

Anesthetic Approach to a Patient with Dystrophic Epidermolysis Bullosa: A Case Report

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Distrofik Epidermolisis Bullosalı Hastaya Anestezik Yaklaşım: Olgu Sunumu

ABSTRACT

Epidermolysis bullosa (EB) describes a rare group of diseases characterized by vesiculobullous lesions and scars which complicate anesthesia practice in terms of airway management and occurrence of new lesions due to friction.

A 5-year-old boy with the diagnosis of dystrophic epidermolysis bullosa (DEB) admitted for an elective release of pseudosyndactyly and circumcision operation. There were lesions on nearly all parts of his body.

Monitorization was performed only with electrocardiograms (ECGs) with the minimal sticky electrode surface and arterial oxygen saturation. A well-lubricated I-gel was preferred for the maintenance of airway. At the end of the surgery, the patient was awakened without any airway problem and new bullae formation, except erythema at electrode adhered regions.

We believe that the use of I-gel is a good alternative in these patients because it is necessary to protect the skin and oropharyngeal mucosa from trauma in order to prevent the formation of new lesions and to limit the monitoring as much as possible in patients with EB.

Keywords: Epidermolysis bullosa, general anesthesia, I-gel, airway management

Öz

Epidermolizis bülloza (EB), anestezi uygulamalarını hava yolu yönetimi ve sürtünmeye bađlı yeni lezyonların oluşması bakımından komplike hale getiren, vezikülobüllöz lezyonlar ve skar dokuları ile karakterize, nadir görülen bir grup hastalığı tanımlar.

Distrofik epidermolizis bülloza (DEB) tanısı olan 5 yaşında 1 erkek çocuk, elektif olarak pseudosindaktili düzeltilmesi ve sünnet için ameliyata alındı. Vücudunun neredeyse tamamında lezyonlar mevcuttu.

Monitörizasyon, yapışkan yüzeyin minimal tutulduğu elektrokardiyogram ve arteriyel oksijen saturasyon takibi ile sınırlı tutuldu. Hava yolu devamlılığı için iyice kayganlaştırılmış I-gel tercih edildi. Cerrahinin bitiminde hasta herhangi bir hava yolu sorunu yaşamadan ve elektrot yapışma yerlerindeki eritemden başka yeni bir lezyon oluşmadan uyandırıldı.

EB'lı hastalarda monitörizasyonun olabildiğince kısıtlanması ve yeni lezyon oluşumunu önlemek için deri ve orofarengeal mukozanın travmadan korunması gerektiğinden I-gel kullanımının bu hastalarda iyi bir seçenek olacağı kanaatindeyiz.

Anahtar kelimeler: Epidermolizis bülloza, genel anestezi, I-gel, hava yolu yönetimi

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BACKGROUND

Epidermolysis bullosa (EB) is an umbrella term which describes a group of hereditary disorders resulting from functional deficiency of structural proteins of the dermo-epidermal junction with a prevalence of 1/30000-1/50000⁽¹⁾. Dystrophic epidermolysis bullosa (DEB) is a subtype characterized by vesiculobullous lesions that arise spontaneously or in response to minimal trauma which leads to subsequent bleeding, secondary infections and healing ulcers⁽²⁾.

Restricted mouth opening and oesophageal stricture due to scar formation, malnutrition, dehydration, anemia, electrolyte imbalances are the other clinical presentations of the disease⁽³⁾. Bullae at larynx and pharynx mucosa and temporomandibular joint involvement can complicate airway management^(2,4).

In this case report, we discuss the anesthetic management of a 5-year-old child with DEB who underwent corrective surgery for pseudosyndactyly and circumcision under general anesthesia.

CASE PRESENTATION

A 5-year-old boy weighing 12 kg was admitted for an elective release pseudosyndactyly correction and circumcision due to phimosis-like scar tissue.

On physical examination there were bullous lesions at various stages of healing all over the body and pseudosyndactyly between 2nd and 3rd digits of his left foot (Figure 1). Cardiovascular and respiratory systems were unremarkable. Therefore, only dermatology consultation was requested for skin protection. We were advised on the use of silicon-based material and lubrication. His mouth opening was not restricted and Mallampati score was I. After parents' consent obtained, operation is programmed.

In the operating room, all staff members were informed about the disease to minimize skin frictions. He had a venous access dressed with a silicon-based non-adhesive tape. An impregnated gauze was placed beneath the pulse oximeter probe (Figure 2). ECG electrodes were trimmed to minimize the sticky surface (Figure 3). NIBP was not applied as there were severe wounds on his upper extremities



Figure 1. Pseudosyndactyly between digits.



Figure 2. Placement of the pulse oximetry probe.

He was preoxygenated with a lubricated face mask without pressure. After induction of anesthesia a well-lubricated silicone-based I-Gel no: 1,5 was

inserted gently and fixed with an elastic ribbon gauze. His eyes were also protected with a methylcellulose-based ocular lubricant. Anesthesia was maintained with sevoflurane in 50% O₂ and 50% air. Paracetamol 10 mg kg⁻¹ was used for postoperative pain relief. At the end of the operation I-Gel was removed and oral aspiration did not applied to avoid trauma. During postoperative 48 hours any respiratory problem and new bullae formation were not observed except erythema at the regions where electrodes were adhered.



Figure 3. Inserted and fixed I-gel.

DISCUSSION

Inherited EB represents a group of disorders that are characterized by spontaneous or traumatic development of blisters over the skin⁽¹⁾. Preoperative medical assessment for difficult ventilation and intubation is imperative and history taking and physical examination, focusing on comorbidities, such as amyloidosis, and neuromuscular diseases, besides clinical presentation of the disease, like growth retardation, anemia, hypercoagulopathy, electrolyte imbalances, hypoalbuminemia and also infections are essential⁽³⁾. Laboratory tests should be ordered. Consultations from departments of pediatrics, dermatology and surgery should be ideally requested to improve patients' health⁽²⁾.

Our patient was diagnosed as DEB four years previously, and there were not any clinical abnormality except for specific lesions and cachexia.

General, regional or both anesthetic techniques can be used⁽⁵⁾. The ultimate goal is patient's safety^(6,7).

In the operating room, skin trauma and airway management are main challenges⁽²⁾. Owing to skin fragility, monitoring and positioning requires taking special measures⁽⁸⁾. We brought our patient gently to the operating table with sheets and preferred minimal monitoring and atraumatic airway management with I-gel. Hereby, apart from erythema at electrode-adhered regions perioperative period passed without any problem.

Difficult ventilation and intubation are expected in EB patients, due to limited mouth opening, temporomandibular joint involvement and oropharyngeal scarring^(2,4-6). Griffin et al.⁽⁴⁾ reported 10 difficult cases of intubation among 390 general anesthesia procedures performed for 44 EB patients within a period of 10 years. In another study, Heuvel et al.⁽⁵⁾ reported 25 difficult cases of intubation in 79 EB patients, where two cases were not anticipated. Therefore, although predictive test results were within normal limits, we have been prepared for potential difficulties.

The induction of anesthesia is preferably performed through intravenous route but, induction with inhalation anesthesia is also possible in cases with difficult venous access⁽⁷⁾. Our patient has come to operating theatre with a venous access, so we used intravenous route for induction.

Propofol seems the best choice for induction. Knowledge about ketamin remains controversial because of its excitatory and hypersalivation effects; increased porfira risk should be kept in mind when using thiopental. Non-depolarizing muscle relaxants should be preferred as neuromuscular diseases can be accompanied with EB and also fasciculations due to depolarizing muscle relaxants may increase trauma to the skin^(2,9). Due to possibility of encountering difficult airway, rocuronium should be the first choice and sugammadex should be kept on hand⁽⁶⁾.

On the other hand, in the choice of an inhalational agent for induction, sevoflurane is the best alternative⁽²⁾. Although use of a smaller endotracheal tube has been suggested⁽²⁾, in a study, authors reported postoperative laryngeal lesions with their use in the presence of an intact trachea⁽⁸⁾. However, laryngeal involvement is too rare as its epithelium is more

unvulnerable than oropharynx and relationship with intubation have not been demonstrated ⁽⁹⁾. Oropharyngeal lesions and vocal cord thickening can cause extubation problems ⁽⁷⁾. Ames et al. ⁽¹⁰⁾ reported only one new lingual lesion in a study group of 57 patients in whom laryngeal mask airways were applied. In a study which compared I-gel and laryngeal mask airway-Proseal, I-gel was found less traumatic with lower airway sealing pressure ⁽¹¹⁾. Therefore, we preferred I-gel.

Although use of multimodal analgesia with opioids and nonsteroidal anti-inflammatory drugs is suggested ⁽⁴⁾, it should not be forgotten that low albumin levels might alter the pharmacokinetics of drugs ⁽¹²⁾. As our patient was cachectic, to avoid postoperative respiratory problems, we preferred paracetamol for postoperative analgesia which was satisfactory.

Children with EB often have to undergo surgical procedures and should be evaluated in terms of disease's characteristics and comorbidities, preoperatively. Airway management and minimizing trauma are the key to the anesthesia. In order to prevent emergence of new lesions, I-gel seems a good choice for airway management, because of its soft consistency.

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Informed Consent: Written informed consent was obtained from the parents for publication case report

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