VASCULITIS FOUNDATION
Support | Awareness | Research
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TAKAYASU'S ARTERITIS

What is Takayasu's arteritis (TAK)?

Takayasu's arteritis (TAK) 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. TAK affects the aorta and its main branches, which carry blood from the heart to the rest of the body. It most commonly occurs in females.

TAK may or may not cause noticeable symptoms in the initial stage of the disease. When early symptoms are present, people may feel generally unwell, with symptoms of mild fever, fatigue, aches and pains, and poor appetite. As the disease progresses, TAK can lead to headaches, chest pain, shortness of breath, high blood pressure, weakness, and light-headedness, among other symptoms.

Early detection and treatment can help lower the risk of serious complications. TAK is usually treated with corticosteroids such as prednisone, and medications that suppress the immune system to control inflammation. Even with treatment, TAK is a chronic condition with periods of relapse and remission, so ongoing medical care and monitoring are necessary.

Causes

The cause of TAK 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 tissues. Researchers believe an infection may set the inflammatory process in motion, but this is not yet proven. Environmental and genetic factors may also play a role.

Who gets Takayasu's arteritis?

TAK primarily affects Asian women and teen-aged girls. Nine out of 10 patients are female, with age of onset between 15 and 40 years old. However, the disease can affect children and adults of both sexes, and all races and ethnic groups. Prevalence is estimated at 1 in 200,000 people.

Symptoms

Symptoms of TAK tend to occur in two stages. In the first stage, patients may feel generally ill, with fatigue, fever, muscle or joint pain, and/or rapid weight loss. However, some may not experience symptoms for months or years—even as inflammation causes damage to blood vessels and organs.

Second-stage symptoms include:

- Pain or weakness with use of limbs
- Lightheadedness, dizziness, fainting
- Headaches
- Visual disturbances
- High blood pressure (or difference in blood pressure between arms and/or legs)
- Diminished or absent pulse in the wrists or ankles
- Anemia (which can cause weakness)
- Chest pain
- Shortness of breath/fatigue

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Seek medical attention right away if you experience signs of a heart attack (chest or arm pain, shortness of breath, nausea/vomiting); or signs of a stroke (face drooping, arm weakness or difficulty speaking). See your doctor for other symptoms listed above that you may be concerned about.

Diagnosis

There is no single test for diagnosing TAK, so your doctor will consider a number of factors, including symptoms, medical history, physical exam findings, laboratory tests, and specialized imaging studies. For many forms of vasculitis, a biopsy—surgical removal and analysis of suspected tissue—confirms the diagnosis. However, biopsy is considered risky with large vessels such as the aorta, and is not done unless the patient is undergoing vascular surgery.

Physical exam: Abnormal exam findings may raise suspicion of TAK: a weak or absent pulse in the limbs and organs (TAK is sometimes called the "pulseless disease"); low blood pressure; different blood pressure between arms; or sounds called "bruits" heard over the large arteries with a stethoscope.

Blood tests: Two common blood tests that can help detect inflammation include the erythrocyte sedimentation rate (ESR) test, commonly called the "sed rate"; and the C-reactive liver protein test (CRP). These tests, however, are not conclusive on their own and may be normal in up to 50 percent of patients with TAK.

Imaging: Non-invasive tests such as magnetic resonance angiogram (MRA) and computed tomography angiography (CTA) are usually enough to establish the diagnosis. An angiogram (an X-ray of a blood vessel that has been injected with a special contrast dye) is rarely needed. Imaging tests can show narrowing or blockage of the vessels, or the formation of an aneurysm—an abnormal bulge in the blood vessel that can burst, with life-threatening consequences.

Your doctor may order other imaging tests as indicated, including the Doppler ultrasound, and positron emission tomography (PET) scan.

Treatment

Goals of treatment include controlling inflammation of the blood vessels, and preventing further damage. For some patients, the disease may go into remission and medication can be tapered or stopped. But for others, the disease may need to be controlled with medicine on a long-term basis. In severe cases, surgery may be required to bypass blocked vessels.

Corticosteroids: Initial treatment typically consists of a steroid such as prednisone, starting at a high dose and then tapered down to minimize side effects.

Immunosuppressants: Medications that suppress the immune system may be prescribed to achieve remission and reduce the need for prednisone. They include methotrexate, which is commonly used to treat rheumatoid arthritis; azathioprine; and mycophenolate mofetil.

Biologics: Those with severe disease who don't respond to traditional treatments may be prescribed biologics. (Biologic medications are complex proteins derived from living organisms. They target certain parts of the immune system to control inflammation.) Among these, tocilizumab and infliximab have

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been used in TAK. Some studies show these drugs are effective at controlling inflammation and reducing the need for steroids in treating TAK, but more research is needed.

Surgery: Vascular surgery may be required in cases where arteries have become severely narrowed or blocked, or to repair aneurysms.

Complications

Even with treatment, damage to the aorta may be permanent. Repeated swelling and healing of arteries can lead to complications. Therefore, it is important to follow up with your doctor on a regular basis. Complications can include:

- Hardening and narrowing of the blood vessels
- High blood pressure
- Heart valve disorder (eg, aortic valve damage)
- Heart failure
- Aortic aneurysm
- Transient ischemic attack (TIA), a "mini-stroke" that doesn't cause permanent damage but serves as a warning sign
- Stroke
- Heart attack
- Pulmonary artery problems

Pregnancy: Successful pregnancies are possible for patients with TAK. If you are pregnant, or planning to become pregnant, talk to your doctor about potential effects of the disease and treatments.

Side effects

The medications used to treat TAK 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.

Relapse

Even with effective treatment, relapses can occur in individuals with TAK. If initial symptoms return or new ones develop, report them to your doctor as soon as possible. Regular check-ups and ongoing monitoring of lab and imaging tests are important in detecting relapses early.

Your medical team

Effective treatment of TAK may require the coordinated efforts and ongoing care of a team of medical providers and specialists. In addition to a primary care provider, you may need to see: a rheumatologist (joints, muscles, immune system); cardiologist (heart doctor); vascular surgeon; or other specialists.

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The best way to manage your disease is to actively partner with your health care providers. Get to know the members of your health care team. It may be helpful to keep a health care journal to track 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 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 Takayasu's arteritis

Coping with TAK 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

There is no cure for Takayasu's arteritis at this time, but with early detection, proper treatment and ongoing monitoring, most patients have a good prognosis. Imaging tests such as MRA and CTA scans remain the diagnostic tools of choice for this chronic disease, and may be used to monitor the blood vessels during and after treatment.

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

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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

Connect with the VF on Social Media

- Instagram: vasculitisfoundation
- Twitter: @VasculitisFound
- VF Facebook Discussion Group: www.facebook.com/groups/vasculitisfoundation

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Ways to Get Involved

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

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

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