

Long QT Syndrome and Anaesthesia

Denise de Noronha^{1†}, Sandra Maurício,² Idalina Rodrigues³

¹Anaesthesia Trainee, Centro Hospitalar Universitário Lisboa Norte, Portugal

²Anaesthesia Trainee, Instituto Português de Oncologia, Portugal

³Consultant Anaesthetist, Centro Hospitalar Universitário Lisboa Norte, Portugal

Edited by: Dr. Lara Herbert, Consultant Anaesthetist, Royal Cornwall Hospitals NHS Trust, UK

†Corresponding author e-mail: denise.d.noronha@gmail.com

Published 8 June 2021



KEY POINTS

- The QT interval on the electrocardiogram reflects the time taken for the ventricle to depolarise and repolarise.
- Long QT syndrome is a conduction disorder characterised by prolongation and dispersion of ventricular repolarisation, making torsade de pointes ('twisting of the points') more likely.
- Both congenital and acquired forms are recognised.
- Symptoms include palpitations, syncope and sudden cardiac arrest. Asymptomatic cases are common.
- Drug-induced QT prolongation is common in the hospital setting, particularly in the perioperative period.
- Appropriate avoidance of drugs that can prolong the QT interval, together with careful selection of anaesthetic drugs and avoidance of arrhythmogenic triggers, helps to ensure an uneventful perioperative course.
- Immediate recognition and treatment of torsade de pointes may be life-saving. Treatments include intravenous magnesium, overdrive pacing, electrical cardioversion and defibrillation depending on haemodynamic stability.

INTRODUCTION

The prolongation of the ventricular repolarisation time and QT interval dispersion (measured as interlead variability of QT) are known risk factors for the development of malignant arrhythmic episodes and cardiac arrest.

The true prevalence of long QT syndrome (LQTS) remains unknown, as it is often undiagnosed. The prevalence of congenital LQTS is estimated to be close to 1:2000 live births,¹ but if genotype-positive/phenotype-negative individuals are included, the numbers would probably be considerably higher. Likewise, acquired LQTS without torsades de pointes (TdP) is more common than TdP events, which may further explain underreporting. In one retrospective single-centre study, 0.7% of all hospitalised patients had a QTc \geq 500 ms, but less than 6% of these patients experienced a life-threatening arrhythmia.²

LQTS is significant in surgical patients because many perioperative drugs are known to lengthen the QT interval. It has been shown that at the end of surgery, 80% of patients experience a significant QTc prolongation, Δ QTc 23 ms (95% confidence interval [CI] 20-25 ms).³

DEFINITION

The QT interval reflects both the ventricular depolarisation and repolarisation times in the electrocardiogram (ECG) and is measured from the onset of the QRS complex until the end of the T wave (Figure 1). Since the QT interval varies inversely with the heart rate (longer at slower rates and shorter at faster rates), the QT duration must be corrected for the heart rate (QTc). The simplest and most commonly used formula was developed by Bazett: $QTc \text{ (ms)} = QT \text{ interval (ms)} \div \sqrt{RR \text{ interval (ms)}}$.⁴

An online test is available for self-directed continuous medical education (CME). It is estimated to take 1 hour to complete. Please record time spent and report this to your accrediting body if you wish to claim CME points. A certificate will be awarded upon passing the test. Please refer to the accreditation policy [here](#).

[TAKE ONLINE TEST](#)

Subscribe to ATOTW tutorials by visiting <https://resources.wfsahq.org/anaesthesia-tutorial-of-the-week/>

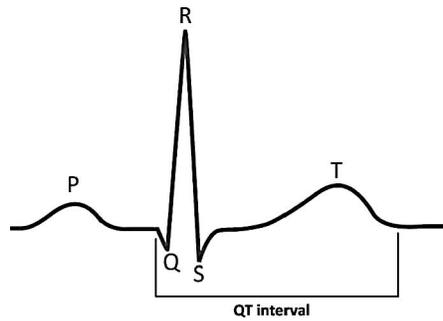


Figure 1. QT interval.

The QTc in healthy subjects varies widely, with a mean and standard deviation of 412 ± 25 ms, respectively, and a 99th percentile value of 470 ms in the adult population.⁵ A QTc interval greater than 480 ms is indicative of an LQTS.⁶ However, several limitations must be considered:

- The Bazett formula is less accurate at heart rate extremes, resulting in undercorrection at low heart rates and overcorrection at high heart rates.⁷
- Accurate measurements of the QT interval can be technically difficult, especially regarding the termination of the T wave and in cases of irregular rhythms. Therefore, the QT interval must be measured manually in several leads (eg, II and V5), averaged over different beats and adjusted for the heart rate.
- U waves should not be included, as they lead to overestimation of QTc.⁸
- It has been suggested that using a threshold of 500 ms for a prolonged QTc or using the JT interval ($JT = QT - QRS$) rather than the QT interval are more appropriate approaches when QRS prolongation is present.⁷

PATHOPHYSIOLOGY

An understanding of the normal myocardial action potential (Figure 2) is required to understand the pathophysiology behind QT prolongation:

- Phase 4, or the resting membrane potential, is when the cell membranes during diastole are polarised at -90 mV. This is mainly due to the constant outward leak of potassium ions through inward rectifier potassium (or I_{K1}) channels.

Action potential of cardiac muscles

Grigoriy Ikonnikov and Eric Wong

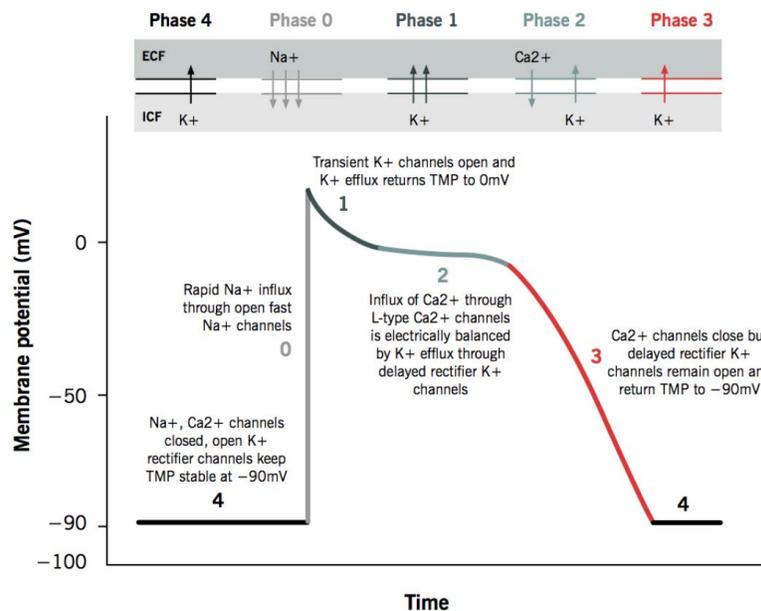


Figure 2. Myocardial action potential. TMP indicates transmembrane potential.¹³ Permission granted by Eric K. C. Wong, MD. This image/content is not covered by the terms of the Creative Commons license of this publication. For permission to reuse, please contact the rights holder.

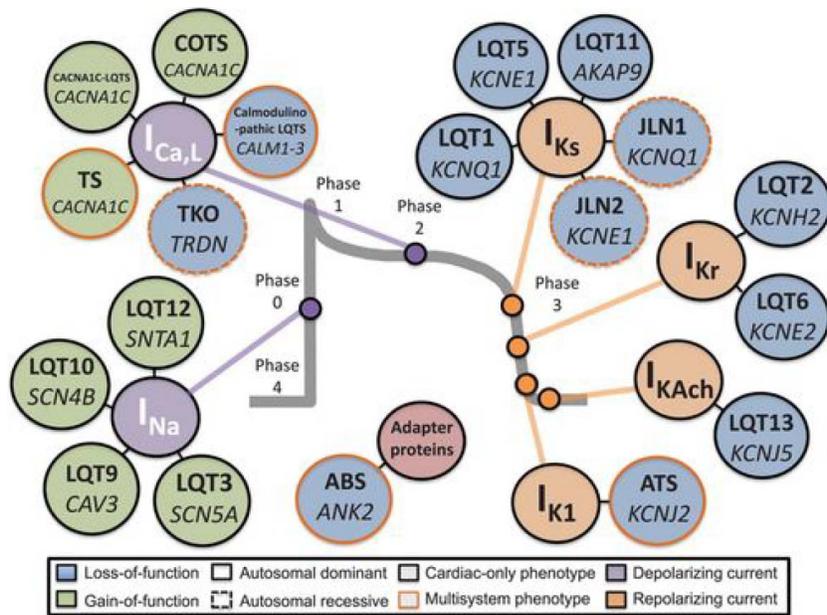


Figure 3. Current-centric classification of long QT syndrome genotypes.¹⁴ Reprinted from Giudicessi JR, Ackerman MJ. Determinants of incomplete penetrance and variable expressivity in heritable cardiac arrhythmia syndromes. *Translational Research*, 161(1):1–14. Copyright 2013, with permission from Elsevier.

- Phase 0 is the phase of rapid depolarisation. The membrane potential reaches +20 mV due to a rapid inward flow of sodium ions through inward sodium (or I_{Na}) channels.
- Phase 1 is due to initial repolarisation caused by potassium ion efflux via transient outward potassium (or I_{to}) channels.
- Phase 2, or the plateau phase, represents an equilibrium between the entry of calcium ions into the cell via long-lasting and transient calcium (or I_{Ca-L} and I_{Ca-T}) channels, and the outward potassium currents via rapidly and slowly delayed rectifier (or I_{Kr} and I_{Ks}) channels.
- Phase 3 is the phase of rapid repolarisation where outward potassium currents (I_{Ks} and I_{Kr}) dominate.

Prolongation of the QT interval, as seen in LQTS, results from prolongation of repolarisation mainly in phase 3. This increases the propensity for early after-depolarisations, some of which reach threshold potential and can lead to TdP.

AETIOLOGY

The prolongation of repolarisation can be due to congenital mutations or acquired factors.

Congenital LQTS

Mutations in more than 15 genes have been identified in patients with congenital LQTS. Mutations in the *KCNQ1*, *KCNH2* and *SCN5A* genes account for 75% to 80% of all congenital LQTS. Mutations in these genes lead to suppression of I_{Ks} channels, suppression of I_{Kr} channels and increased number of I_{Na} channels, respectively. Figure 3 illustrates several gene mutations in the context of the specific current perturbed.

In general, 2 syndromes characterised by congenital LQTS have been described⁹:

- The more common autosomal dominant form, Romano-Ward syndrome, has only cardiac manifestations.
- The autosomal recessive form, Jervell and Lange-Nielsen syndrome, is characterised by both LQTS and sensorineural deafness.

Patients with multiple mutations exhibit significantly longer QT intervals and have a greater risk for life-threatening cardiac arrhythmias.

Acquired LQTS

Acquired LQTS usually requires exposure to drugs or electrolyte disturbances that induce an iatrogenic prolongation of the myocardial repolarisation time. Most drugs that lead to LQTS block the I_{Kr} current.¹⁰ In these patients, ventricular tachycardia usually follows short-long RR intervals (a premature ventricular contraction followed by a compensatory pause) or is associated

Factor	Condition or Trigger
Metabolic factors (“hypos”)	Hypokalaemia, hypomagnesaemia, hypocalcaemia, hypothyroidism, hypothermia
Bradyarrhythmias	Sinus node dysfunction, atrioventricular block
Other clinical factors	Female sex, myocardial ischemia, intracranial disease, HIV, autoimmune diseases
Androgen deprivation	Bilateral orchiectomy, leuprorelin
Antianginal drugs	Ivabradine, ranolazine
Antiarrhythmic drugs	Disopyramide, hydroquinidine, procainamide, quinidine, flecainide, pilsicainide, propafenone, amiodarone, dofetilide, dronedarone, ibutilide, nifekalant, sotalol, vernakalant
Anticholinergic drugs	Solifenacin, tolterodine
Anti-infective drugs	
Fluoroquinolones	Ciprofloxacin, gatifloxacin, gemifloxacin, grepafloxacin, levofloxacin, moxifloxacin, sparfloxacin
Macrolide antibiotics	Azithromycin, clarithromycin, erythromycin, roxithromycin
Antituberculous drugs	Bedaquiline
Azole antifungals	Gluconazole, voriconazole
Antiparasitic drugs	Pentamidine
Antimalarial drugs	Chloroquine, delamanid, halofantrine, hydroxychloroquine, quinine, piperaquine
Antiretrovirals	Saquinavir
Antihistamines	Astemizole, hydroxyzine, terfenadine
Antineoplastic drugs	Aclarubicin, arsenic trioxide, ceritinib, crizotinib, dasatinib, encorafenib, gilteritinib, inotuzumab ozogamicin, ivosidenib, lenvatinib, midostaurin, nilotinib, osimertinib, oxaliplatin, ribociclib, toremifene, vandetanib, vemurafenib
Diuretics	Via electrolyte changes
Gastrointestinal drugs	
Antiemetics	Droperidol, ondansetron
Promotility drugs	Cisapride, domperidone
Neuromuscular blockers	Succinylcholine, pancuronium
Neurologic drugs	Apomorphine, deutetrabenazine, donepezil, ezogabine, fingolimod, pimavanserin, tetrabenazine
Opioids	Buprenorphine, hydrocodone, levacetylmethadol, methadone, sufentanil
Psychotropic drugs	
Antipsychotics	Amisulpride, chlorpromazine, chlorprothixene, clozapine, flupentixol, haloperidol, levomepromazine, mesoridazine, olanzapine, pimozide, quetiapine, risperidone, sulpiride, thioridazine, ziprasidone
Antidepressants	Citalopram, clomipramine, doxepin, escitalopram, imipramine
Other	Atomoxetine, trazodone, valbenazine
Anaesthetics and sedatives	Chloral hydrate, ketamine, propofol, thiopentone, desflurane
Vasodilator drugs	Bepidil, cilostazol, intracoronary papaverine
Miscellaneous	Anagrelide, alfuzosin, cesium chloride, clofazimine, cocaine, eliglustat, ibogaine, lofexidine, mifepristone, oxytocin, pasireotide, probucol, terlipressin, terodiline, organophosphate intoxication

Table 1. Some Causes and Triggers of Long QT Syndrome (Both Congenital and Acquired)

with bradycardia. In fact, lower heart rates result in less potassium moving out of the cell during repolarisation, and the reduction in extracellular potassium further contributes to the drug-induced inhibition of I_{Kr} .¹⁰

Overlap may be present, in which some patients with acquired LQTS have a *forme fruste* of congenital LQTS, a mutation or polymorphism that is clinically unapparent until exposure to an additional insult occurs. QT prolongation often goes unnoticed until symptoms occur, so that acquired LQTS without TdP is more frequent than TdP events. Table 1 includes common causes and triggers of LQTS. A more complete list of such drugs is available at <http://crediblemeds.org>.¹¹

DIAGNOSIS AND TREATMENT

Most individuals are asymptomatic, being diagnosed because of an affected family member or a prolonged QT interval that is detected by an ECG obtained for sports participation or anaesthesia. The primary symptoms, usually precipitated by a trigger (Tables 1 and 2), include palpitations, arrhythmic syncope with or without generalised seizures and sudden cardiac arrest.¹²

After obtaining a comprehensive personal and family history, as well as ECG monitoring, a Schwartz score should be calculated (Table 3) to determine the probability of the presence of congenital LQTS. Diagnosis of acquired LQTS can be made

Category	Drug or Condition
Appetite suppressants	Benzphetamine, diethylpropion, phendimetrazine, phentermine, phenylpropanolamine, sibutramine
Antiepileptic	Fenfluramine
Antibiotics	Sulfamethoxazole-trimethoprim
Attention deficit disorder	Amphetamine, dexamethylphenidate, dextroamphetamine, lisdexamphetamine, methamphetamine, methylphenidate
Beta-2 agonists	Arformoterol, fenoterol, formoterol, indacaterol, levosulbutamol, olodaterol, metaproterenol, ritodrine, salbutamol, salmeterol, terbutaline, vilanterol
Cardio and vasoactive	Atropine, dobutamine, dopamine, droxidopa, ephedrine, epinephrine, glycopyrrolate, isoproterenol, midodrine, norepinephrine, phenylephrine
Nasal decongestants	Oxymetazoline, pseudoephedrine, xylometazoline
Activity and environmental factors	Catecholamine surge, exercise, particularly swimming (LQTS 1) Acute arousal, including emotional and auditory stimuli (LQTS 2) Postpartum period (LQTS2) Rest and sleep (LQTS 2 and LQTS 3)

Table 2. Special Risk Factors in Congenital Long QT Syndrome (LQTS)

in a patient with sufficient QT prolongation that is associated with a known risk factor and that is reversible upon removal of the risk factor. If congenital LQTS is suspected, a cardiologic evaluation and genetic testing are advised.

Whilst most patients with congenital LQTS are started on a beta-adrenergic blocker, the cornerstone of the management of acquired LQTS is addressing the underlying cause. An implantable cardioverter defibrillator (ICD) should be considered for congenital forms in cases of extremely prolonged QTc or recurrent arrhythmias despite maximally tolerated doses of beta blocker.

Condition	Points
History	
Family*	
History of LQTS	1
Unexplained sudden death in a family member younger than 30 y	0.5
Patient	
Congenital deafness	0.5
Prior history of syncope [†]	
Postexercise	1
With stress	2
Probability score	
Low	≤ 1
ECG [‡]	
QTc (Bazzett's formula)	
450-459 ms (males)	1
460-479 ms	2
≥480 ms	3
≥480 ms fourth minute of recovery from exercise stress test	1
Other	
Torsade de pointes [†]	2
T-wave alternans	1
Notched T waves in >3 leads	1
Bradycardia	0.5
Probability score	
Intermediate	1.5-3
High	≥ 3.5

Table 3. Schwartz Score: Diagnostic Criteria for Congenital Long QT Syndrome (LQTS). *The history of LQTS and unexplained sudden death should occur in 2 distinct family members to qualify for each set of points. [†]Mutually exclusive. [‡]After excluding iatrogenic causes

PREOPERATIVE MANAGEMENT

The preanaesthetic evaluation should include review of personal or familial history of LQTS-related symptoms as well as all medications. Asymptomatic patients without a history of arrhythmic symptoms carry lower risk.

Preoperative 12-lead ECG is indicated to establish a baseline ECG. Cardiology consultation before elective surgery is warranted in cases with prior life-threatening arrhythmias, significant QT prolongation or anticipated interference with the normal function of cardiac implantable devices.

To prevent further QT prolongation and resulting arrhythmias, the following perioperative approach is warranted:

- **Avoidance of QT prolonging drugs.** The list is extensive and includes widely used drugs including antiarrhythmics, antibiotics, antihistamines, antidepressants, antipsychotics and antineoplastic agents (Table 1). An individual risk-benefit analysis should be undertaken so that all nonessential treatments are paused.
- **Metabolic and electrolyte homeostasis.** Hypokalaemia, hypomagnesaemia, hypocalcaemia and hypothyroidism further prolong repolarisation time. These imbalances should be identified and corrected prior to the surgery.
- **Chronic beta-adrenergic blocker therapy when indicated.** Beta-adrenergic blocking agents are the mainstay of treatment for congenital LQTS (class I recommendation in both the European and American Cardiological Societies' guidelines). Propranolol is suggested given its superior efficacy, likely because of the additional inhibition of late sodium currents. This therapy inhibits both prolongation and dispersion of repolarisation, reducing the rate of malignant arrhythmias. It should be continued throughout the perioperative period (change to a parenteral formulation if nil by mouth). In contrast, there is no strong evidence to support chronic beta-adrenergic blockade in purely acquired LQTS.

INTRAOPERATIVE MANAGEMENT

The perioperative period is potentially life-threatening, since the patient is exposed to myriad risk factors, including haemodynamic instability, catecholamine surges, metabolic imbalances and exposure to drugs known to prolong the QT interval (although not necessarily torsadogenic).

Premedication and General Conditions

Benzodiazepines (in particular, midazolam) decrease the sympathetic tone and should be considered for anxiolysis, particularly in congenital LQTS. In addition, genotype-specific steps can be taken. For instance, loud noises may act as a trigger for those with the LQT2 genotype.⁶ Therefore, a calm environment in the operating theatre should be prioritised, including reduction of alarm sounds during induction and emergence.

Equipment and Monitors

The American Society of Anesthesiologists' guidelines recommend standard monitoring comprising noninvasive arterial blood pressure, heart rate, continuous ECG (consider 5-lead), pulse oximetry, end-tidal carbon dioxide and body temperature. Before starting the case, it is important to check that a working defibrillator is located in theatre and that intravenous (IV) magnesium sulphate is available. In high-risk patients, the use of self-adhesive multifunction pads should be considered.

Overall Goals of Anaesthesia

Regardless of the surgery or anaesthesia technique chosen, maintenance of homeostasis is the priority. Avoid hypoxaemia, hypocapnia, hypercapnia and high-pressure ventilation. Sudden changes in heart rate and blood pressure should be anticipated and avoided. Maintain normothermia throughout the case and promptly correct any electrolyte disorders (especially hypokalaemia, hypomagnesaemia and hypocalcaemia).

Induction and Maintenance of General Anaesthesia

Evidence suggests that propofol and opioids, such as fentanyl, alfentanil and remifentanyl, are safe options for induction. In contrast, ketamine is not recommended because it may exacerbate haemodynamic adverse effects. A lidocaine bolus (1.5 mg/kg) before laryngoscopy may help to prevent catecholamine surges.

If muscle relaxants are required, rocuronium, vecuronium, atracurium and cisatracurium are safe. Succinylcholine and pancuronium may, however, cause undesired haemodynamic changes. Reversal agents with anticholinesterase activity should be used with caution. Sugammadex used to reverse aminosteroid muscle relaxation is potentially a good option.

For maintenance of general anaesthesia, total IV anaesthesia with propofol is a good choice, although the QT prolongation by volatile anaesthetics (namely, isoflurane and sevoflurane) is probably clinically insignificant.

Regional Anaesthesia

Neuraxial and peripheral techniques are valid given their attenuation of the stress response and the effective analgesia they provide. However, adding epinephrine to the local anaesthetic solution is not advised. With regard to neuraxial anaesthesia, epidural or combined spinal-epidural anaesthesia might allow better titration and, as such, is preferred over spinal anaesthesia because of the propensity for spinal anaesthesia to rapidly drop blood pressure. Consider sedation in awake patients.

Analgesia

Perioperative pain should be anticipated and adequately prevented and treated. Painful stimuli might induce a number of psychological and physiological responses, including enhanced sympathetic activity. In addition to regional techniques, morphine, paracetamol and anti-inflammatory drugs are considered safe.

Antiemetics

Prophylactic antiemetic therapy is limited because of further prolongation of the QT interval by ondansetron and droperidol. As a result, dexamethasone, cyclizine and metoclopramide are preferred in this group of patients.

POSTOPERATIVE MANAGEMENT

All patients should be admitted to a postoperative care unit. Here, management should focus on the following:

- bed rest in a calm environment,
- continuous standard monitoring,
- adequate analgesia,
- control of postoperative nausea and vomiting and
- restarting chronic beta-adrenergic blockade in stable patients.

MANAGEMENT OF CRITICAL ARRHYTHMIAS

Arrhythmias can occur at any time during the perioperative period. Prolongation and dispersion of repolarisation provide the substrate for early after depolarisations, triggered activity and reentry mechanisms. These events range from auto-limited episodes of TdP to sustained ventricular fibrillation.

TdP is a polymorphic ventricular tachycardia, with sinusoidal and cyclic alteration of the QRS axis, so that the peaks of the QRS complex seem to twist around the isoelectric line, as depicted in Figure 4. QT prolongation of the preceding beat is a typical feature and occasionally T-wave alternans (beat-to-beat variation in the shape or amplitude of T waves), a marker of cardiac electrical instability, may be seen.

The torsadogenic potential of a drug is determined not only by prolongation of the QT interval but also by its ability to increase dispersion of repolarisation. This arrhythmia is usually short lived and terminates spontaneously; however, rapidly recurring episodes may degenerate into ventricular fibrillation and cardiac arrest. See Tables 4 and 5 for the prevention and treatment of LQTS and TdP.

The management of various TdP arrhythmia scenarios is as follows:

- Single episode and haemodynamically stable: Regardless of magnesium levels, IV magnesium sulphate is the first-line therapy and helps to prevent recurrent arrhythmias. A single dose of 2 g or 8 mmol (4 mL of 50% magnesium sulphate mixed with dextrose 5% in water to a total volume of 50-100 mL) over 15 minutes is recommended. The mechanism is still unknown, but enhanced potassium efflux and inhibition of late calcium influx are suggested. Potential side effects of hypermagnesaemia include enhanced neuromuscular blockade and respiratory depression as a result.
- Multiple self-terminating episodes and haemodynamically stable: Besides IV magnesium, consider temporary transvenous overdrive atrial pacing (set to 100-140 beats per minute) and/or IV isoproterenol (also known as isoprenaline) infusion: start at 2 µg/min and titrate to a heart rate of 100 beats per minute. Both strategies aim to reduce the RR interval and repolarisation time.
- Haemodynamic instability: Haemodynamically unstable patients due to ventricular arrhythmia require prompt synchronised electrical cardioversion, as well as 2 g IV magnesium.
- Pulseless arrhythmia: If TdP degenerates into ventricular fibrillation, commence cardiopulmonary resuscitation and attempt immediate defibrillation; 2 g IV magnesium should be administered. Avoid amiodarone as it has a proarrhythmic effect due to the additional prolongation of the QTc interval. Give IV lidocaine instead.

Given the likelihood of arrhythmia recurrence, continuous monitoring (consider self-adhesive multifunction pads) and arrhythmia prevention and treatment are mainstay during the rest of the hospital stay.

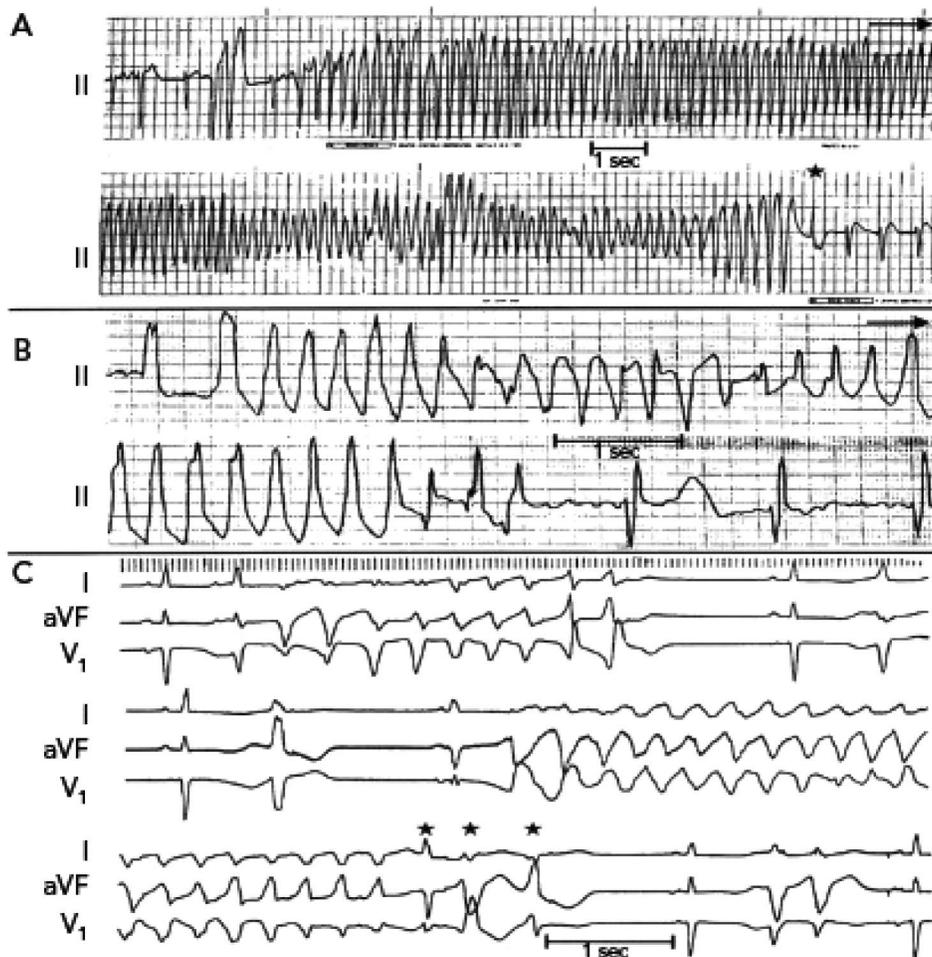


Figure 4. Three electrocardiographic examples of long QT syndrome and torsades de pointes.¹⁵ Permission granted by Nabil E. El-Sherif, MD. This image/content is covered by the terms of the Creative Commons license (the CC-BY-NC 4.0) of this publication.

General Preventive Measures		
Avoidance of QT-prolonging drugs and correction of electrolyte abnormalities After implementing treatment, physical activity can continue recreationally		
Management of LQTS		
Asymptomatic	Symptomatic	
Congenital LQTS Beta-blocker (propranolol or nadolol) If extremely prolonged QTc: consider prophylactic ICD	Beta-blocker (propranolol or nadolol) If previous SCA: ICD Recurrent arrhythmias despite maximally tolerated doses of beta-blocker: mexiletine, LCSD and/or ICD	
Acquired LQTS Decisions regarding use of QT-prolonging drugs based on individualized risk-benefit analysis Monitoring as an outpatient	Decisions regarding use of QT-prolonging drugs based on individualized risk-benefit analysis Telemetry observation during withdrawal of toxic agent	

Table 4. Management of Patients With LQTS. LQTS indicates long QT syndrome; ICD, implantable cardioverter defibrillator; LCSD, left cardiac sympathetic denervation; SCA sudden cardiac arrest

Haemodynamically Stable
Single episode IV magnesium (2-g bolus) Correction of electrolyte abnormalities Removal of QT-prolonging drugs With multiple self-terminating episodes IV magnesium (2-g bolus) Temporary transvenous overdrive atrial pacing (100-140 bpm) and/or IV isoproterenol infusion (initially 2 µg/min, titrated to 100 bpm)
Haemodynamically Unstable
Electrical cardioversion IV magnesium (2-g bolus)
Pulseless
Defibrillation as per European Resuscitation Council guidelines (consider lidocaine over amiodarone) IV magnesium (2-g bolus)

Table 5. Acute Management of TDP

SUMMARY

QT prolongation is a frequent phenomenon in the perioperative setting. Although most patients have an unremarkable course, some high-risk patients may develop malignant arrhythmias such as TdP. Table 6 summarises steps to be taken in the perioperative management to prevent these events.

Category	Action
Preoperative assessment	Check 12-lead ECG, if QTc >480 ms, enquire about personal and family history of LQTS syndrome; refer for cardiology evaluation and optimisation if high risk Risk-benefit analysis of QT-prolonging drugs; hold off if possible Continue chronic beta-blocker therapy
Premedication	Promote a calm environment; consider midazolam for anxiolysis
Equipment and monitors	Standard monitoring, reduce alarm sounds (loud noises are possible triggers for congenital LQTS), and ensure immediate availability of IV magnesium and a defibrillator; consider self-adhesive multifunction pads
Regional anaesthesia	Consider neuraxial (epidural) and peripheral techniques; avoid adding epinephrine
General anaesthesia	
Induction	Opioid: fentanyl, alfentanil, remifentanyl (avoid sufentanyl) Induction agent: propofol (avoid thiopental, ketamine) Neuromuscular blocking agent: rocuronium, vecuronium, atracurium, cisatracurium (avoid pancuronium or succinylcholine) Consider lidocaine before laryngoscopy
Maintenance	Ventilation: avoid hypoxaemia, hypocapnia, hypercapnia and high ventilatory pressures Circulation: prevent sudden changes in heart rate and blood pressure Other: ensure normothermia, electrolyte homeostasis, adequate analgesia Maintenance agent: Total intravenous anaesthesia with propofol is preferred; cautious use of halogenated volatile anaesthetics Analgesia: consider regional techniques as an opioid (and emesis)-sparing strategy Antiemesis: choose dexamethasone and metoclopramide over ondansetron and droperidol
Postanaesthetic care unit	Bed rest, calm environment, adequate analgesia, monitor postoperative arrhythmias; consider low-dose propofol if nausea/vomiting

Table 6. Summary of Perioperative Management

REFERENCES

1. Schwartz P, Stramba-Badiale M, Crotti L et al. Prevalence of the congenital long-QT syndrome. *Circulation*. 2009;120:1761-17677.
2. Yu H, Zhang L, Liu J et al. Acquired long QT syndrome in hospitalized patients. *Heart Rhythm*. 2017;14:974-978.
3. Nagele P, Pal S, Brown F et al. Postoperative QT interval prolongation in patients undergoing noncardiac surgery under general anesthesia. *Anesthesiology*. 2012;117:321-328.
4. Bazett H. An analysis of the time-relations of electrocardiograms. *Heart*. 1920;7:353-370.
5. Vink A, Neumann B, Lieve K, et al. Determination and Interpretation of the QT interval. Comprehensive analysis of a large cohort of long QT syndrome patients and controls. *Circulation*. 2018;138:2345-2358.
6. Priori S, Blomström-Lundqvist C, Mazzanti A, et al. 2015 ESC fines for the management of patients with ventricular arrhythmias and the prevention of sudden cardiac death. *Eur Heart J*. 2015;36:2793-2867.
7. Al-Khatib S, LaPointe N, Kramer JM, et al. What clinicians should know about the QT interval. *JAMA*. 2003;289:2120-2127.
8. Taggart N, Haglund C, Tester D et al. Diagnostic miscues in congenital long-QT syndrome. *Circulation*. 2007;115:2613-2620.
9. Schwartz P, Spazzolini C, Crotti L, et al. The Jervell and Lange-Nielsen syndrome: natural history, molecular basis, and clinical outcome. *Circulation*. 2006;113:783-790.
10. Yang T, Roden D. Extracellular potassium modulation of drug block of IKr. Implications for torsade de pointes and reverse use-dependence. *Circulation*. 1996;93:407-411.
11. CredibleMeds®. Search for Drugs that Prolong QT & induce Torsades de Pointes (TdP). Accessed on July 31, 2020. <https://www.crediblemeds.org/index.php/drugsearch>
12. Moss A, Schwartz P, Crampton R, et al. The long QT syndrome: prospective longitudinal study of 328 families. *Circulation*. 1991;84:1136-1144.
13. Ikonnikov G, Yelle D. Cardiac conducting system. Accessed November 8, 2020. <http://www.pathophys.org/physiology-of-cardiac-conduction-and-contractility/>
14. Giudicessi J, Ackerman M. Calcium revisited new insights into the molecular basis of long-QT syndrome. *Circ Arrhythm Electrophysiol*. 2016;9:e002480.
15. El-Sherif N, Turitto G, Boutjdir M. Acquired long QT syndrome and electrophysiology of torsade de pointes. *Arrhythm Electrophysiol Rev*. 2019;8:122-130.



This work by WFSA is licensed under a Creative Commons Attribution-NonCommercial-NoDerivatives 4.0 International License. To view this license, visit <https://creativecommons.org/licenses/by-nc-nd/4.0/>

WFSA Disclaimer

The material and content provided has been set out in good faith for information and educational purposes only and is not intended as a substitute for the active involvement and judgement of appropriate professional medical and technical personnel. Neither we, the authors, nor other parties involved in its production make any representations or give any warranties with respect to its accuracy, applicability, or completeness nor is any responsibility accepted for any adverse effects arising as a result of your reading or viewing this material and content. Any and all liability directly or indirectly arising from the use of this material and content is disclaimed without reservation.