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Case Report

Anaesthesia Section

Challenging Anaesthetic Management of a Child with Hunter Syndrome Undergoing Adenotonsillectomy and Hernia Repair: A Case Report

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ABSTRACT

Hunter syndrome, also known as Mucopolysaccharidosis type II (MPS II), is a rare X-linked lysosomal storage disorder caused by a deficiency in iduronate-2-sulfatase. This deficiency leads to the accumulation of Glycosaminoglycans (GAG) which affects multiple organ systems in the body, presenting significant challenges in conduct of anaesthesia. The present report presents a case of four-year-old male child with MPS II who required two surgical procedures six months apart-adenotonsillar resection and hernia repair (umbilical and inguinal). The uniqueness of the present case lies in the use of two different anaesthetic approaches for the same patient: general anaesthesia with challenging airway management for the first surgery and spinal anaesthesia with sedation for the second, thus avoiding repeated airway instrumentation. Preoperative evaluation revealed characteristic features such as coarse facial features, an enlarged head, macrocephaly, macroglossia, a short neck, a Mallampati grade III airway, an enlarged liver and mild mitral valve prolapse with regurgitation. Anticipating airway difficulties, spontaneous ventilation was maintained during induction of general anaesthesia; direct laryngoscopy revealed a Cormack-Lehane grade III view and intubation was successfully performed on the first attempt using a paediatric bougie and a flexometallic endotracheal tube. For the subsequent hernia repair, regional anaesthesia was deliberately chosen to circumvent the known airway risks and the procedure proceeded uneventfully. Both surgeries resulted in smooth recoveries. The present case highlights the importance of comprehensive preoperative assessment, careful airway management and adaptable anaesthetic planning, including the strategic use of regional techniques, to ensure safety in children with MPS II who require multiple surgeries during childhood.

Keywords: Adenotonsillar resection, Difficult airway, Hernia Surgery, Mucopolysaccharidosis type II, Paediatric anaesthesia, Paediatric spinal anaesthesia

CASE REPORT

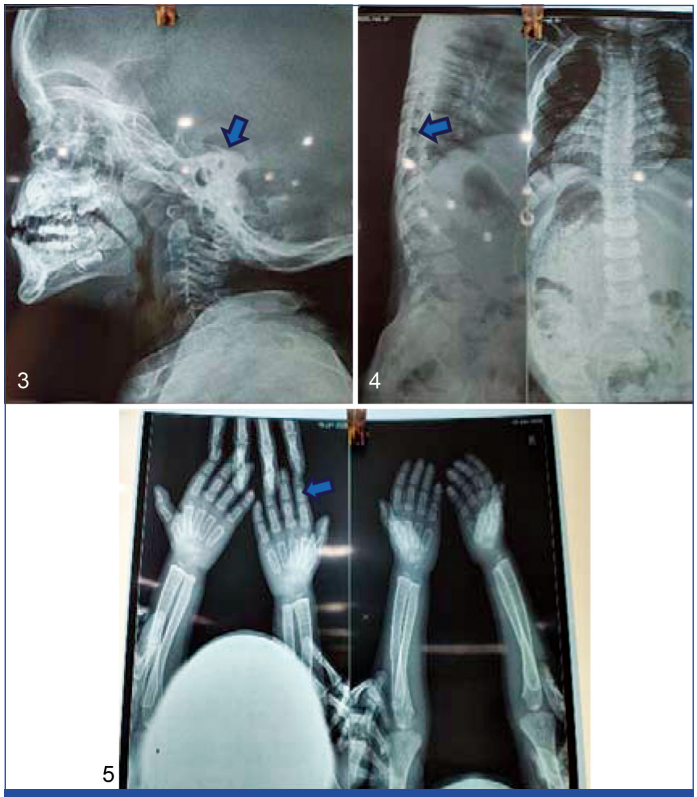
A four-year-old male child, fourth born of a non consanguineous marriage, diagnosed with mucopolysaccharidosis type II, presented to Pre-Anaesthesia Check-up (PAC) unit of Anaesthesia Department for his assessment for scheduled adenotonsillar resection. The child had a history of recurrent episodes of cough, cold and rhinorrhea and current episode had been present for five days. Birth history revealed term vaginal delivery at home with active cry and cord clamping by midwife. However, no antenatal visits and ultrasonography had been performed during pregnancy. Developmental milestones were delayed. Immunisation status was unknown, although Bacillus Calmette-Guérin (BCG) vaccine scar was present. Family history revealed death of the first-born male sibling at the age of three years due to an unknown cause for which no work-up or investigations were done as family was illiterate and belonged to lower socio-economic class.

On general examination, the child appeared well-built and nourished but had short stature (weight 16 kg, expected 16.5 kg; height 90 cm, expected 104 cm). He had active rhinorrhea and was on nebulisation with 3 mL 3% NaCl (Sodium Chloride) three times a day, Syrup Ambroxol Hydrochloride 7.5 mg three times a day and Syrup Cetirizine 10 mg at night and intravenous Augmentin 1.2 gm three times a day antibiotic course. His respiratory rate was 24/min with bilateral coarse crepitations on auscultation with 96% room air saturation. He had characteristic dysmorphic features including coarse facial features, broad nose with depressed nasal bridge, macrocephaly with prominent forehead and short neck. Thick coarse hair and pigmented thickened skin were present. The child also had short broad fingers and corneal clouding. Abdominal examination revealed distension with a pot-belly appearance [Table/Fig-1,2].

[Table/Fig-1,2]: Typical phenotype feature of the child showing dysmorphism.

Airway examination revealed mouth opening of approximately 2.5 cm, macroglossia, Mallampati grade III airway and grade III enlarged tonsils. Neck flexion and extension were mildly restricted. The child also had a history of mouth breathing and snoring. Urine examination using the toluidine blue spot test confirmed the presence of mucopolysaccharides. Enzyme assay revealed deficiency of iduronate-2-sulfatase. Routine haematological investigations were within normal limits. Two-dimensional echocardiography revealed mild mitral valve prolapse with mild mitral regurgitation and normal ventricular function without pulmonary arterial hypertension. Ultrasonography of the abdomen revealed hepatomegaly (12 cm) and enlarged lymph nodes in the pre-aortic, bilateral para-aortic and mesenteric regions. Radiographic evaluation demonstrated bilateral proximal pointing of the second and fifth metacarpals and anteroinferior beaking of

multiple vertebral bodies, findings consistent with dysostosis multiplex associated with Mucopolysaccharidosis II. Chest radiograph showed increased transverse cardiac diameter suggestive of cardiomegaly with cardiothoracic ratio of 0.61. Multiple oar-shaped ribs were noted. Skull X-ray demonstrated macrocephaly with a J-shaped sella. Lateral nasopharyngeal radiograph revealed prominent posterior nasopharyngeal soft tissue (~16 mm) causing narrowing of the nasopharynx, suggestive of adenoid hypertrophy [Table/Fig-3-5].



[Table/Fig-3-5]: Radiological examination of the child. **[Table/Fig-3]:** A large skull with a thickened vault and a J-shaped sella turcica and narrowing of nasopharynx with enlarged adenoids. **[Table/Fig-4]:** Mild thoracolumbar kyphosis notched anterior part of the vertebral bodies. **[Table/Fig-5]:** Sharply pointed metacarpal bones in the proximal part.

After detailed evaluation and discussion with the paediatric team, otorhinolaryngology {Ear, Nose and Throat (ENT)} team and parents, high-risk informed consent was explained and signed by the parents. The child was classified as American Society of Anaesthesiologists (ASA) III because of the anticipated difficult airway and the presence of an active Upper Respiratory Tract Infection (URTI), both of which increased the anaesthetic risk. The recurrent URTI episodes were attributed to adenotonsillar hypertrophy and were expected to resolve only after definitive surgical treatment. General anaesthesia with endotracheal intubation was planned for the present case with difficult airway cart prepared in view of anticipated difficult airway. Difficult airway cart included paediatric fibre-optic bronchoscope, channelled/non channelled anatomical videolaryngoscope (size 3,4), direct laryngoscopes with miller and macintosh blades (size 2,3,4), supraglottic airway devices including I-gel and ProSeal laryngeal mask airway (sizes 1.5, 2 and 2.5), oral/nasal airways, different sizes of endotracheal portex and flexometallic tubes, cricothyroidotomy set, tracheostomy tray (to be performed by ENT surgeons).

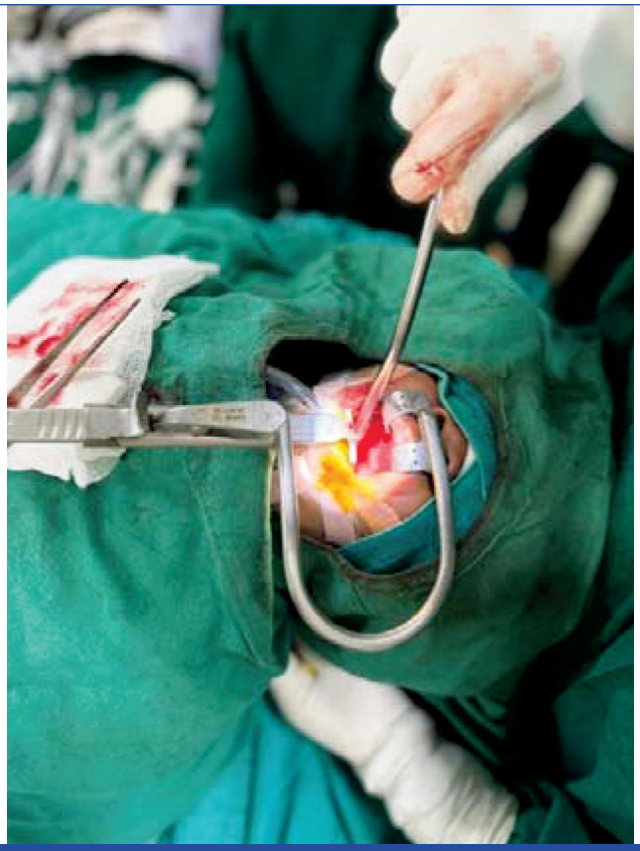
Primary plan of securing airway was fibre-optic followed by videolaryngoscope and direct laryngoscope in case of failure to achieve glottic view and securing airway.

The patient was kept nil per oral for six hours before surgery for solids and two hours for liquids. On the morning of surgery, the child was extremely uncooperative. Dexmedetomidine 1 µg/kg was given (0.5 µg/kg per nostril) intra-nasally through atomiser and Eutectic Mixture Of Local Anaesthetic Agent (EMLA) patch was applied at pre-decided cannulation site in preoperative area. Patient was shifted in operation theatre but was extremely uncooperative. As the patient was not allowing application of monitors also, 22 gauge intravenous line was secured using mild restraint. Glycopyrrolate 0.1 mg, Ondansetron 4 mg, Midazolam 0.5 mg and Ketamine 10 mg were given intravenously. After mild sedation, the child became calm and cooperative, allowing application of routine monitoring including electrocardiogram, non invasive blood pressure, pulse oximetry and temperature monitoring. Baseline parameters were heart rate 106/min, blood pressure 110/76 mmHg and oxygen saturation 97%. Although fibre-optic bronchoscopy was primary plan for securing airway, but due to poor cooperation of the child and risk of airway collapse with excessive sedation, it was reserved as a backup option. Pre-oxygenation with 100% oxygen was performed for three minutes. Induction was achieved with propofol 20 mg followed by assessment of mask ventilation. After confirming ability to ventilate, an additional 10 mg propofol was administered to deepen the plane of anaesthesia. Muscle relaxant was withheld initially to avoid airway collapse.

Once adequate depth of anaesthesia was achieved, channelled videolaryngoscope with blade no. 3 was used to visualise the glottis; however due to macroglossia and comparatively limited mouth opening, its blade could not reach the vallecula and glottic visualisation could not be achieved despite changing the size and type of videolaryngoscope blade (non channelled). Direct laryngoscopy with Macintosh curved blade size 4 was then attempted. Laryngoscopic view was Cormack-Lehane grade III and the epiglottis could not be lifted. Optimum External Laryngeal Manipulation (OELM) did not improve visualisation.

A blind attempt was made to introduce a paediatric bougie beneath the epiglottis. Tracheal ring sensations were appreciated on passing bougie, confirming tracheal placement. A 4.5 mm flexometallic endotracheal tube was rail-roaded over the bougie and successfully placed in the trachea. Correct placement was confirmed by bilateral equal air entry and capnography. The tube was fixed at 16 cm and cuff inflated with 3 mL air. Following endotracheal tube fixation and stabilisation in midline, neuromuscular blockade was achieved with vecuronium 2 mg and the patient was placed on mechanical ventilation with tidal volume 130 mL, respiratory rate 18/min, Positive End-Expiratory Pressure (PEEP) 5 cm water (H_2O) and Fraction of Inspired Oxygen (FiO_2) 0.5 [Table/Fig-6,7].

[Table/Fig-6]: Images of management of difficult airway with direct laryngoscope with bougie-guided intubation.



[Table/Fig-7]: Conduct of adenotonsillar resection surgery.

Adequate suctioning was performed and oropharyngeal packing was done to prevent aspiration during surgery. Anaesthesia was maintained with Sevoflurane (0.8-1.5%) in a fresh gas flow of 2 litres/min (containing mixture of 50 % oxygen and 50% medical air) along with intermittent maintenance doses of vecuronium.

The patient was positioned in Rose's position with a bolster placed under the shoulders. Bilateral equal air entry was reconfirmed after positioning. Because the adenoids and tonsils were markedly enlarged, the procedure took 120 minutes to complete. 450 mL intravenous fluids (Ringer lactate) were administered. Estimated blood loss during surgery was approximately 100 mL. For postoperative analgesia, Lignocaine 2% soaked cotton pellets were placed at tonsillar bed for 2-3 minutes and Paracetamol 240 mg given intravenously. Dexamethasone 4 mg and Hydrocortisone 30 mg was given intravenously to reduce oedema.

At the end of surgery, Sevoflurane was gradually tapered and haemostasis was meticulously ensured by the surgeons. Neuromuscular blockade was reversed with Sugammadex 40 mg.

After adequate spontaneous breathing, restoration of muscle tone and spontaneous eye opening, the patient was successfully extubated. The child was active and crying and was transferred to the post-anaesthesia care unit for monitoring. After six hours of observation, oral intake was initiated and the patient was shifted to the paediatric ward in stable condition.

Later after six months, the child again presented with umbilical and right-sided inguinal hernia in Paediatric surgery unit and then was referred to PAC unit for assessment. Umbilical hernia was measuring approximately 1.5×2 cm and inguinal hernia was 7×15×12 mm containing bowel loop and omental fat. All other preoperative anaesthetic findings were similar to previous examination which were suggestive of an anticipated difficult airway. The authors planned this surgery under spinal anaesthesia to avoid airway instrumentation. In case of failure of spinal anaesthesia, plan B was to administer general anaesthesia with supraglottic airway device (I-gel No. 2). After adequate starvation period, on the day of surgery as done before, EMLA patch was applied at anticipated cannulation site and spinal injection site. The child was uncooperative for application of monitors; intravenous cannulation was done using mild restraint and then he was premedicated and mildly sedated with Glycopyrrolate 0.1 mg, Ondansetron 4 mg, Midazolam 1 mg and Ketamine 15 mg. Patient's breathing was spontaneous and regular. Sevoflurane 1-2% along with 6 litres/min Oxygen was given through Jackson Rees circuit and then child was given lateral position. After aseptic and antiseptic precautions, Bupivacaine Heavy 0.5 % 1.2 mL (6 mg) with 2 µg Dexmedetomidine was given intrathecally in L1-L2 intervertebral space through midline approach.

Patient was then made supine. As the patient was sedated, onset of motor block was assessed by blunting of superficial abdominal reflexes. Sensory block was assessed with pin prick method and facial grimacing was observed. After achieving adequate surgical anaesthesia till eighth thoracic dermatome level, surgical commencement was allowed. Oxygen supplementation with nasal prongs with 1 litre/min was given. Patient remained mildly sedated throughout the surgery without any requirement of anaesthetic/sedation supplementation. The surgical procedure got completed in 60 minutes. At the end of surgery, child was awake, free of pain and spinal anaesthesia had regressed to partial tenth thoracic dermatome and bromage III motor block. Patient was shifted to post-anaesthesia care unit for further observation. Patient had full sensory and motor recovery 3.5 hours after spinal injection. Patient was given Paracetamol 240 mg intravenously for postoperative analgesia.

DISCUSSION

Mucopolysaccharidosis type II (MPS II) was first described by Dr. Charles Hunter in 1917 and after him it is also referred as Hunter syndrome. It is an X-linked lysosomal storage disorder characterised by GAG accumulation in the lysosomes. [1,2] In this disorder, there is genetic mutation of Iduronate-2-sulfatase (IDS) gene coding for iduronate-2-sulfatase enzyme which helps in breakdown of GAGs. Being X-linked genetic disorder, it predominantly affects males, who inherit the mutated gene from their asymptomatic carrier mothers. Female carriers are usually unaffected but may rarely manifest symptoms due to skewed X-chromosome inactivation. Onset, severity, manifestation and progression of the disease vastly vary among the affected male individuals [3]. GAG accumulations gradually cause coarse facial features, macroglossia, adenotonsillar hypertrophy, pharyngomalacia, glossoptosis, laryngomalacia, arytenoid, epiglottis and vocal cord thickening, tracheomalacia and bronchomalacia. These changes lead to distortion of the airway anatomy, progressive airway narrowing and reduced airway compliance, increasing the risk of airway obstruction both during induction of anaesthesia and at the time of extubation [4]. Difficult mask ventilation and endotracheal intubation have been reported to be as high as 35 - 50% in these patients [3]. These patients are also susceptible to potential "Cannot Intubate, Cannot Ventilate" (CICV) scenarios, necessitating the need for emergency front-of-neck surgical access [5].

Apart from airway-related anatomical changes, GAG accumulation leads to progressive multisystem involvement, including macrocephaly, a short neck with restricted cervical mobility, skeletal abnormalities (dysostosis multiplex), hepatosplenomegaly and valvular heart disease [1]. Together, these features make anaesthetic management highly challenging. Consequently, the expected benefit of any surgical intervention must be carefully weighed against the anaesthetic risk [3,6].

With disease progression, as multiple organs and systems get involved, multiple surgical procedures are required to treat associated symptoms. These include ear, throat and nose problems, skeletal and articular malformations, hernias, hydrocephalus and heart valve disorders, needing multiple anaesthesia exposures [7]. Very often these children undergo these surgeries under anaesthesia even before the diagnosis of MPS [8]. Another important finding

is the higher recurrence rate of adenoid hypertrophy after adenoidectomy and recurrent herniations after hernia repair in the MPS population is higher than in healthy children [8]. Thus, meticulous pre-anaesthetic assessment is vital to identify potential risks involved and preparedness for unforeseen outcomes [8]. It should involve a detailed airway examination, systemic examination and investigations like complete blood profile, echocardiography and other radiological imaging, when indicated to evaluate skeletal abnormalities. Cardiac involvement is frequently seen in patients with MPS II in form of valvular dysfunction, cardiomyopathy, or conduction abnormalities [9]. These cardiac manifestations influence anaesthetic drug selection and require perioperative cardiovascular monitoring. In the present case, there were typical phenotypic, radiological features [5] and mild mitral valve prolapse with mitral regurgitation on echocardiogram. Diagnosis was confirmed by urine examination using the toluidine blue spot test and Enzyme assay revealing deficiency of iduronate-2-sulfatase.

Enzyme replacement therapy when introduced earlier has been found to greatly reduce the incidences of airway-related complications due to reduction in GAG accumulation in airway tissues [10]. In the present case, due to unawareness and lower socio-economic class, parents had not realised possible reason for death of first-born son with similar phenotypic features due to MPS. Diagnosis of MPS was made in the second child after identification of typical phenotypic features and enzyme assay study. Parents were explained about the available enzyme replacement treatment options and requirement of genetic counselling.

Children with MPS exhibit a high prevalence (up to 89%) of Obstructive Sleep Apnoea (OSA) due to upper airway obstruction caused by adenotonsillar hypertrophy, which leads to snoring and mouth breathing. Additionally, cervical spine instability and odontoid dysplasia may compress the spinal cord, resulting in central sleep apnoea [8]. In the present case, parents had confirmed mouth breathing, snoring, sleep disturbances and day-time sleepiness of the child, although no sleep study was done on the child. As these symptoms get noticed/identified early, adenotonsillectomy is one of the most frequently performed surgical procedures in MPS II patients [10].

Many anaesthetists who have encountered and reported cases of MPS II have emphasised the high incidence of difficult laryngoscopy and tracheal intubation due to abovementioned causes [10,11]. While dealing with anticipated difficult airway in such cases, maintenance of spontaneous ventilation during induction is highly advisable mainly to minimise the risk of complete airway obstruction. Fibre-optic guided intubation is usually considered gold standard technique for difficult airway management. It aids visualisation and securing the airway while preserving spontaneous breathing. However, in MPS II paediatric patients, fibre-optic intubation may become technically challenging due to poor patient cooperation, inability to provide deep sedation due to risk of airway collapse, limited airway space and the presence of excessive secretions [8,12].

In the present case, comprehensive difficult airway plan was prepared prior to induction of anaesthesia. Plan A was fibre-optic guided intubation, but it could not be performed due to poor patient cooperation and excessive secretion (due to URTI). Thus, following plan B, videolaryngoscope was used but glottis visualisation could not be achieved. As final airway rescue plan, direct laryngoscopy was done which revealed a Cormack-Lehane grade III view and successful endotracheal intubation was achieved using a bougie-assisted technique. The availability of alternative airway devices and a prepared difficult airway cart played a crucial role in ensuring safe airway management.

Although supraglottic airway devices have been successfully used in some surgical procedures in patients with mucopolysaccharidosis, their insertion can be difficult due to reduced mouth opening and macroglossia and also applicability may be limited in oral surgeries [9]. In the present case, supraglottic airway devices such as I-gel and ProSeal LMA (sizes 1.5, 2 and 2.5) were kept ready as part of the difficult airway preparation. However, because adenotonsillectomy requires a shared airway and unobstructed surgical access to the oropharynx, placement of a supraglottic airway device was not feasible unless required as a rescue device for difficult ventilation or failed intubation.

Various airway management techniques have been described in patients with mucopolysaccharidosis, reflecting the complexity and variability of airway anatomy in these individuals. A review of previously published reports demonstrates that successful airway management often depends on anticipating difficulty and preparing multiple backup strategies. Selected case reports and series describing airway management approaches in MPS patients are summarised in [Table/Fig-8] [6,10,12-18].

Author's name/ year	Patient age	Surgery/procedure	Specific airway instrument/ technique	Airway outcome	Perioperative complications/notes
Lao HC et al., 2022 [6]	51 patients (mean age ~7 years)	ENT surgery, orthopaedic surgery, neurosurgery, dental procedures, abdominal surgery	Macintosh direct laryngoscope, GlideScope videolaryngoscope, fiberoptic bronchoscope, Laryngeal Mask Airway (LMA)	Advanced airway devices improved intubation success; videolaryngoscopy and fiberoptic techniques frequently required	Difficult mask ventilation, difficult laryngoscopy, failed first-pass intubation, postoperative Intensive Care Unit (ICU) monitoring in severe airway disease
Machado APG et al., 2022 [10]	10 patients (mean age 8.8 years)	Hernia repair, ENT procedures, orthopedic surgery, imaging procedures	Fiberoptic bronchoscope-guided intubation, videolaryngoscope, supraglottic airway devices	Successful airway management achieved with individualised airway planning	Difficult intubation, airway oedema, need for postoperative ICU observation in selected patients
Frawley G et al., 2012 [12]	17 patients (Children aged 0.8-18 years)	ENT surgery, orthopaedic surgery, Magnetic Resonance Imaging (MRI), dental procedures	Direct laryngoscopy, fiberoptic bronchoscopy, classic LMA, intubating LMA	Difficult airway encountered in large proportion of anaesthetics; supraglottic devices useful rescue adjuncts	Desaturation, difficult mask ventilation, multiple intubation attempts, postoperative airway obstruction, ICU admission
Altunkaya N et al., 2019 [13]	18-years	Incarcerated inguinal hernia repair	Spinal anaesthesia with no airway instrumentation	Surgery completed successfully while avoiding airway manipulation	No airway-related complication reported
Kumar KR et al., 2016 [14]	12-years	Inguinal hernia repair	Spinal anaesthesia; difficult airway cart and fiberoptic bronchoscope kept ready as backup	Successful surgery under spinal anaesthesia without need for intubation	Avoided airway instrumentation because of anticipated difficult airway
Kaur J et al., 2012 [15]	4-years	Tonsilletomy, inguinal and umbilical hernia	Ventilation with LMA, second attempt intubation with direct laryngoscopy	Successful ventilation with LMA followed by intubation	Difficult airway due to macroglossia, short neck, restricted neck movement
Seetharamaiah S et al., 2016 [16]	8-years	Surgery under general anaesthesia	Ambu laryngeal mask airway (Ambu LMA)	Successful ventilation and airway maintenance using Ambu LMA	Demonstrated effectiveness of supraglottic airway in anticipated difficult airway

Zahra R et al., 2023 [17]	7 children (3-12 years)	Multiple surgeries including hernia, ENT and orthopedic procedures	C-MAC videolaryngoscope, fiberoptic bronchoscope, supraglottic airway devices	Successful airway management with careful preoperative planning and using videolaryngoscope, LMA, fibre-optic	Difficult mask ventilation, difficult laryngoscopy, postoperative airway observation required
De Vuyst R et al., 2023 [18]	23 male patients (3-17 years receiving idursulfase)	Diagnostic airway evaluation study	Flexible bronchoscopy, CT airway imaging	Persistent airway abnormalities identified despite enzyme replacement therapy	Tracheomalacia, airway narrowing, macroglossia, supraglottic thickening documented

[Table/Fig-8]: Literature review – Paediatric Hunter Syndrome (MPS II) anaesthesia case reports [6,10,12-18].

Also, incidences of postoperative airway complications in patients with mucopolysaccharidosis is higher due to airway oedema, residual airway obstruction and underlying obstructive sleep apnoea which may lead to postoperative respiratory compromise [6]. Therefore, meticulous extubation planning and postoperative monitoring are equally essential to ensure safe anaesthetic outcome. These patients may require prolonged observational stay in the post-anaesthesia care unit or intensive care unit depending on the severity of airway abnormalities, co-morbid conditions and surgical needs.

When such patients require abdominal or lower limb surgeries, selection of anaesthetic technique has to be meticulously decided considering anticipated difficult airway, skeletal deformities, other systemic involvements, duration of surgery along with experience of anaesthesiologist and surgeon. Accumulation of GAGs can impact both peripheral and central neural structures, thereby affecting the safety and efficacy of neuraxial anaesthesia. It can get deposited within the meninges, epidural tissues, ligaments, vertebral column and peripheral nerves resulting in spinal canal stenosis, thickening of the dura and ligamentum flavum, vertebral deformities and compression neuropathies. These pathological changes can distort normal spinal anatomy, reduce cerebrospinal fluid volume and alter the distribution of local anaesthetics, leading to technically challenging neuraxial block placement as well as unpredictable sensory and motor blockade. Furthermore, these patients may have pre-existing cervical or thoracolumbar cord compression and neurological deficits, raising concerns regarding potential neurological deterioration following neuraxial techniques and complicating postoperative neurological assessment. Skeletal abnormalities such as kyphoscoliosis and gibbus deformity may further exacerbate procedural difficulty [12]. GAG also produces toxic effects on neurons leading to failure of regional techniques [11,12].Thus, spinal anaesthesia in patients with mucopolysaccharidosis necessitates careful patient selection, comprehensive neurological evaluation and an individualised anaesthetic plan. Despite these challenges, successful use of spinal anaesthesia has been documented in selected patients with mild disease and no significant spinal pathology, offering the advantage of avoiding airway manipulation in a population known for difficult airway management [13,14].

But as the patient may not be cooperative as seen in the present case, they may require sedation during administration of regional anaesthesia (subarachnoid block) along with mild sedation throughout the surgery [6,12]. Given its relative safety and ensuring the availability of a difficult airway cart in case conversion to general anaesthesia, spinal anaesthesia was selected as primary plan of anaesthesia for this child and general anaesthesia with supraglottic airway device was secondary plan. Sedation was administered while preserving spontaneous breathing and intrathecal bupivacaine with dexmedetomidine as an adjuvant was used to prolong the duration of the spinal block [19]. Some authors have also documented case reports of hernia repair under general anaesthesia using supraglottic airway device and endotracheal intubation [15,16].

The present case adds to the existing literature by demonstrating that even in the presence of a poor laryngoscopic view, successful intubation can be achieved using a bougie-guided technique when a structured difficult airway plan is implemented. Additionally, the case highlights the practical challenges of fibre-optic intubation in paediatric patients, particularly due to limited cooperation and restricted airway space. Spinal anaesthesia with mild sedation was found to be safe and effective for abdominal surgeries. The authors experience reinforces the importance of maintaining spontaneous ventilation during sedation given in spinal anaesthesia and induction of general anaesthesia. The authors also emphasise preparation of multiple alternative airway strategies beforehand in case initial plan fails. These considerations are especially relevant in ENT procedures such as adenotonsillectomy where supraglottic airway devices may not be feasible because of the shared surgical airway.

CONCLUSION(S)

Mucopolysaccharidosis type II presents major anaesthetic challenges because of progressive multisystem involvement, especially of the airway and cardiovascular system. Anticipation of a difficult airway, thorough preoperative assessment and a clear airway strategy are essential to reduce perioperative risk. This case highlights the importance of maintaining spontaneous ventilation during induction, ensuring availability of multiple airway devices and using alternatives such as bougie-assisted intubation when laryngoscopic view is poor. It also supports the safe use of regional anaesthesia with sedation for short abdominal procedures. Overall, a structured, multidisciplinary approach with close postoperative monitoring is key to optimising outcomes.

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