2011 Eosinophilic Esophagitis Updated Consensus Recommendations: Summary Statements

Below is a summary version of the 2011 Consensus Recommendations for EoE diagnosis and treatment.[1] Since 2007, the number of eosinophilic esophagitis (EoE) publications doubled, providing new disease insight. A panel of 33 physicians with expertise in pediatric and adult allergy/immunology, gastroenterology and pathology conducted a systematic review of the EoE literature (since September 2006) using electronic databases. Based on the literature review and expertise of the panel, a summary of the recommendations is provided here.

Conceptual Definition- To refine perceptions and hypotheses for future EoE studies, the following conceptual definition was developed: **"**Eosinophilic esophagitis represents a chronic, immune / antigen mediated, esophageal disease characterized clinically by symptoms related to esophageal dysfunction and histologically by eosinophil-predominant inflammation."

Diagnostic Guidelines- Taking into account increasing clinical experiences and research, the following guidelines were proposed: "Eosinophilic esophagitis is a clinico-pathological disease. Clinically, EoE is characterized by symptoms related to esophageal dysfunction. Pathologically, one or more biopsies must show eosinophil predominant inflammation.[+] With few exceptions, 15 eosinophils/hpf (peak value) is considered a minimum threshold for a diagnosis of EoE. The disease is isolated to the esophagus and other causes of esophageal eosinophilia should be excluded, specifically PPI-responsive esophageal eosinophilia.*(Table A). The disease should remit with treatments of dietary exclusion and / or topical corticosteroids. EoE should be diagnosed by clinicians taking into consideration all clinical and pathologic information; neither of these parameters should be interpreted in isolation.

[+]For optimal pathological evaluation, multiple biopsies from the proximal and distal esophagus should be obtained and evaluated for a variety of pathological features. Pathologists should report all abnormalities associated with EoE such as the peak eosinophil count (obtained from the area with the highest density of eosinophils,) eosinophilic microabscesses, surface layering of eosinophils, extracellular eosinophil granules, basal cell hyperplasia, dilated intercellular spaces, and lamina propria fibrosis. In a few circumstances, patients may have strong clinical evidence for EoE and have less than 15 eosinophils/hpf with other histological features indicative of eosinophilic inflammation.

*An emerging body of literature and clinical experience describes a subset of patients whose symptoms and histopathologic findings are responsive to PPI treatment and who may, or may not, have well documented gastroesophageal reflux disease (GERD). Until more is known, these patients should be diagnosed as "PPI-responsive esophageal eosinophilia."

Summary Statements

History and Physical- History should focus on difficulties with eating and swallowing (Table B) and a thorough physical examination should focus on growth and nutrition parameters and to assess potential other causes of esophagitis (Table A).

Endoscopy-Endoscopy with esophageal biopsy is considered the only reliable EoE diagnostic. Two to four biopsies each from the proximal and distal esophagus should be obtained. Gastric and duodenal biopsies should be examined to exclude other potential causes of eosinophil associated gastrointestinal disease. (Table A) Endoscopic features can suggest but cannot diagnose EoE.

Radiography-An upper gastrointestinal series is useful to characterize anatomic abnormalities that may escape endoscopic detection such as proximal strictures and long segment narrowing.

Histopathology- See Diagnostic Guidelines and Table C.

Allergic evaluation- An evaluation by an allergist or immunologist is recommended to document aeroallergen sensitization and seasonal variability as it may pertain to EoE and to control concurrent atopic diseases. Serum IgE and/or skin prick testing for immediate type hypersensitivity reactions to foods are warranted to help identify food allergic disease in patients with EoE. Medically supervised food reintroduction may be necessary for patients with previous allergic reactions to a food or IgE-mediated sensitivity documented by IgE testing. Skin prick tests, serum IgE tests, and food patch tests may be used to help identify foods that associated with EoE, but are *not* sufficient to make the diagnosis of food allergy driven EoE. Foods that trigger EoE can only be identified by documenting disease remission and recrudescence after specific food elimination and addition.

Genetics-EoE runs in families and although specific genes that pre-dispose to EoE susceptibility have been identified (TSLP, eotaxin-3), they are not yet ready for usage in clinical settings.

Treatments- (Table D)

Dietary therapy- Amino acid based formulas and dietary elimination are effective therapies for children with EoE and their use in adults requires further study. Patient's lifestyle, adherence to therapy and family resources should be considered when instituting these treatments. Consultation with a registered dietitian is strongly encouraged. EoE foods triggers may need to be restricted indefinitely.

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Steroids- Topical corticosteroids are effective therapy for EoE in children and adults. Systemic corticosteroids may be used for emergent situations (severe dysphagia, hospitalization, weight loss) but caution is warranted for chronic management of EoE.
Other treatments- Cromolyn sodium, leukotriene receptor antagonists, and immunosuppressives (azathioprine or 6-MP) are not recommended treatments for EoE. Biologic agents require further clinical studies and are currently not recommended for routine use.
Dilation- Esophageal dilation can provide relief of dysphagia in selected EoE patients. If high-grade esophageal stenosis is not present, a trial of medical or dietary therapy prior to esophageal dilation is reasonable.

Table A.[1] Conditions associated with *esophageal eosinophilia*

Gastroesophageal reflux disease (GERD)
Eosinophilic Esophagitis (EoE)
Eosinophilic gastrointestinal diseases (EGIDs)
Celiac disease
Crohn's disease
Infection
Hypereosinophilic syndrome (HES)
Achalasia
Drug hypersensitivity
Vasculitis
Pemphigoid Vegetans
Connective tissue disease
Graft versus host disease

Table B.[1] Symptoms Related to EoE

Dysphagia and Feeding dysfunction
Coping mechanisms -avoiding highly textured foods such as meats and bulky foods such as bagels, cutting food in small pieces, lubricating foods before eating with liquids or butter, extensive chewing of foods, washing food down with liquids, prolongation of mealtimes
Food impaction
Coping mechanisms - drinking liquid to wash food down, raising hands above head, jumping up and down, waiting for food to dissolve or to pass
Chest pain
Coping mechanisms-avoiding foods or liquids that exacerbate pain such as highly textured or bulky foods, alcohol or acidic drinks
GERD like symptoms recalcitrant to medical and surgical GERD management
Abdominal pain
Vomiting
Anorexia and early satiety

Table C.[1] Histological Characteristics of EoE

Mucosal eosinophilia
Eosinophil microabscesses
Superficial layering of eosinophils
Extracellular eosinophil granules
Surface epithelial desquamation
Basal zone hyperplasia
Rete peg elongation
Dilated intercellular spaces
Subepithelial fibrosis / sclerosis / lamina propria fibrosis

Table D.[1] Recommended Treatments for EoE

Dietary Therapy (Elimination diet, amino acid based elemental formula)
Topical swallowed corticosteroids
Initial doses[2-4]
Fluticasone (puffed and swallowed via a metered dose inhaler)
Adults – 440-880 mcg twice daily
Children – 88-440 mcg twice to four times daily (to a maximal adult dose)
Budesonide (as a viscous suspension)
Children (< 10 yrs) – 1 mg daily
Older Children & Adults – 2 mg daily
Following administration, patients should not rinse the mouth or eat or drink for 30 minutes.
Systemic corticosteroids
Prednisone – 1-2 mg/kg

Education, Advocacy, and/or Research Support Resources:

American Academy of Allergy, Asthma, and Immunology www.aaaai.org
American Partnership for Eosinophilic Disorders www. apfed.org
Campaign Urging Research for Eosinophilic Disorders www.curedfoundation.org
Children's Digestive Health and Nutrition Foundation www.cdhnf.org
Food Allergy Initiative www.faiusa.org
Food Allergy & Anaphylaxis Network www.foodallergy.org
North American Society of Pediatric Gastroenterology and Nutrition www.naspghan.org
Registry for Eosinophilic Gastrointestinal Disorders www.regid.org
The International Gastrointestinal Researchers www.tigers-egid.cdhnf.org

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