Local Guideline

Epidermolysis Bullosa (EB) management in NICU

Sites where Local Guideline applies: Neonatal Intensive care Unit JHCH.

Target audience: All clinical staff in NICU, who provide care to neonates.

Description: This guideline provides information for staff in the management of Epidermolysis Bullosa whilst in NICU.

This Local Guideline applies to:
1. Adults: No
2. Children up to 16 years: No
3. Neonates – less than 29 days: Yes.

Keywords: Blisters, dressings, EB (Epidermolysis Bullosa), Herlitz variant, skin, wound, JHCH, NICU

Replaces Existing Local Guideline and Procedure: No

Registration Number(s) and/or name and of Superseded Documents: N/A

Related Legislation, Australian Standards, NSW Health Policy Directive, NSQHS Standard/EQuIP Criterion and/or other, HNE Health Documents, Professional Guidelines, Codes of Practice or Ethics:

Prerequisites (if required): N/A

Local Guideline Note: This document reflects what is currently regarded as safe and appropriate practice. The guideline section does not replace the need for the application of clinical judgment in respect to each individual patient but the procedure/s requires mandatory compliance. If staff believes that the procedure/s should not apply in a particular clinical situation they must seek advice from their unit manager/delegate and document the variance in the patient’s health record.

Position responsible for the Local Guideline and authorised by: Dr Paul Craven, Director of Newborn Services JHCH

Contact Person: Jennifer Ormsby Guideline Development NICU CNS NICU JHCH
Contact Details: Phone: 02 4985 5304
Email: Jennifer.Ormsby@hnehealth.nsw.gov.au

Date authorised: 06/10/2015

This Local Guideline contains advice on therapeutics: N/A

Date of Issue: 06/10/2015

Review due date: 06/10/2018
Note: Over time links in this document may cease working. Where this occurs please source the document in the PPG Directory at: http://ppg.hne.health.nsw.gov.au/

**RISK STATEMENT**

This local guideline has been developed to provide guidance to clinical staff in NICU to assist in assessment and management of Epidermolysis Bullosa (EB) in the neonate. It ensures that the risks of harm to the infants whilst caring for an infant being assessed and managed for EB are identified and managed.

Any unplanned event resulting in, or with the potential for injury, damage or other loss to infants/staff/family as a result of this management must be reported through the Incident Information management System and managed in accordance with the Ministry of Health Policy Directive: Incident management PD2007_061. This would include unintended injury that results in disability, death or prolonged hospital stay.

**RISK CATEGORY**: Clinical Care & Patient Safety

**OUTCOMES**

<table>
<thead>
<tr>
<th>Number</th>
<th>Description</th>
</tr>
</thead>
<tbody>
<tr>
<td>1</td>
<td>An infant with suspected EB is identified so management can occur to avoid further damage to the skin</td>
</tr>
<tr>
<td>2</td>
<td>Initial blister and wound care management care will be established</td>
</tr>
<tr>
<td>3</td>
<td>A selection of dressing products will be used in order to achieve optimal wound care management for each EB patient in the NICU-50% liquid paraffin and 50% soft paraffin will be ordered from pharmacy as soon as the baby presents</td>
</tr>
<tr>
<td>4</td>
<td>Referral to specialist dermatologist and multi-disciplinary team (such as wound care nurse, physiotherapist, occupational therapist, dietician and social worker) will occur</td>
</tr>
</tbody>
</table>

**ABBREVIATIONS & GLOSSARY**

<table>
<thead>
<tr>
<th>Abbreviation/Word</th>
<th>Definition</th>
</tr>
</thead>
<tbody>
<tr>
<td>DEB</td>
<td>Dystrophic Epidermolysis Bullosa</td>
</tr>
<tr>
<td>EB</td>
<td>Epidermolysis Bullosa</td>
</tr>
<tr>
<td>EBS</td>
<td>Epidermolysis Bullosa Simplex</td>
</tr>
<tr>
<td>GI</td>
<td>Gastrointestinal tract</td>
</tr>
<tr>
<td>ID</td>
<td>Identification</td>
</tr>
<tr>
<td>JEB/ JEB-H</td>
<td>Junctional Epidermolysis Bullosa / Herlitz variant</td>
</tr>
<tr>
<td>JHCH NICU</td>
<td>John Hunter Children’s Hospital / Neonatal Intensive care Unit</td>
</tr>
</tbody>
</table>
Epidermolysis Bullosa (EB) management in NICU JHCH_NICU_17.01

Epidermolysis Bullosa (EB) management in NICU- One Page Summary and Checklist

(Ctrl+Click on Coloured words to jump to that section)

Rationale

Background

Simplex
Junctional
Dystrophic

Newborn care

Wound care

References

GUIDELINE

This Guideline does not replace the need for the application of clinical judgment in respect to each individual patient.

Rationale

This guideline aims to support the JHCH NICU Nursing and Medical team in the initial management of a baby born with EB or suspected EB. Continued management will be in consultation with a multidisciplinary team.

Background

A newborn baby may be born with the rare genetic disease Epidermolysis Bullosa (EB). EB is characterized by extremely fragile skin and mucosae, resulting in chronic wounds and blisters. Blisters are usually present at birth or appear during the neonatal period. Secondary infection is the primary complication. There is currently no cure.

There are three broad categories of EB: Simplex, Junctional and Dystrophic. There are several subtypes within these categories which are both genetically and clinically different. Simplex: (EBS)

- The most superficial type
- Blisters arise intra epidermally
- May be localized or generalized
- Transmission is autosomal dominant
- Most infants with EBS experience recurrent blistering
- Relatively normal life span

Simplex EB  http://emedicine.medscape.com/article/909549-overview
The most severe form of EBS is Dowling Meara. There may be blisters in the mouth and finger and toenails may be thickened or missing. It can confuse clinicians in the way it presents, and infants who are severely affected are very challenging to manage and will require a multidisciplinary team approach.

Junctional: (JEB)
- Varies from mild to severe
- Blisters arise from the lamina lucida in the dermal-epidermal junction
- Least common type of EB
- May be localized or generalized
- All forms are autosomal recessive
- In all forms the GI tract is often affected
- Patients may die from strictures/obstructions/airway obstructions (blisters in the tracheolaryngeal area)

The most severe form of JEB is Herlitz variant (JEB-H), seen at birth with classic findings of atrophic scarring and dystrophy or absence of nails. Mucosal, tracheolaryngeal and GI tract blistering may cause gut and airway obstruction with mortality rates high secondary to malnutrition, sepsis and airway obstruction.

Dystrophic: (DEB)
- Autosomal recessive and dominant forms.
- Blister formation is below the dermal-epidermal junction in the superficial dermis
- May be localized or generalized
- Oesophagus may be involved
- Atrophic scarring and dystrophic nails and milia are present
- The oesophagus is often involved

Early identification is vital as there is tremendous difference in mortality and morbidity between the types. Autosomal recessive DEB has a much poorer outcome. Death during early childhood is common secondary to overwhelming sepsis.

Although EB is not often seen in the NICU the early management is crucial in the supportive care of and prevention of complications. The management of EB wounds requires frequent application of specialized dressings to promote healing, protect wounds and reduce infection risk.

Family education, support and follow up are essential in ensuring the best outcome possible for infants with EB and their families.

The National EB dressing scheme (An Australian Government initiative), provides a resource tool (compiled by Bright Sky Australia) for the wound care and dressing application, where a Nurse Consultant is available to provide education and support to families and health care professionals. The role is to deliver and develop educational
programs regarding optimal use of dressings for families of those diagnosed with EB and to support Health care professionals in the treatment and management of EB patients.

Contact the Nurse consultant on 1300 290 400 or eb@brightsky.com.au

A resource box is stored in NICU next to the schedule 8 drug cupboard. This box includes Bright sky information and resource tool, guidelines, dressing products and visual guides for dressing application.

_Disclaimer from BrightSky states that all information and material included in their publication is made available as a general guide and expert medical advice must always be sought._

**Newborn care**

<table>
<thead>
<tr>
<th>Action</th>
<th>Rationale</th>
</tr>
</thead>
<tbody>
<tr>
<td>At birth use commercial plastic cling film as a temporary dressing</td>
<td>Other materials may stick to damaged skin</td>
</tr>
<tr>
<td>Avoid attaching plastic ID tags directly to limb-infants to have an ID label with 3 identifiers attached to cot, IV arm board, and gastric tube. Document in notes/patient rounding form that no ID labels on limbs.</td>
<td>The rubbing will create blistering and trauma</td>
</tr>
<tr>
<td>Clamp cord with a ligature instead of a clamp</td>
<td>To avoid trauma to surrounding skin</td>
</tr>
<tr>
<td>Nurse the baby in an incubator only if medically necessary</td>
<td>Heat can exacerbate blistering</td>
</tr>
<tr>
<td>Ensure adequate pain relief is given prior to skin care dressings or interventions</td>
<td>Dressing changes are painful</td>
</tr>
<tr>
<td>Background pain relief may be required</td>
<td>Movement and handling can be painful</td>
</tr>
<tr>
<td>Washed and gloved hands and Non touch technique is accepted practice</td>
<td>To prevent skin infections</td>
</tr>
<tr>
<td>Wet gloved fingertips with water or apply 50% liquid paraffin and 50% soft paraffin to fingertips</td>
<td>To prevent fingers sticking to some dressings</td>
</tr>
<tr>
<td>Avoid bathing in the first few weeks of life</td>
<td>Allows time for healing of damage present at birth</td>
</tr>
<tr>
<td>Lance and drain all blisters using a sterile needle *refer to blister management appendix 1</td>
<td>The blisters will extend if left. The blister roof will facilitate healing</td>
</tr>
<tr>
<td>Apply primary contact layer dressings to raw areas and leave for 3-4 days. Then apply secondary dressings and secure. (Refer to Table of recommended dressings)</td>
<td>To protect the wound with a non-adherent dressing and to encourage healing</td>
</tr>
<tr>
<td>Dress fingers and toes individually if there is skin loss</td>
<td>To avoid digital fusion of open wounds</td>
</tr>
<tr>
<td>Avoid any adhesive dressings or tapes. Use non-adherent silicone tape for IV lines and NG silastic tubes (encourage oral feeding if possible)</td>
<td>Avoid trauma Due to trauma of feeding tube.</td>
</tr>
</tbody>
</table>
Cleanse nappy area with 50% liquid paraffin, 50% soft paraffin  | To avoid friction and reduce potential sting from water
Use disposable nappies with a soft nappy liner  | To prevent trauma for friction
Cover open lesions in the nappy area with a hydrogel impregnated dressing  | Soothing, comfortable and healing
Nurse in a cot on a soft pad where possible. Lift baby on this pad. Never lift a baby from under the arms, use a roll and lift technique  | Reduce friction and shearing forces
Dress baby in a loose jumpsuit turned inside out  | Naked babies with EB tend to cause damage to their skin when kicking or rubbing their arms across their skin. The inside out jumpsuit prevents friction against seams.
If not breast feeding, use a Pigeon cleft palate bottle and protect lips with 50% liquid paraffin and 50% soft paraffin  | To avoid the teat sticking to the lips

<table>
<thead>
<tr>
<th>Item</th>
<th>Use</th>
</tr>
</thead>
<tbody>
<tr>
<td>Mepitel/Urgotul</td>
<td>Primary dressing to raw areas. Use Mepitel for difficult to conform areas</td>
</tr>
<tr>
<td>Mepilex/Mepilex light</td>
<td>Secondary dressing to absorb exudate</td>
</tr>
<tr>
<td>Mepiform</td>
<td>For securing cannulae/gastric tubes</td>
</tr>
<tr>
<td>Mepitac</td>
<td>As above but less adhesive</td>
</tr>
<tr>
<td>Needles</td>
<td>Lancing and draining blisters</td>
</tr>
<tr>
<td>Cornflour</td>
<td>Dab onto small blisters that have been lanced but do not need dressing</td>
</tr>
<tr>
<td>Tubifast</td>
<td>To secure dressings</td>
</tr>
<tr>
<td>Webrill</td>
<td>To wrap over secondary dressing prior to tubifast to assist with securing (if necessary)</td>
</tr>
<tr>
<td>Intrasite conformable</td>
<td>Soothing comfortable dressing product for groin/nappy area. Effective dressing all over body for a HJEB diagnosis</td>
</tr>
<tr>
<td>Aquacel</td>
<td>For use if EBS-Dowling Mearra is suspected</td>
</tr>
<tr>
<td>Roll of cling wrap</td>
<td>Emergency dressing to protect wounds while other dressings and pain relief is prepared. (at birth)</td>
</tr>
<tr>
<td>Pigeon cleft palate feeder</td>
<td>Soft teat to reduce blistering if bottle feeding</td>
</tr>
</tbody>
</table>

The principle of wound management is specific to individual wounds and to the type of EB. The role of the dressing is to:
- provide a barrier
- help reduce infection
- provide optimal healing environment
- relieve pain
- limit friction and protect vulnerable skin
No single approach has proved totally effective and most patients need a variety of dressings to manage their wounds.

Prior to dressing selection clinical assessment of the wound should take place noting:
- wound edge
- wound appearance/fragility
- exudate (may demonstrate infection)
- odour (may demonstrate infection)
- Inflammation
- Wound pain

If dressings adhere to the wound moisten or soak the dressing prior to removal in order to minimise trauma to the wound bed.

Exudate management is crucial to avoid wetness and maceration. Alternatively if the wound is too dry scab formation will prevent wound healing, hydrogels may be required to donate moisture to the wound.

Wound Infection may be defined as the clinical syndrome of bacteria and other microbial organisms impairing wound healing. Topical antimicrobials and the possible need for systemic antibiotics should be considered.

References


3. DebRA UK. Epidermolysis Simplex (EBS) 2003

Author
Julie Gregory CNE NICU JHCH

Reviewed by
Viv Whitehead CNE NICU JHCH
Koert de Waal Neonatologist NICU JHCH
Jo Kent Biggs NE NICU JHCH

Approved by
NICU Management Executive Committee 11/09/2015
Clinical Quality & Patient care Committee 22/09/2015

Feedback
Any feedback on this document should be sent to the Contact Officer listed on the front page.