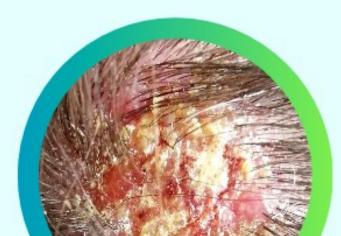
## **PEM Friends**

Enables patients and their loved ones to share information and advice on how to manage their symptoms and to get emotional support.

We also raise awareness of PEM in the medical and research community and can help Health Care Professionals access information and support.

Enquiries and requests are all welcome and PEM Friends would be delighted to hear from anyone willing to provide support and share their experiences.



## **GETTING IN TOUCH**

Read more in our magazine PEM Lives, available on our website: <u>https://www.pemfriends.org.uk/pem-lives</u> <u>-magazine</u>

We welcome any contacts via our website, our Facebook page or by e-mail at: <u>mail@pemfriends.org.uk</u>

Scan QR code for PEM Friends website:





### https://www.bad.org.uk



PEM Friends is a registered charity; Number 1207029

# Pemphigus Vulgaris



# Patient Information Leaflet



### What is Pemphigus Vulgaris?

Pemphigus Vulgaris is a rare autoimmune blistering disease (AIBD) causing severe blistering of the skin and the mucous membranes lining the mouth, nose, throat and genitals.

The blisters are fluid-filled sacs that develop in the upper layer of skin and mucosa and are very thin and fragile, breaking easily to leave raw areas that can be extensive and painful. Pemphigus Vulgaris (PV) always needs treatment by a specialist. PV affects males and females equally and can start at any age but most often in adults aged 50-60.

It affects people of all races but is more common in some. The blisters and erosions often start in the mouth and then on the skin. They may also affect other mucous membranes. Those in your mouth can interfere with eating and drinking leading to weight loss. Sites often overlooked include around your nails, the pharynx and larynx (pain on swallowing and hoarseness), and your nasal cavity (nasal congestion and a bloody mucous discharge). As many as 49% of patients were shown to have laryngeal and nasal involvement.

### How is Pemphigus Vulgaris diagnosed?

A biopsy of a blister may be taken to confirm the suspected diagnosis. This will be processed in a laboratory and examined under a microscope. Part of the biopsy sample will be examined using a technique known as direct immunofluorescence.

Pemphigus Vulgaris autoantibodies measured in the blood (by a specialist technique known as ELISA) is another test which monitors how active your disease is.

### How long will my Pemphigus Vulgaris last?

PV is a chronic autoimmune disease with no known cure. Suppressing your immune system keeps the disease under control and remission is possible.

The initial goal of treatment is to clear existing blisters and help prevent relapses. Treatments deployed typically depend on the severity and stage of your disease. Before corticosteroids, PV had a high fatality rate but nowadays, with treatment, your lesions can heal normally.



Most patients treated for PV will gain remission in 2 to 5 years.

### How is Pemphigus Vulgaris treated?

The key to bringing your PV under control is by suppressing your immune system. This can only be achieved by a range of drugs, and can take time and the use of different drugs and/or a combination of drugs to find a treatment that works best for you. It can take longer for mucosal lesions such as those inside your mouth to heal.

Treatment usually starts with a corticosteroid and once your disease is under control with no new blisters forming, tapering the steroids down can begin with an alternative drug replacing them. These "steroid-sparing" drugs are regarded as safer than steroids but they have their own side effects. Drugs most commonly prescribed for PV include Mycophenolate, Azathioprine, and Cyclophosphamide. A treatment called rituximab, given as an infusion, has been shown to be highly effective in bringing PV into remission. Topical steroid treatments also help your lesions heal on the skin and the mucus membranes. Mouth blisters and erosions may be treated with steroid mouthwashes or sprays.