JACOBS PUBLISHERS

OPEN ACCESS

Jacobs Journal of Physical Rehabilitation Medicine

Short Communication

Amyotrophic Lateral Sclerosis (ALS) and Physical Rehabilitation: Double Standards

Marco Orsini[1,2,3*], André Palma Matta[4], Pedro Ribeiro[1], Juliana Bitencourt[1], Mauricio de Sant Anna Jr[5], Olivia Gameiro de Souza[4], Monara Nunes[6], Victor Hugo Bastos[6], Silmar Teixeira[6], Rossano Fiorelli[2], Bruna Velasques[1], Pedro Moreira Filho[4], Osvaldo JM Nascimento[4], Roberto Dias Batista Pereira[7], Acary Bulle Oliveira[7], Juliane Rocha[8]

[1]*Rio de Janeiro Federal University – UFRJ – IBUP, Rio de Janeiro, Brazil*

[2]*Severino Sombra University – Faculty of Medicine, Vassouras, Brazil*

[3]*Augusto Motta University – Masters Program in Science Rehabilitation – Bonsucesso, Rio de Janeiro, Brazil*

[4]*Fluminense FederalUniversity – Neurology Service – Rio de Janeiro, Brazil*

[5]*IFRJ – Federal Institute of Rio de Janeiro – Physical Therapy, Brazil*

[6]*Piaui Federal University – Physical Therapy Service – Brazil*

[7]*São Paulo Federal University – UNIFESP, São Paulo, Brazil*

[8] *Federal University of Rio de Janeiro – UFRJ, Brazil*

**Corresponding author:* *Dr. Marco Orsini, Rio de Janeiro Federal University – UFRJ – IBUP, Rio de Janeiro, Brazil;*

Email: *orsinimarco@hotmail.com*

Received: *06-13-2016*

Accepted: *07-26-2016*

Published: *09-08-2016*

Amyotrophic lateral sclerosis (ALS) is usually considered a relentlessly and rapidly progressing neurological disease that destroys motor neurons in the cerebral cortex, brainstem and spinal cord [1]. Its incidence varies from 1 and 3 cases per 100,000 inhabitants per year being more prevalent between the sixth and seventh decade of life[2]. In one third of the persons with ALS, the disease starts with bulbar symptoms as the deterioration of the corticobulbar tract and affects the innervation of the muscles responsible for speech functions[3].

Health professionals often encounter with patients newly diagnosed ALS and, undoubtedly after clinical diagnosis comes the following question: How will the rehabilitation be? Initially, is necessary to understand all the natural history of the disease and outline the short, medium and long-term goals. Rehabilitation is an art, therefore requires a full mastery of the pathophysiological framework of the disease. In this sense, we conducted a review in databases of Pubmed, Lilacs, Cochrane and Scielo, to address about clinical evidences in the different aspects of rehabilitation in ALS.

The first concept to be taken into consideration by professionals dealing with rehabilitation is the "overtraining"[4]. This is defined as the installation of symptoms that reflect a non-ideal relationship between physical effort and tolerance to it, externalizing with decreased physical performance, increase in muscle injuries and even immunosuppression, increasing susceptibility to infections. Patients suffering from ALS should conduct their activities in submaximal limits, because about 80% of myotomes in the corresponding spinal segments have been denervated. Why demand them in excess if the motor units are already battered? That fact is just to increase the metabolic demand in the anterior horn neurons of the spinal cord and accelerate the disease process. Unfortunately there are few controlled randomized studies in humans with neuromuscular disorders that verify the effects of resistance therapeutic exercise, which makes them inconclusive[5].

Another change in the paradigm of ALS patients is to understand the respiratory function in disease. Any difference between intrapulmonary pressures seems to generate metabolic consequences (favorable or unfavorable). Numerous

studies have highlighted the effectiveness of treatment (when performed carefully) in the quality of life of these patients[6-7]. The exact time of use of non-invasive ventilation in patients with tracheostomy is dependent on continuous respiratory function tests and polysomnography evaluations. Calzada et al. (2016) assessed the predictive factors that influenced survival in 213 patients after NIV initiation. They observed that the two prognostic factors in ALS patients following NIV were the severity of bulbar involvement and ALSFRS-R at the time on NIV initiation. However, a better assessment of bulbar involvement, including evaluation of the upper airway, and a careful titration on NIV are necessary to optimize treatment efficacy[8].

Others adjunctive therapies in the rehabilitation of ALS is the Respiratory Muscle Training (RMT)[9], botulinum neurotoxin and radiotherapy[10]. The MRT significantly increase the Maximum Inspiratory Pressure (23.50cmH_2O; 95% CI: 7.82 to 39.19), Maximal Expiratory Pressure (12.03cmH_2O; 95% CI: 5.50 to 18.57) and Forced Expiratory Volume in one second (0.27L; 95% CI: 0.12 to 0.42). However, did not differ in Forced Vital Capacity (0.48L; 95% CI: -0.15 to 1.10) and distance in Functional Capacity (6-minute walk test - 6MWT (17.95m; 95% CI: -4.54 to 40.44)[9].

Botulinum neurotoxin (BoNT) injections in the salivary glands and radiotherapy (RT) on these glands are commonly used to alleviate severe drooling in patients with amyotrophic lateral sclerosis (ALS). Weikamp et al. (2016) compared the two therapies and there were not observed statistically significant between-treatment differences for the drooling status after treatment. However, with RT more saliva reduction was achieved and the patients described more transient negative experiences (like pain in mandible) directly after treatment[10].

The big strategy is not to use protocols in the rehabilitation treatment. We must respect the particularities of the disease and especially the patients. Some have a more prolonged evolution, whereas others an inexorable decline in motor function, breathing and swallowing. In that case, the application profiles, measures and evaluation indicators are used to guide the professionals involved in the transdisciplinary team and review of treatment goals. When we characterize the term transdisciplinary, the patient is at the center of decisions, while other professionals surround them and define the process together [11- 12] .

Speech therapy approach is also crucial because dysphagia is one of the most important problems faced in ALS. The association between dysphagia and aspiration pneumonia are usually the greatest damage to the quality of life, risk of malnutrition and dehydration[13]. Treatment in the early stages of dysphagia, this defined by physical examination and endoscopy of swallowing, may provide muscle adaptation mechanisms and minimize the risk of aspiration. The speech therapist performs compensatory procedures, based on learning and functional exercises, as well as techniques that stimulate oral proprioception, postural disorders and swallowing maneuvers [14-15].

As described by our group in the article entitled: "Rehabilitation in amyotrophic lateral sclerosis: risks and benefits"[16], the main suggestions for professionals who deal with ALS are: (a) the appropriate therapeutic exercise is very significant 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 [16].

References

1. Rianchoa J,Gonzalo I, Ruiz-Soto M, Bercianoa J. ¿Por qué degeneran las motoneuronas? Actualización en la patogenia de la esclerosis lateral amiotrófica. Neurología. 2016.

2. Makkonena T, Korpijaakko-Huuhkac AM, Ruottinena H, Puhtoa R, Holloa K et al. Oral motor functions, speech and communication before a definitive diagnosis of amyotrophic lateral sclerosis. J Commun Disord. 2016, 61: 97-105.

3. Yorkston KM, Strand E, Miller R, Hillel A, Smith K. Speech deterioration in amyotrophic lateral sclerosis: implications for the timing of intervention. Journal of Medical Speech-Language Pathology. 1993, 1(1): 35–46.

4. Orsini M, Hasue RH, Leite MAA, Menezes SLS, Silva JG et al. Doenças neuromusculares: rediscutindo o "overtraining". Fisioter Pesqui. 2014, 21 (2): 101-102.

5. Dal Bello-Haas V, Florence JM. Therapeutic exercise for people with amyotrophic lateral sclerosis or motor neuron disease. Cochrane Database Syst Rev. 2008, 16;(2): CD005229.

6. Pinto S, Carvalho M. Breathing new life into treatment advances for respiratory failure in amyotrophic lateral sclerosis patients. Neurodegen Dis Manage. 2014, 4 (1): 83-102.

7. Pinto S, de Carvalho M. Can inspiratory muscle training increase survival in early-affected amyotrophic lateral sclerosis patients?. Amyotroph Lateral Scler Frontotemporal Degener. 2013, 14 (2): 124-126.

8. Calzada G, Soro P, Gomez M, Bulta G, Izquierdo C et al. Factors predicting survival in amyotrophic lateral sclerosis patients on non-invasive ventilation. Amyotroph Lateral Scler Frontotemporal Degener. 2016, 17(5-6): 337-342.

9. Ferreira GD, Costa AC, Plentz RD, Coronel CC, Sbruzzi G. Respiratory training improved ventilatory function and respiratory muscle strength in patients with multiple sclerosis and lateral amyotrophic sclerosis: systematic review and meta-analysis. Physiotherapy. 2016.

10. Weikamp JG, Schinagl DA, Verstappen CC, Schelhaas HJ, de Swart BJ. Botulinum toxin-A injections vs radiotherapy for drooling in ALS. Acta Neurol Scand. 2016.

11. Oliveira ASB, Pereira RD. Amyotrophic lateral sclerosis (ALS): three letters that change the people's life. Forever. Arq Neuropsiquiatr. 2009, 67 (3A): 750-782.

12. Güell MR, Antón A, Rojas-García R, Puy C, Pradas J. Comprehensive care of amyotrophic lateral sclerosis patients: a care model. Arch Bronconeumol. 2013, 49 (12): 529-533.

13. Erdem NS, Karaali K, Ünal A, Kızılay F, Öğüş C. The interaction between breathing and swallowing in amyotrophic lateral sclerosis. Acta Neurol Belg. 2016.

14. Tabor L, Gaziano J, Watts S, Robison R, Plowman EK. Defining Swallowing-Related Quality of Life Profiles in Individuals with Amyotrophic Lateral Sclerosis. Dysphagia. 2016, 31(3): 376-382.

15. Plowman EK, Tabor LC, Robison R, Gaziano J, Dion C. Discriminant ability of the Eating Assessment Tool-10 to detect aspiration in individuals with amyotrophic lateral sclerosis. Neurogastroenterol Motil. 2016, 28(1): 85-90.

16. Orsini M, Oliveira AB, Leite MAA, Freitas MRG, Bastos VH. Reabilitação na esclerose lateral amiotrófica: riscos e benefícios. Rev Bras Neurol. 2014, 50 (3): 57-59.