

Epidermolysis Bullosa – Tread Carefully!!

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Patients with epidermolysis bullosa (EB) provide a unique and complex challenge for the anaesthesiologist. This is a rare inherited group of diseases characterized by excessive fragility of skin and mucous membranes. These patients tolerate compressive forces to the skin but barely tolerate friction and shearing forces. Major challenges to the anaesthetist include establishing adequate monitoring without damaging the skin, issues with positioning, high possibility of difficult airway, compromised immunity and nutritional deficiencies, and other comorbidities. We present a case of a 4-month-old baby boy with epidermolysis bullosa simplex presented with blisters and ulcers all over the body, posted for thoracoscopic plication for diaphragmatic eventration. We highlight that with meticulous preoperative planning and extreme attention to prevent any shearing or frictional forces, anaesthesia can be delivered safely.

Keywords: Epidermolysis bullosa, child, anaesthesia

Introduction

Epidermolysis bullosa (EB) is a group of inherited diseases characterized by severe skin fragility of the skin and mucous membranes. Three major subtypes include Epidermolysis bullosa simplex, Junctional epidermolysis bullosa, and Dystrophic epidermolysis bullosa. There is defective protein formation leading to separation of the epidermis from the basement membrane and transudation of fluids into disrupted skin (bullae formation) and subsequent scar formation from healing.¹The skin is thus extremely fragile and blisters and tears occur with minor friction or trauma. There are unique challenges in the anaesthetic management of patients with EB regarding maintaining the airway and the use of monitoring equipment without damaging epithelial surfaces. This case report describes the successful anaesthetic

management of a patient with EB presenting for thoracoscopic plication of the diaphragm.

Case report

A 3 kg, 4-month-old male child with congenital EB, was admitted with fever and cough for 5 days and rapid breathing for one day. He was diagnosed with EB on 2nd day of birth. On examination, the child was malnourished, sick, and lethargic. The body was covered with eruptions and bullae (Figure 1a) We noticed large ulcers over the occipital area, ear lobes, and cheeks. (Figure 1b) Toenails and fingernails were torn and disrupted. (Figure 1c) The contrast-enhanced CT chest suggested right hemidiaphragm eventration and right middle lobe consolidation. Blood investigations revealed anaemia and hypoproteinaemia. Thoracoscopic plication for diaphragmatic eventration was planned after a week of optimization with antibiotics and intravenous (IV) fluids and packed red blood cell transfusion. Preoperative vitals were as follows. Pulse rate - 164/min, respiratory rate - 42/min, oxygen saturation – 96% (on 2 l/min oxygen by nasal prongs) with bilateral conducted sounds on chest auscultation. Airway examination was normal. Airway anomaly and cardiac abnormalities were excluded by fiberoptic bronchoscopy and 2D echocardiography.

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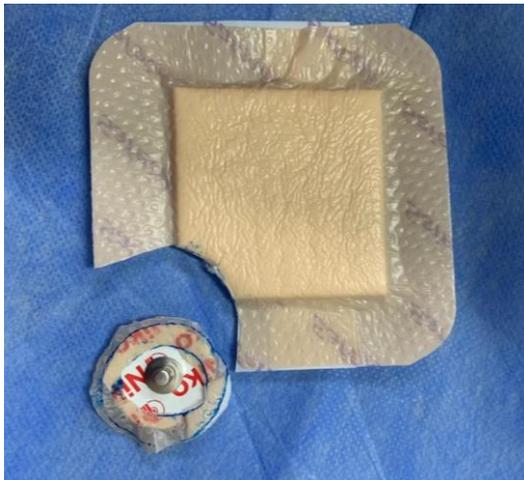


Figure 1: a) The body was covered with eruptions and bullae. b) large ulcers were noticed over the occipital area, ear lobes, and cheeks. c) Toenails and fingernails were torn and disrupted.



On the day of surgery, the trolley and operation theatre table were padded adequately. Sterile gloves were used to maintain asepsis. All pressure points were padded with silicon-based dressings (Mepilex) and paraffin gauzes were placed over the ulcers. A 24 G IV cannula was secured and fixed with silicon dressing. For ECG monitoring, the adhesive part of the ECG electrode was trimmed and used with a cut out from silicon dressing (Figure 2)

Figure 2: Makeshift of ECG



A well-lubricated (water-soluble gel) clip-on type of pulse-oximeter probe was used for saturation monitoring. For blood pressure monitoring, a thick layer of cotton padding was used underneath the BP cuff and the interval was set to 30 minutes. The temperature was monitored with

a contactless infrared thermometer. Intravenous fluid warmers and warm blankets were used to maintain normothermia.

A paraffin gauze was kept under the mask for preoxygenation and atraumatic mask ventilation. (Figure 3)

Figure 3: Placement of paraffin gauze



General anaesthesia was induced with IV fentanyl (2µgm/kg) and propofol (2 mg/kg) and maintained with oxygen and air and sevoflurane 1.5- 2%. Minimal pressure was used for head tilt and lift chin lift. Endotracheal intubation was facilitated with IV rocuronium using a well-lubricated size 2 laryngoscope blade, and tracheal tube was fixed with a tie and paraffin gauge underneath to prevent any shearing on skin. Ocular lubrication was done with Neosporin ointment and silicon-based eye padding was used. Child was carefully positioned for surgery. It

lasted for 2.5 hours with a total blood loss of about 50 ml. The patient's trachea could not be extubated due to poor preoperative chest condition and he was shifted to the Paediatric intensive care unit for continued mechanical ventilation. Postoperative pain was managed with IV paracetamol. He was extubated after 24 hours and was discharged from the hospital after a week.

Discussion

Epidermolysis bullosa possess various challenges for an anaesthesiologist. Apart from the typical cutaneous manifestation in EB, these patients have extracutaneous manifestations of concern to the anaesthesiologist such as progressive webbing (mitten deformities) contracture of the hands and feet, osteoporosis, muscle wasting, muscular dystrophies, dilated cardiomyopathy, chronic anaemia, intraoral blisters, tooth abnormalities, microstomia, ankyloglossia and nutritional compromise with low serum albumin levels.² Our patient had only cutaneous manifestations, muscle wasting, and nutritional deficiencies.

Preoperative evaluation should specifically focus on assessing the severity and degree of skin lesions, presence of contractures, which could lead to reduced mouth opening, reduced neck mobility, difficult positioning, and intravenous access. Since these patients can have airway abnormalities, laryngeal stenosis, and blisters intraorally, it is important to prepare a difficult airway cart and adequately lubricate the instruments like laryngoscope blade. A preoperative fiberoptic bronchoscopy under sedation was performed in our case same is advised in literature to rule out any laryngeal stenosis.³ Our patient had anaemia and hypoproteinaemia as these patients have reduced oral intake, high-energy requirement, and malabsorption (gastrointestinal involvement) requiring preoperative optimization. Cardiac and genito-urinary involvement should be evaluated. Optimization of systemic infection and electrolyte imbalance is also needed.⁴

Our intraoperative goals included extremely gentle handling of the patient to maintain skin

integrity.⁵ We premedicated our patient to avoid any friction from a combative agitated child. Padding of transportation trolley and the operating table is required and there should be no folds and creases on the sheets. We recommend covering ulcerative lesions with a paraffin gauge piece or lubricating them with hydrocortisone cream and pad all pressure points as we did in our case. Use of sterile gloves, while handling the patient is advised to maintain strict asepsis but rubbing alcohol swabs, should be avoided, antiseptic spray can be used instead.⁶ Minimal standard monitoring is advised, and indigenous modifications can be done as we did for the ECG electrode. Clip on-type pulse oximetry probe and frequent change of its position with adequate lubrication are preferred.⁷ We increased our blood pressure monitoring interval to 30 minutes and used cotton padding underneath the cuff. Singh S et al also suggested avoiding continuous BP monitoring in minor surgery with limited fluids loss.⁸ Intravenous access can be challenging in such patients. Use of a tourniquet is not recommended.⁹ All fluids used must be temperature regulated, we used iv fluid warmer and warming blankets as well to maintain normothermia.

Mask ventilation must be gentle avoiding the use of a sharp-edged mask. Succinylcholine is undesirable as patients are at risk of myopathies and the fasciculations can lead to shearing of skin. Careful endotracheal tube fixation can be done with a soft tie with paraffin gauge underneath to avoid any adhesives tapes over the skin as we did. A well-lubricated laryngeal mask airway can be used.⁶ Postoperatively pain management is crucial to prevent shearing forces on the skin as a pain-free patient would be less combative. Intravenous analgesia remains the mainstay as regional anaesthesia is usually not possible due to the presence of local site ulcers and difficulty in positioning but there are reports of its safe use.¹⁰ We managed postoperative pain with Intravenous Paracetamol and avoided local infiltration of the incision site and insertion of rectal suppositories for the fear of local site bullae formation.

Patients with EB provide a unique and complex challenge for the anaesthesiologist. With

meticulous preoperative planning and keen attention to prevent any shearing or frictional forces, anaesthesia can be delivered safely.

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