



General Anaesthesia for Cesarean Section in a Parturient with Long QT Syndrome: A Case Report and a Review of Literature

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Authors' contributions

This work was carried out in collaboration between all authors. Authors NK and BK wrote the first draft of the manuscript. Authors NK, BK and UKI managed the anesthesia period of patient during the operation. All authors read and approved the final manuscript.

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Case Study

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ABSTRACT

Long QT syndrome patients are at high risk of developing ventricular arrhythmia and cardiac arrest, so that the anesthetic technique used for these patients must avoid anything that will induce an arrhythmia such as tachycardia, hypotension or increased catecholamine release by pain or stress.

A 28 -yr-old woman was scheduled for an elective, repeat cesarean section at 36 weeks gestation. She was diagnosed long QT syndrome at age 22 and an automatic implantable cardiac defibrillator (AICD) was implanted. During her pregnancy, parturient was hospitalized at 35 weeks gestation because of fetal bradycardia and obstetrician scheduled cesarean section at 36 weeks gestation. Before induction of anaesthesia, esmolol 200mcg.kg.min⁻¹ was started for prevention of ventricular dysrhythmia during laryngoscopy and tracheal intubation. After preoxygenation, anaesthesia was induced with fentanyl 100mcg, propofol 200mg, rocuronium 100 mg and trachea was intubated at 45th seconds. Esmolol infusion rate was reduced gradually to parturient's

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haemodynamic parameters during surgery and was stopped at end of the surgery. At 4th minutes of the surgery, fetus was delivered but there is no heart rate and breathing of baby. Following cardiac compression for 45 seconds, heart rate and breathing of baby returned. Anaesthesia was maintained with 1 MAC sevoflurane and 100 mcg fentanyl. Parturient's blood pressure and heart rate remained within normal limits during surgery. Consequently, if parturient does not accept regional anaesthesia, in case of an emergency cesarean section, general anaesthesia can be safely used with optimized preoperative evaluation, close monitoring and carefully anaesthetic management.

Keywords: Long QT syndrome; anesthesia.

1. INTRODUCTION

The long QT syndrome, is a rare disorder with a prevalence of 1:1100-3000. This condition is thought to be secondary to mutations in the genes encoding the ion channels in the heart [1]. These patients are predisposed to polymorphic ventricular tachycardia known as Torsades de Pointes (TdP). The principal danger of TdP is deterioration into pulseless ventricular tachycardia or ventricular fibrillation and death [1-4]. Surgical stress, anxiety, inadequate anaesthesia, bradycardia, tachycardia, hypo or hypercarbia, hypothermia, inadequate analgesia may cause sympathetic stimulation increasing the risk of TdP [2,3]. Long QT syndrome patients are at high risk of developing ventricular arrhythmia and cardiac arrest, so that the anesthetic technique used for these patients must avoid anything that will induce an arrhythmia such as tachycardia, hypotension or increased catecholamine release by pain or stresses [1,5].

The cardiovascular changes of pregnancy can cause clinical decompensation in patients with structural heart disease, but little is known about the effect of pregnancy on patients with cardiac rhythm abnormalities [6,7]. The physiological increase in heart rate may be protective during pregnancy in long QT syndrome women who have an increased QT interval at slower heart rates [8], but postpartum the QT interval may lengthen as the heart rate slows, increasing the risk of a cardiac event [6].

In this case report, we aimed to inform that our general anaesthesia experience in a parturient with a long QT syndrome and an automatic cardiac defibrillator (AICD).

2. CASE REPORT

A 28 -yr-old woman, gravida 2, parite 1 was scheduled for an elective, repeat cesarean section at 36 weeks gestation. In her past medical, she

was diagnosed long QT syndrome at age 22 because of repeated syncopes and an automatic implantable cardiac defibrillator (AICD) was implanted. The information about her AICD is as follows: Medtronic GEM 7227CX (pacer/defibrillator); Medtronic 6936; AICD pacemaker mode VVI. She had no any syncope in the last year. Spinal anaesthesia was administered for cesarean section in her first pregnancy. Intra and postoperative courses were uneventful in the first delivery. Her physical examination revealed a fit looking woman with normal airway and spine. Complete blood count, electrolytes and coagulation profile were within normal limits. An ECG indicated a heart rate of 63 per minute and a corrected QT interval (QTc) was noted as 440msec. During her second pregnancy, parturient was hospitalized at 35 weeks gestation because of fetal bradycardia (FHR: 100 beats.min) and obstetrician scheduled cesarean section at 36 weeks gestation. In the preoperative assessment, the cardiologist was contacted for information about patient and AICD. Her medication consisted of metoprolol tablet 50 mg 2x1 per day. The information obtained from cardiologist was that the program of the AICD will not be affected if surgery area be far. Also, cardiologist recommended bipolar cother using and duration of cotherization should be limited to 10 seconds.

We used general anaesthesia because the patient refused regional anaesthesia. In the operating room, routine electrocardiogram, non invasive blood pressure and peripheric oxygen saturation monitoring were applied and baseline values were recorded (heart rate: 65beats.min, blood pressure: 140/70 mmHg, and SpO₂: 100%). Before induction of anaesthesia, esmolol 200 mcg.kg.min⁻¹ was started for prevention of ventricular dysrhythmia during laryngoscopy and tracheal intubation and magnesium sulfate was only kept ready. After preoxygenation, anaesthesia was induced with fentanyl 100 mcg, propofol 200 mg, and rocuronium 100 mg and trachea was intubated at 45th seconds.

Haemodynamic parameters after intubation were within normal limits (BP:104/64mmHg, HR:64beats.min and SpO₂: 100). Anaesthesia was maintained with %100 oxygen and 0.8 MAC sevoflurane until delivery of baby. After delivery of baby, oxygen concentration was decreased to 50% and anaesthesia was maintained with 1 MAC sevoflurane, 100 mcg fentanyl. After removal of the placenta, oxytocin infusion was started. Esmolol infusion rate was reduced gradually to parturient's haemodynamic parameters during surgery and was stopped at end of the surgery. For antibiotic prophylaxis, cefazoline 2gr was administered.

At 4th minutes of the surgery, fetus was delivered but there is no heart rate and breathing of baby. Following cardiac compression for 45 seconds, heart rate and breathing of baby returned. Parturient's blood pressure and heart rate remained within normal limits during surgery. At the end of surgery, baby was transported to neonatal intensive care unit and mother was transported anaesthesia intensive care unit. Mother was extubated at 3rd hour and monitored for 24 hr postoperatively. The later assessment of neonate, LQTS was diagnosed by pediatric cardiology.

3. DISCUSSION

The physiological changes of pregnancy produce an increase in cardiac output and heart rate, increasing the risk of dysrhythmias in normal healthy women. The pregnant women with long QT syndrome are more susceptible to ventricular dysrhythmias during pregnancy, labour and delivery. The long QT syndrome is a disease of young people with significant morbidity and mortality, making the management of pregnant long QT syndrome patients challenging [5].

The anaesthetic management of these patients with long QT syndrome starts with assessment, focusing on the patient's history, medication use and any other pertinent details, such as pacemaker or AICD. A cardiology consultation is advisable and the patient should be advised to continue medication such as beta-blockers, including on the day of surgery. Hypokalemia, hypomagnesemia, and hypocalcemia should be corrected preoperatively and drugs that prolong QT interval should be avoided in the preoperative period [1].

During the intraoperative period, the goal is to prevent and treat any dysrhythmia that can result

from prolonged QT interval. Prolonged QT interval can be precipitated by drugs (thiopentone, succinylcholine, epinephrine, and norepinephrine), electrolyte disturbances, hypothermia and any cause of sympathetic stimulation [9]. An external pacemaker and defibrillator must be available in the operation room before induction of anaesthesia. Any episode of TdP should be treated by cardioversion/defibrillation and magnesium sulphate 30 mg.kg⁻¹ iv over two to three minutes followed by an infusion of 2 to 4 mg.min⁻¹ [10].

Intravenous magnesium is the agent that may be used for the immediate treatment of TdP for long QT syndrome [1,2]. So that magnesium sulfate was only kept ready for possibility of arrhythmia but we did not need to use it.

The anaesthesiologist should be cautious during induction of general anaesthesia. The risk of dysrhythmia is increased due to sympathetic stimulation and increased catecholamine release during intubation and skin incision. Although halothane, enflurane, isoflurane and sevoflurane have been used as a component of uneventful anaesthesia in known long QT syndrome patients who were beta-blocked [1] these drugs prolong the QT interval in normal people when used as the sole induction or maintenance agent [11,12]. Midazolam does not affect QT interval [3]. Thiopental prolongs whereas propofol has been found to have little or no effect on the QT interval and may even reverse sevoflurane induced QT prolongation [1,3]. No inhalational agent is completely safe in long QT syndrome as all can pro0, long the QTc. However, isoflurane is inherently safe and has been reported to shorten QT interval in a patient with long QTc syndrome and therefore is regarded as the agent of choice [3]. In several case reports, involving pregnant [13] and non-pregnant patients, volatile anaesthetic agents have been used uneventfully [3], while in other reports ventricular dysrhythmias [14] and torsades [15] occurred in beta blocked long LQT syndrome patients. Sevoflurane may prolong QTc interval in healthy patients but there is a little information about the concentration-response relationship of the effect of sevoflurane on the QTc interval. In a study, authors informed that among patient receiving sevoflurane, QTc interval changes correlate to anaesthetic level of sevoflurane [16]. We do not have isoflurane in our hospital. So that we used low concentration of sevoflurane (1 MAC) and high dose fentanyl for maintenance of general anaesthesia to avoid the effect on QTc.

Some authors suggest that total intravenous anesthesia with propofol, rather than the use of inhalational agents, may reduce the incidence of ventricular dysrhythmias [17].

In a case report, thiopental, rocuronium and remifentanyl were used for induction of anaesthesia and isoflurane was used for maintenance of general anaesthesia [13]. In another case report, diazepam, thiopental, succinylcholine and halothane were used [18]. There were no dysrhythmias in either case.

Vecuronium and atracurium have been found to have no effect on the QT interval. Fentanyl and morphine have been used without adverse effects in patients with long QTc syndrome [3]. The use of anticholinergic and anticholinesterase has been shown to prolong QT interval in healthy subjects and hence should be used with caution [1,3]. Inadequate plane of anaesthesia, bradycardia or tachycardia, hypertension, hypoxi, hypo or hypercarbia, inadequate analgesia, hypothermia may cause sympathetic stimulation increasing the risk of TdP [3,5].

Long QT syndrome parturients should be thoroughly assessed by an anesthesiologist during their pregnancy as cesarean delivery may be required on an emergent basis. In the untreated, anesthetized, long QT syndrome patient, ventricular dysrhythmias may be refractory to treatment [1,2].

Beta blockers are the mainstay of drug therapy in patients with long QT syndrome and should be continued in the perioperative period. There is no evidence that beta-blockers are teratogenic [8] but fetal effects include intra-uterine growth restriction, bradycardia, hypoglycemia and premature uterine contractions [6]. The risk of dysrhythmia in a pregnant woman with long QT syndrome outweighs any risk to the fetus or newborn of beta blocker therapy [6]. The fetus and neonate are at risk for inheriting LQTS. As there are reports of neonatal long QT syndrome, the neonate of patients with long QT syndrome should have ECG screening at birth [1,19]. The baby of our patient also received a diagnosis of long QT syndrome in the postpartum follow-up. In our case, beta blocker was started during pregnancy and we give esmolol before induction of anaesthesia and continued during the operation with gradually decreasing doses.

In a multicenter retrospective chart review of perioperative management of children with

clinically diagnosed long QT syndrome aged 18 years or younger, who received general anaesthesia, authors suggest that the risk of perioperative TdP is concentrated in neonates and infants requiring urgent interventions after failed first-line management of long QT syndrome [20].

Regional analgesia/anaesthesia is advantageous for long QT syndrome parturients as reduction of the stress response and provision of effective analgesia moderates catecholamine release, reducing the risk of torsades. The disadvantages of regional anaesthesia in long QT syndrome is the potential for a high block, causing hypotension and bradycardia-induced parasympathetic override [5]. Most long QT syndrome parturients described in case reports received neuroaxial anaesthesia for labour analgesia or surgical delivery [19,21-24].

It has been suggested that regional anaesthesia may be more effective than general anaesthesia in reducing sympathetic activity in non-long QT syndrome parturients with severe preeclampsia [25]. There are significant changes in the QTc level at one, five and 15 min after induction of spinal anaesthesia in normal healthy males and in some, the QTc was ≥ 440 msec. This prolongation was thought to be secondary to hypotension as the heart rate did not change [26]. In a study comparing preeclamptic patients (not long QT syndrome parturients) with healthy controls, the QTc was prolonged prior to induction of spinal anaesthesia in the preeclamptic group [27]. When the spinal anaesthesia was administered the QTc normalized in the preeclamptic group and remained stable in the control group. The prolonged QTc at baseline was possibly secondary to hypocalcemia and hypertension, or to autonomic imbalance and sympathetic over activity [27]. Because the patient refused regional anaesthesia, we used general anaesthesia. She was very stressful and wanted to have general anaesthesia.

The major advantage of regional anaesthesia is the reduction in catecholamine levels during surgery. Because of the potential for rapid onset hypotension which could lead to a ventricular arrhythmia and cardiac arrest, most anaesthesiologists have avoided spinal anaesthesia in patients with long QT syndrome [5]. Al-Rafai and colleagues applied spinal anaesthesia safely in a parturient with long QT syndrome and an AICD for cesarean section [5].

Epidural anaesthesia has been used for cesarean delivery in a parturient with long QT syndrome [22] and spinal anaesthesia has been reported in a non-pregnant patient [28]. The postoperative course should include recovery in a monitored setting for 24hr. If the patient has an AICD or a pacemaker, the device must be interrogated by the cardiologist to recheck the program once surgery is finished [5].

Prevention of pain, anxiety, hypoxia, hypercarbia, hypothermia, shivering and hypo/hyperglycemia are important management principles [1,9,29]. High positive airway pressure may increase the QTc [30] and hypothermia prolongs the QT interval [29]. If the patient has an AICD or pacemaker, a full history about the device must be documented including the indication for insertion (if other than long QT syndrome), the type of device, the information and the date of last interrogation, the date of last battery change, any symptoms that indicate malfunction and the response to a magnet [31]. In patients with a pacemaker or AICD, the usual intraoperative precautions should be taken to avoid disruption of function. This includes using bipolar electrocautery, instead of monopolar, and placing the current return pad as far as possible from the device. Mangar reported a pacemaker failure intraoperatively when a magnet was placed on the pacemaker with the use of monopolar electrocautery [10].

General anaesthesia for the long QT syndrome parturient can be challenging due to the risk of prolonging the QT interval and precipitating torsades [1]. The anesthesiologist should aim to avoid the sudden release of catecholamines which can precipitate torsades de pointes. Anxiolysis may be beneficial [1,32], and in healthy patients the QT interval is not prolonged nor are dysrhythmias induced by midazolam alone [33] or in combination with fentanyl [34]. By administering additional beta blockers and /or opioids, or by topical anesthesia to the vocal cords or intravenous lidocaine, catecholamine release can be reduced during laryngoscopy, tracheal intubation and extubation [35]. Esmolol is thought to be problematic in parturient as its administration has been associated with fetal bradycardia and acidosis [36], but in the long QT syndrome parturient, the risk to the fetus may be justified. Low lipid solubility and rapid metabolism of esmolol may have limited placental transfer, which may confer an advantage over long established beta blocking agents whose use during pregnancy and labour has been limited by concern over fetal effects [37]. Eisenach and

Castro found that brief maternal infusion of esmolol did not change fetal blood pressure and produced only minor decrease in fetal heart rate [38]. Because of rapid metabolism and limited placental transfer, we used esmolol to avoid sudden release of catecholamine during laryngoscopy and tracheal intubation. The risk of dysrhythmia in a pregnant woman with long QT syndrome outweighs any risk to the fetus or newborn of beta blocker therapy [6].

Many drugs prolong the QT interval, so that these potential triggers should be avoided perioperatively, if possible. Epinephrine increases the QTc in healthy and long QT syndrome patients [9,11]. Phenylephrine has been used successfully in a long QT syndrome patient who developed hypotension following a CSE for Cesarean delivery [5]. There are no published data on the effects of ephedrine on the QTc in patients with long QT syndrome due to its sympathomimetic effects. Dobutamine and dopamine are relatively contraindicated in long QT syndrome [3]. Oxytocin is a potentially dysrhythmogenic, especially in patients with long QT syndrome [39]. However, oxytocin has been used for induction of labour in long QT women without effect [13].

In a case report, the patient with no medical history, an episode of TdP occurred in recovery room. The ECG after cardiopulmonary resuscitation showed a QTc interval prolongation, whereas it was normalized after 48 hours. Authors reported that a ventricular fibrillation occurred after a TdP, due to drug-induced long QT syndrome during general anaesthesia with probably drug interaction [40].

4. CONCLUSION

Parturients with long QT syndrome need a multidisciplinary approach for their management and care throughout pregnancy and delivery. Anaesthetic management of these patients aims to prevent prolongation of the QTc and anesthesiologist must be prepared to treat immediately any episode of torsades. If parturient does not accept regional anaesthesia, in case of an emergency cesarean section, general anaesthesia can be safely used with optimized preoperative evaluation, close monitoring and carefully anaesthetic management.

CONSENT

As per international standard or university standard written patient consent has been collected and preserved by the authors.

ETHICAL APPROVAL

All authors hereby declare that all experiments have been examined and approved by the appropriate ethics committee and have therefore been performed in accordance with the ethical standards laid down in the 1964 Declaration of Helsinki.

COMPETING INTERESTS

Authors have declared that no competing interests exist.

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