Lambert–Eaton myasthenic syndrome (LEMS)

Version: 2.1
Published: 09 July 2016

Condition for which IVIg has an established therapeutic role.

Specific Conditions	• Lambert–Eaton myasthenic syndrome
Indication for IVIg Use	• Short-term therapy for severely affected LEMS patients.
Level of Evidence	Evidence of probable benefit – more research needed (Category 2a)
Description and Diagnostic Criteria	LEMS is a disorder of neuromuscular transmission first recognised clinically in association with lung cancer, and subsequently in cases in which no neoplasm was detected. Patients with LEMS have a presynaptic neuromuscular junction defect. The clinical picture is characterised by proximal muscle weakness with augmentation of strength after exercise, mild oculomotor signs, depressed deep tendon reflexes and autonomic dysfunction (dry mouth, constipation, erectile failure).
Justification for Evidence Category	In the Biotext (2004) review, one systematic review (containing one randomised controlled trial [RCT] with 9 patients) and one case series of 7 patients with a crossover design were included. Intravenous immunoglobulin (IVIg) appeared to provide some benefit to patients with LEMS. However, both studies only included a small number of patients. Expert consensus states that IVIg produces temporary improvement in patients with LEMS. It therefore has a role as second-line therapy (Kornberg 2004, Asia–Pacific IVIg Advisory Board). One submission to the National Blood Authority reported on a randomised controlled trial that showed significant improvement in strength associated with a decline in the level of pathogenic antibodies (NSW IVIg User Group).
Diagnosis Requirements	A diagnosis must be made by a Neurologist.
Qualifying Criteria for IVIg Therapy	• Severely affected LEMS patients, as demonstrated by functional scores of activities of daily living (ADL) or quantitative muscle scores or Medical Research Council (MRC) muscle assessment. AND • Alternative therapy has failed.

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. 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 annually thereafter. A trial off period should be considered at each review. Documentation of effectiveness is necessary for continuation of IVIg therapy. **On review of the initial authorisation period** Clinical effectiveness of Ig therapy can be demonstrated by: • Improvement in symptoms compared to the qualifying assessment, as measured by activities of daily living (ADL) or quantitative muscle scores, or MRC muscle assessment. **On review of a continuing authorisation period** For stable patients on maintenance therapy, review by a Neurologist is required at least annually. Clinical effectiveness of Ig therapy can be demonstrated by: • Improvement in, or stablilisation of, symptoms compared to the previous review assessment, as measured by activities of daily living (ADL) or quantitative muscle scores, or MRC muscle assessment.
Dose	• **Induction Dose** - 2 g/kg in 2 to 5 divided doses. • **Maintenance Dose** - 0.4–1 g/kg, 2–6 weekly. Aim for minimum dose to maintain optimal functional status. **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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Generated on: 2 April 2019