

Direct composite full mouth rehabilitation in a patient with Junctional Epidermolysis bullosa: A case report

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Abstract

Background: Epidermolysis Bullosa is an autosomal recessive disease that refers to a group of inherited bullous disorders characterised by blistering and erosion of the skin and mucous membranes. There are four major types recognised and 30 subtypes. This case study will focus on the non-Herlitz Junctional Epidermolysis bullosa type. Mutations are present for the basement membrane mediated cell adhesion specific for the oral mucosa and the developing tooth bud. These patients have a normal lifespan and dental intervention is critical. Scientific literature is scarce on the management of these patients and is mainly recorded as case reports. **Methods:** A direct composite was used to restore the aesthetics and function of this patient. **Conclusions:** These patients are not easy to treat due to severe blistering and a staged approach should be employed. The use of direct composite restorations provides the opportunity to conserve the tooth structure as enamel is critical for a predictable bond. This treatment modality is a feasible treatment option for patients with non-Herlitz Junctional Epidermolysis bullosa disease.

Introduction

Epidermolysis Bullosa is an autosomal recessive disease with an incidence of 1 in 50000 worldwide and 1 in 500000 live births.¹ This disease refers to a group of inherited bullous disorders and is characterised by blistering and erosion of the skin and mucous membranes². Diagnosis is made during childhood by examining the epidermal basement membrane using scanning electron microscopy, DNA analysis or immunofluorescence antigen mapping.²

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There are four major types recognised and 30 subtypes. The four major types include intraepidermal (simplex), junctional, dermolytic (dystrophic) and mixed (Kindler syndrome)¹. Tissue separation takes place within the basal keratinocytes in the intraepidermal (simplex) type. The junctional type presents with tissue separation within the lamina lucida of basement membrane. The dermolytic (dystrophic) type presents with tissue separation below the lamina densa. Lastly the mixed or Kindler syndrome present with a mixed cleavage plane of tissue separation.¹

Junctional Epidermolysis bullosa is divided into two subtypes: Herlitz and non-Herlitz disease. This case study will focus on non-Herlitz Junctional Epidermolysis bullosa. Patients with non-Herlitz Junctional Epidermolysis bullosa have a normal lifespan and the disease slows down with age.³

This disease presents with a mutation for the genes that codes for basement membrane mediated cell adhesion (laminin-332) specific for the oral mucosa and the developing tooth bud.^{1,4} Mutations of Type XVII collagen and



Figure 1: Right ear showing scarring and scabbing



Figure 2: Wounds on hand showing different stages of healing.



Figure 3: Scarring and blisters on the outside of the hand.

$\alpha 6\beta 4$ integrin may also be present. The main characteristics of the junctional type is marked soft tissue scarring and alterations in the developing hard and soft tissue in the mouth.³

Extra-oral manifestations include fingernail dystrophy, focal scarring alopecia of the scalp and loss of eyelashes. Intra-oral manifestations include blistering, enamel hypoplasia, pitting and furrowing of the enamel.³

Enamel defects are the most severe in Junctional Epidermolysis bullosa. Normal mineralisation takes place, but there is a significantly decreased mineral per volume contact that leads to enamel hypoplasia.⁵ The teeth are characterised by local or generalised enamel hypoplasia with pitting and furrowing defects. The enamel of Junctional Epidermolysis bullosa individuals differs from normal individuals with abnormal prismatic structure, orientation of the enamel rods and pitting.⁵

Main complaints from patients include rapid dental deterioration, severe aesthetic concerns, hypersensitivity to thermal, chemical, and mechanical stimuli and alterations in vertical dimension of occlusion.⁶

Little scientific literature is available in the management of these patients and is mainly recorded as case reports. A wide range of treatments have been reported in the literature. Treatment includes orthodontic treatment to align the teeth⁶ to implant supported restorations.⁷⁻¹⁰ Some authors used a combination of composite restorations and stainless steel crowns to restore the teeth.⁷ Other clinicians used stainless steel crowns in the paediatric patient.⁸ The restorations of the anterior teeth have been managed using telescopic abutments on the natural teeth and an acrylic bridge¹² and/or porcelain fused to metal fixed partial dentures². Lastly, Kramer (2010) reported the use of removable partial dentures to restore function.⁹

Excellent oral hygiene, regular professional cleanings with

topical Fluoride (alcohol free) and fissure sealants is essential to maintain oral health. Fluoride rinses (alcohol free) can be used as an adjunct.⁹

Case report

A female patient of the age of 18 years with non-Herlitz Junctional Epidermolysis bullosa, presented at the Dental Faculty of the University of the Western Cape.

The patient presented extraorally with multiple blisters, scarring and wound healing at various stages (Figures 1 - 3).

Intra orally the patient presented with an intense erythematous band of attached gingiva on the buccal aspect of the teeth (15-21; 32-44) (Figure 4). The teeth presented with multiple defective restorations, caries and generalised enamel hypoplasia with pitting and furrowing (Figure 5-8).

Sequence of treatment

The patient's intraoral soft tissues were coated with copious amounts of petroleum jelly to obtain primary study and working models. Working models were articulated and the bite was opened 3 mm with the use of a diagnostic wax-up (Figure 10).

Putty indices were made of the upper and lower anterior



Figure 4: Intense erythematous band of attached gingiva on the buccal aspect.



Figure 5: Loss of tooth structure (13) and pitting and furrowing (33-43).



Figure 6: Maxilla showing an ulcer in the hard palate, with defective restorations, loss of tooth structure and trauma of the gingiva of the palate (12-22 area).



Figure 7: Mandible showing defective restorations, loss of tooth structure, pitting and furrowing of teeth (34-44).



Figure 8: Pre-operative smile.

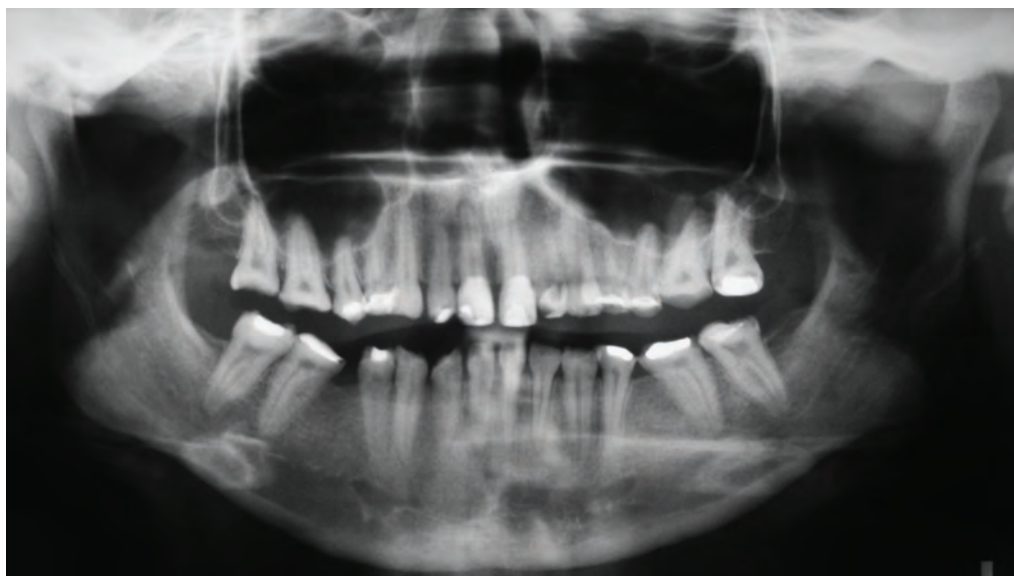


Figure 9: Pre-operative pantomogram.



Figure 10: Diagnostic wax-up

six teeth on the working models (Figures 11 and 12). These indices were used to transfer the new heights of the teeth from the models to the mouth.

During the rehabilitation period endodontic treatment was first performed on teeth 11, 21 and 37 due to irreversible pulpitis (Figures 13 and 14).

The two maxillary central teeth (11, 21) were first restored



Figure 11: Putty index of maxillary teeth



Figure 12: Putty index of mandibular teeth

using Filtek Block out agent (3M ESPE) to block out discolourations of these teeth. A2E colour (Filtek Supreme XTE, 3M ESPE) followed to complete the final restoration. The following appointment the 12,13,22,23, 33-43 teeth were restored with A2E colour (Filtek Supreme XTE, 3M ESPE) (Figures 15 and 16).

Rubberdam was used to isolate the posterior teeth and all

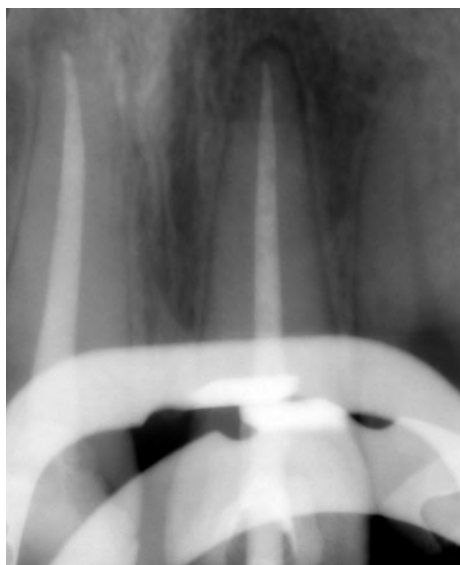


Figure 13: Endodontic treatment of teeth 11 and 21.



Figure 15: Anterior teeth restored with A2E (Filtek Supreme XTE, 3M ESPE).



Figure 14: Endodontic treatment of tooth 37.



Figure 16: Occlusal vertical dimension was increased by 3mm on the anterior teeth.



Figure 17: Restoration of the maxillary teeth 15-25.



Figure 18: Restoration of the mandibular teeth 34-44.



Figure 19: Right lateral view in maximum intercuspation



Figure 20: Left lateral view in maximum intercuspation



Figure 21: Restoration of the maxillary teeth 16,17,26 & 27.



Figure 22: Restoration of the mandibular teeth 36,37,46, & 47.

defective restorations and caries were removed, before each individual tooth was restored. A glass ionomer A3 colour (Vitremer, 3M ESPE) was used to replace lost dentine and A2E colour (Filtek Supreme XTE, 3M ESPE) for enamel. The teeth were built up in stages, first the cusps were built up

and cured, followed by the marginal ridges and lastly the inclines of the cusps.

Only two posterior teeth could be restored at a time, due to extreme blistering during the procedures. The posterior teeth were restored in sessions every two weeks in the



Figure 23: New aesthetic smile



Figure 24: Aesthetic result with the use of A2E Filtek Supreme XTE (3M ESPE).

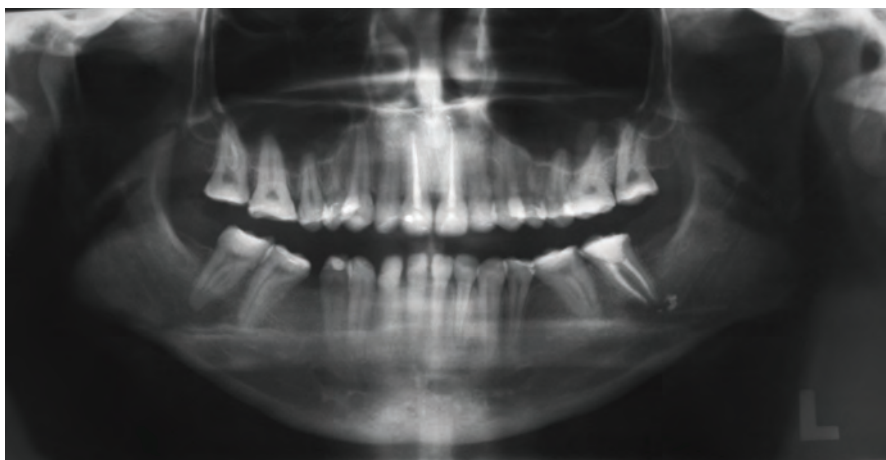


Figure 25: Post-operative pantomogram

following sequences to allow for healing: 14 and 44; 15, 24 and 25, 34 (Figures 17 - 20). The molar teeth were restored in the same sequence as the premolars. First the first molars were restored, followed by the second molars (Figures 21-25).

Complications

Peri-oral and intra-oral blistering were the main complications this patient experienced. Composite fractures developed (after 6 months) on the cervical areas of the 31 and 32 and were repaired (Figures 26 - 28).

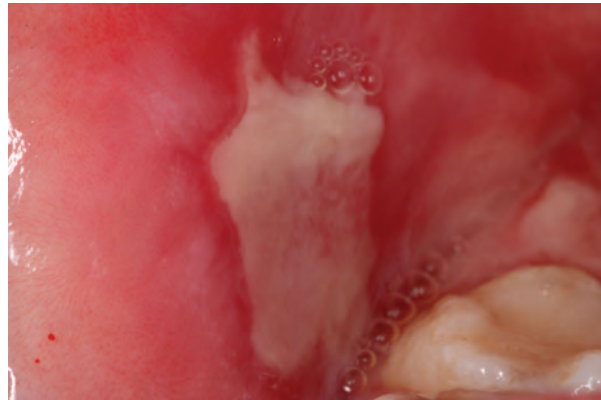


Figure 26: Intraoral ulcer after one month of dental procedures.



Figure 27: Any trauma leads to excessive blistering.



Figure 28: Healing after 1 week.

Conclusion

These cases are not easy to treat due to severe blistering and a staged approach should be employed. The use of direct composite restorations provides the opportunity to conserve the tooth structure as enamel is critical for a predictable bond. Special considerations, such as the lubrication of the soft tissues to reduce mucosa irritation during impression taking will reduce the iatrogenic effects of the treatment. Patient motivation is of utmost importance as the patient will have to content with ulceration between visits. A holistic treatment approach for these patients include regular follow up visits for maintenance and prophylactic hygiene.

Disclosure

The authors do not have any financial interest in the products used in this case.

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Figure 29: A biteplate was provided for use at night to prevent any chipping of the composite during possible night time grinding.

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