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# Steven Johnson Syndrome and Toxic Epidermal Necrolysis in a burn unit: A 15-year experience☆

M. McCullough [a,*], M. Burg [a], E. Lin [a], D. Peng [b], W. Garner [a]

[a] Division of Plastic Surgery, Keck School of Medicine of University of Southern California, Los Angeles, CA, United States

[b] Department of Dermatology, Keck School of Medicine of University of Southern California, Los Angeles, CA, United States

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ABSTRACT

*Introduction:* The diffuse epidermal exfoliation seen in Steven Johnson Syndrome (SJS) and Toxic Epidermal Necrolysis (TEN) is similar to skin loss in second degree burns, and many of these patients are referred for treatment at burn centers. Treatment can differ markedly from center to center, and mortality can range from 25% to 70%, including a considerable morbidity. However, our experience over a 15-year period from 2000 to 2015 with 40 patients found a mortality rate of only 10% (4/40). The purpose of this paper is to discuss our treatment algorithm as a model for other centers treating SJS/TENs patients.

*Methods:* Records were reviewed for all patients admitted to the LAC + USC burn unit between 2000 and 2015 and 40 patients were identified with biopsy-proven SJS or TENS. These cases were reviewed for age, gender, initial and greatest TBSA, causative drug, pre-existing medical conditions, and morbidity and mortality. All data were entered into the SPSS statistical software package and all statistical analyses were performed using this program.

*Results:* Our treatment algorithm focused on early referral to a specialty burn unit, immediate discontinuation of the offending drug, fluid resuscitation, nutritional supplementation, and meticulous wound care. Average time to transfer to a burn unit was 3.36 days. Silver-releasing antimicrobial dressings were applied to the affected skin surface and changed every 3 days. Mupirocin coated petroleum gauze was used for facial involvement. Steroids were tapered and discontinued if initiated at an outside facility (58% of patients), and starting after 2001, all patients received a course of IVIG. All patients received fluid resuscitation and the majority received supplemental tube feedings (69%). Average length of total stay was 17.1 days and length of ICU stay 15.9 days. While 44% were transferred to another facility for further rehabilitative care, 37% of patients discharge to home. In patients discharged home with complete resolution of skin lesions, time to healing was an average of 14 days.

*Discussion:* With our 10% mortality rate in 40 patients, our study represents a relatively large study population while maintaining a relatively low mortality rate. The demographic data from our study largely aligns with the existing literature, and we therefore feel that our low mortality rate is due to our treatment algorithm, rather than to a less severe pathology in our

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* *Corresponding author at*: 1510 San Pablo Street, Suite 415, Los Angeles, CA 90033, United States. Fax: +1 323 442 7901.
E-mail address: Meghan.McCullough@med.usc.edu (M. McCullough).

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patient population. This claim is supported by a standard mortality ratio of 1.68. This ratio proves a significantly improved mortality than would be expected based on disease severity on admission.



## 1. Introduction

The diffuse epidermal exfoliation seen in Steven Johnson Syndrome (SJS) and Toxic Epidermal Necrolysis (TEN) is similar to skin loss in second degree burns, and therefore many of these patients are referred for treatment at burn centers. The epithelial loss induced by these syndromes underlies their high morbidity and mortality as it predisposes the patient to bacterial and fungal infections as well as respiratory, gastrointestinal, nutritional and ocular complications [1]. Treatment can differ markedly from center to center, but most authors agree that optimal outcomes are achieved through early transfer to a burn center, appropriate wound care and vigorous nutritional support [2–5]. Mortality of SJS and TEN has been described to range from 25% to 70% [6], including a considerable morbidity [7]. However, our experience over a 15-year period from 2000 to 2015 with 40 patients found a mortality rate of only 10% (4/40). The purpose of this paper is to discuss our treatment algorithm.

## 2. Methods

Records for all patients admitted to the LAC + USC burn unit between 2000 and 2015 were reviewed for a diagnosis of either SJS or TEN. Forty-five patients were collected and the diagnosis confirmed by tissue biopsy specimen in 42. Three patients were excluded from the study as their biopsy results were non-confirmatory. Two of these patients were diagnosed with pemphigus and the third patient was diagnosed with an autoimmune thrombotic vasculitis. Two other patients were additionally excluded for co-morbid lupus and another for co-morbid DRESS (drug reaction with eosinophilia and systemic symptoms) syndrome. The entity of Rowell's disease, or a cutaneous manifestation similar to SJS and TENS in the setting of a lupus exacerbation, has been well described. The histologic features of this condition are analogous to SJS and TENS; however, it is differentiated as a distinct clinical entity based on the distribution of the cutaneous involvement and the absence of a provoking drug [8]. DRESS syndrome similarly shares histologic features with SJS and TENS but is also differentiated by several clinical features including organ involvement and affected skin distribution [9].

The remaining 40 patient's charts were then reviewed for age, gender, initial and greatest total body surface involvement (TBSA), pre-existing medical conditions and causative drug. Evaluation for the causative drug was undertaken upon admission with a thorough review of all patient medications. The offending agent was determined by the temporal relationship between use of the drug and symptom onset. Complications over the course of the patient's stay were recorded, including respiratory failure as defined by the necessity of intubation, renal failure as defined by the necessity of continuous renal replacement therapy (CRRT) or hemodialysis (HD), shock defined as hypotension utilising vasopressor support, and infection as defined by a positive culture. Infectious complications were further evaluated by specific location and bacteriology. Additionally, ocular complications were examined as well as interventions by the Ophthalmology service. The use of steroids and intravenous immunoglobulin (IVIG) was noted as well as other drug use including antibiotics. The time to transfer to the burn unit, total length of stay, total length of ICU stay and discharge disposition were also recorded.

All data were entered into the SPSS statistical software package and all statistical analyses were performed using this program.

## 3. Treatment algorithm

The treatment of our patients was based on our experience treating patients with thermal burns. Most patients received resuscitation on admission to maintain generalized tissue perfusion. Resuscitation was initiated with Ringer's lactate, then changed to standard IV fluid support thereafter. The goal was to maintain a urinary output of 0.5–1.0 cc/kg/h. Because adequate nutrition has been found essential to recovery from burn, all patients were intensely monitored for adequate calorie and protein intake. Goals were based on the recommendations of a certified nutritional specialist and titrated according to prealbumin and C-reactive peptide levels monitored every 3 days. If patients were not spontaneously eating at the recommended levels, supplements of nasogastric tube feedings were provided. The need for administered tube feedings was common for patients with more than 20% TBSA epidermal loss and those with mucosal involvement. Enteral nutrition was started within 24 h on high risk patients and titrated per nutrition recommendations based on the total body surface area affected. All non-essential medications were stopped upon arrival at our burn center. Patients did not receive glucocorticoid treatment; if the patient was transferred on steroids, the steroids were weaned quickly and then discontinued. Starting after 2001, all patients were given either a 3 or 5-day course IVIG at 1 mg/kg/day, which was initiated within 24 h of arrival to the unit, or of confirmative biopsy if already in house. If IVIG was initiated at an outside hospital, the course was completed. Only proven infections were treated with appropriate antibiotic coverage based on low risk of the exacerbation of skin loss.

On admission the patient underwent wound debridement to remove all sloughed epidermis, fibrinous exudate and