Systemic capillary leak syndrome

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Condition for which IVIg use is in exceptional circumstances only

Specific Conditions	• Systemic capillary leak syndrome
Indication for IVIg Use	• Prevention of recurrent life-threatening episodes of hypotensive shock with hypoalbuminaemia in diagnosed Systemic capillary leak syndrome (SCLS)
Level of Evidence	Insufficient data (Category 4a)
Description and Diagnostic Criteria	Systemic capillary leak syndrome (SCLS) is an extremely rare condition that is characterised by recurrent life-threatening attacks of reversible capillary hyperpermeability accompanied by haemoconcentration and hypoalbuminaemia.
Justification for Evidence Category	Based on clearly documented studies, intravenous immunoglobulin (IVIg) prophylaxis was associated with an 89 percent decrease in the number of systemic capillary leak syndrome (SCLS) flares (252 pre-treatment to 29 post-treatment) in 27 patients for periods of up to 13 years (median duration of follow up 32 months). 15 out of 27 patients receiving IVIg experienced no SCLS episodes for periods of up to 12 years, and 24 out of 27 subjects had at least a 50 percent reduction in the number of flares. The optimal dose, schedule, and duration of IVIg therapy remain to be determined. While most patients (78 percent) received 2 g/kg/month, three patients have remained episode-free for greater than two years on 1 to 1.25 g/kg/month. Only a small number were controlled with 0.4 g/kg/month.
Diagnosis Requirements	A diagnosis must be made by an Immunologist, General Medicine Physician, Intensivist or an Emergency Medicine Physician.
Qualifying Criteria for IVIg Therapy	• Recurrent episodes of unexplained hypotension and oedema requiring hospital admission AND • Haemoconcentration, shock and hypoproteinemia as a result of the loss of plasma into the extravascular space AND • Other causes of shock, haemoconcentration and hypoproteinemia have been excluded Review by a General Physician or Immunologist is required within six months to assess the evidence of clinical benefit, and annually thereafter.

Review Criteria for Assessing the Effectiveness of IVIg Use	Review by a General Physician or Immunologist is required within six months to assess the evidence of clinical benefit, and annually thereafter. **Clinical effectiveness of Ig therapy may be assessed by:** **On review of the initial authorisation period** • Reduction in SCLS-related symptoms including oedema and hypotension post Ig treatment compared to the qualifying assessment AND • A reduction in the number of episodes requiring hospital admission compared to pre-treatment **On review of a continuing authorisation period** • Further improvement in or stabilisation of the number of episodes requiring hospital admission compared to the previous review period AND • A trial of Ig weaning towards cessation of Ig therapy is planned for patients who are clinically stable to identify those in remission or a reason provided as to why a trial is not planned
Dose	• **Induction Dose** - 1 - 2g/kg in single or divided doses. • **Maintenance Dose** - 0.4 - 2g/kg monthly. 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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