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Neurologopedic Therapy in a Child with Moyamoya Disease

Terapia neurologopedyczna dziecka z chorobą moyamoya

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Abstract

Introduction. Moyamoya is a rare disease of unknown etiology which leads to strokes resultant from occlusions of intracranial arteries. As a result of the blockage of the arteries in the brain a lateral network of blood vessels develops, forming a characteristic angiographic image.
Case Report. In the paper a case is presented of a child who in the course of a month has suffered two strokes in two cerebral hemispheres. The damage in the central nervous system led to paresis in four limbs, speech impairment (later: lack of speech), problems with swallowing, and limited visual and audial contact.
Discussion. The main aim of the therapy was to improve feeding, drinking, and chewing and an attempt to introduce alternative communication. Elements of sensory integration were employed, and regulatory therapy of Castillo Morales was used to allow swallowing, shutting the mouth fully, and controlling the mandible while eating and drinking. Furthermore, the child's agility was being simultaneously enhanced the motor skills rehabilitation, the child would also benefit from a Room of World Experiences. Contrary to the initial assumptions, it has not been possible to reach all the aims of the therapy. Epileptic seizures have led to the loss of the acquired skills.
Conclusions. Successfulness of therapy is dependent on a number of factors, which are often independent of the therapist. What is crucial is to adjust the level of the difficulty of the sessions and their duration to the child's state of feeling on a particular day. After the conclusion of the programme improved eyesight was noticed. However, in the case of a child with such vast damage to the central nervous system the aim of the therapy is to retain the acquired skills and to carry out further attempts to develop new skills through their frequent repetition. **(JNNN 2018;7(2):75–79)**
Key Words: moyamoya, logopedic therapy, speech disorders, communication disorders, case report

Streszczenie

Wstęp. Moyamoya to rzadka choroba o nieznanej etiologii, prowadząca do udarów niedokrwiennych, na skutek niedrożności tętnic wewnątrzczaszkowych. Wskutek zamknięcia tętnic w mózgu rozwija się poboczna sieć naczyń krwionośnych, które składają się na charakterystyczny obraz angiograficzny.
Opis przypadku. W pracy przedstawiono przypadek dziecka, które w przeciągu miesiąca przeszło dwa udary w obu półkulach mózgu. Uszkodzenia ośrodkowego układu nerwowego doprowadziły do niedowładu czterech kończyn, zaburzeń mowy (później jej braku), problemów z połykaniem, ograniczenia kontaktu wzrokowo-słuchowego.
Dyskusja. Głównym celem terapii było usprawnienie karmienia, picia i żucia oraz próba wprowadzenia komunikacji alternatywnej. Zastosowano elementy integracji sensorycznej, wykorzystano terapię regulacyjną Castillo Moralesa w celu uzyskania połykania, domykania buzi, kontroli żuchwy w trakcie karmienia i picia. Dodatkowo dziecko było równolegle usprawniane rehabilitacją ruchową, korzystało z Sali Doświadczeń Świata. Wbrew początkowym założeniom nie udało się osiągnąć wszystkich celów terapii. Napady padaczkowe spowodowały utratę osiągniętych umiejętności.
Wnioski. Sukces w prowadzeniu terapii jest zależny od wielu czynników, często niezależnych od terapeuty. Ważne jest dostosowanie poziomu trudności zajęć, ich długości do możliwości i samopoczucia dziecka w danym dniu. Po zakończeniu programu stwierdzono poprawę kontaktu wzrokowego. Jednak w przypadku dziecka z tak bardzo uszkodzonym centralnym układem nerwowym celem terapii jest już utrzymanie uzyskanych umiejętności i dalsze próby wypracowania kolejnych, poprzez ich wielokrotne powtarzanie. **(PNN 2018;7(2):75–79)**
Słowa kluczowe: moyamoya, terapia logopedyczna, zaburzenia mowy, zaburzenia komunikacji, opis przypadku

Introduction

Moyamoya is a rare progressive disorder in the formation and proper functioning of cerebral blood vessels. The disease can be congenital or acquired [1]. It is characterized by a progressing occlusion of major intracranial arteries, in particular, the final section of the internal carotid artery, the middle cerebral artery, and the anterior cerebral artery, leading to their compete blockage [2]. As a result, a lateral network of blood vessels develops in the brain, forming a characteristic angiographic image [3]. The disease afflicts mainly individuals of Asian ethnicity [4]. It is most frequently diagnosed in Japan [5]. Causes have not been found, it has been noted, however, that in 7% of the cases they are familial (in persons of Japanese origin up to 10%), which suggests genetic predispositions [6]. Comorbidity of the moyamoya disease is frequently reported [7].

The therapy of a child afflicted by moyamoya is long and difficult. Its difficulty depends to a great extent on the age and the condition of the patient and to the cerebral damage suffered as the result of a stroke or strokes. Independently of age, cerebral ischemia results in prolonged or permanent disorders which influence the functioning [8]. Children who have suffered a cerebrovascular accident require rehabilitation involving a number of specialists, including a neurologist, a physiotherapist, a speech and language therapist, and a psychologist. In working with the children after a stroke the following methods are employed: the Vojta therapy, the NDT-Bobath method, the SI therapy, the orofacial regulatory therapy of R. Castillo-Morales, the Veronica Sherborne Developmental Movement, and proprioceptive facilitation [9].

The aim of the paper is to present speech and language therapy proceedings (diagnosis and programme of the therapy) in the case of a child with the moyamoya disease who suffered two cerebral strokes in both hemispheres within the period of a month.

Case Report

A girl delivered through a cesarean section from a high-risk pregnancy (threat to pregnancy starting in the fifth month) developed correctly until the 30 month of life when speech impairments appeared — slurring speech and facial asymmetry: lowering of the left corner of the mouth with smoothing of the nasal-temporal fold. The symptoms subsided on the same day. The situation recurred on the following day. On a subsequent day, two incidents occurred as well as partial paresis of the right upper limb. On the following day paresis of the lower right limb was observed.

The child was admitted to a pediatric neurology ward. In the hospital, the child was lucid, did not report any ailments, and did not run a fever. The parents reported that about three weeks previously the child had fallen from the height of ca. 40 cm, hitting the occipital part. CT and MRI scans were conducted in which ischemia of the left hemisphere of the brain was discovered. Subsequent tests discovered a progression of the changes. In the following days of the hospitalization neurological incidents occurred in which facial paresis, as well as pareses of the right upper and lower limbs, were observed. Five days after hospital admission the condition of the child suddenly worsened. Stroke of the left hemisphere occurred, which led to paralysis of the right side of the body and motor aphasia. In the hospital it was suspected that the ischemia of the left hemisphere was caused by an infection, probably borreliosis. Tests allayed these suspicions.

A month later the girl was again admitted to the Pediatric Neurology Ward with mild paresis in the upper left limb. The image generated in an MRI scan presented a new ischemia in the frontal lobe. A new stroke had occurred, this time in the right hemisphere of the brain. After about four hours the symptoms of the paresis of the left limb subsided. However, they returned exacerbated two days later. The child, unconscious and in a grave state, was admitted to the Neurology Clinic in Warsaw. The following were certified: bilateral spastic paresis, pyramidal signs, trismus, lip-smacking movements, impaired swallowing, opening of the mouth, muscle contractions in arms and legs, lack of visual and audial contact, weakened reaction to touch, deflected position and tension of limbs and trunk, motor and sensory ataxia.

Diagnostic tests indicated the moyamoya disease as the cause of the strokes. The girl underwent systematic motor therapy. Her condition improved slightly during her hospital stay, she began to swallow unassisted, however, she remained tube-fed. Improved contact with the environment and decreased extrapyramidal signs were observed. The recumbent patient with bilateral spastic paresis was transferred to a rehabilitation clinic. Half a year after the onset of first symptoms a encephaloduroarteriosynangiosis, i.e., revascularization — removing the blockages and widening the narrowed arteries in both the hemispheres of the brain was performed. After the surgery, the girl was conscious, with a visual fixation for brief periods. Aphasia, heightened muscle tension in all limbs, bending position of upper limbs, tightened fists, hyperextension of lower limbs, and clubfeet were observed. Nine days after the surgery the girl was in good condition, with proper vascular and respiratory functions, and she was discharged for further treatment at home.

Rehabilitation Procedures

After the surgery, the girl was entered into motor and speech therapy. The aims of the motor therapy were to improve control of the head and the trunk in the sitting position, as well as regulation of the muscle tension in the limbs and in the trunk. In the speech therapy focus was placed on sensory stimulation, stimulation of hearing, touch, and sight, as well as comprehension of simple instructions to be carried out passively. After four months of intensive four-hour therapy sessions, a slight improvement of the control of the head and the trunk was achieved, and the girl's ability to follow objects with her eyes increased. After speech therapy sessions stimulation of the senses was practiced, along with understanding simple instructions and introduction to nonverbal communication. Polysensory stimulation, elements of speech therapy massage, comprehension exercises supported by audial and motor reinforcement, biting, chewing, and swallowing practice, as well as orofacial regulatory therapy were employed. One could observe spontaneous smiles, general improvement in agility (particularly of the hands), attempts to react to audial and visual stimuli, slight improvement in eye contact, attempts to position the mouth to produce sounds "a" and "o", as well as vocalization of the "a" sound as echolalia, and improved reaction to verbal messages. In 2012 epileptic attacks took place, which led to a return of the patient's state previous to therapy.

Speech and Language Pathology Diagnosis

Speech and language pathology diagnosis were conducted on the basis of medical documentation, an interview with the parents, and observation of the child. In the course of the examination, the girl was 10 years old and she was undergoing individual education in an integrated private preschool where she was monitored during meals, sessions with a physiotherapist, and activities involving other children. On this basis dependent impaired speech development as a result of the ischemic stroke of the brain, severely impaired the development of passive speech, and lack of active speech was noted. Neurogenic dysphagia was observed as well. Primary actions of biting, chewing, and swallowing were impaired. The sucking and biting reflexes remained. The movements of the tongue are reduced (movements to the front and to the back, no lateral movement). These impairments make it difficult to properly collect food from a spoon, to form and move bites of food, to drink out of a cup, and to swallow. The low tension of the orbicularis oris is persistent, and the mouth is open. Salivation and lockjaw recur, making feeding difficult. Flaccidity of the esophagus is observed, which is dependent on the antiepileptic medication. This leads to choking. The girl cannot cough up the mucus from the respiratory tract. This causes vomiting and frequent infections which weaken the organism. The parents have attempted to remove mucus through the nose with an aspirator, which turned out to be ineffective.

The tendency to breathe with the top of her lungs is problematic because it allows the girl to only partially fill the lungs with air. The breath is deepened in stress situations exclusively, but the breathing phase is not prolonged. The child is unable to independently perform the abdomen and diaphragm reflex because of the sitting position in which she is placed for most of the day and because of the flaccidity of thorax muscles. The girl breathes through her mouth. Breathing through the nose is made difficult by her small nostrils which have not been stimulated since the onset of the disease; consequently, the girl has not breathed through her nose for 7 years.

The Programme and the Course of the Therapy

Therapy sessions with the child were conducted daily. In the speech and language therapy programme particular emphasis was placed on primary functions, improvement of the quality of breathing and eye contact. Sensory stimulation and elements of face massage were employed and attempts to introduce alternative communication were made.

Therapy feeding training was conducted daily during the meals in the preschool. The position of the body of the child was made correct by the rehabilitation chair which secures the proper position of the spine and the pelvis. The consistency of the food was smooth and pulpy. The child would eat slowly, the food was administered in small amounts. The consumption of a meal consisting of two courses, e.g., a small bowl of soup and two dumplings took 30 minutes. When about a month has passed it was prolonged to 40 minutes. When the girl would chew harder foods, such as pieces of potatoes, a banana, a baked apple, or biscuits, she received them in a way allowing her to chew with the molars, using both sides interchangeably. In every meal techniques facilitating administration of food, biting, swallowing, and control of the mandible were used. Drinking from a Medela cup was introduced.

The nasal stimulator (MFS) was introduced to the therapy programme as a first step to facilitate breathing through the nose. Originally the child would use the stimulator for 5 minutes. The duration was prolonged daily. After a week and a half, the stimulation had to be discontinued because of fissures in the epidermis of the openings of the nostrils. After the epidermis healed

an attempt was made to reintroduce the stimulation. Ultimately it was decided that the stimulation is discontinued because it had led to significant painfulness.

External face massage was combined with relaxation exercises. The child was highly hypersensitive in the orofacial sphere. It was not frequently feasible to perform a full massage. The relaxation exercises were mainly conducted in moments of increased muscle tension.

Tactile stimulation was administered with the use of elements of sensory integration and therapy of the hand. Massage of the hand was typically conducted with fabrics of various texture, opening spastic hands while playing with kinetic sand, submerging hands in water, groats, rice, noodles, hand painting, placing clips on fingers. In audial stimulation attempts were made to trigger a reaction to the girl's name, with varied results, to localize the source of sounds, e.g., with acoustic boxes and musical instruments. Following objects with the eyes and prolonging concentration were practiced. Speech comprehension practice was also included in the programme.

Attempts of alternative communication were made through the formation of the notion of "yes/no" with reinforcements: gesture, movement, sound, color (light/dark). They did not produce any results.

Current State of the Child

After six years of therapy, the child maintains visual contact with the environment. When she is in good condition she observes the world around her, especially the games and the behavior of other children. She has the ability to produce a social smile in reaction to her immediate environment. She reacts to nonverbal communication: the tone of voice, facial expressions, the position of the body. Her concentration is low. She may become interested in her environment but only for a brief period. She is fully dependent on her caretakers. She requires constant care of an adult in basic functions such as moving, eating, washing, etc.

The girl does not speak, she sits only with her back and head supported, her circadian rhythm is disordered. The prolonged state of increased muscle tension has led to muscle contracture in all limbs. Manual agility is significantly reduced. The hands are contracted, and fists are tightened. The frequency of epileptic attacks has reduced from 1–2 daily to 1–2 weekly.

Implications for Nursing Practice

1. Speech and language therapy procedures with a proper diagnosis in children with moyamoya disease show beneficial effects.
2. Neurologopedic programme should be focused on the primary functions, improvement of the quality of breathing and eye contact.
3. The main purpose of the therapy is to facilitate feeding, drinking, and chewing, and to introduce alternative communication.
4. Moyamoya remains a significant clinical challenge for multidisciplinary health care providers (neurologists, nurses, logopedists, and physiotherapists).

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