Pure red cell aplasia (PRCA)

Version: 3.0
Published: 20 October 2018

Condition for which IVIg use is in exceptional circumstances only

Specific Conditions	• Pure red cell aplasia – associated B19 infection • Pure red cell aplasia – autoimmune mediated
Indication for IVIg Use	• Pure red cell aplasia (PRCA) associated with Parvo B19 infection in immunocompromised patients or autoimmune mediated refractory to immunosuppressant medication
Level of Evidence	Evidence of probable benefit – more research needed (Category 2a)
Description and Diagnostic Criteria	Pure red cell aplasia (PRCA) is a rare syndrome of severe anaemia, reticulocytopenia and a selective deficiency of erythroid progenitors. IVIg should be considered as first-line therapy for viral PRCA associated with parvovirus B19 in immunocompromised patients. IVIg is a reasonable option for patients with immunological PRCA who have failed other therapies (e.g. prednisone or cyclosporine).
Justification for Evidence Category	Whilst there is no randomised controlled trials for this condition there are many published case studies demonstrating benefit of Ig in the therapy of Parvovirus B19 associated Pure red cell aplasia (PRCA) in over 130 patients with defined Immunosuppressed states. These case series suggest doses of 2g/kg are most effective, with most response demonstrated with 1–3 doses. Several small case series suggest potential benefit in patients receiving Ig therapy for refractory PRCA following immunosuppressive therapies.
Diagnosis Requirements	A diagnosis must be made by a Haematologist.
Qualifying Criteria for IVIg Therapy	• Parvo B19 virus associated pure red cell aplasia (PRCA) in immunosuppressed patient proven by bone marrow biopsy OR • Immune-mediated PRCA proven by bone marrow biopsy refractory to treatment with at least two Immunosuppressant therapies OR • Immune-mediated PRCA proven by bone marrow biopsy and immune-suppressant medications are contraindicated or have resulted in unacceptable side effects or significant toxicity
Review Criteria for Assessing the Effectiveness of IVIg Use	Review is not mandated for this indication however the following indications may be useful in assessing the effectivness of Ig therapy. • Recovery of bone marrow Reduced transfusion dependence

Dose	• **Induction Dose** - 2 g/kg given by divided doses over 2 or 5 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.

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