Anesthetic and alternative airway management of an adolescent with Dystrophic Epidermolysis Bullosa undergoing Esophageal Dilatation

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Introduction: First described by Fox in 1879 Epidermolysis Bullosa Dystrophica (DEB) is an autosomal recessive disorder characterized by separation of the skin at the dermal level and mucosa from underlying tissue spontaneously or by minor trauma. The sequela of this process is formation of bullae, scarring and eventual contractures. This disease process presents many unique challenges for the anesthesiologist, not the least of which is airway management. We discuss a case series of a patient who presented with a special need for unintubated airway during anesthesia for esophageal dilatation (ED). These patients routinely get intubated to protect their airway and secure their patency as the flexible fiberoptic scope is passed through the oral cavity.

Case Series: A 12 year old, 24 KG girl with DEB presented for frequent (every four to eight weeks) ED for esophageal strictures. Preoperative evaluation found an otherwise healthy teen with scarring over her body and several bullae over her ankles, knees, legs, thighs, arms and forearms. Her airway was favorable on prior intubations. The patient and family noted severe sore throat, hemoptysis, and decreased PO intake after each of her prior surgeries in which she was intubated for up to four weeks. In the preoperative area, PO midazolam (0.7mg/kg) was given to minimize the teen’s anxiety. She was taken to the GI suite and positioned herself supine on the endoscopy table, which was padded with gel foam. Monitors were applied in the following fashion: EKG- gel electrodes with sticky pads removed placed under the patient; BP Cuff- upper extremity padded with webril, then size appropriate cuff placed and cycled every 5 minutes; Pulse Oximeter – clear backing/tape not removed, probe placed on finger and secured with webril then Coban nonadhesive silicone dressing. The patient was mask induced after generously coating the face with Vaseline and lightly applying facemask with 8 LPM O2 and 8% sevofluorane. A 22 G angiocath was placed with tourniquet applied over webril and secured with mepiform or mepeitl, webril and then Coban Dressing. A #26 Nasal Trumpet with an endotracheal tube adapter was placed in the nare after generous lubrication with KY jelly. This was then attached to the anesthetic circuit and machine and the patient spontaneously ventilated through the entire procedure under 3 % end expiratory sevofluorane. Medications given IV included propofol, glycopyrolate, dexamethazone and Zofran. At termination of the procedure, the nasal trumpet remained while the patient emerged on 100% oxygen. The patient had a brief, uneventful PACU course and was discharged home. At 1 and 4 weeks follow-up, the patient had no symptoms of hemoptysis or severe sore throat. A similar strategy had been employed for the subsequent three anesthetics after which she was noted to be getting violent after the PO versed. To prevent self inflicted injury an IV was placed pre-induction on the subsequent four anesthetics and IV induction was performed.

Discussion: As noted, the patient with DEB presents several challenges to the anesthesiologist. This particular patient had several prior anesthetics where the trachea was intubated and was found to develop post-procedure hemoptysis presumably secondary to bullae forming and rupturing in the upper airway. For this particular patient and family, this complication severely affected quality of life, especially since she had this procedure every four to eight weeks and had complications for 4 weeks after. Though many of these modifications appear trivial, they have direct impact on outcome. The cycle of trauma, secondary infection, and scarring in DEB leads to many complications. Endotracheal intubation can result in airway bullae (though infrequently) and possibly tracheal scarring leading to strictures even though tracheal epithelium is different. For these reasons, we chose this alternative to intubation. The nasal trumpet allowed the airway to remain patent and ventilation remained adequate. Since the anesthetic circuit is attached via an ETT adapter, we can deliver volatile agent and even assist ventilation if the need arose. Two obvious concerns are potential for aspiration during the procedure and possible need for intubation. Through this series of cases (N= 8) only one possible aspiration pneumonitis was noted. The patient had a nonproductive cough and fever (though normal chest radiograph and WBC count) 2 weeks after an ED. She was treated as an outpatient empirically with antibiotics and soon improved. Our strategy to address need for intubation was to lubricate the laryngoscope and ETT, select an ETT 1 to ½ size smaller for her age, and use a minimal manipulation/touch technique on direct laryngoscopy (DL). Since we knew from recent DL that she had a Grade 1 airway, we were not concerned with a potential difficult airway. It should be noted that this population can have difficult intubations due to old scarring causing limited neck mobility and microstomia. Also, we note specific modifications to monitors, positioning, and IV access which minimize shearing trauma and eventual bullae formation.

Ref:
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