

Tips for Educators

Suggestions:

Meet with the parents of the student with EB to review both videos and discuss a school plan for the student. Address, in a written plan, the issues presented in “Your Welcoming Classroom.” Discuss with the parents and student when to show the movie “What is EB? in the classroom and decide who (including any medical professionals) will participate in a question and answer period with the class.

Understanding the Issues of Importance for students with EB

Swallowing

Esophageal blisters or scarring

Respect “no” if unable to eat

Have snacks available (i.e., puree foods, liquids)

Bandages

Protect skin

Help heal skin

Overheating

Odour

Help other students understand

Speak with nurse and/or parent

Be aware of infections

Depression

Be aware of psychiatric symptoms (i.e., withdrawn, isolated, sadness)

Talk with parents

May need psychiatric evaluation and support

Pain

Pain control management

Rate pain, on scale 0-10

Pain Medications

Socialization

Welcome EB students and involve them in activities
Educate other students about EB (i.e., go to different classrooms)
Educate other students about abilities and limitations
How can other students help?

Bathroom

Set up bathroom time (i.e., 10 minutes)
Reward program or goal sheet
Parents
Communicate regularly
Written back and forth log
Common knowledge and goals

Control

Need sense of control
Self-advocacy
Recognizing talents and strengths

Good Days and Bad Days

Status Fluctuates
Pain
Schedule time to check in with the nurse
Allow breaks

Answers to most commonly asked questions about Epidermolysis Bullosa

What is Epidermolysis Bullosa (EB)?

EB is a rare genetic condition in which the skin, and sometimes the mucous membranes (such as the lining of the mouth) blister in response to mild friction or trauma. In the most severe form, called recessive dystrophic EB, the child inherits from both parents an altered gene needed to make the collagen that anchors the layers of skin together. Because the collagen is defective, the outer skin layer separates easily from the deeper layer, forming a blister and a sore.

Are there different types of EB?

There are three main forms of EB: EB Simplex, Junctional EB and Dystrophic EB. These different subtypes are defined by the depth of blister location within the skin layers. With EB Simplex, blistering may be localized to the hands and/or feet, or may be generalized and affect the entire body. While blistering can be continuous, the skin heals without significant scarring. There may be few to no lesions visible but the child may face considerable problems of immobility due to the potential for or the actual development of blisters. Frequently, Junctional EB is fatal in infancy or early childhood. However, there are some forms of Junctional EB which are not fatal. These children develop blisters and heal with scars. They may have significant mobility

problems and may have airway involvement, as well. The child with Dystrophic EB has very obvious damage to his or her skin. Blistering is more likely to be extensive, both internally and externally, and heal with scarring. Fingers may fuse and contract, causing mitten deformities of the hand. Feet may be similarly affected. Under the clothing the child may have very extensive blisters and erosions which require lengthy and painful daily skin care regimes including extensive bandaging and wrapping.

Is EB contagious?

No, EB is a genetic disease that is not contagious. However, while EB is not contagious, it is important to remember that blood and body fluid precautions (Universal Precautions) must be maintained at all times. This presents a special problem, especially in preschool and in the younger grades, where children share art supplies, toys and often hold hands. If a child with EB has frequent hand lesions, it is advised that the child have his/her own "tools" with which to work. Parents must be mindful that the school is obliged to protect other students. Student with EB may not come to school with open, draining wounds that are not bandaged. The exception to this is if the lesions are located on the face or neck, where bandaging is difficult. When situations such as this arise, they should be handled on a case-by-case basis involving the parent, the teacher and the school nurse or administrators.

Why do children with EB have so many bandages?

The bandages protect the skin from friction and prevent blisters. They also cover areas that have erosions to reduce the risk of infection and reduce pain. However, these bulky bandages are uncomfortable and seal in body heat so that a child with EB gets overheated easily, especially in warm weather.

Should the school nurse have dressings or other material?

Parents may be asked to provide an "EB kit" of dressings, with an explanation of procedure, just in case an injury occurs at school. Parents should also provide telephone numbers of people who can be contacted in an emergency. However, it has been found that most children with EB cope remarkably well with school life and quickly learn how to avoid injury and trauma.

How can we lift a child?

When handling any child with EB, remember that friction may cause blisters and skin damage; direct pressure usually will not. Children with EB must never be lifted from underneath the arms, as this will likely result in painful blistering and this area is very difficult to bandage and heal. If lifting is required from a sitting position, ask the child to lean forward slightly and place one hand under the bottom and the other behind the shoulders and lift in one movement.

Will hand splints help prevent finger flexion deformities in children with EB?

Not all children with dystrophic EB will develop flexion contractures of the fingers or pseudosyndactaly (webbing) of the fingers. Although a large number of different splints have been tried with children with EB, there really are no splints that prevent flexion. It is important to remember that what causes sores to develop on the skin is friction. Using splints may not be helpful as they may cause friction and more blister formation.

Are there adaptive devices that help children with EB and do activities such as writing, cutting, etc?

Often children with EB have difficulty with strength in their hands. Using keyboards for written assignments is very helpful. Adapting cutting activities by using loop scissors that open by themselves may be helpful. A consultation with an Occupational Therapist is very helpful when dealing with these issues.

Can the student with EB participate in P.E. and Games?

The objective should be to allow the child to participate as much as possible. For floor activities, the child can stand, sit or lie on a foam mat. Blistering is precipitated by friction so exercises involving kneeling, running or jumping may be problematic. Games involving hand holding and fast activity may also be difficult. As a rule, the parents and the child should be allowed to educate the school as to the types of activities that can be performed without the development of blisters. This may include modifying the physical education requirements or excusing the child from all formalized physical education activities.

What are the physical limitations of a student with EB?

Children with severe EB may suffer from weakness or fatigue for many reasons including anemia, pain and the increased nutritional demands necessary for wound healing. Blistering and contractures of the feet, knees and hips may make walking difficult. Children may need to limit the amount of walking they do at school. Similarly, writing may be difficult due to joint contractures, webbing of the fingers and pain. Children with EB will have good days and bad days. It is important that they stay as active as possible, but without being pushed beyond their comfort level. An exercise program at school should be flexible depending on their pain and sores. Consider coordinating the school PT/OT with the child's EB medical team.

What activities can a child with EB participate in without increasing blistering?

This varies tremendously from child to child. Usually the child knows what activities he or she can participate in safely. However, activities that have a high friction component should be avoided, such as placing hands on rough surfaces including playgrounds and highly abrasive rugs. Occasionally, a child with EB will benefit from having custom made gloves for their hands that they can wear during these activities to protect their skin.

What foods can children with EB eat without hurting their mouths and throats?

Some children can eat anything without any difficulty. However, there are some children who will develop sores in their mouths and throats if they eat anything that is rough or has sharpness to it. Foods that melt in the mouth are good, such as Gerber Graduates® Lil' Crunchies™. These are very much like cheese puffs but they literally melt in the mouth. Cooked veggies such as carrot sticks can be used for dipping instead of chips or pretzels. Talking with the child and parents about finding foods that the child can eat; so that he or she can have fun foods, and participate in activities such as school parties, etc., is important.

When does the student need a gastric tube for feeding?

Unfortunately eating is not easy for a child with EB. Painful sores in the mouth and blisters in the upper part of the esophagus can occur. The blisters in the esophagus cause scars that narrow the opening and make it difficult to swallow. Children with EB also may have abnormal teeth. Cavities and tooth abscesses are common since EB weakens the enamel and gum sores make tooth brushing painful. When a child with EB cannot eat and drink enough food to maintain proper nutrition, a special gastric tube may be placed through the skin of the abdomen directly into the stomach. A feeding tube can be attached to the gastric tube so that a nutritional supplement can be

administered either by gravity or with a pump. An advantage of this system is the ability to give continuous night time feeds using a pump. These night time pump feeds are less disruptive to normal activities than daytime supplemental feeding. The tube can also be used for medications, such as laxatives and vitamins that have a bad taste.

How much should a child with EB be encouraged to move?

Children with EB have varying degrees of pain depending on their sores and blisters at the time. When a student with EB usually walks around the classroom or in the halls but occasionally prefers not to walk, ask if they are having pain and honour their wishes.

Why do I detect an odd smell from my student with EB?

Students with EB have open and healing wounds of all stages of healing all the time. Since their wounds heal more slowly than wounds in normal skin, they seem to have an odour. This is extremely difficult to prevent. If, however the odour becomes more pronounced, discussing this with a parent will help them treat a wound that may be critically ``colonized`` or ``infected`` and decrease the odour. Adding 1-2 lbs. of swimming pool salt to the student's bath at home may reduce the skin odour, help the sores to heal faster, and reduce the stinging and pain caused by the water in the bath.

Why is scheduling longer bathroom breaks for my student with EB important?

Due to many layers of dressings and some decrease of their dexterity, bathroom time may take more time for the EB student. A child with EB may resist going to the bathroom if they are constipated. Allotting more time for the child to use the bathroom allows him or her to relax, be less anxious and have less pain.

Publications/Website

DEBRA Canada produces newsletters and e-blasts and distributes them to 600+ registered members. Copies of archived newsletters are available for download on the DEBRA Canada website, www.debracanada.org. Visit our website for information/resources on EB.

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