Celiac Disease

What is celiac disease?

Celiac disease is an inherited autoimmune disorder that causes a reaction in your body to the protein, gluten. Gluten in your digestive system triggers your immune system to produce antibodies against it. These antibodies damage the lining of your small intestine (the mucosa). Damage to the mucosa in your small intestine impairs its ability to absorb nutrients from your food, causing nutritional deficiencies.

Gluten is a protein found in grains — particularly wheat, barley and rye. These grains, especially wheat, make up many of the staple foods of the standard Western diet, from breads and cereals to pasta and baked goods. Besides that, gluten often appears as an additive in food products where you wouldn't expect to find it, such as sauces, soups and packaged foods. Beer is usually made from barley or rye.

How does celiac disease affect my body?

Celiac disease affects your small intestine. This is where most of the nutrients from your food are absorbed, including proteins like gluten. But when you have celiac disease, gluten in your small intestine triggers an immune response. Your immune system sends inflammatory cells and antibodies to destroy the gluten molecules. These cells damage the mucous membrane lining your small intestine (mucosa).

The mucosa that lines your small intestine is vast but is scrunched up into many folds and fingerlike projections called villi. If you stretched it all out, it would cover a tennis court. The folds and projections increase the surface area in order to absorb as many nutrients as possible during digestion. But immune cells triggered by celiac disease erode and flatten these projections, diminishing the surface area.

Is celiac disease serious?

Damage to your small intestine can have serious consequences. Your small intestine absorbs nutrients from your food through the mucosa. If the mucosa is damaged, it won't be able to absorb nutrients as it should. This is called malabsorption. It can lead to malnutrition and to many other conditions that follow from the lack of different nutrients. In children, it can cause stunted growth and development.

Who gets celiac disease?

Celiac disease is most commonly found in people of Northern European descent. It's estimated to affect 1% of the populations of Europe and North America. You have a 10% chance of developing the disease if you have a first-degree relative, such as a parent or child, who has it. About 97% of people diagnosed with celiac disease have a recognizable gene mutation associated with it (*HLA-DQ2* or *HLA-DQ8*).

What other risk factors contribute?

Celiac disease is more common in people with certain inherited chromosomal disorders, such as Down syndrome. It's also more common in people with certain other autoimmune diseases. These diseases often share common genes, and they also have a way of triggering each other. Like other autoimmune diseases, celiac disease is more common in people assigned female at birth (AFAB). The ratio is at least 2:1.

You're more likely to have celiac disease if you:

- Are white.
- Have a relative who has it.
- Have a chromosomal disorder such as Turner syndrome, Williams syndrome or Down syndrome.
- Have another autoimmune disease, such as Type 1 diabetes, rheumatoid arthritis, microscopic colitis or Addison's disease.
- Were assigned female at birth.

Symptoms and Causes

What causes celiac disease?

Many autoimmune diseases, like celiac disease, are at least partly inherited (genetic disorders). That means a particular gene mutation that's passed down through family lines makes you more susceptible to developing it. But not everyone with the gene mutation develops the disease, and not everyone who develops it has one of the known genes. Other factors appear to be involved in triggering it.

How does a person get celiac disease?

One theory is that it's triggered by some type of significant physical stress that overextends your immune system. Healthcare providers have observed that the disease often shows up after a physical event such as surgery, illness or pregnancy, or a severe emotional event. Another theory is that microorganisms living in your gut are involved. More research is needed to explore these theories.

When does celiac disease develop?

Celiac disease can appear at any age after you or your child has begun eating gluten. Healthcare providers most commonly see it appear during two distinct age windows: early childhood, between 8 and 12 months old, and mid-life, between the ages of 40 and 60. The window in early childhood is typically when children begin eating solid foods that may include biscuits or cereals with gluten in them.

What are the symptoms of celiac disease?

Symptoms of celiac disease vary widely among people, which can make it hard to recognize. Some people don't notice any symptoms at all. Some experience indigestion and other gastrointestinal (GI) symptoms after eating gluten. Some only have vague symptoms of nutritional deficiencies later on, when real damage has been done. In these people, symptoms of anemia may be the first to present.

You might have:

Gastrointestinal symptoms, such as:

- Stomach pain.
- Bloated stomach.
- Gas.
- Constipation.
- Diarrhea.
- Fatty stools.

Symptoms of iron-deficiency anemia, such as:

- Weakness and fatigue.
- Pallor (pale complexion).
- Cold hands.
- Brittle or concave nails.
- Headaches.
- Mouth sores.

Other symptoms of malnutrition, such as:

- Unintended weight loss.
- Growth delays and failure to thrive in children.
- Muscle wasting or low muscle tone.
- Dental enamel defects, such as pitting, mottled or translucent-looking teeth.
- Abnormal periods or difficulty getting pregnant.
- Mood changes, most commonly irritability in children and depression in adults.

Dermatitis herpetiformis: About 15% of people with celiac disease develop this chronic skin condition as a side effect. Also called the "gluten rash" or the "celiac rash," the same gluten antibodies that damage your small intestine in celiac disease cause this condition. Dermatitis herpetiformis manifests as an itchy rash that looks like clusters of bumps or blisters. It typically affects your elbows, knees, buttocks or scalp.

Diagnosis and Tests

How do you know if you have celiac disease?

You might suspect you have celiac disease if you have gastrointestinal symptoms after eating gluten. Many people appear to have a sensitivity to gluten or wheat products in their diet. Food intolerances can cause uncomfortable symptoms after eating, but they don't damage your intestines the way celiac disease does. To diagnose celiac disease, healthcare providers will look for evidence of this damage.

It's important to get tested for celiac disease before you try a gluten-free diet, so the tests can reveal how gluten actually affects your body. Once you begin avoiding gluten, your gut will begin to heal. Healing is good, but it'll erase the evidence of celiac disease. You and your healthcare provider need to know for sure if you have it in order to know what kind of care you'll need going forward.

How do healthcare providers test for celiac disease?

Healthcare providers use two methods of testing for celiac disease. They prefer to use both together to confirm the diagnosis. The first method is blood testing. Providers test a sample of your blood for the gluten antibodies that damage your intestines. Then, they'll look for the damage itself. This requires taking a small tissue sample from your small intestine (biopsy) to examine under a microscope.

To take the sample, a gastroenterologist will perform an endoscopic exam of your small intestine. Endoscopic procedures involve passing a tiny camera through your body on the end of a long, thin catheter. An upper endoscopy passes the endoscope down your throat into the first part of your small intestine. Guided by the camera, the endoscopist can pass tools through the catheter to take a biopsy.

Additional tests

After confirming celiac disease, your provider will want to test your blood for specific vitamin and mineral deficiencies. Severe deficiencies can have wide-ranging effects on your body and may need to be treated directly with supplements. Common findings include iron-deficiency anemia, vitamin-deficiency anemia and vitamin D deficiency. You may also be low in electrolytes, such as calcium.

Management and Treatment

How do you treat celiac disease?

The first and most important step in treating celiac disease is to stop eating gluten. You can't change the way your body reacts to gluten, but you can prevent gluten

from triggering that reaction. When you stop eating gluten, your small intestine will begin to heal and will soon be able to absorb nutrients again. You have to maintain a strict gluten-free diet for life, though, to avoid hurting your small intestine again.

Additional treatment may include:

- Nutritional supplements to replace any serious deficiencies.
- Specific medications to treat dermatitis herpetiformis, such as dapsone.
- Corticosteroids for severe inflammation that's not responding fast enough to the diet.
- Continuous follow-up care, including regular testing to make sure the disease is controlled.

How long does the treatment take to work?

Most people find their symptoms begin to improve almost immediately after starting a gluten-free diet. It may take several weeks to replace your nutritional deficiencies and several months for your gut to fully heal. It can take longer in some cases, depending on the extent of the damage and how long it's been going on. You can also prevent your body from healing if you aren't strict with your diet.

Outlook / Prognosis

What is the outlook for people with celiac disease?

Most people who've been diagnosed and have stopped eating gluten have an excellent prognosis. Most of the damage done by celiac disease can be undone. If you continue to have symptoms, it may be that you're consuming small amounts of gluten without realizing it, or you may have a secondary condition. Only 5% of people have truly refractory celiac disease that doesn't respond to diet.

What happens if you don't avoid gluten?

If you go many years before being diagnosed or you don't succeed in avoiding gluten afterward, the effects of celiac disease can be more severe and long-lasting. Malnutrition can affect your nervous system and skeletal system and some of these effects are hard to reverse, especially when they occur during childhood development. Chronic inflammation can also lead to other problems in your intestine.

What are the possible long-term complications of celiac disease?

Complications of chronic malnutrition can include:

- Rickets in children or osteomalacia in adults.
- Osteopenia and osteoporosis.

- Permanent dental enamel defects.
- Nervous system effects (peripheral neuropathy), including tingling and numbness, muscle spasms and balance and coordination problems (ataxia).
- Growth and developmental delays in children, short stature.
- Attention and learning disabilities.

Complications of chronic inflammation can include:

- Compromised immunity. When your immune system is chronically overactive, it's left with fewer resources to address an acute attack, such as an infection. This can make you more vulnerable to getting sick. It also makes other autoimmune diseases more likely to trigger. Studies show your chance of developing another autoimmune condition goes up the longer celiac disease goes untreated. Celiac disease itself may also become increasingly slow to respond to treatment. Those who go untreated longer are most at risk of developing refractory disease.
- Additional food intolerances. When your intestinal mucosa is continuously diminished, you may lose the ability to break down nutrients that you previously could break down. People with untreated celiac disease frequently develop lactose intolerance as well, and other common intolerances can follow. As your mucosa diminishes, so does the variety of foods you can eat.
- Ulceration and scarring. Severe inflammation can lead to ulcers in your small intestine. A severe ulcer may wear all the way through your intestinal wall (gastrointestinal perforation), which is a medical emergency. Inflammation can also lead to scarring inside your intestine. Excessive scar tissue can cause your intestine to narrow (stricture,) which can lead to bowel obstruction.
- Collagenous sprue. This rare disease of the small intestine is strongly linked to celiac disease, although the link isn't entirely clear. It causes permanent collagen deposits in the lining of your small intestine, which prevents it from absorbing or secreting. Unlike the erosion caused by celiac disease, these collagen deposits don't heal. They cause permanent malabsorption problems.
- Liver disease. Celiac disease affects some people's livers more than others. Early blood tests may show elevated liver enzymes. Over time, they develop chronic liver disease and progressive liver damage. Healthcare providers aren't sure why this is, but they suspect it's related to chronic inflammation.
- Cancer. Chronic inflammation leads to an increased risk of cancer in your small intestine. Studies show about 7% of people with celiac disease develop intestinal lymphomas, usually after several decades. There's also a slightly increased risk of intestinal adenocarcinoma and esophageal cancers.

Living With

How do I stick to a gluten-free diet?

Avoiding gluten in all its forms can be daunting at first. You'll have to learn to read labels carefully and watch out for accidental contamination. The good news is that there's an abundance of resources available to help you navigate your new diet. Your healthcare provider will refer you to a registered dietitian to get you started. There are also many dedicated support groups and references online.

What foods trigger celiac disease symptoms?

Gluten is found naturally in certain grains and products made from grains, including:

- Wheat (including semolina, durum, emmer, bulgur, spelt, farina, Kamut® or Khorasan wheat, couscous).
- Barley (and malt made from barley).
- Rye.

These grains are used to make:

- Bread and baked goods.
- Hot and cold cereals.
- Noodles and pastas.
- Beer, lager and ale.
- Malted liquors.
- Malt vinegar.

It's also added to many processed food products, such as:

- Soups and sauces.
- Dressings and condiments.
- Processed meats, such as hot dogs and deli meats.
- Processed cheese, yogurt and ice cream products.
- Packaged dinners.
- Sweets.

Make sure to check labels for hidden gluten before eating processed foods.

Quick tips for living with celiac disease:

- Develop a go-to home menu. Consult some recipe guides and come up with a few recipes that you think you could eat often and throw together easily. Get in the habit of keeping those ingredients stocked so you always have something you can eat without thinking about it.

- Keep a few gluten-free snacks on hand in your car, bag or office desk in case you can’t find anything else to eat when you’re away from home.
- Find a few favorite restaurants with reliable gluten-free menus or where you know you can order certain meals safely. Check menus online before arriving and speak with the staff in advance if necessary to make sure they can accommodate you.
- Carry gluten test strips that can test the gluten content of new foods when you aren’t certain what’s in them.