CANADIAN CELIAC ASSOCIATION

Celiac Disease... Hidden and Dangerous

What is Celiac Disease?

Celiac disease (CD) is an immune mediated injury to the small intestine that is caused by ingestion of gluten (a name for multiple proteins in wheat, rye and barley) in genetically susceptible individuals. This can cause a variety of symptoms and result in poor absorption leading to deficiencies of nutrients including fat, protein, carbohydrates, vitamins and minerals such as iron and calcium.

Dermatitis herpetiformis (DH) is "celiac disease of the skin". It is characterized by a blistering, intensely itchy skin rash. The rash is usually symmetrical and is found most frequently on the elbows, knees, buttocks and upper back. Patients with DH often have mild or no gastrointestinal symptoms, but villous atrophy is present in the majority of cases.

Pathogenesis

The pathogenesis of CD involves three factors: genetic, environmental and immunologic. Almost all individuals with celiac disease have the HLA-DQ2 and/or HLA-DQ8 genetic markers. Gluten is the trigger for the immunologic response of CD. Pregnancy, surgery, gastrointestinal infection or severe emotional stress sometimes triggers the disease in genetically predisposed individuals. Celiac disease is an inherited condition and, therefore, first-degree and to a lesser extent second-degree relatives are at higher risk of developing the disorder.

Prevalence

Celiac disease affects about 1% of the population, making it one of the most common chronic gastrointestinal disorders.

Symptoms

Celiac disease can manifest at any age once foods containing wheat, barley or rye are introduced in the diet. Symptoms of classical CD include diarrhea, abdominal pain/distension and weight loss. Patients can be severely malnourished. Some patients present with non-intestinal symptoms, others may have an associated condition with or without celiac-related symptoms. Iron or folate deficiency anemia can occur due to malabsorption. Children may present with short stature, delayed puberty or dental enamel defects. Many symptoms (e.g., anemia, weight loss, bone pain, paresthesia, edema, skin changes) are secondary to nutrient deficiency states. The number and severity of symptoms can vary greatly from person to person. In many cases, the disease is sub-clinical (silent) and is discovered only by blood screening. The presence of obesity does not exclude the diagnosis of CD.

Symptoms of CD may include one or more of the following:

Classical Symptoms

- ☐ Abdominal distension
- ☐ Abdominal pain
- ☐ Chronic diarrhea
- ☐ Loss of appetite
- ☐ Irritability
- ☐ Weight loss (or failure to thrive in children)
- ☐ Muscle wasting
- ☐ Dermatitis herpetiformis (DH)

Associated Conditions (% affected)

- ☐ Relative of individual with CD (8-15%)
- ☐ Type 1 diabetes mellitus (4-8%)
- ☐ Autoimmune thyroiditis (2-5%)
- ☐ Trisomy-21 (Down syndrome) (2-5%)
- ☐ Turner syndrome (2-5%)
- ☐ IgA deficiency (2-5%, up to 30% in patients with gastrointestinal symptoms)

Non-classical Symptoms and Signs

- ☐ Unexplained iron or folate deficiency anemia
- ☐ Aphthous stomatitis (oral canker sores)
- ☐ Dental enamel defects
- ☐ Persistent/recurrent vomiting
- ☐ Irritable bowel syndrome
- ☐ Chronic constipation
- ☐ Elevated liver enzymes (ALT/AST)
- ☐ Arthritis, arthralgia
- ☐ Osteoporosis/Osteopenia
- ☐ Short stature
- ☐ Delayed puberty
- ☐ Infertility

Neurological presentations

- ☐ Unexplained ataxia
- ☐ Peripheral neuropathy
- ☐ Epilepsy with occipital calcifications
- ☐ Depression/anxiety
- ☐ Chronic fatigue

Diagnosis

Studies from Canada and other countries report significant delays in diagnosis of CD. The similarity of the symptoms of CD with those of other diseases often leads to misdiagnoses such as irritable bowel syndrome, lactose intolerance and chronic fatigue syndrome, resulting in delayed diagnosis. Excellent serological blood tests are now available to screen for CD. The IgA tissue transglutaminase antibody (IgA-TTG) or IgA endomysial antibody (IgA-EMA) tests are recommended for initial testing performed by experienced laboratories. The choice of test depends on availability and laboratory preference. Since these tests are IgA based, they will be falsely negative in patients with selective IgA deficiency. The prevalence of IgA deficiency is higher in patients with CD; therefore, screening for selective IgA deficiency should be performed at the same time as the serological tests. In individuals with IgA deficiency, the laboratory may be able to perform IgG-TTG or IgG-deamidated gliadin peptide antibody (IgG-DGP).

The IgA and IgG anti-gliadin antibody (AGA) tests are no longer recommended as screening tests for CD because of poor sensitivity and predictive values.

The diagnosis of CD MUST be confirmed with endoscopic small intestinal biopsy while consuming a regular GLUTEN containing diet. It is strongly recommended that the biopsy be done BEFORE starting a gluten-free diet in order to avoid a false negative biopsy result. The diagnosis of DH can be confirmed with a skin biopsy.

Over 99% of patients with CD are positive for HLA-DQ2 or DQ8 genes. A negative HLA-DQ2 or DQ8test is helpful to exclude the diagnosis of CD. However, approximately 30% of the general population has one of these HLA types and most do not develop CD. Therefore, presence of these genes does not confirm CD.

Treatment

The treatment of celiac disease and dermatitis herpetiformisis a STRICT GLUTEN-FREE DIET FOR LIFE. Patients with DH may also require treatment with medications (e.g. dapsone). A gluten-free diet enables the intestine to recover with resolution of symptoms and can also reduce the risk of developing many of the complications of untreated CD.

Because of the complexity of the diet, patients should be referred to a qualified dietitian with expertise in gluten-free diet for nutrition assessment, education and follow-up. Regular annual follow-up with a physician is also recommended.

The safety of oats in CD has been extensively investigated. Clinical studies have shown that pure, uncontaminated oats are safe for most adults and children. Most commercially available oats are contaminated with wheat, rye or barley. Patients with CD must ensure that the oats they are eating are free from gluten contamination.

All patients with CD and DH should be encouraged to join the Canadian Celiac Association (CCA) and their local CCA chapter for valuable practical information and ongoing support. Detailed information on diagnosis and management of CD and DH is available for public and health care professionals at **http://www.celiac.ca**

References and recommended reading

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