

Darragh Flannery<sup>a\*</sup>, Claire Doyle<sup>a</sup>, Avril Kennan<sup>b</sup>, Sinead Hickey<sup>c</sup> and Fiona Aherne<sup>c</sup>

<sup>a</sup> Kemmy Business School, University of Limerick, Limerick, Ireland.

<sup>b</sup> Medical Research Charities Group, Dublin, Ireland.

<sup>c</sup> DEBRA Ireland, La Touche House, 1 Grove Road, Rathmines, Dublin 6, Ireland

## Introduction

The condition Epidermolysis Bullosa (EB) is a rare, genetic skin condition, where the skin of a patient is abnormally fragile. Due to a genetic abnormality, the layers of the skin do not adhere to each other adequately and thus even minimal physical contact can cause blistering and severe pain. It is estimated that the prevalence of EB in Ireland is approximately 1/17000. There are currently no effective treatments or cures for EB and management focuses on amelioration of the symptoms. Treatments are, however, in development and because the condition is rare, it will be necessary to compare the cost of new treatments with the cost of not treating, in order to make informed budget decisions. Given the rarity of EB, there has been minimal research conducted from a cost viewpoint.

## Aim

To estimate the cost of EB for young people in Ireland for the first time and compare with available international data.

## Methods

Similar to Angelis et al. (2016), we used a prevalence based approach using a sample of households with a child <18 diagnosed with EB who received outpatient care to estimate the resources used. The survey was anonymous and the participants were contacted by their patient organisation. As the information related to those under 18, the main care provider of the patient completed the survey in either a face to face manner or by post. Information on drug, wound care and various medical service utilisation as well as basic socioeconomic information was collected. Unit costs were taken from the HSE guide on service prices and Primary Care Reimbursement Service list of reimbursable items.

## Participant profile

Six participants

Five aged  
2-10 years

One aged  
9 months

Three mild - moderate  
severity

Three  
Severe – very severe



Clinical photograph of patient with RDEB-severe generalised

Courtesy of Dermatology Dept, Our Lady's Children's Hospital Crumlin.

## Results

### Summary of Average Costs across 12 month period for those aged 2-10 by severity

|                      | Severe-Very Severe | Mild-Moderate |
|----------------------|--------------------|---------------|
| Wound and drugs cost | €59,991            | €11,859       |
| Hospital costs       | €47,894            | €0            |
| Clinic costs         | €1,500             | €300          |
| Primary care cost    | €2,583             | €325          |
| Total Medical costs  | €111,888           | €12,484       |

### Summary of Average Costs for first 12 months

|                      | Severe   |
|----------------------|----------|
| Wound and drugs cost | €8,303   |
| Hospital costs       | €104,560 |
| Clinic costs         | €450     |
| Primary care costs   | €3,200   |
| Total Medical costs  | €116,513 |

## Discussion

- The wound and drug costs for those with severe and very severe EB are significant.
- The hospital costs in the first 12 months of life with severe EB are also significant.
- There is a high degree of heterogeneity in these costs depending on severity.
- The cost estimates for the mild-moderate sample are very comparable to available international data, Angelis et al. (2016).
- Our research is limited by a relatively small sample size.

## Future Research

- In the process of gathering information on the informal costs related to households caring for someone with EB.
- These informal costs may add considerably to the overall social cost.
- There are also interesting policy questions around the lost productivity from the disease which are difficult to estimate.

## Relevant References:

Angelis, A., Kanavos, P., Lo'pez-Bastida, J., Linertova, R., Oliva-Moreno, J., Serrano-Aguilar, P., Posada-de-la-Paz, M., Taruscio, D., Schieppati, A., Iskrov, G., Brodsky, V., von der Schulenburg, JMG., Chevreul, K., Persson, U., Fattore, G. (2016) Social/economic costs and health-related quality of life in patients with epidermolysis bullosa in Europe, *Eur J Health Econ*, 7, 31-42.

Fine JD, Bruckner-Tuderman L, Eady RA, Bauer EA, Bauer JW, Has C, et al. Inherited epidermolysis bullosa: updated recommendations on diagnosis and classification. *J Am Acad Dermatol*. 2014;70:1103-26.