**Stevens-Johnson Syndrome and Toxic Epidermal Necrolysis in Adults: Improving Your Diagnostic and Management Skills**

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**Epidemiology**

- **Incidence:** 2-13 cases per million people per year\(^1\)\(^3\)
- **Mortality:** 15 – 23%\(^3\)\(^4\)
- **US Multi-institution Cohort\(^3\), N = 377**
  - 52% female
  - Mean (SD) age: 49.0 (19.2) years
  - Median BSA on admission: 15% (6-30%)
  - Mortality: 14.7%

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Causative Agents

- Allopurinol (Europe\(^3\))
- Anticonvulsants
- NSAIDs
- Nevirapine (sub-Saharan Africa\(^3\))
- Sulfa antibiotics (US\(^2\))


Pathogenesis

- Delayed immunologically-mediated drug reaction
- Antigen-mediated or direct T-cell stimulation
- Activation of cytotoxic T cells and NK cells \(\rightarrow\) cytokines
- Keratinocyte apoptosis

Clinical Presentation

- 1-3 day flu-like prodrome
- Dusky atypical targetoid lesions → epidermal detachment/bullae
- Classification:
  - SJS: < 10% epidermal detachment
  - SJS/TEN overlap: 10-30%
  - TEN: > 30%
- 2 or more mucous membranes

Differential diagnosis

- Erythema multiforme
- *Mycoplasma*-induced rash and mucocysitis
- Staph Scalded Skin Syndrome
- Generalized Bullous Fixed Drug Eruption
- Severe Bullous Lupus (Rowell’s Syndrome)
- Autoimmune blistering diseases
- Severe Graft vs Host disease
Diagnosis

- Stop any possible causative agent
- Biopsy (+/- DIF)
  - Frozen section biopsy or jelly roll
  - Consider any labs to rule out mimickers

Supportive Care

- Multi-disciplinary care at a specialized center
  - Standard Ophthalmology and Gynecology consults
- Careful management of fluid balance, electrolyte disturbances, infection surveillance and pain control
- Consider early airway protection
- Supplemental nutrition
Skin & Wound Care

- Wound care varies by institution and provider
- Goals:
  - Minimize shearing forces
  - Maintain thermoregulation / minimize insensible losses
  - Prevent secondary infection
  - Minimize scarring

Infection surveillance

- Empiric antibiotics are NOT recommended.
- Careful monitoring for signs of infection
- Low threshold for cultures
- Most common organisms:
  - Initially → Staph aureus
  - Prolonged hospitalization → pseudomonas
Eye Care

- Ophthalmology eval within 24hrs
  - Standardize grading
  - Daily eye exam
- Treatment based on severity
  - None: artificial tears
  - Mild – Mod: moxifloxacin, cyclosporine, dexamethasone & tobramycin/dexamethasone ointment
  - Severe: Topicals + amniotic membrane transplantation (AMT).
- AAO Treatment Guidelines:

Gynecology Care

- Prompt evaluation of ALL female patients
- Treatments:
  - Intravaginal steroid ointments (Class III)
  - Dilatory therapy to prevent adhesions
  - Menstrual suppression to prevent adenosis (metaplastic endometrial epithelium in the vaginal wall)¹
- Up to 30% of women with vaginal involvement suffer from long-term sequelae.²

Pharmacotherapies

- No clear beneficial treatment for ALL patients
- Treat should be individualized based on the patient and clinical scenario

Systemic Steroids

- First recognized treatment
- Subsequent small case series showed increased complications, higher mortality\(^1,2\)
- A meta-analysis found survival benefit vs supportive care alone\(^3\)
- Shorter duration (5-7 days) is probably better.

### IVIg

- Initial studies showed a benefit\(^1\)
- Beneficial results have not been replicated
- Reduced “recovery time” seen in IVIg + steroids vs steroids alone
- No mortality benefit: IVIg vs supportive care \(^2,3\)


### Cyclosporine

- Several uncontrolled studies\(^2-3\) & meta-analyses\(^4-5\) have shown a mortality benefit compared to SCORTEN-predicted mortality
- Patients treated with cyclosporine are typically younger and healthier.

**TNF Inhibitors**

- **Thalidomide**
  - associated with excess mortality\(^1\)
- **RCT: etanercept 2x/week vs 1-1.5mg/kg prednisolone\(^2\)**
  - Primary Outcome:
    - Healing time
    - No difference overall
  - Sub-analysis: Improved healing time in BSA \(>10\%\) with etanercept (14 vs 19 days)


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**Take Home Points**

- Dermatologists play a critical role in making the correct diagnosis and coordinating care.

- In addition to multi-disciplinary supportive care, pharmacotherapy should be based on the individual patient.