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Case Report

Cardiocentro ERNESTO CHE GUEVARA SANTA CLARA CUBA

Dual-chamber pacemaker implantation through persistent left superior vena cava

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ARTICLE INFORMATION

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The authors declare no competing interests

Acronyms
AV: atrioventricular
CS: coronary sinus
PLSVC: Persistent left superior vena cava
PM: pacemaker
RA: right atrium
RSVC: right superior vena cava
RV: right ventricle

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ABSTRACT
Persistent left superior vena cava is a congenital anomaly that occurs as a result of a degenerative condition in the left anterior cardinal vein. It is found in 3% of healthy subjects and up to 10% of those with congenital heart disease. It is generally an incidental finding, while performing invasive procedures, which may complicate the implantation of cardiac antiarrhythmic devices. The case of a patient with persistent left superior vena cava and absent right superior vena cava is reported. The patient had a history of surgical correction of anomalous pulmonary venous connection, developed sinus node disease and required permanent dual-chamber pacemaker implantation.
Key words: Persistent left superior vena cava, Sinus node disease, Pacemaker

Implantación de marcapaso bicameral a través de vena cava superior izquierda persistente

RESUMEN
La persistencia de la vena cava superior izquierda es una anomalía congénita que se presenta como resultado de un trastorno degenerativo en la vena cardinal anterior izquierda. Se encuentra en el 3 % de los sujetos sanos y hasta el 10 % de los que presentan cardiopatías congénitas. Su hallazgo es generalmente accidental, durante la realización de procedimientos agresivos, que pueden complicar la implantación de dispositivos cardíacos antiarrítmicos. Se presenta el caso de una paciente con vena cava superior izquierda persistente y ausencia de vena cava superior derecha, con antecedentes de corrección quirúrgica de conexión anómala de venas pulmonares, que evolutivamente desarrolló enfermedad del nodo sinusal y requirió implantación definitiva de marcapaso bicameral.
Palabras clave: Vena cava superior izquierda persistente, Enfermedad del nodo sinusal, Marcapaso

INTRODUCTION

Persistent left superior vena cava (PLSVC)[1] is the most common form of anomalies of the systemic venous return. The prevalence in healthy subjects is between 1 and 3%, and up to 10% in patients with congenital heart disease[1,2], and is associated with increased frequency with coarctation of the aorta, ventricular septal defect, mitral atresia, atrial septal defect, anomalous pulmonary venous connection and tetralogy of Fallot[1,2].

Persistent left superior vena cava occurs due to faulty involution of the left anterior cardinal vein. It is rarely unique, while there is regularly a double system of superior venae cavae, which communicate through the innominate vein, and PLSVC opens into the right atrium (RA) through a dilated coronary sinus (CS)[1-3]. Although this anomaly has no clinical relevance, the importance of its knowledge lies in the consequences brought about by its finding during implantation of cardiac antiarrhythmic devices. Technical difficulties in the introduction and stabilization endocardial systems increase complications during implantation of a pacemaker (PM) or cardioverter defibrillator, especially when there is absence of right superior vena cava (RSVC) and implantation of bicameral systems is required.

This is the case of a patient with absent RSVC and PLSVC and with a history of surgical correction of anomalous pulmonary venous connection that evolutionarily developed sinus node disease and required the final implementation of a bicameral PM.

CASE REPORT

23-year-old female patient, with a history of total anomalous pulmonary venous connection and PLSVC. Surgical correction is performed at 8 months of age, and the absence of RSVC and the presence of a PLSVC draining into the RA through the CS is confirmed during the procedure. After 22 years she starts presenting episodes of dyspnea and dizziness on exertion, to which a syncopal episode during the last crisis was associated. She was assessed at the William Soler Pediatric Cardiology Hospital where sinus bradycardia was evident (**Figure 1**). Complementary tests (Holter, exercise test) were performed, which showed a sinus bradycardia with inadequate symptomatic chronotropic response, so that implantation of a permanent endocardial PM is decided, with DDD dual-chamber pacing mode.

Implantation technique

The patient was premedicated with 1g cefazolin as antibiotic prophylaxis. At the left deltopectoral groove level a surgical incision of 4 cm was performed. Opening was made in layers and the cephalic vein was not visualized, so using fluoroscopic vision a double percutaneous puncture of the left subclavian vein was performed by the Seldinger technique, which was cannulated using 9-French venous sheaths. By moving the guide wires to the right atrium it was radiologically found that they fell on the left paravertebral edge before reaching the RA, confirming the path through the PLSVC (**Figure 2**). After removing the dilator and guide wires two active bipolar electrodes, of passive

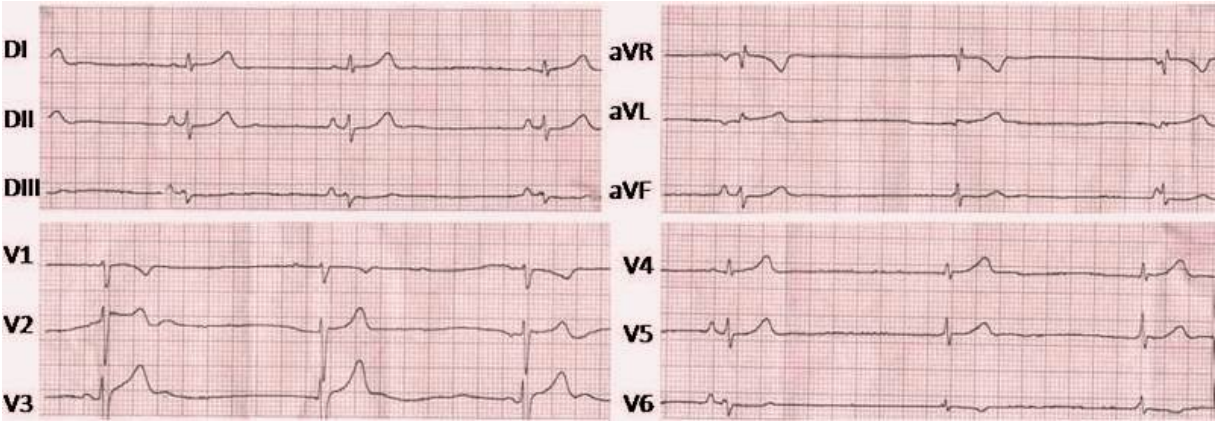

Figure 1. 12 leads electrocardiogram, before implantation of the pacemaker, where 45 bpm sinus bradicardia is seen, PR 120 ms, cQT 350 ms.

(Medtronic Capsure SP Novus 4092), and active (Medtronic CapsureFix Novus 5076) fixation were introduced to right ventricle (RV) and RA, respectively. After preforming the RV electrode guide wire, with a loop in RA, progress was made up to the outflow tract and it was positioned in the RV apex. With the use of guide wire at J, it was unsuccessfully attempted to locate the atrial probe at the level of the appendage, so a straight guide wire was used and the stylet was preformed in L-shaped, so it could be inserted at the level the free wall of the right atrium (**Figure 2**). The following intraoperative measurements were recorded: ventricular stimulation threshold of 0.4 V and atrial of 0.6 V, ventricular probe impedance of 807 ohms and atrial of 974 ohms, while the detected ventricular intracavitary signal was 8.6 mV and atrial of 3.1 mV. After fixation of the leads with nonabsorbable suture, a Medtronic ADAPTA ADDR01 generator was connected, which was positioned subcutaneously. Closure was performed in layers and 80 mg of gentamicin was instilled into the pocket of the PM, as part of antibiotic prophylaxis.

Twenty-four hours after the procedure the patient was discharged after radiologically confirming the correct position of the electrodes, as well as the presence of optimal pacing thresholds.

COMMENTS

Persistent left superior vena cava is a rare congenital anomaly, despite being considered the most frequent thoracic venous malformation[1-5]. The cardinal veins are the main venous drainage system of the embryo, and begin to develop in the eighth week of pregnancy[2]. The anterior cardinal veins drain the cephalic portion of the embryo and the posterior, the caudal region. The right anterior cardinal veins and right primitive must persist to become the RSVC, while the left anterior cardinal is obliterated, caudal to the binding site of the brachiocephalic vein; if the atrophy does not occur then the persistence of the left superior vena cava is produced[1-3]. Atresia or absence of RSVC by involution of the right anterior cardinal vein appears in 10% of subjects with PLSVC, so the right subclavian and jugular veins drain into the left superior vena cava[2-4]; and this is the reason, as in our case, for the implementation of the devices through the PLSVC.

Different morphological variations have been described, in 90% the PLSVC is connected to the CS whereby it drains into the RA; in the remainder percentage, the left superior vena cava can drain directly into the right atrium, into the left superior pulmonary vein or into the left inferior. Another variant is the connection to the roof of the left atrium with absence of septation between this and the CS, which is known as "roofless" coronary sinus[1,2].

As a remnant of the left superior vena cava there remains the oblique vein of Marshall, located in the posterior wall of the left atrium[1,2], which has been identified as generating arrhythmias[6,7]. However, other anatomical substrates could be related to the development of cardiac arrhythmias, especially sinus node dysfunction and atrioventricular (AV) block. The embryological development of the sinus and AV nodes and the Bundle of His, can be influenced by the involution of the cardinal veins, especially the left one[5,7]. In patients with PLSVC and absent RSVC changes in the location and histological organization of the

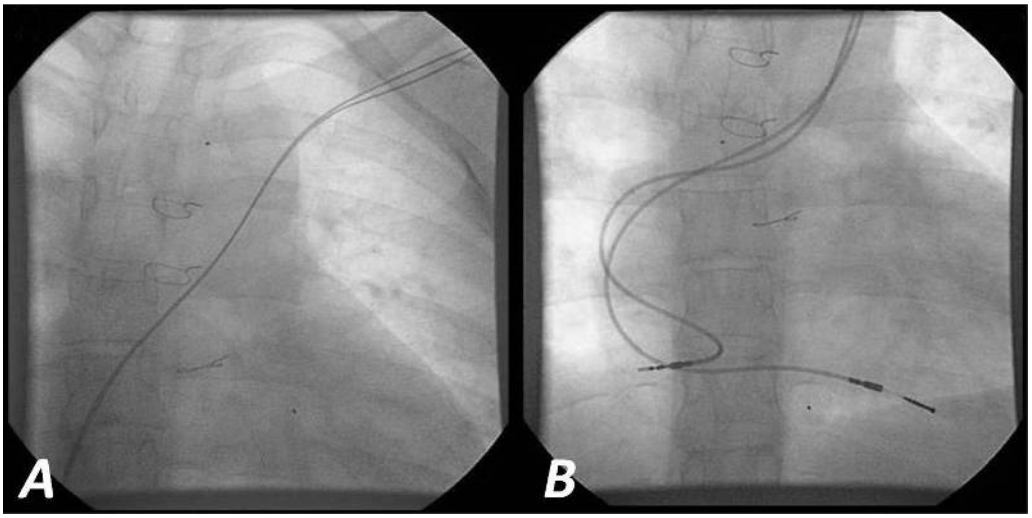

Figure 2. Fluoroscopic images show: **A.** Anomalous pathway of the guide wires, which descend near the left sternal edge. **B.** Final location of electrodes on right atrium free wall (active fixation) and left ventricle apex (passive fixation).

sinus node have been described as well as its hypoplasia, fetal dispersion of the AV node and the Bundle of His in the central fibrous body, small diameter of the bundle of His and arterial hypoperfusion with insufficient irrigation to the conduction system structures[7]. In the case presented, the cause of sinus node dysfunction may be related to the abovementioned changes; however, in patients with surgically corrected anomalous connection of pulmonary veins, the presence of sinus node disease in the long term is reported as a result of injury during surgery, atrial scars that interrupt internodal conduction pathways and the development of late fibrosis of the excito-conductor system[8].

Persistent left superior vena cava may complicate the implant procedures of endocardial pacemakers, that is why aids such as tomography, magnetic resonance imaging and angiography may be useful in the research of this anatomical variant. However, although the routine methods such as echocardiography can diagnose it in 70% of cases[1], or it can be suspected on chest radiography by the presence of a semilunar structure that appears close to the aortic trunk and goes to the middle third of the left clavicle[1,6], it usually constitutes, due to its asymptomatic progression, an incidental finding during placement of the electrodes.

Formerly, PLSVC diagnosis contraindicated the left approach, so that the contralateral approach via RSVC or implanting epicardial electrodes was recommended. Several methods to achieve proper placement of catheters have been published: at the ventricular level a guide wire can be used and preform the end in a "pigtail"[9] or in L shape[10], or, as in our case, making a loop on the wall of the RA that supports the electrode to facilitate the entry to the tricuspid valve[6]. At the atrial level the use of a conventional J-shaped stylet has been reported without technical difficulties in placing the catheter at the appendage level[4,5], or using a straight guide wire and position one of active fixation in the free wall[10]; however, in the patient in question, the stylet was preformed in an L shape and allowed us to place the electrode on the free wall.

At present the characteristics of the endocardial probes and experiences during the implantation described in the various reports have allowed not only bicameral stimulation[4,10], but also the placing of placing cardiac defibrillators[6,11] and resynchronizers[12] and with a low rate of complications related to the approach through the coronary sinus.

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