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CASE REPORT: CLINICAL CASE

Cardiac Resynchronization Therapy in a Chagasic Patient With Persistent Left Superior Vena Cava

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ABSTRACT

This report describes the case of a woman with Chagas' cardiomyopathy with severe left ventricular dysfunction and persistent left superior vena cava who presented with episodes of syncope without prodromes and who was referred for cardiac resynchronization therapy. Despite this venous anomaly, electrodes were safely positioned in suitable locations. (**Level of Difficulty: Intermediate.**) (J Am Coll Cardiol Case Rep 2019;1:387–90) © 2019 The Authors. Published by Elsevier on behalf of the American College of Cardiology Foundation. This is an open access article under the CC BY-NC-ND license (http://creativecommons.org/licenses/by-nc-nd/4.0/).

HISTORY OF PRESENTATION

A 71-year-old woman with Chagas' disease was referred for evaluation because of episodes of syncope with no prodromes. The electrocardiogram (ECG) showed a third-degree atrioventricular block.

MEDICAL HISTORY

Previous medical records revealed a left ventricular ejection fraction of 20% and a bifascicular block on ECG (right bundle block and a left anterior fascicular block). The patient underwent 24-h Holter monitoring, and multiple episodes of paroxysmal third-degree atrioventricular block were found. Because >40% of ventricular pacing was expected and to avoid additional degeneration of left ventricular dysfunction, cardiac resynchronization therapy (CRT) was indicated according to the current guidelines (1,2).

LEARNING OBJECTIVES

- PLSVC is a rare anatomical anomaly that is often incidentally detected by electrophysiologists or other interventionists during procedures.
- Anatomical knowledge of this venous anomaly and subtle maneuvers may be useful in successfully positioning electrodes in the heart for CRT.

MANAGEMENT

The patient was prepared according to standard aseptic protocols, followed by puncture of the left axillary vein. During advancement of the guidewire, an unusual route was observed that by-passed the left lateral border of the cardiac silhouette to the inferior vena cava. Venography was performed using a 7-F vascular sheath, and a dilated venous path (coronary sinus) between the brachycephalic trunk and the right atrium was delineated; persistent left superior vena cava (PLSVC) was diagnosed (**Figure 1**). A right ventricular electrode with active fixation (Biotronik Solia S 60 5,6F, Berlin, Germany) was placed in the

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ABBREVIATIONS AND ACRONYMS

CRT = cardiac resynchronization therapy

PLSVC = persistent left superior vena cava

right lower septum, and a right atrial electrode with active fixation (Biotronik Solia S 53 cm 5,6F) was placed in the right atrial appendage (Figures 2A to 2C). A new venography of the coronary sinus was performed, and lateral and posterior coronary veins were found (Figure 2A). Using a 0.035-mm flexible guidewire, this coronary vein was cannulated and a coronary sinus sheath (Biotronik Selectra MPEP-45) advanced over the wire. A left ventricular pacing electrode (Biotronik Sentus OTW BP 4,8F) was advanced inside the coronary sinus sheath, and the posterior and lateral branches of the coronary vein were reached using a 0.014-mm hydrophilic guidewire. The left ventricle electrode was positioned over the wire (Figures 2A to 2C). Sensing and stimulation threshold tests were performed in all electrodes with acceptable results. Last, all electrodes were fixed to the major pectoralis muscle through the sleeves, and the overlying tissue was sutured by planes.

DIFFERENTIAL DIAGNOSIS

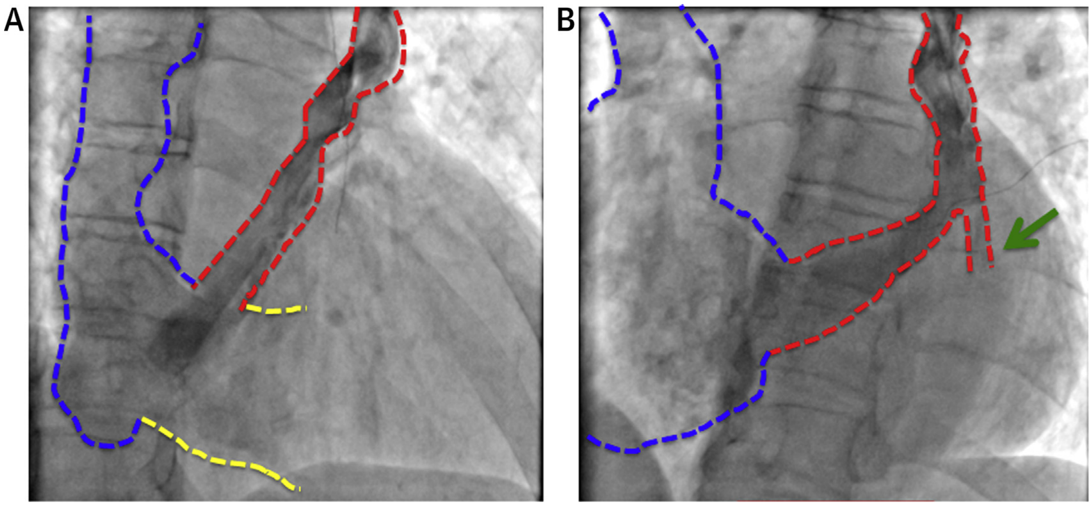
The unusual path of the guidewire along the left lateral border of the cardiac silhouette should raise the possibility of a subclavian artery puncture and placement of the guidewire on the left side of the heart through the aorta. Placement of the guidewire in the left internal thoracic vein, in the left superior intercostal vein, or in the left pericardiophrenic vein should be considered. Extravascular possibilities, such as mediastinum and pleural spaces, should be excluded.

DISCUSSION

PLSVC, although rare, is the most common congenital thoracic venous anomaly, with an estimated prevalence of 0.3% in general population. Briefly, it is the result of patency of the embrionary left anterior cardinal vein, leading drainage into the right atrium through the dilated coronary sinus (3). CRT has been supported by the guidelines for patients with heart failure without left bundle branch block when >40% of right ventricular pacing is expected. This aims to prevent further loss of ventricular function by dyssynchrony caused by right ventricular stimulation (1,2). Because multiple episodes of third-degree atrioventricular block were identified during 24-h Holter monitoring and a high rate of ventricular pacing was expected, CRT was one of the best modalities of cardiac stimulation for this patient.

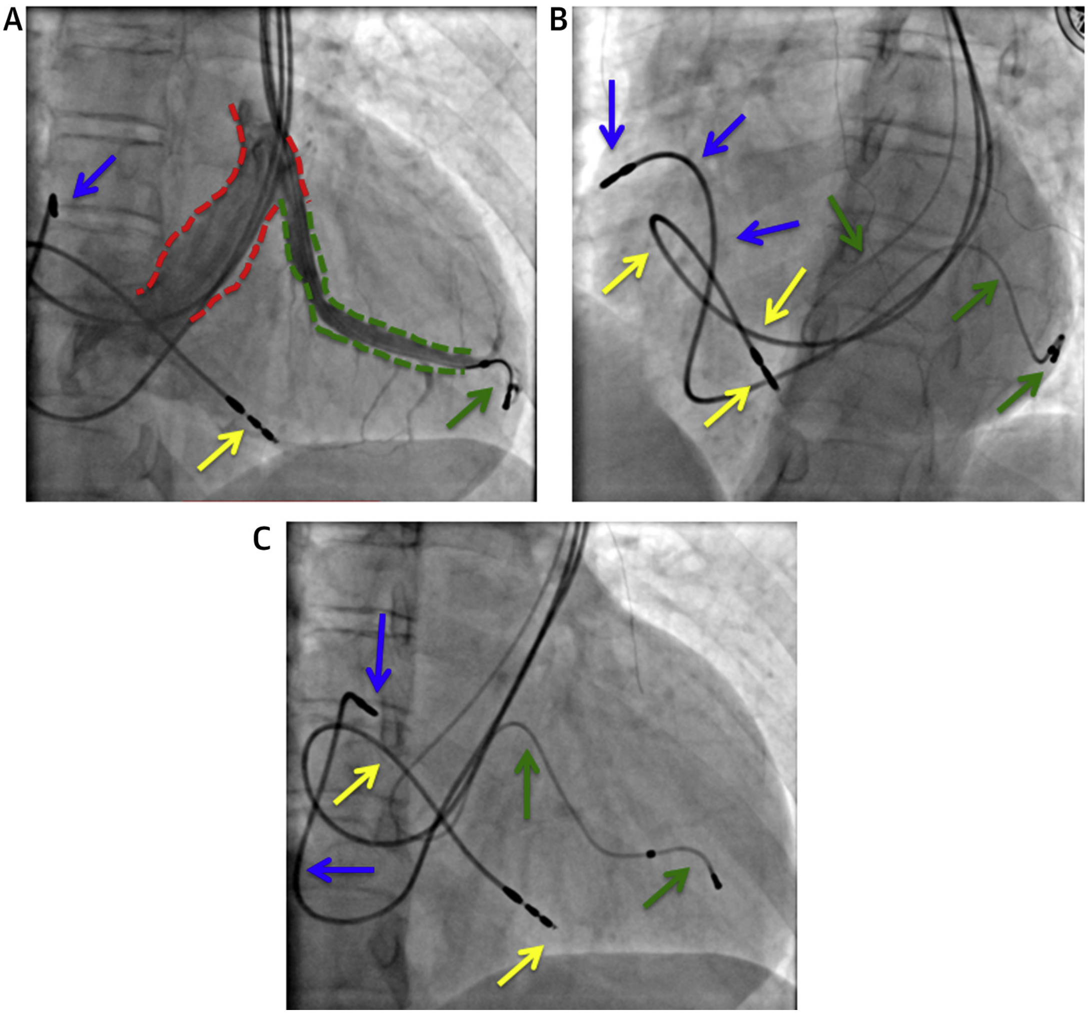
Implantation of cardiac devices in this scenario might be challenging and time consuming due to the need for transpassing sharp angles for positioning the electrodes in the right atrium and ventricle. In addition, delivering an electrode for left ventricular stimulation in a favorable coronary

FIGURE 1 **Anatomical Design of the PLSVC**



(A) Cardiac silhouette shown in right anterior oblique view during contrast infusion in the subclavian vein. The borders of the coronary sinus are delimited by the **red dotted line,** and the **blue dotted line** delimits the boundaries of the right atrium and the superior vena cava. The **yellow dotted line** delimits the initial portion of the right ventricle. **(B)** The previously described structures are shown in left anterior oblique view. The **green arrow** signals the initial portion of a posterolateral coronary vein. PLSVC = persistent left superior vena cava.

FIGURE 2 **Final Positioning of CRT Electrodes**



(A) Anteroposterior view of the coronary sinus **(red dotted lines)** and the posterolateral coronary vein **(green dotted lines).** The right atrial electrode **(blue arrow)** positioned in the anterior portion of the atrium, the right ventricular electrode **(yellow arrow)** in the basal portion of the interventricular septum, and the left ventricular electrode **(green arrow)** in a posterolateral coronary vein. **(B and C)** Left anterior oblique and right anterior oblique views, respectively, showing the right atrial electrode **(blue arrows)**, the right ventricular electrode **(yellow arrows),** and the left ventricular electrode **(green arrows)** in the previously mentioned positions. CRT = cardiac resynchronization therapy.

vein might be an Herculean endeavor even for the most experienced operator. Because most patients with PLSVC are asymptomatic, the operator should be aware of the challenge at the time of the procedure.

CRT implantation in a patient with PLSVC is an unexpected and rare situation faced by the electrophysiologist during the procedure; knowledge of some details may make the procedure easier to perform. First, understanding the anatomy is of vital importance for delivering the correct maneuvers to transpose the sharp angles between the coronary sinus and both the atrium and ventricle with electrodes. In this regard, using different projections in fluoroscopy is useful and time-saving. Second, accessing the right atrium can be challenging and time-consuming. For example, using a "S"-shaped stylet inside the right electrode after positioning it in the basal portion of the right atrium can provide better angulation for screw delivery and good electrode fixation. Third, the right ventricle may be accessed more easily if the lateral portion of the right atrium is used as an anchor to allow the formation of a bend in the electrode to allow its entry in the ventricular chamber. Fourth, because the coronary sinus is a delicate structure, smooth movements should be made to avoid inadvertent injuries, such as perforation and cardiac tamponade. Fifth, because the coronary sinus sheath is a coarse tool, the coronary vein should be, whenever possible, first accessed with a 0.035-mm guidewire. After that, the sheath can be carefully advanced over the 0.035-mm guidewire into the vein. Thereafter, the sheath can be removed and the left ventricle electrode properly positioned using a 0.0144-mm hydrophilic guidewire. Finally, the coronary sinus sheath can be removed as usual with little risk of displacement.

CONCLUSIONS

As long as the operator has a good knowledge of PLSVC anatomy and its differential diagnosis, CRT implantation is feasible and can be performed safely with similar results to procedures performed in patients without a venous anomaly.

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KEY WORDS cardiac resynchronization, Chagas' disease, coronary sinus