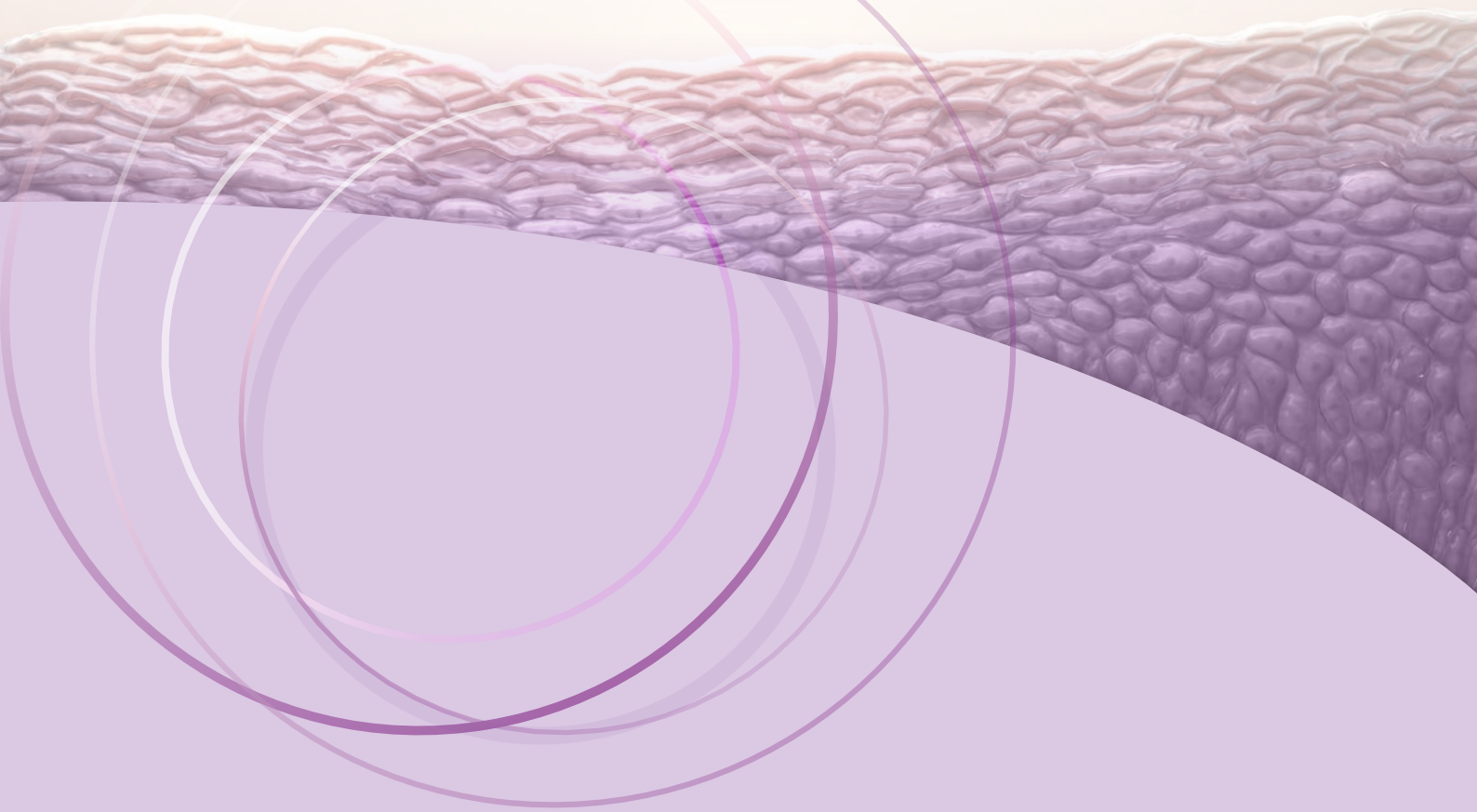
EADV

EUROPEAN ACADEMY OF
DERMATOLOGY AND VENEREOLOGY

Information Leaflet
for Patients



BULLOUS PEMPHIGOID

The aim of this leaflet:

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

BULLOUS PEMPHIGOID

What is bullous pemphigoid?

Bullous pemphigoid is a chronic, blistering, autoimmune disease of the skin. The occurrence of this disease rises considerably with age, and it mainly affects patients over 70 years old. With about 1-4 newly diagnosed patients per 100,000 individuals per year, bullous pemphigoid is a rare disease.

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

Patients almost always suffer from severe itching accompanied by *tense* (taut or stretched) blisters on red or otherwise normal skin (**Fig. 1**). Itching may start weeks or even months before the first skin manifestations arise. The blisters contain a clear or bloody fluid, and develop further into *erosions* (skin breakdown) and *crusts* (scabs) when damaged, but they normally heal without scarring. In about one-fifth of patients, *erosions* may develop in the mouth or on the genitalia.

Initially, blisters may not be present, and skin lesions may look more like *eczema* (inflamed, often itchy skin) erythema, or *urticaria* (hives) (**Fig. 2**).

While blisters only appear on some parts of the body in some patients, in others, the whole body is affected. Apart from intensive itching, general health is not affected in most patients. However, loss of appetite, body weight, fever, infections, and general weakness can develop during the course of the disease.

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, and protect us from these infections as well as cancer. In patients with bullous pemphigoid, the immune system mistakenly produces *antibodies* that bind to certain structures in the skin and mucous membranes, termed BP180 and BP230. These disease-causing *antibodies* that bind to the skin are called *auto-antibodies* (*antibodies* against one's own body). This binding triggers a complex inflammatory reaction that finally leads to the separation of the upper layer of the skin from the deeper layers. So far, it is unclear what

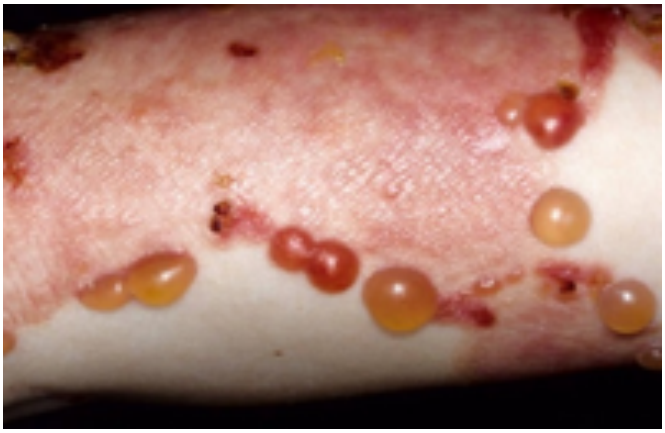


Fig. 1 Clear and haemorrhagic (bloody) *tense* blisters, *crusts* and *erythema* (red skin) on the arm

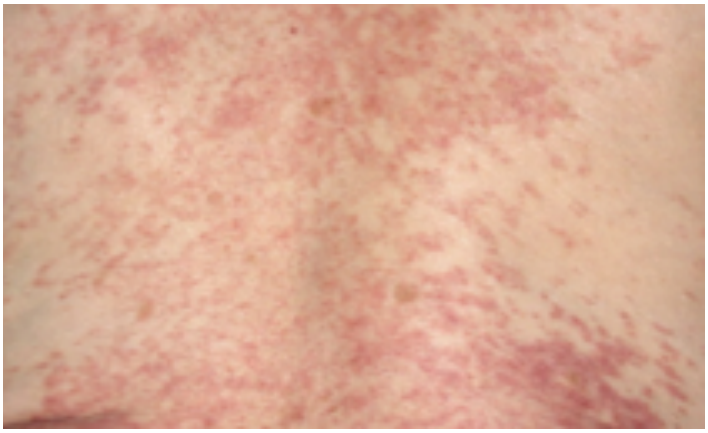


Fig. 2 *Erythematous* (reddish) *macules* (spots) on the back

factor(s) trigger(s) the immune system to produce the disease-causing *auto-antibodies* against BP180 and BP230. In some patients, certain medications have been associated with the onset of the disease (see below in the section "**What are risk factors and associated diseases?**").

How is bullous pemphigoid diagnosed?

The clinical characteristics and older age of patients already give important clues to the diagnosis of bullous pemphigoid. However, for a specific diagnosis, the detection of *auto-antibodies* against BP180 and BP230 is required. These *auto-antibodies* can either be detected in the skin (**Fig. 3**) or blood. Therefore, a skin biopsy (usually 4 mm in diameter) is necessary to detect the *auto-antibodies* in the skin. This procedure is done under local anesthesia in a few minutes.

For the detection of *auto-antibodies* against BP180 and BP230 in the blood, a blood sample is taken. As we know that a higher level of *auto-antibodies* against BP180 in the blood is associated with more skin lesions, blood may also be taken during the course of the disease to better adjust the treatment dose.

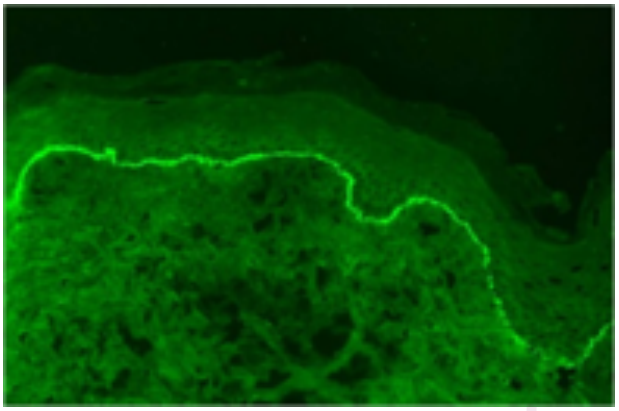


Fig. 3 Immunoglobulin G (IgG) *auto-antibodies* can be detected as a linear band in a skin biopsy (by direct immunofluorescence microscopy)

How does bullous pemphigoid develop?

Bullous pemphigoid usually progresses to become a chronic disease, and treatment may be required for months and sometimes years. During this time, new blisters and *erosions* can appear. For this reason, regular follow-up visits, ideally in a specialized outpatient department, are recommended to allow dermatologists to assess disease activity, adapt the medication, check for possible side effects, and measure the blood levels of *auto-antibodies*.

What are risk factors and associated diseases?

Some medications, such as gliptins used in the treatment of diabetes, or chronic use of spironolactone or phenothiazine, may trigger the onset of bullous pemphigoid. If bullous pemphigoid is diagnosed, these drugs may be replaced if possible by your general practitioner in collaboration with your dermatologist.

Approximately 30-50% of patients with bullous pemphigoid suffer from neurological disorders including Parkinson's disease, stroke, multiple sclerosis, or dementia. No data are available yet about the risk of developing neurological problems after the diagnosis of bullous pemphigoid has been made. In some patients, the disease is associated with cancer of the blood and lymph nodes. Your dermatologist or general physician should rule out those malignancies.

How is bullous pemphigoid treated?

In general, therapeutic options include local (e.g., ointments and creams) and systemic (oral) treatments (tablets), often in combination. In mild disease, daily treatment with strong corticosteroid ointments twice a day is sufficient. In

BULLOUS PEMPHIGOID

moderate to severe disease, an additional systemic treatment may be required. As systemic treatment, cortisone tablets are usually given. In order to use the lowest dose of cortisone as possible, additional anti-inflammatory drugs (like doxycycline or dapsone) or immunosuppressants (such as methotrexate, azathioprine or mycophenoles) are given. Upon treatment, skin lesions will usually heal within a few weeks, and the dose of medication can be decreased. However, skin lesions may come back in up to 30% of patients with bullous pemphigoid, and treatment must then be adjusted or restarted.

Can bullous pemphigoid be prevented?

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

What is practical advice for taking care of bullous pemphigoid?

- Bullous pemphigoid is a rare disease and is best treated in a specialized clinic experienced with this disorder.
- Since some drugs may interfere with those prescribed for bullous pemphigoid, please inform your dermatologist about any new drug prescribed since your last visit.

Patient support groups

There are country-specific and international patient support groups for patients suffering from bullous pemphigoid, and may help you or your friend/family member find further help.

Several national patient support group for BP and related diseases have been established including

- France: www.pemphigus.asso.fr; pemphigus.asso77@laposte.net
- Germany: www.pemphigus-pemphigoid-selbsthilfe.de; pemphix-shg@gmx.de
- Italy: www.pemfigo.it
- Netherlands: www.pemphigus.nl; mail@pemphigus.nl
- Spain: www.aeppeva.com; esthercamaramartinez@gmail.com

In addition, there is an US-based international patient support group, the *International Pemphigus and Pemphigoid Fondation* (IPPF; www.pemphigus.org). ■

EADV

EUROPEAN ACADEMY OF DERMATOLOGY AND VENEREOLOGY

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.

PRODUCED BY THE EADV AUTO-IMMUNE BULLOUS DISEASES TASK FORCE

Publication date: 2019