Pemphigus foliaceus (PF)

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Condition for which IVIg has an emerging therapeutic role.

Specific Conditions	• Pemphigus foliaceus
Indication for IVIg Use	• PF resistant to corticosteroids and immunosupressive therapy, or when these agents are contraindicated.
Level of Evidence	Insufficient data (Category 4a)
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 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.
Diagnosis Requirements	A diagnosis must be made by a Dermatologist.
Qualifying Criteria for IVIg Therapy	• Severe widespread PF disease involving at least 30% body surface area. AND • Unresponsive to standard corticosteroid and immunosuppressant therapy. OR • Severe side effects prohibit the continuation of corticosteroid and immunosuppressant agents. OR • Corticosteroids and/or immunosupressant agents are contraindicated.

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. Cessation of treatment should be considered at each review. **On review of an authorisation period** Clinical effectiveness of Ig therapy may be demonstrated by: • Reduced percentage of body surface area affected compared to the qualifying, or previous, assessment. AND • The auto-antibody titres and/or direct immunofluorescence reflect the response to systemic therapy. Clinical progression: treatment is stopped when patients are clinically free from disease and have a negative finding on direct immunofluorescence.
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

Habif, TP 2004. ' Vesicular and bullous diseases. Chapter 16' in *Clinical dermatology: a color guide to diagnosis and therapy* [electronic resource], 4th edition. Mosby Inc, Edinburgh.

Sami, N, Bhol, KC & Razzaque, A 2002, 'Influence of IVIg therapy on autoantibody titres to desmoglein 1 in patients with pemphigus foliaceus', *Clinical Immunology,* vol. 105, no. 2 pp. 192–8.

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