

Treatment recommendations for active, nonsevere 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.

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



Healthy blood vessel



Inflamed blood vessel

Treatment recommendations for active, nonsevere EGPA

If you have active, nonsevere EGPA, we recommend:

➔ Use mepolizumab and prednisone over:

- Methotrexate and prednisone
- Azathioprine and prednisone
- Mycophenolate mofetil and prednisone
- Why?
 - There are studies showing that mepolizumab works to treat active, nonsevere EGPA.
 - The other medicines have not been studied in a clinical trial to test how well they work in EGPA.
- When would I **not** use mepolizumab?
 - Certain individual factors, such as an allergic reaction or serious side effects. In this case, you can use either methotrexate, azathioprine, or mycophenolate mofetil. There are no studies that show one works better than the others.

➔ Use methotrexate and prednisone, azathioprine and prednisone, or mycophenolate mofetil and prednisone over:

- Prednisone alone
- Rituximab and prednisone
- Cyclophosphamide and prednisone
 - Why?
 - Prednisone can cause negative side effects. Combining methotrexate, azathioprine, or mycophenolate mofetil with prednisone lessens the amount of prednisone you take. This lowers the chances you will have these side effects.

- Rituximab can cause serious side effects.
- Cyclophosphamide can cause serious side effects and should be the last option.
- When **might** I use prednisone alone?
 - If you have mild asthma.
 - If you have allergic reactions to other medicines.
 - If you are pregnant.
 - If you have other individual situations.
- When **might** I use rituximab and prednisone?
 - If you have nonsevere vasculitis and a positive ANCA test.
 - If you aren't improving on other treatments.

*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.
- **Azathioprine**: An immunosuppressant. Lowers inflammation (swelling) in the body.
- **Clinical trial**: A research study designed to learn if new medicines or treatments are safe and work well in people.
- **Cyclophosphamide**: An immunosuppressant. Lowers inflammation (swelling) in the body.
- **Mepolizumab**: A treatment that lowers the number of eosinophils, a type of white blood cell, to lower inflammation (swelling).
- **Methotrexate**: An immunosuppressant. Lowers inflammation (swelling) in the body.
- **Mycophenolate mofetil**: An immunosuppressant. Lowers inflammation (swelling) in the body.
- **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).