Autoimmune retinopathy (AIR)

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Condition for which IVIg use is in exceptional circumstances only

Specific Conditions	• Autoimmune retinopathy
Indication for IVIg Use	• Autoimmune retinopathy (AIR) that is sight threatening and refractory to corticosteroid and immunosuppressant therapy
Level of Evidence	Insufficient data (Category 4a)
Description and Diagnostic Criteria	Autoimmune retinopathy (AIR) is the main disease within the broader condition of uveitis. AIR can be associated with severe, progressive and irreversible visual loss and demonstration of anti-retinal antibodies. Symptoms may include, blurred vision, dark floating spots in the vision field and sub-acute visual loss. Immunoglobulin (Ig) therapy may be used to treat autoimmune sight threatening retinopathy that is refractory to both corticosteroid and immunosuppressant therapy. Although rituximab has been used to treat AIR, it has had mixed success, and its access in Australia remains limited. The value of treatment may lie in arresting decline rather than causing an improvement in function. Indicators of disease status include visual acuity, visual field, electroretinography and integrity of macular anatomy by optical coherence tomography testing.
Justification for Evidence Category	While there is little high level evidence conclusively demonstrating benefit of Ig therapy, autoimmune retinopathy (AIR) is a very rare condition and open label studies and case series do demonstrate some benefit including recent publications (Castiblanco & Foster, 2014 and Garcia-Geremias, 2015). It is important to acknowledge that AIR is sight threatening and when refractory to corticosteroid and immunosuppressant therapy, observational studies conclude that Ig therapy is well-tolerated and effective in arresting disease in some patients.
Diagnosis Requirements	A diagnosis must be made by any specialist. The diagnosis must be verified by an Ophthalmologist.

Qualifying Criteria for IVIg Therapy	• Persistent severe disease threatening eyesight limited to the retinal plane confirmed by electroretinography (ERG) with integrity of macular anatomy confirmed by optical coherence tomography testing AND • Poorly responsive to oral or intravenous corticosteroid therapy for at least 12 weeks OR • Unable to tolerate corticosteroids for a period of 12 weeks due to unaccepatable side effects or significant toxicity OR • Corticosteroid therapy is contraindicated AND • Unresponsive to at least two immunosuppressant agents OR • Immunosuppressant medication is contraindicated or has resulted in unacceptable side effects or significant toxicity Review by a specialist in collaboration with an ophthalmologist is required within three months of treatment to determine whether the patient has responded, and annually thereafter. Documentation of clinical effectiveness is necessary for continuation of IVIg therapy.
Exclusion Criteria	Uveitis with features of anterior or posterior chamber inflammation
Review Criteria for Assessing the Effectiveness of IVIg Use	Review by a specialist in collaboration with an ophthalmologist is required within three months of treatment to determine whether the patient has responded, and annually thereafter. Documentation of clinical effectiveness is necessary for continuation of IVIg therapy. **Clinical effectiveness of Ig therapy can be assessed by:** **On review of the initial authorisation period** • Improvement in visual function or an arrest in the decline of visual function as determined by an ophthalmologist **On review of a continuing authorisation period** For stable patients on maintenance treatment, review by a specialist in collaboration with an ophthalmologist is required at least annually. **Clincial effectiveness of Ig therapy can be assessed by:** • Improvement in visual function or an arrest in the decline of visual function as determined by an ophthalmologist

Dose	• **Induction Dose** - 1.5g/kg in divided dose over 3 days. • **Maintenance Dose** - 0.4 to 1.5 g/kg in single or divided doses monthly. 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.

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