EPIDERMOLYSIS BULLOSA

Surgical Specialty:	Multidisciplinary
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Background:

- General Considerations
  - Epidermolysis Bullosa (EB) is a group of rare genetic conditions characterized by severe skin fragility (1/50,000 – 1/500,000 live births).[1,2]
  - EB is inherited in an autosomal recessive manner. [1]
  - Patients with EB are extremely prone to developing bullae, which occur when the skin and mucosa separate from the underlying tissue due to shearing forces, resulting in blisters, peeling skin, ulcerations, chronic inflammation, nonhealing wounds, and scarring. Shearing forces will cause sloughing and damage, as opposed to perpendicular forces, which are tolerated.
  - There are four major types (over 30 varieties in total) based on the plane of tissue cleavage: Epidermolysis bullosa simplex (EBS) (within the basal layer of keratinocytes in the epidermis), Dystrophic epidermolysis bullosa (DEB) (in the upper dermis), Junctional epidermolysis bullosa (JEB)(within the lamina lucida of the basement membrane), and Kindler Syndrome (at various levels). [1,2]
  - Pediatric patients with EB, particularly those with severe forms, should be preferentially cared for at large tertiary pediatric centers.
  - Patients with EB come to the operating room most commonly for: esophageal dilations, dental work, gastrostomy tube placement, dressing changes, plastic and orthopedic procedures. [1,2]
  - Therapy is mainly supportive, although currently there are experimental gene- and cell-based therapies, as well as stem-cell treatment and protein replacement. [1,2]
- Patient Considerations/Associated Comorbidities
  - Any epithelial layer can be affected, therefore making it a multi-system disease with involvement throughout the body.
  - Squamous epithelium in the mouth is highly vulnerable to trauma, while pseudostratified respiratory epithelium (e.g, tracheal) is more resistant.
  - Over 50% of patients with EB may be difficult to intubate due to microstomia and ankyloglossia from repeated trauma to the oral cavity, fragile dentition, and contractures of the neck and mouth from scar tissue. [3]
  - Granulation tissue from blisters and erosions can cause laryngeal stricture or stenosis, hoarseness, and stridor. Therefore, an upper airway evaluation and possible surgical intervention, including tracheostomy, may be needed.

- There is a high risk for esophageal strictures and dysmotility, leading to esophageal stasis and an increased risk of aspiration.
- Anemia is common in patients with severe EB from chronic inflammation, losses from skin erosions, and reduced dietary intake. Some subtypes (simplex and dystrophic) may be associated with dilated cardiomyopathy due to iron overload from repeated blood transfusions for chronic anemia, dietary selenium, and carnitine deficiency. [1,2]
- Patients with EB may have frequent respiratory infections from ongoing aspiration as well as decreased pulmonary function from chest wall scarring.
- Recurrent blistering of the urogenital tract can lead to hydronephrosis and renal impairment, with end-stage renal disease reported in some severe recessive types.
- Patients with EB have extremely fragile skin and have extensive blistering, scarring, and fibrosis from healing wounds, leading to digital webbing, flexion contractures, and pseudosyndactyly.
- Malnutrition results in osteopenia, fractures, and slowed skeletal growth. Patients will have severe malnutrition, growth retardation, or failure to thrive, which can compromise immunity and lead to chronic infections. [1,2]
- Careful eye protection is necessary because scarring can lead to incomplete closure of the eyes.

Anesthetic Planning:

- Pre-Anesthetic Evaluation
  - Patients with EB who present for surgery require meticulous planning and multi-disciplinary coordination (anesthesiologist, pediatrician, surgeon, pain management, dermatologist, infectious disease) and a thorough preoperative assessment of all comorbidities. Multidisciplinary care should occur in large pediatric tertiary centers.
  - The clinical subtype should be identified, and the severity and extent of the disease should be investigated to guide preoperative testing, optimization, and diagnostic studies. [1,2]
    - Electrolyte disturbances
    - Anemia
    - Hypoalbuminemia
    - Hypercoagulability
    - Renal insufficiency
    - Echocardiography
  - Physical exam with a detailed airway exam is vital in preparation for all EB patients, given the risk of airway complications. The presence of a muffled voice or stridor should prompt an ENT evaluation.
  - Discussions to have with the surgeon/family
    - Extensive discussion with the families is needed regarding the risks of anesthesia and provocation of blistering or skin sloughing with airway management, intravenous access, and procedure-related risks.
  - Patient's current medication regimen (analgesics, antipruritics) should be reconciled to assess for chronic pain requirements.
- Specific or Unique Room Set-Up Requirements
  - Airway
    - Endotracheal and nasotracheal intubation, supraglottic airways, and nasal cannulas have been used safely.

- Mask ventilation is usually not difficult; however, the mask can result in significant shearing forces and trauma. The anesthesia mask should be well-lubricated.[5]
- Airway management can be challenging (>50% of EB patients); therefore, a difficult airway plan and cart with a video laryngoscope, fiberoptic bronchoscope, and airway algorithm should be at the bedside. Multiple sizes of tracheal tubes, supraglottic airways, and airway adjuncts with proper lubrication should be readily available. [3]
- Knowing the type of EB is crucial for airway management.
- The nasal passages are lined by respiratory pseudostratified columnar epithelium, which is less likely to blister than the stratified squamous epithelium of the oral mucosa. Therefore, a nasotracheal intubation with a fiberoptic bronchoscope is preferred in the more severe subtypes of EB.
- Direct laryngoscopy and oral intubation can be performed in patients with recessive dystrophic EB patients without bullae formation, postoperative stridor, or airway obstruction.[6]
- Supraglottic airways should only be used in the event of an airway emergency since they can cause blistering in the mouth and oropharynx.
- Prepare for a difficult airway complicated by fragile skin and mucous membranes.[1,2,6]
- The airway may be secured by covering the skin in a nonadhesive dressing such as a soft silicone tape (Mepitac™ or Mepiform™) and lubricated soft cotton ties behind the patient's occiput. Alternatively, a surgical mask may be placed under the patient's head and the ties brought anteriorly around the endotracheal tube.[3,4]
- ***Close attention should be given to lubricate anything that comes in contact with the patient- instruments, anesthesia mask, anesthesia provider's hands, and points of contact on the patient's face.*** [6]

- Medications
  - Peripheral intravenous access is usually difficult.
  - All induction and maintenance medications have been and can be used successfully. Ketamine is especially attractive for dressing changes or procedures in which airway manipulation can be safely avoided. [2] Parenteral or intramuscular ketamine may limit the need for peripheral intravenous access.
  - Avoid medications associated with post-operative pruritic (e.g, morphine). [1,2]
- Monitors
  - Modification of standard American Society of Anesthesiologists monitors is needed to prevent trauma to the fragile skin.
  - "Hydrogel/Sensitive" EKG electrodes should be used, but if unavailable, standard EKG electrodes should be removed from their adhesive and placed on the skin with lubricant and gauze posteriorly. Alternatively, EKG leads can be placed over 3M defibrillator pads (3M® Defib pads 2346N), or needle electrodes can be used.
  - Clip-on pulse oximeters are preferred. If not available, the adhesive side of the disposable pulse oximeter can be covered with a clear film prior to application to the digit.
  - Non-invasive blood pressure cuffs can be placed over gauze to minimize trauma, and, if safe to do so, less frequent cycling may be warranted.
  - Invasive arterial pressure monitoring can be used for longer procedures.
  - Well-lubricated axillary temperature probes are recommended.

        - ***Adhesives are strictly avoided.*** [1,2,4]
    - Postoperative monitoring
        - Admission to the ICU may be indicated based on the severity of the disease or the type of procedure performed.
    - Special Equipment:
        - Mepitac soft silicone "tape"
        - Hydrogel (sensitive)
        - 2" by 2" gauze for padding around lines and monitors
        - Curad petrolatum dressing
        - Coban wraps
        - Web roll
        - Large container of water-based lubricating jelly
        - Sheepskin or egg crate mattress
        - Fine scissors

Intraoperative Considerations:

- Primary perioperative management involves reverence for the skin and mucous membranes, acknowledging that any pressure, friction, or contact of any kind may cause blistering and sloughing. [2,3]
- Positioning
    - Operating room tables should be meticulously padded with sheepskin or an egg crate mattress. No pressure points should be left at risk from friction or rough surfaces.
    - Patients who can move with little assistance may be asked to move and position themselves. Otherwise, the parents or usual caregivers are of great help. For example, the child can be lifted using a draw sheet placed on the trolley, moved over, and placed on the OR table without any dragging.
    - Skin should be prepped by gently dabbing betadine when placing intravenous or other lines. Care should be taken to utilize perpendicular force rather than shearing forces, which cause tissue trauma. Lines should be secured with a non-adhesive dressing such as Coban, gauze, or, most preferably, with the material of choice that your patient/patient's family suggests is the best for the individual patient (they may present with their own supplies). Alternatively, lines can be sutured into place if needed.
    - Eyes should be lubricated but left untaped. In addition, or in place of, Vaseline-soaked gauze may be placed over the eyes.
- Induction
    - Consider premedication with midazolam, as it reduces anxiety and promotes a calm induction with minimal movement.
    - Mask ventilation is typically easy despite a potentially difficult airway. Pressure from mask placement may cause tissue injury, and lubrication should be used to reduce the shearing forces of mask placement. Maintaining a calm induction is crucial to prevent injury during mask application.
    - Inhaled induction may be preferred if vascular access is challenging.
    - The laryngoscope blade should be lubricated (e.g., KY® jelly).
    - For vascular access, the use of ultrasound or gentle direct pressure to distend the veins may be utilized. Elastic tourniquets should be avoided. IVs may be secured using a non-adhesive dressing or sutured in place if needed for a prolonged period of time.

    - To secure airway devices such as endotracheal/nasotracheal tubes or supraglottic airways, the skin should be covered in Mepiform and then secured using soft cotton ties. [2,3]
- Maintenance
    - Procedures have been successfully done with general, neuraxial, and regional techniques. The choice of technique is individualized to the patient and surgical procedure. [2] Local anesthetic infiltration should be avoided because it induces blistering.
    - Maintain euvolemia and replace blood loss.
    - Maintain normothermia.
    - For EB patients on opioids for chronic pain, higher narcotic doses may be required, and ketamine or alpha 2 agonists (clonidine, dexmedetomidine) may be considered. [1,2]
- Emergence
    - A smooth emergence is as important as a smooth induction.
    - If the airway was not difficult, consider a deep extubation to avoid coughing. Deep extubation may not be appropriate in all scenarios, and consideration should be given to a smooth awake extubation to avoid emergent airway manipulation.
    - There is a theoretical risk of airway obstruction from bullae formation; however, cases of postoperative obstruction are rare.
    - Airway adjuncts should be removed before emergence, and oxygen can be delivered with a cushioned and lubricated nasal cannula.
    - A smooth emergence, minimal postoperative nausea and vomiting, and adequate analgesia are of utmost importance to minimize trauma to the skin and mucosa. [1,3,4]
- Post-op Care/Disposition
    - For uncomplicated procedures, many patients may be able to be discharged home following recovery from anesthesia. [2]
    - Admission may be indicated based on the severity of the disease or the type of procedure performed.
    - Multimodal pain management is necessary to address both the acute and chronic pain of EB. [1,5]

Case-Specific Complications/Pitfalls

- It is helpful to keep a prepared “Epidermolysis Bullosa Kit” with all specialized equipment and printed protocol or handbook readily available for the purpose of caring for EB patients. [4]

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