

# Stevens - Johnson Syndrome in a 9 - Year - Old Child: A Case Report

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**Abstract:** *The incidence of Stevens - Johnson Syndrome in pediatric is 6.3 per 100, 000 children for SJS cases. SJS is classically associated with drug hypersensitivity reactions; however, infectious etiology is increasingly recognized as a triggering agent. The purpose of this case report is to raise awareness among clinicians about cases of SJS in children beginning with symptoms of upper respiratory tract infection. We present a 9 - year - old boy came to the hospital's emergency department, with the chief complaint of swollen and blistered lips with red spots on the chest, abdomen, and hands. The patient was clinically diagnosed as Stevens - Johnson syndrome (SJS) overlapping with upper respiratory tract infection. The success of SJS treatment is largely determined by early recognition of symptoms, stopping or overcoming the causative factors and providing adequate supportive therapy.*

**Keywords:** Stevens - Johnson Syndrome, hypersensitivity reactions, upper respiratory tract infection

## 1. Introduction

Stevens - Johnson Syndrome (SJS) is a disease mediated by an immune reaction to the body followed by a prodromal symptoms, that is the symptoms of severe mucocutaneous. SJS and its more severe form, toxic epidermal necrolysis (TEN) are the result of an inflammatory response that results in keratinocyte necrosis and perivascular lymphocytic infiltration. SJS was defined as the presence of epidermal detachment <10% of body surface area (BSA) and TEN >30% of BSA; cases with skin involvement between 10% and 30% were classified as overlapping SJS/TEN. The incidence in pediatric is 6.3 per 100, 000 children for SJS cases; 0.7 per 100, 000 children for overlapping SJS/TEN cases, and 0.5 per 100, 000 children for TEN cases. The highest incidence occurred in children aged 11 - 15 years ( $p < 0.001$ ). The highest mortality occurred in children aged 0 - 5 years and in children with TEN.<sup>3</sup>

SJS is classically associated with drug hypersensitivity reactions; however, infectious etiology is increasingly recognized as a triggering agent. SJS/TEN is induced by drugs in approximately 60% - 90% of children.<sup>2, 4, 5</sup> A limited number of drugs are responsible for the majority of cases, especially in children, even if more than 100 drugs have been associated with this disease.<sup>2</sup> Anticonvulsants, antibiotics, and nonsteroidal anti - inflammatory drugs (NSAIDs) are the more common triggers. The group of antibiotics includes erythromycin, cefotaxime, trimethoprim - sulfamethoxone, cloxacillin, and amoxicillin.<sup>2, 4</sup> In children, various pathogens, particularly *Mycoplasma pneumoniae* and Herpes virus have been found to induce SJS in 5% - 31% of cases.<sup>2, 4</sup> Infections caused by viruses (influenza, Epstein - Barr, cytomegalovirus, coxsackie, human herpes viruses 6 and 7, parvovirus), bacteria (*-haemolyticum streptococcus*, group A), mycobacteria, and rickettsiae are also associated with pediatric SJS/TEN. Infection can also act as a potential cofactor. SJS/TEN has been reported to be idiopathic in 5% - 18% of children and 25 - 50% of adults.<sup>2</sup> SJS due to infection is suspected if the symptoms of

infection precede the onset of skin or mucosal lesions and the serological diagnosis for the suspected organism is positive. Constitutional symptoms appear in the early stages followed by mucocutaneous involvement. Mucosal lesions are more common than skin lesions and commonly seen on the oral, genital, and ocular mucosal surfaces. Histopathological diagnosis of SJS is the presence of epidermal necrosis.<sup>1</sup>

The purpose of this case report is to raise awareness among clinicians about cases of SJS in children beginning with symptoms of upper respiratory tract infection.

## 2. Case Report

A 9 - year - old boy came to the hospital's emergency department, with the chief complaint of swollen and blistered lips with red spots on the chest, abdomen, and hands (**Figure 1**). The patient had symptoms of upper respiratory tract infection 4 days before admission to the hospital, ie cough, runny nose, mouth sores, and fever up to 37.8°C. The patient had a history of going to two different doctors before entering the hospital. The first treatment the patient received was paracetamol, pseudoephedrine, chlorphenamine maleate and multivitamin syrup. One day after taking these drugs, the patient's complaints of cough and fever were still accompanied by reddish spots starting to appear on the chest, abdomen, both arms and palms, the reddish spots were said to be itchy with no water came out. Furthermore, the patient received a second treatment in the form of paracetamol, dextromethorphan HBr, chlorphenamine maleate, phenylpropanolamine HCl, cefadroxil, hexetidine mouthwash, betamethasone and dexchlorpheniramine maleate. One day after patients took the treatment, patient complain of a burning sensation in the eyes and oral cavity, the lips began to blister, difficulty opening the mouth and swallowed. Patients cannot eat, drink water in small increments and feels weak. In the ER, the results of the general examination showed general weakness, moderate pain, GCS consciousness 15 compos mentis, blood

pressure 107/80 mmHg, heart rate 101/min, respiratory rate 20/min, axillary temperature 37.6°C, and body weight 18 kg. The results of physical examination showed multiple macular erythema in the thoracoabdominal region, multiple erythema palmar manus dextra et sinistra. Erosions of the upper and lower lip mucosa were seen with edema and crusting, also stomatitis. Approximately <10% skin detachment from the body surface area is involved. The patient's Nikolsky sign was positive. Complete blood count laboratory examination showed the number of leukocytes  $15.27 \times 10^3$ ; hemoglobin 13.1 g/dL; HCT 38.1%; PLT  $386 \times 10^3$ /L.

The patient was clinically diagnosed as Stevens - Johnson syndrome (SJS) overlapping with upper respiratory tract infection. Initial therapy in the form of IVFD D5 1/2 NS 20 macro drops per minute, methylprednisolone 10 mg every 12 hours intravenously, ranitidine 20 mg every 12 hours intravenously (given 15 minutes before giving methylprednisolone), aloclair mouthwash 3 times a day, vaseline oral gel is applied to the lips 3 times a day, nystatin drop 3 times a day (1 ml), and history of previous oral therapy was discontinued.

The first day of treatment, the complaints of weakness decreased, the skin efflorescence remained the same, but the patient was able to drink a little bit, still had difficulty opening his mouth. On the 3rd day of treatment, there were more white stomatitis in the mouth, the skin efflorescence remained the same and did not spread. On the 4th day of treatment the patient was able to drink and eat, received additional antibiotic therapy of gentamicin 2x40 mg IV injection. On the 6th day of treatment, the crust on the lips peeled off, eating and drinking ordinary children. The spots on the chest and hands are still there but not getting bigger. The patient was discharged from the hospital and get a return in the form of drug azithromycin syrup 1x4 ml, mouthwash 3 times a day, vaseline oral gel applied to the lips 3 times a day, nystatin drop 3 times a day as much as 1 ml. On the 10th day, follow - up was carried out on the patient's parents over the phone and they said there were no complaints, the crusts in the mouth peeled off completely, the spots on the chest and hands were dry and peeling.



**Figure 1:** Macular erythema of the thoracoabdominal region; erosion of the mucosa of the upper and lower lips with edema and crusting

### 3. Discussion

Stevens - Johnson syndrome (SJS) is a rare, potentially life - threatening, delayed - type severe mucosal hypersensitivity reaction characterized by epidermal detachment, mucosal erosions, and severe systemic symptoms that require immediate medical intervention.<sup>7</sup> Although 74 - 94% of SJS cases are associated with adverse drug reactions and responses to infection, the etiology of this pathological process is complex and not fully understood.<sup>8</sup>

SJS and TEN are characterized by blistering of the skin and mucous membranes. One to three days before the onset of skin and mucosal lesions, prodromal symptoms begin to appear, namely fever, general malaise, unproductive cough, sore eyes, and sore mouth. These symptoms are often mistaken for an upper respiratory infection.<sup>9</sup> In our case the patient had a history of cough, runny nose, mouth sores, and fever 4 days before coming to the hospital. The presence of these symptoms led us to suspect the involvement of ARI that overlapped with the patient's medical history. In children, various pathogens, particularly *Mycoplasma pneumoniae* and Herpes virus have been found to induce SJS in 5% - 31% of cases.<sup>2,4</sup>

Skin lesions have varying severity and multiple appearances of vesicles, bullae, and detachable skin necrosis.<sup>10</sup> If the primary skin lesion is erythema, the diagnosis can be based on Nikolsky's sign, though not exclusively of SJS/TEN. Nikolsky's sign is defined as detachment of the epidermis caused by the application of tangential pressure to erythematous, non - blistered skin.<sup>11</sup> In this case the patient's Nikolsky sign was positive.

All cases suspected of SJS and TEN should be confirmed by skin biopsy for histopathology and immunofluorescence examination. Early lesions show keratinocyte apoptosis in the suprabasal layer. The lesion will eventually show thick epidermal necrosis and detachment of the epidermis from the dermis. A moderate density of mononuclear cell infiltration in the dermal papilla can be seen, mostly represented by lymphocytes and macrophages.<sup>12</sup> In this case, no histopathological examination was performed on the patient. Only a complete blood count examination showed an increase in leukocytes in a patient indicating the involvement of a bacterial infection.

The success of SJS treatment is largely determined by early recognition of symptoms, stopping or overcoming the causative factors and providing adequate supportive therapy. Immediately stop the suspected drug (eliminating the drug) will reduce the mortality rate from 26% to 5%. Supportive therapy for SJS and TEN patients includes treatment in a special place (burn unit or intensive care unit) with a room temperature of more than 30°C to prevent hypothermia, administering intravenous fluids, providing adequate nutrition, using dressings and other supportive therapies such as use of eye drops, mouthwash, and use of antacids.<sup>13</sup> In this patient, the previous oral therapy was discontinued and immediately given fluid therapy in the form of IVFD D51/2 NS 20 macro drops per minute. The patient also received intravenous corticosteroids, ranitidine, mouthwash, lip balm, and nystatin drops.

Determining the prognosis of patients with SJS and TEN is important. Prognostic assessment using SCORTEN should be performed within 24 hours of the patient being admitted. The higher the SCORTEN value, the higher the risk of mortality in the patient.<sup>18</sup> Variables assessed in SCORTEN include age, extent of epidermal necrolysis, heart rate, blood urea nitrogen (BUN), random blood sugar, and serum bicarbonate. A SCORTEN assessment could not be performed in this patient because serum bicarbonate, BUN, and random blood sugar tests were not performed so that SCORTEN could not be evaluated.<sup>14</sup> In non-severe cases the prognosis is good and healing occurs within 2 - 3 weeks. In severe cases with multiple complications or with delayed and inadequate treatment, the mortality rate ranges from 5 - 15%.<sup>15</sup> In this case, the patient improved clinically and was discharged home after 6 days of hospitalization.

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