

Elective Postoperative Ventilation in a Child with Goldenhar Syndrome Undergoing Adenotonsillectomy: A Case Report and Review of Type II Negative Pressure Pulmonary Edema

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ABSTRACT

Background: Goldenhar syndrome is associated with craniofacial abnormalities that may complicate airway management and contribute to obstructive sleep apnoea (OSA). Children with severe OSA undergoing adenotonsillectomy are at risk of postoperative respiratory complications, including type II negative pressure pulmonary edema (NPPE, a rare condition that occurs following relief of chronic upper airway obstruction¹⁻⁴).

Keywords: Goldenhar syndrome; Obstructive sleep apnoea; Negative pressure pulmonary edema

Introduction

Goldenhar syndrome (ouculo- auriculo -vertebral spectrum) is a congenital disorder characterized by cranio facial anomalies including micrognathia, ear anomalies (e.g skin tags, prominent ear, hearing loss), vertebral defects and occasionally cardiac or renal malformations. Airway management in these patients is challenging due to mandibular hypoplasia, short neck and OSA¹.

Negative pressure pulmonary edema also known as post obstructive pulmonary edema is a rare but potentially life-threatening complication following upper airway surgery, particularly adenotonsillectomy in children with OSA, it is classified in two types:

- **Type I NPPE:** occurs with acute upper airway obstruction (eg. Post extubation laryngospasm) where forceful inspiratory efforts against closed glottis generates a highly negative intrathoracic pressure.
- **Type II NPPE:** develops after surgical relief of a chronic airway obstruction (eg. Hypertrophied tonsils, adenoid in long standing OSA). chronic obstruction leads to modest intrinsic peep, maintaining elevated end-expiratory lung volume, sudden release causes rapid drop in intrathoracic pressure, altering hydro static forces and capillary permeability, resulting in pulmonary edema²⁻⁴.

Children with syndromic craniofacial anomalies and severe OSA are at elevated risk. Literature reports Type II NPPE after

ADT. We describe a case of proactive ventilation successfully prevented this complication⁵⁻¹⁰.

Case Presentation

We report the case of a 3 years old weighing 10 kg girl with genetically confirmed Goldenhar syndrome was scheduled for Drug induced sleep endoscopy because of severe OSA requiring nocturnal continuous positive airway pressure (CPAP). She had previously undergone adenoidectomy and bilateral myringotomy but continued to experience significant sleep disordered breathing. Physical examination demonstrated micrognathia, a short neck, vertebral anomalies and bilateral hearing impairment suggesting potentially difficult airway.

She underwent drug induced sleep endoscopy (DISE) followed by adenotonsillectomy (ADT) after severe tonsillar obstruction was identified. Airway management was challenging because of micrognathia and marked tonsillar hypertrophy, necessitating Glidoscope-assisted intubation with bougi guidance.

Anesthetic Management

Anesthesia was conducted using an opioid sparing total intravenous anesthesia (TIVA) because severe OSA increases the susceptibility to opioid- induced respiratory depression.

DISE was initially performed under intravenous dexmedetomidine 4 mcg and titrated propofol infusion 60 mg/hr providing adequate spontaneous ventilation and satisfactory procedural conditions. DISE demonstrated severe upper airway obstruction primarily caused by markedly enlarged tonsils and the decision was made to proceed with ADT during the same anesthetic session. Anesthesia was continued using the opioid sparing TIVA technique, additional medications included glycopyrolate 10 mcg, low dose ketamine (10 mg) dexmedetomidine 4 mcg, dexamethasone 2 mg and cisatracurium to facilitate endotracheal intubation. Airway management was challenging because of micrognathia and marked tonsillar hypertrophy and intubation was successfully achieved using a glidoscope video-laryngoscope using bougi assistance. The surgical procedure lasted approximately two hours, during which the patient remained hemodynamically stable without significant intraoperative complications.

Given the combination of severe OSA, craniofacial abnormalities, anticipated airway edema and the reported risk of type II NPPE after ADT, the patient remained electively intubated and mechanically ventilated in the pediatric intensive care unit for 24 hours. Extubation on postoperative day 1 was uneventful and no respiratory complications occurred.

Discussion

To our knowledge, this is among the first reported cases describing the perioperative management of a child with genetically confirmed Goldenhar syndrome with severe OSA requiring home CPAP, DISE guided ADT, anticipated difficult airway and elective postoperative ventilation undertaken because of the anticipated risk of post operative of post operative respiratory complications, including type IINPPE although no casual relationship can be inferred from a single case, this report highlights a multi-disciplinary, individualized perioperative strategy that resulted in uncomplicated postoperative course¹⁻⁵.

Moser, et al. reported a pediatric patient with severe OSA who developed type II NPPE after immediate extubation following ADT, ultimately requiring invasive ventilation and HFOV. In contrast, our patient was electively ventilated for 24 hours and experienced an uncomplicated recovery without evidence of NPPE⁵.

And similar to Poland-Moebius syndrome case reported by Van Kooy and Gargiulo, our patient exhibited craniofacial abnormalities associated with airway vulnerability. These reports collectively suggest that syndromic children with chronic upper airway obstruction may represent a subgroup requiring heightened postoperative surveillance⁷.

Li and Zhou described a 21 months old child who developed type II NPPE following removal of a chronically obstructing inhaled foreign body. Despite the different etiology, the mechanism was similar with NPPE occurring after sudden relief of long-standing airway obstruction, so it's not limited to ADT but rather represents a physiological consequence of abrupt changes in airway resistance and intrathoracic pressure relationships⁶.

Befikadu also described NPPE post ADT in previously healthy 12 years old child who was eventually transferred to ICU for stabilization⁸.

The concern of type II NPPE was supported by several previously published reports. One of the most relevant is the case reported by Moser and colleagues, similar to our patient this child had severe chronic upper airway obstruction and underwent ADT as a definitive treatment, however our patient remained electively intubated and mechanically ventilated for 24 hours post operative in PICU⁵.

The decision of opioid sparing anesthesia and to maintain postoperative mechanical ventilation was based on a combination of factors rather than concern of single complication. Difficult airway raised concerns regarding emergency reintubation should respiratory failure occur after extubation. Severe OSA increased risk of post-operative respiratory compromise, while published reports of type II NPPE following ADT highlighted the potential consequences of abrupt relief of obstruction. Accordingly, a multidisciplinary discussion among anesthesiology, otolaryngology and pediatric intensive care teams resulted in the decision to continue controlled ventilation overnight. Extubation was subsequently performed under controlled conditions in PICU after ensuring adequate recovery and stability²⁻⁹.

Limitations

The principal limitation of this report is that it represents a single case and cannot establish that elective postoperative ventilation prevents type II NPPE. The favorable outcome may have been influenced by multiple factors, including meticulous airway planning, opioid sparing anesthesia and multidisciplinary postoperative care. Nevertheless, this case demonstrates a rational perioperative strategy that may be considered in carefully selected high-risk patients.

Conclusion

This case illustrates the complex interaction between syndromic craniofacial abnormalities, severe OSA, difficult airway management and postoperative respiratory risk.

Comparison with previously published reports, particularly the case described by Moser et al., underscores the potential severity of Type II NPPE after relief of chronic upper airway obstruction. Careful preoperative assessment, preparation for a difficult airway, multidisciplinary decision-making and individualized postoperative ventilatory support contributed to a favorable outcome in our patient and may represent valuable considerations for similar high-risk pediatric populations.

Although it cannot be concluded from a single case that postoperative ventilation prevents type II NPPE, our experience demonstrates that individualized delayed extubation may be considered in carefully selected high-risk patients¹⁻⁵.

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