



# PATIENT FACT SHEET

## Granulomatosis with Polyangiitis (Wegener's)



### CONDITION DESCRIPTION

**Granulomatosis with polyangiitis (GPA), formerly called Wegener's granulomatosis, is a rare blood vessel disease.** It's a type of vasculitis. GPA can be very serious, but there are effective treatments available.

GPA involves inflammation of small- and medium-sized blood vessels. Blood cannot flow properly and deliver oxygen to cells around the body. Granuloma or cellular

inflammation occurs, causing organ damage. Recurring disease is common.

GPA often affects the sinuses, lungs and kidneys, but may also affect eyes, ears, skin, nerves, joints and other organs. Only 3 out of every 100,000 people get GPA, and it affects women and men equally. It occurs at any age, but is very rare in children. It usually develops between the ages of 40 and 65.



### SIGNS/ SYMPTOMS

**GPA symptoms may develop over days or months. Early signs are nasal congestion, frequent nosebleeds, shortness of breath and coughing up bloody phlegm.** Other possible signs are joint pain, decreased hearing, skin rashes, eye redness or vision changes, fatigue, fever, loss of appetite and weight, night sweats, and numbness or loss of movement in fingers, toes or limbs.

A rheumatologist can diagnose GPA with a physical examination, lab tests, imaging scans and biopsy.

A positive blood test for anti-neutrophil cytoplasmic antibodies (ANCA) suggests but doesn't confirm GPA. Up to 20% of people with GPA test negative for ANCA.

If GPA is suspected, urinalysis and blood testing of creatinine levels can check kidney function, and a chest x-ray or CT scan of the lungs may show signs of granuloma. Tissue biopsy confirms GPA diagnosis.



### COMMON TREATMENTS

**GPA can lead to kidney or lung failure without prompt treatment. Treatment choice depends on the organs involved, disease severity and a person's overall health.** Patients with active, severe disease are treated with a high-dose corticosteroid such as prednisone [Deltasone, Orasone] and cyclophosphamide [Cytoxan], a type of chemotherapy. The prednisone dose is gradually tapered. Then, patients may take either methotrexate

[Rheumatrex, Trexall, Orastrup, Rasuvo] or azathioprine [Imuran, Azasan] for two or more years.

Another option for severe GPA is rituximab [Rituxan] and prednisone. Rituximab is a biologic that is injected. Less active disease may be treated with methotrexate and prednisone. The rheumatologist and patient can talk about the best treatment option, as these medications may have serious side effects.



### CARE/ MANAGEMENT TIPS

**People with GPA need regular check-ups, as disease relapses are common.** It can also have serious complications that are even life-threatening if not treated promptly. Patients need regular laboratory tests, imaging scans and clinical visits with a rheumatologist.

Relapses of GPA may involve different symptoms than earlier episodes, so patients should report any new symptoms to their doctor right away. Regular doctor's visits and testing should spot any problems early so prompt, effective treatment can be started. A rheumatologist can also monitor and manage any treatment side effects.

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