







A two-in-one anomaly – persistent left superior vena cava combined with left brachiocephalic vein agenesis – detected during pacemaker implantation

Dwa w jednym, czyli przetrwała lewostronna żyła główna górna połączona z agenezją lewostronnej żyły ramiennie-głowowej, wykryte podczas implantacji stymulatora serca

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Abstract

A great majority of cardiac implantable electronic device (CIED) implantation procedures require the transvenous introduction of leads into the heart. Thus, any developmental anomalies or anatomical variations of these veins may affect the course of the procedure.

This paper presents a case of double superior vena cava in combination with brachiocephalic vein (innominate vein) agenesis. The asymptomatic nature of this systemic venous anomaly made it difficult to detect before the CIED implantation procedure.

Keywords: venous anomalies, persistent left superior vena cava, brachiocephalic vein, venography, transthoracic echocardiogram, cardiac implantable electronic device (CIED)

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Introduction

Transluminal electrode placement continues to be the predominant technique used for pacemaker and cardioverter-defibrillator implantation. Effective transluminal lead advancement requires both vascular patency and favourable morphometric parameters between the site of lead insertion and to final lead position within the heart.

A developmental variation of vessels, which occasionally affects their course, may pose additional challenges during an implantation procedure. This is also true for any developmental anomalies of systemic veins located in the mediastinum, e.g. persistent left superior vena cava (PLSVC). The lack of clinical manifestations that would indicate a need for preoperative diagnostics causes such anomalies to be detected accidentally, for example during cardiovascular procedures [1–3].

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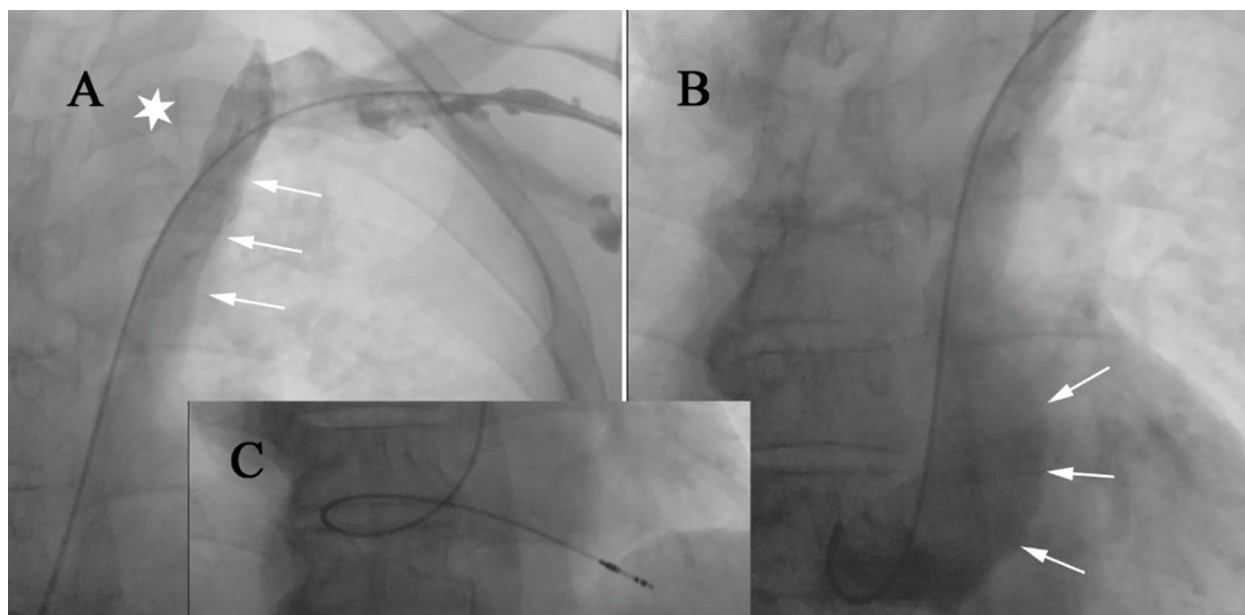


Figure 1. Intraoperative venography: **A.** Persistent left superior vena cava (arrows) in a PA fluoroscopy view. No contrast enhancement of the left brachiocephalic vein (star); **B.** The flow of contract from the left superior vena cava to an enlarged coronary sinus (arrows); **C.** Positioning the lead in the shape of the Greek letter alpha facilitated its introduction into the right ventricle

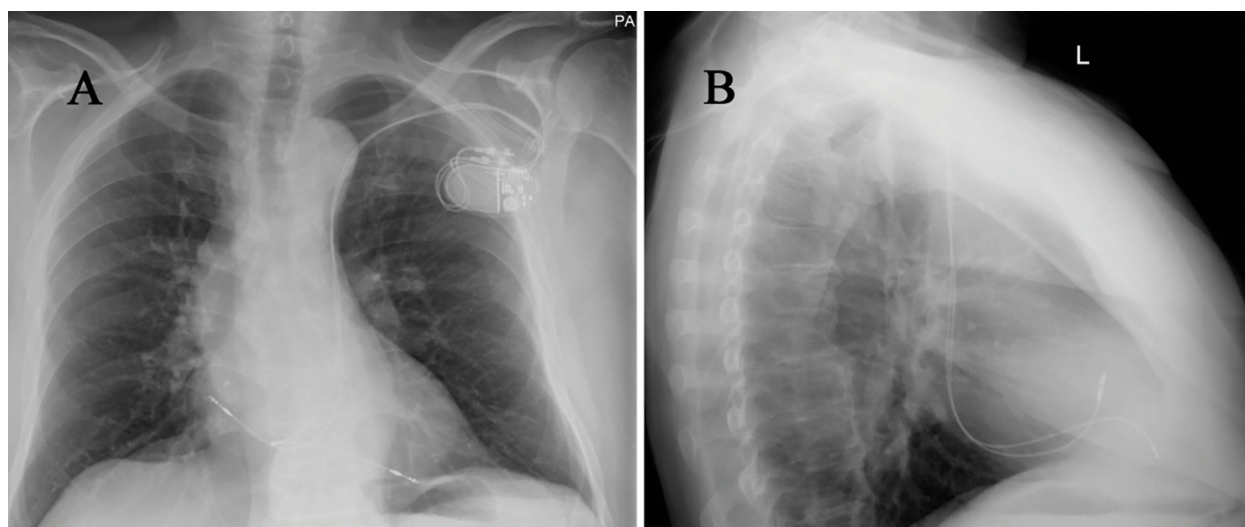


Figure 2. A post-procedure chest X-ray showing the course of both leads and the position of their tips in the chambers of the right heart: **A.** PA view; **B.** Lateral view

This paper presents a rare mediastinal systemic vein anomaly, namely double superior vena cava (DSVC) combined with brachiocephalic vein (BCV) agenesis, detected during cardiac implantable electronic device (CIED) implantation.

Case report

A 73-year-old male was admitted to the ward to undergo CIED implantation due to paroxysmal complete

atrioventricular block with Stokes–Adams attacks. A dual-chamber Assurity MRI DR pacemaker (St. Jude Medical), with a Tendril STS 58 lead placed in the ventricle and Tendril STS 52 lead placed in the atrium, was implanted on March 21, 2023.

The procedure was performed under local anaesthesia, and venous access was achieved via cephalic vein cutdown. Fluoroscopy showed an unusual lead course during ventricular lead advancement, which suggested the existence of PLSVC. This tentative diagnosis was supported once



Figure 3. Transthoracic electrocardiogram. The orifice through which the right superior vena cava opens into the right atrium (arrow)

a contrast agent was administered, clearly showing the lumen of this anomalous vessel (Figure 1A–B)

Discussion

The estimated prevalence of PLSVC is up to 3% of cases in the healthy population, and up to 10% in patients diagnosed with congenital heart defects. There are three types of vascular anomalies involving PLSVC, which differ in terms of morphology and/or anatomy [4–6]. DSVK, with the two vessels bridged with the left BCV. DSVK is present in approximately 85% of adults with PLSVC. DSVK without a bridging left BCV; this anomaly – which constitutes 65% of the remaining PLSVC cases – is a result of no bridging vessel developing between the two anterior cardinal veins during embryonic development. Single superior vena cava (SSVC); this anomaly is a result of right SVC agenesis due to disrupted development of the right cardinal vein (10–20%

of cases). SSVC is haemodynamically significant because the venous blood from the upper part of the body drains into the right atrium solely via the single left superior vena cava.

The presence of a PLSVC may pose a challenge in achieving the optimal ventricular pacing threshold if leads are advanced through this vessel during lead placement in the right heart. Lead advancement towards, and manoeuvring within, the right ventricle requires successfully navigating the acute angle between the opening of the coronary sinus and that of the tricuspid valve. This manoeuvre may be facilitated by using the guidewire to shape the lead into what resembles the Greek letter alpha (α) (Figure 2A).

The images presented in this paper illustrate the presence of the two independent venous drainage routes characteristic of DSVK: PLSVC with no left BCV (shown via intraoperative venography; Figure 1) and the right SVC (shown via transthoracic echocardiography; Figure 3).

Additional information

Ethics statement

No ethical concerns related to the submitted work.

Author contributions

All authors worked together on the final image of the article.

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Conflict of interest

The authors declare no conflict of interest.

Supplementary material

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Streszczenie

Zdecydowaną większość zabiegów stałej elektroterapii serca stanowią procedury wymagające doprowadzenia elektrod do serca drogą przeżylną. Obecność anomalii rozwojowych i odmienność od typowej topografii anatomicznej tych naczyń może wpływać na przebieg zabiegu.

Opracowanie prezentuje przypadek obustronnej żyły głównej górnej w połączeniu z jednoczesną agenezją żyły ramienno-głowej, inaczej bezimiennej (*vena innominata*). Bezobjawowość kliniczna tej systemowej wady rozwojowej naczyń żylnych śródpiersia utrudniła jej wykrycie w okresie poprzedzającym procedurę implantacji wszczepialnego urządzenia elektronicznego.

Słowa kluczowe: anomalie żylnie, przetrwała lewostronna żyła główna górna, żyła ramienno-głowa, wenografia, echokardiografia przezklatkowa, wszczepialne urządzenia elektroniczne (CIED)

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