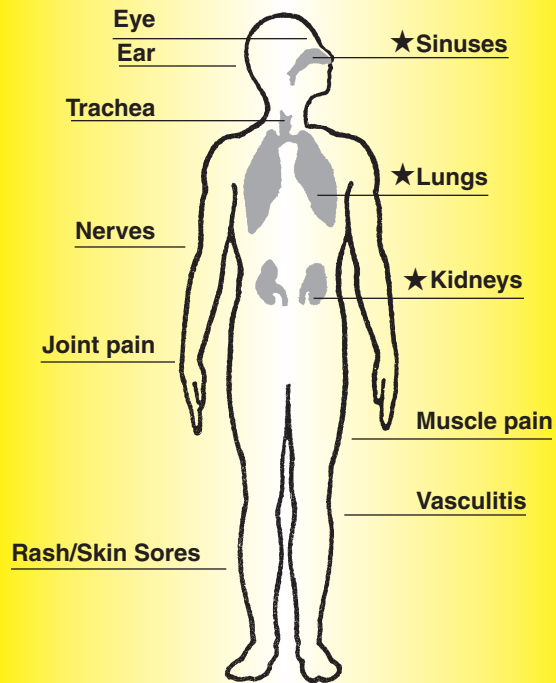


Wegener's Granulomatosis



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Founded May, 1986
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The Disease

Wegener's Granulomatosis (WG) is an uncommon disease that affects about 1 in 20,000 to 1 in 30,000 people. Symptoms are due to **inflammation** that can affect many tissues in the body, including blood vessels (**vasculitis**). It is also considered a disease of abnormal immune function. Some consider it an autoimmune disease, meaning the body's immune system attacks its own body tissues.

There is no known cause of Wegener's; but it is **not contagious**, and there is **no evidence it is hereditary**. It is systemic, meaning it affects the body as a whole. It affects the upper (sinuses and nose), and lower (lungs), respiratory system and frequently involves the kidneys, lungs, eyes, ears, throat, skin and other body organs. For reasons not clear, blood vessels in those areas may become inflamed and clusters of certain cells (**granulomas**), may occur. Patients who do **not** have renal (kidney), involvement are said to have **Limited Wegener's**. However the term "limited" has become unpopular because certain forms of the illness may still be very serious even though the kidneys are not involved.

As awareness of Wegener's Granulomatosis grows, more patients are diagnosed in the early stages of the disease when effective treatment can result in early remission and prevent organ failure.

History of Wegener's

In 1931, a medical student, Heinz Klinger, working with Professor Roessle at the University of Berlin, reported postmortem accounts of two patients who were found to have **inflammation, with granulomas**, affecting many tissues, including the airways and blood vessels.

Five years later, Friedrich Wegener, at the University in Breslau, described the same syndrome in three patients. These patients were also found to have **granulomas and vasculitis** of the upper and lower respiratory tract, as well as other tissues.

In 1954, seven more patients were described. This resulted in the establishment of definite criteria for the diagnosis of the disease described by Friedrich Wegener and hence the name Wegener's Granulomatosis.

Who Gets WG?

Wegener's Granulomatosis is an uncommon disease, which can occur at any age. It most often occurs in the 4th and 5th decade of life. Patients are divided equally between males and females. It appears that Caucasians are far more commonly affected than other racial groups.



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Signs and Symptoms

Onset of Wegener's Granulomatosis may be indolent, slow moving with few symptoms, or have a rapid and severe onset.

About **90% of patients** have symptoms of a 'cold', 'runny nose' or sinusitis **that fail to respond** to the usual therapeutic measures **and last considerably longer** than the usual upper respiratory tract infection. Other symptoms include nasal membrane ulcerations and crusting, saddle-nose deformity, inflammation of the ear with hearing problems, inflammation of the eye with sight problems, cough (with or without the presence of blood), pleuritis, (inflammation of the lining of the lung), rash and/or skin sores, fever, lack of energy, weakness, fatigue, loss of appetite, weight loss, arthritic joint pain, night sweats, and blood in urine which may or may not be indicated by a change in urine color.

Be aware that **not all** Wegener's patients experience all symptoms. Different patients experience different symptoms, and the severity of the disease is also different for each Wegener's patient. **If any of the above symptoms persist**, consider a possible diagnosis of Wegener's Granulomatosis and arrange to have a complete evaluation, including health history, physical exam, laboratory studies, including a urinalysis and an ANCA test.

Diagnosis

Diagnosis is established by clinical and laboratory findings such as the ANCA blood test, other blood and urine tests, x-rays, and tissue biopsy, if needed.

Antineutrophil Cytoplasmic Antibody (ANCA) is an abnormal protein. ANCA is part of a large family of molecules called immunoglobulins, (including antibodies), that are made by all animals and are normally intended to protect you. There are two types of ANCA: 'c' (cytoplasmic), and 'p' (perinuclear). **The vast majority of WG patients test positive for c-ANCA** while a small percentage of patients test positive for both 'p' and 'c' ANCA. **C-ANCA** reacts with a normal human enzyme contained in white blood cells (called proteinase 3 or PR3), and when seen with a special microscope has a yellow-green color outside of the nucleus and throughout the fluid in the cell called the **Cytoplasm**, hence the term **c-ANCA**. **The quantity of c-ANCA roughly correlates with disease activity.** Very rarely, c-ANCA can be found in other diseases and even in some normal individuals.

The c-ANCA/PR3 antibody test is a helpful diagnostic tool that is used most effectively when patients are thought to **possibly** have WG. A positive test is supportive of the diagnosis. However, **a negative test does not guarantee that WG is not present.** Some physicians use the ANCA test to monitor treatment and disease status, while others caution that test results may be misleading in up to 1/3 of patients and therefore should not be the primary guide to decision making. Decisions to change treatment should be based on convincing features of illness activity or remission, and complete laboratory tests.

Treatment

Treatment of Wegener's Granulomatosis usually consists of cytotoxic agents (a form of chemotherapy), often using relatively low doses of **Cyclophosphamide (Cytoxan)**, and/or **Methotrexate** and/or **Azathioprine (Imuran)**, and glucocorticoids (**Prednisone**).

Although basic treatment is the same, it will vary based on the individual patient symptoms, disease activity, organ involvement and lab test results. **Patients with kidney involvement and or more severe disease are commonly prescribed Cyclophosphamide and Prednisone as initial treatment.** Those with **milder forms of Wegener's are commonly prescribed Methotrexate and Prednisone.**

Treatment...continued

These medications will be reduced over time, and even eliminated, if the patient remains in a stable remission. Wegener's patients may also be prescribed calcium supplements or other similar medication to prevent osteoporosis from extended prednisone use. Many patients will also be prescribed the antibiotic **Bactrim** to help prevent secondary lung infections with a dangerous 'bug' called **Pneumocystis carinii pneumonia (PCP)**. In addition, there is some evidence that Bactrim, used cautiously, can have the beneficial effects of reducing relapses and upper airway infections.

Effective treatment should also include a **'team approach' with medical specialists** according to the patient's organ involvement. It is common for a Wegener's patient to regularly see the following Doctors: Nephrologist (Kidney), Otolaryngologist (Ear, Nose/Sinus, Throat), Ophthalmologist (Eye), Pulmonologist (Lung), and **always a Rheumatologist/Immunologist**. Other specialists are involved as needed.

Wegener's patients must maintain a good relationship with their doctors and understand and follow instructions carefully. Many patients find it very useful to maintain a diary listing medications, test results and notes on how they are feeling and what, if any, symptoms they are experiencing. This material can then be reviewed as necessary during a patient/doctor appointment.

It is imperative to have a close, continuous and long-term doctor follow-up, even when in remission and off therapy as relapses or flare-ups are common.

Remission

There is no cure for Wegener's Granulomatosis, but early diagnosis and proper treatment will be effective and the disease can be brought into remission with complete absence of all signs of disease.

Long-term remission can be induced and maintained with medications, close management and regular lab tests to help monitor the disease. Treatment can produce symptom-free intervals of 5 to 20 years or more. Some patients will achieve a drug-free remission. However, **relapses are common** but can be caught at their earliest and most treatable stage, for most patients, by paying attention to patient symptoms and lab tests. **WG patients in remission must not hesitate to see a doctor if any WG symptoms return or if they are not feeling well.**

Reply/Donation Form

Yes, I want to become a WGA Member!

Name (Ms./Mrs./Miss/Mr.) _____

Address _____ Apt. _____

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Individual or Family: \$20. per year for USA;

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Yes, I want to further support the efforts of WGA!

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Please send an acknowledgement of my gift to: 

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Designate Support for:

Awareness/Education Research

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I would like to speak with a WGA representative to learn more about how planned giving opportunities can benefit my tax, financial and estate planning process.

WGA accepts: Checks, Credit Cards, Cash, Money Orders, Postal Money Orders and Western Union Money Orders

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Tax receipts will be issued for all donations.

Thank you for your generosity!

Return to: WGA, P.O. Box 28660, Kansas City,
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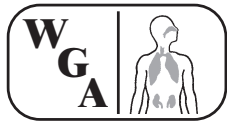
WGA is a registered 501(C)(3) Organization

The Future

We believe Wegener's Granulomatosis is not so rare, but rarely diagnosed. In areas where doctors are aware of the disease, more patients are diagnosed and treated. Early diagnosis and treatment are essential to improve patient outcome and prevent organ failure.

WG treatment has come a long way. In 1958, WG patients had an 82% mortality rate at one year, with an average patient survival of 5 months. With the advent of Cyclophosphamide treatment during the 1970's survival rates improved dramatically. However, treatment side effects and drug toxicity presented new challenges. Today, drug toxicity is managed more carefully and long-term remissions are possible. Some patients are able to lead relatively normal lives and have been in remission for 20+ years after treatment!

Research into new medications, treatment options and the cause of WG are being investigated at leading medical centers throughout the world. Of course, more needs to be done and with your help quicker WG diagnosis and even better, and less toxic, treatments will become a reality.



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Info Packets Available

Patient information packets are available for the benefit of patients, families and care givers.

Doctor's information packets of medical information and papers are also available on request by a medical professional.

Both information packets can be obtained by contacting the Wegener's Granulomatosis Association in Kansas City, Missouri, USA.

Consultants

To be contacted by physicians only *

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Consultants

* Our medical consultants are recognized authorities throughout the world on the diagnosis and treatment of WG. They will consult with a patient's doctor at no cost. Due to time limits, and other factors, our **consultants cannot take patient phone calls.** If a consultation is desired, please ask your doctor to contact one of the medical consultants on your behalf. Only in the case where a patient's doctor refuses to contact a consultant would it be acceptable for a patient to call the consultant's office and ask a staff member how to proceed.

About WGA

Wegener's Granulomatosis Association, formerly the Wegener's Granulomatosis Support Group International Inc. was founded in May, 1986 by Marilyn Sampson (1933-1997), a Wegener's patient and registered nurse.

WGA was established to alleviate the isolation of having a rare life-threatening disease for patients and their families. We understand what each patient is going through, and we are here to help. We want to give each patient hope, a strong positive outlook and the strength to fight and not give up. We want patients to know and believe they can survive. And, we want to be there to celebrate their recovery.

WGA Mission Statement

Wegener's Granulomatosis Association offers comfort and support to WG patients and families through education, awareness and research.

Our mission consists of the following objectives:

- Establish rapport with all known WG patients, to alleviate the isolation of having an uncommon life-threatening disease.
- Assist WG patients and their families with clinical information and coping strategies, to help them gain a strong positive outlook.
- Create greater awareness about WGA within the medical community, as well as the general public.
- Support research efforts into the cause, treatment and cure for WG.