

I have been working as a nurse for children with epidermolysis bullosa for 18 years. I am fortunate to be fully funded by the charity DebRA as this has meant the role has not been eroded by pressures within the health service and enables me to continue to work exclusively to help children with this devastating condition.

Epidermolysis bullosa (EB) is a rare genetically determined condition. There are many distinct types within this large group but the common factor is the extraordinary fragility of the skin, causing it to blister or shear away in response to the minimal traumas associated with everyday living. Internal membranes are also affected and those with severe forms of EB repeatedly suffer blistering in the mouth, throat and eyes.

My contact with the children and their families begins when I receive a phone call from a maternity hospital to say they have delivered a baby they think may have EB. My heart still sinks after so many years and I start hoping the baby will have a mild form of EB. I give advice for immediate care and arrange to see the baby as soon as possible, when I will take a skin biopsy to determine the type of EB and start teaching dressing, handling and feeding techniques. Often the parents have been told not to touch the baby and enabling mum to have her first cuddle is a very special moment.

One of the hardest things I have to do is to give the results of the skin biopsy. To tell parents that their new baby is likely to die before they reach their first birthday, or to become progressively disabled and die as a young adult is so cruel and I take away their dreams of watching their child grow up. I hope through practical and emotional support to recreate some of that dream even though time is so limited.

I am often asked “how can you still do this work?” As well as it being rewarding in many ways, one reason is that so much progress has been made in dressings and other materials which have really improved quality of life for the children.

I would say the most exciting new product in my career is Niltac™; a sting free silicone medical adhesive remover. Niltac™ effectively disables adhesives and has changed the way we work with the children. In the past we were unable to use any adhesive material because when the tape is removed the skin comes with it. This restriction made it difficult to secure drips and tubes and to monitor cardiac function. Now we are able to use sticky tapes to secure the equipment in the knowledge that we have the means to remove it safely. I used to dread getting a call to say someone had inadvertently stuck tape on a child, now I am quite relaxed about it as I know it can be removed without harm.

Niltac™ also works on non -adhesive materials which have stuck to the wound – clothing, bedding and dressings. Imagine waking up to find your ear has blistered in the night and has stuck to the pillow – a common occurrence for those with severe forms of EB. This used to be a long painful; process to release it –

soaking with water and gently easing greasy ointment under the ear. Now a quick spray with Niltac™ and the child can sit up!

Recently I was called on a Sunday evening by a paediatrician who was caring for a newly born baby with extensive wounds over both his legs. He told me the nurses had dressed the wounds with a “non adherent dressing”. I knew the dressing would have adhered to the wounds and had a night filled with worry over what I would find when I saw him the following day. My worries were well founded – the dressings seemed welded to the wounds. My options were limited – bathe the baby and try to soak the dressings off, use greasy ointments or try Niltac™. Bathing may have worked, but it is not advisable to expose all the wounds at any one time as it is not possible to protect the exposed skin and the baby would have suffered a lot more damage. Greasy ointments take a long time to free the dressings as they must be carefully worked under the dressings. The residual grease makes it difficult for new dressing to stay in place risking extension of the wound.

I applied Niltac™ to the surface of the dressing and gradually sprayed it under the edges. The dressings were removed easily and without trauma.

Niltac™ is now part of the recommended formulary for those with EB and I know parents won't leave home without it!

And of course what is better for the child improves the nature of their EB nurse as well!

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