

Eosinophilic Esophagitis

Clinical Presentation in Children

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KEYWORDS

• Eosinophil • Esophagitis • EoE • Children • Pediatric

KEY POINTS

- Symptoms of eosinophilic esophagitis (EoE) vary by age. They range from failure to thrive in toddlers, to abdominal pain in school age children, to dysphagia in adolescents.
- Symptom overlap exists between EoE and other disorders, including gastroesophageal reflux disease, asthma, and primary eating disorders.
- Diagnosis is made by clinical history combined with results of upper endoscopy with biopsy.

INTRODUCTION

Our understanding of eosinophilic esophagitis (EoE) has evolved over the past 30 years from isolated case reports of patients with prominent esophageal eosinophilia (often misclassified as gastroesophageal reflux [GERD]) to a well-defined clinical disorder. In the past, EoE was described differently, including allergic esophagitis, primary EoE, or idiopathic EoE. We now know that EoE is a distinct disease. It is defined as a clinicopathologic diagnosis characterized by a localized eosinophilic inflammation of the esophagus (with no other gastrointestinal involvement), symptoms of esophageal dysfunction, the presence of 15 or more eosinophils in the most severely involved high-powered field (HPF) isolated to the esophagus, and failure to respond to adequate proton-pump inhibitor (PPI) therapy. Other recognized causes of esophageal eosinophilia should be excluded before making the diagnosis (**Box 1**).

DEMOGRAPHICS

EoE is characterized by eosinophilia of the esophagus, an organ typically devoid of eosinophils, without infiltration in other parts of the gastrointestinal tract.^{1,2} First

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Box 1
Causes of esophageal eosinophilia

- EoE
- GERD
- PPI-responsive esophageal eosinophilia
- Celiac disease
- Eosinophilic gastroenteritis
- Crohn disease
- Hypereosinophilic syndrome
- Achalasia
- Vasculitis
- Pemphigus
- Connective tissue disease
- Infection
- Graft-versus-host disease

described in 1993, EoE has been increasing in incidence and prevalence in Western nations with an estimated annual incidence equal to Crohn disease.^{3–8} The increase in EoE mirrors the increased prevalence of allergic diseases (asthma, allergic rhinitis, atopic dermatitis, and food allergy) over the last few decades. Data from the CDC National Health Information Survey (NHIS) confirm the increase in all atopic diseases.^{9,10} The reported prevalence of asthma increased from 3% in 1990% to 7.7% in 2007.¹¹ An estimated 25% to 30% of the population in industrialized countries has atopic dermatitis, food allergy, or allergic rhinitis.¹² EoE has now been reported in every continent.^{12–15} Five to 10% of pediatric patients and 6% of adult patients with poorly controlled GERD are thought to have EoE.^{16–18} In our cohort at The Children’s Hospital of Philadelphia, the authors have seen a 70-fold increase, from 1994 to 2011.¹⁹ In Olten Country, Switzerland, this increase has also been observed in the adult population with increased prevalence from 2 per 100,000 in 1989 to 23 per 100,000 in 2004.⁵ Patients who have EoE typically are male (by a 3:1 ratio)^{8,19} and 75% have a personal history of atopy.^{19–22} Like other atopic diseases, such as asthma and eczema, EoE is a chronic disease. Most patients will continue to have the disease into adulthood. In a 12-year study of adults, no patients had remission.²³ In a 14-year study of pediatric patients, only 2% had remission of disease.¹⁹

CLINICAL PRESENTATION: GENERAL CONSIDERATIONS

EoE should be suspected in patients describing symptoms consistent with GERD but not responding to adequate reflux medications. Presentation in children varies depending on the age of the child. Characteristic symptoms in infants and young children are feeding difficulties, failure to thrive, and classic GERD symptoms. In contrast, school-aged children are more likely to present with vomiting, abdominal pain, and regurgitation. Dysphagia and food impaction are more prevalent in adolescents and adults.^{1,19,20,22,24–26} One study looked at age of presenting symptoms over a 14-year span and found feeding difficulties or failure to thrive in young children (median age 2.8 years), vomiting and GERD symptoms in older children (median age

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