Anaesthetic Management of Hunter Syndrome with AmbuLMA

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ABSTRACT

Patients with Hunter Syndrome have multisystem involvement and difficult airway due to infiltration of tissues with mucopolysaccharides. A eight year old, male child with Hunter Syndrome weighing 20 kg was admitted for repair of umbilical hernia and right inguinal hernia. Anaesthetic management was planned with IV induction and a 2.5 size Ambu Laryngeal Mask Airway (LMA). Post-operatively child was observed for 24 hrs and LMA was removed when the child was fully awake. Children with mucopolysaccharidosis are prone to atlantoaxial subluxation. Airway management with AmbuLMA in this case demonstrated safe alternative to endotracheal intubation.

Key words: Hunter Syndrome, AmbuLMA, Difficult Airway, caudal epidural

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INTRODUCTION

Hunter Syndrome (Mucopolysaccharidosis Type II) is a rare x-linked lysosomal storage disorder with an estimated incidence of 1.3 in 100,000 male live births.1 The absence or deficiency of enzyme iduronate 2 sulfatase leads to accumulation of heparan and dermatan sulphate and usually presents at 2-4 years of age with structural abnormalities, airway obstructions, coarse facies, abdominal wall hernia, and mental retardation. Other features include hepatosplenomegaly, pulmonary dysfunction, cardiomyopathy and valvular dysfunction. Patients present with difficult airway due to tissue infiltration of mucopolysaccharides and other systemic involvement that are important concerns for anaesthesiologist. We describe the anaesthetic management of a child with Hunter Syndrome posted for repair of umbilical hernia and right inguinal hernia.

CASE REPORT

An eight year old, 20 kg male child with hunter syndrome was scheduled for surgery of umbilical hernia and right inguinal hernia. The child also had history of delayed milestones, easy fatiguability, mouth breathing, snoring, and episodes of sleep apnea. The patient was a known case of cardiomyopathy and was on diuretics, digoxin and enalapril.

On examination, he had boat shaped skull, short neck, coarse facies, stiff joint, and thick epicanthic folds. Airway examination revealed macroglossia and Mallampatti grade III. Mouth opening, neck movements and thyromental distance were adequate. The cardiovascular and respiratory examinations were normal. The pre-operative hemogram, renal function tests, liver function tests and serum electrolytes showed results within normal range. Other investigations and their findings are mentioned in table 1.

Table 1 Investigations and findings in Hunter Syndrome patient

Sl No	Investigations	Findings
1	Echo-cardiography	Dilated cardiomyopathy with ejection fraction of 40% and moderate arotic regurgitation.
2	Ultrasound Abdomen	Hepatosplenomegaly
3	CT scan Brain	Fused sagittal and lambdoid sutures, cerebral atrophy, non-specific hypo density in right parietal subcortical white matter
4	Polysomnography	Obstructive sleep apnea with Apnac hypapnea index (API) of 2
5	X-ray Phalanges	Broadening of phalanges

Anaesthetic Management: An informed consent was taken for parents explaining about the surgical and anaesthetic procedures. Pre-operatively, different sizes of endotracheal tubes were made available anticipating narrow airway along with Laryngeal Mask Airway (LMA), laryngoscope blades, oral airway, bougie and paediatric fibre optic bronchoscope. Child was not premedicated in order to avoid sedation and airway obstruction. We planned an intra-venous (IV) induction with titrated doses of Propofol. After confirming adequate mass ventilation and jaw relaxation a 2.5 size Ambu LMA was inserted. Fentanyl 40 µg and atracurium 5 mg was administered later. Anaesthesia was maintained with 1.5% isoflurane in 50:50 oxygen air mixtures and minimum alveolar concentration (MAC) maintained at 1-1.2. Caudal epidural block was given with 15 ml of 0.2% bupivicane. The duration of surgery was 60 minutes with <50 ml blood loss. Fluid deficits and losses were replaced with balanced salt solution. Analgesia was supplemented with 300 mg of IV paracetamol. Intra-operative period was uneventful.

Post-operatively, after reversal of neuro muscular blockade, LMA was removed when the child was fully awake and monitored for 24 hrs. Postoperative analgesia was maintained with I.V pracetamol 300 mg 6th hourly. Child was comfortable and postoperative period was uneventful.

DISCUSSION

Patients with Hunter syndrome commonly present with dysmorphic facial features and short stature with multisystem involvement.2 Airway management in Hunter syndrome is a challenge and incidence of difficult intubation and failed intubation is reported to be 25% and 8% respectively.3 In addition to the features outlined, our patient was also a diagnosed case of Obstructive Sleep Apnea (OSA) which further increased the risk of airway obstruction. Evidences show that difficult airway could be managed by inhalational route of induction.4 However, our patient was non co-operative and therefore we used intravenous induction as an

alternative. Intravenous induction has been done successfully using a combination of ketamine, midazolam and glycopyrrolate.5

Ambu Laryngeal Mask Airway (LMA) was inserted to gain additional advantage so as to prevent atlantoaxial subluxation which can occur in patients with Mucopolysaccharidosis.6 LMA has been used in patients with Hunters syndrome both as a definitive airway and for rescue in difficult intubation.3,7 However, narrowing of the lower airway and tracheomalacia may cause airway obstruction and cannot be managed by LMA.8 Therefore smaller size endotracheal tubes along with fibreoptic bronchoscope were kept standby. In patients with difficult mask ventilation, fiber optic intubation and LMA has been used successfully.9 LMA has the advantage that the child accepts it till he is fully awake thus there are less chances of airway compromise.

The patient was a known case of cardiomyopathy and medications were continued in the perioperative period. Intraoperative fluid administration (200 ml) was as per calculated fasting deficits and maintenance fluids. Caudal epidural bupivacaine along with intravenous paracetamol provided adequate intraoperative and postoperative analgesia. Caudal analgesia decreases the need for inhalation agent and opioids thus resulting in a comfortable recovery and awakening of child in the postoperative period. Monitoring in the postoperative period may result in early identification of obstructed airway and is key factor in preventing postoperative airway obstruction. It is important to note that Hunter Syndrome is progressive disorder and prone for anaesthetic complications with increase in age.10

CONCLUSION

General anaesthesia poses a high risk for patients with Hunter Syndrome due to difficulties in airway management. A thorough pre-operative assessment, optimization of systemic illness; and minimal use of opioids is needed. Use of AmbuLMA along with caudal epidural block with local anaesthetics provides airway management, adequate analgesia and in this case demonstrated as safe alternative to endotracheal intubation. A meticulous monitoring in the postoperative period enables early recognition of respiratory obstruction.

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