



## Opening Remarks to the Oireachtas Committee on Health Health services for persons with EB (epidermolysis bullosa)

Wednesday the 28th of February 2024

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### Introduction

Thank you, Chairman, and thank you to the committee for the invitation to speak on health services for people with Epidermolysis Bullosa, EB, on the eve of Rare Disease Day. I'm Sinéad Hickey Head of Advocacy, Research and Policy at Debra, and I am joined this morning by my colleagues Deirdre Callis Head of Family Support Services at Debra, and Dr Rosemarie Watson, Debra Board member and former Director of EB Services in Ireland.

### Background

Debra is a charity that provides support for people living with this extremely painful, rare and incurable, genetic skin condition called EB. People living with EB are missing proteins that bind the layers of their skin together, so any minor friction causes it to break and blister. In some cases, EB can also affect the internal organs causing internal injuries. An average of 300 people in Ireland live with the condition.

### Implications of EB

Different forms of EB vary in severity, but they all carry debilitating life-limiting symptoms and pain with implications for wellbeing, and quality of life. It is an extremely complex condition that can require intensive specialised care.

Wound management is of particular importance to prevent the development of

secondary infections, cancer and loss of limbs. In some cases, people need three to four hours of extremely distressing, painful bandage changes every other day.

A 39-year-old woman living with one of the worst forms of EB is best placed to describe the impact of bandage changes. Her condition is so severe it has led to limb loss as a result of EB-related skin cancer. She says:

*“I dreaded my mother coming into the room because the pain you endure, there are actually no words in the dictionary.”*

These necessary bandage changes must often be undertaken by relatives, and our report has shown that this has a significant negative impact on the family and carers quality of life too.

A mother who cares for her 20 year old daughter, who also lives with one of the worst forms of EB, describes the condition as having a profound effect on physical and mental health, not only of the person living with it but also of their family members.

## The Report

These experiences are in line with the findings of the [report](#) we launched with the University of Limerick at the end of 2023. This report reviews the evidence related to the impact EB has on the quality-of-life for people affected by this condition. It also examines access to services and supports across five countries, including Ireland. We identified a set of key recommendations based on the results of this research, some of which we will discuss today.

The research finds that people with EB in Ireland do have access to excellent support in the specialist clinics in Children’s Health Ireland at Crumlin and St. James’s Hospital and community health teams, and I wish to commend the work of those dedicated teams. However, many families continue to face

significant challenges, and unmet need for support while managing this difficult condition.

Dr Fiona Browne, Lead Dermatologist for the paediatric EB service in Ireland says:

*“As physicians, it is incredibly frustrating to have nothing to offer patients other than supportive care. While there are treatments on the horizon, they are still some years away. We therefore need to ensure that the supportive care that we do offer is of the best standards, equitable and accessed without obstacles.”*

### Home Nursing Care & Outreach Support

The report found that challenges remain in the unreliable provision of home nursing care for people with EB, including the lack of a contingency plan for when nursing staff are unavailable or move on. As a result, family members often have to step in and perform bandage changes, inflicting tremendous pain and damaging fragile relationships, often between mother and child.

To try and address this urgent need, Debra currently supplements nursing care at home for people in most need. However, we are seeking ring-fenced funding, and a commitment to recruit for these hours and establish a sustainable service to ensure that people with severe EB can access this essential service in line with their growing needs.

### Psychological Support

The report also highlights the significant impact of EB on the mental health and quality-of-life of those with EB and their families. This condition can lead to a loss of mobility, impact on daily tasks, and lead to social isolation, or loss of work opportunities. This can have a significant impact on mental health. In fact, our findings show that people with EB have far higher rates of anxiety and depression, compared to the general population.

As a key feature of this condition is living with ongoing pain and distressing wound management practices, we advocate for specialist support to help cope with this debilitating condition. A key recommendation based on the research was that extra funding should be provided to help appoint a clinical psychologist dedicated to supporting adults with EB.

The findings of this report clearly outline the burden of disease, emotional, and social costs of this debilitating condition, and inform our asks for ringfenced funding, commitment to recruit for a sustainable service and dedicated specialist support to meet the particular needs of those living with EB, and the people that care for them.

We hope that the findings of this report can inform the implementation of the recently published HSE National Rare Disease Office [Inherited Epidermolysis Bullosa \(EB\) Paediatric and Adult Care Pathway](#), a crucial document outlining the breadth of support needed by people with EB across acute and community settings as well as the HSE Rare Disease Plan currently being developed.

To further support this goal, we are currently developing a second project aimed solely at generating in depth evidence on access to EB services and supports in the Irish context to identify strengths, challenges, and gaps in provision. Chairman, would welcome the opportunity to return to the Joint Committee on Health to present our findings next year.

However, EB does not wait, it continues to wreak havoc on people and their families living with the condition in Ireland daily and we hope that these people can be granted the much-needed services I've mentioned while waiting for the HSE care pathway to be fully implemented. We are grateful for Minister Rabbitte's support which has led to the nomination of Debra as an organisation to receive some support in the current Health Service Plan that

will go some way to relieve the pressure to deliver care for children with EB. We still need significantly more support for specialist health services so that we don't leave anyone suffering with this rare and complex condition with unmet needs.

We would like *to thank the Health Committee for inviting us here today to talk about our report's findings, and its implications for the needs of people with EB and their families.* We welcome the opportunity to discuss this topic further and answer any questions you might have. Thank you.