



PEMPHIGOID GESTATIONIS

The aims of this leaflet

This leaflet is designed to tell you more about pemphigoid gestationis. It tells you what the condition is, what causes it, what can be done about it and where to find out more about it.

What is pemphigoid gestationis?

Pemphigoid gestationis is a rare skin blistering disorder that is associated with pregnancy. It usually presents in mid to late pregnancy (between the 13th and 40th week of pregnancy, known as the second and third trimesters) with an itchy rash that develops into blisters. It may recur in subsequent pregnancies and occasionally recurs in women who go on to take oral contraceptive therapy or during menstruation when their periods restart after pregnancy.

It was previously known as herpes gestationis but this name has been changed as there is no association with herpes virus (cold sore) infection and unlike herpes, pemphigoid gestationis does not spread from person to person.

What causes pemphigoid gestationis?

Pemphigoid gestationis is an auto-immune blistering disease. This means that the mother's immune system starts reacting against her own skin causing the skin to split and form blisters. Current knowledge suggests that some very small parts of the placenta tissue enters the mother's blood stream and causes her immune system to activate, producing antibodies which attack the skin.

Female hormones (particularly oestrogen) are thought to aggravate the condition and this may be why the condition presents during pregnancy when

oestrogen levels rise and recurs when oral contraceptives containing oestrogen are taken.

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.

What are the symptoms of pemphigoid gestationis and what does it look like?

Itching is common and the rash often starts around the belly button. It starts with urticarial wheals (like hives from nettles) which are raised, red areas (plaques) commonly occurring on the trunk, back, buttocks and limbs. The face, scalp and mucous membranes (mouth and genital area) are usually not involved. Large tense blisters then occur on the red plaques within 1-2 weeks, and may also occur on palms and soles. The blisters contain clear fluid though occasionally this can be blood-stained. The blisters usually heal without scarring.

How will pemphigoid gestationis be diagnosed?

Usually the appearance and behaviour of the rash is very typical and easily diagnosed by a dermatologist. However, before the blisters appear, the rash can look similar to other skin diseases. Your doctor may take a sample of skin (a biopsy) and a blood test to look for the antibodies that will confirm the diagnosis.

Can pemphigoid gestationis be cured?

No, but it can be controlled with treatment. Symptoms often improve towards the end of pregnancy but 80% of women will experience a flare of the rash at the time of delivery. Symptoms usually resolve days or weeks after giving birth, but in some women the disease can remain active for months and may require continued treatment. Restarting menstrual periods, use of oral contraceptive therapy and further pregnancies can all cause flare-ups of the condition.

How can pemphigoid gestationis be treated?

The primary aim of treatment is to relieve itching, prevent blister formation and treat any secondary infection of the blisters.

Steroid creams can be applied to the affected areas of skin.

Dressings can be applied to weepy or raw areas of skin. Emollient creams or ointments can also be applied to reduce itching and soothe sore areas.

CAUTION: This leaflet mentions 'emollients' (moisturisers). Emollients, creams, lotions and ointments contain oils which can catch fire. When emollient products get in contact with dressings, clothing, bed linen or hair, there is a danger that a naked flame or cigarette smoking could cause these to catch fire. To reduce the fire risk, patients using skincare or haircare products are advised to be very careful near naked flames to reduce the risk of clothing, hair or bedding catching fire. In particular smoking cigarettes should be avoided and being near people who are smoking or using naked flames, especially in bed. Candles may also risk fire. It is advisable to wash clothing daily which is in contact with emollients and bed linen regularly.

Oral antihistamines (those suitable for use during pregnancy) can be used to relieve itching.

Steroid tablets may be needed to control the blisters and itching. The dose of steroid tablets may need to be increased at the end of pregnancy to prevent the disease flaring after delivery.

Occasionally, if symptoms are severe and persist after delivery, in spite of steroid tablets, then alternative medications such as immunosuppressants or concentrated antibodies (IVIg) may be considered. These have been reported to be successful in a few cases, and may be considered by the medical team.

Will the baby be affected?

Sometimes the baby will develop a blistering rash following delivery due to transfer of the mother's antibodies across the placenta. This only occurs in 5-10% of cases. The rash can last up to 6 weeks until the mother's antibodies are cleared from the baby's system. Usually the rash will improve with creams such as steroid or antibiotic creams and emollients.

The baby is at increased risk of premature delivery and may be relatively small as compared to a normal full-term baby. It is important that the obstetrician, and midwives monitor the pregnancy closely.

Is normal delivery possible?

Yes, Caesarean section is not routinely necessary for this condition unless there are other indications. Blistering of the vulva and vagina occurs in 20% of affected women.

Are the treatments safe for the baby and mother?

Taking steroids by mouth means an increased risk in the mother of developing diabetes (raised sugar levels) and hypertension (raised blood pressure). Regular checks of blood pressure, urine and ultrasound scans are important. Women who have been taking steroid treatment for a prolonged period should not stop them suddenly but should follow their doctor's advice and reduce treatment gradually.

Can women with pemphigoid gestationis still breastfeed?

Yes. It is safe to breastfeed even while taking steroid tablets as only very small amounts of steroid get into breast milk.

Where can I get more information about pemphigoid gestationis?

Patient support groups providing information:

PEM Friends

www.pemfriends.org.uk

Weblinks to other relevant sources:

www.dermnetnz.org/immune/pemphigoid-gestationis.html (includes photographs)

Jargon Buster: www.skinhealthinfo.org.uk/support-resources/jargon-buster/

Please note that the British Association of Dermatologists (BAD) provides web links to additional resources to help people access a range of information about their treatment or skin condition. The views expressed in these external resources may not be shared by the BAD or its members. The BAD has no control of and does not endorse the content of external links.

This leaflet aims to provide accurate information about the subject and is a consensus of the views held by representatives of the British Association of Dermatologists: individual patient circumstances may differ, which might alter both the advice and course of therapy given to you by your doctor.

This leaflet has been assessed for readability by the British Association of Dermatologists' Patient Information Lay Review Panel

BRITISH ASSOCIATION OF DERMATOLOGISTS

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