Novel Use of C7 Spinal Nerve for Moebius

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Background: The purpose of this study was to introduce the use of selective ipsilateral C7 spinal nerve transfer to the armamentarium of the dynamic procedures used for facial reanimation in Moebius patients.
Methods: Between 1991 and 2007, the selective ipsilateral C7 technique was used in four patients. In three patients with bilateral paralysis, both C7 spinal nerves were utilized as nerve donors. Thus, evaluation of outcomes was carried out in seven hemifaces.
Results: Short-term paresthesia on the index pulp of the donor upper extremity was observed in four limbs, which recovered spontaneously. Motor deficits were never noticed. Neurotization of the free-muscle transfer for smile was performed in five hemifaces, with good results in three hemifaces ($p < 0.01$, $z = 2.61$). Eye closure neurotization was made in six hemifaces, with good results in four hemifaces ($p < 0.01$, $z = 2.88$). Depressor neurotization was made in one case and resulted in improved depressor complex function. In two cases of free muscle transfer for masseter substitution, electromyographic interpretations revealed full motor activity after bilateral latissimus dorsi transfer. Tongue neurotization was performed in two instances. Postoperatively, the patients' speech intelligibility improved as well as tongue motility and bulk.
Conclusion: The use of the C7 as a motor donor in Moebius cases with multicranial nerve involvement supplies the typical mask-like face with an abundance of motor fibers for facial reanimation and, if there is a coexisting twelfth nerve palsy, for speech restoration. (*Plast. Reconstr. Surg.* 126: 106, 2010.)

The introduction of magnification (1964) in the treatment of peripheral nerve injuries established new possibilities for the field of facial nerve repair. Although substantial progress has been made,[1–6] substitution of the affected facial nerve remains a challenge, especially in Moebius cases.

In 2003, our center reported a series of 43 patients, the largest series of Moebius patients in North America, 20 of whom have been operated on by a single surgeon (J.K.T.).[7] The surgical strategy for dynamic restoration in these unfortunate patients is based on the degree of involvement of the fifth, eleventh, and twelfth nerves. In Moebius patients with simultaneous involvement of the fifth, eleventh, and twelfth nerves, the senior author (J.K.T.) has introduced the use of motor fibers from the ipsilateral C7 spinal nerve as motor donors for dynamic voluntary expression.

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Gu et al.[8] introduced the use of the contralateral C7 as a motor donor in cases of brachial plexus avulsions by utilizing the entire contralateral root. Terzis[9] modified this technique. In November of 1991, Terzis introduced the "selective contralateral C7 technique," which involves the use of anterior and posterior division fibers for reinnervation of contralateral flexors and extensors, respectively. This refinement prevented unnecessary loss of the entire C7, provided gratifying degrees of functional return, and made rehabilitation much easier as the

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regenerating fibers were reaching similar corresponding targets.[10,11]

To our knowledge, the use of the ipsilateral C7 for rendering dynamic facial motion in severe Moebius cases has not been previously described. The purpose of this report is to introduce an already tested technique for brachial plexus reconstruction to the armamentarium of the dynamic procedures used for facial reanimation in Moebius patients.

PATIENTS AND METHODS

Between 1978 and 2007, 25 patients with Moebius syndrome underwent facial reanimation in our center. The selective ipsilateral C7 technique was used in four patients (of 25). In three patients (of four) with classic Moebius, bilateral C7 spinal nerves were utilized as motor donors. Thus, evaluation of outcomes was carried out in seven hemifaces.

Eligibility criteria included (1) a clinical diagnosis of Moebius syndrome, (2) facial reanimation with the selective ipsilateral C7 technique, and (3) follow-up of 2 years or more. Patients' demographic characteristics are summarized in Table 1.

Preoperative Evaluation

The preoperative evaluation consisted of a detailed history, physical examination, and full neurologic assessment of the cranial nerves. The muscles of facial expression, if any, were examined and videotaped in detail. Ancillary investigations included needle electromyography and nerve conduction studies, and when indicated, computed tomography of bilateral facial nerve canals within the bilateral temporal bones. At all subsequent follow-up visits, physical examination, neurologic assessment, and repeated electromyography were performed.

Our center, in 2002, introduced a new classification system for Moebius[12] that highlights the side of involvement and degree of nerve palsy of the cranial nerve motor pools and uses the CLUFT (cranial nerve, lower limb, upper limb, face, and thorax) system, as proposed by Abramson et al.,[13] to indicate associated deformities in the rest of the body.

Moreover, a thorough neurologic assessment of the donor upper extremity was carried out to verify the integrity of the ipsilateral C7. Evaluation of the donor upper extremity was repeated at each follow-up visit.

The Terzis facial grading system, published in 1997,[14] was used by three independent reviewers who each reviewed separately all the preoperative and postoperative videotapes. Results were graded as poor, fair, moderate, good, or excellent. Electromyographs of the targets neurotized, relating to the preoperative consultation and the last follow-up, were also analyzed by using the Terzis grading scale for the electromyography interpretations.[15]

Evaluation of selective neurotizations of targets related to other cranial nerve deficits (tongue neurotization for improving speech) was also performed based on reviewing electromyography records and patients' videotapes relating to the preoperative consultation and the last follow-up visit.

C7 Spinal Nerve Anatomy

The C7 emerges from the corresponding foramen and continues on as the middle trunk. In 1987, the senior author (J.K.T.)[16] reported a quantitative microanatomy of the brachial plexus in humans. In this study, the relative amount of C7 neural tissue was constant at 24 percent (24.1 ± 1.4 percent). The distribution of C7 neural tissue to the posterior, medial, and lateral cord of the plexus was 44.6 to 44.8 percent, 8.2 percent, and 44.0 to 44.2 percent, respectively. C7 fibers to the lateral cord were mainly distributed to the median nerve (34.0 to 43.1 percent of its fibers) and were sensory. On the other hand, C7 fibers to the posterior cord were distributed to the radial nerve (23.9 to 56.3 percent of its fibers) and were mostly motor. Bonnel[17] reported that the total average count of myelinated axons for C7 was 23,781 (range, 16,000 to 40,000).

Table 1. Demographic Characteristics of the Patient Population

Hemiface	Age (yr)	Moebius Type*	Sex	Side	Denervation Time (mo)	Follow-Up (mo)	Nerve Involvement
1	15	I	Female	Right	182	95	c7, c5, p6, c9, c11, c12
2	15	I	Female	Left	180	97	c7, c5, p6, c9, c11, c12
3	43	II	Female	Left	519	72	c7, p6, p12
4	3	I	Male	Right	44	26	c7, p6, p12
5	4	I	Male	Left	33	37	c7, p6, p12
6	4	I	Female	Right	52	28	c7, c6, p9, p10, p12
7	4	I	Female	Left	48	32	c7, c6, p9, p10, p12
Mean ± SD	12.5 ± 14.1				151.2 ± 175	55.9 ± 31.8	

c, complete paralysis; p, partial paralysis. *For Moebius type, type I refers to classical Moebius; type II refers to incomplete Moebius.

Dykes and Terzis[18] found that dermatomal and myotomal distribution of the spinal nerves in monkeys varied but always maintained the same relative cephalo-caudal orientation, supporting the first law of Herringham.[19] The C7 myotome includes fibers serving the postaxial musculature and musculature of the lower part of the shoulder girdle (triceps, pectoralis major, latissimus dorsi, wrist, and finger extensors).

Terzis' Selective C7 Technique

The patient is placed in the supine position, with the head turned away from the operative side. A curved incision along the posterior border of the sternocleidomastoid muscle is carried out, and the supraclavicular plexus is identified between the anterior and the middle scalene muscles. The C7 is located caudally and posterior to the C6, and it is mobilized from the surrounding tissues. Working from proximal to distal, the C7 is exposed until its bifurcation into the anterior division and posterior division.

After longitudinal epineuriotomy and using intraoperative electrical stimulation, each bundle within each division of the C7 is mapped.[10,11] The intraoperative mapping of the components of each C7 division is a mandatory step. Bundles supplying the pectoralis major in the anterior division and the latissimus dorsi and triceps muscles in the posterior division are isolated with vessel loops. Bundles that supply wrist extensors are never sacrificed.

Under high magnification, perineurial windows are performed with partial neurotomies. Interposition nerve grafts are brought in the operating field, and these are coapted with selected C7 components in an end-to-side fashion. In the classic Moebius cases (Table 2), corresponding bundles of each C7 division are guided to similar targets on each side of the face. For example, motor fibers that innervate the pectoralis major (anterior division) are used for direct neurotization of sphincters (e.g., orbicularis oculi muscle for enhancing eye closure), whereas those that innervate the latissimus dorsi and triceps muscles (posterior division) are used as motor donors for levators (tongue, free-muscle neurotization for smile, and depressor complex restoration).

Table 2. Terzis' Classification System of Moebius Syndrome

Type	Cranial Nerve Impairment
Classical Moebius	
I	Complete bilateral facial and abducens nerve paralysis
Incomplete Moebius	
II	Clinical picture of Moebius, with residual motor function on one side of the face
Moebius-like	
III	Unilateral facial paralysis with additional cranial nerve palsies

Nerve Grafts: Targets

The number and length of nerve grafts are assessed by the distance between the C7 and each target that needs neurotization. Bilateral sural nerves are harvested, and if needed, bilateral saphenous nerves are harvested as well. In the adult, the length of each saphenous graft can easily reach 50 to 60 cm. Each nerve graft is tunneled separately using custom-made flexible metal passers to the respective targets to ensure rapid revascularization.

Under high magnification, coaptations between nerve grafts and C7 components take place at sites of perineurial windows and microneurotomies. Selective neurotizations of the orbicularis oculi muscle, tongue, orbicularis oris muscle, and depressor are performed. In cases in which free muscle transfer for smile restoration is planned, nerve grafts are banked at the ipsilateral cheek area so that they can be used at a later stage for innervation of free muscles. The time interval between C7 transfer and free muscle transfer is approximately 6 to 9 months, depending on the relative distance between C7 and the target, the degree of the elicited Tinel's sign, and the conduction velocity results. Direct neurotization of the selected target is accomplished by implanting the distal end of the interposition nerve graft into the substance of the muscle. The nerve end is split into two to three fascicles, and each fascicle is implanted through a separate incision within the epimysium.

Postoperative Care

A specially designed, custom-made head restrainer splint is used to protect the microneural coaptations and is removed 4 weeks later. If tongue neurotization has been performed, the patients are placed on a blended diet for a month.

Our center's postoperative protocol, especially in facial reanimation cases, includes massage and ultrasound therapy, which is applied over the coaptation sites in the neck and in the cheek where the grafts have been tunneled, or over the transferred muscle at the beginning of the sixth postoperative week for a duration of 6 weeks. This strategy has been shown to be beneficial in preventing scar formation around the nerves or free muscles and lessens adhesions with the overlapping skin envelope.

Statistical Analysis

Each side of the face (hemiface) was evaluated independently from the other. The Wilcoxon signed rank test for paired samples was used to compare preoperative and postoperative groups by grade and by electromyographic interpretations. All the analyses were performed using SAS 9.1.3 software (SAS, Gary, N.C.). The significance level was set at 0.05. The sample size limited the power of this study; thus, statistical comparisons were made only for neurotizations with an adequate number of cases ($n \geq 3$) for statistical analysis.

RESULTS

Patients' mean age, denervation time, and follow-up at the time of the first visit were 12.3 years, 151 months, and 55.9 months, respectively. None of the patients had a family history of Moebius syndrome. The sixth and seventh nerves were involved in all cases (Table 1). Associated findings were micrognathia ($n = 2$), microglossia ($n = 2$), phocomelia ($n = 2$), and speech problems ($n = 2$). The reconstructions performed in all cases are summarized in Table 3.

A total of 19 neurotizations were performed in seven hemifaces (four patients) by utilizing 19 interposition nerve grafts. The type of nerve graft and the different donor (anterior division or posterior division of C7) are summarized in Table 4. The selected targets in regard to the donors are presented in Table 5. The selected donors according to the target neurotized on each hemiface and patient are summarized in Table 6. Preoperative and postoperative grading for eye closure, smile, and depressor by the three reviewers is summarized in Table 7. Preoperative and postoperative electromyographic interpretations for all the targets neurotized by C7 are presented in Table 8.

Four of seven hemifaces experienced short-term paresthesia on the index pulp of the donor upper extremity, which recovered spontaneously in all cases. The recovery period ranged from 1 to 5 months. Motor deficits were never noticed.

The gracilis was used as a free muscle for nasolabial fold excursion on five occasions and the latissimus dorsi as a free muscle for masseter substitution in two hemifaces. All the free muscles were neurotized by fascicles from the ipsilateral posterior division (higher motor content) of the C7. The free muscle transfer for smile increased the average preoperative scores in all the patients ($p < 0.01$, $z = 2.61$). Good results were observed in three of five hemifaces. The statistical evaluation of the electromyographic interpretations before and after free muscle transfer for smile restoration was significant ($p < 0.01$, $z = 2.61$). In cases of masseter substitution, electromyographic interpretations revealed full motor activity after bilateral latissimus dorsi transfer.

Neurotization of the orbicularis oculi muscle was performed in six hemifaces from motor fascicles of the ipsilateral anterior division of the C7. In one patient (hemifaces 1 and 2), two nerve grafts were used for each orbicularis oculi muscle

Table 4. Selected Donors (Anterior and Posterior Division of the C7 Root) and Type of Nerve Graft

	Nerve Grafts		
Donors	Sural	Saphenous	Total
AD	8	1	9 (47%)
PD	5	5	10 (53%)
Sum (%)	13 (68%)	6 (32%)	19 (100%)
Total	19 (100%)		

AD, anterior division of C7 spinal nerve; PD, posterior division of C7 spinal nerve.

Table 3. Description of Reconstruction for Each Hemiface

	Donor Nerves				Targets						Postoperative Result					
	AD		PD		AD		PD				AD		PD			
Hemiface	Su	Sa	Su	Sa	OOM	OOrM	Bi	To	Sm	De	OOM	OOrM	Bi	To	Sm	De
1	1	1	2	1	1 (×2)	1	1	—	1	—	Good	Good	Good	—	Good	—
2	1	1	1	1	1 (×2)	—	1	—	1	—	Good	—	Good	—	Good	—
3	1	—	1	—	1	—	—	—	1	—	Mod	—	—	—	Mod	—
4	1	—	1	—	1	—	—	1	—	—	Good	—	—	Good	—	—
5	1	—	1	—	1	—	—	—	—	1	Good	—	—	—	—	Exc
6	1	—	—	1	—	—	—	—	1	—	—	—	—	—	Mod	—
7	—	—	—	2	1	—	—	1	1	—	Mod	—	—	Good	Good	—

AD, anterior division of C7 spinal nerve; PD, posterior division of C7 spinal nerve; Su, sural nerve graft; Sa, saphenous nerve graft; OOM, orbicularis oculi muscle; OOrM, orbicularis oris muscle; Bi, bite; To, tongue; Sm, smile; De, depressor; Mod, moderate; Exc, excellent.

due to a severe deficit of multiple cranial nerves bilaterally. Eye closure after neurotization was improved in all cases, with good results in four of six hemifaces ($p < 0.01$, $z = 2.88$). The statistical evaluation of the electromyographic interpretations before and after orbicularis oculi muscle neurotization was significant ($p < 0.01$, $z = 2.88$).

Tongue neurotization was performed in two hemifaces with speech impairment due to severe hypoglossal nerve deficit. Tongue muscle was neurotized through interposition nerve grafts which coapted to fascicles of the posterior division of the C7. The last follow-up electromyography showed full motor activity of the tongue musculature (at 26 and 32 months). Also, the volume and mobility of tongue

Table 8. Mean ± SD Preoperative and Postoperative Electromyographic Interpretations for All the Targets Neurotized by C7 Root Components

	EMG		
Factor	Preoperative	Postoperative	p
Eye closure	0.5 ± 0.5	2.67 ± 0.5	<0.01*
Smile	0.6 ± 0.5	2.6 ± 0.5	<0.01*
Tongue	1	2.5	—
Bite	0	2	—
Depressor	0	3	—
Orbicularis oris muscle	0	2	—

EMG, electromyography.

Table 5. Selected Donors (Anterior and Posterior Division of the C7 Root) per Target Neurotized

	Targets					
Donors	Smile	Tongue	Eye	OOrM	Bite	Depressor
AD	—	—	8	1	—	—
PD	5	2	—	—	2	1
Sum (%)	5 (26%)	2 (11%)	8 (42%)	1 (5%)	2 (11%)	1 (5%)
Total			19 (100%)			

Smile, free gracilis transfer neurotization for smile restoration; Eye, orbicularis oculi muscle neurotization for eye closure; OOrM, orbicularis oris muscle neurotization for mouth closure; Bite, free latissimus dorsi transfer neurotization for masseter substitution; AD, anterior division of C7 spinal nerve; PD, posterior division of C7 spinal nerve.

Table 6. Selected Donors (Anterior and Posterior Division of the C7 Root) According to the Target Neurotized on Each Hemiface and Patient

			Anterior Division of C7		Posterior Division of C7			
Patient	Hemiface	Side	OOM	OOM	Smile	Bite	Tongue	Depressor
1	1	R	2	1	1	1	—	—
	2	L	2	—	1	1	—	—
2	3	L	1	—	1	—	—	—
3	4	R	1	—	—	—	1	—
	5	L	1	—	—	—	—	1
4	6	R	—	—	1	—	—	—
	7	L	1	—	1	—	1	—
4	7		8 (42%)	1 (5%)	5 (26%)	2 (11%)	2 (11%)	1 (5%)

OOM, orbicularis oculi muscle neurotization for eye closure; OOrM, orbicularis oris muscle neurotization for mouth closure; Smile, free gracilis transfer neurotization for smile reanimation; Bite, free latissimus dorsi transfer neurotization for masseter substitution.

Table 7. Preoperative and Postoperative Grading, Using Terzis' Facial Grading System for Eye Closure, Smile, and Depressor, by the Three Reviewers

	Mean Preoperative Scores			Mean Postoperative Scores			
Factor	Reviewer 1	Reviewer 2	Reviewer 3	Reviewer 1	Reviewer 2	Reviewer 3	p
Eye closure	1.16 ± 0.4	1.66 ± 0.8	1.16 ± 0.4	3.66 ± 0.5	4.33 ± 1.03	4.0 ± 0.89	<0.01*
Sum		1.33 ± 0.59			4.0 ± 0.84		
Smile	1	1.6 ± 0.5	1.2 ± 0.4	3.6 ± 0.5	3.8 ± 0.4	3.6 ± 0.5	<0.01*
Sum		1.26 ± 0.4			3.6 ± 0.5		
Depressor	1	1	1	5	5	5	—
Sum		1			5		

Fig. 1. The patient was born with bilateral global facial paralysis. The patient also had a partial deficit of abducens nerve (sixth) bilaterally and complete paralysis of the fifth, eleventh, and twelfth nerves bilaterally. The diagnosis of Moebius syndrome was made. The patient was first seen in our center at the age of 15 years. The patient underwent her first-stage reconstruction (15 years' denervation time), in which 30 percent of the anterior division of the left C7 root and 80 percent of the posterior division were used as motor donors. Direct neurotization of left upper and lower orbicularis oculi muscle took place by the anterior division. Two grafts were tunneled to the temporal area and coapted proximally to the posterior division. Two months later, the patient underwent her second stage of reconstruction, in which 75 percent of the anterior division and 80 percent of the posterior division of the right C7 root were used as motor donors. Direct neurotization of the right orbicularis oculi muscle and the orbicularis oris muscle took place from the anterior division. Similarly, two grafts were tunneled to the right temporal area and coapted proximally to the right posterior division of C7. Six months later a free gracilis muscle for nasolablal fold excursion was transferred to the left cheek neurotized by a banked nerve from the left posterior division. Two months later a second gracilis muscle was transferred for smile restoration into the right cheek neurotized by a banked nerve from the posterior division of the right C7 root. Twenty months later, a proximal segment of the contralateral free latissimus dorsi muscle was transferred for bite restoration to the right cheek neurotized by the second banked nerve from the right posterior division. Two months later, a second free latissimus dorsi was transferred for bite restoration to the left face neurotized by a banked nerve from the left posterior division. Despite a long denervation time (15 years) and severe multicranial nerve lesions, both aesthetic and functional improvement was noted. (*Above, left*) Patient during her first office visit to our center. (*Above, center*) Patient during last follow-up visit. Note improvement in mouth closure (*above, center*) and smile (*above, right*). (*Below, left*) Patient's attempt at eye closure during first office visit. (*Below, right*) Patient's eye closure postoperatively, 2 years after direct muscle neurotization of right and left upper and lower eye sphincters.

musculature were restored and speech improved in both instances in follow-up visits and according to speech therapists' reports. Depressor neurotization was carried out in one case and resulted in improved depressor complex function. Patient photographs are presented in Figures 1 through 5.

See Video, Supplemental Digital Content 1, which demonstrates patient 3, who was born with bilateral facial palsy and impairment of bilateral sixth and seventh cranial nerves and was diagnosed with Moebius syndrome, attempting to speak preoperatively, ***http://links.lww.com/PRS/A186.*** Speech intelligibility was graded as poor. **See Video, Supplemental Digital Content 2,** which shows the patient postoperatively. Two years after the tongue neurotization procedure, in his last office visit, his tongue mobility and speech showed improvement, and for the first time he acquired understandable speech, ***http://links.lww.com/PRS/A187.*** **See Video, Supplemental Digital Content 3,** which demonstrates patient 4, a 3-year-old girl with Moebius syndrome, micrognathia, microglossia, and phocomelia of the right upper extremity, whose speech, preoperatively at her first office visit, consisted of gurgling sounds, ***http://links.lww.com/PRS/A188.*** **Supplemental Digital Content 4** shows the patient 11 years after the bilateral tongue neurotization procedure, demonstrating that her speech is more understandable and intelligible, ***http://links.lww.com/PRS/A189.***[20]

DISCUSSION

The concept of cross-facial nerve grafting does not apply to the classic Moebius patient because of bilateral facial nerve involvement. Static procedures are not advocated if the main goal is to bring movement to the face.[21] A standard procedure for dynamic restoration in Moebius should not be promoted; instead, a careful preoperative objective and quantitative assessment should guide the surgeon to the optimal strategy. An electromyographic survey of all the potential motor donors is

Fig. 2. This 43-year-old woman was born to a 47-year-old mother as a fifth child. The facial asymmetry was noted since birth. Exploration of the right cheek revealed an extratemporal facial nerve that was found to be atrophic and underdeveloped. An equally tiny branch of the sural nerve was used as a cross-facial nerve graft that was coapted to the tiny right main zygomatic branch in an end-to-side fashion in an effort not to downgrade the smile she had on the right face. In addition, two sural nerve grafts were coapted to the left C7 root by selective neurectomies of bundles in the posterior and anterior divisions. The motor fibers from the latter went for direct neurotization of the left eye sphincter, whereas the nerve graft from the posterior division was banked in the left cheek. A year later, a free gracilis was transferred to the left cheek and was neurotized by the previous placed cross-facial nerve graft and also by the nerve graft from the posterior division of the ipsilateral C7. (*Left*) The patient during her first office visit in our center. Note the asymmetry in her smile. (*Right*) Patient's appearance during her last office visit while attempting to smile. Improvement in functional and aesthetic appearance is noted.

Fig. 3. The patient was born with bilateral facial palsy and impairment of bilateral 12th and sixth cranial nerves. The diagnosis of Moebius syndrome was made. The fifth nerves were intact bilaterally. The patient presented to our center at 1 year and 3 months of age (*above, left*). He underwent strabismus correction surgery 2 years and 3 months after birth. At 2 years and 9 months' denervation time, the patient had a free gracilis muscle transfer for smile restoration into the left cheek. The muscle was neurotized by the ipsilateral masseteric nerve via an end-to-side coaptation. Twelve months later, a second free gracilis muscle was transferred to the right cheek for smile restoration, which was also neurotized via end-to-side coaptation from the ipsilateral masseteric nerve. Eight months later, the patient underwent bilateral C7 root exploration and harvesting of both sural nerves. Four interposition nerve grafts (two on each side) were coapted in an end-to-side fashion to the anterior and posterior divisions of the ipsilateral C7 root. Neurotizations of both orbicularis oculi muscle took place from the anterior divisions of bilateral C7 roots. The right side of the tongue and left depressor were neurotized from the posterior divisions. Preoperative smile, eye closure (*below, left*), and speech intelligibility were graded as poor (see Video, Supplemental Digital Content 1). The postoperative results for smile (*above, right*) and eye closure (*below, right*) were graded as good.

mandatory to avoid further downgrading of function. In contrast to other authors,[6] the twelfth nerve is rarely used in our center because speech and swallowing are severely impaired in the vast majority of Moebius patients. In the current series, the twelfth nerve was involved in all cases.

The fifth nerve, mostly the masseteric branch, is a tempting source because free muscle transplants can be connected directly, thus sparing the stage of nerve transplantation.[22] On the other hand, in severe cases, the fifth nerve is also involved, and it is wise to conduct needle electromyographies so that harvesting of this nerve will not increase oral crippling. Signs of involvement of the temporalis or the masseter muscle exclude the use of this potential donor.[23]

Selective Ipsilateral C7 Spinal Nerve Transfer

The concept of the selective ipsilateral C7 transfer for facial reanimation in Moebius cases

Fig. 4. Same patient as in Figure 3. His left depressor was substantially improved by the direct neurotization procedure (*above, left,* preoperative view; *above, right,* postoperative view). His tongue bulk, mobility, and speech showed remarkable improvement, and for the first time, he acquired understandable speech (*below, left,* preoperative view; *below, right,* postoperative view) (See Video, Supplemental Digital Content 2).

allows the neurotization of multiple targets and also provides motor fibers for future free-muscle transfers. The use of extensive intraoperative microstimulation and mapping of the motor territories of C7 is important to select noncritical motor donor bundles.

Moreover, by using this "mapping" of C7, selective bundles can be used for synergistic muscles in the involved face (anterior division motor fibers for the ipsilateral orbicularis oculi muscle and posterior division for the ipsilateral free-muscle for smile or the depressor complex). This approach minimizes the possibility of co-contractions between sphincter muscles (orbicularis oculi muscle) and levator muscles (free muscle for smile).

In two recent reports from our center[10,11] dealing with the selective contralateral C7 transfer in adult brachial plexus reconstruction, the postoperative morbidity was very limited; 71 percent of the patients experienced numbness in the median nerve that resolved within 6 months, and motor deficit was never encountered.

The selective ipsilateral C7 transfer has been used by the senior author (J.K.T.) in Moebius cases with multiple cranial nerve involvement. In Moebius patients with bilateral or unilateral twelfth nerve involvement, restoring speech intelligibility (with tongue neurotization) is of equal importance as facial motion restoration, as this promotes psychological and social development in these severely afflicted patients. In the current series, tongue neurotization was performed in two instances with severe speech impairment. Postoperatively, the patients' speech intelligibility improved as well as the appearance, motility, and bulk of the tongue.

The selective ipsilateral C7 transfer can also be used in Moebius cases with bilateral partial or complete fifth nerve palsy for neurotization of free muscle transfer for masseter substitution to restore bite and achieve mouth closure. In the current series, this neurotization was performed in one patient (two hemifaces) with total bilateral fifth nerve palsy (Fig. 1). Postoperatively, the patient's jaw movements and mouth closure were

Fig. 5. (*Above, left*) This 3-year-old girl was born with Moebius syndrome, micrognathia, microglossia, and phocomelia of the right upper extremity. At the first office visit her speech consisted of gurgling sounds (see Video, Supplemental Digital Content 3). In two operations, 4 months apart, the patient had an exploration of the left and right supraclavicular brachial plexus. Bilateral sural and saphenous nerves were harvested. She had direct neurotization of the right orbicularis oculi muscle from the right lower trunk and direct neurotization of the left orbicularis oculi muscle from the anterior division of the left C7 root. She had direct neurotization of the right and left sides of her tongue, which was totally denervated and atrophic, by the right lower trunk and the left posterior division of the left C7 root via sural nerve grafts. Two saphenous nerve grafts were banked to the right and left cheeks. Each was coapted proximally to the ipsilateral posterior division of the C7 root. Over the next 2 years, two free gracilis muscles were transferred to the left and right cheeks. (*Above, right*) At 11 years after her first office visit, the patient had an adequate smile, and her speech was now understandable as a result of the bilateral tongue neurotization procedure (see Video, Supplemental Digital Content 4). Her tongue is shown preoperatively (*below, left*) and postoperatively (*below, right*). Note the improvement in its bulk and shape.

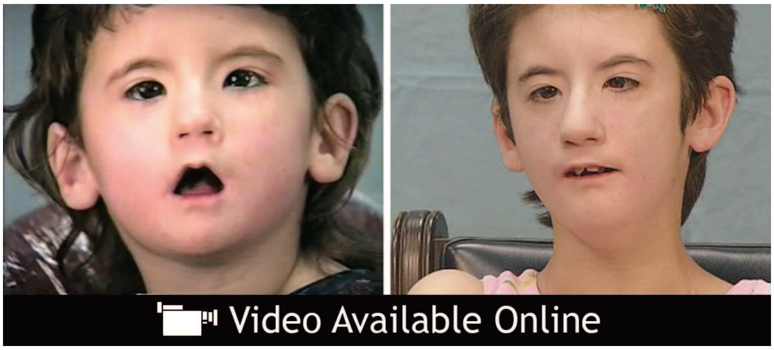
improved. This also improved dental hygiene, which previously was plagued by chronic dessication due to chronic exposure (Fig. 1).

Despite the small sample size of this study, the results of direct neurotizations for orbicularis oculi muscles (four of six hemifaces with good results) and for free muscle transfer for smile (three of five hemifaces with good results) using the selective C7 transfer are encouraging. In the current series, all the patients had late reconstruction (mean denervation time ≥151.2 months) and severe multicranial nerve deficits.

Cortical Adaptation of Selective C7 Transfer

One difficult task after C7 transfer is brain adaptation to coordinate the motor activity of smiling. Gu et al.[8] suggested that in cases in which the contralateral C7 is used as the motor donor for brachial plexus reconstruction, the contraction of the reconstructed

Videos 1 and 2. Supplemental Digital Content 1 shows patient 3, who was born with bilateral facial palsy and impairment of bilateral sixth and twelfth cranial nerves and was diagnosed with Moebius syndrome, attempting to speak preoperatively, ***http://links.lww.com/PRS/A186***. Speech intelligibility was graded as poor. Supplemental Digital Content 2 shows the patient postoperatively. Two years after the tongue neurotization procedure, in his last office visit, his tongue mobility and speech showed improvement, and for the first time he acquired understandable speech, ***http://links.lww.com/PRS/A187***. From Terzis JK, Karypidis D. Direct tongue neurotization: The effect on speech intelligibility in patients with Moebius syndrome. *Plast Reconstr Surg.* 2010;125:150–160.



Videos 3 and 4. Supplemental Digital Content 3 shows patient 4, a 3-year-old girl with Moebius syndrome, micrognathia, microglossia, and phocomelia of the right upper extremity, whose speech at her first office visit consisted of gurgling sounds, ***http://links.lww.com/PRS/A188***. Supplemental Digital Content 4 shows the patient 11 years after bilateral tongue neurotization procedure, demonstrating that her speech is more understandable and intelligible, ***http://links.lww.com/PRS/A189***. From Terzis JK, Karypidis D. Direct tongue neurotization: The effect on speech intelligibility in patients with Moebius syndrome. *Plast Reconstr Surg.* 2010;125:150–160.

muscles needs to be initiated by the simultaneous movement of the intact limb. Moreover, when the power of the reinnervated muscles has reached the M3 level, the patient can achieve active contraction of these muscles without synchronic movement of the healthy limb, which for most patients took about 5 years.[24]

Early age at operation may allow greater plasticity of the cerebral cortex in general and would therefore optimize conditions for cortical plasticity after facial reanimation with a noncranial nerve donor.[25] Cortical plasticity has been demonstrated in multiple instances of sensory and/or motor reinnervation. Merzenich and Jenkins[26] demonstrated remapping of the hand somatosensory cortex after repair of the median nerve in owl monkeys. Similarly, young adults with brachial plexus injuries who underwent biceps neurotization from intercostal nerves show recovery of elbow flexion within 2 years after surgery, and in most cases, elbow flexion and ventilatory movements become segregated.[27]

Beaulieu et al.[27] suggested that after partial neurectomy of the contralateral C7 for biceps neurotization, postoperative elbow flexion on the affected side is controlled by the same areas within both cortex hemispheres. The contralateral regions involved in the new movement network do not seem to belong to a network specifically allotted to a given action and its activation might partly depend on the individual's skills or on motor reeducation procedures.

In the current series, two (hemifaces 1 to 2 and 6 to 7 in Table 1) of the three patients with free muscle transfer for smile restoration developed independent, spontaneous facial motion on follow-up clinical examination. One might speculate that early age at operation in these two patients (4 and 15 years, respectively) resulted in greater plasticity of the cerebral cortex. Another reason could be the neurotization of synergistic muscles from selected donors (posterior division of C7 for free muscles for smile, anterior division for orbicularis oculi muscle). Thus, co-contractions between orbicularis oculi muscle and levators (free muscle for smile) were minimized, which facilitated patient reeducation.

CONCLUSION

The use of the C7 as a motor donor in Moebius cases with multiple cranial nerve involvement supplies the typical mask-like face with an abundance of motor fibers for facial reanimation and, if there is a coexisting twelfth nerve palsy, for speech restoration.

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PATIENT CONSENT

The patients in this study or their parents or guardians provided written consent for the use of their photographs.

REFERENCES

1. Scaramella LF, Tobias E. Facial nerve anastomosis. *Laryngoscope* 1973;83:1834–1840.
2. Smith JW. A new technique of facial animation. In: Huston JT, ed. *Transactions of the 5th International Congress of Plastic Surgery*. London: Butterworth; 1971:83.
3. Harii K, Ohmori K, Torii S. Free gracilis muscle transplantation, with microneurovascular anastomoses for the treatment of facial paralysis: A preliminary report. *Plast Reconstr Surg.* 1976;57:133–143.
4. Terzis JK. Pectoralis minor: A unique muscle for correction of facial palsy. *Plast Reconstr Surg.* 1989;83:767–776.
5. Terzis JK. Babysitters: An exciting new concept in facial reanimation. In *Proceedings of the 6th International Symposium on the Facial Nerve*. Rio de Janeiro, Brazil: Kugler and Ghendini Publications; 1988.
6. Zuker RM, Manktelow RT. A smile for the Moebius' syndrome patient. *Ann Plast Surg.* 1989;22:188–194.
7. Terzis JK, Noah EM. Dynamic restoration in Möbius and Möbius-like patients. *Plast Reconstr Surg.* 2003;111:40–55.
8. Gu YD, Zhang GM, Chen DS, et al. Cervical nerve root transfer from contralateral normal side for treatment of brachial plexus root avulsions. *Chin Med J (Engl).* 1991;104:208–211.
9. Terzis JK. Experience with selective contralateral C7 technique in fifty cases of devastating brachial plexus paralysis. Paper presented at: Annual Meeting of the American Society for Surgery of the Hand, Nashville, Tenn., September 30 to October 3, 1996.
10. Terzis JK, Kokkalis ZT. Selective contralateral C7 transfer in posttraumatic brachial plexus injuries: A report of 56 cases. *Plast Reconstr Surg.* 2009;123:927–938.
11. Terzis JK, Kokkalis ZT, Kostopoulos E. Contralateral C7 transfer in adult plexopathies. *Hand Clin.* 2008;24:389–400, vi.
12. Terzis JK, Noah EM. Möbius and Möbius-like patients: Etiology, diagnosis, and treatment options. *Clin Plast Surg.* 2002;29:497–514.
13. Abramson DL, Cohen MM Jr, Mulliken JB. Mobius syndrome: Classification and grading system. *Plast Reconstr Surg.* 1998;102:961–967.
14. Terzis JK, Noah EM. Analysis of 100 cases of free muscle transplantation for facial paralysis. *Plast Reconstr Surg.* 1997;99:1905–1921.
15. Terzis JK, Olivares FS. Long-term outcomes of free-muscle transfer for smile restoration in adults. *Plast Reconstr Surg.* 2009;123:877–888.
16. Slingluff CL Jr, Terzis JK, Edgerton MT. The quantitative microanatomy of the brachial plexus in man: Reconstructive relevance. In: Terzis JK, ed. *Microreconstruction of Nerve Injuries*. Philadelphia: Saunders; 1987:285–324.
17. Bonnel F. Microscopic anatomy of the adult human brachial plexus: An anatomical and histological basis for microsurgery. *Microsurgery* 1984;5:107–117.
18. Dykes RW, Terzis JK. Spinal nerve distributions in the upper limb: The organization of the dermatome and afferent myotome. *Philos Trans R Soc Lond B Biol Sci.* 1981;293:509–559.
19. Herringham MB. The minute anatomy of the brachial plexus. *Proc R Soc Lond.* 1887;41:423–441.
20. Terzis JK, Karypidis D. Direct tongue neurotization: The effect on speech intelligibility in patients with Moebius syndrome. *Plast Reconstr Surg.* 2010;125:150–160.
21. Rubin LR. The Moebius syndrome: Bilateral facial diplegia: A discourse on the symptoms, pathology, and a surgical dynamic animation of the paralyzed face. *Clin Plast Surg.* 1976;3:625–636.
22. Zuker RM, Goldberg CS, Manktelow RT. Facial animation in children with Möbius syndrome after segmental gracilis muscle transplant. *Plast Reconstr Surg.* 2000;106:1–8; discussion 9.
23. Terzis JK, Konofaos P. Nerve transfers in facial palsy. *Facial Plast Surg.* 2008;24:177–193.
24. Gu Y, Xu J, Chen L, Wang H, Hu S. Long term outcome of contralateral C7 transfer: A report of 32 cases. *Chin Med J (Engl).* 2002;115:866–868.
25. Lifchez SD, Matloub HS, Gosain AK. Cortical adaptation to restoration of smiling after free muscle transfer innervated by the nerve to the masseter. *Plast Reconstr Surg.* 2005;115:1472–1479.
26. Merzenich MM, Jenkins WM. Reorganization of cortical representations of the hand following alterations of skin inputs induced by nerve injury, skin island transfers, and experience. *J Hand Ther.* 1993;6:89–104.
27. Beaulieu JY, Blustajn J, Teboul F, et al. Cerebral plasticity in crossed C7 grafts of the brachial plexus: An fMRI study. *Microsurgery* 2006;26:303–310.