Initial Assessment and Follow-up Care of Celiac Patients
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INITIAL ASSESSMENT

Dr. Green sees a lot of patients who, either through their own frustration or because of physician advice, have started a gluten-free diet without obtaining a biopsy-proven diagnosis of Celiac Disease. However, the need for a biopsy to establish a diagnosis of Celiac Disease must be emphasized. Celiac Disease is a lifelong illness with serious potential implications. In addition, sensitivity to gluten doesn't go away, and a radical lifestyle change is involved. You also need to be certain of the diagnosis because Celiac patients' families should be screened. The initial biopsy is also needed to serve as a baseline because one doesn't know what the future may involve.

Basic blood work is also included in the initial assessment. Such things as anemia and liver function need to be looked for. But it's very important to go further than that, and knowledge of the physiology of the small intestine should lead a physician to measure those nutrients that could be malabsorbed. **Celiac Disease involves the small intestine, where iron, folic acid, calcium, fat soluble vitamins (Vitamins K, A, D, and E) and zinc are absorbed**. **These nutrients should be measured in the initial assessment and also during the course of the illness**. Physicians will see patients who present with malabsorption of just one of these nutrients. If they are aware of the consequences of all these nutrient deficiencies, it will help them consider Celiac Disease as a possible diagnosis.

The patient should also have the Celiac antibodies blood testing, but the diagnosis is still established on the biopsy pathology. In Dr. Green's experience, **about 30% of Celiacs have negative antibodies at diagnosis; so positive antibodies are not required to make the diagnosis.** Antibodies testing often helps establish the need for a biopsy, but their great value is also in establishing a baseline so that an assessment can be made on how the patient is doing later on. All the antibodies should normalize, in time, when gluten is eliminated from the diet.

What about the patient who seeks a diagnosis, but has already eliminated gluten from the diet? It is very difficult for many patients to go back on a gluten-containing diet to secure a biopsy-proven diagnosis. This can often take three to six months or longer. Columbia-Presbyterian has been talking about setting up alternative means of securing a diagnosis, such as a rectal challenge. The physician can take a biopsy of rectal tissue, and then instill gliadin extract into the rectum and do a repeat biopsy a certain number of hours afterward to demonstrate an inflammatory response similar to that in the small bowel. However, interpreting the results of the gluten challenge would require a pathologist who is very experienced, and sophisticated immunology on the cells of the rectal biopsy may be needed.

FOLLOW-UP CARE

Soon after diagnosis and adhering to a gluten-free diet, patients will often report an increased feeling of well-being. How well they feel - and how quickly -will also depend on what the manifestations of their disease were. For example, if the patient was iron deficient, it will take time for the iron stores to be repleted.

An assessment of vitamin and mineral levels should be part of the follow-up care. Specific deficiencies need to be addressed, treated, and monitored. Patients have been seen who have been ingesting too much of the fat-soluble vitamins, with resulting problems such as liver disease (Vitamin A toxicity), and hypercalcemia (Vitamin D toxicity), which can cause confusion, constipation, and kidney problems. Certain vitamins and minerals may need to be administered, but the patient should be under a physician's guidance as to how much should be taken.

After a diagnosis of Celiac Disease, a bone mineral density should be performed to assess the condition of the bones. Reports have shown that between 50-100% of people at initial diagnosis of CD will have osteopenia or osteoporosis. Ostopenia is thinner bones, usually less than 2 standard deviations from normal. Osteoporosis involves an even greater deviation from normal. In Dr. Green's experience, nearly 100% of the Celiac patients at diagnosis will have osteoporosis. Surveys of Celiac patients have shown an increased incidence of fractures prior to diagnosis and after diagnosis.

If the bone mineral density is low, the patient should be referred to a bone mineral expert for assessment and specific individual treatment. For example, calcium and Vitamin D needs will be addressed and monitored, and exercise and hormone replacement (in post-menopausal women) will be considered.

At diagnosis, patients should get a Pneumovax, because it is very common for Celiacs to have poor splenic function, which puts them at risk of developing certain bacterial infections such as pneumoccal pneumonia and meningitis.

Since there is a genetic predisposition to Celiac Disease, another important issue in the follow-up is screening family members for CD. Children and other first-degree relatives should have their antibodies status measured. About 10-15% of first-degree relatives have positive antibodies, and the bulk of the people with positive antibodies will have the disease, even though 50% of those people will be asymptomatic, even with a flat biopsy.

What annual follow-up care should the Celiac patient be getting? The most important thing is a good physical examination. Blood work, x-rays, CAT scans, mammograms and PSA tests, while valuable, do not replace a physical examination. The physical exam should include a breast exam for women, prostate exam for men, and a rectal exam for everyone. **Blood work should include measurements of folic acid, calcium, and iron, and antibodies testing**, Bone mineral density testing should be repeated annually for those with abnormal results, and every several years for those with normal results.

Finally, patients with Celiac Disease should have at least one follow-up biopsy to confirm response - normalization of the biopsy sample. Patients who are non-responders, or whose clinical situation is somewhat confusing, may need more repeated biopsies at intervals.

NON-RESPONDERS

What about the non-responders or people who relapse? The first thing is to check the diet with antibodies testing. People may be ingesting gluten, such as in medications, and not be aware of it. They may be getting gluten from licking stamps or envelopes. They may have misinformation from food labels or manufacturers.

However, the antibodies can normalize and the biopsy still look quite flat, so once again, the antibodies have only a limited value - but they are still important to measure. It is also important to check the original biopsy to make sure of the diagnosis. Not all pathologists are experienced enough to properly diagnose Celiac Disease. Pathology departments, by law, have to keep the biopsies for a lengthy period of time - some keep them for 50 years. So it is important for the physician to review the biopsy sample with a pathologist who understands the spectrum of Celiac Disease. The pathologist needs to know, for example, how to identify latent Celiac Disease and different subtle aspects of the biopsy, such as increased intraepithelial lymphocytes.

A problem that comes up in non-responders is other food sensitivities. It's very rare for people with CD to also have sensitivities to other foods that result in the abnormal biopsy. There are, however, reports of ingestion of soy protein or egg or some kind of meats that cause the biopsy not to normalize.

There are other conditions that can co-exist with Celiac Disease and confuse physicians. For example, pancreatic insufficiency can cause diarrhea, steathorrhea, and malabsorption of fat, and bacterial overgrowth can affect absorption of nutrients. Patients may have colonic pathology. Having one disease doesn't mean you can't have another disease, and other conditions need to be investigated in the CD patient who isn't doing well.

If there is no improvement in the biopsies, patients remain at the risk of developing or maintaining bone disease and vitamin deficiencies, and they are at a higher risk for malignancy. Patients who are refractory may need other therapies such as corticosteroids or immunosuppressent drugs such as cyclosporin. One doesn't engage these therapies lightly (for example, steroids will thin the bones); being closely evaluated while on these drugs is important.

PROGNOSIS FOR THE CELIAC PATIENT

The studies that have indicated increased mortality in CD are from other countries where people have different smoking and dietary habits. It is hard to extrapolate these studies to our patient population.

The existing studies indicate that the mortality rate among adult Celiacs is about two to three times that of the general population, and the increased mortality is found mainly in the first five years after diagnosis. After that, the mortality rate approaches that of the normal population. That tends to suggest that it is the continued ingestion of gluten that is responsible for the increased mortality. This is especially so in regard to malignancies, where the risk of diagnosis of malignancy such as lymphoma is usually highest in the first year after diagnosis, and then decreases in instance downward until it equals that of the normal population after about five years. There is certainly the suggestion that adhering to a gluten-free diet reduces the risk of developing a malignancy.

A FINAL WORD - LOOKING FOR CELIAC DISEASE

Traditionally, the incidence of Celiac Disease in this country, based upon epidemiological work, suggests that CD occurs in about 1 in 4,600 people. Certainly it's much more common that that. Serology testing of blood donors by Dr. Fasano suggests the same prevalence as in European countries, about 1 in 300 people. Dr. Green, who does a lot of endoscopies, has found an incidence of Celiac Disease in about 1 in 280 patients who were having endoscopies for reasons other than suspicion of Celiac Disease. **It is important, therefore, for the gastroenterologist to have a higher suspicion for the possibility of Celiac Disease, and for physicians to screen for Celiac Disease, particularly among their patients who have associated diseases such as Insulin Dependent Diabetes, Sjogren's, and Autoimmune Thyroid Disease.**

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