

CASE REPORT



Emergency dental management of a child with epidermolysis bullosa under general anesthesia - A case report

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Abstract

The dental treatment of a medically compromised child presents a dilemma even to a seasoned clinician. The dental community has approached this dilemma by applying techniques that work in the normal population or by applying non-standard often untested techniques which work in these special groups. Epidermolysis bullosa (EB) is one such medical condition which when affecting the patient requires a scientific as well humanitarian approach. This case report of a 3-year-old patient suffering from recessive dystrophic EB highlights the above approach and the extreme care and precautions that need to be taken while handling such patients.

Keywords: Dental management, epidermolysis bullosa, general anesthesia

Introduction

Epidermolysis bullosa (EB) is a group of genetic inherited diseases characterized by blisters and vesiculobullous formations on skin and mucous membranes caused by minor instances of trauma or sometimes with no apparent etiology.^[1] While the specific pathogenesis for these disorders remains unknown, blisters usually occur due to defects in structure or biochemical abnormalities of keratin, hemidesmosomes, anchoring fibrils, anchoring filaments, and physiochemically altered skin collagenase.^[2]

EB can be classified into various types based on whether the condition expresses itself with a dominant or recessive inheritance pattern [Table 1].^[3]

Milder forms may display smaller vesicles which heal faster without scarring. However, in severe forms, the entire oral mucosa is affected and may cause severe blistering, scarring, microstomia, obliteration of vestibule, and ankyloglossia.^[4]

Case Report

A 3-year-old male child, weighing 8 kg, born of a non-consanguineous marriage reported to the dental outpatient department with a complaint of severe and acute pain in the lower lip region. The child was a known case of recessive

dystrophic EB (RDEB) with blistering since 3 days of age. On examination, blisters, bullae, and erosions were noted over the trunk (bilateral axillae, back, abdomen, elbow, and dorsum of foot) [Figure 1a]. Anonychia with tapering fingers and toes was also present [Figure 1b].

Intraoral examination revealed multiple carious lesions affecting almost the entire dentition, with severe gingival inflammation, microstomia, and elimination of buccal and vestibular sulci [Figure 2]. Due to recurrent oral blisters, the patient was unable to maintain his oral hygiene. The roots of lower incisors were exposed and protruding in the lip mucosa causing repeated ulceration and pain, because of which the patient was unable to eat and was on liquid diet [Figure 3].

Since the patient belonged to pre-cooperative age group, management was planned under general anesthesia. The pre-operative investigations showed the hemoglobin levels to be 8.6 g% and the rest were within normal limits.

The patient was premedicated with ketamine 6 mg/kg and glycopyrrolate 4 mcg/kg orally. Induction was done using sevoflurane in oxygen with Jackson Rees circuit. The face mask was lubricated with water-based jelly and Vaseline gauze was kept beneath the face mask. The electrocardiogram leads and the IV line were fixed to the limbs using Coban dressing [Figure 4]. The blood pressure cuff was lined with a thin layer

Table 1: Classification of EB

Characteristics	Dominant inheritance			Recessive inheritance	
	EB simplex (Koebner type)	EB simplex (Cockayne type)	Dystrophic (Cockayne-Touraine type)	Dystrophic	Junctional/Herlitz type
Age of onset	Neonatal	First to third decade	Neonatal	Birth	Birth
Site of primary bullae formation	Basal layer	Mid-epidermis	Papillary dermis	Papillary dermis	Lamina lucida
Lesion location	Generalized	Localized (hands/feet)	Generalized	Generalized	Generalized
Nail defects	Rare	Rare	Yes	Yes	Yes
Scarring	No	No	Yes	Yes	No
Oral lesions	Some	No	Rare	Yes Complete obliteration of vestibule, ankyloglossia, with age palatal rugae and lingual papillae, may become unrecognizable due to continuous blister and scarring	Yes Microstomia may occur in Herlitz
Dental defects	No	No	No	Yes	Yes (generalized enamel hypoplasia and rampant caries)

EB: Epidermolysis bullosa



Figure 1: (a) Skin lesions noted all over the trunk (b) anonychia along with tapering of toes noted



Figure 3: Exposed root of the lower incisor causing blistering and ulceration of the lower lip mucosa



Figure 2: Poor oral hygiene due to reoccurring oral lesions seen



Figure 4: Extreme care taken to avoid tissue trauma during and post-anesthetic procedure

of soft cotton before application. The temperature probe was also lubricated. The patient's eyes were moistened with a methyl cellular-based lubricant and padded with wet gauze. The patient was intubated using a 4.5 mm 1 D north polar (to avoid pressure on external nares) endotracheal tube and secured using roller gauze. The throat packing was also done using moist roller gauze.

Before starting the dental procedure, the patient's lips were lubricated with petroleum jelly to reduce the likelihood of tissue damage due to shear forces. The local anesthesia was injected deeper and slower than usual to avoid tissue distortion which may cause mechanical tissue separation and blistering. On manipulating the tissues, lateral forces which could induce tissue separation were avoided.^[5] The lower central incisors were then extracted uneventfully. The gauze used during extraction was moistened before use. Coronoplasty of sharp and attrited tooth surfaces was done to avoid further traumatic blistering. The post-operative recovery was uneventful.

In this case, the patient's nutritional condition was deteriorating further due to the ulceration, and hence, an emergency dental procedure was performed which enabled the patient to eat food. The patient was the American Society of Anaesthesiologists' category III due to which further dental treatment and/or prolonged general anesthesia was not recommended at present until further stabilization of his systemic condition.

Discussion

RDEB is typically more generalized and severe form of EB. Our patient had majority of the common manifestations including malnutrition, anemia, esophageal strictures, growth retardation, webbing or fusion of fingers and toes causing mitten deformity (pseudosyndactyly), development of muscle contractures, malformation of teeth, and microstomia.

Most interventions are palliative but help in increasing the life of patients with EB. Treatment consists of covering eroded skin surfaces with antibiotic dressings. Nutritional supplements with zinc and iron are important in treating anemia. Surgical interventions help to reduce mitten deformities and digital webbing.

Complete patient care in our patient required the services of a competent anesthetic team to support the dental staff. The standard of protocol for the general anesthesia procedure was devised concurrently by both dental and anesthetic teams which included few added precautionary measures to avoid any further peri and post-operative systemic complications.

Patients with minimal soft tissue involvement or limited dental problems can be treated under local anesthesia as an outpatient in the clinic. Nerve blocks are safer than infiltrations since they do not place the mucosal surface under pressure by depositing a bolus of fluid near tissue surface.^[6]

However, patients with multiple restorations or surgical procedures are best treated under general anesthesia. Extreme

care should be taken to avoid tissue trauma during anesthesia. General rule for management is the "no touch" principle which was strictly adhered to in our patient.^[7]

Excessive dental caries in EB is a result of presence and severity of the soft tissue involvement which leads to alterations in diet (soft and frequently high carbohydrates), increases oral clearance time (secondary to limited tongue mobility and vestibular constrictions), and creates an abnormal soft tissue and tooth relationship (i.e., buccal and lingual mucosa, which is firmly positioned against the tooth).^[8] Furthermore, these individuals lack the ability to routinely practice normal preventive oral hygiene measures or use of oral rinses.

Preventing dental caries in children with EB can be most challenging, especially due to their highly cariogenic diet and poor oral care. Brushing and oral hygiene measures may be hampered due to hand contractures and scarring, leading to poor oral hygiene. In such cases, parents should be counseled to assist the child in oral care measures.^[9] Soft bristled small headed toothbrushes, softened by dipping in warm water can also be used.

Mild chlorhexidine mouth rinses or better still swabbing chlorhexidine directly on the teeth may be very helpful. A nutritionist should be involved to help with the diet of these patients.

In our case, the oral ulcers healed after extraction of the offending teeth and this greatly improved the child's nutrition and general health status.

Conclusion

Although there is no specific cure for children suffering with the EB, as proper dental management can help by improving the quality of life of these children and improving their functional dentition and nutrition.

Furthermore, reducing soft tissue trauma by either extraction or restorations allows for efficient mastication and better nutrition.

Managing these patients are a teamwork and dentists form an integral part of this team by providing these patients with optimal oral care.

Clinical Significance

The global incidence of EB is low and the chances of encountering an affected child in the dental operatory are even lower. The severity of the effect of this condition on the general health of a patient and quality of life cannot be underestimated. This article sheds light on the medical considerations to be examined in such cases and highlights the extreme precautions to be followed.

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