

# Recommendations for EGPA\*



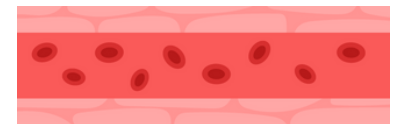
These are recommendations written by a group of doctors and patients based on research studies for people with EGPA.

These recommendations suggest the best treatment for most people, but your individual situation and treatment may be different. Talk to your doctor about what treatment is best for you.

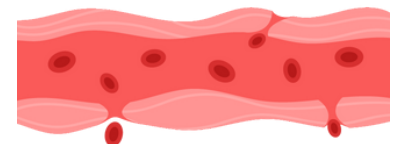
## What is vasculitis?

**Vasculitis:** A group of conditions that involve inflammation (swelling) of your blood vessels. ANCA-associated vasculitis (AAV) is a group of disorders that include:

- **EGPA:** One of the rarest types of vasculitis that mostly affects the small blood vessels. It can affect your lungs, sinuses, skin, heart, digestive tract, kidneys, nerves, and other organs.
- **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 your kidneys, lungs, nerves, skin, and joints.



Healthy blood vessel



Inflamed blood vessel

## Words to know about vasculitis

- **Active disease:** New, ongoing, or worsening signs or symptoms.
- **Severe disease:** Symptoms that may cause death or organ failure (when a major organ stops working, such as the heart).
- **Nonsevere disease:** Symptoms not likely to cause death or organ failure.
- **Refractory:** When a disease does not get better with treatment.
- **Relapse:** When symptoms return after a period of improvement (or remission).
- **Remission:** When the signs and symptoms of a disease decrease or go away.
  - **Remission induction treatment:** Treatments for active disease to get to a place where there are no active symptoms and no new damage to your body from the disease.
  - **Remission maintenance:** Treatments given after a disease goes into remission. These treatments help keep the disease in remission.

See a glossary of health terms at the end of this document.

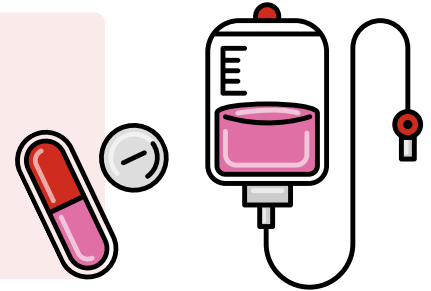


# Treatment recommendations for active EGPA

If you have active, **severe EGPA**, we recommend:

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

- 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.**

- 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, which can suggest a higher risk of death in EGPA.
  - 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 active glomerulonephritis.
  - If you have positive ANCA results.
  - 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 over mepolizumab.**

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



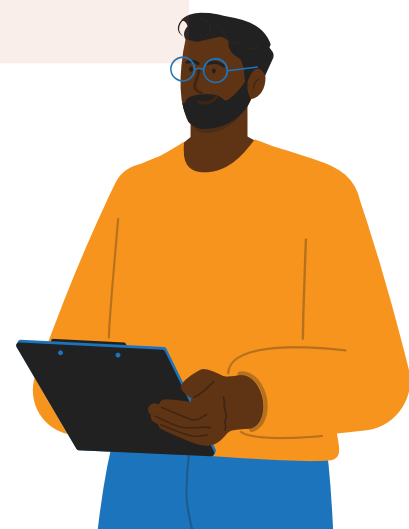
## If you have **active, nonsevere EGPA**, we recommend:

### ➔ Use mepolizumab and prednisone over:

- Methotrexate and prednisone
- Azathioprine and prednisone
- Mycophenolate mofetil and prednisone
  - 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
  - Prednisone can cause negative side effects. Combining methotrexate, azathioprine, or mycophenolate mofetil with prednisone lessens the amount of prednisone you take. This lowers the chance 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.
- 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.



# Treatment recommendations for remission maintenance

If you have **severe EGPA** and it has entered remission, we recommend:

➔ **Use methotrexate, azathioprine, or mycophenolate mofetil, over mepolizumab or rituximab for remission maintenance.**

- We know less about how well mepolizumab or rituximab work for remission maintenance.
- When **might** I use rituximab?
  - If rituximab was your remission induction treatment.
  - If you don't tolerate other treatments.

➔ **Regarding prednisone:**

- The length of time you use prednisone for remission maintenance should depend on your clinical condition, preferences, and values.
- With prednisone, remission maintenance treatment should be balanced with potential side effects. Using prednisone for a long time gives you a higher chance of infections, high blood pressure, weak bones, weight gain, and eye problems. But it may help you control your vasculitis.

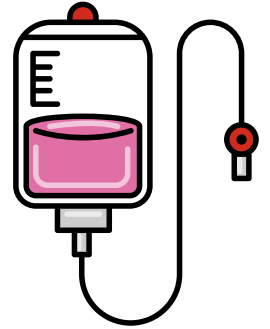


# Treatment recommendations for disease relapse

## If you have **EGPA** and it relapsed with severe symptoms after remission induction treatment, we recommend:

### ➔ If you used cyclophosphamide for remission induction:

- Use rituximab over cyclophosphamide to treat the relapse.
  - It is best to avoid cyclophosphamide again because it can cause serious side effects. The more cyclophosphamide you use, the higher the chance of serious side effects.
- When **might** I use cyclophosphamide?
  - If EGPA is affecting your heart.



### ➔ If you used rituximab for remission induction:

- Use rituximab again over switching to cyclophosphamide to treat the relapse.
  - Cyclophosphamide can cause serious side effects.
- When **might** I use cyclophosphamide?
  - If you have heart-related symptoms.
  - If the relapse happened shortly after rituximab treatment.

## If you have **EGPA** and it relapsed with nonsevere symptoms (such as asthma and/or nose or sinus symptoms), we recommend:

### ➔ If you are using methotrexate, azathioprine, or mycophenolate mofetil:

- Add mepolizumab over switching to another treatment.
  - We know mepolizumab works well for relapsing nonsevere EGPA.

### ➔ If you are using only low-dose prednisone:

- Add mepolizumab over methotrexate, azathioprine, or mycophenolate mofetil.

### ➔ If you have high IgE levels and are using methotrexate, azathioprine, or mycophenolate mofetil:

- Add mepolizumab over omalizumab.
  - We know less about how well omalizumab works for EGPA.



# General recommendations

## 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.**

- 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.**

- FFIS is 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 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.
- We suggest seeing an ENT doctor who has experience treating EGPA to see if these may help you and to choose the best option.

## If you have **EGPA** and active asthma and/or nose or sinus symptoms, we recommend:

### ➔ **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.

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

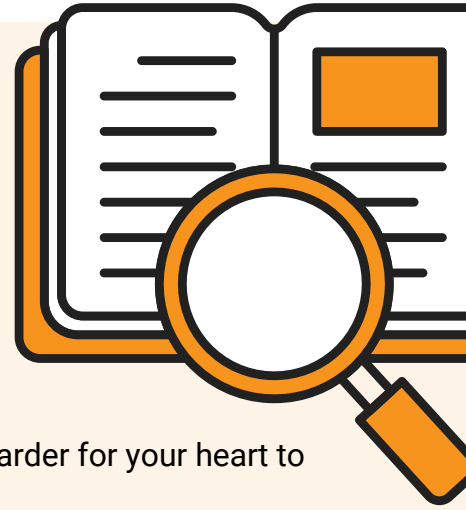
### ➔ Use certain antimicrobial medicines (such as trimethoprim/sulfamethoxazole) to prevent pneumonia caused by a fungus called *Pneumocystis jirovecii*.

- 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.



## Health terms

- A**
  - **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.
  - **Antimicrobial medicines:** Medicines that fight microbes such as the bacteria and fungus that cause infections like pneumonia.
  - **Azathioprine:** An immunosuppressant. Lowers inflammation (swelling) in your body.
- C**
  - **Cardiomyopathy:** Problems with the heart muscle that can make it harder for your heart to pump blood.
  - **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 your body.
- E**
  - **Echocardiogram:** An ultrasound that uses sound waves to show how blood flows through your heart and heart valves.
  - **ENT doctor:** A doctor who treats diseases of your ear, nose, and throat.
- G**
  - **Glomerulonephritis:** Inflammation (swelling) of the tiny blood vessels (glomeruli) in your kidneys that filter waste and extra fluid from your blood.
- I**
  - **IgE:** A type of antibody, which are proteins in your immune system that fight infections.
  - **Immunosuppressant:** A medicine that lowers your body's immune response to stop your immune system from causing inflammation (swelling) and damaging your body.
- L**
  - **Leukotriene inhibitors:** A type of medicine to treat allergies, asthma, or other nose or sinus symptoms.
- M**
  - **Mepolizumab:** A medicine that lowers your body's immune response by lowering the number of eosinophils. Eosinophils are a type of white blood cell in your immune system. This medicine helps lower inflammation (swelling).
  - **Methotrexate:** An immunosuppressant. Lowers inflammation (swelling) in your body.
  - **Mycophenolate mofetil:** An immunosuppressant. Lowers inflammation (swelling) in your body.





**P**

- ***Pneumocystis jirovecii* pneumonia (PCP):** A serious lung infection caused by the fungus *Pneumocystis jirovecii*.
- **Prednisone:** A medicine that lowers inflammation (swelling) in your body, and can be given as:
  - **IV pulse** – A tube into a vein as an IV.
  - **Oral** – A pill by mouth.

**R**

- **Rituximab:** A medicine that lowers the number of B cells (white blood cells) in your immune system to lower inflammation (swelling).

**T**

- **Trimethoprim/sulfamethoxazole:** 2 medicines taken together that kill bacteria or fungi that cause infections.

\*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).

