



## Pediatric Anesthesia & Pain Management

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Guidelines for the Anesthetic Management of Epidermolysis Bullosa (EB)

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## Guidelines For The Anesthetic Management Of Epidermolysis Bullosa (EB)

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## Preoperative Assessment

### ***Establish the primary diagnosis***

There are several types of EB; they all result from a biochemical defect in the skin layers or basement membrane of squamous epithelium. This results in weakness at the basement membrane, a predilection to separation of the squamous cell layer from the BM after minor traumas to the skin, transudation of fluid into the disrupted skin (bullae formation), and scarring during healing. Children with EB are like chronically burned children. Most frequently the varieties that require surgical procedures and our involvement are:

1. **Junctional EB.** Autosomal recessive. Present at birth or soon after; variable severity; large ulcers are common; loss of nails, dysplastic teeth; infection, growth failure. Incidence = 1/50,000
2. **Dystrophic EB (Figure 1).** Autosomal recessive. Onset at birth; progressive scarring and deformity; oropharyngeal and esophageal involvement; mitten-hand deformity from progressive skin loss on hands during infancy and scarring with fusion of the digits; severe skeletal contractures secondary to scarring. Incidence = 1/1,000,000



**Figure 1.** A girl with dystrophic EB prior to a bath. Note the muscle wasting and growth failure of chronic malnutrition and the presence of extensive weeping open skin lesions.

### ***Establish all secondary diagnoses***

#### **Complications of EB include:**

1. **Hypermetabolic state.** Children with EB are chronically injuring and healing their skin. Caloric demands are significant, and growth failure is virtually uniform.
2. **Infection.**
3. **Oral scarring** with limited oral opening.
4. **Esophageal strictures.**
5. **Anemia.**

#### **Most Common Indications for Surgery**

1. Plastics procedures to correct pseudosyndactyly of the hands or feet (Figure 2).
2. Balloon esophageal dilatation (performed under fluoroscopy).
3. PEG or open gastrostomy.
4. GI Endoscopy.
5. Dental restorations.
6. Plastics procedures to increase oral opening.



**Figure 2.** Pseudosyndactyly of the hands of a child with Dystrophic EB.

## **General Principles of Management**

1. Shearing forces applied to the skin will result in bulla formation. Compressive forces to the skin are tolerated.
2. The columnar mucosa of the nares, and the larynx and trachea distal to the vocal cords, are not affected. There is no contraindication to tracheal intubation.
3. A special EB kit is kept in the Anesthesia Workroom that contains:
  - i. Koban® tape, Webril®, Ace® wrap, etc.
  - ii. Lubriderm® and other emollients
  - iii. Surg-o-Flex bandaging



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**Figure 3.** The "EB Kit" containing all the accoutrements needed to do a case on a child with EB.

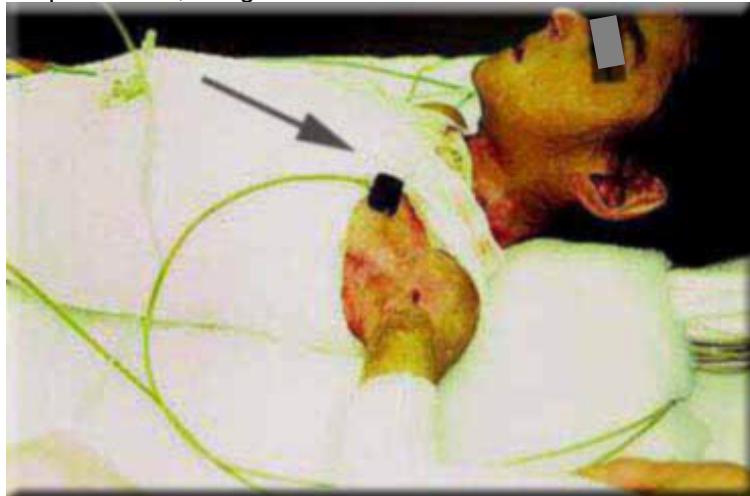
4. Adhesive tape, adhesive ECG electrodes, adhesive pulse oximeter probes are not used under any circumstances.
5. Padding should be generously used. Children should lie on a sheepskin. The sheepskin may be used for moving the children, as if in a hammock, to avoid injuring the skin. All skin should be wrapped with soft cotton dressings (Webril®).
6. All instruments placed into the mouth (laryngoscope, oropharyngeal airways) must be generously lubricated with a water based lubricant (KY® jelly, Lubifax®). Do not lubricate with Lidocaine® jelly. Avoid pharyngeal suctioning.
7. Face masks should be lubricated with Lubriderm® or other similar emollient.

### Monitoring

Routine monitors include noninvasive BP, ECG, core temperature, SpO<sub>2</sub>, and ETCO<sub>2</sub> if intubation is performed.

1. **ECG:** Using scissors, carefully trim the adhesive from adult ECG electrodes. The electrodes can be placed on the limbs and Koban tape can be used to

wrap the limbs, fixing the electrodes to the skin.



**Figure 4.** Notice the ECG electrode tucked under the Surg-O-Flex dressing to secure it to the skin. Also note the facial scars and resultant oral stricture.

2. **BP:** Wrap the extremity with Webril® cotton, and apply the BP cuff over the wrap.
3. **Oximetry:** Using scissors, carefully trim the adhesive off the pulse ox probe, wrap is around the palm of one hand, and wrap Koban® around the probe.
  - i. Or, use the probe without removing the adhesive backing.
  - ii. Or, use an adult clip-on probe.
4. **Temperature:** Use a lubricated axillary probe.
5. **Invasive monitors** may be required, and intra-arterial monitoring is recommended for lengthy cases.
6. **Intra-arterial monitoring** is reserved for lengthy cases lasting several hours to avoid the potential trauma of repeated inflation of the BP cuff.
7. **CVL placement** is infrequently needed if peripheral access cannot be established.

## Induction of anesthesia

1. **Premedication:** Sedative premedication is appropriate, and is nearly always indicated. Larger doses than usual are required. For a heavy premedication, use pentobarbital 5 mg/kg p.o. or [ketamine 10 mg/kg + midazolam 0.6 mg/kg] p.o.
2. **IV:** The goal of the anesthetic is to provide lack of awareness and analgesia with minimal handling of the patient. Placement of an IV followed by an IV induction is generally less traumatic than a mask induction.
  - i. IV placement is easier than one anticipates. The parchment-thin skin over the forearms allows veins to be easily visualized, and cannulated with small bore cannulae.
  - ii. Secure the IV by making a small slit in a 6 inch piece of Koban® tape 1 inch from the end of the tape, inserting the T-piece of the IV into the slit, then wrapping the tape around the extremity. Including the IV tubing in the Koban® wrap secures the IV very well.



**Figure 5.** Technique for securing an IV in an EB patient with Koban and cotton bandage. Note the adhesive tape is applied not to the skin, but only to the dressing itself.

3. **IM Induction:** If IV access cannot be established, then IM injection of Ketamine avoids mask induction.
4. **Inhalation Induction:** Inhalation induction is not contraindicated, particularly if the facial skin is not severely involved with EB. Lubricate the mask well, and beware of the trauma the fingers of your left hand will cause on the skin overlying the mandible. Use the gentlest pressure possible and avoid touching the face altogether if you can.

### Management of the Airway



**Figure 6.** Limited oral opening in a young boy with EB. Nasal fiberoptic intubation is the only alternative in this case for management of the airway.

1. **Less is more.** For peripheral surgery, continuous infusion of propofol ± remifentanil or ketamine to provide analgesia, with oxygen wafting by the face, is the best technique. Most children with EB will have a natural airway that is adequate to maintain oxygenation, and minor degrees of soft tissue airway obstruction are usually relieved by positioning the head to one side, or extending the neck with a towel.



**Figure 7. Inhalation induction of a child with EB.** Note the vaseline gauze applied to the face to prevent friction of the facemask against the facial skin. Also note the pulse oximeter attached to the hand with Koban tape, and the absence of ECG monitoring.

2. **LMA**s. There is nothing in the literature regarding the use of LMAs in EB, however it seems intuitive that a large silicon structure sitting on the larynx might cause mucosal damage. I do not recommend using LMAs for this reason.
3. **Intubation.** Direct laryngoscopy and intubation is usually not difficult provided the mouth will open sufficiently. Nevertheless, fiberoptic intubation causes less trauma to the mucosal epithelium than insertion of a laryngoscope, and should often be the first choice. Oral fiberoptic intubation may be performed after induction of anesthesia and establishment of TIVA so that you are not reliant on inhalation of gases to maintain anesthesia. Nasal fiberoptic intubation may be necessary for dental restorations, but the dentist is often able to work around an oral ETT. If nasal intubation is necessary, bullae may be induced at the opening of the nares by a tight ETT. Generous lubrication (not with Lidocaine® !!) will help to avoid this.



**Figure 8.** One technique for securing the endotracheal tube without tape, by snugly tying a surgical mask around the back of the head.

### Maintenance of Anesthesia

1. The ideal EB anesthetic for hand surgery: Propofol 50-100 µg/kg/hr - remifentanil 0.05-0.1 µg/kg/hr infusions with axillary nerve block. The nerve block may be performed with 0.5 ml/kg of 0.25% bupivacaine, or 1.5% lidocaine mixed with tetracaine 0.5 mg/cc after the establishment of the TIVA.
2. For dental/esophageal/GI procedures requiring intubation, any technique that permits a still patient and rapid arousal is ideal.

### Postoperative analgesia

1. Excellent postoperative analgesia is important to prevent thrashing in the bed/crib and further skin trauma. Anticipate the needs, administer local anesthetic or systemic analgesia before emergence, remembering the rapid metabolism the child will demonstrate. Children with EB will usually need unusually large doses of benzodiazepines and opioids to achieve a clinical effect.
2. Empiric use of acetaminophen and/or ketorolac is recommended



**Figure 9.** Children with EB live a tortured existence of daily pain and frequent medical procedures, with little hope of recovery or normalcy. Yet we must always remind ourselves that inside the disfigured body lives a child who wants to enjoy all the things in life a normal child enjoys, to play, to laugh, and to learn, a child with all the potential and need for joy of a normal child.

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### 25 Web Page Links:

- [Stanford University Blistering Disease Clinic](#)
- [National Epidermolysis Bullosa Registry \(NebR\)](#)
- [EB Medical Research Foundation](#)
- [Apligraf artificial skin](#)

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