

“Takayasu Syndrome.”

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What is Takayasu Syndrome?

Takayasu Syndrome (also known as Takayasu Arteritis and Pulseless Disease) is a rare form of vasculitis. It causes the inflammation of the aorta and its major branches (1). It can also affect pulmonary and coronary arteries. The inflammation narrows the lumen of the arteries and can eventually cause thrombosis. In some cases, the weakness of the walls results in aneurysm leading to death (2). The disease was discovered by Dr. Mikito Takayasu, an ophthalmologist, in 1908 in Japan. He noticed a “wreathlike” formation of blood vessels behind the retina. This new blood vessel growth is a response to the narrowing of the arteries in the neck. Some of Takayasu’s colleagues also found the pulse in the wrist to be absent due to the narrowing of the arteries in the arm (3).

Who does it affect?

Takayasu Syndrome is mostly prevalent in Asians although cases have been reported all over the world. Females are eight times for likely to be affected and the typical age of onset is between 15 and 30 years old (2).

What causes it?

The cause of Takayasu’s is unknown. Some researches believe it is an autoimmune disease in which the body attacks its own cells (4). Some other studies suggest it may be

caused by an infection (viral or bacteria) occurring in a person with predisposing genetic factors (3). Research, however, is still being conducted to find a definite cause.

What are some symptoms?

Symptoms at first include myalgia, arthralgia, malaise, fever and arthritis. However since blood flow is being obstructed weakness in the legs and arms eventually result (5). Other symptoms include dizziness, weight loss, chest pain, circulatory deficit, rash, high blood pressure, night sweats, decreased or absent radial pulses and stroke (6).

How can it be diagnosed?

Although a diagnosis of Takayasu Syndrome is hard to determine some tests can be used to reach a diagnosis. Blood tests can be used to check for a high white blood cell count or high levels of C-reactive protein, which are signs of inflammation. An increased erythrocyte sedimentary rate is another sign of inflammation. An angiography can be done to see if blood flow is normal. A sign of Takayasu's will be stenosis of several arteries. A CT angiography can be used to look at the structure of the aorta and monitor its blood flow. Doppler ultrasound can produce high-resolution images and detect subtle changes of artery walls. It can be used to distinguish between Takayasu Syndrome and atherosclerosis (3).

How is it treated?

Early detection and proper treatment can make the chronic Takayasu Syndrome very manageable. Treatment can include medications and/or surgery. The most common form

of treatment is administering the corticosteroids, such as prednisone. After about a month, the dose is gradually lowered to the lowest level possible to control the inflammation. If corticosteroids are not effective, the patient may be given cytotoxic drugs to reduce blood vessel inflammation. Examples of cytotoxic drugs include Rheumatrex, Imuran and Cytoxan. A more recent treatment used is the use of TNF-alpha inhibitors. These medications reduce inflammation by inhibiting the TNF-alpha protein. This protein plays a role in initiating an immune response. Since many of the medications mentioned above have side effects if taken long-term the dosage must be monitored by the physician. Sometimes surgery is necessary to fix narrowed or blocked arteries. These procedures include bypass surgery, percutaneous angioplasty and stenting (3).

What's the prognosis?

When Takayasu Syndrome is treated properly, the prognosis is usually good. The long term survival rate is greater than 90% (1). However Takayasu Syndrome is still dangerous. About 50% of those affected suffer major complications such as stroke, heart attack, heart failure or aneurysm (2).

Can Takayasu Syndrome be prevented?

Since the cause is still unknown, nobody is completely sure if it can be prevented. What researchers do know is complications can be prevented with early diagnosis and proper treatment (3).

What now?

Those with Takayasu Syndrome should be fully aware of their risks. Since the aorta and its branches are inflamed, they are at greater risk of atherosclerosis which leads to cardiovascular problems such as heart attack and/or stroke. Patients should visit their physician regularly for check-ups. Another thing the patient must keep in mind is managing the side effects of the medications. The best thing the patient can do to curb the side effects is to eat a healthy diet and exercise regularly (3).

Where can I find more information?

Medline Plus Encyclopedia:

<http://www.nlm.nih.gov/medlineplus/encyclopedia.html>

Takayasu's Arteritis Research Association:

<http://www.takayasus.org/>

MayoClinic.com

<http://www.mayoclinic.com/health/DiseasesIndex/DiseasesIndex>

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