Toxic epidermal necrolysis (TEN; Lyell syndrome) / Stevens–Johnson syndrome (SJS)

Condition for which Ig has an emerging therapeutic role.

Specific Conditions	• Stevens–Johnson syndrome / toxic epidermal necrolysis overlap (SJS/TEN) • Toxic epidermal necrolysis (TEN)
Indication for Ig Use	• Toxic epidermal necrolysis (TEN) or Stevens–Johnson syndrome / toxic epidermal necrolysis overlap (SJS/TEN) with rapid evolution and >10% body surface area affected
Level of Evidence	Insufficient data (Category 4a)
Description and Diagnostic Criteria	Stevens–Johnson syndrome / toxic epidermal necrolysis overlap (SJS/TEN) and Toxic epidermal necrolysis (TEN) are severe mucocutaneous adverse reactions, most commonly triggered by medications such as sulphonamides, antibiotics, non-steroidal anti-inflammatory drugs (NSAIDs) and anti-convulsants. SJS and TEN are characterised by severe bullous reaction with extensive destruction of the epidermis, and morphologically by ongoing apoptotic keratinocyte cell death that results in the separation of the epidermis from the dermis. SJS and TEN are considered a disease continuum and are distinguished chiefly by severity, based upon the percentage of body surface involved with skin detachment. The term SJS describes patients with blistering and skin detachment involving a total body surface area of less than 10 percent, SJS/TEN overlap describes patients with 10–30 percent, and TEN refers to patient with greater than 30 percent skin detachment.
Justification for Evidence Category	Data surrounding the use of intravenous immunoglobulin (IVIg) for Stevens–Johnson syndrome / toxic epidermal necrolysis overlap (SJS/TEN) are limited and conflicting. Review of the literature provides no high-quality evidence supporting the use of Ig in SJS/TEN. Systematic review indicates that Ig therapy for patients with severe disease should be given in the early phase of the disease (i.e. within 24 to 48 hours of symptom onset). Systemic corticosteroids and Ig have been administered in combination to patients with SJS/TEN, but the data are too limited to draw any firm conclusions.
Diagnosis Requirements	A diagnosis must be made by an Immunologist or a Dermatologist.
Qualifying Criteria for Ig Therapy	• Onset of significant skin manifestations (painful red skin with or without blisters and/or any mucosal/conjunctival involvement) has occurred within the last 48 hours AND • Erythema and/or erosions affecting >10% body surface area OR • Significant mucosal lesions, including conjunctival erosions and labial blisters require early treatment IVIg should be initiated as early as possible, preferably within 24 hours of diagnosis. Urgent skin biopsy should be performed for confirmation but should not delay IVIg therapy if indicated.
Exclusion Criteria	Stevens–Johnson syndrome (SJS) alone
Review Criteria for Assessing the Effectiveness of Ig Use	**Review is not mandated for this condition however the following criteria may be useful in assessing the effectiveness of Ig therapy.** • Clinical assessment one month after immunoglobulin treatment

Dose	

- **Induction Dose (IVIg)** - Up to 3g/kg divided over 3 days (1g/kg/day), or up to 2g/kg delivered as a single dose. Dosages exceeding 2g/kg are not recommended due to the increased risk of adverse events.

IVIg should be initiated as early as possible, preferably within 24 hours of diagnosis.

Skin biopsy should be performed for confirmation, but should not delay IVIg therapy if indicated.

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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