Fragile Skin/Epidermolysis Bullosa Care Guideline

Inclusion Criteria:
- Known diagnosis Epidermolysis Bullosa (EB)
- Blistering fragile skin condition of unknown etiology
- Presence of Congenital localized Absence of Skin (CLAS)

Definition
Epidermolysis Bullosa (EB) is a chronic genetically inherited disorder

EB is characterized by extreme skin fragility and blistering caused by any amount of friction, trauma or heat

Assessment
- Vital signs
- Respiratory status
- Nutritional status
- Pain level assessment
- Duration/Extensiveness of blisters/lesions
- Assess risk for infection
- Assess need for intensive monitoring/nursing care

Recommendations/Considerations
Appropriate RN staffing to allow adequate time to minimize trauma due to admission process and care

Additional interventions
- For infants encourage oral feeds with fortified BM/formula
- Wound care, dressing changes utilizing 2 persons
- Stepwise approach to pain control
- For infants DO NOT use humidity
- Keep bed flat to prevent shear
- DO NOT apply ID band – place at bedside
- Gentle handling at all times
- Utilize Mepilex transfer with Aquaphor for open blisters and wounds, secure with Mepitac or stockinette
- If IV therapy is needed, secure only with Mepitac, or secure adhesive tape to gauze or Mepilex transfer.
- DO NOT use adhesive tape directly on skin
- Venous lab draws only - NO heel sticks please

Immediate Interventions
- Contact Isolation
- Apply thin coat of Aquaphor to gloves and all medical equipment prior to physical contact with patient
- NO adhesives of any kind should be used (tape, duoderm, tegaderm, stat lock, etc.)
- Least invasive measures for monitoring/intervention should be utilized
- Mandatory Wound Care consult
- Notify Clinical Nurse Specialist on admission to Unit

Discharge Criteria
- Family has mastered Wound Care and application technique
- Family has supplies needed for wound care
- Family has adequate/appropriate support
- Appropriate Emergency Letter issued

Patient Education
- EB handout
- EB resource list
  Link posted on PAWS PFE page

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Reassess the appropriateness of Care Guidelines as condition changes and 24 hrs after admission. This guideline is a tool to aid clinical decision making. It is not a standard of care. The physician should deviate from the guideline when clinical judgment so indicates.
References

Epidermolysis Bullosa Care Guideline


Caring for Newborns

To all new parents, here are some helpful hints and detailed medical information to assist you. Please contact Debra's National Office with further questions or concerns. At present, there is no specific treatment for EB. Current therapy is directed toward the prevention of skin trauma, prevention of infection, and the treatment of complications.

**Prevention of Blisters**

**Handle gently:** Avoid lifting babies or children from under the arms; instead, place one hand beneath the bottom and another beneath the head/neck to lift. A pillow, egg-crated (foam) pad or sheepskin may be used beneath the baby to prevent friction against the skin while lifting and holding.

**Do not rub skin:** Since blisters can be caused by friction, the skin should be patted rather than rubbed. Before blood tests or immunizations, the area can be cleansed by gently pressing or patting the alcohol pad against the skin.

**Dress in loose-fitting clothes:** Clothing that rubs the skin may cause blisters. Avoid or alter clothing with binding elastic. Avoid harsh buttons, snaps and zippers. Non-binding diapers or cloth diapers may be used. Elastic may be cut from the legs of disposable diapers to help minimize blistering.

**Avoid excessive heat:** Overheating tends to increase skin fragility. Maintain a moderate environmental temperature (including your car) and do not over-dress.

**Do not use adhesives on the skin:** Tape or band-aids should not be applied to the skin because they may cause blistering.

**Lubricate the skin:** Aquaphor or Vaseline will help decrease friction.

**Blister Care and Prevention of Infection**

**Wash hands before administering skin care:** Hand washing is the most effective measure to control infection.

**Do not remove clothing or dressings that are stuck to the skin:** Materials that are stuck to the skin should be soaked until they can be removed easily. This can be done at bath time (in the tub) or by applying room temperature water or a soaked compress directly to the bandage.

**Cleanse skin daily:** Skin may be cleaned with a very mild soap such as Dove.
**Drain blisters:** Blisters tend to increase in size if they are allowed to remain intact. For this reason, most blisters should be drained when they are about the size of a dime or if they appear tense. Sterile needles or lancets may be used to puncture the side of the blister roof. A tiny pinhole may re-seal and allow the blister to refill, so take care to open the blister roof adequately. The blister roof should be left intact to facilitate healing and comfort. A mild antibiotic ointment may be applied to the area to aid in the prevention of infection.

**Apply topical antibiotics to lesion:** Mild, over-the-counter antibiotics (Polysporin, Bacitracin) are effective in preventing infection and may be rotated every month or two to discourage bacterial resistance. Bactoban, like all antibiotics, should be used only when infection is present. (signs of infection include increased redness, swelling, pain and warmth). Prolonged use of Bactroban has been associated with the development of resistant Staph infections.

**Apply non-adherent dressings:** After application of topical antibiotics, a non-stick dressing such as Mepitel, Vaseline Gauze or Telfa should be applied to denuded or unprotected areas. Next, rolled gauze is wrapped around the non-adherent dressing and is then secured with a tubular dressing retainer. The dressing retainer will prevent the occurrence of tape accidents.

**Change dressing daily:** Daily dressing changes are recommended and can be coordinated with a bath or cleansing of the skin. Some contact layer dressings, such as Mepitel (Molnlycke Health Care) may be left in place for several days, although the secondary or outer dressing should be changed and the wound assessed daily.

**Nutrition**

**Wash hands before administering skin care:** Hand washing is the most effective measure to control infection. While "breast is best" for most babies, breast feeding presents a special challenge for infants with EB. Infants with blisters and lesions in the mouth may have difficulty nursing or sucking from a regular baby bottle. Mothers may decide to provide expressed breast milk to their infants. The Haberman feeder (http://www.medela.com) is a cleft palate nipple which incorporates a valve that facilitates easy delivery of the formula without the need to suck hard. This works well for babies with a fragile oral mucosa.

- Infant mouth care may include gentle cleansing with a spongy toothette
- A dry nipple will stick to lips and blistered areas, causing more damage. You may moisten the nipple with water or Vaseline prior to feeding

**Keep Growth Charts:** It is important to chart the growth of an infant/child with EB. Such charts provide essential information for evaluating the adequacy of the diet. A nutritionist should be consulted about the nutritional status of an infant with EB if there are questions or concerns about weight gain. Many infants with EB require fortified formula to meet their increased caloric and protein needs.

**Nurturing**

It is not unusual for nursing staff and parents to avoid handling a baby with EB, with the hopes of minimizing blisters. Learning the correct way to hold the baby will instill confidence in the caregivers and allow the baby to receive the emotional support and closeness he/she needs. It's a good idea to use the crib as a "Safe Place." All dressing changes and other unpleasant activities should be performed at places other than the crib (for example, on the changing table). The infant should learn to connect the crib with comfort.
**Continuity of Care**

Continuity of care by the nursing staff and other care-givers is essential.

You may contact Debra of America with questions or concerns:

(866) 332-7276  
nursegeri@debra.org

*Please note that all medical information given by Debra is for informational purposes only. Our information is not intended to substitute the care and guidance given by a qualified physician. All regimens of care should be discussed with the patient's physician. Always check with your physician prior to starting any medications or treatment regimens.

- See more at: [http://www.debra.org/newborns#sthash.9jqYNMta.pdf](http://www.debra.org/newborns#sthash.9jqYNMta.pdf)

**Frequently Asked Questions**

**Can you tell by my baby’s wounds what type of EB this is?**

No. A medical professional can give you an educated guess as to what type of EB your child has, but there have been many cases of these initial guesses being incorrect.

Think of all the symptoms that happen in an EB newborn as moving within each of the circles below - many symptoms are common in one form, but there is overlap of all the main forms of EB. Because of the overlap, the clinical picture cannot give the whole answer. Only a skin biopsy can confirm the type of EB a child has.

**Should I wrap my baby’s fingers individually?**

It is incredibly difficult to individually wrap a newborn’s fingers separately, and attempting to do so may actually cause more damage to the baby’s hands.

You may need to wrap the hands to protect your child from additional damage. Consider adding a ball of gauze impregnated with lubricant (ie white petroleum) to the palm of the hand so that the baby’s fingertips cannot rub on the base of the palm, causing damage.

**Which is better for my baby, cloth diapers or disposables?**

Some babies with EB do well with disposable diapers that are slightly larger and have the elastic cut out. Others do well with microfiber cloth diapers that wick moisture away from the skin. The most important thing is to ensure that urine is being absorbed and not sitting against the skin, causing breakdown.
**Can I breastfeed my baby?**

Some moms have been successful breastfeeding an EB baby. The things to consider are the calories your baby needs to heal and grow and the potential of damage to the baby’s mouth and lips that can occur during breastfeeding. If the nutritionist you work with says the baby can get enough calories, and the baby’s mouth tolerates breastfeeding, then you can certainly do so.

**Is it okay to use a pacifier?**

Some individuals believe the risk of oral damage is too high to allow use of a pacifier in an EB newborn. With that said, a pacifier can often be helpful in pain management during painful events such as dressing changes. Each family needs to weigh the risks and the benefits before deciding on pacifier use.

**What is the right way to wrap my baby’s wounds?**

There is no “right” or “perfect” way to dress EB wounds; many families have had success with a wide variety of methods of wrapping and types of medical products. It is important to have a primary contact layer, a secondary layer for padding and an outer layer to secure the dressing in place.

Additionally, some families find benefit from wrapping some “high risk” areas even when there are no wounds, in order to prevent damage.

**Will my baby get better with age?**

Some children with EB do improve with age, but for others the opposite is true. Unfortunately, it is not always possible to determine the prognosis for a child based on the diagnosis within the EB spectrum.

**My baby wiggles and rubs the back of his head on the sheet. I think he is itchy. What can I do?**

Itching is a common problem in EB. Using an emollient like white petroleum and keeping the skin moist can help. Some parents will also use a cool compress to reduce itching, but if symptoms persist, you can discuss medication options with your physician.
To Care Providers,

My child has been diagnosed with **Fragile Skin/Epidermolysis Bullosa**.

Please follow the **Fragile Skin/Epidermolysis Bullosa Care Guideline** for all patient care interventions.

Thank you for your assistance,

Wound Care Team

(Parents please bring this letter with you to all medical appointments including Emergency room visits and hospitalizations)