

CASE REPORT

Anaesthetic Management of a Case of Long QT Syndrome Undergoing Laparoscopic Cholecystectomy

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ABSTRACT

Patients with Long QT Syndrome (LQTS) carry a very high risk of intraoperative malignant ventricular arrhythmias and are always a challenge to the anesthesiologist. We report An uneventful surgery under general anaesthesia in a patient with long QT syndrome (LQTS)- a 17 years old male posted for laparoscopic cholecystectomy for symptomatic cholelithiasis with intracardiac defibrillator (ICD) in situ. JMS: 2021; 24(4):40-45 DOI: <https://doi.org/10.33883/jms.v24i4.1118>

INTRODUCTION

The long QT syndrome (LQTS) is a disorder of cardiac ion channels producing prolonged ventricular repolarization, usually diagnosed by prolonged QT interval on electrocardiograph (ECG). The heart-rate-corrected QT intervals (QTc) varies by age and sex, with slightly longer values in females than in males.¹ Prolonged QTc is defined when the duration exceeds 450 ms in males and 460 ms in females.¹ In addition, increasingly prolonged QTc values portend a higher risk of potentially lethal arrhythmic events.^{2,3} To confirm the QTc, Leads II and V5 are optimally used.⁴ LQTS may be congenital (due to mutation of genes encoding cardiac ion channel) or acquired (caused by metabolic abnormalities or drugs). Congenital LQTS with associated deafness is known as Jervell and Lange-Nielsen syndrome.⁵ The absence of deafness with prolonged QT interval is called Romano-Ward syndrome.^{6,7} A patient with LQTS is at risk of ventricular tachyarrhythmia, especially torsade's de pointes.⁸ Most often, an episode of torsade's de pointes is self-terminating and may result in a syncopal episode due to the decrease in cerebral perfusion. Still, it may lead to sudden death when it deteriorates to ventricular fibrillation.⁹ The patients, whether treated or untreated,

remain at risk of life-threatening episodes of torsade's de pointes in the perioperative period. The anesthetic management of patients with LQTS, therefore, include immediate management of torsade's de pointes and, in known cases, avoidance of factors that may increase the risk of precipitating arrhythmias (torsade's de pointes). We report a 17-year-old male, a diagnosed case of long QT syndrome, posted for laparoscopic cholecystectomy.

CASE REPORT

A 17-year-old male was admitted to the Sher-I-Kashmir Institute of medical sciences (SKIMS) Srinagar, Kashmir, with the diagnosis of cholelithiasis. The patient has a significant medical history of multiple episodes of syncope at the age of 10 years, for which cardiac evaluation was done, and the patient was labelled as a case of long QT syndrome. Initial medical management was unsuccessful in preventing ventricular tachyarrhythmias and frequent syncopes. At the age of 11 years, a decision to place an in

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situ Intra Cardiac Defibrillator (ICD) was made, and the patient underwent the same successfully. Since then, the patient has also been continued with beta-blocker therapy. The patient reports infrequent episodes of palpitations and impending doom since the time ICD was placed but has not experienced any episodes of syncope, as such dysarrhythmic episodes have been taken care of by ICD. The patient has no other significant comorbidity. There was no history of hearing loss. The patient was planned for laparoscopic cholecystectomy under general anaesthesia. A pre-anaesthetic visit was made a day before surgery. The patient had the following vitals: Heart rate (HR): 64 bpm, Blood Pressure (BP): 110/70 mmHg, SpO₂: 98% on room air and in his usual state of health. General physical, systemic and airway examination was thoroughly carried out with no abnormal findings detected, as were laboratory results. The latest electrocardiogram showed a wide QT interval with a heart rate of 66 beats per minute (bpm) and R-R interval of 91 milliseconds (ms), with QTc of 480 ms. The latest ICD analysis was done on 15 February 2021, which revealed two episodes of ventricular tachycardia/ventricular fibrillation (VT/VF) @ 250 and 286 beats per minute (bpm) and received two shocks of 35 j each on 30 January 2021. Cardiology consultation was advised for risk stratification and ICD mode change prior to surgery. The patient was instructed to take the routine oral dose of beta-blocker in the morning on the day of surgery. The patient was taken to the pacing lab for mode change to asynchronous. All defibrillator settings were disabled, asynchronous pacing rate was set at 40 bpm. The patient was shifted to the preoperative holding area.

The patient was planned for induction with general anaesthesia. In the preoperative holding area, intravenous 2 mg of midazolam was used for anxiolysis and patient was transferred to the operating room on an oxygen face mask. All standard ASA monitors (ECG, pulse oximetry, NIBP) were applied. An 18 G intravenous (IV) cannula was secured on the dorsum of the left hand, and ringer lactate (RL) started. For monitoring beat to beat hemodynamics and arterial blood gas and serum electrolyte analysis, the left radial artery was cannulated and transduced. Pre-induction ABG analysis revealed - pH: 7.38 Na⁺: 136mmol/L Glu: 121 mg/dl, pO₂: 155 mmHg, pCO₂: 42 mmHg, K⁺: 3.7 mmol/L Lactate: 1mmol/L. HCO₃⁻: 24.8 Ca⁺: 0.87 Hct:pO₂: 116 Cl-112mmol/L. In addition to standard two-

lead ECG monitoring (Leads II and V), ECG was also monitored on an external defibrillator. External defibrillator pads were applied over the chest at standard locations. The patient was pre-medicated with injection (inj.) ranitidine 50mg IV and midazolam 1 mg IV before surgery. Pre-oxygenation was done with 100% O₂ for 3 minutes, anaesthesia was induced with inj. fentanyl 100mcg IV (to attenuate sympathetic response), inj. propofol 120 mg IV and tracheal intubation was facilitated with inj. rocuronium 50 mg IV. The patient's vocal cords were sprayed with 4-5 puffs of 10% lidocaine and trachea intubated with a cuffed endotracheal tube (7.5mm internal diameter (ID)). Tracheal intubation was confirmed with five-point auscultation and end-tidal capnography. The patient was connected to an anaesthesia workstation (Mindray WATO EX 65), and oxygen in the air (avoiding inhalation anaesthetics) was used to provide volume control ventilation (VCV) with VT 475 ml, rate 14 breaths per minute, I: E 1:2, PEEP 5 cm H₂O. Total intravenous anaesthesia (TIVA), with Propofol infusion @ 0.1 0.5 mg/kg/min, fentanyl 1 mcg/kg/hr infusion and inj. Rocuronium 5mg IV boluses every 20 minutes, was chosen to maintain anaesthesia and avoid unwanted cardiac effects of inhalational anaesthetics. For laparoscopy, pneumoperitoneum was created using low flow of CO₂ while the intraabdominal pressure (IAP) was strictly ensured to be kept in a lower acceptable working range (10-12mmHg). Total surgical time was 70 minutes, longer duration was ascribed to dense peri-gallbladder adhesions, while total anaesthesia time was 80 minutes. During Anaesthetic Management patient remained hemodynamically stable with heart rate remaining in the range of 60 to 75 bpm with no episode of any tachy/bradyarrhythmia. Intraoperative blood gas analysis on FiO₂ of 40% was performed with values; pH 7.42, pCO₂ 39 mmHg, pO₂ 210 mmHg, HCO₃⁻ 23 mmol/L, Ca²⁺ 0.75mmol/L, K⁺ 3.10 mmol/L, Hct 39%, Lactate 1.5 mmol/L. Hypocalcaemia was corrected with an infusion of Calcium gluconate 1gm IV over 15 minutes, while hypokalemia was corrected with Potassium chloride -15 mEq infused over 30 minutes. In view of hypokalemia, 1 gm of magnesium sulfate was also infused intravenously over 30 minutes. Dexamethasone (6mg IV), instead of 5HT₃ blockers, was used as an antiemetic. At the end of the surgery, propofol and fentanyl infusions were stopped, and the patient was extubated after reversal of neuromuscular

blockade with neostigmine 2.5 mg and glycopyrrolate 0.4 mg IV. Periods of reversal, extubation and immediate post-extubation were uneventful. The patient was fully conscious and comfortable in the immediate post-operative period. The patient was transferred to the post-anaesthesia care unit (PACU) for close observation. Arterial-line was kept in place for continuous hemodynamic monitoring in PACU. Meanwhile, the pacing laboratory was informed beforehand about the arrival of the patient to switch back the ICD settings to the pre-surgical mode.

DISCUSSION

Long QT syndrome (LQTS) is a abnormality of cardiac ion channels resulting either from mutations involving genes (encoding critical ion channels of the heart) or metabolic abnormalities/drugs and referred to as congenital and acquired LQTS, respectively.⁸ Clinically, it can present as syncope, seizures, or sudden cardiac death secondary to a characteristic polymorphic ventricular tachycardia known as torsade's de pointes (TdP) often precipitated by exertion or emotional stress.^{10,11} Certain drugs, including anaesthetic agents, are known to contribute to QT prolongation and thus can precipitate arrhythmias.¹² Long QT syndrome can be congenital or acquired; congenital LQTS was first described by Jervell and Lange-Nielsen in 1957, characterized by prolonged QT interval and congenital deafness.⁵ A similar syndrome with prolonged QT interval, syncope, and sudden death but without deafness was described by Romano et al. and Ward later.^{6,7} The age at presentation of patients with congenital LQTS is variable from early life to adulthood. The family history of sudden death due to cardiac conditions may be present. The frequency of these attacks varies from once or twice in a lifetime to once to twice per week.^{13, 14} As there is a significant risk of morbidity and mortality in patients with suspected or diagnosed LQTS due to cardiovascular compromise, anesthesiologists should be aware of necessary interventions and treatment modalities including the pharmacology of drugs that can cause QT prolongation.

Children, adolescents, and young adults presenting for surgery/anaesthesia with a history of syncope (including during exertion or auditory stimulation), epilepsy, or family history of sudden death should raise suspicion for c-LQTS and be evaluated preoperatively, including a 12 lead electrocardiogram. Increased physical activity, emotional

stress, anxiety, or fear can often trigger symptomatic events by amplified sympathetic outflow. So can also numerous pharmacological agents that prolong the QT interval. Management options include beta-blockers, implantable cardioverter-defibrillator therapy and left cardiac sympathetic denervation. The use of beta-blockers has effectively reduced mortality in patients with c-LQTS.¹⁵ A significant reduction in the mean rate of cardiac events has been shown with beta-blocker therapy in a study by Moss et al.¹⁶ Despite beta-blockade some patients continue to have symptomatic cardiac events Implantable cardioverter defibrillators (ICDs) have been used in this patient population to prevent sudden cardiac death.¹⁷ Left cardiac sympathetic denervation has a significant therapeutic effect in patients who continue to have arrhythmias despite therapy and have received multiple shocks from ICD.^{16, 18, 19} Torsade's de Pointes may arise at any point in the perioperative period and may be self-terminating and can be managed effectively with overdrive pacing and magnesium sulphate infusion. However, prolonged episodes may occur and can degenerate into ventricular fibrillation. Such episodes should be treated following the essential life support and advanced cardiovascular life support guidelines by the American Heart Association.²⁰ Available literature is scant, and there are no consensus guidelines for anaesthetic management for patients with LQTS. From the available literature, whether case reports, case series in patients with c-LQTS or studies in healthy patients, we will endeavour to draw a reasonable approach to anaesthesia for patients with congenital LQTS. Irrespective of anaesthetic modality, care must be taken to prevent stimulation of the sympathetic system as it can trigger TdP. Similarly, bradycardia, tachycardia, hypoxemia, hypertension, light anaesthesia and hypocapnia or hypercapnia must be avoided. Drugs known to increase the QT interval duration should also be avoided in the perioperative period whenever possible. Patients receiving b-blockers should continue their b-blockers through the perioperative period.²¹ Premedication with midazolam will provide anxiolysis and thus prevent sympathetic stimulation; midazolam does not produce QT prolongation.^{22, 23} We used total intravenous anaesthesia with propofol and fentanyl in our patient. Studies have shown Propofol to have slight or no effect on the QT interval in healthy patients undergoing minor surgery, as shown by some studies.^{24, 25} Aswell propofol can

sometimes rapidly reverse sevoflurane-induced QTc prolongation in healthy patients.²⁶ We avoided the use of volatile agents as all volatile agents are known to prolong QT interval,¹² although there are studies that demonstrated the safe use of these agents in patients with c-LQTS receiving beta-blocker therapy.^{27,28,29,30} Rocuronium provides more suitable conditions for rapid tracheal intubation than atracurium, is associated with a shorter clinical duration, has a more negligible effect on mean arterial blood pressure and heart rate, and has less histamine-releasing potency when compared to atracurium, for these advantages we preferred rocuronium in our patient.³¹ In our case, we chose fentanyl to avoid sympathetic stimulation during laryngoscopy and tracheal intubation, although the effects of fentanyl on the QT interval are conflicting.^{15,23} As there are studies showing prolongation of the QT interval due to interaction between 5-HT₃ receptor antagonist and different human cardiac ion channels and also ondansetron being recorded as a medication with “possible” risk for TdP in patients with c-LQTS, we avoided 5-HT receptor antagonist in our patient.^{32,33} To minimize sympathetic stimulation and systemic absorption of carbon dioxide while creating and maintaining pneumoperitoneum, IAP was strictly maintained between 10-12 mmHg without compromising the laparoscopic vision and working field.³⁴

CONCLUSION

Patients having LQTS are at higher risk of perioperative morbidity and mortality. Anaesthesiologists taking care of patients with LQTS should be aware of necessary interventions and treatment modalities for the management of fatal arrhythmias, including TdP. Care must be taken to prevent emotional stress and anxiety related to surgery to avoid sympathetic overactivity. Detailed history taking and preoperative management of any electrolyte imbalances are indispensable.

External defibrillator pads should be kept attached and ready to use in case an arrhythmic event occurs.

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