

Successful anaesthesia management of a child with hunter syndrome for adenotonsillectomy

Jyotsna Punj^{1,*}, Paridhi Kaler¹, Basavaraj Ankalagi¹, Arya Prasad¹, Renu Sinha¹, Prem Sagar²

¹Department of Anesthesiology, All India Institute of Medical Sciences, New Delhi, India;

²Department of ENT, All India Institute of Medical Sciences, New Delhi, India.

Summary

Airway management in a child with hunter syndrome is a challenge to the anesthetists. Various methods to achieve this are reported in literature. Here we describe another method in a three year old male child posted for adenotonsillectomy and myringotomy. After check videolaryngoscopy with C Mac blade size 2, vocal cords were not visible even with various manoeuvres. Thus a larger blade size 3 was used to place it under the epiglottis after which posterior part of vocal cords became visible and bougie guided endotracheal intubation was successful. Thus we recommend that in a child with hunter syndrome if vocal cords are not visible, a larger blade can be utilized to place under the epiglottis to visualize the vocal cords for successful endotracheal intubation.

Keywords: Endotracheal intubation, hunter syndrome, airway, C Mac blade

Mucopolysaccharidoses (MPS) is a rare, inherited, lysosomal storage diseases characterized by accumulation of glycosaminoglycans in various tissues including airways which makes airway management a challenging (1,2). Amongst the seven types of reported MPS, highest incidence of difficult intubation is encountered in MPS II also called hunter syndrome (3,4). Here we describe successful anaesthesia management in a child with hunter syndrome and sleep apnea.

A three year-old, 14 kg male child, diagnosed case of hunter syndrome was scheduled for adenotonsillectomy with bilateral myringotomy and grommet insertion. He presented with breathlessness with lower respiratory chest infection. On admission, child had yellow thick nasal discharge and was breathing through mouth in sitting position. He had respiratory rate of 24/min with bilateral coarse crepitations on auscultation with 96% room air saturation and was receiving oral antibiotics and salbutamol nebulisation. He had history of recurrent

chest infections, mouth breathing, snoring, episodes of sleep apnea, frequent night awakenings and delayed milestones. On examination child had large head, coarse facies, macroglossia, receding mandible and short neck with limited neck extension. He could not sleep in supine position and needed pillow under the shoulders. Enzyme replacement therapy was not being taken by the patient due to financial constraints. Cardiovascular system examination revealed no abnormality and echocardiography was normal. His preoperative routine investigations including hemogram and urine analysis were within normal limits. Computed tomography of head and neck revealed nasopharyngeal airway lumen of 2 mm, grade IV adenoid hypertrophy, grade II tonsillar hypertrophy and no hydrocephalus. Surgery was rescheduled after resolution of chest infection. Risk of difficult airway was explained to parents with informed written consent for tracheostomy.

On the day of surgery, before shifting to the operating room, a 24 gauge cannula was secured on left dorsum of wrist, xylometazoline drops were instilled in both nostrils and nebulization with 3 mL of 2% xylocaine was done. Difficult airway cart was prepared and airway adjuncts were kept ready according to the age and weight of the patient.

Plan A was induction of anaesthesia with sevoflurane in 100% oxygen and a check laryngoscopy

Released online in J-STAGE as advance publication November 25, 2019.

*Address correspondence to:

Dr. Jyotsna Punj, Room number 5016, Department of Anesthesiology, All India Institute of Medical Sciences, Ansari Nagar, New Delhi 110029, India.

E-mail: jyotsna_punj@yahoo.com

with a CMAC videolaryngoscope under spontaneous breathing. Plan B was intubation with oral pediatric fiberoptic bronchoscope (FOB) under spontaneous ventilation. Plan C was insertion of air Q ILA and FOB guided intubation through it under spontaneous ventilation. If at the time of induction of anesthesia, difficult mask ventilation would be encountered, it was planned to insert air Q ILA as plan A. In case of failure, tracheostomy was planned as the last resort.

In the operating room, video games on phone were shown to the child and standard monitors (pulse oximeter, ECG, BP cuff) were attached. Intravenous glycopyrolate 0.1 mg and ketamine 15 mg were administered and child was placed supine with a roll under his shoulders without a pillow. Inhalational anaesthesia was induced with 100% oxygen at 4 L/min fresh gas flow with gradual increase in sevoflurane concentration by face mask. Size 2 oral airway was inserted for effective mask ventilation. Check laryngoscopy with C-MAC blade 2 revealed cormack lehane (CL) grade III b view with no improvement after optimal external laryngeal manipulation (OELM) (Figure 1A). 10% lignocaine was sprayed over epiglottis, arytenoids and below the epiglottis to anesthetize vocal cords (Figure 1B) and bag-mask assisted spontaneous ventilation was resumed. Repeat laryngoscopy was performed with CMAC size 3 blade which was now kept below posterior surface of epiglottis to reveal posterior part of larynx. Bougie guided endotracheal intubation with size 5mm south pole preformed cuffed endotracheal tube was successfully inserted into trachea at first attempt (Figure 1C). Bilateral air entry was confirmed and atracurium 7 mg was administered. ETT was fixed in midline and stomach was decompressed with a suction catheter. Lungs were ventilated with volume control mode 150 mL tidal volume with oxygen:air (FiO₂ 0.5) and 2-5% sevoflurane dial concentration to achieve 1.0-1.3 MAC. Intraoperatively peak airway pressure remained at 20 mmHg. Intravenous dexamethasone 4mg and paracetamol 200 mg were administered for analgesia. At the end of surgery, no active bleeding from tonsillar fossa was ensured, gauze soaked with 0.25 % ropivacaine was kept in peritonsillar bed bilaterally for a minute. Surgery lasted for one hour and was uneventful. Sevoflurane was switched off, and at start of spontaneous ventilation; muscle relaxation was reversed with injection neostigmine and glycopyrrolate. After achieving adequate tidal volume, trachea was extubated when child was fully awake and placed in lateral position. He was shifted to high dependency pediatric unit for overnight monitoring which was uneventful. Postoperative analgesia was managed with paracetamol and ketoralac. The child was discharged after three days and was asked to follow up in the genetics clinic of our hospital for further management. Consent from the parents was obtained for publishing

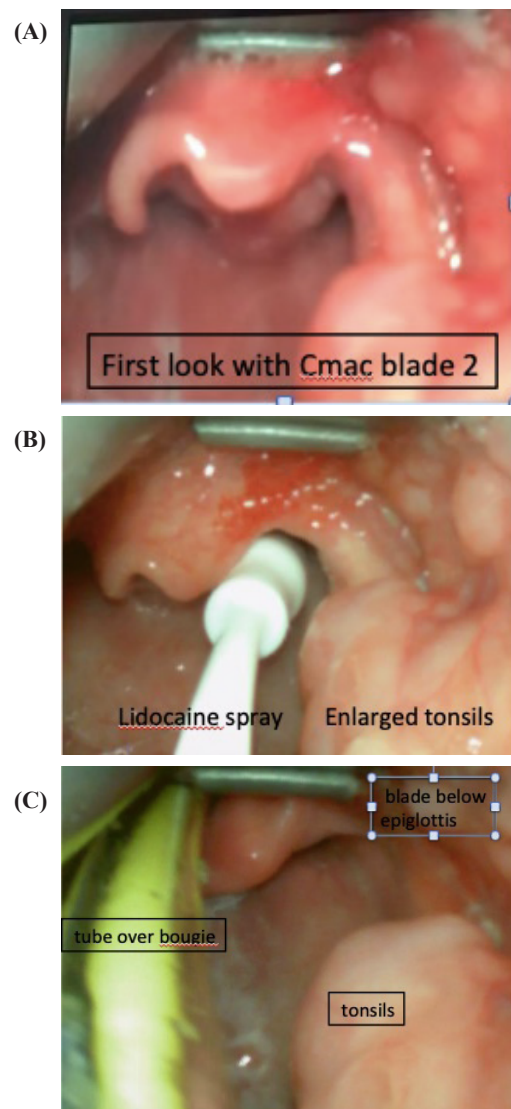


Figure 1. (A), CL IIIb view with Cmac blade 2 with OELM; (B), Lidocaine spray around vocal cords; (C), Visible posterior larynx with Cmac blade 3 kept below epiglottis.

this case report.

Difficulty in airway management reported in these patients include endotracheal intubation with a smaller size ETT after multiple attempts at intubation, stylet guided endotracheal intubation, view of CL grade 3 or 4 on laryngoscopy, ill fitting LMA, ineffective ventilation with LMA, difficult or unsuccessful FOB guided endotracheal intubation and obstructed laryngeal inlet due to incidental epiglottis polyp covering the larynx (5-10).

In the present patient as laryngoscopy view of CL grade IIIb did not improve with OELM due to large and fixed epiglottis, we planned to lift posterior surface of epiglottis with blunting of airway reflexes with topical lignocaine. During second laryngoscopy with size 3 C Mac blade posterior surface of epiglottis was lifted along with OELM which helped to visualize posterior part of glottis and bougie guided endotracheal intubation was successful. Ketamine administration helped in

maintaining spontaneous ventilation and deepening of anaesthesia without respiratory depression.

To conclude, airway in a patient of hunter syndrome can be secured by placing the a larger videolaryngoscope blade at posterior surface of epiglottis to view posterior part of larynx to facilitate bougie guided intubation. An opioid free anesthesia is helpful to prevent postoperative respiratory obstruction due to presence of airway abnormality and sleep apnea.

References

1. Walker RW, Darowski M, Morris P, Wraith JE. Anaesthesia and mucopolysaccharidoses. A review of airway problems in children. *Anaesthesia*. 1994; 49:1078-1084.
2. Walker R, Belani KG, Braunlin EA, Bruce IA, Hack H, Harmatz PR, Jones S, Rowe R, Solanki GA, Valdemarsson B. Anaesthesia and airway management in mucopolysaccharidosis. *J Inherit Metab Dis*. 2013; 36:211-219.
3. Frawley G, Fuenzalida D, Donath S, Yaplitto-Lee J, Peters H. A retrospective audit of anesthetic techniques and complications in children with mucopolysaccharidoses. *Paediatr Anaesth*. 2012; 22:737-744.
4. Mendelsohn NJ, Harmatz P, Bodamer O, Burton BK, Giugliani R, Jones SA, Lampe C, Malm G, Steiner RD, Parini R; Hunter Outcome Survey Investigators. Importance of surgical history in diagnosing muco-polysaccharidosis type II (Hunter syndrome): Data from the Hunter Outcome Survey. *Genet Med*. 2010; 12:816-822.
5. Herrick IA, Rhine EJ. The mucopolysaccharidoses and anaesthesia: A report of clinical experience. *Can J Anaesth*. 1988; 35:67-73.
6. Kaur J, Swami AC, Kumar A, Lata S. Anesthetic management of a child with Hunter's syndrome. *J Anaesthesiol Clin Pharmacol*. 2012; 28:255-257.
7. Suzuki K, Sakai H, Takahashi K. Perioperative airway management for aortic valve replacement in an adult with mucopolysaccharidosis type II (Hunter Syndrome). *JA Clin Rep*. 2018; 4:24.
8. Taguchi S, Kusunoki S, Fukuda H, Hamada H, Kawamoto M. Difficult tracheal intubation using airway scope in a pediatric patient with Hunter syndrome. *Masui*. 2009; 58:1278-1281. (in Japanese)
9. Busoni P, Fognani G. Failure of the laryngeal mask to secure the airway in a patient with Hunter's syndrome (mucopolysaccharidosis type II). *Pediatr Anaesth*. 1999; 9:153-155.
10. Kodani Y, Matsuura T, Mori T, Nishikawa K, Asada A. Anesthesia for three patients with Hunter syndrome. *Masui*. 2011; 60: 846-849. (in Japanese)

(Received June 3, 2019; Revised September 18, 2019; Accepted November 1, 2019)