

EPI DERMOLYSIS BULLOSA

A GUIDE FOR PARENTS AND SCHOOLS



DeBRA of America, Inc
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EPIDERMOLYSIS BULLOSA

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This booklet is intended to be a guide for parents and schools when a child with Epidermolysis Bullosa (EB) is of school age. All children are individuals; their needs, aptitudes and strengths will vary considerably. The best guide to a child's needs are the experiences of the parent and the child.

Above all, children with EB are just that; children who happen to have EB. They are, in all other respects, normal children and students.

A Note to Teachers

Many schools and preschools facing the prospect of teaching a child with EB for the first time will understandably feel concerned.

- What is this condition which they have never heard of before?
- Will the child be able to cope and keep up?
- Will the crowded classroom be too dangerous an environment?

Many of your colleagues have been there before you, have shared the same concerns. The overwhelming majority of children with EB attend public schools successfully. Teachers, staff, volunteers and the other students quickly learn the "do's and don'ts" of working beside a child with EB. Understanding and meeting the unique needs of an EB child becomes part of daily classroom life. We hope this booklet will help you learn strategies and skills to meet EB children's needs, just as you identify and meet the individual needs of every other child in the class. Integrating the child with EB can become a natural part of the school experience for all involved.

Some children with EB will need extra help in the classroom, others will just need the teacher to be aware of their special situation and plan accordingly. This booklet gives an overview of some of the issues and suggests the ways in which these issues can best be dealt. The parent(s) will be the primary source of information about how EB affects their child.

What is Epidermolysis Bullosa?

Epidermolysis Bullosa 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. A genetic defect prevents the layers of the skin from adhering properly. Blisters form as the layers of the skin split apart in response to friction or trauma.

This condition is not contagious.

An estimated 1 out of every 50,000 Americans is born with some form of EB. The disorder occurs in every racial and ethnic group throughout the world and affects both sexes equally.

There are three main forms of inherited 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. These children frequently have a hidden disability. 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.

There are various forms of Dystrophic EB. Blistering is more likely to be extensive, both internally and externally, and heal with scarring. The process is cumulative so that the child with Dystrophic EB may have very obvious damage to his or her skin. 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.

Mild/Severe

Each subtype may range from mild to severe. Because of this, the terms mild and severe will be used rather than referring to the specific subtype. Classification of severity is defined by medical information and the patient's own perception.

Treatment of EB

Although there is no cure for EB, many complications can be minimized or avoided through early intervention. In all cases, treatment of EB is directed toward the symptoms and is largely supportive. This care should focus on prevention of infection, protection of the skin against trauma, attention to nutritional deficiencies and dietary complications, minimization of deformities and contractures, and the need for psychological support for the entire family. Many persons with milder forms have minimal symptoms and may require little or no treatment.

All children with EB need special skin care. Families develop a daily routine of inspecting the skin and taking necessary action. In the mildest forms of EB, this will involve identifying new blisters, draining them and keeping the affected area clean. In the most severe forms, where large areas of the body are affected with numerous new blisters daily, care is very time consuming, often taking hours each morning and/or evening. This may involve soaking off dressings which have stuck to wounds, draining blisters, bathing and applying ointments and several layers of non-adherent dressings.

It is not difficult to imagine the stress and pain a child (and parent) may have suffered before arriving at school in the morning. Even the child with very mild EB may come to school with a couple of newly drained blisters and several healing ones which, although not visible in school clothes, may be causing considerable discomfort

and limitations of mobility.

Starting School

Parents are encouraged to contact the local the local school system's central office prior to their child's 3rd birthday. They should consult with the Exceptional Children Preschool Coordinator and explain their child's diagnosis and needs and request a screening evaluation. Parents should provide literature explaining what EB is and how it affects their child as well as documentation from the physician confirming the diagnosis. Upon request, the Coordinator will guide the family and teachers through preschool and will schedule a transition meeting for the parents prior to kindergarten and "pave the way" for beginning elementary school. Depending on the exact situation and the screening results, the Coordinator and the parents will decide on an appropriate plan for the child. This may include developing a 504 Plan in order to address accommodations and modifications of the environment or an IEP (Individual Education Plan) (<http://www.ed.gov/parents/needs/speced/iepguide/index.html>) which addresses special education or environmental needs.

Within the public school system, the term 'special education' means specially designed instruction, at no cost to parents, to meet the unique needs of a child with a disability. This includes instruction conducted in the classroom, in physical education, in the home, in hospitals and institutions, and in other settings.

The term 'supplementary aids and services' means, aids, services, and other supports that are provided in regular education classes or other education-related settings to enable children with disabilities to be educated with nondisabled children to the maximum extent appropriate in accordance with the law.

The Individuals with Disabilities Education Act states "...To the maximum extent appropriate, children with disabilities, including children in public or private institutions or other care facilities, are educated with children who are not disabled, and special classes, separate schooling, or other removal of children with disabilities from the regular educational environment occurs only when the nature or severity of the disability of a child is such that education in regular classes with the use of supplementary aids and services cannot be achieved satisfactorily..."

Most children with EB attend regular public school classrooms. As stated above, educational legislation clearly states that when possible, children with special education needs will be integrated into ordinary schools.

If the child has not been involved at the preschool age, some time during the spring prior to beginning kindergarten, the parents should contact the principal of the school the child will attend and request a meeting. At this point, the parents should provide the information mentioned above and, with the principal's help, decide on a plan that will meet the child's needs.

The parents should meet with the teacher and, if possible, the school nurse, prior to the beginning of school. This gives the teacher and nurse the opportunity to learn about EB, how it affects the child, how blisters are caused, the best ways to avoid them, and how to deal with them if they occur. The school staff will want to do what's best for the student with EB and the establishment of a partnership with the

parent(s) will help all concerned. During this early meeting, strategies and modifications should be discussed. At this time, you may want to provide an "EB kit" (gloves, ointments, dressing, doctor's orders) which may be used in an urgent situation. We also encourage children with EB to keep such a kit in his/her backpack.

It may be helpful for the parent(s) to visit the classroom during the first week of school and explain EB to the other students. If children understand why a child is wearing bandages or why a child is limited in activity, they are more supportive, kind and helpful. It may be necessary to attend a parent's meeting and provide education and literature to the parents, as well.

Preschool

Parents of a child with EB are encouraged to involve their child in pre-school education and to help their child develop social skills by mixing with other children. This can often be a time of considerable anxiety for parents as their child ventures outside the protected home environment.

Sometimes preschools and playgroups are reluctant to accept a child with EB because they feel they cannot cope with the special needs the child may have, they are worried that children's play may be too dangerous for the child with EB or the mistaken fear of contagion. These fears are understandable and thought must be given regarding how to deal with the perceived and actual problems, but they should not be allowed to prevent the child from getting the stimulation they needs. The parent(s) and staff must combine their efforts to create the safest environment possible. As pre-schoolers, children with EB must learn how to protect themselves, how to ask for what they need, how to function as part of a group and how to trust that someone other than the parent can care for them .

Again, the parent(s) are the best guides to what special arrangements may be needed in the preschool or school setting. They will have learned from experience the best ways of finding the right balance between stimulation and protection. It may be helpful for the parent to attend the first week or two of preschool or kindergarten in order to model interactions and activities which are tolerated by the child, as well as how to handle skin problems which are likely to arise during the course of a day. The child and parent will feel reassured, as will the teachers. In the long term, the child will benefit from the experience of being away from parents (as other children do) and it is important that the belief "I'm only safe with Mom and Dad" be modified as soon as practical. Coping with a minor accident which results in skin damage, while being traumatic initially for child and helper, can lead to great benefits as the child learns that other adults can cope with some of his/her care. In later childhood and adulthood this will be very valuable.

Day to Day Issues

Blood and body fluid precautions: 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 obligated

to protect other students. Students 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 between the parent, the teacher and the school nurse or administrators.

Practical care at school: Parents may be asked to provide an "EB kit" of dressings, with an explanation of procedure, just in case an injury occurs at school. The school will also want 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.

Lifting the 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 neck and lift in one movement.

PE and Games: The objective should be to allow the child to participate as much as possible. The school must be sensitive to those situations when an adaptation is necessary. For floor activities, the child can stand, sit or lie on a foam mat. Blistering is precipitated by friction so exercises which involve 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.

Physical limitations: Children with severe EB may suffer from weakness or fatigue for many reasons, including anemia and nutritional deficiencies. Blistering of the feet is enough to make walking painful and some children will need to limit the amount of walking they do at school. Similarly, writing may be slow due to joint contractures, pain and damage to the hands. The use of computers for schoolwork is helpful for some children. Referral to the school PT/OT services may be helpful.

Crafts: If recess and PE are restricted there must be other opportunities for leisure and enjoyment, such as arts and crafts and music. The hands of children with EB can be subject to constant stress and injury but a balance is needed between protection and participation. Adaptive devices are available through an Occupational Therapist. These may minimize damage and help to avoid friction on fingers and thumbs. Some aids, such as a foam-tube covering for pencils, may be of assistance. Whether clay, paint or other craft materials can be used may depend on the day to day condition of the hands. Parents will know when activities should be restricted and should advise when appropriate.

Middle and High School

Entering middle or high school brings an entire set of new issues into focus. On a practical level, the child will have to move from classroom to classroom for different subjects, which may cause more frequent blistering or may exacerbate existing mobility difficulties. In most schools, lockers are utilized and may be difficult for the child with EB to access. Carrying a heavy backpack is another challenge for the child with EB. Provisions will have to be made to accommodate the needs of the individual. The physical layout of the school may become an obstacle and when school choices are available, the parents will want to involve the child in the decision of which school he/she will attend.

On the social and psychological level, the child with EB is entering a new phase of development where he or she is expected to be more independent and responsible. Additional help, which may be needed, must be provided in a sensitive manner to avoid adding to the child's feelings of being different.

For the more severely affected student, there can be significant obstacles to overcome. Extended absences for hospital treatment may occur. Time must be found each evening for both homework and skin care. With commitment from the student, the parents and the school, these obstacles can be overcome.

All parents want the best for their children — education is important to us all. Children with EB, who may not be able to undertake manual occupations in adulthood need the qualifications and education which will enable them to have the broadest range of career options possible. A wide range of careers has been achieved by people with EB and the condition should not be a barrier to higher education and training.

Strategies to address the daily challenge of being in school with EB

- Discuss the child's dietary requirements with school staff if the child has difficulty swallowing. The school should be made aware of the child's swallowing problems and should be cautioned about offering various foods, such as chips, crackers, nuts, etc. Parents may choose to provide a packed lunch/snack or dietary supplement but, where school lunches are chosen, a soft alternative may be required. Ice cream is usually a favorite!
- Avoid crowds and crush situations by allowing the child with EB to be first or last in line. If the child has very severe EB it may be appropriate for the child to arrive and leave a few minutes before or after the rest of the class.
- Check the classroom for obstacles which may cause tripping and falling.
- Consider how best to explain EB to the rest of the class so that they are aware of why they should not push or grab from the child with EB.

- Inspect classroom furniture for sharp edges so that a change can be made or extra care exercised. In the case of a severe form of EB, the child may bring a piece of sheepskin or a pillow to place on the chair.
- Avoid placing the child next to a heater, heat vent or next to a sunbaked window since heat can exacerbate blistering.
- Consider whether the requirements of PE class will have to be modified or eliminated.
- Think about recess and whether the child will need an uncrowded area and, if so, how this can be achieved while maintaining socialization.

Suggestions for modifications for students with EB

- Teacher's aid to assist the child - full or part time as needed
- Student partner or aide to carry backpack
- Air conditioned classrooms
- Elevator as needed
- Extended time for tests
- Homework modifications (e.g. student will do half of assigned math problems and not be penalized)
- Class notes provided by the teacher to the student
- Home instruction for long periods of illness
- Dismissal from class 5 minutes early so student can navigate in empty halls and avoid crowds
- Tape record lectures
- Books on tape provided to the student who experiences frequent eye erosions
- Arrange classrooms to be adjacent or in close proximity when possible
- Evacuation plan for fire drills and emergencies
- Tutor/home instructors, as needed because of frequent absences (especially in middle and high school. Tutors/home instructors may be needed in the home on a regular basis for challenging courses such as math or science.
- Modify the school day. During middle and high school, schedule PE and other electives during first or last period so the student may come late or leave early if needed without missing the core curriculum.

Beyond the scope of this booklet, advice and support is available through DebRA of America.

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