



Vasculitis

Vasculitis refers to inflammation of blood vessels. There are multiple types of vasculitis. Most types of vasculitis are rare and the causes are generally unknown. Vasculitis can affect persons of both sexes and a broad range of ages from children to older adults.

Fast Facts

- Vasculitis is a general term for a group of rare diseases that have in common inflammation of blood vessels.
- There are many types of vasculitis, and the different diseases may vary significantly in terms of symptoms, severity, and duration.
- The severity of vasculitis can range from mild to extremely serious and life-threatening.
- Early diagnosis and treatment of severe vasculitis can prevent permanent damage.
- Glucocorticoids (prednisone and others) are the cornerstone of treatment.
- Other medications that suppress the immune system are also frequently used in severe disease or to allow lower doses of glucocorticoids to be used.

What is vasculitis?

Vasculitis refers to inflammation of blood vessels. The wide range of symptoms seen in vasculitis reflects the potential for damage from poor blood flow to a variety of affected tissues throughout the body. For example, vasculitic damage to the lungs may cause shortness of breath and cough; damage to nerves may lead to numbness or weakness in a hand or foot; and damage to skin may produce red spots (purpura), nodules, or ulcers. On the other hand, vasculitis of the kidneys may produce no symptoms at first but is a quite serious problem.



Vasculitic diseases can be mild or life-threatening, and either limited to a single episode, or multiple ones.

Most patients with vasculitis can be diagnosed with a specific vasculitic disease based on the size of the affected blood vessels and the organs most commonly involved (see table). However, for some patients, a specific type of vasculitis cannot be determined.

Most types of vasculitis are rare.

Types of Vasculitis	
Largest arteries: aorta and major branches	Giant cell arteritis Takayasu's arteritis Aortitis in Cogan's syndrome Aortitis in spondyloarthropathies Isolated aortitis
Medium-sized arteries	Kawasaki's disease Polyarteritis nodosa
Small and medium-sized arteries	ANCA-associated vasculitis Wegener's granulomatosis Microscopic polyangiitis Churg-Strauss syndrome Primary angiitis of the central nervous system
Small arteries	Henoch-Schönlein purpura Vasculitis associated with rheumatoid arthritis , lupus , Sjögren's syndrome Cryoglobulinemic vasculitis Goodpasture's syndrome Drug-induced vasculitis
Arteries and veins of various sizes	Behcet's disease Relapsing polychondritis

This list includes the most frequently diagnosed forms of vasculitis but is not complete and does not include some forms of vasculitis associated with other diseases such as cancer-associated vasculitis or



Types of Vasculitis

come types of infection-associated vasculitis.

What causes vasculitis?

The causes of most types of vasculitis are not known. However, some cases of vasculitis are directly caused by reactions to medications. Additionally, some chronic infections, especially with hepatitis C virus or hepatitis B virus, can cause vasculitis. No other environmental exposures have been convincingly demonstrated to lead to vasculitis.

Vasculitis can also be a part of several other rheumatic diseases, especially [lupus](#), [rheumatoid arthritis](#), and [Sjögren's syndrome](#). However, most patients with vasculitis have none of these underlying problems. Vasculitis is not hereditary.

Who gets vasculitis?

A few forms of vasculitis are relatively common and affect particular groups of people. For example, Kawasaki's disease is only seen in children, and Henoch-Schoenlein purpura is much more common in children than adults.

On the other hand, [giant cell arteritis](#) occurs only in adults over 50 years of age, and becomes more common with older age.

Most forms of vasculitis are rare and affect persons of both sexes and a broad range of ages.

How is vasculitis diagnosed?

Physicians consider the possibility of vasculitis when a patient has symptoms, physical exam signs, and/or laboratory tests suggesting damage to one or more organ systems in the absence of another clear cause.

Methods to diagnose vasculitis depend on the size of the vessel involved. Small-vessel vasculitis is usually diagnosed by biopsy (for example, of skin or kidney). Medium-vessel vasculitis is diagnosed by either biopsy (for example, of skin, nerve, or brain) or angiography (an type of x-ray in which dye is used to look for abnormalities of blood vessels). Large-vessel vasculitis is also often diagnosed by angiography, although giant cell arteritis is usually diagnosed by biopsy of the temporal artery in the scalp.

A few forms of vasculitis, such as Behcet's disease and Kawasaki's disease, are usually diagnosed on the basis of a collection of clinical findings rather than biopsy or angiography. Some blood tests are so suggestive of a particular type of vasculitis that a positive test can sometimes allow for diagnosis without biopsy or angiography. The most useful of these tests is for anti-neutrophil cytoplasmic antibodies (ANCA); a positive ANCA test can be extremely helpful in establishing a diagnosis of [Wegener's granulomatosis](#), microscopic polyangiitis, or Churg-Stauss syndrome. Other laboratory tests



are important for showing damage to internal organs but the tests are not enough to establish a diagnosis of vasculitis.

How is vasculitis treated?

Glucocorticoids (prednisone, prednisolone, or other similar drugs), often referred to as “steroids,” are an important part of therapy for most forms of vasculitis, but dose and duration vary with the severity and chronicity of the disease.

Other immunosuppressive agents are used either because the disease is so severe that glucocorticoids alone are insufficient treatment, or because the long-term side-effects of these other drugs are less severe than those of glucocorticoids (“steroid-sparing”). [Cyclophosphamide](#) is the strongest of these immunosuppressive drugs and is used to treat severe disease that endangers vital organs. [Methotrexate](#), [azathioprine](#), and other drugs that are used more widely in treating other rheumatic diseases are also used to treat non-life-threatening vasculitis and are useful as steroid-sparing agents.

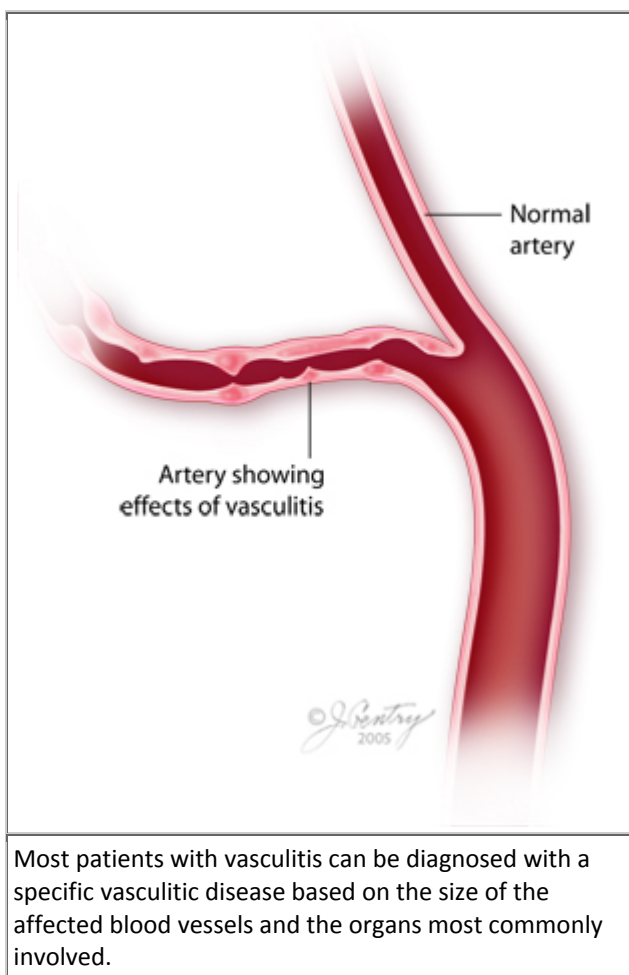
New drugs that are developed for treating other autoimmune and inflammatory diseases are often subsequently investigated in vasculitis. Plasma exchange (plasmapheresis) and intravenous immunoglobulin (IVIG) are sometimes used in severe cases of particular forms of vasculitis that are thought to be caused by circulating autoantibodies.

Finally, permanent damage from severe vasculitis sometimes requires surgical treatment, such as vascular bypass grafting, sinus surgery, or kidney transplantation.

Living with vasculitis

Vasculitis can be a temporary or a life-long problem, and symptoms and complications of vasculitis vary from being merely annoying, to disabling, to life-threatening.

Physicians often focus, with good reason, on preventing permanent damage to critical organ systems such as the lungs, kidneys, brain, and nerves. It is crucial to work to prevent death and long-term





disability from vasculitis, but other issues often trouble patients with vasculitis more on a day-to-day basis, including fatigue, pain, arthritis, nose and sinus problems, and many other problems. Side effects from medications, especially glucocorticoids, can be particularly troubling.

With current treatment strategies, the outcome for patients with vasculitis is often quite good. Patients with vasculitis are encouraged to try to live completely normal lives, but to remember that they may be at risk of recurrence of vasculitis, or at risk of infection if they need to remain on immunosuppressive medications.

Points to remember

- Vasculitic diseases are inflammatory conditions that often require treatment with immunosuppressive drugs including glucocorticoids.
- Several different types of vasculitis have been defined, most of which are rare.
- Diagnosis of vasculitis usually requires either biopsy of affected tissue or angiography, depending on the size of the affected blood vessel.

The rheumatologist's role in treating vasculitis

Rheumatologists are usually the specialists with the most overall knowledge about vasculitis and thus direct the care of patients, particularly those with chronic or severe disease. Patients with vasculitis often benefit greatly from seeing multiple specialists with expertise in the organ systems that are or might become affected. Such specialists have expertise in physical exam techniques, diagnostic testing, and procedures for diagnosis and treatment that are critical for optimal management of patients with vasculitis. Thus, many other medical specialists are often involved in the care of patients with vasculitis, including dermatologists, nephrologists, neurologists, ophthalmologists, otolaryngologists, pulmonologists, and others.

To find a rheumatologist

For a listing of rheumatologists in your area, [click here](#).

Learn more about [rheumatologists](#) and [rheumatology health professionals](#).

For more information

The American College of Rheumatology has compiled this list to give you a starting point for your own additional research. The ACR does not endorse or maintain these Web sites, and is not responsible for any information or claims provided on them. It is always best to talk with your rheumatologist for more information and before making any decisions about your care.

Vasculitis Foundation

www.vasculitisfoundation.org

Vasculitis Clinical Research Consortium (VCRC)

rarediseasesnetwork.epi.usf.edu

Churg-Strauss Syndrome Association

www.cssassociation.org



National Medical Research Foundation

www.nmrfoundation.com

Takayasu's Arteritis Research Association

www.takayasus.org

Polyarteritis Nodosa Support

www.pansupport.org

American Behcet's Disease Association

www.behcets.com

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Written by Paul A. Monach, MD, PhD and Peter A. Merkel, MD, MPH and reviewed by the American College of Rheumatology Patient Education Task Force.

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