Section: Miscellaneous

Case Report

ANAESTHETIC MANAGEMENT OF AN INFANT WITH CONGENITAL MYASTHENIC SYNDROME UNDERGOING PERCUTANEOUS ENDOSCOPIC GASTROSTOMY: A CASE REPORT

Abhijit Sen[1], Febin Sathar[2], Ahmed Mohamed Nabil[3], Bharat Sapra[4]

[1]Head of the Department Anaesthesia, Zulekha hospital, Sharjah
[2]Specialist Department Anaesthesia, Zulekha hospital, Sharjah
[3]Paediatric Specialist Surgery Zulekha hospital, Sharjah
[4]Consultant Gastroenterologist Zulekha hospital Sharjah

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Corresponding Author:
Dr. Febin Sathar,
Email: drfebz@live.com

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ABSTRACT

Background: Congenital myasthenic syndrome (CMS) is a rare inherited disorder of the neuromuscular junction characterized by defective synaptic transmission, resulting in muscle weakness, hypotonia, and risk of respiratory insufficiency. Perioperative management is challenging due to unpredictable responses to sedatives, opioids, and neuromuscular blocking agents. **Case Presentation:** We report the anaesthetic management of a 7-month-old infant with genetically confirmed CMS who presented with feeding difficulties and failure to thrive and was scheduled for percutaneous endoscopic gastrostomy (PEG) tube insertion. Preoperative evaluation revealed generalized hypotonia, multiple joint contractures, unilateral bronchopneumonia, impaired gag reflex, inability to swallow properly, and increased susceptibility to respiratory depression. Anaesthesia was induced with titrated intravenous propofol and minimal-dose fentanyl. Neuromuscular blocking agents were avoided. Maintenance was achieved using sevoflurane in an oxygen–air mixture and remifentanil infusion. Standard ASA monitoring, including capnography, was employed throughout the procedure. **Result:** The PEG insertion was completed uneventfully without intraoperative complications. The patient maintained stable hemodynamics and adequate ventilation throughout. Postoperatively, the patient shifted to the ICU on a ventilator and the next day demonstrated effective spontaneous respiration and was weaned from ventilatory support, with an overall smooth recovery. **Conclusion:** This case highlights that safe anaesthetic management in infants with CMS can be achieved through careful drug selection and avoidance of neuromuscular blockade. Vigilant perioperative monitoring and individualized planning are essential to minimize respiratory complications in this high-risk population.

INTRODUCTION

Congenital myasthenic syndromes (CMS) represent a group of rare inherited neuromuscular disorders caused by defects in proteins essential for effective neuromuscular transmission. The reported prevalence is approximately 9 per million population, although the true burden may be underestimated due to underdiagnosis.[1] These disorders arise from genetic mutations affecting different components of the neuromuscular junction, including presynaptic vesicle release mechanisms, synaptic basal lamina proteins, or postsynaptic acetylcholine receptor function, ultimately resulting in impaired signal transmission and muscle weakness.

In contrast to Myasthenia Gravis, which is mediated by autoantibodies targeting neuromuscular junction proteins, CMS is non-immune in origin and does not involve antibody-mediated receptor destruction.[2,3] It is also distinct from transient neonatal myasthenia, which occurs due to placental transfer of maternal antibodies and resolves spontaneously.[3] The clinical presentation of CMS varies widely depending on the underlying genetic defect but commonly includes generalized hypotonia, feeding difficulties, respiratory insufficiency, and recurrent infections, particularly in infancy.[1,2]

Anaesthetic management of patients with CMS is particularly challenging due to altered pharmacodynamic responses at the neuromuscular junction. These patients are known to exhibit increased sensitivity to sedative and opioid agents, which may precipitate respiratory depression even at low doses.[4,5] Additionally, both depolarizing and

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non-depolarizing neuromuscular blocking agents can produce unpredictable and often exaggerated effects, including prolonged paralysis and delayed recovery.[5–7] The lack of robust evidence regarding safe dosing, monitoring, and reversal of neuromuscular blockade further complicates perioperative management.[4,6]

Another important consideration is the variability in medical treatment among CMS subtypes. While some patients benefit from acetylcholinesterase inhibitors, others may show minimal or even adverse responses, necessitating individualized perioperative planning.[2] Furthermore, concerns have been raised regarding the potential effects of certain anaesthetic agents, including local anaesthetics, on neuromuscular transmission, although evidence remains limited.[5]

Infants with CMS frequently develop feeding difficulties due to poor coordination of swallowing and respiratory compromise, often requiring enteral feeding support such as percutaneous endoscopic gastrostomy (PEG). Although PEG insertion is a relatively short procedure, it carries significant anaesthetic risk in this population due to the potential for perioperative respiratory failure.[6,7]

Given these challenges, anaesthetic strategies in CMS focus on avoiding neuromuscular blocking agents, minimizing respiratory depressant drugs, and preserving spontaneous ventilation whenever possible. Careful titration of short-acting agents and vigilant perioperative monitoring are essential to ensure safe outcomes.[4–7]

In this report, we describe the successful anaesthetic management of an infant with congenital myasthenic syndrome undergoing PEG tube insertion, highlighting key perioperative considerations and the importance of individualized anaesthetic planning in this high-risk group.

Case Presentation

A 7-month-old male infant, weighing approximately 4.9 kg, with a known diagnosis of congenital myasthenic syndrome (CMS), was admitted with complaints of recurrent respiratory distress, cough, and episodes of oxygen desaturation. The infant had a history of generalized hypotonia, feeding difficulties, and recurrent lower respiratory tract infections, requiring frequent hospitalizations and respiratory support.

Birth and Neonatal History: The patient was born at 30+5 weeks of gestation via lower segment cesarean section due to fetal distress. Birth weight was 1.32 kg, and the infant required immediate ventilatory support for respiratory distress syndrome. The neonatal course was complicated by apnea of prematurity, chronic lung disease, and multiple joint contractures consistent with arthrogryposis. The infant also had a history of neonatal jaundice requiring phototherapy and transient metabolic abnormalities, including hypocalcemia and hyponatremia.

Diagnostic Evaluation: Given persistent hypotonia and feeding difficulty, genetic evaluation was performed. Whole exome sequencing revealed a mutation in the CHRND gene, suggestive of congenital myasthenic syndrome (fast-channel syndrome variant). Additional neurological evaluation demonstrated reduced muscle tone, weak cry, poor suck–swallow coordination, and decreased spontaneous movements. Brain MRI showed immature myelination appropriate for age, with no structural abnormalities.

Clinical Course: The infant continued to have recurrent episodes of desaturation and required high-flow nasal cannula (HFNC) support intermittently. Feeding was maintained via nasogastric tube due to poor oral intake and risk of aspiration. The child was started on pyridostigmine, along with supportive therapies including bronchodilator (salbutamol) and steroid (budesonide) nebulization.

Due to persistent feeding difficulties and failure to thrive, percutaneous endoscopic gastrostomy (PEG) tube insertion was planned to ensure long-term nutritional support.

Preoperative Assessment: On preoperative evaluation, the infant was hemodynamically stable. Airway assessment revealed no anticipated difficulty. Respiratory examination showed a history of compromised respiratory reserve with prior desaturation episodes, although the child was stable at the time of surgery on minimal oxygen support. Neurological examination was significant for generalized hypotonia. The patient was receiving pyridostigmine at a dose of 6 mg every 4 hours, which was continued perioperatively in accordance with standard management guidelines for congenital myasthenic syndrome.

Given the underlying neuromuscular disorder, high-risk informed consent was obtained from the parents, explaining the possibility of perioperative respiratory depression, need for postoperative ventilation, and intensive care monitoring.

Anaesthetic Management

A carefully tailored anaesthetic plan was formulated with the following objectives:

- Avoidance of neuromuscular blocking agents
- Minimization of respiratory depressant drugs

Anaesthesia was induced with intravenous propofol in titrated doses. Glycopyrrolate 20 mcg was administered. The total titrated dose of propofol was 10 mg. Sevoflurane 6% was used, along with minimal-dose fentanyl 2 mcg for analgesia. No neuromuscular blocking agents were administered. The airway was secured with an endotracheal tube size 3 mm ID uncuffed.

Anaesthesia was maintained using sevoflurane in an oxygen–air mixture, with careful titration to maintain adequate depth. Remifentanil infusion (25 mcg/kg/min) and propofol infusion were administered. Paracetamol support 125 mg was given. Intraoperative monitoring included electrocardiography (ECG), non-invasive blood pressure (NIBP), pulse oximetry (SpO_2), and end-tidal carbon dioxide ($EtCO_2$).

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Intraoperative Course: The PEG procedure was completed successfully over approximately 3 hrs. The patient remained hemodynamically stable throughout the procedure, with no episodes of desaturation or airway complications. Neuromuscular blocking agents were not required at any stage. The procedure was converted from endoscopic-assisted PEG to open gastrostomy, resulting in a prolonged surgical duration.

Postoperative Course: At the end of the procedure, the patient was shifted to the ICU on ventilatory support and was successfully extubated the next day by the neonatologist. The infant remained in the intensive care unit for close observation. There were no episodes of postoperative apnea or respiratory compromise. After extubation, enteral feeding via the PEG tube was initiated successfully, and the patient showed a stable recovery.

Ethical Approval and Patient Consent: Written informed consent was obtained from the patient for publication of this case report and accompanying images. Formal Institutional Ethics Committee approval was not required for this single case report, as per institutional policy.

DISCUSSION

Congenital myasthenic syndromes (CMS) pose significant perioperative challenges due to impaired neuromuscular transmission and variable responses to anaesthetic agents. The primary concern in these patients is the risk of perioperative and postoperative respiratory compromise, which may arise from both disease-related muscle weakness and pharmacologic effects of anaesthetic drugs.

A key consideration in CMS is the unpredictable response to neuromuscular blocking agents. Previous reports have demonstrated exaggerated or prolonged effects following administration of both depolarizing and non-depolarizing muscle relaxants, with difficulties in reversal and risk of recurarization.[1,6,7] The case described by Knio et al. highlights this concern, where repeated doses of sugammadex were required to reverse rocuronium-induced blockade, and the patient subsequently developed severe postoperative hypoxia and bradycardia requiring resuscitation.[1] The possibility of recurarization or central apnea was considered, emphasizing the vulnerability of this population even after apparent intraoperative recovery.

In contrast, neuromuscular blocking agents were completely avoided in our case. This strategy was chosen to eliminate the risk of prolonged paralysis and unpredictable recovery. This approach is supported by existing literature, which recommends minimizing or avoiding muscle relaxants whenever feasible in patients with neuromuscular junction disorders.[6–9]

Another important aspect is the increased sensitivity of CMS patients to sedatives and opioids. Even small doses can lead to significant respiratory depression due to compromised neuromuscular transmission.[7,8] In our case, anaesthetic induction was achieved with carefully titrated propofol and minimal-dose opioid, ensuring adequate anaesthesia while avoiding excessive respiratory suppression. Similar strategies have been reported in other neuromuscular disorders, including mitochondrial myopathies, where short-acting agents and controlled sedation have been successfully used for procedures such as PEG insertion.[10]

The use of inhalational agents like sevoflurane provides additional advantages, including rapid titratability and minimal residual effects. Maintaining anaesthesia with sevoflurane has been described as a safe and effective approach in such patients. Intraoperative monitoring, including end-tidal CO_2 , is essential to ensure adequate ventilation and early detection of hypoventilation.

Postoperative respiratory events remain a major concern in CMS. The case reported by Knio et al. demonstrated delayed respiratory deterioration occurring two hours after extubation, requiring emergent intervention.[1] This highlights that even when intraoperative and immediate postoperative periods appear uneventful, delayed complications can occur. In our patient, careful postoperative monitoring in a high-dependency setting allowed early detection of any potential complications, although the recovery remained uneventful without episodes of apnea or respiratory failure.

Feeding difficulties are common in infants with CMS due to poor coordination of swallowing and respiratory muscles, often necessitating PEG placement for long-term nutritional support. Although PEG insertion is a relatively short procedure, it carries significant anaesthetic risk in this population due to compromised respiratory reserve. Therefore, an individualized anaesthetic plan focusing on minimal drug use, avoidance of neuromuscular blockade, and preservation of spontaneous ventilation is essential.

This case reinforces several important principles in the anaesthetic management of CMS:

- Avoidance of neuromuscular blocking agents to prevent prolonged paralysis
- Use of short-acting anaesthetic agents with careful titration
- Maintenance of spontaneous ventilation whenever possible
- Vigilant postoperative monitoring for delayed respiratory complications

Additionally, this case contributes to the limited literature on anaesthetic management of infants with CMS undergoing PEG insertion. Compared to previously reported cases with significant postoperative complications, the favorable outcome in our patient underscores the importance of tailored anaesthetic strategies based on pathophysiological understanding.

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CONCLUSION

Anaesthetic management of infants with congenital myasthenic syndrome requires a cautious, individualized approach due to impaired neuromuscular transmission and the risk of perioperative respiratory compromise. This case demonstrates that safe outcomes can be achieved by avoiding neuromuscular blocking agents, minimizing the use of respiratory depressant drugs, and maintaining spontaneous ventilation or controlled ventilation with carefully titrated short-acting anaesthetic agents. Vigilant intraoperative monitoring and close postoperative observation are essential, as delayed respiratory events may occur even after an apparently uneventful procedure. A multidisciplinary strategy and thorough understanding of the disease physiology are key to optimizing perioperative care in this high-risk population.

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