

Case Report

Anesthetic Management of Jervell and Lange-Nielsen Syndrome With Long QT Undergoing Surgical Sympathectomy: A Pediatric Case Report

Mohsen Ziyaeifard¹, MD; Seyed Hasan Attarzadeh¹, MD; Maryam Moradian^{1*}, MD

ABSTRACT

Jervell and Lange-Nielsen syndrome (JLNS) is an uncommon hereditary condition. Characterized by deafness at birth, JLNS is associated with anomalies that distress the electrical system of the heart and causes long QT syndrome. Patients with long QT syndrome are at risk of severe ventricular arrhythmias. Every change in autonomic balance, particularly in the perioperative period, renders patients prone to the risk for torsades de pointes and sudden cardiac arrest and death. Herein, we describe a 6-year-old girl scheduled for left cardiac sympathetic denervation due to frequent implantable cardioverter-defibrillator shocks. (*Iranian Heart Journal 2019; 20(4): 103-107*)

¹ Rajaie Cardiovascular, Medical, and Research Center, Iran University of Medical Sciences, Tehran, IR Iran.

*Correspond Author: Maryam Moradian, MD: Rajaie Cardiovascular, Medical, and Research Center, Iran University of Medical Sciences, Tehran, IR Iran.

Email: maryam_moradian2008@yahoo.com

Tel: 02123922170

Received: January 6, 2019

Accepted: March 8, 2019

Jervell and Lange-Nielsen syndrome (JLNS) is an uncommon hereditary condition characterized by deafness at birth. It is associated with anomalies that distress the electrical system of the heart and causes long QT syndrome. JLNS varies from person to person in the rigorousness of the cardiac symptoms that are associated with it. Some patients may be asymptomatic, while others may develop abnormally increased heart rates or tachyarrhythmias—resulting in syncope, coma, cardiac arrest, and possibly sudden death.¹ Physical activity, seizure, excitement, or anxiety may activate the onset of these symptoms. JLNS is typically distinguished during early infancy, and it is inherited as an autosomal recessive genetic abnormality.

Before the age of 15, the outcome in more than 50% of the untreated cases of JLNS is death. These congenital cardiac channelopathies are, albeit life-threatening, temporarily treatable situations.² Patients with long QT syndrome are at risk of severe ventricular arrhythmias. Every change in autonomic balance, particularly in the perioperative period, places patients at risk for torsades de pointes and sudden cardiac arrest and death. The perioperative management of these patients, particularly when the sympathetic nervous system is manipulated, merits further research.³ We describe a 6-year-old girl with JLNS that underwent surgical left cardiac sympathetic denervation due to recurrent implantable cardioverter-defibrillator (ICD) shocks.

Case Report

Herein, we describe a 6-year-old girl scheduled for left cardiac sympathetic denervation due to frequent ICD shocks. These shocks not only made her anxious but also led to a recurrent need for generator replacements. The patient was a known case of JLNS, a form of long QT syndrome, with a past medical history of cochlear implantation at the age of 20 months owing to congenital sensorineural hearing loss. Additionally, she had undergone ICD placement 1.5 years earlier because she had experienced frequent syncopal attacks despite beta-blocker therapy.

Her parents, who were relatives, reported that their first daughter had died after sudden cardiac arrest at the age of 7 years old. They revealed that their late child also suffered from hearing loss and had a cochlear implant. Their second child, an 11-year-old boy, is completely healthy and his hearing, cardiac, and genetic evaluations are normal. Their third child, our patient, exhibited the same symptoms as her late sister. Nonetheless, previous experience prompted her parents to seek medical attention early and genetic testing confirmed that she suffered from JLNS when she was 10 months old. The patient was regularly examined by an otolaryngologist and a pediatric cardiologist. Her cochlear device was implanted under the supervision of a cardiac anesthesiologist when she was 20 months old, and she received propranolol (3 mg/kg per day). She was well for 4 years; however, afterward, she experienced frequent syncopal episodes despite beta-blocker use. Consequently, an ICD was implanted to prevent sudden cardiac death when she was 4.5 years old. One and a half years after the ICD implantation, the child suffered repetitive ICD shocks, leading to the end of life of the generator and necessitating a new device replacement twice. Physical examinations were normal except for the fact that she was mildly underweight (16 kg). In addition, her blood pressure, respiratory rate,

and heart rate were normal for her age. Cardiac auscultation revealed a 2/6 systolic murmur at the left sternal border without radiation. On chest X-ray, the cardiothoracic ratio and the cardiac silhouette were normal and the ICD could be seen in the left hemithorax (Fig. 1). On electrocardiography, the QT interval was increased and the corrected QT interval was 0.47 second, indicating long QT syndrome (Fig. 2). The only finding on echocardiography was mild tricuspid regurgitation, with a 20-mm Hg pressure gradient. The cardiac anatomy and both ventricular functions were normal. Finally, her physicians decided that sympathetic denervation might reduce the ICD shocks.

Fifteen minutes before the patient was transferred to the operating room, 0.25 mg/kg of midazolam hydrochloride syrup was administered as premedication. In the operating room, defibrillator pads were applied to the chest; and after ECG, pulse oximetry, and noninvasive blood pressure monitoring, an intravenous line was inserted. Anesthesia was induced using 5 µg/kg of fentanyl, 1.5 mg/kg of propofol, and 1 mg/kg of vecuronium bromide. Cuffed endotracheal intubation (No. 5) was performed, and arterial and central lines were inserted. Anesthesia was maintained with propofol and fentanyl infusion with boluses of vecuronium. Left lateral thoracotomy was carried out, with the patient on 2 lung ventilators and end-tidal CO₂ monitoring. (The average value of end-tidal CO₂ was 45–50 mm Hg.) The fourth intercostal space was opened via surgical sympathectomy and thoracotomy, and half of the satellite ganglia and sympathetic chain at T₂–T₄ level were removed. For postoperative pain management, 0.125% bupivacaine was injected into the intercostal nerve.

Intraoperatively, there was no major cardiac event, and the child was transferred to the intensive care unit (ICU) on mechanical ventilation. The patient was extubated at 6 hours after the operation as she did not have

any further episodes of postoperative ventricular tachycardia. One day after the operation, she suffered left myosis and ptosis. Neurological consultation indicated that the complications were the iatrogenic effects of

sympathectomy and no further work was required apart from following the symptoms. The child was discharged from the ICU on the third postoperative day.

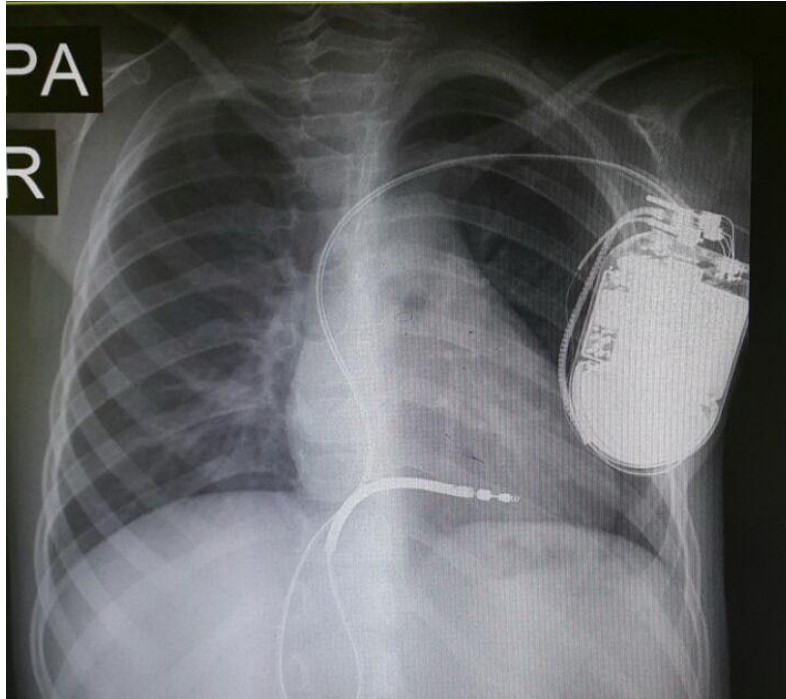


Figure 1. Chest X-ray, showing the position of the implantable cardioverter-defibrillator in its pocket

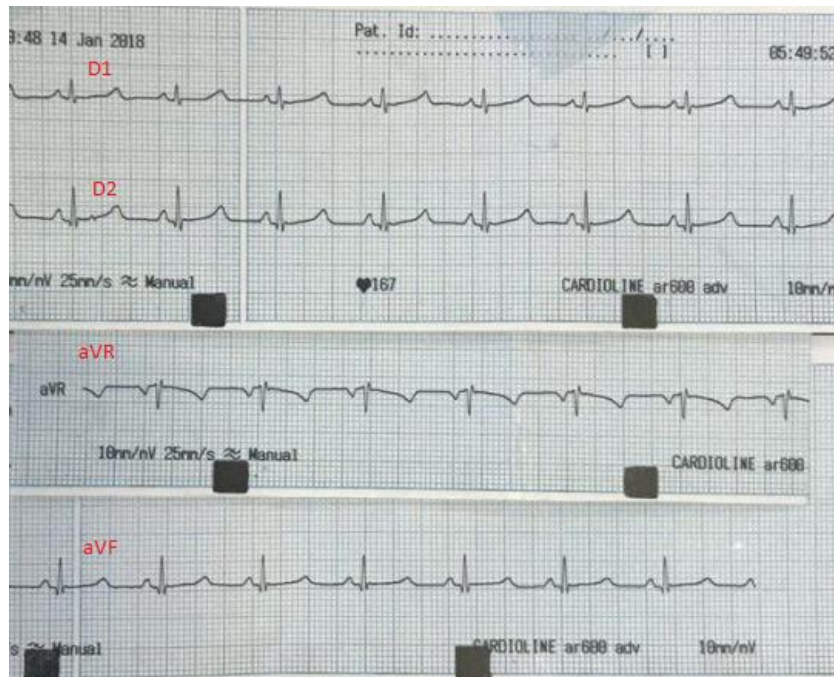


Figure 2. ECG tracing, showing QT prolongation

DISCUSSION

Although JLNS is uncommon, it is considered one of the most common syndromic types of sensorineural hearing loss with autosomal recessive inheritance.⁴ It is also a form of long QT syndrome, a familial condition that can lead to syncope and sudden death through fatal polymorphic ventricular tachycardias (torsades de pointes) in young people. Two genes, namely *KCNQ1* and *KCNE1*, which encode the regulation of the potassium flow both in cardiac cells and inner ears are known to be responsible for this syndrome.⁵⁻⁹

The first step in the evaluation of a child who experiences syncope is to take a thorough history with special attention to the cardiac and nervous systems. Among the 3 major categories of syncope—namely neurally mediated syncope, cardiovascular syncope, and noncardiovascular syncope—the second category (ie, cardiovascular syncope) is, albeit not common, potentially life-threatening.

Family history of syncope, sudden death at a young age, deafness, and seizure are potential clues in favor of cardiac causes necessitating more cardiovascular evaluation. Any left or right ventricular outflow tract obstructions like aortic or pulmonary valve stenoses, tumors in these regions, hypertrophic or congestive cardiomyopathies, pulmonary hypertension, and hypercyanotic attacks in cyanotic heart defects may lead to syncope; nevertheless, more commonly, arrhythmias are responsible for cardiac syncope.^{10,11}

In our case, a family history of deafness and sudden cardiac death in the elder sibling potentially helped to direct to the diagnosis of congenital cardiac channelopathy associated with hearing loss, JLNS. ECG and genetic study confirmed the diagnosis, and cochlear implantation was done to prevent future disabilities. Still, the problem was frequent syncopal attacks despite beta-blocker therapy, which led to ICD implantation. After the

implantation, our patient suffered frequent ICD shocks, rendering her depressed and anxious. Additionally, repetitive shocks led to the end of life of the generator, necessitating new device replacements twice.

In the case of frequent ICD discharges due to refractory ventricular tachycardias, while the patient is on beta-blockers, sympathetic denervation can be considered. The history of cardiac sympathetic denervation to reduce the occurrence of life-threatening ventricular arrhythmias goes back to 1960s and 1970s.¹²⁻¹⁴

It is proven that cardiac sympathetic denervation blocks norepinephrine release, reducing the initiation and continuation of ventricular arrhythmias.^{15,16} Accordingly, we scheduled our patient for sympathectomy and the removal of stellate and T2–T4 thoracic ganglia. Intraoperatively, there was no major cardiac event and the child was transferred to the ICU on mechanical ventilation. She was extubated 6 hours after the operation and suffered no further episodes of ventricular tachycardia following surgery. The child was discharged from the ICU on the third postoperative day. The only complication of surgery was transient Horner syndrome for a few months, which is relieved now. At 1 year's follow-up, she feels very well and has no ICD discharge.

REFERENCES

1. Peter J. Schwartz, MD Marco Stramba-Badiale. Prevalence of the Congenital Long-QT Syndrome (Circulation. 2009;120:1761-1767)
2. P. D. Booker, Whyte SD, Ladusans Ej. Long QT syndrome and anaesthesia Br J Anaesth 2003; 90: 349-66
3. <https://ghr.nlm.nih.gov/condition/jervell-and-lange-nilsen-syndrome> Genetics Home Reference
4. GeneReviews® [Internet]. Seattle (WA): University of Washington, Seattle; 1993-2018. 2002 Jul 29 [updated 2017 Aug 17]

5. Yuka Mizusawa, MD; Minoru Horie, MD, PhD; Arthur AM Wilde, MD, PhD Genetic and Clinical Advances in Congenital Long QT Syndrome *Circulation Journal* Vol.78, December 2014
6. Kenshi Hayashi, MD, PHD,.. Impact of Updated Diagnostic Criteria For Long QT Syndrome on Clinical Detection of Diseased Patients Results From a Study of Patients Carrying Gene Mutations *JACC: CLINICAL ELECTROPHYSIOLOGY* VOL.2,NO.3,2016
7. Guidelines for the diagnosis and management of Familial Long QT Syndrome The Cardiac Society of Australia and New Zealand
8. Update on the Diagnosis and Management of Familial Long QT Syndrome *Heart, Lung and Circulation* (2016) 25, 769–776
9. Peter J. Schwartz Michael J. Ackerman The long QT syndrome: a transatlantic clinical approach to diagnosis and therapy *European Heart Journal* (2013) 34, 3109–3116
10. Peter J. Schwartz Lia Crotti, MD, PhD QTc Behavior During Exercise and Genetic Testing for the Long-QT Syndrome *Circulation*. 2011;124:2181-2184
11. Khorgami MR, Moradian M, Omid N, Yousef M. Management of Cardiovascular Disorders in Patients with Noonan Syndrome. *J Teh Univ Heart Ctr* 2017;12(4):184-187.
12. <http://Thecardiologyadvisor.com/icd-shocks-evaluation-and-management>
13. Kathleen Kopecky MD ShelleyA.HallMD.. Bilateral sympathectomy for treatment of refractory ventricular tachycardia *Pacing Clin Electrophysiol*. 2018;41:93–95.
14. Sophie C. Hofferberth, ..Left thoracoscopic sympathectomy for cardiac denervation in patients with life-threatening ventricular arrhythmias *The Journal of Thoracic and Cardiovascular Surgery* 2014 Volume 147, Number 1
15. Christopher A.. Left cardiac sympathetic denervation for the treatment of long QT syndrome and catecholaminergic polymorphic ventricular tachycardia using video-assisted thoracic surgery *Heart Rhythm* 2009;6:752–759
16. Yongkeun Cho Left cardiac sympathetic denervation: An important treatment option for patients with hereditary ventricular arrhythmia *journal of Arrhythmia* 32 (2016) 340–343