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The role of therapeutic exercise as a means of intervention for the treatment of amyotrophic lateral sclerosis

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Abstract

Background: Amyotrophic lateral sclerosis (ALS) leads to paralysis and premature death within an average of 3-5 years from the onset of symptoms. Therapeutic exercise helps to slow down the onset of muscle weakness. The aim of this review is to describe recent research data on the applications of therapeutic exercise in patients with ALS. The Google Scholar and PubMed databases were searched with the following keywords: motor neurone disease, amyotrophic lateral sclerosis, exercises, rehabilitation. Both clinical trials and reviews were included in the review. In conclusion, the application of therapeutic exercise helps to better compensate the deficits and to minimize the muscle weakness and atrophy caused by the disease. Exercises such as aerobics and flexibility and mobility exercises help maintain work ability and endurance and prevent lack of use in the upper and lower limbs.

Keywords: Amyotrophic lateral sclerosis, therapeutic exercise, rehabilitation

Introduction

Amyotrophic Lateral Sclerosis (ALS) or motor neurone disease is a rapidly evolving, late-onset neurodegenerative disorder that is currently incurable and leads to death. It occurs with fatigue, cramps and muscle weakness in the upper and/or lower limbs or manifests itself with dysphagia or difficulty in speech and leads to paralysis and premature death within an average of 3-5 years from the onset of symptoms [1]. ALS is a relatively rare disease that mainly affects adults, is more common in men (1/350) than in women (1/472) and is evenly distributed worldwide. The potential risk in the life of a person for the development of motor neurone disease is up to 1/300. Population studies in the UK reveal that 1-2 cases of ALS per 100000 people are diagnosed annually which corresponds to a diagnosis of six cases per day. The median survival is 20-48 months, which means that about 5000 people are affected by motor neurone disease at any given time. With the exception of a few geographical areas, the overall incidence of ALS is between 1-2.5 cases per 100000 individuals. The typical age of onset is after 40 years, while the incidence rate increases with age up to the seventh decade of life, followed by a rapid decline. The prevalence is greater than the incidence and ranges from 3.1 to 8.4 cases per 100000 people. Estimates are lower in the Americas and Asia, with the exception of Japan, where the highest rates are observed. In the western Pacific, the numbers are 50-100 times higher [2]. In Greece, in 2003 the impact is estimated at 1.1 per 100000 people [3]. Mortality rates range from 1.5 to 2.5 per 100000 people [2]. It is estimated that 58-82% show ALS with spinal onset, i.e., the points of weakness start from the upper or lower limbs and with the progression of the disease the stem symptoms will also appear. Second is ALS with longitudinal onset, where the stem nuclei are affected and dysfunction of the cerebral nerves occurs, thus leading to dysarthria and dysphagia and progressively affecting the limbs. This category has a bad prognosis. Finally, there is a percentage of 8-23% that belongs to other forms of onset [4]. In terms of prognosis, 50% of patients with ALS die within about three years, while 90% (of those that have surpassed the three-year mark) die within six years from the onset of the disease. A small percentage can exceed 10 years of life [5].

Physiotherapy is an established symptomatic treatment for ALS that relieves the symptoms and compensates for the deficits of the disease. The application of special therapeutic exercise is very important for these patients. Throughout the spectrum of the disease, the physiotherapist is on the side of the patient, constantly giving them new stimuli to maintain their independence [6]. Exercise helps minimize muscle weakness and atrophy caused by motor neurone disease. The aim of this review is to describe recent research data on the application of specific therapeutic exercises in patients with ALS.

Literature review

The Google Scholar and PubMed databases were searched with the following keywords: motor neurone disease, amyotrophic lateral sclerosis, exercises, rehabilitation. Below are the main conclusions of the articles included in this review.

In their review, Lopes *et al.* [7] refer to the role of therapeutic exercise in patients with motor neurone disease. More specifically, in the early stages of the disease, exercise prevents muscle atrophy and maintains good cardiorespiratory function. However, due to the small size of the scientific findings, more research needs to be done to document its benefits. The types of therapeutic exercise that are applied are balance, elasticity and mobility, resistance, aerobic and breathing exercises.

In a randomized clinical trial with 57 participants, Van Groenestijn *et al.* [8] studied the efficacy of aerobic exercise in the treatment of ambulatory patients with motor neurone disease. The participants were divided into two groups: 27 performed aerobic exercise and 30 followed the usual therapeutic approach applied to patients with ALS. The results showed that aerobic exercise can have some positive effects, if applied in combination with conventional treatment, but more studies are needed to better document these effects. Aerobic exercises that can be performed, according to Lennon *et al.* [5], include recumbent cycling and walking, designed to maintain work ability and endurance. The intensity ranges at 30-50% of the maximal work rate (W_{max}), is applied on a daily basis and should last as long as possible without causing excessive exhaustion of the patient. The start of aerobic exercise should be done in the very early stages of the disease due to the rapid decline that is then observed. According to the same author [5], Some additional goals of aerobic exercise are to prevent lack of use and improve mood, appetite, sleep quality and psychological well-being.

Dal Bello-Haas *et al.* [9] in a randomized clinical trial, studied the efficacy of functional exercises on the functioning and quality of life of patients with motor neurone disease. Out of 27 patients, in 13 resistance exercises were applied and in 14 the usual medical care provided to the specific patients for a period of six months. The results showed that individuals in the resistance exercise group had significantly greater upper and lower limb muscle strength, improved functioning and quality of life than individuals in the medical care group. Regarding resistance exercises, Dustine and Moore [10] emphasize that exercises with weights and active range of motion (AROM) can be applied to maintain muscle strength in the upper and lower limbs and core. One set of 8-12 repetitions with light weights should be performed 3-5 times a week and the resistance and repetitions should be reduced as the

weakening progresses. Lennon *et al.* [5] add some sub-objectives of resistance exercises, such as preventing lack of use and delaying the time of dysfunction. Resistance exercises should only be applied to muscles with strength >3 (at the Manual Muscle Testing scale) and patients should be monitored for overuse.

Concerning mobility exercises, Dustine and Moore [10] mention some forms, which are stretching and active and passive range of motion exercises (AROM and PROM) in order to increase or maintain range of motion. They recommend performing 1-2 sessions a day. Lennon *et al.* [5] point out that these exercises are only acceptable for muscles with strength >3 (Manual Muscle Testing scale). In terms of functioning, Dustine and Moore [10] suggest performing the activities of daily living, in order to maintain the ability to perform more and more skills, which should be applied when a conventional exercise program is no longer possible.

Another major issue with exercise therapy for patients with motor neurone disease is their ability to breathe. For this reason, in the study of Plowman *et al.* [11], 48 patients with certain or probable ALS and a predicted Forced Vital Capacity (FVC) rate of $\geq 60\%$ and a Revised ALS Functional Rating Scale (ALSFRS-R) score of > 30 were randomized. The researchers studied Expiratory Muscle Strength Training (EMST) programs at home for eight weeks to improve breathing, swallowing and exhaling function. The tools used to evaluate the above were changes in the value of Maximal Expiratory Pressure (MEP), the Global Swallowing Function scale, the respiratory cough spirometry, the predicted FVC rate and the ALSFRS-R scale scores. A respirator was used to perform the program and in one group, which was identified as placebo, there was no resistance. Both groups performed 25 targeted violent exhalations (five sets of five repetitions) in 20 minutes five times a week with moderate exercise intensity (50% of MEP). In this study, the intervention group had a significant improvement in the primary outcome measures. Specifically, MEP increased ($p = .009$) and swallowing improved by 14.4%, while in the placebo group swallowing disorders worsened by 11.8%. There were no statistically significant differences in the other outcome measures. Although the results were positive, the researchers stressed that the intervention needs more investigation to validate it.

Discussion-Conclusions

The results of this review show that the various forms of therapeutic exercise can significantly contribute to slowing down the time of onset of muscle weakness in patients with motor neurone disease. Exercises such as aerobics and flexibility and mobility exercises help maintain work ability and endurance and prevent uselessness in the upper and lower limbs. Indicatively, aerobics includes activities such as walking and cycling, while in elasticity and mobility stretching and AROM and PROM exercises can be applied. Other types of exercise include strengthening, either by resistance or by weight, functioning to maintain the ability for more and more activities, as well as breathing exercises to delay the onset of lung complications.

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