

## WHAT ARE THE SYMPTOMS OF EGPA?

While almost all patients with EGPA have asthma, the symptoms people experience with EGPA may vary greatly and may affect different organ systems such as the lungs, sinuses, and nerves. It can also affect the heart, gastrointestinal tract, reproductive organs, skin, urinary system, and other organs.

People with EGPA may experience the following symptoms:

- Asthma
- Allergic rhinitis (and sinusitis)
- Fever
- Loss of appetite/weight loss (may be rapid and sudden)
- Fatigue/malaise
- Shortness of breath and/or coughing
- Abdominal pain
- Diarrhea, nausea, and vomiting
- Gastrointestinal bleeding
- Rashes or skin sores
- Joint aches and/or muscle pain
- Severe pain, numbness and tingling in the hands and feet
- Chest pain
- Irregular heartbeat
- Blood in urine and/or stools
- Kidney disease

## ABOUT APFED

Founded in 2001, the American Partnership for Eosinophilic Disorders (APFED) is a 501(c)(3) nonprofit organization with a mission to passionately embrace, support, and improve the lives of patients and families affected by eosinophil-associated diseases through education and awareness, research, support, and advocacy. To learn about the impact we are making in these areas, visit our website at [apfed.org](http://apfed.org).

Our work is funded by grants, membership dues, and donations. If they wish, donors may choose to direct their gifts to specific programs, such as research or education.

APFED operates with integrity and transparency in accordance with nonprofit guidelines. APFED is an accredited charity of the BBB Wise Giving Alliance and is a Platinum Member of GuideStar. View our independently audited financial reports and annual reports on [guidestar.org](http://guidestar.org) and [apfed.org](http://apfed.org). On average, APFED directs more than 85% of its annual revenue to programs and services. This is 20% higher than the BBB Wise Giving Alliance standard of 65%.

Visit [apfed.org](http://apfed.org) for educational materials, community news, or information about events such as APFED's annual patient education conference and National Eosinophil Awareness Week (third week of May). To enroll in Eosinophil.Connect Patient Registry, visit [apfed.org/connect](http://apfed.org/connect).



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## JOIN US

Become a member of APFED to stay up to date on news and initiatives related to eosinophil-associated diseases. For information about membership and benefits or to make a donation to support our work, visit [apfed.org](http://apfed.org).

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# EOSINOPHILIC GRANULOMATOSIS WITH POLYANGIITIS (EGPA)



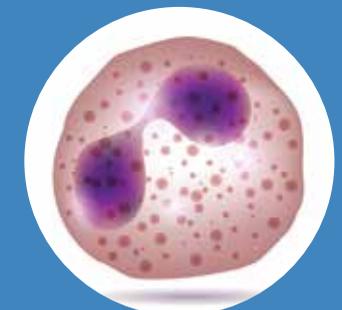
## WHAT IS EOSINOPHILIC GRANULOMATOSIS WITH POLYANGIITIS?

Eosinophilic Granulomatosis with Polyangiitis (EGPA), formerly known as Churg-Strauss Syndrome, is a rare autoimmune disorder characterized by inflammation of blood vessels and the presence of high numbers of white blood cells known as eosinophils. Almost all patients with EGPA have asthma, which is often severe. The inflammation can restrict the flow of blood to organs and tissues, in turn causing damage. It primarily affects small blood vessels. Without treatment, the disease may be fatal.

## WHAT IS AN EOSINOPHIL?

Eosinophils are a type of white blood cell that are part of our immune systems, helping us fight off certain types of infections.

Many different problems can cause high numbers of eosinophils in the blood including allergies (food and environmental), parasitic infections, and certain cancers, to name a few.



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## HOW IS EGPA DIAGNOSED?

EGPA can be difficult to diagnose because its symptoms are similar to other diseases. There are no specific tests to confirm EGPA. High levels of blood eosinophils along with asthma and pneumonia are often the first clues that lead to a diagnosis. The disease is considered present when a person has at least four of the following six features:

- Asthma
- Elevated number of eosinophils
- Nerve damage (numbness and pain in hands/feet)
- Pulmonary infiltrates
- Sinus problems
- Presence of eosinophilic vasculitis

EGPA often presents in three phases, although not everyone will develop all of the symptoms, nor in order. Phase 1 usually presents with allergic conditions such as asthma, hay fever, and sinus or nasal pain and inflammation. Phase 2 often presents with high levels of eosinophils in the blood and tissue. In phase 3, patients may experience inflammation of blood vessels (vasculitis). A skin rash may develop, and patients may experience numbness and/or weakness.

To help guide the diagnosis, your doctor may order blood tests, imaging tests, nerve studies, and may biopsy tissue and/or organs.

## HOW IS EGPA TREATED?

When treating EGPA, the goal is to reduce inflammation and the number of eosinophils in

### RESOURCES

- APFED, [apfed.org](http://apfed.org)
- EOS Connections, [apfed.inspire.org](http://apfed.inspire.org)
- Vasculitis Foundation, [vasculitisfoundation.org](http://vasculitisfoundation.org)

the blood. Many patients who have EGPA respond well to systemic corticosteroids and some also need immunosuppressant drugs. Other patients may have symptoms that are resistant to these therapies.

- **Systemic corticosteroids** (e.g., prednisone) are often prescribed to treat EGPA. This medication changes the way the immune system functions and reduces inflammation. A higher dose may be prescribed initially to get symptoms under control quickly, and then gradually decreased to the lowest effective dose.
- **Mepolizumab** can help reduce the amount of corticosteroids that are needed to keep EGPA under control. This is important since corticosteroids can have many unwanted side effects. Mepolizumab is given as monthly injections.
- **Immunosuppressant drugs** (e.g., azathioprine, mycophenolate, cyclophosphamide, methotrexate, mycophenolate mofetil) can be used in conjunction with systemic corticosteroids to get symptoms under control.

For those who have not responded favorably to these treatments, the following may be considered:

- **Intravenous immune globulin** is an antibody therapy to help fight infections. It is given as an infusion.
- **Interferon-alpha** (or interferon-alfa) is a targeted therapy given as an injection to suppress EGPA symptoms. It may slow down

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or stop eosinophil cells from dividing. It has shown some success inducing remission.

- **Rituximab** is a targeted therapy that alters the immune system's response. It is given as an infusion and has been shown to improve symptoms of EGPA. It has not been studied in large trials, and long-term safety and efficacy are not known.
- **Omalizumab** is a targeted therapy that blocks the allergy antibody, IgE, lessening allergic reactivity. It is approved for use in the U.S. to treat moderate to severe persistent asthma that does not respond to inhalers and other usual asthma medications. This therapy is sometimes given to patients with EGPA to help control asthma.

**Benralizumab** is currently being studied as a potential therapy for EGPA. It is an antibody that binds to a receptor on eosinophils and attracts other cells to destroy eosinophils. It was recently approved in the U.S. and several other countries as an add-on maintenance treatment in severe, eosinophilic asthma.

Work with your healthcare provider(s) to determine the type of treatment, dosage, and duration of treatment based on your individual needs, as well as potential side-effects.

## WHAT CAUSES EGPA?

The cause of EGPA is unknown. It is believed to result from an interaction of genetics and the environment, complicated by an overactive immune system.

Because all patients with EGPA have high levels of eosinophils at some point, it is thought that there may be some dysregulation of eosinophil production, maturation, or development.

Anti-neutrophil cytoplasmic antibodies (ANCA) are also present in many patients with EGPA and may play a role in disease development. The exact role of ANCA remains unclear.

## WHO IS AFFECTED BY EGPA?

The exact prevalence of EGPA is unknown, however, it is believed that there are roughly 14-15 cases per million people worldwide, equating to roughly 5,000 people in the U.S., though the number may be higher. EGPA can happen in a broad age range, though on average is diagnosed in adults 35-50 years old. It has rarely been seen in children. EGPA equally affects males and females.

## WHAT IS THE PROGNOSIS?

There is no cure for EGPA. It is estimated that more than 90% of patients will achieve remission and resolution of symptoms. Complications depend on the organs affected and may include peripheral nerve damage, scarring of the skin, heart disease, and kidney damage.

Relapses are common and asthma may persist. The involvement of the heart and airways will warrant ongoing follow-up with a healthcare provider who can closely monitor symptoms. Patients who have EGPA should receive ongoing medical care to maintain optimum health and for early detection of relapses.