FEATURE EDITOR: Maureen Hanlon, RN, MN, CETN

Options in Practice presents different management approaches to the same clinical situation. You are invited to submit a brief case description, including the specialty nursing care provided, and several glossy, color photographs of the clinical situation. The case material will then be sent to another wound, ostomy, or continence care nurse, who will also address management concerns. Alternative solutions to difficult wound, ostomy, or incontinence clinical situations will be published.

SKIN AND WOUND CARE MANAGEMENT FOR A CHILD WITH EPIDERMOLYSIS BULLOSA

Case Situation

A 3-year-old girl was born with epidermolysis bullosa (EB). Her EB lesions have been generalized, affecting her feet, elbows, buttocks, hands, face, neck, and chest (Figures 1 and 2). She has had oral lesions in the past and currently has lesions on her lower extremities and eyes. She also has a history of wound infections, which are usually manifested by increased wound drainage.

Epidermolysis bullosa is a disabling skin condition that has a wide range of clinical severity and manifestations.[1] In mild cases, patients experience limited areas of blistering, usually affecting the hands, feet, elbows, and knees. More severe cases are characterized by extensive, generalized skin and mucosal lesions, with blistering, erosion, and scarring. Severe EB lesions resemble serious burn wounds, but they recur. The trauma associated with friction or normal childhood activities, such as running, playing, and falling, intensify the blistering process. Webbing of the digits is also common among patients with EB.

According to the Dystrophic Epidermolysis Bullosa Association of America (DEBRA), an estimated 25 000 to 50 000 Americans have some form of EB (Box).[1] Blisters often form at birth or during infancy. The blistered skin produces erosions, which heal slowly and carry a risk of scarring and subsequent skin atrophy. Repeated cycles of blistering and scarring can lead to digital fusions and flexion contractures. Pigment changes, that include either hyperpigmentation or hypopigmentation, can accompany this process as a result of the permanent disruption of the melanocyte activity.

Mucosal involvement of the mouth, pharynx, esophagus, and anus is common. The painful oral and pharyngeal lesions can interfere with eating, leading to nutritional deficiencies, impaired health, and painful or difficult elimination. Esophageal erosion and stricturing can further complicate the patient's fragile nutritional status. Gingival regression and periodontal disease may occur if the lateral gingiva adheres to the buccal mucosa. Milia formation is common in chronically traumatized and scarred skin. Nail dystrophy also occurs and includes thickened, discolored deformity of the nail, leading to nail loss with residual scarring of the nail bed.[1]

Blood loss associated with the denuded and eroded skin can lead to anemia. The patient with EB is also at risk for infection and negative nitrogen balance, owing to the loss of the protective layer of skin and mucosa as serum protein is lost by draining lesions. In all cases, treatment of EB is directed toward the symptoms and is largely supportive. Care should focus on prevention of infection, protection of the skin against trauma, attention to nutritional deficiencies and dietary complications, minimization of deformities and contractures, and psychologic support for the entire family.

Ann Lapioli-Zufelt, RN, BSN, CWOCN: The goals of skin and wound care for this patient included reducing the incidence of infection, facilitating wound healing of the involved areas, and providing protective cushioning against friction. Throughout the first 3 years of her life, the patient's family applied a topical antibiotic ointment on the lesions, followed by a nonadherent cover dressing. The choice of dressing ranged from highly to minimally absorbent. When the dressing adhered to the lesions or skin, its

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removal was painful and caused trauma to the budding epidermis. A multitude of dressings, including hydrogels, petroleum-impregnated, foams, and nonadherent dry dressings, had been tried but were painful and did not achieve healing. Therefore the patient feared dressing changes, and they proved stressful for her parents and other care providers.

To reduce the pain and trauma produced by dressing changes, I decided to try the Mepitel nonadherent silicone dressing (Mölnlycke Health Care, Eddystone, Pa), a fine-mesh dressing coated with a medical-grade silicone. The dressing did not adhere to the moist wound sites, but it did adhere to the surrounding dry intact skin. Removal, therefore, proved nontraumatic. This dressing also was pliable and conformed to contoured body surfaces such as the fingers, toes, and elbow. The porous design of the dressing allowed wound exudate to pass into an outer absorbent cover dressing, and it was permeable to the antibiotic ointment. This topical antibiotic was important, because it effectively managed secondary skin infections.

The cover dressing was removed and reapplied daily to allow application of the topical antibiotic. The Mepitel dressing was changed on an as-needed basis, after cleansing the lesions and drying the surrounding skin; it was usually left in place for 3 to 4 days. The most dramatic response to the new dressing was the alleviation of the pain and anxiety produced by dressing changes, which relieved both the patient and her caregivers. The patient's primary caregiver, her mother, learned the dressing technique quickly. The Mepitel dressing also provided a moist environment that promoted wound healing.

The parents were taught skin care principles designed to decrease the development of additional lesions. In EB, even slight friction can cause blisters; therefore, minimal and gentle handling is absolutely necessary. A cool environment, avoidance of overheating, and skin lubrication to reduce friction can lessen blister formation. Clothing must be made of a soft, nonirritating fabric that is easy to put on and simple in design to facilitate removal.

In addition to topical therapy for her wounds, we also addressed systemic factors including her nutrition and her blood count. This active, growing girl routinely drinks a protein and calorie supplement to maintain a positive nitrogen balance for tissue regeneration and to replace fluid and protein loss associated with blistering. Fortunately, she has not had a problem with anemia. Nonetheless, this patient and her parents will continuously need to learn to adapt and live with her chronic skin condition.

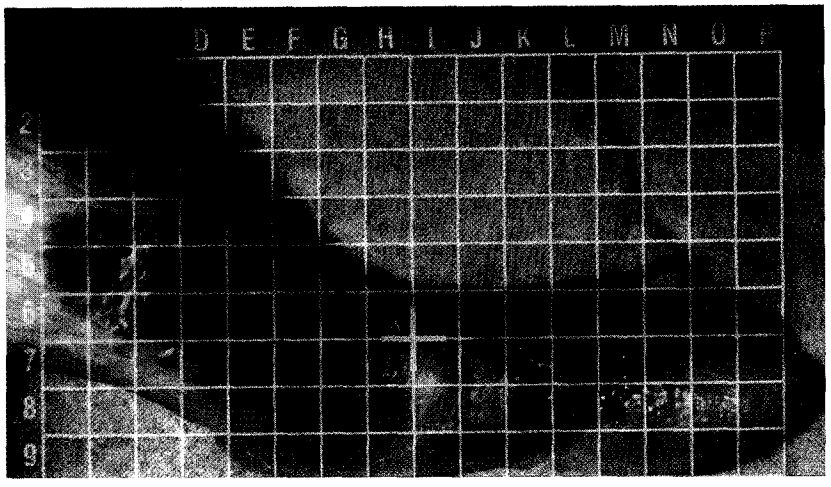

Figure 1. Epidermolysis bullosa (EB) lesions on left foot and lower leg.

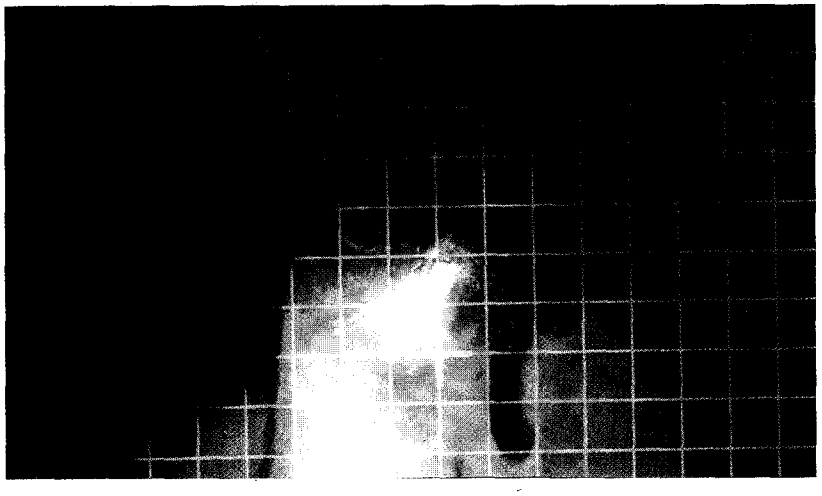

Figure 2. Moist, red, denuded epidermolysis bullosa (EB) lesions on right knee.

Box. Epidermolysis bullosa resources

The Dystrophic Epidermolysis Bullosa Association of America (DEBRA),
40 Rector Street,
New York, New York 10006

EB Treatment Centers:
The Children's Hospital of Philadelphia
Rockefeller (NY) University Hospital
Washington University in St. Louis

An excellent pamphlet, entitled *Living with Epidermolysis Bullosa,* can be obtained from the US Department of Health and Human Services.

E. Jill Morris, RN, MSN, FNP-C, ET: Fortunately, EB is a disease rarely seen in general WOC clinical practice. Genetic counseling, in many cases, has been beneficial, and it is possible to prenatally diagnose EB by fetal skin biopsy. Biopsy is helpful in determining prognosis, which can ultimately assist in dressing choices. There are 3 types of EB, which are differentiated by biopsy: simplex, junctional, and dystrophic.[2] All types are typically represented during the newborn period. *EB simplex* is an inherited autosomal dominant trait. Blistering may be mild to severe and present over the entire body or localized to the extremities. This type does not scar, and infection is the major complication. *Junctional EB,* the most common form, is autosomal recessive and presents as generalized bullae. This nonscarring form is usually fatal, as a result of infection and serum loss, within the first year of life. Oral and esophageal complications are common. *Dystrophic EB* may exhibit as either autosomal dominant or recessive. The recessive form is associated with scarring, developmental and growth delay, nail loss, and severe oral manifestations. Syndactyl is occasionally found and is secondary to chronic cycles of blistering and scarring.

I certainly agree with the skin and wound care protocols described in this article. I would suggest also including oral and dermal pain control measures, as well as interventions to prevent heat loss. Any significant disruption of the skin integrity results in diminished capacity to control and maintain body temperature. Many types of dressings are available that may serve to minimize or eliminate hypothermia. In older children and adults, a slurry of equal parts 1% lidocaine viscous and Maalox is useful in alleviating oral pain. Emla cream is useful for dermal analgesia in intact skin.

Supportive care may also include medications commonly used for burn care, such as silver sulfadiazine cream, secured with a variety of dressings, keeping in mind the degree of intact skin. I have also used both gel-sheet dressings and nonstick gauze without peripheral disruption of skin integrity. One of my colleagues has used large transparent dressings that successfully prevented friction and hypothermia. The dressings were applied and overlapped to most of the body surfaces. As her patient, a child, grew, the dressings lost tenacity and were replaced without traumatic removal.

A necessary consideration when using Mepitel is the potential stripping of weak epidermal layers in the early stages of scarring, because the dressing can adhere to intact skin. Mepitel does appear to have many positive characteristics for application to denuded skin. Cost is always a consideration, and we should continue to search for a combination of reasonable cost and clinical efficacy.

REFERENCES

1. Living with epidermolysis bullosa. Prepared by the Office of Health Research Reports, National Institute of Arthritis, Diabetes and Digestive and Kidney Diseases. Bethesda (MD): US Department of Health and Human Services, Public Health Service, National Institute of Health.

2. Cohen BA, et al. Dermatology. In: BJ Zitell, HW Davis, editors. Atlas of pediatric physical diagnosis. St. Louis: Mosby-Wolfe; 1997.