



# Epidermolysis Bullosa Acquisita

## **What is Epidermolysis Bullosa Acquisita (EBA)?**

EBA is a rare blistering disease which produces deep, tense blisters on the skin and mucosal surfaces (mouth, genitals, nose and eyes) which typically heal with scarring. EBA can occur at any age but more frequently affects elderly persons.

## **What causes Epidermolysis Bullosa Acquisita?**

EBA is an autoimmune disease. It arises when the white blood cells of the immune system produce antibodies which attack the collagen proteins (known as anchoring fibrils) which “sew” the outer skin (epidermis) onto the inner skin (dermis). The destruction of these fibrils leads to the development of a blister due to the separation of the two layers of the skin. The exact trigger for this immune attack is unknown.

## **What does Epidermolysis Bullosa Acquisita look like?**

EBA can present with a number of different clinical signs. Mostly it evolves slowly with noticeable skin fragility and small or large blisters that affect the trauma-prone extensor skin surfaces (hands, feet, elbows, knees and buttocks).

In a subset of affected people, it may be more generalised and progress more rapidly with widespread itchy red tense blisters. In this group of affected people, blisters occur in both trauma-prone and non-trauma-prone areas. This can be mistaken for other more common blistering diseases such as bullous pemphigoid or linear IgA disease.

EBA can also affect the nails, mouth, eyes, genitals, larynx and oesophagus.

Sometimes these are the main areas involved. The blistering characteristically heals with scars which can lead to difficulty with vision, eating, swallowing and speaking.

## **What other problems can occur with Epidermolysis Bullosa Acquisita?**

Although the exact relationship is uncertain EBA has been associated with Crohn’s disease and lupus erythematosus. These are also autoimmune diseases.

## **How is Epidermolysis Bullosa Acquisita?**

- Skin biopsies – biopsy of the edge of a blister for routine pathology; biopsy of skin close to a blister can be sent for direct immunofluorescence which looks for deposition of the antibodies in the skin
- Blood tests – these test for antibodies in the blood

## **How is Epidermolysis Bullosa Acquisita treated?**

EBA often requires coordinated care from a number of specialists depending on the areas of involvement.

These may include dermatologists, ophthalmologists, ear nose and throat specialists, and dentists. As this is an autoimmune disease, medications that target and suppress this abnormal immune response are necessary. The aim of treatment is to control and stop the development of new blisters, promote healing and prevent scarring. EBA response to treatment is not always predictable and treatment tends to be necessary for months to years depending on disease activity. Treatments are tailored to each individual.

*Local treatments* for the skin include:

- Protecting the skin from damage
- Relieving pressure on new blisters – sterile needles may be used to release fluid
- Non-stick dressings over existing wounds with secondary absorbent dressings and bandages
- Bathing to keep skin clean and soak off dressings – every 1 to 2 days
- Cortisone creams for the localised blisters

*Oral medications* are often required. These can include:

- Oral prednisolone
- Dapsone for milder cases
- Immune suppressants such as azathioprine, mycophenolate, cyclosporine, cyclophosphamide
- Immune modulators such as intravenous immunoglobulin
- Biologic response modifiers such as rituximab

## **What is the likely outcome of Epidermolysis Bullosa Acquisita?**

EBA tends to run a chronic course and long-term follow-up is often necessary. The disease activity may fluctuate at times. The aim of treatment is to manage the disease to prevent long-term problems from scarring.

## **Further information about Epidermolysis Bullosa Acquisita**

[www.pemphigus.org](http://www.pemphigus.org)

[www.blisters.org.au](http://www.blisters.org.au)

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