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Robotic bilateral cardiac sympathetic denervation in a patient with severe long QT syndrome: First experience in Poland

Short title: Robotic bilateral CSD

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Cardiac sympathetic denervation (CSD) is an effective therapy in long QT syndrome (LQTS). We present a case of 36 year-old female with LQTS type 2 with recurrent implantable cardioverter-defibrillator (ICD) discharges despite adequate beta-blocker (βB) treatment, in whom for the first time in Poland robotic-assisted bilateral CSD (BCSD) was performed.

The patient was diagnosed at the age of 19 after syncopal episodes and *torsade de pointes* in the postpartum period. Previously, she had been experiencing syncope following arousal and loud noise, and being treated with anti-epileptic drugs. Genetic testing revealed a likely pathogenic variant in KCNH2 gene p.(Gly925Cysfs*49), and p.(Arg176Trp) in KCNH2, considered a risk factor. Due to high-risk arrhythmic profile and persistent QTc >500 ms dual-chamber ICD was implanted in 2007. Despite adjustment in βB and pacing rate she experienced appropriate ICD shocks and electrical storm. In 2013 and 2021, new ICD systems were

replaced due to battery depletion and damage to the defibrillation leads. Finally, in year 2022, due to ineffective nadolol 80 mg/day treatment the patient was referred for BCSD.

After a double lumen intubation, the patient was placed in a lateral decubitus position. The procedure was performed in the same fashion on both sides using three 10-mm thoracoscopic ports in the 6th and 3rd intercostal space (ICS) in the midaxillary line (robotic arms with instruments, DaVinci Xi robot, Intuitive Surgical, Mountainview, CA, US) and 4th ICS in the posterior axillary line (3D camera). The sympathetic chain was identified in T2–T4 portion, dissected, elevated, and excised (including Kuntz nerve). After completion (procedural time 90 minutes), the robot arms were removed and a chest tube was placed in the 1 cm incision.

Post-procedural intermittent ptosis was observed probably due to edema of tissues surrounding right upper stellate ganglion. The patient was discharged on the 3rd day after the BCSD procedure, that is a standard time to discharge. Interestingly, a loss of „fight or flight" response was considered a major positive effect by the patient (she was much calmer in stressful situations e.g. while driving a car or at work). The QTc shortened by 60 ms on a surface ECG to the value below 500 ms. She remains free of symptoms and complications six months after BCSD procedure.

Familial LQTS is a genetic disease (RCDD code: VI-1B-1.2, ORPHA number: 768) predisposing to syncope, polymorphic ventricular tachycardia and sudden cardiac death [1]. Minimally invasive thoracic sympathectomy is recommended as a class I treatment intensification in patients with LQTS experiencing breakthrough cardiac events while on adequate βB therapy [2]. Currently, left or bilateral CSD is an integral part of the management strategy for both LQTS , CPVT and structural heart disease [3–5].

The management of LQTS oscillates between βB and ICD, often adversely affecting the quality of life (QOL). CSD seems to be the therapeutic approach combining efficacy of treatment and QOL, replacing the need of ICD in some cases [2, 3]. Our patient repeatedly experienced serious ICD adverse effects, including ICD discharges, arrhythmic storms and failure of defibrillation leads. Earlier application of CSD would possibly allow to avoid or at least to prolong time to these adverse effects in our patient. We think there is a need for implementation of CSD in prevention of adverse effects and improvement in QOL of LQTS patients.

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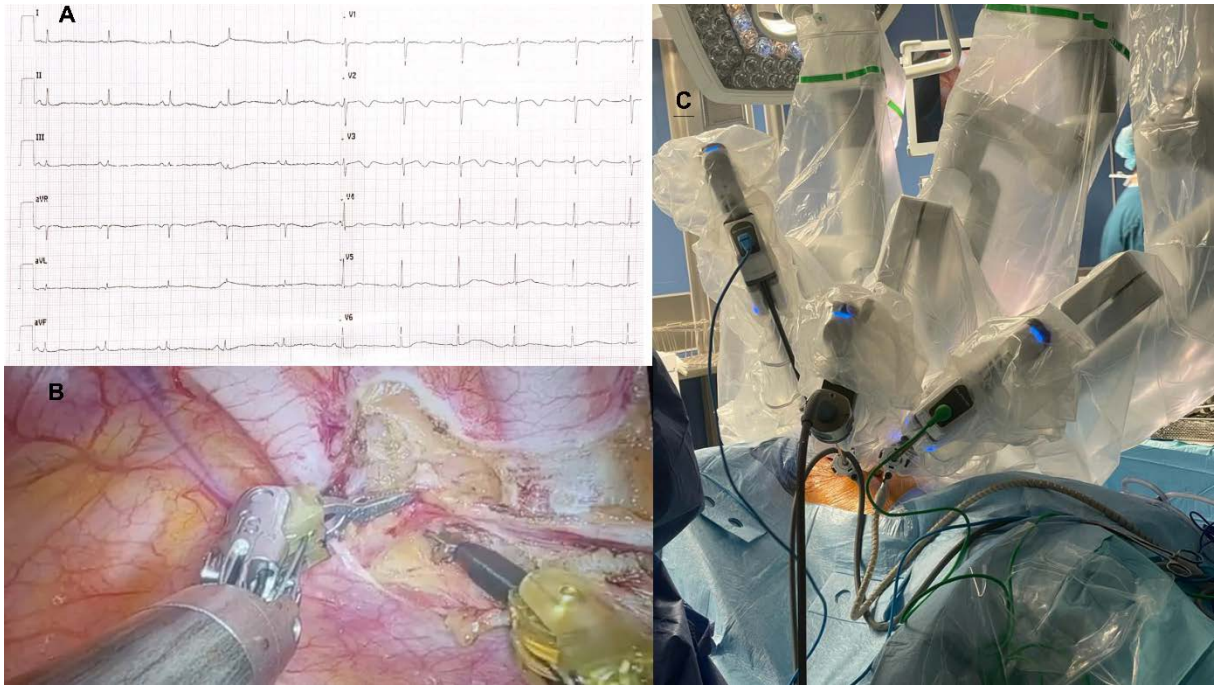

Figure 1. A. Standard, 12-lead electrocardiography, 25 mm/s. Normal sinus rhythm 60 bpm, normal PQ interval, narrow QRS complexes, QTc 540 ms. See abnormal T wave morphology, particularly in precordial leads. **B.** Intrathoracic camera view of T2–T4 sympathetic chain dissection. **C.** Access and robotic arms configuration during sympathectomy