

Left cardiac sympathetic denervation for recurrent ventricular tachyarrhythmias in children with congenital heart disease



Erica D. Bonura, MD,^{*} Christopher Moir, MD,^{*†} Michael J. Ackerman, MD, PhD,^{*‡§} Philip Wackel, MD^{*}

From the ^{*}Division of Pediatric Cardiology, Department of Pediatric and Adolescent Medicine, Mayo Clinic, Rochester, Minnesota, [†]Division of Pediatric Surgery, Department of Surgery, Mayo Clinic, Rochester, Minnesota, [‡]Division of Heart Rhythm Services, Department of Cardiovascular Medicine, Mayo Clinic, Rochester, Minnesota, and [§]Windland Smith Rice Sudden Death Genomics Laboratory, Department of Molecular Pharmacology & Experimental Therapeutics, Mayo Clinic, Rochester, Minnesota.

Introduction

The first sympathetic denervation was described in 1961 as a treatment for ventricular tachycardia (VT).¹ Following this, left cardiac sympathetic denervation (LCSD) was first applied as a treatment for long QT syndrome (LQTS) in humans in 1971.² Currently, LCSD is recognized as an effective treatment for LQTS and catecholaminergic polymorphic VT (CPVT).^{3,4} However, there are increasing data that bilateral denervation surgery or LCSD may be useful in non-channelopathy-related ventricular arrhythmias.^{5,6} Although denervation therapy in adult congenital heart disease (CHD) has been reported previously, there are no reports on utilizing LCSD in pediatric patients with CHD and refractory ventricular tachyarrhythmias after surgical repair.⁷ We present a series of 2 patients with surgically corrected CHD and refractory arrhythmias who experienced resolution of their arrhythmias following videoscopic LCSD.

Case report

Case 1

A now 13-year-old male patient was born with D-transposition of the great arteries, atrial septal defect, ventricular septal defect, and patent ductus arteriosus. On day of life 4, he underwent an arterial switch procedure with a LeCompte maneuver, ventricular septal defect and atrial septal defect

suture closure, and patent ductus arteriosus ligation. He was noted to have congenitally small but patent left anterior descending and diminutive circumflex coronary arteries with a right-dominant coronary artery system. Postoperatively, he had depressed left ventricular (LV) function (ejection fraction 40%) and complete atrioventricular block, for which a single-chamber epicardial pacemaker was placed.

At age 7, his epicardial lead failed and his pacemaker was revised to a transvenous single-chamber pacemaker. At age 11, he had his first episode of monomorphic VT and was upgraded to an epicardial implantable cardioverter-defibrillator (ICD). He trialed multiple antiarrhythmic combinations but had increasingly frequent episodes of VT requiring ICD therapies. Catheter ablation was performed, but under anesthesia VT was not inducible and therefore only substrate modification was done.

At the age of 13, he presented with VT storm, having more than 60 episodes of VT in 24 hours, most of which were terminated with antitachycardia pacing. During a second ablation attempt, there was again suppression of his VT with anesthesia administration. Substrate modification was again attempted, but his incessant VT returned after he emerged from anesthesia. Amiodarone combined with beta-blockers were initiated but only slowed the VT rate. Concurrently, he experienced significant anxiety and posttraumatic stress disorder, and intravenous anxiolytics would temporarily suppress the VT.

Because his VT appeared catecholamine sensitive, he underwent LCSD as a last effort before listing for cardiac transplant. Performing the LCSD as a minimally invasive thoroscopic procedure was successful, although it was more technically difficult because of the prior cardiac surgeries with significant adhesions (Figure 1) and the presence of an ICD lead coursing through the pleural space. The patient was placed in a right lateral decubitus position and via 2 small incisions to accommodate a 3 mm trocar, the sympathetic chain was thoroscopically dissected and visualized.

KEYWORDS Congenital heart disease; Pediatric; Sympathetic denervation; Ventricular fibrillation; Ventricular tachycardia (Heart Rhythm Case Reports 2019;5:392–394)

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KEY TEACHING POINTS

- Recurrent ventricular tachyarrhythmias can be a challenge for pediatric patients with surgically corrected congenital heart disease and can lead to significant morbidity and mortality. This case report demonstrates that left cardiac sympathetic denervation (CSD) may be helpful in effectively controlling these arrhythmias.
- Left CSD is typically performed via a thoracoscopic approach that can be more challenging in patients who have undergone prior thoracic procedures. Despite these challenges, left CSD was able to be performed without complications in the patients within this case report.
- This case report demonstrates a novel application of left CSD, a well-known surgical procedure, which may allow patients to avoid or delay heart transplantation.

Removal of the sympathetic chain beginning below the ganglion at T4 and extending up to the midpoint of the stellate ganglion was confirmed with histopathology. Postoperatively, he required vasopressor support for less than 24 hours. There were no additional complications. After the LCSD, he was taken off amiodarone and has continued with his current medication regimen of carvedilol, lisinopril, and digoxin. He has had no recurrence of his VT and no ICD therapies in more than 2 years post-LCSD. There have been no immediate or short-term side effects during follow-up.

Case 2

A now 8-year-old male patient was born with tetralogy of Fallot and significant LV noncompaction with preserved LV systolic function (Figure 2). He underwent surgical repair of his tetralogy of Fallot at 3 months of age. At age 3, he had his first episode of ventricular fibrillation (VF) arrest, which prompted placement of an epicardial ICD. Genetic testing was performed and revealed no pathogenic mutations. After trials of multiple combinations of antiarrhythmic medications, he continued to have recurrent VF, including a VF arrest requiring cardiopulmonary resuscitation and defibrillation. By age 6 he had undergone multiple revisions of his epicardial ICD system secondary to infections and lead malfunction.

After less than 1 year on amiodarone, in addition to lisinopril and carvedilol to treat his underlying cardiomyopathy, he once again began having recurrent VF. At the age of 7, LCSD (as outlined in the previous case) was performed as a last attempted therapy for recurrent VF before he was to be listed for transplant. The procedure was successfully performed with a minimally invasive thoracoscopic approach, with the patient in the right lateral decubitus position, via 3 small

incisions to accommodate a 3 mm trocar. The sympathetic chain was thoracoscopically removed beginning below the ganglion at T4 and extending up to the midpoint of the stellate ganglion and was confirmed with histopathology. Following the LCSD, he has had no more ventricular arrhythmias, has been taken off of amiodarone, and has had a negative ventricular stimulation test. He has now been arrhythmia free with no ICD therapies while off amiodarone for more than 1 year. There have been no immediate or short-term side effects noted during follow-up.

Discussion

Ventricular tachyarrhythmias are an uncommon but well-recognized life-threatening complication following surgical repair of CHD. Traditional management often includes a combination of antiarrhythmics, ICD implantation, and catheter ablation. Although LCSD has been utilized extensively in patients with either LQTS or CPVT,^{4,5} it has been reported to be beneficial in some patients with various types of cardiomyopathy. However, surgically repaired pediatric CHD has not typically been thought of as a targeted substrate for LCSD. Both of our patients had undergone maximal therapy for incessant ventricular tachyarrhythmias and were being considered for cardiac transplantation before LCSD resulted in complete resolution of the arrhythmia.

It is clear that autonomic cardiac regulation plays an important role in ventricular arrhythmias via a highly complex network of interactions. Complete review of the interactions of the autonomic nervous system with the heart and the effects of manipulating it are beyond the scope of this paper, but Shivkumar and colleagues⁸ summarize this nicely and explain that LCSD reduces efferent and alters afferent signaling to and from the heart, thereby reducing localized

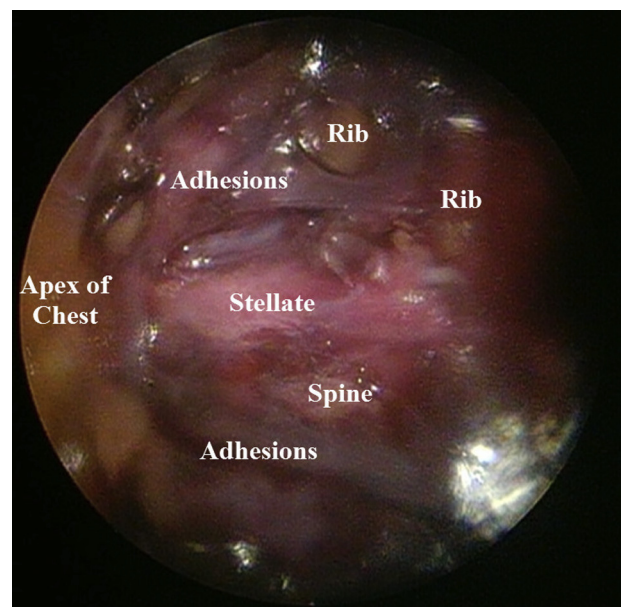


Figure 1 Thoracoscopic view of dense adhesions overlying the left thoracic stellate ganglion.

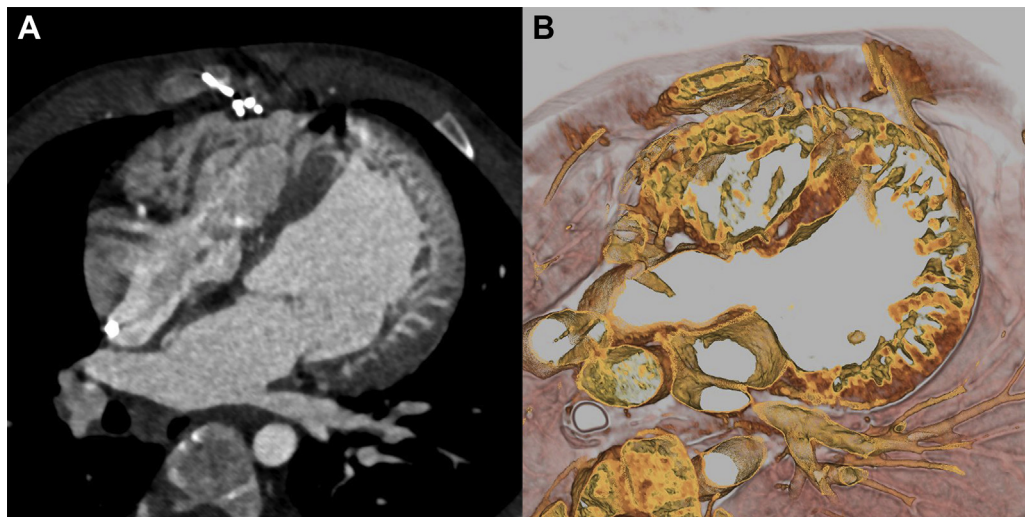


Figure 2 Computed tomography scan (A) with 3-dimensional reconstruction (B) demonstrating left ventricular noncompaction.

norepinephrine release to the LV myocardium. This increases the ventricular fibrillation threshold without reducing the heart rate or myocardial contractility.⁹ In patients with LQTS, CPVT, and some cardiomyopathies, the reduction in myocardial sympathetic stimulation following LCSD can result in a marked reduction in arrhythmia burden and cardiac events. Our first patient had suppression of VT under anesthesia with propofol and intravenous lorazepam, suggesting a benefit to decreasing catecholaminergic tone. However, the second patient did not have any clearly identified clinical catecholaminergic association with his VF, suggesting his clinical improvement was likely related to effects of the LCSD other than simply a decrease in adrenergic tone. The exact mechanism by which ventricular arrhythmia is reduced in patients with scar-mediated ventricular arrhythmia is complex and incompletely understood, but in addition to reduced localized myocardial sympathetic stimulation, LCSD and bilateral denervation also affect repolarization time and QT dispersion that may in part explain the reduction of ventricular arrhythmias associated with scar-related VT.^{10–12}

Despite LCSD not being well studied in the pediatric CHD population, denervation therapy has thus far allowed both of these patients to avoid transplant and have no VF or VT recurrences. In addition, both of these patients were also able to discontinue amiodarone therapy, thus avoiding potentially toxic side effects, without recurrence of ventricular arrhythmia thus far. The adhesions from prior cardiac surgeries made LCSD more technically challenging, but it was still accomplished via a minimally invasive thoracoscopic approach without complications. Considering the marked response observed in these patients, there may be a role for utilizing LCSD as an earlier intervention for recurrent ventricular tachyarrhythmias in the pediatric CHD population. These cases also demonstrate that LCSD may have benefit for a wider variety of substrates with recurrent ventricular tachyarrhythmias in pediatric patients and should be considered even in those with prior surgical repair of CHD. This

expanded utilization of LCSD has yet to be thoroughly studied, but should be considered in difficult cases before transplant.

Conclusion

Thoracoscopic LCSD may be beneficial in pediatric patients with CHD and refractory ventricular tachyarrhythmias.

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