

Basic Care Tips for Epidermolysis Bullosa (EB): A Parent's Guide

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Basic DOs and DON'Ts:

DO love your child with all your heart and soul. **DO NOT** be afraid of them; they will not break. With the following tips and the utmost of care, you can give your child the best life possible, despite the challenges of EB. Different types of EB call for different levels of care, and not all children thrive despite our best efforts. **DO** your best and never give up.

DO educate yourself and those who care for your child on the best methods to use for your child's care.

DO NOT be afraid to question anything and everything in regards to your child's needs, even if that means questioning the medical community or those advising you. Each child is an individual, and there is no "one size fits all" treatment for EB. Some things work well for one child, yet may be damaging to another.

DO experiment to see what works best for your child and ask other parents of EB children what works best for them. They are often your best resource.

DO NOT use adhesives, such as adhesive bandages or tapes on your child's skin.

DO pop blisters as they occur and/or recur. A sterile needle or very sharp clean scissors work best. See **Blister Popping** below. **DO NOT** remove the blister cap from the freshly popped blister.

DO use soft, non-abrasive materials for bedding and clothing. Satins and silks or very soft cottons are especially comfortable. It is often a good idea to turn clothing inside-out so that the seams do not rub against your child's skin.

DO NOT pick up your child from under his/her armpits. Pay attention to where his/her wounds are, and pick him/her up from his/her bottom or upper thighs and across his/her back provided these areas are not severely damaged. Take care to distribute pressure to the palms of your hands rather than just your fingertips. Cradling small infants on a piece of sheepskin can be very safe and comfortable, especially when moving them from place to place.

DO NOT take your child's temperature rectally. Use an ear thermometer (without pulling the ear) or take the temperature from under the armpit. When speaking with your child's health care provider, be sure to mention the method used to check the temperature.

An overview of treatment

EB is caused by missing or dysfunctional connecting structures between the layers of the skin. It is as if the glue that holds the skin together is missing. Another example is to imagine two pieces of Velcro stuck together; in the case of EB, the hooked, receiving-side is either missing its hooks or the hooks are improperly shaped, making for a weaker connection. Understanding this, the next question that often comes to mind is “how is EB treated?” Currently, there is no cure or specific treatment to alleviate the complications of EB. However, great strides have been made in the understanding and treatment of this disease.

Our job as parents or caregivers of an EB child is to provide protection and the resources for good health in order to provide our children with as normal a life as possible. The different types of EB require different approaches to care. Many persons with milder forms of EB have minimal involvement and need little or no treatment. Some of the less severe forms also improve with age. The more severe forms of EB often require hours of intensive nursing care, similar to the care used on burn victims. A team of many doctors and health professionals is often required. It is best to take a team approach in caring for an EB child. The members of this team may include the primary care physician (often the pediatrician), the dermatologist, any nurses involved, the pediatric dentist, the gastrointestinal doctor (deals with digestive disorders), the dietitian or nutritionist, the plastic surgeon, the psychologist or social worker, and the genetic counselor. In the non-medical area, some of the care team may include siblings, relatives, babysitters, teachers and others.

The primary goals of EB care are: protection of the skin against trauma, prevention of infection, maintaining the highest possible level of nutrition and avoiding dietary complications, minimizing deformities and contractures, and lastly, sustaining a strong support system and a positive attitude. Many of the complications of EB can be lessened or even avoided with proper early intervention and care.

The following are suggestions for approaching the care of all forms of EB. Each recommendation may or may not pertain to every form of EB, so discretion and careful experimentation should serve as a guide in each individual case.

Section 1: Protecting the Skin

EB is caused by abnormalities in the connecting structures that hold the different layers of skin together. Because of these abnormalities, friction on the skin leads to blistering. The amount of friction needed to produce blistering depends on the type of EB. In some cases, blistering is only produced after vigorous exertion, but in others, something as trivial as a seam rubbing against the skin may produce a blister. A primary goal of EB care is to reduce the likelihood of developing new blisters. Preventing all blisters from occurring is impossible, but by using these suggestions, your child's general comfort will be improved. Use common sense, and keep these ideas in mind in your daily care of your child.

1a. Clothes

In many forms of EB, even slight friction on the skin causes blisters. Dress your child in breathable, soft clothing with minimal seams to reduce the likelihood of blistering. For babies, turn clothing inside-out so that the seams are not directly against the skin. Fabrics such as satins, silks and soft cottons are ideal materials for both bedding and clothing. Loose-fitting and lightweight clothes are typically comfortable; however, snug tights or leggings help to hold bandages in place and also help to protect the legs. Leggings can be used instead of stretch netting (see **Bandaging**). Always keep in mind that clothing should be easy to put on and take off. Be especially attentive to the head area, ears and armpits when dressing or undressing your child.

1b. Shoes

Shoes are a challenge for some EB children. Firm or poorly-fitting shoes may produce painful blisters on the feet. Using bandages to pad the child's feet is helpful, but this also makes it more difficult to find shoes that fit properly over the padding. When looking for shoes, note where the seams are, whether or not the shoes are wide enough to accommodate extra padding if necessary, and if they will tend to rub in certain areas. Soft slippers, especially those lined with sheep skin, often work better than conventional shoes. Make sure the slippers or shoes have a wide opening for easy access. Children with severely affected feet often prefer not to wear shoes most of the time and just walk around with extra padding on their feet.

1c. Newborn comfort

For newborn babies, it may be necessary to lay them on an absorbent pad rather than use a diaper. Cloth diapers may be preferred in some cases, although many of the disposable diapers with Velcro closures are desirable because of their better absorbency. With a little extra padding at the pressure points, these often work just as well or better than cloth. Another suggestion is to cut the elastic leg cuffs out of disposable diapers. It is very important to keep this area of the baby as clean and dry as possible.

For **bedding** and **car seats**, sheepskin is an excellent padding material. Both natural and synthetic sheepskin work well and can be obtained fairly easily. Egg-crate foam or other foam materials can be used to add extra padding beneath bedding as well. Do not lay a baby directly on the sheepskin or foam to sleep. Silk, satin or soft cotton sheets should be used over the foam or sheepskin. Water or air mattresses can also help to reduce friction and add comfort.

Section 2: Skin Care and Bandaging

As mentioned in **Section 1**, despite our best efforts, it is impossible to prevent all blisters from forming. Areas of skin breakdown will also occur. Maximal skin care, with attention to these areas, is very important in EB for several reasons. Proper wound care helps to minimize pain, promotes healing, and reduces the likelihood of developing skin infections. This section outlines a recommended approach to skin care and bandaging.

2a. Setting up your supplies

Once you are familiar with the bandaging materials that work best for your child and know at what step of the bandaging process to use each item, bandaging set up becomes fairly routine. It is very important that all set up be done ahead of time. You do not want to be looking for items or cutting bandages while a limb or other area is exposed and unprotected. Setting up your supplies before bath time is helpful. Alternatively, if your child is old enough to be in the bathtub unsupervised, supplies could be set up at that time.

Have a clean surface set aside for your bandage supplies. For a list of suggested products, see **Section 3**. All scissors and tools should be pre-sanitized. Open all packages and pre-cut dressings to size. If needed, coat dressings with additional ointment at this time. Applying topical products (ointments, antibiotics, etc.) to the dressings is easier and less damaging to the skin than applying them directly to the wound. Setting up your supplies is an exercise in estimation. Think about the areas to be dressed and visualize how much of each product will be used and what sizes will work best. For instance, a small child's leg with wounds on the knees, feet, and shin area, will likely require several strips of Vaseline gauze, perhaps some Mepilex Transfer, or some other non-adherent dressing. It will also need 2-3 rolls of 3- or 4-inch rolled gauze, and tubular netting, cut to fit the length of the limb.

Your child will need to sit or lay on a bed, table or chair during the bandaging process, and this also needs to be clean and ready to go before bandaging starts. The surface should be soft, comfortable and clean. A clean towel over a washable sheepskin works well. Blue chux pads, or a similar product, add a waterproof layer, and can be added on top of the towel, and then covered with another towel. After bathing, the top towel and chux can be removed, leaving a dry layer for bandaging.

Bandaging may seem tedious, but it is a necessary part of your child's care. Allowing your child to listen to music or watch a video during bandaging will help combat the boredom. Also, being organized helps to streamline the bandaging process. It also minimizes disruption to your child, and reduces the likelihood of contaminating your supplies. Remember to always keep cleanliness a top priority. Bandaging is not a surgical procedure, and items will not be completely sterile. Once packages are opened, they are no longer sterile. However, with common sense and care, contamination with bacteria and germs can be kept to a minimum.

2b. Bathing

Bathing is very important for children with EB. Washing the skin helps remove dry, dead skin and the crusts from areas of skin breakdown. It also helps to reduce the amount of bacteria on the skin, helping to prevent infection. Lastly, soaking in water helps soften bandages that are stuck to the skin, making their removal less painful. Your child's doctor may recommend adding household bleach, vinegar or medicated washes to your child's bath water. Be sure to sanitize the bathtub / shower with bleach before and after your child bathes.

When helping with your child's skin care, first make sure your hands are scrupulously clean! Wash them with soap and water and dry them with a clean towel.

Before your child gets into the bath / shower, begin removing their bandages. Netting and the layers of padding can be cut off. Be careful when removing the bandages in direct contact with the skin. Any bandage that does not come off easily can be left on and soaked off in the water.

Newborn babies are often difficult to handle, so use extreme care and try not to let your hands slip on their damp skin. Rather than immersing them in a shallow tub, it may be necessary to bathe them in sections. Remove a portion of the baby's bandages, and using a basin, wash the exposed areas. A new, clean turkey baster can be used to gently flush wounds. A second rinse with clean water or water with bleach or vinegar (if recommended by your child's doctor) is a good idea as well. This process is then repeated for the other areas.

Older babies and young children can sit on a soft towel and be bathed gently, or use the method described for newborns if bandage removal seems too damaging. Older children may choose either to bathe or shower. For all ages, soaking off bandages helps to prevent sticking, minimizing injury to the surrounding skin. If soaking does not help, a dab of Aquaphor® ointment or a similar product such as petroleum jelly will often release the bandage. In the bath, pay attention to any raw area on the skin. Note any areas that are moist and oozing as they are potential sights for infection.

2c. Blister Popping

After your child's skin is clean, any new blisters need to be popped before bandaging begins. Contrary to what is taught in regards to normal skin, in EB it is important to pop blisters as they occur. In normal skin, the connections between the layers help keep the blister contained in a small area. However, in the absence of proper connections, there is little resistance to keep blisters from growing bigger and bigger. The pressure of the fluid in the blister lifts the nearby skin, making the blister larger. Popping blisters helps to keep them from spreading. It is important to leave the blister cap on the wound, as it serves as natural protection or a covering for the area. The blister cap will eventually dry up and lift off, but leaving it in place often allows the lower layer of skin to heal a bit before being exposed.

There are several good methods to pop blisters. These include using a sterile needle, scalpel, or very sharp tiny scissors. Keep in mind where the blister will be draining, and pop it at the lowest point along its edge so that gravity will further assist it in draining the fluid. If using a needle or scalpel, be very careful not to poke directly towards the patient. Instead, slide the needle or scalpel carefully in a parallel direction with the skin. You will be far less likely to jab your child, and it allows the blister to drain through the slit. When using a needle or scalpel, your goal is to make a small tear in the blister. Avoid simply making a small hole, as this will close up quickly. If using scissors, you can actually cut a small slit by basically pinching a very small section of the blister cap between the scissor blades. Unlike needles and scalpels, scissors are not disposable, so be sure to clean and sanitize your scissors frequently (see **Section 3a**). Sterile gauze can be used to absorb the blister fluid. Another effective and less expensive option is a clean tissue. Gently touch a tissue to the blister opening, and the fluid will be wicked out easily. It is best to avoid pressing on or rubbing the blister to try to empty it. This may force the fluid to move away from the opening and expand the size of the blister. If you must press on it at all, be sure to press the fluid towards the opening.

2d. **Moisturizing the skin**

Using lotions, creams or ointments will help keep your child's skin moisturized, keeping your child comfortable. Dry skin is more likely to itch (see **Itching**). Moisturizers can be used all over your child's body if desired. Ointments are also applied to wounds to promote wound healing and reduce scarring. It may be necessary to rotate the products you use. EB tends to cycle and what works well for a while may become less effective after a long period of time. As an example, you could alternate between using Aquaphor® Healing Ointment for a month and then A&D ointment for a month. For particularly stubborn wounds that are not responding to these ointments, try some Zinc Oxide ointment for a couple of days.

2e. **Bandaging**

There is no "one size fits all" care plan for EB. You will find certain bandages or routines that work best for your child. These suggestions are designed to highlight the rationale for the steps in wound care and serve as guiding principles for your child's care plan. A list of suggested wound care products can be found in **Section 3**.

Bandaging wounds is important for several reasons. Firstly, dressings help reduce the pain of open wounds. They also provide a barrier that keeps irritating substances, which may be painful as well, out of the wound. The skin also heals better in a moist, covered environment. New skin grows in from the edges of wounds, and this is hampered if the wound is dry. Bandages also protect wounds, making infection less likely. Additionally, bandaging and padding on non-wounded skin helps prevent damage from trauma.

The first layer of bandages is in direct contact with the wound. For this reason, it is important to use products designed not to stick to the skin. Used with ointments, these **non-adherent dressings** provide the first layer of coverage over the wound.

Conventional dressings, such as gauze, dry out and stick to wounds, leading to pain and damage to healing skin when removed. Because non-adherent dressings do not stick to the wound, they are less damaging to the skin and allow quicker wound healing.

After the non-adherent layer is in place, **absorbent dressings** make up the second layer. Various types of gauze or soft wraps are absorbent dressings. They help to absorb drainage from the wounds and also function as padding or protection for delicate parts of the body. Some non-adherent dressings also have absorptive properties, but those dressings that are not specifically non-adherent should not be used directly on top of wounds.

Finally, a layer of **netting** can be used over the top of the absorbent dressings to keep everything in place. This breathable, stretchable material conforms to the shape of your child's body. Netting is only to be used over other layers of bandages; it should not be used directly against the skin. If possible, try to avoid using tape to secure bandages. If tape must be used, use paper tape and apply it to a dressing such as rolled gauze. Never use tape directly on the skin.

2f. **Helpful hints**

Medical gloves (e.g. latex or vinyl gloves) are not recommended for handling an EB child's skin. They tend to grab the skin and can actually cause more damage. Minimize the risk of infection by frequently washing your hands, and if gloves must be used, lubricate them with an ointment such as petroleum jelly. Gloves are handy for coating bandages with ointments during the set up process.

Remember, soaking the bandages off helps to prevent sticking. Also, a dab of Aquaphor® Ointment or a similar product such as petroleum jelly will often release the stuck bandage.

Scrupulous cleansing of wounds and frequent dressing changes are very important to prevent overt infection. The use of anti-bacterial washes, ointments, modified Deacon's solution (2 tsp regular strength household bleach/1 gallon water), or wet soaks should be directed by a physician.

It is a good idea to rinse an area with clean water once the bandages are removed, especially if the bandages must be kept on for protection during the bathing process. A clean turkey baster works great for this job.

If netting is not available, a section of rolled gauze can help bind bandages in place. After wrapping the affected area, leave a 6 inch tail of gauze hanging. Cut this tail up the center lengthwise, and then twist and wrap the 2 strips around the limb. Tie the 2 strips in a knot to secure them.

Section 3: Supplies

This section contains recommendations on products that some families have found to be helpful. The brands and products listed are just a handful of what is available to help you get started. Your supplier may carry similar products in other brand names that work just as well. This list is not meant to promote a particular company or brand.

There are too many available products to describe every one, but this list gives you some general ideas to get started. They are listed in order of application, layer by layer. It may be necessary to contact individual companies for more details on sizes and order #s, however, most bandaging supply companies can work off of the information given here and find the specifics for you. Never hesitate to ask for samples. Many companies will ship directly to you (at a great savings) once they become aware of the intense needs of an EB patient. Also keep in mind that there is a huge mark up on products, and most companies and suppliers will work with you if the situation is explained. If your insurance company is reluctant to work with you, sending pictures of the types of wounds you are dealing with may help. Pictures are worth a thousand words. Remember that EB is a very rare disease, and most people have no idea what you are dealing with.

3a. Tools

Sharp scissors are needed to cut and trim bandages. Bandaging scissors can be ordered or found in medical supply stores. Regular scissors also work for cutting dressings to size. A pair of small, sharp, fine scissors can be used to pop blisters and trim dead skin. These can be found in the grooming section of any drug store. Manicure scissors work great. Be sure to clean and sanitize your scissors after use. Wash them in soap and water or wipe them down with alcohol. Neither of these processes will kill all germs, however, so periodically boiling scissors in water is also recommended. Needles or scalpels are useful for popping blisters but are sometimes hard to obtain. Most medical supply companies or pharmacies require a prescription from your doctor in order to obtain needles or scalpels. When using any of these tools on more than one area, avoid cross-contamination by sterilizing the tool between uses.

3b. Ointments

Several non-medicated ointments are available over-the-counter without a prescription.

Aquaphor Healing Ointment is manufactured by Beiersdorf, Inc. A 14 oz. tub costs about \$17. For more information, see www.AquaphorHealing.com.

Over the counter **petroleum jelly** and **zinc oxide** are also handy ointments. Zinc oxide should not be overused on open wounds, however. It is best to rotate its use with other ointments. **A & D ointment** is also recommended.

It is possible to get ointments covered by your insurance company. This may take a battle, but it is well worth it. Start by having your physician write a prescription, as well as a letter, explaining how important these products are in your child's skin care routine.

Any product containing **active ingredients** (anti-itch creams, for example) should only be used after consultation with your physician. Active ingredients may be excessively absorbed through skin wounds. Likewise, **antibiotic ointments** should be used with caution and with the supervision of your physician. Some people develop allergic reactions to topical antibiotics over time, and the prolonged use of certain topical antibiotics can lead to the development of infection with bacteria that are more difficult to treat.

3c. Non-adherent dressings

Vaseline® Gauze is petrolatum-impregnated gauze. This dressing is very easy to use, relatively inexpensive and readily available. It comes in a variety of sizes. The commonly used sizes and the order numbers for the Kendall brand of Vaseline Gauze are listed. The 1/2" X 72" strips are handy for the web spaces between fingers and toes and for wrapping wounds on fingers and toes. For this size, the order # is 8884-421600. The 3" X 9" size is order # 8884-413605, and the 3" X 36" size is order # 8884-415600.

Aquaphor® Gauze is another good non-adhering dressing. However, it often needs some added ointment and does not conform quite as easily on small, tight areas. It is less stringy than Vaseline® Gauze. For a 3" X 8" size the order # is 45588000. This product is manufactured by Beiersdorf-Jobst, Inc.

Mepitel® is an advanced, non-adherent silicone dressing that has been very beneficial in wound healing. Made by Molnlycke Health Care, a Swedish company, its main drawback is its cost. It is not recommended for infected wounds. This product can be re-used by washing it with anti-bacterial soap, soaking it briefly in a bleach solution, and then drying it in a clean environment. For the 3" X 4" size, use order # 290710. For the 9" X 12" sheet, use order # 292005. Two other products made by this same company are **Mepilex®** and **Mepilex Transfer®**. Mepilex® has a 1/4 inch-thick foam pad backing that draws infection and exudate from the wound and into the pad. This product is especially useful on oozing wounds, but can be overly sticky on healthier skin. For the 4" X 8" size, use order # 294290. Mepilex Transfer® is similar, but has a thinner backing and conforms more easily to tighter, bendable areas. The thinner foam also draws moisture out of the wound. For the 8" X 20" size, use order # 294592. More information is available at www.tendra.com.

3d. Absorbent dressings

Rolled Gauze comes in many brands and sizes. One of the most common brands is **Kendall Conform®**, which is sold in non-sterile bags of 12 rolls per bag. This is a practical and economical way to order these bandages. For a bag of 2" rolls, use order #2242, for 3" rolls, use order #2244, and for 4" rolls, use order #2247. These also come

in 1" and 6" sizes. Another brand is **Elastomull®**, made by Beiersdorf. For the 1" rolls, use order # 02088000. Cypress Medical Products (McHenry, IL 60050) also makes a conforming gauze that is also latex-free. For the 4" rolls, use order # 41-54. Another brand is **Duform® Conforming Bandage** (Dumex U.S.A. Marietta, GA. www.dumex.com). For the 4" size, use order # 75124. Most brands of rolled gauze come in a variety of sizes, in addition to those listed here.

Padding is often necessary for areas like the knees, elbows, and toes of small babies learning to crawl. A useful product in this regard is **Kendall Kerlix™** (Large Roll). It is a 6-ply, loose-weave, rolled material similar to cheesecloth. It works well as padding because of its fluffy texture, but it is not recommended for use directly against the skin. It is a bit coarse and may cause irritation. It can be folded in layers for thicker padding, if necessary. For a 4" roll, use order #6715.

3e. Netting

Stretch Netting is super handy as the final dressing product that holds everything neatly in place. There are several sizes and brands of netting. Netting is an open-weave, tubular, elastic material that can be cut to any length needed. One brand is **Surg-O-Flex**, made by Surg-O-Flex of America, P.O. Box 34, Wheatland, Wyoming 82201, phone number 1-800-334-2592. Also see their website: www.surgoflex.netfirms.com. It is ordered by size. Size A is for small extremities, and so forth. Western Medical Ltd., (64 North Summit Street, Tenafly, NJ 07670) makes 2 netting products: **Surgilast®** and **BandNet®**. Go to www.westernmedical-ltd.com for more information. The order #s for **Surgilast®** are GL-701 for size 1 (very small), GL-702 for size 2, and so on. **Ezy Wrap®**, made by Professional Products, Inc., P.O. Box 589, De Funiak Springs, Florida 32433, is another brand. This comes in a variety of sizes from 1 through 11. For size 7, use order #04807. Yet another brand is **Se Pro Net**, made by Acme United Corporation, Medical Products Division, Fairfield, CT 06430. For a size 2, use order #53120.

Section 4. Other skin concerns

4a. Infections

It is fundamentally important to do everything possible to prevent infections and to act quickly in addressing infections when they do occur. Infections tend to worsen wound areas, slowing healing time and increasing the tendency to itch. Infections may also be life-threatening. Signs of skin infection include increased, thick, and foul-smelling drainage from the wound. The appearance of infection should be monitored and watched carefully. Fever is a good indicator of the seriousness of an active infection in an EB child. When treating with antibiotics, it is recommended to culture the area first, so that the proper antibiotic can be used. Antibiotics are best used sparingly, but it is very important to finish any antibiotic program prescribed. Stopping an antibiotic treatment short may give bacteria the chance to multiply, and these new bacteria often have a stronger resistance to antibiotics.

4b. Body Temperature Considerations

In all forms of EB, it is important to monitor body temperature. Most EB patients lose large amounts of body fluid through wound drainage. This can lead to dehydration. In addition, children with lots of bandages may become overheated. Obviously, the problems of heat stroke and dehydration are more likely in warmer climates. Encourage your child to drink lots of water. Always watch for signs of overheating in your child and take their temperature often. This will give you an idea of their baseline temperature. Do not take your child's temperature rectally. Instead, carefully use an ear thermometer, but do not pull on the ear. Taking the temperature gently under the armpit is another method, however, it is less accurate. Tell your physician how the temperature was measured when communicating your concerns.

4c. Itching

Itching is a major problem in all forms of EB. All of the causes of itching remain a mystery, but as wounds heal, or as infections flare up, itching can become extremely troublesome. Keeping the skin well moisturized can be helpful. Also, as wounds heal, they often start to get dry and crusty along the edges. Carefully trimming away these areas of dead, dry skin with very clean scissors is helpful. Over-the-counter anti-itch sprays and creams should be used under the supervision of your physician. In some cases, prescription medications may be used as well.

Section 5: Nutritional Concerns

Nutritional needs should not be overlooked in the care of an EB child. Good nutrition is fundamentally important for all children, but it is even more important for a child with EB. The EB child has nutritional needs similar to those of a patient with skin ulcers or burns. Extra protein and calories are required to help the skin regrow. Protein loss and fluid loss from blistering and skin breakdown also contribute to these demands. However, simply getting an EB child to eat more is a definite challenge as blisters and sores in the mouth and throat often make feeding painful.

5a. Feeding Tips

Attention to the nutritional needs of an EB child should begin at birth. Blistering in the newborn period may produce fluid and protein loss that should be monitored closely. Unless the baby requires isolation for medical reasons, bonding with the mother should be encouraged and will help in the early feeding process. Oral bottle or breast feedings should be encouraged as soon as the sucking reflex is apparent, unless the doctor suggests otherwise. EB babies may demonstrate difficulty in sucking due to blistering and pain in the mouth and throat. A **Haberman® Feeder** can be helpful in these cases. This special feeder was developed to help children born with cleft palates. The nipple contains a disc insert and valve that helps control the flow of liquid and lessens vacuum within the bottle. Alternatively, a larger slit or additional holes in a soft, silicone nipple such as a preemie nipple may be helpful. A rubber-tipped medicine dropper could also be used.

For older children, introducing many tastes and textures is important. Mouth and throat involvement that produces discomfort can cause EB children to develop an aversion to foods. Keep experimenting to find foods your child will eat! In the more severe forms of EB, the mouth and esophagus (food pipe) are often heavily involved. Soft foods should be encouraged. Hard or sharp foods, such as corn chips, may further damage the delicate oral tissues. Pureed foods are a good idea and can be made even easier to swallow if additional liquid is added. Remember that a mouth with sores will be more sensitive to temperature extremes, so avoid serving foods that are too hot or too cold. Acidic foods should also be avoided. During family meals, make sure there is some form of food that is both easy and appealing for the EB child to eat. It is important to encourage them to participate in eating meals, even if only small quantities are consumed. Frequent feedings and snacks are needed to keep nutritional needs met.

5b. Supplements

For many EB children, eating enough regular food to meet nutritional needs is difficult and supplements may be required. There are many ways to help boost caloric and protein intake as well as vitamin and mineral requirements. Carnation® Instant Breakfast® can be added to whole milk or canned supplements such as PediaSure®, Ensure®, or Nutren®, to name a few. These canned supplements can also be taken separately and are generally prescribed according to the child's age and nutritional needs. Fortified milk can be made by adding powdered milk to whole milk. This can be added to soups,

sauces, cereals, puddings and so forth, boosting the calories, fat and protein consumed. If milk is not tolerated, other nutritional supplements may be recommended. Vitamin and mineral supplements may also be recommended by your dietician or physician. Be skeptical of any dietary regimen or supplement product that promises miraculous results. While these approaches are often attractive to parents of children with chronic diseases, alternatives to a varied, nutritious diet can result in malnutrition.

5c. Feeding Tubes

Even with our best efforts, it is often difficult or impossible to keep up with the nutritional needs of the severely-affected EB child. Placing a feeding tube into the child's stomach may be necessary. This procedure has been performed with great success and has helped many patients improve their general health and healing. Discuss whether your child may benefit from a G-Tube with your dietician / nutritionist and doctor, especially a gastrointestinal doctor if possible. Children with G-Tubes can be fed throughout the night by way of a pump that slowly drips a controlled flow of formula directly into the stomach via the G-Tube. The placement of a G-Tube often seems overwhelming, but it can be truly beneficial and may be removed when your child no longer needs to have it.

Section 6: Anemia

Anemia (decreased oxygen-carrying capacity of the blood) is a common complication seen in all forms of EB. Many factors contribute to the development of anemia, the most common being chronic loss of blood and fluids due to blistering and open skin lesions. Limited diets and poor intake and absorption of blood-building substances also contribute to this problem. Iron supplementation may be an effective treatment for mild forms of anemia. More severe cases may require other medications or blood transfusions. In these cases, it is necessary to have your physician and possibly a hematologist (blood specialist) involved.

Iron supplementation can significantly contribute to the problem of constipation (**see Section 7**). If your child is on iron, encourage him/her to drink plenty of liquids.

Section 7: Constipation

Constipation may be a difficult problem for children with severe forms of EB and deserves attention as a distinct complication. It is important to address constipation issues as soon as they arise. Several factors contribute to constipation, and its psychological implications should not be overlooked. Like the mouth and esophagus, the rectum is delicate in children with EB, and both it and the surrounding skin may have damage and open wounds. This leads to severe pain during bowel movements, and children will then intentionally hold back from having a bowel movement, worsening the constipation. A combination of dietary modifications, psychological support, and supplements or medications will help your child avoid constipation.

7a. Diet and Nutrition

A diet with a wide variety of foods from all food groups is extremely important. Include fruits, vegetables, whole-grain breads and cereals, dairy, meat and poultry products, and legumes. Most of these foods can be prepared in a manner that is tolerable to the EB child's fragile mouth and esophagus. Try to increase the amounts of food high in fiber, including vegetables and fruits, such as prunes, in your child's diet. Encouraging a high intake of fluids such as water and juice, is also a helpful way to reduce constipation. Soups are a wonderful way to provide many nutritious and high-fiber foods in a manner that is easily tolerated by children with EB.

7b. Supplements and medications

There are many products on the market today for the relief of constipation. Your gastrointestinal doctor is a great resource for recommending the appropriate supplement or medication, regarding your child's age, weight and individual problems. The use of supplements should be closely monitored so that the child does not become dependant on them. When possible, use dietary modification and fluid supplementation to try to correct the problem first.

7c. Physical activity

Many children with severe EB become less active. This may contribute to constipation and other complications (see **Sections 8a and 8b**). Whenever possible, encourage your child to walk, play, and be as active as their individual condition allows. It is better to have a child who plays and may get a blister or skin injury now and then, than to have a child who is afraid to move and ends up with a variety of other complications. Playing is a normal part of childhood, and while most physical wounds can be dressed and treated, emotional wounds are difficult to heal.

Section 8: Additional Complications and Considerations

This section briefly touches on some of the other complications seen in the severe forms of EB, especially recessive dystrophic EB. Although they are not common to all cases, it is good to be aware of these potential problems so that your child can be observed and monitored, helping to prevent them as much as possible.

8a. Contractures and Deformities

Contractures are shortened, tight areas of skin, ligaments, and tendons that often lead to decreased function in an extremity. Decreased function leads to decreased use, or even nonuse, which also contributes to weakening or atrophy of the muscles. Keeping your child active prevents muscle atrophy and helps to slow the development of contractures. Encourage your child to play and participate in normal childhood activities whenever possible. Swimming is a wonderful sport for EB children and is a good form of exercise. It may be necessary to let your child swim in his/her bandages. This may actually make wound care easier, as it helps release sticking bandages, and the chlorine in swimming pools is a wonderful antiseptic. Normal activities and playtime are also good for your child's mental health. Adequate bandaging, with extra padding at the knees and elbows will help avoid injury, and bandaging, especially of the hands, may also help prevent contractures. In some cases, physical or occupational therapy may be necessary to lessen the complications of contractures.

8b. Fusion of Fingers and Toes (Syndactyly)

Repeated friction, blistering and scarring between the digits leads to fusing or webbing between the fingers and toes called syndactyly. This is often accompanied by down-curling of the digits as well. This type of scarring can be very aggressive and is one of the most debilitating effects of EB in terms of normal day-to-day functions. In the most severe form of syndactyly, complete encasement of the fingers and toes (mitten deformity) renders them useless. Although this is less critical on the feet, it is extremely disabling for the hands. While not always completely avoidable, there are methods that may prevent or dramatically delay this complication. A method of wrapping the hands and individual fingers with tension in the web space is helpful. Important points to remember when wrapping the hands are to always wrap up the fingers from the back of the hand and to try to keep the fingers as straight as possible during the wrapping. This helps support them and encourages them not to curl down into the palm. Using a 1" roll of gauze, this task becomes simple and routine with practice. Children will grow to appreciate the wrapping and the added protection it provides. It is best to start hand wrapping at an early age in the severe dystrophic forms of EB. This helps make hand wrapping a comfortable part of your child's day-to-day routine and will help slow or stop webbing before it has a chance to begin. **Click on this sentence to see streaming video of a hand wrapping method.** Surgery to release the thumbs and fingers, if necessary, should be performed by a hand surgeon familiar with EB.

8c. Eye Problems

In certain forms of EB, particularly the dystrophic forms, the cornea (the clear outer layer of the eye) and the conjunctiva (the mucous membrane covering the eyeball and the underside of the eyelids) can be damaged. These tissues contain the same inefficient connecting fibrils as the skin and easily tears and scars. For example, rapid eye movements that occur during sleep cause friction in the eye, especially if there is not enough moisture present. This results in scratches and/or tears of cornea. This is extremely painful and may take several days to heal. Similar to skin wounds, eye wounds may become infected as well. EB children with eye involvement may have excessive tearing or discharge from the eyes. A regimen of eye ointments or eye drops helps add lubrication to the delicate eye tissues. This is especially helpful at night. Antibiotic eye ointments are sometimes necessary if an infection settles in the eye. Patching the eye can be helpful when an injury to the eye occurs, but it is important not to damage the surrounding skin when securing the patch. It is advisable to have an annual check-up with an ophthalmologist if these problems are present.

8d. Dental Problems

Cavities and other tooth problems occur in several forms of EB for a variety of reasons. Many EB children have their tongues fused to the bottom of their mouths, making it difficult for them to move food and saliva, a natural rinsing agent, around. This retained food is a good environment for the development of cavities. Avoiding sugary substances is recommended, but this is often difficult due to the need for high caloric intake. Another common difficulty of oral hygiene for all forms of EB is the sensitivity and fragility of the oral tissues. Gentle brushing with a very soft toothbrush or foam brush is highly recommended, but be especially careful not to damage the gums and tongue. Fluoridated water or fluoride supplements are recommended. Regular dental care is important, but this must be extremely gentle. A dentist who is familiar with EB will better understand your child's needs. Tooth sealants may be helpful in preventing cavities, and this should be discussed with your dentist.

8e. Immunizations

Immunizations are designed to prevent common childhood infections that are potentially dangerous or deadly. These "shots" help keep our children healthy. Children with EB should handle immunizations as well as a normal child. Every child, including those with EB, should receive regular, scheduled immunizations.

Section 9: Genetic Implications

It is estimated that there are approximately 25,000 people in the United States suffering from one of the inherited forms of EB. There is also a non-inherited form of EB that is caused by auto-immune damage to certain structures in the skin, although this is not very common.

The inherited forms of EB are caused by abnormalities in the genes that produce certain structural proteins in the skin. A gene is like a blue-print or program that provides the information for how a protein is made. If the information is faulty, then the protein will not work correctly. In the case of EB, faulty genes lead to proteins that do not hold the layers of skin together properly. In general, for every protein in the body, there is a set of 2 genes. For some diseases, only one gene needs to be abnormal to produce disease. These are called **dominant** disorders, and they may be passed to the child from an affected parent. Other genetic diseases, called **recessive** disorders, are caused by abnormalities in both genes of the set. In these cases, parents may be **carriers** of one abnormal gene, even though they do not have symptoms of disease. Affected children receive one abnormal gene from each parent who is a carrier of the abnormal gene.

The simplex forms of EB are typically dominantly inherited.

Junctional EB is recessively inherited.

Dystrophic EB has both dominant and recessive forms.

The dominant forms of EB are more common than the recessive forms, which are typically more severe as well. It is important for parents of an EB child to understand that there is a significant chance for subsequent pregnancies to result in another child with EB. The likelihood varies with the different forms of EB, but in general, there is a 50% chance of passing a dominant form of EB to the baby, while for the recessive forms of EB, the chance is 25%.

Unaffected brothers and sisters of children with a recessive form of EB may also be carriers of the disease. If this is the case, they could pass the disease to their children if their partner also carries an abnormal gene. Depending on the type of EB, affected individuals are likely to pass the abnormal gene to their offspring as well, resulting in a carrier or a child who is affected by that type of EB. Genetic counseling is strongly advised for families affected by EB who are considering additional pregnancies.