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Improving mortality outcomes of Stevens Johnson syndrome/toxic epidermal necrolysis: A regional burns centre experience

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ABSTRACT

Introduction: Stevens Johnson Syndrome/toxic epidermal necrolysis (SJS/TEN) are rare, potentially fatal desquamative disorders characterised by large areas of partial thickness skin and mucosal loss. The degree of epidermal detachment that occurs has led to SJS/TEN being described as a burn-like condition. These patients benefit from judicious critical care, early debridement and meticulous wound care. This is best undertaken within a multidisciplinary setting led by clinicians experienced in the management of massive skin loss and its sequelae. In this study, we examined the clinical outcomes of SJS/TEN overlap & TEN patients managed by our regional burns service over a 12-year period. We present our treatment model for other burn centres treating SJS/TEN patients.

Methods: A retrospective case review was performed for all patients with a clinical diagnosis of TEN or SJS/TEN overlap admitted to our paediatric and adult burns centre between June 2004 and December 2016. Patient demographics, percentage total body surface area (%TBSA), mucosal involvement, causation, severity of illness score (SCORTEN), length of stay and survival were appraised with appropriate statistical analysis performed using Graph Pad Prism 7.02 Software.

Results: During the study period, 42 patients (M26; F: 16) with TEN (n=32) and SJS/TEN overlap (n=10) were managed within our burns service. Mean %TBSA of cutaneous involvement was 57% (range 10-100%) and mean length of stay (LOS) was 27 days (range 1-144 days). We observed 4 deaths in our series compared to 16 predicted by SCORTEN giving a standardised mortality ratio (SMR) of 24%.

Conclusion: Management in our burns service with an aggressive wound care protocol involving debridement of blistered epidermis and wound closure with synthetic and biological dressings seems to have produced benefits in mortality when compared to predicted outcomes.

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1. Introduction

Stevens Johnson syndrome (SJS) and toxic epidermal necrolysis (TEN) are severe mucocutaneous exfoliative reactions characterized by fever and extensive necrosis and detachment of the epidermis, most commonly as a result of an adverse drug reaction [1]. SJS and TEN are considered a disease continuum distinguished chiefly by severity, based upon the percentage total body surface area (%TBSA) involved [1].

SJS is the less severe condition, with <10%TBSA involvement. TEN involves detachment of >30%TBSA. Mucous membranes are affected in over 90% of patients for both conditions, usually at two or more distinct sites (ocular, oral, and genital). SJS/TEN overlap describes patients with skin detachment of 10-30%TBSA. For the purposes of this article the term “SJS/TEN” will be used to refer collectively to SJS, TEN, and SJS/TEN overlap.

The extensive degree of epidermal detachment that occurs during the disease progression has led to SJS/TEN being described as a burn-like condition with a number of studies suggesting that the multidisciplinary approach to management of SJS/TEN patients in burns centres may improve survival [2-4]. Treatment can differ markedly between centres, and mortality rates have been described in the literature ranging from 10% to 75% [5,6]. SCORTEN is a scoring system used to predict mortality in toxic epidermal necrolysis (TEN) patients and has been found to be accurate for estimation of mortality among TEN patients treated in a burn centre setting [7]. The optimal therapeutic approach to wound care in SJS/TEN remains controversial, with different approaches used at different centres [8-11]. The results in the literature for the use of IVIG for SJS/TEN are limited and conflicting [12-17]. In this study, we examined the clinical outcomes of SJS/TEN patients managed by our regional burns service over a 12-year period to appraise outcomes, compare these to SCORTEN mortality prediction scores and assess local performance against international comparators. We present our treatment algorithm and discuss the rationale for using this approach as a model for other burns centres treating SJS/TEN patients.

2. Materials and methods

A retrospective case review was performed for all patients with a coding diagnosis of ‘Non-Burn’ skin loss admitted over a 12 year period to our regional burns service between June 2004 and December 2016. Case notes were manually reviewed to identify patients with TEN or SJS/TEN overlap. All other cases of non-burn skin loss were excluded from the study (Fig. 1). Patients included were confirmed as SJS/TEN by a tissue biopsy and consultant dermatologist review. Patient demographics, extent of necrolysis (%TBSA), mucosal involvement, eye involvement, causation, severity of illness score (SCORTEN), time to transfer to burns unit, length of stay (LOS), admission lactate level, admission base excess levels, and survival outcomes were recorded. For the purposes of this study acute kidney injury was defined by the criteria set out by the Acute Kidney Injury Network [18], organ insufficiency and immunosuppression was defined by the APACHE 2 severity of disease classification system [19]. Statistical analysis was performed using Graph Pad Prism 7.02 software.

The burns unit utilised a multidisciplinary team (MDT) approach to care, including plastic surgeons, dermatologists, intensivists, paediatricians, microbiologists, pain team, dieticians, physiotherapy, pharmacy, psychology and specialist skincare nursing. Additional clinical input to the MDT was received from other specialists including respiratory medicine, gastroenterology, gynaecology, urology, ophthalmology and oral medicine on an individual basis. Following referral early transfer to the burns intensive care unit (BICU) was encouraged and barrier nursing was used to prevent nosocomial infections. All patients transferred to the burns centre were given care consisting of clinical assessment, analgesia and fluid resuscitation titrated to achieve a urine output of 0.5-1ml/kg/h. Individualized fluid management was adjusted on a daily basis. The causative agent was immediately discontinued if not already stopped prior to admission. When necessary, continuous invasive haemodynamic monitoring through arterial and central venous lines were utilized to guide fluid resuscitation. Intravenous immunoglobulin (IVIG) was administered at a dose of 0.5-1mg/kg/day for 3 days. Nutritional estimations and input were given based on the

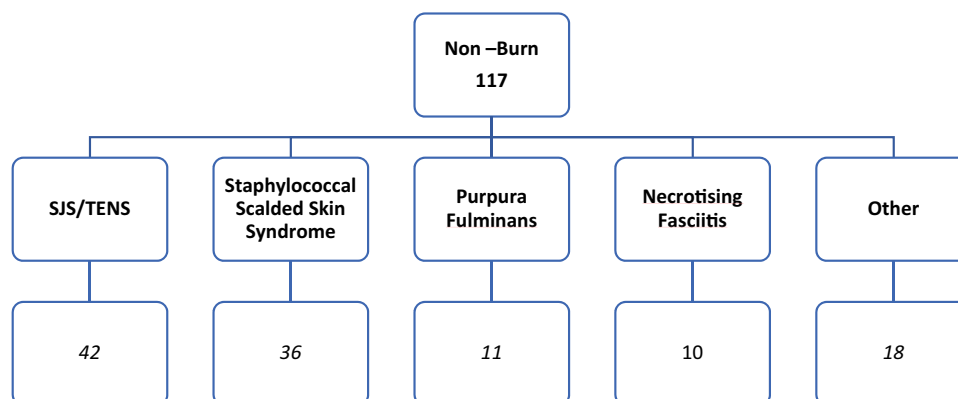


Fig. 1 – Non-burn skin loss cases.

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