



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. One type affects adults, usually over the age of 60; another (also known as chronic bullous disease of childhood) affects children, usually under the age of five.

The condition is called linear IgA disease because a line of immunoglobulin of type A (IgA) can be seen 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?

Infections and antibiotics sometimes 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 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 infectious.

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 rings. The blisters can come up all over the body and limbs, or just affect a few smaller patches. The 'string of pearls' sign is the presence of blistering lying around the rim of wheal-like red patches. In children, the rash usually occurs in the genital region. 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, 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 blood test.

Can it be cured?

Treatment suppresses linear IgA disease but does not cure it. However, the disease does tend to go away by itself, taking 3 to 6 years to do so.

How can it be treated?

Most patients with linear IgA disease do well with tablets such as [dapsons](#), sulphonamides or steroids. Most will need to continue seeing a dermatologist as well as their own GP.

- *In adults*, the tablet used most often is dapsons. 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 dapsons 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 dapsons 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. Dapsons also causes headaches in some patients.

- *In children*, the tablet used most often is one called sulphapyridine. This is generally safe, but can cause some side effects like those of dapsone.
- Many patients will get better slowly over several years and treatment can then be reduced.
- Other tablets such as tetracyclines, colchicine, or erythromycin have also been used. If severe, [steroid](#) or [mycophenolate mofetil](#) tablets may be considered.

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 (includes photographs)

www.emedicine.com/derm/topic240.htm (includes photographs)

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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