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Stevens Johnson Syndrome and Toxic Epidermal Necrolysis Updates on Management

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ABSTRACT

Stevens-Johnson Syndrome (SJS) and Toxic Epidermal Necrolysis (TEN) are dermatologic emergencies characterized by extensive epidermal necrolysis and sloughing. The incidence rates vary depending on location, with the UK experiencing an annual incidence of 5.76 cases per million individuals from 1995 to 2013. Females are more frequently afflicted than males, with mortality rates ranging from 4.8-9% to 14.8-48%.

Nonpharmacologic treatment for SJS/TEN patients includes supportive care, fluid and electrolyte management, infection control, and wound care. The most critical component is the identification and cessation of the causative substance. Management of fluid, electrolytes, and nutrition is crucial for SJS/TEN patients, as they have lower fluid requirements than burn patients.

Prophylactic antibiotics do not enhance outcomes, but infection prevention requires appropriate wound care and antiseptic handling. The role of surgical debridement has been a topic of controversy, and the decision to pursue this treatment option depends on the location of care. Anti-shear therapy, which involves aspiration of blister fluid and preservation of denuded epidermis, has been shown to be effective in reducing mortality rates.

Pharmacologic treatment for SJS/TEN has been the subject of few prospective studies due to the disease's rarity. Various treatment regimens involving corticosteroids, IVIg, cyclosporine, and TNF-alpha inhibitors have been reported, but it is challenging to determine whether the disease's remission was due to a specific treatment or the natural course of the disease.

KEYWORDS: Stevens–Johnson Syndrome; Toxic Epidermal Necrolysis; cutaneous adverse drug reactions

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INTRODUCTION

Stevens–Johnson Syndrome (SJS) and Toxic Epidermal Necrolysis (TEN) are dermatologic emergencies that are distinguished by extensive epidermal necrolysis and sloughing. They are classified according to the body surface area (BSA) affected and are assumed to have the same pathophysiology. The reported incidence rates of these rare diseases are subject to variation depending on the location. The mortality rates for SJS, SJS/TEN, and TEN are 4.8–9%, 19.4–29%, and 14.8–48%, respectively.

Nonpharmacologic treatment

The primary treatment for patients with SJS/TEN is supportive care, which encompasses the cessation of the causative substance, fluid and electrolyte management, infection control, and wound care. The most critical of these components is the identification and cessation of the causative substance. However, in order to achieve the most favorable results, it is imperative to optimize each measure.

The management of fluid, electrolytes, and nutrition is crucial for SJS/TEN patients, as it mirrors the requirements of burn patients due to insensible losses through wounds. However, the fluid requirements of SJS/TEN patients are approximately 30% lower than those of burn patients with similar degrees of cutaneous involvement. Due to the skin's loss of thermoregulatory function, the environment should be maintained at a temperature of 30–32 °C. Fluid replacement should be facilitated by urine output, with a target of 0.5–1 mL/kg/h. Enteral nourishment should be initiated as soon as practicable, and if necessary, through nasogastric tube feeds.

Prophylactic antibiotics do not enhance outcomes, but infection prevention necessitates appropriate wound care and antiseptic handling. The role of surgical debridement has been

a topic of controversy, and the decision to pursue this treatment option is dependent on the location of the care. Anti-shear therapy is a viable alternative to surgical debridement and has the potential to mitigate both pain and hospital expenses. Nevertheless, the absence of high-quality evidence to inform decision-making regarding surgical debridement necessitates additional research to gain a comprehensive understanding of the therapy's role.

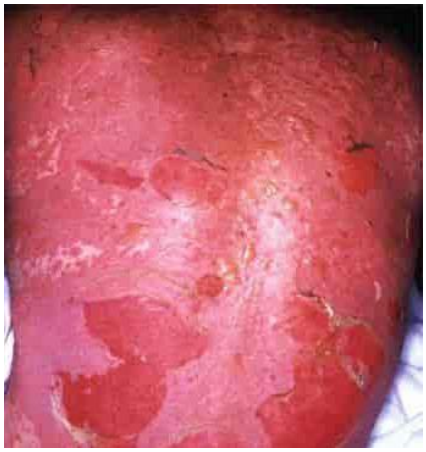

Figure 1. Stevens Johnson Syndrome

Pharmacologic Treatment

The efficacy of specific adjunctive therapies for SJS/TEN has been the subject of few prospective studies due to the disease's rarity. Consequently, there is no established standard of care for pharmacologic treatment. Many case reports have reported positive results with varying treatment regimens involving different combinations of corticosteroids, IVIg, cyclosporine, and TNF-alpha inhibitors, as it is believed that immunosuppressive therapies will aid in treatment due to the immunologic nature of the disease. Nevertheless, it is challenging to ascertain whether the disease's remission was the result of a specific treatment or the natural course of the disease. Numerous systematic reviews and meta-analyses have endeavored to address these methodologic constraints and elucidate the function of pharmacologic therapies in the treatment of SJS/TEN. The monotherapy function of corticosteroids remains a topic of debate. The role of IVIg has also been a topic of controversy, and it seems that monotherapy does not appear to be associated with any mortality benefit.

TNF-alpha inhibitors are also of relevance because of their immunosuppressive properties.

Category	*SJS/TEN Management Updates*
Diagnostic Criteria	- **Early recognition**: Enhanced by tools like the SCORTEN score to predict severity and mortality. - **Biopsy**: Histopathological confirmation is critical for diagnosis.
Initial Management	- **Immediate cessation of causative agents**: Early identification and discontinuation of offending drugs (e.g., anticonvulsants, antibiotics) is crucial. - **Referral to burn unit/ICU**: Patients with extensive skin involvement should be managed in specialized units.
Supportive Care	- **Fluid and electrolyte management**: Crucial to prevent dehydration and renal failure. - **Nutritional support**: Enteral feeding is preferred to maintain gut integrity. - **Wound care**: Use of non-adherent dressings and topical antimicrobials to prevent secondary infections. - **Temperature control**: Maintain a warm environment to prevent hypothermia.
Pharmacologic Treatments	- **Systemic corticosteroids**: Controversial but may be beneficial in the early stages for some patients. - **IV Immunoglobulin (IVIG)**: Conflicting evidence, some studies suggest benefit in halting disease progression. - **Cyclosporine**: Increasing evidence supports its use for reducing mortality and promoting faster recovery. - **TNF-α inhibitors**: Agents like etanercept are being explored for their potential benefits in managing SJS/TEN.
Emerging Therapies	- **Mesenchymal stem cell therapy**: Research is ongoing into its potential for promoting healing and reducing inflammation. - **Amniotic membrane grafts**: Used in ocular involvement to prevent scarring and vision loss. - **Biologics**: Newer biologic agents are being investigated for their immunomodulatory effects.
Monitoring and Follow-up	- **Long-term follow-up**: Essential for detecting late complications such as ocular sequelae, scarring, and psychological impact. - **Regular ophthalmologic exams**: To monitor and manage eye involvement and prevent blindness.

- **Individualized Care**: Treatment should be tailored to the patient's specific needs and the severity of their condition.
- **Multidisciplinary Approach**: Involvement of dermatologists, ophthalmologists, intensivists, and other specialists is crucial for comprehensive care.
- **Research and Guidelines**: Ongoing research and updates to clinical guidelines are essential for optimizing management strategies.

CONCLUSION

Practical considerations, including cost, must be considered when determining the most effective pharmacological treatment for SJS/TEN, as there is a lack of consensus on the most effective treatment.

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