

Stevens-Johnson Syndrome: Case Report and Literature Review

Ángel de Jesús Ramos Fuentes¹, Alejandra Mariel Ceja Vallejo², Paola Gonzalez Castelan ³, Miriam Aylin Matus⁴, Arleth Castillo Aguilar⁵, María de Lourdes Toto Márquez⁶, Samara Estefanía Loya Hernández⁷, Gloria Montserrat Sierra Torres⁸, Diana Rubi Velarde Flores ⁹, Andres Salvador Prado Alanis¹⁰, Carlos Eduardo Garduño Corbella⁹

¹Universidad Veracruzana

²Universidad Vasco de Quiroga

³Universidad Autónoma de Tlaxcala Facultad de Ciencias de la Salud.

⁴Universidad Autónoma de Tamaulipas

⁵Hospital General de Cuernavaca Dr. José G Parres

⁶HGZ1 Zacatecas, Zac. (IMSS)

⁷Universidad Nacional Autónoma de México

⁸ISSSTE Hospital General Santiago Ramón y Cajal

⁹Universidad Michoacana de San Nicolás de Hidalgo

¹⁰Universidad Autónoma de Durango

ABSTRACT

Stevens-Johnson Syndrome is an acute and potentially life-threatening mucocutaneous disease characterized by extensive epidermal necrosis and severe mucosal involvement, usually associated with drug-induced hypersensitivity reactions. We present the case of a 9-year-old female patient with a recent history of treatment with trimethoprim/sulfamethoxazole for a respiratory infection, who initially developed fever, malaise, odynophagia, and bilateral conjunctivitis, subsequently progressing to painful erythematous bullous lesions with epidermal detachment and oral and ocular involvement. Physical examination revealed extensive skin lesions on the dorsal region and upper extremities with a positive Nikolsky sign, involving approximately 12% of the total body surface area. Laboratory studies showed leukocytosis and elevated acute phase reactants. Due to the clinical suspicion of Stevens-Johnson syndrome secondary to a drug reaction, the antibiotic was immediately discontinued and intensive multidisciplinary management was initiated, including fluid replacement, analgesia, advanced wound care, systemic corticosteroids, and intravenous immunoglobulin therapy. In addition, a skin biopsy and histopathological study were performed, reporting full-thickness epidermal necrosis and basal vacuolar degeneration, findings compatible with Stevens-Johnson syndrome. Clinical evolution was favorable, with progressive reepithelialization and improvement of skin lesions without severe systemic complications. This case highlights the importance of early recognition, immediate withdrawal of the causative agent, and timely multidisciplinary management in order to reduce morbidity and mortality and prevent sequelae associated with this highly complex dermatological entity.

KEYWORDS: Stevens-Johnson syndrome, toxic epidermal necrolysis, drug eruption, drug hypersensitivity, epidermal necrosis, bullous lesions, mucocutaneous involvement, trimethoprim/sulfamethoxazole, pediatric dermatology, histopathological study.

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INTRODUCTION

Stevens-Johnson Syndrome is a rare, acute, and potentially life-threatening mucocutaneous disorder characterized by

extensive epidermal necrosis and skin detachment associated with severe mucosal involvement. It forms part of a clinical spectrum together with toxic epidermal necrolysis, differing

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mainly according to the percentage of affected body surface area. Stevens-Johnson syndrome is classically defined by involvement of less than 10% of the total body surface area, whereas toxic epidermal necrolysis involves more than 30%, with overlap forms also described between both entities.¹

Although its annual incidence is low, it represents one of the most severe dermatological emergencies due to its high morbidity and mortality and the complexity of hospital management. Mortality rates range between 5% and 15% in Stevens-Johnson syndrome and may exceed 30% in cases of toxic epidermal necrolysis, particularly in elderly patients, those with comorbidities, or extensive systemic involvement. Prognosis depends on multiple factors, including the extent of skin involvement, the timing of withdrawal of the causative agent, and the presence of infectious or respiratory complications.²

The pathophysiology of Stevens-Johnson syndrome involves an immune-mediated response mainly driven by cytotoxic T lymphocytes, release of granulysin, perforins, and several inflammatory cytokines that induce massive keratinocyte apoptosis. This mechanism leads to progressive epidermal necrosis and dermoepidermal separation, resulting in extensive skin lesions similar to severe burns. Furthermore, genetic associations with certain major histocompatibility complex alleles have been demonstrated, particularly in specific populations exposed to certain medications.³

Drugs represent the main triggering factors and are responsible for the majority of reported cases. The medications most frequently implicated include anticonvulsants such as carbamazepine, phenytoin, and lamotrigine; antibiotics such as sulfonamides, penicillins, and quinolones; nonsteroidal anti-inflammatory drugs; allopurinol; and some antiretroviral agents. Nevertheless, cases associated with viral and bacterial infections have also been described, especially those related to *Mycoplasma pneumoniae*, herpes simplex virus, and respiratory viruses.⁴

The clinical presentation usually begins with nonspecific prodromal symptoms such as fever, malaise, headache, odynophagia, and conjunctivitis, resembling an acute viral infection. Subsequently, painful erythematous skin lesions, purpuric macules, and flaccid blisters appear and rapidly progress to epidermal necrosis and detachment. Mucosal involvement occurs in more than 90% of patients, mainly affecting the oral cavity, ocular conjunctiva, and genital region. Nikolsky sign is frequently positive and reflects the characteristic epidermal fragility of the disease.⁵

Diagnosis is predominantly clinical, although it may be complemented by skin biopsy and histopathological examination, where characteristic findings include full-thickness epidermal necrosis, basal vacuolar degeneration, and sparse dermal inflammatory infiltrate. Early recognition of the disease is essential because immediate withdrawal of

the causative agent constitutes the most important therapeutic measure and is directly associated with decreased mortality.⁶ Management of Stevens-Johnson syndrome requires a multidisciplinary approach in specialized hospital units, frequently similar to the management of major burn patients. Treatment includes strict fluid and electrolyte balance control, nutritional support, analgesia, prevention of secondary infections, advanced wound care, and prompt ophthalmologic management. Several immunomodulatory therapies such as intravenous immunoglobulin, cyclosporine, and systemic corticosteroids have been used, although their efficacy remains controversial in the literature.⁷

Despite therapeutic advances, long-term sequelae continue to represent a significant problem. Ophthalmologic complications may include symblepharon, severe dry eye, corneal ulceration, and permanent visual loss. Likewise, cicatricial skin changes, mucosal dysfunction, and psychological repercussions significantly affect the quality of life of surviving patients.⁸

Due to the low frequency of this disease and the complexity of its clinical and reconstructive management, case reports remain relevant to broaden knowledge regarding its evolution, complications, and therapeutic options. We present the case of a patient with Stevens-Johnson syndrome, describing the clinical findings, diagnostic approach, multidisciplinary treatment, and hospital evolution, accompanied by a current review of the literature related to this highly complex dermatological entity.

CASE REPORT

A 9-year-old female patient without significant family medical history and with a recent history of upper respiratory tract infection treated with trimethoprim/sulfamethoxazole seven days prior to symptom onset presented with fever of 38.5 °C, malaise, odynophagia, and bilateral conjunctivitis, subsequently associated with the appearance of painful erythematous lesions on the dorsal region and thorax, which rapidly progressed to violaceous plaques, flaccid blisters, and areas of epidermal detachment.

During the following 48 hours, the patient experienced progressive worsening of the skin lesions accompanied by severe pain, feeding difficulty, and ulcerated lesions involving the oral mucosa. She was brought to the emergency department due to clinical deterioration and the appearance of extensive cutaneous desquamation. Physical examination revealed an alert but irritable patient with signs of moderate dehydration and multiple erythematous bullous lesions involving the back, shoulders, and lumbar region, with epidermal detachment affecting approximately 12% of the total body surface area. Nikolsky sign was positive. Oral examination showed hemorrhagic erosions and painful crusted lips, while ophthalmologic evaluation demonstrated bilateral conjunctival hyperemia.



Figure 1: Initial clinical image

Laboratory studies revealed leukocytosis of 15,200 cells/mm³, elevated C-reactive protein levels, and mild electrolyte disturbances secondary to cutaneous fluid loss and reduced oral intake. Due to the clinical suspicion of Stevens-Johnson syndrome overlapping with toxic epidermal necrolysis secondary to a drug-induced reaction, the antibiotic was immediately discontinued and multidisciplinary management was initiated by the dermatology, pediatrics, ophthalmology, and plastic surgery departments.

The patient was admitted to the pediatric intermediate care unit, where intravenous fluid replacement, multimodal analgesia, nutritional support, and advanced wound care with

daily saline irrigation and nonadherent dressings were initiated. Systemic corticosteroids and intravenous immunoglobulin therapy were administered, resulting in progressive stabilization of the skin lesions. Intensive ocular lubrication was also indicated to prevent ophthalmologic sequelae.

In order to confirm the diagnosis, a skin biopsy and histopathological examination of a lesion from the dorsal region were performed, revealing full-thickness epidermal necrosis, vacuolar degeneration of the basal layer, and sparse dermal inflammatory infiltrate, findings compatible with Stevens-Johnson syndrome/toxic epidermal necrolysis.

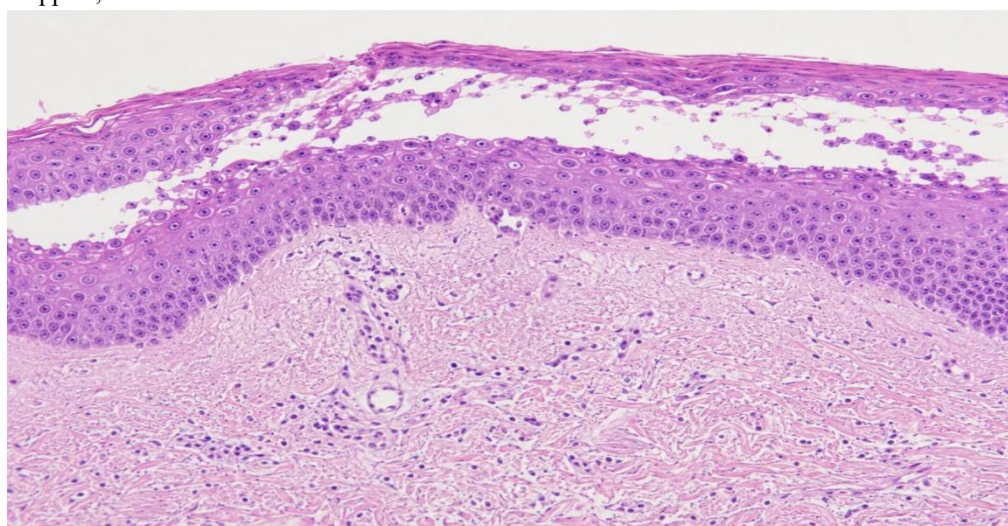


Figure 2: Histopathological study

Clinical evolution was favorable, with progressive reduction of lesions and partial reepithelialization following treatment initiation. After 21 days of hospitalization, the patient was discharged with satisfactory clinical improvement, presenting

only residual areas of cutaneous hyperpigmentation without major functional sequelae, and continued outpatient follow-up by dermatology and ophthalmology services.



Figure 3: Clinical image 21 days after hospitalization

DISCUSSION

Stevens-Johnson Syndrome represents one of the most severe mucocutaneous reactions within the spectrum of drug hypersensitivity disorders, characterized by extensive epidermal necrosis and significant mucosal involvement. Despite its low incidence, it constitutes a dermatological emergency with high morbidity and mortality due to rapid clinical progression and the risk of severe systemic complications. Its presentation in the pediatric population is less frequent than in adults; however, it may be associated with aggressive evolution and require intensive multidisciplinary management.⁹ In the present case, the patient developed progressive cutaneous lesions following the use of trimethoprim/sulfamethoxazole, one of the medications most

frequently associated with Stevens-Johnson syndrome and toxic epidermal necrolysis. The temporal relationship between the initiation of pharmacological treatment and the appearance of mucocutaneous lesions allowed early suspicion of a severe adverse drug reaction, favoring immediate withdrawal of the causative agent and prompt initiation of hospital treatment.¹⁰ The pathophysiology of Stevens-Johnson syndrome involves an immune-mediated response mainly driven by cytotoxic T lymphocytes and the release of inflammatory mediators capable of inducing massive keratinocyte apoptosis. This mechanism results in dermoepidermal separation and loss of the skin barrier, causing alterations similar to those observed in burn patients.¹¹

Table 1. Medications most frequently associated with Stevens-Johnson syndrome

Pharmacological group	Common medications
Antibiotics	Sulfonamides, penicillins, quinolones
Anticonvulsants	Carbamazepine, phenytoin, lamotrigine
Nonsteroidal anti-inflammatory drugs	Oxicams, ibuprofen
Antigout agents	Allopurinol
Antiretrovirals	Nevirapine

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The clinical presentation usually begins with nonspecific prodromal symptoms, including fever, malaise, and upper respiratory symptoms, subsequently progressing to painful skin lesions and mucosal involvement. In our patient, the presence of extensive bullous lesions, positive Nikolsky sign, and oral and ocular involvement were characteristic findings of advanced disease.¹²

Mucosal involvement occurs in more than 90% of patients and constitutes one of the main factors associated with morbidity. Ophthalmologic complications represent one of the most significant sequelae and may range from mild conjunctivitis to corneal ulceration and permanent visual loss.¹³

Table 2. Common clinical manifestations in Stevens-Johnson syndrome

Clinical manifestation	Approximate frequency
Fever	80–90%
Painful skin lesions	>90%
Oral involvement	70–100%
Ocular involvement	50–80%
Genital involvement	20–60%
Positive Nikolsky sign	Frequent

Diagnosis is predominantly clinical; however, histopathological examination remains an important tool for confirming the disease and excluding other bullous disorders.

In the present case, a skin biopsy was performed, revealing full-thickness epidermal necrosis and basal vacuolar degeneration, classic findings described in the literature.¹³

Table 3. Histopathological findings in Stevens-Johnson syndrome

Histopathological finding	Characteristic
Epidermal necrosis	Extensive and full thickness
Basal vacuolar degeneration	Frequent
Dermoepidermal separation	Present
Dermal inflammatory infiltrate	Sparse
Keratinocyte apoptosis	Marked

Management of Stevens-Johnson syndrome requires multidisciplinary treatment and care similar to that used in burn patients. Immediate discontinuation of the triggering drug represents the most important therapeutic measure and is associated with improved prognosis. Fluid support, pain management, prevention of infections, and specialized wound care constitute fundamental pillars of treatment.¹⁴

Several immunomodulatory therapies have been used in patients with severe disease, including systemic corticosteroids, intravenous immunoglobulin, and cyclosporine; however, evidence remains controversial regarding which strategy provides the best clinical outcomes.¹⁴

Table 4. Therapeutic options used in Stevens-Johnson syndrome

Treatment	Objective
Discontinuation of the causative drug	Stop disease progression
Fluid replacement	Maintain hemodynamic stability
Advanced wound care	Promote reepithelialization
Systemic corticosteroids	Reduce inflammatory response
Intravenous immunoglobulin	Inhibit epidermal apoptosis
Ocular lubrication	Prevent ophthalmologic sequelae

In the present case, the patient showed favorable clinical evolution after the established management, with progressive reepithelialization and absence of severe infectious complications. Early intervention and multidisciplinary

management probably contributed significantly to the favorable clinical outcome.¹⁵

Despite clinical recovery, surviving patients may develop long-term cutaneous, ophthalmologic, and psychological sequelae, making continuous outpatient follow-up essential.¹⁵

Table 5. Complications and sequelae associated with Stevens-Johnson syndrome

Complication	Consequence
Cutaneous superinfection	Sepsis
Dehydration	Electrolyte disturbances
Ocular involvement	Scarring and visual loss
Respiratory alterations	Respiratory failure
Cutaneous sequelae	Hyperpigmentation and scars
Psychological impact	Anxiety and depression

CONCLUSION

In conclusion, Stevens-Johnson Syndrome represents a highly complex and potentially life-threatening dermatological emergency that requires early diagnosis and timely multidisciplinary management in order to reduce morbidity and mortality and prevent permanent sequelae. The rapid progression of skin lesions and mucosal involvement requires maintaining a high index of clinical suspicion, especially in patients with recent exposure to high-risk medications.

In the present case, appropriate clinical correlation, immediate discontinuation of the triggering drug, and intensive treatment allowed favorable patient evolution, achieving clinical stabilization and progressive reepithelialization of skin lesions without severe systemic complications. Furthermore, histopathological examination confirmed the diagnosis, demonstrating characteristic findings of extensive epidermal necrosis and dermoepidermal junction damage.

Hospital management based on fluid support, pain control, advanced wound care, and prevention of ophthalmologic sequelae remains the cornerstone of treatment. Although several immunomodulatory therapies have demonstrated usefulness in some patients, controversy still exists regarding the optimal pharmacological treatment; therefore, management should be individualized according to clinical severity and patient evolution.

This report highlights the importance of early recognition of severe drug-induced cutaneous reactions in the pediatric population, as well as the need for comprehensive management and long-term follow-up to detect possible cutaneous, functional, and psychological sequelae. Finally, case reports continue to be relevant for expanding clinical and therapeutic knowledge regarding this uncommon but highly severe disease.

REFERENCES

- I. Harr, T., & French, L. E. (2010). Stevens-Johnson syndrome and toxic epidermal necrolysis. *Orphanet Journal of Rare Diseases*, 5(39), 1–11. <https://doi.org/10.1186/1750-1172-5-39>
- II. Creamer, D., Walsh, S. A., Dziewulski, P., Exton, L. S., Lee, H. Y., Dart, J. K. G., Setterfield, J., Bunker, C. B., Arden-Jones, M. R., Watson, K. M., Wong,

G. A., Philippidou, M., Vercueil, A., Martin, R. V., & du Vivier, A. W. P. (2016). U.K. guidelines for the management of Stevens-Johnson syndrome/toxic epidermal necrolysis in adults 2016. *British Journal of Dermatology*, 174(6), 1194–1227.

<https://doi.org/10.1111/bjd.14530>

- III. Schwartz, R. A., & McDonough, P. H. (2013). Toxic epidermal necrolysis: Part I. Introduction, history, classification, clinical features, systemic manifestations, etiology, and immunopathogenesis. *Journal of the American Academy of Dermatology*, 69(2), 173.e1–173.e13. <https://doi.org/10.1016/j.jaad.2013.05.003>
- IV. Schwartz, R. A., & McDonough, P. H. (2013). Toxic epidermal necrolysis: Part II. Prognosis, sequelae, diagnosis, differential diagnosis, prevention, and treatment. *Journal of the American Academy of Dermatology*, 69(2), 187.e1–187.e16. <https://doi.org/10.1016/j.jaad.2013.05.002>
- V. Roujeau, J. C., Stern, R. S., & Severe Adverse Cutaneous Reactions to Drugs. (1994). *Severe adverse cutaneous reactions to drugs*. *New England Journal of Medicine*, 331(19), 1272–1285. <https://doi.org/10.1056/NEJM19941103311906>
- VI. Schneider, J. A., & Cohen, P. R. (2017). Stevens-Johnson syndrome and toxic epidermal necrolysis: A concise review with a comprehensive summary of therapeutic interventions emphasizing supportive measures. *Advances in Therapy*, 34(6), 1235–1244. <https://doi.org/10.1007/s12325-017-0530-y>
- VII. Lerch, M., Mainetti, C., Terziroli Beretta-Piccoli, B., & Harr, T. (2018). Current perspectives on Stevens-Johnson syndrome and toxic epidermal necrolysis. *Clinical Reviews in Allergy & Immunology*, 54(1), 147–176. <https://doi.org/10.1007/s12016-017-8654-z>
- VIII. Hsu, D. Y., Brieve, J., Silverberg, N. B., Paller, A. S., & Silverberg, J. I. (2017). Stevens-Johnson syndrome and toxic epidermal necrolysis in children. *Journal of the American Academy of Dermatology*, 76(5), 811–817. <https://doi.org/10.1016/j.jaad.2016.11.042>
- IX. Bastuji-Garin, S., Fouchard, N., Bertocchi, M., Roujeau, J. C., Revuz, J., & Wolkenstein, P. (2000).

- SCORTEN: A severity-of-illness score for toxic epidermal necrolysis. *Journal of Investigative Dermatology*, 115(2), 149–153. <https://doi.org/10.1046/j.1523-1747.2000.00061.x>
- X. Mockenhaupt, M. (2011). The current understanding of Stevens-Johnson syndrome and toxic epidermal necrolysis. *Expert Review of Clinical Immunology*, 7(6), 803–815. <https://doi.org/10.1586/eci.11.66>
- XI. Sekula, P., Dunant, A., Mockenhaupt, M., Naldi, L., Bouwes Bavinck, J. N., Halevy, S., Kardaun, S. H., Sidoroff, A., Liss, Y., Schumacher, M., Roujeau, J. C., & RegiSCAR Study Group. (2013). Comprehensive survival analysis of a cohort of patients with Stevens-Johnson syndrome and toxic epidermal necrolysis. *Journal of Investigative Dermatology*, 133(5), 1197–1204. <https://doi.org/10.1038/jid.2012.510>
- XII. Sotozono, C., Ueta, M., Koizumi, N., Inatomi, T., Shirakata, Y., Ikezawa, Z., & Kinoshita, S. (2009). Diagnosis and treatment of Stevens-Johnson syndrome and toxic epidermal necrolysis with ocular complications. *Ophthalmology*, 116(4), 685–690. <https://doi.org/10.1016/j.ophtha.2008.10.048>
- XIII. Yang, M. S., Lee, J. Y., Kim, J., Kim, G. W., Kim, B. K., Kim, J. Y., Park, H. W., Cho, S. H., & Min, K. U. (2016). Incidence of Stevens-Johnson syndrome and toxic epidermal necrolysis: A nationwide population-based study using National Health Insurance Database in Korea. *PLoS ONE*, 11(11), e0165933. <https://doi.org/10.1371/journal.pone.0165933>
- XIV. Auquier-Dunant, A., Mockenhaupt, M., Naldi, L., Correia, O., Schröder, W., & Roujeau, J. C. (2002). Correlations between clinical patterns and causes of erythema multiforme majus, Stevens-Johnson syndrome, and toxic epidermal necrolysis. *Archives of Dermatology*, 138(8), 1019–1024. <https://doi.org/10.1001/archderm.138.8.1019>
- XV. De Risi-Pugliese, T., & Wolkenstein, P. (2019). Stevens-Johnson syndrome and toxic epidermal necrolysis: Diagnosis, clinical manifestations, and management. *Clinics in Dermatology*, 38(6), 702–712. <https://doi.org/10.1016/j.clindermatol.2020.06.006>