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 topical and systemic glucocorticoids and immunosuppressive therapy.
Level of Evidence	Insufficient data (Category 4a)
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 six 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 intravenous immunoglobulin (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 IVIg therapy for at least three months benefited from the therapy. The same article mentioned another small study (15 cases) where patients with 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.
Diagnosis Requirements	A diagnosis must be made by a Dermatologist.
Qualifying Criteria for IVIg Therapy	• Moderate to severe disease. AND • Corticosteroids or immunosuppressive agents are contraindicated. OR • Condition is unresponsive to corticosteroids and immunosupressive agents. OR • Presenting with severe side effects of therapy. The Australasian College of Dermatologists recommends IVIg use in BP only in severe cases where improvement with conventional therapy is not readily achieved.

Review Criteria for Assessing the Effectiveness of IVIg Use	Review is required by a Dermatologist or Clinical Immunologist every six months. Response must be demonstrated at the initial review and improvement demonstrated for continuation of supply. A trial off period should be considered at each review. Response can be measured by a reduction in recurrence of disease or relapse, ability to reduce dose or discontinue other therapies, improved quality of life, resolution of blisters and healing of affected skin or resolution of pruritus. **On review of the initial authorisation period** Evidence of clinical effectiveness of Ig therapy, and criteria for continued use may include: • Reduction in the number and severity of lesions compared to the qualifying assessment. AND • Active disease persists. **On review of a continuing authorisation period** Evidence of clinical effectiveness of Ig therapy, and criteria for continued use may include: • Reduction in the number and severity of lesions compared to the previous review.
Dose	• **Maintenance Dose** - At least 2 g/kg per monthly treatment cycle. Consideration should be given to trial-off immunoglobulin (Ig) therapy once the patient has achieved stabilised disease or clinical remission. The minimal effective dose should be prescribed. **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

Ahmed, AR & Dahl, MV 2003, 'Consensus statement on the use of intravenous immunoglobulin therapy in the treatment of autoimmune mucocutaneous blistering diseases', for the Consensus Development Group, *Archives of Dermatology,* vol. 139, pp. 1051–9.

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.