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 skin disease. It affects around 3 cases per 100,000 population. Pemphigus vulgaris may cause severe blistering of the skin and the mucous membranes lining the mouth, nose, throat and genital area. Blisters develop in the upper layer of the skin, and have a thin and fragile outer surface that breaks away easily to leave raw areas (erosions) that can be extensive and painful. Pemphigus vulgaris does not go away by itself, and always needs assessment and treatment supervised by a Dermatologist.

What causes pemphigus vulgaris?

Our immune system makes antibodies that are required to fight infection. Normally these antibodies do not attack our own body systems. However, in an autoimmune disease, the immune system attacks our own body. In pemphigus vulgaris the immune system makes antibodies (auto antibodies) that attack the skin and mucous membrane leading to damage that causes blisters to develop.

- The auto antibodies in pemphigus vulgaris attack proteins called desmogleins. The desmoglein proteins are present on the cells in the outer layer of the skin (the epidermis). Desmogleins normally act as a glue to hold the cells of the epidermis together.
- When the auto antibodies formed in pemphigus vulgaris attack the desmoglein protein, the cells in the skin and mucous membranes no longer hold together and separate. 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.

Pemphigus vulgaris affects males and females equally. It can start at any age but is most common in adults between 50-60 years old. It affects people of all races but is more common in some people, for example, people of Jewish origin. Pemphigus vulgaris is not an infection and so it cannot be caught or passed on to anyone else.

Is pemphigus vulgaris hereditary?

Pemphigus vulgaris does not pass from generation to generation. However, genetic factors are thought to be important in combination with other factors.

What are the symptoms of pemphigus vulgaris?

Raw erosions are painful and can affect quality of life including disturbing sleep. Mouth lesions can interfere with eating and drinking and so cause weight loss. Pemphigus vulgaris can also affect other mucous membranes such as the genital area (leading to painful sexual intercourse) and the conjunctiva of the eyes (requiring assessment by an ophthalmologist).

What does pemphigus vulgaris look like?

In most people, blisters and erosions start in the mouth, and appear later on the skin. In some patients the skin is affected first. Most people develop erosions in the mouth but some never get blisters or erosions on the skin.

The skin:

- The skin lesions start as thin-walled superficial blisters (collections of clear fluid within the skin), arising on a background of normal-looking skin. Because they are so fragile, pemphigus blisters break easily, leaving raw areas known as erosions.
- Erosions appear raw and feel sore - like a burn. Erosions can join together to create larger areas of raw skin. Erosions can become crusty, and infected. Skin lesions may heal and lead to discoloured marks.

The mouth:

- Erosions form easily in the mouth because blisters in the mouth are easily broken. There may be one or two, or several that join together.

Will pemphigus vulgaris go away?

Pemphigus vulgaris will usually not clear without treatment. There will be occasions when it flares up and other occasions when it improves. There is no way of predicting when these flares will occur or how severe they will be. However, pemphigus vulgaris can be controlled by long-term treatment which may result in control of these flares.

How will pemphigus vulgaris be diagnosed?

- Your general practitioner may refer you to a dermatologist or an oral medicine specialist. These specialists will be able to make a provisional diagnosis of the pemphigus vulgaris on the basis of the changes seen on your skin and in your mouth.
- A *biopsy* will be taken of a blister to confirm the diagnosis. The biopsy sample will be processed in the laboratory and examined under the microscope. Part of the biopsy sample will be examined by a technique known as *direct immunofluorescence* to demonstrate the presence of pemphigus vulgaris autoantibodies in the skin.
- Pemphigus vulgaris autoantibodies can be measured in the blood *(indirect immunofluorescence)*. Measurement of autoantibodies in the blood is a useful way of monitoring how active the pemphigus vulgaris is and can help your dermatologist advise on changes to your treatment required to control the condition.

How can pemphigus vulgaris be treated?

General aims. Treatment is important because pemphigus vulgaris is a serious and potentially life threatening condition if not treated early and effectively. Treatment entails using medicines that are usually taken by mouth but are sometimes given in severe cases into a vein through a drip requiring admission as an inpatient. Because pemphigus vulgaris is an autoimmune disease, most treatments used work by suppressing the immune system.

The aims of treatment are to prevent new blisters forming and to heal existing ones. It usually takes about 2 to 3 weeks of treatment to stop new blisters developing, and sometimes as long as 6 to 8 weeks for healing to occur. Once control has been achieved, the dose of the medication can be reduced slowly to the lowest level needed to keep the pemphigus vulgaris under control. Treatment usually starts with an oral corticosteroid and may later be combined with a so-called ‘steroid-sparing’ drug.

Corticosteroids. Pemphigus vulgaris is usually treated first with an oral corticosteroid. Oral corticosteroids are effective and work quicker than most other treatments. The corticosteroids used are synthetic versions of a natural hormone produced in smaller quantities by the adrenal gland and work by suppressing the immune system.

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

It is very important to consult your doctor if you are considering stopping your corticosteroid treatment. It is dangerous to stop oral corticosteroid treatment suddenly because your body may have stopped making its own corticosteroids and become dependent on the corticosteroid tablets you are taking for its daily requirement.

Nearly all patients gain weight on steroid treatment, however disciplined they are about food. It's important not to get too worried at the start of treatment when the crucial thing is to get the Pemphigus under control.

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

They include the following:

1. *Immunosuppressive drugs*: azathioprine, cyclophosphamide, mycophenolate mofetil, ciclosporin and rituximab.
2. *Additional drugs that may be used include:* gold, methotrexate, tetracyclines (minocycline or doxycycline) combined with nicotinamide and dapsone.

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

Skin and Mouth treatments (topical treatments). Emollients and steroid cream may be used on skin blisters and may help so that the dose of steroid tablets can be reduced.

Mouth blisters and erosions may be treated with steroid sprays or mouthwashes and mouthwashes containing an antiseptic or local anaesthetic.

Other treatments. Plasmapheresis, intravenous immunoglobulin therapy and rituximab may be considered if high doses of oral steroids and other treatments are not effective. These may also be tried in combination with steroid tablets. Further research continues to find better treatments or combinations of treatment for pemphigus vulgaris.

Self care (What can I do?)

- You should keep all of your appointments with your hospital specialist.
- You should take medicines as advised by your specialist and never stop corticosteroids suddenly without informing your specialist.
- If you have erosions in your mouth it may be sensible to avoid eating spicy or hard foods.
- You should have regular check-ups with your dentist.
- You should tell your doctor about your condition if your nose, throat or genital skin are involved.

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.dermnetnz.org/immune/pemphigus-vulgaris.html

For details of source materials used please contact the Clinical Standards Unit (clinicalstandards@bad.org.uk).

This leaflet aims to provide accurate information about the subject and is a consensus of the views held by representatives of the British Association of Dermatologists: individual patient circumstances may differ, which might alter both the advice and course of therapy given to you by your doctor.

This leaflet has been assessed for readability by the British Association of Dermatologists' Patient Information Lay Review Panel

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