

Vasculitis

This booklet provides information and answers to your questions about this condition.

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What is vasculitis?



Vasculitis means inflammation of the blood vessels. There are several different types of the condition, many with unknown causes, but treatments can be very effective. In this booklet we'll briefly explain the main types of vasculitis, how they're diagnosed and treated, what you can do to help yourself and where to get more information.

At the back of this booklet you'll find a brief glossary of medical words – we've underlined these when they're first used.

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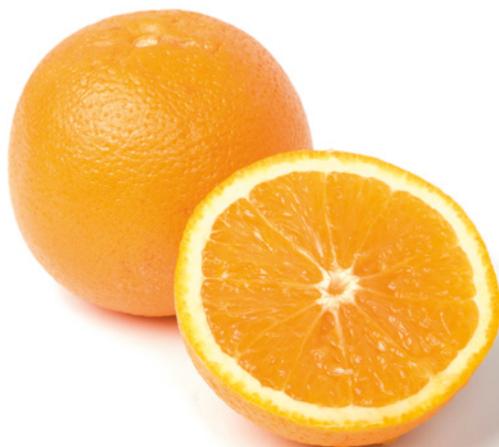
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At a glance

Vasculitis

What is vasculitis?

Vasculitis means inflammation of the blood vessels. It can affect any of the body's blood vessels, causing a variety of different symptoms and problems.

What are the symptoms?

The main symptom of vasculitis is inflammation, and this can be painful. With many forms of the condition the inflammation is internal and you can't see it. Because vasculitis takes different forms, the symptoms vary from person to person. **Many people with vasculitis feel unwell with fever, sweats, fatigue and weight loss. Sometimes these can be the first symptoms you feel, so it's important to be seen by your GP who will consider whether you need to be referred to a specialist.**

Other symptoms vary according to which part of the body is affected, for example:

- **skin** – rash of spots that can rupture, leaving open sores (ulcers)
- **lungs** – coughing or breathlessness
- **nerves** – tingling (pins and needles), pain and/or weakness in the arms and legs
- **kidneys** – problems passing urine, or blood in the urine

There are several different types of vasculitis but all are treatable.

- **fingers and toes** – can turn white or blue, tingle or hurt when exposed to cold conditions (Raynaud's phenomenon).

Headaches, jaw pain and eye problems (such as double vision or blurring) are all possible symptoms of a type of vasculitis called giant cell arteritis (GCA).

What types of vasculitis are there?

Vasculitis can be primary, where it occurs on its own, or secondary, where it occurs with another condition. The following are types of primary vasculitis:

- **Takayasu arteritis (TA)** – a disease that causes inflammation within the large arteries. TA particularly affects the aorta and its main branches. The aorta is the main artery carrying blood from the heart to the rest of the body.
- **Giant cell arteritis (temporal arteritis)** – affects the large arteries that supply the head and neck, especially the temporal artery (very rare in people under 50).

- **Polyarteritis nodosa (PAN)** – inflammation in the medium-sized arteries, such as those supplying the gut and kidneys.
- **Kawasaki disease** – affects small and medium-sized arteries in young children (usually aged under five years).
- **Granulomatosis with polyangiitis (Wegener’s)** – involves inflammation of small and medium-sized arteries in the skin, lungs, eyes and kidneys. If a biopsy is taken, under a microscope, doctors can see swellings called granulomata in this type of vasculitis.
- **Behçet’s syndrome**, or Behçet’s disease – is a rare autoimmune condition that often involves mouth ulcers, genital ulcers, skin problems and eye inflammation. Different parts of the body can be involved, so it can be treated with different medications.
- **Eosinophilic granulomatosis with polyangiitis (Churg–Strauss)** – asthma developing in adult life, followed by inflammation of the blood vessels; this form of vasculitis usually affects the nerves, causing weakness, pins and needles and numbness. If a biopsy is undertaken, under a microscope, doctors can see swellings called granulomatoma.
- **Microscopic polyangiitis** – causes kidney problems that without treatment can lead to kidney failure, and can also involve the lungs, with bleeding that can cause breathlessness.
- **Cryoglobulin-associated vasculitis** – causes a reduction in blood flow or even a blockage, causing damage to the organs or body tissues.
- **Immunoglobulin A vasculitis (Henoch–Schönlein)** – affects small blood vessels (capillaries) and mostly affects children between 2–10 years; symptoms include a skin rash, arthritis, abdominal pain, and/or vomiting, passing blood in the stools and/or urine, fever, headaches and loss of appetite.

How is it diagnosed?

Vasculitis is diagnosed using blood tests, x-rays or other scans and biopsies (getting a tissue sample to look at under a microscope).

What tests are there?

The following tests may be used to check for types of vasculitis:

- blood tests – to test for inflammation, low blood count, kidney and liver function, or to test for antineutrophil cytoplasmic antibodies (ANCA)

Only 10–15 out of every 100,000 people will develop vasculitis in the UK each year.

- urine tests – to test for kidney inflammation
- x-rays, computerised tomography (CT) and magnetic resonance imaging (MRI) scans to test for chest or sinus problems
- heart scans such as echocardiograms and electrocardiograms (ECGs) to check your heart is healthy
- a biopsy may be advised to check if your temporal arteries, liver, kidneys, muscles or lungs are affected
- an ear, nose and throat (ENT) examination to check if these parts of the body are affected
- a special x-ray or MRI scan called an angiogram to see which blood vessels are involved.

What treatments are there?

Treatments for the various types of vasculitis include:

- drug treatments, including steroid tablets or injections, immunosuppressive drugs, biological therapies, immunoglobulin infusions, antiviral treatments and antibacterial drugs
- plasma exchange (sometimes called blood washing), which may be needed by a small number of people with the most severe forms of vasculitis
- dialysis in cases of severe kidney damage.

What else might help?

There are some things you can do to help yourself if you have vasculitis, including:

- not smoking
- balancing regular exercise with rest
- eating a healthy balanced diet, including enough calcium and vitamin D if you're on steroids
- keeping warm (especially if the vasculitis has caused Raynaud's phenomenon)
- learning about the treatment you're on and what you need to do to reduce your risk of side-effects, including vaccinations against seasonal flu and pneumonia.

Who diagnoses and treats vasculitis?

If you have vasculitis it's important you have one consultant who is in overall charge of your care. The management and care of patients with vasculitis is usually undertaken mainly by a rheumatologist, as they specialise in inflammatory diseases, or less frequently by a nephrologist (kidney doctor), especially if the kidneys are affected.

However vasculitis in some circumstances may affect a variety of different organs.

If required, your care should involve a multi-disciplinary team of consultants with different specialities, working together. For example, if your lungs and airways are seriously affected, care may involve a respiratory consultant. In some cases an ear, nose and throat (ENT) consultant, an ophthalmologist, neurologist, gastro-enterologist, immunologist or dermatologist may also be involved.



What is vasculitis?

Vasculitis means inflammation of the blood vessels, the tubes that carry blood around your body. There are three types of blood vessel which can be affected by vasculitis (see Figure 1):

The organs and tissues in your body need a regular blood supply to work properly. Inflammation causes swelling of the blood vessel walls, reducing or even blocking the flow of blood to the tissues and organs (see Figures 2 and 3).

Figure 1: The three types of blood vessel that can be affected by vasculitis

Arteries	Veins	Capillaries
take blood from the heart to different parts of the body such as organs (for example kidneys) and tissues (for example skin).	take blood back to the heart.	tiny vessels between the arteries and the veins where oxygen and other materials pass from the blood into the tissues.

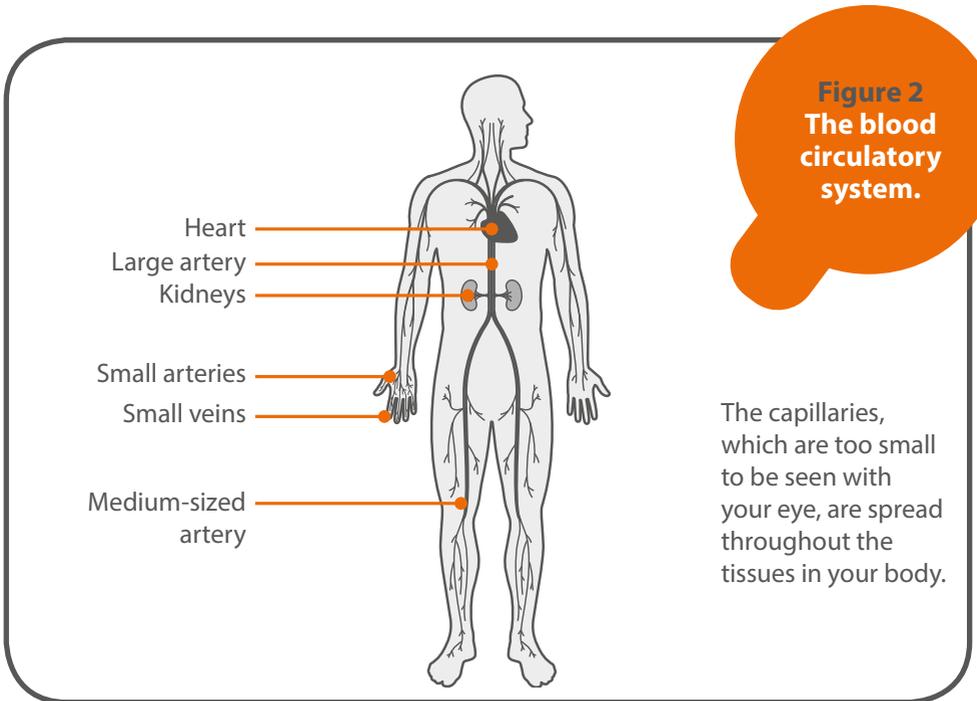


Figure 2
The blood circulatory system.

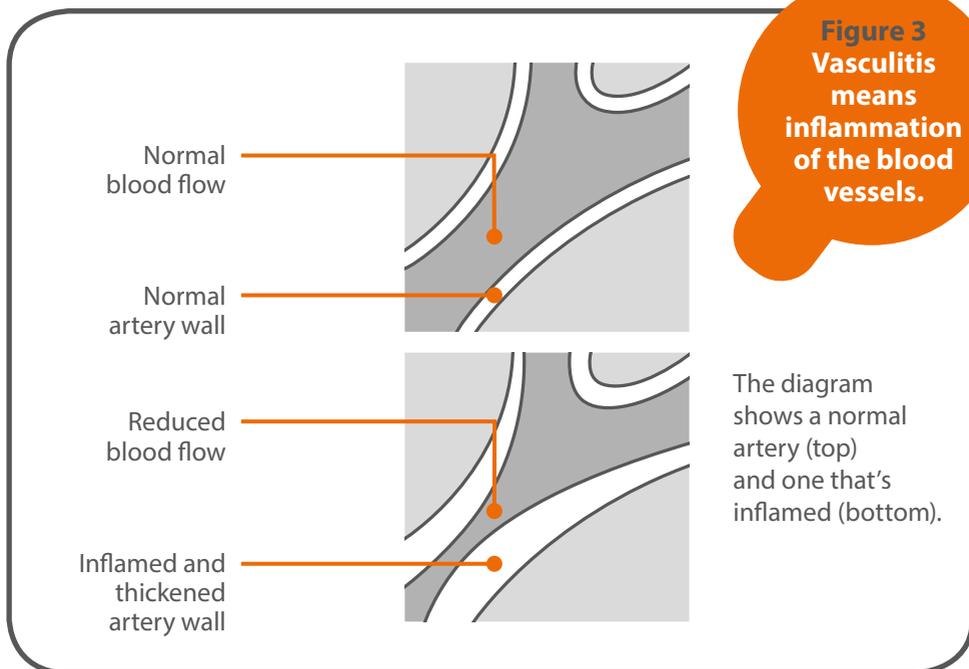
The capillaries, which are too small to be seen with your eye, are spread throughout the tissues in your body.

Vasculitis can cause a range of symptoms and possible complications. The amount of damage vasculitis causes depends on which part of the body is affected. The larger the affected blood vessels, the more damage there may be; the more important the affected body tissue, the more serious the damage will be.

The walls of affected blood vessels can swell and bulge (this is called an aneurysm) and may even burst, causing bleeding inside your body. Apart from the damage to the blood vessel itself, this can lead to damage in the tissues or organs that are supplied by the blood vessel.

Vasculitis can appear suddenly in someone who has previously been completely well – when it occurs on its own, doctors call this primary vasculitis. Vasculitis can also occur alongside other conditions (including rheumatoid arthritis, lupus or Sjögren's syndrome) in which case it's known as secondary vasculitis.

i See Arthritis Research UK booklets
*Lupus (SLE); Rheumatoid arthritis;
Sjögren's syndrome.*



The symptoms of vasculitis vary according to which part of the body is affected.

What are the symptoms of vasculitis?

When any part of your body is inflamed, it swells and is uncomfortable or painful. With many types of vasculitis the swelling is inside the body and you can't see any symptoms on the outside.

Vasculitis takes different forms according to which blood vessels are affected, and symptoms vary. Many people with vasculitis feel unwell and have fever, sweats, fatigue and weight loss. These can be the first symptoms experienced, so it's important to be seen by your GP. Other symptoms vary according to which part of the body is affected:

- **Skin** – vasculitis in the skin causes spots that can burst, leaving open sores (ulcers). When vasculitis only affects the skin, long-term effects aren't usually serious, and symptoms normally clear up once inflammation has settled.
- **Nose** – vasculitis here causes crusting inside the nose and nosebleeds. The shape of your nose can change.

- **Fingers and toes** – some people with vasculitis experience Raynaud's phenomenon, where the fingers or toes turn white or blue and may tingle or hurt when exposed to cold conditions.
- **Eyes** – some types of vasculitis can suddenly affect your vision or cause your eye/s to become red or painful. It's very important to see a doctor quickly if this happens.
- **Nerves** – inflammation of the nerves can cause tingling (pins and needles) pain and burning sensations or weakness in the arms and legs.
- **Joints** – vasculitis can cause joint pain or swelling.
- **Muscles** – inflammation here causes muscle aches, and eventually your muscles could become weak.
- **Lungs** – inflammation of the lungs causes coughing and shortness of breath.
- **Brain** – occasionally the blood vessels in the brain can be affected, causing problems like strokes.
- **Kidneys** – when vasculitis affects the kidneys there may be problems passing urine or blood in the urine. Vasculitis of the kidneys can be dangerous as symptoms may not appear until the kidneys have been damaged. In severe cases treatment on an artificial kidney (dialysis) machine may be necessary.

Headaches, pain in the jaw and problems with the eyes can be serious symptoms of giant cell arteritis (GCA).

What types of vasculitis are there?

Doctors usually define the types of vasculitis according to the size of the blood vessels involved (see Figure 4). The most serious types of vasculitis involve both small and medium arteries.

Takayasu arteritis (TA)

Takayasu arteritis (TA) is an inflammatory disease that affects the main artery from the heart (the aorta) and its large branches, usually in younger women. It's rare in the UK (there are only 100 new cases a year) but is more common in the Far East and Africa.

TA causes the arteries to narrow, and this can reduce the blood supply. The narrowing develops slowly and the arteries don't usually block completely, so there isn't usually a dangerous loss of blood supply to the arms or legs or any major organs. Other major arteries can also be affected, including the carotid arteries in the head and brain, coronary arteries in the heart, renal arteries to the kidneys and arteries that take blood to the arm.

Figure 4 Types of vasculitis can be defined according to the size of the blood vessels involved.

Vasculitis in large arteries:

- includes giant cell arteritis (temporal arteritis) and Takayasu arteritis.

Vasculitis in medium-sized arteries:

- includes polyarteritis nodosa and Kawasaki disease.

Vasculitis in small vessels:

- includes granulomatosis with polyangiitis, microscopic polyangiitis and eosinophilic granulomatosis with polyangiitis
- can happen with infections including hepatitis and, very occasionally, with different types of cancers
- can also be a result of rheumatic diseases
- includes immunoglobulin A vasculitis and cryoglobulin-associated vasculitis
- usually involves the skin and is also sometimes caused by a reaction to certain drugs.

Giant cell arteritis (temporal arteritis)

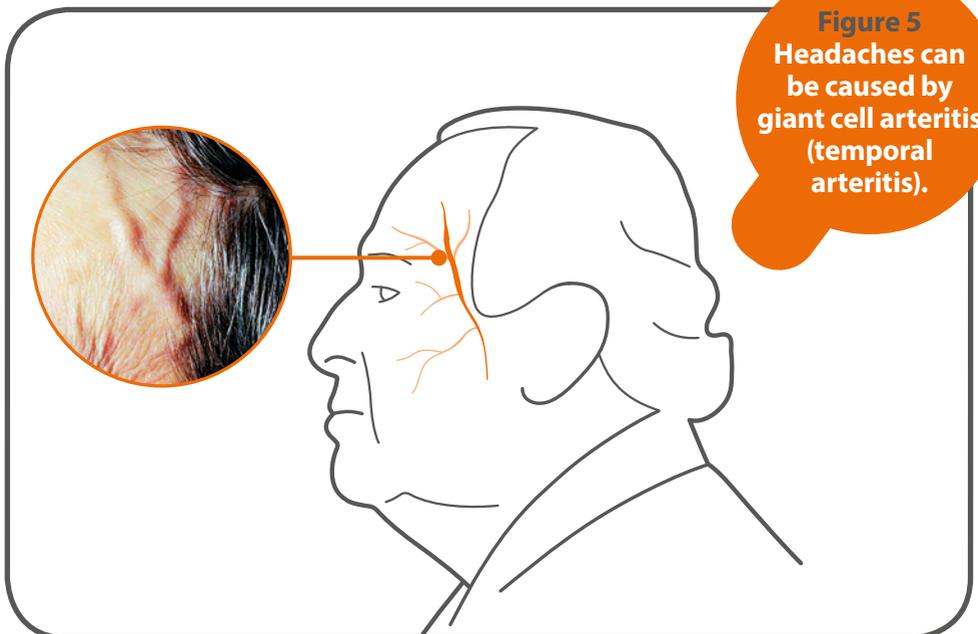
Giant cell arteritis, or GCA, affects the large arteries that supply the head and neck, especially the temporal artery which is found over the temples (see Figure 5). There are around 5,000 new cases a year in the UK and it's more common in northern Europe. GCA doesn't normally affect people below the age of 50.

GCA can cause headaches and is often associated with a condition called polymyalgia rheumatica (PMR), which causes inflammation and stiffness in the muscles of the shoulders and hips.

- i** GCA occasionally involves the blood supply to the eye, where it can cause blindness. If you develop symptoms in your eyes, such as blurring or double vision, you should see your doctor straight away as you'll need to be treated urgently.

Other blood vessels, such as the major arteries, can less commonly be involved in GCA.

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- i** See Arthritis Research UK booklets *Giant cell arteritis (temporal arteritis); Polymyalgia rheumatica (PMR)*.
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The type of vasculitis is usually defined by the size of the blood vessels involved.

Polyarteritis nodosa (PAN)

PAN can be very serious but is very rare – only about one out of every two million people in the UK develop PAN each year. In some cases PAN can be associated with hepatitis B virus infection. It causes inflammation in the medium-sized arteries, particularly those supplying the gut and kidneys. This may only affect part of the wall of the artery, which becomes weak and may bulge, forming an aneurysm. If it bursts it can cause serious internal bleeding. PAN can also involve the whole wall of the artery at a particular point, which causes a blockage.

Kawasaki disease

Kawasaki disease affects small and medium-sized arteries most commonly in children under five. It's sometimes called mucocutaneous lymph node syndrome (because it involves the mucous membrane).

Children with Kawasaki disease will feel unwell – they may have a high temperature, swollen glands in the neck (lymphadenopathy), an inflamed area around the eye and the mouth, and a skin rash similar to measles.

This condition is quite rare but can be serious if the arteries supplying the heart are inflamed (coronary arteritis). Up to 60% of people with Kawasaki disease have coronary arteritis.

Granulomatosis with polyangiitis (Wegener's granulomatosis)

The condition granulomatosis with polyangiitis is quite rare – altogether there are only 1,000 new cases of

granulomatosis with polyangiitis, eosinophilic granulomatosis with polyangiitis and microscopic polyangiitis a year in the UK. It's slightly more common in men than in women.

It usually develops with ear, nose and throat problems, including nosebleeds and crusting of the nose, and occasionally coughing up blood (haemoptysis). These symptoms can appear a year or two before more general vasculitis starts. This general vasculitis usually involves the skin, lungs, eyes and kidneys. The kidney problems can sometimes lead to kidney failure if they're not recognised early.

The name of this disease has changed recently. It was previously called Wegener's granulomatosis after the doctor who was thought to have discovered it. Polyangiitis means that many (poly = many) arteries are involved and granulomatosis means that when a biopsy of the inflamed tissue is studied under a microscope you can see swellings called granulomata. Remember that the terms Wegener's granulomatosis, granulomatosis with polyangiitis, and GPA describe exactly the same disease.



Behçet's syndrome

Behçet's syndrome, or Behçet's disease (pronounced betchets) is a rare autoimmune condition that can involve mouth ulcers, genital ulcers, skin problems and eye inflammation. It's not an infection and can't be passed from one person to another. It can also involve other areas of the body such as the gastrointestinal tract, as well as the pulmonary, musculoskeletal, cardiovascular and neurological systems. Because of the many different parts of the body that can be affected with Behçet's, many different medications can be used to control symptoms.

Eosinophilic granulomatosis with polyangiitis (Churg–Strauss syndrome)

Eosinophilic granulomatosis with polyangiitis causes asthma to develop in adults, followed by inflammation of the blood vessels caused by swellings called granulomas. There will also usually be a high number of eosinophils (a particular type of white cell) in the blood.

The condition differs from granulomatosis with polyangiitis because of the asthma. Unlike granulomatosis with polyangiitis, eosinophilic granulomatosis with polyangiitis rarely causes damage to your ears and nose.

Eosinophilic granulomatosis with polyangiitis can also affect the nerves, causing weakness, pins and needles or numbness. There's also a higher risk of your heart being involved, which can sometimes cause damage to the heart

muscle similar to the damage that occurs during a heart attack.

This disease was previously called Churg–Strauss syndrome after the doctors who were thought to have discovered it. The new name describes the features of the disease – eosinophils, granulomata and involvement of many blood vessels.

Microscopic polyangiitis

Almost all people with microscopic polyangiitis have kidney problems that can lead to raised blood pressure and kidney failure. People may find that they're tired because of anaemia. Blood tests will show that the kidney is inflamed. Microscopic polyangiitis can also involve the lungs, with bleeding that can cause breathlessness.

Cryoglobulin-associated vasculitis

In cryoglobulin-associated vasculitis, small-vessel vasculitis is associated with cryoglobulins – these are proteins in the blood that stick together in the cold. Having cryoglobulins can reduce the flow of blood or even block the blood vessels, causing damage to the organs or body tissues.

Immunoglobulin A vasculitis (Henoch–Schönlein purpura)

Immunoglobulin A vasculitis, also known as IgA vasculitis, affects the small blood vessels. It often follows a chest infection and may be an allergic reaction to a virus, food or drugs. It mostly affects children aged 2–10 years, and boys are affected more often than girls. It can also affect adults, but most of the 2,500 new cases a year in the UK are children.

The symptoms of IgA vasculitis include:

- a rash, often over the buttocks (starts red but develops into a bruised purple colour and appears over several days or even weeks)
- short-lived arthritis, especially of the larger joints
- stomach pain and/or vomiting or passing blood in stools
- passing blood in urine (indicating kidney problems)
- fever, headaches and loss of appetite.

The rash seen in IgA vasculitis is called purpura and was described by Drs Henoch and Schönlein, which explains the previous name of the disease.

In most cases the condition doesn't need specific treatment, although relapses are possible for up to a year after the original illness. Kidney problems are quite common, but serious kidney damage is rare. Occasionally other blood vessels are involved, and rarely more serious complications can occur, sometimes affecting the bowels or causing seizures.

Who gets vasculitis?

Vasculitis is uncommon. In every 100,000 people in the UK, only 10–15 will develop vasculitis each year. However, about 22 people per 100,000 aged over 50 years will develop GCA. The different types of vasculitis tend to affect different age groups, for example:



- GCA (temporal arteritis) is much more common in people over 50, and it's fairly common for it to be associated with a condition called polymyalgia rheumatica (PMR).
- Takayasu arteritis tends to affect younger Asian women.
- IgA vasculitis is much more common in children than in adults.
- Kawasaki disease only affects children under five.

What causes vasculitis?

There's no single cause of vasculitis, and in most cases the exact cause is unknown. We know that vasculitis isn't directly inherited through the genes we get from our parents, but genetic factors do play a part as several cases can occur in the same family. Genes could make you more likely to develop the condition, in which case it may only take a small trigger (such as an infection or drugs) to start this off.

We also know that some types of vasculitis – for example, those affecting the small blood vessels – can be related to infections, particularly those associated with hepatitis. Some cases of vasculitis happen after certain drugs have been used, for example propylthiouracil (used to treat thyroid disease) and allopurinol (used to treat gout), as well as non-steroidal anti-inflammatory drugs (NSAIDs) and antibiotics.

It's thought that most forms of vasculitis are a type of autoimmune disease.

This means that your body's defence mechanisms aren't doing their normal job of fighting infections, but instead attack a healthy part of the body, causing inflammation.

i See **Arthritis Research UK booklet and drug leaflet** *Gout; Allopurinol*.

What is the outlook?

The most severe types of vasculitis can be life-threatening. Early diagnosis and treatment is essential for the best chances of avoiding permanent damage to tissues and organs. Most types respond well to treatment, and for many of them you're likely to make a full recovery, although relapses in the future are possible. The outcome depends on the type of vasculitis and how it affects you. Overall, the best way to learn more about what might happen to you in the future is to talk to your doctor or another healthcare professional.

How is vasculitis diagnosed?

If you think you may be developing vasculitis, you must see your doctor as soon as possible. Infections, drugs and some foods can sometimes cause vasculitis, so your doctor will probably ask about the medications you've been taking and your general health during the past few weeks.

Treatments are given in stages to keep your vasculitis under control.

What tests are there?

There are many tests that may be done to help diagnose the condition. In this section we'll look at the most common, but there are others that you may need. Ask your rheumatologist, other specialist or GP if you're not sure about what a certain test will involve.

Blood tests may be used to measure inflammation, for example:

- erythrocyte sedimentation rate (ESR)
- C-reactive protein (CRP).

A full blood count can help to find out whether you have anaemia and whether you have normal levels of white blood cells (which fight infections) and platelets (which are involved in clotting).

Blood tests for antineutrophil cytoplasmic antibodies (ANCA) are important in the diagnosis of some types of vasculitis, particularly granulomatosis with polyangiitis, eosinophilic granulomatosis with polyangiitis and microscopic polyangiitis.

A urea and electrolytes (U&E) test, an estimated glomerular filtration rate (eGFR) test or a creatinine test may be used to check how your kidneys are working. Liver function tests may also be carried out to check how your liver is working.

If you have vasculitis along with other conditions, such as rheumatoid arthritis or lupus, then blood tests might be used



to assess how active these other diseases are. Blood tests can measure the level of rheumatoid factor in rheumatoid arthritis, or the levels of complement (an enzyme system or group of proteins in the blood) and antibodies in lupus.

Blood tests may be repeated from time to time to check how your condition is responding to treatment.

Other tests may be carried out to see how the affected body organs are working – for example:

- **Urine tests** will show the presence of blood and/or protein, which are often the first signs of an inflamed kidney. People with eosinophilic granulomatosis with polyangiitis, granulomatosis with polyangiitis or microscopic polyangiitis will have regular urine tests for blood and protein.
- **X-rays, CT and MRI scans** can be used to check for chest problems.
- **Echocardiograms and electrocardiograms (ECGs)** can be used to assess the heart.
- **A biopsy** may be needed to confirm whether the kidneys, the muscles, skin or lungs are affected by vasculitis. A small piece of tissue is removed from the organ in question for examination or testing in a laboratory.
- **An ear, nose and throat (ENT) assessment** may be needed for people with granulomatosis with polyangiitis who have symptoms in these parts of the body.

- **An angiogram** is often done where abdominal organs such as the kidney and gut are involved. This involves injecting dye into the arteries so that they show up on an x-ray. They can also be done in Takayasu arteritis and giant cell arteritis to see how much the large blood vessels are involved.

What treatments are there for vasculitis?

The treatments used for vasculitis will depend upon which blood vessels and organs are affected, as well as how much body tissue is affected. If the vasculitis only affects the skin, it may be enough to treat any underlying infection or to remove the drug that triggered the vasculitis. However, in most cases, drug treatment will be needed to control the disease and its symptoms and to stop or limit the damage caused by vasculitis.

Drugs

The two main types of drug used to treat vasculitis are steroids and immunosuppressant agents. Both act to dampen down the immune system to reduce the strength of its attack on the tissues of the body.

If you have vasculitis affecting the large blood vessels, then you'll probably be given steroid tablets. These are very effective for GCA and Takayasu arteritis, and often immunosuppressant agents aren't needed in these conditions.

Vasculitis varies from one person to the next, so it's important that you understand your doctor's advice clearly...

...and to make sure your treatment isn't interrupted.

If you have vasculitis affecting small and/or medium-sized blood vessels, then you may only need a small dose of steroids to control it. However, you may be given a combination of steroids and immunosuppressive drugs, probably over several years, especially if vasculitis affects the internal organs. For many types of vasculitis, including those affecting the kidney, lungs or other vital organs (especially if it involves both small and medium-sized blood vessels), your treatment will be given in stages (see Figure 6).

If you have vasculitis that mainly affects medium-sized arteries, then other treatments can help, depending on the condition:

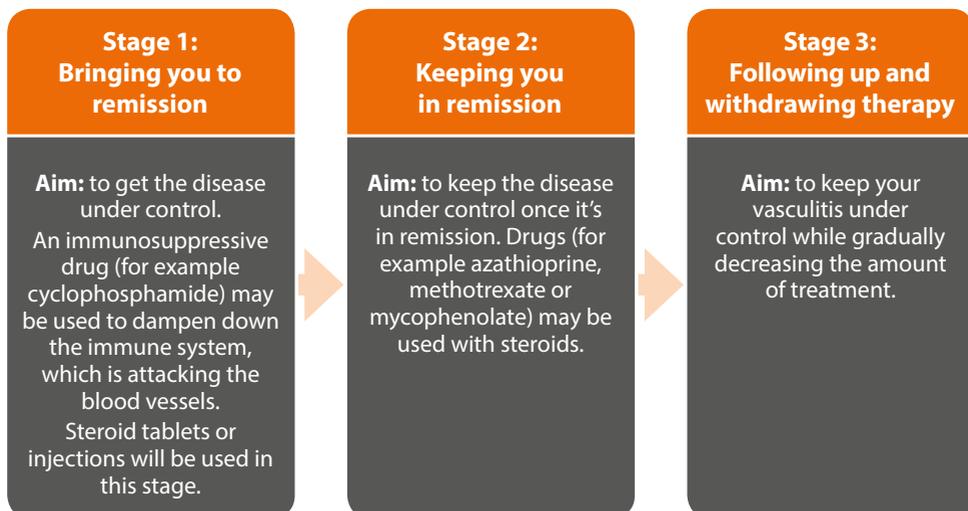
- Kawasaki disease can be treated effectively with injections of immunoglobulin (a type of protein).

- Hepatitis-associated PAN can be treated with antiviral treatment and plasma exchange.

Plasma exchange (also known as blood washing) involves being connected to a machine that your blood passes through before being returned to you so it can be cleaned of the factors causing the vasculitis. Only a few people with the most severe types of vasculitis – for example who have very severe kidney or lung disease – will need plasma exchange. This will be done in specialist centres.

Possible side-effects of steroids include weight gain, indigestion, diabetes, thinning of the skin and thinning of the bones (osteoporosis). If high doses of steroids are given, then you'll also be given drugs like bisphosphonates to help prevent osteoporosis.

Figure 6: The stages of treatment for some types of vasculitis



Gentle exercise can help keep muscles and joints healthy.

i See Arthritis Research UK booklet and drug leaflet *Osteoporosis; Steroid tablets.*

In some types of vasculitis (such as granulomatosis with polyangiitis), an infection may trigger a relapse. You may therefore be given antibacterial drugs such as co-trimoxazole to protect against this. These drugs can also help to protect against the increased risk of infection caused by the stronger immunosuppressive drugs.

As with all medications, there may sometimes be side-effects. Cyclophosphamide, for example, can cause bleeding from the bladder, hair thinning and an increased risk of infection. Unfortunately there's also a significant risk that it can reduce fertility in both men and women. Because of these risks, cyclophosphamide will be stopped or exchanged for a different immunosuppressive drug as soon as your vasculitis is controlled. This is usually azathioprine, but methotrexate or mycophenolate might be used instead. Rituximab, a biological therapy given by intravenous infusion, can also help to encourage remission in some types of vasculitis.

i See Arthritis Research UK drug leaflets *Azathioprine; Cyclophosphamide; Methotrexate; Mycophenolate; Rituximab.*

Self-help and daily living

If you do need treatment then it's very important that you follow your doctor's instructions carefully. It's important to learn and understand as much as you can about your illness and the treatment options; you can discuss alternative treatment options with your medical team. Vasculitis varies from one person to the next and from one type of vasculitis to another. It's important to speak to your doctor or other healthcare professional about any new symptoms you may have.

Exercise

Vasculitis can cause tiredness, and it's important to rest when you need to. However, you should also try to keep muscles and joints healthy by exercising. Start gently and gradually increase the amount of exercise you do. Include some weight-bearing exercise (anything that involves walking or running), and swimming is also recommended. Ask your doctor for advice on how much exercise you should expect to be able to do.

i See Arthritis Research UK booklet *Keep moving.*



Diet and nutrition

You won't usually need to keep to any special diet. A healthy, low-fat, nutritious and balanced diet is important for everyone, but if you're on steroids it's particularly important because these can increase your appetite and cause weight gain. Try not to over eat, and cut down on fatty and sugary foods and others which are particularly high in calories. Instead, eat lots of fresh fruit and vegetables and starchy foods like potatoes and wholemeal bread, pasta and rice.

Drinking plenty of water is helpful. It's healthy for everyone not to drink more than the recommended units of alcohol a day; 3-4 a day for men and 2-3 a day for women. Having at least two alcohol-free days a week is advisable. If you're taking steroids then you're more at risk of osteoporosis; having plenty of calcium in your diet is important to help prevent this from developing. Foods that are good sources of calcium include tinned sardines (with bones), skimmed milk, yoghurt and certain vegetables such as broccoli.

Most forms of vasculitis can now be controlled and sometimes completely cured.

i See Arthritis Research UK booklet *Diet and arthritis*.

Stop smoking

Smoking makes the blood vessels become narrower inside and can make vasculitis symptoms worse. Smoking can also worsen lung inflammation. It's therefore very important to try to stop smoking. This will also improve any symptoms of Raynaud's phenomenon. If you want to stop smoking see your doctor who will be able to give you advice and help about quitting.

Keep warm

If your fingers or toes turn blue in response to the cold, this may be due to Raynaud's phenomenon. Wearing warm clothes, including warm socks and gloves, should improve blood circulation to your hands and feet by helping to keep the blood vessels open.

Research and new developments

In the past, some types of vasculitis were very serious diseases, especially if they affected both small and medium-sized arteries. Advances in treatment over the last two decades have completely changed this and most forms of vasculitis can now be controlled and sometimes completely cured. The new problem to be faced is that some treatments have unpleasant side-effects – some are almost

as damaging as the disease itself, though in different ways.

Researchers are currently trying to better understand what causes vasculitis.

Comparing what happens in different countries may help to find the answer – for example:

- Takayasu arteritis (TA) is much more common in the Far East and in Africa than in the UK.
- GCA is extremely rare in India but very common in northern Europe.
- Granulomatosis with polyangiitis is more common in northern Europe than in southern Europe.
- Microscopic polyangiitis is more common in southern Europe than northern Europe.

Researchers at the University of Cambridge have received funding from Arthritis Research UK to carry out a large genetic study to investigate whether common differences that we all carry in our genes might predispose some people to developing a disease called ANCA-associated Vasculitis.

Other work also being carried out by scientists at the University of Cambridge is looking to increase our understanding of the role played by bacteria in the nose on the development of granulomatosis with polyangiitis. It is hoped the research may enable clinicians to predict those patients with a high risk of disease recurrence.

Glossary

Anaemia – a shortage of haemoglobin (oxygen-carrying pigment) in the blood which makes it more difficult for the blood to carry oxygen around the body. Anaemia can be caused by some rheumatic diseases such as rheumatoid arthritis or lupus, or by a shortage of iron in the diet. It can also be a side-effect of some drugs used to treat arthritis.

Aneurysm – a balloon-shaped sac that bulges out of a blood vessel wall. Some diseases or infections can weaken the vessel wall, and the pressure of blood causes the weakened section to balloon outwards. The bigger the aneurysm gets, the greater the risk of it bursting, which can result in internal bleeding and other complications.

Angiogram – a type of scan that looks inside the arteries in your heart to find out where and how severe any narrowed areas are. It's also known as a coronary angiogram or a cardiac catheterisation.

Antibodies – blood proteins that form in response to germs, viruses or any other substances that the body sees as foreign or dangerous. The role of antibodies is to attack these foreign substances and make them harmless.

Antineutrophil cytoplasmic antibodies (ANCA) – a group of antibodies which attach themselves to molecules in white blood cells rather than to foreign substances. They're commonly found in blood tests for a number of autoimmune disorders such as vasculitis, although they can't definitely confirm the diagnosis.

Antiviral treatment – drugs used to treat infections caused by particular viruses.

Autoimmune disease – a disorder of the body's defence mechanism (immune system), in which antibodies and other components of the immune system attack the body's own tissue rather than germs, viruses and other foreign substances.

Biological therapies – drugs that reduce joint inflammation in people with rheumatoid arthritis and some other inflammatory diseases. They work by targeting specific molecules involved in the inflammatory process and include anti-TNF drugs (adalimumab, etanercept and infliximab) and rituximab.

Biopsy – the removal of a small amount of living tissue from your body. The sample can help diagnose illness when examined under a microscope.

Bisphosphonates – drugs used to prevent the loss of bone mass and treat bone disorders such as osteoporosis and Paget's disease. They work by reducing high levels of calcium in the blood and by slowing down bone metabolism.

Complement – an enzyme 'system' in the blood. An enzyme is a substance that speeds up a biological reaction. Complement consists of at least 19 separate proteins and plays an important part in the body's immune system. It allows foreign particles or micro-organisms to be made harmless, but also generates inflammation. Blood tests can show how much of each of the major elements of complement is present.

Computerised tomography (CT) scan – a type of scan that records images of sections or 'slices' of the body using x-rays. These images are then transformed by a computer into cross-sectional pictures.

C-reactive protein (CRP) – a protein found in the blood. The level of C-reactive protein in the blood rises in response to inflammation and a blood test for the protein can therefore be used as a measure of inflammation or disease activity.

Creatinine test – a urine or blood test used to monitor the kidneys. Creatinine is a waste product from your muscles that's processed by your kidneys, so by checking the levels of creatinine a doctor can tell how well your kidneys are working.

Dialysis – a method of separating particles in a liquid by passing them through a membrane. In kidney dialysis the blood is circulated through a special machine that uses this method to remove waste materials or poisons from the blood.

Echocardiogram – a type of scan that uses ultrasound waves to create detailed pictures of the inside of the heart. This test helps show the structure and movement of the heart.

Electrocardiogram (ECG) – a test that records the electrical activity of the heart using a machine called an electrocardiograph. The aim of an ECG is to detect unusual heart rhythms and to identify heart problems.

Eosinophil – a type of white blood cell which is able to absorb foreign matter. These cells are involved in allergic responses in the body.

Erythrocyte sedimentation rate (ESR) – a test that shows the level of inflammation in the body and can help in the diagnosis of rheumatoid arthritis. Blood is separated in a machine with a rapidly rotating container (a centrifuge), then left to stand in a test tube. The ESR test measures the speed at which the red blood cells (erythrocytes) settle.

Estimated glomerular filtration rate (eGFR) test – a test which estimates the volume of blood filtered by your kidneys over a given period of time to assess how well your kidneys are working.

Fatigue – a feeling of weariness that's more extreme than simple tiredness. It can affect you physically, but it can also affect your concentration and motivation, and often comes on for no apparent reason and without warning.

Gout – an inflammatory arthritis caused by a reaction to the formation of urate crystals in the joint. Gout comes and goes in severe flare-ups at first, but if not treated it can eventually lead to joint damage. It often affects the big toe.

Hepatitis – inflammation of the liver. This can cause yellowing of the skin (jaundice), fever, abdominal (tummy) pain and an enlarged liver.

Immunoglobulins – a class of blood proteins that are responsible for immunity to specific infections.

Immunosuppressive drugs – drugs that suppress the actions of the immune system. They're often used in conditions such as rheumatoid arthritis where

the immune system attacks the body's own tissues.

Inflammation – a normal reaction to injury or infection of living tissues. The flow of blood increases, resulting in heat and redness in the affected tissues, and fluid and cells leak into the tissue, causing swelling.

Infusion – a procedure where fluid is transferred directly into a vein through a thin plastic tube (cannula) over a period of hours or days. Intravenous infusions are used to transfer blood, fluids and essential salts. It's commonly known as a drip.

Liver function tests – blood tests used to check the healthy functioning of the liver. They're frequently performed to monitor drug treatment.

Lupus (systemic lupus erythematosus or SLE) – an autoimmune disease in which the immune system attacks the body's own tissues. It can affect the skin, hair and joints and may also affect internal organs. It's often linked to a condition called antiphospholipid syndrome (APS).

Magnetic resonance imaging (MRI) scan – a type of scan that uses high-frequency radio waves in a strong magnetic field to build up pictures of the inside of the body. It works by detecting water molecules in the body's tissue that give out a characteristic signal in the magnetic field. An MRI scan can show up soft-tissue structures as well as bones.

Mucous membrane – the type of membrane that lines areas of the body such as the mouth, nasal passages,

stomach and gut, vagina, and passages to the lungs.

Osteoporosis – a condition where bones become less dense and more fragile, which means they break or fracture more easily.

Plasma exchange – a procedure to separate blood into its different parts and remove the plasma and the autoantibodies it contains which cause disease. Plasma from a donor or a plasma substitute is then returned in its place.

Polymyalgia rheumatica (PMR) – a rheumatic condition in which you have many (poly) painful muscles (myalgia). It's characterised by pain and stiffness of the muscles of the neck, hips, shoulders and thighs, which is usually worse in the mornings.

Raynaud's phenomenon – a circulatory problem that causes the blood supply to certain parts of the body to be greatly reduced. It can make the fingers and toes go temporarily cold and numb and they turn white, then blue, then red. Raynaud's phenomenon can also occur with the condition scleroderma.

Rheumatoid arthritis – an inflammatory disease affecting the joints, particularly the lining of the joint. It most commonly starts in the smaller joints in a symmetrical pattern – that is, for example, in both hands or both wrists at once.

Rheumatoid factor – a blood protein produced by a reaction in the immune system. About 80% of people with rheumatoid arthritis test positive for this protein, and it can be present in some

other conditions. However the presence of rheumatoid factor can't definitely confirm the diagnosis.

Sjögren's syndrome – an autoimmune disorder that's characterised by dry eyes and/or a dry mouth, aching and fatigue.

Steroids – drugs that have a very powerful effect on inflammation. The adrenal glands in the body produce a natural supply but much larger doses are used to treat autoimmune diseases. Prednisolone is the most commonly used steroid.

Urea and electrolytes (U&E) test – a blood test that mainly monitors the kidneys. This is frequently carried out to monitor drug treatment.

Where can I find out more?

If you've found this information useful you might be interested in these other titles from our range:

Conditions

- *Giant cell arteritis (temporal arteritis)*
- *Gout*
- *Lupus (SLE)*
- *Polymyalgia rheumatica (PMR)*
- *Raynaud's phenomenon*
- *Rheumatoid arthritis*
- *Sjögren's syndrome*

Self-help and daily living

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Vasculitis

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You can download all of our booklets and leaflets from our website or order them by contacting:

Arthritis Research UK

Copeman House
St Mary's Court
St Mary's Gate, Chesterfield
Derbyshire S41 7TD
Phone: 0300 790 0400
www.arthritisresearchuk.org

Related organisations

The following organisations may be able to provide additional advice and information:

Arthritis Care

Floor 4, Linen Court
10 East Road
London N1 6AD
Phone: 020 7380 6500
Helpline: 0808 800 4050
Email: info@arthritiscare.org.uk
www.arthritiscare.org.uk

Churg–Strauss Syndrome Association (USA)

www.cssassociation.org

PMRGCAUK (Polymyalgia Rheumatica and Giant Cell Arteritis UK)

London WC1N 3XX
Phone: 0300 111 5090
Email: info@pmrgcauk.com
www.pmrcauk.com

National Kidney Federation

The Point, Coach Road
Shireoaks, Worksop
Notts S81 8BW
Helpline: 0845 6010 209
www.kidney.org.uk

Vasculitis Foundation

PO Box 28660
Kansas City
MO 64188-8660
USA
www.vasculitisfoundation.org

Vasculitis UK

John Mills, West Bank House
Winster, Matlock
Derbyshire DE4 2DQ
Helpline: 0300 365 0075
Email: john.mills@vasculitis.org.uk
www.vasculitis.org.uk

Vasculitis UK also produces the Vasculitis Roadmap, which is a guide covering every aspect of the condition. You can download it from www.vasculitis.org.uk/about/routemap

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We're here to help

Arthritis Research UK is the charity leading the fight against arthritis.

We fund scientific and medical research into all types of arthritis and musculoskeletal conditions.

We're working to take the pain away for sufferers with all forms of arthritis and helping people to remain active. We'll do this by funding high-quality research, providing information and campaigning.

Everything we do is underpinned by research.

We publish over 60 information booklets which help people affected by arthritis to understand more about the condition, its treatment, therapies and how to help themselves.

We also produce a range of separate leaflets on many of the drugs used for arthritis and related conditions. We recommend that you read the relevant leaflet for more detailed information about your medication.

Please also let us know if you'd like to receive an email alert for our quarterly online magazine, *Arthritis Today*, which keeps you up to date with current research and education news, highlighting key projects that we're

funding and giving insight into the latest treatment and self-help available.

We often feature case studies and have regular columns for questions and answers, as well as readers' hints and tips for managing arthritis.

Tell us what you think

Please send your views to:

**bookletfeedback@
arthritisresearchuk.org**

or write to us at:

Arthritis Research UK, Copeman House, St Mary's Court, St Mary's Gate, Chesterfield, Derbyshire S41 7TD

A team of people contributed to this booklet. The original text was written by Dr Richard Watts, who has expertise in the subject. It was assessed at draft stage by rheumatology nursing lecturer Dr Janice Mooney, rheumatology nurse practitioner Karina Blunn and chairman of Vasculitis UK John Mills. An **Arthritis Research UK** editor revised the text to make it easy to understand. An **Arthritis Research UK** medical advisor, Dr Luke Gompels, is responsible for the content overall.

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Arthritis Research UK

Copeman House
St Mary's Court
St Mary's Gate, Chesterfield
Derbyshire S41 7TD

Tel 0300 790 0400

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