Eosinophilic Esophagitis in Children and Adults

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KEYWORDS

• Eosinophilic esophagitis • Children • Adults • Clinical features • Treatment
• Diet therapy • Dilation

KEY POINTS

- Adults and children with eosinophilic esophagitis (EoE) have distinct clinical and endoscopic presentations.
- Clinical presentation depends greatly on patient age.
- Treatments for EoE are effective across all age groups but used differently among pediatric and adult specialists.

INTRODUCTION

Eosinophilic esophagitis (EoE) is a clinicohistologic diagnosis requiring both esophageal dysfunction and esophageal mucosal biopsies that demonstrate eosinophilia, without an alternative cause.[1] EoE is a prevalent cause of esophageal dysfunction in both children and adults.

Rarely described before 1993, EoE is the most prevalent eosinophilic gastrointestinal illness. It is the second major cause of esophagitis after gastroesophageal reflux disease (GERD) and EoE is the dominant cause of food impaction in young patients.[2] Initially, EoE was thought to be primarily a pediatric illness but later it was recognized in adults.

At first, esophageal eosinophilia was thought to be associated with reflux.[3–7] After the first multidisciplinary conference on EoE in Orlando, Florida, in 2005, EoE was recognized with increased frequency as a common cause of chronic dysphagia and esophageal symptoms in both children and adults.[7,8]

EoE affects all ages; clinical presentation depends greatly on the patient's ability to report symptoms associated with esophageal dysfunction. Recognition of clinical

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signs, along with laboratory and endoscopic findings, is critical for the identification of patients with EoE because delay in diagnosis has been associated with esophageal remodeling and stricture formation.[9–11]

This article describes the similarities and differences in clinical presentation of children and adults with EoE, including areas of epidemiology, clinical and endoscopic presentation, pathophysiology, and treatment.

EPIDEMIOLOGIC ASPECTS OF EOSINOPHILIC ESOPHAGITIS

EoE was likely first described in the adult population as anecdotal cases of small-caliber esophagus found radiographically. However, the first identification of EoE as a separate and distinct disease entity began when Kelly and colleagues[12] identified a cohort of pediatric subjects with refractory esophagitis, characterized by eos, who failed to respond to acid suppression but went into remission when fed an elemental diet. From that point forward, esophagitis characterized by eosinophil (eos)-predominant inflammation has been studied extensively in both the pediatric and adult populations.

The first population-based studies of EoE in children were published in the early 2000s, with numerous additional cohort-based studies that have provided insight into the clinical features of patients with EoE, as well as population-based metrics. EoE has been described in people living in North America, South America, Europe, Asia, and Australia. EoE seems to be more prevalent among developed countries with higher socioeconomic development. However, within these countries, there is variation in prevalence rates, suggesting additional factors, such as climate, urban versus rural environment, and diet. An analysis of an esophageal biopsy database demonstrated a higher prevalence of EoE in cold climate zones of the United States.[13]

EoE has consistently demonstrated a strong male predominance in both adults and children, with a male to female ratio usually between 2.5:1 and 3:1. EoE has been diagnosed in every age group, ranging from infants to the elderly. In the pediatric population, the average age of diagnosis is typically between 6 and 10 years of age. Traditionally, there was a relatively long time lag before diagnosis (approximately 4 years), although this may be decreasing as awareness of the disease increases, not just among specialists who confirm the diagnosis but among primary care providers who make referrals.

Noel and colleagues[14] published the first population-based results on pediatric EoE in 2004. Evaluating a pathology database for histologic cases of isolated, esophageal eosinophilia (>24 eos per microscopic high-power field [HPF]), 315 cases of possible EoE were identified between 1991 and 2003. Almost all of those patients (~97%) were found after the year 2000, providing evidence that the disease was undergoing a significant increase in incidence. Further, in the 4-year period from 2000 to 2003, the incidence increased 4-fold (from 1 case to 4.3 cases per 10,000).

The largest detailed pediatric cohort with EoE was described in 2009 by Spergel and colleagues.[15] In this group of subjects followed at the Children's Hospital of Philadelphia, there was a steady increase in new diagnoses each year from 1996 to 2006. Excluding the patients referred from outside of their local area (PA, NJ, and DE) did not affect the pattern of newly diagnosed patients, suggesting referral bias was not a factor in the increase. The average age of diagnosis in this group was 6 years, with more than one-third of the patients younger than age 3 years at the time of diagnosis. Environmental allergies were thought to be a contributor to EoE findings in about 10% of the patients based on seasonal variation in biopsy findings without changes in diet or medical therapy.