Inform Rebecca Saad NEBDS Podcast Ep 20

SAMSARA:

Hello and welcome to Inform, a podcast where you'll be hearing from people with disabilities, as well as industry experts, on a range of topics.

I'm your host, Samsara.

Epidermolysis Bullosa (or EB) is a rare disease characterized by fragile and blistering skin which requires the frequent application of dressings to repair and protect wounds. It is estimated that there are around 1,000 people in Australia who have some form of EB and over 500,000 worldwide.

In this episode of Inform, we are talking with Rebecca Saad about the National Epidermolysis Bullosa Dressing Scheme. NEBDS is a government funded scheme which supports people with EB by providing affordable access to specialized bandages and dressings. Rebecca will take us through the scheme and provide us with some insights on what it means to be a clinician nurse supporting people with a rare disease.

Hi Rebecca, why don't we start with you telling us a little bit about yourself?

REBECCA:

My name is Rebecca. I'm the nurse specialist for the National EB Dressing Scheme. It's a federally funded role with the National EB Dressing Scheme and at the moment, I work with Independence Australia who are the administrators of the scheme. I've been in this role for the last two years. I've worked with families affected by EB for the last six years in New South Wales. I primarily work with children in that clinical role.

SAMSARA:

How long have you been a nurse? Is this something that you've been doing for a while?

REBECCA:

I've been a nurse for about 20 years now. I started off in pediatrics, and I moved into working with families with EB about six years ago now. I work at Sydney Children's Hospital as the Clinical Nurse Consultant for EB looking after families that have a child affected by EB all over New South Wales and the ACT, and I was introduced to Independence Australia and the National Dressing Scheme in March 2019.

SAMSARA:

Let's start with the big question. What is EB?

REBECCA:

So EB is a rare genetic skin disease. People who have EB are missing one of the genes in one of the layers of the skin. There's many genes involved. They're holding all the different layers of the skin together and they're important just to bring elasticity and allowing any type of frictional movement on the outside of your skin. So when you're missing one of the genes in the layers of the skin, depending on which gene it is you can develop blisters and wounds from any type of friction on the outside of your body. And that might be from wearing shoes or having elastic in your waist, or from being held too tightly, or picked up under the arms. All of those mindless activities that we do every single day in life can actually cause wounds for people who live with EB.

SAMSARA:

What's the occurrence in Australia? Do we know how many people live with a variant of EB here?

REBECCA

The occurrence in Australia is pretty similar to the global statistics, which is approximately one in 20,000 so it's classified as a rare disease. We do think that there are a lot more people living in the community who have the minor subtypes such as EB simplex, or dominant dystrophic EB. We will get a new referral for an infant or a child with one of those conditions and through that new referral, we often come into contact with say, 10 or 15 extended family members who have grown up with it without knowing that they had EB. The way that you test for EB is looking at the layers of the skin and looking for the absence of one of those genes. It's not a defect in a gene that's present, it's actually the people will be missing one of the genes such as Keratin five, or Keratin 14, or Collagen 7. There's quite a few genes that can be missing and depending which gene is missing, that will depend on the subtype and the severity of the disease process for that individual.

SAMSARA

So you've said it's not uncommon to discover that there are a number of people in one family that are living with a variant of EB, but are undiagnosed?

REBECCA

Yes, so with each new diagnosis, if it's the dominant form of the disease, then one parent will have the disease as well, and often will be showing symptoms of that disease and they may have had a diagnosis, they may have never formally been diagnosed, they just may have grown up knowing that they have sensitive skin. With the recessive forms of the disease, you'll find that both parents are carriers of this genetic change but they don't show any signs of the disease so often, they've never heard of it and it's only through testing both parents and both of them being carriers that they pass on the recessive form of the disease.

SAMSARA:

You did mention that it's a rare disease so does that suggest that there's not a lot of understanding in the broader community about what EB is?

REBECCA:

Yeah. That can be a really difficult process for a newly diagnosed baby, for the parents, to try and go through that process of looking after their child and learning about the disease. As a rare disease, it can be really tricky trying to find healthcare professionals who understand what's going on. Or they may have heard of EB, but they may not have any experience with EB. So it can be really quite a difficult experience in that first year of life for new families with a new diagnosis, coming to terms with trying to upskill themselves clinically and educationally, and then very quickly realizing that they possibly know more than the health professionals that they're going to see. That's actually where my role comes into play often - so having another health professional who can speak on the behalf of the parents and liaise with other health care professionals, and help to upskill the health professionals to take that burden off the new families who need to focus on the emotional connection with the child and also their own emotional journey of coming to terms with a diagnosis. They're not usually in a place where they're able to stand up and fiercely advocate for their child or even provide education for health professionals.

SAMSARA:

Let's talk a little bit more about the National EB Dressing Scheme. Can you give me a little bit of a rundown of how the scheme works?

REBECCA:

Yeah, so it's called the NEBD scheme. It's quite a long name. So that stands for National Epidermolysis Bullosa Dressing Scheme, and it's funded by the federal government. We can help to reduce the financial burden of accessing wound care. But we can also help to reduce the burden of trying to find EB appropriate wound care. So if you stick the wrong dressing onto a person who has EB, if it’s an inappropriate dressing, if it has too much stickiness, or glue in the borders of the dressing, it can actually pull their skin off. All of the products on the national dressing scheme are appropriate for people living with EB. They usually have a very low tackiness in them. A lot of them are silicon based. They are non-adhesive dressings. Families can apply through an EB specialist. You can apply to the national EB Dressing Scheme to become a dressing scheme provider. So we have providers located all around Australia who are experienced in EB care. The family will need to find a provider and as a child, it's usually through the main hospital of the state, the multidisciplinary team, where those providers are located and the provider will submit an application on behalf of the family and will write a list of dressing that the family will need, and that family will be offered a monthly delivery.

SAMSARA:

Can you tell us a bit more about the background of the scheme?

REBECCA:

The scheme was set up by a group of concerned healthcare professionals who could see that not having access to appropriate dressings was causing people's wounds to get worse and really impacting on the quality of life of people living with EB. It's actually quite a unique scheme. I was fortunate enough to participate in a study tour in 2016 and review a few different countries around the world and how people living with EB had access to wound care. And there's not many other EB specific schemes that exist anywhere else in the world so the people in Australia living with EB are quite fortunate to have such a program as this. Some countries around the world do include the dressings needed in their national health care system so they can access them for free of charge from the GP.

SAMSARA:

Can you tell us a bit more about how the Scheme provides support to patients and their families?

REBECCA:

The link it provides to healthcare professionals, the interaction that it provides between federal funding body, health care professionals and the families is quite unique. I think it's really supportive of the families and also to support people living with EB with other areas of their life, such as their school or their daycare, and work with other healthcare professionals, like their GPs, or the hospital of birth, pediatricians, things like that. It helps to reduce that burden of disease in a really significant way, so these families don't need to explain themselves and explain why their child needs certain things to everybody that they meet every day, which you can imagine would be a massive burden. If you can walk into a school or walk into a GP, and know that they've been informed about EB and educated by an EB professional, it will give you a lot of confidence in going there and it would also probably encourage you to go to your GP more regularly, if you knew that they were going to be able to support you, rather than you walk in with a sick child and have to take on the educator's role. Where actually you need to go to the doctor to get support, a lot of these families find that when they go to a new doctor, they need to educate the doctor first before the doctor can then support them.

SAMSARA:

Would you tell us a little bit more about the importance of wound care for EB patients?

REBECCA:

Wound care is a very large part of managing EB, and keeping wounds covered and promoting an ideal healing environment for wounds is really, really important. Ideally, appropriate wound care sets up an environment where that wound will continuously get better, where the bacteria is managed and the wound progresses and healthy skin grows. With a lot of wound care products, if we were to put products that are used on a majority of wounds in a plastics clinic or a burns clinic in a hospital, while they're in place it may actually be causing more harm than good for somebody living

with EB. So with the genetic change in the skin, if you stick something to the skin that's too adhesive - and we talk about friction in the skin and that causing blistering and wounds - simply removing a wound dressing from skin may actually pull all the healthy skin from around that wound. You may actually pull that off when you remove the dressing. So it's really a major part of preventative care with EB - making sure that anything you apply to the skin can be gently removed without causing new trauma. Another specific area of EB, depending on the subtype is that sometimes dressings can move a little bit on the skin and that can be quite tricky on bony prominence, or areas that we use a lot in life, such as your palms or elbows. By moving around a lot and having normal dressings in place, that friction underneath the dressing may actually be causing new blisters so people living with EB usually have the dressings changed a lot more frequently than somebody who had all the genes present in their skin might do. You need to monitor the blisters and the wound underneath to make sure that it's not getting worse.

SAMSARA:

I presume having a list of appropriate dressings your doctor can select from makes it easier for people living with EB because they don't have to worry about whether these dressings will cause more harm to them?

REBECCA

Yes, correct. It also sets up a process of management. So it sets up a process of communication between the person living with EB and their healthcare provider. The families can't change their dressings or change their order themselves. The process and the way it's been set up also promotes regular interaction with the person living with EB and the healthcare provider, which is also really great to promote ongoing clinical care and ongoing wound evaluation. The dressings that are being required are being requested by a healthcare professional who has decided that that's the best form of management for the person living with EB.

SAMSARA:

I suppose, as they get older and have new experiences and are doing new things, that the conversation has to shift?

REBECCA:

Exactly, and as the parents are becoming more competent, so as they're learning about the disease, once they reach school age these families managing a child with EB are quite inspirational. They know a lot about their child, they know a lot about how to find that balance between encouraging their child to participate in everything they can in life, but also working out how to protect and prevent friction and protect their skin at the same time.

SAMSARA:

It's not just about supplying dressings, though, is it? it's also about those ancillary services that you mentioned, so perhaps we could talk a little bit about them?

REBECCA:

Yeah, sure. At the moment, we're doing a very large project on education support. When a child with EB is about to start preschool or start kindergarten, or even an older child is about to start High School - whenever somebody is moving into a new area for education - the scheme is able to provide a management plan and education for the centre or the school that they're going to attend. So the parents can have confidence that the staff looking after them will know how to manage them. The staff looking after them will be able to handle either ongoing prevention, but also an emergency if that happens at the centre or the school, and the family don't need to do this. So again, reducing that burden of them needing to upskill everybody that their family interacts with. For example, for somebody with quite severe EB we would be able to interact with their local allied health professionals. We would interact with the school. We would make sure that if they were going to hire a teacher's aide to work with this child all the time we'd be able to help upskill anybody who's working with the child, and also looking at the clinical side of things - making sure that they were comfortable handling the dressings and comfortable knowing how to reapply things and when to communicate with the family. So it can be quite involved with a child whose symptoms are quite severe and who needs a lot of physical support. But interestingly, there are also a lot of children who have very mild EB who are really well managed because their parents are doing an amazing job. To look at them from, you know, running around in the playground, you wouldn't notice anything different. You might notice a few little scars on their knees or they might have something that looks different to the other kids around one of their hands, but you wouldn't necessarily know that they have EB. But the way these parents are able to keep these children so well is through a lot of prevention, a lot of protective equipment, a lot of education – of sports coaches, of teachers - and so helping schools to recognize the mild forms and understand the importance of prevention, putting a healthcare professional’s face to that process is really important to help the schools to respect that this child has an incurable disease. They look well. They are well managed. But they are well managed because all of these prevention strategies work really well.

SAMSARA:

It seems like the scheme takes quite a holistic approach to EB. It's not just about providing financial relief for these very important dressings that you need day to day. It's also about how do we assist you to move through the world in a way that works for you, and where you can get the support that you need?

REBECCA:

It's been a big focus of mine since I've been in this role. I think that I mean, wound care is a really big part of life, but living life is really important. These children are going to get blisters and wounds anyway. We really need to support them to live the best life possible while they're having their wounds managed. Hopefully, we can help to reduce the burden on the person with EB, also on the extended family, and help so that they can fulfill their roles as a parent or, you know, a husband or a wife of somebody with EB. They can move back into the role that they're supposed to be,

which is to love and care and interact with their family member, rather than needing to step out into that clinical or educators role too often

SAMSARA:

It seems like such a significant thing that the scheme does, providing dressings at such a reduced rate?

REBECCA:

Making appropriate dressings available will mean that families are more likely to manage wounds more effectively. They're more likely to use the preventative methods that we recommend. If they're needing to pick and choose which wounds to manage and when to manage them because they have to buy the dressings themselves, often people wait. They'd be less likely to dress very small wounds that are almost healed, but still really should be covered because old wounds that have just healed with a person with EB who's actually missing the genes that fold those layers together, effectively, just a simple bump will open that wound up again. So they need to have that wound dressed for quite a lengthy time while that wound is healing, to give it time to heal really, really effectively before we can have them walk around with that area of skin exposed again. The preventative care means that the wounds heal better, there's less infections, there's less time off school, there's less time off work for the extended family, there'll be less hospitalizations for infection management, there'll be less engagement with the healthcare system. So the financial benefits for the health system as a whole on helping wound care to be managed effectively from the beginning, is quite significant.

SAMSARA:

So how do you think all these supports, as well as the dressings, helps improve the quality of life for someone with EB?

REBECCA:

I would say when infection rates are lower and there's less wounds, then your pain levels will be lower, your ability to participate in life will be higher, you will spend more time at school or more time at work or more likely to participate in social activities. So it would have quite a significant impact on the individual's mental health and also their ability to just participate, particularly for a young child. When they're in pain they're less likely to explore and move around and develop all those muscles that are needed to pass through their normal developmental milestones. So appropriate wound care, and preventative wound care make quite a significant difference in helping people to be able to get on with their lives.

SAMSARA:

I think we've probably covered all the questions that I had. I there anything that we haven't discussed before we go?

REBECCA:

I guess something that I would like people to know about managing a rare disease would be to recognise the emotional burden of the disease - that the inability to get the support that you need at the drop of a hat really impacts on the life of the individual and of the family. We're slowly getting there. Raising awareness of EB and raising awareness of the importance of the scheme is ongoing, and helping people to access the support that they need - it's really, really important in managing a rare disease. I think that the burden on the families is so significant.

SAMSARA:

Thank you so much Rebecca, for all of that wonderful knowledge and insight you have shared with us – not just about the practical financial aspects of the government funded National EB Dressing Scheme, but also providing such wonderful insights as to how the scheme helps people with EB and their families to sustain a significantly improved quality of life.

You have been listening to Inform. You'll find links to any resources mentioned in this episode, plus more, in the show notes at informonline.org.au

Our thanks to Rebecca Saad for being a part of this episode, and for our colleagues at the Department of Health who work with her on this Scheme.

Thank you for listening to Inform, a production of Independence Australia. Inform is hosted and produced by me, Samsara. Our managing editor is Alison Crowe.

This episode of Inform was recorded and produced on the lands of the Wurundjeri people of the Kulin nation. We pay our respects to elders past and present.

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