

Salamon, Gudrun; Strobl, Sophie; Dufresne, H el ene; Bodemer, Christine; Diem, Anja

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Psychometric Properties and Factor Structure of the Revised Epidermolysis Bullosa Burden of Disease (EB-BoD-R) Family Score

Gudrun Salamon¹ · Sophie Strobl^{1,2} · H el ene Dufresne³ · Christine Bodemer³ · Anja Diem⁴

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Abstract

Epidermolysis bullosa (EB) is a rare genetic disease defined by high fragility of the skin. The impact of this chronic disease extends beyond the physical and affects the psychosocial well-being and quality of life of not only the affected individuals but also their families. The Epidermolysis Bullosa Burden of Disease (EB-BoD) questionnaire aims to assess the impact of the disease on the family of those affected. Whereas the questionnaire has been partially validated in French, there is no validated English translation. Additionally, the original version is limited to assessing the quality of life of parents only. The objective of this study was to revise the EB-BoD to ensure its applicability to all relatives of individuals with EB, and to translate, culturally adapt and validate it in English. The EB-BoD revised was developed in three stages: (1) translation and cross-cultural adaptation, (2) content revision and thematic adjustment and (3) sample-based testing with 64 relatives of patients with all EB types to establish the psychometric properties of the English EB-BoD-R. The revised EB-BoD exhibits excellent reliability and internal consistency, with a Cronbach's α of 0.93. Exploratory factor analysis resulted in two distinct factors, *Circumstances* and *Emotions*. Convergent validity was demonstrated by strong correlations with the iscorEB-p and the SWLS (both $p < .001$). Divergent validity was found in relation to social support F-SozU ($\rho = -0.17$, $p = .228$). In conclusion, the English EB-BoD-R is an effective and reliable instrument for assessing the quality of life of relatives of EB patients.

Keywords Epidermolysis bullosa · Relative-reported outcome measures · Quality of life · EB-BoD · Family burden · Burden of disease · Test validity · Psychometric properties

Gudrun Salamon and Sophie Strobl are shared first author.

Extended author information available on the last page of the article

1 Introduction

Epidermolysis bullosa (EB) is a rare genetic skin disorder with an estimated prevalence of 1:90,000 worldwide. It is characterised by the formation of wounds and blisters on the skin and mucous membranes, even at minimal mechanical trauma (Bardhan et al., 2020). There are four main types of EB, depending on the skin layers affected: EB simplex (EBS), dystrophic EB (DEB), junctional EB (JEB) and Kindler EB (KEB), along with various subtypes (Has et al., 2020). Even individuals with the same underlying subtype present heterogeneous disease courses and experience individualised physical and psychosocial challenges (Feinstein et al., 2019; Fine, 2013; Korte et al., 2024; Togo et al., 2020). As EB is still incurable, the principal objective of treatment is to provide symptom reduction through wound care and pain relief, as well as to prevent blister formation (Bruckner et al., 2020). In most cases, family members play a central role, providing invaluable support in managing the condition's daily challenges (Chateau et al., 2023; de Azevedo Silva et al., 2020; Wu et al., 2020). They undertake extensive wound care routines and pain management, which are essential for their children's well-being (Adni et al., 2012; Bruckner et al., 2015; Mauritz et al., 2019). Their role extends beyond medical care, offering vital emotional support and advocating for their children's needs within the healthcare system (Dufresne et al., 2018; Gowran et al., 2015). At the same time, EB has a major impact on the lives of relatives, affecting all areas of life: individual emotions and personal growth, family life and leisure, as well as any kind of interaction with the environment, such as relationships or social integration (Chateau et al., 2023, 2024; De Stefano et al., 2020; Fine et al., 2005; Ireland et al., 2021; Mącik & Kowalska-Dąbrowska, 2015; van Scheppingen et al., 2008).

Health-related quality of life addresses the impact of a chronic condition on physical, psychological and social levels (Augustin et al., 2012; Karimi & Brazier, 2016). In the given context, it operationalises the various burdens of EB described above and their impact on relatives of people with EB (Sebaratnam et al., 2012). The variety of symptoms, the heterogeneous clinical picture and the psychosocial factors that are associated with EB, such as dealing with stigma, require EB-specific questionnaires not only for affected individuals themselves but also for their relatives (Choi et al., 2017; Frew et al., 2009; Mariani et al., 2024). The Epidermolysis Bullosa Burden of Disease (EB-BoD) questionnaire fills this gap.

1.1 The Epidermolysis Bullosa Burden of Disease (EB-BoD) Questionnaire

The questionnaire was developed by Dufresne, Hadj-Rabia, Taieb and Bodemer with the aim to assess the impact of EB on the family of those affected (Dufresne et al., 2015). The EB-BoD questionnaire comprises 20 items and was validated with 55 parents in France. The items are rated on a six-point Likert scale, ranging from "never" to "always" (-1 to 5). A higher sum score indicates greater burden and lower quality of life for relatives. The EB-BoD questionnaire demonstrated good internal consistency (Cronbach's $\alpha=0.9$) and convergent validity (SF-12, $r=-.61$). Known-groups validity was established by a significant differentiation between EB subtypes (Dufresne et al., 2015). Whereas the questionnaire has been partially validated in French, its English translation, which is widely used and cited (Alheggi et al., 2024; Sampogna et al., 2017), has not been validated with psychometric properties. Hence, the aim of this paper is to

establish a validated English version of the EB-BoD to assess and evaluate the burden that EB poses on families in English speaking countries.

2 Materials and Methods

The EB-BoD revised was developed in three stages: (1) translation and cross-cultural adaptation, (2) content revision and thematic adjustment and (3) sample-based psychometric testing. Ethical approval was granted by both the Medical and the Psychological Ethics Committees of the Sigmund Freud University. The reporting followed the cross-sectional study guidelines outlined by the Strengthening the Reporting of Observational Studies in Epidemiology (STROBE) initiative.

2.1 Translation of the EB-BoD to English

The translation process followed the COSMIN checklist (Mokkink et al., 2019) and combined recommendations for translation and cross-cultural adaptation of medical and psychological scales (Borsa et al., 2012; Epstein et al., 2015; Sousa & Rojjanasrirat, 2011).

In a first step, two English translations of the original questionnaire were produced by two qualified independent translators. A third non-validated English version was provided by the original authors (Dufresne et al., 2015). A multidisciplinary team with psychological, medical and linguistic expertise compared and merged the translations, resulting in a provisional consensus English version. Two independent translators conducted back translations of the preliminary English version. To ensure conceptual equivalence with the original, linguistic and conceptual ambiguities and discrepancies from the original French questionnaire were identified and resolved in collaboration with the original authors. Finally, feedback on clarity of the English items was sought from the EB patient organisation DEBRA Ireland and no further changes were considered necessary. A detailed documentation of all steps of the translation and linguistic validation process can be found in Table 1.

2.2 Content Revision and Thematic Adjustment

In consultation with the original authors, the initial EB-BoD was revised on a content level. In order to address not only parents of EB patients but all relatives being affected by EB, the original wording “your child” was changed to “your relative”. In addition, items that only referred to parents of underage EB patients were critically discussed in a multi-professional panel. Finally, the original wording “skin disease” was changed to “EB” in order to reflect the multisystemic dimension of EB and to exclude possible co-morbidities.

With regard to the interpretation of the questionnaire, the coding of the questionnaire was adjusted from the original -1 to 5 to 0 to 5 (0-0-1-2-3-4-5) for statistical and content-related reasons. In the original French version, those items for which the answer option ‘not applicable’ was selected were coded as -1 for not being considered in the sum score. In order to include as much collected data as possible, the EB-BoD-R uses the total average score instead of the total sum score. Even though the two options ‘not applicable’ and ‘never’ may differ semantically, they can be considered similar from a statistical point of view. Hence,

Table 1 COSMIN translation process

COSMIN translation process recommendations		The English EB-BoD translation process	COSMIN evaluation
1.	Describe both the original language in which the PROM was developed, the source language (if different from the original language) and the language in which the PROM will be translated.	The aim of this study was a translated and validated English version of the original French EB-BoD.	very good
2.	Ensure that the items will be translated forward and backward.	Multiple forward and multiple backward translations: Three English forward translations were obtained (two conducted for this study, one already existing non-validated translation). Two backward translations were conducted.	very good
3.	Ensure that both forward translators have a mother tongue in the target language in which the PROM will be translated.	The translators of our forward translations were English native speakers.	very good
4.	Ensure that one of the forward translators has expertise in the diseases involved, and in the construct measured by the PROM; the other forward translators is naïve on the construct measured by the PROM	One of our translators was an EB expert. The other was naïve.	very good
5.	Ensure that both backward translators have a mother tongue in the original or source language	One of the backward translators was a native French speaker and the other was bilingual (French, English).	very good
6.	Ensure that both backward translators are naïve in the disease involved and the construct to be measured	Both backward translators had never seen the original test and were naïve concerning EB.	very good
7.	Ensure that the translators will work independently from each other	All translators were strangers to each other, had no contact and were working independently.	very good
8.	Provide a clear description on how differences between the original and translated versions will be resolved	To ensure content validity and conceptual equivalence with the original, linguistic and conceptual discrepancies in the backward translations of the original French questionnaire were identified and presented to the original authors. Some minor ambiguities were corrected in co-operation with the original authors. Most of the differences were only in wording, not in content, and did not require any adjustments.	very good
9.	Ensure that the translation will be reviewed by a committee (including the original developers of the PROM)	In a group discussion with a multidisciplinary research team with psychological, medical and linguistic experts, a consensus was reached between the three forward versions. Additionally, feedback was obtained from the EB patient organisation DEBRA Ireland. The final version of the English questionnaire was approved by the original authors.	very good
10.	Write a feedback report of the translation process	The whole process was carefully documented, with every step and decision taken. All decisions involving differences in wording or content and their rationale were described and reported in detail.	very good

Table 1 (continued)

COSMIN translation process recommendations	The English EB-BoD translation process	COSMIN evaluation
11. Perform a pilot study (e.g. cognitive interview study) to check (1) the relevance of each item for the patients' experience with the condition, AND (2) the comprehensiveness of the PROM, AND (3) the comprehensibility of the PROM instructions, items, response options, and recall period. Widely recognized or well justified method for qualitative research will be used	We collected direct feedback from the patient organisation DEBRA Ireland on the relevance, clarity and comprehensibility of the final English EB-BoD. Additionally, quantitative survey methods (e.g. floor and ceiling effects) were used to assess the relevance.	adequate
12. Perform the pilot study in a patient population representing the target population	Our study is based on data collected from $n=64$ relatives of EB patients across all EB types. To ensure cross-cultural consistency, we included participants from 19 countries with a high level of English.	very good

in the revised version of the EB-BoD both responses 'not applicable' and 'never' are coded as 0. The final English EB-BoD-R can be found in the appendix (supplementary file S1).

2.3 Recruitment and Design

In a cross-sectional online study, participants were recruited over the course of six months in collaboration with DEBRA International, DEBRA Ireland and local stakeholders (e.g., GPs). Inclusion criteria were a minimum age of 14 years, fluency in English and having a close relative diagnosed with EB. Informed consent was obtained from all participants. The online survey consisted of questions on general demographics and data relating to EB as well as the English EB-BoD-R. In addition, the health status of the EB patients was assessed via the Instrument for Scoring Clinical Outcomes of Research for Epidermolysis Bullosa patient score (iscorEB-p) (Bruckner et al., 2018; Salamon et al., 2024b; Schwieger-Briel et al., 2015), satisfaction via the Satisfaction with Life Scale (SWLS) (Diener et al., 1985; Hinz et al., 2018; Pavot & Diener, 2008), and social support via the Social Support Scale (F-SozU) (Kliem et al., 2015; Lin et al., 2019).

2.4 Data Analysis and Psychometric Properties

The sample was described using frequencies and standard deviations, means or medians and percentiles (interquartile range IQR: 25th and 75th). Data analysis of the psychometric properties was performed at both the individual item and a construct level. In regard to ceiling and floor effects, values <0.8 were considered as acceptable. For item-total correlations, values >0.3 indicate a good consistency of each item (Irwing et al., 2018).

An exploratory factor analysis (principal component analysis, varimax rotation) was conducted to evaluate the construct validity both from statistical and content perspectives. To confirm convergent validity, Spearman's rank correlations were calculated with established tests based on a similar construct, such as medical state via iscorEB-p and satisfaction with life via SWLS. A correlation coefficient of ≥ 0.3 was considered good and ≥ 0.5

excellent (Cohen, 1988). Cronbach's α was used to assess the construct validity and internal consistency of the EB-BoD-R and its subscores. An $\alpha \geq 0.7$ was considered acceptable, ≥ 0.8 good, and ≥ 0.9 was considered excellent (Nunnally, 1994). Finally, the sample was analysed in relation to the final EB-BoD-R and its factors. An inspection and analysis of the data distribution was performed using histograms, Kolmogorov-Smirnov test, and skewness statistics. For group comparisons, Mann-Whitney U-test and Kruskal-Wallis tests were performed. The statistical analyses were performed using IBM SPSS Statistics version 27, assuming a significance level of 5%.

3 Results

3.1 Study Population

A total of $n=64$ relatives of EB patients, consisting of 54 parents, six partners, two grandparents and two aunts, participated in the study. Regarding the gender distribution of the total sample, 84.4% were female. Most of the study participants were from Ireland (42.2%), with smaller percentages from Australia (14.1%), New Zealand (9.4%), the USA (6.2%), the Netherlands (4.8%), Belgium (3.2%), and one participant from each of 13 countries. Concerning the language level, 60.9% were native speakers; besides one, the rest understood English well to very well. The average age was 47.03 years ($SD=12.03$), with a range from 19 to 76 years. The mean age of the participants' relatives with EB was 15.95 ($SD=15.20$) years, ranging from 1 to 59 years. All four main types of EB are represented in the sample, with the majority reporting DEB (50.0%) or EBS (43.8%). The degree of severity (mild: 26.6%, moderate: 35.9%, severe: 37.5%) was well distributed. Regarding the visibility of EB, most relatives (60.9%) stated that their patient's EB was very visible. At the time of the survey, 31.3% patients had an acute challenging phase in regard to EB, four were in their palliative phase. On average, the relatives spent 11.54 h ($SD=11.3$) per week with EB-specific things, ranging from 0 to 47 h. An overview of the demographic data can be found in Table 2.

3.2 Psychometric Properties

Across all participants, only five EB-BoD items had not been answered, resulting in a missing data of <1%. At item level, all items were examined for floor and ceiling effects. The item 'My family does not come to see us because of my relative's EB' demonstrated a floor effect, leading to its exclusion from the revised version of the EB-BoD. In agreement with the original authors, the item 'I have great difficulty in finding childcare for my relative on account of his/her EB' was removed due to content and statistical parameters. The remaining 18 items indicated sufficient item variance and a good item-total correlation.

At construct level, an exploratory factor analysis was performed on the remaining 18 items to evaluate the structure of the EB-BoD revised. A significant Bartlett test of Sphericity ($\chi^2(153)=648.52, p<.001$) and a Kaiser-Meyer-Olkin Measure of 0.828 indicated that the sample was appropriate for factor analysis. An exploratory factor analysis resulted in a four-factor solution that accounted for 66.16% of the variance. However, given several double loadings and little differentiation in content between the factors, a two-factor model

Table 2 Sociodemographic description of the English study sample

Sample size		64 relatives of EB patients	
Average age		47.03 (SD=12.03; MD=46.50) Range from 19 to 76 years	
Average time per week spent with EB-specific things (in hours)		11.54 (SD=11.30; MD=10.0) Range from 0 to 47 hours	
		participants (n)	percent
Gender	Female	50	78.1 %
	Male	14	21.9 %
Nationality	Ireland	27	42.2 %
	Australia	9	14.1 %
	New Zealand	6	9.4 %
	USA	4	6.2 %
	Netherlands	3	4.7 %
	Belgium	2	3.1 %
	Other*	13	20.3 %
Language level	Native	39	60.9 %
	Very good	18	28.1 %
	Good	6	9.4 %
	Adequate	1	1.6 %
Relation to EB	Parent/Stepparent/Foster parent from a person with EB	54	84.4 %
	Partner from a person with EB	6	9.4 %
	Grandmother/Grandfather from a person with EB	2	3.1 %
	Aunt/Uncle from a person with EB	2	3.1 %
<i>Concerning their relative with EB:</i>			
Average age of the relative with EB		15.95 (SD=15.20; MD=12.00) Range from 1 to 59 years	
		participants (n)	percent
EB type	EBS	28	43.8 %
	DEB	32	50.0 %
	JEB	3	4.7 %
	KEB	1	1.6 %
EB severity	Mild	17	26.6 %
	Medium	23	35.9 %
	Severe	24	37.5 %
EB visibility	Very	39	60.9 %
	Somewhat	15	23.4 %
	Rather not to not at all	10	15.6 %
Last challenging phase in regard to EB	At the moment	20	31.3 %
	Less than 6 months ago	25	39.1 %
	Less than 1 year ago	5	7.8 %
	More than 1 year ago	13	20.3 %
	Could not estimate it	1	1.6 %
Palliative phase	Yes	4	6.3 %
	No	59	92.2 %
	Missing	1	1.6 %

*Other: one participant each from Aruba, Belarus, Colombia, France, Hong Kong, Italy, Kenya, Malaysia, Portugal, Romania, Spain, Sweden, and the United Kingdom

was chosen instead, yielding 53.24% of the variance. Factor 1 *Circumstances* (explained variance: 26.74%) addresses all external aspects that can be impacted by EB, such as work, living situation and future plans, whereas factor 2 *Emotions* (explained variance: 26.50%) encompasses the intrinsic effects of EB and in particular the emotions and thoughts evoked by EB, such as the feelings and thoughts that arise before and during treatments or feelings caused by the reactions of others. Both factors demonstrate good internal consistency and reliability. With Cronbach’s $\alpha=0.93$, the reliability and internal consistency of the entire EB-BoD revised is excellent.

In regard to convergent validity, our sample of the EB-BoD revised showed a statistically significant negative correlation with a high effect size with the SWLS ($\rho=-0.52, p<.001$), addressing satisfaction with life (see Fig. 1). Furthermore, the EB-BoD-R statistically correlated significantly with a high effect size with the EB patient’s medical state as assessed via iscorEB-p ($\rho=0.73, p<.001$). Divergent validity was shown with a statistically non-significant correlation in regard to social support, as assessed via the FSoz-U ($\rho=-0.17, p=.228$).

An overview of the psychometric properties of the EB-BoD revised can be found in Table 3.

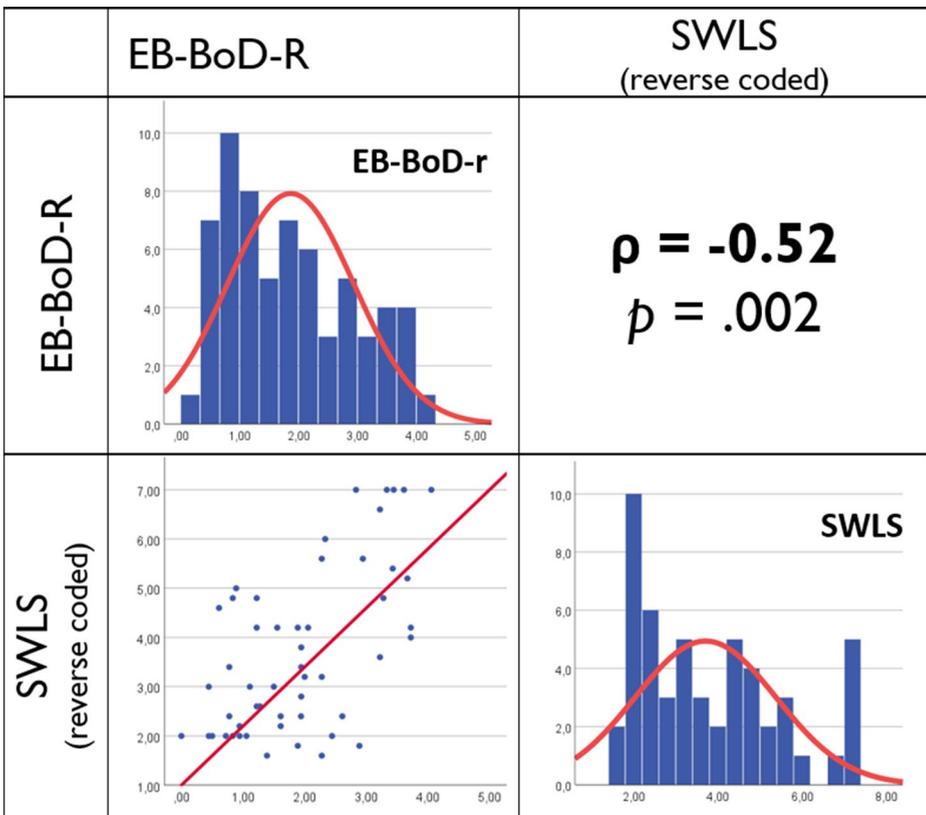


Fig. 1 Convergent validity of the English EB-BoD-R

Table 3 Psychometric properties of the English EB-BoD-R

Psychometric properties of the English EB-BoD-R	Item level		Construct level & factorial validity			
	Floor effect %	Ceiling effect %	Factor 1 Circumstances	Factor 2 Emotions	Item total correlations ¹	Cronbach's alpha if item deleted ¹¹
1. My relative's EB caused us to want to move.	56.3	4.7	0.43	0.21	0.41	0.88
2. My relative's EB led me to want to stop working.	42.2	14.1	0.54	0.37	0.58	0.87
3. I think of my relative's EB all day long.	15.6	14.1	0.40	0.62	0.65	0.85
4. I try to protect my relative because of his/her EB.	4.8	42.9	0.33	0.58	0.63	0.85
5. My relative's EB prevents us from going on vacation.	34.9	9.5	0.49	0.55	0.64	0.87
6. My relative requires more attention than other people due to his/her EB.	3.1	42.2	0.67	0.43	0.72	0.86
7. Our relative's EB forced us to question our future plans.	25.0	21.9	0.77	0.46	0.84	0.85
8. My relative's EB prevents me from visiting my family.	65.6	3.1	0.56	0.51	0.69	0.87
9. My relative's EB creates problems in our relationship.	50.0	9.4	0.56	0.36	0.49	0.86
10. The medical consultations for my relative's EB often leave me feeling frustrated.	23.4	12.5	0.42	0.44	0.53	0.86
11. People's reactions to my relative's EB are difficult to accept.	14.1	10.9	0.51	0.58	0.71	0.84
12. I have feelings of guilt about my relative's EB.	25.0	14.1	0.12	0.82	0.68	0.84
13. I have a hard time getting used to the odor produced by my relative's EB.	61.5	1.6	0.37	0.23	0.36	0.89
14. My relative has great difficulty in school/at work on account of his/her EB.	33.3	9.5	0.81	-0.21	0.44	0.88
15. I am afraid for my relative's future due to his/her EB.	15.6	25.0	0.65	0.39	0.68	0.87
16. Daily care is starting to weigh on me.	35.9	12.5	0.63	0.49	0.74	0.86
17. Each time I accompany my relative to the hospital, I do not feel well the day before.	57.1	4.8	0.27	0.76	0.64	0.85
18. Each time I accompany my relative to the hospital, I do not feel well the day after.	57.1	6.3	0.18	0.71	0.60	0.85
-- (excluded) My family does not come to see us because of my relative's EB.	81.3	1.6				
-- (excluded) If (relative is a child) I have great difficulty in finding child care for my relative on account of his/her EB	58.7	14.3				
			Item excluded due to floor effects		Explained variance of the questionnaire	
			Item excluded due to content and statistical parameters		53.24%	
Content validity	Convergent validity		Divergent validity		Internal consistency & reliability	
From original test development and obtained through the translation process.	iscorEB-p $p = 0.73, p < .001$ SWLS $p = -0.52, p < .001$	F-SozU $p = -0.17, p = .228$	Factor 1: $\alpha = 0.88$ good	Factor 2: $\alpha = 0.87$ good	entire questionnaire: $\alpha = 0.93$ excellent	

ρ =Spearman's rank correlation coefficient; α =Cronbach's alpha, iscorEB-p=Instrument for Scoring Clinical Outcome of Research for Epidermolysis Bullosa (Patient Score); SWLS=Satisfaction with Life Scale; F-SozU=Perceived Social Support Questionnaire

*¹a grey background indicates factor alignment; *²with the respective factor of the item

3.3 The EB-BoD-R

The EB-BoD-R was slightly skewed to the right, with a median of 1.9 on the total average score (IQR=0.94–2.79). In our sample, no differences were found between the quality of life of men and women, neither overall nor on the factors ($p > .05$ for all). Similarly, concerning the three EB subtypes EBS, DEB and JEB, no differences could be found ($p > .05$ for all). However, differences were found in regard to the degree of severity ($H(2) = 19.01, p < .001, \eta^2 = 0.28$; *Circumstances*: $H(2) = 22.99, p < .001, \eta^2 = 0.34$; *Emotions*: $H(2) = 10.99, p = .004, \eta^2 = 0.15$). The results of post-hoc U-tests showed that the burden experienced by relatives of patients with severe EB (MD=2.75) in our sample was higher than that of relatives of patients with moderate (MD=1.61; $U = 126.00, Z = -3.19, p < .001, r = -.40$) or mild (MD=0.94; $U = 62.0, Z = -3.76, p < .001, r = -.47$) severity, with differences between moderate and mild severity found as well ($U = 119.50, Z = -2.08, p = .037, r = -.26$). The perceived burden also differs between relatives of underage (MD=1.94) and adult EB patients (MD=0.94; $U = 110.50, Z = -2.12, p = .034, r = -.27$). The distribution of the entire EB-BoD-R questionnaire and an item-by-item overview of the distribution can be found in Table 4.

Table 4 Description of the study sample in relation to the EB-BoD-R scores

	EBS <i>n</i> =28	DEB <i>n</i> =32	JEB <i>n</i> =3	KEB <i>n</i> =1	Total <i>n</i> =64
EB-BoD-R: MD					
Average score	1.4	2.0	2.9	2.8* ¹	1.9
Sum score	23.0	35.0	53.0	51.0* ¹	31.0* ²
EB-BoD-R scales: MD					
Factor 1: <i>Circumstances</i> (10 items)	1.3	2.0	2.0	3.2* ¹	1.8
Factor 2: <i>Emotions</i> (8 items)	1.5	2.1	2.1	2.4* ¹	1.6
EB-BoD-R items: MD					
1. My relative's EB caused us to want to move.	0.0	0.0	0.0	5.0* ¹	0.0
2. My relative's EB led me to want to stop working.	0.0	2.0	5.0	2.0* ¹	2.0
3. I think of my relative's EB all day long.	2.0	2.0	4.0	5.0* ¹	2.0
4. I try to protect my relative because of his/her EB.	3.0	4.0	5.0	5.0* ¹	4.0
5. My relative's EB prevents us from going on vacation.	1.0	1.0	5.0	1.0* ¹	1.0
6. My relative requires more attention than other people due to his/her EB.	3.0	5.0	5.0	5.0* ¹	4.0
7. Our relative's EB forced us to question our future plans.	1.5	3.0	4.0	5.0* ¹	2.0
8. My relative's EB prevents me from visiting my family.	0.0	0.0	2.0	0.0* ¹	0.0
9. My relative's EB creates problems in our relationship.	0.0	1.0	1.0	0.0* ¹	0.5
10. The medical consultations for my relative's EB often leave me feeling frustrated.	2.0	2.0	4.0	5.0* ¹	2.0
11. People's reactions to my relative's EB are difficult to accept.	2.0	2.0	4.0	2.0* ¹	2.0
12. I have feelings of guilt about my relative's EB.	2.0	2.0	4.0	2.0* ¹	2.0
13. I have a hard time getting used to the odor produced by my relative's EB.	0.0	0.0	2.0	1.0* ¹	0.0
14. My relative has great difficulty in school/at work on account of his/her EB.	1.0	2.0	0.0	5.0* ¹	1.0
15. I am afraid for my relative's future due to his/her EB.	2.0	3.0	2.0	5.0* ¹	3.0
16. Daily care is starting to weigh on me.	0.5	2.0	2.0	3.0* ¹	1.0
17. Each time I accompany my relative to the hospital, I do not feel well the day before.	0.0	0.0	3.0	0.0* ¹	0.0
18. Each time I accompany my relative to the hospital, I do not feel well the day after.	0.0	0.0	1.0	0.0* ¹	0.0

EBS=Epidermolysis bullosa simplex; DEB=Dystrophic epidermolysis bullosa; JEB=Junctional epidermolysis bullosa; KEB=Kindler epidermolysis bullosa

*¹ only one underlying patient dataset; *²*n*=62

4 Discussion

The aim of this study was to revise the EB-BoD in order to include all relatives of EB patients and to validate and culturally adapt its English version. In addition to the involvement of a multidisciplinary team at every stage, recommendations for the translation of medical and psychological scales were followed jointly, ensuring a more holistic approach to assessing the burden on EB families throughout the whole process.

Since EB is a rare disease, the sample size is adequate and consistent with other test validations for EB (Dufresne et al., 2015; Schwieger-Briel et al., 2015). The inclusion of participants with a high level of English from a diverse range of countries ensures that the data reflect the linguistic and cultural diversity of the population. The age range of the

participants was well distributed, encompassing individuals from 19 to 76 years of age. In contrast to the original French version, which only included parents of EB patients aged between 0 and 18 years, the age range of EB patients in our study was considerably wider, at between 1 and 59 years. This ensures inclusion of a more diverse spectrum of life phases of the EB patients and of their relatives. The study encompasses all EB types, with DEB and EBS representing the most prevalent. Yet, the EB type does not seem to be directly related to the relatives' burden (Mauritz et al., 2022; Tabolli et al., 2009, 2010). Given the considerable inter- and intra-individual heterogeneity in symptoms and disease progression even within each EB type, the degree of severity can be considered a proxy for the amount of care required. According to our results, an evaluation of the impact of EB on the relatives' burden can be more accurately made by considering the degree of severity rather than the EB type. In accordance with findings of previous studies (Salamon et al., 2024a; Tabolli et al., 2009), the present study demonstrated that the family burden increased with increasing disease severity and seemed to be higher for underaged EB patients.

The revision of the EB-BoD was conducted in close collaboration with the original authors and through discussions with a multi-professional panel. By including psychologists, psychotherapists and medical experts with expertise in EB, the questionnaire was critically reviewed from a variety of perspectives and backgrounds. Amending the wording of the EB-BoD to include not only the parents' perspective, but also that of all relatives of EB patients, enables the whole family to be considered. Still, in order to achieve the closest possible similarity with the original French version, we have sought to minimise any deviations from the original. A purely statistically orientated analysis would probably have led to a greater reduction of the questionnaire. However, to minimize changes, a more conservative approach was chosen, which in turn covers a greater variety of burdens. This decision was supported empirically, as the final items of the EB-BoD-R showed neither floor nor ceiling effects, thus demonstrating their relevance. Similarly, our a priori decision to combine the options 'not applicable' and 'never' was confirmed by the data: 'not applicable' was selected in only 2.6% of cases. Concerning the test translation process, the English translation that was published along with the original test had not been validated (Dufresne et al., 2015). It was hence only considered as one out of three forward translations, and a full back-and-forth translation process was undertaken to ensure the content validity of the translation.

Concerning the psychometric properties: In the original publication, items are grouped in four dimensions based on content-related decisions only (Dufresne et al., 2015). For the EB-BoD-R, construct validity was established through an exploratory factor analysis. The selection of a two-factor model, comprising two distinct factors and explaining 53.24% of the variance, was made due to double loadings and the lack of clear content differentiation in the original four-factor model (Dufresne et al., 2015). By distinguishing between the factors *Circumstances* and *Emotions*, it is possible to compare external circumstances with internal psychological aspects and to investigate the impact of EB on relatives in a more nuanced way. Further support for the two-factor model can be found in comparable patient assessment, such as the QOLEB (Frew et al., 2009; Yuen et al., 2014; Cestari et al., 2016; Danescu et al., 2019; Salamon et al., 2025), which identified also a two-factor solution with similar underlying constructs. This distinction not only facilitates meaningful comparisons across studies but also provides practical value by identifying specific targets for therapeutic intervention. Furthermore, the two distinct factors found in our study demonstrate good internal consistency and reliability and can be differentiated by EB severity. All items demonstrated

satisfactory discriminative values, with the majority even exhibiting excellent discrimination. Similar to the French version (Cronbach's $\alpha=0.9$) (Dufresne et al., 2015), the entire questionnaire exhibits excellent internal consistency and reliability (Cronbach's $\alpha=0.93$). In contrast to the French questionnaire, no significant differences were found between EB types. This might be due to the uneven distribution of EB types in the sample, requiring further analysis in future research to confirm known-groups validity. In the meantime, severity may be considered as a proxy. In our sample, the EB-BoD-R showed excellent convergent validity, as evidenced by the SWLS and the iscorEB-p.

5 Limitations

While this study provides valuable insights into the psychometric properties of the EB-BoD-R, several limitations should be acknowledged. Given the rarity of EB, our sample size is relatively large, yet concerning the subject-to-variable ratio with 3.5:1, we must consider the possibility of sample bias. Participants were recruited via DEBRA International, DEBRA Ireland and local stakeholders, which might have resulted in a self-selection bias, yet ensured a diagnosis of EB within the family unit. A possible distortion may arise from the considerable percentage of Irish participants due to the different healthcare systems and cultural contexts and needs to be investigated in future studies. The proportion of female participants in our sample is higher than that of male participants, which reflects the higher proportion of female family members taking over the care work (Chogani et al., 2021a, b). Previous studies have produced conflicting results, with some finding a lower quality of life for male relatives (Salamon et al., 2024a) and others finding a lower quality of life for female relatives (Chateau et al., 2023; Chogani et al., 2021a, b). In the present study, however, no statistically significant effect of gender on the EB-BoD-R results was observed. Our sample included a large proportion of parents, but also 15% of other relatives (grandparents, siblings, partners). This is not that many overall, but based on feedback from EB clinicians, this roughly corresponds to the proportion of other active relatives in clinical practice. Still, the questionnaire may require further validation for use with other family members. Future research should aim to replicate and confirm these psychometric properties in independent samples and using alternative methods, such as test-retest reliability and confirmatory factor analysis, to strengthen the psychometric properties of the EB-BoD-R.

Despite these limitations, this study lays important groundwork for the psychometric properties of the EB-BoD-R. Although translated versions of the EB-BoD are already in use, its underlying English version had not previously been validated. This research therefore addresses a crucial gap by providing comprehensive validation to ensure the measure's reliability and validity.

6 Conclusion and Outlook

The revised English Epidermolysis Bullosa Burden of Disease (EB-BoD-R) questionnaire is a rigorously developed instrument that effectively measures the impact of EB on the entire family. Demonstrating strong psychometric properties in our sample, it offers a valid and reliable means of capturing the nuanced experiences and quality of life of not only parents

but also other family members affected by EB. While this paper represents a first step in validating the EB-BoD-R, further research is needed to establish its use longitudinally, ultimately contributing to improved outcomes for patients and their families alike.

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Data Availability The data underlying this article cannot be shared due to the rarity of EB, which limits anonymity even with pseudonymisation or exclusion of personal data. The data will be shared on reasonable request.

Reasons: *Despite the quantitative study design, data publication would violate ethical principles: given the rarity of the disease, the large number of subtypes and the strong interconnectedness within the patient community, already a combination of basic demographic data such as country, age and EB subtype could lead to recognition of individuals. Thus, even anonymisation would not allow sufficient de-identification of the data as required by ethical and data protection regulations.*

Declarations

Ethics Approval This study was performed in line with the principles of the Declaration of Helsinki. Ethical approval was granted by both the Medical and the Psychological Ethics Committees of the Sigmund Freud University Vienna (LBVCR72KAUBQ6188306).

Conflict of Interest None.

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Authors and Affiliations

Gudrun Salamon¹  · **Sophie Strobl**^{1,2}  · **Hélène Dufresne**³ · **Christine Bodemer**³  · **Anja Diem**⁴ 

✉ Gudrun Salamon
gudrun.salamon@sfu.ac.at

¹ HEALTH Lab | Competence Center for Medical and Health Psychology, Faculty of Psychology, Sigmund Freud University, Vienna, Austria

² Department of Dermatology, Medical Center, Faculty of Medicine, University of Freiburg, Freiburg, Germany

³ Department of Dermatology, Reference Center for Genodermatoses (MAGEC), Hôpital Necker-Enfants Malades, APHP, Paris, France

⁴ Department of Dermatology and Allergology, EB House Austria, University Hospital of the Paracelsus Medical University, Salzburg, Austria