Information sheet for patients affected by **BULLOUS PEMPHIGOID**

These information sheets were written in collaboration with the Reference Centre for Autoimmune Bullous Diseases and the Pemphigus-Pemphigoïde France patient association.

Dear Sir or Madam,

This sheet is intended to provide you with information on bullous pemphigoid. It is not a substitute for a medical consultation.
Its purpose is to promote dialogue with your doctor. Feel free to ask him or her to specify any points that do not seem sufficiently clear to you and to ask for additional information on your particular case.
Indeed, some of the information contained in this sheet may not be adapted to your case: it is important to remember that each patient is different. Only your doctor can give individualised and adapted information.

The disease

What is bullous pemphigoid?

Bullius pemphigoid is an autoimmune skin disease, which means that the body, following a disruption of the immune system*, produces antibodies against its own skin (autoantibodies). It mainly affects people over 70 years of age.

Lesions are generally confined to the skin (very rarely affecting the mucous membranes).

They are characterised by the appearance of blisters on red patches (erythematous plaques), located mainly on the limbs and often causing intense itching (pruritus) that can appear even before the first skin lesions.

**Immune system: the set of defences of an organism (including white blood cells and antibodies) that normally allows it to recognise what belongs to its body and to defend itself against what is foreign to it (microbes for example). In this disease, it is mistaken and removes by mistake the skin and/or mucous membranes.*

Is the disease serious?

It is sometimes a serious disease requiring treatment for several months or even years.

What is it caused by?

Bullous pemphigoid is an autoimmune disease, caused by autoantibodies that prevent the attachment systems between the first two layers of the skin (epidermis and dermis) from functioning.

The causes of this immune system disorder are not yet well known. Some specific drugs have been suspected as possible triggers of the disease.

Who can be affected?

Bullous pemphigoid mainly affects people over 70 years of age.
Exceptionally, younger adults, or even children, may be affected.
Both sexes may be affected, although women are slightly more affected.

How many people have the disease?

Bullous pemphigoid is the least rare of all autoimmune bullous diseases. Its incidence (number of new cases per year) is around 1000 to 1500 new cases per year in France, or 15 to 23 new cases per year per million inhabitants. This incidence is much higher among the elderly. It reaches 200 cases per million inhabitants per year among people over 70 years of age.

How do we explain the symptoms?

Bullous pemphigoid is an autoimmune disease caused by autoantibodies directed against two proteins (AgPB230 and AgPB180), normally present at the junction between the dermis and the epidermis (dermo-epidermal junction), which are the first two layers of the skin (see diagram below).
These autoantibodies are responsible for the disease because they cause a detachment between the epidermis (upwards) and the dermis (downwards) and, consequently, the formation of blisters, commonly called "bubbles" and containing a clear liquid.

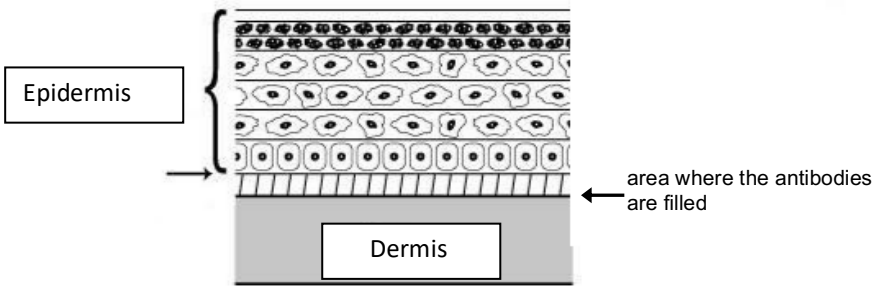

Diagram representing the epidermis (surface layer of the skin) and the dermis (deep layer).

Excerpt from the International Pemphigus Foundation website http://www.pemphigus.org

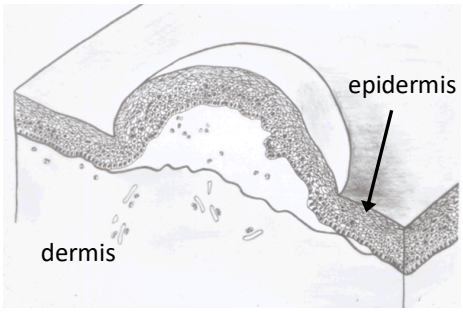

Diagram representing a blister.
C. Prost-Squarcioni

Is it contagious?

No, this disease is not contagious.

Is it hereditary?

No, this disease is not hereditary.

How is it diagnosed?

The dermatologist examines the appearance, number and location of blisters and small wounds left by bubbles (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 the detachment is located and 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 amount of autoantibodies. This is the biological analysis (most often using ELISAs).

How does it evolve?

Untreated, the disease lasts from several months to several years, evolving over time and often with successive relapses. It can be severe if left untreated.

As the blisters disappear, , they sometimes leave coloured spots (pigmented) or small white cysts (milium cyst) but they do not leave scars. With the treatment given at the beginning (attack treatment), blisters and plaques cease to appear in 15 days to 1 month on average. The disease is said to be "controlled" and treatment is gradually reduced over 2 to 3 months. A lighter treatment (maintenance treatment) is required for several months (6 to 18 months) to avoid relapses. A definitive cure is possible within 1 to 5 years.

Widespread and generalised forms of the disease often require admission to hospital in order to begin treatment and improve skin condition as quickly as possible with appropriate local care.

For more localised forms, follow-up treatment can be done in day hospital or even during a consultation (without hospitalisation) if the person's condition allows it, often with the help of home care nurses.
Patient follow-up, with blister count, must be performed in order to be able to monitor progress while on treatment.

Treatment, care, follow-up

Is there a treatment for this disease?

The treatment of bullous pemphigoid is very effective.

It is based on cortisone (also called corticosteroids). Its use during treatment is called corticosteroid therapy.

It is applied topically to the skin with high doses of cortisone cream all over the body, daily and over a long period of time. This treatment requires a lot of attention from the affected person and his or her family and friends. It is often desirable to have this treatment performed by a nurse at home. Treatment will be gradually reduced over 3 to 4 months and continued for an average of 10 to 12 months.

Be careful, relapses are always possible.

In some cases, corticosteroids can be used generally (in tablet form but not in cream).

Other treatments are used when corticosteroid therapy is ineffective (the patient is said to be corticosteroid-resistant, which is very rare) or if the patient relapses when stopping or lowering steroid doses (the patient is said to be corticosteroid-dependent).

These are immunosuppressants (drugs that inhibit the production of antibodies) such as methotrexate or mycophenolate mofetil (CellCept® or its generics) but also doxycycline (Tolexine®, Doxy 100®, Doxylis® or many other generics). These drugs can be very useful in limiting corticosteroid doses in case of relapse.

Is psychological support desirable?

Psychological support may sometimes be considered, at the beginning of the disease and in its severe forms, when there is a strong impact on the patient's quality of life.

Can this disease be prevented?

The onset of the disease cannot be prevented.

Living with bullous pemphigoid

People with the disease may have a lack of appetite, loss of weight and sleep, especially due to itching. This often happens at the beginning of the disease when it is not yet diagnosed.

In general, when the disease is controlled through treatment, the patient’s daily life returns to its previous state without the constraints of treatment and possible side effects.

The usual treatments are continued, but it is desirable to check for possible interference (= combinations of drugs not recommended) with corticosteroids and/or immunosuppressants or exceptional drug inductions.

A detailed fact sheet with tips for the daily life of patients with bullous disease (skin hygiene, clothing, nutrition, sun protection, physical activity)

is available on the website of the National Reference Centre for Autoimmune Bullous Diseases http://www.chu-rouen.fr/crnmba.

Where can you be treated?

In a dermatology department located in a hospital centre, at least initially.

In France, there is a reference centre and several competence centres for the management of autoimmune bullous diseases (contact details on the website of the national reference centre for autoimmune bullous diseases http://www.chu-rouen.fr/crnmba).

To know more

Where can I get more information?

How to connect with other patients with the same disease?

There is a French patient association "Association Pemphigus Pemphigoïde France" created by patients and their families in March 2005.

URL: http://www.pemphigus.asso.fr
Contact Numbers for questions and information requests:
*Hélène Facy (Paris area): +33 (0)687 114 026
* Bernadette Dejean (Western and Southern regions): +33 (0)683 395 320
*Jenny Vernet (Eastern and Northern regions): +33 (0)634 686 874 57
Telephone number of the association's headquarters: +33 (0)143 252 5 42 88

There are also websites dedicated to all rare diseases:

http://www.orpha.net

http://www.maladiesraresinfo.org (+33 (0)156 535 38136 non surcharged call)

This website provides answers to any questions that you may have during corticosteroid treatment:

http://www.cortisone-info.fr