Ten-Year Follow-Up of Cardiac Sympathectomy in a Young Woman with Catecholaminergic Polymorphic Ventricular Tachycardia and an Implantable Cardioverter Defibrillator

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Sympathectomy for CPVT. Current recommendations for therapy of catecholaminergic ventricular tachycardia (CPVT) include beta blockade and implantable cardioverter defibrillators (ICDs). Patients may experience recurrent arrhythmias, ICD shocks and, rarely, sudden death despite optimal medical therapy. We report a young woman with CPVT who received frequent ICD shocks despite beta blockade, who subsequently underwent cardiac sympathectomy with a dramatic reduction in shocks over 10 years of follow-up. (J Cardiovasc Electrophysiol, Vol. 20, pp. 1167-1169, October 2009)

Introduction
Catecholaminergic polymorphic ventricular tachycardia (CPVT) is a heritable arrhythmogenic disease characterized by adrenergically mediated bidirectional or polymorphic ventricular tachycardia, a normal resting electrocardiogram, and a structurally normal heart.1,2 Typically, physical exertion or psychological stress provokes syncope or sudden cardiac death in susceptible individuals, often children or young adults. Current recommendations for therapy include beta blockade and implantable cardioverter defibrillators (ICD).3 We report a young woman with CPVT who received frequent ICD shocks despite beta blockade therapy who subsequently underwent cardiac sympathectomy.

Case
A 14-year-old girl presented after a syncopal episode while running in a field. She had previously had an episode of syncope with “near drowning.” On both occasions, she recovered consciousness within a few minutes without sequelae. There were no preceding palpitations or chest pain. A great uncle died suddenly at the age of 22 years. Her mother experienced syncope after “near drowning.” On both occasions, she recovered consciousness. She had previously had an episode of syncope while playing basketball, skating, or running (Fig. 2). These shocks did not abate despite ICD reprogramming to minimize inappropriate detection (VF detection zone decreased to ≤273 ms and confirmation extended from 2.5 to 5 seconds) and beta-blockers titrated to maximum tolerable doses (Atenolol 25 mg daily). The patient was initially programmed to a single therapy zone to detect and treat ventricular fibrillation (cycle length <300 ms). Despite compliance with medication (Atenolol 25 mg daily), she experienced recurrent appropriate ICD shocks while playing basketball, skateboarding, or running (Fig. 2). These shocks did not abate despite ICD reprogramming to minimize inappropriate detection (VF detection zone decreased to ≤273 ms and confirmation extended from 2.5 to 5 seconds) and beta-blockers titrated to maximum tolerable doses (Atenolol 50 mg bid). The patient received her first shock after cardiac sympathectomy 18 months later. This occurred as a result of nonadherence to beta-blockade therapy that was discontinuing because of fatigue. She renewed her participation in sports. Nine years later she had an uneventful pregnancy and delivery, and has remained well for a year, 10 years after cardiac sympathectomy.

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sympathectomy. Her genetically affected 6-month-old son has a normal ECG and Holter monitor.

Discussion

Catecholaminergic polymorphic ventricular tachycardia can be highly lethal, with a risk of sudden cardiac death of 30–50% by age 20–30 years in the absence of treatment. Gene mutations in the cardiac ryanodine receptor (RyR2) and calsequestrin protein, both of which are important in calcium trafficking from the sarcoplasmic reticulum into the cytosol, lead to intracellular calcium overload. In the face of sudden sympathetic stimulation, this is hypothesized to lead to delayed afterdepolarizations, triggered activity and consequent arrhythmogenesis. Cardiac sympathetic denervation is an intuitively attractive strategy in these patients as it reduces norepinephrine release in the heart and consequent arrhythmogenesis. Cardiac sympathetic denervation is an intuitively attractive strategy in these patients as it reduces norepinephrine release in the heart and consequent arrhythmogenesis.

The clinical utility of cardiac sympathectomy in patients with CPVT has recently been reported. Wilde et al. recently described 3 pediatric cases of CPVT with recurrent exertional syncope and documented polymorphic ventricular tachycardia that responded to sympathectomy. This report describes a young woman with CPVT and recurrent ICD discharges despite concomitant medical therapy. ICD implantation with the use of beta-blockers is currently recommended in patients with CPVT if they have had a cardiac arrest, syncope, or documented sustained ventricular tachycardia. The use of ICDs in this young population is, however, potentially problematic and does not necessarily protect from sudden death.

ICD implantation does not decrease adrenergically driven arrhythmic events, especially in patients who may not have ideal compliance with their beta-blockers, as was the case with our patient. Exercise may at times provoke both appropriate and inappropriate ICD shocks. This may lead to a significant impairment in the quality of life, as was the case with our patient. Inappropriate shocks may also cause endogenous release of catecholamines, which in turn can precipitate a potentially fatal arrhythmic storm. This is in contrast to cardiac sympathectomy or an antiarrhythmic strategy that has the potential to ameliorate the arrhythmic burden.

Our patient experienced an excellent outcome after cardiac sympathetic denervation with rare ICD discharges in the ensuing 10 years in the context of less than fastidious beta-blocker compliance. In keeping with the recommendations of Wilde et al., the invasive nature of the procedure suggests that it should be reserved for patients that are either intolerant of beta-blockers or symptomatic despite beta blockade. Given the rarity and gravity of the condition, a randomized trial may never be feasible. Nonetheless, cardiac sympathectomy is worthy of consideration in the individual CPVT patient with frequent ICD therapies in spite of appropriate medical management, and arguably earlier in the therapeutic cascade.

References