



EUROPEAN
ACADEMY OF
DERMATOLOGY &
VENEREOLOGY

Information Leaflet
for Patients

Pemphigus vulgaris and pemphigus foliaceus



The aim of this leaflet

This leaflet is designed to help you understand more about pemphigus. It tells you what this condition is, what causes it, how it is diagnosed and treated, and practical advice for managing this condition.

Pemphigus vulgaris and pemphigus foliaceus

What is bullous pemphigus?

Pemphigus is a rare group of bullous diseases of the skin and mucous membranes of autoimmune origin. It is a very rare disease whose incidence (number of new cases per year) is about 1 to 2 people per million inhabitants in Central Europe with higher numbers (about 5 per million per year) in the Mediterranean countries. Pemphigus affects people of all ages, ethnic backgrounds and genders. Nevertheless, people between 50 and 60 years of age with Mediterranean background are more often affected. There is no difference between men and women.

What does pemphigus look like, and what are the signs and symptoms?

It is characterised by the appearance of painful blisters and superficial wounds on the skin and/or mucous membranes (mouth, nostrils, throat, eyes, oesophagus, genital mucosa, anus). Not all skin and mucous membranes are necessarily affected.

There are three forms of pemphigus:

1. Pemphigus vulgaris (or common pemphigus)

The term "vulgaris" means "common". Pemphigus vulgaris is the most common form of pemphigus in Europe. It can affect either the mucous membranes (the most frequent) or the skin and mucous membranes, rarely the skin in isolation.

It appears as blisters that are pierced almost immediately after their appearance and leave superficial wounds which are often painful. The face, scalp and chest are often affected. All the mucous membranes mentioned above can be affected, but it is the mouth that is most frequently affected.

2. Pemphigus foliaceus (or pemphigus superficialis)

In the case of pemphigus superficialis, there is no damage to the mucous membranes, only the skin is affected. Crusty wounds or fragile blisters usually appear on the face and scalp, on the chest, back, arms and legs.

Another rare pemphigus form, **paraneoplastic pemphigus**, is described in a separate leaflet.



Fig. 1 Mucosal wounds seen in a patient with *pemphigus vulgaris*



Fig. 2 Cutaneous lesions seen in a patient with *pemphigus vulgaris*

What causes the blistering?

Our immune system produces specialised proteins called *antibodies*, which bind to bacteria, viruses, fungi (the plural of fungus) and tumor cells, thus protecting us both from infections and cancer. In patients with pemphigus vulgaris/ foliaceus, the immune system mistakenly produces antibodies that bind to certain structures in the skin and mucous membranes. These disease-causing antibodies are called autoantibodies (antibodies against one's own body). This binding triggers a complex immunological process that finally leads to the cell detachment and erosions in skin and mucous membranes.

How is pemphigus diagnosed?

The dermatologist examines the appearance, number and location of blisters and small wounds (erosions). This is the clinical analysis.

Under local anaesthesia, (s)he takes two samples of small skin fragments (biopsies). These analyses will allow us to see:

- at what depth in the epidermis (uppermost part of the skin) the blisters are formed. This is the *histological analysis*.
- autoantibodies that attack the junction membrane (basement membrane) between the epidermis and dermis. This is the *direct immunofluorescence analysis*.

A blood test may be performed to determine the type and number of autoantibodies. This is the biological analysis (most often using ELISAs).

How does pemphigus develop?

Pemphigus evolves over time by successive outbreaks. It can be severe if left untreated.

As blisters disappear, they sometimes leave coloured (pigmented) spots but

they do not leave scars.

With the treatment given at the beginning (attack treatment), blisters and erosions cease to appear in 15 days to 1 month on average. The disease is said to be "controlled" and treatment is gradually reduced, becoming milder. This lighter treatment (maintenance treatment) is necessary for several years to prevent relapses.

Before diagnosis, and between the initiation of treatment and the control of the disease, hospitalisation can be necessary. Lesions can be widespread, infected or responsible for severe skin and mucous membrane pain requiring specific local treatment and care. Eating may be difficult because of lesions in the mouth and throat.

When the disease is controlled, follow-up treatment is done during a medical consultation and/or in day hospital.

What are risk factors and associated diseases?

The triggering factor(s) of this disease are not known. It may be caused by a highly stressful event. In exceptional cases, the disease may be caused by certain specific drugs, such as D penicillamine, ACE inhibitors, beta blockers, penicillin, phenylbutazone, pyritinol, thiopronine. Most often there is no cause.

Family forms (several cases occurring in the same family) are extremely rare and there is no reason to worry family members about this. No genetic testing is required.

Most often there is no cause.

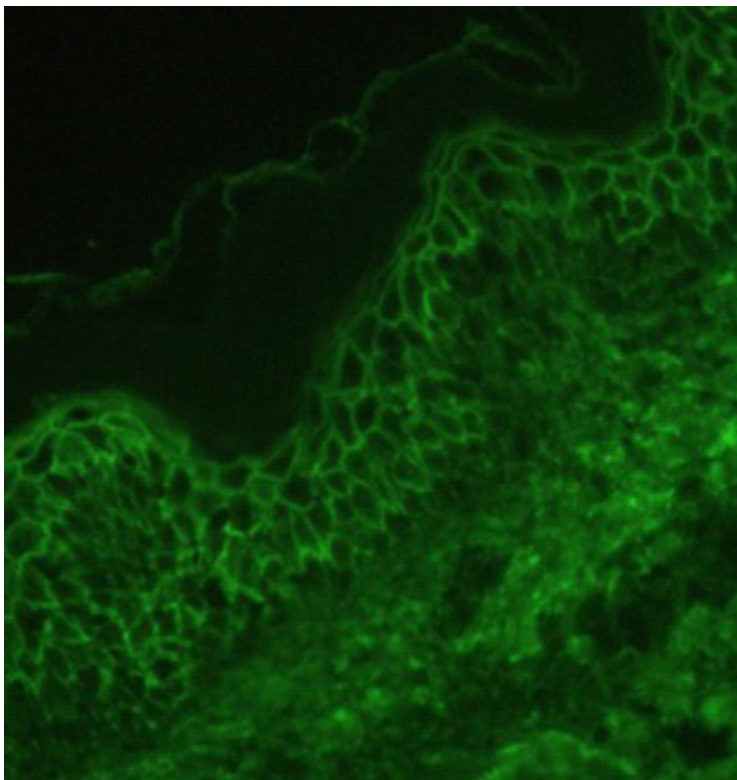


Fig. 3 Aspect of a patient's skin biopsy examined using direct immunofluorescence microscopy, showing the autoantibodies in a net-like pattern typical of pemphigus.

Pemphigus vulgaris and pemphigus foliaceus

How is pemphigus treated?

The treatment of pemphigus is very effective.

It generally takes place in two phases:

- Initial treatment (attack treatment) usually involving high doses of medication to stop the progression of the disease and achieve healing of the lesions.
- Disease maintenance treatment to maintain remission (no relapse), usually continued for several years.

Attack treatment is based on systemic corticosteroid therapy alone or in combination with an immunosuppressive drug (which blocks the production of autoantibodies), most often rituximab, more rarely mycophenolate mofetil or azathioprine. This treatment is then reduced very gradually to avoid relapses.

Local treatments with corticosteroids in cream or mouthwash can be very useful to accelerate the healing of lesions and improve comfort. In case of pain, analgesic (pain relief) treatments should be used.

Discontinuation of maintenance treatment should be discussed on a case-by-case basis and should only be done in consultation with the dermatologist. If treatment is ended too early, it usually results in a relapse of the disease requiring resumption of treatment.

Can pemphigus be prevented?

So far, no preventive measures to avoid the occurrence of pemphigus are known.

What is practical advice for taking care of pemphigus?

- Pemphigus is a rare disease and is best treated in a specialized clinic experienced with this condition.
- Since some drugs may interfere with those prescribed for pemphigus, please inform your dermatologist about any new drug prescribed since your last visit.
- There are country-specific and international patient support groups for patients suffering from pemphigus, and they may help you or your friend/family member find further help.

Patient support organisations

UK: www.pemfriends.co.uk and www.pemphigus.org.uk

Germany: www.pemphigus-pemphigoid-selbsthilfe.de

Italy: www.pemfigo.it

France: www.pemphigus.asso.fr

Turkey: www.turkdermatoloji.org.tr

International Pemphigus and Pemphigoid Foundation: www.pemphigus.org

While every effort has been made to ensure that the information given in this leaflet is accurate, not every treatment will be suitable or effective for every person. Your own clinician will be able to advise in greater detail.