

Epidermolysis Bullosa



McKesson Patient Care Solutions

**Products and Resources
for Care of a Newborn with EB**

The EB Advocate Program

McKesson Patient Care Solutions Inc. (MPCS) offers exceptional service, more quality wound care products and more resources to help you provide more effective care for Epidermolysis Bullosa (EB) patients. Learn more about the benefits of choosing a supplier with extensive experience serving the EB community.



Exceptional Service

- MPCS has a dedicated EB Advocate Team and toll-free phone number 855.5EB.line (855.532.5463) to better serve patients who have EB.
- The EB Advocate Team’s product and insurance specialists are available to answer questions about wound care products or insurance coverage guidelines.
- MPCS is a contracted supplier with hundreds of managed care payers and is a participating provider with Medicare and most Medicaid plans.
- MPCS bills patients’ insurance on their behalf and ships supplies directly to patients. Orders and reorders are shipped within 1-2 business days.¹ Standard shipping is free.

More Quality Wound Care Products

- MPCS supplies quality wound care products from a wide range of manufacturers. Brand requests are typically honored if products are stocked by MPCS.
- The EB Advocate Team works with healthcare providers to support continuity of care and help them provide more effective care to their EB patients.
- Phone and fax ordering options are available to healthcare professionals and patients.

More Resources for the EB Community

- MPCS offers the informative and nationally respected EB Nurse website.
- Subscription is available to the e-newsletter, “The EB Advocate.” The newsletter spotlights people and events in the EB community. “The EB Advocate” is also on Facebook and Twitter.
- MPCS also serves the community that provides resources to patients who have EB.

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¹ Shipping is contingent upon receiving product authorization from the insurance payer and does not apply to incontinence products.

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McKesson Patient Care Solutions Inc. (MPCS) provides access to medical supplies, insurance payers, distribution services and technology to better serve our customers. MPCS is a leading provider of wound care, urological, ostomy, tracheostomy, incontinence and diabetic care supplies¹ as well as breast pumps.

Unless otherwise noted, the recommendations in this document were obtained from professional dermatologists. Be advised that information contained herein is intended to serve as a useful reference for informational purposes only and is not complete clinical information. McKesson cannot be held responsible for the continued currency of or for any errors or omissions in the information



¹ As a result of regulatory changes effective July 1, 2013, McKesson Patient Care Solutions Inc. is not able to provide diabetic care supplies to patients using Medicare Part B benefits.

The Newborn with EB

My child was born with possible signs and symptoms of epidermolysis bullosa. What do I do?

It is difficult to nearly impossible to diagnose a newborn with EB by physical exam alone, especially at birth and early in life. While it is true that the various types of EB have similar features at birth and during infancy, there is significant overlap within the clinical presentations that even the most experienced dermatologists cannot always accurately diagnose EB (and the subtype) without a skin biopsy or genetic testing.

Until the diagnosis is confirmed, parents are encouraged to focus on learning to care for their new baby rather than on the various subtypes of EB and possible challenges ahead. It is typical and understandable to feel overwhelmed and scared with a diagnosis you may likely know nothing about and may never have heard of.

One strategy for coping with these feelings is to try to learn what you need to provide the best care possible for your child while at home. As a parent you may find it helpful to research various support groups in your local community or state. Many helpful resources can be found on the internet. If possible, delegate a family member or friend to research EB and reach out to support groups in your area.

Focus for new parents of a child with EB include:

1. Care of wounds
2. Feeding and nutrition

Skin and Wound Care Treatment Steps

Covering affected areas with soothing bandages will make the baby more comfortable and make it possible for the parents to hold their child by protecting open skin areas. Bandages will also help the baby's wound heal.

Vaseline gauze is available in every hospital nursery and should immediately be wrapped around any affected areas. This will provide comfort and help prevent further damage to the baby's skin.

No tape or adhesives.

Place a LARGE sign over the baby's crib to alert all hospital staff that tape and other adhesives are not ever permitted on the baby's skin.

Wash hands.

This is the single, most effective way to minimize the spread of infection. Before and after dressing changes and handling the baby, wash your hands.

Set out all supplies.

Supplies will vary depending on the child's condition and which products are available.

Commonly used products include:

- Sterile needles or lancets for draining blisters
- Antibiotic ointment such as mupirocin, also called Bactroban™
- Ointment such as Vaseline or Aquaphor™
- Contact layer (first layer to touch the skin) such as Mepitel®, Restore, Vaseline Gauze, Mepilex® Transfer
- Soft, conforming, stretch rolled gauze such as one or two-inch second layer
- Tubular dressing third layer retainer such as Tubifast™ or Surgilast®

Drain blisters.

Blisters should be drained if they are tense or ½ inch in diameter or larger. This is because the fluid inside the blister puts pressure on the surrounding skin, causing the blister to grow. A larger blister takes longer to heal and is at greater risk of becoming infected. It is also more painful than a smaller one.

The side of the blister should be punctured parallel with the skin using a sterile needle. Be sure to lance the blister all the way through from side to side. You may have to lance the blister in multiple places to release all of the fluid. Be sure the opening in the blister is large enough that it will not seal and refill.



Every effort should be made to leave the blister roof (top of the blister) in place, as this improves comfort and healing. Do not denude (unroof) the blisters. Do not attempt to pop any blisters on the face or around the eyes. Parents should contact their baby's healthcare providers for assistance with this.

Remove dressings.

It is best to remove the dressings on one limb or area at a time. Further skin damage may be prevented if all dressings are not removed at once. The baby will naturally kick and rub the hands or feet causing bleeding and injury. Also, the wounds are more painful when they are exposed to air.

Remove the dressings from one limb at a time, drain any blisters, clean the wounds and then apply the new dressing to that limb before proceeding to the next area. Do not pull off dressings that are stuck to a wound. Clothing or bandage materials that have adhered to a wound must be soaked off with warm water to help prevent further damage to the skin, bleeding and pain. Often, soaking with water or applying a warm, wet compress will be enough to remove an adhered bandage. Using sterile saline solution (often supplied by the hospital or wound care company) may sting less when used on open wounds than tap water. If these techniques are not effective, apply an ointment such as Vaseline or Aquaphor™ to the adherent dressing instead. This will allow easier and less traumatic removal of the dressing.

Cleanse skin.

The intact skin and wounds may be cleansed with a mild, non-perfumed, non-soap cleanser, such as Cetaphil™ gentle skin cleanser or Dove™ body wash (for sensitive skin, unscented). Areas with intact blisters, open sores or missing skin may be rinsed or irrigated with a "soapy" solution (Cetaphil™ or Dove™ and mixture of water) and then with plain water. If necessary, the areas may be patted gently. DO NOT RUB the skin dry.

Apply ointment.

Antibiotic ointment (such as mupirocin ointment) may be applied to areas that appear infected if instructed by your doctor. The use of antibiotic creams or ointments, whether prescription or over the counter, as prevention of infection, is not recommended in most cases and should be discussed with your child's physician.

Some patients may develop bacterial "resistance" to topical antibiotic preparations when used for long periods of time. Rarely, an allergic reaction to some topical antibiotic agents can occur and cause a skin rash and itching. Some parents prefer to use non-medicated ointments, such as Vaseline or Aquaphor™ to lubricate dressings prior to application to the skin.



Apply dressings.

A non-adherent contact layer should be applied. Examples of contact layers include Mepitel®, Mepilex®, Restore, Vaseline Gauze or Telfa™ (non-stick) pads. Then, the conforming rolled gauze is wrapped around the contact layer and is secured with the tubular dressing retainer.

Not every small blister requires a dressing. Simply applying an ointment on the affected areas may be enough for small, isolated lesions. Bandaging is necessary if a blister or an open wound:

- Requires protection from further trauma or contact or friction with clothes
- Exhibits draining or bleeding, or has a “scab”
- Requires topical treatment for infection
- Pain is present and a dressing will improve comfort

At first, until it is determined exactly how fragile the newborn’s skin is, daily or every other day dressing changes are recommended. New blisters may develop under the dressings and therefore, the skin must be inspected and the blisters drained as necessary. Some dressings tend to hold less moisture and may stick to the skin if not changed every day. Vaseline or Aquaphor™ ointment on the wounds or a contact layer can also help prevent the dressing from drying out between changes.

It is not uncommon for some EB wounds to bleed or leak a clear light yellow fluid (also called serous drainage). If dressings become soiled or there is a significant amount of drainage from the wounds, then the dressings should be changed at least every day. Dressings that become soiled with fluids from the wounds that are not changed regularly may be a source for bacteria to grow and a possible wound infection could occur.

Missing Skin

Many babies with EB are born with missing skin on the legs, feet, hands and/or wrists but this can occur anywhere on the body. This is referred to by various names:

- Congenital Localized Absence of Skin (CLAS)
- Bart’s Syndrome
- Aplasia Cutis or Aplasia Cutis Congenita

Gently cleanse these areas by irrigating with sterile saline or soapy water (mild soap such as Dove™). Rinse with water. Gently pat dry. Apply a contact layer such as Mepitel® or Vaseline gauze to cover the entire denuded area. Do not overlap the dressing or the area will become too wet.

You may choose to “butter” the Mepitel® or Vaseline gauze with Aquaphor™ or petrolatum jelly before applying. Be sure not to coat the edges of the Mepitel® dressing with ointment or it may become too wet and “float” all over rather than remain in place over the wound. Wrap with conforming rolled gauze and secure with a tubular dressing retainer.

DO NOT USE TAPE!

These areas remain fragile for extended periods of time, even after they have healed. Because they are typically located on the feet or legs of EB newborns, they are subject to trauma caused by kicking and movement. They may require protective wrapping for months or longer to prevent repeated re-injury.

Pain

EB wounds are painful. Covering the areas with comforting ointments and bandages can provide sufficient pain relief. Having assistance from a family member or friend and working fast and smart is helpful when performing dressing changes.

Pre-medicating before dressing changes and baths may be necessary for some infants initially. Using a non-sedating medication, ONLY if instructed to do so by a physician, such as acetaminophen (Tylenol™) or ibuprofen (such as Motrin™) may be helpful. Narcotic medications (such as

morphine) are usually not needed and are often avoided because they cause drowsiness and affect the child’s ability to stay awake and take part in feedings. They can cause significant constipation that can lead to problems, such as stomach discomfort, pain while defecating and anal fissures (deep cracks in the skin). Long-term use can lead to increased levels of pain. When such medications are necessary, caregivers should try to time the dose so that it will have minimal impact on feeding schedule. Narcotic medications should only be given at the direction of the physician during a very limited time period for severe pain.

Oral sucrose (sugar) is another alternative for short-term pain in infants under the age of six months but only if used very sparingly. The effects last approximately five minutes, making it ideal for intense short-term pain such as injections, blood tests, suture removal, etc. Dipping a pacifier into a 24% sucrose solution has the added soothing benefit of non-nutritional sucking.

Oral sucrose should not be used for fussiness or other non-pain related difficulties. Sucrose used this way will significantly limit its effectiveness and may cause it to stop working entirely. Sucrose is ineffective if administered directly into the stomach. There is little research in the use of sucrose in extremely preterm, unstable and/or ventilated babies and should only be used with caution.

Oral Blisters or Lesions

In the mouth, a drained blister may look like a white patch and can sometimes be mistaken for thrush (a common yeast infection in babies) by those unfamiliar with EB. Most blisters that occur in the mouth will rupture on their own when a child eats or drinks.

For oral lesions that result from mouth blisters, your provider may prescribe Magic Mouthwash, a combination of one part viscous lidocaine, one part liquid diphenhydramine (Benedryl®) and one part liquid aluminum hydroxide or magnesium hydroxide (Maalox®).

In older children this is often used as a swish-then-spit preparation. The mixture can be applied to painful areas of the mouth with a Q-tip or toothette (small sponge on a stick) in infants and young children. Apply cautiously because the viscous lidocaine may numb the gag reflex and allow choking. For this reason, use of Magic Mouthwash in infants and very young children is usually not recommended.



Infection

For infants or children with EB, it is extremely important to recognize an infection, make your child's physician aware and treat it promptly.

Many medications, including some topical antibiotics (silver sulfadiazine), may not be used on newborns. You MUST consult with and follow the advice of your primary care practitioner or dermatologist if you suspect your baby has an infection.

Signs of infection include:

- Increased drainage (fluid from wounds) that is thick yellow or green
- Increased pain
- Increased swelling
- Increased warmth of skin compared to surrounding area
- Malodorous (unpleasant smell)
- Easy bleeding
- Fever greater than 101 degrees without another cause

Nutrition

Babies and children with EB require more calories for normal growth and development in comparison to a child without EB. Children with EB require more calories to heal open wounds and grow. They lose important nutrients and fluids from the blisters and open wounds. Infants with blisters and lesions in the mouth may have difficulty latching on and maintaining a strong suck. Because nutrition is so important in babies with EB, expressed breast milk is often provided rather than actual breastfeeding.

If mother decides to breast feed, the baby's weight should be monitored closely. If the baby fails to gain weight, feeding breast milk from a bottle and/or supplementing with formula will likely be necessary. Practitioners should help the mother understand that most babies with EB have difficulty breastfeeding successfully, but bonding will occur if mother holds and cuddles the baby during feedings.

Many babies with EB have difficulty sucking from a standard nipple. Cleft palate nipples, such as the SpecialNeeds® Feeder (known in the past as the Haberman feeder) www.medelabreastfeedingus.com/products/576/special-needs-feeder can be used.

SpecialNeeds feeders or the Pigeon feeder (<http://www.cleftadvocate.org/feeders.html>) have a valve that facilitates easy delivery of the formula without the need for a vigorous suck.

Milk may even be squeezed into the baby's mouth if sucking is still traumatic for the baby when using a cleft palate feeder.

Infant mouth care may include gentle cleansing with a spongy toothette. Often, the blisters in the mouth appear white and may be mistaken for thrush (yeast infection in the mouth). Most babies are vigorous eaters, even with blisters in the mouth. As long as the baby is eating well, the blisters should be left alone and allowed to rupture and heal on their own.

If weight gain slows, stops or if the baby does not begin to gain weight after a brief period of weight loss (usually occurs within the first week of life), a pediatric registered dietitian should be consulted.

There are many strategies that can help the child gain weight, including fortifying breast milk or formula, providing higher calorie formula or providing formula that is easier for the baby to digest. The registered dietitian should work together with the pediatrician to develop a weight gain plan for the child. At minimum, weekly weight checks should be monitored for the infant who is not on an upward growth curve.

Some babies with EB require gastrostomy feeding tubes, also known as "G-tubes," if they are unable to take in sufficient calories by mouth. This is determined by tracking the child's height and weight on a growth chart. The pediatrician will keep these charts as part of the baby's medical record, but it is helpful for parents to track the weight as well. Parents should be aware of their baby's height and weight and should seek support if the baby is approaching the tenth percentile or lower. A feeding tube should be discussed with the child's pediatrician or family doctor, a pediatric gastroenterologist and/or a pediatric surgeon. Feeding tubes have been used with significant success in patients with EB.

Circumcision

It is safe to circumcise a baby with EB as long as the baby does not have any eroded or missing skin in the diaper area.

The child will require padding before being harnessed for the procedure. Healing from the procedure is reported to be comparable to that of babies without EB. The American Academy of Pediatrics very strongly recommends taking precautions to manage the pain associated with the procedure. Talk with your obstetrician or child's pediatrician about options.

Diapers

Diapers can often cause blistering and skin shearing around the thighs and waist due to repeated friction. Liberally apply Vaseline or Aquaphor™ ointment to reduce friction.

Many parents prefer disposable diapers because they keep the urine off open wounds on the buttocks. Some disposable diapers have been reported to cause less damage than others due to their cloth-like softness. Some brand examples include:

- White Cloud (Walmart brand)
- Huggies® Supreme
- Huggies® Ultratrim
- Pampers® Swaddlers

Many parents cut the elastic off the legs of disposable diapers to prevent irritation, blistering and skin shearing. Some parents take fleece or a soft fabric and extend it through the leg and waist area to cut down on friction. Another option includes the use of Johnson & Johnson nursing pads, coated with Vaseline or Aquaphor™ ointment to pad areas of friction.

Some parents prefer cloth diapers and cloth diaper covers. Two brands that EB parents have used successfully are:

- Fuzi Bunz (www.nurturedfamily.com)
- Happy Heiny's Pocket diapers (www.happyheiny.com)

Mepilex® Lite or Mepilex® Transfer may be used to protect the skin, but this is costly, as the dressings will require frequent changing when soiled.

Hand Care

If the fingers are not significantly blistered or eroded, lubricating the hands with Vaseline or Aquaphor™ ointment and applying soft, infant mittens may provide adequate protection.

Individuals with specific types of EB are at risk for "mitten-like" deformities of the hands and feet where the fingers and/or toes eventually fuse together due to repeated blistering and inflammation. During the immediate newborn period it is almost impossible to individually wrap each finger without causing significant issues for the child. The focus should be on wrapping the hand (if needed) with the thumb out and separate from the rest of the hand. When wrapping the hand, it is helpful to add a small well-lubricated ball of Vaseline gauze or other dressing in the palm of the hand. This ensures the fingers will not rub on the base of the palm when the baby makes a fist and help prevent blisters on the fingertips.

As the child gets older and is able to relax the hands and feet, wrapping the fingers individually will significantly help prevent "mitten-like" deformities of the hands and feet.

Bedding and Crib Mattresses

No benefit has been reported from using special mattresses. Soft, jersey-knit sheets are found to be soft and comfortable.

While some parents prefer to use satin sheets, they are not recommended due to the risks of overheating, sticking and blistering from rubbing against any blood that may have dried on them.

If a wound is present on the back of the scalp, some parents have found it helpful to actually apply a thin layer of Vaseline directly to the sheet in an effort to reduce friction.

Bath and Bathtubs

When the baby is a newborn, provide sponge baths as needed to prevent skin damage. When you are ready to give your baby a bath and depending on how fragile the skin is, you may choose to begin the bath with the dressings in place.

Rather than placing the baby in a typical baby bathtub, you may choose to use a bath sponge. These are inexpensive and will provide padding, slip resistance and comfort for the parents and the baby. Parents should follow the pediatrician's recommendations regarding water temperature.

Bath time is not always fun for kids with EB. Wounds may be painful when exposed to air and water. Damage may occur to intact skin or to wounds during the bath. Pre-medicating the baby with Tylenol™ before the bath and dressing change will help with pain control under the direction of the baby's doctor. Being well prepared and having an extra person available will make the event less stressful. Additionally, colorful bath toys available for the baby to look at or interact with can be very helpful to keep the baby distracted from the bath.

Immunizations

Immunizations should be given on schedule, unless the pediatrician decides otherwise.

When giving injections to an individual with EB, there should be no vigorous rubbing or massaging of the site. Tape or adhesive bandages should never be placed over the injection site.

Home Health Nurses

Home Health nurses should be ordered by the physician if parents need assistance with dressing changes or with help understanding the specific care required.

Home Health nurses are able to monitor the nutritional status of the infant.

Parent Self-Care

It is very normal for parents to feel overwhelmed and nervous with any new baby, but this can be especially true of parents who have a baby with EB. Some parents feel guilty, thinking they did something to cause the disease or they should have done something to prevent it. While these thoughts are not uncommon, nothing could be further from the truth. Additionally, self-blame is not productive and can hurt the relationship between the parent and the baby by interfering with the parent-baby bond.

It is very important that parents ask their baby's physician all of their questions. Don't worry about sounding "silly" or "stupid." Knowing how to care for the child's needs will only help parents and caregivers feel more confident in caring from the baby.

Parents and caregivers often say they will take care of themselves "later," but "later" often never arrives. Parents who regularly schedule time for themselves to recharge, typically have more energy, patience and efficiency compared to those who do not take time for themselves. Some things that some parents have found helpful are:

- Going for a walk
- Exercising
- Meditating
- Yoga
- Reading a book
- Going to a movie

Sometimes parents find they need more help with their feelings than what friends and family can provide. Parents who have concerns about depression, anxiety, anger or other emotional or behavioral issues should talk to their doctors about how to get professional help with these difficulties. Family therapy can be useful in helping the entire family work together to cope with changes and feelings that come with a new baby who requires special care. Asking for help early will make it easier to address concerns and help everyone return to a healthier lifestyle quicker.



Gentle Handling

Every effort should be made to prevent trauma and friction to the skin.

Babies and young children with EB should not be picked up from under the arms. Rather, they should be lifted by placing one hand behind the head/neck and one hand beneath the bottom.

Every baby needs to be held and nurtured. It is perfectly normal to feel afraid, but your baby will feel safe. Babies with EB need the added comfort and security of being held and touched, just like any baby.

Blisters are a part of an EB child's life and cannot be avoided. Parents will find a comfortable way to hold their child. Once the baby is properly bandaged and protected as much as possible, the child should be held and cuddled the same as any baby. Swaddling the baby will offer added protection. Parents should cut their own fingernails short, remove large rings and jewelry, as well as wear soft clothing that will not irritate the baby's skin.

Clothing

It is helpful to choose clothing that is made of soft fabric that is loose fitting without tight elastic bands and has easy access for diaper changes and bandage checks.

Clothing tags, buttons, snaps or zippers may cause blisters or skin shearing and should be used cautiously.



Some parents choose to dress their child with clothing worn inside out to prevent the seams from irritating the skin. Additionally, bandages or soft undergarments (t-shirts or leggings) may be necessary to protect the skin. Often the baby will have facial blisters from scratching or rubbing. Placing mittens or socks over lubricated hands will help prevent this and offer protection. The toes may be cut from larger socks and pulled over the elbows to protect from rubbing and shearing.

Lubricate the Skin

You may find it helpful to apply Vaseline or Aquaphor™ ointment to normal skin to help reduce friction and help prevent further blistering.

Cool Environment

Children with EB may be more sensitive to hot, humid conditions, largely due to the amount of dressings they wear. It is best to keep them in a cool environment when possible.

Before placing your child into a car, be aware of the temperature and cool it in advance, if needed. Similar attention should be paid to assure the baby is not kept too cool either. Babies may need an extra layer to maintain body heat.

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