Physical and Psychological Consequences of Left Cardiac Sympathetic Denervation in Long-QT Syndrome and Catecholaminergic Polymorphic Ventricular Tachycardia

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on behalf of the Cardiac Inherited Disease Group New Zealand*

Background—Left cardiac sympathetic denervation reduces risk in long-QT syndrome (LQTS) and catecholaminergic polymorphic ventricular tachycardia. Side effects and patient satisfaction have not been systematically analyzed in patients who underwent left cardiac sympathetic denervation. Aims of this study included documenting physical and psychological consequences and patient satisfaction after left cardiac sympathetic denervation in LQTS or catecholaminergic polymorphic ventricular tachycardia.

Methods and Results—Patients with LQTS (N=40) and catecholaminergic polymorphic ventricular tachycardia (N=7) underwent video-assisted thoracoscopic left cardiac sympathetic denervation, with a median follow-up of 29 months (range, 1–67 months). Clinical records were reviewed; 44 patients completed a telephone survey. Of 47 patients (53%), 25 were preoperatively symptomatic (15 syncpe, 7 near-drowning, and 3 resuscitated sudden death). Indications for left cardiac sympathetic denervation included β-blocker intolerance (15; 32%) or nonadherence (10; 21%) and disease factors (18; 38%; catecholaminergic polymorphic ventricular tachycardia [6], near-drowning [2], exertional syncpe [1], symptoms on therapy [2], LQT3 [1], QTc>520 ms [6]). Other indications were competitive sports participation (2), family history of sudden death (1), and other (1). Median QTc did not change among patients with LQTS (461±60 to 476±54 ms; P=0.49). Side effects were reported by 42 of 44 (95%). Twenty-nine patients (66%) reported dryness on left side, 26 (59%) a Harlequin-type (unilateral) facial flush, 24 (55%) contralateral hyperhidrosis, 17 (39%) differential hand temperatures, 5 (11%) permanent and 4 (9%) transient ptosis, 5 (11%) thermoregulation difficulties, 4 (9%) a sensation of left arm paresthesia, and 3 (7%) sympathetic flight/fright response loss. Majority of the patients were satisfied postoperatively: 38 (86%) were happy with the procedure, 33 (75%) felt safer, 40 (91%) recommended the procedure to others, and 40 (91%) felt happy with their scar appearance.

Conclusions—Despite significant morbidity resulting from left cardiac sympathetic denervation, patients with LQTS and CPVT have high levels of postoperative satisfaction. (Circ Arrhythm Electrophysiol. 2015;8:1151-1158. DOI: 10.1161/CIRCEP.115.003159.)

Key Words: adverse effects ■ long-QT syndrome ■ polymorphic catecholergic ventricular tachycardia ■ sympathectomy ■ video-assisted thoracoscopic surgery

Video-assisted thoracoscopic sympathectomy is used to treat various disorders, most commonly focal hyperhidrosis and facial blushing.1 The procedure (where the lower part of the left stellate ganglion and first 4–5 thoracic ganglia are ablated) also significantly reduces the occurrence and frequency of symptoms in long-QT syndrome (LQTS) and catecholaminergic polymorphic ventricular tachycardia (CPVT), even in high-risk populations.2–7
WHAT IS KNOWN

- Left cardiac sympathetic denervation reduces risk of cardiac events and sudden death in long QT syndrome and catecholaminergic polymorphic ventricular tachycardia
- Side effects have generally been reported as mild and uncommon but have not been reviewed systematically

WHAT THE STUDY ADDS

- Post-operative morbidity is common, most commonly due to dry skin on the left side of the body, a Harlequin-type (unilateral) facial flush with exercise, contralateral hyperhidrosis, differential hand temperatures, transient and permanent ptosis, thermoregulation difficulties, sensation of left arm paresis, and loss of sympathetic flight/fright response.
- Post-operative satisfaction is generally high, despite the side effects, and patients feel safer following the procedure. Most patients recommend that a similarly affected person have the procedure.

to counsel our patients to help make an informed choice, clinicians need to appreciate the experience of patients who have had the procedure previously for the same indications. To our knowledge, this is the first study to report the physical and psychological consequences of left cardiac sympathetic denervation (LCSD) and satisfaction among patients who had undergone video-assisted thoracoscopic LCSD for either LQTS or CPVT.

Methods

Study Population

Forty-seven patients who had undergone a minimally invasive, video-assisted LCSD for the treatment of LQTS or CPVT in New Zealand were included. All were enrolled in the New Zealand Cardiac Inherited Diseases Registry and consented to their data being used for research. Procedures were performed between 2008 and 2014 by 1 of 2 surgeons; median age at the time of LCSD was 17 years (range, 2–64 years); 34 patients were women (72%), and 13 were men. Patients who had the same procedure for other indications, or who had open surgery, were excluded. Pretreatment counseling was performed by the senior author and the surgeon performing the procedure.

An experienced clinician made a clinical diagnosis of LQTS or CPVT, and genotyping was subsequently attempted in all but 1 patient. Referral for LCSD was considered clinically appropriate.

Data Collection

Patient Information

Demographic and clinical data were obtained from medical records, most of which were stored prospectively as a part of the registry. Clinical diagnosis, genotype, mutation, most severe symptom before LCSD, medical therapy before and after the procedure, and indications for and details about procedure were recorded. Age at LCSD was recorded, and mean/median QTc is for LQTS patients only.

Physical and Psychological Consequences of LCSD

A single questionnaire was administered via telephone by the first author to patients aged >18 years (N=25) or to the parents if patients were younger at the time of the survey (N=18) (Data Supplement). One teenager responded to the questionnaire, but with parental consent. Questions were constructed to retrospectively assess baseline level of psychosocial stress, overall satisfaction, and physical and psychological sequelae from the procedure. Half of the questions provided scores out of 5. With regard to feelings of anxiety or depression, adult and pediatric patients and parents were asked to rate how often they felt anxious or depressed, where 1 = none of the time and 5 = all of the time. Answers were documented and were analyzed for common themes and notable side effects. Institutional ethical approval was obtained before survey.

ECG Analysis

Twelve-lead ECG analysis was performed by the first author blinded to patient identity, genotype, and clinical situation. The QT interval was measured from the beginning of the QRS complex to the end of the T wave (defined using the tangent technique, where the tangent of the steepest slope of the second limb of the T wave crosses the isoelectric line). Bazett correction was used, and the longest measurement of lead II or V5 was taken from ECGs taken the day before and after LCSD or as near to this time as possible.

Statistical Analyses

Assumptions of the t test were tested, and all data were analyzed by unpaired parametric and nonparametric tests as appropriate, including 2-way ANOVA, unpaired t test, and column statistics. Statistical analyses were performed using GraphPad Prism version 6.0e for Mac (GraphPad Software, La Jolla, CA; www.graphpad.com) and using SAS version 9.4 (Cary, NC).

Surgical Technique

Surgeons performed the procedure using video-assisted thoracoscopic surgical approaches with double-lumen endotracheal intubation and selective deflation of the left lung. Resection of the sympathetic chain was performed using minimally invasive techniques via either 1 or 3 axillary ports. The proximal extent of sympathectomy was either aggressive, which included sacrificing the lower third to lower half of the stellate ganglion (n=13; 28%) or conservative, which involved sparing the majority of the lower third of the stellate ganglion (n=34; 72%). Patients’ mean age was lower in the aggressive group than in the conservative group (16.5 versus 26 years; P=0.04). When comparing the aggressive and the conservative groups, there were no significant differences with reference to sex (with a preponderance of women in both groups; 8 [69%] versus 25 [74%]; P=0.3) or underlying disease (LQTS versus CPVT; P=1.0). Intraoperative intercostal drains were not used, and at the end of the procedure, air was evacuated. Patients were admitted to the intensive care unit postoperatively and discharged home after a median of 1 day (range, 0–13 days). One patient self-discharged on day 0, and the 13-day admission was because of complex management issues in a patient with Jervell and Lange-Nielsen syndrome, unrelated to LCSD. All other patients had 1 to 2 days of postoperative stay.

Results

Clinical Cohort

All patients had unequivocal phenotypic evidence of disease, 40 with LQTS and 7 with CPVT. Additional details of the study cohort are shown in Table 1 and Figure 1. The most common indications for LCSD related to medical therapy included inability to take β-blockers because of intolerance or contraindication such as asthma in 15 (32%) patients, and nonadherence to therapy in 10 (21%) patients.

Baseline Psychological Status

Self-reported retrospective scores of anxiety and depression were provided by adult patients, and by parents of the affected...
children for themselves and for their children if the patients were aged <18 years at the time of the survey.

**Adult Patients (>18 Years)**
Twenty-four of 25 adults answered the questions about preoperative anxiety and depression; median anxiety score was 2.5 (range, 1–4: ie, anxious none to most of the time, respectively), and baseline depression median score was 1 (range, 1–4). Seven adults (29%) reported feelings of anxiety related to LQTS/CPVT most of the time, and 6 (25%) reported feeling depressed most of the time.

**Pediatric Patients**
Nineteen of the 26 pediatric patients were still <18 years old at the time of the survey, and for this cohort, parents were asked about their own and their children’s baseline scores. However, 1 child was too young (4 years old) at the time of surgery for the parents to assess psychological status, 1 parent was unable...
Emotional and psychological sequelae

Now I get butterflies in my stomach instead of fast heart beats and faints/seizures. I feel more detached, and don’t feel

Difficulties in temperature regulation
One hot side and one cold side make it difficult in bed

Differential hand temperatures
Ice cold left hand

Compensatory hyperhidrosis
It’s really bizarre. I’m a freak, a smelly freak on the right hand side

Harlequin-type facial split
Jekyll and Hyde

Dry skin
Old lady skin on the left side

My left hand doesn’t crinkle, even in the pool

Embarrassing

It’s really bizarre. I’m a freak, a smelly freak on the right hand side

Embarrassing! I’d get the other side done, but then where would all the sweat go?

Ice cold left hand

One hot side and one cold side make it difficult in bed

Now I get butterflies in my stomach instead of fast heart beats and faints/seizures. I feel more detached, and don’t feel

I used to hate getting a fright, but now there is much less of a jolt, and I’m much calmer with frights

I don’t actually feel sad when I hear sad/bad news. I recognize that the situation is a sad one, so cognitively adjust my

behavior and response accordingly

I don’t get really anxious anymore, no more sudden adrenaline surges

I used to hate getting a fright, but now there is much less of a jolt, and I’m much calmer with frights

Bold-blocker intolerance

Beta-blocker non-adherence

Disease related factors

CPVT

LQT3

Symptoms on therapy

QTc=520ms

Figure 1. Major indication for each patient for left cardiac sympathetic denervation included β-blocker intolerance/contraindication, β-blocker nonadherence, disease-related factors, and patient choice. Disease-related factors include those with catecholaminergic polymorphic ventricular tachycardia (CPVT), long-QT syndrome 3 (LQT3), those who experienced symptoms while compliant with medical therapy, or those with a prolonged QTc (≥550 ms or >520 ms with another indication). Patient choice includes those patients who had family history of sudden cardiac death, wished to perform high-level sport, or others.

General Recovery

Majority of the patients (79%) were satisfied with their operation overall with regard to pain relief, side effects, physical and emotional recovery, and economic considerations (Table 4). There was no significant relationship between satisfaction and preoperative depression or anxiety (P=0.12 and P=0.08 respectively), length of postoperative follow-up (P=0.17), or severity of preoperative symptoms (P=0.61). Figure 2 and Table 4 show data on 44 patients who completed the survey, their side effects, and survey response scores.

Follow-up occurred over a median of 29 months postoperatively (range, 1 month to 5 years and 7 months). There was no perioperative mortality or major complications requiring surgical reintervention. One death occurred 47 months after LCSD, but this was noncardiac and unrelated to the procedure.

Cardiac Events

During 116 patient-years of follow-up, 1 patient with LQTS, 1 with Jervell and Lange-Nielsen LQTS, and 3 with CPVT experienced cardiac events (ie, 1+1/40 patients with LQTS, 5%; 3/7 patients with CPVT, 43%). This did not vary with the degree of sympathetic resection (LQTS, P=0.45; CPVT, P=0.43). The patient with single-mutation LQTS was 17 years old at LCSD and carried the KCNQ1 c.797T>C missense mutation, and her longest preoperative QTc was 522 ms. QTc before the procedure was 483 ms, and 431 ms after 2 years. She adhered to controlled-release metoprolol both pre- and postoperatively, and her worst symptom (in both time periods) was classic arrhythmic syncope, which resulted in implantable cardioverter defibrillator insertion after the postoperative event. Five of the 7 patients with CPVT had their procedure performed before flecainide was known to be therapeutic.13 Of the 3 patients with CPVT who experienced postoperative cardiac events, 1 refused medical therapy, 1 was on low-dose β-blocker without uptitration because of failure to attend follow-up appointments, and the third was on both nadolol and flecainide at the time of her arrest. There was no relationship between degree of sympathetic resection and occurrence of postoperative symptoms.

Postoperative Course

Follow-up by telephone survey and review of case notes were performed after a median follow-up of 29 months after LCSD (range, 1–67 months). Tables 2 and 3 show patient comments on side effects.

Table 2. Patient and Caregiver Comments Describing Side Effects Occurring After Left Cardiac Sympathetic Denervation for Long-QT Syndrome or Catecholaminergic Polymorphic Ventricular Tachycardia

<table>
<thead>
<tr>
<th>Side Effect</th>
<th>Patient Comments</th>
</tr>
</thead>
<tbody>
<tr>
<td>Dry skin</td>
<td>Old lady skin on the left side</td>
</tr>
<tr>
<td></td>
<td>My left hand doesn’t crinkle, even in the pool</td>
</tr>
<tr>
<td>Harlequin-type facial split</td>
<td>Jekyll and Hyde</td>
</tr>
<tr>
<td>Compensatory hyperhidrosis</td>
<td>It’s really bizarre. I’m a freak, a smelly freak on the right hand side</td>
</tr>
<tr>
<td>Differential hand temperatures</td>
<td>Ice cold left hand</td>
</tr>
<tr>
<td>Difficulties in temperature regulation</td>
<td>One hot side and one cold side make it difficult in bed</td>
</tr>
<tr>
<td>Emotional and psychological sequelae</td>
<td>Now I get butterflies in my stomach instead of fast heart beats and faints/seizures. I feel more detached, and don’t feel embarrassed, sad, angry, or disappointed anymore. Not getting angry is a bonus with a 15-year-old daughter. At times I know that I’m angry, but I don’t have a fright/fight response, I have no starblt response, and have a dull thud feeling instead. I don’t actually feel sad when I hear sad/bad news. I recognize that the situation is a sad one, so cognitively adjust my behavior and response accordingly</td>
</tr>
<tr>
<td></td>
<td>I don’t get really anxious anymore, no more sudden adrenaline surges</td>
</tr>
<tr>
<td></td>
<td>I used to hate getting a fright, but now there is much less of a jolt, and I’m much calmer with frights</td>
</tr>
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Eleven patients (25%) reported significant differences in the temperature of their hands; however, they were not all able to state which hand was warm or cold.

Electrocardiographic Changes
Median QTc among patients with LQTS was 461±60 ms preoperatively and was 476±54 ms postoperatively (P=0.49).

Physical Sequelae

Dry Skin
The most common effect reported from the procedure was dry skin on the left side of the body (67%). Twenty-nine patients reported a dry left hand, including 3 who also reported a dry face (on left side), and 3 reported dry left foot. Seven patients used extra (or stronger) moisturizer on the left side.

Harlequin-Type Facial Flush
The second most common effect was having a marked Harlequin-type demarcation in color on, at least, the face (63%): 17 reported this on the face only, 3 on the hand only, and 5 on the whole body, and 8 patients reported worsening of this effect after exertion (Figure 3A).

Compensatory Hyperhidrosis
Fifty-five percent of patients reported problems with excessive sweating on the right side (Figure 3B). Lifestyle adjustments included using heavy-duty antiperspirant (including the lumbar back), carrying a towel always, and using grip aids to play sport (with the dry left hand losing grip). One patient used grip aids because of sweaty right hand, one slipped because of dry left hand, and a third used sticky stuff on the hands while playing netball (parent unsure of side).

Pain
Five patients reported severe pain in hospital, necessitating extra analgesia (11%). Three patients (7%) experienced a shooting type of pain down the left side, which resolved after 1 to 8 months. No patients had chronic pain, and 1 patient reported resolution of pre-existing chronic upper back pain.

Hand Temperature
Eleven patients (25%) reported significant differences in the temperature of their hands; however, they were not all able to state which hand was warm or cold.

Table 3. Patient and Caregiver Comments Describing Satisfaction Having Had Left Cardiac Sympathetic Denervation for Long-QT Syndrome or Catecholaminergic Polymorphic Ventricular Tachycardia

<table>
<thead>
<tr>
<th>Comment</th>
<th>Percentage</th>
</tr>
</thead>
<tbody>
<tr>
<td>I see sympathectomy as a passage back to normal life</td>
<td>26 (59)</td>
</tr>
<tr>
<td>Peace of mind for parents. Do it the “sooner the better.” Extra insurance</td>
<td>20 (45)</td>
</tr>
<tr>
<td>Made my life better. I wish it was done earlier. Ends suffering</td>
<td>17 (38)</td>
</tr>
<tr>
<td>Reduces risk of sudden death, its lifesaving, and the benefits far outweigh the side effects</td>
<td>16 (35)</td>
</tr>
<tr>
<td>Might as well have it done, because no difference afterward (back to normal). I’m safe; it’s an extra thing to be safe</td>
<td>14 (31)</td>
</tr>
<tr>
<td>Eliminates worry, and if something happens in the future, I don’t have to think “what if?”</td>
<td>13 (29)</td>
</tr>
<tr>
<td>Helps keep you alive, wouldn’t want to be left wondering</td>
<td>11 (24)</td>
</tr>
<tr>
<td>It’s given me confidence; I’ve done something to make me safer</td>
<td>10 (22)</td>
</tr>
</tbody>
</table>

Thermoregulation Difficulties
Five patients (11%) reported a definite hot and cold side of the body (right:left, 50:50). They commented that it was difficult to regulate their body temperature, particularly in bed or in cold weather.

Ptosis
Nine patients reported ptosis on the left side. In 4 individuals, this was transient lasting between 3 days and ≈6 months. Five patients (11%) had permanent ptosis (at median follow-up of 26 months; range, 9–55 months), all reported as very mild, and none reported disappointment (Figure 4). One patient reported that the ptosis worsened with fatigue.

Paresthesia
Two women (5%) reported a sensation of reduced feeling and tingling in the left fingers and arm up to the elbow.

Emotional/Psychological Sequelae

Loss of Sympathetic Flight/Fright Response
Three women (7%) reported that they were much calmer in situations that previously would have been alarming or frightening. One also felt detached in sad or angry circumstances.

Satisfaction
The majority of patients were satisfied postoperatively and they felt positive, felt safer, and were happy to recommend the procedure to others (Figure 2; Table 3 for patient comments).

Table 4. Postoperative Physical and Psychological Consequences of LCSD, Postoperative Satisfaction

<table>
<thead>
<tr>
<th>Consequence of LCSD</th>
<th>N (%), Postoperative Physical and Psychological Questions</th>
</tr>
</thead>
<tbody>
<tr>
<td>Dry skin</td>
<td>29 (67)</td>
</tr>
<tr>
<td>Harlequin-type flush</td>
<td>27 (63)</td>
</tr>
<tr>
<td>Hyperhidrosis</td>
<td>24 (56)</td>
</tr>
<tr>
<td>Pain</td>
<td>8 (19)</td>
</tr>
<tr>
<td>Difference in hand temperature</td>
<td>11 (26)</td>
</tr>
<tr>
<td>Thermoregulation difficulties</td>
<td>5 (12)</td>
</tr>
<tr>
<td>Ptosis (permanent plus transient)</td>
<td>9 (21)</td>
</tr>
<tr>
<td>Ptosis (permanent)</td>
<td>5 (12)</td>
</tr>
<tr>
<td>Ptosis (transient)</td>
<td>4 (9)</td>
</tr>
<tr>
<td>Paresthesia</td>
<td>2 (5)</td>
</tr>
<tr>
<td>Loss of sympathetic flight/fright response</td>
<td>3 (7)</td>
</tr>
<tr>
<td>Postoperative satisfaction</td>
<td>34/43 (79)</td>
</tr>
<tr>
<td>Feel happy with their surgical scar appearance (score 1, 2, or 3 of 5)</td>
<td>41/44 (93)</td>
</tr>
<tr>
<td>Feel positive after procedure (positive vs negative)</td>
<td>35/41 (85)</td>
</tr>
<tr>
<td>Feel safer after procedure (score 1 or 2 of 5; the remainder felt the same as preoperatively, score 3/5)</td>
<td>33/44 (75)</td>
</tr>
<tr>
<td>Feel happy the procedure happened (score 1 or 2 of 5)</td>
<td>38/44 (86)</td>
</tr>
<tr>
<td>Feel no regret that the procedure happened (score 4 or 5 of 5)</td>
<td>40/44 (91)</td>
</tr>
<tr>
<td>Would recommend the procedure to someone like you/your child (yes vs no, unsure)</td>
<td>40/44 (91)</td>
</tr>
</tbody>
</table>

LCSD indicates left cardiac sympathetic denervation.
Symptom Duration
Three patients reported that their symptoms were worst in the first year (or second), and then improved.

Comparison of Side Effects According to Degree of Resection
When comparing the aggressive versus conservative resection groups, there were no significant differences in dry skin ($P=0.14$), Harlequin-type facial flush ($P=0.72$), compensatory hyperhidrosis ($P=1.0$), hand temperature ($P=0.47$), pain ($P=0.32$), thermoregulation difficulties ($P=1.0$), paresthesia ($P=0.56$), or loss of sympathetic drive ($P=0.55$). When both transient and permanent ptosis were included, then there was a significant difference between aggressive ($n=5/10$) and conservative ($n=3/31$; $P=0.009$) groups.

Discussion
The left cardiac sympathectomy, first described in 1971, has recently re-emerged as an important therapeutic option for both LQTS and CPVT and can now be performed thoracoscopically. The procedure is both safe and efficacious. For some patients, it is first-line therapy when β-blockers are contraindicated or cannot be tolerated. In our New Zealand population, poor long-term adherence to therapy, a major hazard in those severely affected, remains a significant problem. The fact that 3 of our patients who underwent LCSD for nonadherence can no longer be traced underscores the potential value of this procedure in patients who only intermittently engage with health services. Nevertheless, this is a procedure with side effects, most of which are permanent, so accurate counseling before the procedure is essential.

The present study documents high rates of morbidity related to side effects, sometimes associated with significant levels of embarrassment and distress. The side effects described by our cohort are consistent with those when the sympathectomy is done for other reasons, and may be more severe because of the ablation of the second thoracic ganglion, which is associated with more pronounced compensatory hyperhidrosis. However, most other indications, such as hyperhidrosis, involve bilateral sympathectomy, so the harlequin effect does not occur. Furthermore, patients with LQTS and CPVT tend to have underlying anxiety and sometimes depression related indirectly to their condition and also to the sacrifices and lifestyle changes they must make. They often also have a traumatic personal or family history. This background, confirmed by the baseline psychological profile in this study, may explain the overall satisfaction among patients with LQTS and CPVT who have undergone LCSD. The sympathectomy gives them a sense of safety and a sense that they can lead a relatively normal life, which is evident from a patient’s comment: “I see sympathectomy as a passage back to normal life.”

Before this investigation, we and others have counseled our patients that permanent ptosis was the most significant (but uncommon) side effect. However, in this study, patients often recalled their side effects with some distress. They described embarrassment, inconvenience, and annoyance. We are now able to give a more thorough and honest account of life after an LCSD. The findings of this review have resulted in a change of emphasis in our counseling practice; we highlight the compensatory hyperhidrosis, Harlequin-type facial flush, and the less common and very mild ptosis. Advising the patients about overwhelming patient satisfaction is as important as warning them of the side effects. Regarding the occurrence of Horner syndrome, we found no difference between aggressive and conservative surgical technique, but given the low prevalence, a larger series may be needed to clarify this.

This study adds a new perspective to the side effects of sympathectomy. Studies on LQTS and CPVT populations suggest that the side effects of LCSD are mild and limited. Previous reports of side effects from the larger cohorts where sympathectomy is performed for hyperhidrosis or facial flushing were based on physician-only assessments; patient commentary was excluded and may anyway be different since the procedure was largely for cosmetic reasons. The majority...
of patients in our cohort would strongly disagree that side
effects are minimal, but agree that they are outweighed by the
benefits.21

This study was not designed to assess the efficacy of
reducing cardiac events; nevertheless, there are still impor-
tant findings. In case reports and small series, LCSD has
been reported to have significantly reduced the occurrence
of cardiac events in patients with CPVT.2,7,22–27 In the larg-
est and most recent series published, 54 symptomatic patients
with CPVT underwent LCSD. Although the number of
patients who had an incomplete LCSD was small, the authors
reported that those who had a complete LCSD were much
less likely to have postoperative cardiac events compared
with those who had an incomplete denervation (8/47 [17%]
versus 5/7 [71%]; P<0.01).27 Six patients (86%) were preop-
eratively symptomatic, and 3 of 7 (43%) were postoperatively
symptomatic (postoperative median follow-up of 45 months;
range, 6–67 months; 24 patient-years). Therefore, it should
be emphasized that long-term adherence to medical therapy,
importantly flecainide therapy, is paramount in the care of
individuals with CPVT.

A limitation of this study is the reliance on patient report-
ing and lack of objective measures. This may result in a higher
incidence of reported side effects compared with other series.
Nonconfidential responses may introduce bias, but given
the high morbidity reported, we feel that the impact of this
would be minimal. Furthermore, no validated questionnaire
was suitable for this study cohort. Although there are inher-
ent challenges in interviewing children and there may be a
discordance between responses of parents and children, we
performed an ordinal logistic regression excluding parental
data to minimize this issue. Follow-up is variable and <1 year
in 23% of the cohort, a time when side effects are more pro-
nounced (both in our series and in others).8

Conclusions
This study documents, for the first time, side effects and
patient satisfaction associated with minimally invasive video-
assisted thoracoscopic LCSD in LQTS and CPVT populations.
Although morbidity was high (most commonly because of dry
skin, compensatory hyperhidrosis, and Harlequin-type facial
flush), so too was patient satisfaction, with 91% of patients
recommending the procedure to other affected patients. Extent
of surgical resection influenced risk of ptosis, but not of other
outcomes. This study enables us to counsel patients better
before they undergo this treatment.

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Disclosures

None.

References


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CIDG (the Cardiac Inherited Disease Group)

CIDG (the Cardiac Inherited Disease Group) is a national network of clinicians and scientists in New Zealand. In particular regarding the present work we would like to acknowledge the following clinicians for contributing patients to the study and assisting with referral planning.

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Hamilton
# Questionnaire

**TO BE COMPLETED PRIOR TO TELEPHONE CONTACT**

<table>
<thead>
<tr>
<th>PATIENT DETAILS</th>
<th>STICKY LABEL</th>
</tr>
</thead>
<tbody>
<tr>
<td>Treating cardiologist</td>
<td></td>
</tr>
<tr>
<td>Date of procedure</td>
<td></td>
</tr>
<tr>
<td>Surgeon / Anaesthetist</td>
<td></td>
</tr>
</tbody>
</table>
| Surgical technique  
- number of ports  
- patient position |               |
| Anaesthetic/Intubating technique  
- CO2 insufflation  
- apnoea |               |
| Age, height, weight at op, ethnicity |               |
| Date of follow up: clinical |               |
| Date of follow up: phone |               |
| Medications pre op |               |
| Medications post op |               |
| Other medical problems/PMH |           |
| Type of LQTS/  
 gene result |               |
| Significant LQTS history (ICD/arrest etc) |            |
| Indication: Primary/Secondary |             |
| Family history |               |
| Pre-operative QT and QTc |             |
| Pre-operative T wave morphology |            |
| Post-operative QT & QTc in hospital |          |
| Post-operative QT & QTc follow up (date) |           |
| T wave morphology post operative |            |

**COLLECT AT TIME OF TELEPHONE CONTACT**

<table>
<thead>
<tr>
<th>Name of person completing questionnaire</th>
<th></th>
</tr>
</thead>
<tbody>
<tr>
<td>Name of person completing questionnaire</td>
<td></td>
</tr>
<tr>
<td>----------------------------------------</td>
<td>--</td>
</tr>
<tr>
<td>Age/Sex/Clinically affected</td>
<td></td>
</tr>
<tr>
<td>Relationship to person having LCSD</td>
<td></td>
</tr>
</tbody>
</table>

<table>
<thead>
<tr>
<th>How did you (adult) feel BEFORE the procedure:</th>
<th>1= none of the time, 2= a little of the time, 3 = some of the time, 4 = most of the time, 5= all of the time</th>
</tr>
</thead>
<tbody>
<tr>
<td>Not anxious 1 2 3 4 5 Anxious all the time</td>
<td></td>
</tr>
<tr>
<td>Not sad/Depressed 1 2 3 4 5 Sad/depressed all the time</td>
<td></td>
</tr>
</tbody>
</table>

<table>
<thead>
<tr>
<th>How did your child feel BEFORE the procedure:</th>
<th>1= none of the time, 2= a little of the time, 3 = some of the time, 4 = most of the time, 5= all of the time</th>
</tr>
</thead>
<tbody>
<tr>
<td>Not scared 1 2 3 4 5 Scared all the time</td>
<td></td>
</tr>
<tr>
<td>Not flat/down 1 2 3 4 5 Flat/down all the time</td>
<td></td>
</tr>
</tbody>
</table>

<table>
<thead>
<tr>
<th>Was it a difficult decision to have the procedure?</th>
<th>Not hard 1 2 3 4 5 Hard</th>
</tr>
</thead>
<tbody>
<tr>
<td>Why?</td>
<td>Risks and worry about side effects of sympathectomy?</td>
</tr>
<tr>
<td></td>
<td>Worry about operation/anaesthetic</td>
</tr>
<tr>
<td></td>
<td>Worry about what your child would think later in life?</td>
</tr>
<tr>
<td>If prompting needed, eg:</td>
<td>Other.......</td>
</tr>
</tbody>
</table>

<table>
<thead>
<tr>
<th>Why did your child have the procedure?</th>
<th>Because they/I wanted it</th>
</tr>
</thead>
<tbody>
<tr>
<td></td>
<td>Because the doctor recommended it</td>
</tr>
<tr>
<td></td>
<td>Couldn’t take beta blockers</td>
</tr>
<tr>
<td></td>
<td>Needed more protection</td>
</tr>
<tr>
<td></td>
<td>Other.......</td>
</tr>
</tbody>
</table>

<table>
<thead>
<tr>
<th>Any effects from the procedure?</th>
<th>Overall recovery: Considering pain relief, side effects, physical and emotional recovery and economic considerations, how satisfied are you with the operation? Not satisfied 1 2 3 4 5 Very satisfied</th>
</tr>
</thead>
<tbody>
<tr>
<td>Scars:</td>
<td>Very unhappy 1 2 3 4 5 Very happy</td>
</tr>
<tr>
<td>Droopy eye lid:</td>
<td>Immediate post op Yes / No</td>
</tr>
<tr>
<td></td>
<td>Now: Yes / No</td>
</tr>
<tr>
<td></td>
<td>How long before it resolved?.................</td>
</tr>
<tr>
<td>Sweating:</td>
<td>Left hand: Dry / Normal</td>
</tr>
<tr>
<td></td>
<td>Left arm pit: Dry / Normal</td>
</tr>
<tr>
<td></td>
<td>Right hand.........................</td>
</tr>
<tr>
<td></td>
<td>Excessive sweating post op....................</td>
</tr>
<tr>
<td></td>
<td>Change in colour................................</td>
</tr>
<tr>
<td>Any others..........................</td>
<td>.............................................</td>
</tr>
<tr>
<td>Question</td>
<td>Response Options</td>
</tr>
<tr>
<td>-------------------------------------------------------------------------</td>
<td>-------------------------------------------------------</td>
</tr>
</tbody>
</table>
| Are any effects negative? From a mood or body image or physical perspective? | Mood is different: Embarrassed/Sad/Angry/Disappointed  
Feel different about body image:  
Feel different physically:  
Other |
| How do you/your child feel overall after the procedure?                  | Positive/Negative                                     
Safer 1 2 3 4 5 Less safe  
Happy it happened 1 2 3 4 5 Not happy it happened  
Regret it happened 1 2 3 4 5 No regret  
Any faints or cardiac arrest since sympathectomy?  
Other |
| Having gone through the sympathectomy, would you recommend someone like you/your child have the procedure? | Yes/No |
| Why?                                                                    |                                                      |
| Could we do anything to make it easier for you and your child?           |                                                      |
| Do you have any concerns or issues regarding your LCSD or LQTS Tx you would like us to contact your treating cardiologist about? |                                                      |