Case Report

Anaesthesia management of a case of Jervell and Lange-Nielsen syndrome for minimally invasive bilateral thoracoscopic cervicothoracic sympathectomy

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ABSTRACT

Long QT syndrome (LQTS) is an arrhythmogenic cardiac disorder resulting from the malfunction of cardiac ion channels. Patient with LQTS may present with syncope, seizures or sudden cardiac death secondary to polymorphic ventricular tachycardia (VT) or torsades de pointes. Patient may be asymptomatic in the pre-operative period but may develop VT for the first time in operation theatre. We are reporting anaesthetic management of a child with LQTS planned for bilateral thoracoscopic cervicothoracic sympathectomy.

Key words: Cervicothoracic sympathectomy, long QT syndrome, ventricular tachycardia

INTRODUCTION

Long QT syndrome (LQTS) is an arrhythmogenic cardiac disorder resulting from the malfunction of cardiac ion channels.[1,2] Congenital LQTS results from mutations in the genes encoding cardiac ion channels and acquired LQTS is caused by metabolic abnormalities or drugs.[1,2] Patient with LQTS may present with syncope, seizures or sudden cardiac death secondary to polymorphic ventricular tachycardia (VT) or torsades de pointes (TdP).[1,2] Patient may be asymptomatic in the pre-operative period and have VT for the first time in operation theatre (OT).

CASE REPORT

A 3-year-old boy presented to the emergency ward in an unconscious state after an episode of convulsions at home. Airway was secured with endotracheal intubation, and intravenous access was established. Monitors showed that the child had unstable VT for which immediate electrical cardioversion was done. He continued to have repeated episodes of unstable VT requiring defibrillation energy of shock. Intravenous lignocaine and β-blocker infusions were started. The patient was shifted to Intensive Care Unit (ICU). His parents gave the history that the child was deaf, and there was a history of sudden death of the elder sibling. Electrocardiogram (ECG) showed prolonged QTc interval (533 mSec) [Figure1]. Diagnosis of Jervell and Lange-Nielsen Syndrome was made. Since the child was unresponsive to medical management, a plan for cervicothoracic sympathectomy was made.

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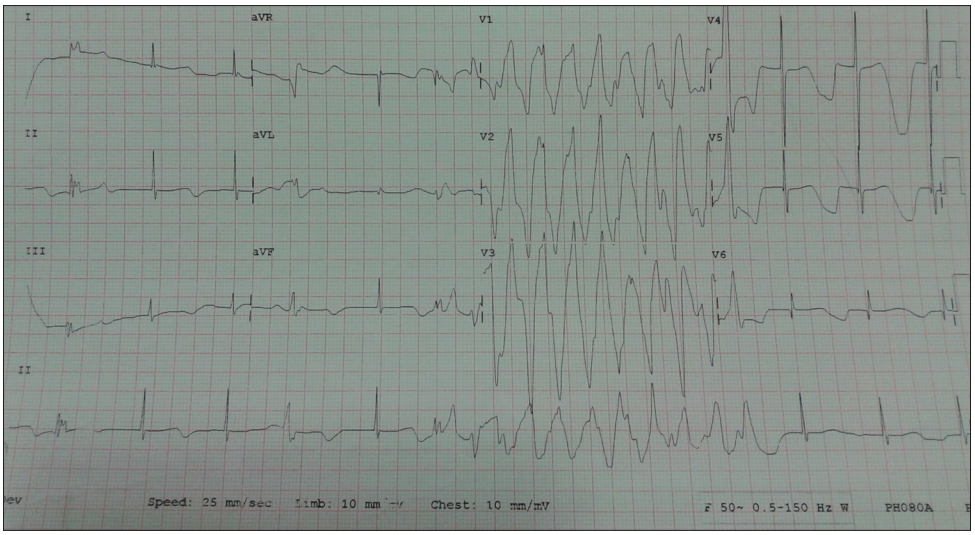

Figure 1: Pre-operative electrocardiogram of the child

During pre-anaesthesia workup, the child was still on mechanical ventilation with 100% oxygen. Chest auscultation revealed bilateral coarse crepitations (possible aspiration at the time of convulsions). The child continued to have repeated episodes of polymorphic VT in the ICU.

A left-sided stellate ganglion block was performed under ultrasound guidance with bupivacaine 0.25% (4 ml) in the ICU, and ipsilateral Horner's syndrome was noted. Following this, the ECG of the child reverted to sinus rhythm with occasional premature ventricular contractions for next 7–8 h. Forty-eight hours later, it was noted that the chest condition had improved and serum electrolytes were within normal limits. At this stage, it was decided to perform bilateral thoracoscopic cervicothoracic sympathectomy.

The child was shifted to the theatre on a mechanical ventilator with arterial and central lines *in situ* and defibrillator pads applied to the chest. The child was on metoprolol, lignocaine, fentanyl, calcium and potassium infusions. The child was also on meropenem, vancomycin, amikacin, mexiletine, spironolactone and frusemide medications. In the theatre, prophylactic temporary pacemaker insertion was performed. The defibrillator pads were repositioned anteroposteriorly in the midline to facilitate surgery in the supine position. Anaesthesia was maintained with propofol and fentanyl infusion with boluses of vecuronium. Norepinephrine infusion was started to maintain the blood pressure. It was decided to perform the surgery without capnothorax as cardioversion would not be possible with capnothorax if need arose. Considering the poor chest condition of the child, it was decided to keep the patient on two lung ventilation with low tidal volume and fast rate while allowing permissive hypercapnoea. Apnoea was provided at crucial steps to facilitate surgery with continued insufflation of 100% oxygen. The average value of end-tidal CO_2 was 52–55 mm Hg with the peak value of 57 mm Hg. Intraoperatively, there was no major cardiac event except for T-wave inversion which reverted to normal spontaneously at the end of the surgery. The child was shifted to the ICU on mechanical ventilation and anti-arrhythmic drugs. Trachea was extubated on the next day as the child did not have any further episodes of VT following surgery. The child was discharged from the hospital on the eighth post-operative day, on metoprolol and mexiletine tablets.

DISCUSSION

LQTS is an arrhythmogenic condition of the heart resulting from mutations in cardiac ion channels.[1] Congenital LQTS can be inherited as an autosomal recessive Jervell and Lange-Nielsen syndrome (JLNS) or dominant (Romano–Ward syndrome) condition. The former is associated with sensorineural deafness, whereas the latter is not.[1] Such children may present with convulsions, syncope or sudden cardiac death because of polymorphic VT or TdP.[2] JLNS patient may present in the OT for cochlear implantation or some other surgery and manifest with polymorphic VT intraoperatively. Therefore, getting an ECG for children posted for cochlear implantation may be a good idea. These patients may present for implantable cardioverter defibrillator placement or left cervical ganglionectomy.

Table 1: Effect of anaesthesia drugs on QTc interval and their implications[1,2,4]

Drugs	Effect on QTc interval	Effect
Midazolam	No effect	Can be used for anxiolysis
Opioids	Conflicting results	Has been used without any adverse effect
Ketamine		To be avoided as causes sympathetic stimulation
Propofol	Little or no effect	Reverse sevoflurane-induced QTc prolongation
Inhalational agents	Prolongation with little clinical effect	Isoflurane as agent of choice, halothane to be avoided as cause sensitisation of myocardium
Succinylcholine	Prolongation	Pre-treatment with non-depolarising muscle relaxant prevents prolongation
Vecuronium and atracurium	No effect	Atracurium causes histamine release and hence better avoided
Anticholinesterase + anticholinergic	Prolongation	Reversal avoided
Haloperidol	Prolongation	Avoid
Amiodarone	Prolongation	Avoid

Hypokalaemia, hypomagnesaemia and hypocalcaemia predispose the myocardium to delayed repolarisation.[1-3] Therefore, serum electrolytes should be within normal limits before surgery. Medications such as β-blockers have to be continued in the perioperative period. Noise level in the OT prior to induction of anaesthesia should be kept very low as any sympathetic stimulation may lead to VT. Similarly, all the factors which may lead to sympathetic stimulation such as anxiety, hypothermia, hypoxia, hypercarbia, laryngoscopy and pain should be taken into consideration and anaesthesia planned accordingly.[1-3] Drugs which are known to cause prolongation of QT interval [Table 1] should be avoided.

Cervicothoracic sympathectomy may not be able to correct the cardiac condition in all the patients with JLNS.[4] However, pre-operative positive response to stellate ganglion block may provide useful information on the therapeutic efficacy of the surgery. Stellate ganglion block has been used in the management of electrical storm due to ischaemic heart disease in adult population,[5] but there is no mention of its use in the paediatric age group with LQTS.

CONCLUSION

Maintenance of electrolyte balance, avoidance of sympathetic stimulation and drugs that cause prolongation of QT interval can lead to uneventful course in patients with Jervell and Lange-Nielsen syndrome undergoing cervicothoracic sympathectomy. Stellate ganglion block can act as a bridging therapy in patients who present in critical condition, until optimisation is achieved.

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Conflicts of interest
There are no conflicts of interest.

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