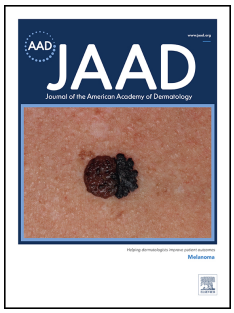

Oral status in patients with inherited epidermolysis bullosa: a multicentric observational study.

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Title page

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Research letter

Inherited Epidermolysis bullosa (EB) represents a group of genodermatoses characterized by skin and mucosal fragility leading to blistering and erosions [1]. We performed a comprehensive assessment of oral health in EB patients, in order to optimize their care.

After institutional approval (NCT04217538), an observational multicenter study was conducted in three EB expert centers, in France (Nice and Toulouse) and Belgium (Leuven), between 2017 and 2019. The main objective of this study was to compare the oral health status of Dystrophic (DEB), Junctional (JEB) or Simplex EB (SEB) patients with an age/gender - matched control group. Practitioners involved in the clinical examinations were specialists in oral pathology and/or pediatric dentistry.

Forty-two patients (mean age 13 years [range 2 to 78]) with EB (25 dystrophic, 12 simplex and 5 junctional) and 42 healthy controls were included. Overall, individuals with DEB and JEB were most severely affected by mucosal blisters, erythema and erosions/ulcerations, which is consistent with the greater fragility of their mucosa (Table I). The localization of oral lesions depended on EB type: in SEB patients, the oral floor was never affected, while in DEB patients the lesions were mainly seen on the inner cheek and palate (80% and 76%) and in JEB patients, on the lips (40%) and oral floor (40%).

The evaluation of the oral hygiene of EB patients showed a 1.5 times greater dental plaque accumulation than in controls (Plaque Index (PI): 1.7 ± 0.7 vs 1.1 ± 1.0 ; p=0.004) while the prevalence of caries was comparable in both groups [2,3,4], probably because of their regular dental follow-up during annual/semestrial medical check-ups.

However, gingival inflammation (characterized by a high gingival index (GI) score and bleeding when brushing), usually associated with excessive dental plaque accumulation [5], deviated from this pattern in DEB participants. They displayed a slight-to-moderate PI score

despite high GI (1.5 ± 0.8 vs 0.4 ± 0.6; $p < 0.001$) and frequent/strong "bleeding when brushing". Therefore, the accumulation of dental plaque cannot explain on its own this enhanced inflammatory reaction. With increasing severity, clinical features become quite different from plaque-induced gingivitis of equal severity: the erythematous area is wider and the swelling more diffuse, extending over the whole attached gingiva, with the free gingiva appearing ulcerated (Fig. 1). These findings suggest that this type of gingival inflammation could be a specific feature of DEB reflecting the intrinsic fragility of the gingival tissue. Indeed, gingivitis extent, severity and progression are known to be affected by genetic mutations that underlie changes in the organization of periodontal tissues [5]. Gingival inflammation is a serious condition that needs to be taken into consideration in the management of these patients, as it may evolve to periodontitis and loss of teeth.

Overall, we showed that gingival inflammation represents a phenotypic trait of dystrophic EB and that oral mucosa lesion localization depends on the type of EB. Joint care by dermatologist and dentist and close dental follow-up are needed to protect the oral health of patients with EB.

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Table and figure

Table I. Oral mucosal lesions in individuals with epidermolysis bullosa (EB).

	Total EB N=42	*DEB N=25*	*JEB N=5*	*SEB N=12*
Blister	19 (45.2%)	12 (48.0%)	3 (60.0%)	4 (33.3%)
Erythema	14 (33.3%)	11 (44.0%)	2 (40.0%)	1 (8.3%)
Erosion/ulceration	15 (35.7%)	10 (40.0%)	2 (40.0%)	3 (25.0%)
At least one lesion	32 (76.2%)	23 (92.0%)	4 (80.0%)	5 (41.7%)
No lesion	10 (23.8%)	2 (8.0%)	1 (20.0%)	7 (58.3%)

DEB, dystrophic epidermolysis bullosa; JEB, junctional epidermolysis bullosa; SEB, simplex epidermolysis bullosa.

Fig. 1. Intra-oral photograph of an 18-year-old patient with dystrophic epidermolysis bullosa showing large deposits of dental plaque leading to severe gingivitis with a wide area of erythema and ulceration of the gingiva, totally lifted from the tooth surface (black arrow). Microstomia and ankyloglossia were present.