TAKAYASU ARTERITIS (TAK)

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What is Takayasu Arteritis (TAK)?

TAK, also referred to as "**the pulseless disease**", is a type of vasculitis. "Vasculitis" is a rare condition where the body attacks its own blood vessels. This leads to vessel wall inflammation and disruption of blood flow to the affected areas, which subsequently causes pain and local tissue injury.

Which blood vessels are affected?

TAK primarily affects the aorta and the large arteries arising from it, including the:

- Subclavian arteries – arms
- Carotid arteries – head & neck
- Renal arteries – kidneys
- Hepatic arteries – liver
- Mesenteric arteries – intestines
- Pulmonary arteries – lungs
- Coronary arteries – heart
- Iliac & femoral arteries - legs

Who is affected?

TAK is a rare condition that affects 4-8 per million people in Western countries, with much higher numbers in Asia (especially Japan and India). It is most common in young women, with females being affected 8X more often than males. The age of onset is typically between 15-40 years.

What symptoms are seen in TAK?

Depending on the arteries affected, TAK can cause:

- Crampy pain in the arm and leg muscles that worsens with activity and improves with rest. This is known as **claudication.**
- Chest pain and/or palpitations
- Shortness of breath with activity
- Abdominal discomfort, nausea or bloating, especially after eating
- Cold and numb extremities
- Pain along the carotid arteries on either side of the neck, often radiating up to the jaw or ear
- Headaches
- Pulsatile whooshing sound
- Dizziness or fainting with activity
- Vision loss or double vision
- Stroke-like symptoms
- High blood pressure, or a major difference in blood pressure between the arms

Other clinical features NOT related to the blood vessels include:

- Erythema nodosum, which is a skin rash appearing as painful raised red nodules usually over the shins
- Uveitis and other forms of eye inflammation

How is TAK diagnosed?

Diagnosis of TAK is made based on a combination of:

- Compatible symptoms & physical exam findings
- Blood tests showing elevated inflammatory markers (ESR and/or CRP)
- Imaging of the arteries from the neck down to the upper legs by CT scan or MRI with vessel imaging (angiography)
- Conventional angiography to visualize the arteries better. This is performed by puncture of an artery in the arm or thigh with injection of a contrast dye medium, under a radioscopic imaging device. Note that conventional angiography is not commonly done as it is more invasive than regular imaging with CT or MRI.

Diagnosing TAK may be challenging when very few arteries are affected, or if the pattern of artery involvement is unusual. Part of the diagnostic process is to rule out other diseases that disrupt the arteries. These may include other types of vasculitis, infections and rare genetic conditions.

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How is TAK treated?

- **Prednisone** is a steroid medication that is highly effective at reducing inflammation. You will start on a high dose and slowly reduce the dose over time.
- **Other immunosuppressive drugs** are often needed, and/or used to replace prednisone in the long-term. These include methotrexate, leflunomide, azathioprine, anti-TNF agents (such as infliximab, etanercept or adalimumab) and tocilizumab. Some of these are taken as pills while others are injections or infusions.
- **"Baby Aspirin" (81 mg per day)** – This may be added if there is vision loss, stroke or other significant cardiovascular risk factors.
- **Bone and stomach protectant** medications to prevent certain complications of high-dose prednisone.
- **Surgical procedures** such as bypass and stenting may be needed in specific emergency situations to "reopen" the blood vessels when there is dangerously low blood flow to an organ. They are normally avoided since the arteries often "close up" again or **restenose** after the procedure. Therefore, the mainstay of therapy in TAK is still with medications alone.

How is TAK monitored?

Common tests used to monitor the disease include:

- Lab tests for blood counts and inflammatory markers (CRP and/or ESR)
- Repeat CT scans and/or MRI with angiography every 6-12 months
- Echocardiogram to assess heart function
- PET scans are available in certain centres however results can be difficult to interpret. There is ongoing debate on how to use the results effectively.

Your rheumatologist may work with a **cardiologist** and/or **vascular surgeon** to treat and monitor your TAK. Your **family physician** also plays a crucial role in monitoring you for hypertension, high cholesterol, diabetes and osteoporosis.

What will happen to me?

TAK is treatable but not yet curable. Some people have a single severe episode of inflammation at the beginning of the disease and can eventually come off treatment after a few years, while others need lifelong treatment due to recurrent flares.

I was diagnosed with TAK after the age of 40. Is this possible?

Some people with TAK have very few symptoms, which causes the disease to stay undetected for years. In these cases, it is not uncommon to have a CT scan or MRI done for a different reason down the road, and it just happens to show the characteristic artery narrowing seen in TAK. By then, the inflammation in the arteries has already entered the irreversible occlusive phase.

Can I get pregnant if I have TAK?

It is important to discuss pregnancy plans with your rheumatologist early. It is recommended to be in **remission** (no active disease) for at least 6 months before trying to get pregnant. Active disease during pregnancy can increase the risk of complications for both mom and baby. If your disease is very severe and has already led to damage in your organs (eg. the heart, lungs, or kidneys), then pregnancy may be very risky.

Some medications used to treat TAK are also unsafe to take before and/or during pregnancy. Once pregnant, you will need to be followed by a medical team that specializes in high-risk pregnancies.

TAK DISEASE SUMMARY TOOL

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My symptoms included:

- ☐ Arm claudication (left / right)
- ☐ Leg claudication (left / right)
- ☐ Cold and numb extremities
- ☐ Chest pain or palpitations with exertion
- ☐ Shortness of breath with exertion
- ☐ Dizziness or fainting
- ☐ Headaches
- ☐ Abdominal discomfort after eating
- ☐ Pain along the neck arteries (carotids)
- ☐ Pulsatile whooshing noise in the ears
- ☐ Vision loss or double vision
- ☐ Erythema nodosum skin rash
- ☐ Weakness of a part of the body
- ☐ Major difference in blood pressure between the arms
- ☐ Joint and muscle aches
- ☐ Fevers and/or night sweats
- ☐ Weight loss
- ☐ Severe fatigue

Medications that I have been prescribed:

	Date started	Date stopped
Prednisone: (Starting dose _____ mg)		
Aspirin 81mg		
Methotrexate ____ mg		
Folic acid (only if on methotrexate)		
Leflunomide ____ mg		
Azathioprine ____ mg		
Mycophenolate mofetil _____ mg		
Tocilizumab 162 mg		
Etanercept 50 mg		
Infliximab _____ mg		
Adalimumab 40 mg		
Others:		

I had the following tests done:

- ☐ Blood tests showing elevated ESR / CRP
- ☐ Conventional angiography
- ☐ CT scan (circle all that apply):
  Brain | Abdomen + pelvis | Legs
- ☐ MRI scan (circle all the apply:
  Brain | Abdomen + pelvis | Legs
- ☐ Echocardiogram
- ☐ PET scan

What do I need to do?

- ✓ Attend follow-up visits with my rheumatologist and other specialists
- ✓ Do regular blood tests to monitor my disease and medication effects.
- ✓ Ensure my medical team is monitoring me for diabetes, high blood pressure and high cholesterol levels.
- ✓ **Seek urgent medical attention (call 911) if I have new stroke-like symptoms, fainting spells or recurrent chest pain with exertion.**