

Improved Scoring System for the Food Allergic Disease Eosinophilic Esophagitis in Children

CINCINNATI - Researchers have developed and validated a histologic scoring system to objectively analyze pathologic features of eosinophilic esophagitis (EoE), a severe, often painful food allergy that renders children unable to eat a wide variety of foods.

This study, published online on February 9 in *Diseases of the Esophagus*, was led by researchers at Cincinnati Children's Hospital Medical Center.

Eosinophils are normal cellular components of the blood, but when the body produces too many eosinophils they can cause a variety of eosinophilic disorders, such as EoE. These are disorders involving chronic symptoms and resulting in tissue damage, often in the gastrointestinal system, such as the esophagus.

The diagnosis of EoE is currently made using the peak esophageal eosinophil count coupled with specific clinical symptoms and a lack of response to proton pump inhibitor therapy. Greater than or equal to 15 intraepithelial eosinophils in a least one high-power microscopic field of an esophageal biopsy signifies active disease. However, using peak eosinophil count as the only pathologic diagnostic feature is limiting. For instance, rereviewing slides with peak eosinophil counts of 1-14 eosinophils yields counts at or above the diagnostic threshold in about 20% of these biopsies, according to the research team.

“This scoring system encourages pathologists to evaluate more than eosinophilic inflammation, and reduces our dependence on a single feature for diagnosis,” says Margaret H. Collins, MD, professor of Pathology and Laboratory Medicine at the University of Cincinnati, member of the Cincinnati Center for Eosinophilic Disorders at Cincinnati Children's and first author of the study. “By having validated tools to measure other pathologic features of eosinophilic esophagitis, our reported findings provide a new opportunity for evaluating disease activity and treatments going forward.”

The histology scoring system (HSS) that the researchers developed for EoE evaluates eight features: eosinophil density, basal zone hyperplasia, eosinophil abscesses, eosinophil surface layering, dilated intercellular spaces (DIS), surface epithelial alteration, dyskeratotic epithelial cells, and lamina propria fibrosis. The authors of this study analyzed esophageal biopsies of pediatric patients with EoE with the HSS, scoring the severity (grade) and extent (stage) of abnormalities using a 4-point scale (0 normal; 3 maximum change).

“This histology scoring system puts numbers to previously qualitative observations of histologic features,” says Marc E. Rothenberg, MD, PhD, Director of the Cincinnati Center for Eosinophilic Disorders and Division of Allergy and Immunology at Cincinnati Children's. “It is a nice step forward in assessing eosinophilic esophagitis as the complex disease entity that it is.”

The study demonstrated that HSS is reliable and could differentiate between treated and untreated patients, with untreated patients having higher scores. Notably, the HSS can be used by pathologists after minimal training, which would aid timely diagnosis of this often misunderstood disease.

This study is the first to validate the HSS. This work is an essential step toward improving the outcomes of patients with eosinophilic esophagitis. With the HSS, researchers can better evaluate esophageal

biopsies for features in addition to peak eosinophil count, and this instrument can be used to test the effect and benefit of new therapies.

Allergic diseases have been on the rise over the past 20 years, with approximately one of every 13 children having food allergies and over 2.5 million children suffering from allergic asthma. Only recently recognized as a distinct condition, the incidence of EoE has also been increasing. Rothenberg and his laboratory team pioneered research showing that EoE has a reported incidence estimated to be approximately 1 out of 1,000 people, is caused by a combination of genetic and environmental factors, and is primarily mediated by an immunologic response to foods. The hallmark of EoE is swelling and inflammation in the esophagus, accompanied by high levels of immune cells called eosinophils.

EoE can affect people of any age but is more common among young men who have a history of other allergic diseases, such as asthma and eczema. EoE is often first discovered in children with feeding difficulties and failure to thrive, but it is often misunderstood and not well known, delaying proper diagnosis and treatment.

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