

The need of social support, life attitudes and life satisfaction among parents of children suffering from epidermolysis bullosa

Zapotrzebowanie na wsparcie społeczne, postawy życiowe i satysfakcja z życia rodziców dzieci chorych na *epidermolysis bullosa*

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wsparcie społeczne, rodzice, satysfakcja z życia, postawy życiowe, *epidermolysis bullosa*.

ABSTRACT

Introduction. Epidermolysis bullosa (EB) is a group of rare genetic skin disorders. Much is known about the disease from the medical perspective, but there are still only a few studies about the psychological condition of the patient or his family. The birth of a child with EB may become a trauma for the parents. Parents' feelings also vary depending on the type of EB.

Objective. The main aim of the study was to determine whether, and in what way, having a child suffering from EB influences parents' life and psychological well-being. Due to this aim, life satisfaction, life attitudes and the need for social support were measured.

Material and methods. Twenty-one parents of ill children were studied (13 mothers, 8 fathers; 16 parents had one child with EB and 5 parents two). Six children suffered from EB simplex, 1 child from junctional EB and 14 from dystrophic EB. The course of the disease was mild in 9 children and severe in 12. As the control group 22 parents of healthy children were chosen.

Results. The parents of ill children have a greater need for social support from family, both general, and instrumental, appraisal and emotional. The need for support from medical institutions is at a low level. In life attitudes there are also significant differences. Parents of ill children have a higher level of existential vacuum, and a lower level of life control and life purpose. Also the life satisfaction is lower.

Conclusions. The results of our study indicate that a child suffering from EB is a heavy burden for the parents.

STRESZCZENIE

Wprowadzenie. *Epidermolysis bullosa* (EB) jest grupą rzadkich genetycznych chorób skóry, pojawiających się raz na kilkaset urodzeń. Z medycznego punktu widzenia choroba jest dobrze poznana, ale wciąż niewiele jest opracowań dotyczących problemów psychologicznych chorego i jego rodziny. Narodziny dziecka z EB mogą stać się specyficznym rodzajem traumy dla jego rodziców, a ich odczucia zależą również od typu i przebiegu choroby. W części piśmiennictwa sugeruje się, że EB jest obciążeniem nie tylko dla chorego, lecz także dla jego rodziny.

Cel pracy. Głównym celem prezentowanych badań było sprawdzenie, czy i w jaki sposób posiadanie dziecka chorego na EB wiąże się z różny-

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mi zmianami w życiu i dobrostanem psychicznym. Uwzględniając cel, mierzono satysfakcję z życia, postawy życiowe oraz zapotrzebowanie na wsparcie społeczne.

Materiał i metodyka. Przebadano 21 rodziców chorych dzieci (13 matek, 8 ojców, 16 rodziców miało jedno dziecko chore na EB, 5 – dwoje). Typ EB prosty wystąpił u 6 dzieci, EB łączący – u 1, EB dystroficzny – u 14. Przebieg choroby był łagodny w 9 przypadkach i ciężki w 12 przypadkach. Grupę kontrolną stanowiło 22 rodziców zdrowych dzieci.

Wyniki. Rodzice dzieci chorych mają wyższe zapotrzebowanie na wsparcie od rodziny, zarówno w wymiarze ogólnym, jak i w wymiarach instrumentalnym, wartościującym i emocjonalnym. Zapotrzebowanie na wsparcie od instytucji medycznych jest na niskim poziomie w obu grupach. Istotne statystycznie różnice zaobserwowano również w postawach życiowych. Rodzice dzieci chorych mają istotnie większą pustkę życiową, a także mniejsze poczucie kontroli życia i jego celowości. Satysfakcja z życia jest także istotnie niższa.

Wnioski. Wyniki naszych badań wskazują, że dziecko chore na EB jest istotnym obciążeniem dla rodzica.

INTRODUCTION

Epidermolysis bullosa (EB) is “a group of rare genetic skin conditions, which is characterised by extremely fragile skin and recurrent blister formation, resulting from minor mechanical friction or trauma” (DEBRA International http://debra-kd.pl/?page_id=26).

Three main types of EB can be distinguished:

- EB simplex (EBS) – the blisters are formed within keratinocytes of the basal layer and above the basal layer;
- junctional EB (JEB) – the blisters are formed within the lamina lucida of the basement membrane;
- dystrophic EB (DEB) – the blisters are located within the sublamina densa of the basement membrane [1].

These types divide further into subtypes in regard of the causes of blister formation in a specific location.

Epidermolysis bullosa is a rare disease – it occurs once in several hundred births. In Poland there are currently around 500 people (children and adults) suffering from it. The treatment of EB is entirely symptomatic, with no possibility of remission. It leads to many problems with functioning, both physical (discomfort, pain, necessity to be careful all the time, changes such as contractures etc., using dressings) and mental (a perspective of life with disease and pain, often bad prognoses, troubled social relations caused by the visibility of the disease and the lack of social awareness).

Currently there are many studies conducted on EB patients, but most of them focus on the medical aspect of the disease, its sources and seeking ways

of treatment. A relatively small number of studies relate to the psychological aspects of living with the disease [2–4]. Appreciating the value of medical research, it is worth noting that the progress in therapy is a distant matter whereas people suffering from EB and their families encounter their difficulties now; therefore, it is important to pay attention to their psychological well-being as well. A large proportion of persons suffering from the disease consists of children and young adults. The studies are conducted on:

- the demand for multi-dimensional care [5], including psychological support [3, 4, 6];
- the feeling of differentness and exclusion from peer groups [6, 7];
- the influence of EB on the quality of life [2, 8, 9],
- the influence of EB on the patient’s family [8–10].

The birth of a child who has been diagnosed with EB is a special kind of trauma for the parents. Parents’ feelings also vary depending on the type of EB. While EB simplex usually takes a mild form, dystrophic EB has a various course with possible complications (e.g. problems such as fixation of the tongue, fusion of the fingers, shrinkage of the mouth and oesophageal stenosis) and in severe cases often leads to significant shortening of lifetime. Junctional EB always has a severe course and usually ends with death during the first years of life [11]. Each situation affects the parents of ill children differently. Taking care of an incurably ill child often puts a parent in a hard situation and requires an assessment of the child’s needs at every stage of being with it [12–16]. In the case of EB, there are practically no such studies

at all. Only a few studies mention the influence of the child's disease on family life [10, 17, 18].

In Poland no research focused on parents of children suffering from EB has been carried out. The specific situation regarding difficulties in financing healthcare in Poland, the repeatedly difficult access to specialists, the costs of taking care of a child (including the necessity for one parent to resign from work or to supply specialist medicines and dressings) and often a lack of a holistic attitude to the problem of a family with an incurably ill child can all be significant factors influencing the mental condition of a parent.

The present study focused on estimating parents' need for social support (both from medical institutions and from family), estimating life satisfaction of parents and assessing their life attitudes in relation to their life conditions.

OBJECTIVE

Due to the lack of research and the need of helping the ill child's parents, two objectives of the study were set: 1) testing whether, and in regard to what aspects, a child's disease differentiates the parents' life attitudes and their need for support; 2) testing which of the analysed variables are significant for parent's life satisfaction.

MATERIAL AND METHODS

Regarding the objectives stated above, in the case of the first one, the independent variable was having a child suffering from EB or having only healthy children, whereas the dependent variables were life satisfaction level, life attitudes and the need for support.

In order to accomplish the second objective, it was decided to carry out stepwise regression method analyses, where the dependent variable was life satisfaction and the independent variables were life attitudes and the need for support.

The variables were measured using standardised methods: Life satisfaction - with the Satisfaction With Life Scale (SWLS) by E. Diener, R. A. Emmons, R. J. Larson, S. Griffin in the Polish adaptation by Zygfryd Juczyński [19]; Life attitudes - the Life Attitudes Profile - Revised (LAP-R) scale by Gary T. Reker in the Polish adaptation by Ryszard Klamut [20]; Support - support from family and medical institutions was evaluated with the scale of social support by Kmiecik-Baran on four levels: information, instrumental, appraisal and emotional support [21].

The following hypotheses were proposed:

- H1. The child's incurable disease leads to a decrease of parents' life satisfaction and to difficulties in defining their life attitudes.

- H2. The child's incurable disease is associated with a greater need for support from family.

- H3. The child's incurable disease is associated with a greater need for support from medical institutions.

- H4. A model presenting the influence of the variables on the life satisfaction of the parents of ill children can be defined.

In order to test the hypotheses, the participants of a rehabilitation camp organised by 'Debra' Kruchy Dotyk Society in Poland were examined. The person studied was a parent staying there as a child's guardian. There were 21 parents studied, including 13 mothers and 8 fathers, whose age varied between 30 and 59 years with the average of 39. About 66% of the respondents had 2 children, two families had more than 2 children, and 3 families had 1 child. Among the people studied, 16 parents had one child suffering from EB and 5 parents had 2. Six children suffered from epidermolysis bullosa simplex, 1 child from junctional epidermolysis bullosa and 14 from dystrophic epidermolysis bullosa. The course of the disease was mild in nine children and severe in 12 (the course was evaluated by a physician according to medical criteria).

The data concerning the type of EB and its course were gathered from the child's medical records. Only 2 parents knew before that they were EB gene carriers. All of the families were full families. In two cases both parents worked, and in other cases the mother had resigned from work and was taking care of the ill child. The material status of the family was defined as "average" by 19 respondents, and as "rather bad" by two. The education level of the studied parents was at least secondary.

The control group consisting of parents of healthy children was selected in a way that it maintained the demographic variables structure and it was composed of 22 parents of healthy children.

RESULTS

Initially it was decided to test whether the fact of having a child suffering from EB differentiates the parents in regard of their life satisfaction, life attitudes and the need for social support.

Table 1 presents the differences together with their significance between parents of healthy and ill children in life satisfaction and life attitudes, and Table 2 presents differences in regard to the need for support. Statistical parameters of the data allow for the use of parametric tests.

It can be noted that the parents of ill children have greater difficulties in the area of life attitudes, and their life satisfaction is significantly lower than in the case of parents of healthy children.

Table 1. Differences in the range of life satisfaction and life attitudes**Tabela 1.** Różnice w zadowoleniu i podejściu do życia

Variable	Parents of children suffering from EB (N = 21)		Parents of healthy children (N = 22)		Value of t	Value of p
	Mean	SD	Mean	SD		
Life purpose	4.52	2.46	6.18	1.73	2.562	0.014
Coherence	4.66	2.51	5.77	1.87	1.639	0.109
Life control	4.23	2.25	5.50	1.73	2.060	0.046
Death acceptance	5.42	1.63	5.04	1.78	-0.734	0.467
Existential vacuum	5.90	2.11	4.45	1.18	-2.753	0.010
Goal seeking	4.61	1.85	3.81	1.89	-1.400	0.169
Personal meaning	4.38	2.53	5.90	1.82	2.275	0.028
Life attitude balance	4.57	2.01	6.27	1.93	2.827	0.007
Life satisfaction	4.57	2.08	6.27	1.80	2.863	0.007

Note: all scale's theoretical scores: 1–10.

Table 2. Statistical differences between parents in the need of support**Tabela 2.** Różnice statystyczne pomiędzy rodzicami w zapotrzebowaniu na wsparcie

Scale of Social Support	Parents of children suffering from EB (N = 21)		Parents of healthy children (N = 22)		Value of t	Value of p
	Mean	SD	Mean	SD		
Support from family						
General result	50.90	13.39	39.81	11.75	-2.891	0.006
Information support	11.95	4.15	10.45	3.60	-1.264	0.213
Instrumental support	14.28	3.93	10.81	3.93	-2.887	0.006
Appraisal support	14.23	4.13	10.68	2.74	-3.336	0.002
Emotional support	10.42	4.26	7.86	3.39	-2.187	0.034
Support from medical institutions						
General result	60.85	9.40	65.27	12.98	1.272	0.211
Information support	13.28	3.80	14.09	4.63	0.620	0.538
Instrumental support	17.33	3.05	18.95	3.30	1.669	0.103
Appraisal support	16.90	3.63	16.50	2.77	-0.412	0.683
Emotional support	13.33	4.38	15.72	4.36	1.793	0.080

Note: scores for general: 24–120; scores for subscales: 6–30. A lower score means higher need for support.

The studied groups of parents do not differ in the area of the need for support from medical institutions apart from the emotional one (but it is only a tendency level). But it is worth noting that the need for this support is a little higher in the case of parents of ill children.

The indicators representing expectations from medical institutions are higher than expectations from family, both for parents of ill and healthy children. It means that parents are quite satisfied with the medical support they receive. What is more, the

parents of children with EB have significantly higher need for support from family than the parents of healthy children.

It was also tested what correlates of life satisfaction are present in both groups of parents (Table 3).

It appears that a relation between life satisfaction and the need for support from family is characteristic for the parents of ill children, both generally and in the information dimension, and to a lesser degree with instrumental and appraisal support in which statistical tendencies were observed and also with

Table 3. Correlations between life satisfaction, and life attitudes among two groups of parents**Tabela 3.** Korelacja pomiędzy zadowoleniem i podejściem do życia w obu grupach rodziców

Variable		Life satisfaction parents of children with EB		Life satisfaction parents of healthy children	
		<i>r</i>	<i>p</i>	<i>r</i>	<i>p</i>
The need for support – family	General	-0.504	0.020	0.126	0.576
	Information	-0.620	0.003	0.046	0.839
	Instrumental	-0.410	0.065	0.222	0.321
	Appraisal	-0.422	0.057	-0.250	0.261
	Emotional	-0.192	0.405	0.332	0.131
The need for support – institution	General	-0.120	0.603	-0.066	0.769
	Information	-0.210	0.360	-0.111	0.622
	Instrumental	-0.369	0.100	0.082	0.716
	Appraisal	0.093	0.688	-0.048	0.833
	Emotional	0.104	0.654	-0.111	0.623
Life attitudes	Life purpose	0.659	0.001	0.668	0.001
	Coherence	0.647	0.002	0.695	< 0.001
	Life control	0.713	< 0.001	0.486	0.022
	Death acceptance	0.248	0.279	0.011	0.962
	Existential vacuum	-0.032	0.889	-0.417	0.053
	Goal seeking	0.485	0.026	0.155	0.492
	Personal meaning	0.646	0.002	0.746	< 0.001
	Life attitude balance	0.620	0.003	0.510	0.015

the dimension of goal seeking in the area of life attitudes.

In the last step a method of stepwise regression was applied which allowed estimation of the model explaining life satisfaction of parents of ill children to the highest degree. As the model variables, both the analysed psychological dimensions (support and life attitudes) and the demographic variables were used. The results indicated that the perceived life satisfaction depends mostly on the sense of life control, the amount of information support from family and the

course and symptom intensification of EB. The statistics of the model are presented in Table 4.

The model with significant values consists of three variables: life control, information support from family and clinical course of EB in a child. All of the variables in total account for around 70.7% of the variability of life satisfaction. *F* for the model is 13.694 ($p < 0.001$).

Regarding the parents of healthy children, the analysis of regression with stepwise method did not allow separation of the variables explaining the level

Table 4. Regression model for life satisfaction of parents of child with EB**Tabela 4.** Model regresji dla zadowolenia z życia rodziców dziecka z EB

Variable	<i>R</i> ²	<i>R</i> ² change	<i>F</i> change	Significance of <i>F</i> change
(Constant)				
Life control	0.508	0.508	19.629	0.000
Information support – family	0.617	0.109	5.118	0.036
Course of EB in a child	0.707	0.090	5.244	0.035

of life satisfaction, which means that this is a variable specifically understood in different life situations.

As a supplement to the questionnaire study, a short interview with the studied parents was conducted concerning the way they perceive their life. The categorised statements of the parents from both groups are presented in Table 5.

DISCUSSION

Having a child suffering from epidermolysis bullosa significantly changes the way the parents live. Most of all, the level of life satisfaction is significantly lower, though it still remains within the av-

erage range for the Polish population [19]. The data presented in Table 1 support hypothesis H1, that the child's incurable disease leads to a decrease of parents' life satisfaction, and to difficulties in defining their life attitudes. Most (15 out of 21 people) parents of a child with EB, when asked directly if they were satisfied with their life, confirmed that, though in the group of healthy people almost everyone was satisfied with their life (see Table 5). It is also worth noting that a large number of parents of ill children are not able to give any reason for life satisfaction and the total number of positive answers among them is almost three times smaller than the number given by the control group. This suggests that the declared

Table 5. Comparison of life assessment for parents of ill and healthy children

Tabela 5. Porównanie samooceny dotyczącej satysfakcji z życia rodziców dzieci chorych i zdrowych

Question and categorised answers			EB N = 21	Healthy N = 22	
<i>Are you satisfied with your life?</i>					
		<i>Why?</i>			
	EB	healthy			
Yes	15	21	Positive: family, work, self-realisation, support	12	25
			Positive: health, home, financial stability, friendship	3	16
No	3	0	Negative: stress, lack of self-realisation, hard life, financial problems	4	0
I don't know	3	1			
Total				19	41
<i>Do you feel you realise yourself in life?</i>					
		<i>Why?</i>			
	EB	healthy			
Yes	9	20	Positive (professional and personal self-realisation)	4	11
			Negative (lack of professional and personal self-realisation)	4	2
No	7	1			
I don't know	5	1			
Total				8	13
<i>What is the hardest in life?</i>					
Everyday life (lack of time, raising children, supporting family, occupational and financial stability, everyday decisions)			4	10	
Own habits, character, fears			3	8	
Lack of acceptance from others and a feeling of helplessness			2	7	$\chi^2(3) = 18.29$ $p < 0.001$
Child's illness			1	0	
Total			20	25	
<i>What is most important in life?</i>					
Family			14	20	
Health			4	12	$\chi^2(2) = 1.57$ $p = 0.454$
Other			10	13	
Total			28	45	

Note: the numerical value is given, not the percentage. The numbers do not add up to N – people studied, everyone could give any number of answers.

satisfaction measured by Satisfaction With Life Scale (SWLS) (which remains at a relatively high level varying around the average) is a kind of external rationalisation; for this declaration is not followed by the internal belief of its authenticity. When asked about the greatest difficulties which they experience in life, the parents indicated everyday, small troubles concerning life or their own character, though in the control group there were over two times more of these answers than in the group with EB. The greatest difficulty for the parents of ill children is definitely the disease of a child and coping with it (see Table 5). This is consistent with Lansdown *et al.* [11], and also Heringhaus *et al.* [16]. The significantly lower life satisfaction, in comparison to the control group, is probably also determined by the necessity for limitations to be set due to the child's disease. These limitations make self-realisation of a parent difficult or simply impossible. While in the control group almost all parents declare a feeling of self-realisation by achieving professional and personal goals (see Table 5), in the group with EB only nine people declared that; 7 people feel a complete lack of self-realisation and 5 people are not able to define it, which is also an indirect indicator of the lack of self-realisation. Similar results were presented by Tabolli *et al.* [10].

The difficulties and limitations which are the consequences of the incurable disease of a child require a support system to help effectively cope with it. In the present study, support from two sources was analysed: from family and from medical institutions.

The support from institutions is important because, in the beginning, it enables one to adapt to the requirements of a specific disease, and later it gives a feeling that there is access to professional care in sudden, unclear and difficult situations. The need for support was studied in four dimensions: emotional (understanding emotions, calming), information (full information concerning the situation, dealing with it, possible course and ways to cope with it), instrumental (expressed by providing access to all possible institutions, medicines, support groups, etc.) and appraisal (expressed by the need for feedback information from others giving faith in own capabilities, effectiveness, resourcefulness, etc.).

We decided to study both groups of parents because of the fact that healthy (meaning not chronically ill) children also use healthcare from time to time. At the same time, the level of healthcare was tested in the assessment especially of parents of children with EB, taking into account the numerous difficulties in financing treatment, difficulties in accessing specialists, and also a rare disease and limited number of doctors and nurses who specialise in this type of disease.

The obtained results, presented in Table 2, support hypothesis H2, the child's incurable disease is associated with a greater need for support from the family, and reject hypothesis H3, The child's incurable disease is associated with a greater need for support from medical institutions. The results, quite univocally, indicate that both parents of healthy and of ill children have a similar level of need for support. The general level of the need for support from medical institutions is average, as well as of appraisal and information support (Table 2). It means that the doctors and medical staff give similar attention to patients despite the severity of the disease. The parents obviously notice some inconveniences and imperceptions of some of their needs, but it is most likely a result of the functions and capabilities of the healthcare system as a whole. The parents of severely ill children receive more emotional support than the parents of healthy children ($t = 1.793$; $p = 0.080$), which probably results from a little greater empathy and concern from medical staff caused by the incurable disease of a small child. The greatest need concerns instrumental support, and it is even a little higher for the parents of healthy children. This can be confirmed by the fact of specific difficulties in the access to medicines and expert diagnosis, while in the case of children with EB the vital problem is acquiring e.g. specialist dressings. However, it is worth noting that some researchers [18] have identified, in their full-scale interviews with the parents of children suffering from EB, several distinct needs that should be provided for by the medical clinics such as information about diagnosis, treatment or a possibility of making decisions concerning the child. The dependency on professionals while taking care of an ill child is similarly indicated by other authors [22–24]. Polish parents did not explicitly indicate a greater need for support from medical institutions. It is possible that these differences are due to different functioning of healthcare between countries.

The second kind of support examined is a support from family. It was studied in the same four dimensions, in both groups of parents. It is much more fulfilled than the support from medical institutions (general result for the parents of children with EB = 50.9). In this case there were significant differences revealed between the parents of healthy and of ill children in all dimensions apart from information support ($t = -1.264$; $p = 0.213$) and, simultaneously, it is worth noting that the parents of healthy children expect very little from family (general result 39.8 with a minimal result of 24).

Emotional support is fulfilled to the highest extent, though the parents of children with EB need it significantly more ($t = -2.187$; $p = 0.034$) than the parents of healthy children (whose needs are almost

completely fulfilled). In the case of appraisal support ($t = -3.336$; $p = 0.002$) and instrumental support ($t = -2.887$; $p = 0.006$) the parents of ill children have greater expectations than the parents from the control group. This means that in regard of raising a child suffering from a serious illness, it is harder for the family to offer the same amount of support as for the parents of healthy children. This partly results from a lack of competence, and personal difficulties in taking care of an ill child which would allow e.g. to ease a parent (instrumental support). The parents of children with EB are generally strongly involved in the care of their own child. Some researchers have pointed out that even professionals who specialise in EB report a significant decrease in their quality of life due to strong involvement and working with limited resources. Due to this problem these professionals require supervision and general recognition of their work as valuable [25]. The parents of children with EB do not receive such support; they take care of their child full-time and feel very tired, which results in increased need for support from family (Table 2). Also they are expected to give their children support for social and emotional life, because of some problems in these areas [26]. It is one more task to do, with no support for parents. What is more, as other studies show [17], having a child suffering from EB has a strong negative effect on the family's quality of life, including quality of the marriage, which along with the severity of the disease grows worse and leads to a divorce. Taboli *et al.* [8] also indicate that the family problems grow bigger along with the noticeable intensification of the disease and the amount of body surface area affected by it. Family problems are connected with less support from loved ones. It should also be remembered that the parents of children with EB perceive the support they get differently [12], and they sometimes see it as a source of stress. The above data are consistent with the results obtained for the model of life satisfaction (Table 4), in which also the information support from family and the severity of the child's disease were found to be variables of significant influence on the sense of life satisfaction.

It was also tested whether giving birth and raising a child suffering from a severe, incurable disease is associated with differentiation of attitudes toward life. The results of the study univocally indicate greater difficulties in the area of life attitudes for the parents of children with EB (Table 1). Their life goals are more vague ($t = 2.562$; $p = 0.014$), which is also confirmed by the conviction of a lack of self-realisation capabilities and life satisfaction (see above), whereas their goals are often connected with child care. The unpredictability of the disease, temporary aggravations, and uncertain prognoses intensify a feeling of less control over one's life ($t = 2.060$; $p = 0.046$),

which becomes dependant on the current condition of the child. This makes not only planning everyday activities and holidays but also planning professional career and making long-term decisions concerning family life much more difficult. The situation in life is also associated with the fact that the feeling of personal sense and life attitude balance are significantly lower than in the control group too (respectively: $t = 2.275$; $p = 0.028$ and $t = 2.827$; $p = 0.007$). This, along with the increased sense of existential vacuum ($t = -2.753$; $p = 0.010$), reveals an image of a parent who sometimes lacks distance of one's life, who perceives it as quite chaotic, hard to control and at the same time giving little personal satisfaction. This leads to frustration which, combined with a generally hard situation of an ill child, makes full acceptance of one's existence difficult.

It should also be emphasised that despite the fact that life attitudes of the parents of children with EB are significantly weaker than the attitudes of parents of healthy children, which means that child's disease has a specific influence, the results obtained remain in the range of average results. So both groups of parents cope similarly, though in case of child's disease it is harder, which is reflected in significant differences presented in Table 3. These results support hypothesis H1. But it is difficult to compare our results to data from other studies because of the above-mentioned lack of publications concerning this problem in regard of epidermolysis bullosa. Such studies have never been conducted in Poland.

The difficulties in the area of life attitudes are also reflected in the relation between life satisfaction and other variables (Table 3). Therefore, regarding the parents of ill children, the increase in life satisfaction is significantly related to a smaller need for support from family in general ($r = -0.504$) and the information dimension ($r = -0.620$), whereas in the case of instrumental and appraisal support the correlation ratios reach as high as 0.4 but still remain only on a statistical tendency level. However, life satisfaction has no relation to the support from medical institutions. And regarding the parents of healthy children, life satisfaction has no relation to the support at all. This suggests the conclusion that unless the parents examined know that they receive as much support from medical institutions as they are able to obtain (taking into account the objective possibilities of care and treatment of a child with EB), they feel very tired with the constant care of an ill child and they expect support from the people who are closest to them, which would help them gain some distance from the situation. Information support provided, expressed in good communication, has the strongest correlation with perceived quality of life. These results support hypothesis H2 and lead to rejection of hypothesis H3.

The relations between life satisfaction and life attitudes are generally similar for both groups of parents (Table 3). The only difference is observed in the case of goal seeking, where the relation with life satisfaction is significant in the group of parents of ill children. This means that for the parents of these children it is satisfying enough that they can at least think of other possibilities, development or future plans which are not necessarily connected to the child's disease. If the number of such possibilities is smaller (which is indicated by the answers given during the interview - Table 5), the life satisfaction decreases.

The above deliberations, enabling us to establish a thesis on a direct influence of analysed variables on life satisfaction, allowed us to create an influence model based on a linear regression method and using the stepwise method. This model, explaining satisfaction, was estimated with this method only for the group of parents with children suffering from EB. Regarding the parents of healthy children, none of the analysed variables was included in the model with significant value. This means that life satisfaction in this group of parents results from other factors than previously assumed, not considered in this study. The presented model supports hypothesis H4: A model presenting the influence of the variables on the life satisfaction of the parents of ill children can be defined.

In the group with EB the model with significant values included three variables: life control, information support from family and clinical course of EB in a child (Table 4). All of the variables together explain 70.7% of the variability of life satisfaction. F for the model is 13.694 ($p < 0.001$). This means that a sense of life control is the most responsible for the life satisfaction of the parents dealing with the incurable and severe disease of a child. The clearly weaker life satisfaction in comparison with the parents of healthy children, expressed in a lack of self-realisation, focusing on everyday problems and submitting to the child's disease, is a consequence of a conviction that life cannot be controlled to a sufficient level. This conviction results mostly from the nature of the child's disease. Most of the studied parents had children suffering from dystrophic EB with a severe course. Such a situation increases the anxiety connected to the unpredictability of development and advancement of the disease which makes all plans concerning development, professional work and family life uncertain. This puts the parents in a worse situation and requires increased efforts to cope not only with tasks but also cognitively and emotionally. They need more support from the family, which also significantly affects the level of the declared life satisfaction.

CONCLUSIONS

The parents of children suffering from EB cope with the burden which is a disease. Their life attitudes remain in the range of average results, similarly as the level of life satisfaction. However, the observed differences in the area of these variables in comparison with the parents of healthy children indicate that the disease has an influence.

The need for support from family is increased. Support from medical institutions is generally assessed as positive, and these parents do not expect more than the parents of healthy children.

It seems that the greatest difficulty for the parents of ill children is the unpredictability of a life situation which limits plans and possibilities due to the unpredictable course of the disease. For the parents, this becomes a strong brake in the area of their self-realisation, which significantly lowers the level of life satisfaction.

The significant limitations of the study are the number of examined parents of ill children and the place of study. The rehabilitation clinic is a place of intensive child care and receiving large amounts of institutional support and, at the same time, it is a place for a parent to rest from everyday life. This situation could have significantly affected the emotional and cognitive change. Therefore, the obtained results should be treated carefully. However, because of the lack of studies on the parents of children suffering from EB (the nature of this disease is poorly extrapolatable to other childhood diseases, which makes referring to other results on this specific group impossible), even this type of preliminary study reveals specific problems which are experienced by parents.

Conflict of interest

The authors declare no conflict of interest.

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