

CASE REPORT

## Successful use of ketamin combined with remifentanil in two patients with epidermolysis bullosa

### *Epidermolizis büllozalı iki hastada remifentanil ile ketaminin birlikte başarılı kullanımı*

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#### ABSTRACT

Epidermolysis Bullosa (EB) is characterized by the presence of extremely fragile skin and painful blister formations in the skin. We present the anesthetic management of 15-years-old, twin sisters with EB during the syndactyly operation. Premedication was done with 0.5 mg/kg nasal midazolam. Anesthesia induction was performed with sevoflurane without touching the facial masks to the skin. After intravenous 2 mg/kg ketamine and 50 µg fentanyl, anesthesia was maintained by intravenous 1 mg/kg/hr ketamine and 0.1 µg/kg/min remifentanil. We injected 2 mg intravenous morphine for postoperative pain control. Ketamine with remifentanil seems to be effective, simple, and safe for airway management and practical way to provide general anesthesia and analgesia of EB. *J Clin Exp Invest* 2012; 3 (3): 395-397

**Key words:** Epidermolysis bullosa, ketamin, remifentanil, anesthesia

#### INTRODUCTION

There are more than 20 different described subtypes of epidermolysis bullosa. Recessive Dystrophic Epidermolysis Bullosa (RDEB), the most common subgroup of epidermolysis bullosa, is characterized by the presence of extremely fragile skin and painful blister formations in the skin in response to minor trauma, friction or pressure.<sup>1</sup> Other common features that can be seen are extremity contractures, severe digit deformities, difficult airways and severe wound infections.<sup>2</sup>

The common surgical procedures for patients with RDEB are balloon dilation of esophageal strictures, pseudosyndactyly release with or without skin graft; postsurgical or skin care related dressing changes, percutaneous endoscopic gastrostomy tube placement and circumcision.<sup>1</sup> Because of the

#### ÖZET

Epidermolizis bülloza (EB) aşırı hassas cildin oluşu ve ciltte ağırlı sıvı kabarcıkları oluşumuyla karakterizedir. Epidermolizis bülloza'lı 15 yaşında ikiz kız kardeşlerin sindaktili operasyonu sırasındaki anestetik yaklaşımımızı sunuyoruz. Premedikasyon 0.5 mg/kg nasal midazolam ile yapıldı. Yüz maskesi cilde temas ettirilmeden anestezi induksiyonu sevofluran ile yapıldı. İntravenöz 2 mg/kg ketamine and 50 µg fentanyl veridikten sonra, intravenöz 1 mg/kg/st ketamine and 0.1 µg/kg/dk remifentanil ile anestezi idamesi yapıldı. Postoperatif ağrı kontrolü için 2 mg intravenöz morfin uygulandı. Remifentanil ile ketamin EB'nin genel anestezi ve analjezi uygulaması için pratik bir yol ve hava yolu yönetiminde etkili, basit ve güvenli olarak gözükmektedir.

**Anahtar kelimeler:** Epidermolizis bülloza, ketamin, remifentanil, anestezi

fragile skin and mucous membranes all of the anesthetic procedures during these operations should be handled gently in these children. For all these reasons, we used a combination of ketamine-remifentanil in order to minimize intervention to airway control.

#### CASE REPORT

We present the anesthetic management for twin patients with epidermolysis bullosa carried out during the syndactyly operation. 15-year-old, 24 kg and 25 kg, twin sisters with epidermolysis bullosa underwent operation for syndactyly. Premedication was done with 0.5 mg/kg nasal midazolam. Routine monitorization was done during the operation. The electrocardiography electrodes were placed with using electrode gel over wet sponches and so

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they did not directly touch the patients' skin. Pulse oximetry was placed on both patients' anterior part of lower leg after oxytetracycline and polymyxine B eye pomade (Terramycin®, Pfizer, Istanbul, Turkey) was applied and that part was covered with wet sponch. Pulse strength was evaluated by palpating the carotid arteries of the patients instead of using routine non-invasive blood pressure monitorization cuffs. After pomade was applied and covered with wet sponch to the facial parts which could be in contact with facial masks, anesthesia induction was performed with 8% sevoflurane in 50% O<sub>2</sub> - 50% N<sub>2</sub>O, gently without touching patients' faces.

After the anesthesia induction, we covered patients' faces with wet sponches and gently put the facial masks on their faces. Intravenous cannulation was done through the anterior part of lower leg and was fixed with only sponches, avoiding plaster usage. We made a small incision in the middle of the sponch to put over the intravenous line from that hole and after covering the iv line with sponches, we fixed it with plasters above the sponches. For the maintenance of anesthesia 2 mg/kg ketamine bolus and later infusion of 1 mg/kg/hr was given intravenously. Also 50 µg fentanyl and 0.1 µg/kg/min remifentanyl infusion was given to both patients. Endotracheal intubation was not done. Because the patients' spontaneous breathing was preserved enough, there was no need for ventilatory support, only O<sub>2</sub> was appropriate. Both operations were ended approximately in 4 hours. At the end of the operations, we stopped ketamine and remifentanyl infusions and injected 2 mg intravenous morfin for postoperative pain control.

## DISCUSSION

Since epidermolysis bullosa is an incurable disease, the current management is mainly focused on supportive care and prevention of complications.<sup>3,4</sup> Under the circumstances, adequate sedation and analgesia while maintaining spontaneous ventilation as in our cases is superior to laryngeal mask airway placement or endotracheal intubation. Intubation or LMA placement would increase the risk of damage to mucosal membranes and laryngeal bullae formation and direct pressure and friction caused by a facemask can blister or peel off the extensively fragile skin.<sup>2,5</sup> The anesthesiologists also try to minimize the trauma to the skin in order to prevent chronic blood loss in these patients who are probably anemic because of this reason.<sup>1</sup>

If intubation is needed in patients with RDEB, it should not be forgotten that the involvement of the

skin and the mucous membranes in the regions of the face, the neck, and the oropharynx can produce considerable difficulties in their intubation because of limited mouth opening due to scarring and contractures at the corners of the mouth.<sup>6</sup>

Since ketamine allows stable hemodynamics and causes less respiratory depression, it can be the drug of preference in children with epidermolysis bullosa. In a case report of two children with epidermolysis bullosa published by Wu,<sup>2</sup> it was shown that during dressing changes and whirlpool bath propofol and ketamine infusions provided satisfactory sedation with significantly reduced narcotic requirements compared with propofol alone. And they did not observe any side effects like delirium, agitation or hallucinations which are the most common side effects of ketamine. Also we did not observe these side effects during intraoperative and postoperative management of our patients.

We have preferred to use the combination of ketamine with remifentanyl and fentanyl in our patients similarly to the combination of ketamine with propofol in the study of Wu.<sup>2</sup> Ketamine may be a good choice for epidermolysis bullosa.

Brachial plexus anesthesia, epidural anesthesia, and spinal anesthesia allow the maintenance of airway patency with minimal epidermal-dermal damage and postoperative pain relief in some study with RDEB.<sup>1</sup> Baloch et al<sup>7</sup> used to spinal anesthesia and epidural analgesia for labour in a pregnant patient.

We preferred oral route for preoperative sedation of our patients. Our patients were 17 years old but also in small children intramuscular or rectal medication should be thought twice because of blister formation or perianal trauma.

Combining ketamine with remifentanyl seems to be effective, simple, safe for airway management and practical way to provide general anesthesia and analgesia of EB patients without respiratory depression.

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