

ANCA-ASSOCIATED VASCULITIS: EOSINOPHILIC GRANULOMATOSIS WITH POLYANGIITIS (formerly Churg-Strauss syndrome)

What is eosinophilic granulomatosis with polyangiitis (EGPA), formerly Churg-Strauss syndrome?

Eosinophilic granulomatosis with polyangiitis (EGPA), formerly called Churg-Strauss syndrome, is a form of vasculitis—a family of rare diseases characterized by inflammation of the blood vessels, which can restrict blood flow and damage vital organs and tissues. EGPA is one of the rarest forms of vasculitis and primarily affects the small blood vessels. Individuals diagnosed with EGPA usually have a history of asthma or allergies.

EGPA can affect the lungs, sinuses, skin, heart, intestinal tract, kidneys, nerves and other organs. EGPA is known as an ANCA-associated vasculitis, referring to a blood protein (anti-neutrophil cytoplasmic antibody) that attacks the body's own cells. Other forms of ANCA-associated vasculitis include granulomatosis with polyangiitis (GPA) and microscopic polyangiitis (MPA). The detection of ANCA in the blood can support a diagnosis of EGPA in some patients.

EGPA is a serious but treatable disease. Treatment typically includes corticosteroids such as prednisolone (prednisone) used in combination with medications that suppress the immune system. A new medication has been added to the treatment toolbox in Australia. The biologic drug mepolizumab is approved (but not funded) for the treatment of EGPA in Australia.

Even with effective treatment, EGPA is a chronic illness with cycles of relapse and remission that can cause serious health problems, so ongoing medical care is necessary.

Causes

The cause of EGPA is not fully understood by researchers. Vasculitis is classified as an autoimmune disorder—a disease which occurs when the body's natural defense system mistakenly attacks healthy tissues. Researchers believe an infection may set the inflammatory process in motion. Genetic and environmental factors, such as inhaled allergens, or certain medications or vaccinations, may also play a role in triggering an onset or relapse of EGPA.

Who gets EGPA?

EGPA can occur at any age, however the average age of diagnosis is between 35 and 50 years old. Women and men appear to be affected equally. EGPA is rare, with an incidence in the United States of 1 to 3 cases per 100,000 adults per year. The international incidence of EGPA is estimated at 2.5 cases per 100,000 adults per year.

Symptoms

The symptoms of EGPA can range from mild to life-threatening, depending on which organs are involved and the extent of disease. Symptoms may vary among individuals, however almost all patients have asthma and/or sinus polyps, and a higher-than-normal level of white blood cells called "eosinophils."

ANCA-ASSOCIATED VASCULITIS: EOSINOPHILIC GRANULOMATOSIS WITH POLYANGIITIS (formerly Churg-Strauss syndrome)

Other symptoms can include:

- Fever
- Fatigue/malaise
- Rapid and sudden weight loss
- Muscle and joint pain
- Skin rashes
- Numbness or tingling of the hands or feet
- Sudden loss of strength in the hands or feet
- Chest pain or palpitations
- Increasing shortness of breath or coughing
- Abdominal pain
- Presence of blood in the stools
- Kidney disease: (Note: A patient can have kidney disease without having symptoms, so patients with vasculitis of any form should have regular urine tests.)

Diagnosis

There is no single test for diagnosing EGPA, so your doctor will consider a number of factors, including a detailed medical history; physical examination; laboratory tests; specialized imaging studies; and, when indicated, a biopsy of an affected tissue or organ.

- **Urinalysis:** Excessive protein or presence of red blood cells may indicate inflammation of the kidneys. Your doctor may use this test to help diagnosis EGPA, and to monitor the kidneys during and after treatment.
- **Blood tests:** The ANCA test, when positive, can be helpful in confirming diagnosis, however, it is positive in only 30-50 percent of individuals with EGPA. Also, it can be positive in the other ANCA-associated forms of vasculitis (GPA and MPA). Other common tests include the erythrocyte sedimentation rate (ESR) test and the C-reactive protein (CRP) test. All of these tests may support a diagnosis of EGPA, but are not conclusive on their own. A tissue biopsy is typically needed.
- **Tissue biopsy:** This surgical procedure removes a small tissue sample from an affected vessel or organ, which is examined under a microscope for signs of inflammation or tissue damage.
- **Imaging studies:** X-rays and computed tomography (CT) scans may reveal changes in your lungs or sinuses that are characteristic of EGPA, such as nasal and sinus polyps.
- **Heart echocardiogram:** All patients with EGPA should be screened for heart involvement.

Treatment

EGPA is traditionally treated with high-dose steroids such as prednisolone, to reduce inflammation. For more severe cases, prednisolone is used in combination with drugs that suppress the immune system's response, such as methotrexate azathioprine, mycophenolate mofetil, and/or cytotoxic agents, such as cyclophosphamide. But long-term use of these drugs can put patients at risk for side effects, complications and relapse.

ANCA-ASSOCIATED VASCULITIS: EOSINOPHILIC GRANULOMATOSIS WITH POLYANGIITIS (formerly Churg-Strauss syndrome)

In Australia, the biologic drug mepolizumab has been recently approved by the Therapeutic Goods Administration, but is not publicly funded as at July 2019. (Biologic medications are complex proteins derived originally from living organisms. They target certain parts of the immune system to control inflammation.) In EGPA, mepolizumab works by reducing the number of eosinophils in the body. When present in normal levels, eosinophils can help protect the body against infection, but over-production can cause inflammation and potentially damage vital organs and tissues.

Side effects

The medications used to treat EGPA have potentially serious side effects, such as lowering your body's ability to fight infection, and potential bone loss (osteoporosis), among others. Therefore, it's important to see your doctor for regular checkups. Medications may be prescribed to offset side effects. Infection prevention is also very important. Talk to your doctor about getting vaccinated against influenza, pneumococcal (pneumonia) vaccination and/or shingles vaccination, which can reduce your risk of infection.

Relapse

Even with effective treatment, relapses are common for individuals with EGPA. If your initial symptoms return or you develop new ones, report them to your doctor as soon as possible. Regular doctor visits and ongoing monitoring of lab and imaging tests are important in detecting relapses early.

Complications

Among the most serious potential complications are heart problems, treatment-induced infections, persistent asthma and, for some, long-lasting numbness or decreased sensitivity in feet or hands. Long-term follow-up is important.

Your medical team

Effective treatment of EGPA may require the coordinated efforts and ongoing care of a team of medical providers and specialists. In addition to a primary care provider, you may need to see the following specialists: rheumatologist (joints, muscles, immune system); pulmonologist (lungs); cardiologist (heart); nephrologist (kidneys); dermatologist (skin); neurologist (brain/nervous system); or others as needed.

The best way to manage your disease is to actively partner with your health care providers. Get to know the members of your health care team. It may be helpful to keep a health care journal to track medications, symptoms, test results and notes from doctor appointments in one place. To get the most out of your doctor visits, make a list of questions beforehand and bring along a supportive friend or family member to provide a second set of ears and take notes.

Remember, it's up to you to be your own advocate. If you have concerns with your treatment plan, speak up. Your doctor may be able to adjust your dosage or offer different treatment options. It is always your right to seek a second opinion.

ANCA-ASSOCIATED VASCULITIS: EOSINOPHILIC GRANULOMATOSIS WITH POLYANGIITIS (formerly Churg-Strauss syndrome)

Living with EGPA

Living with a chronic condition such as EGPA can be overwhelming at times. Fatigue, pain, emotional stress, and medication side effects can take a toll on your sense of well-being, affecting relationships, work and other aspects of your daily life. Sharing your experience with family and friends, connecting with others through a support group, or talking with a mental health professional can help.

Outlook

There is no cure for EGPA at this time, but with early diagnosis and proper treatment, many patients can lead full, productive lives. The majority of EGPA patients respond well to treatment. Because relapses are common with EGPA, follow-up medical care is essential.

Clinical studies are ongoing at multicenter research centers, including in the USA the Vasculitis Clinical Research Consortium (VCRC), to better understand the risk factors and causes of vasculitis, investigate more effective and safer treatments, and work toward a cure. The Vasculitis Foundation encourages patients to consider participating in clinical research studies to help further understanding of vasculitis. Patients are also encouraged to join the Vasculitis Patient Powered Research Network (VPPRN), where they can provide valuable disease insight and information.

ANZVASC, the Australian and New Zealand Vasculitis Society, was formed in late 2018 to promote collaboration and excellence in vasculitis care and research in Australia and New Zealand. Patients and consumers are welcome to participate and can join as associate members.

For more information on vasculitis and vasculitis research, visit:

www.anzvasculitis.org

www.vasculitisfoundation.org

**ANZVASC thanks the Vasculitis Foundation for permission to use this information, with minor changes
for use in Australia and New Zealand**

ANCA-ASSOCIATED VASCULITIS: EOSINOPHILIC GRANULOMATOSIS WITH POLYANGIITIS (formerly Churg-Strauss syndrome)

About Vasculitis

Vasculitis is a family of nearly 20 rare diseases characterized by inflammation of the blood vessels, which can restrict blood flow and damage vital organs and tissues. Most forms of vasculitis are autoimmune diseases, that occur when the body's natural defense system mistakenly attacks healthy tissues. Triggers may include infection, medication, genetic or environmental factors, allergic reactions, or another disease. However, the exact cause is often unknown.

A Family of Diseases

- Anti-GBM (Goodpasture's) disease
- Aortitis
- Behcet's syndrome
- Central nervous system vasculitis
- Cogan's syndrome
- Cryoglobulinemia
- Cutaneous small-vessel vasculitis (formerly hypersensitivity/leukocytoclastic)
- Eosinophilic granulomatosis with polyangiitis (EGPA, formerly Churg-Strauss syndrome)
- Giant cell arteritis
- Granulomatosis with polyangiitis (GPA, formerly Wegener's)
- IgA vasculitis (Henoch-Schönlein Purpura)
- Kawasaki disease
- Microscopic polyangiitis
- Polyarteritis nodosa
- Polymyalgia rheumatica
- Rheumatoid vasculitis
- Takayasu's arteritis
- Urticarial vasculitis

About the Vasculitis Foundation

The Vasculitis Foundation (VF), based in the USA, is the leading organization in the world dedicated to diagnosing, treating, and curing all forms of vasculitis. The VF provides a wide range of education, awareness and research programs and services. To learn more, and get the most updated disease and treatment information, visit www.vasculitisfoundation.org

The VF on Social Media includes:

- Instagram: [vasculitisfoundation](https://www.instagram.com/vasculitisfoundation)
- Twitter: [@VasculitisFound](https://twitter.com/VasculitisFound)
- VF Facebook Discussion Group: www.facebook.com/groups/vasculitisfoundation

Ways to Get Involved

- Participate in research
- Join social media
- Host or participate in an event
- Attend a regional conference and/or symposium

ANCA-ASSOCIATED VASCULITIS: EOSINOPHILIC GRANULOMATOSIS WITH POLYANGIITIS (formerly Churg-Strauss syndrome)

- Give a donation toward long-term solutions
- Become an advocate for yourself, or for others
- Share your journey

Vasculitis Foundation Mission

Building upon the collective strength of the vasculitis community, the Foundation supports, inspires and empowers individuals with vasculitis, and their families, through a wide range of education, research, clinical, and awareness initiatives.

The VPPRN and VCRC

The Vasculitis Patient-Powered Research Network (VPPRN) seeks to improve the care and health of patients with vasculitis by exploring research questions that matter most to patients, and by advancing medical knowledge about vasculitis. For more information, visit: www.VPPRN.org. The Vasculitis Clinical Research Consortium (VCRC) is an integrated group of academic medical centers, patient support organizations, and clinical research resources dedicated to conducting clinical research in different forms of vasculitis. For more information, visit: www.rarediseasesnetwork.org/cms/vcrc

Vasculitis Foundation

PO Box 28660, Kansas City, Missouri, 64188-8660. USA

www.VasculitisFoundation.org

Email: vf@vasculitisfoundation.org

ANZVASC

The Australian and New Zealand Vasculitis Society, was formed in late 2018 to promote collaboration and excellence in vasculitis care and research in Australia and New Zealand.

Patients and consumers are critical in the work of ANZVASC and are most welcome to participate and join as associate members.

ANZVASC is a registered health promotion charity with the Australian Charities and not for Profit Commission as has Deductible Gift Recipient Status

- www.anzvasculitis.org
- anzvasc@gmail.com
- Twitter: @ANZVASC

The VF gratefully acknowledges Alexandra Villa-Forte, MD, MPH, Cleveland Clinic, for her expertise and contribution to this brochure. This brochure was supported by an unrestricted education grant from Genentech to the Vasculitis Foundation. The Vasculitis Foundation is solely responsible for all content. The Vasculitis Foundation (VF) is a 501(c)(3) nonprofit organization governed by a Board of Directors and advised on medical issues by a Medical and Scientific Advisory Board. VF's educational materials are not intended to replace the counsel of a physician. VF does not endorse any medications, products or treatments for vasculitis, and advises you to consult a physician before initiating any treatment.

(Updated October 2018, ANZVASC [R Kitching] July 2019)