



Vasculitis

Vasculitis refers to inflammation of the blood vessels. There are many types of vasculitis. Most types of vasculitis are rare, and the causes are generally not known. Vasculitis can affect persons of both sexes and a broad range of ages.

FAST FACTS

- Vasculitis is a term for a group of rare diseases that have in common inflammation of blood vessels.
- There are many types of vasculitis, and they may vary greatly in symptoms, severity and duration.
- Vasculitis can range from mild to life-threatening.
- Early detection and treatment of severe vasculitis can prevent permanent damage.
- Glucocorticoids (prednisone and others) are the main treatment.
- Patients also may be prescribed other medicines that suppress the immune system. These can help severe disease or let patients take lower doses of glucocorticoids.

WHAT IS VASCULITIS?

Vasculitis refers to inflammation of the blood vessels. These vessels include arteries and veins.

Vasculitis can result in poor blood flow to tissues throughout the body, such as the lungs, nerves and skin. Thus, vasculitis has a wide range of signs and symptoms (what you see and feel), such as:

- Shortness of breath and cough
- Numbness or weakness in a hand or foot
- Red spots on the skin (“purpura”), lumps (“nodules”) or sores (“ulcers”)



On the other hand, vasculitis of the kidneys may produce no symptoms at first but is still a serious problem.

Vasculitis can be mild or disabling, or even lead to death. Patients can have one episode of vasculitis or have repeated episodes over several years. Most types of vasculitis are rare.

WHAT CAUSES VASCULITIS?

We do not know what causes most types of vasculitis. You cannot inherit it. Vasculitis is thought to be an autoimmune disease, which means the body comes under attack by its own immune system. In vasculitis, the immune system attacks blood vessels.

Some cases of vasculitis are caused by reactions to medicines. Also, some chronic (long-term) infections, including with hepatitis C or hepatitis B virus, can cause vasculitis.

Vasculitis can be a part of other rheumatic diseases. These mainly include [systemic lupus erythematosus](#), [rheumatoid arthritis](#) and [Sjögren's syndrome](#). Most patients with vasculitis have none of these diseases.

WHO GETS VASCULITIS?

Vasculitis affects persons of both sexes and all ages. A few forms of vasculitis affect certain groups of people. For instance, Kawasaki disease occurs only in children. Henoch-Schönlein purpura is much more common in children than adults. On the other hand, giant cell arteritis occurs only in adults over 50 years old.

HOW IS VASCULITIS DIAGNOSED?

Physicians suspect vasculitis when a patient has symptoms and abnormal results of the physical exam, lab tests or both, and there is no other clear cause.

The most common tests are:

- Biopsy—surgical removal of a small piece of tissue for inspection under a microscope
- Angiography—a type of X-ray to look for abnormalities of blood vessels
- Blood tests

For most patients, doctors can detect the type of vasculitis based on the size of the affected blood vessels (see Table) and the organs involved. To find small-vessel vasculitis, doctors most often do a biopsy, such as of skin or kidney. Detection of medium-vessel vasculitis happens by either biopsy (for instance, of skin, nerve or brain) or angiography. Angiography also is the test that often finds large-vessel vasculitis. This is not true, though, for detecting [giant cell arteritis](#). It tends to need biopsy of an artery in the scalp.

A few forms of vasculitis, such as Behçet's disease and Kawasaki disease, are usually detected on the basis of a collection of clinical findings rather than biopsy or angiography.



Some blood tests are so suggestive of a certain type of vasculitis that a positive (abnormal) test can be enough evidence to help doctors make the diagnosis. The most useful of these tests is for “antineutrophil cytoplasmic antibodies”—often referred to as ANCA. A positive ANCA test can help detect these types of vasculitis: [granulomatosis with polyangiitis \(Wegener’s\)](#), microscopic polyangiitis or Churg-Strauss syndrome. Other lab tests can show damage to organs, but the tests are not enough to prove vasculitis.

Types of Vasculitis	
Largest arteries: aorta and major branches	Giant cell arteritis Takayasu’s arteritis Aortitis in Cogan’s syndrome Aortitis in spondylarthropathies Isolated aortitis
Medium-sized arteries	Kawasaki disease Polyarteritis nodosa
Small and medium-sized arteries	ANCA-associated vasculitis Granulomatosis with polyangiitis (former name: Wegener’s granulomatosis) Microscopic polyangiitis Churg-Strauss syndrome Primary angiitis of the central nervous system
Small arteries	Henoch-Schönlein purpura Vasculitis related to rheumatoid arthritis, systemic lupus erythematosus and Sjögren’s syndrome Cryoglobulinemic vasculitis Goodpasture’s syndrome Drug-induced vasculitis
Arteries and veins of various sizes	Behçet’s disease Relapsing polychondritis
Abbreviation: ANCA, antineutrophil cytoplasmic antibodies.	
This list is not complete. It does not include some forms of vasculitis related to infection or diseases such as cancer.	



HOW IS VASCULITIS TREATED?

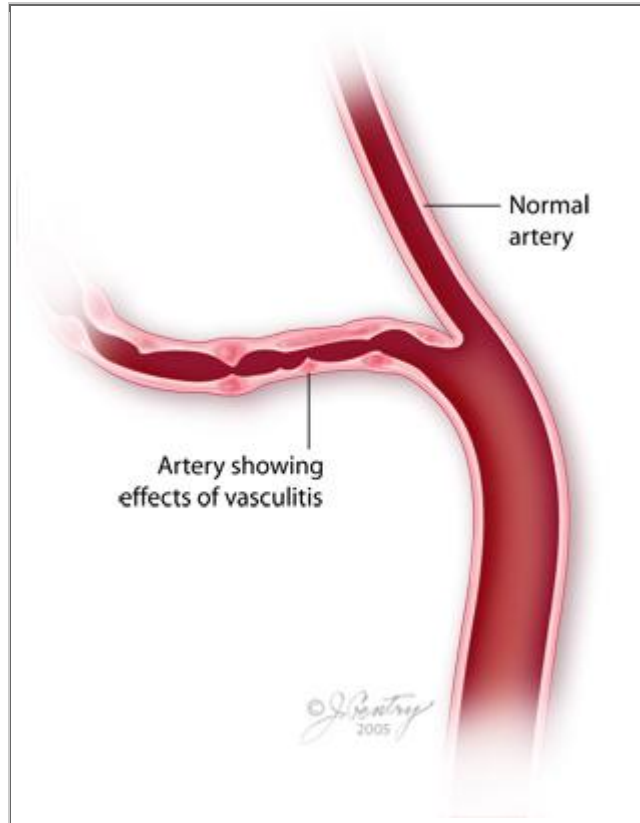
Glucocorticoids. Glucocorticoids (prednisone, prednisolone or others), often referred to as “steroids,” are an important part of treating most forms of vasculitis. The dose and length of treatment depend on how bad the disease is and how long the patient has had it. These drugs help reduce inflammation but can have long-term side effects.

Other drugs. Doctors sometimes prescribe immune-suppressing drugs because their side effects may be less serious than those of glucocorticoids. This is called “steroid-sparing” treatment. [Cyclophosphamide](#) is the strongest of these drugs, and doctors may prescribe it when severe disease endangers vital organs.

For less serious vasculitis, patients may receive [methotrexate](#), [azathioprine](#) or other immune-suppressing drugs. Doctors often prescribe these drugs to treat other rheumatic diseases, but they are useful for vasculitis, too.

Newer drugs designed to treat other autoimmune and inflammatory diseases may also help vasculitis. Researchers found that one of these drugs, [rituximab](#), effectively treats severe cases of certain forms of vasculitis. These include granulomatosis with polyangiitis, microscopic polyangiitis and cryoglobulinemic vasculitis. Some patients with the most severe cases of these diseases may receive plasma exchange (“plasmapheresis”) or intravenous immunoglobulin (often called “IVIg”).

Surgery. Damage from severe vasculitis sometimes requires surgery. This may involve vascular bypass grafting (a surgery to redirect blood flow around a blockage in a blood vessel). Depending on where the damage is, other possible operations are sinus surgery or a kidney transplant.



Doctors can detect the type of vasculitis in most patients with the disease based on the size of the affected blood vessels and damage to the organs most often involved.

LIVING WITH VASCULITIS

Vasculitis can be short term or lifelong. Doctors often focus, with good reason, on preventing permanent damage to vital organs (such as the lungs, kidneys and brain) and the nerves. It is crucial, of course, to prevent death and long-term disability from vasculitis. Yet, other issues often trouble patients. These include fatigue (feeling very tired), pain, arthritis, nose and sinus problems, and many other problems.

Side effects from medications, especially glucocorticoids, also can be troubling. Patients taking immunosuppressants are at risk of infections. Follow your doctor's advice on how to reduce your infection risk.

Fortunately, with current treatments, the outcome for patients with vasculitis is often good.

POINTS TO REMEMBER

- Vasculitic diseases are inflammatory health problems that often need treatment with immunosuppressive drugs. The most common medication used is glucocorticoids.
- Though there are many types of vasculitis, most are rare.
- Detection of vasculitis most often requires biopsy of affected tissue or angiography.

THE RHEUMATOLOGIST'S ROLE IN TREATING VASCULITIS

Rheumatologists are usually the specialists with the most overall knowledge about vasculitis. Thus, they direct the care of patients, particularly those with chronic or severe disease.

Patients with vasculitis often benefit from seeing experts in the organ systems that are or might become affected. Doctors that patients may need to see include a dermatologist (skin doctor), neurologist (expert in nervous system diseases), ophthalmologist (eye doctor), otolaryngologist (ear, nose and throat doctor), nephrologist (kidney doctor) or pulmonologist (lung doctor).

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
www.rarediseasesnetwork.org



Churg-Strauss Syndrome Association

www.cssassociation.org

National Medical Research Foundation

www.nmrfoundation.com

American Behçet's Disease Association

www.behcets.com

American College of Rheumatology Research and Education Foundation

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www.rheumatology.org/REF

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