

Recommendations for managing giant cell arteritis (GCA)



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

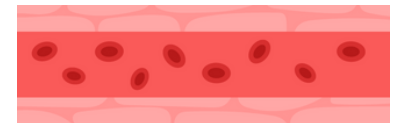
These recommendations suggest the best treatment for most people, but your individual situation and treatment may be different. Talk to your doctor about what treatment is best for you.

What is giant cell arteritis (GCA)?

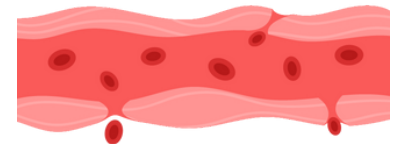
Giant cell arteritis (GCA): A type of vasculitis that mostly affects the arteries in your scalp and head, and large vessels in your body. It often affects the arteries over the temples (temporal arteries).

Words to know about vasculitis

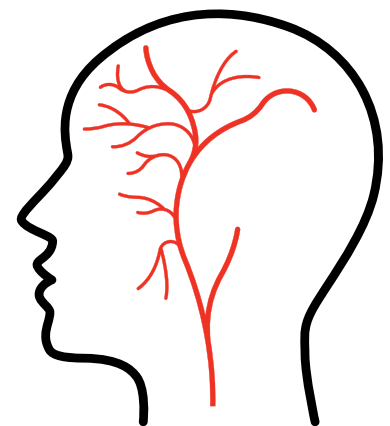
- **Vasculitis:** A group of conditions that involves inflammation (swelling) of your blood vessels.
- **Suspected disease:** Symptoms that suggest GCA and are not explained by other conditions.
- **Active disease:** New, ongoing, or worsening signs or symptoms.
- **Severe disease:** Symptoms that may 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.



Healthy blood vessel



Inflamed blood vessel



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



Recommendations for diagnosing GCA

If you have **suspected GCA**:

➔ **First get a temporal artery biopsy on one side of your head, over both sides of your head.**

- When **might** I get a biopsy on both sides of my head?
 - If your symptoms happen on both sides.
 - If you had a temporal artery biopsy on one side and it was negative, but you have other symptoms that suggest GCA.

➔ **During a temporal artery biopsy, the surgeon should take a section that is longer than 1 cm (about the size of a pea).**

- Since GCA affects only parts of the artery, not the whole artery, a longer section is more likely to have a part affected by GCA.

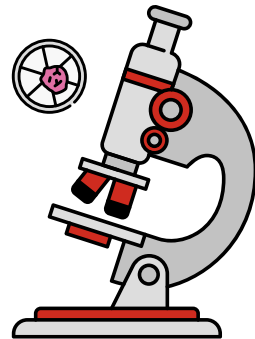
➔ **Get a temporal artery biopsy within 2 weeks of starting oral prednisone, when changes to your artery caused by GCA are more likely to be found.**

➔ **Get a temporal artery biopsy over:**

- A temporal artery ultrasound.
- An MRI of the arteries in your brain.

Why?

- Doctors in the US have less experience diagnosing GCA with temporal artery ultrasound or an MRI.
- We also do not know how well an MRI works for this.



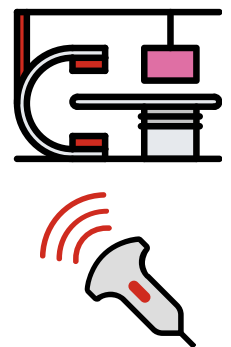
If you have **suspected GCA and a negative temporal artery biopsy result**:

➔ **Get noninvasive vascular imaging of the large vessels and a clinical assessment. Possible noninvasive vascular imaging procedures:**

- MR angiography
- CT angiography
- Vascular ultrasound
- PET scan

Why?

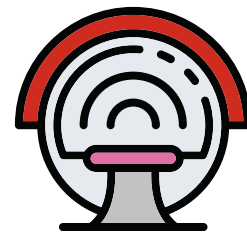
- Imaging of large vessels in your neck, chest, belly area, or pelvis may give more evidence of GCA (especially of GCA outside of the brain).



If you have newly diagnosed GCA:

→ Get noninvasive vascular imaging to see if large vessels are also affected.

- Imaging with an MRI or CT angiography of your neck, chest, belly area, or pelvis can see if large vessels in your body are affected.
- If you do have affected large vessels, you may get stronger treatment and repeat imaging studies to help monitor your disease.



Medical treatment recommendations

If you have newly diagnosed GCA:

→ Use oral prednisone daily dose, over an alternate-day (every other day) dose.

→ Start treatment with high-dose oral prednisone, over moderate-dose oral prednisone because a high-dose can control the disease more quickly.

- When **might** I use moderate-dose oral prednisone?
 - If you are highly likely to have serious side effects from prednisone.
 - If you have a low chance of vision loss or other life-or organ-threatening problems from GCA.

→ Use oral prednisone and tocilizumab, over oral prednisone alone.

→ Do not use a statin specifically to treat GCA, because statins do not work well to treat GCA. However, you may still use statins if you are at risk for heart disease.

→ If you do not have symptoms of cranial ischemia such as vision loss or strokes:

- Start treatment with high-dose oral prednisone, over IV pulse prednisone, because IV pulse prednisone can raise the chance of infections and serious side effects from prednisone.

→ If you have symptoms of cranial ischemia and are therefore at risk for vision loss:

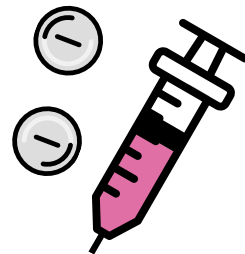
- Start treatment with IV pulse prednisone, over high-dose oral prednisone.
 - IV pulse prednisone may help control the disease more quickly to prevent further damage.
 - IV pulse prednisone can have risks (as mentioned above), so the decision to use it should be based on your condition and personal preferences.



If you **have GCA and affected large vessels** in other parts of your body:

→ Use oral prednisone and immunosuppressants, over oral prednisone alone.

- We recommend tocilizumab, but you could use methotrexate instead.
- When **might** I use methotrexate?
 - If you can't tolerate tocilizumab, have limited access to it, or if it is too expensive.



If you **have GCA and narrowed arteries** in your neck that cause lack of blood flow:

→ Use aspirin, because aspirin may help prevent strokes due to restricted blood flow to the brain.

If you **have GCA disease relapse**:

→ While on moderate-to high-dose prednisone add another immunosuppressant such as tocilizumab or methotrexate.

- Relapse while on moderate-to high-dose prednisone makes it less likely that the GCA will remain under control as you taper to a lower dose of prednisone.
- You can take a lower dose of prednisone, which will help lower the chance of serious side effects from prednisone.

→ With symptoms of cranial ischemia add another immunosuppressant such as tocilizumab or methotrexate and a higher dose prednisone, over a higher dose prednisone alone.

→ With symptoms of cranial ischemia while on prednisone add tocilizumab and use a higher dose prednisone, over methotrexate and a higher dose prednisone.

- With tocilizumab, you may be able to use a lower prednisone dose than you would with methotrexate.
- When **might** I use methotrexate?
 - If you can't tolerate tocilizumab, have limited access to it, or if it is too expensive.



In general:

- ➔ We do not know the best length of time to take prednisone. How long you take it should be based on your condition and personal preferences.

Surgical treatment recommendations

If you need surgery for GCA:

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

If you have severe GCA, worsening signs of limb (arms, hands, legs, or feet) or organ ischemia, and are on immunosuppressants:

- ➔ Increase immunosuppressants instead of surgery.
 - When **might** I get surgery?
 - If you have an aortic aneurysm that is highly likely to rupture (burst open).
 - If you have signs of tissue death or damage, or organ damage.

If you are getting surgery because of a GCA health problem (such as an aneurysm or narrowing of an artery):

- ➔ Use high-dose prednisone shortly before, during, and after the surgery.

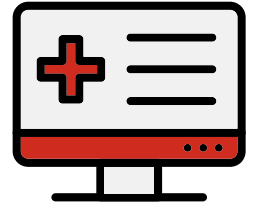


Monitoring recommendations

If you have **GCA 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 how often you need monitoring will depend on your specific situation.



If you have **GCA, and higher levels of inflammation markers with no other symptoms of GCA:**

➔ **Have clinical monitoring, over increasing immunosuppressants.**

- Higher levels of these markers may not be related to GCA. But you may need to have more frequent tests that look for signs of active disease.



Health terms

- A**
 - **Aorta:** The main artery in your body that carries blood from your heart to the rest of your body.
 - **Aortic aneurysm:** A bulge in the wall of your aorta.
- C**
 - **Cranial ischemia:** When there is not enough blood flow to your brain, which can cause vision loss and strokes.
- 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 as a sign of inflammation.
 - **Ischemia:** Lowered blood flow to a part of your body.
- M**
 - **Methotrexate:** An immunosuppressant that 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. Types of noninvasive imaging:
 - **Computed tomography (CT) angiography:** Uses multiple X-rays to make pictures of blood vessels.
 - **Magnetic resonance (MR) angiography:** Uses strong magnets to make pictures of blood vessels.
 - **PET scan:** Uses a radioactive tracer to show how well certain parts of your body are working. A radioactive tracer is a material that gives off low levels of radiation that the machine can detect.
 - **Vascular ultrasound:** Uses sound waves to show how blood flows through vessels.
- P**
 - **Prednisone:** A medicine that 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.
- S**
 - **Statins:** Medicines that help lower cholesterol.
- T**
 - **Temporal artery biopsy:** A procedure to remove a small section of your artery to test for GCA.
 - **Tocilizumab:** An immunosuppressant that lowers inflammation (swelling) in your body.



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.

