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(CASE REPORT)

Case report: Left cardiac sympathetic denervation via thoracotomy in a newborn with congenital long QT syndrome

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Abstract

Objective: Congenital Long QT Syndrome (LQTS) is a rare channelopathy associated with malignant arrhythmias and sudden cardiac death. This case report aims to highlight the role of Left Cardiac Sympathetic Denervation (LCSD) as a rescue therapy in neonates with drug-refractory LQTS.

Methods: We present the case of a female neonate diagnosed with congenital LQTS type 8 (Timothy syndrome), complicated by 2:1 atrioventricular block and recurrent torsades de pointes despite pacemaker implantation, beta-blockade, mexiletine, and magnesium. Due to persistent arrhythmias and a transient response to left stellate ganglion block, LCSD was performed via thoracotomy.

Results: The surgery involved resection of the left stellate ganglion and thoracic ganglia T2–T4 through a left thoracotomy approach. The immediate postoperative course was uneventful. Genetic analysis confirmed a CACNA1C mutation. Despite the intervention, the patient died on postoperative day seven due to progressive cardiac failure.

Conclusion: This case underscores the potential role of LCSD in severe neonatal LQTS unresponsive to conventional therapy. However, its efficacy appears to depend heavily on genetic subtype and timing of intervention. Early genetic testing and timely surgical decision-making may improve outcomes in such high-risk patients. Further investigation is needed to determine optimal indications and timing for LCSD in neonates with LQTS.

Keywords: Cardiac sympathetic denervation; Congenital arrhythmia; Long QT syndrome; Neonate; Pediatric cardiac surgery

1. Introduction

Long QT Syndrome (LQTS) is a rare genetic disorder of cardiac repolarization caused by mutations affecting ion channels. This condition can lead to torsades de pointes and cardiac arrest, with an estimated prevalence of 1 in 2,000 live births [1]. Diagnosis is based on electrocardiographic findings, notably a prolonged corrected QT interval (QTc), and may be confirmed by genetic testing. First-line treatment consists of beta-blockers, with pacemaker or implantable cardioverter-defibrillator (ICD) therapy considered in more severe cases. Left Cardiac Sympathetic Denervation (LCSD), by reducing sympathetic input to the myocardium, represents an alternative therapeutic option in cases refractory to conventional treatment.

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2. Materials and Methods

A female neonate was delivered at 37 weeks of gestation by cesarean section due to fetal bradycardia detected as early as 23 weeks (2:1 atrioventricular block). At birth, she presented with persistent bradycardia (50–60 bpm), a corrected QT interval (QTc) > 600 ms, and 2:1 AV block. Electrocardiography (ECG) confirmed the diagnosis of congenital Long QT Syndrome (LQTS). In the absence of a family history, a pacemaker was implanted on day 1. Propranolol was initiated at 4.7 mg/kg/day, and the infant was discharged home on day 18.

On day 21, the patient experienced a cardiac arrest due to torsades de pointes, which was successfully resuscitated after 20 minutes of advanced life support. Upon admission to the intensive care unit, torsades de pointes persisted despite pacemaker optimization, initiation of mexiletine, and intravenous magnesium sulfate. Echocardiography revealed biventricular dysfunction secondary to the cardiac arrest, as well as a persistent ductus arteriosus.

A temporary left stellate ganglion block using Naropeine (2 mg/ml at 1 mg/kg) resulted in transient improvement, prompting the indication for surgical Left Cardiac Sympathetic Denervation (LCSD).

Surgical Procedure: LCSD was performed via a left thoracotomy under general anesthesia. The infant was positioned in a right lateral decubitus position, with the left arm abducted above the head and a roll placed under the right shoulder. A posterolateral skin incision was made in the fourth left intercostal space, between the mid-axillary and paravertebral lines. Muscle layers (latissimus dorsi, serratus anterior, and intercostal muscles) were dissected using electrocautery.

The left lung was covered with moist gauze and gently retracted anteriorly and inferiorly using a soft tissue retractor. The parietal pleura was carefully opened and exposed. The sympathetic trunk was first identified along the posterior aspect of the thoracic cavity, where it was readily visualized. The left stellate ganglion, along with the T2 to T4 thoracic ganglia, were identified, resected, and sent for histopathological analysis.

The procedure was uneventful, and the patient was transferred to the intensive care unit for postoperative monitoring.

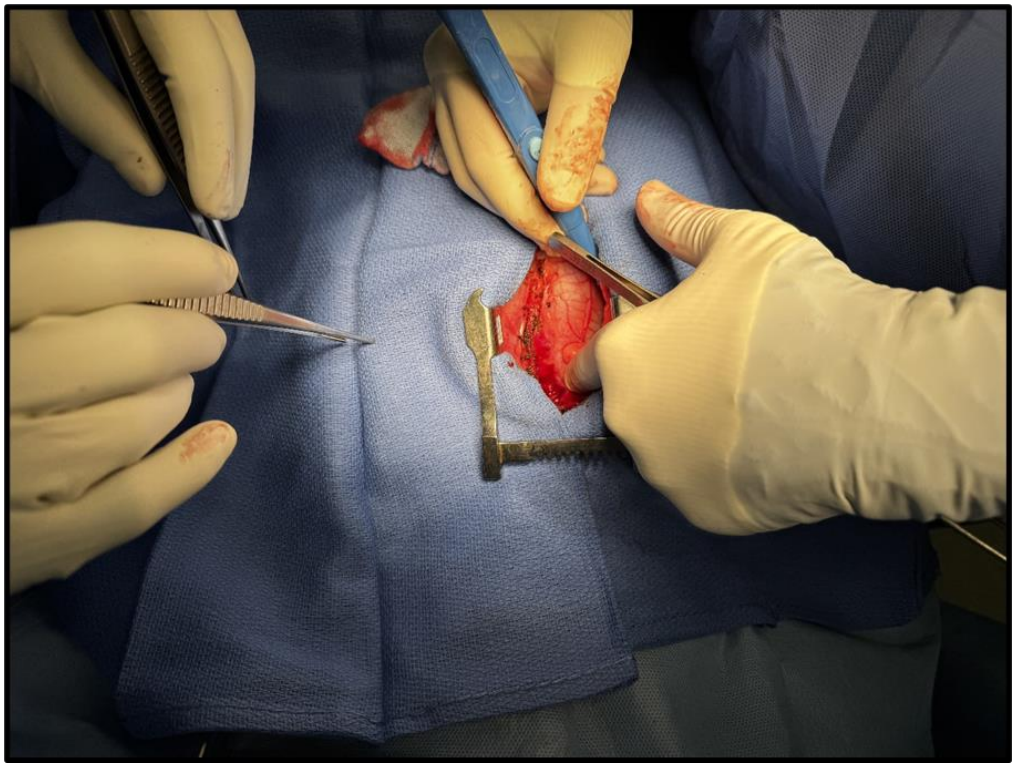

Figure 1 Exposure of the left thoracic sympathetic chain during pediatric LCSD

3. Discussion

Histopathological and genetic analysis of the resected ganglia confirmed the diagnosis of type 8 Long QT Syndrome (LQT8), also known as Timothy syndrome—an extremely rare form (<1 in 1,000,000), associated with multisystem involvement and mutations in the *CACNA1C* gene [2]. The severity of this genotype may explain the lack of response to conventional therapy.

LCSD is considered a last-resort treatment in drug-refractory LQTS. While well-documented in older children and adults, its use in neonates remains rare. The procedure reduces sympathetic tone, thereby lowering the risk of life-threatening arrhythmias. Several studies have demonstrated that LCSD significantly reduces arrhythmic episodes, including recurrent syncope and the risk of sudden cardiac death [3]. However, its efficacy is genotype-dependent and varies based on the extent to which the autonomic nervous system contributes to arrhythmia generation [4].

A comprehensive literature review, including the article by Duthoit titled *"Sympathetic denervation in the management of ventricular tachycardia"* [5], highlights the role of sympathectomy in controlling refractory arrhythmias. This study supports the concept that sympathetic modulation via denervation may provide therapeutic benefit in selected patients with drug-resistant ventricular rhythm disorders. Although most data come from adult and older pediatric populations, such an approach could be considered more systematically in neonates with severe forms of LQTS.

An alternative to the thoracotomy approach has been proposed by Odero et al. in their article *"Left cardiac sympathetic denervation for the prevention of life-threatening arrhythmias: The surgical supraclavicular approach to cervicothoracic sympathectomy."* This technique uses a supraclavicular approach to access and transect the sympathetic chain, making the procedure less invasive and avoiding thoracotomy-related complications [6]. Although promising, this approach remains understudied in neonatal LQTS, and further research is required to assess its long-term efficacy and safety.

In our case, the decision to proceed with LCSD, although delayed, was justified by the persistence of arrhythmias despite maximal therapy. Unfortunately, the clinical outcome was unfavorable: the patient died seven days postoperatively.

This case raises several important questions:

- Would earlier LCSD have improved the outcome?
- Could a neonatal ICD implantation have been feasible?
- Would genotype-guided therapy have altered the disease course?

Abréviations

- LQTS : Long QT interval Syndromes
- LCSD : Left Cardiac Sympathetic Denervation
- ICD : Implantable Cardioverter-Defibrillator
- AV BLOCK : AtrioVentricular Block
- ECG : ElectroCardioGraphy
- LQT8 : Type 8 of Long QT Syndrome
- QTc : Corrected QT interval
- HRS : Heart Rhythm Society
- EHRA : European Heart Rhythm Association
- APHRS : Asia Pacific Heart Rhythm Society

4. Conclusion

Severe congenital Long QT Syndrome is a pediatric emergency that requires specialized management. LCSD may represent a therapeutic option in refractory cases; however, its efficacy is highly dependent on the patient's genotype and the timing of the intervention. This case underscores the importance of early genetic testing and the need for tailored therapeutic strategies in severe neonatal presentations.

Compliance with ethical standards

Disclosure of conflict of interest

The authors declare no conflicts of interest.

Statement of ethical approval

This publication complies with confidentiality standards. Ethics committee approval was not required for this anonymous clinical case report.

Statement of informed consent

Written informed consent was obtained from the patient's parents for the publication of this case.

Author Contributions:

All authors had full access to the data and contributed equally to the analysis and writing of the manuscript. All authors approved the final version and are accountable for all aspects of the work.

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