

Caring for Pediatric Patients With Epidermolysis Bullosa in the Emergency Department

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Abstract: Epidermolysis bullosa (EB) refers to a heterogeneous group of genetic disorders characterized by epithelial fragility. We provide guidelines for management of pediatric patients with EB in the emergency department based on a review of literature, as well as insights from our own experiences caring for patients with EB. The purpose of the guidelines proposed is prevention of avoidable iatrogenic trauma to the skin and mucosa of patients with EB who are presenting to the emergency department for a variety of reasons.

Key Words: epidermolysis bullosa, iatrogenic trauma, mucosa, skin, wound care, immunobullous disease, health care delivery

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Epidermolysis bullosa (EB) refers to a heterogeneous group of genetic disorders characterized by epithelial fragility. Characteristically, the skin and in some cases the mucosa of patients with this condition form bullae and/or erosions in response to minimal trauma (Fig. 1). Because of resultant disruptions of the skin barrier, these patients are at increased risk of poor wound healing and infection.¹ Patients may develop numerous long-term sequelae of chronic wounds and scarring including joint contractures, esophageal strictures, pseudosyndactyly, and aggressive cutaneous squamous cell carcinomas.² Common reasons for patients with EB to seek emergency care include pain, wound infections, and feeding difficulties, but they may also present with the same non-disease-specific complaints as the general pediatric population.

Based on data from the National Epidermolysis Bullosa Registry, prevalence and incidence of EB per 1 million live births are 11.07 and 19.57, respectively.³ Epidermolysis bullosa is caused by mutations in at least 18 genes encoding structural proteins involved with adhesion of epidermis and dermis.^{4,5} Epidermolysis bullosa has been divided into 4 main subtypes based on the level of skin cleavage; these are EB simplex, junctional EB, dystrophic EB, and Kindler syndrome (Fig. 2).⁶ Epidermolysis bullosa simplex involves intraepidermal blistering, most commonly due to mutations of keratins 5 and 14.^{5,7} Junctional EB involves cleavage at the lamina lucida level due to defects in a variety of basement membrane zone proteins, most commonly the components of laminin 332.^{5,8} Dystrophic EB involves blistering just below the lamina densa secondary to defects in collagen VII.^{5,9} Kindler syndrome has variable levels of involvement.⁵ The specific manifestations and severity of individuals with a given subtype are variable. However, the information in this report will be most relevant to patients with dystrophic EB, in which the level of cleavage is deeper and thus involves a greater thickness of skin damage.

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In this report, we provide guidelines for management of pediatric patients with EB in the emergency department (ED) based on a review of literature, as well as insights from our own experiences consulting on patients with EB. There are few trials in this patient population relevant to this topic; thus, much of the data consist of providers' experiences and observations. The guidelines below are not meant to address specific reasons for patients seeking emergency care, but rather general considerations for care of EB patients with moderate to severe disease presenting for a variety of reasons.

GUIDELINES

The purpose of these guidelines is to provide education regarding the pathophysiology of EB and to describe the risks that this vulnerable population faces with standard care. It is also to provide concrete suggestions regarding prevention of iatrogenic trauma to the skin and mucosa of patients with EB, who are susceptible to extensive cutaneous injury from routine interventions, such as transferring a patient from one surface to another, line placement, and monitoring of vital signs. Of course, life-sustaining measures take precedence over protection of skin and mucosa in the management of these patients.

Approach to Patients With EB

The approach to management of a patient with EB will differ based on subtype and severity of disease as well as comorbidities. It is often beneficial to reach out to patients' primary care physicians and dermatologists early on when planning care. Organizing family meetings to determine how to handle more frequently encountered issues and avoid complications may additionally be beneficial for patients who frequently visit the ED. On a larger scale, we encourage development of institutional protocols regarding care of these patients that address such issues as wound dressings, managing infections, and managing pain in order to prevent medical errors and ease timely provision of optimal care.

Bedding, Transfers, and Patient Handling

The handling and transferring of EB patients are to be minimized. If handling is required, use of gloves lubricated with petrolatum jelly or an aqueous lubricant may help to prevent skin injury. These patients are particularly vulnerable to friction and shearing forces, whereas compressive forces are better tolerated.¹⁰ Thus, use a "lift and place" approach during transfers and never slide patients.⁶ The "lift and place" approach requires 2 or more individuals and is accomplished by firmly grasping on either side the top sheet below the patient and lifting, ensuring that the patient's weight is evenly distributed across the sheet. The patient can then be moved to the desired location and carefully lowered directly to intended site.

Use of electric beds that allow patients old enough to do so to adjust their own positioning is preferred to manual beds.⁶ Pressure-redistributing mattresses, in particular, are recommended for use by these patients.² Egg crate padding may be used for padding other surfaces used for supporting weight, including toilet seats and bed railings.²



FIGURE 1. Complete denudement of the knee in a patient with recessive dystrophic EB. There are no signs of infection.

Adhesives and Adhesive Removal

Use of adhesives is to be avoided in patients with EB as removal may result in significant skin trauma. Unfortunately, this creates numerous challenges in the emergency setting as adhesives are frequently relied upon for securing placement of wound dressings, monitoring devices, and indwelling lines. These specific issues are discussed in greater detail below.

If an adhesive is placed on the skin of a patient with EB, either accidentally or out of necessity, extreme care should be taken in its removal, and medical adhesive remover should be used. Depending on the circumstances and the preferences of patient and family, the adhesive may instead be left in place for patient or family member to remove according to their own established methods. Another alternative is to leave the adhesive in place and allow it fall off by itself.

In settings with limited EB-appropriate supplies, a regular adhesive can be placed on the skin to act as a “second skin” to which other regular adhesives can be applied. This anchoring adhesive should then be removed following the same guidelines discussed previously when no longer needed.

Monitoring Devices and Patient Identification

A first consideration is whether a monitoring device is truly indicated for an individual patient; minimal monitoring has been advocated in patients with EB.¹⁰ A second consideration is the frequency of monitoring that is required, as more frequent monitoring may result in increased skin trauma.

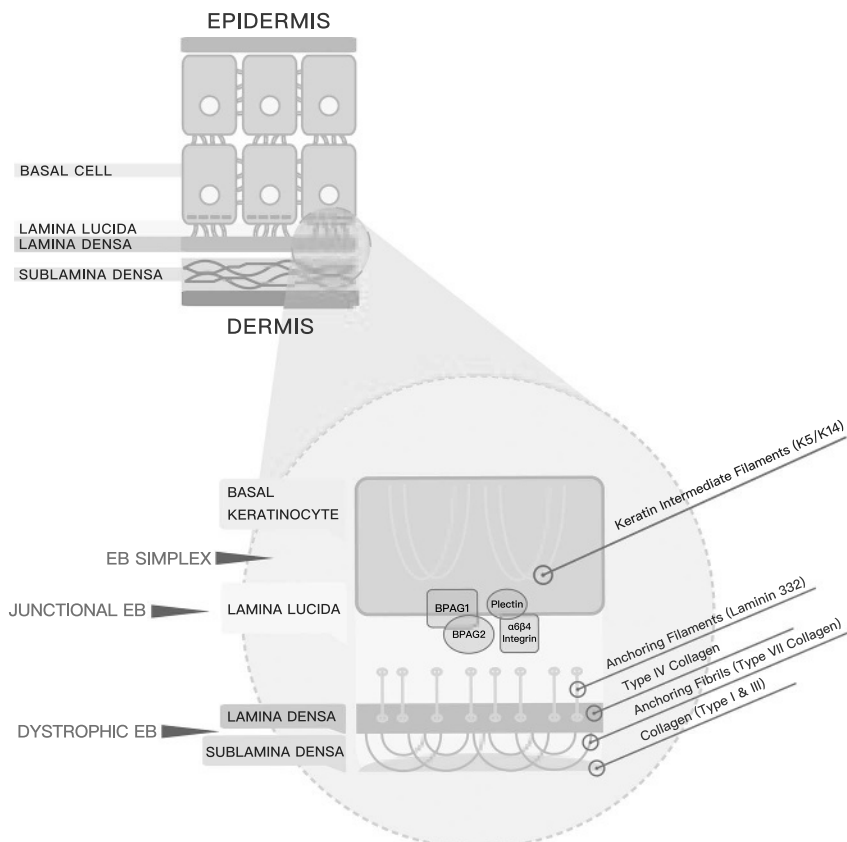


FIGURE 2. Schematic of the dermoepidermal junction zone and the ultrastructural elements of basal keratinocytes, the basement membrane zone, and superficial dermis. Epidermolysis bullosa simplex involves intraepidermal blistering. Junctional EB involves cleavage at the lamina lucida level. Dystrophic EB involves blistering just below the lamina densa.

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Clip-on pulse oximeters applied with lubrication may be used in patients with EB.¹¹ Alternatively, adhesive oximeters may be altered to prevent contact of adhesive with skin. For example, the adhesive surface of an adhesive oximeter may be covered with the adhesive surface of a Tegaderm dressing (3M, St Paul, Minn) (Fig. 3). The nonadhesive side of the Tegaderm dressing may then be secured to skin with a silicone bandage. Alternatively, the adhesive surface may be removed completely, and the sensing device may be applied to skin and secured with a dressing, or a pulse oximeter can be applied directly over white cotton gloves (Fig. 4).⁶ Notably, hand deformities are common in patients with EB, particularly those with recessive dystrophic EB, and it may be necessary to use a patient's palm, digit stump, or ear lobe rather than a finger.^{6,12}

Adhesive electrocardiogram electrodes may be attached to defibrillator gel pads, such as Defib-Pads (3M), rather than directly to skin.¹³ Alternatively, the adhesive may be removed and the lead placed directly on skin, then secured in place with a dressing (Fig. 5). For monitoring temperature, lubricated axillary probes are preferred to oral probes.^{2,11} Nasopharyngeal and rectal measurements of temperatures are to be avoided.¹

Blood pressure cuffs should be placed over multiple layers of soft padding rather than directly on skin. If frequent monitoring will be required, placement of an arterial line should be considered.⁶ Identification wristbands should also be placed over multiple layers of soft padding or simply placed at the patient's bedside.^{2,6}

Wound Dressings

Adhesive dressings should not be used in patients with EB because their removal may result in significant damage to skin. Siliconized dressings are optimal because they are tacky without being true adhesives and can be easily removed with water. If appropriate products are not readily available, Vaseline petrolatum gauze (Medtronic, Dublin, Ireland) can be placed over wounds and kept in place with multiple layers of soft padding. We recommend keeping abundant appropriate wound dressing materials in the patient's room, so these are readily available to staff at all times.

A typical wound dressing will include 4 layers. The first layer is ointment, typically petroleum jelly, AquaPhor (Beiersdorf Inc, Hamburg, Germany), or an antibiotic ointment applied directly to the wound, followed by the second layer, which is in contact with wound, such as Mepitel (Mölnlycke Health Care,



FIGURE 4. The pulse oximeter is placed directly over white cotton gloves.

Gothenburg, Sweden) or Vaseline petrolatum gauze. The role of the third layer is to provide cushioning, often achieved with several layers of stretch rolled gauze, and the fourth layer will secure the dressing in place.² Tubular dressing retainers are commonly used for this purpose. Often patients and their families are a great resource for learning what products have and have not worked for them in the past.

Wound Dressing Changes

Dressing removal may require premedication with analgesics and/or anxiolytics. It is helpful to have all the materials required for the new dressing open and tailored to fit at bedside prior to starting dressing removal. Removal of outer bandages should be done carefully in the event that there are adherent areas. In such a case, soaking affected areas in water or applying a wet compress can loosen bandages to allow for removal.



FIGURE 5. Electrocardiogram leads wrapped in gauze, placed on skin, and secured with a tubular retainer dressing.

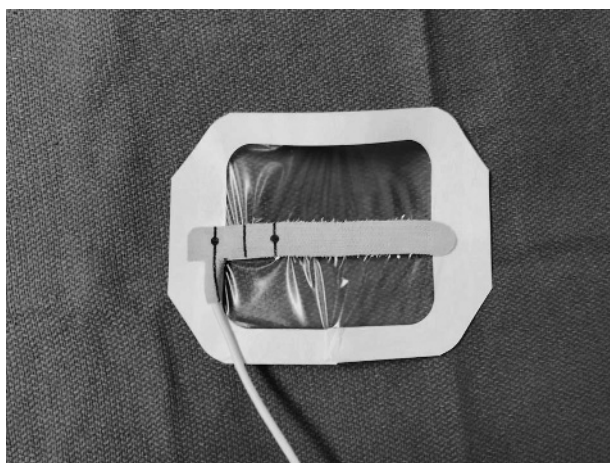


FIGURE 3. The adhesive surface of the Tegaderm is attached to the adhesive surface of the oximeter. The nonadhesive side of the Tegaderm may then be placed against skin and secured with a silicone bandage.

Dressings should be removed and then reapplied 1 limb at a time to avoid further trauma and to prevent wounds from drying out. The skin can be cleansed with a mild, fragrance-free agent; rinsed with water; and then patted dry. Blisters that are tense or greater than a 1 cm in diameter should be sterilely drained with a needle or sterile scissors, and the roof of the blister should be left in place to act as a natural bandage. The wound can then be redressed as described previously.

Not every lesion requires a dressing. Wounds should be dressed if they are exudative or painful, or if they are in areas of high friction. Site-specific dressing options that are engineered specifically for more difficult-to-dress locations, such as ears, interdigital spaces, elbows, axillae, are also available.

Assessing Wounds for Evidence of Infection

Antibiotic therapy is not indicated for patients with wound contamination or colonization. Critical colonization, defined as bioburden sufficient to impair wound healing, is typically treated with topical antibiotics.² A retrospective review of 30 patients indicated that the 4 most common wound colonizers isolated from bacterial cultures in EB patients are *Staphylococcus* species (both *Staphylococcus aureus* and coagulase-negative *Staphylococcus*), *Streptococcus* species, diphtheroids, and *Pseudomonas aeruginosa*.¹⁴ The decision of whether to start systemic antibiotic therapy should be guided by physical examination. Hand washing prior to examining a patient with EB is important to prevent iatrogenic infection.

Vascular Access

Considerations to be made before placement of intravenous (IV) lines in patients with EB include whether the line is truly necessary and how long IV access will be required. If there is reason for prolonged use of IV medications, these patients may benefit from placement of a central rather than a peripheral line, to prevent trauma associated with repeat placement of peripheral IV lines.² Suturing may be used to secure central and arterial lines in place.⁶

Use of tourniquets for placement of IV lines is to be avoided when possible. Gentle pressure from a lubricated hand to distend veins may be attempted instead. If a tourniquet is necessary, padding should be placed beneath it so that it does not make direct contact with skin.¹¹ Peripheral IV lines should be secured using silicone-based rather than adhesive tapes.¹⁵ Based on the complexities of line placement and the potential for harm caused by multiple attempts, it is advisable that lines be placed by hospital staff members with expertise in placement of difficult IV lines.²

For hospitals without on-call anesthesiologists, utilizing a vascular access team or experienced nurse may be necessary, and temporizing measures, such as medication administration and hydration via gastrostomy tube should be considered.

Urinary Catheters

Urinary catheters should be well lubricated prior to insertion; smaller-gauge urinary catheters are preferred.⁶ Epidermolysis bullosa may cause genitourinary complications, including strictures. If there is resistance with placement of catheter or suspicion of urinary anomaly, urology consultation should be considered. Health care providers should pay attention to the position of external tubing so that it does not cause friction against skin, and be mindful that external tubing should not be secured to skin with adhesives.^{5,6}

Enteral Tubes

Many patients with EB require gastrostomies due to chronic feeding difficulties. External tubing should be secured with

silicone tape rather than regular adhesives. For those without gastrostomies, nasogastric (NG) tubes may be required for feeding, decompression, and/or medication administration. Small-gauge, flexible NG tubes should be used for these patients and lubricated liberally prior to insertion.⁶ Clinicians should be aware that patients may have esophageal strictures; thus, tubes should be placed with care and never with excessive force. Gastroenterology consultation may be required if there is suspicion of stricture preventing feeding or enteral tube placement.

Pain and Anxiety

As in other pediatric patients, acute, mild pain may be managed with acetaminophen or ibuprofen, and moderate to severe pain may be managed with opiates administered orally, intravenously, or via gastric tube.^{2,16} Important considerations regarding opiate use in patients with EB are that these medications promote pruritus and constipation, 2 issues that frequently affect this patient population.¹⁶ Topical morphine and lidocaine gel or cream may also be helpful.¹⁶ Anxiolytics may be considered for treatment before certain anxiety-inducing activities such as dressing changes or procedures.¹⁶

Pruritus

Patients with EB often suffer from pruritus. In addition to causing patient discomfort, pruritus may lead to scratching and subsequent blister and erosion formation.¹⁶ Use of emollients may be effective in controlling pruritus in some patients; however, many will find additional benefit from use of antihistamines.¹⁶ Topical corticosteroids may be useful as well, but clinicians should be aware of the possibility of increased systemic absorption due to impairment of skin barrier in these patients.² Heat and sweating may exacerbate pruritus, so temperature of patient's environment should also be considered in managing pruritus.¹⁶ Gabapentin has not been well studied in EB, although some providers report better control of patient's discomfort with this medication.¹⁷

Medication Administration

We prefer administration of medications via the oral route (or via gastric tube, if patient has one) unless there is a specific indication for delivery of medication via the IV route. Rectal administration of medications is relatively contraindicated in patients with EB because of concerns of perianal trauma.¹

Procedures

There is no specific contraindication to the use of usual topical antiseptics in most EB patients. When preparing skin for procedures, cleansing agents, such as povidone-iodine or chlorhexidine gluconate solutions, should be poured or sprayed onto skin and then patted dry, or a cleansing swab may be applied with gentle downward pressure and then removed.^{1,6} It is important to avoid rubbing or stroking the skin.^{6,11} Clinicians should avoid leaning against or resting objects on patients during procedures.⁶ Adhesives should not be used to secure drapes to skin; towel clips may be used to secure drape position instead.⁶

Oxygenation and Intubation

Nasal cannulas should be lubricated prior to placement. Plastic air masks with sharp edges should be avoided; however, anesthesia face masks may be used when lubricated or lined with paraffin gauze.¹

Intubation may be difficult in patients with EB for a number of reasons. Patients with EB may have microstomia and ankyloglossia

TABLE 1. Emergency Department Management of Patients with EB

Transfers and patient handling	<ul style="list-style-type: none"> • Minimize transfers • Use lubricated gloves when handling patient's skin • Use "lift and place" approach during transfer, never slide • Utilize pressure redistributing mattresses and/or egg crate padding
Use of adhesives	<ul style="list-style-type: none"> • Avoid use of all adhesives
Adhesive removal	<ul style="list-style-type: none"> • Use medical adhesive removal spray and remove with extreme caution
Monitoring devices	<ul style="list-style-type: none"> • Assess if truly necessary • Use clip-on pulse oximeter with lubrication • Place pulse oximeter over white cotton gloves • Remove adhesives from electrocardiogram leads or adhesive pulse oximeter and secure with nonadhesive dressings
Patient identification	<ul style="list-style-type: none"> • Consider placing at bedside, otherwise place over soft padding
Wound care	<ul style="list-style-type: none"> • Use nonadhesive dressings • Take care with removal of previous dressings • Watch out for wound infections
Vascular access	<ul style="list-style-type: none"> • Avoid use of tourniquet • Secure with nonadhesive dressings • Consider arterial line placement if frequent blood pressure readings will be needed • Consider central line placement if longer-term IV access is required
Urinary catheters	<ul style="list-style-type: none"> • Use the smallest effective size and abundant lubrication • Do not secure catheters to skin with adhesives
Enteral tubes	<ul style="list-style-type: none"> • Use small-gauge, flexible NG tubes with liberal lubrication
Pain and anxiety	<ul style="list-style-type: none"> • For mild pain, use acetaminophen or ibuprofen • For moderate to severe pain, use opiates • Consider topical morphine and lidocaine
Pruritus	<ul style="list-style-type: none"> • Consider anxiolytics prior to dressing changes • Use topical corticosteroids judiciously given risk for systemic absorption • Use emollients • Use systemic antihistamines • Control ambient temperature as heat and sweating exacerbate pruritus
Medication administration	<ul style="list-style-type: none"> • Rectal administration is relatively contraindicated
Procedures	<ul style="list-style-type: none"> • Cleanse skin with agent by pouring or spraying onto skin and then patting dry • Avoid using drapes with adhesives; use towel clips instead
Intubation and oxygenation	<ul style="list-style-type: none"> • Lubricate nasal cannulas prior to placement • Avoid regular oxygen masks • Lubricate anesthesia face masks prior to use • Use smaller ETTs, preferably with nasal fiber-optic intubation • Utilize only direct vision suctioning

due to scarring, as well as severe dental disorders. Head and neck contractures may prevent adjustment of positioning.¹⁰ Most importantly, trauma from instrumentation may result in potentially life-threatening formation of bullae within the airway. Given these challenges, intubation in patients is ideally planned and performed by anesthesiologists with specific training in caring for this unique patient population. However, making these arrangements may not always be possible in the emergency setting.

Smaller endotracheal tubes (ETTs) are preferred in patients with EB versus the general population. Our preferred technique for airway management is nasal fiberoptic intubation because of improved visualization of the airway and because fiber-optic intubation causes less damage to mucosa than the use of a laryngoscope. In addition, nasal mucosa is composed of respiratory epithelium, which is different than and more resistant to blistering compared with the mucosa of the oropharynx. When laryngoscopy is performed, water-based lubricant should be used. After placement, ETTs may be secured using umbilical tape or ribbon gauze tied around the tube and patient's head.^{1,11}

Patients with EB may have abundant secretions due to esophageal strictures. However, only direct vision suctioning should be performed. Blind suctioning may result in bullae formation within the airway^{5,6} (Table 1).

FURTHER INFORMATION

Specific recommendations for management may be requested from your institution's consulting dermatology department. Another fantastic resource for questions that may arise regarding care of these patients is the nurse educators at Dystrophic Epidermolysis Bullosa Research Association of America (debra of America), a nonprofit devoted to support of the EB community. Nurse educators may be reached via phone or e-mail. Information about the organization and contacting nurse educators is available at the organization's Web site (<http://www.debra.org/>).

CONCLUSIONS

Patients with EB may experience iatrogenic trauma to skin and mucosa in the ED with even basic handling, monitoring, and

procedures. By being aware of EB and the special considerations necessary when caring for patients with the condition, we believe patient suffering may be avoided, and better outcomes achieved.

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