

DYSARTHRIA

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Methods supporting logopedic therapy of patients with dysarthria in Moebius syndrome

SUMMARY

The aim of the article is to present neuromuscular methods supporting logopedic therapy for patients with dysarthria in Moebius syndrome and to describe Moebius syndrome: its etiopathogenesis, pathomechanism and symptomatology. The therapy methods which were presented in the article: logopedic kinesiotaping, orofacial regulation therapy described by C. Morales and electrical stimulation were characterized from the logopedic angles, taking account of different ways of how each method affects neuromuscular regulation. The use of these methods has an impact on the facial muscle tone regulation, improvement of the functioning of the articulators, mandibular mechanics, respiration quality, thereby contributing to diminishing the dysfunctions in the respiratory-phonatory-articulatory apparatus.

Keywords: Moebius syndrome, dysarthria, logopedic kinesiotaping, orofacial regulation therapy, electrical stimulation

INTRODUCTION

The aim of the article is to present methods supporting logopedic therapy of dysarthric patients in Moebius syndrome as well as to describe the syndrome (MBS), whose etiopathogenesis, identified about one hundred thirty years ago, still remains unclear. Factual sources show that in 1880 this pathological syndrome was probably first characterized, the scholar who gave his name to the syndrome being Paul Julius Moebius, German neurologist and philosopher, who indisputably described this problem in 1888–1892.¹

¹ Cudziło D., Obłój B., Obersztyn E., Bocian E., Matthews-Brzozowska T., 2012, *Moebius syndrome with facial-dental impairments – rare or rather seldom diagnosed syndrome?*, “Developmental Period Medicine”.

At present, neurologists define the disease as a malformation sequence whose specific feature is a series and multiplicity of anatomical anomalies resulting from a single developmental defect.² The syndrome is commonly called “a disease of persons without a smile”. This term refers to the most characteristic symptom of the disease: a partial or complete paralysis of cranial nerves that immobilizes facial movements, thereby preventing, inter alia, the ability to smile. The co-occurrence of numerous musculoskeletal anomalies, congenital defects that impede free functioning, is also reported.³ The Moebius syndrome is characterized by a diversified and wide range of clinical symptoms, particularly neurological aberrations, which makes it difficult to properly diagnose patients.

ETIOPATHOGENESIS OF MOEBIUS SYNDROME

Moebius syndrome is one of rare, complex congenital defects. The International Classification of Diseases ICD 10 classifies this disease entity under developmental defects pertaining mainly to facial appearance (Q.87.0)⁴.

According to the sources, it is estimated that Moebius syndrome occurs with an incidence of from 1:50 000 to 1:500 000 in live-born children of either sex.⁵ In their publications, the Dutch scholars who studied the problem, K. Stromlad and K. Sjogreen, presented a view that Moebius syndrome affects the male sex to a vastly greater degree, but this fact is called into question by scientific communities.⁶

Scholars are not entirely certain about the origin of the disease. There are many, often contradictory, theories. Currently, the most probable hypothesis about the etiopathogenesis of Moebius syndrome is the conception that suggests disorders in the development of primary subclavian arteries and their branches before the formation of vertebral arteries delivering blood to the brain at the early stage of embryogenesis⁷ because the mother took an abortifacient that does not abort the

² Cudziło D., Obłój B., Obersztyn E., Bocian E., Matthews-Brzozowska T., 2012, *Moebius syndrome with facial-dental impairments – rare of rather seldom diagnosed syndrome?*, “Developmental Period Medicine”.

³ Osborne G.A., 1999, *Moebius syndrome*, “JADA”, 130.

⁴ Zielińska A., 2006, *Zespół Moebiusa. Problemy terapii logopedycznej*, www.logopedia.org.pl, no 3.

⁵ Cudziło D., Obłój B., Obersztyn E., Bocian E., Matthews-Brzozowska T., 2012, *Moebius syndrome with facial-dental impairments – rare of rather seldom diagnosed syndrome?*, “Developmental Period Medicine”.

⁶ Strömmland K., Sjögren L., Miller M., Gillberg C., Wentz E., Johansson M., Nyllén O., Danielsson, Jacobsson C., Andersson J., Fernell E., 2002, *Mobius sequence – a Swedish multidiscipline Study*, “European Journal of Paediatric Neurology”.

⁷ Cudziło D., Obłój B., Obersztyn E., Bocian E., Matthews-Brzozowska T., 2012, *Moebius*

fetus but damages it. These convictions are based on the results of studies made on animal models and the examinations conducted using methods of neuroimaging of the brain. None of the presented theories, however, has been confirmed by scholars studying Moebius syndrome.⁸

In the United States, research is conducted on the extent of participation and role of genetic factors in the pathogenesis of the disease. Many theories assume that there are multiple factors underlying Moebius syndrome although it cannot be ruled out that the disease is monogenetically determined, which means mutations in a specific gene.⁹ Genetic factors could play a role in the etiology of hypoplasia, or underdevelopment of an organ, which is frequently accompanied by impairment or aplasia, agenesis of an organ with the formation of its germ, nuclei of cranial nerves.¹⁰

Moebius syndrome is an incurable disease: what can largely enhance the patient's comfort is a surgical operation, which, however, is complicated and involves substantial expenses. It consists, inter alia, in transplanting muscles into an immobile area of the face, combining muscles together and making muscular plexuses function as efficiently as possible. Operations of this type are not performed in Poland; at present they can be carried out exclusively abroad, e.g. in the Czech Republic, Germany or the USA.

CLINICAL SYMPTOMS AND PATHOMECHANISM OF MOEBIUS SYNDROME

Moebius syndrome, as a malformation sequence, induces many neurological and anatomical abnormalities, which, consequently, produces a wide range of clinical symptoms, thereby making it difficult to identify and correctly diagnose the disease. The most characteristic feature of Moebius syndrome is a unilateral, bilateral and complete paralysis of cranial nerves responsible for facial expressions, blinking, and lateral movements of eyeballs, which often occur together with dysfunctions of the other cranial nerves such as:

- oculomotor nerve III,
- trochlear, innervating superior oblique muscle I in the orbit,

syndrome with facial-dental impairments – rare of rather seldom diagnosed syndrome?, “Developmental Period Medicine”.

⁸ Cudziło D., Obłój B., Obersztyn E., Bocian E., Matthews-Brzozowska T., 2012, *Moebius syndrome with facial-dental impairments – rare of rather seldom diagnosed syndrome?*, “Developmental Period Medicine”.

⁹ Osborne G.A., 1999, *Mobius syndrome*.

¹⁰ Cudziło D., Obłój B., Obersztyn E., Bocian E., Matthews-Brzozowska T., 2012, *Moebius syndrome with facial-dental impairments – rare of rather seldom diagnosed syndrome?*, “Developmental Period Medicine”.

- trigeminal sensorimotor nerve,
 - ophthalmic nerve, which sensorily innervates the skin of the head above the palpebral fissure and the lateral nasal surface,
 - maxillary nerve, which sensorily innervates the skin below the maxillary fissure and above the oral fissure,
- mandibular sensorimotor nerve, which innervates the skin below the oral fissure and the temples, and innervates masticatory motor muscles,
- glossopharyngeal nerve IX, with sensorimotor innervations of the pharynx and tongue,
 - vagus nerve X, the longest cranial nerve, which conducts sensory, motor and parasympathetic fibers.¹¹

In consequence of damage to these nerves the face becomes mask-like and expressively inefficient. Patients are unable to show even the simplest emotions such as laughter or crying. The malfunctioning of the facial skeleton is accompanied first of all by difficulties in the speaking function. Disorders that are part of the picture of Moebius syndrome also embrace:

- eye hypersensitivity,
- inability to narrow the eyes,
- complete lack of eyeball movements or difficulties in this area,
- characteristic slanting eyes,
- occurrence of squint,
- short or deformed tongue,
- limited tongue movements, lack of tongue elevation,
- high palatal vault,
- cleft palates,
- hearing problems,
- syndactyly (fusion of fingers or toes),
- adactyly (congenital lack of fingers or toes),
- brachydactyly (shortness of fingers or toes),
- ectrodactyly, (fusion of both fingers and toes - lobster claw syndrome),
- varus deformity of lower limbs,
- congenital heart defects,
- defects of the urinary system,
- underdeveloped pectoral muscle,
- anosmia, or total loss of the sense of smell,
- dental-occlusal disorders,
- muscle tone disorders.¹²

¹¹ Cudziło D., Obłój B., Obersztyn E., Bocian E., Matthews-Brzozowska T., 2012, *Moebius syndrome with facial-dental impairments – rare or rather seldom diagnosed syndrome?*, “Developmental Period Medicine”.

¹² Cudziło D., Obłój B., Obersztyn E., Bocian E., Matthews-Brzozowska T., 2012, *Moebius*

Apart from the characteristic dysmorphia, in Moebius syndrome there is a likelihood of psychomotor retardation, yet cases of accompanying mental retardation are comparatively few.

The lack of knowledge about Moebius syndrome often results in an erroneous assessment of the patient; dysmorphia problems and difficulties in verbal communication with the environment cause overinterpretation of the patient's intellectual problems.

SPEECH DISORDERS IN MOEBIUS SYNDROME

The main difficulties faced by Moebius syndrome patients are neuromuscular disorders; they manifest themselves in speech at the articulatory-phonatory-prosodic level, being connected with the diagnosis of dysarthria, dysglossia and dysphagia in those patients.

Because of the paralysis of nerves V, IX, X, and XII, which control the functioning of the muscles of the mandible, tongue, pharynx and larynx, the most frequently found disorders that affect the quality of communication are:

- difficulties in primary functions, respiration, sucking, chewing, which influence the development of orofacial reflexes,
- difficulties associated with the motor activity of speech organs, affecting articulation quality,
- difficulties associated with mandibular mechanics, causing hypersalivation (excessive salivation)
- swallowing difficulties,
- respiratory difficulties, disorders of laryngeal functions which affect the process of phonation,
- disorders of the tension of facial expression muscles, decreased muscle tone affecting the quality of functioning of the articulators,
- cleft palates affecting the quality of the phonic substance at the segmental and suprasegmental levels,
- maxillary-occlusal defects, which affect the quality of articulation .

Due to the progressive nature of Moebius syndrome and the escalation of difficulties connected with neuromuscular disorders, it is vital to quickly begin appropriate therapeutic management, particularly speech therapy.

Methods supporting logopedic therapy of dysarthric patients in Moebius syndrome: logopedic kinesiotaping, orofacial regulation therapy, electrostimulation

At present, in the logopedic treatment of patients with dysarthric disorders that are part of the picture of Moebius syndrome, where difficulties manifest

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themselves mainly in neuromuscular functioning, alternative supporting methods are used more and more often, inter alia logopedic kinesiotaping, orofacial regulation therapy or muscle electrostimulation, which, by strengthening facial expression muscles or by regulating the facial muscle tone, improve the functioning of the articulators, mandibular mechanics and respiration quality, thereby enhancing communication.

The method of logopedic kinesiotaping is a world-recognized form of therapy that uses specialist cotton tapes, their extensibility being comparable with the extensibility of human muscles. The appropriate tension strength of tapes plays an essential role in particular techniques of applying tapes which, through contact receptors, impact on the proprioceptive system. Applications are performed, depending on the set goal, using the muscle, ligament, corrective or lymph techniques, with an appropriate tension along the length of the whole tape, its base, and its shape, a fan-like form Y, X and a single form I. In logopedic kinesiotaping the most often used application technique is the functional-muscular technique, which is used in cases of hypertonia, hypotonia, and in muscle injuries, the tape being applied depending on a specific goal, from the base to the insertion or reversely, from the insertion to the muscle base, with adequate tension. Logopedic applications require not only the knowledge of the facial nerves but also the nerves active in the respiratory process. The logopedic kinesiotaping method is utilized inter alia in :

- hypersalivation – logopedic application strengthens the tension of the masseter, and zygomatic, digastric muscles, and the orbicular muscle of the mouth, which inter alia improve mandibular mechanics, closing of the mouth, the quality of swallowing ,
- improvement of facial muscles: the application regulates tone: detoning – reduction of tone or toning – strengthening of the muscle tone which enhances the quality of functioning of the articulators and the mandible ,
- dysfunction of the temporomandibular joint: appropriate application is used to improve the functioning of the mandible ,
- dysphagia – appropriate application can lower, “draw back the larynx”, thus improving the swallowing process,
- regulation of the respiratory pattern: applications are used on breathing muscles, thereby deepening breathing and regulating the correct posture.

Contraindications against the use of logopedic kinesiotaping therapy are :

- allergy to acrylic, which coats the tapes on the inside,
- unhealed wounds in the application area.¹³

¹³ Kumbrink B., 2013, *K-taping w logopedii*, Dortmund.

An important aspect of logopedic kinesiotaping is the possibility of prolonging the effect upon muscles by means of an application, which actively stays on a fixed skin area until the tape comes unstuck, which enables faster therapeutic effects.



Muscle application, toning of the zygomatic muscle



Muscle application, toning of the orbicular muscle of the mouth

For esthetic reasons and depending on the need to support muscles, logopedic applications, usually beige-colored, are used in the course of logopedic therapy, thereby supporting muscle performance during therapy classes, or prolonging the process of interaction on the muscle – by leaving the tape on the skin until it comes unstuck.



Muscle application, detoning of the masseter in the course of the temporomandibular joint

Another method used in patients with neuromuscular disorders is Castillo-Morales orofacial regulation therapy. It is a neurophysiological method using electric stimulation, developed in the 1970s by the Argentine rehabilitation physician Dr Rodolfo Castillo Morales. The method is used especially in neurological diseases, neuromuscular paralyses. The reference point of the orofacial therapy is a function understood as activity, interaction of particular parts of the orofacial complex, which enables coordination of the functioning of the six elements of Morales method:

- movement understood as a function, at the level of biochemical reactions and changes of body position,
- matter understood as the instrument of activity arising inter alia within muscles, nerves, and receptors,

- time, referring to the stage between the beginning and ending of function,
- space as favorable conditions necessary for a function to occur,
- energy understood as motivation for therapy,
- regulation pertaining to regulation mechanisms that ensure interaction at the histochemical and biochemical levels.¹⁴

The therapeutic technique involves stimulation in different sensory systems, utilizing such stimuli as touching, stroking, pulling, pressure, and vibration used in a specific internal or external sequence of movements which impact on individual muscles through the activation of selected neuromotor points (i.e. the sites where a nerve penetrates into a muscle) in order to inter alia normalize muscle tension, mold orofacial reactions, regulate mandibular mechanics, and improve the respiratory function.¹⁵

Another form that supports the therapy of dysarthria in Moebius syndrome is electrostimulation using appropriate electrical impulses conducted through electrodes, usually placed in pairs, between two attachments of a muscle in order to stimulate it. Depending on the number of electrostimulation channels, therapy can be conducted simultaneously on a large muscle area. After ruling out contraindications against therapy, an electrical impulse with suitable amplitude is applied as a series of 20 impulses onto fixed motor points in the facial area; with a severe nerved paralysis the impulse time is ca. 150–400 ms/ with a 1000–3000 ms interval.¹⁶ During the electrostimulation of small muscles, as in the case of the facial muscles, the active electrode, connected to the negative pole of the source of current, impacts on a particular point, stimulating the selected muscle, causing its contraction and strengthening.



Application of a single electrode in the area of the oral orbicular muscle

The purpose of electrostimulation is to improve communication by strengthening the paralyzed facial muscles with an appropriate electrical impulse that affects the quality of the functioning of the articulators, mandibular mechanics, and the quality of swallowing, which is improved through electrostimulation of the larynx and the posterior wall of the pharynx, using specialist ball electrodes.

¹⁴ Morales R. C., 2009, *Ustno-twarzowa terapia regulacyjna*, Wrocław.

¹⁵ Morales R. C., 2009, *Ustno-twarzowa terapia regulacyjna*, Wrocław.

¹⁶ www.rehabilitacja.pl, portal rehabilitacji medycznej.

CONCLUSIONS

Currently, the speech therapy treatment of dysarthria is oriented towards enhancing the quality of communication; it uses special measures that improve respiratory-phonatory-articulatory functions by the application of, inter alia, alternative methods of neuromuscular support.

In terms of speech therapy, the presented methods are meant to improve communication through different ways of neuromuscular stimulation; the method of logopedic kinesiotaping uses adequate tension and the technique of tape application; the Castillo-Morales neurodevelopmental concept presents the manual regulation by selecting the appropriate sequence of movements and types of touching neuromotor points, whereas electrostimulation uses appropriate voltage, thus intensively contributing inter alia to strengthening selected muscles.

In the treatment of speech disorders arising from dysfunctions of the peripheral or central nervous system, logopedic therapy increasingly often carries out the principle of complex regulations by combing the conventional approach with the supporting methods of muscular neurostimulation such as electrostimulation, neurophysiological manual techniques or logopedic kinesiotaping. As there are many difficulties in the treatment of dysarthria, a significant aspect is the individual approach to the patient's therapeutic needs by selecting one technique or combination of techniques, thus enabling faster achievement of the expected therapeutic effects.

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