Two years later, the patient experienced ICD shocks due to VF triggered by emotional stress. She was offered bilateral cardiac sympathetic denervation (CSD), which had not yet been performed in Poland in adults for arrhythmic indications. The patient was referred to a regional thoracic surgery center experienced in video-assisted thoracoscopic CSD in hyperhidrosis and Raynaud disease. Although left CSD was considered, recent studies and persistent coldness in the upper extremities supported the use of a bilateral approach.

Bilateral CSD was performed within 60 minutes with a single-port access for each site from the third intercostal space in the medial axillary line. Sympathetic ganglia and fibers were removed from the level of Th1 to Th5, with the upper level of right-sided dissection marked with a titanium clip used for bleeding prevention (FIGURE 1B). The results were confirmed histologically (FIGURE 1C). Interestingly, the patient reported an immediate change in temperature perception, allowing her to stop wearing gloves at temperatures below 10°C despite continued β-blocker therapy. There were no ICD interventions during the 6-month follow-up.

Our case demonstrates several pitfalls of CPVT management. First, the CPVT diagnosis was delayed owing to incomplete workup during the initial hospitalization. The exercise test was only performed at follow-up to confirm antiarrhythmic efficacy of metoprolol. Second, whereas in the largest pediatric CPVT registry, flecainide and CSD were used in 24% and 8% of patients, respectively, nadolol and flecainide were the only medications used in our patient.
FIGURE 1  A – bidirectional or polymorphic ventricular tachycardia runs;  B – chest X-ray: the arrow points to a titanium clip used for bleeding prevention, marking the upper level of right-sided dissection of sympathetic ganglia.  C – histological staining for lipofuscin in sympathetic ganglia.

are not widely available in Poland. Moreover, the largest Polish registry of patients with long QT syndrome reported no cases of CSD. The limited access to subcutaneous ICDs, genetic testing, flecainide, and nadolol affects the quality of care in patients with CPVT and other inherited arrhythmogenic diseases. Although rare prescription drugs can be imported (so called target import), administrative barriers limit their availability in clinical practice.

To the best of our knowledge, this is the first case of bilateral CSD in an adult with CPVT in Poland. Despite diagnostic and therapeutic challenges, the patient was diagnosed and managed as part of a multicenter interdisciplinary heart team including specialists in electrophysiology, inherited arrhythmias, and cardiothoracic surgery.

ARTICLE INFORMATION

CONFLICT OF INTEREST  The authors declare no conflicts of interest. SS and PS coauthored patents on electrophysiology catheters. SS is a stockholder of Medicina S.A.

OPEN ACCESS  This is an Open Access article distributed under the terms of the Creative Commons Attribution-NonCommercial-NoDerivatives 4.0 International License (CC BY-NC-ND 4.0), allowing third parties to download articles and share them with others, provided the original work is properly cited, not changed in any way, distributed under the same license, and used for non-commercial purposes only. For commercial use, please contact the journal office at kardiologiapolska@ptkardio.pl.

REFERENCES