



## **LINEAR IgA DISEASE**

### **What are the aims of this leaflet?**

This leaflet has been written to help you understand more about linear IgA disease. It tells you what it is, what causes it, what can be done about it, and where you can find out more about it.

### **What is Linear IgA disease?**

Linear IgA disease is a very rare blistering condition of the skin in adults. A similar condition affecting children is known as Chronic Bullous Disease of Childhood (CBDC).

The condition is called 'Linear IgA disease' because a type of protein called immunoglobulin A (IgA) is deposited under the outer layer of the skin (the epidermis) in a line (linear). This can be seen when a skin biopsy is prepared specially and examined under the microscope.

### **What causes Linear IgA disease?**

Often no cause can be identified. Sometimes, infections and medicines (most commonly the antibiotic vancomycin) may trigger linear IgA disease. It is not due to food allergy. The disease is thought to be related to the immune system attacking structures in the skin layers (i.e. it is an autoimmune disease). When these structures are damaged, skin blisters may form. There are some reports that linear IgA is more common in people with other autoimmune diseases, such as ulcerative colitis, and certain cancers.

### **Is it hereditary?**

No.

## **What are the symptoms of Linear IgA disease?**

These range from mild itching to a severe burning sensation.

## **What does it look like?**

Patients have an itchy rash with small blisters, often arranged in clusters or rings. The blisters can come up all over the body and limbs, or just affect a few localised areas. The blisters may show the 'string of pearls sign' which describes blisters on the rim of red patches. Blisters may heal to leave temporarily discoloured areas of skin, particularly in patients with darker skin. Most affected adults have blisters or ulcers in the mouth, eyes or the genital region. It does not affect internal organs.

In children, the rash often occurs in the genital region and on the face especially around the mouth.

## **How will it be diagnosed?**

It can be difficult for doctors to diagnose linear IgA. The diagnosis requires a skin biopsy in which a line of IgA under the epidermis can be shown up by a special test (direct immunofluorescence). A skin biopsy involves removal of small pieces of skin after the skin has been numbed with a local anaesthetic injection. Afterwards one or two stitches will usually be put in the skin.

## **Can it be cured?**

Treatment suppresses linear IgA disease but does not cure it. However, in the majority of patients the disease tends to eventually resolve after years. If it has been caused by medication, stopping the drug usually clears it up. Skin lesions tend to heal without leaving permanent scars. However, ulcers in the eyes, mouth or genital region can leave scars or cause permanent damage.

## **How can it be treated?**

Treatment choice depends on the severity of the disease. Mild disease can be treated with topical steroids alone. Most patients with linear IgA disease do well with tablets such as [dapsons](#). Other medications such as sulphonamides, steroids, [ciclosporin](#), [mycophenolate mofetil](#), [intravenous immunoglobulins \(IVIg\)](#) and [colchicine](#) may be used. Oral antibiotics such as tetracyclines, or erythromycin have also been used and can be very helpful especially in children.

## Self care (What can I do)?

- Ask to see an eye specialist if you are getting problems with your eyes; for example, [redness](#) and a sore gritty feeling.

Where can I get more information about linear IgA disease?

Web links to detailed leaflets:

[www.thedoctorsdoctor.com/diseases/linear\\_iga\\_disease](http://www.thedoctorsdoctor.com/diseases/linear_iga_disease)

[www.dermnetnz.org/immune/linear-iga.html](http://www.dermnetnz.org/immune/linear-iga.html)

<https://www.pcds.org.uk/clinical-guidance/linear-iga-disease-syn-chronic-bullous-disease-of-childhood-linear-iga-bul>

For details of source materials used please contact the Clinical Standards Unit ([clinicalstandards@bad.org.uk](mailto:clinicalstandards@bad.org.uk)).

**This leaflet aims to provide accurate information about the subject and is a consensus of the views held by representatives of the British Association of Dermatologists: individual patient circumstances may differ, which might alter both the advice and course of therapy given to you by your doctor.**

*This leaflet has been assessed for readability by the British Association of Dermatologists' Patient Information Lay Review Panel*

**BRITISH ASSOCIATION OF DERMATOLOGISTS**

**PATIENT INFORMATION LEAFLET**

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