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CASE REPORT

Pre-anesthesia evaluation leading to a diagnosis of long QT syndrome – a case study

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ABSTRACT

Long QT Syndrome is a rare disorder characterized by prolongation of QT interval on an ECG. Patients with this disorder have an increased risk of developing life-threatening arrhythmias such as torsades de pointes, and occasionally sudden cardiac arrest.

We present this case of undiagnosed Long QT Syndrome in a 7 years old boy, who presented to our service for incision and drainage of an abscess in the left axilla under general anesthesia. This case highlights the importance of proper and meticulous pre-anesthesia evaluation, even in low risk patients, so that potentially harmful medical conditions are diagnosed well in time and managed accordingly.

Key word: Long QT Syndrome; Anesthesia; Pediatrics; Torsades de Pointes

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INTRODUCTION

Long QT Syndrome (LQTS) is an inherited or acquired disorder resulting from cardiac ions channel disorders. During repolarization, potassium enters the myocardial cells through both the rapid (I_{KR}) and slow (I_{KS}) potassium channels. Genetic mutations, medical conditions or drugs that alter the activities of these channels cause abnormalities in repolarization of the myocardium that results in long QT interval as seen on an ECG.[1]

The continuous advances in health care management have led to a relatively increased number of diagnoses of long QT syndrome in recent years. The prevalence of LQTS has been reported to be around 1 in 2000 in the general population.[2]

The QT interval is measured on ECG trace from the beginning of the QRS complex to the end of T wave. This interval depicts the ventricular depolarization and repolarization. Long QT is described as a QTc of more than 0.44 seconds in both genders from birth until adolescence, and as more than 0.45 sec in adult males and more than 0.47 in adult females.[3] Since QT interval is inversely related to heart rate, a correction of heart rate to 60 bpm is made in calculation.[4] In this case, we use the widely popular Bazetts formula ($QTc = QT\sqrt{RR}$) for calculation of the corrected QT interval.[5]

Presentation in patients with long QT is either based on the symptoms of syndromes associated with long QT such as congenital deafness in Jervell and Lange-Nielsen syndrome, or symptomatology due to the QT interval such as syncope.[6] This makes it a challenge to investigate and diagnose asymptomatic individuals who are found to have an incidental finding of long QT interval on ECG, and so a full list of investigations may be warranted.

We present a case of a young child who was incidentally found out to be suffering from this syndrome.

CASE REPORT

A 7-year-old boy (Weight: 19 kg, Height: 116 cm) got admitted from the emergency department for incision and drainage of left axillary abscess. On history taking, there was no significant past medical or surgical history. He had a positive family history of sudden death of his mother at the age of 24 years, preceded by an episode of syncope.

During routine pre-anesthesia evaluation, it was noticed that the patient was having episodes of bradycardia in the range of 50-60 bpm during sleep and a regular heart rate of 70-80 bpm when awake. A subsequent ECG was requested that showed normal sinus rhythm, heart rate of 65 and a QTc of 447 ms. Patient was referred to pediatrics department for evaluation and was provisionally diagnosed to have LQTS after thorough investigations including ruling out electrolyte abnormalities and scoring as per the Schwartz Criteria. The patient had no new active complaints and was asymptomatic from a cardiac standpoint.

The anesthesia team reviewed the patient again before the surgery and a high-risk consent was obtained because of the increased risk of sudden cardiac arrest among these patients. Repeat lab investigations were reviewed before the surgery to rule out any correctable abnormalities.

A calm and comfortable environment was provided before induction of anesthesia to minimize anxiety factor. All precautionary measures were taken before the start of the surgery, including preparation of an emergency trolley inside the theatre with the availability of a defibrillator. ECG leads were applied to the patient for continuous observation. Upon arrival in the operating room, standard monitoring was applied as per AAGBI Guidelines.[7] Chest pads were applied, in case defibrillation was required intra operatively. Patient was induced with fentanyl 25 µg and propofol 80 mg IV. Airway was secured with size 2.5 LMA, maintenance of anesthesia was achieved using sevoflurane in oxygen and air mixture to reach MAC 1.0. Patient remained stable during the surgery with normal variability in his heart rate ranging from 80 -110 bpm. It was a short surgical procedure and surgical time was about 10 min. Child woke up smoothly, and postoperatively he was shifted to the pediatric ICU for standard monitoring for 24 hours.

Continuous monitoring and repeat ECG postop showed QT interval to be 460 ms. Patient remained asymptomatic and was discharged from the hospital in good health after recovering completely from the surgery with a follow up appointment in the outpatient clinics with pediatrics services. Serial repeat ECGs were obtained in the OPD that showed the QTc ranging from 450 ms to 480 ms, all of which are higher than what is acceptable for his age.[7] Diagnosis of LQTS was confirmed, and the patient was referred to pediatric cardiology department for further evaluation and management.

DISCUSSION

Routine monitoring in asymptomatic patients, if carefully observed may help in the early diagnosis of potentially fatal medical conditions. This case presented here highlights the importance of proper and scrupulous evaluation in these asymptomatic patients who are undergoing anesthesia. Timely diagnosis in these patients may lead to early intervention in the disease process and a better long term outcome.

LQTS is a rare genetic or acquired cardiac channel disorder that renders the individual more susceptible to developing fatal arrhythmias such as torsades de pointes and ventricular fibrillation, which may result in sudden death. It may either present with very vague symptoms or may be an incidental finding in asymptomatic individuals, as was the case with our patient. A normal variation exists in QT interval due to different physiological factor, as it is inversely correlated to the heart rate. All these factors make it very challenging to diagnose and establish a clinical pathway for patients who are not in any acute cardiac distress.[8]

After ruling out reversible causes of prolonged QT, the best initial test that helps in establishing a diagnosis is repeat ECGs that show consistently prolonged long QT intervals. This initial suspicion is then complemented by the use of the Schwartz score (Table 1) that gives points based on ECG findings, clinical findings and family history to help establish the diagnosis of long QT syndrome.[5]

Score of < 1 indicates a low probability of having the disease. Score of 1.5 to 4 signifies an intermediate risk and a score $\geq$ 3.5 increases the probability of the patient having LQTS. After a careful evaluation of our patient, the Schwartz score was calculated to be 3.5, which indicated a high probability of having the disease.

Anesthesia management of LQTS can be challenging because of the limited literature available. We summarize the anesthetic management of patients with known LQTS in Table 2.[9,10]

Table 1: Schwartz Criteria for the Diagnosis of LQTS

1. ECG findings	Score
• QTc (Bazettes formula, QTc = QTv RR)	
o =480 msec	3 points
o 460 to 470msec	2 points
o >450 to 460msec (in males)	1 point
• QTc =480 at 4th minute of recovery from exercise stress test	1 point
• Torsades de pointes (mutually exclusive points of syncope)	2 points
• T-wave alternans	1 point
• Notched T wave in three leads	1 point
• Resting heart rate lower for age (in children)	0.5 points
2. Medical history	**Score**
• Syncope (mutually exclusive points of torsades)	
o With stress	2 points
o Without stress	1 point
o Congenital deafness	0.5 points
3. Family history	**Score**
• Family members with LQTS	1 Point
• Unexplained sudden cardiac death in immediate family members <30 years of age	0.5 points

Table 2: Anesthetic management of patients with known LQTS

Preoperative	Perioperative	Postoperative
• Ensure therapeutic beta blockade continues. • Ensure normal electrolyte profile (K^+, Ca^+, Mg^{++}). • Avoid drugs that further prolong QT interval. • Drugs to avoid o *Antiarrhythmic agents.* o *Phenothiazine antipsychotics* o *Selective serotonin reuptake inhibitors* o *Macrolides antibiotics* o *5-HT1 agonists* o *Antimalarial agents* o *Anti histaminics (terfenadine)* o *Prokinetic agents (cisapride)* • Continue genotype directed therapy. • Prescribe anxiolytics premedication • If symptomatic, consider pacing in liaison with cardiologist. • If pacemaker or ICD in situ, check settings.	• Per-induction monitoring of >1 ECG lead. • Low threshold for intra-arterial monitoring. • Establish central venous access to facilitate emergency pacing. • Thiopentone or propofol for induction. • Consider propofol maintenance. • Avoid halothane; all volatiles prolong QTc. • Vecuronium is probably safe. • Cisatracurium theoretically attractive but no clinical experience. • Avoid reversal of muscle relaxant if possible. • Minimize sympathetic stimulation, topical Local Anesthesia during laryngoscopy and intubation. • Apply regional technique where appropriate. • Maintain normoxia, normocarbia, normothermia and normoglycemia • Maintain normal serum K^+, Ca^{++}, and Mg^{++}	• Continuous ECG monitoring • Recovery in quiet environment • Ensure dependency /intensive care unit monitoring. • Good analgesia.

Once the diagnosis of LQTS is established, it is important to initiate the most effective therapy. One study reports that the mortality of the patients who are adequately treated is around 1%.[11]

The cornerstone for the management of LQTS is beta-blockade, with propranolol being the most favorable drug of choice. Although the protective role of beta-blockade in these patients is not fully understood, it has been shown to reduce mortality in patients with LQTS.[11]

The second line in the treatment is automated implantable cardioverter-defibrillators (AICDs). The role of AICDs is not to prevent the incidence of fatal arrhythmias in these patients, but to reduce the mortality associated with them. AICDs are recommended in patients who continue to remain symptomatic with episodes of syncope despite adequate beta-blockade therapy, patients who required resuscitation from cardiac arrest, or patients that have a QTc > 550.[11,12]

The third line of treatment in LQTS is left sided cardiac sympathetic denervation, which involves the removal of the first 5 left thoracic ganglia and the left stellate ganglion. This approach is recommended in high-risk infants and children in whom AICDs are not a very feasible option, and also in patients who cannot be adequately managed with beta-blockade and AICDs.[11,12]

CONCLUSION

LQTS represents a group of cardiac ion channel disorders. Although relatively rare, its important lies in the significant morbidity and mortality associated with failure to recognize and treat symptomatic patients, and the potential for anesthesia to induce life-threatening arrhythmias. The perioperative period is a time of high risk for patient with LQTS. The anesthetist is faced with difficult decisions about the best way to conduct anesthesia, and has scant information or evidence on which to base those decisions.

LQTS pathophysiology illustrates the clinical relevance of basic scientific research into the effects of anesthetic management of LQTS patients, future investigation and pharmacological influences on transmural dispersion of repolarization.

Conflict of interest: None declared by the authors

Authors' contribution:

MA–Case writing, literature search, final editing and revision
SMY– Case writing, literature search
MMC– Case writing, editing and revision
SA– Final editing and revision.

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