Steven Johnson Syndrome (SJS) and Toxic Epidermal Necrolysis (TEN) are rare, acute, life-threatening dermatological diseases characterized by diffuse superficial sloughing of the epidermis (<20% sloughing in patients with SJS, and >40% sloughing in patients with TEN). Approximately 95% of TEN cases and 50% of SJS cases are reported to be drug related.

Key Principles

- Ensure prompt cessation of any of the following suspicious medications, especially antibiotics or anti-convulsants:

<table>
<thead>
<tr>
<th>Medication</th>
<th>Substitute</th>
</tr>
</thead>
<tbody>
<tr>
<td>Allopurinol</td>
<td>Minocycline</td>
</tr>
<tr>
<td>Carbamazepine</td>
<td>Nevirapine</td>
</tr>
<tr>
<td>Fluoroquinolones</td>
<td>Non-Steroidal: anti-inflammatory drugs</td>
</tr>
<tr>
<td>Lamotrigine</td>
<td>Phenobarbital</td>
</tr>
<tr>
<td>Phenytoin</td>
<td>Sulfaalazine</td>
</tr>
<tr>
<td>Trimethoprim-sulfamethoxazole</td>
<td></td>
</tr>
</tbody>
</table>

- Transfer to a Hospital with a verified Burn Center and obtain early consultation with below:
  - Consult Burn Center: 293-BURN (293-2876)
  - Consult Dermatology to rule out other blistering disease
  - Consult Ophthalmology when diagnosis has been confirmed if concern for ocular involvement

- Treatment is primarily supportive.
- Protect skin while it heals, with special emphasis on care of eyes, oral mucosa, gastrointestinal, and respiratory epithelia.

Diagnosis

- Perform full-thickness punch biopsy, from a border of intact epidermis surrounding bullous lesions.
- Obtain thorough history including medication to determine possible cause.

Prognosis: Stratify Severity of Illness and Predict Mortality

Use the SCORTEN Scoring System within 24 hours of admission and daily for the first 5 days of hospitalization.

<table>
<thead>
<tr>
<th>SCORTEN Variables</th>
<th>Prognostic Factors</th>
<th>Values</th>
<th>Weight</th>
</tr>
</thead>
<tbody>
<tr>
<td>Age</td>
<td>&gt; 40 years of age</td>
<td>1</td>
<td></td>
</tr>
<tr>
<td>Malignancy</td>
<td>YES</td>
<td>1</td>
<td></td>
</tr>
<tr>
<td>Body surface area detached</td>
<td>&gt; 10%</td>
<td>1</td>
<td></td>
</tr>
<tr>
<td>Heart rate</td>
<td>&gt; 120 bpm</td>
<td>1</td>
<td></td>
</tr>
<tr>
<td>BUN</td>
<td>&gt; 10 mmol/L or &gt; 28 mg/dl</td>
<td>1</td>
<td></td>
</tr>
<tr>
<td>Serum glucose</td>
<td>&gt; 14 mmol/L or &gt; 250 mg/dl</td>
<td>1</td>
<td></td>
</tr>
<tr>
<td>Serum bicarbonate</td>
<td>&lt; 20 mmol/L or mEq/dl</td>
<td>1</td>
<td></td>
</tr>
</tbody>
</table>

Probability of Death:

- 0 points = 3%  
- 1 points = 3%  
- 2 points = 12%  
- 3 points = 25%

Discontinue the likely offending pharmacological agent.

Etiology Assessment

- Algorithm of Drug causality for Epidermal Necrolysis (ALDEN) criteria
- Naranjo Criteria
- Labs:
  - Mycoplasma pneumoniae serologies
  - Complete Blood counts
  - History driven testing (HSV, other infections).

Fluid Resuscitation

Epidermal Loss 15-30% Total Body Surface Area

Initial Fluids – Parkland Formula

- Lactated Ringer’s (LR) 2ml x % TBSA x weight (kg.)
- For the first hour only, calculate infusion rate based on one-half the total fluid over 8 hours.
- After the first hour, titrate LR, based on Urine Output (U.O.):
  - U.O. < 0.5 ml/kg/hr, increase by 30%
  - U.O. > 1 ml/kg/hr, decrease by 30%
  - Target: U.O. 0.5-1 ml/kg/hr, no change

Epidermal Loss > 30% Total Body Surface Area

Initial Fluids / Labs – West Penn Formula

- Fresh frozen plasma (FFP), 75 ml x weight (kg) / 24 hr = hourly rate for initial rate then titrate based on U.O.
- LR 83 ml/hr x 48 hr.
- Type and cross match STAT.
- FFP, based on U.O.:
  - U.O. < 0.5 ml/kg/hr, increase FFP by 30%.
  - U.O. > 1 ml/kg/hr, decrease FFP by 30%.
  - Target: U.O. 0.5-1 ml/kg/hr, no change

Pain Management

Consider lower opioid doses for opioid-naive and elderly.

<table>
<thead>
<tr>
<th>Medication</th>
<th>Normal Starting Dose/Route</th>
<th>Onset (min)</th>
<th>Peak Effect (min)</th>
<th>Time (hr.)</th>
</tr>
</thead>
<tbody>
<tr>
<td><strong>Opioids</strong></td>
<td></td>
<td></td>
<td></td>
<td></td>
</tr>
<tr>
<td>Morphine</td>
<td>2-6 mg IVP</td>
<td>5-10</td>
<td>20-30</td>
<td>4-5</td>
</tr>
<tr>
<td>First-line opioid therapy</td>
<td>30 mg oral, immediate acting</td>
<td>15-60</td>
<td>60-120</td>
<td>4-5</td>
</tr>
<tr>
<td>Hydromorphone</td>
<td>0.5-2 mg IVP</td>
<td>10-15</td>
<td>15-30</td>
<td>2-3</td>
</tr>
<tr>
<td></td>
<td>4 mg oral</td>
<td>30</td>
<td>90-120</td>
<td>4</td>
</tr>
<tr>
<td>Fentanyl</td>
<td>25-100 mcg IVP</td>
<td>1-2</td>
<td>3-5</td>
<td>0.5-1</td>
</tr>
<tr>
<td>Methadone</td>
<td>2.5 mg IVP</td>
<td>10-20</td>
<td>1-2</td>
<td>6-12</td>
</tr>
<tr>
<td>Not for acute pain</td>
<td>5 mg oral</td>
<td>30-60</td>
<td>90-120</td>
<td>4-12</td>
</tr>
<tr>
<td>Oxycodeine / Acetaminophen</td>
<td>1-2 tablets oral</td>
<td>15-45</td>
<td>60-90</td>
<td>3-6</td>
</tr>
<tr>
<td><strong>Adjunctives</strong></td>
<td></td>
<td></td>
<td></td>
<td></td>
</tr>
<tr>
<td>Midazolam (Versed)</td>
<td>SICU: 2-5 mg IVP</td>
<td>1-5</td>
<td>30</td>
<td>2-6</td>
</tr>
<tr>
<td>First-line adjunctive therapy</td>
<td>9W Doan: 1-3 mg IVP</td>
<td></td>
<td></td>
<td></td>
</tr>
<tr>
<td>Lorazepam (Ativan)</td>
<td>1-2 mg IVP</td>
<td>5-10</td>
<td>15-20</td>
<td>6-8</td>
</tr>
<tr>
<td></td>
<td>1-2 mg oral</td>
<td>15-45</td>
<td>60-90</td>
<td>6-8</td>
</tr>
<tr>
<td>Ketamine*</td>
<td>SICU: 50-200 mg IVP</td>
<td>≤ 1</td>
<td>1</td>
<td>5-15</td>
</tr>
</tbody>
</table>

* Must administer with concomitant benzodiazepine to avoid emergence
**Medication Management**
Prospective trials have not been conducted other than to confirm increased mortality with thalidomide. Treatments vs supportive care are still contentious and should be discussed between the patient, dermatology, medicine, and burn surgery. However, the disease is a cytotoxic T-cell mediated disease and multiple immunosuppressive options have been considered:

<table>
<thead>
<tr>
<th>Drug Name</th>
<th>Dosage</th>
<th>Frequency</th>
</tr>
</thead>
<tbody>
<tr>
<td>Cyclosporine</td>
<td>3-5 mg/kg</td>
<td>14-30 days</td>
</tr>
<tr>
<td>IVIG</td>
<td>1 g/kg</td>
<td>Daily x 4 days</td>
</tr>
<tr>
<td>Methylprednisolone</td>
<td>250 mg q6 h</td>
<td>3 days</td>
</tr>
</tbody>
</table>

**Nutrition**
- Enteral better than parenteral.
- May be proportional to TBSA involved.
- Immune modulating nutrition with glutamine.

**Skin Care**
- Debridement of sloughed epidermis.
- Application of Xeroform gauze to denuded areas.

**Consults**
- Dermatology
- Nutrition
- PT
- Ophthalmology
- OT
- Depending on mucosal involvement:
  - GYN
  - ENT
  - GI

**Order Sets**
- OSU IP BURN: Admission Non-ICU Burn [2109]
- OSU IP BURN: Admission Critical Care Burn [2111]
- OSU IP BURN: Focused Wound Care [2171]
- OSU IP BURN: Post Burn Surgery [2169]
- OSU IP BURN: Burn Pain Management [1768]

**Quality Measures**
- Frequency and timing of punch biopsies
- Percentage of patients with the following consults upon admission:
  - Burn
  - Ophthalmology
  - Dermatology
- Survival (by SCORTEN)

**References**

**Authors**
- Larry Jones, MD
- Benjamin Kaffenberger, MD
- Rebecca Coffey, RN, CNP
- Claire Murphy, PharmD, BPS

**Guideline Approved**

**Disclaimer:** Clinical practice guidelines and algorithms at The Ohio State University Wexner Medical Center (OSUWMC) are standards that are intended to provide general guidance to clinicians. Patient choice and clinician judgment must remain central to the selection of diagnostic tests and therapy. OSUWMC's guidelines and algorithms are reviewed periodically for consistency with new evidence; however, new developments may not be represented.

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