Indian Journal of Pharmacy Practice., 2024; 17(2):119-121.
https://www.ijopp.org

Review Article

Unveiling Moebius Syndrome: Understanding the Rare Neurological Condition

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ABSTRACT

Moebius Syndrome is a rare congenital neurological disorder that affects the sixth and seventh cranial nerves, leading to facial paralysis, impaired eye movement and other associated symptoms. The exact etiology of Moebius Syndrome is not fully understood, but genetic factors, exposure to toxins during fetal development, vascular anomalies or acquired ischemia events have been proposed as potential contributing factors. Clinical manifestations include unilateral or bilateral nonprogressive congenital facial palsy, deficiencies in ocular abduction and limb abnormalities, among other cranial nerve palsies. It is primarily clinical, as there are no specific laboratory tests available for evaluation. Pharmacological management for Moebius Syndrome is limited, and there is no specific pharmaceutical treatment available. Multivitamins may be added to the treatment regimen. Surgical methods, such as the "smile operation," temporalis tendon transfer and bilateral selective neurolysis can be used to correct lagophthalmos and strabismus. Children with Moebius Syndrome can live almost normal lives with the support of a multidisciplinary team. Moebius Syndrome presents a unique set of challenges due to its rarity and the diverse range of symptoms it encompasses. Understanding its pathogenesis, clinical signs, diagnosis, and management is crucial for early diagnosis, intervention, and management to improve the quality of life for individuals affected by this condition.

Keywords: Congenital Neurological Condition, Cranial Nerve Dysfunction, Moebius Syndrome, Seventh Cranial Nerve, Sixth Cranial Nerve.

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Received: 24-01-2024;
Revised: 15-02-2024;
Accepted: 05-03-2024.

INTRODUCTION

A rare congenital neurological condition called Moebius syndrome is characterized by the weakening or palsy of several cranial nerves, usually the sixth and seventh cranial nerves. It is a nonprogressive condition that can result in a variety of symptoms such as facial paralysis, newborns that have issues with feeding and weight gain, as well as trouble moving their eyes. Limb abnormalities and various neurological and developmental problems are also associated with the Moebius syndrome. Most diagnoses are made clinically, and there isn't a particular laboratory test available for evaluation. Therapy is supportive and may involve speech, physical, or occupational therapy.[1,2] Figure 1 indicates the facial nerve plasy in a full term boy at 3 days of age at rest and on crying.

Epidemiology

The epidemiology of Moebius Syndrome is difficult to quantify due to its rarity and the multiple factors that influence data gathering. The prevalence of Moebius Syndrome has been found to be extremely low, with estimates ranging from 0.06 to 0.3 per 100,000 live births. Moebius Syndrome is regarded as a very uncommon condition, with sporadic instances and no gender or regional predilection detected.[4]

Etiology

Although the exact origin of Moebius Syndrome is unknown, genetic factors are thought to have a role. Certain genes implicated in the formation of the cranial nerves may be mutated, according to some studies. Additional theories include exposure to toxins during fetal development, vascular anomalies throughout development, or an acquired ischemia event that happens after the fifth week of pregnancy. One or more localized sites of injury may be involved in the brainstem, where the neurons of the face, abducens, and lacrimal (salivary) nuclei are physically coincident during this stage of development, has been proposed as the initial insult initiating a series of events. To completely understand the pathogenesis of Moebius Syndrome, more research is necessary.[5,6]

DOI: 10.5530/ijopp.17.2.19

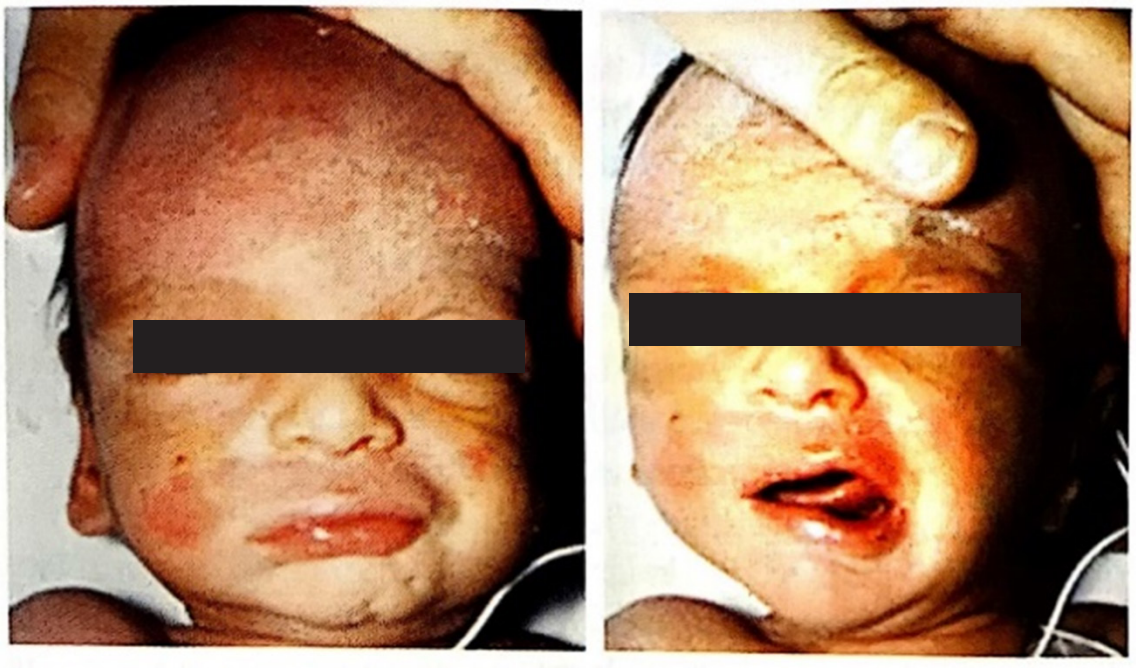

Figure 1: Facial Paralysis. Facial nerve plasy in a full-term boy at 3 days of age, at rest and on crying.[3]

Risk Factors

While the exact cause of Moebius Syndrome's malfunctioning cranial nerves is unknown, certain medications or illicit drug used during pregnancy are risk factors that may contribute to the illness.[7]

Pathophysiology

The VI and VII cranial nerves responsible for controlling face muscles are underdeveloped, which is part of the pathophysiology of Moebius syndrome. This underdevelopment results in facial muscle paralysis, trouble speaking and eating, and the inability to use facial gestures to convey emotions. It is unclear what specifically causes these cranial nerves to be underdeveloped.[8]

Clinical Manifestations

Unilateral or bilateral nonprogressive congenital facial palsy (VII cranial nerve) accompanied by deficiencies in ocular abduction (VI cranial nerve) are the hallmarks of Moebius syndrome.

People who have Moebius Syndrome experience weakness or paralysis of the muscles in their faces, which can impair their ability to create other facial emotions like a frown or a grin.

Additionally, they could find it difficult to move their eyes side to side.

Moebius Syndrome may also be linked to limb abnormalities, orofacial malformations, and other Cranial Nerve (CN) palsies.

Additional symptoms include speech and feeding difficulties, hearing loss, and developmental delays can be present in some Moebius Syndrome sufferers.[9]

Diagnosis

As per the International Group of Experts at the Moebius Syndrome Foundation, the diagnostic criteria for Moebius syndrome are as follows:

Congenital facial diplegia or uniplegia, involving a lower motor neuron type; and

Paralysis of lateral eye movements and strabismus resulting from sixth cranial nerve palsy.

The purpose of developing these criteria in 2007 was to encourage consistency in diagnosis. But the majority of diagnoses are made clinically, and there isn't a specific laboratory test available for diagnosis.[10,11] Figure 2 indicates a computed tomography scan which shows symmetrical dorsal pontine calcifications.

Management

Pharmacological Management

For Moebius syndrome, there is no specific pharmaceutical treatment. Multivitamins were added, though.

Non-pharmacological Management

The non-pharmacological treatment of Moebius syndrome involves occupational, speech, and physical therapy to improve motor control, speech, and eating habits; special attention to the eyes to prevent exposure keratitis; and feeding assistance and careful nutritional management to ensure adequate postnatal weight gain.

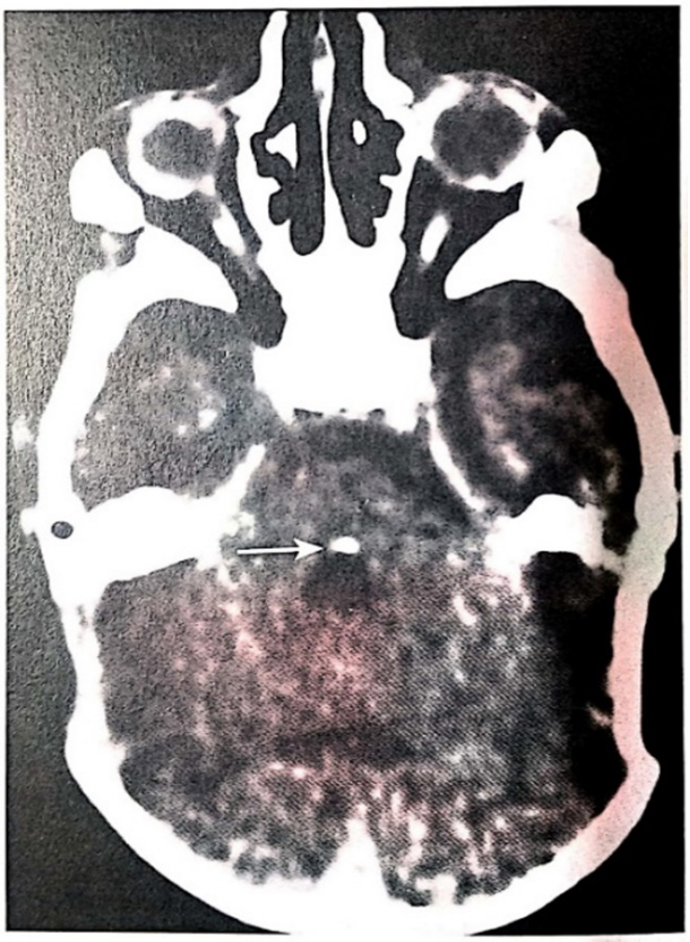

Figure 2: Moebius syndrome. This computed tomography scan shows symmetrical dorsal pontine calcifications (arrow).[12]

Surgical methods

It includes the "smile operation," temporalis tendon transfer, and bilateral selective neurolysis can be used to correct lagophthalmos and strabismus.

Children with Moebius syndrome can live almost normal lives with the support of a multidisciplinary team that includes skilled physicians.[13,14]

CONCLUSION

A rare neurological condition called Moebius Syndrome mainly affects the muscles of the face and the movement of the eyes. Based on the presence of facial paralysis and the incapacity to move the eyes laterally, it is usually diagnosed at birth or in the early stages of infancy. Although Moebius Syndrome's precise cause is unknown, aberrant cranial nerve development is thought to be a contributing factor. Moebius Syndrome presently has no known cure; instead, care aims to manage symptoms and enhance quality of life.

A multidisciplinary team of medical specialists may be involved in this, and services such as feeding assistance, physical therapy, speech and language therapy, eye care, dental care, and psychological support may be provided. Although Moebius Syndrome can significantly impair a person's quality of life, symptoms can be managed, and results can be improved with an early diagnosis and intervention.

ACKNOWLEDGEMENT

The authors are grateful to express gratitude to College of Pharmaceutical Sciences, Government Medical College, Thiruvananthapuram for providing access to resources and facilities essential for conducting this review.

CONFLICT OF INTEREST

The authors declare that there is no conflict of interest.

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Cite this article: Sarath D, Jayakrishnan SS, Sajitha G, Sajna SJ. Unveiling Moebius Syndrome: Understanding the Rare Neurological Condition. Indian J Pharmacy Practice. 2024;17(2):119-21.