Dermatitis herpetiformis

FACT SHEET

What is dermatitis herpetiformis?

Dermatitis herpetiformis (DH) is a chronic skin condition associated with coeliac disease. It is intensely itchy, even when only appearing as a mild rash. The name, dermatitis herpetiformis, is a descriptive name. The rash is not related to either dermatitis or herpes, but is a specific chronic skin condition.

DH has a genetic basis and is not contagious. The rash may be small lumps, like insect bites, some with tiny fluid filled blisters on top, called vesicles. However, it can also appear hive-like, persisting in one area, or it may look like a pink and scaly dermatitis.

DH can flare and subside even without treatment. The rash has a characteristic distribution, over the knee caps, on the outer surface of the elbows, on the buttock area, around the ears, the shoulder blades and in the hairline and eyebrows. It tends to appear symmetrically on both the left and right sides of the body. When the rash subsides, which often occurs spontaneously, it may leave brown pigmentation or pale areas where pigmentation is lost.

Who gets dermatitis herpetiformis?

People are born with a genetic predisposition to develop coeliac disease and/or DH. DH is slightly more common in males than females and generally presents in adult life (15-55 years). It is uncommon to see DH in children, but it can occur.

Less than 10% of patients with DH will also have the gastrointestinal symptoms associated with coeliac disease. However, biopsies show that people with DH will have some degree of villous atrophy (small bowel damage) on consumption of a high gluten load. There is a small group of patients with coeliac disease who later develop DH even though they have been on a strict gluten free diet for years.

Like coeliac disease, it is unclear why the condition develops at a particular time. Both genetic and environmental factors play important roles in DH.

Does dermatitis herpetiformis occur in families?

About 10% of patients with DH have first degree relatives with either DH or coeliac disease. There is the possibility that atypical or silent coeliac disease could be undiagnosed in the families of patients with DH.

How is the condition diagnosed?

The variable presentation of the rash can make diagnosis very difficult, and a referral to a dermatologist is required. When your doctor suspects DH, a skin biopsy for histopathology and immunofluorescence is performed. This involves taking a small piece of skin, which is sent to a pathologist for testing.
The sample of skin will often include a small papule and blister and some normal surrounding skin. This is done prior to commencing treatment. With DH the immunofluorescence test will show deposits of IgA

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in the skin. Whilst IgA can be found in the skin in other skin diseases, a particular pattern in the IgA immunofluorescence is seen in DH. A positive immunofluorescence test can be obtained in many patients with DH even when the skin disease is in remission and the rash is not visible. There are a number of abnormal serum (blood) antibodies that occur in DH, just as they do in coeliac disease.

How is the condition treated?

DH is treated in the same way as coeliac disease, by maintaining a strict gluten free diet for life. It may take six months to achieve moderate improvement in the skin condition and up to two years or more to achieve total control by diet alone.

An oral medication called Dapsone is available to relieve the rash of DH. Within twenty four hours of taking this medication, the itch is often better and the rash may disappear within a few days. Dapsone, however, does not improve the small bowel damage. Dapsone is generally well tolerated but requires careful medical supervision. It has a number of side effects, especially if taken in large amounts for long periods of time, e.g. haemolysis resulting in anaemia and chronic tiredness. Sulphapyridine and other sulphonamides are also used. Maintaining a strict gluten free diet is essential to reducing and eliminating the use of medication. Even occasional unintentional gluten intake reduces successful elimination of Dapsone for many patients.

Where compliance with the gluten free diet is strict, the average time for reduction in the use of Dapsone has been shown in one study to be less than a year and can generally be stopped within about two years.

What are the implications of dermatitis herpetiformis?

The implications of DH are the same as for coeliac disease.

- Before diagnosis, both DH and coeliac patients may suffer from nutritional deficiency caused by malabsorption, including anaemia secondary to iron or folate deficiency.
- There is a small, but statistically higher risk of developing lymphoma of the small intestine, particularly when the condition has been untreated for many years.
- Patients with DH, particularly females, have an increased risk of autoimmune thyroid disease. Twenty per cent of patients with DH have clinical hypothyroidism and about 10% have asymptomatic goitres.
- There is also an increased incidence of other autoimmune diseases, like type 1 diabetes mellitus, vitiligo, sjogrens disease, lupus erythematous and sarcoidosis in those with DH.