

# 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**)

## 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

## 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

## 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*

## 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

## 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*

**References**  
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- Caring for Newborns. (n.d.). *Debra of America-Dystrophic Epidermolysis Bullosa Research Association*. Retrieved from <http://www.debra.org/newborns>
- DebRA :: Dystrophic Epidermolysis Bullosa Research Association of America :: . (n.d.). *DebRA :: Dystrophic Epidermolysis Bullosa Research Association of America :: .* Retrieved November 19, 2009, from <http://debra.org>
- Denyer J, Pillay E. *Best practice guidelines for skin and wound care in epidermolysis bullosa*. International Consensus. DEBRA, 2012. Retrieved from [http://www.debra-international.org/fileadmin/media\\_data/4\\_DEBRA\\_International/Documents/CPG/Wound\\_Care\\_-\\_DEBRA\\_UK.pdf](http://www.debra-international.org/fileadmin/media_data/4_DEBRA_International/Documents/CPG/Wound_Care_-_DEBRA_UK.pdf)
- Epidermolysis Bullosa Clinic - GSDC - Dermatology - Stanford University School of Medicine. (n.d.). *Dermatology - Stanford University School of Medicine*. Retrieved October 12, 2009, from [http://dermatology.stanford.edu/gsd/eb\\_clinic](http://dermatology.stanford.edu/gsd/eb_clinic)
- Fine, J., & Mellerio, J. E. (2009). Extracutaneous manifestations and complications of inherited epidermolysis bullosa. *Journal of the American Academy of Dermatology*, 61(3), 367-384.
- Freeman, E., Kogelmeier, J., Martinez, A., Mellerio, J. E., Haynes, L., Sebire, N. J., et al (2008). Gastrointestinal complications of epidermolysis bullosa in children. *British Journal of Dermatology*, 158, 1308-1314.
- Frequently Asked Questions. (n.d.). *Debra of America-Dystrophic Epidermolysis Bullosa Research Association*. Retrieved from <http://www.debra.org/parentfaq>
- Gannon, B. A. (2004). Epidermolysis bullosa: pathophysiology and nursing care. *Neonatal Network*, 23(6), 25-32.
- Haynes, L. (1998). Nutritional support for children with epidermolysis bullosa. *Journal of Human Nutrition & Dietetics*, 11(2), 163-173.
- Haynes, L. (2007). Clinical Practice Guidelines for Nutritional Support in Infants and Children with Epidermolysis Bullosa. *eb Clinical Practice Guidelines*. Retrieved from [http://nutricia.co.uk/files/uploads/documents/EB\\_Guidelines\\_Final\\_for\\_web.pdf](http://nutricia.co.uk/files/uploads/documents/EB_Guidelines_Final_for_web.pdf)
- Langan, S. M., & Williams, H. C. (2008). A systematic review of randomized controlled trials of treatments for inherited forms of epidermolysis bullosa. *American Journal of Clinical Dermatology*, 34, 20-25.
- Lin, Y., & Golianu, B. (2006). Anesthesia and pain management for pediatric patients with dystrophic epidermolysis bullosa. *Journal of Clinical Anesthesia*, 18, 268-271.
- Ly, L., & Su, J. C. (2008). Dressings used in epidermolysis bullosa blister wounds: a review. *Journal of wound care*, 17(11), 482-492.

**References**  
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- Moss, C., Wong, A., & Davies, P. (2009). The Birmingham epidermolysis bullosa severity score: development and validation. *British Journal of Dermatology*, 160, 1057-1065.
- Newborn Care for EB. (n.d.). *EBnurse.org*. Retrieved from [http://ebnurse.org/assets/files/EBNurse\\_NewbornInfo\\_c.pdf](http://ebnurse.org/assets/files/EBNurse_NewbornInfo_c.pdf)
- Pai, S., & Marinkovich, M. P. (2002). Extracutaneous manifestations and complications of inherited epidermolysis bullosa. *American Journal of Clinical Dermatology*, 3(6), 371-380.
- Pope E, Lara-Corrales I, Mellerio JE, Martinez AE, Schultz G, Burrell R, Goodman L, Coutts P, Wagner J, Allen U, Lee M, Tolar J, Sibbald RG.(2012). *A Consensus Approach to Wound Care in Epidermolysis Bullosa*. Retrieved from [http://www.debra-international.org/fileadmin/media\\_data/4\\_DEBRA\\_International/Documents/CPG/Wound\\_Care\\_-\\_DEBRA\\_Canada.pdf](http://www.debra-international.org/fileadmin/media_data/4_DEBRA_International/Documents/CPG/Wound_Care_-_DEBRA_Canada.pdf)
- Price-Douglas, W., & Diehl-Svrjcek, B. (2007). Epidermolysis bullosa: a case study in transport, treatment, and care. *Advances in Neonatal Care*, 7(6), 289-294.
- Schachner, L., Feiner, A., & Camisulli, S. (2005). Epidermolysis bullosa: management principles for the neonate, infant, and young child. *Dermatology Nursing*, 17(1), 56-59.
- Schober-Flores, C. (2003). Epidermolysis bullosa: the challenges of wound care. *Dermatology Nursing*, 15(2), 135-144.
- Sibaud, V., Roul, S., Leaute-Labreze, C., Meneguzzi, G., & Taieb, A. (2002). Atopic dermatitis: therapeutic challenge in an infant with dystrophic epidermolysis bullosa. *British Journal of Dermatology*, 147, 350-352.
- Tabolli, S., Sampogna, F., Pietro, C. D., Paradisi, A., Uras, C., Zotti, P., et al (2009). Quality of life in patients with epidermolysis bullosa. *British Journal of Dermatology*, 161, 869-877.

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