

CELIAC DISEASE AND DERMATITIS HERPETIFORMIS

Celiac Disease

Celiac disease, also known as gluten-sensitive enteropathy or celiac sprue, is a chronic autoimmune intestinal disorder. When genetically susceptible individuals consume specific proteins in the grains wheat, rye and barley, which are collectively known as “gluten,” the absorptive surface of the small intestine is damaged. This surface contains tiny finger-like projections called villi that become inflamed and flattened (known as villous atrophy) due to the immunologic reaction to gluten, causing malabsorption of nutrients needed for good health. Iron, calcium and folate are key nutrients often affected since they are absorbed in the first part of the small intestine. If the damage progresses further down the small intestinal tract, malabsorption of carbohydrates (especially lactose), fat and fat-soluble vitamins (A, D, E, K), protein and other nutrients may also occur. The development of celiac disease involves a combination of genetic, environmental and immunological factors. Celiac disease can occur at any age, including the elderly, and may be triggered by a gastrointestinal or viral infection, severe stress, surgery or pregnancy.

Continued exposure to gluten can result in vitamin and mineral deficiencies causing conditions such as anemia and osteoporosis; neurological disorders (e.g., ataxia, seizures and neuropathy); and an increased risk for developing other autoimmune disorders (e.g., thyroid disease, type 1 diabetes, connective tissue diseases, Addison’s disease) and certain types of cancer, especially gastrointestinal malignancies. Also, there is an increased risk of miscarriage or having a low-birth-weight baby, and infertility in both women and men. For more information about celiac disease, see the references listed on page 18, nutritional concerns on pages 73-75, 91, 92, 97, 101-103, celiac organizations on pages 343-344 and other resources on pages 345-356.

Prevalence

Originally thought to be rare disorder, celiac disease is now recognized as one of the most common inherited diseases, with a world prevalence estimated at 1:266 people. Recent studies have revealed that celiac disease affects approximately 1% of the U.S. population (1:100 individuals) which is similar to data from European countries.

Symptoms

Celiac disease not only affects the gastrointestinal system but many other systems in the body, resulting in a wide range and severity of symptoms that can vary greatly from one person to another (see page 16). These symptoms may occur singly or in combination in children and adults. Many individuals have “silent celiac disease” (i.e., have no or very subtle symptoms) in spite of gluten sensitivity.

Symptoms of Celiac Disease

- ♦ Iron, folate and/or vitamin B₁₂ deficiency
- ♦ Other vitamin and mineral deficiencies (A, D, E, K, calcium)
- ♦ Chronic fatigue and weakness
- ♦ Abdominal pain, bloating and gas
- ♦ Indigestion/reflux (“heartburn”)
- ♦ Nausea and vomiting
- ♦ Diarrhea, constipation or intermittent diarrhea and/or constipation
- ♦ Lactose intolerance
- ♦ Weight loss (note that CD can also occur in obese individuals)
- ♦ Bone/joint pain
- ♦ Easy bruising of the skin
- ♦ Edema (swelling) of hands and feet
- ♦ Migraine headaches
- ♦ Depression
- ♦ Mouth ulcers (canker sores)
- ♦ Menstrual irregularities
- ♦ Infertility (in both women and men)
- ♦ Recurrent miscarriages
- ♦ Elevated liver enzymes

Additional symptoms in children

- ♦ Irritability and behavioral changes
- ♦ Concentration and learning difficulties
- ♦ Failure to thrive (delayed growth and short stature)
- ♦ Delayed puberty
- ♦ Dental enamel abnormalities

Associated Conditions

Celiac disease can also occur more frequently in a variety of other disorders. Individuals with any of the following conditions and symptoms of celiac disease should be screened for celiac disease:

- ♦ Type 1 diabetes
- ♦ Other autoimmune disorders (e.g., autoimmune thyroid disease, autoimmune liver disease, Sjögren’s syndrome, Addison’s disease, alopecia areata)
- ♦ Osteoporosis
- ♦ Down syndrome
- ♦ Turner Syndrome
- ♦ Selective IgA deficiency

Diagnosis

The diagnosis of celiac disease is often very difficult because of the broad range of symptoms that can vary from mild to severe or none at all. Individuals are often misdiagnosed with irritable bowel syndrome, lactose intolerance, fibromyalgia, chronic fatigue syndrome or ulcers. Recent studies by Columbia University in New York and the Canadian Celiac Association have reported that many people have suffered with symptoms for more than 10 years and have seen numerous physicians before a correct diagnosis of celiac disease was made.

There are specific blood tests, including IgA endomysial (EMA) and IgA tissue transglutaminase (tTG) antibody tests, to detect celiac disease. Unfortunately these tests are not 100% accurate and some individuals test negative in spite of having celiac disease. Therefore, the only definitive test for diagnosing celiac disease is the small intestinal biopsy. **A gluten-free diet should never be started before the blood tests and biopsy are completed as this can interfere with making an accurate diagnosis.**

Treatment

Once a diagnosis for celiac disease is confirmed, it is essential to follow a strict gluten-free diet for life. Additional vitamin and mineral supplements may be necessary to correct the malnutrition. Some individuals may also need to eliminate lactose until the damaged bowel is healed (see pages 101-102).

Dermatitis Herpetiformis

Dermatitis herpetiformis (DH) is another form of celiac disease. This chronic skin condition is characterized by an intense burning, itchy and blistering rash. The rash is symmetrically distributed and commonly found on the elbows, knees and the buttocks, but can also occur on the back of the neck, upper back, scalp and hairline. Initially, groups of small blisters are formed that soon erupt into small erosions. Most people with DH will also have varying degrees of small intestinal villous atrophy although many will have no bowel complaints. A small percentage may present with bloating, abdominal pain and diarrhea, especially if the bowel involvement is severe, and some individuals may show evidence of malabsorption and malnutrition.

Prevalence

Approximately 10% of individuals with celiac disease have DH with a male to female ratio of 2:1. The age of onset is typically between 25-45 but can also occur in children and older adults.

Diagnosis

Individuals with DH are frequently misdiagnosed with other skin conditions such as eczema, contact dermatitis, allergies, hives, herpes or psoriasis and treated with a variety of topical creams. The only way to correctly diagnose DH is a skin biopsy from unaffected skin adjacent to blisters or erosions. A small intestinal biopsy is not essential if the skin biopsy is positive for DH.

Treatment

Treatment for DH is a **strict gluten-free diet for life**. For some individuals, Dapsone, a drug from the “sulphone family,” may be prescribed to reduce the itching. Response to the medication can be dramatic (usually 48-72 hours). However, Dapsone has no effect on the ongoing immune response or intestinal atrophy. Following a strict gluten-free diet will result in:

- ♦ Improvement in the skin lesions.
- ♦ Major reduction in drug dosage for those people initially started on Dapsone. After a time, it is often possible to discontinue the drug to control the skin rash. Flare-ups due to inadvertent or intentional gluten consumption may require temporary use of Dapsone.
- ♦ The gut function will return to normal.

NOTE: Once a diagnosis of celiac disease or dermatitis herpetiformis is confirmed, it is essential to consult with a registered dietitian with expertise in celiac disease and the gluten-free diet for nutritional assessment, diet education, meal planning and assistance with social and emotional adaptation to the new gluten-free lifestyle. Also, joining a celiac support organization for further information and ongoing support is highly recommended.

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