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 and affects around 3 people per 100,000 population. Pemphigus vulgaris may cause severe blistering of the skin and the mucous membranes lining the mouth, nose, throat, eyes and genital area. Blisters develop in the upper layer of the skin and have a thin and fragile outer surface that breaks away easily leaving raw areas (erosions) that can be extensive and painful.

What causes pemphigus vulgaris?

Our immune system makes antibodies to fight infection. Normally these antibodies do not attack our own body. However, in an autoimmune disease, such as pemphigus vulgaris, the immune system makes antibodies (autoantibodies) that work against tissues in the body.

- The autoantibodies in pemphigus vulgaris attack proteins called desmogleins. These proteins are present on the cells in the outer layer of skin (the epidermis) and form the “glue” that holds the cells of the skin together.
- When the autoantibodies formed in pemphigus vulgaris attack the desmoglein proteins, the cells in the skin and mucous membranes no longer hold together and separate. This causes 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.

Pemphigus vulgaris affects males and females equally. It can start at any age but is most common in adults between 40-60 years old. It affects people of all races but is more common in some people, for example, people of Jewish or Indian origin. Pemphigus vulgaris is not an infection and therefore 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?

Skin erosions (sores) are painful and can affect quality of life including disturbing sleep. Mouth and throat 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, urination and bowel movements, the nose causing stuffiness and blood-stained crusts; and the conjunctiva of the eyes causing sore, red eyes (requiring assessment by an ophthalmologist).

The most common area to be affected is the inside of the mouth. This is most commonly the first area to be affected, and skin lesions often appear later. Other areas, as described above, are less commonly affected.

What does pemphigus vulgaris look like?

The skin:

- The skin lesions start as thin-walled fragile blisters (collections of clear fluid within the skin) that burst easily, leaving raw areas known as erosions.
- Erosions are sore, burn-like areas that can ooze fluid or become crusty and infected and when they heal, the skin may be discoloured.

The mouth:

- Blisters in the mouth quickly burst to form erosions. There may be one or two, or several, that join together.

Can pemphigus vulgaris be cured?

Pemphigus vulgaris is a long-lasting (chronic) disease and will not usually settle without treatment. There will be occasions when it flares up and other occasions when it improves. Currently, there is no way of predicting when flares will occur

or how severe they will be. Treatment is aimed at controlling the disease and preventing flares rather than a cure.

How is pemphigus vulgaris diagnosed?

- It is advisable your general practitioner refers you to a dermatologist or an oral medicine specialist. These specialists will be able to make a provisional diagnosis of pemphigus vulgaris by examining the affected area of skin and in the mouth.
- A *biopsy* of a blister may be taken 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 test to monitor how active the disease is and can help determine if a change of treatment is needed.

How can pemphigus vulgaris be treated?

General aims. Treatment is important because severe pemphigus vulgaris is a serious and potentially life-threatening condition if not treated early and effectively. Treatment involves long-term use of oral medicines. In severe cases, intravenous medication may be needed as a hospital in-patient. Most treatments work by suppressing the immune system.

The aims of treatment are to prevent new blisters forming and to heal broken areas of skin. It usually takes about 2 to 3 weeks to stop new blisters developing, and 6 to 8 weeks for healing to occur. Once the disease is controlled, the dose of the medication can be reduced slowly to the lowest effective level. Treatment usually starts with a corticosteroid and is often combined with a 'steroid-sparing' drug (see below).

Corticosteroids. Pemphigus vulgaris is usually treated first with an oral corticosteroid. 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 corticosteroids, usually prednisolone, are given to bring pemphigus vulgaris under control. The dose is then reduced slowly to minimise

side effects. Most people need to continue taking a small daily dose to keep the disease under control.

It is dangerous to stop oral corticosteroid treatment suddenly because your body may have become highly dependent on this medication as the body stops making its own natural corticosteroids. If you have concerns about your steroid treatment, please consult your doctor.

Steroid-sparing drugs. corticosteroids have many side effects when taken long-term at high doses. For this reason, other drugs are prescribed to allow the dose of steroid to be minimised. These are known as steroid-sparing drugs.

They include the following:

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

Side effects. These medications can have serious side effects, so people who take them must be monitored carefully including regular blood tests.

Other treatments. Plasmapheresis and intravenous immunoglobulin therapy may be considered if other treatments are not effective. They may be used in combination with steroid tablets. Further research continues to find better treatments or combinations of treatment for pemphigus vulgaris.

Skin and mouth topical treatments. Moisturisers and *strong* steroid creams may be used on skin blisters to help reduce the dose of steroid tablets.

Maintaining good oral hygiene is very important. The use of soft toothbrushes and mint-free toothpaste may be helpful. Mouth blisters and erosions may be treated with steroid sprays or mouthwashes containing an antiseptic or local anaesthetic. Sometimes, yeast over growth (thrush/candida) in the mouth may need anti-fungal treatment.

Self-care (What can I do?)

- Keep all of your appointments with your doctor and hospital specialist.
- Take medicines as advised by your specialist and never stop taking corticosteroids suddenly without talking to your doctor.
- If you have mouth erosions avoid eating spicy, acidic or hard foods.

- See your dentist or hygienist regularly for check-ups.
- Tell your doctor if your nose, throat or genital skin are affected.

Where can I get more information?

References:

British Association of Dermatologists guidelines on the management of Pemphigus Vulgaris 2017 (in press)

Link to patient support groups:

The Pemphigus Vulgaris Network
http://www.pemphigus.org.uk/

PEM Friends
www.pemfriends.org.uk

The international Pemphigus and Pemphigoid Foundation
http://www.pemphigus.org/

Web links to detailed leaflets:

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

Jargon Buster: www.skinhealthinfo.org.uk/support-resources/jargon-buster/

Please note that the BAD provides web links to additional resources to help people access a range of information about their treatment or skin condition. The views expressed in these external resources may not be shared by the BAD or its members. The BAD has no control of and does not endorse the content of external links.

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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