**INTRODUCTION**

Stevens-Johnson syndrome (SJS) and toxic epidermal necrolysis (TEN, Lyell's disease) are severe, episodic, acute mucocutaneous reactions that may be caused by various factors particularly drugs. Treatment is primarily supportive and there are no specific therapy regimens. Intravenous immunoglobulin (IVIG) has recently been shown to be a useful and safe therapy in paediatric patients with SJS/TEN, though no such case has so far been reported from Pakistan. The authors report the experience with low dose IVIG (0.1 g/kg/day for four consecutive days for treating a 3 years old boy with toxic epidermal necrosis with favourable outcome without early complications.

**CASE REPORT**

A 3 years old boy was admitted with 5 days history of fever and 3 days history of skin eruption. The patient had received some medications from some local paramedic on second day of his illness, the nature of which could not be ascertained. Two days after the onset of fever, the patient developed painful lesions in the mouth and developed dysphagia. This was accompanied by dysuria, cough, photophobia and a painful skin eruption that appeared on the face and rapidly progressed to involve the upper trunk and extremities. Physical examination revealed an acutely ill child, with a temperature of 38.5°C, pulse rate of 126 per minute and respiratory rate of 20 per minute. The skin rash consisted of multiple targetoid lesions, and erosions discrete at places and confluent at others, scattered all over the body with peeling of skin over neck. A few intact blisters were also present on his limbs and upper back. Mucous membrane examination revealed bilateral, non-purulent conjunctivitis, extensive hemorrhagic erosions and crusting of lips, erosions of tongue, hard palate and buccal mucosa. The clinical diagnosis of drug-induced toxic epidermal necrosis (TEN) was made (Figures 1, 2). The authors describe their experience in a 3 years old boy with TEN, in whom low dose IVIG was used successfully.

**ABSTRACT**

Stevens-Johnson syndrome (SJS) and toxic epidermal necrolysis (TEN, Lyell's disease) are severe, episodic, acute mucocutaneous reactions that may be caused by various factors particularly drugs. Treatment is primarily supportive care and there are no specific therapy regimens. Intravenous immunoglobulin (IVIG) has recently been shown to be a useful and safe therapy in paediatric patients with SJS/TEN, though no such case has so far been reported from Pakistan. The authors report the experience with low dose IVIG (0.1 g/kg/day for four consecutive days for treating a 3 years old boy with toxic epidermal necrosis with favourable outcome without early complications.

**Key words:** Stevens-Johnson syndrome. Toxic epidermal necrolysis. Intravenous immunoglobulin. Paediatrics.

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**Figure 1:** Patient showing multiple targetoid lesions and erosions scattered all over the body, discrete at places and confluent at others, on day 1 of hospitalization.

**Figure 2:** Close up of the lesions on day 1 of hospitalization.
Intravenous immunoglobulin (IVIG) is a highly purified IgG prepared from pooled human plasma. It consists mainly of unmodified IgG (more than 95%) with trace amounts of IgA or IgM.²,⁴ It is available as sterile solution of 4.5-5.5% human protein in 9-11% maltose having no preservatives. It is to be administered at an initial rate of 0.01-0.02 ml/kg body weight/minute for initial 30 minutes. If it is tolerated well, the rate may be gradually increased to maximum of 0.08 ml/kg body weight/minute. It may be diluted with 5% dextrose solution.⁵ Side effects from IVIG occur in < 5% of cases and are generally mild and self-limiting.⁶

Viard et al. in a landmark study used IVIG in 10 consecutive individuals with TEN and demonstrated a favourable outcome without significant adverse effects.⁴ A number of retrospective and prospective studies have since been published reporting varying levels of evidence for the efficacy of IVIG.⁵,⁷-¹⁰

However, in paediatric patients with SJS/TEN, IVIG seems to be a useful and safe therapy.⁵,⁷,¹⁰ Metry et al. reviewed 28 previous reports in which IVIG was used in paediatric patients with SJS/TEN and discussed their experience in 7 children with SJS. They concluded that IVIG (0.5-1.0 g/kg/day for 3 days) was a useful and safe therapy for children with SJS/TEN,⁹ Tristani-Firouzi et al., in a retrospective study in paediatric population, reported favourable results with zero mortality at dose of 0.5-0.75 g/kg/day for 4 days in 8 children having TEN.¹⁰ These studies have examined the efficacy and safety of high dose IVIG.⁴,⁷-¹⁰ whereas Mangla et al. were the first to administer low dose IVIG (0.05-0.1 gm/kg/day for 5 consecutive days) to 10 paediatric patients with TEN and reported favourable results.⁵

No paediatric patient of SJS/TEN has so far been reportedly treated with IVIG in Pakistan. The major factors limiting widespread use of IVIG for SJS/TEN and related disorders in resource-poor countries like Pakistan are cost and availability. Under these circumstances, it becomes all the more important to find out cost effective approaches to provide health care facilities. Keeping that in mind, low dose IVIG (0.1 g/kg/day for 4 consecutive days) was prescribed in this patient with favourable outcome. However, further studies are required to find out the minimum effective dosage of IVIG and its safety for treatment SJS/TEN.

REFERENCES


