

## PEMPHIGOID

### What are the aims of this leaflet?

This leaflet has been written to help you understand more about pemphigoid. 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 pemphigoid?

Pemphigoid is a rare blistering disorder, which usually occurs in later life, the average age of onset being over 70 years.

The blisters come up on the skin and, less often, in the mouth too. This is in contrast to a related condition known as 'mucous membrane pemphigoid' in which the brunt of the trouble is borne by the moist surfaces of the body (the mucous membranes) such as the eyes, inside the nose and mouth, and the genitals. Yet another type of pemphigoid (pemphigoid gestationis) occurs during pregnancy. This leaflet will not discuss mucous membrane pemphigoid or pemphigoid gestationis further.

### What causes pemphigoid?

Nobody fully understands its cause, but pemphigoid is classed as an autoimmune disease (a group of diseases that are caused by the reaction of the immune system of the person to produce antibodies against substances which occur naturally in the body). For an unknown reason, antibodies (natural substances important in your body's defences) form in the blood and then attack the structures holding the outer layer of the skin (the epidermis) onto the deeper layers. This leads to splitting of the skin and so to blisters.

Sometimes an outside event, such as sunburn or a reaction to a drug, seems to trigger the rash but usually it comes out of the blue. It is not infectious (or contagious), due to allergies, or affected by diet or lifestyle.

### Is pemphigoid hereditary?

No.

### What are the symptoms of pemphigoid?

Itching is common. The raw areas left when the blisters break can be sore, both on the skin and in the mouth.

### What does pemphigoid look like?

- A rash may be present for some weeks before any blisters come up. At that stage, the rash may look like an odd eczema or, more commonly, like the red weals of nettle rash (urticaria).
- When blisters do come up, they often do appear on red patches. Any part of the skin can be involved, but the most common sites for the blisters are the body

folds and the skin on the abdomen. In severe cases, the blisters can occur all over the body.

- The blisters have thick roofs, and can get quite large and tense before they burst. Most contain clear fluid but in a few this is bloodstained. When the blisters heal up, they do so without leaving scars.
- Roughly a quarter of patients with pemphigoid have blisters or raw areas in the mouth.

### **How will pemphigoid be diagnosed?**

Usually the look of the rash is enough to make the diagnosis, but it is essential to get further proof.

- Examination of a biopsy taken from a small and early blister will show that it has come up just under the outermost layer of the skin (i.e. it is a 'subepidermal' blister).
- Special testing (immunofluorescence) of a sample of normal skin (i.e. taken from an area where there is no blister) will show up a layer of antibodies that also lies just under the outer layer of the skin.
- The same pemphigoid antibodies can be detected circulating in the blood.

### **Can pemphigoid be cured?**

No. Treatment helps a lot, but controls the condition rather than curing it completely. However, pemphigoid does often go away by itself after one to five years.

### **How can pemphigoid be treated?**

Treatment has three aims – to stop new blisters coming up; to heal the blisters that are already there; and to use the smallest possible doses of the medicines, as side effects can be common and severe in the elderly.

- A steroid cream can help if only a small area of skin is affected. Even if the rash is more extensive, a trial of a strong steroid cream may be worthwhile before tablets are given, as creams are much less likely to cause side effects.
- Antibiotics called tetracyclines, sometimes combined with nicotinamide, may help and can be combined with other treatments.
- Treatment for more severe blistering is usually with high doses of steroid tablets to get the pemphigoid under control quickly. This needs careful monitoring, especially in elderly patients or those taking other medicines. Treatment may sometimes be started in hospital to ensure good control and so that proper dressings can be used on the blisters and raw areas. Some patients can come off their treatment gradually, usually after a few months; others will need to go on with lower doses for a long time. Calcium and vitamin D supplements, or other medications, should be given to keep your bones strong.
- Other tablets that affect the immune system can be used at the same time as the steroid tablets, the aim being to reduce the steroid dose (i.e. to have a 'steroid sparing' effect). The idea is that the side effects will be less troublesome if a low dose of two tablets is used rather than a high dose of a single type of tablet. Examples of additional tablets that can be used in this way are azathioprine and, dapsons. However, they can all cause side effects too, and their use has to be considered carefully. Your dermatologist will discuss this aspect of your treatment with you.

### **What can I do?**

- Never change the dosage of the tablets you are taking without talking first to your doctor.
- Avoid hard or rough foods if you have sore areas in your mouth.

**Where can I get more information about pemphigoid?**

*References:*

*British Association of Dermatologists' guidelines on the management of bullous pemphigoid 2012*

*Web links to detailed leaflets:*

[www.emedicine.com/derm/topic64.htm](http://www.emedicine.com/derm/topic64.htm)

[www.medicinenet.com/Bullous Pemphigoid/index.htm](http://www.medicinenet.com/Bullous_Pemphigoid/index.htm)

[www.dermnetnz.org/immune/pemphigoid.html](http://www.dermnetnz.org/immune/pemphigoid.html)

*Links to patient support groups:*

*The International Pemphigus and Pemphigoid Foundation*

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

*Other relevant leaflets issued by the British Association of Dermatologists:*

[Azathioprine](#)

[Dapsone](#)

[Oral pemphigoid](#)

[Oral corticosteroids](#)