

What is Vasculitis?

Vasculitis is a general term that refers to inflammation of the blood vessels. Inflammation of the blood vessel wall causes thickness of the vessel wall in small and medium size vessels, and in medium and large size vessels it can cause aneurysms. Thickness of the blood vessel wall usually results in narrowing or blockage of the vessel and aneurysms can result in vessel rupture and/or bleeding.

Vasculitis is used to describe a family of nearly 20 rare diseases, characterized by narrowing, weakening or scarring of the blood vessels. This can slow or stop the blood flow to vital organs and tissues.

Vasculitis can affect any of the blood vessels of the body, including:

- Arteries that take blood from the lungs and heart to the tissues
- Veins that bring the blood back to the lungs and heart
- Capillaries and the smallest blood vessels that feed the many tissues

What Causes Vasculitis?

The cause of vasculitis is not fully understood. 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 autoimmune 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.

MOST COMMON TYPES OF VASCULITIS (Classified by vessel size)

Large vessel	<ul style="list-style-type: none">• Aortitis• Giant cell arteritis• Polymyalgia rheumatica (when associated with GCA)• 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 (normo or hypocomplementemic)
Small- and medium-sized vessel	<ul style="list-style-type: none">• Central nervous system angiitis• Eosinophilic granulomatosis with polyangiitis (formerly Churg-Strauss)• Granulomatosis with polyangiitis (formerly Wegener's)• Microscopic polyangiitis• Rheumatoid vasculitis
Arteries of various sizes	<ul style="list-style-type: none">• Behçet's syndrome• Cogan's syndrome

What are the Symptoms of Vasculitis?

Vasculitis symptoms vary from patient to patient. They 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.

Some forms of the disease are mild and may improve on their own, while others involve critical organ systems and may require life-long medical care. Early diagnosis and treatment are extremely important to avoid potential life-threatening complications.

Complications

Serious vasculitis complications can occur, especially if the disease goes undiagnosed or untreated. Depending on the type of vasculitis and severity of the 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
- X-rays
- Computed tomography
- Magnetic resonance imaging scans
- Lung function tests
- Biopsy, when indicated

A biopsy involves surgical removal of a small sample of affected organ or tissue that is analyzed for signs of inflammation or tissue damage. A biopsy is usually obtained to confirm a

diagnosis; however, it is not always possible. 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: 1) controlling the inflammation to achieve remission, and 2) 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 sometimes used alone but most often in combination with other immunosuppressive medications.
- For most 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 may include plasmapheresis (plasma exchange), intravenous gamma globulin, or surgery to restore blood flow.

Side Effects of Treatments

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 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. They may include:

- Your primary care provider
- 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)
- Others as needed

The best way to manage your disease is to actively partner with your health care providers and get to know the members of the health care team.

It may be helpful to keep a health care journal to track your 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, if necessary, to provide a second set of ears and take notes.

Remember, it is up to you to be your own advocate. If you have concerns with the treatment plan, be sure to speak with the medical team. It is always your right to seek a second opinion.

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.

This can affect 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.

Your outlook depends on a number of factors, including:

- The form of vasculitis
- The affected organs
- The severity of disease
- How soon it is diagnosed and treated
- Whether there is an underlying condition, among others

Most forms of vasculitis are chronic – meaning it lasts a long time. There can be 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.

(Revised October 2020)