Article published online: 2022-09-20

LETTER TO EDITOR

Hunter's syndrome and the airway: Implications for the anesthesiologist - A correspondence

Sir,

A 7-year-old boy, a known patient of Hunter's syndrome, presented to our hospital with symptoms of bilateral sudden onset blindness and headache since last 1 month. Magnetic resonance imaging revealed obstructive hydrocephalus and subsequently the placement of a ventriculo-peritoneal (VP) shunt was planned. Preanesthetic evaluation revealed soft tissue hypertrophy of pharynx, short neck and a large tongue. On arrival to the operation theatre, an intravenous access was secured, and he was premedicated with glycopyrrolate and made sedated with titrated doses of propofol and midazolam. He being extremely uncooperative, awake fiber-optic bronchoscope intubation was not an option. Ventilation with mask was increasingly difficult, and patient started desaturating. We attempted intubation via Ambu® Pentx Airway Scope (PENTAX Europe GmbH; Hamburg, Germany), but no anatomical structure was identifiable, and the internal anatomy was found to be completely distorted. Within 3 min SpO_2 dropped to 45 and heart rate to 35/min. Immediately a number 2 laryngeal mask airway (LMA) was placed, and ventilation was found possible, following which the saturation and the heart rate improved. An endotracheal tube (ETT number 5.5, cuffed) was passed through the LMA; however, it went into the esophagus. Meanwhile the stomach was inflated to the extent that acceptable ventilation was almost impossible. We removed the LMA, and quickly deflated the stomach by passing a nasogastric tube. Immediately following this an intubation was reattempted with standard McIntosh laryngoscope. We could visualize only the tip of the epiglottis and could slide a pediatric bougie below it. An ETT (5.5 cuffed) was railroaded, connected to the breathing circuit and end-tidal CO_2 tracing and auscultation confirmed the correct placement of the tube. A pediatric bronchoscope was introduced through the tube, and it was adjusted after visualizing the carina. Anesthesia was deepened; muscle relaxant was injected and surgery was resumed. Rest of the surgical and postoperative course was uneventful.

Hunter's syndrome (mucopolysaccharidoses II) is a rare, X-linked disorder caused by a deficiency of the lysosomal enzyme iduronate-2-sulfatase. This leads to an accumulation of glycosaminoglycans in many tissues and organs, particularly affecting the nervous, cardiovascular, respiratory, and musculoskeletal systems. Thickening of the soft tissues enlarged tongue, short immobile neck and restricted mobility of the cervical spine, and temporo-mandibular joints make laryngoscopy and intubation difficult.

Published studies on anesthesia in Hunter syndrome reported failed or difficult intubation in 42% of patients.[1,2] A recent study has reported the incidence of difficult intubation in children with Hunter syndrome is 20 times more frequent.[3] Bronchoscopic intubation is the most appropriate technique for intubation. However, it is prudent to have a back-up plan for the establishment of an emergency airway. Following successful surgery, edema of the larynx and other tissues can make extubation difficult. Those who are unable to maintain an airway after extubation, require urgent reintubation or tracheostomy. For brief procedures and considering difficult intubations, LMA may provide adequate control of the airway.[4] However, LMA may not be quite safe for VP shunt placement.[5] Intravenous sedation in patients with Hunter's syndrome should only be administered only in a setting appropriate for general anesthesia.

Financial support and sponsorship

Nil.

Conflicts of interest

There are no conflicts of interest.

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Access this article online

Quick Response Code:

Website: www.asianjns.org

DOI: 10.4103/1793-5482.145534

How to cite this article: Chatterjee N, Davis J, Ganesamoorthi A. Hunter's syndrome and the airway: Implications for the anesthesiologist - A correspondence. Asian J Neurosurg 2017;12:334-5.