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Principles of management in the case of dysphagia in the course of amyotrophic lateral sclerosis

Zasady postępowanie w przypadku dysfagii w przebiegu stwardnienia bocznego zanikowego

Piotr Ożóg¹, Dawid Natański¹, Walery Zukow²

¹Department of Physiotherapy, Collegium Medicum, Nicolaus Copernicus University in Torun, Poland

²Department of Spatial Management and Tourism, Faculty of Earth Sciences, Nicolaus Copernicus University, Torun, Poland

Streszczenie

Stwardnienie zanikowe boczne jest postępującą chorobą neurodegeneracyjną, w której dochodzi do uszkodzenia obwodowego jak i ośrodkowego neuronu ruchowego. Schorzenie obejmuje głównie osoby w wieku średnim - w szóstym i siódmym dekadzie życia. Częściej występuje u płci męskiej i trwa średnio 2-3 lata (kończy się śmiercią). Choroba ma indywidualny postępujący przebieg i jest trudna do zdiagnozowania. Częstym powikłaniem wśród pacjentów są zaburzenia połykania (dysfagia). Problem może być związany z samym procesem przyjmowania posiłku, jak również jego przemieszczaniem do żołądka. Opieka paliatywna oraz związana z nią fizjoterapia zajmuje się łagodzeniem objawów choroby. Indywidualne postępowanie ma na celu zapobieganie niedożywieniu pacjenta oraz aspiracji pokarmu do układu oddechowego, co może doprowadzić do zapalenia płuc i innych powikłań. Proces usprawniania ma na celu dążenie do jak najdłuższego

utrzymania fizjologicznego procesu połykania, a w późniejszym etapie do połykania funkcjonalnego. Głównym celem rehabilitacji u pacjentów z problemem dysfagii jest odtworzenie skutecznego odżywiania drogą ustną z zachowaniem lub kompensacją odruchami obronnymi umożliwiającymi ochronę dróg oddechowych. Wyróżnia się trzy rodzaje metod terapii czynnościowej: odtwórczą, kompensacyjną oraz adaptacyjną. W momencie, gdy wsparcie fizjoterapeuty nie będzie w stanie zapewnić możliwości funkcji połykania, wskazane jest zastosowanie innych metod wspomagających, jak na przykład gastrostomii.

Słowa kluczowe: dysfagia, zaburzenia połykania, stwardnienie zanikowe boczne.

Abstract

Amyotrophic Lateral Sclerosis (ALS) is a progressive neurodegenerative disease which causes damage to the peripheral or central motor. The disease mainly includes middle-aged persons in the sixth and seventh decades of life. It is more frequent in males and takes an average of 2-3 years (causing death). ALS has an individual growing course and is difficult to diagnose. A frequent complication among patients is dysphagia (swallowing disorders). The issue may be related to the process of fasting as well as its move to the stomach. Palliative care and linked with her physiotherapy deals with symptom relief. Individual treatment aims to prevent patient malnutrition and aspiration of food into the respiratory system, which can lead to pneumonia and other complications. The process of improvement consists in striving for the longest possible physiological swallowing and later for functional swallowing. The main goal of rehabilitation for patients with dysphagia is to restore the nutrition orally effective while maintaining or compensation defensive reflexes enable respiratory protection. There are three types of therapies functional: restorative, compensatory and adaptive. When the physiotherapist's support will not be able to provide the swallowing function, it is advisable to use other supportive methods, such as gastrostomy.

Key words: dysphagia, swallowing disorders, amyotrophic lateral sclerosis.

Admission

Neurodegenerative diseases are a group of many diseases of the nervous system of the progressive course. Included are congenital and acquired. Their common feature is the loss of nerve cells in the course of the disorder. One of these is amyotrophic lateral sclerosis (Charcot's disease, motor neuron disease, lat. Amyotrophic Lateral Sclerosis - ALS called. Amyotrophic lateral sclerosis - ALS), which causes damage to both the peripheral and central motor [1]. The incidence of ALS is about 1-2.6 / 100 000 people per year, while the overall incidence is approximately 6/100 000 people. The disease mainly involves people in middle age - in the sixth and seventh decade of life (58-60 years on average), but are also known cases of younger people, as well as the elderly. It is more common in males, and the median survival time from the beginning to the death of 3 -4 years. [2].

In the period from 19 October 2010. 31 December 2011. In the United States alone reported 12187 cases of ALS. G. Logroscino in his work (2019) presents epidemiological data of the European Consortium Registry amyotrophic lateral sclerosis (EURALS). By. EURALS across Europe is people with ALS in 1028, and the estimated incidence of 2.2 per 100 000 people in the general population. The lowest incidence was observed in East Asia (0.89 / 100 000), South Asia (0.79 / 100 000). A large part of Africa, Latin America and Asia, there are no population studies [3, 4]. The initial clinical picture depends on the level of damage and the type of the damaged neuron. The etiology of ALS is not completely understood. About 5 to 10% of cases are hereditary origin. Other cases are more complex origins. The etiology is multifactorial SLA as evidenced by the differences in the course and many types of diseases. SLA pathogenic mechanisms are still unclear, despite many studies showing changes such as mitochondrial dysfunction, glutamate excitotoxicity, oxidative stress and inflammation, neurological. [5] Often it is hard primary reason for its creation, as both the mechanisms and the causes are superimposed on each other. Some of putative mechanisms underlying ALS include genetic mechanisms, senile changes, viral infections, environmental factors, as well as toxic compounds [2,5,6]. The first person who described the SLA was Jean Martin Charcot. Researchers, comparing the twenty-first century and the nineteenth, and a small quantity of the patients and a minimal number of autopsies conducted, a description of the disease was fully accurate. Number 5 described in steps (Table I) [1].

Table I. The original description of the ALS Charcot [1]

1. Initially, there is a weakening of the upper limb, without paresthesia, accompanied by rapid muscle atrophy. At this stage, spasticity may outweigh the flaccid muscle wasting.
2. In the second stage comes to the involvement of muscle of the lower limb. Progressive muscle weakness (without paresthesia) quickly leads to difficulty in standing and walking. Muscle atrophy in the lower limb appears later and rather does not reach such a high degree as in the upper limb. Sphincter disorders do not occur ever again. There is also a tendency to bedsores despite the immobilization of the patient.
3. In the third phase of the disease symptoms worsen muscle weakness in the upper limbs and lower, there are also signs of bulbar.
4. After about 6-12 months, the disease symptoms are already all the damage of the upper and lower motor neuron and bulbar.
5. Death occurs usually after 2-3 years as a consequence of bulbar disorders.

In this disease entity is not entitled. It is concluded that one particular clinical picture, however, patients in the early stages may face problems such as failure to maintain various objects, stumbling or even overturning, muscle cramps, fasciculation and aphasia (language disturbance). Patients are beginning to use crutches or walkers, but with time increasing weakness and muscle atrophy lead to having to use a wheelchair. At this stage, patients begin to have a problem with keeping the head in the axis of the body. When problems begin to include the surrounding muscular torso observed orthostatic difficulty, twisting from side to side and even with a seat. Speech is becoming more so incomprehensible. "Nasal". Increased muscle tension, which can lead to contractures. In a further process step, salivation and swallowing disorders which increases the risk of chokings. There are also problems with breathing and expectoration, and in the case of extreme hypoxia patient may have hallucinations or be confused. In such situations, patients are often not able to fully control their emotions (crying or unwanted laughs). These are symptoms of supposedly bulbul caused physical damage to the brain [6].

In published in 2018. Study by Nicholson K. et al. Describes the most commonly reported symptoms among sufferers of ALS (study group consisted of 567 persons). These include: fatigue (90%), muscle stiffness and cramps (84% and 74%), and dyspnea (66%). The respondents also marked the difficulty sleeping, pain, anxiety and depression, an increased amount of saliva, constipation, loss of appetite, and weight loss. Interestingly, fatigue was not only the most frequently reported symptom, but also the least treated.[7].

Unfortunately, for the time JM Charcot that little has changed in terms of the treatment of ALS. Scientists have not yet created a drug to guarantee recovery. Pharmacology recommended ALS is only to alleviate the symptoms and the ability to prolong life by a few months. Disease inevitably leads to death of the patient within an average of 2.5-3 years [2].

In patients with ALS leads to symptomatic treatment, including m.in following problems [1]:

- Excessive salivation (recommended pharmacological treatment, portable, domestic mammals, with no effect that pharmacological treatment is recommended that exposure of the salivary glands).
- Excess bronchial secretion (Recommended Power cough performed by a physiotherapist, and family use suction devices, humidifiers, e.g., drug therapy. Mucolytics, breathing aids espólnie bronchial infections).
- Emotional lability (resulting from an organic injury brain, in the case of disruptive syndromes recommended pharmacotherapy in the form of antidepressant).
- Muscle cramps (recommended kinesitherapy and physical procedures like. Hydrotherapy).
- Spasticity (recommended regular physiotherapy treatment).
- Anxiety, depression, insomnia (recommended pharmacotherapy).
- Changes in venous thrombosis (recommended physiotherapy, leg elevation and the use of compression stockings).

One form of the proposed care is palliative care, what to improve the quality of life of patients. In published in 2013. Report of the American Academy of Neurology on the quality of services for patients with ALS have been identified gaps in health care and identified a number of recommendations following [8]:

- Each patient with the SLA requires an interdisciplinary plan of care (doctor, nurse, physiotherapist, psychologist).
- Care should focus primarily on leveling symptoms.
- In the event of respiratory failure, non-invasive ventilation should be provided.
- Patients with ALS in the presence of indication should be given enteral nutrition.

The involvement of individual members of the team is dependent on the current needs of the patient. Physiotherapist using many techniques and methods, the patient is able to help in many aspects, including support the symptomatic treatment of dysphagia. [9].

Dysphagia (dysfalgia)

The problem may be related to the process of fasting as well as the movements of the stomach [9,10]. They arise mainly from damage to the lower neuron (corresponding with symptoms of so-called bulbar syndrome) resulting in weakness, and later muscle atrophy also necessary for the proper conduct of the process of swallowing. Simultaneous influence on the damage pseudobulbar syndrome is associated with a defect of the upper neuron. Manifested by increased muscle tone throat, palate and tongue. Scientific reports indicate that the problem can also result from damage to centers of swallowing as a result of the disease. Extremely dangerous symptom of dysphagia is a problem of aspiration (food entering the contents of the respiratory system). Silent aspiration, ie that at which there is no cough reflex, may cause aspiration pneumonia or even death of the patient. The larger the cough reflex disorder, the greater becomes the likelihood of aspiration. However, if this reflex is fully abolished any interference ends aspiration of food. [10,11]. Important for the early diagnosis of the problem of dysphagia in ALS research have laryngologiczno - phoniatic, which also allow you to exclude any other type of peripheral causes of bulbar symptoms such as throat or larynx cancer. Rapid implementation of rehabilitation proceedings may have a positive impact on improving the quality of life of the patient [12]. The larger the cough reflex disorder, the greater becomes the likelihood of aspiration. However, if this reflex is fully abolished any interference ends aspiration of food. [10,11]. Important for the early diagnosis of the problem of dysphagia in ALS research have laryngologic - phoniatic, which also allow you to exclude any other type of peripheral causes of bulbar symptoms such as throat or larynx cancer. Rapid implementation of rehabilitation proceedings may have a positive impact on improving the quality of life of the patient [12]. The larger the cough reflex disorder, the greater becomes the likelihood of aspiration. However, if this reflex is fully abolished any interference ends aspiration of food. [10,11]. Important for the early diagnosis of the problem of dysphagia in ALS research have laryngologiczno - phoniatic, which also allow you to exclude any other type of peripheral causes of bulbar symptoms such as throat or larynx cancer. Rapid implementation of rehabilitation proceedings may have a positive impact on improving the quality of life of the patient [12]. Important for the early diagnosis of the problem of dysphagia in ALS research have laryngologic - phoniatic, which also allow you to exclude any other type of peripheral causes of bulbar symptoms such as throat or larynx cancer. Rapid implementation of rehabilitation proceedings may have a

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Proceedings physiotherapy

The main purpose of rehabilitation of patients struggling with the problem of dysphagia is to restore effective nutrition orally or maintaining compensation defensive reflexes that will enable respiratory protection. There are three types of therapies functional [10]:

1. reconstructive (restitution);
2. compensation;
3. adaptive.

The techniques used in the treatment of reproductive are intended normalization tonicity restoration activities higher integration and a reduced to a minimum, and if possible the complete elimination of pathological reflexes while priming physiological reflexes. The therapy is based on the passive stimulation, movement exercises and autonomous mobilization techniques. Given the assumptions of neurophysiological should focus on stimulating links proprioreceptywno-neuromuscular. It is used to form various types of stimulation (stretching vibration, temperature afflictions stimulation). Mobilization techniques are used to stimulate muscle contraction, strengthening their forces and the restoration of normal motor coordination.

First, when the disease is not yet fully advanced, pay special attention to the flexibility and strength of the facial muscles involved in the process of biting and forming the billet. Mainly it will masseter, upper lip levator, orbicularis oris muscle, cheek muscles and reduce lower lip. Exercise is best done in a sitting position several times of repetitions, avoiding fatigue. Biofeedback element may comprise a mirror arranged in front of the patient which is able to monitor and control the performance of each of the exercises in a correct way [13]. In studies Hägglund P. et al. [14] showed a significant impact of this type of exercise on swallowing function among the elderly over 65 years of age (N = 116). We evaluated the rate of swallowing (swallowing test water at a certain time), changes in aspirations, as well as the quality of life for swallowing function. The study was conducted by the initiation of therapy, at the end of the 5-week exercise program, as well as 6 months later. At the end of the program in the study group (neuromuscular training the muscles of the face and throat) demonstrated significantly better results compared to a control group that uses classical care. The results in test group proved to be 60% better ($p = 0.007$). 6 months after the end

of exercise index swallowing intervention group remained significantly better ($p = 0.031$). Symptoms of aspiration also significantly reduced in the study group compared with the control group ($p = 0.01$). There were no significant differences between the groups in terms of quality of life related to swallowing.

Another equally important element is the hyoid bone. During the process of swallowing as a result of muscle over and sub hyoid bone is lifted while pulling behind the larynx, thus protecting the inlet into the trachea prior to aspiration of food.

Patients with ALS due to increased muscle tone in aligning the larynx and the hyoid bone in the upper position and the impossibility of their return to the intermediate position after the swallow, which interferes with the phase of swallowing reflex. It is therefore extremely important to mobilize and stabilize the hyoid bone with the ligament-muscle connections with other structures in the neck. Manual stretching of muscles and over and sub hyoid bone beneficial effect on the loss of muscle tone of these areas [13].

In the case of increased distance between the mandible and the hyoid bone, there is a higher risk of aspiration especially in the elderly. The cause may be related to cardiac over hyoid bone, and the therapy consists in performing exercises to increase the connection thereof. The effectiveness of this type of exercise is confirmed by the results of the latest research. Yano J, et al. [15] who have analyzed the effect of strengthening exercises tongue geniohyoid muscle in healthy adults. Results exercise was evaluated by measuring the maximum pressure into a device for measuring the pressure of language and observation of muscle-drone-borne rest and during contraction using ultrasound imaging. After the 8 week training program maximum pressure has increased significantly from 44.9 to 61.6 kPa. Muscle area drone-borne at rest also increased significantly from 2.3 to 2.6 cm². A limitation of the study mentioned is the fact that the test was carried out in a group of healthy individuals.

An effective solution is to pave the exercise movements of the hyoid bone in diagonal directions, for example, using the PNF method (Proprioceptive Neuromuscular Facilitation). It is a method of dealing with neuromuscular priming by stimulating proprioceptors and eksterceptorów body. For this purpose various types of patterns muscle based on the principles in the field of neurophysiology motor body [16].

Technique "fast ice" allows more muscle stimulation during therapy. Therapist before initiating the movement of the patient takes vigorous strokes of an ice cube to the skin of the patient in the course of the weakened muscle [13].

Compensation methods focused on facilitating the swallowing act while preventing aspiration of the billet to the airways using a modification of behavior garbage and changing the

position of the body, especially the head or with certain maneuvers garbage. Proper positioning of the head allows as many as 81% of patients significantly reduces the likelihood or completely prevent aspiration [10,17].

The main items during the meal is sitting or standing position, and the head should be in an intermediate position in the crook of light, which stimulates deep flexors, answers for stabilizing the neck, and these in turn affect the muscles of taking an active part process swallowed. Sometimes in the later stages of the disease of the cervical spine muscles they are weakened to the extent that it starts to touch chin to the sternum. However, this can be corrected by performing a delicate symmetrical reclining. In the case of patients who are at high risk for aspiration pneumonia is recommended to set the headrest at an angle of 30-45 degrees [13].

Commonly used are the direct rehabilitation exercises, which are specific maneuvers garbage. The most frequently used are [10,18]:

1. Maneuver Mendelssohn - aware extension of the elevation of the larynx and hyoid bone shift base of the tongue towards the throat of a deliberate extension of pressure on the palate).
2. Supraglottic swallowing - conscious breathing cessation during swallowing exerting maximum pressure in current throat in order to strengthen the closure of the glottis through better retraction back of the tongue).
3. Swallowing strength - improves the pressure bolus and pharyngeal phase of transportation, increases the movements of the back of the tongue towards the throat, clearing holes linguistically epiglottidean makes it difficult to shorting throat.
4. Shaker Maneuver - the patient is in the supine position and maneuver involves lifting the head which causes muscle strengthening suprahyoid.
5. Masaco maneuver - is to switch while maintaining the language between the incisors.

In case of swallowing abnormality they are associated with a feeling of necessity or retention perform more than one swallow should focus on strengthening the work base of the tongue and throat. For this purpose an exercise such as loud swallowing, swallowing with increased strength and swallowing linguistically epiglottidean which discontinues the air during swallowing with the additional feature of a strong support with both hands on the seat [18].

The above-mentioned techniques can be used in patients who do not have paresis. Otherwise, so-called a compensation maneuver, which consists in carrying out the bending of the head forward by an angle of 30 degrees. This enables the "closure" of the trachea and reduces the risk of aspiration of fluids. Whereas administration of meals maneuver involves bending the side of the head towards the body of weakness (in a situation where there is one-sided hemiparesis) In addition, the therapist can do cheek muscle stimulation [19].

Adaptive methods use aids, enabling adaptation to the changed conditions of structurally and functionally swallowing. These include the proper diet, using appropriate spoons and blood meal or to facilitate placement of the fluid in the mouth. Spoon used for feeding the patient should be large, flat and have a smooth rounded edges. If the patient is unable to take fluids can with a cup of raised edges, it must be remembered constantly filling the vessel to avoid tilting your head to the rear. It may be preferable to drink liquids through a straw or a cup of low-placed dziubku (so will avoid tilting your head to the rear).

The effects of comprehensive therapy in the case of neurologic dysphagia are published in 2019 results of Tarameshlu M. et al. [20] studies in patients with multiple sclerosis. Patients were randomly divided into the study group (using a comprehensive therapy including synergistic interaction of all the methods of interaction mentioned in this article) and a control group in which diet and posture correction were used. Both groups continued therapy for 6 weeks (3 visits per week). The results were evaluated using the following scales: MASA (Penetration-Aspiration Scale) and PRRS (Pharyngeal Residue Rating Scale). There was a significant improvement in both scales in both groups ($p < 0.001$), but a significantly greater difference was found in the study group.

When the above-mentioned methods become insufficient, the use of a gastric probe should be considered, which should also not be a solution longer than 30 days, after this time gastrostomy is recommended [10,18]. Currently, the most common non-surgical method is PEG (endoscopic percutaneous gastrostomy), which is an effective solution for patients who have a high risk of complications with laparotomy and general anesthesia. PEG is a relatively safe procedure for enteral feeding of SLA patients, but in some patients it may be associated with complications. Carbó Perseguer J. et al. [21] analyzed the mortality and complications in patients with ALS ($n = 49$) who received PEG. Mortality during surgery and after 30 days was 2% ($n = 1$). Six patients (12.2%) experienced serious complications, while 17 (34.7%) had less serious complications that were easily solved in a conservative way.

The question arises as to the patients who are unable to perform gastrostomy for reasons such as removal of the stomach, the occurrence of gastroesophageal reflux, which can lead to aspiration pneumonia or the gastrointestinal defects themselves. In these people, jejunostomy is performed, i.e. stoma on the intestine partitions [11,22].

Summary

The problem of ALS, despite the growing knowledge about the disease itself, still requires constant attention. As the results of the latest research show, physiotherapeutic methods definitely improve the function of structures responsible for the swallowing process, also among people whose disorders result from neurological disorders such as multiple sclerosis. Therefore, it seems appropriate to use this type of procedure also for people suffering from ALS, whose dysphagia limits daily functioning, thus affecting the quality of life.

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Adres do korespondencji:

Piotr Ożóg,

Katedra Fizjoterapii,

ul. Techników 3, 85-801 Bydgoszcz

tel. 52 585-34-64

katfizjoter@cm.umk.pl