

Congenital long QT syndrome and patent ductus arteriosus: A rare surgical scenario

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ABSTRACT

Congenital long QT syndrome (LQTS) is a rare cardiac condition characterized by abnormality of either sodium or potassium ion channels resulting in prolongation of QT interval and thereby predisposing to life-threatening arrhythmia. Once the syndrome is diagnosed, measures should be taken to avoid sudden cardiac death. We present a rare case of LQTS associated with patent ductus arteriosus in a child, and a unique approach was used in managing both conditions.

Keywords: Long QT syndrome, patent ductus arteriosus, ventricular arrhythmia

INTRODUCTION

Congenital long QT syndrome (LQTS) is a channelopathy characterized by abnormality of either sodium or potassium ion channels that result in the prolongation of QT interval and predispose to ventricular arrhythmia and sudden cardiac death. Clinical manifestations vary from asymptomatic to life-threatening arrhythmia. Once diagnosed, measures to avoid sudden cardiac death should be initiated that could often change the lifestyle and improve the longevity of the patient.^[1] We present an interesting case of LQTS associated with patent ductus arteriosus (PDA) in a child who presented with refractory ventricular tachycardia.

CASE REPORT

A 3-year-old girl was diagnosed with LQTS during infancy. Electrocardiogram (ECG) revealed a prolonged QT interval of 670 ms [Figure 1]. Holter reported prolonged QT with T-wave alternans (TWA) throughout the study.

Brainstem-evoked response audiometry showed bilateral sensorineural hearing loss. Echocardiography revealed the presence of a 5-mm PDA (left to right shunt) with mild pulmonary arterial hypertension. She was started on tablet propranolol 30 mg twice daily for LQTS. At 6 months of age, attempted PDA device closure was unsuccessful, as she developed torsades de pointes (TdP) at the time of induction of anesthesia and got reverted with defibrillation. She had poor weight gain and two episodes of syncope, and ECG was showing persistent TWA despite high-dose propranolol, and hence, a surgical approach to manage both PDA and LQTS was employed.

Left posterolateral thoracotomy incision was made, and the pleural cavity was entered through the left third intercostal space. Mediastinal pleura was opened, left sympathetic chain was identified, and T1-T5 ganglia were excised for cardiac sympathetic denervation [Figure 2]. PDA was identified and doubly

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ligated [Figure 3]. The pericardium was opened above the left phrenic nerve. Epicardial leads were placed on the left atrium and left ventricle. A pulse generator was connected, checked, and placed in the left pleural cavity fixed to the posterior chest wall. A pacemaker was interrogated, and the heart rate was set at 90/min. Postoperative ECG showed pacing rhythm with regular T-wave morphology, and the child was discharged on the 3rd postoperative day and is on regular follow-up [Figure 4].

DISCUSSION

PDA interruption, left cardiac sympathetic denervation (LCSD), and permanent pacemaker implantation are all common procedures. However, all three procedures in the same patient through left posterolateral thoracotomy have not been reported in the literature. LQTS has two hereditary variants, namely^[1] Jervell and Lange-Nielsen (JLN) syndrome and Romano-Ward syndrome. Our patient belongs to the first type as the child had deafness.

There are more than 300 mutations identified in eight LQTS genes, of which JLN syndrome is commonly due to homozygous or compound heterozygous mutation of *KCNQ1* and *KCNE1* genes. These channel genes encode the alpha- and beta-subunits of delayed rectifier potassium I_{ks} channel and cause LQT1 and LQT5, respectively.^[2] The most common subtype is LQT1 that accounts for nearly 50% of LQTS patients and is associated with maximum cardiac events.^[3] LQT2 is the second common form accounting for 40% and LQT3 occurring in 10% of LQTS. Timothy syndrome, LQT8, the most malignant form of LQTS is due to a mutation involving L-type calcium channels and is associated with neuropsychiatric manifestations, immune system dysfunction, intermittent hypoglycemia, cardiac malformations, and syndactyly.^[3]

Moss *et al.* described the first genotype-phenotype correlation study that highlighted the difference in the shape of the T-waves in the ECG among the three genotypes. The T-waves in LQT1 were tall and

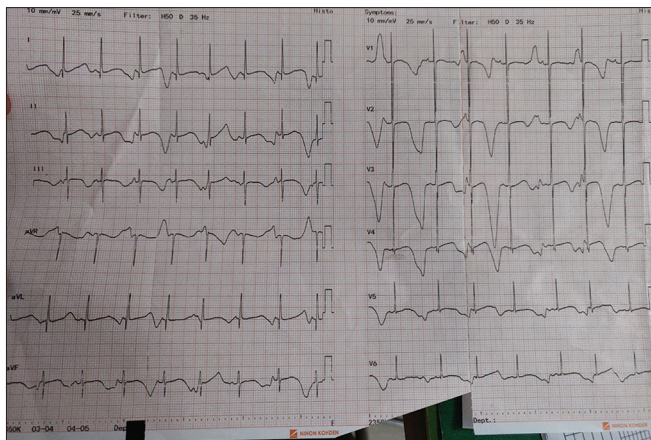


Figure 1: Electrocardiogram showing QT interval of 670 ms with T wave alternans

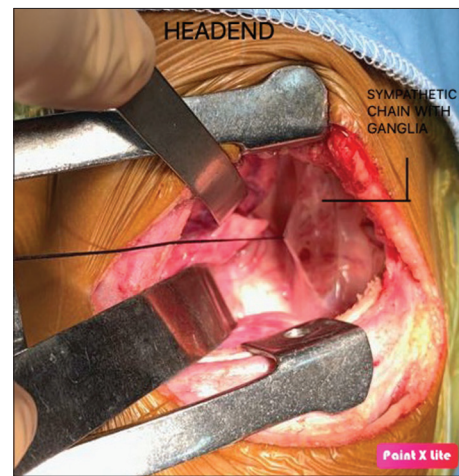


Figure 2: Cervical sympathetic chain with ganglia

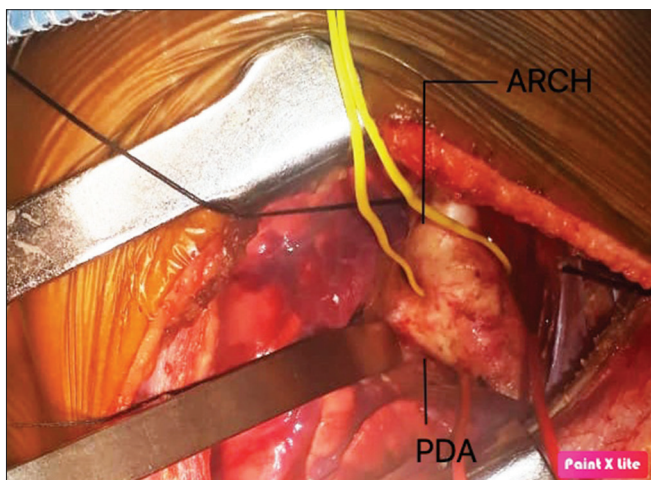


Figure 3: Large patent ductus arteriosus

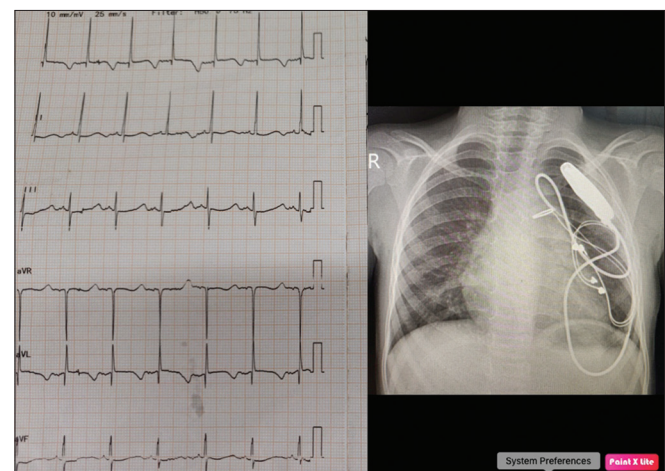


Figure 4: Postop X-ray showing pacemaker leads with pulse generator in the left pleural cavity and electrocardiogram showing sinus rhythm

broad, but in LQT2, they were notched, bifid, and of low amplitude. In LQT3, there was a long isoelectric segment with narrow and peaked T-waves. This correlation had more than 80% sensitivity and 70% specificity in LQT1 and LQT2, however, it was much lower in LQT3.^[4]

Beta-blockers represent the first choice therapy once the diagnosis is confirmed. Propranolol is the most commonly used drug at a daily dosage of 2–3 mg/kg.^[5] LCSD is an effective option for patients in whom beta-blockers are unsuccessful or contraindicated.^[5] There are multiple effects of LCSD that contribute to its clinical efficacy. Importantly, it raises the threshold for ventricular fibrillation, thus making it more difficult for a heart to fibrillate.^[5,6]

The fact that the onset of TdP is often preceded by a pause might add to the rationale of using a pacemaker as an adjunct to the therapy of LQTS patients.^[7,8] The ACC 2017 guidelines^[9] also recommend LCSD and/or ICD for symptomatic patients refractory to beta-blockers. Due to increased lead-related complications and psychosocial complications such as anxiety, depression associated with ICD, we preferred to do LCSD with pacemaker implantation for our patient.^[10,11] The peculiarity in our patient is that the association of LQTS with PDA is rare and all the procedures were performed through a single posterolateral thoracotomy incision which has not been reported in the literature so far.

Declaration of patient consent

The authors certify that they have obtained all appropriate patient consent forms. In the form the patient(s) has/have given his/her/their consent for his/her/their images and other clinical information to be reported in the journal. The patients understand that their names and initials will not be published and due efforts will be made to conceal their identity, but anonymity cannot be guaranteed.

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Conflicts of interest

There are no conflicts of interest.

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