

Electromyographic evaluation of the facial and motor donor nerves before facial reanimation surgery in Moebius syndrome

Evaluación electromiográfica del nervio facial y de nervios donadores previo a cirugía de reanimación facial en pacientes con síndrome de Moebius

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Abstract

Background: Moebius syndrome is a clinical entity characterized by bilateral facial and abducens nerve palsies; other cranial nerves might be affected as well. So far, no studies have reported the electromyographic responses of the facial musculature in this group of patients. **Objective:** The objective of our study is to describe the electromyographic responses of the facial muscle and the main donor nerves for facial reanimation in patients with Moebius syndrome. **Methods:** We analyzed electromyographies from the facial, hypoglossal, masseterine (trigeminal), and accessory nerves from patients with Moebius syndrome treated between 2010 and 2016. Results are presented as percentages and central tendency measures. **Results:** Twenty-four patients were included, mean age 11.79 ± 9.39 years. The facial nerve showed complete unilateral recruitment in four patients, partial bilaterally in 11, seven showed no activity bilaterally, and two had unilateral inactivity. The masseterine was normal in 14 patients, which had partial recruitment bilaterally in four and unilaterally in four cases. The accessory nerve was normal in 20 patients, which showed partial recruitment bilaterally in three and unilaterally in one patient. The hypoglossal was normal in 22 patients and altered in two cases. **Conclusions:** Patients with Moebius syndrome show several degrees of alteration in electromyographic evaluation of the facial nerve. Electromyography is a useful tool in evaluating potential motor donor nerves for facial reanimation surgery.

Key words: Electromyography. Facial nerve. Neurophysiology. Facial paralysis. Moebius syndrome.

Resumen

Antecedentes: Las alteraciones en el perfil de lípidos son parte de la respuesta de fase aguda, lo que corresponde a la denominada lipemia de la sepsis. **Objetivo:** Determinar si la concentración sérica de lipoproteínas de alta densidad (HDL) se relaciona con la gravedad y la mortalidad. **Método:** Estudio retrospectivo, descriptivo, transversal, de pacientes con diagnóstico de sepsis abdominal, durante el periodo de abril de 2016 a febrero de 2017. Se determinó la gravedad mediante APACHE II, SOFA, Mannheim, CONUT, la presencia de fallas orgánicas y la mortalidad. **Resultados:** Se incluyeron 154 casos, 59 mujeres y 95 hombres. El principal órgano causante de sepsis abdominal fue el apéndice (41.6%). La mortalidad global fue del 14.3%. La presencia de fallas orgánicas fue del 35.1%. El valor medio de HDL se situó en 37.64 mg/dl (desviación estándar:

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± 16.16). Los hallazgos, sometidos a verificación estadística mediante la prueba *t* de Student, mostraron significancia entre los casos con SOFA > 4 ($p = 0.01$) y Mannheim > 26 ($p = 0.001$), CONUT > 6 ($p = 0.001$), presencia de fallas orgánicas ($p = 0.001$) y mortalidad ($p = 0.003$). **Conclusión:** Los valores de HDL se relacionan con la gravedad, con el desarrollo de fallas orgánicas y con la mortalidad en la sepsis.

Palabras clave: Gravedad. Lipoproteínas de alta densidad. Mortalidad. Sepsis.

Introduction

Moebius syndrome is a rare disease of unknown etiology, characterized by congenital bilateral paralysis of the facial and abducens nerves. Approximately 300 cases have been reported in the international literature and it is estimated to have an incidence of one case per 50,000 live births¹. Our research group published the largest patient series to date, including 115 who were treated throughout 5 years².

In addition to the presence of paralysis of the sixth and seventh cranial nerves, other cranial nerves may be paralyzed, with the 12th, 10th, 9th, 3rd, 8th, 5th, 4th, and 11th nerves being the most affected, in decreasing order. Clinical manifestations often go unnoticed at birth, and therefore, it is usually diagnosed during the breastfeeding period when feeding problems, such as difficulty sucking or swallowing, sialorrhea, and absence of facial movements when crying, are found¹.

Etiology of the syndrome is so far unknown. The main theories correspond to vascular or genetic alterations, with potential toxic factors associated. The vascular theory proposes that the syndrome is caused by disruption of the basilar or trigeminal arteries during embryonic development, which generates areas of ischemia at the central level, which would also explain the development of dysfunction in other cranial nerves³. The genetic theory is based on reports of chromosomal alterations in some patients, such as chromosome 13 deletion or a 1:11 translocation; in other cases, an autosomal dominant, recessive, or X-linked inheritance have been reported⁴. Due to this heterogeneity, determining a genetic cause for the condition has not been able. Prenatal events such as exposure to thalidomide, misoprostol, cocaine, ergotamine, and benzodiazepines have also been proposed in the pathogenesis of the syndrome⁵⁻⁷.

There are multiple associated malformations, especially those of the musculoskeletal system, such as brachydactyly, syndactyly or arthrogryposis, as well as the presence of other syndromes, such as Poland and Klippel-Feil syndrome. Septal ventricular defects, dextrocardia, and transposition of the great vessels

have been reported in the chest. Orofacial malformations include cleft palate, bifid uvula, microstomy, micrognathia, and velopharyngeal insufficiency due to 9th cranial nerve paralysis¹.

Clinical diagnosis is based on the findings on physical examination. According to the presentation of facial alterations, Terzis classified patients in three groups: complete Moebius if there is bilateral involvement of the 7th and 6th cranial nerves, incomplete Moebius if compromise of the 7th cranial nerve is unilateral, and Moebius-like if there is involvement of the 7th cranial nerve with another cranial nerve excluding the 6th. So far, there are no validated scales that assess facial nerve paralysis severity in patients with Moebius syndrome⁸.

Electrophysiological studies objectively evaluate facial nerve function and its target muscles, which allow determining the severity of its involvement, estimating its prognosis and guiding treatment⁹. Due to the rarity of the disease, there are only few studies evaluating facial muscles neurophysiology in the population with Moebius syndrome, and thus, available information is limited and, in many cases, originated in other populations with facial paralysis. So far, only Verzijl et al.¹⁰, Cattaneo et al.¹¹, and Jaradeh et al.¹² have reported electromyographic results from patients with Moebius syndrome, but studies have focused exclusively on the neurophysiological impact of the disease and potential donor nerves have not been assessed for pre-operative planning purposes.

The purpose of this work is to describe the electromyographic activity of the facial muscles and the main donor nerves for dynamic facial reconstruction in patients with Moebius syndrome cared for in our department.

Methods

Retrospective, observational, and cross-sectional study that included patients with Moebius syndrome cared for in our institution between 2010 and 2016. The syndrome was defined as the presence of non-progressive facial paralysis and ocular abduction

incapacity, present since birth. The patients were divided into three groups according to Terzis and Noah classification⁸:

- Classic Moebius: Facial and abducens bilateral paralysis.
- Incomplete Moebius: Abducens bilateral paralysis and facial paralysis with unilateral residual movement.
- Moebius-like: Unilateral facial paralysis accompanied by paralysis of some other cranial nerve.

Only patients with available electromyography of muscles innervated by the facial, masseter, hypoglossal and accessory nerves were evaluated. The electromyography tests were performed with bipolar needle electrodes placed on the muscles to be assessed. Registration points are placed on the frontal muscle, on the orbicular muscle of the eyes at the height of the lateral edge, on the zygomatic muscles, mouth orbicularis, and mentonian muscles, which are points that correspond to the temporal, zygomatic, and mandibular branches of the facial nerve^{9,13}. To assess donor nerves, electrodes were placed on the masseter nerves to evaluate the trigeminal masseter branch, on the trapezius to evaluate the spinal or on the tongue to evaluate hypoglossal nerve function^{9,13}.

During muscle activity, the percentage of recruited motor units in comparison with the healthy or standard side is determined¹³, which is why grouping the results into three categories was decided, namely, complete, partial, or no recruitment.

The information was collected in a Microsoft Excel database, including age, gender, Moebius type according to Terzis and Noah classification⁸, and muscle activity on electromyography for each muscle group on both sides of the body.

Statistical analysis was performed using descriptive statistics of the patient demographic characteristics; continuous variables are presented as central tendency measurements, and categorical variables as percentages.

Results

During the study period, 115 patients with Moebius syndrome were cared for at the clinic. For the present study, 24 subjects (14 females and 10 males) who had electromyographs of the muscle groups of interest were included; mean age was 11.79 years (standard deviation: ± 9.39) (Table 1). Fifteen patients (62%)

Table 1. Demographic characteristics of the sample

	n	%
Gender		
Males	10	41.6
Females	14	58.3
	Mean	SD
Age	11.79	9.39
	n	%
Classification		
Classic Moebius	15	62
Incomplete Moebius	9	37
Moebius-like	0	0

SD: Standard deviation.

were classified as classic Moebius and nine (37%) as incomplete Moebius.

Facial nerve muscles showed complete unilateral recruitment in four patients (16.6%) and partial bilateral recruitment in 12 (50%); in five patients (20.8%), there was an absence of bilateral recruitment, and unilateral inactivity in three (12.5%). The masseter muscles had complete bilateral recruitment in 14 patients (58.3%), partial bilateral in six (25%), and partial unilateral in four (16.6%).

The trapezius muscle showed complete bilateral recruitment in 20 patients (83.3%), partial bilateral in three (12.5%), and partial unilateral in one (4%). The lingual muscle had normal bilateral activity in 22 patients (91.6%), partial bilateral in one (4%) and partial unilateral in another one (4%) (Table 2).

When analyzing the results according to the type of syndrome presented by the patients, among the 15 subjects with complete Moebius syndrome, bilateral partial recruitment in the facial muscles was observed in nine, total absence of recruitment in four, unilateral partial recruitment in one, and complete unilateral recruitment in one. In the nine patients with incomplete Moebius syndrome, there was partial bilateral recruitment in three, partial unilateral recruitment in two, complete unilateral in three, and absence of recruitment in one.

Discussion

Moebius syndrome encompasses a set of clinical malformations and dysfunctions that warrant multidisciplinary management. First described in 1880 by von Graefe and in 1888 by Paul Möbius, it is currently defined as the combination of facial and abducens nerves

Table 2. Electromyographic activity of the evaluated muscle groups

Muscle group	Complete bilateral	Complete unilateral	Partial bilateral	Partial unilateral	Null bilateral	Null unilateral
Facial (7 th)	-	4 (16.6%)	12 (50%)	-	5 (20.8%)	3 (12.5%)
Masseters (5 th)	14 (58.3%)	-	6 (25%)	4 (16.6%)	-	-
Trapezius (11 th)	20 (83.3%)	-	3 (12.5%)	1 (4%)	-	-
Lingual (12 th)	22 (91.6%)	-	1 (4%)	1 (4%)	-	-

paralysis at birth¹. Facial muscle function correction is important since in addition to restoring social smile, it improves language problems, oral incompetence, facial skeleton growth, and dental occlusion^{14,15}.

Every patient requires a multidisciplinary evaluation protocol implemented by specialists in pediatrics, ophthalmology, orthodontics, otorhinolaryngology, rehabilitation, medical genetics, and orthopedics. The procedure of choice today is microsurgical transfer of the gracilis muscle, but since there is no function of the facial nerve, extra-facial donor nerves must be used to neurotize the gracilis, with the masseteric, spinal accessory, and hypoglossal nerves being the most widely used. In our department, the masseteric nerve is the first choice, followed by spinal nerve, while hypoglossal is reserved for cases of anterior nerve paralysis since its use may be related to speech and swallowing difficulties¹⁴.

Donor nerve selection will depend on a thorough clinical examination, complemented by the results of an electromyographic evaluation. The literature related to the neurophysiological aspects of Moebius syndrome is limited; so far, only three studies on the subject have been published. The oldest study is that by Jaradeh et al.¹², who found a reduction in facial muscle activity in seven patients. In 2005, Verzijl et al.¹⁰ reported the results of six patients, with an absence of facial muscles bilateral activity being found in three (50%) and unilateral in the other three (50%); however, in the latter, only one side of the face was evaluated. In 2006, Cattaneo et al.¹¹ assessed 17 patients with electromyography and reported three (17.6%) without bilateral responses, four (23.5%) with normal bilateral activity, and ten (58.8%) with variable activity depending on the studied muscle; in addition, they assessed the activity of the masseters and found normal function in 94% of patients.

Compared with previous studies, our work shows interesting findings. Normal results were found in < 20% of patients, which is comparable to previous reports, as well as variations in muscle activity

ranging from partial reductions to absence of response in practically 80% of cases, which is slightly higher than in previous studies. In our study, the fact that it is the first to comprehensively evaluate the donor nerves that are most widely used in dynamic microsurgical reconstructions stands out, as well that it reports the state of the trapezius and lingual muscles, whose electromyographic results correspond to the degrees of involvement observed in other clinical series. Interestingly, in the Verzijl et al.¹⁰ series, the masseter muscles showed normal activity in 94% of cases, while in our series, this only occurred in 58%.

The present study has multiple strengths: it is the first to comprehensively assess electromyography results of facial muscles and donor nerves used in dynamic reconstruction, as well as the study with the largest sample size so far. Its disadvantages include the retrospective nature of the study, as well as the fact of limiting the reporting of results to three global categories for the entire face since the lower third is known to generally have residual activity, which might affect final assessments.

Conclusions

Electromyography is a useful resource in the pre-operative evaluation of patients with Moebius syndrome since it allows objectively assessing the function of facial expression muscles and donor candidate nerves for dynamic reanimation procedures.

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Conflicts of interest

The authors declare that there are no conflicts of interest.

Ethical disclosure

Protection of people and animals. The authors declare that no experiments were performed on humans or animals for this research.

Confidentiality of data. The authors declare that they have followed the protocols of their work center on the publication of patient data.

Right to privacy and informed consent. The authors have obtained informed consent from the patients and/or subjects referred to in the article. This document is in the possession of the corresponding author.

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