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Clinical Paper
Congenital Craniofacial Anomalies

Facial animation in patients with Moebius and Moebius-like syndromes

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Abstract. Moebius syndrome, a rare congenital disorder of varying severity, involves multiple cranial nerves and is characterised predominantly by bilateral or unilateral paralysis of the facial and abducens nerves. Facial paralysis causes inability to smile and bilabial incompetence with speech difficulties, oral incompetence, problems with eating and drinking, including pocketing of food in the cheek and dribbling, as well as severe drooling. Other relevant clinical findings are incomplete eye closure and convergent strabismus. The authors report on 48 patients with Moebius and Moebius-like syndromes seen from 2003 to September 2007 (23 males and 25 females, mean age 13.9 years). In 20 cases a reinnervated gracilis transplant was performed to re-animate the impaired sides of the face. In this series, all free-muscle transplantations survived the transfer, and no flap was lost. In 19 patients complete reinnervation of the muscle was observed with an excellent or good facial symmetry at rest in all patients and whilst smiling in 87% of cases. In conclusion, according to the literature, the gracilis muscle free transfer can be considered a safe and reliable technique for facial reanimation with good aesthetic and functional results.

Keywords: Moebius syndrome; paralysis; animation; face.

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Moebius syndrome/sequence is a rare congenital disorder of varying severity characterised by bilateral or unilateral paralysis of the facial and abducens nerves[1,3,4,7,14,17,19,20]. Its aetiology is unknown, but environmental and genetic factors have been implicated. Recent studies suggest the occurrence of a vascular disruption causing a hypoxic/ischaemic insult to the brain stem during the first trimester, which could affect some of the cranial nerves[4,5,7,14,19,20]. The hypoxic/ischaemic insult might result from uterine contractions due to a variety of causes. An abortifaciant drug, misoprostol, has been linked to self-induced but unsuccessful abortions that result in some subjects with the clinical findings of Moebius sequence[14]. Although evidence of a genetic mechanism has been noted in a few subjects, most cases of Moebius sequence appear to be sporadic[14].

Paralysis of the VI and VII cranial nerves leads to lack of function in the muscles they supply. Lateral gaze and facial animation are absent. When the paralysis is bilateral, it can be asymmetric, and facial movement, when present, is always located in the lower face with platysmal activity or depressor anguli oris activity. Effective lower-lip support, lower-lip elevation for bilabial speech production and commissure movement and upper-lip elevation for smiling and emotional expression are absent, and the inability of these patients to smile often leads to the mistaken impression that they are dull and disinterested. Speech difficulties are complex and multifaceted, and facial paralysis often leads to bilabial incompetence, which causes the characteristic speech pattern of flaccid dysarthria consisting of substitution, distortion or omission of the bilabial phonemes/p/,/b/ and/m/and the alveolar phonemes/t/,/d/

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and/n/[1,4,17,19]. Paralysis of the lower face often causes problems with eating and drinking, including pocketing of food in the cheek, dribbling and severe drooling[1,6,12]. Other relevant clinical findings are incomplete eye closure and convergent strabismus. In addition to the abducens and facial nerves, the hypoglossal nerve is the most commonly involved cranial nerve, being affected in 25% of cases[1,6]. This manifests with further problems of articulation, primarily with vowels. Velopharyngeal incompetence is present in about 11% of patients because of glossopharyngeal nerve palsy, which results in a hypernasal voice, glottal substitutions and increased nasal air emission[6]. Mental disability is overdiagnosed and is only present in 14% of patients[6]. This mistake is often due to the frequent inability of people to understand what these patients are saying or feeling. When these patients begin school, they are often teased by other children, which can cause them to become withdrawn and reluctant to speak, despite normal intelligence. The inability to show happiness, sadness or anger frequently results in severe introversion and a reclusive personality with low self-esteem[17]. Impairments of the II, V, X and XI cranial nerves have also been observed in rare instances[1,6,17,20].

Patients with Moebius syndrome have been associated with additional deformities that include limb malformation (clubfeet and agenesis, rudimentary fingers or toes and syndactyly or brachydactyly), malformations of the orofacial structures (bifid uvula, micrognathia, cleft palate, small palpebral fissures, epicanthic folds, ocular hypertelorism, microstomia, external ear deformity with occasional hearing loss and airway problems with aspiration), musculoskeletal malformations (absence of the sternal head of the pectoralis major muscle, rib defects and arthrogryposis; cases with dextrocardia have been described) and dysfunction of the cerebrum (mental retardation and epilepsy)[13,17]. Moebius syndrome may also be associated with Poland syndrome, Klippel–Feil anomaly, Kallmann syndrome and Hanhart syndrome[17].

The restoration of even a small degree of volitional facial movement can be rewarding in terms of verbal and nonverbal communication. In this study, the authors report on patients with Moebius and Moebius-like syndromes seen and treated surgically from 2003 to September 2007. The authors evaluate the effectiveness of gracilis transplant to restore facial movement and indications and results in the use of the controlateral facial nerve or of the masseter motor nerve in providing adequate innervation to the muscle transfer. The complications and the outcomes of the different techniques, focusing on functional and aesthetic issues such as oral competence, speech and the degree of movement and its impact on these patients, is analysed.

Table 1. Patient classification.

	n	%
Moebius	19	39.6
Moebius incomplete	8	16.7
Moebius-like	21	43.7
Total	48	100

Table 2. Associated findings in patients with Moebius syndrome.

	n
Poland syndrome	2
GERD	2
Cleft palate	3
Syndactyly	2
Bifid uvula	1
Clubfeet	3
Micrognathia	3
Testicle anomalies	2
Pituitary dysfunction	1
Angel wing	1
Mental retardation	2
Persistent ductus arteriosus	1
Hearing reduction	1
Epicanthic fold	2

Materials and methods

The authors reviewed the records of 48 patients with Moebius and Moebius-like syndromes seen between 2003 and September 2007. They comprised 23 males and 25 females with a mean age, when first seen, of 13.9 years (range 1–48 years). 27 patients had bilateral Moebius syndrome, a monolateral form was present in 21 patients with involvement of the right side in 8 and the left side in 13. The patients were classified (Table 1), following the classification proposed by Terzis et al.[16,17,23]: Moebius (complete bilateral facial and abducens nerve paralysis); Moebius incomplete (clinical picture of Moebius with the exception that some residual motor function was noted on one side of the face); Moebius-like (unilateral facial paralysis, but additional cranial nerve palsies present).

In those with the Moebius incomplete form, some facial movements were evident, which were always located in the lower face with platysmal activity or depressor anguli oris activity. The abducens nerve was involved in 70% of the patients. In 13 patients, additional cranial nerves were involved, of which the hypoglossal nerve was impaired in 25% and the motor branches of the fifth cranial nerve in 4%.

A standardised neurological and logopaedic examination was performed in all cases. Facial expression, oral motor function and speech were evaluated clinically, and most patients underwent electromyographic examinations during their first visit. Special attention was directed toward the identification of possible motor donor nerves. Examination of the facial nerve included needle electromyography of the facial musculature, including the upper, middle and lower facial territories. The temporalis and masseter muscles were tested clinically. All patients were videotaped and photographed with particular attention to facial expression, oral motor function and speech.

The logopaedic evaluation showed alterations of speech patterns with articulation difficulties and substitution or distortion of the bilabial phonemes/p/,/b/and/m/in 40% of the patients. Speech was severely altered in 10%. The associated findings are shown in Table 2.

Patients satisfying the following criteria were excluded from 'smile surgery': age less than 6 years; low collaboration due to mental retardation; good functional repair of the residual motor units; anomalies in walking; absence of a collaborating family.

Twenty patients underwent microsurgical reconstruction to re-animate the impaired sides of the face. A segment of the gracilis muscle was transplanted in all cases. The facial artery and vein were used as recipient vessels in most of the procedures. In one patient, the facial vein was not found so the transverse facial vein was used. Ten patients underwent a bilateral free-muscle transplantation, with a total of 30 gracilis free flaps. The contralateral facial nerve was used as a motor donor nerve in 7 procedures, the motor nerve to the masseter muscle in 13 patients, 10 with bilateral and 3 with monolateral facial paralysis.

Cross-facial nerve graft

On the normal side through a preauricular incision, the branches of the facial nerve are identified as they exit the anterior portion of the parotid fascia. With the aid of a nerve stimulator, a map of the muscles they innervate can be made, identifying the buccal and zygomatic branches. Segments of these branches can be sacrificed as their activity is dupli-