

## Physiotherapy in the first three months after allograft face transplantation – case report

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### Abstract

**Objective:** The aim of the study was to present physiotherapy procedures of the patient with von Recklinhausen's disease in the first three months after allograft face transplantation (PT).

**Methods:** During hospitalization lasting 2.5 months intensive functional rehabilitation was conducted. It ran in two stages: early (up to 30 days after surgery) and recovery (1 to 3 months). In physiotherapy the method with elements of PNF (Prioprioceptywne Neuromuscular Facilitation) was used. During 2 months from PT an electrostimulation of orbicularis oris muscles was included. Physiotherapists did not focus only on rehabilitation of the face transplant, but also paid attention to functional problems caused by von Recklinhausen's disease. When planning therapy, they took into consideration personal, physical and emotional features. In therapy we used exercises in different starting positions, such as rollers, elastic bands, balls and sensory rings were employed. Lymphatic drainage of the face and neck was performed.

**Results:** During three months of the rehabilitation an improvement of upper and lower limbs' muscle strength was achieved; cervical spine range of motion increased, pain and discomfort caused by scar restrictions were reduced. There was also an improvement of the overall functionality of the patient. However, due to short time after PT no improvement in the functioning of facial expressions and sensory feeling was noticed, nevertheless the patient accepted the new face. In our opinion, rehabilitation is an inevitable element after face transplant (PT). The use of an individual and comprehensive program of physiotherapy, and it's early and systematic implementation allow to achieve an improvement of the patient's physical function after PT. Continuing rehabilitation is necessary to achieve full facial functionality and maintain physical capacity.

**Key words:** neurofibromatosis type I, face transplantation, rehabilitation

### Introduction

In this article treatment proceedings in the first three months after allograft face transplant in a female patient with a diagnosis of neurofibromatosis type I were presented.

Neurofibromatosis type I (von Recklinhausen's disease) is a disease from the phacomatoses group (Greek "phakoz", i.e. marked at birth); it is genetically determined, and autosomally and dominantly inherited. It occurs once in every 2,500-3,500 people with identical frequency in both sexes and all races. The course of disease varies in different patients. Anomalies of the skin and subcutaneous tissue, the central nervous system, within the musculoskeletal system, endocrine, ophthalmic and cardiovascular disorders, and anomalies within the gastrointestinal tract are typical for this group of patients. In the patients there is also an increased risk of malignant tumours of soft tissues [1]. Typical symptoms include brown

spots (color: coffee with milk) spread on the skin of the trunk and limbs, and freckled spots with a 2-3mm diameter in the area of armpits and groins. In young people and adults subcutaneous nodules, which in the histological examination show hematoma or neurofibroma tissue, are also observed. In 30-50% of patients skeletal disorders, most frequently bone loss due to the oppression of neurofibroma, scoliosis, and congenital bent long bones disease are observed. Epilepsy occurs in 10-15% of the patients, and delayed psychomotor or mental development – in 15-20%. In approx. 70% of patients optic nerve gliomas develop; brain and spinal cord meningioma, as well as astrocytomas can also occur [2].

An indication to perform allograft face transplant includes serious, complex facial deformations or trauma in cases where it is not possible to apply any other treatment methods restoring lost functions, aesthetic appearance and well-being of the patient. The procedure of allograft face transplant requires principles of microvascular surgery and organ transplantation to be combined as well as application of immunosuppressive therapy the aim of which is to prevent transplant rejection.

### Abbreviations:

PT – allograft face transplantation

The first successful partial facial transplant was performed in 2005 in France [3]. Within the last 9 years thirty-two facial transplantations in 25 men and 7 women were conducted throughout the world. The amount of transplanted tissue varied: in 16 cases it was partial transplantation, and in the remaining 16 cases the entire face was transplanted.

During the initial post-surgery period, due to transplantation of muscle, vascular and nerve structures, the patient does not feel the new face. Within the transplant exteroceptive sensation is disturbed or suppressed, and the brain still “sees and remembers the old face”, it does not reflect the new one [4-6]. Long hours of surgery, anaesthesia procedures and immobilization in bed decrease overall fitness and exercise capacity of the patient. Additionally, pain and swelling disturb patterns of movement and motor control throughout the movement system.

A key element of treatment of the patient after facial transplantation is rehabilitation, an important component of which is physiotherapy [7, 8]. The ultimate goal of rehabilitation is to restore optimal physical, mental and social functioning of the patient. Regarding physical functioning the goal is to restore the following functions: chewing, swallowing, and smell, speech and facial expressions. It is equally important to restore and maintain the highest possible level of functioning of the entire body, to eliminate or decrease unwanted movement patterns and other effects of immobilization, to support healing of tissue in the area of the transplant. Obtaining the proper performance and physical fitness positively impacts the mental condition of the patient which translates into improvement of their quality of life.

**Case report**

A woman, age 29, suffering from neurofibromatosis type I since her birth with advanced destructive craniofacial changes: head and neck external surfaces were covered in full by neurofibromas, came to the Department of Oncological and Reconstructive Surgery of the Oncology Institute. The patient was visually impaired and she had troubles moving independently. Due to large tumours, deformities of the face and mouth the patient had problems with breathing, talking, eating and drinking. The patient did not have an active movement of closing her mouth. It was impossible to evaluate the facial muscles due to deformation. The size and severity of changes caused compensatory setting of the shoulders in elevation, large muscle tension of the shoulder girdle manifested in activation of trigger points of the levator scapulae muscle and trapezius muscles of the ridge.

Earlier the patient underwent many surgeries of her facial soft tissue which were not performed by the Oncology Institute. She underwent 36 plastic surgeries involving partial resections of deformed facial external surfaces which did not result in the expected functional and aesthetical effect (Fig. 1).

Because it was not possible to apply conventional reconstructive surgical methods improving the patient’s quality of life, in October 2013 she was qualified by the Team for Facial Transplantation of the Oncology Institute in Gliwice for allograft face transplantation (PT).

The surgery (PT) was conducted in December 2013. During surgery which took 23 hours resection of the affected soft facial tissue at all levels (skin, blood vessels of muscles and nerves) was performed with concurrent reconstruction through allogeneic multi-tissue whole face transplant collected from a dead donor in the same scope. Microanastomoses of the external carotid arteries of the donor with the facial arteries of the recipient as well as facial veins with internal carotid veins were performed bilaterally. Additionally, on the right



Figure 1. The patient’s face before transplant

the retromandibular vein was connected with the external jugular vein. Also the nasal cartilage frame of the donor was connected with the margin of the recipient’s nose. Bilaterally the facial nerves were micro-combined within the branches, and mental nerves of the donor with the great auricular nerves of the recipient.

After surgery the patient was hospitalized for 2.5 months, during which intense functional rehabilitation took place consisting of two stages: early stage up to 30 days after surgery and recovery stage from 1 to 3 months. The treatment program is presented in tables no. 1 and 2.



Figure 2. Exercises of the upper and lower limbs and the torso using three dimensional resistance – transferring tension (irradiation) on the facial muscles



Figure 3. Exercising control of proper setting of the head

Table 1. Physiotherapy program in the early period after the face transplant

	Goals of physiotherapy	Applied therapeutic techniques
Early rehabilitation (1-30 days)	Preventing negative effects of immobilization	<ul style="list-style-type: none"> <li>– Changing positions (active and passive)</li> <li>– Breathing exercises: through the chest and diaphragm, tapping</li> <li>– Assisted exercises of the upper and lower limbs</li> <li>– Active exercise of the upper and lower limbs</li> </ul>
	Transplant protection	<ul style="list-style-type: none"> <li>– Active learning to change positions by the patient from lying on the back to lying on the stomach with head compensation and returning to the initial position – learning to protect her face</li> <li>– Learning to protect her face, including touch (the patient delicately touched her new face)</li> </ul>
	Verticalization	<p>On the fifth day after surgery the patient started to learn to sit</p> <ul style="list-style-type: none"> <li>– Exercises described above were continued</li> <li>– Learning to change from the position of lying on the side to sitting with lowered legs and returning to the initial position</li> <li>– Exercises restoring proper setting of the pelvis, head, shoulder girdle when sitting down</li> <li>– Exercises stabilizing the torso in the sitting position</li> </ul>
	Facial sensorimotor re-education	<ul style="list-style-type: none"> <li>– Exercising physical memory within facial muscles (the patient received recommendations to imagine her facial expressions occurring in the case of expressing feelings and emotions, among others: joy, laughter, surprise, anger, smelling pleasant and unpleasant odours)</li> </ul>
	Re-education of gait	<p>On the eighth day the patient started to learn to walk</p> <ul style="list-style-type: none"> <li>– Exercises described above were continued</li> <li>– Active exercises of the upper and lower limbs in the position of lying on the back applying three dimensional facilitating resistance</li> <li>– Exercises of changing positions: from sitting to standing</li> <li>– Exercises of alternating axial loading of the lower limbs</li> <li>– Learning to walk forwards</li> </ul> <p>On the fifteenth day the patient walked 1.5m for the first time</p> <ul style="list-style-type: none"> <li>– Improving walking forwards, walking backwards and sideways</li> </ul>
	Learning active movement of closing the mouth Learning correct operation of the tongue and facilitating the swallowing function	<ul style="list-style-type: none"> <li>– Assisted and self-assisted exercises of temporo-mandibular joints: projecting the lower jaw forward, moving the lower jaw from side to side, closing the mouth.</li> <li>– Sensorimotor exercises of the proper setting of the lower jaw</li> <li>– Exercising the tongue using facilitating resistance with the use of a spatula</li> </ul>

Table 2. Physiotherapy program in the period of 1-3 months after the face transplant

	Goals of physiotherapy	Applied therapeutic techniques
Recovery period from the 30 <sup>th</sup> day – 3 months	Mobilization of the postoperative scar and reduction of swelling of the face	– Fascial techniques, facial lymphatic drainage
	Re-education of the functions of mimic muscles	<ul style="list-style-type: none"> <li>– Passive and self-assisted exercises within facial muscles combined with exercises of facial expressions occurring during communication, expressing feelings and emotions</li> <li>– Exercises of lower and upper limbs, torso using three dimensional resistance to transfer tension on the facial muscles (Fig. 2).</li> <li>– Electrostimulation of the orbicularis oris muscle using unidirectional current. Triangular impulses lasting 20 ms (muscle chronaxie was 20 ms) was applied with an interval of 1 sec. 3.5 cm/2.5 cm electrodes were placed on opposite edges of the mouth so that current passes along the orbicularis oris muscle. Concentration of 12 mA was applied which caused visible muscle cramps. Treatment time was 2 min.</li> </ul>
	Strengthening of the muscles of lower and upper limbs	– Active and resistance exercises using elastic band
	Body posture correction	<ul style="list-style-type: none"> <li>– Activation of the transversus abdominis muscle</li> <li>– Control exercises of the proper head position (Fig. 3)</li> <li>– Control exercises of the proper scapula position (activation of the lower part of serratus anterior and the descending part of trapezius muscle)</li> <li>– Exercises for the correct position of the pelvis</li> </ul>
	Preventing limited mobility of the cervical spine, contractures of muscles of the neck and shoulder girdle	– Stretching and postisometric muscle relaxation: trapezius muscle – descending part, scapula levators, suboccipital muscles, scalene muscles, rhomboids, Sternocleidomastoid muscle (Fig. 4)
	Strengthening of the deep neck flexors	– Active exercises with resistance strengthening of the following muscles: longus colli muscle, longus capitis muscle, anterior rectus capitis muscle, lateral rectus capitis muscle, (Fig. 5)
	Exercises of the tongue and learning to swallow	– Exercises of the tongue with facilitating resistance using a spatula mobilization of the hyoid bone
	Improving gait, balance, coordination	– Balance and coordination exercises using sensorimotor cushions, balls, rollers



Figure 4. Stretching and postisometric relaxation of the scapula levators



Figure 5. Active exercises with resistance strengthening of the following muscles: longus colli muscle, longus capitis muscle, anterior rectus capitis muscle and lateral rectus capitis muscle

Treatment commenced on the fourth day after surgery after disconnecting the patient from the ventilator, with stable cardio-respiratory parameters. Physiotherapy sessions took place twice a day and they took 30-45 minutes each. The sessions were conducted in the patient’s hospital room due to her reduced immunity caused by administration of immunosuppressive drugs. Due to impaired vision the patient required constant assistance and support of a third person. In the initial period treatment was also impeded due to large swelling in the area of the transplant, lack of the movement of closing the mouth, and habitual compensatory settings of shoulders and trunk. Elements of the PNF (Proprioceptive neuromuscular facilitation) were used in the treatment procedure. Rehabilitation of the transplanted face was not the only focus, but also the use of the entire functional potential of the patient [9, 10]. Attention was drawn to positive commencement of the therapy through work without pain, and use of the strong parts of the body to stimulate the functional reserves of the patient. Planning the therapy physical and emotional features of the patient were taken into account. To teach movement the following were used: irradiation (transfer of the muscle tone from the strong parts of the body to the weakened parts), manual contract, verbal commands, facilitating resistance, relief (traction) and pressure (approximation) of the joint surfaces of limbs and torso.

In the therapy various starting positions were applied, using the force of gravity to facilitate or hinder movement. In exercises rollers, elastic bands, sensory rings and balls were used. Also lymphatic drainage of the face and the neck was applied as well as manual techniques on soft tissue. Two months after PT electrostimulation of the orbicularis oris muscle was additionally applied. The method of treatment was established based on electrodiagnostics results. The patient was also receiving psychological care.

Applying the described physiotherapy program during three months of rehabilitation the muscular strength of the upper and lower limbs was improved, the scope of mobility of

Table 3. Functional evaluation of the patient after 30 days and 3 months after surgery

	Evaluation of the functional condition of the patient after 30 days after surgery	Evaluation of the functional condition of the patient after three months after surgery
Muscle strength of upper limbs – Lovett’s test	4	5
Muscle strength of lower limbs – Lovett’s test	3+	4+
Facial muscles – Lovett’s test	0 no sign of tension within the facial muscles	0 no sign of tension within the facial muscles
Mobility of the temporo-mandibular joint	lack of active movement of closing the mouth	incomplete active movement of closing the mouth
Range of motion of the cervical spine using CROM device: extension-0-flexion	40-0-30	60-0-45
Range of motion of the cervical spine: left side bow-0-right side bow	15-0-15	30-0-30
Range of motion of the cervical spine: counterclockwise rotation -0- clockwise rotation	25-0-30	60-0-60
Pain of the neck and shoulders (VAS 0-10)	7 at the VAS scale	1 at the VAS scale
30-second Chair Stand CS	Impossible to perform – the patient gets up from the chair helping herself with her hands	5 repetitions
Eating	The patient was fed though PEG (percutaneous endoscopic gastrostomy), additional oral ingestion - semi-liquid diet, eating 100ml of food takes 25 minutes	The patient was fed though PEG (percutaneous endoscopic gastrostomy), additional oral ingestion, eating 200 ml of food takes 15 min
Gait	The patient walks independently (due to impaired vision she requires assistance) difficulties with walking backwards and sideways	The patient walks forwards, backwards and sideways independently (due to impaired vision she requires assistance)

the cervical spine was increased, pain and discomfort caused by tightening of postoperative scars were reduced. Also the general fitness of the patient improved (Table 3). No improvement was observed in the functioning of facial expressions and exteroceptive sensation due to too short a time after surgery. In spite of that the patient fully accepted her face.

The patient with her family was educated in the field of exercise techniques and discharged with a recommendation to continue rehabilitation under outpatient conditions with periodical follow-ups and functional evaluation in the Oncology Institute.

## Discussion

Allograft face transplantation (PT) is a new achievement in medicine and rehabilitation giving hope to people whose face was deformed by disease, injury or as a result of congenital defects of development [11].

In literature there are no publications presenting detailed schemes of physiotherapeutic proceedings of patients after allograft face transplantation. Devauchelle et al. (2006) describing the first successful partial face transplant say that in the patient whose face was severely bitten by a dog, when she waited for her partial transplant, intense physiotherapy was applied in order to decrease scarring within the damaged area of the face and to prevent atrophy of facial muscles of the preserved part of the face. The authors say that physiotherapy was also applied after PT for four months after surgery as of the second day. Therapy took place twice a day and it involved: passive and active exercising of the face mostly aimed at restoring mobility of the lips and closing the mouth [7, 8].

Also Roche et al. (2015) point out the necessity to apply early and intense rehabilitation in patients after PT involving: speech therapy, physical activity and sensory re-education [3].

Siemionow et al. (2009) say that physiotherapy in the patient after the total face transplant was commenced after 48 hours after surgery and it was conducted once a day for six weeks, and then three times a week. Active and passive physical exercises, delicate massage, sensory re-education and learning to accept the transplanted face were conducted. Functions, such as chewing, laughter, speech, swallowing and facial expressions were evaluated [6].

In our opinion in the early period after the PT surgery in the patient with neurofibromatosis type I, apart from direct re-education of functions within the transplanted face, it is important to improve functioning of the entire musculoskeletal system of the patient and using strong parts of her body to facilitate mobility within the transplanted tissue. Application of the presented treatment program allowed improvement of the general fitness of the patient to be achieved and conditions for further rehabilitation within the transplanted face to be created. The achieved improvement motivated the patient and her family to continue therapy.

A face transplant is a significant venture requiring cooperation of surgeons, transplant specialists, physical therapists, speech therapists, psychologists, a nursing team and many other specialists.

In treatment of patients after PT it is particularly important that the transplantation team takes into account the needs to conduct complex rehabilitation after surgery, and consider the possibility to commence it before the planned transplant.

In order to achieve fitness in the field of facial functions and maintain the correct fitness and physical capacity application of an individual and complex physiotherapy program is necessary. Physiotherapy proceedings require numerous studies and an exchange of experience, which might be impeded due to a small number of this type of procedures performed in the world.

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