

Epidermolysis bullosa — dental management and anesthetic considerations: case report

Patricia A. Lanier, DDS William R. Posnick, DDS, MPH, MS
Kevin J. Donly, DDS, MS

Abstract

Epidermolysis bullosa represents a group of hereditary skin disorders manifested by an exceptional tendency of the skin and mucosa to form bullae and vesicles as a result of trauma or friction.

Comprehensive dental care for preoperative-age children with this disorder frequently is managed in the operating room under a general anesthetic.

This case report describes an anesthetic protocol for dental management which was effective and resulted in minimal postoperative trauma. Eucerin® cream, Gauztex® bandages, and DuoDerm® pads were used to lubricate and stabilize anesthetic armamentarium. Preoperative and postoperative erythromycin antibiotic therapy was instituted to prevent infection of the bullae present.

Early diagnosis and institution of preventive measures can minimize the need for restorative and surgical management in these children. However, when anesthetic management is necessary, the use of appropriate consultants and adjuncts can provide valuable support.

degree of severity and phenotype of the disorder is related to the uniformity and pattern of the localized structural abnormalities (Sedano and Gorlin 1989).

Classification of EB can be based on phenotypic characteristics, mode of inheritance, or microscopic assessment of the specific tissue defect. Table 1 presents a summary of diagnostic criteria of the most common forms of EB.

Dental management of individuals with EB has been reported previously by several authors (Howden and Oldenburg 1972; Endruschat and Keenen 1973; Crawford et al. 1976; Kahn and Trieger 1976; Album et al. 1977; Carroll et al. 1983; Wright 1984). Dental restorations and extractions generally follow a variety of anesthetic protocols focusing on the need to minimize skin and mucosal tissue trauma.

The following case report presents a multidisciplinary approach to comprehensive dental management

Introduction

Epidermolysis bullosa (EB) represents a group of genetically transmitted vesiculobullous skin disorders of varying severity (Gorlin 1971). It is manifested by an exceptional tendency of the skin and mucosa to form bullae and vesicles as a result of trauma, friction, or spontaneous eruption. The etiology of the disorder is due to ineffective or absent connective tissue anchoring fibrils, resulting in a loosely bound epithelial/subepithelial connective tissue interface. The

TABLE Diagnostic Criteria and Classification of Epidermolysis Bullosa

Phenotypic Characteristics	Dominant Inheritance			Recessive Inheritance	
	Simplex Koebner Type	Simplex Weber-Cockayne Type	Dys-trophic Cockayne-Touraine Type	Dystrophic	Junctional (Letalis) Herlitz Type
Age of onset	neonatal	first to third decade	neonatal	birth	birth
Oral bullae	some	no	rare	yes	yes
Dental defects	no	no	no	yes	yes
Nail defects	rare	rare	yes	yes	yes
Scarring	no	no	yes	yes	no
Lesion location	generalized	hands/feet	generalized	generalized	generalized
Site of primary Bullae formation	basal layer	mid-epidermis	papillary dermis	papillary dermis	lamina lucida

of a child with Cockayne-Touraine type epidermolysis bullosa dystrophica.

Case Report

Preoperative Management

A three-year-old white female, CS, presented to the University of Texas Dental Branch Pediatric Dental Clinic for initial examination with multiple carious teeth and a medical diagnosis of epidermolysis bullosa dystrophica.

Past medical history included an uncomplicated pregnancy and delivery, with a diagnosis of Cockayne-Touraine type epidermolysis bullosa dystrophica made at birth. During the neonatal period she was placed on a diphenylhydantoin therapy regimen of two months on/one month off. She remains on this protocol with dosages being monitored and modified. With the exception of the dermatologic disorder and its sequelae, a review of systems proved noncontributory. Family history revealed that her father also is affected by the same disorder.

Upon physical examination, bullae were generalized and in various stages of eruption and healing (Figs 1 and 2).

Oral examination revealed a complete primary dentition with severe dental caries secondary to *ad libitum* and nocturnal bottle feeding of sweetened juice for 2 1/2 years. Although the bottle feeding had been discontinued, a pacifier habit resulted in a severe anterior open bite and perioral bullae formation. Intraoral soft tissue examination revealed plaque accumulations, generalized marginal gingivitis, and a distinct lack of oral mucosal bullae (Figs 3 and 4).

Clinical examination and charting was completed, noting that the child's behavior was difficult to manage. An unsuccessful attempt was made at obtaining

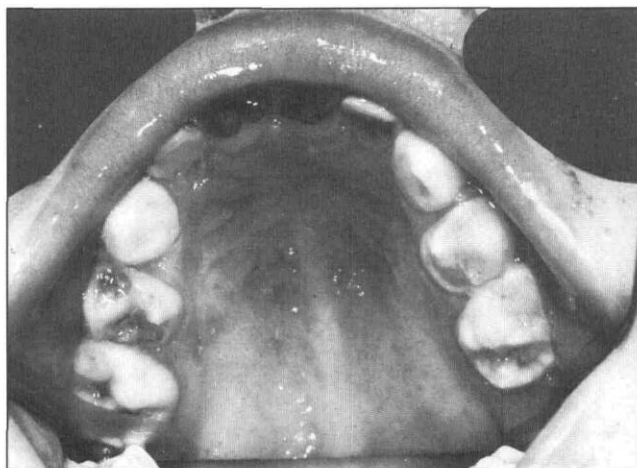


Fig 3. Preoperative maxillary dentition.



Fig 4. Preoperative mandibular dentition.

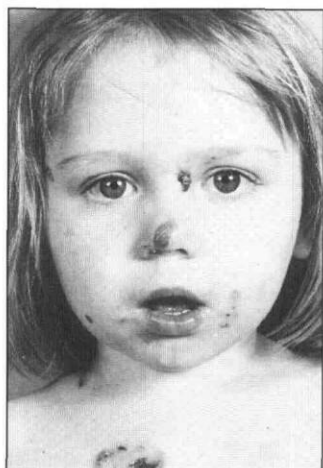


Fig 1. Facial bullae.



Fig 2. Generalized bullae in various stages of healing.

bite-wing and periapical radiographs. Although no oral bullae erupted as a consequence of the attempt to expose radiographs, placement of the lead shield resulted in bullae formation on her legs.

At CS's second visit, preventive counseling was initiated, including oral hygiene instructions, diet counseling, and home fluoride therapy. The decision was made to pursue consultations from pediatric medicine, dermatology, and anesthesiology directed toward providing comprehensive dental care under general anesthesia.

Appropriate consultations were obtained, and CS was scheduled for general anesthesia with specific consideration for minimizing bullae formation and infection.

Hospitalization and Dental Treatment

Prophylactic antibiotic coverage, erythromycin (125 mg every 6 hr), was instituted by the pediatric dermatologist six days prior to surgery to prevent infection of existing bullae at the time of admission.

The patient was admitted one day prior to surgery to review and update the history and physical examination and to verify her preoperative status by all medical consultants. On the morning of surgery the patient was taken to the holding area where 2 mg of diazepam were administered orally. She was carried sedated to the operating room and placed in a supine position on sterile lambswool bedding.

Halothane mask induction was accomplished using a thick layer of Eucerin® cream (Beiersdorf, Inc., Norwalk, CT) applied to the mask borders to prevent trauma. Gauztex® gauze (General Bandages, Inc., Morton Grove, IL) was placed on the patient's arm, and a blood pressure cuff was secured. An intravenous line was started and stabilized with tape applied over DuoDerm® pads (ER Squibb and Sons, Princeton, NJ, Fig 5).

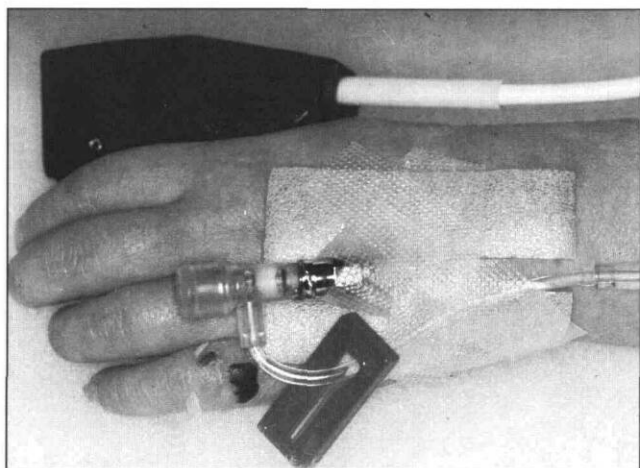


Fig 5. Stabilized intravenous line.

A precordial stethoscope was lubricated with Eucerin cream and stabilized with DuoDerm pads. The eyes were protected with petroleum jelly, and no head cover was used. Orotracheal intubation was achieved, and the tube was stabilized with tape to DuoDerm pads which had been adapted to protect the circumoral and facial skin (Fig 6).



Fig 6. Stabilized orotracheal tube.

A throat pack lubricated with sterile saline was placed conventionally, and intraoral radiographs were obtained. Restorative dentistry, including pulpotomies, stainless steel crowns,

and composite resin restorations, was performed with rubber dam isolation and a mouth prop for jaw stabilization. Maxillary primary incisors were extracted, and hemorrhage was controlled with light pressure and infiltration of a local anesthetic containing epinephrine. All procedures were performed using normal dental operating room protocol, augmented with additional sensitivity to tissue manipulation and pressure.

Postoperative Care and Follow-up

Following completion of the dental procedures, the mouth and oropharyngeal areas were carefully suctioned, and the throat pack was removed. All mucosal surfaces were inspected for bullae formation and none was noted. The patient was aroused and extubated in the operating room and transferred to postoperative holding. Sterile lambswool blankets were used over eggcrate foam cushions for all postoperative bedding.

The patient's postoperative course was uneventful. The intravenous line was maintained for a 36-hr postoperative period until adequate oral fluid intake was assured. All adhesive materials including DuoDerm pads were kept lubricated with Eucerin cream and atraumatically removed approximately 48 hr postoperatively. The patient was discharged 72 hr postoperative with a minimum of treatment-related bullae and was maintained on erythromycin for one week.

A follow-up oral examination was performed one week postoperatively, and normal healing was noted. Preventive measures were reinforced, and the patient was placed on three-month recall. Two subsequent recall examinations have taken place with no new dental caries and a noticeable improvement in hygiene and behavior.

Discussion

A disorder such as epidermolysis bullosa greatly compromises a dentist's ability to provide necessary dental treatment. The successful delivery of dental care involves consultation and cooperation with multiple medical specialists and knowledge and appreciation of the characteristics of the various categories of the disorder.

Dental care for the young uncooperative child with dystrophic epidermolysis bullosa can be delivered effectively in the operating room with inhalation anesthesia. The use of Eucerin cream, DuoDerm pads, and Gauztex gauze enable conventional anesthesia techniques, including mask induction, intravenous line placement, and stabilization of monitoring devices, to be used with minimum tissue trauma. In addition, oral and pharyngeal protection can be assured by careful placement of a throat pack and rubber dam.

The benefits of early consultation, preventive measures, and ongoing patient monitoring must be stressed

to minimize the need for restorative therapy.

Dr. Lanier is in private practice in pediatric dentistry in Houston, TX. Dr. Posnick is in private practice in pediatric dentistry in Galveston, TX, and is adjunct professor, pediatric dentistry at the University of Minnesota School of Dentistry. Dr. Donly is assistant professor, Department of Pediatric Dentistry at the University of Texas Dental Branch, Houston, TX. Reprint requests should be sent to: Dr. William R. Posnick, 2501 65th Street ~ Suite A, Galveston, TX 77551.

Album MM, Gaisin A, Lee KWT, Buck BE, Sharrar WG, Gill FM: Epidermolysis bullosa dystrophica polydysplastica: a case of anesthetic management in oral surgery. *Oral Surg Oral Med Oral Pathol* 43:859-72, 1977.

Carroll DL, Stephan MJ, Hays GL: Epidermolysis bullosa—review and report of case. *J Am Dent Assoc* 107:749-51, 1983.

Crawford EG Jr., Burkes EJ, Briggaman RA: Hereditary epidermolysis bullosa: oral manifestations and dental therapy. *Oral Surg Oral Med Oral Pathol* 42:490-500, 1976.

Endruschat AJ, Keenen DA: Anesthetic and dental management of a child with epidermolysis bullosa dystrophica. *Oral Surg Oral Med Oral Pathol* 36:667-71, 1973.

Gorlin RJ: Epidermolysis bullosa. *Oral Surg Oral Med Oral Pathol* 32:760-66, 1971.

Howden EF, Oldenburg TR: Epidermolysis bullosa dystrophica: report of two cases. *J Am Dent Assoc* 85:1113-18, 1972.

Kahn SB, Trieger N: Epidermolysis bullosa hereditaria letalis—a case report with special emphasis on oral manifestations. *J Oral Med* 31:32-35, 1976.

Sedano HO, Gorlin RJ: Epidermolysis bullosa. *Oral Surg Oral Med Oral Pathol* 67:555-63, 1989.

Wright JT: Epidermolysis bullosa: dental and anesthetic management of two cases. *Oral Surg Oral Med Oral Pathol* 57:155-57, 1984.

Healthy Snacks

According to a recent report by the U.S. Surgeon General, what you eat may affect your long-term health. On the average, about 40% of our daily calories come from fat. Experts say that figure should be 30% or less. You can help your children develop good eating habits, and improve your own, by choosing healthy snacks.

Examples of high-calorie, high-fat foods to avoid:

	Calories	Fat (gm)
Salted peanuts (1 oz.)	170	15
Potato chips (1 oz)	150	10
Corn chips (1 oz)	160	10
Candy bar (1-1/2 oz.)	150	8-10
Ice cream (1/2 cup, chocolate)	195	16
Pie (1/8 pie, apple)	181	11
Cheddar cheese (1 oz.)	114	9.4
Cheesecake (1 piece)	257	16.3

Examples of low-fat, low-calorie alternatives

Popcorn (1 cup, plain)	23	2
Three graham crackers	120	3
One apple	80	0.5
One pear	98	0.7
One carrot	42	0.2
Fruit yogurt (6 oz.)	190	4
Mozzarella (1 oz.)	72	0.7