



PEMPHIGOID GESTATIONIS

What are 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 auto-immune skin blistering disorder that occurs in pregnant women after 13 weeks gestation. It appears with an itchy rash that develops into blisters. It usually recurs in subsequent pregnancies and once triggered tends to occur in women who go on to take oral contraceptive therapy, or rarely during menstruation when periods restart following pregnancy.

It was previously known as herpes gestationis, but this name has been abandoned as there is no association with herpes (cold sore) virus infection.

What causes pemphigoid gestationis?

As noted, 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 of the placenta tissue enters the mother's blood stream and causes her immune system to become activated and cross react with her skin and attacks it causing the blisters.

Female hormones (particularly oestrogen) are thought to aggravate the reaction; this may be why it often presents during pregnancy, when oestrogen levels rise, and recurs when oral contraceptives containing oestrogen are taken and during the periods later on.

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?

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

What does pemphigoid gestationis look like?

There is often an urticarial rash with wheals (like hives from nettles) and large raised red patches (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 patches within 1 to 2 weeks, and are also seen 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 trained dermatologist. However, in cases presenting early before the blisters appear, the rash can look similar to other skin diseases. Your dermatologist may take a sample of skin, and a blood sample for immunofluorescence tests, to detect the antibodies that will confirm the diagnosis.

Can pemphigoid gestationis be cured?

No, but it can be suppressed with treatment. Symptoms often improve towards the end of pregnancy but then 80% of 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 or years and may require continued treatment. Restarting periods, use of oral contraception and further pregnancies can all cause flare-ups of the disease.

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.

Topical steroid creams can be used in mild cases if only a limited area of skin is affected. Even if the rash is quite extensive using a strong steroid cream may be worthwhile before steroid tablets are given.

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

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 the obstetricians and paediatricians as well as the dermatologists, to look after the health of both mother and baby. The dose of steroid tablets may need to be increased at the end of pregnancy to prevent the disease flaring after delivery.

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. Emollient creams or ointments can also be applied to reduce itching and soothe sore areas.

Other drugs may be used in more severe cases.

Will the baby be affected?

Occasionally 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% to 10% of cases with the rash lasting up to 6 weeks until the mother's antibodies are cleared. Usually only mild treatment is required such as antibiotic creams and dressings.

The baby is at increased risk of premature delivery and may be relatively small for dates. With this in mind it is important that the obstetrician, paediatricians and dermatologist monitor the pregnancy closely with careful observation of the baby's size and growth, particularly if the mother is taking steroid tablets.

Is normal delivery possible?

Yes, Caesarean section is not routinely recommended for this condition unless there are other indications. Blistering of the vulva and vagina can occur but only in about 20% of cases.

Are the treatments safe for the baby and mother?

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

Can women with pemphigoid gestationis still breastfeed?

Yes. It is safe to breastfeed even while taking steroid tablets as only negligible amounts of steroid get into breast milk. There is some evidence that breastfeeding may improve the disease activity.

Is any special monitoring required?

Yes, regular review at an antenatal clinic is even more important if the mother is taking steroid tablets orally and blood tests, urine tests, blood pressure checks with ultrasound scans are all extremely important to monitor the mother and baby's wellbeing.

Where can I get more information about pemphigoid gestationis?

Link to patient support group:

<http://health.groups.yahoo.com/group/PemGest/> (requires registration/log in)

Web links to detailed leaflets:

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

<http://emedicine.medscape.com/article/1063499-overview> (includes photographs)

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: its contents, however, may occasionally differ from the advice given to you by your doctor.

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

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