RESEARCH PAPER | Medical Science | Volume : 3 | Issue : 12 | Dec 2013 | ISSN - 2249-555X

Perioperative care of a patient with Congenital Epidermolysis Bullosa

KEYWORDS

Dr Suhas Ramaswamy
Assistant Professor, KMC,Manipal University, Mangalore,Karnataka,India

Dr Mahadeva Kalegowda
Consultant Anaesthesiologist, Vikram group of hospitals, Mysore, Karnataka, India

ABSTRACT *"Few conditions are more devastating to the physical, emotional and social health of an infant or child than Epidermolysis bullosa". The disease is a group of congenital abnormalities of the skin and it encompasses a variety of genetic abnormalities of proteins that mediate adhesion of the skin. We discuss the successful anesthetic management of a child with Epidermolysis Bullosa Dystrophica for upper gastro intestinal scopy.*

INTRODUCTION

Epidermolysis bullosa is an inherited connective tissue disease causing blisters in the skin and mucosal membranes, with an incidence of 1/50,000[1]. It is a result of a defect in anchoring between the epidermis and dermis, resulting in friction and skin fragility. The separation of skin layers occurs after application of friction or shearing forces and results in intradermal fluid accumulation and bullae formation[2]. This may lead to scarring and debilitating, life-threatening medical conditions. In addition to considerations associated with positioning, monitoring, infection, and prevention of skin and mucosal trauma, anesthetic management of Epidermolysis bullosa is uniquely challenging because of the effects on the airway. Careful clinical monitoring and no touch principle is the key for safe anesthetic management[3].

Case report

15 year old boy known case of Congenital Epidermolysis Bullosa Dystrophica presented with history of difficult swallowing since 15days.He was diagnosed to have esophageal soft strictutes.Child had crusty and scaly lesions all over the abdomen,chest,back and thighs. Fingers and toes were fused and dystrophic. Airway and other system examination were normal. Patient's relatives were explained about the procedure, anaesthetic concerns, risks involved and Written Informed consent was obtained. Child was shifted to the endoscopy table along with the child's attender. Intravenous access was obtained in right upper limb using 20G intravenous cannula. Roller bandage was used to secure the intravenous line. Pulse oximeter was connected to left upper limb. We did not record the BP as the cuff inflation can cause skin to peel off[4]. Lignocaine 10% was used to spray the oral cavity. Child was premedicated with Inj Midazolam 1mg, Inj Glycopyrrolate 0.1mg and Inj Fentanyl 50 mcg. Child was induced with incremental doses of Inj Propofol upto 30mg. Oxygen was delivered using gently and loosely applied nasal prongs @ 2lpm. Serial dilation of esophagus was done. Anesthesia was maintained with intermittent boluses of propofol 5mg until the procedure was over. Post procedure recovery was smooth.

DISCUSSION

Epidermolysis bullosa is a group of inherited disorders in which skin blisters develop in response to minor injury. There are four main types of Epidermolysis bullosa: Dystrophic Epidermolysis bullosa Epidermolysis bullosa simplex, Junctional Epidermolysis bullosa, Kindler syndrome (mixed levels of blistering)[5]. Epidermolysis bullosa can vary from minor blistering of the skin to a lethal form involving other organs. Anesthetic challenges that may be faced will be due to oral, pharyngeal, mucosal and skin involvement. Airway management and maintenance of skin integrity are problematic[6]. Avoiding damage to skin and mucosae are basic considerations in caring for these patients. Airway management requires particular care not to damage the skin of the face, the eyes, the oropharynx, larynx, and trachea. To minimize friction, a lubricant (e.g. Vaseline or liquid paraffin) may be used on the face mask as well as on the anesthetist's hands. Intubation must be performed carefully, with lubrication of the laryngoscope blade to minimize friction on the lips, gums, and palate. The endotracheal tube must be well lubricated and softened in warm water. In those patients in whom airway difficulties are anticipated and a bougie is required to facilitate intubation, damage to any part of the airway may occur. An endotracheal tube, uncuffed and half to one size smaller than would normally be selected, is usually required. All tubing should be padded and Vaseline gauze wrapped around any part which may come in contact with the patient. The endotracheal tube may be secured with Vaseline gauze, held by the anaesthesiologist, or remain unsecured during surgery. Laryngeal mask airways (LMA) have been used successfully in patients with EB but, must be placed extremely carefully to avoid damaging the lips and palate. They should also be very well lubricated, with only minimal air inflated into the cuff. Pulse oximetry and capnography are recommended for all cases, but non-invasive blood pressure should only be performed when precautions are taken to avoid damage to skin underlying the cuff (e.g using Vaseline gauze) around the arm before placing the cuff[7]. The placement of ECG electrodes may also cause damage on removal. Surgical staples and crocodile clips may be preferred. General anesthesia is generally preferred. Regional anesthesia has been proposed and, after initial concerns over causing more bullae, more techniques are now being used[8]. Areas where the skin is infected or likely to break down should be avoided. Care should be taken when cleaning the skin: rubbing should be avoided and antiseptic spray used instead. Infiltration of local anesthesia should be avoided. Pain should be anticipated and treated pre-emptively and aggressively.

CONCLUSION

Children with epidermolysis bullosa, some more than others, pose a formidable challenge to the anaesthesiologist. Implementing measures to prevent damage to skin and mucosa, and assuming a "no-touch" approach, ensures that anesthesia for children with EB may be undertaken with few untoward consequences. Anaesthetizing children with Epidermolysis Bullosa pose a challenge of balancing "least touching and least monitoring techniques" on one side and "deeper planes of anesthesia" required on the other side.

Volume : 3 | Issue : 12 | Dec 2013 | ISSN - 2249-555X

REFERENCE

1.Fine JD, Johnson LB, Wright JT. Inherited blistering diseases of the skin. Pediatrician 1991; 18: 175-87. | 2.Carter D, Lin A, Varghese M. Treatment of junctional epidermolysis bullosa with epidermal autografts. J Am Acad Dermatol 1987; 17: 246-50. | 3.Chevaleraud E, Ragot JM, Glicenstein J. (Anesthesia for hand surgery in patients with epidermolysis bullosa). Ann Chir Main Memb Super 1995: 14: 296-303. | 4.Association of Anaesthetists of Great Britain and Ireland. Recommendations for Standards of Monitoring during Anaesthesia and Recovery. London: Association of Anaesthetists of Great Britain and Irelaand, 1988. | 5.Holzman RS, Worthen M, Johnson KL. Anaesthesia for children with junctional epidermolysis bullosa (letalis). Can J Anaesth 1987; 34: 395-9. | 6.Iohom G, Lyons B. Anaesthesia for children with epidermolysis bullosa: a review of 20 year experience. Eur J Anaesthesiol 2001; 18: 745-54. | 7.Hagen R, Langenberg C. Anesthetic management in patients with epidermolysis bullosa dystrophica. Anaesthesia 1988; 43: 482-5. | 8.Doi S, Horimoto Y. Subcutaneous tunneling of an epidural catheter in a child with epidermolysis bullosa. Acta Anaesthesiologica Scandinavia 2005; 50: 394-5.