Overview:
Stevens Johnson syndrome is a rare, serious disorder in which your skin and mucous membranes react severely to a medication or infection. Medications include:
- Barbiturates
- Penicillins
- Phenytoin
- Sulfonamides
Infections include:
- Herpes simplex
- Mycoplasma

Often, the syndrome begins with flu-like symptoms, followed by a painful red or purplish rash that spreads and blisters, eventually causing the top layer of your skin to die and shed. Nikolsky's sign – separation of the epidermis from the dermis with pressure—is usually present. It can also affect the mucus membranes of the body, such as the oral cavity or gastrointestinal tract. The condition is extremely painful, and there is a high risk of mortality or complications (60-75% of cases).

Disease manifestations: range from mild SJS (1-10% TBSA) to moderate SJS-TEN overlap (10-30%), to severe TEN (>30% TBSA affected). Extent of TBSA affected correlates with mortality.

<table>
<thead>
<tr>
<th>Classification of TEN/SJS</th>
</tr>
</thead>
<tbody>
<tr>
<td><strong>SJS</strong></td>
</tr>
<tr>
<td>• Skin detachment &lt;10% TBSA</td>
</tr>
<tr>
<td>• Widespread erythematous or purpuric macules or flat atypical targets</td>
</tr>
<tr>
<td><strong>Overlap SJS/TEN</strong></td>
</tr>
<tr>
<td>• Detachment between 10% and 30% of BSA</td>
</tr>
<tr>
<td>• Widespread purpuric macules or flat atypical targets</td>
</tr>
<tr>
<td><strong>TEN with spots</strong></td>
</tr>
<tr>
<td>• Detachment &gt;30% of BSA</td>
</tr>
<tr>
<td>• Widespread purpuric macules or flat atypical targets</td>
</tr>
</tbody>
</table>
**TEN without spots**

- Detachment of >10% of BSA
- Large epidermal sheets and no purpuric macules

**Mechanism:** Not completely understood still, believed to be related to an acute lymphohistiocytic inflammatory infiltrate around blood vessels and degenerative changes in endothelial cells of capillaries. Onset is usually within a week if caused by antibiotics, or within a month for anticonvulsants.

**Complications:** Cellulitis, sepsis, eye problems, damage to internal organs, respiratory tract complications due to mucus retention and sloughing of tracheobronchial mucosa

**Treatments:** Stopping all medications associated with SJS, Supportive Care (wound care, nutrition and fluids), Medications (pain medications, anti-histamines, antibiotics, topical steroids, etc.), Skin Grafting (rare)

**Nutrition and TEN:** Purpose of effective nutrition management in SJS is to promote wound healing, positive nitrogen balance, weight preservation, immune function, and host defense mechanisms – necessary for survival! TEN is a highly catabolic state, and most clinicians/dietitians agree that the energy/protein needs are similar to those of a partial thickness burn patient – although there is debate over what route of getting nutrition in is best. Eating can be very painful and difficult if there is facial or oral involvement, and nutrition needs may exceed the amount that the patient is able to get in. Enteral nutrition is used very often in these cases, while parenteral nutrition is often discouraged because of the risk of sepsis. Most of the more recent literature in this area includes case studies, but it seems the best route is individualized for each patient and their needs, including assessment of the areas affected by TEN and TBSA affected.

**Resources:**

- www.mayoclinic.com
- www.dermnetnz.org
Brief Review of Recent Literature:


This is a more recent review of the literature surrounding the management of toxic epidermal necrolysis. It briefly mentions nutrition under “supportive care”, and encourages the use of early enteral or parenteral nutrition to support wound healing if a patient is unable to ingest food orally. It also urges caution when inserting nasogastric tubes in a patient who has oropharyngeal involvement of the condition. The authors also recommend keeping the room temperature around 30-32 degrees Celsius (or use air-heat body warmers) to prevent excessive caloric expenditure due to epidermal loss.


Compared measured resting energy requirements of children with Stevens-Johnson syndrome or toxic epidermal necrolysis with those of children with burns of similar size. Researchers used indirect calorimetry to find the measured resting energy expenditure (MREE) in 30 pediatric patients. The researchers found that energy needs of the SJS/TEN group were 22%, or around 600 kcal, less than the burn group with comparable wounds, but still above the basal metabolic rate. This study revealed that pediatric patients with TEN/SJS do not exhibit the same degree of hypermetabolism as burn patients with comparable wounds. In conclusion, they recommend the application of a 30% factor to the MREE in pediatric patients. They came up with a formula for use in this population (24.6 x weight in kg) + (% wound x 4.1) + 940. Recommend using caution until formula can be studied further.


Case study of 30 yr old woman who developed TEN over 60% TBSA. Even after her exterior blisters had healed, she continued having bloody diarrhea. An endoscope revealed the presence of ulcers, detachment of villi, and edematous mass on the mucus membrane of the intestine. Some reports show that the development of mucus membranes being affected can be delayed. Important to note this possible effect, especially as it relates to nutrition. There is potential for malabsorption and gastric discomfort.

Retrospective study that looked at 14 patients’ medical records to describe the swallow function in TEN and SJS and define the role of speech pathology in management. Of the 14 patients, 12 had severe mucosal involvement (bleeding from the lips and gums, severe ulceration of all the structures within the oral cavity and pharynx, anorexia, complaints of oral and pharyngeal pain, and poor oral hygiene). Twelve of the 13 patients on an oral diet suffered odynophagia resulting in poor oral intake, and 11 demonstrated ability to tolerate fluids more easily than solids. Enteral feeding had to be instituted in 8 of the 14 patients because of inadequate oral intake and dehydration.


In this letter to the editor, the author discusses the difficulties of randomized control studies in this patient population, due to the rarity of the disease. He also notes his observations that TEN patients do not have the exact same requirements as burn patients (for example, their fluid requirements are not as high), although states that the results on the differences in nutrition requirements are inconclusive. He also makes a case against using the enteral route of nutrition, and notes two patients of his who developed gastrointestinal hemorrhages requiring transfusions during the insertion of nasogastric tubes. In his hospital, a protocol was developed where no nasogastric tubes or central venous lines are used. Instead, two phases of nutrition are administered: first, low osmolarity total parenteral nutrition through the peripheral vein, then a second phase of oral supplements when the patient can tolerate them. There is no discussion on the success of such protocol.


A case report is presented of a 63 year old woman who was admitted to the burn unit with 60% TBSA affected, with excessive oropharyngeal hemorrhage and laryngeal sloughing. She was started on parenteral nutrition on day 1 with 20 grams of glutamine added for immune support. The authors discuss the possibility of a use for immune-enhanced nutrition for patients with this disease. On day 3, oropharyngeal bleeding subsided, and the patient was started on nasogastric feeds with an immune modulating formula that contained added glutamine, arginine, and Omega-3 fatty acids. The patient developed sepsis on day 7, although she made a full recovery and was off the
ventilator by day 26. The patient also had maintained her nutrition status and weight upon discharge. It is extremely difficult to assess the effectiveness of an immune modulating formula against a standard high protein, high calorie formula due to the rarity of the condition and lack of literature or documentation on the types of enteral formulas used. However, the researchers suggest that this type of immune modulating formula might have a place in this patient population. More research is needed in this area to show that the benefit is worth the extra costs.


Researchers looked at data on 199 patients admitted to 15 different burn centers to examine correlates between different factors and survival rates between 1995 and 2000. They examined number of complications, renal dialysis, intubation, and nutrition (parenteral or enteral), and how these factors correlated with mortality after a patient was admitted to a burn unit. They found that early enteral nutrition was instituted in 60% of cases in burn centers, compared with 11.6% in non-burn facilities. The burn centers had much higher survival rates, and this was attributed to the more aggressive wound care, nutrition, critical care, and topical therapies, things that need to be instituted promptly by trained personnel. The researchers also found that early initiation of parenteral nutrition (before patients were transferred to a burn unit) was associated with lower survival rates, and this is not recommended.

Chowdhury, M.M.U. and Holt, P.J.A. (2001), Burning fingers, but where is the fire?. British Journal of Dermatology, 144: 1286. doi: 10.1046/j.1365-2133.2001.04263.x

Correspondence letter – discusses the risks of TPN causing sepsis, and yet the nasogastric tube poses a risk of perforation for those with gastrointestinal involvement. The authors present a case study of a 56 y.o. male with >90% TBSA affected, including sloughing of the entire oral cavity and esophagus. Because of this involvement a PEG was placed, and though the rest of his recovery course was complicated, the PEG seemed to be tolerated well. There is a potential higher risk of infection, however, at the insertion site of a PEG. The authors recommend TPN if the small bowel or colon is affected.

An older review of the literature that contains a small section on nutrition with this condition. Discusses the gastrointestinal involvement in the condition – many patients suffer with dysphagia, and the gastrointestinal tract can be affected for months after recovery. It recommends using the enteral route over parenteral because of the risk of sepsis.

In conclusion, from the slight amount of literature on this patient population:

TEN patients are in a very catabolic state and have elevated and urgent nutrition needs, although probably not quite as much as burn patients, and are often unable to take in adequate nutrients orally. Some of the potential nutrition concerns include malabsorption, pain with chewing or swallowing due to ulcers, hypermetabolic state, and increased risk of sepsis with aspiration. In these cases, enteral nutrition might be very effective in wound healing and maintaining positive nitrogen balance. However, caution should be used when inserting nasogastric/duodenal tubes due to the risk of perforation or irritation of mucus membranes. Administration of parenteral nutrition should be used cautiously, given the small data showing increased mortality associated with early TPN. Clinical judgment should be used on a case by case basis until more conclusive research can be performed, and these judgments should be based upon the total body surface area affected, the involvement (or not) of mucus membranes, and co-existing conditions or presence of sepsis.