

Management of Patient with Epidermolysis Bullosa: A Case Report

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Introduction

Epidermolysis bullosa (EB) is a genetic disorder characterized by blistering, erosions, and scar formation of the skin and mucosa following mechanical trauma. The incidence of epidermolysis bullosa is 19.6 per 1 million births. There are four major types of EB (EB Simplex, Junctional EB, dominant dystrophic EB, and recessive dystrophic EB) that differ genotypically and phenotypically. The most common forms of EB present with skin blisters and secretion of colorless exudate. These blisters and erosions can occur due to trauma or any friction to the skin. Complications of the disorder include sepsis, failure to thrive, and squamous cell carcinoma, which continues to be the leading cause of death for these individuals.

Individuals with EB not only present with skin lesions, but also many have oral manifestations of the disorder. Recurrent oral blisters commonly occur due to mucosal fragility. These can create difficulties in feeding and oral hygiene. Additional oral manifestations include microstomia, ankyloglossia, vestibule obliteration, denuded tongue, enamel hypoplasia, periodontal disease, and increased risk of dental caries. Oral care of these children is challenging as they need a vigorous caries management plan guided by risk assessment at home and dental office as well as staged restorative and surgical intervention in dental office and under general anesthesia.

Case Presentation

An 18-year-old male patient with recessive dystrophic epidermolysis bullosa (RDEB) is of record at UCSF Pediatric Dentistry clinic since 2011. The patient has followed caries management program and received dental treatment of permanent first molars under general anesthesia in 2013. With the eruption of third molars, the patient returned for full mouth rehabilitation under general anesthesia in 2020. This case report highlights the prevention and management of oral diseases in children with EB. Emphasis is placed on anesthesia and soft tissue management.



Fig. 1 Extraoral Photo
Perioral granulation tissue

CLINICAL AND RADIOGRAPHIC FINDINGS

Extraoral: scar and granulation tissue, sloughing of skin

Intraoral soft tissue: generalized blistering and ulceration in palate and oral mucosa, denuded tongue, gingival inflammation

Intraoral hard tissue: Permanent dentition

#3, 7, 10, 14, 19, 26, 30 – missing

#1, 2, 4, 5, 12, 13, 15, 16, 17, 18, 20, 21, 28, 29, 31 (gross), 32 – dental caries

#1, 4, 13, 17, 18, 20, 32 – SDF-treated

ADDITIONAL FINDINGS

Patient was previously solely G-tub fed but has started eating by mouth

Oral Hygiene: poor. Toothbrush: 1x/day assisted by parents. Floss: 0x/week. Plaque and gingival bleeding noted

Caries risk assessment: High



Fig. 2 Extraoral photo Scarring and Mitten deformities in RDEB

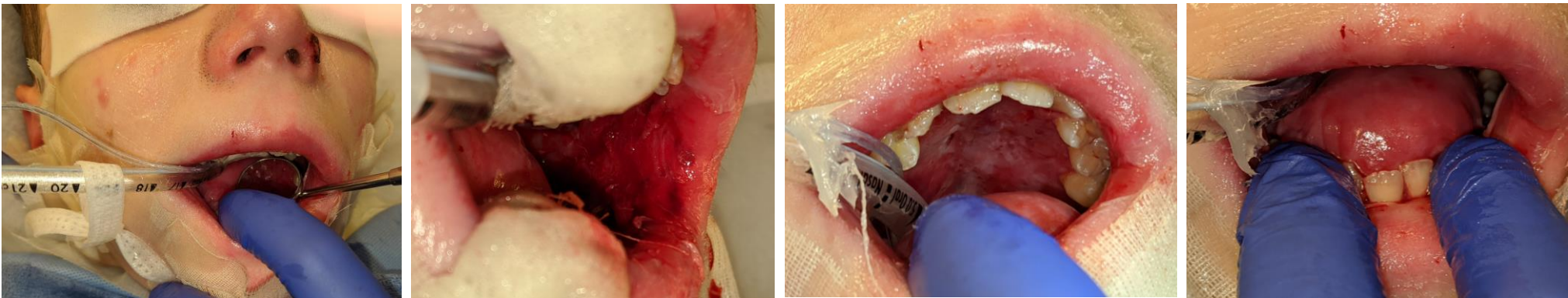


Fig. 3 Intraoral Photos Non-adhesive foam dressing protecting the contact areas, lips well lubricated. Microstomia, limited mouth opening. Bullae, erosions, and scars present on the palate and buccal mucosa. Absence of tongue papillae in RDEB

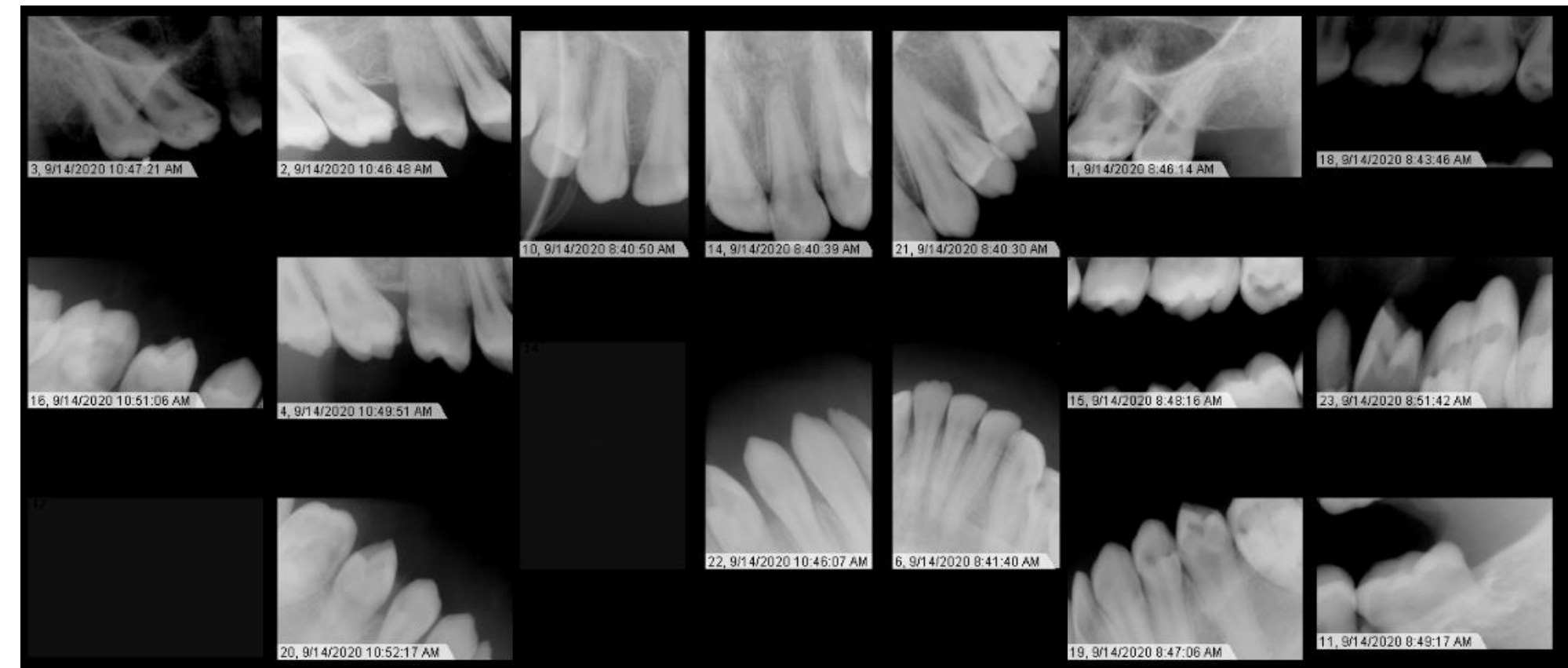


Fig. 4 Intraoral and Panoramic Radiographs (2020) Caries and missing permanent teeth

Treatment Plan

#18 and 21: amalgam restorations

#1, 4, 5, 12, 13, 17, 18, 20, 21, 28, 29, 32: composite restorations

#2, 15, 16, 31: prefab stainless steel crowns

#1, 4, 13, 17, 18, 20, 32: SDF

PREVENTIVE REGIMEN

- Oral hygiene instruction, including strategic use of electric toothbrush
- Diet counseling, including frequent hydration and rinse
- Fluoride toothpaste (5000ppm), baking soda regimen
- Chlorhexidine (0.12%, 1 week / month)
- 3-month recall interval

Discussion

For patients with EB, early referral and close follow-up are the keys to maintaining oral health. As in the present case, many reports have shown that patients visit the dentist only when they already have several carious lesions or pain¹. Although patients with milder oral involvement do not require extensive modifications to dental treatment, each patient should be evaluated individually. Patients with the severe generalized RDEB subtype of EB, which is presented here, require the most specific precautions during treatment to minimize soft-tissue damage¹.

Treatment under general anesthesia allows full mouth rehabilitation regardless of the severity of soft-tissue fragility and limited mouth opening. Coordination with an experienced anesthesia team is essential to prevent intraoperative generalized mucosal sloughing. Proper use of lubrication and non-adhesive dressing, positioning suction tip only on hard tissue, careful handling of the fragile tissue (minimal compression without shear forces), and drainage of intra-operative bullae are critical modifications in the dental management of patients with RDEB². In the present case, oral intubation, frequent application of lubricant, and full-body dressing were employed.

The present case also demonstrates the importance of long-term treatment planning in conjunction with rigorous preventive regimen. At appropriate stage of dental development, the extractions of the first permanent molars and teeth #7 and #26 under general anesthesia in 2013 orchestrated the alignment of present permanent dentition. This allowed the dental team to restore all carious permanent teeth under general anesthesia in 2020 providing a sound basis for long-term preventive regimen through patient's early adulthood.



Fig. 5 Panoramic Radiograph (2013) Extractions for molar substitutions and crowding

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