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

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AATS 2013
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 years\(^1\)
  – 14% who survived a SCD event will have a recurrence within 5 years\(^1\)
  – Medication intolerance, poor compliance

• Implantable Cardioverter Defibrillators (ICDs)
  – Device malfunction\(^2\)
  – Inappropriate ICD discharges/electrical storms\(^2\)
  – Reduced quality of life

Rationale for Left Cardiac Sympathetic Denervation (LCSD)

- Sympathetic nervous system plays “triggering role” 1

- Antiarrhythmic effects
  - Reduced release of norepinephrine at the ventricular level 1
  - Raises fibrillation threshold, increases refractoriness
  - Highly arrhythmogenic potential of left-sided cardiac sympathetic nerves 2

- Permanent

Background: LCSD Experience

- 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

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

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