Evans syndrome - autoimmune haemolytic anaemia (AIHA) with immune thrombocytopaenia

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Condition for which IVIg has an emerging therapeutic role.

Specific Conditions	• Evans syndrome
Indication for IVIg Use	• To reduce platelet destruction and improve haemolysis in patients not responding to corticosteroid therapy.
Level of Evidence	Insufficient data (Category 4a)
Description and Diagnostic Criteria	Evans syndrome is a rare but serious autoimmune disease defined by the simultaneous or sequential occurrence of autoimmune haemolytic anaemia (AIHA) and immune thrombocytopenic purpura (ITP) without underlying aetiology. As such, it is a diagnosis of exclusion and other disorders, such as collagen vascular diseases, especially systemic lupus erythematosus (SLE) and scleroderma should be ruled out. The 2005 review by Norton and Roberts provided perspective on diagnosis, clinical features and management.
Justification for Evidence Category	A 2005 review on the management of Evans syndrome, based on Massachusetts Hospital data and a literature review, showed a transient response in all patients unless intravenous immunoglobulin (IVIg) was given every three weeks (Norton and Roberts 2006). The review concluded that the data supported a role for IVIg in first-line therapy. It was not clear whether it was important for steroids to be given at the same time, although this is common practice. A total dose of 2 g/kg in divided doses appeared to be sufficient. The review also stated that there might be a role for IVIg in preference to steroids in the acute setting in very young children.
Qualifying Criteria for IVIg Therapy	In patients with Evans Syndrome • Refractory to conventional therapy with corticosteroids. OR • Corticosteroids are contraindicated. OR • As a temporising measure before splenectomy.
Exclusion Criteria	Patients in whom a trial of corticosteroids has not been undertaken (providing corticosteroids are not contra-indicated and can be tolerated at the required doses).

Review Criteria for Assessing the Effectiveness of IVIg Use	Maintenance therapy is rarely required. In the rare circumstance where maintenance therapy is required, a six monthly review should be undertaken. Documentation of clinical effectiveness is necessary for continuation of IVIg therapy. Cessation of Ig treatment should be considered at each review. Clinical effectiveness of Ig therapy may be demonstrated by: • Improvement in clinical symptoms and signs, including resolution of haemolytic anaemia and improvement in platelet count
Dose	• **Initial Dose** - 0.8 - 2g/Kg in a divided dose. • **Maintenance Dose** - When indicated, 0.8-2g/kg as a single dose or divided dose 3 to 6 weekly **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

Darabi, K, Abdel-Wahab, O & Dzik, WH 2006, 'Current usage of intravenous immunoglobulin and the rationale behind it: the Massachusetts General Hospital data and review of the literature', *Transfusion*, vol. 46, no. 5, pp. 741–53.

Mathew, P, Chen, G & Wang, W 1997, 'Evans syndrome: results of a national survey', *Journal of Pediatric Hematology/Oncology*, vol. 19, no. 5, pp. 433–7.

Norton, A & Roberts, I 2006, 'Management of Evans syndrome', *British Journal of Haematology*, vol. 132, no. 2, pp. 125–37.

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