

## AORTITIS

### What is aortitis?

Aortitis is a form of vasculitis—a family of rare disorders characterized by inflammation of the blood vessels, which can restrict blood flow and damage vital organs and tissues. Aortitis refers to inflammation of the aorta, which carries oxygenated blood from the heart to the rest of the body. It is seen in a variety of conditions, including certain forms of vasculitis, other inflammatory conditions, and infections. When aortitis occurs in isolation without an underlying cause, it is called “isolated aortitis.” Aortitis is a serious condition that can lead to pain and weakness of the arms and legs, kidney failure, stroke, heart failure, and heart attack.

Most symptoms of aortitis are associated with the underlying disease. They include back pain, abdominal pain and fever, along with headaches, weakness, weight loss, joint pain, chest pain, shortness of breath, fainting, visual disturbances, and others. Aortic aneurysm, an abnormal bulge in the wall of the aorta, is a potentially life-threatening complication.

Treatment depends on whether the aortitis is caused by an infection or an underlying condition. Aortitis caused by infection is rare but can be life-threatening, and must be treated promptly with antibiotics. Aortitis caused by other inflammatory conditions or unknown reasons is typically treated with corticosteroids such as prednisone, and medications that suppress the immune system. In some patients, surgery is needed to repair aneurysms or bypass blocked arteries. Patients with aortitis require ongoing medical care, including repeated imaging of the aorta and its main vessel branches.

### Causes

The exact cause of aortitis is not fully understood by researchers. Vasculitis is classified as an autoimmune disorder—a disease which occurs when the body’s natural defense system mistakenly attacks healthy tissue.

The causes of aortic inflammation can be divided into three categories:

- **Noninfectious:** Caused by an underlying rheumatic disease—an umbrella term for inflammatory disorders that cause chronic pain of the joints, muscles and connective tissue. Some forms of vasculitis fall into this category, including giant cell arteritis (GCA) and Takayasu’s arteritis, Behcet’s syndrome, and Cogan’s syndrome. Other inflammatory disorders that can affect the aorta include systemic lupus erythematosus, rheumatoid arthritis, ankylosing spondylitis, sarcoidosis, and others.
- **Infectious:** Infections associated with aortitis include tuberculosis, salmonella, syphilis, Rocky Mountain spotted fever, herpes virus, hepatitis B and hepatitis C, among others.
- **Isolated:** This is inflammation of the aorta without any identifiable underlying cause. Other vessels are not affected.

### Who gets aortitis?

Prevalence of aortitis is not well-documented. It can affect both men and women of any age. In the United States and Europe, the incidence of aortitis is estimated at 1 to 3 new cases per million people per year. Isolated aortitis is very rare.

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### Symptoms

Symptoms of aortitis depend on the underlying condition causing the inflammation, and in early stages there may be no symptoms at all. Sometimes the aortic inflammation is discovered only at the time of surgery for an aneurysm.

General symptoms may include:

- Severe headaches that don't go away
- Back pain
- Chest pain
- Abdominal pain
- Fever

When the aortitis is associated with an underlying condition such as vasculitis or other rheumatic disorder, symptoms include:

- Headaches
- Tenderness of the scalp, especially the temples
- Fatigue
- Fever
- Pain and weakness of the arms and legs
- Chest pain
- Shortness of breath
- Loss of vision/double vision
- Unexplained weight loss
- Dizziness/fainting
- Difficulty with coordination and balance
- Night sweats
- Joint and/or muscle pain

### Complications

Inflammation of the aorta and its major arteries can lead to kidney failure, aortic aneurysm, stroke, heart failure, and heart attack. In cases where there is an underlying disease or infection causing the aortitis, complications are related to the specific cause.

### Diagnosis

In diagnosing aortitis, your doctor will consider a number of factors, including a detailed medical history; physical examination; laboratory tests; and specialized imaging studies. A physical exam may reveal abnormal heart sounds, and/or sounds over the major arteries; abnormalities (discrepancy) of blood pressure; difference in blood pressure between the arms and legs; or reduced or absent pulse in the wrists or ankles.

Determining the cause of the aortitis is critically important because the treatments vary depending on the source of the inflammation. For example, immunosuppressant medications typically used to treat vasculitis are not appropriate for aortitis caused by infection, as they can aggravate an active infection.

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Diagnosis of isolated aortitis involves ruling out diseases or conditions that can cause aortic inflammation and present with similar symptoms.

Depending on organs affected, the following diagnostic tests may be ordered:

- **Blood tests:** To detect abnormal levels of antibodies and proteins in the blood, which can indicate inflammation or infection. People with inflammation typically have elevated levels of C-reactive protein (CRP) and erythrocyte sedimentation rate (ESR, or “sed rate).
- **Imaging tests:** Echocardiogram uses ultrasound waves to examine the heart. Other tests include computed tomographic angiography (CTA), and magnetic resonance angiography (MRA), which reveal abnormalities of the aorta such as aneurysms or narrowing of the vessels. A dye-based angiography is rarely needed for diagnosis.
- **Tissue biopsy:** A biopsy involves surgical removal of tissue from an affected vessel, which is sent to a laboratory and analyzed for signs of inflammation. Biopsy of the aorta is not possible unless a surgical procedure is needed for aortic aneurysm.

### Treatment

The course of treatment for aortitis treatment depends on what is causing the inflammation. While infectious aortitis is rare, it can be life-threatening and requires prompt treatment with appropriate antibiotics, sometimes given intravenously.

In cases of isolated aortitis, and aortitis associated with systemic vasculitis or other autoimmune disorders, treatment is aimed at controlling the inflammation, typically with corticosteroids such as prednisone. Other treatments may include immunosuppressant drugs such as methotrexate, azathioprine, mycophenolate mofetil, and cyclophosphamide. Biologic agents such as infliximab or rituximab may be prescribed. Biologic medications are complex proteins derived from living organisms. They target certain parts of the immune system to control inflammation.

Surgery is sometimes needed to repair an aneurysm, or to bypass blocked arteries.

### Side effects

The medications used to treat aortitis have potentially serious side effects, such as lowering your body’s ability to fight infection, and potential bone loss (osteoporosis), among others. Therefore, it’s important to see your doctor for regular checkups. Medications may be prescribed to offset 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.

### Medical follow-up/relapse

Aortitis requires ongoing medical care and repeated imaging studies of the heart and aorta. Patients who have aortitis may experience relapse of their vasculitis symptoms, as most forms of vasculitis are chronic conditions. New or returning symptoms should be reported to a doctor as soon as possible.

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### **Your medical team**

Effective treatment of aortitis may require the coordinated efforts and ongoing care of a team of medical providers and specialists. In addition to a primary care provider, aortitis patients may need to see the following specialists: rheumatologist (joints, muscles, immune system); infectious disease specialist; cardiologist (heart); cardiovascular surgeon; ophthalmologist (eyes); or others as needed.

The best way to manage your disease is to actively partner with your health care providers. 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's up to you to be your own advocate. If you have concerns with your treatment plan, speak up. Your doctor may be able to adjust your dosage, or offer different treatment options. It is always your right to seek a second opinion.

### **Living with aortitis**

Living with a chronic condition such as aortitis 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**

Aortitis is a serious condition. The outlook for patients with this condition depends on whether the aortitis is caused by an infection or another underlying condition, and also how quickly the disease is diagnosed and treated. Patients with aortitis may experience relapse of their vasculitis symptoms. Aortitis requires ongoing medical care and repeated imaging studies of the heart and aorta.

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](http://www.vasculitisfoundation.org/research)

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### **About Vasculitis**

Vasculitis is a family of nearly 20 rare diseases characterized by inflammation of the blood vessels, which can restrict blood flow and damage vital organs and tissues. Vasculitis is classified as an autoimmune disorder, which occurs when the body's natural defense system mistakenly attacks healthy tissues. Triggers may include infection, medication, genetic or environmental factors, allergic reactions, or another disease. However, the exact cause is often unknown.

### **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](http://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](https://www.facebook.com/groups/vasculitisfoundation)

### **Ways to Get Involved**

- Participate in research
- Join social media
- Host or participate in an event

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- 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](http://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](http://www.rarediseasesnetwork.org/cms/vcrc)

### **Vasculitis Foundation**

PO Box 28660

Kansas City, Missouri 64188-8660

USA

Phone: 816.436.8211

Toll-free: 800.277.9474

Fax: 816.656.3838

[www.VasculitisFoundation.org](http://www.VasculitisFoundation.org)

Email: [vf@vasculitisfoundation.org](mailto:vf@vasculitisfoundation.org)

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