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Long-QT syndrome

orphan anesthesia

a project of the German Society
of Anaesthesiology and Intensive Care Medicine

SUPPLEMENT NR. 1 | 2026

OrphanAnesthesia –
ein krankheitsübergreifendes Projekt des Wissenschaftlichen Arbeitskreises Kinderanästhesie der Deutschen Gesellschaft für Anästhesiologie und Intensivmedizin e.V.

Ziel des Projektes ist die Veröffentlichung von Handlungsempfehlungen zur anästhesiologischen Betreuung von Patientinnen und Patienten mit seltenen Erkrankungen. Damit will OrphanAnesthesia einen wichtigen Beitrag zur Erhöhung der Patientensicherheit leisten.

Patientinnen und Patienten mit seltenen Erkrankungen benötigen für verschiedene diagnostische oder therapeutische Prozeduren eine anästhesiologische Betreuung, die mit einem erhöhten Risiko für anästhesieassoziierte Komplikationen einhergehen. Weil diese Erkrankungen selten auftreten, können Anästhesistinnen und Anästhesisten damit keine Erfahrungen gesammelt haben, sodass für die Planung der Narkose die Einholung weiterer Information unerlässlich ist. Durch vorhandene spezifische Informationen kann die Inzidenz von mit der Narkose assoziierten Komplikationen gesenkt werden. Zur Verfügung stehendes Wissen schafft Sicherheit im Prozess der Patientenversorgung.

Die Handlungsempfehlungen von OrphanAnesthesia sind standardisiert und durchlaufen nach ihrer Erstellung einen Peer-Review-Prozess, an dem eine Anästhesistin bzw. ein Anästhesist sowie eine weitere Krankheitsexpertin bzw. ein weiterer Krankheitsexperte (z. B. Pädiaterin bzw. Pädiater oder Neurologin bzw. Neurologe) beteiligt sind. Das Projekt ist international ausgerichtet, sodass die Handlungsempfehlungen grundsätzlich in englischer Sprache veröffentlicht werden.

Ab Heft 5/2014 werden im monatlichen Rhythmus je zwei Handlungsempfehlungen als Supplement der A&I unter www.ai-online.info veröffentlicht. Als Bestandteil der A&I sind die Handlungsempfehlungen damit auch zitierfähig. Sonderdrucke können gegen Entgelt bestellt werden.

Bisher in A&I publizierte Handlungsempfehlungen finden Sie unter:

www.ai-online.info/Orphsuppl
www.orphananesthesia.eu

OrphanAnesthesia –
a project of the Scientific Working Group of Paediatric Anaesthesia of the German Society of Anaesthesiology and Intensive Care Medicine

The target of OrphanAnesthesia is the publication of anaesthesia recommendations for patients suffering from rare diseases in order to improve patients' safety. When it comes to the management of patients with rare diseases, there are only sparse evidence-based facts and even far less knowledge in the anaesthetic outcome. OrphanAnesthesia would like to merge this knowledge based on scientific publications and proven experience of specialists making it available for physicians worldwide free of charge.

All OrphanAnesthesia recommendations are standardized and need to pass a peer review process. They are being reviewed by at least one anaesthesiologist and another disease expert (e.g. paediatrician or neurologist) involved in the treatment of this group of patients.

The project OrphanAnesthesia is internationally oriented. Thus all recommendations will be published in English.

Starting with issue 5/2014, we'll publish the OrphanAnesthesia recommenations as a monthly supplement of A&I (Anästhesiologie & Intensivmedizin). Thus they can be accessed and downloaded via www.ai-online.info. As being part of the journal, the recommendations will be quotable. Reprints can be ordered for payment.

Find a survey of the recommendations published until now on:

www.ai-online.info/Orphsuppl
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Anaesthesia recommendations for

Long-QT syndrome

Disease name: Long-QT syndrome

ICD 10: I49.8

ICD 11: BC65.0

ORPHA:768, Alpha-SE ID I117711

Synonyms: LQTS

Disease summary:
Long-QT syndrome (LQTS) is characterized by a prolonged, heart-rate corrected QT (QTc) interval in the ECG. Typical QTc values are >480 ms. Two different forms of LQTS are known: an inherited and thereby often familial form of LQTS (congenital form / cLQTS) and the acquired, often reversible form (aLQTS). For typical ECG recordings see references [1, 7, 8, 9, 12]. The disease is characterized by a dysfunction of cardiac ion channels responsible for myocellular repolarization. In consequence, patients with either cLQTS or aLQTS have an increased risk of polymorphic ventricular tachyarrhythmias of the so-called Torsades de Pointes type (TdP).
An estimated 10% of sudden infant deaths and 20% of unexplained adult deaths are thought to be due to LQTS.
The congenital form has an estimated prevalence of 1: 2,000 and is caused by pathogenic variants in cardiac ion channel genes that directly lead an altered ion channel function due to ion current reduction or protein-protein interactions influencing correct ionic current flow. There are >10 genetic subforms known; the LQT1, LQT2 and LQT3 are the most common ones.
The most common subform is LQT1 (42-54%) where TdP are often triggered by high physical exercise (e.g., swimming) or emotional stress, i.e. adrenergic conditions. In contrast, in the second most common type, LQT2 (35-45%), known arrhythmia triggers are rest, emotions, acoustic triggers, and physical stress. In the subform LQT3, resembling 7-8% of all, possible triggers are rest or sleep, vagotonic conditions.
The first clinical manifestation, often syncope, may occur in childhood or young adulthood due to exposure to triggers. In the majority (95%), the ventricular arrhythmia is self-terminating and spontaneously converts into sinus rhythm. In a minority, however, TdP may degenerate in ventricular fibrillation and cardiac arrest, fatally without outside help. The diagnosis is often difficult and requires precise ECG assessment and genetic diagnostics. This is beyond the scope of this short review.
In LQT1, β-receptor blockers are effective in preventing clinical events. This effect is reduced in LQT2 patients and only slightly pronounced in LQT3 where sodium channel blocking agents such as mexiletine might be effective. Left cardiac sympathetic denervation (LCSD) is an ultimate and additive therapy to β-receptor-blockers. The use of an ICD for high-risk patients is recommended, but should not replace β-receptor blocker therapy, life-style modification and trigger prevention.

▶ **Citation:** Graf M, Kleinschmidt S: Long-QT syndrome. Anästh Intensivmed 2026;67:S1–S8.
DOI: 10.19224/ai2026.S001

Acquired LQTS is typically reversible after correction of the acquired, clinical reason for impairment of myocardial repolarization, e.g. by ion channel blockade or dysregulation. Many reasons may underlie and include severe electrolyte disturbances, endocrine disorders, structural heart disease and by the administration of QT-prolonging drugs (see www.crediblemeds.org) that finally trigger TdPs [3]. The condition is potentially reversible by cessation of the cause impairing myocardial repolarization; the surface ECG then might normalize. The triggering medication should be strictly avoided and a prolonged effect of the medication due to inhibition of the degradation pathways (cytochrome inhibition) should also be taken into account. The presence of liver failure, heart failure and female gender make the occurrence of TdP more likely. The most effective way to prevent the occurrence of aLQTS is to avoid triggering drugs and, if necessary, to stop the intake.

Medicine is in progress

Perhaps new knowledge

Every patient is unique

Perhaps the diagnosis is wrong

Find more information on the disease, its centres of reference and patient organisations on Orphanet: www.orpha.net

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Emergency information

A	**AIRWAY / ANAESTHETIC TECHNIQUE**	No specific recommendations beside standard care. A difficult airway may be anticipated in the case of co-morbidity.
B	**BLOOD PRODUCTS (COAGULATION)**	No specific recommendations.
C	**CIRCULATION**	Sudden onset of ventricular tachycardia as Torsades de Pointes (TdP).
D	**DRUGS**	- The administration of droperidol, ketamine, succinylcholine, epinephrine and pancuronium is contraindicated. - In general, drugs that cause CYP3A4 inhibition (e.g. cimetidine, ketoconazole, clarithromycine, also grapefruit juice in the nutrition of patients!) should be avoided [7, 8]. - No known risk of MH to date. - Avoid the use of sympathomimetics and cholinesterase inhibitors such as flecainide, sotalol, amiodarone, cafedrine/theodrenaline and neostigmine [1, 4, 7, 8]. - Avoid volatile anaesthetics, prefer TIVA. - In the peripartum period, be aware of arrhythmogenic effects of tocolytic and uterotonic drugs such as fenoterol or oxytocine [6]. Please avoid if possible.
E	**EQUIPMENT**	Ensure postoperative surveillance on IMC or ICU.

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Typical surgery and procedures

Apart from general operations, typical surgical procedures in LQTS patients may include insertion of implantable cardioverter defibrillators and pacemakers.

Type of anaesthesia

General and regional anaesthesia techniques (including a combination of both) are required in accordance with the nature of surgery or intervention (e.g. local anaesthesia in dental procedures) and postoperative pain therapy.

Necessary additional pre-operative testing (beside standard care)

A detailed family history, especially focussing on sudden deaths of young, healthy relatives is an essential part of preoperative evaluation. A prolonged QT interval is defined as a heart rate-corrected QT interval (QTc) of >450 ms in males and >460 ms in females. The European Society of Cardiology suggests using a QTc of ≥480 ms for diagnosing LQTS and using a QTc of 460-479 as a borderline range where a diagnosis may be considered (see references 6, and 16). Anaesthesiologists must be aware of pre, intra and postoperative ECG changes in respect to the diagnostic criteria of LQTS. If LQTS has already been diagnosed, a cardiac consultation should be performed and potential arrhythmia triggering factors have to be determined; the potential LQTS genotype might be evaluated due to different prognostic and clinical features. The preoperative implantation of an ICD device may be necessary after cardiologic consultation [1, 7, 8, 9, 11]. Discontinue QT-prolonging medication (since contraindicated), if justifiable and continue β-blocking therapy even on the day of surgery. In the laboratory panel, determine electrolyte levels (Potassium, magnesium and calcium are of highest importance), which should be at the higher normal reference range and may need preoperative substitution. Administration of anxiolytic premedication is recommended (e.g., midazolam), but please pay attention to possible drug interactions.

Particular preparation for airway management

There are no pre-existing associations of patients with LQTS and difficult airway unless other comorbidities exist (e.g. Andersen-Tawil-Syndrome (LQT7) with retrognathia, hypertelorism). The standard guidelines for the management of an expected or unexpected difficult airway are applied. An adequate level of anaesthesia and analgesia should be warranted in order to prevent any sympathetic activation and potential hazardous tachyarrhythmias.

Particular preparation for transfusion or administration of blood products

There are no specific recommendations regarding the administration of blood products in Patients with LQTS. In general, maintenance of haemostasis and tissue oxygen delivery is essential, especially in patients with significant comorbidities. Potential electrolyte imbalances (e.g., calcium, potassium, magnesium) as a side effect of e.g., packed red cell transfusion have to be considered.

Particular preparation for anticoagulation and/or platelet inhibition

No recommendation for anticoagulation or antiplatelet therapy in patients with LQTS have yet been published. No coagulopathies have been described in a close association with LQTS. The use of anticoagulants or antiplatelet drugs is recommended due to specific co-morbidities and types of surgery in accordance with current guidelines.

Particular precautions for positioning, transportation and mobilisation

There are no specific recommendations for positioning, transportation and mobilization of patients with LQTS. Standard monitoring in accordance with guidelines maintaining homeostasis and effective stress protection (including noise reduction) are the cornerstones of adequate care of patients [9].

Interactions of chronic disease and anaesthesia medications

During the perioperative period, drugs known to prolong the QT interval with the consequence of induction of Torsades de Pointes (TdP) must be avoided. In the case of polypharmacy, possible interactions have to be considered.
In brief, the following anaesthetics or adjuvants should be strictly avoided: Ketamine, succinylcholine, pancuronium, droperidol and epinephrine. An adequate access to an up to date list of “critical drugs” is mandatory, e.g. at https://crediblemeds.org/healthcare-providers

Anaesthetic procedure

Obtain anaesthesia induction in a strictly quiet environment using propofol or thiopentone with QT interval monitoring with at least a 5-lead ECG. Consider a transient pacemaker implantation for emergency pacing.
Take care of excessive sympathetic stimulation. Ensure an adequate depth of anaesthesia. If necessary, the administration of a short-acting β-receptor blocker (e.g. esmolol, landiolol) is advisable.
Avoid epinephrine as an adjunct to local anaesthetics for regional anaesthesia procedures. Prefer dexamethasone for a prolongation of the duration of nerve blocks for pain therapy.
For maintenance of anaesthesia preferably propofol (via TIVA or TCI mode) may be applied. Volatile anaesthetics should be used with caution. Use fentanyl or sufentanil for analgesia, other opioids are also adequate. For muscle relaxation, vecuronium, (cis)atracurium or rocuronium seem to be save. The use of neuromuscular monitoring is recommended. Cholinesterase inhibitors for antagonism of neuromuscular blockade may cause bradycardia. The use of Sugammadex seems to be without relevant side effects [9].
Intraoperatively, high ventilation pressures and recruitment manoeuvres should be avoided as far as possible, as these can lead to bradycardia due to a reduction in venous return flow.
Flecainide, sotalol and amiodarone are contraindicated in patients of LQTS and should be strictly avoided in the treatment of intra- or postoperative tachycardia. The same refers to systemic application of epinephrine or cafedrine-theodrenalin (Akrinor®).

Particular or additional monitoring

The need for invasive monitoring (e.g. arterial and central venous line) is determined by the type of surgery and co-existing diseases. The threshold to the indication for invasive monitoring in patients with LQTS should be low in order to ensure an adequate diagnosis and therapy in the case of malignant tachyarrhythmias, enable the detection of electrolyte imbalances via laboratory analysis or monitoring of volume status. Enable continuous ECG monitoring and availability of a defibrillator and adhesive defibrillation paddles during the whole perioperative period.

Possible complications

Despite all precautions taken, the occurrence of TdP or PVT is possible. The episodes are usually short and self-limiting. However, transitions to ventricular fibrillation are possible.

If TdP occurs, magnesium sulphate is the first-line treatment. For adult patients, an initial bolus of 30 mg/kg is recommended, followed by an infusion of 2-4 mg/min. Pulseless TdP must be treated according to current Advanced Cardiac Life Support guidelines and algorithms including the use of epinephrine [10, 13]. Episodes of TdP associated with persisting bradycardia despite magnesium sulphate administration, a transvenous pacemaker is a therapeutic option.

Post-operative care

In the post-operative period, continuous ECG monitoring in a high dependency ward with adequate emergency resuscitation equipment is recommended. Strictly obtain a maximal noise reduction e.g., by reducing sound levels of monitor alarms. If possible, a separate patient box for post anaesthesia care is advisable. Effective pain control (e.g. with regional anaesthesia and/or with combined opioid/non-opioid agents in accordance with current guidelines), of normothermia should be part of the therapeutic strategy to avoid or at least significantly reduce sympathetic stimulation and emergent agitation also during patient transfers. In the case of paediatric surgery, the consultation and the availability of a paediatric cardiologist and postoperative care in a specialized paediatric intensive care unit is obligatory or at least highly recommended [11]. Laboratory abnormalities (e.g. potassium, magnesium, calcium, glucose, pH) should be monitored and must be corrected immediately. Polypharmacy has to be avoided, especially in respect to QTc prolongation or “torsadogenicity” of various drugs (see https://crediblemeds.org/healthcare-providers, [3]). Beta-receptor blocking medication should be continued if part of the routine treatment.

Disease-related acute problems and effect on anaesthesia and recovery

The occurrence of TdP and PVT is possible. See chapter “possible complications”.

Ambulatory anaesthesia

Recommendations or guidelines for ambulatory anaesthesia do not exist to date. Due to the nature of the underlying disease, the variety of surgical procedures and the various challenges in the whole perioperative period, ambulatory discharge cannot be recommended. The general advantages of early mobilisation have to be considered. Implantable cardiac devices should be restored to their baseline settings before discharge.

Obstetrical anaesthesia

A variety of case reports exist for obstetric anaesthesia in patients with LQTS. For elective Caesarean Section (CS), regional anaesthesia techniques, e.g. epidural or combined spinal epidural anaesthesia (CSE) may be beneficial because of an adequate stress response with only gradual alterations of arterial blood pressure [2]. If general anaesthesia is mandatory, e.g. in the case of emergency CS, an adequate management of stress response and minimal alteration of QT interval during and after rapid sequence induction may be achieved with propofol, opioids, rocuronium, and maintenance of anaesthesia with propofol in favour of volatile anaesthetics. For the prevention of postoperative nausea and vomiting, dexamethasone alone or in combination with ramosetrone or ondansetrone in doses according to current guidelines seem to be the agents of choice [5]. In the case of uterine atony, uterotonic drugs such as oxytocine should be used with caution [6] and their application needs an interdisciplinary consensus between anaesthesiology and obstetrics during the whole peripartum period. The same refers to the use of tocolytics such as fenoterol.

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