



PODIATRY

Dystrophic nail care for people living with epidermolysis bullosa (EB)

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



LOWE GGGK

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



Ingrid Vitoria Aparecida Martins, living with recessive dystrophic EB, aged 25, Brazil (photo credit: Suelen Szymanski)



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Diane Laverne Inns, living with EB simplex, aged 51, United Kingdom

Foreword

In an EB registry study involving males and females of all ages, dystrophic nails were reported on 33.3% of patients with EB simplex, 90% of patients with junctional EB, 83.25% of patients with dominant dystrophic EB, and 94.8% of patients with recessive dystrophic EB (Rogers, 2002).

Who is this booklet for?

This booklet is for people living 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 provides information on dystrophic nail care for people for people living with EB.

Topics covered in this booklet include:


- ▶ what are dystrophic nails in EB?
- ▶ what are the recommended treatments?
- ▶ helpful tips
- ▶ wound care and pain management
- ▶ staying active and mobile
- ▶ how your/your child’s podiatrist can help

Where does the information in this booklet come from?

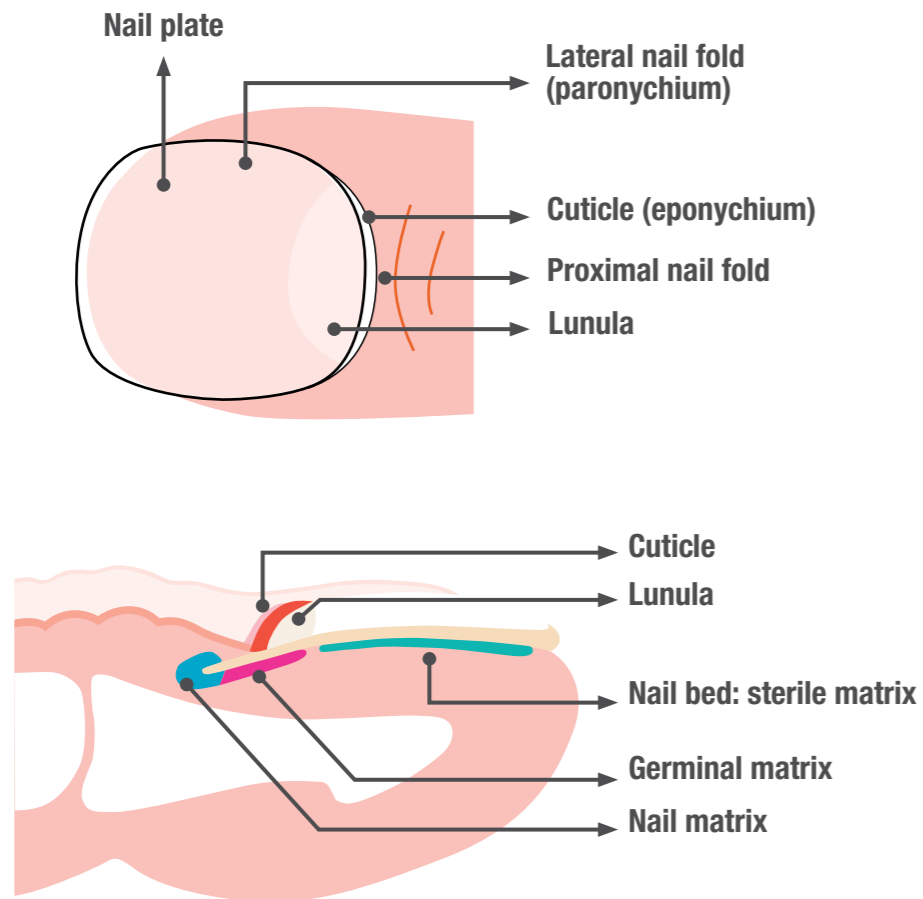
The information and recommendations in this booklet are derived from the “Foot care in Epidermolysis bullosa: Evidence-based Guideline”. The guideline was written in 2018 by a group of EB healthcare professionals, individuals living with EB, and family members. The information and recommendations in the guideline come from a variety of sources including clinical research and expert opinion.

There are three different types of recommendation in this booklet:

 <p>STRONG RECOMMENDATION based on good-quality research evidence</p>	 <p>RECOMMENDATION based on research evidence</p>	 <p>RECOMMENDATION based on clinical experience</p>
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 This icon signposts to recommendations that overlap with those from other EB clinical practice guidelines (CPGs). These are referenced to on page 21.

Anatomy of a nail and cross section



“ Growing up with very thick and sharp fingernails was sometimes a challenge. I had to be extra careful whenever I changed my clothes, as they would get caught in things, which can be extremely painful. Sometimes I’d wake up with scratch marks on my face and body. ”

Male, living with junctional EB, aged 35, Australia

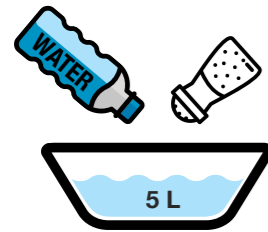
Dystrophic nails refer to changes and abnormalities to the nail resulting from trauma and scarring of the nail plate and matrix. This can cause the nail (toenails, fingernails, or both) to appear damaged, misshapen, discoloured, thickened, and curvy. Some nails may even start to lift from the nail plate or come off completely.



Dystrophic nails can be very problematic in EB and may affect all EB subtypes. There is a large range of severity from nail abnormalities that may cause severe disability and pain to only mild cosmetic problems.



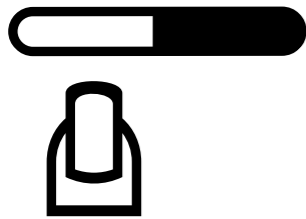
Nail care for toes and fingers



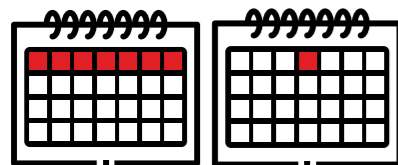
01 Add 5 litres of clean lukewarm water and 2 tablespoons of salt (45 g)



02 Soften the nails by soaking them in lukewarm water with salt

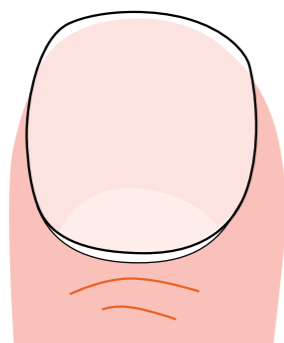


03 File nails with an emery board and keep nails trimmed straight across

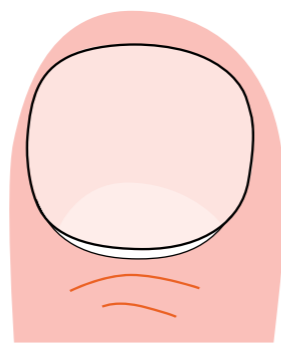


04 For adults and thick nails = daily
For children = once per week

Trimming nails



Correct



Incorrect

If you or your child has dystrophic fingernails or toenails, you are advised to:

- ▶ keep fingernails and toenails trimmed straight across
- ▶ file nail surfaces with an emery board after softening the nails by soaking them in warm saline water, baby oil, or in the bath
- ▶ apply emollient or moisturiser daily for adults and weekly for children. A urea-based cream is recommended to reduce the thickness of the nail. This will also hydrate the nail, softening it and making it easier to cut.

Strong recommendation

Manage problematic dystrophic nails by applying a topical therapy and implementing a nail reduction treatment

Podiatrists are sometimes asked to deal with fingernails as well as toenails by their EB consultant. Podiatrists should ensure that this activity is within the scope of the podiatric practice act for their country and check whether certification is required.

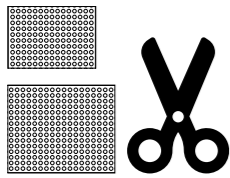
Helpful tips

- ▶ Many people living with EB have also reported bad experiences regarding podiatrists who have no understanding of the nature of the condition. Those with access to a can pass on the podiatry service contact details to provide any additional support/information/advice to their local podiatrist.
- ▶ Please direct your/your child's podiatrist to the areas of treatment you wish to be managed and instruct them as to how much you would like the nail cut back or filed.
- ▶ Ensure footwear is the appropriate size and depth to accommodate dystrophic nails to avoid rubbing.

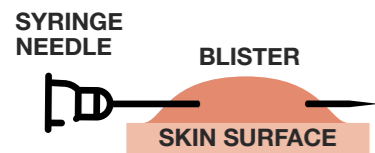
“As an EB patient, my nails are much more prominent and more difficult to cut, it can be time consuming and painful. But with the help of the special equipment podiatrists use, it helps make the procedure pain-free.”

Male, living with junctional EB, aged 35, Australia

Bigger blisters = bigger wounds! Pop a blister as soon as you see it. Cleaning and removing debris/dead tissue helps to reduce bacteria in wounds and promote wound healing.



01 Prepare all dressing material in suitable sizes and amounts in advance to keep dressing changing times shorter.



02 Use a clean needle to create an entry and exit hole so the fluid can escape



03 Roll a soft clean cloth over the blister to expel fluid



04 The roof should be left on the blister to protect the wound



05 Remove dead skin around the open wound

As EB blisters are not self-limiting and can spread, intact blisters should be lanced and fully drained even at the areas of the feet and toes. Your/your child's specialist EB trained podiatrist or EB team may have shown you how to do this but you can also find advice in the skin and wound care CPG and from the illustration on page 12 of this booklet.



Recommendation from the skin and wound care CPG

You should lance intact blisters and fully drain them or they will refill.



Wound dressings, insoles, and socks will also need to be considered when using footwear. If dressings are worn daily on the feet then foot size should be measured with dressings in place when choosing suitable footwear.

Painful feet and neuropathic pain are common complaints with EB. Neuropathic pain is caused by damage or injury to the nerves that transfer pain information. This pain is often described as a 'burning' pain on the soles of the feet. Some of these issues are addressed in the pain management CPG. They emphasise that an important aspect of pain care for individuals is to have careful or professional attention to their footwear, nails, orthotics, and hyperkeratosis management. Hyperkeratosis refers to thickening of the outer skin layer made of keratin protein.



Strong recommendation from the pain management CPG

Discuss how to optimise your/your child's pain control with your/your child's specialist EB trained podiatrist or EB team.



Recommendation from the pain management CPG

Discuss with your/your child's specialist EB trained podiatrist or EB team to see if there are any topical therapies you could use.





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

Problems such as blistering, hyperkeratosis (callus), nail loss, altered gait, and deformity can result in reduced mobility and, eventually, increased wheelchair use. Preventing these problems can help you/your child to stay mobile for longer and improve your/their quality of life.



Recommendations

- ▶ Preventing these problems can help children and adults of all subtypes of EB to stay mobile for longer and improve their quality of life.
- ▶ Podiatric treatment can help you/your child stay active and mobile for longer.
- ▶ Discuss how to access programs to optimise mobility with your/your child's specialist EB trained podiatrist or EB team.

“ In the absence of any current ability to switch off EB, good quality and regular podiatry is the single most useful intervention to improve quality of life. ”

Consultant Dermatologist, United Kingdom



How your/your child's podiatrist can help

Specialist EB trained podiatrists should be available to:

- ▶ file nail surfaces with an emery board
- ▶ extract a difficult nail
- ▶ help with dystrophic nail management
- ▶ examine your/your child's feet
- ▶ develop podiatry treatment plans and recommend suitable footwear
- ▶ offer treatment at a specialist clinic, such as for the development of bespoke footwear
- ▶ recommend appropriately trained podiatry services near your home.

Please contact your national DEBRA group for more information on your local EB specialist clinic.

When people want to go privately we advise them to check the podiatrist is registered with their governing body.



Strong recommendation

Get specialised podiatry support to treat and manage dystrophic EB nails.

“Patients who have never had Podiatry care before are often hesitant about allowing someone to tend to their feet but, even after simple treatments such as nail care, the reactions are overwhelmingly positive!”

Mark O'Sullivan, Podiatrist, United Kingdom

Janaína Rodrigues Pereira, living with recessive dystrophic EB, aged 40, Brazil (photo credit: Suelen Szymanski)



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.

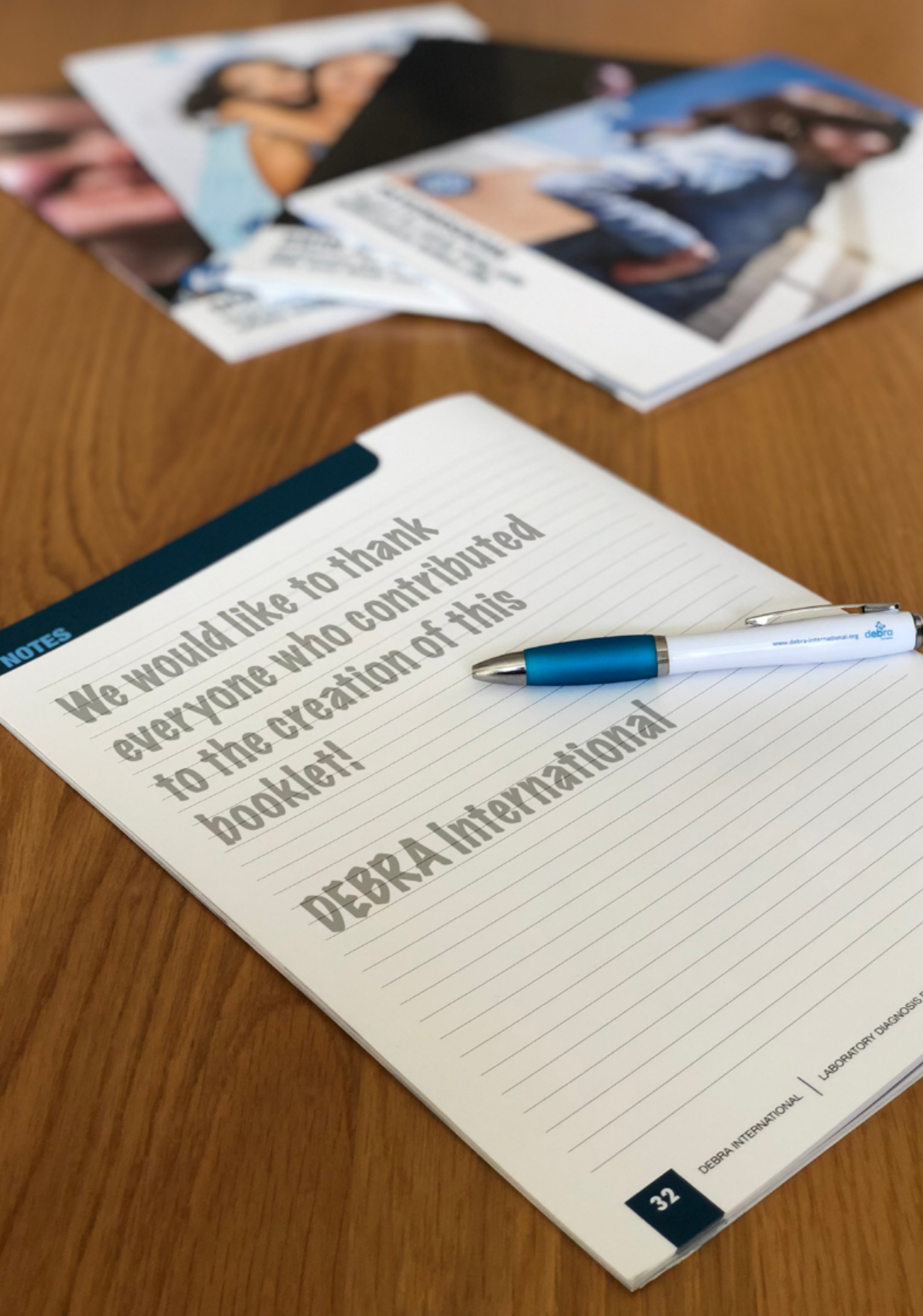
Help us create new CPGs and patient versions

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, Katty 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.surveymhero.com/c/PatientVersionsSurvey





Miguel Kingerski da Silva Calgaro, living with recessive dystrophic EB, aged 5, 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 UK, 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

Foot care in Epidermolysis bullosa: Evidence based guidelines

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

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



Podiatry booklets

- ▶ Podiatry hyperkeratosis (callus) care for adults living with epidermolysis bullosa (EB)
- ▶ Podiatry footwear advice for parents caring for a child living with epidermolysis bullosa (EB)
- ▶ Podiatry footwear advice for adults living with epidermolysis bullosa (EB)

Other CPG topics referred to in this booklet



International Consensus Best Practice Guidelines for Skin and Wound Care in Epidermolysis Bullosa



Pain care for patients with epidermolysis bullosa: Best care practice guidelines

Other CPG topics

- ▶ Clinical Practice Guidelines for Epidermolysis Bullosa Laboratory Diagnosis
- ▶ 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
- ▶ Psychosocial recommendations for the care of children and adults with epidermolysis bullosa and their family: evidence based 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 Foot care guideline produced?

- ▶ The CPG development group consisted of EB experts: podiatrists, dermatologists, a physiotherapist, a dietitian, and patient representatives.
- ▶ All panel members completed written conflict of interest and code of conduct declarations.
- ▶ During guideline development, they conducted several panel meetings where six to nine members would meet in person and virtually. They discussed the clinical questions and methodology; reviewed the evidence (publications in the field of EB podiatry research); formulated recommendations; and agreed 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 podiatry and foot care research together with the personal experiences of the panel members.
- ▶ To identify publications, seven electronic search engines were accessed, including Wiley Online Library, Google Scholar, Athens, ResearchGate, Net and PubFacts.com. The search period ended December 2018.
- ▶ Out of 46 papers appraised, 36 were chosen for the final recommendations by six panel members according to the Critical Appraisal Skills Programme (CASP) and Scottish Intercollegiate Guidelines Network (SIGN) quality rating.

Panel group

- ▶ M T Khan - CPG Lead (United Kingdom / Australia)
Specialist EB Paediatric Podiatrist - Great Ormond Street Hospital for Sick Children, London; Royal London Hospital for Integrated Medicine, UCLH, London; St George Hospital, Sydney, NSW; Barts and The London NHS Foundation Trust, London
- ▶ M O'Sullivan - CPG Co-lead (United Kingdom)
Specialist EB Adult Podiatrist - University Hospitals Birmingham NHS Trust, Solihull Hospital, Solihull
- ▶ B Faitli - CPG Member (United Kingdom)
Person living with EB
- ▶ J E Mellerio - CPG Member (United Kingdom)
Consultant - St John's Institute of Dermatology, Guy's and St Thomas' Hospital NHS Foundation Trust, London
- ▶ R Fawkes - CPG Member (United Kingdom)
Specialist EB Podiatrist (Retired) - St Thomas' Hospital NHS Foundation Trust, London
- ▶ M Wood - CPG Member (United Kingdom)
Specialist EB Paediatric Physiotherapist - Great Ormond Street Hospital for Sick Children, London
- ▶ L D Hubbard - CPG Member (United Kingdom)
Principal dietitian in adult EB - St Thomas' Hospital NHS Foundation Trust, London

- ▶ A G Harris - CPG Member (Australia)
Dermatologist - St George Hospital, Sydney, NSW; Department of Dermatology, Concord Hospital, Sydney, NSW
- ▶ L Iacobaccio - CPG Member, CPG patient version Lead (Australia)
Specialist EB Adult Podiatrist - The Royal Melbourne Hospital, Melbourne, VIC
- ▶ T Vlahovic - CPG Member (United States)
Specialist EB Podiatrist - Temple University, Philadelphia, PA
- ▶ L James - CPG Member (United Kingdom)
Specialist EB Adult Podiatrist - University Hospitals Birmingham NHS Trust, Solihull Hospital, Solihull
- ▶ L Brains - CPG Member; Patient version Co-lead (Australia)
Person living with EB
- ▶ M Fitzpatrick - CPG Member (Australia)
Person living with EB, DEBRA Australia
- ▶ K Mayre-Chilton - Methodologist, CPG Member (United Kingdom)
DEBRA International CPG Coordinator; Specialist Adult Research Dietitian - Guy's and St Thomas' Hospitals NHS Foundation Trust, London

External review group

- ▶ C Pankhurst (UK)
Clinical Specialist - Guy's and St Thomas' Hospitals NHS Foundation Trust, London; Clinical Network: Foot Care (Co-chair); British Association of Prosthetists and Orthotists; (Professional Affairs Committee Co-vice-chair); Foot in Diabetes UK (Vice-chair)
- ▶ K A Bø (Norway)
Person living with EB

DEBRA International project team

- ▶ K Mayre-Chilton - CPG patient version project lead (United Kingdom)
CPG Coordinator - DEBRA International
- ▶ O Mullins - Editorial and format lead (United Kingdom)
Business Manager - DEBRA International
- ▶ L A Taguchi - Branding and design lead (Brazil)
Mother of a person born with EB; Marketing Director - DEBRA Brazil
- ▶ M Cardoza - Graphic designer (United Kingdom)
- ▶ A Winter - Design assistant (United Kingdom)
Fundraising Events Coordinator - DEBRA UK

-  ▶ **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
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www.debrabg.net
contact@debrabg.net
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-  ▶ **Canada**
www.debracanada.org
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-  ▶ **China**
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-  ▶ **Japan**
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www.debra.nl
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-  ▶ **New Zealand**
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+886 03 5734493
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-  ▶ **Ukraine**
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info@debra-ukraine.org
svitdebra@gmail.com
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-  ▶ **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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