

## Case Report

# Anesthetic management using an i-gel™ supraglottic airway for a pediatric patient with Hunter syndrome-a case report

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**Abstract:** Mucopolysaccharidoses (MPS) are a group of inherited, metabolic disorders caused by a lack of lysosomal enzymes. Among the various types of MPS, Hunter syndrome (MPS type II) is a rare X-linked disorder in which patients lack the lysosomal enzyme iduronate-2-sulfatase (IDS), which leads to the accumulation of glycosaminoglycans (GAGs). This can be a great concern to the anesthesiologist due to the involvement of several regions related to airway management. Here, we report a case of successful anesthetic management using a size 2 i-gel™ supraglottic airway in a 5-year-old child with non-suppurative otitis media and middle ear effusion who arrived at our operating room for myringotomy with ventilation tube insertion.

**Keywords:** Airway management, Hunter syndrome, i-gel™, pediatric

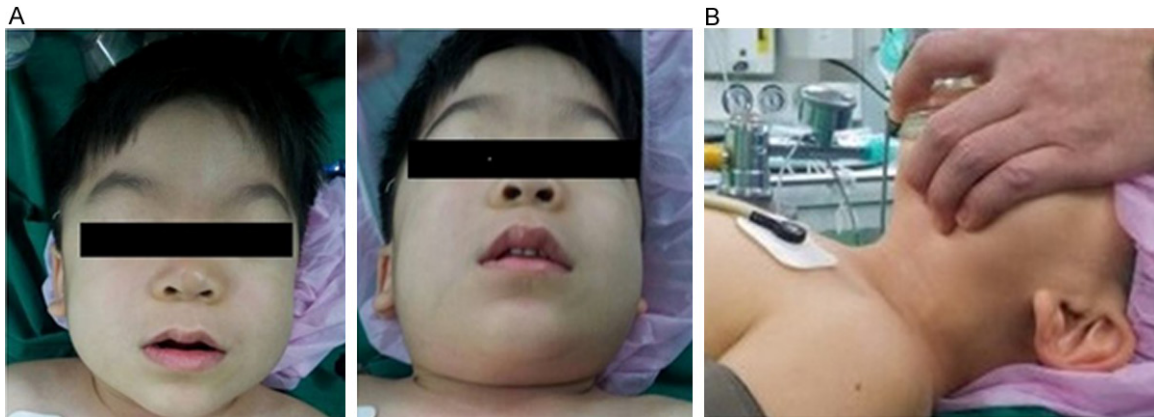
### Introduction and summary

Hunter syndrome, also called mucopolysaccharidosis type II (MPS II), is a rare X-linked metabolic disease caused by a deficiency of iduronate-2-sulfatase (IDS), a lysosomal enzyme that cleaves glycosaminoglycans (GAGs). Accumulation of GAGs within multiple organs and tissues can threaten safe perioperative management. Anesthesiologists must be aware of difficult airway management in such cases due to multiple risk factors: craniofacial abnormalities, limited mouth opening, hypertrophic tongue, short neck with difficult extension, hypertrophic tonsils and adenoids, and tracheal deformities [1]. Here, we present a case of successful airway management in a 5-year-old Hunter syndrome patient who was scheduled for bilateral myringotomy with ventilation tube insertion. Supraglottic mask devices have been reported to provide assistance in Hunter syndrome patients. However, use of the i-gel™ supraglottic airway in pediatric Hunter syndrome patients has not been reported previously. The airway in our patient was well managed with a size 2 i-gel™ during the operation,

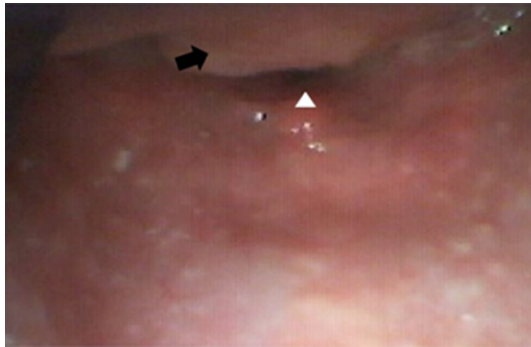
and the patient left the recovery room without any complications.

### Case report

A 5-year-old male (108 cm, 19.2 kg) was scheduled for elective myringotomy with ventilation tube insertion due to bilateral non-suppurative otitis media and middle ear effusion. The patient had been diagnosed with Hunter syndrome 2 years previously during evaluation for hepatosplenomegaly, coarse facies, and heart murmur. He also had a surgical history of bilateral inguinal herniorrhaphy and left hydrocelectomy at 2 months old. Physical examination revealed coarse facies, micrognathia, and a short neck with limited neck extension (**Figure 1A, 1B**). Mouth opening was normal and Mallampati grade was 4. The patient did not have any developmental disorders or intellectual disabilities. The results of preoperative laboratory tests, including common blood tests, liver function tests, and urinalysis, were all normal. MRI of the abdomen showed hepatomegaly with minimal splenomegaly, and echocardiography revealed moderate mitral regurgitation with mitral valve prolapse.



**Figure 1.** The patient had several airway management risk factors. A. Micrognathia, short neck, and a coarse face. B. Limited neck extension.



**Figure 2.** View obtained with a video-assisted laryngoscope (GlideScope®). Only the epiglottis (black arrow) and pharynx (white arrowhead) could be seen.

Difficult airway management was anticipated and various devices for airway control were prepared: endotracheal tubes of various sizes, a Macintosh laryngoscope blade, a video-assisted laryngoscope (GlideScope®; Saturn Biomedical Systems, Burnaby, BC, Canada), and an i-gel™ supraglottic airway (Intersurgical Ltd., Wokingham, UK). Intravenous access was obtained after monitoring electrocardiogram, noninvasive blood pressure, pulse oximeter, and end-tidal carbon dioxide (ETCO<sub>2</sub>). Smooth mask ventilation was confirmed after intravenously administration of 30 mg of ketamine. Rocuronium (10 mg) was administered intravenously and endotracheal intubation was first attempted with a Macintosh laryngoscope blade. However, only the tip of the epiglottis was visible. Blind intubation was not performed to avoid airway trauma. Instead we used a GlideScope® video laryngoscope, but there was only a slight improvement. Although we were

able to see the whole epiglottis, the vocal cords were still not visible (Figure 2). We next employed an i-gel™ (size 2) supraglottic airway as the operation duration was not only short but also did not involve the oral cavity (Figure 3). The patient was well ventilated (normal ETCO<sub>2</sub> wave and lung sounds by stethoscope) without any leakage in the oral cavity. Anesthesia was maintained with sevoflurane (1.5-2.0 vol%, FiO<sub>2</sub> 60%) and intravenous fentanyl (20 µg). The patient was ventilated using volume-control ventilation (tidal volume 8 ml/kg, respiratory rate 18/min) and the ETCO<sub>2</sub> was maintained at 34-38 mmHg. Total duration of anesthesia was 40 min, and pyridostigmine (0.3 mg/kg) and glycopyrrolate (0.08 mg/kg) were administered at the end of surgery for reversal of neuromuscular blockade. The i-gel™ was removed in the operating room after confirming full neuromuscular recovery. The patient was observed in the recovery room for 40 min and discharged to a general ward without any complications.

#### Discussion

Hunter syndrome, also called mucopolysaccharidosis type II (MPS II), is a rare X-linked metabolic disease caused by a deficiency of iduronate-2-sulfatase (I2S), a lysosomal enzyme that cleaves glycosaminoglycans (GAGs). The incidence is believed to be 1.3 per 100000 male live births [2, 3]. The lack of this enzyme leads to the accumulation of GAGs, and patients with Hunter syndrome show multisystemic symptoms, some which can be a great concern for the anesthesiologist [1].



**Figure 3.** Successful ventilation using a size 2 i-gel™.

The many risk factors of mucopolysaccharidosis—such as cardiac, pulmonary, spinal cord, and airway anomalies—have been well described previously [1, 4]. Indeed, although a different subtype from Hunter syndrome, mucopolysaccharidosis type I, has been shown to have a high rate of mortality [5]. In the present case report, we will focus on airway management, which is probably the matter of greatest concern during anesthesia in these patients. The continuous accumulation of CAGs in the upper airway can lead to several levels of obstructive risk factors: craniofacial abnormalities, limited mouth opening, hypertrophic tongue, short neck with difficult extension, hypertrophic tonsils and adenoids, and tracheal deformities [4]. Tracheostomy was reported to be necessary in Hunter syndrome patients for emergency airway management of non-ENT surgical procedures [6, 7]. A history of obstructive sleep apnea (OSA) is often present and may be a predictor of high risk during airway management [8, 9]. The presence of a progressive decrease in pulmonary function can also compromise airway management [4]. The patient described here had coarse facies, micrognathia, and a short neck with limited extension. Difficult intubation was anticipated and various airway management devices were prepared before anesthesia. Although we were able to mask ventilate the patient, the laryngoscopic view was grade 3 and the vocal cord was still not visible with a GlideScope®. Due to another important risk factor, endotracheal intubation itself, we did not

perform blind intubation. Even with successful intubation, post-obstruction pulmonary edema may occur after extubation and compromise the patient [10]. Supraglottic airway devices, such as laryngeal mask airway (LMA), have the advantage of obtaining a secure airway without direct trauma to the larynx. LMA has also been reported to be effective when performing intubation with a fiberoptic bronchoscope in mucopolysaccharidosis patients [11]. The second-generation LMA, i-gel™, has been shown to be adequate or possibly more effective in providing

airway management in normal, spontaneous breathing pediatric and adult patients when compared with previous LMA devices [12, 13]. The i-gel™ supraglottic airway has also been reported to be effective in an adult Hunter syndrome patient when performing intubation with a fiberoptic bronchoscope [14]. However, there have been no previous reports of the successful use of an i-gel™ in a pediatric Hunter syndrome patient. In the present case, a size 2 i-gel™ was effective for providing an adequate airway for myringotomy with ventilation tube insertion. Due to the relatively short surgical duration, a more definite airway was not attempted. Our present case suggests that i-gel™ may be an effective, non-traumatic airway management device in pediatric Hunter syndrome patients. However, as there have been only limited reports and the severity and consequences differ among patients, the anesthesiologist must prepare various and emergency airway management devices when providing anesthesia in Hunter syndrome patients.

### Disclosure of conflict of interest

None.

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