

## Inflammatory Myopathies: Inclusion Body Myositis (IBM)

Condition for which Ig has an established therapeutic role.

<b>Specific Conditions</b>	<ul style="list-style-type: none"><li>• Inclusion Body Myositis (IBM)</li></ul>
<b>Indication for Ig Use</b>	<ul style="list-style-type: none"><li>• Patients with inclusion body myositis (IBM) who have dysphagia limiting dietary intake</li></ul>
<b>Level of Evidence</b>	Evidence of probable benefit – more research needed (Category 2a)
<b>Description and Diagnostic Criteria</b>	<p>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.</p>
<b>Justification for Evidence Category</b>	<p>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.</p> <p>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.</p> <p>Long term treatment does not appear justified.</p>
<b>Diagnosis Requirements</b>	A diagnosis must be made by an Immunologist, Neurologist or a Rheumatologist.

## Qualifying Criteria for Ig 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 myopathy - see [Inflammatory myopathies: polymyositis, dermatomyositis and necrotising autoimmune myopathy](#)

## Review Criteria for Assessing the Effectiveness of Ig 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 (IVIg)** - 2 g/kg in 2 to 5 divided doses
- **Maintenance Dose (IVIg)** - 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 smaller 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.

## Bibliography

Association of British Neurologists, 2005, 'Guidelines for the use of intravenous immunoglobulin in neurological diseases', *The Association, London*, Available from: [www.theabn.org/abn/userfiles/file/IVIg-guidelines-final-July05.pdf](http://www.theabn.org/abn/userfiles/file/IVIg-guidelines-final-July05.pdf). [cited 7 Dec 2007]

Biotext 2004, 'Summary data on conditions and papers', in *A systemic literature review and report on the efficacy of intravenous immunoglobulin therapy and its risks* <https://www.blood.gov.au/system/files/A-systematic-literature-review-and-report-on-the-efficacy-of-IVIg-therapy-and-its-risks.pdf>

Cherin, P, Pelletier, S, Teixeira, A, et al 2002, 'Results and long-term follow up of intravenous immunoglobulin infusions in chronic, refractory polymyositis: an open study with thirty-five adult patients', *Arthritis & Rheumatism*, vol. 46, no. 2, pp. 467–74.

Choy, EHS, Hoogendijk, JE, Lecky, B, et al 2005, 'Immunosuppressant and immunomodulatory treatment for dermatomyositis and polymyositis (Cochrane Review)', in *The Cochrane Library*, Issue 3, John Wiley & Sons, Ltd, Chichester, UK.

## Bibliography

- Dalakas, MC, Illa, I, Dambrosia, JM, et al 1993, 'A controlled trial of high-dose intravenous immune globulin infusions as treatment for dermatomyositis', *New England Journal of Medicine*, vol. 329, no. 27, pp. 1993–2000.
- Dalakas, MC, Sonies, B, Dambrosia, J, et al 1997, 'Treatment of inclusion body myositis with IVIg: a double-blind, placebo-controlled study', *Neurology*, vol. 48, no. 3, pp. 712–6.
- Dalakas, MC, Koffman, B, Fujii, M, et al 2001, 'A controlled study of intravenous immunoglobulin combined with prednisone in the treatment of IBM', *Neurology*, vol. 56, no. 3, pp. 323–7.
- Dalakas, MC, 2004, 'The use of intravenous immunoglobulin in the treatment of autoimmune neuromuscular diseases: evidence-based indications and safety profile', *Pharmacology & Therapeutics*, vol. 102, no. 3, pp. 177–93.
- Dalakas, MC, 2005, 'Polymyositis, dermatomyositis, and inclusion body myositis', in DL, Kasper, E, Braunwald, AS, Fauci, et al (eds), *Harrison's Textbook of Medicine*, 16th edn, McGraw-Hill, New York, pp. 2540–45.
- Kornberg, AJ, for the Asia–Pacific IVIg Advisory Board, 2004, '*Bringing consensus to the use of IVIg in neurology. Expert consensus statements on the use of IVIg in neurology*', 1st edn, Asia–Pacific IVIg Advisory Board, Melbourne.
- Walter, MC, Lochmuller, H, Toepfer, M, et al 2000, 'High-dose immunoglobulin therapy in sporadic inclusion body myositis: a double-blind, placebo-controlled study', *Journal of Neurology*, vol. 247, no. 1, pp. 22–8.
- Wiles, CM, Brown, P, Chapel, H, et al 2002, 'Intravenous immunoglobulin in neurological disease: a specialist review', *Journal of Neurology, Neurosurgery and Psychiatry*, vol. 72, no. 4, pp. 440–8.

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