

# PRACTICAL MANAGEMENT OF THE NEWBORN INFANT WITH SEVERE EPIDERMOLYSIS BULLOSA (EB)



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## INTRODUCTION

Epidermolysis bullosa (EB) comprises a group of genetically determined skin disorders. The common factor is the tendency for the skin and mucous membranes to break down in response to minimal everyday trauma and friction. Affected infants may present with extensive wounds resulting from inter-uterine movements and damage during the birthing process.

There are few specialised centres for EB and therefore care generally takes place in local hospitals where experience of this rare disorder is limited. We do not recommend transfer of a severely affected infant to a specialised EB centre because the journey and handling are hazardous for a severely affected infant.

## AIM

The study aims and objectives were to evaluate the ease of use of Guidelines for Immediate Care of the Infant with EB (the Guidelines) and the anticipated improvement in wound healing and reduction of traumatic injuries.

## METHOD

Newborn infants with skin and mucosal fragility who required modified handling, feeding and specialised wound care were selected for the study. Following telephone or email contact with the specialist centre for EB (Great Ormond Street Hospital (GOSH), London UK) the nursing and medical staff based at the referring hospital were directed to the Guidelines.

If recommended dressings were not available immediately suggestions were made to modify existing materials, e.g. by application of a greasy emollient to reduce the risk of traumatic removal.

One of the EB Specialist Nurses based at GOSH arranged to travel to the referring hospital within 48 hours of the referral to modify care to suit the individual infant, teach handling, feeding and dressing techniques and take a diagnostic skin biopsy.

Subsequent visits took place 1-2 times weekly depending on current workload. Factors considered included:

- Ease of application and removal of dressing materials.
- Healing
- Duration of dressing changes
- Pain control (Neonatal Infant Pain Scale)
- Minimal trauma from handling
- Adequate nutritional intake

## CONCLUSION

Using the Guidelines correctly will minimize trauma from handling and promote wound healing, pain control and general well-being. The Guidelines are now widely used in our practice.

## DISCUSSION

The high numbers of staff employed in a neonatal unit and the 12 hour shift pattern can result in inconsistency in allocation of staff to each baby, often a different nurse each day. Although the Guidelines are prominently displayed by the cot there is not always compliance with them. Parents are fundamentally important in ensuring correct care is given but this places an additional burden on them during this stressful time.

### Baby with fragile skin - handle with care!

- No shearing forces or friction!
- Remove cord clamp and replace with a ligature to avoid trauma to surrounding skin
- Nurse in cot/bassinette unless incubator required for medical reasons such as prematurity
- No adhesive products or name-bands (use photographic ID for consent and medication)
- If policy dictates wearing gloves then apply greasy ointment or lubricant in aerosol form to the fingertips to prevent friction with the skin

## Immediate care of newborn infant with epidermolysis bullosa

If recommended products are not available discuss with EB nurses for advice on adaptation of alternatives. These suggestions are for immediate care and will be adapted by the EB nurses during their first and subsequent visits.

### Handling

Lift on soft pad. Avoid sliding your hands under the baby as shearing forces cause damage. Use a "roll and lift technique" - The infant is gently rolled onto their side, the carers' hands placed behind the baby's head and bottom, the infant rolled back onto the carers' hands and lifted.

### Nappy area

- Cleanse with 50% liquid paraffin, 50% white soft paraffin mix or emollient spray and soft gauze
- Line nappy with soft liner to prevent elastic from rubbing.
- Apply an emollient / barrier cream
- Cover open lesions with hydrogel impregnated gauze and change at every nappy change

### Feeding

- Use a Special Needs Feeder if mouth is sore. Protect lips with petroleum jelly.
- Moisten teat with cooled boiled/sterile water prior to feeding to avoid sticking, or use teething gels if mouth is very sore.
- Avoid naso-gastric tube if possible. If naso-gastric feeding essential, use tube suitable for long-term feeding and secure with soft silicone tape.

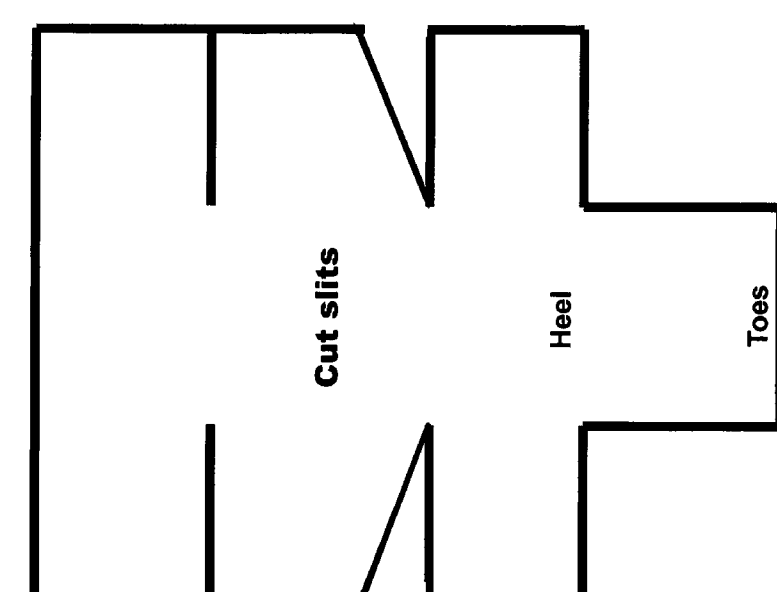


### For removal of tape without skin stripping

Use a Silicone Medical Adhesive Remover (SMAR). If SMAR not available, cover with 50% liquid / 50% white soft paraffin, which will help to dissolve the adhesive and enable safe removal.

If adhesive tape has been used in error but it is holding in an important cannula then do not remove it until the cannula can be removed.

### Dressing tips



Example of a template for the foot and leg



Digits wrapped with hydrofiber and soft silicone, hand wrapped with PMD



Change dressings before they become too wet to prevent hypothermia.



Change PMD when exudate is visible from the top of the dressing.



Remove cord clamp. to avoid damage



Cut through tape before removal as pulling can cause blistering.

### Wound healing



All infants achieved good healing using PMD which was easy to apply and remove. Images show a newborn with Herlitz Junction EB. The first image is prior to application of PMD, the second shows the improvement two days later.



### Clothing

Dress in soft, front fastening baby suit over dressings and nappy. Turn baby suit inside out to avoid damage from seams and labels.

### How to cannulate\*

- Do not rub area when cleaning as blisters or skin loss will result
- Do not use a tourniquet or stretch the skin
- Protect skin with soft gauze if assistant needed to squeeze the limb
- Secure cannula with a Soft Silicone Tape

*\*(IV fluids / antibiotics only necessary in the presence of sepsis or dehydration) Raised CRP level in a baby with EB is not necessarily an indication of infection in the presence of wide-spread inflammation*

### Analgesia

Regular analgesia is required with additional doses prior to dressing changes. A combination of paracetamol and oral morphine is effective. 24% sucrose solution is helpful in reducing procedural pain in combination with pharmacological management. Feeding the infant during dressing changes has a calming effect and is encouraged.

### Management of blisters

Blisters are not self-limiting and will enlarge if not lanced.

- 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
- Slide the needle through the blister to create an entry and exit point
- Withdraw the needle and gently press the blister with the gauze to expel the fluid
- It is not necessary to dress the blister site if the roof has remained on the blister



### Wound care

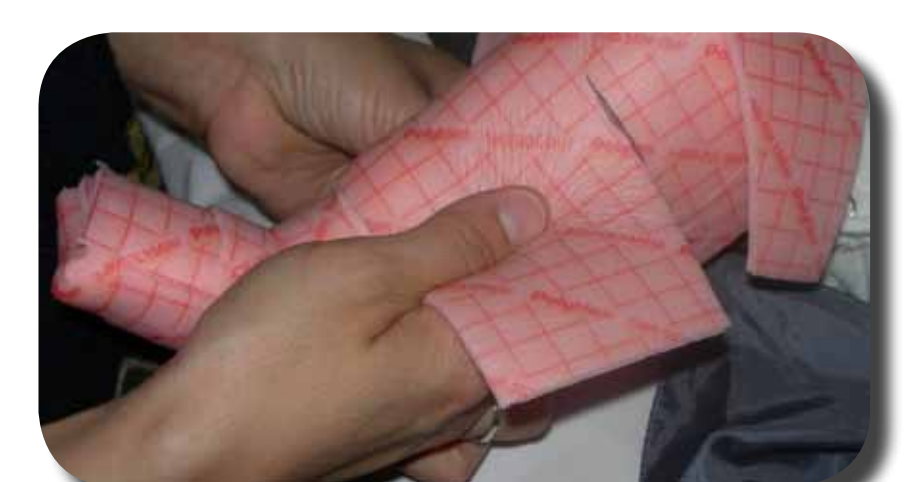
*\*Ensure adequate analgesia given prior to wound care*

- Prepare a clean trolley with clinical waste bag, hypodermic needles, all dressings (cut to shape) and tape cut into short lengths.
- Carefully remove soiled dressings using the medical adhesive removers or greasy emollient if stuck.
- Lance any new blisters.
- Raw wounds: Apply polymeric membrane dressings\* (PMDs).
- Further secure dressing with wrap-around bandage and or tubular bandage.
- Change dressing when "strike through" observed.
- Dress fingers and toes individually if raw to avoid digital fusion- use lipidocolloid / hydrofiber/ one-sided soft silicone dressings.
- Secure dressing by overlapping and taping to itself. Take care that no tape comes into contact with the skin.
- Avoid bathing until inter-uterine and birth damage have healed.

### How to apply PMD to the newborn



Overlap PMD and secure to itself



Cut slits over the joints to allow movement.



Secure dressing tightly to itself



Secure with tubular bandages.