Sympathectomy for Patients With Catecholaminergic Polymorphic Ventricular Tachycardia
Should We Have the Nerve?

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Since the early descriptions almost 4 decades ago, there has been considerable expansion in the knowledge base for catecholaminergic polymorphic ventricular tachycardia (CPVT), with identification of underlying genetic mutations and a better understanding of mechanisms leading to ventricular arrhythmias. However, as a malignant entity predisposing mostly young, apparently healthy individuals to sudden cardiac arrest, CPVT continues to pose a management challenge to the clinical cardiologist, often compounded by emotionally fraught situations. Risk stratification for CPVT remains problematic, particularly because long-term follow-up data in adequate numbers of patients are hard to obtain in this rare entity, yet clinical experience suggests fairly high event rates in diagnosed subjects. The therapeutic approach to CPVT relies mainly on countering sympathetic stimulation as the key trigger of arrhythmia in this syndrome. β-Blockers have conventionally been the cornerstone of management with sizable reductions in arrhythmia burden; unfortunately, breakthrough events despite β-blocker therapy are not uncommon. Side effects such as lethargy related to β-blockade also result in noncompliance in this young population. Calcium channel blockers such as verapamil have limited efficacy and the Class 1c agent flecainide has shown some promise, although long-term data are lacking. At the present time, implantable cardioverter-defibrillators (ICDs) are advocated for those with sustained VT/syncope or aborted cardiac arrest despite β-blockers. However, concerns have been raised about the possibility of VT storm and death resulting from the sympathetic surge after ICD shocks and the relatively high rate of inappropriate shocks and device complications in these young patients. Left cardiac sympathetic denervation (LCSD) has emerged as an alternative approach to sympathetic blockade and has been used effectively in other inherited arrhythmia syndromes such as the long-QT syndrome. Although scattered reports of the success of LCSD in abolishing arrhythmia in CPVT exist, recommendation for its adoption as standard therapy has been hampered by lack of data on long-term efficacy.

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on flecainide. Flecainide, a drug that potentially directly targets the underlying molecular defect in ryadonide receptor 2,20 has generated considerable interest recently, with early reports of significant reductions in arrhythmia burden in CPVT over that conferred by β-blockers alone.11,21 A multicenter, randomized, controlled trial of flecainide in CPVT currently under way (http://www.clinicaltrials.gov; identifier, NCT01117454) should provide more definitive answers on the efficacy of flecainide. The present study cannot answer the question of whether LCSD is superior to flecainide. If the early promise of this drug holds up in larger trials, it would be important for clinicians to factor this into the management scheme as a desirable noninvasive option compared with LCSD. Surgical expertise in performing LCSD is currently available only in selected centers, representing a possible bottleneck to its widespread adoption. In addition, whether LCSD can be “substituted” for an ICD when symptoms are refractory to medical therapy (as the authors suggest) may need further investigation.

De Ferrari and colleagues are to be congratulated for their careful work, which represents an important advance in the therapeutic options available for the difficult CPVT patient. For rare syndromes such as CPVT, randomized trials can be difficult to conduct, and multicenter, long-term, observational studies are a key resource in advancing the field. This useful cooperative study also shows the way forward for clinicians to continue evaluating management options in CPVT. Considering the results of this study, we agree with the authors that it is reasonable to proceed with LCSD for the CPVT patient with recurrent ICD shocks, despite maximally tolerated β-blocker or flecainide therapy. It is not unreasonable to consider LCSD in the CPVT patient with optimal medical therapy and recurrent syncope if a particular clinical situation demands it, but some residual risk will need to be factored in (9% major events in this study). Future studies should focus on whether LCSD can be universally moved up the management ladder in front of an ICD and whether it affords any long-term benefit in the presently asymptomatic, genotype-positive patient.

Disclosures
None.

References


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