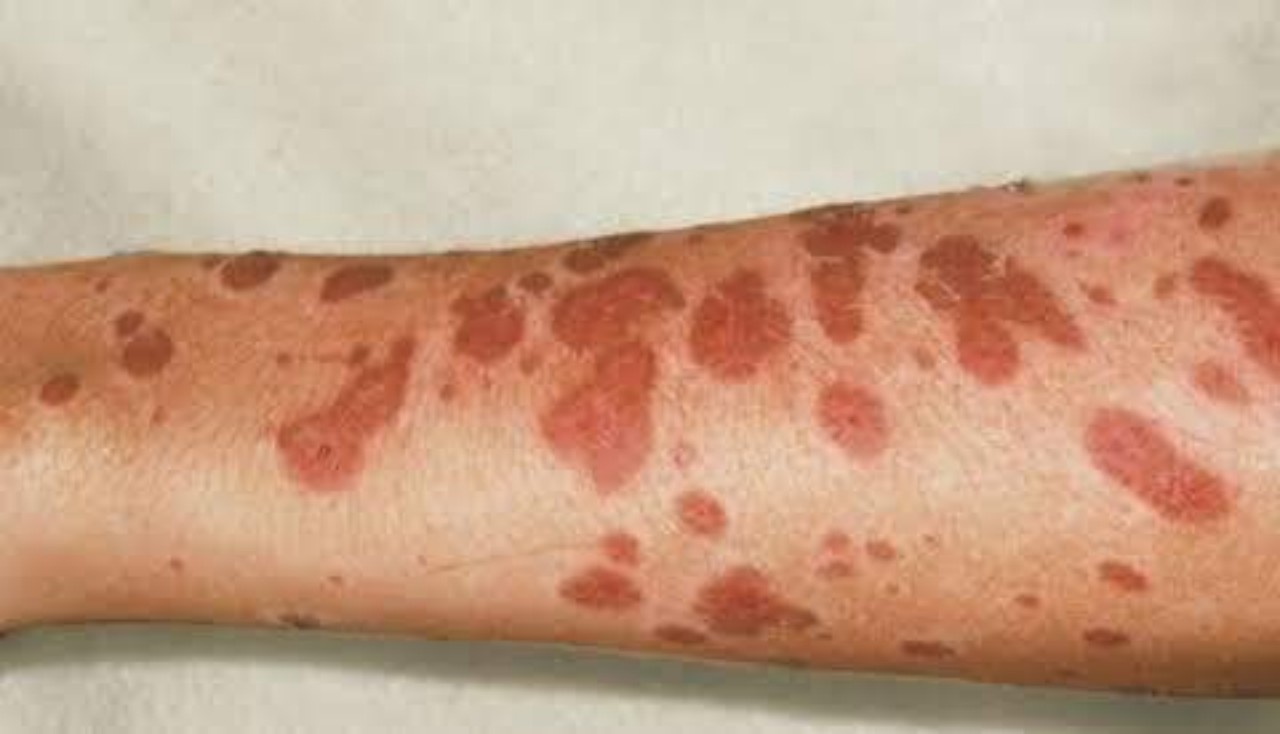

STEVENS-JOHNSON SYNDROME

- Stevens-Johnson syndrome (SJS) is an immune-complex–mediated hypersensitivity complex that typically involves the skin and the mucous membranes. While minor presentations may occur, significant involvement of oral, nasal, eye, vaginal, urethral, gastrointestinal, and lower respiratory tract mucous membranes may develop in the course of the illness. GI and respiratory involvement may progress to necrosis. Stevens-Johnson syndrome is a serious systemic disorder with the potential for severe morbidity and even death.

STEVENS-JOHNSON SYNDROME

- The syndrome was first described in 1922, when the American pediatricians Albert Mason Stevens and Frank Chambliss Johnson reported the cases of 2 boys aged 7 and 8 years with "an extraordinary, generalized eruption with continued fever, inflamed buccal mucosa, and severe purulent conjunctivitis." Both cases had been misdiagnosed by primary care physicians as hemorrhagic measles.

STEVENS-JOHNSON SYNDROME

- Although several classification schemes have been reported, the simplest breaks the disease down as follows:
- Stevens-Johnson syndrome - A "minor form of TEN," with less than 10% body surface area (BSA) detachment
- Overlapping Stevens-Johnson syndrome/toxic epidermal necrolysis (SJS/TEN) - Detachment of 10-30% BSA
- Toxic epidermal necrolysis - Detachment of more than 30% BSA

EPIDEMIOLOGY

- The incidence rate is 7 cases per million population per year.
- Cases tend to have a propensity for the early spring and winter.
- Stevens-Johnson syndrome has been described worldwide in all races, although it may be more common in whites. Interestingly, disease is not limited to humans; cases have been reported in dogs, cats, and monkeys.

12.Stevens - Johnson syndrome

- Severe form of Erythema multiforme
- Minor form of Toxic Epidermal Necrolysis involving < 10 % of body surface area
- Muco-cutaneous, immune-complex-mediated hypersensitivity disorder causing separation of epidermis from dermis

ETIOLOGY

- Idiopathic: 25 - 50 % cases
- Drug reaction: Penicillin, Sulfonamides, Macrolide, Ciprofloxacin, Phenytoin, Carbamazepine, Valproate, Lamotrigine, NSAIDs, Valdecoxib, Allopurinol
- Viral infection: herpes simplex, HIV, influenza
- Malignancy: carcinoma, lymphoma

GENETIC FACTORS

- There is strong evidence for a genetic predisposition to severe cutaneous adverse drug reactions such as Stevens-Johnson syndrome. Carriage of the certain human leukocyte antigens has been associated with increased risk.

INFECTIOUS CAUSES

- Viral diseases that have been reported to cause Stevens-Johnson syndrome include the following:
- Herpes simplex virus (possibly; remains a debated issue)
- AIDS
- Coxsackie viral infections
- Influenza
- Hepatitis
- Mumps

INFECTIOUS CAUSES

- Bacterial etiologies include the following:
- Group A beta-hemolytic streptococci
- Diphtheria
- Brucellosis
- Lymphogranuloma venereum
- Mycobacteria
- *Mycoplasma pneumoniae*
- Rickettsial infections
- Tularemia
- Typhoid

TABLE 1

Common Medication Causes of SJS/TEN

Sulfonamides	Trimethroprim-sulfamethoxazole Sulfamethoxazole eye drops Sulfapyridine Sulfadoxine Silver sulfadiazine
Other antibiotics	Penicillins Cephalosporins Minocycline Ciprofloxacin Terbinafine
Anticonvulsants	Phenobarbital Carbamazepim Phenytoin Lamotragine
Other medications	Allopurinol Nevirapine Non-steroidal Anti-inflammatory agents (NSAIDs) Sertraline

Pathophysiology

- *A delayed hypersensitivity reaction has been implicated in the pathophysiology of Stevens-Johnson syndrome.*
- *Antigen presentation and production of tumor necrosis factor (TNF)–alpha by the local tissue dendrocytes*
- *results in the recruitment and augmentation of T-lymphocyte proliferation and enhances the cytotoxicity of the other immune effector cells.*

Signs and Symptoms

- Facial swelling
- Tongue swelling
- Hives
- Skin pain
- A red or purple skin rash that spreads within hours to days
- Blisters on skin and mucous membranes
- Shedding (sloughing) of skin

CLINICAL PRESENTATION

- Typically, Stevens-Johnson syndrome (SJS) begins with a nonspecific upper respiratory tract infection. This usually is part of a 1- to 14-day prodrome during which fever, sore throat, chills, headache, and malaise may be present. Vomiting and diarrhea are occasionally noted as part of the prodrome.
- Mucocutaneous lesions develop abruptly. Clusters of outbreaks last from 2-4 weeks. The lesions are typically nonpruritic.

WORKUP

There are no specific laboratory studies (other than biopsy) that can definitively establish the diagnosis of Stevens-Johnson syndrome.

- Serum levels of the following are typically elevated in patients with Stevens-Johnson syndrome:
- Tumor necrosis factor (TNF)-alpha
- Soluble interleukin 2-receptor
- Interleukin 6
- C-reactive protein
- However, none of these serologic tests is used routinely in diagnosing and managing Stevens-Johnson syndrome.

TREATMENT & MANAGEMENT

- Management of patients with Stevens-Johnson syndrome is usually provided in intensive care units or burn centers. No specific treatment of Stevens-Johnson syndrome is noted; therefore, most patients are treated symptomatically. In principle, the symptomatic treatment of patients with Stevens-Johnson syndrome does not differ from the treatment of patients with extensive burns.

TREATMENT & MANAGEMENT

- Identify and stop the offending agent.
- Isolate the patient.
- Give IV fluids.
- Give corticosteroids.
- IVIG if available.
- Antibiotic to treat secondary infection.
- Use minimum drugs for its treatment.

PROGNOSIS

Survivors of Stevens-Johnson syndrome may experience numerous long-term sequelae; the most disabling are those of the eye. Cicatrization of conjunctival erosions may lead to the following:

- Inverted eyelashes
- Photophobia
- A burning sensation in the eyes
- Watery eyes
- A siccalike syndrome
- Corneal and conjunctival neovascularization

COMPLICATIONS

Of patients with Stevens-Johnson syndrome, 27-50% progress to severe ocular disease. Ocular complications of Stevens-Johnson syndrome include the following:

- Chronic cicatrizing conjunctivitis
- Corneal epithelial defects
- Corneal stromal ulcers
- Corneal perforation
- Endophthalmitis