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LEFT THORACOSCOPIC SYMPATHECTOMY FOR CARDIAC DENERVATION IN CHILDREN WITH LIFE-THREATENING VENTRICULAR ARRHYTHMIAS

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Disclosures

- **None of the authors in this study have any commercial relationships to disclose**



Inherited Ventricular Arrhythmias

- **10-20% of all sudden cardiac deaths occur in the absence of structural heart disease**
- **Inherited ion channel disorders/channelopathies**
 - **Long QT Syndrome (LQTS)**
 - Highly susceptible to ventricular arrhythmias
 - Estimated risk of SCD 1-2% per year
 - **CPVT (Catecholaminergic polymorphic ventricular tachycardia)**
 - Exercise/emotion-induced bidirectional ventricular tachycardia
 - 30% experience ≥ 1 cardiac arrest and up to 80% ≥ 1 syncopal spells
 - **Idiopathic recalcitrant VT, Brugada Syndrome**

Existing Treatment Options

- **Beta-blockers ± other antiarrhythmic**
 - 32% have further cardiac events within 5 years¹
 - 14% who survived a SCD event will have a recurrence within 5 years¹
 - Medication intolerance, poor compliance
- **Implantable Cardioverter Defibrillators (ICDs)**
 - Device malfunction²
 - Inappropriate ICD discharges/electrical storms²
 - Reduced quality of life

1. Moss et al Circ 2000; 2. Tung et al, J Am Coll Cardiol 2008.

Rationale for Left Cardiac Sympathetic Denervation (LCSD)

- **Sympathetic nervous system plays “triggering role”¹**
- **Antiarrhythmic effects**
 - Reduced release of norepinephrine at the ventricular level¹
 - Raises fibrillation threshold, increases refractoriness
 - Highly arrhythmogenic potential of left-sided cardiac sympathetic nerves²
- **Permanent**

1. Schwartz PJ et al. Am J Cardiol 1976; 2. Schwartz PJ Circ Res 1977.

Background: LCSD Experience

Clinical Investigation and Reports

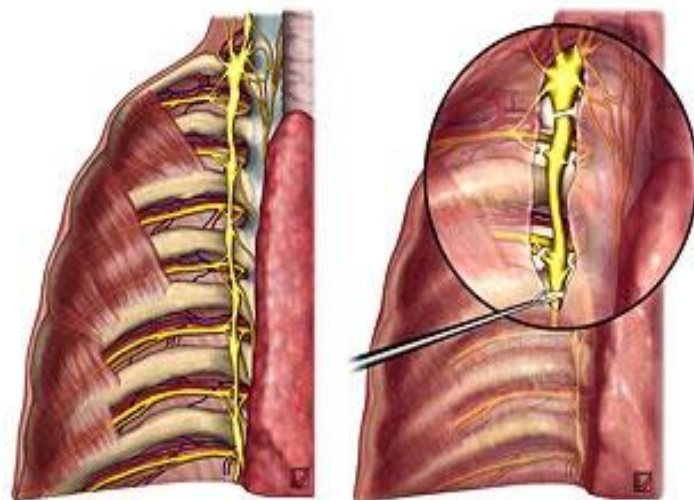
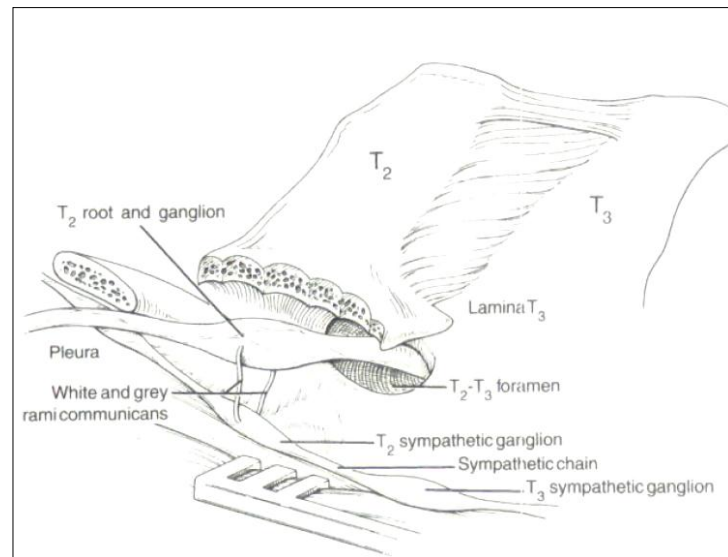
Left Cardiac Sympathetic Denervation in the Management of High-Risk Patients Affected by the Long-QT Syndrome

Peter J. Schwartz, MD; Silvia G. Priori, MD, PhD; Marina Cerrone, MD; Carla Spazzolini, PhD; Attilio Odero, MD; Carlo Napolitano, MD, PhD; Raffaella Bloise, MD; Gaetano M. De Ferrari, MD; Catherine Klersy, MD, MS; Arthur J. Moss, MD; Wojciech Zareba, MD; Jennifer L. Robinson, MS; W. Jackson Hall, PhD; Paul A. Brink, MD; Lauri Toivonen, MD; Andrew E. Epstein, MD; Cuilan Li, MD; Dayi Hu, MD[†] (*Circulation*. 2004;109:1826-1833.)

- **147 patients with refractive Long QT Syndrome**
- **Cardiac sympathectomy significantly reduced the mean annual rate for any cardiac event and for SCD or aborted cardiac arrest**

History of Technique

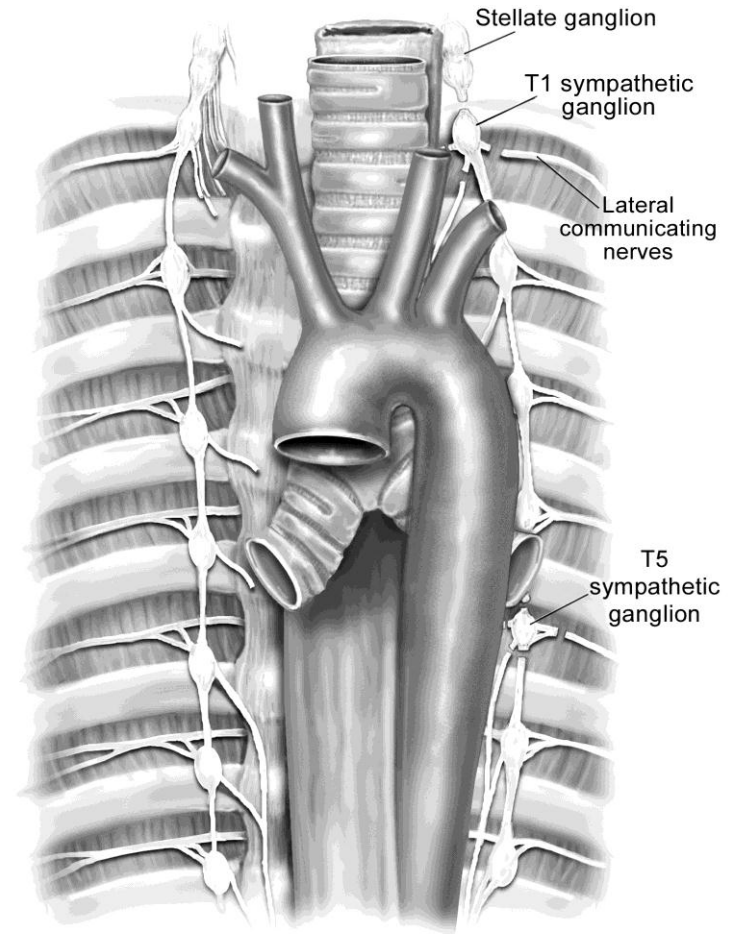
- **Standard technique: anterior supraclavicular and/or posterior thoracotomy incision**
- **Resection of all or part of the left stellate ganglion and most of the upper thoracic sympathetic ganglia**
- **Associated with development of Horner's syndrome ¹**



Schneider et al. Clin Res Cardiol 2013
Coleman et al. Circ 2012

VATS – High Thoracic Sympathectomy

- Left lung isolation
- Right lateral decubitus position
- Standard 3-port technique
- Transection of left sympathetic chain to base of left stellate ganglion at the level of T1 and T5.
- Transection of lateral communicating nerves of Kuntz between T1 and T5
- No chest drains left in situ



Methods

- **24 patients underwent VATS-LCSD from 2000-2011**
 - 13 congenital LQTS (2 Jervell and Lange-Nielson syndrome)
 - 9 CPVT
 - 2 idiopathic recalcitrant VT
- **Study Endpoints**
 - Baseline patient characteristics
 - Indications for treatment
 - Persistent symptoms (Multiple ICD shocks/arrhythmic events despite beta-blockade)
 - Failure to tolerate beta-blockers
 - Primary prevention
 - Procedural events
 - Response to therapy

Patient Characteristics

	Long QT Syndrome	CPVT	Idiopathic VT
Number	13	9	2
Median age, yrs (range)	8 (2-22)	17 (8-27)	1 (5 wks-2 yrs)
Sex (M/F)	6/9	5/4	1/1
Initial Presentation			
Syncope with VT	6	4	1
Aborted SCD	5	1	1
Recurrent VT/TdP	2	4	-
Previous Intervention			
ICD	5	7	-
ICD + Pacemaker	1	1	-
Other	-	-	1 (ablation)



Indications for VATS-LCSD

- **Persistent Symptoms (n=18)**
 - LQTS = 8
 - CPVT = 8
 - Idiopathic VT = 2
- **Failure to tolerate medical therapy (n=4)**
 - LQTS = 3
 - CPVT = 1
- **Primary Prevention (n=2)**
 - LQTS = 2



Surgical Outcomes

- **Concomitant procedures**
 - ICD implantation: 8 patients
 - Dual chamber pacemaker: 1 patient
- **No intraoperative complications**
- **3 minor post-operative complications**
 - 1 patient with prominent “harlequin” facial flushing (stellate ganglion)
 - 2 patients with small apical pneumothorax (did not require chest tube)
- **Median length hospital stay = 2 days**



Outcomes

- **Median F/U: 28 months (range, 4-131 months)**
- **Response based on LCSD Indication**
 - **Persistent symptoms (n=18, 2 lost to follow-up)**
 - Reduced arrhythmia burden: 3/16
 - Arrhythmia free: 10/16
 - Response rate: 13/16 (81%)
 - **Failed Medical Therapy (n=4)**
 - Reduced arrhythmia burden: 1/4
 - Arrhythmia free: 2/4
 - Response rate: 3/4 (75%)
 - **Prophylactic Therapy (n=2)**
 - Patient 13: 3 ICD shocks, 1 antiarrhythmic agent at 82 months
 - Patient 18: Multiple runs TdP, increased B-blocker at 26 months

Overall Response

- **16 (73%) Responders**
 - LQTS = 9
 - CPVT = 6
 - IVT = 1
- **6 (27%) Non-responders**
 - LQTS = 4
 - CPVT = 2

Limitations

- **Retrospective Study**
- **Small patient numbers**
- **Incomplete follow-up**
- **Lack of quantitative marker of arrhythmia burden**



Conclusions

- **VATS-LCSD is a safe and effective adjunctive therapy in most children with life-threatening ventricular arrhythmias**
- **Main indication is in patients with persistent symptoms despite optimal medical therapy/frequent ICD shocks and in those who fail to tolerate beta-blockers**
- **Effectiveness as prophylactic therapy remains equivocal**
- **VATS approach is minimally invasive, low-risk intervention**
 - **Treatment of asymptomatic patients based on genotypic characteristics**
 - **Subset of non-responders patients may benefit from bilateral sympathectomy**