



Takayasu's Arteritis

Takayasu's Arteritis most often starts in young adults, but children and middle-aged people may get it, too

Takayasu's arteritis, also called TAK, is a rare disease involving inflammation in the walls of the largest arteries in the body: the aorta and its main branches. The disease results from an attack by the body's own immune system, causing inflammation in the walls of arteries. The inflammation leads to narrowing of the arteries, and this can reduce blood flow to many parts of the body.

TAK can result in a weak pulse or loss of pulse in arms, legs and organs. For this reason, people used to refer to the illness as "pulseless disease."

Fast Facts

- TAK is much more common in women than men.
- The disease most often starts in young adults, but children and middle-aged people may get it, too.
- Doctors find TAK on angiograms. Angiograms are types of X-ray tests that look at arteries. In TAK, angiograms show narrowing of large arteries.
- Narrowed or blocked arteries cause problems that range from mild to serious.
- Treatment of TAK almost always includes glucocorticoids (prednisone and others), which help reduce the inflammation.
- Patients also may be prescribed other medications that suppress the immune system.

What is Takayasu's arteritis?

This disease is one of many types of vasculitis. [Vasculitis](#) refers to inflammation of blood vessels, and arteries are a type of blood vessel. In TAK, this inflammation occurs in the walls of large arteries: the aorta and its main branches. These blood vessels supply blood to the head, arms, legs and internal organs, such as the kidneys. Inflammation may cause the vessels' walls to thicken. With time, this thickening results in a narrowing inside the artery, called a "stenosis." If severe enough, such narrowing can reduce blood flow and result in less oxygen sent to the body parts or organs that the artery supplies.

Stenosis can cause symptoms (what you feel) and problems ranging from annoying to dangerous:

- Pain with use of an arm or leg (called "claudication")
- Dizziness, headaches or fainting
- Weakness and fatigue
- High blood pressure
- Chest pain
- Heart attack
- Stroke

Stenosis occurs slowly with time, and smaller vessels may grow and expand to carry blood around the blockage. These new vessels are called "collateral vessels." Collateral vessels may help prevent major organ damage.

Sometimes inflammation in the artery weakens the vessel wall, causing vessel expansion rather than narrowing (stenosis). This expansion is called an aneurysm (a bulge in the artery). The aorta as it emerges from the heart is one of the more common areas where an aneurysm can form. An aneurysm in the aorta might lead to heart valve dysfunction or rupture (bursting) of the aorta.

What causes Takayasu's arteritis?

As with most types of vasculitis, the cause of TAK is not known. It is rare to see more than one case in a family and the role of genetics is unclear. A link between TAK and an infection has also not been proven.

TAK is thought to be an autoimmune disease, which means that the body comes under attack by its own immune system. In TA, the immune system is attacking the blood vessels.

Who gets Takayasu's arteritis?

TAK is rare, affecting perhaps one in 200,000 people. It most often occurs in people ages 15–40 years, but sometimes affects younger children or middle-aged adults. Nine of 10 patients are female. TAK seems to be more common in East Asia, India and, perhaps, Latin America, than in other regions. Yet, it is rare even in these regions and occurs in a wide range of ethnic groups.

How Is Takayasu's arteritis diagnosed?

Doctors most often find TAK on an angiogram, a test that shows how well blood flows in arteries. A doctor often orders an angiogram when a patient has symptoms and abnormal results of the physical exam. These include loss of pulse or low blood pressure in an arm, or abnormal sounds ("bruits") heard over large arteries with a stethoscope.

There are various types of angiograms, including standard ones that involve injection of dye directly into an artery while X-ray test are taken. Less invasive types of angiography use another imaging technique such as computed tomography, and this is CT angiography or CTA. When MRI—magnetic resonance imaging—is used, it is called magnetic resonance angiography or MRA.

Angiograms may show narrowing of one or more large arteries. It is important for the doctor to try to distinguish between narrowing due to vasculitis (inflammation of arteries) and narrowing due to atherosclerosis ("hardening" of the arteries). At times, this can be challenging. There are other causes of arterial narrowing as well, including fibromuscular dysplasia, another rare disease that mainly affects women.

Large arteries can also become inflamed in a few other diseases. Examples include other types of vasculitis: [giant cell arteritis](#) (a disease of older adults), relapsing polychondritis, Cogan's syndrome and Behçet's disease. Some infections can also cause inflammation in large arteries.

Blood tests for inflammation include measurements of the erythrocyte sedimentation rate (sometimes referred to as the "sed rate" or ESR) and C-reactive protein (often called CRP). Results of these tests are often, but not always, high in patients with TAK. However, these tests are also abnormal in a large number of other inflammatory diseases. Patients with TAK may also have anemia due to chronic (long-term) inflammation. Anemia is also tested for with a blood test. None of these blood tests can tell you for sure if you have TA, and these blood tests may be abnormal in many other diseases.

Patients with TAK may have no symptoms, and the disease is so rare that doctors may not easily recognize it. Thus, there is often a delay in detecting it, sometimes several years.

How is Takayasu's arteritis treated?

TAK most often needs treatment to prevent further narrowing of affected arteries. Yet, the narrowing that has already occurred often does not improve, even with drug treatment.

Glucocorticoids (prednisone, prednisolone or others), often referred to as "steroids," are an important part of treatment. The dose and length of treatment depend on how bad the disease is and how long the patient has had it. However, these drugs can have long-term side effects.

Doctors sometimes prescribe immune-suppressing drugs because their side effects may be less serious than those of glucocorticoids. This is called "steroid-sparing" treatment. These medicines include [methotrexate](#), [azathioprine](#), [mycophenolate mofetil](#), [cyclophosphamide](#) and [drugs that block tumor necrosis factor](#) (such as etanercept, adalimumab or infliximab). Doctors frequently prescribe these drugs to treat other rheumatic diseases, but they also use them to treat TAK. There is not enough proof that these drugs are definitely effective in treating TAK. Research studies are ongoing to find new drugs to treat TAK.

Some experts advise routine use of low-dose aspirin. The thought is that it will help prevent blood clots from forming in damaged arteries.

Therapy for TAK also includes screening for high blood pressure and high cholesterol, and treatment if these problems are present.

Lasting damage to arteries sometimes needs a vascular procedure or surgical treatment. This may involve angioplasty (widening a narrowed or blocked blood vessel), with or without placement of a stent, to prop open the vessel. Another treatment option is bypass grafting, a surgery to redirect blood flow around a blockage in a blood vessel.

What is the broader health impact of Takayasu's arteritis?

Because TAK can cause heart problems, high blood pressure and stroke, patients with TAK should talk to their doctor about ways to lower the risk of these serious problems.

Living with Takayasu's arteritis

TAK is a chronic disease and may need long-term treatment. Some patients have no symptoms or only mild symptoms, but others are disabled or need surgery more than once. Side effects from medicines, mainly glucocorticoids, can be troubling. Patients taking immunosuppressants are at risk of infections.

Blood pressure measurement is often not correct (falsely low due to blocked arteries) in the arm. So, your health care provider may need to measure your blood pressure in a leg.

The disease can recur after treatment or can silently get worse. It is often very hard to know whether TAK is active again. Thus, most patients need frequent doctor visits and angiograms.

Points to remember

- TAK is a rare inflammatory disease of large arteries.
- These patients often need treatment with glucocorticoids and immunosuppressive drugs.
- Symptoms of TAK reflect poor blood flow to tissues and organs.
- Detection of TAK most often requires testing by use of angiograms.

The rheumatologist's role in treating Takayasu's arteritis

Rheumatologists are usually the experts with the most overall knowledge about TAK. Thus, they direct the care of these patients, particularly those patients needing immunosuppressive drugs. Other doctors that patients may need to see include a cardiologist (heart doctor) and a vascular surgeon. A team approach can offer the best care to patients with this disease.

To find a rheumatologist

For a listing of rheumatologists in your area, [click here](#).

Learn more about [rheumatologists](#) and [rheumatology health professionals](#).

For additional 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.

National Institutes of Health

www.nlm.nih.gov/medlineplus/ency/article/001250.htm

Vasculitis Foundation

www.vasculitisfoundation.org

Updated February 2013

Written by Paul A. Monach, MD, PhD, and Peter A. Merkel, MD, MPH, and reviewed by the American College of Rheumatology Communications and Marketing Committee.

This patient fact sheet is provided for general education only. Individuals should consult a qualified health care provider for professional medical advice, diagnosis and treatment of a medical or health condition.

© 2013 American College of Rheumatology