

Parents' perception of challenges of caring of children with Epidermolysis Bullosa: A qualitative study

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ABSTRACT

Introduction: Epidermolysis bullosa (EB) severely affects the lives of children and their families; however, the severity of the disease, its dramatic effects on the lives of parents, and the challenges of parents of children with the disease have not been investigated. Therefore, the present study was conducted to investigate parents' experiences of children with EB.

Material and methods: This descriptive phenomenological qualitative study was conducted on 17 parents of children with EB from five hospitals affiliated with the University of Medical Sciences, west of Iran. Data was collected through semi-structured interviews. COLAIZZI's method was also used to analyze the data.

Results: Three main themes were extracted in this study: invasion of psychological crisis, alive but no living, and comprehensive support, including 10 categories.

Conclusion: The parents of these children are the target of psychological crises that severely reduce their quality of life. In this regard, they need extensive support to improve their living conditions. Therefore, it is suggested to provide cultural and institutional contexts in which all components of the children's dignity are emphasized.

Keyword: Parents, Challenge, Caring, Children, Epidermolysis Bullosa, Qualitative Study

Introduction

EB is an inherited disease of skin tissues [1], which causes changes in the skin such as thinning or blistering. Minor skin damage, such as warming, rubbing, and scratching, can cause blistering in people with the disease. In acute cases, even blisters can occur in various body organs, such as the mouth, esophagus, stomach, etc., severely affecting people's life [2].

⁷ The overall prevalence of EB in the United States is 11.1 per million live births, with an estimated ⁷ 1 in 51,000 live births. Others estimate the incidence of EB at ⁷ 1 in 20,000 live births, affecting approximately 30,000 in the United States [3]. In Iran, 950 EB patients have been identified so far [4]. This disease usually manifests itself in infancy or childhood. Although, in very mild cases, the disease improves with age, no cure has

been found for EB. Disease management primarily focuses on blister prevention, wound care, pain relief, early diagnosis and prevention of external complications (5, 6).

Caring for children with EB is very difficult and exhausting, affecting not only the life of the child but also the life of parents [7]. Despite the obvious needs of these patients and their caregivers and families, there are limited studies on the challenges of this population in providing health care. A study in Ireland examines the challenges of patients of children with EB. It states that one of the most important problems for these patients is access to appropriate care and treatment services in the community. It also highlighted the financial, emotional, and social challenges [8].

In their study, Van Scheppingen ¹⁷ et al. also stated that children with EB have reported many physical, psychological, and social challenges severely affecting their social interactions [9]. In their study, Donohoe et al. described the experiences of 10 to 14 year-olds with EB. On the other hand, due to the occurrence of this disease in childhood, caring for a baby with the disease at home is not easy. The children sometimes cannot move or play due to blisters on their palms and soles. As primary caregivers, parents experience psychological crises that severely affect their lives [10]. According to ²⁰ Van Scheppingen et al., parents of the children with EB had reported that people avoidance of them and their children [9]. The results of these studies highlighted the importance of paying attention to ¹² the needs and problems of these patients and their families. Meanwhile, despite the spread of the disease in Iran, the affected children and their families have received less attention, and the challenges of these parents in caring for and living with the children with EB have not been investigated. Although in recent years, ¹² studies have examined the life experience of parents with a sick and disabled child in Iran [11], the experiences of parents of

children with EB were not evaluated. Hence, according to the purpose of this study, a qualitative study with a phenomenological approach was used to explore the experiences of these parents. Therefore, the present study investigates the experiences and challenges of parents of children with EB. It is hoped that the results of this study will help managers and policymakers provide a supportive environment while protecting the rights of these children and their families.

Methods

Study design and research question

This descriptive phenomenological qualitative study was conducted in two centers affiliated with the University of Medical Sciences, using purposeful sampling from February to April 2022. Notably, purposeful sampling is a method used in a qualitative study to identify and select the individuals or groups of individuals who are especially knowledgeable about or experienced in a phenomenon [12,13].

Participants

The participants were 17 parents of children with EB, who selected from two centers. Inclusion criteria were parents of children with EB that their child is alive, at least 6 months have passed since the definitive diagnosis of EB in their children, having Iranian nationality, speaking Persian language, and being able to provide appropriate and sufficient information on the subject. There was an attempt to select the participants of this study with maximum variation; therefore, parents of children with EB were selected from a wide range of age, gender, financial status, education, household dimension, and living status. Exclusion criteria included having a known history of physical and mental illness and reluctance to continue the interview.

Data Collection

Data was collected through semi-structured interviews and field notes. The interview guide was prepared with the participation of all research team members. For this purpose, a meeting was held in the presence of all members of the research team in the office of the corresponding author, and the interview guide was prepared in the following steps: 1- Based on research objectives and the literature review, three key questions were initially considered for this study. Two interviews were conducted in the presence of all research team members to develop and test the questions. Based on the process of two interviews, the questions of the interview were developed. 2- Semi-structured interview approach was considered as a method of data collection. 3- Interviews should be done face-to-face. 4- By the rules of ethics in the research, the study's objectives should be explained to the participants and told that all their information would be kept confidential, and then written consent should be obtained from the participants. 5- Data should be analyzed by COLAIZZI's method. Therefore, data were collected through individual and semi-structured interviews. These interviews were conducted face-to-face in a quiet environment in the corresponding author's office with general questions. Then, follow-up questions were asked based on the answers (Table 1). The interviews lasted between 35 to 50 minutes and continued until they reached saturation. Saturation happens when new data do not appear and categories reach saturation regarding features and dimensions [14].

Data analysis

The collected data were analyzed according to COLAIZZI's method, which consist of seven steps. Step one; Interviews were listened to and transcribed, and each transcript was studied several times to gain a sense of the whole content. Step two; Identifying significant statements were extracted. Step three; Meanings were formulated from the significant statements. Step four; Organizing and categorizing

similar units.⁵; Presenting comprehensive descriptions of the extracted categories.

Step six; Creating, a basic paradigm of the subject under study according to the extracted categories. Step seven; Confirming, the basic paradigm by having the participants verify the categories and themes [15]. The data from this study were translated into English by native English.

² **Rigor**

Guba and Lincoln criteria were used to ensure reliability, including credibility, dependability, transferability, and conformability [16]. For this purpose and for increasing the acceptability and accuracy of data, data collection was done using semi-structured interviews, prolonged engagement, and data immersion. For reducing bias in data collecting were used member and peer checking. To this end, the extracted themes were submitted to 3 participants and 5 peers, who stated that the results were consistent with their interpretations.

Results

The present study interviewed 17 parents (10 mothers and 7 fathers) of children with EB. All parents participating in this study had only one child with EB. The mean age of the participants was 36.64 ± 2.74 years. Most of them had a diploma and were self-employed. In addition, most of the children in this study were suffering from dystrophic EB (DEB) and the mean age of these children was 7.35 ± 2.42 (Table 2). Three main themes were the invasion of psychological crisis, alive but no living, and comprehensive support, including 10 subthemes (Table 3).

1. Invasion of psychological crisis

The parents of the participants in the study stated that they had experienced severe psychological crises since the onset of the disease in their children. They saw prominent blisters, inflamed, red skin, and scary appearance of their children every

day and were unable to caress them. They reported disappointment and depression, maternal affection and hidden grief, fear of the future, and shock of euthanasia and child death are the most important psychological crises.

1.1 Disappointment and depression

14 participants stated that although they were looking for a solution for their child's disease, there was no cure, and they experienced living a life full of bitterness, disappointment, and depression.

'I was madly seeking treatment for my child. I was ready to do whatever I had ... but, I finally faced a great pain that this disease has no cure ... disappointment and depression predominated on my life' (participant 3, male 44years).

1.2 maternal affection and hidden grief

15 mothers stated that they see their children's pain, sorrow, and tears, but they have to hide their grief and comfort their child.

'My baby's appearance is very upsetting. In some parts, the skin on his hands and feet is peeled. She has great blisters on her skin. Other children do not play with her. I see the grief on her face ... she sometimes shed tears all daylong ... she says why the children do not love me I am a mother ... I am overwhelmed by motherly affection and have to hide my grief and comfort my child' (participant 7, female 48 years).

5 1.3 Fear of the Future

17 participants also stated that the fear of their children's future does not leave them alone for a moment. They were concerned about their child's health and, in milder cases, about their living, working, and marriage.

'My child's disease is not very severe. Sometimes he has skin blisters, but most of the time, he has inflamed skin. I am always worried about his health. Sometimes he has severe scaling. I am afraid of what will happen in the future. Like being expelled from

school ... even her marriage and future life worry me terribly. I am strangely afraid of the future'. (Participant 14, male 51 years).

1.4 The shock of Euthanasia and Child Death

One of the most important themes of psychological crisis in this study was the shock of euthanasia and child death. Participants stated that sometimes their children's health deteriorated, they had multiple infections, and they were hospitalized in intensive care units. Medical staff and relatives talked to them about the seriousness of the condition, euthanasia, and child death. While they had not yet recovered from the indescribable pain of their child's disease, they had to face the shock of euthanasia and child death.

'I still have not come to terms with the indescribable pain of my child's incurable disease ... I searched all websites every day to find a solution and treatment ... Suddenly, my child's condition worsened in a few months, and he was hospitalized. He is not conscious. And others talk to me about death and euthanasia ... I'm still seeking treatment ... How can I bear the shock of his imminent death' (participant 4, female 38 years)

2. Alive but not living

The important theme from the parents' perspective of children with EB was alive but no living children. They reported exhausting care, limited social interactions, and regret of a true smile.

2.1 Exhausting Care

⁹ From the perspective of the all parents participating in this study, exhausting care is the most obvious life experience of these parents with a child with EB. They have to spend a lot of time every day taking care of the child, including feeding, dressing, etc.

while enduring the terrifying appearance and serious physical condition of the child makes caring for them difficult and exhausting.

'My baby's appearance has become frightening with large tufts. It is hard and tedious for me to take care of him'. (participant 9, male 37 years)

2.2 Limited Social Interactions

Limited social interactions were ²¹ one of the most important themes in this study. 17 participants stated that their child's disease severely affected their social, family, and professional relationships, making them unable to communicate with others.

'Our communication with others, relatives, and the community are overshadowed by our child's disease. Sometimes, we do not want to get in touch with relatives and friends. Sometimes, we like to be in contact with someone, but they are afraid because of our child. They are worried that the disease is contagious, and they avoid us. ... Our social interactions are limited, and this adds to our grief "(participant17, female 28 years).

2.3 Regret of a True Smile

14 participants stated that their lives are full of regrets. They are even longing for a true smile. Although life is full of joys and sorrows, the sorrows have affected their lives much more than joys and socializing. Taking their child to school and watching them grow up has become their biggest regret.

'I am overwhelmed with regret ... life has good and bad things ... but the bitterness of this child who cannot even eat, run and play has made our lives like hell. If anything joyful happens and a smile comes to my lips, my baby's face comes to my mind, and the smile disappears from my lips' (participant 13, female 39 years).

3. Comprehensive Support

According to the participants, they need comprehensive support to deal with their existing conditions and improve their sick child's life quality. Accordingly, parents of children with EB must receive extensive information support from medical staff, emotional support from others, and financial support from charities.

3.1 Information Support

16 participants stated that they need extensive information support from medical staff to become well acquainted with the disease, and its progression to provide the best care, and take the most appropriate action to improve their child's life.

'This disease is very rare and unknown. Exact information about it and providing care for these children is unavailable. I needed basic and accurate information about the disease. Obviously, the medical staff is the best people who can guide me, and I follow their instructions in caring for my child safely and without fear' (participant 8, female 30 years).

3.2 Emotional Support

15 parents of children with EB have stated that widespread emotional support is one of the most important factors in enduring this painful and stressful life. The emotional support of friends and relatives could significantly improve these parents' endurance to adapt to the child's disease. Accordingly, emotional support was one of the most important themes extracted in this study.

'Living with a sick child, especially a child with EB is really hard and exhausting. I could not have survived without the 9 years of psychological and emotional support of those around me. My wife was by my side all these years. He cared for me impeccably. He comforted, wiped away my tears, hugged me, and talked to me to calm me down (participant 6, male 30 years).

3.3 Financial Support by Charities

16 parents also stated that caring for children with EB comes at a high cost. Sometimes, the costs of treatment impose high costs on families. Therefore, these parents need financial support from charitable associations to take care of their children.

'The costs for these children are very high. The cost of caring for, treating, and dressing this child is very terrible, especially in this economic situation of the country. We cannot afford it. The EB association and other charities are really needed to recognize this rare disease, the huge costs of caring for these patients, and help us pay for it' (Participant 11, female 27 years).

Discussion

In the present study, parents of children with EB reported experience of the invasion of psychological crisis, alive but not a life, and comprehensive support. Few studies have qualitatively examined the experiences and psychological crises of the parents of these patients as their caregivers. Therefore, the present study was conducted to investigate the experiences and challenges of parents of children with EB. In this regard, other studies assessing the experiences or emotional stresses of patients of children with EB were used. In this study, the experiences of parents of children with EB were investigated in three main themes: invasion of psychological crisis, Alive but not life, and comprehensive support.

The onset of psychological crisis was the first theme extracted in this study. The parents of children with EB sought a treatment. Eventually, they experienced disappointment and depression, maternal affection and hidden grief, fear of the future, the shock of euthanasia, and child death.

The rarity of EB has led many parents to seek a solution for their children. Although they have been hoping for a cure all this time, they have finally experienced only

depression and disappointment. Several other studies indicated that patients of these children reported frustration [17-20]. In this regard, Mauritz et al. (2021) stated that the parents of children with EB have experienced a lot of depression, sadness, and despair in caring for and finding a way to save their children [18]. Ireland et al. (2021) also stated that these parents reported pain, discomfort, and depression during the grueling care of their children, which severely reduced their resilience and ability to care for their children [17].

On the other hand, Tang et al. (2021) stated that patients with EB and their families have experienced a lot of suffering, depression, and despair, which has severely affected their quality of life [20]. The similarity in this finding indicates that apart from cultural, environmental, and economic differences in any society, the experience of having a sick child, especially with a severe and incurable disease, makes parents sad, despaired, and depressed.

However, the parents of children with EB, especially mothers, in the height of depression and despair, with motherly affection, hide their grief and appear in front of their children with a smile to reduce their pain and discomfort. They reported that when these children were overwhelmed with crying and restlessness due to not socializing with other children and being confined to the house, they had to hide their grief and comfort their child with a smile. Mauritz et al. (2021) stated that mothers of children with EB try to control their emotions in interacting with their children and comfort them with compassion [21]. Accordingly, emotion control has been reported as an important category of exploring the experience of these parents, which is consistent with the results of the present study. On the other hand, the parents participating in this study, especially children with mild EB expressed a strange fear of their children's future. They are worried about their child's health, work, emotional needs, and marriage.

Consistent with this study, other studies stated that caregivers, families, especially parents of children with EB are always afraid of their children's future and do not know what consequences this unknown disease will bear on their child's life [17,19,21].

The shock of euthanasia and child death was another category extracted in this study. The parents of these children, while not yet adapting to their children's illness, sometimes faced the worsening of the disease and their children being in the later stages of life. They described the shock of euthanasia and their children's death as the most painful experience of their lives. Active euthanasia is illegal in Iran because most of the Iranian people are Muslim and the country's laws are based on Islamic rules, but passive voluntary euthanasia has been noted in recent years. In passive voluntary euthanasia a patient, who is suffering from an incurable disease, or a first-degree relative of the decide patient not to continue the treatment. The suffering of the patient and the lack of hope for survival have caused patients and their families to think about passive euthanasia and think about which one is more suitable for the patient and brings the least suffering. Although the parents in this study stated that they had thought about euthanasia, none of them had attempted passive euthanasia of their children even with very bad conditions. This can be due to emotional relationships between parents and children or the religious beliefs of these parents. In this regard, Zeydi et al (2022) stated, although in recent years, this opposition was less than similar studies in Iran in the past but most of Iranian nurses in the intensive care unit had a negative attitude toward euthanasia for patients in the later stages of the disease [22].

Alive but not living, another theme in this study included three categories: exhausting care, limited social interactions, and regret of a true smile. Providing care for children with EB is difficult, time-consuming, and exhausting. Obviously, parents, especially

mothers, are the most important caregivers of children, and they spend a lot of time caring for and educating their children. However, mothers with sick children, especially with diseases such as EB, due to the physical condition of the child, experience exhaustive care. In this regard, Ireland et al. (2021) stated that caring for patients with EB is very tedious and time-consuming and severely affects the lives of all family members. Sangha et al. (2021) also stated that life with EB is very exhausting and puts a lot of mental and emotional stress on patients and their families, making life painful and difficult for them [17-23]. While being time-consuming and difficult, care severely limits parents' opportunities to interact with relatives, friends, and colleagues. Although, in most cases, these parents do not have the time and motivation for entertainment, sometimes those around them do not want to associate with these parents due to the child's illness. In this regard, several studies have reported limited social interactions between these patients and their families. According to Mauritz et al. (2019), mothers of children with EB do not tend to socialize due to the child's disease. Mauritz et al (2019) and Kearney et al. (2020) also stated that although child disease had severely limited their social interactions, they needed to expand their social interactions to promote their health and awareness [18,24]. Another category extracted in this study was a regret of a true smile. The parents said their children could not play, go to school, eat or dress like other children. Hence, these parents' lives are drowned in longing for a normal life. Accordingly, Sangha et al. (2021), Tang et al. (2021), and Mauritz et al. (2021) stated that patients with EB and their families had reported psychological stress and longing for a normal life [20,21,23].

Another theme in the study was comprehensive support. Emotional support from relatives and friends, information support from experienced medical staff, and financial support from charities help these parents to adapt more quickly to the situation. These

parents reported the information support of the medical staff was very helpful. When they get accurate information about the child's disease, its stages, and how to take care of them, they can go through the shocking stages of their child's disease faster and with full awareness. Meanwhile, Kearney et al. (2020) stated that patients with EB and their parents as the primary caregivers need information support and counselling by medical staff to improve their care [24]. Hammersen et al. (2021), Kearney (2020) and Martin et al. (2019) also stated that the information support of families, especially the parents of these children is essential to improve the quality of their life [24,26].

On the other hand, these parents reported that the emotional support of family and relatives facilitated of them to endure this suffering. Concurring with this study, Kearney et al. (2020) and Mauritz et al. (2019) stated that sympathy and empathy with patients with EB and their families is reducing on their mental hardships and they better adapt to their living conditions [18-24]. They need charities and grassroots organizations to help fund them. Many studies emphasize the importance and impact of financial support for these patients and their families. These studies show that financial support makes it more bearable for them to endure stressful conditions and, consequently, improves their quality of life [17,24].

Tang et al. (2021) stated that the cost of treating patients, especially patients requiring special wound dressings, is high. Annual total medical costs for these patients are in Ireland were \$84,534 and in Korea were \$7392. Also, annual costs in US for wound dressings ranging from \$4000 to \$245,000. Meanwhile, Iranians do not have access to many medical and dressing due to the embargo. They should buy drugs and dressing with huge cost. However, the increase in the number of patients with EB in recent years in Iran, the care needs of patients and their families have been taken into consideration [20].

Although there is the support association of patient with EB in several big cities of Iran, but there is no support association in other cities these patients. On the other hand, health insurances in Iran only cover part of the treatment costs, and the cost of medicines and dressings for these patients, which is imported from abroad, is very high and not covered by insurance. Therefore, these patients and their families in Iran need extensive financial support.

Finally, it can be said that the parents of children with EB experience many psychological crises, live a grieving life, and need extensive support to improve their living conditions. One of the limitations of this study was that the participants were only parents of children with EB. Clearly, the involvement of caregivers, teachers, and others who interacted closely with these children could broaden the generalizability of the results. Other limitations in this study were data collection using individual interviews and field notes. Other data collection methods, such as focus groups and observation, can enrich the results of this qualitative study. Therefore, it is suggested that future studies, in addition to individual interviews, use other data collection methods, including observation and focus groups.

Conclusion

Parents of children with EB experience many psychological crises, live a grieving life and need extensive support to improve their living conditions. In this regard, it seems that officials and policymakers can take basic steps to improve the quality of their life by using the results of this study and providing a suitable supportive environment for them.

Declarations

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Conflicts of interest/Competing interests No conflicts of interest for any of the authors of this manuscript, which would interfere with the integrity of this research, are reported.

Ethics approval: This study received ethics code from Hamadan University of Medical Sciences.IR.UMSHA.REC.1401.217.

Consent to participate: At the beginning of study, the researchers introduced themselves and explained the aim of the study. All the participants were informed that all information would remain confidential and that they could withdraw from the study at any time without any negative impact on their care in the future. Participants agreed to participate in the study. Written consent was obtained from participants to participate in the study.

Consent for publication: Participants were told that their information would be kept confidential and data would be published in the article without being named

Availability of data and material: The data obtained are clearly and comprehensively expressed in this study.

Code availability:

The authors have purchased SPSS software.

Authors' contributions

FM, SZM, MRS, FCH, SKH, MB were involved in the conception of the study and designed the study. They are responsible for data collection. Then S KH analyzed data. FM, MB drafted the primary manuscript.

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Table 2. Individual characteristics of the participants									
Part icip ants	Gen der	Age of paren ts	Total num ber of child ren	Type s of EB	Age of chil dren	Marital status	Duration of the child's illness (years)	Parents' educati on level	Parent s' Job

P1	Male	29	1	DEB	11	Married	2	Diploma	Governmental
P2	Female	32	3	EBS	10	Married	3	Diploma	Housewife
P3	Male	44	2	DEB	3	Married	4	Bachelor	Self-employed
P4	Female	38	2	DEB	6	Divorced	2	MSc	Housewife
P5	Female	26	2	JEB	4	Married	1	Diploma	Housewife
P6	Male	30	1	EBS	12	Married	3	MSc	Self-employed
P7	Female	48	1	DEB	8	Married	5	Diploma	Self-employed
P8	Female	30	2	EBS	3	Married	6	Diploma	Self-employed
P9	Male	37	2	JEB	8	Married	3	Bachelor	Governmental
P10	Female	41	3	EBS	9	Divorced	5	Bachelor	Self-employed
P11	Female	27	1	JEB	5	Married	6	Diploma	Governmental
P12	Male	41	2	EBS	7	Married	7	MSc	Governmental
P13	Female	28	1	DEB	13	Divorced	2	Diploma	Self-employed

P14	Male	51	2	JEB	10	Married	9	Bachelor	Governmental
P15	Female	40	1	DEB	4	Married	2	MSc	Self-employed
P16	Male	42	2	DEB	7	Married	5	Diploma	Self-employed
P17	Female	39	2	JEB	5	Divorced	8	Bachelor	Governmental
EBSimplex (EBS) Dystrophic EB(DEB) Junctional EB(JEB)									

Table3: Themes and Sub-themes extracted from content analysis

Invasion of psychological crisis	<ul style="list-style-type: none"> ▪ Disappointment and Depression ▪ Motherly affection and hidden grief ▪ Fear of the future ▪ Shock of euthanasia and child death
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Alive but not living	<ul style="list-style-type: none"> ▪ Exhausting care ▪ Limited social interactions ▪ Regret of true smile
Comprehensive support	<ul style="list-style-type: none"> ▪ Information Support ▪ Emotional support ▪ Financial support by charities

Table 1: *the main questions and their subsequent probing questions,*

<i>general questions</i>	<i>follow-up questions</i>
What is the nature of life with the children?	Can you explain further?
What are the challenges and concerns of family members and parents of children with epidermolysis bullosa?	Can you give an example? What do you mean by that?
What are the changes in the lives of family members and parents of children with epidermolysis blouse?	Can you explain further?