

THE ROLE OF PHYSIOTHERAPY IN THE SCHOOL – CHILD WITH EPIDERMOLYSIS BULLOSA

The symptoms of EB can vary greatly. The skin is extremely fragile and blisters form when the tissues are subject to friction. Layers of skin move apart and fluid fills this created space. The severity of the condition will depend on the type of EB the child has and thus the depth at which the blisters form.

Each child is unique in their presentation.

Blister formation can result in large raw areas or deep cracks in the skin. The deeper the blisters the more pain and difficulty moving the child will often have.

When these areas form the child is at risk of several problems:

1. Reluctance and fear of moving joints/limbs due to pain.
2. Difficulty walking – certain types of EB cause very painful feet.
3. Difficulty moving due to dressings and bandages which often cause restriction.
4. Increased periods of immobility.
5. Reduced amount of movement a joint has available due to muscle tightness.
6. formation of a joint deformity, when a joint becomes stuck in a particular position.

If a child has a particularly sore area they will be reluctant to move it and over time, if they are not moved through their full normal range the muscles will become tight. This is called a contracture. If they can't move the joints through their full range of movement the muscles do not get the chance to work properly and can become weak.

If the muscles are weak the child is then often not able to walk as far as usual and function can be affected.

If the function is affected the child is less active and therefore the muscles can get tight through disuse and then you have a vicious circle.

This vicious circle can obviously have a big impact on the child and significantly effect their independence. This is often a large problem at school.

Physiotherapy is concerned with maintaining function promoting the most amount of activity possible and trying to prevent these problems from occurring.

The difficulty of course is doing this with such fragile skin. It can be done but care obviously needs to be taken and therapy needs to be tailor made to the individual. It is very important carers are able to recognise signs of deteriorating function and are able to help improve it safely.

This will then help the child to maximise their abilities and independence.

Physiotherapy can play a very important role.

The aims of therapy are to:

- maintain and improve mobility
- maintain and improve joint movement
- maintain and improve muscle strength
- encourage activity and motor development
- give advice on equipment and splinting.

This can be done with:

1. Specific exercise regimes, targeting relevant joints.
2. Active and passive stretches. These will maintain the length of the muscles. The passive type (carried out by the carer) have to be done with great care. The active type are done by the child.
3. Prone lying. This is lying on the stomach to stretch the hip muscles. This is very important to keep the child walking straight.
4. Mouth and tongue exercises. These get tight with blistering, keeping them as mobile as possible will help speech and feeding.
5. Hydrotherapy – swimming is an excellent way of maintaining movement as the child can move freely with the water supporting their limbs.
6. Splintage, we will often use splints at night or sometimes during the day to give a joint or muscle a stretch.
7. Developmental therapy, for the much smaller children and babies we also aim to promote developmental milestones. Children can become delayed due to fear of moving due to pain, restriction of dressings and understandable overprotection by carers. we aim to allow them to experience ALL positions and will often supply therapy programmes to encourage various aspects of development.
8. Use of equipment. Standing frames etc can be used to good effect to stretch and strengthen joints and muscles. Extra padding may be required for vulnerable areas but the frames can be used safely. Trikes etc can also be used to give more severely affected children some independent movement.

The more opportunity the child has to move their limbs the better. At school there is ideal opportunity to do this. Sometimes PE with their classmates may be too dangerous but this time can be spent joining in where possible and doing specific exercises.

Taking the joints through their full range of movement on a daily basis will highlight any problem areas. At any hint of tightness active exercises can be encouraged within the child's pain limits to stretch specific muscles.

The most severely affected children will have their own physiotherapist and they can help with specific exercise regimes. For hydrotherapy, advice from a medical practitioner is always advisable.

Due to the nature of EB there will be days when pain is a limit and activity will be impossible but by monitoring these children closely we can aim to help them have the best quality of movement possible.

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