

Eosinophilic Esophagitis

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Eosinophilic esophagitis (EoE) is a chronic allergic inflammatory disease of the esophagus that affects children and adults. Typical symptoms of EoE typically involve failure to thrive and vomiting in small children, and dysphagia and food impaction in adolescents and adults. However, throat clearing, choking, gagging, and hoarseness have also been observed and these issues often bring patients to first seek

otolaryngology and pulmonary evaluation. Diagnosis is only made by esophagogastroduodenoscopy with biopsy, so clinicians must have a low threshold for considering eosinophilic esophagitis even in the absence of frank gastrointestinal symptoms.

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Eosinophilic esophagitis (EoE) is an allergic esophagitis in which exposure to food and environmental antigens causes eosinophil predominate inflammation of the esophagus. It was first described in 1995 when Kelly et al¹ described a cohort of 10 children with eosinophilic infiltration of the esophagus that was refractory to typical therapies including antacids, prokinetic agents, and Nissen funduplications. All patients had resolution of inflammation and symptoms with implementation of a diet consisting of only amino acid based formula. Subsequently, Liacouras et al² showed that a similar cohort of patients had reduced inflammation with oral prednisone therapy for a period of 6 weeks. Unfortunately, both cohorts of patients had relapse of disease after termination of therapy.

Over the last 20 years, a tremendous amount of research progress has been made toward understanding some of the pathobiological and epidemiologic aspects of this disease. Currently, large database analysis has shown that the prevalence of EoE is 56.7/100,000.³ The majority of EoE patients are male (72%), with an overwhelming predominance of Caucasian (83%) followed by African American (10%).⁴ Patients with EoE are more likely to have asthma, eczema, IgE-mediated food allergy, chronic rhinitis, and environmental allergies.⁵ In fact, recent studies show that patients with EoE may have some

degree of increased airway hyperresponsiveness compared to normal subjects.⁶

The symptoms of EoE vary greatly, making diagnosis challenging. Patients can have general symptoms of GERD including regurgitation, heartburn, stomach pain, and vomiting. Others will have more frank dysphagia reporting feelings of globus, delayed transit of food, gagging on food, and feelings of food getting stuck. One common presenting symptom is food bolus impaction with patients requiring emergent endoscopic removal.⁴ EoE is diagnosed in both pediatric and adult populations however, the symptomatology of the pediatric and adult populations varies greatly with children presenting with feeding refusal, vomiting, and failure to thrive and adolescent/adult patients presenting with dysphagia and food impaction.⁷ This paradox seems to be related to duration of disease prior to diagnosis⁸ with adults having presenting after years of unchecked inflammation. During maturation, many pediatric and adult patients develop coping mechanisms that cause, on average, a 6-year delay in diagnosis from the onset of symptoms. These coping mechanisms include increase fluid intake during meals to try to lubricate food boluses, extended meal times with careful chewing and swallowing, and avoidance of tough meats such as steak and chicken.

Due in large part to this broad symptomatology, patients often present to otolaryngologists and pulmonologists prior to gastroenterologists and allergists. In two separate retrospective studies, approximately 4% of patients who presented for care at tertiary pediatric multispecialty aerodigestive centers were found to

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have EoE after endoscopic evaluation. Over 40% of these patients presented with cough as their predominant symptom. Other common symptoms included choking, throat clearing, and hoarseness.^{9,10} Of patients with EoE, up to 18% of patients presented first to otolaryngology with symptoms including stridor, cough, croup, or dysphonia.¹¹ Gastrointestinal symptoms are not always necessary in EoE. Prospective evaluation of patients presenting to a tertiary care center with primary respiratory symptoms of cough and wheeze determined that 8% of patients had EoE¹² even in the absence of frank gastrointestinal complaints.

At this time the only way to diagnosis EoE is by esophagogastroduodenoscopy (EGD). Because acid-related esophagitis causes eosinophilia of the esophagus, patients must be adequately treated for GERD with twice daily proton pump therapy for at least 6 weeks prior to endoscopy.⁵ The diagnosis of EoE is made when biopsies reveal >15 eosinophils in one high-powered field. Other inflammatory findings in the esophagus are common including basal cell hyperplasia, elongated papilla, and dilated intracellular spaces.^{5,13} EoE is a patchy disease, and eosinophil count can vary from one part of the esophagus to another. In order to accurately diagnosis EoE and decrease the risk of falsely negative biopsies, it is recommended to obtain at least 4–6 biopsies from the esophagus. Furthermore, endoscopic appearance in pediatrics can be quite variable ranging from normal appearance to furrows or rings. Normal endoscopy findings should not deter the clinician from obtaining adequate number of biopsies if EoE is a consideration.

Therapy for EoE involves a multidisciplinary approach with allergic and gastrointestinal specialist involvement. Possible therapies include diet-directed therapy and topical steroids.¹⁴ Diet-directed therapy involves empiric or allergy testing directed elimination of possible offending food antigens.^{15,16} The only reliable way to determine if a food is causing EoE is to perform subsequent endoscopies after food elimination and reintroduction. Steroid therapy involves topical preparations of budesonide or fluticasone. Both have both been used with success in EoE. This allows patients to maintain a regular diet with minimal effects on the adrenocortical axis.¹⁷ Both diet

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elimination and topical steroids induce remission, but some children will continue to have ongoing feeding issues with prominent choking and gagging despite improved inflammation. Patients often develop aversions to textures and may require formal feeding therapy to help reintroduce these textures even after medical treatment.¹⁸

In summary, the heterogeneous presentation of EoE makes it diagnostically challenging for gastroenterologists, pulmonologists, and otolaryngologist alike. Clinicians must have a low index of suspicion and consider this entity even in the setting of isolated respiratory illness.

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