# Bilateral cardiac sympathetic denervation in catecholaminergic polymorphic ventricular tachycardia

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Catecholaminergic polymorphic ventricular tachycardia (CPVT) is a rare inherited disease linked to syncope, bidirectional or polymorphic ventricular tachycardia (bidVT/pVT), ventricular fibrillation (VF), and sudden cardiac death during exercise or emotional stress in patients without organic heart disease.[1] There are several diagnostic and therapeutic options, although their availability in different countries is limited.

We present a case of a 30-year-old white woman after aborted cardiac arrest, which occurred at her brother's funeral. During hospitalization, bidVT/pVT and VF reoccurred, requiring 5 defibrillations. Serial electrocardiograms revealed no features of short QT, long QT, or Brugada syndrome, whereas telemetry and Holter recordings documented bidVT/pVT runs (FIGURE 1A). Echocardiography and coronary angiography were normal.

A single-chamber ProMRI® implantable cardioverter defibrillator (ICD; Iforia 5 VR-T, Biotronik, Berlin, Germany) was implanted, and the patient was discharged on metoprolol (50 mg twice daily) with recommendations for genetic and family screening. Until now, only her 6-year-old daughter was screened (no abnormalities were detected), as no other family members agreed to be screened. Genetic testing is still underway due to lack of reimbursement. Although a follow-up exercise test revealed no significant arrhythmia, the patient still complained of cold sensation in the upper extremities, which she linked to metoprolol use.

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.[2] 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.[1] Secondly, whereas in the largest pediatric CPVT registry, flecainide and CSD were used in 24% and 8% of patients, respectively,[3] nadolol and flecainide

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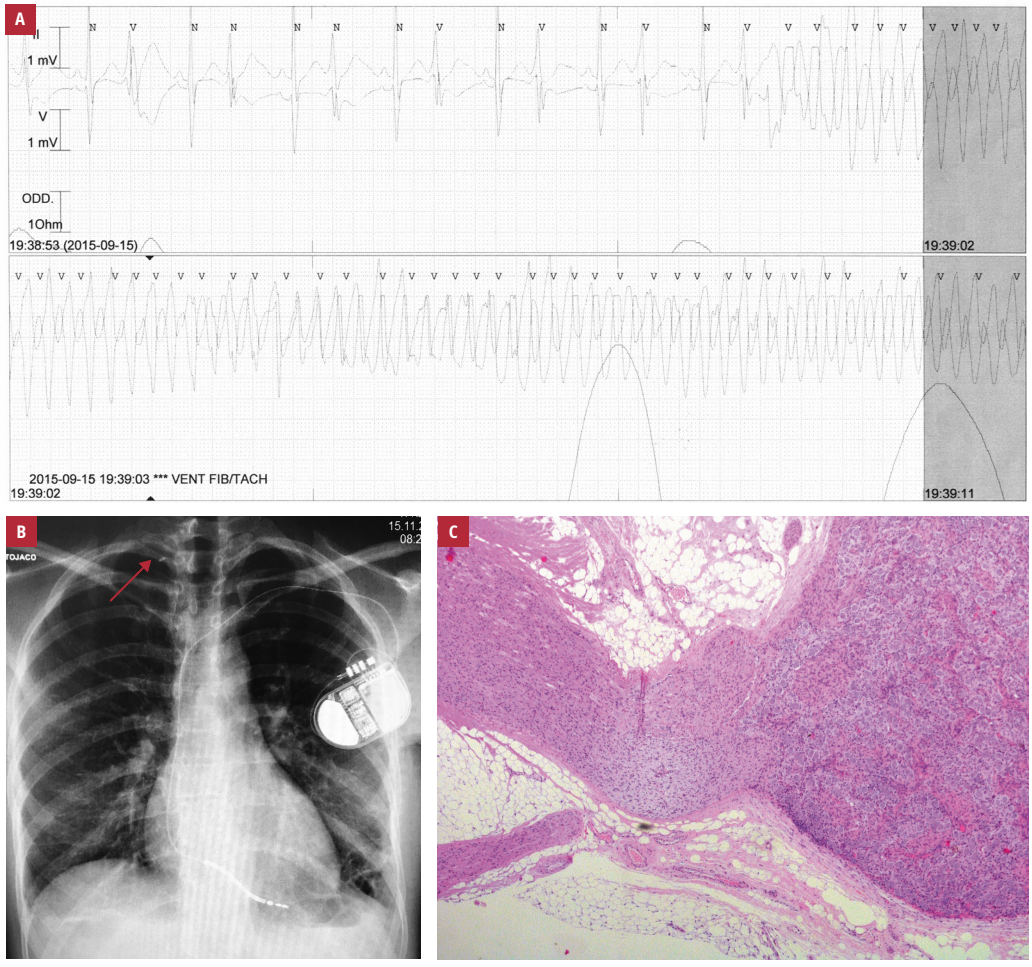


**FIGURE 1** **A** – bidirectional or polymorphic ventricular tachycardia runs; **B** – chest X-ray: the arrow points to a titanium clip used for bleeding prevention, marking the upper level of right-sided dissection of sympathetic ganglia. **C** – histological staining for lipofuscin in sympathetic ganglia

are not widely available in Poland. Moreover, the largest Polish registry of patients with long QT syndrome reported no cases of CSD.[4] 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 Medinice S.A.



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## REFERENCES

1 Priori SG, Blomström-Lundqvist C, Mazzanti A, et al. ESC Guidelines for the management of patients with ventricular arrhythmias. Eur Heart J. 2015; 36: 2793-2867.

2 Shivkumar K, Ajijola OA, Anand I, et al. Clinical neurocardiology defining the value of neuroscience-based cardiovascular therapeutics. J Physiol. 2016; 594: 3911-3954.

3 Roston TM, Vinocur JM, Maginot KR, et al. Catecholaminergic polymorphic ventricular tachycardia in children. Circ Arrhythm Electrophysiol. 2015; 8: 633-642.

4 Zienciuk-Krajka A, Sterliński M, Filipecki A, et al. Implantable cardioverter-defibrillators in patients with long QT syndrome. Kardiol Pol. 2018; 76: 1687-1696.