

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

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Cardiac sympathetic denervation (CSD) is an effective therapy in long QT syndrome (LQTS). We present a case of a 36-year-old female with LQTS type 2 with recurrent implantable cardioverter-defibrillator (ICD) discharges despite adequate beta-blocker (β B) 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 syncope episodes and *torsade de pointes* in the postpartum period. Previously, she had been experiencing syncope following sensory stimulation and loud noise and was treated with anti-epileptic drugs. Genetic testing revealed a likely pathogenic variant in the *KCNH2* gene p.(Gly925Cysfs*49), and p.(Arg176Trp) in *KCNH2*, considered a risk factor. Due to a high-risk arrhythmic profile and persistent QTc >500 ms, dual-chamber ICD was implanted in 2007. Despite adjustment in β B 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 the year 2022, due to ineffective nadolol 80 mg/day treatment, the patient was referred for BCSD.

After 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 the nerve of Kuntz). 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 the right upper stellate ganglion. The patient was discharged on the 3rd day after the BCSD procedure, which is a standard time to discharge. Interestingly, the elimination 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 was shortened by 60 ms on a surface electrocardiogram to a value below 500 ms. She remained free of symptoms and complications six months after the 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 β B 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 β B and ICD, often adversely affecting the quality of life (QOL). CSD seems to be the therapeutic approach combining the efficacy of treatment and QOL improvement, replacing the need for ICD in some cases [2, 3]. Our pa-

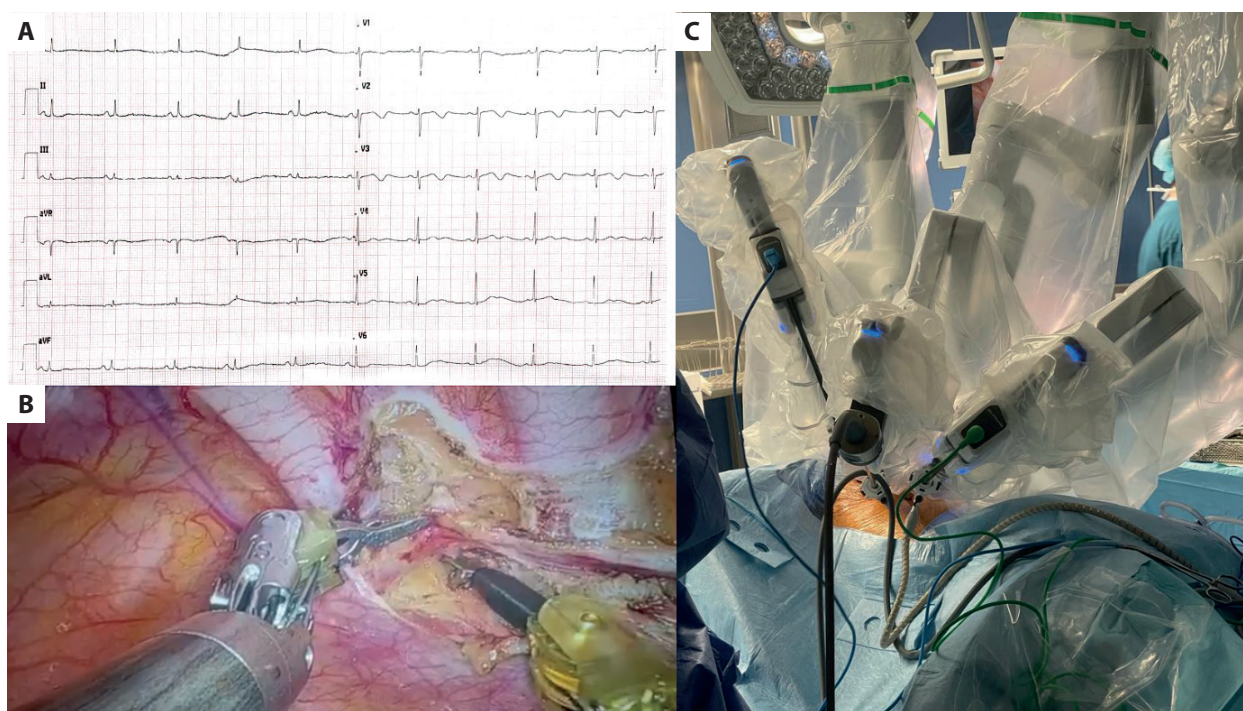


Figure 1. **A.** Standard, 12-lead electrocardiography, 25 mm/s. Normal sinus rhythm 60 bpm, normal PQ interval, narrow QRS complexes, QTc 540 ms. See abnormal T wave morphology, particularly in the precordial leads. **B.** Intrathoracic camera view of T2–T4 sympathetic chain dissection. **C.** Access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

tient repeatedly experienced serious ICD adverse effects, including ICD discharges, arrhythmic storms, and failure of defibrillation leads. Earlier use of CSD would possibly have allowed her to avoid or at least reduce the number of those adverse effects. We think there is a need for CSD implementation in the prevention of adverse effects and improvement in QOL of LQTS patients.

Article information

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