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

Polyarteritis nodosa (PAN)

This information sheet explains about polyarteritis nodosa (PAN), 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 polyarteritis nodosa?

Polyarteritis nodosa (shortened to PAN) is a rare form of vasculitis in which the medium and/or smaller sized arteries become damaged and inflamed. It can sometimes be triggered by an infection, particularly if this infection was with a bug called streptococcus but we are unsure of the exact cause.

How is it diagnosed?

As with most types of vasculitis, it is diagnosed by the signs and symptoms that indicate inflammation and when doctors have ruled out other diseases that can have these symptoms. A blood test will be necessary and often a skin biopsy. Other biopsies may also be needed to check the extent of any damage to organs like the liver or kidneys. A specific test called an arteriogram (an x-ray test of the arteries) can show up changes in the arteries.

What are the symptoms and signs?

General health	A persistent unexplained fever or temperature with a general feeling of being unwell
Skin	Several types of skin rashes or palpable (able to be felt) lumps due to inflammation in the skin
Digestive system	Tummy aches and weight loss
Kidneys	Protein is often found in the urine which indicates that there is inflammation in the kidneys, and sometimes the blood pressure may be too high
Other organs: heart, lungs, brain, nerves and liver	Can be affected and damaged depending on which arteries are inflamed

How is it treated?

It is treated with steroids to reduce the inflammation in the body and a disease modifying drug such as cyclophosphamide, azathioprine, mycophenolate mofetil, or methotrexate. Your doctor will decide which of these is most appropriate for your child. More information about these medicines is available in our general vasculitis information sheet.

What is the outlook for people with polyarteritis nodosa?

There is a range of possible outcomes depending on the severity of the disease and how much damage is already there in vital organs such as the kidneys. Early diagnosis and prompt treatment improves the outcome, but despite this PAN can be a serious disease with a small risk of death, particularly if diagnosis is delayed. Many children will respond well to the medicines and not have further relapses. Others will have a more chronic, relapsing course and require long-term maintenance on medications. Overall, most children require treatment for two to three years, after which treatment can be withdrawn usually without return of the disease. Your doctor may invite your child to join a long-term follow up study looking for early hardening of the arteries.

Is there a support group?

There is no support group in the UK for polyarteritis nodosa but the following organisations provides information and support.

Vasculitis UK
Website: www.vasculitis.org.uk

Lauren Currie Twilight Foundation
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

