

# Epidermolysis Bullosa: A Guide for Parents, Schools and Playgroups

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DEBRA is the national charity that supports individuals and families affected by Epidermolysis Bullosa (EB) – a painful genetic skin blistering condition.

A charity registered in England and Wales (1084958) and Scotland (SC039654)

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*Please note: minor amendments were made to the text in this edition in August 2014 and this booklet now incorporates the 'Guidelines for Playgroup (2011)' information sheet.*

## **Introduction**

This short booklet is intended to be a guide for parents and schools on the issues to be considered when a child with Epidermolysis Bullosa (EB) is of school age. It is by no means exhaustive and it must be read with the recognition that all children are individuals and their needs, aptitudes and strengths will vary considerably. There are a number of different forms of EB with a wide variation in symptoms and severity. The best guide to an individual child's needs is the experience of the parent(s) and, where of an age to express a view, the child him/herself.

Above all, children with EB are just that; children who happen to have EB but are, in all other respects, the same as any other child or pupil.

## **A Note to Teachers and Playgroup/Nursery Supervisors**

Many schools and playgroups facing the prospect of teaching a child with EB for the first time will feel understandably nervous. What is this condition of which they have never heard before and about which they are hearing so many dire warnings of the vulnerability of their new pupil's skin? 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, shared the same concerns and made it work. The overwhelming majority of children with EB go to mainstream schools and do well. Staff quickly learn the 'do's and don'ts' of looking after a child with EB and it becomes part of daily classroom life, just as the individual needs of every other child in that class are recognised and met.

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 makes suggestions about how they can be dealt with. The parent(s) will be a mine of information about how EB affects their particular child and will be able to pass on what they have learned. DEBRA is also more than happy to offer support and information.

## **What is Epidermolysis Bullosa?**

Parents reading this booklet will already know what Epidermolysis Bullosa is and will be able to give to the school detailed information about how the condition affects their child. However, the following brief explanation may be of assistance to school staff.

Epidermolysis Bullosa is a genetic condition in which the skin, and sometimes the mucosal linings of the body (such as the oesophagus), blister at the slightest knock or rub. The skin layers do not adhere properly so that friction causes them to part and blister. It would be difficult to exaggerate the vulnerability of the skin in the most severe forms of the condition.

There are a number of different forms of EB which can be broadly categorised into three main groups: EB Simplex, Dystrophic EB and Junctional EB. Junctional EB can be fatal in early childhood so children with this type of EB may not reach school age. However, there are some forms of Junctional EB which are not fatal and advice can be obtained by schools on these children's needs from their parents, their EB Clinical Nurse Specialist or the DEBRA office.

This booklet concentrates on children with EB Simplex and Dystrophic EB.

### **EB Simplex**

In these forms of EB blistering may be localised to hands and/or feet, or generalised so that the whole body can be affected. Whilst blistering can be continuous, the skin does heal normally. Blistering may occur in the mouth or throat and, sometimes, a child who is not visibly very affected can be in considerable discomfort internally.

These children frequently have a hidden disability – they look perfectly 'normal' but face considerable problems of mobility due to the pain of blistered feet.

## **Dystrophic EB**

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. Feet may be similarly affected. Under their clothing the child may have very extensive blisters which require lengthy and painful skin care night and morning.

### **Mild/Severe**

In general, Dystrophic EB is seen to be more severe than EB Simplex. However, there are forms of EB Simplex which have unusually severe symptoms and forms of Dystrophic EB where the symptoms are not unduly intrusive.

As a consequence, the terms 'mild' and 'severe' will be used in this general booklet rather than referring to the diagnostic categories. The experience of the parent(s) and the school will soon determine what is relevant to a particular child.

### **Skin care**

At present there is no cure or effective treatment for EB, although there are good grounds for optimism that this will change within the next few years. Current treatment involves protecting the skin from injury and infection and limiting the extent of blisters.

All children with EB need skin care which is not required by other children. Families develop a daily routine of inspecting the skin and taking any necessary action. In the mildest forms of the condition this will involve identifying new blisters, bursting these so that the blister fluid can escape and keeping the affected area clean. In the most severe forms, where large areas of the body are affected with numerous new blisters appearing every day, the process will be very time consuming, often taking over an hour each morning and evening. This may involve soaking off dressings which have stuck to the skin overnight, bursting blisters, bathing the child, drying very gently and applying several layers of different types of dressings.

It is not difficult to imagine the stress and pain which a child (and parent) may have experienced before arriving at school in the morning, or will face in the evening. However, even the child with very mild EB may be coming to school with a couple of newly burst blisters and several healing ones on the feet which, although not visible in school clothes, will be causing considerable discomfort.

### **Pre-school**

Parents of children with EB are encouraged to involve their child in pre-school education at playgroup or nursery class 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 makes the first steps outside the protected home environment. However, it is essential that the child with EB has as much pre-school experience as possible. The value of pre-school education for all children is generally accepted and is perceived as important by most parents. This is even more the case for the child with EB.

In some areas there is a shortage of good quality pre-school facilities. Sometimes, playgroups may be reluctant to take a child with EB because they feel that they cannot cope with any special needs which the child may have, or are worried that the 'rough and tumble' of young children's play may be too dangerous. These fears are understandable and thought must be given to how to deal with the perceived problems, but they should not be allowed to prevent the child from getting the stimulation he or she needs. After all, the pre-school child will rapidly become the schoolchild, the teacher and the adult; they cannot be protected all of their life and they will need a good start.

Again, the parent(s) are the best guides to what special arrangements may be needed in the pre-school setting and will have learned from experience the best ways of finding the right balance between stimulation and protection.

It is always tempting to ask a parent to be present whilst their child is at playgroup, so that he or she is on hand in case of problems and the child and parent feel reassured. For some children this may be the best option, at least in the short term. However, in the longer term, the child will benefit from the experience of being away from parents (as other children do) and it is important that the belief that 'only Mum can do my dressings' is modified as soon as it is appropriate. Coping with a minor accident which results in skin damage, whilst being traumatic initially for child and helper, can lead to great benefits as the child learns that other adults can cope with some of their care. In later childhood and adulthood this will be of crucial significance.

## **Starting school**

### **Mainstream schooling?**

Some parents or Local Education Authorities (LEAs) may ask themselves whether a particular child with EB can cope with attending a local primary school or whether a special school is needed. A general booklet such as this cannot deal with the specific situation of every child and each family will have its own view. However, it is exceedingly rare for children with EB to be unable to manage in a mainstream school and education legislation emphasises the duty of the LEA to integrate children with special educational needs into mainstream schools unless there are particular reasons why this is not desirable. The wishes of the parent should always be given special consideration by the LEA.

### **Special Educational Needs**

Whilst a mainstream school will probably be the best option, it may be that a child with EB will need some particular forms of help to get the best out of school. Where an LEA feels that a child may require special educational provision it can decide to undertake an assessment to determine this. Parents may also request an assessment.

Following assessment, the LEA may feel that it is necessary to make a Statement of Special Educational Needs which defines the Authority's view of the special educational needs and the special provision which it believes is necessary to meet those needs. This may cover the type of school recommended or any special help or aids needed. Computer facilities, for example, may help children with severe EB in some situations.

This process, known as 'statementing', can be complicated and advice is available from DEBRA if parents so wish. DEBRA is always willing to discuss the requirements of children with EB with school and Educational Department staff.

### **Selecting the right school**

Unless they have already been in touch with the LEA during pre-school years, and have discussed special provisions, the normal procedure is for the parent(s) to contact the primary school of their choice and talk to the Head Teacher about their child when he or she is three or four years old. It is unwise to leave this any later since special arrangements, if needed, can take months to arrange.

Parent(s) and school staff may wish to consider some of the following factors, depending on how severe the form of EB the child has:

1. Is the school building spacious or overcrowded, affecting the possibility of bumps and falls?
2. Is the school building on one level, or are there many steps and stairs to manage?
3. Is the playground suitable with a quieter area available, if needed?
4. Would there be the possibility of the child staying indoors with one or two school friends if his or her skin is in poor condition or on windy days to avoid eye problems?

If the school and parent(s) are happy that the school meets the child's needs, consideration will have to be given to whether the school can manage with just giving extra attention or whether extra resources will be needed.

Some measures may be very straight forward, just requiring some forethought, for example avoiding crowds, adapting participation in PE and games or making simple adaptations to tools and equipment. Children with more severe forms of EB may need special arrangements to be made for them, such as the provision of a classroom helper to watch over the child and give help and 'protection' where needed. Some children may require special equipment to enable them to benefit fully from school activities. The LEA may consider making an assessment of special educational needs where this will be helpful in mobilising support and services for the child.

### **Early days**

Once the arrangements for admission have been made, it is helpful for parents, Head Teacher and Class Teacher to meet in the term before admission. This gives the opportunity for staff to learn about how EB 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 wish to do the best for their new pupil with EB and the establishment of a partnership with parent(s) from early on will help all concerned.

This early meeting may be the time to look at possible areas of difficulty and agree strategies for overcoming them. These may include:

1. Avoiding crushes by allowing the child with EB to be first or last in the queue. Where the child has very severe EB it may be appropriate for the child to arrive and leave a few moments before or after the rest of the class.
2. Watching out for objects on the floor to avoid tripping.
3. Considering how best to explain about EB to the rest of the class so that they are aware why they should not push or snatch.
4. Inspecting classroom furniture for sharp edges so that a change can be made for the child or extra care exercised. In the case of a severe form of EB, the child may bring a piece of sheepskin to place on the chair.
5. Avoiding placing the child next to a radiator or next to a sun-baked window since heat can exacerbate blistering.
6. Considering whether the activities undertaken in PE will have to be adapted for that particular child.
7. Thinking about playtime and whether the child will need to have the possibility of an uncrowded area when necessary and, if so, how this can be achieved without isolation.
8. Discussing the child's dietary requirements with school canteen staff if the child has difficulty in swallowing. Parents may choose to provide a packed lunch or dietary supplement but, where school dinners are chosen, food may need to be liquidised or a soft alternative substituted. Ice cream is always popular!

## **Day to day needs**

### **Practical care at school**

Parents may be asked to provide a 'First Aid 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 and details of the GP. However, it has been found that most children with EB cope well with school life, quickly learning how to avoid getting blisters.

### **Lifting the child**

Before handling any child with dystrophic EB, remember that friction will cause blisters and skin damage, but direct pressure will not. Children with EB must never be lifted from underneath the arms as painful blistering will always result and this area is very difficult to dress and to heal.

When needing to be lifted from a sitting position, ask the child to lean forward slightly and place one hand under the bottom, the other behind the neck and lift in one movement.

### **Craft subjects**

If sport is restricted there must be education for leisure and enjoyment and fulfilment in other areas, such as art, craft 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. Attention may need to be given to the best tools or equipment to minimise damage, for example plastic-handled, very sharp scissors or battery-aided scissors can, under supervision, help to avoid pressure on fingers and thumbs. Some aids, such as a foam-handled tap turner or a foam tube covering for pencils, will be of assistance to some children and these can be obtained via the Social Services Department.

Whether clay, paint and Plasticine can be used may depend on the day to day condition of the hands. Parents will know from experience when these sorts of activities should be restricted and can advise on appropriate skin care afterwards.

### **PE and Games**

The objective should be to allow the child to join in as much as possible but to be sensitive to those situations when an adaptation should be made. For floor activities the child can stand, sit or lie on a foam mat. All joints need to be gently stretched and flexed and, if possible, such exercises should be incorporated into PE lessons. Blistering is precipitated by friction so exercises which involve kneeling, handstands or jumping may be problematical. Games involving hand holding and fast activity may also be difficult but a lightly held wrist can be used in any pairing activity. On the other hand, swimming is an ideal sport and arm bands on a foam strip, or polystyrene floats, can be used for the learner. Care should be taken to avoid the danger of slipping on wet tiles is sometimes of help when getting in or out of the pool.

Teachers may wish to read the DEBRA booklet 'A Guideline to Physiotherapy for Children with Epidermolysis Bullosa' to see what exercises may be beneficial to the child with severe EB. The specialist physiotherapist at Great Ormond Street Hospital may also be able to offer specific advice in some cases.

### **School meals**

Eating may be difficult if the mucous membranes of the mouth and gullet are affected. Children with severe EB lose a great deal of protein in blister fluid so they need to pay special attention to diet. Swallowing may vary from day to day and the child may need a near liquid, or very soft, diet until healing occurs. At these times, meals may need to be liquidised. Over recent years dietary intervention has greatly improved with the use of high protein, high fibre drinks or even gastrostomies, where the child is fed overnight through a tube into the stomach. Parents will be able to advise schools if their child has any particular dietary needs.

Teachers may be interested to read the DEBRA booklet 'Nutrition in Epidermolysis Bullosa for children over 1 Year of age' to gain greater insight into this issue.

## **Physical challenges**

Children with severe EB may experience exhaustion due to loss of protein from blisters and loss of iron (anaemia) whilst their bodies are fighting infection and trying to heal. Blistering to the feet and other areas of the body can make walking painful so that some children will need to limit the amount of walking they do around the school. Similarly, writing may be slow due to joint contractures, pain and damage to the hands; the use of computers for schoolwork has been found helpful for some children.

## **Secondary school**

### **A time of change**

Much of what has been discussed above about primary school will be applicable to secondary school as well. Parents will need to satisfy themselves that they pick the right school which can meet their child's physical needs and provide the best education possible. Staff at the new school will need to be satisfied that they can provide for the child and the question of 'statementing' may arise again.

In other ways, things will be different. On a practical level, the child will have to move around the school more, from classroom to classroom for different subjects which may exacerbate existing mobility difficulties. In some schools there will be no coat peg or secure desk where books and equipment can be kept, with the expectation that pupils will carry everything with them in a (heavy) bag. The physical layout of the school will become more important and special arrangements to avoid these difficulties may have to be made.

On the social and psychological level, the child with EB is entering a new life, where he or she is expected to be more independent and responsible. Additional help which may be needed must be given sensitively to avoid the child standing out.

### **Balance**

The group of children entering Year 7 with the child with EB will be his or her friends and peer group for the next 5 – 7 years. The local secondary school may not be ideal in terms of accessibility but it may be the school which most of the other children in the neighbourhood attend. If the child goes to a more accessible, but more distant school will he or she lose touch with friends from primary school and have difficulty in seeing new school friends socially because they live too far away? If the child stays with friends from primary school, will he or she find it too difficult to cope with an unsatisfactory layout in the new school? There is no right answer in all cases. Each child and family will have to come to their own conclusions, based on their priorities. Once the choice has been made the need for partnership between home and school will be as important as ever.

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 which will enable them to have the broadest range of career options possible.

For the more severely affected pupils, there can be significant obstacles to be overcome. Extended absence for hospital treatment may need to be dealt with, time may have to be found each evening both for homework and skin care. With commitment from pupil, parents and school they can be overcome as the success of many children with EB shows.

### **Educational needs**

Examination Boards can be contacted well in advance to make concessions for children with EB who have physical difficulties, such as slow handwriting because of injury to the hands, or any hearing loss. Extra time can be given in written examinations, or in science, art or craft practicals. An amanuensis can be employed by the school or education authority if the student cannot write because of injury in the hands: answers can be dictated.

Education Authorities have the power to provide a further year's education in school for youngsters who have been held back through physical disabilities or hospitalisation. The decision whether to do so will depend on individual needs.

Special provision of aids, such as computers and calculators with instantly responsive keys, may help not only with immediate needs but may give rise to career possibilities.

A wide range of careers has been achieved by people with EB and the condition should not be a barrier to Further Education and training. Although beyond the scope of this booklet, advice and support is available from DEBRA and other agencies to the student with EB and for their universities, colleges and future employers.

### **Further support**

DEBRA is the national charity that supports individuals and families affected by Epidermolysis Bullosa (EB).

DEBRA provides an enhanced EB Healthcare Service, in partnership with the NHS, to deliver optimal healthcare to children and adults living with EB and community support staff to work directly with individuals and families. DEBRA also provides information and help to anybody living or working with EB, including carers and healthcare professionals, and funds pioneering research to find effective treatments and, ultimately, a cure for EB.

To find out more about how DEBRA can support you please visit [www.debra.org.uk](http://www.debra.org.uk), call 01344 771961 or email [debra@debra.org.uk](mailto:debra@debra.org.uk).

## Guidelines for Playgroup

- **NEVER** pick up under arms.
- When lifting, support under nappy/bottom and other hand on back or front.
- No hard biscuits or food – try jaffa cake type biscuits, quavers.
- Food can sometimes get stuck in the throat, no need for alarm, it will be coughed up, occasionally the child will vomit when this happens and need reassurance. There may be streaks of blood in the vomit – this is normal for children with dystrophic EB.
- If drinking with a straw only use the bendy type as straight straws scrape the top of the mouth.
- Baby wipes are best for wiping hands.
- After painting, sand or glue wash in water and dry with a soft towel – **NO PAPER TOWELS**. If the paint or glue does not come off do not scrub, leave it will come off in the next bath.
- Don't use anything sticky – Sellotape, sticky labels, plasters. If something does accidentally become stuck leave, **DO NOT** attempt to take off.
- When sitting on the floor if possible sit on sheepskin or cushion with the group. Watch fingers, feet and legs when others are walking around or getting up.
- When sitting at table avoid kicking feet. Place a beanbag or cushion under table if necessary.
- Sand – only if damp and there are no sore areas on hands, wash hands well after use and watch eyes.
- If the eyes are sore, try to put the child somewhere dark with sunglasses on. The child will need to go home. **NEVER** attempt to open eyes or to take anything out.
- If very windy please allow to stay indoors or wear sunglasses to protect eyes.
- If bright, or the eyes have been sore recently, please allow to wear sunglasses.
- If itchy please try to distract or put cream on
- If the child falls or knocks themselves, check area for blister, if the area is red a blister may appear in next half-hour, this will need bursting. If area has lost the skin then it will need dressing
- Socks or tights must always be inside out
- Pants and vest if they do not have flat seams must be turned inside out
- Nappy changing – follow parent's instructions, nappy liners are commonly used to prevent friction
- If bare skin is exposed when changing nappy or putting on a dressing make sure it is on something soft, a towel or sheepskin – avoid plastic as the skin can stick
- Always support when walking up and down steps, may need extra help if knees are sore
- If wearing apron, watch for any straps rubbing around the neck or throat
- If dressing up watch for zips, buttons, poppers, Velcro, when putting things on and taking things off especially over the head (nose, lips and forehead are very vulnerable)
- Watch for sharp or rough edges on tables, display boards and so on
- Watch for objects on the floor to avoid tripping and slipping
- Be careful that jewellery and watches do not catch the child's skin

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