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PEDIATRIC ANESTHESIA (R AGARWAL, SECTION EDITOR)

Anesthesia for Long QT Syndrome

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Abstract

Purpose of Review This review article seeks to discuss the salient information about the known genetic variants associated with congenital long QT syndrome (LQTS) and describe the current anesthetic considerations.

Recent Findings LQTS describes a heterogeneous group of patients with varying genetic mutations that cause an increased risk of torsade de pointe and sudden death. Research continues to identify more genetic mutations associated with LQTS subtypes. Publications of clinical experience with LQTS patients under general anesthesia provide new insights on the risk of medication inducing arrhythmias in these patients.

Summary Many medications used during general anesthesia are known to prolong the QTc interval. Patients with LQTS are at increased risk of arrhythmias under general anesthesia.

Keywords Congenital long QT syndrome · Torsades de pointes · Pediatric anesthesia

Introduction

Congenital long QT syndrome (LQTS) has an estimated prevalence of 1 in 2000 people and is associated with increased risk of ventricular arrhythmias and sudden cardiac death [1]. Since 1957, when the first case of LQTS was described, over 17 subtypes with a distinct gene mutation have been identified [2, 3•]. In 2017, Bohnen et al. published a thorough review of the many genetic mutations that affect each of these ion channels and the associated subtypes of LQTS [4]. These numerous gene mutations all result in the prolongation of the ventricular action potential making the myocytes prone to early or delayed afterdepolarizations that may cause torsades de pointes (TdP), ventricular fibrillation (VF) and/or sudden death. The aim of this article is to explore implications of congenital long QT syndromes for patients undergoing general anesthesia.

Acquired long QT syndrome is attributed to causes other than the genetic syndromes mentioned above and include, electrolyte abnormalities (hypokalemia, hypocalcemia, hypomagnesemia), and medications (ondanestron, diphenhydramine, furosemide) [5]. Though this is not the focus of this paper, it is relevant to mention since such patients are at risk for arrhythmias and do present for general anesthesia. These include oncology patients in whom chemotherapeutic regimens have resulted in a prolongation of the QT interval or critically ill patients from the intensive care unit [5, 6]. Lim et al. published a single-center retrospective study of the prevalence of QTc prolongation and risk of TdP in hospitalized pediatric oncology patients. In this study of 287 pediatric oncology inpatients on whom an electrocardiogram (ECG) was performed, 9% had a prolonged QTc interval defined as QTc > 500 ms and 2 of these patients experienced an episode of TdP [6]. This patient population should be screened for prolonged QTc intervals with ECGs, monitoring and correction of electrolytes, avoidance of QTc prolonging medications, and clinicians should be prepared to respond to arrhythmias under general anesthesia [7].

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Search Strategy

A PubMed database search was completed for all types of published articles using the following keywords: "pediatric anesthesia and long QT syndrome," "congenital long QT syndrome," "prolonged QTc interval," to identify most recent and relevant publications. Further publications were identified from the references of the original articles.

Pathophysiology of LQTS

Genetic mutations of LQTS impact the function of different ion channels to disrupt the ventricular myocyte's ability to create and maintain the ion milieu needed for a normal action potential. Figure 1 illustrates the temporal movement of ions during an action potential and where some of the most common genetic mutations of LQTS occur during the action potential [4, 8]. During the depolarization phase, sodium channels are activated to bring sodium into the cell. In the plateau phase, sodium channels are not active, and potassium and calcium ion flows are balanced. Potassium channels are active during the repolarization phase moving positive charge out of the cell at a greater magnitude than the influx from calcium channels during the preceding phase of the action potential for a net effect of bringing the cell back to the resting potential. Loss of function mutations in the potassium channels or gain of function mutations in the sodium and calcium channels will result in prolongation of the action potential by delaying repolarization.

The interaction of the autonomic nervous system on the specific ion channel activation and deactivation during the action potential has implications for the pathophysiology of the specific LQTS subtypes [8]. Ninety percent of the LQTS patients with identified genetic mutations have LQT1, 2 or 3 subtypes [9]. LQTS 1, the most prevalent subtype at 35% frequency, results in a loss of function of one of the potassium channels such that ion flow is not increased by sympathetic stimulation and the QT interval does not shorten in response to tachycardia [10, 11]. Thus, affected patients are at risk for arrhythmias triggered by sudden increases in heart rate as seen with increased sympathetic tone, exercise, stress, or stage 2 of inhaled induction of volatile anesthesia. Beta blocker therapy to blunt the tachycardic response to increased sympathetic tone has proven effective in decreasing incidence of sudden and lethal cardiac arrhythmias and is the mainstay for management of LQTS [3•]. Other treatment modalities including left cardiac sympathetic denervation (LCSD) or placement of an implantable cardiac defibrillator (ICD) may be used in patients with arrhythmias despite optimal beta blocker therapy or with known high risk LQTS subtypes [3•]. Table 1 shows relevant characteristics of the most common or lethal LQTS subtypes.

Diagnosis of QT Prolongation

The QT interval, defined by the start of the Q wave to the end of the T wave, is the time for full ventricular depolarization and repolarization. Corrected QT interval (QTc) is referenced because it refers to the QT interval after correction for variations in heart rate [12]. A normal QTc interval on a 12-lead ECG tracing is < 440 ms for males and < 460 ms for females. QTc intervals greater than these

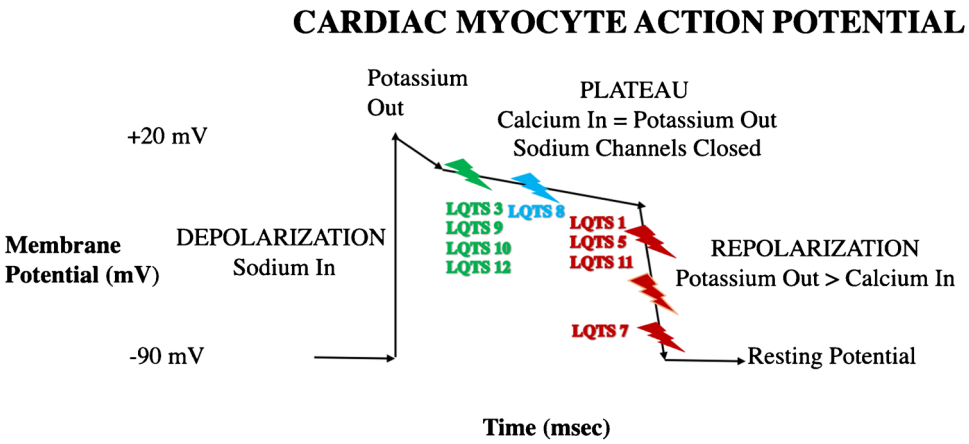

Fig. 1 Resting potential of the cardiac myocyte is − 90 mV. During the action potential, with the change ion flow in and out of the cell the membrane, potential peaks at + 20 mV before returning to resting potential. In the depolarization phase, sodium channels are activated to bring sodium into the cell. In the plateau phase, sodium channels are inactivated and potassium and calcium ion flows are balanced. Repolarization begins as the potassium channels moving positive charge out of the cell overcomes magnitude of the calcium influx for a net effect of bringing the cell back to the resting potential. Genetic mutations in each of the ion channels have been associated with LQTS subtypes as noted in color coding (green = sodium channel, blue = calcium channel, red = potassium channels)

Table 1 LQTS subtypes

Subtype	Frequency	Affected ion channel	Clinically Significant Characteristics
LQTS 1	40–55%	IKs – loss of function	Triggers for arrhythmia: Stress, exercise, increased SNS output
LQTS 2	30–45%	IKr – loss of function	Triggers for arrhythmia: Sudden startle/fear, increased SNS output
LQTS 3	5–10%	INa – gain of function	Triggers for arrhythmia: Pause dependent arrhythmia in sleep
LQTS 5	< 1%	IKs – loss of function	Jervel and Lange-Nielsen syndrome: autosomal recessive, deafness, 90% have cardiac events
LQTS 8	< 1%	ICa – gain of function	Timothy syndrome: syndactyly, congenital heart disease

IKs delayed rectified potassium current channel, *SNS* sympathetic nervous system, *PPM* permanent pacemaker, *ICD* implantable cardiac defibrillator, *IKr* rapid rectified potassium current channel. *INa* sodium current channel, *ICa* calcium current channel [8]

ranges are considered prolonged and QTc interval greater than 500 ms put a patient at significant risk of TdP [6, 13].

One nuance of the LQTS is that the correlation between genotype and phenotype is variable for all subtypes and thus clinical presentation can vary in terms of type and severity of symptoms and patient age. Amongst patients with identified LQTS gene mutations, resting ECG often demonstrates QTc interval in the normal range [9]. This makes diagnosis and more importantly prediction of prognosis more difficult in these patients. Patients who present with a history of syncopal episode, epilepsy of unknown etiology, with a history of sudden cardiac death in a family member, or drug-induced prolonged QT syndrome should raise a clinician's suspicion for congenital long QT syndrome. The Schwartz criteria, assigns a point system between 0.5–3 based on 5 separate ECG findings, clinical histories of syncope and/or congenital deafness, and history of a family member with LQTS and/or sudden death in a family member younger than 30 years [8, 14]. The summation of the numerical scores results in one of three conclusions about probability of LQTS: low probability (score < 1),intermediate probability (score = 1.5–3), and high probability (score ≥ 3.5) [8]. This provides a quantitative score to determine which patients with suspicion for LQTS are likely to be affected by LQTS and thus should undergo genetic testing [8].

Anesthetic Management

The anesthetic management of patients with LQTS is based on preventing further lengthening of the QTc interval and treating ventricular arrhythmias. Perioperative QT prolongation is relatively common, but thankfully Torsades de pointe (TdP) under anesthesia is rare [15]. In 2014, Whyte, et al. published a retrospective chart review of LQTS patients receiving general anesthesia. The overall incidence of TdP was low at 3.1% and all events occurred in infants undergoing surgery specifically for LQTS [16]. They also found no correlation with arrhythmia events and anesthetic drug choice. Case reports, reviews and expert opinion currently provide guidance for perioperative management and can support intraoperative decision making in patients with LQTS. Table 2 lists commonly used anesthetic drugs and preference of use in LQTS.

Preoperative Care

The first step in assessment of a patient with LQTS is to understand the etiology. As described earlier, there are many genetic mutations associated with LQTS. Therefore, when possible, this should be delineated to understand the risk and potential arrhythmia triggers. For example, patients with LQT1 genotype are more likely to suffer from arrhythmia events during sympathetic stimulation (i.e.: anxiety or intubation), while LQT3 patients will classically have arrhythmia events during periods of bradycardia (i.e.: sleep

Table 2 Common anesthetic drugs and LQTS risk [11, 12, 17•, 18]

Considered Safe	Use caution or avoid
Midazolam	Dexmedetomidine
Acetaminophen	Methadone
Ibuprofen	Sufentanil
Ketorolac	Propofol
Fentanyl	Etomidate
Morphine	Ketamine
Remifentanil	Volatile anesthetics (Sevoflurane, isoflurane, desflurane)
Rocuronium	
Vecuronium	Nitrous oxide
Cisatracurium	Succinylcholine
Sugammadex	Glycopyrrolate
Lidocaine	Atropine
Bupivacaine	Neostigmine
Ropivacaine	Ondansetron
Dexamethasone	Droperidol
Vasopressin	Epinephrine
Esmolol	Norepinephrine
	Dopamine
	Phenylephrine
	Ephedrine
	Albuterol

or sedation) [3•]. In addition to genotype awareness, there are certain patients that carry a higher baseline risk for malignant arrhythmias which the anesthesiologist should be aware of. Table 3 describes patient characteristics associated with high risk of arrhythmias [16, 19, 20]. Because LQTS describes a diverse patient population, it is difficult to define risk for every patient. Caution should be utilized for all patients with LQTS. An anesthesiologist with experience in cardiac resuscitation should care for patients with LQTS. In addition, electrophysiology consultation should be available for patients with LQTS and high risk characteristics.

As always, a thorough history and physical exam should be done, with emphasis on cardiac symptoms such as dizziness, palpitations, syncope, and family history of sudden death. Patients with new or worsening cardiac symptoms suggestive of LQTS should not undergo elective procedures until evaluation has been initiated. A recent ECG should be reviewed, and the patient's cardiologist made aware of the intended procedure. It is also important to note that some patients will have a normal QTc interval on resting ECG despite having a genetic mutation known to cause LQTS. Even with a normal QTc interval, these patients are still at risk of having an arrhythmia event [21]. An echocardiogram is not required for every procedure, but previous documentation of a structurally normal heart is necessary. Electrolytes should be normalized and may necessitate preoperative laboratory testing if there are concerns or medication changes. Furthermore, it is imperative that patients continue medications used to treat LQTS on the day of surgery. Beta-blockers, sodium channel blockers, and potassium supplements are commonly used (depending on the patient's genotype variation) and should not be stopped. Pacemaker/ICD interrogation and reprogramming should be arranged based on the intended procedure.

Because stress and anxiety can induce arrhythmias in certain LQTS genotypes, a calm environment and anxiolysis plan is highly recommended. Minimizing NPO time and planning for a first case start time can decrease stress and dehydration. Preoperative midazolam does not prolong the QT interval and should be given when appropriate [12]. In contrast, dexmedetomidine can cause bradycardia and has been shown to lengthen the QT interval [22, 23]. There are conflicting reports on the QT prolonging effect of dexmedetomidine, so its use in LQTS patients may be limited until further studies have defined its effect [24].

Table 3 High-risk patient characteristics [16, 19, 20]

QTc interval > 500 ms; extremely high risk if > 600 ms
T – wave alternans on 12 lead ECG
Diagnosis of Timothy syndrome (LQT 8)
Diagnosis of Jervell and Lange-Nielsen syndrome
Patients < 7 years with syncope and/or cardiac arrest; extremely high risk < 1 year
Persistent arrhythmias despite full therapy
Patients presenting for surgical intervention to treat LQTS (ex: ICD or sympathectomy)

Intraoperative Care

The intraoperative anesthetic plan targets avoiding sympathetic stimulation, avoiding medications known to prolong the QTc interval and managing ventricular arrhythmias. Both inhalation and IV induction have been reported in patients with LQTS [16, 25]. Sympathetic stimulation will increase the likelihood of ventricular arrhythmias, so minimizing this risk should be prioritized during induction. The patient is especially vulnerable during intubation, both during endotracheal tube placement and transition to positive pressure ventilation. Hypoxia and hypercapnia will cause sympathetic stimulation and high peak airway pressures have been shown to lengthen the QTc [12]. Using topical lidocaine on the vocal cords prior to intubation may be beneficial.

Induction Agents All volatile anesthetics prolong the QTc interval and some case reports have described arrhythmias in association with their use, but many have not [13, 16, 25, 26]. Nitrous oxide is a known sympathomimetic so its use may be limited [11]. With regard to IV induction agents, propofol is probably the most commonly used agent in LQTS. However, CredibleMeds® still considers propofol a known risk for TdP despite the many reports of propofol use that are not associated with arrhythmias or QTc prolongation [27–30]. Ketamine will cause sympathetic stimulation and is best avoided at large doses, and etomidate has limited and conflicting data similar to propofol [31]. It therefore appears that there is no anesthetic induction agent without some theoretical risk of QT prolongation. Sympathetic stimulation will increase the likelihood of ventricular arrhythmias, so efforts should be made to minimize this risk since both inhalation or IV induction have been described [16, 25].

Neuromuscular Blockers and Reversal Agents Succinylcholine, a depolarizing neuromuscular blocker, will increase the QTc interval and therefore should be avoided when possible. Non-depolarizing neuromuscular blockers (rocuronium, vecuronium and cisatracurium) are considered safe as they do not pharmacologically lengthen the QTc interval [12]. Pancuronium administration classically causes tachycardia and should be avoided, and the histamine release commonly seen with atracurium makes it an inferior choice as well. It should be emphasized that when administering a paralytic, one must ensure that the patient has an adequate depth of anesthesia to avoid increased sympathetic stimulation which

could induce a ventricular arrhythmia. Sugammadex is the preferred reversal agent for patients with LQTS because it has not been shown to lengthen the QTc [32]. Anticholinesterase-anticholinergic reversal technique (neostigmine and glycopyrrolate) does prolong the QTc so it is best avoided, especially given a superior alternative [33].

Pain Management Providing adequate pain control is paramount when treating patients with LQTS. Acetaminophen and NSAIDs can be given safely and should always be considered for multi-modal pain management. Some commonly used opioids (fentanyl, morphine, remifentanil) will not prolong the QTc and are viewed as safe. In contrast, methadone and sufentanil are known to lengthen the QTc and should be used with caution [12]. Whenever possible, regional anesthesia should be considered to help with pain management. In addition, neuraxial anesthesia has been shown in the literature to be safe in patients with LQTS [18]. Epidural anesthesia has the advantage of medication titration. Local anesthetics can be used safely, but an epinephrine additive is best avoided [11].

Vasoactive Support It is often necessary to prepare for hemodynamic shifts in patients with known cardiac disease. Unfortunately, the common vasoactive agents used in anesthesia may length the QTc and should be avoided. Epinephrine, dopamine, phenylephrine and ephedrine have all been classified as agents that can prolong the QTc [12]. Phenylephrine has somewhat conflicting reports, so should be used with caution [34, 35]. As such, vasopressin and fluid bolus are the author's preferred strategies for intraoperative hypotension.

Postoperative Care

Vigilance must continue in the postoperative period because of the continued risk of sympathetic stimulation associated with pain, delirium and nausea/vomiting. A deep extubation should be considered and pain aggressively treated. Patients should be monitored with telemetry during the recovery period. Patients with a pacemaker or ICD will need post-operative interrogation and programming.

Patients who require treatment for arrhythmias should be admitted for observation and cardiology should be consulted to assist with clinical management. Overnight admission should otherwise be at the discretion of the anesthesiologist, with input from the patient's cardiologist and surgeon. It is the author's opinion that even patients who had an uneventful anesthetic should still be monitored for an extended period of time in the recovery unit to ensure there are no post-operative arrhythmias.

Antiemetics Post-operative nausea and vomiting (PONV) plays a significant role in perioperative care. The different agents commonly used to prevent PONV are not typically used in patients with LQTS because of the concern about their QTc prolongation effect. Ondansetron, the most common 5-hydroxytryptamine type 3 serotonin receptor antagonist, has an FDA warning against its use in patients with LQTS [17•, 36]. Caution should be taken if ondansetron is administered with other QT prolonging medications [12]. Promethazine and droperidol will lengthen the QTc interval and should be avoided [36, 37]. Dexamethasone and scopolamine patch for PONV can be used in patients with LQTS [37].

Arrhythmia Management

When caring for a patient with known LQTS, it is important to anticipate rhythm disturbances during the entire perioperative period. The patient should have ASA monitors prior to induction, including at least a 3-lead ECG. Permanent pacemaker/ICDs need perioperative interrogation and reprogramming. If the patient is pacemaker dependent, conversion to an asynchronous mode for surgery may be necessary. ICD function may need to be deactivated, especially if cautery is used, and the external defibrillation pads must then be applied to the patient and connected to the defibrillator. In addition, operating room staff will need to be familiar with the defibrillating and pacing functions of the device in use.

Torsade de pointe is the most classically associated rhythm disturbance in LQTS, but the anesthesiologist should also be prepared to treat tachycardia, bradycardia, ventricular fibrillation and cardiac arrest. The first line treatment for torsade de pointe is administration of intravenous magnesium sulfate (30–50 mg/kg bolus followed by an infusion). If TdP persists and the patient is stable with a pulse, temporary pacing could be attempted to terminate the arrhythmia [18]. If the patient has TdP without a pulse, defibrillation (2–4 J/kg) is necessary along with cardiopulmonary resuscitation. Defibrillation should not be delayed in the event of transformation to ventricular fibrillation. Amiodarone is not recommended for patients with LQTS, but lidocaine can be used instead for treatment of ventricular arrhythmias. Epinephrine should be used cautiously because of its direct sympathetic effect that could worsen the arrhythmia.

Treatment of bradycardia may necessitate pacing, either transcutaneous or with the patient's device. The use of isoproterenol or dobutamine for treatment of bradycardia should only be considered in consultation with a cardiologist due to the increased risk of QTc prolongation [18]. Sinus tachycardia in the perioperative period can have multiple etiologies. Once the common perioperative causes of sinus

tachycardia have been addressed, the use of short acting beta blockers, such as esmolol, may be warranted.

Conclusion

Congenital long QT syndrome comprises a challenging group of patients for anesthesiologists. Hemodynamic perturbations, like tachycardia, hypoxia, pain, etc. are commonly seen under general anesthesia and could induce a malignant arrhythmia in LQTS patients. In addition, because of the known risk of ventricular arrhythmias such as Torsades de pointes, these patients and family members may be very fearful of undergoing anesthesia and will require additional time and counseling. This review discusses some of the current concepts around anesthesia and long QT syndrome and provides the reader with background knowledge of the pathophysiology and guidance on anesthetic care in patients with long QT syndrome.

Compliance with Ethical Standards

Conflict of Interest The authors declare no competing interests.

Human and Animal Rights and Informed Consent This article does not contain any studies with human or animal subjects performed by any of the authors.

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