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Chronic Ocular Sequelae of Stevens-Johnson Syndrome in Children: Long-term Impact of Appropriate Therapy on Natural History of Disease

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ABSTRACT

Purpose: To describe the long-term ocular and visual morbidity in children with chronic sequelae of Stevens-Johnson Syndrome (SJS) and visual outcomes of various management strategies.

Design: Retrospective comparative case series.

Methods: This study included 568 eyes of 284 children with SJS who presented between 1990 and 2015. Affected eyes either received conservative therapy (n=440) or definitive management (n=128) including lid margin mucous membrane grafting (MMG), prosthetic replacement of the ocular surface ecosystem (PROSE) contact lenses, allogeneic limbal transplantation or keratoprosthesis using an algorithmic approach based on the severity of dryness and cause and extent of corneal damage. The primary outcome measure was best corrected visual acuity (BCVA).

Results: Two-thirds of patients presented more than a year after acute SJS, 99% without prior amniotic membrane grafting, with low-vision or blindness in 60% of eyes. Children 8-years or younger in age had significantly worse ocular and visual morbidity ($P \leq 0.037$). At 5-years of follow-up, definitive therapy significantly altered the natural history of the disease by improving BCVA and preventing the development or progression of keratopathy, as compared to conservative therapy ($P \leq 0.002$). In eyes with lid-related keratopathy, MMG was significantly more effective than PROSE, although both were significantly better than conservative therapy and the combination of MMG followed by PROSE provided the best results ($P < 0.0001$).

Conclusion: Children receiving sub-optimal care during acute SJS presented later with severe ocular and visual morbidity. Timely therapy, particularly with PROSE and MMG in eyes with lid-related keratopathy changed the natural course and helped in preserving and improving vision.