

Eosinophilic Esophagitis Is Enigmatic in Adults

BY BETSY BATES

Los Angeles Bureau

LOS ANGELES — Eosinophilic esophagitis appears to be a different disease in adults than it is in children, sharing similar pathophysiologic features and perhaps an allergic etiology, but displaying a different pattern of symptoms, Dr. David A. Katzka said at the annual Digestive Disease Week.

"Certainly this is a new kid on the block, with many of us feeling this is a totally new disease," said Dr. Katzka, director of the swallowing program at the University of Pennsylvania in Philadelphia.

It is generally agreed that eosinophilia is on the rise. Children with eosinophilic esophagitis complain of a "plethora" of symptoms, including nausea and vomiting, epigastric pain, heartburn, and dysphagia. "In adults, by far and away, the most dominant symptom is dysphagia," Dr. Katzka stressed. "It may be intermit-

tent. It may be catastrophic," he said, noting that it has become a highly prevalent cause of food impaction, despite the fact the disease was only described in 1993.

At first believed to be a variant of gastroesophageal reflux disease (GERD), the disease is clearly a distinct entity with a genetic component in some families. On the other hand, many adults with the disease have GERD symptoms and some respond to aggressive acid suppression with proton pump inhibitors, making the connection between the two entities vexing.

In both children and adults, the diagnosis is pathologic and based on an ill-defined elevated rate of eosinophils found in a patchy pattern in the esophageal mucosa.

He emphasized the importance of taking multiple biopsies, since there may be 5 eosinophils per high-powered field in one spot, and "50 in another."

Longitudinal furrows are very common findings on endoscopy in all age groups.

In addition, rings throughout the esoph-

agus and strictures are commonly seen in adults (and less often, in children) even on radiographic films.

Eosinophilic abscesses, visualized as white specks, are seen in 17%-20% of children with the disease and are "almost pathognomonic" in adults. The esophagus may have a "firm, woody feel," he said.

Debate rages as to whether a person with a normal-appearing esophagus can have the disease. Dr. Katzka said he believes it is possible.

The peripheral eosinophil count is normal in about 90% of patients.

Although studies have not been done in adults to direct management, Dr. Katzka recommends RAST testing, patch testing, and skin testing to try to identify an allergen or combination of allergens that may be responsible. However, he warned of an "imprecise correlation between skin, blood, and esophageal findings" and said some adult patients do not respond to avoidance of known allergens.

The biggest problem is convincing patients to avoid foods that may be contributing to the condition.

In children, treatment with steroids, leukotriene inhibitors, and mast cell stabilizers have been shown effective.

In adults, "we're flying by the seat of our pants" in regard to treatment, he said.

He recommends a 2-month course of fluticasone propionate and possibly, maintenance with montelukast, noting that some specialists also suggest a 1- to 2-month course of proton pump inhibitors prior to performing a second endoscopy.

"We will treat these patients very aggressively for 2 months before thinking about dilation," he said.

In children, eosinophilic esophagitis seems to stabilize and improve over time, while in adults, limited studies suggest it persists or worsens.

"One of our fears is that in adults, this is a progressive or static disease that has to be recognized and treated early." ■

Mouth Ulcers and Defects in Tooth Enamel May Suggest Celiac Disease

NEW YORK — Dental enamel defects and aphthous ulcers are both strongly associated with celiac disease, and their presence should be followed up with a full investigation for the disorder in undiagnosed people, Theologos Malahias, D.D.S., suggested at an international symposium on celiac disease.

Dental enamel defects that are seen in permanent teeth form when the teeth first develop, and thus will not reverse when celiac patients are treated with a gluten-free diet, said Dr. Malahias, a dentist from Groton, Conn.

"For example, the 6-year molars—also known as adult permanent first molars—start forming enamel when a baby is 3-4 months old," he said in an interview. "So if celiac disease is present at that time, it can affect the enamel, but you won't see the defect until the tooth erupts at 6 years old." Patients with these problems also may experience a delay in the eruption of permanent teeth, he added.

The bilateral, symmetrical markings are most commonly seen on central incisors and molars and are evident in all four quadrants of the mouth, Dr. Malahias explained. They are opaque and can be white, yellow, or brown, causing the enamel to look mottled, and not shiny.

"There are other causes for this type of dental appearance, so I always stress that celiac disease should be considered in the context of the rest of the patient's medical history," he said.

His study of 136 patients, 67 with celiac disease and 69 without, found dental enamel defects in 51% of the celiac cohort and in 30% of the nonceliac group. Among the pediatric

subgroup of 47 children aged 6-16 years, 87% of the 23 celiac patients had enamel defects, compared with 33% of the 24 who did not have celiac disease. In children younger than age 12 with mixed dentition, the enamel defect rate was 90% among those with celiac disease and 44% among the others.

The rate of dental decay was similar for children with and without celiac disease, but was higher in the 44 adult celiac patients than in the 45 adults without celiac disease, he reported. Decay rates need to be studied further because other studies have not found an increased decay rate in adult celiac patients, he said.

According to Dr. John Zone, a dermatologist from the University of Utah, Salt Lake City, who also spoke at the meeting, about 5% of people with unexplained aphthous ulcers have occult celiac disease. "They are presumably due to chronic stimulation of the immune system by gluten," he suggested.

The ulcers continue to occur, but less frequently, after celiac patients start a gluten-free diet, Dr. Malahias noted.

—Kate Johnson



If celiac disease is present when adult teeth are forming, it can affect the enamel.

COURTESY DR. THEOLOGOS MALAHIAS

Textbook Celiac Disease Occurs Less Often, Eluding Diagnosis

BY KATE JOHNSON

Montreal Bureau

NEW YORK — Celiac disease doesn't look like it used to, and consequently the diagnosis is frequently missed by physicians who think of it as a disease of wasting, reported experts at an international symposium on celiac disease.

Traditionally, textbook pictures of celiac patients have shown the wasted limbs and swollen abdomens characteristic of malnutrition—but today, up to 40% of patients are overweight and 13% are obese at presentation, according to Dr. William Dickey of Altnagelvin Hospital, Londonderry, Northern Ireland.

Dr. Dickey's study of body mass index at presentation in 371 celiac patients over a 10-year period found that only 5% were underweight, and few presented with the classic symptoms of diarrhea and anemia (*Am. J. Gastroenterol.* 2006;101:2356-9). The well-documented problem of missed or delayed diagnosis of celiac disease may be partially explained by physicians' failure to recognize its modern presentation, he suggested.

In response to physicians' "significant lack of awareness and very substantial underdiagnosis of patients," the National Institutes of Health has embarked on an awareness campaign aimed mainly at primary care providers, said Dr. Stephen James, director of the digestive diseases and nutrition division of the NIH, who also spoke at the meeting.

"Physicians do not perceive the underdiagnosis of celiac to be problematic or to have long-term consequences," he said, citing findings from the recent NIH Consensus Development Conference on Celiac Disease (www.consensus.nih.gov/2004/2004CeliacDisease118html.htm).

Although celiac disease was once regarded as a disorder found only in people of European descent, recent evidence suggests

that its prevalence—about 1%—is the same in both Europe and North America (*Arch. Intern. Med.* 2003;163:286-92). Higher prevalence rates are seen in first-degree relatives (5%-15%), monozygotic twins (70%-75%), and patients with other autoimmune disorders.

The prevalence of celiac disease is slightly higher than that of either type 1 diabetes or Crohn's disease, but many physicians still have the misperception that celiac disease is a rare disorder, Dr. James said. Although many primary care physicians surveyed said they had never seen a case of celiac disease, they probably had several patients in their practice who were undiagnosed, he said.

Extragastrintestinal signs and symptoms of celiac disease, such as dermatitis herpetiformis, neurologic disorders, osteoporosis, recurrent miscarriage, and dental and oral problems, often go unrecognized, he noted.

The ambiguity of diagnostic tests contributes to the elusiveness of celiac disease, once called a "clinical chameleon" by Dr. Alessio Fasano, a leading expert from the University of Maryland Medical Center, Baltimore. In addition to the nonspecific spectrum of clinical symptoms, diagnostic criteria have hinged on positive serum antibodies and small bowel biopsies, but equivocal serologic and histologic results are frequently found, experts agreed.

The combination of positive serology followed by a positive bowel biopsy and response to a gluten-free diet confirms the disease with roughly 95% accuracy, said Dr. Edward J. Hoffenberg, of the University of Colorado Health Sciences Center in Denver. But his work, and that of others, has shown that an individual's degree of immune response can fluctuate, especially if they are not ingesting gluten at the time of testing, and the characteristic villous atrophy seen on bowel biopsies is not always present. ■