Journal of Comprehensive Surgery

Case Report

DOI: 10.51271/JOCS-0080

Difficult airway management in a patient with mucopolysaccharidosis type IVA (Morquio A syndrome): a case report

Aleyna Calap Ocak*[1], Şemsi Mustafa Aksoy[1,2], Burak Nalbant[1,2]

[1]Department of Anesthesiology and Reanimation, Faculty of Medicine, Ankara Yıldırım Beyazıt University, Ankara, Turkiye
[2]Department of Anesthesiology and Reanimation, Ankara Bilkent City Hospital, Ankara, Turkiye

Cite this article as: Calap Ocak A, Aksoy ŞM, Nalbant B. Difficult airway management in a patient with mucopolysaccharidosis type IVA (Morquio A syndrome): a case report. *J Compr Surg.* 2025;4(3):74-76. doi:10.51271/JOCS-0080

Received: 15.07.2026 ◆ **Accepted**: 18.08.2026 ◆ **Published**: 25.08.2026

ABSTRACT

Mucopolysaccharidosis type IVA (Morquio A syndrome) is a rare lysosomal storage disorder characterized by glycosaminoglycan accumulation, causing progressive skeletal deformities and complex airway abnormalities that complicate anesthetic management. We report the perioperative airway management of a 27-year-old woman with MPS type IVA undergoing elective posterior cervical laminectomy for cervical myelopathy secondary to odontoid hypoplasia. Despite a preoperative Mallampati score of II, tracheal intubation was unexpectedly difficult. Attempts using both video laryngoscopy and a Macintosh laryngoscope failed because of a markedly enlarged, thickened epiglottis obstructing glottic visualization. Intubation was successfully achieved with a Miller laryngoscope. Anesthesia was maintained with total intravenous anesthesia, and the patient was extubated after complete recovery and a successful cuff leak test. Postoperative upper airway edema resolved with medical treatment. This case highlights that conventional airway assessment may underestimate airway difficulty in MPS type IVA and emphasizes the importance of alternative airway devices, meticulous perioperative planning, and vigilant postoperative monitoring.

Keywords: Miller laryngoscope, Morquio A syndrome, difficult airway

INTRODUCTION

Mucopolysaccharidosis type IVA (MPS IVA, Morquio A syndrome) is a rare autosomal recessive lysosomal storage disorder caused by deficiency of the enzyme N-acetylgalactosamine-6-sulfatase (GALNS), resulting in the accumulation of keratan sulfate and chondroitin-6-sulfate in various tissues. Progressive glycosaminoglycan deposition leads to characteristic skeletal abnormalities, short stature, and a short neck, as well as severe respiratory, cardiovascular, and neurological manifestations.[1]

Airway management represents one of the greatest anesthetic challenges in patients with Morquio A syndrome, and respiratory complications remain a major cause of morbidity and mortality in this population.[1] Glycosaminoglycan (GAG) deposition within the airway results in progressive airway narrowing, while macroglossia, adenotonsillar hypertrophy, and a short neck further increase the difficulty of tracheal intubation.[2] In addition, odontoid hypoplasia and atlantoaxial instability, which are frequently encountered in these patients, substantially increase the risk of spinal cord compression and sudden death during airway manipulation requiring cervical spine movement.[1]

Previous studies have demonstrated that the incidence of difficult tracheal intubation in patients with MPS increases with age and becomes particularly pronounced in children older than 36 months.[3] Conventional airway management techniques are often inadequate because of narrowed nasal passages, limited mandibular mobility, and restricted cervical extension.[3] Therefore, perioperative airway management should be carefully planned, with alternative airway devices such as videolaryngoscopes, flexible fiberoptic bronchoscopes, and emergency tracheostomy readily available, supported by an experienced multidisciplinary team.[2,3]

In this case report, we present the perioperative management of a patient with MPS type IVA, with particular emphasis on the challenges of difficult airway management in light of current evidence and contemporary airway management strategies.

CASE

A 27-year-old woman with mucopolysaccharidosis type IV-A, weighing 18 kg and measuring 98 cm in height, was scheduled for elective posterior cervical laminectomy because of cervical myelopathy secondary to C1 dens hypoplasia. She underwent a comprehensive preoperative anesthetic evaluation. Physical examination revealed coarse facial features, hypertelorism, macroglossia, and a short neck. Neurological examination demonstrated flaccid weakness of both the upper and lower extremities. The patient had been unable to use her lower extremities since 2022 and had lost the ability to use her hands during the preceding 3 weeks.

***Corresponding Author:** Aleyna Calap Ocak, aleynacalap@gmail.com

Cerebral angiography showed no evidence of intracranial vascular pathology, and no cardiac abnormalities were identified. Laboratory investigations, chest radiography, and electrocardiography were unremarkable. The patient was classified as American Society of Anesthesiologists (ASA) Physical Status III. Written informed consent for anesthesia and publication of this case report was obtained from both the patient and her parents.

Although the patient's Mallampati score was II, preparations for anticipated difficult airway management were made because unexpected difficult tracheal intubation may occur despite apparently reassuring bedside airway assessment, in accordance with the recommendations of the 2022 American Society of Anesthesiologists Difficult Airway Guidelines.[4] Upon arrival in the operating room, standard monitoring, including electrocardiography, pulse oximetry, and noninvasive blood pressure monitoring, was applied, while invasive monitoring was intentionally avoided. A 22-gauge intravenous catheter was inserted into the left antecubital vein.

General anesthesia was induced with propofol (40 mg), lidocaine (10 mg), and remifentanil (60 μg), without the administration of a neuromuscular blocking agent, in order to facilitate reliable intraoperative motor evoked potential (MEP) and somatosensory evoked potential (SSEP) monitoring. Initial airway assessment using a C-MAC video laryngoscope (Karl Storz, Germany) equipped with a difficult-airway blade revealed a markedly enlarged and thickened epiglottis; however, the vocal cords could not be visualized. As an adequate glottic view could not be obtained with the video laryngoscope, direct laryngoscopy was attempted using a No. 2 Macintosh blade, but elevation of the epiglottis was unsuccessful because it remained positioned over the glottic opening. A No. 2 Miller laryngoscope was subsequently used to directly lift the epiglottis, allowing clear visualization of the glottic opening and vocal cords (**Figure**). External laryngeal manipulation using the BURP maneuver and a stylet were used to facilitate tracheal intubation, whereas a bougie was not required. Tracheal intubation was then successfully achieved using a 5.5-mm cuffed reinforced endotracheal tube.

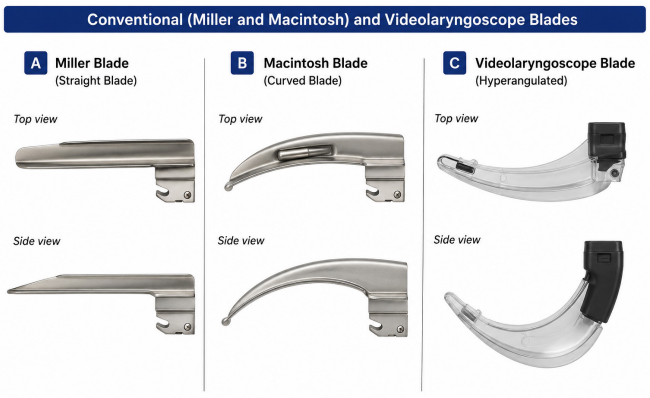

Figure. Conventional (Miller and Macintosh) and video laryngoscope blades[9]

Correct endotracheal tube placement was confirmed by bilateral chest auscultation and capnography. Anesthesia was maintained with total intravenous anesthesia (TIVA) using propofol and remifentanil. Intravenous fluids were administered at a rate of 10 ml/kg/h. At the completion of surgery, the patient was fully awake and was extubated after a successful cuff leak test.

The patient was transferred to the post-anesthesia care unit (PACU), where she was monitored for approximately 1 hour. During the early postoperative period, the patient developed upper airway mucosal edema accompanied by laryngospasm. Treatment consisted of intravenous theophylline, intravenous methylprednisolone (20 mg), and intranasal lidocaine combined with 1% diluted epinephrine. The airway symptoms resolved completely within 10 minutes without further complications, and the patient was subsequently transferred to the surgical ward after 1 hour of observation.

DISCUSSION

Mucopolysaccharidosis type IVA (Morquio A syndrome) is characterized by the accumulation of keratan sulfate within bone, cartilage, and connective tissues, leading to severe skeletal abnormalities that directly influence anesthetic management, including short stature, a short neck, odontoid hypoplasia, and consequent atlantoaxial instability.[5]

Previous studies have consistently demonstrated that airway management in patients with mucopolysaccharidoses is associated with a high incidence of difficult mask ventilation and tracheal intubation because of progressive glycosaminoglycan deposition, macroglossia, thickened supraglottic tissues, and cervical spine abnormalities.[1,3] As airway involvement progresses with age, airway management becomes increasingly challenging, emphasizing the importance of meticulous preoperative assessment, careful planning, and the availability of alternative airway devices.[3,6] Similar airway-related challenges have been reported in both pediatric and adult patients with Morquio A syndrome.[5,6]

These anatomical and pathological changes translate into a high incidence of difficult airway management in clinical practice. Previous studies have reported an incidence of difficult tracheal intubation ranging from 28% to 44% in patients with MPS.[7] As observed in our patient, progressive GAG accumulation with advancing age further increases the complexity of airway management. This observation is consistent with the 2022 ASA Difficult Airway Guidelines, which emphasize that no single bedside airway assessment reliably predicts difficult tracheal intubation and recommend comprehensive airway evaluation together with preparation for alternative airway strategies.[4]

Identification of cerebrovascular abnormalities reported in patients with MPS during the preoperative evaluation may contribute to safer perioperative management of cerebral perfusion. Although cerebral angiography is not routinely recommended, it may provide valuable information for anesthetic planning in selected patients with neurological manifestations or suspected cerebrovascular pathology.[5]

GAG deposition within the airway results in macroglossia, pharyngeal narrowing, laryngeal involvement, and thickening of the epiglottis.[5] In our case, both video laryngoscopy and direct laryngoscopy using a No. 2 Macintosh blade failed because the enlarged epiglottis could not be adequately displaced to expose the glottic opening. Although video laryngoscopy is currently recommended as a first-line technique for airway management in patients with MPS,[1,3] it may be insufficient in cases of severe soft-tissue hypertrophy, limited mouth opening, or marked epiglottic enlargement. Successful tracheal intubation was ultimately achieved using a Miller laryngoscope, suggesting that its ability to directly

elevate the epiglottis may provide a distinct advantage in selected patients with difficult airway anatomy, consistent with previous reports describing improved glottic visualization with the Miller blade in patients with a large or elongated epiglottis.[8]

Because our patient underwent surgery for cervical myelopathy secondary to odontoid hypoplasia, excessive cervical movement during airway management had to be strictly avoided. Patients with MPS IVA are at increased risk of cervical spinal cord compression and atlantoaxial instability; consequently, even minimal neck manipulation may result in catastrophic spinal cord injury or sudden death. Therefore, maintaining the cervical spine in a neutral position during tracheal intubation and considering alternative techniques, such as flexible fiberoptic bronchoscopy when appropriate, are essential components of safe airway management.

Postoperative laryngospasm and upper airway edema are well-recognized airway complications in patients with MPS. Structural airway fragility, mucosal thickening secondary to GAG deposition, and a narrowed airway lumen predispose these patients to upper airway obstruction, even following minimal airway irritation after extubation. In our patient, postoperative upper airway edema was successfully managed with prompt medical treatment, without the need for reintubation. This case highlights that safe airway management in patients with MPS extends beyond successful tracheal intubation. Extubation should be performed only after complete recovery of consciousness and airway reflexes, followed by close postoperative monitoring in an environment where airway complications can be rapidly recognized and treated.

CONCLUSION

- Laryngoscopy should be performed as gently as possible to avoid airway trauma.
- All types of laryngoscopes should be readily available to the anesthesiologist, considering the possibility that visualization of the glottic opening may be hindered by GAG accumulation.
- Clinicians should be prepared for airway edema that may develop during extubation and in the postoperative period.

Successful anesthetic management of patients with MPS type IVA requires meticulous preoperative planning, an individualized airway management strategy, and close postoperative monitoring following extubation.

ETHICAL DECLARATIONS

Informed Consent

Written informed consent was obtained from the patient included in this report. Signed consent forms are retained by the authors and are available upon request.

Peer Review Process

This report underwent external peer review.

Conflict of Interest

The authors declare no conflicts of interest.

Financial Disclosure

This case report did not receive any financial support.

Author Contributions

Concept: ACO, ŞMA, BN; Design: ACO, ŞMA, BN; Control: ACO, ŞMA, BN; Resources: ACO, ŞMA, BN; Materials: ACO, ŞMA, BN; Data Collection and/or Processing: ACO, ŞMA, BN; Analysis and/or Interpretation: ACO, ŞMA, BN; Literature Review: ACO, ŞMA, BN; Writing the Article: ACO, ŞMA, BN; Critical Review: ACO, ŞMA, BN.

REFERENCES

1. Hendriksz CJ, Berger KI, Giugliani R, et al. International guidelines for the management and treatment of Morquio A syndrome. *Am J Med Genet A*. 2015;167A(1):11-25. doi:10.1002/ajmg.a.36833
2. Hanalıoglu D, Yeke B, Bırbılen AZ, Tekşam Ö, Sivri S. Bir mukopolisakkaridoz tip II olgusunda zor hava yolu yönetimi. *Çocuk Acil Yoğ Bak Derg*. 2019;6(2):113-116. doi:10.4274/cayd.galenos.2018.22931
3. Tümer M, Yılbaş AA, Debbağ S, Sarıcaoğlu F, Canbay Ö. Airway management in mucopolysaccharidosis: a retrospective case series review. *Braz J Anesthesiol*. 2023;73(5):680-682. doi:10.1016/j.bjane.2021.10.013
4. Apfelbaum JL, Hagberg CA, Connis RT, et al. 2022 American Society of Anesthesiologists Practice Guidelines for management of the difficult airway. *Anesthesiology*. 2022;136(1):31-81. doi:10.1097/ALN.0000000000004002
5. Theroux MC, DiCindio S, Averill LW, et al. Tracheal narrowing and its impact on anesthesia care in patients with Morquio A (mucopolysaccharidosis type IVA): an observational study. *Anesth Analg*. 2023;137(5):1075-1083. doi:10.1213/ANE.0000000000006513
6. Walker RWM, Darowski M, Morris P, Wraith JE. Anaesthesia and airway management in mucopolysaccharidosis. *J Inherit Metab Dis*. 1994;17:509-517.
7. Garcia YKG, Reyes CRB. Anesthetic and airway management in a pediatric patient with Morquio syndrome: a case report. *Acta Med Philipp*. 2024;58(9):35-38. doi:10.47895/amp.v58i9.8284
8. Anderson P, Espinaco Valdés J, Vorster JG. Successful difficult airway intubation using the Miller laryngoscope blade and paraglossal technique-a comparison with the Macintosh blade and midline technique. *South Afr J Anaesth Analg*. 2015;21(2):28-30. doi:10.1080/22201181.2015.10