Steven Johnson Syndrome and Toxic Epidermal Necrolysis

SJS and TEN are part of the same spectrum of skin reaction resulting in sheet like skin loss with mucosal involvement. It is nearly always medication related and is more common in association with HIV infection.

Drugs that most commonly cause SJS/TEN

Antibiotics Antifungals Antivirals	▪ Sulfonamides, e.g., cotrimoxizole; beta-lactams i.e., penicillins, cephalosporins ▪ Imidazole antifungals ▪ Nevirapine (non-nucleoside reverse-transcriptase inhibitor)
Allopurinol	
Nonsteroidal anti-inflammatory drugs (NSAID) (oxicam type mainly)	▪ Naproxen ▪ Ibuprofen
Anti-convulsants	▪ Carbamazepine ▪ Phenytoin ▪ Phenobarbital ▪ Valproic acid ▪ Lamotrigine

Clinical Features

- Prodromal illness – flu-like with high fever.
- Tender rash, trunk initially then face and limbs.
- Macules (flat red areas), purple spots, target lesions (bulls eye-like, as in erythema multiforme) and flaccid blisters.

Mucosal involvement is prominent and severe, although not forming actual blisters. At least 2 mucosal surfaces are affected including:

- Eyes (conjunctivitis) – red, sore, sticky
- Lips/mouth (cheilitis, stomatitis) – red crusted lips, mouth ulcers
- Oesophagus – causing difficulty eating
- Upper respiratory tract (trachea and bronchi) – causing cough and respiratory distress
- Genital area and urinary tract – ulcers
- Gastrointestinal tract – causing diarrhoea.

Diagnosis

Classification of TEN

SJS	▪ Skin detachment <10% of body surface area (BSA) ▪ Widespread erythematous or purpuric macules or flat atypical targets
Overlap SJS/TEN	▪ Detachment between 10% and 30% of BSA ▪ Widespread purpuric macules or flat atypical targets
TEN with spots	▪ Detachment >30% of BSA ▪ Widespread purpuric macules or flat atypical targets
TEN without spots	▪ Detachment of >10% of BSA ▪ Large epidermal sheets and no purpuric macules

Diagnosis is clinical but skin biopsy is usually required to exclude Staphylococcal Scalded Skin Syndrome (SSSS).

SCORTEN

SCORTEN is an illness severity score that has been developed to predict mortality in SJS and TEN cases. One point is scored for each of seven criteria present at the time of admission. The SCORTEN criteria are:

- Age >40 years
- Presence of a malignancy (cancer)
- Heart rate >120
- Initial percentage of epidermal detachment >10%
- Serum urea level >10 mmol/L
- Serum glucose level >14 mmol/L
- Serum bicarbonate level <20 mmol/L

The risk of dying from SJS/TEN depends on the score.

SCORTEN predicted mortality rates

SCORTEN 0-1	>3.2%
SCORTEN 2	>12.1%
SCORTEN 3	>35.3%
SCORTEN 4	>58.3%
SCORTEN 5 or more	>90%

Treatment

- Stop suspected causative drug(s)
- Hospital admission – via Burns and Plastics to burns unit, with input from Dermatology. ITU care may be needed.
- Nutritional and fluid replacement (crystalloid) by intravenous and nasogastric routes
- Temperature maintenance – as body temperature regulation is impaired
- Pain relief – as pain can be extreme
- Sterile handling and reverse isolation procedures
- Skin care:
  - topical antiseptics e.g. silver nitrate or chlorhexidine, (but not silver sulfadiazine as it is a sulfa drug)
  - dressings such as gauze with petrolatum or non-adherent nanocrystalline-containing gauze
  - avoid using adhesive tapes
  - preferable not to remove the dead skin; leave the blister roof as a 'biological dressing'
  - daily examination and skin culture to detect bacterial infection
- Eye care:
  - daily assessment by ophthalmologist,
  - frequent eye drops/ointments (antiseptic, antibiotic, cortisone)
- Mouth care:
  - mouthwashes
  - topical oral anaesthetic
- Lung care:

        - may include aerosols, bronchial aspiration, physiotherapy
        - may require intubation and mechanical ventilation if trachea and bronchi are involved
    - Urinary catheter because of genital involvement and immobility
    - Psychiatric support for extreme anxiety and emotional lability
    - Physiotherapy to maintain joint movement and reduce risk of pneumonia
    - Regular assessment for infection including of skin, mucous membranes, catheter sites:
        - Staphylococcal infection is common; gram negative infection may also arise
        - appropriate antibiotic should be given if infection develops
        - prophylactic antibiotics are not recommended and may even increase the risk of sepsis

Adjuvant therapies including systemic corticosteroids, Ciclosporin and immunoglobulin may be considered but their benefits are currently unclear.

Complications

This condition can be fatal due to complications in the acute phase. The mortality rate is up to 10% for SJS and at least 30% for TEN.

During the acute phase, potentially fatal complications include:

- Dehydration and acute malnutrition
- Infection of skin, mucous membranes, lungs (pneumonia), septicaemia (blood poisoning)
- Shock and multiple organ failure including kidney failure
- Thromboembolism and disseminated intravascular coagulopathy

Reference:
Stevens Johnson Syndrome & Toxic Epidermal Necrolysis
www.dermnetnz.org/reactions/sjs-ten.html