
Granulomatosis with Polyangiitis Factsheet

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This was formally known as Wegener's Granulomatosis, GPA is a form of Vasculitis that affects the small to medium sized blood vessels in the body. GPA is an ANCA associated Vasculitis, meaning there are usually antineutrophil cytoplasmic antibodies (ANCA) present in the blood. It is these ANCAs which are thought to activate cells which cause inflammation and consequent damage to the blood vessel wall.

Who gets granulomatosis with polyangiitis (GPA)?

GPA is a rare illness. About 60 per million people in the UK have the condition. It is slightly more common in men than in women. It usually develops between the ages of 35-55, but has become increasingly recognised in older age groups. Less than 15 in a hundred people who get GPA are children.

What causes GPA?

One theory is that GPA is caused by infections, another is that it is caused by an allergy. No one knows for sure.

What are the symptoms of GPA?

GPA can affect different parts of the body and you can therefore get a whole variety of symptoms. You may start feeling generally unwell with fever, weakness, tiredness, loss of appetite and sweating at night. Some people get cold-like symptoms with a stuffy nose, cough and hoarse voice or blockage of the sinuses with pain in the face. You may notice crusting and ulcers around the nostrils. Deafness sometimes occurs. If your lungs become involved you may feel breathless and wheezy. Many other signs and symptoms are commonly observed, including a blistering rash and blood in the urine

How is GPA diagnosed?

Because the symptoms are so vague and varied GPA can be difficult to diagnose. It can typically take several months before the diagnosis comes to light.

The most important test is a blood test for antineutrophil cytoplasmic antibodies (ANCA). These antibodies are of 2 types - C-ANCA and P-ANCA. During an attack of the illness, people often test positive for ANCA. Blood counts, kidney function, inflammation and a urine test is also routinely undertaken.

You may be offered other tests, depending on which part of the body is affected. These may include nasal endoscopy (a telescope examination of the nose) breathing tests, chest x-rays, a scan of your sinuses and lung or kidney biopsies.

What is the treatment for GPA?

The first medicine you may be offered to control the symptoms of GPA is called cyclophosphamide. This belongs to a group of medicines called immunosuppressants. They act by lessening the activity of the immune system. Cyclophosphamide may be given in short bursts every 2-4 weeks by a drip or in daily tablet form. You will be closely monitored while taking this medication since a build up of the drug could cause side effects such as damage to blood cells and consequent infections. You will also be offered steroids such as prednisolone to help lessen your symptoms. The dosage is usually tapered off after a month although you may continue to take lower doses for a long time.

Once your symptoms have settled, your doctor may suggest changing the cyclophosphamide to another immunosuppressant medicine called azathioprine which will help to cut down future attacks. Depending on your response and your body's ability to tolerate these medications, you may be offered alternatives including rituximab, mycophenolate mofetil and methotrexate.

Does GPA get better?

Medicines such as cyclophosphamide have dramatically improved the lives of people with GPA. In about 8 out of 10 people the symptoms settle down completely, although in 5 out of 10 people they return from time to time. Infection, inflammation of the blood vessels and kidney failure are the main dangers to health, particularly in the early stages. However, early monitoring of complications and use of medicines with a low risk of side effect has made the prospects for people with GPA much better than they used to be.

Further reading

Papadopoulos P Wegener Granulomatosis , Medscape, 2012

<http://emedicine.medscape.com/article/332622-overview>