*Arch Health Invest (2025)14(7):2013-2017*
http://doi.org/10.21270/archi.v14i7.6629

**Clinical Case**

# Oral Management of Recessive Dystrophic Epidermolysis Bullosa in Childhood: From Clinical Fragility to Functional Oral Care

*Manejo Oral da Epidermólise Bolhosa Distrófica Recessiva na Infância: da Fragilidade Clínica ao Cuidado Oral Funcional*

*Manejo Oral de la Epidermólisis Bullosa Distrófica Recesiva en la Infancia: de la Fragilidad Clínica al Cuidado Oral Funcional*

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**Abstract**
Introduction: Hereditary epidermolysis bullosa (EB) is a group of heterogeneous genetic disorders characterized by skin fragility, leading to blistering, erosions, and wounds on the skin and mucous membranes in response to minimal mechanical trauma with rupture at the dermoepidermal junction. Objective: To describe the oral manifestation and dental management of a patient with recessive dystrophic EB. Case report: A 6-year-old boy, diagnosed at birth with recessive EB, was referred to the dental service of the referral hospital for follow-up. Extraoral examination revealed synechiae and hand contractures, as well as blisters and crusted lesions on the upper and lower limbs. Intraoral examination showed ulcerated and erosive lesions on the lips, blisters and ulcerations on the buccal mucosa and lateral border of the tongue, tongue depapillation, petechiae on the palate, ankyloglossia, microstomia and mixed dentition with active carious lesions, gingival erythema and biofilm accumulation. Dental management included prophylaxis sessions, restorative procedures, photobiomodulation and oral hygiene instruction. Final considerations: Oral health management in EB patients requires early integration of dental care within a multidisciplinary team, adaptation of clinical techniques, use of assistive technologies, and strong emphasis on caregiver education, in order to significantly improve the quality of life for individuals living with EB.
**Descriptors:** Epidermolysis Bullosa Dystrophic; Dental Care for Children; Preventive Dentistry; Low-Level Light Therapy; Dental Staff, Hospital.

**Resumo**
A epidermólise bolhosa (EB) hereditária é um grupo de doenças genéticas heterogêneas caracterizadas pela fragilidade da pele, levando ao surgimento de bolhas, erosões e feridas na pele e nas mucosas em resposta a traumas mecânicos mínimos, com ruptura na junção dermoepidérmica. O objetivo deste artigo é descrever as manifestações orais e o manejo odontológico de um paciente com EB distrófica recessiva. Um menino de 6 anos, diagnosticado ao nascer com EB recessiva, foi encaminhado ao serviço odontológico de um hospital de referência para acompanhamento. O exame extraoral revelou sinéquias e contraturas nas mãos, além de bolhas e lesões crostosas nos membros superiores e inferiores. O exame intraoral mostrou lesões ulceradas e erosivas nos lábios, bolhas e ulcerações na mucosa bucal e borda lateral da língua, despapilação da língua, petéquias no palato, anquiloglossia, microstomia e dentição mista com lesões cariosas ativas, eritema gengival e acúmulo de biofilme. O manejo odontológico incluiu sessões de profilaxia, procedimentos restauradores, fotobiomodulação e instrução de higiene oral. O manejo da saúde oral em pacientes com EB exige a integração precoce do cuidado odontológico dentro de uma equipe multidisciplinar, adaptação de técnicas clínicas, uso de tecnologias assistivas e uma forte ênfase na educação dos cuidadores, a fim de melhorar significativamente a qualidade de vida de indivíduos que vivem com EB.
**Descritores:** Epidermólise Bolhosa Distrófica; Assistência Odontológica para Crianças; Odontologia Preventiva; Terapia com Luz de Baixa Intensidade; Equipe Hospitalar de Odontologia.

**Resumen**
La epidermólisis ampollosa (EA) hereditaria es un grupo de trastornos genéticos heterogéneos que se caracterizan por la fragilidad de la piel, lo que produce ampollas, erosiones y heridas en la piel y las membranas mucosas en respuesta a un traumatismo mecánico mínimo con ruptura en la unión dermoepidérmica. El propósito de este artículo es describir la manifestación oral y el tratamiento odontológico de un paciente con EA distrófica recesiva. Un niño de 6 años, diagnosticado al nacer con EA recesiva, fue remitido al servicio odontológico del hospital de referencia para seguimiento. El examen extraoral reveló sinequias y contracturas en las manos, así como ampollas y lesiones costrosas en las extremidades superiores e inferiores. El examen intraoral mostró lesiones ulceradas y erosivas en los labios, ampollas y ulceraciones en la mucosa bucal y el borde lateral de la lengua, depapilación lingual, petequias en el paladar, anquiloglosia, microstomía y dentición mixta con lesiones cariosas activas, eritema gingival y acumulación de biopelícula. El tratamiento odontológico incluyó sesiones de profilaxis, procedimientos restauradores, fotobiomodulación e instrucciones de higiene bucal. El manejo de la salud bucal en pacientes con EB requiere la integración temprana de la atención odontológica en un equipo multidisciplinario, la adaptación de las técnicas clínicas, el uso de tecnologías de asistencia y un fuerte énfasis en la educación de los cuidadores, con el fin de mejorar significativamente la calidad de vida de las personas con EA.
**Descriptores:** Epidermólise Ampollosa Distrófica; Atención Dental para Niños; Odontología Preventiva; Terapia por Luz de Baja Intensidad; Personal de Odontología en Hospital.

## INTRODUCTION

Hereditary epidermolysis bullosa (EB) is a group of heterogeneous genetic disorders characterized by skin fragility, with blistering, erosion, and wounds on the skin and mucous membranes in response to minimal mechanical trauma. The tissue cleavage occurs at the dermoepidermal junction[1]. Its classification is related to the layer of skin in which blistering occurs, namely: simple EB, junctional EB, dominant or recessive dystrophic EB, and Kindler's EB.

Approximately 20 different genes are responsible for the genetic heterogeneity of EB. These genes are associated with coding intracellular, transmembrane, or extracellular proteins, which are mainly structural components of the cytoskeleton, cell matrix, or cell-to-cell adhesion[2,3], Recessive dystrophic EB is the most severe form of the disease. It is caused by mutations in the COL7A1 gene, which encodes collagen VII. Collagen VII is the main component of the anchoring fibrils in the cutaneous basement membrane zone. The main extracutaneous complication is pseudosyndactyly of the feet and hands. Other complications include nail loss, hair loss, eye changes, gastrointestinal changes, nutritional deficiencies, growth changes, and secondary bacterial infections followed by sepsis[1,4].

Oral manifestations commonly seen in patients with EB are mainly the presence of ulcers and blisters, microstomia, tongue depapillation, ankyloglossia, periodontal disease, gingivitis, caries, occlusal abnormalities, dental agenesis, and risk of developing squamous cell carcinoma, which is the leading cause of death in patients with the recessive dystrophic form of the disease[4,5].

Dental treatment can be challenging, primarily due to the mucosal fragility, which increases the risk of bullous lesions development, in addition to the presence of pre-existing lesions and microstomia. The main precautions during dental care involve reducing shear forces and preventing adhesions by constantly lubricating instruments, thereby reducing the risk of tissue damage. Although there is no cure for EB, management is centered on supportive care aimed at improving the patient's quality of life[4].

This article presents a case of recessive dystrophic epidermolysis bullosa, detailing the associated oral manifestations and the dental management provided.

## CASE REPORT

A 6-year-old male patient presented to the Dentistry Department of a tertiary hospital, accompanied by his mother, with a diagnosis of recessive dystrophic epidermolysis bullosa (RDEB), confirmed since birth. He was the only affected individual in the family and was under clinical follow-up with a pediatrician and a dermatologist. He was not on continuous medication and had no other comorbidities. His daily care routine included the use of special dressings on skin lesions, glycerin soap, and body moisturizers. The caregiver also reported a cariogenic diet.

On physical examination, erosive and crusted lesions with areas of desquamation were observed on the neck and upper limbs, along with pseudosyndactyly in both hands (Figures 1A and 1B), for which he had undergone two corrective surgeries. On the lower limbs, extensive bullae, erosions, crusts, and skin desquamation with exudate were noted, protected by dermal dressings (Figure 1C).

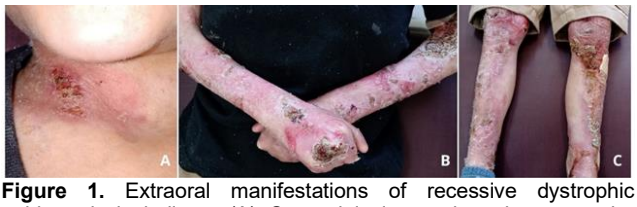


**Figure 1.** Extraoral manifestations of recessive dystrophic epidermolysis bullosa. (A) Crusted lesion and erythema on the cervical region. (B) Multiple crusted and erosive lesions with desquamation on the upper limbs and pseudosyndactyly of both hands. (C) Crusted lesions and blisters on the lower limbs, partially covered by dermal dressings.

Intraoral examination revealed an ulcerated lesion on the lower lip. Intraorally, multiple bullae were seen on the bilateral buccal mucosa, labial mucosa, and tongue, as well as petechiae and ulceration on the hard palate. The patient was in the mixed dentition stage, with carious lesions on teeth 52, 62, and 64, exfoliation of tooth 72, gingival erythema, and generalized biofilm accumulation. Ankyloglossia and microstomia were also observed (Figure 2).

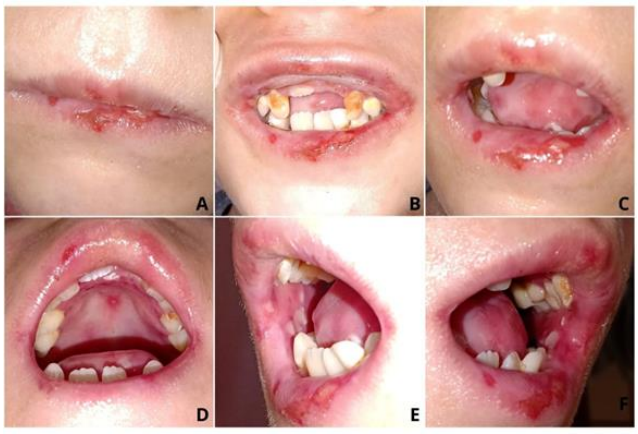


**Figure 2.** Intraoral manifestations of recessive dystrophic epidermolysis bullosa. (A–C) Extensive ulceration affecting the lower lip and perioral region. (D–F) Ulcerations affecting the oral mucosa, with presence of mixed dentition.

During the entire dental examination, all instruments and gloves were lubricated with essential fatty acid (sunflower oil) to minimize mucosal trauma and prevent the formation of new

bullae. The panoramic radiograph revealed a mixed dentition stage, with permanent teeth exhibiting age-appropriate development and primary teeth in various stages of exfoliation. Despite the normal dental development, reduced arch space was evident, particularly in the anterior regions, suggesting a potential for crowding as eruption progresses (Figure 3).

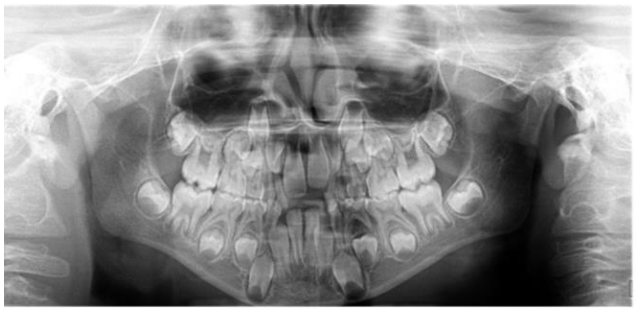

**Figure 3.** Panoramic radiograph of a 6-year-old boy, demonstrating normal mixed dentition development. Multiple permanent teeth are in various stages of formation, while several primary teeth are undergoing exfoliation. Limited arch space is evident, especially in the anterior region, indicating a tendency toward crowding.

Based on clinical and medical history, a comprehensive treatment plan was developed, which included dietary counseling to reduce intake of cariogenic foods and improve oral hygiene. The use of a soft-bristled, small-headed toothbrush, preferably single-tuft, was recommended. The patient underwent careful professional prophylaxis and selective caries removal, followed by atraumatic restorative treatment using light-cured glass ionomer cement on teeth requiring restorations. Relative isolation was used to minimize trauma, especially given the presence of microstomia, and the procedures were performed in two sessions, one week apart. Additionally, topical fluoride application was carried out, and a daily 0.05% sodium fluoride mouth rinse was prescribed to prevent caries progression, particularly in posterior teeth, due to hygiene difficulties.

As part of the management protocol for ulcerated and erosive lesions triggered by contact—even with lubricated instruments—photobiomodulation therapy (low-level laser therapy) was employed using a low-power laser device (Laser DUO MMO, São Carlos, Brazil), with the following parameters: 660 nm wavelength, 100 mW power, and 1 J of energy per point. The goal was to enhance healing, aid in tissue repair, and modulate the inflammatory process.

Six months after the initial visit, clinical examination revealed fewer ulcerative lesions on the lips and intraorally, with remaining ulcers on the tongue and hard palate. Oral hygiene had improved, with better biofilm control. New carious lesions were detected on teeth 63 and 64, with restoration fracture and eruption of permanent teeth (Figure 4). Atraumatic restorations were performed on teeth requiring them, and photobiomodulation therapy was repeated using the same parameters. The patient and caregiver reported dietary improvements and better adaptation to oral hygiene routines.

One year after the initial consultation, the patient exhibited milder ulcerations on the lips and tongue, effective biofilm control, and no new carious lesions, reflecting a significant improvement in oral health and quality of life (Figure 5). The patient has remained under continuous dental care at the reference hospital for two years, with routine follow-ups scheduled every six months or as needed.

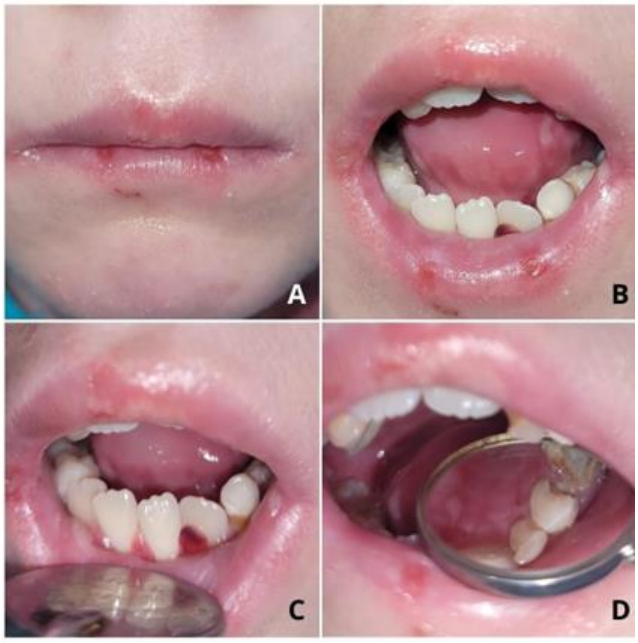


**Figure 4.** Six-month dental follow-up, fewer ulcerated oral lesions, presence of eruption of permanent teeth (A-B); hemorrhagic blister between the incisors (C); carious lesions on teeth 63 and 64, with restoration fracture (D).

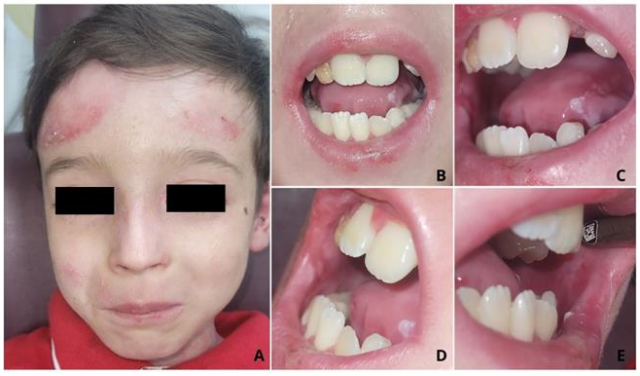


**Figure 5.** One year dental follow-up, observed improvement in extraoral and intraoral lesions, improved oral hygiene, and eruption of permanent teeth.

## DISCUSSION

Recessive dystrophic epidermolysis bullosa (RDEB) is a severe form of epidermolysis bullosa (EB) caused by mutations in the *COL7A1* gene, leading to defective anchoring fibrils and mucocutaneous fragility. Oral involvement is common and often extensive, with patients presenting early in life with blistering, ulceration,

and progressive scarring of the oral mucosa, as illustrated in this case.

The patient, despite ongoing medical monitoring since birth, had never received a dental evaluation until age six, when already presenting with severe caries, poor oral hygiene, and painful ulcerations. This delay exemplifies a significant gap in interdisciplinary care for EB patients. The absence of preventive oral measures during critical developmental years likely contributed to the deterioration observed. Literature emphasizes that dental professionals should be integrated into the care team from infancy to establish preventive routines, such as dietary guidance, atraumatic hygiene methods, and fluoride-based protocols, aiming to reduce early tooth decay and preserve mucosal integrity[5,6].

Anatomical alterations like microstomia, ankyloglossia, and vestibular obliteration, frequently seen in RDEB—as in this case—pose substantial clinical challenges. Restricted access limited both visualization and instrumentation, requiring careful planning and modified techniques. The use of selective caries removal, atraumatic restorative procedures, and glass ionomer cements under relative isolation were essential to minimize discomfort and avoid tissue trauma. Additionally, lubrication of instruments and gloves with sunflower oil played a pivotal role in reducing friction and blister formation, aligning with best practices recommended for EB patients[7].

One of the most impactful strategies adopted in this case was photobiomodulation therapy (PBM). Its analgesic, anti-inflammatory, and healing-promoting properties have been described in dermatologic and mucosal management of EB[8]. PBM sessions were well tolerated and contributed significantly to the resolution of mucosal ulcerations, improving comfort during and after procedures. While robust pediatric evidence is still limited, growing reports reinforce PBM's potential as a non-invasive adjunctive tool in EB oral care[9].

After one year of follow-up, the patient showed no new carious lesions, improved hygiene, and a notable decrease in mucosal lesions, demonstrating the effectiveness of a tailored and preventive oral health approach. These outcomes reinforce the notion that, even in medically complex and high-risk conditions like RDEB, early intervention and continuity of care can lead to tangible health improvements.

Beyond clinical adaptations, this case also emphasizes the essential role of family support and caregiver involvement. The psychological burden and technical challenges of daily oral care in EB can overwhelm unprepared families. Structured education and guidance empower caregivers to detect lesions early, implement trauma-reducing routines, and collaborate effectively with healthcare providers. Studies have shown that informed and supported families are instrumental in ensuring adherence to dental regimens and improving oral health outcomes in children with EB[10,11].

## CONCLUSION

In conclusion, managing oral health in RDEB requires early dental care integration within multidisciplinary team, modifications to clinical techniques, use of supportive technologies like PBM, and a strong focus on caregiver education and support. This comprehensive, patient- and family-centered approach has the potential to not only preserve oral function but also meaningfully enhance quality of life in patients with severe EB.

## REFERENCES

1. Has C, Bauer JW, Bodemer C, Bolling MC, Bruckner-Tuderman L, Diem A, et al. Consensus reclassification of inherited epidermolysis bullosa and other disorders with skin fragility. Br J Dermatol. 2020;183(4):614-627.
2. Has C, Fischer J. Inherited epidermolysis bullosa: New diagnostics and new clinical phenotypes. Exp Dermatol. 2019;28(10):1146-1152.
3. Has C, Liu L, Bolling MC, Charlesworth AV, El Hachem M, Escámez MJ, et al. Clinical practice guidelines for laboratory diagnosis of epidermolysis bullosa. Br J Dermatol. 2020;182(3):574-592.
4. Krämer S, Lucas J, Gamboa F, Peñarrocha Diago M, Peñarrocha Oltra D, Guzmán-Letelier M, et al. Clinical practice guidelines: Oral health care for children and adults living with epidermolysis bullosa. Spec Care Dentist. 2020;40 Suppl 1(Suppl 1):3-81.
5. Wright JT. Oral manifestations in the epidermolysis bullosa spectrum. Dermatol Clin. 2010;28(1):159-64.
6. Smith, Z., Nath, S., Javanmard, M. Salamón Y. The dental needs of children with Epidermolysis Bullosa and service delivery: a scoping review. BMC Oral Health. 2024;24:1131.
7. Siqueira MA, Silva JS, Silva FWGP, Diaz-Serrano KV, Freitas AC, Queiroz AM. Dental treatment in a patient with epidermolysis bullosa. Spec Care Dentist.2008;28(3):92-95.
8. Souza MS, Lopes LC, Almeida LG, Dantas MTC, Moura MDG, Almeida RB et al. Health challenges and importance of using photobiomodulation therapy in patients with epidermolysis bullosa: a cross-sectional study. Lasers Med Sci. 2025;40:285.
9. Oliveira NTJ, Paulo AC, Corona SAM, Borsatto MC. Effectiveness of low-laser therapy on cutaneomucosal lesions in patients with epidermolysis bullosa: a scoping review. Lasers Med Sci. 2025 21;40(1):204.
10. Togo CCG, Zidorio APC, Gonçalves VSS, Hubbard L, de Carvalho KMB, Dutra ES. Quality of life in people with epidermolysis bullosa: a systematic review. Qual Life Res. 2020;29(7):1731-1745.

11. Smith Z, Nath S, Javanmard M, Salamon Y. The dental needs of children with Epidermolysis Bullosa and service delivery: a scoping review. BMC Oral Health. 2024;24(1):1131.

## CONFLICT OF INTERESTS

The authors declare no conflict of interest.

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**Received** 08/07/2025
**Accepted** 30/07/2025