Thames Valley Immunoglobulin Assessment Panel: Department of Health Clinical Guidelines –

Blue and Grey Indications Approved for Short Term Emergency Use Only

Short-term emergency use of IVIg may be appropriate in critically ill patients with the above conditions. Short term use of IVIg (as defined in the table for each condition) can be started without prior approval from the IVIg Panel (or the designated immunologist) provided the information requested in the table is clearly set out on the Immunoglobulin Request (National Database) Form. Please note the following:

- Short term use is defined as up to 2g/kg given in up to three doses at appropriate clinical intervals with all treatment occurring within three months. Any treatment given beyond the three month cut off (whether or not within the 'three dose' definition)will be considered as a separate episode and will require full authorisation by the Panel (ie the arrangements for automatic approval described in this policy will not apply).
- The prescribing clinician is responsible for providing the baseline (selection criteria) information and for providing subsequent outcome data.
- The selection criteria data must be included in the request form BEFORE pharmacy issue the IVIg.
- Outcome data must be provided by the clinician to the IVIg Panel AND recorded on the National Database.
- Invoices for patients which lack either or both selection criteria (baseline) data and/or outcome data will not be accepted by the Specialised Commissioning Group or the PCTs (the commissioners).
- The commissioners will not accept an invoice for any patient for whom a full, accurate data-set is not recorded on the National Database.

Table of Approved Short Term Emergency Indications for Immunoglobulin

Condition	Designation	Selection criteria	Outcome measures (must provide baseline and subsequent follow up values)	Dosing
Haematology				
Haemophagocytic syndrome	Blue	Diagnosis confirmed by consultant haematologist with bone marrow biopsy AND pancytopenia (provide details)	Pancytopenia Survival	Up to 2g/kg as a single or divided dose
Post-transfusion purpura	Blue	Sudden severe thrombocytopenia 5-10 days post-transfusion of blood products AND active bleeding (provide details)	Resolution of bleeding Increase in platelet count	2g/kg in divided doses over 2-5 consecutive days

Acquired red cell aplasia (including foetal hydrops)	Blue	Patients with parvovirus B19 infection confirmed by PCR; AND failure of other therapies (corticosteroid and at least one other immunosuppressive therapy) Foetal hydrops if likely to be associated with parvovirus B19 infection.	Correction of anaemia	2g/kg in two to five divided doses; repeated on relapse and for a second relapse
Auto-immune haemolytic anaemia (including Evans syndrome and post-transfusion hyperhaemolysis	Blue	Symptomatic or severe anaemia (Hb<6g/dL, except patients with co-morbidities) or thrombocytopenia (Evans syndrome, platelets $<20x10^9$/L) refractory to conventional treatment with steroids (or steroids contra-indicated); OR temporising measure prior to splenectomy.	Correction of anaemia/thrombocytopaenia	Up to 2g/kg as a single or divided dose
Neurology				
Myasthenia gravis (includes Lambert-Eaton myasthenic syndrome – LEMS)	Blue	Diagnosis of MG or LEMS by a neurologist; AND acute exacerbation (myasthenic crisis) for which other immunosuppressive treatments are ineffective/inappropriate. Short term use for other indications in MG/LEMS should be discussed/approved by the Trust designated immunoglobulin lead clinician before use.	Prescribing clinician must specify at least one of the following: • Forward arm abduction time • Quantitive MG Score (Duke) • Respiratory function measure (eg FVC) • Specific MG muscular score	2g/kg given over 2-5 days
Acute disseminated encephalomyelitis/idiopathic transverse myelitis	Grey	Diagnosis confirmed by a neurologist; Reduced level of consciousness (GCS<14); Plasma exchange not available or contra-indicated; Failure to respond to IV steroids and plasma-exchange.	• Improvement in level of consciousness (GCS) • Improvement in Expanded Disability Status Score (EDSS)	2g/kg

Auto-immune encephalitis	Grey	Diagnosis by a neurologist Probable/definite autoimmune encephalitic syndrome associated with autoantibodies where plasma exchange is inaccessible or contra-indicated Unexplained limbic encephalitis (LE) unresponsive to acyclovir Relapse in a patient with known LE	• Improvement in cognitive function • Resolution of seizures • Improvement in level of consciousness (GCS)	2g/kg
Infectious Diseases				
Necrotising (PVL-associated) sepsis	Blue	Diagnosis of streptococcal or staphylococcal toxic shock syndrome, preferably with isolation of organism; AND failure to achieve rapid improvement with antibiotic therapy and other supportive measures; AND life threatening	FBC, ALK and CPK Hospital in-patient stay (duration) Survival	2g/kg as a single dose
Severe or recurrent *Clostridium difficile* colitis	Blue	In severe cases not responding to oral vancomycin 125 mg qds, high dosage oral vancomycin +/- iv metronidazole 500mg tds is recommended. The addition of rifampicin or IVIg to this regime may be considered. Automatic authorisation for IVIg use in this situation applies only where situation is life-threatening. If situation less severe or if IVIg is being considered for use in a patient with multiple recurrences, FULL assessment by the IVIg panel is required.	Confirmation of C diff present at baseline followed by subsequent presence/absence at follow-up Hospital in-patient stay (duration)	0.4g/kg in one dose, may be repeated
Staphylococcal or streptococcal toxic shock syndrome	Blue	Diagnosis of streptococcal or staphylococcal toxic shock syndrome, preferably with isolation of organism; AND failure to achieve rapid improvement with antibiotic therapy and other supportive measures; AND life threatening	FBC, ALK and CPK Hospital in-patient stay (duration) Survival	2g/kg as a single dose

Rheumatology (Adult and Paediatric)				
Catastrophic antiphospholipid syndrome (adults)	Grey	Diagnosis confirmed by haematologist or rheumatologist, based on: • Demonstration of multiple small vessel thromboses • Presence of one or more phospholipid antibodies (ACL, anti-B_2GPI, LA) Plasmapheresis not available or contra-indicated; Unresponsive to plasmapheresis or conventional anti-thrombotic management	Survival	2g/kg

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