

Medication recommendations for active GPA/MPA



These are recommendations written by a group of doctors and patients based on research studies* for people with a new active diagnosis of GPA or MPA 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:** A type of vasculitis that can affect blood vessels in your nose, sinuses, throat, lungs, and kidneys.
- **MPA:** A type of vasculitis that mostly affects small to medium-sized blood vessels. It can affect the kidneys, lungs, nerves, skin, and joints.

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

Severe disease: Symptoms that may cause death or organ failure.

Nonsevere disease: Symptoms not likely to cause death or organ failure.



Healthy blood vessel



Inflamed blood vessel

Medication recommendations for GPA/MPA

If you have active GPA/MPA, in general, we recommend:

⌚ **Not changing your dose of immunosuppressive therapy based on ANCA test results alone. Instead, base all treatment decisions on symptoms along with diagnostic tests, such as labs, imaging, and biopsy findings.**

- Why?
 - A high ANCA level does not always predict if you will have a disease flare (worsening symptoms).
 - If immunosuppressive therapy is increased based on ANCA levels alone, it can lead to suppressing the immune system too much, which can cause harmful side effects like infections.

If you have active GPA and you take immunosuppressive therapy (such as cyclophosphamide or rituximab), we recommend:

⌚ **Use antimicrobial medicines (such as trimethoprim/sulfamethoxazole) to prevent pneumonia caused by a fungus called *Pneumocystis jirovecii***

- Why?
 - Medicines for vasculitis can lower your body’s ability to fight infection. This can make you more likely to get pneumonia, which is a serious lung infection.
 - Antimicrobial medicines fight microbes such as the fungus that causes pneumonia.
- When **should** I take trimethoprim/sulfamethoxazole?
 - If you take cyclophosphamide or rituximab.

- When **might** I take trimethoprim/sulfamethoxazole?
 - If you are taking prednisone with methotrexate, azathioprine, or mycophenolate mofetil.

If you have active GPA/MPA and can't have other immunosuppressive therapy (such as cyclophosphamide or rituximab), for example for an infection, we recommend:

➔ Get IVIG (intravenous immunoglobulin)

- Why?
 - IVIG is an immunomodulatory therapy (which changes your immune system), not an immunosuppressant. It does not raise your chance of infection.



*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

- **ANCA** (antineutrophil cytoplasmic antibodies): An antibody that recognizes a type of white blood cell called neutrophils. ANCA tests help doctors diagnose certain types of vasculitis.
- **Antibodies**: Proteins in your immune system that fight infections.
- **Antimicrobial medicines**: Fight microbes such as the bacteria and fungus that cause infections like pneumonia.
- **Azathioprine**: An immunosuppressant. Lowers inflammation (swelling) in the body.
- **Cyclophosphamide**: An immunosuppressant. Lowers inflammation (swelling) in the body.
- **Immune system**: A system of cells, tissues, and organs that helps the body fight infection or illness.
- **Immunosuppressive therapy**: Lowers the body's immune response to stop the immune system from causing inflammation (swelling) and damaging the body.
- **IVIG** (intravenous immunoglobulin): Immunoglobulin given as an IV through a vein in the arm. Immunoglobulin is a part of your blood that has antibodies to help you fight germs and disease.
- **Methotrexate**: An immunosuppressant. Lowers inflammation (swelling) in the body.
- **Mycophenolate mofetil**: An immunosuppressant. Lowers inflammation (swelling) in the body.
- ***Pneumocystis jirovecii* pneumonia** (PCP): A serious infection caused by the fungus *Pneumocystis jirovecii*.
- **Prednisone**: Lowers inflammation (swelling) in the body, and can be given as:
 - **IV pulse**: A tube into a vein as an IV
 - **Oral**: A pill by mouth
- **Rituximab**: An immunosuppressant that lowers the number of B cells (a type of white blood cell) to lower inflammation (swelling).
- **Sepsis**: A dangerous reaction to an infection that can cause inflammation (swelling) and low blood pressure, which can lead to tissue damage, organ failure, and even death.
- **Trimethoprim/sulfamethoxazole**: 2 medicines taken together that kill bacteria or fungi that cause some infections.