

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 disease (up to 3.2 cases per 100,000 population) that causes severe blistering of the skin and of the mucous membranes lining the mouth, nose, throat and genitals. Blisters are sacs with fluid that develop on the upper layer of the skin so their roofs are very thin and fragile, 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 Dermatologist.

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, called 'auto antibodies' that damage them.

- The auto antibodies 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 auto antibodies formed in pemphigus vulgaris combine 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 auto antibodies in the first place.
- Pemphigus vulgaris affects both males and females equally. It can start at any age but is most common in adults usually between 50-60 years old. 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?

Information provided by British Association of Dermatologists

The raw unhealing erosions are painful and can disturb sleep. Those in the mouth can interfere with eating and drinking and can lead to weight loss, hence the advice of the dentist would help especially those with dentures. It can also affect other mucous membranes such as the genitalia (causing painful intercourse) and the conjunctiva of the eyes (in which case the help of an ophthalmologist would be useful).

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 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 look raw and feel sore - like a burn. Erosions can join together to create larger areas of raw skin. Erosions can become crusty and scabbed. When they heal, those on the skin may leave discoloured marks.
- *The mouth:*
- Erosions usually form in the mouth because blisters here are easily broken. 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 which may eventually result in no further flare ups.

How will pemphigus vulgaris be diagnosed?

- Your general practitioner will send you to see a dermatologist or an oral medicine specialist, who will make a provisional diagnosis of the condition 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. As further confirmation of the diagnosis, part of the biopsy specimen is likely to be examined by a technique known as *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 taking a blood sample and subjecting it to *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 is serious and can be life threatening if not treated early and properly. It means using medicines that are usually taken by mouth prescribed in outpatients but are sometimes given into a vein through a drip requiring admission as an inpatient. As pemphigus vulgaris is an autoimmune

disease, most treatments work by suppressing the immune system, which, although they carry the risk of developing side effects (see below) they are life saving.

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.

Be sure to consult your doctor if you are thinking about ceasing to take your corticosteroids. It is dangerous to do so suddenly, as your body may have stopped making its own corticosteroids and come to depend on the tablets you are taking for its daily requirement.

Steroid-sparing or 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.

Immunosuppressives: azathioprine, cyclophosphamide, mycophenolate mofetil, ciclosporin and rituximab.

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.

Skin and Mouth treatments (topical treatments) - A steroid cream may be used on skin blisters so that the dose of steroid tablets can be kept lower. Mouth blisters and erosions may be treated with steroid sprays or mouthwashes containing antiseptic or local anaesthetic.

Other treatments - Plasmapheresis, intravenous immunoglobulin and rituximab may be considered if high doses of steroids are not effective. These may also be tried in combination with steroid tablets. Further research continues to find better treatments or combinations to treat PV.

What can I do?

- You should keep your appointments with the hospital
- You should take medicines as advised by the dermatologists and never stop corticosteroids suddenly on your own without informing your dermatologist
- If you have erosions in your mouth it may be sensible to avoid eating heavily spiced or hard foods.

- You should tell your doctor about your condition if your nose, throat or genitalia are examined

Where can I get more information?

Link to patient support group:

The Pemphigus Vulgaris Network

Web: www.pemphigus.org.uk

Web links to detailed leaflets:

www.dermnetz.org/immune/pemphigus-vulgaris.html