Case report

VOLATILE INDUCTION AND MAINTENANCE OF ANAESTHESIA IN A PATIENT WITH ADVANCED EPIDERMOLYSIS BULLOSA

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Abstract Epidermolysis bullosa dystrophica is characterized by cutaneous bullae formation and subsequent scarring caused by relatively minor trauma to the skin and mucous membranes. These patients grow up with more complicated sequelae that pose multiple challenges to anaesthetists. There is a paucity of data concerning perioperative management in patients with advanced epidermolysis bullosa dystropica, who present with airway problems, particularly when coinciding with seizures from severe electrolyte disturbance, and lack of venous access. Therefore, we report perioperative anaesthetic management by using the volatile induction and maintenance of anaesthesia in a patient with advanced epidermolysis bullosa dystropica. Chiang Mai Medical Journal 2008;47(1):39-44.

Keywords: epidermolysis bullosa, volatile induction and maintenance of anaesthesia

Epidermolysis bullosa dystrophica (EBD) is an autosomal recessive disorder, which is characterized by mechanically induced bullae formation caused by minor friction followed by subsequent scarring.(1) It is an important disease for anaesthetists because unfortunate sequelae may occur during anaesthesia if the necessary precautions are not taken. Patients frequently present with anaemia, hypoproteinaemia, and electrolyte imbalance, which require preoperative correction. Intubation in patients with advanced EBD might be difficult because of mouth contractures, poor dentitions, and intraoral bullae, which can impede laryngoscopy. Trauma from adhesive tape, blood pressure cuffs, or adhesive ECG electrodes may cause new bullae formation. Anaesthetic management of patients with advanced EBD is directed toward avoiding even minor friction to the skin and mucous membranes. However, there is a paucity of data in the literature concerning the perioperative management in patients with advanced EBD, who present with accumulative problems in the upper airway, particularly when coinciding with seizures due to severe electrolyte...
disturbance, and preoperative status can not be corrected because of failure of preoperative venous access. Therefore, we report the volatile induction and maintenance of anaesthesia in a patient with advanced EBD.

Case report
A 20-year-old female patient (20 kg, 120 cm), with a background of EBD since birth, presented with a five-day history of severe diarrhoea and vomiting, and developed generalized tonic clonic seizures due to hyponatraemic dehydration. Because of poor cooperation and failure in multiple attempts to establish venous access for fluid resuscitation, she was scheduled for central venous access under general anaesthesia. On physical examination, her body surface was covered with numerous blisters and fresh and scattered bullae, particularly involving the face, extensor sides of the extremities, and around the mouth and nose. There was evidence of microstomia, poor dentations, intraoral bullae and limitation of temporomandibular joint mobility. There were also flexion contractures and fusion of the fingers of both hands (Fig. 1). Capillary blood sampling demonstrated severe anaemia (haemoglobin 3.2 g/dL), prolonged partial thromboplastin time (2.1 times control value), and severe hyponatraemia (119 mEq/l). She had been treated with corticosteroids in the past, but at the time of admission she was taking phenytoin and applying 0.5% hydrocortisone ointment to her lesions.

She was taken to the operating room after an adequate fasting period without premedication. The respiratory circuit was primed for 3 minutes using 8% sevoflurane in 100% oxygen with 6-l/min fresh gas flow. Her face and chin, gloves and the anaesthetic face mask were well lubricated with hydrocortisone ointment. Spare laryngoscope blades, airways and intubating forceps were also well lubricated. A pulse oximeter probe was positioned on the left earlobe. Anaesthesia was induced with vital capacity induction using the face mask with sevoflurane starting at 8% in 100% oxygen, with fresh gas flow decreasing from 6-l/min to a 3-l/min during maintenance (50% oxygen), with up to 3.5% sevoflurane. Spontaneous breathing via the face mask was maintained throughout the whole procedure.

After induction of anaesthesia, needle electrodes were applied subcutaneously to the extremities for electrocardiographic monitoring, and non-invasive blood pressure was measured with paraffin gauze covering under the cuff of the sphygmomanometer. Her eyes were carefully closed with a gel pad. A central venous catheter was then inserted into the right internal jugular vein and sutured to the skin. During the procedure, the Trendelenburg position and passive leg raising were used to facilitate central venous access and treat hypotension associated with hypovolaemia. The total anaesthesia time was 30 minutes. There was no new bullae formation and perioperative complication. After her level of consciousness gained to preoperative level, she was transferred back to the intensive care unit. Her initial postoperative course was notable for the requirement of extensive fluid resuscitation, blood component replacement, and electrolyte balance. Her recovery was uneventful and she was discharged on the sixth postoperative day.

Discussion
EBD is a group of inherited disorders characterized by mechanically induced blistering
Figure 1. Photograph shows a 20-year-old female patient (20 kg, 120 cm), with advanced epidermolysis bullosa dystrophica.
and bullae formation occurring immediately below the lamina densa of the basement membrane. These blisters heal with atrophic scar formation. This structural abnormality affects the stratified squamous epithelium of the skin, oropharynx and upper oesophagus, while the ciliated columnar epithelium of the larynx and trachea is spared. Certain precautions and meticulous attention to several details are required throughout the operation to avoid anaesthetic related morbidity. Although direct pressure to the skin is not as damaging as frictional or shearing forces, all persons involved in handling the patients before, during and after an operation must be made aware of the extreme vulnerability of their skin.

Routine monitoring is not always possible, so these patients are often poorly monitored. Pulse oximetry is the preferred method of monitoring pulse rate and there have been no complications reported with its use. Electrocardiogram monitoring is not easy because the adhesive tapes can cause friction at application or removal. In our patient, needle electrodes were placed subcutaneously, and non-invasive blood pressure was measured with paraffin gauze covering without new bullae formation.

Chronic anaemia resulting from chronic blood loss associated with frequent wound dressing is an additional problem for perioperative management. Red cell preparation and replacement should be considered in a major operation. However, in accordance with a previous review, minimal blood loss associated with this procedure was not a major problem.

Airway patency may be a problem in some patients with advanced EBD because of microstomia, poor dentations, severe strictures, webbing, or large bullae in the oropharynx, which can make intubation impossible. However, several cases, without intraoral bullae, have been reported, in which endotracheal intubation was performed successfully with no adverse sequelae after extubation. The use of a laryngeal mask airway (LMA) during the procedure has also been described. Nevertheless, a review of 44 patients over a 10 year period has warned against the risks. If tracheal intubation is necessary, it is important to ensure that both laryngoscopy and intubation are non-traumatic. A well-lubricated laryngoscope and endotracheal tube are preferred to avoid trauma. A retrospective study reported that a minority of these patients required fiberoptic endotracheal intubation secondary to limited opening of the mouth and contractures in the neck. However, these intubations were achieved with an intravenous anaesthesia. In our patient, failure of preoperative venous access and poor cooperation were reasons to anaesthetize the patient with the VIMA technique. Fresh intraoral bullae proved on important problem for airway manipulation. However, because the procedure had a non-airway involvement and a relatively short duration, less oropharyngeal manipulation was achieved using gentle application of a well-lubricated face mask, and spontaneous breathing was kept throughout the procedure, without airway complication. By using sevoflurane, postoperative involuntary movements and shivering were minimal, with less risk of inadvertent friction and trauma, and this was preferable in patients with difficult or no venous access.

Because of coagulation abnormality, a right internal jugular approach for central venous access was planned instead of a subclavian one, in which bleeding or haematoma is less controllable. The perioperative challenge in this
patient was the haemodynamic compromise from anaesthesia together with preoperative hypovolaemia. However, the position required in this procedure was trendelenburg, which suited the patient’s status. In addition, further volume requirement was treated with gentle passive leg raising. The haemodynamic situation was within the preoperative range, and awakening to the level of preoperative consciousness was fast and uncomplicated.

In conclusion, as a result of the findings in this and other case reports, anaesthetic management in patients with advanced EBD should include careful airway manipulation, reduction of mucocutaneous contacts, prevention of pressure or friction skin trauma, appropriate haemodynamic balance, and the use of non-adhesive material. We successfully managed the anaesthesia in a patient with advanced EBD.

References
การนำสลบและรักษาระดับความลึกของการสลบด้วยยาดมสลบในผู้ป่วยโรค epidermolysis bullosa ชนิดรุนแรง

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บทคัดย่อ  Epidermolysis bullosa dystrophica เป็นโรคทางพันธุกรรมที่มีลักษณะเด่น คือ มีผิวหนังและเยื่อบุต่าง ๆ เปราะบาง เนื่องจากการกระแทกหรือการสัมผัสเบา ๆ ก็อาจก่อให้เกิดการสลบ ทำให้ผิวหนังและเยื่อบุแตก จนกลายเป็นแผลเป็น พื้นที่แผลเป็นอาจขยายขึ้น มักจะมีร่องริมของแผลเป็นติดตาม การทำให้แผลด่างว่างระหว่าง ๆ หรือเป็นแผลเป็นตามมาโดยผู้ป่วยที่มีรูปแบบหน้าผาก หรือศีรษะและเป็นอุปสรรคสำคัญในการเปิดทางเดินหายใจได้ นอกจากนี้ ผู้ป่วยยังมีค่าอิเล็กโทรไลท์ที่ไม่ปกติ ซึ่งยากที่จะแก้ไขได้ ดังนั้นเทคนิคทางวิสัญญีวิทยาในการรักษาผู้ป่วยดังกล่าวจึงมีความสำคัญมาก การรักษาผู้ป่วยดังกล่าวต้องมีการเตรียมการให้ล่วงหน้า เช่น การเตรียมยาดมในรูปแบบที่มีประสิทธิภาพในการทำให้ผู้ป่วยผ่านการลูกเรียนโดยไม่ต้องใช้การเจาะหูหรือการเจาะหน้าผาก การใช้ยาดมในรูปแบบ volatile induction and maintenance of anaesthesia ในการรักษาผู้ป่วย epidermolysis bullosa dystrophica เชียงใหม่เวชสาร 2551; 47(1): 39-44.

คำสำคัญ: epidermolysis bullosa, volatile induction and maintenance of anaesthesia