

Recommendations for managing Takayasu arteritis (TAK)



These are recommendations written by a group of doctors and patients based on research studies for people with Takayasu arteritis (TAK).

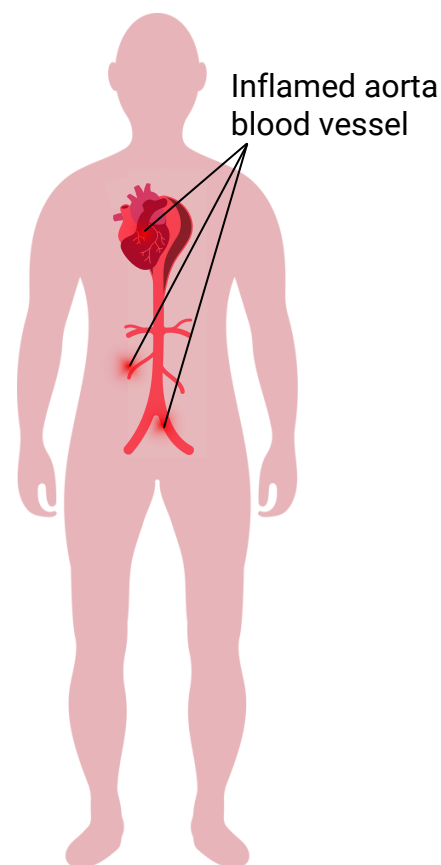
These recommendations suggest the best treatment for most people, but your individual situation and treatment may be different. Talk to your doctor about what treatment is best for you.

What is Takayasu arteritis (TAK)?

Takayasu arteritis (TAK): A type of vasculitis that affects the aorta and its main branches. The aorta is the largest blood vessel in your body. It carries blood from the heart to the rest of the body.

Words to know about vasculitis

- **Vasculitis:** A group of conditions that involves inflammation (swelling) of your blood vessels.
- **Suspected disease:** Symptoms that suggest TAK and are not explained by other conditions.
- **Active disease:** New, ongoing, or worsening signs or symptoms.
- **Severe disease:** Symptoms that may cause death or organ failure (when a major organ stops working, such as the heart).
- **Nonsevere disease:** Symptoms not likely to cause death or organ failure.
- **Remission:** When symptoms go away.
- **Refractory disease:** When symptoms do not get better with treatment.
- **Relapse:** When symptoms return after a period of remission.



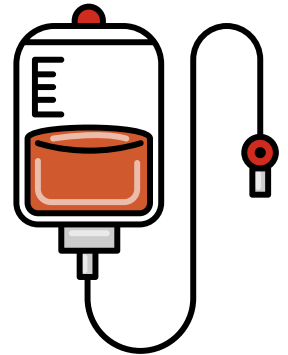
See a glossary of health terms at the end of this document.



Medical treatment recommendations

If you have **active TAK**:

- ➔ For initial treatment, use another immunosuppressant (such as methotrexate, tumor necrosis factor inhibitor (TNFi), or azathioprine) over tocilizumab, because we do not know how well tocilizumab works for TAK.
- ➔ Use an additional immunosuppressant (such as methotrexate or azathioprine) and prednisone, over prednisone alone. You can take a lower dose of prednisone, which will help lower the chance of serious side effects from prednisone.



If you have **newly diagnosed or relapsed, severe TAK**:

- ➔ Start treatment with high-dose prednisone, over low-dose prednisone, because a higher dose can help control the disease quicker, lowering the chance of organ damage or life-threatening events.

If you have **active, severe TAK and are not on immunosuppressants**:

- ➔ Start treatment with high-dose oral prednisone, over IV pulse prednisone followed by high-dose oral prednisone. IV pulse prednisone does not work better than high-dose oral prednisone.
 - When **might** I use IV pulse prednisone?
 - If you have life- or organ-threatening disease.

If you have **TAK that is in remission while on prednisone for 6–12 months or longer**:

- ➔ Lower the dose of prednisone until you are no longer on it, instead of long-term treatment with low-dose prednisone. Use of prednisone should be limited because of the potential serious side effects.



If you have **TAK** that is refractory to treatment with only prednisone:

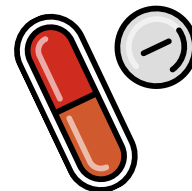
- ➔ Add a tumor necrosis factor inhibitor (TNFi) over tocilizumab, because we have more experience with TNFi than tocilizumab for TAK.

If you have **TAK** and a vascular lesion that is getting worse, but no signs of ongoing active disease:

- ➔ Continue your current treatments, over increasing or changing immunosuppressants, because the progression could be due to things other than TAK.
 - When **might** I increase or change immunosuppressants?
 - If the lesion progression happens quickly after a period where your disease had been stable (not getting worse).

If you have **active TAK** that affects blood vessels in your brain:

- ➔ Add aspirin or another antiplatelet drug, because this helps lower the chance of stroke caused by lowered blood flow to your brain.



Surgical treatment recommendations

If you need **surgery for TAK**:

- ➔ You, your surgeon, and your rheumatologist should decide together which surgery is best for you and when to get it.

If you have **TAK** and muscle pain when using your limbs (arms, hands, legs, or feet), but no signs of ongoing active disease:

- ➔ Do not get surgery.
 - Your body may start using other blood vessels to go around the blocked vessel that is causing pain, so you may not need surgery.
 - When **might** I get surgery?
 - If the pain greatly effects your activities.



If you have **TAK** with worsening signs of limb or organ ischemia:

- ➔ Wait to get surgery until your disease is in remission, over getting surgery while you have active TAK.
 - When **might** I get surgery when I have active TAK?
 - If you have life- or organ-threatening symptoms such as stroke, lack of blood flow to your heart, or not being able to use a limb.

If you have **TAK** with worsening signs of limb or organ ischemia while on immunosuppressants:

- ➔ Consider increasing immunosuppressants, over getting surgery and increasing immunosuppressants.
 - When **might** I get surgery?
 - If you have life- or organ-threatening symptoms such as stroke, lack of blood flow to your heart, or not being able to use a limb.

If you have **TAK**, high blood pressure, and narrowed blood vessels in your kidneys:

- ➔ Manage with medicines instead of surgery, because this may be treated effectively with blood pressure medicines and immunosuppressants.
 - When **might** I get surgery?
 - If your high blood pressure does not get better with medications.
 - If your kidney function gets worse.

If you have **TAK** and narrowed blood vessels in your head or neck, but no symptoms:

- ➔ Do not get surgery.
 - Manage with medicines instead of surgery, because there is a high chance of health problems from this surgery.
 - When **might** I get surgery?
 - If multiple blood vessels in your head or neck are affected.



Monitoring recommendations

In general, if you have TAK:

- Use inflammation markers to help monitor your disease.

If you have TAK that is in remission:

- Have long-term clinical monitoring, over no clinical monitoring.
 - Clinical monitoring may include exams, lab tests, and imaging. Monitoring can help find new symptoms early, before they become severe.
 - How long and often you need monitoring will depend on your specific situation.

If you have TAK with no symptoms, but have higher levels of inflammation markers:

- Have clinical monitoring, over increasing immunosuppressants.
 - Higher levels of these markers may not be related to TAK. But you may need to have more frequent tests that look for signs of active disease.

Imaging recommendations

In general, if you have TAK:

- Use noninvasive imaging instead of catheter angiography, because noninvasive imaging gives better information about the walls of blood vessels.
- Have routine noninvasive imaging in addition to routine clinical monitoring, because changes in vessels can happen even if you are not having symptoms.

If you have TAK and it is in remission, but imaging shows that you have inflammation in a new area:

- Use immunosuppressants, because this can be a sign of active disease.



Health terms

- A**
 - **Antiplatelet drug:** Medicines that stop platelets (cells that help you stop bleeding) from sticking together and forming blood clots.
 - **Azathioprine:** An immunosuppressant. Lowers inflammation (swelling) in your body.
- C**
 - **Catheter angiography:** A procedure used to see the blood vessels in your body. A catheter (a thin, flexible tube) is inserted into a blood vessel, and contrast material is injected through the catheter. Contrast material is a substance that makes your blood vessels more visible on an x-ray. An x-ray machine then takes a movie of how the contrast material moves through your vessels.
- I**
 - **Immunosuppressant:** A medicine that lowers your body's immune response to stop your immune system from causing inflammation (swelling) and damaging your body.
 - **Inflammation markers:** Blood tests doctors use to look for signs of inflammation.
 - **Ischemia:** Lowered blood flow to a part of your body.
- M**
 - **Methotrexate:** An immunosuppressant. Lowers inflammation (swelling) in your body.
- N**
 - **Noninvasive imaging:** Scans or tests that do not involve putting an instrument through your skin or into your body.
- P**
 - **Prednisone:** Lowers inflammation (swelling) in your body, and can be given as:
 - **IV pulse:** A tube into a vein as an IV.
 - **Oral:** A pill by mouth.
- T**
 - **Tocilizumab:** An immunosuppressant that lowers inflammation (swelling) in your body.
 - **Tumor necrosis factor (TNF) inhibitors (TNFi):** A medicine that lowers inflammation (swelling) in your body.
- V**
 - **Vascular lesions:** Abnormal growths in your blood vessels.



*Maz M, Chung SA, Abril A, Langford CA, Gorelik M, Guyatt G, Archer AM, Conn DL, Full KA, Grayson PC, Ibarra MF, Imundo LF, Kim S, Merkel PA, Rhee RL, Seo P, Stone JH, Sule S, Sundel RP, Vitobaldi OI, Warner A, Byram K, Dua AB, Husainat N, James KE, Kalot MA, Lin YC, Springer JM, Turgunbaev M, Villa-Forte A, Turner AS, Mustafa RA. 2021 American College of Rheumatology/Vasculitis Foundation Guideline for the Management of Giant Cell Arteritis and Takayasu Arteritis. *Arthritis Rheumatol*. 2021 Aug;73(8):1349-1365. doi: [10.1002/art.41774](https://doi.org/10.1002/art.41774). Epub 2021 Jul 8. PMID: 34235884.