Haemophagocytic syndrome

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Condition for which IVIg has an emerging therapeutic role.

Specific Conditions	• Haemophagocytic syndrome
Indication for IVIg Use	• Management of severe haemophagocytic syndrome not responding to other treatments.
Level of Evidence	Insufficient data (Category 4a)
Description and Diagnostic Criteria	Haemophagocytic syndrome is characterised by fever, splenomegaly, jaundice, rash and the pathologic finding of haemophagocytosis (phagocytosis by macrophages of erythrocytes, leukocytes, platelets and their precursors) in bone marrow and other tissues with peripheral blood cytopenias. Haemophagocytic syndrome has been associated with a wide range of infectious, autoimmune, malignant and other disorders (modified from Fisman 2000). Mortality is high.
Justification for Evidence Category	No randomised controlled trials (RCTs) have been done, although many, mostly small, case series show evidence of benefit from intravenous immunoglobulin (IVIg) treatment.
Qualifying Criteria for IVIg Therapy	• Bone marrow diagnosis or other laboratory evidence supporting a diagnosis of haemophagocytosis. AND • Clinical features characteristic of haemophagocytic syndrome. AND • Non-response or ineligibility for other treatments.
Exclusion Criteria	Children with haemophagocytic lymphohistiocytosis (HLH) and hypogammaglobulinaemia — see secondary hypogammaglobulinaemia unrelated to haematological malignancy. - see Secondary hypogammaglobulinaemia (including iatrogenic immunodeficiency)
Review Criteria for Assessing the Effectiveness of IVIg Use	Review is not mandated for this indication however the following criteria may be useful in assessing the effectiveness of therapy. Clincial effectiveness of Ig therapy may be demonstrated by: • Survival and improvement in clinical and laboratory features.

Dose	• **Dose** - 2 g/kg is the most widely published dose. Emmenegger et al (2001) reported that better outcomes were associated with early administration of IVIg in their small case series (10 patients). **The aim should be to use the lowest dose possible that achieves the appropriate clinical outcome for each patient.** **Dosing above 1g/kg per day is contraindicated for some IVIg products.** **Refer to the current product information sheet for further information.**

Bibliography

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