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 suspicious medications, especially antibiotics or anticonvulsants.
- Obtain early consultation or transfer to Burn Center.
  - 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
- Protect skin while it heals, with special emphasis on care of eyes, oral mucosa, gastrointestinal, and respiratory epithelia.
- Treatment is primarily supportive.

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></td>
<td>1</td>
</tr>
<tr>
<td>Body surface area detached</td>
<td>&gt; 10%</td>
<td></td>
<td>1</td>
</tr>
<tr>
<td>Heart rate</td>
<td>&gt; 120 bpm</td>
<td></td>
<td>1</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:
- 0-1 points = 3% (4 points = 58%)
- 2 points = 12% (5-7 points = 90%)
- 3 points = 35%

Discontinue the likely offending pharmacological agent.

Fluid Resuscitation

**Epidermal Loss 15-30% Total Body Surface Area**

**Initial Fluids – Parkland Formula**
- Lactated Ringer’s (LR) 4 ml 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 U.O.:
  - U.O. < 0.5 ml/kg/hr, infuse 350 ml/hr.
  - U.O. 0.5-1 ml/kg/hr, infuse 175 ml/hr.
  - U.O. > 1 ml/kg/hr, infuse 75 ml/hr.

**Epidermal Loss > 30% Total Body Surface Area**

**Initial Fluids / Labs – West Penn Formula**
- LR 83 ml/hr x 48 hr.
- Type and cross match STAT.
- Fresh frozen plasma (FFP), based on U.O.:
  - U.O. < 0.5 ml/kg/hr, infuse 350 ml/hr.
  - U.O. 0.5-1 ml/kg/hr, infuse 175 ml/hr.
  - U.O. > 1 ml/kg/hr, infuse 75 ml/hr.

Pain Management
Consider lower opioid doses for opioid-naïve 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>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 Not for acute pain</td>
<td>2.5 mg IVP</td>
<td>10-20</td>
<td>1-2</td>
<td>6-12</td>
</tr>
<tr>
<td>Oxycodone / 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) First-line adjunctive therapy</td>
<td>SICU: 2-5 mg IVP</td>
<td>1-5</td>
<td>30</td>
<td>2-6</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>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 phenomena.*
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

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

- Percentage of patients admitted to Burn Center
- Frequency and timing of punch biopsies
- Percentage of patients with the following consults upon admission:
  - Burn
  - Ophthalmology
  - Dermatology

References


Authors

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- Benjamin Kaffenberger, MD

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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