Epidermolysis Bullosa

Alyssa Brzenski
Case

- A 4-year-old female with epidermolysis bullosa presents for orthopedic repair of pseudo-syndactyly release.
Figure 1
Level of cutaneous blisters in different types of epidermolysis bullosa. Adapted with permission (3).
Epidermolysis Simplex

- Autosomal dominant
- 1-2 in 100,000
- Most Common overall
- Mild disease
- Affects epidermis superficial to the basement membrane
- Blisters of then heal without scarring
Junctional Epidermolysis Bullosa

- Severe autosomal recessive disorder
- Mutation of the laminin 5 gene allowing separation between the dermis and epidermis
- Death often before 2 years of age
- Airway involvement
  - Larynx affected—recurrent stridor and risk for asphyxiation
  - Recurrent oral lesions making feeding difficult
- Sepsis
  - Poor nutritional state
  - Frequent severe blisters which can become colonized
Kindler Syndrome

- Most recent classification
- Autosomal recessive
- Blistering and photosensitivity
Dystrophic Epidermolysis Bullosa

- Most frequent type of EB seen by anesthesiologists
- 2 in 100,000
- Defect of the basement membrane and the dermis due to mutations of collagen 7
- Two forms:
  - Autosomal recessive (RDEB)- more common
  - Autosomal dominant (DDEB)
Airway

- Oral and pharyngeal blisters
  - Contraction of the mouth - Limited mouth opening
  - Fixation of the tongue

- Dental caries
  - Poor dental hygiene from pain of brushing
  - Poor nutrition
  - Defective enamel
GI

- Gastroesophageal reflux common
- Scaring leads to strictures and webs
  - Need frequent esophageal dilations
Cardiac

- Risk for Dilated cardiomyopathy
  - May be secondary to selenium or carnitine deficiencies
- ECHO screening frequently performed
Cutaneous

- Scarring common resulting in contractures and fusion of fingers and toes
- May present for orthopedic procedures
- Bacterial colonization- frequently MRSA
### Other Complications

**Table 1**
Complications of EB

*screening for squamous cell carcinoma begins at 10 years of age*

<table>
<thead>
<tr>
<th>Acute</th>
<th>Chronic</th>
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<tbody>
<tr>
<td>Pain</td>
<td>Pain</td>
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<tr>
<td>Fluid loss</td>
<td>Feeding difficulties with failure to thrive</td>
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<tr>
<td>Heat loss</td>
<td>Anaemia</td>
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<tr>
<td>Severe pruritus</td>
<td>Depression</td>
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<tr>
<td>Secondary bacterial infection</td>
<td>Constipation</td>
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<tr>
<td></td>
<td>Scarring and contracture formation</td>
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<td></td>
<td>Difficulties in mobility</td>
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<td></td>
<td>Osteopaenia</td>
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<td>Dental decay and periodontal disease</td>
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<td>Ophthalmological</td>
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<td></td>
<td>Stridor</td>
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<td></td>
<td>Renal dysfunction</td>
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<td></td>
<td>Cardiomyopathy</td>
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<td></td>
<td>Squamous cell carcinoma (second, third and fourth decade)</td>
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</tbody>
</table>
Table 3
Common surgical procedures in EB

<table>
<thead>
<tr>
<th>Procedure</th>
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<td>Change of dressing/Plaster of Paris</td>
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<tr>
<td>Repair of pseudosyndactyly/surgery to contractures</td>
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<tr>
<td>Dental extraction/conservation</td>
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<tr>
<td>Oesophagoscopy and dilatation</td>
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<tr>
<td>Open gastrostomy</td>
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<tr>
<td>Insertion of intravenous access (Portacath)</td>
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<tr>
<td>Ophthalmic surgery</td>
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<tr>
<td>Skin biopsy</td>
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<tr>
<td>Nissen’s fundoplication</td>
</tr>
<tr>
<td>Excision squamous cell carcinoma and skin grafting (adults and rarely children)</td>
</tr>
</tbody>
</table>
Bone Marrow Transplantation for Recessive Dystrophic Epidermolysis Bullosa

John E. Wagner, M.D., Akemi Ishida-Yamamoto, M.D., Ph.D.,
John A. McGrath, M.D., Maria Hordinsky, M.D., Douglas R. Keene, B.S.,
David T. Woodley, M.D., Mei Chen, Ph.D., Megan J. Riddle, B.A.,
Mark J. Osborn, Ph.D., Troy Lund, M.D., Ph.D., Michelle Dolan, M.D.,
Bruce R. Blazar, M.D., and Jakub Tolar, M.D., Ph.D.
General Considerations

- Shearing forces are traumatizing
- Pressure should not cause tissue damage
- Only squamous cell lined tissues are affected
- Columnar respiratory epithelium NOT affected so nasopharynx and trachea unaffected
Anesthetic Considerations- Premedication

- Should consider a premed due to
  - Frequent procedures
  - Thrashing could cause new blisters
Bedding

- Mere wrinkled sheets can lead to new blister formation
- Sheepskin minimizes friction and should be placed on the beds
- Patients should self-position if possible
Adhesives

- All adhesives are contraindicated
- Non-adhesive monitors should be used if possible
- Silicone based products should be used to secure all lines and monitors
- Silicone based products are easily removed with water
Lubrication

- Anyone or anything touching the patient should be lubricated

- Aqueous lubricants such as vaseline products or lacrilube should be liberally applied to hands, masks, and any instruments entering the mouth
EKG

- No EKG pads directly on the patients
- May not place EKG leads for a short case
- For longer cases, cut old defib pads and place on the patient with the EKGs on top.
Pulse Ox

- The easiest way to remove the sticky from the pulse-ox is to place a tegaderm over the adhesive side and secure it with coban.
Blood Pressure Cuff

- Shear forces, not pressure, causes new bullae formation

- Blood pressure cuffs should be used sparingly and dressings or unwrinkled web-roll should be under the cuff
Eye Protection

- Ocular lubricant should be used
- Mepitel sheeting can keep the eyes shut
IV Access

- IV access can be difficult due to
  - multiple IV placements in the past
  - limited access due to dressings
  - scaring

- Central lines/PICC lines are often a last resort as infection/sepsis is common in EB kids

- Malnutrition minimizes subcutaneous fat and visualization may be easy

- Tourniquet use is controversial—should place web-roll or dressing below the tourniquet

- Secure with Mepitac
Airway Management

- Inhaled induction tolerated well
- Small, scared opening with fixed tongue
  - Difficult oral intubation
  - Rarely obstructs
- Short procedures can be performed with a well lubricated fully inflated mask anesthetic
  - Minimize shearing—steady gentle pressure without moving your hand
LMA?

- Well lubricated LMAs have been used
- Placement may be difficult with minimal mouth opening
- Possible shearing force to the oral cavity
Intubation

• Early in life a direct laryngoscopy may be possible
  • Must lubricate the blade well

• Fiberoptic intubation preferred
  • Intubation through the mouth possible
  • FOB through the nare may be preferred- only the entrance of the nares is squamous epithelium
Anesthetic Choice

- Many different anesthetics used—neuroaxial, regional, general
- Even IM injections have been used

Caudal Epidural Anesthesia in an Infant with Epidermolysis Bullosa

Lawrence L. Yee, M.D.,* Joel B. Gunter, M.D.,† Charles B. Manley, M.D.‡

Bilateral ultrasound-guided axillary plexus anesthesia in a child with dystrophic epidermolysis bullosa
PACU

• Ensure good pain management
  • Thrashing can cause new blisters

• No oxygen facemasks

• Must give a good sign-out to the PACU nurses to ensure no complications
How would you provide anesthesia?
Sources


- Goldschneider K et al. *Perioperative care of patients with epidermolysis bullosa: proceedings of the 5th international symposium on epidermolysis bullosa, Santiago Chile, December 4-6, 2008*. Pediatric Anesthesia 2010; 20: 797-804.


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