What is Takayasu Arteritis?
Takayasu arteritis (TAK) is a form of vasculitis, which is an 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.

TAK affects the aorta, which carries blood from the heart to the rest of the body. It can also involve the aorta’s main branches.

Causes
The cause of TAK is not fully understood. It is classified as an autoimmune disorder—a disease that occurs when the body’s natural defense system mistakenly attacks healthy tissues. It is thought that an infection may set the autoimmune process in motion, but this is not yet proven. Environmental and genetic factors may also play a role.

Who Gets TAK?
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.

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
- Rapid weight loss

Some people may not experience symptoms for months or years—even as inflammation causes damage to blood vessels and organs.

Second-stage symptoms include:

- Pain, numbness, 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 arms and/or legs
• Chest pain
• Shortness of breath
• Fatigue

Seek medical attention right away if you experience signs of a:

• Heart attack (chest or arm pain, shortness of breath, nausea/vomiting)
• 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 your symptoms and medical history, as well as a:

- **Physical exam:** Abnormal exam findings may raise suspicion of TAK, including:
  - A weak or absent pulse in the limbs and organs (TAK is sometimes called the “pulseless disease”)
  - Low or high blood pressure
  - Different blood pressure between arms and/or between legs
  - Abnormal vascular sounds called “bruits” heard over the large arteries with a stethoscope

- **Blood tests:** Three common blood tests that can help detect inflammation include:
  - High erythrocyte sedimentation rate test, commonly called the “sed rate”
  - High C-reactive protein test
  - Complete blood cell count showing anemia and/or high platelets

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, which is life-threatening. Your doctor may order other imaging tests as indicated, including the Doppler ultrasound and positron emission tomography scan.

• **Biopsy:** This is the surgical removal and analysis of suspected tissue used to confirm 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.

**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, which are complex proteins derived from living organisms. They target certain parts of the immune system to control inflammation. Among these, tocilizumab and infliximab have 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. Recurrent inflammation 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, which is 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
- Potential bone loss (osteoarthritis), 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
- Other specialists
The best way to manage your disease is to actively partner with your health care providers and get to know the members of your health care team.

A health care journal is helpful to track medications, symptoms, test results, and notes from doctor appointments.

Make a list of questions before your doctor visit. Bring along a supportive friend or family member to provide a second set of ears and take notes.

Remember, it is 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 TAK
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. This can affect 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 TAK 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.

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