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Living with dystrophic epidermolysis bullosa in Poland: first national study of health-related quality of life and symptom burden

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Abstract

Background: Dystrophic epidermolysis bullosa (DEB) is a rare genetic disorder associated with substantial and multidimensional disease burden. Although therapeutic options are advancing globally, patients in Central and Eastern Europe continue to face significant systemic barriers, including limited access to specialized care and innovative treatments. This study aimed to comprehensively assess health-related quality of life (HRQoL) and symptom burden in a national cohort of patients with DEB in Poland, with the objective of identifying key unmet needs.

Methods: A cross-sectional study was conducted in collaboration with the EB Polska Foundation. Data were collected through a structured, multi-modular online survey. Symptom severity, including pain and pruritus, was evaluated using an 11-point Numeric Rating Scale (NRS). HRQoL was assessed using validated dermatology-specific instruments (DLQI/CDLQI, Skindex-29) alongside a generic measure (SF-36). Statistical analysis was limited to descriptive methods.

Results: A total of 33 patients with DEB were included (mean age: 23.7 years; 75.8% female). The cohort exhibited a high symptom burden, with mean NRS scores of 5.7 for pain and 5.9 for pruritus. Dermatology-specific instruments indicated severe impairment in HRQoL (mean DLQI: 13.9; CDLQI: 14.3), with the highest burden observed in the symptoms domain of Skindex-29 (mean: 48.9). In contrast, the SF-36 revealed a marked discrepancy between physical and mental health domains: physical functioning remained relatively preserved (mean: 94.5), whereas mental health scores were markedly reduced (mean: 22.4), indicating substantial psychological morbidity.

Conclusions: Patients with DEB in Poland experience a considerable symptom burden and pronounced psychological distress, despite maintaining relatively preserved physical functioning, potentially reflecting adaptive coping mechanisms. This discordance highlights persistent disparities in care. Addressing these gaps requires systemic healthcare interventions, including the development of specialized reference centers for rare diseases, integration of routine psychological assessment, and improved access to disease-modifying therapies.

Background

Epidermolysis bullosa (EB) encompasses a heterogeneous group of rare, inherited genodermatoses characterized by extreme skin and mucosal fragility. Dystrophic EB (DEB), caused by mutations in the *COL7A1* gene encoding type VII collagen, represents one of the most severe subtypes [1, 2]. The clinical presentation extends far beyond recurrent blistering and chronic, non-healing wounds. Patients frequently suffer from severe systemic complications, including syndactyly (pseudosyndactyly), esophageal strictures, severe malnutrition, and a highly elevated risk of aggressive squamous cell carcinoma [2, 3].

While the therapeutic landscape for DEB is rapidly evolving with the recent introduction of gene therapies (such as beremagene geperpavec (B-VEC) or prademagene zamikeracel), novel topical agents like birch

triterpenes, and emerging stem cell therapies that show significant promise in accelerating wound healing, global access remains highly unequal [4, 5, 6, 7, 8]. In many regions, particularly in Central and Eastern Europe, these transformative treatments are not yet reimbursed or clinically available. Consequently, for the majority of patients in Poland, clinical management remains predominantly symptomatic and palliative, centering on extensive wound care, pain management, and nutritional support [9]. While the physical toll of DEB is undeniable, the chronic pain, relentless pruritus, and time-consuming nature of these daily care routines inflict a profound psychosocial and economic burden on both patients and their families [10, 11, 12]. Ultimately, this underscores a significant global disparity in care, where Health-Related Quality of Life (HRQoL) remains severely compromised due to a lack of access to disease-modifying therapies that are becoming standard in other parts of the world.

Despite growing international recognition of the HRQoL burden in rare genodermatoses, comprehensive data regarding the DEB population in Central and Eastern Europe remains scarce [12, 13]. In Poland, the management of DEB is compounded by significant systemic challenges. These include the absence of a dedicated national rare disease reference center and limited access to novel, disease-modifying treatment options, which delay definitive diagnosis and restrict future access to advanced targeted therapies. In this fragmented healthcare landscape, patient advocacy organizations, such as the EB Polska Foundation, play an indispensable role in bridging the gaps in systemic care and providing essential support.

Therefore, this study aimed to evaluate the multidimensional HRQoL and symptom burden in a national cohort of patients with DEB in Poland. To capture both the direct clinical impact and the broader psychosocial consequences, we employed a combination of dermatology-specific and generic assessment tools: Dermatology Life Quality Index (DLQI) and Children's Dermatology Life Quality Index (CDLQI), Skindex-29, Short Form Health Survey (SF-36), and Numeric Rating Scale (NRS). The DLQI (adults) and CDLQI (children) are 10-item questionnaires assessing the impact of skin disease on daily life over the previous week. Each item is scored from 0 to 3, yielding a total score between 0 and 30. Higher scores indicate a greater negative impact on HRQoL [14, 15]. The Skindex-29 is a validated skin-specific instrument containing 29 items across three domains: symptoms (7 items), emotions (10 items), and functioning (12 items). An additional non-scored question assesses treatment side effects. Responses on a 5-point scale are linearly transformed into a score from 0 to 100, where 0 represents no impairment and 100 represents maximum impairment. The recall period for this instrument is the previous 4 weeks [16, 17, 18]. The SF-36 is a generic 36-item questionnaire evaluating overall health status across eight domains: physical functioning, role limitations due to physical health, role limitations due to emotional problems, energy/vitality, emotional well-being, social functioning, pain, and general health perceptions. Scores for each domain range from 0 to 100, with higher scores reflecting better health status and functioning [19]. Finally, the NRS is an 11-point scale utilized to quantify the subjective intensity of specific symptoms, namely pain and pruritus, with scores ranging from 0 (no symptoms) to 10 (worst imaginable symptoms) [20, 21]. By utilizing this comprehensive set of instruments, we sought to provide a nuanced analysis of the patient experience. Ultimately, this research aims to identify critical

unmet needs, providing empirical data to inform and advocate for targeted healthcare policy improvements for the rare disease community.

Methods

Study Design and Ethical Considerations

A cross-sectional, national cohort study was conducted between June 2025 and October 2025 to evaluate the multidimensional health-related quality of life (HRQoL) and symptom burden in patients diagnosed with dystrophic epidermolysis bullosa (DEB) in Poland. The study was conducted in accordance with the principles of the Declaration of Helsinki. The protocol was formally reviewed and approved by the Bioethics Committee of Medical University of Lodz (Resolution No. RNN/46/25/KE).

Participant Recruitment and Advocacy Collaboration

Given the rare nature of the disease and the absence of a centralized national clinical registry in Poland, patient recruitment was conducted in close collaboration with the EB Polska Foundation, one of the leading national patient advocacy groups.

Inclusion criteria for the study were: (1) a clinically or genetically confirmed diagnosis of DEB, and (2) active membership or registration with the foundation. Exclusion criteria included incomplete survey responses that precluded meaningful data analysis.

The survey was distributed electronically via email to all registered members with a confirmed clinical diagnosis of DEB. Participation was strictly voluntary. Prior to participation, informed consent was obtained from all adult patients. For pediatric patients unable to complete the forms independently, parents or legal guardians acted as proxy respondents and provided parental informed consent.

Outcome Measures and Survey Instruments

To comprehensively capture both the disease-specific symptoms and the broader psychosocial impact of this multisystemic condition, data were collected using a secure online platform with a multi-modular questionnaire:

- Sociodemographic and Clinical Profile: Data regarding age, sex, extent of disease (anatomical locations involved), specific complications (e.g. syndactyly, gastrointestinal involvement), and previous treatments.
- Symptom Severity: The intensity of disease-specific hallmarks, pain and pruritus, experienced over the preceding week was quantified using an 11-point Numeric Rating Scale (NRS) (0 = no symptoms, 10 = worst imaginable symptoms).

- Self-reported health: Patients used a 10-point scale to evaluate general health, with higher scores indicating worse health perception.
- HRQoL Assessment: A combination of generic and dermatology-specific validated instruments was utilized to ensure a holistic evaluation:
    - Dermatology Life Quality Index (DLQI) for adults and Children's Dermatology Life Quality Index (CDLQI) for pediatric patients, focusing on the direct impact of skin fragility.
    - Skindex-29, to assess the emotional, symptomatic, and functional burden of skin lesions.
    - Short Form Health Survey (SF-36), a generic measure used to evaluate overall physical and mental health domains that might be overlooked by skin-specific tools.

Statistical analysis

Statistical analysis of the collected data was performed using Statistica software (version 13.3).

Results

Sociodemographic Profile and Clinical Characteristics

A total of 33 patients with a confirmed diagnosis of dystrophic epidermolysis bullosa (DEB) were included in the study. Detailed demographic and clinical characteristics of the cohort are summarized in Table 1. The cohort exhibited a strong female predominance (75.8%) and a mean age of 23.7 years, with nearly half of the respondents being pediatric patients. Genetic confirmation of the diagnosis was reported by the majority of the cohort (72.7%). Regarding disease management, all participants utilized non-adhesive dressings, and the vast majority relied on wound-healing adjuvants, whereas systemic therapy was extremely rare.

The clinical burden of DEB in the studied cohort was considerable. Patients reported an average involvement of nearly seven anatomical locations, most frequently affecting the hands and feet. As shown in Table 1, the clinical profile was characterized by a high prevalence of multisystemic manifestations. Nail dystrophy and dysphagia were the most common complications, followed by syndactyly and malnutrition. Beyond dermatological and gastrointestinal symptoms, systemic comorbidities such as anemia and anxiety disorders were also present.

Self-Reported Health and Symptom Burden

On a 10-point scale evaluating self-reported general health, the cohort demonstrated an overall moderate perception, with a mean score of 5.74 (SD 1.76). However, the specific symptom intensity was substantial. The distribution of self-reported pain and pruritus severity is illustrated in Figure 1. The mean pain intensity on the Numeric Rating Scale (NRS) was 5.7, with a significant proportion of patients rating

their pain between 5 and 8 points (Figure 1, Panel A). Pruritus intensity was marginally higher, presenting a mean score of 5.9, indicating an itch burden that generally ranged from moderate to severe (Figure 1, Panel B).

Dermatology-Specific Quality of Life

The comprehensive assessment of HRQoL, utilizing both dermatology-specific and generic instruments, is summarized in Table 2. Skin-specific tools consistently revealed profound impairment across all age groups. In both the adult (DLQI) and pediatric (CDLQI) cohorts, the disease exerted a "very large" negative effect on daily life. Furthermore, the Skindex-29 highlighted that the physical symptoms of the skin lesions constitute the most severe burden for the patients, closely followed by emotional and social distress.

Conversely, the generic SF-36 health survey revealed a striking divergence in the cohort's general health profile (Table 2). While self-reported physical functioning was remarkably preserved—indicating a retained capacity for basic daily activities—mental health, vitality, and general health perception were severely diminished. This stark contrast underscores the profound, often hidden psychological toll of DEB, even in patients who maintain a relatively high level of physical independence.

Generic Health-Related Quality of Life

Results from the SF-36 generic health survey (higher scores indicate better HRQoL) demonstrated a striking divergence across different health domains. Patients reported remarkably preserved Physical Functioning, with a mean score of 94.5 (SD 12.0). Conversely, severe impairments were recorded in nearly all other areas.

The lowest mean score was observed in the Mental Health domain (22.4; SD 6.4), underscoring a profound psychological burden within this cohort. Other significantly impacted domains included Vitality (41.0; SD 8.5), General Health Perception (49.0; SD 10.5), Social Functioning (54.4; SD 13.0), and Pain (55.0; SD 15.0). Furthermore, Role Limitations due to Physical Health (56.2; SD 18.8) were more pronounced than Role Limitations due to Emotional Problems (70.0; SD 23.3), suggesting that daily productivity is restricted primarily by the physical constraints of the disease rather than emotional distress alone.

Discussion

To our knowledge, this study represents the first evaluation of HRQoL among patients with dystrophic epidermolysis bullosa (DEB) in a Polish cohort using multiple assessment tools. While previous research by Mącik and Kowalska-Dąbrowska has highlighted the significant reduction in life satisfaction among parents and caregivers of children with epidermolysis bullosa (EB) in Poland, the HRQoL of patients

themselves has not yet been researched [13]. HRQoL is a critical outcome measure, particularly in rare genetic conditions such as EB, for which treatment remains largely symptomatic [22].

Our findings confirm that DEB imposes a significant burden, affecting virtually all aspects of patients' lives. Dermatology-specific instruments, the DLQI and CDLQI, demonstrated a very large impact on quality of life, with mean scores of 13.9 and 14.3, respectively. These results are consistent with previous studies identifying DEB, especially the recessive subtype, as one of the dermatological conditions associated with the greatest HRQoL impairment [23-29].

In contrast, generic measures suggested a more moderate overall health perception, with a mean self-reported score of approximately 6/10 and an SF-36 general health score of 49.0. This discrepancy likely reflects the limited sensitivity of generic HRQoL instruments, which may overlook key disease-specific factors such as non-healing wounds and the substantial time burden associated with daily wound care. As a result, generic tools may fail to fully capture the burden of DEB [22,25]. Nevertheless, used alongside dermatology-specific questionnaires they still provide valuable insights into specific HRQoL domains.

In our cohort, a notable finding is the discrepancy between preserved physical functioning and high symptom burden. According to the SF-36, patients reported very high physical functioning (94.5), suggesting retained capacity for daily activities. In contrast, the Skindex-29 identified symptoms as the most impaired domain (48.9). This suggests that patients develop coping strategies and undergo a psychological 'response shift', recalibrating their internal baseline for health, that allow them to remain functionally active despite chronic discomfort [30,31].

The physical burden in DEB patients is primarily driven by skin fragility, recurrent blistering and chronic non-healing wounds, resulting in persistent pain and pruritus [11, 25-27, 31-35]. In our cohort, both symptoms were reported at moderate levels, with mean NRS scores of 5.7 for pain and 5.9 for pruritus. Daily care routines, specifically bathing and dressing changes, are perceived as particularly painful and burdensome, as they are time-consuming and often require assistance [25, 32-35, 36]. Pain also directly limits basic activities such as walking and eating, underscoring the critical need for the systemic implementation of standardized, state-funded pain and wound management protocols, which could substantially improve patient outcomes and reduce healthcare disparities [26].

Disease extent further contributes to HRQoL impairment. Most patients in our cohort reported involvement of approximately 4 – 7 anatomical locations, most commonly the right foot, right hand, and left foot. Syndactyly was among the most frequently reported complications, affecting 13 patients (39.4%). Hand and feet involvement is particularly impactful, as it significantly limits mobility and self-care, thereby increasing dependence in daily activities and significantly lowering QoL [35].

Beyond skin manifestations, patients reported a broad spectrum of complications, including microstomia, dental involvement, dysphagia, malnutrition, anaemia and anxiety, reflecting the multisystem nature of DEB. Gastrointestinal involvement, along with associated dietary restrictions and

nutritional deficiencies, is well documented and further contributes to physical and psychological burden [33, 36-37]. Optimizing nutritional intake, with a high-energy and high-protein diet, remains essential to support wound healing and overall health [37].

The cumulative burden of these physical, nutritional, and systemic complications inevitably takes a profound toll on patients' psychological well-being. Mental health emerged as the most severely affected domain in the SF-36 (22.4) followed by vitality (41.0). These findings were further evidenced by the Skindex-29 scores, where both emotions (40.3) and social functioning (41.5) indicated severe impairment. Chronic pain, visible skin lesions and other functional limitations lead to significant psychological distress. This is consistent with literature identifying high rates of depression and anxiety in this population [28, 34-35, 36, 38]. Notably, Margari et al. reported that while up to 80% of EB patients exhibit psychiatric symptoms, only a minority receive professional treatment [24]. Therefore, routine screening for mental health disorders and the integration of psychological support into standard medical care are imperative [28].

The visible nature of DEB further contributes to stigmatization, social isolation, and reduced participation in everyday life [38,39]. This, combined with the time-intensive nature of wound care, limits engagement in school, work, and leisure activities [36]. Social support, particularly from family members, has a strong positive influence on coping [24,28,30]. Furthermore, engagement with the broader EB community proved beneficial, as patients can share adaptive strategies and treatment information [33].

Despite the markedly reduced mental health scores, role limitations due to physical health (56.2) were more pronounced than those due to emotional problems (70.0) in the SF-36, indicating that productivity is more affected by physical symptoms, rather than emotional distress. This observation is consistent with previous findings [28, 40]. While symptom scores in our cohort are comparable to previous reports [25, 41], mental health outcomes appear substantially worse than those in other studies, which have reported relatively preserved mental health [11, 25, 41]. One explanation is adaptation, as some patients habituate to their condition and develop coping strategies to maintain a "normal" life, thus reducing perceived stress [31, 42]. However, in other patients, psychological burden may increase with age, as it was observed that anxiety and depression are more common in adults [40]. Importantly, these findings may also reflect broader systemic challenges in mental healthcare in Poland, including low mental health literacy and reduced help-seeking behavior [43].

To contextualize our findings, we compared the HRQoL and symptom burden of our Polish DEB cohort with previously published international studies (Table 3) [11, 25, 41, 44]. Notably, while our patients reported remarkably preserved physical functioning (SF-36 PF: 94.5) compared to Dutch (PF: 70.5) and Spanish (PF: 33.6–36.9) dystrophic EB cohorts, their mental health scores were substantially lower (SF-36 MH: 22.4 vs. 79.1 and 40.5–45.9, respectively). This stark divergence further reinforces the hypothesis of a strong physical 'response shift' and adaptation, overshadowed by a profound, unaddressed psychological toll. Furthermore, the substantial symptom burden and QoL impairment observed in our study are highly consistent with the severe clinical impact reported in other international

DEB cohorts (Table 3), confirming that despite high physical independence, the daily realities of the disease remain devastating.

This divergence between our findings and international literature may also be partially explained by healthcare system differences. In Poland, access to comprehensive and multidisciplinary EB care remains limited. Although specialized dressings are currently fully reimbursed, they previously imposed a financial burden on patients, a factor linked to diminished QoL [23, 26]. Moreover, treatment is largely restricted to symptomatic management and access to advanced therapies, such as birch triterpenes (Filsuvez®) and beremagene geperpavec (Vyjuvek®), is limited due to high costs and restricted availability. Furthermore, the absence of a dedicated national EB reference centre perpetuates a model of fragmented care, which is a critical unmet need in national rare disease policies. As a result, patients often face profound logistical challenges, including long travel distances to consult experienced clinicians, which amplifies both the financial burden of the disease and psychological distress [33]. In the absence of comprehensive systemic solutions, patient advocacy organizations act as a crucial buffer, striving to fill the existing gaps in care. Organizations such as the EB Polska Foundation and DEBRA Polska play a vital role by fostering a sense of community and providing essential psychological support for both patients and their families. Beyond these efforts, they organize workshops, facilitate access to qualified medical professionals, and promote disease awareness in both society and the medical community.

This study has several limitations that must be acknowledged. The small sample size reflects the rarity of DEB, and the lack of subtype stratification limits detailed comparisons. The relatively large standard deviations observed across HRQoL measures indicate considerable inter-individual variability. Additionally, the cross-sectional design precludes the assessment of changes over time. Finally, the lack of EB-specific validated questionnaires in Polish resulted in the use of less specific general tools, which may not fully detect the impact of EB symptoms on QoL [22, 25].

Conclusions

Despite relatively preserved physical functioning, patients with dystrophic epidermolysis bullosa in Poland experience a substantial and persistent symptom burden, accompanied by pronounced psychosocial impairment. This apparent discordance suggests the presence of adaptive coping mechanisms that enable maintenance of daily functioning, while simultaneously masking significant underlying physical discomfort and psychological distress. The findings point to considerable unmet clinical and supportive care needs within this population. These results underscore the urgent need for systemic healthcare improvements, particularly the establishment of a dedicated national reference center for epidermolysis bullosa to provide coordinated, multidisciplinary, and patient-centered care. Such a framework should integrate dermatological management with structured psychological support, pain control, and long-term follow-up. In parallel, improving equitable access to emerging disease-modifying therapies remains critical to reducing disease burden and addressing disparities between regions.

Overall, this study provides the first comprehensive national insight into the health-related quality of life and clinical burden of patients with DEB in Poland. It establishes an important evidence base for future longitudinal research and supports the development of targeted healthcare policies aimed at optimizing clinical outcomes and quality of life in this vulnerable population.

Abbreviations

EB – Epidermolysis bullosa

DEB – Dystrophic epidermolysis bullosa

HRQoL – Health-Related Quality of Life

NRS – Numeric Rating Scale

DLQI – Dermatology Life Quality Index

CDLQI – Children's Dermatology Life Quality Index

SF-36 - Short Form Health Survey

MH – mental health

PF – physical functioning

DEBRA – dystrophic epidermolysis bullosa research association

Declarations

Ethical approval and consent to participate

The study was conducted in accordance with the principles of the Declaration of Helsinki. The protocol was formally reviewed and approved by the Bioethics Committee of Medical University of Lodz (Resolution No. RNN/46/25/KE**).** Only individuals (or caregivers of minors) who had previously provided consent to be contacted were invited to participate in the study. For participants under 18 years of age, consent was obtained from a parent or legal guardian, who completed the survey on their behalf.

Availability of data and materials

The datasets generated and analyzed during the current study contain sensitive or potentially identifiable information and are therefore not publicly available due to ethical restrictions. Data are available from the corresponding author upon reasonable request.

Competing interests

The authors declare that they have no competing interests.

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Author contributions

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Tables

Table 1. Demographic and clinical characteristics of the study cohort (n = 33).

Characteristic	Value (n = 33)
Demographics	
Sex, female, n (%)	25 (75.8%)
Sex, male, n (%)	8 (24.2%)
Age, mean (SD), years	23.7 (±12.4)
Age range	5 weeks – 43 years
Age group: Pediatric (<18 years), n (%)	16 (48.5%)
Age group: Adult (≥18 years), n (%)	17 (51.5%)
Diagnosis Confirmation	
Genetic confirmation, n (%)	24 (72.7%)
Clinical diagnosis only, n (%)	9 (27.3%)
Clinical Manifestations & Complications	
Affected anatomical locations, mean (median; range)	6.76 (6.0; 1–12)
Nail dystrophy, n (%)	26 (78.8%)
Dysphagia, n (%)	16 (48.5%)
Syndactyly (pseudosyndactyly), n (%)	13 (39.4%)
Malnutrition, n (%)	9 (27.3%)
Alopecia, n (%)	7 (21.2%)
Anemia, n (%)	4 (12.1%)
Anxiety disorder, n (%)	4 (12.1%)
Management and Therapy	
Use of non-adhesive dressings, n (%)	33 (100%)
Use of wound-healing adjuvants, n (%)	27 (81.8%)
Systemic therapy, n (%)	1 (3.0%)

Table 2. Comprehensive overview of Health-Related Quality of Life (HRQoL) scores across dermatology-specific and generic instruments.

Instrument / Domain	Mean (SD)	Median	Range (Min–Max)
Dermatology-Specific Instruments			
DLQI (Adults)	13.9 (±6.99)	13.0	2.0 – 29.0
CDLQI (Children)	14.3 (±7.60)	12.0	4.0 – 28.0
Skindex-29 (Scale 0–100)*			
Symptoms	48.9 (±17.6)	48.6	22.9 – 80.0
Emotions	40.3 (±19.5)	42.0	8.0 – 80.0
Functioning	41.5 (±11.8)	41.7	10.0 – 75.0
Total Score	42.9 (±26.3)	41.4	15.2 – 77.9
Generic Health Survey (SF-36) (Scale 0–100)**			
Physical Functioning	94.5 (±12.0)	95.0	70.0 – 100.0
Role Limitations (Physical)	56.2 (±18.8)	62.5	25.0 – 100.0
Bodily Pain	55.0 (±15.0)	50.0	22.0 – 90.0
General Health Perception	49.0 (±10.5)	50.0	25.0 – 77.0
Vitality (Energy/Fatigue)	41.0 (±8.5)	40.0	25.0 – 60.0
Social Functioning	54.4 (±13.0)	55.0	25.0 – 87.5
Role Limitations (Emotional)	70.0 (±23.3)	66.7	33.3 – 100.0
Mental Health	22.4 (±6.4)	22.0	12.0 – 36.0

Table 3. Comparative analysis of Health-Related Quality of Life (HRQoL) and symptom severity between the current cohort and international dystrophic epidermolysis bullosa studies.

BP, Bodily Pain; CDLQI, Children's Dermatology Life Quality Index; DDEB, Dominant Dystrophic Epidermolysis Bullosa; DEB, Dystrophic Epidermolysis Bullosa; DLQI, Dermatology Life Quality Index; EBS, Epidermolysis Bullosa Simplex; GH, General Health; JEB, Junctional Epidermolysis Bullosa; MH, Mental Health; N/A, Not Available; NRS, Numeric Rating Scale; PF, Physical Functioning; RDEB, Recessive Dystrophic Epidermolysis Bullosa; RE, Role Limitations due to Emotional Problems; RP, Role Limitations due to Physical Health; SF, Social Functioning; SF-36, 36-Item Short Form Health Survey; VAS, Visual Analogue Scale; VT, Vitality

STUDY	STUDY POPULATION	DLQI / CDLQI (Mean / Distribution)	SF-36 (All 8 Domains: Mean)	SKINDEX-29 (Sub-scores: Mean)	NRS / VAS PAIN & PRURITUS (Mean)
Current Study (2026, Poland)	**n = 33** (DEB: 33)	**13.9** (DLQI mean) **14.3** (CDLQI mean)	**PF:** 94.5 **RP:** 56.2 **BP:** 55.0 **GH:** 49.0 **VT:** 41.0 **SF:** 54.4 **RE:** 70.0 **MH:** 22.4	**Symptoms:**48.9 **Emotions:**40.3 **Functioning:**41.5	**Pain:** 5.7 (NRS) **Pruritus:** 5.9 (NRS)
Yuen et al. (2014, Netherlands)	**n = 55** (EBS: 29, JEB: 8, DDEB: 13, RDEB: 5)	**N/A**	**PF:** 70.5 **RP:** 62.7 **BP:** 53.4 **GH:** 65.5 **VT:** 60.8 **SF:** 70.7	**Symptoms:**45.0 **Emotions:**24.9 **Functioning:**26.0	**N/A**

STUDY	STUDY POPULATION	DLQI / CDLQI (Mean / Distribution)	SF-36 (All 8 Domains: Mean)	SKINDEX-29 (Sub-scores: Mean)	NRS / VAS PAIN & PRURITUS (Mean)
			RE: 81.2 **MH:** 79.1		
Jeon et al. (2016, South Korea)	**n = 13** (RDEB: 13)	**N/A**	**N/A**	**Symptoms:**86.31 **Emotions:**75.23 **Functioning:**76.69	**Pain:** 6.54 (VAS) **Pruritus:** 7.54 (VAS)
Villar Hernández et al. (2022, Spain)	**n = 16** (DDEB: 6, RDEB: 10)	**N/A**	**DDEB:** **PF:** 36.9 **RP:** 38.8 **BP:** 34.1 **GH:** 35.9 **VT:** 33.8 **SF:** 37.3 **RE:** 42.7 **MH:** 40.5 **RDEB:** **PF:** 33.6 **RP:** 38.1 **BP:** 37.0 **GH:** 30.0 **VT:** 39.0 **SF:** 34.4 **RE:** 43.3 **MH:** 45.9	**N/A**	**N/A**
Paller et al. (2022, Multinational)	**n = 28** (DEB: 28) 15 Adults, 13 Children	**DLQI:** 60.1% reported moderate to extremely large effect. **CDLQI:** 61.6% reported moderate to extremely large effect.	**N/A**	**N/A**	**N/A**

Figures

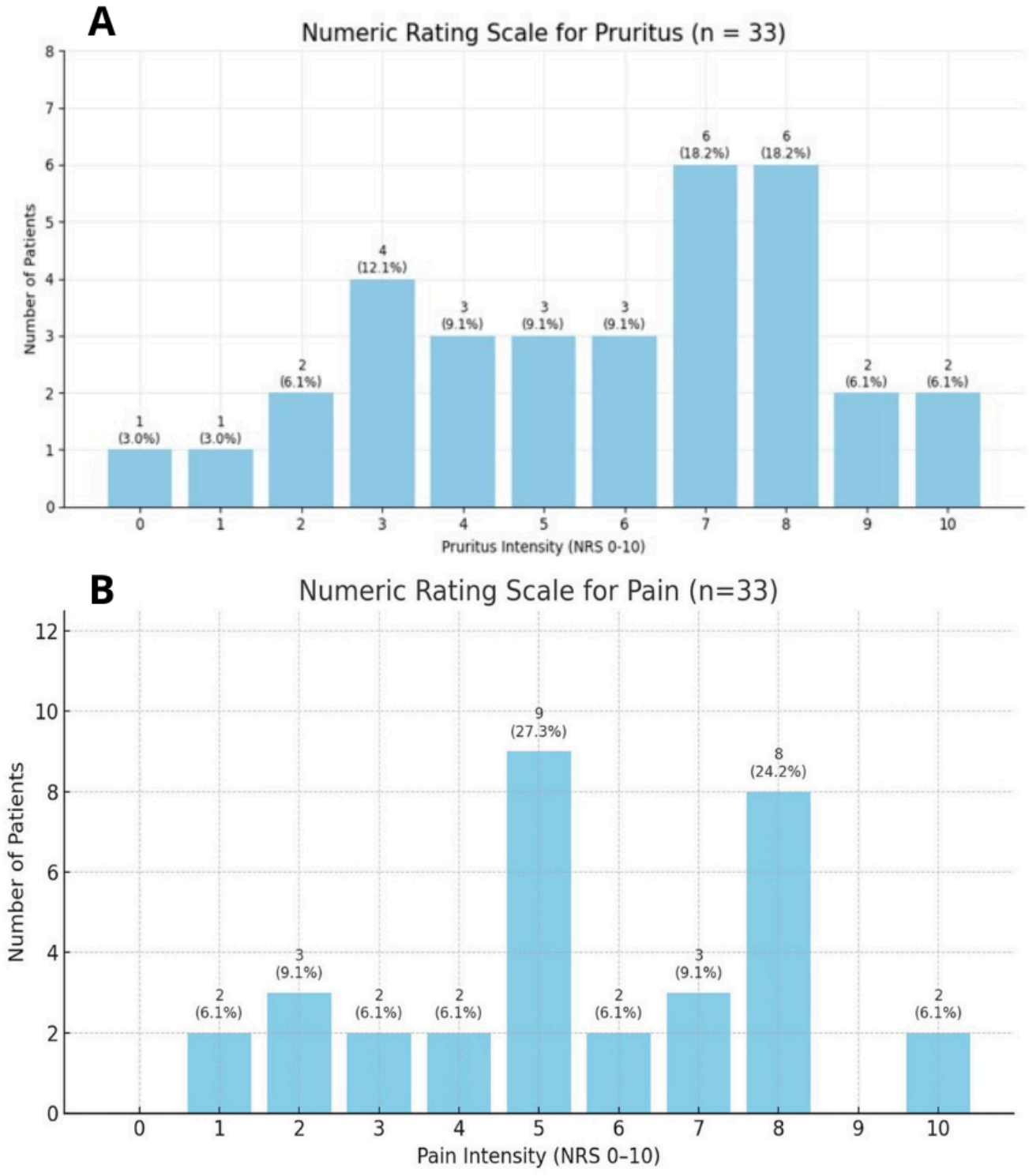


Figure 1

Distribution of pain (**A)** and pruritus (**B)**severity in the study cohort.