

## 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"> <li>• Take a detailed history from the patient and/or relatives</li> <li>• Perform a full physical examination, including baseline body weight and record the vital signs, including oxygen saturation</li> <li>• Order a set of investigations: FBC, U&amp;E, LFT, glucose, magnesium, phosphate, bicarbonate, mycoplasma serology, CXR, skin biopsy and baseline body weight</li> <li>• Initiate a primary management plan:             <ol style="list-style-type: none"> <li>1. establish peripheral venous access</li> <li>2. if patient cannot maintain adequate nutrition orally, insert a nasogastric tube and institute nasogastric feeding</li> <li>3. insert a urinary catheter if urogenital involvement is causing significant dysuria/retention</li> </ol> </li> </ul> <p><b>(Strength of recommendation D (GPP))</b></p>
<p>Determination of drug causality</p>	<ul style="list-style-type: none"> <li>• Identify causative agent and withdraw immediately</li> </ul> <p><b>(Strength of recommendation D)</b></p>
<p>Prognostic scoring</p>	<ul style="list-style-type: none"> <li>• Calculate SCORTEN within the first 24 hours</li> </ul> <p><b>(Strength of recommendation C)</b></p>
<p>Care setting</p>	<ul style="list-style-type: none"> <li>• 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</li> <li>• 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</li> <li>• 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</li> </ul> <p><b>(Strength of recommendation D (GPP))</b></p>
<p>Skin management regimen 1</p> <p><i>Applicable to all patients in all settings</i></p>	<ul style="list-style-type: none"> <li>• Employ strict barrier nursing to reduce nosocomial infections</li> <li>• 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</li> <li>• Administer systemic antibiotics only if there are clinical signs of infection</li> </ul> <p><b>(Strength of recommendation D (GPP))</b></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><b>Institute a conservative approach in all patients as follows:</b></p> <ul style="list-style-type: none"> <li>• Regularly cleanse wounds and intact skin by irrigating gently using warmed sterile water, saline or an antimicrobial such as chlorhexidine (1/5000)</li> <li>• 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</li> <li>• Apply a topical antimicrobial agent to sloughy areas only (choice should be guided by local microbiological advice). Consider Ag-containing products/dressings.</li> <li>• 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.</li> <li>• Apply non-adherent dressings to denuded dermis (suitable dressings include Mepitel<sup>i</sup> or Telfai<sup>i</sup>).</li> <li>• A secondary foam or burn dressing should be used to collect exudate (suitable dressings include Exu-Dry<sup>®</sup>).</li> </ul> <p><b>Consider transfer to a Burn Centre in patients with TEN (&gt;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.</b></p> <ul style="list-style-type: none"> <li>• Remove necrotic/loose infected epidermis and clean wounds using a topical antimicrobial agent (e.g. betadine or chlorhexidine) under general anaesthetic</li> <li>• Consider debridement with Versajet<sup>™</sup></li> <li>• Physiological closure with Biobrane/ allograft /xenograft skin in patients with early presentation involving non infected and large confluent areas</li> </ul> <p><b>(Strength of recommendation D (GPP))</b></p>
<p>Fluid replacement regimen</p>	<ul style="list-style-type: none"> <li>• Site venous lines through non-lesional skin, whenever possible, and change peripheral venous cannulas every 48 hours</li> <li>• Monitor fluid balance carefully: catheterize if appropriate/necessary</li> <li>• 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.</li> <li>• With improvement of SJS/TEN mouth involvement, oral administration of fluids should be progressively increased</li> </ul> <p><b>(Strength of recommendation D)</b></p>

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