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ORIGINAL ARTICLE

Facial reanimation surgery in Möbius syndrome: Experience from 76 cases from a tertiary referral hospital in Latin America[☆]

Chirurgie de réanimation faciale au syndrome de Möbius : expérience de 76 cas d'un hôpital de référence tertiaire en Amérique latine

A. Cardenas-Mejia, D. Palafox^{*}

Plastic and Reconstructive Surgery Department, Hospital General "Dr. Manuel Gea González", Avenida Calzada de Tlalpan #4800, Col. Sección XVI, Delegación Tlalpan, 14080 Mexico City, Mexico

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KEYWORDS

Möbius syndrome;
Facial reanimation;
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Summary

Introduction. — Möbius syndrome is defined as a combined congenital bilateral facial and abducens nerve palsies. The main goal of treatment is to provide facial reanimation by means of a dynamic surgical procedure. The microvascular transfer of a free muscle transplant is the procedure of choice for facial animation in a child with facial paralysis.

Observation. — Between January 2008 and January 2017, 124 patients with the syndrome have been approached at our institution. Distribution according to Möbius Syndrome classification presents as follows: Complete Möbius syndrome ($n = 88$), Incomplete Möbius syndrome ($n = 28$), Möbius-Like syndrome ($n = 8$). Seventy-nine female and 45 male patients. Sixty-one percent have undergone a microsurgical procedure ($n = 76$), in all of them, a free gracilis flap transfer was performed.

Discussion. — Our proposed treatment protocol for complete Möbius syndrome is determined by the available donor nerves. We prefer to use the masseteric nerve as first choice, however, if this nerve is not available, then our second choice is the spinal accessory nerve. For this purpose, all patients have an electromyography performed preoperatively. Overall, dynamic facial reanimation obtained through the microvascular transfer of the gracilis muscle have proved to improve notoriously oral commissure excursion and speech intelligibility.

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^{*} Corresponding author.

E-mail address: palafoxdamian@hotmail.com (D. Palafox).

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MOTS CLÉS

Syndrome de Möbius ;
Réanimation faciale ;
Paralysie faciale

Conclusion. — The free gracilis flap transfer is a reproducible procedure for patients with Möbius syndrome. It is of utmost importance to select the best motor nerve possible, based on an individualized preoperative clinical and electromyographic evaluation. To our best knowledge, this is the largest series of patients with Möbius syndrome globally, treated at a single-institution.

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Résumé

Introduction. — Le syndrome de Möbius se caractérise par une paralysie bilatérale congénitale combinée du nerf facial et du nerf moteur oculaire externe. L'objectif principal du traitement est d'offrir une réanimation faciale au moyen d'une intervention chirurgicale dynamique. Le lambeau libre musculaire pur de gracilis est la méthode de choix pour la réanimation faciale chez un enfant atteint de paralysie.

Observation. — Entre janvier 2008 et janvier 2017, 124 patients avec le syndrome de Möbius ont été approchés dans notre institution médicale. La répartition selon la classification du syndrome se présente comme suit : syndrome de Möbius complet ($n = 88$), syndrome de Möbius incomplet ($n = 28$), syndrome de Möbius-Like ($n = 8$). Soixante-dix-neuf femmes et 45 patients hommes. Soixante et un pour cent ont subi une intervention microchirurgicale ($n = 76$), dans tous les cas, un lambeau libre musculaire pur de gracilis a été effectué.

Discussion. — Notre protocole de traitement proposé pour le syndrome de Möbius complet est déterminé par les nerfs disponibles des donneurs. Nous préférons utiliser le nerf masséterin en première intention, cependant, si ce nerf n'est pas disponible, alors notre deuxième intention est le nerf spinal accessoire. À cette fin, tous les patients ont une électromyographie effectuée préopératoire. La réanimation faciale dynamique obtenue grâce au lambeau libre musculaire pur de gracilis améliorer notablement la course de la commissure orale et l'intelligibilité de la parole.

Conclusion. — Le lambeau libre musculaire pur de gracilis est une procédure reproductible et reproductible pour les patients atteints du syndrome de Möbius. Il est de la plus haute importance de choisir le meilleur nerf moteur possible, sur la base d'une évaluation clinique et électromyographique préopératoire individualisée. À notre connaissance, c'est la plus grande série de patients atteints du syndrome de Möbius à l'échelle mondiale, traités de façon unicentrique.

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Introduction

Möbius syndrome is defined as a combined congenital bilateral facial and abducens nerve palsies [1]. Etiology is unknown, however, several theories have been proposed as the cause of the syndrome, such as the use of misoprostol during pregnancy [2]. Terzis et al. created a classification system which allows surgeons to identify precise cranial nerve deficiencies and establish a reconstructive surgery [3]. This is the most recognized classification globally and the one we use in our institution. Classic Möbius syndrome patients have complete bilateral facial and abducens nerve paralysis. In patients with incomplete Möbius syndrome, although they have the clinical picture of the syndrome, some residual motor function is encountered on one side of the face. Möbius-like patients, have unilateral facial paralysis, with additional cranial nerve palsies found [3]. Aside from treating other malformations present in these patients (limb and palate anomalies amongst others), the main goal of treatment is to provide facial reanimation by means of a dynamic surgical procedure. The microvascular transfer of a free muscle transplant is the procedure of choice for facial animation in a child with facial paralysis [4,5]. The

gracilis muscle is ideal as it is accessible, leaves no functional deficit and has reliable surgical anatomy [1]. The aim is to obtain a symmetric, spontaneous and powerful smile. Nonetheless, as experts have demonstrated, it has great importance to consider that one of the critical factors of this procedure is the selection of a motor nerve to innervate the transplanted muscle [5]. We herein present our experience from 76 cases surgically treated at our institution.

Observation

Between January 2008 and January 2017, 124 patients with Möbius syndrome have been approached at our Facial Paralysis and Peripheral Nerve Clinic, from the Plastic and Reconstructive Surgery Department at Hospital General "Dr. Manuel Gea González" in Mexico City. We retrospectively studied the charts and clinical files of all patients. Distribution according to Möbius Syndrome classification presents as follows: Complete Möbius syndrome ($n = 88$), Incomplete Möbius syndrome ($n = 28$), Möbius-Like syndrome ($n = 8$). Gender distribution is as follows: 79 female and 45 male patients. We further decided to classify patients according to age group < 4 years old ($n = 40$), 4–15 years old

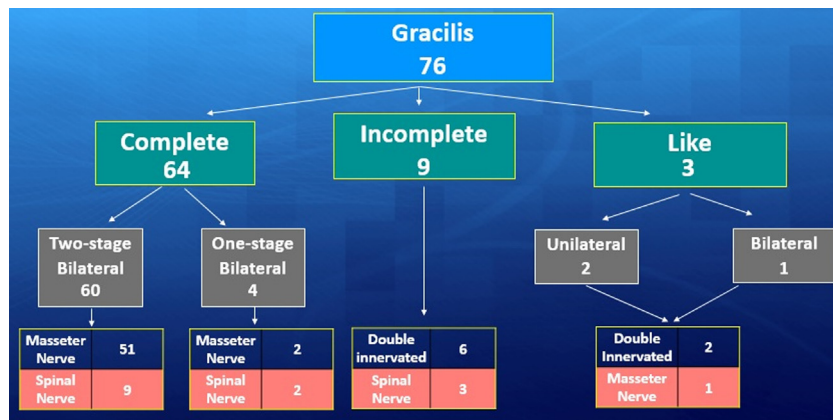


Figure 1 Description of the free gracilis flap transfers, according to Möbius syndrome type and donor nerve used for the procedure.



Figure 2 Male patient who underwent a bilateral free gracilis flap transfer as a one stage procedure. A. Preoperative condition. B. Postoperative condition.

($n = 63$), > 15 years old ($n = 21$). All patients have a thorough multidisciplinary clinical and electrophysiological examination before surgery consideration. From the cohort of patients, 61% have undergone a microsurgical procedure ($n = 76$). From the rest of the patients (39%), which have not undergone surgery, some are still awaiting for optimal conditions for the procedures. Some of these patients are still too young for the procedure and some others (most likely adult patients, have decided not to undergo surgery for personal reasons).

In all patients ($n = 76$), a free gracilis flap transfer was performed. Fig. 1 demonstrates the surgeries performed on the patients depending on the Möbius syndrome type. It also shows if a bilateral procedure (both hemifaces) was performed in one stage or most commonly, in two stages. The most common donor nerve used for facial reanimation is the masseteric nerve, however, in some cases we had to perform spinal nerve elongation. Also, in some patients with incomplete and möbius-like syndrome we performed a double innervated free functional muscle transfer. We encountered a total of 19 complications. Free flap re-exploration (causes associated with the microsurgical procedure itself) ($n = 8$), flap failure ($n = 6$), hematoma ($n = 1$), gracilis flap re-setting ($n = 4$). The patients with flap failure or flap loss were first treated with a contralateral gracilis flap transfer if the clinical conditions were stable and if authorization was provided by the patient and the family in cases of a minor, otherwise, patients with a microsurgical procedure failure were successfully treated with an orthodromic transfer of

the temporalis muscle. In 4 patients, we performed a bilateral free gracilis flap transfer in one stage (both hemifaces treated during the same procedure) (Fig. 2).

Discussion

Derived from our experience, our proposed treatment protocol for complete Möbius syndrome is determined by the available donor nerves [6]. For segmental gracilis reinnervation we prefer to use the masseteric nerve as first choice, however, if this nerve is not available, then our second choice is the spinal accessory nerve. For this purpose, all patients have an electromyography performed preoperatively (Fig. 3). Also, in cases of incomplete Möbius syndrome and möbius-like syndrome, our trend is to perform a double innervated free functional muscle transfer, as this procedure provides an additional axonal load to the paralyzed hemiface. We have published our results with the technique in the past [7]. We consider that the end-to-end cross face nerve graft to the obturator nerve coaptation carries the main impulse, creating a more symmetrical and spontaneous smile, in contrast, the end-to-end masseteric nerve to the obturator nerve coaptation provides an extra axonal load, thus, these patients have a more powerful smile although they might achieve less spontaneity [7]. Surgical Treatment protocol in patients with one side of the face affected, also considers availability of donor nerves supported by a pre-operative electromyography (Fig. 4). In our cohort of

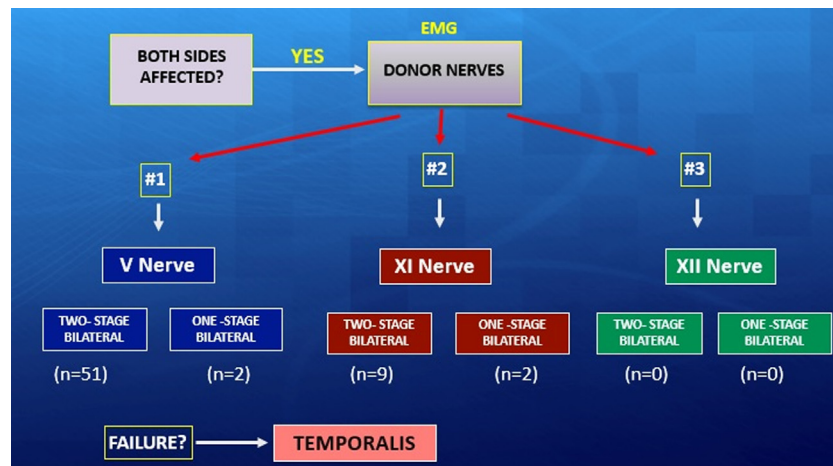


Figure 3 Surgical treatment protocol considering availability of donor nerves in patients with both sides of the face affected.

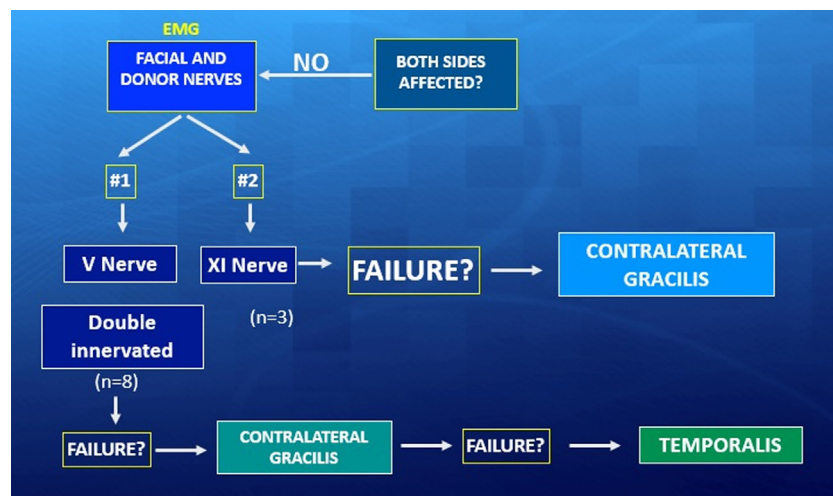


Figure 4 Surgical treatment protocol considering availability of donor nerves in patients with one side of the face affected.

patients, we have encountered that 7.5% have an abnormality, compromise or involvement of the trigeminal nerve in the electromyography. This is the main reason we had to use the spinal accessory nerve in some patients. Inspired by the results obtained by Dr. Chuang and his team, we decided to perform one stage facial reanimation surgery with the transfer of 2 gracilis flaps within the same procedure [8]. We performed surgery in 2 female and 2 male patients for a total of 8 free flaps. According to the Chuang scale, we obtained good results (grades II and III) without synkinesis. As for the patients with a microsurgical procedure failure, we performed an orthodromic transfer of the temporalis muscle, as described by Dr. Viterbo. Acceptable results are found in the overall appearance of the face, particularly in rest, nonetheless smile excursion is inferior when compared to the one achieved by a free flap transfer, despite this last limitation, we have found the transfer of the temporalis muscle as a reliable salvage surgery.

Overall, dynamic facial reanimation obtained through the microvascular transfer of the gracilis muscle have proved to improve notoriously oral commissure excursion and speech intelligibility [9]. Recently, researchers have demonstrated that the natural contraction of the masseter muscle during

normal smile production helps to explain the high rate of spontaneous smile development in patients with facial paralysis who have undergone a microsurgical reanimation procedure [10]. In fact, it has been proposed that cortical plasticity mechanisms play an important role in the restoration of smiling after these type of procedures [11,12]. At our institution, all patients who undergo a facial reanimation surgery receive biofeedback therapy in order to improve both coordination and facial expression. Patients who underwent surgery using the spinal nerve, begin with exercises based on shoulder elevation with resistance. We have observed that with time and with therapy, patients ultimately learn how to smile without having to activate the muscles innervated by the spinal accessory nerve. In patients who undergo surgery using the masseteric branch from the trigeminal nerve, therapy is quite easier as the exercises involve having to "bite" (masseter activation) with the result of a powerful immediate smile with good excursion (Fig. 5). They all also receive follow up by a multidisciplinary team, with specialists from Rehabilitation, Speech Therapists, Orthodontics amongst others.

It is quite satisfactory that our patients not only do they obtain an improvement in speech and feeding, but also have

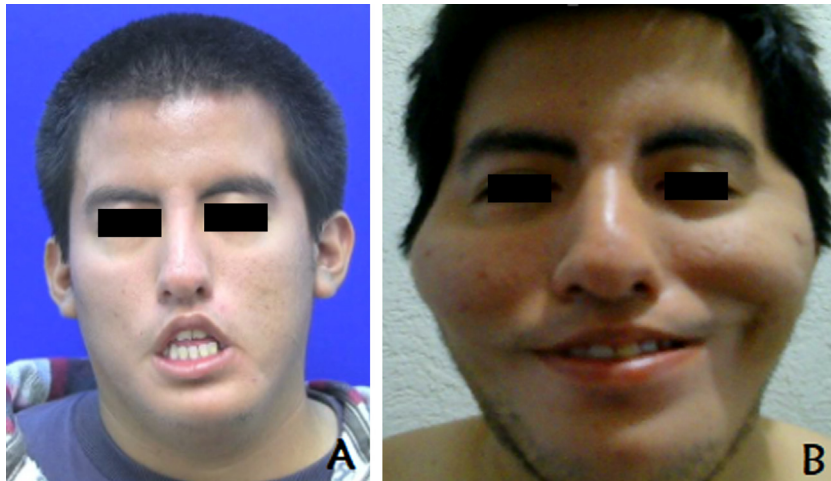


Figure 5 Male patient who underwent a free gracilis flap transfer, double innervated. A. Preoperative condition. B. Postoperative condition.

we observed a notable and better self esteem, as well as development of enhanced interpersonal relationships.

Conclusion

The free gracilis flap transfer is a reproducible procedure for patients with Möbius Syndrome. It is of utmost importance to select the best motor nerve possible, based on an individualized preoperative clinical and electromyographic evaluation. To our best knowledge, this is the largest series of patients with Möbius syndrome worldwide treated at a single-institution.

Disclosure of interest

The authors declare that they have no competing interest.

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