

Bilateral Cardiac Sympathetic Denervation in a Patient with Electrical Storm Ventricular Arrhythmia and Non-Ischemic Dilated Cardiomyopathy

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INTRODUCTION

Cardiac sympathetic denervation (CSD), also referred to as the left thoracic sympathectomy, is a new treatment option for the management of recurrent ventricular arrhythmia (VA) in patients with VT storm and ICD shocks, in whom ablation and medications have not produced expected results[1]. While effects of CSD in patients with long QT syndrome and catecholaminergic polymorphic ventricular tachycardia have been well described, data regarding its use in other cardiac conditions is scarce [1,2,3].

Keywords: Cardiac sympathetic denervation; Ventricular arrhythmia; VT storm; ICD shocks; Long QT syndrome

We present the case of a 38-year-old physically active male with a family history (father) of myotonic dystrophy type 2 without concomitant cardiomyopathy. The initial presentation included a sudden loss of consciousness with broad QRS complex tachycardia (VT), recurring during hospitalization (Figure 1A).



Figure 1 A: 12-lead standard electrocardiogram (speed 25 mm/s). Wide QRS tachycardia diagnosed as ventricular tachycardia of epicardial origin.

Upon the suspicion of non-ischemic dilated cardiomyopathy (NIDCM, EF 40%) associated with familial myotonic dystrophy, the EP study and catheter ablation were performed urgently. The endocardial ablation was ineffective, due to the epicardial substrate situated within the lower portion of the lateral wall of the left ventricle, and it actually induced a clinically significant VT. Coronary angiography excluded significant anomalies and coronary artery disease. Due to recurrent VT and electrical storm, the ICD device (Protecta DR) was implanted without prior cardiac magnetic resonance imaging.

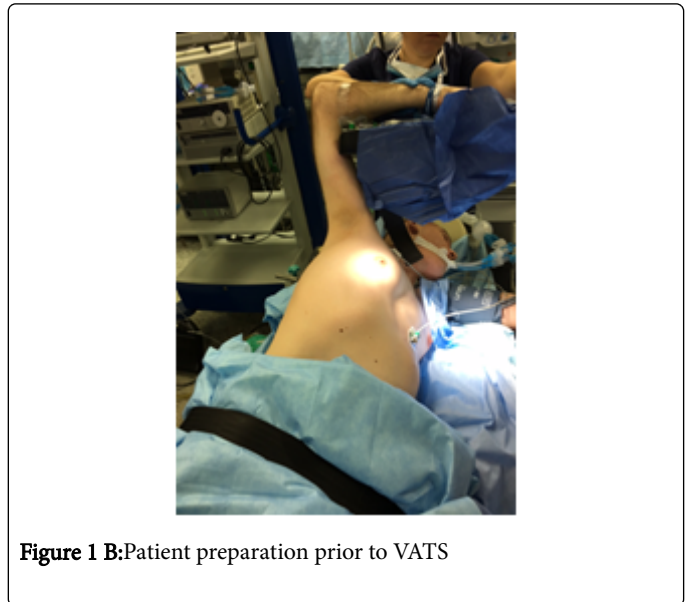


Figure 1 B: Patient preparation prior to VATS

Despite pharmacological treatment, there were further episodes of electrical storm encompassing rapid VT and ventricular fibrillation (VF), which required additional external shock and resuscitation on the top of 6 ICD discharges. The emergency epicardial ablation from the subxiphoid window was performed, leading to the resolution of the most frequent epicardial VT from the lower-right wall and the absence of inducible ventricular arrhythmia in electrophysiologic study. Throughout the inpatient stay, however, the patient had recurrent episodes of NSVT without electrical storm, which depended on

emotional arousal. He was found eligible for the first in Poland video-assisted thoracoscopic (VAT) bilateral cardiac sympathectomy (BCSD) performed by the EP-HEART-TEAM (Figure 1 B, C, D).



Figure 1 C: Video assisted thoracoscopic sympathectomy (VATS) (access from 3rd intercostal space)



Figure 1 D: Resected right-sided lower part of stellate sympathetic ganglion from Th1 to Th4 (6cm)

BCSD was performed under general anesthesia from the bilateral intercostal access to remove the stellate ganglion between Th1-Th4. This procedure was performed for the first time in a cardiac patient in a EP-HEART-TEAM setting in the Centre with expertise in thoracic surgery for hyperhidrosis and Raynaud's syndrome.

The 18-month follow-up, without Amiodarone treatment, was uneventful. The patient returned to long-distance walking. Furthermore, genetic tests confirmed familial myotonic dystrophy type 2 (RCDD code: III-5A.2g). Our case shows that life-threatening ventricular arrhythmias may be the first clinical manifestation of muscular dystrophy type 2. The presentation, physical examination, family history, creatine kinase (CPK) and genetic tests may help to diagnose the disease [4].

Previous studies show that supplementing ICD implantation with BSCD in patients with structural HD and resistant VT reduced the number of ICD shocks and ES episodes. After bilateral sympathectomy, the number of ICD shocks decreased significantly in the 12-month follow-up compared to left-sided sympathectomy alone [5].

The case documented complex antiarrhythmic interventions such as endo-epicardial mapping and catheter ablations, failed drug and ICD therapy and VATS BCSD performed by the multidisciplinary EP-Heart-Team.

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