# Stevens-Johnson Syndrome

## Introduction

Stevens-Johnson syndrome (SJS) and toxic epidermal necrolysis (TEN) are severe mucocutaneous adverse reactions, most commonly triggered by medications, characterized by fever and extensive necrosis and detachment of the epidermis. 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.

SJS is the less severe condition, in which skin detachment is <10% of the body surface. Mucous membranes are affected in over 90 percent of patients, usually at two or more distinct sites (ocular, oral, and genital).

TEN involves detachment of >30% of the body surface area. Mucous membranes are also involved in over 90% of cases.

SJS/TEN overlap describes patients with skin detachment of 10 to 30% of body surface area. Mucous membranes are also involved in over 90% of cases.

Here, "SJS/TEN" is used to refer collectively to SJS, TEN, and SJS/TEN overlap.

## Target User

* Doctors
* Nurses

## Target area of use

* Ward
* Outpatient department

## Key areas of focus/new additions/changes

These guidelines addresses the management of Stevens Johnson.

## Limitations

Lack of an intensive care unit to nurse patient with extensive lesions.

## Presenting symptoms and signs

Typical prodromal symptoms of SJS are as follows:

* Cough productive of a thick, purulent sputum
* Headache
* Malaise
* Arthralgia

Patients may complain of a burning rash that begins symmetrically on the face and the upper part of the torso. This may be accompanied by ocular symptoms.

In addition to the skin, lesions in SJS may involve the following parts of the body:

* Oral mucosa
* Oesophagus
* Pharynx
* Larynx
* Anus
* Trachea
* Vagina
* Urethra

Ocular symptoms include the following: red eye, tearing, dry eye, pain, blepharospasm, itching, grittiness, heavy eyelid, foreign body sensation, decreased vision, burning sensation, photophobia, diplopia

## Examination findings

The rash can begin as macules that develop into papules, vesicles, bullae, urticarial plaques, or confluent erythema. The centre of these lesions may be vesicular, purpuric, or necrotic.

Lesions may become bullous and later rupture, leaving denuded skin. The skin becomes susceptible to secondary infection.

Urticarial lesions typically are not pruritic. Infection may be responsible for the scarring associated with morbidity.

Although lesions may occur anywhere, the palms, soles dorsum of the hands, and extensor surfaces are most commonly affected.

The rash may be confined to any one area of the body, most often the trunk. Mucosal involvement may include erythema, oedema, sloughing, blistering, ulceration, and necrosis.

The following signs may also be noted on examination:

* Fever
* Tachycardia
* Hypotension
* Altered level of consciousness, epistaxis
* Conjunctivitis
* Corneal ulceration
* Erosive vulvovaginitis or balanitis
* Seizures
* Coma

## Differential Diagnoses

* Trauma
* Irradiation
* Porphyria cutanea tarda
* Staphylococcal scalded skin syndrome
* Sebaceous cell carcinoma
* Adenoviral conjunctivitis
* Intraepithelial epithelioma
* Acute generalized exanthematic pustulosis
* Chemical burns

## Investigations

There are no specific laboratory studies (other than biopsy) that can definitively establish the diagnosis of SJS but the following investigations can be done:

* FBC, severely elevated WBC count indicates the possibility of a superimposed bacterial infection.
* U&E, LFT may be needed to help manage related problems
* Skin and blood cultures have been advocated because the incidence of serious bacterial bloodstream infections and sepsis contribute to morbidity and mortality.
* CXR may indicate the existence of a pneumonitis when clinically suspected. Otherwise, routine plain films are not indicated.
* Skin biopsy specimens demonstrate that the bullae are subepidermal. Epidermal cell necrosis may be noted. Perivascular areas are infiltrated with lymphocytes.

## Management

The main principles of supportive care are the same as for major burns and include wound care, fluid and electrolyte management, nutritional support, temperature management, pain control, and monitoring or treatment of superinfections

### Initiate a primary management plan:

* Establish peripheral venous access: Site venous lines through non-lesional skin, whenever possible, and change peripheral venous cannulas every 48 hours.
* 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, but replacement volumes are approximately one-third lower than those needed for burn victims.
* With improvement of SJS/TEN mouth involvement, oral administration of fluids should be progressively increased.

If patient cannot maintain adequate nutrition orally, insert a nasogastric tube and institute nasogastric feeding.

* Insert a urinary catheter if urogenital involvement is causing significant dysuria/retention.

### Prognostic scoring

The prognosis of individual patients can be rapidly evaluated on admission by applying a prognostic scoring system called **SCORTEN**. SCORTEN is based upon seven independent and easily measured clinical and laboratory variables and has been validated for use on days one and three of hospitalization for SJS/TEN.

|  |  |  |  |  |
| --- | --- | --- | --- | --- |
| **Prognostic factors** | **Score** |  | **SCORTEN** | **Mortality(%)** |
| Age > 40 years | 1 |  | 0-1 | 3 |
| Tachycardia >120 bpm | 1 |  | 2 | 12 |
| Neoplasia | 1 |  | 3 | 35 |
| Initial detachment > 10% | 1 |  | 4 | 58 |
| Serum urea > 10 mmol/L | 1 |  | ≥5 | 90 |
| Serum bicarbonate < 20 mmol/L | 1 |  |  |  |
| Blood glucose > 14 mmol/L | 1 |  |  |  |

The decision to refer the patient to an intensive care or burn unit should be made on a case-by-case basis, based upon the extent of skin involvement and the presence of comorbidities. Patients with a limited skin involvement, a SCORTEN score of 0 or 1, and disease that is not rapidly progressing may be treated in nonspecialized wards. Patients with more severe disease (skin detachment >30% of the body surface area) **or** a SCORTEN score ≥2 should be transferred to intensive care units, burn units, or specialized dermatology units, if available.

### Skin Care

* 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
* 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 emulsifying ointment, 50% white soft paraffin with 50% liquid paraffin, over the whole epidermis, including denuded areas
* Apply a topical antimicrobial agent to sloughy areas only (choice should be guided by local microbiological advice·
* The detached, lesional epidermis may be left *in situ* 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

### Analgesia

* 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

### Supportive therapeutic measures

* 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

### Ocular Care

* 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
* Application of topical corticosteroid drops (e.g. non-preserved dexamethasone 0.1% twice a day) may reduce ocular surface damage
* Administer topical antibiotic as prophylaxis (e.g. ciprofloxacin drops four times a day) in the presence of corneal ulceration
* In the unconscious patient, prevention of corneal exposure is essential.

### Mouth care

* 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

### Urogenital Caret

* Daily urogenital review is necessary during the acute illness
* Apply white soft paraffin ointment to the urogenital skin and mucosae every four hours throughout the acute illness
* Use a potent topical corticosteroid ointment once a day to the involved, but non-eroded surfaces.

### Nutritional care of airway

* 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.

### Discharge and follow up

* Give the patient written information about drug(s) to avoid
* Drug allergy should be documented in the patient’s notes; all doctors involved in the patient’s care should be informed
* 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

## Key Issues for Nursing care

* Sterile handling and/or reverse-isolation nursing techniques are essential to decrease the risk of nosocomial infection
* Daily ocular and mouth hygiene
* Effective and timely skin care

## References

1. [Bastuji-Garin S, Rzany B, Stern RS, et al. Clinical classification of cases of toxic epidermal necrolysis, Stevens-Johnson syndrome, and erythema multiforme. Arch Dermatol 1993; 129:92.](https://www.uptodate.com/contents/stevens-johnson-syndrome-and-toxic-epidermal-necrolysis-management-prognosis-and-long-term-sequelae/abstract/1)
2. [Creamer D, Walsh SA, Dziewulski P, et al. U.K. guidelines for the management of Stevens-Johnson syndrome/toxic epidermal necrolysis in adults 2016. Br J Dermatol 2016; 174:1194.](https://www.uptodate.com/contents/stevens-johnson-syndrome-and-toxic-epidermal-necrolysis-management-prognosis-and-long-term-sequelae/abstract/2)
3. [Harr T, French LE. Toxic epidermal necrolysis and Stevens-Johnson syndrome. Orphanet J Rare Dis 2010; 5:39.](https://www.uptodate.com/contents/stevens-johnson-syndrome-and-toxic-epidermal-necrolysis-management-prognosis-and-long-term-sequelae/abstract/3)
4. [Bastuji-Garin S, Fouchard N, Bertocchi M, et al. SCORTEN: a severity-of-illness score for toxic epidermal necrolysis. J Invest Dermatol 2000; 115:149.](https://www.uptodate.com/contents/stevens-johnson-syndrome-and-toxic-epidermal-necrolysis-management-prognosis-and-long-term-sequelae/abstract/4)
5. 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.
6. *Br J Dermatol* **2016**; 174: 1194-1227 & *J Plast Reconstr Aesthet Surg* **2016**; 69: e119e153

|  |  |  |
| --- | --- | --- |
| **Written by:** | Name: Amie Secka | Date: 30 November 2018 |
| **Reviewed by:** | Name: Orighomisan Agboghoroma | Date: 06 December 2018 |
| **Version:** | **Change history:** | **Review due date:** |
| 1.0 | New document | 31 January 2021 |
| 1.1 | Executive summary added | 31 January 2021 |
| Review Comments (*if applicable)* |  |  |