Anaesthetic Management of Long QT Syndrome

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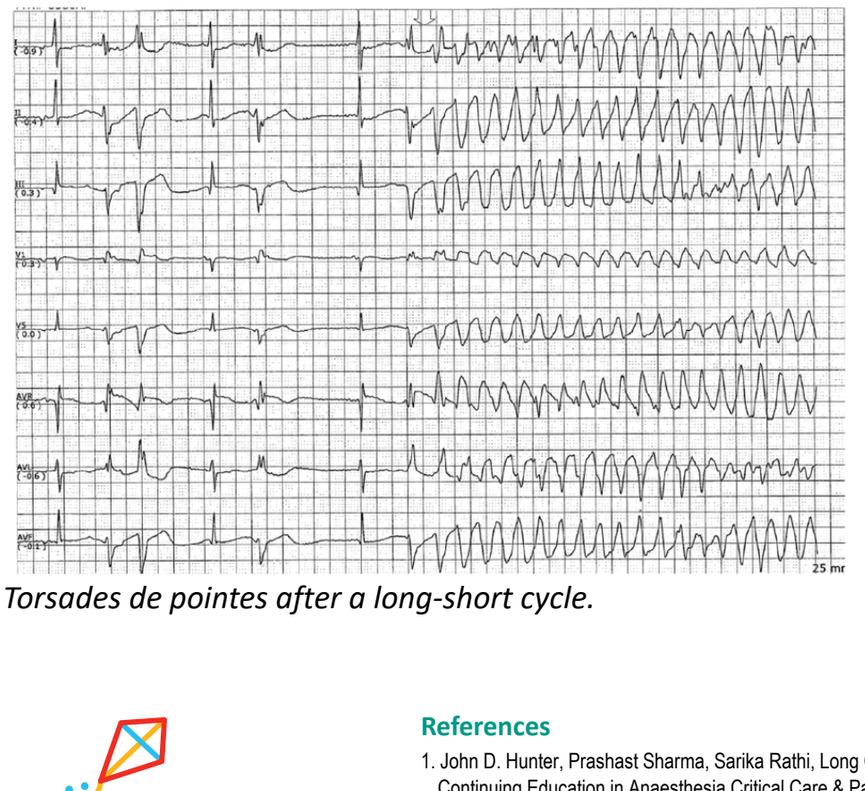
Background

- Long QT syndrome LQTS is a congenital or acquired channelopathy.
- Impairment of myocardial electrical conduction results in impaired ventricular repolarization
- Prolonged QT intervals need to be corrected for heart rate (QTc) and are highly variable, but defined as > 460ms prepuberty average, >470ms in boys, >480ms in girls (adult values: males > 450ms, females >460ms)
- Diagnosis of long QT syndrome should be made by a paediatrician with an expertise in cardiology or a paediatric cardiologist
- Several genetic mutations have been identified for this syndrome
- Clinical symptoms are recurrent syncope, pseudo-seizures, or sudden death, due to malignant arrhythmias
- Patients with QT prolongation and LQTS are susceptible to the development of a characteristic polymorphic ventricular tachycardia, called **Torsade de Pointes** (TdP)
- 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 TdP to occur for the first time during anaesthesia
- Acquired long QT can be due to medications, electrolyte disturbances (low magnesium, potassium, calcium), female sex (due to gonadal steroids), co-morbidities like heart disease or neurological injury, severe starvation, etc
- 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.

Torsade de Pointes

- Polymorphic ventricular tachycardia
- Twisting of QRS axis around iso-electric line
- Predictors of risk for TdP are:
  - Age (increased risk >40yrs)
  - Sex: pre-puberty: boys are more at risk than girls
    post-puberty: females more at risk that males
    >60yrs: equal risk
  - Genotype
  - Decreased short-term variability in QTc in genetic LQTS
  - Avoidance of triggers like increased sympathetic activity or QT prolonging drugs.



Torsades de pointes after a long-short cycle.

Pre-operative Management

- Patient should be discussed with cardiologist and have a good quality ECG
- 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-operative Management

- Paediatric Cardiologist or Paediatrician with Expertise in Cardiology (PEC) should be contacted for advice and to be made aware of patient
- Patients with ICDs need an electrophysiologist in theatre for safety of patient and device as well as for transvenous pacing if needed.
- Premedication is advised, as stress/ anxiety can trigger arrhythmias
- Induction room should be calm and quiet as auditory triggers can cause Torsades
- Full non-invasive monitoring should be attached before induction
- For best drugs, see drug list in table below
- **The drugs in the "Avoid" list will prolong QT in all patients** and might cause Torsades in patients with borderline QTc.
- Avoid hypothermia, hypoxia and hypercarbia
- Resuscitation equipment should be readily available in theatre.

Preferred Agents	Exercise Caution	Avoid
Sedation Midazolam 0.05-3mgkg^{-1} i.v. Analgesia Lidocaine 1.5 mg kg^{-1} i.v. Fentanyl 2 µg kg^{-1} i.v. Alfentanil 0.5-3 µg kg^{-1} min^{-1} Remifentanil 0.1-0.5 µg kg^{-1} min^{-1} i.v Morphine 0.05-0.1 mg kg^{-1} i.v. I.V. anaesthetic agents Volatile anaesthetics Isoflurane 1-3% MAC inspired Neuromuscular blockers and reversal agents Rocuronium 0.6-1.2 mg kg^{-1} i.v. Vecuronium 0.04-0.1 mg kg^{-1} i.v. for intubation 0.8-1.2 µg kg^{-1} min^{-1} for maintenance Cisatracurium 0.15-0.2 mg kg^{-1} min^{-1} for intubation 0.06-0.18 mg kg^{-1} min^{-1} for maintenance Spinal and epidural anaesthesia Epidural preferred over spinal Combined spinal-epidural anaesthesia Bupivacaine Spinal: 0.8-1.6 ml of 0.75% Epidural: 3-5 ml boluses of 0.25-0.5% Ropivacaine Epidural: 5-25 ml of bupivacaine 0.5% Postoperative care and anti-emetics	 Buprenorphine 0.3 mg i.v. Methadone 0.1e0.3 mg kg^{-1} i.v. (do not exceed 200 mg day^{-1}) Propofol 6 mg kg^{-1} h^{-1} i.v. Etomidate 0.3 mg kg^{-1} i.v. Thiopental 2-6mgkg^{-1} i.v. Sevoflurane 0.5-3% MAC inspired Nitrous oxide 25-70% inspired Anticholinesterase-anticholinergic reversal agents Combined spinal-epidural anaesthesia Oxytocin Droperidol 0.625-1.25 mg i.v. Ondansetron 4 mg i.v. (do not exceed 16 mg) Metoclopramide 10-20 mg i.v. Dexamethasone 0.1 mg kg^{-1} i.v.	 Dexmedetomidine Epinephrine Ketamine Sufentanil Succinylcholine Pancuronium Glycopyrrolate Atropine Spinal anaesthesia to level of T10 Epinephrine Clarithromycin and erythromycin Grepafloxacin, levofloxacin, and moxifloxacin

Treatment of Torsade de Pointes

- 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 (for both treatment and prevention, even with a normal Magnesium level)
- In bradycardia or in magnesium-resistant TdP, transvenous pacing at 90bpm or higher can be effective
- Deepen anaesthesia to decrease sympathetic activity.

Post-operative Management

- Post-op monitoring dependent on case
- Analgesia and a calm environment essential
- No evidence for best practice length of monitoring or stay

References

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