Left thoracoscopic sympathectomy for cardiac denervation in patients with life-threatening ventricular arrhythmias

Sophie C. Hofferberth, MBBS, BSc,^a Frank Cecchin, MD,^b Dan Loberman, MD,^a and Francis Fynn-Thompson, MD^a

Background: We reported the outcomes of a single-institution experience using video-assisted thoracoscopic left cardiac sympathetic denervation as an adjunctive therapeutic technique in pediatric and young adult patients with life-threatening ventricular arrhythmias.

Methods: We conducted a retrospective clinical review of all patients who underwent left cardiac sympathetic denervation by means of video-assisted thoracoscopic surgery at our institution. From August 2000 to December 2011, 24 patients (13 with long QT syndrome, 9 with catecholaminergic polymorphic ventricular tachycardia, and 2 with idiopathic ventricular tachycardia) were identified from the cardiology database and surgical records.

Results: There were no intraoperative complications. The median postoperative length of stay was 2 days (range, 1-32 days). There were no major perioperative complications. Longer-term follow-up was available in 22 of 24 patients at a median follow-up of 28 months (range, 4-131 months). Sixteen (73%) of the 22 patients experienced a marked reduction in their arrhythmia burden, with 12 (55%) becoming completely arrhythmia free after sympathectomy. Six (27%) of the patients were nonresponsive to treatment; each had persistent symptoms at follow-up.

Conclusions: Video-assisted thoracoscopic left cardiac sympathetic denervation can be safely and effectively performed in most patients with life-threatening ventricular arrhythmias. This minimally invasive procedure is a promising adjunctive therapeutic option that achieves a beneficial response in most symptomatic patients. These results support the inclusion of thoracoscopic cardiac sympathetic denervation among the treatment armamentarium in all patients with ventricular arrhythmias refractive to conventional medical therapy. (*J Thorac Cardiovasc Surg* 2014;147:404-11)

The sympathetic nervous system plays a prominent role in the genesis of many life-threatening ventricular arrhythmias. β -Blockade is the mainstay of therapy; however, despite reducing arrhythmia frequency, many patients experience persistent cardiac events and remain at risk of sudden death.^{1,2} Intolerance to antiarrhythmic agents is also a common problem.¹ In some patients, a more aggressive strategy to prevent sudden cardiac death is required. Implantable cardioverter-defibrillator (ICD) implantation has frequently been used in these cases. Although ICDs effectively protect against lethal arrhythmias, associated morbidities are prominent and include procedural risks, device malfunction, inappropriate shocks, and psychological

distress, particularly among adolescents.³ Left cardiac sympathetic denervation (LCSD), first described in 1971,⁴ has been a safe and effective procedure to reduce fatal arrhythmias and prevent cardiac death. The surgical technique has undergone several modifications,⁵ with variations in strategy among different centers.

Video-assisted thoracoscopic (VATS) LCSD was first reported by Reardon and colleagues⁶ in 2000, with Li and colleagues⁷ reporting the first small series in patients with LQTS in 2003. Several groups have demonstrated positive intermediate outcomes using adjunctive thoracoscopic LCSD to treat intractable ventricular arrhythmias in pediatric patients,⁸⁻¹¹ with the largest experience in children with refractory LQTS (long QT syndrome).^{8-10,12} The indications for performing VATS-LCSD continue to evolve, with recent reports demonstrating its utility in children with non-LQTS arrhythmias.^{2,8,12} Herein, we report our single-center experience performing adjunctive VATS-LCSD in 24 pediatric and young adult patients with life-threatening ventricular arrhythmias.

PATIENTS AND METHODS

This retrospective study was performed after approval from the Institutional Review Board. Patients were identified using the cardiology database and surgical notes. Medical records were reviewed for baseline

From the Departments of Cardiac Surgery^a and Cardiology,^b Boston Children's Hospital, Harvard Medical School, Boston, Mass.

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Address for reprints: Francis Fynn-Thompson, MD, Department of Cardiac Surgery, Boston Children's Hospital, 300 Longwood Ave, Boston, MA 02115 (E-mail: francis.fynn-thompson@cardio.chboston.org).

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Abbreviations and Acronyms

CPVT	= catecholaminergic polymorphic ventricular tachycardia
ICD	= implantable cardioverter-defibrillator
IVT	= idiopathic ventricular tachycardia
JLNS	= Jervell and Lange-Nielsen syndrome
LCSD	= left cardiac sympathetic denervation
LQTS	= long QT syndrome
VATS	= video-assisted thoracoscopic
VF	= ventricular fibrillation
VT	= ventricular tachycardia

characteristics, treatment indications, details of operative strategy, intraoperative events, and postoperative course. Between August 2000 and December 2011, VATS-LCSD was performed in a total of 24 patients (11 males; median age, 13 years; range, 5 weeks to 27 years) at Boston Children's Hospital (Boston, Mass). The cohort includes the 9 patients in our initial study from 2008 reporting the VATS-LCSD technique.⁸ Thirteen were diagnosed with congenital LQTS (2 had *Jervell and Lange-Nielsen syndrome*, defined as severe QT prolongation and congenital hearing loss), 9 with catecholaminergic polymorphic ventricular tachycardia (CPVT), and 2 with idiopathic recalcitrant ventricular tachycardia (VT).

Surgical Technique

In all patients, LCSD was performed via a left-sided VATS approach under general anesthesia. The operative technique is largely unchanged from that described previously,⁸ with the following exceptions: dissection is performed using a harmonic scalpel in place of electrocautery, and chest tube placement in the left pleural cavity is no longer undertaken. The stellate ganglion was intentionally spared in 23 of the 24 patients.

Patient Descriptions

Long QT syndrome. Thirteen patients were identified with LQTS (Table 1). The median age at LCSD was 8 years (range, 2-22 years). Six (patients 1, 2, 4, 5, 6, and 8; Table 1) of the 13 had undergone previous ICD implantation. Indications for LCSD included delivery of recurrent appropriate ICD shocks despite optimal antiarrhythmic therapy in 4 (patients 4, 5, 6, and 8), ongoing arrhythmic events despite optimal antiarrhythmic therapy in 4 (patients 9, 11, 12, and 13), failed medical therapy (unable to tolerate β -blocker dose increase) in 3 (patients 1, 2, and 3), with 2 (patients 7 and 10) high-risk patients treated prophylactically. Of the 13 patients, 9 had available genotypic information: 7 were LQTS-1 and 2 were LQTS-2 genotype.

Catecholaminergic Polymorphic VT. Nine patients were identified with CPVT (Table 1). The median age at LCSD was 17 years (range, 8-27 years). Seven had prior ICD implantation, with 1 (patient 19) also receiving a dual-chamber pacemaker. Delivery of recurrent ICD shocks despite optimal medical therapy was the indication for LCSD in 6 of these 7 patients. Patient 17 had recurrent ICD shocks associated with failure to tolerate medical therapy. The remaining 2 (patients 14 and 21) were symptomatic despite optimal medical therapy.

Intractable VT. Two patients (patients 23 and 24) had intractable VT. Both had persistent symptoms despite optimized medical therapy. Patient 23 had recurrent VT despite maximal antiarrhythmic therapy and multiple electrophysiology studies for ablation of a left-sided accessory pathway and 3 distinct VT foci. Follow-up was limited to the time of discharge, and he had ongoing runs of VT during a prolonged postoperative hospital

stay. Patient 24 was treated at 5 weeks of age in the setting of recalcitrant ventricular arrhythmias requiring repeated defibrillation.

RESULTS

Left Cardiac Sympathetic Denervation and Video-Assisted Thoracoscopic Surgery

Video-assisted thoracoscopic LCSD was performed in 24 patients. Eight patients received concomitant ICD implantation; 1 received a dual-chamber pacemaker. There were no intraoperative complications, and blood loss was minimal. Continuous cardiac rhythm monitoring (telemetry) was used in all patients postoperatively, with a pediatric electrophysiologist reviewing the telemetry twice daily. Two (9%) of the patients experienced arrhythmias before discharge. Patient 2 developed VT necessitating cardioversion and began taking esmolol and magnesium infusion. No further arrhythmic episodes occurred. Patient 17 experienced recurrent runs of VT, which led to a prolonged hospital stay of 32 days.

There were no major postoperative complications. Minor postoperative complications occurred in 3 (13%) of the patients. Patient 16 developed a small apical pneumothorax, necessitating chest tube suction. She was discharged 1 day later. Patient 18 developed a small left apical pneumothorax and was successfully treated with 24 hours of oxygen therapy. Her length of stay was 2 days. Patient 8 developed prominent harlequin facial flushing, but not Horner syndrome, and this had resolved at follow-up. She is the only patient to have had the left stellate ganglion included in the LCSD. No patients developed Horner syndrome. Eleven patients were initially managed in the postoperative intensive care unit (ICU). The median length of postoperative hospital stay was 2 days (range, 1-32 days).

Follow-up Outcomes

Long QT syndrome. Of the 13 patients with LQTS, 4 were treated with LCSD for recurrent appropriate ICD shocks despite optimal medical therapy. Of the 4 patients, 2 were symptom free post-LCSD. Patient 4 was event free at 33 months, whereas patient 8 had complete resolution of her frequent VT episodes at 131 months post-LCSD treatment (Table 2). One patient (patient 5) demonstrated some reduction in arrhythmia burden. She experienced multiple ICD discharges in the months before LCSD, and at latest follow-up, she had experienced 2 further ICD firings and required 4 antiarrhythmic medications. The fourth patient treated for recurrent ICD discharges (patient 6) did not respond to LCSD, experiencing ongoing recurrent ICD shocks at 23 months. In the 4 patients who were symptomatic despite optimal medical therapy, patients 11 and 12 were both asymptomatic and required no antiarrhythmic therapy at 31- and 29-month follow-up, respectively. Patient 13 experienced 2 ICD shocks during 13 months of follow-up, although this occurred in the setting of poor medication

TABLE 1. Patient baseline characteristics

Patient no.	Sex	Age, y	Diagnosis	Initial presentation	Antiarrhythmic medication(s)	Daily dose, mg/kg	Indication for LCSD
1	M	22	LQTS	Syncopal episodes with VT	Nadolol	NA	Multiple ICD shocks, unable to tolerate medication increase
2	F	22	LQTS	VF arrest	Labetalol	7.6	Multiple ICD shocks, unable to tolerate medication increase
3	F	19	LQTS	Single syncopal episode	Betaxolol	0.2	Failed medical therapy
4	F	22	LQTS	Syncopal episodes with VT	Labetalol	3.7	Multiple recent ICD shocks, optimal medical therapy
5	F	15	LQTS	Recurrent VT/TdP	Nadolol Labetalol	1.1 16.0	Recurrent VT, maximal medical therapy
6	M	4	LQTS	VT	Nadolol NA	0.8 NA	Multiple ICD shocks for VT, on optimal medical therapy
7	M	4	LQTS (JLN)	Asymptomatic	Atenolol	NA	Primary prevention
8	F	6	LQTS (JLN)	Recurrent TdP	Nadolol	2	Multiple ICD shocks for TdP
9	M	3	LQTS	Aborted SCD	Propranolol	2.5	Multiple aborted SCDs despite optimal medical therapy
10	F	2	LQTS	Aborted SCD	Nadolol	0.8	Primary prevention
11	M	10	LQTS	TdP, cardiac arrest	Esmolol	0.4	Frequent VT/TdP, symptomatic despite optimal medical therapy
12	M	8	LQTS	Repeated episodes of LOC	Nadolol	NA	Symptomatic despite medical therapy
13	F	2	LQTS	Aborted SCD with VT	Nadolol	2.8	Recent VT arrest
14	M	21	CPVT	Polymorphic VT	Betaxolol	1.6	Symptomatic on optimal medical therapy
15	M	15	CPVT	VT, syncopal episodes	Atenolol	0.8	Multiple ICD shocks, on optimal medical therapy
16	F	19	CPVT	Aborted SCD and VT	Atenolol	2.1	Multiple ICD shocks, on optimal medical therapy
17	M	11	CPVT	VT	Nadolol	1.5	Multiple AICD shocks for VT, poor medication tolerance
18	F	15	CPVT	Family history, asymptomatic	Atenolol	0.8	Multiple ICD shocks for VT, on optimal medical therapy
19	F	27	CPVT	Exercise-induced syncope, VT	Labetalol	6.3	Multiple ICD shocks for VT, optimal medical therapy
20	M	17	CPVT	VF arrest	NA	NA	Multiple ICD shocks for VT/VF with optimal medical therapy
21	F	8	CPVT	Syncopal episode	Nadolol	1.2	Symptomatic despite optimal medical therapy
22	M	17	CPVT	Exercise-induced syncope, VT	Nadolol	2.4	Persistent ICD shocks for VT, optimal medical therapy
23	M	2	IVT	Syncopal episodes with VT	Esmolol	NA	Intractable VT despite multiple ablations, medical therapy
24	F	5 wk	IVT	VT, aborted SCD with VF	Amiodarone	NA	Symptomatic despite optimal medical therapy

LCSD, Left cardiac sympathetic denervation; LQTS, long QT syndrome (without congenital deafness); VT, ventricular fibrillation; NA, not available; ICD, implantable cardioverter-defibrillator; VF, ventricular fibrillation; TdP, torsades de pointes; JLN, Jervell Lange-Nielson-type LQTS; SCD, sudden cardiac death; LOC, loss of consciousness; CPVT, catecholaminergic polymorphic ventricular tachycardia; AICD, automatic implantable cardioverter defibrillator; IVT, idiopathic ventricular tachycardia.

compliance. This patient had experienced a ventricular fibrillation (VF) arrest and multiple runs of VT in the months preceding LCSD intervention. Patient 9 was lost to follow-up. The 3 (patients 1, 2, and 3) treated with LCSD after failing medical therapy were all asymptomatic

at latest follow-up, with patients 1 and 4 not requiring any antiarrhythmic medications. Of the 2 (patients 7 and 10) treated prophylactically with LCSD and concomitant ICD implantation, patient 7 had experienced 3 ICD shocks and was being managed on a single antiarrhythmic agent at 82

TABLE 2. Patient outcomes

No.	QTc before/after, ms	Hospital stay, d	Follow-up, mo	Response to LCSD therapy
1	467/460	1	4	No events, no medications
2	467/422	4	37	No events, tolerating 3 antiarrhythmic agents
3	472/497	1	27	Asymptomatic, no medications
4	520/537	1	33	No events, 2 antiarrhythmic agents
5	542/560	1	11	2 ICD firings/year, 4 antiarrhythmic agents
6	392/479	1	23	Multiple ICD shocks/year, 2 antiarrhythmic agents
7	535/515	5	82	3 events in 6 years, 1 low-dose antiarrhythmic
8	630/588	2	131	No ICD shocks, 2 antiarrhythmic agents
9	476/456	5	1	NA
10	462/NA	4	26	6 episodes of TdP, increased antiarrhythmic
11	600/500	6	31	Asymptomatic, no medications
12	600/503	3	29	Asymptomatic, no medications
13	566/530	2	13	2 ICD shocks, 1 antiarrhythmic agent
14	461/421	1	13	No events, 2 antiarrhythmic agents
15	394/364	1	39	1 ICD firing/year, 1 antiarrhythmic agent
16	433/410	2	37	No events, 1 antiarrhythmic agent
17	422/399	1	47	1 ICD firing/year, 1 antiarrhythmic agent
18	419/NA	2	51	No events, medications unknown
19	403/412	2	7	No events, 3 antiarrhythmic agents
20	447/463	2	34	3 events in 3 years, 2 antiarrhythmic agents
21	399/NA	1	12	Asymptomatic, 1 antiarrhythmic agent
22	360/372	1	15	No events, 1 antiarrhythmic agent
23	506/NA	32	1	NA
24	440/396	3	46	No events, 2 antiarrhythmic agents

The mean $\pm$ SD QTc, before/after, is $476 \pm 72/465 \pm 61$ ms. QTc, corrected QT interval; LCSD, left cardiac sympathetic denervation; ICD, implantable cardioverter-defibrillator; NA, not available; TdP, torsades de pointes.

months. Patient 10 experienced 6 episodes of torsades de pointes and required increasing β -blockade at 26 months post-LCSD (Table 2).

Catecholaminergic polymorphic VT. Of the 9 patients with CPVT, 6 underwent VATS-LCSD for recurrent appropriate ICD shocks despite optimal antiarrhythmic therapy. Patients 16, 18, 19, and 22 had no further ICD shocks at 37, 51, 7, and 15 months' follow-up, respectively (Table 2). Of the remaining 2 patients with this indication, patient 15 averaged 1 ICD shock per year and was managed on 1 antiarrhythmic agent at 39 months post-LCSD. Patient 20 experienced 1 ICD shock per year over 34 months, with stable doses of 2 antiarrhythmic agents. Two patients (patients 14 and 21) underwent LCSD for persistent symptoms despite optimal medical therapy; both were asymptomatic at 13 and 12 months, respectively. In 1 (patient 17), LCSD was indicated in the setting of recurrent ICD shocks with failure to tolerate escalating β -blockade. At 47 months post-LCSD, he averaged 1 ICD shock per year, and was being treated with a single antiarrhythmic agent.

Intractable VT. Two patients with idiopathic ventricular tachycardia (IVT) were treated with LCSD and concomitant ICD implantation for persistent symptoms despite optimal medical therapy. Patient 23 was lost to follow-up, whereas patient 24 was event free at 46 months post-LCSD procedure.

Overall Response to LCSD Therapy

Longer-term follow-up was available in 22 of the 24 patients at a median of 28 months (range, 4–131 months). Two patients referred from overseas centers were lost to follow-up. In 10 (4 LQTS and 6 CPVT) patients, the indication for LCSD was recurrent ICD shocks despite optimal medical therapy. Of these patients, 8 (80%) experienced a marked reduction in their arrhythmia burden after LCSD, with 6 (60%) becoming arrhythmia free.

Longer-term follow-up was available in 6 of the 8 (4 LQTS, 2 CPVT, and 2 IVT) patients who were symptomatic despite optimal medical therapy before LCSD procedure. Five patients (83%) had a marked reduction in arrhythmia burden, whereas 4 (66%) were arrhythmia free. Patient 22 (LQTS) had 2 ICD shocks in the first year post-LCSD, necessitating an escalation of antiarrhythmic therapy.

Four patients (3 LQTS and 1 CPVT) received LCSD after failing to tolerate medical therapy. Three patients (75%) had a marked reduction in arrhythmia burden; 2 were arrhythmia free and no longer required medical therapy, whereas 1 was able to tolerate lower doses of antiarrhythmic agents while remaining asymptomatic.

Of the 2 patients who received LCSD therapy as a prophylactic measure, 1 had 3 arrhythmic events over 6 years

of follow-up, whereas the other appeared to only marginally benefit from the LCSD. This patient had recurrent arrhythmias and required escalation of medical therapy during the available 2-year follow-up period.

DISCUSSION

Video-assisted thoracoscopic LCSD is becoming increasingly recognized as a safe and effective adjunctive therapy in patients with life-threatening ventricular arrhythmias.^{8-10,12} Herein, we describe our single-center experience using the minimally invasive VATS-LCSD approach in children and young adults with malignant ventricular arrhythmias. Of the 24 patients in this cohort, 13 had LQTS (including 2 patients with Jervell and Lange-Nielsen syndrome) and 9 had CPVT, with 2 diagnosed with idiopathic VT. Patient selection for LCSD therapy consisted of 3 subgroups of patients, presenting with 1 of 3 characteristics: persistent symptoms (defined as recurrent appropriate ICD shocks despite optimal medical therapy or arrhythmic events, despite optimal medical therapy), failure to tolerate medical therapy, or prophylactic therapy. The absence of procedure-related complications, and minimal ($n = 3$) incidence of mild/transient postoperative complications, is consistent with previous reports^{8,9} using the minimally invasive VATS-LCSD approach. Overall, we demonstrate VATS-LCSD is a low-morbidity procedure that achieves a marked response in most patients with symptomatic life-threatening arrhythmias, irrespective of underlying arrhythmogenic etiology.

Left Cardiac Sympathetic Denervation and Video-Assisted Thoracoscopic Surgery

Left cardiac sympathetic denervation was initially described by Moss and McDonald⁴ in 1971 to treat patients with medically refractory LQTS. Animal and human studies, performed soon thereafter, suggested left cardiac sympathetic nerves are proarrhythmogenic, because the ventricular refractory period was prolonged and fibrillation thresholds increased after stellate ganglia denervation.^{13,14} Stimulation of the left stellate ganglion has also triggered T-wave alternans, which is frequently associated with LQTS.¹³⁻¹⁵ Because LCSD involves both preganglionic and postganglionic denervation, postoperative supersensitivity and reinnervation are unlikely to occur, thereby ensuring the antifibrillatory effects are permanent. Permanence is important in the context of poor medication compliance, an issue of particular relevance in the adolescent population. With greater understanding of its antifibrillatory properties, LCSD has emerged as a safe adjunctive therapeutic option for patients with ventricular arrhythmias refractory to standard therapy.^{5,9,16} The largest experience is a multicenter series reporting more than 80% reduction in cardiac events post-LCSD in 147 patients with LQTS.⁵

The LCSD procedure has undergone several modifications since it was first described. Variations in surgical technique include high-thoracic left sympathectomy,¹³ left cervicothoracic sympathectomies (stellate ganglion and upper thoracic ganglia), bilateral sympathectomies, and a posterior approach.¹⁷⁻¹⁹ Since VATS-LCSD was first reported by Reardon and colleagues⁶ in 2000, several small series have used the technique in pediatric patients with intractable ventricular arrhythmias.⁸⁻¹¹ Most of the global experience has been in patients with refractory LQTS. Nearly 80% of all reported cases using VATS-LCSD in patients with LQTS demonstrate resolution of arrhythmias/symptoms at 1 year.⁸⁻¹¹ Similarly, it has effectively treated CPVT.^{8,9,12,16} A smaller experience exists in patients with Jervell and Lange-Nielsen syndrome (JLNS), a more malignant autosomal recessive variant of congenital LQTS, that is associated with deafness. Because of the severity of this phenotype, pharmacologic therapy alone is often inadequate and more aggressive treatment is required.²⁰ Both of the series that included patients with JLNS^{8,12} report a high response rate; in the study by Coleman and colleagues,¹² only 1 of 5 patients with JLNS experienced a cardiac event post-LCSD. Two of the patients in our initial series had JLNS; one achieved complete resolution of symptoms, whereas the other experienced 1 event at 26-month follow-up.⁸ These results are not surprising given the marked response rate among patients with LQTS. Although fewer data are available for patients with IVT, it appears VATS-LCSD offers benefit to this population also.^{8,12} Among all reported series undertaking VATS-LCSD, procedural morbidity and surgical complication rates, including operative blood loss, have been minimal.^{8-10,12} The outcomes of this study confirm that VATS sympathectomy is a safe and feasible therapeutic option that carries minimal procedural risk.

Indications for Treatment and Outcome of LCSD

Symptomatic patients. This report documents an overall response rate of almost 80% in patients who were symptomatic at LCSD intervention. Patients treated after experiencing multiple ICD discharges despite optimal medical therapy, and those who had persistent symptoms despite optimal medical therapy, received the most benefit from adjunctive LCSD therapy.

Failure to tolerate medical therapy. In the 4 patients who developed significant adverse effects at maintenance antiarrhythmic doses or who failed to tolerate escalating β -blocker therapy, 3 appeared to benefit markedly. Performance of adjunctive VATS-LCSD appears a promising strategy to enable this patient subgroup to be maintained at a lower medication dose and still remain symptom free. Furthermore, for some of these patients, adjunctive LCSD may eliminate the requirement for ongoing antiarrhythmic therapy altogether.

Asymptomatic, primary prevention. Two patients (patients 13 and 18) in this cohort underwent VATS-LCSD as a prophylactic measure. The outcomes were equivocal as to whether any real benefit was derived from undergoing the procedure. With such limited experience and indeterminate results, our data do not support undertaking prophylactic VATS-LCSD in all children identified at high risk of fatal arrhythmias. Nevertheless, there may be a subset of patients who possess a high-risk genotype that would benefit from prophylactic LCSD treatment. A recent study by Jons and colleagues²¹ demonstrated that analysis of mutant-specific ion channel characteristics in LQTS patients may be useful for clinical risk stratification. Further investigation may reveal a specific genotype and phenotype that are associated with a heightened risk profile and would, therefore, benefit from adjunctive VATS-LCSD as a primary preventative strategy. Nevertheless, we cannot recommend LCSD as a prophylactic therapy.

CONCLUSIONS

In summary, we have documented that video-assisted thoroscopic LCSD can be safely and effectively performed in most children and young adults with life-threatening ventricular arrhythmias. This minimally invasive procedure is a promising adjunctive therapeutic option that achieves a beneficial response in most symptomatic patients. We advocate the use of this treatment in all patients who remain symptomatic with recurrent life-threatening arrhythmias, syncope, or frequent ICD discharges, despite conventional medical therapy. This treatment strategy should be considered as part of the treatment armamentarium in all patients with recalcitrant ventricular arrhythmias. The utility of LCSD as a prophylactic therapy in high-risk pediatric patients must be further elucidated before definitive recommendations can be made.

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Discussion

Dr Joseph Dearani (Rochester, Minn). Thank you, Dr Backer and Dr Reddy.

Dr Hofferberth and colleagues have summarized their results of a small series of 24 children with VATS sympathetic denervation to treat life-threatening ventricular arrhythmias. Their technique intentionally spares the entire left stellate ganglion. They demonstrated a “marked reduction” of arrhythmias in 73% and elimination of arrhythmias in 55%. Two were lost to follow-up, and 2 had 1-month follow-up; this should be factored into the recurrence equation.

Surgery was performed safely; however, I believe there are shortcomings with this review. Most centers with the greatest experience in treating these cardiac channelopathies intentionally remove the lower half of the left stellate ganglion. The literature has demonstrated that the optimal cardiac denervation includes the removal of T4, T3, T2, and the lower pole of the left stellate ganglion (T1). In fact, the greatest density of norepinephrine-containing vesicles resides in the stellate ganglion (T1) and a portion of T2. So, ideally, cardiac denervation would include a complete

stectomy. However, the upper pole of the left stellate ganglion is preserved to minimize the potential risk of developing Horner syndrome.

Among the largest programs that perform this specific left cardiac sympathetic denervation operation (ie, taking the lower half of the stellate ganglion and T2 through T4), there exceeds a 90% reduction in arrhythmia burden that includes breakthrough faints and breakthrough ICD shocks overall. Furthermore, the antifibrillatory (ie, protective effect) of denervation therapy is disease and disease genotype dependent, where it has been shown to be most effective in LQT1 and CPVT, emphasizing the importance of genotyping for all of these patients. For example, among the greater than 30 LQT1 patients denervated in our practice, there have been no breakthroughs to date with longer follow-up. Unfortunately, we do not know the genotypes of the long QT patients in this small series. If many or most were LQT1, then the higher observed breakthrough rate further underscores the critical importance of including the lower pole of the left stellate ganglion in the operation.

I believe cardiac denervation surgery for channelopathies should not be viewed as a simple adaptation of minimally invasive surgery performed for hyperhidrosis, and it probably should not be performed at a pace of approximately one procedure per year. Despite performing approximately 20 per year, we still encounter pretty variable anatomic variation with the left stellate ganglion. In addition, in our experience, we have now performed left cardiac sympathetic denervation in over 110 patients with these potentially life-threatening disorders. In the early part of our reported series, there were 3 patients with eyelid ptosis. Importantly, to date, there is no patient with a complete Horner (facial droop), in more than 110 patients.

In closing, I agree with your VATS approach to sympathectomy, and I congratulate your team for performing it with low perioperative morbidity. However, I respectfully disagree with your technique to intentionally spare the left stellate ganglion, leaving up to a third of your patients with residual ventricular arrhythmias, resulting in ICD discharges. This study demonstrates that this technique is less effective for this difficult problem, and I would encourage you to acknowledge this higher incidence in arrhythmia recurrence when the lower half of the stellate ganglion is preserved.

Dr Hofferberth. Thank you very much for your comments, Dr Dearani.

First, I would like to address the issue of the indications for treatment in this study. The vast majority of the literature that exists using the VATS-LCSD procedure is in patients with cardiac ion channelopathies, who are treated for secondary prevention. Looking at our data, we had a 73% response rate among all-comers with 2 patients in the series of 24 treated for primary prevention. If you exclude those 2 patients out, that means that we have an 82% response rate among the patients that were treated for secondary prevention.

Published data from other centers that perform the VATS-LCSD technique certainly show that an 82% response rate is equivalent to the results attained across the entire global experience to date. There was a review article published 2 years ago out of Texas Children's Hospital by Hwang and colleagues, who reviewed all cases using VATS-LCSD procedure in children

with congenital long QT syndrome. This article demonstrated that, of the global experience, there was a 77% response rate, defined in terms of arrhythmia reduction. The vast majority of data available on this topic defines response to treatment as reduced arrhythmia burden. As I alluded to in the presentation, it is difficult to quantify arrhythmia burden; however, that is a limitation across all of the studies in this area. Nevertheless, based on the current criteria for treatment response, our results are equal to the global experience.

Dr Dearani. There is an important difference between a 90% to 95% success rate, with ICD discharges going off in children with refractory ventricular arrhythmias compared to 55% to 70%. This is an evolving science. Genotyping has helped understand expectations in abolishing arrhythmias substantially. We have not had a single case of a permanent Horner syndrome with facial droop in over 100 cases when the lower pole of the stellate is removed. Life-threatening ventricular arrhythmia recurrence, resulting in ICD discharge, is different than recurrence of atrial fibrillation. It is a truly life-threatening problem, and striving for a 90% to 95% arrhythmia reduction rate without causing complete Horner syndrome seems to be a better goal to aim for.

It would be helpful for other surgeons in the audience to comment. As we discussed earlier, general pediatric surgeons and pediatric cardiac surgeons do this procedure. It is important that the surgical community understand the differences and expectations with the various sympathectomy techniques.

Dr Hofferberth. Thank you, Dr Dearani. Our institution does not agree with the notion that removal of part or all of the left stellate ganglion is a morbidity-free approach. The most recent article published out of the Mayo Clinic in 2012, which described their experience performing VATS-LCSD with resection of the lower half of the left stellate ganglion, demonstrates this is not a morbidity-free approach, with just over 10% of patients developing a perioperative Horner syndrome.

There was also an article published last month by Schneider and colleagues from a center in Munich, who reported their results performing LCSD with resection of the lower half of the left stellate ganglion in 10 patients with long QT syndrome. They reported that 7 of the 10 developed perioperative Horner syndrome. Our approach has been to transect the sympathetic chain up, at the base of the stellate ganglion, and in the process remove all of the interior radiating nerve fibers, which is, as you state, the location that releases the highest concentration of norepinephrine. By performing the procedure in this fashion, we are effectively eliminating the morbidity risks associated with this procedure without sacrificing the treatment efficacy.

Dr James Tweddell (Milwaukee, Wis). Thank you. It was a nice presentation. And although I would agree with Dr Dearani that this is not a simple extension of a thoracoscopic sympathectomy for hyperhidrosis, it is probably closer to that than the operations typically performed by most of the people in this room. So, when we have encountered this problem, we have actually asked our adult thoracic surgery colleagues to help us with this procedure. They have tremendous experience with this. And, I think it is easier for us to work with them as a team approach rather than try to reinvent the wheel on these patients; I did enjoy your presentation much.

Dr Hofferberth. Thank you much for your comments, Dr Tweddell.

Dr Laureano Molins (*Barcelona, Spain*). We have experience with sympathectomy or better clipping for hyperhidrosis mainly. We have little experience, 5 or 6 cases with babies. It is difficult to decide if the stellate ganglion should be removed or not because we do not want to have those Horner syndromes.

I think that the theory is to transect the lower part of the stellate ganglion, but it is not easy to respect the whole one. So, in fact, I have not done it, to resect the stellate ganglion, and the percentage of the patients that went well was similar, 75% to 80%.

But, I would like first to congratulate you for your elegant presentation and to talk a little bit about bilateral approach. Could this bilateral approach reach a high level of success? I really do not know. And we begin always with left side, of course. We go through until T6. But, I would like to know not only your experience but your opinion on that.

Dr Hofferberth. Thank you for your comments and question. At this point in time, the experience of performing a bilateral sympathectomy is limited at our center; however, the reason we did include the comment on the conclusion slide is that most recent patients treated with VATS-LCSD at our center had undergone a left-sided sympathectomy and remained symptomatic. In this particular case, we then decided to proceed with a second operation to perform a right-sided sympathectomy, and since that time, that patient has been completely arrhythmia free. It is, obviously, a limited experience to date. However, this may be a strategy that should be considered further in the future.

Dr Carl Backer (*Chicago, Ill*). I have 2 questions. The first relates to a patient of ours with ventricular tachycardia treated by our chief of anesthesia with a temporary sympathetic nerve block in the left neck. This completely cleared up the arrhythmia and then we electively took the patient for a thoracoscopic sympathectomy several days later. Do you have any experience with temporary nerve blocks in the neck as a predictive study for this patient population?

Dr Hofferberth. As far as I am aware, there has not been any experience using that as a temporary measure.

Dr Backer. Yes, and it worked, it was unbelievable, it was night and day.

The second question I have relates to the fact that some of these children are pretty small and putting in an epicardial AICD is not without issues. Can we use sympathectomy as the primary therapy? We had one patient in whom we did the thoracoscopic sympathectomy, the arrhythmias went away, and we observed the child for a long time. We are still discussing whether or not we should put in an AICD. Do you have patients in whom you have simply done the sympathectomy and then not proceeded with an AICD?

Dr Hofferberth. There are certainly patients that have had a sympathectomy with additional medical treatment and who did receive an ICD. We view this procedure as an adjunctive therapeutic strategy that should always be implemented in conjunction with established therapies. In patients treated with VATS-LCSD for primary prevention, we would still treat them with optimal medical therapy; however, there have been some patients who have been spared from undergoing ICD implantation.

Dr Backer. I failed to mention we did treat this patient medically, and we did watch in the hospital for over a week. I noticed your mean hospital stay was 2 days. How long would you watch a patient with a sympathectomy and medical therapy before you would send them out?

Dr Hofferberth. We discharge all patients on medical therapy and then proceed to have them followed up regularly with a cardiologist. So, at this point, a patient that has received a sympathectomy is always going to be on some form of antiarrhythmic therapy.

Dr Backer. Thank you much. This really was an eye-opening presentation. This was not even on my radar screen 10 years ago, and certainly it has now become an effective therapy. Thank you much.