



# Integumentary System - Disorders

## Stevens-Johnson Syndrome (SJS) and Toxic Epidermal Necrolysis (TEN)

### What are they?

Both SJS and TEN are severe hypersensitivity reactions of the skin. If not treated early enough mortality rates are as high as 7.5% in children and 25% in adults.

### Discerning the Difference

SJS and TEN are similar but defined by the distribution on the body:

- SJS is <10% of body surface area (BSA)
- TEN is >30% of body surface area (BSA)
- Anything in between (15-30%) is considered overlap

### Incidence

The diseases affect 1-5 people per million and incidence may be higher in bone marrow recipients, certain HIV patients, and patients with chronic rheumatologic disease.

### Causes

Drugs are responsible for 50% of SJS cases and up to 95% of TEN cases, common drugs include:

- Sulfa Drugs [Sulfamethoxazole]
- Aminopenicillins [Amoxicillin, ampicillin], Fluoroquinolones, and Cephalosporins
- Antiepileptics [Phenytoin, Phenobarbital, Valproate]
- Miscellaneous drugs [Allopurinol]

Non-drug related causes might include:

- Infection [usually Mycoplasma pneumoniae]
- Vaccination
- Graft-vs-Host Disease

### Risk Factors

While the mechanisms are unknown there has been some research to indicate that genetics may be a factor

### Signs and Symptoms

Within 1-3 weeks of taking responsible drug the patient might feel malaise (feeling of unwellness), headaches, fever, cough, and keratoconjunctivitis (inflammation of cornea and conjunctiva [clear part of eye and lining of the eyelid]).

Shortly after macules (roundish discolorations of skin that appear flat) appear suddenly, usually on face/neck/upper trunk.

These macules will appear simultaneously elsewhere on the body and turn into large fluid filled sacs (bullae) and slough (drop off) over the next 1-3 days. This process may involve the loss of nails and eyebrows.

The first skin abnormality of TEN is widespread erythema (redness).

In the most severe cases of TEN large sheets of epithelium (top layer of skin) slide off body at common pressure points known as Nikolsky's Sign. The exposed skin will be weepy and reddened. This may be followed by oral, genital, pulmonary, kidney, and liver issues.

### Diagnosis

SJS and TEN present similarly early but TEN may develop toxic shock syndrome, rash on palms and soles that develops to peeling (desquamation), widespread peeling skin (exfoliative erythroderma), and blistering mucous membranes.

### Treatment

SJS/TEN are treated as soon as possible in an ICU setting and are treated similarly to a burn.