

# What is Vasculitis?

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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.

## Causes

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.

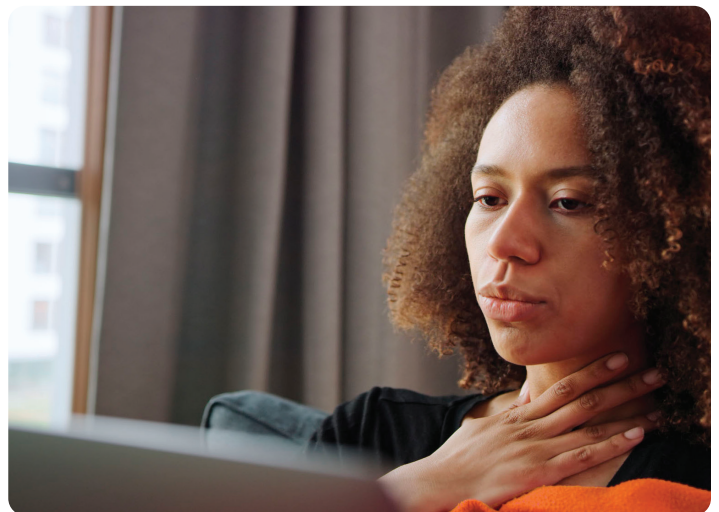
## Who Gets Vasculitis?

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.

## Symptoms

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, sinus and/or ear problems
- ▶ Shortness of breath
- ▶ Cough (or coughing up blood)
- ▶ Abdominal pain
- ▶ Severe headaches
- ▶ Nerve problems (numbness, weakness, pain)
- ▶ Bloody or dark-colored urine, potentially indicating kidney problems (Note: A patient can have kidney disease without having symptoms; therefore, patients with vasculitis should have regular urine tests.)



# Vasculitis

## Types of Vasculitis

There are many types of vasculitis, which are classified by the predominant size of involved blood vessels. Your doctor will help determine the type of vasculitis you have and the most appropriate treatment.

### MOST COMMON TYPES OF VASCULITIS (Classified by vessel size)

<b>Large vessel</b>	Aortitis
	Giant cell arteritis (GCA)
	Polymyalgia rheumatica (PMR)
	Takayasu arteritis (TAK)
<b>Medium vessel</b>	Kawasaki disease
	Polyarteritis nodosa (PAN)
<b>Small and medium sized vessel</b>	Cutaneous small-vessel vasculitis (CSVV, formerly hypersensitivity/leukocytoclastic)
	Eosinophilic granulomatosis with polyangiitis (EGPA, formerly Churg-Strauss syndrome)
	Granulomatosis with polyangiitis (GPA)
	Microscopic polyangiitis (MPA)
	Primary angiitis of the central nervous system (PACNS)
	Rheumatoid vasculitis
	Anti-GBM disease (formerly Goodpasture's syndrome)
	Cryoglobulinemic vasculitis
	IgA vasculitis (formerly Henoch-Schönlein purpura)
	Urticarial vasculitis (normocomplementemic or hypocomplementemic)
Central nervous system vasculitis (CNSV)	
<b>Arteries of various sizes</b>	Behçet's syndrome
	Cogan's syndrome

## Complications

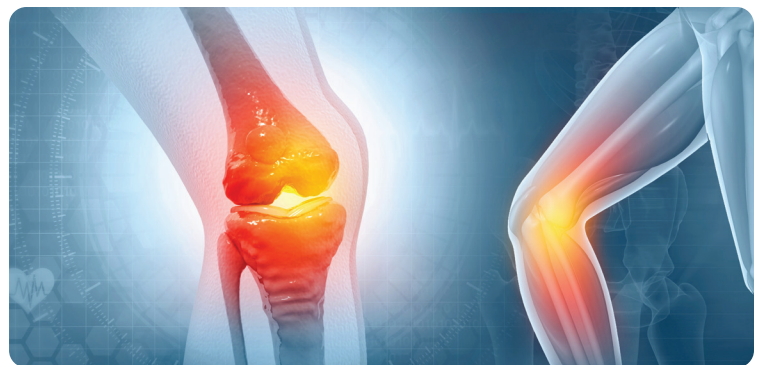
Serious vasculitis complications can occur, especially if the disease goes undiagnosed or untreated. Depending on the type of vasculitis and severity of condition, complications can include organ damage or failure, blood clots, an aneurysm (an abnormal bulging of a weakened blood vessel that can burst), heart problems, vision loss, neuropathy, and lung bleeding, 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 your symptoms and the type of vasculitis suspected, your doctor may order laboratory tests such as urinalysis and blood tests, imaging studies such as X-rays, computed tomography (CT) or magnetic resonance imaging (MRI) scans, or lung function tests. A biopsy of the affected tissue or organ is usually obtained to confirm diagnosis but is not always feasible. Biopsy is not always required to confirm the diagnosis before starting treatment, which shouldn't be delayed.

## How is Vasculitis Treated?

Treatment is based on numerous factors including the specific type of vasculitis, symptoms, organs affected, disease severity, laboratory test 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. Corticosteroids are also immunosuppressive medications.
- ▶ For more serious forms of vasculitis, other 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.



## Side Effects of Treatment

All the medications used to treat vasculitis have side effects. These include 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 vaccines (e.g., flu shot, pneumonia and/or shingles vaccination), which can reduce your risk of infection.

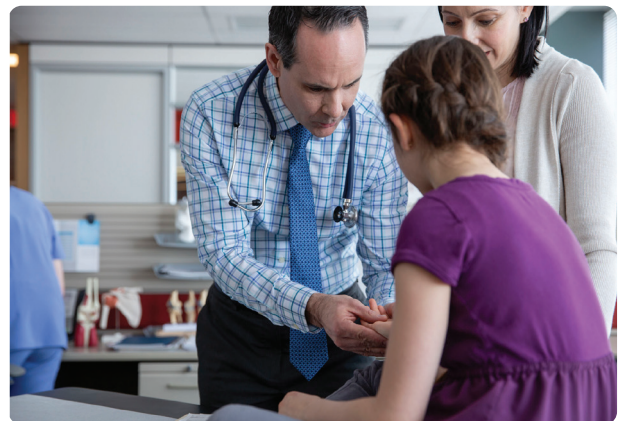
## Medical Follow-up/Relapse

Even with effective treatment, relapses of vasculitis are common. Regular doctor visits and ongoing monitoring of laboratory 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/nervous system) or others as needed





# Vasculitis

## 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 several 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.

## About the VF

The VF is the leading organization in the world dedicated to diagnosing, treating, and curing all forms of vasculitis. The 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. The VF does not endorse any medications, products, or treatments for vasculitis, and advises you to consult a physician before initiating any treatment.

The VF gratefully acknowledges Alexandra Villa-Forte, MD, Cleveland Clinic Center for Vasculitis Care and Research, for her expertise and contribution to this brochure.

To access additional VF support and educational resources, please scan the QR code below.



## VF 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.



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