PEMPHIGOID GESTATIONIS

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Pemphigoid gestationis (PG) is a skin condition that usually presents in the second trimester of pregnancy; however, it may occur at any time during the pregnancy, as well as in the days post-partum. PG was formerly called herpes gestationis, but it not related to the herpes virus. PG is caused by the production of autoantibodies to proteins found in the skin. It is not surprising that this condition occurs more frequently in those with other autoimmune diseases such as Graves' disease. Other risk factors include older age, Caucasian race, and multiple pregnancies. PG occurs in approximately 1 in 50,000 pregnancies in the United States.

PG presents as a red, itchy rash that forms plaques and papules around, and often including, the belly-button. The lesions resemble hives. Within days to weeks the rash may spread to the extremities, chest, back, buttocks, palms and soles. The rash usually spares the face and mucosal surfaces. It is often intensely pruritic. After a couple of weeks, blisters, also called vesicles, may form within the plaques. The fluid filled vesicles are often arranged in a circular configuration. Some of the lesions may look like targets. Ruptured vesicles do not leave scars, although scarring may occur secondary to scratching or bacterial infections. The disease progression is variable, but it often flares shortly after delivery, and then spontaneously resolves within 3 months.

For unknown reasons, the body creates antibodies to proteins in the skin that are integral parts of a structure called the hemidesmosome. The hemidesmosome attaches the outer layers of the skin, the epidermis, to the deeper layer of the skin called the dermis. The deposition of antibodies at the junction of the dermis and epidermis causes inflammation. The inflammation results in redness, swelling, and fluid accumulation within the skin. As the fluid builds-up, it separates the layers of the skin and forms a blister.

PG is diagnosed by taking a **biopsy** of the skin and a pathologist examines the tissue under a microscope. However, the biopsy alone is not enough to distinguish PG from another condition called **pruritic urticarial papules and plaques of pregnancy** (PUPPP). PUPPP can look similar clinically and histologically. Thus a test called direct immunofluorescence is used in conjunction to detect antibodies deposited in the skin. Additionally, your doctor may use a test called indirect immunofluorescence to detect specific antibodies circulating in the blood stream. Because PG is more common in those with Grave's disease, your physician may order tests to assess your thyroid function as well.

Although PG and PUPP may have similar symptoms, it is important to differentiate between the two. PG may recur with subsequent pregnancies, menstruation cycles, or with the use of oral contraceptives. This suggests a hormonal role in disease manifestation. Rarely, cases of persistent disease have been reported. In less than 5% of cases, circulating maternal IgG antibodies cross over the placenta resulting in transient urticarial lesions or blisters in the newborn. Most cases are mild and do not require treatment. Severe cases are rare, and can increase the risk for infection, fluid loss, and electrolyte abnormalities. PG is associated with premature delivery; however, data does not suggest that there is an increased risk for death of the baby. Additionally, newborns may be small for gestational age. Other differential diagnoses include epidermolysis bullosa acquisita, acute **urticaria**, bullous **scabies**, **allergic contact dermatitis**, **drug eruption**, and **erythema multiforme**.

Mild cases are treated with **topical steroids**. Moderate to severe cases are treated with **oral steroids**. Steroids are tapered to the lowest effective dose and given on alternate days once the blisters stop appearing. First generation **antihistamines**, such as diphenhydramine, can be used for itching. Pyridoxine (B6) is reported to be effective in some patients. Other treatments used after delivery in severe cases include IVIG, plasmapheresis, rituximab, **dapsone**, **cyclophosphamide**, **methotrexate**, and **tetracyclines** with nicotinamide. Secondary bacterial infections are a potential complication and should be treated with antibiotics.

This information has been provided to you compliments of the American Osteopathic College of Dermatology and your physician.