Persistent left superior vena cava discovered during pacemaker implantation in a paediatric patient

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Introduction

A 12-year-old boy with a history of congenitally corrected transposition of the great arteries (ccTGA) underwent pacemaker (PM) implantation due to advanced conduction disorders (numerous episodes of 1st, 2nd and 3rd degree atrioventricular (AV) block). During the procedure, persistent left superior vena cava (PLSVC) was discovered. Although this finding required supplementary manoeuvring, the ventricular lead was successfully placed into the sub-pulmonary chamber and the procedure was completed without complications.

The patient’s profile overview

The patient was a 12-year-old boy, with congenital heart anomaly (CHA) – ccTGA. In this defect heart ventricles are reversed – the left, systemic ventricle is morphologically the right ventricle, while the right, sub-pulmonary ventricle is morphologically left. The patient developed multiple arrhythmias – episodes of 1st, 2nd and 3rd degree AV block as well as self-limiting supraventricular tachycardia. The patient was qualified for pacemaker implantation during multi-specialist consultation. Pre-implantation diagnostic measures were taken – Holter ECG monitoring confirmed AV blocks, while transthoracic echocardiography (TTE) and magnetic resonance imaging (MRI) ruled out potential obstacles to the procedure.

Procedure description

The implantation procedure took place in the operating theatre, under general anaesthesia, and was performed by a paediatric cardiac surgeon and an electrophysiology cardiologist. An incision in the left subclavian e.g. region was made and the cephalic vein was found. Selective venography was done, and presence of the PLSVC was discovered (Fig. 1 & 2), although such a finding was not described in any of the previous imaging
Due to non-standard anatomy of the heart with ccTGA and existence of PLSVC, multiple attempts to introduce the lead into the sub-pulmonary ventricle were made with pigtail and J-shape stylets. During one such unsuccessful attempt the lead retrogradely positioned itself in the right superior vena cava (Fig. 4). Finally, the lead was positioned in the apical region of the sub-pulmonary ventricle, stimulation parameters were checked, and the lead position was secured (Fig. 5). The lead was connected to the pacemaker. The procedure was finished and periprocedural outcome was uneventful.

**Discussion**

PLSVC is the most common venous anomaly, presenting in 0.3%-0.5% of patients in the general population\(^1\text{-}^3\), but with much higher incidence in patients with CHA, up to 11\(^\%\)\(^4\). PLSVC is usually asymptomatic and has no haemodynamic significance but can create a challenge during PM implantation\(^5\text{-}^6\). Dilated coronary sinus and sharp angle of passing through the tricuspid valve require use of multiple shapes...
Pacemaker implantation in paediatric patient with PLSVC

of stylets and additional modelling of this tool[6]. Although
the mentioned methods apply to adult patients, they can be
successfully implemented in a paediatric procedure.

Our patient was considered complicated in advance due
to ccTGA. This anomaly causes the heart to be rotated and
shifted to the right (Fig. 6), creating unusual angles of access to
the ventricle. Intraprocedural discovery of the PLSVC added
even more complexity to the procedure. It is unclear why the
PLSVC was not noticed during the MRI examination. Later
re-examination of the MRI sequence allowed selection of a
structure that may be the PLSVC (located laterally to the left
atrium) and dilated CS (Fig. 7 & 8). ccTGA in combination
with PLSVC required skilful lead manoeuvring, so that it could
be placed in the sub-pulmonary ventricle. These anomalies
significantly extended the duration of the procedure, which
is typical for procedures on patients with PLSVC[9].

PLSVC is usually diagnosed during invasive procedures,
such as central venous catheterization, PM implantation or
retrograde cardioplegia for cardiac surgery. Fluoroscopic or
X-ray images reveal the catheter or PM lead to descend on
the left side of the spine (Fig. 9), not crossing the spine and
descending in the right superior vena cava field. PLSVC may
be also suspected if a dilated coronary sinus is discovered
in TTE. A contrast injection test during TTE is suggested for
diagnosis of PLSVC[7]. If it is present, contrast injected into
the left antecubital vein will be visible in the CS first and RA later.

**Conclusion**

Combination of congenital heart anomaly and existence
of PLSVC significantly prolonged the duration of the pro-
cedure and required excessive measures to achieve optimal
lead location and pacing parameters. PLSVC may be sus-
pected in patients with dilated CS and suggestive position
of intravenous instruments, such as catheters or PM leads. A
simple contrast echocardiographic test may be beneficial for
diagnosis of PLSVC.
References


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