# Medical or Research Professionals/Clinicians

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## EFFICACY OF INTRAVENOUS IMMUNOGLOBULIN THERAPY IN REFRACTORY DYSPHAGIA IN PATIENTS WITH IDIOPATHIC INFLAMMATORY MYOPATHIES

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**Background:** Polymyositis (PM) and Dermatomyositis (DM) are systemic inflammatory diseases affecting skeletal muscles and other organs, particularly hypopharynx and the upper tract of the esophagus, causing motility dysfunction. The current treatment for PM and DM relies on steroids and immunosuppressants, but esophageal involvement is often refractory to standard therapies.

**Objectives:** To assess the efficacy and safety of intravenous immunoglobulins (Ig-iv) as therapy of refractory dysphagia (1) in patients affected with PM/DM who have received conventional treatments for at least three months. These are the extended data of a previous abstract entitled “Improvement of refractory dysphagia in patients with idiopathic inflammatory myopathies receiving immunoglobulin intravenous therapy” accepted to EULAR 2015 Congress.

**Methods:** We enrolled 17 patients (15 F), 8 with DM, 9 with PM according to Bohan and Peter criteria, treated with prednisone (mean dose 20 mg/day) ± Methotrexate (15 mg/wk) or Azathioprine (100 mg/day) for at least 3 months, without improvement of dysphagia. All patients received one infusion of Ig at a standard dose (2 g/kg in a day) every two months for 3 cycles. In all patients, at entry and before each infusion, creatine kinase (CK) levels and Manual Muscle strength Test on 12 muscle groups (MMT12) were evaluated. Dysphagia assessment included: Eating Assessment Tool (EAT) impaired for score ≥ 3; Three-oz Swallow Test (3oST) and fibrolaryngoscopy (FEES) for pharyngeal muscle propulsion and stasis of liquid/solid bolus detection. Student T-test and Chi-square test were used to compare baseline and follow-up data.

**Results:** At entry, all patients had an EAT score ≥ 3 with a mean score of 16.2±9, 3 patients presented a 3oST for solid (STPNS) impaired, and only 1 patient also for liquid (STPNL). CK levels were slightly increased (474±720 UI/L), and MMT12 score was 4.45±0.35. Following Ig-iv therapy we observed a significant improvement of EAT score after the first dose (9±8, p=0.006) with a further reduction after the second dose (7.4±8, p=0.007) and stability after the third one (7.6±8, p=0.008). An improvement of pharyngeal muscle propulsion, with a significant resolution of solid stasis and a partial resolution of liquid stasis were observed. In addition, MMT12 score progressively improved after each dose, increasing at 4.86±0.16 after the third dose (p= 0.001). Noteworthy, after the third Ig-iv dose, three patients stopped the steroid and the remaining 12 were treated with a significant lower dose of 5 mg/day of prednisone (p= 0.04).

**Conclusions:** According to literature, our findings provided evidence that Ig-iv induced a meaningful improvement of dysphagia, also having a steroid-sparing effect. An improvement of muscle strength was also detected. Although further studies of larger cohort of patients and a longer follow-up time are needed, our findings corroborate the utility of Ig-iv in the treatment of more resistant idiopathic inflammatory myositis related dysphagia.

**References:** 1. Marie I et al. Intravenous immunoglobulins for steroid-refractory esophageal involvement related to polymyositis and dermatomyositis: a series of 73 patients. Arthritis Care Res. 2010;62(12).

**Disclosure of Interest**: None declared