Bullous pemphigoid (BP)

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Condition for which IVIg has an emerging therapeutic role.

Specific Conditions	• Bullous Pemphigoid
Indication for IVIg Use	• Bullous pemphigoid (BP) resistant to corticosteroids and immunosuppressant therapy or when these agents are contraindicated
Level of Evidence	Evidence of probable benefit – more research needed (Category 2a)
Description and Diagnostic Criteria	Bullous pemphigoid (BP) is a rare disease of elderly people characterised by tense blisters and vesicles, with a prominent inflammatory component. The cause is unknown. Lesions result from a failure of basal keratinocytes to adhere to the epidermal basement membrane. The course of BP is characterised by exacerbations and remissions. Pruritis is a common feature and an increase in pruritis may herald an exacerbation. In most patients, BP is not a life-threatening disease. The side effects of systemic immunosuppressive therapy need to be managed. In most patients, the disease spontaneously clears within two to three years and all medication can be stopped. In a small group, the disease recurs after treatment is stopped. Skin infection is the most common complication. A submission by the Australasian College of Dermatologists recommends IVIg use in BP only in severe cases where improvement with conventional therapy is not readily achieved.
Justification for Evidence Category	The 2003 Harvard consensus statement (Ahmed and Dahl 2003) identified a small study (17 cases) where patients who were on intravenous immunoglobulin (IVIg) therapy for at least three months benefited from the therapy. The same article mentioned another small study (15 cases) where patients with Bullous pemphigoid (BP) could not be controlled with high-dose systemic corticosteroids and multiple immunosuppressive agents. IVIg produced prolonged clinical remission sustained after IVIg therapy was discontinued. In 2012, a published small case series (including pooled case reviews from literature), reported that approximately 75 percent of patients with BP responded to IVIg treatment, especially when used early on in the course of disease.
Diagnosis Requirements	A diagnosis must be made by an Immunologist or a Dermatologist.

Qualifying Criteria for IVIg Therapy	• Moderate to severe BP disease confirmed by biopsy or demonstration of autoantibodies AND • Unresponsive to standard corticosteroid and immunosuppressant therapy (using steroids and at least two alternative medications or rituximab) OR • Corticosteroids and immunosuppressant agents are contraindicated or have resulted in unacceptable side effects or significant toxicity Review is required every six months by a dermatologist or immunologist. Response must be demonstrated at the initial review at six months and improvement must be demonstrated for continuation of supply. Dosing should be reduced progressively and consideration should be given to a trial-off immunoglobulin (Ig) therapy once the patient has achieved stabilised disease or clinical remission.
Review Criteria for Assessing the Effectiveness of IVIg Use	Review is required every six months by a dermatologist or immunologist. Response must be demonstrated at the initial review at six months and improvement must be demonstrated for continuation of supply. Dosing should be reduced progressively and consideration should be given to a trial-off immunoglobulin (Ig) therapy once the patient has achieved stabilised disease or clinical remission. **Clinical effectiveness of Ig therapy may be demonstrated by:** **On review of the initial authorisation period** • A reduction in the number and severity of lesions with greater than 30 percent improvement compared to the qualifying assessment **On review of a continuing authorisation period** • A reduction in the number and severity of lesions compared to the previous review or disease has stabilised AND • A trial of weaning towards cessation of Ig therapy is planned for patients who are clinically stable to identify those in remission or reason is provided as to why a trial is not planned
Dose	• **Maintenance Dose** - 2g/kg each 4 weeks Dosing should be reduced progressively and consideration should be given to a trial-off immunoglobulin (Ig) therapy once the patient has achieved stabilised disease or clinical remission. 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 on dose administration and contraindications.

Bibliography

Ahmed, AR & Dahl, MV, for the Consensus Development Group, 2003, 'Consensus statement on the use of intravenous immunoglobulin therapy in the treatment of autoimmune mucocutaneous blistering diseases', *Archives of Dermatology*, vol. 139, no. 8, pp. 1051–9.

Gaitanis, G, Alexis, I, Pelidou, SH, et al 2012, 'High-dose intravenous immunoglobulin in the treatment of adult patients with bullous pemphigoid', *European Journal of Dermatology*, vol. 22, no. 3, pp. 363–9.

Orange, JS, Hossny, EM, Weiler, CR, et al 2006, 'Use of intravenous immunoglobulin in human disease: A review of primary evidence by members of the Primary Immunodeficiency Committee of the American Academy of Allergy, Asthma and Immunology', *Journal of Allergy and Clinical Immunology*, vol. 117, no. 4, pp. S525–53.

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