

## Paradigms and perspectives

# Eosinophilic gastrointestinal diseases make a name for themselves: A new consensus statement with updated nomenclature

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Eosinophilic gastrointestinal (GI) diseases (EGIDs) are characterized by tissue eosinophilia and symptoms of esophageal or GI dysfunction. Consensus recommendations for diagnosis of

eosinophilic esophagitis (EoE), the most common EGID, have been developed and updated since 2007.<sup>1-3</sup> The prevalence of non-EoE EGIDs is much lower, and currently, there are no consensus guidelines for diagnosis.<sup>4,5</sup> One of the first priorities for developing diagnostic guidelines is standardization of EGID nomenclature. Particularly problematic has been variable use of the term *eosinophilic gastroenteritis* to describe EGIDs involving the stomach, small intestine, large intestine, or groupings of these anatomic sites. Consistent terminology with greater specificity is critical to convey accurate clinical information and enhance data collection for research.

A large multidisciplinary group of researchers and clinicians<sup>6</sup> sought to achieve international consensus for EGID nomenclature by using an iterative process with formalized feedback using Delphi methods.<sup>7</sup> Major goals of the process focused on retaining useful terms, removing or deemphasizing ambiguous terms such as *eosinophilic gastroenteritis*, creating a basic and intuitive nomenclature useful for clinical practice, and including a more detailed nomenclature tier for research purposes. The authors administered a 42-question online survey that was followed by the consensus meeting. After incorporation of feedback, a second round of Delphi questions was administered through a subsequent 29-question online survey. The participants consisted primarily of experts in gastroenterology (70%), followed by experts in allergy/immunology (15%), and experts in pathology (6%) who were practicing in academic and/or university settings (91%). Both Delphi rounds achieved at least 90% participation among the 91 individuals invited to participate.

The Delphi process resulted in development of a new tiered framework for EGID nomenclature.<sup>6</sup> The guidelines recommend that *EGID* be used as the umbrella term for diseases involving pathologic eosinophil infiltration into any segment of the GI tract that occurs in the absence of secondary causes of eosinophilia (Fig 1, A). In routine clinical practice, physicians should utilize the first tier of the nomenclature to designate the GI tract location with active eosinophilic inflammation: esophageal involvement alone remains termed *EoE*, whereas any other location of involvement should be termed *non-EoE EGID*. Further delineation of the term *non-EoE EGID* can be completed by using the segment of the GI tract with disease, with involvement of the stomach termed *eosinophilic gastritis*, involvement of the small intestine termed *eosinophilic enteritis*, and involvement of the colon termed *eosinophilic colitis*. The second tier of the nomenclature provides additional granularity with naming EGIDs, particularly when considering small bowel involvement and when multiple overlapping nonesophageal locations have active disease. This tier can be used clinically, but it was developed to decrease ambiguity and heterogeneity in EGID terminology within the research setting.

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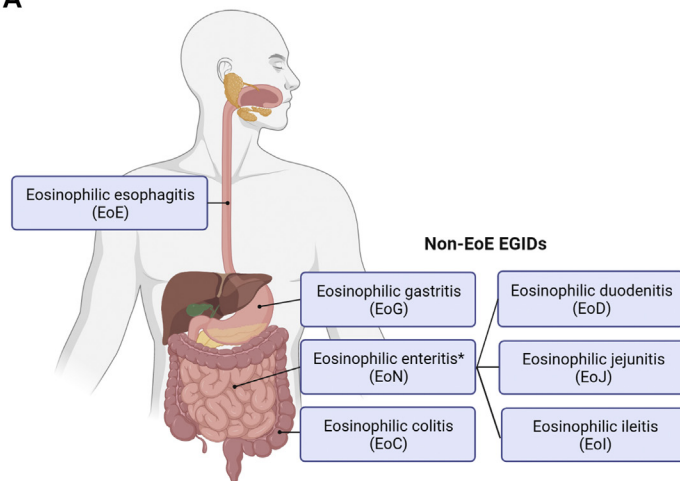
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## Eosinophilic Gastrointestinal Diseases (EGIDs)

A



## Nomenclature for Multiple Segment Involvement

B

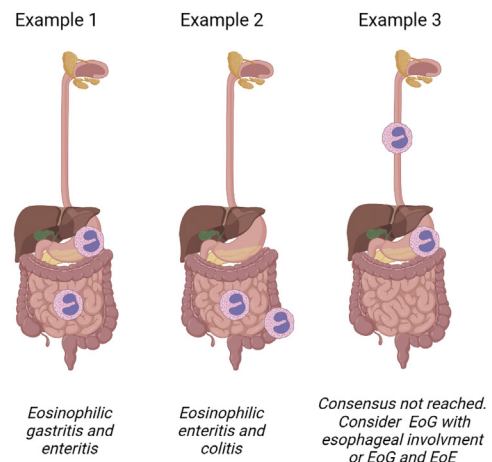


**FIG 1.** Schematic of the consensus nomenclature for EGIDs. *EGID* is recommended as the umbrella term for pathologic infiltration of the GI tract with eosinophils without secondary causes. **A**, Esophageal involvement alone is termed *EoE*, and any other location of involvement can be termed a *non-EoE EGID*. Individual involved areas of the GI tract should be named specifically, and an “Eo” abbreviation convention should be used in clinical and especially research settings. **B**, If the esophagus and stomach are both affected, use of the terms *eosinophilic gastritis with esophageal involvement* or *eosinophilic gastritis and EoE* should be considered. The term *eosinophilic gastroenteritis* should be used only to indicate both stomach and small bowel involvement. For non-EoE EGIDs, the recommendation is to indicate the GI wall layer involved when it is known. Figure created with [BioRender.com](https://www.biorender.com).

In this tier, small bowel involvement can be further differentiated into the specific segment involved (ie, eosinophilic duodenitis when the duodenum is involved, eosinophilic jejunitis when the jejunum is involved, and eosinophilic ileitis when the ileum is involved). Furthermore, when multiple nonesophageal locations are involved, multiple terms should be applied; for example, stomach and small bowel involvement should be termed *eosinophilic gastritis and enteritis* and small bowel and colon involvement should be termed *eosinophilic enteritis and colitis* (Fig 1, B).

There was considerable debate among the expert panel on how to best term multiple segment involvement, given insufficient data to determine whether more diffuse disease represents a disease spectrum with a shared pathogenesis. Many experts believed that it was important to identify a “primary” location of involvement by taking into account the histology, symptomatology, endoscopic features, and clinical complications (eg, protein-losing enteropathy, malabsorption, anorexia, weight loss). It was acknowledged that additional research is needed to refine this part of the nomenclature, which will likely evolve as future data emerge. No consensus was reached on how to designate involvement of multiple segments including the esophagus. The authors recommended that patients with lower GI and esophageal disease be designated by using the term for the lower GI involvement with the additional qualifier “with esophageal involvement” or “with EoE” (Fig 1, B). The Delphi process also demonstrated that even among the expert panel, use of the term *eosinophilic gastroenteritis* was variable. There was consensus agreement to deemphasize this term and redefine it to be used only in situations in which both stomach and small bowel are involved, although the preferred terminology would be *eosinophilic gastritis and enteritis*.

Revision of the EGID nomenclature stands to advance both clinical care for patients with EGID and EGID research. Regarding

clinical care, accurate nomenclature reflecting the affected areas of the GI tract is expected to help inform clinicians about potential symptoms and side effects that patients may experience more readily than a catch-all terminology would, and it may help to target therapies in the future. Additionally, specific nomenclature will help to limit diagnostic testing such as colonoscopy to that which is indicated and limit overinvestigation once the diagnosis has been made. With respect to research applications, precise naming of EGID will help to standardize studies. How EGID differs in immunologic and molecular phenotype and response to therapy based on those segments of the GI tract that are affected remains unclear. Precision in diagnostic nomenclature is the first step to establishing whether there are clinically significant differences in patients’ response to therapy based on the segment of the GI tract involved. Future directions for consensus diagnostic guidelines will likely require selection of specific disease markers and thresholds (eg, eosinophils/hpf), definition of symptoms of GI dysfunction, exclusion of alternative diagnoses resulting in GI eosinophilia, and (possibly) duration of disease with persistence of tissue pathology.

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