

Multidisciplinary Behavioral Management of Anomalies in Moebius Syndrome

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Abstract

Introduction: Moebius syndrome presents the following alterations: facial paralysis, ocular abduction, superior ankyloglossia and loss of integrity of the palatal platform among other alterations.

Objective: To systematically review the multidisciplinary behavioral management of Moebius Syndrome anomalies from 2019 to 2023 for the purpose of integrating a guiding protocol.

Methodology: A systematic review was carried out considering the prism method. The following databases were used: Web of Science, Scopus and Scielo.

Results: A total of 25 articles were selected according to the systematization process where it is evident that 40% deal with facial paralysis and the most frequent area is Surgery.

Conclusion: Early diagnosis is essential for adequate management of the syndrome. It is necessary to address the facial paralysis, as well as the other alterations. It is recommended to provide emotional support to the patient and caregivers.

Keywords: *facial paralysis, multidisciplinary, Moebius syndrome.*

INTRODUCTION

Julius Moebius discovered Moebius Syndrome (MS) in 1888. The main factor of this syndrome is not known, but the most common cause is miscarriage. This results in the absence of somatic sensory core formation and calcium deposits in the body's tissues. (1) In addition to limited mouth opening, breathing disorder, and heart disease. It was also found to be related to the intake of medications such as sedatives, alcoholic beverages, drugs, as a result of abnormal embryonic maturation. Comments (2)

Therefore, the recorded data show that mothers of children with MS during the gestational period have a significant link with misoprostol and thalidomide. (3) Although teratogens are associated with a higher prevalence of autism, the distinguishing feature is the poor movement of the muscles of the face without being able to smile or wrinkle the forehead. They are generally perceived as unintelligent and sociable. However, they have

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average intelligence, but they do not express emotions because of their condition. Comments (4)

For this reason, it is called a "rare disease", because it is found in few people. Experts estimate 5 in 50,000 to 1 in 500,000 newborns with MS. A close relationship was found with non-progressive, symmetrical or asymmetrical facial paralysis and impaired ocular abduction. (5) This is due to weakness of the facial nerves VII and VI. According to neuropathological and neuroradiological evidence, there is a maldevelopment of the hindbrain that causes hypoxia and causes dysfunction of the cranial nerves. Comments (6)

MS involves the sixth cranial nerve, which is responsible for controlling the mobility of the eyeballs, in turn including other nerve structures causing alterations at the level of the eyes, mouth and ears. In addition, it hinders the expressiveness in their face, limiting themselves to expressing their state of mind. As a result, their emotions and feelings are not known, which affects socialization among their peers. Comments (7)

The study by Milone et al. (8) He verified through functional thermography how facial immobility decreases the intensity of emotional perception. Conduct disorders are closely related to affective involvement and lack of emotions.

It is not yet understood what the main multidisciplinary conflicts with Moebius Syndrome would be. However, they experience social isolation, frustration, behavioral problems, and reduced virtual networks affect their quality of life as they grow up. (9) On the other hand, there are studies that show that parents with MS reflect more social and emotional problems than the general population. They are often compared to people with cognitive difficulties. Comments (10)

This syndrome consists of the delimited way in which they interact in social situations, leading to misunderstandings and awkward situations. Facial paralysis does not allow them to express their emotions openly because of the immobilization it presents. This study will help to compile the different management recorded in different areas. (1,2,3,4)

Finally, a multidisciplinary behavioral management protocol can guide parents to better understand treatments. Likewise, the strategies found will be effective to be used in their daily lives. Such patients will be able to reduce behavioral problems and anxiety. In addition, it will contribute to future reviews on the subject. (6,7)

The following question was posed: What is the protocol in multidisciplinary behavioral management in Moebius Syndrome between the years 2019 to 2023? Therefore, the objective is to systematically review the multidisciplinary behavioral management in Moebius Syndrome between 2019 and 2023 in order to integrate a guiding protocol.

METHODOLOGY

Type of study

An exhaustive study of scientific articles was carried out based on the PRISMA method(11)

The questions established to initiate the methodological process were the following:

What abnormalities are most common in Möbius syndrome?

What is the multidisciplinary behavioral management in patients with Moebius syndrome?

What is multidisciplinary behavioral management in patients with Moebius syndrome?

What are the most common medical areas?

Search Process

To carry out the systematic search and ensure the results, the following descriptors were selected: Moebius syndrome, Moebius Syndrome, behavior, behavior. The selected languages were Spanish, English and Portuguese. To detail the results, search combinations were designed with the Boolean terms and operators: Moebius syndrome OR Moebius syndrome. AND behavior OR behavior.

Inclusion and exclusion criteria

We included research published in scientific databases in Portuguese, English and Spanish. Keywords were taken into account in order to obtain adequate information. The year of publication was considered from 2019 to 2023. The search was conducted from March to June 2023. Articles that have open access. On the other hand, articles that do not correspond to the year of publication were also excluded. Languages other than English, Spanish, and Portuguese. Books, journals, these were also not considered. In the Web of science, Scopus and Scielo databases, research articles were considered as shown in Figure 1.

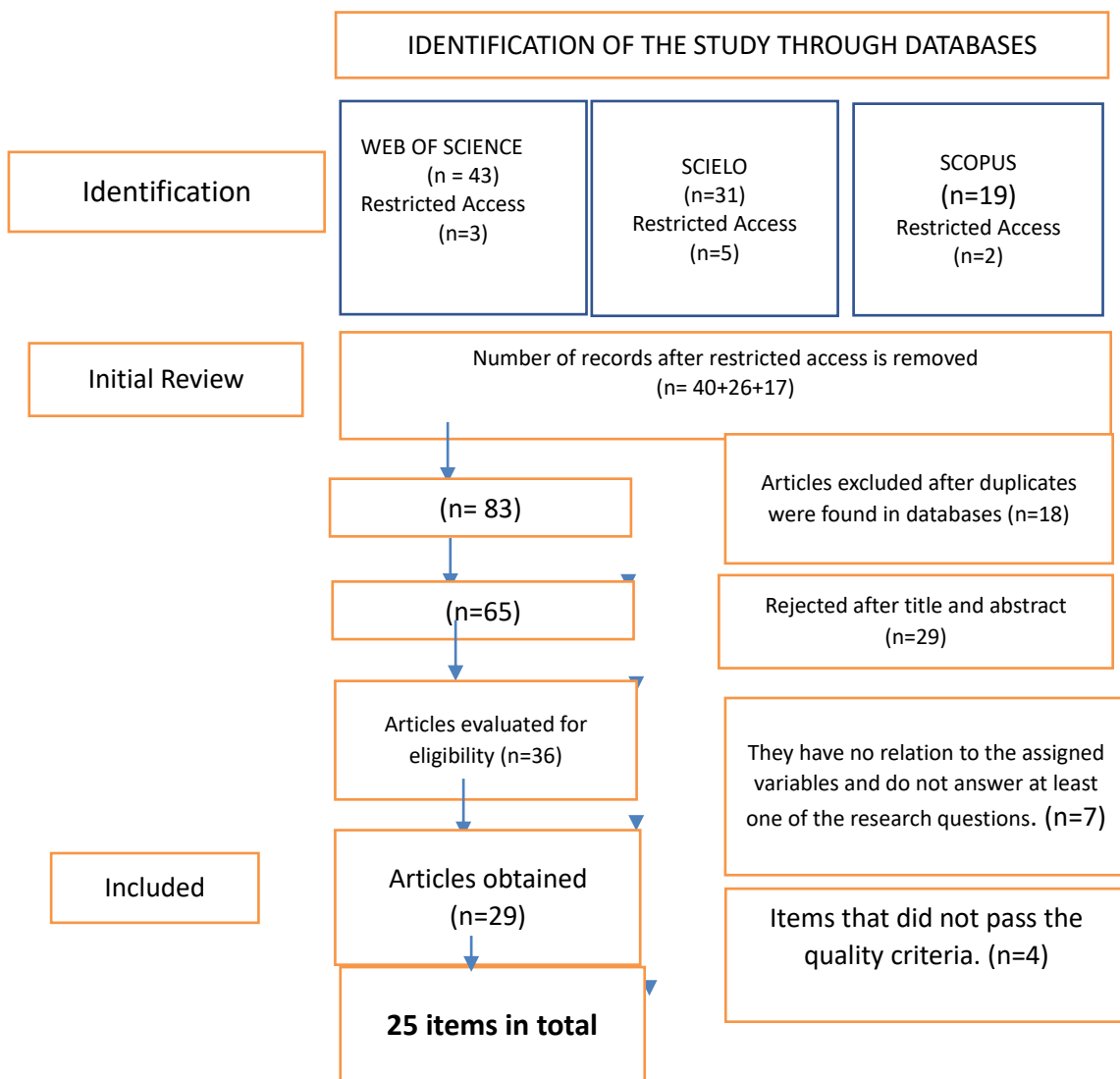


Figure 1. Selection process of articles based on the unit of analysis.

RESULTS

Through the article search, 25 articles were selected for the presentation of results. As for the database used, the one with the highest number was Web of science (72%), followed by Scopus (24%) and Scielo (4%).

Articles by year of publication

Figure 2 shows that the largest number of articles were selected from the year 2020, representing (40%), followed by articles from the year 2022 with (28%). In addition, articles from 2019 and 2021 were selected with the same amount (16%).

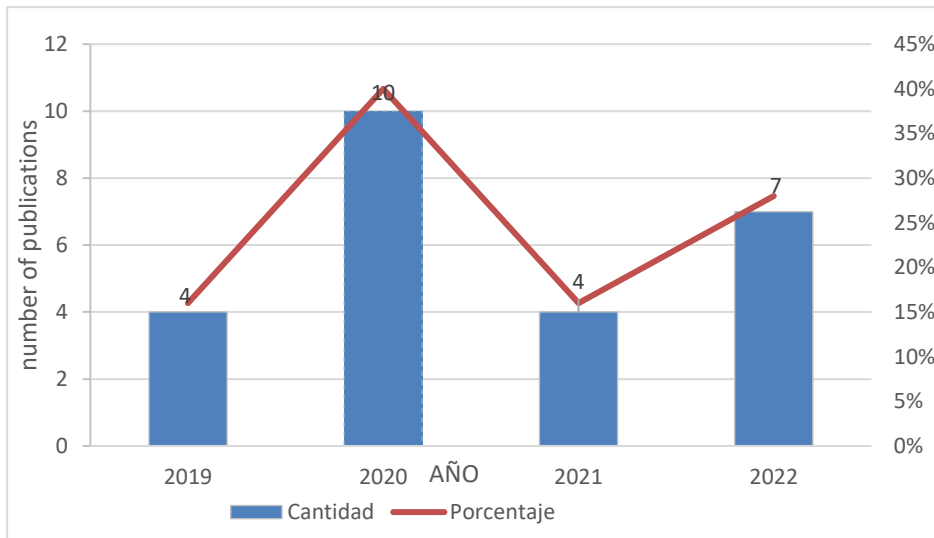


Figure 2. Sources by year of publication

Sources by place of origin

In Figure 3, the largest number of articles is from the United States (16%); it is followed by Brazil, Italy and Colombia (12%); Iran, Mexico and Poland (8%); Japan, Romania, Korea, France, Germany and Turkey have the lowest percentage, with one item each (4%). On the other hand, Europe and Asia found the highest number of articles (69.3%); in Latin America it was only found (30.7%).

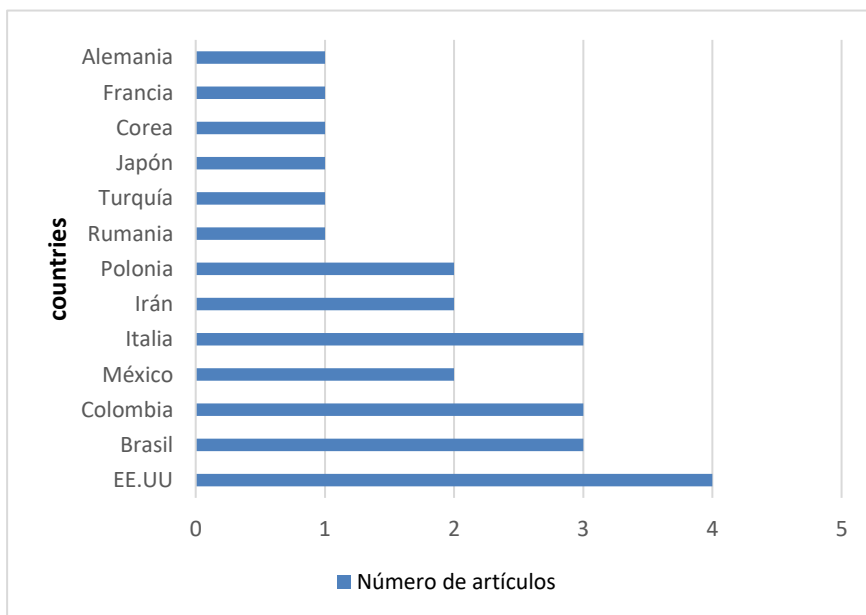


Figure 3. Sources by place of origin

Research Response

This unit shows the result in relation to the number of articles for each type of question. This made it possible to answer the questions raised.

P1. Multidisciplinary behavioral management in patients with Moebius syndrome is due to facial paralysis; dental malocclusions; breathing and feeding difficulties; hypoplasia of the left pectoralis major muscle and hand; ocular abduction of one or both eyes; superior ankyloglossia and loss of integrity of the palatal platform; delayed mental development, hypertrophic and dysmorphic features; flattening of the floor of the fourth ventricle secondary to a bilaterally absent facial colliculus; obstructive sleep apnea and sleep-related hypoventilation due to medical disorders; early childhood caries and impaired facial growth; bilateral hip dysplasia; abducten nerve palsy; strabismus, brainstem anomaly, growth retardation, impaired swallowing, pathway difficult area, malignant hyperthermia.

P2: The most common medical areas: Radiology and imaging, Pediatric Dentistry, Maxillofacial Surgery, Plastic Surgery.

Table 1 Response to the research.

Questions	Articles	Answer
What abnormalities are most common in Moebius Syndrome?	[1],[2],[7],[9],[16],[17],[19],[21],[22],[25]	Facial paralysis
	[3]	Dental malocclusions
	[4]	Breathing and feeding difficulties
	[5]	Hypoplasia of the left pectoralis major muscle and hand
	[6],[21],[22]	Ocular abduction of one or both eyes.
	[10]	Flattening of the floor of the fourth ventricle secondary to a bilaterally absent facial colliculus.
	[11]	Obstructive sleep apnea
	[8],[12],[14]	superior ankyloglossia and loss of palatal shelf integrity
	[15]	Bilateral hip dysplasia
	[18]	Strabismus
	[20]	Brain stem abnormality
	[24]	Malignant hypertemia
	[23]	Impaired swallowing and pathway difficult area
[13]	Early Childhood Cavities	
What is the multidisciplinary behavioral management in patients with Moebius Syndrome?	[2],[16],[7]	Maxillofacial Surgery
	[1],[19],[22]	Plastic surgery
	[3]	Orthodontics
	[4],[11]	Otolaryngologist
	[5],[19]	Pediatric Neurology
	[8],[13],[12],[14]	Pediatric dentistry
	[6],[10],[17],[8]	Radiology & Imaging

[15]	Pediatric Orthopedic Surgery
[17]	Neuroimaging
[9]	Biochemistry and Biophysics
[23],[24]	Anesthesiology
[21],[25]	Diagnostic
[18]	Ophthalmology
[20]	Urology

DISCUSSION AND CONCLUSION

In this systematic review, 25 study articles were selected where it is evident that early diagnosis is the key to starting treatment as soon as possible. (21,25) On the one hand, facial paralysis in the (MS) is a rare congenital disease. (1,2,7) It is characterized by the lack of development of cranial nerves VI and VII, (9,16,17) which modulate eye movements and facial muscles, respectively. (19,21) For this reason, the treatment of facial paralysis is a top priority. (1,2,7,9,16,17,19,21,22,25) Additionally, it connects with emotional well-being. (21,22,25). The Surgery area treats the lack of facial expression and the inability to smile, (1,2,7,16) through grafts. (4,19,22,23,25). These treatments are carried out with the purpose of improving the social aspect, such as: interaction with peers and their social environment. On the other hand, they found ocular abduction that does not allow them to move their eyes sideways. (6,21,22) Other anomalies related to this syndrome included hypoplasia of the left pectoralis major muscle and hand, flattening of the floor of the fourth ventricle secondary to a bilaterally absent facial colliculus, obstructive sleep apnea, bilateral hip dysplasia, strabismus, brainstem anomaly, and malignant hyperthermia. (5,10,11,15,18,20,24) Also, in the oral cavity there are alterations, such as upper ankyloglossia, dental caries, malocclusion, loss of integrity of the palatal platform in minors, so the intervention of the pediatric dentist is opportune. (3,8,12,14,13) Likewise, in the area of anesthesiology, there is difficulty at the time of endotracheal intubation due to impaired swallowing and rigid airway. (23,24) To perform these procedures, the auxiliary examinations referred to the area of Radiology and Imaging were magnetic resonance imaging, tomography, and neuroimaging. (6,10,17,18) In this regard, it is important to know that this syndrome involves different areas of the body, so it requires multidisciplinary management. This study was limited to the period 2019 to 2023, the information obtained did not take into account other medical areas such as Cardiology, Psychology. From the search in Scielo only one article remained, this is due to using the same search syntax.

In conclusion, early diagnosis of Moebius syndrome is indispensable for proper management of the condition. Likewise, early multidisciplinary intervention to correct the different alterations is essential for the general well-being of the patient. In these cases, a support group is recommended to promote inclusion and understanding in the community.

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