INTRODUCTION

Epidermolysis bullosa (EB) is the generic term for a large group of inherited skin fragility disorders. There are many types of EB but the common factor is a tendency for blistering and stripping of skin and mucous membranes following minimal everyday friction and trauma. EB varies in its effects from relatively minor blistering of the hands and feet (Localised EB Simplex) to increasing disability resulting from internal and external contractual scarring (Severe Generalised Dystrophic EB) in its most extreme form (Herlitz Junctional) EB leads to death in early infancy from complications including laryngeal blistering and failure to thrive.

Those affected with severe forms of EB have a tendency to develop chronic wounds and the later complication of squamous cell carcinoma often lead to death in early adulthood. At present there is no cure or effective treatment for this painful and devastating condition, but research is progressing towards availability of both gene and cell therapies. Treatment is symptomatic with emphasis placed on skin and wound management, pain relief and nutritional and psychological support. The presence in a Neonatal Unit of an infant with a severe form of EB poses challenges for the staff as knowledge of the condition and previous experience of specialised care may be lacking. Traditionally EB dressings are complex, requiring a multi-layered system in order to achieve non-adherence, correct positioning (to avoid deformity), exudate management, protection from limb movements and handling and anti-microbial cover. Such complex dressing regimens pose difficulties for staff who are not experts in wound care or management of infants with EB.

Aim

Dressing evaluation in regards to;
- Healing
- Control of odour and exudate
- Ease of application and removal
- Duration of dressing changes
- Reduction in pain (Neonatal Infant Pain Scale / FLACC pain score)
- Infection frequency

Method

Polymeric membrane* dressings are designed to be used as stand alone dressings negating the need for a separate primary and secondary dressing. Initially we were concerned about adherence and used polymeric membrane in conjunction with a trusted non-adherent wound contact layer. As our confidence grew in the product we were able to recommend the dressing be placed directly over the wound thus reducing dressing time, trauma and emotional distress for both patient and nurse.

6 severely affected neonates with forms of junctional or dystrophic EB were selected for the study. The four presented here show typical results representing the entire group.

Results

The dressing was easy to apply and to remove. More rapid healing was noted than with previously recommended dressings. Daily dressing changes were required initially due to the copious exudate. Addition of polymeric membrane wic reduced the frequency of changes.

Pain scores reduced in comparison to previously recommended dressings. Antibiotic and anti microbial treatments have not been required to treat skin infections or critical levels of colonization in any of the 6 infants selected for the case studies, with one infant not requiring such treatment for over 18 months. With previously recommended regimens regular courses of oral antibiotics and topical therapies have been necessary.

Discussion

The study demonstrated a rapid improvement in wound size in all cases. Of particular importance is the reduction of time during dressing changes in this vulnerable age group. The study has highlighted the lack of specialized dressings being readily available to neonatal units meaning inappropriate dressing materials are applied initially.

The initial study included infants with severe generalized dystrophic EB only, and now has been extended to children with other forms of severe EB. A significant factor has been the ability of polymeric membrane dressings to prevent critical colonization and infection. Infants with severe forms of EB are unable to be bathed until prenatal and birth induced wounds have healed as it is not possible to protect from additional damage from handling or from contact with the surface of the bath. The cleansing ability of the polymeric membrane dressings appears to offer a solution to this aspect of wound management.

Case 1

“Baby M” was the first neonate with severe generalized dystrophic EB to use polymeric membrane dressings. He is now 17 months old with. At birth Baby M had extensive wounds over both legs as a result of prenatal and birth induced trauma. His wounds were immediately covered with paraffin tulle and dry gauze resulting in additional trauma when these were removed. The lesions were then dressed using recommended atraumatic dressings for infants with EB. However, the wounds kept on deteriorating. Polymeric membrane dressings were then applied and changed on alternate days due to the copious amounts of exudate. Rapid healing was observed and we were able to reduce dressing changes to every 3-4 days in response to the reduction in exudate. Dressing change time decreased greatly due to the ease of application and removal of polymeric membrane dressings and the use of a single sheet dressing rather than a wound contact layer plus a secondary dressing. We have found that it is not necessary to add antimicrobial agents when using polymeric membrane dressings.

Infants with this level of severity of dystrophic EB generally demonstrate a higher incidence of chronic wounds by this age. The quality of skin over healed areas is better than generally predicted in infants with this form of EB who have suffered such extensive prenatal damage. By continuing to use polymeric membrane dressings for protection of vulnerable healed areas we have noted that when a new lesion develops the action of the polymeric membrane dressings appears to inhibit progression of the wound.

Case 2

MK is the first child of unrelated parents. At birth she presented with a deep wound over the dorsum of her foot. The great toe was shortened indicating the wound had been present for several weeks and suggesting a diagnosis of dystrophic EB. However, skin biopsy results point to Kindler syndrome which was included in the EB classification in 2008. The wound was dressed shortly after birth using an soft silicone wound contact layer and a secondary foam dressing. After 2 days the wound became wet and sloughy and the dressing was replaced with polymeric membrane and polymeric membrane wic dressings. These were changed daily. The wound looked much cleaner after a couple of days and healed rapidly.

Case 3

Baby H was born by caesarean section. She had deep sloughy wounds on both hands and both feet. Her diagnosis was later shown to be Non-Herlitz Junctional EB. Lack of Collagen XVII results in delayed and poor healing for those affected with this type of EB.

Case 4

Baby G has Herlitz junctional EB. He was born with a deep, extensive wound covering his left leg. Experience has shown that prenatal wounds in this type of EB do not heal and are generally present until the infant dies. In general we find that many of the atraumatic dressings used today prevent extension of the wound but have no impact on healing. Polymeric membrane dressings were applied at the first dressing change and changed daily due to the large volume of exudate. The wound remained clean and is healing well. We have never had this positive experience in a baby with this form of EB before.