EPIDERMOLYSIS BULLOSA IN DENTISTRY: REPORT OF THREE CASES AND REVIEW OF THE LITERATURE**

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Abstract

Epidermolysis Bullosa is a congenital, genetic, vesiculobullous disease. It may be autosomal dominant and recessively inherited. The major types of Epidermolysis Bullosa are Epidermolysis Bullosa Simplex, Junctional Epidermolysis Bullosa, and Dystrophic Epidermolysis Bullosa. Due to the systemic disorders specific to the Epidermolysis Bullosa patient, approach to dental care has gain importance in these cases.

This article presents improving of patient comfort with oral health maintenance and atraumatic dental interventions in three cases diagnosed with Epidermolysis Bullosa and reviews the literature regarding clinical applications in brief.

Three patients - one with Epidermolysis Bullosa Simplex and two with Dystrophic Epidermolysis Bullosa - were referred to our clinic by other medical institutions because of discomfort due to dental complaints. Intra and extra oral symptoms were examined and treatment plans were arranged for each one of them. Dental treatment was carried out mainly by means of establishing non-invasive methods like oral hygiene motivation along with professional debridement and radical methods like tooth extraction. All the treatments were done with precision by experienced specialists in the field of dentistry. Treated successfully the patients were taken under control for regular observation of the disease's oral symptoms.


Keywords: Epidermolysis bullosa, dystrophic epidermolysis bullosa, epidermolysis bullosa simplex, congenital disease, dental care.

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Introduction

Epidermolysis Bullosa (EB) is an autosomal genetic disease in which vesiculobullous lesions mainly affect mucosa and skin of oral, ocular, nasopharyngeal, respiratory, and genitourinary surfaces. Bullae usually develop following trauma to the skin.1 The symptoms include dystrophy or absence of nails, alopecia, exuberant granulation tissue, congenital skin loss, palmoplantar keratoderma, mottled pigmentation, and pigmented nevi as regard to the severity of the disease.1 EB presents in four main types determined according to the location of bulla in regard to the dermoeidermal junction; Intraepidermal EB (EB Simplex), Junctional EB, Dermolytic (Dystrophic) EB and Mixed EB (Kindler Syndrome).2 Intraepidermal EB also called Epidermolysis Bullosa Simplex (EBS) is an autosomal dominant type of the disease. It may appear at birth or during neonatal period with lesions in response to trauma on hands, feet, legs, knees, elbows and scalp. Bullae may heal without scarring with

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decreasing incidence within time. Prognosis is good in long term for this type of EB. Junctional EB is very rare and seen in generalized-severe (Herlitz type) or generalized-intermediate (non-Herlitz type) forms. The severe subtype usually causes death in the first 5 months after birth corresponding to developmental disorders, respiratory malfunction and sepsis. Dystrophic (Dermolytic) EB type expresses in two subtypes; dominant and recessive. In Recessive Dystrophic EB (RDEB) severe nutritional disorders may occur based on frequent mucosal involvement of the disease. Corneal erosions, dental caries, alopecia and dystrophy of nails are observed. Exposure to chronic trauma and healing can predispose for carcinoma of the skin. Based on fusion of fingers and scar tissue, flexion contracture may develop in joints. Dominant Dystrophic subtype is the most frequent form of EB with light attack of the disease, manifesting general or local alopecia, atrophic scaring and dystrophic nail formation. The Kindler Syndrome (Mixed EB type) is characterized by early formation of acral bullae, dermal atrophy and photosensitivity. This type is recessively inherited and very rare form of the disease that needs differential diagnosis with RDEB.

Independent of the type, all EB patients need special medical and dental care. The course of the disease must be monitored by professionals regularly and urgent measures taken if symptoms complicate. The dental care in patients with Epidermolysis Bullosa has changed dramatically in the last 30 years. In the past, considering the systematic complications of the disease, dental treatment was limited to only tooth extraction. Nowadays, dental treatments for these patients have become varied along with the improvements in dentistry.

This study presents clinical care of three patients with Epidermolysis Bullosa and evaluates measures to be taken from dental point of view.

Case History

Case 1: An eleven-year-old RDEB diagnosed male patient consulted to our clinic with complaint of a toothache. The RDEB was defined by a dermatologist but the need of periodical medical care was ignored. Family history revealed consanguineous marriage and lost two siblings from the same disease. In extra oral examination, bullae and erosions in patient’s head, neck and oral surfaces were observed, besides, fusions of fingers and toes related to bullae were seen (Figure 1, 2 & 3).
Alopecia was evident in patient's scalp. Based on the destructive effect of the disease there was an intensive hand and leg disability, and the patient was completely dependent upon his mother’s support in regard to basic daily activities such as feeding and toilet needs. In intra oral examination, dental plaque, caries, and many bullae were found, especially in palate. Because of patient's limited mouth opening (microstomia), dental intervention could not be done initially and only oral rinse containing 0.15% benzydamine and 0.12% chlorhexidine was prescribed. Cotton swabs were indicated instead of a tooth brush for the provision of gentle dental cleaning. Oral hygiene motivation was achieved and daily exercises were suggested for the improvement of mouth opening. A threaded cone shaped custom apparatus made of clear self-curing dental acrylic was prepared for this purpose. (Figure 4).

When mouth opening reached an optimal level so that a mini-head handpiece can reach the posterior teeth, patient’s periodontal treatment started with atraumatic and thorough ultrasonic scaling and polishing techniques. Fluor was applied once a week for the initial month and every third month following. All of the first molars were extracted as were aching and hopeless regarding treatment. Conservative needs like endodontic and restorative treatments were spread out in time as the patient was non compliant. Routine frequent follow-ups were scheduled.

Case 2: A twelve-year-old male patient was referred to our clinic because of toothache and gingival bleeding. In anamnesis, a story of EBS was seen. There was not consanguinity within the family and none of the blood related members suffered such a condition before. Although the restrictions of the disease the patient was able to meet his everyday needs alone and was socially active, even with sense of humour. In extra oral examination, scar tissue related to recovered bullae and proportional skin fragility in some surfaces was observed (Figure 5).
started with giving oral hygiene instructions. Debridement was performed and dental plaque eliminated using ultrasonic scaling devices. Residue on teeth was removed and restorative treatment was carried out in a few appointments. Upper central incisor teeth were restored with hybrid composite resin, whereas resin-modified glass-ionomer restorations were used for the upper and lower premolar and molars (Figure 6). Semi-annual controls were scheduled for the patient.

**Case 3:** Extensive caries and gingival bleeding were observed in 21-year-old male patient with RDEB, consulting to our clinic. The disease was diagnosed in a university hospital and the patient was under control at the university dermatology unit. In extra oral examination, hair loss, bullae in neck, fusions of fingers and toes, and lip and cheek tension were found (Figure 7, 8 & 9).

The patient could not use his hands effectively because of reduced flexible binding due to cicatrization in the skin and was having difficulties with holding and using tools like spoon or tooth brush. In intra oral examination which was limited because of restricted mouth opening, excessive tartar and plaque in anterior lower region were observed as well as dental caries detected in posterior region in radiological examination (Figure 10).

Upper anterior teeth were formerly missing due to extensive caries and unsuccessful conservative interventions. Any restorative procedure could not be performed on the patient, since the decayed teeth were impossible to access. However, tartar and plaque in anterior region were removed with atraumatic methods by means of ultrasonic debridement. Lower right first molar and second premolar were extracted carefully. Oral hygiene instructions were given by suggesting very soft and small-head sized
toothbrush and oral rinse was prescribed. Monthly controls were arranged for the patient.

Discussion

EB is an inherited disease characterized by vesicle and bullae especially in skin and its severity varies according to its type.\textsuperscript{5} The lesions may be local or generalized affecting patient’s life functions and may have serious consequences that might end in death.\textsuperscript{5}

The hereditary nature of EB expresses even at birth, which describes the relation with early morbidity and mortality of the disease.\textsuperscript{8} EBS can sometimes bring about serious infections, even sepsicaemia; the junctional type will often take a life. The RDEB patients generally live to 30 years of age while suffering from serious cardiopulmonary and renal complications, as well as metastatic squamous-cell carcinoma.\textsuperscript{8} The life expectancy of an EB patient decreases rapidly with age.\textsuperscript{5}

In present cases, intra and extra oral symptoms of patients diagnosed with EBS or RDEB were examined. In the EBS patient scar tissue based on healing bullae with proportional skin fragility was observed; however, newly formed bulla was not detected. On the other hand, any contraindication which prevented dental procedures was not identified. Due to limited mouth opening, and intense bullae and erosion in head, neck and oral areas of the patients with RDEB, dental intervention was performed with difficulty.

In the younger RDEB patient, feeding difficulties and accordingly, growth deficiency by the reason of stenosis due to mucosal involvement of oesophagus and temporomanibular limited joint mobility related to facial trauma scars were observed. Also, as the patient had fusion of fingers, thus, difficulties in manipulation; he was unable to perform oral care on his own.

The microstomia condition in patients with RDEB may result from adherence occurring during healing of vesiculobullous lesions. This is a problem leading to difficulties in performing self-activities, such as eating, speaking and maintaining oral hygiene,\textsuperscript{9,10} and is the biggest clinical obstacle in carrying out dental treatment.\textsuperscript{11}

EB patients with poor prognosis should do daily exercises to develop and maintain optimal mouth opening. For the purpose, wooden spatula or plastic threaded cone can be used.\textsuperscript{1} Exercises to be taken for 30-60 min prior to intervention will facilitate accessibility.\textsuperscript{12} Threaded cone made of dental acrylic was used in the present study to gradually increase mouth opening. When mouth opening reached a level facilitating for intra oral performance, unavoidable processes such as extraction of aching and hopeless teeth were carried out.

Crawford et al.\textsuperscript{13} stated that for patients with RDEB, extraction of all teeth was a treatment of choice. Wright,\textsuperscript{10} declared that it was possible to manage dental abnormalities successfully with the combination of local anaesthetics and restorative techniques. Torres et al.\textsuperscript{14} carried out caries prevention in RDEB patients with dietary recommendations maintained consistently, favouring for adopting habit of oral hygiene with frequent professional dental cleaning and fluoride therapy.

Olsen and Bourke,\textsuperscript{15} showed that in experienced professional hands, ordinary ultrasonic scaling can be very effective method in maintaining oral health and also harmless to even RDEB patients considering their fragile nature.

Marini and Vecchiet,\textsuperscript{16} expressed that sucralfate suspension reduced lesions to form and shortened the duration of oral mucosal bullae and ulceration; conditions associated with oral pain decreased and reduction in plaque development and gingival inflammation were observed. In present cases, patients’ comfort was increased by eliminating pain symptoms, thus, their nutrition got better and the chance to compensate the loss of nutrients, fluids and electrolytes through the collapsed integrity of the bullous skin improved. Skin moisturizers were suggested to prevent lesions forming in response to trauma and oral hygiene motivation along with professional debridement was carried out successfully.

It is possible to brush teeth in all patients with EB despite the predisposition to tissue damage. Brushing teeth can be performed even in individuals with RDEB by means of determining a proper tooth brush. Soft and small head sized toothbrushes are ideal for this purpose. It is important to wet and soften brush before brushing. Tooth brush handle is required to be specially adapted for the patient with poor manual dexterity due to pseudo-syndactyly.
During severe oral pain, in the event that tooth brush cannot be used temporarily, dental cleaning should be attempted using cotton-tipped swabs, disposable micro brushes, clean cottons or gauzes.\textsuperscript{10,17} Families and/or companions also have responsibilities for the maintenance of patients’ oral hygiene.

EB patients are individuals with need of special care. As the patients are highly susceptible to trauma, families must be aware of the consequences therefore careful in order to prevent even accidents that can normally be overlooked. Special clothes and tools with soft linings are indicated to be use by these patients and taping of the body parts with bandages for preventing potential lesions is essential. Frequent epidermal injuries require appropriate wound dressings, and if the wound surfaces facing each other are not separated during healing, fusion of the connective tissue is inevitable causing particularly pseudo-syndactyly.\textsuperscript{18} Patients should periodically be referred for medical and dental aid and preventive maintenance provided.

Dentist should list frequency of clinic visits individually for each patient according to the amount of present plaque and risk of tooth decay in patients with EB. While it is sufficient for some patients to be seen every 3-6 months, some patients need to be seen monthly. We see our patient with EBS in 3 month-periods, whereas check the patients with RDEB on a monthly basis. Rate of progression of caries and requirement for professional plaque removal should be assessed at these appointments, and application of topical fluoride and dietary recommendations should be made. Especially, recommending the use of antiseptic mouthwash liquids after each meal will help improve oral hygiene notably.\textsuperscript{19}

Consequently, as there are different subtypes of EB, it may show a variable course in each patient. RDEB has the most serious progression. Oral symptoms are severe. Particularly, the presence of intense plaque and dental caries related to microstomia are significant. Not only maintenance of oral treatment but also healthy nutrition is difficult due to microstomia, therefore, it is vital to enable maximum mouth opening with exercises. Carrying out dental treatment and maintenance of oral care will increase patients’ comfort that is already poor, which will help patients hold on to the life. Dentists, dieticians and dermatologists should work together on treatment of this rarely encountered disease the existence of which is reality and specialized centres should be established to treat complex cases.

Declaration of Interest

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References