

Wegener's Granulomatosis is a disease that still claims too many victims. It is a disease which, according to American studies, affects 2 per million of population a year. However, a British survey carried out by Dr D M Carruthers for the Norwich Health Authority puts the figure at 8.5 per million, while other studies put the incidence of Wegener's Granulomatosis at 11 per million of population per annum.

Though often referred to as a rare disease, there is a growing belief that Wegener's Granulomatosis is an uncommon disease, not so rare but rarely diagnosed. It is becoming apparent that, in areas where doctors are aware of the disease, more patients are diagnosed and treated.

It is the purpose of Wegener's UK, through this leaflet and through any other possible means, to increase awareness, among both the general public and the medical profession, as to the life threatening nature of this disease, and the paramount importance that should be given to early diagnosis.

WG is not at all straightforward. It presents diagnostic problems. It looks like this, it appears to be that, and eventually it turns out to be something else. In fact it is very much like the logo on the front of this leaflet, seeming to be neither one thing nor the other, but a bundle of things which can turn an individual's life upside down. Like the logo, WG may be a mixture but, unlike the logo, it is certainly not imaginary.

The Background

In 1931 the first patients were reported by Heinz Klinger of the University of Berlin. They had died having prolonged sepsis with inflammation of the blood vessels throughout the body.

Five years later, Frederick Wegener in Breslau found a distinct syndrome in three patients. They had narcotizing (gradually degenerating) granulomas involving the upper and lower respiratory tract.

In 1954 seven further patients were described, and as a result definite criteria were established for the diagnosis of the disease now known as Wegener's Granulomatosis.

The Disease

Wegener's Granulomatosis (WG) is an uncommon

disease. It is a form of vasculitis - inflammation of the blood vessels, and an auto-immune disease - the body's immune system attacks its own body tissues.

There is no known cause for WG, but it is not contagious, and there is no evidence that it is hereditary.

It is systemic which means that it affects the body as a whole. This includes the upper and lower respiratory system, which are the sinuses and nose (upper) and the lungs (lower). It may also affect the kidneys, eyes, ears, throat, skin and other body organs. For reasons not yet understood the blood vessels in those areas become inflamed and cell debris, called granuloma, appears. People who do not have kidney (renal) involvement are said to have Limited WG.

Early diagnosis and treatment can result in early remission and prevent organ failure.

Who gets WG?

WG is an uncommon disease that can occur at any age, and appears to be divided equally between male and female.

Signs and Symptoms

The early onset of WG may be rapid or non-apparent, i.e. it does not exhibit early symptoms. Some patients have a steady decline in health for a year or more before WG becomes apparent; while others become seriously ill within a matter of weeks.

About 90% of patients have symptoms of a 'cold' or 'sinusitis' that fails to respond to the usual therapeutic measures, and lasts considerably longer than normal.

Other symptoms may include:

Nasal ulcerations and crusting,
Heavy nose bleeds,
Saddle nose deformity,
Inflammation of the ear with hearing problems,
Inflammation of the eye with sight problems,
Cough with/without blood and/or pus,
Pleuritis (inflammation of the lining of the lungs),
Rash,
Skin lesions

Subglottic stenosis
Shortness of breath,
Fevers/bad night sweats,
Lack of energy/lethargy/weakness,
Loss of appetite,
Weight loss,
Severe joint pain,
Changes in colour / blood in urine,
Pericarditis (inflammation of the membrane around the heart),
Peripheral neuropathy (numbness or discomfort affecting the nervous system outside the brain and spinal cord).

Please note:

Not all patients experience all these symptoms. Much depends on the speed of diagnosis, and the start of correct treatment. The sooner this takes place the greater is the possibility that the symptoms will be less severe.

Diagnosis

Diagnosis is established by clinical and laboratory findings, such as the ANCA test (a blood test) and tissue biopsy.

Antineutrophil Cytoplasmic Antibody (ANCA) is manufactured by the body's immune system. ANCA is found in the majority of WG patients and its level usually correlates with the disease activity - a positive ANCA test is more likely to occur when symptoms worsen.

The ANCA test is helpful in monitoring treatment, since the amount of ANCA in the blood tends to fall in some patients as the disease goes into remission.

Treatment

Treatment usually involves using a combination of drugs to suppress WG. The dosage depends on the severity of the disease. Also **Plasmapheresis** may be necessary, and **Dialysis** if the kidneys are badly involved.

The drugs mainly used at present are **Prednisolone**, which is an anti inflammatory, and **Cyclophosphamide**, originally used to treat cancer, but has been found to be very effective in suppressing diseases of the immune system. **Azathioprine** is also used as an alternative to

Cyclophosphamide, especially when bad side effects are experienced. **Methotrexate** is another drug which is used instead of Cyclophosphamide, and **Mycophenolate mofetil** is a more recent addition to the list of drugs found to be effective.

Even when the disease is in remission and the drugs are at a maintenance level, it is necessary to continue visiting the relevant clinic for tests as the disease can flare up again.

The side effects caused by the drugs can be devastating for many patients

Prednisolone (a corticosteroid) encourages weight gain and swelling of soft tissue, particularly in the upper body, resulting in a moon-shaped face and, in some cases, buffalo hump.

Patients may also experience thinning of bones (osteoporosis), as well as thinning of the skin, thrush in the mouth, and an increase in blood sugar levels. Mood swings may also be experienced - periods of tearfulness or low spirits followed by periods of well-being or hyperactivity.

Cyclophosphamide, though effective in bringing the disease under control, is responsible for nausea, sickness, hair loss and reduced blood cells in bone marrow.

The combination of drugs which the WG patient may require for the management of the disease may well cause problems with fertility in both male and female patients. This should be discussed with the consultant as early as possible.

NB. The ANCA blood test will monitor treatment in order to check whether the symptoms suffered by the patient are caused by WG or are side effects of the drugs.

Remission

There is no cure for WG, but early diagnosis and the correct treatment will mean that

there is more chance of the disease going into remission. Long-term remission can hopefully be achieved and maintained, but regular ANCA tests and visits to the consultant are essential ingredients in this process.

Finally

Wegener's Granulomatosis leaves patients feeling isolated, freakish, in a strange kind of way separated from those around them. Sadly the average GP and, perhaps, even the local hospital will never have heard of the illness. The patient gets shut in and only has the specialist at the clinic to talk to. It is essential for the WG patient to have support from family and friends, as his/her body goes through physical and emotional turmoil. Hence the need for publicity which is the purpose of this brochure.

If there is anything you can do to spread the word concerning this most dreadful disease it will improve the possibility of earlier diagnosis and treatment for those who will inevitably contract Wegener's Granulomatosis, and fewer will die as a consequence.

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Updated: 24th January 2000



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