



Safe Anaesthesia Management in a Child with Congenital Long QT Syndrome

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Long QT syndrome is a cardiac repolarisation disorder that can occur with clinical symptoms such as dizziness, fainting, life-threatening arrhythmias and sudden cardiac death, and its incidence is increasing in the general population. A careful anaesthetic management is required for patients with this syndrome because of the risk of torsades de pointes and malignant arrhythmias. In this case report, we discuss the anaesthetic management of a seven-year-old patient with congenital long QT syndrome that was diagnosed during the preoperative evaluation.

Keywords: Long QT syndrome, anaesthesia, torsades de pointes

Introduction

Long-QT syndrome (LQTS) is a cardiac repolarization disorder that occurs with the clinical symptoms such as dizziness, fainting, life-threatening ventricular arrhythmias and sudden cardiac death. Its aetiology may be congenital or acquired (1, 2). Hereditary form of LQTS is associated with the mutation in genes responsible for the formation of potassium channels. The iatrogenic form depends more on drugs and electrolyte imbalance. Idiopathic or heritable LQTS is a disease characterized by QT interval prolongation, tachyarrhythmia triggered especially by emotional or physical stress, syncope attacks and even sudden death (3). The incidence is between 1/3000 and 1/5000 (4). When a careful history taking is not performed, it may often be confused with epileptic seizures. Delay in diagnosis may cause life-threatening conditions. The anaesthetic management we performed for a seven-year old patient who had congenital LQTS and was diagnosed with LQTS in preoperative evaluation was discussed in this case report.

Case Presentation

Seven-year-old male patient was admitted to our ear nose and throat (ENT) polyclinic because of persistent nasal congestion, continuing for about three months, and frequent attacks of snoring and tonsillitis. Following physical examination, the patient was referred to anaesthesia outpatient clinic for adenoidectomy and tonsillectomy. History taken from the relatives of the patient revealed that father, grandmother and close relatives had dysrhythmias, and an etiologically unknown sudden death had occurred in one of his cousins. The patient was asked to consult with paediatric cardiology. The echocardiography of the patient revealed 72% of ejection fraction, 1st degree tricuspid regurgitation and 1st degree mitral regurgitation. After examination and preliminary evaluation, inherited LQTS (QT = 0.447) was diagnosed because of long QT interval in the electrocardiography (ECG). After prescribing propranolol HCl 20-mg tablets (Dideral®) 2×1 perorally, surgery was not considered to be risky in terms of cardiology with the suggestions of intraoperative close monitoring and administering anaesthetic agents that do not prolong the QT interval. The preoperative evaluation revealed normal vital signs; in addition, the lungs were well ventilated, crackles and rhonchi were absent, mouth opening was good, head and neck movement was not restricted, and no pathology was detected. In the laboratory tests, Hb was found to be 12.4 g dL⁻¹, haematocrit 35.3% and platelet count 193×10³ µL⁻¹; other electrolyte and biochemical results were within normal limits. The relatives of the patient were informed in terms of the anaesthesia risks, and written informed consent was received for the intervention to be performed.

In the morning of the surgery, the patient was premedicated with midazolam 0.7 mg kg⁻¹ by the nasal route and taken to the operating room. After establishing vascular access, lidocaine 1 mg kg⁻¹, fentanyl 1 µg kg⁻¹, propofol 2.5 mg kg⁻¹ and rocuronium bromide 0.5 mg kg⁻¹ were administered intravenously. Together with a mixture of 50% oxygen and 50% air, 8 mg kg⁻¹ propofol infusion [Total intravenous anaesthesia (TIVA)] was administered during the trial. No complications were seen during the induction phase or the surgical procedure. In the cardiac monitoring of the patient, whose vital signs remained stable, no change occurred in heart rate and rhythm. The duration of the intervention was recorded as 35 minutes. Towards the end of the surgical procedure, TIVA was terminated and the rebreathing process was initiated. The effect of the muscle relaxant was reversed at a dose of sugammadex 2 mg kg⁻¹ intravenously. The patient was taken to the recovery room after a comfortable awakening and extubation, and he was monitored for 2 hours. The patient remained stable after 24 hours of observation in the intensive care unit, and he was transferred to the ENT department on the second day.

Discussion

LQTS is a rarely seen ventricular repolarization disorder that causes prolonged QT interval in a structurally normal heart, recurrent syncope attacks, tachyarrhythmia and sudden death. In this syndrome, characteristic polymorphic ventricular tachycardias occur because of inadequate ventricular repolarization resulting from cardiac ion channel dysfunction (5). LQTS more commonly manifests in the third decade; however, it can manifest at any age. Patients diagnosed with LQTS were also reported to be in new-born and intrauterine life (6). LQTS is clinically important; because it is a risk factor for ventricular arrhythmias. The observed arrhythmias are “torsade de pointes (twisting of the points; TdP)” type arrhythmias that progress along with axis changes in ECG; these arrhythmias have a characteristic of a polymorphic ventricular tachycardia and a high mortality rate. The main symptom of LQTS is the syncope caused by the ventricular tachycardia of TdP type. TdP is observed at a rate of 1–8% in treatment with antiarrhythmic drugs that prolong the QT; it is also observed after QT interval prolongation primarily while returning to sinus rhythm with antiarrhythmic drugs or during sinus rhythm. In our patient, the absence of any history of drug use and the presence of arrhythmias in close relatives suggest that LQTS is hereditary.

Stress, which causes sympathetic stimulation, and many drugs including anaesthetic agents can induce TdP. Halogenated volatile anaesthetics (halothane, isoflurane, desflurane and sevoflurane) were shown to prolong the QT interval in the studies in the literature (7-9). However, although the effects of the findings of this study are not very clear in the clinical use, it is recommended that these agents should be avoided in

LQTS patients. Paventi (10) stated in his study that sevoflurane prolongs the QT interval and propofol shortens it. On the basis of all these findings, TIVA administration was preferred in our case; this is because we considered that propofol infusion would be safer instead of volatile anaesthetics.

The use of beta-blockers in patients with LQTS is recommended before and after the surgery, and magnesium sulphate administration and defibrillator application are recommended to be available in the operating room (11). In our case, preoperative beta-blocker therapy was initiated and maintained during the trial; in addition, magnesium sulphate, short-acting beta-blockers that could be administered as infusions and defibrillator were kept ready in the room.

Anxiety, crying, shouting, loud noises and operating environment can trigger the TdP by causing sympathetic stimulation (12). Therefore, midazolam was administered to the patient in the preoperative period as a premedication. In the postoperative period, paracetamol was administered intravenously at a dose of 2 mg kg⁻¹ in order to avoid agitation because of pain.

Conclusion

In the preoperative evaluation of the patients, LQTS should be kept in mind and ECG should be examined more carefully. In paediatric patients with the story of syncope and arrhythmias, LQTS must be taken into consideration. In patients diagnosed or suspected with LQTS, propofol infusion (TIVA) can be considered as a safe option in the maintenance of anaesthesia.

Informed Consent: Written informed consent was obtained from patients' parents who participated in this case.

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References

1. Schwartz PJ, Periti M, Malliani A. The long Q-T syndromes. *Am Heart J* 1975; 89: 378-90. [\[CrossRef\]](#)
2. Camm AJ, Janse MJ, Roden DM, Rosen MR, Cinca J, Cobbe SM. Congenital and acquired long QT syndrome. *Eur Heart J* 2000; 21: 1232-7. [\[CrossRef\]](#)
3. Moss AJ. Long QT syndrome. *JAMA* 2003; 289: 2041-4. [\[CrossRef\]](#)

4. Crotti L, Spazzolini C, Schwartz PJ, Shimizu W, Denjoy I, Schulze-Bahr E, et al. The common long-QT syndrome mutation KCNQ1/A341V causes unusually severe clinical manifestations in patients with different ethnic backgrounds: toward a mutation-risk stratification. *Circulation* 2007; 116: 2366-75. [\[CrossRef\]](#)
5. Fazio G, Vernuccio F, Grutta G, Re GL. Drugs to be avoided in patients with long QT syndrome: Focus on the anaesthesiological management. *World J Cardiol* 2013; 5: 87-93. [\[CrossRef\]](#)
6. Villain E, Levy M, Kachaner J, Garson A Jr. Prolonged QT interval in neonates: benign, transient or prolonged risk of sudden death. *Am Heart J* 1992; 124: 194-7. [\[CrossRef\]](#)
7. Yildirim H, Adanir T, Atay A, Katircioğlu K, Savaci S. The effects of sevoflurane, isoflurane and desflurane on QT interval of the ECG. *Eur J Anaesthesiol* 2004; 21: 566-70. [\[CrossRef\]](#)
8. Karagöz AH, Basgul E, Celiker V, Aypar U. The effect of inhalational anaesthetics on QTc interval. *Eur J Anaesthesiol* 2005; 22: 171-4. [\[CrossRef\]](#)
9. Owczuk R, Wujtewicz MA, Sawicka W, Lasek J, Wujtewicz M. The Influence of desflurane on QTc interval. *Anesth Analg* 2005; 101: 419-22. [\[CrossRef\]](#)
10. Paventi S. Effects of sevofluran versus propofol on QT interval. *Minerva Anesthesiol* 2001; 67: 637-40.
11. Kumar M, Baidya DK, Mohan VK, Mamta. Safe anesthesia management protocol of a child with congenital long QT syndrome and deafness (Jervell and lange-nielsen syndrome) for cochlear implant surgery. *Saudi J Anaesth* 2015; 9: 98-9. [\[CrossRef\]](#)
12. Kies SJ, Pabelick CM, Hurley HA, White RD, Ackerman MJ. Anesthesia for patients with congenital long QT syndrome. *Anesthesiology* 2005; 102: 204-10. [\[CrossRef\]](#)