Case Report

Anaesthetic Management of Circumcision and PEG Tube Replacement in an Infant with Congenital Myasthenic Syndrome: A Case Report

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Abstract

Background: Congenital myasthenic syndrome (CMS) is a rare genetic disease of neuromuscular transmission with abnormal synaptic transmission, which causes weakness, hypotonia and risk of respiratory insufficiency. It is accompanied by enhanced sensitivity to anaesthetic agents and a significant risk of perioperative respiratory effects. There are further complications to anaesthetic management due to repeated surgical procedures. **Case presentation:** We present an infant with CMS, who had undergone percutaneous endoscopic gastrostomy (PEG) insertion earlier and again presented for circumcision and PEG tube replacement. Persistent hypotonia was found preoperatively while the respiratory status was stable. Sevoflurane and minimal opiate supplementation titrated with intravenous propofol were used for induction of anaesthesia. Neuromuscular blocking agents were not used. Sevoflurane in an oxygen–air mixture and remifentanil infusion were used with spontaneous ventilation for maintenance. **Results:** No complications during the procedure, both procedures completed successfully. Throughout, the patient was hemodynamically stable and had adequate spontaneous respiration. There were no complications in the treatment of the patient, and ventilatory support was not needed during the recovery period. **Conclusion:** It is safe to repeat anaesthetic exposure in infants with CMS provided that the exposure is carefully planned, neuromuscular blocking agents are avoided and ventilation remains spontaneous. To achieve the best results, individual anesthetic plans should be developed and carefully monitored.

Keywords: Congenital myasthenic syndrome, circumcision, PEG tube replacement, paediatric anaesthesia, spontaneous ventilation

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Introduction

Congenital myasthenic syndromes (CMS) are heterogeneous syndromes of congenital defects in proteins involved in the transmission across the neuromuscular junction. These defects affect how the signal is transmitted between the two neurons, leading to muscular weakness, fatigue and variable levels of respiratory dysfunction. CMS is not an immune disorder and has varying sensitivity to drugs in relation to the underlying genetic form of the disease.[1–3]

Given the lack of evidence specific to CMS, principles of anaesthetic management are extrapolated from autoimmune MG, such as careful titration of anaesthetic agents (including sedatives, opioids and muscle relaxants), neuromuscular monitoring, and close postoperative monitoring for the risk of prolonged neuromuscular blockade and respiratory insufficiency.[4,9]

If a patient needs multiple surgeries, then the repeated drug exposure, and disease progression, can affect anaesthetic response. In paediatric patients and minor procedures, many clinicians choose not to use a neuromuscular blocking agent as a precautionary measure to reduce risk.In minor procedures and paediatric patients, however, it was many clinicians' preference to avoid using a neuromuscular blocking agent to avoid risk.

Further, minor procedures like circumcision necessitate proper anaesthetic planning in infants with CMS, who often have feeding difficulties and failure to thrive, which stems from their swallowing coordination problems.[1,2] Anesthetic options for regional techniques (e.g., caudal blocks), short-acting sedative agents, and airway devices are commonly used during circumcision to assure safety and good pain control, particularly in the presence of underlying neuromuscular disorders as evidence from anaesthesia practice surveys suggests.[11]

In light of this, anaesthetic techniques in CMS emphasize avoiding the use of neuromuscular blocking agents when possible, limiting the use of respiratory depressant drugs, maintaining spontaneous respiration and ensuring close perioperative monitoring.

This report outlines the successful anaesthetic management of two repeat surgical procedures (circumcision and PEG tube replacement) in a baby with congenital myasthenic syndrome (CMS) to highlight the peri-operative techniques that were used and the importance of individualised anaesthetic planning in this high-risk cohort.

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Case Presentation

Patient Profile: An 8-month-old male infant with a history of congenital myasthenic syndrome (CMS) presented for replacement of his percutaneous endoscopic gastrostomy (PEG) tube and elective circumcision, two months following initial placement of a PEG. The child was known to have generalized hypotonia and feeding issues, and PEG placement was conducted previously, and the child had recovered.

Clinical Status: On presentation, the baby was clinically stable and had no history of recent respiratory distress or any infections or hospitalisations. There was however a long-standing hypotonia noted which was consistent with the underlying neuromuscular disorder. All previous anaesthetic experience had been uneventful, and reassuring but not without risk of perioperative events.

Preoperative Assessment: Preoperative assessments had good hemodynamics and good baseline respiratory function. The patient was at high risk for perioperative respiratory depression due to increased sensitivity to anaesthetic agents because of the diagnosis of CMS. Parents were obtained informed consent, including informed consent for postoperative ventilatory support, that was deemed to be high-risk. Standard fasts were adhered to and the patient continued receiving pyridostigmine 6 mg every 4 hours and syrup glycopyrrolate until the procedure.

Anaesthetic Management: A specific anaesthetic protocol was designed which focused on minimizing respiratory compromise. Analgesia was provided using a Paracetamol suppository of 60 mg, and induction with titrated intravenous Propofol and also intravenous infusion of Remifentanil. A 1.5 LMA was placed. Deliberate avoidance of neuromuscular blocking agents to avoid prolonged muscle weakness. Spontaneous ventilation was maintained throughout the procedure and anaesthesia was maintained with a mixture of oxygen and air containing sevoflurane and a remifentanil infusion.

Intraoperative Course: A penile block 2 mL of 0.125% bupivacaine was administered by the paediatric surgeon prior to the surgery.

The skin was circumcised first and then the replacement of the PEG tube. During the operation, no desaturations or airway problems occurred, and the hemodynamics remained stable. Pulse oximetry, electrocardiography, non-invasive blood pressure and end-tidal carbon dioxide were used to continuously monitor for any changes in physiology.

Postoperative Course: Infant's spontaneous respiratory effort and oxygen saturation levels were adequate at the end of the procedure. The recovery period after surgery was uneventful, without any episodes of apnea, desaturation or respiratory distress. The patient was observed appropriately and did not need ventilatory support and so was discharged with a good outcome. The infant after extubation was moving all four limbs with good muscle tone.

This case serves as an example of how repeated anaesthetic exposure can be safely handled in infants with Congenital Myasthenic Syndrome (CMS) using careful planning, avoiding Neuro-Muscular Blockers (NMBs) and maintaining spontaneous ventilation, and careful perioperative monitoring.

Ethical Approval and Patient Consent: Written informed consent was obtained from the patient for this case report and the accompanying images. This is an individual case report, and formal Institutional Ethics Committee approval was not required as per institutional policy.

Discussion

Congenital myasthenic syndromes (CMS) are a diverse group of inherited disorders in which genetic defects at the neuromuscular junction cause impaired neuromuscular transmission. The clinical spectrum is diverse, depending on the type of underlying mutation, and most patients have clinical signs of hypotonia, fatigable weakness and varying degrees of respiratory involvement and this is important for anaesthetic management, especially in the infant population who have limited respiratory reserve.

An important perioperative issue in CMS is how the body reacts to the anaesthetic drugs, and this reaction can be different from what is predicted. This indicates the possibility of both a prolonged and exaggerated effect following neuromuscular blockade or "recurarization" after rocuronium use in CMS patients, as well as difficulties in achieving a full reversal after neuromuscular blockade, and can result in severe postoperative hypoxia and bradycardia requiring resuscitative measures as was seen in the patient reported by Knio et al.[1]

With these concerns in mind, it is sometimes recommended to avoid the use of neuromuscular blocking agents, particularly in shorter procedures where adequate conditions for use of an OCA can be obtained without the use of paralysis. In the present case, the neuromuscular blockers were intentionally withheld and spontaneous breathing was ensured throughout the procedure. This will reduce the risk of prolonged neuromuscular weakness, postoperative ventilatory failure, and is in line with the literature on neuromuscular disorders.[7,8]

Anaesthetic management is often extrapolated from that of MG, as there is limited CMS specific evidence. The latter consist of careful titration of short-acting agents, reduction of opioid usage and careful monitoring of the patient during the perioperative period. We used induction with titrated propofol and minimal-dose fentanyl with maintenance with sevoflurane to ensure adequate anaesthetic depth, while maintaining respiratory function. Advantageous use of inhalational agents like sevoflurane, because the onset and offset is rapid, thus recovery is smooth.

Additional considerations are presented when ingesting a drug repeatedly, as occurred in this patient. Previous uneventful anaesthesia is helpful but cannot guarantee that there will not be some risk because this may be changed over time with disease and cumulative drug effects. In more complex procedures, like scoliosis surgery, it is demonstrated that careful use of reduced-dose muscle relaxants (NMBB) with neuromuscular monitoring and postoperative ventilatory support (VM) is successful.[10] but in minor procedures, physicians prefer to avoid using muscle relaxants.

Although a minor surgical procedure, procedures like circumcision still require proper anaesthesia and analgesia,

especially in CMS children, where feedings are often difficult and failure to thrive is a common occurrence. Based on survey data, airway devices including laryngeal masks, caudal blocks and short-acting sedative drugs are used to ensure safety and are commonly practised for paediatric circumcision, with airway devices being tailored towards ensuring spontaneous ventilation, and avoiding excessive sedation.[11]

Respiratory problems continue to be the biggest risk for patients undergoing CMS. Postoperative monitoring is necessary, as delayed respiratory depression, apnea and hypoxia have been reported after seemingly uneventful courses during surgery.[1,9] The patient recovered without respiratory compromise in the present setting, probably because the patient did not receive neuromuscular blockers, had minimal use of opioids and did not require the use of artificial ventilation.

In summary, this case highlights the importance of tailored planning according to the physiologic characteristics of CMS, as well as the need to avoid or use carefully neuromuscular blocking drugs, to favour short acting anaesthetic drugs and to monitor closely all the peri-operative periods. The following successful procedure confirms the safety of an individualised and careful anaesthetic management of CMS patients.

Conclusion

Care of infants with congenital myasthenic syndrome who are undergoing multiple surgeries should be handled with caution and individualized approach because of a potential for perioperative respiratory compromise and impaired neuromuscular transmission. The case shows that a good result can be obtained if the dosage of drugs which depress the respiratory function is kept to a minimum and neuromuscular blocking drugs avoided, with a combination of carefully titrated doses of short-acting anaesthetic drugs, allowing spontaneous respiration. Past uneventful anaesthetic will guide, but every anaesthetic should be treated individually and closely monitored. It is important to plan and prepare for the perioperative period carefully and to ensure safety in this high-risk population.

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Conflicts of interest

There are no conflicts of interest.

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