

## Oral Management of Dystrophic Epidermolysis Bullosa: A Multidisciplinary Approach

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**Abstract:** Dystrophic epidermolysis bullosa is a rare genetically determined disease characterized by cutaneous and mucosal blistering secondary to minor trauma. The recessive form presents the greatest oral alterations leading to microstomia, ankyloglossia, vestibule obliteration and multiple decayed teeth associated to poor oral hygiene. The aim of this article is to describe the specific clinical manifestations of a patient diagnosed with dystrophic epidermolysis bullosa as well as dental treatment difficulties including the measures that dentists should adopt in order to provide an effective treatment and improve the patient's life quality.

**Keywords:** Epidermolysis Bullosa Dystrophica, Microstomia, Blister, Dental Care.

### INTRODUCTION

Epidermolysis bullosa (EB) belongs to a group of a rare hereditary genetic disease which is characterized by blistering of tissues containing stratified epithelia following minor trauma [1]. EB is divided into four broad categories based on the level of blistering within the dermal-epidermal junction, namely, EB simplex (EBX), junctional EB (JEB), dystrophic EB (DEB), and Kindler syndrome, which is a mixed level of cleavage [2].

Dystrophic EB is related to abnormal enzyme activity and type VII collagen gene mutations (a fibrous protein that is the main component of the anchoring filaments) producing serious mucosal lesions due to blistering below the lamina densa of basement membrane zone [3].

DEB has either autosomal recessive or dominant subtype. Individuals with the recessive DEB have extreme fragility of the oral and perioral mucosa [2]. The aim of this case report is to describe the oral health condition of a patient diagnosed with RDEB and to focus on measures that dentists should take into consideration to provide a safe and effective dental care.

### CASE REPORT

A 29-year-old patient was referred from the dermatology department with a chief complaint of painful teeth and oral ulcerations. The patient was diagnosed with an autosomal recessive DEB since his childhood. History revealed that our patient was born from an inbreeding marriage. According to the medical records, he was hospitalized several times for deep anemia and mal nutrition likely secondary to esophageal strictures.

Physical examination revealed dystrophic hands with syndactyly, alopecia, blistering lesions and healed scars especially in the back, elbows and knees (fig1<sub>b</sub>).



Fig-1: hyperkeratosis of palms, blistering lesions and healed scars in elbows

Intra oral examination showed poor oral hygiene with high plaque index and gingival inflammation, multiple decayed teeth (fig -2), bullae extending throughout the oral mucosa and atrophic

lesion of the tongue associated to ankyloglossia .The patient also had fibrous scars causing vestibule obliteration and microstomia (fig- 3).



**Fig-2: poor oral hygiene with multiple decayed teeth and microstomia**



**Fig-3: vestibule obliteration and blisters all over the oral mucosa**

Oral hygiene measures were not possible as tooth brushing trauma led to the eruption of new oral bullae. In addition to the clinical examination,

panoramic radiograph revealed a good crown-to-root ratio with no bone pathology or teeth agenesis (fig 4).



**Fig-4: panoramic radiograph showing unrecoverable teeth but a good crown-to-root ratio for residual teeth**

Based on the clinical and radiological findings, a logical treatment planning was developed including a preventive and supportive approach.

Preventive measures consisted of recommendations to eat soft food and avoid sticky food with high acidic content as well as gentle and daily tooth brushing.

The treatment strategies included at first 0.12 % chlorhexidine mouth wash associated to dental and root scaling using ultrasonic inserts. Unrecoverable teeth were removed atraumatically with uneventful postoperative recovery.

Oral lesions therapy was based on the sucralfate powder application over all oral mucosa

particularly on blisters 4 times a day for 3 months. Bioadhesive oral gel (Kin Care 15 ml) containing aloe vera and hyaluronic acid was also applied daily contributing in the oral mucosa healing.

At one month follow-up, decreased number of blisters and improved oral hygiene were noticed. The patient experienced a pain relief associated to re-epithelialization of the damaged mucosa (fig 5).



**Fig-5: decreased number of blisters, improved oral hygiene and re-epithelialization of the damaged mucosa**

The restorative and rehabilitative procedures were carried out under local anesthesia with slow injection to avoid epithelium detachment. Protection by the application of petroleum jelly was performed on perioral tissues, comissures and intra oral mucosa before each procedure.

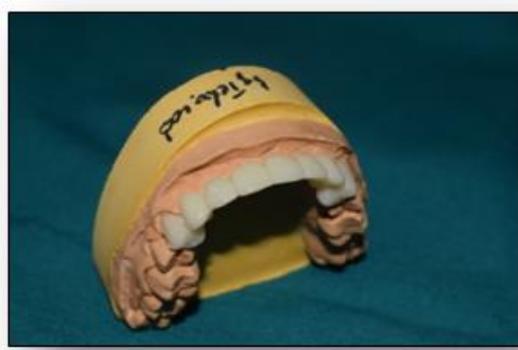
Maxillary first premolars and molars were restored with glass ionomer filling. Regarding the vestibule obliteration and the oral mucosa fragility to mild trauma and to avoid denture stomatitis, removable partial denture was discarded in favor of the fixed

prosthesis rehabilitation in the anterior maxillary area. Microstomia made the prosthetic steps difficult and challenging.

Individual impression tray was made and a provisional fixed denture using polymethyl methacrylate (PMMA) was designed by indirect laboratory technique to avoid the exothermic reaction and the chemical irritation on the oral tissues (fig 6a). A final cream metallic restoration was sealed (fig 6b). Instructions about oral hygiene and dietary habits were given.



**Fig-6a: provisional fixed denture**



**Fig-6b: final result showing the ceramo metallic restoration**

## DISCUSSION

Epidermolysis bullosa is a rare disease with multiple oral manifestations that require a special approach in terms of dental care. It presents a wide range of clinical phenotypes with over 1000 mutations identified in 13 structural genes [5].

The extent of oral involvement varies from one EB variant to another [4]: those with the severe types have widespread cutaneous and mucosal blistering that may lead to septicemia and may even be fatal [6]. In the simplex form the teeth are not affected and oral ulcers and blisters are rare, however enamel hypoplasia has been described in patients with JEB caused by mutations in the genes of laminin-332,  $\alpha 6\beta 4$ -integrin, and type XVII collagen [5].

The dystrophic types often display malformed teeth, dental decay and generalized blistering at birth [3] associated to extra oral manifestations including corneal erosions, scarring alopecia, erosions in tracheal epithelium, esophageal strictures, deformities of the hands and feet, loss of nail beds, congenital pyloric atresia, and late onset of muscular dystrophy [6].

Cycles of bullae formation may occur at any site in the oral cavity. Minor traumas to the oral mucosa often lead to a transudation of fluid into the epithelium, causing fluid-filled blisters [6].

Over time, oral blistering may lead to vestibule obliteration, ankyloglossia and microstomia. The mucous membrane of the mouth and the entire gastro intestinal tract are intensively compromised by scarring making difficult to eat. Defective enamel, poor oral food clearance and inability to achieve adequate oral hygiene may lead to rampant decay [4].

Perioral and cutaneous squamous cell carcinoma can occur by mid adulthood; therefore we should be extra vigilant in monitoring changes in oral ulcerations, such as the development of raised indurated borders [2]. EB has no known treatment, thus, systemic corticosteroids are minimally effective.

As a result, health care providers are able to offer palliative therapy only. Patients should be referred to a dentist as early as possible to identify any complications that needs special attention in order to start preventive programs and reduce the risk of developing dental diseases. Many published cases have shown that patients with DEB used to visit the dentist only for acute pain related to advanced teeth decay [8-5].

Although oral bullae, ulcers, and erosions are the most common oral manifestations of EB, there are only two published studies dealing with the management of these complications:

At first Marini *et al.* [9] have found that sucralfate suspension reduced the number of blisters, oral pain and gingival index. Usually used for the treatment of duodenal ulcers and reflux esophagitis, sucralfate seems to be effective in the prevention and treatment of oral lesions caused by DEB.

The cytoprotective activity of sucralfate may be related to the strengthening of some defense mechanism of the mucosa through the stimulation of mucus and bicarbonate secretion and the activation of endogens prostaglandins [9].

Another recent study [10] has shown the effectiveness of cord blood platelet gel and low level laser therapy in reducing long term intra oral discomfort related to ulcerations, however, randomized controlled trials are still needed to provide a promising option for the treatment of DEB oral lesions.

Adjuvant therapies has been proposed to prevent or treat secondary infections like topical and/or systemic antibiotics, Chlorhexidine 0.12% including mouthwashes, swabs, sprays, gels.... and a protein-rich diet, iron and zinc [7-1].

Local trauma during dental interventions is unavoidable, but it could be limited by mucosa and perioral lubrication with petroleum jelly. It's probably best to use soft and small sized instruments and the local anesthesia must be injected deeply and slowly to avoid blister formation.

Oral prosthetic rehabilitation can be fixed or removable depending on the state of residual teeth, oral condition and financial possibilities. Whenever possible, fixed rehabilitation is recommended as the tolerance to bear tissue-supported dentures is low. Endosseous implants can be successfully placed and restored with overdentures; peri implant soft tissues remained in good condition with no peri implant bullae observed [11]. All restorations should be well adapted and highly polished.

Microstomia can be a real challenge: in our case an individual impression tray was made since the mouth opening was restricted. Another alternative is to do a first impression with hard silicone and to use this as a tray adding light body silicone on a second step.

## CONCLUSION

Regarding the challenging multidisciplinary treatment steps, surgical procedures and dental care for patients with DEB, it's noteworthy to promote continued dental care and re-evaluation three times at least each year.

A preventive program should be introduced as soon as possible to preserve the teeth and offer a better quality of life since childhood.

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