Catastrophic anti-phospholipid syndrome (CAPS)

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Condition for which IVIg use is in exceptional circumstances only

Specific Conditions	• Catastrophic anti-phospholipid syndrome
Indication for IVIg Use	• Confirmed diagnosis of CAPS with clinical deterioration post conclusion of steroids and plasmapheresis therapy or where therapies are contraindicated or plasmapheresis is not available
Level of Evidence	Insufficient data (Category 4a)
Description and Diagnostic Criteria	Catastrophic anti-phospholipid syndrome (CAPS) describes a rare accelerated form of anti-phospholipid syndrome characterised by widespread small vessel thrombosis leading to multi-organ failure. Occurring twice as often in women than men, diagnosis is demonstrated by clinical evidence of multiple organ involvement within seven days; histopathological evidence of multiple small vessel occlusions, and laboratory confirmation of the presence of antiphospholipid antibodies (aPL), usually in high titre. Although patients with CAPS represent less than one percent of all patients with anti-phospholipid syndrome (APS), the situation is usually life-threatening with mortality up to 40 percent. Optimal treatment uses steroids, plasmapheresis and anti-coagulation, however where plasmapheresis in not available or is contraindicated or where there is a deterioration after plasmapheresis, 2 g/kg Ig therapy over five days is indicated. Ig therapy is not indicated for chronic recurrent thrombosis; however, Rituximab has been shown to be effective in some individuals.
Justification for Evidence Category	Given the rarity of this condition, the evidence level is unlikely to extend beyond case reports and series. An international registry of patients with catastrophic anti-phospholipid syndrome (CAPS) (Cervera et al, 2016) was established in 2000 by the European Forum on anti-phospholipid antibodies and contains data from around 400 patients. Retrospective data analysis demonstrates that patients treated by plasmapheresis and Ig therapy demonstrate superior clinical outcomes.
Diagnosis Requirements	A diagnosis must be made by an Immunologist or a Haematologist.

Qualifying Criteria for IVIg Therapy	• Rapidly evolving thrombosis involving three or more organs within seven days AND • Laboratory evidence of anti-phospholipid antibodies (at least one of anticardiolipin, beta 2 glycoprotein I antibody or lupus anticoagulant) AND • Inadequate response to steroid therapy OR • Steroid therapy is contraindicated or has resulted in unacceptable side effects or significant toxicity AND • Clinical deterioration post plasmapheresis OR • Plasmapheresis is unavailable OR • Plasmapheresis is contraindicated
Exclusion Criteria	Chronic recurrent thrombosis
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.** • Survival of patient
Dose	• **Dose** - 2 g/kg over 5 days. Retreatment may be required in early relapse or occurrence of a second episode. A new request is required. 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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