Haemolytic transfusion reaction (hyperhaemolysis syndrome)

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Condition for which IVIg use is in exceptional circumstances only

Specific Conditions	• Hyperhaemolysis syndrome
Indication for IVIg Use	• Haemolytic transfusion reaction not due to alloantibodies when haemolysis of both donor and recipient red cells is suspected (hyperhaemolysis syndrome)
Level of Evidence	Insufficient data (Category 4a)
Description and Diagnostic Criteria	Hyperhaemolysis is an uncommon but potentially fatal type of delayed haemolytic transfusion reaction characterised by a drop in haemoglobin to below the pre-transfusion levels due to haemolysis of both donor and recipient red cells, reticulocytopenia and an absence of allo red cell antibodies. Although it is characteristically seen in sickle cell disease in childhood, there are reports in thalassaemia, myelofibrosis and lymphoma. The pathogenesis is poorly understood. Recommended treatment includes avoidance of transfusion where possible and immune modulation with high dose steroids and IVIg concurrently.
Justification for Evidence Category	There is a growing awareness of this uncommon complication of transfusions through the number of case reports over the last few years including in Australia. In the UK, hyperhaemolysis has been reportable to serious hazards of transfusion (SHOT) as a defined event since 2012.
Diagnosis Requirements	A diagnosis must be made by a Haematologist.
Qualifying Criteria for IVIg Therapy	• Severe haemolytic transfusion reactions where there is suspected haemolysis of both donor and recipient red cells as evidenced by a fall in haemoglobin below pre-transfusion levels AND • Ig therapy is given in conjunction with high dose corticosteroids
Exclusion Criteria	Autoimmune haemolytic anaemia (AIHA) - see Autoimmune haemolytic anaemia (AIHA) Alloimmune delayed haemolytic transfusion reaction with a positive direct agglutination test where haemolysis of only transfused red cells is suspected
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 Ig therapy. • Resolution of haemolysis, or stabilisation or rise in haemoglobin

Dose	• **Dose** - Up to 2 g/kg over 1 to 2 days. 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

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