Anaesthesia recommendations for patients suffering from

Long QT syndrome

**Disease name:** Long QT syndrome

**ICD 10:** I45.8

**Synonyms:** LQTS

**Related syndromes:** Romano-Ward S., Jervell and Lange-Nielsen S., Timothy S.

Long QT syndrome (LQTS) is a cardiac disorder resulting from malfunction of cardiac ion channels. It can be divided in congenital (c-LQTS) and acquired (a-LQTS) forms. Typical is a prolongation of the QT interval on ECG and a propensity to ventricular tachyarrhythmias which can lead to a characteristic polymorphic ventricular tachycardia known as “torsades de pointes”. There is a risk of syncope, cardiac arrest or sudden death.

The diagnosis is based on ECG findings, clinical and family history. The Schwartz score defines the probability of LQTS according to three categories: (1) point, low probability of LQTS; (2) 2 or 3 points, intermediate probability of LQTS; and (3) ≥3.5 points, high probability of LQTS.

**THE TABLE OF THE SCHWARTZ SCORE** should be added here:

Table 2 of the paper Schwartz PJ and Crotti L. QTc behavior during exercise and genetic testing for the Long-QT Syndrome: Circulation 2011;124:2181-2184.

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Medicine in progress

Perhaps new knowledge

Every patient is unique

Perhaps the diagnostic is wrong

Find more information on the disease, its centres of reference and patient organisations on Orphanet: [www.orpha.net](http://www.orpha.net)
Typical surgery

The typical surgery for LQTS is left cardiac sympathetic denervation, which is recommended in patients symptomatic for cardiac events despite beta-blocker (BB) therapy or in patients in whom BBs are contraindicated or not tolerated. For therapeutic reasons the implantation of a cardioverter defibrillator (ICD) is recommended in patients with a previous cardiac arrest or in patients with recurrent syncopal events despite antiadrenergic interventions. Only exceptionally, asymptomatic patients should be implanted with an ICD, and are those with very prolonged QT interval and signs of high electrical instability. Cardiac events in LQTS can occur at any age, mostly in childhood but also from in utero up to adulthood.

Type of anaesthesia

Patients with LQTS can receive either general or regional anaesthesia. Despite the chosen technique the major issue should be the complete avoidance of any stress or sympathetic activation. There are only few reports about regional anaesthesia but spinal anaesthesia has been successful used. During caesarean section epidural anaesthesia with or without morphine has been used. Epinephrine must be avoided as an adjuvant medication in regional anaesthesia because it could favour life-threatening arrhythmias. Atropine should also always be avoided because of its effect on heart rate and because it removes the cholinergic-adrenergic antagonism.

Necessary additional diagnostic procedures (preoperative)

- The diagnosis of LQTS in mainly clinical and is based on ECG measurements, and on the collection of personal and family history. The Schwartz score used for the diagnosis has already been discussed. A comprehensive overview is given on [http://en.wikipedia.org/wiki/Long_QT_syndrome](http://en.wikipedia.org/wiki/Long_QT_syndrome). Whenever the LQTS is acquired, the causes of QT prolongation should be corrected before surgery and anaesthesia.

- The anamnestic talk with the patient or care taker should include questions about family and personal history for cardiac events and possible triggers of LQT-related arrhythmias, like swimming, running or startle like loud alarm horns, ringing phone as well as stressful emotions like anger or crying.

- Additionally, a genetic evaluation should be performed. For clinical purposes KCNQ1, KCNH2 and SCN5A should be screened according to current consensus documents.

- Before anaesthesia laboratory examination of the blood should focus on K⁺. Potassium levels <4.0 mmol should be corrected to normal or “supranormal” values (4.5-5.5 mmol)

Particular preparation for airway management

No special preparation for airway management is necessary in patients with LQTS. Every manipulation during intubation must consider complete stress reduction requiring adequate anaesthesia before intubation.
Anaesthesiologic procedure

A comprehensive and structured list of recommended or restricted drugs is available online (see literature and Internet links).

Premedication
A sedative premedication preferably with midazolam to blunt the sympathetic response is mandatory. Drugs that should be avoided for premedication include: Chloral hydrate, Droperidol and Ketamine. Anticholinergic adjuvants like Atropine can precipitate torsade de points and should not be given in LQTS patients. If already diagnosed the patient should be under b-blockade, which has to be continued during the operative procedure.

Induction of anaesthesia
Induction of anaesthesia should be made with special care of a quiet environment. Loud noises or a scaring atmosphere must be avoided. A prophylactic treatment with magnesium sulphate can be considered. Defibrillator paddles should be attached or at least a defibrillator should be close by. If the parents could be of help during induction of anaesthesia providing a familiar atmosphere must be considered individually according to the judgment of the anaesthesiologist. If possible iv-induction should be considered as first choice.

Volatile anaesthetics
All of the clinical available inhalation agents could be considered in patients with LQTS except Halothane and Desflurane. Halothane is known to sensitize the heart to catecholamines and Desflurane causes a sympathetic activation and QT interval prolongation. Among the others, Isoflurane has been reported as the agent of choice because of its apparent safety.

Nitrous oxide
Nitrous Oxide has been used in conjunction with other inhalation agents without adverse events.

Intravenous anaesthetics
If no contraindications Propofol seems to be the agent of choice in patients with LQTS because of little or no effect on the QT-interval. Ketamine should be avoided in any case because of its central sympathetic activation.

Opiates
Morphine as well as fentanyl are used safely in patients with LQTS although there are conflicting reports on the effect on the QT-interval. The advantages of stress suppression during endotracheal intubation must be considered when thinking of pro´s and con´s. Sufentanil is known to prolong the QT-interval in adult patients. There is one report of Kweon and co-workers that the administration of remifentanil before intubation can prevent the prolongation of the QTc interval associated with tracheal intubation during induction of anaesthesia with sevoflurane.

Muscle relaxants
Succinylcholin is obviously not recommended any more in paediatric anaesthesia as well as in patients with long QT-syndrome. Vecuronium and Atracurium are reported to be safe in those patients. Rocuronium was reported to be safe in one patients undergoing c-section. Pancuronium should be considered with special care because of its vagolitic properties and
is not the agent of first choice especially as there are alternatives as mentioned above. Special care should be taken before using any neuromuscular reversal agent because the combination of anticholinergic agents like atropine with anticholinesterases is prolonging the QT-interval.

Local anaesthetics
Local anaesthetics (lidocaine and chloroprocaine) can be safely used, avoiding addition of epinephrine. Spinal as well as epidural anaesthesia was successful used in patients with LQTS. Lidocaine and chloroprocaine are used in these procedures with and without additional morphine. Levobupivacaine was thought to be of favour according to the racemic bupivacaine to decrease the risk of drug-induced cardiotoxicity.

Termination of anaesthesia
The same basic principles for termination of anaesthesia in patients with LQTS are of importance as compared to induction of anaesthesia. A calm, quiet, and relaxed atmosphere will prevent cardiac arrhythmias due to stress reduction. If experienced, the anaesthesiologist could consider to extubate the child while still sleeping but with sufficient spontaneous breathing. Before, during and after anaesthesia the patient should kept warm. Hypothermia of the patient has to be strictly avoided because it will prolong QT-interval. It is mandatory to ensure adequate pain control to avoid physical stress.

Particular or additional monitoring
The ECG is part of the main monitoring before during and after anaesthesia. It is necessary to record a baseline ECG before induction of anaesthesia to document the baseline QT-interval. Every change of the QT-interval during anaesthesia can now be compared to baseline value. A continuous perioperative QT-Interval monitoring is possible and recommended (Pfizenmayer and Friederich 2010, Anton and Friederich 2010) until normalization of the qt-interval in the recovery room.

Possible complications
Possible complications are life-threatening arrhythmias that could cause a sudden cardiac arrest. Because of this possibility, the patient should be continuously monitored with an ECG and a defibrillator should always be available as well as magnesium sulphate to treat arrhythmias. To reduce the probability of life-threatening arrhythmias avoidance of sympathetic stimulation and correction of any abnormal electrolyte should be performed.

Postoperative care
Postoperative care should focus on stress reduction. That means an adequate stress reduction with sufficient pain therapy is needed. After the surgical procedure at least one check of blood potassium should be made and corrected if necessary. ECGs should be performed on a regular basis. Avoid QT-prolonging antiemetic agents like Droperidol or Ondansetron. Dexamethason and Dimenhydrinat can be used alternatively.
Ambulatory anaesthesia

Ambulatory anaesthesia or surgery is not recommended in patients with LQTS. Despite some reports that even dental care should be done in hospitals under general anaesthesia to ensure complete stress reduction in children with LQTS, in reality this is almost never necessary. What matters is to avoid vasoconstrictor agents and to have a reassuring behaviour, besides limiting pain as much as possible.

Obstetrical anaesthesia

Obstetrical anaesthesia can be done in spinal, epidural or general anaesthesia with the limitations mentioned above.
Literature and internet links

1. Atlee JL 2001 Cardiac arrhythmias: drugs and devices. Curr Opin Anaesthesiol 14:3-9
11. Kim DH, Kweon TD, Nam SB, Han DW, Cho WY, Lee JS 2008 Effects of target concentration infusion of propofol and tracheal intubation on QTc interval. Anaesthesia 63:1061-1064

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