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Management of Difficult Airway in a Child with Morquio Syndrome: A Case Report

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Abstract: ***We are reporting a case of Morquio syndromic child with a challenging airway, a 14-years-old boy known case of morquio syndrome with a fixed cervical spine planned for extension of fusion and decompression of sub axial spine under IONM, successfully managed with fibreoptic bronchoscopy (FOB) guided by video laryngoscopy (VL).***

Keywords: Morquio syndrome, difficult airway management, fiberoptic bronchoscopy, video laryngoscopy, cervical spine fixation

1. Introduction

Morquio A, also known as Morquio-Brailsford syndrome, is a rare lysosomal storage disorder caused by deficiency of the enzyme N-acetyl-galactosamine-6-sulphatase (GALNS), an enzyme that catalyses the breakdown of two glycosaminoglycans (GAGs), keratan sulphate (KS) and chondroitin-6-sulphate(C6S)

Morquio syndrome is characterized by the progressive accumulation of KS and C6S in tissues. Therefore, Morquio syndrome presents most commonly as skeletal dysplasia which leads to short stature with bone deformities like dysostosis multiplex. Patients also suffer from odontoid hypoplasia and consequent atlantoaxial instability and neuromuscular fragility. Even minor extension of the neck can result in subluxation, spinal cord injury, and death.

2. Case Report

A 14-year-old male child of BMI 20.1 KG/M2 known case of mucopolysaccharidosis -morquio syndrome, undergone occipital condyle to C2 fixation in view of atlanto occipital dislocation and thrombolysis to vertebral artery following posterior circulation stroke before was admitted for extension of fusion with or without laminectomy of sub axial joint.

Anticipated Difficult Airway in View of Limited Extention –Flexion of Cervical Joint with Thoracic Kyphoscolisosis
During pre-anaesthesia assessment patient was evaluated as potential difficult airway and hence OT Preparedness was done accordingly. Difficult airway cart was kept ready including fibre optic bronchoscopy. Besides anaesthesia preparedness surgical team was informed about the anticipated difficult airway and also possibility of invasive airway access.

Preoperatively, Parents were counselled and risk of difficult airway and post op ventilation was explained

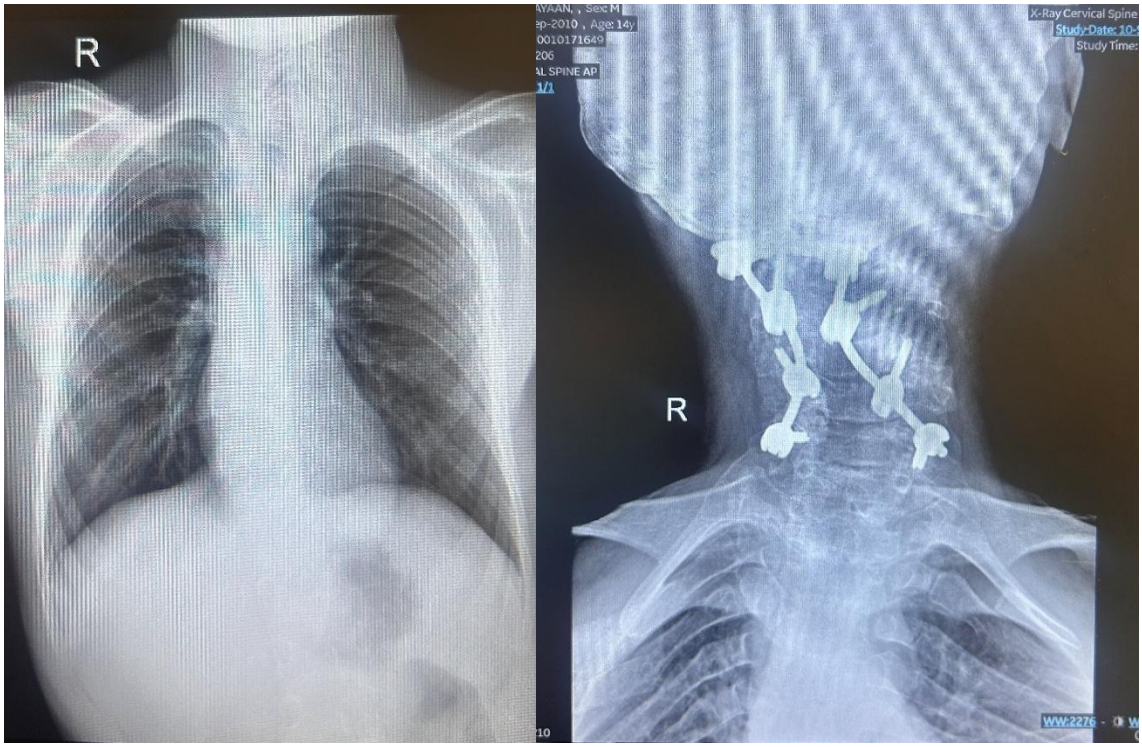
Intraopertively, following attachment of all standard ASA monitors, child was preoxygenated till ETO2 was achieved above 95. Careful positioning for intubation was given considering the limited mobility of cervical joint. IV line was flushed and checked. Induced with fentanyl 30mcg, midazolam 1mg, glycopyrrolate 0.1 mg, propofol 100mg and muscle relaxation achieved with atracurium 15mg.

First attempt of intubation was done with cmac blade 3 which showed Cormac lehane grading of 4 view and intubation was unsuccessful. As bag and mask ventilation is possible, child was ventilated again with FIO2 of 1 .0 again for another 2 minutes.

Second attempt was done with Fibre optic bronchospy guided orally with flexo metallic tube of size 6.5 mm passed and guided over FOB and fixed at 16 cm mark. Tube position confirmed with ETCO2 and B/l chest equal air entry on auscultation

GA was maintained by total intravenous anaesthesia (TIVA) in the form of propofol 150 mcg/kg /min marsh model. Neuromonitoring was done intraop and any reduction in SSEP signals were noted and taken care of.

After the end of surgery, Neostigmine 0.05mg/kg and glycopyrrolate 8mcg/kg was given intravenous before extubation. Child was gradually weaned off ventilator and extubated over bougie maintaining saturation 100% on room air, observed for 5mins on table with bougie insitu. No desaturation noted, bougie removed and patient kept on Hudson's mask. He was maintaining 100% saturation with Hudson's 6lo2/min. Shifted to PACU for further observation. After uneventful observation child was sent to ward ensuring vitals stable.



a) Scoliosis of the thoracolumbar spine is seen with its convexity to the left.
b) Status post laminectomy is noted from the C3 to C6 levels and surgical fusion of occipito cervical spine is noted [bilateral assimilation of the occipital condyles, C1 and C2].

3. Discussion

Morquio syndrome (Mucopolysaccharidosis type IV) arises from *GALNS* or *GLB1* enzyme defects, causing keratan sulphate accumulation and resulting in a variable skeletal phenotype characterized by short-trunk dwarfism, odontoid hypoplasia, and progressive kyphoscoliosis (1). Population-based reviews estimate that Morquio A is rare but globally distributed, with variable regional prevalence and numerous private *GALNS* mutations (2).

Multiple case series and reviews highlight significant anaesthetic challenges. Difficult airway management due to macroglossia, limited mouth opening, and restricted cervical mobility is well-documented (3,4). Severe cervical spine instability and atlantoaxial subluxation increase the risk of spinal cord injury during airway manipulation, while progressive tracheal and large-airway narrowing may not be apparent on clinical examination (5). These anatomical challenges contribute to a high incidence of difficult mask ventilation and intubation, often necessitating the use of awake or fiberoptic techniques (6).

Large multicentre trials and long-term follow-ups of enzyme replacement therapy (ERT) with elosulphase alfa demonstrate significant improvements in endurance (6-minute walk test), reduced urinary keratan sulphate levels, and some pulmonary benefit; however, reversal of established skeletal deformities remains minimal (7,8). Consequently, airway and cervical risks persist despite ERT, and perioperative precautions remain crucial.

Survey and registry-based analyses reveal that Morquio patients frequently undergo multiple surgical procedures-particularly orthopaedic and otolaryngological- with a high rate of perioperative complications linked to airway and

cervical instability (9,10). Published series consistently document frequent use of video-assisted or fibreoptic intubation and emphasize the need for multidisciplinary coordination (11). Expert guidelines recommend preoperative cervical spine imaging (flexion–extension radiographs or MRI), airway imaging when symptomatic tracheal narrowing is suspected, preparation for fibreoptic/video-laryngoscopy, readiness for surgical airway access, and the involvement of experienced anesthetic and ENT teams (12,13).

For anaesthetists, practical implications include comprehensive airway and cervical spine evaluation, avoidance of neck hyperextension, maintaining spontaneous ventilation during airway management, preparing smaller endotracheal tubes, and ensuring postoperative high-dependency monitoring for respiratory compromise (14,15). Registry data and systematic reviews reinforce the importance of individualized, multidisciplinary perioperative protocols to minimize morbidity and improve outcomes in this complex patient population (16).

Challenges of Airway

Oral Cavity
- Reduced mouth opening, abnormal dentition, large tongue,
- Increased copious secretions, mandibular abdnormalities

Hypoharyngx
- Narrow due to redundant tissue, enlarged adenoids and tonsils

Neck
- Cervical spine compression, atlanto axial joint instability, restricted mobility

Chest
- Kyphoscoliosis, tracheomalacia and tracheal narrowing,
- Airway edema, decreased pulmonary function, weakness of diaphragm

Challenges in this Case:
1) Fixed cervical spine due to the syndrome involved
2) Pre-existing Spine fixation complicating airway further
3) Thoracic kyphoscoliosis

Airway Management
Airway management in a child with Morquio syndrome (Mucopolysaccharidosis type IV) poses significant challenges due to the characteristic skeletal and soft tissue abnormalities associated with the disorder. These children often present with a short neck, limited neck extension, and atlanto-axial instability secondary to odontoid hypoplasia and cervical spine deformity. In addition, macroglossia, adenotonsillar hypertrophy, a short mandible, and tracheal narrowing contribute to a potentially difficult airway. The combination of these factors increases the risk of difficult mask ventilation, intubation, and possible cervical cord injury during airway manipulation.

Preoperative preparation must begin with a thorough multidisciplinary discussion involving the anaesthesiologist, otolaryngologist, paediatric intensivist, and neurosurgeon. Two anaesthesiologists is required to perform video laryngoscopy and to provide cricoid pressure for better view of vocal cords. A detailed review of the patient's previous anesthetic history, airway difficulties, and perioperative complications is invaluable. A comprehensive airway assessment should include imaging studies such as lateral cervical spine radiographs or MRI in flexion and extension to evaluate for atlanto-axial instability or spinal cord compression. Computed tomography (CT) may be used to assess the tracheal anatomy and identify areas of narrowing. An ENT evaluation is advisable during preoperative evaluation to assess for upper airway obstruction due to enlarged tonsils, adenoids, or macroglossia. Pulmonary function testing and sleep studies may be indicated if there are symptoms suggestive of obstructive sleep apnea.

Premedication should be administered with caution. Heavy sedation and opioid premedication are best avoided due to the risk of airway obstruction. But anxiolysis are generally unavoidable in paediatric population especially when Fibre optic bronchoscopy has to be performed. Small doses of midazolam may be used under close monitoring for anxiolysis. Antisialogogues such as glycopyrrolate can help reduce airway secretions, particularly when fibreoptic intubation is anticipated.

A difficult airway cart must be fully prepared before induction of anaesthesia. Essential equipment includes a fibreoptic bronchoscope (**the gold standard for airway management in these patients),** paediatric video laryngoscopes, a range of supraglottic airway devices, bougies, intubating stylets, and smaller-than-expected endotracheal tubes due to tracheal narrowing. A rigid bronchoscope and tracheostomy set should be immediately available, and an experienced ENT surgeon should be on standby for emergency surgical airway access. Cervical spine immobilization should be maintained with a collar or manual in-line stabilization throughout airway manipulation to prevent cord injury.

Fibre Optic Bronchoscopy
The preferred approach for securing the airway is awake or minimally sedated fibreoptic intubation while maintaining spontaneous ventilation. Topical anaesthesia with nebulized or sprayed lidocaine facilitates patient comfort and cooperation. Neck extension should be strictly avoided.

Flexible bronchoscopy is generally well tolerated. Appropriate measures to optimize the patient's condition should be taken to minimize risk. The absolute contraindications that impede performing bronchoscopy are severe refractory hypoxemia, hemodynamic instability, uncorrected haemorrhagic diathesis and the lack of authorization for the procedure by the parent or guardian. The relative contraindications depend on the experience of the team and the level of critical care in the hospital.

Sedation and Monitoring during FOB:
The aim of sedation in a child is to ensure that the patient is safe, comfortable and reasonably still during the procedure while maintaining adequate oxygenation and ventilation. Bronchoscopy can be performed in sedation with benzodiazepines or narcotics or under general anaesthesia.

An anaesthesiologist or intensivist must be present in addition to the person preforming bronchoscopy to administer drugs and monitor the patient continuously during the procedure. Spontaneous respiration is preferred during diagnostic procedures; hence level of sedation should be appropriate.

Flexible bronchoscopy is usually performed trans-nasally and can be performed orally or *via* artificial airway. Oxygen mask or nasal prongs can be used simultaneously while using the nasal route or the oral route. Oral route is used when nasal route is not feasible as in choanal atresia, nasal bleeding or trauma. (17)

Preferred drugs for local anaesthesia are 2% lidocaine jelly for the nose and 1% lidocaine spray for the pharynx and larynx. For analgesia and sedation, midazolam (0.05-0.2 mg/kg), fentanyl (1-3 mcg/kg) or ketamine (1-3 mg/kg) are used.

If fibreoptic intubation is not feasible, video laryngoscopy may be attempted cautiously with manual in-line stabilization, provided imaging has ruled out instability. Supraglottic devices, such as the laryngeal mask airway (LMA), can also serve as conduits for fibreoptic intubation. In cases where non-invasive techniques are predicted to fail, elective tracheostomy should be considered.

During the intraoperative period, it is essential to maintain a neutral neck position and avoid excessive movement. Muscle relaxants should not be administered until the airway is definitively secured. Gentle positive pressure ventilation is recommended to prevent distal airway collapse, and care should be taken to avoid high airway pressures. Continuous monitoring is crucial, especially if cervical instability is present.

Extubation should only be performed when the child is fully awake with intact protective airway reflexes and adequate spontaneous ventilation. Due to the high risk of postoperative airway obstruction, all patients should be closely monitored in a high-dependency or paediatric intensive care unit setting. In some cases, delayed extubation may be safer, particularly if airway edema or significant upper airway obstruction is anticipated.

In summary, airway preparation for a child with Morquio syndrome requires meticulous planning, multidisciplinary coordination, and readiness for difficult airway management. The use of fiberoptic bronchoscopy, maintenance of spontaneous ventilation, strict neck immobilization, and careful postoperative monitoring are key principles to ensure patient safety and successful outcomes.

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