**Takayasu’s Arteritis**

**Fast Facts**
- Takayasu’s arteritis is a rare disease involving inflammation in the walls of the largest arteries in the body: the aorta and its major branches.
- Takayasu’s arteritis can result in a loss of pulses in arms, legs, and other areas; for this reason, the disease was sometimes referred to as “pulseless disease.”
- Takayasu’s arteritis is much more common among women than men and usually starts when patients are young adults, although children may also get Takayasu's arteritis.
- Takayasu’s arteritis is diagnosed by angiography, which shows narrowing of large arteries.
- The problems caused by narrowed or blocked arteries range from mildly annoying to life-threatening.
- Takayasu’s arteritis is almost always treated with glucocorticoids (prednisone and others) but other medications that suppress the immune system are also sometimes prescribed.

**What is Takayasu's arteritis?**

Vasculitis refers to inflammation of blood vessels. In Takayasu’s arteritis, one of many types of vasculitis, this inflammation occurs in the walls of large arteries: the aorta and its major branches, which supply blood to the head, arms, legs, and internal organs. Inflammation may cause arterial walls to thicken resulting in narrowing of the inside diameter of the artery. If severe enough, such narrowings, also called “stenoses,” can result in reduced blood flow and decreased oxygen delivery to the body areas or organs supplied by the artery and cause symptoms and problems ranging from mildly annoying to life-threatening. For example, poor blood flow to an arm or leg might not cause problems at rest but would lead to pain with use of that limb—this symptom is called “claudication.” Poor blood flow to the brain can lead to dizziness, headaches, or even a stroke; similarly, blockages of the coronary arteries can lead to a heart attack. Takayasu’s arteritis that affects the renal arteries (which supply the kidneys) may produce no symptoms but may lead to severely high blood pressure.
Since stenosis occurs gradually over time, smaller vessels grow and expand to carry blood around the obstruction—these are called “collateral” vessels. Collateral vessels may help prevent major organ damage. Sometimes, the inflammation in the artery weakens the wall and the vessel expands, causing an aneurysm. The aorta as it emerges from the heart is one area of artery that typically expands to form an aneurysm rather than narrowing, potentially leading to heart valve dysfunction or rupture of the aorta.

Vasculitis of large arteries can also occur in a few other diseases, such as giant cell arteritis (a disease of older adults), relapsing polychondritis, Cogan’s syndrome, and Behçet’s disease.

What causes Takayasu’s arteritis?
As with most types of vasculitis, the cause of Takayasu’s arteritis is not known. It is quite rare to see multiple cases within a family. No association with an infectious agent has been convincingly identified. The observation that this is predominantly a disease of young women has not yet resulted into an insight into the cause of the vasculitis.

Who gets Takayasu’s arteritis?
Takayasu’s arteritis is rare, affecting perhaps one in 200,000 people. It is diagnosed in young adults (ages 15-40 years), occasionally in children or middle-aged adults, and 90% of patients are female. It appears to be more common in East Asia, India, and, perhaps, Latin America than in other regions, but it is rare even in these regions and is seen in a wide range of ethnic groups.

How is Takayasu’s arteritis diagnosed?
Takayasu’s arteritis is usually diagnosed by an angiogram, a test that demonstrates how well blood flows in arteries. Various types of angiogram are now available, including conventional angiograms involving injection of dye directly into an artery, as well as less invasive techniques such as computerized tomography angiography (CTA) or magnetic resonance angiography (MRA). An angiographic procedure is usually requested on the basis of symptoms in combination with physical exam findings, such as loss of pulse or blood pressure in an arm, or abnormal sounds (“bruits”) heard over large arteries with a stethoscope. Angiographic studies reveal narrowing of one or more large arteries. It is important to try and distinguish between narrowing due to vasculitis (inflammation of arteries) and narrowing due to atherosclerosis (“hardening” of the arteries). There are other causes of arterial narrowing, including another rare disease called fibromuscular dysplasia that leads to narrowing of major arteries, usually in women.

Blood tests for inflammation (including measurements of the erythrocyte sedimentation rate [ESR] or “sed rate,” and C-reactive protein [CRP]) are often, but not always, elevated in patients with Takayasu’s arteritis, and a large number of other inflammatory diseases produce similar abnormalities in these tests.

Because patients with Takayasu’s arteritis may have no symptoms and the disease is so rare that doctors may not easily recognize a case, there is often a delay in diagnosis, sometimes for several years.
How is Takayasu’s arteritis treated?
Takayasu’s arteritis is usually needed in order to prevent further narrowing of affected arteries, but the narrowing that has already occurred often does not improve, even with treatment. Glucocorticoids (including prednisone, prednisolone, or other similar drugs), often referred to as “steroids,” are an important part of therapy, but dose and duration vary with the severity and chronicity of the disease. Other immunosuppressive agents are used because the long-term side-effects of these other drugs are thought to be usually less severe than those of glucocorticoids (“steroid-sparing”). Methotrexate, azathioprine, and inhibitors of tumor necrosis factor (including etanercept, adalimumb, infliximab, and others), are used more widely in treating other rheumatic diseases, but they are also used to treat Takayasu’s arteritis; however, strong evidence is lacking for the effectiveness of these drugs in treating this disease. Research studies are ongoing to find new drugs to treat Takayasu’s arteritis.

Some authorities advocate routine use of low-dose aspirin with the thought that such treatment will help prevent formation of blood clots in damaged arteries. Screening for both high blood pressure and high cholesterol, and treatment for these disorders if they are found, are additional important parts of therapy for Takayasu’s arteritis.

Permanent damage sometimes requires vascular intervention or surgical treatment, such as angioplasty with or without stenting, or vascular bypass grafting.

Living with Takayasu’s arteritis
Takayasu’s arteritis may require long-term treatment. Some patients have no or only mild symptoms, whereas others have chronic disability or require repeated surgical interventions. Side effects from medications, especially glucocorticoids, can be particularly troubling. Blood pressure measurement in the arms is often inaccurate (artificially low due to blockage of arteries), so patients and their doctors need to know that blood pressure may need to be measured in a leg. It is often very difficult for patients and their physicians to determine whether Takayasu’s arteritis is currently active, and thus frequent doctor visits and repeated angiograms are needed. Patients are encouraged to try to live normal lives, but to remember that they may be at risk of recurrence of disease, or at risk of infection if they need to remain on immunosuppressive medications.

Points to remember
- Takayasu’s arteritis is a rare inflammatory disease of large arteries that often requires treatment with immunosuppressive drugs including glucocorticoids.
- Symptoms of Takayasu’s arteritis reflect poor blood flow to tissues.
- Diagnosis of Takayasu’s arteritis usually requires angiography.

The rheumatologist’s role in treating Takayasu’s arteritis
Rheumatologists are usually the specialists with the most overall knowledge about Takayasu’s arteritis and thus direct the care of patients, particularly those requiring immunosuppressive medications. A patient may also benefit from seeing a cardiologist and/or a vascular surgeon, depending on the individual’s clinical situation. A team approach can be critical for optimal management of patients with Takayasu’s arteritis.
For more information
If you want more information on this or any other form of arthritis, contact the Arthritis Foundation at (800)283-7800 or visit the Arthritis Foundation Web site at www.arthritis.org.

Links to ACR fact sheets on vasculitis, Wegener’s granulomatosis, and giant cell arteritis
2. Vasculitis Clinical Research Consortium (VCRC) www.RareDiseasesNetwork.org/vcrc

To find a rheumatologist
For a listing of rheumatologists in your area, click here. Learn more about rheumatologists and rheumatology health professionals.

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