Case Report

Recurrent syncope in a child and video assisted thoracoscopic surgery - The long and short of it

Address for correspondence:
Dr. Rajeshwari Subramaniam, Department of Anaesthesiology, Pain Medicine and Critical Care, All India Institute of Medical Sciences, New Delhi, India.
E-mail: drsrajeshwari@gmail.com

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Manpreet Kaur, Nirupa Ramakumar, Sangam Yadav, Rajeshwari Subramaniam
Department of Anaesthesiology, Pain Medicine and Critical Care, All India Institute of Medical Sciences, New Delhi, India

ABSTRACT

Long QT syndrome (LQTS) is a myocardial repolarisation disorder caused by cardiac ion channelopathy and one of its common presentations is recurrent syncope. This reduced repolarisation reserve in LQTS can be unmasked by perioperative factors like electrolyte imbalance, drugs, hypothermia and changes in cardiac autonomic tone. We report the anaesthetic management of left thoracoscopic sympathectomy in a 5-year-old child with LQTS and epicardial pacemaker *in situ*. It is very challenging to isolate the lung on one hand and prevent the predisposition to torsadogenic potential on the other.

Key words: Long QT, thoracoscopic left sympathectomy, uninvent

INTRODUCTION

Long QT syndrome (LQTS) is a cardiac ion channelopathy with incidence of congenital LQTS: 1 in 5000. There is scarcity of adequate literature on its perioperative management.[1,2] About 60% of the patients with congenital (c)-LQTS will have QT prolongation on the electrocardiogram [QTc >470 ms in males and 480 ms in females].[1]

Patients with latent LQTS have a reduced repolarisation reserve that can be unmasked by perioperative factors like electrolyte imbalance, drugs, hypothermia and changes in cardiac autonomic tone. We report the peri-operative management of left thoracoscopic sympathectomy in a 5-year-old child with LQTS and epicardial pacemaker *in situ*.

CASE HISTORY

This 5-year, 20 kg, 130 cm-male child (written informed consent for publication obtained from his parents) was diagnosed with LQTS1 (Romano Ward Syndrome) at the age of 4 years due to persistent syncopal episodes. He was born of non-consanguinous marriage, and two of his older male siblings had experienced sudden cardiac death. His younger sibling also had similar syncopal attacks and was on β-blockers. There were no associated hearing abnormalities in the child. He was on oral propranolol for last one year and was recently started on phenytoin. Despite the placement of epicardiac pacemaker (VVIR mode) 5 months back, syncopal episodes persisted and a decision for thoracoscopic left sympathectomy was made. His

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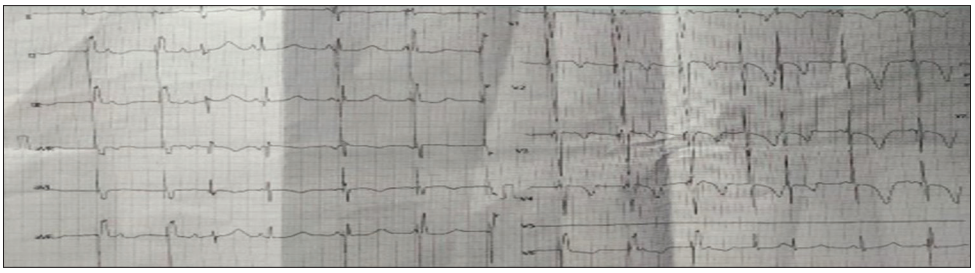

Figure 1: 12-Lead electrocardiogram showing that most of the beats are paced

preoperative heart rate (HR) was 70 beats/min (bpm), and non-invasive blood pressure (NIBP) was 98/54 mm Hg. The electrocardiogram (ECG) showed that most of the beats were paced and the QT interval was prolonged (526 msec) [Figure 1]. Investigations including electrolytes (K^+, $Ca2^+$, $Mg2^+$) were normal. Pacemaker interrogation revealed normal function with adequate battery reserve. Close communication with the paediatric cardiologist and pacemaker technician was maintained throughout the peri-operative period.

The child was transferred to the operating room (OR) after premedication with 1 mg midazolam intravenous (IV) (cannula inserted preoperatively). Monitoring included a continuous ECG, NIBP, invasive blood pressure (IBP) using a right radial arterial cannula and SpO_2 prior to induction and end-tidal carbon dioxide and nasopharyngeal temperature monitoring after intubation. Paediatric automated external defibrillator (AED) pads were applied at right infraclavicular and interscapular region [Figure 2]. Intravenous (IV) magnesium sulphate in the dose of 100 mg (5 mg/kg) was administered over 15 min and an IV lignocaine infusion was started at a rate of 1 $mg.kg^{-1}$ hr^{-1}. Anaesthesia was induced with IV 20 μg of fentanyl, 80 mg of propofol and 10 mg of atracurium. IBP remained stable and we did not encounter any arrhythmia during induction. The trachea was intubated with Univent tube (No. 3.5) (Fuji system corporation, Tokyo, Japan) with left bronchial blocker under fibre-optic guidance (2.8 mm). Position of the blocker was confirmed in supine and right lateral position. The lungs were ventilated with pressure-controlled ventilation (inspiratory pressure18 cm H2O, frequency 14/min and inspiratory time 1.5 sec) and anaesthesia maintained with O_2-air and sevoflurane to maintain a minimum alveolar concentration of 0.8–1.0. An epidural catheter was inserted at L3/L4 space and the catheter tip threaded up to T6 vertebral level (15 cm). Intraoperative analgesia was provided by IV 1 $\mu g.kg^{-1}$ fentanyl boluses, 300 mg paracetamol and epidural analgesia with 5 ml of 0.125% ropivacaine with 1 mg morphine bolus. The

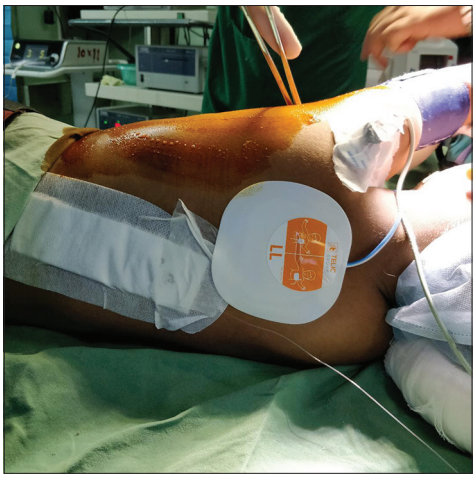

Figure 2: Paediatric automated external defibrillator (AED) pads applied interscapular region

pacemaker mode was left unchanged, and surgeons were advised to use the ultrasonic harmonic scalpel instead of the regular electro-cautery. Dexamethasone 2 mg was administered IV as antiemetic. The surgery was uneventful and lasted for 120 min, with blood loss of approximately 100 ml and child was extubated in the OR. Postoperative analgesia was provided with epidural 1 mg of morphine in 5 ml 0.1% ropivacaine bolus at 12 hourly intervals for 2 days. Rest of the postoperative course was uneventful.

DISCUSSION

Congenital LQTS is characterised by prolonged ventricular repolarisation due to cardiac ion channels (especially sodium, potassium and calcium) dysfunction. It predisposes to dysrhythmias especially torsades de pointes (TdP).[3] Anaesthetic management of patients with LQTS is challenging due to innate predisposition to arrhythmias with anaesthetic agents.

There are 12 different variants of LQTS described.[4] Two common variants are Romano Ward syndrome (Autosomal Dominant) and Jervell and Lange- Nielsen syndrome (Autosomal recessive).[1] Romano Ward

syndrome was diagnosed in our patient by autosomal dominant pattern suggested by sudden death in other siblings and absence of ear anomalies.

Anaesthetic and peri-operative management of these patients is aimed at prevention of further prolongation of QT interval, prevention of pacemaker failure, achieve one lung ventilation and effective postoperative analgesia.

Liaison with the child's cardiologist and pacemaker programmer is important for the assessment of pacemaker function and any decision to change the mode in the perioperative period. Antiarrhythmic drug therapy (β-blockers, phenytoin) should be continued on the day of surgery and well into the postoperative period to minimise the risk of perioperative arrhythmia.

Implantation of a cardioverter defibrillator is a treatment modality in children above 7 years and was not an option in this patient due to large size of the pulse generator. Thus, an epicardial VVIR pacemaker without internal cardioversion was inserted. We did not change the pacemaker mode to asynchronous mode since a decision to use the harmonic scalpel was taken. Further, the asynchronous mode is known to precipitate the R-on-T phenomenon.

Electrolyte abnormalities such as hypokalaemia, hypomagnesaemia and hypocalcaemia can predispose the heart to delayed repolarisation. Serum electrolytes should be within normal limits before surgery. Other physiologic and biochemical stressors of myocardial repolarisation reserve (pain, fear, dehydration, hypothermia) should be avoided.[5]

Adequate premedication and a calm atmosphere is essential. Continuous arterial pressure monitoring is recommended to detect any haemodynamic instability related to arrhythmias. The preoperative evaluation should also include pacemaker interrogation including details of the manufacturer and model, the programmed settings (e.g., mode and rate), the indication for the implant and need for changing the mode and avoid disruption of function.

One-lung ventilation provides good operating conditions for thoracoscopic sympathectomy.[6] Adult double-lumen tubes (DLTs) were too large for this patient; hence, the 'Univent' tube that has a movable bronchial blocker was used. Selective blocking of the left main bronchus was our goal. Use of Univent tube for paediatric thoracoscopic sympathectomy has not been described in literature.[7,8]

IV propofol was used as an induction agent as it does not prolong QT and has shown to reverse sevoflurane-induced QT prolongation in some studies.[9] Hence, maintenance with Propofol infusion is a reasonably good option; however, we were concerned about the hypotension caused by propofol infusion. All volatile anaesthetics agents prolong the QT interval.[10] Incidence of adverse events with inhalation agents in a recent cohort study of children with LQTS was 29.13% with desflurane, 48.54% with isoflurane and 22.33% with sevoflurane for maintenance of anaesthesia.[4] Due to minimum propensity of sevoflurane for adverse events, we chose to maintain anaesthesia with sevoflurane. In fact though sevoflurane prolongs QTc in healthy children, is not torsadogenic as it does not increase the Tp-e (The interval between the peak and end of the T wave) which is a predictor of transmural dispersion of repolarisation (TDR).[11]

Drugs that prolong QT interval should be avoided. Intraoperative use of lignocaine infusion can help with steady intraoperative course as we did.[6] Kenyon *et al*. successfully used lignocaine infusion in 3 patients to reduce arrhythmogenic potential.[7] We used magnesium bolus in this patient as it is a membrane stabilizing agent and it has been used in management of torsades de pointes,[12,13] but did not use magnesium infusion to avoid augmentation of neuromuscular blockade. Minimising neostigmine dose was also our goal (vide infra). Besides, magnesium infusion (2–4 mg/kg) was reserved for the treatment of torsades de pointes if it did occur.

Periods of enhanced sympathetic activity like intubation and emergence from anaesthesia should be smooth. The use of anticholinesterase and anticholinergic combinations, can precipitate fatal arrhythmias and should be avoided.[14] Antiemetic's like antidopaminergic drug droperidol and the 5-hydroxytryptamine (HT) 3-antagonist ondansetron prolong the QTc and should be avoided.[1] Hence, we used dexamethasone as antiemetic.

Good intraoperative and postoperative analgesia decreases the perioperative stress and provides pain-free awakening. It was apparent in this patient that the catheter tip was located in the correct place evidenced by lack of pain. We threaded the epidural

catheter till we had no resistance which corresponded to a distance of 15 cm (roughly to T6). Thoracic catheters under general anaesthesia is still controversial due to risk of cord injury. We used thoracic epidural so that in the event of conversion to open thoracotomy it would be used even more judiciously. We could not check for the sensory level after administration of local anaesthetics as the child was already under general anaesthesia. Pain following thoracic sympathectomy can be controlled with combination of opioids, acetaminophen, ketorolac, regional blocks such as intercostal nerve block, paravertebral block, lumbar epidural block with catheter extending to thoracic region.[6-8] Thoracic epidural in fact decreases the QTc and TDR.[8]

CONCLUSION

Preoperative optimisation, maintenance of β-blockade, detailed discussion with cardiologists and pacemaker technician to understand pacemaker function, avoidance of offending drugs and vigilant monitoring in the perioperative period can prevent occurrence of adverse cardiac events in patients with LQTS.

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Conflicts of interest

There are no conflicts of interest.

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