

## DENTISTRY APPROACH OF EPIDERMOLYSIS BULLOSA: TWO CASE REPORTS

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### Abstract

Epidermolysis bullosa (EB) represents a group of genetically determined disorders characterized by development of blisters after minimal mechanical trauma or friction of the skin or mucosa with multiple oral manifestations.

In the following we presented oral manifestations and therapy of two case reports with EB. Firstly we describe a 24 years-old caucasian female with diagnosis of epidermolysis bullosa, presence of vesiculobullous lesions on the hands and oral cavity. Secondly a 55 years-old caucasian female with diagnosis of epidermolysis bullosa, presence of vesiculobullous lesions on the body and oral papilloma.

In conclusion patients with Epidermolysis bullosa (EB) can and should receive dental care, independent of the EB subtype, and prevention programs and oral diagnosis have a great effectiveness.

*Case report (J Int Dent Med Res 2013; 6: (3), pp. 109-112)*

**Keywords:** Epidermolysis bullosa.

**Received date:** 16 November 2013

**Accept date:** 26 November 2013

### Introduction

Epidermolysis bullosa (EB) represents a group of infrequent genetically skin disorders characterized by development of blisters in the region cutaneous and mucosal throughout the body<sup>1</sup>. Physiopathologically is caused by anomalous adhesion of epithelial components and occurs at different levels, depending on the subtype of EB. Due to the lack of cohesion and mechanical fragility, blisters form following minimal trauma or friction<sup>1,2</sup>.

Nowadays epidermolysis bullosa is classified into four major groups of skin disease: epidermolysis simplex, junctional, dystrophic and a mixed type (Kindler syndrome), based on distinct differences in

the ultrastructural level within which blisters develop in affected tissues<sup>3,4</sup>.

Oral manifestations include repeated occurrence of blisters, erosions and scars, which lead to limited mouth opening, ankyloglossia and elimination of buccal sulci. Severe periodontal disease and early loss of dentition are frequently observed, such as alveolar bone resorption and jaw atrophy. Routine dental treatment as well as methods of oral hygiene can result in formation of bubbles in the oral mucosa<sup>5,6</sup>.

The dental treatment of the patient with epidermolysis bullosa (EB), in particularly for those with the most severe has changed dramatically in the last 30 years<sup>7</sup>.

Actually researchers showed that caries and periodontal disease can be successfully prevented in patients with EB by continuous follow-up and oral hygiene habits<sup>8</sup> and reports published recently demonstrated successfully of dental implants in patients with EB<sup>9,10</sup>.

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The aim of this case report is to describe oral diagnosis of different oral manifestations associated or not to epidermolysis bullosa of two patients with junctional EB and the treatments provides.

## Case Report

### Case 1

A 24-years-old caucasian female was referred to medical and dentistry care at the Center of Studies and Care to Special Patients (CEAPE) at Paulista University (UNIP) in São Paulo. In the anamnesis, the patient medical report/geneticist to be suffering of junctional epidermolysis bullosa in mild form an inherited autosomal recessive, reporting that since its birth occurred scraping blistering in the whole body and the oral cavity, leaving scar marks, confirmed in clinical, principally in the areas of hands and mouth (Fig. 1 and 2). The oral examination revealed ankyloglossia microstomy, which lead to limited mouth opening.



**Figure 1.** Hands exhibiting multiple scars.

The patient was partially edentulous, with bullous lesions in the region of the buccal mucosa and lips at both sides (Fig. 2). In the region of the palate bullous lesions progressed to a painful ulcer (Fig.3), which has been treated with the application of laser therapy (GaAlAs) at a wavelength of 660 nm, 0,03 W of power with application of 4 J per point, during 133 seconds, presenting lesion remission during the period of 7 days. All dental intervention was implemented with

patient's care could not cause blisters in the oral cavity of the patient, care those in lubricate the instrumental for which will make them less traumatic, while also avoiding the use of suction system and total isolation.



**Figure 2.** Buccal Vestibule with microstomia and multiple scars with leukoplakia characteristics.



**Figure 3.** Oral palate with bullous lesions and multiple scars and ulcer.

### Case 2

A 55-years old caucasian female referred of University of Cuiabá, from Mato Grosso do Sul – Brazil, during the clinical interview, the patient reported a white lesion on the tongue, painless and with recurrence after 4 years. In the anamnesis reported being treated hypertensive with Atenolol® 50 mg submitted dermatologist report to be suffering from junctional epidermolysis bullosa, reporting that occurred since birth bullous lesions throughout the body and oral

cavity, leaving scar marks, confirmed by physical examination, especially in regions of hands, feet and knees (Fig. 4-5).



**Figure 4.** Hands with bullous lesions and loss of fingernails.



**Figure 5.** Knees with multiple scars.

The oral examination revealed microstomia, which had limited her ability to open her mouth, actinic cheilitis on lower lip, as well as lesion on the dorsum of the tongue leukoplakia painful (Fig. 6). The patient was edentulous with bullous lesions of oral mucosa, using removable dentures. The clinical diagnosis was papilloma and was planned surgical removal by biopsy, the piece was placed on the formaldehyde solution 10%, and sent to pathological examination, where the material was processed and examined, confirming the

diagnosis of squamous papilloma. The patient is currently with weekly application of 80% trichloroacetic acid and after four applications and follow-up of 6 months the lesion showed partial remission.



**Figure 6.** Tongue squamous papilloma.

### Discussion

In the past extractions were the treatments of choice for patients with different subtypes of epidermolysis bullosa<sup>7</sup>. Nowadays, preventive protocol is dental management approach of choice<sup>8</sup>, with different technologies such as in the first case presented with laser therapy in oral manifestations of EB.

Then laser therapy promotes biostimulation, analgesic and anti-inflammatory effects and promote tissue repair in different oral lesions<sup>11</sup>. In addition oral bullae, ulcers and erosion are the most common oral manifestations of EB<sup>8</sup>. Recently laser therapy was applied with the same protocol of the first case reported in the recurrent aphthous ulcer in HIV patients and showed an important alternative therapy to improve the quality of life of the patients as well as the present study<sup>11</sup>.

Rehabilitation with dental implants has been reported with success in the scientific literature. Peñarrocha et al. 2007<sup>9</sup> installed 38 dental implants with a success rate of 97.9 %, with a follow-up of 1 to 9 years. The peri-implant mucosa remained with good

health conditions in all patients<sup>9</sup>. After that Peñarrocha-Oltra et al., 2012<sup>10</sup> describe rehabilitation with dental implants place simultaneously with particulate bone graft in 4 patients diagnosed with epidermolysis bullosa provide support for fixed prostheses and improving these patients' quality of life.

Moreover in the oral cavity can occur in regions polymorphic bulbous lesions of oral mucosa, gingiva, palate and lips. In junctional and dystrophic forms for susceptibility to injuries, such as erosion and denaturation of tongue and besides ankyloglossia and microstomia<sup>1,2</sup>. In the second patient oral papilloma was an aberrant finding associated with EB, and then was treated by surgical removal with an adjunctive therapy of 80% trichloroacetic acid (TCA), promoting oral health. Actually human papillomavirus (HPV) have been treated with trichloroacetic acid with two different concentrations, and they concluded that TCA 80% was more effective, but this solution must be used only with careful consideration<sup>12</sup>.

As consequence treatment should be multidisciplinary, although it's not necessary adopt any specific therapy, and a careful approach benefits all patients. In patients with generalized subtype requires specific precautions during treatment to prevent tissue injury though the use of lubrication of instruments, lip and oral mucosa with petrolatum or hydrocortisone, drainage of blisters, as well as to avoid pressure on the tissues as described in the first case report<sup>3,7</sup>.

Furthermore Kramer et al., 2012<sup>8</sup> presents a guideline on dental care with people living with EB have been development by an international team of dentist, using an standard methodology based on a systematic review of the currently available systematic review.

## Conclusions

All patients with Epidermolysis bullosa (EB) can and should receive treatment, regardless of subtype and prevention

programs have a great effectiveness, these patients present multiple oral manifestations. Consequently multidisciplinary approach provides the interaction of professional staff and comprehensive care improving patients' quality of life.

## Declaration of Interest

The authors report no conflict of interest and the article is not funded or supported by any research grant.

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