

Anaesthetic Guideline for the management of children with long QT Syndrome

SOP/Protocol Detail

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Aims

To ensure the safety of paediatric patients with congenital or acquired long QT syndrome who need a general anaesthetic. It is anticipated that most patients will undergo anaesthesia in Leeds and this guideline is to support those patients who do need to have a general anaesthetic at their local hospital.

Objectives

- 1.To provide guidance on the management of children with LQTS
- 2.To enable children to have minor surgery in their local hospital where appropriate
- 3.To improve equity and consistency in care across the Yorkshire and Humber CHD Network

Background

Long QT syndrome is a congenital or acquired channelopathy, impairing myocardial electrical conduction that results in impaired ventricular repolarization and can present clinically as recurrent syncope, pseudo-seizures, or sudden death. Patients with QT prolongation and LQTS are susceptible to the development of the characteristic polymorphic ventricular tachycardia, called **Torsades de Pointes TdP**.

The prolongation of the QT interval caused by anaesthetic drugs and the sympathetic response to anaesthesia and surgery can trigger malignant arrhythmias in patients with long QT syndrome.

Patients with a genetic predisposition to LQTS may be asymptomatic and **may have a normal resting QTc interval**, it is possible for an episode of torsade de pointes to occur for the first time during anaesthesia.

Diagnosis

The diagnosis of long QT syndrome should be made by a paediatrician, a paediatrician with an expertise in cardiology or a paediatric cardiologist.

QT intervals need to be corrected for heart rate (QTc – routinely defined using the Bazett formula) and are highly variable, but the abnormal corrected values are defined as a pre-puberty average of >470ms in males, >480ms in females. (adult values: males > 450ms, females >460ms)

Pre-op management

- Patient should be discussed with cardiologist and have a good quality ECG
- Patients with ICDs need a defibrillator check by a cardiac physiologist prior to theatre. This will ensure that the device will be set to correctly diagnose and treat arrhythmias during surgery if they occur.
- Medication for LQTS should be optimized prior to surgery
- Electrolytes (Potassium, Magnesium and Calcium) should be checked and corrected before anaesthesia to high normal levels
- Premedication is advised, as increased sympathetic activity caused by stress and anxiety can trigger TdP

Intra-op management

- Paediatric Cardiologist or Paediatrician with Expertise in Cardiology (PEC) in the unit where anaesthesia will occur should be contacted for advice and to be made aware of the patient
- For patients with devices, a cardiac physiologist does not need to be present during the induction or operation, but should be available to return for device support during the case if required.
- Premedication is advised, as stress/ anxiety can trigger arrhythmias
- Induction room should be calm and quiet as auditory triggers can cause TdP.
- Full non-invasive monitoring should be attached before induction
- For best drugs, see drugs list at end of guideline
- **The drugs in the “Avoid” list will prolong QT in all patients** and might cause TdP in patients with borderline QTc.
- Avoid hypothermia, hypoxia and hypercarbia
- Resuscitation equipment should be readily available in theatre

Treatment of TdP:

- Mostly self-limiting
- if sustained or haemodynamic compromise: asynchronous cardioversion
- Magnesium 30mg/kg as a slow bolus over 10min, then infusion at 10mg/kg/hr
- In bradycardia or in magnesium-resistant TdP, ventricular pacing at 90bpm or higher can be effective
- Deepen anaesthesia to decrease sympathetic activity
- If there is a need to discuss the anaesthetic management of an elective case with the Specialist Surgical Centre in Leeds please email j.scheffczik@nhs.net
- If urgent advice is required please call Leeds General Infirmary switchboard 0113 243 3144 and ask for the Paediatric Cardiac Anaesthetist or the Paediatric Cardiologist.

Post-op management

- Post-op monitoring dependent on case
- Analgesia and a calm environment essential
- No evidence for best practice length of monitoring or stay
- If there have been no arrhythmic issues with the ICD during anaesthesia/ surgery a further check prior to discharge will not be required.

Dental procedures
Dental procedures like extractions can use local anaesthetic, but Adrenaline as an additive should be avoided , because increase in sympathetic activity can trigger TdP ^{4,5} .
Conflicts of Interests
None
Provenance:
Author name(s)/Post and address: Dr Jutta Scheffczik, Paediatric Cardiac Anaesthetist, Leeds General Infirmary, Great George Street, Leeds LS1 3EX Clinical condition: Anaesthesia for Congenital Heart Disease Target patient group: Children with CHD Target professional group: Anaesthetists in non-specialist centres
Evidence Base:
References and Evidence levels: C. Expert consensus.

References:

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Addendum: (from O'Hare et al)**Perioperative medications**

Preferred Agents	
Sedation	
Midazolam	0.05-3mg/kg IV
Analgesia	
Lignocaine	1.5mg/kg IV
Fentanyl	2µ/kg IV
Alfentanil	0.5-3µ/kg/min IV
Remifentanyl	0.1-0.5 µ/kg/min IV
Morphine	0.05-0.1mg/kg IV
Volatile anaesthetics	
Isoflurane	1-3% minimum alveolar concentration
Neuromuscular blockers and reversal agents	
Rocuronium	0.6-1.2mg/kg IV
Vecuronium	0.04-0.1mg/kg IV for intubation
	0.8-1.2 µ/kg/min IV for maintenance
Cisatracurium	0.15-0.2mg/kg IV for intubation
	0.06-0.8mg/kg IV for maintenance
Spinal and epidural anaesthesia	
Epidural preferred over spinal	
Combined spinal-epidural anaesthesia	
Bupivacaine	Spinal 0.8-1.6ml of 0.75%
	Epidural 3-5ml boluses of 0.25-0.5%
Ropivacaine	Epidural 2-25ml of bupivacaine 0.5%

Exercise Caution	
Analgesia	
Buprenorphine	0.3mg IV
Methadone	0.1mg/kg IV (do not exceed 200mgs per day)
IV Anaesthetic Agents	
Propofol	6mg/kg/h IV
Etomidate	0.3mg/kg IV
Thiopental	2-6mg/kg IV
Volatile anaesthetics	
Sevoflurane	0.5-3% minimum alveolar concentration
Nitrous oxide	25-70% inspired
Neuromuscular blockers and reversal agents	
Anticholinesterase-anticholinergic reversal agents	
Spinal and epidural anaesthesia	
Combined spinal-epidural anaesthesia	
Oxytocin	
Postoperative care and antimetics	
Droperidol	0.625-1.25mg IV
Ondanestron	4 mg IV (do not exceed 16 mg)
Metoclopramide	10-20mg IV
Dexamethasone	0.1 mg/kg IV

Avoid	
Sedation	Neuromuscular blockers and reversal agents
Dexmedetomidine	Succinylcholine
Analgesia	Pancuronium
Epenephrine	Glycopyrrolate
Ketamine	Atropine
Sufentanil	Spinal Anaesthesia
	Spinal Anaesthesia to T10
	Epenephrine

Drugs to be avoided: (from <https://www.sads.org.uk/drugs-to-avoid/>)

Antiarrhythmics
Class 1: ajmaline*, cibenzoline*, dihydroquinidine*, disopyramide, encainide*, flecainide, mexiletine, pirmenol*, procainamide, propafenone quinidine* Class 3: almokalant*, amiodarone, azimilide*, bretylium, dofetilide*, dronedarone*, d-sotalol*, ersentilide*, ibutilide*, nifekalant*, sematilide*, sotalol, terikalant*
Anti-anginals/vasodilators
bepidil*, lidoflazine*, prenylamine*, ranolazine, terodiline*, vardenafil
Anti-Cancer
aclarubicin
Anti-hypertensives
indapamide, isradipine, moexipril/hydrochlorthiazide, nicardipine
Antihistamines
astemizole*, azelastine, diphenhydramine, ebastine*, hydroxyzine, terfenadine
Serotonin agonists and antagonists
cisapride*, dolasetron, granisetron, ketanserin*, ondansetron
Antimicrobials
<i>Macrolide antibiotics:</i> azithromycin, clarithromycin, erythromycin, roxithromycin*, spiramycin, telithromycin <i>Quinolone antibiotics:</i> ciprofloxacin, gatifloxacin*, gemifloxacin*, grepafloxacin*, levofloxacin, moxifloxacin, ofloxacin, sparfloxacin* <i>Antifungals:</i> cotrimoxazole, fluconazole (caution with itraconazole), ketoconazole, voriconazole <i>Others:</i> pentamidine, trimethoprim sulfa (bactrim) <i>Antiviral:</i> foscarnet (HIV)
Antimalarials
amantidine, chloroquine, halofantrine*, quinine
Psychiatric drugs
Tricyclic antidepressants: amitriptyline, amoxapine*, clomipramine, desipramine*, doxepin, imipramine, nortriptyline, protriptyline*, trimipramine Phenothiazines: chlorpromazine, fluphenazine, prochlorperazine, thioridazine*, trifluoperazine Others: atomoxetine, citalopram, clozapine, droperidol*, fluoxetine, haloperidol, levomethadyl*, lithium, maprotiline, mesoridazine, methadone, paroxetine, pericycline, pimozide, quetiapine, risperidone, sertindole, sertraline, trazodone, venlafaxine, zimeldine*, ziprasidone
Anticonvulsant
felbamate*, fosphenytoin (prodrug of phenytoin)
Anti-migraine
naratriptan, sumatriptan, zolmitriptan
Anti-cancer
arsenic trioxide, geldanamycin*, sunitib, tacrolimus, tamoxifen
Others
alfuzosin, chloral hydrate, clobutinol*, domperidone, galantamine, octreotide, organophosphates*, perflutren lipid microspheres, probucol, solifenacin, tizanidine, tolterodine, vasopressin
Stimulant drugs
Some cold remedies contain these drugs so it is important always to check the label. adrenaline (epinephrine), amphetamine, cocaine, dexmethylphenidate, dobutamine, dopamine, ephedrine, fenfluramine, isoprenaline (isoproterenol), levalbuterol, metaproterenol, methylphenidate, midodrine, norepinephrine (noradrenaline), phentermine, phenylephrine, phenylpropanolamine, pseudoephedrine, ritodrine, salbutamol (albuterol), salmeterol, sibutramine, terbutaline