GENETICS AND GENODERMATOSES

ALOPECIA IN EPIDERMOLYSIS BULLOSA: A SYSTEMATIC REVIEW OF PATHOGENESIS AND CLINICAL FEATURES OF DISEASE

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Introduction: Epidermolysis bullosa (EB) is a group of rare genetic skin diseases characterised by the gene mutations encoding adhesion proteins within the skin. These adhesion proteins are also present in normal hair follicles. Anecdotally, there have been reports of scalp alopecia as a complication of EB and there are scattered cases in the literature, but alopecia has generally been overlooked in severe blistering diseases because it is regarded as a cosmetic issue. Therefore, there is no consensus about the natural history and clinical manifestations of alopecia in EB to allow potential intervention.

Objective: To review the current literature detailing the pathogenesis and clinical presentations of alopecia in EB patients.

Materials and Methods: Relevant studies were searched in Medline, PubMed and EMBASE electronic databases up to September 2018, and included according to clinical presentations of disease in humans.

Results: Only 15 cases detailed 29 EB patients with demographic and clinical manifestations of alopecia. Vertical biopsy sections were the most common method of alopecia diagnosis, and the most common pattern was patchy scalp alopecia (45%) followed by diffuse alopecia (41%). The most robust finding was nonspecific scarring alopecia in all dystrophic EB (DEB) patients and nonspecific nonscarring alopecia in most patients with EB Simplex (EBS).

Conclusion: Hair abnormalities observed in EB are of variable severity despite there being no universal validated alopecia scoring system, with alopecia occurring secondary to blistering, or in areas prone to friction.