Is it celiac disease?

Help your patients develop defenses against celiac disease through proper diagnosis and treatment

BY ANKUR A. CHUGH, MD

Celiac disease is the abnormal immune response to the ingestion of gluten (found in wheat, barley and rye). Symptoms within the GI tract can include abdominal pain, diarrhea, vomiting, weight loss and failure to thrive. Extra-intestinal (EI) manifestations can include iron-deficiency anemia, osteoporosis, arthritis, headaches and

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dermatitis herpetiformis. EI symptoms typically present after the age of 5. GI and EI symptoms typically resolve in 12 to 24 months once the patient is on a gluten-free diet.

**How is celiac disease diagnosed?**
A typical diagnosis is made with abnormal celiac serologies and an abnormal intestinal biopsy. There are many celiac serologies, which include the anti-gliadin antibody (AGA), deaminated gliadin peptide (DGP) antibody and tissue transglutaminase (TTG) antibody, all of which have an IgG and IgA test. The sensitivity and specificity of the serologies are different, and as such, current recommendations are to only get the TTG IgA antibody, along with the total IgA. Patients who are IgA deficient may not mount an appropriate TTG IgA antibody response, and in these cases, the TTG IgG antibody should be obtained. For patients under 2 years old, the DGP IgG antibody should be obtained along with the TTG IgA antibody.

**What if my patient wants to stop eating gluten?**
Obtain the patient’s celiac serologies before they stop eating gluten. This will help clarify if they have celiac disease or gluten sensitivity. If the celiac serologies are abnormal, they should be referred to a pediatric gastroenterologist for a scope. For the scope to be accurate, the patient needs to keep eating gluten. North American Society for Pediatric Gastroenterology, Hepatology, and Nutrition (NASPGHAN) guidelines suggest two pieces of bread a day is enough to have accurate biopsy results.

**What if my patient is avoiding gluten?**
First, try to understand how much gluten, if any, the patient is consuming. If it’s equivalent to at least two pieces of bread a day, both lab and biopsy results should be accurate. Some gastroenterologists might be OK with doing a scope if the patient is eating at least one cracker a day for three weeks prior to the scope.

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**At-risk patients for celiac disease**

- IgA deficiency: 2%
- Williams syndrome: 8%
- Type 1 DM: 3–12%
- Multiple sclerosis: 11%
- Autoimmune thyroiditis: 2–15%
- Sjogren’s syndrome: 2–15%
- Turned syndrome: 4–8%
- Down syndrome: 5–12%
- Autoimmune hepatitis: 6–15%
- 1st degree relative with celiac disease: 11%
- Addon disease: 6%
- Autoimmune liver disease: 6–8%
- IgA nephropathy: 4%

The risk of developing celiac disease is increased by the percentages noted for patients with:
If the patient won’t eat any gluten, consider genetic testing for celiac disease. While 20 to 30 percent of the population has genes that put them at risk for celiac disease, the lack of these genes effectively rules it out. Genetic testing, however, can be expensive and may not be covered by insurance, so this should be discussed before ordering the test.

**Is a scope really necessary to diagnose celiac disease?**

The European guidelines indicate that the diagnosis of celiac disease can be made with elevated celiac serologies >10x normal, positive celiac genetics and a positive anti-endomysial antibody (EMA), a very specific test for celiac disease. However, NASPGHAN recognized the European criteria without affirming it, citing celiac disease as a lifelong diagnosis with concomitant disorders, such as H. pylori and eosinophilic esophagitis, that can be diagnosed on biopsy.

**My patient was diagnosed with celiac disease. What’s next?**

The physician making the diagnosis should educate the patient and their family about the disease and refer them to a dietician to learn about the gluten-free diet. The gluten-free diet is difficult, and it can take time for patients and families to learn and strictly follow.

Celiac serologies (TTG IgA) should be checked three to six months after diagnosis to ensure they are improving, and annually after that. The 2016 NASPGHAN guidelines suggest additional labs be obtained at diagnosis (CBC, LFTs, calcium, thyroid, vitamin D, iron) and annually thereafter (CBC, vitamin D, thyroid); however, they acknowledge that these recommendations are based on expert consensus opinion rather than strong evidence. In addition, a recent study suggests that even if lab abnormalities are detected, they resolve within one year of being on a gluten-free diet.

**My patient is asymptomatic. Do they need a gluten-free diet?**

The short answer is yes. Untreated celiac disease can lead to both GI and EI symptoms down the road. Symptoms such as osteoporosis, iron-deficiency anemia and short stature can develop. In addition, the risk of intestinal lymphoma, while low, is increased by about three to six times in patients with untreated celiac disease.

**Who needs to be screened for celiac disease and how often?**

First-degree relatives of patients with celiac disease should be screened. European guidelines suggest starting with celiac genetics because, if they are negative, you can stop screening. However, not all insurance covers genetic testing for celiac disease, and siblings of patients with celiac disease often have the genetics that put them at risk for celiac disease, making the test less helpful. Another option is to obtain the TTG IgA every three years, or sooner if symptoms...
The Neurogastroenterology, Motility and Autonomic Disorders Program at Children's Hospital of Wisconsin offers one of the world’s largest teams of experts in the diagnosis and treatment of these disorders. Families from around the United States and beyond seek treatment at Children’s because of our team’s deep level of expertise, high volume of patients with motility disorders and cutting-edge diagnostic tools.

Children’s is recognized as a Gastrointestinal Motility Center of Excellence by the American Neurogastroenterology and Motility Society. In addition, ours is one of the few pediatric programs in the nation that specializes in autonomic disorders that have a gastrointestinal component.

For example, our Cyclic Vomiting Syndrome Program is the first and largest program to focus on treating children who are experiencing severe, recurring vomiting episodes.

We offer a full range of state-of-the-art testing and treatment options, including manometry studies and gastric pacing. Our comprehensive, interdisciplinary approach incorporates the expertise of pediatric specialists across many specialties, including general surgery, neurology, genetics, nutrition, immunology and behavioral health. These specialists use the best, safest equipment for kids, including imaging and scoping equipment that is the right size for children.

In addition, our team is actively involved in research on functional GI, motility and autonomic disorders, which deepens our understanding of how to best treat these conditions.

Learn more about the Neurogastroenterology, Motility and Autonomic Disorders Program at chw.org/neurogi.
Are there new developments for celiac disease?
Lots! There are enzymes (similar to Lactaid) on the market that allow patients to digest small amounts of gluten. These are best used when patients are trying to avoid contamination in their diet, as they do not digest large amounts of gluten. There is a product on the market that can measure gluten in the urine and stool, which can detect or confirm accidental ingestion. There is a vaccine in development that would allow patients to consume small amounts of gluten. There is also a test that might be able to diagnose celiac disease when a patient is not eating gluten. Finally, there are a few drugs in clinical trials that are trying to alter the pathogenesis of celiac disease.

What’s new at Children’s for celiac disease?
We have a dedicated team of 17 pediatric GI providers who are well trained in caring for patients with celiac disease. We also have a team of dietitians, and all our patients diagnosed with celiac disease have a one-on-one education session with one of them. The session includes the patient and any family members who care for the patient. Our guidelines for celiac disease should be available on chw.org in late 2018.

In addition, with Martin J. Hessner, PhD, we have begun translational research attempting to identify a unique inflammatory signature in patients with celiac disease before they develop celiac disease itself. A clinical trial is also underway in healthy children with a sibling affected by type 1 diabetes to determine the role of dietary gluten and other grain proteins as potential risk factors in type 1 diabetes progression. The study is sponsored by the American Diabetes Association and led by Susanne M. Cabrera, MD, a pediatric endocrinologist at Children’s.