

Treatment recommendations for active, severe EGPA



These are recommendations written by a group of doctors and patients based on research studies* for people with a new active diagnosis of EGPA 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 includes EGPA. EGPA is one of the rarest forms of vasculitis that mostly affects the small blood vessels. It can affect the lungs, sinuses, skin, heart, digestive tract, kidneys, nerves, and other organs.

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

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



Healthy blood vessel



Inflamed blood vessel

Treatment recommendations for active, severe EGPA

If you have active, severe EGPA, we recommend:

➔ Use either IV pulse prednisone or high-dose oral prednisone as a part of initial therapy

- Why?
 - We recommend either because there are no studies that compare how well they work.
 - Either choice should be used with cyclophosphamide or rituximab.

➔ Use either cyclophosphamide or rituximab

- Why?
 - We recommend either because there are no studies that show one is better than the other for EGPA.
- When **might** I use cyclophosphamide?
 - If you have heart problems, such as cardiomyopathy. This symptom can suggest a higher risk of death in EGPA.
 - There is more experience treating EGPA with cyclophosphamide.
 - If you have negative ANCA results.
 - If you have severe neurologic (nervous system) or gastrointestinal (digestive) symptoms.
- When **might** I use rituximab?
 - If you have positive ANCA results.
 - If you have active glomerulonephritis.
 - If you previously had cyclophosphamide treatment, and it hasn't worked, or you want to try another treatment.
 - If you are concerned about becoming infertile (unable to have children) from cyclophosphamide.

➔ Use cyclophosphamide or rituximab instead of mepolizumab

- Why?
 - There are no studies that show how well mepolizumab works for severe EGPA.



*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 attacks a type of white blood cell called neutrophils. ANCA tests help doctors diagnose certain types of vasculitis.
- **Cardiomyopathy**: Problems with the heart muscle that can make it harder for the heart to pump blood.
- **Cyclophosphamide**: An immunosuppressant. Lowers inflammation (swelling) in the body.
- **Glomerulonephritis**: Inflammation (swelling) of the tiny blood vessels (glomeruli) in the kidneys that filter wastes and extra fluid from the blood.
- **Mepolizumab**: A treatment that lowers the number of eosinophils, a type of white blood cell, to lower inflammation (swelling).
- **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**: A treatment that lowers the number of B cells, a type of white blood cell, to lower inflammation (swelling).