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Original Case Report

Anaesthetic Challenges and Strategies in a High-Risk Infant with Hydrocephalus, Corpus Callosum Dysgenesis, Severe Gerd, and Airway Anomalies: A Case Report

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Abstract: Managing anaesthesia in infants with multiple congenital anomalies is highly challenging. We report the perioperative management of a 7-month-old male (3.87 kg) with aqueductal stenosis, corpus callosum dysgenesis, hydrocephalus, severe GERD, retrognathia, and recurrent pneumonia, who underwent laparoscopic fundoplication. The main anaesthetic concerns were airway difficulty, aspiration risk, and brain protection. Careful planning, advanced airway strategies, and vigilant multidisciplinary care resulted in a safe outcome, highlighting the importance of individualized approaches in high-risk paediatric anaesthesia.
Case Presentation: A 7-month-old, 3.87 kg patient with severe GERD, hydrocephalus, corpus callosum dysgenesis, retrognathia, and recurrent pneumonia was scheduled for laparoscopic fundoplication. He had developmental delay, hypotonia, and a Mallampati III airway. Preoperative preparation included strict fasting, continuation of proton-pump inhibitors, and IV ranitidine. Airway equipment, including video laryngoscope and fibreoptic bronchoscope, was prepared. Inhalational induction with 6% sevoflurane preserved spontaneous breathing. Video laryngoscopy enabled successful intubation with a 3.0 mm ETtube. Anaesthesia was maintained with sevoflurane in air/oxygen, pressure-controlled ventilation, and PEEP; nitrous oxide was avoided. Analgesia included IV paracetamol (15mg/kg) and fentanyl (2mcg/kg), with atracurium (0.1-0.5mg/kg) for muscle relaxation. Intra-abdominal pressure was kept below 10 mmHg. The patient was extubated awake in a semi-upright position after thorough suctioning, with dexmedetomidine for smooth emergence. Postoperatively, he was monitored in ICU, received chest physiotherapy, and recovered without complications.This case demonstrates that meticulous planning, skilled airway management, and multidisciplinary teamwork are vital for safe anaesthesia in infants with multiple congenital anomalies.

Keywords: Paediatric anaesthesia, Hydrocephalus, GERD, Airway difficulty, Laparoscopic fundoplication, Retrognathia

INTRODUCTION

Infants with multiple congenital anomalies present significant anaesthetic challenges due to the combined involvement of neurological, gastrointestinal, and airway systems. The physiological immaturity of vital organ systems in early infancy, coupled with anatomical variations and coexisting comorbidities, increases the risk of perioperative morbidity and mortality (1). Anaesthesia providers must therefore anticipate and prepare for a range of complications including difficult ventilation, impaired airway access, delayed recovery, aspiration, altered drug responses, hemodynamic instability, and inadequate respiratory reserve (2).

Hydrocephalus, a common congenital neurological disorder, results from an imbalance between cerebrospinal fluid (CSF) production and absorption, causing ventricular enlargement and increased intracranial pressure (ICP) (3). The impaired cortical development and disrupted cerebrovascular autoregulation associated with hydrocephalus make infants particularly vulnerable to fluctuations in CO_2, hypoxia, hypercarbia, and elevated intra-abdominal pressure during laparoscopy (4). Additionally, corpus callosum dysgenesis, a structural malformation involving partial or complete absence of the corpus callosum, may contribute to developmental delay, muscle hypotonia, impaired airway reflexes, and abnormal physiological responses under anaesthesia (5).

Severe gastroesophageal reflux disease (GERD) in infants is often associated with recurrent aspiration, respiratory infections, and growth retardation. During induction of anaesthesia, the loss of airway reflexes markedly increases the risk of gastric content regurgitation and aspiration pneumonitis (6). In this patient population, early and reliable airway control becomes a primary objective, yet this is complicated further by craniofacial anomalies. Retrognathia, characterised by a posteriorly displaced mandible, is a well-known predictor of difficult mask ventilation, laryngoscopy, and endotracheal intubation. Infants also have large tongues, high anterior larynxes, and compliant airways, all of which make securing the airway inherently challenging even in otherwise normal subjects (7).

Laparoscopic fundoplication is a preferred surgical intervention for severe GERD and recurrent aspiration in infants; however, carbon dioxide insufflation and pneumoperitoneum introduce additional anaesthetic concerns (8). Increased intra-abdominal pressure can impair venous return, elevate diaphragm position, reduce lung compliance, and exacerbate ventilation-perfusion mismatch. In the presence of hydrocephalus, these physiological alterations may trigger acute rises in intracranial pressure if not carefully managed. Avoiding hypercarbia, hypoxia, excessive airway pressures, and nitrous oxide become essential to maintain stable cerebral dynamics (8).

The combination of respiratory vulnerability due to recurrent pneumonia, impaired respiratory muscle tone from neurological disease, and difficult airway anatomy demands meticulous perioperative planning. Strategies must involve minimising aspiration risk during induction, ensuring the availability of advanced airway tools such as video laryngoscopes and fibreoptic bronchoscopes, closely regulating ventilatory parameters, and adopting a cautious approach to extubation and postoperative respiratory monitoring (9).

This case report describes the detailed anaesthetic management of a high-risk infant with aqueductal stenosis-induced hydrocephalus, corpus callosum dysgenesis, severe GERD, retrognathia, and a history of recurrent aspiration pneumonia undergoing laparoscopic fundoplication. It highlights the importance of a personalised strategy integrating airway security, neuroprotection, and respiratory optimisation. Through vigilant planning, multidisciplinary collaboration, and utilization of modern airway and monitoring technologies, a safe perioperative outcome was achieved in a patient with extremely complex needs. This case reinforces that paediatric anaesthesia for multisystem congenital disorders requires careful anticipation, readiness for complications, and a comprehensive approach tailored to each patient's unique anatomical and physiological challenges.

CASE REPORT

Patient Presentation:

A 7-month-old male infant, weighing 3.87 kg, was referred for surgical correction of severe gastroesophageal reflux disease (GERD) following persistent feeding difficulties, recurrent choking episodes, and multiple hospitalizations for aspiration pneumonia. He was born at term after an uncomplicated pregnancy and delivery; however, from early infancy he exhibited recurrent non-bilious vomiting and inadequate weight gain despite medical management. Developmental delays were apparent, with poor head control, hypotonia, and lag in achieving motor milestones. The infant was the first child of healthy, non-consanguineous parents, and family history for congenital anomalies was unremarkable.

Neuroimaging previously performed due to

macrocephaly demonstrated aqueductal stenosis causing obstructive hydrocephalus, along with corpus callosum dysgenesis. Ventricular enlargement was persistent, although he did not require shunt placement at this stage. Severe reflux symptoms continued despite upright feeding, thickened feeds, and ongoing treatment with proton-pump inhibitors; hence, laparoscopic fundoplication was planned to improve feeding tolerance and reduce pulmonary complications.

Preoperative Assessment:

During pre-anaesthetic evaluation, the infant appeared malnourished and irritable. He exhibited tachypnoea with mild intercostal retractions suggestive of compromised pulmonary reserve from repeated aspiration events. Auscultation revealed bilateral coarse crepitations but no active wheeze. Airway assessment identified significant retrognathia, micrognathia, a large tongue, and reduced mandibular space, predicting difficult laryngoscopy. Mallampati grading, although limited in infants, approximated Grade III.

Neurological assessment confirmed global developmental delay and hypotonia, increasing the risk of airway collapsibility under anaesthesia. Hydrocephalus raised concerns about maintenance of optimal cerebral perfusion and avoidance of intracranial pressure (ICP) surges.

Laboratory investigations including complete blood count, electrolytes, and coagulation profile were within acceptable ranges. Chest X-ray showed residual bilateral patchy infiltrates from previous pneumonia. The patient was deemed high-risk for aspiration, airway difficulty, and postoperative respiratory complications; a tailored anaesthetic plan was formulated accordingly.

Figure 1: Anaesthetic induction in a high-risk infant with hydrocephalus, severe GERD, and retrognathia. The image demonstrates gentle mask ventilation using a paediatric face mask and self-inflating bag while maintaining spontaneous respiration, along with standard intraoperative monitoring and readiness for difficult airway management.

Intraoperative Anaesthetic Management:

After strict preoperative fasting, pharmacologic aspiration prophylaxis was administered using proton-pump inhibitors and intravenous ranitidine. Standard monitoring was applied in the operating theatre. A comprehensive difficult-airway cart was kept ready including a video laryngoscope, paediatric fibreoptic bronchoscope, different-sized endotracheal tubes, and emergency tracheostomy equipment.

Anaesthetic induction was initiated with sevoflurane (6%) in 100% oxygen, carefully maintaining spontaneous respiration to reduce the risk of airway obstruction and regurgitation. Gentle jaw thrust and minimal mask pressure were used to prevent gastric insufflation. Once optimal depth was reached, video laryngoscopy was performed and provided an adequate view of the glottis despite anatomical challenges. Endotracheal intubation using a 3.0 mm cuffless tube was achieved smoothly without oxygen desaturation or regurgitation.

Maintenance of anaesthesia was achieved using sevoflurane in a mixture of air and oxygen. Nitrous oxide was deliberately avoided to prevent bowel distension and ICP elevation. Pressure-controlled ventilation with low peak airway pressures and

appropriate PEEP was employed to optimize ventilation while minimizing barotrauma. $ETCO_2$ was maintained between 35–40 mmHg to avoid cerebral vasodilation.

Intra-abdominal pressure during laparoscopy was strictly limited to <10 mmHg to mitigate risks of reduced venous return, decreased lung compliance, and increased ICP. Analgesia was provided using intravenous paracetamol (15 mg/kg) and fentanyl (2 mcg/kg). Atracurium supplementation facilitated surgical exposure and minimized diaphragmatic movement.

Throughout the procedure, haemodynamics remained stable and oxygen saturation was consistently above 95%. No episodes of bronchospasm or regurgitation occurred.

Figure 2: Use of video laryngoscopy and standard monitoring during anaesthetic management of a high-risk infant with anticipated difficult airway. The image highlights real-time airway visualization, continuous ECG and oxygen saturation monitoring, and preparedness for advanced airway management.

Figure 3: Video laryngoscopy

Postoperative Care and Outcome:
At surgical completion, residual secretions were thoroughly suctioned before extubation. Extubation was performed in a fully awake state with the infant positioned semi-upright to further reduce aspiration risk. Dexmedetomidine was used to ensure smooth recovery, minimizing coughing and agitation.

The infant was immediately transferred to the paediatric intensive care unit for close respiratory and neurological monitoring. Humidified oxygen therapy, chest physiotherapy, and careful head positioning were continued postoperatively. Analgesia was maintained with paracetamol.

The patient remained clinically stable with no signs of respiratory distress, aspiration, or raised ICP. Feeding was re-initiated gradually under anti-reflux positioning. He was discharged home in a stable condition after an appropriate recovery period with planned follow-up for nutritional rehabilitation and neurological evaluation.

DISCUSSION

Infants with multiple congenital anomalies, particularly those involving craniofacial, neurological, and gastrointestinal systems, pose significant risks during the perioperative period (10). In the present case, the patient had hydrocephalus with corpus callosum dysgenesis, severe GERD leading to recurrent aspiration pneumonia, and retrognathia resulting in a potentially difficult airway. Each of these conditions independently increases anaesthetic complexity, and their combination necessitated highly strategic management throughout the perioperative period.

The foremost challenge was securing the airway while

minimizing aspiration risk. Severe GERD compromises laryngeal protective reflexes, increasing gastric fluid entry into the airway at any point during induction or extubation. Moreover, retrognathia and a diminutive mandibular space can hinder visualization of the glottis, making direct laryngoscopy difficult (11). The choice of inhalational induction with preserved spontaneous ventilation was appropriate, as this avoided airway collapse and prevented positive-pressure mask ventilation from forcing gastric contents upward (12). Video laryngoscopy offered superior visualization in the anatomically challenged airway and decreased the likelihood of multiple intubation attempts, which are known to increase aspiration risk. Avoidance of nitrous oxide was a prudent choice since it can expand bowel gas volume and indirectly increase the risk of regurgitation and intracranial pressure (13).

The patient's hydrocephalus required diligent neuroprotective strategies. Laparoscopic procedures inherently risk hypercarbia from carbon dioxide absorption and elevated intra-abdominal pressure, both of which may critically increase intracranial pressure (14). Pressure-controlled ventilation, maintenance of normocapnia, and keeping pneumoperitoneum pressure below 10 mmHg effectively mitigated these risks. In addition, opioids were carefully titrated to provide analgesia while preventing exaggerated respiratory depression, which could impair cerebral venous drainage and augment intracranial pressure (15).

Pulmonary optimization was another essential consideration due to the infant's history of recurrent pneumonias. Reduced functional residual capacity, airway hyper-reactivity, and secretion pooling in infants heighten the probability of perioperative hypoxemia (16). Gentle ventilation and postoperative physiotherapy helped maintain airway clearance while avoiding barotrauma and atelectasis. Extubation in a fully awake state was critical to ensure the return of protective airway reflexes before the patient resumed spontaneous handling of secretions (17).

Multidisciplinary collaboration between anaesthesiologists, paediatric surgeons, and ICU caregivers was essential for anticipating complications and optimizing overall recovery. The favourable postoperative course demonstrated how careful planning, skilled execution of airway and ventilatory strategies, and vigilant monitoring contribute to safe outcomes in extremely high-risk infants (18).

This case reinforces important principles in paediatric anaesthesia: anticipate airway challenges, avoid factors that elevate intracranial pressure, implement aggressive aspiration prophylaxis, and ensure continuity of respiratory support in the postoperative period. The success achieved highlights the value of individualized anaesthetic approaches informed by a clear understanding of pathophysiology and refined by modern airway and monitoring technologies.

CONCLUSION

This case highlights the significant anaesthetic challenges associated with infants having multisystem congenital anomalies such as hydrocephalus, severe GERD, airway deformities, and recurrent aspiration-related lung compromise. A tailored anaesthetic plan emphasizing secure airway management, aspiration prevention, meticulous neuroprotection, and gentle ventilatory strategies was essential in ensuring safe perioperative care. The use of video laryngoscopy for atraumatic airway access, avoidance of nitrous oxide, strict control of intra-abdominal pressure during laparoscopy, and judicious postoperative monitoring were pivotal in preventing complications. Effective multidisciplinary coordination further contributed to a smooth clinical course and favourable outcome. This case underscores the importance of comprehensive planning and evidence-based strategies for managing high-risk paediatric patients, reaffirming that successful anaesthesia in complex settings depends on vigilance, preparedness, and individualized clinical decision-making.

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