

## Guidance from the literature: Inclusion Body Myositis

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Disclaimer: This document is not intended to provide definitive guidance on diagnosis and treatment of patients with Inclusion Body Myositis. 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

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.

IBM is considered to be a partly inflammatory and partly degenerative muscle disease and is the focus of this review.<sup>1</sup>

### Pathophysiology and Presentation

Inclusion Body Myositis is very similar to polymyositis and is often diagnosed in cases of PM that are unresponsive to therapy.<sup>1</sup> However, IBM has its own distinctive features. The onset of muscle weakness in IBM is generally gradual (over months or years). IBM affects both the proximal (closest to the center of the body) and distal (farthest from the center of the body) muscles.

IBM and PM share some common features, especially the initial sequence of immune system activation. However, polymyositis comes on over weeks or months and does not display the subsequent muscle degeneration and protein abnormalities as seen in IBM. PM tends to respond well to treatments, while IBM does not. IBM is often misdiagnosed as polymyositis, but polymyositis that does not respond to treatment is likely IBM.<sup>2</sup>

**Onset, Prognosis:** Symptoms usually begin after the age of 50. IBM is generally resistant to all therapies, and its rate of progression also appears to be unaffected by the present treatments.<sup>1</sup>

Falling and tripping are usually the first noticeable symptoms. For some patients the disorder begins with weakness in the hands causing difficulty with gripping, pinching, and buttoning. Dysphagia occurs in approximately half of IBM cases.<sup>1</sup>

### Typical dysphagia dysfunction

Dysphagia is estimated to occur in one-third of patients who have IBM.<sup>1</sup> Dysphagia in IBM 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

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the cricopharyngeal muscles. While dysfunction of the esophagus and stomach may occur, it is usually overshadowed by the pharyngeal dysfunction.

### Management

There is no standard course of treatment for IBM, and this disease is unresponsive to corticosteroids and other immunosuppressive drugs. Therefore, therapy is symptomatic and supportive. Bed rest is necessary during the active stage of the disease, and active therapy should be reserved until the inflammation has subsided. When the disease is under control, exercise may help keep the muscles from becoming too weak (see *Literature review* below). 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 a patient may be appropriate for exercise (see above) should be followed in deciding if and when to begin dysphagia treatment using NMES. If a patient is not appropriate for exercise, then NMES should not be used. Very little information is available about the possible benefits of NMES with this IBM.

### Literature review

Little, if any, research is available about the effects of exercise on dysphagia with IBM. However, some research has been published about the impact of exercise with this diagnosis in general (see below).

Spector SA, Lemmer JT, Koffman BM, Fleisher TA, Feuerstein IM, Hurley BF, Dalakas MC. **Safety and efficacy of strength training in patients with sporadic inclusion body myositis.** *Muscle Nerve* Oct; 20 (10): 1242-8 (1997).

This study examined the effects of a 12-week progressive resistance strength training program in weakened muscles of 5 patients with sporadic inclusion body myositis (IBM). After 12 weeks, the values of repetition maximum improved in the least weakened muscles, 25-120% from baseline. Repeat muscle biopsies did not reveal changes in the number and degree of degenerating fibers or inflammation. The size of the trained muscles did not change. We conclude that a supervised progressive resistance training program in IBM patients can lead to gains in dynamic strength of the least weak muscles without causing muscle fatigue and muscle injury or serological, histological, and immunological abnormalities. Even though the functional significance of these gains is unclear, this treatment modality is a safe and perhaps overlooked means of rehabilitation of IBM patients.

Alexanderson H, Lundberg ID. **The role of exercise in the rehabilitation of idiopathic inflammatory myopathies.** *Curr Opin Rheumatol* March; 17 (2): 164-71 (2005).

**FINDINGS:** Polymyositis, dermatomyositis and inclusion body myositis are rare conditions with muscle weakness as a common prominent feature. Earlier, these patients were discouraged from active exercise due to a fear of increased muscle inflammation with recommendations to rest, perform range of motion exercises and in

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some cases, isometric exercises. Patients with active, recent onset disease seem to benefit from mild/moderate muscular exercise without signs of increased muscle inflammation. There is no evidence of increased muscle inflammation following exercise in inclusion body myositis. However the beneficial effects are unclear as one study report increased muscle strength, while the other could not achieve impairment reduction. **SUMMARY:** Studies evaluating active exercise in IIM support the notion of safety and benefits. However, large multi-center studies are needed to fully establish the safety and benefits of different types of exercise. Data indicate that active exercise, adapted to disease activity and disability should be included in the rehabilitation of patients in all stages of IIM. The newly developed and validated outcome measures for patients with polymyositis and dermatomyositis help assess the effects of interventions on disease activity and disability in clinical trials and in clinical practice. However, there are no sensitive and valid outcome measures for patients with inclusion body myositis.

### References

1. NINDS Inclusion Body Myositis Information Page. National Institute of Neurological Disorders and Stroke. April 9, 2008. [http://www.ninds.nih.gov/disorders/inclusion\\_body\\_myositis/inclusion\\_body\\_myositis.htm](http://www.ninds.nih.gov/disorders/inclusion_body_myositis/inclusion_body_myositis.htm)
2. Facts About Inflammatory Myopathies (Myositis). MDA Publication. Muscular Dystrophy Association. November, 2007. [www.mda.org/publications/fa-myos](http://www.mda.org/publications/fa-myos)