Pemphigus vulgaris (PV)

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Condition for which IVIg has an emerging therapeutic role.

Specific Conditions	• Pemphigus vulgaris
Indication for IVIg Use	• Moderate to severe PV as an adjuvant to prolonged corticosteroid treatment.
Level of Evidence	Evidence of probable benefit – more research needed (Category 2a)
Description and Diagnostic Criteria	Pemphigus vulgaris (PV) is a rare but potentially fatal condition accounting for approximately 70% of pemphigus cases. While the cause is unknown, an immuno-genetic predisposition is well established. PV may also be drug-induced. Drugs reported to be most significantly associated with PV include penicillamine, captopril and other thiol-containing compounds. Rifampicin and emotional stress have recently been reported as triggers for PV. The oral cavity is almost always affected and erosions can be scattered and extensive, with subsequent dysphagia. Blistering and erosions secondary to the rupture of blisters may be painful and limit the patient's daily activities. Pemphigus may occur in patients with other autoimmune diseases, particularly myasthenia gravis and thymoma. **Prognosis** The severity and natural history of PV are variable. Before the advent of steroids, most patients with PV died. Treatment with systemic steroids has reduced the mortality rate to 5–15%. Most deaths occur during the first few years of disease and, if the patient survives five years, the prognosis is good. Early disease is easier to control than widespread disease, and mortality may be higher if therapy is delayed. Morbidity and mortality are related to the extent of disease, the maximum dose of corticosteroid required to induce remission, and the presence of other diseases.
Justification for Evidence Category	In a retrospective cohort study, 15 corticosteroid-dependent patients with moderate to severe PV were treated with intravenous immunoglobulin (IVIg) and followed over a mean period of 6.2 years. All 15 patients had a satisfactory clinical response to IVIg therapy. IVIg had a demonstrable corticosteroid-sparing effect and was considered a safe and effective alternative treatment in patients who were dependent on systemic corticosteroids or who developed significant adverse effects as a result of their use (Biotext 2004).
Diagnosis Requirements	A diagnosis must be made by a Dermatologist.

Qualifying Criteria for IVIg Therapy	• Moderate to severe disease. AND • Corticosteroids or immunosupressive agents are contraindicated. OR • Condition is unresponsive to corticosteroids and immunosuppressive agents. OR • Presenting with severe side effects to immunosuppressive therapy.
Review Criteria for Assessing the Effectiveness of IVIg Use	Review is required every six months by a Dermatologist and improvement must be demonstrated for continuation of supply. **On review of the initial authorisation period** Evidence of clinical effectiveness of Ig therapy, and criteria for continued use may include: • Reduction in the severity of disease compared to the previous review period **On review of a continuing authorisation period** Evidence of clinical effectiveness of Ig therapy, and criteria for continued use may inlcude: • Reduction in the severity of disease compared to the previous review period Consideration should be given to trial-off immunoglobulin (Ig) therapy once the patient has achieved stabilised disease or clinical remission.
Dose	• **Maintenance Dose** - efficacy demonstrated with doses of at least 2 g/kg per monthly treatment cycle. **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.**

Bibliography

Biotext 2004, 'Summary data on conditions and papers', *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. 240–1. Available from: http://www.nba.gov.au/pubs/pdf/report-lit-rev.pdf

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Bibliography

Sami, N, Oureshi, A, Ruocco, E, et al 2002, 'Corticosteroid-sparing effect of intravenous immunoglobulin therapy in patients with pemphigus vulgaris', *Archives of Dermatology*, vol. 138, pp. 1158–62.

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