



Anesthetic implications for cochlear implant in a child with cardiac anomaly

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ABSTRACT

The symptomatic or incidental finding of one or more congenital anomalies warrants a detailed history and systemic examination to find out or exclude the presence of other associated anomalies. In the present case report, a young child presented with hearing loss and a cardiac anomaly. A pre-emptive interventional management resulted in successful outcome after the surgery under a tailored general anesthesia.

Key words: Cochlear Implant; Cardiac anomaly; General anesthesia

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INTRODUCTION

Cochlear implantation has paved new ways for treatment in patients with deaf-mutism and irreversible hearing loss. Till now more than 400 genetic conditions have been found to be associated with hearing loss. The universal newborn hearing screening has played an important role in detection, diagnosis and intervention of hearing loss at an early stage of life hence allowing improved speech perception and sound localization at an early age. We report a case of a young child with hearing impairment and heart block for cochlear implant surgery.

CASE REPORT

A 9 year old boy weighting 30 kg admitted with chief complaints of inability to hear and speak since childhood and planned for cochlear implant. Child

had a history of birth asphyxia and delayed milestones.

On examination child had severe to profound bilateral hearing loss. All hematological and biochemical tests were within normal limits. A routine electrocardiogram showed Wenkebachs phenomenon. Child was asymptomatic and had a good effort tolerance. A 24 hour Holter monitoring revealed type 1 AV block. 2D echocardiography revealed a structurally normal heart with episodes of bradycardia. Prophylactically, a temporary pacemaker (TP) was inserted before surgery via right femoral vein, with output of 4MV and a back up pacing of 70/min.

Child was premedicated with midazolam 2.5 mg PO, induced and maintained with propofol infusion. Muscle relaxation was achieved with atracurium,

while for analgesia fentanyl 2 µg/kg was given IV. A close monitoring of 12 lead ECG along with monitoring of NIBP, SpO₂, ETCO₂ and temperature was done. For postoperative analgesia intravenous paracetamol was used. Perioperative period remained uneventful. A day after surgery TP was removed and child was discharged home.

DISCUSSION

Cochlear implant is an electronic prosthesis which partially replaces the functions of cochlea in patients with severe sensorineural hearing loss. Of all the patients with hearing loss 50% cases are hereditary. Among these 30% have additional abnormalities e.g. craniofacial, skeletal, ocular, neurologic, renal and cardiovascular system defects and are a part of a syndrome.¹ Utmost significance among these are cardiac co-morbidities which influence in a spectrum of ways ranging from minor intra operative issues to major life threatening complications.

The Jervell Lange-Nielsen syndrome first described in 1957, is an assemblage of syncope, sudden death, congenital sensorineural deafness, prolonged QT interval and cardiac arrhythmias.² Factors which trigger symptoms include emotional stress, exercise, or loud noise. Pathophysiology involves mutations in KCNE1 and KCNQ1 genes which alter the structure and function of potassium channels disrupting the movement of potassium ions in the inner ear and cardiac muscle, hence leading to hearing loss and irregular cardiac rhythm. These patients are treated with beta blockers to reduce risk of arrhythmias and may require an implantable cardioverter defibrillator (ICD) insertion or pacing.^{3,4} Goldenhar syndrome is an anomaly of first and second branchial arches. The spectrum of cardiac malformations found in this condition include tetralogy of fallot, Double outlet right ventricle, ventricular septal defect, total anomalous pulmonary venous return, single ventricle and atresia of pulmonary artery. Congenital rubella syndrome encompasses cardiac, cerebral, ophthalmic and auditory defects. Various other syndromes with hearing impairment and a spectrum of other organ systems involvement include- Treacher Collins syndrome, branchio- oto- renal syndrome, CHARGE syndrome, Pierre Robin syndrome, stickler, Aperts, Crouzon and Velocardiofacial and mitochondrial disorders.

A thorough knowledge on the part of physician is required to be able to rule out and diagnose associated disorders and manage appropriately because most of pediatric patients appear asymptomatic. Pre anesthetic assessment should include complete

medical history, including risk of prenatal infections and family history. Complete general and systemic examination is required including an airway examination to rule out any syndromes. Routine blood investigations plus electrocardiogram should be mandatory in pediatric patients to rule out possibility of underlying cardiac pathology. And a complete cardiac work up in selected cases.

Literature search showed that Kies et al. offered recommendations for anesthetic management of patients with congenital long QT syndromes.⁵ Use of pre medication calms the child and lessens anxiety, as in our case we premedicated with midazolam. In our case TP was inserted to maintain adequate heart rate. Choice of induction agent and other drugs is guided by concern for their action on QT interval, myocardium and postoperative nausea and vomiting. Use of opioids provides analgesia and reduces stress response. Hypothermia has to be taken care of by active heating with forced warm air and use of warm fluids. Minimum blood pressure fluctuations are required to provide a still and bloodless field to the surgeon. Inhalational agents are best avoided as these sensitize myocardium to catecholamines. Total intravenous anesthesia remains a better choice. Adequate anti-emetic prophylaxis has to be given as ear surgeries carry a high rate of 40-50% of postoperative nausea vomiting. Anticholinergic and anticholinesterases lengthen QT interval and cause significant bradycardia and should be used with caution. Extubation should be smooth, with minimum coughing and bucking to avoid dislodgement of implant and sympathetic stimulation. Adequate analgesia should be given to ensure calm child in postoperative area with adequate monitoring.

CONCLUSION

It is a specialized surgery which requires expertise on behalf of surgeon and anesthetist to provide best possible results. Identifying associated problems like cardiac issues needs meticulous investigative protocols and elaborative cardiac workup to prevent untoward events. Judicious pre-operative planning will help us to surge over the risks during the procedure. Appropriate counselling to the parents and family needs to be emphasized in such scenarios. Cochlear implantation is safe in cardiac co-morbid conditions provided they are diagnosed early and treated appropriately.

Conflicts of interest: None

Authors' contribution: Both authors took part in managing the case, preparation of the manuscript and literature review.

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