A child with Jervell and Lange-Nielsen syndrome for permanent pacemaker implantation and sympathectomy: Anesthesia management and considerations

Sir,

Jervell and Lange-Nielsen (JLN) syndrome is an autosomal recessive condition presenting with bilateral deafness, long QT syndrome (LQTS), and sudden death.

A 6½-year-old male child weighing 15 kg with history of recurrent episodes of syncopal attacks and having bilateral deafness was diagnosed with Jervell and Lange-Nielsen (JLN) syndrome. His 12 lead electrocardiography (ECG) showed a QTc interval of 441 ms [Figure 1]. The child had a positive family history of sudden cardiac death (in one of his siblings). The syncopes persisted inspite of beta blockers (20 mg of propranolol tablet twice a day for more than 6 months). The electrophysiologist planned for a permanent pacemaker implantation (PPI) first, followed by a left cervicodorsal sympathectomy 3 days later. Routine investigations including serum electrolytes were normal. Baseline heart rate was 54/min. The patient was classified as American Society of Anesthesiologists Grade III physical status and an informed consent was obtained. Tablet propranolol was continued as per schedule.

We monitored ECG, invasive arterial pressure, oxygen saturation, and end tidal CO₂ intraoperatively. Transdermal paddles were attached and defibrillator was kept ready. On both occasions, induction of anesthesia was done with propofol and a single dose of atracurium as muscle relaxant and trachea was intubated with 4.5 sized cuffed endotracheal tube. We used intravenous (IV) fentanyl as analgesic agent (30 μg) and propofol infusion as maintenance agent (100 μg/kg/min). Cardiologist accomplished single chamber PPI (AAI) through left subclavian vein under fluoroscopy guidance and a rate of 90/min was set. In the 12 lead ECG after PPI, QT interval was still prolonged, but as patient was on beta blockers and the heart rate was fixed at 90/min, the possibility of prolonged QT leading to fatal arrhythmias gets reduced [Figure 2]. During video assisted thoracoscopic surgery (VATS), lung isolation was achieved with 4 Fr Fogarty catheter placed in left bronchus under fiberoptic guidance. Before VATS, pacemaker was converted to asynchronous mode (AOO). No reversal agent was used and trachea was extubated uneventfully. Vital parameters were stable throughout procedure. The child was discharged from the hospital after 5 days.

Long QT syndrome (LQTS) is an arrhythmogenic cardiovascular condition characterized by prolonged ventricular repolarization. The prolonged ventricular repolarization is due to malfunction of cardiac ion cells. It manifests as recurrent syncpe, pseudo - seizures, cardiac arrest, but often it is an incidental finding either on ECG or noticed on precipitation following drug administration.[1]

QT interval varies with heart rate. With bradycardia, it increases and gets shortened with tachycardia. Hence, QT interval is corrected for heart rate, using Bazette’s formula.[2]

\[ QTc = \frac{\text{Measured QT}}{\sqrt{R-R \text{ interval}}} \] (all in seconds)

A QTc interval of >440 ms is prolonged QTc. However, in 6% patients of symptomatic LQTS, QTc is normal. QT interval is the total duration of depolarization and repolarization phase of ventricular action potential. Therefore, increase in QT interval due to prolongation of QRS constitute LQTS. Hence, QT interval is used as an indicator of ventricular repolarization as it avoids incorporation of QRS duration.

There are seven ion channels known to cause LQTS. But for the sake of discussion, they are classified as congenital
and acquired LQTS.\textsuperscript{[3]} Congenital LQTS includes two syndromes. JLN syndrome also known an cardioauditory or surdocardiac syndrome is an autosomal recessive condition, which constitutes prolonged QT interval, congenital deafness, and sudden death.\textsuperscript{[4]} Romano-Ward syndrome is an autosomal dominant condition, which presents as prolonged QT interval only and is 3 times more common than JLN syndrome.\textsuperscript{[5]} Acquired LQTS usually manifests because of drugs, electrolyte abnormalities, starvation, neurological injury, etc.

Electrophysiologically, there are three distinct types of cell in the ventricular myocardium: Epicardial, endocardial, and M cells. M cells are different from the endocardial and epicardial cells. The action potential of M cells tends to prolong disproportionately leading to the development of a transmural dispersion of repolarization (TDP) which is estimated on ECG as the interval between the peak and the end of T wave (QT peak-QT end interval). This is referred as Tp-e. The prolongation of QT interval will evolve into torsades only if TDP is prolonged. On several occasions, torsades de points (TdP) is of a short duration and terminates by itself, but it can turn out to be fatal if it progresses to ventricular fibrillation. In that case, it can be terminated only by DC shock.\textsuperscript{[6]}

Optimal doses of narcotics, beta blockers, deep anesthesia plane should be used during induction and intubation to prevent sympathetic hyperactivity. Normocarbia, euthermia, euvolemia should be the aim as any derangement can trigger arrhythmia. High airway pressures should be avoided as it mimics valsalva maneuver and can prolong QTc. Beta blockers, magnesium sulfate, equipments like hardware for transcutaneous pacing, transvenous pacing, and defibrillator should be available throughout surgery.

DC cardioversion is the only treatment for torsades. Once detected, a loading of 30 mg/kg of magnesium sulfate over 2-3 min should be administered intravenously followed by an infusion at 2-4 mg/min. IV magnesium acts by membrane stabilization and as a calcium blocker. It activates sodium-potassium ATPase in cell membrane and leads to resting repolarization (Phase 4) and reduction of arrhythmia.

However, it doesn’t shorten QT interval. Magnesium infusion can lead to potentiation of neuromuscular blockade due to use of muscle relaxant.

Propofol neither prolongs QTc or Tp-e hence is clearly nontorsadogenic and is therefore the safest anesthetic agent in prolonged QTc. Benzodiazepines and narcotics can be safely used as it doesn’t interfere with QTc. Thiopentone sodium prolongs QTc, but as it decreases TDP it can be used. Ketamine has no direct effects on QTc or TDP, but owing to its sympathomimetic effects it should be avoided. Not much information is available regarding effect of dexmedetomidine on QTc prolongation leading to TdP. It may theoretically help as it reduces heart rate, like beta blockers. Succinylcholine prolongs QT interval in recommended doses. Nondepolarizing muscle relaxants can be safely used. Anticholinergics (atropine, glycopyrrrolate) can prolong QTc and precipitate torsades. Neostigmine can also prolong QTc. Volatile anesthetics can prolong QTc, especially in children with congenital QT prolongation. Studies with sevoflurane in children’s with LQTS has shown that although QT interval is prolonged, there is no effect on Tp-e interval. Regional techniques can be safely used provided the patient is euvolemic. In case of hypotension, avoid sympathomimetics like ephedrine and mephentermine. Phenylephrine is the drug of choice.\textsuperscript{[7]}

Beta blockers are the mainstay in the treatment of LQTS. By reducing the heart rate, shortening QT and sympatholysis, they prevent the development of torsades in a patient with LQTS.\textsuperscript{[8]} automated implantable cardioverter-defibrillator (AICD) is the treatment of choice for patients with LQTS, especially those who are refractory to beta blockers by preventing sudden death in high risk patients.\textsuperscript{[9]} When proper sized AICD is not available (as in pediatric patients), a PPI can be placed and after setting a high heart rate, beta blockers are started. It’s called as antiarrhythmic pacing.

Pacing also prevents bradycardia and pauses, which can lead to arrhythmias. AICD can be planned once the child grows up. Refractory patients who do not benefit from beta blockers or PPI can be offered a left cervical sympathectomy/left stellate ganglionectomy. After sympathectomy, it has been observed that there is reduction in QT dispersion and decreased incidence of torsades.\textsuperscript{[10]}

Patients with LQTS are a challenge to anesthesiologist and undiagnosed patients can be a nightmare. Avoid drugs that prolong QT/TDP/Tp-e, avoid sympathetic overactivity, and maintain normothermia, normocarbia, euvolemic and electrolyte levels.

Crash cart should be equipped with essential drugs and equipments. Beta blockers should be continued perioperatively.
phenomenon includes three different elements, namely, the
its early description in the 16th century.[1] The phantom limb
for the patient, so has been the fascination of physicians since
As disturbing and distressing “phantom limb phenomenon” is

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