Stevens Johnson Syndrome in Children: Consider Monitoring for Bronchiolitis Obliterans

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Background

- Stevens Johnson Syndrome (SJS) is a disorder characterized by widespread inflammation, typically triggered by an infection or drug exposure.
- SJS can manifest in cardiovascular, pulmonary, gastrointestinal, and genitourinary organ systems.
- A small percentage of patients with SJS develop bronchiolitis obliterans (BO), a disease characterized by obstruction of the bronchioles and subsequent air trapping.

Methods

- The Children’s Hospital Colorado electronic medical record was queried from 1/1/2004-12/31/2016 for patients with erythema multiforme, erythema multiforme unspecified, erythema multiforme major, Stevens Johnson syndrome, Stevens Johnson Syndrome-toxic epidermal necrosis, toxic epidermal necrosis, and other erythema multiforme.
- Patient charts with a recorded diagnosis of SJS were included and reviewed for demographics and potential factors associated with developing BO such as smoke exposure, birth history, prior oxygen use, and other significant respiratory history.
- Etiology of SJS, presence of respiratory symptoms on presentation, level and duration of respiratory support, and medications were described.
- Patients were classified as “likely developed BO,” “unlikely developed BO,” or “did not develop BO.”
- Analyses of descriptive statistics for both continuous variables (mean and SD) and categorical variables (frequency and percentage) were performed.

Results

- Of the 68 patients identified with SJS, 6 (9%) likely developed BO.
- There was no significant difference in age, sex, race, ethnicity, history of asthma, nor oxygen requirements between SJS patients who likely developed BO (SJS/BO) and unlikely/did not develop BO (SJS/No BO).
- The odds ratio of developing BO after recurrent SJS was 3.4.
- SJS patients treated with mechanical ventilation were 86% more likely to develop BO.

Conclusions

- BO is a serious and potentially fatal complication of SJS.
- Some patients may be at higher risk of developing BO following SJS and should therefore have close pulmonary follow-up.
- Patients with recurrent SJS should be closely monitored for development of BO.
- Patients who require mechanical ventilation for treatment of SJS should be closely monitored for development of BO.
- Although our data did not show an increased risk of BO development following SJS in patients with a history of asthma, it is reasonable to consider close follow up in this subset of patients as well.

Future Directions

- The distinction between Mycoplasma-induced rash and mucositis (MIRM) and SJS was proposed in 2015. We do not feel comfortable making this distinction in our patient sample, but further research on the development of BO following MIRM is needed.
- Further identification of the associations between other manifestations of SJS (i.e., other mucosal involvement) and BO development will be important to help providers more thoroughly understand the risk of BO in their patients.

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Figure 1: Expiratory image from chest computed tomography two months after Stevens Johnson Syndrome diagnosis demonstrating mosaic perfusion and vascular attenuation consistent with bronchiolitis obliterans