

# Recommendations for conditions related to 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

## Recommendations for conditions related to GPA/MPA

**If you have active, severe GPA/MPA with glomerulonephritis, we recommend:**

### ➔ Not everyone needs plasma exchange

- Why?
  - Plasma exchange can raise your chance of serious infection.
  - The risk of infection may outweigh the benefit of plasma exchange if you have a low chance of kidney failure.
- When **should** I get plasma exchange?
  - If you have anti-glomerular basement membrane disease.
- When **might** I get plasma exchange?
  - If you have a high chance of kidney failure, then the benefit of plasma exchange may outweigh the risk of infection.
  - If you are critically ill and other treatments have not worked.

If you have active, severe GPA/MPA with alveolar hemorrhage, we recommend:

➔ **Not adding plasma exchange to your treatment**

- Why? Studies have **not** shown that plasma exchange helps people with alveolar hemorrhage, and it can raise your chance of serious infection.
- When **should** I get plasma exchange?
  - If you have anti-glomerular basement membrane disease.
- When **might** I get plasma exchange?
  - If you have glomerulonephritis with a high chance of kidney failure.
  - If you are critically ill and other treatments have not worked.

\*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](http://www.vasculitisfoundation.org).



## Health terms

- **Alveolar hemorrhage:** Bleeding in the lungs that fills the alveoli. Alveoli are tiny air sacs in your lungs that fill with oxygen as you breathe.
- **Anti-glomerular basement membrane disease:** An autoimmune disease that affects your kidneys and lungs.
- **Blood thinners:** Medicines that prevent blood clots. Blood clots can lead to heart attacks or strokes.
- **Glomerulonephritis:** Inflammation (swelling) of the tiny blood vessels (glomeruli) in the kidneys that filter wastes and extra fluid from the blood.
- **Kidney failure or end-stage renal disease (ESRD):** The last stage of chronic kidney disease in which the kidneys do not work well enough for someone to live without dialysis or a transplant.
- **Plasma exchange:** Removes the harmful materials in the blood by removing a person's plasma (the liquid part of blood) and replacing it with donor plasma or plasma substitute.
- **Venous thrombotic events:** Blood clots that block the flow of blood through your veins.