

General recommendations for EGPA



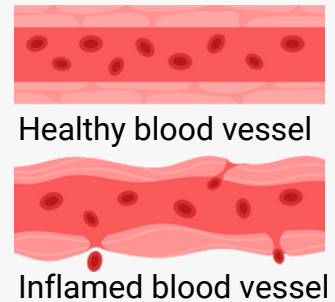
These are recommendations written by a group of doctors and patients based on research studies* for people with a new active diagnosis of EGPA vasculitis. They are for “remission induction,” which means getting to a place where there are no active symptoms and no new damage to the body from vasculitis.

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.



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.

*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.

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

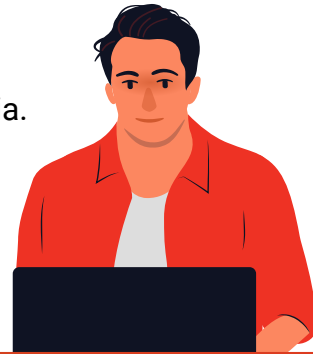
➔ Try nasal rinses and topical nasal therapies, such as antibiotics, lubricants, and glucocorticoids

- 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.