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 eruption 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) can be seen deposited in a line in the skin when a small sample of skin (a biopsy) is prepared specially and examined under the microscope. This line of IgA lies just under the outer layer of the skin (the epidermis).

What causes linear IgA disease?

Often no cause can be identified. Sometimes, infections and occasionally medicines may trigger linear IgA disease. The IgA lying in the skin is made by the body’s immune (defence) system and seems to be directed against structures in the skin attaching the outer layer of the skin to the deeper tissues (i.e. it is an autoimmune disease). When these structures are damaged, the skin blisters more easily than it should. Linear IgA disease is not contagious.

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 smaller patches. The blisters may show the ‘string of pearls sign’ which describes the presence of blisters lying around the rim of wheal-like red patches. In children, the rash often occurs in the genital region and on the face especially around the mouth. Blisters and ulcers can affect the mouth and eyes, but the internal organs remain clear.

How will it be diagnosed?

The diagnosis can be difficult clinically, as linear IgA disease can mimic several other disorders. Confirmation of 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). Sometimes, these antibodies can be picked up by a special blood test.

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.

How can it be treated?

Treatment choice depends on the severity of the disease. Most patients with linear IgA disease do well with tablets such as dapsone. Other medications such as sulphonamides, steroids, cyclosporin, mycophenolate mofetil and colchicine may be used. Oral antibiotics such as tetracyclines, or erythromycin have also been used.

Dapsone is used most often. This has some potential side effects, so treatment will not usually start until the diagnosis has been confirmed by a skin biopsy. All patients on dapsone get some decrease in haemoglobin (the oxygen-carrying part of red blood cells), which is usually dose-related and can be monitored with blood tests. However, a few patients on dapsone get a rapid fall in their blood counts. For this reason, weekly blood tests are performed to begin with, and any unusual symptoms (e.g. a sore throat,
dizziness or faintness) should be reported to your doctor. Dapsone also causes headaches in some patients.

Many patients will get better slowly and treatment can then be gradually reduced and then stopped. During treatment most patients will need to continue seeing a dermatologist as well as their own GP.

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.
- Cover your blisters and raw areas with sterile dressings.

Where can I get more information about linear IgA disease?

Web links to detailed leaflets:

www.thedoctorsdoctor.com/diseases/linear_iga_disease
www.dermnetnz.org/immune/linear-iga.html

For details of source materials used please contact the Clinical Standards Unit (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: its contents, however, may occasionally differ from the advice given to you by your doctor.

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

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