COMBINED ANESTHESIA TECHNIQUE IN A CHILD WITH DYSTROPHIC EPIDERMOLYSIS BULLOSA

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INTRODUCTION
Epidermolysis Bullosa (EB) represents a group of skin diseases, the main characteristic of which is the appearance of bullae after the slightest skin trauma and subsequent scarring. Dystrophic Epidermolysis Bullosa (DEB) is a severe form of EB, which involves a mutation in the gene encoding type VII collagen, which fixes the epidermis to the dermis. This structural abnormality affects the stratified squamous epithelium of the skin, oropharynx, and the esophagus. We present a 3-year-old male child with Dystrophic Epidermolyis Bullosa, who required anesthesia for left hand syndactyly release secondary to chronic scarring.

CASE REPORT
A 3 year-old male, 13.5 kg presented requiring anesthesia for left hand syndactyly release secondary to chronic scarring. His past medical history was significant for the diagnosis of Dystrophic Epidermolyis Bullosa at six months of age. The patient had a previous right hand syndactyly release at an outside institution, which the mother reported “did not go well”. We were unable to obtain the anesthetic records from the previous institution.

Preoperative evaluation one week prior to surgery revealed a cooperative child in no apparent distress with bandages on all extremities. Vital signs were normal for age, however no blood pressure reading was taken. Airway examination revealed a Mallampati class II airway with obvious oral scarring, and bullae present in the oropharynx. The cardiorespiratory evaluation was unremarkable. No preoperative labs were ordered, and the mother was given instruction concerning npo status for the day of surgery. A long discussion with the mother concerning anesthesia techniques and complications was undertaken, and all risks were explained.

On the day of surgery the patient was allowed clear liquids until two hours prior to surgery. The child was extremely uncooperative in the preoperative area, thus it was decided to proceed with IM premedication. The patient was given 30 mg ketamine, 1.3 mg midazolam, and 130 mcg glycopyrolate IM. The child was taken to the operating room sedated in no apparent distress. After oxygenation with 100% oxygen via a lubricated face mask, monitors were placed with mepilex adhesive to avoid skin trauma. The anesthesia was induced with a 8% sevoflurane in 40% oxygen and 60% nitrous oxide via face mask. Venous cannulation was achieved with a 24 gauge catheter in the right hand, and secured with mepilex adhesive. Once IV access was obtained 30mg of propofol was given to facilitate intubation. Intubation was performed with a lubricated miller 1.0 blade, and 4.0 uncuffed ETT. During laryngoscopy it was noted the child had multiple oropharyngeal ulcers extending into the tonsillar pillars. The ETT was secured with the same mepilex adhesive. The surgery proceeded with a mixture of 3% sevoflurane, 30% oxygen, and 70% nitrous oxide technique without complication. At the conclusion of surgery an axillary block was performed for postoperative pain management. The block was performed using a 22 gauge stimex needle and peripheral nerve stimulator, a total of seven ml of 0.25% levobupivacaine with 1:200,000 epinephrine was injected in 1ml increments. The child was extubated without complication, was stable in the PACU, and subsequently went home one hour later in no obvious distress.
DISCUSSION
The choice of anesthesia technique in DEB, as well as the need for postoperative pain management need to be discussed with the parent and surgeon preoperatively. The aggressiveness of monitoring depends on the operation, and the overall general health of the child. Care should be taken to minimize skin and mucous membrane trauma. Venous access is invariably difficult, so inhalational induction techniques,
especially in pediatric patients are most successful. The greatest challenge is airway management, and the propensity to cause new oral bullae. Problems leading to difficulties in intubation such as restricted mouth opening or dental disorders are common. When possible, regional anesthesia techniques, which provide satisfactory postoperative analgesia may be useful.

REFERENCES

