
Staphylococcal
Scalded Skin
Syndrome

Staphylococcal Scalded Skin Syndrome

A spectrum of superficial blistering skin disorders caused by the
. exfoliative toxins of some strains of *Staphylococcus aureus*

It is a syndrome of acute exfoliation of the skin typically following an
.erythematous cellulitis
Severity of SSSS varies from a few blisters localized to the site of
.infection to a severe exfoliation affecting almost the entire body

Staphylococcal Scalded Skin Syndrome

- Certain strains of *S. aureus* secrete **exfoliative toxins (toxemia)** that cause dissolution of epidermal desmosomes
- Cause **staphylococcal scalded skin syndrome (SSSS),** in which the epidermis peels off in sheets
- **Person-to-person spread**
  - The bacterium penetrates cuts and abrasions
- **Susceptible patients**
  - Infants, elderly, immunosuppressed patients

Fluid in blisters does not contain S. aureus

Staphylcoccal Scalded Skin Syndrome (SSSS) *(Cont'd)*

- Usually under age 5
- Staph toxin, with scarlatiniform rash after onset of fever, irritability and then exfoliation
- Positive Nikolsky's sign
- Treated with antibiotics (beta-lactamase resistant) but this does not change the skin effects
- Better prognosis than toxic epidermal necrolysis (TEN)

Epidemiology

- predominantly in infants and children younger than 5 years of age and rarely occurs in adults
- Due to circulating antibodies and renal excretion of toxins
- most cases are caused by type 71 strain (75%)
- no differences in incidence based on gender nor economic status

Nelson's Textbook of Pediatrics. 19th Edition
Schwartz, M. William. *The 5 minute pediatric consult*. 2nd ed.

Exfoliative toxins

- Staphylococcal scalded skin syndrome (SSSS), a spectrum of diseases characterized by exfoliative dermatitis, is mediated by exfoliative toxins.
- The prevalence of toxin production in *S. aureus* strains varies geographically but is generally less than 5% to 10%.

Etiology

- caused predominantly by phage group 2 staphylococci, particularly strains 71 and 55
- found in nasopharynx and, less commonly, the umbilicus, urinary tract, a superficial abrasion, conjunctivae, and blood
- spreads hematogenously

Nelson's Textbook of Pediatrics. 19th Edition

Pathophysiology:

Predisposing factors

- Staphylococcus aureus strains produce an exfoliative toxin (ETA, ETB).
- ETs target the cell adhesion protein **desmoglein 1** (DG1) resulting in separation of keratinocytes just beneath the granular layer in the epidermis (intraepidermal).
- In bullous impetigo, the ETs remain local in the infected skin but in SSSS the ETs spread haematogenously resulting in widespread skin involvement.

Staphylococcal Scalded Skin Syndrome - Clinical Features

- Starts Abruptly
  - Perioral erythema
  - Sunburn like, tender rash → spreads over entire body
- Bullae Appear Rapidly
  - Nikolsky sign
  - Flaccid bullae slough off → Denuded areas
- Exfoliated Areas Eventually Dry
  - Flaky desquamation lasting 3-5 days
- Within 10 days After Onset →Complete Recovery
  - New epidermis has replaced the denuded areas

Clinical variants

- Localized SSSS:
  - Favours the flexures, especially axillae, groin and limb flexures.
  - Healing of localized form of disease leaves wrinkled desquamating skin with hyperpigmentation.
  - Localized form is considered to be less dangerous than the generalized form of the disease.

Staphylococcal scalded skin syndrome in an adult.

Localized staphyloccocal scalded skin syndrome (SSSS) which subsequently healed with wrinkling desquamation and hyperpigmentation.

DIAGNOSIS

- Gram stain or Culture: from the remote site of infection
- Skin biopsy or Frozen Section
- PCR

Treatment

- Proper hygiene
  - This condition is primarily treated with advocating cleaning of the affected area regularly
  - Warm water and mild shampoo for eyelashes
- Antibiotics
- Steroid Eyedrops
- Artificial Tears

Review of Medication:

- Hydroxyzine 2mg/ml, 2.5 ml every 6 hours PRN for pruritus
- Mupirocin ointment, apply over nasal mucosa using cotton buds, 3x a day for 7 days
- Erythromycin eye oitment, 1 strip to both lower lids 2x a day
- Cloxacillin 250mg/ml, 2ml every 6 hours on an empty stomach, 1 hour prior to meals

Staphylococcal Scalded Skin Syndrome (SSSS)

- Treatment, prevention, and control
  - isolation and identification based on catalase test, coagulase test, serology, DNA fingerprinting, and phage typing
  - antibiotic therapy
    - many drug-resistant strains
  - personal hygiene, food handling, and aseptic management of lesions

Prognosis

- Recovery is usually rapid, but complications such as excessive fluid loss, electrolyte imbalance, faulty temperature regulation, pneumonia, septicemia, and cellulitis may cause increased morbidity.

Toxic Epidermal Necrolysis

What is T.E.N.?

- Definition:
  - **Toxic** = "pertaining to, due to or the nature of a poison or toxin, manifesting the symptoms of severe infection"
  - **Epidermal** = "pertaining to or resembling the epidermis"
  - **Necrolysis** = "separation or exfoliation of tissue due to necrosis"

- Rare
- Life threatening
- Drug induced
  - adverse drug reaction: 5-15% drug treatments
  - cutaneous reactions: most common
    - morbilliform type: most common
      - symmetrical, erythematous rash, macules & papules, lasts few days
    - can proceed to serious cutaneous reactions:
      - serum sickness
      - hypersensitivity syndrome
      - **T.E.N.**

DEFINITION

- Definition:
  - **Toxic epidermal necrolysis** (**TEN**), also known as **Lyell's syndrome**, is a rare, life-threatening skin condition that is usually caused by a reaction to drugs. The disease causes the top layer of skin (the epidermis) to detach from the lower layers of the skin (the dermis), all over the body, leaving the body susceptible to severe infection.

- morbilliform type: most common
  - symmetrical, erythematous rash, macules & papules, lasts few days

- can proceed to serious cutaneous reactions:
  - serum sickness
  - hypersensitivity syndrome
  - **T.E.N**

Toxic Epidermal Necrolysis (TEN)

- TEN is a rare, potentially fatal, adverse drug reaction characterized by tenderness and erythema of the skin and mucosa, and extensive mucocutaneous exfoliation.
  - This exfoliation is due to extensive death of keratinocytes via apoptosis.
- Incidence: 0.4 to 1.2 cases per million per year.

Causes

- Adverse drug reaction
  - over 100 drugs implicated

Sulphonamide antibiotics
Anticonvulsants
NSAIDs
Allopurinol
Corticosteroids

Newest Causative Drugs
Nevirapine (antiretroviral)
Lamatrogine

- Other causes:
  - immunisations; bone marrow transplants; solid organ transplants

Clinical Features

- Prodromal phase; **1 – 14 days**
  - flu-like symptoms
- Inflammation:
  - eyelids
  - conjunctiva
- Tenderness:
  - oral mucosa
  - general cutaneous

- Generalised macular erythma:
  - progresses to flaccid blisters and bullae
  - join to form large bullae
  - large areas of epidermis are "sloughed off"
- Mucous membranes often involved
  - usually 1-3 days earlier then skin lesions
  - eyes; oropharynx; respiratory tract; GI tract; genital tract; anus

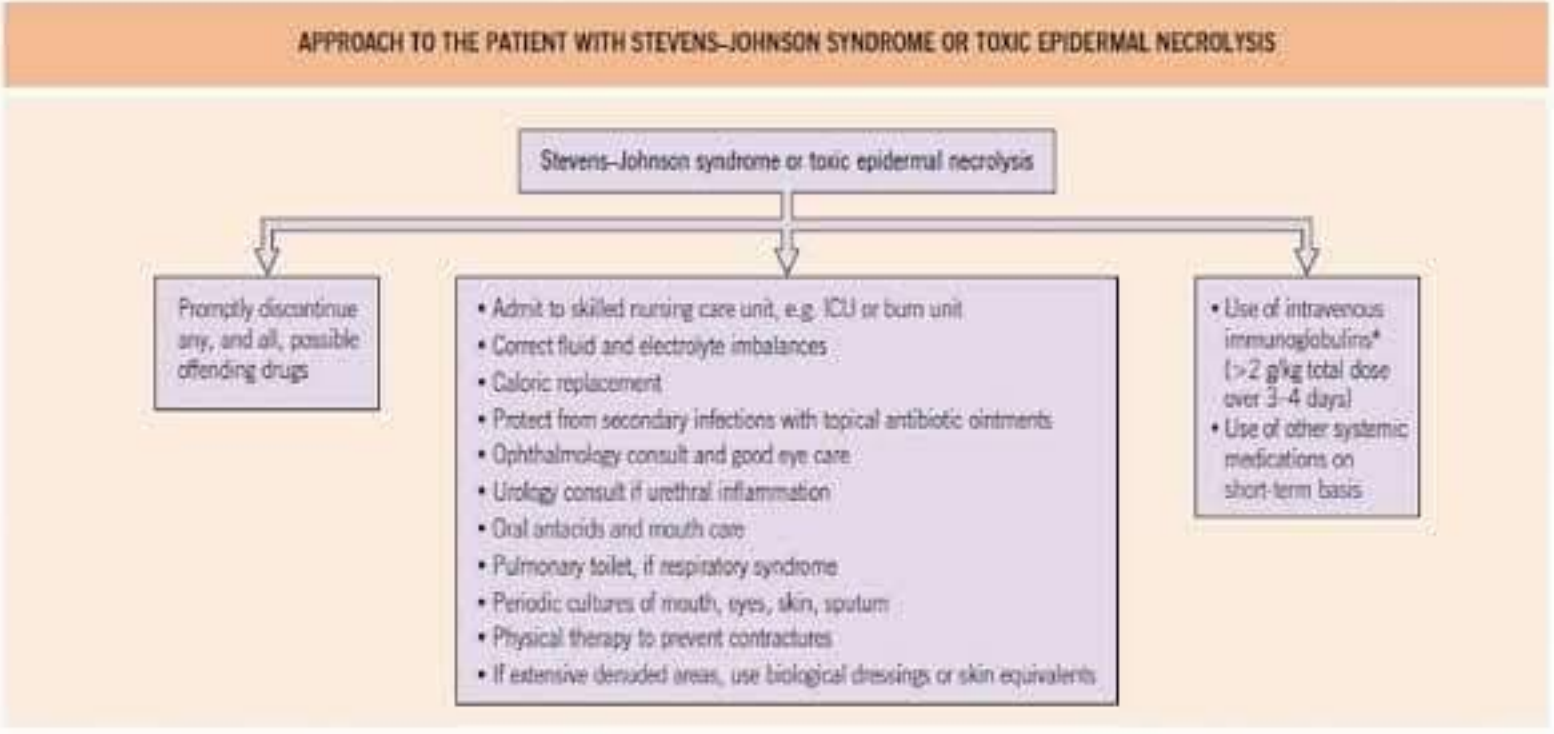
Rapid progression over days. 10 – 100% of body's surface area involved

Investigations

- MICROBIOLOGY
  - blood culture
  - MSU
  - swabs; including MRSA screening swabs
- GENERAL
  - FBC
  - ESR
  - U&E and creatinine
  - LFTs
  - Albumin
  - Glucose
  - Calcium
  - CRP
  - Urine dipstick (protein & blood)
- IF INDICATED
  - coagulation studies
  - CXR

Treatment

- Optimal medical management of SJS and TEN requires early diagnosis, immediate discontinuation of the causative drug(s), supportive care, and specific therapy.



Treatment

- Optimal medical management requires early diagnosis, immediate discontinuation of the causative drug(s), and supportive care.
- Careful daily wound care, hydration, and nutritional support are essential, and preferably, done in an ICU.
- No specific treatments for TEN have met evidence-based medicine standards of acceptance.
- Cyclosporine, cyclophosphamide, plasmapheresis, and N-acetylcysteine have shown promising results.
- The use of corticosteroids is controversial and they may even increase mortality.