

Review

Caring for a child with Epidermolysis Bullosa: a scoping review on the family impacts and support needs

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Keywords Epidermolysis Bullosa, family, supportive care needs, parents, service delivery models

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Abstract

Aims Epidermolysis Bullosa (EB) is a rare genetic disorder characterised by recurrent skin blistering. Wound care and nursing are critical to everyday lives of EB patients. The aim of this review was to identify the support needs of parents of a child with EB and to assess the impact EB has on the family unit, irrespective of subtype of condition severity.

Methods We conducted a scoping review comprising 11 studies (2005–2021) to examine the research literature related to the support needs of parents with a child with EB, and the impact on family unit wellbeing.

Results Most common needs identified were emotional needs, followed by practical needs, social needs and physical needs. Many parents also reported a lack of informational and psychological support. Common findings

included emotional stress, lack of respite and physical strain on caring responsibility, financial stress, guilt and impact on relationships and family unit.

Conclusions Few studies exist that explore the support needs of parents of a child with EB. More attention should be paid to the support needs of parents to provide adequate care to those diagnosed with EB as well as their families.

Introduction

Epidermolysis Bullosa (EB) is a complex group of genetic disorders producing various degrees of recurrent skin blistering and epidermal detachment from the basement membrane in response to mechanical trauma¹. The latest consensus reclassification of inherited EB and other disorders with skin fragility describes major classical types of EB including EB simplex (EBS), junctional EB (JEB), dystrophic EB (DEB) and Kindler EB (KEB)². The clinical features for each of the four main types of EB are provided in Figure 1.

In addition to the four major types there are over 30 different subtypes of EB and varying degrees of severity ranging from mild local skin involvement with minimal impact on quality of life and overall longevity to severe forms involving multiple organs with early postnatal death or chronic progression and painful and life-threatening complications³. Chronic skin blistering commonly leads to the development of skin cancer, a major cause of death in severe forms of EB, hence significantly impacting a patient's quality of life and resulting in decreased life expectancy⁴. There is currently no cure for EB; however, concurrent advances in gene and stem cell therapies and significant focus on the design of symptom relief therapies over the last decade are converging towards multimodal combination therapies that hold the promise of clinically meaningful and lifelong improvement⁵.

The characteristics of severe EB demand higher than average levels of support compared to other children with chronic needs as disease management is focused on symptom

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Figure 1. The four main Epidermolysis Bullosa types and associated clinical features.

EBS: EB simplex with generalised blistering and superficial bullae over the body and blistering on the foot

JEB: Junctional EB with extensive shoulder blistering and vesiculobullous erosions and milia formation on the forearm

DEB: Dystrophic EB with severe scarring and pseudosyndactyly of the hands and atrophic areas on the lower extremities

KEB: Kindler Syndrome with poikiloderma on the hands and erosions on the back

Images provided by DEBRA

control and extensive wound care⁶. Wound management and nursing are an important aspect of support for families with a child with EB; however, many health professionals do not adequately understand the impact EB has on the individuals and the family unit. Numerous studies have described the needs of patients with EB and support from nurses, and qualitatively and quantitatively assessed the impact of disease on a patient's quality of life⁷⁻⁹. Additionally, many studies to date have acknowledged the impact of EB on caregiving parents, including the impacts on family size/planning and relationships¹⁰⁻¹³. The latest evidence-based psychosocial recommendations for the care of EB patients published in 2019 highlight the significant impacts the disease places on individuals with EB and their families, including all aspects of psychosocial life¹⁴. Additionally, this international guideline identified the research gap in better

understanding the support needs of parents caring for a child with EB and the impact on the family unit¹⁴.

Dystrophic Epidermolysis Bullosa Research Association (DEBRA) International is an umbrella patient advocacy and support organisation with member organisations in over 50 countries worldwide aimed to support best practices for EB care, increase professional knowledge of the condition, coordinate research efforts to better understand EB, find effective treatments and develop cures for all EB types. DEBRA groups in different countries have been critical in working with governments and healthcare systems to develop and implement the best models of care for patients and their families with EB, and implementing pivotal family support programs that aim to address the gap in the support offered by governments. However, there exists a wide difference in levels and range of support provided (for example, nursing support, access to dressings and medical and support needs, psychology programs, youth mental health programs, respite programs, family camps) for patients and their families with EB across different countries and sometimes even different regions or states in the same country¹⁵⁻¹⁷.

The objectives of the current review are three-fold:

- To identify the support needs of parents with a child with EB and the subsequent impacts EB has on the family unit irrespective of EB subtype of condition severity.
- To help inform future research on the impacts of EB on families, as well as help develop and revise future guidelines for care and development of an effective framework of support for patients and families with EB.
- To advance wound care practices by nurses and DEBRA groups in developing effective and tailored models of family support in their communities.

The novelty of this review includes development of the support care needs framework that can guide healthcare professionals (HCPs) in providing patient-centric support models for the person/child with EB and their family unit as a whole.

Methods

A scoping review was undertaken guided by the five-stage framework approach described by Arksey and O'Malley¹⁸, and subsequently enhanced and reinforced over the past few years¹⁹⁻²¹, in order to undertake a comprehensive literature search. The benefits of using the scoping review framework included: the ability to consider a broad array of literature including qualitative, quantitative and grey literature; and the capacity for a broad research question to be addressed in what is otherwise a limited area of focus within a specific rare disease^{18,20}. A systematic process was used to ensure rigour and transparency. The results are reported following the PRISMA-ScR guidelines²².

Research question

What are the support needs of parents caring for a child with Epidermolysis Bullosa (EB)?

Search strategy

It was agreed that the search for relevant literature would be limited to peer-reviewed published articles to adequately address the stated research question. Electronic databases searched were Medline, Embase, PsychInfo, Health and Society, Scopus, as well as the search engine Google Scholar.

Key search terms and phrases were Epidermolysis Bullosa* or EB Simplex or Junctional EB or Dystrophic EB or Kindler Syndrome and parents or mothers or fathers or caregivers or carers or family* and emotional or support* or burden* or impact* or need or care or wellness or wellbeing or cope or coping. Searches were limited to articles written in English between January 2005 through to March 2021. Review articles were excluded to minimise duplication of reporting. As we were focusing on the needs of parents, articles were excluded if they appeared to focus on caregivers other than parents (mothers and fathers); studies that were considered too medically focussed or psychologically obscure were also excluded. This resulted in 255 potential articles which were imported to the full text screening software Covidence® for a title and abstract review. Authors independently reviewed all articles.

Study selection

Title and abstract screening were undertaken by authors, including both review of all articles independently and selection to exclude or include respectively. If there was any conflict, for example one excluded and one included, then these were reviewed together, and a consensus was reached. Following final agreement on included articles, full texts were obtained and reviewed by all authors independently. Conflicts were reviewed as a team and the final decision to include or exclude was made. Remaining literature sources from each of the database searches were exported into the references and bibliographic management software program EndNote® for detailed examination and charting of relevant information to address the research question.

Charting of the data

The remaining full-text articles were then included in the extraction of information using a standardised data extraction form. The data extraction form included author, year, country, study aim, study design, method, sample size, main findings and critique. These were then reviewed by all authors for validation of domains and areas of support needs. Following this, all authors confirmed the support needs identified within each article and this was then added to the data extraction table as an additional column.

Results

A total of 255 articles were identified during the initial search;

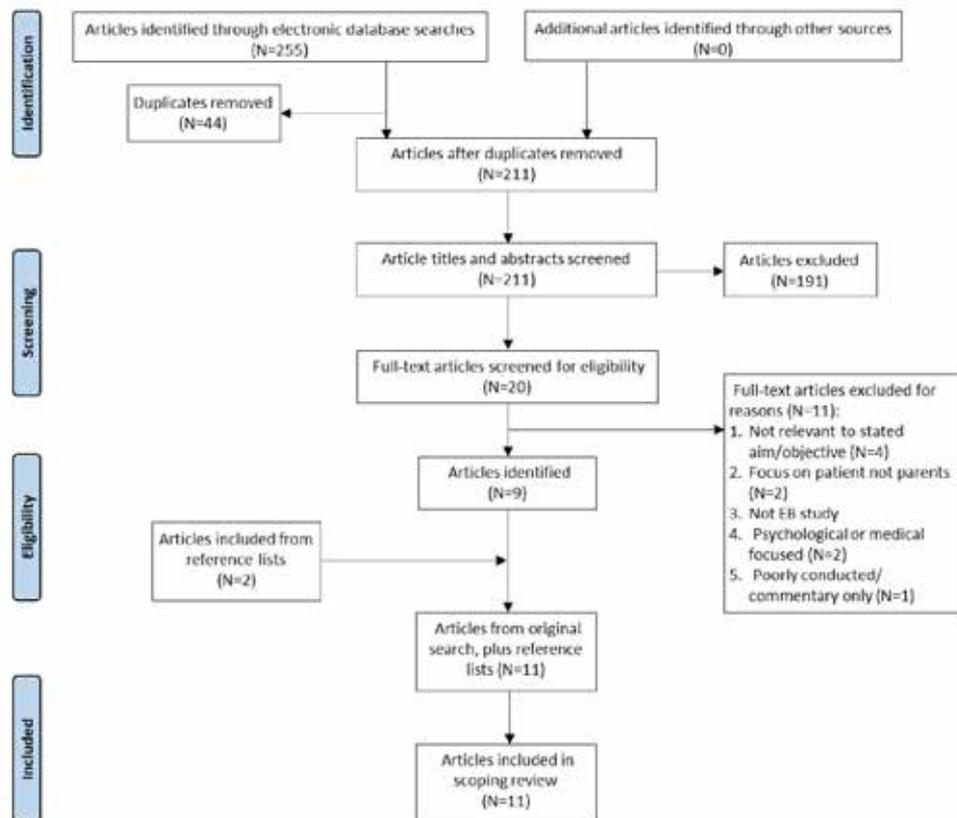


Figure 2. PRISMA flow diagram of search and selection

44 duplicates were removed. Following title and abstract and full-text screening, 11 studies were included in the final review, covering all subtypes of EB. Numbers of sources of evidence assessed for eligibility and included in this final review are summarised in Figure 2 in accordance with the PRISMA-ScR guidelines²².

Out of the 11 studies which met the inclusion criteria, six used qualitative methods (interviews), three used quantitative (questionnaires), and two used a combination of both to gather information. All included studies reported on the perceptions of support needs from a caregiver perspective. While seven studies also reported on the support needs of patients, these details were not included in the reporting of results of this scoping review. Eight reported studies were conducted by support organisations and local community groups, and three were conducted by hospital and university dermatology departments. Eight studies characterised findings based on support need perceptions as viewed by the mother or father of the child, one study mentioned the inclusion of a family which had a healthy child in the family unit, and one study reported findings of the siblings.

Six overarching support needs domains were identified – emotional needs, practical needs, social needs, physical needs, informational needs and psychological needs. The most common needs identified were emotional needs (n=10; 90%), followed by practical needs (n=8; 72%) and social needs (n=7; 63%) (Table 1). The majority of studies (n=10, 90%) reported on three or more supportive care need domains. Table 2 provides a synthesis of the domains of support needs discussed within the studies.

Emotional needs

Emotional needs were the most identified support needs and overwhelmingly encompass many support need domains for parents caring for a child with EB. Parents described the most common emotions associated with their child's EB to include frustration, embarrassment, worry, anxiousness and depression^{12,23-25}. Many parents expressed difficulties in accepting their child's condition and the daily burden that the disease demanded. The emotional impacts of the daily wound care routine place a significant toll on parents, mostly mothers, with feelings of powerlessness and sadness at seeing their child in pain and suffering²⁷. This leads parents to feelings of guilt about being responsible and causing pain to their child during dressing changes²³. The impacts of their child's disease on the family unit were also reported by parents as having a negative effect on personal relationships with their spouse/partner and other family members^{13,29}. The most significant ongoing emotional need reported by parents was feelings of guilt, with some parents also reporting guilt towards neglect of other siblings due to the ongoing care burden associated with looking after their child with EB²⁹.

The majority of parents reported a range of emotional stresses as a result of perceived inequality and lack of

consistency across both hospital- and community-based support healthcare services²⁴. In part, parents did not feel well supported by HCPs in addressing their emotional needs and would have valued a higher level of care and understanding from them in this area^{29,30}.

Compounding effects of emotional needs on parents caring for a child with EB were identified, influencing life choices and leading to a decreased life satisfaction; this was correlated with the severity of EB and stage of life (newborn vs childhood vs palliative care) and family support^{23,26,28}. Parents often reported the need for respite from caring responsibilities; however, a barrier to this was their emotional trust issues around relying on others to take carriage of their child's complex wound care regime in their absence²⁷. Fear of the unknown was also identified as a significant emotional need each time their child fell ill or during the period it took to confirm an infection and/or receive diagnosis/treatment; these were significantly traumatising for parents, adding to the ongoing concern about care burden²⁵. Interestingly, parents also reported the need for discussion around death and euthanasia of their child with EB, although they never initiated these discussions themselves³⁰.

Practical needs

The most commonly reported practical need identified by parents was that of financial concerns. Largely, this was associated with issues around employment and income/earning capacity due to the high burden of care associated with their child's disease. Some parents had to reduce working hours or terminate employment altogether in order to care for their child with EB²⁹. The impact on employment was more pronounced by mothers who were often the main caregivers; however, the financial implications of EB on the family as a whole were a significant source of stress for both parents as fathers had an added burden of being the sole provider for the family^{11,24}.

Compounding the parents' financial concerns was the expense of having to purchase their child's dressings; the protracted process they had to go through to acquire these was considered an unnecessary practical need for them²⁴⁻²⁶. Parents reported that wound care influenced all other daily life routines, making planning and organising family life extremely challenging²⁷. Additionally, having to coordinate and navigate local health services and appointments, and planning care for their child with EB was a constant source of stress and concern for parents²⁹. Parents reported constant challenges around not only the accessibility of wound care dressings but also in accessing local health services and this was further influenced by the provision of government-funded dressing schemes in certain countries or the metropolitan vs rural placement of the family^{24,25}.

Parents felt that, in order for others to care for their child in their absence, this would require a significant amount of training and trust for parents to feel comfortable leaving their

Table 1. Details of primary studies identified and reviewed

Author (year)	Country	Study aim	Study design/method/sample	Main findings/critique of a study	Support needs
Bruckner et al. (2020) ²³	United States of America	To understand the current manifestations and impact of EB from the perspectives of patients and caregivers and the difficulties for patients between the various subtypes.	Quantitative study; telephone survey; conducted by a third-party research firm; n=156 participants (63 patients, 93 caregivers); participants were recruited through two local EB patient advocacy organisations; ethics approval statement given.	Caregivers reported the most helpful sources of information were personal experiences, the patient community, family members and the EB specialist treatment centres. Many caregivers reported EB has had a profound impact on their quality of life and life choices. Reduced employment hours or quitting work altogether, resulting in a considerable financial burden for several parents. Information on reproductive family planning important. Impact of EB on partner relationships with nearly half of parent relationships ending in separation or divorce. Emotions reported include frustration, embarrassment, worry, anxiety and depression. No details of the process for the development of the survey or its validity/reliability. No statistical analysis was performed in reporting findings. Much of the impacts of EB are discussed from the perspective of patients, and the responses of caregivers are considered only as a proxy to patients.	Informational (disease, family planning), social (freedom/choices), practical (financial), emotional.
Fine et al. (2005) ¹³	United States of America	To assess the impact of EB in children on the personal relationships between their parents.	Quantitative study; questionnaire; administered via telephone interview; 109 items based from existing validated questionnaires, plus additional questions added; Likert style; participants recruited via the American-based National EB Registry (NEBR) and as part of a larger longitudinal study; n=374 (234 adults, 140 children); ethical approval reported.	Overall, parents of children with severely affected EB reported having greater relationship problems with their spouse/partner compared to parents with children with milder forms of EB. Many parents felt that they no longer had a private life. Parents stated that they communicated less like a couple and only really spoke about their EB child's needs. A negative impact on their sexual relationship and intimacy/closeness with one another was discussed. Around a third of parents said they were divorced, primarily due to their child's disease. A quarter-half of parents chose not to have more children because of their child with EB. Financial stressors were identified for some parents. And some said that their partners did not understand and or could not accept the daily burdens of their child's disease. Additional questions tested for validity but not reliability. Details including several parents who responded to the questionnaire not disclosed in the total numbers.	Social (relationship), practical (financial), psychological (reality)
Gowran et al. (2015) ²⁴	Ireland	To explore the experiences of key stakeholders including individuals with EB, their families, service providers, and policy developers currently.	Mixed methods study; qualitative (step 1) individual face-to-face interviews; lasting up to 1 hour; n=30 participants; qualitative (step 2) participatory action research workshop using an Soft Systems Approach (SSA); n=12 participants; quantitative; national survey; n=20 (11 parents, 3 were current parents, 6 patients); participants were recruited via DEBRA membership; ethics approval reported.	The majority of participants described feeling frustrated and emotionally challenged with the inequality and lack of consistency across the hospital- and community-based healthcare services and staff regarding communication, level of EB-specific expertise, lack of local services, and having to go through a protracted process to access medical supplies (e.g. bandages/dressings). Concern about the expense associated with having to purchase own dressings was discussed. HCPs need to understand the care burden on families and the time pressures placed on parents each day. Parents desire professional awareness by HCPs to consider life as an EB sufferer when undertaking tests/treatments. Details of the development and dissemination of the survey not provided. Questions not shown to be valid or reliable. Respondent demographics limited for the qualitative phase and not provided for quantitative. Results rarely differentiate between the views of parents, patients or healthcare workers.	Emotional, informational, practical (financial, accessibility)

Author (year)	Country	Study aim	Study design/method/sample	Main findings/critique of a study	Support needs
Kearney, Donohoe & McAuliffe (2020) ²⁵	Ireland	To identify the needs of the EB population to inform the development of a community EB outreach service aimed at supporting adults with EB and parents who are caring for children with EB.	Qualitative study; inductive exploratory using a participatory action research approach; semi-structured interviews; n=16 (8 mothers, 2 fathers, 6 adult patients); participants were recruited through local EB centres; ethics approval statement provided.	Physical care needs related to the disease identified by parents. Dressing changes and the time it takes to undertake treatment regime impacting. Parents value having an EB nurse present during the wound treatment as it helps relieve the physical and emotional burden on them. Need for more locally based care services was discussed. Some parents reported difficulties with having to travel significant distances to access community-based EB services. Others voice concern that their children might get sick when attending hospital appointments and would prefer to attend private services. Parental need for up-to-date practical information regarding their child's physical care needs and everyday aspects of living such as wound care, topical aids and skills. Parents fear the unknown, their child falling seriously ill and the time it takes to confirm infections is emotionally traumatising. Parents value informal support of family/others but are reluctant to leave their child in the care of others out of guilt, meaning they never get a break. Parents reported frustration with having to battle health service bureaucracy, a lack of empathy towards their child/situation, and unnecessary administration. Details of how the interviews were conducted were not provided. A well-conducted study overall.	Practical (wound care), social (accessibility, support), emotional, informational (family, disease), physical (services, respite).
Macik & Kowalska-Dąbrowska (2015) ²⁶	Poland	To determine if having a child suffering from EB influences the life and psychological wellbeing of parents.	Quantitative case-control study; standardised measures; Satisfaction With Life Scale (SWLS), Life Attitudes Profile – Revised (LAP-R) and Scale of Social Support; following short interview with participants; participants recruited through a rehabilitation camp organised by DEBRA / Kruchy Dotyk Society in Poland; n=21 parents of children with EB (13 mothers, 8 fathers) and n=22 parents of healthy children (without EB).	Not surprisingly, parents of children with EB reported greater difficulties in the areas of life attitudes, life satisfaction and the need for social support compared to parents of healthy children. Increased need for social and emotional support, especially from family members. Parents of both case and control groups were equally happy with the medical support received for their child. No details of the interviews conducted with participants. No ethical considerations were provided.	Social and emotional.
Mauritz et al. (2019) ²⁷	The Netherlands	To explore how adult patients and parents experienced the impact of wound care and examine coping strategies during childhood.	Qualitative study; individual semi-structured interviews; lasting 45–90 minutes each; participants recruited through the EB dermatology database; n=13 participants (7 patients, 6 mothers); ethics approval reported.	Physical and emotional impacts of wound care reported by all participants. Parents specifically reported wound care influenced all other daily routines. Mothers reported being more involved with wound care compared to fathers. Consequently, mothers made the decision not to work. The wound care process of preparation, performing and clean-up is tiring for parents, resulting in their need for respite/recovery afterward but was not often possible. Emotional impacts of wound care reported by parents include feelings of powerlessness and sadness. Seeing their child in pain further leads to feelings of suffering and distress. All parents acknowledged their need for respite, though some parents made the decision not to rely on others for wound care due to complexities and trust issues towards others. Fathers' voices were not represented in the study.	Physical, emotional, social, and practical.

Author (year)	Country	Study aim	Study design/method/sample	Main findings/critique of a study	Support needs
Sampogna et al. (2013) ¹¹	Italy	To evaluate the burden of recessive dystrophic EB (RDEB) on family caregivers, and to validate the FDLQI tool.	Quantitative study; postal surveys using two existing validated questionnaires; Family Strain Questionnaire (FSQ) and Family Dermatology Life Quality Index (FDLQI) which was translated and cross-culturally adapted to Italian; participants recruited from the EB databases of Dermopathy Institute of the Immaculate Conception (IDI-IRCCS) and DEBRA Italy; n=62 patients, n=59 caregivers of children with RDEB (50 mothers, 8 partners, 1 brother); ethical approval statement provided.	The most frequent problem reported by caregivers was time spent looking after the patient/relative. Emotional distress included affected physical wellbeing associated with disease severity and increased household expenditure. Family members reported a higher impairment on their quality of life when the patient/relative had psychological distress. The term 'caregiver' is used to mostly refer to mothers. A small sample of Italian patients with RDEB and caregivers. Study results primarily focused on the validation of the scale.	Physical (caring), emotional, practical (financial).
Tabolli et al. (2010) ²⁸	Italy	To assess the family burden of EB in children aged between 0–7 years.	Quantitative study; observational, cross-sectional; postal surveys based on three existing validated questionnaires; Patient Global Assessment (PGA), Family Strain Questionnaire (FSQ) and General Health Questionnaire (GHQ) plus a short demographic questionnaire; participants recruited through the EB database of the Dermopathy Institute of the Immaculate Conception (IDI-IRCCS); n=28 families (23 mothers, 5 fathers); ethics approval reported.	Most families reported needing more information about their child's disease. Reliance on family members to cope with caregiver burden was considered by many. The greatest emotional need reported by families was feeling worried for their child, with many relating this to the chronicity of the disease. Unsurprisingly, caregiver burden and the physical and emotional impacts of disease in families were associated with the severity of their child's EB. Small sample size with fathers under sampled.	Emotional, informational, physical.
van Scheppingen et al. (2008) ²⁹	The Netherlands	To identify and specify the problems experienced by parents caring for their child with EB.	Qualitative study; semi-structured interviews; lasting 1–1.5 hours; participants recruited from a university medical centre dermatology database; n=11 families (11 mothers, 5 fathers); ethics approval provided.	The visibility of their child's disease leads parents to experience public awkwardness, unkind remarks/questions by others. Lack of understanding of the disease causes others to act insensitively. Parents struggled emotionally seeing their child in pain, and guilt when they were the ones causing the pain while attending to wounds/blisters. Parents most expressed feelings of uncertainty and the unpredictability of their child's disease and the long-term health of their child. Fear of losing their child when their condition worsened. Mothers quit work to care for their child. Financial difficulties discussed. Impact on partner relationship, guilt about neglecting the other siblings. Challenges coordinating services/planning care discussed. Parents felt physically exhausted, need respite/leisure time. Health professionals lack knowledge and skills of condition. Parental need for more information. A well-conducted study.	Emotional, social, practical, physical, and informational.

Author (year)	Country	Study aim	Study design/method/sample	Main findings/critique of a study	Support needs
Wu, Sun & Lee (2020) ¹²	Taiwan	To explore the lived experience of family caregivers of EB patients.	Qualitative phenomenological study; in-depth face-to-face interviews lasting 90–120 minutes; followed by an informal in-home interview to observe caregivers; purposive sampling of participants aided by the Taiwan Foundation for Rare Disorder (TFRD); n=10 caregivers (7 mothers, 2 fathers, 1 grandmother); ethics approval reported.	Caregivers described emotional and mental impacts of stress, anxiety, overwhelmed, depression and helplessness. Physical burden leads caregivers to feel exhausted and mentally fatigued; the impacts considered greater for mothers. Impact of disease on employment leading to increased financial pressure. Social impacts include work-life balance, feeling embarrassed and humiliated when in public, and effects on members of the family relationships and, in some instances, causing relational breakdown. Some caregivers have guilt towards neglecting siblings due to ongoing care burden. A well-conducted and reported study.	Emotional, physical, practical (financial), and social.
Yuen, Duipmans & Jonkman (2012) ³⁰	The Netherlands	To identify the needs of parents who have lost a child to lethal EB within the past year.	Qualitative study; semi-structured telephone interviews; lasting 30–150 minutes; participants recruited from the Dutch Epidermolysis Bullosa Registry; n=16 (6 mothers, 5 fathers, 5 couples).	Parents felt that HCPs lacked the required knowledge and ability to properly care for their child with lethal EB causing injury and suffering. A lack of knowledge by HCPs also leads to incorrect/delayed referral to specialist care for some families. Parents expressed the need to make the ultimate decision for whether their child received hospital- or home-based care and treatment. Parents also desired to be actively involved in all aspects of care and decisions for their child; this enabled them to feel in control, have decreased feelings of helplessness, and adjust to the pending death of their child. Many parents expressed the need for discussions around death and dying and euthanasia, although most never initiated the discussions themselves. The reported number of participants is unclear. No ethical consideration statement was provided.	Informational, psychological, and emotional.

child in their care³⁰. Together, the practical needs of parents caring for their child with EB highlighted the need for better support services for parents to allow them much-needed respite and time free from their caring responsibilities.

Social needs

Parents most frequently voiced the need for more support in caring for their child with EB, particularly from family members. Issues with relationships and a lack of support from family members were reported by parents to have affected their quality of life and freedom of life choices^{13,23,26,29}. Access to a support group, such as DEBRA, was considered by parents as an important means of support and respite²⁷. There was an overarching feeling of embarrassment and humiliation when in public and a feeling that others did not fully understand or appreciate the impacts that their child’s EB has on them and on their family unit^{25,29}. These feelings were further compounded by the negative impact associated with a lack of work-life balance, adding additional strain on the family unit. This included effects on both partner relationship and relationships with other children where parents felt guilty about neglecting the other children^{12,29}.

The daily burden of care and missed opportunities to spend quality time together often had a negative impact, with one study reporting nearly half of all parent relationships ending in separation or divorce²³. Unsurprising, parents caring for a child with a severe form of EB reported having greater relationship problems with their spouse/partner compared with parents with children with milder EB forms. Many parents report not having an active social life. This further affected their communication/relationship with a partner which predominantly centred on either providing care or discussing the needs of the child with EB¹³. As a result, compounding impact also affected the sexual relationship and intimacy with each other, with a third of parents in one study saying they divorced primarily due to their child with EB while a quarter to half of parents in the same study reported not wanting to have more children as a result of their child with EB¹³.

Accessibility to support services was also an important factor affecting the social wellbeing of parents as some highlighted the need for more locally based services to access support and reported having to travel significant distances to access community-based EB services²⁵.

Physical needs

Caring for a child with EB often left parents feeling physically and emotionally fatigued^{25,27}. Parents reported this mainly related to all consuming aspects

Table 2. Summary of support needs

Study	Needs					
	Emotional	Informational	Physical	Practical	Psychological	Social
Bruckner et al. (2020) ²³	●	●		●		●
Fine et al. (2005) ¹³				●	●	●
Gowran et al. (2015) ²⁴	●	●		●		
Kearney, Donohoe & McAuliffe (2020) ²⁵	●	●	●	●		●
Macik & Kowalska-Dąbrowska (2015) ²⁶	●					●
Mauritz et al. (2019) ²⁷	●		●	●		●
Sampogna et al. (2013) ¹¹	●		●	●		
Tabolli et al. (2010) ²⁸	●	●	●			
van Scheppingen et al. (2008) ²⁹	●	●	●	●		●
Wu, Sun & Lee (2020) ¹²	●		●	●		●
Yuen, Duipmans & Jonkman (2012) ³⁰	●	●			●	

of providing wound care for their child with EB, the process of wound preparation, performing and cleaning-up following which was a very physically demanding responsibility, combined with the emotional aspects of seeing their child in pain and discomfort²⁷. The physical needs of parents caring for their child with EB also extended to general caregiving, leaving parents emotionally distressed which attributed to their physical needs, especially mothers who acknowledged that they largely assumed the primary carer role¹¹. Some parents had EB themselves and related the physical problems associated with it with that of their child, making caregiving even more physically challenging²⁸. Parents cited feeling overwhelmed, exhausted and mentally fatigued, highlighting the greater need for better community support services providing respite^{12,29}. They reported feeling as if they were never off duty, which exacerbated their exhaustion²⁹.

Informational needs

Parents considered the most helpful sources of information to include personal experiences of other parents, the patient community, family members, and accessing EB specialist centres^{23–25,28–30}. Information on reproductive family planning was an important aspect raised by some parents²³. Many parents describe feeling frustrated with inequality and lack of consistency across hospital- and community-based services, and information provided by HCPs, including lack of EB-specific expertise and knowledge among various health professionals^{24,25,29}. Parents identified a requirement for better support in providing up-to-date information on their child's physical care needs and everyday living aspects, including wound care, topical treatments and skills so that they could more adequately plan for the future²⁵. The need for better informational support was most evident in the groups of parents caring for a newborn where a definitive diagnosis

was pending, and parents caring for a child with lethal EB, resulting in delayed referrals to specialists in some cases³⁰. In addition, parents also commented that they required accurate information regarding disease progression and were not prepared for the suffering at the end of life³⁰.

Psychological needs

Psychological needs were discussed much less frequently in the studies compared to other domains, with only 18% of studies identifying parental psychological need as an important aspect of support for families with a child with EB. Psychological needs noted included parents' perception of reality and their struggle to understand or accept the daily burden of their child's disease¹³. The psychological needs of parents are related to the level of social and family support, relationship status and severity/progression of the child's disease³⁰. Many caregivers expressed that EB had a profound effect on their quality of life and life choices, including psychological/spiritual wellbeing²³. Two studies identified the need for better coping strategies to manage the disease burden, depression and mental health wellbeing of parents^{23,28}.

Discussion

This review of the literature is the first to comprehensively and systematically identify the documented support needs of parents caring for a child with EB, and to categorise these needs under six domains to identify areas of unmet need in parents.

It is important to acknowledge that support needs are dynamic and can alter over time because of factors such as the disease journey, the severity of disease, age and personal or environmental changes³¹. This was evident

from several studies included in this scoping review, which are not necessarily unique to EB, where parents reported that socioeconomic factors, patient age, severity/stage of EB and access to medical resources and supports all negatively impact on their support needs in caring for their child with EB^{23,24}. This is consistent with previous findings showing that support needs of parents and individuals living with a rare disease are dependent on the country/medical system³², while EB patient/family access to dressings and in-home nursing support can also be affected by families' metropolitan versus rural location^{15,16}.

From this review, we have identified that parents, irrespective of the severity of their child's EB, share common support needs. Aspects relating to emotional needs were identified by parents focusing on stress and guilt but also the uncertainty and fear of the unknown. Largely, the emotional needs of parents were directly related to their child's disease and the daily caregiver burden. However, it could be argued that emotional needs, if gone unmet, can influence other needs, including mental and psychological needs, often leading to the development of depression and mental health issues¹⁴. Therefore, facilitating the emotional support needs of parents caring for a child with EB should be an important consideration by HCPs. If parents are not coping emotionally, then their ability to care for their child with EB is challenged, placing themselves, their family unit and their sick child at further risk.

Interestingly, practical needs were the second most documented need in the current literature, with financial concerns being the most commonly identified aspect, as the cost of dressings in caring for the child with EB can place a significant burden on the family. The long-term sacrifice parents make in caring for their child with EB has been indicated in several studies as parenting a child with EB is a life-long commitment, including impact on work-life balance,

physical/mental exhaustion and feelings of isolation^{13,24,26,27,29}. These findings were in agreement with the recent report on the psychosocial recommendations for the care of children and adults with EB and their families published by DEBRA International¹⁴. All these factors were more predominantly observed in mothers of children with EB who are often the primary carers; data on fathers' support needs are not often reported to date.

Social, physical, emotional and psychological needs are often compounding and interrelated, while there is also a close relationship between the practical and informational needs of parents caring for a child with EB focused mainly on access to EB-specific services and planning at different stages of disease progression^{29,30}. This highlights the complex interrelationship of different support needs of parents caring for a child with EB, including the interplay between emotional, practical, social, physical and psychological needs.

Importantly, both emotional and informational needs identified highlighted that death, euthanasia and not being prepared for end of life are significant issues affecting parents of children with EB; this was in agreement with findings of the evidence-based psychosocial guidelines for care of children and adults with EB and their families¹⁴. Overall, the identified support needs are similar to those of other parents caring for a child with a rare disease where patient numbers are small, support is varied or lacking, and families are geographically scattered³². Importantly, the needs will vary vastly based on access to family support or in-home nursing care and the child's age¹⁵.

Based on the findings of the cited literature, we propose a novel parent support needs framework (Figure 3) which considers the complexity and interrelationship of the different support needs identified here. This is a suggested framework; however, more research in this area is needed to validate the domains of support needs and finalise the framework.

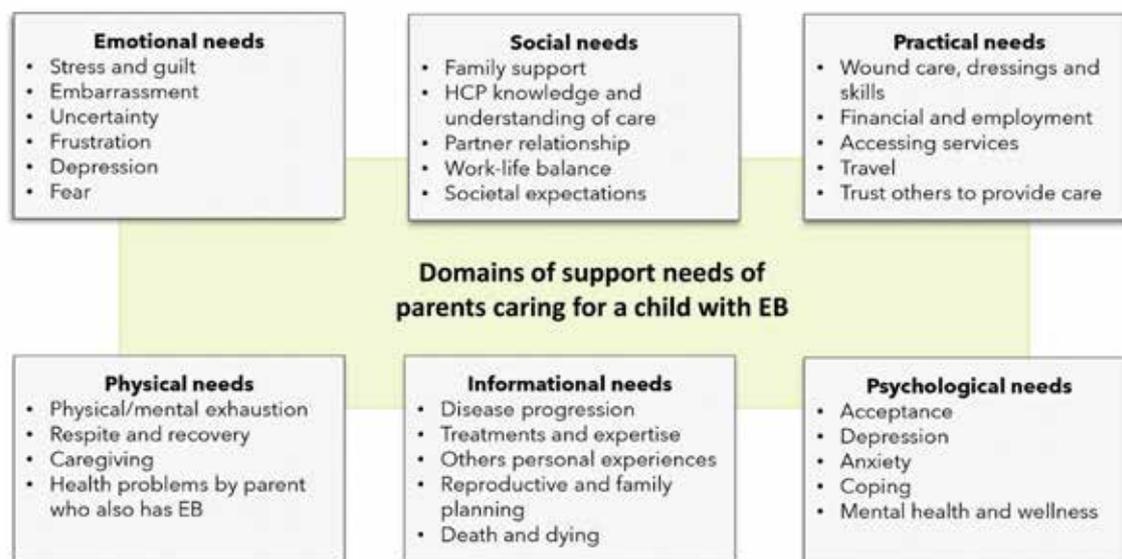


Figure 3. Proposed domains of parent support needs

Limitations of the review

The review followed the seminal scoping study framework proposed by Arksey and O'Malley¹⁸. A systematic process was used to ensure rigour and transparency following the PRISMA-ScR guidelines²². The authors engaged with each of the five stages of the scoping review process in an iterative manner and, where necessary, repeated steps to ensure the review of literature in this area was comprehensive. Nevertheless, there are some limitations with the current process that need to be acknowledged, including targeting the primary search to English language studies. We appreciate that there might be other published studies addressing the unmet support needs of parents caring for a child with EB in another language that were not included in this review. Importantly, to the topic of EB and support needs of parents, we also found that majority of studies reviewed the mothers' voices more prominently; this may reflect the fact that mothers are mainly the primary caregivers, and hence more likely to engage in studies exploring aspects of need in them. While some studies acknowledged this and outlined the need for a better understanding of the support needs of fathers, others did not address this and still used the word 'parent' in the title to reflect the views of mothers. We acknowledge that the support needs of parents are not homogeneous.

Implications for further research and practice

Despite reviewing a large timeframe (16 years), the paucity of studies in this review reflects the lack of focus on support needs of parents caring for a child with EB, especially fathers. This is not entirely surprising considering the rare nature of EB disease; however, the review emphasises the lack of evidence and highlights the need for future research on this important area. The development of new supportive care models that address the six domains of support needs identified in this review is urgently needed in the community to better support parents caring for a child with EB. Additional qualitative research and integrative reviews of the qualitative and quantitative literature is required to better understand the needs of families. Such studies will allow identification of the most important support needs of families with a child with EB, allowing the development of a multifaceted framework of support for these families.

Conclusion

Six domains of support needs were identified in this review with emotional, social and practical needs being the most common for parents caring for a child with EB. The caregiving burden is disproportionately affecting the mothers of children with EB, and fathers support needs are less identified, with parents' support needs not being homogeneous. We have proposed a novel parent support needs framework which could be used to guide future provisions of care for families. We hope that this review will help guide future research and development of support services in the community to allow support organisations like DEBRA and wound care HCPs to offer a better quality of life for parents and families with EB.

Author contributions

The authors confirm they had full access to the data in the study and take responsibility for the integrity and the accuracy of the data analysis. All the authors gave final approval of this version to be published and agree to be accountable for all aspects of the work in ensuring that the questions related to the accuracy or integrity of the work are appropriately investigated and resolved.

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Conflict of interest

Zlatko Kopecki is a Board member of DEBRA International and DEBRA Australia and a member of the Editorial Board of the *Wound Practice and Research Journal*.

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