

Eosinophilic Esophagitis (EE)

Apfed
American Partnership for Eosinophilic Disorders
Connecting the Eosinophilic Community

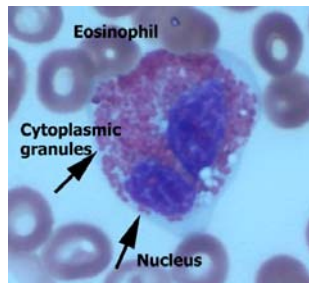
Origin of APFED

The diagnosis of Eosinophilic disorders, once considered rare, has risen dramatically. However, many families still experience a delay in obtaining a correct diagnosis, appropriate treatment, or even up to date information on these disorders. This prompted mothers of children with eosinophil-associated gastrointestinal disorders to take action, and in 2001 APFED was created in an effort to connect and assist other families affected by eosinophilic disorders. Since then, APFED has grown to include both children and adults affected by eosinophilic esophagitis, eosinophil-associated gastrointestinal disorders, hypereosinophilic syndrome, and Churg-Strauss Syndrome. APFED is a 501(c) 3 nonprofit organization dedicated to patients and their families coping with eosinophilic disorders. Our mission is to provide **Education, Awareness, Support and Research to patients with these diseases and to physicians who care for them.**

Our education and awareness efforts are made possible by educational grants from the American Academy of Allergy, Asthma and Immunology and the American College of Allergy, Asthma & Immunology. We are also indebted to our Medical Advisory Board for their continued support.

What is an Eosinophil?

Eosinophils, a type of white blood cell, are an important part of the immune system, helping us fight off certain types of infections, such as parasites. Many different problems can cause high numbers of eosinophils in the blood including allergies, parasitic infection, eosinophil associated gastrointestinal disorders, leukemia, and other problems. When eosinophils occur in higher than normal numbers in the body, without a known cause, an eosinophilic disorder may be present. Eosinophil disorders are further defined by the area affected. For example, eosinophilic esophagitis means elevated numbers of eosinophils and inflammation in the esophagus.



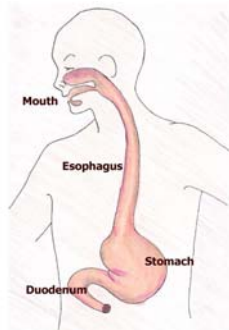
Eosinophil, Courtesy of Dr. Margaret Collins

What is EE?

Eosinophilic esophagitis (**EE**) is a relatively newly recognized disease that over the past decade has been increasingly diagnosed in children and adults. This increase is thought to reflect an increase in diagnosis as well as a true increase in EE cases. Fortunately, the medical community is responding and new scientific information is emerging to guide management of this disorder, which often persists with ongoing or recurrent symptoms.

Eosinophilic esophagitis is characterized by a large number of eosinophils and inflammation in the esophagus (the tube connecting the mouth to the stomach). People with EE commonly have other allergic diseases such as rhinitis, asthma, and/or eczema. EE affects people of all ages and ethnic backgrounds. Males are more commonly affected than females. In certain families, there may be an inherited (genetic) tendency.

In individuals with EE, the eosinophils cause injury to the tissue in the esophagus. EE can be driven by food allergy or intolerance: most patients who eliminate food proteins from their diet (by drinking only an amino-acid based formula) improve.



Upper gastrointestinal tract

Eosinophils are not normally present in the esophagus, although they may be found in small numbers in other areas of the gastrointestinal tract. Diseases other than EE can cause eosinophils in the esophagus including gastroesophageal reflux diseases (GERD), drug hypersensitivity response, and inflammatory bowel disease (Crohn's).

What are the **symptoms** of EE?

Symptoms vary among individuals and age groups. Vomiting may occur more commonly in young children and difficulty swallowing in older individuals.

Common symptoms include:

- Reflux that does not respond to usual therapy (medicines which stop acid production in the stomach)
- Dysphagia (difficulty swallowing)
- Food impactions (food gets stuck in the esophagus)
- Nausea and Vomiting
- Failure to thrive (poor growth, malnutrition, or weight loss)
- Abdominal or chest pain
- Feeding refusal/intolerance or Poor appetite
- Difficulty sleeping

How is EE **diagnosed**?

At present, the only way to definitively diagnose EE is through endoscopy with biopsies. The endoscopy is often performed after treatment with reflux medications (acid suppressors) have failed to relieve the symptoms. During an upper endoscopy, the gastroenterologist looks at the esophagus, stomach, and duodenum (first part of the small bowel) through an endoscope (small tube inserted through the mouth) and takes multiple biopsies (small tissue samples) which a pathologist reviews under the microscope.

The gastroenterologist may be able to see a problem through the endoscope, but eosinophilic esophagitis may be present even if the esophagus looks normal to the doctor. That is why the biopsy samples are important to making the diagnosis of EE. A high number of eosinophils (> 15 per high power field) suggest the diagnosis of EE. GERD also causes eosinophils in the esophagus, but typically far fewer and only in the part of the esophagus closest to the stomach. The pathologist will also look for tissue injury, inflammation, and thickening of the esophageal layers. With EE, the increased eosinophils are limited to the esophagus and not found in other areas of the intestinal tract. Once the diagnosis of EE is confirmed, food allergy testing is typically recommended to guide treatment. Skin prick testing to different foods is the most common type of allergy testing.

Treatment

At present, the two main treatments recommended are dietary management and topical corticosteroids. A comprehensive allergic evaluation to foods and aeroallergens is recommended in the 2007 Consensus Recommendations for Diagnosis and Treatment.

Dietary Therapy

Most children and adults with EE respond favorably to dietary treatments. Dietary treatment may include:

- **Elimination** diets, in which all 'positive' foods on allergy testing are removed from the diet, may be the only treatment needed for some individuals with eosinophilic esophagitis.
- **Six-food elimination diet** is another type of elimination diet that has shown success in some EE patients. Instead of basing dietary elimination on skin testing, patients eliminate the top 6 most allergenic foods (dairy, eggs, wheat, soy, peanuts/other nuts, fish/shellfish).
- **Elemental diets**, in which all sources of protein are removed from the diet, are another dietary therapy. An elemental diet includes only an amino acid formula (building blocks of protein), no whole or partial proteins. Simple sugars, salt and oils are permitted on an elemental diet. Because these formulas are not palatable, the use of a strict elemental diet may require a feeding tube. Amino-acid based formulas are very expensive and are only sometimes covered by health insurance. Elemental diets are effective in treating most children with EE.
- **Food trials** involve adding back one ingredient at a time to one's diet to determine specific foods causing a reaction. Food trials begin after symptoms resolve and eosinophils have cleared. Food trials are handled differently by different professionals, but may involve repeat endoscopies with biopsies as foods are introduced to determine which foods are 'safe' for an individual.

Medications

Medications for EE most commonly include steroids to control inflammation and suppress the eosinophils. Steroids can be taken orally or topically (swallowed asthma medicine). For many patients, **topical corticosteroids** (fluticasone, budesonide) have led to **EE remission** (improvement while on treatment), though their long-term use for maintenance treatment has not been studied. Acid suppressors may also help symptoms in some patients with reflux.

EE requires ongoing care

Patients with EE may require additional endoscopies to assess how the esophagus is responding to specific treatment. Physicians and scientists are working to discover other methods of monitoring and diagnosis. Novel medicines are also being tested that may help with future management of EE.



Prognosis

EE does not appear to limit life expectancy and there is currently no data suggesting EE causes cancer of the esophagus. In some patients, EE is complicated by the development of esophageal narrowing (strictures) which may cause food to lodge in the esophagus (impaction). It is not clear how long EE has to exist before strictures form. However, since the natural history of EE is only emerging, careful monitoring and long-term follow-up is advised. The initial diagnosis of EE can be overwhelming and often affects the entire family. A positive attitude and a focus on non-food activities go a long way in learning to live with EE. With proper treatment, individuals with EE can lead a normal life.

About APFED

American Partnership for Eosinophilic Disorders (APFED) is a non-profit organization dedicated to patients and their families coping with eosinophilic disorders. Our mission is **Education, Awareness, Support and Research**. All medical information is reviewed for accuracy by our medical advisory board. For more information on eosinophilic disorders, or to find information about our annual education conferences, visit www.apfed.org. Our educational efforts would not be possible without educational grants from the American Academy of Allergy, Asthma and Immunology and the American College of Allergy, Asthma & Immunology.

Membership Application

American Partnership for Eosinophilic Disorders

Please print clearly

Name: _____ Phone number (____) _____

Address: _____

City: _____ State/Province: _____ Zip: _____

Country: _____ Email address: _____

____ I would like to become a member ____ \$30 US ____ \$45 Canada ____\$55 International

____ Professional membership (physicians, healthcare providers) ____ \$100 US ____ \$125 Canada
____ \$150 International

____ I would like to make a tax-deductible donation ____\$50 ____ \$100 ____\$250 ____\$500 ____ \$1000
____ other \$_____

Please send the completed application, with check or money order to:

APFED PO Box 29545 Atlanta, GA 30359 or enroll [online at www.apfed.org](http://www.apfed.org) "Join APFED"