Great Ormond Street Hospital for Children NHS Foundation Trust: Information for Families

Granulomatosis with polyangiitis (GPA)
previously known as Wegener's Granulomatosis

This information sheet explains about granulomatosis with polyangiitis, what causes it and how it can be treated. It also gives details of what to expect when your child comes to Great Ormond Street Hospital (GOSH) for assessment and treatment.

What is Granulomatosis with polyangiitis?

Granulomatosis with polyangiitis (GPA) is a rare disease that affects the upper respiratory tract (sinuses, ears, nose and throat), lungs, kidneys, eyes, skin and other body organs.

As with most types of vasculitis, we do not know exactly what causes the disease to develop.

How common is it?

The exact frequency in children is not known, but it can and does occur at any age, including in children. It is slightly more common in males than females.

How is it diagnosed?

GPA is usually diagnosed by looking at your child's history of symptoms and with a physical examination. It is diagnosed by the symptoms present and a positive blood test result for a marker called ANCA (anti-neutrophil cytoplasmic antibodies).

What are the symptoms?

Ears, nose and throat	Nosebleeds are common along with crusting of blood and mucus in the nose, and sinusitis Recurrent ear infections and possible hearing loss can occur
General health	A fever or high temperature with weight loss, tummy pain, and joint pains
Chest	Chest pains and coughing, bringing up blood at times
Kidneys	Can be affected, shown by abnormal urine tests and/or high blood pressure when measured

How is it treated?

The treatment is with steroids and an immunosuppressant drug, usually cyclophosphamide or mycophenolate mofetil which dampens down the body's immune system. Your child will also be asked to take a regular long-term antibiotic to help prevent infections. You may be offered the possibility of joining in with a clinical trial. More information about these medicines is available in our general vasculitis information sheet.

What is the outlook for children with GPA?

The key to a good outcome is early diagnosis, but GPA is still a serious disease with a small risk of death in some patients. If your child receives treatment early, the overall outlook is good, however. Once he or she goes into remission (the disease gets better) and the treatment is reduced, about half of children with the disease may have a relapse (the disease returns). Most children and adolescents lead normal lives but will have to continue to see their doctor regularly to check for signs of a relapse, take their medicines regularly, and have blood tests every few months. The outlook for children whose kidneys have been affected varies from child to child, depending on the level of damage sustained.

Is there a support group?

There is no support group in the UK for Granulomatosis with polyangiitis but the following organisation provides information and support.

Vasculitis UK
Tel: 0300 365 0075
Website: www.vasculitis.org.uk

Lauren Currie Twilight Foundation
Tel: 08456 005 855
Website:
www.thelaurencurrietwilightfoundation.org

Please note: The mention of a particular support group or website does not constitute an endorsement by Great Ormond Street Hospital.

Compiled by the Vasculitis team in collaboration with the Child and Family Information Group
Great Ormond Street Hospital for Children NHS Foundation Trust, Great Ormond Street, London WC1N 3JH
www.gosh.nhs.uk

