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Case report

Anesthetic implications of Möbius syndrome

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Abstract Möbius syndrome is a rare disorder characterized by unilateral or bilateral facial paralysis and defective extraocular eye movements secondary to congenital paresis of the facial (VII) and abducens (VI) cranial nerves. Associated dysfunction of other cranial nerves, orofacial abnormalities, and skeletal muscle hypotonia are common accompanying features. Given the multisystem involvement of Möbius syndrome, there are several potential perioperative concerns. Of primary importance to the anesthesia provider are the propensity for postoperative respiratory failure due to several pathological mechanisms, associated orofacial abnormalities that may make routine airway management difficult, the presence of gastroesophageal reflux and other factors that increase the risk of perioperative aspiration, and associated hypotonia of the skeletal musculature. The authors present a 10-month-old infant with Möbius syndrome who required anesthetic care during a Nissen fundoplication. The potential anesthetic implications of this syndrome are discussed.

1. Introduction

Möbius (or common English spelling Moebius) syndrome is a rare neurological disorder characterized by unilateral or bilateral facial paralysis and defective extraocular eye movements secondary to congenital paresis of the facial (VII) and abducens (VI) cranial nerves [1,2]. These classic features of the syndrome are often accompanied by other cranial nerve palsies [3-5]. Affected infants typically present with congenital esotropia and immobile, expressionless facies. Depending on the pattern of cranial nerve involvement, there may be a wide range of clinical expression. Feeding difficulties due to poor coordination of sucking and swallowing may be present with IXth and Xth cranial nerve involvement. This may be associated with dysphagia and retention of oral secretions leading to recurrent bouts of aspiration pneumonia. Inadequate function of the soft palate can also result in dysarthria. Möbius syndrome may also occur in association with various craniofacial, limb, and musculoskeletal malformations as well as multiple ophthalmic abnormalities [4,6,7]. Other associated manifestations include seizure disorders, hypogonadotropic hypogonadism, hypotonia, and some degree of mental retardation [8].

To date, there are only 3 published reports addressing the relevant anesthetic issues in patients with Möbius syndrome [9-11]. Krajcirik et al [9] described the anesthetic care of a 13-year-old girl with Möbius syndrome during a diagnostic fiberoptic bronchoscopy for persistent pneumonia.

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The anesthetic regimen included intravenous induction with fentanyl and diazepam, neuromuscular blockade with pancuronium and D-tubocurarine, followed by maintenance anesthesia with nitrous oxide and fentanyl. Airway management and tracheal intubation were uneventful except for the presence of copious tracheal secretions. Postoperative problems included acute respiratory failure secondary to excessive airway secretions, which required tracheal intubation and postoperative mechanical ventilation for 48 hours. Another case report of a single patient undergoing dental surgery under general anesthesia was presented by Ha and Messieha [10]. Aside from the need for general anesthesia due to the patient's situational anxiety and violent behavior, no other perioperative complications were noted.

Ferguson [11] retrospectively reviewed the anesthetic experience of 59 anesthetics in 19 children with Möbius syndrome over a 15-year period. The most common surgical procedures included either strabismus surgery or orthopedic procedures to improve limb function. They emphasized the potential airway implications of Möbius syndrome as several of the children had associated micrognathia, microstomia, or limited mouth opening. Endotracheal intubation was uneventful in 28 of 41 attempts and considered difficult in 11 attempts in 8 children. In these 8 children, there were 2 failed intubations and the airway was managed with a laryngeal mask airway, although placement was noted to be difficult requiring more than 1 attempt in both cases. Other perioperative issues included the potential for copious airway secretions, upper airway obstruction, and respiratory failure. We report the anesthetic management of a 10-month-old infant with Möbius syndrome during a repeat Nissen fundoplication. The potential anesthetic implications of this syndrome are discussed.

2. Case report

Review of the patient's hospital records and presentation of this case report were approved by the Institutional Review Board of the University of Missouri. A 10-month-old, 12.8-kg, white male infant presented for repeat fundoplication for treatment of recurrent aspiration pneumonia. Recurrent gastroesophageal reflux (GER) was demonstrated by a pH probe study after the recurrent bouts of pneumonia. The infant had been diagnosed with Möbius syndrome as a neonate because of the presence of congenital paresis of cranial nerves VI and VII. Other manifestations during the neonatal period included poor pharyngeal muscle control, upper airway obstruction, and central apnea. His neonatal course was complicated by poor feeding, GER, and respiratory failure, which necessitated tracheostomy with home mechanical ventilation, Nissen fundoplication, and feeding gastrostomy. Anesthetic care at that time included fentanyl and pancuronium. The postoperative course was complicated by an episode of pneumonia and bacteremia. The patient had a history of seizures during the neonatal period, which were controlled with oral phenobarbital (30 mg per gastrostomy tube twice a day). Other medications included a multivitamin preparation and ranitidine (15 mg per gastrostomy tube twice a day). Previous anesthetic and surgical history were negative except for the tracheostomy, G-tube, and Nissen fundoplication during the neonatal period.

Current home ventilator settings included room air, positive end expiratory pressure of 5 cm H_2O, and pressure support of 5 cm H_2O. Physical examination revealed an infant in no acute distress. His vital signs were normal for his age. The mother did report episodes of bradycardia (heart rate of 60 beats per minute) during sleep, which were noted on his home cardiac monitor. The physical examination revealed the stigmata of Möbius syndrome, including absent facial expression and no extraocular eye movements. Significant hypotonia was present, and the infant was in a frog-leg position. Micrognathia was present as well as a well-healed tracheostomy site with a 3.5-mm–internal diameter pediatric tracheostomy tube in place. The cardiorespiratory examination was unremarkable. Echocardiography 2 months earlier revealed normal ventricular function. The patient was held *nil per os* for 4 hours before the surgical procedure except for his usual morning dose of phenobarbital. Ranitidine and metoclopramide were administered via the gastrostomy tube 1 hour before the surgical procedure. Inhalation induction was performed using incremental doses of sevoflurane in oxygen via the tracheostomy. Once an acceptable depth of anesthesia was obtained, a peripheral intravenous cannula was placed and glycopyrrolate (0.05 mg) and mivarcurium (0.2 mg/kg) were administered. His eyes were protected by the application of artificial tears and tape. Maintenance anesthesia consisted of desflurane in air/oxygen and fentanyl (3 μg/kg). The surgical procedure lasted 90 minutes, and the infant was transported to the Pediatric ICU to continue postoperative positive pressure mechanical ventilation until the afternoon of postoperative day (POD) 1. At that time, he was transitioned back to his home ventilator and his baseline ventilator settings. Phenobarbital was administered intravenously (30 mg every 12 hours) until enteral feeds were resumed on POD 3 at which he was transitioned back to phenobarbital per the gastrostomy tube. Pain control was provided by fixed interval dosing of acetaminophen (initial dose of 40 mg/kg PR followed by 15 mg/kg every 6 hours) supplemented as needed with ketorolac (0.5 mg every 6 hours prn) and nalbuphine (0.05 mg/kg every 2 hours prn). Given the patient's lack of facial expression and hypotonia, the assessment of pain was based on physiological parameters (alterations in heart rate and blood pressure) as well as significant input from the mother regarding his level of comfort. On POD 3, enteral feeds were restarted and increased to full feeds on POD 5. There were no postoperative complications, and he was discharged home on POD 6.