Rehabilitation in amyotrophic lateral sclerosis: risks and benefits

Reabilitação na esclerose lateral amiotrófica: riscos e benefícios

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Amyotrophic lateral sclerosis (ALS) is a progressive, degenerative disease that affects the neurons of the anterior horn of the spinal cord, brainstem and motor cortex.[1] The main initial complaint is muscle weakness, with amyotrophy, reduced muscle strength, and myofasciculations revealed by physical exam. It is worth mentioning that when the early signs of paresis are noticed, commonly as loss of dexterity in the hands or unexplained tripping, the patients have already lost about 80% of the motor neurons in this region. This principle is valid for the muscles of the upper and lower limbs, for breathing and swallowing. Therefore, any kind of activity can effectively overload an already ailing system.[1]

Nowadays, there is no consensus regarding the prescription of therapeutic exercises. The possibility of inducing an injury by "overtraining", that may cause intense metabolic demand on already ailing motor units is the primary concern of the professionals that deal with motor rehabilitation. The exercises (type, frequency and intensity) must respect the peculiarities of patients, by performing frequent revaluations.[1,2] Unfortunately, there is an extreme lack of randomized or quasi-randomized clinical trials examining aerobic exercise in such population.[2] Poor cooperation of patients when the disease begins to cause complete dependence, small sample size, uncontrolled and short-duration trials, remain the main restraints.

The next future for motor rehabilitation in ALS may be facing approaches that seek to stimulate the cerebral cortex or other regions of the nervous system, without generating direct neuromuscular impacts on an already debilitated system. This can happen with the use of imagery as an associated therapeutic resource.[3] Other studies also attempt to investigate, as a clinical possibility, transcranial magnetic stimulation, a proposal that still requires analysis of many forms of sedation and effective stimulation of the nervous system.[4,5] The association with the mirror neuron system seems to be another strategy in the search for more effective results for these patients.[4]

Other major challenges are the programs of pulmonary rehabilitation. Any difference in intrapulmonary pressure seems to generate metabolic consequences (favorable or harmful). In the context of respiratory training, recent studies have demonstrated its effectiveness in improving the quality of life as long as it is implemented with caution.[6,7]

Moreover, in addition to muscle weakness and amyotrophy, immobility syndrome in bed must be considered, which can result in muscle contractures, joint stiffness, pain and deformity. Besides, these

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problems also decrease tolerance to physical activity and increased fatigue. The sum of these factors characterizes a cycle that promotes prolonged stay in bed, and therefore a poorer quality of life.[7]

Educating patients and families about rehabilitation in ALS aims to:

(a) advise of energy conservation, avoiding burden when planning activities;
(b) encourage light intensity daily activities;
(c) guidance on the use of aid equipment and/or support when needed;
(d) face rehabilitation in aquatic environment as a good strategy for this clientele,[8] and
(e) provide communication options for people with ALS who cannot rely on speaking and writing.[9]

In contrast to the discouraging view that "there is nothing we can do about it", a broad approach to management, through collaboration with a multidisciplinary team, will permit the ALS physicians to make a meaningful difference in the lives of individuals surviving with ALS.[8,9]

The main suggestions for professionals who deal with ALS are:

(a) the appropriate therapeutic exercise is very important in the management of atrophy by misuse – crucial point to maintain mobility for as long as possible;
(b) the therapist should seek to work with the already ailing muscles at submaximal limits, preferably linking them to the basic and instrumental activities of daily living;
(c) never cause pain and/or unusual tiredness;
(d) body weight-supporting systems should be faced as a potential non-monotonous way for contributing for better health-related quality of life;
(e) the use of non-invasive ventilation provides more comfort, reduces dyspnea and may improve survival with quality of life, and
(f) there is no consensus on the application of electric currents for muscle stimulation, therefore they should not be used, considering that the primary problem is not in the skeletal striated muscle, but in the anterior horn neurons of the spinal cord.[7-11]

There are theories that also support the idea that stimulation of partially denervated muscle groups may, through retrograde axoplasmic flow, further impair the function of the entire motor unit.[7-11]

In ALS, although the natural history of the disease is considered ruthless, patients have particularities and various functional levels within that spectrum of presentations.[10-13] These functions should be evaluated by profiles, measurements and evaluation indicators, to subsequently be optimized by the physiotherapist.

The application of scales in ALS is used to guide professionals involved in disciplinary work, measure the clinical and functional losses that arise with disease progression and review/analyze the effectiveness of treatment goals (short, medium and long term). Some are specific as the ALSFRS, the Tufts Quantitative Neuromuscular Exam (TQNE), the ALS Severity Scale, the Appel Scale, the Norris Scale, the ALS Health State and the Severity and Functional Ability Scale. Others, although general, can also be used (Functional Independence Measure (FIM), the Barthel Index, the Manual Muscle Test (Medical Research Council – MRC), and Maximal Voluntary Isometric Contraction (MVIC).[14]

That is the key for the treatment.

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