



Epidermolysis Bullosa (EB): Care of the newborn with suspected EB

Epidermolysis Bullosa (EB) is a genetically determined (inherited) skin fragility disorder.

Usual care of a newborn must be modified in order to reduce trauma and resulting damage to the skin and mucosae where possible ([Denyer, 2009b](#)).

The DEBRA NZ Nursing Team deliver a National Nursing Service to advise and support the Inter-disciplinary team from the hospital to home.

These guidelines are intended to support health care professionals looking after the newborn with suspected EB and to improve the care and safety of this group of patients.

Please contact one of the DEBRA NZ Nursing Team below for support:

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Background

There are four major types of EB, each with different clinical outcomes. The common factor is an extreme fragility of the skin and mucous membranes ([Fine et al, 2014](#)). The type of EB is determined by analysis of a skin biopsy. Regardless of the type of EB, care of the affected newborn will be the same. Severely affected newborns often present with extensive wounds over their limbs resulting from inter uterine movements and compounded by trauma during delivery.

Heat, pressure and friction can lead to the development of new blister formation & shearing of the skin. Modifications to the management of care for these newborns will reduce the risk of avoidable additional trauma to the skin and mucous membranes.

Immediate care after birth

([Denyer et al 2017](#))

- Remove the newborn's cord clamp and replace with a ligature (**Rationale** - to reduce the risk of causing trauma to the surrounding skin).
- Nurse the newborn in a bassinet unless an incubator is required for medical reasons such as prematurity (**Rationale** - heat and humidity can exacerbate blistering).
- Handle the newborn with care – avoid all rubbing or friction of the skin (**Rationale** - friction and shearing forces result in blistering and skin stripping).
- Do not use adhesive tapes or name bands. Photographic identification could be used (**Rationale** - name bands can rub causing trauma and blistering).
- If available, use latex-free gloves (people with chronic skin conditions exposed to excessive use of latex are at risk of developing a latex allergy) & apply 50% liquid paraffin, 50% white soft paraffin mix to the fingertips before handling the infant (**Rationale** – emollient applied to gloves reduces the risk of drag and subsequent trauma to the skin).

General care

Handling

- Lift the newborn on a soft pad/blanket. Avoid sliding your hands under the baby. Use a roll and lift technique (**Rationale** - shearing forces result in skin stripping).
- Avoid bathing until inter-uterine and/or birth damage has healed (**Rationale** - it is difficult to prevent further trauma with the newborn moving in the bath and to safely hold the newborn without causing skin loss - [Stevens & Denyer, 2010](#)).
- A pressure relieving mattresses such as a Roho should be considered (**Rationale** – to help relieve pressure and prevent further blistering).

For removal of tape without damaging skin

- Use a Silicone Medical Adhesive Remover (SMAR) such as Niltac® (**Rationale** - removal of adhesive products can result in skin stripping).
- If SMAR is not available, cover with 50% liquid paraffin, 50% white soft paraffin mix, which will dissolve the adhesive and enable safe removal (Denyer, 2011).
- When removing the tape, roll the tape back on itself rather than lifting it up from the skin (**Rationale** - reduces risk of epidermal stripping).
- Do not attempt to remove tape whilst it is functional, e.g. securing an IV cannula. If a new cannula is required secure with a soft silicone tape (**Rationale** - tape will not damage skin whilst in situ; it is the removal of the tape that has the potential for harm).

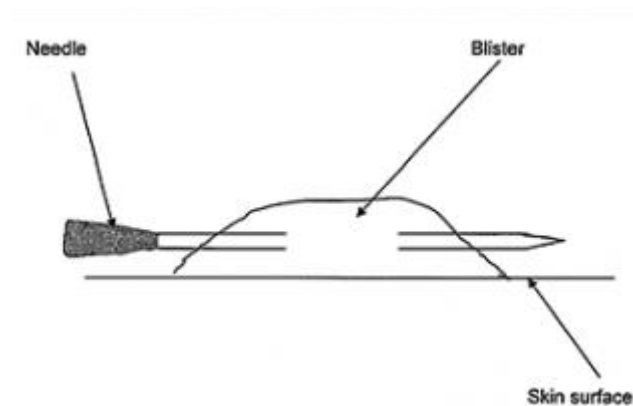
Nappy area care

- Cleanse with 50% liquid paraffin, 50% white soft paraffin mix and soft gauze (**Rationale** - cleansing with water can cause pain from contact with open lesions. Using emollients to cleanse the skin reduces trauma).
- Line the nappy with a soft cloth liner (**Rationale** - the edges of the nappy & elastic can rub leading to blisters and skin stripping).
- Apply a barrier cream such as Cavilon® or Bepanthen® (**Rationale** - to protect wounds and blisters from faecal contamination).
- Cover open lesions with Intrasite Conformable® dressings and change these at every nappy change (**Rationale** - to protect wounds & blisters and aid healing - Denyer, 2009a).

Blister care

- Blisters are not self-limiting and will enlarge if not lanced (Abercrombie et al, 2008).
- Prepare equipment – hypodermic needle, gauze and dressing if required.
- Wash hands. If not wearing gloves, please follow the 5 moments for hand hygiene (**Rationale** - to minimise the risk of cross infection for the newborn & staff - WHO, 2006).
- Position and prepare the newborn.
- Use a piece of soft gauze to gently compress the blister from the side to increase tension.
- Use an orange or blue hypodermic needle and pierce the blister at its lowest point (**Rationale** - lancing at the lowest point allows maximum drainage of the blister).
- Slide the needle through the blister to create an entry and exit point (**Rationale** - to allow the blister roof to lie flat after drainage).

- Withdraw the needle and gently press the blister with the gauze (**Rationale** - to ensure that all fluid is expelled).
- There is no need to dress the blister site if the roof has remained on the blister (**Rationale** - the underlying wound is protected by the roof of the blister).



When using needles there is a risk of needle stick injury. Therefore, avoid re-sheathing any used needles and dispose of any sharps in accordance with the hospital/community's waste management policy. In the event of a needle stick injury occurring, please seek immediate advice as per the hospital/community policy.

Wound care

Use a validated neonatal pain assessment tool to ensure adequate analgesia is given prior to wound care e.g. Sucrose & Morphine (opioid analgesia is usually required). Avoid the administration of rectal medication if possible as this can blister the anal margin (Herod et al, 2002).

- Prepare a clean trolley with clinical waste bag, hypodermic needles, sharps container, gauze, wound cleansing solution, dressings & tape cut into short lengths. It can be useful to cut limb dressings to shape using the supplied template below (**Rationale** - advanced preparation of dressings reduces time required to undertake dressing change and reduces the trauma for the newborn). The DEBRA NZ Nurse can help you obtain the specialised dressings required.
- Wash hands according to the WHO 5 moments for hand hygiene (**Rationale** - to minimise the risk of cross infection for the newborn and staff). Ensure lubricated gloves are worn.
- Expose one limb at a time (**Rationale** – to prevent movements of the newborn causing trauma to other exposed skin and also the wounds experiencing temperature loss that delays healing).
- If there is a need for more than one limb to be exposed at the same time e.g. for a medical review, then cling film can be used to protect the skin and keep air away from any wounds (**Rationale** – wounds exposed to air for extended periods leads to increased pain and delay in wound healing).
- Carefully remove soiled dressings using a Silicone Medical Adhesive Remover (SMAR) if the dressing is stuck (**Rationale** - to avoid the risk of skin stripping from the removal of adherent dressings). Dispose of all waste and sharps in accordance with the hospital/community's waste management policy.

- Lance any new blisters.
- Raw wounds: apply primary dressing of silicone mesh i.e. Mepitel/Mepitel One [Molnlycke], Siltex [Advancis from Bamford] or other silicone mesh dressing to raw areas as the skin contact dressing. Secure by taping it to itself (if required) and not directly on to the skin (**Rationale** - taping dressings to the skin risks skin stripping on removal of the tape. Larger dressing sizes offer protection from trauma of limb movements).
- Cover with a secondary dressing of Mepilex Lite/Mepilex Transfer/Mepilex [Molnlycke] (depending on amount of exudate). If using Mepilex Transfer a further layer of dressing is always required to allow the exudate to wick into. Secure with a cotton tubular bandage i.e. Tubifast [Molnlycke] or Comfifast [Bamford]. Soffban can be used under the tubular bandage for extra protection, or as the additional layer, if required. Change dressing when 'strike through' is observed - staining visible on the outside of the dressing (**Rationale** - 'strike through' indicates saturation of the dressing, loss of protection from microbes in the environment and a risk of maceration of the wound).
- Dress the newborn's fingers and toes individually if these are raw with one of the following recommended dressings: Mepitel/Mepitel One® (**Rationale** - raw digits will fuse together creating pseudo syndactyly - Denyer & Murrell, 2010).
- Take care that no tape comes into contact with the skin (**Rationale** - removal of adhesive products can result in skin stripping).
- Refer to suggested dressing template below for dressing limbs.

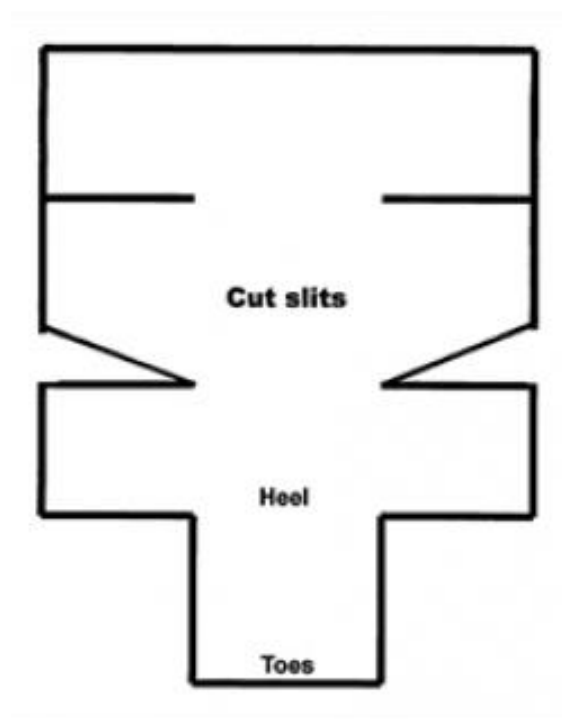


Figure 1. Template for foot and lower leg dressing (NB: this template can be used for upper limbs as well)

Feeding

- If the newborn is being breast fed, lubricate the nipple and newborn's lips with e.g. Lansinoh cream or 50% liquid paraffin, 50% white soft paraffin mix - 50:50 (**Rationale** – to avoid the baby's lips sticking to the breast).
- If the baby is being bottle fed and experiencing pain from oral blistering use a Haberman Feeder (**Rationale** - the valve in the teat of a Haberman Feeder allows a weak suck to generate a good volume of milk).
- Protect the newborn's lips with Lansinoh cream or 50% liquid paraffin, 50% white soft paraffin mix - 50:50 (**Rationale** - to reduce the risk of trauma from the bottle teat).
- Moisten teat with cooled boiled/sterile water prior to feeding or use teething gels if the mouth is very sore (**Rationale** - a dry teat can adhere to the oral mucosa and lead to skin stripping).
- Avoid using naso-gastric tube if possible (**Rationale** - mucosal blistering can result both from passing the tube and when the tube is in situ).
- If naso-gastric feeding is essential, use a tube suitable for long-term feeding and secure with soft silicone tape (**Rationale** - the long term tube is softer and will reduce the trauma for the infant).
- Pacifiers are not recommended (**Rationale** – movement of the pacifier will cause friction).

Clothing

Dress the baby in soft clothing turned inside out (**Rationale** - to reduce friction from internal seams, labelling & fastenings).

Cannulation

- IV fluids/antibiotics should be administered to the newborn only when necessary e.g. in the presence of sepsis or dehydration (**Rationale** - the majority of newborns with EB are able to feed orally. Prophylactic use of antibiotics is not recommended).
- Raised C Reactive Protein in the newborn with suspected EB is not necessarily an indication of infection in the presence of widespread inflammation (Mellerio, 2010).
- Clean the cannulation area by dabbing the skin with an antiseptic rather than rubbing (**Rationale** - rubbing may result in skin stripping).
- Do not use a tourniquet (**Rationale** - tourniquet use may cause blistering).
- Protect skin with soft gauze/soffban when squeezing the limb (**Rationale** - direct contact with the skin may cause blistering and skin stripping).

- Secure the cannula with soft silicone rather than an adhesive tape. Mepitel Film can be used as a protective landing zone for fixation tapes. If soft silicone tape or Mepitel film dressing are not available, use an adhesive tape but remove this following the guidelines above to reduce the risk of trauma.

Medical Devices

The use of medical devices on newborns with suspected EB causes an increased risk of friction & pressure injury. If these are essential, care should be taken when using devices such as a naso-gastric tube, oxygen, CPAP, IV/central lines, pulse oximeter, urinary catheter etc.

Continuity of care

Caring for a newborn with suspected EB is challenging and requires an interdisciplinary approach. Continuity of care is beneficial to the newborn, the family/whanau and the health care professionals.

It is recommended that wherever possible a primary nursing team is confirmed as soon as possible after birth and care is planned with support/involvement of a DEBRA NZ Nurse.

Family

It is important that the family have the chance to bond with their newborn. By implementing the recommended management of care as outlined above, the family can be involved in their baby's care, thus encouraging bonding, and the newborn can be safely handled with the support of nursing staff. It is also recommended that a family photo be taken at the earliest opportunity. This may be possible alongside any medical photography being taken.

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