## Guidance from the literature: Myasthenia Gravis

<u>Disclaimer</u>: This document is not intended to provide definitive guidance on diagnosis and treatment of patients with Myasthenia Gravis. 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.

#### **Background**

An autoimmune disease is characterized by an abnormality of the immune system that causes the production of antibodies against one's own tissues and other body materials. One such autoimmune disease in which dysphagia is common is Myasthenia Gravis (MG).

MG is a chronic autoimmune neuromuscular disease characterized by varying degrees of weakness of the skeletal (voluntary) muscles of the body. The key sign is muscle weakness that improves with rest.<sup>1</sup>

### **Pathophysiology and Presentation**

Patients with MG have antibodies to the acetylcholine (ACh) receptors. ACh is a neurotransmitter that is located in areas of muscle tissue that receive nerve impulses (called neuromuscular junctions) where it acts to control muscular contraction. The ACh receptor antibodies in patients with MG block acetylcholine from binding to muscle cell receptors. The impulse transmission is therefore impaired and muscle fibers are incapable of contracting.

Patients with MG have weakness in facial, oropharyngeal, limb, and trunk muscles without any other signs of neurological deficit, such as sensory loss, change in deep tendon reflexes, or muscle atrophy. Muscle weakness increases during periods of activity and improves after periods of rest. In MG the defective neuromuscular transmission causes most of the symptoms of fatigue.<sup>3</sup>

# **Typical Dysphagia Dysfunction**

Dysphagia with MG is primarily characterized by general weakness of the swallowing system which includes decreased hyolaryngeal excursion and weakness in constriction. The muscles which provide traction force to open the UES may not produce sufficient force to pull the UES open.

# Management

Medications used to treat the disorder include anticholinesterase agents and immunosuppressive drugs. These allow for reception of the signal to the muscle by preventing enzymatic removal of ACh after it has been released in the post-synaptic cleft. This increases the availability of free ACh and maximizes the potential for binding to a free receptor.<sup>1</sup>

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Therapists have as their primary goal and challenge to maintain and possibly increase muscle strength in order to maintain function. A large question with this disease is whether exercise is indicated or contraindicated. 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 support* section below).

#### Management of dysphagia:

- 1. Modification of food and fluid consistency as needed
- 2. Exercise aimed at strengthening the weak muscle groups
  - a. Patients may benefit from 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.
- 3. Manage food intake with regards to endurance status
- 4. Smaller meals with high nutrition and calories

**Role of NMES**: The use of electrical stimulation should be considered in the context of not wanting to overly fatigue the muscles. Since not all motor end plates are affected to the same degree in MG, some motor units may have greater capacity than others. The motor units in whom the changes are most severe will not respond whereas the less affected motor units may respond favorably. There is however no known research to suggest to what extent this modality may be helpful or harmful. The following would be prudent guidelines:

- 1) Initiate a conservative exercise program at moderate exercise intensities while closely monitoring for fatigue and functional decline
- 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 is to further facilitate the muscle strengthening process. If the use of electrotherapy is going to cause a decline in neuromuscular transmission because of an 'exhausted' motor end plate, the decline will be noticeable during or immediately after the very first session. In this case the use of the modality is not indicated.

#### Literature review

There is limited published research specific to Myasthenia Gravis and swallowing therapy. Information from the PT literature provides information about the possible benefits and effects of exercise with MG in general.

• Lohi EL, Lindberg C, Anderson O. **Physical training effects of myasthenia gravis**. *Arch Phys Med Rehabil*. 74 (11): 1178-80 (1993).

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<u>Findings</u>: 11 patients with mild to moderate MG received strength training program for 10 weeks. The patients demonstrated a 23% increase in strength on the trained side vs. only a 4% increase in the untrained side. The subjects did not experience any adverse affects due to training. The authors concluded that physical training can be carried out safely in mild MG and provides some improvement in muscle force.

#### References

- 1. Myasthenia Gravis. Mayo Clinic, Mayo Foundation for Medical Education and research, 2008. <a href="https://www.mayoclinic.org/myasthenia-gravis">www.mayoclinic.org/myasthenia-gravis</a>
- 2. Myasthenia Gravis Fact Sheet. National Institute of Neurological Disorders and Stroke, July 25, 2008. www.ninds.nih.gov/disorders/myasthenia\_gravis
- 3. Awwad, Shady, MD, Myasthenia Gravis, July 13, 2007. http://www.emedicine.com/oph/topic263.htm