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Eosinophilic Esophagitis (EoE)

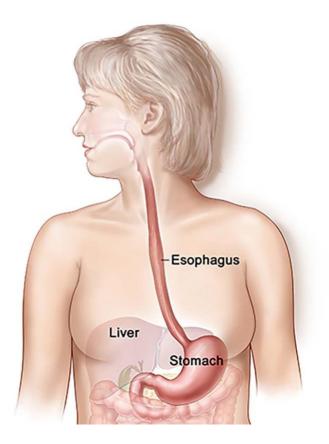
Eosinophilic Esophagitis, commonly known as EoE, is a chronic inflammatory condition of the esophagus. Think of your esophagus as a smooth highway that carries food from your mouth to your stomach. In EoE, this highway becomes inflamed and narrowed due to an unusual buildup of white blood cells called eosinophils. These cells help fight infections throughout your body; usually, very few are seen in the esophagus. But in EoE, they gather in large numbers in your esophagus, where they don't belong.

When these cells accumulate, they cause inflammation, making eating and swallowing difficult and sometimes painful. The condition might sound complicated, but breaking down the name helps understand it better: "eosinophilic" refers to the type of white blood cell involved, and "esophagitis" means inflammation of the esophagus. Together, they describe this unique condition that affects people of all ages, from infants to adults.

What causes EoE ?

The causes of eosinophilic esophagitis are not fully understood, and researchers continue to study the condition. Many medical scientists believe that an imbalance in the immune system causes EoE . Many theories about what might cause this imbalance include environmental factors, infections, dietary factors, or even genetic predisposition. However, the exact cause of EoE is not yet known.

Your immune system works like a highly trained security team, constantly patrolling your body to protect you from harmful invaders while maintaining peace with harmless substances. However, in EoE, this security team becomes overzealous, particularly in your esophagus. The immune system begins treating certain foods or environmental substances as threats, even though they're harmless. This misguided immune response leads to chronic inflammation in your esophagus. It's similar to what happens in allergic conditions like asthma and eczema,



which is why many people with EoE also have these conditions. Over a period of years, this chronic inflammation can cause the esophagus to become narrow and rigid, making it harder for food to pass through. Unfortunately, EoE is a lifelong condition once symptoms occur.

Who gets EoE ?

EoE is a relatively rare disease but is increasing in prevalence, with an estimated 1 in 2,000 Americans affected. More and more people are being diagnosed with EoE these days. This is partly because EoE is becoming more common, and doctors are better at recognizing it. EoE affects people of all ages and ethnic backgrounds. We see it in infants, children, and adults. It attacks more men than women, with a peak incidence in the 30s and 40s. About 2/3 of patients with EoE have some allergy or have a close family member with allergies, such as food allergies, asthma, or eczema. The risk of siblings being affected is less than 5% unless twins are involved.

What are the symptoms of EoE ?

Symptoms vary depending on the age and duration of the disease and include heartburn, chest pain, and vomiting. In adults, the most frequent symptom is <u>dysphagia</u> or difficulty swallowing. Initially, there is an odd sensation as if food is sticking on the way down. In extreme cases, a food bolus may become lodged in the esophagus. It won't come up, and it won't go down! This is called a "food impaction" and, if persistent, is a medical emergency. A food impaction signifies long-standing disease and advanced stages of EoE but, sadly, is often present when the diagnosis is first made.

Public health data suggest that, on average, there is a delay of about five years from the onset of symptoms until the diagnosis is made. The diagnosis is often made only after a food impaction and a trip to the Emergency Room. We must improve our approach to diagnosing EoE to start treatment earlier and prevent long-term damage to the esophagus. We need to do a better job, including better patient education, as in this article.

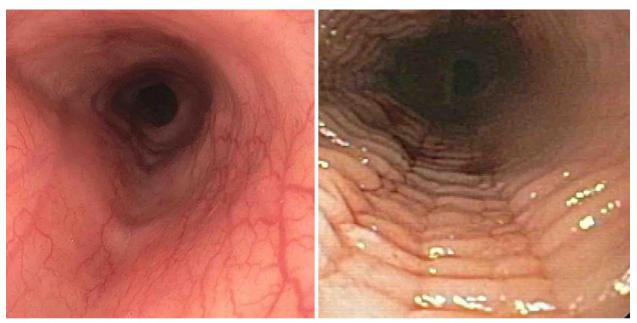
How does your doctor know?

The gold standard for diagnosing EoE is an upper endoscopy. During this procedure, a doctor uses a thin, flexible tube equipped with a tiny camera to examine your esophagus in detail. Though this might sound intimidating, you'll be under sedation during the procedure and won't feel any discomfort.

When performing the endoscopy, your doctor will look for characteristic signs of EoE that experienced physicians often describe as "rings and railroad tracks" in the esophagus lining. These visual changes happen because inflammation causes the esophagus to develop circular rings and vertical lines that look like a railroad track. Your doctor will also notice if there's any narrowing or structuring of the esophagus that might be making it difficult for food to pass through.

During the same procedure, your doctor will take several small tissue samples, called biopsies, from different areas of your esophagus. These samples are sent to a laboratory where a pathologist examines them under a microscope. The pathologist specifically looks for and

counts eosinophils in the tissue. In a healthy esophagus, very few, if any, eosinophils should be present. However, if the pathologist finds more than 15 eosinophils in a high-powered microscopic field, this strongly suggests a diagnosis of EoE.



Normal Esophagus Lining

Eosinophilic Esophagitis

Note the linear tracks and circular rings classic for EoE at Endoscopy

It's important to note that other conditions can cause eosinophils to infiltrate the esophagus, including chronic gastroesophageal reflux disease or acid reflux. There is a bit of overlap between these conditions. are specific guidelines that doctors follow to diagnose EoE. They will evaluate your symptoms, upper endoscopy results, and what the pathologist sees when they examine your tissue samples.

Treatment

Successfully managing EoE usually requires a multifaceted approach, like solving a puzzle with several pieces that must fit together. Your healthcare team will work with you to develop a treatment plan tailored to your specific symptoms, lifestyle, and needs. EoE is a lifelong disease, and we have no cure at present. Treatment aims to reach a manageable point, keeping the disease in remission and minimizing esophageal damage and scarring. Early diagnosis and treatment are crucial. If left untreated for a long time, EOE can lead to permanent scarring of the esophagus, which makes it difficult for food to pass down.

We can avoid these long-term complications if a therapeutic regimen is started early and the disease is controlled. It usually takes 2–3 months to see a noticeable improvement with

treatments. Often, at that time, repeat endoscopy and biopsies are performed to assess the degree of esophageal infiltration and the response to treatment.

Elimination diets

Research has shown a strong connection between food allergies and eosinophilic esophagitis (EOE). The following six foods are most commonly associated with this allergic response: cow's milk, dairy, wheat, soy, eggs, nuts, and seafood or shellfish. *Unfortunately, routine skin and blood allergy tests cannot accurately determine which foods might be the problem, so formal allergy testing is not very useful.*

Elimination diets can help identify food allergies and improve EOE symptoms. Strict diets are hard to maintain; unfortunately, no accurate test determines which food allergies are connected with EOE . <u>The most common food trigger identified in EoE is cow's milk.</u> Any food can trigger EoE, but some are more common than others. In addition to cow's milk, wheat and soy are common triggers. Fish, nuts, and rice are less common triggers. If a long-term special diet is necessary, meeting with a registered dietitian can help ensure that nutritional needs are met with dietary restrictions in place.

Medications

Proton Pump Inhibitors (PPIs)

Proton Pump Inhibitors (PPIs) are often used as a first-line treatment. These medications reduce stomach acid production and help decrease inflammation in your esophagus. About 30-40% of adults with EoE see significant improvement with PPIs alone. Proton Pump Inhibitors such as Nexium (esomeprazole), Prilosec (omeprazole), and Protonix (pantoprazole) are often prescribed. Daily PPI use is most helpful in patients with chronic heartburn and those with stricture formation after esophageal dilation.

Topical Corticosteroids

Topical steroids, such as **Flovent (fluticasone) or Pulmicort (budesonide)**, are a common treatment option for EoE that can put the condition into remission. These medications are similar to those used to treat asthma *but are swallowed rather than inhaled* to coat the esophagus and reduce irritation and inflammation in the lining. Fluticasone is administered with an asthma inhaler, but the patient is advised to swallow the medication, not inhale it. Budesonide is usually given as a slurry made by a compounding pharmacy. Patients are advised not to eat or drink anything for at least one hour after each dose.

Eohilia (2024)

Eohilia represents an innovative treatment option for eosinophilic esophagitis (EOE). Unlike Dupixent, which requires injections, Eohilia is an oral solution containing budesonide, a well-established corticosteroid used to treat inflammation. It's worth noting that while Eohilia's formulation is new, budesonide has been used in compounded prescriptions for esophagitis for years, taken as an "off-label" compounded prescription made by a specialty pharmacy.

What makes Eohilia unique is its specialized formulation of budesonide designed specifically for the esophagus. The medication has been engineered to change consistency - becoming more fluid for easy



swallowing, returning to its original form to effectively coat the esophagus and deliver medication directly where needed. It comes in convenient premixed stick packs, requiring no refrigeration, making it portable and suitable for on-the-go use. While the exact mechanism isn't fully understood, research shows that budesonide reduces eosinophils and other inflammatory cells in the esophagus, leading to symptom relief. The medication requires specific administration steps - including shaking, snipping, squeezing, swallowing, and rinsing all designed to ensure effective delivery and minimize side effects in the mouth. Steroid therapies are very effective but may cause side effects such as thrush or yeast infections. Once esophageal inflammation is adequately controlled, the dose of swallowed corticosteroid is decreased to the smallest dose necessary to maintain control of EoE to reduce the risk of steroid side effects. However, it's important to note that inflammation typically returns in 90% of patients if steroid therapy is discontinued

Dupixent

As of May 2022, the FDA approved a new drug called Dupixent[®] (dupilumab) to treat EoE in adults and children 12 years and older. Cytokines are small proteins that control the activity of other immune system cells, including eosinophils. This medication works by blocking cytokines that contribute to esophageal inflammation. Dupixent[®] can help alleviate dysphagia symptoms and may be an effective treatment



option for patients who have not responded to other therapies. Dupixent is an injectable medication called a monoclonal antibody (or biologic) administered weekly through a subcutaneous injection placed under the skin.

When comparing Eohilia to Dupixent, both treatments effectively target inflammation through different mechanisms: Dupixent blocks specific inflammatory signals, while Eohilia's budesonide acts on a broader range of inflammatory cells. The main practical difference is administration - Dupixent requires injections weekly, while Eohilia is taken orally twice daily. The choice between these treatments should be made in consultation with your healthcare provider, considering your personal preferences and lifestyle needs.

Esophageal Dilation

If the esophagus has become too narrow due to EoE, esophageal dilation may be recommended to treat it. Performed during upper endoscopy, this procedure involves temporarily inflating a special elongated balloon into the restricted section of the esophagus and stretching it out. In severe cases, repeated dilations may be necessary.

Common misconceptions

- 1. The idea that EoE is just another term for acid reflux or GERD (Gastroesophageal Reflux Disease) is among the most common fallacies. Both disorders are unique, have separate underlying causes, and call for different treatment strategies, even if they can have comparable symptoms, such as trouble swallowing and chest pain. EoE is an imbalance of the immune system in which eosinophils produce inflammation, unlike GERD, which is predominantly caused by stomach acid impacting the esophagus. This implies that effectively managing EoE may require more than just using antacids or acid-reducing drugs.
- 2. Another widespread misunderstanding is the idea that a person will eventually "grow out of" EoE . Regretfully, this isn't true. EoE is a chronic illness that needs to be managed continuously throughout a person's life. There isn't a definitive cure for EoE at this time; while therapies can successfully manage the condition, the symptoms may alter with time. Knowing this makes it easier for patients and their families to plan long-term care instead of waiting for the illness to disappear.
- 3. It's also a common misconception that food allergies are the only cause of EoE. Dietary cues are important in many situations, but the truth is more nuanced. Environmental allergens, genetic predispositions, and immune system abnormalities are some of the variables that contribute to the development of EoE. Because other variables may contribute to their disease, some individuals may still experience symptoms even after identifying and removing food triggers.

Eating Techniques and Meal Planning

Taking control of your eating habits can make a significant difference in managing EoE symptoms. This means being mindful of what you eat and how you eat. Consider eating your meals in a relaxed environment without rushing. Take smaller bites and chew your food thoroughly before swallowing. Some people find it helpful to cut their food into small pieces, especially meats or other foods that might be more challenging to swallow. Keep plenty of

liquids available during meals. Small sips between bites can help food move more easily through your esophagus. However, avoid using liquids to "wash down" poorly chewed food, as this can increase your risk of food becoming stuck.

Emergency Planning for EoE

Food getting stuck in your throat (food impaction) is the most serious concern with EoE. Having a plan helps you stay calm and get help quickly when needed. First, it's essential to recognize the difference between food feeling temporarily stuck (which might resolve with small sips of water) and true food impaction. In a true impaction, you'll find that nothing will go down past the stuck food, and you might experience excessive drooling, difficulty swallowing your saliva, or chest discomfort. If this happens, this is a medical emergency requiring immediate attention. Work with your doctor to make an emergency plan. Find out which ER to use (ideally one familiar with EoE) and carry a medical card listing your condition, medications, and allergies. This helps emergency staff treat you properly, especially if you have trouble speaking.

The Emotional Impact of EoE

Living with Eosinophilic Esophagitis (EoE) extends far beyond its physical manifestations, impacting mental health and emotional well-being across various aspects of daily life and social interactions. Many individuals with EoE experience significant anxiety surrounding meals, particularly in social situations or unfamiliar environments where food choices may be limited or uncertain. The constant need to be vigilant about food choices creates a persistent state of hyperawareness during what should be enjoyable social occasions.

The development of phagophobia, an intense fear of swallowing, is common among EoE patients due to past experiences with food impaction or difficulty swallowing. This fear manifests as hesitation during meals, avoidance of certain foods or textures, reduced portion sizes, extended meal times, and increased stress during everyday eating. These challenges often lead to social isolation, as dietary restrictions create barriers to participating in food-centered gatherings, leaving individuals feeling different or excluded during shared meals, repeatedly explaining their dietary needs, and missing out on cultural or traditional food experiences.

The condition's impact extends into various aspects of daily living, affecting work situations involving business lunches or team meals, dating and romantic relationships, family dynamics and meal planning, and travel experiences. It's crucial to recognize these emotional responses as normal and valid, as living with a chronic condition like EoE requires both physical and emotional adaptation. These feelings are completely normal and valid. It's important to acknowledge them and seek support when needed. Consider talking with a mental health professional who has experience working with chronic health conditions. They can help you develop coping strategies and maintain a positive outlook while managing your condition.

Building Your Support Network

Living with EoE becomes easier when you have a strong support network. This network can include your healthcare team, family members, friends, and others living with EoE. Support groups, in-person or online, can be valuable resources for sharing experiences, learning new management strategies, and finding emotional support from others who truly understand your challenges. When you first find out you have EoE, it can be overwhelming. Families often benefit from participating in support groups and organizations. Visit the American Partnership for Eosinophilic Disorders (APFED) and Campaign Urging Research for Eosinophilic Disease (CURED). These are two lay organizations that provide valuable, reliable resources.

What is the long-term prognosis?

While being diagnosed with eosinophilic esophagitis (EoE) can feel overwhelming, the longterm outlook is generally favorable. EoE does not affect life expectancy and has no association with esophageal cancer. Most patients achieve significant symptom improvement through proper medical management, dietary adjustments, and lifestyle modifications. While the condition may require periodic endoscopic procedures to monitor or treat esophageal narrowing, these interventions are typically well-tolerated and effective. The disease course often fluctuates, with periods of symptom improvement and worsening, making ongoing medical supervision essential.

Though developing esophageal strictures and eating difficulties may necessitate lifestyle adjustments, patients can maintain a good quality of life with proper support and medical care. Since EoE 's natural history is still being understood, careful monitoring and long-term follow-up are recommended. Stay informed, maintain open communication with your healthcare team, and don't hesitate to seek support when needed. Your journey with EoE is unique to you, and with the right tools and support, you can navigate this chronic condition while maintaining your quality of life.

The Future...

For years, more research was needed on EoE. But in the last decade, scientific breakthroughs have led to a better understanding of the underlying pathology. The field of EoE research is rapidly evolving, with scientists and medical professionals constantly working to understand this condition better and develop more effective treatments. Current research focuses on several promising areas that could improve care options. Scientists are investigating the genetic factors that might make some people more susceptible to developing EoE. This research could lead to more personalized treatment approaches based on an individual's genetic profile. Other studies are exploring new medications that target specific aspects of the immune response involved in EoE, potentially offering more precise treatment options with fewer side effects. Researchers are also developing better ways to monitor EoE activity without requiring frequent endoscopic procedures. This might include blood tests or other less invasive methods that could make tracking how treatments are working easier.



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Robert Fusco MD December 2024



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