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STUDIUM PRZYPADKU / *CLINICAL VIGNETTE*

Implantable cardioverter-defibrillator placement via a single persistent left superior vena cava in secondary prevention of sudden cardiac death in a patient with Turner syndrome

Implantacja kardiowertera-defibrylatora przez pojedynczą lewostronną żyłę główną górną w prewencji wtórnej nagłego zgonu sercowego u pacjenta z zespołem Turnera

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A 66-year-old female with Turner syndrome (TS), long QT syndrome, and episodes of haemodynamically unstable ventricular tachycardia was admitted to our Clinic in order receive a permanent cardiac pacing device. Prior to hospitalisation, the patient suffered an episode of sudden cardiac arrest caused by ventricular fibrillation, which was diagnosed and successfully managed by the paramedic team. At the time, the patient did not receive any QT-prolonging drugs and her laboratory tests revealed no electrolyte imbalance. The patient had a history of long-standing hypertension, corrective surgery of coarctation of the aorta (in 1973), and chronic atrial flutter. Computed tomography scans of the chest showed a systemic vein anomaly in the form of a persistent left superior vena cava (PLSVC) in the absence of a right superior vena cava (RSVC) (Fig. 1A). The patient received a Cardia VR device (Medtronic) with a 6935-65 lead. The lead was inserted into the right ventricle using a guidewire moulded into an 'α' shape, with its final position in the right ventricle presented in Figure 1B. The set of inborn genetic abnormalities comprising TS includes coarctation of the aorta in 12.5% of TS patients. Approximately 20% of adult TS patients display a prolonged corrected QT (QTc) interval associated with paroxysmal polymorphic ventricular tachycardia of the torsade de pointes, which increases the risk of sudden cardiac death (SCD). A systemic vein anomaly in the form of a PLSVC affects approximately 0.3–0.5% of the population, more commonly occurring in patients with congenital heart defects (3–10%). The prevalence of a PLSVC in TS patients is estimated at 5%. A single PLSVC (with RSVC agenesis) is a rarely reported variant, affecting only 10–15% of patients with a PLSVC. This anomaly is usually detected during procedures requiring catheter or lead insertion through the RSVC because it poses problems during such procedures. As far as we have been able to ascertain, this is the first case report of implantable cardioverter-defibrillator implantation via a single PLSVC in secondary prevention of SCD in a patient diagnosed with TS.

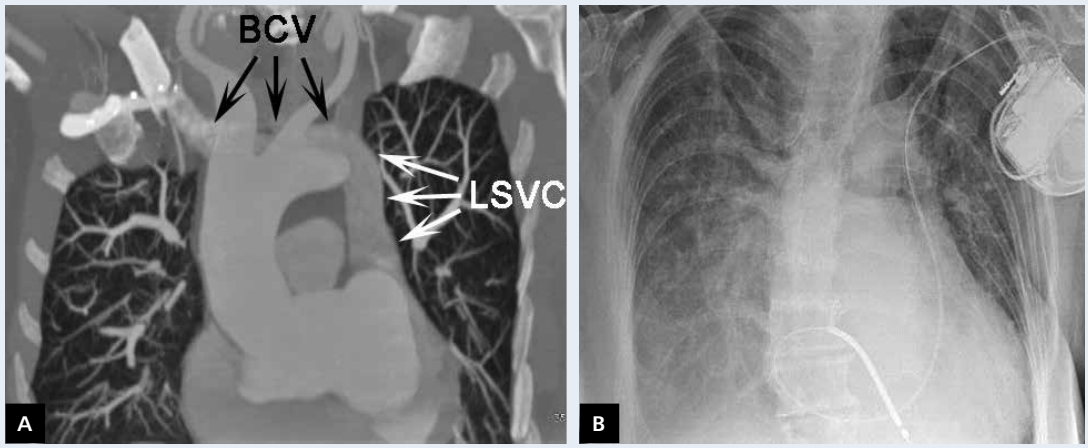

Figure 1. **A**. Spatial relations between the brachiocephalic vein (BCV) and left superior vena cava (LSVC; arrows) visualised via a computed tomography scan (2012); **B**. Chest X-ray: lead position as it courses through the LSVC, coronary sinus, and right atrium, forming the characteristic alpha (α) shape as it enters the right ventricle (AP view)

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