THE AUSTRALASIAN COLLEGE
OF DERMATOLOGISTS

Childhood Bullous Pemphigoid

What is childhood bullous pemphigoid?

Childhood bullous pemphigoid is an extremely rare form of the bullous pemphigoid which is an autoimmune blistering disease. It occurs in children under 18 years of age. Given its rarity, the exact incidence of the condition is unknown.

What causes childhood bullous pemphigoid?

Like adult bullous pemphigoid, childhood bullous pemphigoid occurs when cells of the body's immune system produce proteins (autoantibodies) which attack and damage specific proteins of the basement membrane zone (known as BP180 and BP230). These proteins are critical in attaching the top layer of skin cells (the epidermis) to the underlying layer (the dermis). If the basement membrane proteins are damaged the top and bottom layers of the skin break apart causing blister formation.

The cause of the immune attack on the child's skin is unknown.

What does childhood bullous pemphigoid look like?

Similar to adult bullous pemphigoid, childhood bullous pemphigoid is characterised by tense fluid-filled blisters accompanied by redness and itching. There may also be patches of itchy redness without blisters before the onset of blisters

The eruption may occur anywhere on the child's body. Younger children (under one year of age) may have a more extensive rash and their extremities and facial areas are more likely to be affected. There have been some reports of mucous membrane involvement.

What other problems can occur with childhood bullous pemphigoid?

The initial presentation of childhood bullous pemphigoid can be severe. However, unlike adult bullous pemphigoid, the condition usually improves over time.

How is childhood bullous pemphigoid diagnosed?

Diagnosis may be delayed as it often takes some time for the blisters to emerge.

The following tests are usually required:

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

How is childhood bullous pemphigoid treated?

There is currently no cure but it is expected that symptoms will gradually improve over time. Treatments are personalised to the individual and directed towards reducing blister formation, relieving the pruritus (itchiness), promoting wound healing and minimising the side effects of treatment.

Individuals with mild disease

- Potent topical corticosteroid

Individuals with more aggressive disease

Oral medications are often required. These can include:

- ***First-line treatment***
    - Oral prednisolone
- ***Second-line treatments***
    - Topical immunomodulators (e.g. topical tacrolimus ointment)
    - Dapsone
    - Azathioprine
    - Mycophenolate
    - Methotrexate
    - Intravenous immunoglobulin
    - Rituximab

What is the likely outcome of childhood bullous pemphigoid?

Childhood bullous pemphigoid seems to improve with time. In previously reported cases, almost all children reached complete remission within 1 to 2 years. The relapse rate is generally low. However, it can be triggered by infections or if oral corticosteroid treatment is weaned too quickly.

This information has been written by Dr Cathy Zhao & Professor Dedee Murrell