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RESEARCH ARTICLE

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PARTICULARITIES OF THERAPEUTIC EXERCISES IN AMYOTROPHIC LATERAL SCLEROSIS

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ABSTRACT

Amyotrophic lateral sclerosis (ALS) can be defined as a progressive and inexorable disease that affects the neurons of the last horn of the spinal cord and the pyramidal tract to varying degrees. The purpose of rehabilitation programs should be tailored to the individual's needs and goals and focused on addressing symptoms and maximizing function and allowing people with ALS to live their lives to the fullest and with quality. The best practices for the management of ALS include an interdisciplinary approach aimed at addressing the physical and psychological needs and desires of patients and their families and caregivers. Exercise can significantly improve the functional ability and pulmonary function of patients with ALS safely, as long as directed to the particularities of the patients.

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INTRODUCTION

Amyotrophic lateral sclerosis (ALS) can be defined as a progressive and inexorable disease that affects the neurons of the last horn of the spinal cord and the pyramidal tract to varying degrees. Approximately two-thirds of patients with typical ALS have a spinal form of the disease (limb onset) and present with symptoms related to focal paresis and wasting, where the symptoms may start either distally/proximally in the upper and lower limbs. Spasticity may develop in the weakened atrophic limbs, affecting grip strength and gait. Patients with bulbar onset usually present with dysphagia and dysarthria for solids or liquids, and limbs symptoms can develop almost simultaneously with bulbar symptoms, and in the vast majority of cases will occur within 2 or 3 years. Commonly, after the onset, about 80% of the population of motor neurons have already been involved in corresponding myotomes. The same principle is also valid for the spinal cortical tract neurons¹. In view of this, we can use, as an example, a binomial of possibility and necessity.

Is it "necessary" to promote metabolic stress – anterograde and retrograde? What would be the "alarms" for a possible signal about overtraining? Some authors cite abnormal peripheral and central fatigue, tiredness, and pain after therapeutic activities with "submaximal" limits. We know, based on the current literature and according to experiences, that there is a great question mark when the subject is frequency, intensity, and duration of exercises in ALS². This and some other questions depend on the particularities of the patients, the degree of evolution of the disease, the forms of clinical presentation, and, certainly, the genetic participation in response to exercise. We already know that repeat expansion of C9ORF72 predisposes to exercise-induced ALS³. The purpose of rehabilitation programs should be tailored to the individual's needs and goals and focused on addressing symptoms and maximizing function and allowing people with ALS to live their lives to the fullest and with quality. A major question that still needs to be well-exemplified concerns the "phases" of the disease and possible standardized protocols². We believe that patients with Amyotrophic Lateral Sclerosis fit into only three phases: disease onset, partial disability, and total dependency (Figure 1)

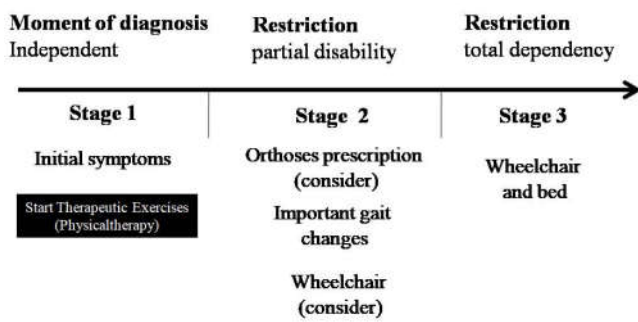


Figure 1. The Stages of ALS in the authors' view

The role of bracing, exercise, assistive devices, and adaptive equipment, as well as all technical support for patients, families, and caregivers, is essential⁴. "Sometimes not doing it can be better than doing it wrong, even if muscle strength exercises are not implemented". Doing nothing is not always a sign of a lack of understanding about the pathophysiology that involves ALS. It is, undoubtedly, a disease that needs to be dealt with in detail. A few and small studies showed that the resistance exercise in ALS patients can promote better function, as measured by total ALS Functional Rating Scale and upper and lower extremity subscale scores, and quality of life without adverse effects as compared with subjects receiving usual care⁵. We questioned how long resistance exercises are really effective in the course of the disease. Initially, they could induce positive responses, but later, rapid and irreparable damage would remedy the residual functional capacity of adaptation in this population. Mendelian randomization experiments suggest a positive causal relationship between ALS and exercise. Exercise can cause motor neuron damage only in patients with an at-risk genotype. Consistent with this we have shown that ALS risk genes are activated in response to exercise. In particular, we propose that G4C2-repeat expansion of C9ORF72 predisposes to exercise-induced ALS⁶. The understanding of genotypic variation and its relationship to physical activity is a question mark that makes us shoot in the dark.

A lifestyle assessment during the pre-symptomatic phase of ALS was performed after stratification by the C9orf72 mutation, followed by an examination of the evidence supporting the causality of lifestyle factors⁷. This is another situation that we do not pay attention to when we delegate or suggest certain functional activities and/or exercises for these patients; the "memory" to exercise. Lifestyle during the presymptomatic phase differs between patients with ALS and controls decades before onset, depends on C9-status, and is probably part of the presymptomatic causal cascade. Identification of modifiable disease-causing lifestyle factors offers opportunities to lower the risk of developing neurodegenerative disease. Interesting results were founded in a longitudinal, population-based, case-control study that used data from the Prospective ALS patient's reports. The researchers included patients with a C9orf72 mutation (C9+ group), patients without a C9orf72 mutation (C9-group), and controls. Smoking, alcohol, physical activity, body-mass index (BMI), and energy intake were relevant when compared to the three groups; one more epigenetic factor that multiplies the range of questions about physical activity in ALS⁸. Due to the collection of information from the participation of patients with ALS in registries, biomarkers and genes associated with ALS have been discovered.

The best practices for the management of ALS include an interdisciplinary approach aimed at addressing the physical and psychological needs and desires of patients and their families and caregivers. Exercise can significantly improve the functional ability and pulmonary function of patients with ALS safely, as long as directed to the particularities of the patients⁹. Despite all the satisfactory evidence, we still need to advance with regard to the prescription of frequency, intensity, duration, the interval between sets, and other variables related to exercise prescription¹⁰.

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