debra International.

PSYCHOSOCIAL

Support from your EB team/healthcare providers

WHAT IS EPIDERMOLYSIS BULLOSA?

EB is a group of rare genetic disorders characterised by fragility of the skin and mucous membranes and mechanically induced blistering. EB comprises four main types - EB simplex (EBS), junctional EB (JEB), dystrophic EB (DEB), and Kindler EB (KEB), with more than 30 subtypes. EB is clinically heterogeneous including a broad spectrum of severity.

COVER PHOTO: Carmelita Kelly Vieira de Sousa, living with recessive dystrophic EB, aged 33, Brazil
(photo credit: Suelen Szymanski)

Paula de Almeida Alves Santos, living with dominant dystrophic EB, aged 27, Brazil (photo credit: Suelen Szymanski)

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Artwork by Deanna Molinaro, living with recessive dystrophic EB, aged 27, Canada (© d'innovation art, @_dinnovation)

What is psychosocial support?

Psychosocial support means helping to support psychological, social, emotional, and spiritual well-being. This is formed during a person's lifetime and is influenced by environmental factors as well as factors that are unique to each individual.

Who is this booklet for?

This booklet is for people living with any of the following types of EB or for parents of a child with any of the following types of EB:

- EB simplex (EBS)
- dystrophic EB (DEB)
- junctional EB (JEB)
- Kindler EB (KEB)
- EB acquisita (EBA) - autoimmune, not genetic*
  *There was no evidence found in this population group for the CPG. However, it is assumed that they would require the same support.

What is this booklet about?

This booklet is designed to explain how a range of healthcare professionals might be able to support you/your child in managing and/or coping with EB.

Topics covered in this booklet include:

- Who are the EB team/healthcare providers?
- What can the EB team/healthcare providers do?
- The role of the EB team/healthcare providers
- The importance of working together with the EB team/healthcare providers
- The importance of EB awareness education
- The importance of working on pain management together

Where does the information in this booklet come from?

The information and recommendations in this booklet are derived directly from the "Psychosocial recommendations for the care of children and adults with epidermolysis bullosa and their family: evidence-based guidelines". The guidelines were written in 2018 by a group of EB healthcare professionals, individuals living with EB and family members. The information and recommendations in the guidelines come from a variety of sources including clinical research and expert opinion.

There are two different types of recommendations in this booklet:

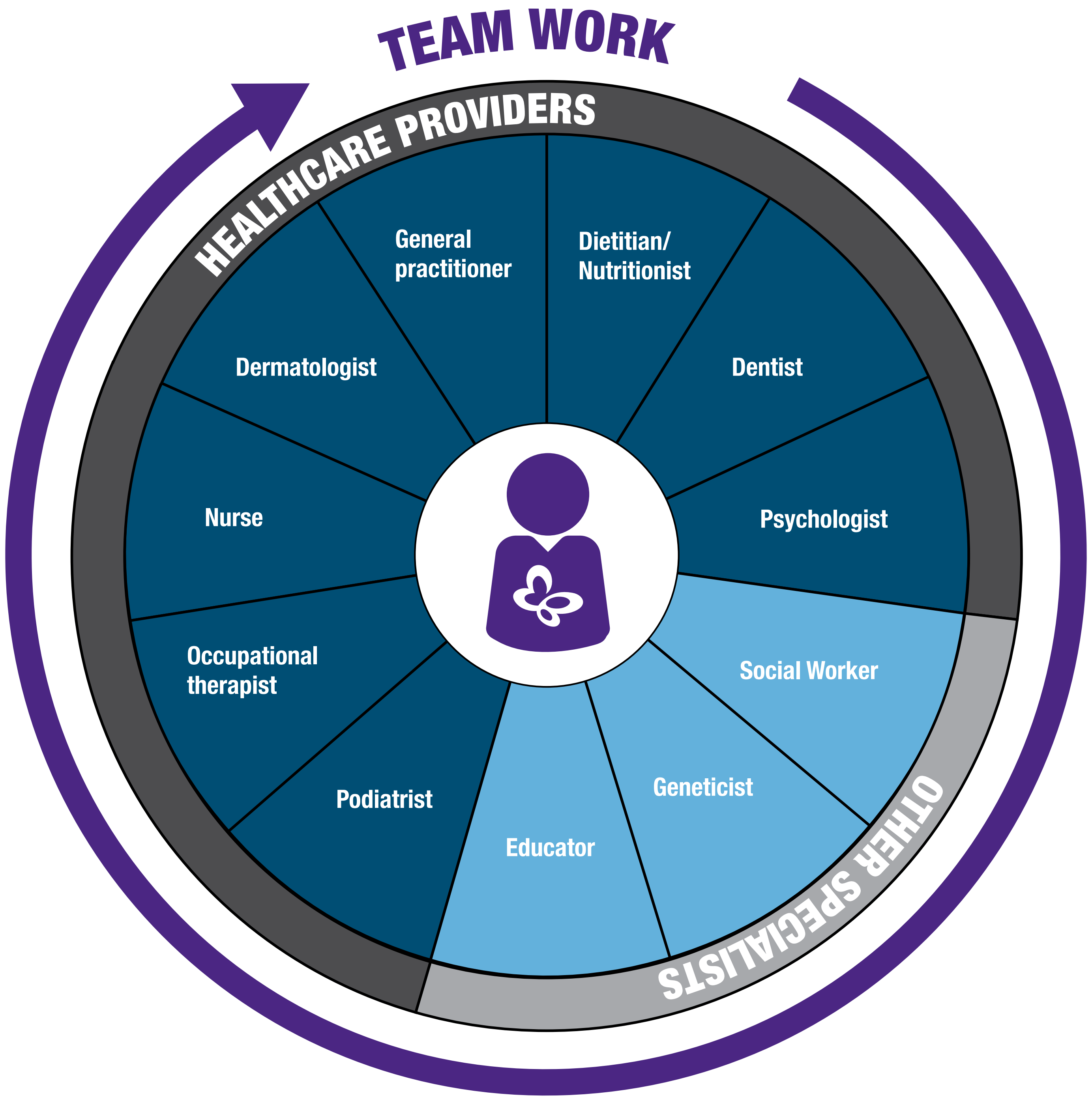
RECOMMENDATION
based on research evidence

RECOMMENDATION
based on clinical experience

This icon signposts to recommendations that overlap with those from other EB clinical practice guidelines (CPGs). These are referenced to on page 31.

WHO ARE THE EB TEAM/HEALTHCARE PROVIDERS?

Several different healthcare providers can become involved in EB care because it is such an extremely complex and rare condition. You may meet healthcare providers in different settings including at a hospital or specialist EB centre, community health service, or at your home. Different countries have different healthcare provider support systems, which can be complicated and seem difficult to access. Your national DEBRA group can help you find healthcare provider support in your country. Working together and open communication between people with EB and healthcare providers are very important. They can help maximise your physical, emotional, and social well-being.



WHO ARE THE EB TEAM/HEALTHCARE PROVIDERS?

The range of different providers that you have access to will vary greatly from individual to individual, depending on available medical and psychosocial needs. Healthcare providers that can help with EB may include:

Healthcare providers	Other specialists
Nurse specialist (in EB)	Social worker
Dermatologist	Educator
Physiotherapist	Play specialist
Occupational therapist	School specialist
Psychologist/psychotherapist	Needs assistant
Dietitian/nutritionist	Counsellor
Dentist	Geneticist
Speech and language therapist	Psychiatrist
Ophthalmologist/optician	DEBRA
Podiatrist/chiropodist	
Surgeon	
General practitioner	
Pharmacist	
Endocrinologist	
Anaesthetist	
Practice nurse	

This list is not exhaustive, there may be other healthcare providers and other specialists involved in your care, such as when you have blood tests and other investigations.

Theo Muzzi, living with EB, aged 1, Brazil (photo credit: Márcio Rodrigues/Lumini)

What can the EB team/healthcare providers do for you/your child?

Healthcare providers specialise in several different aspects of physical health care for people with EB, which may be needed at different stages of life. Social workers, psychologists, psychotherapists and other mental health and social care professionals have expertise in providing support in relation to emotional, behavioural, and social challenges that can arise. These professionals can provide direct support to you/your child with EB, your immediate family, and support network. They can also assist you in accessing more specialist psychosocial support when needed. This applies to all types of EB.

The EB team/healthcare providers can support your/your child's well-being and the well-being of your family, caregivers, and support network. This may include practical support with accessing education and social/leisure groups, support to help build confidence in talking about EB, as well as providing direct EB education to professionals who may not have experienced EB before.

They can also provide psychological support to help with difficult emotional challenges that may arise in connection with managing EB. Psychosocial support may be needed particularly at key times of change, for example the change from childhood to adolescence and adolescence to young adulthood. Psychosocial support can also be especially important in the early stages of infancy when parents, family, and caregivers are adjusting to an EB diagnosis.

Miguel Miranda Cecato, living with EB simplex, aged 4, Brazil (photo credit: Guilherme Gobbi)

WHAT CAN EB TEAM/HEALTHCARE PROVIDERS DO?

> When I was at school I had to talk to all of my teachers and it's the same at my son's school. For example, when I was at school, in gym classes they compared me to normal kids, I could skip gym class if I wanted to but they didn't make any changes or arrangements for me to be included. I loved sports and really wanted to play. It can leave you feeling on the outside. Maybe professionals can help you and others to understand EB and be confident talking about it, they may have ideas on how to help include someone who has EB and to make adjustments so we can do things.

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

> Recently, I've moved away from home and married somebody else with EB (RDEB). This has presented further issues, such as worrying about each other's health as well as just our own. Also, the very real possibility of having to bring up a child or children alone, and how the surviving parent would cope psychologically and with having EB. The introduction of care agencies and social services into my life (providing my husband's care) and the added restrictions they impose on our life together, as well as the all-consuming focus on our health as at least one of us is always unwell, and a sudden lack of parental support.

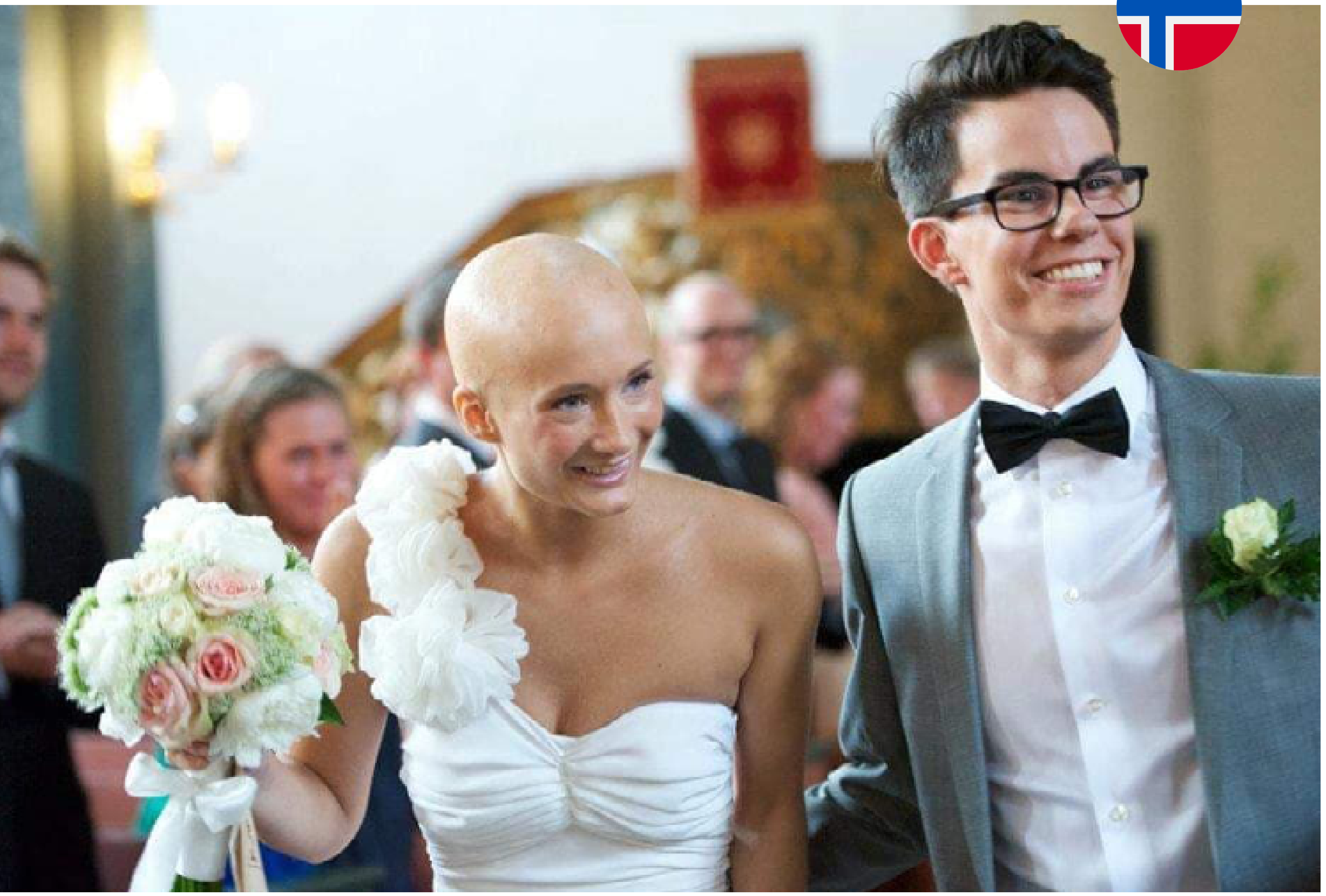
Gemma Jaega, living with recessive EB simplex, aged 28, United Kingdom;
Mike Jaega, born with recessive dystrophic EB (1971-2019), United Kingdom

> During the transition process with our young patients with EB we also prepare parents for their child's transition to the adult services. Transition to adult care can be worrying and daunting for parents as over the years they have been their child's advocate and main carer. In adult services the young person is treated as an independent.

Senior EB Clinical Nurse Specialist in association with DEBRA UK, United Kingdom

Recommendations

- Get access to a team of healthcare providers to:
  - ensure optimal pain management
  - receive medical and psychosocial care
  - optimise personal well-being, and the well-being of family, and caregivers.
- The EB team/healthcare providers should be aware of psychosocial challenges that can arise and be prepared to help, guide, and support.



> ❝ I’ve dealt with depression, and bad anxiety is still part of my life. But I always got help dealing with it. It does get better. ❞

Ida Steinlein, living with recessive dystrophic EB inversa, aged 31, Norway

What is the role of the EB team/healthcare providers regarding your/your child’s psychosocial needs?

It is important that the EB team/healthcare providers ask about your/your child’s emotional or social well-being. It is part of caring for you/your child as a whole person and not just focussing on physical needs. We would also encourage you to talk to your/your child’s EB team/healthcare providers about any physical, emotional, or social needs so they can help provide support. Sometimes it can be helpful to talk to a psychologist or psychotherapist in the specialist EB team or in the community to give extra help and emotional support. They might also suggest other services that can help.

> ❝ I have lived experience of anxiety, low mood and post-traumatic stress disorder due to the uncertain nature of EB, 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

You can ask the EB team/healthcare providers questions and be direct in suggesting ways of getting support. This is important for building a positive working relationship. You can make suggestions for your/your child’s EB team/healthcare providers to connect with DEBRA International and EB-CLINET if they have not done so already.

Recommendations

- Reach out to other people with EB. This can improve your psychosocial well-being.
- Identify and prepare for potential challenges. Healthcare professionals and other people with EB may be able to help.

> There are a lot of concerns which I didn't voice growing up, but if professionals were to ask it may have helped. These change over time, for example, there are often a lot of questions around EB in relationships; you may not have questions about this until you are in a relationship. One thing healthcare providers can offer is a broader experience, they know what is clinically best, but they need to individualise that to the patient's needs and what they want to know. So keep on asking and inviting patients to discuss how EB fits into their lives and their aims, knowing that it's ok to share questions and inviting them to talk when they want to.

Matt Lightfoot, living with dominant dystrophic EB, aged 26, United Kingdom

The importance of working together with the EB team/healthcare providers

It is important for people living with EB, their families/carers, and all healthcare providers involved in their care to adopt a collaborative approach and work together. Healthcare providers acknowledge that people with EB become experts in their own condition. Personal choice and involvement in care-related decisions are important throughout a person's life.

The EB team/healthcare providers and your national DEBRA group can work jointly with other non-EB professionals and people connected to you/your child to help provide support with different aspects of your life, such as schools, leisure activity providers, and employers.

Having a team of people who can work together with you/your child can help provide a sense of support and reduce the burden of EB in everyday life.

Photo credits: Karla Nogueira, projeto DIV


Tauani Vieira, living with recessive dystrophic EB, aged 25, Brazil

> “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

Expert opinion

A collaborative patient-professional relationship can help provide a sense of support.

The EB team/healthcare providers have an important role in providing education about EB. They can help you/your child to understand more about EB and increase awareness and knowledge among those who may have little to no knowledge of the condition, such as general practitioners, school staff and classmates, employers and colleagues. Increasing EB awareness has been found to be helpful in improving well-being in some people with EB.

National DEBRA groups hold numerous events throughout the year to promote EB awareness and people with EB are encouraged to tell their personal stories. Being able to relate to other people with EB can bring a sense of not being alone and enhances psychological well-being.

Recommendation

Ask your EB team/healthcare providers to help promote awareness and provide education about EB.

> **“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.”**

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

You and your EB team can work together to promote EB awareness

Tip

What you can do:

Ask for help - work together with your EB team/healthcare providers to gain the most positive outcomes in your physical and psychosocial care. They are a good resource to help teach about EB. Talk to them about any emotional or psychological concerns that you have.

Building confidence to talk to others about EB

The EB team/healthcare providers can provide education about EB, and support people with EB and their families to build their confidence when talking about the condition:

- **Teaching others** – they can provide information and teach anyone involved in caring for you/your child about EB.
- **Transition** – they can provide information during times of change, such as moving to a new school/university, or workplace. They can provide school or work visits, letters of support, education for staff and employers.
- **Information** – they can give you information about EB and services that provide support, including national DEBRA group contact details. Some information leaflets may be available to help explain EB and specific details, such as types of EB, pregnancy in EB, and wound care.

> I am the only one who knows my pain. How are they going to know unless I tell them? Professionals can help. Everyone is different. I had treatment for my feet and it really helps with the pain, I can walk much better.

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

The importance of working on pain management together

Pain can be very distressing and affect daily activities and social interactions. The EB team/ healthcare providers have an important role in working together with people with EB and their care networks to establish optimal pain management. It is important to discuss pain management support and get advice from them so that the impact of pain can be reduced where possible.

Expert opinions

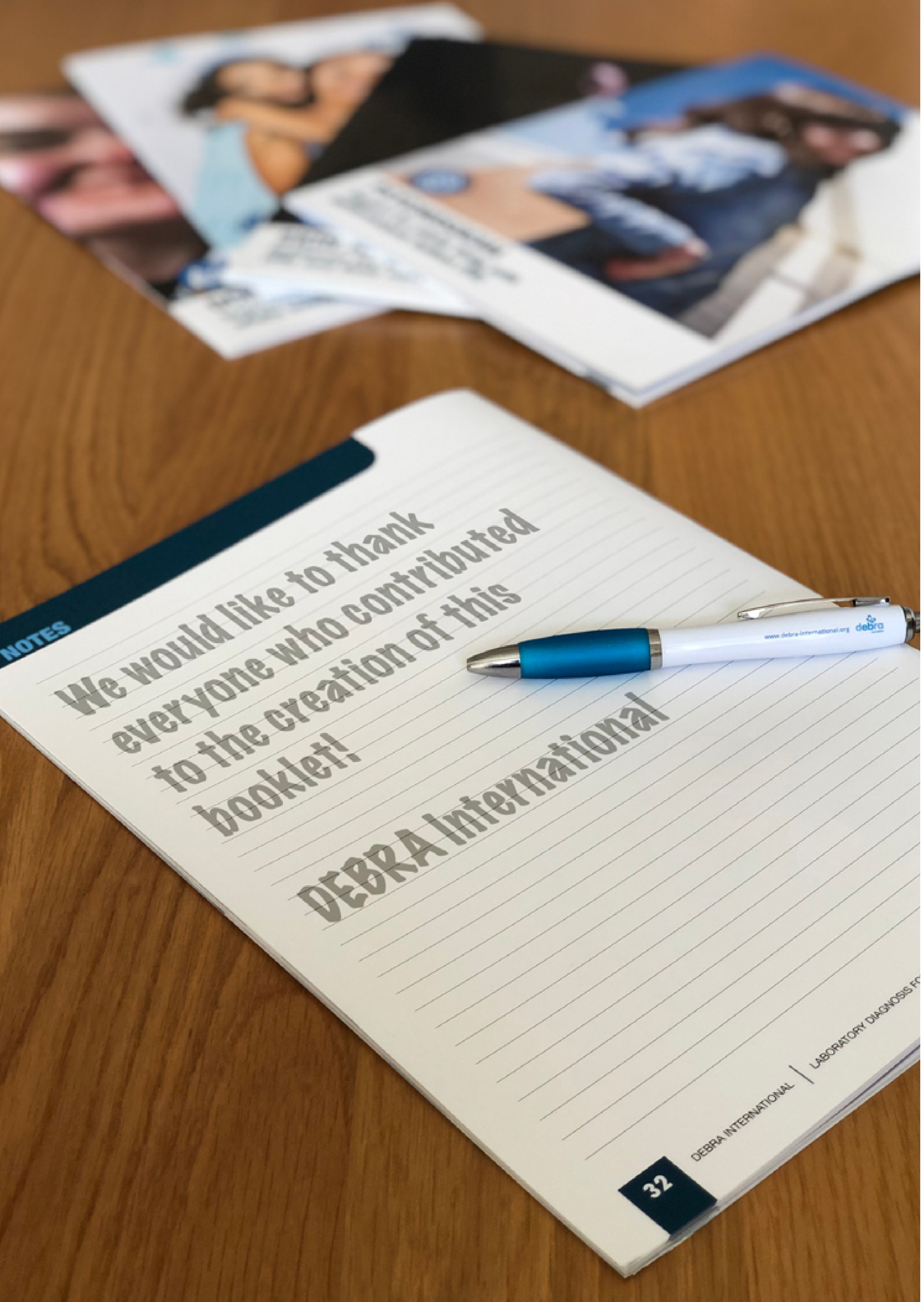
- The EB team/healthcare providers have an important role in managing pain to help people with EB physically and emotionally.
- When dressing changes have the potential to cause pain, your/your child’s EB team/healthcare providers should consider the need for medication or therapies for treatment including psychological support.

Summary

- Everyone’s individual experiences are unique - you are an expert in your EB; your child an expert in theirs.
- Working together with the EB team/healthcare providers is vital to gain positive outcomes in care.
- The EB team/healthcare providers can help with your/your child’s physical and psychosocial well-being.
- The EB team/healthcare providers are a good resource to help teach about EB.

> Maybe some people feel they don’t need support because they just get on with life and living with EB. Maybe they don’t know what help they can get. Maybe that’s why there’s less research into simplex; nobody tells about their struggles so the resources aren’t put into EB simplex. We need to be open and tell professionals about what we are struggling with, maybe they can help, maybe not, but it is worth a try.

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



We would like to thank everyone who contributed to the creation of this booklet!

DEBRA International

Tell us what you think

Have your say in the future patient versions of clinical practice guidelines (CPGs) for epidermolysis bullosa (EB)

The purposes of this survey are to:

- assess the quality of the information, presentation, and delivery of the patient versions
- help us to develop a standard for all patient versions now and in the future.

The data collected will help us to improve the information provided and experience of the user in all future CPG patient versions. The data may be used to report the development steps taken to improve their quality; this may be done through conference presentations, posters, abstracts, or studies.

We want to make sure that all patient information provided meets the needs of everyone living with EB.

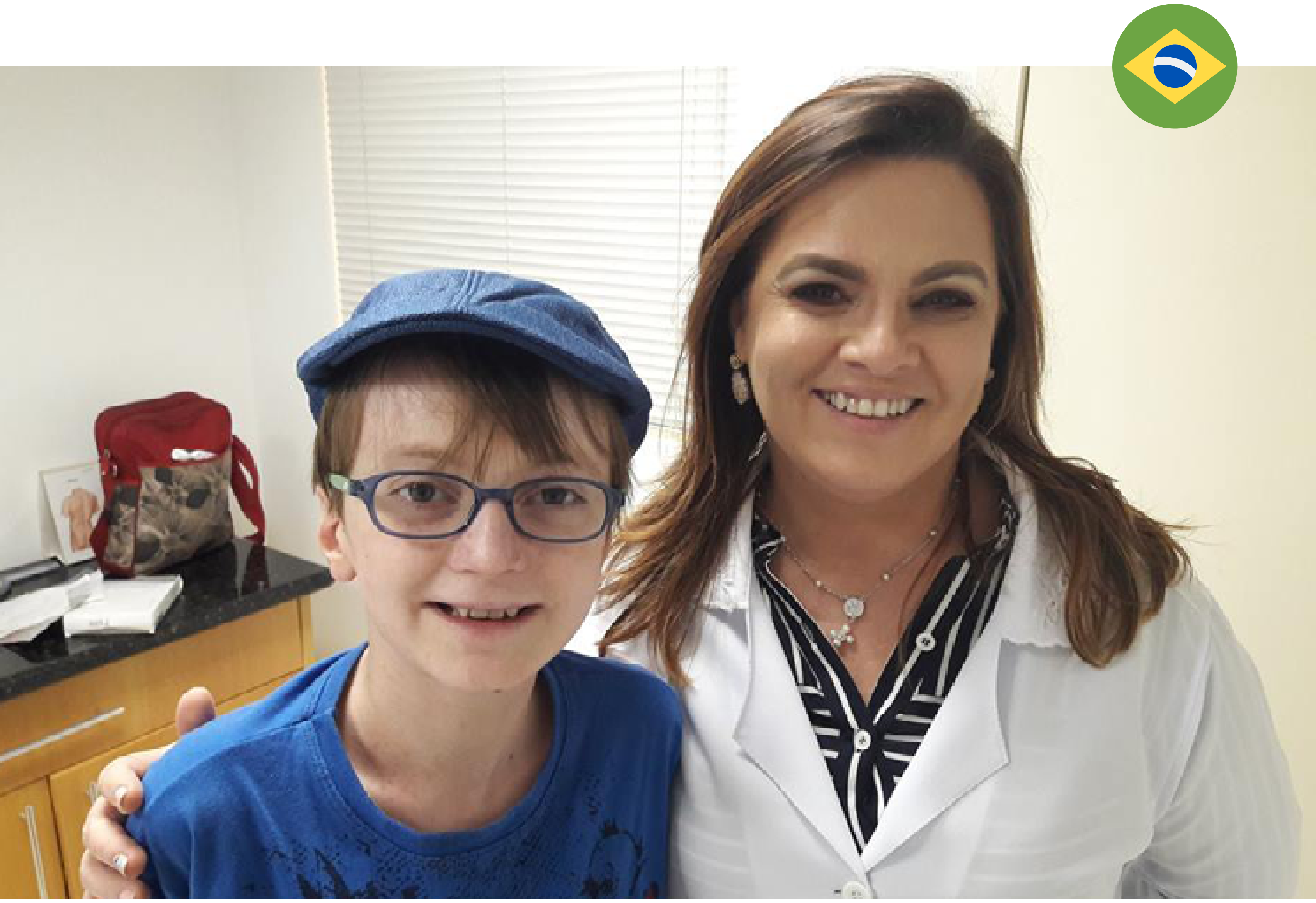
Help us create new CPGs and patient versions

All responses to the above survey are confidential unless you decide to join the DEBRA International CPG network. Please consider joining the network if you are interested in becoming involved in the development of CPGs and patient versions in the future. To join the network, please complete question 1 on page 7. If you do not complete this question, we will not receive any of your personal details and you will remain anonymous. Joining the CPG network is entirely voluntary and you may choose to opt out at any time by contacting DEBRA International.

If you have any questions when completing this survey or about joining the CPG network, please contact the DEBRA International CPG Coordinator, Kattya Mayre-Chilton by email at: kattya.mayre-chilton@debra-international.org

TELL US WHAT YOU THINK!

Answer the survey to help us improve the patient versions of the CPGs:
www.surveyhero.com/c/PatientVersionsSurvey



Emanuel Kluge, living with recessive dystrophic EB, aged 18, Brazil (photo credit: Suelen Szymanski)

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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Development source

Psychosocial recommendations for the care of children and adults with epidermolysis bullosa and their family: evidence based guidelines.

This CPG was approved by DEBRA International and funded by DEBRA Norway.

To access the following CPGs and patient version booklets, please visit: www.debra-international.org

Psychosocial booklets

- Psychosocial support for parents caring for a child living with epidermolysis bullosa (EB)
- Psychosocial support for adults living with epidermolysis bullosa (EB)

Other CPG topics

- Clinical Practice Guidelines for Epidermolysis Bullosa Laboratory Diagnosis
- Foot care in Epidermolysis bullosa: Evidence-based Guideline
- International Consensus Best Practice Guidelines for Skin and Wound Care in Epidermolysis Bullosa
- Management of Cutaneous Squamous Cell Carcinoma in Patients with Epidermolysis Bullosa: Best Clinical Practice Guidelines
- Occupational therapy for epidermolysis bullosa: clinical practice guidelines
- Oral Health Care for Patients with Epidermolysis Bullosa - Best Clinical Practice Guidelines
- Pain care for patients with epidermolysis bullosa: Best care practice guidelines

Other languages

We are happy to consider requests for this booklet to be provided in other languages. Please send all enquiries to: office@debra-international.org

How was the Psychosocial guideline produced?

- The CPG development group 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, CINHAL, 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.

Panel group

- K Martin - CPG Chair (United Kingdom)
  Consultant Clinical Psychologist, adults and families - Solihull Hospital
- S Geuens - CPG Co-chair (Belgium)
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- J K Asche - CPG Member (Norway)
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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
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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.

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EB-CLINET

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

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Get involved with DEBRA International

THE WORLD'S LEADING EB PATIENT ADVOCACY AND SUPPORT NETWORK

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