

Wegener's granulomatosis

Wegener's granulomatosis is a rare type of inflammation that targets the arteries, veins and capillaries of vital organs within the body. The two organs that it mainly targets are the kidneys and the respiratory system, including the lungs, trachea, nose and sinuses.

There is no cure but appropriate treatment is usually successful in controlling the inflammatory process and allows good health to be restored for many years.

Inflammation (redness, heat and swelling) of blood vessels is called 'vasculitis'. Wegener's granulomatosis is a rare type of vasculitis. If only the blood vessels of the respiratory system are affected, the disease is known as 'limited Wegener's granulomatosis'.

Wegener's granulomatosis may be fatal without prompt medical treatment. This is because the inflammation within the walls of blood vessels reduces the blood's ability to flow through the vessels and carry oxygen, which impairs the functioning of the associated organs. In severe cases, tissue death (necrosis) can occur. Men and women of any age can be affected. The disease tends to be more common in middle-aged Caucasian (white) people. The cause for this is unknown.

Symptoms

The symptoms and signs of Wegener's granulomatosis depend on which blood vessels are affected but may include:

- Fatigue
- Unexplained weight loss
- Recurrent fever
- Night sweats
- Breathlessness
- Persistent cough
- Painful joints
- Painful muscles
- Chronic runny nose
- Sinusitis (sinus inflammation, blockage and pain)
- Nasal passage ulcers
- Hole (perforation) in the tissue that separates the nostrils (septum)
- Traces of blood in nasal mucus, sputum or urine
- Chest discomfort.

Other areas of the body may be affected

While Wegener's granulomatosis tends to target the blood vessels of the kidneys and respiratory system, other areas of the body may be affected by inflammation too. Wegener's granulomatosis may cause:

- Eye problems such as conjunctivitis, scleritis or vision changes including double vision
- Nerve pain or dysfunction
- Middle ear blockage, pain and hearing loss
- Skin lesions that tend to look like purple or red blister-like lumps
- Skin ulcers (localised areas of tissue loss)
- Joint pain.

Cause

The cause for Wegener's granulomatosis is unknown. Since inflammation is usually the result of immune system activity, researchers believe the cause may be that the immune system cells begin to attack the blood vessel walls in the body. However, the cause of the immune system attack is not clear.

Diagnosis

Early diagnosis is crucial to avoid serious and potentially life-threatening complications. However, the signs and symptoms of Wegener's granulomatosis are very common to other diseases. Diagnosis relies, in part, on first testing to exclude other possible causes of the signs and symptoms.

Tests used in the diagnosis of Wegener's granulomatosis may include:

- Medical history
- Physical examination
- Urine tests that check for unusual signs such as the presence of red blood cells or proteins
- X-ray examinations of the chest, sinus cavities or both
- General blood tests to check for anaemia and inflammation
- Kidney scans
- A specific blood test to check for Wegener's granulomatosis – this test looks for unusually high levels of the immune system cell known as 'antineutrophil cytoplasmic antibody' (ANCA), which may indicate Wegener's granulomatosis
- Biopsy – this is the definitive test and involves taking a small sample of suspect tissue for laboratory examination – the positive finding of a granuloma (area of inflammatory damage) indicates Wegener's granulomatosis.

Complications

With medical treatment, about 95 per cent of patients diagnosed with Wegener's granulomatosis will survive for at least five years. Without medical treatment, however, nine out of 10 patients will die within two years. Causes of death can include:

- Heart attack
- Heart failure
- Respiratory failure
- Kidney (renal) failure.

Other complications of Wegener's granulomatosis may include:

- Blindness
- Deafness
- Haemoptysis – bleeding of the lungs, expelled in blood-stained phlegm
- Neuropathy – problems with nerves outside of the brain and spinal cord.

Treatment

Treatment for Wegener's granulomatosis aims to reduce inflammation within the blood vessels, which prevents further damage to associated organs and reduces the risk of complications. Prescription medications that inhibit the action of the immune system are commonly used.

These medications may include:

- **Corticosteroids** – steroid drugs such as cortisone are used in relatively high doses for the first few months then gradually reduced as the inflammation subsides.
- **Antibiotics** – these may be helpful in cases of limited Wegener's granulomatosis.
- **Cytotoxic drugs** – such as cyclophosphamide, which suppress the activity of the immune system and greatly extend the person's potential life span. The dose may be calculated on various factors including the patient's age and blood test results. Cytotoxic drugs may be prescribed for up to one year after remission.

- **Other drugs** – such as methotrexate, azathioprine or intravenous (given through a vein) immunoglobulin therapy to reduce the risk of flare-ups once the inflammatory process has been controlled.
- **Regular blood tests** – the average patient is advised to have an ANCA blood test every six weeks to help monitor the success of treatment.

Possible side effects of treatment

While taking medication, a patient with Wegener's granulomatosis needs careful and regular medical monitoring to maintain the best possible health. This is because the treatment may cause a wide range of unwanted side effects including:

- Nausea
- Mouth ulcers
- Hair loss
- Oedema (fluid retention)
- Insomnia
- Unwanted weight gain
- Increased susceptibility to infection
- Cystitis (bladder inflammation)
- Easily broken bones
- Diabetes
- Cognitive changes such as mood swings
- Sterility
- Cancer of the bladder.

It is important to discuss potential side effects of your medication with the doctor. See your doctor immediately if you suspect that your medications may be causing adverse reactions. The doctor may alter the dose of particular medications. Dose alterations are usually made over a period of time to reduce the risk of withdrawal symptoms such as low blood pressure and dizziness.

Long-term outlook

There is no cure for Wegener's granulomatosis but the long-term outlook, with appropriate medical treatment, is very good. In many cases, prompt treatment can bring about a remission, which means the patient has no signs or symptoms of disease.

Relapses may occur after the end of medical treatment. In most cases, relapses occur some two years after treatment is stopped. Regular check-ups are important.

Where to get help

- Your doctor
- Wegener's Granulomatosis Support Group of Australia
- Kidney Health Information Service Tel. 1800 4 KIDNEY (543 639), or TTY users phone 1800 555 677 then ask for 1800 454 363

Things to remember

- Wegener's granulomatosis is a rare type of inflammation that targets the arteries, veins and capillaries of the kidneys and the respiratory system, including the lungs, trachea, nose and sinuses.
- Wegener's granulomatosis may be fatal without prompt medical treatment.
- In many cases, prompt treatment can bring about a remission, which means the patient has no signs or symptoms of disease.

This page has been produced in consultation with, and approved by:

Kidney Health Australia

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