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Anesthetic Management of an Adult Patient with Moebius Syndrome for Strabismus Surgery: A Case Report

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Abstract:
Moebius syndrome is a rare non progressive congenital neurologic disorder mostly involving cranial nerve dysfunction and craniofacial, limb, musculoskeletal and ophthalmic deformities that vary in presentation and severity. These pose several challenges to the anesthesiologist especially in terms of airway management. Currently, there is limited literature discussing the anesthetic management and considerations of adult patients with the syndrome for surgery. This paper presents a case of a 33 year old female presenting with congenital medial deviation of the eyes, features of Moebius syndrome and bronchial asthma who underwent strabismus surgery under general anesthesia. Due to anticipated difficult airway, preoperative interventions such as prophylaxis for airway hypersensitivity and preparation in line with a difficult airway algorithm were done. Airway management during induction of general anesthesia and the intraoperative course were unremarkable. The patient was able to tolerate the procedure well.

Keywords: Moebius syndrome 1, Congenital Oculofacial Paralysis, Difficult Airway, Strabismus Surgery, Total Intravenous Anesthesia (TIVA)

Introduction:

Moebius syndrome or congenital oculofacial paralysis is a rare, non-progressive congenital disorder characterized by unilateral or bilateral facial paralysis, extraocular muscle movement problems due to affectation of CNVII (facial nerve) and CNVI (abducens). Other features of the disease include multiple cranial nerve palsies (III, IV, V, IX, X, XII), craniofacial malformations (mandibular hypoplasia, microstomia, temporomandibular joint dysfunction, cleft palate, external ear deformities), limb (clubfoot, syndactyly), musculoskeletal malformations and ophthalmologic problems[1]. It has an estimated prevalence of 1/250,000 live births with equal incidence in both sexes. Most cases are noted to be sporadic in nature but a few are documented to be familial. The pathophysiology of the disease remains to be controversial but some theories behind its cause include fetal toxic exposure, genetic rhombencephalic disturbances or ischemic event during the 5th week of pregnancy[2].

As mentioned, features of the disease include facial paralysis, cranial nerve palsies, orofacial abnormalities and respiratory impairment due to hypotonia, among others. These contribute to perioperative concerns to the anesthesiologist such as difficult airway management in terms of intubation and ventilation as well as the risk for postoperative pulmonary complications when general anesthesia is performed[3][4]. Careful selection of a neuromuscular blocking agent (NMB) is suggested due to their ability to exacerbate poor

respiratory function and the risk for malignant hyperthermia[1][5]. The possibility of these complications therefore requires perioperative planning.

Case Description:

We present a report on the anesthetic management using TIVA (total intravenous anesthesia) with an LMA (laryngeal mask airway) of a 33 year old female with Moebius syndrome for strabismus surgery - bilateral medial rectus recession. The patient presented with esotropia since birth with alternating eye fixation as well as congenital non progressive facial weakness on the right side of the face. She did not present with any respiratory problems, intellectual disturbances and did not complain of any difficulties in feeding or speaking. She has undergone esotropia correction, OD (oculus dexter) at 4 years of age under general anesthesia with no reported complications. As for comorbidities, she has bronchial asthma maintained on Salmeterol + Fluticasone metered dose inhaler as needed.

On preoperative physical assessment, the patient weighs 50kg, with a height of 146 cm for a BMI of 23. The patient is assessed as American Society of Anesthesiologists Physical Classification (ASA) 2 for her history of controlled bronchial asthma. Furthermore, the patient is unable to abduct the left eye, strabismus, hearing loss on the right ear and a short left arm. Cranial nerve palsies that were observed in the patient were CN VI bilateral (inability to abduct OU), CN VII R (right facial asymmetry with weakness), and CN VIII R (impaired hearing on the right ear). Other systemic findings were found to be normal. Difficult airway was noted as manifested by a Mallampati score of 4, with a mouth opening of 2 fingerbreadths, adequate thyromental distance, no loose teeth and no dentures. Diagnostics revealed a normal 12L ECG, normal brain MRI, blood and chemistry parameters.

Figure 1. Patient's Airway Assessment. Note Mallampati 4 and Mouth Opening of ~2 Fingerbreadths.

Preoperatively, the patient was instructed to use prescribed bronchodilator inhaler and was given Hydrocortisone IV to avoid exacerbation of asthma. Prior to induction, a standby plan of securing the airway via endotracheal intubation using a video laryngoscope and with Rocuronium as a neuromuscular blocking agent was prepared. The patient was induced with Propofol IV bolus (total of 80mg) and Remifentanil dose rate starting at 0.05mkm. Once the patient was in a deep plane of anesthesia, confirmed by checking for eyelash reflex and the trapezius squeeze test, an appropriately sized LMA (Supreme size 4) was then inserted and secured. The absence of any leak and presence of breath sounds were then checked. Insertion of the LMA was uneventful and the patient was then maintained through TIVA using

propofol dose rate (5-10 mg/kg/hour) and remifentanil dose rate at 0.5-0.1 mkm. Ventilation was provided using Pressure Control setting. Emergence was smooth with no occurrence of laryngospasm and desaturations. Total OR was 2 hours and 15 minutes with no significant intraoperative adverse events. The postoperative course of the patient was insignificant and she was eventually discharged well one day after the procedure.

Figure 2. Post-induction Showing Placement of LMA Supreme Size 4

Discussion:

Patients with Moebius syndrome present several challenges to the anesthesiologist depending on their presenting features. These problems mainly involve managing a difficult airway in terms of ventilation and intubation and post-operative respiratory complications due to aspiration pneumonia secondary to palatopharyngeal dysfunction from cranial nerve impairment[4]. In 90% of patients, tracheal intubation is difficult due to craniofacial changes such as micrognathia, retrognathia, mandibular hypoplasia. In a study by Ferguson et al, difficulty was encountered in 11 of 41 intubation attempts with failure in intubating 2 patients mainly due to craniofacial abnormalities of the patients included in the study[3][5]. Hence, preparation for a difficult airway is key. According to Budic et al, facemask with or without an oral airway are appropriate choices for ventilation as it is usually adequate. In addition, a combination of methods may be used to facilitate intubation such as performing cricoid pressure or 2-person ventilation as well as use of airway adjuncts like a gum elastic bougie and a fiberoptic scope[1]. It is also suggested that algorithms for a potentially difficult airway should be followed, maintenance of spontaneous ventilation and avoidance of NMBA until confirmation of bag mask ventilation, and availability of adjuncts mentioned above should be on standby[3]. In our patient, factors contributing to a possible difficult airway include a high Mallampati score of 4, mouth opening of 2 fingerbreadths and a slightly large tongue. Additional preoperative imaging for airway evaluation such as a CT scan or MRI could have been done due to the anticipation of a difficult airway[8]. A CT scan would have helped in predicting a difficult airway by showing delineation of soft tissues in features such as in macroglossia[8][9]. The possibility of a difficult intubation was avoided through the use of a laryngeal mask airway (LMA). The absence of craniofacial

deformities and adequate mouth opening upon induction has facilitated the use a supgraglottic airway device. The absence of OSA also ensures patency of airway after anesthesia[10]. In case of failure of the LMA, a standby plan of intubation with Rocuronium using a videolaryngoscope was in place. The use of Rocuronium as a relaxant is to ensure reversibility using Sugammadex if unable to intubate and ventilate. Despite adequate preparation, planning could have been improved by availability of other pathways in a difficult airway algorithm such as the availability for call for help and trained personnel for an invasive airway and equipment such as a fiberoptic scope. As per 2022 American Society of Anesthesiologists guidelines for a difficult airway, other preparations that could have been done include an emergency pathway in cases of inability to ventilate and intubate (See Appendix) [6].

According to the same study by Budac et al., induction of anesthesia may be done intravenously or via inhalational route[1]. For a rapid induction, intravenous technique using propofol and remifentanil was done in the patient. No neuromuscular blockers were given during the procedure to allow the performance of a forced duction test intraoperatively and to avoid possible respiratory complications from their use. Patients with Moebius syndrome, with unclear pathophysioiology of their hypotonia, are considered at risk for rhabdomyolysis, hyperkalemia and malignant hyperthermia secondary to associated myopathy. Residual neuromuscular blockade may also contribute to postoperative respiratory complications due to associated hypotonia of the thoracic musculature and diaphragm [3]. Although there is still no causation regarding Moebius syndrome and malignant hyperthermia, Fernandes et al reports of such case after the infant was exposed to succinylcholine and sevoflurane [7]. Therefore, it is still prudent to avoid these agents as much as possible. The risk for these complications were decreased in our patient as no NMBs and halogenated agents were through the use of TIVA by Remifentanil and Propofol.

Conclusion:

In this case report, we were able to safely administer GA-TIVA using an LMA to an adult patient with Moebius syndrome despite the possibility of a difficult airway. The risk of other possible complications such as malignant hyperthermia and pulmonary aspiration were mitigated through avoidance of using neuromuscular blockers and inhalational agents by using solely TIVA. This highlights the importance of a thorough preoperative assessment and knowledge of the features of a patient with Moebius syndrome for ample preparation in case a difficult airway is encountered and for the prevention of perioperative complications.

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Appendix 1: 2022 American Society of Anesthesiologists Algorithm for Difficult Airway in Adults