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CASE REPORT

Anesthetic Management of a Two-Year-Old Child with Mucolipidosis using the I-gel® for Bronchoscopy: A Case Report

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ABSTRACT

The lysosomal storage disease mucolipidosis presents formidable challenges for the pediatric anesthetist, not only due to progressive multi-system involvement but also because of intricate airway abnormalities. A two-year-old boy with mucolipidosis type II (GNPTAB mutation) presented with history of T.B under treatment and posted for flexible bronchoscopy with bronchoalveolar lavage (BAL). This report demonstrates the complexity of perioperative airway management in such children as guided by modern difficult airway algorithms, and the pivotal role of the I-gel® supraglottic airway device as both primary and rescue tool.

KEYWORDS

• Bronchoscopy • Mucolipidosis • Igel • Anaesthesia

INTRODUCTION

Mucolipidoses are a rare group of autosomal recessive lysosomal storage disorders clinically and genetically distinct from mucopolysaccharidoses caused by defective lysosomal enzyme targeting and trafficking, with four recognized types.[1,2] Type II (I-cell disease) and type III (pseudo-Hurler polydystrophy) are most frequently due to biallelic pathogenic variants in the GNPTAB gene, which encodes N-acetylglucosamine-1-phosphotransferase, a critical enzyme for mannose-6-phosphate tagging of lysosomal hydrolases.[1,3,4]

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This molecular defect impairs lysosomal enzyme delivery and leads to extracellular secretion of hydrolases, chronic substrate accumulation within the lysosomes, and profound multi-organ involvement.[2,5,6] Affected children characteristically manifest coarse facial features, macroglossia, gingival hypertrophy, high-arched palate, skeletal dysostosis, and hepatosplenomegaly. Airway compromise is progressive and compounded by tissue thickening, macroglossia, pharyngeal infiltration, thoracic cage deformity, and limited cervical mobility, producing perioperative respiratory vulnerability that closely mimics but is molecularly distinct from mucopolysaccharidoses.[3,5,7,8]

Recent consensus guidelines from leading societies including the Difficult Airway Society (DAS, UK, 2025) and the updated American Society of Anesthesiologists (ASA, 2022)—emphasize thorough risk assessment, multidisciplinary planning, preservation of spontaneous ventilation, judicious use of supraglottic airway devices, and early escalation to advanced techniques, especially in the pediatric population.[9-12] Knowledge of disease-specific airway features and adherence to modern algorithms are essential for safe management in mucolipidosis.

Case Presentation

We present a two-year-old boy (weight 5 kg) with genetically confirmed mucolipidosis II (GNPTAB mutation), a history of failure to thrive, and recurrent lower respiratory tract infections. On examination, he exhibited coarse facies, depressed nasal bridge, macroglossia, high-arched palate, pectus carinatum, hepatosplenomegaly, and notably limited neck extension. Respiratory assessment revealed crepitations; his SpO_2 was 96% on room air. CT imaging demonstrated right upper lobe atelectasis, history of TB and under treatment.

Preoperative planning and we assembled a comprehensive difficult airway cart with multiple I-gel® sizes, video laryngoscope, pediatric fiber optic scope, and emergency tracheostomy kit. After careful IV induction (propofol 2 mg/kg, fentanyl 2 µg/kg), standard ASA monitoring and glycopyrrolate (0.004 mg/kg) were administered. Sevoflurane was used to preserve spontaneous breathing, deliberately avoiding muscle relaxants until airway security was established.

A size 1.5 I-gel® supraglottic airway was placed, verified by chest rise and capnography. Bronchoscopy and BAL were successfully performed via the I-gel® conduit. During early recovery, acute airway obstruction developed with rising end-tidal CO_2. Two direct laryngoscopy attempts failed due to distorted anatomy and ventilation was maintain with mask and I gel in between attempt. As Per guidelines, further attempts were stopped and escalation ensued: the I-gel® was reinserted, restoring airway patency, and fiberoptic-guided intubation with a 4.0 mm tube was placed and Bilateral ait entry and Capnography confirmed and I gel was removed carefully with placing the tube in situ. Elective ventilation in the PICU overnight preceded uneventful extubation.

Figure 1: Typical Physical Features of Mucolipidosis of Child

Figure 2: Bronchoscopy with I gel in situ

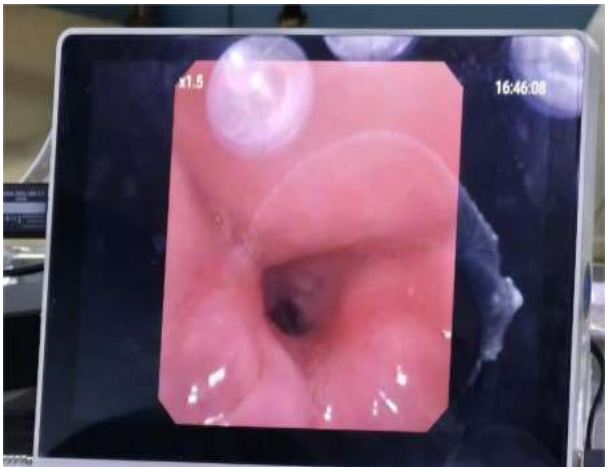

Figure 3:

DISCUSSION

Children with mucolipidosis remain at the highest risk for perioperative airway events, and often present with a constellation of craniofacial, pharyngeal, laryngeal, and cervical anomalies.[3,5,8,17] Airway narrowing due to chronic substrate accumulation, thickened tongue, pharyngeal walls, limited neck movement, and a crowded oral cavity pose well-recognized challenges to both mask ventilation and tracheal intubation.[4,5,7,18] These risks are not only anatomical but also compounded by recurrent respiratory tract infections, inadequate cough, and excessive secretions.

Our perioperative strategy was shaped by recent airway guidelines that prioritize patient safety through (a) prediction and preparation, (b) maintenance of spontaneous ventilation, (c) selection of advanced airway devices, and (d) limitation of repeated intubation attempts.[9-12] The I-gel® is particularly suited to pediatric cases, offering superior sealing properties and the unique ability to serve as both a primary airway and a reliable fiberoptic conduit.[6,16,18] Unlike first-generation LMAs, the i-gel® offers minimal tissue pressure and gastric drainage, decreasing aspiration risk and aiding in the management of children with airway edema and secretions.[18,19]

When direct laryngoscopy failed, escalation to fiberoptic intubation via SGA is advocated by both DAS and ASA algorithms in children.[9,10,12,16] This approach converted a potentially catastrophic situation into a controlled rescue, allowing oxygenation and bronchoscopic guidance, and avoiding trauma from repeated instrumentation.[11,14,16] Post-operative elective ventilation is justified in children at risk for airway edema, and a staged approach to extubation should be routine in mucolipidosis.[8,19] Collaboration among anesthesia, pediatric, and surgical teams is vital to effective airway rescue.[13-15]

Alternative strategies such as awake fiberoptic intubation, video laryngoscopy, and nasal intubation may be considered but have specific limitations in this population, including intolerance, poor views due to macroglossia, and increased risk of trauma. The i-gel® platform, with its second-generation features, continues to gain support as the airway management device of choice in pediatric patients with syndromic difficult airways.[6,16,19,20]

CONCLUSION

Management of the difficult pediatric airway in mucolipidosis demands sophisticated planning, multidisciplinary coordination, and adherence to current international guidelines. The I-gel® has demonstrated its value as both a primary airway device and a rescue conduit for fiberoptic intubation, making it fundamental in the management strategy for syndromic airway disease. Future research into airway algorithms and device innovation will further refine care standards for these complex children.

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