

Reduction of Friction and Shearing Forces in Epidermolysis Bullosa

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Introduction

This poster describes the use of products made from Parafricta™ fabric for children with a severe form of epidermolysis bullosa.

Epidermolysis bullosa (EB) is a group of genetically determined skin disorders for which there is no effective treatment and as yet no cure. The common factor is an extreme fragility of the skin and mucous membranes with the susceptibility of blistering and wounds in response to minimal friction and everyday trauma.

There are many different types of EB, the 3 main groups being simplex, junctional and dystrophic. Severity ranges from minor blistering of the hands and feet to death in early infancy.

Management of the skin is by lancing blisters, dressing open wounds and protection of vulnerable areas. Nutritional supplements are necessary in many cases and often enteral feeding is required. Those severely affected require input from a large multi disciplinary team. In the absence of gene therapy treatment is largely symptomatic and with emphasis placed on prevention of trauma and the minimising of disability.

Adequate pain management is an essential part of daily care, although effective management is difficult due to variation in symptoms on any one day and the multiple contributing factors.

We have recently been using products made from Parafricta™ material in an attempt to reduce the amount of blistering caused by trauma from traditional bedding. Parafricta™ is a unique fabric in that its friction coefficient is like ice. The fabric was initially developed to reduce the potential development of pressure ulcers on patients with compromised skin.

Method

A pillowcase incorporating Parafricta™ fabric was given to 11 infants and children who have a severe form of EB.

Infants with a severe form of EB cannot be handled in the traditional way as extensive skin damage may result. We recommend infants are carried on a suitable soft pad, such as a Spenco Incubator Mattress. For the purpose of the study the mattress was placed inside a pillowcase made from Parafricta™ fabric. Older children who used a pillow at night were given a Parafricta™ pillowcase to use in place of their regular pillowcase.

They were asked to fill in a questionnaire following one month's continuous use of the pillowcase.

Results

- 11 children or parents reported the fabric to feel cool to the touch
- 1 child stopped using the pillowcase as she turned her pillow over in the night and suffered skin trauma from contact with the cotton fabric side
- 8 children reported the pillowcase to be comfortable (2 children were too young to comment)
- 10 children (or parents of infants) reported a reduction in the amount of blisters and skin stripping to the head and neck
- 2 children experienced chronic wounds sticking to the fabric
- 2 parents found the fabric to become thinner and worn following repeated washing
- 10 children (or parents of infants) wished to continue using the pillowcase

Case Study



Fig 1

A 12 year old girl with severe recessive dystrophic EB (RDEB) has chronic wounds all over her body. The most extensive of these is an area covering the whole of her back (fig 1). This wound has been present for several years and is unlikely to heal due to the underlying gene defect. Despite using a suitable mattress she found moving in bed very difficult as the resulting friction caused her dressings to slip and the previously intact peri-wound skin to tear. Recently she has been sleeping on a bed sheet containing Parafricta™ material and finds she is able to turn unaided during the night without pain and minimising risk of further skin damage.

Repeated blistering following trauma from her bedding and the subsequent scarring associated with RDEB caused her ear to fuse to her head, making it difficult for her to wear her glasses. Her ear was surgically separated and a skin graft applied. The result was excellent, but we were concerned trauma would cause fusion to occur again. Following one year of sleeping on a Parafricta™ pillowcase her ear has remained unblistered (fig 2).



Fig 2



Fig 3: Trauma to neck



Fig 5: Mittens made from Parafricta



Fig 4: Corneal abrasions in a neonate



Fig 6: Trauma to ear

Discussion

Further work will need to be done to establish the effectiveness of Parafricta™ across a larger group of patients. It may also be of benefit to establish whether it is both possible and beneficial to incorporate the anti friction and anti shear properties of Parafricta™ into other products used by this patient group.

Conclusion

Parafricta™ has proved to be a useful adjunctive therapy in the prevention of blistering and management of wounds seen in severe epidermolysis bullosa. Mittens made from Parafricta™ fabric have reduced the amount of damage caused by infants rubbing their faces and may reduce the rate of friction induced corneal abrasions from contact with regular cotton baby mittens. Products are available from www.parafricta.com

DebRA

DebRA is a registered charity set up to help people affected by EB. They provide information, practical help, and professional help from nurses, social workers and welfare workers and a means of communicating to families and healthcare professionals via publications and conferences. The majority of the work into finding gene therapy treatments for EB is funded by DebRA.

