

## Wegener's Granulomatosis

Wegener's granulomatosis (WG) is a rare blood vessel disease that can cause symptoms in the nasal sinuses, lungs and kidneys as well as other organs. This is a complex and potentially serious disease. However, with prompt diagnosis, WG can be treated effectively.

### Fast Facts

- Wegener's granulomatosis often affects the sinuses, lungs and kidneys, and can lead to kidney failure.
- To prevent complications, prompt diagnosis and treatment are essential.
- Recurrences of this form of [vasculitis](#) are common.

### What is Wegener's Granulomatosis?

WG belongs to a group of diseases characterized by the presence of blood vessel inflammation (vasculitis). In WG, organ damage occurs as a result of [vasculitis](#) involving the small- and medium-sized blood vessels, and from a type of tissue injury called granulomatous inflammation. [A granuloma is a mass or nodule of inflamed tissue, usually seen on biopsies of affected organs.]

Most commonly, WG affects the nasal sinuses, lungs and kidneys, but can also affect the eyes, ears, skin, nerves, joints and other organs. Because of the variety of potential organ involvement, a wide range of symptoms can develop over days to months. For 90% of people, the first symptoms appear in the respiratory tract (e.g., nose and lungs) and include nasal congestion, frequent nosebleeds, shortness of breath, and cough that may produce bloody phlegm. Other early symptoms can include joint pain, decreased hearing, skin rashes, eye redness and/or vision changes, fatigue, fever, appetite and weight loss, night sweats, and numbness or loss of movement in the fingers, toes or limbs.



Well-studied therapies have proven effective in treating WG. Although permanent organ damage can still occur, the medications used to treat WG can improve or even resolve organ injury in many instances.

### **What causes Wegener's Granulomatosis?**

The cause of WG is unknown.

### **Who gets Wegener's Granulomatosis?**

This rare disease is estimated to affect 3 out of every 100,000 people, and men and women equally. The condition can occur at any age, but most often between the ages of 40 and 50.

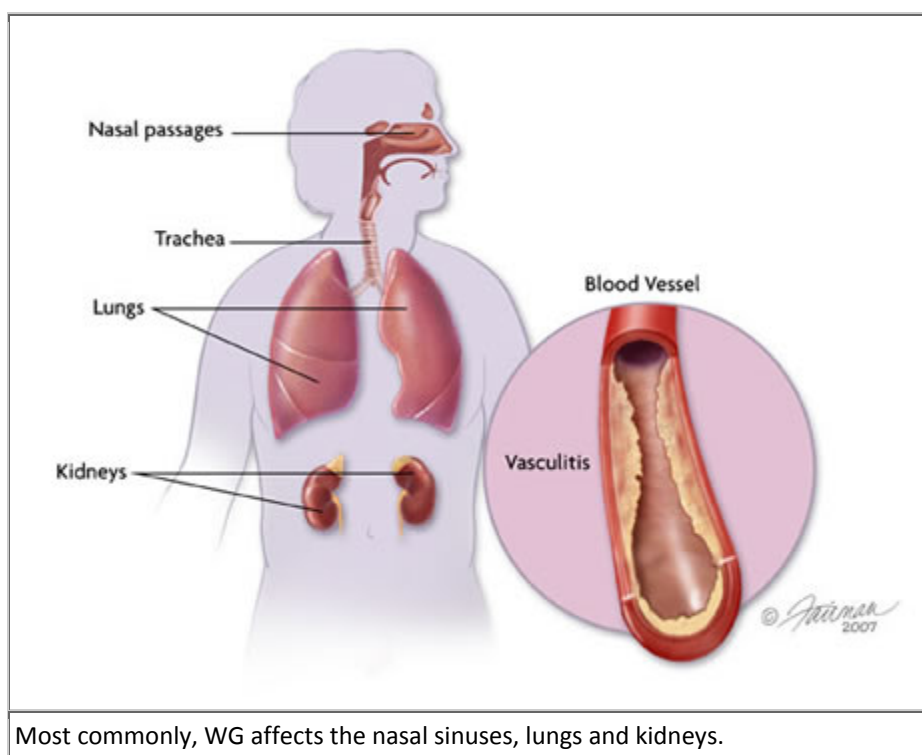
### **How is Wegener's Granulomatosis diagnosed?**

History of symptoms, physical

examination, and laboratory testing and imaging studies, such as x-rays, computed tomography (CT), or magnetic resonance imaging (MRI), each play an important role in detecting organ involvement in WG. As symptoms may not develop in the kidneys and the lungs, two of the most important sites of disease, lab tests are used to look for problems in these organs. For example, the doctor may order a urinalysis, measurement of renal function (drawing blood to test levels of creatinine) and lung imaging through a chest x-ray or CT if WG is suspected.

The diagnosis of WG is based on clinical features affecting typical organ sites together with the identification of blood vessel and tissue inflammation in those organs. Because of this, most physicians will recommend a biopsy of the tissues involved to look under the microscope for evidence of characteristic changes.

Many people with WG test positive for special antibodies in their blood called antineutrophil cytoplasmic antibodies (ANCA). A positive ANCA blood test supports the need for further examination for WG but usually does not replace the need for a tissue biopsy. In rare instances where people have certain clinical features, a positive ANCA may be sufficient to make a diagnosis of WG. However, ANCA cannot be found in up to 20% of people with well-diagnosed WG so a negative ANCA test does not throw out the possibility of WG in patients with other specific clinical features.





### How is Wegener's Granulomatosis treated?

Active WG involving a critical organ site requires treatment to prevent ongoing inflammation that can result in organ failure. In the absence of treatment, WG can worsen rapidly, leading to potentially life-threatening kidney or lung failure. Treatment choices are based on the organs affected, disease severity and individual medical factors. Typically those with active severe disease are given a glucocorticoid (steroid) medication such as prednisone combined with daily oral [cyclophosphamide](#), a chemotherapy type of medication. Prednisone is begun at a high dose and gradually reduced. [Cyclophosphamide](#) is given for 3-6 months. Those who improve are then switched to another medication such as [methotrexate](#) or [azathioprine](#) for 2 or more years. Those with active but not severe disease may be treated with prednisone and [methotrexate](#). Each medication has significant side effects that must be considered to minimize or prevent the risk of serious side effects.

### Living with Wegener's Granulomatosis

The impact of WG varies greatly between individuals. It's influenced by the severity of their illness, the organs involved, and any complications related to the disease or its treatment. People with WG need ongoing checkups that include laboratory studies, imaging tests, and clinic visits with their doctor. Such monitoring is essential to follow the course of WG and to detect or prevent short- or long-term treatment-related complications.

Even with effective treatment, recurrences (relapses) are common. Relapses may resemble or differ from the initial onset so new symptoms should be reported to the medical practitioner as soon as possible. Regular doctor visits together with monitoring of laboratory tests and imaging studies can help to detect relapses early.

### Points to remember

- WG is serious but treatable.
- Significant side effects of medications used to treat WG can be minimized with preventive strategies.
- While disease relapses frequently occur, early detection and treatment can lessen the potential for serious organ injury.

### The rheumatologist's role in treating Wegener's Granulomatosis

Depending on the organ sites involved, people with WG frequently require care from numerous physician specialists, each of whom contributes in a valuable way. Within this working team, however, it remains important to identify a specific doctor to prescribe and check for problems with related medications. This is typically the rheumatologist who is trained to use these medications to effectively treat WG while minimizing the potential for side effects.



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### To find a rheumatologist

For more information about rheumatologists, [click here](#).

For a listing of rheumatologists in your area, [click here](#).

### For more information

The American College of Rheumatology has compiled this list to give you a starting point for your own additional research. The ACR does not endorse or maintain these Web sites, and is not responsible for any information or claims provided on them. It is always best to talk with your rheumatologist for more information and before making any decisions about your care.

Center for Vasculitis Care and Research, Cleveland Clinic

<http://www.clevelandclinic.org/arthritis/vasculitis>

John's Hopkins Vasculitis Center

<http://vasculitis.med.jhu.edu/>

Vasculitis Clinical Research Consortium

<http://rarediseasesnetwork.epi.usf.edu/vcrc/>

Vasculitis Foundation

<http://www.vasculitisfoundation.org/>

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Written by Carol A. Langford, MD MHS, and reviewed by the American College of Rheumatology Patient Education Task Force.

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