Safe anesthesia management protocol of a child with congenital long QT syndrome and deafness (Jervell and lange-nielsen syndrome) for cochlear implant surgery

Sir,

Jervell and Lange-Nielsen syndrome (JLNS) is an autosomal recessive variant of congenital long QT syndrome (c-LQTS) associated with deafness.[1] These patients are predisposed to polymorphic ventricular tachycardia (torsades de pointes [TdP]) leading to syncope and sudden death. Surgical stress in the perioperative period and general anesthesia *per se* may trigger fatal arrhythmia.[1,2]

A 2-year-old, weighing 10 kg, male child with congenital deafness was scheduled for cochlear implant surgery. There was no history of seizure, cyanosis or syncope, but one sibling had sudden death at 3 months of age. Electrocardiography (ECG) revealed prolonged QT interval (QTc 519 ms). He was diagnosed with JLNS and tablet propanolol 10 mg twice daily was started. Hematology, blood biochemistry, chest X-ray, and echocardiography were normal.

On the day of surgery, he was premedicated with oral midazolam 5 mg and eutectic mixture of local anesthetics (EMLA) was applied on the left dorsum. After 45 min an intravenous 22G canula was inserted with the child in mother's lap. He was shifted inside the operating room (OR) accompanied by mother. Standard monitors and debrillator-cum-transcutaneous pacer pads were placed. Anesthesia was induced with fentanyl 30 mcg, propofol 25 mg and vecuronium 1 mg and trachea was intubated with 5.0 mm uncuffed endotracheal tube. Anesthesia was maintained with O_2:N_2O 50:50 with isoflurane (minimum alveolar concentration 1-1.2). Dexamethasone 2 mg was given intravenously. Analgesia was provided with infusion fentanyl at 15 mcg/h and intravenous paracetamol 150 mg. Magnesium sulfate was kept ready. Surgery lasted for 3 h and intraoperative vitals were stable. At the end, neuromuscular block was reversed with neostigmine and glycopyrrolate and trachea was extubated when the child was awake. Patient was kept in a quiet area in postanesthesia care unit with continuous ECG monitoring with defibrillator pads placed *in situ*. Postoperative analgesia was provided with intermittent morphine 0.5 mg boluses and intravenous paracetamol 150 mg every 6 h. Subsequent postoperative course was uneventful and child was discharged after 10 days.

Children with congenital deafness coming for cochlear implant surgery may be associated with various syndromes and c-LQTS may be one of them.[3] Any history of sudden cardiac death in a sibling like in the present case; a history of syncope or a diagnosis of epilepsy should arouse suspicion

of c-LQTS.[2] In a suspected case "Schwartz score" should be calculated and documented. Any score of 3.5 signifies a high probability of LQTS.[4] In the present child, Schwartz score was 4.0 (QTc >480 = 3; congenital deafness = 0.5 and unexplained sudden cardiac death in young family member = 0.5).

Although beta blocker has been found to reduce mortality, nearly one third patients may experience cardiac arrest or sudden death even on beta blocker therapy. In patients with recurrent symptoms even on beta blocker, implantable cardioverter-defibrillator or pacemaker or left cardiac sympathetic denervation may be considered.[2,5]

Beta blocker should be continued in the perioperative period. Magnesium sulfate and defibrillator should be readily available, the latter may be required in case TdP is not responding to Mg or causing hemodynamic compromise. Some short TdP episodes, which occur in association with bradycardia may require overdrive pacing.[2,5] In our case, beta blocker was continued, magnesium was kept ready, pads of defibrillator with transcutaneous pacemaker was attached.

Any sympathetic stimulation, including anxiety, cry, loud noise can precipitate TdP.[2] Hence, the child was premedicated with oral midazolam, intravenous canula was obtained in mother's lap after applying EMLA cream cram and he was shifted to OR with mother. In the postoperative period also he was kept in quiet environment. Inadequate plane of anesthesia, brady or tachycardia, hypertension, hypoxia, hypo or hypercarbia, inadequate analgesia, hypothermia may cause sympathetic stimulation increasing the risk of TdP.[2] All these factors were avoided and adequate analgesia was provided. Serum electrolytes should be normal, as hypokalemia, hypomagnesaemia and hypocalcaemia predispose to delayed ventricular repolarization and TdP.[5]

Drugs which prolong the QT interval should be avoided. Midazolam does not affect QT interval.[2] Thiopental prolongs,[2] whereas propofol has been found to have little or no effect on the QT interval and may even reverse sevoflurane induced QT prolongation.[2,5] All volatile agents have the potential to prolong QT interval. However, Isoflurane is inherently safe and has been reported to shorten QT interval in a patient with c-LQTS and therefore is regarded as the agent of choice.[2] Vecuronium and atracurium have been found to have no effect on the QT interval.[2] Fentanyl and morphine have been used without adverse effects in patients with c-LQTS.[2] Ondansetron has possible risk of TdP and *in vitro* studies have shown that 5HT3 receptor antagonist may potentially prolong QT interval.[2] Therefore ondansetron was not used and only dexamethasone was used as antiemetic. The use of anticholinergic and anticholinesterase as reversal agent has been shown to prolong QT interval in healthy subjects and hence should be used with caution.[2,5]

To conclude, safe anesthesia management in children with c-LQTS with deafness (JLNS) would include:

a. Peri-operative continuation of beta blockade,
b. Avoidance of sympathetic stimulation, which may trigger ventricular arrhythmia (TdP),
c. Providing adequate premedication, peri-operative analgesia and careful use of anesthetic and other drugs to avoid QT prolongation,
d. Ready availability of magnesium sulfate and defibrillator-pacer in case any arrhythmia occurs.

Mayank Kumar, Dalim Kumar Baidya, Virender Kumar Mohan, Mamta

Department of Anesthesia and Intensive Care, All India Institute of Medical Sciences, New Delhi, India

Address for correspondence:
Dr. Dalim Kumar Baidya,
Department of Anesthesia and Intensive Care, 5th Floor, Teaching Block, All India Institute of Medical Sciences, New Delhi - 110 029, India.
E-mail: dalimkumarb001@yahoo.co.in

REFERENCES

1. Schwartz PJ, Spazzolini C, Crotti L, Bathen J, Amlie JP, Timothy K, *et al*. The Jervell and Lange-Nielsen syndrome: Natural history, molecular basis, and clinical outcome. Circulation 2006;113:783-90.
2. Kies SJ, Pabelick CM, Hurley HA, White RD, Ackerman MJ. Anesthesia for patients with congenital long QT syndrome. Anesthesiology 2005;102:204-10.
3. Baidya DK, Dehran M. Anesthesia for cochlear implant. Trends Anaesth Crit Care 2011;1:90-4.
4. Schwartz PJ, Crotti L, Insolia R. Long-QT syndrome: From genetics to management. Circ Arrhythm Electrophysiol 2012;5:868-77.
5. Booker PD, Whyte SD, Ladusans EJ. Long QT syndrome and anesthesia. Br J Anaesth 2003;90:349-66.

Access this article online	
Quick Response Code:	Website: www.saudija.org
	DOI: 10.4103/1658-354X.146328