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Foot care in Epidermolysis bullosa: Evidence-based Guideline

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Abstract

To provide service providers and users with an evidence-based set of current best practice guidelines for people and their families and carers, living with Epidermolysis bullosa (EB). A systematic literature review relating to the podiatric care of patients with EB was undertaken. Search terms were used, for which the most recent articles relating to podiatric treatment were identified as early as 1979 to present day, across seven electronic search engines: Medline, Wiley online library, Google Scholar, Athens, Researchgate, Net and pubfacts.com. The Scottish Intercollegiate Guidelines Network (SIGN) methodology was used. The first guideline draft was analysed and discussed by clinical experts, methodologists and patients and their representatives at four panel meetings. The resulting document went through an external review process by a panel of experts, other health care professionals, patient representatives and lay reviewers. The final document will be piloted in three different centres in United Kingdom and Australia. Following an EB community international survey the outcomes indicated six main areas which the community indicated as a priority to foot management. These included blistering and wound management; exploring the most suitable footwear and hosiery for EB; management of dystrophic nails; hyperkeratosis (callus); maintaining mobility; and fusion of toes (pseudosyndactyly). Evidence here is limited but several interventions currently practised by podiatrists show positive outcomes.

Introduction

The Dystrophic Epidermolysis Bullosa Research Association (DEBRA) International is a worldwide network of national groups working for people affected by the genetic skin blistering condition. EB is a group of rare heritable skin fragility disorders, typically

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presenting as blistering of the skin from minor trauma¹. While there are currently over 30 known subtypes of EB, there are four primary types including EB Simplex (EBS), Dystrophic EB (DEB), Junctional EB (JEB), and Kindler syndrome (KS)¹⁻². EB can be the result of either inherited or spontaneous dominant mutations, as seen in most forms of EBS and Dominant DEB (DDEB); or from inherited recessive mutations as is the case with rare forms of EBS, Recessive DEB (RDEB), JEB, and KS² (Fig. 1). Ninety percent of EB patients have one or more podiatric manifestation, including blistering, hyperkeratosis, flat feet, nail dystrophy or structural abnormality affecting foot positioning³⁻⁴. EB requires specialised podiatric care but because of its rarity many podiatrists have limited knowledge of the disorder. Furthermore there is a dearth of evidence regarding podiatric care of EB and management decisions are usually based on experience and expert opinion.

The recommendations outlined in this clinical practice guideline (CPG) contain general information on foot care of people living with EB, Table 1. They explain the precautions that should be taken when treating people with EB, as well as recommendations for podiatry treatment.

Objectives of the CPG

- To describe foot problems in people of all ages with EB
- To outline current EB podiatry practice in the UK and Australia
- To highlight specific considerations for different subtypes of EB
- To provide guidance for foot care in EB

Users These guidelines are intended for podiatrists, other health professionals, people with EB (all ages and subtypes), their families and carers, teachers, employers, shoe manufactures, stakeholders and policy makers.

Target group These guidelines comprise information relating to people with EB of all ages and subtypes.

Recommendations

Blistering and wound management

Podiatric education

We strongly recommend offering podiatry education programmes to prevent blistering and wounds (*Strength of recommendation Grade: B*)

Foot blistering is a common problem in all subtypes of EB and all age groups may be affected (Quality evidence level ranged from 4 to 2+) ⁴⁻⁸. Blistering in EB usually results from friction or minor trauma ^{3,9}. Blisters on the feet can be caused by a dressing, socks, shoe, or boot rubbing against the skin ³, but sometimes appear spontaneously. The size of a blister depends on the type of EB and the degree and duration of friction ³. In EB, defective skin adhesion means that a shearing force causes skin components to separate, creating a space which fills with fluid. The resulting blister easily enlarges under pressure because there is a plane of weakness in the skin, so it should be burst to avoid this (Appendix 1a).

The usual technique is to lance intact blisters with a sterile needle at their lowest point to facilitate fluid drainage by gravity ⁹ and to stop blisters from refilling ³. Some patients prefer to use sterile scissors. Gauze or other absorbent material may be used to wick the fluid from the blister. A saline soak, non-medicated and medicated dressings and the use of topical antiseptics or antibiotics could be used to prevent secondary infection until the skin heals (Quality evidence level ranged from 3 to 1-) ^{3,5}, existing guideline \Rightarrow ⁹ (Expert opinion *Grade: D*).

Generally the management of EB tends to be supportive and is aimed at preventing blistering by reducing friction and the amount of mechanical trauma to the feet (Quality evidence level 3) ⁵. Prevention is key and involves minimising friction and mechanical trauma to the feet (Quality evidence level 3) ⁵. The evidence here supports the training of staff, patients and carers to improve understanding of the causes of blistering and wounds on feet.

Prevention of blisters is facilitated by an assessment tool which the podiatrist can use to address the adequacy of hosiery (silver-lined socks) and footwear (Quality evidence level 4) ⁴. This study presented the development of a universal assessment tool which

requires validation (Appendix 2; Section A). The foot health status questionnaire is seen as a universal assessment validate tool which is not EB-specific (Quality evidence level 1-)¹⁰, (Appendix 2; Section B).

In a cohort of 57 patients with a localized form of EBS (EBS-l) all reported localized pain in the feet related to blisters, of these 6 (11%) tried 5% lidocaine plasters on their feet, with good efficacy (Quality evidence level 2+)⁶.

Footwear and Foot biomechanics

Selection of appropriate footwear and the use of appropriate insoles can help to reduce blisters and improve foot function in EB. In a prospective study of 6 EBS patients, 3 reported no new blisters while wearing shock-absorbing moulded orthoses (Quality evidence level 3)⁵.

In a qualitative study of 79 adults with all subtypes of EB, all patients improved in 2 to 4 key variables when using shock absorbing insoles, custom orthotics and bespoke footwear. They used gait analysis system to capture static and dynamic in-shoe foot pressure of bespoke footwear in 7 patients, provided an objective, quantifiable technique to identify biomechanical discrepancies and pathological foot function and to assess gait. Furthermore the improved mobility and independence correlated to reduced numbers and severity of blisters (Quality evidence level 2-)⁷, demonstrating that the use of insoles and orthotics are also important (Quality evidence level 2-)⁴ (Appendix 3 & 4).

- Socks are helpful to provide ventilation, wick away moisturise and reduce friction (Quality evidence level ranged from 4 to 2-)³⁻⁴.
- Footwear for EB patients ideally should be firm, comfortably fitting with appropriate length and width, a rounded toe, a flexible flat sole with heel support, have laces/straps or equivalent to prevent excessive movement or slipping of the foot inside the shoe and have a seamless internal lining (Quality evidence level ranged from 4 to 2-)³⁻⁴.

Practical point; although not mentioned in the literature, healthcare professionals and patients alike have reported the benefit of using cornflour on the soles of the feet and in-between the toes to help control excessive moisture and reduce friction. Both of which can help control blistering on a day to day basis.

Dystrophic nails

We strongly recommend offering podiatry support to treat and manage EB Dystrophic nails (*Strength of recommendation Grade: B*)

- Dystrophic nails can be very problematic in EB and may affect all EB subtypes (Quality evidence level 2-)¹²⁻¹⁵
- Dystrophic nails may be effectively managed by the application of a topical keratolytic agent and the nail thickness can be further reduced by an expert podiatrist (Quality evidence level 2-)^{3-4,12-15}

Nail changes occur in all subtypes of EB. In an Australian EB registry study involving males and females from childhood onwards, dystrophic nails were reported in 33.3% of patients with EBS, 90% of those with JEB, 83.25% of DDEB and 94.8% of RDEB patients (Quality evidence level 3)¹². Most reports focus on toenails rather than fingernails, although both can be dealt with by podiatrists, Box 1. A retrospective qualitative study reported on 201 adults (Quality evidence level 2+)⁸; most other cases discuss the diagnosis, characteristics and familial inheritance in childhood.

BOX 1	Disclaimer: Podiatrists are sometimes asked to deal with finger nails as well as toe nails by their EB consultant. Podiatrists should ensure that this activity is within the scope of the Podiatric practice act for their country and whether certification is required
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Toenails should be preserved where possible because they protect the tips of digits from friction and pressure (Quality evidence level ranged from 4 to 2-)³⁻⁴. The treatment and management of dystrophic nails presented the strongest evidence in this CPG, and encourages podiatrists to be involved with all EB patients (Quality evidence level ranged from 4 to 2-)³⁻⁴. As EB nails are a rare condition, community podiatrists are unlikely to have the disease-specific knowledge and expertise to deal with EB-related

complications (Quality evidence level 2+)⁸. Therefore podiatry is an essential component of EB multidisciplinary care (Quality evidence level 2+)⁸. (Appendix 1b).

EB podiatrists should be available to assess newly diagnosed patients, develop care plans, offer treatment at the specialist centre, and to recommend appropriately trained podiatry services near the patients' home.

EB Patients with dystrophic nails should be advised to (Quality evidence level ranged from 4 to 2-)³⁻⁴:

- keep toenails trimmed straight across
- file nail surfaces with an emery board after softening the nails by soaking in warm saline water or a bath
- Apply daily to weekly depending on age of the individual and thickness of the nail a urea based cream, such as a keratolytic agent, to reduce the thickness of the keratin layer and hydrate the nail (Quality evidence level 4 Expert opinion).
- The removal of the toenails can be performed via chemical or laser ablation to prevent future problems. If an EB Podiatrist is not undertaking this procedure it is advisable for them to provide some guidance/advice to the Podiatrist who is performing the nail surgery (Quality evidence level 2-)⁴.
- Surgical procedure can be carried out, please refer to pseudosyndactyly section.

Hyperkeratosis (Callus)

We strongly recommend assessment of hyperkeratosis with a validated tool to facilitate monitoring. (Strength of recommendation Grade: B)

- Hyperkeratosis and fissuring of the feet has been reported in all EB subtypes (Quality evidence level 2+)⁸
- The use of a validated tool can help to monitor, evaluate and manage EB hyperkeratosis (Quality evidence level 1-)¹⁰⁻¹¹.
- Pressure redistribution and cushioning is helpful to prevent hyperkeratosis development (Quality evidence level 2-)^{3-4,7-8}

Hyperkeratosis (Callus) has been reported in all subtypes of EB. EBS is often associated with mild to moderate hyperkeratosis (palmoplantar keratoderma), particularly of the

soles^{4-5,7}. In a retrospective qualitative study, carried out in an EB podiatry clinic covering both males and females with all subtypes from childhood onwards, 74 out of 201 patients (36.8%) were treated for hyperkeratosis (Quality evidence level 2-)⁸.

Hyperkeratosis (also called keratoderma or callus) may be defined as hard, thickened areas of the skin located on the tip of toes or between the toes and soles underneath the metatarsal heads³. If the skin is hard and yellow with a nucleus or plug of keratin, it is called a corn or helom³. A corn or callus will appear red if it is inflamed³. The central core of a corn extends downwards in a cone-shaped point and can cause notable pain/discomfort. Patient's will often compare this to walking on a small stone or pebble³. A corn or callus enlarges if there is continuing friction as a direct hyperproliferative response of keratinocytes³. Hyperkeratosis is, to a limited extent, protective³. However, in EB blisters can form under the thickened tissue and painful cracks can develop. (Appendix 1a). The foot health status questionnaire is currently being used in studies with EB patients with Hyperkeratosis (Quality evidence level 1-)¹⁰.

Podiatric management of hyperkeratosis and corns involves the following³:

- Debridement of the lesions is a procedure performed regularly by Podiatrist using manual debridement/paring of hyperkeratosis (Quality evidence level 2+)⁸. The forms of debridement can include self-management using an emery-board/nail file. If the area is too painful or too thick then patients need to be seen by a Podiatrist for a blade/scalpel debridement. However, in dealing with EB patients Podiatrists are advised to be more conservative in their approach as over-debridement can make the underlying skin susceptible to increased blistering and tenderness. After debridement careful use of emollients and non-adherent dressings should be used to protect the debrided skin.
- **Practical point;** in the experience of the panel, many EB patients have reported bad experiences regarding over-debridement from Podiatrists who have not understood the nature of the condition or sought advice from the patient themselves. This is why Podiatry education in EB has been highlighted as a priority by DEBRA and a specific training programme is currently being developed.

- Assessment and correction of weight distribution, with cushioning to try and reduce hyperkeratosis build up (Quality evidence level ranged from 2- to 2+)^{4,7-8}
- The use of EB-specific assessment tools(s) (eg pressure assessment platforms and Foot Health Status Questionnaire; Appendix 2, section B) which assess the distribution of pressure on the skin leading to hyperkeratosis and evaluating how best to manage the condition by further assessing the quality of everyday function being carried out (Quality evidence level ranged from 2- to 1-)^{4,7-8,10}.

Special considerations:

- Heloma (corn): The common corn is Heloma durum. Heloma miliare (seed corns) are frequently seen in EB due to toe and foot deformity. Heloma neurovascular are encountered but to a lesser degree (Quality evidence level 4 Expert opinion).
- Neurovascular hyperkeratosis: This is a form of callus in which nerve endings and blood vessels become prominent in the epidermis in response to trauma and treatment. This condition can present in EB patients (estimated < 1%). The area is sensitive, painful and difficult to treat as normal debridement causes pain and bleeding. Although rare it is very debilitating in the small number of EB patients affected. It probably results from long standing gross hyperkeratosis and usually affects skin overlying the calcaneum and hallux (Quality evidence level 4 Expert opinion). Debridement of these lesions by a Podiatrist is also recommended and can provide similar relief but the patient should be advised that due to the nature of the lesion, treatment can often be more uncomfortable than is experienced with standard hyperkeratosis.

Footwear

We recommend suitable footwear and appropriate insoles/orthotics to manage the EB foot to podiatrists, patients, carers and healthcare professionals (*Strength of recommendation Grade: C*)

Evidence that advice on footwear is beneficial has been reported in all subtypes of EB: EBS^{4,5,8}, JEB^{4,8}, DDEB^{8,21}; RDEB^{7,8}.

EB footwear advice suggests that wherever appropriate, footwear should be supportive. Its primary focus should be aimed at minimizing blistering by reducing friction (Quality evidence level ranged from 3 to 2-)^{3,5,7}. Once blisters have formed, the use of dressings and topical antiseptics or antibiotics may be used to prevent secondary infection until the wound heals (Quality evidence level ranged from 4³ to existing guideline^{⇒9}). Therefore suitable shoe/footwear is essential to accommodate dressings and not lead to further trauma to the damaged area. Footwear which is adjustable maybe beneficial in these circumstances.

Recommendations regarding footwear in EB are based on expert opinion since evidence is lacking. The overriding recommendation is to minimize mechanical trauma to the feet by emphasising the need for suitable footwear and appropriate insoles/orthotics (Quality evidence level 2-)⁴.

Footwear advice should address the following:

- Socks should be selected to improve ventilation. Silver fibred cotton socks and silver vinyl covering (for example Coolsorb) can be used with simple insoles and orthoses (Appendix 3). These conduct heat away from the feet, reducing sweating and friction (Quality evidence level ranged from 4 to 2-)⁴. Silver fibre socks also have an additional anti-bacterial action (Quality evidence level 2-)⁴. Silver socks technology is readily available across the world through hiking and trekking products. Additional options for patients living with EB include bamboo socks and double layer socks.
- Shoes should ideally have the following features: firmness (Appendix 4), comfortable fit, appropriate length and width, rounded toe, plenty of room for the toes, flexibility, flat heel, heel support, laces or straps, to prevent excessive movement or slipping of the foot inside the shoe and flat or absent seams (Quality evidence level 4)⁴. The upper covering should be leather or fabric mesh to allow air to circulate, rather than plastic or synthetic (Quality evidence level 2-)⁴. Shoes (Quality evidence level ranged from 4 to 2+)^{3,8}.

The gray literature supports this by showing how different types of cushioning materials and insoles provide benefit in non-related Diabetic foot ulcers. This can have a subjective relevance to support the benefits of footwear and orthotics in EB¹⁶⁻¹⁸.

Special considerations

- Care must be taken when a child starts walking, acknowledging that shoes are not always necessary indoors (Quality evidence level 4)³. Allowing a child to walk barefoot or just in socks helps feet to grow normally and develops muscular joint strength. A child will also benefit from proprioceptive feedback when walking barefoot (Quality evidence level 4)³. These benefits must be balanced against the risk of damage to the unprotected skin. Outside, children's feet should be protected in lightweight flexible footwear made of natural materials (Quality evidence level 4)³. The soft cartilage within their feet can easily be bent whilst walking and the layer of fat tissue will offer support and shock absorption, potentially masking abnormal development (Quality evidence level 4)³.
- Babies with EB may have one foot smaller than the other due to prenatal loss of skin and subcutaneous tissue: this can be managed by correctly fitting shoes of different sizes (Quality evidence level 4)³. The child's foot should be measured at a reputable shoe store every 2 to 4 months (Quality evidence level 4)³, or at the EB clinic, and it may be necessary to change the shoes and the socks every few months to allow room for growth (Quality evidence level 4)³. Children with severe types of EB needing bulky dressings to the feet may have difficulty finding shoes to fit and rely on lightweight plastic shoes such as Crocs. (Appendix 4).
- All severe EB patients would not be walking barefoot but would have layers of protective dressings. Special consideration should be given to the length of the Achilles tendon, which can tighten in response to pain on walking and application of dressings.
- Cost implication for appropriate footwear is a large consideration in the global EB world. Some countries may use specific shoe funded services for "suitable" patients, whilst others are unable to provide such a service. Recommending suitable footwear can have its limitations and cost implications due to the type of footwear available, the age of the individual, their foot deformity, type of EB they have, their occupation, and changes in fashion. Not all services will cover or have

international availability. However, the improvement in mobility, independence and quality of life in the long term outweigh this (Quality evidence level 2-)⁷. (Appendix 4).

Mobility

We recommend measures to improve mobility, with assessment and monitoring for all subtypes of EB (*Strength of recommendation Grade: C*)

- Longitudinal mobility assessment and monitoring is essential in EB as disease-related factors such as scarring and contractures can change an individual's ability to mobilise over time (Quality evidence level ranged from 4 to 2+)^{3,5,7,19-20}
- Gait-analysis systems can be safely used in EB and can be helpful to diagnose pressure areas, walking patterns and evaluate the effect of therapeutic interventions (Quality evidence level ranged from 2- to 2+)⁶⁻⁷
- Podiatry interventions differ between EB subtypes and must be tailored to the individual patient to prolong mobility (Quality evidence level ranged from 4 to 3)^{3,5}

The evidence of podiatric care can improve mobility. Both children and adults of all subtypes of EB may have affected feet¹⁸. Problems such as blistering, hyperkeratosis (callus), nail loss, altered gait and deformity (Quality evidence level ranged from 3 to 2+)^{8,20-21} can result in reduced mobility and eventually, wheelchair use (Quality evidence level ranged from 3 to 2+)^{8,21}. Preventing these problems can help children and adults of all subtypes of EB to stay mobile for longer and improve their quality of life.

A study of 425 EB patients of whom 140 were children reported the percentage of children being able to walk independently compared to dependently within the different EB subtypes (Fig. 2)¹⁹. Their results showed children who were independent had differing walking abilities¹⁹. It was noted that DDEB showed a higher percentage of independent walking compared to EBS, JEB and RDEB¹⁹. The percentage of patients in the dependent group showed very little ability for walking with RDEB requiring the most support compared to EBS and JEB, with DDEB requiring no support (Fig. 2)¹⁹. The impact of dependency should not be diminished by these results as the use of occasional wheelchairs is present amongst all forms of EB¹⁹.

Painful foot blistering is a common problem exacerbated by walking or standing in EB (Quality evidence level ranged from 3 to 2+) ^{6,19,22-23}. A DDEB study reported that pain on walking was reduced in 6 of the 7 patients by correcting the foot's balance and eliminating areas of abnormal weight bearing (Quality evidence level 3) ²². In a more recent EBS-I cohort localized foot pain occurred before, during or after the onset of a blister (Quality evidence level 2+) ⁶. Blisters triggered by friction, walking, heat, trauma and hyperhidrosis tend to be worse in the summer (Quality evidence level 2+) ⁶.

Practical point; A case controlled study reported that plantar injections of botulinum toxin effectively reduced pain from walking, and was a long-lasting and safe treatment for painful blistering and callosities in EBS (Quality evidence level 2-) ²³. Blisters disappeared after botulinum toxin therapy and the pain reduction was sufficient to permit the patient to start walking more freely (Quality evidence level 2) ²³. However, the procedure is painful and not tolerated by all patients (Quality evidence level 2-) ²³.

The gait analysis/pressure measurement systems assess foot step pattern ² (Quality evidence level 2-) ⁷ (Appendix2; Section C). Analysis of a person's manner of walking (gait) in EB facilitates the diagnosis and appropriate management of foot problems ³ (Quality evidence level 2-) ⁷. These platforms have proved essential in podiatry practice in diabetic foot management. The podiatrist can use gait assessment to identify areas which have more focused pressure when walking and translate this information into the development of patient specific insoles (Appendix2; Section C). This practice can also be extended, where appropriate in the development of bespoke footwear. This is particularly useful when the type of EB results in such deformity, that standard off the shelf footwear will not fit ³ (Quality evidence level 2-) ⁷. The physiotherapist can also use gait analysis/assessment, to help improve posture while walking and provide further advice/exercises to help develop core strength (Quality evidence level 4) ²⁴. Furthermore this approach can evaluate the effectiveness of the therapy (Quality evidence level 4) ²⁴. From a practical view the Gait platform mats are portable and can be fun to use and above all do not damage the skin (Quality evidence level 4) ²⁴.

The overarching recommendations here are:

- Referral for podiatry assessment, treatment and monitoring to minimise blisters and pain while walking
- Where appropriate/affordable, consideration of patient specific insoles and bespoke shoes
- Multidisciplinary therapy (MDT) management to include the podiatrist, occupational therapist and physiotherapist to reduce pain while walking and encourage mobility (Quality evidence level ranged from 4 to 2++)^{25,28}
- Use of a gait analysis system to assess the patient's walking pattern, monitor and evaluate therapy intervention (Appendix 2; Section C)
- Plantar injections of botulinum toxin have been highlighted as providing therapeutic benefits in a small cohort of patients. Further research is required in this area but may be a consideration for EBS adult patients who can tolerate the procedure, if conservative therapies have not worked.
- Tailoring podiatry interventions to the subtype of EB to prolong mobility.
 - EB Simplex patients tend to require debridement of hyperkeratosis (callus), blister care management, or simple insole and footwear advice
 - JEB patients may require blister and wound management, also simple insoles and footwear advice
- All EB patients require nail management from birth, plus wound care and footwear advice and insoles as they become older

Pseudosyndactyly

We suggest consideration of surgery for pseudosyndactyly and mitten deformities of the feet as well as contractures of the lower extremities in patients with DEB

(Strength of recommendation Grade: D)

- Foot surgery for joint contracture deformity release and digital amputation can be successfully performed in EB and may benefit patients by reducing pain, improving ability to wear shoes and improving mobility (Quality evidence level 3)^{22,29-32}

The evidence for the surgical management of pseudosyndactyly, mitten deformities and contractures of the lower extremities largely concerns patients with DEB. The relevant literature is heterogeneous and consists mainly of case reports and case series. Surgery in this patient cohort is primarily undertaken in the hand to improve function. Surgery can be considered for pseudosyndactyly, mitten deformities and contractures of the lower extremities in patients with DEB but due to the relative short term nature of any cosmetic improvement and limited functional improvement (if any) then other surgical procedures that will help accommodate the foot in certain footwear, such as selective digital amputation may be more appropriate. Patients should therefore consider any benefit versus the risks very carefully. Similar complications from other EB subtypes do not cause mitten deformities/pseudosyndactyly deformities and are therefore not highlighted in any of the literature (Appendix 1a).

RDEB-GS is characterized by progressive fusion of digits leading to pseudosyndactyly and a mitten-like deformity of the hands and feet. These complications occur to a lesser degree and later in RDEB GI. Cutaneous scarring can also lead to joint contractures and deformities in feet resulting in reduced mobility and pain (Quality evidence level 3)^{22,29-31}.

Procedures reported

- Clawed toes were surgically released in 3 patients, by making extensive transverse incisions across the dorsal and/or plantar surface of the toes and distal forefoot, extending into the subcutaneous tissue (Quality evidence level 3)²². This improved the foot contour, reduced pain on walking and allowed shoes to be worn (Quality evidence level 3)²². The release of pseudosyndactyly makes it easier to wear normal footwear and it is of psychological benefit to the patient to observe a “normal” foot with five toes (Quality evidence level 3)²². Improvements were seen for several years but due to the progression of the EB reoccurrence was likely.
- Despite the long term complications of surgery, early extension procedures to address contractures of the toes, equinus and cavus deformities using soft tissues surgery was recommended by the experts reviewing 6 cases (Quality evidence level 3)³⁰.

- In a case study of foot syndactyly in 6 patients with RDEB, the main foot surgery goal was to reduce extreme flexion or extension contractures to allow the patient to wear shoes and ambulate comfortably (Quality evidence level 3)²⁹.
- Most surgical procedures to the foot in EB involved mitten release, although this procedure is used more for hands (Quality evidence level 4)³². Syndactyly release to the foot reduced pain or difficulty in standing and walking, and the inability to wear shoes due to hyperextension contractures of the toes (Quality evidence level 4)³². The numbers are small with only 6 patients benefitting out of 25 who had surgery in a New York cohort, and 6 of 50 patients in the St. Thomas Hospital cohort, who underwent foot surgery (Quality evidence level 4)³².
- Contractures almost always recur, because of the lack of long-term benefit and patients often refused further surgical interventions (Quality evidence level 3)³³.
- RDEB mice models shown Losartan reduced TNF- β mediated inflammation and supported matrix remodelling. The RDEB mice with injured fore paws administered with Losartan seems to prevent digit fusion (Quality evidence level 2++)³⁴. Clinical trial to establish safety, tolerance and efficacy of losartan in children with RBED is currently ongoing³⁵.

Podiatrist Professional Development

An enhanced proficiency in the functional treatment of people with EB is recommended. It is expected that clinicians always use great care as an integral part of their professionalism as a podiatric clinician. However, EB is a condition requiring specialist intervention beyond just 'being more careful'. It necessitates specialist training and provision and a recognition of podiatric practitioners of the extent of their scope of practice and experience. This has important ramifications for undergraduate podiatry training in relation to informed knowledge of inter-professional referral pathways in instances where newly qualified students may inadvertently meet patients with EB for the first time.

Podiatrists managing EB must avoid causing secondary injury, by:

- Handling feet and limbs with great care
- Avoiding the use of highly adhesive tapes, dressings and felt padding

- Removing any adherent dressings, ideally with silicone spray

Continued professional development for podiatrists is encouraged, for example undertaking a specialist EB podiatric management course to integrate their professional knowledge and clinical skills in managing EB-related podiatric conditions.

The guideline is focused on helping people living with EB and their families to manage their foot problems. In countries where podiatrists are unavailable or not part of the healthcare system, a healthcare professional can aid to some degree. Nurses can offer expertise with wound care management, and they can offer advice on footwear and foot-care with the guideline as a reference. There are restrictions with blade debridement as this is not part of their scope of practice. Specialist podiatric knowledge to address biomechanical issues of the feet and prescription orthotics can only be delivered by a podiatrist, physiotherapist or musculoskeletal doctor. These key areas may also be addressed by a dermatologist, paediatrician, surgeon or “doctor”. In countries where there is no podiatrist support, the healthcare professional offering this care should make sure that they legally adhere to that country's scope for their profession.

Key Limitations

An extensive literature review of syndactyly surgery revealed mainly case studies with few controlled studies. Recommendations are therefore based on expert opinion of current clinical practice.

Conclusion

We can conclude that podiatric intervention improves EB foot-care. The key intervention of clinical debridement of hyperkeratotic (callused) skin, dressings of wounds and reduction and cutting of nails greatly improved the well-being of patients. Advice given by the podiatrist helped patients to identify suitable footwear, insoles and socks, benefitting patients on a daily basis. A podiatrist should routinely be included as part of the multidisciplinary management of EB.

Future Research

This CPG highlights the need for further high quality research (Table 2).

Implementation of Guideline recommendations

DEBRA International aims to ensure that the EB guidelines address the needs of patients internationally. These guidelines will be translated into other languages and a patient version will be made to make them more accessible. These guidelines could be disseminated and promoted through the education of professionals, and eventually incorporated into clinical practice. This guideline was presented at the DEBRA Australia EB camp 2018. The implementation of these recommendations could be monitored and evaluated through audits, education programme registration, and the CPG Evaluation Form: Pre implementation (Appendix 8).

Development of the guideline and methodology used for formulating the recommendation

In 2016, an International panel of multidisciplinary health professional and people living with EB was coordinated through DEBRA International (DI), through a voluntary membership. The panel represented clinical or personal experience of EB covering both specific adult and paediatric knowledge bases. All panel members were encouraged to actively participate in all stages of the guideline development, so that the co-construction of knowledge and experience of the condition could be seen to move beyond tokenism in relation to the incorporation of the expert patients who live with EB on a daily basis.

Following the SIGN³⁶ methodology the panel decided on the clinical question, “Can podiatry support help improve the quality of life of people living with EB?” and used this to focus their search through considering participants, interventions, comparisons, and outcomes (Fig. 3) (“PICO”)³⁶⁻³⁷. This process was informed by priorities raised by people living with EB from an international survey using DEBRA International, EB-CLINET databases and distribution of hard copies of the survey in clinics in Australia (Appendix 6), and a preliminary literature search. The panel voted for the relative importance of the outcomes and selected the top 6-7 priorities which matched those raised from the survey³⁶⁻³⁷.

Literature search

A systematic literature search regime was adopted with no language restrictions. The literature search was conducted by the two panel leads using seven electronic search engines: Medline (PubMed MeSH), Wiley online library, Google Scholar, Athens, Researchgate, Net and pubfacts.com. The search terms and inclusion criteria utilised PICOS (Fig. 3). The boolean AND and OR operators were used to combine these terms as appropriate (Fig. 3). Cited reference searches were conducted on eligible papers. Updating of the available literature was continued up to publication.

Criteria of inclusion applied to all articles identified by the searches (Appendix 7). These were discerned from the papers' abstract and title, or the full articles in cases of uncertainty. Papers which were unpublished or did not meet the methodological filters were retained as gray literature. These were examined to provide context or considered divergence within the main recommendations.

Research Appraisal

All published papers meeting this filtering stage were then subject to a systematic quality appraisal and risk of bias assessment. This appraisal was modified from the Critical Appraisal Skills Programme³⁸ and SIGN³⁶ quality rating. This allowed both quantitative and qualitative research to be appraised using one list of questions, yielding one quality rating scale to allow a comparison of studies as required (Appendix 7). The study limitation and indirectness were taken into account through the appraisal tool. The precision and statistical consistency could not be evaluated as the EB articles had no statistical values. Most studies reviewed had more than a 50% risk of bias as EB is a rare condition, there are no double-blind randomised clinical studies and most people would know they have EB.

All selected papers were filtered and appraised by the two panel leads (MTK and MOS). In those instances where consensus could not be reached between the two panel leads, a third appraisal from the panel was allocated until this could be assured. This was conducted to reduce bias, increase content validity checks of the literature and most importantly to ensure the consistency of the reviews undertaken. The research quality score was obtained with a high percentage being indicative of the higher quality of the

paper. Levels of bias were also measured in percentage values and all papers were graded in accordance with the SIGN method “Level of evidence and Grades of Recommendations” 1++ to 4 and Grade A to D³⁶.

The papers were then divided into outcome topics. All papers and gray literature were allocated to these outcomes. The two panel leads and a member summarised the appraisals per outcome and rated the strength of the recommendation. Outcome summary tables were presented to highlight the population subtypes, numbers of subjects, study type, percentage quality and risk of bias in accordance with SIGN. The panel checked the emerging strength of the recommendation, desires and undesired effects, costs related to benefits and the feasibility of implementation. They confirmed and discussed the recommendations elicited using the Grading of Recommendations Assessment, Development and Evaluation (GRADE) framework for the recommendation table³⁷. All recommendation summaries were circulated to the panel and final agreement and feedback were included. The Appraisal of Guidelines for Research & Evaluation (AGREE) II tool⁴⁰ was consulted to increase the quality of practice guidelines in rare diseases and this CPG acknowledges existing guidelines by signposting with the symbol \Rightarrow through this manuscript.

The guidelines were peer-reviewed by a representative cross-section of EB MDT specialists and people living with EB. Five out of 8 health professionals and 1 person living with EB reviewed the guideline draft to assess the degree to which the recommendations presented addressed patients’ concerns and identified good practice points (Table 4).

The lead and co-lead compiled a reviewers’ feedback report for discussion with the guideline panel. Each point was addressed and any resulting change to the guideline was noted or, if no change was made, the reason for this was recorded. The panel conducted a final proof read of the manuscript before submission.

Implementation of Guideline Recommendations

The implementation of these recommendations could be monitored and evaluated through audits, education programme registration, and the “CPG Evaluation Form: Pre

implementation" [Appendix 7]. The panel recommends clinical sites to conduct pre-practice audit, implement the CPG and re-audit to test improvements. Audit tools can be used from SIGN³⁶. DEBRA International would value your feedback on the site findings to continue to improve CPG quality.

Guideline Dissemination and Update

DEBRA International is aiming to ensure that the EB guidelines address the needs of patients internationally. These guidelines will be translated into other languages and a patient version is planned to make them more accessible. The guidelines will be updated every 3-5 years or if there is a significant breakthrough in EB podiatry care from the publication date. We recommend iterative updating of search terms to see if a full review is warranted at any stage.

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Table 1. Summary of key recommendations for podiatry management of EB foot and nail disorders

Table 2. Possible future research per outcome

Table 3. Overview evidence per outcome

Table 4. Roles of panel and reviewer list

Table 5. Abbreviations list

Figure 1. EB foot manifestations

Figure 2. Percentage of EB children who reported independent and dependence for major activities for daily living¹⁹

Figure 3. Search terms and inclusion criteria

Figure 4. Flow of papers searched and appraised

Appendix List

1. A) Images of related foot and nail disorders with children and adults with Epidermolysis Bullosa
B) Hand Dystrophic nails management
2. Assessment tools
3. Foot-care devices and products
4. Footwear and hosiery products for children and adults
5. Bespoke footwear information
6. EB Survey
7. Appraisal and risk of bias tool
8. Implementation survey

a) Key recommendation	Grade strength of recommendation	Quality of evidence (Rate Average)	Key references
Desirable consequences clearly outweigh undesirable consequences in most settings for this reason we recommend offering these options:			
Avoidance of blistering and wounds: a podiatry education programme should be offered from birth, enabling carers, patients and staff to recognise and avoid causes of blistering and wounds, including <ul style="list-style-type: none"> • Footwear • Dressings • Foot biomechanics • Heat and sweating 	B	2+	3- 10*
Management of dystrophic nails: podiatric support can include <ul style="list-style-type: none"> • Topical keratolytics • Trimming, reducing or removing nails 	B	2+	3-4, 8, 12-15
Management of hyperkeratosis (callus): podiatric support should include <ul style="list-style-type: none"> • Assessment and monitoring of weight distribution • Appropriate cushioning to prevent hyperkeratosis • Use of a validated assessment tool (Appendix 2) 	B	2+	3-5, 7-8, 10*-11
Footwear advice: information should be provided regarding suitable shoes and the appropriate use of <ul style="list-style-type: none"> • Insoles • Cushioning materials • Orthotics 	C	3	3-5, 7-9, 21
Assessment and monitoring of mobility: podiatric care should focus on maintaining mobility, adapting to the specific needs of different subtypes and different age-groups, within an MDT	C	3	3-9, 18-28

The balance between desirable and undesirable consequences were uncertain for this reason we suggest consideration of this option:				
Assessment of pseudosyndactyly and contractures: podiatric support should include:				
<ul style="list-style-type: none">Advice on preventative measuresAssessment of functional impairmentReferral for surgical correctionPost-operative management to prevent recurrence and promote mobility	D	3	22, 29-34	
Key * Articles with no EB population				
bi) Grades Descriptions in accordance to SIGN ³⁶				
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 1++ or 1+			
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++			
D	Evidence level 3 or 4; or Extrapolated evidence from studies rated as 2+			
bii) Rate Descriptions in accordance to SIGN ³⁶				
2++	High quality systematic reviews of case control or cohort studies; High quality case control or cohort studies with a very low risk of confounding or bias and a high probability that the relationship is causal			
2+	Well conducted case control or cohort studies with a low risk of confounding or bias and a moderate probability that the relationship is causal			
3	Non-analytic studies, e.g. case reports, case series			
biii) ✓ Recommended best practice based on the clinical experience of the guideline development group ³⁶				
Notes:	There was no disagreement on the quality of the appraised articles or the strength of the recommendations.			

Blistering and wound management

- Comparative studies can be used to assess dressing types used on the feet on different EB groups.

Dystrophic nails

- Evaluation to assess the benefit for a podiatrist to manage both finger and toenails.
- A review of the nail conditions affecting EB patients is needed and then a study to examine the treatment protocols: with topical keratolytic agents, urea-based agents and daily filing with an emollient to follow.

Hyperkeratosis (Callus)

- Evaluate the benefits of Callus debridement between manual techniques (scalpel) over keratolytic agents.
- Comparative studies assess different keratolytic agents when treating hyperkeratosis in EB Patients.

Footwear

- Examine different podiatry materials to offer shock absorption and redistribution within footwear being worn
- Studies on footwear for EB patients and engaging with footwear and hosiery manufactures to make friendly footwear and hosiery s more accessible for EB sufferer.
- Evaluation of specific footwear funding by the service for "suitable" patients, the outcome of this would be useful
- A study would be required to show any quantifiable benefit of silver vinyl insole material.

Mobility

- Further assessment with larger EB groups monitoring mobility using gait analysis platforms and fitbits to assess total distances achieved (patients to record their steps just using their mobile phones. It's not as accurate as a fitbit but less expensive and also not everyone can wear something around their wrist)
- Assessing the impact aids, suitable footwear, and insoles/orthotics and dressings have on aiding distances achieved by EB individuals

Pseudosyndactyly

- Benefit of no surgical implementation of losartan in slowing down fibrosis in RDEB patients

Other areas

- Botox injections in EBS
- Pedagogical implications for the contextual positioning of EB education and training in both undergraduate and CPD/postgraduate podiatric specialisms.

Outcome	# allocated papers	Participants with EB in the articles	Methodology	Average quality rate	Quality appraisal (range)	Benefits and limitations
Blistering and wound management	6	347* EBS 171 JEB 11 DDEB 31 RDEB 22	1 qualitative 1 quantitative 1 cohort 2 case studies 1 chapter	2+	58% (52-86%)	Blisters can be reduced in size and frequency of new blisters forming but the know how is still limited to a few centres
Dystrophic nails	8	234* EBS 137 JEB 11 DDEB 38 RDEB 24	2 qualitative 1 quantitative 3 case studies 1 observational 1 chapter	2+	67% (17-90%)	Mainly toenails rather than fingernails and their use for diagnosis
Hyperkeratosis	5	286* EBS 137 JEB 11 DDEB 33 RDEB 22	1 qualitative 1 quantitative 2 case studies 1 chapter	2+	58% (52-64%)	Highlights occurrence in clinic not complexity
Footwear	6	291* EBS 114 JEB 11 DDEB 31 RDEB 22	1 qualitative 1 quantitative 1 cohort 2 case studies 1 chapter	3	56% (48-69%)	Mainly on advice no audits
Mobility	14	1067* EBS 396 JEB 71 DDEB 148 RDEB 105	3 qualitative 2 quantitative 1 cohort 3 observational 4 case studies 1 chapter	3	60% (48-90%)	Early stages of new approaches to assess and treat.
Pseudosyndactyly	8	3401* DEB 96	Out of 96 DEB only 7 DEB cases were on toe fusion 1 laboratory biological and animal models	3	54% (24-95%)	Low evidence with only case reports/series of poor quality and high risk of bias.
Key	EB: epidermolysis bullosa; RDEB: recessive dystrophic epidermolysis bullosa; JEB: Junctional epidermolysis bullosa; DDEB:					

dominant dystrophic epidermolysis bullosa; EBS: Epidermolysis bullosa simplex; KS Kindler syndrome; n: number of; *total number of persons with EB in all papers combined

Rate

Descriptions in accordance to SIGN³⁶

2+

Well conducted case control or cohort studies with a low risk of confounding or bias and a moderate probability that the relationship is causal

3

Non-analytic studies, e.g. case reports, case series

CPG working Panel

Panel member	Country of origin	Speciality with EB	Role in panel
Lisa Brains	Australia	Person living with EB	Member
Beata Faitli	UK	Person living with EB-Mum	Member
Rodney Fawkes	UK	Podiatrist	Member
Michael Fitzpatrick	Australia	Person living with EB-Dad	Member
Adam Harris	Australia	Dermatologist	Member
Lynne D Hubbard	UK	Senior specialist dietitian for adults with EB	Member
Laura Iacobaccio	Australia	Podiatry	Member
Lisa James	UK	Podiatry	Member
M Tariq Khan	UK	EB Podiatrist	Lead
Jemima Mellerio	UK	Professor and Consultant	Member
Mark O'Sullivan	UK	Podiatry	Co-lead
Tracey Vlahovic	USA	Dr Podiatric	Member
Michelle Wood	UK	Physiotherapist specialised in EB	Member
Katty M Mayre-Chilton	UK	CPG Coordinator and Research Dietitian	Member

Reviewer Panel List

Country of origin	Title
Norway	Person living with EB
UK	Reader in Health Professions Pedagogic Practice
UK	MD Consultant
UK	Consultant Dermatologist
Australia	Professor and Dermatologist
Australia	Consultant Dermatologist

Key: UK- United Kingdom; USA- United States of America; EB- Epidermolysis bullosa; DI- DEBRA International

AGREE	Appraisal of Guidelines for Research and Evaluation
CASP	Critical Appraisal Skills Programme
DDEB	Dominant Dystrophic Epidermolysis Bullosa
DEBRA	Dystrophic Epidermolysis Bullosa Research Association
EB	Epidermolysis Bullosa
EB-CLINET	Epidermolysis Bullosa Clinical network
EBS	Epidermolysis Bullosa Simplex
EBS-I	Localized form of Epidermolysis Bullosa Simplex
GRADE	Grading of Recommendations Assessment, Development and Evaluation
JEB	Junctional Epidermolysis Bullosa
KS	Kindlers syndrome
PICOS	Participants, interventions, comparisons, outcomes and study design
QoL	Quality of life
RDEB	Recessive Dystrophic Epidermolysis Bullosa
SIGN	Scottish Intercollegiate Guidelines Network
UK	United Kingdom

Podiatry Guideline: Figure 1

Primary types of EB	Blistering and scarring	Dystrophic nails	Hyperkeratosis, callus and corns	Pseudosyndactyly and mitten deformities
EB Simplex	Yes		Yes	
Dominant Dystrophic EB	Yes	Yes	Yes	
Recessive Dystrophic EB	Yes	Yes		Yes
Junctional EB	Yes	Yes		
Kindler Syndrome	Yes	Yes		

Podiatry Guideline: Figure 2

Walking Independent Dependent	ES	JEB	DDEB	RDEB
	31.2	30.8	66.7	24.4
	2.1	7.7	0	13.3

Podiatry Guideline: Figure 3

Population: Epidermolysis Bullosa, rare diseases

Intervention: Podiatry

Compared: to no service available for podiatry management





Outcomes: Blistering; wound, footwear, hosiery, nails, hyperkeratosis, callus, mobility, fusion of toes, pseudosyndactyly, mitten deformities, mitten contractures

Studies: Any design. No language restrictions. From 1 January 1979 to 25 October 2018 and inclusive.

Podiatry Guideline: Figure 4

Number of papers	
46	Full articles
10	duplicates
36	Selected articles

Images of related foot and nail disorders with children and adults with Epidermolysis Bullosa	
Outcome	Type of EB
Blistering	
	a. Generalized intermediate junctional EB: Scarring
Feet dystrophic nails	
	b. EB Simplex: Loss of nails
	c. Generalized severe EB simplex: (former EB simplex Dowling Meara)

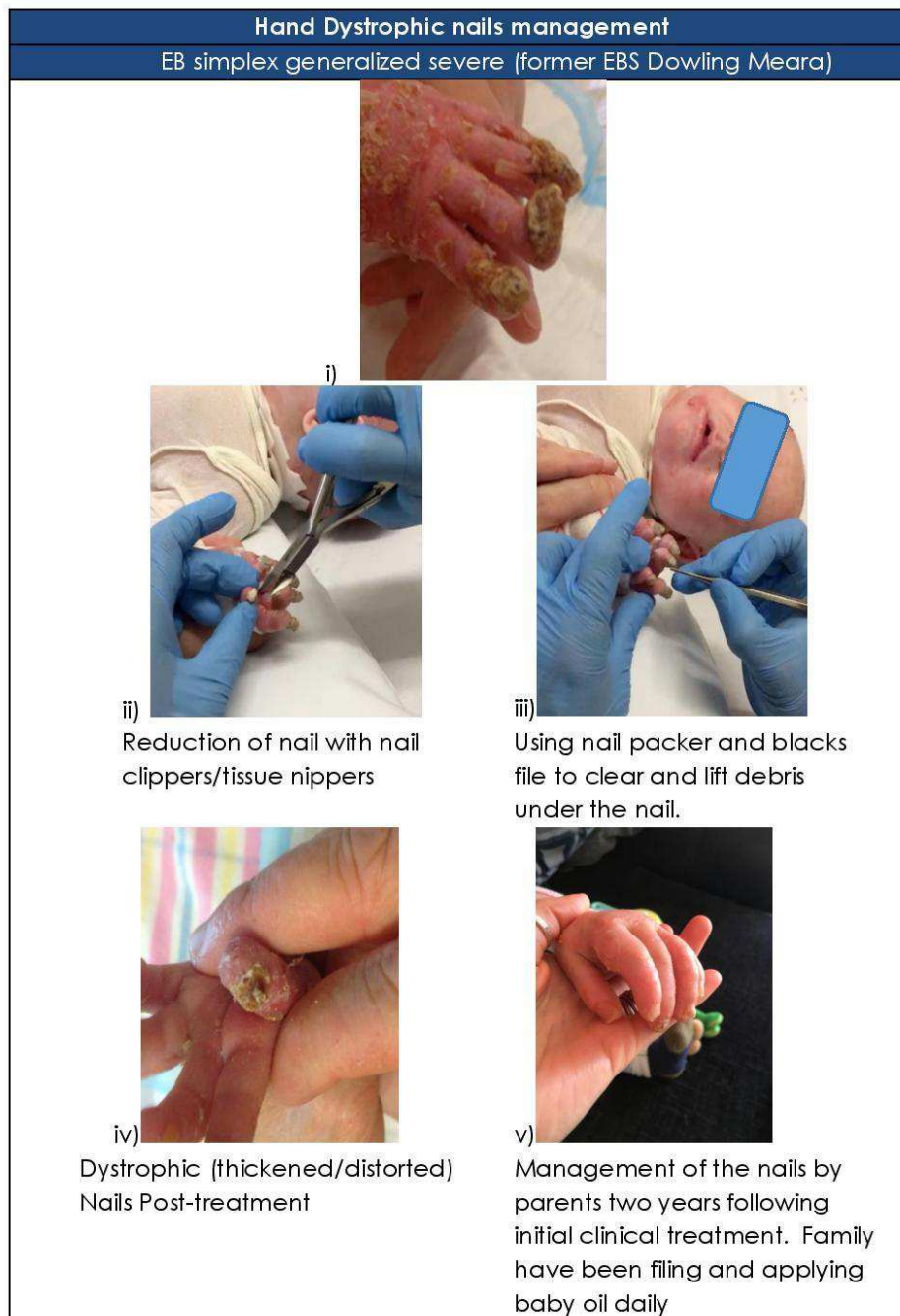
	<p>d. Generalized intermediate EB simplex: Loss of nails</p>
Hyperkeratosis/ callus	
	<p>e. EB simplex</p>
	<p>f. Generalized intermediate junctional EB</p>
	<p>g. Generalized intermediate EB simplex</p>

Fusion of toes (pseudosyndactyly)

h. Recessive dystrophic epidermolysis bullosa: pseudosyndactyly



i. Chronic wound in Recessive dystrophic epidermolysis bullosa



Podiatry CPG: Appendix 2

Foot assessment tools

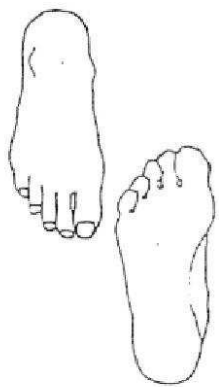

A) EB Podiatry assessment form

Name		DOB	
Address		Sex	
Tel.No		Email	
GP		Age	

Past Medical History & Drug History

--

Feet Assessment

<p>Right</p> 	<p>Left</p> 
<p>Comment</p>	<p>Comment</p>

Dr M Tariq Khan, Dr Parita Patel and Prof Dedee F Murrell



Podiatry CPG: Appendix 2

Hands Assessment	
Right	Left
Comment	Comment

	Hands	Feet
Hyperkeratosis, calluses		
Keratoderma		
Nail abnormalities		

Treatment Plan			
Foot care advice	<input type="checkbox"/> Foot cleanliness	<input type="checkbox"/> Foot exercise	<input type="checkbox"/> Carefully fitted shoes
Foot wear	<input type="checkbox"/> Functional orthotics	<input type="checkbox"/> Insoles	<input type="checkbox"/> Silver lined socks
Medication	<input type="checkbox"/> Debridement	<input type="checkbox"/> Marigold tagetes	

	Initial	Visit 1	Visit 2	Visit 3
Date				
Appearance				

Dr M Tariq Khan, Dr Parita Patel and Prof Dedee F Murrell

Podiatry CPG: Appendix 2

EB Podiatry Questionnaire

Age of onset of blisters on feet?	
Age of starting to walk?	
If you use a wheelchair, how many hours a day do you spend on your feet?	
Do you have pain on walking?	
Do you get painful blisters after walking?	
Where on your feet are most your blisters?	
Do you use any walking aids? If so, what?	
How do you deal with blisters? Do you pop them for example?	
What previous treatment have you had for your feet?	
Have you seen a podiatrist before? If so, did they help?	
Do you wear specialised footwear? If so, what?	
Would you like to have more help with your feet?	

My feet make me feel:



Ultra happy



Happy



No different



Slightly worse



Miserable

Any other comments:

Dr M Tariq Khan, Dr Parita Patel and Prof Dedee F Murrell



Podiatry CPG: Appendix 2

B) International foot health status questionnaire

**THE FOOT HEALTH STATUS
QUESTIONNAIRE**



Thank you for taking the time to fill out this important questionnaire.

The answers you provide will help your podiatrist to understand how to care for your foot problems.

The questionnaire is very simple to complete and there are no right or wrong answers. The questionnaire takes less than 10 minutes to complete.

Foot health status questionnaire Version 1.04:
Analysis is available from www.fhsq.org and supported by Microsoft Windows™

The Foot Health Status Questionnaire

INSTRUCTIONS

- This questionnaire asks for your views about your foot health.
- All you need to do is circle your answer to each question.
- If you are unsure about how to answer a question, please give the best answer you can.

The following questions are about the foot pain you have had during the past week.

1. What level of foot pain have you had during the past week ?

(circle number)

- None..... 1
- Very Mild..... 2
- Mild..... 3
- Moderate..... 4
- Severe..... 5

(circle a number for each question below)

DURING THE LAST WEEK...

	Never	Occasionally	Fairly Many Times	Very Often	Always
2. How often have you had foot pain ?	1	2	3	4	5
3. How often did your feet ache?	1	2	3	4	5
4. How often did you get sharp pains in your feet ?	1	2	3	4	5

Foot health status questionnaire Version 1.04: Analysis is available from www.fhsq.org and supported by Microsoft Windows™



Podiatry CPG: Appendix 2

These questions are about how much your feet interfere with activities you might do during a typical day.

(circle a number for each question below)

DURING THE LAST WEEK.....

	Not at All	Slightly	Moderately	Quite a bit	Extremely
5. Have your <u>feet</u> caused you to have difficulties in your work or activities ?	1	2	3	4	5
6. Were you limited in the kind of work you could do because of your <u>feet</u> ?	1	2	3	4	5

DURING THE LAST WEEK...

	Not at All	Slightly	Moderately	Quite a bit	Extremely
7. How much does your <u>foot health</u> limit you walking ?	1	2	3	4	5
8. How much does your <u>foot health</u> limit you climbing stairs ?	1	2	3	4	5

9. How would you rate your overall foot health ? (circle number)

- Excellent..... 1
 Very Good..... 2
 Good..... 3
 Fair..... 4
 Poor..... 5

Please turn to the next page

Foot health status questionnaire Version 1.04: Analysis is available from www.fhsq.org and supported by Microsoft Windows™



Podiatry CPG: Appendix 2

The following questions are about the shoes that you wear. Please circle the response which best describes your situation.

	Strongly Agree	Agree	Neither Agree nor Disagree	Disagree	Strongly Disagree
10. It is hard to find shoes that do not hurt my feet.	1	2	3	4	5
11. I have difficulty in finding shoes that fit my feet.	1	2	3	4	5
12. I am limited in the number of shoes I can wear.	1	2	3	4	5

13. In general, what condition would you say your feet are in ?

(circle number)

Excellent..... 1
 Very Good..... 2
 Good..... 3
 Fair..... 4
 Poor..... 5

Please write some comments about the current state of your feet:

.....

.....

.....

.....

.....

Foot health status questionnaire Version 1.04: Analysis is available from www.fhsq.org and supported by Microsoft Windows™

Podiatry CPG: Appendix 2

14. In general, how would you rate your health :

(circle number)

Very Good..... 1
Fair..... 2
Poor..... 3

15. The following questions ask about activities you might do during a typical day. Does your health now limit you in these activities? If so, how much?

(circle a number on each line)

<u>ACTIVITIES</u>	Yes, Limited A Lot	Yes, Limited A Little	No, Not Limited At All
a. Vigorous activities , such as running, lifting heavy objects, or (if you wanted to) your ability to participate in strenuous sports	1	2	3
b. Moderate activities , such as cleaning the house, lifting a chair, playing golf or swimming	1	2	3
c. Lifting or carrying bags of shopping	1	2	3
d. Climbing a steep hill	1	2	3
e. Climbing one flight of stairs	1	2	3
f. Getting up from a sitting position	1	2	3
g. Walking more than a kilometre	1	2	3
h. Walking one hundred meters	1	2	3
i. Showering or dressing yourself	1	2	3

16. This next question asks to what extent has your physical health or emotional problems interfered with your normal social activities with family, friends, neighbours or social groups?

(circle number)

Not at all..... 1
Slightly..... 2
Moderately..... 3
Quite a bit..... 4
Extremely..... 5

Please turn to the next page

Podiatry CPG: Appendix 2

17. These questions are about how you “feel” and how things have been with you during the past month. For each question, please give the one answer that comes closest to the way you have been “feeling”. How much of the time during the past 4 weeks:

	All of the time	Most of the Time	Some of the Time	A little of the Time	None of the Time
a. Did you feel tired?	1	2	3	4	5
b. Did you have a lot of energy?	1	2	3	4	5
c. Did you feel worn out?	1	2	3	4	5
d. Did you feel full of life?	1	2	3	4	5

18. During the past 4 weeks, how much of the time has your emotional problems or physical health interfered with your social activities (like visiting with friends, relatives, etc.)?

(circle number)

- No time at all..... 1
A small amount of time..... 2
Moderate amount of time..... 3
Quite a bit of the time..... 4
All of the time..... 5

19. How TRUE or FALSE is each of the following statements for you?

(circle a number on each line)

	True or Mostly True	Don't Know	False or Mostly False
a. I seem to get sick a little easier than other people	1	2	3
b. I am as healthy as anybody I know	1	2	3
c. I expect my health to get worse	1	2	3
d. My health is excellent	1	2	3

Foot health status questionnaire Version 1.04: Analysis is available from www.fhsq.org and supported by Microsoft Windows™

Please complete the following details.

20. Full Name: _____

21. Address: _____ Postcode: _____

22. Date of Birth: _____ Sex: Male ☐ Female ☐

23. What is the date when you filled out this survey? Please write here → _____

24. Do you currently take any medicine prescribed by your doctor for any of the following conditions ;

(please tick the appropriate box/s)

Diabetes	<input type="checkbox"/>	Hormone Replacement Therapy	<input type="checkbox"/>
Osteoarthritis	<input type="checkbox"/>	High Cholesterol	<input type="checkbox"/>
Blood Pressure	<input type="checkbox"/>	Rheumatoid Arthritis	<input type="checkbox"/>
Heart Disease	<input type="checkbox"/>	Back Pain	<input type="checkbox"/>
Lung Disease	<input type="checkbox"/>	Depression	<input type="checkbox"/>

Any other conditions you take
medicine for, please list

1.
2.
3.

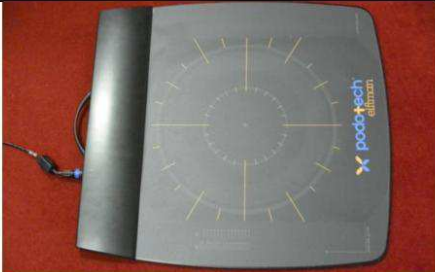


For the next questions, please tick either YES or NO

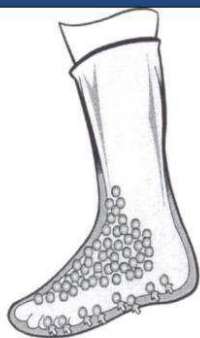
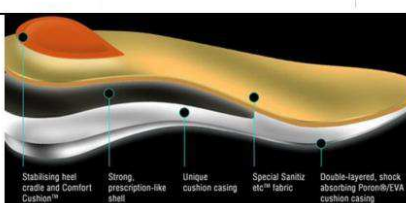


	Yes	No
25. Are you a pensioner or health care cardholder ?	<input type="checkbox"/>	<input type="checkbox"/>
26. Do you smoke cigarettes ?	<input type="checkbox"/>	<input type="checkbox"/>
27. Do you do any regular physical exercise ?	<input type="checkbox"/>	<input type="checkbox"/>
28. Do you have private health insurance ?	<input type="checkbox"/>	<input type="checkbox"/>
29. Have you completed a trade certificate or any other educational qualification since leaving school ?	<input type="checkbox"/>	<input type="checkbox"/>

**Thank you for completing this
questionnaire**

Podiatry CPG: Appendix 2


C) Foot and walking assessment tools

i)	Elfman gait analysis platform																																									
ii)	Fscan inshoe analysis system																																									
iii)	Gaitrite System	<div></div> <div><p>GAITRite</p><p>Our standard Portable Electronic Walkway.</p><table><tr><th>GAITRite Walkway Model</th><th>Active Area Width (in/cm)</th><th>Active Area Length (in/cm)</th><th>Overall Outer Width (in/cm)</th><th>Overall Outer Length (in/cm)</th></tr><tr><td>PP12</td><td>24.0/61.0</td><td>180.0/457.2</td><td>35.0/89.0</td><td>180.0/457.2</td></tr><tr><td>PP14</td><td>24.0/61.0</td><td>204.0/518.1</td><td>35.0/89.0</td><td>204.0/518.1</td></tr><tr><td>PP16</td><td>24.0/61.0</td><td>228.0/579.1</td><td>35.0/89.0</td><td>228.0/579.1</td></tr><tr><td>PP18</td><td>24.0/61.0</td><td>252.0/640.0</td><td>35.0/89.0</td><td>252.0/640.0</td></tr><tr><td>PP20</td><td>24.0/61.0</td><td>276.0/701.0</td><td>35.0/89.0</td><td>276.0/701.0</td></tr><tr><td>PP24</td><td>24.0/61.0</td><td>324.0/822.9</td><td>35.0/89.0</td><td>324.0/822.9</td></tr><tr><td>PP26</td><td>24.0/61.0</td><td>348.0/883.9</td><td>35.0/89.0</td><td>348.0/883.9</td></tr></table></div>	GAITRite Walkway Model	Active Area Width (in/cm)	Active Area Length (in/cm)	Overall Outer Width (in/cm)	Overall Outer Length (in/cm)	PP12	24.0/61.0	180.0/457.2	35.0/89.0	180.0/457.2	PP14	24.0/61.0	204.0/518.1	35.0/89.0	204.0/518.1	PP16	24.0/61.0	228.0/579.1	35.0/89.0	228.0/579.1	PP18	24.0/61.0	252.0/640.0	35.0/89.0	252.0/640.0	PP20	24.0/61.0	276.0/701.0	35.0/89.0	276.0/701.0	PP24	24.0/61.0	324.0/822.9	35.0/89.0	324.0/822.9	PP26	24.0/61.0	348.0/883.9	35.0/89.0	348.0/883.9
GAITRite Walkway Model	Active Area Width (in/cm)	Active Area Length (in/cm)	Overall Outer Width (in/cm)	Overall Outer Length (in/cm)																																						
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PP26	24.0/61.0	348.0/883.9	35.0/89.0	348.0/883.9																																						

Socks and Insoles Products	
Adults	
<p>Figure 1: Sock contains pure silver a naturally occurring element Silver acts as a transmitter removing heat and regulating temperature</p>	
<p>Figure 2: Functional orthotics</p>	
<p>Figure 3: Cushioning insoles – Coolsorb. Keep in the fridge.</p>	
<p>Figure 4: Microporous cellular structure allows feet to breathe Absorbs shock Rebounds to original thickness step after step</p>	

Shoe Products	
Children	
 <p>Figure 1: Startrites shoes https://www.startriteshoes.com/</p>	T/A
 <p>Figure 2: Clarke shoes https://www.clarks.co.uk</p>	All
 <p>Figure 3: Kid-o-fit shoes https://www.happylittlesoles.co.uk/products/brand-1/kidofit-1.html</p>	T/ A
Key	
All	All subtypes of EB
T	Toddlers
A	Adolescent
Ad	Adult

	
<p>Figure 4: Geox shoes https://www.geox.com/en-GB/</p>	All
	
<p>Figure 5: Crocs https://www.crocs.co.uk</p>	All <u>T/A/Ad</u>
	
<p>Figure 6: UGG boots https://www.ugg.com</p>	All <u>T/A/Ad</u>
<p>Key</p> <p>All All subtypes of EB</p> <p>T Toddlers</p> <p>A Adolescent</p> <p>Ad Adult</p>	

B. Shoe Products	
Adults	
Rockport: http://www.rockportuk.com	Ad
Timberland: https://www.timberland.co.uk	Ad/A
Crocs: https://www.crocs.co.uk	Ad/A
UGG boots: https://www.ugg.com	All/ Ad
New balance: https://www.newbalance.co.uk/	All
https://www.newbalance.co.uk/kids/shoes/children-size-10-2.5/	T/A
Asics: https://www.asics.com/gb/en-gb/	All
https://www.asics.com/gb/en-gb/kids-grade-school-gear/c/as30603000/	T/A
Brookes: https://www.brooksrunning.com/en_gb	Ad
	
Figure 1: Fitflop https://www.fitflop.com/uk/en/cart	A/Ad
Key	
All	All subtypes of EB
T	Toddlers
A	Adolescent
Ad	Adult

C. Trainers Products	
Adults	
	
Figure1: Running trainers Sketchy Sketchers: https://www.skechers.com/en-gb/	All

Nike.com	All
	
Figure 2: Running trainers Nike Nike free trainers: https://store.nike.com/gb/en_gb/product/free-rn-flyknit-2018-id-shoe/?piid=44870&pbid=190565551	
Adidas: https://www.adidas.co.uk/	All
https://www.adidas.co.uk/pod-s3.1-shoes/B42073.html?pr=nosearch_rr&slot=9	A
Key	
All	All subtypes of EB
T	Toddlers
A	Adolescent
Ad	Adult

D. Bespoke foot-wear

Footwear semi-bespoke/measured for/modified if needed:

<https://www.reedmedical.co.uk/>

<http://www.saltstechstep.co.uk/footwear/diabetic>

Bespoke shoes:

<http://www.saltstechstep.co.uk/footwear/bespoke>

I can send some photos of what we've provided in the past

bespoke safety boots

<http://www.bolton-bros.co.uk/index.php/bespoke-footwear/bespoke-safety-footwear>

Bespoke Footwear

The podiatry team provide bespoke footwear that are tailored to meet each patients individual needs.

We work closely with local orthotics and some of the elite footwear manufacturers across the country to provide personalised and stylish footwear.

Safety shoes/footwear are made to measure and adhere to the national safety requirements.



Seamless footwear: can cut down on blistering and friction.



E.B. Podiatrists

The Podiatrists are part of the multidisciplinary team.

We specialise in providing an individual podiatry assessment for each patient and then implement appropriate specialist treatment in order to alleviate symptoms and promote optimum mobility.


We also specialise in biomechanics and can provide bespoke insoles and/or footwear to help improve long term foot health and comfort for our patients.

Contact us

0121 424 5232
ebteamsolihull@heartofengland.nhs.uk

Heart of England **NHS**
NHS Foundation Trust

The E.B. Podiatry Team

 Building healthier lives

E.B. Podiatry

Blistering is a regular occurrence and usually can't be prevented. However, with advice and treatment from our team mobility can be greatly improved.

Regular podiatry care

There maybe increased prevalence of hyperkeratosis (thick hardened skin) and blistering on the soles of the feet. We can provide ongoing Podiatry care or referrals to local podiatrists with specific technical advice.



Certain products such as padding, strapping and E.B. specific hosiery are also available through our service. We are continually trying to source and sample cutting edge materials in order to optimise the quality of the service we provide.

Further advice

The aim of the Podiatry team is to try and alleviate symptoms, by providing footwear that the patient is both comfortable and confident with and in turn providing relief from pain.

The footwear we offer our patients is extremely varied and is ultimately based on individual needs and personal preference.



Once footwear has been reviewed and discussed we will then assess the possible additional benefit of in-shoe orthotics.



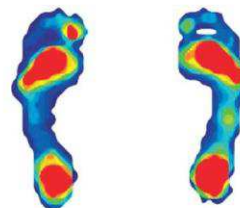
Such devices are varied and will be individually tailored and prescribed by us depending on symptoms, footwear and activity.

Bespoke Orthotics

As part of our desire to provide bespoke interventions for each patient, we also offer an in shoe plantar pressure measurement (tekscan) service.



This entails using electronic insoles inserted into the patients current footwear which provides readouts of varying pressures under the soles of the feet as shown below.



With this information we are then able to produce custom made off-loading insoles to help prevent or reduce blister build up.



Podiatry care in epidermolysis bullosa

DEBRA International is undertaking a long-term initiative to develop clinical practice guidelines (CPGs) for EB. These guidelines will help improve the clinical and social care of EB patients across the globe.

This guideline is specifically focused on providing insight and recommendations into the topic of Podiatry in Epidermolysis bullosa.

Your responses to the following survey will directly affect the questions and issues addressed in the International Clinical Practice Guidelines for Podiatry in people living with Epidermolysis Bullosa.

This survey is designed for people living with EB, families, carers supporting people living with EB.

Confidentiality

Participation in this survey is voluntary. No compensation will be provided for completion. Any identifying information you provide will be used solely for interpretation of survey results to direct the clinical practice guideline process. By completing this survey, you consent to the use of any information you provide in this process.

1. Do you require feet care/podiatry care due to EB?

- ☐ yes
☐ no

2. What are your problem areas? (Select all that applies)

- ☐ Dystrophic nails
☐ Blistering and Wound management
☐ Mobility
☐ Shoes
☐ Hyperkeratosis (thickening of the outer layer of the skin.)
☐ Fusion of toes
☐ Dry and hardened areas
☐ Other (please specify)

3. How often problems with your feet affect your activities?

	Never	Rarely	Sometimes	Often	Always	N/A
walking	<input type="radio"/>	<input type="radio"/>	<input type="radio"/>	<input type="radio"/>	<input type="radio"/>	<input type="radio"/>
doing sports	<input type="radio"/>	<input type="radio"/>	<input type="radio"/>	<input type="radio"/>	<input type="radio"/>	<input type="radio"/>
working	<input type="radio"/>	<input type="radio"/>	<input type="radio"/>	<input type="radio"/>	<input type="radio"/>	<input type="radio"/>
independence	<input type="radio"/>	<input type="radio"/>	<input type="radio"/>	<input type="radio"/>	<input type="radio"/>	<input type="radio"/>
socialising	<input type="radio"/>	<input type="radio"/>	<input type="radio"/>	<input type="radio"/>	<input type="radio"/>	<input type="radio"/>
relying on others for help	<input type="radio"/>	<input type="radio"/>	<input type="radio"/>	<input type="radio"/>	<input type="radio"/>	<input type="radio"/>

4. Which problem areas do you think the guideline should concentrate on the most?

☐

Dystrophic nails

☐

Blistering and wound management

☐

Mobility

☐

Exploring the most suitable shoes for EB

☐

Hyperkeratosis (thickening of the outer layer of the skin)

☐

Fusion of toes

5. Do you have any other concerns relating to feet care that could be relevant to the guideline?

6. What do you use on your feet to prevent blistering? (Select all that applies)

- ☐ dressings
- ☐ vaseline gauze wrap
- ☐ seamless socks
- ☐ custom made shoe
- ☐ creams
- ☐ insoles or orthotics
- ☐ don't use anything

Other (please specify)

7. How do you think EB patients could be best supported relating to feet care?

- ☐ supported in the community by local health professionals
- ☐ supported in the hospital during regular hospital check-ups
- ☐ supported in patients own home by community staff and referred to hospitals if necessary
- ☐ Other (please specify)

8. What type of EB do you have? (for easier analysis only using the main types not the subtypes)

- ☐ EB Simplex
- ☐ Junctional EB
- ☐ Dystrophic EB
- ☐ Kindler Syndrome

9. What age group do you belong to?

- ☐ 0 - 5 years
- ☐ 5 - 10 years
- ☐ 10 - 18 years
- ☐ 18 - 25 years
- ☐ 25 - 35 years
- ☐ 35 - 45 years
- ☐ 45 - 55 years
- ☐ above 55 years

10. Where do you live?

- ☐ Europe
- ☐ North America
- ☐ South America
- ☐ Asia
- ☐ Australia Oceania
- ☐ Africa
- ☐ Other (please specify)



Podiatry CPG: Appendix 6

Thank you!

Thank you for taking the time to complete this survey; your feedback will be used by the Podiatry CPG panel to develop the guideline.

DECLARATION

I understand that the information I provide will be used only for the discussion, direction, and development of the Podiatry Clinical Practice Guidelines.

I understand that all data collected through this survey will be anonymised and will not become identifiable in the guideline development process. For more details on how DEBRA International uses your information, please visit www.debra-international.org.

Podiatry CPG: Appendix 7

PART ONE: ELIGIBILITY		Answer
Participants	Do the participants in the study have EB, are family members of someone with EB, are carers (professional/personal) of person(s) with EB? (what proportion sufficient for inclusion?)	Yes (1) /No (0)
Outcomes	Is the outcome(s) one or some of the following: Dystrophic nails; Blistering and wound management; Mobility; Exploring the most suitable shoes for EB; Hyperkeratosis (thickening of the outer layer of the skin); Fusion of toes?	Yes (1) /No (0)
Design	Is the methodology one of the following: Quantitative, Qualitative, Systemic reviews, Meta-analysis, RCTs, Cohorts, Case control, Diagnostic studies, Observational? If the paper is a dissertation, position (opinion) paper	Yes (1) /No (0)
ACTION		If you have answered NO to any of these questions please STOP HERE . If you have answered YES for all questions, please proceed to Part Two .
PART TWO: QUALITY ASSESSMENT		Answer
Overview	1. Does the title reflect the content? 2. Are the authors contact details and institute reported? 3. Does the abstract summarise the key components? 4. Is the rationale for undertaking the research clearly outlined? 5. Has there been a comprehensive literature review with a clearly outlined process? 6. Is the aim of the research clearly stated? 7. Has it been approved by an ethical board?	Yes (1) /No (0) Yes (1) /No (0) Yes (1) /No (0) Yes (1) /No (0) Yes (1) /No (0) Yes (1) /No (0) Yes (1) /No (0)
with EB, people who work with people with EB?	Was everyone included who should be included? Or was there something particular (biased) about the sample? E.g., only people with RDEB? Or, only people with hand problems? Only people who have been referred to a psychologist? What are the sources of sampling bias.....	Yes (1) /No (0)
Method Design Section A	Is this Paper Quantitative research? (including mixed method)	If you have answered NO go to section B. Yes complete the quantitative section A.
	8. Those this study have a control group? 9. Is the study design clearly identified, and is the rationale for choice of design evident? 10. Is there an experimental hypothesis clearly stated? 11. Are the key variables clearly defined? 12. Were the outcome measures valid? Did they actually measure what the study set out to measure?	Yes (1) /No (0) Yes (1) /No (0) Yes (1) /No (0) Yes (1) /No (0) Yes (1) /No (0)
Method Design Section B	Is this a paper on Qualitative research? (including mixed method)	If you have answered NO go to FINAL section. Yes complete the qualitative section B.
	8. Is the study design clearly identified, and 9. Is the selection of participants described and the sampling method identified? 10. Is the method of data analysis credible and confirmable? 11. Are the results presented in a way that is appropriate and clear?	Yes (1) /No (0) Yes (1) /No (0) Yes (1) /No (0) Yes (1) /No (0)
Final section	12. Is there a good critical discussion linked to relevant evidence? 13. Are the results generalizable? 14. Are the results transferable? 15. Is the conclusion comprehensive? 16. Have they reported any limitations to the study? 17. Have there any conflicts of interest with the authors or the funding?	Yes (1) /No (0) Yes (1) /No (0) Yes (1) /No (0) Yes (1) /No (0) Yes (1) /No (0) Yes (1) /No (0)
Final Quality Framework questions (these need discussion in your outcome groups)		
Per outcome summary table	Sample populations: What type of EB Number of subjects with EB Study Design/ Method 18. Was the chosen study design appropriate to answer the question posed in the study? 19. Is the method of data analysis valid and reliable? 20. Did the measurements have proven satisfactory reliability and validity?	Yes (1) /No (0) Yes (1) /No (0) Yes (1) /No (0)
Framework totals	Maximum score for study types Qualitative studies Quantitative studies Mixed methods studies	20 21 25
How much population sample selection bias is present		
	1. Does the study use random selection to put participants into groups? 2. Is the person undertaking the research or data analysis unknown to the subject e.g. involved in their care? 3. Are all patients in study reported on 4. Is there Selective out-come reporting? e.g. not all outcomes or patients are reported on	Yes (1) /No (0) Yes (1) /No (0) Yes (1) /No (0) Yes (1) /No (0)
Rating the Strength of Psychosocial Outcome article summaries & Recommendations		
	Does this OUTCOME address a subject that is a priority for our patient population? (vote) How strong is the evidence that what the article is saying is true = the overall certainty of the Look at SIGN and quality score to do this and consider any grey literature. Are the desired effects relevant? Discuss Are there any undesirable effects? Discuss Are the costs small relative to the benefits in carrying out the recommendations? Discuss Are the recommendations feasible to implement?	1. very weak, to 4. very strong 1. very weak, to 4. very strong 1. very weak, to 4. very strong 1. very weak, to 4. very strong 1. very weak, to 4. very strong 1. very weak, to 4. very strong 1. very weak, to 4. very strong



EB Podiatry Guideline: Appendix 8

CPG Evaluation Form: Pre implementation

Please answer the questions below to help us evaluate the usefulness and effectiveness of our guidelines, as well as learn more about those using the information contained in the publication.

Section A: General Information			
How did you hear about this guideline? <i>Please tick all that apply and provide details where applicable</i>	<input type="checkbox"/> Journal searches (e.g. PubMed) <input type="checkbox"/> Event (e.g. DEBRA conference, EB study day) <input type="checkbox"/> Recommendation (e.g. family member, clinician) <input type="checkbox"/> DEBRA communication (e.g. newsletter, website, social media) <input type="checkbox"/> Other (please specify)		
Why have you chosen to review this guideline?			
City and country: <i>Residence</i>			
DEBRA group affiliation: <i>If applicable</i>			
Which of the following best describes you? <i>Please tick all that apply</i>	Group A <i>Professionals</i> <input type="checkbox"/> Clinician <input type="checkbox"/> Allied health professional <input type="checkbox"/> Professional carer	Group B <i>Non-professionals</i> <input type="checkbox"/> Person living with EB <input type="checkbox"/> *Caregiver (family member) <input type="checkbox"/> Caregiver (friend) <input type="checkbox"/> Friend	Group C <i>Other</i> <input type="checkbox"/> Other (please provide details)
	<i>Please continue with Section B</i>	<i>Please continue with Section C</i>	<i>Please continue with Section B or C (as appropriate)</i>

Section B: Professionals	
Your institute: <i>Name and address (including country)</i>	
Tell us about the type of EB services you offer: <i>Please tick all that apply and provide details</i>	<input type="checkbox"/> Hospital <input type="checkbox"/> Community <input type="checkbox"/> Home service <input type="checkbox"/> Other (please specify)

* For the purposes of this document, a caregiver indicates a non-professional. If you are a carer in a professional capacity, please tick the 'Professional carer' option in Group A.



EB Podiatry Guideline: Appendix 8

<p>Please tell us who works with EB patients at your institute:</p> <p><i>Please tick all that apply and circle the number of relevant people</i></p>	<input type="checkbox"/> Consultant <input type="checkbox"/> Dermatologist <input type="checkbox"/> Surgeon <input type="checkbox"/> Nurse (specialist) <input type="checkbox"/> Psychologist, psychotherapist <input type="checkbox"/> Dietician, nutritionist <input type="checkbox"/> Dentist <input type="checkbox"/> Physiotherapist <input type="checkbox"/> Podiatrist <input type="checkbox"/> Occupational therapist, hand therapist <input type="checkbox"/> Social worker/social support <input type="checkbox"/> Other (please specify)	1 1 1 1 1 1 1 1 1 1 1 1	2 2 2 2 2 2 2 2 2 2 2 2	3 3 3 3 3 3 3 3 3 3 3 3	4+ 4+ 4+ 4+ 4+ 4+ 4+ 4+ 4+ 4+ 4+ 4+																																																
<p>Please give a summary of the EB service(s) your institute provides:</p> <p><i>300 words maximum; please attach and use additional pages, if needed</i></p>																																																					
<p>Target groups:</p> <p><i>Please tick all that apply and provide approximate numbers, where applicable</i></p>	<table border="1"> <thead> <tr> <th>Living with EB (years of age)</th> <th>EBS</th> <th>JEB</th> <th>DDEB</th> <th>RDEB</th> <th>KS</th> </tr> </thead> <tbody> <tr> <td><input type="checkbox"/> Newborn and neonatal</td> <td></td> <td></td> <td></td> <td></td> <td></td> </tr> <tr> <td><input type="checkbox"/> Children (0-7)</td> <td></td> <td></td> <td></td> <td></td> <td></td> </tr> <tr> <td><input type="checkbox"/> Children (8-12)</td> <td></td> <td></td> <td></td> <td></td> <td></td> </tr> <tr> <td><input type="checkbox"/> Teenagers (12-18)</td> <td></td> <td></td> <td></td> <td></td> <td></td> </tr> <tr> <td><input type="checkbox"/> Young adults (18-25)</td> <td></td> <td></td> <td></td> <td></td> <td></td> </tr> <tr> <td><input type="checkbox"/> Adults (25+)</td> <td></td> <td></td> <td></td> <td></td> <td></td> </tr> <tr> <td><input type="checkbox"/> Other (please specify)</td> <td></td> <td></td> <td></td> <td></td> <td></td> </tr> </tbody> </table>					Living with EB (years of age)	EBS	JEB	DDEB	RDEB	KS	<input type="checkbox"/> Newborn and neonatal						<input type="checkbox"/> Children (0-7)						<input type="checkbox"/> Children (8-12)						<input type="checkbox"/> Teenagers (12-18)						<input type="checkbox"/> Young adults (18-25)						<input type="checkbox"/> Adults (25+)						<input type="checkbox"/> Other (please specify)					
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EB Podiatry Guideline: Appendix 8

	<p>How confident are you/your team in providing care for EB patients relating to this issue?</p> <p>1 2 3 4 5</p> <p>Not at all confident Extremely confident</p> <p>How often do you/your team manage this clinical issue?</p> <p>1 2 3 4 5</p> <p>Less than yearly Yearly Monthly Weekly Daily</p>
<p>Describe how you think this guideline will benefit your service users:</p> <p><i>Including patients, clients, families, etc.</i></p>	

Section C: Non-professionals	
<p>Which type of EB do you (your family member/friend) have?</p>	<p><input type="checkbox"/> EBS <input type="checkbox"/> JEB <input type="checkbox"/> DDEB <input type="checkbox"/> RDEB <input type="checkbox"/> KS</p> <p><input type="checkbox"/> I (my family member/friend) have not officially been diagnosed</p> <p><input type="checkbox"/> Other (please specify)</p>
<p>Before reading the guideline, what information do you already know about this clinical issue?</p> <p><i>Please circle the number you feel best reflects your experience and/or tick the most appropriate box, where applicable.</i></p>	<p>How confident are you to get appropriate care in this clinical area?</p> <p>1 2 3 4 5</p> <p>Not at all confident Extremely confident</p> <p>Why did you indicate this level of confidence?</p> <p><input type="checkbox"/> No one is available to help</p> <p><input type="checkbox"/> I only have written information about the clinical area</p> <p><input type="checkbox"/> I have a person I can reach by phone, email, or other means of communication</p> <p><input type="checkbox"/> I only have a local, general doctor</p> <p><input type="checkbox"/> My local EB association gives me support (please specify)</p> <p><input type="checkbox"/> My doctor has experience</p> <p><input type="checkbox"/> My centre has EB experts</p> <p><input type="checkbox"/> I do not require care</p> <p><input type="checkbox"/> Other (please specify)</p>



EB Podiatry Guideline: Appendix 8

	How often do you (your family member/friend) experience this problem?				
	1	2	3	4	5
	Less than yearly	Yearly	Monthly	Weekly	Daily
How do you (your family member/friend) normally manage this problem?					

DEBRA International (DI) would like to further contact respondents to this guideline survey a year later for post evaluation measurement. The purpose of this additional survey is to evaluate the use of the guideline after its principles have been applied in practice.

Declaration

I understand that the information I provide will be used for data collection and guideline evaluation purposes only.

I understand that all data collected through these surveys will be anonymised and will not become identifiable in the evaluation process.

I understand that if DI would like to contact me in the future, they will only be able to do so for the purposes of being asked to complete the post evaluation survey, unless consent has otherwise been obtained for additional purposes.

I understand I am under no obligation to complete the post evaluation survey once I receive it, and that I am able to change or withdraw my consent at any time by notifying DI (office@debra-international.org). I understand if consent is withdrawn, DI will destroy all identifiable records (i.e. contact details provided with the pre or post evaluation surveys).

I understand I have the right to ask to see any information and records held about me by DI.

☐ I give my consent for DI to contact me after one year with a post evaluation guideline survey.

For more details on how DI uses your information, please visit www.debra-international.org.

Contact Details			
Full name			
Signature		Date	
Email			