

# Stevens - Johnson syndrome

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Steven-Johnson syndrome (SJS) is a type IV hypersensitivity reaction that affects the skin and mucous membranes. Usually happens after an infection or exposure to medication including antiepileptics (phenytoin, carbamazepine, lamotrigine, and phenobarbital), NSAIDs, allopurinol, and some antibiotics such as trimethoprim-sulfamethoxazole . It starts as flu-like symptoms, such as productive cough, fever,

headache, malaise, and arthralgia, and progress to burning rash that appears on the face and the upper trunk. In addition, the palms, soles, dorsum of the hands, and extensor surfaces are commonly affected. This rash is characterized by erythema, edema, sloughing, blistering, ulceration, and necrosis.

Stevens-Johnsons syndrome is differentiated from toxic



**Therapeutic drug monitoring (TDM) is necessary when treating SJS because most therapy is considered narrow therapeutic index**

epidermal necrolysis (TEN) by the percentage detachment of the body surface area (BSA), as follow:

1. Stevens-Johnson syndrome: A minor form of toxic epidermal necrolysis, with less than 10% body surface area (BSA) detachment
2. Overlapping Stevens-Johnson syndrome/toxic epidermal necrolysis: Detachment of 10-30% of the BSA
3. Toxic epidermal necrolysis: Detachment of more than 30% of the BSA

SJS is considered a medical emergency that usually necessitate hospitalization. The management focuses on eliminating the underlying cause, controlling symptoms and minimizing complications as your skin regrows. Recovery can take weeks to months, depending on the severity of the condition.



# Pharmacist role in managing Stevens-Johnson syndrome

**There is no specific guidelines for managing SJS, for this reason, pharmacist have a great impact on treatment options as they are experts in medicines.**

Pharmacists have a great role in planning the management of SJS because there are no definite guidelines to guide the treatment.

We have encountered a 14 years old female who experienced SJS that was induced by the rapid titration of Lamotrigine to treat her epilepsy. First, the offending agent (lamotrigine) was discontinued. Most studies recommend starting the treatment with IVIG infusion based on body weight (1gm/kg/day) [1]. To depress the immune response, cyclophosphamide, azathioprim, and cyclosporine were used. Since cyclophosphamide can caused SJS by itself, it was recommended by most papers to use cyclosporine [4]. Cyclosporine is a narrow therapeutic index medication that involves regular TDM and regular adjustments for doses. The dose is calculated according to patient's weight (3mg/kg/day) and a trough level is obtained every 5<sup>th</sup> day. Some papers reported using systemic corticosteroids however, their use is especially

controversial. Once steroids are used, ensure using a high dose (2mg/kg/day) for short duration to avoid impairing the skin healing. In addition to the immunosuppressants, dermatological and ophthalmological involvements are very common.

Dermatological consult is a must to assess the severity of SJS as well as assess the improvements in lesions and the prognosis. Supportive therapy includes skin-substitutes dressing that should be applied on lesions, antiseptic and antibiotics cream, and moisturizers. Topical mouthwashes including antiseptics and steroids should be included as part of the treatment.



Ophthalmology consultations are necessary to due to the involvement of eye. Topical steroids, topical antibiotics, and surgery maybe indicated.

Additionally, pain management is as important as the management of SJS because patients in acute distress and pain may be difficult to manage. Since sedatives and analgesia are tailored according to patient situation, pharmacist will have major impact in decision.

Infection control is important because patients are treated with immunosuppressant and because they will have reduced immune response caused by SJS. Treatment of an infection is critical and based on the suspected organism and the pan-culture results. However, prophylactic antibiotics use is discouraged.

Anticoagulation based on kidney function and patient's characteristics is also considered.

Other systems may be involved such as kidneys and lung and treating each system involved is part of the therapy.

In conclusion, SJS is a life-threatening condition that requires an ICU stay. Pharmacists have a great role as part of the multi-disciplinary team in the management as there are no specific guidelines to be followed and literature review is needed. Additionally, SJS is mostly caused by medicines and management of comorbidities is needed too.

