



Pemphigus Vulgaris (PV)

What are the aims of this leaflet?

This leaflet has been written to help you understand more about Pemphigus. It tells you what it is, what may cause it, how it affects the skin and mouth, what can be done about it and where you can find out more about it. (Additional information may be found in the British Association of Dermatology patient information leaflet – see link at the end of this leaflet)

What is Pemphigus?

Pemphigus is a rare disease that may lead to blistering or ulceration of the mouth, throat, nose and genitals (mucous membranes). Some patients also develop blisters on the skin. Although it can start at any age it most commonly affects people between 40 and 60 years. Women and men are affected equally. Some races are more prone to the disease e.g. those from the Indian subcontinent.

What causes Pemphigus?

Pemphigus is an autoimmune disease. For an unknown reason, antibodies (natural substances normally important in defending your body against infections) develop in the blood and mistakenly react with the surface layer of your mouth (or other mucous membranes) or skin causing blisters, which usually break down to leave ulcers. These 'autoantibodies' target proteins that in health hold the layers of the skin and mucous membranes together. As a consequence, the tissues become fragile and first blister and then ulcerate.



Is Pemphigus hereditary?

There is some evidence to suggest that the tendency to develop pemphigus is inherited in some patients but there is very rarely more than one family member affected.

What are the symptoms of Pemphigus?

The ulcerated areas of the mouth are very painful and can make eating and wearing dentures difficult. The pain can be made worse by eating, particularly hot, salty, spicy or hard / abrasive food. If the gums are affected, they may become sore, particularly when brushing the teeth. Blistered or ulcerated areas of skin and genitals can be painful and affect sleep.

What does pemphigus look like?

In the mouth, pemphigus causes shallow blisters which burst easily to form painful areas of ulceration. Ulceration can affect the lips, tongue, inside the cheeks, floor of mouth and palate extending to the throat. The gums can also be affected (desquamative gingivitis), and often bleed easily when brushed. Pemphigus in the skin presents as blisters which can burst to form ulcerated areas known as erosions. These can weep and sometimes become infected. Healing areas will crust over particularly if the scalp is affected. Lesions on the genitals are similar to the mouth.

How is Pemphigus diagnosed?

A skin or mouth biopsy will be carried out to confirm the diagnosis. This involves taking one or two small samples of tissue under local anaesthetic and sending these to the laboratory for analysis. The diagnosis of pemphigus can be confirmed by testing for specific antibody markers. A blood test is also taken to check for the presence of these antibodies. Your doctor may wish to monitor the level of antibodies in the blood as treatment progresses.

Can Pemphigus be cured?

There is no curative treatment for pemphigus, but it can be well controlled in the majority of patients. The severity of the condition varies widely among patients. In some patients the disease may be very mild and in others it may be more severe. It is usually well controlled with medication and in some patients, may go into long term remission i.e. they are symptom free and the treatment can be discontinued. There is always the chance however that it might relapse at a later stage.

How is Pemphigus treated?

Pemphigus is treated with a combination of tablets and topical treatments (mouthwashes or creams on the skin). The choice of treatment will depend upon the severity of the disease and how well a patient can tolerate a specific treatment. It may be necessary to try different treatments before finding the most suitable for an individual patient. The aims of treatment in pemphigus vulgaris are to decrease blister formation, promote healing of blisters and erosions. Patients need careful monitoring by a specialist particularly at the start of treatment to control the disease quickly and effectively. Once the disease is in remission, the strength of treatment will be slowly reduced to the minimal dose of medication necessary to control the disease process.

Topical treatments

For the mouth:

- Anaesthetic (analgesic) mouthwashes are available if your mouth becomes sore and are particularly helpful if used before meals. Benzydamine (Difflam) mouthwash may be helpful.
- Topical steroids which can be applied locally to the mouth are helpful for most patients but often require additional systemic medication (tablets). These are

available as mouthwashes, sprays, pastes and small pellets which dissolve in your mouth. Topical steroids can sometimes cause oral thrush which can be easily treated or prevented with topical antifungals.

- An antiseptic mouthwash or gel may be recommended to help with your plaque control, particularly at times when your gums are sore. Daily hydrogen peroxide mouthwash (Peroxyl) or occasional chlorhexidine (Corsodyl) twice per week are examples. If possible, avoid a mouthwash containing alcohol.

For skin, scalp or genital lesions:

- Corticosteroid ointments or creams are applied regularly

Systemic treatment:

In most cases of PV, treatment taken by mouth may be required for several months or years. Your specialist will discuss with you risks and benefits of the different drug options available. Regular blood tests are required, to screen for drug toxicity, when taking systemic drugs, particularly during the early stages of treatment.

- Oral corticosteroids may be used for a few months in high doses or, by some specialists, for much longer periods at low doses. However long-term treatment with corticosteroid tablets has many potential side-effects and therefore precautionary measures are required (for example medication for bone protection)
- Other drug treatments which further 'dampen down' the immune system are added so that the dose of corticosteroid can be reduced as soon as possible. These include azathioprine and mycophenolate mofetil. They are usually well tolerated but also require careful monitoring and can be associated with a number of side-effects which should be discussed with your specialist. Regular blood tests are often required when taking these drugs, particularly during the early stages of treatment.

- The most severe cases of pemphigus may require drug treatments given directly into a vein. These include corticosteroids, intravenous immunoglobulin, cyclophosphamide and rituximab.

What can I do?

- It is very important to attend your appointments regularly to ensure that your pemphigus is managed optimally and safely. The sooner the condition is diagnosed and treated effectively the better the chance of a successful remission.
- Please do not stop your corticosteroid (prednisolone) tablets suddenly as this may be harmful. Please discuss any side effects of treatment with your doctor or specialist
- Avoid spicy, acidic or salty foods if these make your mouth sore.
- Have your mouth checked on a regular basis by a dentist, or oral specialist.
- If your gums are affected (desquamative gingivitis), it is important that you keep your teeth as clean as possible. If not, a build-up of debris (known as plaque) can make your gum condition worse. Your dentist/dental hygienist will be able to give oral hygiene advice and will arrange for scaling of your teeth as necessary. Keep your teeth clean by using a soft brush and small interdental brushes.
- Choose a toothpaste with a mild flavour and free from the foaming agent, sodium lauryl sulphate (SLS).
- It is advisable to stop smoking and reduce your alcohol intake to recommended limits (currently 14 units a week for both men and women).



Further Information

www.pemphigus.org.uk - The Pemphigus Network

www.bad.org.uk (British Society of Dermatologists BAD)

www.pemphigus.org (International Pemphigus Foundation)

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

www.e-medicine.com

This leaflet has been prepared by the British & Irish Society for Oral Medicine (BISOM) in conjunction with the British Association of Dermatologists (www.bad.org.uk). It is reviewed periodically to reflect relevant advances and improved understanding. Not all the information will be relevant to all patients. For individual advice please see your Oral Medicine specialist. The BISOM is not responsible for information on web sites where links are provided.



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