Perioperative Recommendations 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 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)

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
* Difficult IV access and securement

Common Surgical Procedures

- 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
- 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, adhesive 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.***
- ***Nasal tracheal intubation is acceptable.***

OR Preparation:

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

Suggested Supply Kit

- Non adhesive dressing: Mepitel®, Mepiform®, Mepitac tape®, Mepilex®, Mepilex Transfer® (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 or use Mepilex Transfer to create custom mask to protect face
- Water based lubricant to lubricate oral airways, laryngoscope blade
- Clip or Velcro pulse oximeter probe. If clip or Velcro pulse ox is not available, remove adhesive and secure with silicone-based tape.

Consider Oral / g tube Premedication to minimize struggling for mask induction or to facilitate IV placement preop

Monitoring: Reasonable Minimum
- Gauze padding under BP cuff
- Non adhesive pulse oximeter probe
- Non adhesive ECG leads (needle electrodes or normal electrode pads applied directly over non adhesive gel defibrillator pad applied to the skin).
- Lubricated temperature probe if needed

Induction
- Mask induction common for pediatric patients
- Gentle pressure with protective face mask cut from Mepilex Transfer or well lubricated face mask
- Consider IV placement preop, with ultrasound as needed and IV induction, especially for older children / adults

IV Access
- Tourniquet placed over gauze padding
- Secure IV with silicone-based dressings such as Mepiform®,
- If recommended silicone-based products are not accessible, ask family if they have supplies on hand to use.
- Secure IV with adhesive dressing such as Tegaderm over Mepiform. No adhesive on skin!
- No adhesives!
- Check IV site frequently since IVs tend to become dislodged more easily.

Airway Management
- Mask lubricated with ointment (eg Aquaphor®)
- Alternatively, fashion protective mask to cover face with Mepilex Transfer

- Oral airway may cause blistering in mouth and may be difficult to insert
- Gentle intubation with well lubricated laryngoscope and small ETT possible in younger patients
- Fiberoptic intubation often necessary, especially in older children, adolescents, adults
- Anticipate difficult intubation
- Nasotracheal intubation may be easier and less traumatic than oral fiberoptic intubation. Smaller ETT may be needed to accommodate narrow nasal airways.
- Laryngeal Mask Airway (LMA) may be difficult to place, may cause mucosal trauma,
- Secure ETT with nonadhesive cotton tape. Can also be secured to head wrap or to teeth.

Anesthetic Techniques

- General endotracheal anesthesia advisable for
  Abdominal surgery, major operations
  Patients at risk for aspiration, difficult airway
- Mask anesthesia for brief procedures as appropriate
- Consider natural airway, spontaneous ventilation for appropriate procedures o
  Dental rehab, esophageal dilation, peripheral procedures such as lesion excision, hand surgery
  Total Intravenous Anesthesia (TIVA) with propofol ± narcotic, ketamine supplementation
  Nasopharyngeal airway, shoulder roll may help maintain natural airway
  Regional anesthesia acceptable
- Muscle relaxants including succinylcholine are acceptable
- 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
Use nonadhesive gel surgical grounding pad under patient
Appropriate perioperative antibiotics
When prepping the skin, apply without friction and remove excess betadine gently blotting with alcohol

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