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Indications for intubation and early tracheostomy in patients with Stevens–Johnson Syndrome and Toxic Epidermal Necrolysis

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Abstract

BACKGROUND: Stevens–Johnson Syndrome (SJS) and Toxic Epidermal Necrolysis (TEN) result in epidermal sloughing and mucositis. There are no published guidelines for intubation and early tracheostomy in this patient population.

METHODS: A retrospective chart review of 40 patients admitted from 2010 to 2015 with SJS and TEN was conducted. Descriptive statistics and significance were calculated.

RESULTS: Of the 43% of patients who underwent early tracheostomy, 100% had oral involvement while the initial total body surface area (TBSA) was 70% or more in 41% of patients ($P < .05$). TBSA progressed 15% or more in 53% of patients with 6% having airway involvement and a neurologic diagnosis mandating intubation. Mortality was 17%.

CONCLUSIONS: Indications for intubation and early tracheostomy for SJS and TEN are documented oral involvement plus one of the following: initial TBSA 70% or more; progression of TBSA involved from hospital day 1 to hospital day 3, 15% TBSA or more; underlying neurologic diagnosis preventing airway protection; and documented airway involvement on direct laryngoscopy.

Stevens–Johnson Syndrome (SJS) and Toxic Epidermal Necrolysis (TEN) are severe cutaneous adverse reactions that result in skin sloughing at the dermal–epidermal junction with associated mucositis. Lyell was the first to describe TEN when he reported four cases of toxic eruptions and sloughing of the skin in 1956.[1] Stevens and Johnson were the first to report two pediatric cases of skin sloughing and conjunctivitis in 1922.[2] SJS is defined as epidermal detachment of 10% or less of the total body surface area (TBSA). TEN is epidermal detachment of 30% or more of the TBSA. There is a syndrome of SJS/TEN overlap with epidermal detachment between 10% and 30% of the TBSA.[2–4] The onset of these syndromes is triggered by a preceding medication, oftentimes an antibiotic, leading to apoptosis of keratinocytes.[5–7] Patients experience a prodromal phase with flu-like symptoms 1 to 3 days before the onset of the acute phase. During the acute phase of the disease, mucocutaneous blistering lesions appear followed by epidermal sloughing leading to various degrees of exposed dermis[8,9] (Supplementary Figs. 1 and 2).

These diseases are rare but fatal with 1 to 2 cases per million per year reported.[10,11] Mortality rates as high as

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10% and 40% have been noted for SJS and TEN, respectively.[12–14] There is great variability in the management of patients with SJS and TEN. The mainstay of treatment involves the discontinuation of the offending agent in addition to supportive care.[15–18] At our institution, we use supportive care and hydrotherapy wound care to treat our patients. We do not use corticosteroids or intravenous immunoglobulin in the treatment of our patients. Intravenous immunoglobulin is thought to inhibit keratinocyte apoptosis by binding at the Fas ligand receptor, but success rates are markedly variable and not well defined in the literature.[19–24]

A particular challenge a clinician faces in the management of SJS and TEN stems from oral and upper airway involvement. Painful oral lesions in the acute phase of the disease result in crusting lesions at the vermillion border (Supplementary Fig. 3) with excessive salivation and dysphagia.[25] The mucous membrane involvement may extend to the larynx causing inflammation and swelling resulting in a compromised airway with additional bronchial epithelial sloughing leading to infection, edema, and atelectasis.[26–30] It has been estimated that 40% of patients with SJS and TEN will experience a respiratory complication with an estimated 20% of SJS and TEN patients requiring mechanical ventilation.[31–36] In addition to acute complications, chronic pulmonary complications including bronchiolitis obliterans have been described.[37–42]

Although there are general clinical criteria for endotracheal intubation including respiratory failure, inability to manage secretions, shock, altered mental status, and pain control, there are no published guidelines for intubation and early tracheostomy specific to the SJS and TEN patient population. Because this patient population has a high prevalence of respiratory complications, we sought to identify criteria for intubation and early tracheostomy in patients with SJS and TEN before the onset of respiratory failure.

Patients and Methods

An IRB-approved retrospective chart review of 40 adult patients admitted to our burn unit at Grady Hospital between the years of 2010 and 2015 with a biopsy-confirmed diagnosis of SJS and TEN was performed. Those patients who underwent tracheostomy by ventilator day 10 were defined as having undergone an "early" tracheostomy. We compared the rates of oral involvement, initial TBSA sloughed at presentation, progression of sloughing, and score of toxic epidermal necrosis (SCORTEN) values between SJS and TEN patients we elected to intubate and perform early tracheostomy on vs those who were not intubated. Outcomes including mortality, bacteremia and rates of bacteremia clearance, pneumonia, disposition location, and decannulation rates were tabulated. Descriptive statistics, unpaired t test, and calculation of P values were performed.

Results

We identified 40 adult patients who were admitted to our institution between 2010 and 2015 with a biopsy-confirmed diagnosis of SJS or TEN. Of those patients we admitted, 43% (n = 17) underwent intubation and early tracheostomy before ventilator day 10. These patients were intubated by hospital day 3 ± 2.2 with an average P:F ratio of 335 ± 17.1 postintubation. Overall, 45% (n = 18) of our admitted patients did not undergo intubation or tracheostomy. We found that those patients in our intubation and early tracheostomy group were slightly older (51 ± 18.7 vs 41 ± 17.1 years) and were more likely to have an admission diagnosis of TEN (82% vs 23%) in comparison with our nonintubated group ($P < .05$; Table 1). We did not find a statistically significant difference concerning the causative agent. In both groups, antibiotic exposure was more commonly reported.

When we retrospectively examined the patients in our intubation and early tracheostomy group a pattern of clinical characteristics emerged. Concerning those patients who underwent intubation and early tracheostomy, documented oral involvement was present in 100% of those patients. The initial TBSA of epidermal sloughing was noted to be 70% or more in 43% of patients who were intubated. Progression of TBSA evolved from hospital day 1 to hospital day 3, 15% or more TBSA, was noted in 53% of our intubated SJS/TEN patients, whereas 6% had an underlying neurologic diagnosis of stroke and documented airway involvement on direct laryngoscopy (Fig. 1). We found these clinical parameters to be present in our patients who were intubated and underwent early tracheostomy compared with those who did not undergo these procedures.

SCORTEN is a predictive probability score for mortality in patients with SJS and TEN.[43–45] It factors age, malignancy, heart rate, body surface area involved, blood urea nitrogen, bicarbonate, and glucose levels into its prediction of mortality.[46] We did not find SCORTEN values to be predictive of our decision to intubate and perform early

Table 1 Patient demographics

Demographic	Intubated and early tracheostomy	Not intubated	P value
Age (average)	51 ± 18.7 y	41 ± 17.1 y	.02
Sex	F-62% M-38%	F-67% M-33%	.03
Diagnosis	SJS-6% SJS/TEN-12% TEN-82%	SJS-39% SJS/TEN-33% TEN-28%	.002
Agent	Antibiotic-50% Anti-seizure-12% Other-38%	Antibiotic-55% Anti-seizure-27% Other-18%	.30

F = female; M = male; SJS = Stevens–Johnson Syndrome; TEN = Toxic Epidermal Necrolysis.