Eosinophilic Esophagitis

What is Eosinophilic Esophagitis?

Eosinophilic Esophagitis, also known as EE is an important disorder of the swallowing tube (esophagus) that is different from Gastroesophageal Reflux Disease (GERD). EE is an inflammatory condition of the esophagus that is characterized by having above normal amounts of eosinophils in the esophagus.

Eosinophils are a type of white blood cells that are an important part of the immune system. Eosinophils are found in small quantities in the blood and intestine, but are not normally found in the esophagus. Eosinophils help fight off certain types of infections such as parasites and are one of the types of cells involved in allergic reactions.

Eosinophils can cause damage when they appear in large numbers in areas other than the blood or intestine. For example, if one has seasonal allergies, eosinophils are found in nose; if one has asthma, they are found in the lung; and if one has EE, they are found in the esophagus.

EE is a relatively uncommon disorder, although there has been an increase in the frequency of EE in the pediatric population in recent years. However, EE affects people of all ages, race and gender. The exact cause of EE is unclear, but a link between allergies and EE is probable. The allergen may be food or environmental (aeroallergens). 60% of patients with EE have a personal history of allergy, i.e. allergic rhinitis, asthma or eczema and 20%-40% of patients with EE have a family history of atopy, against breast cancer.

What are the symptoms associated with Eosinophilic Esophagitis?

The symptoms of EE may vary with age. The clinical presentation of EE may be confused with GERD especially in younger children. Infants often present with vomiting, irritability and poor weight gain. In the older child and adolescent, difficulty swallowing and food obstruction or impaction in the esophagus may be more common. Other symptoms might include reflux not responsive to standard medical therapy, nausea, vomiting, abdominal or chest pain, poor appetite, and sleeping difficulties.

How do you diagnose Eosinophilic Esophagitis?

An upper endoscopy with biopsies is necessary for diagnosis of EE. The appearance of the esophagus in EE is quite characteristic. The esophagus appears wrinkled or furrowed and ringed and covered with whitish material or exudate. However, the esophagus may look normal. The gold standard for diagnoses of EE is examining the biopsies (small tissue samples) taken at upper endoscopy under microscope.

The cause of EE can not be determined by the biopsy alone. Once EE has been diagnosed your gastroenterologist may refer you to an allergist for allergy testing. Allergy testing includes a skin prick test (scratching a small amount of food or environmental allergen into the skin) and a RAST test (used to confirm immediate reactions to a specific food).

Treatment of Eosinophilic Esophagitis

The treatment is either dietary or medication or both.

Diet

If a specific food(s) (allergen) are detected by allergy testing, removal of the food(s) may be all that is required. The most common foods to consider include milk, egg, peanuts, soy, wheat, tree nuts, fish and shellfish. However, in many patients, especially older children and adults, no specific allergen is found. In this group of patients, an elimination diet can be considered in which all sources of protein are removed from the diet and only an amino acid (building blocks of protein) based formula is allowed. Simple sugars, salts and oils are permitted on this diet. Once the patient is symptom free, a food challenge with one new food reintroduced every 4-7 days can be considered. The food challenge helps to identify the responsible food(s), and is the basis of long-term dietary treatment. It is important to work with a nutritionist.

Drug Treatment

Your physician may recommend use of medications. To date no medications have been approved specifically for the treatment of EE, however there is evidence that corticosteroids may be of benefit. Corticosteroids can be taken topically (swallowed from an asthma inhaler) or orally.

Topical corticosteroids: Fluticasone propionate and beclomethasone are used at various doses, twice a day, depending on age. The medication is swallowed and not inhaled. Oral and esophageal thrush (candidiasis) are the major side effects.

Systemic corticosteroids (i.e. prednisone) may be necessary. Symptoms usually improve within 1-2 weeks. Your physician will review with you the dose and tapering schedule as well as the side effects.

Patients with EE may require additional endoscopies with biopsies to assess how the esophagus is responding to treatment.

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Complications of Eosinophilic Esophagitis

Failure to thrive
Esophageal strictures

For more information or to locate a pediatric gastroenterologist in your area please visit our website at:
www.naspghan.org

Additional helpful website: www.apfed.org/healthcare.htm

IMPORTANT REMINDER: This information from the North American Society for Pediatric Gastroenterology, Hepatology and Nutrition (NASPGHAN) is intended only to provide general information and not as a definitive basis for diagnosis or treatment in any particular case. It is very important that you consult your doctor about your specific condition.
**Eosinophilic Esophagitis**

**What is it?**

An eosinophil is a type of white blood cell that works as a part of the body’s immune system. Usually they are found in a small number in the blood and in the GI tract, but not usually in the esophagus. When eosinophils are present in the esophagus they may contribute to inflammation and injury.

**Who gets it?**

This disorder can affect anyone; the majority of cases have been men in their 20-30s. In children, males also tend to be more affected than females and more often in Caucasian individuals. The number of cases of eosinophilic esophagitis is increasing but this may be partly due to increased recognition. Many people have a history of allergies to food or the environment and/or a history of asthma and atopic dermatitis. These individuals may also have an increased number of eosinophils in your blood.

**Why does it happen?**

The cause of this disorder is not completely understood, but is likely related to both genetic and factors in the environment. Once the eosinophils are in the esophagus they may cause local inflammation and this might cause difficulty with swallowing.

**What are the symptoms?**

The symptoms are variable depending on the patient’s age:

**In adults:**

- Trouble swallowing. Most often this is noticed with solid foods.
- Food impaction. 54% of people with eosinophilic esophagitis patients will experience episodes of food getting stuck in the esophagus.
- Chest pain, often located in the center
- Heartburn
- Upper abdominal pain

**In children:**

- Feeding dysfunction, such as not developing normal eating patterns, not progressing to other foods after liquids, or refusing solid foods
- Vomiting
- Abdominal pain
- Trouble swallowing
- Food impaction
How is it diagnosed?

Based on symptoms, the diagnosis is usually made by **upper endoscopy** where a tube with light and camera is inserted through the mouth, into the throat, down to the stomach and first part of your small intestine. Tissue samples can be taken during this procedure. There are some specific findings associated with this condition as listed below, but sometimes the esophagus looks completely normal:

- Stacked circular rings (“feline” esophagus)
- Strictures, or narrowing, especially near the top of the esophagus
- Changes in the appearance of blood vessels of the esophagus
- Linear furrowing
- White bumps
- Narrow esophagus

As mentioned, taking tissue samples of the esophagus is important. When looking under the microscope at the tissue there are often over 15 eosinophils per high power field.

Other tests that may be performed include:

- **Esphagram/Barium swallow**: this is an x-ray that is taken during and after swallowing a thick chalky liquid that covers the walls of the esophagus and stomach.
- **Blood work**: There is no test that will specifically diagnose eosinophilic esophagitis. Over half of people will have an elevated IgE level (a specific protein that the immune system uses) and 40-50% of patients have increased eosinophils in their blood

How is it treated?

Treatment can include diet, medicines and procedures during an upper endoscopy.

- **Dietary therapy** is more often used, and successful, in children than adults. This is based on the fact that many patients have food allergies and possibly these allergies lead to eosinophilic esophagitis. While avoidance of the problem foods can be effective, at times this can cause a lack of nutrients and often symptoms return if the special diet is stopped. This treatment is typically only tried with the help of a dietician or allergist. There are a couple specific dietary approaches, **elimination diet** where complete food groups are avoided (often specifically 6 foods: milk, soy, eggs, peanuts/tree nuts, wheat and seafood) or an **elemental diet** where a formula is consumed either through the mouth or a tube that has all the necessary nutrients. The elemental diet is expensive and often does not taste good. Elimination diets have also been tried in adults but require substantial lifestyle changes.

- **Medicines**
  - **Acid suppression**. Proton pump inhibitors (PPI), medicines that decrease acid levels, may help, because the inflammation in the esophagus makes the tissue more prone to injury and more sensitive to acid. Also heartburn or gastroesophageal reflux disease
(GERD) can be present with eosinophilic esophagitis and a PPI can work to treat this or perhaps works by an alternative anti-inflammatory way.

- **Topical glucocorticoids.** Swallowed steroids work to cut down inflammation right at the tissue of the esophagus. Most people respond quickly, within several days or a week and are treated for 6-8 weeks. Sometimes symptoms return when the treatment is stopped and maintenance treatment is necessary.

- **Esophageal Dilation.** An upper endoscopy is performed with a lighted tube placed down the throat into the stomach. This time, a balloon is also inserted and when it is inflated, it will stretch the esophagus. This can also be done with a bougie (a soft tapered instrument dilator). Although this can provide relief, it does not impact the inflammation that initially caused the narrowing. Often it is only used if medicines do not work or the narrowing is very significant.

What can I do?

Because of the strong link of eosinophilic esophagitis with allergies, often a referral to an allergist or immunologist is made to help guide treatment decisions.

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