

UK guidelines for the management of Stevens-Johnson syndrome/toxic epidermal necrolysis 2016

Creamer D, Walsh SA, Dziewulski P, Exton LS, Lee HY, Dart JKG, Setterfield J, Bunker CB, Ardern-Jones MR, Watson KMT, Wong GAE, Philippidou M, Vercueil A, Martin RV, Williams G, Shah M, Brown D, Williams P, Mohd Mustapa MF, Smith CH. *Br J Dermatol* 2016; 174: 1194-1227 & *J Plast Reconstr Aesthet Surg* 2016; 69: e119-e153.

<p>Initial assessment on presentation</p>	<ul style="list-style-type: none"> • Take a detailed history from the patient and/or relatives • Perform a full physical examination, including baseline body weight and record the vital signs, including oxygen saturation • Order a set of investigations: FBC, U&E, LFT, glucose, magnesium, phosphate, bicarbonate, mycoplasma serology, CXR, skin biopsy and baseline body weight • Initiate a primary management plan: <ol style="list-style-type: none"> 1. establish peripheral venous access 2. if patient cannot maintain adequate nutrition orally, insert a nasogastric tube and institute nasogastric feeding 3. insert a urinary catheter if urogenital involvement is causing significant dysuria/retention <p>(Strength of recommendation D (GPP))</p>
<p>Determination of drug causality</p>	<ul style="list-style-type: none"> • Identify causative agent and withdraw immediately <p>(Strength of recommendation D)</p>
<p>Prognostic scoring</p>	<ul style="list-style-type: none"> • Calculate SCORTEN within the first 24 hours <p>(Strength of recommendation C)</p>
<p>Care setting</p>	<ul style="list-style-type: none"> • A multi-disciplinary team should be convened, co-ordinated by a specialist in skin failure, usually dermatology and/or plastic surgery, and including clinicians from intensive care, ophthalmology and skin-care nursing • Patients with greater than 10% BSA epidermal loss should be admitted without delay to a Burn Centre or ICU with experience of treating patients with SJS/TEN and facilities to manage the logistics of extensive skin loss wound care • Patients must be barrier-nursed in a side room controlled for humidity, on a pressure-relieving mattress with the ambient temperature raised to between 25° and 28°C <p>(Strength of recommendation D (GPP))</p>
<p>Skin management regimen 1</p> <p><i>Applicable to all patients in all settings</i></p>	<ul style="list-style-type: none"> • Employ strict barrier nursing to reduce nosocomial infections • Take swabs for bacterial and candidal culture from three areas of lesional skin, particularly sloughy or crusted areas, on alternate days throughout the acute phase • Administer systemic antibiotics only if there are clinical signs of infection <p>(Strength of recommendation D (GPP))</p>
<p>Skin management regimen 2</p> <p><i>This may involve a conservative and/or surgical approach based on the specialist multi-disciplinary team's daily review of the individual needs of the patient</i></p>	<p>Institute a conservative approach in all patients as follows:</p> <ul style="list-style-type: none"> • Regularly cleanse wounds and intact skin by irrigating gently using warmed sterile water, saline or an antimicrobial such as chlorhexidine (1/5000) • Apply a greasy emollient, such as 50% white soft paraffin with 50% liquid paraffin (50/50 WSP/LP), over the whole epidermis, including denuded areas • Apply a topical antimicrobial agent to sloughy areas only (choice should be guided by local microbiological advice). Consider Ag-containing products/dressings. • The detached, lesional epidermis may be left <i>in situ</i> to act as a biological dressing. Blisters should be decompressed by piercing and expression or aspiration of tissue fluid. • Apply non-adherent dressings to denuded dermis (suitable dressings include Mepitelⁱ or Telfaiⁱ). • A secondary foam or burn dressing should be used to collect exudate (suitable dressings include Exu-Dry[®]). <p>Consider transfer to a Burn Centre in patients with TEN (>30% BSA epidermal loss) and evidence of the following: clinical deterioration, extension of epidermal detachment, sub-epidermal pus, local sepsis, wound conversion and/or delayed healing. In a Burn Centre conservative measures may be supplemented with a surgical approach.</p> <ul style="list-style-type: none"> • Remove necrotic/loose infected epidermis and clean wounds using a topical antimicrobial agent (e.g. betadine or chlorhexidine) under general anaesthetic • Consider debridement with Versajet[™] • Physiological closure with Biobrane/ allograft /xenograft skin in patients with early presentation involving non infected and large confluent areas <p>(Strength of recommendation D (GPP))</p>
<p>Fluid replacement regimen</p>	<ul style="list-style-type: none"> • Site venous lines through non-lesional skin, whenever possible, and change peripheral venous cannulas every 48 hours • Monitor fluid balance carefully: catheterize if appropriate/necessary • Establish adequate intravenous fluid replacement initially. Fluid replacement can be guided by urine output and other endpoint measurements. Individualized fluid management should be adjusted on a daily basis. • With improvement of SJS/TEN mouth involvement, oral administration of fluids should be progressively increased <p>(Strength of recommendation D)</p>

Nutrition regimen	<ul style="list-style-type: none"> • Provide continuous enteral nutrition throughout the acute phase • Deliver up to 20 to 25 kcal/kg/day during the early, catabolic phase and 25 to 30 kcal/kg/day during the anabolic, recovery phase <p>(Strength of recommendation C)</p>
Analgesia	<ul style="list-style-type: none"> • Use a patient appropriate validated pain tool to assess pain in all conscious patients at least once a day • Patients should receive adequate analgesia to ensure comfort at rest, with the addition of supplementary opiates, as required • Additional analgesia may be needed to address increased pain associated with patient handling, re-positioning and dressing changes <p>(Strength of recommendation D (GPP))</p>
Supportive Therapeutic Measures	<ul style="list-style-type: none"> • Immobile patients should receive low molecular weight heparin • Patients in whom enteral nutrition cannot be established should receive a proton pump inhibitor to reduce the risk of stress-related gastro-intestinal ulceration • Neutropenic patients may benefit from recombinant human G-CSF <p>(Strength of recommendation C)</p>
Treatment of eye involvement	<ul style="list-style-type: none"> • Daily ophthalmological review is necessary during the acute illness • Apply an ocular lubricant (e.g. non-preserved hyaluronate or carmellose eye drops) every two hours through the acute illness • Ocular hygiene must be carried out each day by an ophthalmologist or ophthalmic-trained nurse • Application of topical corticosteroid drops (e.g. non-preserved dexamethasone 0.1% twice a day) may reduce ocular surface damage • Administer a broad-spectrum topical antibiotic as prophylaxis (e.g. moxifloxacin drops four times a day) in the presence of corneal fluorescein staining or frank ulceration • In the unconscious patient, prevention of corneal exposure is essential <p>(Strength of recommendation D (GPP))</p>
Treatment of mouth involvement	<ul style="list-style-type: none"> • Daily oral review is necessary during the acute illness • Apply white soft paraffin ointment to the lips every two hours through the acute illness • Clean the mouth daily with warm saline mouthwashes or an oral sponge • Use an anti-inflammatory oral rinse or spray containing benzydamine hydrochloride every three hours, particularly before eating • Use an anti-septic oral rinse containing chlorhexidine twice a day • Use a potent topical corticosteroid mouthwash (e.g. betamethasone sodium phosphate) four times a day <p>(Strength of recommendation D (GPP))</p>
Treatment of urogenital involvement	<ul style="list-style-type: none"> • Daily urogenital review is necessary during the acute illness • Apply white soft paraffin ointment to the urogenital skin and mucosae every four hours through the acute illness • Use a potent topical corticosteroid ointment once a day to the involved, but non-eroded, surfaces • Use a silicone dressing (e.g. Mepitel™) to eroded areas <p>(Strength of recommendation D (GPP))</p>
Treatment of airway involvement	<ul style="list-style-type: none"> • Respiratory symptoms and hypoxaemia on admission should prompt early discussion with an intensivist and rapid transfer to an ICU or Burn Centre, where fibre-optic bronchoscopy should be undertaken <p>(Strength of recommendation D (GPP))</p>
Active therapy	<ul style="list-style-type: none"> • If active therapy is instituted it should be given, ideally, under the supervision of a specialist skin failure MDT in the context of clinical research and/or case registry <p>(Strength of recommendation D)</p>
Discharge and follow-up	<ul style="list-style-type: none"> • Give the patient written information about drug(s) to avoid • Encourage the patient to wear a MedicAlert bracelet • Drug allergy should be documented in the patient's notes; all doctors involved in the patient's care should be informed • Report the episode to the national pharmacovigilance authorities • Organize an out-patient clinic appointment, and if required an ophthalmology out-patient appointment, within a few weeks of discharge • Refer for review to unit with appropriate sub-speciality interest <p>(Strength of recommendation D (GPP))</p>
Diagnostic testing	<ul style="list-style-type: none"> • Routine drug hypersensitivity testing is not recommended following an episode of SJS/TEN. • Seek specialist advice on hypersensitivity testing where: <ol style="list-style-type: none"> 1. the culprit drug is not known or 2. medication avoidance is detrimental to the individual or 3. accidental exposure is possible <p>(Strength of recommendation D (GPP))</p>