Birmingham Children's Hospital NHS
NHS Foundation Trust

Information Leaflet for parents/carers

Kindler Syndrome

Kindler Syndrome

Contents:

Kindler Syndrome is part of the group of fragile skin conditions called Epidermolysis Bullosa (EB). This leaflet is designed to give you a brief overview of the condition and what you can do to alleviate the symptoms.

What is Kindler syndrome?

Kindler Syndrome is a rare genetic skin condition in which there is blistering of the skin, changes in the appearance of the skin (poilkiloderma) and sometimes involvement of the lining of the mouth, food pipe and eyes.

What are the symptoms?

Kindler Syndrome is usually identified before a child is 1 year old as the skin has a tendency to blister in infancy, mainly on the hands and feet. By the age of 5 the skin tends to appear thin and wrinkled (atrophic). The internal membranes may also be fragile so there may be problems with the gums, eyes, food pipe and bowels. This can cause problems with discomfort, dental problems and constipation.

How is Kindler Syndrome diagnosed?

Kindler Syndrome is usually diagnosed based on how the skin looks and can be confirmed by taking a small piece of skin (a skin biopsy) and sometimes by doing a blood test.

Why has my doctor never heard of Kindler Syndrome?

Kindler Syndrome is very rare and is often confused with other skin disorders.

What causes Kindler Syndrome?

Kindler Syndrome is caused by a defective gene, KIND1. Everybody carries two copies of every gene, one in inherited from each parent. Kindler Syndrome is an autosomal recessive condition. This means that a person with Kindler Syndrome has inherited two defective copies of the gene, one from each parent. With Kindler Syndrome, the parents usually have one defective copy and one normal copy: they are said to be 'carriers' and do not have Kindler Syndrome.

Parents who have had a child with Kindler Syndrome have a one in four chance of any future baby developing Kindler Syndrome.

How is Kindler Syndrome treated?

There is no cure for Kindler Syndrome but symptoms can be relieved.

Blisters should be burst with a sterile needle and the fluid drained. The area should then be covered with special dressings which do not stick to the skin. These can help wounds heal and prevent the open wounds getting infected. Your child's EB Nurse will advise you on the best dressings for your child's skin.

Moisturisers help to keep the skin in good condition.

Avoiding excessive exposure to sunlight will help to slow down the development of skin damage. Use of a high factor (SPF30+) sunscreen is recommended. This should be applied on all sunny days, even in Winter.

Teeth: It is important that you establish a good dental health routine for your child from an early age. This includes encouraging your child to eat a healthy diet, brushing their teeth daily and regular visits to the dentist (at least every 6 months).

Constipation: When weaning your child, a varied diet rich in fruit and vegetables should be introduced. They should also be encouraged to drink plenty of fluids. This may help prevent constipation. Your health visitor can give advice on how treat constipation. Sometimes medications may be prescribed.

Will my child get better?

Your child will always have the condition but the blisters usually improve after the first year, although the skin remains fragile.
By the age of 5 the appearance of the skin tends to change. It looks thin and papery (atrophic) and there are changes in colour that look like freckles. These changes tend to be noticed first on sun exposed areas like the back of the hands and tops of the feet. These changes are known as poikiloderma.

Some children with Kindler Syndrome are more sensitive to the sunlight which may make them more likely to get skin cancers in adult life than they otherwise would be.

Children with Kindler Syndrome are otherwise well and should be encouraged to lead as normal and full a life as possible.

Looking after and sharing information about your child

We have a duty of care to help patients and families understand how information about them is kept and shared and we include the following information in all our patient leaflets:

Information is collected about your child relevant to their diagnosis, treatment and care. We store it in written records and electronically on computer. As a necessary part of that care and treatment we may have to share some of your information with other people and organisations who are either responsible or directly involved in your child's care. If you have any questions and/or do not want us to share that information with others, please talk to the people looking after your child or contact PALS (Patient Advice and Liaison Service) on 0121 333 8403/8611.

Please use this space to write down any notes or questions you might have.

Support Groups

EB team at Birmingham Children's Hospital
Tel: 0121 333 8224/8757
eb.team@bch.nhs.uk
www.bch.nhs.uk

EB patients' support group DEBRA
Tel: 01344 771961
debra@debra.org.uk
www.debra.org.uk

Health information and internet access is also available in the Family Health Information Centre at the hospital.
Tel: 0121 333 8505
Email: child.infoctr@bch.nhs.uk

Further Information

We hope this leaflet will help you to understand about your child's condition. This leaflet was produced using the most up to date evidence available. Further details are available upon request. If you feel you need any more information please speak to your child's dermatologist or specialist nurse who will try to answer any further questions you may have.

Dermatologist Tel: ....

Specialist Nurses Tel: ....

Community Nurses Tel: ....

Produced by Natalie Kingsbury, EB Nurse Specialist, Dr F.Browne and Prof C. Moss, Dermatology Consultants, Birmingham Children's Hospital in association with Debra. With thanks to the DEBRA charity for helping to fund the production this leaflet.

Birmingham Children's Hospital NHS Foundation Trust
Steelhouse Lane Birmingham B4 6NH
Telephone 0121 333 9999
Fax: 0121 333 9998
Website: www.bch.nhs.uk

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for people whose skin doesn't work we do
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Birmingham Children's Hospital

The Information Standard
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