AB078. Ocular involvement in epidermolysis bullosa

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Background: Epidermolysis bullosa (EB) is a heterogenous group of skin disorders characterized by formation of blisters and erosions of the skin in response to minor trauma. Subtypes include EB simplex (EBS), junctional EB (JEB), dystrophic form of EB (DEB) and finally Kindler syndrome (KS). In addition to dermal manifestation, patients can present with various ophthalmic pathologies.

Methods: We reviewed the pathobiology, epidemiology and management of ocular manifestations as well as current and future innovative therapies for EB.

Results: The severity and incidence of ocular involvement were the highest in the recessive DEB-generalized severe and JEB-generalized severe subtypes. Recurrent corneal erosions and blisters were the most common finding and seem to correlate with skin disease. Other manifestations include corneal scaring, blepharitis, ectropion, symblepharon, infantile cataracts, lacrimal duct obstruction as well as meibomian gland deficiency.

Conclusions: Ophthalmology consult as well as regular follow-up are essential in the multi-disciplinary approach of this disease. Indeed, parents’ and patients’ education as well as early diagnosis and treatment are crucial to prevent permanent and long-term visual disabilities.

Keywords: Review; epidermolysis bullosa (EB); eye; cornea

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