

# Role of Endoscopy in Diagnosis and Management of Pediatric Eosinophilic Esophagitis



Amanda B. Muir, MD<sup>a,b,1</sup>, Jamie Merves, MD<sup>a,b,1</sup>,  
Chris A. Liacouras, MD<sup>a,b,\*</sup>

## KEYWORDS

• Eosinophilic esophagitis • Dilation • Food impaction • Food allergy • Dysphagia

## KEY POINTS

- Endoscopy is currently the only way to diagnose eosinophilic esophagitis (EoE) and monitor disease activity.
- Food impaction occurs frequently in undiagnosed or chronic EoE, especially in the adult population, and may result in emergent endoscopic disimpaction.
- Stricture dilation in EoE may relieve symptoms of dysphagia but does not attenuate underlying inflammation.
- Endoscopic tools are being studied to find additional invasive biomarkers of disease activity.
- Less invasive techniques to diagnose and evaluate disease activity are currently being investigated.

## INTRODUCTION

Eosinophilic esophagitis (EoE) is a chronic immune-mediated and antigen-mediated disorder characterized by an isolated eosinophilic infiltration of the esophagus resulting in esophageal dysfunction.<sup>1</sup> Although there are many causes of esophageal eosinophilia, EoE must be primarily distinguished from gastroesophageal reflux disease (GERD) and proton-pump inhibitor (PPI)-responsive esophageal eosinophilia

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<sup>a</sup> Division of Gastroenterology, Hepatology, and Nutrition, The Children's Hospital of Philadelphia, 34th and Civic Center Boulevard, Philadelphia, PA 19104, USA; <sup>b</sup> Department of Pediatrics, Perelman School of Medicine, University of Pennsylvania, Philadelphia, PA 19104, USA

<sup>1</sup> Equal contributions.

\* Corresponding author. The Children's Hospital of Philadelphia, 34th and Civic Center Boulevard, Philadelphia, PA 19104.

E-mail address: [Liacouras@email.chop.edu](mailto:Liacouras@email.chop.edu)

(PPI-REE). EoE is an emerging disorder associated with other allergic conditions<sup>2</sup> and currently affects approximately 56.7 per 100,000 people in the United States.<sup>3</sup> EoE pathophysiology is complex, involving a variety inflammatory cells and cytokines in a non-immunoglobulin E (IgE)-dependent allergic model.<sup>4-6</sup> EoE is primarily caused by the ingestion of one or more food antigens,<sup>7,8</sup> but may also be triggered by inhaled aeroantigens.<sup>9</sup> EoE and associated complications have a significant impact on patient quality of life<sup>10</sup> and patients with EoE in the United States have an annual health care cost of up to \$1.4 billion.<sup>11</sup> EoE therapy is limited to dietary modification, steroids, and endoscopic dilatation.<sup>1</sup> At present, esophagogastroduodenoscopy (EGD) is the gold standard for diagnosis and disease monitoring.

## EOSINOPHILIC ESOPHAGITIS DIAGNOSIS AND SURVEILLANCE

EoE requires a clinicopathologic diagnosis. EGD is often performed in children for a variety of reasons, including chronic reflux symptoms with or without a poor response to acid suppression therapy, feeding problems, poor growth, intermittent or persistent vomiting and regurgitation, chest or epigastric abdominal pain, gastric or duodenal ulcers, dysphagia, a history of food impaction, or any other chronic indication of esophageal or gastric dysfunction. Because the current gold standard for the diagnosis of EoE is histologic evidence of esophageal eosinophilia, when performing an EGD it is paramount to perform biopsies, even in the presence of normal-looking mucosa.<sup>1</sup>

### *Endoscopic Findings*

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Although there are no pathognomonic findings for EoE, esophageal edema, longitudinal furrows, mucosal fragility, whitish exudates, transient esophageal rings (feline folds), fixed esophageal rings (trachealization), diffuse esophageal narrowing, and small-caliber esophagus are its typical macroscopic findings (**Fig. 1**).<sup>1</sup> Up to one-third of children with EoE may have visually normal esophageal mucosa.<sup>12</sup>

A novel endoscopic classification and grading system called the EoE Endoscopic Reference Score (EREFS) was recently developed and validated in adults to define common nomenclature, severity description, and disease assessment among providers.<sup>13</sup> The EREFS score is generated based on the presence and/or severity of transient or fixed esophageal rings, exudates, furrowing, mucosal fragility (so-called crepe-paper esophagus), edema (and the associated vascular pattern), as well as stricture.<sup>13</sup> Although this system has good interobserver agreement between practitioners, a validated scoring system in children has yet to be established.

There are many past and present differences in EoE diagnostic practices between pediatric and adult gastroenterologists. For example, adult gastroenterologists have traditionally based the diagnosis of esophageal disorders primarily on symptoms and endoscopic findings, rather than histopathology.<sup>14</sup> Alternatively, pediatric gastroenterologists have been trained to obtain mucosal biopsies in all diagnostic procedures, even if the mucosa is visually normal. The pediatric approach to diagnosis may be extremely important to identifying the presence of EoE, because it is common for patients of all ages to have a normal-appearing esophagus.<sup>12</sup>

### *Radiology*

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Although not recommended routinely for EoE diagnosis, imaging with upper gastrointestinal series (UGI) or esophagram should be recognized to be a useful test in patients with feeding problems, dysphagia, or food impaction to evaluate for anatomic and mucosal abnormalities such as narrowing, stricture, or formation of rings (**Fig. 2**).<sup>1,15,16</sup> In a cohort of 22 pediatric patients with EoE with strictures who

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