

Anesthetic management of epidermolysis bullosa : a review and report of two cases

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Abstract : Dystrophic epidermolysis bullosa is a rare genetically mechanobullous disorder, characterized by an excessive susceptibility of the skin and mucosa to separate from the underlying tissues after mechanical trauma. Avoiding mechanical stimulation of the skin and mucous membranes is essential in the anesthetic management. These case reports describes bilateral nerve block in two patients with an anticipated airway difficulty due to epidermolysis bullosa. We report the successful applications of intraoperative indirect electrocardiogram monitoring without electrodes and bilateral axillary and midhumeral nerve block using levobupivacaine in combination with lidocaine in patients who had previously undergone repeated general anesthesia. We conclude that regional anesthesia should be preferred in patients with dystrophic epidermolysis bullosa, especially those who had previously undergone repeated general anesthetic procedures.

Key words : Axillary nerve block ; midhumeral nerve block ; epidermolysis bullosa ; levobupivacaine ; lidocaine ; monitoring.

INTRODUCTION

Epidermolysis bullosa (EB) describes a group of genetically determined mechanobullous disorders that vary in course and severity, ranging from relatively minor disability to death in early infancy (12). “*Dystrophic epidermolysis bullosa*” (DEB) is caused by a defect in type VII collagen, where marked scarring leads to deformities of extremities (2). Oral and pharyngeal mucous membranes may form adhesions, which may result in ankyloglossia, microstomia and narrowed oral vestibule. Airway control and endotracheal intubation may be difficult secondary to contractures due to previous repeated anesthetic managements (3, 12). The challenge is to maintain patency of the airway and make effective use of the monitoring device without damaging epithelial surfaces, which may result in permanent scarring. When caring for patients with DEB, precautions to protect the integrity of the skin from trauma, friction injury and adhesive products are essential (3, 14). There is a paucity of data in the

literature on the perioperative management of dystrophic epidermolysis bullosa for both general (7, 9) and regional anesthesia (4, 13). Regional anesthesia is especially suitable in patients with DEB when a difficult airway is anticipated due to prior repeated airway manipulation during general anesthesia.

These case reports describe successful use of regional anesthesia under light sedation and indirect monitoring of electrocardiogram (IME).

CASE REPORT

The patients had typical features of severe DEB, including extremity contractures, severe digit deformity, extensive blisters and broken skin with denuded areas. Typically, they had fusion of all fingers and toes into mittenlike deformities (pseudosyndactyly) (Fig. 1). Previously, the first case had undergone four surgeries and the second case even six surgeries, under general anesthesia. Both patients were admitted to our hospital for elective plastic surgery for pseudosyndactyly repair. Examination of the patients revealed oral bullae and lingual adhesions resulting from previous airway management during general anesthesia. Microstomia was observed and tongue mobility was limited by scarring.

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Fig. 1. — Hands of first case with DEB progression to produce severe scarring and pseudosyndactyly.



Fig. 2. — Limited mouth opening is shown in second case

In both patients, the Modified Mallampati score was 4 and mouth opening was limited i.e. 1.5 and 1.3 cm respectively (Fig. 2). After informed consents were obtained from the parents, all pressure points were cushioned on the operation table and non-invasive blood pressure monitoring was avoided to prevent bullae formation. We removed all adhesive parts of the pulse oximetry probe, then attached the probe to the earlobes of patients covered with vaseline. We carried out the IME without attaching adhesive electrodes to the skin. This was made possible by placing three patches of wet cloth soaked in physiological serum on the anterior thorax. Both patients received phenytoin, a collagenase inhibitor, polivitamin and enteral nutrition product.

First case

A 15-year-old male, weighing 25 kg and with a height of 144 cm, was first diagnosed with DEB at the age of one. Continuous intravenous infusion of propofol ($1-1.5 \text{ mg.kg}^{-1}.\text{h}^{-1}$) for light sedation was carried out during axillary nerve blocks. Both axillae were free of bullae formation. A 25 mm, 24G cannula (Stimuplex® A, B Braun Melsungen AG, D-34209 Melsungen, Germany) were used to perform a brachial plexus block by the axillary approach. Using a nerve stimulator, a bilateral block was achieved with levobupivacaine (Chirocaine amp 0.375%, 7.5 ml at both sides) and lidocaine (Aritmal amp 1%, 5 ml at both sides) combination.

Second case

A 12-year-old male, weighing 24 kg and with a height of 139 cm, was first diagnosed with DEB

at the age of six. Continuous intravenous infusion of ketamine ($0.5-2 \text{ mg.kg}^{-1}.\text{h}^{-1}$) for sedation was carried out during peripheral nerve blocks. Because there were bullae formations at both sides, the mid-humeral approach was selected. Bilateral median, radial and ulnar nerve blocks were carried out with the 25 mm, 24G cannula (Stimuplex® A, B Braun Melsungen AG, D-34209 Melsungen, Germany), using a nerve stimulator. Levobupivacaine (Chirocaine amp 0.375%, 5 ml at both sides) and lidocaine (Aritmal amp 1.5%, 5 ml at both sides) combination.

In the two cases the courses of both anesthesia and surgery were uneventful. In the postoperative period, first analgesic requirement was four hours later and the patients were discharged without any complications.

DISCUSSION

DEB is a genetic disorder which causes blistering and shearing of the skin following even minor trauma, and leads to an array of medical problems. Patients with this condition also suffer from blisterings in mucosal membranes, particularly in esophagus, which leads to strictures and failure of oral feeding. Esophageal blistering also presents a risk of pulmonary aspiration due to regurgitation or reflux. These patients may also have scarring, which results in microstomia, limited opening of the mouth, and limited temporomandibular joint motion. Nasal, oral, laryngeal and tracheal manipulations should be kept to a minimum to avoid airway damage.

With respect to medical therapy, phenytoin, a collagenase inhibitor in the fibroblasts, may provide

short-term improvement (2). Our patients too had been receiving phenytoin. In these cases, right and left hand surgeries were performed in a single session, in accordance with the preferences of both the patients and the surgical staffs. Many authors recommend that patients with DEB are brought to the operation theatre fully awake to assist in optimal and safe positioning (1). Accordingly, our patients were not premedicated.

Minimal monitoring is advocated in these patients due to the risks described above. Placement of pulse oximetry probe on the finger was difficult due to pseudosyndactyly, so the probe was placed on the earlobe instead. It is known that electrocardiogram monitoring is difficult because the adhesive electrodes may cause skin trauma on placement and removal. Whenever possible, surgical tape and adhesive electrodes should be avoided.

Previous studies have reported a number of solutions to the challenge of electrocardiogram monitoring in patients with DEB. These solutions include the placement of adhesive electrodes onto defibrillation pads (1), the removal of the adhesive parts of electrodes (6) and placement of electrodes on the underside of a chest radiograph placed underneath the patient (2). OHARA *et al.* (10) have verified the feasibility of a new indirect method and recommended its usage in patients with epidermolysis bullosa as well as cases of severe burns or severe atopic dermatitis. LIM *et al.* have developed a new indirect contact electrocardiogram measurement method for monitoring electrocardiogram during sleep, which did not require any direct conductive contact between the instrument and bare skin (8). None of these reports had actually used the indirect monitoring method in a patient with DEB.

In our patients, we carried out the IME without attaching adhesive electrodes to the skin. This was made possible by placing three patches of wet cloth soaked in physiological serum on the anterior thorax. Our method is similar to but not identical to those used by other authors. We selected physiological serum above water because of its better conductivity. We conclude that this modified indirect monitoring method may be more useful in patients with DEB.

In our cases, non-invasive blood pressure monitoring was avoided to prevent bullae formation. Intra-arterial blood pressure monitoring has been recommended as the method of choice over non-invasive monitoring (3). However, we avoided intra-arterial blood pressure monitoring; as DEB-related scarrings in the extremities would have made insertion into the radial artery difficult.

We preferred regional anesthesia in these cases for three reasons. First of all, our patients indicated that they would prefer a regional technique above general anesthesia, as their past surgeries were followed by postoperative complications such as dysphagia and severe pain in the oropharyngeal region. Secondly, they had microstomia, lingual adhesions, limited mouth opening and immobile tongue, all of which would complicate airway management. Lastly, the type of surgery was suitable for a regional technique.

Cooperation of the patient is required in order to secure an optimal position on the operating table and minimize pressure or friction to the skin, which is made possible only with regional anesthesia. Peripheral nerve blocks can be an option in patients with this rare disease, as they can minimize the number of repeated general anesthetic sessions needed and eliminate the problems faced in airway handling and potential subsequent mucosal lesions. We preferred a regional technique and even though single stage bilateral hand surgery requires a large amount of local anesthetics, the total amount used in our cases did not exceed the toxic threshold for either levobupivacaine and lidocaine. Although convulsions following axillary brachial plexus blockade with levobupivacaine have been reported (5, 11), we did not observe any convulsions in our patients after receiving levobupivacaine. This may be due to the fact that our patients were under phenytoin treatment, an anticonvulsant, and that the local anesthetic doses were far below the toxic threshold.

In conclusion, applications of indirect monitoring of electrocardiogram without adhesive electrodes and bilateral peripheral nerve block using levobupivacaine in combination with lidocaine are effective in avoiding new blisters and may ensure more safety in patients with DEB. Successful anesthetic management is possible while being prepared to encounter all potential problems and protecting the integrity of skin and mucosa from trauma, friction injury and adhesive products.

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