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Case Report

# Anesthesia Management of the Lower Limb Amputation in an Adult Patient with Recessive Dystrophic Epidermolysis Bullosa

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## Abstract

Epidermolysis bullosa (EB) is a family of hereditary skin disorders resulting from abnormal or absent structural proteins that cause weak or absent connections between the epithelium and underlying layers of the skin and mucosa, which occurs in approximately 20 per 1 million live births. Extreme skin fragility, extracutaneous manifestations such as microstomia and laryngeal stenosis, and little experience accumulated make anesthesia management quite a challenge. Here we report the anesthesia management of the lower limb amputation in an adult patient who is the first EB patient admitted to our department during the past 30 years. The patient went through the peri-anesthesia period without emergency ventilation disorder and new bullous formation after adequate evaluation and careful planning, which may suggest that the patient with EB can safely undergo anesthesia if served meticulously enough even in non- EB specialized institutions.

**Keywords:** Epidermolysis Bullosa; Airway Management; Mucosal Integrity; Skin Protection

## Introduction

Epidermolysis bullosa (EB) is a family of hereditary skin disorders marked by extreme skin fragility, in which mild friction or shear forces on the skin and mucous membranes result in blisters and ulcers. These disorders result from abnormal or absent structural proteins that cause weak or absent connections between the epithelium and underlying layers of the skin and mucosa [1,2]. In addition to blistering and chronic wounds, there are several extra cutaneous manifestations of EB such as microstomia, laryngeal stenosis, cardiomyopathy, esophageal stricture, gastroesophageal reflux, anemia and chronic pain [3]. Common surgical procedures in adult EB patients are esophageal dilation, contracture release, cesarian section and squamous cell carcinoma resection. Both cutaneous and extra cutaneous manifestations pose challenges to anesthesia [4]. Given the rarity of EB that occurs in approximately 20 per 1 million live births [5,6], most medical institutions do not have the opportunity to accumulate experience on anesthesia in such patients. Here we report the experience on anesthesia for the first EB patient admitted to our department during the past 30 years in order to provide reference for medical workers in non- EB specialized institutions. The patient agreed to report his case and signed informed consent.

### Case Description

A 37-year-old, 45-kg man with recessive dystrophic epidermolysis bullosa (DEB) was diagnosed with chronic osteomyelitis and squamous cell carcinoma of the left foot after presenting with persistent skin infection. The patient presented to us for amputation of the left lower extremity at the level of the mid-tibia which was his first time undergoing anesthesia and surgery in April 2022. The focus of this case report is the mechanical and physical problems associated with anesthetic administration related to his EB. The special anesthetic considerations are: (1) a thorough preoperative evaluation; (2) maintaining skin and mucosal integrity; (3) airway management; and (4) postoperative analgesia.

## Preoperative Evaluation

The mouth opening-inter-incisor distance was 1.8 cm, and the tongue showed a partial fixity in the oral cavity (Mallampati score=4). Electronic laryngoscopy showed scarring of the hypo pharyngeal wall. The computed tomography(CT) and three-dimensional reconstruction of the upper airway showed no signs of obvious stricture. The patient presented with extensive skin blisters and bullous formation on the jaw, neck, back, and buttocks and was affected by mitten deformity (pseudo syndactyly) in the hands and feet. The patient was in poor nutritional status with unclear superficial venous vessels and laboratory findings suggested anemia and hyperproteinaemia. Evaluation of the airway and skin is detailed in Figure 1.


**Figure 1:** Pre-anesthesia evaluation on airway and skin. The mouth opening-inter-incisor distance was 1.8 cm (A). Electronic laryngoscopy showed spherical tongue, pharyngeal pseudomembranous and scarring of the hypopharyngeal wall (B, white arrow). Three-dimensional reconstruction of the upper airway showed no signs of obvious stricture (C). Severe pseudosyndactyly in hands (D) and feet (E), the white arrow shows squamous cell carcinoma of the left foot. Extensive skin blisters and bullous formation on the jaw, neck, shoulder, and back. (F).

## Maintaining Skin and Mucosal Integrity

Precautions were taken to minimize the risk of causing sheer stress and the development of new bullous lesions. The adhesive on the electrocardiogram (ECG) electrodes was trimmed. A wrap-around style pulse oximeter sensor was covered in transparent film dressing and then gently wrapped around the stump and held in place with gauze. The blood pressure cuff was placed on a upper extremity over medical cotton pads and noninvasive blood pressure was measured only twice, before and after induction of anesthesia. An arterial line was established for invasive blood pressure monitoring and arterial blood gas analysis after endotracheal intubation. Two experienced nurses tried to establish peripheral intravenous access, but they failed. Finally, a right femoral central venous catheter was placed under ultrasound guidance by the anesthesiologist. Arterial and central venous cannula were sutured in place and protected with gauze. All airway equipment and adjuncts were well lubricated. Facemask in smaller sizes was generously lubricated with petrolatum to minimize friction and blister formation on the face. Additionally, the endotracheal tube was carefully secured with a special endotracheal tube retainer consisting of a dental pad and a wide soft fixing strap; petroleum gauze and padding were placed where the retainer touched the face to minimize trauma. The patient, s eyes were protected with a disposable medical hydrogel eye patch. The measures for the protection of the skin and mucous membranes are shown in Figure 2.

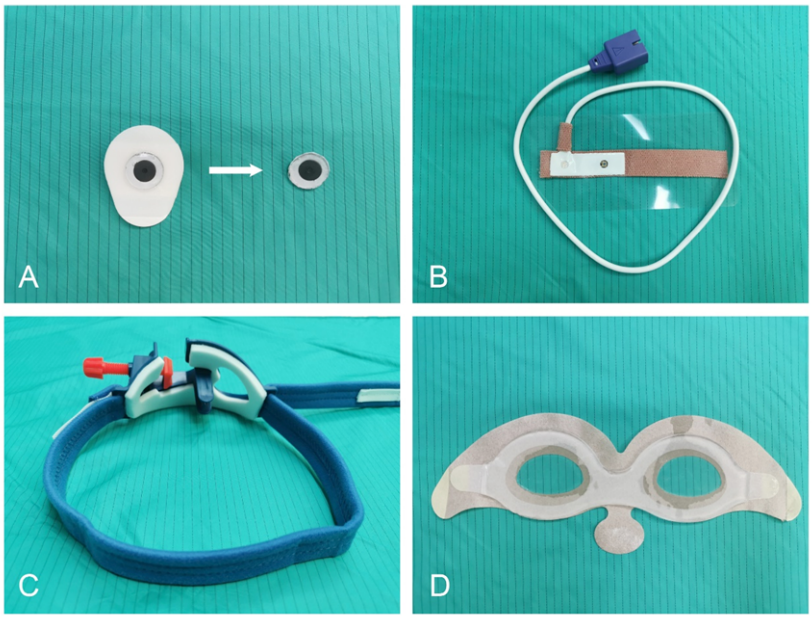


**Figure 2:** Precautions taken to minimize the risk of causing sheer stress. Adhesive on the ECG electrodes was trimmed (A). Wrap-around style pulse oximeter sensor was covered in transparent film dressing before use (B). Special endotracheal tube retainer with foam pad on the side that touches the skin around the mouth and a wide soft fixing strap to make a circle around the neck and face (C). Disposable medical hydrogel eye patch used to prevent exposure keratitis and inadvertent eye friction (D).

When transferred, the patient was levitated, with the aid of a sheet placed under him, moved over, and placed back down without any touching and dragging. Lidocaine was sprayed into the airway before emergence to increase tolerance to endotracheal tube and prevent restlessness during awakening. The endotracheal tube was not removed until the patient's muscle strength was fully recovered and consciousness was completely restored. A soft suction catheter for oral and gastric suctioning was available just in case and the patient was encouraged to spit out pharyngeal secretions himself. The patient went through a safe and smooth emergence without any new bulla formation. Laryngopharynx and tracheal topical anesthesia was performed by nebulization of 2% lidocaine 15 minutes before anesthesia, and then adequate preoxygenation was achieved using 100% oxygen , with a fresh gas flow rate of 6 L/min and breathing for 5 min [7]. Devices for endotracheal intubation under the guidance of flexible fiber-optic bronchoscopy and emergency tracheotomy were at hand. Intravenous propofol 90 mg were injected within 10 seconds to make the patient unconscious and apneic for about 2 minutes, during which an experienced anaesthesiologist tried to gently insert a well-lubricated, smaller endotracheal tube (size 6.0-mm internal diameter) into the glottis using a video laryngoscope; the Cormack-Lehane grade was 4 under direct laryngoscopy views of the glottis and the patient was successfully intubated at the first try. As soon as the assistant confirmed the position of the endotracheal tube by auscultation of both lungs, a bolus administration of 36 mg rocuronium, 27 mcg sufentanil and 2 mg midazolam was performed followed by intravenous remifentanil at 0.2-0.3 μg/kg/min and continuous inhalation of sevoflurane for anesthesia maintenance. The whole intubation process was smooth without any coughing or struggle.

## Postoperative Analgesia

Sufentanil 15 mcg and ketorolac tromethamine 30 mg were injected intravenously during sutures to prevent pain sensitization caused by remifentanil. The wound was locally infiltrated with ropivacaine of 0.2%, 100mg before covered by sterile dressings. Prophylactic antiemetics and antipruritics were given. Patient-controlled intravenous analgesia (PCIA) was implemented with a specialized device containing 100ml solution of sufentanil, dezocine and tromethamine postoperatively, which was then switched to oral analgesics after oral intake resumed.

## Discussion

Airway management is regarded as one of the most challenging parts of caring for patients with EB because prevention of shear forces and management of difficult airways seem to be fish and bear's paw [8-11]. These patients will likely

have a very limited mouth opening, dental caries, and restriction of neck movement owing to recurring contractures. Even so, safe airway management can be achieved if carefully prepared [12]. Limited mouth opening, dental caries, tongue fixation and skin lesions on the face of this patient suggest that his endotracheal intubation is a challenging task. Preventing the pharyngeal and laryngeal formation of new blisters as a result of friction and shear forces is crucial and the ideal anesthesia regimen should be a combination of regional anesthesia and sedation to avoid potential complications associated with airway manipulation [13]. However, the skin of his spine and buttocks is ulcerated and infected, which is a contraindication to regional anesthesia for lower extremity. Supraglottic airway is dismissed due to microstomia, intraoral scarring and ankyloglossia [12]. General anesthesia with endotracheal intubation was ultimately adopted due to the operative duration and risk of aspiration. Sober intubation guided by fiber-optic bronchoscope is the most guaranteed scheme for difficult airway, but may cause discomfort and bucking which will increase the risk of apnea resulting from probable laryngopharyngeal blistering, edema and bleeding for EB group. Direct intubation with video laryngoscopy, the most convenient and skilled method for anaesthesiologists has been successfully used in patients with EB [8,14]. In order to avoid the suffocation after receiving muscle relaxants and skin trauma caused by firm mask oxygen ventilation as much as possible, we have developed an anesthesia protocol that the short-term loss of consciousness and transient breathlessness is achieved by a bolus administration of 2 mg/kg propofol followed by one single shot at endotracheal intubation. Successful attempt is exactly what we want. If not, we can calmly start the plan of sober intubation without any concern and anxiety on oxygenation because the patient is breathing and consciousness will soon be restored. Besides, difficult airway equipment including supplies for emergent surgical airway is readily available in case of an emergency airway situation. It has been reported that high-flow nasal oxygen (HFNO), an oxygen delivery method that is very suitable for patients who need to avoid hand-controlled ventilation with a closed mask after the induction of general anesthesia such as EB, is superior to conventional oxygenation in reducing the risk of O2 desaturation and increasing safe apnea time [15,16]. We used the traditional preoxygenation method because there was no equipment available, but it also worked well. Intravenous access for EB is often difficult and may require ultrasound guidance [17]. The patient presented with severe fingers/toes deformity and extensive skin damage, which makes it more difficult to establish intravenous access. Consequently, we established central venous access under ultrasound guidance to facilitate intraoperative blood transfusion and fluid rehydration. An arterial line was also established for real-time blood pressure monitoring and arterial blood gas analysis to reduce upper arm skin injuries caused by frequent non-invasive blood pressure monitoring. Central venous and arterial catheterization was sutured in place and wrapped with gauze to avoid shear forces when removed. Patients with EB may have corneal erosions, keratitis, and blepharitis, etc. Careless touching and prolonged corneal exposure during surgery can aggravate pre-existing ocular complications [18]. We used hydrogel eye patches which were designed to moisturize the eyes to avoid exposure keratitis and unintentional eyelid or eyeball rubbing and the sticky edges are trimmed off before use. A smooth postoperative course is important for EB patients because agitation and excess patient movement may lead to the formation of new blisters and lesions, which partially depending on multimodal pain management to control the acute exacerbation of chronic pain in EB patients [3]. Longitudinally, we first did analgesic articulation with sufentanil to prevent pain sensitization caused by discontinuity of refentanil, then performed local infiltration with ropivacaine around the wound, and then provided PCIA. Later, the patient switched to oral analgesics prescribed by the pain specialists when oral intake was resumed. Horizontally, we used a multiple combination of local anaesthetics, opioids, and non-steroidal anti-inflammatory drugs for postoperative analgesia. Intravenous opioids may lead to increased itching, nausea, and vomiting. Therefore, antipruritic and antiemetic drugs were given in advance. Although the patient go through the peri-anesthesia period without emergency ventilation disorder and new bullous formation, there is also something inappropriate in what we do. One is that we are petty-minded and obsessed with details to reduce the frequency of non-invasive blood pressure measurements. However, it has been reported that non-invasive blood pressure measurement is safe for patients with EB because it does not apply friction and shear forces [17]. In addition, it is our first time to care for EB patients, lack of experience and over-meticulosity resulting in prolonged induction time.

## Conclusion

Patients with EB can safely undergo anesthesia with minimal morbidity with a thorough preoperative evaluation, careful planning and vigilance, utmost care to avoid shear forces or friction on skin and mucosal surfaces, even in institutions that have never cared for such patients.

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