# Celiac Disease: A Medical Puzzle

A guide to recognizing and managing this highly variable genetic illness.

**OVERVIEW:** Celiac disease is a T-cell–mediated, autoimmune, genetic illness that targets the small intestine and typically resolves with removal of gluten from the diet. More widespread serologic testing indicates that celiac disease affects 0.5% to 1% of the U.S. population, but presentation is highly variable and diagnosis is often missed or delayed. Strict adherence to a gluten-free diet remains the only treatment but can be challenging. This article outlines the pathophysiology of celiac disease, discusses signs and symptoms and the four disease types, describes testing, and addresses treatment and nursing implications.

**Keywords:** celiac disease, celiac sprue, gluten, gluten intolerance, gluten-sensitive enteropathy

*I am a registered nurse. I was treated for irritable bowel syndrome for more than 10 years. Although I was very careful with my diet, I never seemed to get better and felt sick most of the time. One day, I was admitting an elderly woman to home care and was reviewing her many diagnoses. She told me about her celiac disease and the symptoms she'd experienced. As she went down the long list, I put down my pen and looked at her. "I have the same symptoms," I said.*

Celiac disease is a T-cell–mediated, autoimmune, genetic illness that targets the small intestine; in most people the duodenum is affected first, although often the jejunum is also involved.[1] The disease has been described as "a permanent intolerance to ingested gluten that damages the small intestine, characteristically inducing crypt hyperplasia and villous atrophy, and typically resolves with removal of gluten from the diet."[2] (Gluten is a complex protein found in wheat and other grains.) Other names for the illness include celiac sprue, nontropical sprue, endemic sprue, gluten enteropathy, gluten-sensitive enteropathy, and gluten intolerance.

Although a plethora of articles have been written about this illness, many nurses and other clinicians are unaware of its prevalence and its ramifications. Celiac disease affects 0.5% to 1% of the U.S. population, according to a 2004 consensus statement by the National Institutes of Health, which also concluded that the disease "is widely underrecognized."[3] Yet clinicians are still being taught that celiac disease is a childhood illness, so that when adults present with its signs and symptoms, diagnosis is often missed or markedly delayed. The average time from onset of symptoms to diagnosis for adults in the United States is 10 years.[4] The authors of this article have either personally endured or witnessed the misery of misdiagnosis. This article provides an overview of celiac disease, outlines the pathophysiology, discusses signs and symptoms and the four types of celiac disease, and addresses diagnosis and

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disease management. In the interest of raising nurses' awareness, we offer our own illustrative stories as well.

## PREVALENCE AND RISK

**Prevalence.** Green and Jabri note that, although the disease was once believed to occur in only one in 3,345 people worldwide, recent studies show that actual prevalence is closer to one in 266 people worldwide,[5] with many nations showing higher rates.[6-8] And there are indications that prevalence is rising: for example, a retrospective Finnish study found that total prevalence among adults had doubled over two decades of study and that the increase couldn't be attributed solely to better detection.[9]

In Europe, celiac disease is one of the most commonly diagnosed genetic diseases.[7] It has "historically been considered rare" in the United States and elsewhere,[7] but more widespread use of serologic screening tests has revealed that this isn't the case. (For more on the history of how celiac disease came to be recognized, see *Historical Background*.[10-14]) In a five-year study by researchers at the University of Maryland Center for Celiac Research (UMCCR), more than 13,000 people were screened for antibodies; the researchers determined that among people with no known risk factors, the prevalence was 1:133.[7] Similarly, celiac disease has long been thought to be rare in other non-European countries, but now researchers believe it has been underdiagnosed. For example, Wang and colleagues found that, of 118 Chinese children with chronic diarrhea, 14 (12%) were found to have celiac disease.[15] And Bahari and colleagues found a prevalence rate of 1:114 in a convenience sample of "apparently healthy" southeastern Iranians.[6]

**Risk factors.** Celiac disease can manifest at any age.[16] It's unclear whether incidence increases with age. A Finnish study found increased incidence among people over age 55, but the researchers were unsure whether this reflected diagnostic delay, late emergence, or both.[17] Another small study concluded that the disease is indeed underdiagnosed in older adults.[18] Celiac disease appears to be more prevalent in women than in men,[16, 19-21] although this may reflect gender differences in health care–seeking behaviors rather than in prevalence.[22, 23] In a study of 1,436 people with gluten intolerance, Bardella and colleagues found an overall female-to-male ratio of 2.3:1.[24] People with autoimmune disorders may also be at higher risk for celiac disease.[25]

Because the disease is genetic, people with a family member who has celiac disease are at greater risk

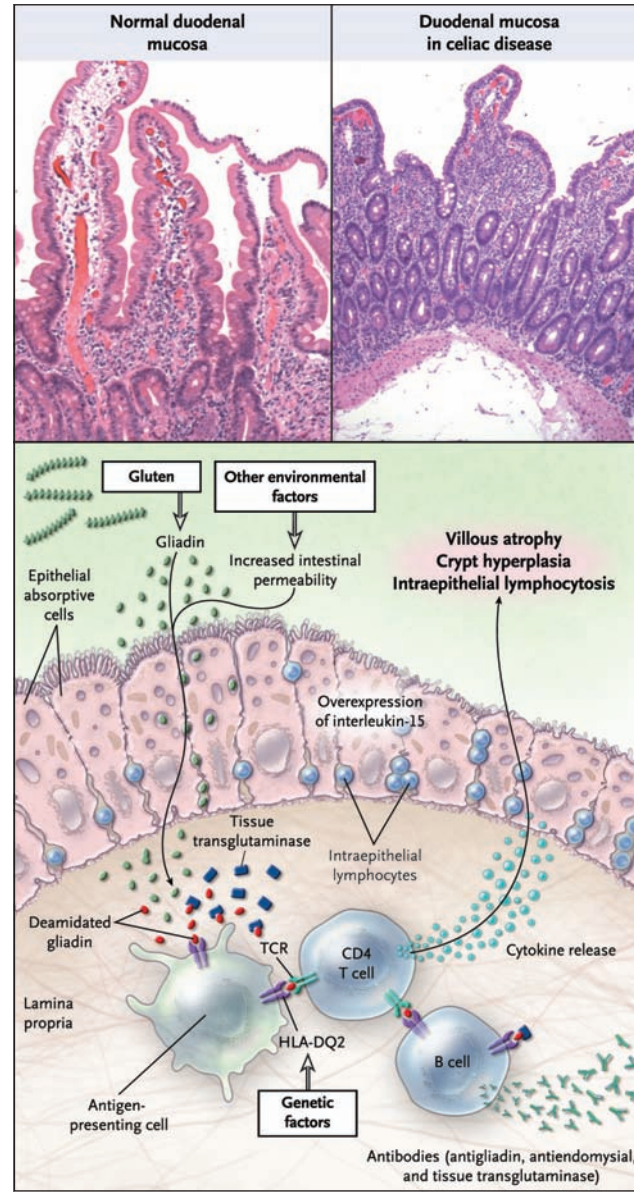


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**Figure 1. Interaction of Gluten with Environmental, Immune, and Genetic Factors in Celiac Disease.** Gluten is digested into amino acids and peptides. The gliadin peptides induce changes in the epithelium, where gliadin damages epithelial cells, resulting in increased expression of interleukin-15, which in turn activates intraepithelial lymphocytes. During infections or as the result of permeability changes, gliadin enters the lamina propria, where it is deamidated by tissue transglutaminase, allowing interaction with HLA-DQ2 (or HLA-DQ8) on the surface of antigen-presenting cells. Gliadin is presented to gliadin-reactive CD4+ T cells through a T-cell receptor, resulting in the production of cytokines that cause tissue damage. This leads to villous atrophy and crypt hyperplasia.

for developing the disease themselves. In the aforementioned UMCCR study, among at-risk subjects, researchers found prevalence rates of 1:22 in first-degree relatives of patients with celiac disease and 1:39 in second-degree relatives.[7] In a study of 168 asymptomatic siblings of people with celiac disease, serologic screening revealed that 24% had celiac disease themselves.[26] And in a concordance study of 47 sets of twins, Greco and colleagues found that in 13 of the 47 pairs identified, both twins had a clear diagnosis of celiac disease.[27]

*I am a registered nurse. My first symptom of celiac disease appeared when I was six, when I began bruising for no apparent reason. Over time, I suffered from 26 vague and multisystem symptoms. I went from doctor to doctor and was repeatedly tested for gallbladder disease and treated for anemia, osteopenia, and other symptoms. But no one ever put the puzzle pieces together. I was finally diagnosed at the age of 63, when my brother's diagnosis of celiac disease led me to believe I had it, too; a duodenal biopsy confirmed that I did. By then, I had a hemoglobin count of 8 g/dL, was enduring frequent migraine headaches and repeated bouts of bronchitis, and was severely depressed with suicidal ideation—all of which disappeared when I eliminated gluten from my diet.*

## PATHOPHYSIOLOGY

Three factors, environment, genetics, and immunology, are important to the pathogenesis of celiac disease (see Figure 1).[28-30]

**Environment.** Celiac disease is provoked by the ingestion of gluten, a protein found in wheat and wheat-related grains, including kamut, rye, spelt, and triticale, as well as barley.[31, 32] Even tiny amounts of gluten can cause damage to the intestines; thus lifetime avoidance is essential. This can be more challenging than one might think. Gluten is also used as a stabilizer, as an emulsifier, and as a thickening agent in a wide variety of processed foods, including many common sauces and marinades, processed meats and meat substitutes, soups, and candies. Cross-contamination can also be an issue. For example, experts disagree about the safety of ingesting oats because of possible cross-contamination with wheat during the growing or processing of oats.[28, 33] Many experts recommend eliminating oats from the diet as a precaution, unless the oats are specifically labeled gluten free.[34-36]

**Genetics.** In order to have celiac disease, a person must have the genetic predisposition for the disease. Although researchers have not yet identified all of the genes involved, such predisposition has been strongly linked to the genes that encode inherited human leukocyte antigen (HLA), specifically the alleles known as HLA-DQ2 and HLA-DQ8. HLAs are proteins found in most cells of the body, particularly white blood cells.[31, 37] According to Fasano, 95% of patients with celiac disease express the HLA-DQ2 allele, and the remaining patients test positive for the HLA-DQ8 allele.[31] However, these alleles are common, found in about one-third of the general population; having either allele is necessary but not sufficient for disease development.[31, 38] Non-HLA genes are thought to play a role as well.[39]

**Immunology.** HLAs guard the body by identifying other cells as *self* or *non-self*. In people with celiac disease, the HLA-DQ2 and HLA-DQ8 alleles identify gluten as an invasive foreign body and produce antibodies (immunoglobulins) that inflame the cells of the small intestines in an attempt to protect the body. The immunoglobulins IgA and IgG are involved, particularly IgA, which is abundant in the intestines.[29, 40, 41]

**Disease process.** The immunoglobulin-induced inflammation triggers an all-out assault by the immune system in an attempt to rid the body of the foreign invader. Several mechanisms are involved. The gluten molecule is resistant to normal protein digestion by peptidases, and a long peptide chain (a toxic fraction of gliadin, which is a component of gluten) remains after digestion. In people with celiac disease, the toxic fraction gets under the epithelial cells that line the intestinal villi, triggering an immune response and inflammation.[29, 36] Tissue transglutaminase (tTG), an enzyme, also appears to play a role. It's believed that tTG interacts with gliadin and converts it into a more toxic molecule, one that triggers the immune response in genetically predisposed people.[5, 25, 29] The actual damage is done by cytokines, specifically interferon gamma.[39, 42]

**In order to have celiac disease, a person must have the genetic predisposition for the disease.**

Over time, chronic inflammation leads to changes in the intestines. Flattening and atrophy of the intestinal villi lead to loss of absorptive ability. The new epithelial cells produced by the crypts are unable to move up the damaged villi, leading to crypt hyperplasia and leakage of serum from the crypts.[25, 39, 42]

## SIGNS, SYMPTOMS, AND DISEASE TYPE

Researchers have used an iceberg model in explaining celiac disease, with the smaller, visible tip representing symptomatic cases (and thus more likely to be diagnosed), while the larger, submerged portion represents asymptomatic and latent cases (less likely to be diagnosed).[43, 44] Diagnosis is all the more challenging because a significant proportion of patients may present with nongastrointestinal symptoms or have no symptoms at all.[3] In a health record review of 220 patients newly diagnosed with celiac disease, Balamtekin and colleagues found that 59% presented with gastrointestinal (GI) symptoms, 36% had non-GI symptoms, and 7% were asymptomatic.[45] For a list of some GI and non-GI symptoms, see *Signs and Symptoms of Celiac Disease*.[4, 41, 46-52]

Presentation is highly variable; taking a thorough patient history is paramount, as more than 200 signs and symptoms have been associated with the disease.[52] Symptoms may come on suddenly or gradually; and patients may present as slightly or gravely ill.[53] Symptom type and severity are related to the amount of intestinal damage and individual patient response to lack of nutrients.[4, 29]

**Types of celiac disease.** According to a 2004 consensus statement on celiac disease by the National Institutes of Health, celiac disease can be classified as classical, atypical, silent, or latent.[3]

*Classical celiac disease* (also called severe) usually involves villous atrophy of much of the small intestine.[3, 29] Because of the extensive intestinal damage, classical celiac disease is characterized by symptoms and sequelae of GI malabsorption, and includes symptoms such as diarrhea, abdominal pain, bloating and distension, constipation, and reflux.[3, 29, 45]

*Atypical celiac disease* may involve only the duodenum and is characterized by few or no GI symptoms; non-GI symptoms predominate.[3, 19, 29] Patients who present with mild GI symptoms such as reflux and bloating may be misdiagnosed with irritable bowel syndrome. Malabsorption of a single nutrient such as calcium or iron may occur, leading to symptoms

### Signs and Symptoms of Celiac Disease[4, 41, 46-52]

**Gastrointestinal**
- Abdominal distention
- Abdominal pain
- Acid reflux
- Diarrhea or constipation
- Flatulence
- Hypoglycemia
- Increased appetite or food cravings
- Lactose intolerance
- Nausea and vomiting
- Steatorrhea
- Unexplained weight loss or gain

**Musculoskeletal**
- Arthralgia
- Bruising
- Muscle cramps
- Osteopenia or osteoporosis
- Short stature

**Neurologic**
- Anger
- Anxiety
- Ataxia
- Attention deficit–hyperactivity disorder
- Behavioral disturbances
- Depression
- Impaired concentration
- Irritability or apathy
- Migraines
- Peripheral neuropathy
- Seizures
- Sleep disturbances
- Suicidal ideation or attempts

**Reproductive**
- Delayed menarche
- Delayed puberty
- Early menopause
- Infertility
- Recurrent abortion

**Dermatologic**
- Alopecia
- Aphthous stomatitis (recurrent)
- Dermatitis herpetiformis
- Follicular keratosis

**Pediatric**
- Behavioral disturbances
- Failure to thrive
- Poor school performance

**Other**
- Anemia
- Asthma
- Blurred vision
- Dental enamel hypoplasia
- Fatigue
- Night blindness
- Uveitis, bilateral

of anemia, osteopenia, or osteoporosis.[5, 16, 19, 25, 45, 54] Patients may initially present with apparently unrelated symptoms, such as dental enamel defects, peripheral neuropathy, irritability, or fatigue.[4, 25, 41, 55] Indeed, fatigue is a major complaint of people with celiac disease,[36] and may be caused by many different factors, including malabsorption of nutrients resulting in malnutrition, anemia, a concomitant autoimmune disease, and depression.[41]

For both classical and atypical celiac disease, diagnosis is confirmed "by serological testing, biopsy evidence of villous atrophy, and improvement of symptoms on a gluten-free diet."[3]

*My wife is a registered nurse. When I was disqualified from donating blood because I was severely anemic, I was sure I had cancer. My wife convinced me to see our family doctor. Blood work revealed that what I had was not cancer but celiac disease.*

*Silent celiac disease* refers to cases that are asymptomatic.[3] Patients learn they have celiac disease when it is detected via serologic screening or when villous atrophy is found during endoscopy or biopsy performed for other reasons.[3, 29]

*Latent celiac disease*. Patients with latent celiac disease have a positive serology but no villous atrophy on biopsy[3] or negative serology that turns positive later.[29]

Some experts distinguish between celiac disease, wheat allergy, and gluten sensitivity.[56, 57] Wheat allergy has been defined as "an adverse immunologic response to wheat proteins"; IgE antibodies play a central role.[57] Gluten sensitivity has been defined as "those cases of gluten reaction in which both allergic and autoimmune mechanisms have been ruled out (diagnosis by exclusion criteria)."[57] Further research is needed to clarify what some call the "spectrum of gluten disorders."[58]

**A closer look at two symptoms.** *Diarrhea* associated with celiac disease may have several mechanisms.[29] As prolonged inflammation destroys the epithelial barrier, the intestines become more permeable to substances usually not absorbed; they also ooze serum in a process called protein-losing enteropathy. Malabsorption of sugars and fats causes excess secretion of water in the colon, with resultant watery stool. Other undigested products, including bile salts and fatty acids, intensify the problem. Bacterial ingestion of undigested food products produces gas, causing flatulence, abdominal distention, and cramps. It's important to note that although diarrhea has long been considered the classic symptom of celiac disease, in recent years researchers have found it to be the main presenting symptom in fewer than 50% of cases.[25, 59]

*Dermatitis herpetiformis* is sometimes called celiac disease of the skin.[29] Characterized as an intensely itchy, blistering rash that may be accompanied by a burning sensation, it can occur anywhere on the body. It may disappear and then recur, and is usually mirrored on both sides of the body. Some people with dermatitis herpetiformis never develop GI or malabsorptive symptoms, and many people with celiac disease never develop the rash. Fortunately, as with the other symptoms of celiac disease, dermatitis herpetiformis responds to the removal of gluten from the diet.[29, 57, 60]

*I am a registered nurse. My path to a diagnosis of celiac disease started at my 41st birthday party. After eating pizza and cake, I had excruciating gas and loose stool. I didn't think much of it, but over the next month these problems worsened. I visited a gastroenterologist who surmised I had irritable bowel syndrome. My symptoms steadily got worse over the next few years. It got so that I could hardly eat anything—but, of course, I was eating mostly bread products to settle my upset stomach. When I developed painful blebs on my elbows and hands, I saw a dermatologist who concluded that I'd had a reaction to something I'd touched at work and recommended wearing long-sleeved shirts and gloves. At my next visit, he concluded that I had scabies. Fed up, and with my hands still covered with blebs and often bleeding, I went to a second dermatologist, who did a punch biopsy. Two weeks later, I finally had a diagnosis: dermatitis herpetiformis secondary to celiac disease.*

## TESTING FOR CELIAC DISEASE

Routine screening for celiac disease is not currently practiced in the United States.[61] Some researchers support screening, especially for people in high-risk categories, such as those with first- or second-degree relatives who have celiac disease and those who have another autoimmune disease.[62-64]

Clinicians should consider celiac disease when a patient has multiple or vague symptoms that don't seem to fit a clear diagnosis of another illness. The presence of any of the following should prompt testing: a first- or second-degree relative with celiac disease, selective IgA deficiency, connective tissue disorders such as rheumatoid arthritis or lupus, Down syndrome, autoimmune endocrinopathies such as type 1 diabetes or Graves' disease, epilepsy, dermatitis, and chronic

fatigue.[41, 65] Children under the age of three years with any of the above or with failure to thrive and persistent diarrhea should also be evaluated for celiac disease.[66]

Testing for celiac disease currently includes serologic testing for autoantibodies, including antigliadin, tTG, deaminated gliadin peptide, and antiendomysium antibodies.[67] Although the antigliadin antibodies test does not have good sensitivity, test panels include it to determine whether a person is IgA deficient. Combination testing identifies patients who are candidates for a jejunal biopsy, which remains the gold standard for diagnosis.[61]

Once gluten is removed from the diet, antibody levels decrease. In a longitudinal study of 20 patients, Midhagen and colleagues found that antibody titers fell sharply within one month after introduction of a gluten-free diet.[68] So it's important for clinicians to teach patients that they should continue to ingest gluten until all testing is completed.

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Genetic testing for the presence of the HLA genes associated with celiac disease is available. A negative result can rule out the possibility that a person will develop the disease, while a positive result indicates

## Historical Background[10-14]

**Tracking down the cause.** As early as the 1st century CE, the Greek physician Aretaeus of Cappadocia described an intestinal disease he named *koiliakos*, after the Greek word for abdomen (*koelia*).[10] In the early 19th century, Scottish pathologist Mathew Baillie wrote about a chronic diarrheal condition in adults that caused malnutrition and improved when patients lived mostly on rice. In 1887, British physician Samuel Gee lectured on "the coeliac affection," describing it as "a kind of chronic indigestion which is met with in persons of all ages, yet is especially apt to affect children between one and five years old."[11] Both Baillie and Gee rightly believed the illness was diet related, but the specific trigger remained unknown.[10, 11]

In 1924, Sidney Haas, a New York City pediatrician, published a paper describing his success in treating children with anorexia and celiac disease with a special diet.[10] Called the "specific carbohydrate diet" or "banana diet," it excluded potatoes and anything made from grain, including bread. The diet's success led Haas to believe that carbohydrates were the cause of celiac disease. But it wasn't until World War II that the real causative agent was identified. A Dutch pediatrician, Willem-Karel Dicke, noticed that, during war-related bread shortages, the death rate among children with celiac disease dropped from 35% to almost zero—and that when bread became available again, the children sickened and the death rate rose.[10, 11] After the war, Dicke and other researchers were able to determine that gluten was the culprit.

**Developing diagnostic standards.** In 1956, another breakthrough occurred. Margot Shiner, a pediatric gastroenterologist working in London, developed a jejunal biopsy device that permitted her to reach and biopsy the distal duodenum.[10, 12] This revealed "a specific, recognizable pattern of damage" to the mucosa,[10] and established the disease's histopathologic identity.[12]

In 1969, at a meeting in Interlaken, Switzerland, the European Society for Paediatric Gastroenterology (now the European Society for Paediatric Gastroenterology, Hepatology and Nutrition) developed a set of diagnostic criteria for celiac disease.[13] The criteria called for eliminating gluten from the diet until any lesions were normalized and then reintroducing gluten; if lesions recurred, celiac disease was confirmed. For over 20 years, this remained the accepted diagnostic standard worldwide. But the Interlaken criteria ignored another important discovery—the presence of certain antibodies in the blood following gluten ingestion. In 1964, Berger and colleagues found antigliadin antibodies in the blood of children with celiac disease.[10] Then, in 1973, Seah and colleagues identified antireticulins, which are autoantibodies,[14] and within a few years it became evident that celiac disease was sometimes associated with other autoimmune disorders.[10] In the late 1980s, Stefano Guandalini conducted a study demonstrating that in 95% of cases, using "strict clinical and laboratoristic criteria," a correct diagnosis of celiac disease could be reached with one initial biopsy.[10] Since about 1990, celiac disease has been accepted as an autoimmune disease associated with a specific gene.

that, whether or not the person has symptoms, regular antibody screening should be done to determine if and when the disease becomes active.[66]

### TREATMENT AND NURSING IMPLICATIONS

Strict adherence to a gluten-free diet remains the only treatment for people with celiac disease, and can be challenging. Fortunately, maintaining a gluten-free diet will usually reverse the trajectory of the disease, allow the intestines to heal, and cause symptoms to disappear.

Nurses can help patients to better understand both the disease and the importance of avoiding gluten. Gluten-free labeling is currently voluntary and not standardized, so "hidden" gluten is a real threat. Patients need to learn how to read labels. Ingredients lists may not list gluten directly. But if a food contains any of the glutenous grains or their derivatives (such as malt, which is made from barley), it shouldn't be consumed. Nurses, perhaps together with the team's dietician, can also teach patients how to eat a balanced diet that supplies essential nutrients while staying within a budget. For example, as they would do with any new food, patients can first buy a small amount of a gluten-free food to determine whether they like it before purchasing it in bulk. Many large supermarket chains now carry a greater variety of gluten-free foods, often at lower prices than health food stores. And most recipes can be adapted to suit a gluten-free diet without much additional expense. There has been scant research on whether people with celiac disease can benefit from vitamins and other supplements; but many people take them, and some manufacturers use wheat-based inactive ingredients in their formulations.[29] Similarly, medications can also contain gluten. Nurses should teach patients to check that any vitamins, supplements, and prescription and over-the-counter drugs they take are gluten free.

**People with celiac disease are at higher risk for other autoimmune diseases.**

Although gluten can be found in some nonfood products such as shampoos and lotions, it is not problematic unless ingested, as gluten molecules are too large to be absorbed through the skin.[69,70] But people with celiac disease should wash their hands after application to avoid hand-to-mouth exposure. Ingredient lists on lipsticks, lip balms, and oral and dental care products must be read carefully, as these products can be ingested. Manufacturers can be contacted directly for further clarification about ingredients in their products.

Many restaurants and restaurant chains are beginning to offer gluten-free choices on their menus. Nurses can encourage patients to ask about such options when eating out, and support them in doing so without embarrassment. Patients should also be taught to ask about possible cross-contamination during food preparation. For example, restaurant staff might be asked "Is gluten-free pasta cooked in water previously used to cook regular pasta?" and "Are gluten-free foods fried in oil previously used to cook foods that contain gluten?"

> *I am a registered nurse. I grew up in an Italian neighborhood and married a woman who only cooked Italian foods. Eliminating wheat from my diet seemed impossible. When I started reading cans, boxes, and labels, I was astounded to find wheat in just about everything—and the fact that gluten or wheat often wasn't mentioned was scary. Eating out at first was a nightmare, too. I've learned to carry gluten-free products when I travel. If an airline doesn't offer a gluten-free meal, I ask for kosher and usually find enough to eat. Now that I've found gluten-free beer and pizza, I'm a happy boy.*

Unfortunately, a small percentage of patients with celiac disease may not respond (or may stop responding) to a gluten-free diet; in such cases, the disease is called *refractory celiac disease*.[2,25] These patients develop complications such as diarrhea and malabsorption, and are at higher risk for ulcerative jejunitis and T-cell lymphoma. Treatment involves managing any nutritional deficiencies, possibly through total parenteral nutrition; corticosteroids or immunosuppressants may also be prescribed.

Patients need good psychosocial support as well. Because their symptoms are often vague, many patients go from provider to provider for years before receiving a diagnosis of celiac disease. (Indeed, many of us believed we were crazy because, for months and even years, no provider could find a reason for our symptoms.) Nurses can listen and provide patients with information and emotional support. They can also refer patients to knowledgeable providers who will take patients' complaints seriously, offer further information, and perform a thorough workup, including biopsy. The Celiac Sprue Association (www.csaceliacs.org) maintains a list of celiac centers that specialize in caring for patients with celiac disease, and will help patients in finding a site close to home.

*I am a registered nurse. During the early 1990s, I experienced severe, unbearable itching for months with intermittent breakouts of fluid-filled bullae. I saw several physicians and was diagnosed with eczema. The third dermatologist I saw biopsied the bullae, and this revealed that I had dermatitis herpetiformis. An initial trial of prednisone just made things worse; I was then started on dapsone, with immediate improvement. The dermatologist stated that there were some indications that the only ways to stop bullae formation were to take dapsone or completely eliminate wheat from my diet; but he felt that removing wheat would be "too difficult" and that I should just take the dapsone.*

*I was well maintained for 13 years. But if I missed even one dose, I developed a flare-up almost immediately. In 2007, I was finally tested for celiac disease, and the results were positive. In all that time, not one provider had recommended that I go wheat free and none offered any education on the disease. I educated myself—and them. When I learned that dapsone increases the risk of cardiac problems, I went wheat free and stopped the medication. I also changed my primary provider to one who actually listens to me, although he admits to having to look up information on celiac disease and dermatitis herpetiformis.*

For some patients, especially those newly diagnosed, involvement in a support group can be helpful. Participants can help support each other in making the transition to and staying on a gluten-free diet, learn more about celiac disease, and share information on available resources. (See *Resources*.)

*I am a registered nurse. One day I was standing in my office preparing to lick an envelope when a colleague who also has celiac disease entered the room and said, "You can't do that—wheat paste!" That was the moment when I really started to understand the ramifications of my new diagnosis.*

It's important that nurses encourage patients to follow up regularly with their primary care provider. People with celiac disease are at higher risk for other autoimmune diseases. Nurses should teach patients about the need for yearly serologic testing to ensure dietary adherence and assess for refractory celiac disease, periodic bone-density testing, and periodic assessment for nutritional deficiencies and possible malignancies.

## MUCH MORE IS NEEDED

Since under- and misdiagnosis of celiac disease are not uncommon, we believe that further education of both health care professionals and the public is

## A Common Tale

I'm a registered nurse. As an adolescent, I remember playing outside and coming in to tell my grandmother I didn't feel well. I would get cold and clammy, start to lose my vision, and eventually black out. When my mom asked my pediatrician what was wrong, she was told that my blood pressure was a little low and that I probably suffered from orthostatic hypotension. The pediatrician suggested I drink more water throughout the day and take care when changing positions. I followed these suggestions and my fainting became less frequent. Around the age of 10, I became borderline anemic. We didn't think much of this at the time since anemia ran in my family.

I went through high school and my first two years of college with few problems—except for the fainting. In my junior year I started fainting every day during my clinicals. I saw my doctor and was again told that it was probably orthostatic hypotension. She actually tried to push me out the door, and I pleaded with her to please run some blood tests because there was no way I could get through nursing school if I was blacking out in hospital hallways. She reluctantly gave me a lab slip, and a few days later she called to tell me that I had to see an endocrinologist because I had a thyroid problem.

I was diagnosed with Graves' disease and started on Synthroid. During my monthly blood tests, my endocrinologist commented that my anemia was getting worse and could be causing Synthroid inefficacy. She recommended that I mention this to my other doctors. After three visits with three more specialists, I was told that the anemia was normal because I was an actively menstruating female and that I should increase my iron intake. A few months later, my endocrinologist found that my anemia was getting much worse and sent me for celiac testing, as celiac disease often coincides with thyroid disease. I was skeptical at first because I didn't have any of the GI symptoms we had learned about in nursing school. But within a month I was diagnosed. If it weren't for my endocrinologist finally making the connection, I would still be looking for answers.

paramount. More articles in professional and lay journals are needed to raise awareness. Celiac disease should also be included in nursing and medical school curricula.

Celiac disease has not been explored widely in the nursing literature, especially in the United States. In particular, further investigation into the psychosocial aspects of celiac disease is essential. Being diagnosed with celiac disease is as life changing as being diagnosed with diabetes. One's health and one's diet—even feelings about one's self—change. Qualitative research about the lived experiences of patients

*My mother is a registered nurse. My high school years were rather stressful for both my family and me. Although I tested above average in intelligence, I struggled to achieve passing grades. I seemed to have more trouble staying out of trouble than my friends did. When I got my driver's license, I had a terrific feeling of freedom; but after I totaled three cars, the excitement was gone. I was diagnosed with attention deficit disorder (ADD) and started on medication that helped me focus, but I still felt like I had a head full of cotton, what some have described as "brain fog."*

*When I started having GI symptoms, I blamed the ADD medication; but since my dad has celiac disease, my parents decided to have me tested. My tests came back positive. Halfheartedly, I began following a gluten-free diet, but as a teenager it was hard for me to accept the restrictions. When I first started college, I continued to struggle, but sophomore year has been easier. I realize I do better and feel better on the diet. The choice and consequences are mine: go off the diet and struggle, or stay on the diet and do well. I know the difference now.*

## Resources

**Health-related Resources**

Celiac Disease Center at Columbia University
www.celiacdiseasecenter.columbia.edu

Celiac Disease Foundation
www.celiac.org

Celiac Sprue Association
www.csaceliacs.org

Gluten-Free Diet
www.glutenfreediet.ca

Gluten Free Drugs
(covers prescription and over-the-counter drugs)
www.glutenfreedrugs.com

Mayo Clinic
www.mayoclinic.org/celiacdisease

National Digestive Diseases Information Clearinghouse
http://digestive.niddk.nih.gov/ddiseases/pubs/celiac/celiac.pdf

National Foundation for Celiac Awareness
www.celiaccentral.com

The University of Chicago Celiac Disease Center
www.celiacdisease.net

The University of Maryland Center for Celiac Research
www.celiaccenter.org

**Other Useful Resources**

Celiac.com
www.celiac.com

Gluten Free Registry
(searchable database of "gluten-free friendly" restaurants, grocers, and other businesses)
www.glutenfreeregistry.com

Gluten-Free Restaurant Awareness Program
(searchable database of "gluten-free friendly" restaurants)
www.glutenfreerestaurants.org

GlutenFreeTravelSite.com
www.glutenfreetravelsite.com

Lab Tests Online
(enter "celiac" in search engine)
http://labtestsonline.org

*Living Without*
(magazine on living with allergies and food sensitivities)
www.livingwithout.com

with autoimmune diseases will further our understanding of the impact of such illnesses on a patient's quality of life. Quantitative research about other aspects of celiac disease is called for as well.

More accurate diagnostic criteria and standardization of serologic tests are needed. This would help to support screening initiatives, thus reducing morbidity and mortality. Several comorbidities are frequently associated with celiac disease, including type 1 diabetes, cardiomyopathy, Down syndrome, peripheral neuropathy, primary biliary cirrhosis, thyroid disease, and cancer.[2, 25, 71-73] In a study of 653 patients with non-Hodgkin lymphoma and 5,720 healthy controls, Catassi and colleagues concluded that celiac disease was associated with a higher risk of lymphoma, particularly gut lymphoma.[74] According to the Celiac Sprue Association, undiagnosed celiac disease increases the risk of cancer by 200% to 300%.[75]

Studies have found significantly higher death rates in people with celiac disease compared with those without the disease. In a retrospective cohort study of more than 46,000 patients with celiac disease, celiac disease with inflammation, or latent celiac disease, all based on duodenal or jejunal biopsies, Ludvigsson and colleagues found an increased risk of mortality in all three groups.[76] And in a retrospective study of 228 patients, using histologic information, Cottone and colleagues found a higher mortality rate in patients with celiac disease compared with the general population.[77] Twelve deaths were observed, whereas only three were expected.

Further research is needed to determine exactly what level of gluten might be ingested without triggering an autoimmune response. Since adherence to the diet is paramount and since unintentional ingestion is possible, a better understanding of the association between even minute amounts of gluten and the disease process will help patients in their efforts to adhere. An accurate testing procedure for detecting gluten in foods and established standards for gluten-free foods in the United States are also essential. These will lay the foundation for accurate, consistent food labeling with regard to gluten. Without such labeling, people with celiac disease must often avoid foods that might actually be safe and nutritious. ▼



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