



EUROPEAN ACADEMY OF
DERMATOLOGY AND VENEREOLOGY

Information Leaflet for Patients

PEMPHIGOID GESTATIONIS

The aim of this leaflet

This leaflet is designed to tell you more about Pemphigoid Gestationis. It tells you what the condition is, what causes it, and what can be done about it.

PEMPHIGOID GESTATIONIS

What is Pemphigoid Gestationis?

Pemphigoid Gestationis is a rare skin blistering disorder that occurs in pregnant women. It usually starts in pregnancy but can also return in women when they take oral contraceptive therapy or have their periods for some time after the pregnancy. It usually starts with an itchy rash that develops into blisters. It is most common during mid- to late-pregnancy (the 2nd and 3rd trimesters).

What causes it?

Pemphigoid Gestationis is an auto-immune blistering disease which means that the woman's immune system reacts against her own skin causing skin blistering. Female hormones (particularly oestrogen) are thought to aggravate the reaction, and this may be why it starts during pregnancy when oestrogen levels rise. *Pemphigoid Gestationis* may come back in following pregnancies, in some women who take oral contraceptive therapy, or with their periods following pregnancy, but later on there is usually no problem.

Does Pemphigoid Gestationis run in families?

No, but there is a link with other auto-immune diseases (which may run in families) such as thyroid disease and pernicious anaemia (anaemia due to B12 deficiency).

What are the symptoms of Pemphigoid Gestationis and what does it look like?

Itching is common and often starts around the umbilicus (belly button) during mid- to late-pregnancy (13 to 40 weeks of the pregnancy). Itching may be followed by a rash with large, red, inflamed areas of skin, and then blisters may develop later.

There is often a rash with *wheals* (like hives, nettle rash or *urticaria*) and large, raised, red patches commonly occurring on the trunk, back, buttocks, and limbs. The face, scalp, mouth, and genital area are usually not involved. Large tense blisters then appear on the red patches within 1-2 weeks, and are also seen on palms and soles. The blisters usually contain clear fluid or occasionally blood-stained fluid. There is usually no scarring when the blisters heal.

How is Pemphigoid Gestationis diagnosed?

Diagnosis requires a skin sample (biopsy) and/or a blood test for special laboratory tests (*immunofluorescence*). Usually the appearance and behaviour of the rash is very typical, but in early disease without blisters the rash can look like other skin diseases.

Can Pemphigoid Gestationis be cured?

No, but it can be suppressed with treatment. The symptoms often improve towards the end of pregnancy, but many women will experience a flare of the rash around the time of delivery. In most cases, symptoms resolve days or weeks after giving birth, but in some women the disease can remain active for months to years and may require continued treatment. Restarting periods and use of oral contraceptive therapy can all cause flare-ups of the disease for some time after delivery, as can future pregnancies.

How can Pemphigoid Gestationis be treated?

The primary aim of treatment is to relieve itching, prevent blister formation, and treat any secondary infection. It is also important during pregnancy to use treatments that are as safe as possible for both mother and baby.

The use of emollients, moisturising creams, and ointments can help with the inflammation and discomfort of the skin, as can bathing or showering in emollient and soap substitutes.

Steroid creams or greasier steroid ointments are necessary to relieve symptoms. The steroid cream or ointment should only be applied to affected areas of skin and a mild (for example hydrocortisone) or moderate steroid cream should be used. The amount should be as small as possible, and ideally only 1-2 small (15-30 g) tubes should be used for a few days intermittently. However, if the condition is severe, then application of a stronger steroid cream or ointment to the skin in larger quantities is still better than oral steroids.

Mild- to moderate-strength steroid creams or ointments appear to be safe during pregnancy, but stronger steroid creams and ointments may cause problems with the growth of the unborn baby, so that they may be born small especially if the mother is using large amounts (more than 50 gm, 1/2 large tube per month or over 200-300 gm, 2-3 large tubes, in the whole pregnancy) of steroid creams or ointments.

Treatment for more severe disease (with blistering) is usually with high doses of steroid tablets to get the disease under control rapidly. This needs careful monitoring and should involve obstetricians to care of the health of the mother and baby. The dose of steroid tablets may need to be increased at the end of pregnancy to prevent disease-flaring after delivery.

Antihistamines may be helpful to treat the itch.

The following are considered safe in pregnancy:

- Non-sedating: Loratadine, Cetirizine
- Sedating (cause drowsiness): Clemastine, Dimethindene, Chlorpheniramine.

Blisters may be burst (with a sterile needle) to offer relief from discomfort, and dressings can be applied to weepy or raw areas of skin. Other drugs may be used in more severe cases, or in women who experience severe disease following delivery.

Will the baby be affected?

Occasionally the baby will develop a blistering rash following delivery due to the mother's *antibodies* crossing the placenta. This only occurs in 5-10% of babies, and the rash is usually only temporary (lasting up to 6 weeks until the *antibodies* are cleared). Usually only mild treatment is required such as emollients, mild steroid creams, or ointments and dressings.

The baby is at increased risk of premature delivery and may be relatively small for dates (or for "gestational age"). With this in mind, it is important that the obstetrician and dermatologist monitor the pregnancy closely with careful observation of the baby's size and growth, particularly if the mother is taking steroid tablets or if the mother is using large amounts (more than 200 gm, 2 large tubes) of steroid creams or ointments.

It has recently been shown that this reduction in foetal growth is not due to treatment with steroid tablets, but due to disease severity. Therefore, early treatment is essential in such cases.

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Is normal delivery possible?

Yes, and Caesarean-section (C-section) is not recommended for this condition unless there are other indications. Blistering can develop at the site of the scar and may require treatment. Blistering of the vulva and vagina can occur, but only in a minority of cases.

Are the treatments safe for the baby and mother?

Mild- to moderate-strength steroid creams or ointments appear to be safe during pregnancy and can be used, but stronger steroid creams and ointments may cause problems with the growth of the unborn baby so that they may be born small.

With steroid tablets, there is always an increased risk of developing diabetes (raised sugar levels) and hypertension (raised blood pressure). Careful observation of blood pressure and urine checks are therefore essential for the mother at an antenatal clinic, while ultrasound scans can look for changes in the baby's growth. Women who have been taking steroid treatment for a prolonged period may not be able to stop the drugs immediately, and should see a clinician for advice about the best way to reduce treatment.

Can women with Pemphigoid Gestationis still breastfeed?

Yes. Even while taking steroid tablets, women should still be encouraged to breastfeed as only small amounts of steroid get into breast milk. There is some evidence that breastfeeding may make the rash disappear more quickly.

Is any special monitoring required?

Yes, regular follow-up at an antenatal clinic is even more important if the mother is taking oral steroids and blood tests, urine tests, and blood pressure checks along with ultrasound scans are all extremely important to monitor the mother and baby's health. ■