Pemphigus foliaceus (PF)

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Condition for which IVIg has an emerging therapeutic role.

Specific Conditions	• Pemphigus erythematosus • Pemphigus herpetiformis • Endemic pemphigus foliaceus • IgA pemphigus foliaceus • Paraneoplastic pemphigus foliaceus • Drug-induced pemphigus foliaceus
Indication for IVIg Use	• Pemphigus foliaceus (PF) 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	Pemphigus foliaceus (PF) is a rare autoimmune blistering skin disease characterised by loss of cohesion of cells (acantholysis) in the superficial (subcorneal) layers of the epidermis. The lesions are generally well demarcated and do not coalesce to form large eroded areas (as seen in pemphigus vulgaris). It is mediated by an autoantibody that targets desmoglein 1, a cell-to-cell protein molecule that binds the desmosomes of neighbouring keratinocytes in the epidermis. The disease has a long-term course with patients maintaining satisfactory health. Spontaneous remissions occasionally occur.
Justification for Evidence Category	Habif (2004) concluded that intravenous immunoglobulin (IVIg) was effective as monotherapy for pemphigus foliaceus (PF) and particularly useful in patients who experienced life-threatening complications from immunosuppression. Sami et al (2002) observed that autoantibody titres to desmoglein 1 in a series of 15 PF patients declined persistently following IVIg therapy. A more recent randomised controlled trial for pemphigus vulgaris (PV) and pemphigus foliaceus (PF) patients (61 patients in total) demonstrated both safety and efficacy of Ig therapy with monthly doses of up to 2 g/kg divided over five days (Amagai, 2009).
Diagnosis Requirements	A diagnosis must be made by an Immunologist or a Dermatologist.
Qualifying Criteria for IVIg Therapy	• Severe widespread PF disease involving at least 30 percent body surface confirmed by biopsy with positive direct immunofluorescence (DIF) test or demonstration of autoantibodies AND • Unresponsive to standard corticosteroid and immunosuppressant therapy using rituximab or two alternative immunosuppressant agents 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 and improvement must be demonstrated for continuation of supply. Dosing should be reduced progressively. Treatment is stopped when patients are clinically free from disease and have a negative finding on direct immunofluorescence.

Review Criteria for Assessing the Effectiveness of IVIg Use	Review is required every six months by a dermatologist or immunologist and improvement must be demonstrated for continuation of supply. Dosing should be reduced progressively. Treatment is stopped when patients are clinically free from disease and have a negative finding on direct immunofluorescence. **Clincial effectiveness of Ig therapy may be assessed by:** **On review of the initial authorisation period** • A reduced percentage of body surface area affected or other clinical improvement compared to the qualifying assessment AND • The direct immunofluorescence test remains positive and/or active disease persists **On review of a continuing authorisation period** • A reduced percentage of body surface area affected compared to previous review AND • The direct immunofluorescence test remains positive and/or active disease persists AND • A trial of weaning towards cessation of Ig therapy is planned for patients who are clinically stable to identify those in remission or a reason is provided as to why a trial-off is not planned
Dose	• **Maintenance Dose** - Efficacy can be achieved with dosing of up to 2 g/kg per month. Dosing should be reduced progressively and consideration should be given to a trial-off immunoglobulin (Ig) therapy once the patient has achieved 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

Amagai, M, Ikeda, S, Shimizu, H, et al 2009, 'A randomized double-blind trial of intravenous immunoglobulin for pemphigus', *Journal of the American Academy of Dermatology*, vol. 60, no.4, pp. 595-603. https://www.ncbi.nlm.nih.gov/pubmed/19293008

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