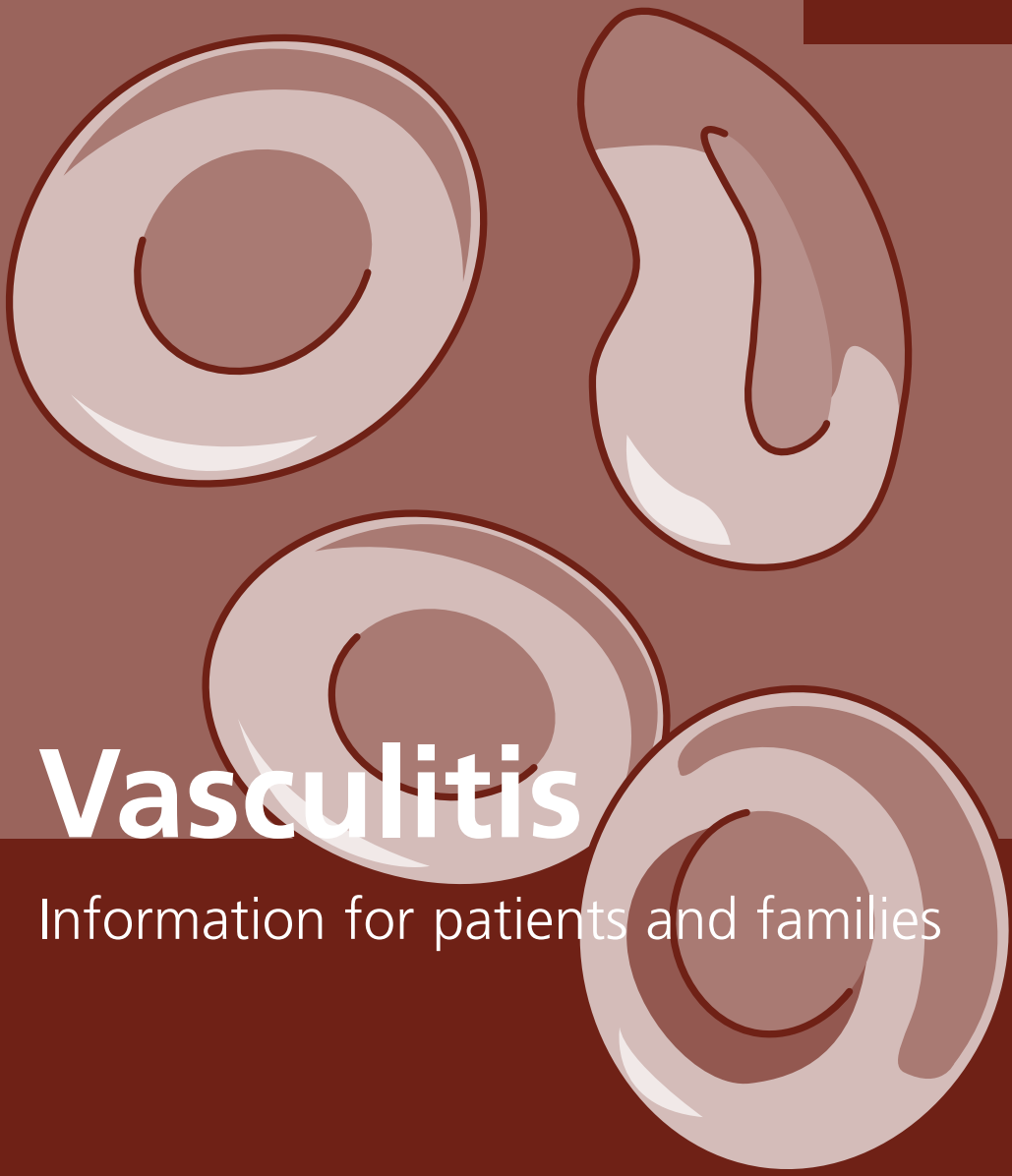
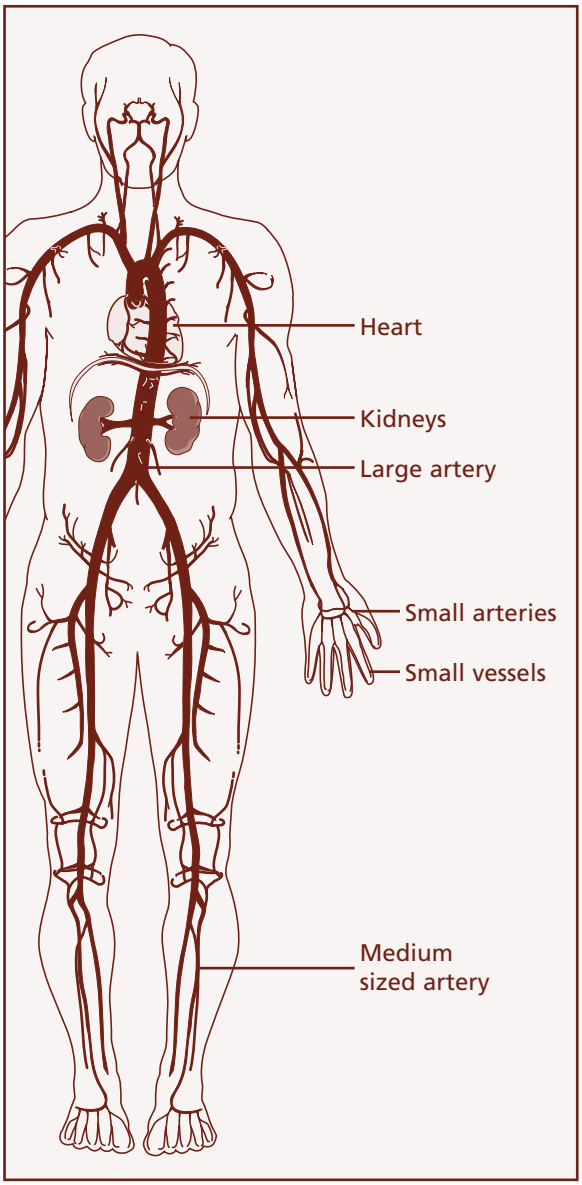


Vasculitis

Information for patients and families

Great Ormond Street Hospital for Children NHS Trust

This booklet explains about the various forms of vasculitis, and what to expect when your child comes to Great Ormond Street Hospital for treatment.



What is vasculitis?

Vasculitis is a word used to describe various diseases that involve inflammation of the blood vessels. The blood vessel system is made up of various sizes of blood vessels, arteries (which carry blood away from the heart), veins (which carry blood back 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. This causes small red or purple dots on the skin. If a larger blood vessel becomes inflamed, it may swell to produce a lump that you 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, affecting different sizes of blood vessel and different parts of the body. The different kinds of vasculitis are defined in different ways. For example, from the size of the affected blood vessels or the way they show up most often.

Some types of vasculitis affect predominantly the skin and others can affect internal organs with more serious complications. Those mainly affecting the skin include conditions such as: urticarial vasculitis; leucocytoclastic vasculitis; panniculitis which involves the fat tissue deep to the skin and pityriasis lichenoides. These are relatively rare disorders and can be associated with some systemic (affecting the whole body) symptoms. Children with these skin conditions are usually referred to a skin specialist (dermatologist). Often a skin biopsy is needed to help confirm the diagnosis. Treatment varies according to the diagnosis and any associated medical problem, but for those with mainly skin vasculitis the outlook is usually good.

The booklet describes different types of systemic vasculitis.

The most common types of vasculitis affecting children:

- Henoch-Schönlein Purpura
- Kawasaki disease

Other types of vasculitis:

- ANCA associated vasculitis, which includes microscopic polyangiitis, Wegener's granulomatosis and renal limited vasculitis
- Polyarteritis Nodosa
- Takayasu's arteritis.

There are other types 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. Other types of vasculitis 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 include:

- skin - red or purple pinprick spots called 'petechiae'. If the spots are larger (about the size of your fingertip) they are called 'purpura'. These are the most common skin symptoms associated with vasculitis, but others occur including itching, hives (nettle rash) or wheals on the skin, and painful lumps
- joints - aching joints are very common with vasculitis. They may swell up and feel warm too
- kidneys - vasculitis often leads to kidney damage which shows up as high blood pressure or blood and protein in the urine
- brain - headaches are a symptom of vasculitis affecting the brain
- peripheral nerves (in the limbs) - numbness, tingling, and loss of strength in a limb
- intestines - if the blood flow to the intestines is reduced, this can cause tummy pain and bloating
- heart - the heart is a muscle that is supplied with blood by the coronary arteries. If the blood supply is affected, it can cause chest pain when exercising, called angina. Pre-school children may not be able to describe this symptom
- lungs - symptoms of fever, cough, chest pain and shortness of breath tend to show up on x-rays as pneumonia
- eyes - the blood vessels in the back of the eye (retina) can be affected, which leads to blurring or even loss of sight. It may be accompanied by a severe headache too.

How is vasculitis diagnosed?

Henoch-Schönlein Purpura and Kawasaki disease are often easy to diagnose because of the fairly typical symptoms. The other diseases, which are much less common, have symptoms that can look very similar to those of other diseases. This may mean it takes a while to achieve a firm diagnosis. As it affects various parts of the body, a multidisciplinary approach is often needed, with input from rheumatologists (specialists of the musculoskeletal system), nephrologists (kidney specialists), neurologists (brain and nerve specialists), cardiologists (heart specialists), dermatologists (skin specialists) and so on. At Great Ormond Street Hospital, these doctors meet regularly to hold clinics to diagnose and treat patients with vasculitis.

In some cases, the doctors can only decide on a diagnosis once other conditions have been ruled out. In other cases, specific blood tests, x-rays or biopsies from affected organs are helpful.

What causes vasculitis?

We do not know 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, but we do not know 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. In most cases of the uncommon and most severe vasculitis diseases, quite strong treatment with different medicines is needed, for instance, steroids that reduce the inflammation and damp down the overactive parts of the immune system. Other medicines, like cyclophosphamide, may also be used alongside steroids to damp down the immune system further. Less powerful medicines or lower doses of stronger medicines will then be used to maintain improvement.

- Steroids – Most severe cases of vasculitis will need treatment with steroids. It will often be started as injections of high doses of methylprednisolone in 'pulses' or blocks of treatment. After that the child will often need high doses of prednisolone as tablets or medicines, which we gradually reduce or 'wean' to as low a dose as possible. Long term treatment, for a year or more, with low doses of prednisolone is often needed. The steroids are often very good at reducing the inflammation and damp down the immune system. Unfortunately, as with many powerful medicines, there are side effects. Most side effects relate to the cumulative total dose received. For more information, please see our leaflet on steroids.

- Immunosuppressant medicines – A number of different immunosuppressant medicines are used in the treatment of severe vasculitis. The most common is cyclophosphamide, which will often be started as injections in 'pulses' or blocks of treatment. After that the child will often need high doses of cyclophosphamide as tablets or medicines, which we gradually reduce or 'wean' to as low a dose as possible. Sometimes, other medicines may be used for maintenance, including azathioprine, cyclosporin A, MMF, colchicine, methotrexate and thalidomide. As in steroid medicines, these can be effective but do have side effects. More information is available in our leaflets. Plasma exchange is also used in severe cases. Please see our leaflet for more information.

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

Henoch-Schönlein Purpura

What is Henoch-Schönlein Purpura?

It is a disease where small blood vessels called capillaries become inflamed and damaged. It is named after the two doctors who first described the disease and is often referred to as HSP for short. It mainly affects four organs:

- the skin causing a purpuric rash, which in severe cases can become swollen and ulcerated;
- the digestive system causing severe stomach pains and blood in the faeces (poo). In a small number of children, this is so severe that surgery is needed.
- the joints and the tissue around the joints causing severe pain and there may be difficulty in walking;
- inflammation in the kidneys causing blood and protein in the urine and sometimes increased blood pressure.

In a few cases, other parts of the body can be affected too.

How common is it?

It occurs mainly in young children of school age, but has also occurred in younger children and adults. It seems to affect slightly higher numbers of males than females. It is the most common type of childhood vasculitis and occurs in about 14 out of every 100,000 children.

What are the symptoms?

In most cases, symptoms occur as above, but often not at the same time. Very typically, the symptoms come and go for several weeks.

How is it diagnosed?

In most cases, doctors will be able to diagnose it easily as the symptoms are very similar from child to child.

How is it treated?

There is no specific treatment for HSP, only treatment for the symptoms. If the joints are uncomfortable, then a mild pain relieving medicine should be taken to reduce any pain. Steroids can also help some children with severe symptoms.

What is the outlook for people with HSP?

The outlook for most children overall is very good. The symptoms tend to disappear within a few weeks, although in about half of the children affected they may return weeks or months later. If the kidneys have been affected, in a small number of cases, this can lead to kidney failure, which would mean that your child might need dialysis later.

Kawasaki disease

What is Kawasaki disease?

It is a disease that affects young children, named after the doctor who first reported it. It usually starts with a fever and a rash like a 'normal' infection, but it tends to last more than two weeks. Other symptoms develop during this time as well.

How common is it?

Kawasaki disease is the second most common type of childhood vasculitis. The exact incidence is not known, but reports suggest that it affects eight in every 100,000 children under five years old in the UK. It tends to affect children under the age of five, and is more common in males than females. It has been reported in all racial groups, although it occurs more often in Japan, where it was first reported.

What are the symptoms?

The main symptoms include fever (over 38°C), redness of the insides of the eyelids, lips, tongue and inside the mouth. The lymph nodes in the neck may also become swollen and there may be a rash, especially on the chest. The palms of the hands and soles of the feet may turn a bright red colour and become puffy. The skin at the ends of the fingers may also turn bright red and start to peel. There may also be swelling of the joints (arthritis) in older children, which usually affects large and small joints on both sides of the body. Other symptoms can include diarrhoea and vomiting, stomach pain and irritability. Some children develop symptoms like breathlessness and chest pain if the heart is affected.

How is it diagnosed?

The doctors will need to rule out other infections, but in most cases, the doctors will be able to diagnose Kawasaki disease because the symptoms are rather similar from child to child. As about 20 to 40 per cent of children develop vasculitis in the blood vessels around the heart, an echocardiogram (ECHO) or an angiogram is often needed. If the blood vessels are affected it may lead to a bulge developing in the wall of a heart blood vessel (coronary aneurysm).

How is it treated?

Kawasaki disease is usually treated aggressively in hospital with high doses of a medication called gamma globulin given directly into a vein (intravenously or IV) through a 'drip'. If this is given early enough in the course of the disease, it can reduce the chance of future heart problems. Your child may also be given aspirin, which reduces the chance of blood clots developing.

Long term follow up treatment is usual for most children, to check how the heart is working and catch any problems that may occur, early.

What is the outlook for people with Kawasaki Disease?

It usually takes children a few weeks to start to feel better. Children who do not have any heart problems usually recover fully. A second attack of the disease is rare, but if it happens, steroid therapy is often started. The outlook for children whose heart has been affected varies from child to child, depending on the level of damage sustained.

ANCA associated vasculitis

ANCA associated vasculitis is a group of uncommon cases of vasculitis affecting the small blood vessels in many parts of the body. Most of these cases have 'ANCA antibodies'. These are antibodies attacking the kind of white blood cells that are called neutrophils. There are two main kinds of ANCA: P-ANCA (also called MPO ANCA) and C-ANCA (also called PR3 ANCA). There are three forms of ANCA vasculitis:

- microscopic polyangiitis;
- Wegeners granulomatosis;
- and renal limited vasculitis.

These diseases are defined from their symptoms and the kind of ANCA antibodies that are present.

Microscopic polyangiitis

What is microscopic polyangiitis?

It is a disease that affects small and medium blood vessels throughout the body. It usually affects the blood vessels in the skin, lungs, digestive system and kidneys.

How common is it?

In the UK, it affects about two in every 100,000 people, usually more males than females. It can appear at any age from childhood onwards, but is much more common in adults.

What are the symptoms?

The generalised symptoms include fever, fatigue, muscle aches and pains and weight loss. The other symptoms depend to an extent on which part of the body is affected. However, common symptoms include purpura, coughing up blood, abdominal pain and kidney problems.

How is it diagnosed?

It is diagnosed by the symptoms present. Most cases have high values of MPO ANCA. A kidney biopsy will often show severe kidney inflammation.

How is it treated?

Microscopic polyangiitis is treated with steroids and immunosupressants.

What is the outlook for people with microscopic polyangiitis?

If the patient receives early treatment, the outlook is fairly good. Most patients go into remission and a high proportion of these do not have a relapse, where the disease returns. Most patients continue to lead normal lives, but will have to continue to see their doctor regularly to check for signs of a relapse.

Wegener's granulomatosis

What is Wegener's granulomatosis?

It is a very rare disease that starts with inflammation of the tissues in the nose, throat and lungs, but may develop into vasculitis affecting blood vessels throughout the body

including severe kidney inflammation. It is named after the doctor who first reported it. As with most types of vasculitis, we do not know exactly what causes the disease to develop.

How common is it?

The exact incidence is not known, but it can occur at any age. It is twice as common in men than in women. It is also more common in white people.

What are the symptoms?

The first symptoms tend to affect the nose, ears and windpipe and may include nosebleeds, sinusitis (inflammation of the sinuses), middle ear infections, and coughing. A high temperature, loss of appetite, joint pains and swelling, and inflammation of the eye or ear may occur as well. The disease may also affect other blood vessels, especially the arteries to the heart, which can cause chest pain or a heart attack, or those leading to the brain and spinal cord, which can cause similar symptoms to a lot of neurological diseases. Rarely the bowel is affected, causing pain and blood in the faeces (poo).

The disease may also spread causing inflammation of blood vessels throughout the body. These can affect the skin, causing sore patches that spread and may leave scars. The disease may also affect the kidneys, which could lead to kidney damage, although this ranges from mild kidney damage to kidney failure requiring dialysis.

How is it diagnosed?

It is diagnosed by the symptoms present and a positive result for PR3 ANCA.

How is it treated?

The treatment is similar to that used for microscopic polyangiitis. Taking the antibiotic co-trimoxazole long term to prevent infections has been show to be beneficial in preventing relapses.

What is the outlook for people with Wegener's granulomatosis?

If the patient receives treatment, the outlook is reasonably good. However, about half of the people with the disease have a relapse where the disease returns. This tends to happen within a couple of years of stopping treatment but could occur anytime. Most patients lead normal lives but will have to continue to see their doctor regularly to check for signs of a relapse. The outlook for children whose kidneys have been affected varies from child to child, depending on the level of damage sustained.

■ Renal limited vasculitis

What is renal limited vasculitis?

Renal limited vasculitis might be a form of microscopic polyangiitis with symptoms only from the kidneys. The main feature is glomerulonephritis (inflammation in their kidneys). The diagnosis is made on the kidney biopsy and with the ANCA antibodies. The treatment is similar to that of MPA.

Other types of vasculitis

■ Polyarteritis nodosa

What is polyarteritis nodosa?

It is a disease where medium and small-sized arteries become inflamed and damaged. This leads to problems with blood supply to various parts of the body, depending on the arteries affected. We do not know exactly what causes the disease to develop. Sometimes an infection can seem to start the inflammation, especially in one type of polyarteritis nodosa where the skin shows symptoms but the internal organs do not.

How common is it?

It tends to develop between the ages of 40 and 50 in adults, but childhood polyarteritis nodosa most commonly occurs around the age of nine years. There are no exact figures for how many children have polyarteritis nodosa but we know that it is rare.

What are the symptoms?

Fever is an early symptom along with tummy pain, pins and needles in the hands and feet, weakness and weight loss. Muscle and joint pain is also common, and the joints may become inflamed and swollen. The blood vessels near the surface of the skin may feel lumpy and rarely, ulcers can develop on the skin over these blood vessels.

Children with polyarteritis nodosa develop kidney damage. Other symptoms depend on the arteries affected, which may be in the digestive system, the heart, the brain or the liver.

How is it diagnosed?

As with most types of vasculitis, it is diagnosed by the symptoms and signs that indicate inflammation and when doctors have ruled out other diseases that can have these symptoms. The diagnosis can be confirmed by a biopsy of an affected blood vessel. This involves taking a small sample of tissue containing blood vessels to examine under a microscope. 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 angiogram can show up damage in the arteries. For more information about this test, please see our leaflet Angiography and angioplasty: information for families.

How is it treated?

The treatment is similar to that used for microscopic polyangiitis and Wegener's granulomatosis.

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. Many will respond to the medicines and not have further relapses. Others will have a more chronic, relapsing course and require long-term maintenance treatment. This may include medicines to treat the damage caused by the disease before it is under control, such as raised blood pressure from damage to the kidneys. Long-term side effects of both the disease and the medicines used to treat it can occur.

■ Takayasu's arteritis

What is Takayasu's arteritis?

It is a disease where the aorta (large artery

leaving the heart) and the blood vessels branching off of it become inflamed and damaged. This usually leads to problems with the blood supply to the head and arms, but can affect other parts of the body. It is named after the doctor who first reported it. As with most types of vasculitis, we do not know exactly what causes the disease to develop.

How common is it?

It tends to develop in people between the ages of 15 and 30, and affects about eight times as many women as men. It can affect all races, although it seems more common in women of Asian origin.

What are the symptoms?

Some patients with the disease seem to start with an acute (or sudden) phase where there is a general feeling of being unwell, fever, night sweats, weight loss, joint pain and tiredness. Some people also have anaemia. This sudden phase seems to improve and the disease then has an ongoing phase. The main symptoms of this phase are temperature changes, pins and needles and pain in the affected arm or leg. These symptoms are caused by damage to the aorta and other arteries as a result of inflammation and narrowing of the vessel, which reduces blood flow through it. Other patients with the disease do not seem to have the sudden phase but develop the ongoing symptoms straightaway.

When blood flows through the aorta and other vessels is reduced, this can lead to fainting and transient ischaemic attacks (TIAs or mini-strokes), along with cramps in the jaw or arms. This reduction in blood supply can also cause the muscles in the face and arms to weaken and grow smaller. Sometimes aneurysms can develop, where the wall of the artery balloons out and weakens. If the artery supplying blood to the kidneys is affected, damage to the kidneys can develop which can lead to high blood pressure and kidney failure. The heart and lungs can also be affected too.

How is it diagnosed?

It is usually diagnosed by a physical examination including checking the patient's pulse and blood pressure. If a person has the disease, the pulse may be very weak and difficult to find, and the blood pressure in the arms is much lower than in the legs. The blood pressure may differ from one side of the body to the other. Occasionally, there may be a murmur where the blood makes a noise as it rushes through a narrowed artery at high pressure.

How is it treated?

The disease tends to be treated with medications like steroids and immunosuppressants, which damp down the immune system. If there is a risk of mini-strokes, then a blood-thinning medication may be suggested. If blood pressure is high, then medications to lower this will also be suggested.

If the arteries are narrowed to a dangerous extent, then they may need to be unblocked (angioplasty), although clear discussion about the possible risks and benefits of this procedure is needed.

What is the outlook for people with Takayasu's arteritis?

The outlook depends on whether serious complications of the arteries have occurred. In people with no serious complications, the outlook is good. The outlook is also good if serious complications are treated quickly before too much damage is done. Otherwise, people may need repeated operations or procedures to correct any damage that occurs as well as medications for a long period.

More information and support groups

General information about vasculitis

There is no support group in the UK for vasculitis but the following organisations provide information and support:

Arthritis Care Youth Service
The Source
18 Stephenson Way
London NW1 2HD
Youth Helpline: 0808 808 2000
for people up to 26 years of age.
Mondays to Fridays 10am to 2pm
Website: www.arthritiscare.org.uk

Arthritis Research Campaign
Copeman House
St Mary's Court
St Mary's Gate
Chesterfield S41 7TD
Tel: 0870 850 5000
Website: www.arc.org.uk

Stuart Strange Vasculitis Trust
12 Acton Road
Mackworth
Derby DE22 4JF
Tel: 01332 521595
Website: vasculitis-uk.org

European Vasculitis Study Group
Website: www.vasculitis.org

Cleveland Clinic (USA)
Website: www.clevelandclinic.org/arthritis/vasculitis/

Johns Hopkins Vasculitis Center (USA)
Website: vasculitis.med.jhu.edu/

There are few support organisations for the specific types of vasculitis, but the following organisations and website might be useful:

Takayasu's Arteritis Foundation International
Website: www.takayasu.org

Wegener's Granulomatosis UK
Email: wegeners.uk@btinternet.com
Website: www.btinternet.com/~wegeners.uk/

Kawasaki Support Group
13 Norwood Grove
Potters Green
Coventry CV2 2FR
Tel/Fax 024 7661 2178

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

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