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Guidelines

Neonatal epidermolysis bullosa: a clinical practice guideline

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Abstract

DEBRA International is undertaking a long-term initiative to develop clinical practice guidelines (CPGs) for epidermolysis bullosa (EB), to improve the clinical care of people living with EB. Current neonatal care is based on evidence, clinical expertise and trial and error, with collaboration between the EB specialist team, parent or carer and patient, and is dependent on the neonate's individual presentation and type of EB.

Early intervention based on research and clinical practice is needed to establish a foundation of knowledge to guide international practitioners to create and improve standards of care and to be able to work effectively with those newly diagnosed with EB. This CPG was created by an international panel with expertise working with persons with EB. The CPG focuses on neonatal care using a systematic review methodology covering four key areas: (i) diagnosis and parental psychosocial support; (ii) hospital management: medical monitoring, wound care and pain; (iii) feeding and nutrition; and (iv) discharge planning and EB education.

These four areas highlight the importance of a multidisciplinary team approach, to provide a patient-specific holistic care model that incorporates the needs and wishes of the parents and carers. The Hospital Implementation Tool included promotes transfer of theory to clinical practice.

What is already known about this topic?

- All subtypes of epidermolysis bullosa (EB) can cause complications for the neonate at birth.
- Implementation of preventative management during handling and medical monitoring will minimize risk of skin damage.
- Many major teaching hospitals around the world that look after neonates with EB have similar guidelines and protocols for management.
- Inpatient management for people living with EB requires specialist EB support.

What does this study add?

- Neonatal EB-specific medical, nutritional and educational inclusive guidelines.
- Improved effectiveness, accessibility and quality of care for neonates with EB.
- Care recommendations that reinforce health provider education, access to specialist multidisciplinary team care, and family inclusion throughout care.
- Inclusion of EB education and continuity of care from hospital discharge to community care.

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The recommendations in these guidelines do not constitute a single approach or standard of medical care. Variations to the recommendations provided may be indicated on an individual, organizational or other basis. Considerable efforts have been made by this panel to ensure the content is accurate and up to date.

It is recognized that under certain conditions it may be necessary to deviate from these guidelines and that the results of future studies may require some of the recommendations herein to be changed. Failure to adhere to these guidelines should not necessarily be considered negligent, nor should adherence to these recommendations constitute a defence against a claim of negligence.

1 Background

Epidermolysis bullosa (EB) is an umbrella term for a group of genetic disorders characterized by blistering and wounds resulting from friction to the skin, due to the absence or reduction of one of the proteins that help bind the layers of the skin together.[1–4] The four major subtypes of EB include EB simplex (EBS), junctional EB (JEB), dystrophic EB (DEB) and Kindler EB (KEB).[4,5] All subtypes have the potential for significant birth trauma that requires intensive monitoring, wound care and feeding support. Some forms of EB can be fatal in infancy due to severe mucosal lining damage, oesophageal atresia or extensive wounds.[4–7]

A neonate, aged up to 28 days past the expected due date,[8] with suspected EB requires treatment from an experienced EB multidisciplinary team (MDT) to implement preventative and clinical strategies to reduce blistering, prevent deterioration of wounds and provide adequate nutrition.[1,2,6,9,10] Signs of EB at birth vary from scant blistering through to extensive wounds including degloving wounds (total or partial absence of skin covering a section of the limb). Development of further wounds or blisters can occur from handling or monitoring procedures. High wound exudate, risk of infection, and pain on feeding and/or movement all impact the clinical outcomes of the neonate.

Parents of a neonate with EB can experience practical and emotional difficulties due to the rarity and complexity of the disease.[11,12] Difficulties include poor access to experienced healthcare professionals, delays in appropriate care and diagnosis and increased financial stress navigating the healthcare system. Access to early psychosocial support, education and discharge planning in hospital, and upskilling local health professionals prior to discharge all contribute to a successful transition from hospital to home.[1,6,9,13,14]

1.1 Purpose and scope

This clinical practice guideline (CPG) highlights management priorities in neonatal EB care and provides clinicians access to recommended treatment and preventative care options. The recommendations were reached following systematic literature review, summary of hospital policies and expert consensus on the management of neonates with EB (details of panel and review members in Appendix S1 and Table S1a, S1b and S1c, see Supporting Information). This guideline is intended to inform and support clinicians of neonates with EB.

For families that include a mother and neonate with EB these recommendations may be used in conjunction with the pregnancy, childbirth and aftercare CPG[2] to support the inclusion of both maternal and neonatal care.

1.2 User and target groups

This CPG has been developed to inform and aid decision making for health professionals and parents and carers of neonates with EB (all subtypes). The target group includes paediatricians, neonatologists, midwives, neonatal and paediatric nurses, dermatologists, anaesthetists, family practice physicians, psychologists, occupational therapists, physiotherapists, dietitians, genetic counsellors, social workers, parents and carers.

1.3 Aims

- To provide a comprehensive, expert, evidence-informed resource for neonatal EB care.
- To decrease the incidence of avoidable neonatal EB complications.
- To promote inclusion of education, preventative care and MDT management for neonatal EB care.
- To highlight resources required for EB care after leaving hospital.

1.4 Methodology

The methodology stages used for the development of this CPG, and for implementation and future research are outlined in Appendix S2 and Tables S2a and S2b (see Supporting Information), with reference to the literature review, GRADE, Critical Appraisal Skills Programme (CASP) and the AGREE II instrument.[15–17]

The CPG development process established four broad clinical questions pertinent to the guideline scope.

A. Should early and EB-appropriate emotional support, compared with standard neonatal care, be used in and throughout the period of EB diagnosis?
B. Should EB standards of wound care, pain and sepsis management, compared with standard care, be used for neonates with EB to prevent complications?
C. Should early nutrition dietary interventions, compared with standard neonatal care, be used for neonates with EB?
D. Should the care of neonates with EB include early education, handling and discharge planning, compared with standard neonatal management?

1.5 Results

A literature search identified 571 papers relevant to the neonatal priority (Figure 1). After screening and appraisal, and two updated literature searches, a total of 43 papers were included in the full appraisal.

The panel used additional evidence including CPGs, hospital protocols and academic grey literature to support the recommendations. See Appendix S2 and Table S2b for appraised article allocation per outcome.

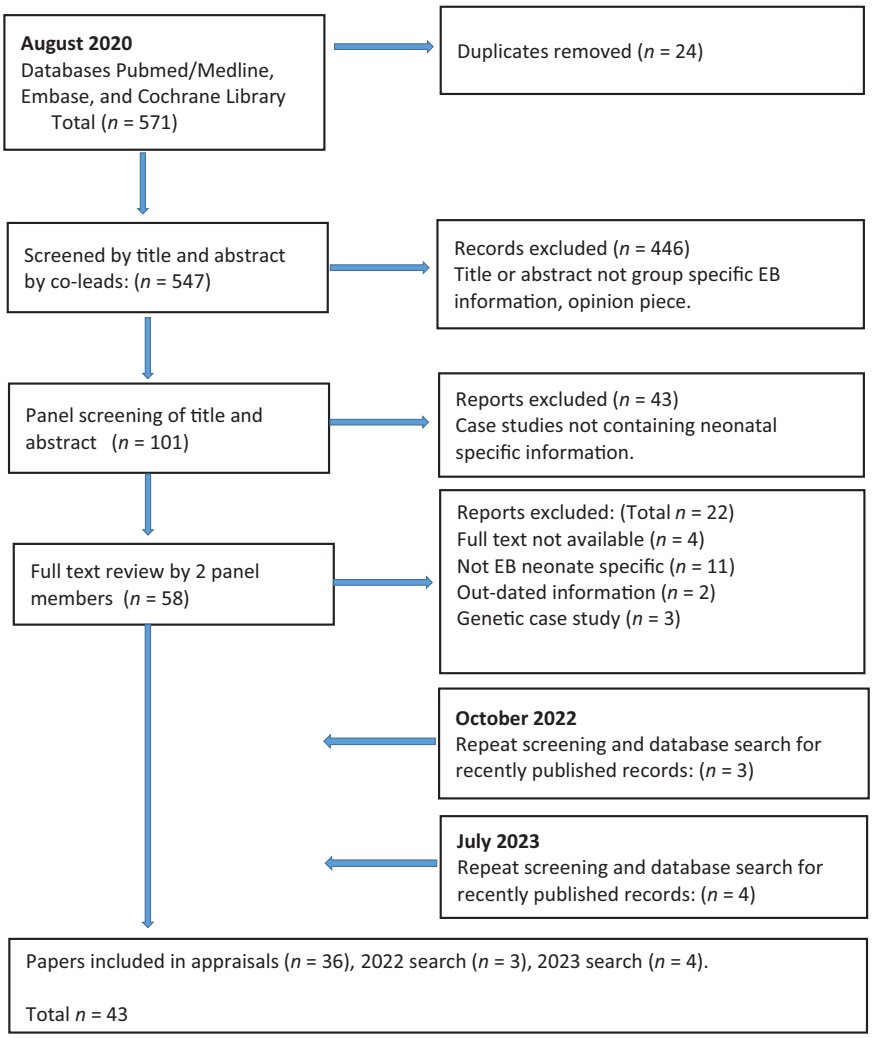

Figure 1 Literature search for epidermolysis bullosa (EB).

2 Recommendations

This CPG will discuss neonatal care in sequential order from birth, through hospital care, to discharge and referral to community care.

Recommendation tables

A summary of recommendations can be found Tables A, B.1–5, C.1–3, D and E.

A detailed key to the recommendations can be found in Appendix S3 (see Supporting Information). Recommendations based on good practice points (GPP) are marked as such. Recommendations for future research are listed in Appendix S4 (see Supporting Information).

Hospital Implementation Tool

Practical resource: use the recommendation table summaries and the 'Hospital Implementation Tool' (Appendix S5; see Supporting Information) to guide use of the CPG in clinical practice.

A: Neonatal diagnosis and parental psychosocial support

Early clinical evaluation and genetic testing can support clinical management and the parent's or carer's understanding of the prognosis. Nursing and wound care can be tailored effectively when diagnosis[5] and EB classification[4] are established. Misdiagnosis or lack of subtype-specific diagnosis can lead to unnecessary monitoring and handling.[18]

Parents and carers of a neonate with EB experience many challenges during hospitalization, complicated by the rarity of the disease. There is a high emotional burden for parents and carers with reduced access to their emotional and social support networks, complicating their ability to cope with and learn about EB.[12,19,20] Extended hospital stays and time away from home support can increase financial burden,[12,19,21] further exacerbating emotional stress. Implementation of a rare disease model of care that includes parents' and carers' wishes, referral to specialist care, access to education, and community support models[21,22] can support parents or carers and the neonate with EB.

R1 ↑↑ Consider providing access to early and appropriate emotional support throughout the period of diagnosis.

Table A Summary of recommendations for neonatal diagnosis and parent psychosocial support in epidermolysis bullosa (EB)

Recommendation	Strength of recommendation	Level of evidence	Key references
R1 ↑↑ Consider providing access to early and appropriate emotional support throughout the period of diagnosis			
Offer family EB education and psychosocial support while waiting for genetic results	↑↑	Very low	1, 3, 9, 10, 12, 18, 23, 25 →13
Access an EB specialist to provide diagnostic and management education for families	↑↑	Very low	1, 6, 14, 18, 20, 25 +26, +27
Promote psychological support for family if transfer away from community support structure is required for medical reasons. Link to local DEBRA organization	↑↑	Very low	11, 12, 14, 19, 20, 22, 28–30 →13
R2 ↑↑ Provide education and support for parents and carers throughout diagnosis and prognosis			
Offer referral to palliative care and psychosocial teams to support moral dilemmas about diagnosis and prognosis	↑↑	Very low	1, 3, 12, 18, 25, 35 →5, →13
Refer to genetic counselling to support accurate understanding of inheritance	↑↑	Very low	1, 6, 12, 14, 31 →5, →13
Extend psychosocial support to immediate family members including siblings	↑↑	Very low	1, 6, 12, 14, 18–20, 25, 33, 34 →13 +26, +27, +32

Strength of recommendation measured using ↑↑, ↑, – or GPP. Good practice point (GPP) decided by expert panel.[16] The level of evidence was assessed using GRADE and was measured as very low, low, moderate or high.[16] Key references: arrows (→) indicate a clinical practice guideline; plus signs (+) indicate hospital guidelines. Increased presence of clinical practice guidelines or hospital guidelines influenced strength over appraised evidence. Detailed descriptions of the grading are given in Appendix S3 (see Supporting Information).

Table B.1 Summary of recommendations for hospital transfer and access to care in epidermolysis bullosa (EB)

Recommendation	Strength of recommendation	Level of evidence	Key references
R3 GPP Hospitals in low-resource settings and/or without access to EB specialist care can contact a specialist hospital for guidance			
Contact EB experts to gain support for appropriate EB treatment and to reduce trauma through handling or monitoring	GPP	Very low	GPP
R4 ↑ Hospital transfer to an EB specialist hospital should be discussed between the birthing hospital and the EB specialist team			
Consider EB severity, medical requirements, birthing hospital resources and staffing, and the availability of safe transport	↑	Very low	1, 14 +43, +45, +47, +48
R5 ↑↑ Neonates with EB require access to a multidisciplinary team			
Utilize management from a multidisciplinary care team to meet complex medical needs of the neonate	↑↑	High	3, 9, 14, 21, 24, 31, 49 →2, →50 +43, +44, +45, +48, +51

Strength of recommendation measured using ↑↑, ↑, – or GPP. Good practice point (GPP) decided by expert panel.[16] The level of evidence was assessed using GRADE and was measured as very low, low, moderate or high.[16] Key references: arrows (→) indicate a clinical practice guideline; plus signs (+) indicate hospital guidelines. Increased presence of clinical practice guidelines or hospital guidelines influenced strength over appraised evidence. Detailed descriptions of the grading are given in Appendix S3 (see Supporting Information).

- Offer ongoing EB education and psychosocial support while waiting for genetic results.[1,3,9,10,12,13,23–25]
- Access an EB specialist to provide diagnostic and management education for parents, carers and extended family members.[1,6,14,18,20,25–27]
- Promote access to psychological support for the family if transfer away from community support structure is required for medical reasons.[11–14,19,20,22,28–30]
- Link to DEBRA organization for community support if available (GPP).

R2 ↑↑ Provide education and support for parents and carers throughout diagnosis and prognosis.

- Offer genetic counselling to outline inheritance patterns and support the family with any feelings of guilt or blame.[1,3,5,6,12–14,31] Prenatal testing and *in vitro* fertilization with preimplantation genetic diagnosis can be discussed as an option to educate families about risk of recurrence in future pregnancies.[1,3,5,6]
- Offer educational support for siblings or extended family. If parents request support, provide resources on diagnosis and ways to safely 'play' or interact with a neonate with EB to help reduce the risk of injury at home.[1,6,12–14,19,20,26,27,32–34]
- Offer psychological and palliative care (if appropriate), to discuss realistic expectations and prognosis[1,3,5,12.13,18,25,35] and to support ethical or moral dilemmas if fatal forms of EB are suspected.[13,35]
- Offer copies of medical images or scans to parents in cases of end-of-life care (GPP).

B: Hospital management: medical monitoring, wound care and pain

Neonatal EB care requires early access to an EB specialist team, at the specialist hospital or through an established specialist EB outreach service. Management can vary from mild to complex, possibly requiring surgical intervention, as well as palliative care. An MDT approach

Table B.2a Summary of recommendations for postnatal epidermolysis bullosa (EB) care

Recommendations	Strength of recommendation	Level of evidence	Key references
R6 ↑ Consider implementing postnatal preventative EB care			
Secure the umbilical cord with a ligature or a rubber cord ring rather than a cord clamp, to prevent trauma around the umbilicus	↑	Low	1 →2 +26, +43, +48, +51, +52
Cover any birth trauma with cling film or a nonadherent dressing to prevent further trauma, and reduce pain and exposure to infection	↑	Very low	+46, +48, +49
Apply padded layer around neonate if stimulation for breathing is required after birth	GPP	–	Expert panel consensus
Apply emollient to suction catheter tips required for mucus or meconium removal; this will help to avoid friction damage to the mucosa	↑↑	High	1, 9, 10, 31, 40 →2, →36, →54 +45, +48
Promote skin-to-skin contact with mother once wounds are covered and neonate is well	↑↑	Low	1, 9, 23 →2, →36
Weigh neonate wrapped in a blanket. Tare weighing scale to zero to include a soft cloth or towel prior to weighing neonate, to reduce movement and avoid trauma from being handled naked	↑	Low	23 +26, +27, +43, +45
Attach hospital ID band over clothing or socks, not directly on skin	↑↑	High	1, 14, 31, 39 →2, →36 +26, +27, +32, +43, +45
Use a venous sample for newborn screening test on day 2–3 of life, to reduce friction to heel and avoid degloving injury. Mark sample as 'venous' on test card	↑	High	9, 40 →2 +44
Cluster any blood tests to reduce use of tourniquet and handling that might cause trauma	↑	Low	3 →2, →5
Give vitamin K, hepatitis B vaccine or any country-specific vaccination required at birth. Use a low-adherent dressing to cover site if required	↑↑	Low	1, 3, 31, 40 →2 +26, +43, +44, +45, +51
R7 ↑↑ Consider the impact of heat and friction if temperature regulation support is required			
Do not nurse the neonate in an incubator, unless for medical reason such a prematurity. Heat or humidity exacerbates blistering	↑↑	High	1, 9, 14, 42 →2 +26, +43, +45, +51, +52
Only use overhead heaters during procedures. Use blankets to maintain temperature	↑↑	High	23, 31 +47, +52
Use a pressure-relieving, soft mattress, or extra soft padding on beds, such as incubator mattress	↑↑	High	1, 14, 31 +26, +27, +43, +45, +51, +52

Strength of recommendation measured using ↑↑, ↑, – or GPP. Good practice point (GPP) decided by expert panel.[16] The level of evidence was assessed using GRADE and was measured as very low, low, moderate or high.[16] Key references: arrows (→) indicate a clinical practice guideline; plus signs (+) indicate hospital guidelines. Increased presence of clinical practice guidelines or hospital guidelines influenced strength over appraised evidence. Detailed descriptions of the grading are given in Appendix S3 (see Supporting Information).

is the gold standard for EB neonatal care (onsite if available, or utilizing telehealth) from birth, throughout the hospital admission, and in transfer of care to community health.[1–3,6,9,10,23,31,34,36–45] Comprehensive documentation by all teams will support MDT management and discharge planning. Documentation should include growth data, fluid input and output, pain scores, medical monitoring, wound status and clinical or genetic diagnosis (if results are available).[1,14,40,46]

B.1 Hospital transfer and access to a multidisciplinary care team

R3 GPP Hospitals in low-resource settings (LRSs) and/or without access to EB specialist care can contact a specialist hospital for guidance.

- Access DEBRA International, EB Without Borders or EB-CLINET for assistance linking with EB health professionals and/or an EB specialist hospital.
- The DEBRA International website provides outlines to emergency management.

Practical resources for LRS management are included throughout this guideline.

R4 ↑ Hospital transfer to an EB specialist hospital should be discussed between the birthing hospital and the EB specialist team.

- Consider the availability of EB outreach or a deployable nurse programme attending the local hospital rather than transferring the neonate (GPP).
- Consider EB severity, monitoring requirements, birthing hospital resources and staffing, and the availability of safe transport.[1,14,43,45,47,48]
- Pressure-redistributing mattresses will provide support during travel (GPP).
- Use barrier creams underneath transport straps, or protective clothing on the neonate to reduce friction.
- Always consider country-specific laws regarding car seat use (GPP).

R5 ↑↑ Neonates with EB require access to an MDT.

- Consider access to complex care and input from many medical disciplines.[2,3,9,14,21,24,31,43–45,48–51] Early inpatient neonatal EB care often includes dermatology, paediatrics or neonatology, genetics, nursing, dietetics, pain and psychosocial.

Table B.2b Summary of recommendations for neonatal epidermolysis bullosa (EB) preventative care and handling during hospital interventions and monitoring

Recommendation	Strength of recommendation	Level of evidence	Key references
R8 ↑↑ Use safe handling practices to protect the neonate's skin			
Place notices around the cot or bassinette to alert people to risk of trauma and need for preventative care	↑↑	High	1, 6, 14, 34, 39 +26, +27, +43, +45, +51
Encourage parents and carers to hold their newborn if they wish. Promote parenting skills to prepare for hospital discharge	↑	Low	1, 6
Gloves: use emollient or padding between gloved hands and neonates to reduce friction on skin	↑↑	Moderate	1, 31, 32, 39 →2, 36 +26, +27, +43, +45
Pick up neonates with EB using side roll technique, holding under neck and nappy. Do not lift holding under the arms	↑↑	Moderate	1, 6, 34 +26, +27, +45, +52
R9 ↑↑ Implement preventative care for all procedural monitoring			
Observation monitoring			
Frequency: individualize blood pressure, $Sa0_2$ and temperature monitoring requirements	↑↑	High	1, 6, 9, 37, 38, 55 +43, +45, +48
Use axillar temperature probes with emollient, lift arm to place and remove. Use digital infrared thermometer if available	↑↑	High	1, 3, 6, 9, 23, 31, 55 →2, →36 +43, +45
Oxygen monitoring: use soft cotton or silicone dressing between monitor and skin	↑↑	High	1, 6, 9, 37, 38, 40, 55 →2, →36 +43, +45, +48, +52, +53
Blood pressure and tourniquets: consider whether medically needed for monitoring. If required use padding or clothing between cuff and skin	↑↑	High	1, 6, 9, 37, 38, 40, 55 →2, →36 +43, +45, +48, +52, +53
Fluid balance: maintain fluid balance records and document bilious or repetitive vomiting or abdominal distention that may indicate ultrasound exploration for pyloric atresia	↑↑	High	1, 7, 24, 31, 40, 56
Urine collection: avoid adhesive bags, attempt clean-catch collection or dipstick testing	GPP	–	Expert panel consensus
Cannulation and blood tests			
Intravenous cannulation: use experienced practitioner to reduce potential for multiple attempts. Use low-adherent tape to secure, and preventative padding under tourniquets	↑↑	High	6, 10, 31, 38–40, 55 →2, →36 +43, +45, +51, +53
Blood tests: precautionary blood tests are not recommended. Only monitor if clinically indicated, to reduce unnecessary friction and blood volume drawn	↑	Low	1, 31, 40 +45, +51
Blood tests: prioritize requested tests in cases of insufficient blood volumes	↑	Low	3, 14, 46, 57 →2
Cluster tests to include genetic bloods, newborn screening, and any monitoring required			
Procedures requiring preventative lubrication			
Feeding tubes and catheters to be lubricated well with a water-based lubricant. Use low-adhesive tape to secure to face	↑↑	High	1, 9, 31, 39 →2, →36 +43, +45, +53
Oral or cavity swabbing to be lubricated with saline before performing swab	↑↑	High	1, 3, 40, 42 →54 +43, +45
Corneal abrasions: refer to ophthalmology, use nonmedicated hydrating eyedrops or gel to reduce friction in and around eyes	↑	Low	23, 58, 59 →41
Anal fissures and blisters: use petroleum-based lubricant around the anus to reduce blisters and pain while passing stools	↑	Very low	23 →2
Management of potential neonatal complications			
Jaundice: if phototherapy is required, gain support from an EB specialist. Consider friction risk of trauma from incubator, or use of a biliblanket	GPP	–	Expert panel consensus
Pyloric atresia: this requires transfer to an EB facility with neonatal intensive care. Surgical and anaesthetic procedures require full preventative care. Provide access to a specialist EB multidisciplinary team and neonatal medical support	↑↑	High	7, 56, 60–64

Strength of recommendation measured using ↑↑, ↑, – or GPP. Good practice point (GPP) decided by expert panel.[16] The level of evidence was assessed using GRADE and was measured as very low, low, moderate or high.[16] Key references: arrows (→) indicate a clinical practice guideline; plus signs (+) indicate hospital guidelines. Increased presence of clinical practice guidelines or hospital guidelines influenced strength over appraised evidence. Detailed descriptions of the grading are given in Appendix S3 (see Supporting Information).

Table B.2c Summary of recommendations for emergency neonatal management in epidermolysis bullosa (EB)

Recommendation	Strength of recommendation	Level of evidence	Key references
R10 ↑↑ Implement measures to minimize harm during anaesthetic and emergency procedures			
Consultation between anaesthetist and EB staff to precede surgery. Ensure inclusion of preventative theatre taping techniques	↑↑	High	1, 6, 9, 10, 23, 40, 57 →2, →36, →54 +53
Intubation: lubricate laryngoscope and endotracheal tube. Use low-adherent tape on skin underneath standard securing tape	↑↑	High	40, 55, 65 →2, →36, →54 +53
Face masks: use soft inflated-edge anaesthetic masks. Use water-based lubricant or foam padding between mask edge and the neonate's face. Apply padding between gloved hands and neonate's jaw while forming seal with mask	↑↑	High	1, 6, 9, 10, 23, 40, 55, 57 →2, →36, →50, →54 +43, +51, +53
An EB nurse escort should follow the patient through medical and surgical procedures	↑↑	High	6, 9, 14 →36 +45

Strength of recommendation measured using ↑↑, ↑, – or GPP. Good practice point (GPP) decided by expert panel.[16] The level of evidence was assessed using GRADE and was measured as very low, low, moderate or high.[16] Key references: arrows (→) indicate a clinical practice guideline; plus signs (+) indicate hospital guidelines. Increased presence of clinical practice guidelines or hospital guidelines influenced strength over appraised evidence. Detailed descriptions of the grading are given in Appendix S3 (see Supporting Information).

Table B.3 Summary of recommendations for neonatal wound care in epidermolysis bullosa (EB)

Recommendation	Strength of recommendation	Level of evidence	Key references
R11 ↑↑ Implement neonatal EB wound care strategies			
Use silicone medical adhesive removers (SMARs) to remove any tape or wound care product adhered to the skin	↑↑	High	1, 3, 6, 9, 14, 23, 31, 38, 40, 55, 65 →2, →36 +32, +45
Use a thin nonadherent contact layer on wounds for improved wound contact on small neonatal bony prominences	↑↑	High	1, 3, 6, 9, 14, 23, 31, 39 →2, →36 +43, +45, +51
Secure contact layer with a thin foam secondary layer (e.g. Mepilex Lite) and bandage in place with a small gauze bandage or tubular bandage	↑↑	High	1, 3, 6, 9, 14, 23, 31, 39 →2, →36 +43, +45, +51
Warm wound cleansing solution to reduce pain while cleaning	↑↑	High	1 →41 +45
Debridement requires dermatology input for EB subtype and pain management, attempt enzymatic and autolytic debridement prior to surgical	↑↑	High	→2, →36, →41, →67
Gradually increase parents' participation in all wound care, consider their emotional distress and birth recovery	↑	Low	1, 9, 19 +27, +43, +45
R12 ↑↑ Use preventative care to reduce friction during neonatal wound care			
Dress wounds one limb at a time. Wrap neonate to secure other limbs to reduce movement and potential new trauma, and to avoid increased pain from friction to exposed wounds	↑↑	High	1, 9, 14, 31 →2 +43, +45, +48
Use tubular (e.g. Tubifast) or gauze bandages to secure dressings or devices rather than tape	↑↑	High	1, 3, 14 →2 +43
Scalp blisters: use lubrication or a foam pad if neonate has increased movement. Avoid shaving hair to reduce risk of long-term damage	↑	Low	1 +26, +27
Use of emollients on wound care products can reduce shearing on wound surface when removed	↑↑	High	1, 6, 14, 23, 42 →2 +43
R13 ↑↑ Include parents in blister education and management			
Include parents in blister management and education with a skilled EB nurse or dermatologist to prepare for hospital discharge	↑↑	Low	1, 9, 14, 20, 37 +26, +27, +43, +45, +51, +68
To reduce pain and blister size, lance and drain neonatal EB blisters as soon as possible. Review for new blisters with each nappy change	↑↑	High	1, 3, 6, 9, 10, 23, 31, 38, 40, 42 →2 +45, +51

Strength of recommendation measured using ↑↑, ↑, – or GPP. Good practice point (GPP) decided by expert panel.[16] The level of evidence was assessed using GRADE and was measured as very low, low, moderate or high.[16] Key references: arrows (→) indicate a clinical practice guideline; plus signs (+) indicate hospital guidelines. Increased presence of clinical practice guidelines or hospital guidelines influenced strength over appraised evidence. Detailed descriptions of the grading are given in Appendix S3 (see Supporting Information).

Table B.4 Summary of recommendations for neonatal infection and sepsis in epidermolysis bullosa (EB)

Recommendation	Strength of recommendation	Level of evidence	Key references
R14 ↑↑ Always include a dermatologist in planning wound management and rotation			
Use antimicrobial soaks in nonsymptomatic positive wounds cultures	↑↑	Low	1, 10, 31 →2, →67
Use antimicrobial soaks in combination with systemic antibiotics if wound shows signs of spreading or presence of systemic infection	↑↑	Low	1, 10 →2, →67
Test for *Staphylococcus aureus* and candida to differentiate between EB oral plaques, feeding pain and candida	GPP	Very low	37, expert panel consensus
Medical-grade honey can be used in neonates	↑↑	Very low	1, 10 →2, →67
Use of diluted bleach or vinegar is not recommended for use in neonates	↓↓	Very low	1, 6, 9, 23 →2, →67
Use of silver is not recommended as first-line treatment of infections in neonates	↓↓	Low	1 →67
R15 ↑↑ Select antibiotics based on swab results and wound symptoms in neonates with EB to reduce risk of resistance			
Antibiotics are not always required for asymptomatic wounds in an asymptomatic neonate with a positive bacterial swab. Utilize antimicrobials first	↑↑	High	1, 6, 10, 23, 31, 38 →2, →67
Limit topical antibiotics and use in rotation to avoid development of microbial resistance	↑↑	High	1, 6, 10, 38 →2, →67
Enteral antibiotics should be used for symptomatic wounds where no systemic symptoms are present. Consider trauma risk from intravenous cannulation	↑↑	High	1, 6, 23, 31 →67
Intravenous antibiotics should be used to treat the systemically unwell neonate to reduce the risk of sepsis and to provide rapid response to treatment	↑↑	High	1, 10, 31 →2
R16 ↑ Monitor neonates closely for signs of sepsis			
Neonatal sepsis should be addressed with urgency due to the risk of rapid deterioration for severe subtypes, such as EBS generalized severe and junctional EB	↑	Moderate	1, 6, 31, 40 →2
Monitor wounds, pain, appetite, fever and lethargy to identify early deterioration. Apply continuous monitoring to the systemically unwell neonate	↑↑	High	1, 9, 10, 31, 40 →2 +32
Urgently address signs of sepsis in neonates with EB, particularly in severe subtypes such as junctional EB with a lack of laminin 332	↑	Very low	1, 69

Strength of recommendation measured using ↑↑, ↑, – or GPP. Good practice point (GPP) decided by expert panel.[16] The level of evidence was assessed using GRADE and was measured as very low, low, moderate or high.[16] Key references: arrows (→) indicate a clinical practice guideline; plus signs (+) indicate hospital guidelines. Increased presence of clinical practice guidelines or hospital guidelines influenced strength over appraised evidence. Detailed descriptions of the grading are given in Appendix S3 (see Supporting Information).

Table B.5 Summary of recommendations for neonatal pain management for neonates with epidermolysis bullosa (EB)

Recommendation	Strength of recommendation	Level of evidence	Key references
R17 ↑↑ Always monitor pain using a validated neonatal pain scale			
Use a validated neonatal pain scale, e.g. Neonatal Infant Pain Scale (NIPS) or Face, Legs, Activity, Cry and Consolability (FLACC), for effective pain management	↑↑	High	1, 10, 40, 44, 70 →41
R18 ↑ Consider providing parents and carers with pain education and psychosocial support			
Provide psychosocial support to help parents and carers understand difficulties with handling their neonate in pain	↑↑	Low	1, 3, 9, 12, 18–20, 25, 71 →2, →13, →31
Include parents and carers in hospital pain management plans and implementation	↑	Low	31, 39, 72
Parents must be competent at identifying, measuring and administering pain medication, and as well as nontherapeutic relief strategies	↑↑	Low	1, 3, 6, 9, 14, 19, 39, 71, 72 →2, →41 +43, +45, +51
R19 ↑↑ Provide appropriate neonatal pain relief			
Use sucrose, paracetamol/acetaminophen, and/or oral morphine/oxycodone as first-line medications for pain relief	↑↑	Moderate	1, 3, 23, 72 →41 +44, +51
Refer to acute pain team if first-line medications are not sufficient	↑↑	Moderate	3, 40 →41 +43, +44, +45, +51
It is recommended that medical monitoring and resuscitation equipment be available	↑↑	Moderate	1, 6, 40 →41
Seek country- or region-specific neonatal prescribing guidelines to ensure safe dosage, monitoring and weaning	↑↑	High	3, 40, 73, 74 →41

Strength of recommendation measured using ↑↑, ↑, – or GPP. Good practice point (GPP) decided by expert panel.[16] The level of evidence was assessed using GRADE and was measured as very low, low, moderate or high.[16] Key references: arrows (→) indicate a clinical practice guideline; plus signs (+) indicate hospital guidelines. Increased presence of clinical practice guidelines or hospital guidelines influenced strength over appraised evidence. Detailed descriptions of the grading are given in Appendix S3 (see Supporting Information).

Table C.1 Summary of recommendations for neonatal dietitian referral and nutritional requirements in epidermolysis bullosa (EB)

Recommendation	Strength of recommendation	Level of evidence	Key references
R20 ↑↑ Refer to a neonatal dietitian to support intake and nutritional status if mucosal blistering and/or pain are present			
It is strongly recommended that a dietitian be linked to neonatal EB management	↑↑	High	8, 10, 24, 38, 46, 72, 77 +26, +53
Include pain relief prior to feeding if oral mucosa is impacted. Consider oral paracetamol and/or analgesic gel on neonate's mouth	↑↑	High	1, 3, 6, 9, 10, 23, 35, 37, 46, 49, 55, 57 →2, →41, →54 +43
Encourage breastfeeding and/or expressing breast milk	↑↑	Moderate	57, 76
A barrier cream or emollient can be applied to the breast nipple, the bottle teat and the neonate's cheek	↑↑	High	6, 9, 10, 23, 38, 39, 42, 55 →2, →36, →50, →54 +26, +45, +51, +52
Use a soft silicone bottle teat or a Haberman feeder. Place bottle teat in warm water prior to use to soften	↑↑	High	1, 6, 9, 10, 14, 24, 31, 35, 49, 57, 76 →2, →50 +43, +52
Refer the neonate with suspected pyloric atresia to surgical teams	↑↑	High	7, 56, 60–64
R21 ↑↑ Calculate nutritional requirements considering wound losses and feeding capacity			
Energy: calculate calorie requirements using ideal bodyweight, ratio of blisters to body surface area, exudate losses, additional injury or activity factor, and the need for catch-up growth. Neonates with severe subtypes of EB will require to 150–200% of their standard nutritional needs	↑↑	High	6, 24, 57, 76, 78
Protein: monitor biochemical protein profile, and wound and blister burden on the body surface when estimating 1.5–4 g protein per kg per day	↑↑	High	31, 57, 76, 78, 79
Sodium: supplement sodium if energy provision appears adequate and exudate losses are high, but weight continues to falter	↑↑	High	10, 24, 31, 76, 80
Vitamins and minerals: for large degloving wounds, monitor iron, zinc, vitamin K and vitamin D levels. Consider prophylactic multivitamin and mineral. Vitamin requirements in children with EB can be 150–200% of the recommended daily intake	↑↑	High	1, 24, 38, 57, 76, 81, 82
If **vitamin K** is not given at birth monitor vitamin K and PIVKA-II. Serum vitamin K is not a specific test and should not be measured in isolation	↑	High	81
Inflammation: interpret serology markers with caution, with careful consideration of impact of inflammation on certain biomarkers and nutritional status. Acute-phase reactants are common with EB due to high levels of inflammation	↑↑	High	57, 80, 82
If **breast milk** is not available and growth is faltering, use a high-energy or high-protein polymeric formula	↑↑	High	3, 76, 78

Strength of recommendation measured using ↑↑, ↑, – or GPP. Good practice point (GPP) decided by expert panel.[16] The level of evidence was assessed using GRADE and was measured as very low, low, moderate or high.[16] Key references: arrows (→) indicate a clinical practice guideline; plus signs (+) indicate hospital guidelines. Increased presence of clinical practice guidelines or hospital guidelines influenced strength over appraised evidence. Detailed descriptions of the grading are given in Appendix S3 (see Supporting Information).

- Neonates with severe subtypes of EB may require access to neonatal surgery, intensive care, anaesthetics or palliative care.
- In addition to inpatient care teams, additional teams are required for ongoing community care, including physiotherapy, occupational therapy, community nursing, general practitioner, and DEBRA organization support.
- Teams and roles may vary depending on geographical location.

B.2 Medical monitoring and risk factors in postnatal epidermolysis bullosa care

Neonatal EB skin fragility can lead to blistering or wounds from any handling, and therefore all medical interventions and monitoring must be adapted appropriately.[1–3,9,23,32,37,38,40,45,48]

B.2a Recommendations for postnatal epidermolysis bullosa care

R6 ↑ Consider implementing postnatal preventative EB care.

- Preventative care is recommended for any neonates with a suspected or confirmed diagnosis of EB within the first 48 h of life. If a mild subtype of EB is suspected, the EB specialist can guide transition to standard neonatal handling.
- Use padding between weighing scales, ID band, nappy or tourniquet and the neonate's skin.[1,2,31,26,27,32,36,39,43,45] Pin identification bands to clothing or attach to the bassinette.
- Tie the umbilical cord with ligature, do not use a cord clamp.[1,2,26,43,48,51,52]
- Cover birth trauma (large wounds) with low-adherent dressing or, if unavailable, use cling film until low-adhesive wound care is available.[51–53]
- Use emollient on oral or nasal suction catheters.[1,2,9,10,31,36,40,45,48,54] Use the lowest effective suction pressure, avoiding the sides of the mouth while suctioning oral secretions (GPP).
- Apply a padded layer around the neonate if stimulation for breathing is required after birth (GPP).
- Encourage postbirth skin-to-skin contact between the mother, parent or carer and the neonate, once wounds are covered.[1,2,9,23,36]
- Give scheduled vaccinations[1–3,31,26,40,43–45,51] and avoid use of adhesive dressings.

Table C.2 Summary of recommendations for neonatal enteral nutrition in epidermolysis bullosa (EB)

Recommendation	Strength of recommendation	Level of evidence	Key references
R22 ↑ Consider using enteral feeding in neonates with EB not able to meet full nutritional requirements			
Attempt all oral feeding strategies for neonates with oral and oesophageal mucosal fragility prior to insertion of a nasogastric tube (NGT)	↑	Very low	1, 14, 31, 35, 49, 57, 76, 83, 84
There should be a low tolerance of reduced feeding for early insertion of an NGT for severe subtypes of EB	↑	Very low	1, 46, 49, 57, 76
R23 ↑ Consider using experienced EB staff for NGT insertion and monitoring			
An experienced staff member should insert the lubricated NGT. This will reduce friction on internal mucosa and reduce the risk of needing multiple passes, which can have long-term effects on the mucosal lining	↑	Very low	1, 9, 10, 31
Use of NGTs (rather than orogastric tubes) in neonates may be indicated where the pharyngo-oesophageal mucosal lining is not at risk	↑	Very low	31, 57, 76
Use NGTs for severe subtypes of EB when oral intake is not meeting requirements, but feed tolerance is normal	↑	Very low	31, 85
Use nasojejunal feeding for neonates with severe reflux, vomiting or risk of aspiration	↑	Very low	80
To secure NGT use a low-adherent film as a contact layer, with a full-adherent tape that secures the tube to the film, avoiding contact with the neonate's skin. Use lasso technique if adherence is an issue	↑↑	High	1, 9, 10 →2, →36 +53
R24 ↑ Suggest maintaining dietetic input throughout enteral feeding use			
Consider EB subtype, hospital capacity, parenting skills and community support after discharge to ensure enteral feeding is sustainable	↑	Low	1, 11, 31, 34, 44, 49 →2 +43, +45
Maintain a degree of oral feeding (if safe) or oral stimulation if enteral tube feeding to support oral stimulation and reduce risk of oral aversion	↑	Low	57, 85
Consider continuous feeding regimens if feed tolerance is poor with gastro-oesophageal reflux or vomiting present	↑	Low	57, 85

Strength of recommendation measured using ↑↑, ↑, – or GPP. Good practice point (GPP) decided by expert panel.[16] The level of evidence was assessed using GRADE and was measured as very low, low, moderate or high.[16] Key references: arrows (→) indicate a clinical practice guideline; plus signs (+) indicate hospital guidelines. Increased presence of clinical practice guidelines or hospital guidelines influenced strength over appraised evidence. Detailed descriptions of the grading are given in Appendix S3 (see Supporting Information).

Table C.3 Summary of recommendations for neonatal gastrostomy use and parental nutrition in epidermolysis bullosa (EB)

Recommendation	Strength of recommendation	Level of evidence	Key references
R25 ↑ Consider gastrostomy feeding to support nutritional requirements for neonates with severe EB			
Provide early education directed at parents and carers to normalize enteral feeding for severe forms of EB to support understanding and improve acceptance if future insertion is required	↑	Low	9, 19, 31
Optimize feeding and nutrition prior to gastrostomy insertion surgery to promote wound healing and improve surgical outcomes	↑	Medium	24, 84, 87 →2
Consider gastrostomy if long-term supplemental nutrition is required and fixation of nasogastric tube or nasojejunal tube is problematic	↑↑	Medium	7, 24, 35, 86–88 →2
Gastrostomy placement should be undertaken by an experienced surgeon using the modified two-port laparoscopic approach using the Seldinger technique, with serial dilatation and tube insertion through a peel-away sheath (LAG technique)	↑	Low	89, 90
R26 ↑ Consider parental nutrition (PN) to support nutrition for neonates with EB			
Consider PN to optimize nutrition prior to insertion of gastrostomy or surgical management of pyloric atresia	↑	Very low	1, 24, 57, 64
Continue trophic enteral feeding or small oral feeds (where possible) during PN use to maintain integrity of intestinal mucosa and gastrointestinal endocrine function	↑	Very low	57
Consider PN use while umbilical line is accessible and the neonate is being stabilized	↑	Very low	1
Perform line care, dressings and monitoring as advised by the EB nurse, with signs of infection or sepsis addressed rapidly	↑	Low	1, 9, 14, 40 +45

Strength of recommendation measured using ↑↑, ↑, – or GPP. Good practice point (GPP) decided by expert panel.[16] The level of evidence was assessed using GRADE and was measured as very low, low, moderate or high.[16] Key references: arrows (→) indicate a clinical practice guideline; plus signs (+) indicate hospital guidelines. Increased presence of clinical practice guidelines or hospital guidelines influenced strength over appraised evidence. Detailed descriptions of the grading are given in Appendix S3 (see Supporting Information).

Table D Summary of recommendations for neonatal epidermolysis bullosa (EB) management for feeding, nappies/diapers, bathing and clothing

Recommendation	Strength of recommendation	Level of evidence	Key references
Feeding			
R27 ↑↑ Support feeding tolerance through pain management and friction reduction			
Always use emollient on pacifier, limit use if blister or wounds near mouth are present	↑↑	High	1, 6, 9, 14, 23 →2, →41, →54 +26, +27, +43, +47, +51
Use emollient on the teat, bottle, breast or nipple as well as the neonate's lips	↑↑	High	2, 6, 10, 23, 38, 39, 42, 50, 55 →2, →36, →54 +26, +45, +51, +52
Use patting, tapping or gentle rubbing to support burping and settling after a feed	↑↑	High	1, 6, 9, 23, 39, 50 +26, +27
Give nonsedating pain relief for 20 min prior to feeding for neonates with oral blistering or plaque	↑↑	High	1, 6, 9, 10, 23, 31, 37 →2, →41 +43
Use a soft silicone or cleft-palate-style teat for neonates with oral pain and blistering. Warming to soften before use may help	↑↑	High	1, 9, 10, 14, 31, 35, 49, 50, 57, 76 →2 +43, +52
Nappies/diapers			
R28 ↑↑ Use emollient and liners to reduce friction from nappy/diaper elastic on skin			
Clean nappy/diaper area with emollient ointment in preference to water or commercial wipes. If skin has blistered use a hydrogel dressing such as Intrasite Conformable dressing	↑↑	High	1, 9 →2, →36, →50 +26, +43, +52
Use well-fitted standard nappies/diapers. Trim off the inner elastic of disposable nappies/diapers to reduce friction	↑↑	High	1, 9, 42 →2, →50 +26, +27
Line nappy/diaper with a soft cloth liner coated with emollient or paraffin-impregnated gauze. Barrier creams and dressings may be useful	↑↑	High	1, 7, 16, 20, 23, 42 →2
Bathing			
R29 ↑↑ Always include a dermatologist, an EB nurse and pain medication in bathing management			
Swaddle neonates with birth trauma and clean limb by limb during wound care until effective pain relief has been established then move to a full bath	↑↑	High	1, 6, 9, 14, 20, 23, 31 →2, →41 +26, +45, +48, +51
If adequate pain relief is available wrap the neonate in a thin cloth, then soak in a deep padded bath, allowing dressings to come loose. To pad the bath place a soft blanket underwater under the neonate	↑↑	High	1, 6, 20, 23, 38 →2 +43, +44, +45
Add 9 g of salt to each one litre of bath water to create a saline bath	GPP	Low	→41
After bath, wrap the neonate in towels (with loose dressings in place) and allow to dry while holding; do not rub dry	GPP	–	Expert panel consensus
When dry, swaddle the neonate in a warm, dry, soft blanket exposing one limb at a time as new for old dressings are attended to	↑↑	High	1, 6, 23, 38 +43, +45, +51
Clothing			
R30 ↑↑ Keep the neonate dressed and wrapped to reduce movement			
Keep the neonate dressed and wrapped to reduce trauma from movement and friction	↑↑	High	1, 3, 6, 10, 14, 20, 23 →41, →50 +26, +27, +45, +47, +51
Dress clothing inside out and, if possible, select natural fibres. Reduce trauma from seams, buttons, press-studs or snap-fasteners touching the skin	↑↑	High	1, 3, 6, 10, 14, 20, 23 →41 +26, +27, +45, +47, +51
Apply wound dressing or barrier cream to bony prominences such as heels and elbows, as well as trauma-exposed sites, to protect the fragile skin	↑↑	High	1, 3, 10, 23, 31 →50 +26, +27, +43, +45, +48

Strength of recommendation measured using ↑↑, ↑, – or GPP. Good practice point (GPP) decided by expert panel.[16] The level of evidence was assessed using GRADE and was measured as very low, low, moderate or high.[16] Key references: arrows (→) indicate a clinical practice guideline; plus signs (+) indicate hospital guidelines. Increased presence of clinical practice guidelines or hospital guidelines influenced strength over appraised evidence. Detailed descriptions of the grading are given in Appendix S3 (see Supporting Information).

- Use a venous sample for newborn genetic screening tests, rather than heel prick.[2,9,40,44]
- Consider pain relief for neonates with large birth trauma.[1–3,6,38]

R7 ↑↑ Consider the impact of heat and friction if temperature regulation support is required.

Heat can exacerbate blistering for neonates with EB.[1,2,9,14,26,42,43,45,51,52]

LRS: Temperature regulation through kangaroo care (holding the neonate skin to skin on an adult's chest) is appropriate for neonates with EB; ensure wounds are covered first.

Temperature

- Consider medical comorbidities and gestational age for frequency of monitoring.

Table E Summary of recommendations for neonatal discharge planning, care coordination and epidermolysis bullosa (EB) education

Recommendation	Strength of recommendation	Level of evidence	Key references
EB education			
R31 ↑ Offer EB education to prepare parents, carers and community providers for complex disease management at home			
Start education in hospital, consider parents' postpartum recovery, pain and sleep	↑	Very low	1, 9, 10, 14, 19, 20, 25, 29, 71
Offer repetitive education in different formats (spoken, practical demonstrations, written and weblinks) to support learning	↑	Very low	9, 19, 20, 25, 28, 29, 31, 71
The EB specialist centre can provide education to community providers listed in the discharge plan prior to discharge	↑	Very low	10, 14, 19, 20, 42 +26, +43, +45, +51
Use of a comprehensive education checklist will assist with parental discharge planning (Appendix S5)	GPP	Very low	+68, expert panel consensus
Discharge planning			
R32 ↑ Consider using a comprehensive discharge plan to support EB management to continue after discharge			
Establish organizational and health system support that includes an individualized care plan with all specialist and local health provider contacts	↑	Very low	10, 20, 31, 40, 42 →2 +44, +68
Hospital discharge can be considered when general health condition is stable, and the parents or carers have demonstrated competence in neonatal EB management	↑	Very low	1, 3, 6, 10 +44
The discharge plan should encourage communication between parents and teams. Provide a single contact point for appropriate EB emergency management	↑	Very low	1, 9, 14, 31, 42 →2
Parents and carers should be offered support from their country-specific DEBRA organization, and peer support, in order to provide peer and practical support away from the clinical setting	↑	Very low	12, 19, 20, 71 →13
Care coordination			
R33 ↑ Consider using a care coordination plan with appointments, contacts and emergency management to support parents, carers and local providers			
The EB specialist team should continue to guide care and review results within the community until all practitioners have received EB education	↑	Very low	10, 14, 19, 20, 31 +26, +43, +45, +51
Open communication between specialist team, general practitioner and homecare team is suggested for effective care coordination	↑	Very low	10, 19, 20, 25
Parents and carers require a single point of contact for a care coordinator role; this needs to be an EB expert	↑	Very low	12, 14, 19, 20, 30
Local providers will benefit from ongoing contact with the specialist hospital through different stages of growth and development	↑	Very low	12, 14, 19, 20, 25, 30

Strength of recommendation measured using ↑↑, ↑, – or GPP. Good practice point (GPP) decided by expert panel.[16] The level of evidence was assessed using GRADE and was measured as very low, low, moderate or high.[16] Key references: arrows (→) indicate a clinical practice guideline; plus signs (+) indicate hospital guidelines. Increased presence of clinical practice guidelines or hospital guidelines influenced strength over appraised evidence. Detailed descriptions of the grading are given in Appendix S3 (see Supporting Information).

- If the neonate is maintaining their own body temperature, use spot checks rather than adhesive continuous monitoring.
- Use emollient on thermometers to reduce friction during axilla insertion and removal.
- If available, consider using an infrared thermometer (GPP).

Incubators and overhead heaters

- Do not nurse the neonate in an incubator unless for coexisting medical conditions.[1,2,9,14,26,42,43,45,51,52]
- Only use overhead heaters for procedures.[23,31,47,52] Depending on gestational age and weight at birth, utilize swaddling and extra blankets to support temperature regulation.

Mattresses and blankets

- Use a pressure-redistributing soft mattress, or extra soft padding on beds, such as an incubator mattress.[1,14,26,27,31,43,45,51,52]
- Swaddle the neonate in a soft cloth to reduce movement to reduce risk of blister development.[1,9,26,27]
- Carefully flatten blankets, and minimize creases when swaddling a neonate with a severe subtype of EB to reduce blisters or wounds resulting from blanket creases on the skin (GPP).

B.2b Preventative care for handling, interventions and monitoring in hospital

R8 ↑↑ Use safe handling practices to protect the neonate's skin.

- Communicate preventative care strategies including handling techniques using printed signage around the cot.[1,6,14,26,27,34,39,43,45,51]
- Encourage parents and carers to hold their neonate to promote confidence with handling.[1,6,26,34,43,45]
- Apply emollient or a padding barrier between gloved hands and the neonate's skin.[1,2,26,27,31,32,36,39,43,45] Take caution to handle and lift the neonate safely when emollient is used (GPP).

- Pick up using flat hands and a side roll technique, holding under the neck and nappy/diaper. Do not lift holding under the arms.[1,6,26,27,34,45,52]

R9 ↑↑ Implement preventative care for all procedural monitoring.

Any handling or monitoring could cause trauma, exacerbation of blistering and increased pain.[1–3,9,10,23,31,32,34,36–45,48,54] Refer to Appendix S5 for the implementation tool.

Implement the following preventative care during monitoring.

Observation monitoring

- Individualize frequency of observation based on severity.[1,6,9,37,38,43,45,48,55]
- Use padding or lubrication between all medical devices and the neonates' skin, including thermometer, blood pressure (BP) cuff, oxygen saturation equipment and tourniquets.[1,2,6,9,36–38,40,43,45,48,52,53]
- Maintain strict input and output fluid balance charts.[1,7,24,31,40,56]
- Use dip stick or clean-catch urine collection, do not adhere urine bag to skin (GPP).

Cannulation and blood tests

- Use preventative padding under tourniquet and gloves, use an experienced blood-draw technician or anaesthesiologist to reduce risk of multiple attempts.[2,6,10,31,36,38–40,43,45,51,53,55]
- Precautionary testing is not advisable, only monitor if clinically indicated.[1,31,40,45,51]
- Consider combining tests to include genetic bloods if appropriate and mark their order of priority in case of insufficient sample.[14,46,57]

Procedures requiring preventative lubrication

- Cover feeding tubes and suction catheter tips with emollient.[1,2,9,31,36,39,43,45,53]
- Oral swabs can be moistened with normal saline.[1,3,40,42,43,45,54]
- Use nonmedicated lubricating drops or gel in eyes to reduce corneal abrasions from rubbing.[23,41,58,59]
- Use emollient around the anus if fissures or blistering are present.[2,23]

Management of potential neonatal complications

- Jaundice treatment should consider potential EB subtype, clinical severity and access to treatment and pathology. Contact an EB specialist if phototherapy is required (GPP).
- Pyloric atresia requires transfer to an EB facility if available, alternatively referral to a paediatric surgery with intensive care unit while utilizing practical advice from the EB centre. Surgical and anaesthetic procedures require full preventative care.[7,56,60–64]
- Link external treating teams to EB specialist for elective procedures, such as male circumcision, to consider clinical stability and subtype-specific management (GPP).

B.2c Emergency management

R10 ↑↑ Implement measures to minimize harm during anaesthetic and emergency procedures.

With the exception of neonates with established end-of-life plans, lifesaving support should always be the priority with unwell neonates. If an antenatal EB diagnosis is suspected, implementing hospital education and access to EB dressings prior to the birth may increase the chances of preventative care being implemented in an emergency.

- Implement preventative taping for all intubations and surgical management including line insertions, gastrostomy insertion, pyloromyotomy and tracheostomy.[1,2,6,9,10,23,36,40,53–55,57]
- Lubricate endotracheal tube and scopes. Use low-adherent padding between gloved hands and neonates' skin, use low-adherent taping underneath standard tape to secure tubes to neonate's face.[2,36,40,53–55,65]
- Use a water-based lubricant under face mask.[1,2,6,9,10,23,36,40,43,50,51,53–55,57]
- Where available, organize escort by an EB nurse for the entire patient procedure.[6,9,14,36,45]

Monitor wound healing closely, particularly if the EB subtype is unknown. Document the presence of milia, overgranulation, or maceration of healthy skin around wounds to support a clinical diagnosis, guide education and wound product selection. Utilize local or online EB specialists, consider EB-CLINET, EB Without Borders or DEBRA International to guide wound care. Always obtain and document family consent to share photos to guide a clinical diagnosis.

LRS: Consider using locally sourced emollient or padding to reduce adherence if low-adherent dressings are not available. Wound dressings can also be locally sourced; examples include banana leaf, potato peel dressings or coconut oil.[66] Other LRS wound care options are outlined in EB Without Borders, DEBRA International and JBI evidence summaries from the Wound Healing and Management Collaborative.[66]

B.3 Neonatal wound care in epidermolysis bullosa

R11 ↑↑ Implement neonatal EB wound care strategies.

Refer to the skin and wound care in EB guidelines[2] for EB subtype dressing choices and care techniques. Neonates benefit from EB subtype-specific wound care performed with preventative handling techniques, to reduce blistering and improve wound healing.[1–3,6,9,10,23,37–45,48,54,58]

- It is strongly advised to use silicone medical adhesive removers (SMARs) to remove any tape or wound care product adhered to the skin for patients with EB, particularly neonates where it is unknown how they will react to the products.[1–3,6,9,14,23,31,32,36,38,40,45,55,65] Alternatively, dressings can be soaked loose in the bath (method outlined in R29).
- Use thin nonadherent contact and secondary layers to improve wound bed contact over small neonatal bony prominences.[1–3,6,9,14,23,31,36,39,43,45,51]
- Warm cleaning solution may reduce pain while cleaning.[1,41,45]

- Neonatal EB wound debridement is rare. Ineffective or delayed EB treatment may increase the need for wound debridement. Follow debridement recommendations in the EB wound care and EB pain management CPGs.[2,36,41,67]

R12 ↑↑ Use preventative care to reduce friction during neonatal wound care.

Encourage parents to prepare wound care products, participate in wound care and hold and soothe their neonate during procedures. Preparation of wound care products, including precut templates, warmed cleaning solution and adequate pain relief, prior to commencing wound care can support procedural care.

- Attend to wound care one limb at a time keeping the neonate wrapped throughout to reduce blistering caused by friction related to movement.[1,2,9,14,31,43,45,48]
- Use gauze bandages or tubular bandages to secure dressings instead of tape.[1–3,14,43]
- Apply emollient or foam pad to scalp wounds, avoid shaving hair.[1,26,27]
- Use emollients on wound care products to reduce shearing on removal.[1,2,6,14,23,42,43]

R13 ↑↑ Include parents in wound care and blister management.

- Gradually increase parents' participation in all wound care; consider their choices, their emotional distress and birth recovery.[1,9,19,27,43,45]
- Teach parents webspace bandaging on hand wounds for neonates with RDEB to slow pseudosyndactyly (GPP).
- Offer repetitive education to promote practice. Parents and carers may find mastering new skills difficult while coming to terms with the EB diagnosis.[1,9,14,20,26,27,37,43,45,51,68]
- Review blisters with each nappy change to respond quickly if new blisters appear throughout the day (GPP).
- Follow EB blister management strategies outlined in the wound care CPG.[2]

B.4 Wound infection and sepsis

Skin infection impairs wound healing and can lead to life-threatening systemic infection.[1,3,10,31,34,39,67] Complete and document a comprehensive wound assessment with each dressing change; include exudate, odour, pain, surrounding erythema and oedema. Use clinical judgement and microbiology results (when available) to determine whether topical antimicrobials or systemic antibiotics are required.[1–3,10,34,38,39,67]

Antimicrobial use

R14 ↑↑ Always include a dermatologist in planning wound management and rotation of neonatally appropriate wound irrigation products.

- Use antimicrobial soaks for nonsymptomatic positive wound cultures. Rotate antimicrobials if using long term.[1,2,10,31,34,67]
- Use antimicrobial soaks in combination with systemic antibiotics if wound shows signs of spreading or presence of systemic infection.[1,2,10,34,67]
- Consider treatment for candida in flexural, moist or macerated areas, including differentiating between EB-related oral plaques, feeding pain and oral candida (GPP).[37]
- Medical-grade honey can be used in the neonate; consider pain and surface area.[1,2,10,67]

Antimicrobials are not recommended as first-line wound treatment for neonates with EB

↓↓ Cleansing solutions such as diluted vinegar to treat pseudomonas or diluted bleach to treat *Staphylococcus* are reserved for older infants and children. Consider site-specific targeted wound soaks rather than full bath submersion to avoid allergy and skin irritation.[1,2,6,9,23,67]

↓↓ Silver-impregnated dressings are generally not recommended in neonates, due to potential toxicity from high ratio of wound size to total body surface. Limited research shows that application to a small area for a short duration may reduce the risk of systemic absorption and may reduce use of systemic antibiotics.[1] Use of silver should not be a first-line infection treatment for neonates.[67]

Antibiotic use

R15 ↑↑ Select antibiotics based on swab results and wound symptoms in neonates with EB to reduce risk of resistance.

- Antibiotics are not always required for asymptomatic wounds, as colonization of wounds is inevitable but does not always impact wound healing.[1,2,6,9,10,23,31,34,38,67]
- Limit topical antibiotics, and use in rotation to reduce microbial resistance.[1,2,6,10,34,38,67]
- Use enteral antibiotics for symptomatic wounds where no systemic symptoms are present.[1,6,31,23,34,67]
- Use intravenous antibiotics for any neonate with systemic symptoms to treat possible sepsis.[1,2,10,31]

Sepsis

R16 ↑ Monitor neonates closely for signs of sepsis.

- Neonatal sepsis should be addressed with urgency due to the risk of rapid deterioration for severe subtypes such as EBS generalized severe and JEB.[1,2,6,31,40]
- Monitoring wounds, pain, fluid and temperature can support early interventions for signs of sepsis.[1,2,6,31,40]
- Apply continuous monitoring to the systemically unwell neonate.[1,2,6,9,10,31,40]
- Always refer to an EB specialist for complex EB sepsis management to support subtype-specific care.
- Urgently address signs of infection in severe subtypes such as suspected JEB. Research suggests compromised thymic development in JEB with an absence of laminin 332.[1,69]

B.5 Pain management for neonates with epidermolysis bullosa

Neonates benefit from a range of pain relief options similarly to older patients with EB.[40]

R17 ↑↑ Always monitor pain using a validated neonatal pain scale.

- Use a validated pain scale such as the Neonatal Infant Pain Scale (NIPS)[70] or Face, Legs, Activity, Cry and Consolability (FLACC).[1,10,40,41,44,70]
- Changes to behaviour and pain scores should prompt investigation into infection and wound care strategies.[38]
- Complex wound care may require debriding, soaking or changes to wound product selection, increasing the wound care time and intensity, and should be factored into pain management.[6,41,67]

R18 ↑ Consider providing parents and carers with pain education and psychosocial support.

- Psychosocial support may assist parents and carers with potential difficulties seeing their neonate in pain.[1–3,9,12,13,18–20,25,41,71] Include palliative care and/or pain team referral if complex pain management or end-of-life support is required.
- Provide education for parents and carers to identify signs of pain and how to use pain management plans.[31,39,72]
- Parents and carers are more likely to engage, feel confident about daily care and participate in learning about EB when their neonate's pain is well managed.[3,31,35,71]
- Teaching parents pain management techniques in hospital and promoting use after discharge can minimize fear and procedural anxiety as the infant enters childhood.[1–3,6,9,14,19,39,41,43,45,51,71,72]

R19 ↑↑ Provide appropriate neonatal pain relief.

Neonates with EB can be born with large painful wounds, where some or all of the skin has peeled away from a limb. They will require appropriate pain relief.[1–3,6,38]

- Use sucrose, paracetamol/acetaminophen and/or oral morphine/oxycodone as first-line medications for pain relief.[1,3,23,41,44,51,72] Do so especially for degloved wounds.
- Refer to the acute pain team for complex and ongoing pain management if first-line medications are not sufficient.[3,40,41,43–45,51]
- It is recommended that medical monitoring and resuscitation equipment be available.[1,6,40,41]
- Seek country- or region-specific neonatal prescribing guidelines to ensure a safe and legal dosage, monitoring and weaning.[3,40,41,73,74]

Parents: implement nonmedication-based pain relief strategies, such as swaddling, patting, rocking, calm music and singing, as standalone relief or in conjunction with medication. Encourage participation in hospital management to support implementation after discharge.

Weaning: neonates often naturally wean from oral morphine[1,41] as their wounds heal, frequency is reduced and weight-prescribed doses remain unchanged after discharge. No literature was found reporting neonatal patients with EB experiencing opioid addiction in the first year of life.

Other analgesics: if other analgesics are used, consider monitoring and weaning guidelines. There was limited research into the use of clonidine, topical ropivacaine,[72] oral ketamine,[75] gabapentin[41] or fentanyl for neonatal pain management. Consider gestational age, prescribing policy and available neonatal resuscitation resources.

C:Feeding and nutrition of neonates with epidermolysis bullosa

Neonates with EB, especially those presenting with extensive wounds, have increased nutritional requirements to compensate for losses, and higher nutritional demands during wound healing and fighting infection.[57]

LRS: In some geographical areas direct breastfeeding may be the only option for neonates born with EB. Seek support through DEBRA International, EB Without Borders or EB-CLINET to access a tertiary hospital if additional nutritional support is needed.

If high-energy or high-protein formula is required but not available, the concentration of standard infant formula can be increased gradually to a maximum strength of 1 kcal mL^{-1}. If blood testing is not available, a (liquid or finely crushed tablet) general multivitamin and mineral may be beneficial.

C.1 Skin fragility and pain can alter nutritional requirements and feeding capabilities, necessitating dietitian referral and nutrition monitoring

Skin fragility, oral mucosal damage and pain in neonates with EB may impact on both their ability to feed orally and their subsequent growth.[6,7,23,24,35,46,57,76,77] Early knowledge of the EB subtype is vital as different subtypes of EB can pose varying nutritional challenges ranging from mild to life threatening. Filoni *et al.* found that 35% of neonates measured below the fifth growth centile at 1 month of age.[77]

R20 ↑↑ Refer to a neonatal dietitian to support intake and nutritional status if mucosal blistering and/or pain are present.

The negative pressure created in the mouth during feeding from breast- or bottle feeding can lead to painful mucosal damage and reduced volume intake.[7,57,76]

- Include nondrowsy pain relief prior to feeding; analgesic gel may be used.[1–3,6,9,10,23,35,37,41,43,46,49,54,55,57]
- Encourage breastfeeding and/or expressing breast milk.[57,76]
- Emollient should be applied to the breast/nipple or bottle teat (if bottle feeding) and to the neonates' cheek and lips to reduce friction while feeding.[2,6,9,10,23,24,26,36,38,39,42,45,50–52,54,55]
- Use a soft silicone teat or Haberman feeder to support bottle feeding.[1,2,6,9,10,14,24,31,35,43,49,50,52,57,76] Teats can be softened in warm water prior to use. Use emollient on the teat and the neonates' mouth to reduce friction.
- Refer the neonate with bilious vomiting or nasogastric tube (NGT) aspirates to neonatal surgical teams for suspected pyloric atresia.[7,56,60–64]

R21 ↑↑ Calculate nutritional requirements considering wound losses and feeding capacity.

The EB dietitian, the MDT and the parent or carer should work together to assess oral intake, feed tolerance, nutritional requirements, wound burdens and feeding strategies. Consider reduced nutritional monitoring, weight checks and interventions for neonates on end-of-life care pathways.

- Biweekly weight measurement can identify growth faltering and prompt early nutritional support. Head circumference and length measures can be taken at birth and once monthly thereafter in the first year.
- Calculate estimated calorie and protein requirements using bodyweight, extent of blisters on body surface, exudate losses, additional stress and activity factors, and the need for catch-up growth.[6,24,57,76,78]
- Monitor biochemical protein profile, and wound and blister burden.[31,57,76,78,79]
- Measure biweekly urinary sodium and potassium if energy provision is adequate and exudate losses are high, but weight continues to falter[1,10,24,31,76,80] to identify electrolyte depletion.
- Monitor iron, zinc and vitamins D and K for neonates with large degloving wounds.[1,24,38,57,76,81,82]
- Consider a prophylactic multivitamin and mineral supplement that contains vitamins A, C and D, iron and zinc.[57]
- Consider the impact of inflammation when interpreting serology markers.[57,80,82]
- If vitamin K is not given at birth, monitor vitamin K and PIVKA-II. Serum vitamin K is not a specific test and should not be measured in isolation.[81]
- If breast milk is not available, or supply does meet requirements, and growth continues to falter, high-energy and high-protein polymeric formula can be given as full or partial (top-up).[3,76,78]

C.2 Enteral feeding

Promote access to EB MDT care that includes a dietitian for comprehensive nutritional assessment.[6,23,31,35,38,46,76,83]

R22 ↑ Consider using enteral feeding tubes in neonates with EB not able to meet full nutritional requirements.

- Trial oral feeding, including soft preterm teats or Haberman feeder, prior to enteral feeding.[1,14,31,35,49,57,76,83,84]
- Consider the subtype of EB, the presence and severity of oral and oesophageal mucosal lining fragility, and tolerance of feeds to determine whether enteral feeding is required.[46,49,57,76]
- Consider early insertion of appropriate feeding tube to optimize nutrition in neonates with severe subtypes with poor feeding or tolerance.[1,57,76]

R23 ↑ Consider using experienced EB staff for NGT insertion and monitoring.

- An experienced staff member should insert the well-lubricated tube to reduce the risk of oesophageal damage.[1,9,10,31] Oesophageal trauma may cause acceleration of oesophageal stricturing in RDEB.[1,7,10]
- Use NGTs rather than orogastric tubes (OGTs) due to increased OGT movement against the oral mucosal lining.[31,57,76]
- Use NGT for severe subtypes when oral tolerance is normal, but intake is low.[31,85]
- Nasojejunal tubes can be utilized if medically indicated (severe vomiting or gastro-oesophageal reflux).[80]
- Secure NGTs with a low-adherent film contact layer, then full-adherent tape to secure NGT to film. Avoid tape contact with skin.[1,2,9,10,36,53]
- Remove emollient from skin to allow tube to be fixed in place.[1,9,10,53]
- A barrier film wipe may be used on the skin first (GPP).

If NGT securement is problematic, the lasso technique can be implemented.[57] Create a flexible bandage ring around the circumference of the neonate's head and under the nasal septum. The NGT is placed as per protocol then subsequently spiralled around the bandage ring to secure in place (GPP) (see photo in Appendix S5).

R24 ↑ Suggest maintaining dietetic input throughout enteral feeding use.

Enteral feeding strategies should be developed using an individualized approach.

- Maintain a degree of oral feeding (if safe) or oral stimulation such as non-nutritive suckling during enteral tube feeding.[57,85]
- Consider slow-pump bolus or continuous feeding if feed tolerance is poor.[57,85]
- Consider EB subtype, hospital capacity, parenting skills and community support after discharge to ensure enteral feeding is sustainable.[1,2,11,31,34,43–45,49]

C.3 Gastrostomy use and parenteral nutrition for neonates with severe epidermolysis bullosa

R25 ↑ Consider gastrostomy feeding to support nutritional requirements for neonates with severe EB with faltering growth.

Gastrostomy use in neonates with EB, while not common, is a treatment option. Nutritional support should continue to prioritize oral feeds, possible NGT placement or intravenous supplemental hydration, and MDT discussion before a gastrostomy is considered in the first 4 weeks of life. Gastrostomy management is outlined in the EB wound care CPG.[2]

- Early use of gastrostomy in neonates with severe EB can be lifesaving and can be reversed in childhood if nutritional status stabilizes.[24,57,63,86]
- Encourage parents and carers to document details and photos of the insertion process to support the child's understanding and ownership of the process when they are older (GPP).
- Provide parents and carers with early information to normalize enteral feeding for severe forms of EB.[9,19,31]
- Consider use if long-term nutritional supplementation is required and NGT is problematic.[2,7,35,86–88]
- Insertion should take place at an EB specialist hospital, or a tertiary children's hospital with intensive care unit support utilizing external EB hospital support for anaesthetic and surgical preventative support (GPP).
- Use modified two-port laparoscopic approach using the Seldinger technique with serial dilatation and tube insertion through a peel-away sheath (LAG technique).[89,90]

R26 ↑ Consider parenteral nutrition (PN) to support nutrition for neonates with EB.

While rare, use of PN can be considered for neonates with EB. Neonates with EB would be at higher risk of line sepsis and should be monitored very closely.

- Use short-term PN to optimize or stabilize nutrition prior to gastrostomy insertion or surgical management for pyloric atresia.[1,24,57,64]
- Trophic NGT or oral feeds should continue (if safe).[57]
- Consider short-term use while the umbilical line is accessible if neonate requires stabilizing.[1]
- Utilize an experienced EB nurse to support insertion and securement. Blistering and wounds around the insertion site should be monitored, and signs of infection or sepsis addressed quickly.[1,9,14,40,45]

D: Epidermolysis bullosa management for feeding, nappies/diapers, bathing and clothing

Parents, carers and local providers must be competent in daily care before the neonate is discharged home.[1,6,9,23,36–38,42,43,45]

D.1 EB neonatal management for feeding, nappies/diapers, bathing and clothing

Neonates with EB require preventative handling, emollient and padding during most activities to reduce the occurrence of new blisters.

Feeding

R27 ↑↑ Support feeding tolerance through pain management and friction reduction.

- Refer to R20 for emollient and teat support.
- Always use emollient on the pacifier, limit use if the oral cavity mucosa or lips are extensively blistered.[1,2,6,9,14,23,26,27,39,41,45,50,51]
- To burp or settle the neonate hold them upright on your chest and gently pat.[1,6,9,23,26,27,39,50]
- Give (paracetamol/acetaminophen) pain relief 20 min prior, or topical medication immediately prior to feed if appropriate.[1,2,6,9,10,23,31,37,41,43] Oral oxycodone/morphine may cause drowsiness and reduce appetite.

Nappies/diapers

R28 ↑↑ Use emollient and liners to reduce friction from nappy/diaper elastic on skin.

- Use emollient to clean the nappy/diaper area.[1,2,9,26,36,43,50,52]
- Use a hydrogel dressing on wounds in the nappy/diaper area.[1,9,43]
- Use a well-fitted nappy/diaper, and remove the outer elastic layer around the legs to reduce friction.[1,2,9,26,27,42,50]
- Use a nappy/diaper liner covered in emollient to reduce movement of nappy on skin.[1,2,7,16,20,23,42]
- Cloth or disposable nappies/diapers can be used in conjunction with the above recommendations.

Bathing

R29 ↑↑ Include a dermatologist and EB nurse and pain medication in bathing management.

- Neonates with birth trauma should initially be swaddled and cleaned using a dabbing technique rather than rubbing, and be cleaned limb by limb during wound care.
- When effective pain relief is established[1,2,6,9,14,20,23,26,31,41,45,48,51] move to submersion in a bath.
- Allow the neonate, wrapped in a thin cloth, to soak in a deep bath lined with a towel or blanket as padding, allowing dressings to come loose.[1,2,6,23,31,38,41,43–45]
- To reduce contamination of other wounds, consider limb-by-limb bathing during wound care if any wound infection is present (GPP).
- Adding 9 g of salt to each litre of water to create a saline bath can reduce pain.[41] Bath oil, emollient or soaps can make it difficult to hold the neonate securely.
- After the bath, leave wet dressings in place to protect the neonate from friction caused by movement until the new dressing can be applied (GPP).
- To dry, wrap in towels (with dressings still on) and allow the neonate to dry while holding; do not rub dry (GPP).
- Swaddle in a warm soft blanket, exposing one limb at a time, as new for old dressings are changed. Ensure the neonate is kept warm throughout.[1,6,23,38,43,45,51]

Clothing and protection

R30 ↑↑ Keep the neonate dressed and wrapped to reduce movement.

- Encourage parents and carers to dress their neonate with EB[1,26,27] to 'normalize' handling their baby.
- Keep the neonate dressed and wrapped to reduce movement.[1,3,6,10,14,20,23,26,27,41,45,47,50,51] Dress clothes inside out to reduce trauma from seams. Prevent buttons, press-studs, or snap-fasteners from touching the skin.[1,3,6,10,14,23,26,27,31,39,43,45,48,50] Smooth out creases from clothing or blankets to reduce skin markings (GPP).
- Apply a nonadherent wound dressing or barrier cream on bony prominences such as heels and elbows to reduce trauma.[1,3,10,23,26,27,31,43,45,48,50]

E: Discharge planning and epidermolysis bullosa education

An EB diagnosis at birth can be a traumatic event for a family and make learning and adjustment to life changes challenging.[19,20,29,71]

E.1 Parent education

R31 ↑ Offer EB education to prepare parents, carers and local providers for complex disease management at home.

Through previous recommendations we have highlighted the importance of including parental education. In addition to those, offer the following:

- Start education in hospital to prepare for discharge.[1,9,10,14,19,20,25,29,71] Include emergency care and health system navigation, among other support.[68]
- Offer repetitive education in different formats (spoken, practical demonstrations, written and weblinks) to support learning.[9,19,20,25,28,29,31,71]
- The EB specialist centre can provide education to community providers listed in the discharge plan prior to discharge.[10,14,19,20,26,42,43,45,51]

E.2 Discharge planning and organizational support

R32 ↑ Consider using a comprehensive discharge plan to support continuation of EB management after discharge.

- Establish organizational and health system support that includes an individualized care plan including specialist and local health provider contacts.[2,10,20,31,40,42,44,68]
- Hospital discharge can be considered when the neonate's general health condition is stable, and the parents and carers have demonstrated competence in neonatal EB management.[1,3,6,10,44]
- Encourage communication between parent and teams. Provide a single contact point for EB emergency management.[1,2,9,14,31,42]
- Parents and carers can benefit from country-specific DEBRA organizations, peer support and ongoing psychosocial support,[12,13,19,20,25,71] as well as DEBRA International patient-version CPGs.
- Consider cultural beliefs and barriers that may hinder ongoing management. Provide respectful, culturally sensitive education for key family members and siblings to dispel myths, and provide accurate information about inheritance, wound care and home management.[25]

E.3 Care coordination

All medical reviews, monitoring and handling are impacted by EB. Communication among the MDT is paramount.

R33 ↑ Consider using a care coordination plan with appointments, contacts and emergency management to support parents, carers and local providers.

- The EB specialist hospital can guide care and results within the community until practitioners have received EB education.[10,14,19,20,26,31,43,45,51]
- Open communication with the general practitioner and homecare team can benefit patients with EB.[10,19,20]
- Parents and carers benefit from ongoing access to their care coordinator or EB expert.[12,14,19,20,30]
- Local providers benefit from ongoing contact with the specialist hospital through different stages of the neonate's growth and development.[12,14,19,20,25,30]

3 Conclusion

All neonates with EB can benefit from specialist EB MDT care, either with face-to-face care, through a deployable EB nurse service, or through virtual support when transport is unavailable. EB education is essential for the family of the neonate and all healthcare providers involved in hospital or community care. Collaboration between hospital teams is essential to reduce handling and monitoring, which can cause or worsen trauma.

With early referral and access to EB-appropriate care, the neonate can be effectively monitored using care coordination between the specialist hospital, the local healthcare providers and community, and charity organizations. The inclusion of parents and carers in EB education, clinical skill development and care coordination planning is essential if appropriate management is to continue at home.

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Supporting Information

Additional Supporting Information may be found in the online version of this article at the publisher's website:

Appendix S1 Stakeholder involvement and peer review.
Table S1a Author panel members.
Table S1b Contributions to authorship.
Table S1c Contribution to external review.
Appendix S2 Methodology.
Table S2a Appraisal tool.
Table S2b Appraised article allocation per outcome.
Appendix S3 Strength of recommendations ratings.
Table S3a Strength of recommendations for interventions key.
Table S3b Level of evidence: GRADE certainty rating.
Table S3c Reference symbol key.
Appendix S4 Future research.
Appendix S5 Neonatal EB Hospital Implementation Tool.

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Data availability

All data are openly available.

Ethics statement

Ethics approval was not required for development of this guideline. This clinical practice guideline was developed

through a systematic literature review. Written consent from parents was obtained for all photos of neonates and children.

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