

What is Granulomatosis with Polyangiitis (GPA)?

Granulomatosis with polyangiitis (GPA) is a form of vasculitis. Vasculitis means inflammation of the blood vessels. Granulomatosis means that “granulomas” (harmful inflammatory tissue) form in tissues and organs. Polyangiitis means many arteries. Inflammation of blood vessels may result in narrowing of the vessel, which can block or slow down blood flow to vital organs and tissues. GPA can worsen rapidly, so early diagnosis and treatment are important to prevent organ damage or failure.

Symptoms

The symptoms of GPA can vary greatly from person to person depending on the organs that are affected. For some, the disease is mild, while for others it may be severe or even life-threatening. GPA symptoms may come on slowly over a period of months, or develop rapidly in a matter of days. Early symptoms can include:

- Runny nose/nasal congestion
- Nosebleeds
- Sinus pain
- Ear pain/hearing loss
- Shortness of breath
- Coughing up blood
- Hoarseness

Other symptoms include:

- Fever
- Fatigue
- Joint pain
- Loss of appetite/weight loss
- Eye problems
- Skin sores
- Night sweats
- Numbness in the fingers, toes, or limbs
- Kidney problems*

*Note: Kidney disease can be present without symptoms; therefore, patients with vasculitis should have regular urine tests.

When to see Your Doctor

See your doctor if you have cold symptoms that don't get better with over-the-counter cold medications, especially if you have nosebleeds or are coughing up blood. Also, if you have any of the symptoms and they don't away.

Causes

The cause of GPA is not yet fully understood. Vasculitis is classified as an autoimmune disorder—a disease which occurs when the body's natural defense system mistakenly attacks healthy tissues. Researchers believe an infection may cause GPA to start. But an infection alone cannot explain this complex disorder.

Environmental and genetic factors may play a role as well. Research is ongoing, but so far, no specific infectious, genetic, or environmental factors have been associated with GPA.

Who Gets GPA?

Although GPA can occur at any age, it usually starts between 40 and 65 years of age. The disease is rare in children but does occur. GPA can affect people of any race or ethnic background but appears to mostly affect Caucasians. Because GPA often goes unrecognized, researchers believe it is underdiagnosed, making it difficult to know how often it happens. GPA affects an estimated 3 out of every 100,000 people.

Diagnosis

There is no single test for diagnosing GPA, so your doctor will review your symptoms, medical history, physical exam findings, laboratory tests, and imaging studies. A biopsy of the affected tissue may be needed to confirm the diagnosis.

- **Urine tests:** A urinalysis can detect red blood cells or excess protein in the urine, which can indicate whether the kidneys are being affected.
- **Blood tests:** The most common blood test for GPA checks for an antibody called ANCA, or anti-neutrophil cytoplasmic antibody. The ANCA test is positive in most individuals with GPA, so it may help support a suspected diagnosis of GPA. However, a positive test alone does not confirm the diagnosis and a negative test does not exclude the possibility of GPA.
- **Imaging studies:** Chest x-rays can show changes in the lungs. Computed tomography (CT) scans and magnetic resonance imaging (MRI) provide more detailed abnormalities of the internal organs.

- **Tissue biopsy:** This surgical procedure removes a small tissue sample from an affected organ, such as the lung, kidney, or skin, which is examined under a microscope for signs of inflammation or tissue damage.

Treatment

GPA is a potentially serious but treatable disease. Treatment goals are to reduce and control the inflammation. It depends on the organs affected and disease severity for what medications will be used. Even with effective treatment, relapse can occur, so follow-up visits with your doctor are important.

To control inflammation, patients with milder disease may take corticosteroids such as prednisone, along with medications that keep the immune system from overreacting. This may be methotrexate, mycophenolate mofetil, or azathioprine.

Patients with moderate to severe disease may be prescribed the biologic drug, rituximab, used with corticosteroids. Biologics target certain parts of the immune system to control inflammation.

Another option for those with severe disease is cyclophosphamide—a chemotherapy-type drug that blocks abnormal growth of certain cells in the body. It is used in combination with prednisone. Cyclophosphamide is usually limited to a three- to six-month period, then replaced with less toxic drugs such as methotrexate, mycophenolate mofetil, or azathioprine.

Once the disease is in remission, you may most likely need to continue taking maintenance medications, such as azathioprine, methotrexate or rituximab, to keep the disease under control. The dose of steroids is usually tapered during remission.

Some individuals may experience kidney failure, a serious complication that requires dialysis and/or a kidney transplant.

Side Effects of Medications

The medications used to treat GPA have potentially serious side effects, such as lowering your body's ability to fight infection, and potential bone loss (osteoporosis), among others. Therefore, it's important to see your doctor for regular checkups. Medications may be prescribed to offset side effects.

Infection prevention is also very important. Talk to your doctor about getting a flu shot, pneumonia vaccination, and/or shingles vaccination, which can reduce your risk of infection.

Relapse

GPA is a chronic disease with periods of relapse and remission. If your initial symptoms return or you develop new ones, report them to your doctor as soon as possible. Regular doctor visits and ongoing monitoring of lab and imaging tests are important in detecting relapses early.

Complications

Serious complications may occur, including bleeding and scarring of the lungs, kidney damage or failure, heart disease, hearing loss, skin scarring, deep vein thrombosis (blood clot), or damage to the bridge of the nose from weakened cartilage.

Your Medical Team

Effective treatment of GPA may require the coordinated efforts and ongoing care of a team of medical providers and specialists. In addition to a primary care provider, you may need to see the following specialists, depending on which organ and systems are involved:

- Rheumatologist (joints, muscles, and immune system)
- Pulmonologist (lung)
- Otolaryngologist (ear, nose and throat)
- Nephrologist (kidneys)
- Dermatologist (skin)
- Cardiologist (heart)
- Neurologist (brain/and nervous system); or others as needed

The best way to manage your disease is to actively partner with your health care providers and get to know the members of your health care team.

It may be helpful to use a health care journal to keep track of medications, symptoms, test results and notes from doctor appointments.

To get the most out of your doctor visits, make a list of questions and bring along a supportive friend or family member to provide a second set of ears and take notes.

Remember, it's up to you to be your own advocate. If you have concerns with your treatment plan, speak up. Your doctor may be able to adjust your dosage, or offer different treatment options. It is always your right to seek a second opinion.

Living with GPA

Living with a chronic disease such as GPA can be challenging at times. Fatigue, pain, emotional stress, and medication side effects can take a toll on your sense of well-being, affecting

relationships, work and other aspects of your daily life. Sharing your experience with family and friends, connecting with others through a support group or talking with a mental health professional can help.

Outlook

There is no cure for GPA at this time, but early diagnosis and effective treatment can bring the disease into remission, and many patients can lead full, productive lives. Left untreated, GPA can lead to potentially life-threatening organ damage or failure. Even with treatment, many patients will experience periods of relapse, so ongoing medical care is important.

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