



Vasculitis

This information sheet explains about vasculitis and what to expect when your child comes to Great Ormond Street Hospital (GOSH) for treatment.

What is vasculitis?

Vasculitis is a word used to describe various diseases causing inflammation of the blood vessels.

Blood is carried around the body in tubes or vessels; these are different in size and have different names. There are arteries (which carry blood away from the heart), veins (which carry blood to the heart) and capillaries (tiny blood vessels) through which the blood travels to all tissues and organs.

When a small blood vessel becomes inflamed, it can break and bleed into the surrounding tissue, causing small red or purple dots on the skin. If a larger blood vessel becomes inflamed, it may swell to produce a lump that you can feel under the skin. The inside of the blood vessel may also narrow, which reduces the amount of blood able to flow through it or it may become blocked by a blood clot. If the blood flow through the blood vessels is reduced or stops, the tissue may begin to die. On rare occasions, vasculitis may cause the wall of a blood vessel to weaken and develop a bulge (aneurysm) that can rupture and bleed.

There are many types of vasculitis and they are usually defined by the size of the blood vessels which are involved.

- Small vessel – usually involves the skin such as Henoch-Schönlein Purpura
- Small artery – includes Granulomatosis with polyangiitis (GPA), Churg-Strauss syndrome
- Medium artery – includes Polyarteritis Nodosa , Kawasaki disease
- Large artery – includes Takayasu's arteritis

Some types of vasculitis affect mainly the skin and others can affect internal organs with more serious complications. There are other forms of vasculitis where it is not clear into which category they fit, but these are very rare in children and will be recognised by specialised children's doctors. Some types of vasculitis which mainly affect older patients are not included in this booklet.



What are the symptoms of vasculitis?

The symptoms of vasculitis depend on which organs are affected. In most cases, children have other symptoms of general illness, including fatigue, fever, weight loss and aches and pains all over. The symptoms associated with some types of vasculitis in the table below.

Skin The most common symptom	Red or purple pinprick spots called 'petechiae' or if they are larger (about the size of your fingertip) they are called 'purpura', but others can occur including itching, hives (nettle rash) or wheals on the skin, and painful lumps
Kidneys	Vasculitis can lead to kidney damage which shows up as high blood pressure or blood and protein in the urine
Eyes	Blood vessels in the back of the eye (retina) can be affected, which can lead to blurring or reduction in sight
Major organs of the body	These can be affected by a reduction in blood flow to them causing pain and sometimes damage to the organ

How is vasculitis diagnosed?

Vasculitis can be difficult to diagnose and may have many symptoms that can look very similar to those of other diseases. This may mean it takes a while to achieve a clear diagnosis. As the disease can affect various parts of the body, a multidisciplinary approach is often needed with input from several different doctors in GOSH.

In some cases, the doctors can only decide on a diagnosis once other conditions have been ruled out and after several specific tests or investigations have been performed such as those in the table below.

Blood tests	These can show if the blood vessels are inflamed. They can be an indicator of inflammation in the body and to help monitor other organ function/damage
Urine tests	To look for any kidney disease
X-rays and scans	To confirm any organ or bone problems
Biopsies	Of the skin/muscle or kidneys to confirm their involvement
Angiogram	Dye is injected into the arteries to show them up around major organs, such as the heart and look for changes

What causes vasculitis?

We can not say for certain what causes vasculitis to develop. The most likely reason is that the white blood cells attack healthy cells instead of foreign invaders like bacteria and viruses; we are unsure why this happens. It may also be related to other autoimmune disorders like arthritis and systemic lupus erythematosus (SLE).



How is vasculitis treated?

Many types, especially Henoch-Schönlein Purpura, do not need treatment. The more severe and uncommon cases of vasculitis will require quite strong treatment with different medicines; a quick guide is given below.

Medication	How is it given	Side effects
Steroids Reduce the inflammation and dampens down the immune system	High doses of methylprednisolone given via a drip in 'pulses' or blocks of treatment and/or as prednisolone tablets given daily	Increase in appetite leading to weight gain Mood changes Thinning of the bones in long term use Can become more prone to infections Reduced growth
Methotrexate Disease modifying anti-rheumatic drug that reduces symptoms and inflammation	Orally by tablet/syrup or by subcutaneous injection given weekly	Stomach upset and feeling sick (nausea) Mouth ulcers and thinning of the hair, which can be treated with folic acid Changes in blood counts, therefore regular blood tests are required especially for monitoring changes in liver function tests
Cyclophosphamide Dampens down the immune system	By drip in hospital from fortnightly to three-monthly	Feeling sick (nausea), mouth ulcers Can become more prone to infections Irritation of the bladder wall Changes in blood counts, requires regular testing and monitoring
Mycophenolate Mofetil (MMF) Dampens down the immune system	Orally twice a day on an empty stomach Doses will be increased slowly	Upset stomach Changes in blood counts requires regular testing and monitoring Can become more prone to infections
Azathioprine Used to maintain improvement	Orally once daily	Upset stomach Can become more prone to infections Changes in blood counts requires regular testing and monitoring

There are newer, even more powerful medicines under trial, which can be fully explained to you if appropriate.



More information and support groups

General information about vasculitis

The following organisations provide 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

Arthritis Care

Helpline: 0808 808 4050

Website: www.arthritiscare.org.uk

Arthritis Research Campaign

Tel: 0870 850 5000

Website: www.arc.org.uk

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