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Assessing pain catastrophizing and functional disability in pediatric epidermolysis bullosa patients

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CONFLICT OF INTEREST

No conflict of interest.

CONSENT STATEMENT

Informed consent was obtained from parent(s)/guardian(s).

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Abstract

Background/Objectives: The primary objective was to assess pain catastrophizing and functional disability in pediatric patients with epidermolysis bullosa (EB) and their parents/guardians. Secondary objectives included examining relationships between pain catastrophizing, functional disability, and correlations with other factors (e.g., age, disease severity, and percent of body surface area (BSA) involved).

Methods: Patients with EB ages 8–16 and their parents/guardians who were English or Spanish speaking completed a one-time online survey. Parent measures included: demographics questionnaire, Pain Catastrophizing Scale-Parent (PCS), and Parent Functional Disability Inventory (FDI). Child measures included: PCS child and child FDI. Higher scores on both scales indicate higher levels of catastrophizing and functional disability.

Results: Of 31 children, the mean age was 11.47 years and the majority (70.97%) had dystrophic EB. Mean scores were: 35.84 = PCS parent; 34.58 = PCS child; 30.87 = parent FDI; 29.77 = child FDI. Total scores for PCS parent, parent FDI, and child FDI increased significantly with disease severity and percentage of involved BSA ($p < .01$ for all). Total scores for PCS child increased significantly with percent of EB skin involvement ($p = .04$) but not disease severity. Older children reported more functional disability than their parents and younger children ($p = .02$).

Conclusions: Our results demonstrate significant positive correlations between negative thoughts related to pain and the experience of functional difficulties in patients with EB and their caregivers. Psychological, psychiatric, and/or behavioral interventions to help managing chronic pain may be effective for patients with EB.

Keywords

children; epidermolysis bullosa; pain; pediatric dermatology; pediatric psychology

1 | INTRODUCTION

Epidermolysis bullosa (EB) is a group of complex genetic diseases resulting in fragile skin and blistering from minor trauma.[1] Most patients with EB experience chronic itch and daily pain.[2] Patients have frequently reported feelings of negative affect (e.g., sadness, frustration,

etc.) and difficulties with interpersonal relationships due to pain.[3] EB has been demonstrated to decrease quality of life (QoL) for many patients, even those with milder disease.[4,5] In addition to decreased QoL, pain related to EB negatively impacts functioning. Children with each of the major forms of EB reported difficulties with age-appropriate and functional activities, including needing assistance with activities of daily living and walking.[6]

Pain catastrophizing refers to exaggerated or perseverative worry about the negative consequences of pain, perceived lack of control over symptoms, and an inability to cope with pain.[7] It includes the constructs of rumination, magnification, and helplessness.[7] Pain catastrophizing is well studied in children with chronic pain (e.g., headache, functional abdominal pain, and inflammatory bowel disease) and is associated with decreased functioning, increased anxiety, and increased depression across populations.[8–10]

A review of the literature did not reveal any prior studies on pain catastrophizing and functional disability in children with EB or their parents. Given the chronic and painful nature of EB, we hypothesized that assessing pain catastrophizing and related functional difficulties in patients and parents/caregivers might provide insight into psychological treatment approaches for EB. Our primary objective was to examine levels of pain catastrophizing and functional disability in these populations. Our secondary objective was to examine the relationships between pain catastrophizing and functional disability, as well as any correlations between these variables and individual factors (e.g., age, severity of disease, percent body surface area affected by EB).

2 | METHODS

Eligible participants were children with EB ages 8–16, and their parents or legal guardians. Participants were also identified through the EB Clinical Research Consortium, whose investigators informed patients and their parents/guardians about the survey and provided survey links as part of the EB Clinical Characterization and Outcomes Database (University of Colorado IRB #12–0321). Survey links were also distributed by EB non-profit organizations. The de-identified survey was completed online through REDCap, at a single time point. The PCS and FDI instruments were translated into Spanish by an approved translation service. This study was approved by the parent site, Children’s Hospital of Philadelphia IRB #19–016442.

Demographic information was obtained for the parent and child (e.g., age, race, ethnicity, location of residence, household income, parent’s highest education, child gender, age at EB diagnosis, time since EB diagnosis, self-reported EB severity). We estimated subjective severity by asking parents to determine body surface area (BSA) involvement and EB subtype (epidermolysis bullosa simplex, junctional EB, dystrophic EB, Kindler syndrome, other). BSA was assessed by asking: Of the following which best describes the area of involvement of EB in your child: 0%–25%, 25%–50%, 50%–75%, 75%–100%. We asked whether the child or caregiver had ever undergone any mental health intervention (i.e., therapy and/or medication). We also asked if the child is currently taking any pain medications (defined as either over-the-counter or prescription medications).

The Pain Catastrophizing Scale (PCS) is a validated instrument that asks participants to reflect on past painful experiences and indicate the degree to which they experienced each of 13 thoughts or feelings when experiencing pain, on 5-point scales from (0) not at all to (4) all the time.[7] The PCS yields a total score by summing responses to all 13 items (range from 0 to 52) and three subscale scores assessing rumination (e.g., “I can’t stop thinking about how much it hurts”), magnification (e.g., “I’m afraid that something serious might happen”), and helplessness (e.g., “There is nothing I can do to reduce the intensity of my pain”). The PCS has been validated in many studies with robust internal consistency (Cronbach’s alpha .87), and each of the subsections have robust internal consistency as well (Cronbach’s alpha: rumination = .87; magnification = .60, and helplessness = .79).[7]

The Functional Disability Inventory (FDA) is a 15-item validated instrument developed to assess disability in children and adolescents by rating the amount of difficulty they have completing each activity on a 5-point Likert scale (from 0 = no trouble to 4 = impossible).[11] The total FDI score is a sum of all of the items and scores range from 0 to 60, with higher scores indicating greater functional disability.[12] Recently, clinical reference points were developed to identify 3 categories of disability in pediatric chronic pain, that is, no/minimal disability (0–12), moderate disability (13–29), and severe disability (30), and these can be used for children and adolescents with a variety of pain conditions, including widespread chronic pain.[13] The FDI has been validated and used in many studies and had similar high internal consistency for both of its sections (Cronbach’s alpha: physical activities = .91 and daily activities = .77).[11]

Summary statistics were calculated including mean and standard deviation for continuous variables and frequencies and percentages for categorical variables. Non-parametric tests were used when assessing associations with characteristic variables. Differences across parent and child scores were determined with paired Wilcoxon rank sum test. Score differences across variable levels were measured with Kruskal–Wallis test. Association between ordinal variables EB severity and BSA and associations with scored outcomes were assessed with Spearman correlations. Associations of the parent and child scores and association with age were evaluated in a Pearson correlation matrix. Statistical analysis was performed in SAS 9.4.

3 | RESULTS

Forty-one parents responded to the survey, one for each child. Of 41 responders, 31 completed all surveys (76%) and were included in the study. Twenty-three were English-speaking and 8 were Spanish-speaking. Demographics of parents and children are shown in Table 1.

Parent and child PCS and FDI scores are shown in Table 2. Children had a significantly lower median PCS Rumination score than parents ($p < .01$). However, there were no other significant differences between parent PCS and child PCS sub-scores.

Median parent total PCS scores increased along with increasing disease severity and BSA ($p < .01$ for both) (Table 3). Median child total PCS scores increased with increasing disease

severity, but this was not statistically significant ($p = .2$). Similarly, median child total PCS scores tended to increase with increasing BSA, but this was not statistically significant ($p = .08$) (Table 3).

The average parent FDI score (30.77) was severe while the average child FDI score (29.77) was borderline severe (Table 2). An FDI score greater than or equal to 30 indicates severe disability. Both median parent FDI and median child FDI scores increased with increasing EB severity and higher BSA ($p < .01$ for all) (Table 3). Parent FDI total scores were higher for those whose children received pain medication ($p = .02$). There were no other significant correlations between FDI or PCS scores and pain medication use, perceived pain medication efficacy, or prior mental health intervention (in either child or parent).

All parent and child scores (PCS total, PCS subscale scores, and FDI Total scores) had significant positive correlations, except for PCS magnification ($p = .06$). Correlations between PCS and FDI measures and child age, disease severity, and BSA are shown in Table 4.

4 | DISCUSSION

Our findings demonstrate the prevalence of negative thoughts related to pain and challenges patients with EB face with daily functioning. Patients with EB and their parents reported higher average PCS and FDI scores compared to pediatric patients with functional abdominal pain.[14] In parents of children with EB, the average parent PCS total score was approximately 8 points higher than parents of children with functional abdominal pain. In patients with EB, the average child PCS total score was approximately 19 points higher and the average child FDI total score was approximately 10 points higher than children with functional abdominal pain.[14] Patients with EB also reported higher PCS scores than pediatric patients with chronic and episodic migraines.[15] The average child PCS scores for chronic and episodic migraine patients were both under 30, while patients with EB had an average PCS score of 34.58.

Child and parent PCS scores were consistent with each other, meaning that both acknowledge the difficulties patients with EB face with negative thoughts about pain. Child and parent FDI scores were also consistent with one another, meaning that both are aware of the functional difficulties patients with EB face. Parents did not overestimate the degree of negative thoughts, or the degree of functional difficulties experienced by their children.

Both parents and children with more severe disease and larger involved BSA reported higher PCS and FDI scores. This is consistent with increased pain associated with larger wound area and those with more severe types of EB.[16,17]

These findings objectively confirm that those with EB are more likely to experience difficulties coping with pain and engagement in functional activity. These increased difficulties are likely multifactorial due to the progressive complications and extracutaneous manifestations of EB such as dental and oral cavity problems (contributing to FDI question of eating regular meals), hand/foot contractures (contributing to FDI question of walking/running). Wound care may take hours and is exhausting for patients.[16] The increased time

for wound care and overall disease management may also negatively affect daily functioning for patients. Our results add to the literature on the challenges that patients with EB and their parents face when managing chronic pain and performing daily tasks.

Parent FDI scores were higher for those whose children received pain medications, perhaps indicating increased disease severity not captured elsewhere. Targeted efforts for patients with more severe and widespread EB are necessary to help effectively cope with pain and functional difficulties due to EB. Although those who received pain medications and mental health interventions primarily reported moderate to severe disease (89.5% and 81.2%, respectively) as expected, pain and functional disability scores still increased with severity of disease and there was no significant difference compared to those who had not received interventions. However, this lack of significance may be due to small sample size.

Mental health resources are underutilized in this population with only 35% of children and 29% of parents having had mental health intervention. This study suggests that children with EB, and their caregivers, are likely to benefit from well-established psychological interventions focused on coping with chronic pain, improving functioning, and modifying negative thinking patterns (e.g., rumination, magnification, helplessness). A previous study found that children who showed more acceptance and distancing showed higher levels of functioning.[18] As EB can affect several organ systems, multidisciplinary care and coping strategies can improve the overall quality of life for both patients and parents.[19] While pain medications can improve pain symptoms, providers should work together to also incorporate referrals to a psychologist for evaluation if signs of stress, anxiety, or depression are present. A recent study highlighted the use of trauma-informed care strategies in pediatric EB patients and could be particularly useful for those experiencing psychological distress from pain.[19] Early intervention is important in preventing worsening of functional ability as time progresses, as we found that older patients reported higher levels of disability compared to younger children and their parents.

Our study is limited by the small sample size and cross-sectional design. The sample size is perhaps too small to assess age as a predictor for catastrophizing either because of neurocognitive development and/or increased painful experience. The majority of those who responded had dystrophic EB, which is a possible recruitment bias. We also did not break down the type of dystrophic EB further (autosomal dominant [DDEB] or autosomal recessive [RDEB]); DDEB patients tend to have milder disease than RDEB patients. Since the highest child PCS scores, parent PCS scores, and parent FDI scores were for BSA 50%–75% and not 75%–100% as would be expected, subjective reporting of BSA may not be standardized across all participants, thus introducing possible inaccuracies. Additionally, extracutaneous pain is not captured by BSA.

Future studies should include assessments of anxiety and depression, as these can certainly influence both pain catastrophizing and functional disability. Further research is warranted with larger sample sizes to advance the complicated management of pain coping and related functional difficulties for patients with EB, as well as determine the effects of psychological intervention and treatment with psychiatric medications on the perception of pain in these patients.

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DATA AVAILABILITY STATEMENT

The data that support the findings of this study are available from the corresponding author upon reasonable request.

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TABLE 1

Parent and patient characteristics ($n = 31$)

Descriptor	Parameter	Value
Parental age, mean (SD)	Years	41 (8.9)
Parental race, *N* (%)	White	19 (61.3)
	Multiple	9 (29.0)
	Other	2 (6.5)
	Prefer not to answer	1 (3.2)
Parental ethnicity, *N* (%)	Non-Hispanic or Latino	19 (61.3)
	Hispanic or Latino	10 (32.3)
	Prefer not to answer	2 (6.5)
Parental education, *N* (%)	Up to 8th grade	4 (12.9)
	High school graduate	7 (22.6)
	Some college, no degree	2 (6.6)
	Trade/technical training	1 (3.2)
	Associate degree	3 (9.7)
	Bachelor's degree	6 (19.4)
	Master's degree	8 (25.8)
Location, *N* (%)	Midwest	3 (9.7)
	Northeast	7 (22.6)
	Southeast	5 (16.1)
	Southwest	3 (9.7)
	West	4 (12.9)
	Non-United States	9 (29.0)
Parent's yearly income, *N* (%)	<$50,000	13 (41.9)
	$50,000-$150,000	13 (41.9)
	≥$150,000	5 (16.1)
Child age, mean (SD)[a]	Years	11.57 (4.4)
Child gender, N (%)	Male	16 (51.6)
	Female	15 (48.4)
EB subtype, N (%)	EB simplex	6 (19.4)
	Junctional EB	2 (6.5)
	Dystrophic EB	22 (71.0)
	Other	1 (3.2)
Body surface area of involvement of EB, *N* (%)	0%−25%	6 (19.4)
	25%−50%	9 (29.0)
	50%−75%	7 (22.6)
	75%−100%	9 (29.0)
Self-reported EB severity, *N* (%)	Mild	8 (25.8)
	Moderate	14 (45.2)
	Severe	9 (29.0)
Anatomic area of EB involvement, *N* (%)[b]	Head/face/neck	23 (74.2)

Descriptor	Parameter	Value
	Mouth/teeth	21 (67.7)
	Trunk	22 (71.0)
	Arms	24 (77.4)
	Hands	27 (87.1)
	Legs	28 (90.3)
	Genitalia	15 (48.4)
Child mental health intervention, *N*(%)	Yes	11 (35.5)
Parent mental health intervention, *N*(%)	Yes	9 (29.0)
Child pain medication, *N*(%)	Yes	19 (61.3)
Pain medication efficacy, *N*(%)[c]	Not at all	1 (6.7)
	Slightly	4 (26.7)
	Moderately	5 (33.3)
	Very	5 (33.3)

[a] Data available for 30 of 31 participants.

[b] Participants were allowed to select more than one choice.

[c] Data available for 15 of 19 children taking pain medication.

TABLE 2

Parent and child pain catastrophizing scale (PCS) total and subscale scores and Functional Disability Inventory (FDI) total scores (n = 31)

Score parameter[a]	Mean (SD)		Mean (SD)
PCS parent total	35.84 (10.3)	PCS child total	34.58 (13.6)
PCS parent rumination	14.35 (3.5)	PCS child rumination	12.45 (4.4)
PCS parent magnification	6.90 (2.6)	PCS child magnification	7.03 (3.2)
PCS parent helplessness	14.58 (5.6)	PCS child helplessness	15.10 (6.9)
Parent FDI total	30.87 (12.8)	Child FDI total	29.77 (13.0)

[a]Higher scores on both the PCS and FDI indicate higher levels of catastrophizing and functional disability.

FDI: no/minimal disability (0–12), moderate disability (13–29), and severe disability (30).

TABLE 3

Median total PCS and FDI scores by disease severity and body surface area of involvement of EB ($n = 31$)

	Disease severity			**Percent BSA**			
	Mild ($n = 8$)	**Moderate ($n = 14$)**	**Severe ($n = 9$)**	**0%–25% ($n = 6$)**	**25%–50% ($n = 9$)**	**50%–75% ($n = 7$)**	**75%–100% ($n = 9$)**
PCS scores							
Parent	29	31	45	28	29	45	39
Child	25.5	34	40	24	27	47	39
FDI scores							
Parent	7	16	24	6	14	25	18
Child	7	18	21	4	15.5	20	21

TABLE 4

Correlations (and *p*-value) between PCS and FDI measures and child age, disease severity, and percent of EB (BSA) ($n = 31$).

	Child age		Disease severity		Body surface area of involvement of EB	
	***R* score**	***p* value**	***R* score**	***p* value**	***R* score**	***p* value**
PCS parent scores						
Total	0.03	.89	0.61	<.01[a]	0.48	<.01[a]
Rumination	0.23	.23	0.60	<.01[a]	0.52	.01[a]
Magnification	−0.24	.21	0.39	.03[a]	0.36	.047[a]
Helplessness	0.02	.91	0.58	<.01[a]	0.35	.054
Parent FDI total	0.04	.82	0.69	<.01[a]	0.53	<.01[a]
PCS child scores						
Total	0.18	.34	0.31	.09	0.38	.04[a]
Rumination	0.04	.83	0.47	<.01[a]	0.54	<.01[a]
Magnification	0.22	.24	0.10	.58	0.20	.27
Helplessness	0.23	.22	0.36	.04[a]	0.34	.058
Child FDI total	0.44	.02[a]	0.57	<.01[a]	0.64	<.01[a]

Note: Correlations with child age were determined using Pearson correlation coefficients. Prob > |*r*| under H0: Rho = 0. Correlations with disease severity and body surface area involvement of EB were determined using Spearman correlation coefficients.

[a]Statistical significance ($p < .05$).