

Emergency Management Steps for Epidermolysis Bullosa

CAUTION: The skin is extremely FRAGILE

Epidermolysis Bullosa (EB) is a rare genetic **SKIN condition** characterised by fragile skin and body linings (mucosae).

- Minimal trauma (friction or knocks) can easily cause **blistering** of the skin and mucosal surfaces, these can lead to erosions and **wounds**. This can increase susceptibility to **infections**.
- **Pain** is constant from birth and can worsen during procedures.
- Most **emergency procedures** can be performed BUT **specific precautions must be taken**.
- **Analgesic and anxiolytic medication** can be administered.

Specific precautions with general procedures:

- **Never slide to transfer:** use a 'lift and place' approach.
- **Avoid sticky dressings and tapes:** whenever possible.
- **Avoid skin rubbing or stroking:** pat with-gentle pressure applied.
- **Avoid friction:** when handling, undressing, or dressing.

Recommended care measures are:

- **Listen to the patient**
- **Skin care and protection**
 - Pat or dab when cleaning or drying the skin, avoid rubbing or stroking the skin.
 - Lance any blisters using a finger prick lancet, hypodermic needle or blade, and gently drain without removing the blister roof.
 - Place identification (ID) bracelets over a protective dressing or bandage.
 - Pad trauma-prone sites
 - for example, pad with 2-3 layers of soft padding (e.g. Softbank) before applying the blood pressure cuff or tourniquet.
 - Alternately, use gentle pressure to distend veins by hand and aid cannula insertion.
 - If removal of dressings is unavoidable, cling film may be used as a temporary covering to the skin.
 - Allow the patient to remove the dressing after the procedure if possible
 - Transfer using "lift and place" technique – never slide or use 'Pat Slides' (is this a sliding board).
 - Minimise the number of transfers.
 - Use of HoverMatt® is highly recommended for all lateral transfers.
- **Care with sticky materials**
 - Avoid adherent dressings where possible.
 - Ensure the safe removal of any sticky dressing, tape or monitoring stickers that may be inadvertently applied.
 - Use Medical Adhesive Removal Spray (MARS) such as silicone-based spray to remove any sticky dressings (examples are: Appeel® or Niltac® (or 50/50 liquid paraffin/white soft paraffin))



- If silicone-based spray is not available, soak dressings with lukewarm water.
- Remove adhesives from electrocardiogram pads and secure with non-adhesive dressings such as Mepitel One.
- Never tape the eyelids
 - Instead close gently and apply simple eye ointment to prevent the eyes drying out.
 - If the eyes are excessively dry, or the eyelids are scarred, artificial tears may be used. Don't open the eyes; apply artificial tears drops, gels, or ointments on eyelashes; and allow the person to stay in a dark room.
 - Use Mepitel One if needed
- **Lubricate, lubricate, lubricate**
 - Gloved hands in contact with the skin can cause damage to fragile skin. Gloves should be lubricated if practical.
 - Lubricate all devices (thermometer, cannulas, catheters, feeding tubes, etc.)
 - Use lubricated clip sensors for pulse oximetry.
- **Selecting devices**
 - Use smallest effective sizes for catheters and tubes.

The following table summarises some emergency management steps for some of the most common EB related events which can be carried out prior to hospitalization with EB. Columns are ordered in descending order of Emergency response (from left to right).

	Acute upper airway obstruction	Sepsis	Corneal erosions	Acute feeding inability in new-borns /infants	Acute oesophageal obstruction	Acute urinary retention
Emergency diagnosis	<p><i>Clinical history</i></p> <ul style="list-style-type: none"> • Hoarseness • Episodes of respiratory stridor 	<p><i>Clinical history</i></p> <ul style="list-style-type: none"> • Feeling unwell • Change in behavior, functioning and/ or consciousness. • ↓ urine output the previous 12-24 hours. • Any risk factors for infection noted. 	<p><i>Clinical history</i></p> <ul style="list-style-type: none"> • Pain, inability to open the eye, photophobia, and excessive tearing. • There may or may not be a history of minor preceding trauma. 	<p><i>Clinical history</i></p> <ul style="list-style-type: none"> • Sudden uncontrollable crying. • Feeding refusal. 	<p><i>Clinical history</i></p> <p>Frequently onset is during a meal, following ingestion of a large or traumatic bolus of food.</p>	<p><i>Clinical history</i></p> <ul style="list-style-type: none"> • Inability to pass urine, dry diapers in infants, abdominal distension, and discomfort. • There may be a history of difficulty starting urination and reduced urinary flow, a deflected stream or of blistering around the urethral meatus.
	<p><i>Clinical features</i></p> <ul style="list-style-type: none"> • Prominent inspiratory stridor with suprasternal and sternal wall retraction, worsened by crying • Shortness of breath, agitation, and distress • Pale to dusky complexion 	<p><i>Clinical scoring (age appropriate) to assess an individual's risk of sepsis:</i></p> <ul style="list-style-type: none"> • Unwell, confused, altered conscious. • ↑ respiratory rate • Hypotension • Tachycardia or bradycardia • Temperature normal, low, or raised • ↓ oxygen saturations • Dusky, mottled skin changes 	<p><i>Clinical features</i></p> <ul style="list-style-type: none"> • Blepharospasm • Excessive tearing • Redness of the eye • Blurred vision • Corneal defect(s) visible on fluorescein slit lamp examination 	<p><i>Clinical features</i></p> <ul style="list-style-type: none"> • Sialorrhea (drooling or excessive salivation) • Presence of one or more large tense blisters in the oral cavity 	<p><i>Clinical features</i></p> <ul style="list-style-type: none"> • Acute complete/almost complete inability to swallow solids or both solids and liquids • Acute painful dysphagia • Onset of a severe sialorrhea (drooling or excessive salivation) or worsening of a pre-existing sialorrhea • Regurgitation 	<p><i>Clinical features</i></p> <ul style="list-style-type: none"> • Enlarged, tender bladder on abdominal palpation • Blistering around urethral meatus, meatal stenosis, labial fusion (females)
ACTION	Emergency call 📞	Emergency call 📞	Go to the hospital *	If trained try at home or if NOT go to the hospital*	After 2-3 days if inadequate fluid intake or untreatable pain go to the hospital *	Referral to hospital *
Who	First aid by emergency service /Paramedics	Local doctor or paramedics	Local doctor, trained parents/ caregivers	Local doctor, trained parents/ caregivers; EB nurse specialist;	Local doctor; EB nurse specialist;	Local doctor

Treatment	<ol style="list-style-type: none"> 1. Proper airway management by positioning via the head tilt-chin lift manoeuvre 2. Administer oxygen therapy, secure an IV line and check vital signs. 3. Non-invasive ventilation by bag-valve mask 4. Invasive airway management via intubation (nasal or endotracheal), or emergency tracheostomy 5. Immediate hospitalization. 	<ul style="list-style-type: none"> • Oxygen therapy by emergency services if saturations < 90% on air, aiming for saturations of 94–98% (or 88–90% if at risk of hypercapnic respiratory failure) • Establishment of peripheral IV access by emergency services if possible 	<ul style="list-style-type: none"> • Frequent application of preservative-free artificial tear drops, gels, or lubricant ointments • When applying artificial tears drops, gels, or ointments: do not open eyes, apply on eyelashes; and allow the person to stay in a dark room. <p><i>Adequate analgesia</i></p> <ul style="list-style-type: none"> • Paracetamol oral solution 15mg/kg/3–4 times a day • Tramadol hydrochloride 1 mg/kg every 6 hours. • If not effective, morphine oral solution 0.2–0.3 mg/kg every 4 hours, with doses increased by 30% if necessary. 	<p><i>Analgesic therapy</i></p> <ul style="list-style-type: none"> • For the nociceptive pain: paracetamol oral solution 15mg/kg, which can be repeated up to 4 times/day • For the more severe pain: tramadol hydrochloride 1 mg/kg every 6 hours • Blister lancing using a finger prick lancet or a hypodermic needle or, if not available, a sterilized sewing needle 	<ul style="list-style-type: none"> • Oral betamethasone 0.1–0.2 mg/kg/day • In case of complete swallowing inability, dexamethasone sodium phosphate 0.2% drops 1 mg/kg for up to 2–3 days. • If neither are available an equivalent dosage of soluble prednisolone can be administered. • Corticosteroid therapy should be accompanied by administration of oral sodium alginate and sodium bicarbonate solution or a proton pump inhibitor. • For complete aphagia (inability or refusal to swallow), an equivalent dose of corticosteroid should be administered IV. <p><i>Analgesic therapy if needed</i></p> <ul style="list-style-type: none"> • paracetamol oral solution (15 mg/kg 3–4 times/day) and tramadol hydrochloride 1mg/kg every 6 hours • if not effective, morphine oral solution 0.2–0.3 mg/kg 5–6 times a day, with doses increased by 30% if necessary. • Switch to liquid or semi-liquid nutrition, preferably cold 	
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ACTION				Emergency call 📞		
				<ul style="list-style-type: none"> • If treatment at home is not recommended or not effective (failure to drain the blister or immediate relapse, or new-born/infant still being agitated and crying): management at hospital. 	<ul style="list-style-type: none"> • In case of persistent inability to swallow in infants, lack of improvement within 2–3 days in children and adults, complete dysphagia with inadequate fluid intake or untreatable pain: hospitalization. • Refer to specialised dietitian 	

KEY: * If you are linked or close to an EB clinical centre, go to that hospital; ↑ HIGH, ↓ LOW; mg: milligrams; kg: kilograms; IV: intravenous;

At the hospital

For emergency management steps of above events at the hospital site or EB Centre.

See the full “Emergency management in epidermolysis bullosa: consensus clinical recommendations from the European reference network for rare skin diseases” Mellerio et al 2020

[<https://ern-skin.eu/wp-content/uploads/2020/08/Emergency-management-in-EB.pdf>]

To understand and know more about EB go to WHAT IS EB?

[<https://www.debra-international.org/what-is-eb>]

References:

- Mellerio J E et al 2020
- Clapham J, Snelson K, et al. Epidermolysis Bullosa – care of adult patients during clinical and surgical procedures. Guy's and St Thomas' Hospital NHS. Guidance 2019
- Greenblatt D et al 2021 [<https://www.debra-international.org/pregnancy-childbirth-aftercare-cpg>]



This was update in February 2022