

## **What is Aortitis?**

Aortitis is a form of vasculitis—which is a group of rare disorders characterized by inflammation of the blood vessels. Inflammation of blood vessels may result in aneurysms or narrowing of the vessel, which can block or slow down blood flow to vital organs and tissues.

Aortitis refers to inflammation of the aorta. The aorta carries blood rich in oxygen from the heart to the rest of the body. Aortitis can happen with an infection or other forms of vasculitis, and when it occurs by itself, it is called “isolated aortitis.” Aortitis is a serious condition and needs to be managed.

## **Who Gets Aortitis?**

Who and how many people having aortitis is not well-documented. It can affect both men and women of any age.

## **Causes**

The exact cause of aortitis is not fully understood. 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 aortitis can be divided into three categories:

### **Noninfectious causes:**

- Rheumatic diseases, such as rheumatoid arthritis, ankylosing spondylitis or lupus
- Some forms of vasculitis, such as Behçets’ syndrome, giant cell arteritis, and others
- Other systemic inflammatory disorders, such as sarcoidosis and others

### **Infectious causes:**

- Infections associated with aortitis include tuberculosis, syphilis, hepatitis B and C, among others.

## Isolated aortitis:

- This is inflammation of the aorta without any identifiable underlying cause. Other vessels are not affected.

## Symptoms

Symptoms of aortitis depend on the underlying condition causing the inflammation, and in early stages there may be no symptoms at all.

General symptoms may include:

- Back pain
- Chest pain
- Abdominal pain
- Fever

When the aortitis is associated with an underlying condition such as vasculitis or another 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

Aortitis and inflammation of the 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 try to determine what is causing it by performing a:

- **Detailed medical history**—are there genetic or predisposing factors?

- **Physical examination**, which may reveal abnormal heart sounds, and/or sounds over the major arteries; difference in blood pressure between the arms and legs; or reduced or absent pulses in the arms and/or legs.

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.
- **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 depends on what is causing the inflammation. In cases of isolated aortitis, and aortitis associated with systemic vasculitis or other autoimmune disorders, treatments may include:

### Medications to control inflammation:

- **Corticosteroids** such as prednisone.
- **Immunosuppressant** drugs such as methotrexate or azathioprine, which keep the immune system from attacking healthy tissue.
- **Biologic agents** such as infliximab or rituximab may be prescribed. They target certain parts of the immune system to control inflammation.

### Medication to control infection:

- Aortitis caused by infection is rare but can be life-threatening, and must be treated promptly with antibiotics.

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

## Side Effects of Treatment

The medications used to treat aortitis have potentially serious side effects, such as:

- Lowering your body's ability to fight infection
- Potential bone loss (osteoporosis), among others

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.

## **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)
- 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. 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.

## **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.

## **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.

(Revised October 2020)