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Direct Costs of Epidermolysis Bullosa by Disease Severity

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The condition epidermolysis bullosa (EB) is a genetic, rare 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, sores and severe pain. EB prevalence is estimated at 1/18,000 live births (1, 2) EB has been separated into different subtypes, based on the ultrastructural level within which blisters develop in affected tissues (3). The 3 most prevalent sub-types are: EB simplex (EBS), junctional EB (JEB) and dystrophic EB (DEB). EB simplex is the most common and usually the least severe form; blistering is often localised and lesions heal without scarring. Junctional EB, recessive dystrophic EB and EB simplex generalised severe, on the other hand, tend to be less common and are often more severe for the patient.

There are currently no effective treatments or cures for EB and management focuses on amelioration of the symptoms. Treatments are, however, in development and are likely to be expensive 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. However, given the rarity of EB, there has been minimal research conducted from a cost viewpoint. Indeed, Angelis et al. (1) highlight the lack of research in this area. Tabolli et al. (4) provide survey evidence of high family burdens related to EB from a psychological distress viewpoint. Brun et al. (5) also use patient survey data and show that those with EB to have frequent and severe pain with neuropathic characteristics, with this pain negatively affecting quality of life. The only study identified to date that considers cost issues related to EB is that of Angelis et al. (1). This study utilised a bottom up patient survey and showed mean costs ranging from €46,000 per annum in Germany to €9,500 in Sweden. It was also notable that informal care costs formed a large proportion of the overall total for each country and that costs for paediatric patients were considerably higher than for adults. We conducted a similar study for the case of Ireland using a sample of paediatric patients. We differentiate our study from that of Angelis et al. (1) by providing more in depth analysis specific to wound care costs and costs across a subjective measure of the severity of the condition.

MATERIALS AND METHODS

Similar to Angelis et al. (1), we used a prevalence based approach using a sample of households with someone with a child <18 diagnosed with EB who received outpatient care to estimate the resources used and thus the social costs of the disease. The study protocol was submitted to the Kemmy Business School, University of Limerick (UL) Research Ethics Committee and received an exemption (respondents were recruited with the assistance of DEBRA Ireland). All patients and caregivers were informed about the study objective, data confidentiality and were asked to indicate their consent to participate. 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 on a total of 6 patients with respect to the previous month; 5 children aged between 2 and 18 and one infant aged 9 months. Cost information for the hospital stays (€813 per night based on semi private room) and day hospital clinic visits (€407) were taken from the Irish Health Service Executive's (HSE) guide on service prices (available at https://www.hse.ie/eng/about/who/acute-hospitals-division/patient-care/hospital-charges/). Primary care usage stemmed mainly from visit to general practitioners but also included physiotherapy, occupational therapy and public health nurse visits. To obtain separate cost information for each of these separately proved extremely difficult, therefore the mean GP visit charge in Ireland of €52.50 (6) for each primary care event was used as a proxy. The HSE's Primary Care Reimbursement Service list of reimbursable items (available at https://www.sspcrs.ie/druglist/pub) provided the unit cost information for wound and drug utilised (e.g. the unit price of a common dressage used by our sample, Mepilex Transfer 20x50cm was €37.20 as per the HSE guidelines).

Table I. Summary of mean costs across 12 month period for those aged 2–18 years by severity

Age, years	2.5	7	10	8	9
EB type	RDEB	RDEB	RDEB	EB simplex	RDEB
Severity	Very severe	Very severe	Very severe	Mild	Moderate
Wound and drugs cost (€)	27,252	89,780	62,699	17,732	5,986
Overnight hospital costs (€)	39,837	56,910	28,455	0	0
Day clinic costs (€)	2,442	1,628	20,350	2,442	814
Other primary care costs (€)	1,890	1,102	5,145	105	577
Total medical costs per annum (€)	71,421	149,421	116,649	20,279	7,377

EB: epidermolysis bullosa; RDEB: recessive dystrophic EB

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RESULTS

The sample is presented on a per patient basis with an infant as a stand-alone study and the older children summarised separately. **Table I** focuses on the latter and presents the costs across a 12-month period for those aged 2.5–10 by severity of illness and EB type. The direct costs estimated for our sample range from €149,421 to €7,377, the former representative of a patient with a very severe condition with the latter reflecting a patient with a milder form. The 3 other very severe patients also exhibit high costs at €71,421 and €116,649, respectively. It is notable that wound and drug costs are considerably higher for those with more severe conditions with the high utilisation of wound dressings such as Mepilex Transfer and Urgotul a significant driver of these high costs. **Table II** presents a summary of the mean costs for the first 12 months of life based upon our case study of one infant with severe EB. This highlights the significant contribution that hospital admittance has in costs at the early stage of life for those with severe EB rather than wound care costs, contributing around 90% of the overall cost of €118,665.

DISCUSSION

This short report contributes to the relatively sparse literature on the economic cost of the rare skin disease EB and suggest significant variation in these costs for young people dependent on severity. It also highlights the potential high hospital costs involved in the first year of life associated with the disease. This study has the obvious limitation of a small sample size, however, the estimates helps to demonstrate the high degree of heterogeneity in social costs from EB depending on the severity of the disease and thus contribute to knowledge in the area. The cost estimates for the mild-moderate sample are very comparable to those estimated in Angelis et al. (1) while the costs for the more severe cases in our sample are considerable higher. However, the Barthel index scores (mean of 85.2) from the Angelis paper indicate that their sample was mainly drawn from more mild to moderate cases and thus help explain the differences seen here. Future research in the area may focus upon more severe cases of EB, particularly the informal hours dedicated.

Table II. Summary of mean costs for first 12 months of life

Severe EB	Euro
Wound and drugs cost	8,303
Overnight Hospital costs	104,560
Day Clinic costs	2,442
Other Primary care costs	3,360
Total Medical costs per Annum	118,665

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