



PEMPHIGUS VULGARIS

What are the aims of this leaflet?

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

Pemphigus vulgaris is a rare autoimmune (see below) disease that causes severe blistering of the skin and of the mucous membranes lining the mouth, nose, throat and genitals. The blisters have thin roofs and break easily to leave raw areas (erosions) that can be extensive and painful. Pemphigus does not go away by itself, and always needs treatment by a specialist.

What causes pemphigus vulgaris?

We all have an immune system. It makes the antibodies that are needed to fight off infections. Normally these antibodies do not attack our own bodies. However, in an autoimmune disease, the immune system makes a mistake and fights our own body instead. The mistake made by the immune system in pemphigus vulgaris is to view the cells in our skin and mucous membranes as 'foreign' and to make antibodies that damage them.

- Pemphigus vulgaris is therefore an 'autoimmune' disease. The antibodies that attack parts of our own body are called 'autoantibodies'.
- The autoantibodies in pemphigus vulgaris target proteins called desmogleins, which are present on the cells in the outer layer of the skin (the epidermis). Desmogleins act as a glue to stick these cells together.
- When the autoantibodies formed in pemphigus vulgaris have combined with the desmogleins, the cells in the skin and mucous membranes no longer stick together properly and fall apart. This causes the blisters and erosions that are typical of pemphigus vulgaris.
- Although it is known that antibodies to desmoglein cause pemphigus vulgaris, it is still not clear why some people develop these autoantibodies in the first place.

Pemphigus vulgaris affects both sexes equally. It can start at any age but is most common in middle-aged adults. It affects people of all races but is more common in some than others – for example in people of Jewish origin. Pemphigus vulgaris is not an infection: it cannot be caught or passed on to anyone else.

Is pemphigus vulgaris hereditary?

Strictly speaking, it is not, as it does not pass from generation to generation. Genetic factors are important, but other factors are needed to trigger pemphigus vulgaris into activity.

What are the symptoms of pemphigus vulgaris?

The raw unhealing erosions are painful and can disturb sleep. Those in the mouth can interfere with eating and drinking, leading to loss of weight.

What does pemphigus vulgaris look like?

In most patients, the blisters and erosions start first in the mouth, and appear later on the skin. In a few, the skin is affected first. Most patients get erosions in their mouth at some time; but some never, or only very rarely, get blisters or erosions on their skin.

The skin:

- The skin lesions start as thin-walled blisters (collections of clear fluid within the skin), arising on a background of normal-looking skin. Because they are so fragile, pemphigus blisters break very easily, leaving raw areas known as erosions.
- Erosions are areas of skin (or mucous membrane), which lack its top (outer) layer. They look raw and feel sore - like a burn. Erosions can join together to create larger areas of raw skin that look as if the top layer has been scraped off.
- Erosions can become crusty and scabbed. When they heal, those on the skin may leave discoloured marks.

The mouth:

Intact blisters are seldom seen in the mouth in pemphigus vulgaris because they get broken so easily. Erosions are seen there instead. There may be just one or two, or several that can join together.

Will pemphigus vulgaris go away?

No, it will not clear without treatment. There will be times when it flares up, and others when it gets better. There is no way of knowing when these flare-ups will happen or how bad they will be. However, pemphigus vulgaris can be

controlled by long-term treatment and may eventually get better completely, so that it does not flare up when treatment stops.

How will pemphigus vulgaris be diagnosed?

- Pemphigus vulgaris is rare and most general practitioners have never seen it. Your own doctor will send you to see a dermatologist or an oral medicine specialist, who will make a provisional diagnosis of pemphigus vulgaris on the basis of the changes seen on your skin and in your mouth.
- Next a *biopsy* will be taken. This is a sample of an unbroken blister. It will be processed in the laboratory and then examined under the microscope. The diagnosis of pemphigus vulgaris will be confirmed by finding cells in the epidermis that are rounding off and separating away from each other.
- As further confirmation of the diagnosis, part of the biopsy specimen is likely to be examined by a technique known *direct immunofluorescence* to demonstrate the presence of pemphigus vulgaris autoantibodies in the skin.
- Another useful test is to measure the levels of pemphigus vulgaris autoantibody in the blood. This is done by *indirect immunofluorescence* and is sometimes called an antibody titre test. It is a useful way of monitoring how active the pemphigus vulgaris is, and the results can suggest changes in treatment.

How can pemphigus vulgaris be treated?

General aims

Treatment is important because pemphigus vulgaris can be serious. It means using medicines that are usually taken by mouth but are sometimes given into a vein through a drip. As pemphigus vulgaris is an autoimmune disease, most treatments work by suppressing the immune system.

The initial aim of treatment is to prevent new blisters forming and to heal the existing ones - this is 'disease control'. It usually takes about 2 to 3 weeks to stop new blisters coming up, and sometimes as long as 6 to 8 weeks for healing to be complete. Once control has been achieved, the dose of the medication will be reduced slowly to the lowest one needed to keep the pemphigus vulgaris under control. Treatment usually starts with a corticosteroid, which may later be combined with a so-called 'steroid-sparing' drug.

Corticosteroids

Pemphigus vulgaris is usually treated first with a corticosteroid. This is effective and works more quickly than most of the other treatments. The corticosteroids used are synthetic versions of a natural hormone produced in smaller quantities by the body. They work by suppressing the immune system.

High doses of the corticosteroid, usually prednisolone, are given to bring the pemphigus vulgaris under control. The dose is then reduced slowly to minimise side effects. However, the prednisolone can seldom be stopped completely and most patients need a small maintenance dose to keep their disease under control. This dose varies from person to person, and depends partly on their weight.

Steroid-sparing and adjuvant drugs

Unfortunately, although they are very effective, corticosteroids do have side effects. For this reason, other medications are often used as well. These are known as steroid-sparing drugs or adjuvant drugs and they work well when combined with corticosteroids.

They include the following:

1. *Immunosuppressives:* azathioprine, cyclophosphamide, mycophenolate mofetil and ciclosporin.
2. *Additional drugs:* gold, methotrexate, tetracyclines (minocycline, or doxycycline) combined with nicotinamide, and dapsone.

Side effects

All these medications can have serious side effects, so patients must be monitored carefully by regular urine and blood tests, and blood pressure readings.

What can I do?

- Be sure to consult your doctor if you are thinking about ceasing to take your corticosteroids. It is dangerous to do so suddenly.
- If you have erosions in your mouth it may be sensible to avoid eating heavily spiced or hard foods.

Where can I get more information?

Patient support group:

The Pemphigus Vulgaris Network is the UK support group for people living with pemphigus and mucous membrane pemphigoid. It can be contacted by:

- Looking at its website – www.pemphigus.org.uk
- Writing to Flat C, 26 St Germans Road, London SE23 1RJ, enclosing a stamped addressed envelope.
- Telephoning 0208 690 6462 (preferably between 8.30am – 10.00am). The group has no funding, and cannot return phone calls.

Reference used in preparing this leaflet:

“Guidelines for the management of pemphigus vulgaris” by KE Harman, S Albert and MM Black. In the British Journal of Dermatology (2003), volume 149: pages 926 to 937.

Links to other Internet sites:

www.pemphigus.org/ (International Pemphigus Foundation)

www.dermnetnz.org/dna.pemphigus/pgus.html

Other relevant patient information leaflets issued by the British Association of Dermatologists:

These include leaflets on azathioprine, ciclosporin, dapsone, methotrexate, mycophenolate mofetil and oral treatment with corticosteroids.

(Whilst every effort has been made to ensure that the information given in this leaflet is accurate, not every treatment will be suitable or effective for every person. Your own doctor will be able to advise in greater detail.)

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