

Guidance from the literature: Amyotrophic Lateral Sclerosis

Disclaimer: This document is not intended to provide definitive guidance on diagnosis and treatment of patients with Amyotrophic Lateral Sclerosis. It provides clinicians with general information on certain disease processes that may assist in clinical decision making. Specifically, Empi/VitalStim is not aware of any published empirical data on the use of NMES for dysphagia in this patient population and has not requested nor received specific clearance from the US FDA for such labeling. Clinicians are advised to consult the professional literature for information specific to that condition and use best practice guidelines in determining treatment intervention.

Description

Amyotrophic Lateral Sclerosis (ALS), otherwise known as “Lou Gehrig’s disease” is a progressive motor neuron disease which affects the nerves and muscles resulting in muscle weakness and atrophy of muscles.¹

Pathophysiology and Presentation

The cause of ALS remains unknown. The disorder causes the upper motor (brain) and lower motor (spinal cord) neurons to degenerate and cease to send messages to muscles. Unable to function, the muscles gradually weaken, develop fasciculations (twitching) because of denervation, and eventually atrophy.

ALS is a quite variable disease, and no two patients will have the same presentation of symptoms or progression of the disease.² As both lower and upper motor neurons (LMN and UMN) can be affected at different rates, patients will have mixed presentations. Signs of LMN degeneration include muscle weakness and atrophy, involuntary contraction of muscle fibers, muscle cramps, weakened reflexes, flaccidity (decreased tone). Signs of UMN degeneration include spasticity (muscle stiffness) and emotional lability.⁴ For some people, the muscles for speaking, swallowing or breathing are the first to be affected. This is known as Bulbar ALS. The term “bulbar” refers to the motor neurons located in the bulb region of the brain stem that control the muscles used for chewing, swallowing, and speaking.⁴ In addition to muscle weakness, patients may have central fatigue (UMN; initiation of movement) and peripheral fatigue (LMN).³

Typical dysphagia dysfunction

Oral: weak tongue or slow movements causing poor bolus formation and oral control, slow and impaired oral transit, decreased tongue base retraction

Pharyngeal: reduced hyo-laryngeal excursion, progressively weaker pharyngeal constriction, aspiration risk before and after the swallow due to residuals

Management

Therapists have as their primary goal and challenge to maintain muscle strength as much as possible in order to maintain function. A big question with progressive neuromuscular diseases is whether exercise is indicated or contraindicated, and the medical community does not have unanimous agreement on this subject. Because of the variability in this disease, efficacy of exercise can be difficult to research.

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Findings in the literature about exercise and ALS include the following concepts:

- Although clinical opinions vary, there is general agreement that exercise to the point of fatigue is not beneficial.
- Patients in the mild to moderate stages of this disease tend to respond better to therapy and are more appropriate for moderate exercise programs (not intensive exercise) than those who are in the severe stages. Increased strength of the muscles that are still intact may improve functioning with patients at the mild to moderate stages of the disease (See *Literature* below).
- Patients with neurodegenerative disease have shown benefits with moderate exercise with no adverse effects. Some research is showing that low intensity exercise (60% of max or less) may be beneficial to maintain and even regain some muscle strength and function (See *Literature* below). Sub-maximal contractions evoked with VitalStim during dysphagia therapy are considered “moderate exercise”.
- Patients in the severe stages may reach a point when therapy can no longer overcome the cumulative effects of the disease. Patients with severe progressive neurodegenerative diseases may not be good candidates for swallowing therapy or use of NMES as they may have lost too much muscle functioning to make appreciable gains.

Dysphagia management:

1. Exercise for strengthening of weak muscles
 - a. Patients should have shorter treatment sessions with frequent rest breaks.
 - b. Exercise at low intensities and closely monitor for immediate post-exercise fatigue. SLPs should monitor for signs of fatigue as evidenced by decreased function. If this fatigue is such that function is impaired, reduce the exercise intensity.
2. Modification of food/fluid consistency and quantity
3. Recommend small, frequent, highly nutritious meals

Role of NMES: Because ALS is a chronic disorder in which there may be periods of spasticity and muscle weakness and then a long term period of lower motor neuron degeneration and "flaccid paralysis," the indications for and the application of electrical stimulation will vary with the symptoms and functional limitations.

Exercise and NMES may improve the timing or recruitment of muscles so that muscles exert force in a more useful and coordinated manner. NMES is not indicated for extremely weak or insensate fibers due to the risk of damage to delicate atrophied muscle fibers- as a rule of thumb, if a patient is no longer able to volitionally move the muscles, which suggests extreme weakness, then NMES should not be used in that region. Therapists should avoid over-fatiguing a patient with a progressive neurodegenerative disease during therapy (with or without the use of NMES).

The following would be a prudent guideline in using NMES during therapy with this population:

- 1) Initiate a conservative exercise program at moderate exercise intensities while closely monitoring for fatigue and functional decline

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2) Add electrotherapy to facilitate the process if no progress is made but only if no functional declines were observed

3) If still no progress is made and/or functional declines are observed, stop electrotherapy

The role of electrotherapy would be to facilitate the muscle strengthening process. If the use of electrotherapy is going to cause a decline in neuromuscular transmission because of fatigue the decline will be noticeable during or immediately after the very first session. If this happens, the use of the modality is not indicated with that patient.

Literature review

Research with ALS and dysphagia is difficult to conduct given the variability of the disease among potential subjects. More studies have been conducted in the PT literature where patients can serve as their own controls by having treatment on one side of the body versus no treatment on the other. These studies can give us suggestions about the possible benefits and effects of exercise with this population.

- Dal Bello-Haas V, Florence JM, Kloos AD, Scheirbecker J, Lopate G, Hayes SM, Pioro EP, Mitsumoto H. **A randomized controlled trial of resistance exercise in individuals with ALS.** *Neurology* 68: 2003-7 (2007).
Findings: 27 subjects diagnosed with ALS were randomly assigned to a resistance exercise group that received a home exercise program consisting of resistance exercises 3x/week or to a usual care group who performed only daily stretching. At 6 months, the resistance exercise group had significantly higher ALS Functional Rating Scale scores with no adverse events related to the intervention. These subjects had less decline in leg strength.
- Aitkens SG, McCrory MA, Kilmer DD, Bernauer EM. **Moderate resistance exercise program: its effect in slowly progressive neuromuscular disease.** *Arch Phys Med Rehabil* 74(7): 711-5 (1993).
Findings: 27 subjects with slowly progressive neuromuscular diseases and 14 controls underwent a trial of 3x/week submaximal regimen of home exercises using ankle and wrist weights. Both the NMD and CRL groups demonstrated significantly increased strength. The authors concluded that this study provides evidence that a 12-week submaximal strength training program is practical and safe in slowly progressive NMD and produces moderate improvements in measured strength
- Kilmer DD, McCrory MA, Wright NC, Aitkens SG, Bernauer EM. **The effect of a high resistance exercise program in slowly progressive neuromuscular disease.** *Arch Phys Med Rehabil* 75(5): 560-3 (1994).
Findings: A high resistance training program although well tolerated in NMD subjects with mild to moderate weakness may cause some deleterious effects to diseased skeletal muscle. A high resistance training program appears to offer no advantage over a moderate resistance training program in this population.

References

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2. What is ALS? ALS Association. 2004. www.alsa.org
3. Kent-Braun JA, Miller RG. Central fatigue during isometric exercise in amyotrophic lateral sclerosis. *Muscle & Nerve* 23-6, 2000: 909-914.
4. Living with ALS. ALS Society of Canada, 2003. <http://www.als.ca/manual-whativals.shtml>