

Dental Management of Patients with Epidermolysis Bullosa

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Summary

Epidermolysis Bullosa (EB) is a group of rare, genetic skin disorders characterized by fragility and blistering to minimal trauma. All oral surfaces may be involved, including the tongue, buccal mucosa, palate, floor of the mouth and gingiva. Common oral findings of the disease include microstomia, intraoral ulcerations and bullae formation, ankyloglossia, tongue atrophy, elimination of buccal and vestibular sulci, lingual depapillation and atrophy of the palatal folds. In these case reports; systemic findings, oral manifestations and preventive measures are described for 3 patients with EB, all of whom required extensive oral management. Early dental management and preventive care to minimize caries development and improve oral health is very important for patients with EB. Pediatric dentists play an especially important role in early intervention. In describing the dental management of three EB cases, this article stresses the importance of an aggressive dental preventive programme with strict oral hygiene instructions for patients and parents along with frequent professional cleaning and fluoride therapy.

Key words: *Epidermolysis bullosa, Dental management, Preventive dentistry.*

Introduction

Epidermolysis Bullosa (EB) is a group of rare inherited disorders, usually detected at birth or early childhood [1-4]. It is characterized by blister formation and extreme fragility of the skin and mucous membranes. Vesiculobullous lesions may form in response to trauma or spontaneously. Scarring of the extensor surfaces of the extremities, hands and feet are typical; milia occur frequently; and nails often become thick and dystrophic or are lost. The disorder affects both sexes equally and occurs in all racial and ethnic groups [5]. Approximately 400,000-500,000 people are affected worldwide, and no definitive treatment has yet been developed [6].

The 4 major inherited forms of epidermolysis bullosa are distinguished by the degree of ultra-structural cleavage, skin fragility and blistering [7-10]. Intraepidermal or EB simplex (EBS) is an autosomal dominant trait that is characterized by relatively mild blistering of the skin and mucous membranes. Of the 3 variants of the disease, EBS has the best prognosis, appearing at birth or shortly thereafter and usually improving at puberty. Dermolytic or Dystrophic EB (DEB) is either dominant or recessive, and like EBS, it usually manifests as bullous lesions that appear at or shortly after birth. However, the onset of DEB may be delayed until puberty. Increased collagenase activity results in excessive collagenolysis, reducing epidermal adherence and causing a separation of the epidermal layer. Junctional EB (JEB), which is the most severe form of EB, is a rare autosomal recessive trait that is characterized by extensive non-scarring skin bullae, dystrophic teeth and nails and oral erosion. Junctional EB can be life-threatening during the neonatal period [4,11].

The effects of EB on tooth formation and structure is unclear, and its clinical expression is highly variable, with involvement of oral soft tissue and in developing teeth varying considerably on a case-by-case basis [8]. Oral involvement can occur on all surfaces, including the tongue, buccal mucosa, palate, floor of the mouth and gingiva. Repeated blistering

with scar formation may lead to a decrease in oral aperture (microstomia), ankyloglossia, tongue atrophy, destruction of buccal and vestibular sulci, lingual depapillation and atrophy of the palatal folds [3,10,11].

The literature on oral health-care in EB subjects is relatively rare. As a result, dentists with no experience may find it difficult to provide appropriate treatment for such individuals [12]. The following case reports present the oral findings and dental treatment of 3 patients with EB.

Case Reports

Case 1

A 14-year-old male patient diagnosed with severe generalized Dystrophic EB (RDEB) was referred to the Department of Pedodontics from Faculty of Medicine, Department of Pediatrics. The patient complaints of dental pain, halitosis, gum bleeding and poor esthetics. No other members of the patient's family were affected by the disease.

Physical examination revealed generalized worn-out skin, blistering and scar formation, with blisters and vesicles present especially on the head and neck (*Figure 1a*). The patient's extremities were affected, with crusts particularly apparent on the skin of the knees and elbows (*Figure 1b*). The patient's fingers were adherent, and his nails were affected (*Figure 1c*). Bullous lesions were also observed on the oral mucosa. Scar formation had resulted in the formation of microstomia and ankyloglossia. The patient's maximum mouth opening was 14 mm. Clinical and radiographic examination showed missing teeth, decay and poor oral hygiene (*Figures 1d and 1e*), due in part to a soft diet and hand contractures.

Parental consent and patient assent for treatment was obtained. Treatment strategy comprised performing restorations and extractions and instructing the patient in proper oral hygiene. Unrestorable teeth were extracted and restoration performed under general anesthesia. Treatment was provided with minimal touching and using a lubricant



Figure 1A. Facial photograph for patient in Case 1.



Figure 1D. Anterior view of upper and lower arches (Case 1).



Figure 1E. Panoramic radiograph (Case 1).



Figure 1B. View of the right elbow showing crusts (Case 1).



Figure 1C. View of the left hand showing adherence of fingers, absence of nails, crusts, scarring and bullae (Case 1).

to protect commissures and intra-oral soft tissue. No pressure was applied to soft tissue, flat malleable retractors were used to separate the cheeks, small cotton rolls were used with a lubricant for isolation, and the use of high-vacuum suction was avoided.

The patient and his parents received instruction on oral hygiene routines, and the parents were advised to assist the child, who had limited manual dexterity. A soft nylon toothbrush with bristles softened under warm water and the regular use of a non-alcohol-based fluoride rinse were recommended. Moreover, a chlorhexidine 0.12% mouthwash rinse was also recommended twice a day for 2 weeks every 3 months, and a clinical appointment was scheduled once every three months for the topical application of a fluoride varnish. The patient was referred to a dietician for dietary counseling. The dietician adjusted the energy requirements for the child according to the need for catch-up growth and applied a high protein/calorie nutrition strategy. Increasing fibre and fluid intake with frequent consumption of meals and snacks were recommended.

Although the child was affected after several extractions, the quality of nutrition was improved because there was no tooth pain during chewing and oral blisters were decreased with the extraction of broken teeth. An improved quality of dental health has been maintained through regular follow-up visits that have continued for approximately 2 years.

Case 2

A 12-year-old female patient diagnosed with generalized DEB (DDEB) applied to our clinic with complaints of halitosis and poor esthetics. The patient was taking daily iron supplements for anemia. A sister who had been affected by EB had died many years earlier.

Physical examination revealed large blisters, vesicles and scar formation, especially around the neck and extremities (Figure 2a). The patient's nails were also affected (Figure 2b). Intraoral and radiographic examination showed generalized decay and poor oral hygiene (Figures 2c and 2d). The tongue was normal, and no oral blisters were observed; however



Figure 2A. Facial photograph (Case 2).



Figure 2D. Panoramic radiograph (Case 2).



Figure 2B. View of the right hand showing absence of nails, crusts, scarring and bullae (Case 2).



Figure 2C. View of upper arch (Case 2).

erosion was present over wide areas, and mouth opening was limited to 15 mm.

Parental consent and patient assent for treatment was obtained. Although the majority of teeth were restored using adhesive materials, unrestorable, transposed maxillary lateral incisors were extracted. Restorative treatment was performed with minimal touching and using a lubricant to protect commissures and intra-oral soft tissue. The patient was also instructed in proper oral hygiene as described in Case 1 above, and follow-up visits were conducted over a 2-year period.

Case 3

A 13-year-old female patient with severe generalized Dystrophic EB (RDEB) applied to our clinic with dental pain. She had no other systemic problem. The patient's cousin was also affected by EB.

Physical examination revealed generalized blistering (Figure 3a) and areas of crusted skin, particularly on the extremities. The patient's fingers were adherent, and the nails were indistinguishable (Figure 3b). Clinical examination showed no intraoral bullae; however, recurrent scar formation had resulted in the formation of microstomia that limited mouth opening to 11 mm. The patient's overall oral hygiene was very poor, and a panoramic radiograph revealed four teeth affected by caries (Figure 3c).

Parental consent and child assent for treatment was obtained. Due to the patient's limited mouth opening, the tooth that was causing pain could not be treated, and it was extracted under general anesthesia. The patient was instructed to perform daily exercises with wooden spatulas (Lanschützer et al. 2009) to improve mouth opening. Following an increase of 4-5 mm in mouth opening, 3 teeth were restored in the clinic. Moreover, the patient's overall oral hygiene has improved as a result of the increased mouth opening, and oral health has been maintained and controlled through routine visits over the past 2.5 years.

Discussion

Epidermolysis bullosa is a rare disease with multiple oral manifestations that require a special approach in terms of dental care [12]. The extent of oral involvement varies from one EB variant to another. In the mild forms, small blisters (<1 cm) may form and heal without scarring. In the more severe forms, cycles of bullae formation, erosion and scarring may occur at any site in the oral cavity. Over time, oral blistering may lead to obliteration of the vestibule, ankyloglossia and microstomia. Defective enamel, poor oral clearance of food and inability to achieve adequate oral hygiene may lead to rampant decay [13] that is most likely attributable to nonsalivary factors such as enamel involvement, soft-tissue alterations and/or diet [5]. Lesions may be distributed over the entire oral cavity, with potential involvement of all mucosal surfaces, and no site-specific pattern of involvement, such as blistering limited to or absent from certain intraoral areas, attributable to any of the different EB categories or subtypes [2].



Figure 3A. Facial photograph (Case 3).



Figure 3B. View of the left hand showing adherence of fingers, absence of nails and scarring (Case 3).



Figure 3C. Panoramic radiograph (Case 3).

Soft-tissue alterations and enamel defects are among the factors contributing to an increased risk of caries in EB populations [2]. Scarring and hand contracture may develop, further complicating oral-hygiene issues. Dental care through plaque control and careful prophylaxis is advised. Home care should include meticulous oral hygiene with a small-headed, soft-nylon bristled toothbrush softened under warm water and regular use of a non-alcohol-based fluoride rinse. Parents should be advised to assist children with manual dexterity problems [12,14-16]. In the cases reported here, regular follow-up that included strict oral hygiene instructions and parent education improved functioning, and the improved chewing subsequently led to improvements in nutrition.

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Limited mouth opening, formation of intraoral bullae and mucosal sloughing with trauma from hard foods and toothbrushing that leads to swallowing difficulties are often seen in children with EB. These children are often on hyper-caloric formulas, which, when combined with poor oral hygiene and swallowing issues, result in high caries rates [13]. For this reason, it is important to receive consultation from a dietician on the nutritional content of the diet and food consistency [12].

While most individuals with EB can tolerate routine dental treatment, some patients who require extensive treatment and those who have severe soft-tissue involvement are best managed under general anesthesia [3,10]. However, positioning and use of instruments such as endotracheal tubes, face masks, blood pressure cuffs, ECG electrodes and adhesives associated with anesthesia and vital-sign monitoring may cause serious post-operative complications such as infection and debilitating scarring [17]. Therefore, care should be taken to avoid tissue trauma during anesthesia and in the clinic in general. Although soft-tissue lesions resulting from restorative treatment has been reported to heal in 1-2 weeks without any specific treatment [18], in the cases presented here, precautions were taken to avoid soft-tissue damage during dental treatment, including lubrication of the lips and buccal mucosa, avoidance of high-vacuum suction, use of flat retractors to separate the cheeks and small-sized, lubricated cotton rolls for isolation [12,17].

The present cases emphasize the importance of recognizing EB and the multidisciplinary treatment required for affected patients. In view of the great difficulties and risks associated with the provision of dental treatment to patients with EB, prevention is crucial [6,12,13]. Good oral hygiene, routine dental control and possible diet standardization are especially important for. In addition to dental and oral problems, hand contractures in Patients with EB complicate their oral hygiene regimes. When providing dental treatment, minimal trauma must be maintained. Minimal touching and minimal interference can help to prevent new pre-operative and post-operative lesions from developing [2,20,21]. Patients may require frequent cleaning and topical fluoride application. Whereas neutral sodium fluoride topical applications and non-alcohol-based rinses may prove efficacious, topical application of a high-dose fluoride varnish may be preferable in patients with EB. Chlorhexidine rinses may also help to eliminate cariogenic microorganisms. Home-care regimens should include brushing with a soft-bristled toothbrush. Dietary counseling may best be managed by a dietician [12,13].

Conclusion

An aggressive dental preventive programme with frequent recall visits that include oral hygiene instruction for patients and parents along with frequent professional cleaning and fluoride therapy are the keys to maintaining patients with EB's oral health to as great an extent as possible.

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