

Möbius Syndrome, What Anesthesiologist Should Consider About: A Case Report

Masoud Nashibi ©
Parisa Sezari ©
Farhad Safari ©
Ahmad Naderi ©
Kamran Mottaghi ©

Möbius Sendromu, Anestezi Uzmanının Nelere Dikkat Etmesi Gerekir: Olgu Sunumu

ABSTRACT

First described in 1888, Möbius syndrome is a variety of cranial nerve palsy combinations mainly involving sixth, seventh and eighth cranial nerves. Although rare, affected infants and children might be referred to anesthesiologists for many types of ophthalmologic or otologic surgeries. Data on anesthetic management of such patients are scarce and case reports can be a source of useful information for anesthesiologists throughout the world.

Since it involves facial muscles and their development, affected patient potentially may be a case of difficult airway as well; although it is rare. Malignant hyperthermia is another potential threat.

We report a known case of Möbius syndrome candidate for cochlear implantation under general anesthesia.

Although our case was uneventful, the anesthesiology team has to be prepared for potential risks such as difficult airway management or malignant hyperthermia in these patients.

Keywords: Möbius syndrome, difficult airway, malignant hyperthermia, anesthetic considerations, cochlear implant

Öz

İlk kez 1888'de tarif edilen Möbius sendromu başlıca altıncı, 7. ve 8. kranial sinirlerin kombinasyonunu içeren kranial sinir felcidir. Nadir de olsa, etkilenen bebekler ve çocuklar birçok oftalmolojik veya otolojik cerrahi için anesteziyologlara yönlendirilebilir. Bu tür hastaların anestezi yönetimine ilişkin veriler azdır, olgu sunumları tüm dünyada anesteziyologlar için yararlı bir bilgi kaynağı olabilir.

Yüz kaslarını ve gelişimini etkilediği için, etkilenen hasta potansiyel zor hava yolu olabilir, ancak nadirdir. Malign hipertermi başka bir potansiyel tehdittir.

Genel anestezi altında kohlear implantasyon adayı bir Möbius sendromu olgusunu sunuyoruz. Olgumuz sorunsuz olmasına rağmen, bu hastalarda anesteziyoloji ekibi zor hava yolu yönetimi veya malign hipertermi gibi potansiyel risklere hazırlıklı olmalıdır.

Anahtar kelimeler: Möbius sendromu, zor hava yolu, malign hipertermi, anesteziyoloji özellikler, kohlear implant

Received: 18 January 2020

Accepted: 18 May 2020

Publication date: 30 July 2020

Cite as: Nashibi M, Sezari P, Safari F, Naderi A, Mottaghi K. Möbius syndrome, what anesthesiologist should consider about: a case report. JARSS 2020;28(3):213-6.

Kamran Mottaghi

Loghman Hakim Hospital,
Makhsous Street, Kargar Avenue
0098 Tehran - Iran

✉ k_mottaghi@sbmu.ac.ir

ORCID: 0000-0003-3371-1047

M. Nashibi 0000-0003-3825-9889

P. Sezari 0000-0003-3874-1050

F. Safari 0000-0002-1479-8477

Shahid Beheshti University of
Medical Sciences,
Tehran, Iran

A. Naderi 0000-0003-1545-4649

Jam Hospital,
Tehran, Iran



INTRODUCTION

German ophthalmologist Albrecht Von Graefe in 1880 first described the concurrence of paralysis in horizontal rotator muscles of one or both eyes with unilateral or bilateral paralysis of facial muscles and Harlem (1881) and Chisholm (1882) referred to it. Firstly full description and classification of MS was done by German neurologist Paul Julius Möbius in 1888 and 1892 after reporting 44 cases. He called it nuclear atrophy. Since then it is called Möbius syndrome ⁽¹⁾. Because of the rare prevalence of the disease, many of the physicians may not be familiar with the course of the disease and its potential hazards. Due to scarcity of cases, clinical trials are not available; hence, the only sources to guide the anesthesiologist are case reports.

CASE PRESENTATION

A 6-year-old 17 kg boy was referred to otolaryngology clinic due to hearing deficiency and scheduled for cochlear implantation. His parents and his two siblings have no hearing deficiency.

Since his birth, left ptosis, left-sided incomplete eyelid closure (Figure 1) and right-sided hemi-facial deviation (left-sided facial palsy) (Figure 2), left external auditory canal stenosis (Figure 3) have been observed. On 40th day postpartum, the infant was admitted to Neonatal Intensive Care Unit (NICU) following reported poor feeding and weakness. In further evaluation, bilateral vesicoureteral reflux and right cystic and dysplastic kidney was diagnosed. He had delayed neck holding, sitting and walking. When he was 7 months old, hearing deficiency was discovered and treated with hearing aid device which had no satisfying outcome. Based on clinical manifestations, Möbius syndrome was diagnosed.



Figure 1. Left sided incomplete eyelid closure.



Figure 2. Right sided hemi-facial deviation (left sided facial palsy)



Figure 3. Left external auditory canal stenosis

Brain CT Scan and MRI did not show any pathologic findings. Laboratory data reported leukocytosis ($12700 \mu\text{L}^{-1}$), and eosinophilia (6.5%). Workup did not conclude in any etiology. Strabismus and maxillofacial abnormalities were not present. In electrophysiologic studies, auditory brain stem response (ABR) was absent for both ears.

On arrival to operating room, patient was sedated with intravenous midazolam $30 \mu\text{kg}^{-1}$ and fentanyl $3 \mu\text{g kg}^{-1}$. Anesthesia induced using 2mg kg^{-1} propofol and atracurium 0.5mg kg^{-1} . Mask ventilation was not

difficult and laryngoscopy was done using Macintosh blade size ⁽²⁾. Cormack-Leyhane grade was IIA and patient was intubated using spiral endotracheal tube with internal diameter of 4.5mm. Anesthesia maintained with propofol 150 µg kg⁻¹ min⁻¹, O₂ and N₂O (FiO₂: 40%). Seventh cranial nerve was monitored during surgery so we avoided using neuromuscular blocking drugs (NMBDs). Since the patient had strabismus we avoided succinylcholine and volatile anesthetics due to the possibility of Malignant Hyperthermia (MH).

Surgery lasted 3 hours and was uneventful, patient transferred to the recovery room and then discharged.

Informed consent for publication was obtained from the patient's guardian.

DISCUSSION

Möebius syndrome is a congenital disorder with some possible etiologies such as abnormal fetal position which exerts pressure on fetal brain that may lead to altered blood supply to cranial nerve centers ⁽²⁾. Some other authors believe that exposure to teratogens (benzodiazepines, misoprostol, cocaine, alcohol, hyperthermia, hypoxia and rubella) during first trimester of pregnancy could play a role in etiology ^(3,4). Its incidence is about 2 in 10⁷ to 2 in 10⁵ ⁽⁵⁾ with a female to male ratio of 1:1 ⁽⁶⁾.

The clinical findings may include unilateral or bilateral facial paralysis, defects of extraocular muscles movements which is due to paralysis of cranial nerves VI and VII and central nervous system involvement, orofacial abnormalities, muscular hypotonia, orthopedic abnormalities (club foot) ⁽⁷⁾, gastroesophageal regurgitation, vertebral anomalies, cerebellar hypoplasia, tracheo-esophageal fistula ⁽⁸⁾, dysarthria and difficulty in chewing, swallowing and coughing which may lead to aspiration and respiratory complications ⁽⁹⁾. Therefore, administration of antisialogogues before emergency intervention may decrease the likelihood of potential aspiration and its consequences ⁽¹⁰⁾. Cardiac and musculoskeletal abnormalities may also co-exist ⁽¹¹⁾.

Like other rare syndromes it may be a burden to

anesthesiologists concerning airway management. Micrognathia, retrognathia, mandibular hypoplasia and cleft palate have been reported in patients with Möebius syndrome and in a substantial number of these patients, anesthesiologists may encounter a difficult or failed airway ⁽¹²⁾; on the other hand, cases of day care management or use of an laryngeal mask airway (LMA) has been reported as well ⁽¹³⁾. In Möebius syndrome patients, 90.5% of tracheal intubations are performed easily. In 8.3% difficulty is encountered during intubating the patients and in 1.2% of cases intubation failure occurred, with the orofacial abnormalities as the possible factor related to difficult intubation ⁽¹¹⁾.

CONCLUSION

All things together, since the prevalence of Möebius syndrome is low, original articles and reviews are scarce in number. Airway management in these patients could be a challenge for anesthesiologists and an equipped facility must be prepared before patient admission in operating theatre. Several plans, various airway devices and expert specialists are required in facilities where surgery is performed. Anesthesiologists must be prepared for potential Malignant Hyperthermia ⁽⁷⁾. Hence, avoiding triggering factors and utilizing a volatile-free anesthetic delivery system are highly recommended. Since aspiration is an important complication among these patients, antisialogogues as premedication or before emergency intervention is highly recommended.

Conflict of Interest: No potential conflict of interest relevant to this article was reported.

Funding: Authors declare that the article was prepared without any funding and financial support.

Informed Consent: Obtained.

REFERENCES

1. Stansbury JR. Moebius's Syndrome: Congenital Oculofacial Paralysis: A Case Report. *Am J Ophthalmol.* 1952;35:256-61. [https://doi.org/10.1016/0002-9394\(52\)90857-X](https://doi.org/10.1016/0002-9394(52)90857-X)
2. Morales-Chavez M, Ortiz-Rincones MA, Suarez-Gorriñ F. Surgical techniques for smile restoration in patients with Mobius syndrome. *J Clin Exp Dent.* 2013;5:e203-7. <https://doi.org/10.4317/jced.51116>

3. Graziadio C, Lorenzen MB, Rosa RF, et al. New report of a familial case of Moebius syndrome presenting skeletal findings. *Am J Med Genet A*. 2010;152A:2134-8. <https://doi.org/10.1002/ajmg.a.33548>
4. Lima LM, Diniz MB, dos Santos-Pinto L. Moebius syndrome: clinical manifestations in a pediatric patient. *Pediatr Dent*. 2009;31:289-93.
5. Briegel W, Schimek M, Kamp-Becker I, Hofmann C, Schwab KO. Autism spectrum disorders in children and adolescents with Moebius sequence. *Eur Child Adolesc Psychiatry*. 2009;18:515-9. <https://doi.org/10.1007/s00787-009-0003-1>
6. Briegel W. Neuropsychiatric findings of Moebius sequence -- a review. *Clin Genet*. 2006;70:91-7. <https://doi.org/10.1111/j.1399-0004.2006.00649.x>
7. Fernandes CR, Pinto Filho WA, Cezar LC, Alves Gomes JM, Florencio da Cunha GK. Fatal recrudescence of malignant hyperthermia in an infant with Moebius syndrome. *Braz J Anesthesiol*. 2013;63:296-300. [https://doi.org/10.1016/S0034-7094\(13\)70234-4](https://doi.org/10.1016/S0034-7094(13)70234-4)
8. Cote G, Bouchard S. Hepatotoxicity after desflurane anesthesia in a 15-month-old child with Moebius syndrome after previous exposure to isoflurane. *Anesthesiology*. 2007;107:843-5. <https://doi.org/10.1097/01.anes.0000287196.15665.16>
9. Krajcirik WJ, Azar I, Opperman S, Lear E. Anesthetic management of a patient with Moebius syndrome. *Anesth Analg*. 1985;64:371-2. <https://doi.org/10.1213/00000539-198503000-00013>
10. Gondipalli P, Tobias JD. Anesthetic implications of Moebius syndrome. *J Clin Anesth*. 2006;18:55-9. <https://doi.org/10.1016/j.jclinane.2005.05.005>
11. Ames WA, Shichor TM, Speakman M, Zuker RM, McCaul C. Anesthetic management of children with Moebius sequence. *Can J Anaesth*. 2005;52:837-44. <https://doi.org/10.1007/BF03021779>
12. Ferguson S. Moebius syndrome: a review of the anesthetic implications. *Paediatr Anaesth*. 1996;6:51-6. <https://doi.org/10.1111/j.1460-9592.1996.tb00354.x>
13. Hobaika AB, Neves BS, Fernandes ML, Guedes VC. Anesthesia in a patient with moebius sequence: case report. *Rev Bras Anesthesiol*. 2009;59:341-3. <https://doi.org/10.1590/S0034-70942009000300009>