CASE REPORT

Difficult Airway Management in Pediatric Anesthesia: Insights from a Mucopolysaccharidosis Case

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ABSTRACT

This case report describes the challenging airway management of a 11-year-old boy with Mucopolysaccharidosis Type II (MPS II) who presented for cervical laminectomy. The combined use of dexmedetomidine and low-concentration sevoflurane effectively ~~in~~ managed a difficult airway in this case. Definitive airway management was achieved through Laryngeal mask airway (LMA) assisted fiber optic intubation providing a safe alternative for handling challenging airway situations in pediatric anesthesia

Key Words: *Mucopolysaccharidosis (MPS II) ŧ, Fiber optic intubation (FOI), Laryngeal mask airway (LMA), Obstructive sleep apnea (OSA), Enzyme replacement therapy (ERT)*

INTRODUCTION

Mucopolysaccharidosis is an inherited and progressive lysosomal disorder caused by a deficiency of enzymes involved in glycosaminoglycan degradation, which leads to the accumulation of glycosaminoglycans throughout the body, including skeletal structures, connective tissues, and internal organs. The patient exhibits features of dysostosis multiplex, heart valve disease, decreased pulmonary function, kyphoscoliosis, hepatosplenomegaly, and obstructive sleep apnea. Additional common issues include cervical spine instability, airway narrowing, hydrocephalus, visual and auditory deficits, intellectual impairments, inguinal hernia, and carpal tunnel syndrome. Patients often require multiple surgical interventions to address organ pathologies and extremity abnormalities. Anesthesia and airway management present significant challenges due to the unique clinical features, organ dysfunction, and facial deformities resulting from glycosaminoglycan accumulation in the body.

The purpose of this case report is to highlight the challenges of airway management in patients with mucopolysacchradosis, as well as to discuss the modern techniques and induction agents used for general anesthesia in cervical laminectomy.

CASE REPORT

An 11-year-old boy weighing 27 kg, diagnosed with MPS II, was scheduled for cervical laminectomy due to craniovertebral junction (CVJ) compression, under general anesthesia. Physical examination revealed a mouth opening of 4.5 cm and severely restricted head and jaw movement. His lower teeth could not protrude beyond the upper teeth, indicating limited mandibular protrusion. The Patient also exhibited limited submandibular space and a Mallappatti classification of grade IV. He had a history of snoring and obstructive sleep apnea but had unremarkable cardiovascular and respiratory findings.

In the pre-op area, the patient was nebulized with 2% lidocaine (4 ml) for 15 minutes. Following standard monitoring, glycopylorrate (0.2 mg) was administered through 22G cannula to manage secretions. The patient inhaled 100% oxygen for 3 minutes. Dexmedetomidine (0.2–1 µg/kg) was

administered incrementally over 20 minutes, followed by sevoflurane, which was initiated at 1% and titrated up to 4% with fresh gas flow of 6 liters/min. Adequate sedation was achieved within 30 seconds after inhalation.

When the end tidal CO_2 was >45 mmHg, a 2.5 Solus™ Standard laryngeal mask airway (LMA) was successfully inserted, allowing for capnography monitoring. The LMA was connected to a semi-closed anesthesia circuit and sevoflurane at 4% was inhaled to maintain anesthesia. After 3 minutes, a flexible bronchoscopy (5mm Olympus, BF-MP190F) was performed through the LMA, with 3 ml of lidocaine 2% sprayed around the vocal cords. A cuffed endotracheal tube (ETT) Hi-Lo curved 5.0 was then smoothly inserted into trachea, guided by the bronchoscope, with intubation confirmed by lung auscultation and CO_2 exhalation. The LMA was subsequently withdrawn, and SpO2 remained >95%. Spontaneous breathing was maintained throughout the procedure, ensuring continuous ventilation and reducing the risk of airway obstruction. Balanced anesthesia was maintained with 50% oxygen and 50% air at a flow rate of 2 liters with sevoflurane (1.5 -2%) and fentanyl (1ug/kg). Controlled ventilation achieved using atracurium (0.5 mg/kg). The patient was recovered in a lateral position with a 6.0-nasopharyngeal airway and was transferred to the postoperative area with an oxygen mask, without any complications.

DISCUSSION

Difficult intubation is a major concern during anesthesia, particularly in pediatric patients due to anatomical variations. In children with Mucopolysaccharidosis (MPS), a genetic disorder with seven types and 11 subtypes, airway complications arise in 25% of cases with an intubation failure rate of 8%. These rates increase to 54% and 23% respectively in Hurler's syndrome (MPS I).[1,2] MPS results from enzyme deficiencies that lead to glycosaminoglycan in tissues, contributing to features such as coarse facial structures, thickened mucosa, macroglossia, hypertrophic tonsils, hypoplastic mandible, and narrowed trachea all of which worsen with age. MPS I and MPS II are the more common subtypes, while MPS IV and MPS VI are associated with odontoid hypoplasia and atlanto-axial subluxation. Imaging techniques like MRI and cervical spine X-rays are crucial for assessing potential myelopathy. Enzyme replacement therapy (ERT) and hematopoietic stem cell transplantation (HSCT) alleviate airway challenges and halt disease progression.[2,3]

Our anesthesia plan involved fiber optic intubation through a LMA while maintaining spontaneous breathing. A key challenge was ensuring a patent airway prior to LMA insertion. We used dexmedetomidine, a centrally acting alpha2-adrenergic agonist, to induce sedation. Its effects include reducing sympathetic activity and inhibiting norepinephrine release. This was followed by sevoflurane, facilitating a balanced approach that lowered the required concentration of sevoflurane, minimized airway irritation, and preserved airway tone and spontaneous ventilation. This technique effectively addressed the anticipated challenges of mask ventilation, upper airway obstruction, and excessive secretions common in MPS patients, while minimizing trauma and hemodynamic instability.[4,5]

The LMA facilitated fiber optic intubation, avoiding neck extension in patients with cervical spine instability while preserving spontaneous breathing to minimize airway obstruction. We used incremental doses dexmedetomidine (0.2 to 1 µg) with sevoflurane, allowing LMA insertion within two minutes without respiratory complications though mild limb movements occurred. A 5mm fiber optic bronchoscope was then inserted, with topical anesthesia applied to prevent-breath holding and laryngospasm allowing smooth intubation. Airway backup plan (Emergency tracheostomy equipment) should always be available. Postoperative care should be carried out in a monitored environment, providing humidified oxygen and early physiotherapy to prevent respiratory complications and promote recovery.[4,5]

Effective pediatric airway management requires a thorough assessment, preparation, teamwork, and expertise in handling difficult airways. Anesthesiologists must be familiar with drugs like sevoflurane and dexmedetomidine to ensure safe anesthesia and airway management. Early identification of difficult airway in patients with MPS and fiber optic intubation with LMA

assistance offers a less invasive, safer approach for pediatric patients undergoing anesthesia for complex surgeries. Continuous education and training are essential for success in challenging pediatric airway management.

Conflict of interest: None

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