



PATIENT INFORMATION LEAFLET

PEMPHIGUS FOLIACEUS

WHAT ARE THE AIMS OF THIS LEAFLET?

This leaflet has been written to help you to understand more about pemphigus foliaceus. 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 FOLIACEUS?

Pemphigus foliaceus is a rare blistering autoimmune skin disease that only impacts the skin. It affects less than 5 people per 1 million people worldwide each year. It should not be confused with [pemphigus vulgaris](#) which is a more severe related condition. The blisters that form in Pemphigus Foliaceus have a delicate outer surface which can be easily damaged leaving extensive raw areas (erosions) which can be painful.

WHAT CAUSES PEMPHIGUS FOLIACEUS?

Our immune system makes antibodies to fight infection. Normally, these antibodies do not attack our own body. However, in pemphigus foliaceus, the immune system makes antibodies (autoantibodies) that attack the "glue" holding the outer layer of the skin (epidermis) together, making it fragile and prone to blistering. It is not clear why some people develop these autoantibodies.

- The autoantibodies in pemphigus foliaceus attack proteins called desmogleins. These proteins form the "glue" that holds the cells of the skin together.
- When the autoantibodies formed in pemphigus foliaceus attack the desmoglein proteins, the cells in the

skin no longer hold together and separate. This causes blisters that are typical of pemphigus foliaceus.

Pemphigus foliaceus affects men and women equally. The average age of onset is usually 50 to 60 years. It can rarely occur in children. More commonly, it occurs in people originating from North Africa, Turkey, and South America. Pemphigus foliaceus is not an infection and therefore cannot be caught or passed on to anyone else.

IS PEMPHIGUS FOLIACEUS HEREDITARY?

Pemphigus foliaceus does not pass from generation to generation. However, genetic factors (the genes we inherit from our parents) are thought to be important if someone develops pemphigus foliaceus in combination with environmental factors.

WHAT ARE THE SYMPTOMS OF PEMPHIGUS FOLIACEUS?

Pemphigus foliaceus commonly affects the scalp, face, torso, armpits and genital areas. The skin lesions are typically small, scattered blisters that rapidly evolve into scaly, crusted erosions. Skin erosions (sores) are painful and can affect quality of life including disturbing sleep. Pemphigus foliaceus does not affect other parts of the body, it only affects the skin.

WHAT DOES PEMPHIGUS FOLIACEUS 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. This can lead to discolouration of the skin.

CAN PEMPHIGUS FOLIACEUS BE CURED?

Pemphigus foliaceus is a long-lasting (chronic) condition. However it usually can be well controlled with 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 condition and preventing flares.

HOW IS PEMPHIGUS FOLIACEUS DIAGNOSED?

- It is advisable that your general practitioner refers you to a dermatologist. A dermatologist can make a provisional diagnosis of pemphigus foliaceus by examining the affected area of skin.
- A biopsy of a blister may be taken for examination and to confirm the diagnosis. Part of the biopsy sample will be examined by a technique known as direct immunofluorescence to demonstrate the presence of pemphigus autoantibodies in the skin.
- Pemphigus autoantibodies can also be detected with a blood test. This can show how active the disease is and determine if a change in treatment is needed.

HOW CAN PEMPHIGUS FOLIACEUS BE TREATED?

General aims. The aims of treatment are to prevent new blisters forming and to heal broken areas of skin. Treatment involves long-term use of topical and oral medicines.

In severe cases, intravenous medication may be needed as a hospital in-patient. Most treatments work by suppressing the immune system.

Topical Corticosteroids

Steroid creams and ointments are the most commonly prescribed treatment for pemphigus foliaceus. **Topical corticosteroids** are anti-inflammatory medications that work by suppressing the immune system. They relieve symptoms and calm inflammation and blistering. Higher strength steroids are usually needed and are typically applied once a day. When applied as directed by your healthcare professional, side effects are rare.

Tetracycline antibiotics

These are helpful for the inflammatory element of pemphigus foliaceus and used as a steroid sparing measure. These are used in the long term for their anti-inflammatory properties. Doxycycline is commonly used and is suitable for people with a penicillin allergy.

Oral Corticosteroids

High doses of **oral corticosteroids**, usually prednisolone, are often given to bring pemphigus foliaceus under control (by suppressing the immune system). Once the condition is under control, the dose is then reduced slowly to minimise side effects. Some people need to continue taking a small daily dose to keep the disease in remission. Rarely, intravenous corticosteroids may have to be used in the form of IV pulse methylprednisolone.

Immunosuppressants

People affected by pemphigus foliaceus often require an additional immunosuppressant agent. This is to allow the **oral steroid** dose to be weaned. Immunosuppressants work by dampening down the immune system and are given under the close supervision of a dermatology consultant. Options include:



- [Azathioprine](#)
- [Mycophenolate mofetil](#)
- [Methotrexate](#)
- [Rituximab](#)

Side effects. These medications can have side effects, so people who take them must be monitored carefully including regular blood tests. More information can be found in the individual patient information leaflets for the corresponding drug.

Other treatments

Cyclophosphamide and intravenous immunoglobulins may be considered if other treatments are not effective. Further research continues to find better treatments or combinations of treatment for pemphigus foliaceus.

Other practical tips:

- Keep all of your appointments with healthcare professionals.
- Adhere to the blood monitoring regimen your doctor gives you.
- Take medicines as advised by your specialist and never stop taking corticosteroids suddenly without talking to your doctor.
- You may be provided with a steroid card to alert other healthcare professionals that you are taking this medication, and provide guidance on what to do if you become unwell.

WHERE CAN I GET MORE INFORMATION?

Patient support groups:

The Pemphigus Vulgaris Network

Web: <http://www.pemphigus.org.uk/>

PEM Friends

Web: <http://pemfriendsuk.co.uk/>

The international Pemphigus and Pemphigoid Foundation

<http://www.pemphigus.org/>

Web links to detailed leaflets:

<https://dermnetnz.org/topics/pemphigus-foliaceus>

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

BRITISH ASSOCIATION OF
DERMATOLOGISTS

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