

Anesthetic Management In Pediatric Patients With Epidermolysis Bullosa

Julio Cezar Mendes Brandão^{1*}, Fernando Vicente de Araujo², Luciana Sales de Araujo³, Patricia Sanches⁴, Itamar Souza de Oliveira Junior⁵

¹Postdoctoral Research Fellow, Harvard Medical School; Administrative Director, Sergipe Society of Anesthesiology - SAESE; Member of Perioperative Medicine Commission of Brazilian Society of Anesthesia(SBA); Anesthesiologist staff at Federal University of Sergipe, Brazil

²Member of the Brazilian Society of Plastic Surgery, Plastic Surgeon at Hospital São Lucas, Sergipe, Brazil

³Active member of the Brazilian Society of Anesthesiology, Member of the Sergipe Society of Anesthesiology and anesthesiologist at São Lucas Hospital, Sergipe, Brazil

⁴Member postgraduate committee in surgery of Federal University of Minas Gerais, Brazil

⁵Professor at Discipline of Anesthesiology, Pain and Intensive Medicine, University Federal of São Paulo, Brazil

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*Corresponding author: Julio Brandão, Federal University of Sergipe (UFS), São Cristóvão, SE, Brazil, Fax : 5579988552321; Tel.: 5579988552321; Email: juliobrand@hotmail.com

Summary

Introduction: Anesthetic therapy in patients with epidermolysis bullosa has several peculiarities. Some choices in the anesthetic procedure can be understood as imprudent, but such isolated or unrelated measures can make a difference in saving them.

Report: The patient presented intermittent remission / exacerbation with infected lesions on the skin. A total of 37 anesthetic / surgical procedures were performed, and the main alterations and peculiarities related to the difficulty of monitoring, obtaining peripheral venous access, hypothermia, analgesia and difficult airway were described.

Conclusions: Hypothermia, difficult airway, difficulty in venous access and monitoring, and infection were frequent problems. Dealing with these procedures repeatedly required of the team a series of adaptations and extrapolations in the absence of data in the literature.

Keywords: Epidermolysis Bullosa; Anesthesia; Hypothermia;

Introduction

Epidermolysis Bullosa (EB) are congenital bullous dermatoses that are described in a rare group of genetic disorders. These have as a common characteristic the development of blisters or vesicles at the least mechanical trauma, or even the separation of the tissues at the lowest friction. It is a rare disease, with an incidence of 1: 300,000, approximately. Patients present involvement of cutaneous and extra-cutaneous structures, as well as mucosal alterations [1,2].

Three groups of the disease are recognized, according to the second international consensus: epidermolysis bullosa simple, junctional and dystrophic1. The case reported is epidermolysis

bullosa of the dystrophic type and the main aspects of the surgical anesthetic outcome will be addressed, besides the particularities of the anesthesia and pre-anesthetic evaluation.

Case Report

A 6-year-old pediatric patient diagnosed from birth with intermittent remission / exacerbation of the disease, presenting with advanced dystrophic lesions in the upper and lower limbs, including hands and feet with pseudoindactyly. No family history.

He was hospitalized for sepsis secondary to infected skin lesions, and after initial therapy for the treatment of sepsis, a surgical approach was programmed for debridement of the lesions. The skin surface without lesions was very small (estimated between 20% in the procedure with the highest percentage of skin compromised and 80% in the procedure with the lowest percentage of compromised skin).

During the hospitalization period (4 months), 37 anesthetic procedures were performed to perform the debridements, and the patient was cooperative. These procedures were performed by 4 different anesthesiologists, but with very similar anesthetic techniques.

Pre-anesthetics were administered in all procedures with oral midazolam 0.5 mg / kg about 30 to 40 minutes before anesthetic induction, monitored with pulse oximetry, capnography, and blood pressure measurement by noninvasive method.

Inflatable silicone disposable facial masks with vaseline were used on the patient's skin contact surface. There was a need for adaptation and placement of petroleum jelly on various contact

surfaces to avoid further skin lesions. In two of the procedures, the patient had no venous access and no attempt was made to obtain their access. In 10 procedures the patient had a deep venous access and in another 25 had peripheral venous access, through which the anesthetic and adjuvant medications were administered.

In the procedures in which no venous access was obtained, the following medications were used intramuscularly: ketamine at a dose of 3-5mg / kg, fentanyl 1-3mcg / kg, morphine 100mcg / kg and atropine 15mcg / kg. Inhaled induction with nitrous oxide (N₂O), sevoflurane and oxygen was used, being maintained in spontaneous ventilation with facial mask in semi-open circular circuit. Ketamine 1mg / kg, fentanyl 1-3mcg / kg, atropine 10-15mcg / kg, ondansetron 100-200mcg / kg, dipyrone 30-

50mg / kg and tramadol were used intravenously. 1mg / kg, diphenhydramine 1mg / kg and dimenhydrate 100-150mcg / kg, with small variations in doses and medications in most of the procedures performed.

In several procedures, scarification areas were observed at the palpation sites for the adequacy of the facial ventilation mask and contact areas with rigid surfaces (Figure 1A and B). There were no significant interurrences during the procedures and the patient woke up without pain symptoms in most procedures. Laryngeal masks and fibroscopy were kept available for any complication, given the possible difficulty in the management of the airway. After a few months of the last surgery, the patient presented septic shock secondary to infection of some of the wounds and died.



Figure 1: A

Figure 1: B

Figure 1: Scarification areas. In A: at the palpation sites for adequacy of the facial ventilation mask; In B: places of contact with rigid surfaces. Disclosure of the photos was allowed by means of the TCLE provided by the mother.

Discussion

The appropriate approach during the pre-anesthesia evaluation may anticipate a number of problems during the anesthetic procedure. Depending on the lesions, venous punctures can be difficult, making it even more difficult to induce anesthesia. In viable cases care should be taken with the dressing chosen in the venoclysis and with its retention so that a new wound and new point of infection is not created. In the case of this patient (37 interventions in 4 months) the peripheral veins became scarcer each day, in addition to some places of puncture have become foci of new wounds.

Thus the inhalation induction was an option, and care with the use of the facial mask was carefully obeyed. The use of gauze soaked in petroleum jelly or other protectors may decrease the chance of the appearance of lesions on the face. In cases where orotracheal intubation is performed, it should be borne in mind that tube fasteners may cause injury to the face and therefore it is usually chosen not to attach or use an external fixation.

Patients with gastro-oesophageal reflux disease should intubate in a rapid sequence. These patients may still have risk factors for a difficult airway, and all preparation and planning should be done to avoid surprises that could be disastrous. These

are patients who may very often have neck injuries and these can make it difficult to extend the head to align the axes of the airway, an important factor for the success of orotracheal intubation.

Nasal intubation should be avoided because of possible minor trauma in this region. Patients with EB are in the group of patients at risk for subglottic stenosis due to chronic lesions and the number of procedures that are exposed in childhood. If it is necessary to use a smaller orotracheal tube, care should be taken when inflating the tube balloon. The use of high pressures is a predictor of mucosal and tracheal lesions, which may lead to subglottic stenosis. If the flask is inflated, it is recommended to use the lowest possible pressures, sufficient to avoid leaks. More modern pediatric tubes now require up to 10cm H₂O to seal the airway. It should also be remembered fundamental points that must be taken into account: microstomia, often associated and injuries; Fragile or even absent dentition; The fragility inherent in the skin and mucous membranes; Ankyloglossia and subglottic stenosis [2].

It is still recommended that, in cases where general anesthesia can be avoided, sedation or regional anesthesia may be used because of possible complications with airway mucosa [3].

It is possible to perform neuraxial blockade or peripheral

blockade, however it should be kept in mind that the cleaning and antiseptic process itself may cause a new lesion in the place where the puncture was performed. Therefore, great care must be taken not to injure the skin at the time of preparation for puncture, as well as contraindication due to injury or infection at the place of insertion of the needle to perform the peripheral or central block [3].

For monitoring, the recommended parameters for each type of anesthesia and procedure should be maintained, according to resolution CFM 1802/2006 (Brazil). However, it is important to fix the intravenous catheters and electrodes of the electrocardiogram with special products based on silicone or non-adherent products, since the usual fixers are sufficient to promote cutaneous lesions in some patients. In the absence of these products it is possible to choose to soak the normal dressing in vaseline, thus reducing the chance of scarification of the skin at the time of removal of the fixatives. It was observed that the procedures in which vaseline was used there were less lesions consequent to manipulation. The use of biopolymer membranes, hydrogel, biosynthetic cellulose and silicone gels are very useful for wound protection and care during placement in different beds for procedures [4].

The sphygmomanometer can be placed on a layer of wool because it reduces contact with the surface of the device, promoting less local trauma. To protect the eyes, ophthalmic gels may be used and arterial puncture may ensure fixation with a suture point or tightly attached fixatives, but the risks of losing arterial line with the latter may be significant. Eventually one should consider not using any of the monitors in some patients when they may bring more damage. This is not an easy choice and must be analyzed in the face of each situation, given the risks that may arise in the event of non-monitoring of any important parameters for anesthesia [4].

Since these patients are generally treated with chronic pains, postoperative analgesia should include this aspect, as well as changes in drug metabolism and possible associated hyperalgesia. There are reports of EB with a need for a surgical approach also in newborns, as shown in the editorial of *Anesthesiology* of Anthony et al. [5], which may amplify the expected and described problems for anesthetic care. Specific pain management and multidisciplinary pain team care may bring less morbidity to these chronic patients, especially in patients who can commonly use a large variety of analgesics, in addition to the eventual use of tricyclic antidepressants [1,5].

There is still no standardized set of anesthetic materials available in patients with EB, but these factors must be taken into account and appropriate care should be taken with manipulation, monitoring, maintenance of homeostasis and special analgesia for these cases. Individualization of care [2,4].

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