

Recommendations for specific symptoms in active GPA



These are recommendations written by a group of doctors and patients based on research studies* for people with a new active diagnosis of GPA vasculitis. They are for “remission induction,” which means getting to a place where there are no active symptoms and no new damage to the body from vasculitis.

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

What is vasculitis?

Vasculitis: A group of conditions that involves inflammation (swelling) of the blood vessels. ANCA-associated vasculitis (AAV) is a group of disorders that include GPA. GPA is a type of vasculitis that can affect blood vessels in your nose, sinuses, throat, lungs, and kidneys.

Active disease: New, ongoing, or worsening signs or symptoms.



Healthy blood vessel



Inflamed blood vessel

Recommendations for sinonasal symptoms in people with GPA

If you have nose or sinus symptoms, such as swelling in your nose or sinuses, runny nose, nosebleeds, or nasal crusting, we recommend:

➔ Try nasal rinses and topical nasal therapies, such as antibiotics, lubricants, and prednisone

- To see if these may help you, we suggest seeing an ENT doctor who has experience treating GPA.

Recommendations for airway symptoms in people with GPA

If you have actively inflamed subglottic and/or endobronchial tissue with stenosis, we recommend:

➔ Use immunosuppressive treatments over surgical dilation with steroid injection alone

- When **might** I get surgical dilation with intralesional steroid injection?
 - If you have had the stenosis for a long time, it is fibrotic (damaged or scarred), or it doesn't get better after immunosuppressive treatment.
 - If the stenosis needs treatment right away, such as because it is stopping your body from getting enough air (called critical narrowing) – in this case, it may be used with other treatments.

Recommendations for mass lesions in people with GPA

If you have mass lesions, we recommend:

➔ Use immunosuppressive treatments over debulking surgery with immunosuppressive treatment

- When **might** I get debulking surgery?
 - If there is an urgent need to lower the pressure from the lesion (called decompression). This can include pressure on the optic nerve that could cause vision loss, or pressure that could threaten an organ or cause death.

*Chung, S.A., Langford, C.A., Maz, M., et al. 2021 American College of Rheumatology/Vasculitis Foundation Guideline for the Management of Antineutrophil Cytoplasmic Antibody–Associated Vasculitis. *Arthritis Rheumatol*, 73: 1366-1383. <https://doi.org/10.1002/art.41773> You can find the full ACR/VF recommendations at www.vasculitisfoundation.org.



Health terms

- **Debulking surgery:** A surgery to remove a lesion.
- **ENT doctor:** A doctor who treats the ear, nose, and throat.
- **Immune system:** A system of cells, tissues, and organs that helps the body fight infection or illness.
- **Immunosuppressive treatments:** Lowers the body's immune response to stop the immune system from causing inflammation (swelling) and damaging the body.
- **Inflamed subglottic or endobronchial tissue with stenosis:** When a part of the windpipe or lungs narrows, usually caused by swelling or scar tissue.
- **Mass lesion:** Non-cancerous, tumor-like masses or swollen tissue.
- **Prednisone:** A type of steroid. It can be given as:
 - **IV pulse** – A tube into a vein as an IV
 - **Oral** – A pill by mouth
- **Stenosis:** When something becomes narrower.
- **Steroid:** Lowers inflammation (swelling) in the body.
- **Surgical dilation with intralesional steroid injection:** Surgery to inject a steroid directly into a lesion.