Inflammatory myopathies: polymyositis (PM), dermatomyositis (DM) and inclusion body myositis (IBM)

Version: 2.1
Published: 09 July 2016

Condition for which IVIg has an established therapeutic role.

Specific Conditions	• Polymyositis • Dermatomyositis • Inclusion body myositis
Indication for IVIg Use	• Polymyositis (PM) or dermatomyositis (DM) with significant muscle weakness unresponsive to corticosteroids and other immunosuppressive agents. • Inclusion body myositis (IBM) with dysphagia affecting function. • Rapidly progressive inclusion body myositis (IBM).
Level of Evidence	Evidence of probable benefit – more research needed (Category 2a)
Description and Diagnostic Criteria	The inflammatory myopathies are a group of three discrete disorders of skeletal muscle: DM, PM and IBM. These disorders are acquired and have in common the occurrence of significant muscle weakness and the presence of an inflammatory response within the muscle. The diagnosis of DM, PM or IBM is usually made by neurologists or rheumatologists, and relies on the combination of careful clinical evaluation, an elevated creatine kinase level, electromyography and muscle biopsy.
Justification for Evidence Category	PM: The Biotext (2004) review included one prospective case-series study of 35 adults with chronic refractory polymyositis. Intravenous immunoglobulin (IVIg) may be of benefit in these patients, improve mean muscle power and allow reduction in dose of corticosteroid. Further research is needed. DM: The Biotext (2004) review included one double-blind, placebo-controlled trial considered of low quality of 15 patients with biopsy-confirmed, treatment-resistant dermatomyositis. IVIg treatment combined with prednisone led to significant improvement in muscle strength and neuromuscular symptoms of patients in the intervention group (n = 8). IBM: The Biotext (2004) review included three small controlled studies, two of which had a crossover design. A total sample of 77 patients diagnosed with IBM was followed for between 4 and 12 months. The three studies showed possible slight benefit in reducing endomysial inflammation, disease progression and severity of IBM. Further research is needed. One submission reported the effectiveness of IVIg therapy for PM and DM as add-on therapy for patients who have not responded to steroids and immunosuppression (NSW IVIg User Group). A further submission confirms a role for IVIg as add-on maintenance therapy in some patients, resulting in an increased chance of complete remission and reduction in corticosteroid dose. A third submission suggests that IVIg can be tried as add-on treatment for patients with PM or DM who have not responded adequately to corticosteroids and second-line immunosuppressive agents (Asia–Pacific IVIg Advisory Board 2004).

	Weak evidence suggests that it may benefit patients with dysphagia associated with IBM (Asia–Pacific IVIg Advisory Board 2004).
Diagnosis Requirements	A diagnosis must be made by an Immunologist, Neurologist or a Rheumatologist.
Qualifying Criteria for IVIg Therapy	Polymyositis (PM) or dermatomyositis (DM) with significant muscle weakness unresponsive to corticosteroids and other immunosuppressive agents. • Significant muscle weakness, as measured by activities of daily living (ADL), medical research council (MRC) or other functional/disability scales. OR • Dysphagia affecting function. AND • No response to corticosteroid treatment. OR • Corticosteroids are contraindicated. AND • No response to immunosuppressive agents. OR • Immunosuppressive agents are contraindicated. Inclusion body myositis (IBM) with dysphagia affecting function. • Dysphagia affecting function. Rapidly progressive inclusion body myositis (IBM). • Rapidly progressive and significant muscle weakness, as measured by activities of daily living (ADL), medical research council (MRC) or other functional/disability scales.
Exclusion Criteria	Expert consensus does not recommend IVIg to treat the limb weakness of IBM.

Review Criteria for Assessing the Effectiveness of IVIg Use	Polymyositis (PM) or dermatomyositis (DM) with significant muscle weakness unresponsive to corticosteroids and other immunosuppressive agents. IVIg should be used for three to six months (three to six courses) before determining whether the patient has responded. If there is no benefit after three to six courses, IVIg therapy should be abandoned. Review by a Neurologist, Rheumatologist, or Clinical Immunologist is required within six months and annually thereafter, and a trial off period should be considered at each annual review. Documentation of clinical effectiveness is necessary for continuation of IVIg therapy. **On review of the initial authorisation period** Clinical effectiveness of Ig therapy may be demonstrated by: • Improvement in muscle weakness compared to the qualifying assessment, as measured by activities of daily living (ADL), medical research council (MRC) or other functional/disability scales. OR • Improvement in symptoms of dysphagia compared to the qualifying assessment. **On review of a continuing authorisation period** Clinical effectiveness of Ig therapy may be demonstrated by: • Improvement in muscle weakness or stabilisation of symptoms compared to the previous review assessment, as measured by activities of daily living (ADL), medical research council (MRC) or other functional/disability scales. OR • Improvement in, or stabilisation of, symptoms of dysphagia compared to the previous review assessment.

Inclusion body myositis (IBM) with dysphagia affecting function.

IVIg should be used for three to six months (three to six courses) before determining whether the patient has responded. If there is no benefit after three to six courses, IVIg therapy should be abandoned.

Review by a Neurologist, Rheumatologist, or Clinical Immunologist is required within six months and annually thereafter, and a trial off period should be considered at each annual review.

Documentation of clinical effectiveness is necessary for continuation of IVIg therapy.

On review of the initial authorisation period

Clinical effectiveness of Ig therapy may be demonstrated by:

- Improvement in symptoms of dysphagia compared to the qualifying assessment.

On review of a continuing authorisation period

Clinical effectiveness of Ig therapy may be demonstrated by:

- Improvement in, or stabilisation of, symptoms of dysphagia compared to the previous review assessment.

Rapidly progressive inclusion body myositis (IBM).

IVIg should be used for three to six months (three to six courses) before determining whether the patient has responded. If there is no benefit after three to six courses, IVIg therapy should be abandoned.

Review by a Neurologist, Rheumatologist, or Clinical Immunologist is required within six months and annually thereafter, and a trial off period should be considered at each annual review..

Documentation of clinical effectiveness is necessary for continuation of IVIg therapy.

On review of the initial authorisation period

Clinical effectiveness of Ig therapy may be demonstrated by:

- Improvement in muscle weakness compared to qualifying assessment, as measured by activities of daily living (ADL), medical research council (MRC) or other functional/disability scales.

On review of a continuing authorisation period

Clinical effectiveness of Ig therapy may be demonstrated by:

- Improvement in, or stabilisation of, muscle weakness compared to previous review assessment, as measured by activities of daily living (ADL), medical research council (MRC) or other functional/disability scales.

Dose	Polymyositis (PM) or dermatomyositis (DM) with significant muscle weakness unresponsive to corticosteroids and other immunosuppressive agents. • **Induction Dose** - 2 g/kg in 2 to 5 divided doses. • **Maintenance Dose** - 0.4–1 g/kg, 4–6 weekly. **The aim should be to use the lowest dose possible that achieves the appropriate clinical outcome for each patient.** **Dosing above 1 g/kg per day is contraindicated for some IVIg products.** **Refer to the current product information sheet for further information.** Inclusion body myositis (IBM) with dysphagia affecting function. • **Induction Dose** - 2 g/kg in 2 to 5 divided doses. • **Maintenance Dose** - 0.4–1 g/kg, 4–6 weekly. **The aim should be to use the lowest dose possible that achieves the appropriate clinical outcome for each patient.** **Dosing above 1 g/kg per day is contraindicated for some IVIg products.** **Refer to the current product information sheet for further information.** Rapidly progressive inclusion body myositis (IBM). • **Induction Dose** - 2 g/kg in 2 to 5 divided doses. • **Maintenance Dose** - 0.4–1 g/kg, 4–6 weekly. **The aim should be to use the lowest dose possible that achieves the appropriate clinical outcome for each patient.** **Dosing above 1 g/kg per day is contraindicated for some IVIg products.** **Refer to the current product information sheet for further information.**

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Generated on: 3 April 2019