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

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

Dear Madam,

This sheet is intended to provide you with information on pemphigoid gestationis. 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 pemphigoid gestationis?

Pemphigoid gestationis (or pregnancy pemphigoid), formerly known as herpes gestationis, is an extremely rare disease whose main triggering factor is pregnancy. It is a bullous skin disease of autoimmune origin. This means that the body, following a disruption of the immune system*, produces antibodies against its own skin (autoantibodies).

It usually begins in the second half of pregnancy, most often in the third trimester, and less frequently in the days following childbirth. The first symptoms are severe itching (pruritus), then lesions appear on the skin that can take several forms: urticaria (like after a nettle sting); blisters. These skin lesions start in most cases around the navel (umbilicus) and can in some cases become widespread. The face and mucous membranes are spared. The general condition is preserved.

**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?

1- IS THERE A RISK FOR THE MOTHER?

No, the lesions regress spontaneously or under treatment. The only potential risk is the occurrence of side effects of the treatment (general or local corticosteroid therapy - see below).

2- IS THERE A RISK FOR THE BABY?

For the baby, there is a risk of premature delivery and a risk that the birth weight will be lower than normal. This justifies close monitoring throughout the pregnancy.
Very rarely (in less than 5% of cases), the child may be born with a few blisters. This eruption will disappear alone and quickly, without sequelae.

What is it caused by?

Pemphigoid gestationis 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.

Who can be affected?

Pemphigoid gestationis only affects women of childbearing age (reproductive age), regardless of their geographical origin. Most often, the disease occurs in the second or third trimester of pregnancy; more rarely within a few days of delivery.

It occurs most often in women who have already had several pregnancies (if the father is the same each time). Exceptional cases of recurrence on the pill have been described.

How many people have the disease?

Pemphigoid gestationis is a rare disease.
Its incidence (number of new cases per year) is poorly known, ranging from one case per 5,000 to one case per 50,000 pregnancies in France.

How do we explain the symptoms?

Pemphigoid gestationis is an autoimmune disease caused by autoantibodies directed against two proteins (AgPB230 and AgPB180), 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.

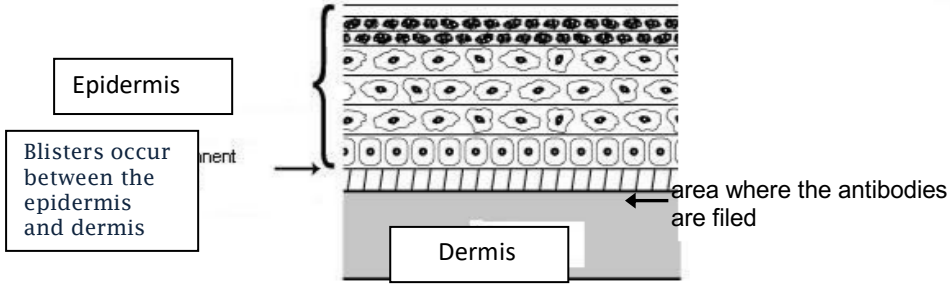

Diagram representing the epidermis (surface layer of the skin) and the dermis (deep layer).

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

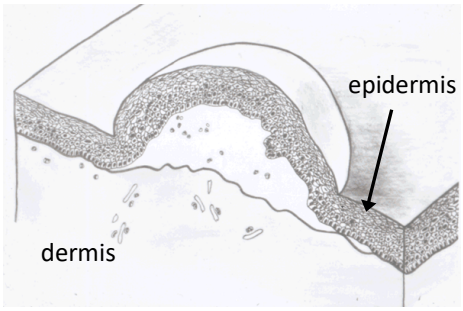

Diagram representing a blister.
C. Prost-Squarcioni

Is it contagious?

No, this disease is not contagious.

Is it hereditary?

Pemphigoid gestationis is not a hereditary disease. Even if the baby is born with a few blisters (less than 5% of cases), because it is the mother's autoantibodies (the baby does not make autoantibodies himself), these will therefore disappear on their own.
There is a genetic predisposition to develop the disease. Some genes transmit a terrain that predisposes to the development of pemphigoid

gestationis; but even in women with this gene, the risk of triggering the disease remains extremely low.

Family forms are therefore extremely rare and there is no reason to worry family members about this. No genetic testing is required.

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?

The rash on the skin evolves over time with successive flare-ups interspersed with periods of calm. An aggravation is possible at the time of delivery.

In general, skin lesions disappear spontaneously within one to two months after delivery. They can sometimes leave spots of colour, either lighter or darker than normal skin, which will then disappear.

Is there a risk for the following pregnancies?

The rash can recur in subsequent pregnancies (if the father is the same), with an earlier and more intense character with each new pregnancy.

Does the child risk having pemphigoid later on?

The child has no greater risk than the general population of later developing pemphigoid (i.e. a low risk: rare disease).

Treatment, care, follow-up

Is there a treatment for this disease?

The main treatment for pemphigoid gestationis is corticosteroids (cortisone-based drugs).

In less severe forms with a small number of new blisters appearing daily, local application of a cream containing corticosteroids (dermocorticoid) may be sufficient.
In more severe forms, oral corticosteroids are most often necessary, without adverse consequences on foetal development.

In both cases, the high doses at the beginning of treatment will be gradually adjusted according to the evolution of the rash. In some cases, treatment may have to be continued for a few weeks, or even exceptionally several months, after delivery.

Is psychological support desirable?

Psychological support may be considered in severe forms of the disease that have 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 pemphigoid gestationis

In general, when the disease is controlled through treatment, the patient's daily life is virtually unchanged outside the constraints (diet ± physical activity) and possible side effects associated with corticosteroid therapy.

Before diagnosis, and in the interval between the initiation of treatment and disease control, quality of life can sometimes be significantly impaired, and hospitalisation may be required.

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 38136non surcharged call)

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

http://www.cortisone-info.fr