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Epidermolysis Bullosa and the National EB Dressings Scheme

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SAWMA Education Evening

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What is Epidermolysis Bullosa?

- EB occurs due to a genetic mutation resulting in protein defects in the dermal-epidermal junction.
- Leads to a lack of bonding between skin layers, causing skin fragility which is particularly sensitive to friction. Can also affect internal mucosa
- It presents in the form of blistering and chronic wounds
- It is estimated that EB affects 1 in 50,000 live births



Types of EB

4 main types with up to 20 sub-types within these classifications:

- **SIMPLEX (EBS)** – EBS LOCALIZED & DOWLING MEARA
- **JUNCTIONAL (JEB)** – HERLITZ & NON HERLITZ
- **DYSTROPHIC (DEB)** – DOMINANT DYSTROPHIC AND RECESSIVE DYSTROPHIC (dystrophic means *mutilating*)
- **KINDLER SYNDROME** - CONGENITAL ACRAL SKIN BLISTERING, PHOTSENSITIVITY AND PROGRESSIVE POIKILODERMA,



EB Simplex (EBS)

- Mechanical defect just *above* the basement membrane affecting the epidermis (keratin 5, 14 and plectin)
- One of the less severe groups of EB



Simplex (EBS)

- DOWLING MEARA is the most severe of EBS – can be very sick in neonatal period
- Blistering tends to improve over time, however blistering on hands & feet, can be disabling

EBS LOCALIZED is the most common of EBS

Thick walled blisters occur on hands and feet



Junctional EB (JEB)

- Mechanical defect *through* the basement membrane (collagen 17, laminin and integrin)
- **Herlitz Junctional EB** is the most severe and infants do not survive infancy.
 - Blistering in the larynx/malnutrition.
- **Non-Herlitz Junctional EB** presents as JEB in the newborn stage, but the patients survives early childhood.
 - Live with chronic wounds

Non-Herlitz Junctional EB

- Non healing areas remain a life long problem for such patients
- Chronic pain
- Corneal abrasions
- Urethral strictures

Herlitz Junctional EB

- Generally do not survive infancy
- Deterioration: Sepsis/respiratory failure/malnutrition

Care & Treatment:

- Adequate pain relief – midazolam, morphine, amitriptyline/gabapentin
- Nebulisers & dexamethasone to treat respiratory distress (morphine will help this too)
- Appropriate dressings
- Care at home with community nursing & palliative care support



Dystrophic EB

- Mechanical defect *under* the basement membrane affecting collagen VII
- Blister formation in this type of EB occurs within the dermis, thus resulting in scarring.



Dystrophic EB

- **Dominant Dystrophic EB (DDEB)** is the less severe form
- **Recessive dystrophic EB (RDEB):**
Pseudosyndactyly, SCC in later life



Complications of Severe EB

- Hands
- Nails
- Constipation
- Teeth
- Eyes
- Genito-urinary tract
- Kidneys
- Anaemia
- Heart
- Nutritional Problems
- Poor Growth



Blister Care and Prevention-

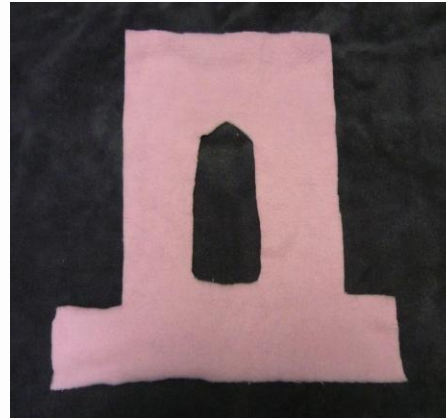
DO NOT:

- X** Rub the skin.
- X** Use adhesive dressings or tape.
- X** Affix plastic ID tags to skin



Newborn Care

- **Avoid bathing in the neonatal period**
(Denyer J, Stevens L 2010 "Bathing in EB – benefit over trauma?" Wounds UK)
- **Assess the need for pain relief**
(Goldschneider K, Lucky A, 2010 "Pain Management in EB" Dermatologic Clinics)
- **Lance and drain blisters - cornflour for EBS**



- Soft nappy liners
- 50/50 (Dermeze) for cleansing nappy area and protecting lips
- Apply appropriate dressings
- Dress the baby for protection – baby suit inside out
- Breast feed or Cleft Palate bottle feeder or Haberman. Avoid NGT/IV if possible
- **EMPOWER PARENTS**

Prior to the National Epidermolysis Bullosa Dressing Scheme (NEBDS)

- Inconsistency in access to dressings Australia-wide
- Dressings can cost up to \$4000 per month
- Relied on DebRA / Local charity groups

National EB Dressing Scheme

- Australian Government Initiative after 2 years of lobbying from DebRA Australia
- First national wound dressing scheme to support EB
- Launched 1st Jan 2010
- Value \$16.4m over four years
- Clinical Advisory Committee in place to advise on best practice
- Supports education for patients, carers and healthcare professionals
- BrightSky is appointed as the Scheme administrator

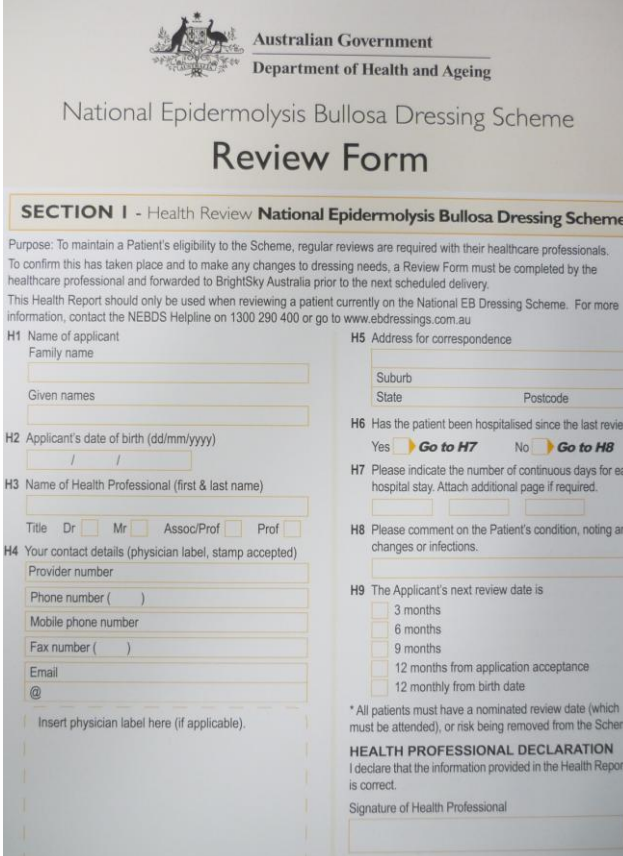
Application Process

- Meet eligibility requirements?
Complete application form & dressings prescription by Treating Healthcare Professional
- Biopsy or DNA result confirming clinical diagnosis of EB (provisional acceptance without this if Approved Healthcare Professional signs off on clinical diagnosis)
- Brightsky send to Canberra for approval
- Once approved, co-payment required (\$5.40 or \$33.30) : dressings sent out direct to patient

The image shows a screenshot of the 'National Epidermolysis Bullosa Dressing Scheme Application Form'. At the top, it features the Australian Government logo and the Department of Health and Ageing. The title is 'National Epidermolysis Bullosa Dressing Scheme Application Form'. Below the title is a 'Quick Eligibility Check' section with a warning: 'If any of the following points apply, you are NOT eligible for the National Epidermolysis Bullosa Dressing Scheme, so please do not complete this Application Form.' The eligibility criteria listed are: 'You are NOT an Australian citizen or permanent Australian resident', 'You do NOT have suspected or confirmed Epidermolysis Bullosa', and 'Your sub-type of Epidermolysis Bullosa is NOT covered by the Scheme*'. A note refers to the Eligibility Guidelines for more details or to call the National Epidermolysis Bullosa Dressing Scheme Helpline on 1300 290 400 or visit www.ebdressings.com.au. Below this is a box for returning the application to the NEBDS Administrator at BrightSky Australia, PO Box 6347, Silverwater DC NSW 1811. Another box explains that 'NEBDS' stands for National Epidermolysis Bullosa Dressing Scheme and that if the patient is already on NEBDS, only a NEBDS Review Form should be used. The form is divided into sections, with 'SECTION I - Applicant's Particulars' visible. It includes instructions for filling out the form (e.g., 'Please complete using a blue or black pen', 'Mark boxes like this [] or [] with a ✓', 'If the box looks like this [] follow the instructions after the arrow', 'If it has an instruction such as **Go to 12** skip to the question number shown. You do not need to answer the questions in between.') and numbered questions. Question 2 asks if the applicant is the applicant or completing the form on behalf of the applicant, with options to 'Go to 6' or 'Go to next question'. Question 3 asks for the relationship to the applicant, with options: 'Applicant's parent/legal guardian', 'Applicant's carer', 'Case manager', 'Healthcare professional', and 'Other' (with a 'Specify below' field). Question 4 asks if the applicant can nominate someone else to be the main contact and to receive correspondence on their behalf, with options to 'Go to 6' or 'Go to next question'. The page number '1 of 4' is visible at the bottom right of the form.

Review Form

- This will encourage regular skin checks, flagging infections and especially in RDEB – risk of SCC



Australian Government
Department of Health and Ageing

National Epidermolysis Bullosa Dressing Scheme
Review Form

SECTION I - Health Review National Epidermolysis Bullosa Dressing Scheme

Purpose: To maintain a Patient's eligibility to the Scheme, regular reviews are required with their healthcare professionals. To confirm this has taken place and to make any changes to dressing needs, a Review Form must be completed by the healthcare professional and forwarded to BrightSky Australia prior to the next scheduled delivery. This Health Report should only be used when reviewing a patient currently on the National EB Dressing Scheme. For more information, contact the NEBDS Helpline on 1300 290 400 or go to www.ebdressings.com.au

H1 Name of applicant
Family name
Given names

H2 Applicant's date of birth (dd/mm/yyyy)
/ /

H3 Name of Health Professional (first & last name)
Title Dr Mr Assoc/Prof Prof

H4 Your contact details (physician label, stamp accepted)
Provider number
Phone number ()
Mobile phone number
Fax number ()
Email
@
Insert physician label here (if applicable).

H5 Address for correspondence
Suburb
State Postcode

H6 Has the patient been hospitalised since the last review?
Yes **Go to H7** No **Go to H8**

H7 Please indicate the number of continuous days for each hospital stay. Attach additional page if required.
/ /

H8 Please comment on the Patient's condition, noting any changes or infections.

H9 The Applicant's next review date is
 3 months
 6 months
 9 months
 12 months from application acceptance
 12 months from birth date

* All patients must have a nominated review date (which must be attended), or risk being removed from the Scheme

HEALTH PROFESSIONAL DECLARATION
I declare that the information provided in the Health Report is correct.

Signature of Health Professional

CNC Role @ Brightsky

- Educate other professionals in dressing application
- Educate people living with EB
- Implement best practice
- Research/Studies on new products
- Develop Resources

The Aim of EB Dressings

- Provide Protection
- Relieve Pain
- Limit friction/slippage
- Promote optimal wound healing environment
- Reduce Infection

Key Points

- EB is very individual – eg cleansing:
- 10-15mls bleach in 10 litres of water
 - (Mellierio J, “Infection and Colonisation in EB” Dermatologic Clinics www.dermtheclinics.com) **or**
- 90g salt in 10 litres water **or**
- showers **or**
- dry dressings changes
- Wound assessment important
- Protect the wound bed
- Modern dressings designed to stay in place longer
- Product rotation beneficial



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National Epidermolysis Bullosa Dressing Scheme

Schedule of Dressings

Contact Layer Dressings – non silicone

- Vas ribbon Gauze – digit dressings
 - Adaptic – Petroleum emulsion coated
 - Urgotul / Urgotul Duo/ Urgotul Duo Border
-
- Primary and secondary dressings often used in EB due to fragility of the skin
 - Lifting secondary dressing allows inspection of the blisters, while protecting the wound bed

Urgotul®

- Non-adhesive lipido-colloid mesh, impregnated with hydrocolloid particles dispersed in a matrix of petroleum jelly
- Can be left in place a few days
- Promotes healing - JEB
- Atraumatic and pain free

Blanchet-Bardon (2005) Urgotul & skin lesions Journal of wound care Vol 14 No10

Primary Dressing



Urgotul Duo

- Lipido-colloid contact layer with a protective pad
- Low to moderate exuding wounds
- Healing in a moist environment
- Atraumatic
- Proliferation of fibroblasts
- Protection

Urgotul Duo Border

- Bordered dressing
- Suitable for DDEB or EBS
- Protection of bony prominences
- Can use silicone adhesive remover to remove (Welland /Niltac)

Contact layers - Silicone

- Mepitel®/Silflex –
DEB

Timmons et al (2009) Silflex soft
silicone wound contact dressing
Wounds UK Vol 5 No2

- Episil (semi-
permeable adherent
film dressing)

- Atraumatic soft
Silicone technology
- Soft & conformable
- Non adherent
- Allows passage of
exudate

Thick Foams & Exudate Transfer

- Mepilex Transfer
- Allevyn Heel
- Biatain non adhesive
- Polymem/ Polymem Max/Wic
- Urgocell
- Mepilex/ Mepilex Border/Sacrum/Heel

Mepilex Range

- Absorbent foam with gentle adhesion
- Reduces pain at dressing change
- Absorbs/Conforms
- Bordered for DDEB & EBS – waterproof
- Mepilex Transfer transfers exudate away for heavily exuding wound
- Conformable for difficult to dress areas

Traditional Foam Dressings

Allevyn Heel

- Moist wound environment
- Soft foam
- Anatomical shape, also useful for elbows
- Manages exudate
- Breathability to prevent unnecessary moisture retention

Caution with slippage and friction in EB

Biatain

- Moist wound environment
- Soft, flexible foam
- Easy to conform
- Highly absorbent
- Bevelled edges
- Protects

Caution with slippage and friction in EB – some adherence noted

PolyMem Quadrafoam[®]

- Cleanses, moisturises, absorbs & fills.
- A safe non-toxic cleanser (F68 surfactant)
- A moisturiser (glycerol) which prevents the dressing drying out and adhering to the wound bed,
- A starch copolymer to enhance the fluid handling properties.



Stevens, L (2010) The management of EB lesions with a polymeric membrane dressing. Poster presentation AWMA

Urgocell

- 3 layers : Urgotul, absorbent polyurethane pad & a semi-permeable polyurethane backing
- Exudate is retained in the dressing
- Non- adherent

Thin Foams - Silicones

- Advasorb Border
- Episil Absorbent
- Mepilex Border Lite
- Mepilex Lite

Highly Absorbent Dressings

- Dry Max
- Eclipse non-adherent
- Exu-Dry
- For increased exudate
- Not recommended as a primary dressing in EB
- HJEB – lay babys' head on (occipital can break down)

Low Absorbent Dressings (traditional)

- Combines
- Melolin
- Telfa/Telfa AMD

Hydrofibres & Hydrogels

- Aquacel – EBS , Dowling - Mearra
- Intrasite Conformable - JEB/nappy area
- Intrasite/Solosite Gel – debriding
- Prontosan Gel - debriding

Hydrofibre: Aquacel

- Soft dressing which converts to a gel when in contact with exudate
- Maintains moist wound healing environment
- Used in EBS – DM newborns
- Useful for blisters which keep re-filling
- Useful for moderately to heavily exuding wounds and sloughy areas



Hydrogel: Intrasite Conformable®

Gauze impregnated with Intrasite Gel: a hydrogel which de-sloughs, debrides and re-hydrates

Indications:

- Nappy area - cooling
- HJEB
- EBS - feet



Antimicrobials

- Honey dressing – Activon Tulle
- Acticoat
- Allevyn AG
- Aquacel AG
- Biatain AG
- Mepilex AG
- Polymem Silver
- Flaminal Forte & Hydro
- Prontosan

Silver Dressings

- CRT sparse – Mainly in vitro studies
- Some evidence to suggest systemic absorption of silver
- Caution with chronic wound use:
 - Not < 1 yr old
 - Not over 4 weeks continuous use

Honey based products



- **Activon Tulle®**

Hon, J (2005) Using honey to heal a chronic wound in a patient with Epidermolysis Bullosa BJN Vol 14 No 19



Actions:

- Debrides
- Reduces bacterial load
- Reduce malodour
- Osmotic pull can be painful

Prontosan

- Wound irrigation solution – Polyhexanide (PHMB)
- Reduces surface tension to dissolve fibrin coatings, exudate and biofilm
- Active against gram –ve and +ve, MRSA and pseudomonas
- High tissue tolerance

Kaehn, K (2009) An invitro model for comparing the efficiency of wound-rinsing solutions Journal of wound Care Vol 18 No 6

Horrocks, A (2006) Prontosan wound irrigation and gel: management of chronic wounds BJA Vol 15 No 22



Flaminal

- Alginate gel dressing (autolytic debridement)
- Antimicrobial enzyme system kills bacteria but does not affect dermal and epidermal cells
- Hydro & Forte



Tapes

- Hypafix
- Micropore
- Transpore – adheres to Polymem
- Yukiban
- Mepitac – safe for direct contact on skin
- Siltape – safe for direct contact on skin

Securing

- Padding – Ortho / Soffban / Webrill
- Bandages – crepe / retention
- Handybands – useful for digits
- Tubular Bandages – Surgifix/Tubifast/Tubigrip
(Preferable in paediatrics)
- Tubifast garments ? Difficult to apply

Ancillaries

- Lancets
- Needles
- Trio Niltac adhesive remover spray
- Welland Adhesive remover Spray
- Welland Barrier Film (WBF)

Silicone Medical Adhesive Removers and Barrier Film

- A non–sting preparation available as a spray or wipe
- Enables adhesive tapes and dressings to be used
- Harmless removal of adhered clothing & dressings
- Welland Barrier Film protects healthy skin from exudate



In Conclusion

- 45 patients now on the NEBDS
- More resources available by Jan 2011
- Working closely with DebRA Australia – training planned for 2 new DebRA nurses

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