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CASE REPORT

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Challenging pacemaker implantation: Persistent left superior vena cava with absent right superior vena cava

Schwierige Schrittmacherimplantation bei persistierender linker oberer Hohlvene und Fehlen einer rechten oberen Hohlvene

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► **Abstract** A persistent left superior vena cava (PLSVC) in combination with an absent right superior vena cava (RSVC) is a rare congenital cardiovascular abnormality which is usually found by chance during pacemaker (PM) implantation. In this case we describe a PM implantation using right cephalic approach through PLSVC and coronary sinus (CS), with lead fixation in right atrium and a posterolateral branch of the CS.

► **Keywords** pacemaker implantation · persistent left superior vena cava · absent right superior vena cava

► **Zusammenfassung** Das Vorliegen einer persistierenden linken oberen Hohlvene in Abwesenheit einer rechten oberen Hohlvene stellt eine seltene congenitale kardiovaskuläre Anomalie dar, die meist zufällig im Rahmen einer Schrittmacherimplantation entdeckt wird. In diesem Fallbericht beschreiben wir eine Schrittmacherimplantation mit Zugang über die rechte Vena cephalica, durch die persistierende linke obere Hohlvene und den Koronarsinus mit Sondenfixierung im rechten Vorhof und in einem posterolateralen Ast des Koronarsinus.

► **Schlüsselwörter** Schrittmacherimplantation · persistierende linken oberen Hohlvene · Abwesenheit rechten oberen Hohlvene

A 56-year-old male was admitted to our cardiology department after experiencing a collapse. He had a history of paroxysmal atrial fibrillation, treated with flecainid. During observation a sick sinus syndrome was diagnosed and the patient was scheduled for dual-chamber pacemaker implantation. On echocardiography a dilated coronary sinus (CS) was observed (Fig. 1); consequently a persistent left superior vena cava (PLSVC) was suspected and later confirmed by angiography (Fig. 2A). After accessing the right cephalic vein, the absence of the right superior vena cava (RSVC) and the existence of an innominate vein were discovered (Fig. 2B). Leads were placed through the innominate vein, the PLSVC and CS. Leads were actively fixed in the right atrium (St Jude Med 1788TC/58, St Jude Med, St. Paul, USA) and posterolateral branch of CS (Medtronic Starfix, Medtronic Inc, Minneapolis, USA) (Fig. 3). Normal sensing and capture were obtained. Two days after implantation the patient suffered from frequent diaphragm stimulation. We decided to replace the ventricular lead with an epicardial lead, which was placed using video-assisted thoracal surgery.

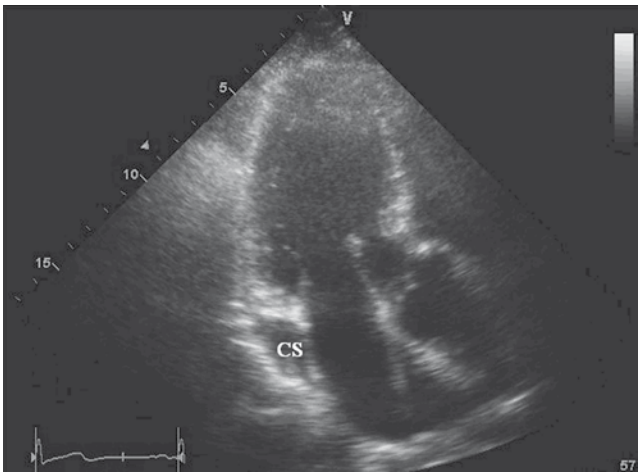

Fig. 1 Two-dimensional echocardiogram. Apical two-chamber view showing dilated coronary sinus (CS)

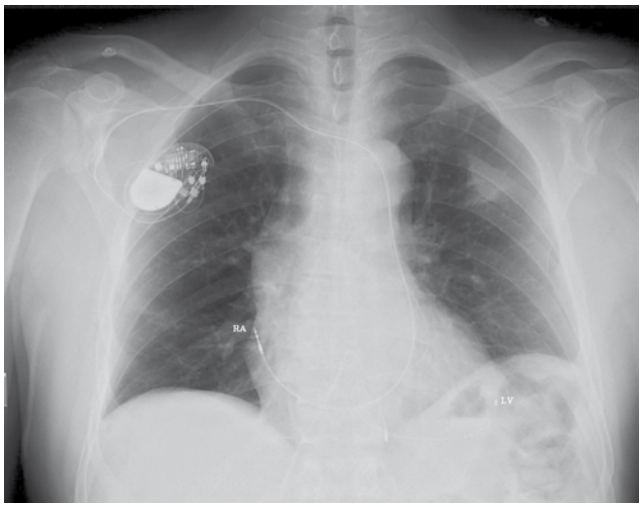

Fig. 3 Chest X-ray. Final position of pacemaker and leads. *RA* right atrial lead, *LV* left ventricular lead

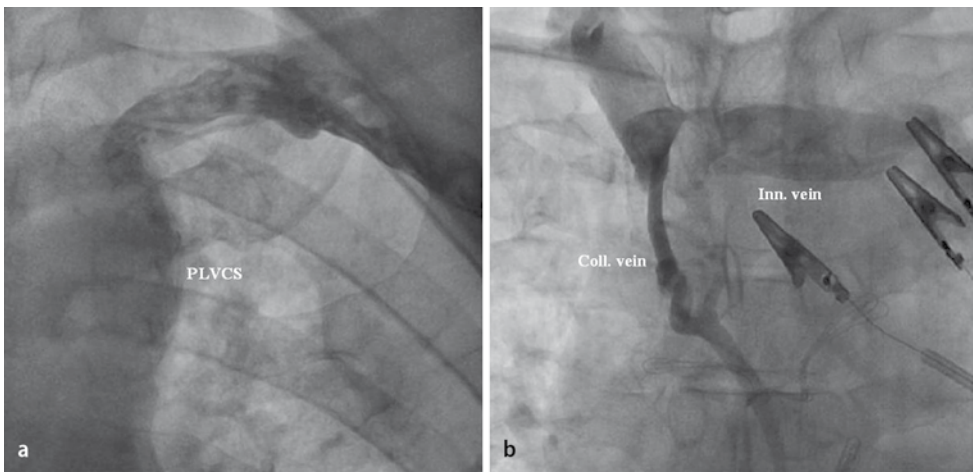

Fig. 2 Venography: **A** persistent left vena cava superior (PLVCS), **B** absent right vena cava superior and presence of innominate vein (Inn. vein) with flow from right to left and a collateral vein (Coll. vein) from right subclavian vein to vena cava inferior

Discussion

PLVCS is the most frequent variation of the thoracic venous system, with a prevalence of 0.3-0.5% in the general population [2,5]. However, an absent RSVC associated with PLSVC is rare, with a prevalence of 0.07-0.13%. In 46% of the cases, absence of RSVC is associated with additional congenital cardiac abnormalities [1]. Embryology of PLVCS with absent RVCS has been published previously [7,9,10].

Because this congenital variation is clinically asymptomatic, several cases were reported as incidental findings, for example, during pacemaker implantations. Due to the abnormal venous anatomy, these implantations were often complicated and technically difficult. Several different techniques of implantation have been described [3-5,8]. For this case, we chose a less common right cephalic approach and fixation of the leads in the right atrium and posterolateral branch of CS. Unfortunately we had to replace the LV lead because of diaphragmatic stimulation, which can be explained by the anatomy of the phrenic nerve and posterolateral branch of CS.

Detection of PLSVC before implantation is possible. PLSVC should be considered when echocardiography shows a dilated CS [4,6,9] or when the opacity of the RVCS is missing on chest X-ray and an additional structure at the aortic knob of the aortic left upper mediastinum is present [6]. Venography, contrast-echocardiography, CT scan or MRI can be used to confirm the diagnosis [6,9].

Conclusion

A PLVCS in combination with absent RVCS is a rare congenital malformation making pacemaker implantation challenging.

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