Going Beyond Gluten-Free

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Celiac disease (CD) is an autoimmune, small intestinal enteropathy caused by a permanent sensitivity to gluten from wheat, rye, and barley in genetically susceptible individuals. Affecting ~1% of children,[1] CD is more common than sickle cell disease or type 1 diabetes mellitus. The prevalence of CD continues to increase, partially due to increasing clinical and public awareness.[2] Even so, it is believed that a large majority of patients remain undiagnosed. Young children often present with gastrointestinal symptoms such as bloating, diarrhea, abdominal pain, and failure to thrive, whereas older children and adolescents are more likely to present with poor growth and extraintestinal symptoms such as anemia and delayed puberty.[3] Once diagnosed, treatment requires lifelong adherence to a strict gluten-free diet and failure to do so places patients at risk of multiple health concerns, including suboptimal growth, progressive osteoporosis, enteropathy-associated T-cell lymphoma, and shortened life expectancy.[3,4] Thus, timely diagnosis and treatment, with regular follow-up to ensure compliance, are essential for the long-term health and well-being of all who have CD.

Current guidelines for the diagnosis of CD recommend the use of the serum tissue transglutaminase immunoglobulin A antibody (tTG-IgA) as the most sensitive, specific, and cost-effective test for identifying pediatric and adult patients who may have CD.[1,5] It is recommended that those with a positive tTG-IgA result undergo endoscopy with small intestinal biopsy to look for the characteristic histologic changes to confirm the diagnosis. Recent guidelines from the European Society for Pediatric Gastroenterology, Hepatology, and Nutrition suggest that in some cases with tTG-IgA levels >10 times the upper limit of normal it may be possible to make the diagnosis without a biopsy.[5] For many reasons, these recommendations have not yet been widely adopted in the United States. Although the current guidelines provide good recommendations for the diagnosis and treatment of CD, they provide little guidance on how patients should be monitored and followed after the initiation of treatment.

In this issue of *Pediatrics* the article by Snyder et al, entitled "Evidence-Informed Expert Recommendations for the Management of Celiac Disease in Children,"[6] attempts to address questions regarding what tests and practices should, or should not, become the standard of care surrounding patients with CD. This group of experts in pediatric CD first identified 7 areas in the management of CD that were believed to be in need of being addressed and then undertook an extensive review of the available literature to develop recommendations. On the basis of the available data and their expert opinion, they used an anonymous voting technique to reach consensus in developing these recommendations, and further reported the quality of data, grade of evidence, and strength of the recommendation in each instance.

Their recommendations for the initial diagnosis of CD are in accordance with published guidelines; IgA-tTG with serum IgA level should be the test of choice followed by endoscopy with biopsies.[1,5] In addition, they support the judicious use of the endomysial antibody and recommend HLA testing

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only for special cases when there may be questions surrounding the diagnosis. Although they comment on the European Society for Pediatric Gastroenterology, Hepatology, and Nutrition's nonbiopsy diagnosis recommendations, this was not a topic on which they voted and may need to be revisited in the future.

Perhaps more helpful were their recommendations on appropriate ancillary testing at the time of diagnosis and subsequently in the areas of bone health, endocrinology, hematologic issues, liver issues, and nutrition in the context of CD. Although some of the recommendations are largely "common sense," as in all patients with CD should have routine assessment of anthropometric measures and all patients should be seen by a dietitian well versed in CD and a gluten-free diet, the majority of topics examined pose problems known to be associated with CD but without clear mechanisms or guidance for further investigation. In addition, they highlighted some issues that may not be well known: for example, the fact (and proposed mechanism) that those with CD may not mount immunity against hepatitis B vaccine[7] and thus should be routinely screened at diagnosis.

Studies clearly show that those with CD who have regular follow-up are more likely to remain adherent to a gluten-free diet and have a better long-term prognosis.[8,9] This article provides a useful roadmap to health care providers for the associated testing at diagnosis and during follow-up visits to monitor for comorbidities associated with the underlying disease, or as a consequence of the dietary restrictions, and to ensure the optimal health and well-being of the patient. These recommendations are supported by current data when available, and on those topics where few data exist, the consensus opinion of experts was used after vigorous debate. These recommendations will help guide the way in which we monitor our patients with CD and ultimately allow us to deliver better health care.

ABBREVIATIONS

CD: celiac disease
tTG-IgA: tissue transglutaminase immunoglobulinAantibody

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