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Double superior vena cava and left brachiocephalic vein agenesis: a rare systemic vein anomaly and potential source of CIED and CVC placement complications

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Double superior vena cava and left brachiocephalic vein agenesis: a rare systemic vein anomaly and potential source of CIED and CVC placement complications

R. Steckiewicz et al., DSVC without LBCV detected on CIED

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Abstract

Abnormal systemic vein development produces anomalous veins, which—in the case of persistent left superior vena cava and/or left brachiocephalic vein (BCV)—exhibit considerable topographic and morphometric differences in comparison with their usual anatomy. The nature and extent of those developmental anomalies—detected during intravenous procedures, such as cardiac implantable electronic device (CIED) lead insertion or central venous catheter (CVC) placement—may hinder the procedure itself and/or adversely affect its outcome, both at the stage of cardiac lead advancement through an abnormally shaped vessel and lead positioning within the heart. This may lead to problems in achieving optimal sensing and pacing parameters and in ensuring that the patient cannot feel the pacing impulses. These events accompanied a de novo CIED implantation procedure in the patient with a double SVC and left BCV agenesis, who ultimately required reoperation.

Key words: persistent left superior vena cava, brachiocephalic vein, venous anomalies, venography, transthoracic echocardiogram, CIED, CVC

INTRODUCTION

The increasing use of minimally invasive transvenous procedures — such as cardiac implantable electronic device (CIED) implantation — facilitates detection of interindividual venous variations. Abnormal systemic vein development produces anomalous veins, which—in the case of persistent left superior vena cava (PLSVC) and/or left brachiocephalic vein (BCV)—exhibit considerable topographic and morphometric differences in comparison with the typical vein anatomy [1, 2, 3, 4, 5]. Discovered incidentally during a CIED implantation or central venous catheter (CVC) placement procedure, such anatomical variations may hinder the procedure itself and possibly adversely affect the recovery period.

One rare mediastinal systemic vein anomaly is a developmental variation in the form of a double superior vena cava (SVC) with concomitant agenesis of the left BCV [6, 7, 8, 9]. Azizowa et al. comprehensively discussed left BCV variations and their clinical aspects [5]. In this paper we focused on one of such variations (encountered during a CIED implantation procedure) along with its ramifications.

CASE REPORT

A 73-year-old woman with hypertrophic obstructive cardiomyopathy and tachycardia-bradycardia syndrome (with sinus bradycardia and paroxysmal atrial fibrillation) was admitted to our ward to undergo an elective implantation procedure of a dual chamber pacemaker (DDD mode).

The patient received an atrioventricular cardiac pacing system with both leads introduced via left subclavian vein (SV) puncture. The administration of the contrast agent directly into the SV visualized a congenital mediastinal systemic vein anomaly in the form of a PLSVC, which determined the route of cardiac lead advancement into the heart (Fig. 1. A–C). The recorded intraoperative cardiac pacing parameters, pacing thresholds, and sensing amplitudes from the site of lead tip placement in the right atrium (RA) and right ventricle were acceptable.

A few days after the procedure, the patient developed pacing-associated nonspecific stabbing pain at the right costal margin, sternum, and epigastrium. Pacemaker lead parameters recorded on the fourth postoperative day were found to be unchanged, apart from a slightly increased pacing threshold in the atrial lead. Nonetheless, due to the persistent intense discomfort reported by the patient and the suboptimal intracardiac position of the leads, which was apparently causing this discomfort, the CIED was removed.

One month later (March 2021), the patient was readmitted into the ward to have a CIED implanted on the right side. Venography images obtained with contrast administration into the veins of the right cubital fossa showed a cephalic vein (CV) morphometry conducive to lead insertion and a normal right SVC (Fig. 2 A). CV cut-down technique was used to insert both leads into the venous system, and their tips were placed at different locations within the right heart than previously (Fig. 2 B, D). The pacing parameter and sensing amplitude values recorded at the new lead locations were normal. Test pacing provoked no negative sensations in the patient. This status remained constant both during the procedure and during further in-hospital and outpatient follow-up (July 2021).

Computed tomography (CT) scans illustrated the nature of the encountered developmental anomaly and the vein’s topography in the mediastinum in coronal (Fig. 3A) and transverse (Fig. 3B) planes. This documented a definitive lack of the left BCV (in its expected location, i.e. anterior to the aortic branches; Fig. 3C).

A computed tomography scan ultimately elucidated the nature of the developmental anomaly, and showed: the mediastinal topography and morphometry of the left SVC (Fig. 3A), the spatial relation of the two superior venae cavae with respect to the adjacent arterial vessels (Fig. 3B), unequivocally showed a lack of the left BCV in the area where it might be expected, that is in front of the main arteries branching off the aortic arch (i.e. the left brachiocephalic trunk, left common carotid artery, and left subclavian artery) (Fig. 3B, C).

Contrast echocardiography confirmed left BCV agenesis and the consequent formation of two independent vessels returning blood from the upper part of the body to the RA (with the blood from the right side returning through the right SVC and from the

left side through the left SVC and coronary sinus (CS). The arrow marked with one asterisk (*)points to the coronary sinus (CS), which is filled with a contrast agent. The arrow marked with two asterisks (**)points to the right ventricle (RV), with no contrast agent, while the arrow marked with three asterisks (***)points to the RV, which is filled with contrast. Figure 4 shows evidence of the presence of a PLSVC draining blood from the left arm to the CS and then, through the right atrium, to the RV.

Subsequent outpatient follow-up showed normal atrioventricular pacing, which was not accompanied by any unpleasant sensations induced by right ventricular stimulation, which had been felt by the patient after the previous procedure.

DISCUSSION

Abnormal embryological development of the systemic mediastinal veins may result in one of three anatomical variations of PLSVC, namely two variations of double left SVC that differ by the presence or absence of the bridging left BCV and a form of single left SVC with right superior SVC agenesis. The prevalence of the PLSVC is approximately 0.3–0.5%, with the anomaly typically detected incidentally during postmortem examination; invasive cardiac, anesthesiologic, or cardiothoracic procedures; echocardiography, computed tomography, etc. [9, 10, 11, 12].

During physiological development of human embryos, the initially symmetrical venous system has the form of two pairs of cardinal veins (anterior cardinal veins and posterior cardinal veins, which drain the cranial and caudal regions of the embryo, respectively). These paired veins combine to form the right and left common cardinal veins.

In its mature form, the terminal segment of the right anterior cardinal vein forms the right SVC. By week 20 of gestation, the left vessel undergoes involution, with the blood flow redistributed to the right side. The PLSVC is a result of the lack of this physiological involution of the left anterior cardinal vein. The PLSVC drains venous blood to the CS or directly to the RA, rarely to the left atrium (90% vs. 10%). In the absence of any other congenital vascular anomalies, the fact that the PLSVC drains into the RA

typically produces no symptoms. However, such hemodynamic conditions increase the risk of supraventricular arrhythmias associated with CS volume overload [13, 14, 15].

The PLSVC more commonly accompanies congenital heart defects (3–10%), including ventricular or atrial septal defect, pulmonary stenosis, complete atrioventricular canal defect, dextrocardia, and triatrial heart. The rate of PLSVC detection, like that of other congenital anomalies, depends on the employed diagnostic method and the stage of development (children vs. adults) [16, 17, 18]. In the case presented here, echocardiography had not shown any evidence of a heart defect.

Approximately 85% of adults with a newly detected PLSVC have both functional routes of venous drainage. The failure of a bridging vessel to form between the two anterior cardinal veins leads in the lack of left BCV (approximately 65%). In 10–20% of cases the right cardinal vein fails to develop, which in consequence leads to right SVC agenesis. This variation has marked hemodynamic consequences, since all the venous blood returning from the upper part of the body to the RA flows exclusively through the single left SVC, which drains into the CS.

On week 8 of normal human gestation, the BCVs form as a result of anastomoses between the right and left anterior cardinal veins. The left BCV is typically located above the aortic arch and in front of the arterial branches (brachiocephalic trunk, left common carotid artery, and left subclavian artery). After crossing over to the right side of the upper mediastinum, the left BCV joins the right BCV thus forming the right SVC (Fig. 4C) [19]. Approximately 1% of left BCV anomalies accompany congenital heart defects, such as tetralogy of Fallot, patent foramen ovale, or ventricular septal defect. The prevalence of this congenital venous anomaly in populations without heart defects has been estimated at less than 0.4%. Left BCV anomalies may be isolated, accompany a PLSVC, or be a result of vessel agenesis (as in the case presented here). This type of developmental systemic vein anomalies affects 0.3–0.5% of the general population.

Due to its topographic and morphometric parameters, a PLSVC may hinder both the transvenous advancement of cardiac leads and their positioning in the right heart. This is particularly true during the stage of cardiac lead advancement towards the RV, which requires negotiating the acute angle between the CS outlet and the tricuspid valve. Sometimes, positioning the lead in the shape of the letter L or the Greek letter alpha (α)

may facilitate this maneuver (Fig. 1C). However, this may lead to problems in achieving optimal pacing parameters, ensuring that hart stimulation is not felt by the patient.

The presence of anatomical variations in the veins used during medical procedures, such as CIED implantation or CVC placement, may significantly affect the course of these procedures, just as in the presented case of a congenital mediastinal systemic vein anomaly [1, 2, 3, 19, 20].

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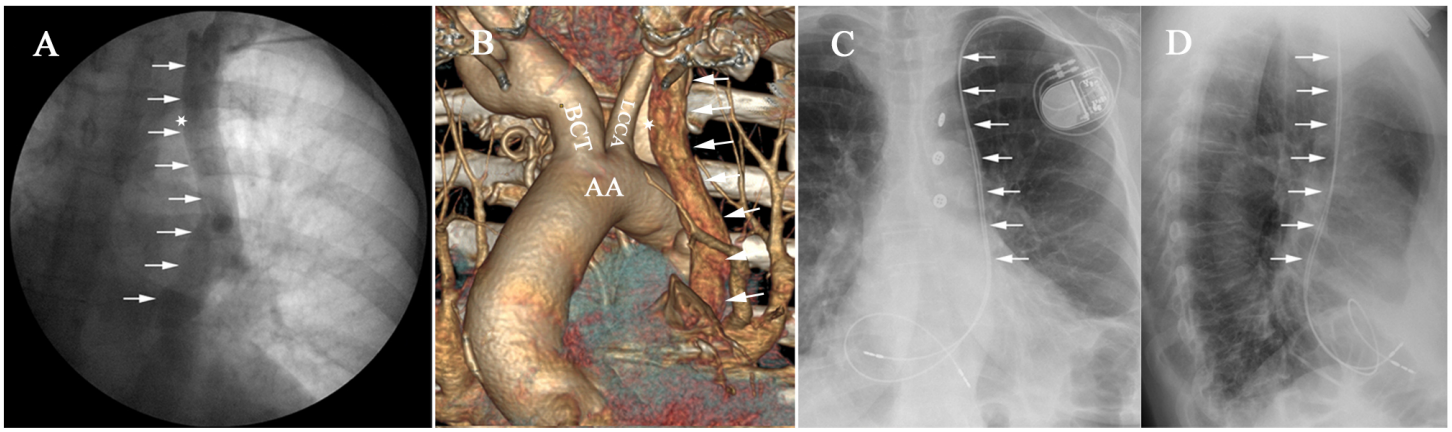
Figure 1. A–D. Cardiac DDD mode pacemaker implantation on the left side in a 73-year-old woman (February 2021). A. Contrast agent administration via left subclavian vein puncture visualized a persistent left superior vena cava (SVC) (arrows) but not the site of its potential junction with the left brachiocephalic vein (asterisk). B. A computed tomography scan showing 3-dimensional topography of the encountered venous anomaly (arrows) with respect to the following arterial vessels: the aortic arch (AA), brachiocephalic trunk (BCT), and left common carotid artery (LCCA). C, D. X-ray film showing the course of both cardiac leads within the left SVC (arrows) and the location of their tips within the right heart chambers (C. PA view, D. Lateral view).

Figure 2. A–D. A CIED (DDD pacemaker) implantation on the right side (March 2021). A. Contrast administration via the peripheral veins of the left forearm showed normal venous return to the right atrium through the right superior vena cava (SVC) (arrows); contrast administration did not visualize any left brachiocephalic vein (BCV) outlets into the right SVC (asterisk). B. The course of cardiac leads within the right SVC (arrows) and the position of lead tips within the RA and right ventricle (follow-up X-ray, PA view). C. A 3-dimensional computed tomography scan illustrating the course of the cardiac leads (arrows) within the right SVC with respect to the aorta (A); BCT – brachiocephalic trunk; LSA – left superior artery. D. The course of the cardiac leads (follow-up X-ray, lateral view)

Figure 3. A–C. Computed tomography scans. A. Coronal scan showing the left superior vena cava (SVC) coursing vertically, anterior to the aortic arch (AA), until it joins the coronary sinus. The site (ring) where a normally developed left brachiocephalic vein

(BCV) would join the left SVC. B. Transverse scan showing the likely course of the absent left BCV (line of arrowheads) between the left SVC and the right SVC, illustrating its relationship to the brachiocephalic trunk (BCT) and left common carotid artery (LCCA). C. Spatial relations of both superior venae cavae to the aortic arch (AA) and its main branches (LSA – left subclavian artery). The site (oval) where the normally developed left and right BCVs would be connected. The right BCV and its continuation, the right SVC, are marked by the cardiac leads within (long arrows).

Figure 4. A–D. Persistent left superior vena cava (PLSVC). Dilated coronary sinus (CS) in a modified four chamber apical view (Panel A) and parasternal long axis view (Panel B). The CS is dilated because the blood from the left arm drains via the PLSVC to the CS and then flows to the right atrium. After intravenous administration of the contrast agent into the left cephalic vein, the dilated CS (*) (Panel C) is filled first, while there is no contrast visible in the right ventricle(RV) (**). A few seconds later (Panel D), the contrast agent is still present in the CS (*), but now the RV is filled with contrast (***) as well (D). This is evidence of a connection between the left cephalic vein and the RV through the CS and the right atrium. CS – coronary sinus. LV – left ventricle. RV – right ventricle.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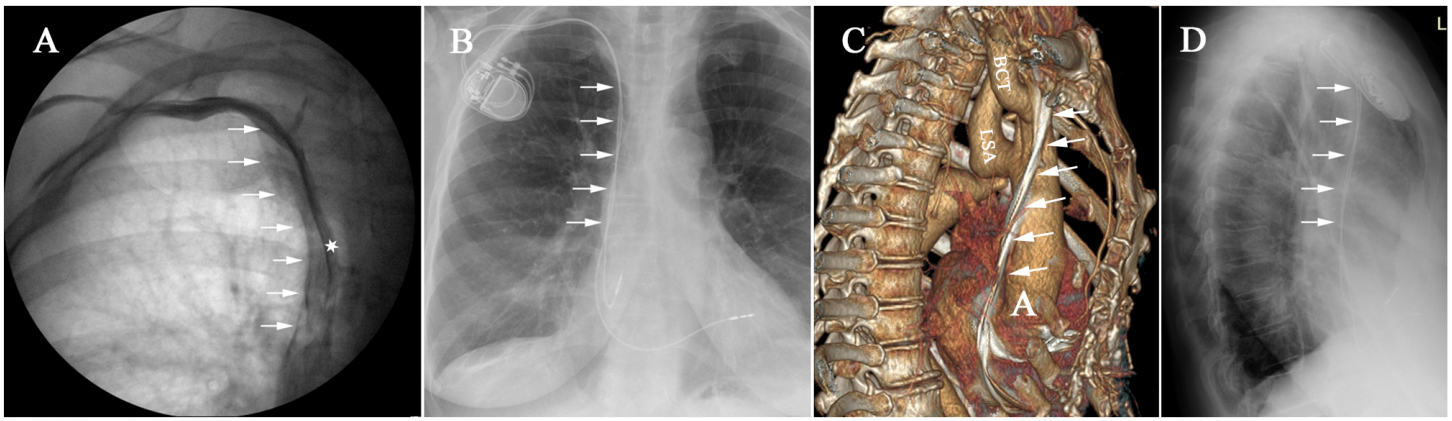
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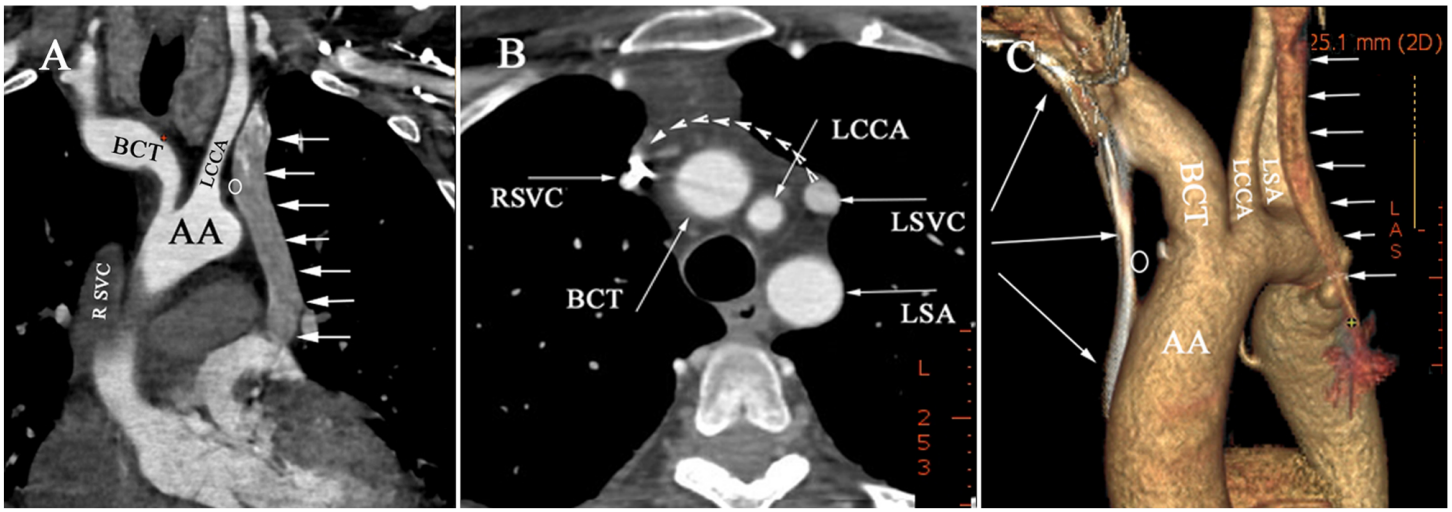
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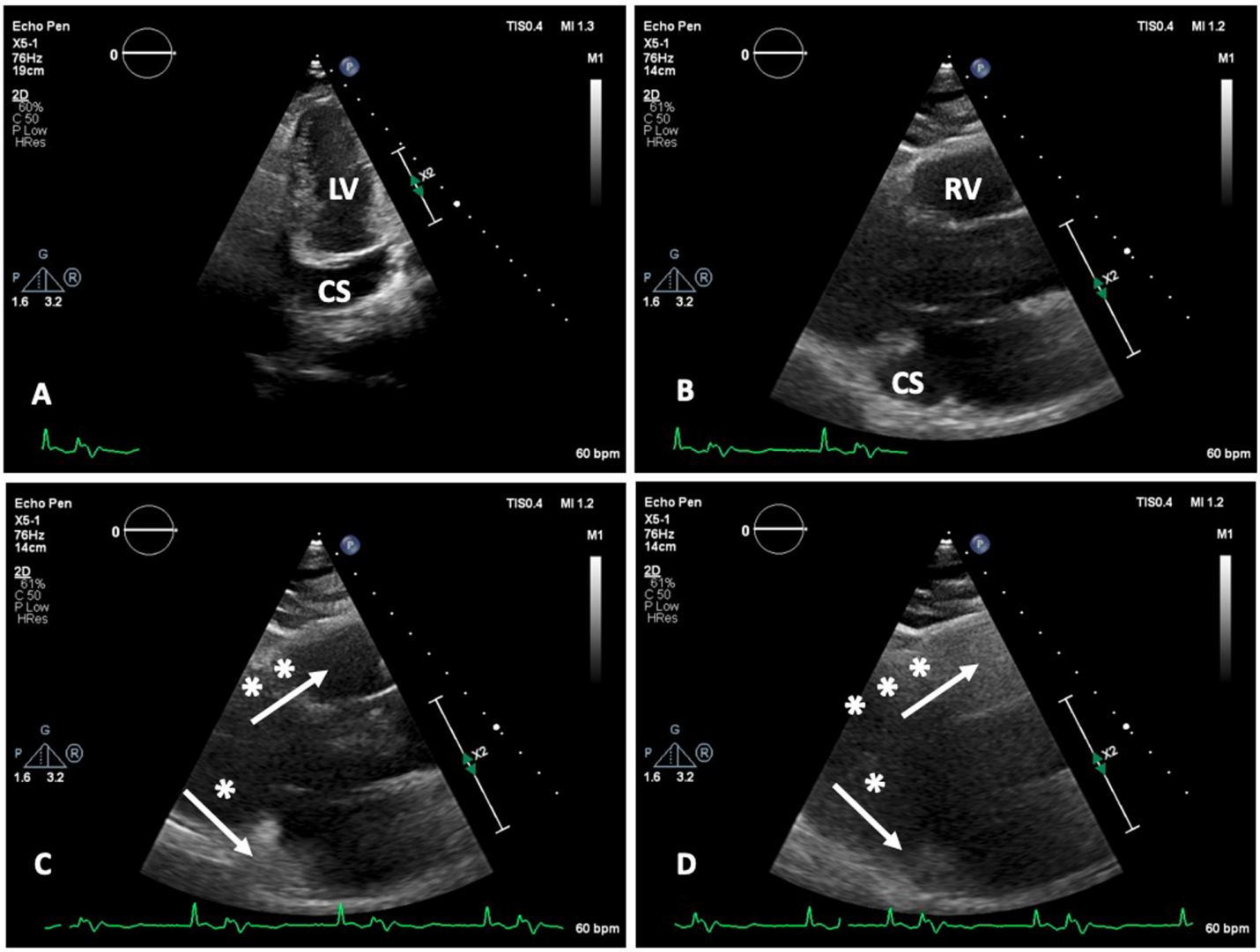
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