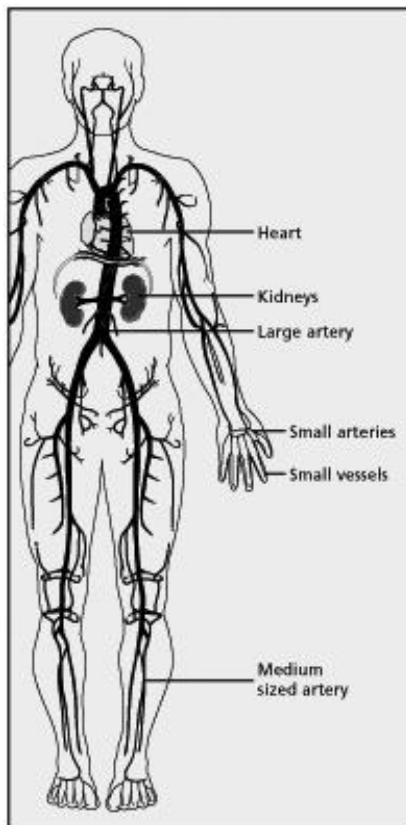


Vasculitis: information for families

Vasculitis is a word used to describe various diseases causing inflammation of the blood vessels. This information sheet from Great Ormond Street Hospital (GOSH) explains about vasculitis, what causes it and how it can be treated.

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.

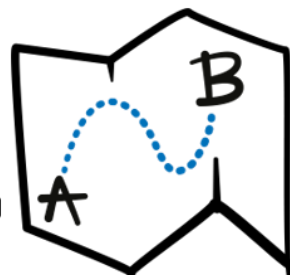
The inside of the blood vessel may also narrow, which reduces the amount of blood able to flow through it or it may become blocked by a blood clot. If the blood flow through the blood vessels is reduced or stops, the tissue may begin to die.

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 described in this information sheet.

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 are described below.

Skin (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

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 below.

- **Blood tests** to 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** (small samples of tissue) from the skin/muscle or kidneys to confirm their involvement
- **Angiogram** to show the arteries up around major organs, such as the heart and look for changes, by injecting a dye into them

What causes vasculitis?

We cannot 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. Details are given in the table on the next page.



Medication	How it is given	Side effects
Corticosteroids to reduce the inflammation and dampen 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	<ul style="list-style-type: none"> • 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 is a disease modifying anti-rheumatic drug that reduces symptoms and inflammation	Orally by tablet/syrup or by subcutaneous (under the skin) injection given weekly	<ul style="list-style-type: none"> • 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	<ul style="list-style-type: none"> • 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	<ul style="list-style-type: none"> • Upset stomach • Changes in blood counts requires regular testing and monitoring • Can become more prone to infections
Azathioprine is used to maintain improvement	Orally once daily	<ul style="list-style-type: none"> • 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.

What is the outlook for people with vasculitis?

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. 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.

Further information and support

- Vasculitis UK – Call their helpline on 0300 365 0075 or visit their website at www.vasculitis.org.uk
- Lauren Currie Twilight Foundation – Call their helpline on 08456 005 855 or visit their website at www.thelaurencurrietwilightfoundation.org
- Arthritis Care – Call their helpline on 0808 808 4050 or visit their website at www.arthritiscare.org.uk
- Arthritis Research Campaign – Call their helpline on 0870 850 5000 or visit their website at www.arc.org.uk

