

GENERAL VASCULITIS

What is vasculitis?

Vasculitis is a general term that refers to inflammation of the blood vessels. It is used to describe a family of nearly 20 rare diseases, characterized by narrowing, weakening or scarring of the blood vessels, which can restrict blood flow and damage vital organs and tissues.

Vasculitis can affect any of the blood vessels of the body, including arteries, veins and capillaries. Symptoms depend on the organs and tissues affected, and can vary from person to person. Some forms of the disease are mild and may improve on their own, while others involve critical organ systems and may require lifelong medical care. Early diagnosis and treatment are extremely important to avoid potentially life-threatening complications.

It is common for people with vasculitis to experience periods of relapse and remission, so regular doctor visits and follow-up monitoring are recommended. Proper treatment and ongoing medical care can improve the quality of life and prognosis for people with vasculitis.

What causes vasculitis?

The cause of vasculitis is not fully understood by researchers. Vasculitis is classified as an autoimmune disorder, which occurs when the body's natural defense system mistakenly attacks healthy tissues. Researchers believe a combination of factors may trigger the inflammatory process, including infections, medications, genetic or environmental factors, allergic reactions, or another disease. However, the exact cause is usually unknown.

Who gets vasculitis?

Vasculitis can affect people of all ages and races, although some forms may be more common among certain age or ethnic groups. Vasculitis usually, but not always, affects women and men in equal numbers.

Types of vasculitis

There are many types of vasculitis, which are classified by the size and location of affected blood vessels. Your doctor will help determine the type of vasculitis you have and the most appropriate treatment (See table on Page 2).

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MOST COMMON TYPES OF VASCULITIS (Classified by vessel size)

Large vessel	<ul style="list-style-type: none"> • Aortitis • Giant cell arteritis • Polymyalgia rheumatica • Takayasu's arteritis
Medium vessel	<ul style="list-style-type: none"> • Kawasaki disease • Polyarteritis nodosa
Small vessel	<ul style="list-style-type: none"> • Anti-GBM (Goodpasture's) disease • Cryoglobulinemia • Cutaneous small-vessel vasculitis (formerly called hypersensitivity) • IgA vasculitis (Henoch-Schönlein purpura) • Urticarial vasculitis (hypocomplementemic)
Small- and medium-sized vessel	<ul style="list-style-type: none"> • Central nervous system angiitis • Eosinophilic granulomatosis with polyangiitis (EGPA/Churg-Strauss) • Granulomatosis with polyangiitis (formerly Wegener's) • Microscopic polyangiitis • Rheumatoid vasculitis
Arteries of various sizes	<ul style="list-style-type: none"> • Behcet's syndrome • Cogan's syndrome

What are the symptoms of vasculitis?

Vasculitis symptoms vary from patient to patient and depend on the type of vasculitis and affected tissues and organs. Symptoms may include:

- Fatigue/weakness
- Fever
- Muscle and/or joint pain
- Lack of appetite/weight loss
- Rashes or skin lesions
- Eye pain and redness/blurred vision
- Chronic nasal, ear and/or sinus problems
- Shortness of breath
- Cough (or coughing up blood)
- Abdominal pain
- Severe headaches
- Nerve problems, such as numbness, weakness, pain (neuropathy)
- Bloody or dark-colored urine, potentially indicating kidney problems (Note: A patient can have kidney disease without having symptoms; therefore, patients with vasculitis of any form should have regular urine tests.)

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Complications

Serious vasculitis complications can occur, especially if the disease goes undiagnosed or untreated. Depending on the type of vasculitis and severity of condition, complications can include organ damage or failure; blood clots; an aneurysm (an abnormal bulging of a weakened blood vessel that can burst); heart problems; vision loss; and neuropathy, among others. If you have the above symptoms, or others that you are concerned about, report them to your doctor as soon as possible.

How is vasculitis diagnosed?

Diagnosing vasculitis can pose a challenge because the symptoms may be similar to those caused by other illnesses or diseases. Your doctor will take a detailed medical history and perform a physical exam. Depending on symptoms and the type of vasculitis suspected, your doctor may order laboratory work such as urinalysis and blood tests; imaging studies such as X-rays, computed tomography (CT), or magnetic resonance imaging (MRI) scans; lung function tests; or biopsy, when indicated.

A biopsy involves surgical removal of a small sample of affected organ or tissue, which is analyzed for signs of inflammation or tissue damage. A biopsy is usually obtained to confirm diagnosis, however it is not always feasible. In addition, a positive biopsy is not always a requirement to confirm the diagnosis before starting treatment.

How is vasculitis treated?

Treatment is based on numerous factors including the specific type of vasculitis, symptoms, organs affected, disease severity, lab results, age, overall health and more. It is essential to work closely with your doctor in developing a comprehensive treatment plan.

Treatment usually involves two phases: controlling the inflammation to achieve remission, and maintenance treatment to prevent relapse. Common treatments include the following:

- Corticosteroids such as prednisone are often the first line of treatment for vasculitis, to reduce inflammation.
- For more serious forms of vasculitis, medications that suppress the immune system are often prescribed, including methotrexate, azathioprine, mycophenolate mofetil, and cyclophosphamide.
- Biologic agents such as rituximab, tocilizumab, and mepolizumab may be prescribed for specific types of vasculitis. Biologic medications are complex proteins derived from living organisms. They target certain parts of the immune system to control inflammation.
- For very severe cases, other additional treatments include plasmapheresis (plasma exchange), intravenous gamma globulin, or surgery to restore blood flow.

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Side effects

All the medications used to treat vasculitis have side effects. These including lowering your body's ability to fight infection, potential bone loss (osteoporosis), and others. Your doctor may prescribe medications to offset these side effects. Infection prevention is also very important. Talk to your doctor about getting a flu shot, pneumonia vaccination, and/or shingles vaccination, which can reduce your risk of infection.

Relapse

Even with effective treatment, relapses of vasculitis are common. Regular doctor visits and ongoing monitoring of lab and imaging tests are important in detecting relapses early.

Your medical team

Effective treatment of vasculitis often requires the coordinated efforts and ongoing care of a team of medical providers and specialists. In addition to a primary care provider, patients may need to see: a rheumatologist (joints, muscles, immune system); dermatologist (skin); pulmonologist (lungs); gastroenterologist (digestive system); otolaryngologist (ear, nose and throat); immunologist (immune system); nephrologist (kidneys); cardiologist (heart); neurologist (brain/and nervous system); or others as needed.

Living with vasculitis

Coping with vasculitis 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 vasculitis at this time, but with early diagnosis and proper treatment, many patients can lead full, productive lives. Outlook depends on a number of factors, including the form of vasculitis; affected organs; severity of disease; how soon it is diagnosed and treated; and whether there is an underlying condition, among others. Most forms of vasculitis are chronic, with periods of relapse and remission. In addition, medications used to treat vasculitis carry the risk of side effects, so follow-up medical care is essential.

Clinical studies are ongoing at multicenter research centers, including 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.

For more information on vasculitis research, visit: www.vasculitisfoundation.org/research

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About Vasculitis

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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) 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

Connect with the VF on Social Media

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

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Ways to Get Involved

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

Join the VPPRN!

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

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