CONDITION

Granulomatosis with polyangiitis

Granulomatosis with polyangiitis is an immune condition that inflames blood vessels, often affecting the airways, lungs, and kidneys.

WHAT IT IS
Inflammation of small and medium blood vessels

BODY SYSTEM
Blood vessels, airways, lungs, and kidneys

MAIN SYMPTOMS
Sinus problems, nosebleeds, cough, fatigue

WHAT CAUSES IT
Misguided immune attack; exact trigger unknown

HOW IT IS DIAGNOSED
Blood and urine tests, imaging, and tissue biopsy

MAIN TREATMENT
Immune-suppressing medicines and glucocorticoids

CAN IT BE CURED?
No - remission is possible, but relapse can occur

IS IT AN EMERGENCY?
Sometimes - coughing blood or severe breathlessness

LONG-TERM OUTLOOK
Remission is common, but relapses and organ damage can occur

Overview

Granulomatosis with polyangiitis, or GPA, is a rare condition that causes inflammation in blood vessel walls. It belongs to a group called vasculitis , meaning inflammation of blood vessels.

GPA most often affects small and medium blood vessels in the nose, sinuses, ears, lungs, and kidneys. It can also affect the eyes, skin, nerves, joints, heart, or other organs.

The inflammation can reduce blood flow and damage nearby tissue. Kidney or lung involvement may become serious without prompt treatment. However, modern treatments can often bring GPA into remission , a period when the condition is controlled and causes few or no symptoms.

GPA was previously called Wegener's granulomatosis. The current name describes two

important features: inflammatory cell clusters called granulomas and inflammation of blood vessels called polyangiitis.

Symptoms

Symptoms differ according to which organs are affected. They may develop gradually over weeks or months, although severe lung or kidney problems can appear more quickly.

Symptoms involving the nose, sinuses, and ears may include:

Lung symptoms can include a persistent cough, chest discomfort, shortness of breath, or coughing up blood. Some lung changes cause no obvious symptoms and are found on imaging.

Kidney inflammation often causes no pain or early warning symptoms. Blood and protein may appear in the urine before you notice any change. More advanced kidney problems can cause swollen ankles, less urine, nausea, breathlessness, or high blood pressure.

Other possible symptoms include:

Many of these symptoms have more common causes. Having one of them does not mean that you have GPA.

- A blocked or persistently runny nose
- Frequent nosebleeds or nasal crusting
- Sinus pain or repeated sinus inflammation
- Ear pain, hearing loss, or a feeling of fullness in the ears
- Hoarseness or noisy breathing
- Tiredness, fever, night sweats, or unintended weight loss
- Aching or swollen joints
- Red or purple skin spots, sores, or tender lumps
- Eye redness, pain, swelling, or changes in vision
- Numbness, burning pain, or weakness in a hand or foot

Causes and risk factors

GPA is an autoimmune condition . This means the immune system mistakenly attacks the body's own tissues, including blood vessel walls. The exact reason this begins is unknown.

Many people with GPA have proteins in their blood called antineutrophil cytoplasmic

antibodies , usually shortened to ANCA. These antibodies target certain white blood cells and are linked with blood vessel inflammation. The ANCA type most strongly associated with GPA targets a protein called proteinase 3, or PR3.

ANCA may play a role in the inflammation, but it does not fully explain why GPA develops. Genes may affect susceptibility, while infections or environmental exposures may act as triggers in some people. GPA is not usually inherited directly from a parent, and it is not contagious.

There is no single lifestyle choice known to cause GPA.

Diagnosis

GPA can resemble infections, cancer, allergies, or other inflammatory conditions. Diagnosis usually combines your symptoms, examination findings, laboratory tests, imaging, and sometimes a biopsy.

Blood tests may look for inflammation, anemia, kidney function, and ANCA. A positive ANCA result can support the diagnosis, especially when symptoms fit GPA. However, some people with GPA have a negative result, and ANCA can be positive in other conditions.

Urine tests are important because kidney inflammation may be present without symptoms. They can detect blood, protein, and abnormal cell clusters called casts. Blood tests measure how well the kidneys are filtering.

Chest imaging may show lung nodules, cavities, or areas of bleeding or inflammation. CT scans can provide more detail than standard X-rays. Imaging of the sinuses may also be useful.

A biopsy removes a small tissue sample for examination under a microscope. Samples may come from a kidney, lung, skin lesion, or another affected area. A biopsy can provide the strongest confirmation, although it is not always possible or necessary before urgent treatment begins.

Doctors may also test for infections and other conditions before starting medicines that suppress the immune system.

Treatment and management

Treatment aims to stop active inflammation, prevent organ damage, and maintain remission. Care is often coordinated by specialists in rheumatology, kidney medicine, lung medicine, or ear, nose, and throat care.

Treatment usually has two stages. Induction treatment brings active GPA under control. Maintenance treatment helps prevent it from returning.

For GPA that threatens the kidneys, lungs, or another major organ, induction commonly includes rituximab or cyclophosphamide along with a glucocorticoid such as prednisone. Rituximab targets specific immune cells. Cyclophosphamide broadly reduces immune activity. An oral medicine called avacopan may sometimes be used as part of treatment to reduce glucocorticoid exposure.

Less organ-threatening GPA may be treated with medicines such as methotrexate, depending on the organs involved and the person's overall health.

Once remission is reached, medicines such as rituximab, azathioprine, methotrexate, or mycophenolate may be used for maintenance. The choice and duration are based on relapse risk, previous treatment, kidney function, pregnancy plans, and side effects.

These treatments can increase the risk of infection and require regular blood and urine tests. Cyclophosphamide can affect fertility and increase certain long-term risks. Glucocorticoids may affect bones, blood pressure, blood sugar, mood, sleep, and infection risk. Your care team can discuss monitoring, vaccination, fertility preservation, and ways to reduce these risks.

Plasma exchange, which filters antibodies from the blood, is not routine treatment for everyone with GPA. Specialists may consider it in selected situations, such as certain rapidly worsening kidney problems or severe lung bleeding.

Complications

GPA can cause lasting damage if inflammation is severe, prolonged, or repeatedly returns.

Possible complications include chronic kidney impairment, kidney failure, lung scarring, or bleeding into the lungs. Inflammation in the upper airway can narrow the windpipe, while long-term nasal damage may change the shape of the nose. Hearing loss may become permanent.

Eye inflammation can threaten vision. Nerve damage may leave ongoing numbness, pain, or weakness. GPA is also associated with an increased risk of blood clots.

Some complications come from treatment rather than the condition itself. These include serious infections, weakened bones, fertility problems, and medicine-specific effects. Regular monitoring helps identify problems early.

Prevention

There is no known way to prevent GPA because its exact cause is unknown. It is not caused by poor hygiene, diet, stress, or lack of exercise.

Once GPA has been diagnosed, taking maintenance treatment as agreed and attending monitoring appointments can reduce the chance that a relapse or treatment complication goes unnoticed. Avoiding tobacco smoke can also help protect lungs that are already vulnerable, although it does not prevent GPA itself.

Living with it and what to expect

Many people reach remission and return to work, education, exercise, and family activities. Improvement may take time, especially if GPA caused kidney, lung, nerve, hearing, or airway damage before treatment.

Remission does not always mean the condition is permanently gone. GPA can relapse months or years later, and some people have more than one relapse. PR3-ANCA-associated GPA tends to relapse more often than some other forms of ANCA-associated vasculitis, but an ANCA result alone cannot reliably predict what will happen to one person.

Follow-up commonly includes symptom reviews, blood pressure checks, urine testing, kidney tests, and blood counts. Imaging or other tests may be repeated when symptoms or earlier findings make this useful.

Fatigue can continue even when inflammation is controlled. Recovery may include gradually rebuilding strength and adjusting to permanent changes such as hearing loss or reduced kidney function. Emotional support, rehabilitation, hearing services, and kidney or respiratory care may all be useful.

Do not stop immune-suppressing medicines suddenly without medical advice. This is especially important for glucocorticoids, which often need to be reduced gradually.

When to get medical help

Contact your usual healthcare team promptly if you have possible signs of a relapse, such as returning sinus symptoms, nosebleeds, unexplained fever, joint pain, a new rash, worsening cough, blood in the urine, swelling, or new numbness or weakness.

Also seek prompt advice for possible treatment complications, including a persistent fever, painful urination, unusual bruising, or exposure to a significant infection. Because

immune-suppressing treatment can make infections harder to recognize, symptoms should not be ignored.

Seek emergency medical help for:

These symptoms can have causes other than GPA, but they need urgent assessment.

- Severe or rapidly worsening difficulty breathing
- Coughing up more than small streaks of blood
- Chest pain, fainting, confusion, or blue lips
- Sudden loss of vision or severe eye pain
- Very little urine with swelling or breathlessness
- Sudden one-sided weakness, facial drooping, or trouble speaking

Common questions

Is granulomatosis with polyangiitis cancer?

No. GPA is an autoimmune blood vessel condition, not cancer. Some symptoms and scan findings can resemble cancer, so testing may be needed to tell them apart.

Is GPA contagious?

No. You cannot catch GPA from another person or pass it to family members through contact, coughing, food, or sex.

Can GPA be cured?

There is no treatment that guarantees GPA will never return. Treatment can often achieve remission, sometimes for many years. Ongoing monitoring remains important because relapse is possible.

Does a positive ANCA test mean I have GPA?

Not by itself. ANCA can support a diagnosis when symptoms and other test results fit, but it may occur in other conditions. Some people with GPA have negative ANCA tests. A clinician interprets the result alongside urine tests, imaging, examination findings, and sometimes a biopsy.

What is the life expectancy with GPA?

Outcomes have improved greatly with immune-suppressing treatment. Many people live for years or decades after diagnosis. An individual's outlook is shaped by which organs are affected, how quickly treatment controls inflammation, permanent organ damage, relapses,

infections, and treatment side effects.

Can GPA come back after remission?

Yes. A return of active inflammation is called a relapse. It may affect the same organs as before or cause different symptoms. Routine follow-up helps distinguish relapse from infection, medicine side effects, and damage left by earlier inflammation.

Sources

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