

## 294502 Mepilex Transfer

Soft silicone exudate transfer dressing

### Product details

**Size :** 20cm x 50cm  
**Descriptive feature :** Exudate transfer, Foam, Non-border, Soft silicone  
**Sterile :** Sterile

### Images



### Delivered items

294502-03

**Sales released in:** Algeria, Australia, Austria, Bahrain, Belarus, Belgium, Bosnia and Herzegovina, Bulgaria, Canada, China, Croatia, Czechia, Denmark, Estonia, Faroe Islands, Finland, France, Germany, Greece, Hong Kong, Hungary, Iceland, Iran (Islamic Republic of), Ireland, Israel, Italy, Japan, Kazakhstan, Kuwait, Latvia, Lithuania, Luxembourg, Macedonia (the former Yugoslav Republic of), Malaysia, Moldova (the Republic of), Netherlands, New Zealand, Norway, Poland, Portugal, Romania, Russian Federation, Saudi Arabia, Serbia, Singapore, Slovakia, Slovenia, South Africa, Spain, Sweden, Switzerland, Taiwan (Province of China), Thailand, Turkey, Ukraine, United Arab Emirates, United Kingdom of Great Britain and Northern Ireland

**Country of origin:** Finland

**Shelf life:** 3 years

**Sterilization method:** EtO

**Packing information:** First packaging layer is a peel-open sterile barrier, plastic/plastic. Once opened the sterile barrier cannot be closed again. Second layer is a cardboard dispenser box. Third layer is a corrugated board transport box.

**Is suitable for Tray:** No

Packing level	Quantity	GS1 code	WxLxH (mm)	Vol (dm <sup>3</sup> )	Weight gross/net (kg)
Consumer pack	1	7332430003775			

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Packing level	Quantity	GS1 code	WxLxH (mm)	Vol (dm <sup>3</sup> )	Weight gross/net (kg)
Dispenser box	4	7323190024032			
Transport box	24	7323190024025			
Pallet	2016	7323190024018			

## Material

Natural rubber latex : No

## Product Composition Wound Contact Layers

Product Component	Composition
Transferring layer	Polyurethane foam
Wound contact layer	Silicone
Protective release liner	Polyethylene film

## Product Performance Wound Contact Layer Products

Characteristics	Test Method	Internal Test Method	Unit	Requirement	Product Performance
Free Swell Absorptive Capacity	EN 13726-1 part 3:2	T-1069	g/100 cm <sup>2</sup>	Not specified	N/A
Free Swell Absorptive Capacity	EN 13726-1 part 3:2	T-1069	g/g	Not specified	N/A
Conformability-Extensibility, MD	EN 13726-4	T-1086	N/cm	Not specified	N/A
Conformability-Extensibility, CD	EN 13726-4	T-1086	N/cm	Not specified	N/A
Conformability-Permanent Set, MD	EN 13726-4	T-1086	%	Not specified	N/A
Conformability-Permanent Set, CD	EN 13726-4	T-1086	%	Not specified	N/A

## Technical

## Dimension

Dimension text	Dimension value
Product	20 cm x 50 cm

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Dimension text	Dimension value
Product	8 in x 20 in

### Classifications

Regulation type(s)	MDD Class I/IIb	Locally Regulated	Unregulated
CE Certificate Number :	CE 01965		
Notified body medical devices/PPE :	BSI (2797)		
Intended use MDD :	Mepilex Transfer is designed for a wide range of exuding and difficult-to-dress wounds. Mepilex Transfer can also be used as a protective layer on non-exuding wounds and/or large areas of fragile skin. Mepilex Transfer can be used under compression.		
Sales released in :	Austria, Belgium, Bulgaria, Croatia, Czechia, Denmark, Estonia, Faroe Islands, Finland, France, Germany, Greece, Hungary, Iceland, Ireland, Italy, Latvia, Lithuania, Luxembourg, Netherlands, Norway, Poland, Portugal, Romania, Slovakia, Slovenia, Spain, Sweden, Switzerland, United Kingdom of Great Britain and Northern Ireland	Algeria, Australia, Bahrain, Canada, China, Israel, Japan, Kazakhstan, Kuwait, Macedonia (the former Yugoslav Republic of), Malaysia, Moldova (the Republic of), New Zealand, Russian Federation, Saudi Arabia, Serbia, Singapore, Taiwan (Province of China), Thailand, Turkey, Ukraine, United Arab Emirates	Belarus, Bosnia and Herzegovina, Hong Kong, Iran (Islamic Republic of), South Africa

**Applied standards :** The standards presented below is a selection of the most essential standards that are adhered to.

EN 1041, EN ISO 9001, EN ISO 13485, EN ISO 10993-1, EN ISO 10993-5, EN ISO 10993-7, EN ISO 11607-1, EN ISO 11607-2, EN ISO 15223-1, EN ISO 10993-11, EN ISO 10993-10, EN ISO 10993-18, ISO 14001

### Removable label

No

### GMDN Code (Global Medical Device Nomenclature)

46855 Wound - nonadherent dressing, permeable

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**284322 Mepilex EM**

Absorbent soft silicone dressing

## Product details

**Size :** 17.5cm x 17.5cm  
**Descriptive feature :** Foam, Non-border, Soft silicone, Thin  
**Sterile :** Sterile

## Images



## Delivered items

284322-01

**Sales released in:** Bosnia and Herzegovina, Bulgaria, Croatia, France, Greece, Hungary, Israel, Macedonia (the former Yugoslav Republic of), Martinique, Moldova (the Republic of), Pakistan, Poland, Portugal, Romania, Serbia, Slovakia, Slovenia, Spain, Turkey

**Country of origin:** Finland

**Shelf life:** 3 years

**Sterilization method:** EtO

**Packing information:** First packaging layer is a peel open sterile barrier, paper/plastic. Once opened the sterile barrier cannot be closed again. Second layer is a cardboard dispenser box. Third layer is a corrugated board transport box.

**Is suitable for Tray:** No

Packing level	Quantity	GS1 code	WxLxH (mm)	Vol (dm <sup>3</sup> )	Weight gross/net (kg)
Consumer pack	1	7332430666642			
Dispenser box	5	7323190126606	26x220x236		
Transport box	35	7323190126590	234x263x215	13.2	1.3 / 0.5

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Packing level	Quantity	GS1 code	WxLxH (mm)	Vol (dm <sup>3</sup> )	Weight gross/net (kg)
Pallet	4200	7323190126583	800x1200x1884		

## Material

**Animal tissues :** No  
**Natural rubber latex :** No  
**Medicinal substances :** No

## Product Composition Non-bordered Foam Products

Product Component	Composition
Backing material	Polyurethane film
Wound pad	Polyurethane foam
Wound contact layer	Silicone
Protective release liner	Polyethylene film

## Product Performance Non-bordered Foam Products

Characteristics	Test Method	Internal Test Method	Unit	Requirement	Product Performance
Free Swell Absorptive Capacity	EN 13726-1 part 3:2	T-1069	g/100 cm <sup>2</sup>	Not specified	N/A
Free Swell Absorptive Capacity	EN 13726-1 part 3:2	T-1069	g/g	Not specified	N/A
Fluid Handling Capacity	EN 13726-1 part 3:3	T-1068	g/10 cm <sup>2</sup> /24 h	Not specified	7.3
Absorbency	EN 13726-1 part 3:3	T-1068	g/10 cm <sup>2</sup> /24 h	Not specified	1.82
Moisture Vapour Transmission Rate (MVTR)	EN 13726-1 part 3:3	T-1068	g/10 cm <sup>2</sup> /24 h	Not specified	5.5
Moisture Vapour Transmission Rate (MVTR) of a wound dressing when in contact with water vapour	EN 13726-2 part 3:2	T-1070	g/m <sup>2</sup> /24 h	Not specified	2282

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Characteristics	Test Method	Internal Test Method	Unit	Requirement	Product Performance
Moisture Vapour Transmission Rate (MVTR) of a wound dressing when in contact with liquid	EN 13726-2 part 3:3	T-1075	g/m <sup>2</sup> /24 h	Not specified	4617
Waterproofness	EN 13726-3	T-1083	Pass/Fail	>500 mm H <sub>2</sub> O for 300 s	N/A
Conformability-Extensibility, MD	EN 13726-4	T-1086	N/cm	Not specified	0.6
Conformability-Extensibility, CD	EN 13726-4	T-1086	N/cm	Not specified	0.5
Conformability-Permanent Set, MD	EN 13726-4	T-1086	%	Not specified	0.3
Conformability-Permanent Set, CD	EN 13726-4	T-1086	%	Not specified	2.7
Resistance to microbial penetration - Wet	ISO 22610	T-1005	BI	6	N/A
Viral penetration	ASTM F 1671	N/A	Pass/Fail	29 out of 32 samples	N/A

#### Technical

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#### Dimension

Dimension text	Dimension value
Product	17.5 cm x 17.5 cm
Product	6.9 in x 6.9 in

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**Classifications**

Regulation type(s)	MDD Class IIb	Locally Regulated	Unregulated
MDD Classification Rule :	4		
CE Certificate Number :	CE 01965		
Notified body medical devices/PPE :	BSI (2797)		
Intended use MDD :	Mepilex Lite is designed for the management of a wide range of non to low exuding wounds, such as leg and foot ulcers, pressure ulcers, partial thickness burns, radiation skin reactions and Epidermolysis Bullosa. Mepilex Lite can also be used as protection of compromised and/or fragile skin.		
Sales released in :	Bulgaria, Croatia, France, Greece, Hungary, Martinique, Poland, Portugal, Romania, Slovakia, Slovenia, Spain	Bosnia and Herzegovina, Israel, Moldova (the Republic of), Pakistan, Serbia, Turkey	Macedonia (the former Yugoslav Republic of)

**Applied standards :** The standards presented below is a selection of the most essential standards that are adhered to.

EN 1041, EN ISO 9001, EN ISO 13485, EN ISO 10993-1, EN ISO 10993-5, EN ISO 10993-7, EN ISO 11607-1, EN ISO 11607-2, EN ISO 15223-1, EN ISO 10993-11, EN ISO 10993-10, EN ISO 10993-18, ISO 14001

**Removable label**

No

**GMDN Code (Global Medical Device Nomenclature)**

46854 Wound - nonadherent dressing, absorbent, sterile

**UNSPSC**

42311510 Foam dressings

**Commodity Code**

3005100000 Wadding, Gauze, Dressings, drapes singlepacked - adhesive articles, Sets mainly consisting thereof

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Epidermolysis  
Bullosa practical  
care guidelines

**Adult Surgical  
Procedures**



**DEBRA is the only charity supporting people living and working with EB (Epidermolysis Bullosa) – a rare genetic condition which causes the skin to blister and shear at the lightest friction, or even spontaneously.**

## **Our purpose**

We have a vision of a world where no one suffers from EB.

Until that day, we offer specialist care to those who need it.

We give support to people and families affected.

And we provide real hope for the future by funding pioneering research which will one day find a cure.

## **Our service**

We provide information, practical help and professional advice through our Nursing and Social Care teams.

In partnership with the NHS, DEBRA's Specialist Children's and Adults' Nursing teams work throughout the UK, providing individual specialist healthcare advice and support to both people with EB and their carers, both in the community and in specialist hospital centres in London, Birmingham and Scotland.

DEBRA's Social Care team works with individuals and families, providing information, advice, advocacy and support on issues such as benefits and finance, housing, education and employment, thereby empowering and enabling people with EB to make their own life choices.

Details for the Nursing and Social Care teams, all DEBRA literature, including our 'In Touch' newsletter, information about our Holiday Homes, local or general meetings, are available on our website or through the DEBRA offices.

## Authors

Karen Snelson (DEBRA EB Clinical Nurse Specialist) – linked to St. Thomas' Hospital

Jane Clapham (DEBRA EB Clinical Nurse Specialist) – linked to St. Thomas' Hospital

These guidelines were originally generated for adult patients under the care of the DEBRA adult EB team & St Thomas' Hospital, London. We would be very happy to offer general advice, however your patient may already be under the care of another EB team. If so, please contact them directly (contact details on back page).

# Guidelines for the practical care of adult patients with Epidermolysis Bullosa during surgical procedures.

## Aim

To provide all staff involved with the care of patients with Epidermolysis Bullosa (EB) undergoing surgical or invasive procedures with clear guidelines and advice to ensure best practice at all times. This is in line with the WHO safe surgical checklist guidance of 2009<sup>(5)</sup>.

## Rationale

EB is a group of rare genetically determined disorders characterised by excessive susceptibility of the skin and mucosa to blister even after trivial shear forces and mechanical trauma. Management of those with EB is often complex and undergoing even routine procedures has the potential to compound their already difficult condition. Whilst in hospital there is a risk of significant skin or mucosal damage and secondary complications as a result of undergoing routine general procedures.

## Introduction to EB

There are 4 main subtypes of EB:

EB Simplex, Junctional EB, Dystrophic EB and Kindler's Syndrome.

It is those affected by Dystrophic EB who will be seen most frequently as they may require, as a consequence of their disease, frequent diagnostic or therapeutic procedures under general anaesthetic.

Common surgical procedures include repair of syndactyly ("mitten glove" deformity), release of contractures, dental extraction, oesophageal dilatation, formation and repair of gastrostomy sites, excision of Squamous Cell Carcinoma, skin grafting and limb amputation.

The EB patient is the expert in managing the condition and will guide health professionals wherever possible. However, they are most vulnerable when asleep as they are unable to self-advocate or advise staff about necessary precautions to be taken<sup>(3)</sup>. Forward planning and communication is the key to a successful outcome.

## Pre-Assessment Guidance

Patients with EB have a number of important issues to address in the pre-operative evaluation. If possible, seeing these patients in consultation a week or two ahead of the operative date is useful because it allows data to be collected and consultation to occur in an unhurried manner that does not risk delaying surgery<sup>(6)</sup>.

Obtain records of previous anaesthesia	Valuable source of information regarding optimal management of the patient with EB undergoing the procedure.
Full Blood Count U&E Clotting Screen	For taking blood samples a gentle pair of hands is often better than a tourniquet.  Iron deficient & anaemia of chronic disease are common.  Renal & cardiac dysfunction may be found in EB.
Assess for possible renal & cardiac complications	May be present in EB & pre-operative echocardiogram should be considered.
BMI	Malnutrition & low body weight & BMI are frequently seen.
MRSA screen Infection control	Treat as per local guidelines.  Infection related to compromised skin integrity & poor immunity related to malnutrition & chronic disease is common in EB.  Treat as per local guidelines & prophylactic antibiotics should be considered.
Gastro-Oesophageal Reflux Disease is common & there is a high risk of aspiration	Patients with EB have a higher risk for gastro-oesophageal reflux <sup>(6)</sup> Antisecretory/mucosal protectant prophylaxis may be required.  Occurrence of oesophageal strictures is common & anatomically these develop high in the oesophageal tract. Those with oesophageal strictures may have pooled secretions & particulate matter that put them at risk of aspiration <sup>(6)</sup> .
Review recent or long term corticosteroid use	Systemic & topical use.
Airway assessment	Microstomia & limited mouth opening, fixed & scarred tongue, limited neck movement due to contractures, poor dentition & oral blistering are all common features.  Dental caries & restorative dental work may be extensive.  For detailed advice please contact the EB nursing team.
Musculoskeletal assessment	Extensive contractures & osteopaenia/osteoporosis may be present. This may result in difficulties achieving optimum procedural positioning.
Psychological preparation	Reassurance & full explanation of the procedure is essential.  Contact the EB Psychotherapist via EB office if appropriate.

## Pre-Operative Preparation and Anaesthetic Management

Contact EB Adult Nursing Team	<p>For specialist advice &amp; support during admission (see details below).</p> <p>In addition Dermatology Outreach Nurses may provide practical help with dressings.</p>
Identity bracelets	Apply with extreme care – ideally over a protective dressing or tubifast.
“Handle with Care” stickers	Available from EB team – ensure these are placed on all patient notes & (if patient consents) they can also be applied to gown as an easy visual reminder.
Anti embolitic management	<p>Avoid TEDS.</p> <p>Flowtron boots are recommended where available.</p>
Supply of suitable dressings & Silicone medical adhesive remover e.g. Apeel® or Niltac® (or a 50/50 preparation) should be taken to theatre with patient	To avoid inappropriate use of adherent dressings & ensure the safe removal of any dressing, tape or monitoring stickers that may be inadvertently applied.
Moving & Handling Issues Pressure Relief	<p>Request assistance &amp; guidance from the patient as appropriate.</p> <p>Minimise the number of transfers.</p> <p>e.g. anaesthetise in operating theatre to avoid at least one episode of patient transfer<sup>(2)</sup>.</p> <p>Transfer using “lift and place” approach<sup>(1)</sup> – never slide.</p> <p>Use of “Pat Slides” is strictly contraindicated.</p> <p>Gloved hands in contact with the skin can cause damage to fragile skin – where feasible gloves should be well lubricated. (Take care to ensure gloves/hands are free from lubrication when handling equipment).</p> <p>EB Nursing team will provide advice appropriate to each individual regarding safest transfer – use of the <i>HoverMatt</i>® is highly recommended for all lateral transfers – contact the EB office or nursing team to arrange use.</p> <p>Use KCI RIK operating table pads for maximum pressure relief.</p>

Skin	Blisters & erosions may be present & dressings should be left in situ wherever possible. If removal of dressings is unavoidable, cling film may be used as a temporary covering to the skin.
Skin preparation	Avoid rubbing or stroking the skin.  Cleansing fluid can be poured over limb & patted dry or a cleansing swab can be placed on skin, gentle downward pressure applied & then removed.
IV access	Use gentle pressure to distend veins & aid cannula insertion. If a tourniquet is used this should be well padded.  Secure cannula with Episil® or Mepitac® tape & k-band®.  In addition, the skin beneath the cannula should be protected from trauma e.g. with Mepilex Transfer®, Mepilex Lite® or similar non-adherent dressing.  To secure central & arterial lines suturing should be considered.
Eyes	Never tape the eyelids – instead close gently & then cover with Geliperm® hydrogel sheet.  Eyelid contractures may be present. There is a risk of corneal abrasion.
Theatre drapes	Secure drapes with a carefully positioned towel clip. Avoid use of sticky tape.
Airway management	After securing the airway, the priority is the avoidance of trauma & further bullae formation – care must be taken when applying face masks, head tilting and lifting chin.  Wrap foam padding around tape ties before securing ET tube to protect the skin on the face & neck.  Cover the areas of face where mask &/or anaesthetist's fingers will rest with a protective layer of suitable non-adherent dressing such as Mepitel One®, Geliperm® or ActiformCool®.  Cricoid pressure is not contraindicated but pressure should be applied evenly and with no sideways movement <sup>(4)</sup> .

Detailed advice & guidance on choice of anaesthesia & airway management (intubation) is available in Guys & St Thomas' NHS Trust Anaesthetic Guidelines<sup>(1)</sup>. Please contact the EB nursing team for more information if required.

## Epidural Management

<p>Skin preparation as above.</p>
<p>Avoid use of “sticky drapes”.</p>
<p>Use of adhesive dressings to safely secure the epidural is unavoidable unless suturing (using a tunnelling method) is an option. Use of medical adhesive removal spray is essential when removing the epidural in order to avoid skin damage.</p>
<p>Protect the skin on the spine from potential damage caused by pressure from cannula by applying Mepilex Transfer® to the back underneath the line.</p>
<p>Wherever possible allow the patient/carer to remove dressings when the epidural is removed.</p>

## Intra-Operative Management and Monitoring

<p>Oxygen saturation monitoring</p>	<p>Nail &amp; hand deformity is common &amp; therefore it may not be possible to apply the probe to a digit. It may be necessary to use the ear lobe.</p> <p>If the finger probe is used it is suggested that the finger is well lubricated &amp; then protected with the tip of a glove before the probe is applied<sup>(2)</sup>.</p>
<p>BP</p>	<p>Apply 2-3 layers of soft padding (e.g. soft-band) beneath the cuff.</p>
<p>ECG</p>	<p>Use non-adhesive electrode pads wherever possible.</p> <p>Adhesive electrodes can be used if the adhesive part is removed &amp; the electrode secured in place with Mepitac®. Alternatively the electrode can be placed onto a defib pad sandwiched between two pieces of Mepitel®<sup>(2)</sup> or stuck directly onto Mepitel One® (Note that the readout can be erratic with these methods).</p>
<p>Temperature control &amp; monitoring</p>	<p>Standard tympanic temperature monitoring advised.</p> <p>Avoid tempadots.</p> <p>To maintain patient body temperature during the procedure an adjustable warming system (e.g. Bair Hugger) may be used.</p>
<p>Trolley, bed &amp; equipment</p>	<p>Ensure that all equipment coming into contact with the patient is well padded &amp; lubricated where appropriate.</p>
<p>Incidental pressure</p>	<p>Avoid staff inadvertently leaning on or resting instruments on the patient.</p>
<p>Diathermy</p>	<p>Consider use of bipolar diathermy or harmonic scalpel as adhesive pads should be avoided wherever possible.</p> <p>If unavoidable then the pad should be removed with extreme caution &amp; generous use of silicone medical adhesive remover spray or 50/50.</p>

## Occasional & non-routine intra-operative procedures

Urinary catheterisation	Use a small gauge silicone catheter (10ch or smaller) & ensure that it is well lubricated.  Position catheter tubing with care to avoid potential skin damage.
Naso-gastric tube insertion	Avoid use of rigid NG tube.  Lubricate small gauge tube well before insertion & position with care.
Use of stirrups for positioning during procedure	If required the legs should be well padded for protection first.

## Post Operative Management & Analgesia

Extubation	Awake extubation should be considered to minimise potential airway obstruction & the need for mask pressure on the unprotected face.  Oropharyngeal suctioning can lead to life threatening bullae formation <sup>(4)</sup> .  Post Operative oxygen should be administered via a face mask padded with Mepilex Transfer <sup>®</sup> . Alternatively protect the face with a dressing such as Geliperm <sup>®</sup> .
Pharyngeal suction	Direct vision suction only.  Avoid yanker suckers where possible.
Nutritional requirements	Special diets may be required & the advice of a dietitian with knowledge of EB should be sought (EB dietitian can be contacted via EB office).  Constipation may be a chronic problem.  Many people with EB will have a gastrostomy.
Beds/ mattresses	Continuous pressure relieving system e.g. Repose <sup>®</sup> should be used. The KCI Visio <sup>®</sup> mattress should be used if the patient is at high risk.  Wherever possible the patient should have an electric bed to enable self positioning & reduce the risk of skin damage as result of manual handling.
Analgesia	Consider use of regional anaesthesia as an adjunct to general anaesthesia <sup>(1)</sup> .  Pain management as per WHO analgesic ladder is recommended.  PR analgesia should be used with extreme caution (risk of damage to fragile anal margins).  Use of morphine is NOT contraindicated in EB <sup>(1)</sup> .

## Theatre Essentials

- SpO2 ear probe
- ECG electrodes placed on defib gel pads
- Mepitel One®, Geliperm® and ActiformCool® to protect face from masks
- Silicone medical adhesive remover e.g. Apeel® or Niltac® spray to remove tapes & dressings safely
- Soft band
- Mepilex Transfer® to protect back if Epidural used
- Mepitel®, Mepilex® or Episil® to secure venflon
- Mepitac® to secure ETT or LMA. Alternatively use foam padding around tape ties
- Cling Film to protect skin temporarily if dressings are removed
- Selection of Classic LMAs size 2- 2.5
- Nasal Mask (Goldman)
- Selection of laryngoscopes
- Fibre optic laryngoscope

## To be avoided...

Anything sticky!

But don't panic! If something has been inadvertently applied then remove using silicone medical adhesive remover spray. If this is not available or appropriate please leave in situ and ask the patient to remove it later. Much damage occurs when people panic and try to remove something immediately – unless it is essential that the item is removed it is far better to leave it to the patient or their carer.



## References

1. Guys & St Thomas' NHS Trust (2010) Guidelines for Anaesthetic Management of Epidermolysis Bullosa Patients
2. Herod et al (2002) Epidermolysis Bullosa in children: pathophysiology, anaesthesia and pain management. *Paediatric Anaesthesia* 12: 388-397
3. Sweeney K (2009) Protocol for the pre-operative, intra-operative and post-operative care of a patient with Recessive Dystrophic Epidermolysis Bullosa. St James's Hospital, Ireland.
4. Ames W, Mayou B & Williams K (1999) Anaesthetic management in epidermolysis bullosa. *British Journal of Anaesthesia* 82 (5): 746-51
5. WHO (2009, January 26). National Patient Safety Agency. Retrieved October 12, 2010, from [www.npsa.nhs.uk/advise](http://www.npsa.nhs.uk/advise).
6. Goldschneider K, Lucky A, Mellerio J et al (2008). Perioperative care of patients with Epidermolysis Bullosa: proceedings of the 5th international symposium on Epidermolysis Bullosa, Santiago Chile, December 4–6, 2008. *Pediatric Anesthesia* 2010 20: 797–804

<b>Product</b>	<b>Company</b>
Flowtron Boots	Huntley Healthcare Limited
Apeel	Clinimed Limited
Niltac	Trio Healthcare International Limited
HoverMatt	Hovertech International
KCI RIK	KCI Medical Limited
KCI Visio	KCI Medical Limited
Episil	Advancis Medical
Mepitac	Molnlycke Healthcare
Mepital One	Molnlycke Healthcare
Mepilex Transfer & Mepilex Lite	Molnlycke Healthcare
K band	Urgo Ltd
Geliperm	Geistlich Sons Limited
ActiFormCool	Activa Healthcare Limited
Bair Hugger	Arizant UK Limited
Repose	Frontier Therapeutics Limited

### Further support and advice

Further details of products listed in Guidelines can be obtained from the adult nursing team contacts listed on the back page.

## Contact details

All Nursing & Social Care Services can be contacted Monday to Friday 9am-5pm.

### DEBRA Adult Nursing Service – Linked to St Thomas' Hospital

Secretary to the EB Adult Nurse team	01527 456968 (8.00am – 2pm Mon – Fri)
Hospital – EB Secretary	0207 188 6399
Out of hours on call dermatologist	0207 188 7188
EB Nurse Consultant (Adults)	07775 688324 (9.00am – 5pm Mon – Thur)

### DEBRA Children's Nursing Service – Linked to Great Ormond Street Hospital

EB team	0207 829 7808
Emergency on call service	0207 405 9200 (ask for EB Nurse on call)

### Children's Nursing Service – Birmingham Children's Hospital

EB team	0121 333 8224
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### Adult Nursing Service – Solihull Hospital

EB team	07846 986987 including out of hours
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### Scottish Nursing Service

Nursing team	01698 477777
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### DEBRA Social Care Managers

South England	01344 771961
Midlands	01299 826999
North England	07920 231271
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# Inpatient management of children with recessive dystrophic epidermolysis bullosa: A review

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## Abstract

Recessive dystrophic epidermolysis bullosa is a disorder marked by skin and mucosal blistering after minimal trauma. Even the most routine procedures in the hospital, if done incorrectly, can precipitate extensive skin loss, pain, and scarring. Most providers have little experience working with patients with this degree of skin fragility. When a person with recessive dystrophic epidermolysis bullosa is admitted to the hospital, there are multiple considerations to keep in mind while strategizing an effective care plan: avoidance of new blisters with a “hands-off” approach; careful consideration of all indwelling devices; symptomatic management of pain, itch, and anxiety; coordination of dressing changes; aggressive treatment of skin infections; environmental and staffing considerations; and awareness of other chronic complications that affect care, such as anemia, malnutrition, and chronic pain. To minimize discomfort for patients with recessive dystrophic epidermolysis bullosa during the hospital stay, inpatient care teams should understand these considerations and modify the care plan accordingly. Prior preparation by the hospital facility and inpatient care team will facilitate the delivery of safe and effective care and greatly improve the overall patient experience.

## KEYWORDS

epidermolysis bullosa, health care delivery, immunobullous disease, quality of life

## 1 | INTRODUCTION

Inherited epidermolysis bullosa (EB) is a heterogeneous group of chronic skin disorders characterized by fragility and blistering of the skin and mucous membranes. EB is complex and multisystemic in severe subtypes. For individuals with recessive dystrophic EB (RDEB), even the most routine procedures, such as moving the patient or measuring vital signs, can precipitate blistering and extensive skin loss.

Most providers outside of EB specialty centers have limited knowledge about the accommodations and specific handling techniques that these individuals require. Although there are guidelines for the general care of children with RDEB, there are none specifically for care in the inpatient setting.<sup>1,2</sup> The purpose of this review is to provide a practical resource to help facilitate inpatient

admissions and safe and effective day-to-day treatment on the hospital floor for this vulnerable group.

The first half of this review will provide an overview of the clinical presentation of RDEB and commonly associated complications. The second half will review general inpatient management, including safe patient handling, initial patient assessment, and inpatient wound care for individuals with RDEB.

## 2 | PATHOGENESIS

Mutations in the gene that encodes collagen VII weaken the structural adhesion within the skin, allowing the skin layers to separate with only minimal trauma, resulting in blisters and erosions.<sup>3</sup>

### 3 | CLINICAL PRESENTATION AND ASSOCIATED COMPLICATIONS

Wounds can occur anywhere on the body. Blistering is often worse in areas subject to repeated trauma, such as the hands, feet, and bony prominences (Figure 1).<sup>4</sup> Chronic wounds can lead to scarring, joint contractures, pseudosyndactyly (mitten hand deformity), and greater risk of cutaneous squamous cell carcinoma.<sup>5</sup>

#### 3.1 | Pain and itch

Pain is an almost constant symptom in RDEB patients and can severely affect quality of life and complicate daily activities.<sup>6</sup> Acute pain results from the formation and expansion of bullae and the irritation of erosions, in addition to anal fissures and reflux.<sup>6</sup> Sources of chronic pain include chronically inflamed wounds, joint contractures, bone pain, and constipation.<sup>6</sup>

Individuals with RDEB can experience severe and debilitating itching, which can originate from healing wounds or intact skin.<sup>7</sup>

The mechanism underlying the itch in RDEB is poorly understood, but it is likely that it is due to the disease itself, as well as certain triggers such as inflammation, concurrent opioid use, heat, sweating, and stress.<sup>7</sup> Controlling itch is particularly important because chronic scratching can result in trauma and blister formation.

#### 3.2 | Infection

The bacterial burden of a wound exists on a spectrum, starting at one end as contamination, with the potential to progress to colonization, critical colonization, and ultimately frank infection.<sup>8</sup> Contamination and colonization are normal states and require no treatment, but critical colonization, a state in which the bioburden is enough to impair wound healing, should be treated with topical antimicrobials.<sup>8</sup> To minimize antimicrobial resistance, systemic antibiotics are typically reserved for frank infections. The diagnosis of infection and appropriate treatment are discussed in a later section in this review.



**FIGURE 1** Clinical presentation of recessive dystrophic epidermolysis bullosa: (A) right lateral back, (B) buttocks, (C) left arm, (D) right knee. Diffuse erosions and scarring typical of a patient with recessive dystrophic epidermolysis bullosa. Blistering can be worse in areas subjected to repeated trauma including the (B) buttocks, (C) elbows, and (D) knees

The most common culprit organisms are *Staphylococcus aureus*, *Streptococcus* species, and *Pseudomonas aeruginosa*.<sup>1,8</sup> Wound infection is generally diagnosed clinically according to wound size, exudate, odor, pain, surrounding erythema, and edema,<sup>8</sup> but these classic signs of inflammation can be diminished or obscured in chronic EB wounds.<sup>8</sup>

Systemic infections are less common than cutaneous infections but can be lethal because of chronic malnutrition and a weakened immune system and most often arise from cutaneous infection.<sup>2</sup>

### 3.3 | Nutrition

Maintaining adequate nutrition is paramount because malnutrition can impair wound healing,<sup>9</sup> but management of nutrition is complicated in EB because of high caloric demand secondary to accelerated skin turnover, blood and protein loss through wounds, recurrent infections, and chronic inflammation.<sup>2,9</sup> Low intake secondary to dysphagia and constipation compounds the risk of malnutrition.<sup>2,9</sup> Iron; zinc; selenium; folate; and vitamins A, D, and B6 deficiencies have been observed in association with EB.<sup>2</sup> Some of these deficiencies have been implicated in impaired wound healing, osteoporosis, and cardiomyopathy.<sup>2</sup> Mixed anemia secondary to iron deficiency and chronic inflammation is frequent in individuals with EB and may also contribute to impaired wound healing.<sup>9,10</sup>

### 3.4 | Gastrointestinal

The gastrointestinal tract is one of the most common sites of extracutaneous complications in EB.<sup>11</sup> Wounds in the oral cavity and esophagus can lead to odynophagia, dysphagia, unwillingness to eat, and reflux.<sup>11</sup> Dental caries and premature loss of teeth can complicate oral hygiene and feeding.<sup>12</sup> In the small and large bowel, wounds can cause protein and blood loss, contributing to anemia, hypoalbuminemia, hypoproteinemia, and malabsorption.<sup>11</sup> Anal erosions and fissures result in painful defecation and constipation, further decreasing willingness to eat.<sup>11</sup> Chronic blistering can lead to strictures and cause ankyloglossia, microstomia, dysphagia, chronic constipation, and megacolon.<sup>11</sup>

### 3.5 | Ocular

Corneal abrasions and blistering may occur because of the fragility of the conjunctiva and cornea.<sup>13</sup> Chronic corneal wounds can precipitate scarring. In the operating room, skin contractures may prohibit complete closure of the eyes, putting the person at risk of corneal damage.<sup>14</sup>

### 3.6 | Cardiac

Dilated cardiomyopathy is a rare complication of EB and is probably due to multiple factors, including micronutrient deficiencies (especially selenium and carnitine), chronic anemia, iron overload from transfusions, and viral myocarditis.<sup>15</sup>

## 3.7 | Renal and genitourinary

Blistering along the genitourinary mucosa can lead to dysuria.<sup>16</sup> Chronic blistering can lead to strictures and urethral meatus stenosis, which can result in urinary retention, bladder distention, hydroureter, and hydronephrosis.<sup>16</sup>

## 3.8 | Psychiatric

Depression is common in individuals with EB, who at times can have suicidal ideations or gestures.<sup>17</sup> They may also experience anxiety and posttraumatic stress disorder secondary to painful dressing changes. Social isolation secondary to the visibility of their skin involvement is common.<sup>18</sup>

## 4 | OVERVIEW OF INPATIENT MANAGEMENT

Successful inpatient management of an individual with RDEB requires a multipronged approach to care. Avoidance of new blisters is critical and is best done with a hands-off approach to patient handling. The use of any indwelling medical devices should be carefully considered, because insertion and removal can precipitate trauma. Symptoms of pain, itching, and anxiety are common in individuals with RDEB and should be regularly assessed and treated. Caring for an individual with RDEB can be a significant time commitment and may require certain staff members to become “specialized” in dressing changes. Dressing changes should be carefully coordinated to minimize discomfort. Skin infections are common and should be treated aggressively, as they can lead to life-threatening systemic infections. The team should be aware of the chronic complications commonly associated with RDEB that can affect care, such as anemia, malnutrition, and chronic pain.

## 5 | EMERGENCY DEPARTMENT CONSIDERATIONS

When an individual with RDEB is received in the emergency department, the most appropriate service to admit them to needs to be considered. In the United States, hospitalist services, for example, are likely to be the most experienced in coordinating inpatient care among consultants and tend to have more availability on the unit. Depending on the reason for admission and the severity of the illness, individuals with RDEB may be admitted to intensive care units, surgical services, or specialty services. Furthermore, this decision will differ for different countries according to the typical distribution of specialty trainees within ward services. Second, although we review general handling procedures below, patients will benefit from carrying a letter or card that outlines individualized care instructions to help facilitate and orient new caregivers.

## 6 | AVOIDANCE OF NEW BLISTERS

A major portion of inpatient care for RDEB is dedicated to preventing new blisters. Although completely preventing new blisters is impossible, adhering to the following suggestions can minimize the risk of developing new wounds and greatly increase patient comfort throughout the hospital stay (Figure 2).

### 6.1 | Pressure relief

Pressure areas are particularly at risk of rapid wound development.<sup>4</sup> A pressure-redistributing mattress should be used for the duration of the stay (Repose, Frontier Medical Group, Blackwood, Caerphilly, UK; AccuMax Quantum, Hill-Rom, Batesville, IN; Rest-Q, Comfortex, Winona, MN).<sup>4</sup> If the person is scheduled for surgery, a padded operating table should be used or the table should be lined with sheepskin.<sup>19</sup> Any furniture that will support the person's weight at any point during the hospital stay should also be padded, including the toilet seat, bath chair, and bed railing.<sup>4</sup> Egg crate padding can be cut to cushion these items.

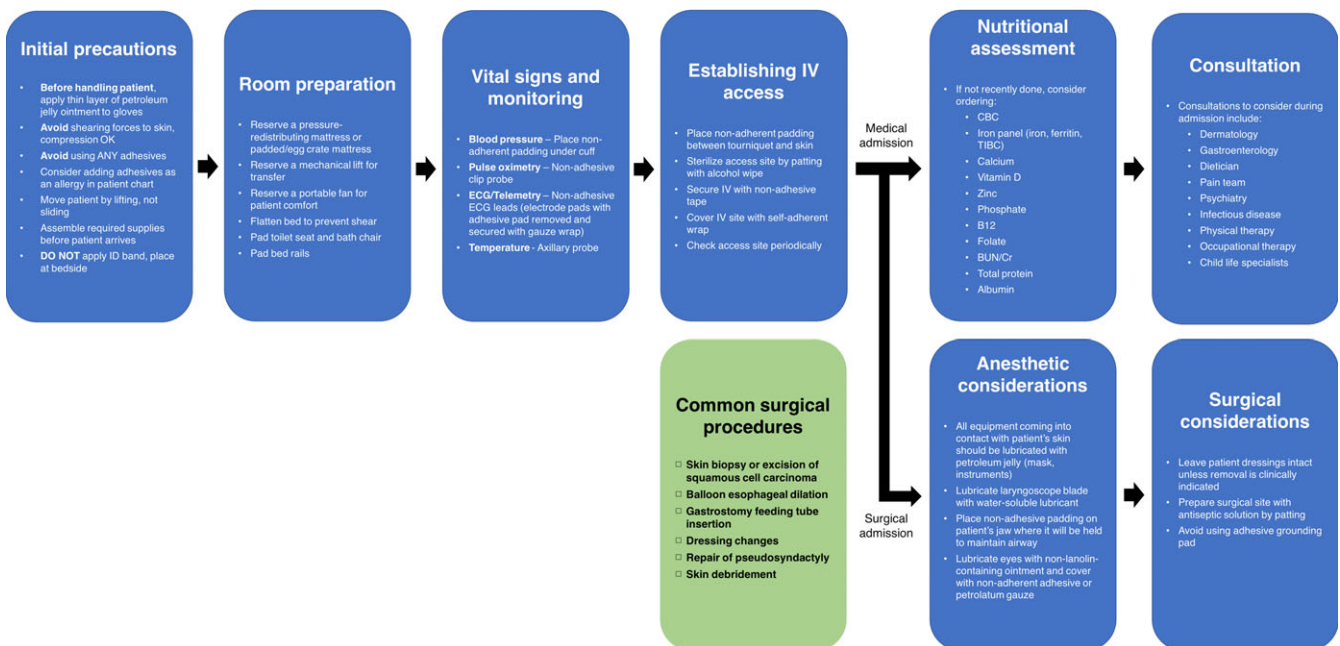
### 6.2 | Patient handling

Given the skin fragility and propensity to form blisters in individuals with RDEB, handling can be a challenge; a hands-off approach is advised whenever possible.<sup>4</sup> This is not to say that the person should be ignored. Essential care needs to be provided, potentially in a consolidated manner. When handling is unavoidable, several general management principles should be followed to minimize trauma (Figure 2):

- Before handling the person, apply a thin layer of ointment on gloves and medical devices that will have direct contact with patient skin.<sup>20</sup>
- Apply firm but gentle pressure; avoid shearing forces.<sup>20,21</sup>
- When moving the person, lift instead of sliding.<sup>20,21</sup>
- Avoid applying any kind of adhesive directly on the person's skin. If adhesives are accidentally used or are required to secure access, they can be removed safely using medical adhesive remover (Niltac, ConvaTec, Deeside, Flintshire, UK; Uni-Solve, Smith & Nephew, London, UK; Detachol, Ferndale Labs, Ferndale, MI).<sup>20,21</sup>
- Avoid rubbing of the skin.<sup>21</sup>

### 6.3 | Patient monitoring

Many of the aforementioned principles should be applied to modify the monitoring setup (Figure 2). Electrocardiographic lead adhesive pads should be removed and leads secured with gauze wrap or non-adhesive dressing (Mepitel, Mölnlycke Health Care, Gothenburg, Sweden; Silflex, Advancis Medical, Nottinghamshire, UK; Adaptic, Acelity, San Antonio, TX).<sup>2,21</sup> An oximeter clip probe should be used instead of an adhesive sensor.<sup>2</sup> Before taking blood pressure, several layers of nonadherent padding should be placed between the blood pressure cuff and skin as a cushion.<sup>2</sup> If monitoring of temperature is required, axillary probes are preferred because oral probes may cause oral blistering.<sup>20</sup> Indwelling urinary catheters should be avoided if possible because of the risk of developing urethral strictures. If urinary catheters are necessary, they should be well lubricated before insertion.



**FIGURE 2** Medical and surgical admissions flowchart. Medical and surgical admissions begin with similar preparatory steps but diverge after establishment of intravenous access

## 6.4 | Indwelling medical devices

Finding and maintaining vascular access can be particularly challenging in individuals with RDEB. Visualization and palpation of their veins can be extremely difficult. The most experienced providers are often needed to secure intravenous (IV) access; consultation with anesthesia or vascular access teams may be necessary. It is important to wrap multiple layers of nonadhesive padding around the limb before placing any tourniquet device.<sup>22</sup> The access site should be sterilized by patting rather than rubbing with an alcohol wipe. The line should be secured with nonadhesive tape and covered with self-adherent wrap.<sup>20</sup> IV lines tend to dislodge easily in individuals with EB, which can be emotionally and physically distressing for the patient and staff, so periodic monitoring is required.<sup>20</sup> When IV placement becomes a problem, finding oral alternatives to medications, if possible, should be attempted.

Central venous access may be preferable in cases in which a long-term hospital stay is anticipated and IV access is necessary. The benefits of establishing central venous access, including less potential trauma from repeated skin puncture and provision of a convenient access site, should be weighed carefully against the greater likelihood of systemic infection. As with peripheral access, central lines in individuals with RDEB are prone to catheter migration and dislodgement. In addition to securing the line with nonadhesive tape and self-adherent wrap, anchoring sutures can be placed for further support.<sup>23</sup> Placing sutures through the full thickness of the skin, deep to the cleavage plane, may minimize secondary trauma. It has been reported that a cuffed line is more secure than an uncuffed line in individuals with RDEB.<sup>23</sup> Tunneled lines are also safe to use, although femoral access should be avoided if possible.<sup>2</sup> Percutaneous IV central catheters are an alternative that allow for long-term access by securing on an extremity instead of a central location.

## 6.5 | Operating room considerations

Cutaneous dressings should be left in place unless removal is necessary (Figure 2).<sup>20</sup> To protect the eyes, a non-lanolin-containing ointment can be used as lubrication.<sup>24</sup> The eyes should then be covered with nonadherent adhesive or petrolatum gauze.<sup>25</sup> While preparing the surgical site with antiseptic, patting motions should be used instead of the typical rubbing motions.<sup>20</sup>

## 6.6 | Airway management

Microstomia, limited neck mobility, and ankyloglossia can all complicate airway management in RDEB.<sup>24,25</sup> Noninvasive ventilation should be avoided because it can precipitate facial trauma. Intubation using a fiberoptic bronchoscope is preferred because it can be less traumatic to the oral mucosa than direct laryngoscopy.<sup>25</sup> Endotracheal tubes should be half to one size smaller than predicted to avoid overinflation and trauma.<sup>25</sup> All equipment coming into contact with the person's skin or mucosa, including the facemask and

laryngoscope blade, should be well lubricated.<sup>14,25</sup> Nonadhesive padding should be placed on the jaw before manipulation.<sup>14</sup> Endotracheal tubes should be secured using cotton tape.<sup>17</sup> For a more comprehensive overview regarding anesthetic and surgical considerations, refer to the referenced articles.<sup>14,20,25</sup>

## 7 | INITIAL ASSESSMENT

### 7.1 | Pain and itch

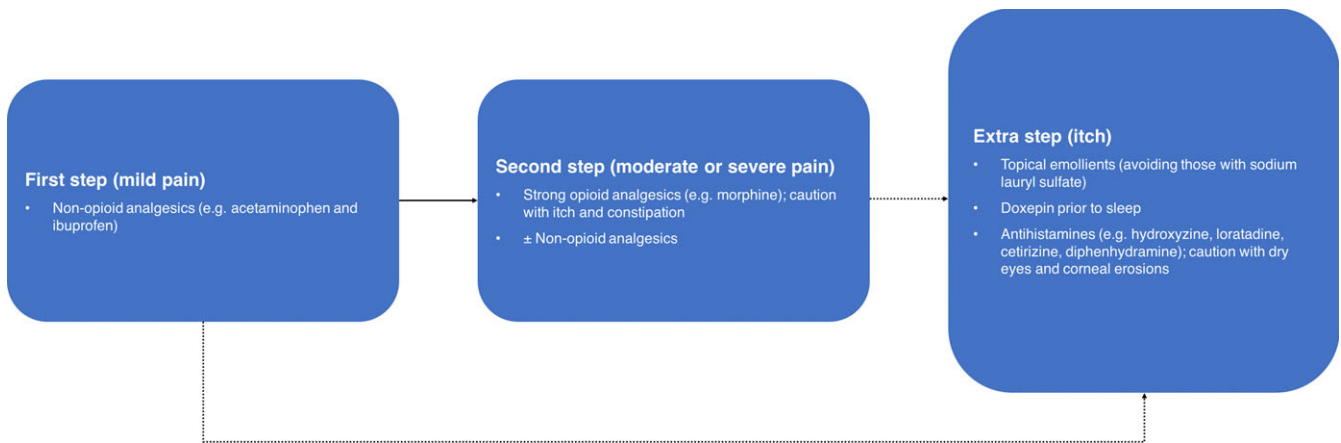
A modified version of the World Health Organization approach to pediatric pain management can be used to treat acute pain in individuals with RDEB (Figure 3).<sup>26</sup> With small wounds or minor pain, nonopioid analgesics (eg, acetaminophen, ibuprofen) can be used alone or together.<sup>26</sup> For moderate or severe pain, an opioid analgesic (eg, morphine) should be used,<sup>26</sup> although opioids can worsen pruritus and constipation.<sup>11</sup> Anxiety can exacerbate anticipatory pain, so anxiolytics such as diazepam and lorazepam taken before a painful procedure may be helpful.<sup>27</sup> When possible for painful or frightening procedures such as biopsies, provide adequate anxiolysis (eg, oral midazolam), local anesthesia, or other forms of conscious sedation (moderate or deep). Conscious sedation, with appropriate bedside monitoring, can be effective and timely, helping avoid general anesthesia and minimizing time in the operating room.

For chronic pain, many individuals with RDEB use long-acting opioids to maintain a base level of comfort.<sup>6</sup> Tricyclic antidepressants and gabapentin have been used successfully in managing chronic wound pain in EB.<sup>6,28</sup> Additionally, nonpharmacologic therapies, including distraction, visualization, and other forms of cognitive behavioral therapy, have been recommended.<sup>6</sup>

For itch, topical emollients may be useful.<sup>2</sup> Topical corticosteroids have been used with some degree of success, but providers should be wary of the greater systemic absorption in individuals with EB because of impaired barrier function.<sup>2,29</sup> Although it is not thought that histamines cause the itching in EB, antihistamines such as doxepin can be tried for their sedative properties, especially before sleep,<sup>1</sup> although they should be used with caution because they can exacerbate dry eyes and corneal erosions.

### 7.2 | Nutrition

As mentioned previously, individuals with RDEB are at risk of developing malnutrition, which in turn will impair wound healing.<sup>9</sup> Because of the gastrointestinal complications associated with RDEB, many children will have had gastrostomy tubes placed for long-term nutritional supplementation.<sup>7</sup> If the person has not had nutritional laboratory tests performed in the last 6 months, the team should consider ordering those listed in Figure 2. If the results suggest underlying malnutrition, supplementary nutrition with nasogastric or gastrostomy tube feeding is recommended. Consultation with a nutritionist is important for each hospital admission.



**FIGURE 3** Pain and itch management flowchart. This approach is based on the World Health Organization's approach to pediatric pain management, which was modified to include an additional step for itch management. The dashed lines indicate that itch management does not necessarily have to follow pain management in a stepwise fashion

### 7.3 | Consultation

Care for individuals with EB requires a multidisciplinary team.<sup>2,30-32</sup> Consultation with the inpatient teams outlined in Figure 2 should be considered during every admission. The child life specialists, in particular, are an important team to consult. They play a crucial role in the care team, explaining indwelling devices to patients and parents, providing nonpharmacologic pain relief through distraction or relaxation techniques, and providing much-needed companionship.

## 8 | GENERAL WOUND CARE

In individuals with RDEB, proper wound care and dressing technique are important for several reasons. Dressings protect the skin as a physical barrier, reduce the risk of wound infection, and promote an environment conducive to wound healing.<sup>21</sup> A properly dressed, noninfected wound will typically heal gradually over time. Any non-healing wound lasting longer than 6 months should be assessed for squamous cell carcinoma.<sup>30</sup>

### 8.1 | Dressing regimen

The specific aspects of a wound care regimen, such as frequency of dressing changes and types of dressings used, vary from person to person depending on several variables, including extent of disease involvement, wound location and characteristics, patient preference, and the presence or absence of infection (Figure 4). Most regimens will include a nonadhesive primary dressing, which provides the contact layer (Mepitel, Molnlycke Health Care, Gothenburg, Sweden; Urgotul, Urgo Limited, Shepshed, Loughborough, UK; Silflex/Siltac, Advancis Medical, Nottinghamshire, UK; Adaptic Touch, Acelyty, San Antonio, TX). Secondary dressings can be placed over primary dressings to provide additional cushion and to

absorb exudate (Mepilex, Tefla, Medtronic, Minneapolis, MN; Polymem, Ferris Mfg. Corp., Fort Worth, TX). Lastly, the dressings are typically reinforced with tubular bandages or self-adhering wrap. Details regarding specific dressing types are outside the scope of this article but can be found in other comprehensive references.<sup>1,21</sup> In adapting a wound care regimen to the inpatient setting, it is important to solicit patient and family preferences. Individuals with RDEB and their caregivers have had a lifetime of experience optimizing their wound care regimen to best prevent injury and maximize comfort,<sup>4</sup> but in certain situations it will be necessary to make changes to a person's preferred wound care regimen to optimize wound healing. In these cases it is important to engage the person and family in dialogue to negotiate a wound care plan that is acceptable to them and the care team.

### 8.2 | Gastrostomy tube site care

Gastrostomy tube sites should be dressed in a fashion similar to that of the rest of the skin. In the case of leakage from the gastrostomy site, sterile saline should be used for cleaning and a topical barrier applied to minimize further irritation.<sup>21</sup> A superabsorbent secondary dressing can help contain leakage and allow the site to heal gradually.<sup>21</sup>

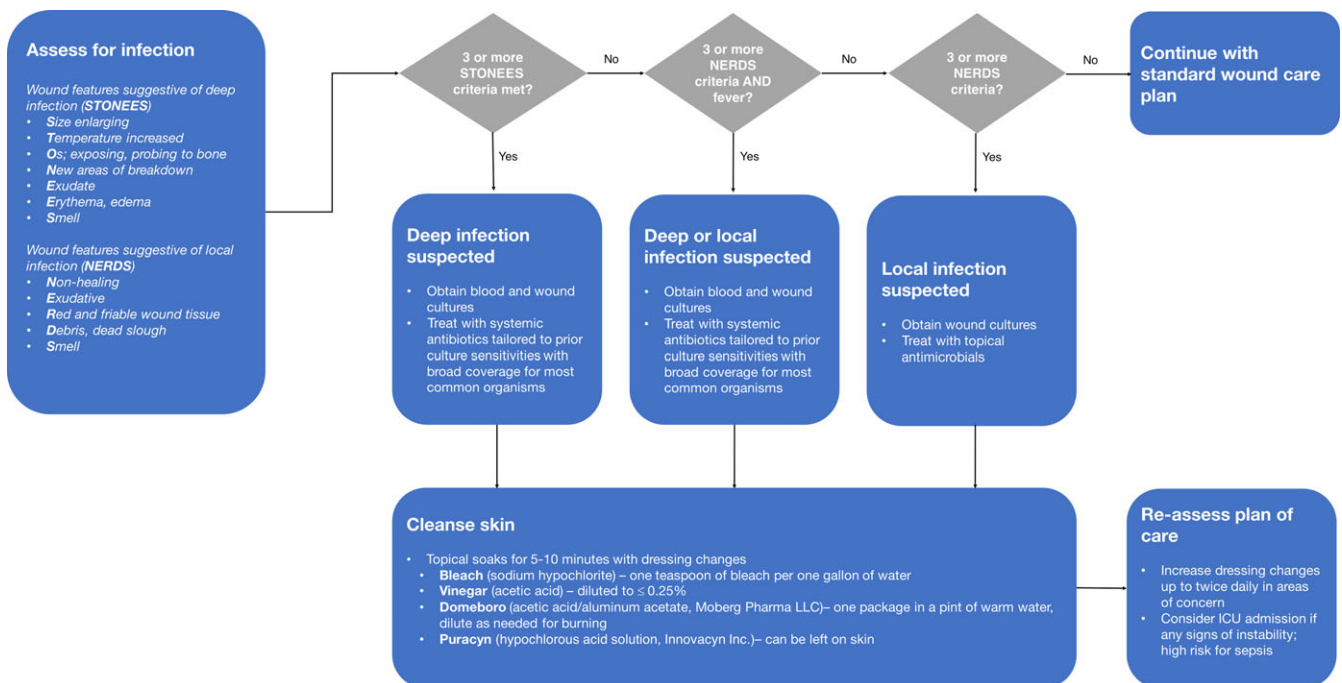
### 8.3 | Patient comfort

Procedural pain management is an important topic to discuss with the person and caregivers because dressing changes can be a major contributor to discomfort.<sup>6</sup> Providers can further contribute to comfort by maintaining a warm room temperature and taking precautions to ensure privacy.<sup>4</sup> Preparing dressings beforehand by unwrapping them, cutting them to shape, and adding petroleum jelly ("buttering") will also facilitate the dressing process. To preserve patient autonomy, the care team should discuss with the individual how much





**FIGURE 4** General inpatient wound care flowchart. General inpatient wound care begins with preparation of the person and the environment, followed by dressing removal, assessment of infected wounds, blister aspiration, and dressing application. Assessment of a potentially infected wound is further elaborated upon in Figure 5



**FIGURE 5** Wound infection management flowchart. Treatment of an infected wound will depend on whether a superficial skin infection or a deep skin infection is suspected. If wound infection is suspected, the skin should be cleansed after cultures are obtained. ICU, intensive care unit

involvement he or she wants in the dressing changes. In academic settings, privacy and clinical care must be carefully balanced with the unique learning opportunity that the person could provide to learners of all levels.

### 8.4 | Staffing considerations

The time commitment for dressing changes is significant in inpatient care because one-on-one nursing is often required for several hours.

It is our experience that a consistent approach to dressing changes increases compliance and facilitates the dressing process. Thus, asking for dedicated staff who are able to specialize in dressing changes and form a primary nursing team can be helpful. Caring for a patient with RDEB can be distressing not only for the patient, but also the staff involved. As an example, the warm room temperature and wound odor during dressing changes can make for a taxing working environment.<sup>4</sup> Awareness and management of psychological trauma is therefore essential to keep these team members effective.

## 8.5 | Bathing

There is no established standard of care regarding bathing frequency.<sup>21</sup> While in the inpatient unit, we recommend bathing or cleansing every day to every other day during dressing changes.<sup>2,19</sup> Removing dressings during a bath may make the dressing change process more comfortable because the bathwater can help dissolve adherent crusting.<sup>27</sup> Tub water can be supplemented with vinegar (7.5 L of 5% acetic acid or 11.5 L of 3% acetic acid in a full 160-L bathtub) or bleach (120 mL of bleach in a full bathtub) to reduce microbial burden and reduce the risk of infection.<sup>2,7,8</sup> Vinegar and bleach have been shown to have antimicrobial properties for Gram-negative and Gram-positive bacteria, respectively.<sup>33,34</sup> Salt (900-1000 g in a full bathtub) can be added to the water to create an isotonic solution and decrease pain.<sup>7</sup> A whirlpool bathtub, if available, allows for concurrent gentle wound debridement.<sup>19</sup> If a bathtub is unavailable, a shower with a cushioned shower seat is also an acceptable option.<sup>7</sup>

## 8.6 | Blister management

Blisters should be sterilely lanced and drained to prevent blister extension.<sup>1</sup> Using a large-gauge sterile needle, puncture the blister through the blister roof, parallel to the skin. Multiple puncture sites followed by gentle pressure with sterile gauze can be used to facilitate drainage. Leave the blister roof in place to minimize infection risk.<sup>2</sup>

## 8.7 | Managing infection

Patients should be assessed for infection during each dressing change because skin infection impairs wound healing and can lead to life-threatening systemic infection.<sup>1,19</sup> Distinguishing a superficial from a deep skin infection is important because the latter necessitates more aggressive treatment (Figure 5). NERDS (nonhealing, exudative, red or bleeding, debris, smell) and STONEES (size increasing, temperature > 3°F above normal, os [probes to or exposes bone], new areas of breakdown, erythema or edema, exudate, smell) are two mnemonics that may help providers clinically assess for wound infection, as they describe the clinical features of superficial and deep skin infection, respectively.<sup>35</sup> Although these mnemonics have not been validated in wounds related to EB, they have been validated for use in chronic wounds and recommended for use in EB in an expert consensus statement.<sup>1,35</sup>

If three or more of the STONEES criteria are met, the person will require a systemic agent.<sup>1</sup> Similarly, if three or more NERDS criteria are met, and the person exhibits systemic symptoms (fever, malaise), he or she should be treated using a systemic agent.<sup>1</sup> Systemic agents should begin with broad coverage for the most common organisms, including *S. aureus*, *Streptococcal* species, and *P. aeruginosa*.<sup>7</sup> Current and past bacterial culture results should then be used to tailor coverage.<sup>1</sup>

If three or more of the NERDS criteria are met, but the person does not have systemic symptoms, the wound should be treated with topical antimicrobials to decrease microbial burden.<sup>1</sup> Lipid-stabilized hydrogen peroxide cream has broad antimicrobial coverage and is well tolerated in individuals with RDEB.<sup>8</sup> Silver sulfadiazine also has a broad spectrum of activity, although it should be used for only a short period of time because argyria has been reported in individuals with RDEB with prolonged exposure to silver.<sup>8,36</sup> Topical antibiotics, including gentamicin and mupirocin, can be effective in the short term but should also be used cautiously in longer hospital stays to avoid the development of microbial resistance and adverse events from systemic absorption.<sup>22,37</sup>

If infection is suspected, the wound care plan should be modified appropriately. The frequency of dressing changes to the area of concern may need to be increased to prevent irritation secondary to excess wound exudate.<sup>21</sup> The wear time for the specific type of secondary dressing used will dictate how frequently dressings will need to be changed, because some manufacturers recommend changing when exudate is observed on the outer surface of the dressing and some recommend changing when wet or heavy.<sup>21</sup> Additionally, infection can amplify wound pain, so the pain regimen should be adjusted accordingly. Deep sedation with IV propofol and ketamine has been used successfully for dressing changes and deep whirlpool baths in individuals with extensive wound infection.<sup>38</sup>

## 9 | CONCLUSION

Individuals with RDEB experience complications in multiple organ systems, which complicates life inside and outside of the hospital. To minimize discomfort for these individuals during a hospital stay, inpatient care teams will need to factor in the multiple considerations addressed in this text while strategizing the care plan. Prior preparation by the hospital facility and inpatient care team will facilitate delivery of safe, effective care and greatly improve the overall experience of an individual with RDEB.

## ACKNOWLEDGMENTS

The authors would like to thank Kristen Corey, MD, for providing the clinical images used in Figure 1.

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**How to cite this article:** Li AW, Prindaville B, Bateman ST, Gibson TE, Wiss K. Inpatient management of children with recessive dystrophic epidermolysis bullosa: A review. *Pediatr Dermatol*. 2017;34:647–655. <https://doi.org/10.1111/pde.13276>