

The journal has had 7 points in Ministry of Science and Higher Education parametric evaluation, Part B item 1223 (26.01.2017).
1223 Journal of Education, Health and Sport eISSN 2391-8306 7

© The Authors 2017;

This article is published with open access at Licensee Open Journal Systems of Kazimierz Wielki University in Bydgoszcz, Poland

Open Access. This article is distributed under the terms of the Creative Commons Attribution Noncommercial License which permits any noncommercial use, distribution, and reproduction in any medium, provided the original author(s) and source are credited. This is an open access article licensed under the terms of the Creative Commons Attribution Non Commercial License (<http://creativecommons.org/licenses/by-nc/4.0/>) which permits unrestricted, non commercial use, distribution and reproduction in any medium, provided the work is properly cited. This is an open access article licensed under the terms of the Creative Commons Attribution Non Commercial License (<http://creativecommons.org/licenses/by-nc/4.0/>) which permits unrestricted, non commercial use, distribution and reproduction in any medium, provided the work is properly cited.

The authors declare that there is no conflict of interests regarding the publication of this paper.

Received: 10.11.2017. Revised: 15.11.2017. Accepted: 11.12.2017.

Kinesitherapy in amyotrophic lateral sclerosis

Joanna Ilżecka

Independent Neurological Rehabilitation Unit, Medical University, Lublin

Keywords: amyotrophic lateral sclerosis, nervous system, kinesitherapy, neuroprotection

SUMMARY

Introduction and purpose of work. Amyotrophic lateral sclerosis (ALS) is a progressive neurodegenerative disease of unknown etiology. As a result, damage to motor neurons in patients with clinical symptoms such as weakness and atrophy of muscles, spasticity, paresis, disorders of speech and swallowing difficulties. In view of the lack of effective treatment, it is important causal symptomatic treatment and rehabilitation, including kinesitherapy. The aim of this study was to review the literature concerning the use of kinesitherapy in the ALS.

Brief description of the state of knowledge. The evidence regarding the benefits of kinesitherapy in the ALS is limited to a few experimental studies and clinical trials. Experimental studies confirm the beneficial neuroprotective effect of kinesitherapy on motoneurons. Clinical trials have shown the beneficial effect of moderate exercise on the state of ALS patients. It is recommended to exercise: stretching, aerobic, resistance, strengthening and balance, as well as special methods such as the method Vojta and PNF. Literature data

show that exercise influence on the activity of motoneurons which are more resistant to apoptosis selectively.

Summary. Kinesitherapy is recommended for patients already in the early stages of ALS. However, despite the presented advantages of this method are the need for a more randomized trials on a larger group of patients.

INTRODUCTION AND PURPOSE OF WORK

Amyotrophic lateral sclerosis (ALS) is a progressive neurodegenerative disease of unknown etiology in which there is damage to motor neurons. The result of the disease is progressive muscle weakness and atrophy, limb paresis, spasticity, speech and swallowing disorders. There are also mood disorders and cognitive disorders. In some patients observed in frontotemporal dementia. There is no effective pharmacological treatment. Only riluzole may slightly lengthen survival. Patients usually die within 2-3 years mainly because of respiratory failure [1,2].

Despite that progressive muscle weakness in the absence of pain is a typical symptom of ALS, over time patients may occur secondary musculoskeletal pain and discomfort due to poor mobility. Pain is reported by some ALS patients even in the early stages of the disease. A typical pain is neck and shoulder area. Especially common are: loss of range of motion, subluxation and cramps within the arms. However, in the lower limbs discomfort may be associated with spasticity, loss of range of motion and muscle spasms. The weakening of the muscles of the mouth and tongue causes difficulty in chewing and swallowing (dysphagia). Dysphagia can lead to malnutrition, which is a negative prognostic factor in ALS [3]. In view of the lack of effective treatment, it is important causal symptomatic treatment and rehabilitation, including kinesitherapy. Evidence of benefit in the use of kinesitherapy in the ALS is limited to a few experimental and clinical studies [4]. The aim of the work is to review the literature concerning the use of kinesitherapy in the ALS.

DESCRIPTION OF KNOWLEDGE

Experimental studies on the neuroprotective effect of kinesitherapy in the ALS.

Experimental studies confirm the beneficial neuroprotective effect in ALS kinesitherapy. It has been shown that kinesitherapy has a protective effect on motor neurons, causing change in the structure of the dendrites, increased protein synthesis, improved axonal transport and synaptic transmission and improve the neuromuscular and change of electrophysiological properties which may affect neuronal gene expression. Increased physical activity leads to an increase in activity, synaptic neuron by normal neurotransmitter release from nerve endings and amplification of synaptic connections to skeletal muscle as evidenced by the increase in acetylcholinesterase and the number of acetylcholine receptors within the muscle fiber [5]. Because the exercises work on strengthening the neuromuscular junction of normal motor neurons, hypothesized they can prevent loss of the neuromuscular junction, causing patronage selectively vulnerable to damage the motor neurons in ALS [6]. Under the influence of exercise in rats showed an increase in protein synthesis and an increase in the activity of glucose-6-phosphate. It was also shown that long-term exercise are not important metabolic stressor of the skeletal muscle [5]. Other studies have shown increased concentrations of growth factors such as VEGF, BDNF, GDNF, CNTF and IGF-1 under the influence of exercise, which is associated with neuroprotection and neurogenesis in the brain and spinal cord, and increased transport of growth factors by the blood - brain barrier [7,8]. It has been shown that a healthy motor neurons of rats subjected to moderate training endurance capacity have better transport protein [5]. It has been observed, that only prolonged period of exercise, lasting a few weeks, had a beneficial effect on axonal transport and rats exposed to exhaustive exercise showed his release [9,10]. Mitochondria within the motor neurons in ALS exhibit irregularities involving their vacuolization, disorganization and expansion of the membrane during presymptomatic stage of the disease. According Mattiazzi et al.[11] exercises do not adversely affect the energy production by mitochondria. The pathology of ALS plays an important role glia. It has been demonstrated that in experimental animals subjected to exercise slowed activation of astroglia and microglia, indicating the protective effects relative to glial [12]. In the brains of rats subjected to exercise it has been shown to increase angiogenesis and proliferation of astrocytes [13]. Exercises in experimental animals can induce angiogenesis in the cortex and striatum involved in motor function [14]. Other studies have also demonstrated a beneficial effect of exercise to increase vascularity [15,16]. Thus, stimulated by the increase in exercise angiogenesis in combination with astrocytosis can

act against damage a blood - brain barrier and may inhibit reactive astrogliosis observed in ALS [17,18]. Deforges et al. [19] compared the efficacy of swimming training and speed training in a mouse model of ALS. The authors showed that swimming sustained motor function and prolonged the survival of mice by about 25 days. In addition, the swimming program protects the behavior of the population of astrocytes and oligodendrocytes in the spinal cord of mice. Thus, it demonstrated that the above training induces neuroprotective mechanisms. Studies in a mouse model of ALS have shown that moderate endurance exercise delayed the onset of the disease and lengthened survival [20-22]. On the other hand, very vigorous exercise were harmful [20,23].

Clinical studies on the impact of kinesitherapy on the state of ALS patients.

For many years, exercise is not recommended for patients with neuromuscular diseases thinking that exercise can accelerate the process of muscle weakness. However, there was no evidence that it is indeed the case. Clinical trials have shown the benefits of physical activity [24]. Conducted on a small group of patients with ALS emphasized the safety of exercise of moderate intensity. The study of Drory et al. [25] randomized 25 ALS patients in order to conduct an intensive daily individualized average exercise program. The aim of the study was to improve muscular endurance. The exercise program was well tolerated and reduced the scale of functional disability evaluated ALSFRS and Ashworth spasticity scale. A study conducted by Bello-Haas et al. [26] showed a beneficial effect of retaining moderate exercise on the functional status of patients measured ALSFRS scale and on the quality of life assessed after 6 months of kinesitherapy. Currently, it is believed that moderate-intensity exercise are safe for ALS patients. Avoid prolonged fatigue after physical exertion and muscle soreness indicating muscle damage caused by overloading them [27]. Contraindicated are intense strength training. Mild exercise can to improve sleep and mood of patients. Aerobic exercise in social environment (eg. the pool, golf) are preferred, and can contribute to promoting social interaction. They have a potential role in reducing symptoms and improving mood, sleep, reduced spasticity, improved quality of life. It should maintain a moderate level of submaximal exercise. If the patient can not speak freely during exercise, it proves too dense their program. Stretching exercise and physical activity should be initiated in the early stages of the disease. Clinical experience suggests that the beneficial effects of straight stretches that prevent contractures and spasticity and also the occurrence of pain and reduced function, especially in the shoulder. Strengthening exercises have a potential role in maintaining muscle strength. It is recommended a specialist training and periodic modification of the exercise

program with the progress of the disease [28]. Majmudar et al. [3] exchange the following types of exercises are recommended in patients with ALS: stretching, aerobic exercise, resistance, strengthening and balance exercises. According to these authors, stretching exercises and movement are part of the standard of care for patients with ALS, which should be encouraged to simple daily stretching exercises at the beginning of the disease. With regard to strengthening and aerobic exercise are not available specific guidelines, since there are only few reports on the benefits of these exercises in the ALS. Keep in mind that fatigue after exercise should not interfere with daily activities. If the patient reported fatigue or pain lasting longer than 30 minutes after exercise, exercise program needs to be modified. By controlling the patient's ability to exercise should take into account factors such as age, cardiovascular and pulmonary state, baseline level of physical activity and the rate of progression of the disease. The latter may be important, because due to the rapidly progressive symptoms of ALS obtained results may be different than expected. Unsuitable exercise in a patient with ALS can lead to serious complications such as falls and sprains. According Krivickas [29] for patients with neuromuscular diseases, including ALS, there are four major types of exercises: stretching exercises, strength, balance and aerobic exercise. According to the author stretching exercises prevent the development of painful contractures hinder daily activities. Contractures are usually present in the knee and shoulder joints, especially in patients sitting in a wheelchair. In ALS patients stretching exercises may be beneficial in reducing spasticity and reduce the risk of falls. Controlled strength training seems to be advantageous in increasing muscle strength and delay muscular atrophy. Increasing evidence confirms the impact of exercise on the activity of ALS patients including elements of quality of life such as mood, appetite and sleep. Studies in ALS patients indicate the average level of the benefits of moderate exercise at an early stage of the disease, without adverse effect on the natural course of the disease and survival of patients. However, no randomized clinical trials examining the effects of aerobic exercise in patients with neuromuscular diseases. Moderate exercise programs can be safely adapted to the abilities, interests, availability and support a family. Type of exercise should be tailored to the patient's clinical status and needs, taking into account the main symptoms and degree of disability [30]. The conducted study of respiratory muscle training ALS patients showed no significant improvement in the inspiratory pressure and lung volume, but suggested a slight improvement of respiratory function [31]. According Sliwinski [32] an important part of the therapy in patients with ALS is the stimulation of the muscles responsible for the control trunk postural, including stabilizing muscles blades and the lumbar spine, which leads to a reduction of increased muscle tension, and improves the

active range and the dynamic movements of the limbs. The author states that in ALS patients with gait disorders must be prevented limitations range of motion in the joints, strengthen weak muscle groups of the trunk and limbs, as well as conduct regular exercises affecting the proper feeling and loading rates in the stance phase. In the case of speech and swallowing disorders occur frequently in patients with ALS, use the stimulation of facial mimic muscles and strengthen weakened muscles and tongue muscles involved in swallowing function. According Sliwinski is essential to the proper development of postural control, thus leading physiotherapist improvement should be aware of choosing the appropriate intensity exercise, which should be no larger than that when performing activities of daily living. The author believes that the strength of muscle strengthening exercises should be contraindicated because they can accelerate the loss of motor neurons.

The use of special methods in kinesiotherapy of ALS patients.

In kinesiotherapy of ALS patients can be used special methods such as the method Vojta and PNF (Prorioceptive Neuromuscular Facilitation - priming neuromuscular).

The basis of the Vojta method is priming (coordination stimulation field in the central nervous system) by the sum of incentives - time (prolonged stimulation) and spatial (simultaneous stimulation of several zones trip). Requires improvement in patient positioning so. activating position, making it easier to achieve a certain reaction during the reflex stimulation of the zones. The most commonly used release specific stimulus causing the reaction pressure. Stimulation causes the central nervous system reaches about receptors muscles and joints. This allows to build a proper body schema [33].

Stimulation by Vojta causes global motion complexes which affect the operation of the motor, autonomic, sensory as well. In the case of a patient with ALS therapy method determines the form of the disease. In the bulbar form of ALS should focus on how to maintain the longest swallowing function by activating the muscle tension in the neck and head, using known and available zone stimulation. Position of the body during treatment is similar to the ingested by the patient with the classic form of ALS, where efforts should be made to maintain the extension and rotation of the cervical spine and upper limb activation using a slightly different firing zone. Diaphragm muscle paralysis and respiratory muscles of patients with ALS leads to respiratory failure. Activating complex abdominal muscle

(abdominal effort concentric) causes resistance to the membrane, stimulating it to work. The effectiveness of Vojta method reduces permanent and irreversible damage to the nervous structures in the ALS. In the initial stage of the disease, there may be the possibility of new synaptic ending with other cells anterior horn of the spinal cord, therefore it is important to start the treatment as early as possible. Vojta therapy method conducted regularly can significantly improve the quality of life of ALS patients [34].

PNF technique is a structured way of priming, the aim of which is to improve the function or activity of the body. The task of the techniques used in PNF is to facilitate the functional movement by means of priming, inhibiting, strengthening and loosening of individual muscle groups. Technology use concentric, eccentric and static muscle work [35].

In the ALS to a reduction in muscle spasticity can be used various kinds of PNF. The first of them is the stabilization of feedback, allowing for stabilization and improvement of balance and muscle strength. Also you can use a rhythmic movement of the stimulation that is used in the absence of ability to activate the specific muscle group is responsible for a specific movement. Another technique is a dynamic continuous maneuverability, wherein due to the contraction of the coaxial exerts influence on the muscles performing pattern; this technique allows you to minimize fatigue. Therapy can also enable vehicles „hold-relax and tense-relax " to permit elimination of structural spasticity. It shall also apply to combinations of isotonic contractions, enhancing the strength of the muscles and affecting the reduction of pathological voltage, as well as relaxation technique breathing exercises which causes muscle relaxation [36,37].

The effects of kinesitherapy in the ALS.

In response to exercise is an adaptation of both the nervous and muscular systems. Among other things, increased activation of motor units and synchronization. Muscular hypertrophy-responsive fibers, increased protein synthesis, and increased capillary density [30]. According Deforges et al. [19] the effect of physical exercise on the nervous system is related to the plasticity and survival of nerve cells, but the mechanisms underlying the neuroprotective role of kinesitherapy are still not clear. Current evidence suggests that exercise influence on the activity of motoneurons, which can selectively become more resistant to apoptosis.

Nevertheless, the effects of exercise in ALS by some authors were considered controversial [38,39]. Moreover, according to Chen et al. [40] exercise contribute to the activation of the motor neuron, which can cause oxidative stress, production of free radicals and glutamate stimulation. The author emphasizes that these results apply to ALS patients who have an increased risk of neurotoxicity caused by the disease.

It is necessary to conduct more randomized clinical trials on a larger group of patients, in order to provide stronger evidence for the beneficial effects of exercise in ALS. There are known benefits of exercise in healthy people, on their impact on the cardiovascular system, musculoskeletal and immune. Therefore, it is reasonable to their use in patients with ALS least because there are no adverse effects of moderate kinesitherapy in the open literature [41].

SUMMARY

ALS is an incurable neurodegenerative disease. Studies in experimental models of ALS indicate a protective effect of exercise not only in motor neurons but also glia and blood vessels in the central nervous system. In the rehabilitation of patients with ALS is indicated among others kinesitherapy. It is recommended to all kinds of moderate exercise to reduce symptoms, improve the functional status of patients, thereby improving their quality of life. You can also use special methods such as the method Vojta and PNF. Exercise must be tailored to the clinical status of patients and modified as the disease progresses. Kinesitherapy should begin early in the ALS. They needed further larger clinical trials to confirm the benefits for the patient resulting from the application of this therapy.

REFERENCES

1. Pasinelli P, Brown RH. Molecular biology of amyotrophic lateral sclerosis: insights from genetics. *Nat Rev Neurosci.* 2006; 7: 710-723.
2. Hogden A, Foley G, Henderson RD, James N, Aoun SM. Amyotrophic lateral sclerosis: improving care with a multidisciplinary approach. *J Multidiscip Health.* 2017; 10: 205-215.
3. Majmudar S, Wu J, Paganoni S. Rehabilitation in amyotrophic lateral sclerosis: why it matters. *Muscle Nerve.* 2014; 50(1): 4–13.

4. Dal Bello-Haas V, Florence JM. Therapeutic exercise for people with amyotrophic lateral sclerosis or motor neuron disease. *Cochrane Database Syst Rev.* 2013; 5: CD005229.
5. Gardiner P, Dai Y, Heckman CJ. Effects of exercise training on alpha-motoneurons. *J Appl Physiol.* 2006; 101: 1228–1236.
6. Wood JD, Beaujeux TP, Shaw PJ. Protein aggregation in motor neurone disorders. *Neuropathol Appl Neurobiol.* 2003; 29: 529–545.
7. Cotman CW, Berchtold NC. Exercise: a behavioral intervention to enhance brain health and plasticity. *Trends Neurosci.* 2002; 25: 295–301.
8. Kleim JA, Jones TA, Schallert T. Motor enrichment and the induction of plasticity before or after brain injury. *Neurochem Res.* 2003; 28: 1757–1769.
9. Jasmin BJ, Lavoie PA, Gardiner PF. Fast axonal transport of acetylcholinesterase in rat sciatic motoneurons is enhanced following prolonged daily running, but not following swimming. *Neurosci Lett.* 1987; 78: 156–160.
10. Jasmin BJ, Lavoie PA, Gardiner PF. Fast axonal transport of labeled proteins in motoneurons of exercise-trained rats. *Am J Physiol.* 1988; 255: 731–736.
11. Mattiazzi M, D'Aurelio M, Gajewski CD, Martushova K, Kiaei M, Beal MF et al. Mutated human SOD1 causes dysfunction of oxidative phosphorylation in mitochondria of transgenic mice. *J Biol Chem.* 2002; 277: 29626–29633.
12. Kaspar BK, Frost LM, Christian L, Umapathi P, Gage FH. Synergy of insulin-like growth factor-1 and exercise in amyotrophic lateral sclerosis. *Ann Neurol.* 2005; 57: 649–655.
13. Li J, Ding YH, Rafols JA, Lai Q, McAllister JP, Ding Y. Increased astrocyte proliferation in rats after running exercise. *Neurosci Lett.* 2005; 386: 160–164.
14. Ding YH, Luan XD, Li J, Rafols JA, Guthinkonda M, Diaz FG et al. Exercise-induced overexpression of angiogenic factors and reduction of ischemia/reperfusion injury in stroke. *Curr Neurovasc Res.* 2004; 1: 411–420.
15. Kleim JA, Cooper NR, VandenBerg PM. Exercise induces angiogenesis but does not alter movement representations within rat motor cortex. *Brain Res.* 2002; 934: 1–6.
16. Swain RA, Harris AB, Wiener EC, Dutka MV, Morris HD, Theien BE et al. Prolonged exercise induces angiogenesis and increases cerebral blood volume in primary motor cortex of the rat. *Neuroscience.* 2003; 117: 1037–1046.
17. Julien JP. ALS: astrocytes move in as deadly neighbors. *Nat Neurosci.* 2007; 10: 535–537.

18. McCrate ME, Kaspar BK. Physical activity and neuroprotection in amyotrophic lateral sclerosis. *Neuromol Med.* 2008; 10: 108–117.
19. Deforges S, Branchu J, Biondi O, Grondard C, Claude C, L'ecolle S et al. Motoneuron survival is promoted by specific exercise in a mouse model of amyotrophic lateral sclerosis. *J Physiol.* 2009; 587: 3561–3572.
20. Carreras I, Yuruker S, Aytan N, Hossain L, Choi JK, Jenkins BG et al. Moderate exercise delays the motor performance decline in a transgenic model of ALS. *Brain Res.* 2010; 1313: 192-201.
21. Kirkinezos IG, Hernandez D, Bradley WG, Moraes CT. Regular exercise is beneficial to a mouse model of amyotrophic lateral sclerosis. *Ann Neurol.* 2003; 53(6): 804-807.
22. Veldink JH, Bar PR, Joosten EA, Otten M, Wokke JH, van den Berg LH. Sexual differences in onset of disease and response to exercise in a transgenic model of ALS. *Neuromuscul Disord.* 2003; 13(9): 737-743.
23. Mahoney DJ, Rodriguez C, Devries M, Yasuda N, Tarnopolsky MA. Effects of high-intensity endurance exercise training in the G93A mouse model of amyotrophic lateral sclerosis. *Muscle Nerve.* 2004; 29(5): 656-662.
24. Abresch RT, Han JJ, Carter GT. Rehabilitation management of neuromuscular disease: the role of exercise training. *J Clin Neuromuscul Dis.* 2009; 11: 7–21.
25. Drory VE, Goltsman E, Reznik JG, Mosek A, Korczyn AD. (2001). The value of muscle exercise in patients with amyotrophic lateral sclerosis. *J Neurol Sci.* 2001; 191(1-2): 133-137.
26. Bello-Haas VD, Florence JM, Kloos AD, Scheirbecker J, Lopate G, Hayes SM, et al. A randomized controlled trial of resistance exercise in individuals with ALS. *Neurology.* 2007; 68(23): 2003-2007.
27. Petrof BJ. The molecular basis of activity-induced muscle injury in Duchenne muscular dystrophy. *Mol Cell Biochem.* 1998; 179(1-2): 111-123.
28. Paganoni S, Karam C, Joyce N, Bedlack R, Carter GT. Comprehensive rehabilitative care across the spectrum of amyotrophic lateral sclerosis. *NeuroRehabilitation.* 2015; 37: 53–68.
29. Krivickas LS. Exercise in neuromuscular disease. *J Clin Neuromuscul Dis.* 2003; 5: 29–39.
30. Lopes de Almeida JP, Silvestre R, Pinto AC, de Carvalho M. Exercise and amyotrophic lateral sclerosis. *Neurol Sci.* 2012; 33: 9–15.

31. Cheah BC, Boland RA, Brodaty NE, Zoing MC, Jeffery SE, McKenzie DK et al. INSPIRATIONAL--INSPIRATORY muscle training in amyotrophic lateral sclerosis. *Amyotroph Lateral Scler.* 2009; 10(5–6): 384–392.
32. Śliwiński Z. Postępowanie fizjoterapeutyczne w szyjnopochodnym bólu głowy, stwardnieniu zanikowym bocznym, hemiplegii i zaburzeniach chodu. *Rehabilitacja w praktyce.* 2014; 6, 32-33.
33. Banaszek G. Metoda Vojty jako wczesna diagnostyka neurorozwojowa i koncepcja terapeutyczna. *Przegl Lek.* 2010; 67(1): 67-76.
34. Baranowska E. Metoda Vojty – wykorzystanie w rehabilitacji chorych na SLA. *Kwartalnik dla chorych na SLA/MND.* 2011; 4: 15-16.
35. Adler SS, Beckers D, Buck M. PNF w praktyce. DB Publishing, Warszawa; 2014. s.34.
36. Chambliss T, Jeremiason C. Proprioceptywne nerwowo-mięśniowe torowanie. W: S. Martin, M. Kessler. *Techniki terapeutyczne w fizjoterapii neurologicznej.* Elsevier Urban & Partner, Wrocław; 2012. s. 267-271.
37. Mraz T. Praktyczne wykorzystanie metody PNF w terapii pacjenta spastycznego. *Praktyczna fizjoterapia i rehabilitacja.* 2011; 13: 8-12.
38. Sanjak M, Paulson D, Sufit R, Reddan W, Beaulieu D, Erickson L et al. Physiologic and metabolic response to progressive and prolonged exercise in amyotrophic lateral sclerosis. *Neurology.* 1987; 37: 1217–1220.
39. Longstreth WT, Nelson LM, Koepsell TD, Van Belle G. Hypotheses to explain the association between vigorous physical activity and amyotrophic lateral sclerosis. *Med Hypotheses.* 1991; 34: 144–148.
40. Chen A, Montes J, Mitsumoto H. The role of exercise in amyotrophic lateral sclerosis. *Phys Med Rehabil Clin N Am.* 2008; 19: 545–557.
41. Lisle S, Tennison M. Amyotrophic lateral sclerosis: the role of exercise. *Curr Sports Med Rep.* 2015; 14: 45-46.