Thames Valley
Strategic Clinical Network

Guidelines for the investigation and management of pure red cell aplasia in adults (age 16 years and above)

Definition
Pure red cell aplasia (PRCA) is a rare bone marrow (BM) disorder (incidence approximately 1 in 1 million per year) characterized by a reticulocytopenic normocytic anaemia and the disappearance of erythroid precursors. This guideline focuses on PRCA where no underlying cause has been found. Potential underlying causes include inherited and acquired conditions.

Inherited:
Diamond-Blackfan anaemia (DBA) ***– if identified, discuss with red cell consultant***

Acquired:
Virally mediated – particularly parvovirus B19 mediated transient red cell aplasia ***– often associated with haemolytic anaemia. Usually transient, consider intravenous immunoglobulin***
Medication-induced – ***review any medications as potential triggers and discontinue if possible, particularly erythropoietin, rifampicin and phenytoin.***
Large granular lymphocytic leukaemia (LGL) or other lymphoproliferative disorders ***– if identified, consider treatment for the underlying LGL***
Autoimmune disease such as SLE - ***treat underlying disorder***
Thymoma ***– consider surgical resection*** *(approx. 42% partial response rates; patients should also receive treatment as for primary PRCA)*
Common variable immunodeficiency (CVID) ***– discuss with immunology consultant***
ABO-incompatible allogeneic stem cell transplant ***– usually self-limiting and resolves within six months, discuss with bone marrow transplant consultant***

Initial investigations:
- FBC, reticulocyte count, DAT, renal and liver prolife, haematinics (ferritin, iron studies, B12, folate), blood film.
- Viral serology – HIV antigen/activity, Parvovirus B19 (IgM, IgG and PCR), EBV, CMV, hepatitis B core antibody, hepatitis B surface antigen, hepatitis C IgG
- Haemoglobin and serum electrophoresis
- TSH
- Erythropoietin level
- Bone marrow aspirate and trephine – morphology, myeloid: erythroid ratio, dysplastic changes, giant pronormoblasts, cytogenetics, T-cell receptor rearrangement

If history or investigations suggestive, consider investigations for:
- DBA - genetic testing on R92 panel (discuss with red cell team)
- LGL – peripheral blood flow cytometry, T-cell receptor rearrangement, Myeloid panel for STAT3 gene mutations
- Autoimmune disease - ANA
- Thymoma - CT thorax
- Consider investigations for malignancy

- Anti-erythropoietin antibodies (only if erythropoietin level is low)

Management:
Treat any known underlying cause.

For idiopathic PRCA:
- First line treatment is with oral ciclosporin 2.5mg/kg twice a day with target ciclosporin levels of 150 to 250 ng/mL.
- Consider prednisolone starting at 30mg once a day alongside ciclosporin
- Transfuse red cells as required
- Transfusion-related iron loading is common and the potential need for iron chelation should be considered early
- Second line treatment
    - Sirolimus 4mg for the first day and then 2mg once a day thereafter with target levels of 5 to 15 ng/m
    - Cyclophosphamide 50 to 100mg once a day

References

Gurnari and Maciejewski. How I manage pure red cell aplasia in adults. Blood 2021; 137(15): 2001-9

Versino F et al. Clinical Heterogeneity and Outcome of acquired PRCA: a Multicenter European Study. Blood Advances 2026; doi: 10.1182/bloodadvances.2025019525

Name	Revision	Date	Version	Review date
Dr Mike Desborough, Dr I-Jun Lau, Dr Noémi Roy	New protocol	Apr 2023	V0.2	Apr 2026
Dr Michael Desborough NSSG meeting	Agreed version at NSSG meeting	Jul 2023	V1.0	Jul 2026
Dr Michael Desborough	Update to include second line treatments	Jul 2026	V1.1	Jul 2029