Neonatal EB Hospital Implementation Tool

Appendix 5

Appendix for the Neonatal Epidermolysis Bullosa clinical practice guidelines.

Neonatal EB Hospital Implementation Tool: an example neonatal hospital admission guideline.

How to use this hospital implementation tool:

1. Print relevant sections to make available for hospital staff, parents/carers, or community health professionals.
2. Consider availability of and access to local products and services.
3. Alter the document as provided to suit your hospital/regions health system and availability of services.

Topics covered in this example hospital guideline.

1. Admission, bedding, and safe handling
2. Psychosocial support for family
3. Pain relief
4. Anaesthetic and emergency management
5. Postnatal care
6. Preventative care for inpatient medical monitoring
7. Wound care and blisters
8. Infection and sepsis
9. Feeding and dietetic support
10. Daily cares
11. Discharge planning: parent competencies
12. Care coordination
13. Contact lists
14. Example signs for hospital room/cot

Procedure photos completed by
R Saad: EB Nurse Consultant, Sydney Children's Hospital, Australia, and CPG Lead.
J Duipmans: Lead EB Specialist Nurse, University Medical centre Groningen, The Netherlands.
K Plevey: Lead EB Clinical Specialist Nurse, Great Ormond Street NHS Foundation Trust, UK
N Yerlett: Dietetic Lead EB and Rare Dermatology, Great Ormond Street NHS Foundation Trust, UK

The document was reviewed by the neonatal EB CPG panel, and journal peer review process. All feedback was addressed and incorporated into this document.

Neonatal EB Hospital Implementation Tool

Recommendations for hospital management for neonate's with diagnosed/suspected epidermolysis bullosa (EB).

References for all recommendations can be found in EB Neonatal Clinical Practice Guidelines, 2024.

1.Admission, bedding, and safe handling

Referral to multidisciplinary care teams for inpatient care

Genetics	☐	Dietetics	☐
Epidermolysis Bullosa/ Dermatology	☐	Pain team	☐
Neonatologist/Paediatrician	☐	Social worker/Psychologist	☐
EB/Dermatology/Wound Nurse	☐	Palliative care	☐
Transport required to specialist EB hospital ?	☐	Contact Debra International or Debra without borders?	☐
Closest EB specialist hospital _____		Informed transport service of EB cares	☐

EB specialist contact Hospital _____ HCP_____

Early care and bedding

Do not nurse the neonate in an incubator, unless for medical reason such a prematurity.
Heat and humidity exacerbate blistering.

Overhead heaters- use only during procedures, blankets are preferred to maintain temperature.

Use a pressure-relieving, soft mattress, or extra soft padding on beds, such as incubator mattress.

Safe handling of a neonate with EB

Gloves- Use emollient or cotton/foam padding between gloved hands and neonates' skin.
To reduce drag of glove leading to increased friction.

Encouraged parents/carers to hold their newborn if they wish.

Lift on a pillow if baby is very fragile. Use flat palm and roll and lift technique – avoid scooping baby.

Place notices around the cot "Speak to nurse prior to handling patient" "Safety measures required for all handling".

Neonatal EB Hospital Implementation Tool

2. Psychological support for the family			
Explore options for EB outreach/deployable nurse program to maintain new family access with local support structures.			
Contact closest EB specialist hospital to discuss EB Nurse specialist support	☐	Local hospital support available?	☐
		Transfer to specialist hospital required?	☐
Establish emotional support structures for families if transfer away from local community is required for medical reasons.			☐
Provide parents/carers with information about Debra International or local Debra community support			☐
Refer for psychological or social work support through genetic testing period.			☐
Support parents with ethical and moral dilemmas linked to rare/fatal forms of EB such as junctional EB with pyloric atresia.			☐
Consider family			
Refer to genetics, palliative care, and psychosocial teams to discuss realistic expectations and prognosis.			☐
Support siblings through diagnosis process, teaching age-appropriate safe play and handling.			☐
Specialist to provide disease specific education opportunities for extended family (if requested/consented by parents).			☐

Neonatal EB Hospital Implementation Tool

3. Pain relief

Use a validated neonatal pain scale e.g., NIPS or FLACC for effective pain management.					
Seek country or region specific neonatal prescribing guidelines to ensure safe dosage, monitoring, and weaning.					
Offer oral feeds prior to opioid pain medication. A sleepy neonate may have reduced feeding leading to poor weight gain.					
Neonatal Pain relief options					
First line weight dosed medications for neonatal pain relief, especially when large degloving wounds are present. Use all individually and/or combined:					
Oral Sucrose	☐	Oral paracetamol/ acetaminophen	☐	Oral morphine/oxycodone	☐
Refer to acute pain team for complex and ongoing neonatal pain management if first line medications are not sufficient.					
Monitoring and resuscitation equipment available.	☐	EB preventative monitoring techniques implemented.	☐		
Considering parents/carers					
Refer parents to psychosocial support to understand difficulties handling their neonate in pain.					☐
Encourage parents/carers to learn, monitor and report signs of pain.	☐	Teach parents to administer medication.			☐
Teach parents wrapping, soothing, distraction, calming techniques.					☐

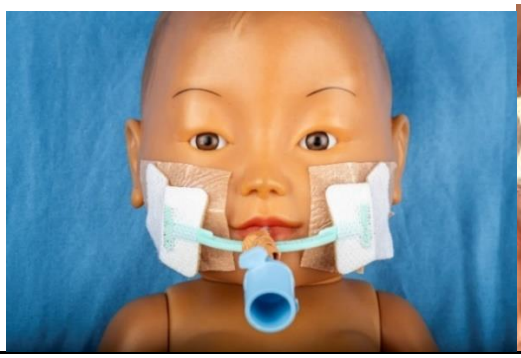
Neonatal EB Hospital Implementation Tool

4. Anaesthetic and emergency management

Follow steps to prompt discussion with emergency management team, anaesthetics, and medical team to prepare preventative care measures.

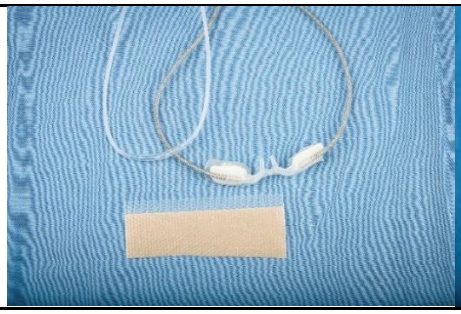
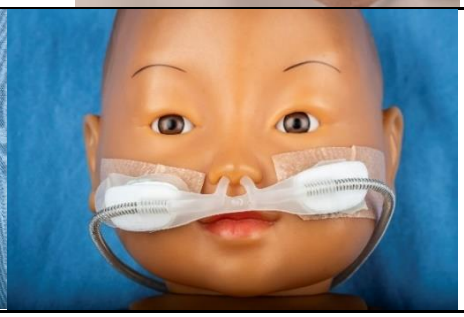
Potential intubation or airway support needed?	☐	Transfer to specialist EB hospital required?	☐
Anaesthetic consultation with EB staff to review available low adhesive tapes and discuss monitoring plan.			☐

Intubation- Lubricate laryngoscope and ET tube with emollient. Apply padding to mask and jaw line. Use low adherent tape on skin under neath standard securing tape.



Face masks -use soft inflated edge anaesthetic masks. Use water-based lubricant or foam padding between mask edge and neonate face. Remove emollient or lubricant from face prior to securing low adherent tape.

Nasal canula oxygen delivery- use low adhesive tape against skin before applying adhesive nasal canula.

An EB nurse escort should follow the patient through medical and surgical procedures.

Other Complications to monitor

Jaundice- Access EB specialist support if phototherapy treatment is required.

Pyloric atresia- requires transfer to an EB facility or surgical facility with neonatal intensive care. Surgical and anaesthetic procedures require full EB preventative cares.

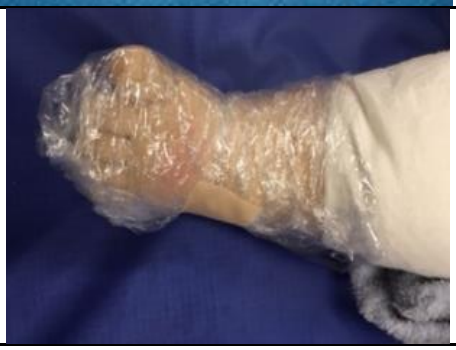
Neonatal EB Hospital Implementation Tool

5.Post-natal EB care

EB preventative care to implement in the first 48hrs for any suspected/diagnosis of neonatal EB (all subtypes).

Secure the umbilical cord with a ligature or a rubber cord ring rather than a cord clamp, to prevent trauma around the umbilicus.

Cover any birth trauma with a non-adherent dressing or cling film or as soon as possible.



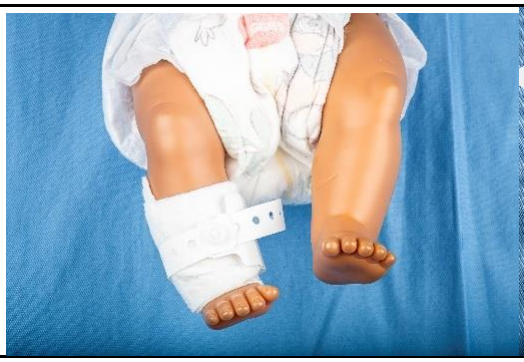
If stimulation is required after birth apply padding or emollient layer around neonate first.

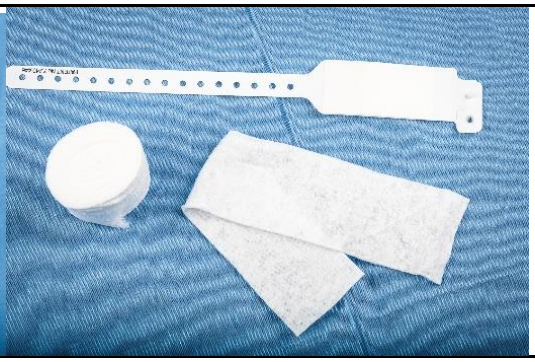
Apply emollient to suction catheters tips required for mucous or meconium removal.

Promote skin to skin contact with mother once wounds are covered and if neonate is well.

Weigh neonate wrapped to reduce movement, tare weighing scale to zero to include a soft cloth or towel.

Attach hospital ID band over clothing or socks, or pin to clothing.





Newborn screening test- use a venous sample on day 2-3 of life. Mark sample as 'venous' on test card.

Cluster any blood tests to reduce use of tourniquet and handling, consider collecting blood for genetic testing.

Give country specific vaccination required at birth. Use a low adherent dressing to cover site if required.

Use relevant sections during hospital admission or for EB education.
Refer to Neonatal CPG and referenced recommendation tables for supporting information.

Neonatal EB Hospital Implementation Tool

6.Preventative care for inpatient medical monitoring

Communicate preventative care techniques to hospital staff and in medical notes to support all monitoring.

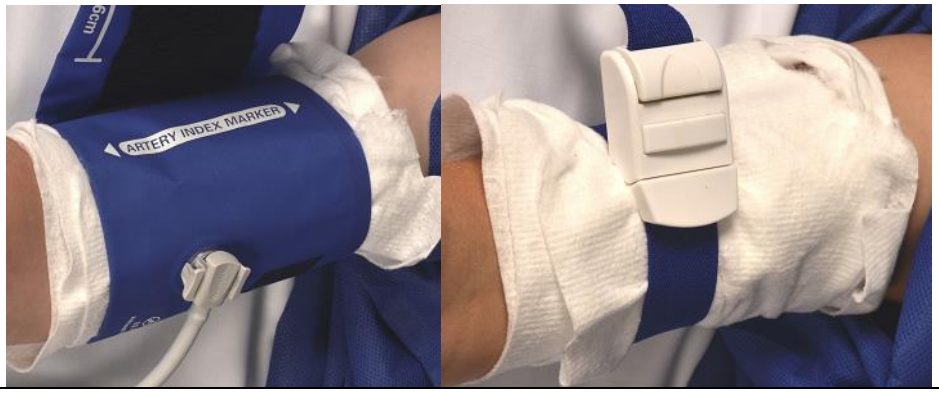
Observation and monitoring

Frequency: Individualise BP, Sa02, and temperature monitoring requirements/frequency.

Use infrared digital thermometer OR axillar temperature probes with emollient, lift arm to place and remove

Oxygen monitoring: Use soft cotton or silicone dressing between monitor and skin, cut hole for the sensor, ensure it is not covered.

Blood pressure- and tourniquets. If required use padding or clothing between cuff and skin.

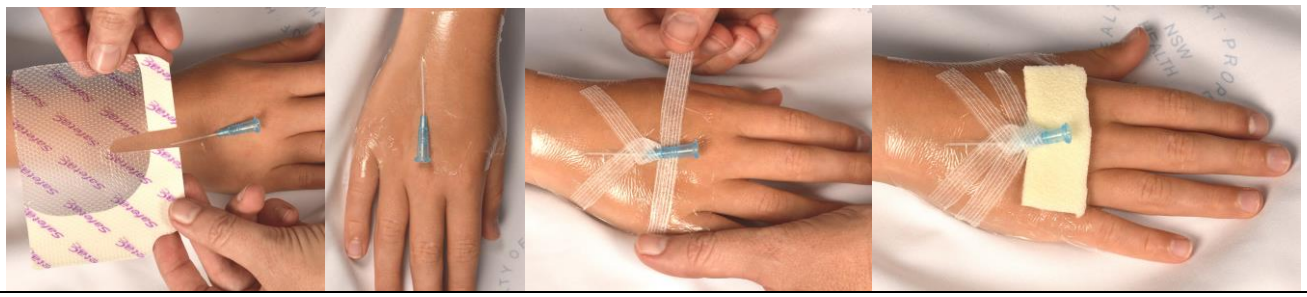


Fluid balance- Maintain strict fluid balance records. Bilious or repetitive vomiting, or abdominal distention may indicate need for ultrasound and exploration for pyloric atresia.

Urine collection- avoid adhesive bags, attempt clean catch collection or dipstick testing.

Canulation and Blood tests

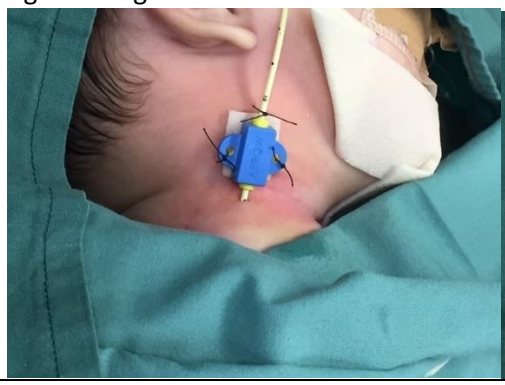
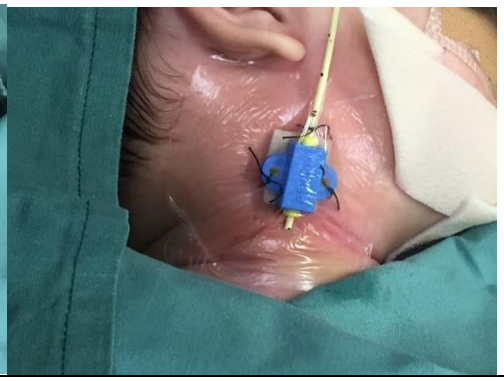
Intravenous canulation- requires the most experienced practitioner to reduce potential for multiple attempts. Use preventative padding for tourniquets, taping, and handling.



Use relevant sections during hospital admission or for EB education.
Refer to Neonatal CPG and referenced recommendation tables for supporting information.

Neonatal EB Hospital Implementation Tool

Central venous access-sterile foam placed below device prior to securing device with sutures. Secure with low adherent sterile film. Consider site with fewer blisters or wounds. Foam placed along jawline to reduce friction from gloves during handling.

Blood tests: Precautionary blood tests not recommended. Only test for clinical indications. To reduce unnecessary friction to skin and reduce blood volume drawn.

Blood tests- Priorities requested tests in cases of insufficient volumes.

Monitoring requiring preventative lubrication

Feeding tubes and catheters- lubricate well with a water-based lubricant.

Oral or cavity swabbing- lubricate with saline before performing swab.

Corneal abrasions use non medicated hydrating eye drops or gel to reduce friction in and around eyes.

Neonatal EB Hospital Implementation Tool

7. Wound care and blisters

Provide page to wound care staff and in medical notes to support EB preventative cares for all wound care.

Name of EB wound care specialist:	Contact details:

Current dressing plan (list products being used)

1. Contact layer: _____
2. Thin Foam layer:_____
3. Any Padding in use: _____

Securing method (type of tape or bandage: _____

Focus areas of care during neonatal wound care

- Contact layer to maintain contact with wound bed.
- Warm cleaning solution prior to use to reduce pain.
- Document length of wound care to inform pain relief requirements.
- Document wound size, presence of ooze, redness, or odour.
- Always refer to dermatologist if debridement required.
- Attempt enzymatic or autolytic debridement with wound care products.

Consider parent capacity during neonatal wound care

- Gradually increase parents' participation in all wound care.
- Consider their emotional distress and birth recovery.
- Parents are required to show competence in wound care prior to hospital discharge.

Neonatal EB blister management

Lance and drain neonatal EB blisters as soon as you can.	If roof remains on the blister, a dressing may not be required.
Include parents in blister management and education with a skilled EB nurse.	Promote Parent/carer competence in blister management prior to hospital discharge.

Reducing friction in neonatal wound care

Provide wound care one limb at a time, wrap neonate to secure other limbs.

Use tubular or crepe bandages or tape to secure dressings or devices to themselves rather than to skin.

Scalp blisters: Use lubrication/barrier cream or place foam pad if neonate has increased movement. Avoid shaving hair to reduce risk of long-term damage.

Neonatal EB Hospital Implementation Tool

8.Infection and Sepsis in neonatal EB
Provide page to ward staff and in medical notes to guide prescribing and infection management.
Preventative antibiotic coverage is generally not required for neonates with EB.
Antimicrobial Use
Use antimicrobial soaks in non-symptomatic wounds with positive wounds cultures. Rotate antimicrobials if using long term.
Medical grade honey can be used in neonates. Perform small patch of skin first as product may sting. Honey may be more readily available than antibiotics or standard antimicrobials in some regions.
Use antimicrobial soaks in combination with systemic antibiotics if wound shows signs of spreading or presence of systemic infection.
Antibiotic Use
Do not prescribe antibiotics for asymptomatic wounds in an asymptomatic neonate with a positive bacterial swab. Utilise antimicrobials first.
Limit topical antibiotics and use in rotation to avoid the development of microbial resistance.
Enteral antibiotics should be used for symptomatic wounds where no systemic symptoms are present.
Intravenous antibiotics should be used to treat the systemically unwell neonate to reduce the risk of sepsis.
Sepsis
Apply continuous monitoring to the systemically unwell neonate. Use preventative monitoring measures.
Rapid and fatal sepsis is not uncommon, monitor wounds, pain, appetite, fever, and lethargy to identify early deterioration.
Urgently address signs of sepsis in neonates with EB, particularly suspected severe subtypes such as JEB with a lack of laminin 332.

Neonatal EB Hospital Implementation Tool

9.Feeding and dietetic support

Provide page to dietitian to support EB preventative cares.

Feeding support for oral mucosal blistering and/or pain

Link neonate to EB specialist dietitian in hospital or via telehealth if not locally available.	☐	Encourage breastfeeding and/or expressing breastmilk.	☐
Provide pain relief if oral blistering/pain is present. Give 20minutes prior to feed.	☐	Pain relief: oral paracetamol and/or lidocaine gel on neonates' mouth. Sucralfate can be used to ease oral mucosal pain.	☐
If bottle feeding provide soft silicone teat or Habermann feeder.	☐	Provide emollient for bottle teat/mothers' breast/ neonates lips/chin.	☐
Provide ongoing feeding education to parents/carer.	☐	Provide parents education about signs of pain/ recording feeding volumes.	☐

Calculating nutritional requirements

Energy- Severe subtypes of EB will require up 150-200% of their nutritional needs.

Calculate calorie requirements using ideal body weight, ratio of blisters to body surface area, exudate losses, additional injury/activity factor and the need for catch up growth.

Protein- Replace protein loss through wounds and blisters. Monitor biochemical protein profile, wound and blister burden on the body surface when estimating between 1.5-4g protein/kg/day.

Sodium- Risk of sodium depletion that can result in suboptimal weight gain. Supplement sodium if energy provision appears adequate, exudate losses are high, but weight continues to falter.

Vitamin and minerals- Vitamin requirements in children with EB can be 150-200% of RDI.
For neonates with large de-gloving wounds, monitor Iron, zinc, Vitamin K and Vitamin D levels. Give iron and zinc at different times of the day. Consider prophylactic multivitamin.

Monitor Vitamin K if the neonate did not receive standard post-partum therapy.

Inflammation- Interpret serology markers with caution, careful consideration of inflammation on certain biomarkers and nutritional status. Acute phase reactants are common with EB due to high levels of inflammation.

If growth is faltering, add a high energy/protein polymeric formula to expressed breastmilk or standard formula bottle feeds.

Formula is an adequate substitute of breast milk and can be readily fortified to meet high nutritional requirements.

Neonatal EB Hospital Implementation Tool

Feeding and dietetic support continued…			
Enteral feeding			
Encourage oral feeding with pain relief, soft teat, and emollient prior to NG insertion (not OG tube).			☐
Support oral stimulation to reduce risk of aversion.	☐	Experienced staff member to insert NG tube.	☐

To insert: Use a well lubricated NG tube. Options to secure tube:

1.Lasso technique

2. Standard technique: Low-adherent film as a contact layer, with a full adherent tape that secures the tube to the film

Consider EB subtype, hospital capacity, parenting skills, and community support post discharge to ensure enteral feeding is sustainable.
Gastrostomy Insertion
Gastrostomy feeding is very rare in the first 4 weeks of life in EB.
Consider optimising nutrition during neonatal period if gastrostomy care is being considered in infancy.
Provide early education directed at parents/carers to normalise any enteral feeding for severe forms of EB.
Gastrostomy placement should be undertaken by an experienced surgeon using the modified 2-port laparoscopic approach using the Seldinger technique with serial dilatation and tube insertion through a peel-away sheath (LAG technique).
Parental nutrition
Use to optimise nutrition post-surgery or prior to insertion of gastrostomy.
Line care, dressings and monitoring as advised by the EB nurse, with signs of infection of sepsis addressed rapidly.
Trophic NGT/oral feeds can continue (if safe).
PN can be considered while the umbilical line is accessible, and the neonate is being stabilised.

Neonatal EB Hospital Implementation Tool

10.Daily cares: Feeding, Nappies/diapers, bathing, and clothing for neonates with EB.

Provide page to parents, hospital, and community staff to outline preventative handling.

Feeding

Limit Pacifier/dummy use if blister/wounds Present in mouth	☐	Encourage breastfeeding/expressing breastmilk.	☐
Use emollient on the teat/bottle/breast/nipple as well as the neonate lips			☐
Use Gentle pat/tap to settling after a feed.	☐	Soak bottle teat in a bowl of warm water to soften before use.	☐
Offer paracetamol 20-30minutes prior to feeding for neonates with oral blistering or plaque.			☐
Use a soft silicone or cleft-palate style bottle teat for formula or expressed breastmilk (e.g. Habermann).			☐

Nappies/Diapers

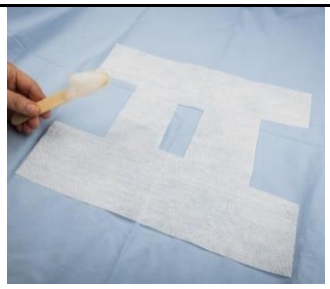
If skin has blistered use a hydrogel dressing over wounds or blisters.

Clean nappy/diaper area with emollient ointment in preference to water or commercial wipes.

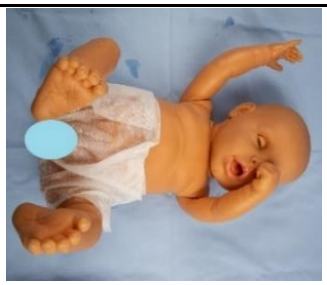
Use a well-fitted standard, disposable nappies/diaper. Trim off the inner elastic around legs to reduce friction.

Line nappy/diaper with a soft cloth liner coated with emollient or paraffin impregnated gauze.

Barrier creams and dressings may need to be changed every nappy/diaper change to keep wounds or blisters clean.

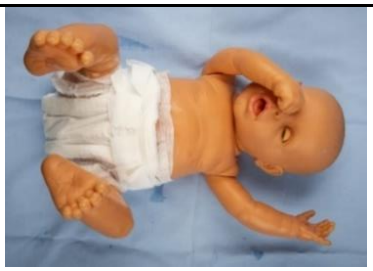


1.Cut out a soft nappy liner and cover with emollient.



2. Apply to neonate in similar fashion to nappy/diaper.

3.Trim the inner elastic of a nappy away, leaving the external elastic in place.



4.Place the nappy over the liner on the neonate.

Bathing

First Bath: Neonates with birth trauma should be swaddled and sponge cleaned limb by limb during wound care.

Bathing when pain relief has been prescribed: wrap the neonate in a thin cloth then soak in a deep padded bath, allowing dressings to come loose. Add salt to bath (9gms/1L) to reduce pain.

After bath wrap the neonate in towels (with loose dressings in place) and allow to dry while holding, do not rub dry.
When dry, swaddle the neonate in a warm soft blanket exposing one limb at a time as new for old dressings are attended. Ensuring the neonate is kept warm throughout.

Clothing
Keep the neonate dressed in padding or clothing and wrapped to reduce movement.
Wear clothing inside out to limit exposure of zippers, buttons, or seams to contact the skin.
Pad bony prominences such as heels and elbows as well as trauma-exposed sites to protect the fragile skin.

11.Discharge planning:

Use a hospital in the home approach to support parents to assume full care of wounds and medications in a supported setting prior to hospital discharge (if available).

EB care competencies checklist for parent

Checklists can be used by care coordinator or primary EB provider to support parents, community education, and effective transfer from hospital to community care.

Basic knowledge of their child's diagnosis of EB	☐	Preparing bath and dressing change	☐
Knowledge of medications, including pain relief prescribed and how to administer.	☐	Dressing changing	☐
Importance of safe storage of medications, dressings and topical treatments	☐	Bathing and skin cleansing	☐
Hand washing procedure	☐	Blister care	☐
Pain and itch assessment	☐	Disposing needles and soiled dressings	☐
Assessment for signs of infection	☐	Nappy care	☐
Knowledge of dressings and topical treatments, and how to use them.	☐	Handling of the baby	☐
Ordering repeat prescriptions	☐	Clothing/dressing the baby.	☐
Basic Life Support	☐	How to use EB care plan in the community	☐

Comments:

Neonatal EB Hospital Implementation Tool

12.Care coordination between hospital and community		
Items to coordinate	**Organisation/Contact name/details**	
Linking home nursing to the neonate.		☐
Sourcing EB wound care products.		☐
Linking local health care providers to the EB specialist hospital		☐
Outlining access and financial coverage available (government, non-government, EB organisations).		☐
Organising ongoing EB specialist appointments.		☐
Extended family and siblings education provided.		☐
Ongoing psychosocial support established for family.		☐
Providing EB education and support for local health care providers such as general practitioner, early childhood nurses, and local paediatrician.	Date of education sessions booked:	
Provision of an emergency management plan including EB specific care and contacts.	Copy of plan sent to (names):	
Comments:		

Use relevant sections during hospital admission or for EB education.
Refer to Neonatal CPG and referenced recommendation tables for supporting information.

13.Contact details for neonatal EB care

Complete to individualise contact list for patient

Role	Name	Contact details	When to contact
Specialist hospital contacts			
EB care coordinator			-to link community specialists with EB specialist hospital providers -to make Specialist hospital appointments
General Paediatrician			-to review wounds of concern -to review growth and illness of concern
Genetics team			-for diagnosis follow up -for genetic counselling for family members
EB Dermatologist			-to review wounds of concern -to community with local providers -to guide clinical diagnosis/wound care
EB Paediatrician			-to support community care
EB Dietitian			-to order feeding supplies -to communicate weekly weight
Other			
Community health care professionals and support agencies			
General Practitioner/ Family doctor			-for community medical review -swab wounds of concern -monitor growth
Psychologist/ social worker			-for ongoing social support through diagnosis and discharge
Wound product supplier			-to order ongoing wound supplies
Community nursing			-to organise community wound care appointments -to review wounds of concern - monitor growth
Local Debra Organisation			-for emotional and financial support -to request clinical EB support for local providers if not available
Nutritional product supplier			-to access feeding supplies
Local Pharmacy			-to organise medication -to receive scripts from hospital
Other			

Neonatal EB Hospital Implementation Tool

14. Example signs for hospital room/cot	
Adapt all monitoring to protect fragile skin	Speak to nurse before handling this patient: fragile skin
Do not remove any tape or wound dressings without speaking to nurse	EB Nurse escort required for all screening and procedures: patients requires extensive wound care support

Neonatal EB Hospital Implementation Tool

Disclaimer

This document provides guidance to support clinical judgment of trained health professionals within a hospital environment, and communication between treating hospitals and EB specialist teams. This tool can be used in conjunction with the recommendation tables from the neonatal EB clinical practice guideline.

This document was created using recommendations identified in the Neonatal EB CPG as well as existing EB hospital guidelines from Great Ormond Street NHS Foundation Trust, UK, Sydney Children's Hospital, Australia, and The Center for Blistering Diseases, University Medical Centre Groningen, The Netherlands.

Product availability may vary internationally, do consult your local resources, and seek local advice as well.