Inflammatory Myopathies: Inclusion Body Myositis (IBM)

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Condition for which IVIg has an established therapeutic role.

Specific Conditions	• Inclusion Body Myositis (IBM)
Indication for IVIg Use	• Patients with inclusion body myositis (IBM) who have dysphagia limiting dietary intake
Level of Evidence	Evidence of probable benefit – more research needed (Category 2a)
Description and Diagnostic Criteria	Inclusion body myositis (IBM) is an idiopathic inflammatory disorder of muscle. It is the most common inflammatory myopathy in individuals older than 50 yrs. Clinically IBM presents with slowly progressive weakness. It is more common in men than women (3:1). Along with proximal muscle weakness, distal muscles are commonly involved. The disease has a predilection for certain muscles, especially the quadriceps and long finger flexors, with prominent atrophy of the quadriceps muscle.
Justification for Evidence Category	The Biotext (2004) review identified three small controlled studies. Two were crossover trials comparing intravenous immunoglobulin (IVIg) to placebo in 19 patients and 22 patients. The outcome was negative even if some symptomatic positive effects were recorded. In one randomised controlled trial (RCT) IVIg plus prednisolone was compared with placebo plus prednisolone in 35 patients – the outcome was negative. Overall a small number of patients reported benefits regarding swallowing difficulties. IVIg in inclusion body myositis (IBM) continues to be controversial. Since there is a question about regional differences in response to IVIg, and persistent case reports about the efficacy of IVIg in IBM, further research is required to determine if a small subset of patients respond. For this reason, the evidence category of “Evidence of probable benefit - more studies needed” has been applied and relates to Ig therapy in the treatment of dysphagia rather than muscle weakness. Long term treatment does not appear justified.
Diagnosis Requirements	A diagnosis must be made by an Immunologist, Neurologist or a Rheumatologist.

Qualifying Criteria for IVIg Therapy	• Biopsy proven inclusion body myositis (IBM) with dysphagia (unless absolute contraindication) AND • Dysphagia limits dietary intake with involvement of pharyngeal muscles as demonstrated by videofluoroscopy OR • Speech pathology assessment indicates that video fluoroscopy is associated with an unacceptable risk of aspiration for this patient AND • Intolerance for solid dietary textures OR • At least two documented episodes of aspiration for which there is no better explanation IVIg should be used for up to four months (induction plus three maintenance cycles) before determining whether dysphagia has improved. If there is no benefit after this treatment, IVIg therapy should be abandoned. Review by a neurologist, rheumatologist, or immunologist is required within four months and annually thereafter. Once patients appear stable a trial off treatment should be considered. Documentation of clinical effectiveness is necessary for continuation of IVIg therapy.
Exclusion Criteria	Inclusion Body Myositis (IBM) with limb weakness without dysphagia affecting function Inflammatory myopathies: polymyositis (PM), dermatomyositis (DM), necrotising autoimmune myopathynecrotising autoimmune myopathy - see Inflammatory myopathies: polymyositis, dermatomyositis and necrotising autoimmune myopathy

Review Criteria for Assessing the Effectiveness of IVIg Use	IVIg should be used for up to four months (induction plus three maintenance cycles) before determining whether dysphagia has improved. If there is no benefit after this treatment, IVIg therapy should be abandoned. Review by a neurologist, rheumatologist, or immunologist is required within four months and annually thereafter. Once patients appear stable a trial off treatment should be considered. Documentation of clinical effectiveness is necessary for continuation of IVIg therapy. **Clinical effectiveness of Ig therapy may be assessed by:** **On review of the initial authorisation period** • Improvement in symptoms of dysphagia including as assessed by speech therapist, improvement in dietary intake and reduction in aspiration episodes, as relevant **On review of a continuing authorisation period** • Continued improvement in or stabilisation of symptoms of dysphagia including improvement in speech therapy assessment and improvement in dietary intake or aspiration episodes, as relevant AND • A trial of Ig weaning towards cessation of Ig therapy is planned for patients who are clinically stable or a reason provided as to why a trial is not planned
Dose	• **Induction Dose** - 2 g/kg in 2 to 5 divided doses • **Maintenance Dose** - 0.4–1 g/kg, 4–6 weekly. A maximum total dose of 1g/kg may be given in any 4 week period. This can be administered in weekly divided doses, provided total maximum is not exceeded. The aim should be to use the lowest dose possible that achieves the appropriate clinical outcome for each patient. Refer to the current product information sheet for further information on dose, administration and contraindications.

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