



## Emergency Difficult Airway Management in a Patient with Severe Epidermolysis Bullosa

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Epidermolysis bullosa (EB) is a rare disease characterised by vesiculobullous lesions with minimal trauma to the skin and mucous membranes. Bleeding, scar tissue, contractures, oedema and lesions that can spread throughout the body can cause a difficult airway and vascular access in patients with EB. Therefore, anaesthetic management in patients with EB is a major problem even for experienced anaesthesiologists. Herein, we report a case of difficult airway management in a patient diagnosed with severe EB who presented for emergency tracheostomy because of respiratory failure under general anaesthesia.

**Keywords:** Epidermolysis bullosa, nasal fiberoptic intubation, difficult airway

### Introduction

Epidermolysis bullosa (EB) is a rare disease characterised by vesiculobullous lesions in the skin and mucous membranes that grow because of minimal trauma. EB occurs during birth or from the first year of life, and its incidence rate is 1/50000–1/500000 (1). Epithelial or subepithelial tissue defects are regarded to be responsible for its pathophysiology (2). Scar formation and tissue contraction occur in the oral, pharyngeal and oesophageal mucosa and cause limited mouth opening and neck movement due to the development of contractures (1). Therefore, the anaesthetic management of patients with EB is a major challenge even for experienced anaesthesiologists. An emergency notification of a difficult airway was not encountered in such a severe case in the review of literature. Difficult airway management in a patient diagnosed with severe EB and for whom emergency tracheostomy was planned under general anaesthesia due to respiratory failure is presented in this case report.

### Case Presentation

It was found out that the 20-year-old, 22-kg male who presented to our hospital due to nausea, vomiting and weakness 10 days ago was staying in the adult intensive care unit due to poor general conditions and that he had an anamnesis of EB that was diagnosed at birth for which he was intermittently hospitalised. No characteristics were found in his family history, and he had no history of previous surgery. During his physical examination, the following were observed: he had a poor general condition, he was unconscious, there were open sores and peelings in 40% of his body and he had respiratory distress. One-finger mouth opening and neck mobility of less than 15° were detected in the evaluation of his airway. His temporomandibular joint movements were restricted. Therefore, Mallampati scores could not be evaluated. The thyromental distance was 5 cm and the sternomental distance was 10 cm. The patient's eyes had an exophthalmic appearance because of extreme oedema. The region of the lower jaw and neck was extremely oedematous and strained such that it could not be gripped with the hand (Figure 1, b). The inside of the mouth was bleeding and had a fragile structure. The laboratory test results were as follows: aPTT, 85.8 s; PTZ, 6%; PTT, 123 s; INR, 10.4; platelet count, 38,000/mm<sup>3</sup>; haemoglobin, 4.7 g dL<sup>-1</sup> and HCT, 16.7%. It was found that a segmental contraction with smooth contours was detected at the C5 level on oesophagography performed a month ago. After obtaining informed consent from the patient's relatives, the patient who was scheduled to undergo urgent tracheostomy for the purpose of airway control due to respiratory failure was taken to the operating room after applying 3 units of fresh frozen plasma. Electrocardiogram (ECG), invasive arterial blood pressure (IABP) and peripheral oxygen saturation (SpO<sub>2</sub>) monitoring were performed. After being reduced in size, electrography electrodes were placed with an additional gel on them. HR was

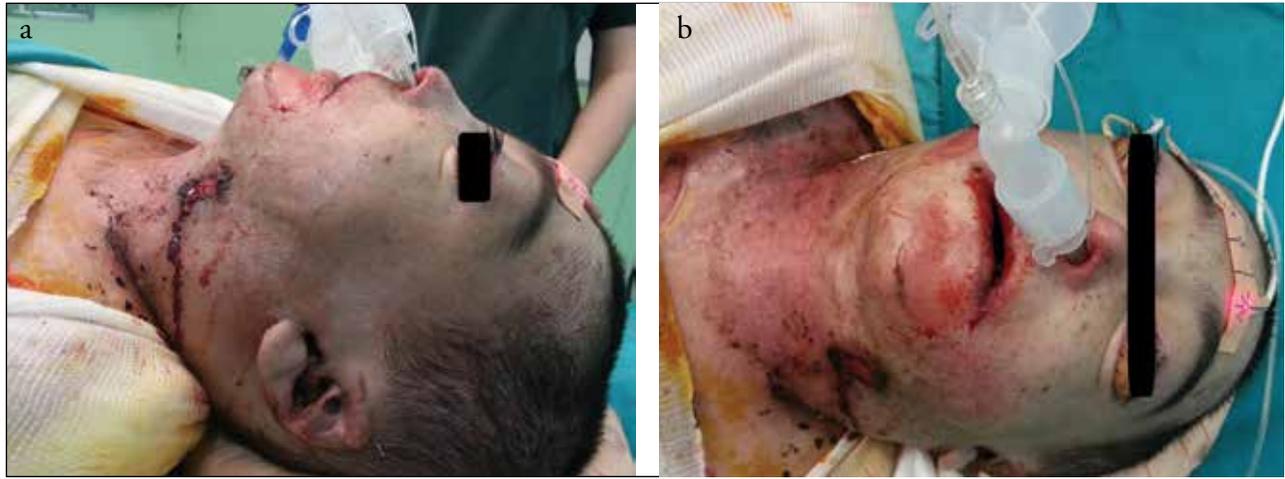


Figure 1. a, b. Short thyromental and sternomental distance, one finger mouth opening and restricted neck movements

120 beats  $\text{min}^{-1}$ , IABP was 90/50 mmHg and  $\text{SpO}_2$  was 80% (10 L  $\text{min}^{-1}$  under oxygen mask support). The central venous catheter previously placed in the right jugular vein was used for the infusion of fluid, and the cannula previously placed in the left radial artery was used for invasive blood pressure measurement. Considering the difficult intubation and ventilation of the patient, intubation was performed under sedation by an experienced anaesthetist through a flexible fiberoptic bronchoscope. Alternative airway tools [video laryngoscopes, laryngeal mask airway (LMA), LMA-Fastrach and i-gel LMA] were kept available. The patient was preoxygenated for 5 min with 100% of oxygen. Sedation was provided with ketamine 0.5 mg  $\text{kg}^{-1}$  and 0.5  $\mu\text{g kg}^{-1}$  fentanyl. An entrance was made through the nasal passage with the fiberoptic bronchoscope on which a number 5.5 spiral intubation tube lubricated with a water-based lidocaine gel was placed and properly inserted. Meanwhile, the pharyngeal tissue and vocal cords were found to be oedematous. After passing the vocal cords with the bronchoscope, the patient was intubated at the first attempt by sliding the tube on the bronchoscope; immediately after that, 2 mg  $\text{kg}^{-1}$  propofol was intravenously administered. It was confirmed by means of the bronchoscope that the tip of the tube was on the carina. Support was provided with an oxygen mask (5 L  $\text{min}^{-1}$ ) during the process, and  $\text{SpO}_2$  values did not fall below 90%. Anaesthesia maintenance was provided with 1–2% of sevoflurane in 50% of oxygen and 50% of air mixture. A neuromuscular blocking agent was not administered to the patient. No complication was observed during the operation that lasted for 45 min. Because the mean arterial pressure fell 50 mmHg, perioperative dopamine infusion was started at a dose was infused at a dose of 10  $\mu\text{kg}^{-1} \text{min}^{-1}$  during the process. The  $\text{SpO}_2$  level remained between 80 and 90%. For the purpose of postoperative analgesia, 20 mg tramadol and 1 g paracetamol were intravenously administered. At the end of the operation, the patient with approximately 50 cc of bleeding was transferred tracheotomised to the intensive care unit to receive prolonged mechanical ventilation. The patient died of multiorgan failure after one day.

## Discussion

Epidermolysis bullosa is a disease that causes blisters and erosions in the skin and mucous membranes (3). It has three types, simple, dystrophic and junctional, according to the level of EB tissue disorders. While the autosomal dominant ones remain benign, the ones that are autosomal recessive and have poor prognosis cause minor trauma, widespread ulcers and vesicobullous lesions. In addition to medical problems that may accompany EB, characteristic factors in anaesthesia applications are challenges between airway management and skin integrity protection associated with the oropharyngeal mucosa and skin involvement as well as challenges in airway and monitoring because the minimal trauma causes the formation of new blisters.

In patients with EB, lesions that can spread to the whole body, bleeding, scar tissue, contractures and oedema can cause a difficult airway and a difficult vascular access. Airway difficulty may be seen due to temporomandibular joint involvement and along with accompanying problems. Due to the 1 cm mouth opening and limited movements of the temporomandibular joint, it was suggested that difficult intubation occurred in a 12-year-old patient who was diagnosed with EB one year after birth; therefore, a LMA was applied, and it was reported that airway security was successfully provided (4). Even minimal traumas occurring during LMA applications may contribute to the formation of blisters; therefore, their use is controversial. To minimise the formation of blisters due to trauma, nasotracheal intubation is performed in patients with EB in whom a difficult airway is expected due to the limited mouth opening (5). Griffin et al. (6) reported that a total of 469 surgical procedures, 390 of which were under general anaesthesia, were performed in 44 patients with EB within 10 years; intubation difficulty was seen in 10 patients, and various methods, such as blind nasal and fiberoptic intubation, were performed in those patients. In another study, oral intubation was performed in 113 of 131 patients and nasal intubation in 18 (7).

Heuvel et al. (8) reported that difficult intubation was seen in 25 of 79 patients with EB in whom general anaesthesia was administered and that an unexpected difficult intubation was encountered in 2 patients. In a patient with EB, who had a mouth opening of 1.5 cm and who was scheduled for a mandibulectomy, a secure airway was established by inserting an intubation tube via a tracheotomy under sedation (9). It was reported that fiberoptic nasal intubation was successfully implemented and that no complications developed in another patient who had a 2.5-cm mouth opening and was expected to have a difficult airway (9). Due to the limited mouth opening and sensitive and bleeding oral mucosa in our patient, fiberoptic nasal intubation was planned and successfully implemented. Deep sedation was avoided because it could make airway management more difficult as the patient would be unconscious. A water-based lidocaine gel (Cathejell, Taymed, Istanbul, Turkey) should be applied on the tube to minimise the trauma in regions through which the intubation tube passes and to facilitate the passage. To minimise trauma during tube fixation, the tube should not be fixed to the skin but instead it should be fixed to the shield of the operation table together with the anaesthesia circuit or to the apparatus on the anaesthesia device. In cases when general anaesthesia may be necessary, it should be noted that difficult mask ventilation may occur; an emergency tracheostomy should (might) be performed. Rocuronium should be used as the neuromuscular blocker agent, and sugammadex should be kept available as an antagonist. In addition to intubation difficulty, it should be noted that ventilation difficulties may also develop in patients with severe EB. Sedation can be applied in manner where spontaneous respiration will be protected in patients in whom intubation is not performed.

While intubation can be smoothly performed, extubation difficulties may occur due to oropharyngeal blistering, laryngomalacia and vocal cord thickening, and it (those) may make early postoperative extubation impossible (10). For the purpose of postoperative analgesia, the use of analgesics that could suppress breathing can also make extubation difficult; therefore, the intravenous application of paracetamol might be a good choice. Because postoperative ventilatory support was necessary in our patient, tramadol was safely used as an analgesic, and the patient was transferred to the intensive care unit without being extubated.

In providing vascular access in these patients, invasive attempts are risky because they can lead to the formation of blisters by traumatising the tissue and because they can lead to infection. In these cases, inhaled anaesthetics and anaesthesia induction are often preferred, and it is recommended to provide controls with minimal intervention. Additional invasive procedures were not required in our patient. Fiberoptic nasal intubation was performed to provide intubation with minimal trauma.

## Conclusion

A detailed and thorough preoperative evaluation should be performed in patients with EB, and appropriate preparations should be made in patients who might have airway difficulty. Nasal fiberoptic intubation can be applied by an experienced anaesthetist as a favourable alternative method in patients in whom an urgent airway should be provided.

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