The Treatment

- **GLUTEN-FREE DIET**
- Sulfa-based medications, Dapsone or Sulfapyridine
- (G-6-PD enzyme level before treatment)
- Response to treatment is slow

The Treatment

The mainstay of treatment for dermatitis herpetiformis is the same as for celiac disease, and, that is, the avoidance of ingesting gluten. The gluten-free diet is a particularly difficult diet because wheat, barley and rye are so common in the Western diet. Avoidance of all gluten is important to control the disease and prevent skin lesions from forming and to prevent the possible development of lymphoma. In addition to avoiding dietary gluten, medications may be used to help treat dermatitis herpetiformis. Two common medications are Dapsone and Sulfapyridine, both sulfa-based drugs. Prior to starting Dapsone, individuals with dermatitis herpetiformis and celiac disease should be tested for an enzyme, glucose 6 phosphodiesterase or G-6-PD. If this enzyme is not present or low, Dapsone will not be used or used very cautiously in low doses. Dapsone has effects on the blood which are amplified when the enzyme is not present. The initial Dapsone dosage will be small and will be gradually increased. The effects on blood must be monitored and the dosage adjusted accordingly. In many people with dermatitis herpetiformis, Dapsone is the most effective drug to supplement treatment by a gluten-free diet. Sulfapyridine may also be used to treat dermatitis herpetiformis. This drug has a tendency to cause kidney stones so it is important to drink plenty of fluids if taking it. It may take a longer time for gluten free diet treatment to be effective. Generally, the longer a patient has had the disease, either untreated or poorly treated, the longer it will take to get dermatitis herpetiformis under control. Gluten avoidance is necessary for the rest of a person's life.

Testing for Dermatitus Herpetiformis may be arranged through the Immunodermatology Laboratory,University of Utah. Specimen kits with paid return postage are available for individuals to take to their medical care providers or, preferably, kits can be sent directly to medical care providers.

Email: http://uuhsc.utah.edu/derm/immunoderm/
or
Phone: (866) 266-5699

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DERMATITIS HERPETIFORMIS

The Disease

Dermatitis herpetiformis is a skin manifestation of celiac disease. The skin lesions of dermatitis herpetiformis are intensely itchy. They typically begin as groups of red bumps, often with tiny blisters on the surface of the bumps, and commonly arise on skin around elbows, knees, buttocks, back of the neck and scalp. The location of the lesions on these skin sites is important for the diagnosis of dermatitis herpetiformis (although skin lesions can occur on almost any part of the skin including face, trunk and other parts of arms and legs). Because itching is so severe, people with dermatitis herpetiformis cannot help but scratch their skin. Blisters may be scratched open and then crust over. Bruising may be found. Also, with chronic scratching, skin becomes thickened. Therefore, the effects of scratching can dominate and be all that is observed. However, scarring rarely or never occurs, and, once dermatitis herpetiformis is controlled, the skin heals and appears normal.

"Dermatitis herpetiformis" is a descriptive name for the disease because "dermatitis" means inflammation of skin as manifested by redness and "herpetiformis" means similar to herpes which shows clusters of lesions. The disease is not related to herpes virus infection at all. An American dermatologist, Louis Duhring, first described the disease in 1884, and, in the past, it was called Duhring's Disease. The association with celiac disease was not recognized for many years; in fact, it was nearly a century after dermatitis

The Disease

- **VERY ITCHY**
- Skin lesions are common on elbows, knees, buttocks, neck and scalp
- Associated with intestinal celiac disease (gluten sensitivity)
- May occur with other auto-immune diseases
- Increased risk of lymphoma, a type of cancer in untreated disease

herpetiformis was described that the link was made in 1967. Individuals with celiac disease can have other skin diseases as well, so that not all skin disease in people with celiac disease is dermatitis herpetiformis. Acne, psoriasis and rosacea are examples of common skin diseases that may occur along with dermatitis herpetiformis or alone in people with celiac disease. Dermatitis herpetiformis can occur at any age. The average age of onset is 28 years old. It is usually not found in children. When it does occur before age 20, there is a slight female predominance. After age 20, it is more common in men than women.

Dermatitis herpetiformis is an inherited disease and is most common in people of northern European descent. It is associated with certain genetic markers of immune expression. These same markers occur in celiac disease. Virtually everyone with dermatitis herpetiformis has at least mild celiac disease. For unknown reasons, intestinal malabsorption in celiac disease is usually mild in patients who have dermatitis herpetiformis. Very recent investigative studies are beginning to provide information on why only some people with celiac disease get dermatitis herpetiformis.

Several other diseases may also be seen in people with dermatitis herpetiformis. These diseases are linked to the same genetically determined immunologic expression as dermatitis herpetiformis and celiac disease, but not so strongly or directly. They include: thyroid dysfunction, pernicious anemia, diabetes mellitus, lupus erythematosus, rheumatoid arthritis, and some liver diseases. These diseases are all characterized by abnormal production of antibodies to one's self (autoantibodies). A very serious association with dermatitis herpetiformis and celiac disease is an increased risk of developing lymphoma, a cancer of the lymph system in the body. As is the case in celiac disease without dermatitis herpetiformis, the risk can be significantly lessened if individuals follow a gluten-free diet.

The Diagnosis

The diagnosis of dermatitis herpetiformis may be delayed because skin lesions can look like other skin problems and because health care providers do not think of the disease and perform the proper testing. The diagnosis is made by skin biopsy and blood testing. When the proper skin site is biopsied in individuals with the disease, a special test, called direct immunofluorescence, is very accurate. The skin biopsy should be obtained near but not directly from a lesion or cluster of lesions.

The Diagnosis

- **SKIN BIOPSY FOR DIRECT IMMUNO-FLUORESCENCE** (if you think that you have a skin disease related to celiac disease, a skin biopsy for a special test is "crucial" see how to arrange testing below)
- Blood studies will help confirm the diagnosis and monitor response to treatment
- Intestinal biopsies may be recommended for associated celiac disease

It may be helpful to have more than one biopsy taken because the findings of dermatitis herpetiformis can be subtle and sparse. The actual procedure to obtain the biopsy is not uncomfortable because a local anesthetic is used to numb the site. A small scar will be present after healing where the biopsy is taken. It is very important that the biopsy specimen be sent in special fixative to a laboratory with specialists, Immunodermatologists, who know how to prepare the direct immunofluorescence test and interpret it. The typical findings are granular clumps of an antibody, called IgA, in certain parts of the skin, but, as already mentioned, the findings can be very subtle, so having an experienced Immunodermatologist analyze the specimen is important. Biopsy of an active lesion preserved in formaldehyde for regular microscopic studies is useful for revealing other findings characteristic of dermatitis herpetiformis.

Blood testing can be helpful to verify the results of skin biopsy testing and to monitor response to therapy. Blood tests detect IgA antibodies to two substances, smooth muscle endomysium and tissue transglutaminase, that are involved in a very complicated process to cause celiac disease. Blood testing may also be performed to detect IgA antibodies to gliadin, the main protein in wheat, which initiates the reaction. IgG antibody testing may be performed if an individual is deficient in IgA. Antibody levels to these factors decrease when dermatitis herpetiformis and celiac disease are treated. Therefore, these blood tests may be used to monitor the response to treatment as well as help to establish the diagnosis. Finally, differences in antibody responses may determine why an individual gets dermatitis herpetiformis. Therefore, in the near future additional antibody testing in blood may be helpful in determining or predicting who will develop skin disease along with celiac disease. Blood testing will likely help predict relatives of disease sufferers who are genetically susceptible to developing disease, too.