Anesthetic management of a difficult airway in a patient with epidermolysis bullosa: A case report

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Epidermolysis bullosa is an inherited skin disease that leads to an array of medical problems. Patients are susceptible to blistering and scar formation following even minor trauma. These patients may present with scarring, limiting the range of motion of their temporal mandibular joint. This case report describes a 15-year-old patient with epidermolysis bullosa presenting for contracture release, with a difficult airway.

Key words: **Difficult airway, epidermolysis bullosa, fiberoptic intubation.**

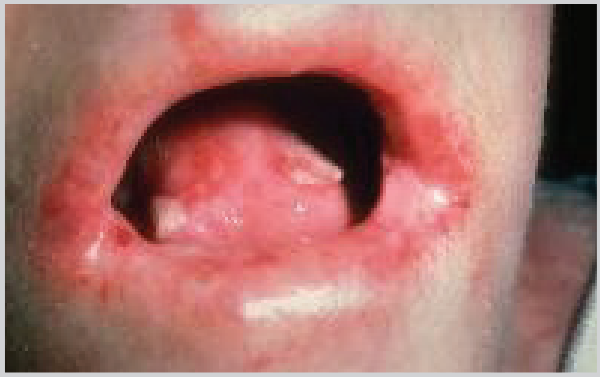
Epidermolysis bullosa (EB) is a rare, inherited disease characterized by excessive susceptibility of the skin and mucosa to blister formation after even minor shearing forces. It encompasses an array of autosomal dominant and recessive conditions that may have either localized or generalized dermatological manifestations.[1] The loss or absence of normal intracellular bridges is due to a collagen abnormality. The separation of skin layers occurs after application of friction or shearing forces and results in intradermal fluid accumulation and bullae formation.[2] The resultant scarring and contractures that develop during healing of the bullous lesions can lead to an array of debilitating, life-threatening medical conditions. Patients with EB frequently encounter infections, chronic anemia, and feeding problems because of oral and esophageal bullae formation.

In addition to the anesthetic considerations associated with positioning, monitoring, infection, and prevention of skin and mucosal trauma, the anesthetic management of patients with EB is uniquely challenging because of the effects on the airway. A difficult airway may result from perioral scarring that can restrict the opening of the mouth.[3] The tongue may be distorted by adhesions and the larynx may be stenosed.[3] Limited neck extension may be present secondary to scar formation. Furthermore, acute airway obstruction can result from bullae formation in the oropharynx secondary to airway instrumentation. This case report describes the anesthetic management for a patient with EB presenting for contracture release with a difficult airway (Figure).

Case summary

A 15-year-old, 56-kg male, with dystrophic EB, presented for an elective right hand flexion contracture release with placement of a bilayered skin substitute. Past medical history was insignificant with the exception of EB. Past surgical history included circumcision as an infant, gastrostomy tube placement at age 10 secondary to feeding difficulties, and release of toe contractures, all of which had been performed under sedation. The patient's trachea had never been intubated. Physical examination was remarkable for small skin erosions in the neck and chest areas, as well as on the patient's arms and legs. These erosions were lubricated with ointments and wrapped in bandages before arrival to the short surgical procedure unit. An airway assessment revealed limited neck extension secondary to scarring, a small mandible, and a severe range of motion limitation in the temporal mandibular joint. The patient's airway was classified as a Mallampati grade 4. After discussion with the patient, family, and

Figure. Recessively inherited dystrophic epidermolysis bullosa, oral cavity blistering, and scarring



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the plastic surgeon, the decision was made to proceed with an awake nasal fiberoptic intubation. A preoperative sedation dose of oral midazolam, 15 mg, was given 20 minutes prior to transporting the patient into the operating room (OR).

Prior to positioning of the patient, the OR table was padded with egg crate foam. Upon arrival into the OR, the patient moved himself to the OR table in order to decrease the risk of any shearing force. Electrocardiogram monitors with the adhesive portion removed were available if necessary, but not applied. Heart rate and rhythm were monitored from a clip on pulse oximeter, which was lubricated. An elastic bandage wrap had been applied to both upper arms in preparation for the placement of the blood pressure cuff and surgical tourniquet. The blood pressure cuff was applied to the left upper arm and cycled every 15 minutes or more frequently if necessary. An axillary temperature probe was lubricated and placed under the left axilla but was not secured with adhesive tape. Nitrous oxide, 50%, was provided via a well-lubricated facemask for peripheral intravenous catheter insertion. A 20-gauge needle was inserted intravenously in the patient's left forearm without the use of a tourniquet and secured with an elastic support wrap.

Intravenous sedation was started with incremental boluses of midazolam and fentanyl titrated to effect. The patient's oropharynx was sprayed with benzocaine spray. Oxymetazoline hydrochloride drops were applied to both nares. A small, lubricated flexible fiberoptic scope was inserted via the patient's right nare. The vocal cords were visualized with the aid of the fiberoptic scope, and a lubricated 5.5-cuffed endotracheal tube (ETT) was advanced using the scope as a guide wire. The ETT position was confirmed with the aid of a fiberoptic scope, positive end-tidal CO_2, bilateral chest excursions, and auscultation of bilateral equal breath sounds. The endotracheal tube was secured with ribbon gauze ties wrapped around egg crate foam, which was placed around the back of the head and the cheeks. The nare was lubricated around the ETT. No direct contact occurred between the gauze ties and the patient's skin. After the ETT was secured, general anesthesia was induced with ultane and nitrous oxide, 70%. Neuromuscular blockade was achieved with rocuronium 0.6 mg/kg. Both eyes were well lubricated but not taped prior to the start of the procedure.

As the surgeon completed the procedure, morphine was titrated while monitoring the respiratory rate. Ondansetron, 4 mg, was administered to prevent postoperative nausea and vomiting. Neuromuscular blockade was antagonized. After meeting extubation criteria, the patient was extubated. The patient was admitted to the postanesthesia recovery room for an uneventful postoperative recovery. The patient had no recall of the OR events and was discharged the next day.

Discussion

The fundamental pathophysiologic abnormality in EB is an increase in collagenase activity, which leads to collagen degeneration.[1] The abnormal collagen does not allow for the various layers of skin to anchor to each other. Bullae formation occurs secondary to transudation of fluid into disrupted skin. As the bullae heal in the dystrophic forms of the disease, significant scarring occurs. Shearing forces and friction present a larger threat than perpendicular pressure.

More than 20 different subtypes of EB have been identified and are broadly grouped into 3 main categories: EB simplex, junctional EB, and dystrophic EB.[4] The incidence of EB is approximately 1:17,000, and 46.5% of the cases are dystrophic. Epidermolysis bullosa simplex is inherited as an autosomal dominant trait that is characterized by a benign course and normal development. Junctional EB is inherited as an autosomal recessive trait. The disease begins at birth. Patients develop severe subepidermal blisters on the scalp and extremities, which may eventually involve the entire cutaneous surface.[3] Junctional bullosa is a progressive disease that causes mortality in early childhood.

Dystrophic EB occurs in infancy. Chronic anemia may be present. The autosomal dominant form is usually limited to subepidermal bullae on the hands, feet, and sacral area, which heal rapidly to leave soft, superficial scars.[3] Children with the autosomal dominant dystrophic form have a good prognosis. In contrast, the autosomal recessive form of dystrophic EB is the most severe form of the disease. Healing occurs with scarring and results in the characteristic deformities that include flexion contractures of the hands, feet, elbows, and knees, pseudosyndactyly, microstomia, ankyloglossia, esophageal strictures and adhesions of various skin surfaces.[5] Oral mucosa is involved in over a third of cases.[5] The overall prognosis for EB dystrophica is poor, and death usually occurs in the first three decades.[3]

Anesthesia is frequently required for dental extractions, removal of scar tissue, relief of esophageal strictures, or correction of pseudosyndactyly.[6] Meticulous attention must be paid to positioning, monitoring, airway management, and infection control when caring for patients with EB. Any mechanical contact with the patient's skin or mucosal membrane could lead to bullae formation. Padding of all surfaces in which the

patient will come in contact, as well as allowing the patient to transfer himself to stretchers and beds, reduces the risk of shearing force. Electrocardiogram monitoring presents a challenge because the adhesive patches can result in trauma with application or removal. Pulse oximetry is the preferred method of monitoring heart rate, and there are no complications reported with its use.[7] A lubricated axillary temperature probe was used in place of the more traumatic esophageal temperature probe in this patient. The patient's extremities were wrapped with an elastic bandage prior to the placement of the blood pressure cuff and tourniquet. Cycling of the blood pressure cuff does not cause the same skin trauma as a shearing force. In addition, scar formation may contribute to difficult intravenous access.

Bullae formation in the airway can result in complications, including bullae formation from application of the mask, airway instrumentation, and endotracheal intubation. Facemasks should be well lubricated to reduce trauma. The use of oropharyngeal and nasopharyngeal airways should be avoided because the squamous epithelium that lines the oropharynx and esophagus is susceptible to bullous separation.[2] In general, these are not necessary to maintain a good airway, as the tongue is relatively immobile and is drawn anteriorly by the adhesions around it.[8] The size of the endotracheal tube should be smaller than the calculated size in order to reduce the risk of bullae formation. In our case, a smaller, well-lubricated endotracheal tube was chosen to minimize the risk of bullae formation around the nare from a tight-fitting tube.

Several anesthetic techniques have been described in the literature for patients with EB. For patients in whom airway management is not anticipated to be difficult and the surgical manipulation is not extensive, infusions of propofol with either remifentanil or ketamine have been used to provide deep sedation. In patients with EB who present with a difficult airway, as described in this case report, awake fiberoptic intubation is the safest option for airway management. Fiberoptic intubation is less traumatic to the mucosal epithelium than direct laryngoscopy and should often be the first choice in EB patients with a difficult airway.[9] General anesthesia is a safe, reliable technique associated with a very low incidence of complications.[7]

Postoperatively, pain control is of utmost importance. Excellent postoperative analgesia is important to prevent thrashing in the bed or crib.[9] Children with EB can be distressed upon waking and struggle, causing skin damage; the parents of these children should be present in recovery to reassure them on waking.[10] A balance must be reached in order to achieve adequate pain relief, yet still maintain an appropriate level of mentation in order to prevent skin trauma from a somnolent, uncooperative patient. Nausea and vomiting should be prevented and treated as soon as possible in order to prevent retching, which can lead to bullae formation.

In conclusion, the patient with EB presents a challenge to the anesthesia provider. In addition to the many demanding aspects of caring for an EB patient, special attention must be paid to the psychosocial issues of living with a debilitating, painful disease. These patients are not strangers to medical institutions and can be quite wary and anxious of staff and procedures. As with any patient, a thorough airway assessment and preparation is a critical component in the delivery of a safe anesthetic.

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