Editorial

Celiac disease: more common yet "atypical" than previously thought

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Jessica W Madden
Boston Children's Hospital, USA

Correspondence: Jessica W. Madden, M.D., Boston Children☐s Hospital, 106 Prescott St., Reading, MA 01867, Boston, MA, USA, Tel 216-313-2016, Email Jessica.madden@childrens.harvard.edu

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Editorial

Celiac disease, an autoimmune enteropathy triggered by the consumption of gluten, can develop in genetically predisposed children (HLA DQ2 and/or DQ8 positive) at any point from 9 months of age through adulthood. Although the incidence of celiac disease in both North America and the bulk of Europe is approximately 1 in 100, patients with a first-degree relative with celiac disease are at a much higher risk of development. For example, up to 25% of children who are homozygous for HLA-DQ2 will develop evidence of celiac autoimmunity by age 6.[1] Additional risk factors for the development of celiac disease include type 1 diabetes, autoimmune thyroid disease, Trisomy 21, Turner syndrome, William's syndrome, and selective IgA deficiency.[2] The celiac genes (HLA-DQ2 and DQ8) contribute 40% of the risk of developing celiac. Environmental risk factors for celiac disease include infant feeding patterns, early infections, gut microbiota, receiving multiple doses of antibiotics during early life and the amount and timing of initial gluten exposure.[3]

Although the "classic" symptoms of pediatric celiac disease include abdominal pain and distension, diarrhea, and failure to thrive there has been increased recognition over the last decade that many children with celiac disease present with "atypical" symptoms. These atypical symptoms include fatigue, iron deficiency anemia, dermatitis herpetiformis, dental enamel defects, aphthous ulcers, arthritis and arthralgias, low bone mineral density, elevated liver enzymes, short stature and delayed puberty.[2] Neurologic and psychiatric symptoms of celiac disease in children include cerebellar ataxia, recurring headaches, peripheral neuropathy, seizures, anxiety, panic attacks, and depression.[2,4] In addition, many children with celiac disease are asymptomatic. Hence, the need for celiac screening in high-risk groups, including those with affected family members, as well as children with any of the aforementioned atypical symptoms of celiac disease.

A basic celiac disease screening panel should include both a serum IgA level and a TTG (tissue transglutaminase) IgA. In children who are younger than age 4 years of age, as well as those with a selective IgA deficiency, a DGP (deamidated gliadin peptide) IgG can be tested. The anti-gliadin IgA is no longer considered to be an adequate celiac screening test due to having low specificity. Children need to be consuming gluten at the time of testing, as it takes only 2 weeks on a gluten-free diet to make the results of a patient's celiac disease testing unreliable.[5] Confirmatory testing for celiac disease consists of endoscopy with small bowel biopsy. At least 4 to 6 tissue samples should be obtained for histologic analysis, including at least one sample from the duodenal bulb, as the villous blunting in celiac disease can be confined to solely this area.[2,5]

The only current treatment for celiac disease is strict adherence to the gluten free diet for life; however, there are multiple therapies in development to augment the gluten-free diet and to help prevent symptoms from inadvertent gluten ingestion via cross-contamination.[6] It is crucial for parents and children diagnosed with celiac disease to meet with dieticians who are well versed in celiac and the gluten free diet as the most common cause of persistent symptoms is accidental gluten ingestion. In addition children with celiac disease may have multiple vitamin and mineral deficiencies at the time of diagnosis that may need to be corrected, including low levels of Vitamin D, folate, and zinc.

In conclusion, celiac disease is a relatively common autoimmune disease with a myriad of presenting symptoms that can develop at any point during life. Early diagnosis and treatment of celiac disease can lead to an improved quality of life and decrease in complications and also presents an opportunity for at-risk family members to be periodically screened.

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Conflicts of interest

The authors declare there is no conflict of interests.

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