EPIDERMOLYSIS BULLOSA GUIDELINE	
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1.0 What is Epidermolysis Bullosa?

Health Care Professionals, who come in contact with children with Epidermolysis Bullosa (EB), should be aware of the general management of the condition and of the necessary techniques to minimise trauma and complications (Denyer et al. 2017). Ideally, management should take place in a specialist centre.

2.0 Definition of Epidermolysis Bullosa

Epidermolysis Bullosa is a genetic disorder characterised by blistering of the skin as a result of friction or trauma (Danial et al, 2014). However, mucus membranes can be affected including pharyngoesphageal mucosa (Denyer et al, 2017).

Inheritance pattern

Inheritance of EB can be autosomal recessive or autosomal dominant.

- Dominant inheritance occurs when one parent has the condition and there is a 50% chance in each pregnancy that their child will be affected. Dominant inheritance can also occur as a new mutation.
- Recessive inheritance occurs when both parents carry one gene for EB. There is a one in four chance for each pregnancy that their children will be affected (Denyer et al. 2017).

There are 4 classifications of Epidermolysis Bullosa (EB)	
Classification	**Subtype**
Simplex (EBS)	▪ Localised (EBS-loc) ▪ Generalised Intermediate (EBS-GI) ▪ Generalised Severe (EBS-GS) *(There are also additional rare basal subtypes)*
Dystrophic **Dominant Dystrophic**	▪ DDEB generalised (DDEB-gen) ▪ DDEB acral (DDEB-ac) ▪ DDEB pretibial (DDEB –pt) ▪ DDEB pruriginosa (DDEB-pr) ▪ DDEB nails only (DDEB-na) ▪ DDEB bullous dermolysis of the newborn (DDEB-BDN)
Dystrophic **Recessive Dystrophic**	▪ Recessive Dystrophic Generalised Severe (RDEB-GS) ▪ Recessive Dystrophic Generalised Intermediate (RDEB-GI) ▪ RDEB bullous dermolysis of the newborn (RDEB-BDN) ▪ Recessive Dystrophic Pruriginosa (RDEB-pr) ▪ Recessive dystrophic Inversa (RDEB-I) ▪ Recessive Dystrophic Localised (RDEB-loc) ▪ Recessive Dystrophic pretibial (RDEB-pt) ▪ Recessive Dystrophic centripetalis (RDEB-ce)
Junctional	▪ Junctional Generalised Severe (JEB-GS) ▪ Junctional Generalised Intermediate (JEB-GI) ▪ Junctional with Pyloric Atresia (JEB-PA)
Kindler Syndrome	

Complications

- Skin trauma resulting in blistering episodes due to improper handling of an infant.
- Infection
- Blistering of the oral, pharyngeal and oesophageal mucosa
- Management of a difficult airway
- Scar tissue formation and contractures
- Nutritional problems
- Chronic pain and pruritus
- Constipation and anal fissures
- Dysphagia
- Anaemia
- Cardiomyopathy
- Osteopenia
- Dental decay
- Renal Dysfunction
- Ophthalmologic issues i.e. erosions
- Pubertal delay
- Squamous cell carcinoma

3.0 Admission of an infant with Epidermolysis Bullosa

Equipment

Ensure the following equipment is available when accepting a baby or child with EB on to your ward.

- Newborn Epidermolysis Bullosa pack (located on Nazareth ward)
- Dressing trolley
- Appropriate dressings e.g., mepitel®, mepilex transfer®, mepilex®, tubifast® Mepitac® polymem®
- Corn flour.
- Height adjustable cot.
- Melolin roll.
- Angel care mat (non-invasive monitoring mat for cot)
- Orange needles/sharps box Digital thermometer, stethoscope, Dynamap
- Soft sterile gauze.
- Weighing scales.
- Skin swabs.

ACTION	RATIONALE AND REFERENCE
Admit infant to a large single room if available.	To minimise the risk of infection to the child with EB and reduce the risk of cross infection (OLCHC 2017b, CHI 2020) To provide the space required for parents, staff, and equipment for dressing changes and to reduce the risk of skin damage caused by caring for the infant in a cramped environment (Atherton and Denyer, 2003)
Remove any adhesive material from the room	Reduce the risk of accidental trauma to the skin
Wear a disposable apron and always decontaminate hands before and after patient contact	To minimise the risk of infection to the child with EB and reduce the risk of cross infection (OLCHC 2017b, CHI 2020)
Consider the temperature of the room. If possible, an air-conditioned room should be used. Only nurse the infant in an incubator if medically necessary e.g. prematurity	A hot humid environment can exacerbate blistering. The risk of developing blisters is increased for infants with EB when they are nursed in a warm environment (Denyer et al. 2017)
Nurse the infant in a height adjustable cot if available	A risk assessment, should be carried out by staff to prevent back strain for staff and parents during dressing changes as they can take some time (CHI 2019)
When handling an infant with EB, remember friction and sheering forces will cause skin damage - direct pressure is safe.	Correct lifting techniques prevent skin damage (Denyer et al. 2017)
Never lift the infant under the arms. When it is necessary to lift the infant, roll the infant onto his/her side, place a hand under the head and neck and a hand under the buttocks, allow the infant to roll onto your hands and lift. Nurse the infant on a soft surface such as a piece of melolin roll. This can be used also when lifting the infant.	To prevent skin damage caused by friction and to minimise trauma to skin (Denyer 2010)
Obtain baseline vital signs on admission. Daily temperature, pulse and respiration rate suffice. Increase frequency, if clinically indicated. **NOTE Do not use tempadot**. If using an axillary digital thermometer lift the infants arm gently and place the probe in the axilla. When removing gently lift the arm and remove thermometer.	Tempadots will adhere to the skin causing trauma and blistering (Nandi and Howard 2010)

When checking the heart rate using a stethoscope place the metal part over an item of clothing e.g. vest or dressings. Never place it directly onto the skin.	To prevent any skin trauma and further blistering (Nandi and Howard 2010)
If a blood pressure reading is required, ensure the cuff is not directly on the skin. Place it over an item of clothing or dressing	Blood pressure cuffs and tourniquets can exert sheering forces and cause skin blistering (Mortell and Azizkhan 2010).
Obtain skin swabs for culture and sensitivity and MRSA screen. Swab should be taken in a zigzag pattern and rotated gently over the wound to maximise contact with wound. A moist swab will help the collection of microorganisms	Early detection of infection can be addressed and treatment can commence (Mellerio 2010, OLCHC 2018). Rubbing the swab on the skin, may cause skin damage (Mellerio 2010).
If nasal and throat swabs are required, they should be taken under direct vision with a swab moistened with 0.9% saline	They may cause serious mucosal detachment and blistering (Nandi and Howard 2010)
Obtain a baseline weight and height. Line the scales with a piece of melolin roll and weigh the infant with dressings in place	The risk of skin damage is increased if the infant is naked and without dressings (Denyer 2014).
The identification band is applied over a piece of Tubifast. Ensure the band does not come into contact with the infant's skin.	To prevent skin damage caused by friction on the skin (Denyer 2010
<u>Pain Management</u> Pain is assessed using a validated, age appropriate pain assessment scale and analgesia administered accordingly. Non-pharmacological methods i.e. adequate preparation and a play specialist can also help with pain. Infants may require morphine prior to dressing changes. Paracetamol and Ibuprofen may be administered for general comfort. A plan should be made for the administration of analgesia based on individual needs.	To ensure patient is pain free and reduce anxiety of parents and child. Optimal pain management, is vital for a patient with EB to reduce the risk of skin damage (Goldschneider and Lucky 2010, Goldschneider et al. 2014, Denyer et al.2017)
Administer analgesia 30-45 minutes prior to examination and dressing change.	To ensure the maximum result of the analgesia is experienced during examinations/dressing changes (Goldschneider et al. 2014)
NOTE: Do not administer suppositories to an infant with EB.	Risk of trauma to anus (Snelson and Clapham 2010)

Skin Care	
On initial examination of the infant clinical photographs are taken, with parental consent.	Photographs can be used as a tool to make an assessment of wound healing (Ahn and Salcido 2008)
Bathing is delayed until the birth damage has healed, and is performed in conjunction with dressing changes.	Patient is most at risk from skin damage when naked (Denyer et al. 2017)
Commercial cling film can be used as a temporary dressing after bathing.	Cling film will adhere to itself and not on the skin of a child with EB (Nandi and Howard 2010)
If an umbilical clamp is present observe the surrounding skin for signs of friction from it. Apply a piece of Mepilex® transfer to the skin to protect the area if required. The clamp can be removed and replaced by a ligature.	Periumbilical cord damage is common from the plastic cord clamps (Denyer et al. 2017)
Avoid heel prick for neonatal screening use peripheral blood.	To avoid a degloving injury (Denyer et al.2017)
The skin must be inspected regularly and blisters burst. Burst all blisters with a sterile 23G orange needle inserting the needle at the base of the blister. The insertion and exit of the needle must must be made through the same entry and exit point. Gentle pressure can be applied with a piece of soft gauze to empty the blister fluid.	Blisters are not self-limiting and will extend if left intact (Lara- Corrales et al 2010, Denyer et al. 2017)
The roof of the blister should be left intact if possible and corn flour can be lightly dusted onto the area however if the blister area denudes a dressing should be applied.	Corn flour helps absorb blister fluid and reduces friction on the skin (Denyer 2010). Regular cornflour is used taking note of the expiry date.
Non-viable skin can be cut away.	To promote healing and prepare the wound bed (Lara- Corrales et al 2010)
Wounds must be dressed with a non-adherent dressing. Regular dressings can cause trauma to the skin. Internationally recommended dressings include Mepilex®, Mepilex transfer®, Mepitel, Polymem and Urgotul	These wound dressings are recommended as they do not adhere to the skin. They are 'sticky' to the touch but are easily removed from the wound without pain or trauma
Dressings are held in place with Mepitac® and Tubifast.	Dressings provide extra protection against injury when handling the child .They are used to protect existing wounds, promote healing, minimise trauma and prevent further blistering (Lara- Corrales et al 2010, Denyer et al. 2017). Dressing retention is important to reduce friction (Denyer et al. 2017)

When carrying out dressings, the infant should not be fully exposed. Dressings should be changed on a limb to limb basis. Paraffin gel can be used • General skin care • On crusted wounds, • On sticky material accidentally applied to the skin.	Naked infants can cause damage to themselves while they are undressed (Denyer 2010) • As a moisturiser • To soften the crust • To aid easy removal of the item adhered to the skin (Denyer 2010)
Infants with EB, should not be nursed naked. Select flat seamed soft clothes or turn garments inside out. Tags can be removed to prevent friction. Disposable nappies, can be used but nappies must be lined with a soft nappy liner, which should extend beyond the elasticised part of the nappy especially around the groin and at the top of the nappy.	Naked infants will cause damage to themselves due to friction from their own skin. Flat seamed clothes will create fewer traumas. Liners will prevent friction and trauma caused by the nappy (Denyer 2010, Denyer et al 2017)
To obtain a urine specimen, a clean catch method should be used.	Never use adhesive material on the skin of a child with EB as trauma will occur upon removal of same (Lara-Corrales et al 2010)
Feeding Some infants have problems with severe blistering of the mouth and sometimes are reluctant to feed. In the infant who is not breast feeding, use of a special feeding bottle is recommended (Haberman bottle).	A Haberman bottle minimises risk of trauma to the gum margin and reduces the need for strong sucking so even a weak suck delivers a satisfactory milk flow (Haynes 2012, Denyer et al. 2017)
Apply paraffin gel ™ to the lips and teat. Analgesia and/or topical applications such as Gelclair may be required to reduce pain during feeding if blisters are present in the mouth.	A dry teat, may stick to the blistered areas and cause more damage Feeding itself can cause trauma to the mucus membranes (Denyer et al. 2017)
Liaise with the dietician regarding ease of feeding, regurgitation, type of feed and weight.	To ensure adequate nutritional intake of the infant as infants with EB has additional nutritional requirements. These children require a high calorie feed to ensure adequate growth because some of the nutrition is diverted into wound healing (Haynes 2012)
Document care given and evaluate effectiveness of treatment provided.	To facilitate communication, to provide evidence of delivery of quality care, and to ensure evaluation of the effectiveness of care provided (NMBI 2015).

4.0 Care of a Child with Epidermolysis Bullosa during IV Cannulation/Bloods

Introduction

The infant / child with E.B., is at greater risk of skin damage from the application of adhesives material directly on to the skin (Lara-Corrales et al 2010). During treatment, IV Cannulation for medications or bloods may need to be performed. Avoiding skin trauma, is a priority therefore, the procedure should be planned where possible and should be discussed with the Dermatology Team. Care, must be taken during the procedure to avoid skin trauma. (Denyer et al.2017)

Equipment

- Cannulation equipment.
- Blood bottles if required
- EB dressing's e.g. Mepitel, Mepilex transfer, Mepitac tape ®, Tubifast.
- Scissors
- Tourniquet

ACTION	RATIONAL AND REFERENCE
Link with EB CNSp or Dermatology Team if a cannula or bloods are required.	Planning of procedures is important to reduce risk of trauma to skin (Denyer et al. 2017)
Explain the procedure to parents.	To ensure child and parents understands the procedure and gain their trust and co-operation (Trigg and Mohammed 2010)
Pre-medication An oral sedative, such as midazolam may be administered, if clinically indicated. **This is assessed on individual patient needs.**	Sedative premedication, is used in younger children to avoid restlessness, particularly during procedures thus reducing the risk of injury and trauma (Nandi and Howard 2010).
An intravenous cannula, if required, it should be performed with gentle pressure to distend the veins. Sheering forces should be avoided. Direct pressure avoids friction.	Prevention of skin damage due to friction (Denyer 2009.)
A tourniquet can be used over dressings or padding on the arm.	Use over dressing or padding to avoid friction. (Denyer 2009)
Secure the cannula with specific EB dressings. Mepitel, Mepitel Mepilex transfer and or Mepitac are very effective for securing devices such as an intravenous cannula.	To protect the skin from the adhesive of regular dressings as these may cause blistering Silicone wound products/non-adherent dressings prevent trauma to skin. (Nandi and Howard 2010, Denyer et al 2017)

Please be aware the securing tapes used are not as secure as regular tapes. Therefore, care should be taken if moving the patient.	To prevent damage to the skin by using non-adhesive methods of securing cannula to the patient. (Lara-Corrales et al 2010).
Document care given and evaluate effectiveness of treatment provided.	To facilitate communication, to provide evidence of delivery of quality care, and to ensure evaluation of the effectiveness of care provided (NMBI 2015).

5.0 Applying Electrocardiograph (ECG) Electrodes or a Respiration Sensor (MRIO Graseby) on a Child with Epidermolysis Bullosa

Introduction

The infant / child with E.B., is at greater risk of skin damage from the application of adhesives material directly onto the skin (Lara-Corrales et al 2010). ECG electrodes and respiration sensors contain adhesive materials, which can cause friction to the skin (Nandi and Howard 2010) and trauma on removal. Avoiding skin trauma is a priority therefore, the introduction of monitoring equipment, should always be discussed with the Dermatology Team. Care must be taken during the application and removal of such monitors (Denyer 2010)

Equipment

- Mepitac tape ®
- Scissors,
- ECG electrodes / respiration sensor.
- Adhesive spray remover/paraffin gel.
- Mepitel®

ACTION	RATIONAL AND REFERENCE
Cut a hole in the Mepitel® to correspond with the desired positions on the chest to fit (a) the size of the gel part of each ECG electrode or (b) the size of the respiration sensor **NB The Mepitel® will be easier to cut when the backing is still in place.**	To prevent damage to the skin by using non-adhesive methods of securing electrodes to the patient. (Lara-Corrales et al 2010).
Apply the Mepitel® to the chest. Attach the electrodes / sensor to the Mepitel® pushing the gel part of the electrode through the hole in the dressing, ensuring that none of the adhesive is in contact with the patient's skin. Secure with mepitac tape if necessary	Prevention of skin damage due to friction (Denyer 2009.)

When an electrode/ sensor needs to be replaced, leave the Mepitel® in place, unless clinically indicated. Remove the used electrode / sensor and replace it with a new one. When monitoring is completed, gently remove the Mepitel® with the electrodes / sensor from the skin. **If electrodes or a sensor are placed directly on the skin in error or in an emergency situation use an adhesive remover to remove same or paraffin gel.**	Ease of adhesive removal prevents further skin trauma and reduces blistering episodes(Lara-Corrales et al 2010)
Document care given and evaluate effectiveness of treatment provided.	To facilitate communication, to provide evidence of delivery of quality care, and to ensure evaluation of the effectiveness of care provided (*NMBI 2015).*

6.0 Bathing or Washing an Infant with Epidermolysis Bullosa

This Guideline, should be used in conjunction with CHI (2020) Guideline on bathing an infant (under 1yr)

Introduction

Although bathing is considered to be a normal activity of daily living it is not an easy task for a patient with EB *(Arbuckle 2010).*The infant with EB is at greater risk of skin damage when unclothed from friction e.g. kicking of legs/rubbing of arms and from adhering to certain surfaces e.g. soft plastic (Denyer 2014). These surfaces can be temporarily covered with cling film as it only adheres to itself but not to skin (Nandi and Howard 2010). Bathing should be delayed until initial birth trauma is healed (Denyer 2010). Health care professionals who come in contact with children with EB should be aware of the general management of the condition and of the necessary techniques to minimise trauma and complications (Denyer et al 2017)

Indications

When bathing infants with EB it is necessary to meet the infants hygiene needs (Trigg and Mohammed 2010), and to prevent infection and facilitate dressing removal (Arbuckle 2010).

Complications

Trauma to skin and further blistering episodes caused by friction (Denyer et al. 2017)

Equipment

- Bath
- Disposable gloves and apron
- Soft padding (for base of bath)
- Soft cotton cloth or gauze for washing.
- Soft towels 3-4
- Emollient for bath e.g. Elave® ,Oilatum plus®
- Non-perfumed shampoo

- Cream / ointment for buttocks / perineal area e.g. paraffin gel
- Fresh nappy and nappy liners
- Rounded end nail scissors / clippers (if required)
- Fresh clothes
- Weighing scales (if required)
- Cotton wool
- 2 Sheets, soft blanket

ACTION	RATIONAL AND REFERENCE
Pain to be assessed prior to bathing using a validated age appropriate pain assessment scale. Analgesia is administered as prescribed.	EB is a very painful condition (Goldschneider and Lucky 2010, Goldschneider et.al 2014,)The role of the nurse is to assess patients pain level, administer analgesia and assess effectiveness of same (NMBI 2007)
Medications are administered in accordance with the hospital medication policy	To ensure safe administration of medications (NMBI 2007, OLCHC 2017a)
Ensure the nurse/carer who will bathe the infant is familiar with the correct way to handle an infant with EB.	The infant is most at risk of trauma when undressed (Denyer 2010)
Two people should be present: one to bath the infant and one to prevent the infant from causing trauma to skin from kicking feet together.	To reduce the risk of trauma to the skin during the bath.
Explain procedure to parents and encourage involvement.Ensure privacy for the infant and parents throughout the treatment.	To ensure child and parents understands the procedure and gain their trust and co-operation (Trigg and Mohammed 2010)
Prepare the environment by gathering the required equipment	Prepare the environment (Trigg and Mohammed 2010) To prevent cross infection (CHI 2019, CHI 2020)
Line the bottom of the bath with some soft padding e.g. a soft towel or a piece cut from a roll of Melolin	Prevention of discomfort and to prevent trauma and blistering episodes(Denyer 2010,Denyer 2014)
Check bath is at correct temperature not too hot, and add emollient to same	
Use an Emollient or antiseptic in the bath as prescribed.	
Hair Washing When tilting the infant's head back to wash the hair place soft padding between the back of the infant's	To Prevent skin trauma (Denyer 2010)

neck and the hand of the person bathing them, to prevent friction.	
Wash the infant using a dabbing method rather than 'wiping' method, then dry the infant on a soft, non-adherent surface using a soft towel. Pat gently with a soft towel to dry.	Prevention of skin damage due to friction (Denyer 2014.)
Document care given and evaluate effectiveness of treatment provided.	To facilitate communication, to provide evidence of delivery of quality care, and to ensure evaluation of the effectiveness of care provided (NMBI 2015).

7.0 Caring for a child with Epidermolysis Bullosa requiring a general anaesthetic for a procedure.

Introduction

Health Care Professionals, who come in contact with children with EB, should be aware of the general management of the condition and of the necessary techniques to minimise trauma and complications (Nandi and Howard 2010). When a child with EB is going to theatre or requiring a general anaesthetic, rigorous planning is required to avoid risk of trauma to the skin and mucus membranes (Denyer et al.2017)

Common Surgical Procedures in Children with EB

- Change of Dressing
- Repair of Pseudosyndactyly / Surgery to contractures
- Oesophagoscopy and Dilatation
- Open Gastrostomy
- Insertion of Intravenous Access (portacath)
- Ophthalmic Surgery
- Skin Biopsy
- Nissen's Fundoplication
- Excision of Squamous Cell Carcinoma and Skin Grafting (rarely for children)
- Dental Treatments
- General surgery

Complications associated with epidermolysis bullosa

- Fluid loss
- Pain
- Heat loss
- Severe pruritus
- Bacterial infection
- Management of a difficult airway, Difficult intubation due to microstomia and tongue tethering
- Skin trauma resulting in blistering

Theatre Equipment

- Full range of airway management equipment, including fibro scope, guedel airway and LMA
- A range of appropriate dressings, including Mepitac®, Mepilex Transfer®, Mepitel, Vaseline gauze,Episil®
- Melolin® Roll
- Crepe and tubular bandages, Tubifast®
- Cling film
- Clip on oximetry probe
- Foam padding for pressure areas
- Bipolar/dry pad diathermy
- Petroleum jelly
- Medical adhesive removal spray

N.B Please consult with the EB CNSp and or the Dermatology Team, when a child with EB needs to go to theatre

ACTION	RATIONAL AND REFERENCE
Preoperative Preparation **Psychological preparation** Standard preoperative procedures apply using the following adaptations when dealing with a child with EB. The child and parents, should be prepared as normal for theatre with reassurance that all involved know about their child's condition and how to care for them. Informed consent, should be obtained as per hospital policy.	Effective preparation of children who undergo anaesthesia and surgical procedures is an important factor in reducing the anxiety experienced by the child and the family/carer. (Trigg and Mohammed 2010). Preoperative planning and care are essential, when dealing with a child that has EB (Denyer et al. 2017).
<u>Assessment</u> A thorough history is essential including a full discussion with the family paying particular attention to airway involvement, neck movement, nasal airway patency, laryngeal involvement, mouth opening, and oral contracture formation. Any current medical problems such as oesophageal reflux, anaemia, cardiac and renal complications, should be identified and treated if possible (e.g. Antacids or H2- antagonists for oesophageal reflux). Need for a premedication is assessed.	Subtypes of EB can influence airway management. Children with dystrophic or junctional EB are more likely to have airway management problems, which can result in difficult intubations (Nandi and Howard 2010). To prevent post-operative complications (Nandi and Howard 2010) Premedication can help promote a calm induction reducing risk of trauma to skin and mucus membranes (Denyer et al. 2017)

Physical Preparation The child's identification (ID) band should be applied over a piece of tubifast® or the normal dressing. Ensure the ID band, does not come into contact with the child's skin.	To prevent skin damage due to friction on the skin (Denyer 2010).
A topical local anaesthetic such as Amethocaine may be applied in preparation for an intravenous cannula. Do not use an adhesive dressing over the Amethocaine and care must be taken when wiping away any residue. Cling film can be used as a temporary dressing.	Cling film will adhere to itself and not on the skin of a child with EB (Nandi and Howard 2010)
If adhesive tapes or dressings are used in error, they should be left in place until removal is required, when 50% liquid paraffin/50%white soft petroleum and or an adhesive remover can be used to aid removal.	Never use adhesive material on the skin of a child with EB as trauma will occur upon removal of same. Adhesive tapes or dressings can cause trauma to the skin resulting in blistering episodes. (Atherton and Denyer 2003, Lara-Corrales et al 2010).
Dressings should be removed only when necessary. Clingfilm may be used as a temporary dressing for open areas when dressings are removed in the theatre department.	Cling film will adhere to itself and not on the skin of a child with EB (Nandi and Howard 2010)
Pre-medication An oral sedative such as midazolam may be administered if clinically indicated.	Sedative premedication is used in younger children to avoid restlessness, particularly during inhalation induction thus preventing injury and trauma (Nandi and Howard 2010).
Atropine may be administered at the discretion of the anaesthetist.	This is used as a drying agent to help control airway secretions, as excessive salivation may occur (Nandi and Howard 2010).
Medications are administered, in accordance with the hospital medication policy. Monitor the child as clinically indicated.	To ensure safe administration of medications (NMBI 2007, OLCHC 2017a)
Transportation to theatre and transfer onto the theatre trolley Where possible transport the child to theatre on his/her own bed.	To reduce the number of transfers from one surface to another thus reducing the possibility of skin damage caused by friction (Nandi and Howard 2010).
The theatre trolley, should be covered with melolin roll or other soft padding. The trolley, should then be covered with cling film, ensuring to cover the sides. All sharp edges should be well padded with melolin roll and cling film.	To reduce the risk of skin damage, the trolley and sharp edges are padded. The use of cling film helps to reduce the possibility of the skin adhering to any surface (Nandi and Howard 2010).
A sheet of melolin roll, should be placed directly	To reduce the number of transfers from one surface to

against the child's skin so that the child may subsequently be lifted unto the operating theatre on this.	another thus reducing the possibility of skin damage caused by friction (Nandi and Howard 2010).
Patient transfer aids, which involve sliding and sheering forces, should be avoided. Use a "lift and place" method of positioning the child. When it is necessary to lift an infant or small child, roll the child away from you onto his/her side, place a hand underneath the head and neck and a hand under the buttocks, allow the child to roll onto your hands and lift.	Sliding and sheering forces cause damage to the skin (Nandi and Howard 2010,Snelson and Clapham 2010) Correct lifting techniques prevent skin trauma (Denyer 2010).
Theatre Preparations Remove all equipment that should be avoided with the child with EB from the operating room e.g. Elastoplast, self-adhesive items such as, oximetry probes, ECG electrodes, diathermy pads and tempadot thermometers. Ensure all equipment coming in contact with the child is well padded or soft and in the case of anaesthetic equipment, well lubricated with materials such as Vaseline gauze or petroleum jelly. Ensure appropriate equipment, as listed previously, is readily available.	To avoid skin trauma and friction to the skin and mucous membranes. Inappropriate equipment should be removed, if possible from the environment to reduce the risk of skin damage (Nandi and Howard 2010). Sheering forces can cause skin damage. Mepitel, Mepilex and mepitac have adhesive properties to hold an intravenous cannula securely, but, in contrast to adherent tapes, removal does not cause damage to the skin (Denyer 2010, Mortell and Azizkhan 2010, Nandi and Howard 2010).
Anaesthesia An intravenous cannula if required it should be performed with gentle pressure to distend the veins. Sheering forces should be avoided. Mepitel, Mepitel one, Velafilm or Mepitac are very effective for securing devices such as intravenous cannula. Non-adhesive elasticated netting and conforming bandages may also be useful to secure cannulae. All equipment coming in contact with skin should be lubricated i.e. masks, gloves and instruments. The anaesthetic facemask should be covered with Vaseline gauze and mepitel should be placed against the child's skin, where the underside of the jaw is held by the anaesthetist and any other area of the child's skin where pressure is likely to be needed. Endotracheal tube should be one size smaller than would be normally used and the tube and laryngoscope blade should be well lubricated with KY gel prior to intubation. The tube should be held	Prevention of skin damage due to friction (Denyer 2009.) Holding the child's skin can cause major damage around the mandible and mouth. This will protect the child's skin from trauma (Mortell and Azizkhan 2010) To avoid trauma to mucus membrane and to avoid friction which may damage the skin (Mortell and Azizkhan 2010, Nandi and Howard 2010)

in place with ribbon gauze or mepitac tape, all tubing should be padded and Vaseline gauze should be applied where it comes in contact with skin.	
Suction and the use of oropharyngeal airways, should be avoided, if possible although in some cases this is unavoidable. Pharyngeal suctioning should be performed under direct vision, taking care not to apply the suction catheter to the mucosa directly.	To avoid trauma to mucus membrane and to avoid friction which may damage the *skin* (Mortell and Azizkhan 2010, Nandi and Howard 2010) This may cause serious mucosal detachment and blistering (Nandi and Howard 2010).
The eyes, should be carefully closed and covered with Vaseline gauze and not adhesive material. Mepitac tape or gel pads can be used to keep eyes closed.	To prevent corneal abrasions and to protect the eyes (Mortell and Azizkhan 2010)
Nursing Observations A non-adhesive pulse oximetry probe of the 'clip-on' variety, should be placed onto the ear lobe.	Direct pressure will not cause damage to the skin(Nandi and Howard 2010)
ECG electrodes should be adapted using Mepitel or Mepilex transfer directly on the skin, underneath the adhesive and securing with mepitac. **NB Do not use tempadot use a digital thermometer to record temperature.**	To protect the skin from the adhesive of the electrodes as this may cause blistering (Nandi and Howard 2010) To prevent any skin trauma and further blistering(Nandi and Howard 2010)
Non-adherent soft gauze padding such as melolin roll, should be placed around the child's arm prior to application of blood pressure cuffs or tourniquets.	Blood pressure cuffs and tourniquets can exert sheering forces and cause skin blistering (Mortell and Azizkhan 2010, Denyer et al.2017).
<u>Skin care</u> Skin is prepared with antiseptic solution as normal for surgery but avoid rubbing. Use patting motions.	Rubbing the skin is to be avoided as this is a severe cause of blistering (Nandi and Howard 2010).
The appropriate dressings should be used for surgical wound sites post-surgery. All dressings normally worn by the child should be left in situ and they should only be removed when clinically indicated.	Recommended dressings are non-adherent and dressings left in situ will protect the child's skin (Denyer 2010).
<u>Post-operatively</u> Postoperative care is carried out as appropriate for the surgical procedure.	To ensure safe recovery from procedure and anaesthetic (Trigg and Mohammed 2010).
A general skin assessment should be performed to document any new blistering episodes. Temporary dressings or open wounds should be re-dressed with the appropriate dressings.	All care is undertaken to ensure minimal blister formation and unnecessary trauma However trauma can occur if child is unsettled or in pain(Nandi and Howard 2010)

Pain is assessed using a validated, age appropriate pain assessment scale and analgesia administered. Medications are administered in accordance with the hospital medication policy. Non pharmacological methods i.e. adequate preparation and involving play specialist can also help with pain.	Effective postoperative analgesia is essential to minimise discomfort and skin trauma (Nandi and Howard 2010). To ensure patient is pain free and reduce anxiety of parents and child. To reduce the risk of skin damage (Goldschneider and Lucky 2010).
N.B The rectal route, frequently used in paediatric postoperative pain management, should be avoided.	Risk of trauma to the mucus membranes of the anus (Snelson and Clapham 2010)
On return to the ward, the child should be transferred safely from the trolley to the bed on the melolin sheet. Should the child need to be lifted, please refer to the correct lifting technique above.	Sliding and sheering forces cause damage to the skin (Nandi and Howard 2010 ,Snelson and Clapham 2010)
Postoperative observations should be carried out as clinically indicated using the techniques outlined above and by referring to the" Guidelines on the care of a new baby with EB on admission".	All care is undertaken, to ensure minimal blister formation and unnecessary trauma (Nandi and Howard 2010).
Document care given and evaluate effectiveness of treatment provided.	To facilitate communication, to provide evidence of delivery of quality care, and to ensure evaluation of the effectiveness of care provided (NMBI 2015).

REFERENCES

Ahn, C and Salcido R.S. (2008) Advances in Wound Photography and assessment methods. ***Advances in Skin Wound Care,*** 21(2) 85-96.

Arbuckle A. (2010) Bathing for individuals with Epidermolysis Bullosa In: Thiers B.H. (eds) *Dermatolologic clinics Epidermolysis Bullosa: part II- Diagnosis and Management*. .WB Saunders Company. United States of America.

Atherton DJ.and Denyer J (2003) *Epidermolysis Bullosa. An Outline for Professionals*. Debra Publication, Crowthorne Berks.

Children's Health Ireland (2019) *Standard Universal Precautions.* CHI, Crumlin Dublin.

Children's Health Ireland (2019) *Safety Statement*. CHI, Crumlin, Dublin.

Children's Health Ireland (2020) *Guideline on Isolation.* CHI, Crumlin, Dublin.

Danial C. Adeduntan R. Gorell E.S. Lucky A.W. Paller A.S. Bruckner A Pope E. Morel K.D. Levy M.L. Shufeng L. Gilmore E.S. and Lane A.T. (2014) Prevalence and Characterization of Pruritus in Epidermolysis Bullosa. *Pediatric Dermatology*, 1-7

Denyer J (2010) *Wound Management for Children with Epidermolysis Bullosa* In; Thiers B.H. (eds) Dermatolologic Clinics Epidermolysis Bullosa: part II- Diagnosis and Management. WB Saunders Company. United States of America

Denyer J (2014) *The New baby with EB. Information for Parents*. DebRA Publication, Crowthorne Berks

Denyer, J Pillay E, Clapham J. (2017) Best Practice Guidelines Skin And Wound care in Epidermolysis Bullosa.International Consensus. Wounds International.

Goldschneider K.R and Lucky A (2010) *Pain Management in Epidermolysis Bullosa* In; Thiers B.H. (eds) Dermatolologic Clinics Epidermolysis Bullosa: part II- Diagnosis and Management. WB Saunders Company. United States of America

Goldschneider, K.R Good., Harrop, J, Liossi4, C, Lynch-Jordan. A, Martinez.A.E, Lynne, K.R, Maxwell. G, and Stanko-Lopp. D, (2014) BMC Medicine, 12:178 Page 2 of 23. Accessed on 08/04/2020 http://www.biomedcentral.com/1741-7015/12/178

Haynes L (2010) *Nutrition for Children with Epidermolysis Bullosa* In; Thiers B.H. eds Dermatolologic clinics Epidermolysis Bullosa: part II- Diagnosis and Management. WB Saunders Company. United States of America.

Himsworth, J (2010) *Caring for Personal Hygiene Needs* .In: Glasper, A. Aylott, M. Battrick C (eds) Developing Practical Skills for Nursing Sick Children and young people .Hodder Arnold, London 188-202.

Lara-Corrales I. Arbuckle A. Zarinehbaf and S Pope E. (2010) Principles of Wound Care in Patients with Epidermolysis Bullosa *Pediatric Dermatology*, **27**(3) 229-237.

Mellerio J.E (2010) *Infection and colonization* In Epidermolysis Bullosa Thiers B.H. (Eds) Dermatologic Clinics Epidermolysis Bullosa: part II- Diagnosis and Management. WB Saunders Company. United States of America

Mortell A.E and Azizkhan RG (2010) *Epidermolysis Bullosa: Management of esophageal strictures and Enteric Access by Gastromy* In; Thiers B.H. (eds) Dermatolologic Clinics Epidermolysis Bullosa: part II- Diagnosis and Management. WB Saunders Company, United States of America

Nandi R and Howard R. (2010) *Anaesthesia and Epidermolysis* Bullosa In; Thiers B.H. (eds) Dermatolologic clinics Epidermolysis Bullosa: part II- Diagnosis and Management. WB Saunders Company. United States of America

Nursing and Midwifery Board of Ireland (2007) *Guidance to Nurses and Midwives on Medication Management.* NMBI, Dublin

Nursing and Midwifery Board of Ireland (2015*) Recording Clinical Practice: Professional Guidance to Nurses and Midwives.* NMBI, Dublin.

Our Lady's Children's Hospital (2015) *Guideline on bathing an infant under 1* .OLCHC Crumlin Dublin

Our Lady's Children's Hospital (2017a) *Medication Policy*. OLCHC, Crumlin, Dublin

Our Lady's Children's Hospital (2017b) *Guideline for Hand Hygiene,* OLCHC, Crumlin, Dublin.

Our Lady's Children's Hospital (2018) *Guidelines on Performing a Wound Swab.* OLCHC Crumlin, Dublin.

Snelson K and Clapham J (2010) *Guidelines for the practical care of Adult Patients with Epidermolysis Bullosa during Surgical Procedures*. Debra Publication, Crowthorne Berks.

Stalder JF (2011) Skin Care of the Newborn In: Harper J, Hoeger P, Irvine A.D, Yan A.C (eds) *Textbook of Pediatric Dermatology* , 3rd edition vol.(**1)**pgs 5.1-5.8. Blackwell Publishing Ltd.

Trigg, E., and Mohammed, T (2010) *Practices in Children's Nursing; Guidelines for Hospital and Community,* Churchill Livingstone, London.