

## Epidermolysis bullosa: facts for the anesthesiologists

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### Keypoints

Epidermolysis Bullosa (EB) is a group of congenital, hereditary disorders of the skin and mucous membranes, characterized by development of blisters on minor trauma or friction. Special attention is needed in patients with epidermolysis bullosa so as to prevent exacerbating existing lesions and avoiding formation of new lesions. Airway should be managed with utmost care and regional technique should be used whenever feasible.

### Abstract

Epidermolysis Bullosa (EB) is a group of congenital, hereditary disorders of the skin and mucous membranes, characterized by development of blisters on minor trauma or friction. We report a case of 3 year male child, a known case of epidermolysis bullosa, posted for dental extraction. Patient had difficulty in swallowing. Examination revealed multiple raw areas over the body and intraoral lesions. Blood investigations revealed low hemoglobin, rest all investigations were within normal limits. Patient was sedated with oral ketamine. Appropriate monitors were attached taking care to avoid any adhesive dressing, tapes or direct pressure on the skin. Induction of anaesthesia was done using Sevoflurane in Oxygen, with Succinylcholine as muscle relaxant. Anaesthesia was maintained with Sevoflurane in Oxygen and Nitrous Oxide with Atracurium. Nasal intubation was done. Intraoperative period was uneventful, patient was extubated on table. New blister formation occurred at the site where NIBP cuff was applied.

**Keywords:** Epidermolysis Bullosa, Anesthesia management

### Introduction

Epidermolysis Bullosa (EB) is a group of congenital, hereditary, blistering disorders of the skin and mucous membranes. The disease usually affects the stratified squamous keratinizing epithelium of the skin, but blistering can occur on any mucosal surface. Patients with EB have associated features that range from malnutrition, anemia, to laryngotracheal and esophageal scarring, and dilated cardiomyopathy in rare instances. Aim is to prevent exacerbation of existing lesions and prevent formation of new lesions.

### Case report

A 3 year old male child weighing 8 kg, born of non-consanguineous marriage was posted for extraction of central incisors in view of dental caries. The child was a known case of EB, with blistering since second day of life. Patient had difficulty in swallowing and was on liquid diet. On examination he had multiple raw areas all over the body. Airway examination revealed intraoral lesions with adequate mouth opening. Pulse rate was 100/min, respiratory rate 16/ min, systemic examination was unremarkable. Patient was on Syrup. Amoxicillin, lignocaine jelly, and calcium supplements.

Pre-operative investigations showed anemia with hemoglobin of 8.6 gm%, rest all investigations were within normal limits.

Patient was pre-medicated with Ketamine 6mg/kg and Glycopyrolate 4mcg/kg orally. Induction was done using Sevoflurane in Oxygen with JR circuit. The face mask was lubricated with water based jelly and Vaseline gauze was kept beneath the face mask. A 24G IV line was inserted and held in place using COBAN® dressing. Inj. Midazolam 0.05mg/kg and Inj. Fentanyl 2µg/kg were given intravenously (iv). Monitoring included electrocardiography, pulse oximetry, non-invasive blood pressure monitoring, capnography and temperature. ECG leads were cut removing the adhesive part and were fixed to the limbs using COBAN® dressing. Blood pressure cuff was lined with a thin layer of soft cotton prior to application. Lubricated temperature probe was placed in the axilla. Eyes were moistened with a methyl-cellulose based lubricant and padded with wet gauze. Patient was intubated after administration of Inj. Succinylcholine 2mg/kg, using 4.5mm ID north polar endotracheal tube and secured using roller gauze. Throat packing was done with wet roller gauze. Anesthesia was maintained using Sevoflurane in Oxygen and Nitrous Oxide 50:50. Inj. Atracurium 0.5mg/kg iv was administered for muscle relaxation. Analgesia was supplemented with Inj. Paracetamol 10mg/kg. Total of 110ml of crystalloid was given to the patient, blood loss was minimal.

At the end of procedure, patient was reversed using Inj. Glycopyrolate 8µg/kg iv and Inj. Neostigmine 0.05mg/kg iv and extubated on table. There was new blister formation at the site where blood pressure cuff was placed. Subsequently patient was shifted to PACU for observation.

### Discussion

Epidermolysis Bullosa was first described by Hebra<sup>1</sup> in the year 1871. Inheritance can be either autosomal dominant or recessive. It is characterized by development

of blisters on minor trauma or friction which can cause debilitating and also life threatening scarring. Incidence is around 1 in 100000. It is classified into three major subtypes depending on the location at which blistering occurs:

- Epidermolysis Simplex
- Junctional EB
- Epidermolysis bullosa dystrophica
- Kindler Syndrome is a newly added fourth group.
- More than 30 subtypes have been identified with help of advances in technology.<sup>2</sup>

Pre operative evaluation should focus on child's present condition with relation to the blistering, nutritional status, infection and the use steroids. Patients with EB can have a difficult airway due to lesions and scarring in the perioral, intraoral and laryngotracheal region.<sup>3</sup> There may be associated anemia and dilated cardiomyopathy.

These patients have mucosal and cutaneous fragility, any type of instrumentation can cause blister formation. Spontaneous blister formation in the larynx has been reported in patients with junctional and dystrophic EB.<sup>4</sup> Care of the skin is an important aspect in perioperative management. Compressive forces are tolerated better compared to shear forces.<sup>4</sup> Bed sheets on which patients are placed should be crease free, sheep skin can also be used. Generous padding of the pressure points should be done with soft cotton. Eye protection is important as EB patients are prone to ocular manifestations like scarring of eyelids, methyl-cellulose based lubricant can be applied to the eyes. Avoid taping of eyes, instead a silicon-based pad placed over the eyes or covering the lubricated eyes with moist gauze is recommended.

Monitoring is as per ASA guidelines, but application of the monitors is to be modified. Pulse oximetry probe should preferably be of clip-on type. ECG electrodes should be fixed without their adhesive part with non-adhesive dressing. The area where blood pressure cuff is to be applied must first be covered with soft cotton over which cuff can be wrapped. Temperature probe to be

lubricated and kept in the axilla, instead of fixing it to the skin. Intravenous access is difficult to establish and should be secured with sutures, silicon based dressings or soft cotton wraps, central venous line can be placed if peripheral access is not possible. Invasive monitoring can be used in cases of prolonged surgery.

All currently available inhalational or IV induction agents can be used in patients with EB. Premedication can be helpful as restraining a child can lead to blister formation. Intramuscular Ketamine has also been used for induction and maintenance of anaesthesia.<sup>5</sup> Both non-depolarizing muscle relaxants and succinylcholine can be used in patients with EB. Airway instruments need to be lubricated with water based jelly, avoid lignocaine jelly. Oropharyngeal airways should be avoided. Face masks should also be lubricated and should be placed on the face gently without pressure. A straight blade laryngoscope is preferred, endotracheal tube should be smaller than expected, softened and well lubricated. Nasal intubation is preferred as the nasal mucosa is not as prone to bullae formation, compared to oral mucosa.<sup>3</sup> The nasal tube can be fixed to the head using a silicon based dressing. Vigorous suctioning of the oral cavity is not recommended.

Regional anesthesia is a good alternative, whenever possible, to avoid airway manipulation. Care is to be taken during cleaning the puncture site. Preparatory solution is to be poured over the area. Avoid rubbing the area and administering local infiltration, both of which can lead to bulla formation.<sup>6</sup>

Struggling during waking up from anesthesia can lead to blister formation. Pain relief should be adequate so as to prevent excessive movement. Oxygen masks can be avoided as the sharp edges of the mask can cause skin injury.

### Conclusion

Special attention is needed in patients with epidermolysis bullosa so as to prevent exacerbating existing lesions and avoiding formation of new lesions. Airway should

be managed with utmost care and regional technique should be used whenever feasible. With adequate knowledge of the disease and thorough preoperative planning a favorable patient outcome is possible.

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