

Toxic epidermal necrolysis (TEN) and (Stevens-Johnson syndrome (SJS))

What are TEN and SJS?

Toxic epidermal necrolysis (TEN) and Stevens-Johnson syndrome (SJS) are extremely rare, sudden onset, often life-threatening reactions to medications occurring in the skin and the linings of the mouth, the gastrointestinal tract, genitalia and eyes. The affected person has a fever and there may be enlargement of the lymph nodes and inflammation in the liver and other organ systems.

SJS is a less severe manifestation involving mainly the lips, eyes and genital mucosa. In TEN large areas of skin are killed by the immune reaction precipitated by the drug, leading to severe pain and loss of skin similar to that seen in burns. In SJS less than 10% of the skin is affected. In TEN more than 30% of the skin is involved. People with 10-30% skin loss are classified as "overlap".

What causes TEN?

TEN is more frequently seen in people who have a specific genetic make-up (genotype) that leads to slow metabolising of certain classes of drugs, or those who have HIV or are immune suppressed.

Various medications have been associated with the development of SJS and TEN. In SJS a drug can be incriminated in 50% of cases. In TEN drugs are involved in 95% of cases. They are mostly systemic drugs (oral or injection forms) but some cases have come from topical use as well.

Commonly associated medications include antibiotics (sulphonamides), antifungals, antivirals, allopurinol (gout medication), non-steroidal anti-inflammatory drugs and anticonvulsants. The onset of the adverse drug reaction usually occurs within 7 to 21 days of commencing treatment.

What does TEN look like?

Flu-like symptoms such as generalised aches and pains, sore throat and runny nose often precede the rash. TEN develops as a flat, tender, red rash that starts on the face and trunk and rapidly spreads to the rest of the body, including mucous membranes (linings of mouth, guts, wind pipes, eyes and genitals). The size of the rash expands and often forms blisters. A gentle touch or rub can peel the skin off and it is very painful. The condition often spreads over hours to days. The active phase can last up to two weeks.

How is TEN diagnosed?

There is no specific laboratory test needed to diagnose TEN or SJS. The history of the development of the rash and a physical examination usually suggest the diagnosis. A skin biopsy may be needed to confirm the diagnosis and to rule out other conditions such as staphylococcal scalded skin syndrome or other blistering diseases.

Blood tests are often required to assess the fluid/nutritional status and to identify complications.

What other problems can occur with TEN?

As the skin is the physical barrier protecting the body against infections and keeping water inside the body, the loss of the top layer of skin in TEN and SJS leads to severe problems of dehydration and increases the risk of infection. Infections are a common cause of death in those affected with TEN. In the short term, individuals can also develop dehydration, multiple organ failure and blood clots.

How is TEN treated?

The first step is to stop the medication. After this, the treatment for TEN and SJS is mainly supportive care until the top layer of skin regenerates. Affected individuals are often transferred to a burns or intensive care unit and treated as if they have suffered from very severe burns. They require life support, fluid and electrolyte replacement and antibiotics to treat infections. Usually, affected individuals are managed in a multidisciplinary team that includes a dermatologist.

Specific treatments

Prompt cessation of the medication suspected of causing the condition

Hospitalisation and admission to a specialised burns unit or intensive care unit

Fluid and electrolyte replacement

Infection control with antibiotics

Pain relief

Skin care such as the use of topical antiseptics and regular wound dressings

Special wound nursing care

Eye, mouth and lung care

Urinary catheterisation

Some reported cases have benefited from immunoglobulins, immunosuppressive agents (cyclosporin), systemic steroids or other biologic agents. However, evidence of their effectiveness is limited and controversial.

What is the likely outcome of TEN?

TEN is a life-threatening condition that is associated with high mortality rate. It is estimated to be about 5-12% for SJS and 30% for TEN. In the long term, those who survive from TEN may be left with pigmented and scarred skin, damaged nails and joints, and eye problems.

What follow up is needed for TEN?

People who develop TEN require follow up to identify the likely cause of the condition to that it can be avoided. To prevent any recurrence, affected individuals should discuss any new medications with a doctor and a pharmacist prior to commencement to check the similarity of the chemical compounds.