What Is Dermatitis Herpetiformis ?

http://www.dermatitisherpetiformis.org.uk/whatisdh.html

The following article, written by Lionel Fry, Emeritus Professor of Dermatology, first appeared in the Summer 2001 Edition of the Crossed Grain, the official magazine of Coeliac UK, and is reproduced here with their kind permission.

Described by one sufferer as "...like rolling in stinging nettles naked with a severe sunburn, then wrapping yourself in a wool blanket filled with ants and fleas...."

Dermatitis herpetiformis (DH) was first described as a distinct clinical entity in 1884 by an American dermatologist, Louis Duhring. The name was descriptive, dermatitis being inflammation of the skin and herpetiformis meaning group. The typical features are small grouped itchy blisters, often on red plaques, situated on the back of the elbows and forearms, buttocks and front of the knees. Although these are the common sites the rash may in addition, occur anywhere on the body including face, scalp and trunk. The eruption is extremely itchy and may keep patients awake at night.

DH may be present at any age but most commonly begins between the ages of 15-40. It is slightly more common in men than females at a ratio of 3:2. DH is a persistent condition and only approximately 10% of patients have a spontaneous permanent remission with no medical treatment.

Role of gluten and association to celiac disease

The first suggestion that patients with DH also have an enteropathy identical to coeliac disease (CD) was made in 1967. This was confirmed by showing the enteropathy cleared with gluten withdrawal from the diet and recurred when gluten was reintroduced. It was subsequently shown that all patients with DH have evidence of a gluten enteropathy. However, in the majority of patients the enteropathy is mild and does not give rise to symptoms such as abdominal pain, weight loss and diarrhoea. Thus, all patients with DH have associated CD although it could be described as latent CD in the majority.

Diagnosis

The diagnosis of DH is made by a simple skin test. A small piece of skin approximately 3 mms in diameter is taken from an unaffected area, ie. normal looking skin. The skin is examined for the presence of a substance called IgA (immunoglobulin A) and is found at a specific site in the skin. Although the test is simple, it is important a laboratory experienced in the procedure undertakes the examination of the skin.

- The blister is subepidermal (it forms underneath the epidermis)
- The inflammatory cells (neutrophils and eosinophils) group in the dermal papillae
- Direct immunofluorescence reveals IgA immunoglobulin in dermal papillae

The diagnosis of DH can also be confirmed with the same tests as used for diagnosing CD, ie. a small intestinal biopsy and blood tests looking for specific antibodies, called anti-endomysial (EMA) and tissue transglutaminase antibodies (tTg. Occasionally in DH, the blood tests may be negative because their positivity correlates strongly with the severity of the intestinal lesion.

Management of DH

Until the discovery of the association of DH with CD the treatment of DH was solely with drugs. Now there is a more satisfactory and less hazardous treatment with a gluten free diet; although, it must be appreciated that drugs will control the rash within days. The gluten free diet will take longer but since it is addressing the underlying cause of the condition, will be a more permanent cure. Initially, both drugs and a gluten free diet are suggested to maximize comfort of the patient.

DRUGS - The drugs more commonly used for the treatment of DH are Dapsone, sulphapyridine, and sulphamethoxypyridazine. All are very early antibiotics dating back to the 1930's and 1940's. However, these drugs do not work as antibiotics but their exact mechanism of action on DH is unknown.

Although the drugs will control the rash of DH very quickly, ie. within days, the rash will recur equally quickly when drugs are discontinued. Thus, drug treatment of DH has to be considered indefinite but it is not a cure for the disease. It is important to find the smallest dose required to control the rash and this varies between patients. Thus, the dose may be increased or decreased after initial treatment.

Unfortunately all three drugs used to control the rash in DH may have side effects.

Dapsone has the highest incidence of side effects. 25% patients will experience an adverse reaction. The commonest reaction is so-called haemolytic anaemia. Others are neuropathy (damage to nerves), depression, headache and (rarely) damage to the liver and bone marrow.

It is important that patients taking drugs for DH are carefully monitored with frequent blood tests at the beginning of treatment. Since side-effects tend to occur early in treatment, patients may only have to attend hospital every six months once established on drug treatment.

GLUTEN FREE DIET - It is important to appreciate that a gluten free diet may have no effect on the rash for approximately six months and sometimes, even longer. It takes this length of time before patients can start to reduce their drug requirements and approximately 2 years before they can discontinue drugs completely. It is also important to realize that these times are only achieved if the diet is *absolutely* strict. Even small amounts of gluten may result in a reoccurrence of DH rash.

Thus, because the drugs control the rash quickly and the gluten free diet does not, it is normal practice to start both drugs and diet together. After six months, the dose of drugs can be slowly reduced. DH is a persistent disorder (and because these patients also have CD, even if mild), the diet must be considered to be life-long. However, one improvement for patients that has occurred in the last five years is that it has been shown in DH (as in CD) that oats do not cause the rash and thus, these can be taken. Wheat, barley and rye must still be omitted. There is no evidence that gluten in flour or wheat products touching the skin can induce or exacerbate DH or CD.

A strict gluten-free diet is strongly recommended.

- It reduces the requirement for dapsone
- It improves associated gluten enteropathy
- It enhances nutrition and bone density
- It may reduce the risk of developing other autoimmune conditions
- It probably reduces the risk of intestinal lymphoma.

Associated disorders

Although, DH is not itself a so-called autoimmune disease, there is an increased incidence of the latter in patients with DH. The three most commonly associated with DH are thyroid disease, pernicious anaemia and diabetes. Thus, patients with DH should be screened for these diseases on a yearly basis. There are other less common autoimmune disorders, which are also increased in DH and these have to be borne in mind during follow-up. There is some evidence that a strict gluten-free diet reduces the risk of developing autoimmune diseases.

Lymphoma (a type of malignancy) also has an increased incidence in DH as in CD. The incidence is approximately 2% but it has been shown that the risk of developing this disorder disappears with a strict gluten-free diet.

Genetics

There is evidence that both DH and CD have a genetic basis. It appears that the genes for the two diseases are fundamentally the same because pairs of identical twins have been described in which one twin has DH and the other only CD. In addition, in large families the relatives of a patient with DH have equal numbers of those affected with DH as with CD. At present, the genes for DH and CD have not been found but there are certain genetic markers for the disease that are known, which appear to increase the susceptibility to the diseases.

The risk of patients with DH having children with the disease is relatively low. In a study of over 1,000 patients with DH in Finland, only 10% of the patients had a family history of the disease and only 14% of these had a child with the disease. However, there are now tests for screening for CD, which is the prerequisite for DH and thus, relatives can be screened if so desired.

The future

It can be said that both patients with DH and CD can now be cured with a treatment, a gluten free diet, which does not require drugs, which is a remarkable achievement compared to many other diseases. However, research will continue into how gluten causes both CD and DH and new treatments may emerge which allow patients to eat a normal diet.

We very much appreciate Professor Fry writing this article. As a footnote, he added that people with Celiac Disease do significantly reduce their risk of contracting DH if they stick to a gluten-free diet.

Dermatitis Herpetiformis from http://www.nutramed.com/skin/herpetiformis.htm

Dermatitis Herpetiformis (DH), a skin disease, is another manifestation of gluten or wheat allergy. It is characterized by urticarial plaques and blisters on the elbows, buttocks, and knees, although other sites may also be involved. The eruption tends to be persistent: only 10-15% of patients have spontaneous remission over a 25-year study period. The disease is characterized by the presence of IgA deposits in the upper dermis. The HLA-B8 gene is found in 80% of patients with gluten enteropathy and dermatitis herpetiformis.

In DH, clusters of itchy skin bumps, and small blisters appear on the legs, sacrum, buttocks and back - the extensor surfaces of the body. Intense, "dives-me-crazy" itching is typical. As the lesions mature, the skin tends to be red, thickened, bumpy and scratching adds injury to insult. The skin inflammation is typical of a delayed, cell-mediated hypersensitivity reaction. Gluten proteins from ingested wheat and other grains find their way to the afflicted skin and trigger an immune attack.

The lesions of DH are visible models of how food antigens can bring an inflammatory attack into an organ system and wreck havoc. In the skin, the inflammation is uncomfortable, but in a more critical organ, the inflammation can have disastrous effects. DH clears completely when all gluten is eliminated from the diet, although healing usually takes several weeks to occur. Although people with DH and celiac disease often come to their diagnosis by different routes and may not recognize their kinship, both groups are allergic to gluten and have similar risks of developing any or many of the associated diseases. Patients with DH have a high incidence of auto-immune disorders, thyroid disease, pernicious anemia, and insulin-dependent diabetes. As with celiac disease, there is an increased incidence of lymphoma and a gluten-free diet appears to protect patients.

The drugs, Dapsone, sulphapyridine or sulphamethoxypyridazine have been used to suppress the skin manifestation. It takes several months for the skin to improve on these drugs and they do not protect against the serious complication of gluten hypersensitivity. The drugs also have numerous side effects and should not be necessary if treatment includes a strict GF diet..

The person with DH may not realize that they have major disturbance in their digestive tract but studies have shown similar if not identical disease processes. T-lymphocytes in the small intestinal mucosa of celiac disease and dermatitis herpetiformis subjects on a normal diet has been studied and compared to normal controls. The small intestine is a site of vigorous T-cell activity in gluten-sensitive individuals and is consistent with the view that the enteropathy of dermatitis herpetiformis and celiac disease is the result of a delayed-type hypersensitivity reactions triggered by wheat proteins.

Additional Websites for further research

http://www.dermnetnz.org/immune/dermatitis-herpetiformis.html

http://www.gluten.net (National Gluten Intolerance Group website)

http://www.glutenfreeway.info (local, Whatcom County Gluten Free support group)

http://digestive.niddk.nih.gov/ddiseases/pubs/celiac/ (National Clearinghouse for Digestive Diseases)

http://www.celiac.com (type Dermatitis Herpetiformis into search window)

http://emedicine.medscape.com/article/1062640-overview - GREAT Article (would be good to print and present to physician)