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Pathophysiology-Stevens-Johnson Syndrome/Toxic Epidermal Necrolysis

Douglas M. Klein

Otterbein University, douglas.klein@otterbein.edu

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Introduction
Stevens-Johnson Syndrome (SJS) and Toxic Epidermal Necrolysis (TEN) are rare, cutaneous reactions triggered by certain drug metabolites, although the exact mechanism of action has been poorly understood. Evidence has established a genetic predisposition for illness to various Asian populations, leading to FDA recommendations for pre-treatment genetic testing. Additional research has focused on the effects Vitamin A derivatives have on disease process.

Pathophysiology-Overview
SJS/TEN disease has long been presumed to appear as a result of an immune-mediated reaction to certain drug metabolites, but the exact mechanism of action has been poorly understood. Evidence has established a genetic predisposition for illness to various Asian populations, leading to FDA recommendations for pre-treatment genetic testing. Additional research has focused on the effects Vitamin A derivatives have on the disease process.

Disease

<table>
<thead>
<tr>
<th></th>
<th>TBSA %</th>
<th>Mortality</th>
<th>Incidence/ million</th>
</tr>
</thead>
<tbody>
<tr>
<td>SJS</td>
<td>&lt;10%</td>
<td>5%</td>
<td>2-6</td>
</tr>
<tr>
<td>TEN</td>
<td>&gt;30%</td>
<td>30%</td>
<td>0.2-1.2</td>
</tr>
</tbody>
</table>

(Mawson et al., 2015) (>10%, <30% is considered not to be SJS/TEN disease) (TBSA=total body surface area)

Implications: Pathophysiology
Understanding pathophysiology is key for treating and limiting the extent of the disease. Provided that retinal toxicity is involved with SJS/TEN, plasmapheresis could be used to reduce circulating levels and mitigate the destructive effects which retinol contributes in this disease. Plasmapheresis has been shown in small samples to reduce mortality versus typical treatment for patients with SJS/TEN (Mawson, 2015). Genetic predispositions have been positively established for people of Asian descent who carry the HLA-B1502 allele when exposed to Carbamazepine. The FDA recommends genetic testing prior to starting Asian patients on Carbamazepine therapy (Tangamornsukan, Chalyakunapruk, Somchon, Lohthaney, & Tassaneeyakul, 2013). Studies have also looked at higher than normal incidence of SJS/TEN within the Indian population, particularly as they are HIV-positive (Pate, Barvaliya, Sharma & Tripathi, 2013). HLA-A3101 has recently been identified as a phenotype-associated risk among Japanese and Europeans (Pirmehdi et al., 2011). Understanding genetic associations can help healthcare professionals mitigate the risk through pre-treatment testing.

Nursing: Implications
Rapid identification of disease is crucial for patients suffering from SJS/TEN as removal of the offending medication is critical for mitigating the effects of the illness. Nursing should familiarize themselves with the symptoms and signs of progression as illness may advance quickly from SJS to TEN in as few as twelve hours (Poulou, Nielsen, & Poulsen, 2013).

- Nursing should be watchful for signs of symptoms of decompensation due to respiratory failure or sepsis.
- Pain management and the use of conscious sedation, especially during debridement and dressing changes will improve healing and health outcomes.
- Treatment is best done in a specialized burn center.
- The nurse should seek to coordinate a multidisciplinary team approach (Cooper, 2012).

Long-term restrictions for the development of disease is frequently seen with SJS/TEN and patients will often require long-term use of bronchodilators (British Journal of Medicine, 2015).

References


Bershan, A. R., Eriator, J., & Karre, K. A. (2015). Rapid identification of disease is crucial for patients suffering from SJS/TEN as removal of the offending medication is critical for mitigating the effects of the illness. Nursing should familiarize themselves with the symptoms and signs of progression as illness may advance quickly from SJS to TEN in as few as twelve hours (Poulou, Nielsen, & Poulsen, 2013).

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Conclusion: Although SJS/TEN has been recognized illness for decades, recent advancements in genetics have identified strong predispositions to certain drug reactions, helping guide prescribing prior to safe and effective treatment. Patients at risk should be tested for susceptibility before starting therapy. The use of plasmapheresis should be studied further to determine whether limiting free protein components could mitigate the disease caused by damage.