Guidance from the literature: Myopathies

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

Description

The idiopathic inflammatory myopathies (IIM) are systemic connective tissue diseases which are characterized by symmetrical, proximal muscle weakness, decreased muscle endurance, and chronic inflammation in muscle tissue. They can be subclassified into dermatomyositis (DM), polymyositis (PM), and inclusion body myositis (IBM) according to differences in clinical as well as histopathological features. As IBM is less similar to DM and PM, this paper will summarize polymyositis and dermatomyositis with IBM being discussed in a separate paper.

Polymyositis is a condition that results in chronic inflammation of the muscle fibers with the most noticeable characteristic being muscle weakness. Dermatomyositis leads to many of the same symptoms as polymyositis and is treated in a similar fashion, but dermatomyositis causes skin inflammation or a rash in addition to muscle inflammation.

Pathophysiology and Presentation

With polymyositis, the disease progresses as the body's immune system spontaneously invades the muscle fibers. In dermatomyositis, the immune system invades the blood vessels that supply the muscles and skin. The cause of both of these conditions is unknown.[1]

The muscles can become weak with both PM and DM and can lose varying degrees of power, as well as atrophy. Fatigue, general discomfort, and weight loss are often present, and some reports suggest the presence of muscle aches and tenderness. In DM, the patient will also present with a reddish, purple rash over the face, neck, and chest.[2]

Typical dysphagia dysfunction

As weakness and atrophy are hallmarks of these diseases, one can assume that the muscles for swallowing can also be affected. Dysphagia in polymyositis/ dermatomyositis is primarily due to weakness of the striated musculature of the posterior pharynx. Dysphagia may also result from cricopharyngeal obstruction secondary to inflammation or fibrosis of the cricopharyngeal muscles. Dysfunction of the esophagus and stomach occurs but the symptoms are usually overshadowed by pharyngeal dysfunction.

Management

Both PM and DM are often treated with medications, such as corticosteroids, that suppress the immune system and limit the production of antibodies which reduces muscle inflammation.[1] Bed rest is necessary during the acute stages of these diseases. Active therapy should be reserved until the inflammation has subsided. When the disease is under

control, exercise can help keep the muscles from becoming too weak. The key is to strike a balance between too much activity (which can strain and tire muscles), and too little activity (which can increase pain and stiffness and lead to further weakness).
Role of NMES: Guidelines regarding when the patient may be appropriate for exercise (see "Management" above) should be followed in determining when to begin dysphagia treatment using NMES.

Literature review

The literature contains one case study of a patient with polymyositis who responded well to NMES treatment as well as research in the PT field demonstrating that exercise was not detrimental to patients with stable polymyositis/ dermatomyositis.

D'Souza K, Krieger R, Kobe C. **Effect of Electric Stimulation on Swallow Function in Patient With Polymyositis: A Case Report.** *Arch Phys Med Rehab*. 2006;87(11):e14

> **Case Description:** A 62-year-old man with polymyositis was admitted to an acute care hospital with progressive weakness and dysphagia. He was started on steroids and put on a liquid diet.
> **Assessment/Results:** At the acute rehabilitation hospital, a PEG tube was inserted and the patient underwent swallowing therapy. Repeat VFSS at discharge showed no improvement in dysphagia. He then underwent ten 45-minute sessions of electric stimulation spread over 3 weeks. Repeat VFSS at the end of this treatment protocol showed improved pharyngeal transit phase. Electric stimulation accounted for earlier and quicker recovery from dysphagia in this subject.

Alexanderson H, Dastmalchi M, Esbjornsson-Liljedahl M, Opava CH, Lundberg IE. **Benefits of intensive resistance training in patients with chronic polymyositis or dermatomyositis**. *Arthritis Rheum* June 15; 57 (5): 768-77 (2007).

> **Objective**: To investigate the benefits and safety of an intensive muscular training program in patients with chronic polymyositis (PM) and dermatomyositis (DM).
> **Methods**: Nine patients with chronic PM or DM (median age 53 years, range 44-61) were included. Assessments of impairment were performed 4 weeks prior to baseline, at baseline, and after 7 weeks of exercise. The patients exercised 3 days per week for 7 weeks on loads allowing 10 VRM. **Results**: The group improved significantly regarding at 7 weeks compared with baseline ($P < 0.05$). Two patients were responders with reduced disease activity and no patient had signs of increased muscle inflammation in the muscle biopsy sample after 7 weeks of exercise.
> **Conclusion**: Patients with chronic, stable PM and DM can perform this intensive resistive exercise program with beneficial effects on impairment and activity limitation without increased muscle inflammation.

Varju C, Petho E, Kutas R, Cairjak L. **The effect of physical exercise following acute disease exacerbation in patients with dermato/polymyositis.** *Clin Rehabil* Feb; 17 (1): 83-7 (2003).

> **Subjects**: Ten patients 2-3 weeks after an acute phase of dermato/polymyositis DM/PM (early recovery group) and 11 patients in the inactive stage of DM/PM for at least three months (chronic stage group). **Interventions**: Isotonic muscle training consisted of several series of different repeated movements at 65-70% of individual maximal repetition limit. **Results**: No disease relapses or decreases in muscle function were seen. By the end of the therapy both groups showed improvements in disability tests ($p < 0.05$). **Conclusions**: Physical training started 2-3 weeks following an acute exacerbation of the disease seems to be useful and safe. Some improvement in muscle strength and respiratory function can be obtained, muscle atrophy due to inactivity may be partially prevented and the level of disability can be decreased.

GF Wiesinger, M Quittan, M Aringer, A Seeber, B Volc-Platzer, J Smolen and W Graninger. **Improvement of physical fitness and muscle strength in polymyositis/dermatomyositis patients by a training programme.** The British Journal of Rheumatology, Vol 37: 196-200 (1998).

> In this prospective, randomized, controlled study, 14 patients with polymyositis (PM) or dermatomyositis (DM) were investigated. The training, consisting of bicycle exercise and step aerobics, took place over a 6 week period. There was no significant rise in disease activity in the training group in comparison to the controls. No rise in inflammatory activity, but significant improvement in muscle strength, oxygen uptake and well-being were found in patients with inflammatory myopathy as a result of physical training. Besides medication, a physical training programme consisting mainly of concentric muscle contractions should therefore be an integral part of therapy particularly in view of the cardiopulmonary risk of these patients.

References

1. Polymyositis. Diseases and Conditions. Mayo Clinic. July 13, 2007. www.mayoclinic.com/health/polymyositis

2. Facts About Inflammatory Myopathies (Myositis). MDA Publications, Muscular Dystrophy Association. November, 2007. www.mda.org/publications/fa-myos