



THE AUSTRALASIAN COLLEGE  
OF DERMATOLOGISTS

## Linear IgA Disease

**Also known as** ... Chronic Bullous Dermatosis of childhood, linear immunoglobulin A dermatosis

### What is linear IgA disease?

Linear IgA disease is a rare skin blistering condition. It affects young children (usually before 5 years of age) or adults (usually after 40 years of age). This condition affects the skin and mucosal (inside the mouth and/or genitalia) surfaces of the body.

### What causes linear IgA disease?

The exact cause of linear IgA disease is unknown; however, it is considered to be an "autoimmune" disease. The body's immune system produces antibodies known as immunoglobulin A (IgA) which are incorrectly deposited in the top layer of the skin in a line – hence the term "linear". Deposits of antibodies in the skin cause damage to the structure of the skin and a blister appears.

Certain medications can cause linear IgA disease. The antibiotic, Vancomycin, is the most commonly known drug to cause linear IgA disease. Other drugs reported to cause linear IgA disease include lithium, phenytoin, trimethoprim/ sulfamethoxazole, Frusemide and diclofenac. In these cases, the blisters appear after the medication has been started, and disappear after the medication has been stopped.

Linear IgA disease was once thought to be related to dermatitis herpetiformis which occurs in coeliac disease, but this has been proven to be incorrect. A gluten-free diet will therefore not control or cure Linear IgA Disease.

### **What other problems can occur with linear IgA disease?**

Linear IgA Disease may occur in association with cancer known as "paraneoplastic phenomenon" or "paraneoplastic syndrome". Lymphoma and haematological (blood) malignancies are the most common types of cancers seen in association with linear IgA disease.

Linear IgA disease may also be associated with inflammatory bowel disease and infection.

### **What does linear IgA disease look like?**

In Linear IgA Disease multiple tense blisters with a clear fluid appear over parts of the body. The blisters can vary in size and the skin may appear red around the blisters. Blisters often occur in clusters giving a "cluster of jewels" or "string of beads" appearance. Blisters and ulcers may occur on mucosal surfaces such as in the mouth or genitalia. Typically, the skin may be itchy or sore or have a burning sensation. Gritty or sore eyes may suggest involvement of the eyes.

In rare cases the larynx and/or pharynx (air passages) may be affected which may cause changes in the sound of the voice.

Sometimes there may be fever, joint pains and feelings of tiredness.

### **How is linear IgA disease diagnosed?**

The diagnosis of Linear IgA Disease is usually confirmed with a skin biopsy. Special staining of a skin sample in the laboratory typically shows deposition of the antibody, Immunoglobulin A (IgA) in the skin. A blood test may show circulating IgA antibodies in the blood.

### **How is linear IgA disease treated?**

In adults, the condition has an unpredictable course but tends to be chronic (continue for years). Long-term treatment is usually needed to suppress the condition and keep the skin clear of any blisters.

Linear IgA disease often responds extremely well to **dapsone**. Other treatments include prednisolone, sulfapyridine, colchicine, **tetracyclines**, erythromycin, **azathioprine**, **cyclosporine** or mycophenolate mofetil. **Topical steroid creams** may also be used. Intravenous immunoglobulin therapy may be used in people who fail to respond to the above-mentioned treatments.

In children, the disease tends to disappear over time with most children going into remission within two years of onset of the disease.

When treated, skin blisters tend to fade away without scarring or leaving a permanent mark on the skin.

Serious complications of linear IgA disease may include infection, dehydration and temperature dysregulation. Mucosal lesions inside the mouth or eyes may heal with some scarring.

***If you notice multiple blisters on your skin, you should see a dermatologist immediately.***

This information has been written by Dr Eleni Yiasemides