Indian Academy of Pediatrics (IAP)

STANDARD TREATMENT GUIDELINES 2022

Stevens–Johnson Syndrome/Toxic Epidermal Necrolysis

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Stevens–Johnson Syndrome/Toxic Epidermal Necrolysis

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Introduction

- ☑ Stevens–Johnson syndrome (SJS) is a rare and serious disorder of the skin and mucous membranes.
- ☑ This disorder affects between one and five people/million and is usually a reaction to medication that starts with flu-like symptoms, followed by a painful rash that spreads and blisters. Subsequently the epidermal layer of affected skin dies, sheds, and begins to heal after several days.
- ☑ Stevens–Johnson syndrome is a medical emergency that usually requires hospitalization.
- ☑ Treatment focuses on removing the cause, caring for wounds, hydration, controlling pain, and minimizing complications as skin regrows. It can take weeks to months to recover.
- ☑ A more severe form of the condition is called toxic epidermal necrolysis (TEN). It involves >30% of the skin surface and extensive damage to the mucous membranes.
- ☑ Stevens–Johnson syndrome and TEN are clinically similar except for their distribution. By one commonly accepted definition, changes affect <10% of body surface area in SJS and >30% of body surface area in TEN; involvement of 10–30% of body surface area is considered SJS/TEN overlap.

Symptoms

Stevens–Johnson syndrome is a rare and unpredictable illness. 1–3 days before a rash develops, patient may show early signs of SJS, including:

- ☑ Fever
- ☑ Sore mouth and throat
- ☑ Fatigue
- ☑ Burning eyes

As the condition develops, other signs and symptoms include:

- ☑ Unexplained widespread skin pain
- ☑ A red or dusky rash that spreads
- ☑ Blisters on your skin and the mucous membranes of the mouth, nose, eyes, and genitals
- ☑ Shedding of skin within days after blisters form

Etiology

Healthcare provider may not be able to identify its exact cause, but usually the condition is triggered by medication, an infection, or both. Patient may react to medication while patient is using it or up to 2 weeks after patient has stopped using it.

Drugs that can cause SJS include: Remember any drug can cause it. Common drugs are:

- ☑ Anti gout medications, such as allopurinol
- ☑ Medications to treat seizures, especially lamotrigine (all anticonvulsants and antipsychotics have got possible chances)
- ☑ Antibacterial sulfonamides (including sulfasalazine)
- ☑ Nevirapine
- ☑ Anti-tuberculous drug (ATT)
- ☑ Antibiotics
- ☑ Acetaminophen, ibuprofen, and naproxen sodium
- ☑ Infections that can cause SJS include pneumonia and human immunodeficiency virus (HIV), *Mycoplasma*, Epstein–Barr virus (EBV), and herpes simplex

Risk Factors

Risk factors for SJS include:

- ☑ HIV infection (among people with HIV, the incidence of SJS is about 100 times greater than among the general population)
- ☑ *Immunodeficiency state:* Postorgan transplant, *Pneumocystis jirovecii*-infected HIV patients, systemic lupus erythematosus, and in patients with other chronic rheumatologic diseases and autoimmune diseases
- ☑ *Malignancy:* Especially related to blood cancer
- ☑ Prior history of SJS, if it is medication-related they are at risk of a recurrence, if they resist that drug
- ☑ A family history of SJS, especially, if an immediate blood relative has had it
- ☑ *Genetic factors:* Having certain genetic variation like HLA-B*1502 puts them at increased risk

Complications

Stevens–Johnson syndrome complications include:

- ☑ *Dehydration:* Fluid is lost from areas of skin shedding; and sores in the mouth and throat can make fluid intake difficult, resulting in dehydration.
- ☑ *Blood infection (sepsis):* Sepsis occurs when bacteria from an infection enters the bloodstream and spreads throughout the body. Sepsis is a rapidly progressing and life-threatening condition that can cause shock and organ failure.
- ☑ *Eye problems:* The rash caused by SJS can lead to eye inflammation, dry eye, and light sensitivity. In severe cases, it can lead to visual impairment and, rarely, blindness. So, early evaluation with ophthalmologist is must. Eye lubricating agents should start early.
- ☑ *Lung involvement:* Acute respiratory failure
- ☑ *Permanent skin damage:* When your skin grows back following SJS, it may have dyspigmentation, scars, alopecia, and growth issue with fingernails and toenails.

Prevention

- ☑ Avoid the incriminating drug.
- ☑ Genetic testing (pharmacogenomics) before taking certain drugs, it may be important but not readily available. The US Food and Drug Administration approved screening for people of Asian and South Asian ancestry for a gene variation called HLA-B*1502 before starting treatment.
- ☑ Avoid precipitating medication if such generic variation is found.
- ☑ If someone had SJS prior and it was caused by a medication, best prevention is to avoid that drug. This is key to preventing a recurrence, which is usually more severe than the first episode and can be fatal.

Treatment

- ☑ Investigations such as complete blood count (CBC), renal function tests, hepatic function tests, and chest X-rays to be done along with skin biopsy.
- ☑ It is supportive care with care of skin with dressings, eye care, bland diet, monitoring vitals, and fluid intake; cyclosporine, intravenous (IV) immune globulin, early corticosteroid therapy in early phase judiciously tapered over 3–7 days, and tumor necrosis factor-alpha inhibitors have been used, only after dermatologist being consulted.
- ☑ Mortality can be as high as 7.5% in children and 20–25% in adults but tends to be lower with early treatment.

Further Reading

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