

Perioperative Guidelines for Anesthesia Care for Patients with EB

Preoperative Assessment:

Airway assessment is critical

- Oral scarring results in limited mouth opening (microstomia)
- Mucosal lesions may be severe
- Dental caries may be present
- Tongue may be fused to floor of mouth
- Teeth may be angled inward
- Esophageal strictures may be very high
- Anticipate difficult intubation, consider oral fiberoptic intubation

Multiple Systems May be involved

Pulmonary:

Frequent respiratory infections
Aspiration
Decreased pulmonary function

Cardiac:

Dilated Cardiomyopathy
in patients with RDEB
possibly related to carnitine and/or selenium deficiency).
Consider preoperative echocardiogram

Musculoskeletal:

Extensive contractures
Digital fusion (mitten deformities)

Difficult IV access

Nutrition:

Increased caloric demand
Growth failure and failure to thrive
Anemia of iron deficiency and chronic disease

Skin

Extreme fragility
Blisters and erosions
Squamous cell carcinoma
Infection related to
compromised skin integrity
poor immunity due to malnutrition

Common Surgical Procedures

- Plastic surgery to correct pseudosyndactyly of hands, feet
- Balloon esophageal dilation under fluoroscopy

- Skin biopsies to rule out squamous cell carcinoma
- Excisional surgery with grafting for squamous cell carcinoma
- Dental rehabilitation
- Whirlpool treatments for skin debridement
- PEG or open gastrostomy
- GI Endoscopy
- Central lines
- Dressing changes

General Management Principles

- *No shearing forces can be applied to skin to minimize bulla formation.*
- *Compressive forces to the skin are tolerated.*
- *Lift, do not slide patient during transfer.*
- *All adhesive tape, ECG leads, adhesive pulse oximeter probes must be avoided.*
- *If patient dressings are in place and not in the way, leave in place.*
- *Columnar mucosa of nares, larynx, trachea distal to vocal cords are not affected.*
- *Tracheal intubation is acceptable.*

OR Preparation:

- Warm room
- Padded OR table
- Gentle transfer of patient.
- Egg crate mattress which stays under patient throughout perioperative period
- Lubricate eyes with preservative-free, non-lanolin ointment (Refresh®) and cover eyes with moistened gauze pads or use non-adherent adhesive (Mepitel® or Mepiform®)
- Assemble all necessary supplies ahead of time.

Suggested Supply Kit

- Non adhesive dressing: Mepitel®, Mepiform®, Mepitac tape® (Molnlycke Healthcare, Goteburg, Sweden, www.molnlycke.com) or Vaseline gauze/Telfa®
- Coflex® (Andover) wrap or Coban® (3M), gauze
- Cotton tape to secure endotracheal tube (ETT))
- Aquaphor® ointment to lubricate anesthesia mask
- Water based lubricant (i.e. Surgilube® by Fougera) to lubricate oral airways, laryngoscope blade
- Clip pulse oximeter probe

Oral Premedication to minimize struggling for mask induction

Monitoring: Reasonable Minimum

- Gauze padding under BP cuff
- Nonadhesive pulse oximeter probe (clip probe)
- Nonadhesive ECG leads (needle electrodes or normal electrode pads with adhesive cut off and secured with Mepitac tape® or gauze wrap)
- Axillary temperature probe if needed

Induction

- Mask induction common for pediatric patients
- Gentle pressure with well-lubricated mask
- IV induction may be difficult due to poor venous access

IV Access

- Tourniquet placed over gauze padding
- Secure IV with Mepitel or Mepitac
- Wrap with Kling® (J&J), Webril® (Kendall) Coban® (3M) or Coflex® (Andover)
- No adhesives!
- Check IV site frequently since IVs tend to become dislodged more easily.

Airway Management

- Mask lubricated with ointment (eg Aquaphor®)
- Avoid oral airway if possible, may cause oral blistering
- Gentle intubation with well lubricated laryngoscope and small ETT
- Anticipate difficult intubation
- Oral fiberoptic intubation if needed, avoid nasal intubation unless absolutely necessary.
- Laryngeal Mask Airway (LMA) may cause pharyngeal bullae.
- Secure ETT with nonadhesive cotton tape or suture to teeth.

Anesthetic Techniques

- General endotracheal anesthesia advisable for
 - Esophageal dilation
 - Dental rehabilitation
 - Abdominal surgery, major operations
- Mask anesthesia for brief procedures as appropriate
- Other options:
 - Total Intravenous Anesthesia (TIVA) with propofol \pm remifentanyl or ketamine for whirlpool treatments, peripheral surgery
 - Regional anesthesia: axillary block, spinal, epidural, caudal
- Muscle relaxants including succinylcholine are fine
- Avoid histamine releasing drugs e.g. morphine to minimize postoperative pruritus

Emergence / Post operative Care

- Emergence should be smooth to avoid airway, skin trauma
- Suction gently when needed with lubricated suction catheter
- Awake extubation to minimize airway obstruction and need for mask pressure on face
- Appropriate analgesia
- Prophylactic antiemetics to prevent post-operative nausea and vomiting
- Care for new skin lesions
- Monitor for airway compromise

Surgical Considerations

Cannot use adhesive grounding pads

Appropriate perioperative antibiotics

When prepping the skin with betadine solution, apply without friction and remove excess betadine gently blotting with alcohol

"A man never stands so tall as when he leans over to help a child." - Abraham Lincoln

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