

Management of Dysphagia in Toxic Epidermal Necrolysis (TEN) and Stevens-Johnson Syndrome (SJS)

Nicola A. Clayton, BAppSc, and Peter J. Kennedy, MB, BS, MDS, FRACS

Burns Unit, Concord Repatriation General Hospital, Hospital Road, Concord, New South Wales, 2139, Australia

Abstract. Toxic epidermal necrolysis (TEN) is a rare and potentially fatal skin disorder, precipitated by severe allergic drug reaction, and is one of a spectrum of conditions, which includes Stevens-Johnson syndrome (SJS). Mucosal involvement is common, resulting in extreme pain on swallowing and poor oral intake. The aim of this study was to describe swallow function in TEN and SJS and define the role of Speech Pathology in management. The Burns Unit database was reviewed for patients that presented over a five-year period with TEN and SJS. Diagnosis of TEN and SJS was confirmed by skin biopsy. Information specific to swallow function, treatment approaches, and adequacy of oral intake was collected. Fourteen patients' medical records were studied: eight TEN, two TEN/SJS spectrum, and four SJS. The majority had mucosal involvement causing odynophagia, poor oral intake, an ability to tolerate fluids more easily than solids, and increased aspiration risk. These symptoms were confirmed by Speech Pathology swallowing assessment. Severe mucosal involvement resulting in odynophagia, dysphagia, and poor oral intake is common in TEN and SJS. The speech pathologist is able to assess swallow function and provide recommendations to promote safe oral intake, minimize odynophagia, and facilitate nutritional input critical to optimizing recovery.

Key words: Toxic epidermal necrolysis — Stevens-Johnson syndrome — Mucosal involvement — Dysphagia — Odynophagia — Management — Deglutition — Deglutition disorders.

The swallowing function of patients with toxic epidermal necrolysis (TEN) or Stevens-Johnson syndrome (SJS) has not been previously described. Toxic epidermal necrolysis is a potentially life-threatening rare skin disorder that occurs as a result of severe allergic drug reaction [1–4], with an incidence of 0.4–1.2 cases per million [3]. This disease is one of a spectrum of skin disorders, which includes Stevens-Johnson syndrome, and is characterized by lesions of the skin and mucous membranes known as bullae, with associated erythema and epidermal detachment [3, 5]. The extent of skin necrosis is often equated to a partial thickness burn [4], as deep blistering occurs initially, followed by exfoliation of the affected skin and subsequent spontaneous wound healing. Although TEN and SJS present with similar symptoms and causative agents [6], epithelial loss is usually greater in TEN [2, 7]. Mortality rates for this disease vary considerably within the literature, ranging from 20% to 75% [4]. Comorbidity is not infrequent and is often the primary cause of death [3].

Mucosal involvement in both skin disorders is common. Mucosal erosions, presenting as inflammation and severe ulceration of the conjunctiva, oral cavity, pharynx, nasal cavity, esophagus, and genitalia, are frequently observed, causing complaints of photophobia and extreme odynophagia [3]. The oropharynx is also reportedly the most common area for mucositis [5], often persisting after other external areas have re-epithelialized [8]. The tenacity of these symptoms may have nutritional ramifications.

Toxic epidermal necrolysis patients are often best treated within a burns unit for expert wound management because of extensive skin detachment and high risk of infection [2, 4]. Studies suggest that early transfer to a burns unit decreases mortality [4]. Improved survival rates in burn centers were attributed to

Correspondence to: Nicola A. Clayton, BAppSc, ; E-mail: ncla3136@mail.usyd.edu.au; nicola.clayton@email.cs.nsw.gov.au

use of enteral nutrition, avoidance of prophylactic antibiotics or corticosteroids, and implementation of clearly defined wound management procedures. In addition, the burns unit provides a multidisciplinary team approach to patient care, thus facilitating efficacious rehabilitation. Research indicates that the ideal management of TEN involves prompt diagnosis, immediate cessation of any potential causative agent, and timely transfer to a burn facility [2–4, 7]. A number of pharmaceutical treatment strategies have also been reported in the literature, including the use of intravenous immunoglobulins (IVIG) [3].

Materials and Methods

The Burns Unit database of Concord Repatriation General Hospital was reviewed for all patients admitted with TEN and SJS between 1999 and 2004. The age and gender of the patient, length of hospital stay, and outcome were noted. Biopsy was performed on all patients and they were classified according to the criteria of Bastuji-Garin et al. (1993) [9]. The medical record and Speech Pathology notes, which are recorded prospectively during the patient's admission, were then reviewed. Information specific to oral and pharyngeal symptoms of dysphagia, odynophagia, dysphonia, treatment approaches, and adequacy of oral intake was noted for each patient.

Results

Fourteen patients' medical records (6 male, 8 female) were studied: eight TEN, two TEN/SJS spectrum, and four SJS. The two patients classified as TEN/SJS spectrum were unable to be definitively diagnosed by skin biopsy as a single disease entity. Data were coded into nominal and ordinal scales and statistical analysis was completed using a software package (SPSS, Inc., Chicago, IL).

Twelve of the 14 patients (85.7%) had mucosal involvement as a symptom of their disease. Documented symptoms of mucosal manifestation included bleeding from the lips and gums, severe ulceration of all structures within the oral cavity and pharynx, anorexia, complaints of oral and pharyngeal pain, and poor oral hygiene. Twelve of the 13 patients (92.3%) on an oral diet at the time of their admission suffered odynophagia resulting in poor oral intake, and 11 (84.6%) demonstrated an ability to tolerate fluids more easily than solids. The one patient who did not commence an oral diet during her stay experienced a prolonged period in the intensive care unit and eventually underwent tracheostomy due to unsuccessful extubation and

respiratory failure. By the time of transfer back to the referring facility, mechanical ventilation still being required, TEN had resolved. There was one death within the series due to severe sepsis that occurred after TEN had resolved.

Six patients (42.9%), all diagnosed as TEN, were referred to Speech Pathology for assessment of swallow function. Of these six patients, 16.7% presented with oral dysphagia only, whereas 83.3% presented with oral and pharyngeal dysphagia on initial examination. Dysphagic features were predominantly of the oral phase: poor mouth opening and lip seal (66.7%), reduced and effortful bolus preparation (100%), poor bolus control (66.7%), and impaired oral clearance (66.7%). Pharyngeal phase dysphagic features were most commonly a delayed swallow initiation (83.3%), reduced hyolaryngeal excursion (83.3%) and clinical symptoms of laryngeal penetration or aspiration on thin fluids (66.7%). Pharyngeal phase dysphagic features were particularly apparent in those intubated earlier in their admission.

Discussion

Toxic epidermal necrolysis (TEN) and Stevens–Johnson syndrome (SJS) are often compared with partial thickness burn injury [4]. The rationale for this comparison is that superficial skin loss occurs in all three conditions. Spontaneous wound healing also takes place without the need for surgical grafting, and patchy pigment changes may be seen in all groups. Patients with TEN are best treated within a burns unit in view of the specialized wound management practices, including appropriate dressings that may not be available in other facilities [2, 4].

There are also several differences between TEN and partial thickness burn injury. TEN results from an immunologic reaction, whereas burns are traumatic in origin. There is separation of the tissues at the dermal-epidermal junction in TEN, while partial thickness burns may be variable in depth depending on the degree of dermal involvement. Mucosal ulceration and odynophagia are predominant features of TEN and SJS, whereas these are seen only in burn patients in whom there is a component of burn caused by inhalation or ingestion. Definitive diagnosis of TEN and SJS is also dependent upon tissue biopsy. Finally, while mortality in partial thickness burns is relatively low, the incidence of death in TEN ranges from 20% to 75% [4], frequently associated with sepsis and organ failure.

There are clearly certain parallels between the conditions of partial thickness burn and TEN. These will now be addressed with reference to the importance of assessing swallow function and facilitating nutrition.

Optimal levels of nutritional intake via the oral route may be difficult to achieve and maintain in the burn patient as dysphagia and odynophagia are common [10]. Dysphagia in the burn patient is often related to inhalation injury, prolonged endotracheal intubation, and tracheostomy [10, 11]. Inhalation injury and intubation may irritate mucosa within the laryngopharynx, potentially causing edema, vocal cord paresis, and consequently impaired airway protection [12, 13]. Presence of a tracheostomy tube has been reported to alter swallowing mechanics by anchoring the larynx, resulting in impaired hyolaryngeal excursion and epiglottic inversion, consequently reducing airway protection during deglutition [14–18]. Adequate airway protection is essential during the act of swallowing; impairment places the patient at greater risk of aspiration and development of pulmonary complications such as pneumonia.

Swallowing evaluation is an integral component in the multidisciplinary treatment of patients with TEN and SJS. Severe sloughing of the oral, nasal, and pharyngeal mucosa occurs in this population resulting in extreme pain at rest and on swallowing. It is postulated that this may potentially affect the dynamics of the swallow mechanism, particularly if the patient has also been intubated as part of the medical treatment.

Ward et al. [10] retrospectively examined the pattern of dysphagia rehabilitation in a group of 30 patients with thermal burn injuries, with and without inhalation injury, diagnosed with dysphagia. All patients were ventilated via endotracheal tube on admission and 80% required subsequent tracheostomy. Results demonstrated that the mean duration between admission and first safe oral intake was 30 days. The mean duration between admission and total safe oral feeding without enteral supplementation was 53 days. Texture modification was used to assist the transition to oral intake in all patients. The largest proportion of patients was initially commenced on thickened fluids, while puree consistency was the most frequently prescribed first diet. Ninety percent of patients were noted to be tolerating a full diet and thin fluids by discharge, no longer requiring any form of alternate nutrition. The mean duration until return to a full diet without restrictions was 57 days.

In the present study texture modification was also used to assist transition to oral intake. Following Speech Pathology assessment of swallow function, management plans were made in conjunction with the dietitian, who frequently recommending a diet of thin fluids and puree consistency foods. The patient and caregiver(s) were also taught safe swallowing guidelines, including optimal positioning, to reduce risk of aspiration at this time. Based on clinical examination of swallowing, videofluoroscopy was not used in the subject group as it was determined that this procedure was not clinically indicated. In future studies data pertaining to measurement of hyolaryngeal excursion, timing of the swallow, pharyngeal contraction, and incidence of laryngeal penetration and aspiration as determined by videofluoroscopy or endoscopy would be useful within this population.

McKinnon-DuBose et al. [11] further examined the pattern of dysphagia recovery after thermal burn injury. A retrospective review of medical records of patients admitted to a burn unit and referred for swallowing evaluation over a six-year period was conducted. Results indicated a strong positive linear relationship between percent total body surface area (TBSA) burnt, number of days with tracheostomy, number of days on a ventilator, and days to oral feeding. The authors also describe another positive relationship, although weak, between presence of inhalation injury, duration of oral endotracheal intubation, and days to oral feeding. These results suggest that patients who suffer respiratory compromise as a consequence of their burn injury will require a greater duration of enteral feeding before transition to oral intake. Another key finding of this study demonstrated that a large proportion of patients (78.6%) require diet and fluid modification in the early stages of feeding to maintain safety of oral intake and adequate nutritional levels.

The aim of this study was not only to describe swallow function in TEN but also define the role of Speech Pathology in facilitating recovery. Strategies implemented by the speech pathologist for the management of odynophagia resulting from severe mucosal involvement and to facilitate ease of oral intake are described below.

Application of topical anesthesia such as xylocaine or lignocaine viscous before oral intake reportedly relieved pain during the oral phase of swallowing, but this was recommended only in the absence of pharyngeal dysphagia so as not to raise aspiration risk. Modifications to the temperature of food and fluid were implemented as patients commented on increased odynophagia with substances of



Fig. 1. Epithelial loss in a TEN patient.



Fig. 2. Inflammation and ulceration of the oral cavity in a TEN patient.

very high or low temperatures. Food and fluid at ambient temperature was accepted with greater ease. Acidity was another factor that influenced the degree of pain on swallowing; patients avoided oral intake of foods with higher acidity. The final factor that appeared to elevate the level of odynophagia was texture of certain foods. This was assumed to be due to “rougher” foods feeling more abrasive and consequently aggravating the mucosal wounds. As a result, the speech pathologist often recommended a diet of “smooth” and/or “moist” foods so as not to irritate the oral lesions.

Specific strategies used to minimize trauma to the mouth and lips during food and fluid delivery included the use of smaller utensils and syringes. Specific guidelines for meticulous oral care frequently requested administration of antifungal agents (e.g., nilstat, mycostatin), medicated mouthwash (e.g., bicarbonated soda, half-strength hydrogen peroxide), and topical cream to the lips (e.g., vasoline, lanolin).

The role of Speech Pathology in management of the burn patient has been briefly described in the literature [19, 20; G. Coffey, G. O’Loughlin, F. Li, unpublished, 1999]. Williams and Baker [19] describe the speech pathologist as a key member of the

multidisciplinary burn team, particularly for those patients with head and neck burns. The patient with head and neck burns often sustains an inhalation injury resulting in damage to the oropharyngeal and potentially laryngeal mucosa, frequently necessitating endotracheal intubation. Consequences of inhalation injury and the need for respiratory support often include dysphagia, odynophagia, and dysphonia. The speech pathologist is able to assess voice and swallow function and provide a rehabilitation program to facilitate the safe transition to oral intake. There are many similarities between the clinical presentation of the head and neck burns patient and the TEN patient. For these reasons, in addition to those above, it is postulated that the speech pathologist also plays a vital role in the management of the patient with TEN.

In the present study all patients demonstrated a slow transition to oral intake. It was noted that once a patient showed signs of ability to tolerate adequate amounts of oral intake of a modified diet, progress toward the patient’s pre-morbid diet was rapid. Twelve of the 13 (92.3%) patients who started oral intake during their admission resumed a normal or near-normal diet by the time of discharge. The remaining patient passed away due to severe sepsis.

Nutrition is well documented to be a key component in facilitating successful rehabilitation of the burns patient [10, 21, 22]. In addition, research into burns and TEN has demonstrated that early enteral, but not parenteral, feeding reduces mortality [4, 10]. Thermal burn injury may induce hypermetabolism due to the release of stress hormone cortisol, catecholamines, and glucagon, which manifest as tachycardia, hyperthermia, and protein wasting [23]. This subsequently results in elevated nutritional demands to assist in preventing excessive weight loss, preserve muscle tissue, promote rapid wound healing following debridement, and sustain immunocompetence [10].

This evidence is supported in the current study as the nutritional status was also of concern in several of the patients examined. Enteral feeding was instigated in 8 (57%) of the 14 patients: 7 of 8 TEN, 0 of 2 TEN/SJS spectrum, and 1 of 4 SJS, largely because of inadequate oral intake and dehydration. Complications related to enteral feeding tubes included difficult insertion, patient refusal of insertion secondary to extreme pain, and tube removal by the patient as a result of accident or acute confusion.

Conclusion

Severe mucosal involvement resulting in odynophagia, dysphagia, and poor oral intake is common in TEN and SJS. These symptoms need to be detected early and warrant prompt referral to the speech pathologist so that an appropriate oral feeding regimen can be implemented as early as possible. The speech pathologist is able to assess swallow function and provide recommendations to promote safe oral intake, minimize odynophagia, and therefore facilitate nutritional input critical to optimizing recovery.

Achieving optimal nutritional input also proves to be a challenging task in the TEN patient as a result of dysphagia and odynophagia. Insufficient oral intake and increased metabolic needs are routine indicators for enteral feeding in this population. However, it should be noted that insertion of a feeding tube may be extremely painful, particularly if the nasopharyngeal mucosa is affected. Additional pain relief may be required to assist tube placement.

Finally, the use of topical anesthesia appears to be effective in reducing odynophagia; however, further investigation is required to examine the effects on pharyngeal swallow function and possible risks associated with its use.

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