CASE REPORT

Flexible bronchoscopy combined with videolaryngoscope for tracheal intubation in a child with Hunter syndrome: A case report

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Abstract

Hunter syndrome (mucopolysaccharidosis type II) has the highest reported prevalence of difficult tracheal intubation among the seven known types of mucopolysaccharidoses. Despite improved difficult airway guidelines and equipment, conventional approaches may fail in some cases. A 10-year-old child with Hunter syndrome, was scheduled for multiple dental extractions. On the first visit, failed intubation was declared as per Difficult Airway Society guidelines in the surgical day-care suite of our institute and the procedure was postponed. The case was then planned to be handled in the main operating room with additional preparation and input from the paediatric otolaryngologist for possible tracheostomy, paediatric intensive care for postoperative need for ventilation, and difficult airway resource faculty for an unconventional approach—videolaryngoscope combined with fibreoptic bronchoscope—which resulted in safe administration of anaesthesia. This case illustrates the importance of meticulous planning in the management of previously failed airway.

Keywords: Mucopolysaccharidosis II; Videolaryngoscope; Fibre optic bronchoscope; Difficult airway; Endotracheal intubation; Case report.

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Introduction

Hunter Syndrome (Mucopolysaccharidosis type II) is a metabolic disorder caused by inherited deficiency of the enzyme iduronate-2-sulfatase that is required for degradation of mucopolysaccharides (MPS).[1] These patients have the highest reported prevalence of difficult intubation among the seven known types of mucopolysaccharidoses. We, herein, present a structured approach to airway management in a child with MPS type II who previously had failed intubation.

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Case Report

A 10-year-old child weighing 25kg, was diagnosed with Hunter Syndrome at the age of three on September 19, 2013, at the Paediatric Clinic, Aga Khan University Hospital. He was then seen at the Dental Clinic, Aga Khan University Hospital on September 11, 2020 and was scheduled for multiple tooth extraction, due to dental abscess, under general anaesthesia. His past medical history was significant for recurrent chest infections, delayed milestones, sleep disordered breathing, and seizures. On examination, the child had coarse facies with large tongue and short stature. He was unable to lie flat and sat hunched forward with his tongue protruded out so he could breathe through the mouth adequately. (Figure 1)

Airway evaluation was compromised due to cognitive impairment. Bilateral occasional wheeze was audible on auscultation. Chest X-ray done two weeks ago showed bilateral air trapping and peri bronchial cuffing, probably representing bronchiolitis for which antibiotics were prescribed. Seizures were controlled with oral Lamotrigine.

The procedure was initially scheduled at the hospital's day

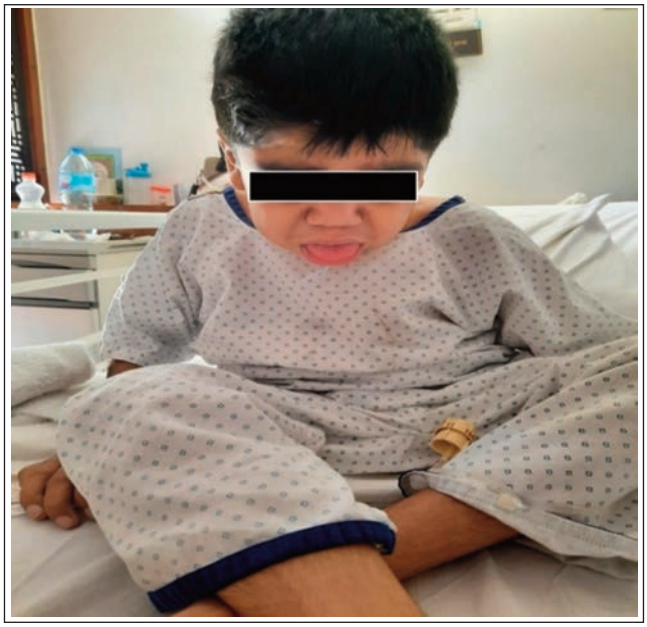

Figure-1: Preoperative status of the patient. Note the hunch forward posture with protruded tongue.

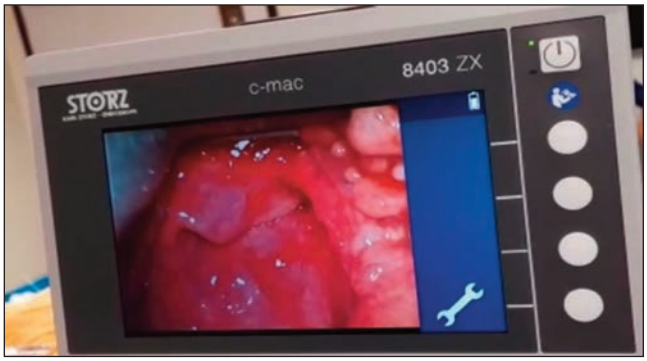

Figure-2: Glottic view with C-MAC videolaryngoscope. Percentage of glottic opening (POGO) visible is 0%.

care surgery unit on September 17, 2020, but the case had to be postponed due to failed intubation. First attempt with conventional Macintosh blade revealed a grade IV view with overhanging floppy epiglottis. A second attempt was made with C-MAC videolaryngoscope (VL), but was not successful due to 0% percentage of glottis opening (POGO) (Figure 2).

A third attempt was made with McCoy blade to help lift the epiglottis out of the way. The glottic view improved with McCoy but the bougie could still not be passed due to high anterior larynx. In between the laryngoscopy attempts, mask ventilation became inadequate and oxygen saturation dropped to < 90%. Positive pressure ventilation was maintained with laryngeal mask airway (LMA). After three unsuccessful attempts, failed intubation was declared. LMA was removed after the patient regained consciousness with no complications.

The case was then planned to be conducted in the main operating room with the child admitted as in-patient with discussion with Difficult Airway Resource Faculty. A Paediatric Otolaryngologist was taken on board, and an intensive care bed was booked. The child's parents were explained the risk of failed airway, need of emergency tracheostomy, and postoperative ventilation. Written and informed high-risk consent was taken. The child was admitted a day before and was nebulised with Ipratropium Bromide eight-hourly.

On the day of surgery (September 24, 2020), a case huddle involving two anaesthesia consultants, a trainee, and a senior technician was done. The preparation was reviewed and availability of difficult airway trolley, paediatric fibre optic bronchoscope (FOB), C-MAC videolaryngoscope with conventional and hyperangulated D-blade and two screens, was assured. Standard acetylsalicylic acid (ASA) monitoring was applied. Before the induction of anaesthesia, 4-L/min oxygen was applied via nasal cannula, Glycopyrrolate 0.1mg and Dexamethasone 2mg were administered intravenously. Inhalation induction with Sevoflurane 4-8% combined with small Propofol Boluses were given. LMA size 2.5 was inserted and Cisatracurium 3mg IV was administered after establishing successful ventilation on supraglottic device. Oxygen flow rate through nasal cannula was increased to 10 litres/min for apnoeic oxygenation. The senior consultant made an initial attempt via videolaryngoscope with hyper angulated D-blade and successfully visualised the glottic opening with moderate anterior lifting force probably helped by muscle relaxation. At this point, an attempt to pass the bougie failed due to steep angle towards anteriorly placed larynx.

Both fibre-optic bronchoscope (FOB) and VL with separate screens were then made ready. Reinforced tracheal tube (ETT) of the size 4.5 was mounted on FOB. One consultant performed videolaryngoscopy with D-blade to create space by moving the tongue away and visualised the glottic opening. The other consultant performed the flexible bronchoscopy and navigated the scope through a steep path and across the vocal cords until the carina was visualised. The tube passed over the bronchoscope into the trachea successfully and fixed at 15cm. Placement of the tube was confirmed with bilateral air entry and continuous waveform capnography. The patient remained haemodynamically stable throughout, and his oxygen saturation was well above 94% due to apnoeic oxygenation. Anaesthesia was maintained with Isoflurane 1.5-2% with oxygen and air. Nalbuphine 1.5mg and Paracetamol 375mg were used for analgesia.

In anticipation of the possible upper airway obstruction owing to large tongue, tissue oedema and floppy structures at extubation, nasopharyngeal airway was inserted before extubation. Due to unavailability of appropriate sized nasopharyngeal airway, a 5.0mm PVC endotracheal tube was used, measured and cut to correct size. It was warmed to make it soft and easy to insert in the nostril. Neuromuscular blockade was reversed and extubation was done after the child was fully awake, though he could not follow commands due to cognitive impairment. Right after extubation, a high-pitched sound was heard due to upper airway obstruction. The ENT team was consulted to rule out stridor that may threaten the airway. However, the sound was due to obstruction from macroglossia. Nebulisation with 0.5% Epinephrine was administered and after vigilant monitoring inside the OR for 40 minutes, the child was shifted to the post-anaesthesia care unit (PACU) with saturation of 100% on two litres of oxygen support, heart rate of 97 beats per minute and non-invasive blood pressure of 123/92mmHg. The patient was later shifted to the ward and discharged on analgesics and antibiotics the next day with no

respiratory complications. Consent to publish the case was duly signed by the parents in the ward.

Discussion

Difficult airway is an important concern for an anaesthetist managing a patient with Hunter syndrome. Thorough knowledge of the airway structure and pathophysiology play a key role in preoperative planning and intraoperative management. The involvement of respiratory system is multifactorial. First, accumulation of glycosaminoglycans (GAGs) in the upper airway causes enlargement of the larynx, adenoids, tongue, and epiglottis leading to reduced mobility and chronic partial obstruction.[2] Second, limited mobility of thoracic cage leads to restrictive lung disease.[3] Third, structural integrity of the trachea is compromised due to softening of the supporting cartilage with progressive accumulation of GAGs; the resulting tracheomalacia can cause critical airway obstruction.[2] Fourth, recurrent respiratory tract infections and copious secretions make the patients prone to laryngospasm. These patients also have coarse facial features, short neck, and obstructive sleep apnoea.[4]

The presence of multiple predictors of difficult airway warrants an awake fibre-optic intubation. However, mental retardation precludes performance of traditional awake fibre optic intubation. Published reports have shown successful use of supraglottic devices in patients with mucopolysaccharidoses to aid ventilation in difficult airway scenario.[4] However, the case was postponed owing to limitations of day care setting.

Videolaryngoscope has revolutionised airway management and become the preferred device in case of anticipated difficult airway. Megens et al compared the success rate of intubation using different devices in children with MPS: videolaryngoscope was on the top (89%), followed by conventional laryngoscope (67%) and fibre-optic scope (20%).[5]

The benefits of combining videolaryngoscope and fibre-optic bronchoscope in this case were two-fold. First, VL displaced the large tongue and created a space to visualise the glottic opening. Second, fibre-optic bronchoscope served the purpose of a flexible stylet or bougie, guided through the vocal cords under vision and avoided trauma to the surrounding structures. Successful utilisation of this technique was also reported in a child with MPS VI (Maroteaux-Lamy syndrome), when three attempts of laryngeal mask airway insertion failed, and intubation was accomplished with videolaryngoscope assisted by fibre optic bronchoscope.[6] Another method worth mentioning is supraglottic airway assisted fibre optic intubation. Chaudhuri et al intubated a patient with Morquio-Brailsford syndrome (MPS type IV) with the help of LMA classic as a conduit for fibre-optic to pass through the redundant pharyngeal tissue and reach the glottis with ease.[7]

One major concern was maintenance of oxygen delivery during intubation attempts. Apnoeic oxygenation with nasal cannula helped to prevent hypoxia throughout the process.[8] In a can't-intubate-can't-oxygenate (CICO) scenario, the front of the neck airway may be the final choice. Cricothyroidotomy is not advised for MPS patients whose laryngeal structures are often deformed by deposition of MPS making it technically difficult.[9] In the current case, the presence of ENT team in the operating room at the time of intubation and extubation was ensured. Special attention was paid to the extubation process as there are reports of unsuccessful wean off from mechanical ventilation after complicated intubation because of tracheobronchomalacia.[10]

Conclusion

The combined use of flexible bronchoscope and videolaryngoscope improves glottic visualisation and achieved successful intubation in our case when a single device had failed on previous occasion. Thorough airway assessment and case record review are essential when addressing challenging airways, as in this case. We recommend keeping a high degree of suspicion of difficulty in airway management for patients with Hunter syndrome. It's important to be extra prepared and have availability of advanced equipment and assistance.

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Conflict of interest: None.

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