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Case Report

Anaesthesia Section

Anaesthetic Management for Patient of Mucopolysaccharidosis Undergoing Bilateral Functional Endoscopic Sinus Surgery: A Case Report

CHARMI HITENBHAI SHAH[1], POOJA ARPAN SHAH[2], JIGISHA BHARATBHAI MEHTA[3], SARA MARY THOMAS[4]

ABSTRACT

Mucopolysaccharidosis (MPS) represent rare, group of inherited lysosomal storage disease characterised by deficiencies in specific lysosomal enzymes involved in the metabolism of glycosaminoglycans. These conditions present significant anesthetic challenges due to progressive airway obstruction, cardiovascular complications, and skeletal abnormalities. Hereby, the authors present the case of a 12-year-old female patient with MPS who underwent bilateral Functional Endoscopic Sinus Surgery (FESS) and was successfully managed under general anaesthesia. The patient presented with characteristic features including short stature, short hands, receding mandible, short neck, corneal clouding, and mouth breathing. Her surgical history included two previous FESS procedures, the first 10 years ago and the second five years ago, both performed under general anaesthesia. She also had a history of recurrent convulsions, managed with antiepileptic therapy. Preoperative assessment revealed stable vital signs with a blood pressure of 95/60 mmHg. Anaesthetic management included premedication followed by induction, complicated by a difficult airway. Despite challenging mask ventilation, endotracheal intubation was successfully achieved. Maintenance of anaesthesia with sevoflurane provided stable intraoperative conditions. Surgery proceeded without complications, and the patient had a favourable postoperative recovery. The present case highlights the importance of thorough preoperative assessment, careful anaesthetic planning, and preparedness for airway management challenges in MPS patients undergoing Ear, Nose and Throat (ENT) procedures.

Keywords: Airway management, General anaesthesia, Lysosomal storage disorders, Paediatric anaesthesia

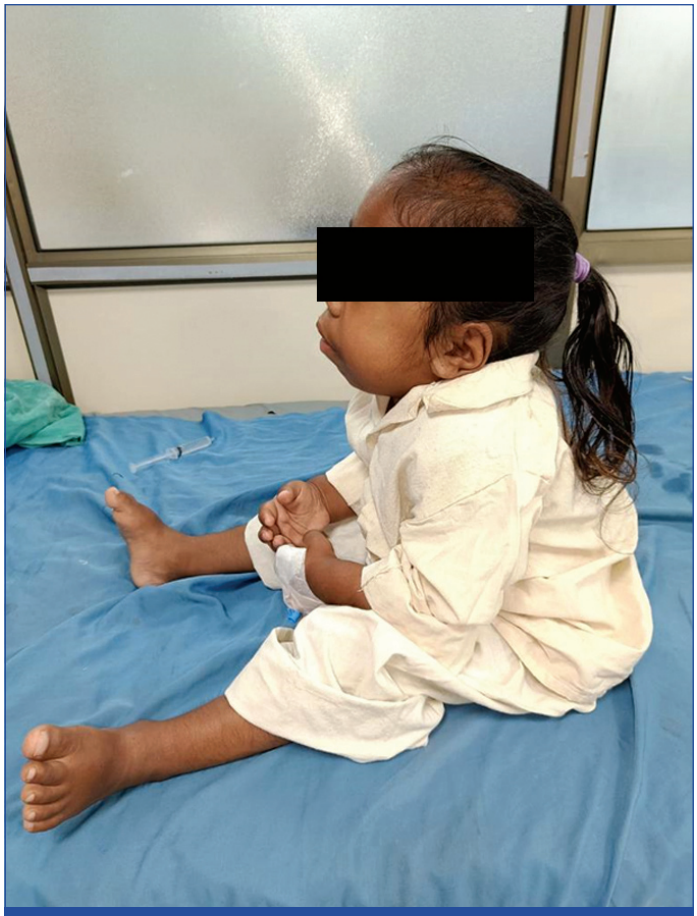
CASE REPORT

A 12-year-old female patient weighing 30 kg, a known case of MPS (Type II, American Society of Anaesthesiology (ASA) Physical Status III), was scheduled for bilateral FESS. She presented with characteristic phenotypic features suggestive of MPS, including short stature (height 125 cm), short hands and fingers, receding mandible, short neck with limited extension, corneal clouding, and persistent mouth breathing (Mallampati score IV) [Table/Fig-1-4].

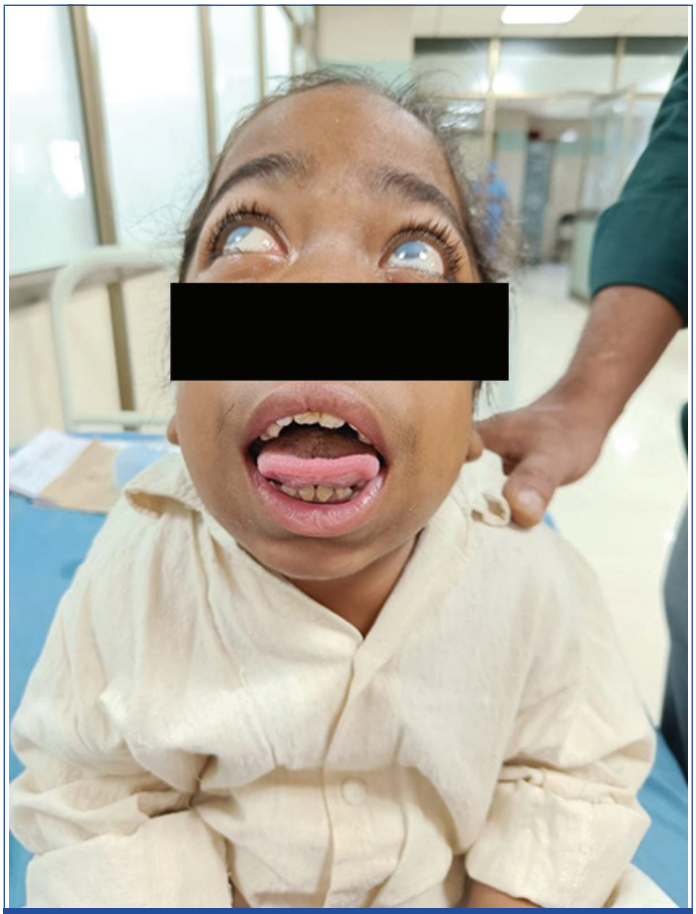
[Table/Fig-1]: Patient's frontal view showing characteristic facial features of MPS including corneal clouding and coarse facial features.

The patient had multiple co-morbidities, including epilepsy for four years, well-controlled on sodium valproate 200 mg twice daily, recurrent upper respiratory tract infections for the past six

[Table/Fig-2]: Patient's lateral profile demonstrating receding mandible and short neck.



[Table/Fig-3]: Patient in sitting position, lateral view showing postural features and skeletal abnormalities.



[Table/Fig-4]: Mouth opening assessment showing limited inter-incisor distance of 2.5 cm and macroglossia.

years, chronic rhinosinusitis for five years, mild hearing impairment noted three years ago, and no evidence of cardiac involvement on echocardiography.

The patient had undergone two previous FESS procedures, one 10 years ago and the other five years ago, both successfully performed under general anaesthesia. She had a significant history of recurrent convulsions since, age eight, managed with sodium valproate 200 mg twice daily (1-0-1). Her seizures were well controlled, with no episodes reported in the past six months leading up to the operation.

Family history was significant for consanguineous marriage (first cousins). Her elder brother (age 15 years) exhibited similar phenotypic features suggestive of MPS but remained undiagnosed. The parents reported a normal birth history with term delivery and a birth weight of 2.8 kg. Developmental milestones were delayed, with walking achieved at 18 months and speech development at 24 months.

Preoperative assessment: Vital signs were as follows: blood pressure 95/60 mmHg, pulse rate 88 beats per minute, respiratory rate 22 breaths per minute, and oxygen saturation 96% on room air. Cardiovascular examination revealed normal heart sounds with no murmurs. Respiratory examination showed bilateral wheeze with a prolonged expiratory phase. Neurological examination was normal, with no focal deficits.

Investigations: Laboratory investigations showed haemoglobin 11.2 g/dL, total leukocyte count 8,500/mm^3, platelet count 2.88 lakh/mm^3, and normal renal function tests [Table/Fig-5]. Chest radiograph demonstrated clear lung fields with no acute pathology. Electrocardiogram showed normal sinus rhythm. Echocardiography revealed normal cardiac function with no structural abnormalities.

Parameters	Value	Normal range
Haemoglobin	11.2 g/dL	11.5-15.5 g/dL
Total leucocyte count	8,500/mm^3	4,000-11,000/mm^3
Platelet count	2.88 lac/mm^3	1.5-4.5 lac/mm^3
Blood urea	25 mg/dL	15-40 mg/dL
Serum creatinine	0.8 mg/dL	0.6-1.2 mg/dL
Sodium	138 mEq/L	135-145 mEq/L
Potassium	4.2 mEq/L	3.5-5.0 mEq/L

[Table/Fig-5]: Preoperative laboratory investigations.

Anaesthetic Management

The patient was shifted to the operating theatre, and intravenous access was secured with an 18-gauge cannula. Total 500 mL of dextrose normal saline was commenced to maintain glucose homeostasis, considering the patient's reduced oral intake and metabolic demands. Standard monitoring, including pulse oximetry, non invasive blood pressure and electrocardiography, was applied.

Premedication included intravenous glycopyrrolate 0.08 mg, midazolam 1 mg, and fentanyl 50 mcg. Additionally, lignocaine 30 mg was administered as premedication for airway comfort. The patient was thoroughly preoxygenated with 100% oxygen for three minutes. During mask ventilation, significant difficulty was encountered due to the patient's anatomical abnormalities, particularly the receding mandible and short neck, which limited adequate neck extension. For induction, propofol 50 mg (1.7 mg/kg) was administered intravenously, followed by succinylcholine 50 mg intravenously for muscle relaxation. Direct laryngoscopy revealed a Cormack-Lehane Grade II view. Successful endotracheal intubation was achieved using a 5.0 mm cuffed endotracheal tube without packing.

Anaesthesia was maintained with oxygen and sevoflurane (2-3%). Intraoperative muscle relaxation was provided with atracurium 15 mg, with an additional 2.5 mg administered as required. Intraoperative vital signs remained stable throughout the procedure. The surgery lasted 90 minutes without complications. Estimated blood loss was minimal (<50 mL). The patient maintained stable haemodynamics, with blood pressure ranging from 90-105/55-65 mmHg, heart rate 85-95 beats per minute, and oxygen saturation 98-100%.

Following completion of the surgery, residual neuromuscular blockade was reversed with intravenous neostigmine 1.2 mg and glycopyrrolate 0.4 mg. The patient was successfully extubated without complications and transferred to the post-anaesthesia care unit in stable condition.

DISCUSSION

Mucopolysaccharidosis (MPS) represent a group of inherited lysosomal storage disorders characterised by the progressive accumulation of glycosaminoglycans in various tissues [1]. Anesthetic management of patients with MPS presents unique challenges due to multisystem involvement, particularly affecting the airway, cardiovascular system, and central nervous system [2].

The most significant concern in the anesthetic management of MPS patients is airway difficulty [Table/Fig-6] [3]. Progressive deposition of glycosaminoglycans in upper airway tissues leads to macroglossia, adenotonsillar hypertrophy, laryngeal narrowing, and tracheal stenosis [3]. These anatomical changes, combined with skeletal abnormalities such as atlantoaxial instability and cervical spine rigidity, create a scenario of anticipated difficult airway management.

System	Challenges	Management strategies
Airway	Macroglossia, short neck, limited mouth opening, laryngeal narrowing	Smaller endotracheal tubes, fibreoptic intubation, Laryngeal Mask Airway (LMA), emergency tracheostomy preparation
Respiratory	Restrictive lung disease, recurrent infections, sleep apnoea	Preoperative optimisation, postoperative monitoring, CPAP
Cardiovascular	Valvular disease, cardiomyopathy, coronary artery disease	Echocardiography, intraoperative monitoring, cardiology consultation
Neurological	Atlantoaxial instability, hydrocephalus, seizures	Careful positioning, antiepileptic continuation, neuroimaging
Other	Difficult venous access, hepatosplenomegaly, joint contractures	Ultrasound-guided access, modified positioning

[Table/Fig-6]: Anaesthetic challenges in Mucopolysaccharidosis (MPS) [3].

Several case reports have documented anesthetic management in MPS patients [4,5]. King DH et al., reported successful anesthetic management using fiberoptic intubation in eight patients with MPS, emphasising the importance of thorough airway assessment [4]. Similarly, Moores C et al., described anesthetic considerations in 31 patients with MPS, 28 of whom required anaesthesia, highlighting the progressive nature of airway involvement [3]. Cohen MA and Stuart GM demonstrated successful delivery of anaesthesia for 43 patients with MPS Type III using various techniques, including mask anaesthesia and laryngeal mask airways [6]. However, they emphasised that endotracheal intubation remains the gold standard for major procedures requiring controlled ventilation. The comparative analysis of anaesthetic management in MPS cases has been depicted in [Table/Fig-7] [2-4,6]. Walker R et al., conducted a comprehensive review of anesthetic management in MPS patients and found that difficult intubation occurred in 25% of cases, with the need for alternative airway devices in 18% of patients [2].

The study demonstrated that careful preoperative assessment and preparation for a difficult airway significantly improve outcomes. Comprehensive preoperative evaluation is crucial in MPS patients. Cardiovascular assessment should include echocardiography to detect valvular involvement and cardiomyopathy, which occur in upto 90% of patients with certain MPS subtypes [7,8]. Respiratory evaluation should assess for sleep apnoea, restrictive lung disease, and recurrent respiratory infections. Neurological assessment should focus on cervical spine stability, as atlantoaxial instability occurs in approximately 20% of MPS patients [9]. Magnetic resonance imaging of the cervical spine may be warranted in cases with suspected instability.

The choice of anesthetic technique depends on the procedure requirements and patient factors. General anaesthesia with endotracheal intubation remains the preferred approach for major procedures requiring controlled ventilation and airway protection. However, alternative techniques, including laryngeal mask airways and fiberoptic intubation, should be readily available. Intraoperative monitoring should be comprehensive, with particular attention to cardiovascular stability and airway pressures. The use of neuromuscular monitoring is essential to ensure adequate reversal of muscle relaxation before extubation, as residual weakness may precipitate respiratory complications in patients with pre-existing airway compromise.

Postoperative management requires vigilant monitoring for respiratory complications, particularly in patients with pre-existing airway abnormalities. Continuous pulse oximetry and consideration for high-dependency unit care may be appropriate. Pain management should balance adequate analgesia with the risk of respiratory depression.

Key considerations for practitioners managing MPS patients:

- Always prepare for difficult airway with multiple backup plans, including video laryngoscopy, fiberoptic bronchoscopy, and surgical airway equipment.
- Ensure the presence of an experienced Anesthesiologist and ENT surgeon for emergency tracheostomy if, needed.
- Consider awake fiberoptic intubation in patients with severe airway compromise.
- Use smaller-than-predicted endotracheal tube sizes due to subglottic stenosis.
- Maintain spontaneous ventilation until the airway is secured.
- Equipment readiness checklist
- Multiple sizes of endotracheal tubes (one to two sizes smaller than predicted)
- Video laryngoscope with Paediatric blades
- Fiberoptic bronchoscope
- Laryngeal mask airways of various sizes
- Emergency cricothyrotomy/tracheostomy kit
- Experienced ENT team on standby

Study	Patient details	Procedure	Airway management	Complications	Outcome
Present case	12-year-old female, MPS	Bilateral FESS	Direct laryngoscopy, ETT 5.0	Difficult mask ventilation	Successful
King DH et al., [4]	8 patients, MPS	Various	Fibreoptic intubation	None reported	Successful
Moores C et al., [3]	31 patients (28 requiring anaesthesia), MPS I	Adenotonsillectomy	LMA then ETT	Postoperative stridor	Required ICU
Cohen MA and Stuart GM [6]	43 patients, MPS III	Dental examinations, MRI scans, and minor surgical interventions	Fibreoptic intubation	Prolonged intubation	Successful
Walker R et al., [2]	Multiple patients	Various	Multiple techniques	25% difficult intubation	Variable

[Table/Fig-7]: Comparative analysis of anaesthetic management in MPS cases [2-4,6].
ETT: Endotracheal tube; ICU: Intensive care unit; MRI: Magnetic resonance imaging

CONCLUSION(S)

The present case demonstrates the successful anaesthetic management of a Paediatric patient with MPS undergoing bilateral FESS surgery. The key to a successful outcome was comprehensive preoperative assessment, meticulous planning for an anticipated difficult airway, and preparedness for potential complications. Despite challenging anatomy resulting in difficult mask ventilation, successful endotracheal intubation was achieved using conventional techniques.

The present case reinforces the importance of a multidisciplinary approach involving Anaesthesiologists, Surgeons, and other specialists in managing patients with complex genetic disorders. Continued research into optimal anesthetic techniques and long-term outcomes in MPS patients remains important for improving patient care. Healthcare providers managing MPS patients should be prepared for airway challenges, have alternative airway management techniques readily available, and maintain a high index of suspicion for cardiovascular and respiratory complications. With careful planning and appropriate precautions, complex surgical procedures can be performed safely in this challenging patient population.

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PARTICULARS OF CONTRIBUTORS:

1. Assistant Professor, Department of Anaesthesiology, Smt. Bhikhiben Kanjibhai Shah Medical Institute and Research Centre, Sumandeep Vidyapeeth (Deemed to be University), Vadodara, Gujarat, India.
2. Professor, Department of Anaesthesiology, Smt. Bhikhiben Kanjibhai Shah Medical Institute and Research Centre, Sumandeep Vidyapeeth (Deemed to be University), Vadodara, Gujarat, India.
3. Associate Professor, Department of Anaesthesiology, Smt. Bhikhiben Kanjibhai Shah Medical Institute and Research Centre, Sumandeep Vidyapeeth (Deemed to be University), Vadodara, Gujarat, India.
4. Professor and Head, Department of Anaesthesiology, Smt. Bhikhiben Kanjibhai Shah Medical Institute and Research Centre, Sumandeep Vidyapeeth (Deemed to be University), Vadodara, Gujarat, India.

NAME, ADDRESS, E-MAIL ID OF THE CORRESPONDING AUTHOR:
Charmi Hitenbhai Shah,
20, Netaji Society, Near Muktanand, Karelibaug, Vadodara-390018, Gujarat, India.
E-mail: charmi28shah@gmail.com

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