1 **(A)** and 3 **(B).** The blue arrows show the isolated superior vena cava running to the left of the midline.

All patients remained asymptomatic until their first contact with a physician, at which point the primary pacemaker implant was decided. Imaging diagnoses of the malformations were made using multidetector angio-CT in three of them and venous angiography in the remaining two.

In the 36-year-old patient, who initially underwent a CT scan, a PLSVC was observed, located to the left of the aortic arch, with the absence of an RSVC. The PLSVC drained into the RA in a posterior plane through the coronary sinus, with no large venous vessels defined in their normal position (absence of RSVC). Patient number 3 was also studied by CT, with a final diagnosis of PLSVC plus absence of RSVC (**Figure 1**).

Knowledge of the probability of associated vascular malformation when faced with difficulty in endocavitary lead implantation led us to establish an institutional diagnostic protocol involving the injection of 20-50 milliliters of contrast from the subclavian vein or cephalic vein of the arm homolateral to the chosen implant site. This is based on the premise of avoiding exposure to additional CT radiation beyond the fluoroscopy necessary for device implantation.

During contrast injection from the right subclavian vein, the venous vessel is not delineated in the normal anatomical location of the vena cava in the right projection of the column (absence of RSVC); instead, it crosses the midline and joins the contralateral homonymous vein (left subclavian) to give rise to a large-caliber vessel (PLSVC) with a left paravertebral anteroposterior projection (**Figure 2**). Visualization of this venous angiography, by performing the correct implantation technique, allows access to the endocardium when drainage is to the coronary sinus and from there to the RA.

Therefore, an important aspect is knowing where the PLSVC drains, because if the final drainage is not into the RA, endocavitary implantation will be impossible. The **video** (patient 5, **supplementary material**) shows the path of the contrast from the cephalic vein of the arm, giving rise to a large-caliber vessel (PLSVC) in the left anterior oblique projection

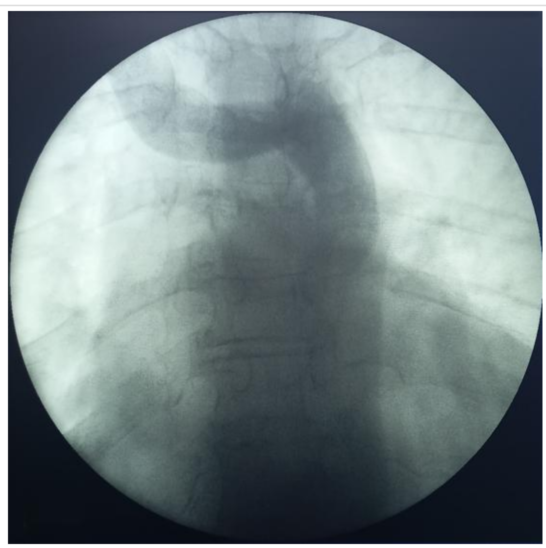

Figure 2. Contrast injection that does not delineate the venous vessel in the normal anatomical location of the superior vena cava in the right projection of the column (absence of right superior vena cava). The contrast crosses the midline and delineates a large-caliber vessel (persistent left superior vena cava) with a left paravertebral anteroposterior projection.

that runs paravertebrally to the left. Note the great dilation of the coronary sinus caused by receiving part of the venous drainage from the upper limbs, head, and neck. The presence of endocavitary leads in the projection occupied by the RSVC can also be observed. This study demonstrates the presence of two superior vena cava.

The specialist performing the endocavitary lead implantation in patients with this type of malformation must have imaging knowledge of the fluoroscopic projections to decide the site of permanent stimulation.

Figure 3A, in a right anterior oblique projection, shows how the permanent stimulation lead in the right ventricular (RV) apex penetrates the inferoposterior portion of the right atrium (RA). Subsequently, it forms a loop towards the anterosuperior portion to the RV inflow tract to end up in the apex region. The lead for RA stimulation makes a reverse loop to end in the posterolateral wall of the RA. **Figure 3B** shows leads running on both sides of the vertebral column, which confirms the diagnosis of dual superior vena cava. On the left side of the column, the leads penetrating through the PLSVC are observed, ending in the following projections: the lead for ventricular stimulation with its tip in a central projection of the image (located in the para-Hisian region) and the lead for RA stimulation is visualized with the tip towards the right lateral portion of that cavity.

DISCUSSION

The low prevalence of superior vena cava system malformations justifies the need and importance of the present study. PLSVC with the absence of RSVC is described as extremely infrequent (0.09-0.13%) in individuals with congenital heart disease[8,9]. However, in 11% of patients with right isomerism, PLSVC and absence of the RSVC are present[10].

In the authors' opinion, the prevalence of this malformation could have a higher incidence than described in the literature, especially in the absence of heart disease, as patients remain asymptomatic for many years[11]. This is confirmed by this series, where patients remained asymptomatic until the diagnosis of complete atrioventricular block (CAVB). Generally, PLSVC, whether as a dual vena cava malformation or with the absence of the RSVC, has been described in 0.3-0.5% of the general population. In the case of patients with congenital heart disease, a higher incidence is described (3-12%), and 0.1-0.3% in unselected patient necropsies[3,12,13].

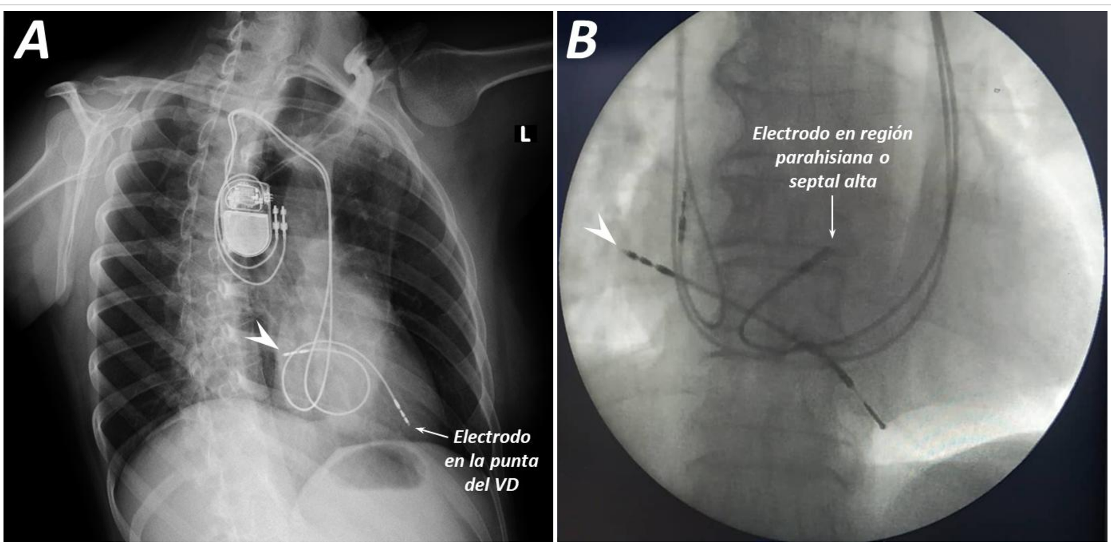

Figura 3.A. Chest X-ray in right anterior oblique projection. The permanent stimulation lead in the RV apex penetrates the inferoposterior portion of the right atrium (RA) and makes a loop towards the anterosuperior portion directed to the RV inflow tract to finally end in the apex region. The lead for RA stimulation (arrowhead), in turn, makes a reverse loop to end in the posterolateral wall of the RA. **B.** Non-contrasted angiography. The definitive leads run along the left side of the column, through the persistent left superior vena cava. The lead for ventricular stimulation was placed in the para-Hisian region and for RA stimulation (arrowhead), in the right lateral wall of that cavity. RV, right ventricle.

The patients in this investigation were studied by echocardiography and showed no other heart malformations, which is a favorable factor for them remaining asymptomatic for a large part of their lives. The center where this research was carried out is a territorial reference center, responsible for the medical care of a population of more than four million patients. If the prevalence of PLSVC with absence of RSVC is 0.09%, this malformation would be expected to be diagnosed in 3600 patients within the population served by this center. This calculation allows us to infer the probability of new cases with this diagnosis appearing in any of the implant centers in this region and country. Therefore, knowledge of the anatomy, diagnostic methods, and techniques for implanting endocavitary leads is of vital importance to achieve procedural success.

We mentioned earlier that one of the patients also presented with PSVT. This arrhythmia has been described in association with these malformations, and ablation has even been performed using non-fluoroscopic navigation systems[14]. However, the presence of CAVB necessitates permanent pacemaker implantation. This patient was treated with a pacemaker and Type IC antiarrhythmic drugs (flecainide), due to the inability to perform ablation as three-dimensional navigation systems were unavailable.

The clinical form of symptom presentation in the studied patients was related to CAVB. In these cases, sinus node dysfunction and heart rhythm originating in the coronary sinus may exist, possibly due to an alteration in development and hypoplasia of the sinus node. Furthermore, the atrioventricular node tissue may be elongated and fragmented, possibly due to the large size of the coronary sinus ostium. These alterations may facilitate the presence of CAVB and/or intranodal reentrant tachycardias. This can lead to clinical situations that frequently present with syncope and necessitate permanent pacemaker implantation[3].

Regarding the implantation of leads in the final position for permanent pacing, there is not much description in the literature on how to carry out the procedure. Different specialists have published techniques to achieve this, as there is currently no universally adopted method[11]. Some propose manually looping the stylet to reach the RV apex; others report having achieved it by preforming an 'L' in the stylet[5,15].

The case series presented is not extensive; however, knowledge of the anatomy and different fluoroscopic projections has allowed us to achieve the placement of leads in the RA by manually preforming the stylet into a 'J' shape, according to the size of the RA. It is also important to know that the entry of the lead into the RA is via its posteroinferior portion; therefore, simply pushing it can displace it to the lateral wall of the atrium. Access to the RV may be more difficult, but the use of different fluoroscopy projections will always help to understand the displacements of the lead by knowing the radiological anatomy.

Reaching the RV for permanent pacing has been achieved as follows: once the RA is accessed, the stylet is withdrawn about 10 cm, so that the lead remains "floating" inside the RA, and in a left anterior oblique view, the lead is rotated counterclockwise while being further introduced. This maneuver allowed penetration into the RV apex (**Figure 3A**).

Our electrophysiology team has also developed techniques for para-Hisian pacing[16], so achieving this pacing site was a challenge with this malformation. After leaving the lead floating in the RA, as explained above, instead of rotating it counterclockwise, we performed a clockwise rotation while introducing the lead. In this way, we reached the RV outflow tract. Once located in that position, an 'L' measuring 2.5 centimeters is made at the tip of the stylet, the stylet is introduced again, and the lead is advanced downwards. As it descends, upon reaching the central area of the fluoroscopic image (para-Hisian or high septal zone) in any of its projections, fixation and threshold checks are performed. This latter pacing site constitutes a more physiological way of conducting the stimulated impulse.

CONCLUSIONS

It is likely that there is an underreporting in the incidence and prevalence of persistent left superior vena cava due to the absence of clinical symptoms. Knowledge of the anatomical variants of this malformation, diagnostic methods, and techniques for implanting endocavitary leads are of vital importance to achieve procedural success. Specialists who face lead implantation, upon identifying that their descent is via the left paravertebral region, should establish diagnostic protocols and master the specific techniques for their implantation.

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