Epidermolysis Bullosa and Dental Management: A Case Report of a 14-Year-Old Patient

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Introduction

- Epidermolysis Bullosa (EB) is a group of chronic genetic disorders causing fragile skin and distinct oral features.² EB is classified into four major types (dystrophic, junctional, simplex, and Kindler Syndrome) with 35 subtypes based on clinical features, inheritance, and severity. ^{1,2}
- Diagnosis involves immunofluorescence mapping and genetic testing.²
- Dystrophic EB, caused by COL7A1 gene mutations, can be inherited dominantly or recessively. Recessive dystrophic EB leads to severe blistering, mitten deformities and digit fusion, oral and GI complications, chronic wounds, and a high risk of squamous cell carcinoma.¹

OBJECTIVE

This case report highlights the precautions and special considerations for dental treatment and management of patients with severe dystrophic EB.

Case Presentation

14y.o. male patient presented to UCSF Pediatric Dentistry for comprehensive dental care since 2015.

MEDICAL HISTORY

- · Genetically diagnosed with recessive dystrophic EB due to 1 recessive and 1 dominant collagen VII mutation (carrier parents, no family history).
- Hx of chronic malnutrition, upper GI hemorrhage, iron deficiency anemia requiring transfusions, corneal scarring, syndactyly, hand surgery in 2022, and history of dilated cardiomyopathy.
- Meds: Aquaphor, doxepin with coconut oil, triamcinolone, artificial tears, NKDA.

SOCIAL HISTORY

- 8th grader from Santa Rosa, CA, with good support system (lives with parents, 5 siblings, and grandparents).
- Has Medi-Cal insurance but no reported resource limits.

DENTAL HISTORY

- Hx of dental work under GA (age 7 and 12)). Planned but cancelled at age 6 due to parental hesitancy/fear of GA.
- **Oral Hygiene:** Poor with heavy plaque accumulation; difficulty brushing at young age; much improved with assistance brushing 2x/day with Waterpik several times a week and baking soda rinse after meals in recent months.
- **Diet:** Soft mashed food, more controlled and less frequent carbohydrate diet recently.
- Care disrupted since 2019 due to health issues, depression, and COVID-19; missed visits and intermittent pain.
- Behavior: In tears but tries to help, <2 cm opening, needs breaks, signals with hand raises. Behavior has drastically improved.

INTRA-GA PATIENT PREPARATION (Intubation, Skin/mucous protection)

Figure 2. OR Prep: (a) The Pink Pad® used. (b) Skin padded below BP cuff; tegaderm avoided for securing IV. (c) Petroleum gauze strip and Aquaphor used to lubricate and protect skin and mucosa. (d) Mask and arm wrapping used to protect skin blisters. (e) Face and eyes protected with petroleum gauze coverage; string used to secure oral tube instead of adhesive tape. (f) Microstomia (less than 2 cm). Mouth prop covered with moist gauze wrap.





OROFACIAL FINDINGS

Figure 3. Pre-op Photos: Generalized rampant caries and plaque accumulation with mucosal ulcers extraorally as well as intraorally. #9 and 24 in crossbite and permanent dentition with crowding.





Figure 1. Syndactyly, Aquaphor



Figure 4. Rubber Dam, Anterior Teeth: Facilitate restoration and protect soft tissues.

Figure 5. Panoramic Radiographs: (a) Age 7 (February 2017) before GA #1. (b) Age 11 (April 2021) before GA #2. (c) Age 14 (November 2024) before GA #3.





Post-Op

Figure 7. Post-op Photos: Composite strip crowns completed for anterior teeth and SSCs completed for posterior teeth.





Treatment GA March 2017: Age 7, early mixed dentition

- Glass ionomer restorations on #3. 14. and 30.
- Amalgam restoration on #19
- Extractions of all remaining primary teeth #A, B, C, D, H, I, J, K, L, M, R, S, and T.

Treatment GA January 2022: Age 11, late mixed dentition

- Composite restorations on #5, 7, 8, 9, 10, 12, 15, 18
- Protective restoration #31.
- Simple Extractions #3, 13, 14, 19, 25, 26, 30.
- Extraction of impacted #6 and 11.
- Frenectomy upper and lower lip due to scarring prohibiting speech and lip movement.
- Gingivectomies UR, UL, LL due to gingival irregularities and overgrowth.

Treatment GA November 2024: Age 14, permanent dentition

- Composite strip crowns on #7, 8, 9, 10, 22, 23, 24, 27.
- Stainless steel crowns on #2, 4, 5, 12, 15, 18, 20, 21, 29, 31.
- Extraction #28

Figure 8. 2mo post-op:

Patient is healing well and adapting to and loving his healthier smile.



Dental Treatment & Management

TREATMENT CONSIDERATIONS

- Empathetic, gentle, and supportive bedside manner and care.
- Definitive treatment requires GA due to microstomia and soft tissue fragility; still can be traumatic despite caution.
- Anterior PA and panoramic radiographs preferred for caries detection
- Severe EB often involves extensive oral scarring, limited mouth opening, and crowding. GA planning includes timed sequential extractions of primary and permanent teeth to facilitate good alignment: early mixed dentition (6-7 years), late mixed dentition (9-10), and early permanent dentition (12-14).
- Extensive precautions include use of Vaseline, petroleum gauze strips, softer bedding, and careful instrument handling to minimize soft tissue trauma (Figure 2a-f).
- Posterior teeth minimally prepped occlusally to increase vertical height and correct anterior crossbite.

POST-OP

- Patient is adapting to his healthier smile and teeth. He adjusted to new occlusion and reports chewing more easily. (Figure 8)
- #23 crossbite corrected to 10% OJ and 2mm OB.
- Patient is more motivated with OH now and in good spirits; OH improving and consistently rinsing with baking soda after meals.
- Improved gingival health with no bleeding in posterior teeth, mild plaque still present in anterior teeth due to lip tightness.

HOME PREVENTION AND FOLLOW-UP

- Lower frequency of carbohydrate diet.
- Parent-assisted brushing with small rotary toothbrush to reduce mucosal friction; frequent breaks.
- Rx 5,000 ppm F toothpaste; baking soda rinse and waterpik after meals.
- 3M recall with F varnish and SDF/SMART for caries prevention.

Conclusion

- Barriers to care: feelings of depression, inability to adhere to 3M recall schedule, COVID-19, and systemic health issues.
- Areas for improvement: better adherence to 3M recall, earlier, more frequent SDF applications/protective restorations (e.g. #28).
- Early prevention and multidisciplinary care are essential for managing oral disease and improving quality of life.²
- Care requires compassion, excellent understanding of EB, and tailored visits.

References

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