

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).

Treatment recommendations for active, nonsevere EGPA



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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

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- **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).

General recommendations for EGPA



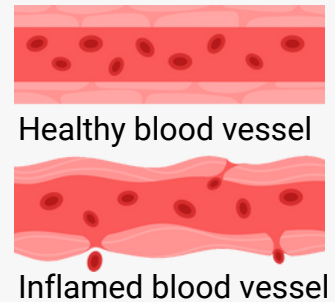
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Active disease: New, ongoing, or worsening signs or symptoms.



General recommendations for EGPA

If you have EGPA, in general, we recommend:

➔ Get an echocardiogram at the time of your diagnosis, even if you don't have heart symptoms

- Why?
 - An echocardiogram can find many heart problems, and heart problems are the main cause of death related to EGPA. If heart problems are found, it can guide treatment choices.
 - Not finding heart problems early in people with EGPA could lead to worse health outcomes.

➔ Your doctor should use the Five-Factor Score (FFS) to guide treatment

- What is the FFS?
 - A tool doctors can use to score symptoms of EGPA. Higher scores can mean a worse outcome. It has also been used to guide treatment choices.
 - However, we don't know how it applies to newer treatments such as mepolizumab or rituximab.

If you have EGPA and active asthma and/or nose or sinus symptoms:

➔ You can start or continue leukotriene inhibitors

- Why was there concern about taking leukotriene inhibitors?
 - Some experts were concerned that leukotriene inhibitors could cause EGPA. But studies have not found a link. There is no reason not to take these medicines, as they can help manage these conditions.
 - However, leukotriene inhibitors should not be used for other symptoms.

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<https://doi.org/10.1002/art.41773> You can find the full ACR/VF recommendations at www.vasculitisfoundation.org.

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

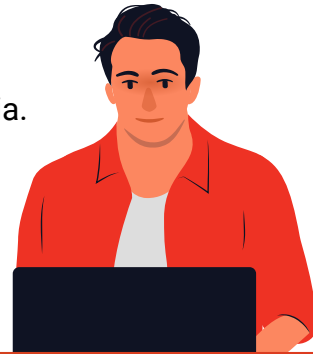
➔ Try nasal rinses and topical nasal therapies, such as antibiotics, lubricants, and glucocorticoids

- You may benefit from these, even though we don't know how well they work in EGPA.
- To see if these may help you and to choose the best option, we suggest seeing an ENT doctor who has experience treating EGPA.

If you have EGPA and you take immunosuppressants, we recommend:

➔ Use certain 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 (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.



Health terms

- **Antimicrobial medicines:** Fight microbes such as the fungus that causes pneumonia.
- **Azathioprine:** An immunosuppressant. Lowers inflammation (swelling) in the body.
- **Cyclophosphamide:** An immunosuppressant. Lowers inflammation (swelling) in the body.
- **Echocardiogram:** An ultrasound that uses sound waves to show how blood flows through your heart and heart valves.
- **ENT doctor:** A doctor who treats the ear, nose, and throat.
- **Immunosuppressant:** A type of treatment that lowers the body's immune response to stop the immune system from causing inflammation (swelling) and damaging the body.
- **Leukotriene inhibitors:** A type of medicine to treat allergies, asthma, or other nose or sinus symptoms.
- **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.
- ***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:** A treatment that lowers the number of B cells, a type of white blood cell, to lower inflammation (swelling).
- **Trimethoprim/sulfamethoxazole:** 2 medicines taken together that kill bacteria or fungi that cause infections in the body.