DermNet NZ

Authoritative facts about the skin from the New Zealand Dermatological Society Incorporated.

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Dermatitis herpetiformis

Dermatitis herpetiformis (also known as 'DH') is a rare but persistent immunobullous skin condition related to coeliac disease. It affects young adults; two thirds of patients are male. There is a genetic predisposition.

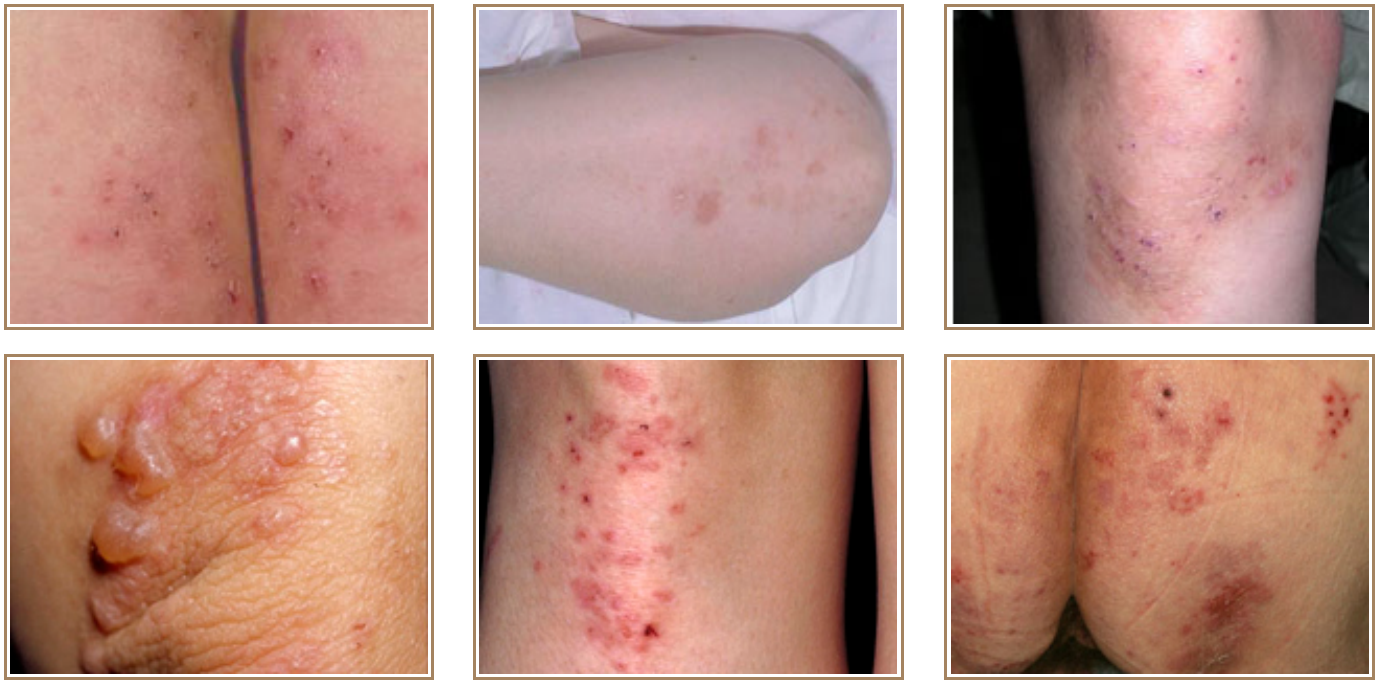
'Immunobullous' means it is a blistering condition caused by an abnormal immunological reaction. All forms of coeliac disease involve IgA antibodies and intolerance to the gliaden fraction of gluten found in wheat; the precise reaction has not been identified.

Eighty percent of patients with dermatitis herpetiformis also have gluten enteropathy, which is the most common type of coeliac disease. There is an association with thyroid disease in one third.

Clinical features

Dermatitis herpetiformis characteristically affects the scalp, buttocks, elbows and knees but lesions may arise on any area of skin. Extremely itchy bumps (papules) and blisters (vesicles) up to 1 cm in diameter arise on normal or reddened skin. The severity can vary from week to week but it rarely clears up without specific treatment.

Dermatitis herpetiformis



Gluten enteropathy

Gluten enteropathy may affect children and adults. It is characterised by villous atrophy. This means that instead of being highly convoluted, the lining of the intestines is smooth and flattened. The result is poor or very poor absorption of nutrients. The patient may feel well or develop the following symptoms:

- Tiredness (80%)

- Abdominal discomfort and bloating (75%)
- Weight loss (30%)
- Constipation (30%) or diarrhoea (50%)
- Pale stools that float on the surface of the toilet pan
- Bone fractures due to osteoporosis

Other associated conditions

The range of conditions less commonly induced by gluten also includes:

- Neurological problems including ataxia (loss of balance), polyneuropathy, epilepsy
- Heart problems including pericarditis and cardiomyopathy
- Thin dental enamel
- Recurrent abortions (miscarriage)
- Fatty liver resulting in abnormal liver function
- Aphthous ulcers

Patients with coeliac disease sometimes suffer from other autoimmune conditions possibly associated with gluten intolerance. These include insulin-dependent diabetes mellitus, thyroiditis, autoimmune hepatitis, Sjögren's syndrome, Addison's disease, atrophic gastritis and alopecia areata.

They may also be affected by conditions that are not related to gluten intolerance. These include IgA deficiency, psoriasis, Down syndrome and primary biliary cirrhosis.

Non-Hodgkin´s lymphoma, affecting the intestines or any part of the body, is a serious complication of gluten enteropathy but is fortunately rare, affecting less than 1% of patients.

Laboratory findings

Although dermatologists may suspect the diagnosis from the clinical appearance, a skin biopsy is usually necessary to confirm it. The microscopic appearance of dermatitis herpetiformis is characteristic.

- 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 results of blood tests are usually normal but some patients have the following abnormalities, due to gluten enteropathy:

- Mild anaemia
- Folic acid deficiency
- Iron deficiency

Specific autoantibody tests are available to confirm the diagnosis.

- Positive transglutamidase screening test (IgA and IgG)
- IgA antigliadin antibodies
- Antiendomysial antibodies

Other tests may include:

- Total IgA
- Histocompatibility antigen typing: HLA-DR3 and DQw2 are present in most patients with coeliac disease. About 5% of those with HLA-DQ are affected by one form or other of coeliac disease
- Small bowel biopsy

The bowel may appear normal because of treatment (medicine or restricted intake of gluten), because there are skip lesions (the sample was taken from an unaffected site) or the intestine may be unaffected by the disease.

Treatment

The medication of choice is dapsone, which considerably reduces the itch within a day or two. The dose required varies from 50 mg to 300 mg daily; refer to DermNet's page about dapsone for potential side effects and monitoring requirements.

For those intolerant or allergic to dapsone, the following may be useful:

- Ultrapotent topical steroids
- Systemic steroids
- Sulfapyridine (not available in New Zealand).

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.

Related information

Other websites:

- Manufactured Food Database (NZ) for gluten free diet
- NZ Coeliac Society
- Gluten Intolerance Group of North America
- Dermatitis herpetiformis - emedicine dermatology, the online textbook

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DermNet does not provide an on-line consultation service.
If you have any concerns with your skin or its treatment, see a dermatologist for advice.