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Fatal Skin Disorder - Stevens Johnson Syndrome

T. Merlin Shiba
Assistant Professor, Sree Balaji College of Nursing, Bharath University, Tamil Nadu, India
V. Hemavathy
Principal, Sree Balaji College of Nursing, Bharath University, Tamil Nadu, India

Abstract:
Toxic epidermal necrolysis (TEN) and Stevens-Johnson syndrome (SJS) are rare (one to two per 10, 00, 00 population per year) but life threatening adverse drug reactions. Drugs commonly implicated are anti-epileptics, anti-microbial and non-steroidal anti-inflammatory drugs (NSAIDS). Amongst anti-epileptics, carbamazepine and phenytoin are the major culprits.
Stevens- Johnson syndrome or toxic epidermal necrolysis are potentially fatal skin disorders and the most severe forms of erythema multiforme. The diseases are mucocutaneous reactions that constitute a spectrum of reactions, with TEN being the most severe. The mortality rate from TEN is 30% to 35%. TEN and SJS are triggered by a reaction to medication. Antibiotics, specially sulfonamides, anti seizure agents, non- steroidal, anti- inflammatory drugs, and sulfonamides are the most frequent medications implicated. Most patients with TEN have an abnormal metabolism of the medication; the mechanism leading to TEN seems to be a cell- mediated cyto-toxic reaction.

Keywords: *Toxic epidermal necrolysis (TEN), Stevens-Johnson syndrome (SJS), non-steroidal anti-inflammatory drugs, severe cutaneous reactions (SCAR)*

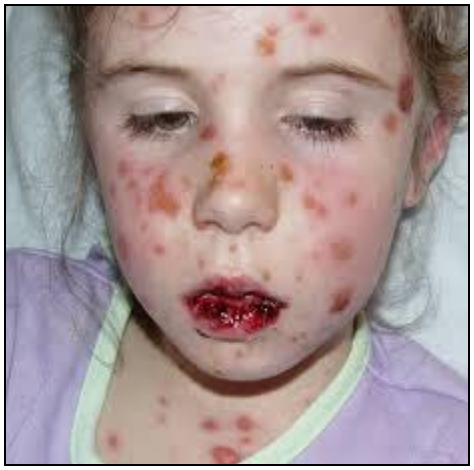


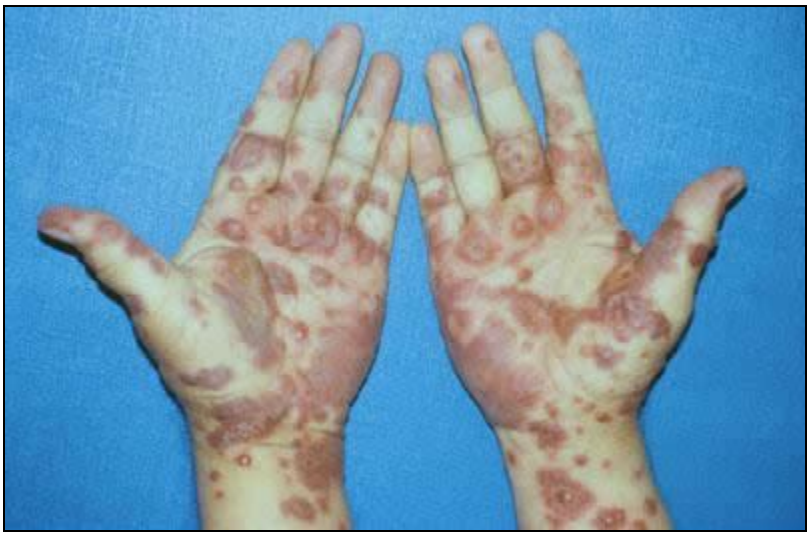
Figure 1

Stevens–Johnson syndrome (SJS) is a milder form of toxic epidermal necrolysis (TEN).[1] These conditions were first recognized in 1922. A classification first published in 1993 that has been adopted as a consensus definition identifies Stevens–Johnson syndrome, toxic epidermal necrolysis, and SJS/TEN overlap. All three are part of a spectrum of severe cutaneous reactions (SCAR) which affect skin and mucous membranes.[3] The distinction between SJS, SJS/TEN overlap, and TEN is based on the type of lesions and the amount of the body surface area with blisters and erosions. Blisters and erosions cover between 3% and 10% of the body in SJS, 11-30% in SJS/TEN overlap, and over 30% in TEN. The skin pattern most commonly associated with SJS is widespread, often joined or touching (confluent), papuric spots (macules) or flat vesicles or bullae which may also be confluent. These occur primarily on the torso**.**

1. Causes

stevens-Johnson syndrome is a rare and unpredictable reaction. Your doctor may not be able to identify its exact cause, but usually the condition is triggered by a medication or an infection.

2. Medication and Therapy Causes



Drugs that can cause Stevens-Johnson syndrome include:

1. Anti-gout medications, such as allopurinol
2. Pain relievers such as acetaminophen (Tylenol, others), ibuprofen (Advil, Motrin IB, others) and naproxen sodium (Aleve)
3. Medications to fight infection, such as penicillin
4. Medications to treat seizures or mental illness (anticonvulsants and antipsychotics)
5. Radiation therapy

3. Infectious Causes

Infections that can cause Stevens-Johnson syndrome include:

1. Herpes (herpes simplex or herpes zoster)
2. Pneumonia
3. HIV
4. Hepatitis

4. Risk Factors

Factors that increase your risk of developing Stevens-Johnson syndrome include:

- Viral infections. Your risk of Stevens-Johnson syndrome may be increased if you have an infection caused by a virus, such as herpes, viral pneumonia, HIV or hepatitis.
- Weakened immune system. If you have a weakened immune system, you may have an increased risk of Stevens-Johnson syndrome. Your immune system can be affected by an organ transplant, HIV/AIDS and autoimmune diseases, such as lupus.
- A history of Stevens-Johnson syndrome. If you've had a medication-related form of this condition, you are at risk of a recurrence if you use that drug again.
- A family history of Stevens-Johnson syndrome. If an immediate family member has had Stevens-Johnson syndrome or a related condition called toxic epidermal necrolysis, you may be more susceptible to developing Stevens-Johnson syndrome too.
- Having a certain gene. If you have a gene called HLA-B 1502, you have an increased risk of Stevens-Johnson syndrome, particularly if you take certain drugs for seizures or mental illness. Families of Chinese, Southeast Asian or Indian descent are more likely to carry this gene.

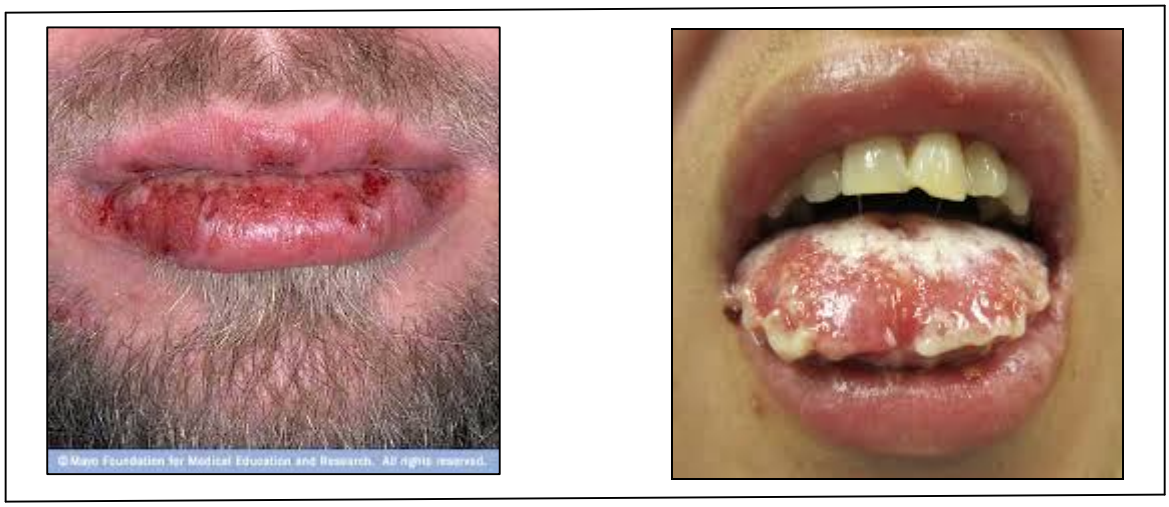


Figure 2: Stevens - Johnson syndrome Symptoms

- Facial swelling.
- Tongue swelling.
- Hives.
- Skin pain.
- A red or purple skin rash that spreads within hours to days.
- Blisters on your skin and the mucous membranes of your mouth, nose, eyes and genitals.
- Shedding of your skin.

4.1. Complications

- Secondary skin infection (cellulitis). Cellulitis can lead to life-threatening complications, including sepsis.
- Blood infection (sepsis). Sepsis occurs when bacteria from an infection enter your bloodstream and spread throughout your body. Sepsis is a rapidly progressing, life-threatening condition that can cause shock and organ failure.
- Eye problems. The rash caused by Stevens-Johnson syndrome can lead to inflammation in your eyes. In mild cases, this may cause irritation and dry eyes. In severe cases, it can lead to extensive tissue damage and scarring that results in blindness.

Figure 3

- Keratoconjunctivis can impair vision and result in conjunctival reaction, scarring and corneal lesion.
- Damage to internal organs. It's unusual for this condition to affect internal organs. But it may cause inflammation of the lungs, heart, kidneys or liver.

5. Tests and Diagnosis

- Physical exam. Doctors often can identify Stevens-Johnson syndrome based on your medical history, a physical exam, and the disorder's signs and symptoms.
- Skin test. To confirm the diagnosis, your doctor may remove a sample of skin for laboratory testing (biopsy).

6. Medical Management

6.1. Stopping nonessential medications

The first and most important step in treating Stevens-Johnson syndrome is to discontinue any medications that may be causing it. Because it's difficult to determine exactly which drug may be causing the problem, your doctor may recommend that you stop taking all nonessential medications.

6.2. Supportive care

Supportive care you're likely to receive while hospitalized includes:

- Fluid replacement and nutrition. Because skin loss can result in significant loss of fluid from your body, replacing fluids is an important part of treatment. You may receive fluids and nutrients through a tube placed through your nose and advanced into your stomach (nasogastric tube).
- Wound care. Cool, wet compresses will help soothe blisters while they heal. Your health care team may gently remove any dead skin and place a medicated dressing over the affected areas.
- Eye care. You may also see an eye specialist (ophthalmologist).

7. Medications

Medications commonly used in the treatment of Stevens-Johnson syndrome include:

- Pain medication to reduce discomfort
- Medication to relieve itching (antihistamines)
- Antibiotics to control infection, when needed
- Medication to reduce skin inflammation (topical steroids)

If the underlying cause of Stevens-Johnson syndrome can be eliminated and the skin reaction stopped, new skin may begin to grow over the affected area within several days. In severe cases, full recovery may take several months.

8. Nursing Management

1. A careful inspection of the skin is made, including its appearance and the extent of involvement.
2. Oral cavity is inspected daily for blistering and erosive lesions.
3. The patient is assessed daily for itching, burning and dryness of eyes.
4. The patient's ability to swallow and drink fluids, as well as speak normally is determined.
5. The patients vital signs are monitored, special attention is given to the presence and character of fever , respiratory rate, rhythm and cough.
6. Assessment for high fever, tachycardia, extreme weakness and fatigue is essential because theses indicate epidermal necrosis
7. Urine volume, specific gravity, and colour are monitored.
8. The insertion sites of IV lines sites are inspected for signs of local infection.
9. Body weight is recorded daily.

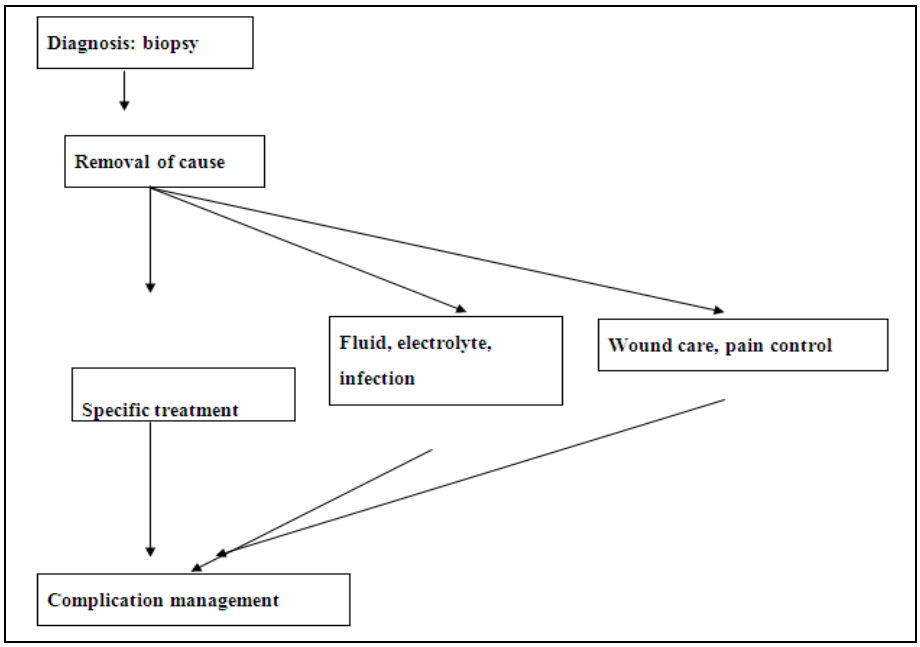


Figure 4: Management of SJS

9. Prevention

1. If you've had this condition, avoid the medication that triggered it. If you've had Stevens-Johnson syndrome and your doctor told you it was caused by a medication, avoid that drug and others like it. This is key to preventing a recurrence, which is usually more severe than the first episode and can be fatal.
2. Your family members also might want to avoid this drug because some forms of this condition have a genetic risk factor
3. Wear a medical information bracelet or necklace. Have information about your condition and what caused it inscribed on a medical information bracelet or necklace. Always wear it.
4. Inform your health care providers. Tell all your health care providers that you have a history of Stevens-Johnson syndrome. If the reaction was caused by a medication, tell them which one.

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