Guillain–Barré syndrome (GBS)

Version: 2.1
Published: 07 July 2016

Condition for which IVIg has an established therapeutic role.

Specific Conditions	• Guillain–Barré syndrome
Indication for IVIg Use	• GBS and its variants with significant disability and progression.
Level of Evidence	Clear evidence of benefit (Category 1)
Description and Diagnostic Criteria	GBS is the commonest cause of acute flaccid paralysis in the West. The syndrome typically presents with rapidly progressive, relatively symmetrical ascending limb weakness consistent with a polyradiculoneuropathy and often with associated cranial nerve involvement. Motor signs and symptoms usually predominate over sensory signs and symptoms. Loss of tendon reflexes occurs in most cases. Major complications include respiratory failure and autonomic dysfunction. The disease is monophasic, reaching its nadir usually within two weeks, although arbitrary definition accepts a limit of four weeks. A plateau phase of variable duration follows the nadir before gradual recovery. Although recovery is generally good or complete in the majority of patients, persistent disability has been reported to occur in about 20% and death in 4 to 15% of patients. Intravenous immunoglobulin (IVIg) has been shown to have the same efficacy as plasma exchange. The choice is based on availability, practicality, convenience, cost, and ease or safety of administration (Asia–Pacific IVIg Advisory Group). **Investigations** There is no biological marker for GBS. It is diagnosed by clinical recognition of rapidly evolving paralysis with areflexia. Investigations include the following: • Cerebrospinal fluid (CSF) protein elevation, although the level may be normal in the first two weeks of illness. The CSF white cell count may rise transiently, but a sustained pleocytosis suggests an alternative diagnosis or association with an underlying illness (e.g. human immunodeficiency virus, HIV). • Electrophysiological studies may show changes after the first or second week of the illness, including conduction block, conduction slowing or abnormalities in F waves.
Justification for Evidence Category	One systematic review of nine randomised controlled trials (RCTs) of moderate quality found IVIg hastened recovery in adults with GBS to the same degree as plasma exchange (Biotext 2004). One low-quality RCT with a small sample size (n = 21), in which the randomisation of patients to the IVIg treatment group was skewed, was identified. Children who received IVIg treatment showed earlier signs of improvement, and disability scores were lower at four weeks than the placebo group (Frommer and Madronio 2006).

Qualifying Criteria for IVIg Therapy	• Significant disability objectively measured by GBS Disability Grade. OR • Bulbar or autonomic features of GBS variant with significant disability AND • Disease progression.
Review Criteria for Assessing the Effectiveness of IVIg Use	Review is not mandated for this indication however the following criteria may be useful in assessing the effectiveness of therapy. Clinical effectiveness of Ig therapy may be demonstrated by: • Improvement in disability at four weeks after Ig treatment as assessed by the GBS Disability Grade. 0 – healthy state 1 – minor symptoms and capable of running 2 – able to walk 10 metres or more without assistance but unable to run 3 – able to walk 10 metres across an open space with help 4 – bedridden or chairbound 5 – requiring assisted ventilation for at least part of the day 6 – dead
Dose	• **Dose** - 2 g/kg in 2 to 5 divided doses. Approximately 10% of patients relapse, which may require a second treatment with IVIg. A second dose must only be on the advice of and after assessment by a Neurologist. **Refer to the current product information sheet for further information.**

Bibliography

Association of British Neurologists 2005, *Guidelines for the use of intravenous immunoglobulin in neurological diseases*, The Association, London. Available from: http://www.theabn.org/media/docs/ABN%20publications/IVIg-guidelines-final-July05.pdf

Biotext 2004, 'Summary data on conditions and papers', in *A systematic literature review and report on the efficacy of intravenous immunoglobulin therapy and its risks*, commissioned by the National Blood Authority on behalf of all Australian Governments, pp.149–50. Available from: http://www.nba.gov.au/pubs/pdf/report-lit-rev.pdf

Frommer, M & Madronio, C 2006, *The use of intravenous immunoglobulin in Australia*. A report for the National Blood Authority, Part B: systematic literature review, Sydney Health Projects Group, University of Sydney, Sydney, pp. 32–4.

Hughes, RAC, Raphaël, J-C, Swan, AV, et al 2006, 'Intravenous immunoglobulin for Guillain– Barré syndrome (Cochrane Review)', in *The Cochrane Library*, Issue 1, John Wiley & Sons, Ltd, Chichester, United Kingdom.

Korinthenberg, R, Schessl, J, Kirschner, J, et al 2005, 'Intravenously administered immunoglobulin in the treatment of childhood Guillain–Barré syndrome: a randomized trial', *Paediatrics*, vol. 116, no. 1, pp. 8–14.

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, pp. 14–20.