Oral Manifestations and Challenges in Dental Treatment of Epidermolysis Bullosa Dystrophica

Thais Regina Kummer, DDS, MSc, PhD
Helen Cristhiane Müller Nagano, DDS, MSc, PhD
Silvia Schaefer Tavares, DDS, MSc, PhD
Bianca Zimmermann dos Santos, DDS, MSc, PhD
Carla Miranda, DDS, MSc, PhD

ABSTRACT

Epidermolysis bullosa (EB) is a rare congenital disease characterized by the formation of blisters following minimal trauma. Oral health can be compromised by oral bullae, perioral fibrosis, ankyloglossia, and a high risk for dental caries, all of which lead to a decreased quality of life. The purpose of this paper is to report the case of an 11-year-old female who presented with autosomal recessive dystrophic EB. Besides the skin manifestations, the extra- and intra-oral exams revealed lesions on the lips and mucosa, microstomia, ankyloglossia, depapillated tongue, and carious teeth. This case emphasizes that patients diagnosed with EB require multidisciplinary care, with the dentist playing an important role in oral health management.

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Epidermolysis bullosa (**EB**) is a rare dermatosis of genetic origin in which an adhesion disorder of the epidermis leads to the formation of mucocutaneous blisters following minimal trauma.[1] The disease can be classified based on the site of blister formation: (1) epidermolytic (simple), which is characterised by discontinuities in the epithelial keratinocyte layer; (2) junctional, which is characterised by separation within the basement membrane (lamina lucida); and (3) dermolytic (dystrophic), which is characterised by discontinuities in the underlying connective tissue (sublamina densa).[1] Epidermolysis bullosa dystrophica (**EBD**) may be transmitted by an autosomal recessive or dominant gene, the recessive form of which is more severe.[1]

EB is the result of mutations within the genes encoding for proteins in the epidermis or skin basement membrane zone (dermoepidermal junction).[1] In contrast, EB acquista (**EBA**) is a very rare, non-inheritable, mechano-bullous condition characterized by the development of autoantibodies that target Type VII collagen found in the basement membrane.[2]

Research has now shifted toward the identification of therapeutic interventions, including gene therapy, recombinant protein infusions, intradermal injection of allogeneic fibroblasts, and stem cell transplantation, that might eventually lead to a definitive cure for EB. Other developing therapies being explored are directed toward the enhancement of wound healing and the prevention of potentially life-threatening skin cancers in these patients.[3]

Dr. Kummer is an associate professor, School of Dentistry, Centro Universitário de União da Vitória, União da Vitória, Paraná, Brazil; Dr. Nagano is an associate professor, School of Dentistry, THUM Dental Studies Center, Joinville, Santa Catarina, Brazil; Dr. Tavares is a staff dentist, University Hospital, Universidade Federal de Santa Catarina, Florianópolis, Santa Catarina, Brazil; Dr. Santos is an associate professor, School of Dentistry, Centro Universitário Franciscano, Santa Maria, Rio Grande do Sul, Brazil; and Dr. Miranda is an associate professor, School of Dentistry, Universidade do Sul de Santa Catarina, Tubarão, Santa Catarina, Brazil.

Correspond with Dr. Miranda at ca_mirand@yahoo.com.br

Dentists should recognize that the oral sequelae of EB lead to high risk for caries, thus a strong prevention program for affected patients is of paramount importance.[4]

CASE DESCRIPTION

Publication of this case report was approved by the Human Research Ethics Committee of the Federal University of Santa Catarina, Florianópolis, Santa Catarina, Brazil (process no. CEPSH/UFSC #259/08).

An 11-year-old Caucasian female with a diagnosis of autosomal recessive EBD was referred for a dental evaluation by her dermatologist following hospitalization for the treatment of anemia, malnutrition, and dehydration. The patient had a history of skin blisters since birth, with no family history of the disease. There were reports of multiple hospitalizations due to bloody cough, digestive tract bleeding, chronic malnutrition, and for blood transfusions. The patient also had a history of esophageal stenosis as well as recurring dermatological and respiratory infections.

The physical exam revealed generalized hemorrhagic blisters, partial fusion of the fingers and toes due to synechiae, anonychia in the hands and feet, contracture in the hands (not affecting pinch or grip functions), and cicatricial alopecia. The extra- and intra-oral exams revealed blisters on the lips and mucosa, microstomia, ankyloglossia, depapillated tongue, and deep carious lesions (Figure 1).

The patient was on multivitamins, hydantoin, and ferrous sulphate. The patient ate a high-calorie diet primarily comprised of soft foods. She had considerable difficulty to perform oral hygiene due to oral lesions. A panoramic radiograph, which was used for caries diagnosis rather than intraoral films due to the patient's intraoral condition, confirmed the caries observed in the clinical exam. The occlusal radiograph revealed a periapical lesion on the permanent maxillary right central incisor and the absence of the permanent maxillary left central incisor (Figure 2). The left central incisor had been extracted under anxiolytic sedation, but the

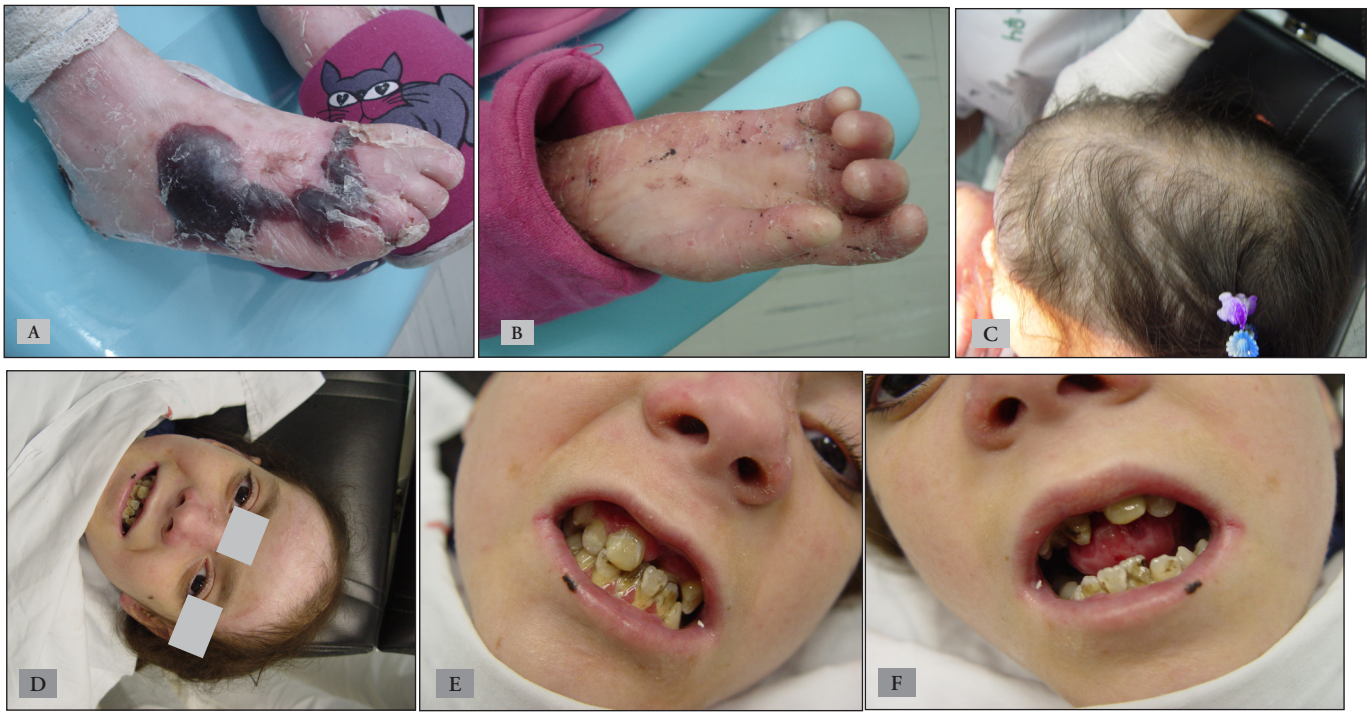


Figure 1. (A–B) Generalized serohemorrhagic blisters, partial fusion of fingers and toes due to synechiae, anonychia in hands and feet, contracture in hands, and (C–D) cicatricial alopecia; (E–F) lesions on the lips and oral mucosa, microstomia, depapillated tongue, and permanent teeth with caries.

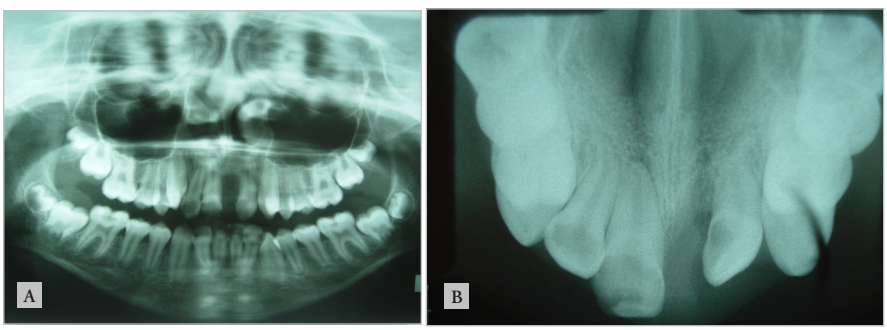


Figure 2. (A) Panoramic radiograph showing caries in the mandibular posterior teeth; (B) occlusal radiograph showing a periapical lesion on the permanent maxillary right central incisor and absence of a permanent maxillary left central incisor.

patient exhibited postoperative excitation and needed to be physically restrained, which led to a considerable increase in blisters.

Dental treatment was a challenge due to the sensitivity of the mucosa, constant presence of blisters, risk of causing new lesions, microstomia, limited cooperation from the patient, and impossibility of physical restraint due to the fragility of the skin. Using behavior management techniques, the patient gained confidence in the dental team and cooperated with treatment. Oral hygiene instruction, prophylaxis with rubber cup, and application of 2% neutral sodium fluoride foam were performed during each appointment, for four consecutive weeks (Figure 3a-c). Carious lesions were cleaned both manually and with low-speed round burs, and filled with glass ionomer cement. There was no need for local anesthesia for the root canal treatment of the permanent maxillary right central incisor, as the tooth was necrotic. A root apex locator was used (Figure 3d,e).

Monthly dental visits were scheduled to check the patient's dental health due to the fact that her underlying condition required a high-calorie diet and caused compromised oral hygiene, leading to high caries risk.

DISCUSSION

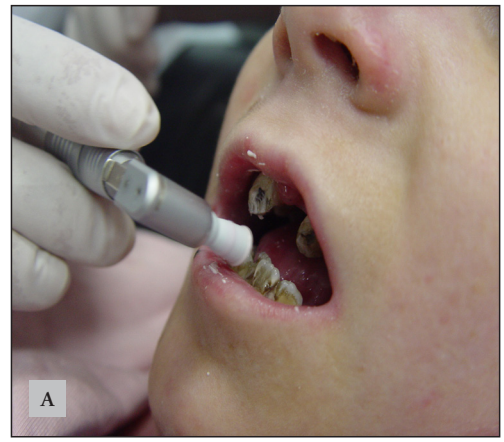
The clinical manifestations of EBD are present at birth. Besides causing skin and mucosal blisters, the joints, digestive system, respiratory system and speech are compromised. Abnormalities may also be found in the cornea and eye lids. Given the chronic loss of electrolytes and proteins due to the constant formation of blisters, EBD is most often associated with chronic malnutrition, leading to systemic complications, such as severe anemia, protein deficiency, infection, and retarded growth.[1,5]

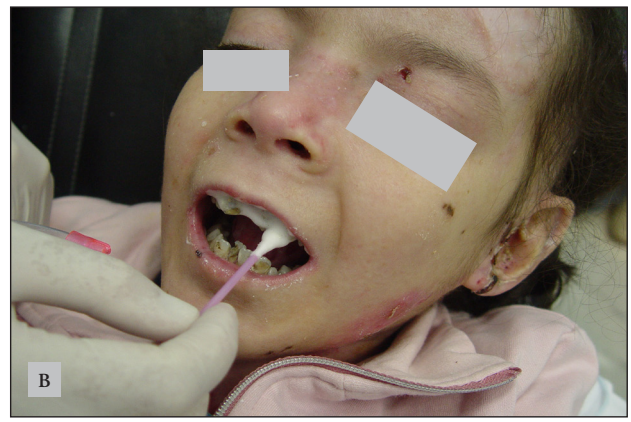
Oral blisters cause atrophic scarring, leading to microstomia, ankyloglossia, loss of the sense of taste, and pain upon minimal contact. Thus, oral hygiene is difficult to perform.[1,4-6] Such patients should, therefore, receive periodic evaluations for maintenance of oral health through diet and oral hygiene review, and for the early detection of dental disease.[4,5]

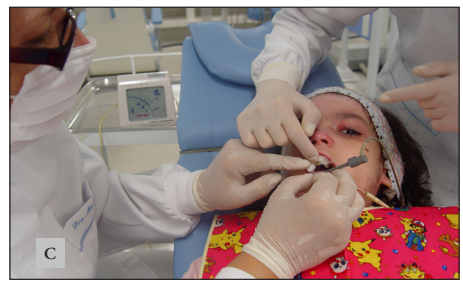
Patient cooperation is fundamental, as ulceration due to minimal trauma during dental treatment is inevitable.[4,7] There are measures that can be taken to minimize injury, such as abundant application of lubricating agents (eg, petroleum jelly) to all dental instruments, reduction in the force applied to the mucosal tissue and slow, and deep local anesthesia to avoid the mechanical separation of the tissues.[7]

General anesthesia is recommended when there is a need for multiple surgical procedures, treatment of severe manifestations of EB or when the patient cannot tolerate dental treatment. Oral or nasal endotracheal intubation may further traumatize the tissues and cause difficulty in airway healing and soreness postoperatively. The pressure of the laryngoscope on the supraglottic area during oral intubation causes more trauma and bulla formation to the tongue and oral mucosa.[4] Nasotracheal fiberoptic intubation is highly recommended, because it minimizes skin and mucosal trauma.[6] Therefore, it is important to combine multiple surgical procedures whenever possible.[6,7]

In conclusion, this case emphasizes that patients with EB require multidisciplinary treatment, with the dentist playing an important role in the prevention of dental disease. Adequate patient management through behavioral conditioning techniques and care to avoid trauma to oral tissues are essential.







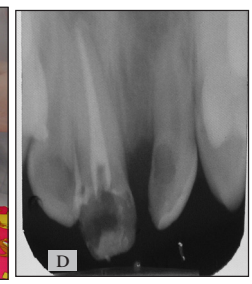


Figure 3. (A–B) Prophylaxis and application of fluoride; (C, D) root canal treatment of the permanent maxillary right central incisor (odontometry) and final radiograph.

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