



LINEAR IgA BULLOUS DERMATOSIS

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Linear IgA bullous dermatosis (LABD) is a rare subepidermal blistering disease due to an autoimmune reaction against basement membrane proteins such as the lamina lucida and sublamina densa. The basement membrane anchors the epidermis to the dermis and helps to stabilize the skin. When IgA antibodies target such proteins, the basement membrane destabilizes resulting in tense blister formation.

Despite its low prevalence, LABD has a bimodal peak of age distribution. The disease can occur in children ranging from infancy to early adolescence while adult-onset LABD tends to occur at a mean age of 50. In children, the disease is sometimes called 'chronic bullous disease of childhood'. Both children and adults may experience prodromal itching and form tense blisters on the base of red or normal-appearing skin. The blisters tend to form in clusters resulting in the classic "cluster of jewels" sign or linearly along the edge of a blister resulting in the "string of beads" sign. Patients may also have scattered red bumps or plaques at sites of inflammation instead of blisters. Lesions in children tend to involve the perioral, perineum, and anogenital areas more frequently than adults. However, both children and adults can experience mucous membrane and ocular involvement including burning and discharge from the eyes.

In the majority of LABD cases, the cause is unknown or idiopathic. Furthermore, more than half of all childhood cases tend to remit over a mean course of two to four years. Adults may have a more protracted course and LABD has been shown to occur in those with internal malignancy, infection, and other autoimmune diseases like rheumatoid arthritis or **dermatomyositis**. Given the rarity of LABD, however, such associations still remain to be proven. Other cases of LABD are drug-induced often due to vancomycin. Patients can break out as early after the first dose of vancomycin in some cases.

To diagnose LABD, the dermatologist will take a punch **biopsy** around the blister site in addition to healthy skin for histological and immunofluorescence studies. Such testing will enable the dermatologist to detect inflammatory cells and autoimmune activity at the basement membrane. Patients with more diffuse involvement or formation of pus-like vesicles may also receive a bacterial and/or viral culture of blister fluid to rule out secondary infections.

Idiopathic cases of LABD have been successfully treated with **dapsone** or sulfapyridine. LABD has also been shown to respond to **mycophenolate mofetil**, **colchicine**, intravenous immunoglobulin, and dicloxacillin. For drug-induced cases, the treatment involves removing the offending drug from one's drug regimen. A short course of **corticosteroids** may also be given to decrease inflammation. Cutaneous lesions can be expected to heal several days to weeks following treatment. However, there is a risk of residual scarring with mucous membrane and ocular involvement.

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