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Anesthesia in Long QT syndrome: A review of pathophysiology, risk factors, and management strategies

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Abstract Long QT syndrome (LQTS) poses significant challenges in anesthetic management due to its potential to provoke life-threatening arrhythmias such as Torsades de Pointes, which can lead to sudden cardiac death if not properly managed. This review examines the pathophysiology of LQTS, its classification into congenital and acquired forms, and the associated anesthetic risks, while providing evidence-based strategies for its safe and effective management. Congenital LQTS is subdivided into LQT1, LQT2, and LQT3, each associated with distinct genetic mutations, ion channel dysfunctions, and specific triggers, whereas acquired LQTS arises from factors such as drug-induced effects, electrolyte imbalances, and underlying medical conditions. Accurate diagnosis hinges on a thorough clinical evaluation, electrocardiogram findings, and, where applicable, genetic testing to guide individualized risk stratification and tailored management plans. Anesthetic management in LQTS demands meticulous planning, vigilance, and a comprehensive understanding of the syndrome's complexities. Preoperative strategies focus on optimizing electrolyte levels, discontinuing QT-prolonging medications, and assessing genetic predispositions that may exacerbate risks. The intraoperative phase requires continuous ECG monitoring to detect QT prolongation, the judicious selection of anesthetic drugs to minimize arrhythmogenic potential, and readiness to address arrhythmic events with therapies such as intravenous magnesium sulfate and immediate defibrillation. Postoperative care emphasizes extended monitoring, strict maintenance of electrolyte balance, and the avoidance of QT-prolonging agents to reduce the risk of delayed complications. This review underscores the importance of a multidisciplinary approach, involving anesthesiologists, cardiologists, and geneticists, in delivering optimal care to patients with LQTS. By focusing on detailed preoperative assessment, careful drug selection, and proactive intra- and postoperative management, clinicians can effectively minimize risks and enhance patient safety. These recommendations provide a robust framework for navigating the challenges of anesthesia in individuals with LQTS.

Keywords: torsades de pointes, arrhythmias, congenital LQTS, genetic mutations, ECG monitoring, drug-induced LQTS

1. Introduction

Long QT syndrome (LQTS) is a cardiac disorder that prolongs the QT interval on the electrocardiogram, increasing the risk for possibly life-threatening arrhythmias such as Torsades de Pointes (TDP). The QT interval measures the time taken for ventricular depolarization and repolarization. With LQTS, this interval becomes prolonged owing to abnormalities in cardiac ion channels responsible for cardiac repolarization (Moss, 2005). The characteristic defect in LQTS is the impairment of normal ion channel function in that it inhibits the ability of the heart to repolarize after each beat properly. Delayed repolarization can prolong the QT interval. Prolongation of this period increases susceptibility to TDP, a polymorphic VT that can trigger syncope or sudden cardiac death (SCD) (Lima et al., 2023). The genetic forms of LQTS are commonly known as congenital LQTS and result from genetic mutations that lead to abnormalities in ion channels or related proteins in these patients. Such forms can be further divided on the basis of the ion channel they affect. On the other hand, the acquired forms result from exogenous factors or underlying conditions that prolong the QT interval and include some medications, electrolyte imbalance, and an underlying medical disorder (Ackerman, 1998).

The management of LQTS, particularly in the perioperative period, is challenging due to the potential for arrhythmias to be triggered by anesthesia-related factors such as certain medications, sympathetic stimulation, or electrolyte disturbances. Identifying patients at risk is crucial, as undiagnosed or poorly managed LQTS can lead to catastrophic outcomes, including cardiac arrest. Advances in genetic testing have enabled more accurate identification of congenital LQTS, helping to stratify patients according to their specific risk profiles. Nevertheless, the acquired forms of LQTS often remain underdiagnosed, highlighting the need for clinicians to have a high index of suspicion in the presence of QT-prolonging factors (Moss, 2005).

Given its implications for anesthesia and perioperative care, LQTS demands a comprehensive understanding of its pathophysiology, diagnosis, and management strategies. Anesthetic considerations include the selection of drugs with minimal

QT-prolonging effects, continuous ECG monitoring, and readiness to address arrhythmic events promptly. A multidisciplinary approach involving anesthesiologists, cardiologists, and geneticists is often essential for optimizing outcomes. This review aims to provide a detailed exploration of these strategies, emphasizing the importance of individualized care for patients with LQTS undergoing surgery (Wallace et al., 2019).

2. Congenital long QT syndrome

Several genetic subtypes are LQTSs, each associated with specific ion channel mutations. The most common among them is LQT1, which is caused by mutations in the gene KCNQ1, affects the potassium channel KV7.1 and is usually exercise inducible with peak incidence swimming. LQT2, caused by mutations in the gene KCNH2 and affecting the potassium channel KV11.1, may be induced by loud noises or stress. LQT3 is caused by mutations in SCN5A, affecting the voltage-gated NaV1.5 sodium channel, and may be exercise inducible, rest inducible, or sleep inducible. Mutations in ANK2 cause LQT4; its role in QT prolongation is under debate. LQT5 results from mutations in KCNE1, which affects the β-subunit of the potassium channel, causing either Romano–Ward syndrome or Jervell–Lange–Nielsen syndrome. LQT6 is caused by mutations in KCNE2, which codes for another β-subunit, although its function is not without controversy. LQT7 is caused by mutations in KCNJ2, which codes for the potassium channel Kir2.1, and is also associated with Andersen–Tawil syndrome. LQT8 is caused by mutations in CACNA1C, encoding the calcium channel CaV1.2, which underlies Timothy syndrome. LQT9 is caused by mutations in CAV3, which codes for caveolae and affects the sodium current. LQT10 has been associated with mutations in SCN4B, which involve the sodium channel β4-subunit. LQT11 is caused by mutations in AKAP9, which interacts with KV7.1. LQT12 is caused by mutations in SNTA1 and affects the regulation of the sodium current. Mutations in KCNJ5 cause LQT13 and affect potassium channels. LQT14, LQT15, and LQT16 are caused by mutations in CALM1, CALM2, and CALM3, respectively, each affecting calmodulin proteins that interact with calcium channels (Modell & Lehmann, 2006; Wallace et al., 2019).

3. Acquired Long QT Syndrome

Drug-induced LQTS is an acquired form of LQTS that occurs due to the use of certain medications, which include those known to prolong the QT interval by affecting cardiac ion channels. These channels play a role in the repolarization phase of the cardiac action potential. Perturbations in the function of ion channels can generate potentially life-threatening arrhythmias, such as TDP. Antiarrhythmic drugs themselves often have the unexpected result of increasing the risk for or even causing arrhythmias by prolonging the QT interval. This appears to be particularly true for Class IA antiarrhythmics, which include quinidine and procainamide, and Class III antiarrhythmics, which include amiodarone and sotalol. Each of these medications depolarizes the heart muscle by blocking potassium channels, thereby delaying repolarization and thus lengthening the QT interval. Antipsychotic medications, particularly first-generation, typical antipsychotics such as haloperidol and thioridazine, are among the most significant risks for QT prolongation. These drugs block various ion channels in the heart, particularly the hERG potassium channels, which significantly participate in the repolarization process. This risk is also associated with second-generation atypical antipsychotics, specifically ziprasidone, although the risk generally is lower than that associated with first-generation antipsychotics. Certain classes of antibiotics, most notably macrolides, are known to prolong the QT interval. These antibiotics either directly act on cardiac ion channels or interact with other medications, which results in QT prolongation and increases the risk of developing an arrhythmia. The most significant risk is in those patients with preexisting electrolyte imbalance, inborn LQTS, or other predisposing conditions. Other drugs that have also been linked to the prolongation of the QT interval include some antidepressant drugs, antiemetic drugs, antifungal drugs, etc. These drugs are known to cause QT prolongation mainly at relatively high doses because of drug interactions through the inhibition of metabolic pathways and in people with other risk factors or conditions, such as electrolyte imbalance and cardiac diseases (Kannankeril et al., 2010).

Electrolyte imbalances, particularly potassium, calcium, and magnesium imbalances, are central to the pathogenesis of LQTS. These electrolytes maintain regular electrical activity in the heart, and their depletion may disrupt the delicate balance required for proper cardiac repolarization. Hypokalemia, or low potassium levels, has an essential function during the repolarization phase of the cardiac action potential. When there is a low level of potassium, this repolarization process slows down, eventually prolonging the QT interval. Hypokalemia increases the sensitivity of the myocardium to certain medications, which extends the QT interval, further aggravating the risk of arrhythmias. It does so to the plateau phase of the cardiac action potential. Low calcium levels can increase the duration of the action potential and thus prolong the QT interval. Although hypocalcemia is less commonly implicated in QT prolongation than hypokalemia is, it can still be a contributing factor, especially in the presence of other predisposing factors. Hypomagnesemia, or low levels of magnesium, contributes to the stabilization of cell membranes and the regulation of ion channels. The risk of QT prolongation is increased with magnesium deficiency, especially when individuals present with hypokalemia. Deficiency of magnesium is common in critically ill patients and is one of the treatable etiologies for QT prolongation (Antzelevitch & Burashnikov, 2011).

Some acquired forms of LQTS occur because they directly or indirectly affect cardiac electrical activity as a consequence of an underlying medical condition. Myocarditis is an inflammation of the cardiac muscle that has been described to lead to QT prolongation, probably through the disturbance of normal heart electrical activity. The process of inflammation likely influences

ion channels and makes the myocardium more arrhythmogenic. A slow heart rate or bradycardia can also lead to QT prolongation, as the phase of cardiac cycle repolarization is prolonged by a declining heart rate. This can be particularly problematic in patients with other risk factors for LQTS, such as those on medications that prolong the QT interval. Other conditions that may lead to an electrolyte imbalance and thus affect the heart's electrical activity include endocrine disorders such as hypothyroidism, hyperparathyroidism, and adrenal insufficiency. For example, hypothyroidism is associated with both bradycardia and QT prolongation. A disease such as hyperparathyroidism can result in hypercalcemia and thus delay repolarization by also prolonging the QT interval. Cardiac structural abnormalities, including left ventricular hypertrophy or heart failure, can also be associated with QT prolongation because of alterations in the architecture of the myocardium and the distribution of ion channels, increasing the subsequent risk of arrhythmias (Lazzerini et al., 2015).

4. Diagnosis and Risk Assessment of Long QT Syndrome

A proper diagnosis of LQTS must be established by considering the combination of clinical assessment, electrocardiographic (ECG) patterns, and genetic testing outcomes. The diagnosis of LQTS is confirmed by EKG, which measures the QT interval. Corrected for heart rate (QTc) via the Bazett or Fridericia correction formulas, the QT interval is prolonged in both males if it is > 450 ms and females if it is > 460 ms, which is suggestive of likely LQTS. The presence of TDP on an EKG can be used to make a diagnosis of LQTS, particularly in symptomatic patients. In general, type-specific EKG findings are characteristic of the different forms of LQTS. In particular, each form of the syndrome has typical patterns and triggers. LQT1 generally shows findings that are most evident during exercise, more so with exercise stress testing, because physical activity can increase the characteristic prolonged QT interval. In LQT2, the EKG may present with TDP, whose onset is precipitated by acoustic stimuli, reflecting increased sensitivity of the electrical system of the heart to loud noises. In contrast, LQT3 typically presents with a prolonged QT interval even at rest, and the episodes often occur during sleep, suggesting a different mechanism that affects the electrical activity of the heart, mainly when the heart rate is low. These type-specific patterns are, therefore, important in the establishment of proper diagnosis and guiding treatment for LQTS (Johnson & Ackerman, 2009). Genetic testing is of utmost importance in establishing the diagnosis of congenital LQTS and in recognizing some gene mutations. This is important, especially in patients with a history of LQTS in the family or unexplained syncope. Genetic testing of some mutations in specific genes, such as KCNQ1, KCNH2, SCN5A, and KCNJ2, can establish a definite diagnosis that can guide risk stratification and management (Aiba, 2019).

5. Clinical Risk Factors

An integrated approach to clinical evaluation, ECG findings, genetic testing, and monitoring is needed for a comprehensive diagnosis of LQTS with risk assessment. History A history of syncope, particularly during effort or emotional stress, suggests an increased risk of arrhythmias and SCD. On the other hand, a family history of SCD or LQTS, especially if this happens at a young age or involves severe cases, further increases this risk. Symptoms such as palpitations, dizziness, or syncope mandate a search for the presence of severe arrhythmias. ECG findings are essential; a prolonged QTc duration is correlated with an increased risk of arrhythmia, and a fragmented QRS complex acts as a marker of increased arrhythmic risk. Genetic factors, type, and mutation have strong interactions that significantly influence risk stratification; for example, LQT3 patients have a higher SCD risk than LQT1 or LQT2 patients do because of a mutation in SCN5A. The appreciation of genotype–phenotype correlations permits individualized risk management. This also includes extended investigations—the exercise test in risk stratification for the occurrence of exercise-induced arrhythmias in LQT1 patients and Holter monitoring for the detection of concealed arrhythmic events not expressed on a regular ECG. These factors are all summed for risk stratification in high-risk individuals among patients with LQTS, particularly those scheduled for treatment decisions against lethal arrhythmias and SCD (Saenen & Vrints, 2008).

6. Pharmacological Considerations in Anesthesia

In patients with LQTS, the effects of anesthetic drugs on the QT interval and general cardiac stability are crucial because they can considerably increase the danger of arrhythmias during surgical procedures. Therefore, their impacts must be understood to safeguard patients and institute the required measures for precaution. Halothane is a common inhalational anesthetic that has the potential to prolong the QT interval. Nevertheless, its clinical relevance in patients with LQTS is not as clearly established as that of other agents. Isoflurane and sevoflurane generally have very minimal effects on the QT; however, they may interact with medications that alter cardiac repolarization, hence mandating careful use in LQTS patients. Another inhalational anesthetic agent is desflurane, which by itself is unlikely to prolong the QT interval. Nevertheless, it may interact with other drugs that affect cardiac conduction and thus needs careful monitoring (Saito et al., 2021). The intravenous anesthetics thiopental and propofol have very minor direct effects on QT prolongation. Nevertheless, propofol is a very well-recognized cause of hypotension, and through this action, it can exert indirect effects on cardiac stability, making it a possible factor in LQTS management. Etomidate has minimal cardiovascular effects, making it safer in terms of the QT interval and general cardiac stability (Saarnivaara et al., 1993). Succinylcholine and nondepolarizing neuromuscular blockers are generally

safe with respect to QT prolongation. Nevertheless, they may interact with other medications that affect cardiac activity and, therefore, must be managed to avoid adverse effects. Opioids, such as morphine, fentanyl, and sufentanil, may depress cardiac function directly or indirectly, depressing cardiac function in the QT interval. Thus, the use of such agents must be carried out very carefully in patients with preexisting cardiac disease to minimize the risk of arrhythmias (Niimi et al., 2022). Many antidepressant and antipsychotic drugs prolong the QT interval. Interactions with anesthetics, which themselves have an independent effect on cardiac function, increase the risk of developing arrhythmias. Adjustments in anesthetic management and close monitoring of cardiac function are essential for mitigation. Macrolides and fluoroquinolones are classes of antibiotics that prolong the QT interval. The additive effect of their interaction with anesthetic agents could further increase the potential risk of developing arrhythmias. One can reduce the risks by using antibiotics that have fewer QT-prolonging effects or monitoring more closely during anesthetic procedures (Aroke & Nkemazeh, 2020). More commonly, however, the interaction between anesthetic drugs and patients' regular medications needs to be considered. Antiarrhythmic medicines, especially Class IA and Class III agents such as quinidine, sotalol, and dofetilide, lengthen the QT interval. When combined with anesthetics that either depress cardiac conduction or delay repolarization, the QT interval may be significantly prolonged, increasing the risk of TDP; thus, such patients need to be monitored carefully, and combination with interacting drugs should be avoided (Ayad et al., 2010).

7. Preoperative Assessment and Management

In the case of a diagnosed LQTS patient, preoperative screening is the most critical step toward a safe outcome from anesthesia and surgery. This is done to evaluate the cardiac condition of the patient in detail, calculate the risk, and plan proper management. A detailed history forms the essential background for screening. A thorough history of symptoms of syncope, palpitations, and episodes of dizziness must be taken and investigated adequately since they might yield signs and symptoms of the underlying arrhythmias of LQTS. The medication history is mandatory; it has to stress antiarrhythmics, psychotropics, and antibiotics that influence anesthetic drugs or change the QT interval. Equally crucial for surgical cardiovascular risk is the understanding of the family history related to genetic predisposition for LQTS or SCD. Obtaining a family history of congenital LQTS or other cardiac diseases will yield information regarding possible genetic risks. A detailed cardiac examination for occult cardiac diseases or arrhythmias should be performed. An electrocardiogram should be performed to record a baseline QT interval and evaluate for prolongation and potential arrhythmias. Suppose significant QT prolongation is detected or that there is a suspicion of substantial arrhythmias. In such cases, long-term monitoring with a 24-hour Holter monitor or an exercise stress test may be appropriate. Patients suspected of having congenital LQTS should undergo genetic testing for diagnostic confirmation and to identify specific mutations. Some situations do, however, require direct consultation with a cardiologist—those in which complex decisions have to be made, for example, or where there is great concern over the cardiac stability of the patient. In such instances, ancillary risk assessment with management recommendations may be delivered by a specialist. Adequate anesthesia and surgical risk management in LQTS patients involves developing strategies to minimize the risk of arrhythmias, thereby promoting safety in such patients. This forms the core of this process, where anesthetic planning takes place, and drugs that are least likely to prolong this interval QT are administered to the patient. Anesthetized drugs that may exaggerate the prolongation of QT or poorly interact with the drugs being taken by the patient are avoided. This can also be controlled by increasing the dosages of medicines used for anesthesia so that this effect can be minimized with respect to cardiac stability. Monitoring during the perioperative period should be performed mainly to detect any changes in cardiac status. Continuous ECG monitoring should be performed to detect any prolongation of the QT interval or the development of arrhythmias at any stage of the procedure. In addition, electrolytes—levels of potassium, calcium, and magnesium—with regular control should be in the normal range. All medications provided in the perioperative period should be checked with caution for potential adverse drug interactions, with very close coordination with pharmacists in tracking such interactions. Another aspect of risk management is preoperative optimization. This may include replacing electrolytes to normalize abnormalities before surgery in an attempt to decrease the risk of arrhythmias. Other medication adjustments should be made in consultation with the patient's cardiologist, particularly those medications that may impact QT prolongation. Emergency preparation is also imperative when preparing patients with LQTS for surgery. One should have available resuscitative equipment and medications in the operating room, along with anesthesia and a surgical team prepared for possible arrhythmias and emergent protocols. Monitoring should continue into the postoperative period, with the ECG and serum electrolytes monitored for late-onset arrhythmic events or complications. Thus, the patient is followed up by his cardiologist regarding surgery results and the need for readjustments in the long-term management plan. This ensures an all-round approach to the safe and effective management of LQTS patients for surgery with the most negligible risks of arrhythmias and the best results in general (Booker et al., 2003).

8. Intraoperative Management of Patients With Long QT Syndrome

Precise intraoperative management in patients with LQTS mainly lies in vigilant monitoring and individual adjustments. Monitoring must include continuation of ECG recording during surgery, with intermittent QT estimation to observe any

lengthening or change that may alert one to the increased risk of arrhythmias. Advanced telemetry systems could constantly evaluate the QT interval and produce an alert if it had become clinically significantly prolonged or if new arrhythmias had developed. In that way, a more proactive approach to the management of patient care could be provided. High-resolution electrocardiography is necessary to detect slight changes in the QT interval and establish early manifestations of potentially dangerous arrhythmias. This is very important in patients with a known history of remarkable QT prolongation. Automated systems with alerts for arrhythmia detection allow timely identification of abnormal rhythms, enhancing patient safety (Drew et al., 2004). Another critical aspect is electrolyte monitoring, entailing the need for routine checks of potassium, calcium, and magnesium at regular intervals. Electrolyte imbalances can shift the QT interval and arrhythmic risk; therefore, correction of these imbalances is very urgent to maintain cardiac stability. Hemodynamic monitoring should include blood pressure and heart rate for the detection of large swings that might decrease cardiac function and induce arrhythmias. Fluid management is essential for avoiding both hypovolemia and fluid overload, conditions that beget cardiac instability (Chen et al., 2018). The depth of anesthesia must also be monitored to prevent excessive sedation or autonomic instability, either of which may affect cardiac performance. In the event of an arrhythmic episode occurring during anesthesia, an institution for patient safety is essential with a well-defined emergency protocol. An immediate response must be made to identify the type of arrhythmia, whether it is TDP or another form of ventricular tachycardia, by instituting continuous ECG monitoring. Airway patency should be ensured, and adequate ventilation should be maintained at the most basic level (Md. Noor, 2022). Appropriate medications should be given according to the type of arrhythmia present. This can include antiarrhythmics, such as intravenous magnesium sulfate for TDP. Defibrillation may be required if drug therapy is insufficient, with defibrillation equipment available and all members of the team trained in its use. In the case of an arrhythmic event, correction of the underlying electrolyte imbalance could also prove urgently necessary. This would further help with effective communication among all the team members for the coordinated management of the patient's condition and would probably require consultation with a cardiologist or cardiac electrophysiologist in challenging cases. Monitoring should continue into the postoperative period to detect late-arriving arrhythmic events or complications. Accurate and complete documentation of the event, including the type of arrhythmia and the patient's response, provides continuity of care and helps for future reference (Moss, 2005).

9. Postoperative Considerations

The care of patients with LQTS after surgery needs to be closely monitored for safe recovery and to establish an early diagnosis of complications that may arise. Continuous ECG monitoring is vital in the immediate postoperative period for the diagnosis of possible arrhythmic events or QT interval prolongation that a patient may experience during recovery from anesthesia. In the setting of intraoperative complications or patients at high risk for arrhythmias, 24-hour Holter monitoring or telemetry may be appropriate for further evaluation. Electrolyte levels should also be monitored regularly; special attention should be given to potassium, calcium, and magnesium, whose imbalances could result in QT prolongation and an increased propensity for the development of arrhythmias. When identified, these imbalances should be urgently corrected in an attempt to ensure cardiac stability. Vital signs, especially blood pressure and heart rate, must be closely monitored for the first onset of cardiac instability. The patient must be watched for the development of palpitations, dizziness, or syncopal symptoms, which would indicate a possible arrhythmia or other complications (Golzari et al., 2007). Effective pain management is necessary, and the techniques used are designed to minimize the risk of QT prolongation. Certain medications that are likely to prolong the QT interval or have harmful interactions with drugs that are prescribed should be avoided. Nonpharmacological interventions are implemented whenever possible to reduce reliance upon potentially damaging medications. Fluid and electrolyte balance is managed carefully to avoid both hypovolemia and fluid overload, both of which may impact cardiac performance. Long-term management involves several essential considerations. This would mandate follow-up with a cardiologist at regular intervals to review the cardiac status of the patient and his response to the recent modifications of their management plan. Follow-up ECGs are necessary to confirm that the QT interval has remained within safe limits following surgery. The medication regimens must be reviewed for potential interactions with anesthesia and changes in the patient's condition (Ahmed et al., 2006). Patient education is crucial in long-term care. Patients and their families need to be educated about the symptoms of arrhythmias and how and where to report them if they return with palpitations or syncope. Adherence to medications and follow-up care is a fundamental issue to discuss. Furthermore, patients need to be educated on lifestyle changes that prevent known triggers of arrhythmias, such as vigorous exercise or emotional stress; however, patients should be encouraged to participate in safe physical activity, as determined by cardiologists. Care coordination is paramount and genuinely interdisciplinary because the cardiology team must work in close conjunction with anesthesiology and primary care providers to develop a care plan that includes acute postoperative as well as chronic management of LQTS (Epstein et al., 1996).

10. Patient Studies and Clinical Outcomes

There are several case reports of great value for obtaining general insights into the anesthetic management of patients with LQTS, both successful and challenging cases.

Ozgur and Koseoglu reported a case of a seven-year-old male patient who has been followed up ever since he was diagnosed with LQTS in the course of preoperative evaluation for adenoidectomy and tonsillectomy with a familial history of dysrhythmias. The treatment modalities were targeted at reducing the risk of possible arrhythmias by ensuring careful anesthetic selection and intraoperative monitoring. He was placed on preoperative beta-blocker therapy, and total intravenous anesthesia (TIVA) with propofol was administered to avoid the QT-prolonging effects of volatile anesthetics. The surgery was uneventful, further supporting the need for individual anesthetic management strategies in LQTS patients (Ozgur & Koseoglu, 2016).

Yaman et al. reported a case of anesthetic management for cochlear implant surgery in a 17-month-old male with congenital LQTS and bilateral sensorineural hearing loss. He had a positive family history of LQTS, with previous syncopal episodes at the age of 37 days; he was already receiving propranolol therapy. The preoperative evaluation confirmed that his vital signs were within normal limits, there was no electrolyte imbalance, and the surgical team was advised to avoid anesthetics that prolong the QT interval. Given the risks of LQTS, the anesthesia plan emphasized propofol-based TIVA to avoid QT-prolonging volatile agents. Sevoflurane was briefly used for induction to ensure better vascular access and subsequently stopped and replaced by TIVA. Lidocaine, propofol, remifentanil, and rocuronium were used for induction and maintenance. During this period, the cardiac status was continuously monitored. A defibrillator and magnesium were available to counter arrhythmias, with emphasis on TDP. The intraoperative course was uneventful, and there was no fluctuation in vital signs, with no variation in pulse rate or the QT interval. Postoperative analgesia was managed by rectal paracetamol, and phenobarbital was given to prevent agitation and smooth recovery (Yaman et al., 2021).

Intraoperative ventricular tachycardia and fibrillation occurred immediately following the induction of anesthesia in a 32-year-old male patient. The immediate interventions included the cessation of sevoflurane, manual ventilation with 100% oxygen, and emergency defibrillation. Following stabilization, the implantable cardioverter defibrillator was implanted after the patient was diagnosed with LQTS. His preoperative ECG was unremarkable, and he had significant arrhythmias during surgery, thus highlighting the role of monitoring and good management during surgery. In the subsequent surgery, very low doses of propofol and remifentanil were used to avoid lengthening of the QTc interval, with continuous ECG monitoring. Postoperative normalization of the QTc interval occurred with the stability of the patient. This case demonstrates that patients with LQTS need prudent anesthesia management and preparation for probable arrhythmias (Kim et al., 2010).

A 4-year-old female weighing 22 kg was admitted for general anesthesia for the treatment of velopharyngeal dysfunction. There were no symptoms noted during the preoperative period, and the patient had no history of arrhythmias. Her preoperative ECG, laboratory results and chest X-rays were normal. On baseline ECG, she had borderline QTc interval prolongation of 445 ms. She was induced with thiopental and rocuronium and maintained with sevoflurane. TDP followed an abrupt increase in blood pressure and heart rate. Immediate measures were taken to stop sevoflurane and administer 100% oxygen, with preparation for resuscitation. The TDP self-terminated within 60 seconds without medication or defibrillation. Dopamine was administered for blood pressure stabilization, and the surgery proceeded without further events under sevoflurane anesthesia. The QT prolongation persisted throughout the surgery, with a QTc measurement of 556 ms. Her postoperative sodium, potassium, and calcium levels were normal, whereas her echocardiogram was normal, with pulmonary edema on chest X-ray. She was diagnosed with a high probability of LQTS on the basis of Schwartz's criteria and started propranolol. Genetic testing for LQT1 was negative, and the patient had a negative epinephrine QT stress test. The patient remained asymptomatic during follow-up, and propranolol therapy was continued for three months (Jeon et al., 2020).

11. Guidelines and Best Practices

AHA guidelines are where one can find a comprehensive framework for the treatment of patients with LQTS. This emphasizes the need to establish adequate preoperative assessments, such as recording the cardiac and family history of the patient and determining the baseline measurement of the ECG. Medication management is also essential, with careful review of medications that may lengthen the QT interval or interact with anesthetics and adjustment where appropriate. Continuous ECG monitoring is recommended to detect any arrhythmic events or changes in the QT interval during surgery; the availability of proper defibrillation equipment in the operating room is a must (Fleisher et al., 2014).

The ESC guidelines build on these principles with a focus on additional considerations specific to anesthesia and surgical procedures. Principal among these is risk stratification on the basis of the type of LQTS and associated genetic mutations, with attendant tailoring of anesthetic and perioperative care accordingly. Vigilant intraoperative monitoring, as well as the use of anesthetic techniques that minimize QT prolongation, is advocated. Furthermore, the assessment of serum electrolytes and cardiac function is underlined. Postoperative monitoring will have to be continued to monitor late-onset arrhythmic events and keep treatment given to the patient in view of a long-term management plan (Zeppenfeld et al., 2022).

12. Future Directions and Research Opportunities

Research into LQTS is currently underway. New studies have identified novel genetic mutations that expand the genetic spectrum and have proposed implications for personalized medicine in terms of anesthesia responses and arrhythmia risks

attributed to specific genetic variants in patients. Studies of the molecular mechanisms underlying cardiac repolarization are revealing the ways by which mutations in ion channels interact with anesthetic agents to influence repolarization; thus, information of this nature assumes a very critical dimension in the development of targeted therapies for enhancing the management of risk. Pharmacologic advances have been made toward the development of new antiarrhythmic drugs and the study of drug interactions that minimize QT prolongation from anesthesia. Advanced risk assessment models, including genetic, clinical, and environmental factors, are in development to provide advanced prediction models of arrhythmic events and personalized management strategies. Next-generation wearable devices are advancing monitoring through the continuous monitoring and recording of high-fidelity ECG data, which could offer early detection of QT prolongation and arrhythmias. The integration of data from wearable devices into electronic health records would allow monitoring in real time, after which intervention can be feasible. Moreover, telemedicine has introduced remote monitoring, which allows for the possibility of continuous surveillance and prompt responses to arrhythmias. High-resolution ECG technology, together with artificial intelligence, significantly enhances the accuracy of arrhythmia detection and its predictive power. Advanced implantable cardiac monitors in LQTS patients have adaptive algorithms that are capable of continuous real-time dynamic monitoring; such a system captures arrhythmic events and manages them outside the hospital (Yu et al., 2023).

13. Educational and Training Needs Anesthesiologist Training Relevance of Subspecialty

An anesthesiologist should know the physiology basics and various types of LQTS, its effects on cardiac function, and its tendency toward arrhythmia, which are all necessary in deciding anesthetic management and minimizing risks associated with surgical procedures. Parents must be informed about the clinical signs and symptoms of LQTS and probable arrhythmic events so that they are able to identify the first signs and symptoms, which will help in early intervention to prevent perioperative complications (Booker et al., 2003). Preoperative assessment will include an analysis of the ECG and genetic information, as well as the identification of potential triggers to assess the risk of arrhythmias in patients with LQTS. Medication management will have to address QT-prolonging drugs and their interaction with anesthetics. Guidelines, such as those provided by the AHA, should be followed for appropriate adjustment or avoidance of such medications. Intraoperative management during this time may include the use of advanced monitoring to track QT intervals, specifically continuous or high-resolution ECG, with early detection of arrhythmias, according to the ESC. The anesthetic team should also be well conversant with protocols for emergencies involving the deployment of defibrillators and antiarrhythmic agents if an episode occurs. Continuous professional development for anesthesiologists through periodic reviews and constant updates from research, technology, and guidelines pertaining to LQTS management is paramount. Primary education about LQTS and its implications for the perioperative environment and preventive measures need to be conveyed to patients. Symptom recognition of arrhythmia should be taught to patients, and they need to understand the importance of early consultation if they suffer from palpitations or dizziness (Aroke & Nkemazeh, 2020). Preoperative preparation should also include education on the management of medications, especially regarding the disclosure of all medications and supplements taken owing to their QT-prolonging effects that dictate appropriate adjustment in doses or avoidance. This is also the right time to discuss lifestyle modifications for LQTS and treatment adherence. The anesthetic plan should be discussed with patients from the time of incidence, including measures that will be undertaken explicitly for the control of LQTS in the course of surgery and steps that are ensured for safety. An emergency action plan for episodes of arrhythmia should be understood. Such aftercare would involve impressing patients with the need for recovery monitoring and follow-up, with an emphasis on long-term vigilance in symptom reporting. The patient should be educated regarding his long-term management plan, which would include the schedule of follow-ups, adherence to medication regimens, and lifestyle modifications. Finally, patients should also be given resources from which continuous support and information can be sought (Seto et al., 2017).

14. Conclusion

LQTS is a heterogeneous cardiac disorder associated with a prolonged QT interval on electrocardiography that is associated with an increased risk of potentially life-threatening arrhythmias. With respect to proper management, congenital and acquired forms of the disorder exist. Congenital LQTS results from genetic mutations in cardiac ion channels. Exogenous factors such as medications, electrolyte imbalances, and underlying medical conditions result in acquired LQTS. The accurate diagnosis and risk assessment of LQTS require minute scrutiny of the patient's clinical history, ECG findings, and data obtained from genetic testing. Thus, there are two major diagnostic criteria for this syndrome: a prolonged QTc interval and episodes of TDP. The risk stratification was based on the clinical symptoms, family history of the patient, changing features on the ECG, and genetic parameters. The proper management of patients with LQTS requires an interdisciplinary approach to treatment planning, careful preoperative assessment, vigilant intraoperative monitoring, and thoughtful postoperative care. Anesthesia and surgery, in the context of their influence on treatment modalities, are essentially related to the selection and dosage of anesthetic drugs, drug interactions, and monitoring of the QT interval and electrolytes. During the postoperative period, ECG monitoring with attention to electrolyte balance and analgesia is continued to safeguard such patients from arrhythmias in

humans. From a highly detailed and patient-specific treatment plan, problems and risks can be reduced for many of the potential issues that can arise with LQTS. The prognosis for such patients is also likely to improve.

Ethical Considerations

This review article is based on the synthesis of existing literature and does not involve direct patient participation or the use of unpublished patient data. As such, no ethical approval or informed consent was required. Efforts have been made to present unbiased and accurate information, adhering to the principles of academic integrity and transparency.

Conflict of Interest

The authors declare no conflicts of interest related to the preparation or publication of this review.

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