debra
for people whose skin doesn't work we do

Epidermolysis Bullosa practical care guidelines

Adult Surgical Procedures

Guidelines originated by Adult Nursing Team

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.

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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[5].

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[3]. 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[6].

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[6] 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[6].
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	For specialist advice & support during admission (see details below). In addition Dermatology Outreach Nurses may provide practical help with dressings.
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	Avoid TEDS. Flowtron boots are recommended where available.
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	Request assistance & guidance from the patient as appropriate. Minimise the number of transfers. e.g. anaesthetise in operating theatre to avoid at least one episode of patient transfer[2]. Transfer using "lift and place" approach[1] – never slide. Use of "Pat Slides" is strictly contraindicated. 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). EB Nursing team will provide advice appropriate to each individual regarding safest transfer – use of the *HoverMatt*® is highly recommended for all lateral transfers – contact the EB office or nursing team to arrange use. Use KCI RIK operating table pads for maximum pressure relief.

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[4].

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

Epidural Management

Skin preparation as above.
Avoid use of “sticky drapes”.
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.
Protect the skin on the spine from potential damage caused by pressure from cannula by applying Mepilex Transfer® to the back underneath the line.
Wherever possible allow the patient/carer to remove dressings when the epidural is removed.

Intra-Operative Management and Monitoring

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

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. Oropharangeal suctioning can lead to life threatening bullae formation[4]. Post Operative oxygen should be administered via a face mask padded with Mepilex Transfer®. Alternatively protect the face with a dressing such as Geliperm®.
Pharangeal 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® should be used. The KCI Visio® 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[1]. 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[1].

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.
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

Product	Company
Flowtron Boots	Huntley Heatlhcare 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 Theraputics 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 Out of hours on call dermatologist	0207 188 6399 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

Adult Nursing Service – Solihull Hospital	
EB team	07846 986987 including out of hours

Scottish Nursing Service	
Nursing team	01698 477777

DEBRA Social Care Managers	
South England	01344 771961
Midlands	01299 826999
North England	07920 231271
Scotland	01698 477777
General enquiries	01344 771961
DEBRA Office – Director of Nursing & Social Care	01344 771961

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