Left cardiac sympathetic denervation (LCSD) is a safe and effective therapeutic option for patients with malignant ventricular arrhythmias stemming from both channelopathies and cardiomyopathies. Although β-blocker pharmacotherapy is the mainstay of medical management for these patients, some are either intolerant or refractory to this therapy. For some patients, a more aggressive sudden cardiac death prevention strategy, such as implantable cardioverter-defibrillators (ICDs), may be required. An ICD will arguably provide the most definitive protection against lethal ventricular arrhythmias; however, there are significant comorbidities, including device-related malfunction, inappropriate therapies, infections, and psychological stress, associated with their use.

Background—Treatment options for patients with recurrent ventricular arrhythmias refractory to pharmacotherapy and ablation are minimal. Although left cardiac sympathetic denervation (LCSD) is well established in long-QT syndrome, its role in non–long-QT syndrome arrhythmogenic channelopathies and cardiomyopathies is less clear. Here, we report our single-center experience in performing LCSD in this setting.

Methods and Results—In this institutional review board–approved study, we retrospectively reviewed the electronic medical records of all patients (N=91) who had videoscopic LCSD at our institution from 2005 to 2011. Data were analyzed for the subset (n=27) who were denervated for an underlying diagnosis other than autosomal dominant or sporadic long-QT syndrome. The spectrum of arrhythmogenic disease included catecholaminergic polymorphic ventricular tachycardia (n=13), Jervell and Lange-Nielsen syndrome (n=5), idiopathic ventricular fibrillation (n=4), left ventricular noncompaction (n=2), hypertrophic cardiomyopathy (n=1), ischemic cardiomyopathy (n=1), and arrhythmogenic right ventricular cardiomyopathy (n=1). Five patients had LCSD because of high-risk assessment and β-blocker intolerance, none of whom had a sentinel breakthrough cardiac event at early follow-up. Among the remaining 22 previously symptomatic patients who had LCSD as secondary prevention, all had an attenuation in cardiac events, with 18 having no breakthrough cardiac events so far and 4 having experienced ≥1 post-LCSD breakthrough cardiac event.

Conclusions—LCSD may represent a substrate-independent antifibrillatory treatment option for patients with life-threatening ventricular arrhythmia syndromes other than long-QT syndrome. The early follow-up seems promising, with a marked reduction in the frequency of cardiac events post-denervation. (Circ Arrhythm Electrophysiol. 2012;5:782-788.)

Key Words: catecholaminergic polymorphic ventricular tachycardia sudden cardiac arrest ventricular fibrillation left cardiac sympathetic denervation denervation

Left cardiac sympathetic denervation (LCSD) is a safe and effective therapeutic option for patients with malignant ventricular arrhythmias stemming from both channelopathies and cardiomyopathies. Although β-blocker pharmacotherapy is the mainstay of medical management for these patients, some are either intolerant or refractory to this therapy. For some patients, a more aggressive sudden cardiac death prevention strategy, such as implantable cardioverter-defibrillators (ICDs), may be required. An ICD will arguably provide the most definitive protection against lethal ventricular arrhythmias; however, there are significant comorbidities, including device-related malfunction, inappropriate therapies, infections, and psychological stress, associated with their use.

With the therapeutic gap and side effects of pharmacotherapy and the comorbidity profile of ICDs, additional therapies may be necessary for those patients with severely arrhythmogenic disease. With the growing understanding of the antifibrillatory potential of LCSD, it has emerged as an additional and safe treatment option for patients with long-QT syndrome (LQTS) and catecholaminergic polymorphic ventricular tachycardia (CPVT). This procedure removes the lower half of the left stellate ganglion (T1) and thoracic ganglia T2 to T4, thereby diminishing the noradrenergic input to the left ventricular myocardium. In a study of 147 patients with LQTS, there was a >90% reduction in cardiac events post-denervation. LCSD raises the threshold for ventricular fibrillation (VF) without impairing myocardial contractility or reducing heart rate and reduces arrhythmias in postmyocardial infarction animal.
models. Although LCSD has a well-established event-attenuating effect in patients with LQTS, its efficacy for non-LQTS arhythmogenic channelopathies and cardiomyopathies is less clear. Herein, we report our single-center experience in performing LCSD in these patients.

**Methods**

In this study, approved by Mayo Clinic’s Institutional Review Board for protection of human subjects, we identified all patients (N=27) with arhythmogenic channelopathies and cardiomyopathies, other than autosomal dominant or sporadic LQTS, who had undergone LCSD at our institution between November 1, 2005, and December 1, 2011, as previously described. A detailed retrospective review of the electronic medical record was performed, specifically looking at demographic variables, genotype, indications for surgery, available follow-up, postoperative complications, presence of ICD or pharmacological therapy, and incidence of aborted cardiac arrest (ACA) or appropriate VF-terminating ICD therapies before and after LCSD. Denervation surgery was considered secondary prevention if the patient had a history of either ACA or VF-terminating ICD therapy. Breakthrough cardiac events (BCEs) were defined as an appropriate VF-terminating ICD therapy, ACA, or syncope occurring after LCSD. Follow-up was censored as of January 1, 2012.

**Procedure**

Using a videoscopic transthoracic approach and single right bronchus intubation, the left-sided sympathetic ganglia were identified through the pleura, which were then divided to expose the sympathetic chain from T4 to T1. The ganglia were removed by dividing the major rami communicans and fully identifying the much smaller sympathetic nerve branches that ravel toward the heart. The lower half of the left stellate ganglion (T1) was then divided along the anatomic fusion between its upper and lower poles. The dissected materials were removed en bloc and sent to pathology for confirmation. A postoperative chest radiograph was performed to confirm the absence of significant pneumothorax, and all patients were monitored postoperatively in the intensive care unit.

**Statistical Analysis**

Statistical analysis was performed using JMP 8.0 statistical software (JMP 8.0; SAS Institute Inc, Cary, NC). All continuous variables were reported as the median (25th, 75th percentile) or were indicated as median (range). P<0.05 was considered statistically significant. Kaplan-Meier cumulative estimates were used to analyze cumulative postoperative event-free survival. Time to first event was characterized by the time to first appropriate ICD therapy, ACA, or sudden cardiac death. For those without a BCE postoperatively, January 1, 2012, was considered the end point for follow-up evaluation, and they were censored for Kaplan-Meier analysis. Event-free survival curves were calculated for the entire cohort, and comparisons were made between asymptomatic patients (ie, 0 cardiac events pre-LCSD) and previously symptomatic patients with 1 to 10 and >10 cardiac events pre-LCSD.

**Results**

Among the 91 patients who underwent videoscopic LCSD at our institution, 27 patients (14 women; median age at diagnosis, 12 [4–16] years; 19 genotype positive) had high-risk malignant ventricular arrhythmia syndromes other than autosomal dominant/sporadic LQTS (Tables 1 and 2). Their underlying causes included CPVT (n=13), Jervell and Lange-Nielsen syndrome (JLNS; n=5), idiopathic ventricular fibrillation (IVF; n=4), left ventricular noncompaction (LVNC; n=2), hypertrophic cardiomyopathy (HCM; n=1), ischemic cardiomyopathy (ICM; n=1), and arhythmogenic right ventricular cardiomyopathy (ARVC; n=1) (Figure 1). Their median age at LCSD was 15 (9–25) years (range, 2 months to 56 years), and the median follow-up post-LCSD was 1.2 (0.5–2.1) years (range, 1 month to 6.2 years). In general, those patients with IVF, ARVC, and ICM were considerably older at age of diagnosis (36 [21–38] years) and at LCSD (48 [30–52] years) than those with CPVT, JLNS, LVNC, and HCM (9 [3–14] years at diagnosis and 14 [9–18] years at denervation). In our cohort, 17 (63%) were pediatric patients.

The majority of patients were taking β-blockers before LCSD (93%); 33% were being treated with sodium channel blockers, 11% with potassium channel blockers, and 48% had an ICD in place before LCSD. Post-LCSD, 59% of the total cohort and 73% of the secondary prevention cohort had an ICD in place. For 27% (n=6) of patients without ICDs in the secondary prevention group, monitoring of BCEs was based on symptom reporting of syncope and ACA and annual 24-hour ambulatory ECG monitoring.

Five asymptomatic patients (19%; 3 with CPVT, 2 with JLNS) had LCSD performed after high-risk assessment and β-blocker intolerance, and none had a sentinel BCE at early follow-up. Among the remaining 22 who had LCSD as secondary prevention, 18 had no recurrences so far, whereas 4 (18%; 1 woman, 1 with CPVT, 1 with JLNS, 1 with HCM, and 1 with IVF) have experienced ≥1 BCE post-LCSD (Figure 2).

Of the 13 patients with CPVT, all were genotype positive for mutations in the CPVT-associated RYR2-encoded cardiac ryanodine receptor or CASQ2-encoded calsequestrin protein, 10 had LCSD for secondary prevention, and 12 had no recurrences post-LCSD. In the 5 JLNS patients, 4 were genotype positive with double KCNQ1 mutations (ie, JLNS1), 3 had LCSD as secondary prevention, and 4 have not experienced any event post-LCSD.

All 4 patients with IVF had LCSD as secondary prevention, and 3 have been event free since their procedure, again at early follow-up. Both the patients with LVNC and the patients with ICM and ARVC were symptomatic before LCSD and have been quiescent since the operation.

**Analysis of BCEs**

Although 85% of patients remain event free post-LCSD at early follow-up (1.5 [0.5–2.1] years), 4 patients had ≥1 BCE post-LCSD. However, as shown in Figure 2, all had a marked decrease in events compared with the number of disease-attributable cardiac events recorded predenervation. The individual characteristics of these 4 patients are illustrated in Table 2.

**Event-Free Survival**

A Kaplan-Meier analysis of cardiac event-free survival for the complete cohort demonstrated 76% of patients to be event free postprocedure (Figure 3). All asymptomatic patients pre-LCSD maintained a 100% event-free survival at short follow-up. When comparing event-free survival based on the number of events preprocedure, the subset with >10 events before denervation had a 55% chance of event-free survival compared with 80% in the group with 1 to 10 events before surgery (data not shown). Although no statistically significant difference was reached, the data suggest that patients with heavier event burden preoperatively might be at higher risk of having BCEs post-LCSD.
Postoperative Complications

Short-term complications predominantly included transient symptoms of Horner syndrome (ie, left eyelid ptosis and miosis) in 3 and mild left-sided apical pneumothorax in 3, with spontaneous resolution in 2 and a chest tube required in 1. All these complications resolved at follow-up. One patient’s procedure was converted to an open thoracotomy because of a bleeding intercostal vessel. No long-term complications were present in our cohort, and technical success was 100%.

Discussion

Although LCSD is an increasingly recognized and well-established treatment option in LQTS, its role in other arrhythmogenic channelopathies and cardiomyopathies is less understood. Here, we report our single-center experience in performing LCSD for patients with malignant arrhythmias, including those with CPVT, JLNS, IFV, LVNC, HCM, ICM, and ARVC. Overall, we demonstrate a marked decrease in the number of cardiac events in the majority of these patients postoperatively. Because LCSD increases the fibrillatory threshold and decreases arrhythmogenicity in the myocardium after an acute ischemic event,\textsuperscript{1,5} LCSD may be a potential adjuvant treatment in patients with malignant ventricular arrhythmias stemming from substrates besides LQTS. In addition, LCSD should be considered in arrhythmogenic conditions, which may be exacerbated specifically by sympathetic activation.

Catecholaminergic Polymorphic Ventricular Tachycardia

CPVT is an autosomal-dominant arrhythmia syndrome caused by disruptions in calcium homeostasis, specifically by mutations in the RYR2-encoded cardiac ryanodine receptor (CPVT1) and more rarely in the CASQ2-encoded calsequestrin protein (CPVT2).\textsuperscript{14,15} Patients with CPVT usually have a normal resting ECG with ventricular ectopy during exercise or catecholamine stress testing, clinically presenting with exertional syncope or sudden death.\textsuperscript{16} Most CPVT patients are treated pharmacologically with \( \beta \)-blockers, calcium channel blockers, flecainide, or a combination thereof. However, some patients do not exhibit a sufficient response to \( \beta \)-blockade and remain symptomatic. In a meta-analysis of 11 studies including 354 patients with CPVT treated with \( \beta \)-blockers, the estimated 8-year rate of arrhythmic events was 37.2% (95% CI, 16.6%–57.7%), near-fatal events was 15.3% (95% CI, 7.4%–23.3%), and fatal events was 6.4% (95% CI, 3.2%–9.6%).\textsuperscript{17} For those patients who are not fully protected with \( \beta \)-blockade, ICDs are generally recommended as additional therapy.\textsuperscript{7} However, given the nature of CPVT, both the fear and pain elicited from an ICD discharge can result in further catecholamine release and cardiac events, ultimately resulting in the administration of multiple shocks in what is called an ICD storm.\textsuperscript{5} In this vicious cycle, the patient may not always be rescued. In addition, inappropriate ICD shocks may trigger an ICD storm. If the patient has sympathetic denervation, this cycle theoretically is short circuited, making LCSD a useful adjunctive therapy for patients with CPVT.

For patients with CPVT who continue to be symptomatic despite \( \beta \)-blocker use, LCSD has been shown to be a safe and effective antifibrillatory therapy in 2 previous case series\textsuperscript{15,18} and 1 case report,\textsuperscript{19} in addition to our success with 13 patients. Wilde et al\textsuperscript{15} had a highly successful outcome with 3 patients with CPVT; Atallah et al\textsuperscript{18} demonstrated complete resolution of...
cardiac events in 3 of 4 patients and partial resolution in 1; and Scott et al described a successful treatment of 1 patient with CPVT with bilateral sympathetic denervation. Here, we present 13 CPVT patients, with thus far no BCEs after LCSD in 12 of the 13 patients and a reduction in events shown in the other patient. Although the average follow-up for these patients is only 1.3 (range, 0.1–4.1) years, these early results are encouraging.

Because LCSD involves postganglionic and preganglionic denervation, postoperative supersensitivity and reinnervation are highly unlikely, and thus the antifibrillatory effects of this procedure should be durable. Although there is no long-term follow-up for patients with CPVT who had LCSD, there are 30-year data on LQTS patients, which demonstrates the successful longevity of this procedure. Denervation may be of additional benefit to those who are poorly compliant with pharmacotherapy. LCSD does not, however, eliminate the need for ICDs for sudden cardiac death prevention in the subjects at highest risk and must not be misconstrued as a therapeutic cure. Indeed, they should be used together, along with β-blockers, to ensure the highest level of protection in patients with CPVT who are deemed at greatest risk. Without treatment altogether, there is a 30% to 50% lethality rate by age 40 years in patients with CPVT.7

### Table 2. Individual Characteristics of All Patients

<table>
<thead>
<tr>
<th>Case No.</th>
<th>Age at LCSD, y</th>
<th>Age at Diagnosis, y</th>
<th>Male/Female</th>
<th>Follow-Up Time, y</th>
<th>No. of Cardiac Events Pre-LCSD</th>
<th>No. of Cardiac Events Post-LCSD</th>
<th>Surgical Complications*</th>
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</tr>
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<td>18</td>
<td>15</td>
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LCSD indicates left cardiac sympathetic denervation; CPVT, catecholaminergic polymorphic ventricular tachycardia; JLNS, Jervell and Lange-Nielsen syndrome; IVF, idiopathic ventricular fibrillation; LVNC, left ventricular noncompaction; HCM, hypertrophic cardiomyopathy; ICM, ischemic cardiomyopathy; ARVC, arrhythmogenic right ventricular cardiomyopathy.

*No long-term complications in any of the 27 patients.
Jervell and Lange-Nielsen Syndrome

JLNS is an autosomal recessive variant of congenital LQTS associated with deafness and a more malignant arrhythmia phenotype. In a study of 186 patients with JLNS, the corrected QT interval was markedly prolonged (>550 ms), 86% of the cohort had a cardiac event, and of those, 95% were triggered by emotional or physical stress. More than 25% of these patients experienced sudden cardiac death. Because of the severity of this phenotype, pharmacological therapy alone is often inadequate, and more aggressive measures need to be pursued. Because cardiac events in JLNS can be triggered by stress, LCSD could be hypothesized to be highly efficacious. In addition, with >90% efficacy in LQTS, it can be expected that LCSD would be highly successful in JLNS.

In our study, we performed LCSD on 5 patients with JLNS, of whom only 1 suffered post-LCSD (follow-up, 1.8 [range, 0.5–4.8] years). In the study by Atallah et al.,2 patients with JLNS were identified: 1 had resolution of symptoms post-LCSD, whereas the other experienced 1 event during the 26-month follow-up. Two other case reports have demonstrated a reduction in symptoms in patients with JLNS after LCSD: 1 with no postoperative symptoms and the other demonstrating ventricular ectopy immediately after surgery but not since the 18-month follow-up.21,22 Again, LCSD should not preclude ICD placement in a patient with JLNS who has been assessed to be at high risk.

Idiopathic Ventricular Fibrillation

IVF is a diagnosis of exclusion identified in patients with structurally normal hearts. These patients generally present later in life, at a mean age of 36 years,23 and have an excellent prognosis as long as the VF is controlled. Therefore, an ICD along with β-blockade is often advised.23–26 In addition, radiofrequency ablation can be used in IVF, and LCSD may also be of benefit. In our 4 patients with IVF, 3 remained symptom-free and 1 had 1 BCE but has an apparently attenuated arrhythmogenic substrate (>200 events over 12 years pre-LCSD [17 events/year] to 1 event post-LCSD in 2.5-year follow-up [0.4 events/year]).

Cardiomyopathies

All the other diseases in our LCSD cohort were cardiomyopathies with severely arrhythmogenic presentations: LVNC, HCM, ICM, and ARVC. In a study conducted by Bourke et al.,2 LCSD was performed in 9 patients who had recalcitrant ventricular arrhythmias, with a decrease in arrhythmia burden in 5 patients. In their study, 2 patients had ICM, with partial improvement in 1 and no measurable reduction in the other. Similarly, 2 patients had HCM, with 1 exhibiting no response to LCSD and the other having a full response with no further ventricular tachycardia at early follow-up. There was also a patient with ARVC who demonstrated complete resolution of symptoms post-LCSD. The other 4 patients with LCSD in this...
cohort included 2 with non-ICM (1 with complete response and 1 with no response) and 2 with sarcoid cardiomyopathy (1 with partial response and 1 with no response). Considering that LCSD’s antibrillatory effect may be somewhat substrate independent, it seems reasonable to consider denervation therapy for some of these malignant cardiomyopathies where life-threatening ventricular arrhythmias persist despite conventional therapy.

Limitations

Due to the nature of tertiary referral to Mayo Clinic’s Congenital Heart Center and the retrospective nature of this study, a time-indexed arrhythmia burden was nearly impossible to obtain from our cohort because a significant number of patients were not able to recollect the exact date of the first arrhythmic event or number of events that occurred before LCSD. Because uniform data were not available for all patients, quantifying this in a time-indexed manner would certainly lead to misinterpretation and be subject to investigator bias.

Conclusions

The early follow-up of LCSD seems promising, with a marked reduction in the frequency of cardiac events observed post-denervation. LCSD should be considered as part of an expanded treatment armamentarium for patients with highly arrhythmogenic non-LQTS channelopathies and cardiomyopathies who remain symptomatic despite standard therapy. However, larger studies for each disease are required before making any definitive recommendations. In addition, a randomized trial or comparison between patients with LCSD and patients without would contribute significantly to our understanding of LCSD efficacy.

Sources of Funding

The Center for Translational Science Activities (CTSA) at Mayo Clinic was consulted for statistical analysis and supported by National Institutes of Health (NIH)/National Center for Research Resources CTSA Grant Number UL1 RR024150. Its contents are solely the responsibility of the authors and do not necessarily represent the official views of the NIH.

Disclosures

Michael J. Ackerman, MD, PhD, is a consultant for Transgenomic and chairs their FAMILION Medical/Scientific Advisory Board (approved by Mayo Clinic’s Medical-Industry Relations Office and Conflict of Interests Review Board). In addition, cardiac channel gene screen and know-how relating to long-QT genetic testing license agreements, resulting in consideration and royalty payments, were established between Genaissance Pharmaceuticals (then PGxHealth and now Transgenomic) and Mayo Medical Ventures (now Mayo Clinic Health Solutions) in 2004. Dr Ackerman, is also a consultant for Biotronik, Boston Scientific Corporation, Medtronic, and St. Jude Medical Inc. However, none of these entities provided financial support for this study. The other authors have no conflicts to report.

References


LCSD in Non-LQTS Arrhythmia Syndromes

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CLINICAL PERSPECTIVE

Left cardiac sympathetic denervation is useful in some patients with long-QT syndrome, but its role in controlling life-threatening ventricular arrhythmias in other arrhythmogenic disorders is less clear. We report the use of left cardiac sympathetic denervation in 27 patients with a spectrum of arrhythmia syndromes, including catecholaminergic polymorphic ventricular tachycardia, idiopathic ventricular fibrillation, and cardiomyopathies. Left cardiac sympathetic denervation was performed as primary preventive therapy in 5 patients who had a high-risk profile and β-blocker intolerance, and all are free of arrhythmic events thus far. In 22 patients with previous arrhythmias, 18 are free of recurrent arrhythmias and the frequency of recurrences was reduced in 4. Thus, left cardiac sympathetic denervation may represent a substrate-independent antifibrillatory treatment option for some patients with life-threatening ventricular arrhythmia syndromes other than long-QT syndrome.
Videoscopic Left Cardiac Sympathetic Denervation for Patients With Recurrent Ventricular Fibrillation/Malignant Ventricular Arrhythmia Syndromes Besides Congenital Long-QT Syndrome

Mira A. Coleman, J. Martijn Bos, Jonathan N. Johnson, Heidi J. Owen, Claude Deschamps, Christopher Moir and Michael J. Ackerman

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