Successful use of hydrophobic technology in neonates with Epidermolysis Bullosa.

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Epidermolysis bullosa(EB) is a family of mechanobullous genetic disorders characterized by fragility of skin in response to mechanical trauma. EB is caused by mutations in the genes that encode proteins of the basement membrane(BM) zone. Minimal, every day friction causes blisters and erosions in EB. The extent and distribution depend on the subtype. 4 main types include EB simplex(EBS), Junctional EB(JEB), dystrophic EB(DEB) and Kindler syndrome.

EBS-- disorder of abnormally coded keratin proteins 5 and 14, necessary for keratin stability within basal epidermal cells. Dysfunction leads to mechanical weakness, breakdown, and blistering, even with minimal friction. EBS is divided into basal and suprabasal subtypes.
JEB has a defect at the lamina lucida level of the (BM). The mutation in JEB arises from components of hemidesmosomes, filaments providing integrity and anchorage across the BM zone. JEB is associated with systemic complications.
Dystrophic EB has a defect in protein collagen 7, resulting in separation at the sub lamina densa part of the BM zone. There are 15 subtypes, depending on the relative amount of collagen7. DEB affects mucous membranes and leads to difficulty with feeding & bowel elimination

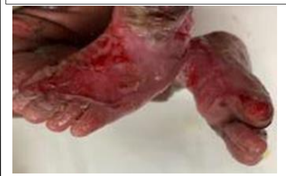
Skin care challenges :
***Blister prevention and management** ***Wounds protection**
***Skin colonization prevention/ treatment**
***Infection treatment** ***Safe securement of devices**

DACC (dialkylcarbamoyl chloride) is a hydrophobic fatty acid derivative
- Bacteria & fungi are inactivated once bound to the dressing and prevented from proliferating or releasing harmful toxins.
- Microorganisms are removed from the wound bed along with the dressing, reducing the bacterial load
- No cell wall disruption/no bacterial death therefore no systemic inflammatory reaction
- Non-hydrophobic (less pathogenic) organisms are left in the wound to stimulate healing

DACC family of dressings use in Epidermolysis Bullosa provide:
- **Skin protection from environment**
- **Bacterial colonization prevention**
- **Moist dressing for wounded areas**
- **Gentle cover for blisters**
- **Digits separation to prevent mitten deformity.**
- **Treatment of topical bacterial infection**

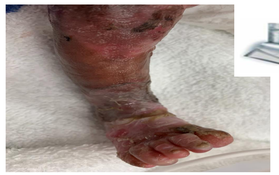
Case1– Dystrophic EB

Neonate with DEB was born with denuded blisters, evolving into wounds. DACC -hydrogel impregnated dressings were useful for moist healing and anti-microbial environment. He was eventually colonized with methicillin-sensitive Staph. Aureus, culminating in systemic infection. Continuous DACC –coated dressing application helped decreased topical colonization. Hydrogel promoted moist healing without too much moisture(excessive humidity/ moisture can promote blistering).

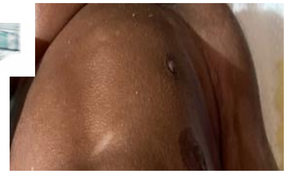


Denuded, bleeding skin Sloughed outer epidermal layer

Denuded blisters due to ongoing friction.



10 days post. Note dry, healing outer epidermal layer.



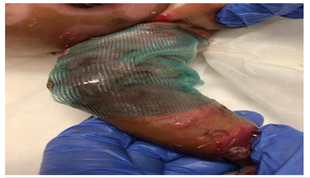
Prior to discharge. About 2 month after initial admission.

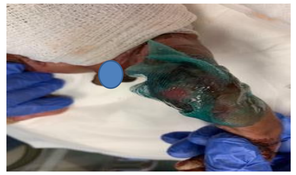
DACC *coated hydrogel dressing/ ribbon gauze or contact layer can be used for patients with deep fissures, dry areas of denuded skin or ruptured blisters. They provide mechanical protection, decrease colonization or treat infected areas/wounds.

*Cutimed Sorbact, Essity, Charlotte, NC

Case2-EB Simplex

Mild case of EBS, with blisters and denuded areas around groin area. DACC technology was protective from friction and bacterial skin colonization. As EB is characterized by lesions that heal and re-appear, dressing application was every 3 days on as needed "active " wounded areas.



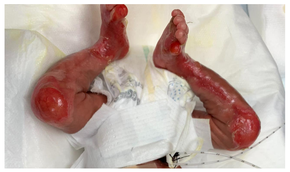


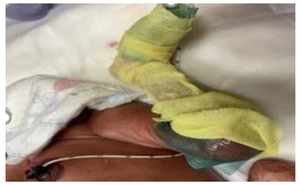
Affected areas covered with hydrogel-impregnated DACC dressing. Secondary dressing (Kerlex)was utilized to secure DACC dressing, avoid premature drying and decrease the need for frequent changes. Dressing was changed every 3 days.

DACC coated ribbon gauze was used often as a soft protective covering once skin lesions were healed.

Case3- EB Simplex

Preterm neonate with EBS had ongoing skin blisters/erosions. Systemic organ-specific complications were significant, including necrotizing enterocolitis, systemic bacterial sepsis and ongoing bacterial cutaneous colonization. Initial prematurity required some humidity to maintain electrolyte balance, contributing to slow blister healing. Many pieces of equipment required securement and maintenance, leading to further friction and blister formation.





In effort to minimize isolette humidity and keep DACC dressing from drying out, secondary petrolatum/ bismuth impregnated gauze was used.

Conclusions

DACC* technology effectively binds and removes hydrophobic organisms <u>without</u>:
Systemic/local absorption of chemicals from an antimicrobial
Endogenous systemic reaction, chemokines production
Release of bacterial by-products, which may act as exogenous inflammatory agents

Hydrophobic technology should be considered in the care of patients with EB as it provides effective skin physical protection, maintains a moist wound environment, physically binds and removes microorganisms without the use of an antimicrobial, while allowing non-toxic, easy-to-apply dressing care that can support healing without the need for daily manipulation

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