



PSYCHOSOCIAL

Guidance for social and healthcare professionals/
epidermolysis bullosa (EB) teams

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Where does the information in this booklet come from?

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Conflict of Interest

Two authors declared potential conflicts of interest (Col): one an associate to DEBRA Norway and the DEBRA International executive team; the other guideline Coordinator for DEBRA International and project manager of the clinical practice guidelines (CPGs). These authors were not involved in the final recommendation manuscript editions post review panel feedback. Two reviewers declared a Col: one reported “to hold to different operating frameworks and professional competitors in a pool of diminishing mental health resources”; for the other, the guidelines directly relate to their work, PhD, and research.

Disclaimer

The information contained in this booklet does not indicate an exclusive course of action or serve as a standard of medical care. Variations, taking individual circumstances into account, may be appropriate. The authors of this booklet have made considerable effort to ensure that the information contained within accurately reflects the content of the guidelines on which it is based. The authors, DEBRA Norway, and DEBRA International accept no responsibility for any inaccuracies, information perceived as misleading, or the success of any recommendations, advice, or suggestions detailed in this booklet. The information provided on the following pages is subject to change without notice. For the most up-to-date information on available clinical practice guidelines, booklets, and contact information, please visit: www.debra-international.org

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CPG panel healthcare experts

- ▶ Kate Martin - CPG Chair, CPG healthcare guidance Lead (United Kingdom)
Consultant Clinical Psychologist, adults and families - Solihull Hospital
- ▶ Sam Geuens - CPG Co-chair (Belgium)
Clinical Psychologist, adults and paediatrics - University Hospitals Leuven
- ▶ Fiona Browne - CPG Member (Ireland)
Dermatologist - Our Lady's Children's Hospital Crumlin
- ▶ Annette Downe - CPG Member (United Kingdom)
EB Clinical Nurse Specialist - Guy's and St Thomas' Hospitals NHS Foundation Trust
- ▶ Nora García García - CPG Member (Spain)
Clinical Psychologist - DEBRA Spain
- ▶ Bronagh Kennedy - CPG Member (Ireland)
Senior Clinical Psychologist, paediatrics - Our Lady's Children's Hospital Crumlin
- ▶ Petra J Mauritz - CPG Member (Netherlands)
Child Psychologist - The University Medical Center Groningen
- ▶ Florence Pérez - CPG Member (Chile)
Clinical Psychologist - DEBRA Chile
- ▶ Kristina Soon - CPG Member (United Kingdom)
Senior Clinical Psychologist, paediatrics - Great Ormond Street Hospital
- ▶ Katty Mayre-Chilton - Project Manager/CPG Member (United Kingdom)
Specialist Adult Research Dietitian - Guy's and St Thomas' Hospitals NHS Foundation Trust

CPG panel people living with EB and PPI representatives

- ▶ Judith K Asche - CPG Member (Norway)
Person living with EB - DEBRA Norge
- ▶ Rebecca Bodan - CPG Member (United States)
Person living with EB - California State University
- ▶ Gemma Jaega - CPG Member (United Kingdom)
Person living with EB - Psychology graduate and person living with EBS, Liverpool

- ▶ Vlasta Zmazek - CPG Member (Croatia)
Mother of a person born with EB - DEBRA Croatia

CPG external review panel healthcare experts

- ▶ Kate Moss (United Kingdom)
Psychoanalytic psychotherapist with adults - Guy's and St Thomas' Hospitals NHS Foundation Trust
- ▶ Sondra Butterworth (United Kingdom)
Regional Community Support Manager - DEBRA UK
- ▶ Gabriele Gößnitzer (Austria)
Psychologist - DEBRA Austria
- ▶ Jacqui Finnigan (North Island, New Zealand)
EB Clinical Nurse Specialist - DEBRA New Zealand
- ▶ Sharon Cassidy (South Island, New Zealand)
EB Clinical Nurse Specialist - DEBRA New Zealand
- ▶ Kalsoom Begum (United Kingdom)
EB Clinical Nurse Specialist - Heart of England Foundation Trust, Solihull Hospital
- ▶ Alex King (United States)
EB Occupational therapy lead - Phoenix Children's Hospital, Arizona

CPG external review panel people living with EB and PPI representatives

- ▶ Matija Zmazek (Croatia)
Born with recessive dystrophic EB (1983 - 2018) - DEBRA Croatia

DEBRA International production team

- ▶ Katty Mayre-Chilton - CPG patient version project lead (United Kingdom)
CPG Coordinator - DEBRA International
- ▶ Olivia Mullins - Editorial and format lead (United Kingdom)
Business Manager - DEBRA International
- ▶ Michael Cardoza - Graphic designer (United Kingdom)



ABSTRACT	06
DEFINITIONS	07
PROVIDING PSYCHOSOCIAL SUPPORT	08
SUPPORTING DEVELOPMENT AND CONNECTIONS	10
ADVOCACY AND EDUCATION	12
PAIN MANAGEMENT AND WOUND CARE	15
PATIENT-PROFESSIONAL PARTNERSHIP	16
SELF-CARE AND TEAM SUPPORT	18
KEY RESEARCH PRIORITIES	21
GUIDELINE DEVELOPMENT METHOD	22
ABBREVIATIONS	23
CONTACT INFORMATION	24
NATIONAL DEBRA GROUPS	26

Epidermolysis bullosa (EB) is a group of rare genetic disorders resulting in skin fragility and other symptoms. EB is a complex disease impacting enormously on every aspect of psychosocial life. People and families living with EB need access to multidisciplinary (MDT) support, including psychological guidance, in order to improve quality of life and psychosocial well-being.

This booklet is a summary of the Martin et al. (2019) Psychosocial recommendations for the care of children and adults with epidermolysis bullosa and their family: evidence based guidelines. It provides recommendations for health care professionals to optimise psychosocial well-being in EB. An international multidisciplinary panel of social and healthcare professionals (HCPs) and people living with EB focussed on six different outcomes:

1. Quality of life (QoL)
2. Coping
3. Family
4. Well-being
5. Access to HCPs
6. Pain

Based on the evidence of appraised papers, recommendations were made for individuals living with EB, family and caregivers, and HCPs working in the field. This guidance booklet focusses on supporting and educating HCPs about the psychosocial complexity of EB and encourages them to work collaboratively with those around the individual with EB (e.g. schools, employers etc.) to provide psychosocial opportunity and care.

What is already known about this topic?

The physical needs of people with EB are often highlighted in research. The nature of living with EB is likely to present psychosocial challenges. The limited research in this area is divided across different specific domains of psychosocial care and focusses mainly on children with EB.

What does this guideline add?

People with EB and their families face psychosocial challenges. Attention should be paid to psychosocial impact as much as physical needs. Few studies have investigated which factors can lead to better outcomes. This guideline has outlined where further research could help in future care. Consensus is, however, that people with EB and their families should have access to multidisciplinary care, be supported to participate in daily life, and prevent isolation. EB-specific knowledge and support is needed for healthcare providers.

Key words

Epidermolysis Bullosa, Rare disease, Psychosocial, Psychology, Social care, Family, Healthcare professional.

The CPG panel agreed on the following published definitions to help focus their outcome searches as psychological health terms can be very broad and the concepts of QoL, coping, and well-being are complex with many interpretations and some overlap:

- ▶ **Psychosocial health:** the American Psychological Association (2018) defined this term as ‘describing the intersection and interaction of social, cultural and environmental influences on the mind and behaviour’.
- ▶ **QoL:** the WHO (2016) defined this term as an ‘individual’s perception of their position in life, in the context of the culture and value systems in which they live and in relation to their goals, expectations, standards and concerns. It is a broad ranging concept affected in a complex way by the person’s physical health, psychological state, level of independence, social relationships, personal beliefs and their relationship to salient features of their environment’.
- ▶ **Coping:** Lazarus et al. (1984) defined this term as ‘thoughts and behaviours that people use to respond to internal or external stressful demands’.
- ▶ **Well-being:** Dodge et al. (2012) defined this term as the ‘balance point between an individual’s resource pool and the challenges faced’.

Whilst some individuals living with EB will be able to successfully navigate their lives without psychosocial intervention or assistance from the healthcare team, many would likely benefit from assessment and intervention. Unfortunately, there was little in the way of guidance for clinicians regarding how to best care for the psychosocial health of patients and families affected by EB. Given the impact that psychosocial health can have on one’s physical health, this is a pressing need for those in the EB community.

The quality of the research which contributes to the recommendations is graded using SIGN (page 21).

Purpose of this guidance

This guidance provides information on the psychosocial support that professionals can offer patients with EB and their family. It is a summary of the main published CPG recommendations.

Guidance users

Social and healthcare professionals/EB teams who are working with patients who have EB and their families.

Target population of guidance

It is relevant for all types of EB: EB simplex (EBS), dystrophic EB (DEB), junctional EB (JEB), Kindler EB (KEB), and EB aquisita (EBA) - autoimmune, not genetic (there was no evidence found in EBA for the CPGs; however, it is assumed that they would require the same support). Other populations are the close family to people living with EB as well as social and healthcare professionals/EB teams.

It is strongly recommended that social and healthcare professionals/EB teams read the main CPG and the three patient versions (Support for adults living with EB, Support for parents caring for a child living with EB, and Support from your healthcare providers/EB team). These highlight specific psychosocial challenges of living with EB and ways to help.

“ I have lived experience of anxiety, low mood and Post-traumatic Stress Disorder due to the uncertain nature of EB and further diagnoses linked to EB – breathing, neuromuscular and ocular issues, previous physical health crises. The general daily restrictions of having a physical illness, such as difficulty finding employment, carrying out household chores and fighting for disability benefits are challenging. This leads to feelings of isolation, hopelessness, frustration and low self-esteem... as I’ve gotten older I’ve realised how fundamentally important it is for integrated physical and psychological healthcare. People often forget that the biggest organ of the human body is the skin, and naively underestimate the impact that the symptoms and limitations impose on the individual living with EB. ”

Gemma Jaega, living with recessive EB simplex, aged 28, United Kingdom

C

Recommendation

We strongly recommend psychosocial expertise to help people cope with EB. This includes having an awareness of the psychosocial impact of EB, asking about this specifically, and helping patients and their families to access support. It also includes supporting opportunities for psychosocial opportunities and development through advocacy, information giving, focussing on pain relief, and wound care.

EB has a profound impact socially, emotionally, and physically for the individual living with EB and their family. It can bring pressure around balancing care with roles, commitments, and relationships; and difficult feelings such as powerlessness or guilt. Parents face the challenge of being both the parent who wants to protect their child and the caregiver who has to inflict pain, which can take a toll on their well-being. For those with EB, the experience of living in pain, of enduring continued physical, social, and emotional challenges and uncertainties can have a profound impact on mental health. It is important to normalise mental health struggles as being part of enduring EB, not a reflection of any weakness or poor coping. The more we learn about mental health and psychological well-being, the more we see that physical and mental health are linked; it is therefore understandable that EB can be linked to difficulties such as anxiety, depression, and difficult psychological symptoms and experiences.

Professionals need to be confident in asking about the psychological and social impact of EB. Training on recognising and responding to psychosocial needs may be helpful. Whilst there are various measures of the psychosocial impact of EB and quality of life, these should not replace a conversation about coping with EB. It is important to integrate psychosocial care and monitoring into the routine care and conversations which professionals have with patients and their families.

Professionals need to recognise that the objective physical severity of a condition does not correlate with the psychosocial impact it can have. It is important not to assume that patients are okay because they physically seem okay. This can be the case, for example, where pain cannot be seen and in conditions such as EB simplex which can carry huge amounts of pain, disability, and psychosocial impact despite limited wounds.

Professionals can help by asking how things are going and explaining early on at initial contact, and at later points, what psychosocial support can be accessed. They could offer help with some of the practical tasks and paperwork or by helping to organise appointments that need to be attended. This includes the option to bring in outside caregivers or respite.

Research suggests that a diagnosis of EB can frequently impact on the family unit and link to the breakdown of relationships. Consider a referral for psychological assessment and intervention for individuals with EB and the family unit, should this be acceptable. Encourage and support families to have time together, for example, by helping families to access respite and outside carers. Some DEBRAs have recommendations and access to specialised holiday homes which are more accessible for people with EB.

SUPPORTING DEVELOPMENT AND CONNECTIONS

The reactions of family members to EB can be assimilated by the child. This then impacts later on in adulthood. Therefore, supporting family members, including the siblings of children with EB, is important. Encourage and support parents to talk to their child about EB and the impact this has. Children need to be given age-appropriate information to help them understand about EB and learn how to deal with it the best they can. Professional support, child booklets, or educative games can help. This information and understanding needs to be revisited in adulthood. Having the confidence to communicate about EB, and having the opportunity to ask for help and to be listened to and supported in response to this request can help patients feel supported in their psychosocial needs.



Artwork by Felipe Pliego Pontin, living with recessive dystrophic EB, aged 7, Brazil

Living with EB is more bearable when people feel supported and integrated in society. Participation in social life, peer support, and a sense of role and self-achievement help people with EB and their family to optimise their social well-being and coping. Professionals need to use their expertise to help with this since this can be really difficult for those living with EB.

Children with EB need to be encouraged to follow their psychosocial developmental milestones, alongside meeting their physical needs. Support is needed to develop a secure attachment to their parents; this is core to them understanding and regulating their emotions, trusting others, learning about relationships, and having confidence to explore the world around them.

As a healthcare professional, you may be able to advise on the physical “dos and don’ts” for people with EB, but it is also important to support individuals to experience new things. Through this process they are able to learn what their limitations are, to shape their own identity and develop a sense of independence, of having value, meaning, and a role. Those with EB should be supported to have a certain grade of mobility and independence: psychologically, socially, and, where possible, physically. There is always the challenge of supporting independence as much as possible whilst balancing physical needs and limitations. Encourage children and adults with EB to face challenges and goals step by step, to develop their interests and opinions, to discover what they are good at and to take time to enjoy the achievements and special times.

“ Very often as a parent I find he is much more capable than I even thought that he would be... it’s a balance between being supportive, encouraging them to experiment, having boundaries and discipline, and pushing your child to do the best they can. ”

Vlasta Zmazek, mother to Matija born with recessive dystrophic EB (1983 – 2018), Croatia

By providing advocacy and education about EB, focussing on pain management and wound care, you can help people overcome some of the challenges to social participation and forming relationships.



“ Despite constant pain he still plays football and is a goalkeeper for a local team. Eden also does Jujitsu and he has the most amazing football coach (Dan) and jujitsu Sensei (Luke). They have both embraced Eden’s condition and made sure they have a good understanding of it to enable him to continue to do what he loves. As parents it is heart breaking knowing he is in pain but also so reassuring knowing that he has the support to succeed! ”

Laura Hummerston, mother of Eden, living with EB simplex, aged 6, United Kingdom

There can be a lack of knowledge and understanding about EB in schools, working places, leisure time groups, or in peer relationships. Many social activities or public places are not sufficiently adapted for the needs of people with EB. Raising awareness of EB in general and the psychosocial needs of people with EB is essential. Providing family members with information can equip them to feel more confident in managing situations where EB is not understood.

“ EB is hard to explain. One day my son fell over and had taken the skin off his toe, I took off his sock and the people at his kindergarden saw it. They finally understood that EB is real. ”

Kari Anne Bø, mother of Adrian living with EB simplex Oгна, aged 3, Norway

You can help by supporting parents, carers, and people with EB to prepare for transitions and ‘new’ experiences, such as starting or changing school, employment, leisure groups, or new relationships. Embarking on new experiences can be less stressful if others are well informed about EB and how to help. The national DEBRA group can help with this, for example, in accessing information sheets. You could provide information about EB and physical needs, help them access medical supplies and equipment that can help, and help to think through some of the challenges, ways to problem solve, and to weigh options in decision-making.

Professionals may be in a position to advocate for the realities of EB on a wider scale. There needs to be a focus not only on the challenges and grief which EB brings, but also on the achievements and attributes it forges. The portrayals of EB in the media impact how people view EB and, for those living with EB, potentially themselves.

“ On the internet everything about EB is negative, always drama, videos of bandage changing and crying; this is not everything that EB is about. There should be useful information, more focus on what people with EB can do and achieve whilst recognising that it is extremely hard to manage EB. Think about us, those actually living with EB and the impact that such images and negativity can have. ”

Adriana Taguchi, mother to Mariana born with junctional EB with pyloric atresia (3 September – 11 November 2009), Brazil



Júlia da Costa Landim, who lives with EB, aged 6, Brazil (photo credit: Rodrigo Landim)



Physical discomfort (pain, itching, wound care, etc.) plays an important role in the limitations people with EB and their family experience. There is a CPG specifically on pain in EB. Although there is limited research on applying psychosocial techniques for managing pain to EB, techniques such as pacing and planning activities, relaxation, distraction, managing anxiety, tension, and cognitive attributions may be helpful.

Think about the care you routinely offer and the messages you are implying and teaching about EB. For example, with wound care it is important that children and adults with EB are involved in this process so they can learn and grow in confidence in managing their EB. The development of a fixed routine in a dedicated space, and a process of acknowledging feelings and pain but also of using techniques to make the experience more bearable can all help patients to recognise that wound care is one part of their life, not its entirety.

Using distractions and partaking in pleasurable activities, such as music, humour, watching a film, telling a story, singing songs together, and, for children, playing with toys all help with enduring the pain. It is important to involve other care providers in wound care at an early stage so that the person with EB can build trust in others tending to their wounds. For children, this use of carers allows the child and parent opportunity to function independently of each other, an essential step going into adulthood.

“ We can trace our gene all the way back to a small place in Norway in 1800. In my family it is normal to have EB. Coming from a family with a long history of EB is helpful, we can ask each other about wound care and they understand. In some ways it is more difficult because we don't talk about the impact of EB because it is a 'normal' part of our lives. It is important to be able to have someone to talk to about EB. ”

Kari Anne Bø, living with EB simplex Oigna, aged 36, Norway

Individuals with a chronic disease will have a lifelong relationship with their EB and the professionals supporting them. People with EB and their caregivers become experts on their own situation. A strong collaboration between the scientific based expertise of professionals and the experience based expertise of the person with EB is recommended. There should be a focus on both parties sharing what they can offer towards managing a situation. It can be hard for professionals to accept when their advice is not followed or dismissed, it is important to have space to process this and to continue to offer support whilst respecting individual decisions. It is also necessary not to assume that you have nothing to offer those people who seem to be managing their EB well. Sometimes having an 'outsider', who is not part of their family, to talk to about EB can be really valuable.

It is important to be transparent about limits to knowledge, treatments, and how sometimes there is no clear right direction or treatment. This, along with patients feeling listened to, understood and supported by professionals can help patients to build confidence and trust in their EB professionals. This lays the foundations for when difficult questions or choices need to be made, like treatment or surgery, or when difficult news has to be broken.

“ It is important to be in a partnership with professionals; we are both experts in EB and we work together. Nobody has all of the answers but we try to figure problems out together. ”

Judith Kristin Asche, living with junctional EB non-Herlitz, aged 57, Norway

C

Recommendation

We strongly recommend a collaborative patient-professional relationship. Training on working collaboratively with patients may be beneficial, especially given the complex dynamics which may present.



*Osmijeh ne košta ništa,
a djeluje čudesno.*



Matija Zmazek (right), born with recessive dystrophic EB (1983 – 2018), Croatia

Helping and treating patients with EB can be incredibly complex and difficult. The burden of EB can be hard to carry for healthcare professionals. Sometimes there are no answers or ways to help and the feeling of powerlessness is overwhelming. This is often difficult for the healthcare professional, the patient, and their family. Having access to adequate training is important so that clinicians can be confident in the care they can offer. It is also important to be aware of the limits and boundaries to professional responsibilities and capabilities, especially given the potentially long-term relationships between professionals, persons living with EB, and their families.

It is essential that healthcare professionals are aware of the need for self-care. They need support at an organisational level to take care of themselves in order to effectively be able to care for others. Self-care may consist of taking time for adequate reflective practice, supervision, managerial support, and making time to connect with those in their own support network outside of work.

Clinical supervision can provide a safe space to process your thoughts and emotions, to understand how these impact on your relationships with patients and colleagues and to share challenges and achievements.

It is necessary to be able to rely on colleagues in a MDT, to share support, learning and difficult decisions. It is important to plan and make time to meet as a team of professionals. For those who are working in isolation, try to connect with others in the same profession, a similar role, or who are working with EB: contacting DEBRA can help you to do this. Reaching out to international networks and forms for example through EB-CLINET can also support those working in isolation.

C

Recommendation

We strongly recommend offering support for professionals working in EB. Emotional support is necessary personally and on an organisational level, by building:

- ▶ an awareness of the importance of self-care
- ▶ support to do this and access to reflective clinical supervision
- ▶ work in isolation, time to link together as a multi-disciplinary team working together with patients
- ▶ feeling equipped through information and training to help with psychosocial needs.

“ I think it’s vital that HCPs look after their own mental wellbeing. Working with individuals who have a condition, such as EB, which causes a great deal of suffering and carries a high mortality rate, has the potential to cause HCPs long-term psychological distress. HCPs are only human, and even with advances in medical research not everybody can be saved. It is important for them to be reminded that they are doing the best they can with the resources that they have and that their tireless work, whether in clinic, the community, or at dermatology/rare disease/DEBRA conferences, doesn’t go unnoticed. ”

Gemma Jaega, living with recessive EB simplex, aged 28, United Kingdom

“ Having supportive colleagues who will always be available to listen or talk through any difficult situations is also very helpful. It is also important to be able to ‘switch off’ from work and have time to relax and enjoy time with family and friends. Getting a work/life balance is essential to continue in the work without ‘burning out’. ”

Annette Downe, EB Clinical Nurse Specialist, Guy’s and St Thomas’ NHS Foundation Trust, London, United Kingdom

“ Our nurse specialist team attend clinical supervision together rather than as individuals. This is really helpful and reassuring for me as I realise that they have the same concerns and worries. We are collectively able to find solutions to difficult situations and if we cannot find a solution at least we feel united as a team and are able to support each other. . . . I don’t realise how much the grief my patients suffer affects me until I attend our team clinical supervision sessions with our clinical psychologist. I always feel emotionally drained after these sessions. As nurses we are so used to keeping our emotions in check and carrying the emotional burden.... It is beneficial to discuss collectively the concerns, burdens and challenges that we face when caring for people with EB. As nurses we develop very close working relationships with the patients and their families This therapeutic relationship develops trust and openness, however, it can be costly when we give so much of ourselves physically and emotionally over time. ”

Catherine Nye, EB Clinical Nurse Specialist, Guy’s and St Thomas’ NHS Foundation Trust, London, United Kingdom



Existing research offers an understanding of the psychosocial challenges presented by EB, but not how to address or manage these effectively. High quality research is needed on:

- ▶ the efficacy of interventions to support and manage psychosocial needs
- ▶ the application of existing psychosocial interventions for difficulties such as pain management, anxiety, and social interaction to an EB specific population
- ▶ EB in adulthood and EB simplex are under-represented
- ▶ the rarity of EB leads to difficulties with small and heterogeneous samples: international collaborations may be an option.

SIGN grading system

Levels of Evidence		Grades of recommendation	
1	++	High-quality meta-analyses, systematic reviews of RCTs, or RCTs with a very low risk of bias	A At least one meta-analysis, systematic review or RCT rated as 1++, and directly applicable to target population; or A body of evidence consisting principally of studies rated as 1+, directly applicable to target population, and demonstrating overall consistency of results
	+	Well-conducted meta-analyses, systematic reviews, or RCTs with a low risk of bias	
	-	Meta-analyses, systematic reviews, or RCTs with a risk of bias	
2	++	High-quality systematic reviews of case control or cohort studies	B A body of evidence including studies rated as 2++, directly applicable to the target population, and demonstrating overall consistency of results; or Extrapolated evidence from studies rated as 2++
	+	High-quality case-control or cohort studies with a low risk of confounding or bias and a high probability that the relationship is casual	
	-	Case-control or cohort studies with a high risk of confounding or bias and a significant risk that the relationship is casual	
3		Non-analytic studies, e.g. case reports, case series	C A body of evidence including studies rated as 2+, directly applicable to the target population and demonstrating overall consistency of results; or Extrapolated evidence from studies rated as 2++
4		Expert opinions	D A body of evidence rated level 3 or 4; or Extrapolated evidence from studies rated as 2+
Good practice points			
			✓ Recommended best practice based on the clinical experience of the guideline development group

Source: SIGN 50 Guideline Developer's Handbook. NHS Scottish Intercollegiate Guidelines, Network 2014

- ▶ The international CPG development panel consisted of EB experts: psychologists, paediatric dermatologists, nurses, a dietitian, a methodologist, and patient representatives.
- ▶ All panel members completed written conflict of interest and code of conduct declarations.
- ▶ During guideline development, the group met twice in face-to-face meetings to discuss the clinical questions and methodology; review the evidence (publications in the field of EB psychosocial research); formulate recommendations; and agree on the guideline's structure and wording.
- ▶ CPGs are based on a critical, extensive, and exhaustive review of the most relevant publications in the field of EB psychosocial research together with the personal experiences of the panel members.
- ▶ To identify publications, 12 electronic search engines were accessed, including Medline (PubMed MeSH), Embase Emtree PsychInfo, CINHALL, Scopus, and the main search engine for the National Institute for Health and Care Excellence (NICE). Searches were performed using the terms “EB and Psychosocial”, “EB and psychologist” and “EB and social” in four different languages with the search period ending December 2018.
- ▶ A total of 601 articles were identified. Finally, 36 papers were appraised, each by two panel members, according to the Critical Appraisal Skills Programme (CASP) and Scottish Intercollegiate Guidelines Network (SIGN) quality rating.

APA	American Psychological Association
CASP	Critical Appraisal Skills Programme
CoI	Conflict of Interest
CPG(s)	Clinical practice guideline(s)
DEB	Dystrophic EB
EB	Epidermolysis bullosa
EBA	EB acquisita
EBS	EB simplex
HCP	Healthcare professional
JEB	Junctional EB
KEB	Kindler EB
MDT	Multidisciplinary team
NHS	National health service
NICE	National Institute for Health and Care Excellence
Orphanet J Rare Dis	Orphanet Journal of Rare Diseases
PPI	Patient and Public involvement
QoL	Quality of life
SIGN	Scottish Intercollegiate Guidelines Network
UK	United Kingdom
US	United States
WHO	World Health Organization

DEBRA International

DEBRA International is the umbrella organisation for a worldwide network of national groups that work on behalf of those affected by the rare genetic skin blistering condition, epidermolysis bullosa (EB). The first DEBRA was created over 40 years ago and is present in over 50 countries around the world.

www.debra-international.org
office@debra-international.org

EB Without Borders

EB Without Borders is a key initiative of DEBRA International. Its mission is to help patients, families, and doctors in countries where there is no DEBRA structure to support them, and to assist new groups to form and develop.

ebwb@debra-international.org

EB-CLINET

EB-CLINET is the global clinical network of EB centres and experts.

www.eb-clinet.org
office@eb-clinet.org

**This is how life feels
to people with EB.**



NATIONAL DEBRA GROUPS



▶ **Argentina**
www.debraargentina.org
info@debraargentina.org
+54 (011) 3965 4298



▶ **Australia**
www.debra.org.au
admin@debra.org.au
+61 (0) 427 937 003



▶ **Austria**
www.debra-austria.org
office@debra-austria.org
+43 1 876 40 30



▶ **Belgium**
www.debra-belgium.org
info@debra-belgium.org
+32 (0) 4 267 54 86



▶ **Bosnia & Herzegovina**
www.debra.ba
djeca.leptiri@hotmail.com
+387 33 328 284



▶ **Brazil**
www.debrabrasil.com.br
web@debrabrasil.com.br
+55 (047) 3237 6243



▶ **Bulgaria**
www.debrabg.net
contact@debrabg.net
+359 (0) 882 919 167



▶ **Canada**
www.debracanada.org
debra@debracanada.org
+1 800 313 3012



▶ **Chile**
www.debrachile.cl
info@debrachile.cl
+56 2 22 28 67 25



▶ **China**
www.debra.org.cn
debra_china@163.com
+86 139 1850 3042



▶ **Colombia**
www.debracolombia.org
info@debracolombia.org
+57 1 62 365 09



▶ **Costa Rica**
www.debracr.org
info@debracr.org
+506 2244 3318



▶ **Croatia**
www.debra.hr
info@debra-croatia.com
+385 1 4555 200



▶ **Cuba**
debra_cuba@yahoo.com



▶ **Czech Republic**
www.debra-cz.org
info@debra-cz.org
+420 532 234 318



▶ **Finland**
www.allergia.fi/suomen-eb-yhdistys-ry
suomenebyhdistys.debrafinland@gmail.com
+358 9 473351



▶ **France**
www.debra.fr
contact@debra.fr
+33 (0)7 78 95 83 44



▶ **Germany**
www.ieb-debra.de
ieb@ieb-debra.de
+49 (0) 6461 9260887



▶ **Hungary**
www.debra.hu
posta@debra.hu
+36 1 266 0465



▶ **Ireland**
www.debraireland.org
info@debraireland.org
+353 1 412 6924



▶ **Italy**
www.debraitalia.com
info@debraitalia.com
+39 02 3984 3633



▶ **Italy (Südtirol - Alto Adige)**
www.debra.it
info@debra.it
+39 335 10 30 23 5



▶ **Japan**
www.debra-japan.com
info@debra.sakura.ne.jp
 +81 (0) 11 726 5170



▶ **Malaysia**
www.debramalaysia.blogspot.com
debramalaysia@gmail.com
 +60 12 391 3328



▶ **Mexico**
www.debra.org.mx
debramexico@gmail.com
 +81 8008 0352



▶ **Netherlands**
www.debra.nl
voorzitter@debra.nl
 +31 030 6569635



▶ **New Zealand**
www.debra.org.nz
debra@debra.org.nz
 +64 04 389 7316



▶ **Norway**
www.debra.no



▶ **Poland**
www.debra-kd.pl
sekretariat@debra-kd.pl
 +48 501 239 031



▶ **Romania**
www.minidebra.ro
contact@minidebra.ro
 +40 0763 141 326



▶ **Russia**
www.deti-bela.ru
info@deti-bela.ru
 +7 (495) 410 48 88



▶ **Serbia**
www.debra.org.rs
office@debra.org.rs
 +381 (0)64 308 6620



▶ **Singapore**
www.debrasingapore.com
debrasingapore@gmail.com
 +65 9237 5823



▶ **Slovakia**
www.debra-slovakia.org
debra.slovakia@gmail.com
 +421 940 566 667



▶ **Slovenia**
www.debra-slovenia.si
info@debra-slovenia.si
 +386 31 348 303



▶ **Spain**
www.debra.es
info@debra.es
 +34 952 816 434



▶ **Sweden**
www.ebforeningen.se
malin.ch.net@gmail.com
 +46 (0)703 146 048



▶ **Switzerland**
www.schmetterlingskinder.ch
debra@schmetterlingskinder.ch
 +41 62 534 16 90



▶ **Taiwan**
www.eb.org.tw
taiwaneb2010@gmail.com
 +886 03 5734493
 +886 0966 521836



▶ **Ukraine**
www.debra-ukraine.org
info@debra-ukraine.org
svitdebra@gmail.com
 +380 50 331 94 97



▶ **United Kingdom (UK)**
www.debra.org.uk
debra@debra.org.uk
 +44 (0)1344 771 961



▶ **United States of America (USA)**
www.debra.org
staff@debra.org
 +1 (212) 868 1573
 +1 (855) CURE-4-EB

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