



## Dental management of a patient with epidermolysis bullosa: a case report

**Ahmed Albariqi**

Lecturer, Department of Periodontology and Dental Public Health, College of Dentistry, King Khalid University, Abha, Saudi Arabia

### Correspondence:

Ahmed Ali Albariqi, BDS, MDPH.

Doctorate of Paediatric Dentistry, University of Leeds, UK

Lecturer, Department of Periodontology and Dental Public Health, College of Dentistry, King Khalid University  
Abha, Saudi Arabia

Address: Guraiger, Abha 62529, Saudi Arabia

Phone: +966553188935

Email: dr.albariqi@hotmail.com

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### General Note

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### ABSTRACT

*Introduction:* Epidermolysis bullosa (EB) is a group of rare inherited disorders characterised by formation of blisters following minor trauma to the skin or mucosal surfaces. There are four major forms of EB- simple, junctional, dystrophic and Kindler syndrome. According to the form of EB, the oral and dental manifestations are varied in the frequency and severity. *Case Report:* A 6-years old girl with a dystrophic form of EB was attended in our dental care centre in 2018. On examination, the child had multiple blisters on

her elbows, feet, knees. Clinical and radiographic examinations revealed multiple carious lesions affecting almost the present primary dentition, with moderate gingival inflammation. *Treatment:* The patient had oral hygiene and dietary instructions, restorative dental treatment, extraction of badly decayed teeth. The patient was routinely followed up every three months for up to 1 year. On the last recall visit, the patient presented with good oral health status.

**Keywords:** Dystrophic Epidermolysis Bullosa, Blisters, Case Report.

## 1. INTRODUCTION

Epidermolysis bullosa is a group of rare vesiculobullous disorder characterised by making the skin and mucosal surfaces more prone to fragile. EB is first reported by Hebra in 1871 (Marini et al., 2001). Patients with this condition illustrate blisters following minor trauma or taction to the skin or mucosal surfaces (Fine et al., 1994). The degrees of severity are varied from blister formation to premature death depends on the defect on epithelial or subepithelial connective tissues (Rao et al., 2012). EB can occur in every racial and ethnic group affecting both genders equally with often becomes manifest at birth or first year of life (Momeni and Peiper, 2005). There are four major types of EB (simplex [KRT5 or KRT14 gene mutations, junctional [LAMA3, LAMB3, LAMC2 gene mutations], Kindler syndrome [FERMT1 gene mutation] and dystrophic type). Also, there are more than 20 distinctive clinical subtypes. However, the pathogenesis of EB remains obscure (Fine et al., 2000). The dystrophic (DEB) is caused by genetic mutations within the human COL7A1 gene encoding the protein type VII collagen. DEB has two subtypes (autosomal dominant and recessive subtypes). The autosomal dominant subtype is benign while the recessive type is the worst form of EB in which minor trauma leads to severe skin blisters and ulcerations (Album et al., 1977). The prevalence rate of dystrophic types of EB ranges between 1% to 2.8% in 50,000 births (Fine et al., 1999). The oral manifestations of DEB vary in severity and frequency between the patients from small to large blisters. The lesions can occur on buccal mucosa, floor of mouth, tongue, palate and gingivae (Hochberg et al., 1993). DEB patients have more susceptibility to dental caries due to their soft diet; tooth malformation and difficulty in maintain oral hygiene procedures due to inability to tolerate the trauma of brushing (Album et al., 1977). Several investigators reported other dental defects associated with DEB such as hypodontia, enamel hypoplasia, and gingivitis. Intraoral examination is difficult in patient with EB due to their continuous blister formation which may result in restricted mouth opening (Fine et al., 2000). The involved patients require particular precautions to be taken in order to avoid causing any trauma to skin and mucosal surfaces by using lubrication (Wright, 1990). Restorative treatment is possible under local or general anaesthesia. Some investigators suggest prescribing pre and postoperative antibiotic therapy in order to prevent infection of any blisters that may arise (Hochberg et al., 1993).

## 2. CASE REPORT

A 6 year-old girl diagnosed with dystrophic Epidermolysis bullosa (DEB) was seen in our dental care centre in 2018 for dental care and management. She had suffered from DEB since birth, and the diagnosis had been established at the Great Ormond Street Hospital in London, UK. She was the first child of unrelated parents. Her Father is also diagnosed with DEB. The patient was cooperative during the time of examination. The patient presented on the day of initial examination with several blisters involving her neck, elbows, hands and knees (Fig 1a and b).



**Figure 1a:** Bullae formation on hands.



**Figure 1b:** Bullae formation on neck.

The range of movement extremities was normal. Patient showed normal cognitive function and attending school. The patient complained of pain on the mandibular right quadrant. Intraoral clinical and radiographic examinations revealed multiple carious lesions affecting all primary molars and all primary canines with different degree of severity (Fig 2a and b).



**Figure 2a:** Right Bitewing x-ray



**Figure 2b:** Left Bitewing x-ray

The oral hygiene (brushing) was not practised daily due to the fear of blisters formation which resulted in Plaque accumulation and moderate inflammatory gingival inflammation.

### 3. TREATMENT

The dental treatment was discussed with patient and her parents. Normally the first visit would include treatment of the gingivitis and improve the patient oral hygiene. But because of spontaneous pain in the mandibular right primary molars, extraction was performed instead under local anaesthesia using cotton rolls with petroleum jelly was applied to all instruments and mucosal surfaces and gingivae in order to prevent trauma because of adhesion of cotton rolls.

In second visit, patient complained from spontaneous pain from mandibular left first molar. Extraction of mandibular left first primary molar was performed and caries removal of second left mandibular molar was done. However, caries extended into pulp tissue. So, a pulpotomy procedure was performed by removing only the 2 mm of the coronal pulp. Bleeding was controlled using ferric sulphate. When haemostasis achieved, Zinc oxide Eugenol (Kalzinol) was placed gently and tooth was restored with a preformed metal crown. There was no access to use a rubber dam, so the isolation was performed by sterile cotton rolls. The suction tip was lean on the occlusal surface of the tooth to prevent bullae formation. The air syringe was used very carefully with lowest pressure to prevent lesion formation.

In third visit, caries removal from maxillary right primary molars was performed using hand excavator and performed metal crown was placed on maxillary right primary molars without teeth preparation (Hall technique).



**Figure 3a:** Right Bitewing x-ray



**Figure 3b:** Left Bitewing x-ray

In fourth visit, caries removal from maxillary left primary molars was performed. However, maxillary left first primary molar had deep caries extended into pulp. So, a pulpotomy procedure was carried out and tooth restored with preformed metal crown. On maxillary left second primary molar, caries removal using hand excavator was performed and preformed metal crown was placed without tooth preparation (Hall technique).

In the fifth visit, a restorative treatment was carried out conservatively on all primary canines using composite restoration. All the dental treatment was done as a traumatically as possible in order to minimise lesion formation on the oral soft tissues. The suturing was not performed to avoid iatrogenic soft tissue trauma. Due to the high caries risk in this patient, dental recall scheduled every three months to emphasise on oral hygiene instructions and application of topical fluoride.

At one year follow up visit, the patient had no complaints of pain. The clinical and radiographic examinations showed good dental and periodontal health (Figures 3a, b, c, d).



**Figure 3c:** Lower occlusal view



**Figure 3d:** Upper occlusal view

#### 4. DISCUSSION

Epidermolysis bullosa is an inherited disorder characterised by skin and mucous membrane fragility. Although EB is a rare disorder, in dental clinic, EB required particular care due to complexity in mouth opening and epithelial fragility (Siqueira et al., 2008; Kramer et al., 2016). Al-Abadi et al., (2016) reported that the morbidity rate of EB is high. Dental treatment can be applied either under local or general anaesthesia. However, general anaesthesia may cause airway complications for the patient as a result of intubation procedure (Yoon and Ohkawa, 2012). Finke et al., (1996) reported that routine dental treatment can be provided for involving patient if particular care was taken. Conservative treatment is favourable in patients with EB in order to minimise the risk of developing oral laceration.

This case report illustrated many features of the dystrophic type of Epidermolysis Bullosa. The oral manifestations of EB vary in frequency and severity according to subtype (Hochberg et al., 1993; Alqarni, 2020). Wright et al., (1994) found that there is no oral or dental defect with simplest form of EB. However, the dystrophic types often showed enamel hypoplasia, high caries incidence, gingival inflammation (Harries et al., 2001).

The susceptibility of dental caries and abscess formation are high compared with healthy people (Wright et al., 1993) and this directly related to their incapability of having good oral hygiene, high consumption of soft diet containing sucrose, severity of soft tissue involvement as well as difficulty in maintaining tooth brushing (Toress et al., 2011).

In the present case, there was a high caries rate, plaque accumulation and gingival inflammation. At time of follow up visits, the gingival inflammation was reduced dramatically. The preventive and therapeutic procedures that were applied resulted in satisfactory oral health status. In this case, dental procedures such as extractions of unrestorable teeth, restorations and pulp therapy were successfully performed under local anaesthesia with minimally invasive methods.

Vital pulp therapy is known as a treatment that aims to preserve the pulp vitality, especially in young patients. Many studies have reported successful treatment outcome in vital teeth with curiously exposed pulp with signs and symptoms of pulpitis (Teixeira et al., 2001). Preformed metal crowns have proved to be an effective restorative treatment for children as they showed satisfactory results in many clinical cases (Zahdan et al., 2018). The Hall technique is an alternative method of treating caries in primary molars by placing a preformed metal crown without tooth preparation (Innes et al, 2015). This approach has steadily increased in popularity worldwide. Unfortunately, the extraction of some primary molars was unavoidable in this case as they were badly broken-down and showing signs and symptoms of chronic infection. The space maintainer was not considered in this case as patient at high risk of developing caries as well as to avoid blisters formation that may occur.

The patient was followed up every three months for one year. No new or recurrent dental caries appeared during this period. On each visit, an intra and extraoral examinations were done, oral hygiene instruction, diet advice and topical fluoride application were applied. A radiographic examination revealed that pulp therapy and restoration were satisfactory.

## 5. CONCLUSION

The oral and dental manifestation of patient with EB disrupts the patient's oral health and exhibit a challenge for the dentist. So, the involved patient should be seen on regular basis to minimise the risk of oral and dental tissues damage. Although dental treatment under local anaesthesia for patient with EB is complex due to restricted mouth opening and risk of soft tissue laceration, it can be safe and beneficial for the patient with preventing repeated general anaesthesia and its adverse effects as well as it is minimally invasive for airway.

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### Informed consent

Written and oral informed consent was obtained from the patient and parent' who included in the study.

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### Conflict of Interest

The author declares that there are no conflicts of interests.

### Data and materials availability

All data associated with this study are present in the paper.

### Peer-review

External peer-review was done through double-blind method.

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