Life with EB in Ireland:

An international review of quality-of-life and comparison of access to healthcare resources and government supports

Epidermolysis Bullosa (EB) is a genetic condition that affects the body's largest organ, the skin. People living with EB are missing the essential proteins that bind the skin's layers together, so any minor friction, movement or trauma causes it to break and blister.

This collaborative report between Debra and the Kemmy Business School at the University of Limerick reviews the impact of EB on quality-of-life and considers access to healthcare resources and government supports internationally, with a view to identifying areas where access to EB supports and services in Ireland can be improved in the future.

In order to achieve this, we conducted (a) an in-depth literature review of the existing international evidence relating to the quality-of-life and economic burden of EB, and (b) a comparative review of access to healthcare resources and government supports across Spain, Australia, Ireland, the UK, and Austria.

Key findings

- **Studies have shown that people with EB exhibit higher than average frequencies of psychiatric symptoms.** For example, depression has a prevalence of 63.6% among people with EB compared to 10.1% for the general population. Despite this, there is relatively poor access to mental health services for those affected by EB.
- **The literature also revealed that quality-of-life for people caring for someone with EB is significantly lower** than the control adult population. For example, in the UK the average EQ – 5D index for carers is 0.696, 21.4% lower than the average adult.
- Further analysis of the available literature gives **an estimated total economic burden for EB of €130,021 per person, per year**. More research into the economics of the condition is needed.
- Relative to other sampled countries, **people in Ireland have poorer access to dressing materials**. Other countries have national bandage schemes or support access to cheap bandaging materials for people living with all severities of EB.
- **Access to dental care is an issue across multiple countries, including Ireland**, and people with EB can be required to travel long distances to receive care in specialist treatment centres.
- Access to medication and medical equipment was good across the sampled countries, but **access to equipment can depend on the severity of the disease**.
- Similar to other countries, **challenges remain in the provision of home nursing care in Ireland**, such as the lack of a contingency plan when nursing staff are unavailable, and a lack of nurses trained in EB to replace those who move on. **Access to respite care is an issue that arose across all the sample countries**.
- Care is available for people with EB through specialist clinics. However, **it can be difficult for people to access additional services crucial to their wellbeing, such as physiotherapy and occupational therapy**, in the community.

EB in Ireland: key recommendations

The following recommendations are based on the findings of this research, alongside recommendations of the Rare Disease Taskforce[1] and those set out in Debra's 2024 pre-budget submission.

- EB is a serious, chronic, incurable condition and, in line with advice from the Rare Disease Taskforce (2020), as such it is recommended that **EB should be included in the Long-Term Illness scheme.**
- In line with countries such as Australia, **funding for an EB-specific bandaging and dressing material scheme should be provided**. Under such a scheme, a monthly supply of approved dressings would be delivered directly to a person's home. An initial estimate based upon the Australian National Epidermolysis Bullosa Dressing Scheme (NEBDS) suggests that €400,000 in funding per annum would be required to run a similar scheme in Ireland.
- It is recommended that extra funding should be provided to help **appoint a clinical psychologist dedicated to supporting adults living with EB**. This funding has been estimated in the Debra pre-budget submission for 2024 at €80,000 per annum.
- The provision of **regular, reliable home nursing care for those living with more severe EB should be significantly enhanced**. The Debra pre-budget submission for 2024 calls for a ring-fenced fund of €580,000 per annum to ensure people with EB can access secure and flexible care packages that adapt as their needs change over time.
- Greater **research into the quality-of-life of people living with EB in Ireland** should be undertaken.

1. Rare Disease Taskforce, 2020. *An Easy Guide to Rare Diseases in Ireland and Consensus for Action*, Dublin: HRCI, IPPOSI, Rare Disease Ireland.

What is Epidermolysis Bullosa (EB)?

EB is a group of painful genetic blistering conditions that affects the body's largest organ, the skin. It's debilitating. Excruciating. Relentless. A disease with no known cure.

There are four main forms of EB that vary in severity, but all carry their own life-limiting symptoms, life-long pain and, in many cases, disability. EB can affect both the external and internal lining of the skin. EB is a classified as a rare disease. An estimated 1 in 18,000 babies born in Ireland are affected by EB. On average, 300 people in Ireland have EB.

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debra.ie

Tel: 01 412 6924

Email: advocacy@debra.ie