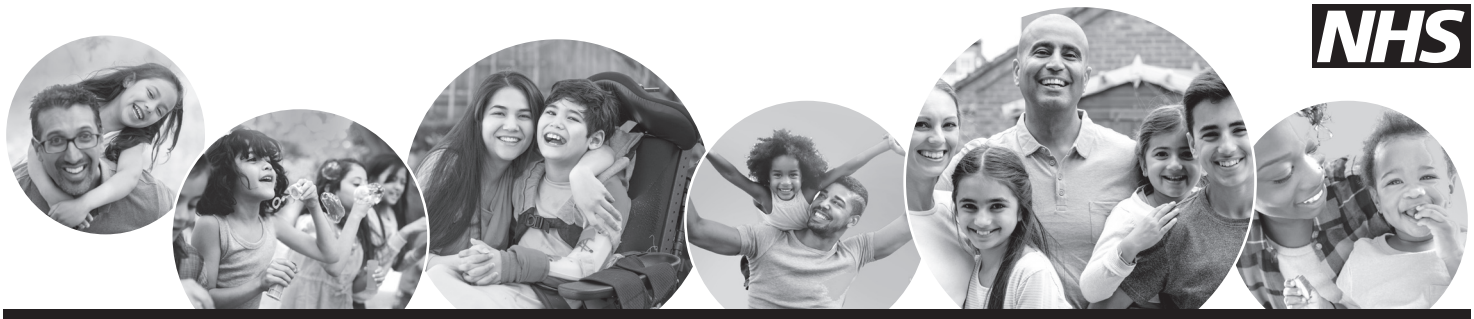

Chronic bullous disease of childhood

This information sheet explains chronic bullous disease of childhood and tells you what to expect when your child comes to Great Ormond Street Hospital (GOSH) for diagnosis and treatment.

What is chronic bullous disease of childhood?

This is a rare autoimmune skin condition which results in clusters of blisters developing in rings often on the face or genitals. In autoimmune disorders, antibodies – which usually seek and destroy foreign invaders to the body, such as viruses – attack the body's own cells instead. This causes inflammation and damage. Other autoimmune disorders include some types of arthritis and inflammatory bowel disease.

Chronic bullous disease of childhood is also known as chronic bullous dermatosis or linear IgA dermatosis of childhood. There is a similar condition called IgA dermatosis which affects adults but its development and treatment are different from that in children.

What are the symptoms?

The main feature is the development of clusters of blisters in specific areas of the body. These blisters are usually itchy and uncomfortable. Some children have many of these clusters, others have only a few. They tend to appear in phases, with new clusters of blisters appearing in the same area as previous ones.

How is it diagnosed?

A skin biopsy is usually needed to confirm the diagnosis. For more information, please see our *Skin biopsy (punch method) information sheet.*

If your child has chronic bullous disease, his or her blood test will show higher levels of antibodies than usual. The skin biopsy will often show IgA antibodies as well which are arranged in a line ("linear" – hence the name that is sometimes used for the condition, linear IgA dermatosis of childhood). Blood tests are often needed to confirm diagnosis.

What causes chronic bullous disease of childhood and whom does it affect?

We are not sure what causes autoimmune disorders such as chronic bullous disease of childhood. More research is needed to discover what triggers antibodies to attack the body instead of fighting foreign invaders like viruses. The condition is not inherited, so cannot be passed on to other children or adults.

Chronic bullous disease of childhood usually appears before the age of five and affects both sexes equally. It also appears to affect all races, but more research is needed to confirm this.

How is it treated?

The aim of treating chronic bullous disease of childhood is to reduce the blistering and associated itching and to prevent any infection. If a blister pops or is scratched so that the surface breaks, this provides a route for infection into the body. If untreated, such an infection may cause serious problems. In the past, before modern medications were developed, this condition and others like it could be fatal if blisters became infected.

A range of treatments are used, including dapsone, ciclosporin and steroids.

Dapsone is a medicine used to treat certain infections but it also has anti-inflammatory effects. Steroids like prednisolone are given at a low dose to prevent blisters forming. Ciclosporin suppresses (damps down) the immune system and also prevents blisters forming.

What is the outlook for children with chronic bullous disease of childhood?

The outlook is very good, with no lasting effects. In most children the condition improves suddenly, usually within two years. The blisters do not usually leave any scarring and should not recur.

Is there a support group for children with chronic bullous disease of childhood?

No, there is no support group specifically for this condition, but the following organisation should be able to offer support and advice:

Contact a Family

Freephone 0808 808 3555
Email: info@cafamily.org.uk
Website: www.cafamily.org.uk

Compiled by the Dermatology Diseases Department in collaboration with the Child and Family Information Group
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