

Cicatricial pemphigoid (CP) or mucous membrane pemphigoid (MMP)

Condition for which Ig has an emerging therapeutic role.

Specific Conditions	<ul style="list-style-type: none">• Cicatricial pemphigoid (CP)• Mucous membrane pemphigoid (MMP)
Indication for Ig Use	<ul style="list-style-type: none">• Cicatricial pemphigoid (CP) / mucous membrane pemphigoid (MMP) resistant to corticosteroid 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	<p>Cicatricial pemphigoid (CP) or mucous membrane pemphigoid (MMP) is a rare, acquired subepithelial blistering disease characterised by erosive lesions of mucous membranes and skin. It is associated with autoantibodies to antigenic proteins in the epidermal basement membrane. Serious complications may occur due to erosions and scarring of affected tissues. Nasopharyngeal or laryngeal involvement may cause hoarseness, pain, tissue loss and even upper airway destruction, and oesophageal and urogenital lesions may lead to stenosis or strictures. CP/MMP is usually a chronic, progressive disorder.</p> <p>The aim of long-term treatment is cessation of the autoimmune process. Failure to do so results in invariable progression of the disease, culminating in progressive scarring. Permanent remission is usually possible if the disease is diagnosed early and treated sufficiently for one to five years.</p> <p>For the 70 percent of patients who have eye involvement, the disease progresses to conjunctival scarring and shrinkage, but may take 10 to 20 years to reach the end stage of bilateral blindness.</p>
Justification for Evidence Category	<p>A review of case reports and reports on small series of patients published in 2012 identified 72 patients who had received IVIg therapy for cicatricial pemphigoid (CP) or mucous membrane pemphigoid (MMP) (Czernik et al, 2012). The majority of patients experienced an improvement of disease manifestations and a decline in serum levels of autoantibodies to epidermal basement membrane antigens, where examined. Disease remissions for at least 12 months were common. Doses of IVIg given were usually higher than used for other autoimmune diseases, at 2-3 g/kg over 3-5 days every 2 weeks, because CP/MMP that is unresponsive to steroids and immunosuppressant therapy may cause considerable disability, particularly blindness from conjunctival ulceration and oesophageal strictures from oesophageal ulceration. Preliminary data suggest that a combination of IVIg and therapeutic B cell depletion through the use of rituximab arrests disease progression and prevents blindness in patients with conjunctival involvement that is unresponsive to corticosteroid and immunosuppressant therapy (Foster et al, 2010).</p>
Diagnosis Requirements	A diagnosis must be made by an Immunologist, Dermatologist or an Ophthalmologist.

Qualifying Criteria for Ig Therapy

Rituximab has been shown to be effective in the treatment of cicatricial pemphigoid (CP) / mucous membrane pemphigoid (MMP). A course of rituximab should be considered for the patient if not already trialled, unless strongly contraindicated.

- Moderate to severe CP/MMP disease with involvement of multiple sites, oesophageal involvement alone or conjunctiva alone, 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

- Corticosteroid and immunosuppressant medication are contraindicated or have resulted in unacceptable side effects or significant toxicity

Review is required every 6 months by a dermatologist, ophthalmologist or immunologist. Response must be demonstrated at the initial review at 6 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 therapy once the patient has achieved stabilised disease or clinical remission.

Review Criteria for Assessing the Effectiveness of Ig Use

Review is required every six months by a dermatologist, ophthalmologist 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 assessed by:

On review of the initial authorisation period

- A reduction in the number and severity of lesions 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 the disease has stabilised

AND

- A trial of weaning towards cessation of Ig therapy is planned for patients who are clinically stable, or a reason is provided as to why a trial is not planned

Dose

- **Maintenance Dose (IVIg)** - Initial treatment at 2 g/kg to gain control and reducing to the lowest dose that still adequately controls disease. A maximum of 3 g/kg in any 4 week period, with fortnightly dosing supported where eyesight is threatened.

Dosing should be reduced progressively and consideration should be given to a trial-off immunoglobulin 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

Bibliography

- treatment of autoimmune mucocutaneous blistering diseases', Archives of Dermatology, 139(8):1051–9, <https://doi.org/10.1001/archderm.139.8.1051>.
- Czernik A, Toosi S, Bystryn JC & Grando SA (2012) 'Intravenous immunoglobulin in the treatment of autoimmune bullous dermatoses: An update', Autoimmunity, 45(1):111-118, <https://doi.org/10.3109/08916934.2011.606452>.
- Daoud YJ & Amin KG (2006) 'Comparison of cost of immune globulin intravenous therapy to conventional immunosuppressive therapy in treating patients with autoimmune mucocutaneous blistering diseases', International Immunopharmacology, 6(4):600–6, <https://doi.org/10.1016/j.intimp.2005.11.002>.
- Foster CS, Chang PY & Ahmed AR (2010) 'Combination of rituximab and intravenous immunoglobulin for recalcitrant ocular cicatricial pemphigoid: a preliminary report', Ophthalmology, 117(5):861–9, <https://doi.org/10.1016/j.ophtha.2009.09.049>.
- Knudson RM, Kalaaji AN & Bruce AJ (2010) 'The management of mucous membrane pemphigoid and pemphigus', Dermatologic Therapy, 23(3):268-80, <https://doi.org/10.1111/j.1529-8019.2010.01323.x>.
- Letko E, Miserocchi E, Daoud YJ et al (2004) 'A nonrandomized comparison of the clinical outcome of ocular involvement in patients with mucous membrane (cicatricial) pemphigoid between conventional immunosuppressive and intravenous immunoglobulin therapies', Clinical Immunology, 111(3):303–10, <https://doi.org/10.1016/j.clim.2003.11.002>.
- 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, 117(4):S525–53, <https://doi.org/10.1016/j.jaci.2006.01.015>.
- Sobolewska B, Deuter C & Zierhut M (2013) 'Current medical treatment of ocular mucous membrane pemphigoid', The Ocular Surface, 11(4):259-66, <https://doi.org/10.1016/j.jtos.2013.02.003>.

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