

Multifocal motor neuropathy (MMN)

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Condition for which IVIg has an established therapeutic role.

Specific Conditions	• Multifocal motor neuropathy with or without persistent conduction block
Indication for IVIg Use	• First-line therapy for MMN.
Level of Evidence	Clear evidence of benefit (Category 1)
Description and Diagnostic Criteria	MMN is a relatively rare disorder characterised by slowly progressive, asymmetric, predominately distal limb weakness without sensory impairment. Weakness often begins in the arms and the combination of weakness, wasting, cramps and fasciculations may suggest a diagnosis of motor neuron disease. However, clinical examination may demonstrate that the pattern of weakness follows the distribution of individual nerves rather than a spinal segmental pattern. Investigations will typically show conduction block on nerve conduction studies. Immunoglobulin M (IgM) anti-GM-1 antibodies have been reported in a large number of patients with MMN and provide confirmatory evidence, but are not essential for the diagnosis.
Justification for Evidence Category	The Biotext (2004) review found six low-quality case studies or crossover randomised controlled trials (RCTs) with a total sample size of 68 patients. A possible benefit of intravenous immunoglobulin (IVIg) treatment in these patients was observed, although five studies were not controlled. The Frommer and Madronio (2006) review found one high-quality systematic review (a Cochrane review) of four crossover RCTs with 34 patients. Evidence for improvement in muscle strength with IVIg and limited evidence of a reduction in disability after IVIg administration. Consensus statements assert that IVIg is the only safe treatment demonstrated to work in patients with MMN. It is recommended in those who have significant disability. Dose and monitoring is similar to chronic inflammatory demyelinating polyneuropathy. IVIg therapy is usually long term, but the minimum effective dose for each patient should be sought. Plasma exchange and steroids appear to cause a worsening in the condition of patients with MMN with conduction block. Regular maintenance doses of IVIg are needed. The National Guideline Clearinghouse recommends the use of IVIg in the treatment of patients with progressive, symptomatic MMN that has been diagnosed using electrophysiology, ruling out other possible conditions that may not respond to IVIg treatment.
Diagnosis Requirements	A diagnosis must be made by a Neurologist.
Qualifying Criteria for IVIg Therapy	• Typical clinical phenotype with or without persistent motor conduction block. AND • Demonstration of functional impairment/disability, as measured by activities of daily living (ADL) or other functional/disability scales.

Exclusion Criteria	Presence of upper motor neuron signs. Significant sensory impairment without an adequate alternative explanation.
Review Criteria for Assessing the Effectiveness of IVIg Use	IVIg should be used for three to six months (three to six courses) before determining whether the patient has responded. Most individuals will respond within three months, unless there is significant axonal degeneration whereby a six-month course will be necessary. If there is no benefit after three to six courses, IVIg therapy should be abandoned. Review by a Neurologist is required within six months and at least annually thereafter. Documentation of clinical effectiveness is necessary for continuation of Ig therapy. On review of the initial authorisation period Clinical effectiveness of Ig therapy can be demonstrated by: • Improvement in focal muscle weakness and disability as measured by at least one of functional activities of daily living (ADL) or quantitative muscle scores or Medical Research Council (MRC) muscle assessment or neuropathy score or other method. On review of a continuing authorisation period For stable patients on maintenance treatment, review by a Neurologist is required at least annually. Clinical effectiveness of Ig therapy can be demonstrated by: • Improvement in, or stabilisation of, focal muscle weakness and disability as measured by at least one of functional activities of daily living (ADL) or quantitative muscle scores or Medical Research Council (MRC) muscle assessment or neuropathy score or other method.
Dose	• Induction Dose - 2 g/kg in 2 to 5 divided doses. • Maintenance Dose - 0.4–2 g/kg, 2–6 weekly. The amount per dose should be titrated to the individual's response. Aim for the minimum dose to maintain optimal functional status. Dosing above 1 g/kg per day is contraindicated for some IVIg products. 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.

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