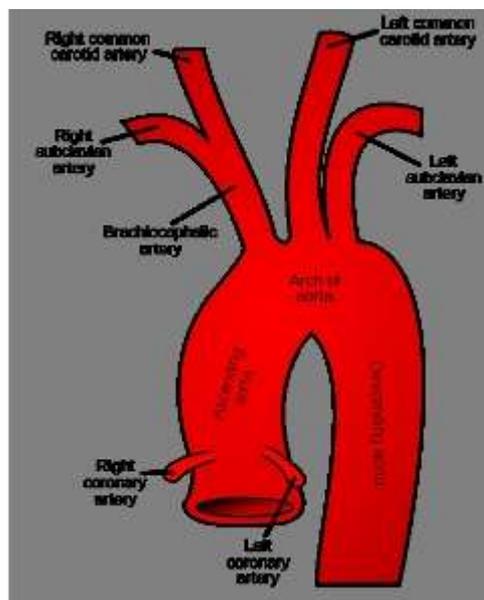
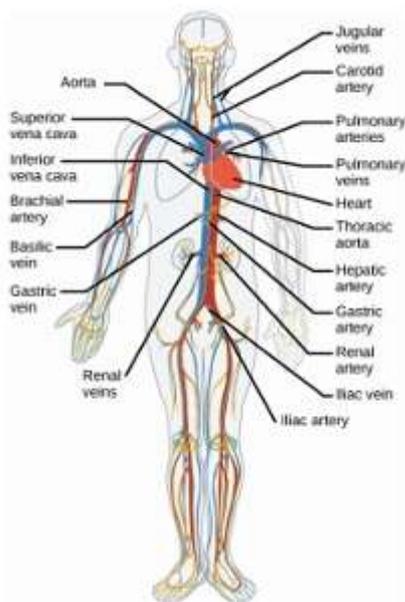


Takayasu Arteritis

What is Takayasu Arteritis?

Takayasu Arteritis (TA) is an inflammatory disease of the large arteries. TA particularly affects the aorta (the main artery taking blood from the heart to the rest of the body), and the pulmonary artery (which sends blood to the lungs). The major arteries that arise from the aorta may also be affected. These include the subclavian arteries that supply the arms, renal arteries to the kidneys, coronary arteries in the heart and carotid arteries in the head and brain. (see illustrations below)

In some patients, widening of the aorta results in failure of the aortic valve in the heart, necessitating replacement. In 90% of patients one or more of these arteries become narrowed or blocked, hence TA is also known as "Pulseless disease". In 25 per cent of patients part of an artery may swell, forming an aneurysm.



Click on the diagrams above to enlarge

Illustrations above: Circulation diagram sourced from the "Arteries, Veins, and Capillaries" section of Robert Bear and David Rintoul's article, "The Circulatory System," *Connexions*, August 19, 2013, [The Circulatory System](#). Aorta - Ascending, Arch & Descending diagram sourced from: "Gray's Anatomy" Wikipedia Commons, [Aorta diagram](#).

Who are affected?

TA is a very rare disease of young people with the initial symptoms arising in those between 5 and 40 years of age. Although both sexes may be affected, 80-90 per cent of patients are female. TA has worldwide prevalence and may affect all races. However, the disease is thought to be more common in patients originating from the Far East, Japan and the Asian sub-continent. As far as we know, TA is not a genetically inherited disease. It is unusual for more than one family member to suffer from the disease. However, the increased prevalence in certain parts of the world may indicate a genetic component to disease susceptibility and current studies are investigating this.

What is the aetiology (cause)?

The cause of Takayasu's arteritis, and why an individual develops the disease at any one time remains unknown. The disease is most likely to be the consequence of environmental factors and a susceptible genetic background. One common but unproven hypothesis is that it is precipitated by

an infection. The rarity of the disease worldwide means that it is very difficult to identify an underlying cause. The inflammation involves white blood cells invading the wall of the artery predisposing to damage and scarring.

What are the symptoms?

The initial symptoms of TA are typically non-specific and may include one or more of:- malaise (a general feeling of being unwell), profound fatigue, fever, night sweats, weight loss, myalgia (muscle pain), arthralgia (painful joints), rash. Additional symptoms can include dizziness/light headedness, shortness of breath, cramping pain in the arms, legs or chest on exertion. Carotidynia, (pain and tenderness over the carotid arteries in the front of the neck) is also found in approximately 25 per cent of patients.

Making a diagnosis

This is made on the basis of formal criteria such as those published by the American College of Rheumatology. For diagnosis the patient should fulfil three or more of the criteria listed below:

Diagnostic Criteria for Takayasu's Arteritis

1. Onset of disease \leq 40 years
2. Claudication** of an extremity
3. Reduced brachial artery pulsation
4. Difference in systolic blood pressure >10 mmHg between the arms
5. Aortic or subclavian artery bruit
6. Angiographic abnormality

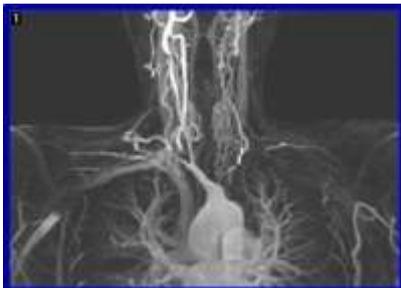
** Claudication - severe muscle pain due to cramp, caused by narrowed blood vessels

In practice clinical examination most commonly detects decreased or absent pulses in the arms and less frequently in the legs. Using a stethoscope, bruits*, (a loud 'whooshing noise') may be detected, over the neck, chest or kidneys indicating narrowed arteries. High blood pressure is commonly found. In recent years specialist scans have been introduced to aid diagnosis and identify inflammation, narrowing and dilatation of arteries and the extent of the disease. These include positron emission tomography (PET scanning), magnetic resonance angiography (MRA), high-resolution ultrasound and CT angiography (see below).

Diagnostic tests

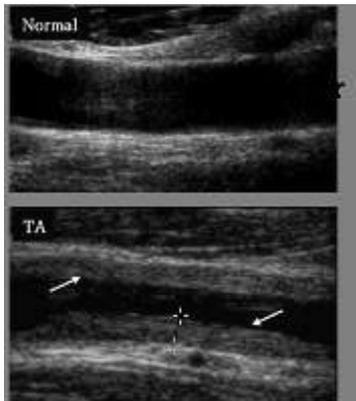
Blood Tests: There is no blood test that can be used to definitively confirm the diagnosis. The erythrocyte sedimentation rate (ESR) and C-reactive protein (CRP) can help measure activity of the illness and your response to treatment.

Imaging: Imaging studies are useful in the diagnosis and monitoring of TA. Their use may allow diagnosis before arterial damage is severe. However, none of them are 100 per cent accurate.



Magnetic resonance imaging (MRI) is commonly used to aid diagnosis and assess the extent of TA. The scan does not involve radiation and the picture of the arteries can be enhanced by using a contrast medium (that only rarely causes an allergic reaction).

MRI is a safe means by which to monitor the progress of disease and response to treatment. The main drawbacks are that the scanner is very noisy and rather claustrophobic. The illustration shows an MRI scan of a TA patient.



Ultrasound - Doppler ultrasound (ultrasound scan): uses a probe which is passed over the carotid arteries in the neck. This allows assessment of blood flow and can detect and monitor the arterial wall and the degree of narrowing.

This illustration shows a normal carotid artery and one from a TA patient. The arrows show the thickening of the artery wall causing the narrowing of the artery.

Positron emission tomography (PET scanning): requires the patient to be injected with radioactive fluorodeoxyglucose. PET scans are particularly good at detecting inflammation in the arterial wall and are often used in combination with CT scanning (CT-PET). PET may identify early active disease and may on occasion help to distinguish active disease (which needs further treatment) from scarring resulting from previous inflammation. The illustration shows two PET images from the same patient; the one on the left is before treatment, with the arrows pointing to areas of inflammation. These areas have disappeared in the image on the right which was taken after six months treatment.



Computed tomography (CT) angiography: Contrast-enhanced CT angiography is useful for the diagnosis of TA and can help identify the extent of damage to the arteries. Unlike MRI/MRA and Doppler US, CT angiography does involve radiation.

Treatment

Treatment is aimed at improving symptoms and preventing further damage and/or scarring to the blood vessels. When there is active inflammation in the arteries, treatment is started with steroids, usually prednisolone, and often in combination with an immunosuppressant drugs. Those commonly used include methotrexate, azathioprine or mycophenolate. These drugs increase the effectiveness of steroid therapy and help to reduce the dose of steroids required. In patients with persistent or very severe disease, cyclophosphamide may be recommended. In recent years, for refractory disease, biologic therapies, including tumour necrosis factor inhibitors such as etanercept and infliximab have been used successfully, as has tocilizumab which targets interleukin-6.

Additional treatment: Drugs to lower blood pressure are often recommended. Statins, the most common of which is simvastatin, are commonly prescribed to lower cholesterol and reduce the risk of heart disease. In TA they may also have a beneficial effect on arterial function, even when cholesterol levels are normal. Low-dose aspirin may be prescribed to reduce the risk of a blood clot developing.

Surgery: Surgical intervention should not be undertaken lightly and is only indicated for the most severe cases of TA. For optimal results surgery should be carried out when the disease is in remission and decisions will involve a team of specialist doctors including the rheumatologist, vascular surgeons and radiologists. Procedures that may be carried out include:

Revascularisation - where a blocked artery is bypassed, typically with a vein graft from somewhere else in your body. Good long-term outcomes have been seen, with 20 year graft survival rates >70 per cent in one study. However, results are variable and further studies are needed for more conclusive evidence.

Percutaneous transluminal angioplasty (PTA) - aims to open narrowed arteries by inserting and inflating a balloon to stretch the narrowed section. In some cases a wire mesh (stent) may be required to keep the artery open.

Drugs and Side effects

For information on the main drugs prescribed for Takayasu Arteritis see:

- [Azathioprine](#)
- [Cyclophosphamide](#)
- [Methotrexate](#)
- [Mycophenolate Mofetil \(Cellcept\)](#)
- [Steroids](#)

Pregnancy and Takayasu arteritis

Many women with Takayasu's arteritis achieve one or more successful pregnancies.

It is very important for a woman planning to have a baby to talk to her specialist before trying to conceive. Depending on how advanced your disease is and how much damage has been done to your arteries, it may on occasion be dangerous for a woman to carry a baby. For example control of blood pressure can be difficult in patients with TA and blood pressure can be increased by pregnancy to a dangerous level. Likewise, high pressure in the pulmonary (lung) circulation can place a woman at significant risk during pregnancy.

Unless the disease is very active, TA itself will not affect your fertility, whether you are a man or a woman. However, some of the drugs used to treat the condition may reduce fertility, Likewise, other medications may increase the risk of defects in the foetus. It is therefore essential for both women and men with TA to consult their specialist to discuss all aspects of starting a family.

Prognosis

The long term prognosis of TA is good. Approximately 20 per cent of patients will have a monophasic self-limiting disease (just one inflammatory episode). More typically, the disease follows a relapsing and remitting course. Limited data is available on long-term outcome and varies significantly between patients, dependent upon the extent of their disease. In the majority of patients the TA appears to 'burn out' after a period of 2-5 years, and treatment can be gradually withdrawn. The figures below summarise data from surveys performed around the world:

- 25 per cent of patients largely unaffected by their disease
- 25 per cent of patients able to perform normal daily activities when disease in remission
- 50 per cent were unable to consistently do or maintain full their full range of daily activities
- Up to 25 per cent of patients are unable to work due to their Takayasu's arteritis

With early intervention and careful monitoring in a specialist centre TA can be effectively treated. Mortality rates are low with survival in the USA and UK up to 98 per cent at 10 years. Mortality directly related to TA is usually the consequence of heart attack, heart failure, aneurysm rupture, stroke or kidney failure.

Key Points

- TA is very rare in the UK and mainly affects young women
- Presenting symptoms are often non-specific and missed
- Treatment with steroids and immunosuppressants is effective in many cases
- Surgical repair or angioplasty to narrowed blood vessels should only be performed in severe cases and after the inflammation is controlled

Related Vasculitis Articles

- [Fertility and Vasculitis](#) - Dr David Jayne

Further reading

- [Large Vessel Diseases](#) - Ashima Gulati, Arvind Bagga