Case Report

A child with Moebius syndrome for squint surgery: Anaesthetic management and literature review

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ABSTRACT

Moebius syndrome is an uncommon clinical disease characterised by unilateral or bilateral facial paralysis and abnormal extraocular eye movements. It is caused by congenital paresis of the facial (VII) and abducens (VI) cranial nerves and other genetic abnormalities affecting different bodily systems. We describe a child with Moebius syndrome who had undergone squint surgery under general anaesthesia and the anaesthetic issues it entailed.

Keywords: Moebius syndrome, general anaesthesia, squint surgery, airway

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Introduction

Moebius syndrome (MS) is a congenital paralysis of the facial (VII) and abducens (VI) cranial nerves that results in unilateral or bilateral facial paralysis and abnormal extraocular eye movements.[1] The syndrome's characteristic symptoms are often accompanied by hypoglossal (XII), trigeminal (V), glossopharyngeal (IX), and vagal (X) nerve palsies. Children affected by this condition usually have congenital esotropia and expressionless facies.[1] Clinical manifestations vary according to the pattern of cranial nerve involvement. Additionally, Moebius syndrome may occur in conjunction with a variety of other disorders. Mandibular hypoplasia may predispose the patient to have a problematic airway.[2]

These patients may require imaging and surgical interventions to correct eye anomalies (strabismus surgery, ptosis repair, tarsorrhaphy), orthopaedic issues (correction of limb abnormalities), plastic/reconstructive (cleft palate, jaw surgery, facial reanimation surgery), otolaryngological, dental (teeth extractions), or general surgical interventions.[3]

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The purpose of this article is to describe a child with diagnosed MS who had undergone squint surgery (MR Recession) under general anaesthesia and the anaesthetic complications that might be ensued.

Case report

A two-year-old male infant weighing 10 kgs came to the ophthalmology outpatient clinic with bilateral inward deviation of the eyes, and left eye lagophthalmos. He exhibited microstomia, drooping of the angle of the mouth (L > R), and mask-like facial characteristics and was thus referred for genetic evaluation, after which MS was identified. His mother was diagnosed with polyhydramnios during the pregnancy. Otherwise, his birth and growth history were normal. The child had bilateral sixth nerve palsy, left seventh nerve palsy, and left median entropion of the upper lid at the time of evaluation (figure 1).

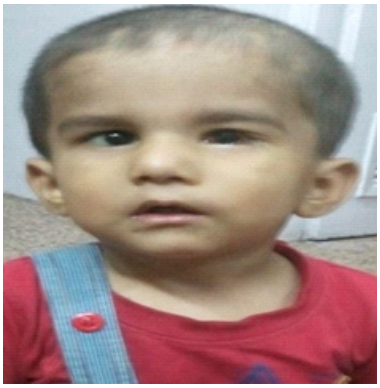

Figure 1: Moebius syndrome : Bilateral inward deviation of the eyes, left entropion of the upper lid and slight inclination of the head to the right

While walking, there was a slight inclination of the head to the right. There were no abnormalities seen in the airway. The cervical spine's X-ray revealed a widening of the interscapular distance. All biochemical analyses were within normal ranges. The systematic investigation revealed no anomalies.

During the preoperative evaluation, informed written permission was acquired from the child's parents.

All standard monitors (ECG, pulse oximetry, and NIBP) were attached in the operating room. We used incremental dosages of 5-8% sevoflurane in combination with oxygen and nitrous oxide for inhalational induction. Following induction, a 24 gauge cannula was secured in the left hand. Intravenous atracurium 2 mg and fentanyl 15 μg were administered when adequate mask ventilation was achieved, and the airway was maintained with a size 1.5 air-Q™ intubating laryngeal airway (ILA) (Cookgas LLC, Mercury Medical, Clearwater, FL, USA). Isoflurane and nitrous oxide at 50% O2 were used to maintain anaesthesia. Intravenous ondansetron 1.0 mg was given for postoperative nausea and vomiting prevention. At the conclusion of the operation, neuromuscular blockade was reversed by using 500 μg neostigmine and 100 μg glycopyrrolate intravenously. Once the child was completely awake, the Air-Q was removed. The patient's recovery and postoperative period were uneventful.

Discussion

While Von Graefe reported a case of congenital facial diplegia in 1880, Paul Julius Möbius, a German neurologist, investigated and characterised the condition further in 1888. It occurs at a rate of 0.002% of births, or one in every 50,000 live births.[1]

Moebius syndrome has a complex aetiology. Two major causative theories for MS include rhomboencephalic maldevelopment and brainstem ischemia during the first trimester.[2]

Additionally, hyperthermia, trauma, thrombus formation, embolism, bleeding, and in utero exposure to certain medicines, including misoprostol, are teratogenic. Moebius sequence inheritance is complex and may be autosomal recessive, autosomal dominant, or even X-linked. Although many potential areas and genes (3q21-q22 and 13q12.2-q13) have been reported, no causal gene has been identified to date.[1] Poland-Mobius syndrome is a rare congenital disease characterised by a combination of Poland and Mobius characteristics. Poland syndrome is characterised by the absence of the pectoralis major muscle, syndactyly, brachydactyly, and hand hypoplasia.1-3

These individuals may have micrognathia, mandibular hypoplasia, cleft palate, and temporomandibular joint dysfunction.3 All of these characteristics may make the bag and mask ventilation, intubation, and supra glottic airway device (SGA) placement problematic. We selected air-Q ILA for airway management in our patient because this SGA allows blind and fiberoptic guided intubation if required. If feasible, a thorough airway examination using indirect laryngoscopy should be performed in advance to map any upper airway abnormality. Oculocardiac reflex (OCR) is more common following extraocular muscle manipulation during squint surgery. MS is linked with the involvement of many organ systems. These issues, as well as their related anaesthetic problems,[1-7] have been summarised in Table-1.

Our patient had no problem with mask ventilation or insertion of the air-Q ILA. Ames et al examined 111 anaesthetic records from 46 MS patients who had undergone various surgical procedures.8 They found that face mask ventilation was easy in all patients, whereas tracheal intubation was uneventful in 76 cases (71.6% of all intubations). External laryngeal manipulation alone was needed in 17 (16%) cases to improve the view with laryngoscopy. In ten (9.4 per cent) cases, a combination of procedures was required, including cricoid pressure, stylet use, two person method, and blade replacement. Three patients needed fiberoptic intubation. Five patients' surgeries were postponed owing to unsuccessful intubation. Telich-Tarriba et al similarly examined 51 individuals with MS who underwent 172 procedures.9 They identified four individuals (7%) with a difficult airway. Endotracheal intubation was successful in all patients; 38 patients were intubated successfully on the first attempt, while the other patients required a second effort. Rasmussen et al described a 23-year-old man with MS who died due to difficulties during intubation and sudden circulatory collapse.[10]

Conclusion

MS may be associated with difficulties with the airway and other anaesthesia-related aspects. It is critical to be aware of these co-morbidities, since it may result in unexpected complications during anaesthesia, if left undiagnosed.

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Conflicts of interest

There are no conflicts of interest.

Table-1: The involvement of different organ systems and its anaesthetic implications in a patient with Moebius Syndrome[1,3,4-7]

Organ involvement	Characteristic findings	Anaesthetic concerns and their management
Airway	Temporomandibular joint dysfunction Cleft palate Congenital bilateral vocal cord paralysis4 Retention of oral secretions Nasal regurgitation	Difficult airway equipment should be available. Aspiration of oral secretions- antisialogogue premedication Non-opioid analgesics should be used preferentially. Postoperatively extended respiratory monitoring may be required
Face	Expressionless (Mask) facies Broad root of the nose Mandibular hypoplasia Microstomia Microglossia Micrognathia Hypertelorism	Difficult to measure and assess pain. The degree of analgesia should be determined by changes in physiological markers (heart rate and blood pressure). Difficulty in bag and mask ventilation
Musculoskeletal deformity	Club foot Smallness of limbs Syndactyly Brachydactyly Missing fingers or toes Arthrogryposis multiplex congenita Klippel-Feil anomaly Poland sequence (I/L abnormalities of the hands and a full or partial absence of the pectoralis muscles and breast) association with 15% of patients Webbing of axilla	Difficulty in positioning on OT table Avoid succinylcholine due to the danger of rhabdomyolysis, hyperkalemia, and malignant hyperthermia5 Difficulty in securing intravenous access
Neurological and skull	Paresis of the facial, abducens, hypoglossal, trigeminal, glossopharyngeal and vagal nerve Seizure disorders Hypotonia, Hydrosyringomyelia6 Holoprosencephaly7 Autism Speech problems Mental retardation (Rare)	Train-of-four monitoring due to hypotonia Hypoglossia or ankyloglossia combined with poor tongue coordination increases the risk of secretion.
Eye and Ear	**Eye**- Congenital esotropia, incomplete eye closure, ptosis, conjunctivitis, corneal opacities, ophthalmoplegia, epicanthus, lateral gaze paralysis **Ear**- External ear deformities, otitis media	Exposure keratopathy Oculocardiac reflex (OCR) and postoperative nausea and vomiting (PONV) are more common after squint surgery. Communication difficulties with child
Cardiovascular	Congenital heart diseases (ventricular septal defect, patent ductus arteriosus, dextrocardia)	Preoperative echocardiography, Anaesthetic considerations based on the cardiac lesion
Miscellaneous	Dysphagia Dysarthria Hypogonadotropic hypogonadism Prematurity Café-au-lait pigmentation	Feeding problems as a result of inadequate sucking and swallowing

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